# NeuroRehabilitation

## A Multidisciplinary Approach

**2nd Edition**


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## A Multidisciplinary Approach

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This book is basically a compilation of information / literature on the available on the topic, from various sources (which have been acknowledged duly). However, this is by no means an exhaustive resource, since the field is evolving at a very rapid pace. Every effort is made to ensure accuracy of material, but the publisher, printer and author will not be held responsible for any inadvertent error(s).

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*This book is dedicated to all rehabilitative therapists, physicians, care- takers, institute, rehabilitative centers, NGOs, societies who have devoted their lives looking after the neurological and musculoskeletal disease and injuries.*

# Acknowledgement

Publishing a book of this nature could not have materialized had it not been from the valuable contribution of all the authors who have spent their valuable time and energy. Most of them are from the staff of NeuroGen Brain & Spine Institute and the visiting consultants from various disciplines. So rightly we can say that it is an effort of a multi disciplinary team. We acknowledge our sincere appreciation to all of them. We also thank all our juniors and family members who have also extended a helping hand in whatever way they could.

**Dr. V. C. Jacob**
**Dr. Hema Biju**
**Dr. Alok Sharma**

# Preface to the First Edition

**NeuroRehabilitation - A Renaissance**

This book (which is the first of its kind published from India) has been written for the following purposes:-

1] To provide in one source a comprehensive overview of all aspects of NeuroRehabilitation

2] To provide both undergraduate and postgraduate students of physiotherapy and occupational therapy with all the information they will need from this field for their examinations.

3] To make available to our younger therapists the knowledge and wisdom of years of experience of the senior authors and contributors of this book.

4] To stimulate younger rehabilitation therapists to take up NeuroRehabilitation as a career choice.

5] To emphasize the importance of a multidisciplinary team approach in NeuroRehabilitation.

6] To introduce students, teachers, junior and senior practicing rehabilitation therapists to the fascinating possibilities of clinical improvements that can occur using Neuro-Regenerative-Rehabilitation-Therapy (which is a combination of Stem cell therapy with NeuroRehabilitation)

Of all the injuries and diseases that have afflicted mankind, those of the nervous system have proved to be the most difficult to treat. Whereas modern medical advances have significantly impacted the survival from infectious diseases and improved the longevity from life threatening cardiological and other critical illnesses, the management of neurological problems has not been as successful. There are still many neurological conditions where no drugs were available and where there are no further neurosurgical options. In such cases the only options for the patients to improve the quality of their lives was rehabilitation. But even in rehabilitation of these conditions, things were not so simple. Having to deal with patients who had no movements or limb strength when combined with other aspects such as spasticity, contractures, loss of sensations, cognitive impairment etc made the rehabilitation process very difficult. The work required to rehabilitate these patients was very physically intensive, requiring many hours of laborious work over long periods of time. The results after all this hard work were most often not upto the patients and their relatives satisfaction since what the patients would want is to start walking normally again and this was in most cases just not possible. The subsequent reluctance of patients to pay for the services appropriate to the efforts that were put in became another factor.

All these facts (the complicated nature of the rehabilitation, the poor response, the lack of willingness to pay) resulted in most younger therapists opting for the less physically demanding and more financially rewarding other options in rehabilitation that were also available more commonly. NeuroRehabilitation in its original sense was there becoming a dying science and skill. Another limiting aspect of treating this was the artificial barriers we had put up between the different branches of rehabilitation mainly physiotherapy and occupational therapy. We all tended to look at the patients from our specialty point of view and this often times did not let us see the whole picture. This was in a sense unfortunate since this was one

aspect of rehabilitation where a real difference could be made.

All these facts (the complicated nature of the rehabilitation, the poor response, the lack of willingness to pay) resulted in most younger therapists opting for the less physically demanding and more financially rewarding other options in rehabilitation that were also available more commonly. NeuroRehabilitation in its original sense was there becoming a dying science and skill. Another limiting aspect of treating this was the artificial barriers we had put up between the different branches of rehabilitation mainly physiotherapy and occupational therapy. We all tended to look at the patients from our specialty point of view and this often times did not let us see the whole picture. This was in a sense unfortunate since this was one aspect of rehabilitation where a real difference could be made.

However all this is now undergoing a major transformation. The availability of regenerative medicine using healthy cells (such as Stem cells) to replace damaged cells has opened up a entire new world of treatment options giving fresh hope to millions of patients who had given up on any hope for improvement. What Stem cells do is that they initiate a process of repair, replacement and regeneration of the damaged cells by a process of release of various growth factors, increased blood supply and cellular replacement. This sort of reenergizes the nervous and musculoskeletal system. However this is only part of the improvement process. The real functional changes in the patients lives come from rehabilitation. What the Stem cell therapy does is that it makes the rehabilitation process more effective and productive. Improvements not seen earlier just with rehabilitation are now visible. This combination of Regenerative therapy and NeuroRehabilitation has therefore opened up an entire new world of opportunities for rehabilitation therapists. Not only is this work exciting and challenging and on the frontline of modern medicine, it is also very rewarding in all aspects i.e. clinically, academically as well as materially. A entire group of patients who had given up on rehabilitation since they were not getting encouraging improvements are now coming back to rehabilitation. The newer fancier corporate hospitals that are just about introducing stem cell therapy are now seeing rehabilitation in a new way. This is an incredible opportunity. A field that had been forgotten and given up as not being useful or remunerative enough is now being looked upon with a whole new interest. It's a rebirth or a renaissance of sorts. Much like how the older radio had almost died in the 80's and 90's with the advent of television but had a rebirth in the last decade with the introduction of FM radio.

In this book all the different aspects of rehabilitation (physiotherapy, occupational therapy, speech therapy, counseling, clinical and surgical aspects ) have all been put together in individual chapters that are focused on an individual diseases or medical condition. This is intentional and meant to highlight the multidisciplinary approach to NeuroRehabilitation. Various theoretical aspects have been covered along with practical advice and suggestions on management. Extensive literature reviews have been done to incorporate the best of whatever is available from all across the world. Valuable individual information gathered from years of experience have also been incorporated. This is important. To highlight this let me state that at the NeuroGen Brain and spine institute we receive patients regularly from all over the world. These patients have already been to the best rehabilitation centers in the USA and elsewhere. And yet despite this virtually all the patients without any exceptions have one thing to say. " that they have never received the kind of rehabilitation in all the centers they had been to, which they received at NeuroGen". What is the difference between what is offered at NeuroGen and what is offered in the bigger rehabilitation centers all over the world. At NeuroGen there is the

human touch. There are virtually no fancy electronic machines or equipment. Our rehabilitation makes a difference because its more hands on with the rehabilitation therapist working intensely and closely with the patients instead of depending on machines and watching from a distance.

This is work that requires dedication, commitment, hard work and perseverance. The joy of this treatment lies not as much in the money but in the small small functional improvements seen in the patients and their gratefulness and gratitude for the same. There were very few therapists who have dedicated their lives to this work. And these therapists belong to an older generation. It was important that the wisdom and knowledge of their years of experience was passed on to today's younger generation. This is therefore one of the important intentions of this book.

It is fortunate for the readers of this book that through this book they will get together in one place the lifetimes wisdom and experience of the senior author of this book Dr. V. C. Jacob. Dr. Jacob, who as you all well know, is one of the countries senior most and highly respected physiotherapist who had devoted his whole life to NeuroRehabilitation through his clinical and teaching work at the LTM Medical college and the charitable work at the Paraplegic Foundation. Inputs from the occupational therapy aspects of the work come from Dr. Hema Biju who has worked for several years in the USA before coming back to India. All the other contributors and authors are experts in their respective fields and we are grateful to their contributions which have resulted in this unique collection of knowledge and wisdom. On behalf of the Neurogen Brain and Spine Institute, I take this opportunity to thank all the authors and contributors. Grateful acknowledgement also goes out to all the members of the NeuroGen team who have despite great difficulties helped and supported all the work that had to be done to make this book a reality. The biggest thanks of course goes to Dr. Nancy Thomas, the scientific and editorial coordinator, who had worked tirelessly for several months to put all this wonderful information together. This book could not have been put together without her sincere efforts. All of us from Neurogen as well as all the contributors now place this book in your hands with the hope that it will inform, enrich an contribute to your lives and the lives of your patients in some important way. Should any of you need any more information please feel free to reach out to us. We are always more than happy to help any of you in any way that your desire. Wish you all students as well a practicing therapists happy reading and happy learning.

# Preface to the Second Edition

It's been 9 years since the release of First Edition of India's first text book on Neurorehabilitation. The last decade has seen a dramatic change in the quality and extent of rehabilitation and newer medical treatments for those suffering from neurological and musculoskeletal disabilities. There is a shift in the wind as we have navigated from a state of hopelessness to one of hope. No more do we have to tell parents of special needs and families suffering from neurological disorders that nothing can be done for you. This shift is especially visible in the thinking process, attitude and treatment approaches of our younger generation of rehabilitation therapists for whom this book has primarily been written. This generation is smart and aggressive and has a hunger for knowledge that will help them improve their professional services. Modern technology has also come to the help of rehabilitation therapists, the availability of stem cell therapy which by improving the functioning of damage nervous system enhances the result of hard work of our therapists. It would not be an understatement to say that availability of stem cell therapy and regenerative therapy has been the single biggest force multiplier in achieving enhanced results from rehabilitation therapies. Other technological advances such as robotic therapy, biofeedback, virtual reality, google glass, motion sensor games, digital aid, gait labs, advanced assistive technologies etc., have also played a major role in the improved results of the present-day rehabilitation. Another major shift that has occurred over the last few years is the multidisciplinary approach to rehabilitation. Whereas, earlier decades had seen clear and separate demarcation amongst the different branches of rehabilitation, today in most of rehab facilities the work is so integrated that it is difficult to make out which therapist is from which branch of rehabilitation. This integrated approach not only benefits the patients, but it is also beneficial to the therapist who learn from each other's thought processes and techniques. Newer training programs such as the fellowship program in advanced occupational therapy in neurorehabilitation, NDTA certification, SI certification, aquatic therapy, reflex integration, ergonomics, etc. are offering opportunities for skill development in the field of neurorehabilitation.

This book has been put together with painstaking efforts of the rehabilitation therapists of Neurogen Brain and Spine Institute. In particular, I would like to thank Dr. Amruta Paranjape and Dr. Khushboo Bhagwanani for all their efforts in bringing out this second edition. We hope that for the undergraduate and post graduate students of various rehab specialties, this book provides adequate information for their studies and exam purposes. For our practicing rehab specialists, we hope that this book provides them updated and comprehensive information to help them in their practice. We hope all the readers enjoy and benefit from this book as much as we have enjoyed putting together all the information.

**Dr. Alok Sharma**
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# About Authors



**Dr. V. C. Jacob,** *B.Sc. D.P.T, MIAP.*

Dr. V.C. Jacob (PT) has been the Deputy Director of NeuroGen Brain and Spine Institute since its inception and is currently the Head of Neurorehabilitation at NeuroGen. He is a Trustee of Nina Foundation which is an NGO set up for the rehabilitation of spinal cord injury persons. He has been a physiotherapist at LTMG Hospital, Sion, Mumbai for 35 years and is now retired. For a period of 38 years, from 1968 to 2006 he was part of the Executive Committee of the Paraplegic Foundation, a charitable organization for the comprehensive care of the paraplegics. He has also organized several sports events for these paraplegics on a national level. In addition, he has also served as President of the Indian Association of Physiotherapists in the past. Apart from all these activities, Dr. Jacob has been involved in disaster management during calamities such as the catastrophic earthquake at Latur and also in Gujarat. He is wholeheartedly committed to the cause of Rehabilitation of patients suffering from neurological disorders.



**Dr. Hema Biju,** *M.Oth (Neuro), OTR (U.S.A)*

Dr. Hema Biju Niravethu is one of the core members that came together under Dr. Alok Sharma when Neurogen was initially conceptualised. With over 25 years of national and international work experience , she has been instrumental in implementing several early practices that have helped in laying the foundation of Neurogen as a pioneer in Research and Development in Neuro-Rehabilitation and Stem Cells.

While leading the Department of OT at Neurogen, she has also authored three books and also jointly published over 20 articles in peer reviewed journals and presented papers at numerous forums related to neurorehabilitation.

After completing her Bachelors in India in 1994, she cleared her OTR (USA) in 1996 and after returning to India, her M.O.Th in 2003. She has since completed her FAOT : NR (2019) and numerous courses in diverse topics of occupational therapy such as office ergonomics, aquatheraphy, hand therapy etc. She remains dedicated to her chosen field of study and enjoys mentoring the future generations of Occupational Therapists



**Dr. Alok Sharma,** MS, MCh

Dr. Alok Sharma is a Neurosurgeon and is presently the [a] Professor & Head of Department of Neurosurgery at the LTMG Hospital & LTM Medical College in Sion, Mumbai as well as the [b] Director of the NeuroGen Brain & Spine Institute in Navi Mumbai and [c] Consultant Neurosurgeon at the Fortis Hospital in Mulund. He completed his graduate and post graduate studies from the Seth G.S. Medical College and KEM Hospital of Mumbai University. Subsequently he did Trained Overseas in Sweden, England, Germany, USA, & Japan. He has written/edited 20 books, has over 150 scientific publications in medical journals and has made over 200 scientific presentations in India and overseas. He has conducted several national and international trials and has been conferred with numerous honors and awards in his distinguished career. He has been committed to both basic as well as clinical research in attempting to

find an answer to the problems of paralysis and neurological deficits that occur following injury and diseases of the nervous system. He has a special interest in the neurobiology of the child's brain and has extensively researched the effects of autism and cerebral palsy on brain function. He has dedicated his life to doing research on incurable disease and injury of the brain and spine and to providing the latest and most advanced treatments for patients who are paralyzed and disabled from incurable neurological conditions. Is the pioneer of Stem cell therapy in India.

He setup the Stem cell and Genetic research laboratory at the LTMG hospital & LTM Medical College which was the first of its type in Mumbai. He has also created India's first dedicated Stem Cell Therapy and Neurorehabilitation center in Navi Mumbai. He is a staunch believer that Stem cell therapy can relieve a lot of human suffering of neurological patients and makes every attempt to popularize this new approach amongst the medical community. He is one of the very few people in the world who does psychosurgery which is a form of brain surgery for people with intractable psychiatric problems. He is also done pioneering work in various forms of minimally invasive Neurosurgery such as Stereotactic surgery and Neuroendoscopy.

He has done innovative work in the field of spinal surgery and Neurotrauma and has a special interest in Revascularization Microvascular surgery for cerebral ischemia. He has a special personal interest in Neurobiology of the mind and lectures extensively on the relationship of the brain/ mind to health and disease. He is a strong advocate of the role of yoga, meditation, natural therapy and diet in the prevention of various modern illnesses. In summary: He is a Neurosurgeon, Medical Teacher and Scientist who is attempting to combine the best of science, medicine and humanity to make a difference to the lives of people who are suffering from neurological disorders.

From the early years of his carrier he has believed in to working towards combining the value of comprehensive rehabilitation with surgical and medical practice. In the 1990's he worked at paraplegic foundation at the LTMG hospital which was one of the rehabilitation center for spinal inury patients. He also started a dedicated center for rehabilitation of head injury practice in the OT department of LTMG hospital.

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Dr. Nandini Gokulchandran is an M.D. in medical Microbiology, presently working in the capacity of Deputy Director & Head of Medical Services at Neurogen Brain and Spine Institute. She did 4 years of post graduate fellowship in the field of Developmental Neurobiology and stem cell research at Tata Institute of Fundamental Research, Mumbai. She brings to NeuroGen Brain and Spine Institute an astute amalgamation of medical/clinical background with deep faith in and understanding of stem cell research & regenerative medicine, especially in neurodevelopment disorder such as autism, cerebral palsy & mental retardation. She has to her credit 97 scientific publication in international peer review journals (1st Author publication in reputed Science magazine as well as 7 papers on autism). She has also co-authored 18 books on stem cell therapy and neurological disorders.



**Dr. Hemangi Sane,** *M.D. (Internal Medicine, USA) (Medical Physician)*

Dr.Hemangi Sane is the Deputy director and Head of the research and development department & Consultant Physician at Neurogen Brain and Spine Institute, Navi Mumbai.

Dr.Sane was practicing Internal Medicine Physician after post-graduation with MD degree from the New York Medical college. She has acquired MBBS graduation from Seth G.S Medical College, KEM hospital Mumbai University. With 20 years of clinical experience, she has more than 90 scientific peer reviewed publications in the field of stem cell therapy for neurological disorders. Some of these are viewed as breakthrough medical articles. She has co-authored more than 12 medical books multiple chapters in International books. She has been recognized as one of the "Leading Physicians of the world" by the International Associations of Internists, USA. She was awarded at the International Women day by the Mayor of Mumbai & as "Iconic Women creating a better world for All" by World Economic Forum. She is an American Board of Internal Medicine certified physician. She is a member of Indian Medical Association, American Medical Association, American College of Physicians and Indian Society of Regenerative Sciences.

She is also the founder of Asha Ek Hope foundation for ALS/MND, India. Her unique, broad and sensitive outlook comes from her experience as a clinician, researcher and as an ALS patient herself. She is dedicated towards finding a cure for neurological disorders, through research. Her positivity, compassion and enthusiasm are an added asset. She envisions independent and better quality of life for patients with neurological conditions.



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Dr. Vishal A Chafale is a Consultant and Interventional Neurologist, practising in Navi Mumbai. Dr. Chafale has completed his MD (Medicine) from SS Medical College Rewa. Post MD he has done his super specialisation in Neurology from Bangur Institute of Neurosciences Kolkata and DNB Neurology from National Board of Examinations. During his tenure in Kolkata he developed special interest in vascular Neurology and to pursue it he did his advanced fellowship in Interventional Neurology SNVI Mumbai. Dr Vishal has 5 years of clinical experience as a Neurologist. He has peer reviewed various publications in the field of Neurology and Vascular Neurology. He has many articles published in indexed Journals.



**Dr. Joji Joseph**, *Head of Physiotherapy Department, NeuroGen Brain and Spine Institute, Navi Mumbai*

Short Profile: Dr. Joji Joseph (PT) heads the department of Adult Physiotherapy and Spinal Cord Injury Walking track unit at Neurogen. He is highly skilled and compassionate professional with more than 20 years of experience in rehabilitation of patients with Neurological Disorders. For a period of over 10 years, he was also an integral part of the rehabilitation team in "Paraplegic Foundation- A charitable organization for the comprehensive care of the paraplegics". He is patient centric and result driven individual with excellent interpersonal and motivational skills from assessing patients condition and developing treatment plan. His experience has continuously helped patients to become functionally independent to the greater extent possible. He also has a number of publications on Neuro-rehabilitation to his credit.



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Dr. Amruta A. Paranjape (PT) is Head of the department, Aquatic Therapy and Chief Scientific Officer at NeuroGen Brain and Spine institute, Navi Mumbai. She has completed MSc in Applying physiotherapy from Sheffield Hallam University, UK; PG certificate in cardiovascular rehabilitation (UK) and is a certified aquatic therapy practitioner (Water specific therapy, Bad Ragaz and Watsu) practitioner from Switzerland. She has more than 35 scientific peer reviewed publications to her name in the field of regerative medicine and rehabilitation. She has authored a guidebook for patients with spinal cord injury and co-authored 4 books on various other neurological disorders. She has conducted training workshops for parents and therapists in treatment of neurological disorders with Aquatic therapy in India, Kenya and Uganda. She is currently also the joint secretary of Aquatic Therapy Network of India.



**Dr. Khushboo Bhagwanani,** *C/NDT(Peads), SI (USC/WPS) (Physiotherapist), Head of Pediatric Physiotherapy Department & Chief Rehabilitation Officer*

Dr. Khushboo Bhagwanani (PT) is a chief rehabilitation officer and head of pediatric physiotherapy department at NeuroGen Brain and Spine Institute. She has completed her advanced certification in Neurodevelopment therapy and Sensory Integration and has 10 years of clinical interest and expertise in the field of pediatric neuro-physiotherapy and her main expertise is treating neurodevelopmental conditions such as autism, cerebral palsy & others, she is also co-author for "Parent Guide Book for Cerebral Palsy" & also for "Parent and patient guidebook" on muscular dystrophy 1st & 2nd edition, Looking after Children with Autism a handbook 1st & 2nd Edition, A Handbook on Neurological Disorders for Special Educator and has 11 scientific publications to her name about in the field of stem cell therapy.



**Dr. Myola D'sa,** *Deputy Head- Pediatric Occupational therapy*

Dr. Myola D'sa , is an Occupational Therapist and has a varied experience in the pediatrics for the last 9 years. She has worked with kids with various developmental disorders, Autism spectrum disorders, Learning disabilities, intellectual disabilities, cerebral palsy, etc. Currently she is working as an occupational therapist at Neurogen Brain and Spine institute. She is certified in Therapeutic Listening, Brain Gym in special needs and is training in other programs like Rhythmic Movement Training (RMT) and The Masgutova Neurosensorimotor Reflex Integration - MNRI® Method. She has co-authored "Parent and Patient guide book" on Muscular dystrophy 1st edition and contributed in the "NeuroRehabilitation – A MultiDisciplinary approach".



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Vishal Ganar is a certified Rehabilitation Psychologist and Counsellor graduated form reputed organisation. He has immensely contributed in the field of disability rehabilitation. Acting Vice President for State Psychologist Association, Maharashtra. And also directing NGO, Bhumika foundation for adults with spl needs, where he train independent living skills and vocational skills to rehabilitate adults with Autism and intellectual disability. He has been honoured with the State Award from Govt. of Maharashtra and also the National excellence award from Rajasthan for his contribution for his contribution in Disability Rehabilitation He has implemented TQR model that has been accepted world wide and has been appreciated at various national and international platform's.



**Nikita D Khokhani,** *BASLP , Post graduate - SLC,*

Ms. Nikita Khokhani is a Speech Language Pathologist at Neurogen Brain Spine Institute Navi Mumbai . She has completed her Bachelors from Manipal University and Post graduation In Advanced Speech and language communication disorder from University of London and has 4-5 years of experience in the field . Her clinical expertise lies in Autism Spectrum disorder with specific oral sensory needs . She has Experience in working with children and teenagers with, RELD, SLI,Apraxia, Hearing impairment, fluency disorders, articulation disorders, Intellectual disability and learning disorders and adults with Aphasia,Dysarthria, Voice Disorders and fluency disorders.

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# Section 1
# General Aspects of Neurorehabilitation

# Ch. 1 International Classification of Functioning, Disability and Health

**Dr. Hema Biju, Dr. V.C. Jacob, Dr. Amruta Paranjape**

The International Classification of Functioning, Disability, and Health (ICF) is a biopsychosocial model for disability. It describes health and health related states to bring standardization of nomenclature and comparability. It involves enlisting of different body structures, functions and activities. Activity, impairment of a function and participation in various tasks shares a non-linear relationship which is different in every individual as it is influenced by personal and environmental factors. There is a prefix for all components followed by two numeric codes. This allows for a clear description of every domain, level of impairment and participation. Although most commonly used in rehabilitation the ICF has multitude applications. In this chapter we aim to outline the structure and use of ICF its advantages and limitations.

ICF evolved from the conflict between medical and social understanding of disease. While medical classifications focused on disease and held the disease processes responsible for the limitation of function, socially the environment and social attitudes were held responsible for such limitation. Therefore, the earlier classification, International Classification of Impairment, Disability and Handicap (ICIDH), could not be applied in both medical and social scenarios. In 1999 World Health Organization (WHO) came up with the conceptual model of ICF. ICF model is by far the best model to suit the needs of both and is currently widely accepted and used, although the adoption of this model was a slow process when it was introduced.

WHO also has a common language for diagnosing disease termed as International Classification of Disease (ICD) but diagnosis alone is not siffucient to determine service needs and level of care required for an individual. Many other factors influence the functional and vocational outcomes post diagnosis. For better policy making it is important to standardized the understanding of these factors and therefore ICF was designed. ICF allows to collect vital data for decision making which is comparable internationally.

ICF focuses on function rather than limitation. It encompasses all the body functions and disabilities ( limitations in performing an activity and participation). It accounts for all the body functions, limitations, activities and environmental factors that restrict participation. It does not account for personal factors.

Schematic representation of ICF model is given in Figure 1

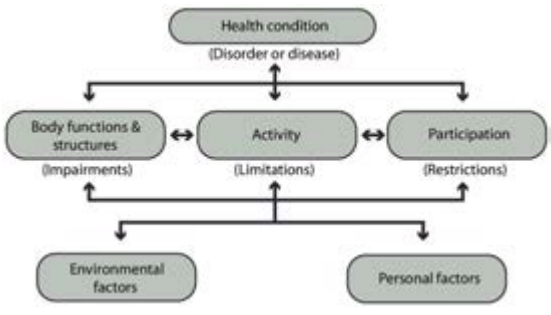



***Figure 1: Schematic representation of the ICF showing various factors that play a role in participation***

**Some terminologies to be understood for the ease of understanding this model are**

1. Body functions, encompass all the physiological and psychological functions
2. Body parts, encompass all the anatomical body structures
3. Impairments, problems with either body function or parts that cause significant deviation of loss of function (Physiological, motor, sensory, cognitive, social etc. )
4. Activity, implementation of body functions and structures to carry out a task.
5. Activity limitations, inability of performing a task and the difficulties encountered in doing so
6. Participation, involvement in life situations
7. Participation restriction, complete or partial inability to involve in life situations
8. Environmental factors, are the physical and social factors that are not in an individuals control
9. Personal factors, include cultural factors, personal attitudes and behaviors, in addition to anatomic, genetic and racial factors
10. While activity and participation describe functional status of an individual, activity limitation and participation restriction describes the disability. These are not coded in the ICF but included in the ICF as they can influence function, activity and participation.

ICF comprises of different domains that have their independent prefixes as follows:

b - body function

s - body structures

d - activities and participation

e - environmental factors.

This is followed by a one-digit code for the chapter number and a further two or three digit or four digit code for level 2, 3 and 4 item.

E.g. b – Body functions

b7 - Neuromusculoskeletal and movement-related functions

b710 – Mobility of joint functions

b7100 – Mobility of a single joint

For describing the severity of difficulty 'qualifiers' are used as suffixes. Without the suffixes only the code in meaningless. The list of domains in ICF becomes a classification when qualifiers are used.

For body function primary qualifier is used to indicate severity. It is a five point scale indicating severity as ( 1 – no problem, 2 – mild problem, 3 – moderate problem, 4 – severe problem, 5 – complete problem )

E.g b7100.2 will mean mild restriction of a single joint

For body structure primary qualifier is used to indicate severity and subsequently secondary qualifier to indicate the change in the respective body structure. Secondary qualifiers are a 9 point scale ( 0 - no change in structure, 1 - total absence, 2 - partial absence, 3 - additional part, 4 - aberrant dimensions, 5 – discontinuity, 6 - deviating position, 7 - qualitative changes in structure, including accumulation of fluid, 8 - not specified, 9 - not applicable )

E.g:

s750 – means structure involved is lower extremity

s750.4 – severe impairment of lower extremity

s750.42 – severe impairment with partial

absence of lower extremity

In the case of the Activity and Participation list of domains, two qualifiers are provided. The Performance qualifier describes what an individual does in his or her current environment. The Capacity qualifier describes an individual's ability to execute a task or an action. This construct indicates the highest probable level of functioning of a person in a given domain at a given moment. Two more qualifiers can be added based on assistance required and optimization of the environment. This allows one to identify what patients do in their current environment using assistance (first qualifier), what they would do in a standard environment (second qualifier), what they could do in an optimized environment (third qualifier), and what they can do in their current environment without assistance (fourth qualifier).

Environmental qualifiers use the 5 point scale for intensity same as body function and a positive sign to denote that the environment is facilitating or a negative sign to denote that the environment is hindering the activity and participation. This is specifically designed to accommodate the needs of all the individuals. An environment that facilitates one individual can become a barrier for another.

ICF is a significant step forward for bringing a common language for all rehabilitation professionals. It helps to describe the disability experienced by individual patient. It has not been widely accepted for reporting and documenting worldwide. It is important to adapt this system in documentation so there is a common language between all. It has a potential to be used far more widely, including in education of both professionals and patients, government policy making, occupational medicine etc. It can be tedious to understand this model and implement it and it doesn't account for personal and cultural factors; these can be the reasons for ICF not being adopted as widely as envisaged. However, the awareness is increasing and it will be used more frequently and uniformly in future.

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# Ch. 2 Pathophysiological Consequences of Injury to Nervous System

**Dr. Vishal Chafale, Dr. Hemangi Sane, Dr. Amruta Paranjape**

Injury or ischemia of nervous system causes two folds damage to nervous tissues. Primary injury is the damage caused by the trauma, ischemia or hypoxia; this leads to neuronal cell death in the area surrounding the impact site. This is followed by cascade of biochemical reactions that lead to inflammation, disruption of blood brain barrier, modulated immune response causing variety of micropathologies like diffuse axonal and neuronal injury and altered microvasculature. These changes are termed as secondary injury. There are different mechanisms at play in Traumatic injury, Ischemic or hypoxic injury, Infection and Inflammation. This chapter elucidates these for a better understanding of the pathophysiological processes.

**Traumatic Brain Injury**

Nervous tissue is protected from trauma by skull, vertebrae, meninges and cerebrospinal fluid; however, when the physical trauma exceeds the physical and functional thresholds it causes Traumatic brain injury or Traumatic spinal cord injury. Neural cells cannot tolerate mechanical forces and deformations like other types of cells. After the initial mechanical insult, secondary injury is caused by inflammation, altered cellular signaling, glutamatergic excitotoxicity, calcium overload, cerebral edema and vascular dysfunction. These mechanisms also trigger mechanisms that activate cell death pathways. Progressive neurodegeneration causing loss of motor, sensory and cognitive functions is observed following traumatic brain injury due to cell death and damage. While contusion and hemorrhage follow a focal injury, which is easier to detect on the radiological investigations; diffuse injury is followed by widespread axonal and neuronal death which is difficult to distinguish on radiological investigations. Neurons exhibit differential susceptibility to damage induced by stress. Some of the factors that determine the susceptibility of neurons to damage are myelination, vasculature, intensity and frequency of trauma. Cellular changes observed acutely after physical trauma include proteolysis, apoptosis, necrosis, disruption of blood brain barrier and loss of synaptic terminals. There is mitochondrial damage to the cells and loss of ionic homeostasis. Some of the mechanisms causing this are calcium influx and indiscriminate release of neurotransmitters typically glutamate. Following the acute response altered vascularity and immune response contribute to subacute and chronic changes.

Some of the vascular insults seen in traumatic brain injury and hemorrhage, edema, vasospasms, fluctuations in cerebral blood flow and altered blood brain barrier. Hemorrhage or the bleeding can be progressive and may continue up to 24 – 48 hours post injury. There can be formation of hematomas. Based on their location they are classified into 3 types Epidural (Between skull and dura matter), Subdural (between dura matter and arachnoid matter) and Subarachnoid (between arachnoid and pia matter) (Figure 2.1).

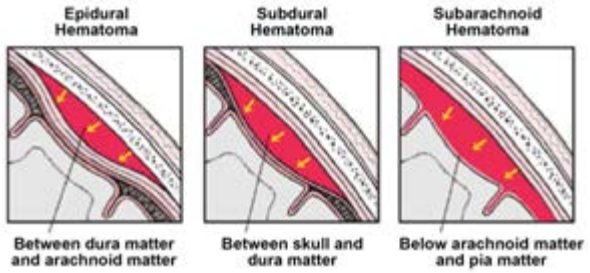



***Figure 2.1 Image showing different types of hemorrhages***

Hematomas usually resolve on their own but microbleeds can persist rendering hemosiderin deposits that can be detected on MRI as black or hypo-intense lesion, signal voids. Hematomas are known to cause grey matter damage, but micro bleeds may affect white matter as well. Hematomas can be considered as direct effect of the injury whereas other vascular dysfunctions are indirect mechanisms.

Acutely after the injury blood brain barrier is disrupted. BBB is a physiological neurovascular unit comprised of anatomical units like endothelial cells, neurons, astrocytes and pericytes. The said barrier is maintained due to tight junctions in the endothelial cells that do not allow certain molecules to permeate in to the neuronal environment. Ionic imbalances, apoptosis, proteolysis, hyperfibrinolysis and coagulation cascade dysfunction disrupts and destroys these tight junction proteins therefore increasing the permeability of the blood brain barrier. Allowing pro-inflammatory molecules to permeate in neuronal microenvironment causing edema. Inflammation hinders normalization of BBB. It may take several weeks, months or even years for the blood brain barrier to be repaired.

Cerebral edema is caused by accumulation excessive fluid extracellularly as well intracellularly. Extracellular edema is caused by fluid accumulation in extracellular spaces due to disruption of blood brain barrier; it usually follows cytotoxic edema. It compresses brain tissue and increases intracranial pressure. Intracellular edema is also known as cytotoxic edema which is caused by ionic imbalances, leads to neuronal apoptosis but does not disrupt blood brain barrier.

Cerebral blood flow fluctuates in acute and subacute phases of the injury. Immediately after the injury there is a rapid decline or cessation of blood flow to the site of the injury. This may lead to reduction beyond the threshold of ischemia; causing secondary ischemic injury. This may be followed of regularization of the blood flow but in some cases increased blood flow or hyperemia. Eventually vasospasm leads to hypoperfusion which remains a leading cause of death in traumatic brain injury. It may take several weeks for the cerebral blood flow to normalize. Vascular and immune responses subside however persistent activation of microglia has shown to cause chronic inflammation and neurodegeneration in even in chronic phases of the injury.

**Ischemic brain injury:**

Ischemia can be defined as a decreased cerebral blood flow that can cause metabolic or functional deficits. Ischemic brain injury can occur post thrombotic or embolic occlusion of one of the arteries of the brain. Ischemic injury results in more focused or localized damaged as compared to traumatic injury or hemorrhagic injury that causes widespread damage. In cerebral ischemia blood flow is completely arrested in a part of the brain, infarct core, tissue in this part dies within minutes. The toxic cellular microenvironment and reduced blood flow causes rapid damage to surrounding tissue. However, the cells in the surrounding cells may not die immediately due collateral perfusion. These cells become dysfunctional but are salvageable and may retain structural integrity. Partially viable tissue surrounding

the core infarct area is known as 'Penumbra'. Figure 2.2 shows penumbral regions in PET-CT scan.

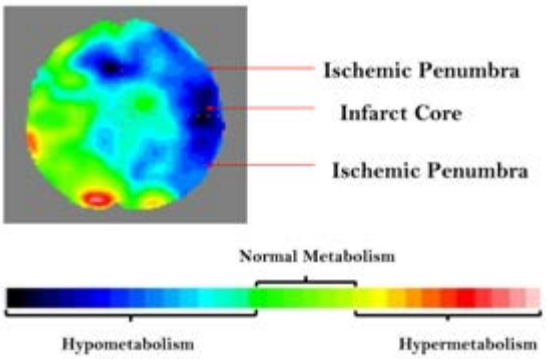



***Figure 2.2 PET-CT scan images showing centrifugally graded tissue damage with black infarct core surrounded by ischemic penumbra in varying shades of blue***

Cell injury and death mechanisms in these two regions show stark differences. Ischemic cascade are the changes that follow ischemia in brain and cause cell apoptosis or necrosis. Having same outcome of cell death, apoptosis and necrosis are two different physiological processes. While infarct core seems to undergo necrosis, penumbral tissue may undergo both but more often apoptosis. Necrotic cell death stimulates paracrine inflammatory response. Apoptotic cell death is a programmed cell death that goes through series of cell shrinking processes and finally phagocytosis preventing cellular debris from entering the microenvironment and eliciting an inflammatory response. Leading pathogenic mechanisms that constitute ischemic cascade are energy failure, increased intracellular calcium levels, excitotoxicity, free radical toxicity, disruption of blood brain barrier, inflammation and finally apoptosis or necrosis.

Brain requires continuous oxidative phosphorylation of to generate ATPs for functioning. It does not have the ability to store energy or substrates and is dependent on a continuous supply of Oxygen and Glucose. Immediately when the blood flow reduces these sources are depleted creating an energy deficit and accumulation of lactic acid due to anaerobic glycolysis. Acidosis hinder intracellular protein synthesis by producing excess free radicals. Energy failure also cause ionic dyshomeostasis leading to calcium ion influx and cytotoxic edema. Calcium ion dyshomeostasis is particularly detrimental to the tissue as Calcium ions can activate variety of enzymes like protein kinase C, phospholipase A2, phospholipase C, cyclooxygenase, calcium - dependent nitric oxide synthase, calpain, proteases and endonucleases; that trigger inflammation, necrosis and apoptosis. which may be Energy depletion causes rapid depolarization of neurons and excessive release of neurotransmitters especially Glutamate. Apart from Glutamate toxicity this causes further increase in intracellular calcium ion, sodium ion and chloride ion concentration worsening the edema. This is also known as excitotoxicity, which can cause necrosis but also stimulate apoptotic processes. Following the cytogenic edema vasogenic edema is caused by blood brain barrier disruption caused to destruction of basal lamina by matrix-metalloproteinases, damaged induced by inflammatory molecules and free oxygen radicals and hypoxic and mechanical damage to endothelium. This amplifies inflammatory response. Inflammatory response is seen with minutes of blood flow occlusion due upregulation of inflammatory mediators such as platelet-activating factor, tumor necrosis factor α, and interleukin 1 β. Ischemic injury triggers inflammatory cascades in the brain parenchyma that may further amplify tissue damage by many mechanisms. Within minutes of occlusion, there occurs upregulation of proinflammatory genes which produces mediators of inflammation such as cytokines, nitric oxide, superoxide and prostanoids. Neutrophils are found to infiltrate first

followed by microglial activation and infiltration of Macrophages and Monocytes after several hours. All of these processes eventually cause necrosis or apoptosis.

**Recovery of neural tissue post traumatic and ischemic injury**

Recovery post stroke is lead by various mechanisms that bring about neuroplasticity. Neuroplasticity is defined as the ability of the nervous system to change its activity in response to intrinsic or extrinsic stimuli by reorganizing its structure, functions, or connections3. This is observed in brain as well as spinal cord injury. Neuroplasticity contributes towards functional recovery after an insult. There are short-term and long-term mechanisms with which this can be achieved. Short term mechanisms are activation of alternate brain pathways and unmasking of silent pathways. Whereas long-term mechanisms axonal and dendritic sprouting of the surviving neurons, synaptic pruning and neoangiogenesis. This leads to physiological restitution of penumbral area, resolution of diaschisis and functional substitution and compensation.

The acute recovery from ischemia is crucial to recovery post stroke. It is found that assessments on day 7 post stroke are the strongest predictors of final recovery4. Degree of vascular insult, extent and location of anatomic damage and inflammatory response post insult can influence the recovery. Understanding the natural history of recovery is important to providing appropriate rehabilitative inputs. Initial recovery in the acute phase is unlikely to be due to large-scale restructuring. Axonal growth and development of new synapses takes several days and therefore rehabilitation in sub-acute phase is of importance. Knowing the exact mechanisms of neuroplasticity at different time points in recovery can aid to provide accurate rehabilitation targeting these mechanisms. Some of the principles of recovery are 1] The recovery is not linear with time, meaning, the speed of recovery varies with time. It is fastest in the first 30 days and then slowly declines at varying speeds 2] Outcome in the chronic phase dependent on the severity of initial insult.

Figure 2.2 shows a hypothetical curve of recovery after ischemic stroke

**Neurogenesis post injury**

It was only recently established that neurogenesis is possible even after birth and to a minor extent even in adult brain. The process of neurogenesis can be reactivated in response to an insult. Subgranular zone of dentate gyrus, subventricular zone of some cortical areas, substantia nigra are usual sites of neurogenesis; after an insult peri-infarct areas also can contribute to neurogenesis. Some of the processes of neurogenesis like differentiation, migration of neural precursor cells and axon sprouting are independent of physical activity and occur irrespectively in response to injury. These processes are thought to be guided along the blood vessels, these vessels provide a structural scaffold for neural networks. Therefore, neurogenesis and angiogenesis go hand in hand. Enhanced neurogenesis post stroke is observed in areas with greater density of blood vessels. Synaptic pruning and plasticity are activity dependent and can be influenced by means of physical exercise. Various molecular mechanisms that can enhance neurogenesis are increased production of anti-inflammatory cytokines, secretion of neurotrophic factors like Brain Derived Neurotrophic Factor (BDNF), Glial Cell Derived Neurotrophic Factor (GDNF), Vascular endothelial growth factor (VEGF), Epidermal growth factor (EGF), erythropoietin and fibroblast growth factor-2 (FGF-2). Apart from this nitric oxide, neurotransmitters like glutamate, serotonin and acetyl cholin may also influence the process of neurogenesis.

Neurogenesis is need based and therefore the areas that are trained more may show enhanced neurogenesis. Rehabilitation therefore plays a crucial role in facilitating neurogenesis and enhancing neuronal plasticity. Neurogenesis is negatively affected by various other factors which are stress, chronic alcoholism, depression, substance abuse, irradiation, high-fat diet, diabetes and chronic inflammation. Exercise, learning new skills, enriched environment, small calorie have a positive influence of neurogenesis.

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# Ch. 3 Multidisciplinary Team Approach

**Dr. V. C. Jacob, Dr. Joji Joseph, Dr. Hema Biju, Dr. Amruta Paranjape, Dr. Ritu Varghese, Dr. Khushboo Bhagwanani, Mr. Vishal Ganar**

**Introduction**

People with prolonged disability, usually benefit from rehabilitation to improve their abilities or to adapt. These kinds of services are provided with the help of rehabilitation. Rehabilitation is focused on improving the functional activity that is limited in people with disability. Rehabilitation goals are to optimize participation in social activities and to curtail distress and discomfort. Therefore, the process of neurological rehabilitation is important in all healthcare professionals.

This chapter discusses about how the services should be organized and what is the process of rehabilitation. It discusses that having access to a specialist team using an interdisciplinary approach is essential for all patients, even people with relatively straightforward problems. Interdisciplinary approach largely benefits the person with neurological disorder.

**Key features of rehabilitation are**

- It is mainly a process, not a single or limited set of treatments.
- It focuses on disability and function, not disease.
- It takes universal view, taking into account and actively considering all influences on a patient's situation, rather than considering such influences as not relevant.
- Therefore it necessarily often involves a wide range of different:
  - Professionals and non-professional people
  - Agencies and organizations

There is difficulty for two reasons in neurological and neuromuscular disorders. The main patient-related processes in rehabilitation are adaptation and learning, and it is the nervous system that is required for these recovery processes. Therefore, the process must be adapted to the patient's cognitive ability, which will often be limited by the disease.

At the same time, because the nervous system is central to almost all human skills and activities, the range of loss is great and that usually includes a perceived or actual change in a person's identity. Thus, the skills and knowledge need to cover a very wide range.

Rehabilitation includes accurate analysis of the presenting problems (diagnosis) and then undertaking targeted interventions aimed at reversing or ameliorating identified problems (treatment) will lead to desired success. The focus of the neurological and neuro surgical services is on the underlying damage to the disease, and of rehabilitation is to improve the functional activity and improve the disability status.

This chapter outlines, the benefits of multidisciplinary and intradisciplinary approach and wide range of expertise that is needed. The team should be working towards achieving a common goal, and this emphasizes the central importance of goal setting when faced with complex problems.

**The biopsychosocial model of illness**

In 1977, Engel wrote an influential paper, upon sociological and other research to formalize a broader approach to understanding the illness, together with the idea of Talcott Parsons which was published In the year 1953. He came up with biopsychosocial model of illness which was later used by world health organization (WHO) as the basis for the International Classification of Impairments, Disabilities and Handicaps (WHO ICIDH, 1980) and then the improved International Classification of Functioning (WHO ICF, 2001).

The basic insight is that any illness must first be divided into contextual factors and patient-related factors. The patient-related factors come from one of four hierarchical levels: the organ, the body, the person interacting with their physical, observable environment, and the person's interactions with other people. The contextual factors encompass four concepts: the physical environment, the social environment, the person's own pre-existing characteristics and time, which is subdivided to cover both the person's stage in their life and their stage in their illness. The descriptive framework is also a systems analytic model.

One strength of this model, of relevance to neurologists is that it predicts the existence of functional illness.

**Recovery from loss of function and adapting to change**

Restitution of the tissue occurs initially following an episode of tissue dysfunction. However, when there is residual dysfunction the body and person adapt, a process of learning to achieve goals in a different way. For most internal, physiological functions this is 'automatic' and beyond conscious control. There are exceptions, such as the use of hormone replacement therapy after failure of an organ when the person must learn to take the replacement appropriately, which is not trivial for insulin.

In conditions where there is a gradual and usually progressive loss of tissue function, such as occurs with multiple sclerosis, Huntington's disease, muscular dystrophies, or motor neuron disease, then there is an unavoidable process of adaptation than, in slowly progressive disorders such as Parkinson's disease, be so successful that the patient may lose significant amounts of tissue without noticing it. However, eventually most people will notice problems, especially in fluctuating conditions such as multiple sclerosis.

Finally, there are conditions that arise before, at or shortly after birth, when the person is naturally totally dependent anyway. If the damage is fixed, then the person will learn and develop and will incorporate the consequences of their losses into their normal development which may thus be different from usual. If the person has an additional progressive loss, they will also adapt to their changing abilities.

**Rehabilitation**

Rehabilitation is no more or less than helping the person to adapt and learn in response to their limited, altered, or changing abilities. Theoretically, it is exactly like education, except that it is set in the context of loss or absence of existing or expected abilities arising from a disease or health disorder.

Sometimes, for example when muscles have simply wasted through disuse or after an acute but reversible injury but are still intact, the process is primarily one of encouraging 'natural' recovery and doing so in a safe environment so that, for example, the person does not fall or develop skin pressure ulcers while recovering. More commonly, the process involves identifying and teaching alternative strategies and allowing practice in a safe environment. Also, quite commonly, the process involves helping the person

recognize that some previous goals or activities are no longer achievable and helping them and their family adjust to this.

This approach emphasizes that rehabilitation is not only applicable to people with recently acquired losses set in the context of premorbid 'normality'. It is also appropriate for people who have limitations imposed by some congenital or other problem present from birth and for people who have a progressive disorder. In these circumstances the goals may be different and the underlying illness processes may be different, but rehabilitation services can still help the person adapt, set appropriate goals, which may be greater than those expected by the patient, and learn new skills to meet goals.

The rehabilitation process depends crucially upon an accurate initial analysis of

1. Identifying underlying pathology (disease, disorder), because it may:
   - Determine prognostic field and prognostic markers
   - Suggest impairments that should be looked for, or do not need testing for
   - Impairments present, nature and severity
2. Levels of activity and social participation
3. Personal contextual factors
   - Expectations of rehabilitation and change
   - Goals / domains of interest
   - Attitudes, strengths, weaknesses etc.
4. Physical contextual factors such as:
   - Equipment and adaptations
   - Accommodation
   - Availability of practical support from people (caring, not social)
5. Social contextual factors, such as
   - Attitudes and expectations of family and important others
   - Benefits and other resources available

**Rehabilitation team**

The list given of important components needed to achieve successful (efficient and effective) rehabilitation illustrates why a multidisciplinary team is needed.

For most patients with more complex problems, this detailed and in-depth analysis can only be achieved by a group of professionals who have between them appropriate expertise. Without a clear, accurate initial analysis it is probable that the process may be misdirected, attempting to achieve inappropriate goals or not attempting to achieve appropriate goals.

Patients with neurological conditions are particularly likely to have complex problems. Complexity is, in fact, difficult to define and in this context, it refers to problems that:

- Are multifactorial (i.e. are influenced by many different factors such as cognition, mood change, altered sensation)
- Have interactions between the different factors that themselves influence the outcome of interest (e.g. combination of blindness and amnesia after posterior cerebral circulation ischemia makes rehabilitation of both impairments much more difficult)
- Have non-linear relationships between the different factors
- Usually include clinical uncertainty about the disease and its prognosis.

This complexity arises particularly in neurological rehabilitation because the nervous system is the central system for functioning as a human being, defined in

rehabilitation as a social being who learns and adapts to changing circumstances. The brain controls almost all conscious and much unconscious behavior. It analyses situations, plans action, responds to changes, etc. It also is the basis for personality, emotion and goals. Indeed, most people now equate brain damage with a change in their personhood—who they are.

There are few if conditions that affect the central nervous system that do not cause complex problems. Consequently, most patients with continuing problems associated with any neuro-logical disorder may benefit from expert rehabilitation.

Given the wide range of problems that may arise from neurological dysfunction, it should be obvious that no single person or profession is likely to have the very large range of expertise needed to lead to an accurate analysis of a patient's situation. However, without a full understanding both deficits and of the areas of preserved or good function, it is likely that each individual professional person involved will not set appropriate goals or undertake appropriate interventions.

**Rehabilitation team and importance of team work**

A team is or should be more than a group of people who simply share factual, analytic information. A team works towards a common goal, each member contributing according to their expertise and ability.

Teams are themselves complex systems. Team members should:

- Share knowledge and skills
- Share clinical information about patients continuously
- Be able to undertake tasks usually undertaken by others within the team; the work is shared and therefore if someone is not available the team can continue to function.

In rehabilitation certain aspects of the team keep evolving, the points of discussion for such evolution are:

- The membership of the team
- The type of teamwork used, usually distinguishing between multidisciplinary and interdisciplinary teams and, more recently transdisciplinary practice
- Whether the patient (and family) are a part of the team
- Who should lead the team.

It is self-evident that no team can include every single expert whoever might be needed by the patients. In other words, teams will always need to seek additional knowledge or skills from other professionals in some situations.

Moreover, in a team of any size the actual group of people involved with a particular patient will be a subset of the whole team in the service. Some patients will have no need for some professions; that patient's team is not the same as the whole team. Even with a highly focused service, every therapist cannot be involved with every patient, and often a person or people or team from elsewhere will be involved to a greater or lesser extent.

The overall group of people who are actively and appropriately involved directly or indirectly can be very large and can come from a wide variety of different organizations and can come from a wide variety of professions. Everyone will have his or her own interests, skills, knowledge, experience and expectations. Success depends upon all of the professionals working together and not against each other (usually accidentally).

The potential for miscommunication and

misunderstanding is great, and highlights the overwhelming importance of liaison and communication, which is discussed later.

Different team dynamics is observed between the professionals depending upon the patients' needs. These are often characterized as one of three types:

- **Multidisciplinary;** work undertaken with other disciplines in parallel or sequentially
- **Interdisciplinary;** work undertaken jointly with other disciplines
- **Transdisciplinary;** work integrated across many disciplines and undertaken collaboratively.

The ideal is to have a fully integrated team who all work collaboratively, because this is likely to lead to a more efficient and effective team.

**Role of a multidisciplinary rehabilitation team**

Most multidisciplinary rehabilitation teams hold meetings at regular intervals to discuss on topics such as:

- Short- and long-term goals
- Length of stay
- Patient and family education needs
- Discharge planning

To help reach patient specific needs and goals, rehabilitation programs often include:

- Advice around medication
- Physical therapy and occupational therapy for wellbeing and recovery
- Help with activities of daily living (adls), such as eating, dressing, bathing, toileting, handwriting, cooking, and basic housekeeping
- Developing skills to enable independent living
- Help with speaking, reading, writing or swallowing
- Psychological support and social and behavioral skills retraining
- Patient and family education
- Nutritional counseling
- Safety and home care needs
- Links to community resources and activities.
- Vocational counseling

**Structure and function of a multidisciplinary rehabilitation team**

*A multidisciplinary rehabilitation team often includes:*

- **Neurologist:**

Neurologists are specialized in diagnosing, treating and managing neurological disorders. They may advise the physician managing the patient's overall health.

They may also be involved in educating patients of possible adversities so that they are able to recognize them when they do arise. wherever appropriate, they may identify and communicate with a professional who is well linked with other services

- **Physicians:**

A physician's role is medical evaluation and treatment. The physician also focuses on establishing a long-term medication regimen and appropriating any change in medications as per changing needs of the patient. Wherever needed, they identify the need for communication with other specialists.

- **Nurses:**

Nurses work in collaboration with physicians to ensure timely medication.

They not only provide medical care but also

provide information such as use and care of respirators, feeding, moving patients to avoid contractures, bed sore prevention and care, etc.

- **Physiotherapists**

Physical therapy plays an important role to improve a patient's quality of life through examination, prognosis and physical intervention. Physiotherapy rehabilitation programs aim at reducing risks of complications and at optimizing function. A rehabilitation program is specifically designed for each individual patient. To reduce the risk of secondary complications. Improve physical function and independence to allow for better participation in life functions and social activities.

Therapists use a variety of therapy techniques to optimize function including:

- Therapeutic exercises for mobility and strength
- Splinting, bracing and use of other specialized equipment
- Gait training with use of assistive devices
- Neurological therapeutic techniques for re-education of movement and activity patterns
- Compensatory techniques and strategies

*Re-evaluation and Reviewing of rehabilitation programs*

Rehabilitation in neurological and progressive diseases is never static. A person newly diagnosed have goals that change in due course of the disease. Later, since needs are constantly changing, reassessments and reviewing of the interventions with changes where required may need to be initiated in response to emergent disabilities.

- **Occupational therapists**

Occupational therapy treatments are patient-centered and are planned to meet individual's specific needs and goals.

- Occupational therapists provide assessment, treatment, education and support.
- Therapy is aimed at enhancing independence and quality of life.
- They help in restoring function for problems related to activities of daily living (ADLs) including work, school, family, community and leisure activities.
- They help patients to return to pre-illness employment or considering alternate or altered employment.
- Occupational therapists also assist with splints, braces, wheelchairs and other equipment selection and adaptations and provide training to caregivers to ensure a smooth transition from the rehabilitation setting to home.

- **Aquatic Therapist**

- The aquatic therapy is the use of water and its chemical and physical properties to perform specifically designed activity given by qualified personnel to aid in the restoration, extension, maintenance and quality of function for persons with acute, transient, or chronic disabilities, syndromes or diseases. (Definition given by aquatic therapy network-India)
- Aquatic therapy means making use of various physical and chemical properties of water for the treatment of various disorders.
- Aquatic therapy refers to treatments and exercises performed in water for relaxation, fitness and other therapeutic benefit.

- Aquatic therapy techniques include Ai Chi, Aqua Running, Bad Ragaz Ring Method, Halliwick, Watsu, and other aquatic bodywork forms.
- An Aquatic therapist will commonly focus on improving the physical, cognitive and psychosocial function associated with illness, injury, or disability.

- **Psychologist:**
  - Performs a neuropsychological assessment
  - Assists individuals and their families to come to terms with the effects of the difficulties they may experience
  - Helps identify and build resources and coping mechanisms for dealing with the effects of the disability
  - General psychiatrists are called where neuropsychiatric complications arise.
  - Genetic counselors: Genetic counseling is the process of advising patients or the family that are at risk of inherited disorder, of the nature of the disorder and the option of family planning. They however do not remain indefinitely involved with the team.

**The team may also include the expertise of other professionals like:**

- **Dieticians**
  - They help individuals manage dietary needs and,
  - Promote healthy eating habits and provide education on how to stay healthy.
  - Speech and language therapists:
  - Assess and treat communication and language impairment
  - Assess and treat swallowing difficulties

- **Social workers:**

Neurological disorders being chronic, economic costs related to caregiving need to be considered. Social workers help with obtaining financial aid and finding support groups for patients and the caregivers.

- **Other specialist services**

Rehabilitation of people with chronic disease is supported by specialist teams such as those advising on continence, enteral feeding, tissue viability, and pain.

Thus, individuals and care givers benefit from a multidisciplinary rehabilitation team with availability of a vast range of specialty services. Interventions are most effective and timely with specialists working together as a team.

**Goal setting**

If a team is defined as a group of people working towards a common goal, then the process of identifying, agreeing and setting those goals must be the central, defining process that distinguishes effective teams from less effective teams.

Goal setting increases motivation and engagement. The evidence is very strong that setting goals for individuals and for teams alters behavior; individuals and teams achieve more when they set appropriate goals.

**An effective collaborative rehabilitation team is likely to have the following characteristics:**

- An agreed, shared framework for understanding and analyzing and describing a patient's situation.
- A shared terminology and vocabulary.
- A similar lay-out of clinical notes.
- The use of an agreed set of measures for frequently measured domains.

- A shared primary clinical record where all professions record all major observations.

**Challenges encountered by Multidisciplinary teams:**

- Some professions are reluctant to share some data
- Another challenge is to achieve a comprehensive complete record that can nonetheless be easily searched to find relevant information quickly
- A single geographic area (office or set of offices) used by all team members
- Shared treatment spaces, not 'belonging' to any profession or department
- Multidisciplinary goal-setting meetings on a regular basis for all patients may be difficult to conduct
- Undertake shared educational activities
- Be managed as a single unit, including having a single budget shared by the whole team

Neurological disorders are complex and challenging leading to severe functional deficits. Long term multidisciplinary neurorehabilitation is a goal-directed process that reduces the functional dependence, neurological deficits and secondary complications to provide better quality of life and re-integration in the community. Therefore, despite the challenges it should be encouraged, and teams should evolve to overcome these challenges.

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# Section 2
# Disorders of Spinal cord

# Ch. 4 Traumatic Spinal Cord Injury

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## INTRODUCTION

*"So many of our dreams at first seem impossible, then they seem improbable and when we summon the will they soon become inevitable." – Christopher Reeve*

In Latin, rehabilitare means make fit again.

**Rehabilitation is defined as**

*"A treatment or treatments designed to facilitate the process of recovery from injury, illness or disease to as normal a condition as possible."*

In other words, it is the process of restoration of skills by a person who has had an illness or injury to regain maximum self-sufficiency and function in a normal or as near as normal manner as possible. Having to give up independence and becoming totally dependent on others is a frustrating and debilitating affair for a person with SCI. With a unified team approach, a person with a SCI himself or herself would benefit in gaining not only physical independence but also economic and social acceptance. The relatives and the individual with SCI are as much a part of the team as professionals as eventually they will have to share the responsibility for the success in getting back the person into the society and not becoming a burden to the society.

Rehabilitation can be a good medium for harmful parasitic thoughts of hopelessness, helplessness and worthlessness. To help patients come out of this dreadful situation, it requires effort from a team of professionals, which is rightly called as a multidisciplinary team.

## MULTI DISCIPLINARY TEAM

An ideal multi-disciplinary team should comprise of all the following members:

1.Spinal surgeon

2.Urologist

3.Physiotherapist

4.Occupational therapist

5.Aquatic Therapist

6.Medical social worker

7.Psychological counselor

8.Rehabilitation nurse

9.Nutritionist and dietitian

10.Care givers

However, the team would not be complete without the SCI person himself whose willingness to co- operate with the team members alone would make the entire process easier. Ideally the team members should take collective decisions in setting a goal or outcome measures, which is nothing but a guideline to achieve the expected level of recovery. For this each member must set a realistic achievable goal. The patient's expectation may be too much, so also of the team members. The expectation may be realistic only if one takes into consideration the medical problems and the psychological status of the patient.

It is advisable and ethical to involve the patient in planning the treatment pathway from time to time. However very few patients have the technical knowledge

regarding the management of their condition. Hence the therapists must make efforts to explain the advantages and disadvantages of various approaches. Many times it is beneficial to show other rehabilitated patients who have successfully undergone the rehab process and how they have solved their own problems, thereby living a productive life within their face of limitations. Before taking the final decision, it is mandatory on the part of the therapist to consult other team members and each one of them has to make a thorough evaluation of the motor skills, functional skills, home assessment, ADL, etc. Rehab begins from the day of the injury. All the interventions planned for the patient from day one focuses on prevention of further damage to the spinal cord and to preserve the functional capabilities, most vital to save the patient. So, a brief review of the SCI is discussed below. In the initial phase after the SCI, surgeon plays a vital role especially, to decompress and stabilize the spine so that the actual physical rehabilitation can commence at the earliest. How big is the problem? To put light to the intensity of the SCI issue, the epidemiology is stated below.

**Epidemiology:**

- Before World War 2, the average life expectancy of a SCI person was just 2yrs. With the advent of antibiotics and improved therapeutic measures, the management of complications of SCI has improved remarkably. Acute care has also improved considerably. After the rehabilitation, they learn to live rather independently or with minimal help of the family members.
- The annual incidence of SCI is about 54 cases per million population
- The life expectancy of SCI patients has increased significantly in last decades and is near normal for injuries of lower cervical and thoracic levels

**Causes and risk factors:**

Spinal cord trauma can be caused by any number of injuries to the spine. They can result from motor vehicle accidents, falls, sports injuries (particularly diving into shallow water), industrial accidents, gunshot wounds, assault, and other causes.

A minor injury can cause spinal cord trauma if the spine is weakened (such as from Rheumatoid Arthritis or Osteoporosis) or if the spinal canal protecting the spinal cord has become too narrow (spinal stenosis) due to the normal aging process.

Direct injury, such as cuts, can occur to the spinal cord, particularly if the bones or the disks have been damaged. Fragments of bone (for example, from broken vertebrae) or fragments of metal (such as from a traffic accident or gunshot) can cut or damage the spinal cord.

Direct damage can also occur if the spinal cord is pulled, pressed sideways, or compressed. This may occur if the head, neck, or back are twisted abnormally during an accident or injury.

Bleeding, fluid accumulation, and swelling can occur inside the spinal cord or outside the spinal cord (but within the spinal canal). The accumulation of blood or fluid can compress the spinal cord and damage it.

Most spinal cord trauma happens to young, healthy individuals. Men ages 15 - 35 are most commonly affected. The death rate tends to be higher in young children with spinal injuries.

Risk factors include participating in risky physical activities, not wearing protective gear during work or play, or diving into shallow water.

Older people with weakened spines (from osteoporosis) may be more likely to have a spinal cord injury. Patients who have other medical problems that make them prone to

falling from weakness or clumsiness (from stroke, for example) may also be more susceptible.

**Symptoms:**

Symptoms vary somewhat depending on the location of the injury. Spinal cord injury causes weakness and sensory loss at and below the point of the injury. The severity of symptoms depends on whether the entire cord is severely injured (complete) or only partially injured (incomplete).

The spinal cord doesn't go below the 1st lumbar vertebra, so injuries at and below this level do not cause spinal cord injury. However, they may cause "cauda equina syndrome" -- injury to the nerve roots in this area.

**1. CERVICAL LEVEL INJURIES**

When spinal cord injuries occur in the neck area, symptoms can affect the arms, legs, and middle of the body. The symptoms may occur on one or both sides of the body. Symptoms can include:

- Breathing difficulties (from paralysis of the breathing muscles, if the injury is high up in the neck)
- Loss of normal bowel and bladder control (may include constipation, incontinence, bladder spasms)
- Numbness
- Sensory changes
- Spasticity (increased muscle tone)
- Pain
- Weakness, paralysis

**2. THORACIC LEVEL INJURIES**

When spinal injuries occur at thoracic level, symptoms can affect the legs:

- Loss of normal bowel and bladder control (may include constipation, incontinence, bladder spasms
- Numbness
- Sensory changes
- Spasticity (increased muscle tone)
- Pain
- Weakness, paralysis

Injuries to the high thoracic spinal cord may also result in blood pressure problems, abnormal sweating, and trouble maintaining normal body temperature.

**3. LUMBO SACRAL INJURIES**

When spinal injuries occur at the lumbo-sacral level, varying degrees of symptoms can affect one or both legs, as well as the muscles that control bladder and bowel:

- Loss of normal bowel and bladder control (constipation, leakage, and bladder spasms)
- Numbness
- Pain
- Sensory changes
- Spasticity (increased muscle tone)
- Weakness and paralysis

**COMPLETE SPINAL CORD INJURY**

In complete lesion, there is no sensory or motor function below the level of lesion. It is caused by a complete transection or severe compression or extensive vascular impairment (due to lack of blood supply to the spinal cord).

**INCOMPLETE SPINAL CORD INJURY**

An incomplete spinal cord injury is the term used to describe damage to the spinal cord that is not absolute. The incomplete injury will vary enormously from person to person and will be entirely dependent on the way the spinal cord has been compromised.

**TYPES OF INCOMPLETE SPINAL CORD INJURY**

- Central Cord Syndrome:

This is the commonest of the incomplete syndrome (CCS). It is characterized by weakness in upper more than lower limbs with sacral sparing. In addition, sensory loss below the lesion with partial sensory deficit. Schneidan and others noted that the etiologic factor was hyperextension with simultaneous compression of the cord by either anterior osteophytes and posterior impingement caused by buckling of the ligament flavum. It is more common with elderly group of patients with cervical spondylosis, than with younger group of traumatic patients. Pathology of the CCS is probably due to the injury to the central part of the cord. Depending on the severity of the lesion both upper and lower limbs getting affected with upper more than lower because in cervical fibers are more centrally located compared to dorsal, lumbar and sacral spine.

Quencer et al by MRI study found that CCS is predominately a white matter peripheral injury and not intramedullary hemorrhage. CCS usually have good prognosis. The pattern of recovery also seen starting with lower limbs followed by bladder bowel function and then Upper limb starting from proximal to distal and finally intrinsic muscle in Upper extremity. However younger patients recover faster than elderly patients. Similarly, bladder recovery is also seen in younger more than elderly patients. Patients with ASIA scale 'D' is able to recover well and are ambulatory.

**Brown –Sequard Syndrome (BSS) :**

- It occurs in cases of stab injuries with ipsilateral loss of all sensory modalities at the level of injury:
- With ipsilateral flaccid motor paralysis.
- With ipsilateral loss of position sense and vibration sense below the lesion.
- With ipsilateral loss of motor function below the lesion.
- With contralateral loss of pain and temperature sensation.

This is because of the crossing of spinothalamic fibers which carry pain and temperature fibers whereas corticospinal tracts cross in the brainstem Brown sequard plus syndrome (BSPS) is much more common compared to pure BSS i.e. with ipsilateral motor weakness (hemiplegia) and contralateral hemiagnosia.

Numerous nontraumatic causes of Brown-Séquard syndrome have also been reported, including the following:

- Tumor (primary or metastatic)
- Multiple sclerosis
- Disk herniation
- Herniation of the spinal cord through a dural defect (idiopathic or posttraumatic)
- Epidural hematoma
- Vertebral artery dissection
- Transverse myelitis
- Radiation
- Type II decompression sickness
- Intravenous drug use

**Anterior Cord Syndrome:**

Damage to the anterior 2 / 3 of the spinal cord preserving the posterior column due to retro-pulsed bone fragments either due to direct injury to the anterior cord or anterior spinal artery which supply blood to the anterior part of the spinal cord.( due to aortic or cardiac surgery, embolism, angioplasty, etc.) Clinically they present loss of motor and pinprick sensation with relative preservation of proprioception, deep pressure sensation. Usually the prognosis is very poor with poor motor recovery and coordination.

**Conus medullaries and Cauda equina lesion:**

It is the terminal part of the cord which lies at the inferior aspect of L1 vertebrae. The segment above conus medullaris is termed as epiconus consisting of cord segments L4-S1. Nerve roots then travel from conus medullaries caudal as the cauda equine.

Injuries to the conus will present LMN deficits of anal sphincter and bladder. If L3-S2 nerve roots are not affected, motor strength in the legs and feet may not be affected. If the roots are affected, it will give LMN damage with diminished reflexes.

Injuries below L1 usually give a LMN weakness supplying the lumbar and sacral segments causing muscle atrophy with bladder bowel involvement with loss of plantar reflexes, anal reflexes and bulbocaverous reflexes and impotence.

Cauda equine lesions have better prognosis than other types of SCI.

Fracture L1 may result in conus damage whereas fracture L2 downwards will affect cauda equina.

Isolated conus injury is very rare.

Incidence of conus and cauda equina injuries are common.

**DIAGNOSTIC MEASURES:**

Spinal cord injury is a medical emergency requiring immediate attention.

The health care provider will perform a physical exam, including a neurological exam. This will help identify the exact location of the injury, if it is not already known. Some of the person's reflexes may be abnormal or absent. Once swelling goes down, some reflexes may slowly recover.

The following tests may be ordered:

- A CT scan or MRI of the spine may show the location and extent of the damage and reveal problems such as blood clots (hematomas).
- Myelogram may be necessary in rare cases.
- Somatosensory evoked potential (SSEP) testing or magnetic stimulation may show if nerve signals can pass through the spinal cord.
- Spine x-rays may show fracture or damage to the bones of the spine.

As discussed earlier, the management of SCI begins at the location of the trauma. The initial focus should be to reach the patient immediately to hospital with utmost care not to aggravate the injury (well supported horizontal position of the spine).

**RADIOLOGICAL INVESTIGATIONS IN SPINAL CORD INJURY**

In addition to the physical examination special investigations should be used to identify associated injuries and to rule out complications that may hinder the rehabilitation of the patients.

**X-ray**

An X-ray is required to identify the problems with the bony structure of the body. In case of chronic spinal cord injury, a physiotherapist should take a detailed history about weight bearing activities performed by the patient and as a precautionary measure may ask for X-rays of the following regions

1. X - ray Spine anteroposterior and lateral view - To identify the surgical fixation and any associated problems with the same.
2. X - ray Pelvis and both hips anteroposterior view - To identify myositis ossificans and bladder stones.

In addition, if the patients present any history of fall since injury screen for the fractures of the relevant bones. Associated fractures at the time of spinal cord injury should also be screened by X -ray before starting rigorous rehabilitation and weight

bearing exercises. Any restriction of the movement with a bony end feel in the regions of elbow, knee and hip should be screened for presence of myositis ossificans using the X-ray.

**Magnetic Resonance Imaging**

MRI of the spine and spinal cord should be viewed to identify the extent of injury whether the transaction is complete or not and to identify any other associated soft tissue injuries of the spine.

**Ultrasound**

Ultrasound of the abdomen and bladder gives information about any associated visceral injuries, bladder stones and other bladder pathologies.

**Urodynamic investigations**

Whether or not urodynamic evaluation is required will be decided by the urologist. Preliminary investigations before consulting a urologist are

- Bowel and bladder history
- Routine urine test
- Urine culture test (in case any signs of infection are evident like fever and uncontrolled spasticity or spasms)
- Ultrasound of the bladder and urethra

If the urologist advices so, the urodynamic evaluation should be done. Urodynamic studies are usually advised in presence of symptoms of lower urinary tract dysfunction. Some of these symptoms are

- Frequent leakages
- Frequent or painful urination
- Uncontrolled urge to pass urine
- Difficulty in micturition
- Inability to void the bladder completely

Urodynamic evaluation is to thoroughly investigate the bladder, urethra and sphincter function. Different evaluations done and the information they provide during the urodynamic testing are as follows:

- Uroflowmetry, is conducted to measure the speed and flow of urine to find out any blockages or weakness of detrusor muscles
- Postvoid residual measurement is performed to evaluate bladder incontinence
- Cystometric test, conducted to identify involuntary bladder contractions. A cystometric test measures how much urine the bladder can hold, how much pressure builds up inside the bladder as it stores urine, and how full it is when the urge to urinate begins.
- Leak point pressure measurement conducted to identify at what pressure the bladder leaks and is performed to find out the type of bladder problem
- Pressure flow study evaluates what is the rate of urine flow when a certain pressure is applied by the bladder and what pressure is required to produce by the bladder to start emptying the urine. These tests help to locate any urine outlet blockages.
- Electromyography of bladder is performed to understand muscles' response to nerve stimulation, if the nerve impulses are reaching the muscles or not and if the muscles are responding to these impulses or not.

**Functional MRI:**

Functional magnetic resonance imaging (fMRI) is a noninvasive method for measuring activity in the brain and spinal cord. fMRI uses MRI technology to measure brain activity at rest as well as while performing a particular task by detecting changes in the blood flow in the area of

activation.

Oxygen in the blood is carried by hemoglobin. Oxygenated hemoglobin is diamagnetic (resistant to magnetism) than deoxygenated hemoglobin which is paramagnetic (attracted to magnetism). When a particular area of brain or spinal cord is active, blood flow to that area increases bringing in more oxygenated hemoglobin. This rise in oxygenated hemoglobin in the activated area is picked up by MRI scanner as an MR signal which is mapped and represented graphically by color coding according to the strength of activation. This way images of active brain areas can be collected at rest and while specific activities (such as visualizing/looking at an object/light, listening to sound/music, moving limbs etc.) are being performed.

**Procedure**

In SCI, fMRI is used to track changes in the function of brain and spinal cord following injury and to monitor effects of any treatment over time. During the fMRI procedure, the patient is asked to perform a specific task or is stimulated to trigger sensory response for a period of 30 seconds followed by rest for 30 seconds. This pattern of task followed by rest is repeated several times and MRI images are obtained which are computed and analyzed by a radiologist. For studying the areas involved in motor (movement) function, tasks like tapping the fingers, toes, moving feet up and down are commanded to perform, whereas for studying the areas involved in sensory function, sensory stimuli like stroking with a brush along a particular part of the patient's body are performed.

fMRI is used clinically and for research purpose in a variety of brain and spinal cord disorders. In SCI, it is used to:

- Help assess the effects of SCI on brain and spinal cord function.
- Map neuronal activity at different levels of the spinal cord in response to various sensory stimuli and motor tasks.
- Study how the brain or spinal cord recovers following an injury.
- Detect a neuronal response in the spinal cord below the site of injury to check areas of impaired and preserved activity.
- Identify the cause and mechanism of chronic neuropathic pain in SCI patients.
- Compare the effects of a treatment intervention with respect to function using a pre intervention and a post intervention fMRI.

**Contraindications of f-MRI:**

- Patients with metal and electronic implants
- Pregnant females, especially in the first trimester
- Patients that are unable to follow commands

**MEDICAL MANAGEMENT:**

A spinal cord trauma is a medical emergency requiring immediate treatment to reduce the long- term effects. The time between the injury and treatment is a critical factor affecting the eventual outcome. Initial treatment of patients with cord injury focuses on two aspects - preventing further damage and resuscitation.

**Prehospital Management:**

Spinal cord injury isn't always obvious. Mechanism of injury, pain in the vertebral column, or neurologic symptoms gives a clue to the emergency medical personnel regarding suspected spinal injury. Injuries involving the head, pelvis and those resulting from falling from heights should be suspected for spinal cord damage.

Emergency medical personnel stabilize and immobilize the spine at the scene of injury and transport the patient to the emergency

department (ED) taking care in not moving the spine. Many a times it is the bad lifting and transport that causes damage to the spinal cord.

**Emergency Department Management:**

A. ABC Resuscitation: Airway, breathing and circulation may be compromised because of spinal cord injury or associated injuries. Resuscitation is aimed at airway maintenance, adequate oxygen saturation of peripheral blood, restoring blood pressure to acceptable limits, preventing bradycardia, done simultaneously to prevent any ischemic damage to the already compromised cord.

   a. Airway management: Spinal cord injury makes the assessment and management of airway complex and difficult. Immobilization of cervical spine must be continued in neutral alignment during airway assessment and management.

      I. Restoring Airway Patency: Oral secretions are cleared to maintain airway patency and to prevent aspiration.

      II. Maintaining Airway Patency: To keep the airway patent, modified jaw thrust, and insertion of an oral airway is required in some patients and, endotracheal intubation in others. When indicated intubation should be done carefully in these patients to avoid spine movement and further spinal cord injury.

   b. Circulation Management: In patients with acute spinal cord injury, shock may be hemorrhagic and / or neurogenic. In these patients, a diligent search for occult sources of hemorrhage must be made before making the diagnosis of neurogenic shock as these patients have high incidence of associated injuries.

      I. Excluding Hemorrhagic shock: Computed tomography (CT) scan or X- Ray will reveal the most common sources of occult hemorrhage like chest, abdominal, retroperitoneal injuries and fractures of the pelvis or long-bones.

      II. Neurogenic shock and its management: History of spinal cord injury with exclusion of hemorrhage suggests spinal shock.

         I. Fluid resuscitation: Fluid resuscitation with isotonic crystalloid solution is the initial treatment of choice for neurogenic shock. These patients are at risk for the acute respiratory distress syndrome (ARDS) and hence volume overload should be avoided with judicious fluid resuscitation.

         ii. Maintaining blood pressure: Systolic blood pressure should be maintained above 90 mm Hg and hypotension should be avoided in these patients.

         iii. Maintaining oxygenation: Supplemental oxygenation and / or mechanical ventilation should be given to maintain adequate oxygenation and perfusion of the injured spinal cord.

         iv. Monitoring heart rate: Heart rate should be 60-100 beats per minute (bpm) in normal sinus rhythm. Atropine may be given for treatment of hemodynamically significant bradycardia.

         v. Maintaining urine output: Foley catheter should be inserted to decompress the neurogenic bladder. Urine output should be monitored and maintained more than 30 mL / h.

vi. Inotropic support: Dopamine or norepinephrine is should be reserved for patients who have decreased urinary output despite adequate fluid resuscitation. A low dose of dopamine in the 2- to 5-mcg / kg / min range is usually enough.

vii. Prevention of hypothermia: Hypothermia should be prevented in these patients.

B. Assessment and management of associated injuries: Twenty five percent of patients with spinal cord injury have associated head injury. A thorough evaluation for intracranial injury should be done in the presence of amnesia, external signs of head injury or basilar skull fracture, focal neurologic deficits, associated alcohol intoxication or drug abuse, and a history of loss of consciousness.

**Corticosteroid Therapy:**

Methyl prednisolone has been shown by the NASCIS-II (National Acute Spinal Cord Injury Studies) trial to be significantly beneficial in causing favorable neurological outcome if administered early enough (within 8 hours) of the primary insult. It appears to work by reducing damage to nerve cells and decreasing inflammation near the site of injury. To be effective corticosteroids should be given within 8 hours of injury. The recommended dose is 30mg / kg bolus to be administered over 15 minutes and after a 45-minute pause 5.4 mg / kg / hour over the next 23 hours.

Nesathurai and Shanker questioned the validity of the results of NASCIS trial. These authors cited concerns about the randomization, clinical endpoints and statistical analysis used in the study. In addition, the investigators noted that even if the benefits of steroid therapy were valid, the clinical gains were questionable. The steroid therapy is not free of side effects. An increased incidence of infection and avascular necrosis has been documented.

The Congress of Neurological Surgeons (CNS) has stated that steroid therapy "should only be undertaken with the knowledge that the evidence suggesting harmful side effects is more consistent than any suggestion of clinical benefit." Every hospital should have a policy regarding administration of steroids to spinal cord injury patients.

Monosialotetrahexosyl ganglioside (GM-1): GM-1 administered following acute spinal cord injury as an adjunct after the administration of methylprednisolone may be of benefit.

Spinal traction: Spinal traction is a more conservative and less invasive approach to allow the bones to heal naturally without surgery. Traction prevents movement of the spine. The skull may be held in place with tongs (metal braces placed in the skull and attached to traction weights or to a harness on the body). The spine braces may need to be worn for a long time.

**Surgical Management:**

Surgery may be required in the treatment of spinal cord injury patients. There are three major goals of surgery-

A. Spinal decompression: This involves removal of fluid tissue, bone fragments, disk fragments, or foreign objects that presses on the spinal cord.

B. Prevention of tethering: Neurological disorders related to pulling of the spinal cord at the base of the spinal canal causing stretching and damage. Surgery may prevent tethetering of the cord. [14]

C. Stabilization of the spine: The weakened vertebrae from fracture may not be capable of supporting the normal weight from the body and protect the spinal cord. Spinal instrumentation and fusion can be

used to provide permanent stability to the spinal column. These procedures correct, join, and solidify the level where a spinal element has been damaged. A combination of metal screws, rods, plates or cage may be necessary to help hold the vertebrae together and stabilize them until the bones heal.

Timing of surgery: There are no established standards regarding when to operate the patients with spinal cord injury because of the lack of well-designed studies. Emergency decompression of the spinal is suggested in the following indications-

1. Acute spinal cord injury with progressive neurologic deterioration, facet dislocation, or bilateral locked facets.
2. Spinal nerve impingement with progressive radiculopathy.
3. Extradural lesions such as epidural hematomas or abscesses.
4. Cauda equina syndrome.

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**Type of surgery:**

The type of surgery performed (anterior vs. posterior), distraction forces during surgery, preoperative grade all influence the outcome. The surgery may be-

**Spinal decompression:**

The major types of surgery for spinal decompression are-

a. Microdiscectomy / Corpectomy

b. Laminectomy

**Stabilization of the spine:**

Can be achieved by spinal fusion with or without instrumentation.

1. In the Craniovertebral junction, stabilization may be achieved by

a. C1-C2 fixation using pedicle screws / trans-articular screw / sublaminar wires

b. Occipitocervical fixation

2. In the cervical spine stabilization may be achieved by

a. Anterior approach using vertebral body plates and screw

b. Posterior approach using lateral mass screws

3. In the dorsal spine stabilization may be achieved by

a. Anterior transthoracic approach for interbody cage placement

b. Posterior approach using transpedicular screw and rod system

3. In the lumbar spine, types of stabilization procedures are-

a. Anterior lumbar interbody fusion (ALIF)- the disc is accessed from an anterior abdominal incision.

b. Posterior lumbar interbody fusion (PLIF)- the disc is accessed from a posterior incision. c. Transforaminal lumbar interbody fusion

c. Transforaminal interbody fusion(TLIF) - the disc is accessed from a posterior incision on one side of the spine. Instrumentation with screws, rods, plates or cages may be required to provide permanent stability to the spinal cord.

Surgery for prevention of tethering: After complete release and reconstruction of the

spinal cord, a Gore-Tex surgical membrane can be placed over the cord and fixed to the lateral dural surface with stay sutures to prevent tethering.

**Post-operative physiotherapy**

Post-operative physiotherapy starts immediately after the surgery and is continued during the convalescent period. Respiratory management plays a very important role to prevent post-operative complications. Restorative physiotherapy starts immediately after the patient is stabilized.

Rehabilitation of patients with Traumatic Spinal Cord Injury

The acute stage of rehabilitation is followed by the convalescent stage, where the patient is thoroughly evaluated by the team for further management. The management is based on classification of SCI as per the ASIA Scale.

The American Spinal Injury Association (ASIA) first published an international classification of spinal cord injury in 1982, called the International Standards for Neurological and Functional Classification of Spinal Cord Injury. Now in its sixth edition, the International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI) is still widely used to document sensory and motor impairments following SCI. It is based on neurological responses, touch and pinprick sensations tested in each dermatome, and strength of ten key muscles on each side of the body, including hip flexion (L2), shoulder shrug (C4), elbow flexion (C5), wrist extension (C6), and elbow extension (C7).Traumatic spinal cord injury is classified into five categories on the ASIA Impairment Scale:

**ASIA IMPAIRMENT SCALE**

- A indicates a "complete" spinal cord injury where no motor or sensory function is preserved in the sacral segments S4-S5.
- B indicates an "incomplete" spinal cord injury where sensory but not motor function is preserved below the neurological level and includes the sacral segments S4-S5. This is typically a transient phase and if the person recovers any motor function below the neurological level, that person essentially becomes a motor incomplete, i.e. ASIA C or D.
- C indicates an "incomplete" spinal cord injury where motor function is preserved below the neurological level and more than half of key muscles below the neurological level have a muscle grade of less than 3, which indicates active movement with full range of motion against gravity.
- D indicates an "incomplete" spinal cord injury where motor function is preserved below the neurological level and at least half of the key muscles below the neurological level have a muscle grade of 3 or more.
- E indicates "normal" where motor and sensory scores are normal. Note that it is possible to have spinal cord injury and neurological deficits with completely normal motor and sensory scores.

After evaluation of the client based on the classification, it is vital to set goals or achieving the realistic expected outcome.

**Prediction of outcome:**

Physical examination is the most important mehtod to predict the outcome.

Initial presence of muscle power would give a clear prediction. The earlier the muscle shows some power the faster muscle could come to functional level i.e above 3.

Within the first 6 months the recovery seen is maximum and the improvement in the first three months is usually more than the next three months.

If one gets back some power initially the chances of improvement is better than the muscles remaining "0".

However, those who have an incomplete injury initially have better prognosis for ambulation. MRI is the most superior radiological test for determining the prognosis. E.g. if a hemorrhage seen initially, the prognosis is poor. Contusion is worse than edema. If no hemorrhage is seen, then damage to the cord is incomplete and the prognosis is better. The degree and the extent of edema also determines the chances of recovery. In other words, along with clinical assessment, MRI can give the prediction better. The other investigations include nerve conduction studies, late responses (H-reflex and F-wave) and somatosensory evoked potentials.

Whatever the prognosis is, rehab has a definite role to play and whenever any scientific advances come, they could try at that stage. Till then the patient has to come to terms and try to become as independent as possible.

**Prescription of the equipments:**

*Things to be considered are:*

1. Easy availability
2. Accessibility
3. Durability
4. Safety
5. Comfort
6. Cost

It is advisable to involve the patients in planning. However very few patients can make their own decisions and therefore therapist has to explain advantages and disadvantages, often trying out different techniques. Perhaps, it will be more beneficial to show other patients who have experienced the same problems and have solved their problems. Many a times it would be essential to set a trial period, trying out trial calipers / shoes. Eventually the therapist can take a final decision in prescribing the necessary equipments. Before taking the final decision, therapist has to make a thorough evaluation of motor skills, functional skills, home assessment, ADL and opinions of other team members and taking patients' consent.

**Evaluation:**

1. Data collection:

- From patients and relatives.
- From medical records including X-rays, MRI, and other reports.

It is essential to explain to patients the need for evaluation and taking proper history so that he actively participates in the actual rehabilitation process.

**2. Evaluation Procedures:**

- Assessment of strength.
- ROM
- Sensations
- Tone
- Girth measurement

**3. Physical Evaluation:**

*a) Skin Evaluation:*

1. Bony prominences and vulnerable area of skin.
2. Scar tissue.
3. Pressure sores.
4. Secondary bad postures because of pressure sores.

*b) Sensory Evaluation:*

1. Superficial and deep sensations like pain, temperature, touch joint position etc.
2. Patient's awareness about sensory deficits.

3. Implications because of sensory deficits namely burns, bruises lack of coordination because of lack of kinesthetic sense.
4. Damage to skin due to faulty equipment like orthosis.

*c) Respiratory Evaluation:*

**4. General observations:**

a) Shape and symmetry of chest.

b) Muscle tone.

c) Wasting.

d) Presence of artificial airways like tracheostomy, intercostals tubes.

Respiratory history and associated medical history may indicate that he may require preventive measures for chest complications. Evaluation of upper and lower chest expansions may give a clue as to which lobes require to be attended. Lack of chest expansion and mobility may lead to decreased vital capacity. (Normal value of chest expansion at xiphoid process should be at least 2 1 / 2 to3 inches.) Lack of ROM at shoulder level may lead to poor chest expansion. It is essential to have normal flexibility of trunk and hips as it is easier for him to cough effectively. Normal posture of trunk is essential for rib mobility as excessive tightness of ribs would lead to poor Vital capacity.

**5. Respiratory Muscle Strength Evaluation:**

- Diaphragm.
- Abdominals.
- Accessory muscles like intercostals, scalenes, serratus anterior, pectorals, erector spinae and sternocleidomastoid.

**Implications:** If abdominal muscles are weak, it may affect in poor coughing because of inability to build up intrathoracic pressure to expel air and therefore bronchial hygiene may be affected very badly.

1. An abdominal binder may be useful.
2. Increased respiratory rate indicate shallow breaths.
3. When abdominals are paralyzed, they breathe best in lying.
4. Hypoventilation in patient may also cause drowsiness and irritability. This usually happens among quadriplegics or high-level paraplegics because of weakness of accessory muscles.
5. Hypoventilation may cause fainting, tingling and numbness.
6. Poor co-ordination of breathing with functional activities may cause lack of endurance.
7. Deep breathing exercises would increase inspiratory capacity which will improve coughing and ventilation of all the lobes thus decreasing respiratory complications.

**Personal hygiene**

1. Evaluation of cough and Vital Capacity
2. Evaluation of patients' knowledge of postural drainage.

**Range of Joints:**

- Rolling may be more difficult with overstretched back and trunk muscles.
- Selective tightness of the long flexors of the hand will help in grasp (Tenodesis action).
- Maintaining of adequate ROM is essential for helping the totally dependent patients to move for changing the positions.
- If a patient can be moved easily, there is less chances for pressure sores.

**Muscle Tone:**

1. Mild increase: Resistance for passive stretching with full ROM, function not

being affected

2. Moderate increase : More resistance and full ROM is possible but function being affected.
3. Severe increase: Full ROM is not possible and many functional skills impossible.

**Description of quality of Tone:**

1. Constant or Fluctuating?
2. Is it related to position change?
3. Symmetrical or Asymmetrical?
4. Fluctuation related to time.
5. Is increased tone beneficial in functional activities? e.g minimal increase in tone in extensors of hip & knee is beneficial in standing.

**Outcome measures**

Outcome measures are designed to carefully assess and gather information from patients with spinal cord injury to be able to monitor their progress and to allow standardization of measurement and comparability of findings of multiple therapists. Outcome measures allow to record the changes quantifiably. Outcome measures are tested for validity, reliability for using again and again by multiple assessors as well as for use by multiple assessors, specificity and sensitivity. It is important to choose a good outcome measure that has all of these psychometric qualities for better assessment and monitoring of the patients. Some of the commonly used outcomes measures in spinal cord injury are ASIA scale and Spinal Cord Injury Independence Measure (SCIM) used for sensori-motor and functional assessment. Apart from this various other outcome measures can be used for measuring, spasms, balance, strength, bed mobility, ambulation etc.

**Strength:**

Complete evaluation of all muscles.

Difficultes in evaluating the muscles:

1. Immobilization device.
2. Poor stabilization because of weak muscles.
3. Trick movements.

**Implications:**

*Some of the key muscles:*

1. Scapular stabilizers useful in one-man pivot transfer.
2. Pectoralis Major essential for rolling.
3. Triceps for bed mobility skills.
4. Wrist Extensors:
   1. Tenodesis.
   2. Moving wheelchair.
   3. Use as a hook in other ADL.
5. Triceps: are essential for Wheelchair push-ups. Lateral transfers.
6. Knee flexors and extensors for ambulation with AFO.
7. Hip flexors: very useful in ambulation.
8. Ankle Dorsiflexors: for clearing the ground and ambulation without AFO

**Identification of the compensatory movements:**

*Common muscle compensations seen in spinal cord injured patients*

1. Neck flexors may be instead of upper abdominals
2. Shoulder internal rotators can compensate for elbow flexion
3. Shoulder external rotators can compensate for elbow extension
4. Shoulder abductors can compensate shoulder flexion in scaption movement
5. Trapezius can for be used shoulder flexion and abduction through shoulder and

scapular elevation

6. Wrist extension can be used for finger flexion through tenodesis
7. Pronation can be used for wrist flexion in gravity assisted plane
8. Supination can be used for wrist extension gravity assisted plane
9. Latissimus dorsi can be used instead of quadratus lumborum for hiking of pelvis
10. External and internal obliques can be used for pelvic hiking as well as to generator hip flexion movement in gravity eliminated plane
11. Trunk side flexors can mimic the movement of hip abduction in gravity eliminate plane

These muscle compensations should be identified and encouraged for functional benefits. If the patients are not able to perform the movements using these compensations these should be taught to the patient.

**Tolerance for Vertical Position:**

1. Lying to sitting in bed.
2. Reclining wheelchair.
3. Tilt table. Implications:

   1. ADL independence, especially

      a) Transfers.

      b) Wheelchair movements.

      c) Dressing.

**Balance :**

- Evaluation of equilibrium reaction.
- Evaluation of static balance / Dynamic balance.

**Implications:**

Unless patient had a head injury also, loss of balance is due to posterior column involvement and weakness of certain group of muscles. Therefore, he needs to learn to use some other group of muscles. Providing a trunk support may also help.

**Coordination:**

a) Kinesthetic sense or awareness and accuracy of movements depend on timing. Find out as to how good was his performing skill prior to injury in comparison to the present skill.

b) It may require a lot of repetitions for improving the skill, also repeated instructions and explanations are beneficial.

c) ADL training is the goal.

**Endurance :**

1) Evaluation of cardiopulmonary status.

2) Evaluation of breathing with different activities.

3) Because of poor endurance, ADL may become slow.

4) Many a times Exercise sessions must be made short for want of endurance.

**Emotional Status:**

1) Evaluation of patients understanding of the disability, whether he is realistic or not in his expectation of the outcome.

2) Evaluation of his accepting the disability.

3) Does he take active interest in solving his problems by himself? Is he interested in having equipments, which would improve the quality of life? e.g An orthosis may be useful for ambulation or perhaps he may have to use a W / C for the rest of his life

4) Is there a desire for him to be independent in his ADL or does he expect all the help from spouse or caretaker?

**Relevant Information required in the Chart:**

A) Age: Younger patients may be able to adapt or perform better. Would like to be independent in ADL

B) Body build :

1) Has he lost weight or put on with heavy weight? With heavy weights the movements are reduced, the chances of getting pressure sores are more. At the same time, the thinner the person more the bony prominences and they are also prone to have pressure sores.

2) Standard wheelchairs are easily available, whereas over size wheelchairs and cushions are to be custom made. Over size wheelchair may not pass through standard doors. Wheelchair movements may also require larger area for mobility.

3) Heavier patients may require a greater number of people for ADL especially transferring. Many people prefer institutional life.

**C) Type of Injury:**

Type of injury and extent of injury would help in knowing the prognosis. e.g. Cases with complete transection of the cord, almost always will not have any chance of recovery and hence the patients have to live with disability. Similarly, the level of injury would also give us an idea about the outcome as well as the chances of complications.

**D) General Health :**

Other injuries like head injury, limb fractures, cardiac, respiratory problems, ectopic bone formation, bladder and bowel problems are to be taken into consideration. They all affect the therapy and functional outcome. e.g. Ectopic bone formation affect ROM. Cardiac & Respiratory problems may affect the performance and endurance.

**E) Personality and Life style:**

1. Whether his work was sedentary nature or not.
2. Educational level / occupation
3. Willingness for change of lifestyle or job.
4. Acceptance of disability: Whether he is flexible or not?

**F) Family Support:**

1. Is family supportive?
2. Will he need another care giver?
3. Source of income for maintenance?
4. Will they continue the same therapy at home also?
5. Will they arrange for Vocational Rehabilitation so that he too can become an earning member and he too can live with dignity?

Evaluation is a continuous process. Initial evaluation is only a baseline evaluation to find out main problems of the patient. It is only to see whether we are able to go towards the expected goal and final evaluation before discharge is to see whether he has achieved the outcomes so that he can lead a meaningful life at home.

**Precautions and Complications of Spinal Cord Injury**

A serious condition like Spinal cord injury results in functional, psychological and socioeconomic implications. Several complications follow the occurrence of Spinal cord injury (SCI). These could be immediate or long-term secondary to the injury. Hence continued care is required for these patients to prevent the complications and to treat the complications of time. Complications may require re-hospitalization and are most often the cause of morbidity and death. They significantly hamper the quality of life and functional abilities and independence of the patients. Prevention, early identification and immediate treatment of these complications

is important for improving quality of life and lifespan of the spinal cord injury patients.

We have discussed some of the significant complications of spinal cord injury which are as follows

1) Autonomic Dysreflexia

2) Pulmonary Embolism

3) Deep Vein Thrombosis

4) Postural Hypotension

5) Edema

6) Pain

7) Abnormalities of temperature regulation

8) Heterotopic (Ectopic) Ossification

9) Contractures

10) Osteoporosis

**Autonomic Dysreflexia:**

Autonomic Dysreflexia is due an over-activity of the Autonomic Nervous system. It is seen in patients at T5 and above level of injuries. Autonomic dysreflexia can develop suddenly and is a possible emergency. If not treated promptly and correctly, it may lead to seizures, stroke, and even death.

Mechanism: It can occur when an irritating stimulus is introduced to the body below the level of spinal cord injury, such as an overfull bladder. The stimulus sends nerve impulses to the spinal cord, where they travel upward until they are blocked by the lesion at the level of injury. Since the impulses cannot reach the brain, a reflex is activated that increases activity of the sympathetic portion of autonomic nervous system. This results in spasms and a narrowing of the blood vessels, which causes a rise in the blood pressure. Nerve receptors in the heart and blood vessels detect this rise in blood pressure and send a message to the brain. The brain sends a message to the heart, causing the heartbeat to slow down and the blood vessels above the level of injury to dilate. However, the brain cannot send messages below the level of injury, due to the spinal cord lesion, and therefore the blood pressure cannot be regulated. This can be a MEDICALEMERGENCY because extremely high blood pressure can result in stroke, seizures, or death.

**Causes:**

There can be many stimuli that cause Autonomic Dysreflexia. Anything that would have been painful, uncomfortable or physically irritating before the injury may cause AD after the injury. Things to consider include

1) Bladder associated triggers (most common)- from overstretch or irritation of bladder wall.

**This includes**

- Urinary tract infections
- Urinary Retention
- Blocked Catheter
- Overfilled collection bag
- Noncompliance with intermittent catheterization program etc

2) Bowel related triggers - over distention or irritation

- Constipation / impaction
- Distention during bowel program( digital stimulation)
- Hemorrhoids or anal fissures
- Infection or irritation ( e.g., appendicitis)

3) Skin related triggers

- Any direct irritant below the level of injury(e.g., prolonged pressure by object in shoe or chair, cut, bruise, abrasion)
- Pressure Sores (Decubitus ulcers)
- Ingrown toe nails

- Burns( e.g., sun burn, burns from using hot water)
- Tight or restrictive clothing or pressure to skin form sitting on wrinkled clothing

4) Sexual Activity related triggers:

- Overstimulation during sexual activity (stimuli to the pelvic area which ordinarily be painful if sensation were present)
- Menstrual cramps
- Labor and delivery

5) Others:

- Heterotopic Ossification
- Acute Abdominal condition (gastric ulcers, colitis, peritonitis)
- Skeletal fractures What are the Signs of AD?

A patient with AD may have one or more of the following findings on physical examination

1) Pounding headache (caused by high blood pressure)
2) Goose bumps/ cool, clammy skin
3) Sweating above the level of injury
4) Nasal Congestion
5) Nausea
6) Slow Pulse/ Heart Rate (< 60 beats per minute)
7) Blotching of the Skin
8) Restlessness

The sudden rise in the blood pressure in AD is usually because of slow heart rate. Normal systolic pressure for SCI above T6 is 90-110mm Hg. Blood pressure 20- 40mmHg above the reference range may be a sign of AD for individuals with SCI. However, patients with AD may display no symptoms, despite elevated blood pressure.

**Prevention of AD:**

Following are the list of precautions that one should take to prevent episodes of AD:

- Frequent pressure relief or change of position in bed/ chair
- Avoidance of sun burn/scalds (avoid overexposure, use of #15 or greater sunscreen, watch water temperatures)
- Maintain a regular bowel program.
- Well-balanced diet and adequate fluid intake
- Compliance with medications
- Persons at risk and those close to them should be educated in the causes, signs and symptoms, first aid, and prevention of autonomic dysreflexia.
- If you have an indwelling catheter:

Keep the tubing free of kinks

- Keep the drainage bags empty
- Check daily for grits (deposits) inside of the catheter.
- If you are on an intermittent catheterization program, catheterize yourself as often as necessary to prevent overfilling.
- If you have spontaneous voiding, make sure you have an adequate output.
- Carry an intermittent catheter kit when you are away from home.
- Perform routine skin assessments. What to do if these signs are coming on? Initial Steps:
- SIT UP or raise the head of the bed immediately
- Look for the cause or alert your nurse, therapist or caregiver so they can look too
- Bladder: Do you need to catheterize? Is the

Foley kinked, plugged or full?

- Bowel: Do you get constipated often? Has it been 3 or more days since your last bowel movement?
- Skin: Do you have pressure sores, burns cuts, bruises? Is your clothing/ undergarments twisted? Do you have an ingrown toenail?
- FIX IT! Catheterize, do your bowel program, adjust clothing or remove whatever offending stimulus is bothering your body! Once the stimulus is removed, you should notice relief from the above symptoms. If not, get to the EMERGENCY ROOM and be ready to explain dysrreflexia because not all health care providers encounter SCI or dysreflexia on a regular basis.
- Consider medical management. If systolic BP remains above 150mm. Further Treatment:

If symptoms persist or BP still remains elevated following the above efforts or a cause cannot be readily identified, pharmacological treatment with a short acting antihypertensive medication should be started simultaneously with the search for and treatment of the noxious stimulus.

NOTE: teaching the patient and care giver about this complication is essential.

**Reminder**

Obtain emergency medical treatment, if you are unable to find the stimulus causing autonomic dysreflexia.

**Pulmonary Embolism:**

**What is pulmonary Embolism?**

Pulmonary Embolism is a potential cardiovascular emergency that occurs when a part of thrombus, usually dislodged from a DVT (called an embolus), passes in the pulmonary circulation and blocks the pulmonary arteries that supply the lungs with blood. Blood clots in the lungs prevent enough oxygen from getting into our body and oxygen is what fuels our brain, muscles, everything. PE is a potentially life-threatening condition which can be preventable.

**Incidence of Pulmonary Embolism:**

Studies show a correlation between the Incidence of deep vein thrombosis (DVT), pulmonary embolism (PE), and death during the initial acute hospitalization after traumatic spinal cord injury (SCI). Studies show that incidence of Pulmonary Embolism is 4.6% and DVT is 14.5% with higher incidence of mortality seen in quadriplegics as compared to paraplegics.

**Diagnosis of PE:**

PE is a potentially life-threatening condition and in severe cases the occurrence of circulatory collapse and cardiac arrest may result in sudden death, hence rapid diagnosis is crucial. Early fatality occurs in up to 15% of patients and thus rapid diagnosis is crucial. However, the diagnosis of PE may be missed because of its non-specific clinical symptom.

Common signs and symptoms of PE are:

- Dyspnoea, Shortness of breath (worse than the breathing problems your SCI may cause)
- Chest pain, also pain in jaw or shoulder (like the signs of a heart attack)
- Heavy feeling in chest
- Cough
- Fever
- Haemoptysis (Red streaks in coughed-up secretions)
- Syncope
- Unilateral leg pain

- Signs of DVT (unilateral extremity swelling)
- Irritable or anxious feeling
- Fast heartbeat (pulse)
- Pale complexion, bluish tint to lips

These symptoms are not specifically diagnostic of PE. Hence careful judgment of the clinician is required regarding the need for imaging tests designed to detect PE. A clinician should maintain a high index of suspicion for this condition, because prompt treatment of PE can dramatically reduce the levels of morbidity and mortality associated with PE.

**How it is diagnosed?**

When you visit your doctor for your symptoms, they'll ask about your overall health and any pre-existing conditions you may have. Your doctor will typically perform one or more of the following tests to discover the cause of your symptoms:

- Chest X-ray: This standard, noninvasive test allows doctors to see your heart and lungs in detail, as well as any problems with the bones around your lungs.
- Electrocardiography (ECG): This test measures your heart's electrical activity.
- Magnetic resonance imaging (MRI): This scan uses radio waves and magnetic field to produce detailed images.
- Computerized tomography (CT) scan: This scan gives your doctor the ability to see cross-sectional images of your lungs.
- Pulmonary angiography: This test involves making a small incision so your doctor can guide specialized tools through your veins. Your doctor will inject a special dye so that the blood vessels of the lung can be seen.
- Duplex venous ultrasound: This test uses radio waves to visualize the flow of blood and to check for blood clots in your legs.
- Venography: This is a specialized X-ray of the veins of your legs.

**What to Do if there are signs for PE?**

THIS IS AN EMERGENCY! CALL the doctor.

- Avoid massaging legs and avoid pressure behind legs
- ecrease activity level (your lungs aren't getting enough fuel…don't use more!)
- Sit up/lean forward or get into a position that makes breathing easier
- Remove tight clothing that may restrict breathing
- Use supplemental oxygen if available.

**How it is treated?**

The treatment for a pulmonary embolism depends on the size and location of the blood clot. If the problem is minor and caught early, you may be advised to take medications to break the clots. Commonly used drugs:

- Anticoagulants: Also called as blood thinners, e.g., heparin and warfarin, these drugs prevent formation of new clots in your blood.
- Thrombolytics( clot dissolvers ): These drugs are used to break the clot. They're used in emergency situations because of its side effects which include dangerous bleeding problems.

Surgery may be necessary to remove problematic clots, especially those that restrict the blood flow to the lungs or heart. Some surgical procedures your doctor may use in the case of a pulmonary embolism

- Vein filter: A small incision will be made to install a small filter in your inferior vena cava (the main vein that leads from your legs to the right side of your heart). The filter prevents blood clots from traveling

from your legs to your lungs.

- Clot removal: A thin tube called a catheter will suction large clots out of your artery. It isn't an entirely effective method because of the difficulty involved, so it's not always a preferred method of treatment.
- Open surgery: Doctors use open surgery only in emergency situations when a person is in shock or medications aren't working to break up the clot.

Follow-Up Care: includes care after you are properly treated for the pulmonary embolism at hospital

- Treatment of the underlying cause. This is typically deep vein thrombosis.
- Blood thinner medications, such as heparin and warfarin, to prevent blood clots from returning.
- Regular use of compression stockings to prevent clots from forming in your legs.
- Regularly passive ROM exercises to prevent future blood clot

**Deep Vein Thrombosis:**

Deep Vein Thrombosis is one of the common complications seen in patients with Spinal Cord Injury. DVT and subsequent Pulmonary embolism remain significant causes of morbidity, mortality and substantial healthcare costs in patients with SCI. Venous thromboembolism remains the third leading cause of death after SCI. Morbidities include thromboembolism, prolonged edema and pressure ulcers. It is a condition in which a blood clot is formed in one or more of the veins in the body, most often in the lower legs or thigh. DVT can result in life threatening danger if the clot breaks loose from the leg vein and finds its way to the lungs causing pulmonary embolism. The risk of DVT is high in the acute phase of SCI.

**Incidence:**

Incidence of DVT varies significantly among the various racial groups and it also varies from population to population and from country to country. Asian patients have been reported to have a significantly lower incidence of idiopathic and secondary venous thromboembolism as compared to the western patients. The clinical symptoms are seen in only 15% of patients with acute SCI and pulmonary edema develops in approximately 5% of these patients.

**Causes:**

- inactivity after SCI
- lack of pumping action of large muscle groups, which causes pooling of blood in the veins and trauma.

Signs and Symptoms : The signs and symptoms of DVT are varied and it depends upon the severity.

- Pain and tenderness in the leg mainly in ankle and foot, it often starts in the calf area and feels like cramping. However, majority of the paraplegics are unable to feel the pain, therefore one has to look for other symptoms to diagnose and treat it immediately.
- Swelling: around the affected calf muscle in the affected leg, including the swelling around ankle and foot
- Skin discoloration: like the limb may turn pale, red or blue.
- Increased warmth of the affected area.

Although the various methods of DVT detection will be discussed, it is very important for the health care professionals, patients, family members and care givers to be educated regarding the early signs and symptoms.

What to do if I have signs of DVT:

1) Call doctor or got to the emergency room

2) Elevate the affected leg/arm.

3) Remove the stockings

4) DO NOT MASSAGE or exercise the affected leg/arm.

5) Doctor will advise tests and give instructions. Always follow them.

**Preventive measures:**

1) Regular inspection twice daily for any unusual increase in the calf/ thigh circumference, low grade fever of unknown origin and/or pain, tenderness or heaviness of the affected extremity.

2) Routine practice of passive and active ROM exercises. Mobilization and movement of the extremities should be essential in the prevention of DVT after SCI.

3) Routine use of graduated compression stockings is common in people with paralysis.

**Treatment : treatment of DVT is aimed at**

1) Prevent the further increase in the size of the clot

2) Prevent the clot from dislodging and causing subsequent pulmonary embolism

3) Reducing the chances of a recurrence of DVT.

**Various treatment options include**

1) Blood thinners: Doctors use anticoagulants, commonly called blood thinners, to prevent blood clots. In spinal cord injury, anticoagulants are generally given with the first 72 hours after injury to all patients. The thinners are usually given for about eight weeks. The most common type of blood thinner used in SCI is a low molecular weight heparin such as enoxaparin or dalteparin. These medications slow the time it takes for blood to clot and also prevent growth of a clot. Blood thinners do not remove existing clots; that sometimes involves surgery, they can prevent clots from getting bigger or reduce the risk of developing additional clots.

As soon as a patient is suspected to have DVT, infusion of blood thinner like heparin is given for a few days. After the course of the heparin injections, oral medications like warfarin are usually given. Oral blood thinners are required for at least a minimum of 3 months. However, these blood thinners have serious side effects like risk of bleeding especially with higher doses, however if the dose is too low there can be chances of getting additional blood clots. Hence it is necessary to get a periodic check up of the blood clotting and bleeding time , so that the physician can accordingly adjust the dosages. In the event of a paraplegic getting a life threatening complication like pulmonary embolism, the physician may give other medicines like tissue plasminogen(TPA)

2) In cases where one cannot be given blood thinners, filters are inserted in the vena cava, the IVC filter is designed to capture blood clots from deep veins (mostly commonly from the leg) that would otherwise travel to the heart and lungs. A vena cava filter is a cone shaped medical device that is inserted into the inferior vena cava (IVC), the largest vein in the body, to prevent devastating pulmonary embolisms.

3) Routine use of graduated compression stockings helps in preventing the swelling that is associated with DVT. The stocking are worn on the leg from foot to knee. They are to be worn for duration of approximately 1 yr.

**Postural Hypotension**

Postural or Orthostatic hypotension is a sudden drop in blood pressure caused by moving from a lying to a sitting or standing position. The patient complains of feeling dizzy or light headedness with fast changes

in position and it often occurs after spending a long time in bed and then suddenly getting up. Although orthostatic hypotension can happen to anyone, it is more common following a spinal cord injury (SCI) and in the first few weeks of rehabilitation. This is because of the loss of nervous system control which works to keep the blood pressure stable, as veins and arteries can't tighten or widen like they used to below the level of injury, also the muscles in the arms/trunk/legs aren't working, to help return the blood to the heart.

Orthostatic hypotension is defined by The Consensus Committee of the American Autonomic Society and the American Academy of Neurology (1996) as a decrease in systolic blood pressure of 20 mmHg or more, or in diastolic blood pressure of 10 mmHg or more, upon the assumption of an upright posture from a supine position, regardless of whether symptoms occur.

Postural Hypotension is more commonly experienced by cervical SCI and high thoracic (levels 1- 6) spinal cord lesions. In most cases, it resolves and happens only after being in bed for long periods of time, and in the morning, however it can continue in few and medications may be necessary. Different patient react differently with postural hypotension. Higher level injury patients suffer with postural hypotension for a longer time after the injury.

**Signs of Postural Hypotension**

- Dizziness
- Lightheadedness
- Nausea
- Fainting
- Sweating
- Cool, clammy skin
- Fatigue
- Muscle Weakness
- Syncope (temporary loss of consciousness) How to Prevent and Manage Postural Hypotension?
- Initially, you may need a wheelchair that leans back, reclining chair or ask somebody to tip you back. The first few times you get out of bed the back of the chair will be tilted back and then slowly raised to an upright position as you are able to tolerate sitting up.
- Check your Wear TED hose, ACE wraps, abdominal binder, and stockings and make sure it is tight enough. They may help to stop the blood pressure from dropping as you sit up.
- Move SLOWLY (especially in the morning`): gradually come to sitting or standing positions once the dizziness has gone, do this for few times until your body becomes used to it. If it continues, return to bed
- Elevate head of bed and sit up in bed for long time before transferring to your wheelchair (progressive sitting)
- Stay active and hydrated, and avoid staying in bed for long periods of time unless medically necessary.
- There are medications that may help raise the blood pressure. If hypotension continues to occur, consult your doctor.

**Pharmacological Management:**

- Fludrocortisone acetate (Florinef)

Mechanism: Salt and water retention/ delayed action

Side effects: HTN, CHF, hypokalemia, hypernatremia, weight gain, edema

- Ephedrine sulphate (Ephedrine)

Mechanism: Alpha & beta sympathetic agonist

Side effects: Tachycardia, arrhythmias, HTN, palpitations, urine retention, insomnia,

sweating, tremors, tachyphylaxis, abuse potential

- Midoridinehcl (ProAmitine)

Mechanism: Alpha-1 adrenoreceptor agonist

Side effects: Headache, paresthesia, dysuria, pruritis, piloerection, supine HTN

**Non-Pharmacological Management:**

- Regulating Sodium and fluid intake: Increases in fluid intake and a diet high in salt/sodium can expand extracellular fluid volume and improve orthostatic responses. Though no sufficient evidences are available
- Use of abdominal binders or elastic stockings improves the cardiovascular responses during the sub maximal arm exercises. Although there are insufficient evidences on the same.
- Whole Body Vibration: it is found to increase the mean arterial pressure in individuals with SCI.
- Use of FES (Functional Electrical Stimulation) can be used as an adjunct to minimize the cardiovascular changes that occur during Postural hypotension.
- Evidence shows that exercise training enhances the sympathetic outflow in individuals and makes them more tolerable to OH as compared to inactive individuals with SCI.
- Also regular standing training has shown to improve the tolerance to OH.

**IMPORTANT NOTE:**

- Attempting to fight with dizziness is not a matter of will power, if you ignore these early signs you might faint. If this happens and you are not able to assume a reclined position, then you will deprive your brain of blood and oxygen and possibly cause brain damage.
- If you find yourself getting dizzy suddenly after a very long time, this might be a sign that you have an infection or some other medical problem. Call your doctor.

**Edema:**

Edema is the medical term for swelling which simply means collection of fluid under the skin.

**Causes:**

- Swelling in lower limbs is caused by gravity. While sitting up in a wheelchair, legs and feet dangle and gravity pulls everything down towards the earth, including blood. Blood pools in the feet and this fluid moves from the blood vessels into the tissues of the feet and ankles. Thus, swelling is usually worse at the end of the day when legs and feet have been hanging down all day.
- Swelling is also made worse when paralyzed muscles in the feet and legs don't work to help veins push blood back to the heart. Injury or surgery can also cause damage to the valves in veins making it even easier for blood to pool in the feet and ankles.

**How to Prevent Swelling/ Edema?**

- Wear TED hose or compressive stockings
- Elevate the legs and feet throughout the day and when it is possible
- MOVE as much as possible (exercise, propel wheelchair, range of motion, walk if possible)
- For quadriplegia: prop hands up on pillows when sitting or massage hands from fingers towards arm

Note: DON'T MASSAGE FEET/LEG, that would dislodge a blood clot and might cause a pulmonary embolism or stroke.

Side-effects of Edema: chronic swelling can lead to

- Soft tissue infections like cellulitis
- Pressure ulcers
- Skin becomes fragile and it is more prone to splitting and cracking

**Impaired temperature Regulation:**

The body regulates temperature by sweating to cool off, shivering to warm up, and by shrinking or expanding blood vessels to give off heat or keep heat in. After a spinal cord injury, the body's ability to control temperature is affected, which can lead to hypothermia or heat stroke. It is due to the loss of internal thermoregulatory responses, due to which there is absence of thermoregulatory sweating. The higher the injury level and the more complete an injury is, the greater it is difficult for temperature regulation. Due to the decreased sensation patient can become severely sunburned or frost bitten. The following precautions are to be taken in extreme conditions to regulate the body temperature.

**In Cold Weather:**

- Wear warm clothing, hat, gloves and socks
- Use extra blankets
- Drink warm fluids
- DON'T use heating pads, electric blankets on skin if sensation is affected. There are chances of burns.
- Don't keep hot plates or cups on the lap without a towel. The skin on your thighs is more sensitive than the skin on your hands. Also check the seat with hands for temperature before transferring on the car seat.

**In Hot Weather:**

- Don't stay out in Sun.
- Use hats and stay in shade
- Spray self with cool water at regular intervals.
- Drink cool fluids
- Use fan or air conditioner
- Avoid hot tubs for bathing
- Be careful not to put your bare feet on hot pavement
- Watch out for decreased urine output, this could signal dehydration, which can contribute to all kinds of other secondary problems/complications.

**Neurogenic Pain:**

After a spinal cord injury, pain is experienced by many paraplegics at or below their level of injury. It is the most challenging medical problems. This neuropathic pain is very difficult to treat and patients spend many sleepless nights. Many of them may experience pain immediately after their injury and then as the body heals the pain subsides. Musculoskeletal, neuropathic, and pain from the trauma related to the injury are common. A lot of individuals with a spinal cord injury suffer with chronic pain to the extent that it may affect their ADLs.

**Types of Pain:**

Musculoskeletal Pain: It usually occurs above the level of injury in complete injuries and below the level in incomplete injuries. It occurs in parts of the body

like the bones, joints, and muscles. And usually it is worsened by movement and eased with rest. This type of pain is usually dull aching in nature.

**Causes:**

- Overuse of joints and muscles
- Overuse syndrome/ pressure syndrome eg: rotator cuff tendinitis seen in individuals using manual wheelchair
- Muscles spasms pain
- Mechanical instability of spine around the

operated site

**Management: Anti-inflammatory medications**

Neuropathic Pain

Neuropathic pain can occur anywhere at or below your injury. There are 3 types, SCI central pain, Segmental, nerve root entrapment pain, pain due to syrinx

Causes: The cause is not really understood. It is believed that neuropathic pain is related to the nerve endings at the site of the injury. It is felt that because the brain cannot receive and send messages through the nerves, that it may think that there is pain below the level of injury when it is not.

**Central pain:**

- Described as sharp, shooting or burning pain, tingling numbness or throbbing
- It is very disturbing in nature.
- It can begin within weeks or months after the injury and is felt at the level of injury or below the level where there is no touch sensation.

**Segmental Pain:**

It often occurs slightly above the level of injury or slightly below. It is often associated with hyperalgesia in the painful region. Hyperalgesia is pain caused by something that does not normally cause pain. E.g., something cold, warm or very light touch to the skin can cause pain.

**Nerve root entrapment pain**

It occurs at or just below the level of injury and has a distant pattern like brief waves of stabbing or sharp pain or a band of burning pain at the point where the normal feelings stops.

**Pain due to syrinx:**

A Syrinx is a hollow fluid filled space in the spinal cord. It develops months and years after the injury. As the syrinx expands it may move up and down the spinal cord and can result in pain along with increased loss of sensory and motor function.

**Treatment:**

Neuropathic pain is very difficult to treat because each individual respond to treatment differently.

Pharmacological Treatment:

- Anticonvulsants
- Antidepressants

Non-Pharmacological Treatment:

- Exercise
- TENS
- Psychological counseling
- Relaxation techniques
- Graded motor imagery
- Breathing Exercises
- Cognitive behavioral therapy/ supportive psychotherapy

Surgical Treatment:

- Dorsal root rhizotomy
- Cordotomy
- Lumbar sympathectomy

Osteoporosis:

Osteoporosis occurs due to loss of mineral from the bone, causing bone tissue to break down. This occurs normally in individuals as they age. An individual's metabolism changes after suffering from SCI. Osteoporosis is one of the inevitable complications seen in individuals after SCI, and it is mainly seen in pelvis and lower extremities. Osteoporosis is due to prolonged bed rest, no weight bearing on lower limbs and lower motor disorder.

Causes:

- Disuse
- Disordered Vasoregulation
- Poor Nutrition
- Hormonal changes
- Metabolic disturbances
- Autonomic Dysregulation

Factors that affect Bone Loss:

- Level of Injury: The severity of the bone loss varies in paraplegics and quadriplegics. Paraplegics usually have bone mineral density preserve din their upper extremities.
- Type of Injury: Individuals with complete injuries have more bone loss than those with incomplete injuries.
- Duration of injury: the longer the time since injury, the greater the bone loss.

**How it is detected?**

- X-rays
- Bone Density testing ( Dexa Scan )
- Blood Calcium, phosphorous levels and blood alkaline phosphatase levels
- 1,25- dihydroxyvitamin D and calcitonin levels
- Urine Calcium levels

**Management:**

- Exercise: weight bearing exercises using standing frame or harness, treadmill training and functional electrical stimulation helps prevent demineralization of bones
- Increase dietary Calcium intake and Vit D levels: calcium helps build strong bones and Vit D improves the absorption of calcium
- Stop Smoking
- Limit Caffeine
- Stop Alcohol
- Medications: Biphosphonates can help prevent bone loss and increase the bone density
- Protect the bones: by doing regular ROM exercises. Spasticity can put some force on the bones that helps to strengthen them, however strong spasms can make the bones weak and hence make it more susceptible to fractures.

Note: Report any concerns to your physician immediately

**Heterotopic Ossification:**

Heterotopic ossification (HO) is a frequent complication in spinal cord injury (SCI). It was first described by Deejerine and Ceillier in 1918 as paraosteoarthropathy. It is characterized by the formation of new extra osseous (ectopic) bone in soft tissue surrounding peripheral joints in patients with neurologic conditions. Heteretopic Ossification is also seen in other neurologic conditions such as Traumatic Brain Injury (TBI) and Stroke, as well as after thermal injuries and other orthopedic procedures ( e.g., total hip replacement).

**Incidence:**

In SCI patients, the incidence ranges from 16%-53% depending on the incidence reports from various institutions. Clinically significant HO develops in about 20% of patients with a SCI. Fortunately, only 3-5% of cases involve joint ankylosis. In SCI, HO always occurs below the level of the lesion, most commonly at the hip (70-97%). Other body segments, including the knee, elbow, shoulder, hand, and spine (in decreasing incidence), may also be involved.

There is no known race or sex predilection for Neurogenic HO, however, the incidence of Neurogenic HO after SCI is less common in children than in adults with an incidence generally ranging from 3% to 10%. In addition, spontaneous resorption/ regression of the Neurogenic HO is often

reported in pediatric patients and young adults than in older adults. HO may develop within days following the spinal cord injury or several months later. HO usually occurs 3-12 weeks after spinal cord injury yet has been known to also develop years later.

**Causes:**

The specific cause of HO after spinal cord injury is unknown, however there are many theories about why it develops after spinal cord injury including:

- Trauma or injury
- Immobilization or paralysis
- Muscle spasms
- Loss of oxygen
- Severe bleeding
- Inflammation - the body's natural response to injury
- Genetics
- Prolonged pressure on the hips Symptoms:
- Decreased Range of the motion
- Swelling and warmth around the joint
- Fever
- Increased Spasticity
- Joint pain, Muscle pain, and Autonomic Dysreflexia

**Secondary Effects on the body from HO:**

It is a vicious cycle that affects the body after one or more joint of the body is affected with HO. HO causes restriction of the joint, affecting the ability to do transfers, bowel and bladder care and will overall affect the performance of ADLs. Also, HO can increase the amount of pressure applied to tissue under certain boney areas, skin breakdown can occur. Pain in the joint can lead to increased spasticity and/or autonomic dysreflexia. Blood clots or deep vein thrombosis (DVT) can also develop due to decreased movement and problems with circulation around the joints. One will have to work closely with a physician to treat HO and prevent its potential complications.

**How is it Diagnosed?**

HO is often diagnosed after you have some or all of the symptoms listed above. Your doctor will order some tests to confirm the symptoms are related to HO. Some of the tests that may be ordered include:

- Blood tests
- CT-scan
- Ultrasound
- Three-Phase Bone Scan
- X-rays

**How is it treated?**

- Work out a treatment plan with the physician and therapist
- Gentle range of motion of the joints and some physical therapy.
- Medications to slow down or stop the abnormal growth of bone.

When HO severely affects your movement or causes excruciating pain, surgery may be needed. Also, radiation has been used in some cases to stop the growth of bone. All other treatment options are explored first before considering surgery and/or radiation.

**Preventative Measures:**

Since the cause of HO is currently unknown, preventive measures are limited.

- Medications to prevent bone growth. The blood thinner Coumadin (Warfarin) is sometimes prescribed because it decreases the activity of Vitamin K, an important component in the development of bone.

- Another type of medication often prescribed is non-steroidal anti-inflammatory (NSAIDS) drugs. These medications prevent formation of bone growth by blocking prostaglandin cells from forming bone.
- Depending on the individual's health and needs, the physician will develop a plan that is right for you. All medications come with side effects, however never start or stop a medication without consulting with your doctor first.

**Contractures:**

What causes Contractures?

- Lack of active muscle function due to imbalance in muscle pull
- Spasticity and muscle spasms
- Flaccidity
- Improper positioning
- Heterotopic Ossification
- Edema
- These cause prolonged shortening of structures across and around the joints and hhese cause limitation of joint ROM

**How to manage contractures?**

- Normalization of Tone
- ROM exercises
- Stretching exercises
- Positioning in bed, chair etc
- Splinting

Bed sore management

Pressure Ulcers commonly known as bed sores are called as decubitus ulcers. It is defined as a localized area of tissue necrosis that tends to develop when soft tissue is compressed between a bony prominence and an external surface for a prolonged period of time.

Incidence: It is one of the most common complications seen in individuals after Spinal Cord injury along with Urinary Tract Infection. It is the most limiting squeal that can confine an otherwise independent individual to bed rest. But 39% of people re-hospitalized in the first year after their SCI are admitted for pressure ulcers. And about one-third, or 34% of people with an SCI end up requiring three or more hospitalizations throughout the rest of their lifetime for treatment of pressure sores. About 7-8% of deaths in the SCI population are related to a pressure sore. These deaths most likely result from sepsis, an infection that spreads throughout the body in the blood and tissues, causes of loss of income due to increased care costs, also causing prolonged bed rest and inactivity.

**Risk Factors:**

- Paralysis and sensory loss causing inability to feel irritants to the skin.
- Due to muscle atrophy there is less padding on the bony prominences, which causes more shear stress on these areas.
- Impaired sensation causing reduced blood supply to the tissues, which delays the healing process.
- Delayed wound healing due to altered collagen metabolism
- Low testosterone ( common in male SCI ) inhibits or delays wound healing
- Diabetes
- Malnutrition may cause impairment in the protein stores, vitamin C and zinc levels that are important to maintain the skin integrity.
- Smoking contributes to peripheral vascular disease causing delayed wound healing
- Aging causes the skin to get thinner and more prone to skin breakdown and less

tolerant to the shearing forces (dragging/ rubbing).

**What Causes Pressure Ulcers in SCI.?**

1)Pressure

- Persistent pressure for long duration: constant low pressure for long duration can cause tissue damage
- Recurrent pressure: e.g., bumping the elbow many times a day against the edge of a table
- Short periods of high pressure e.g., accidently hitting yourself hard against a surface while performing transfers.

2) Abnormal Shear Forces: dragging skin or body parts

3) Abnormal positioning while in wheel chair, vehicle or in bed can cause pressure on the bony prominences.

4) Exposure to moisture for long period can cause maceration and skin breakdown Common Areas for Pressure Sores

- Heels
- Sacrum
- Ischial tuberosity
- Hip bones
- Other Bony prominences like knees, shoulder blades, elbows, spine, pelvis, back of head etc.

**How to prevent pressure ulcers??**

- Inspect the skin for any signs of inflammation

**Methods to do Skin Checks**

- Should be done twice daily , first thing in morning and last thing before bedtime
- Use long handled/ angled mirror for areas that are difficult to visualize or ask for caregiver help
- Check for common areas of skin breakdown
- Check for signs of inflammation,
- If any areas are seen, perform blanch test
- Follow the instructions as to how to take care of various stages of pressure sores
- Consult physician, if the wound is oozing and has a foul smell.
- Perform regular pressure relief techniques, after every 15-30 min

**Pressure relief maneuvers:**

1. Push-ups after every 15 minutes by taking support of the armrest of the wheelchair. Triceps strength should be adequate for this maneuver.
2. Forward leaning: For active forward leaning trunk muscles, pectoral muscles, triceps are essential. However, if weakness persists, passively leaning forward can also relieve pressure to some extent.
3. Side lean by hooking on the rim of the wheelchair on the opposite side and leaning to one side and then the other. For doing this he needs strong biceps.
4. Electrically operated reclining wheelchair can also relieve the pressure.

- Frequent change in position in bed, turning almost after every 2 hrs in bed.
- Use of good wheelchair cushions and mattresses that evenly distribute the pressure
- Keep clothing wrinkle free
- Perform good transfer techniques with good pushups and ground clearance
- Protect the hands, legs and feet from trauma
- Proper posture and positioning in bed, chair
- Maintain proper skin hygiene after bowel

and bladder management

- Pat the skin dry
- Stop on habits like smoking and drinking
- Check testosterone levels every few years
- Diet high in protein, iron and vitamin C

What does it look like when the ulcer is starting?

- The skin gets reddish/ pinkish
- Shiny
- Skin texture changes- cracked, dry, hard, soft, warm

What to do if you suspect an Ulcer/Bedsore?

- Remove the pressure immediately
- Look for the source of pressure
- Perform blanch test: press on the discolored area, the skin should turn pale, remove finger and it will return to normal in a few seconds, if not that means the blood flow to that area is interrupted. And if the area is still discolored then that means the ulcer has started.

Note: What you see on the surface of the skin is often the smallest part of the pressure ulcer.

**Stage Wise Ulcer Management:**

Stage 1: Non Blanchable Erythema: in this stage the skin is intact with no blanching redness over the bony prominence. This stage is difficult to detect on dark skin tones. Individuals are at the risk of developing pressure sore. The skin is red and there is no change in color even after 30 min of pressure removal.

**Management:**

- Keep the pressure off the sore
- Maintaining good hygiene
- Evaluate the diet
- Review the cushions and mattresses
- Also review the turning, transfer and pressure relieving techniques
- If sore doesn't heal in few days even after following these measures, then consult your physician

Stage 2:Partial thickness: in this stage there is partial thickness loss of dermis with a red pink shallow open ulcer without slough

**Management:**

- Follow the same management of ulcer as done for the stage 1
- Keep the wound sterile and dry and use a hydrocolloid dressing or gauze dampened with saline.
- Check for signs of wound healing with every dressing change.
- If signs of infection are seen consult the physician.

Stage 3:Full thickness skin loss: there is full thickness tissue loss with visible subcutaneous fat and ample slough. The depth of the ulcer varies depending on the anatomical site of ulcer. The ulcer extends into the subcutaneous and fat tissues.

**Management:**

- Follow the same steps of maintaining wound hygiene
- Frequent wound cleaning and debridement should be done
- Antibiotics will be required
- Special pressure relieving mattress will be required

Stage 4: Full thickness tissue loss: the ulcer in this stage has extended to the level that underlying bone, muscle and tendon are visible. The depth of the ulcer varies depending on the anatomical site of ulcer

Management: wound is so big that a surgical

procedure like flap surgery will be required to close the wound.

Complications caused by Pressure sores:

- It can be a life-threatening event if infection spreads to blood, heart and bones
- Amputations
- Prolonged bed rest
- Autonomic Dysreflexia

**Spasticity Management**

The most important of the symptoms that requires a lot of attention and may hinder the day to day working of the patients is muscle spasticity and spasms. Various exercises and techniques can reduce and control spasticity. But sometimes spasticity can also be used by the patients to improve their functional performance. A therapist should therefore carefully assess the patient's functional abilities and how these abilities will be altered by spasticity before prescribing exercises to reduce tone. Patients should also take care to prevent some triggering factors that may increase the spasticity and spasms. Let us first understand what spasticity is and how it can be managed and used to the patient's advantage.

**What is spasticity?**

Spasticity is uncontrolled as well as sustained contraction of the muscles. It is very common in individuals with Spinal cord injury 8 out of 10 individuals with spinal cord injury have some grade of spasticity. Depending upon the level and site of injury the grade of spasticity may change. The spasticity is usually more if the injury is at higher level like that of cervical level and sometimes in the upper thoracic regions but as the level of injury moves lower the grade of spasticity also generally reduces.

**How to identify spasticity?**

Spasticity exhibits differently in different people and could present as a combination of symptoms that are mentioned below.

- The most common symptom is resistance to the movement in the opposite group of muscles leading to slow movements
- There could be sudden jerks in the muscles that have spasticity
- Spasms, i.e. sudden flexion (bending) and extension (straightening) of the whole body or some parts of the body
- Uncontrolled movement of the body while initiating a voluntary movement.
- Inability to control the speed and the direction of the movement of the limbs.
- Inability to relax the muscles leading to stiffness and tightness at rest.
- Resistance to a passive movement

**What is the reason for spasticity?**

Brain controls the body movements. These movements are generated in response a stimulus. However sometimes there is a need for a quicker response. Spinal cord is responsible for these quick responses and such responses are called reflexes. These reflexes are protective. An example of reflex is withdrawing the hand when touching a hot object. Such reflexes are inhibited by brain and therefore such sudden, violent and continuous muscle contractions are prevented. In spinal cord injury the communication between the spinal cord and brain is lost. Therefore, the muscles can be in a state of continuous contraction under the influence of uninhibited spinal cord. This gives rise to jerks, spasms and spasticity. Therefore, any external stimulus like touch, movement or any irritation can trigger and sustain an unwanted muscle contraction.

**What can trigger unusual responses of spasticity?**

- Suddenly stretching a muscle

- Initiating a body movement
- Skin irritation
- Pain
- Urinary infection
- Full bladder
- Constipation
- Bed sore
- Tight clothing
- Uncomfortable clothing causing skin irritation in the area with impaired sensations
- Uncomfortable footwear
- Fracture, muscle, tendon or ligament injury

**What are the harmful effects of Spasticity?**

- Pain
- Contractures i.e. loss of joint mobility due to muscle stiffness. If this persists adhesions can be formed in other tissues like fascia, ligaments and other connective tissues. Skin shortening may also occur. This can result in permanent loss of range of motion in a joint requiring surgical intervention.
- Sudden and uncontrolled spasms are a risk to the safety while performing transfers and gait training. A spasm compromises the safety while driving.
- Breathing may be restricted due to spasms and spasticity in the chest muscles; this may further reduce the endurance for performing day to day activities as well as training exercises.
- Sleep disturbances
- Skin irritation and abrasions
- Difficulty in performing movements of daily activities.

**What care to take for preventing harmful effects of spasticity?**

- From the safety perspective it is important to anticipate spasms when driving on an surface or wheeling the wheelchair on an uneven surfaces. Adequate safety measures like putting the seat belt and wheelchair belt should be followed. In case of severe spasm, it is advisable to halt the car by the side of the road till the spasm wears off.
- During therapy, therapists should be aware of the movements trigger spasms and should avoid any unnecessary jerky movement and sudden stretching of the muscles. Also, during training for transfers and gait training adequate safety measures should be taken to prevent falls in case of spasms.
- Regular passive stretching of the muscles is essential to avoid joint stiffness and muscle contractures.

**How can Spasticity be used for the benefit of the patient?**

- Spasticity can be used to trigger muscle contraction and thereby to perform certain tasks of daily activities as shown in the table below. Certain muscles can be triggered to give rise to the spastic contraction helping to perform a task.

| Muscle | Trigger movement | Functional movement |
|---|---|---|
| Finger flexors | Wrist extension | Gripping an object |
| Hip flexors | Backward or sideways leaning of the trunk | Walking |
| Trunk flexors | Pushing back with head | Supine to sit |
| Elbow flexors | No particular trigger movement | Eating |

- In absence of pain spasticity is a warning sign for infection, pain, bed sore and bowel or bladder infection and brings about the attention to such issues which can be treated promptly.
- Spasticity is also a sign of bowel and bladder fullness and therefore can be utilized functionally for toileting activities.

**Management of spasticity**

Spasticity management is multidisciplinary and may require medical, surgical as well as rehabilitative intervention. It is crucial to understand what treatment should be one at what time, for optimal outcome.

- Early phase of spasticity -
  - Grade 1 and1+ on Modified Ashworth scale

Prescribe regular passive slow and sustained active or passive stretching exercises, strengthening of antagonists, facilitation of active movements primarily with antagonists and weight bearing exercises to control the spasticity.

If the antagonists can be trained in weight bearing position after the stretching has been performed it may give optimum results.

  - Grade 2 and 3on modified Ashworth scale

Oral medicines should be given in addition to physiotherapy exercises. For oral antispastic medication refer the patient to a physician or neurologist with a detailed evaluation of spasticity. Dosages of oral medication should be titrated to get optimal results and therefore advice your patient to keep up with the follow up appointments. A therapist should also carry out follow up evaluation of the patient's spasticity and consult with the concerned physician.

Before starting the oral medication rule out any triggering factors for spasticity like urinary tract infection, urinary blockage, systemic infections, bowel impaction, bowel abnormalities and bed sores. If any of these triggering factors are detected it is important to manage them medically before increasing or starting the oral medication.

  - Grade 4 on modified Ashworth scale

When the affected part is rigid and passive movement is impossible or very painful. It is important to address the spasticity in the early phase itself before tissue shortening occurs. Intramuscular injection of botulinum toxins may be considered for this. This will also ensure inhibiting pathological movement patterns caused by spasticity as well as tissue contractures.

Physiotherapists should take care to not over stretch severely spastic muscles to avoid muscle and soft tissue tearing and damage.

- Chronic phase spasticity
  - Grade 1 and 1+ on modified Ashworth scale

Continue with rehabilitation exercises and titrate the dose of oral medication.

  - Grade 2 and Grade 3 on modified Ashworth scale

Rehabilitation management and medical management must be adhered to, to prevent the harmful effects of spasticity. Weight bearing exercises and slow and sustained active or passive stretching should be the mainstay for spasticity reduction.

Intramuscular botulinum toxin injections can be considered even in this stage. The muscles should be carefully selected to break the synergic movement patterns, which by now are learned as new motor patterns of the movement. After the injection physiotherapist must retrain the motor patterns to break the synergic movements and rigorous slow and sustained active stretching should be taught to the patient and

patient's relatives or caretakers. Weight bearing exercises should also be emphasized. Strengthening of the antagonists is very important in this phase and active strengthening should be compared with the weight bearing to inhibit over activity of the agonists with spasticity.

With higher grades of spasticity at a chronic phase it is important to identify the time for stopping the treatment due to non-responsiveness. For example, if there is generalized spasticity which has not reduced with oral medication, oral medication can be discontinued. The decision to discontinue the medication must be taken after consulting with the prescribing physician.

Repeated injections of botulinum toxin can be considered in case of smaller number of muscle groups. However, if there is generalized non responsive spasticity then intrathecal baclofen can be considered. This decision should be taken as a multidisciplinary team in consultation with physician and neurosurgeon. Generally, the pump is considered within one year of the injury. Although in incomplete spinal cord injury if initiation of voluntary movement is limited or inhibited by spasticity an earlier installation of pump device or earlier injection may be considered.

- Grade 4 spasticity on modified Ashworth scale

For rigidity unresolved by medical and rehabilitative management surgical management like rhizotomy and baclofen pump installation can be considered. This decision should be taken in consultation with treating physician and surgeon.

**Neurorehabilitative management of spasticity**

Neurorehabilitation of spasticity is multidisciplinary and involves good nursing care, physiotherapy an occupational therapy intervention and psychological counseling to the patients and family education.

**Nursing care:**

1. Preventing pressure sores by regular skin inspection and decompression of the pressure areas.
2. Appropriate positioning in bed and wheelchair
3. Good techniques for catheterization and monitoring patient's catheterization technique.
4. Monitoring sleep patterns
5. Monitoring pain

**Physiotherapy intervention:**

1. Slow sustained passive stretching and active stretching whenever possible.
2. Teaching correct positioning to caregivers and relatives
3. Facilitating antagonists active movements
4. Inhibiting the synergy patterns
5. Weight bearing exercises
6. Use of ice to reduce spasticity and facilitate voluntary control there after

**Occupational therapy intervention:**

1. Teaching correct patterns for performing activities of daily living and activities relate to personal hygiene
2. Devising an exercise program with the physiotherapist to reduce spasticity so that functional movements can be trained

Spasticity management should be undertaken by multidisciplinary team of professionals including physician, nurse, physiotherapist, occupational therapist, urologist, psychologist and care givers. A good communication within the team is key to facilitating appropriate management strategies at the correct time. Patient's counseling regarding the adherence with the treatment is very important as spasticity

management is a long-term process.

**Bladder and Bowel management**

Spinal cord injury at any level almost always affects bladder and bowel function. This is because of injury to the spinal cord and there is cut off from brain input. A wide range of techniques and tools are available to manage bladder and bowel function.

Changes in bowel function after SCI has a considerable impact on the quality of life of individuals with spinal cord injury. Bowel problems experienced by spinal cord injured patients include prolonged evacuation time, poorly localized abdominal pain, hemorrhoids, abdominal distension, fecal incontinence, and chronic constipation. Bowel Dysfunction is the most significant problem than loss of ambulation in SCI and managing this change is important for an individual's independence and autonomy. Therefore, it is an important area of care and rehabilitation for individuals with SCI for community reintegration and long term health.

**Bowel Management:**

Injury to the spinal cord has a profound impact on the function of the large bowel and on the maintenance of fecal continence. The following are the reasons of bowel incontinence in SCI

- Slow transit of stool through the bowel causing constipation
- Loss of sensory and motor control of ano-rectum causing an inability to feel the need to evacuate the bowel

With no intervention the individual will be chronically constipated with all the secondary complications, hence it is important to manage the function of the large bowel and minimize the health issues.

Keeping in mind the rehabilitation of an individual, the bowel management programme is a process that focuses on the needs and promotes autonomy in an individual with SCI... The individualized programme is supported by educating the injured individual, their family and any carers who may be involved in that person's care. Bowel management program is an ongoing process for lifetime that changes according to the needs of an individual.

**Aims of Bowel Management:**

- To enhance safety, privacy, comfort and dignity of SCI individuals during bowel management episodes
- To assess the needs of the SCI individual in regard to bowel care and to develop and maintain an appropriate individualized bowel management routine.
- To avoid incontinence, constipation, diarrhea and autonomic dysreflexia, and minimize the development of secondary ano-rectal complications associated with bowel management (rectal over-distension, hemorrhoids, anal fissure etc.)
- To achieve a satisfactory bowel management with minimum necessary physical and pharmacological interventions before the discharge from initial rehabilitation
- Identify appropriate transfer methods, equipment and adaptive devices to promote independence in bowel management for the individual.
- To evaluate the outcomes of bowel management. The Bristol Scale. Duration of bowel management episodes and any unplanned bowel evacuations should be recorded
- To educate the individual with a 'toolkit' of knowledge with which they can manage and adapt their bowel care in the long-term

Assessment of the individual, planning interventions and evaluation of outcomes is required for developing an individualized

bowel management program. The need for a bowel management programme after spinal cord injury is life-long, and regular re-evaluation and modification of the programme will be required over the years post injury.

Factors to be considered while assessing an SCI individual for their bowel management and rehabilitation:

- Current bowel function
- Pre-injury bowel habit
- Past medical history - including obstetric history, chronic bowel disease, cancer, any abdominal surgery
- Medication
- Diet and fluid intake
- Ability to eat a full diet
- Activity, general mobility, exercise, standing, passive movements
- Communication
- Cognitive ability
- Level of independence
- Lifestyle - cultural, sexual, work or educational roles
- Psychological and emotional factors
- Manual handling risk assessment
- Home and care circumstances

Outcome measures for evaluating bowel management are:

- Duration of management episodes
- Stool form recorded using the Bristol Scale
- Frequency of episodes of incontinence
- Have pharmacological interventions been minimized as far as possible?
- Is the patient as independent as possible?
- Feedback from the patient regarding satisfaction and perceived autonomy

How Frequently of bowel care should be re-evaluated for an individual undergoing initial rehabilitation or in community living?

- After the stoppage of spinal shock phase
- When starting with mobilization
- When the patient is ready to begin bowel management on the toilet
- During general routine follow up
- If duration of management regularly exceeds an hour following a sudden change in bowel function i.e. Incontinence, change in stool form, color or odor or bleeding per rectum and if these changes persist for more than 4 weeks and have not responded to changes in the usual programme may be indicative of bowel cancer and should be investigated immediately. Rapid appropriate referral for investigation is essential.
- In the light of changing levels of independence/dependence

**Importance of early bowel care for an individual with SCI**

For an early reintegration into the society following an SCI an optimal bowel function and management is required in the longer term immediately following an injury. Prompt and effective bowel management will avoid over distension of the bowel with constipated stool which is thought to result in damage to stretch receptors which in turn may adversely affect the return of reflex ano-rectal activity, decreasing the future capacity for effective reflex evacuation of stool.

**Assessing Bowel Function:**

Individuals with complete spinal cord injury will have no sensation of the need for evacuation of the bowel and no control of the bowel. Depending on the level of injury there are different types of bowel seen

- Reflexic Bowel (also called upper motor neuron or spastic): seen usually in injuries at or above T12 vertebrae. The bulboanal reflex and the anal wink are present if the patient has a reflexic bowel.
- Areflexic Bowel (also called lower motor neuron or flaccid): usually seen in injuries at or below the L1 vertebrae. The bulboanal reflex and the anal wink will be absent if the patient has an areflexic bowel.

For incomplete SCI, the residual bowel function is less predictable and will be established only through examination by a medical practitioner.

**Bowel Management Program:**

The bowel management programme will specify the following:

- Specific interventions to be used
- The frequency (twice daily, daily, alternate day) and timing (morning, evening) of the bowel program.
- The location for bowel management (bed, toilet etc.)
- Who will perform bowel care (the patient, a specific relative, care staff, community Nurse)
- Equipment and adaptations required (shower chair, padded toilet seat etc.) Factors to be considered when forming a Bowel Programme.
- Level of injury
- Type of Bowel Dysfunction
- Evaluation of bowel care during the acute stage
- Mobility of the individual
- Limiting factors such as spasticity and poor balance
- Self-care abilities of the patient
- Age of the patient
- Weight of the patient
- Cultural requirements
- Domestic circumstances
- Availability of carers
- Availability of bowel care aids and equipment
- Accessibility and suitability of facilities at discharge destination
- Any pre- or post-injury bowel related complications
- Pre-injury bowel habit
- Relevant medical history
- Prevention of dysreflexia in susceptible patients
- Supervision requirements (is it safe to leave the patient during bowel management episodes?)
- Planned future family commitments, employment or education
- Other lifestyle considerations

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- Prevention of dysreflexia in susceptible patients
- Supervision requirements (is it safe to leave the patient during bowel management episodes?)
- Planned future family commitments, employment or education
- Other lifestyle considerations

**Guidelines for patients:**

- Patient education regarding the bowel management, its importance, obtains an informed consent. Also, if patient is unconscious Gain verbal consent perform procedure as long as it is in the best interest of the patient after discussion with the multidisciplinary team

- Ensure privacy for performing bowel management and that the individual's dignity is maintained at all times. Consider the practicality of moving the patient (on his bed) to a more private area for bowel care
- Procedure should be performed only by a competent nurse.
- Remove feces from rectum prior to inserting prescribed mild rectal stimulant to assist in removing feces.
- During spinal shock use mild rectal stimulant to lubricate the stool prior to digital evacuation if constipated, or to aid in release of flatus in individuals with any type of bowel function
- Wait for approx. 20 minutes and allow the mild rectal stimulant to work - time bowel management to coincide with log rolling the patient for hygiene and skin checking suppositories can be given whilst patient is flat on bed and bowel emptying can be performed when rolled.
- Maintain spinal alignment at all times and ensure correct positioning when finished
- Check skin around peri-anal area and make sure skin is cleaned and dried adequately and use barrier creams if necessary, especially if bowel movements are loose
- Record bowel management interventions used, duration

Note: A routine and timely follow-up for bowel care is required to achieve an early identification of potential problems, and to promote timely intervention in order to reduce the incidence of chronic bowel complications.

**Reflexic Bowel Management:**

- Establishing a Routine
- Diet and Fluids
- Digital rectal stimulation
- Pharmacological rectal stimulation: suppositories, micro enemas
- Oral laxatives

**Areflexic Bowel Management:**

- Establishing a Routine
- Diet and Fluids
- Manual removal of stool from rectum
- Abdominal massage
- Valsalva maneuvers/straining Non-conservative options:
- Trans-anal irrigation (Rectal Irrigation/Anal Irrigation)
- Ante grade Continence Enema (ACE)
- Percutaneous Endoscopic Colostomy (PEC)
- Colostomy
- Sacral Anterior Root Stimulator (SARS)
- Sacral Nerve Stimulation (SNS)

**Diet and bowel management in spinal cord injury**

Food and fluid intake have a major effect on the bowel actions that affect the bowel management. Fecal weight and consistency is affected by the amount and water- holding capacity of the remaining undigested material that passes into the colon, the amount of bacteria present and colonic transit times. Diet assessment and alteration in diet is required to help maintain appropriate stool consistency while maintaining a balanced diet for life.

Dietary factors that affect the stool consistency:

- Enteral tube feeding
- Fluids
- Manipulate diet to alter stool consistency

**Bladder Management:**

How the bladder normally works:

The Urinary System has three major functions to perform:

- Makes Urine in kidneys
- Stores Urine in bladder
- Expel urine from body through urethra.

**How the bladder works after SCI?**

After SCI the bladder is usually affected in one of two ways depending upon the level of injury:

- Spastic (reflex) bladder/ Upper Motor Neuron type: seen in injury above T12 level. In this type, when your bladder fills with urine and a reflex automatically triggers to empty the bladder. With a spastic bladder you do not know when, or if, the bladder will empty.
- Flaccid (non-reflex) bladder/ Lower motor Neuron type: seen in injury below T12 level. In this type the reflexes of the bladder muscles are sluggish or absent. You will not feel when the bladder is full and the bladder can become over-distended or stretched. The urine can back up into the kidneys (called reflux). Stretching also affects the muscle tone of the bladder.
- Dyssynergia occurs when the sphincter muscles do not relax when the bladder contracts. The urine cannot flow through the urethra, which can result in the urine backing up into the kidneys. The bladder may also not empty completely.

Bladder Management: A bladder management is important to empty your bladder when it is easy for you and helps you avoid bladder accidents and prevent UTIs.

Points to keep in mind when following a bladder management:

- Preparing and following a regular schedule
- Emptying the bladder completely without any residual Nonsurgical Management for Bladder Dysfunction:

1. Continuous Intermittent Catheterization (CIC)

- Urine is drained out of the bladder using a catheter on regular fixed intervals (usually every 4 to 6 hours).
- Advantage: - No need to wear a catheter and urine bag all the time. It is done on need basis. It is more convenient and healthier and UTIs are less common with this method.

2. Indwelling catheter (Foleys): A catheter is left in place in bladder along with a bag for urine collection. There are two basic types of indwelling catheters:

- Urethral indwelling catheters are inserted into the bladder through the urethra. This is a good option for people with poor hand functions or who have limited assistance.
- Suprapubic indwelling catheters are inserted into the bladder through a surgical opening in the lower abdomen, just above the pubic bone. It's recommended for individuals with bladder neck obstruction or urethral abnormalities.

Complications of Indwelling Catheter: bladder or kidney stones (especially in females), deterioration of the upper urinary tract (including increased reflux), and increased risk for bladder cancer especially used for longer than 10 years.

3. External condom catheter (for men only): condom catheters are worn, and it is attached to a bag that collects the urine.
4. Crede Maneuver is used to empty the bladder by manually applying pressure to the lower abdomen, with a closed fist to manually push the urine out through the

urethra. This method is used when the bladder cannot contract on its own or has weak contractions and needs extra pressure to push out urine.

5. Valsalva maneuver: is used in flaccid type bladder. In this type abdominal muscles are used along with diaphragm (the muscles directly below your rib cage) to push down on the bladder in order to empty it.

6. Reflex Voiding: it's used by males with an overactive bladder. There are involuntary contractions of bladder. After SCI, a contracted bladder muscle is not always coordinated with a relaxed sphincter muscle. So, the sphincter relaxes and tightens intermittently, the flow of urine stops and starts involuntarily and thus unpredictably. Reflex voiding can be used in combination with intermittent catheterization or and external condom catheter.

Medical Management of spinal cord injury

1. Alpha Blockers: Helps to improve urination in males who have a lack of coordination between the bladder and sphincter by relaxing the sphincter and prostate. It helps to lower the pressure in the bladder during voiding, relaxes certain muscles and helps small blood vessels remain open and relaxed. This improves blood flow and lowers blood pressure.

2. Anticholinergic Drugs: Commonly prescribed for people who use intermittent catheterization. These drugs help relax the bladder, thus increasing its capacity and reducing contractions that might cause leakage. They work by blocking certain nerve signals that activate muscle activity. Side effects of anticholinergic drugs can include dry mouth, blurred vision, drowsiness, and constipation.

Note: Complications like high bladder pressure causing back pressure on the kidneys, abdominal bruising when using the Crede method, hernia, pelvic organ prolapses, or hemorrhoids are all associated with Crede and Valsalva. They are generally not recommended as primary methods of bladder emptying.

Surgical Management for Bladder Dysfunctions:

- A Mitrofanoff procedure: in this procedure a new passageway for urine is constructed using the appendix, which allows catheterization to be done through the abdomen to the bladder. This procedure is of great advantage for women and for individuals with limited hand function.
- Bladder augmentation/ Cutaneous Ileovesicostomy: this procedure is done to reduce the need for frequent catheterization by surgically enlarging the bladder (using a portion of the intestines).
- Sphincterotomy: Transurethral Sphincterotomy (TURS) is done to reduce the pressure on the valve and allow easy flow of urine out of the bladder. This operation is not normally carried out on women.
- Botulinum Toxin injections: Can be used by individuals using intermittent catheterization to relax an overactive bladder. Botox is injected directly into the bladder muscle, effectively reducing contractions increasing bladder volume and thus reducing leakage.
- Functional Electrical Stimulation and Posterior Sacral Rhizotomy

Common Issues and Concerns with bladder management:

1. Residual Urine: it is seen when there is inadequate bladder emptying causing some amount to be left in the bladder. This is a major cause of UTIs in individuals

with SCI.

2. Urinary Tract Infection: Individuals with SCI are at a high risk of developing UTIs. Bacteria from the skin and urethra get into the bladder with all forms of bladder management.

**Symptoms of UTI:**

- Fever
- Chills
- Nausea
- Headache
- Increased Spasms
- Autonomic Dysreflexia

In incomplete injuries one may also be able to feel

- Burning while urinating
- Pain/ Discomfort in Pelvic area
- Abdominal Pain/ Discomfort
- Lower Back Pain/ Discomfort

**Warning Signs of UTI:**

- Sediment(gritty particles) or mucus in the urine
- Cloudy Urine
- Foul odor of Urine
- Blood stained Urine ( pink or red urine)

Note: One may experience one or more signs/symptoms when having an UTI. An immediately consult your Urologist

Preventing UTIs: UTIs can be prevented by keeping your urinary system "clean"

- Clean the skin regularly
- Wash your hand before and after performing catheterization
- Wash area around the genitals regularly with soap and water
- Change the linen after every urine or bowel accidents
- Drink plenty of fluids helps in washing out the bacteria from the bladder which can help prevent UTIs and other problems of urinary system. Individuals undergoing intermittent catheterization are advised to take 8-10 glasses of liquid per day( primarily between breakfast and dinner)
- Stop drinking beverages with sugar, caffeine and alcohol
- Frequent catheterization for at least every 4-5 hrs. Treatment of UTI:

**Medical Management: Antibiotics**

Note: If UTI is left untreated, bacteria can travel further up multiply, and infect the kidneys. Kidney infection was once a leading cause of death after spinal cord injury and is still a danger.

3. Autonomic Dysreflexia: AD is often triggered by an overly filled bladder or a bladder infection in individuals with SCI at or above T6 levels. It is an important concern for individuals with SCI. Autonomic dysreflexia is a potentially dangerous spike in blood pressure that can be accompanied by pounding headache, sweating, "goose bumps," and blurred vision.

4. Bladder cancer is another concern for some individuals with spinal cord injury. Research shows an increase in the risk of bladder cancer among those who have been using indwelling catheters for a long period of time. Smoking also increases the risk for developing bladder cancer.

**Things to be done**

1. Regular Urologic Check Up: A complete medical check-up is recommended at least once a year. This should include a urologic examination, including a renal scan or ultrasound to know the kidneys are

working properly. The examination may also include a KUB, an X-ray of the abdomen that can detect kidney or bladder stones. An Urodynamic study that will tell how much urine your bladder can hold and how much urine in the bladder can cause the bladder to reflex. Even with a regular bladder management program and proper prevention methods, the risk remains for urinary tract infection. Treatment for a UTI almost always includes an antibiotic medication prescribed by a doctor.

2. Cleaning and Storing the Urinary catheters

**Rubber Catheters**

- After using the catheter, wash it with warm soapy water, rinse under warm running water, lay it on a clean towel to dry and then place it in a plastic bag.
- After you return home, the rubber catheters should be placed in a pan of water on the stove and boiled for 20 minutes, then placed on a clean towel to dry and then stored in a plastic bag.
- These catheters should be cleaned as instructed above after each use. One can continue to use these catheters until signs of wear such as discoloration or cracking is noted.

**Long or Short Clear Catheters**

- After using the catheter wash it in warm soapy water, rinse under warm running water, lay it on a clean towel to dry and place it in a plastic bag.
- After you get home, the catheters should be cleaned again in hot, not boiling, soapy water, rinsed well under hot running water and place on a clean towel to dry. Then place in a plastic bag for storage.
- These catheters should be cleaned as instructed above after every use. One can continue to use these catheters until signs of wear such as discoloration or cracking is noted.

## BED MOBILITY TRAINING

Bed mobility training is the first step towards independence for the patients of spinal cord injury. The patient has to be educated about the importance and benefits of bed mobility. From not being able to move the body at all to managing all the activities in bed independently boosts the confidence. Bed mobility training strengthens the muscles and improves trunk control which will help in increasing the endurance for performing subsequent tasks of transfers and walking.

What is the use of all strengthening exercises if at the end of the day patient still needs assistance to get up and sit?

Therefore, this chapter will be a step by step guide for training of the bed mobility activities. The sequence in which these are given should be followed but alterations can be made based on therapist's judgment. Although these are in sequence the activities should be more or less trained simultaneously and the therapist need not wait for the completion of one activity before starting to train for the next.

Some of the points to note for the therapists are

-muscle strengthening exercises should be performed at the same time as that of the training for bed mobility

-Bed mobility exercises should be functional and compensations by patients can be allowed or encouraged. Compensation can facilitate the firing of muscles to begin with and the movements can be refined later. Repetition of a movement prompts patients to come up with their own compensatory strategies. These compensations can be encouraged to achieve greater functional independence in view of stagnant motor recovery

- care should be taken to not encourage abnormal postures and sudden explosive movements
- Repetition is the key

**1. Rolling**

It is one of the most important and useful activity as rolling can help in preventing the potentially fatal complication of spinal cord injury i.e bedsore. Rolling to one side every 30 minutes helps in relieving pressure on their back reducing the risk of developing bedsore.

There are mainly two components in rolling. Rolling to turn to one side and rolling to turn to prone position. The first task is rolling to the side. The technique to teach the patient is to roll from one side to the other using the momentum of arm movements (Fig 1 and 2).



*Figure 1 - Side to side movement of arms to facilitate rolling*



*Figure 2 - Side to side movement of arms to facilitate rolling*

**Log rolling to the side:**

It can be performed in the acute phase but to ensure safety and prevent torsional forces on the surgical fixation a spinal jacket/brace is used. Protective brace can be used for first 3 to 6 months after injury to perform log rolling and can be later removed after consulting the surgeon. Log rolling is performed by the patient and the therapist together. Patient should be advised to move the arms with force and speed from one side to the other, 2 to 3 times and to then to use the momentum to turn to one side. The therapist should ensure that the lower body is turned at the same time to prevent torsional forces. To make it easier to turn the body the contralateral knee should be bent (Figure 3 and 4).



*Figure 3 - Contralateral knee flexion during log rolling*



*Figure 4 - Log rolling with therapist assistance*

**Segmental rolling:**

Segmental rolling is a technique where the upper and lower part of the body rolls separately one after the other creating a twisting action at the spine (Figure 5 and 6). Segmental rolling is to be encouraged only after the initial 3 to 6 months of the injury, as the torsion force created by the movement can damage the surgical fixation performed on the spine.



*Figure 5 - Assisted Segmental rolling*



*Figure 6 - Assisted Segmental rolling*

To facilitate rolling first let the patient perform sideways arm movements as shown in figures 1 and 2. If the patient is unable to roll with this technique, weights can be added to the arms to increase the momentum Figure 7 and 8.



*Figure 7 - Sideways movement of arms with weight cuffs to facilitate rolling*



*Figure 8 - Sideways movement of arms with weight cuffs to facilitate rolling*

In case of cervical injuries where the triceps are not working, patients can wear elbow splints to keep the arms straight for gaining momentum. If there is severe adductor weakness patients will not be able to hold the position despite wearing the elbow splints. In such cases strengthen shoulder adductors to improved arm swing. This movement strengthens the shoulder muscles and an alternate pattern for movement is generated for turning. Once this pattern is learnt the splints can be removed and patients can turn on their own (Figure 9 and 10).



*Figure 9 - Turning to one side without splints or assistance by a patient with quadriplegia*



*Figure 10 - Turning to one side without splints or assistance by a patient with paraplegia*

**Rolling to prone:**

The progression from side rolling is rolling to prone position. Care should be taken to lift the arms above the head while rolling, to clear the arms from under the body while

turning. Therefore, therapists have to take extra care with cervical injury patients while performing this activity as it can cause excessive pain in shoulder if it gets stuck under the body. If the shoulder flexion range is restricted to 100 degrees or less the therapist can place the ipsilateral hand close to the body and assist the patient into log rolling using one arm; instead of using both arms to swing.

2.Supine to side lying to long sitting

The next step after rolling is to be able to sit up from supine. There are two techniques for this, turning onto one side to sit up and sitting up straight from supine.

Once the patient learns to turn to one side, patients can sit up using elbow extension where the triceps is preserved. The therapist should strengthen triceps, all the three parts of deltoids, side flexors of the trunk and abdominals to facilitate ease in performing this movement.

Patients with higher thoracic injuries may also be taught this technique for better stability.

For quadriplegics with preserved triceps muscle the same technique can be used but for patient without triceps the task becomes really challenging but not impossible. The therapist can still train the patient to get up and sit by strengthening shoulder rotators, deltoid, adductors, biceps, wrist extensors and upper abdomen. Therapist has to teach the patient to weight shift, how to use head and neck muscles and upper back muscles. Since there is no triceps it becomes impossible to push up when you are on your elbow on side lying position. If you guide patient to shift weight on the other arm using head and neck muscles, the patient can straighten the elbow and lock it using shoulder rotators.

Key- Repetition!!

Figure 11, 12 and 13 show a patient with quadriplegia resulted from injury at the level C6-7

performing this movement



*Figure 11 - Supine to side lying*



*Figure 12 - Pushing on the elbows using triceps*



*Figure 13 - Long sitting come to long sitting*

For lower thoracic and lumbo-sacral injuries one arm can be used to push up as the trunk strength and stability is fairly good.

Figure 14 to 18 show a patient with paraplegia performing the movement of side lying to sitting

**3.Supine to long sitting**



*Figure 14*



*Figure 15*



*Figure 16*



*Figure 17*



*Figure 18*

***Figure 14 - 18 Patient transitioning from supine to side lying and then pushing on the elbow unilaterally to come up to long sitting.***

Patients can choose to sit up from the supine position itself, making use of the shoulder rotators and triceps in case of cervical injuries. In case of higher cervical injuries where the triceps is not preserved biceps can be strengthened so that patients can pull themselves up to sitting. A rope, side rail or human support can be given so that patients can pull onto it sit up. In thoracic and lumbosacral injuries patients can sit up using the triceps, shoulder rotators and upper abdominals.



*Figure 19*



*Figure 20*



*Figure 21*

***Fig 19- 21 Patient transitioning from supine to long sitting***

**4. Balance training in long sitting**

Achieving the position of long sitting can be challenging for the patients with higher thoracic and cervical level injuries due to weakness and impairment of the back extensors. As the legs are stretched the pelvic position can only be controlled by the spinal muscles. To be able to sit erect keeping the center of gravity within base of support too much slouching is not helpful. Therefore training in long sitting is very crucial for patients with higher level injuries.

For patients with quadriparesis - Training the patient to move forward and backward in the bed is difficult but can be achieved with training. Therapist has to strengthen upper thoracic and shoulder muscles for better hip clearance.

Therapist has to guide the patient, to sit in long sitting position bending slightly forward, with arms kept at the level of the shoulder, shoulder rotated and elbow locked in extension. In this position ask the patient

to bend further to put weight on both the arms and try to lift the hip up. (Figure 22)

If the shoulder rotators and adductors are weak another technique can be used One stool should be placed on both the sides of the patient. The stool height should slightly exceed his elbow level from surface of the bed. Now ask the patient to keep elbow and forearm on the stool bend forward putting weight on the elbows and try to lift the hip.

Once they achieve hip clearance moving forward and backward becomes easier and this also reduces risk of developing bedsore due to prolonged sitting as they can now push up independently and relieve ischial and sacral pressure.

NOTE- Stretching of hamstring and dorso-lumbar fascia can help in better performance.

paraplegics-Training the patients to move in the bed forward, backward, sideways and in circles improves their mobility in bed and the spine mobility as well as stability. Initially the movements can be performed using the push up bars and then slowly as the shoulders and elbow muscles strengthen perform these movements without push up bars.

Balance training in long sitting must include movements that move the pelvis and hip joints through the whole range of motion and in various directions.

Long sitting facilitates stretching of hamstrings and helps to reduce the flexor spasticity of the lower limbs



*Fig 22 - Rotation from one side to another with dumbells*



*Fig 23 - Rotation from one side to another with dumbells*



*Fig 24 - Lifting bolster in long sitting*



*Fig 25 - Arm flexion with side rotation with weights*



*Fig 26 - Arm flexion with side rotation with weights*

**5. Balance training on the edge of the bed**

Once the patient is sitting on the bed i.e. long sitting is achieved ask the patient to grip the legs with arms and move the led sideways. After repeating this couple of times, the legs can be pushed out of the bed to sit on the edge of the bed. Sitting on the edge of the bed should only be facilitated after training for balance in long sitting.



*Fig 27 - Forward bending in Long sitting*



*Fig 28 - Backward bending in Long sitting*



*Fig 29 - Patient transitioning from side lying to sitting on the edge of the bed*



*Fig 30 - Patient transitioning from side lying to sitting on the edge of the bed*



*Fig 31 - Patient transitioning from side lying to sitting on the edge of the bed*



*Fig 32 - Patient transitioning from side lying to sitting on the edge of the bed*

Balance training at the edge of the bed is extremely important for functional tasks as well performing exercises in sitting. In the traumatic injury patients are apprehensive of falls. In training balance there are two components involved strengthening of the muscles and developing new motor patterns.

The strengthening of 2 group of muscles is required, stabilizers and movers. Stabilizers are trunk and proximal muscles that will provide stability for the peripheral joint movements. For balance training key muscles are the back extensors and scapular retractors. If these muscles are weak patients can attain balance through slouching which may lead to awkward bending of the spine and scoliosis.

Therefore, training for balance with active exercises in sitting should be combined with strengthening of back extensors and scapular muscles in lying down and sitting position.

Factors that hinder the sitting balance are the spasms and tightness of the muscles of lower extremity. Spasm can give rise sudden push or pull difficult to counter if the muscles are weak and if the patient is unable to anticipate the jerk. Balance training with perturbations either by therapist or generated through activities like throwing and catching, rebound from the spring activities etc. should be incorporated in the balance training.



*Fig 33 - Arm flexion and extension sitting at the edge of the bed without support*



*Fig 34 - Arm flexion and extension sitting at the edge of the bed without support*



*Fig 35 - Arm flexion and extension sitting at the edge of the bed without support*





*Fig 36 - Trunk rotation with weights sitting*



*Fig 37 - Supported forward bending and sitting up at the edge of the bed without support*



*Fig 38 - Supported forward bending and sitting up at the edge of the bed without support*



*Fig 39 - Supported forward bending and sitting up at the edge of the bed without support*

**Prone extension-**

Prone extension exercise should be performed only after consulting with the surgeon in the acute phase of spinal cord injury. Due to the extreme extension motion there is a risk of loosening the surgical fixation if performed before the bone healing can take place. This posture should also be avoided for the lumbar spinal cord injury patients as it may loosen the surgical fixation. Although functionally not essential the prone extensions must be trained to strengthen the upper extremity muscles, back extensors, scapular muscles and pectoral group of muscles and to maintain the joint range of motion of the spine.

For Patients with lumbar spinal cord injury- Prone on elbow will serve best to achieve strengthening of similar group of muscles except triceps. Therapist can strengthen triceps separately in such cases.







*Fig 40 to Fig 42 - Prone extension from Supine position*

**6.Prone to quadruped**

To build a good control of the shoulder, weight bearing exercises such as quadruped help a lot.

Especially in higher cervical injuries when the shoulder proprioception may be affected weight bearing facilitates the proprioception as well. It is important to first train for the prone extensions, back extensors strengthening and then progress to quadruped position.

At first therapist should stand behind the patient and support at the pelvis and give backward pull to the pelvis to come to the quadruped position. Ask the patients to perform this movement several times and slowly reduce the support given.







***Fig 43 to 45 Patient with paraplegia transitioning from prone to quadruped***

To unlock the hips from the prone extension position patient would need to bend the neck down and give a backward thrust with arms so that hips can unlock and go in flexion.







***Fig 46 to Fig. 48 Patient with quadriplegia bending down to unlock the hips before pushing up to quadruped position***

Once the patients are able to attain the quadruped position the progression will be to maintain this position and build the endurance for the same to up to 30 seconds before progressing to next level.

7.Quadruped to side sitting

As the endurance to hold the quadruped positions increases to 30 seconds, the next progression will be to move forward and backwards in quadruped further strengthening the shoulder muscles and scapular muscles. Once the patient is able to perform this movement independently, progress to side sitting in quadruped. This position is of importance to strengthen the pectoral group of muscles, trunk side flexors and abdominals in addition to the deltoids.

Start with giving a support of a bolster or a stool on the side to achieve partial range of side sitting followed by complete side sitting on the plinth.





*Figure 49 to 50 - Supported quadruped to side sitting in partial range*



*Fig 51 - Supported quadruped to side sitting through full range*

**8.Crawling-**

In paraplegic patients repeated crawling helps in increasing strength of upper limb muscles, upper back muscle and lower abdomen muscles. Crawling also facilitates hip flexor muscles and proprioception in the hip joint.

**9.Quadruped to kneeling**

Kneeling position simulates walking but due to the lower center of gravity it is easier to perform. The trunk muscles can be strengthened in this position to achieve better balance in standing. The transition from quadruped to kneeling is crucial for back extensor strengthening. Several progressions are possible in kneeling position. Kneeling also facilitates the hyperlordotic posture that enables the patients to lock the hips in standing.

For higher thoracic and cervical level injuries assisted kneeling can be trained by resting the arms on a stool or a platform and supporting the pelvis to achieve the anterior tilt.

**10.Kneel sitting to kneel standing**

Kneel sitting to kneel standing helps to strengthen the abdominal muscles and hip extensors. It can be performed independently only in case of lumbosacral injuries. For higher level injuries assisted movement can be performed. Patients can hold onto a hand rail or push down on the stool or a platform to perform the movement. In assisted exercises, scapula depressors and the shoulder muscles are strengthened these muscles are key to gait training and to maintain the balance in standing. This exercise also develops the pattern for achieving the lordotic posture by pulling on hands. This movement pattern is very important for standing balance.







*Figure 52 to 54 Assisted transition from kneel sitting to kneel standing*

**11.Kneel walking**

A progression of kneel standing is kneel walking in the sideways and rarely in forward direction. This exercise cannot be performed by patients with injuries of the thoracic and cervical spine.

**12.Sitting on the edge of the bed to supine**

Finally, the bed mobility exercises end with being able to lie down in supine position from sitting position. This requires eccentric contraction of the abdominal muscles and trunk side flexors. A therapist should be careful to train these muscles to facilitate a smooth and controlled movement.

Patients should be taught to lift the legs and throw when lying down. Easiest and most energy efficient techniques is to grip the legs under the knee with hands and then to turn backwards with force. This momentum can be used to lift the legs with hands as the patient achieves lying position ask them to throw their legs out and straight holding on the back side of the calves.

Alternatively, patients can also lift the legs and keep them on the bed back in long sitting. This can only be performed by patients who have a good sitting balance



*Figure 55 - 58*

**Relevant strengthening exercises**

The bed mobility training should be accompanied with the relevant muscle strengthening exercises. Following are the examples of some of the exercises. The exercises are not limited to the below exercises.

Important muscles to be strengthened are:

1. Deltoids
2. Rotator cuff muscles
3. Shoulder rotators
4. Shoulder adductors.
5. Scapular muscles
6. Lattisimus dorsi
7. Triceps
8. Biceps
9. Abdominals
10. Back extensors
11. Trunk side flexors
12. Obliques (Internal and External) .



*Figure 59 to 60 - Strengthening of external rotators of the shoulder*



*Figure 61 to 62 - Strengthening of internal rotators of the shoulder*



*Figure 63 and 64 - Strengthening of abdominals*



*Figure 65 and 66 - Strengthening of abdominals*



*Fig 67 - Strengthening of back extensors*



*Fig 69 and 70 - Strengthening of back extensors*



*Figure 71 to72 - Prone pushups for strengthening scapular muscles, deltoids, pectoralis group, serratus anterior muscles, deltoids, pectoralis group, serratus anterior and Abdominals*



*Fig 73 to 74 Sitting pushups for strengthening of shoulder, triceps,pectoral muscles,upper back*



*Figure 75 Strengthening of quadrates lumborum in supine lying*



*Figure 76 Strengthening of quadrates lumborum in supine lying*

Practicing and repeating the movements several times in a day is very important. The exercises should be practiced even in the absence of the therapists throughout the day. It is also important to educate the patient relatives and care takers to not help the patient unless essential to do so and to have patience to let the patients carry out the movements on their own even if it is slow.

Bed mobility training is essential for the patients to carry out activities of daily living. The therapist should take care to inhibit wrong postural patterns but should encourage the muscular compensations.

Further if the patient is unable to use the compensatory movements, therapist should teach these movements and guide them.

**Transfer techniques**

Ambulation in most of the patients with spinal cord injury is wheelchair dependent except for the patients with incomplete and lumbosacral injuries. To be able to get into wheelchair independently the patients require training of special techniques. It is important to train the patients, rather than the patients exploring it on their own, to prevent secondary injuries. Techniques used for transfers should be most energy efficient and safe, not harming the integrity of other joints and soft tissues. There are different transfers that the patients need to do, we can simply classify these in same level transfers, transfers to the surface below, transfers to the higher surface and corner transfers. However, therapists should also focus on functional transfers.

Prerequisites to the transfer training are:

- Patients should be independent in bed mobility
- Sitting balance at the edge of the bed should be good
- Strength of the shoulder stabilizers, triceps, pectoralis and abdominal muscle strength should be adequate

**Physiotherapy training prior to transfers**

Training for same level transfers involves strengthening of the deltoids, rotator cuff muscles, shoulder depressors, triceps, pectoralis, abdominals and back extensors. Following are some of the training exercises to be performed in bed before training for transfers.

Same level transfers with equipment:

- Transfer board
- Push up bars

Training for transfers can initially make use of transfer boards of various sizes and shapes to suit the patient needs.

**Transfer technique**

- Keep the wheelchair at 45-degree angle / 90 degree angle if the shoulder strength is adequate
- Make sure that the breaks of the wheelchair are on
- Remove the hand rest near to the side of the transfer
- Remove the foot rests
- Shift as close to the wheelchair as possible
- Place the transfer board underneath your thighs
- Lift the legs with hands and keep it slanting, to align to the wheelchair
- Lock the elbows using substitute muscles when triceps are weak or impaired due to injury. (Substitution can be done by putting weight on the forearm ensuring the wrist extension and then perform shoulder external rotation to lock the elbow. Supination can also aid in locking of the elbow.)
- With one quick movement of push-up (in case of preserved triceps) and turning the trunk slide onto the transfer board.
- When triceps is impaired move the head and shoulders away from the wheelchair and push the chin downwards to lift the hips and push the hips towards the wheelchair / on the transfer board (Head-hip relationship). Use the same technique to slide down the transfer board by adjusting the direction of arms and relocking the elbows using compensatory mechanisms. (Take precaution to not perform internal rotation of the shoulder as it may unlock the elbow).
- Make sure to keep your balance forward

- Make sure to keep weight on your arms at all times, as that is the only way to control the movement
- Once you are on the transfer board slide slowly onto the wheelchair
- Ensure you are sitting properly in the wheelchair
- Now lean on the back of the wheelchair and on the hand rest on the opposite side
- Lift the leg near to hand rest that was removed with opposite arm
- Remove the transfer board
- Put the arm rest back
- Put the foot rest
- Remove the breaks

Once good shoulder and trunk control is developed, the same transfer can be performed without the transfer board (Figure 1 to 6)





*77, 78 and 79 - Training for bed to wheelchair transfers*





*80, 81 and 82 - Training for same level wheel chair transfer by a patient with paraplegia*

**Same level transfer for quadriplegic**

With practice patients won't require equipment like transfer board and push up bars. This is applicable even for the higher level injuries resulting in quadriplegia.

For patients who do not have good trunk control either due to higher level of injury or due to poor training and muscle weakness, the wheelchair should be kept at an angle of 45 degrees to the bed. For patients with higher levels removing and putting the foot rest back may have to be done by others (Figure 7)










*83 to 89 - Transfer from bed to wheelchair by a patient with quadriplegia*

**Transfers to higher level**

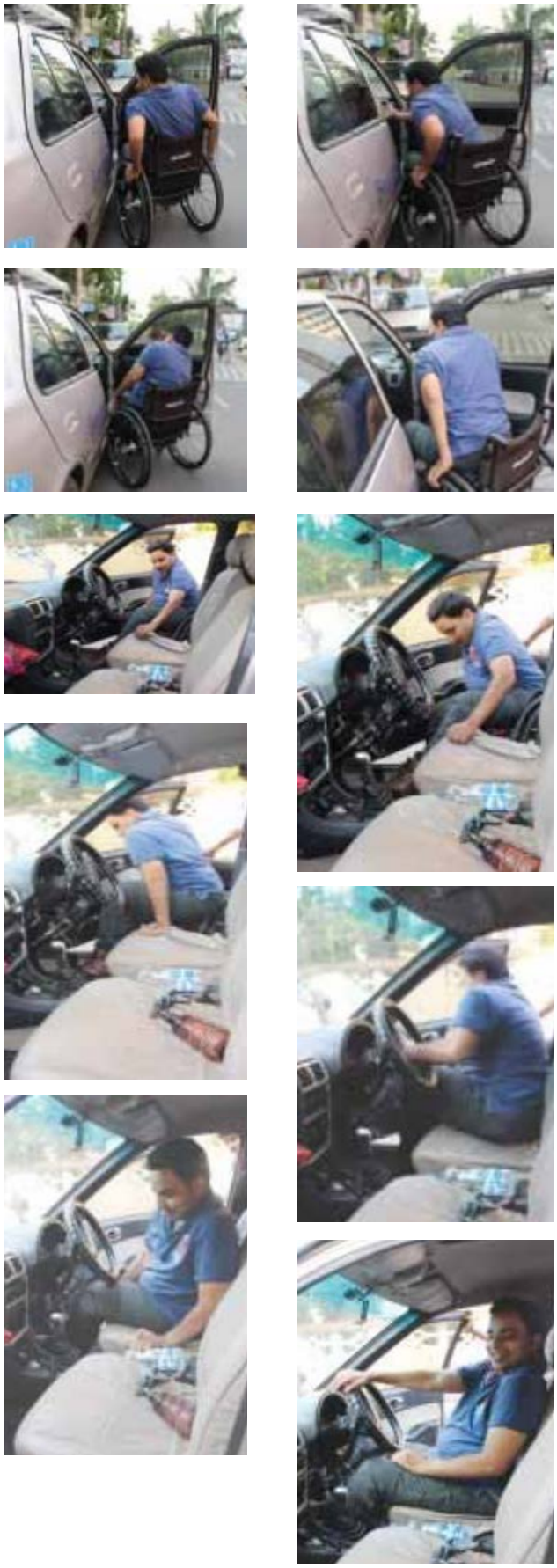
To perform this activity shoulder muscles and triceps needs to be very strong with good trunk control. The wheelchair is to be placed next to the bed horizontally, therapist sits facing the patient providing support at the knee, patient has to keep ipsilateral hand on the bed and contralateral arm on the armrest. Then patient has to push up lifting his hip and then rotate the trunk to get seated on the bed. After practicing for several times the support can be minimized till the patient starts performing it independently (as shown in image 15 to 17)



*90 to 92 - Training for higher level transfers*

**Functional transfers:**

Along with the training for transfers onto higher surfaces, lower surfaces, same level transfers and corner shifting; functional transfers like commode transfers and car transfers must also be taught and practiced. Figure 18 to 27 show the patient with quadriplegia transferring from the wheelchair to the car



*93, 94, 95, 96, 97, 98, 99, 100, 101 and 102 - Transfers to the higher level car seat*

**Transfer from wheelchair to commode-**

Once the patient learns same and higher level transfers, transferring to commode becomes easier. The technique used will be same but to make life easier bar handle can be attached next to commode for better transfer as shown in the image 28 and 29

Note- Lock the wheels every time you perform transfer activity.



*103 to 104 - Transfer from wheelchair to commode*

**Gait training**

There is an ongoing debate about whether it is advisable to teach the paraplegics to walk or not? If you ask the patients, they would want to walk. In the initial days there is poor acceptance of wheelchair use for ambulation but later after experiencing the energy expenditure for walking, patients may choose wheelchair for long distance ambulation as well as indoor mobility. This debate has not settled even amongst the clinicians. Weight bearing exercises especially standing and walking were thought to prevent osteoporosis, but recent investigations have suggested that weight bearing is not the only factor preventing osteoporosis. Only if there is muscle activity and contractive forces of muscles the bone growth is stimulated, and osteoporosis is prevented. It was further noted that non-weight bearing does not alter the calcium excretion in the manner it is altered in neurotypical adults. This has been attributed to the neural feedback for calcium excretion and absorption which is absent in individuals with spinal cord injury. There is unequivocal evidence for the benefits and shortcomings of gait training.

Therapists should make patients aware of the advantages and shortcomings of ambulation with orthotic devices and walker or crutches. Whether to ambulate with orthotic devices and walker and crutches is patient's choice and it should be respected. For functional and strength gains therapists should undertake gait training for patients with spinal cord injury.

Before the gait training can begin, it is important to determine the orthotic devices that are required for the patient.

**Lower Extremity Orthosis for SCI**

Occurrence of pressure sores can be reduced by standing using the lower extremity orthotic devices.

**Goals of Splinting:**

- To prevent contractures
- To prevent compartment syndromes
- To maintain alignment

**Benefits of orthosis assisted standing**

- Prevent joint contractures.
- Decrease osteoporosis.
- Reduce risk of fall.
- Reduce man power.
- Reduce risk of fracture by sudden torsion force.
- Prevent UTI.
- Enhance cardiopulmonary endurance by allowing freedom of movement
- In pediatrics, it enhances the development of trunk and head control.

**Types of orthosis**

- Hip knee ankle foot orthosis (HKAFO) - HKAFO is an extension of a KAFO with a pelvic band. The support provided by the pelvic band improves standing balance

for spastic patients.

- Knee Ankle Foot Orthosis (KAFO): - provides stability at knees and ankles. KAFO is prescribed in case of

1. Hip and trunk extensors weakness
2. Weak quadriceps muscles

- Floor reaction orthosis (FRO)- It provides stability at ankle, prevents foot drop and reinforces knee locking. FRO is mostly prescribed in patients with incomplete cervical level injury with good upper limb strength and in patients with D10 or lower level injury who develop knee locking with minimal hyperextension of the knee.
- Ankle foot orthosis (AFO): Both static and dynamic AFO are available. In case of complete foot drop one may consider static AFO but for foot drop due to spasticity that affects the knee joint control and alignment, a dynamic KFO may be considered.

There are many other orthotic devices available for the patients. Factors to be taken into consideration before choosing the orthotic devices are,

- Ease of availability.
- Accessibility
- Durability
- Safety
- Comfort
- Cost

Therapist should take an informed decision with the patient about the use and type of orthosis. Therapist may also choose to give orthotic devices that are different from the conventional KAFOs, where the knee orthosis and ankle orthosis are separate and therefore are easier to put on. The weight is also considerably less.

The first step towards gait training is the ability of the patients to wear their own orthoses. Patients need a good sitting balance and a good hip range for wearing the orthoses and therapist should target to achieve these before beginning the gait training. Figures 11.1 to 11.6 show a patient with paraplegia wearing a KAFO.



***105 to 110 - Patient of paraplegia wearing her KAFO***

Tolerance to vertical position must be developed for patients with higher thoracic and cervical injuries before making them stand. This can be achieved with tilt table in the initial phases. Once the tolerance has been developed, passive standing can be performed with the help of standing board. Exercises can be performed to train for trunk balance and shoulder stability in the standing board. Figures 7 to 12 show a patient with quadriplegia performing

exercises in the standing frame.

Blood pressure must be monitored continuously, sudden change in posture results in postural hypotension causing black outs. If patient reports any discomfort, excessive sweating or difficulty in keeping the eyes open then the patient is to be taken to supine position immediately. Hence, blood pressure is to be checked before standing and after making the patient stand.



***111, 112, 113, 114, 115 and 116 - Trunk stability and strengthening exercises in standing board***

**Parallel bar exercises before gait training**

Patients have to first learn the paraplegic stance or develop hyper-lordotic stance to maintain the hip stability. This posture is already taught and practiced during bed mobility training. However, once the patient is made to stand in the parallel bar this posture should be reemphasized. Patient can be supported at the pelvis and asked to perform forward and backward movements thrusting the hips back and forth (Figure 117 and 118)



***Figure 117 and 118 - Facilitation of the hyper-lordotic posture***

Once the hyper-lordotic posture has been learnt, shifting weight sideways is facilitated by holding at the pelvis and by guiding the sideways movement of the pelvis. This maneuver is called weight shifting and it is essentially to be able to propel forward.



***Figure 119 and 120 - Weight shifts in hyper-lordotic posture***

As the patients learn to do the weight shifts slowly the trunk rotations are facilitated to be able to propel the legs forward for mimicking walking. For this the therapist should stabilize the pelvis on the contralateral side of the movement, ask the patient to side flex on the contralateral side and move the leg forward (Figure 11.17 and 11.18). As the hip flexors and hikers are not functional in paraplegics with higher thoracic injury; internal and external obliques must be used

for moving the leg forward and clearing the ground. Therefore, these muscle patterns must be developed and muscles should be trained. Once the pelvis is stabilized the muscles can contract to bring about unilateral movement of the lower extremity with trunk rotation. Support given by the therapist at the pelvis acts as a fulcrum for this movement. Slowly as the muscles learn this pattern, trunk rotation and leg movements can be carried out independently.



*Figure 121 - Forward stepping using trunk rotation assisted by a therapist*



*Figure 122 - Backward stepping using trunk rotation assisted by a therapist*



*Figures 123 and 124 - Sidestepping using trunk side flexors*

Therapist should guide not only the forward and backward movement but also the movement of the legs sideways (Figure 121 and 122)

**Gait training**

Depending on the patient's muscle power, swing to and swing through gait can be taught instead of the 'alternating four-point gait'. For swing to and swing through gait very strong shoulder and trunk muscles are required.

For teaching the swing through and swing to gait, support the patient at the pelvis and ask to perform a push up on a parallel bar and propel the body forwards and backwards (fig 21 to 23)

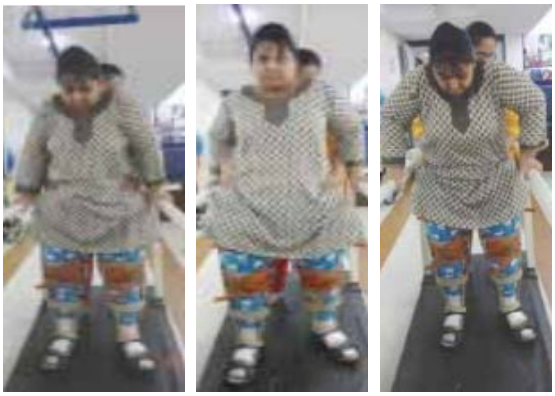


*Figure 125, 126 and 127 - Training for Swing through and Swing to gait*

Once the guided movements are practiced patients are encouraged to perform push-ups on the parallel bars and maintain the position (Fig. 11.24 and 11.25). As the endurance increases swinging movements are added to the push-ups.

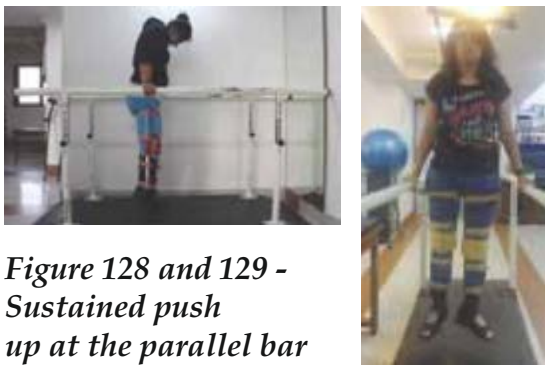


*Figure 128 and 129 - Sustained push up at the parallel bar*

Alternatively, patients can also be given advanced strengthening exercises like pull up in the parallel bars (Fig 26)



*Figure 130 - Assisted pull ups*

Once good shoulder strength is achieved the forward backward bending of the trunk and the forward, backward and sideways movement of the legs can be performed by the patient independently (fig. 27 to 33)



*Figure 131 and 132 forward and backward bending of the trunk*



*Figure 133 and 134 - Leg movements using trunk rotation and side flexion performed independently by the patient.*

Parallel bar training continues to develop the alternate gait pattern. Common exercises to be performed by the patient are forward, backward walking and side walking. These exercises can be performed independently by the patients with lower thoracic and lumbosacral injuries. Patients with higher thoracic injuries may need continued assistance for these exercises. Patients with complete cervical injuries will not be able to perform these exercises independently.

**Gait training in walker**

Once the gait training in parallel bar develops adequate trunk and shoulder control, patients can be trained to walk using a walker. First task before learning to walk with a walker is to be able to stand up from bed with the walker. A stable position is to turn in prone and pushing up to stand up. Initially the legs can be supported and guided to prevent slipping (Fig. 31 to 32)

Two different ways to get up and stand-

1.Prone to standing-



*Figure 135 to 140 - Standing up from bed onto the walker with assistance*

The exercises performed in parallel bar should be repeated in the walker as well i.e. sustained push-ups and weight shifts. To begin with 'swing to' gait can be used on the walker while training for alternate gait pattern. In the same manner as that in

parallel bars patients can walk using alternate gait patterns (Fig. 11.48 and 11.50)



*Figure 141, 143 and 143 - Walking with the help of a walker wearing push knee splints and high boots with posterior steel shank.*

Sit to stand- Patients with better trunk and shoulder control can stand up from the chair may choose to stand up by pulling onto the walker and then pushing down on the walker to perform a push uplifting the legs up and keeping them in desired place (fig 39 to 41)





*Figure 144, 145 and 146 - Standing from the bed independently using a walker*

It is also important to learn correct techniques of sitting down to prevent injuries. The patient should slowly lean back still holding onto the parallel bars with arms (Figure 11.54 and 11.55). Once they are seated on the wheelchair or bed, they can let go of the hands. A quick push up and shifting back will ensure that the patient is securely seated. Once this is achieved orthotic devices can be taken off..



*Figure 147 and 148 - Sitting down on a wheelchair from parallel bar*

**Gait training with Floor reaction orthosis-**

For a patient to become independent in ambulation with FRO, patient needs to master 4 stages of training which are-

1.Achieve knee locking- To initiate knee locking patient must perform sit to stand activity and learn to lock knee in minimal hyperextension of knee and lumbar lordosis.



*Figure 149 and 150 - Sit to stand to initiate knee locking*

2. Gait training with FRO and kunming walker- To make the task easier, initially stepping at the same place must be practiced in which the patients learn to do the weight shifts slowly the trunk rotations are facilitated to be able to propel the legs forward and lock the knee for mimicking walking.



*Figure 151 and 152 - Forward and backward stepping at the same place*

Once patient learns to propel leg forward and lock the knee, forward walking can be adviced. After repeated practice if the patient gains good hip knee flexion pattern and confidence then the herapist can challenge the patient by introducing him to the next step i.e. parallel bars.



*Figure 153 and 154 - Forward walking with FRO and Kunming.*

3.Gait training with FRO in parallel bars- Same protocol is to be followed as we do in gait training with KAFO but at the start the therapist should be sitting behind the patient and provide minimal assistance to prevent buckling of knee (refer figure 153,154). Gradually as the patient gains goo hip and knee control, therapist can reduce the assistance.




*Figure 155 and 156 - Walking in parallel bars with FRO and physical assistance.*

4.Gait training with FRO and 4-point walker-

This stage can be further divided in 4 stages which are-

I.Getting up from bed with walker assistance independently (refer figure 155).

II.Standing with walker and FRO – Balance training in 4 point walker forward and backward shifting of walker (refer figure 156).

III.Walking with FRO and walker with 1

physical assistance (refer figure 157,158).



*Figure 157 and 158*

**Advantages of FRO-**

1. Patient walks with hip-knee flexion pattern.
2. More functional as it becomes easy to sit and stand.
3. Light weighted.
4. Cost effective.

**Disadvantages-**

1) Risk of fall.
2) Soft tissue damage in long run can lead to ligament laxity.

**Gait training in the crutches**

Crutch walking is usually possible for patients with lower thoracic and lumbosacral injuries. Some patients with higher thoracic injuries may also be able to learn crutch walking (figure 159 to 161).



*Figure 159 and 160*

The training for crutch walking remains the same as that of walking in the parallel bar and walker.



*Figure 159, 160 to 161 - Standing up with the help of Elbow crutches*



*Figure 162 and 163 - Gait training with the help of Elbow crutches*

**Stair climbing**

Patients choose to stand up by pulling onto the walker and then pushing down on the walker to lift the legs up and keep them in desired place.

Patients with lower thoracic injuries can climb up the stairs using hip hikers and trunk side flexion to be able to clear the stairs (Fig 164 to fig 166)



*Figure 164 and 165*



*Figure 166*

Another method is facing the railing and holding it with both hands, swing one limb in one pendulum cycle and then hike the hip and place the foot on the higher step; then bring the other foot up. This is possible only if he has strong hip flexors or quadratus lumborum. There is yet another way of climbing up i.e. Jack kniving, Here the patient must lift him up higher than the height of a step with back facing the staircase and swinging both legs backwards to reach the higher step. The person must use one crutch in one hand and holding the railing with the other hand or using crutches in both hands. This is possible only for patients who have very good control of the trunk as well as very strong Triceps and shoulder muscles.

**Occupational Therapy Management in SCI**

Traumatic spinal cord injury (SCI) may occur following road traffic accidents, gun-shot injuries or direct trauma to the spine through a fall from height. Traumatic SCI can be either complete or incomplete, which affects a client's prognosis and level of functional performance.

1. Complete SCI results in affection of sensory and motor functions below the level of injury.
2. Incomplete SCI may involve a number of neurological segments and sensorimotor functions may be partly or completely intact.

Non-traumatic SCI occurs secondary to different diseases such as :-

1. Tumors : may be benign or malignant
2. Spina bifida : congenital SCI, may be myelocele or meningomyelocele
3. Syringomyelia : growth of a cyst in the spinal cord
4. Multiple sclerosis : long-lasting disease that can affect the brain and spinal cord
5. Amyotrophic lateral sclerosis : gradual deterioration and death of motor neurons
6. Transverse myelitis: inflammation within the spinal cord
7. Communicable diseases - Tuberculosis and HIV
8. Degenerative diseases like OA leading to spinal stenosis

**Impairments in Traumatic SCI and their implications for Occupational Therapy (OT)**

1. **Sensory loss :** There is an increased risk of skin breakdown with sensory loss, resulting in pressure sores. These can be prevented through regular self and caregiver skin examination and pressure relieving techniques, like routine weight shifts and use of appropriate bed mattresses or wheelchair cushions.
2. **Vital capacity :** Maintenance of good vital capacity results in performing the exercises and activities of daily living (ADLs) more fruitfully. Breathing difficulty should be managed by an occupational therapist with frequent rest periods in between exercises and training of inspiratory and expiratory techniques while performing ADLs.
3. **Orthostatic hypotension :** Occurs where BP drops down suddenly by 20 mm Hg or more of the baseline systolic blood pressure. If left untreated, client may get unconscious. Symptoms include light-headedness, pallor and visual changes. This can be addressed by client's

positioning in supine position and elevating feet above heart level or by reclining the wheelchair. OTist should be cautious while working on dependent/independent transfers from supine to long-legged sitting, or any quick changes in body positions, to avoid a rapid drop in blood pressure. Slow movements in transfers to avoid orthostatic hypotension are recommended. Also ask the client/caretakers to gradually bring the client to an upright position 10 minutes before the therapy session starts.

4. **Autonomic dysreflexia :** Occurs in injuries at or above T6 level, where there is an increase in systolic blood pressure of 20-40 mm Hg or higher than normal. A pounding headache is usually the first common symptom followed by goosebumps, sweating, blurred vision, anxiety , heavy breathing and chest tightness (Consortium for Spinal Cord Medicine, 1997). It can be triggered due to :- a) bladder problems like overfull bladder, bladder or kidney stones, urinary tract infection or blocked catheter. b) bowel problems like constipation, distension or haemorrhoids. c) skin issues like prolonged pressure, pressure sores, ingrown toenail, deep vein thrombosis or fractures. This can be addressed by making the client sit up/stand up with head elevated, loosening tight clothes or devices, checking catheter for any obstruction and passive stretching.

5. **Heterotopic ossification (HO) :** HO occurs in 16-53% of clients with SCI. This can be controlled by appropriate positioning in bed and wheelchair and maintenance of functional range of motion (FROM) of upper & lower extremity joints through active and/or passive ROM exercises.

6. **Spasticity :** Occurs in clients with an upper motor neuron (UMN) lesion, i.e., an injury above D12 level. Severe spasticity should be managed with medications. It can be managed by OTists through appropriate positioning, splinting, stretching, ROM exercises, serial casting, etc. Spasticity limits functions such as dressing, self-care and transfers. Spasticity may lead to development of contractures; wheelchair positioning is essential to prevent contractures.

7. **Bladder and bowel dysfunction :** Bladder and bowel functions are affected in all injuries at and above S2-S5 level. Using new routines and habits of bladder and bowel elimination is essential to reduce risks of infection and risk of autonomic dysreflexia. Use of intermittent/ indwelling/ supra-pubic catheterization helps complete SCI clients with UMN (above T12) or lower motor neuron (LMN) (below T12) injury to empty bladder. Digital rectal stimulation or medications for bowel management helps to reduce the risk of involuntary defecation. Occupational therapists support the new skills and habit acquisition for safe transfers, clothing management, safety with task performance and bowel elimination and catheter care.

8. Temperature regulation : This is often affected in SCI clients, so they need to avoid exposure to sunlight and severe temperatures to reduce risk of hypothermia, heat stroke or frostbite.

9. **Pain :** Pain may be either nociceptive (caused by reaction to noxious stimuli like inflammation) or neuropathic (originating in the nervous system and may be caused by mechanisms such as misdirected neural sprouting after the injury) below the level of injury. Burning and shooting neuropathic pain often occurs with gunshot injuries. Sometimes cramping visceral pain can occur due to medical issues like constipation, ulcers,

kidney/gall bladder stones, etc.

**10.DVT :** Most commonly occurs in lower extremities, abdominal areas or pelvic areas. Regular skin inspection with attention to asymmetric color, size or temperature should be done.

**11.Sexual function :** Sexual functions are often affected in clients with SCI.

**OCCUPATIONAL THERAPY EVALUATION**

A. Occupational Profile includes occupational history, life roles, values, interests, understanding of causes, their implications and prognosis.

B. Physical assessment includes :-

1. Upper extremity ROM:

a. TAM (Total Active Motion)

b. TPM (Total Passive Motion)

c. AROM (Active ROM) & PROM (Passive ROM)

d. FROM (Functional ROM)

2. Muscle strength : Manual muscle strength (MMT), grip and pinch strength

3. Muscle tone : Modified Ashworth Scale

4. Sensibility :

a. AIS Sensory Assessment

a. AsTex Sensory Test

5. Balance in both sitting and standing

6. Hand/Upper limb functions :

a. Summed Upper Limb Strength

a. The Capabilities of Upper Extremity (CUE) of the target hand

b. Modified Action Research Arm Test (ARAT)

c. AIS Sensory Assessment

d. AuSpinal Assessment

e. Silverman test

7. Pain :

a. Visual Analogue Scale (VAS)

a. Type, location

B) The Occupational Performance assessment is carried out through observations and performing standardised and non-standardised assessment scales, such as :-

1. The Spinal Cord Independence Measure (SCIM)

2. The Quadriplegia Index of Function

3. The Functional Independence Measure (FIM)/ Wee-FIM

4. The Canadian Occupational Performance Measure (COPM)

5. BADLs and IADLs

6. Work evaluation

7. Interest Checklist

**C) Quality of Life -**

1. Assessment of Quality of Life - 8 (AQoL - 8)

2. Health Utilities IndexMark 3 (HUI3)

**FRAMES OF REFERENCES (FOR)**

1. Biomechanical FOR

2. Rehabilitative FOR

3. Occupational Adaptation FOR

4. Occupational Behavioral FOR

**FUNCTIONAL INDEPENDENCE FOLLOWING SPINAL CORD INJURY**

A person's functional independence has a major impact on a client's quality of life, sense of self-worth and self-esteem, and consequential social participation. It is logical that achievable functional independence is dependent on a person's level of injury.

Factors affecting functional performance include :-

1. Neurological level
2. Degree of impairment (complete or incomplete)
3. Age at the time of injury
4. Time period since injury
5. Other injuries or medical conditions (e.g. fractures, nerve injuries, cardiac complications, arthritis etc.)
6. Physique (body proportions and body distributions)
7. Cognition/motivation (if TBI, depressed)
8. Mental illness (e.g. schizophrenia, personality disorder)
9. Drug and alcohol abuse
10. Social support
11. Cultural expectations
12. Financial resources
13. Environmental factors

Rehabilitation should aim at assisting clients with SCI to overcome the limitations caused in daily activities by : improving trunk, upper and lower limb functions, providing adaptive/assistive devices and assistive technologies and other environmental accommodations to enable the clients to continue self, family and work roles. Table 1 to 6 provides a broad summary of the functional outcomes desirable for different levels of complete SCI.

**Table 1 :**

| Task / Activity | Expected Functional Outcome | Functional possibilities & restrictions | Adaptive devices/Assistive technology | Splints |
|---|---|---|---|---|
| Pressure relieving/ Positioning | Complete assist; can be independent with equipment | 1. Limited head control/movement<br>2. Ventilator dependent<br>3. Possible autonomic dysreflexia | • Power wheelchair with tilt-in-space, postural supports and head control systems<br>• Pressure-reducing cushion<br>• Pressure-relieving mattress. | • Resting hand splints for overnight use<br>• Soft wrist cock-up braces during the day |

| | | | | |
|---|---|---|---|---|
| Bed Mobility | Complete assist | | • Electric bed with elevating back break & side rails<br>• Scanning bed controller with sip/puff independent operation<br>• Trendelenburg option may be required with positioning |  |
| Transfers | Complete assist | | • Electric hoist with slings | |
| Mobility | Manual wheelchair (W/C) - Total assist<br>Power W/C - Independent with highly specialized equipment | | • Lightweight manual W/C with tilt-in-space, postural supports and headrest. High back may be indicated<br>• Power W/C with tilt-in-space, ventilator tray (for C1-C3) and control equipment | |
| Feeding | Complete assist | | • Drinking system attached to W/c | |
| Grooming | Complete assist | | Not applicable | |
| Bathing | Complete assist | | • Mobile shower commode<br>• Handheld shower | |
| Dressing | Complete assist | | Not applicable | |

| | | | | |
|---|---|---|---|---|
| Toileting | Complete assist | | • Mobile shower commode with custom padded seat, tilt-in-space, arm troughs for support, head rest (for C1-C3), lateral supports | |
| Transport | Complete assist | | • ·Van with modified access & appropriate docking/tie-down system | |
| Domestic management | Complete assist | | Not applicable | |

**Table 2 :**

| Task/Activity | Expected Functional Outcome | Functional possibilities & restrictions | Adaptive devices/Assistive technology | Splints |
|---|---|---|---|---|
| Pressure relieving/ Positioning | Complete assist; may be independent with equipments | 1. Full head control / movement<br>2. Diaphragmatic breathing<br>3. Possible autonomic dysreflexia<br>4. Able to raise arms & flex elbows | • Power wheelchair with tilt-in-space, postural support and head control systems<br>• Pressure-reducing cushion<br>• Pressure-relieving mattress. | • Resting hand splints for overnight use<br>• Soft wrist cock-up braces during the day<br>• Long opponens splint |
| Bed Mobility | Complete assist | | • Electric bed with elevating back break & side rails<br>• Trendelenburg option may be required with positioning | |
| Transfers | Complete assist | | • Electric hoist with sling | |

| | | | | |
|---|---|---|---|---|
| Mobility | Manual W/C - Complete assist<br>Power W/C - Independent with highly specialized equipment | | • Lightweight manual W/C with tilt-in-space, postural supports and headrest. High back may be indicated<br>• Power W/C with tilt-in-space, headrest, specialized hand control | Wrist cock-up |
| Feeding | Complete assist | | • Drinking system/ drink bottle with C-clips attached to W/C<br>• Palmar band/ universal cuff, ringed cutlery | Wrist cock-up |
| Grooming | Complete assist | | • Palmar band/ Universal cuff | |
| Bathing | Complete assist | | • Mobile shower commode<br>• Handheld shower in cuff | |
| Dressing | Complete assist | | • Ring pull zippers, clothing loops | |
| Toileting | Complete assist | | • Mobile shower commode with custom padded seat, tilt-in-space, padded armrests for support, lateral supports | |
| Transport | Complete assist | | • Van with modified access & appropriate docking/tie-down system | |
| Domestic management | Complete assist | | Not applicable | |

**Table 3 :**

| Task/Activity | Expected Functional Outcome | Functional possibilities & restrictions | Adaptive devices/Assistive technology | Splints |
|---|---|---|---|---|
| Pressure relieving /Positioning | Moderate to Complete assist; may be independent with equipment | 1. Full head control/movement<br>2. Diaphragmatic breathing<br>3. Possible autonomic dysreflexia<br>4. Able to extend wrist to some extent | • Power wheelchair with tilt-in-space, postural support e.g. lateral supports<br>• Pressure-reducing cushion<br>• Pressure-relieving mattress | • Thumb post splint to position thumb in opposition when using tenodesis action<br>• Wrist-driven flexor hinge splint |
| Bed Mobility | Complete assist | | Electric bed with side rails | |
| Transfers | Complete assist | | • Electric hoist recommended<br>• Assisted sliding board transfer | |
| Mobility | Manual W/C - Complete assist Power W/C - Independent with highly specialized equipment | | • Lightweight rigid/folding frame manual W/C with modified push rims (rubber-coated/larger diameter rims) and push mitts<br>• Power W/C with hand control, power tilt-in-space<br>• Headrest for outdoor mobility | |
| Feeding | Complete assist | | • Palmar band/universal cuff, ringed cutlery, drinking bottle with C-clips stable table, plate guard | |

| | | | | |
|---|---|---|---|---|
| Grooming | Complete assist | | • C-clip, palmar band/universal cuff, built-up toothbrush, long-handled aids<br>• Electric toothbrush | |
| Bathing | Complete assist | | • Mobile shower commode<br>• Shower chair without armrests with padded seat<br>• Handheld shower<br>• Soap mitt, long-handled scrub brush | |
| Dressing | Upper : Moderate assist<br>Lower : Complete assist | | • Ring pull zipper, clothing loops | |
| Toileting | Complete assist | | • Mobile shower commode with custom padded seat<br>• May require increased backrest inclination angle and /or padded armrests | |

| | | | | |
|---|---|---|---|---|
| Transport | Complete assist | | • Modified self-drive van with W/C power lockdowns<br>• Hand controls to drive, adaptive techniques to transfer self and chair into vehicle or with chair hoist (e.g. slide board with sheepskin) | |
| Domestic management | Complete assist | | • May complete basic tasks with environmental modifications | |

**Table 4 :**

| **Task/Activity** | **Expected Functional Outcome** | **Functional possibilities & restrictions** | **Adaptive devices/Assistive technology** | **Spints** |
|---|---|---|---|---|
| Pressure relieving/Positioning | Minimum assist to independent | 1. Limited head control/movement<br>2.Diaphragmatic breathing<br>3. Possible autonomic dysreflexia<br>4. Able to flex/extend wrist, partial finger movement | • Wheelchair with postural support e.g. lateral supports<br>• Pressure-reducing cushion<br>• Pressure-relieving mattress | · Static hand splint with lumbrical bar & thumb post<br>· Knuckle bender |
| Bed Mobility | Moderate assist to independent | | • Electric bed; rails may be required<br>• Single/larger beds may be used | |
| Transfers | Minimum assist to independent | | • With/without transfer board | |

| | | | | |
|---|---|---|---|---|
| Mobility | Manual W/C - Independent on even surfaces, assist in uneven surfaces<br>Power W/C - Independent | | • Lightweight rigid/folding frame manual W/C with modified push rims (rubber-coated rims)<br>• Power W/C recommended for long distances/outdoor mobility | |
| Feeding | Independent | | • With/without ringed or built-up cutlery | |
| Grooming | Independent | | • With/without built-up handles, long-handled aids | |
| Bathing | Minimum assist to independent | | • Mobile shower commode with custom padded seat, tilt-in-space, armrests for support, lateral supports<br>• Shower chair without armrests and with padded seat cushion<br>• Handheld shower | |
| Dressing | Minimum assist to independent | | • With/without adaptive equipments | |
| Toileting | Minimum assist to independent | | • Mobile shower commode with custom padded seat with full/partial cut out for access<br>• Adaptive equipment like suppository inserter | |
| Transport | Independent with hand controls | | • Hand controls to drive, adaptive techniques to transfer self and chair into vehicle or with chair hoist | |

| | | | | |
|---|---|---|---|---|
| Domestic management | Moderate to complete assist | | • Long-handled aids, specific kitchen appliances<br>• Environmental modifications | |

**Table 5 :**

| Task / Activity | Expected Functional Outcome | Functional possibilities & restrictions | Adaptive devices /Assistive technology | Splints |
|---|---|---|---|---|
| Pressure relieving/Positioning | Independent | 1.Respiratory capacity & endurance may/may not be compromised<br>2.Normal upper extremity ROM & strength | • Wheelchair with postural support e.g. lateral supports<br>• Pressure-reducing cushion<br>• Pressure-relieving mattress | Reciprocating gait orthosis(high energy expenditure) |
| Bed Mobility | Independent | | • Electric bed; rails may be required<br>• Slide rails may be required | |
| Transfers | Independent | | • With/without transfer board | |
| Mobility | Independent | | • Ultra lightweight rigid/folding frame manual W/C<br>• Independent in W/C, ascending and descending curbs | |
| Feeding | Independent | | Not applicable | |
| Grooming | Independent | | Not applicable | |

| | | | | |
|---|---|---|---|---|
| Bathing | Independent | | • Mobile shower commode with/without custom padded seat, tilt-in-space, armrests for support, lateral supports<br>• Shower chair without armrests with padded seat cushion<br>• Handheld shower | |
| Dressing | Independent | | • Adaptive techniques like log rolling, long sitting for lower extremity dressing | |
| Toileting | Independent | | • Mobile shower commode with custom padded seat with full/partial cut out for access<br>• Over-toilet aid with padded toilet seat | |
| Transport | Independent | | • Hand controls to drive, adaptive techniques to transfer self and chair into vehicle or with chair hoist | |
| Domestic management | Minimum to moderate assist | | • Long-handled brush and broom, reaching aids<br>• Specific kitchen appliances<br>• Environmental modifications | |

**Table 6 :**

| Task/Activity | Expected Functional Outcome | Functional possibilities& restrictions | Adaptive devices/Assistive technology | Splints |
|---|---|---|---|---|
| Pressure relieving/ Positioning | Independent | 1. Normal upper extremity ROM & strength | • Wheelchair with postural support equipment<br>• Pressure-reducing cushion<br>• Pressure-relieving mattress | • W/C gloves/pad-ded bicycle gloves |
| Bed Mobility | Independent | | • Electric bed; rails may be required<br>• Slide rails may be required | |
| Transfers | Independent | | • With/without transfer board | |
| Mobility | Independent | | • Ultra lightweight rigid/folding frame manual W/c<br>• Independent in wheelie, ascending and descending stairs | • Knee ankle foot orthosis (KAFOs)/ Vannini-Rizzoli Ankle foot orthosis (AFOs) and crutches($T_{11}$-$L_2$ level) |
| Feeding | Independent | | Not applicable | |
| Grooming | Independent | | Not applicable | |
| Bathing | Independent | | • Mobile shower commode recommended<br>• Shower chair without armrests with padded seat<br>• Padded transfer bench<br>• Handheld shower | |
| Dressing | Independent | | • Adaptive techniques like log rolling, long sitting for lower extremity dressing | |

| | | | | |
|---|---|---|---|---|
| Toileting | Independent | | • Mobile shower commode with custom padded seat with full/partial cut out for access<br>• Over-toilet aid with padded toilet seat | |
| Transport | Independent | | • Hand controls to drive, adaptive techniques to transfer self and chair into vehicle or with chair hoist | |
| Domestic management | Minimum assist | | • Long-handled brush and broom, reaching aid<br>• Specific kitchen appliances<br>• Environmental modifications | |

**Table 7 :**

| Task/ Activity | Expected Functional Outcome | Functional possibilities & restrictions | Adaptive devices/Assistive technology | Splints |
|---|---|---|---|---|
| Pressure relieving/ Positioning | Independent | 1. Little bladder-bowel control | • Wheelchair with postural support equipments if indicated<br>• Pressure-reducing cushion | • W/C gloves/padded bicycle gloves |
| Bed Mobility | Independent | | • Normal cushioned bed | |
| Transfers | Independent | | • With/without transfer board | |
| Mobility | Independent | | • Ultra lightweight rigid/folding frame manual W/c | AFOs(at/below $L_3$) |
| Feeding | Independent | | Not applicable | |
| Grooming | Independent | | Not applicable | |

| | | | | |
|---|---|---|---|---|
| Bathing | Independent | | • Mobile shower commode recommended<br>• Shower chair without armrests with padded seat<br>• Padded transfer bench<br>• Handheld shower | |
| Dressing | Independent | | • Adaptive techniques | |
| Toileting | Independent | | • Mobile shower commode with custom padded seat with full/partial cut out for access<br>• Over-toilet aid with padded toilet seat | |
| Transport | Independent | | • Hand controls to drive, adaptive techniques to transfer self and chair into vehicle or with chair hoist | |
| Domestic management | Minimum assist | | • Long-handled brush and broom, reaching aid<br>• Specific kitchen appliances<br>• Environmental modifications | |

**ASSISTIVE TECHNOLOGY**

Assistive technology can be defined as "any item, piece of equipment or product system, whether acquired commercially off the shelf, modified or customized, that is used to increase, maintain, or improve the functional capabilities of individuals with disabilities"

**ACTIVITIES OF DAILY LIVING (ADLs)**

Mobile Arm Support (MAS) [formerly termed as "ball bearing feeders" or "balanced forearm orthosis"] can be used for independent feeding. It should be installed by a skilled and experienced occupational therapist and requires adjustments and training to the client.

**Physical prerequisites :-**

1. C4 quadriplegic
2. Fair to good sitting stability and endurance
3. No upper limb stability
4. Ability to coordinate contraction and relaxation of innervated muscles at will

5. Control of shoulder movement patterns
6. A small degree of elbow flexor sparing





*Fig. 167 and fig 168*

MAS is usually used with a scoop dish & plate guard, a Dycem friction mat to stabilize the plate and a raised platform to bring plate up to a suitable level. Swivel cutlery can also be used. Initially a spoon can be used during training. With repeated practice and increased strength and coordination, a spork/fork may be used. Mirrors should be used during adjustments, donning the MAS and training where client participates actively in modifications and location of components.

With absent elbow and wrist flexors (Whalley 1981), a custom-moulded plastic arm trough (extends from fingertips to elbow) is used. MAS bracket can be used on wheelchair tubing of 22 mm diameter, which can be fixed to wheelchair backrest at a suitable height.

**Adaptive and Assistive Devices**

Following is a list of few adaptive and assistive devices prescribed and trained to clients with SCI in ADLs :-

**Bathroom**



- Commode
- Elevated toilet seat
- Grab bars
- Shower seat
- Bath mat
- Toilet safety fram
- Transfer bench
- Other





**Feeding/Drinking**

- Built-up Utensils
- 2-handled Mug
- Insulated Mug
- Guarded Plate
- Plate With Suction Cups
- Nosey Cups
- Weighted Utensils
- Foam Tubing
- Universal Cuff





**Personal care**



- Lotion / cream applicator
- Shampoo tray
- Long-handled sponge bath
- Auto-trimmer
- Toileting aids



**Dressing**



- Dressing stick
- Button hook
- Stocking aid
- Dressing stick
- Long shoehorn
- Sock aid
- Velcro shoes

**Kitchen**

- Adjustable portable table
- Jar opener
- Door knob extender
- Key holder
- Large remote control

**Mobility**



- Walker
- Crutches
- Rollator
- Transfer board
- Wheelchairs

**Bedroom**

- Bed tray
- Bed assist
- Bed rope ladder
- Other



**Miscellaneous**

- Adapted pen
- Card holder
- Leg lifter
- Reacher



*Fig. 169*

Biofeedback Training can be used to maintain upper extremity ROM, maintain and/or improve muscle strength and to improve sensibility in both upper extremities. The rationale behind using biofeedback is to stimulate neuroplasticity. It is known that, mass repetitive upper extremity task practice with sensory stimulation can bring about greater brain excitability in the corresponding brain sites and some improvement in function (Hoffman & Field-Fote, 2009).

Robotics arms can be used with the client for the same purpose.

**LEISURE ACTIVITIES**

Indoor games: A simple adaptation to mouth stick can allow clients to manoeuver chessman, draughts etc. A bird peak or pincer mouth stick, or a suck/blow vacuum wand can be used to manipulate playing cards or remove them from a holder. Several electronic/computer games are available that can be played with use of virtual reality with use of a helmet. This helps the clients to socialize with friends and also can be competitive.

Reading : Reading books or newspapers for a high-lesion quadriplegic is possible using a large bookstand or Posturite book ledger or a newspaper holder which holds the book/newspaper at a suitable height so that the pages can be turned with use of a mouth stick. Several companies also make electric page turners for people with severe disabilities.

Cassette tape books : In 21st century almost 99% of books are available online. Amazon kindle can be read using I-pad/laptop having touch options that can be controlled by a mouthstick. Moreover, Environmental Control Units (ECUs) can be used to control functions of a cassette player.

Photography : High-lesion quadriplegics can use several available devices to operate an electronic camera. It may be placed on a tripod that stands on a lap tray, or a mobile arm that can be mounted on to the side of the wheelchair.

Spotting scope : High-lesion quadriplegics who are interested in bird-watching, horse racing or such other activities can use mounted binoculars or a spotting telescope on a camera tripod. Different models of binoculars and telescopes are available with tripods that can move the telescope sideways and tilt it up and down.

Hunting, shooting and fishing : Hunting rifles can be modified for a high-lesion quadriplegic to allow the use of a mouthpiece to control the movement of a rifle sideways and upwards/downwards. The gun fires when the client sips on a vacuum tube. Fishing equipments are modified to enable client to operate a rod with a combination of chin and pneumatic controls.

Remote control vehicles : A simple joystick manipulated by mouth or chin can be used to operate remote control vehicles such as yachts or cars. The client can also compete at an outdoor activity alongside his able-bodied.

Music : Mouth organ (harmonica) or saxophone can be played as a form of incentive spirometry. Computer generated music is largely used worldwide. Self-supporting harness for a mouth organ or a guitar (where client is unable to hold the guitar) are available in music stores.

Blow darts : Lots of leisure activities can be performed with use of mouthsticks e.g reading, playing draughts and chess. Batavia (1987) has discussed the merits of using a short blow gun for target shooting darts which requires expiratory exertion (this in return increases the respiratory strength). Velcro darts can also be used.

Computers/Laptops : Several adapted items for computers, high-profile laptops and different softwares are available in the market to enhance access to communication, study, vocational opportunities and domestic duties (e.g. banking, online shopping). Some of these items are trackball, on-screen keyboard, voice-activated softwares, etc.

Spinal Orthosis : The type of spinal orthosis needed depends on the level and severity of SCI and whether it is with or without spinal bony injuries. These are used to provide support, restrict motion, facilitate fracture healing and maintain the spinal alignment. Following are the spinal orthosis generally prescribed according to the level of injury :-

1. Cervical : a) Rigid collars - Aspen collar/Philadelphia collar
   b) Halo brace
   c) Sternal-occipital-mandibular immobilizer (SOMI)
   d) Minerva orthosis
   e) Yale brace
   f) Soft cervical collar
2. Thoracic
3. Lumbar : Boston brace
4. Sacral

Upper limb orthosis/splints : Different types of splints are prescribed and fabricated for clients with SCI depending on their level of injury:-

1. Resting hand splint
2. Functional cock-up splint
3. Wrist-driven flexor hinge splint
4. Tenodesis splint
5. Cock-up with flexor assist splint

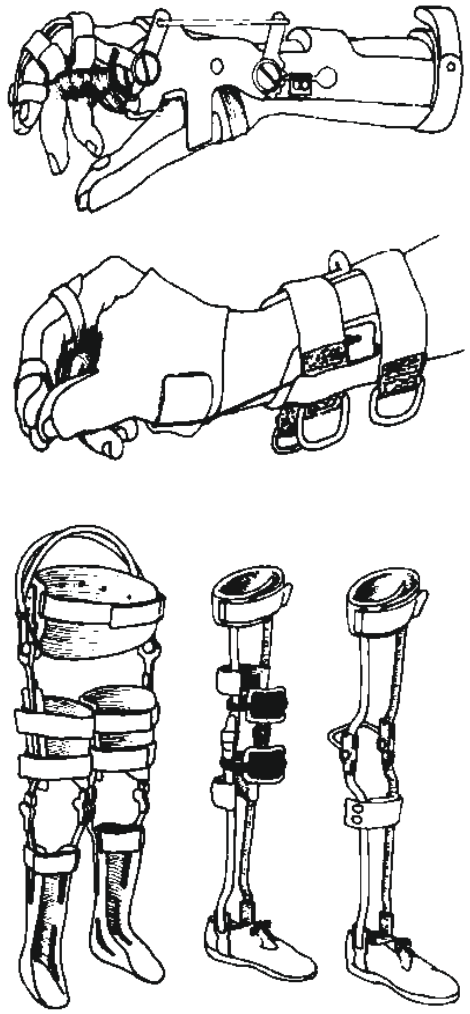


*Fig. 170, 171, 172, 173, 174 & 175*

a) Wrist driven flexor hinge splint

a) Left - Reciprocating gait orthosis

b) Tenodesis splint

b) Middle - Standard KAFO

c) Right - Scott-Craig orthosis

Lower limb orthosis : The external devices that are used to maintain functional limb position and joint ROM, to stabilize weak lower limbs, protect insensate areas against injury and allow client to walk independently with the help of walking aids.

Precautions :

a. Avoid orthosis that give less stability, protection or immobilization

b. Avoid contact with open wounds or pressure to soft tissues

c. Caution while accommodating areas with a fluctuating shape and size (like oedema, weight change, dialysis)

Following are the lower limb orthosis usually prescribed for functional mobility :-

1. AFO : used by clients with neurological level at or below L3
2. KAFO : can be used by clients with neurological level T11-L2 along with the walking aids
3. Hip knee ankle foot orthosis (HKAFO)

Walking Aids : Prescription and selection of the type of walking aid depends on stability, strength of upper and lower limbs and extent of improvement.

1.Crutches - These are used to reduce weight-bearing on one or both legs and to give additional support when lower limb strength is inadequate. Before prescribing crutches, the occupational therapist should assess the muscles needed to walk with crutches and even strengthen them. The muscles involved are :-

i) depressors of scapula

ii) extensors, flexors and adductors of shoulder

iii) elbow extensors

iv) wrist dorsiflexors

v) finger and thumb flexors

**Crutches used are :-**

a. Elbow crutches and,

b. Gutter crutches



*Fig 176 - Elbow crutch*



*Fig 178 - Gutter crutch*

**2. Walkers/ walking frame- Walking frames can be :-**

a. A gutter frame is the same as a standard walking frame. The tops of upper horizontal tubes on each side is modified by adding two gutters for the client to rest his forearms which are then fastened with velcro straps. Client takes more weight over forearms. This can be used when the client is unable to extend elbows fully, or unable to take weight on his hands/ has a weak or absent grasp.

b. The Reciprocal walking frame is selected when a client is unable to lift the walker or needs more stability. There are swivel joints between the horizontal and vertical tubes in the front making each side of the frame to move forward alternately.

Selection of walking aids depends on the level of injury and functional capabilities of walking. The functional outcomes of gait depends on the client's age, body built, spasticity and any existing medical condition and motivation. Table shows the possible gait patterns in a client with an SCI at different levels.

**Table 7 :**

| Level of Injury | Possible functional gaits |
|---|---|
| D1-D8 | Swing to gait with calipers & rollator; may use crutches if controlled spasticity |
| D8-D10 | Swing through/swing to gait with full length calipers and crutches |
| D10-L2 | Swing through/4-point gait with calipers and crutches |
| L2-L4 | 4-point/2-point gait with AFOs with crutches or sticks |
| L4-L5 | May need sticks/other aids with or without AFOs |

**Transfer aids :**

1. Slide sheets
2. Transfer boards
3. Hoists

**Wheelchairs :**

Wheelchair prescription and modification depends on the client's age (child/adult), level of injury (quadriplegia/diplegia), requirement (sports/vocation) and whether motorised controls (puff, joystick, voice-activated) are needed. It should be decided along with the client for achieving the highest level of independence in mobility. Types of wheelchairs include :-

1. Manual wheelchair (self- or attendant-propelled),
2. Powered wheelchairs (head, chin or hand-controlled),
3. Hand-powered tricycles and
4. Scooters.

Wheelchair mobility Skills : This includes propulsion, falling and negotiation of ramps, curbs and stairs. A wheelie is essential for managing uneven surfaces, ascending-descending ramps, curbs and to prevent front casters from getting stuck.

a. Independent wheelchair propelling

b. Ascending ramp :-

i) Lean forward as much as possible

ii) use short quick arm strokes to maintain forward momentum

c. Descending ramp :-

i) Lean backward as much as possible

ii) Allow pushrims to glide smoothly

iii) Grasp pushrim to slow down

iv) If grasp is weak, push medially on hand rims

v) On steep ramps, use a wheelie to prevent falling out of the chair or follow a zig-zag path

d. Ascending curb :-

i) approach in forward direction

ii) perform a wheelie to place casters up on a curb

iii) Lean forward and firmly propel rear wheels onto curb

e. Descending curb forward :-

i) Approach curb backward to the edge

ii) Lean forward and slowly propel backwards to lower rear wheels down

iii) Make a sharp turn to lower caster wheels down

f. Descending curb forward :-

i) Approach curb forward in a wheelie position

ii) Slowly lower rear wheels down curb

g. Ascending stairs :-

i) Approach stairs backwards

ii) Tilt wheelchair back against the firststep

iii) Use one hand to pull handrail while the other hand pulls on the handrail or the pushrim

iv) reposition hands for the next step and repeat

h. Descending stairs backwards :-

i) Approach the first step backwards

ii) Lean forward in chair

iii) Use both the hands on the handrail or one hand on the handrail and the other hand on the pushrims

I. Descending stairs forward :-

i) Approach stairs forward in a wheelie position

ii) Maintain hands on pushrims to hold the wheelie position

iii) Lower rear wheels down the steps

j. Ascend and descend stairs with assistance :-

i) The1st assistant stands behind the wheelchair and pulls on the pushrims with the wheelchair in a wheelie position

ii) The 2nd assistant stands in front of the wheelchair and ensures that the chair doesn't roll forward

k. Ascending and descending on buttocks :-

i) transfer from wheelchair onto floor

ii) bump up and down each step on buttocks

iii) pull the wheelchair to step beside buttocks after each step

Seating and positioning systems : These are the seating modifications and adaptations in wheelchairs required for being safe during wheelchair propulsion.

1. Pressure relief cushions

2. Head supports

3. Standing tables
4. Positioning belts
5. Supports for thorax, pelvis, hips and legs
6. Abduction wedge

## HOME MODIFICATIONS

The occupational therapist should visit the client's home to assess the architectural barriers that are restricting the client from being more independent in transfers, mobility and ADLs and reduce the risk of falls. After assessment of the client's needs, the occupational therapist, with the help of a contract with an architect and builder, makes modifications to the client's home. The home should be accessible to all persons regardless of their mobility status.

### General features

- Single floor dwelling is optimal, at least the bedroom, bathroom and kitchen should be on the same floor
- Doorways : 32"-36" wide
- Hallways: > 36" wide
- Light switches: 36"-42" from floor
- Electric outlets : > 15" from floor
- Window sills : <30" fom floor
- Thermostats : 36" from floor
- W/c 360 turning space : 60"*60"
- Adequate and even lighting in all areas
- Doors with lever-style handles (not knobs)
- Level or beveled threshold < ½" in height
- Doors with offset hinges to maximize width of open doorways

### Entrances

- Ramp inclination : 1:20 or at least 1:12 (for every 1" of vertical raise, 12" ramp)
- Railings : 29"-36"
- Curbs on ramps : 4" height
- Landings/platforms : 90 turn - 4'*4' and 180 turn - 4'*8'
- Ramp width : 36"
- Divide long ramps into 10-12 foot sections with platforms in between
- Nonslip ramp surface with fire-resistant
- 5*5 feet2 platform space for door management at ramp's top and bottom
- Easy-to-reach door locks

### Bathrooms

- Installed next to the bedroom
- Size : 8*10 feet2
- Sinks with an open space underneath for knee clearance
- Sink pipes behind walls
- Faucet handles are of lever-type
- Standard height toilet, if commode chair used
- Tub/shower seat, built-in or set-in
- Roll-in shower : > 5*5 feet2, nonslip floor
- Hand-held shower head with 6 foot hose and built-in on/off control
- Thermostat set for < 120 water temperature
- Mirrors and cabinets are set low
- Strategically placed grab bars

### Kitchen

- Countertops : 30"-35" above floor with 24" space underneath for knee clearance
- Cabinet doors and drawers easy to open/close and placed at proper heights
- Stove height : 30"-32" with large front-mounted control knobs
- Ovens set low at the individual's

discretion

- Side-to-side refrigerator with self-defrosting

**Bedroom**

- Maximum size : 10*14 feet2 with 5*5 feet2 clear area and a 3 foot wide passage to bed and bathroom
- Closets with accessible shelves < 4 feet 6 inches from floor and rods for hanging clothes < 4 feet from floor
- Telephone and light switches should be easily reachable from the bed

**Stairs**

- Sturdy handrails extending beyond the top and bottom of stairs
- Light switches placed at the top and bottom of stairs
- Step height : 6"-7" with width > 10"

**Laundry**

- Preferably located on the main floor
- Front-loading washers and dryers
- Table for folding clothes : 30''-32'' height

## DRIVER REHABILITATION AND VEHICLE MODIFICATIONS

After SCI, a driving assessment is done by an occupational therapist qualified in driving to determine whether the client is ready for driving and recommend certain adaptations and modifications with driving simulation training to enable the client in independent driving. OT driving assessments can be done through the local CRS office or through a specialized driving center.

Models :-

1) Model of Human Occupation (MOHO)
2) The Person-Environment-Occupation Model
3) The Canadian Model of Occupational Performance (CMOP)
4) Occupational Adaptation Model

On the basis of these models, occupational therapists assess the measurable driving potential of the clients with SCI and select an appropriate vehicle-type and the appropriate vehicle modifications. Assessment of the individual's driving readiness includes :-

a. History of medical problems, present medical conditions and medications

b. Sensibility assessment :-

1) Tactile sensation

2) Light and deep pressure sensation

3) Stereognosis

c. Motor assessment :-

1) ROM

2) MMT

3) muscle tone

4) proprioception

5) Kinesthesia

6) Endurance/tolerance

7) Sitting balance

8) Standing balance

9) Ambulation status

d. Brake Reaction Time

e. Visual assessment :-

1) Acuity

2) Peripheral Vision

3) Color

f. Cognitive and visual perceptual skills assessment

Standardized driving assessments :-

1) Cognitive Behavioral Drivers Inventory

2) driveABLE

3) Driving Health Inventory

4) Driver Performance Test

5) MVPT

6) Hazard Perception Test

A spinal cord injury client can even drive a truck based on their functional abilities




*Fig 179, 180, 181*

If a client with SCI does not meet the criteria to be fit for driving, he/she should undergo rehabilitation process. Occupational therapists aim at treatment in following areas :-

1. Visual rehabilitation
2. Cognitive behavior therapy
3. Transfer training
4. Driving simulation training
5. Visual perceptual rehabilitation
6. Vehicle modifications

**Car Modifications**

All car modifications should meet RTA guidelines. All modifications are recommended after assessment of clients level of functioning and needs, which are carried out by private companies or local car modification centres.

- Hand controls to operate accelerator and brake. This may include the MPS Monarch, Push-Pull or (twist grip) controls
- Steering modifications and adaptations :- A spinner knob, fork attachment or C bar attachment to the steering wheel
- Primary and secondary control devices like dimmer, horn, parking brake, door locks, mirror, head rest, wheelchair securements, etc.
- Some vans have automatic wheelchair lockdown restraints installed, automatic ramp, swivel seat and many options.
- A mobile phone is recommended in case of emergency whilst driving
- Disabled parking - a sticker is available from RTA and enables a client with disability to park in Accessible parking spots

**TENDON TRANSFER**

Tendon transfers with or without nerve transfers is an option for treating hand and

arm dysfunctions in clients with a SCI. Early hand recovery is necessary for performing and restoring ADLs. In complete injuries, where there is little chance for upper limb recovery, until the clients achieve a stable functional level, it is necessary to evaluate the client and classify for the group of tendon transfers according to the International Classification (IC) of surgery of the Hand in Tetraplegia. Evaluation should be done thoroughly to find the donor tendon or nerves to be transferred. Post-operativey, the occupational therapists or hand therapists collaborate with the hand surgeon to decide the most possible transfer to be performed and carry on the post-operative rehabilitation to restore function in upper limbs of clients with quadriplegia.

**Evaluation : This includes :-**

1. Muscle strength of donor muscle(s)
2. Joint ROM
3. Sensibility testing
4. Others : Identification of brachial plexus injuries and entrapment neuropathies, paralytic spine deformity, thoraco-scapular stability, spasticity, contractures, stiffness and joint instability

**Table 8 : International Classification of surgery of the Hand in Tetraplegia - with addition of sources for nerve transfers**

| *Group* | *Spinal Cord Segment* | *Possible Muscle transfers* | *Possible Axon sources for Nerve transfers* |
|---|---|---|---|
| 0 | ≥ C5 | No transferable muscle below elbow | Musculocutaneous nerve branches to Coracobrachialis & Brachialis muscle |
| 1 | C5 | Brachioradialis (BR) | Axillary nerve branches to deltoid and teres minor muscles |
| 2 | C6 | • Extensor Carpi Radialis Longus (ECRL) | Radial nerve branches to supinator muscle |
| 3 | C6 | • Extensor Carpi Radialis Brevis (ECRB) | |
| 4 | C6 | • Pronator Teres (PT) | |
| 5 | C7 | • Flexor Carpi Radialis (FCR) | |
| 6 | C7 | • Extensor Digitorum | |
| 7 | C7 | • Extensor Pollicis Longus | |
| 8 | C8 | • Flexor Digitorum | |
| 9 | | Lacks intrinsics only | |
| 10(X) | | Exceptions | |

Reconstructive surgeries (RCS) for upper limb motor function in clients with quadriplegia involves multiple surgeries to restore shoulder, arm and hand functions so that the client is able to perform ADLs independently. The main goals of RCS are to provide :-

a. Elbow extension

b. Grip function (flexion phase)

c. Opening of hand (extension phase)

**Prerequisites :** The following are the prerequisites for RCS for upper limb function in quadriplegia :-

1. Neurological functional plateau - No further recovery expected
2. Emotional stability - accepting the consequences of injuries
3. No open wounds or pressure sores/ decubitus ulcer or infections (e.g. bladder)
4. Motivation and ability of the client to take part in post-op treatment
5. Treatment plan based on clinical examination and counselling of the client
6. Available donor muscles (muscle strength >4/5)
7. Free joint passive mobility

**Table 9 : Summary of possible surgical procedures to achieve patients' ability goals**

| Ability goal | Functional goal | Procedures | Post-op rehabilitation (9) |
|---|---|---|---|
| Stabilizing elbow in space, reaching overhead, pushing wheelchair, stabilizing trunk with arms | Elbow extension | <u>Reconstruction of Triceps function</u><br>• Posterior deltoid ->triceps<br>• Biceps ->Triceps | 4 weeks cylindrical cast with elbow fully extended<br>4 weeks posterior elbow conformer |
| | | Axons of axillary nerve -><br>Brachialis branch of musculocutaneous nerve -> | |
| Use of utensils, handwriting, pushing wheelchair | Grip | <u>Reconstruction of grip</u><br>1. Reconstruction of passive key grip<br>Brachioradialis->Extensor CarpiRadialisBravis<br>FlexorPollicisLongus->Radius (Moberg procedure)<br>1st carpometacarpal arthrodesis<br>Reconstruction of active key grip<br>Brachioradialis (BR)->FlexorPollicisLongu (FPL)<br>1st carpometacarpal (CMC) arthrodesis<br>Split FlexorPollicisLongus-ExtensorPollicisLongus(EPL) tenodesis | 1. 4 weeks cast with flexed thumb and wrist<br>4 weeks active exercises of gross grasp |

| | | | |
|---|---|---|---|
| | | 1.Brachialis motor branch->ExtensorCarpiRadialisLongus motor branch<br>2. FlexorPollicisLongus (FPL)->Radius | |
| Feeding, writing etc. needs pronated arm | Forearm pronation | 1. Distal transposition of biceps tendon (if needed, with interosseous membrane release)<br>2. Dorsal transposition of brachioradialis (during BR->FPL)<br>3. Derotation osteotomy of radius | |
| Reaching for objects to hold like cup, glass | Opening of hand | <u>Reconstruction of thumb & finger extensors</u><br>1. Passive opening<br>1st CMC arthrodesis<br>EPL->Extensor retinaculum<br>Active opening<br>PronatorTeres->extensor digitorum communis (EDC) & EPL/APL<br>Thumb stabilization<br>1st CMC arthrodesis<br>Reconstruction of intrinsics<br>Zancolli-Lasso tenodesis<br>ExtensorDigitiMinimi->AbductorPollicisBrevis | 1. 4 weeks cast with thumb and wrist<br>2. 4 weeks cast with wrist, fingers and thumb<br>6 weeks splint for IP/1st CMC<br>4 weeks cast with fingers in intrinsic plus position<br>4 weeks cast with thumb opposed and wrist flexed |
| | | Supinator motor branch (C6) to Posterior Interosseous nerve (C7-C8) | |

**AQUATIC THERAPY FOR SPINAL CORD INJURY PATIENTS**

Aquatic therapy means making use of different physical and chemical properties of water to facilitate functional recovery and independence in individuals with Spinal Cord Injury. Aquatic therapy is defined by The Aquatic Therapy and Rehabilitation Institute as "The use of water and specifically designed activity by qualified personnel to aid in the restoration, extension, maintenance and quality of function for persons with acute, transient, or chronic disabilities, syndromes or diseases".

Aquatic therapy does not mean just swimming in water. Aquatic therapy includes purposeful therapeutic movements or exercises performed in order to achieve optimum benefits for the individuals with Spinal Cord Injury. There are various techniques in aquatic therapy. An exercises session will consist of a combination of these

techniques and approaches like Water specific therapy, Bad-Ragaz ring method, Clinical Ai-Chi, Aquatic exercises, Aqua aerobics and Passive relaxation or Watsu. Mostly in Spinal Cord Injury an exercise session will be conducted one - on - one by the therapist but group sessions may also be conducted to improve participation and peer interaction.

**Aim of Aquatic Therapy-**

1. To improve muscle strength- Water has some amazing physical and chemical properties out of which viscosity is one. Viscosity can be an excellent strengthening tool. The resistance provided by the water is dependent upon the speed of the movement and direction of the movements which are patient dependent. In painful conditions if the patient stops the movement the resistance drops to zero and therefore the strengthening activities can be performed within the limits of patient's tolerance.

Several equipments like aquatic webbed gloves, aquatic dumbells, Fins, noodles and aquatic weight cuffs can be used for strengthening muscles.



*Fig. 182 and fig 183*

- Aquatic webbed gloves (greater the surface area greater the resistance)- It is used in both paraplegics and quadriparetic patients to strengthen upper limb muscle.



*Fig.184*

- Water Noodles (it can also be used for balance training)



*Fig.185*

- Fins- Mainly used in patients with paraparesis. It provides great resistance for lower limb muscles while walking and also helps in initiating dorsiflexion of ankle.





*Fig.186 and 187*

- Water Dumbbells- It is a great tool for strengthening shoulder girdle.



*Fig.188*

**Water weight cuff-** Osteoporosis is very common in patients with spinal cord injury. These water weight cuffs are used to prevent floating of the legs and keep them grounded during gait training.

2. **To improve trunk control-** Water specific therapy and Bad-ragaz ring method can be well used to improve trunk control by strengthening trunk musculature. The waves in pool make it difficult to balance due to which they have to constantly use muscle to prevent from falling.





*Fig 189 and Fig 190*

**WST (Sagital rotation)**

3. To improve aerobic capacity- Patients with spinal cord injury in the higher level will be having breathing difficulty. Compressive forces of the water provide resistance to the respiratory muscles (muscles required for breathing) and help strengthening these muscles. Expiratory flow is poor because of poor abdomen muscles. Water has a property known as Hydrostatic pressure which provides pressure in all the part of the body which is immersed in water. Therefore, it provides assistance to take the residual air out of the lungs and improving the intake. Aquatic therapy also improves cardiovascular endurance

4. To reduce spasticity- A session in warm water can help reduce the spasticity. Watsu is a passive relaxation technique which can be used to bring the tone down and improve the performance on land.

To improve balance while sitting and standing- Turbulence can be used very well to give balance training.





*Fig 191 and Fig 192*

- Sitting balance training



*Fig 193*

- Standing balance training

5. To Improve gait in patients with paraparesis, quadriparesis and initiate walking in complete paraplegia- Water resistance helps improving lower limb muscle strength required for ambulation. Buoyancy gives assistance in lifting the leg hence making it easier to perform. Fins can be used for gait training as the resistance provided by fins results in better hip knee flexion pattern.

In patients with complete paraplegia splints may be required for gait training to prevent knee buckling.

6. Other benefits of aquatic therapy-

- Aquatic exercises increase serum endorphin (hormone responsible for mood elevation) levels and levels of neurotransmitter serotonin which are responsible for reducing pain and controlling many functions like sleep, appetite but mainly mood. Aquatic exercises have a psychological mood elevating effect.
- Increased blood supply, hydrostatic pressure of the water, improved endorphins and serotonin levels also helps to improve appetite and bowel movements. In the later stages of the disease it can prevent complications like severe constipation.

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# Ch. 5 Non-Traumatic Spinal Cord Injury

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**Medical management Non-traumatic spinal cord injury**

Non-traumatic spinal cord injury can be the result of infection to the spinal cord or infection to the spinal bones that compress the spinal cord and may damage it permanently. It could also be due to cysts and tumors of the spinal cord or spinal bones and other connective tissue surrounding the cord.

Medical Management of acute spinal cord injury: After the spinal cord injury, emergency care is the most important predictor for the further recovery. A spinal cord trauma is a medical emergency requiring immediate treatment to reduce the long-term effects. To prevent further neurological injury and enhance neural recovery short-term medical treatment includes anatomical realignment and stabilization interventions of the spine and pharmacological management.

**Prehospital management:**

At the sight of the accident it is difficult to know about the spinal cord injury and its extent. Mechanism of injury, pain in the vertebral column, or neurologic symptoms can help the emergency medical personnel regarding assumed spinal injury. Complex and severe trauma those resulting from falling from heights should be suspected for spinal cord damage involving the head and pelvis. Immediate stabilization and immobilization of the spine at the sight of injury and transporting the patient to the emergency department is very much important. Most often damage to the spinal cord is due to the poor handling and transport.

**Emergency department management:**

A.ABC resuscitation: airway, breathing and circulation should be assessed at sight of the accident and if needed it should be started then and there. Resuscitation is focused at airway maintenance, adequate oxygen saturation of peripheral blood, restoring blood pressure to acceptable limits, preventing bradycardia. Performed simultaneously to prevent any further ischemic damage to the previously injured spine. Oral secretions are cleared to maintain airway patency and to prevent aspiration. When indicated intubation should be carefully performed to avoid spine movement and further injury to the spinal cord.

Before diagnosing for neurogenic shock patients with acute spinal cord injury should be over ruled for hemorrhagic and/or neurogenic shock.

I. Excluding hemorrhagic shock: the most common sources of occult hemorrhage like chest, abdominal, retroperitoneal injuries and fractures of the pelvis or long-bones may be revealed in computed tomography (ct) scan or x-ray.

II. Neurogenic shock: history of spinal cord injury with exclusion of hemorrhage suggests spinal shock

**Management of neurogenic shock**

I. Fluid resuscitation: fluid resuscitation

with isotonic crystalloid solution is the initial treatment of choice for neurogenic shock. These patients are at risk for the acute respiratory distress syndrome (ARDS) and hence volume overload should be avoided with judicious fluid resuscitation.

ii. Maintaining blood pressure: maintain systolic blood pressure above 90 mm hg and avoid hypotension.

iii. Maintaining oxygenation: mechanical ventilation and/or supplemental oxygenation is required to maintain adequate oxygenation and perfusion of the injured spinal cord.

iv. Monitoring heart rate: heart rate should be 60-100 beats per minute (bpm) in normal sinus rhythm. Atropine can be given for treatment of hemodynamically significant bradycardia.

v. Maintaining urine output: insert Foley catheter to decompress the bladder. Urine output should be monitored and maintained more than 30ml/h.

vi. Inotropic support: a low dose of dopamine in the 2-to 5-mcg/kg/min range is usually sufficient for patients who have decreased urinary output despite adequate fluid resuscitation.

B. Assessment and management of associated injuries: Spinal cord injury is most commonly associated with head injury. A detailed evaluation for intracranial injury should be done in the presence of amnesia, external signs of head injury or basilar skull fracture, focal neurologic deficits, associated alcohol intoxication or drug abuse, and a history of loss of consciousness. Corticosteroid therapy: Methyl prednisolone is beneficial in causing favorable neurological outcome if administered early enough (within 8 hours) of the primary insult. It helps by reducing damage to nerve cells and by decreasing inflammation near the site of injury. Monosial-otetrahexosyl ganglioside (gm-1): gm-1 administered following acute spinal cord injury as an adjunct after the administration of methyl-prednisolone may be of benefit. Methyl-prednisolone enhances the flow of blood to the injured spinal cord, preventing the typical decline in white matter, extracellular calcium levels, and evoked potentials, thus preventing progressive post traumatic ischemia.

The therapist must be aware of side effects that may occur with such high doses of steroids, including gastric ulcers, decreased wound-healing time, hypertension, cardiac arrhythmias, and alteration in mental status.

Deep vein thrombosis prophylaxis: low molecular weight heparin can be used to prevent deep vein thrombosis.

Spinal traction: spinal traction is a more conservative and less invasive approach to allow the bones to heal naturally without surgery. Traction prevents movement of the spine. The skull may be held in place with tongs (metal braces placed in the skull and attached to traction weights or to a harness on the body). The spine braces may need to be worn for a long time. The other type is halo traction device which has three parts: the ring, the uprights, and the jacket. The ring fits around the skull, just above the ears and is held in place by four pins that are inserted into the skull. The uprights are attached to the ring and jacket by bolts. The jacket is usually made of polypropylene and lined with sheepskin. It is left in place for 6 to 12 weeks until bony healing is satisfactory. The advantage of using the halo device is the ability to mobilize the client as soon as the device has been applied without compromising spinal alignment. This allows the rehabilitation program to commence more rapidly. It also allows for delayed decision making regarding the need for

surgery.

Surgical management: The treatment of spinal cord injury patients may require surgery.

**There are three major goals of surgery**

A. Spinal decompression: Involves removal of fluid tissue, bone fragments, disk fragments, or foreign objects that compresses the spinal cord.

B. Prevention of tethering: Surgery may prevent tethering of the spinal cord. Neurological disorders related to pulling of the spinal cord at the base of the spinal canal can cause stretching and damage which can be prevented.

C. Stabilization of the spine: The fractured vertebrae may not be capable of supporting the normal weight from the body and protect the spinal cord and can further lead to severe injuries. Spinal instrumentation and fusion can be used to provide permanent stability to the spinal column. The procedure helps to correct, join, and solidify the level where a spinal element has been damaged. Metal screws, rods, plates or cage or combination of all may be necessary to hold the vertebrae together and stabilize them until the bones heal.

D. The type of surgery performed (anterior vs. Posterior), distraction forces during surgery, preoperative grade all influence the outcome.

E. The surgeries for Spinal decompression: The major types of surgery for spinal decompression are

Microdiscectomy / corpectomy: Laminectomy Stabilization of the spine: can be achieved by spinal fusion with or without instrumentation.

i. In the craniovertebral junction, stabilization may be achieved by

a. C1-c2 fixation using pedicle screws/ transarticular screw/ sub laminar wires

b. Occipito cervical fixation

ii. In the cervical spine, stabilization may be achieved by

a. Anterior approach using vertebral body plates and screw

b. Posterior approach using lateral mass screws

iii. In the dorsal spine, stabilization may be achieved by

a. Anterior transthoracic approach for interbody cage placement

b. Posterior approach using transpedicular screw and rod system

iv. In the lumbarspine, types of stabilization procedures are

a. Anterior lumbar interbody fusion (alif)

b. Posterior lumbar interbody fusion(plif)

c. Transforaminal lumbar interbody fusion (tlif)

d. Transforaminal interbody fusion

**Postoperative physiotherapy:**

Post-operative physiotherapy starts immediately after the surgery and is continued during the convalescent period. Respiratory management plays a very important role to prevent post-operative complications. Restorative physiotherapy starts immediately after the patient is stabilized. Acute neurorehabilitation for spinal cord injury Therapeutic rehabilitation should be effectively delivered at the beginning in an acute care setting at the time of injury. Inpatient rehabilitation begins during the critical and acute care stages after an sci. The primary emphasis of early rehabilitation is to lessen the adverse effects of neuro trauma and immobilization. This focus may last from a few days to several

weeks, depending on the

severity and level of injury and other associated injuries.

During the acute phase patients may begin participating in early therapy that should include,

a. Respiratory care,

b. Prevention of indirect impairments and complications

c. Range-of-motion (rom) exercises and initiating active exercises in available musculature

d. Normalization of tone

e. Early strength training

f. Gaining upright tolerance,

g. Out-of-bed activities

h. Sensory reeducation,

I. Skin-management education

Goals during this phase should focus on prevention of secondary complications and preparing the client for full rehabilitation participation. The treatment team must begin discharge planning and family training in this phase. Respiratory care should include deep breathing exercises, glossopharyngeal breathing, strengthening respiratory muscles, assisted coughing, abdominal support and stretching of the thorax. Full range of motion exercises should be completed daily by the patient if not possible should be performed by the therapist except in those areas that are contraindicated. With paraplegia, motion of the trunk and some motions of the hip are contraindicated. Straight leg raising more than 60 degree and hip flexion beyond 90 degrees (during combined hip and knee flexion) should be avoided. To normalize the muscle tone facilitatory or inhibitory techniques should be used as per the need. Every 2 hourly patients should be positioned and splinted in the neutral position. The use of an abdominal binder, compression garments and elastic stockings will retard venous pooling and prevent deep vein thrombosis. Upright activities should be initiated by gradually elevating the head of the bed and progressing to tilt table or reclining wheelchairs. Vital sign should be observed and recorded during these movements. This will also help to decrease postural hypotension after the continuous bed rest. Gradual strengthening program should be followed for the weak muscle which will help in the later phase of rehabilitation. The bed mobility should be initiated with proper precautions. Activity based training should be initiated to reinforce the movements from the paralyzed muscle. Mental imagery, mirror therapy and contemporary task-oriented approach of training for the paralyzed limb can be focused to get better results in active phase of rehabilitation. Careful monitoring of autonomic dysreflexia, postural hypotension and hypothermia is important and should be reported to other medical care professionals.

**Intervention for sensory issues:**

Sensory training is a vital part of neurorehabilatation in the individuals with spinal cord injury as the sensory impairment becomes the major issue and makes these individual prone to many other risk factors. The sensory rehabilitation emphasize to normalize the sensation and can help to get better motor response.

The techniques used are Compensatory techniques: used when there is loss or impairment of protective sensation.

The primary goal of compensatory techniques is to prevent injury

- Safety first
- Increase awareness of deficit
- Minimize risk of tissue damage (like continuous low pressure, concentrated

high pressure, excessive heat or cold, repetitive mechanical stress, pressure on infected tissue)

**Compensatory methods and techniques include:**

- Use of other senses like vision (observe for signs of inflammation from repetitive pressure, excessive force, and if these signs occur rest the hand) and hearing to rubbing sounds while propelling wheelchair)
- Should not handle sharp objects, or directly check temperature of hot water or it may cause cuts or burns respectively in hand
- Using adaptive devices like hand cuff to prevent callous formation due to prolonged use of manual wheelchairs, built up handles to distribute the pressure as well as to enhance the grasp
- Skin care: use of protective glove mitts to prevent blister formation while handling hot utensils, regular moisturizing of skin to prevent dryness of hand
- Avoid gripping for long duration instead use modified handles and grip aids

**Sensory reeducation techniques:**

It can be started when the individual can appreciate deep, moving touch. The modalities are first used with eyes closed, then eyes open and then again eyes closed. This technique can be used to develop good touch localization after the individual has developed a faint perception of light moving touch.

Modalities for sensory reeducation include the following:

- Blunt end of pencil for training for graphesthesia
- Dowels of different textures
- Different fabrics
- Objects of different rough/ smooth edges
- Games
- Performing activities of daily living with vision occluded

**Desensitization techniques:**

Few patient experience hypersensitivity at or above the level of injury, or neurogenic pain above the level of injury Goal of this technique is to increase the pain threshold of an individual and reduce the discomfort due to pain. The technique progresses from desensitizing the affected area using a soft texture, followed by coarse and finally rough texture and one can gradually increase the force, duration and frequency of application.

Following modalities are used for desensitization:

- Therapeutic massage
- Percussion/ tapping/ stroking with different textures
- Vibration
- Pressure/ compression
- Tens
- Fluid therapy

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# Section 3

# Disorders of Central Nervous System

# Chp. 6 Stroke

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**Definition:**

Stroke is defined as sudden neurological deficit caused by focal vascular lesion in the brain. The term 'stroke' is synonymous with cerebrovascular accident (CVA) and is purely clinical definition which, according to the World Health Organisation, can be defined as a 'rapidly developed clinical sign of a focal disturbance of cerebral function of presumed vascular origin and of more than 24 hours duration' Included within this definition are most cases of cerebral infarction, cerebral haemorrhage and subarachnoid haemmorhage, but deliberately excluded are those cases in which recovery occurs within 24 hours. These are designated as 'transient ischaemic attacks' (TIA). The vascular lesion could be either hemmorhagic or thromboembolic phenomena (leading to ischaemia) involving the blood vessels supplying various parts of the brain. The extent of neurological involvement may range from mild motor deficit to gross involvement of various function, namely Sensorimotor, perceptual, emotional, behavioural, memory intelligence, speech & language function.

**Etiology:**

The brain is a unique organ in which functioning of the neurons depend on a continuous blood supply because metabolism is almost exclusively aerobic. Different mechanisms have been found to cause vascular insufficiency to the brain resulting in stroke. However, the most common causes are:

1. Thrombus: This is mainly due to the presence of atherosclerotic plaque in the cerebral arteries as a result of severe platelet adhesion, fibrinous coagulation & decreased fibrinolysis activity.

2. **Emboli:** These are free flowing bodies in the cerebral blood stream in the form of dislodged thrombus, fats, air, tissue particle, etc which gets trapped at any one point along their course, frequently at the bifurcation of the arteries, & cause occlusion to the cerebral Vessel.

3. **Hemorrhage:** It occurs due to the rupture of the blood vessels in the brain, commonest type being subarachnoid haemmorhage. Following hemorrhage tissue death occurs due to both ischemia & mechanical injury to the brain substance as a result of compression by the clot. Hemorrhage usually occurs either due to hypertension, arteriovenous malformation or due to trauma.

4. There are some risk factors that can predispose to stroke. The common ones are diabetes, high blood pressure & cardiac disorders. Modifiable risk factors are cigarette smoking, obesity, sedentary lifestyle & excessive alcohol intake.

5. *Pathophysiology of Cerebral Ischemia &* consequent infarction:

6. The two pathophysiological changes leading to cerebral infarction are loss in the supply of oxygen & glucose due to vascular occlusion, & various changes in cellular metabolism resulting in consequent collapse of energy producing processes with disintegration of cell membrane.

Complete occlusion to brain substance causes severe damage to it with a zone of infarction, which is however found to be smaller than the actual area supplied by the involved artery. The margin of this infarcted zone consists of cells that are alive but metabolically less active. These surrounding areas are termed as ischemic penumbra. These areas are nourished by meningeal collaterals.

The necrotic tissue swells rapidly mainly due to excessive intercellular water content. Also, lack of oxygen is another factor that could contribute to swelling. This vascular lesion to the brain causes release of neurotransmitters like glutamate & aspartate by the ischemic cells, which excites neurons & produces an intracellular influx of Na & Ca leading to irreversible cell damage. Thus recent research attempts at blocking this action of gluatamate & aspartate on nearby cells, which will reduce the secondary involvement of surrounding viable cells.

Cerebral edema begins within few minutes & reaches a maximum by about 4 days, however it mostly disappears by 3 weeks. This edema can increase the intracranial pressure & can even cause contralateral & caudal shift of brain structure.

*Neurovascular Syndromes:*

Cerebral Blood flow (CBF) is controlled by a number of autoregulatory mechanisms (cerebral) that modulate a constant rate of blood flow through the brain. These mechanisms provide homeostatic balance, counteracting fluctuations in systolic blood pressure while maintaining a normal flow of 50 to 60 ml / 100 gm of brain tissue per minute. The brain has high energy requirements & very little metabolic reserves. Thus, it requires a continuous rich perfusion of blood to deliver oxygen & glucose to the tissues. Cerebral blood flow represents approximately 17 % of available cardiac output.

Depending on the areas of the brain & the arteries affected clinical symptoms vary.

**Synergy Patterns:**

Stroke being a central Nervous system disorder, following the acute stage of flaccidity, there is severe spasticity in the affected parts of the body. This leads to abnormal,

**A) Occlusion of proximal Middle Cerebral Artery (MCA)**

Table 1 : Clinical Manifestations of Middle Cerebral Artery Syndrome

| **Structures involved** | **Neurological deficit** |
|---|---|
| Motor area of face, arm, and fibres descending from leg area to enter the coronal radiata | Paralysis of contralateral face, arm and leg |
| Somatosensory area of face, arm and face leg | Sensory impairment over contralateral and leg |
| Motor (Broca's) area on dominant hemisphere | Motor speech disorder |
| Central language area and parieto-occipital cortex of the dominant hemisphere | Central aphasia, word deafness, anomia, jargon speech, alexia, agraphia, acalculia, and finger agnosia |
| Non-dominant parietal lobe | Perceptual disorder like unilateral neglect, anosognosial unawareness of hemiplegic side, apraxia, and spatial disorganization |
| Occipital lobe | Homonymous hemianopia and loss of conjugate gaze to the opposite side |
| Parietal lobe | Ataxia of contralateral limbs |
| Bilateral frontal lobe | Brun's ataxia or apraxia of gait |
| Supramarginal gyrus or inferior parietal lobe | Loss or impairment of optokinetic nystagmus |
| Posterior limb of internal capsule and adjacent corona radiata | Pure motor hemiplegia without sensory and visual involvement |

**B) Clinical Manifestations of Anterior Cerebral Artery Occlusion are as follows:**

Table 2 : Clinical Manifestations of Anterior Cerebral Artery Syndrome

stereotyped, primitive,

mass movement pattern, called Synergy patterns. Synergy can be either flexor or extensor. Synergy normally involves abnormal movement patterns that are not useful for functional activities.

The abnormal hemiplegic attitude is due to combination of strongest component of the flexor and extensor synergy in both upper and lower limbs. The strongest components for the upper limb is the flexor synergy and for lower limb is extensor synergy. Hence, a hemiplegic patient has an attitude of shoulder retraction and depression, shoulder adduction and internal rotation, elbow flexion, pronation and wrist and finger flexion. The lower limb adopts an attitude of pelvic rotation, hip extension, adduction and external rotation, knee extension, and ankle plantarflexion and inversion.

**Comparison of Normal and Hemiplegic Gait :**

Abnormal Synergy patterns of lower limbs leads to abnormal Gait. Therefore, understanding the influence of synergy patterns on gait can help to retrain for proper gait patterns. Walking is an independent, automatic, symmetrical and economical event. During

**C) Clinical Manifestations of Posterior Cerebral Artery Occlusion are as follows:**

Table 3 : Clinical Manifestations of Posterior Cerebral Artery Syndrome

| **Structure involved** | **Neurological deficit** |
|---|---|
| Thalamus | Hemianesthesia (contralateral sensory loss) or thalamic sensory syndromes (unpleasant hemibody sensation with spontaneous pain) |
| Occipital cortex | Homonymous hemianopia, visual agnosia, prosopagnosia |
| Bilateral occipital cortex involvement | Cortical blindness |
| Temporal lobe ischaemia | Amnesic syndrome with memory defect |
| Midbrain | Skew deviation, athetoid posturing, postural tremor, hemiballismus) |
| Cerebral peduncle | Contralateral hemiplegia |
| Motor tract between red and vestibular nuclei | Decerebrate attacks |

**D) Lacunar Syndromes:**

Lacunar syndromes are caused by small vessel disease deep in the cerebral white matter **(penetrating artery disease). Lacunar syndromes are consistent with specific anatomic sites.**

Table 4: Clinical Manifestations of Lacunar Syndrome

| **Structure Involved** | **Neurological deficit** |
|---|---|
| Posterior limb of internal capsule, pons & pyramid | Pure motor lacunar stroke |
| Venterolateral thalamus or thalamo-cortical projections | Pure sensory lacunar stroke |
| Base of pons, genu of anterior limb or the internal capsule. | Dysarthria / Clumsy hand syndromes |
| Pons, genu of internal capsule, coronal radiate or cerebellum. | Ataxic Hemiparesis |
| Junction of internal capsule & thalamus | Sensory / Motor Stroke |
| Putamen, global pallidus, subthalamic nucleus. | Dystonia Movements |

a normal walking there is both vertical and horizontal displacement of center of gravity in the form of a sinusoidal wave that causes horizontal displacement of 1.7 inch and vertical displacement of 1.8 inch. The gait determinants are as follows:

- Knee flexion at heel strike on loading which lowers the CG.
- Pelvic tilting
- Pelvic Rotation which increases with increase in the heel strike.
- Foot ankle mechanism
- Knee flexion on heel rise or heel off.
- Lateral displacement of pelvis

Under normal circumstances walking is an automatic process which requires contribution and integration of various mechanisms, however after the brain damage like in cases of stroke all the automatic adaptation is lacking and the patient tries to compensate for it by putting more voluntary efforts. The phasic action

Table 5: Synergy patterns

| **Synergy type** | **Upper Limbs** | **Lower Limbs** |
|---|---|---|
| Flexion Synergy | Shoulder girdle retractor and elevation, shoulder abduction, external rotation, supination flexion of elbow, wrist and finger flexion. | Hip flexion abduction and lateral rotation, knee flexion, dorsiflexion and inversion |
| Extension Synergy | Shoulder Girdle protaction and depression, Shoulder adduction, internal rotation, elbow extension, pronation, wrist and finger flexion. | Hip extension, adduction, internal rotation, knee extension, ankle plantarflexion, inversion and toe plantarflexion. |

of muscle group seen normally is replaced by abnormal muscular combination characteristic of primitive reflexes. The flexion and extension synergy interferes with the walking and greatly influence the hemiplegic gait.

The abnormality of gait in hemiplegic patient can be compared with normal gait taking into consideration the contribution of the joints for various phases of gait cycle.

***Ankle Joint:***

*Early stance (heel strike to foot flat):*

In normals when the heel strikes the ground, the angle at the ankle is 90 degrees once the weight is transmitted to the fore foot, the sole is gradually lowered which is controlled by eccentric contraction of the Dorsiflexors to prevent slapping of foot.

In hemiplegics, the anticipation of the weight bearing excites the extensor synergy which induces plantar flexion instead of dorsiflexion because dorsiflexion is not a part of the extensor synergy. The hemiplegic limb comes down on the entire sole and if the plantarflexion is marked then on the toes. Thus the heel strike is completely missing in hemiplegics. If there is pronounced inversion there is more weight borne by the lateral aspect of the foot and when it is moderate it gets corrected on weight bearing.

*Mid stance:*

In normal when the sole is firm on the ground the leg starts pivoting forwards above the ankle causing a relative dorsiflexion. This happens in response to the weight shift from hindfoot to the forefoot.The movement of relative dorsiflexion is controlled by eccentric work of the plantarflexors which gradually gets elongated.

In hemiplegics when the calf muscle is stretched by body weight, it stimulates the stretch reflex causing increase in spasticity of plantarflexors and it prevents any further stretch. Thus the forward shift is prevented in hemiplegics.

*Late Stance (Unloading to push off):*

In normals towards the end of the stance phase the heel raises from ground and the knee flexes. This is achieved by forceful concentric contraction of plantarflexors for initiating a forward swing.

In hemiplegics, the knee does not flex due to lack of dorsiflexion at the ankle, in the earlier phase and also due to influence of quadriceps spasticity. Sometimes even if it does flex it flexs gradually. Moreover the combination of plantarflexion with knee flexion neither belongs to flexion or extensor synergy, hence is very difficult for a hemiplegic to propel himself. Due to lack of push off in hemiplegics the force required to initiate a swing is very less that causes slowness in walking.

***Knee Joint:***

*Early and midstance:*

In normals as heel strikes the ground the knee is extended but as the body weight shifts on to the loading foot, there is 15 degree flexion at the knee under the influence of body weight and it is controlled by the eccentric contraction of the quadriceps to prevent buckling. Immediately afterwards due to concentric work of quadriceps there is extension at the knee.

In hemiplegic patient in flaccid stage there can be buckling due to weight transmission. In stage of spasticity, the initial flexion does not take place and the knee continues to remain in hyperextension throughout. Some patients do have a tendency to buckle initially but immediately rectify by going into hyperextension.

*Late stance:*

In normals knee flexes with plantarflexion

to give momentum to the swing phase.

In Hemiplegics the strong linkage between quadriceps and plantarflexors prevents the swing and the affected limb is moved forward by other compensatory mechanism.

***Hip Joint:***

*Early to midstance:*

In normals during weight bearing the abductors of the hip prevent the pelvis from sagging on the opposite side along with trunk flexors. The hip extensors work from heel strike to midtsnace. The short burst of activity reduces just before midstance.

In hemiplegics due to activation of extensor synergy, the adductors contract in place of abductors and hence there is tredelenberg's sign. Moreover when the adductor spasticity is very severe the affected leg is placed very close to normal limb or may even cross over interfering grossly with the forward propulsion.

*Swing Phase:*

In normals this begins in the late stance and is due to combined effort of muscles, gravitational force and momentum.EMG studies have shown that muscle activity level is minimal. The hip and knee flexors along with Dorsiflexors act to help in adequate ground clearance.

In hemiplegics the extensor synergy does not let go its grip and if it does then it is very gradually so that the body moves forward in slow manner. Hip and knee flexion is absent because of plantarflexion at the ankle and retracted pelvis. Thus, there is inadequate ground clearance. In the absence of forward rotation of the pelvis and hip flexion, the limb is brought forward by circumduction.

**Assessment of Stroke Patient**

***History Taking***

Date of onset, progression of the condition, about the medical management the patient underwent, check results of previous investigations for example MRI, CT and blood reports.

*Past Medical History*

- Check if patient has history of surgery
- Does he have dependency on any kind of technology like pacemaker, home oxygen etc.
- Risk factors like hypertension, DM and cholesterol to be checked.
- History of neurological disorders like TIA.

***Subjective***

The subjective and objective assessment together provides a detailed picture of how the present condition affects the patient. Proper subjective and objective evaluation helps to set realistic goals to guide treatment and maintain patient motivation.

Social History-

It is important to understand social situation of the patient as it helps to plan discharge and the treatment plan during and after the treatment.

***Things to checked are-***

Patients background

- Family support
- Accommodation (Gives the therapist an idea if any modification to be done inside house for better mobility and plan a suitable home program)

- Ask if the patient is linked with any social service organization

***Activity***

Mainly 3 areas to be checked i.e. Activities of daily routine, employment and other leisure activities like sports, art, reading etc. Understanding the involvement and difficulty faced by the patient to perform bed mobility like turn to one side, getting up to sit, sit to stand, difficulty in taking shower, difficulty in clothing and toileting helps the therapist to set goals for the patient in a step by step manner.

***Mobility***

- Can the patient walk without brace and with physical assistance?
- Can the patient walk independently with a knee brace and walker support?

***Transfers***

- Therapist has to check if the patient can transfer independently from wheelchair to couch. Note if the patient requires partial assistance, complete assistance or can perform it independently.

Other areas of involvement-

- **Vision**
- **Hearing**
- **Communication**
- **Cognition**
- **Swallowing**
- **Fatigue**
- **Pain**

***Objective assessment*** *Posture*

- In posture first thing to be noted is head, trunk and pelvic alignment and then check for neglect.

*Voluntary and involuntary movement*

*Range of Movement*

Measurement of movement around the joints of affected area is import and and is done with the help of a Goniometer. Range can be measured in 3 ways-

- Passive range of motion- If the joint is inactive due to poor functioning of the muscle, in this situation the range has to be checked passively by the therapist.
- Active assisted range of motion- Patient actively performs the activity and the therapist provides minimal assistance to complete the range.
- Active range of motion- Movements performed by the patient independently.

*Strength*

The maximum force a muscle can generate to perform a particular pattern can be called as normal muscle strength.

*Involuntary movements*

- Clonus
- Tremors
- Associated reactions

***Reflexes***

**Deep tendon reflexes should be checked of upper and lower extremities the classification of the reflexes are as given below. Brisk and Exaggerated reflexes suggest hypertonia.**

(-absent, +present but depressed, ++ normal, +++ brisk, ++++ exaggerated)

***Sensations-***

Sensations are affected due to the damage in the brain. Tingling and numbness can be felt in the affected part of the body. Light touch, pin prick/pain, pressure, two point discrimination, vibration and proprioception is to checked thoroughly as poor sensation can affect safety, activities of dailiy living and self care.

***Balance-***

- It is the ability to maintain the body in equilibrium.
- Balance is to be checked in sitting as well as standing. Grading given below can be used for assessing balance in these positions

Table 6: MRC grading for manual muscle testing

| Grade | Function |
|---|---|
| **0** | **No contraction** |
| **1** | **Feeble contraction can be felt** |
| **2** | **Full range of motion in gravity eliminated position** |
| **3** | **Full range of motion against gravity** |
| **4** | **Full range of motion against gravity and minimal ressistance** |
| **5** | **Full range of motion against gravity and maximum assistance** |

***Co-ordination***

Co-ordination is the ability to use different parts of the body together both smoothly and efficiently and involves the complex coordination of multiple joints and muscles. Equilibrium and non equilibrium test is used to challenge the patient and note the areas of difficulty.

***Tone***

Normal tone is high enough to hold the body in upright posture against gravity and should allow voluntary movements.

Tone can be measured using modified ashworth scale

Modified ashworth scale

**Voluntary control (Brunnstrom grading) -**

Voluntary control is an activity that is done by the will of the individual. After stroke patients develop synergy patterns/ abnormal pattern and brunnstrom grading helps to use these abnormal pattern to their advantage.

Given below are the 6 different stages of recovery as per brunnstrom approach that can be used to assess the voluntary control

1 – Flaccidity no voluntary movement

2 – Synergies or minimal voluntary movement

3 – Synergies performed voluntarily

4 – Some deviation from Synergy

5 – Independent or isolated movement

6 – Individual joint movement nearly normal with minimal spasticity

Table 7: Sitting balance assessment

| Static Sitting | | Dynamic sitting | |
|---|---|---|---|
| Normal | Able to maintain balance against maximal resistance | Normal | Able to sit unsupported & weight shift across midline maximally |
| Good | Able to maintain balance against moderate resistance | Good | Able to sit unsupported & weight shift across midline moderately |
| G-/F+ | Accepts minimal resistance | G-/F+ | Able to sit unsupported and weight shift across midline minimally |
| Fair | Able to sit unsupported without balance loss and without UE support | Fair | Minimal weight shifting ipsilateral/front, difficulty crossing midline |
| Poor+ | Able to maintain with minimal assistance from individual or chair | Fair- | Reach to ipsilateral side and unable to weight shift |
| Poor | Unable to maintain balance – requires mod/max support from individual or chair | Poor+ | Able to sit unsupported with min A and reach to ipsilateral side, unable to weight shift |
| | | Poor | Able to sit unsupported with Mod A and reach ipsilateral/front – can't cross midline |

***Table 8 : Standing balance assessment***

| Static Standing | | Dynamic Standing | |
|---|---|---|---|
| Normal | Able to maintain standing balance against maximal resistance | Normal | Stand independently unsupported, able to weight shift and cross midline maximally |
| Good | Able to maintain standing balance against moderate resistance | Good | Stand independently unsupported, able to weight shift and cross midline moderately |
| G-/F+ | Able to maintain standing balance against minimal resistance | G-/F+ | Stand independently unsupported, able to weight shift across midline minimally |
| Fair | Able to stand unsupported without UE support and without LOB for 1-2 min | Fair | Stand independently unsupported, weight shift, and reach ipsilaterally, LOB when crossing midline |
| Fair- | Requires Min A or UE support in order to stand without LOB | Poor+ | Able to stand with Min A and reach ipsilaterally, unable to weight shift |
| Poor+ | Requires Mod A and UE support to maintain standing without balance loss | Poor | Able to stand with Mod A and minimally reach ipsilaterally, unable to cross midline |
| Poor | Requires Max A and UE support to maintain standing balance without loss | | |

**Few assessments scales that are used as a standard tool are:**

- STREAM Scoring. The Barthel Index.
- Functional Independence Measure (FIM)
- NIH Stroke Scale Work sheet (www. strokecentre.org)
- DASH scale

**Table 9: Modified Ashworth Scale**

| Grade | Description |
|---|---|
| 0 | No increase in muscle tone |
| 1 | Slight increase in muscle tone, manifested by a catch and release or by minimal resistance at the end of the range of motion when the affected part(s) is moved in flexion or extension |
| 1+ | Slight increase in muscle tone, manifested by a catch, followed by minimal resistance throughout the remainder (less than half) of the ROM |
| 2 | More marked increase in muscle tone through most of the ROM, but affected part(s) easily moved |
| 3 | Considerable increase in muscle tone, passive movement difficult |
| 4 | Affected part(s) rigid in flexion or extension |

- The Modified Rankin Scale
- The Berg Balance Scale
- Disability of arm shoulder and hand questionnaire

**Rehabilitation after Stroke:**

Stoke rehabilitation has experienced tremendous growth in recent years. Much of the impact of stroke on functional independence is due to its effects on motor function. One of the main reasons of the impaired quality of life of these patients is not only related to the actual lesion but also to the result of immobilization and segregation from the community.

The type of rehabilitation and outcomes are determined by a variety of clinical and social factors, including length of time since injury, level of dependency, characteristics of the residual impairment, age of the patient, and resources available.

***Is there any scope of remedial training in stroke?***

Earlier the adult central nervous system was rigid and unalterable. Stroke rehabilitation was focused on compensatory approaches for motor impairments and most clinicians believed that therapeutic interventions had little impact on the process of neurological recovery. But research indicates that the adult central nervous system has great neuroplasticity and tremendous potential for reorganization, which has offered new hope to those treating patients with long-term disability and underlies the increasing interest in finding new and more effective ways to maximize this potential. In the recent years, it has become clear that a variety of interventions have the potential to favorably influence motor recovery.

The ICF model provides a conceptual basis and a universal common language for understanding and describing patients' health status, reaching beyond mortality, diseases, and rehabilitation. In the field of neuro-rehabilitation, it facilitates

multidisciplinary team communication, to structure the rehabilitation process, for goal setting and assessment, for documentation, and for reporting.

ICF model defines various problems at four levels:

a) Pathophysiology of the HEALTH CONDITION,

b) Impairments at the BODY FUNCTIONS / STRUCTURE level,

c) Disability at the ACTIVITY level, and

d) Handicaps at the PARTICIPATION level. Based on the first three levels, Levin et al defined recovery and compensation to explain the differences and allow better understanding about motor recovery after stroke

***The mechanism of recovery:***

Spontaneous recovery is the norm after stroke. This recovery is typically most rapid & early after a stroke and generally proceeds at a decelerating pace. In case of mild deficits due to small infarcts and transient ischemic attacks, recovery of near normal motor function may result. The mechanism of neurological recovery after stroke involves more than one process.

Recovery of the ischemic penumbra (brain tissue that has been affected by ischemia but not infarcted) and resolution of cerebral edema early after stroke may account for much of the recovery that occurs within the first few days. The role of diaschisis, a loss of brain functions due to reduced neural input from remote infarcted tissue, may contribute to deficits that resolve during the early recovery period.

Stroke recovery may continue for months after a stroke. The latter portions of neurological improvement appear to be substantially due to cortical plasticity. The plasticity consists of various mechanisms like denervation super sensitivity, unmasking of silent synapses, regenerative and reactive synaptogenesis.

Basically, it is the rewiring of synaptic connections within the surviving brain tissue to restore some degree of motor functioning. Cerebral plasticity has been clearly documented in animal model of cortical stroke and appears active in humans as well.

Animal studies of cerebral function have found changes in cortical function that result from exercise training for motor tasks in both the intact and the damaged brain and in the absence of exercise of the paretic limb, the cortical plasticity provides important evidence of the impact of exercise training on the brain. Thus, cortical maps are dynamic However one should not forget to consider the various factors such as the patient's age, severity of the lesion, effects of drugs post injury that will have an impact on the outcome.

***Mechanisms that induce recovery (plasticity i.e., at neuronal level) after brain damage:***

Till date researchers have found following 10 principles, relevant to rehabilitation outcomes which might augment brain plasticity leading to neuronal recovery. Patients need to be explained, in details all of them to gain maximum benefit.

*Principles*

1. Use It or Lose It : Failure to drive specific brain functions with volitional movements can lead to functional degradation.

2. Use It and Improve It : Training that drives a specific brain function can lead to an enhancement of that function.

3. Specificity; The nature of the training experience dictates the

nature of the plasticity.

4. Repetition Matters: Induction of plasticity requires enough repetition.

5. Intensity Matters: Induction of plasticity requires enough training intensity.

6. Time Matters: Different forms of plasticity occur at different times during training. Earlier the rehabilitation intervention better the prognosis.

7. Salience Matters: The training experience must be sufficiently salient to induce plasticity.

8. Age Matters: Training - induced plasticity occurs more readily in younger brains.

9. Transference: Plasticity in response to one training experience can enhance the acquisition of similar behaviors.

10. Interference: Plasticity in response to one experience can interfere with the acquisition of other behaviors.

It is becoming clear that the amount and type of physical and mental activity as well as attitude and motivation of the patient has an impact on reorganization of the brain. Hence, application of these new approaches with better understanding of recovery and compensation and mechanism for plasticity will help us in gaining better outcomes after stroke rehabilitation.

### *The role played by physiotherapists in stroke rehabilitation*

Physical therapists specialize in treating disabilities related to motor and sensory impairments. Rehabilitation helps stroke survivors relearn skills that are lost when part of the brain is damaged. For example, these skills can include coordinating leg movements in order to walk or carrying out the steps involved in any complex activity.

Physical therapists help stroke survivors regain the use of stroke-impaired limbs and establish exercise programs to help people retain their newly learned skills. Disabled people tend to avoid using impaired limbs, a behavior called learned non-use. However, the repetitive use of impaired limbs encourages brain plasticity and helps reduce disabilities. Rehabilitation teaches survivors, new ways of performing tasks to circumvent for any residual disabilities.

There is a strong consensus among rehabilitation experts that the most important element in any rehabilitation program is carefully directed, well- focused, repetitive practice - the same kind of practice used by all people when they learn a new skill, eg: playing the guitar or learning new game.

For some stroke survivors, rehabilitation will be an ongoing process to maintain and refine skills and could involve working with specialists for months or years after the stroke. Over time rehabilitation can be quite remarkable. It does take a team of professionals and the family to accomplish this.

Physiotherapists take a leading role in developing appropriate policies and strategies with other exercise professionals and services to address the transition from rehabilitation to an active lifestyle following stroke.

During this recovery phase, there are many caveats which physical therapists have to address to enhance the progression of recovery. Few of these issues are as follows:

- Paralysis or muscle weakness
- Impaired coordination

- Apraxia ( patients loose their ability to plan the steps involved in a complex task and to carry the steps out in the proper sequence, having problems following a set of instructions.)
- Impaired muscle tone
- Loss of feeling
- Speech and language impairments
- Memory and reasoning problems
- Swallowing difficulties
- Psychological trauma (fear, anxiety, frustration, anger, sadness, and a sense of grief, clinical depression
- Problems with vision and visual perception Goals of Treatment in Stroke patient:
- To normalize muscle tone
- To restore muscle function
- To control compensation strategies
- To maintain muscle length
- To re-educate balance
- To retrain walking and restore mobility
- To maximize functional ability while allowing on-going neuromuscular recovery

To deal with these issues, physical therapists adopt various approaches during the rehabilitation process.

***Approaches used in Clinical Setting – Early Rehabilitation***

Rehabilitation therapy begins in the acute-care hospital after the patient's medical condition has been stabilized, often within 24-48 hours after the stroke.

Goals of treatment in acute stage are :

a) Prevent ignorance or unawareness of hemiplegic side.

b) Decrease the tendency to develop synergy in the chronic stage.

c) Prevention of any joint restriction or stiffness.

d) Prevention of complications due to immobilization like chest complications, deconditioning of bone muscles, etc.

e) Early weight bearing.

f) Psychological counselling.

g) Education to the family.

The first steps involve promoting independent movement because many patients are paralyzed or seriously weakened. Patients are promoted to change positions frequently while lying in bed and to engage in passive or active range-of-motion exercises to strengthen their affected limbs. Exercises are generally begun with mat activities like rolling, sitting up from lying, in sitting position activities like side shifting are encouraged. Patients progress from sitting up and transferring between the bed and a chair to standing, bearing their own weight, and walking, with or without assistance.

Weight bearing exercises are necessary to promote development of tone in muscles and to maintain the absorption of calcium in the bones. So, activities like bridging, supine to elbows, sitting with weight bearing on affected arms and standing should be given as soon as possible after stroke. Oromotor exercises are also started to inhibit drooling and initiate adequate swallowing without chocking.

Rehabilitation nurses and therapists help patients perform progressively more complex and demanding tasks, such as bathing, dressing, and using a toilet, and they encourage patients to begin using

their stroke-impaired limbs while engaging in those tasks. Beginning to reacquire the ability to carry out these basic activities of daily living represents the first stage in a stroke survivor's return to functional independence.

Exercises on vestibular ball are also started to improve balance and also normalize tone of the trunk. Vestibular ball may be used to exercise the upper extremity mainly to achieve control at the proximal joint and facilitate extension of fingers through stimulation of the proprioreceptors at all the joints of upper limbs. Once the patient is comfortable in doing these exercises, then standing and gait training are initiated emphasizing on symmetrical weight bearing on both lower limbs. Traditional therapeutic exercise program also consists of strengthening, mobilization, compensatory techniques, endurance training (e.g., aerobics). Traditional approaches for improving motor control and coordination: emphasize need of repetition of specific movements for learning, the importance of sensation to the control of movement, and the need to develop basic movements and postures.

***Approaches used in Clinical Setting-Late Rehabilitation***

*Bobath Approach (Neurodevelopmental Therapy)*

The Bobath approach is widely utilized in rehabilitation following stroke and other neurological conditions. It is a problem-solving concept that allows for a variety of strategies flexible enough to be adapted to the strengths and impairments of the individual client rather than a prescribed treatment of exercise.

Traditional Bobath approach was based on the belief that voluntary movements are built on reflexive movement and that treatment must follow the normal developmental sequence. Over the years, the Bobath approach has been modified with many changes and till today alterations continue.

Current Bobath concept has accepted that neural control is not a simple hierarchical function, but it is more complex where multiple body systems participate in executing movement that is organized by the specific task and constrained by physical laws and the environment.

Limitation:

There is currently no evidence that Bobath interventions are more or less effective than other therapy approaches (kollen, et al., 2009, Luke, et al., 2004; Paci, 2003).

Hence, NDT approach requires evidence for its effectiveness over other treatment approaches.

Proprioceptive (or peripheral) Neuromuscular Facilitation (PNF)

PNF approach uses spiral and diagonal components of movements rather than the traditional movements in cardinal planes of motion with the goal of facilitating movement patterns that will have more functional relevance than the traditional technique of strengthening individual group muscles.

It uses resistance with the goal of facilitating "irradiation" of muscles to other parts of the body associated with the primary movement.

*Constraint Induced Movement Therapy (CIMT)*

CIMT is a behavioral approach consisting of different compo-

nents derived from neuro rehabilitation from basic neuroscience. The treatment for humans after neurological injury has three components:

a) Repetitive, task oriented training of the impaired extremity or function following shaping principles for several hours a day for 10 or 15 consecutive weekdays (depending on the severity of the initial deficit);

b) Constraining patients to use the impaired extremity or function during walking hours over the course of treatment, sometimes by restraining the unimpaired extremity; and

c) Applying a package of behavioral methods designed to transfer gains made in the clinical setting to the real-world.

*Brunnstorm Approach (Movement Therapy)*

Brunnstrom approach is based on the concept that damaged CNS regressed to phylogenetically older patterns of movements (limb synergies and primitive reflexes); thus, synergies, primitive reflexes, and other abnormal movements are considered normal processes of recovery before normal patterns of movements are attained. Hence, based on this principle, Brunnstorm approach uses primitive synergistic patterns in training to improve motor control through central facilitation. Treatment includes facilitation of specific synergies through cutaneous / Proprioceptive stimuli.

*Motor Relearning Program/Carr and Shepard Approach*

This approach is based on cognitive motor relearning theory and influenced by Bobath's approach. Goal is for the patient to relearn how to move functionally and how to do problem solving while attempting new tasks.

In addition, this approach teaches general strategies for solving motor problems, instead of emphasizing repetitive performance of a specific movement for improving skills.

***Motor Learning*** is defined as a set of processes in which the patient learns a skilled activity by repeated practice and experience.

5 key factors in motor learning are

*1. Repetition* - With practice an individual is able to develop appropriate sequencing of movement component with better timing and concentration.

*2. Retention* - Ability to perform an activity over time and with no practice. Initially can be difficult but the individual gets the hold of it after few trials.

*3. Generalization* - Ability to use a learned skill for learning other similar task. For example, if the patient learns to transfer from wheelchair to bed, he should be able to implement the same technique to transfer from wheelchair to commode or car.

*4. Resistance to contextual change* - Ability to perform a motor task in altered environment.

*5. Motor capacity* - As the impairment in learning is common in CNS dysfunction, rate and degree of learning may be different in individuals.

Stages of motor learning

Fitts and posner for better understanding explained motor learning in 3 stages

1. Cognitive stage-

- To develop skill termed as 'cognitive mapping' ie decision making phase of what to do.
- By repetition retain stratergies that are successful and discard which are not.
- Initial stage learner relies on vision

2. Associative stage-

- In this stage the learner focuses on 'how to do' the movement
- Proprioceptive cues required more while dependence on visual cue reduces.
- Movement develops into coordinated pattern as spatial and temporal aspects become organised.

3. Autonomous stage-

- In this stage after repeted practice now the activity is performed automatically.
- Minimal level of cognitive monitoring required.
- Learner is able to perform much more complex task with better control and coordination.
- Can perform tasks in predictable and unpredictable environment.

*Sensorimotor Approach/Rood Approach (Noll, Bender, and nelson, 1996)*

This approach is based on modification of muscle tone and voluntary motor activity using cutaneous sensory motor stimulation.

To facilitate muscle tone following cutaneous stimulation techniques are used: quick stretch, brisk icing, fast brushing, tendon tapping, vibration, and joint compression to promote contraction of proximal muscles. Similarly, to inhibit muscle tones, following cutaneous stimulation approaches are used: slow sustained stretch, prolonged icing, etc.

All of these approaches can be used in combination or independently as deemed necessary by the therapist. These approaches should be used for functional training. Focus should be on improving function, independence and participation of the patient. Functional training should start from bed mobilities followed sitting, activities in sitting, transfers, standing, ambulation, vocational movements and recreational movements.

**Bed mobility training:**



*Figure 1: Side to side movement by clasping both hands to facilitate turning and rolling.*



*Figure 2: Pushing up on the elbow of unaffected side to come to sitting. When there is motor weakness due to stroke, the contralateral side can be trained to achieve functional independence.*

**Exercises to improve motor control**
***Strengthening***



*Figure 3: Suspension exercises: these exercises are performed when the patients are unable to perform movements against gravity but are able to perform the movements in gravity eliminated plane. It can be used only for single joint movements giving an opportunity to the therapist to prevent involuntary for synergistic movements in other joints.*



*Figure 4 – When there is sufficient strength to perform movements against gravity. Active movemetns are encouraged. As strength improves resisted activities can be performed.*

***Proprioceptive neuro-muscular facilitation***



*Figure 5: Sit to stand training using PNF technique.*

**Weight bearing exercises**

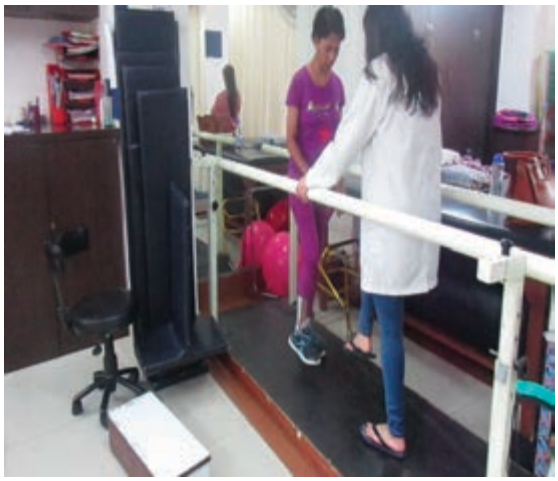


*Figure 5: Stepping exercise in parallel bar to improve weight bearing in the affected leg.*



*Figure 6: Kneel walking exercise- Improve hip knee flexion pattern while walking.*



*Figure 7: Quadruped position to facilitate proprioception in upper and lower extremities.*



*Figure 8: Pelvic bridging (unilateral and bilateral) to aid trunk strengthening and weight bearing.*

## Gait Training



*Figure 9: Gait training with cane and physical assistance.*




**Figure 10: Training for ascending and descending slopes**




**Figure 11: Training for ascending and descending stairs.**

**Newer approaches in late rehabilitation**

***Treadmill mediated gait training***

*Gait restoration is a major goal in post-stroke neurological rehabilitation.* For this reason, the recovery of independent walking is important in rehabilitation studies. Recently, gait training on a treadmill with body weight support (BWS) has received special attention. It consist of a suspension system to which a patient is connected so that weight shifting, balance, and stepping can be controlled; walking is facilitated by a treadmill. Increasing evidence has suggested that treadmill training in older subjects with hemiparesis improves locomotor capabilities during over-ground walking and motor relearning, because it provides task-oriented practice of walking and active repetitive movement training. It has been suggested that through training, functional movements of locomotors patterns, sensory inputs, and therefore central neuronal circuits, become activated. Hence, even with several studies having shown the feasibility of supported treadmill ambulation training in patients with stroke, whether it is superior to other therapies is still under dispute.

***Functional electrical stimulation (FES)***

People with hemiparesis often display abnormal gait patterns, such as equinovarus (excessive plantar flexion and inversion) or foot drop (excessive plantar flexion), in which selective Liberson et al, uses electrical signals to activate peripheral nerves and control functional movements. This technique makes use of afferent feedback during contraction, a process that, with a patient's help, may minimize motor relearning during active repetitive movement training.

In routine clinical practice Functional electrical stimulation time (in minutes) can be adjusted according to verbal feedback from the patients during the 20 to 45 minute stimulation period. The patient should be interested to say when they felt fatigue related to dorsiflexion and eversion movements of the stimulated leg. In that situation, FES should be discontinued for 5 minutes and then activated again. As volitional control improves, the FES amplitude can be reduced.

Two main advantages of using FES combined with treadmill training.

- The first advantage was that the patients often report a preference for walking on the treadmill with BWS combined with FES. They report that gait training was more comfortable because it was easier to place their foot during early stance.
- The other advantage was that training with FES decreased the participation of the physical therapists. Manual assistance can be provided

to help the subjects optimize gait quality during training.

***Task-oriented training:***

No conclusive definition of a task-oriented approach exists in the literature. In the task-oriented approach, movement emerges as an interaction between many systems in the brain and is organized around a goal and constrained by the environment. (Shumway Cook & Woollacott 2001). Task-oriented training includes a wide range of interventions such as treadmill training, walking, training on the ground, bicycling programmes, endurance training and circuit training, sit-to-stand exercises, and reaching tasks for improving balance.

Task oriented training needs to be repetitive and meaningful for the individual. Many interventions in task-oriented training that have proved to be effective are usable in daily nursing practice, such as walking on the ground, moving from sitting to standing from different chairs, and sitting and reaching.

Hence, such training is task and patient focused and not therapist focused. Active use of task-oriented training in the daily nursing care of stroke survivors, will lead to improvements in functional outcomes and overall health-related quality of life.

***Robotic-Assisted Therapy (Krebs et al., 1998)***

Robotic-Assisted Therapy for stroke rehabilitation started only since 1990s. in general, robotics interfaces with computer software and hardware for improving upper extremity function. Most robots that have been developed for rehabilitation allow several modes of operation including acting as a low friction passive support to the upper extremity, providing active range of motion and / or soft guidance in a movement pattern when the individual is unable to initiate or complete a movement, and offering some resistance to the movement.

*Five types of robots have been tested:*

1) Assisted Rehabilitation and Measurement (ARM) Guide- made for straight line trajectory reaching (Reinkensmeyer,et al.2000)

2) Mirror Image Movement Enabler (MIME) - provides unilateral or bilateral shoulder / elbow movement (lure, et al., 2002; Kahn, et al., 2006)

3) Bi-Manu-Track- facilitates bilateral passive and active movement of the forearm (Prange et al., 2006)

4) NeReBot- enable movement of the shoulder, elbow, and wrist (Masiero, et al., 2007)

5) MIT-Manus/inMotion2- allows the person to person movement without interference from the robot (although the robot can help perform the motion when the person is unable to initiate or complete the motion)

Robotic-assisted therapy has the potential to facilitate improvements in motor control post stroke, but it is not clear whether this therapy improves outcomes to a greater extent than conventional therapy.

***Bilateral Movement Training (Stewart et al., 2006)***

A primary reason to perform bilateral arm training is that much of what we do everyday involves the use of both arms and therefore, bilateral re-training is necessary. For example, daily living activities such as bathing, dressing, feeding, toileting, as well as, many other mobility functions such as carrying ob-

jects, getting up from bed or chairs, and in driving requires use of both arms and hand. Instrumental activities such as keyboarding, shopping and cooking also rely heavily on bilateral arm use. Hence, bilateral training has potential to improve arm function post-stroke.

*Mechanism for Bilateral Movement Training:*

1) Bilateral coordinated movement, the upper extremities couple and act nearly as a single unit, reflecting inter-hemispheric coupling at the supplementary motor area (SMA), lateral premotor cortex, the premotor and sensorimotor areas that may facilitate movement in the affected extremity.

2) There is a reduction in intra-cortical inhibition with bilateral movement that may facilitate reorganization in the damaged hemisphere.

*Limitation:*

Studies of bilateral training have shown that people improve their motor skills with bilateral training after stroke, but the results are completely mixed on whether such training produces greater motor gains than unilateral training.

To design rehabilitation protocols for functional gains with bilateral movements is a challenge.

***Mental Imagery Therapy (Page et al., 2006: 2007)***

Mental practice, using first-person visual and kinesthetic imagery to practice performance of tasks without activating the muscles involved in physically performing those activities, activates many of the same neural areas as does physical practice. In stroke, mental practice has been paired with either practice of daily common tasks or nonfunctional motor tasks and has been performed from 3-5times per week. Studies have reported a positive effect of adding mental practice to therapy.

Thus, mental practice after stroke appears a promising adjuvant to motor rehabilitation. Although unexplored, the ability of individuals with stroke to produce quality detailed images may influence response to this therapy. How well motor imagery can be accomplished may depend on stroke location in addition to pre-stroke imagery ability.

***Virtual reality therapy (Burdea, et al., 1994)***

Virtual reality technology provides the capacity to create an environment in which the intensity of feedback and training can be systematically manipulated and enhanced in order to create the most appropriate, individualized motor learning paradigm. The potential benefits of training in virtual reality environment would be the ability to increase the duration, frequency, and intensity of therapy that could be provided to patients by using semi-automated programs. Virtual reality rehabilitation exercises can be made to be engaging, such that the patient feels immersed in the stimulated world, which is extremely important in terms of the patient motivation.

*Limitation:*

Subjects trained on a motor task in a virtual environment demonstrated the ability to improve performance on the task in that environment, but the learning did not always transfer to the real-world task.

Hence, these protocols should be explored more in order to ascertain the use of virtual reality training as an enhancement to traditional stroke therapy.

**Management of Complications:**

1. ***Complex regional pain syndrome-*** many hemiplegic patients develop pain in the upper extremity. Pitting edema, cyanosis, and limitation of extension in the wrist and finger joints, associated with pain, are common and can be attributed to malpositioning and lack of therapy

   *Treatment :*

   Paraffin bath may be utilized prior to the passive assistive exercises, provided the paraffin glove is molded with the fingers in maximum flexion. The use of a platform or cockup splint, with the fingers in extension is advised.

2. ***Subluxation of Shoulder:*** is very common,and sling suspension along with weight bearing exercises are recommended.

   Heterotopic calcification - extra articular calcification has been described in other neurologic disorders and it may occur in hemiplegic patients. It is seldom encountered in the patient who has been quite active and conscious during the early petiod following the stroke. Repeated insignificant trauma may play a role in the development of this, progressive refractory complication.it may be associated with parietal lobe dysfunction causing the patient to neglect the limb, predisposing it to repeated injury.

   The extra-articular calcification occurs about the lesser trochanter of the femur producing a severe adduction flexion contracture of the hip similarly, it can produce a complete extra-articular bony ankylosis of the shoulder joint.The extensor surface of the elbow just above the olecranon process may also be the site of involvement. The elbow is held in an atypical attitude of relative extension rather than flexion as commonly seen in the hemiplegic patient. Initially the elbow is quite painful, warm and slightly swollen, without any joint effusion. A small amorphous calcific deposit at the insertion of the triceps tendon, seen in the lateral X-ray film,may be the initianl finding in other patients this complication may not recognized until the acute phase has subsided the calcification may progress to complete ossification

   Treatment of this complication is very unsatisfactory, particularly when it is advisable to immobilize the elbow in flexion and avoid all motion until the swelling and pain have subsided has been recommended in traumatic conditions but has not been described in hemiplegic patients.

3. ***Frozen Shoulder / Adhesive Capsulitis:*** The painful shoulder is probably the most frequent and most disturbing complication encountered in the stroke patient. It can be quite disabling since it intereferes with active function, including the performance of simple dressing activities, and may even secondarily immobilize the hand.

The cause of the painful shoulder in the hemiplegic patient has been the subject of consider able discussion basically, several factors may be involved:

1) preexisting degenerative changes in the rotator cuff;
2) repeated small trauma to the periarticular structures, incurred as a result of traction on the shoulder when the patient lies unconscious on the hemiplegic arm or when he tries to move in bed with the arm flaccid at his side;
3) excessive traction on the shoulder capsule, with marked inferior displacement of the humeral herd in a flaccid limb;
4) vigorous stretching of the “tight” spastic shoulder during physical

therapy, producing further trauma to the cuff and long head of the biceps tendon;

5) unrecognized trauma such as an impacted fracture of the surgical neck of the humerus, incurred as a result of falling during the onset of the stroke;and

6) contractures that occur in the untreated patient or develop as a result of peripheral nerve lesions and heterotopic calcification.

The treatment of the painful shoulder should be predicated on an accurate diagnosis rather than consist of routine procedures and exercises. Although the use of a sling may be of considerable value in supporting the flaccid shoulder, it may only aggravate the adduction internal rotation contracture in a painful spastic shoulder. Mobilisation of the head of humerus and manual therapy gives great results in breaking adhesions between the capsule.

Local infiltration of the tender long head of the biceps tendon with procaine and corticosterois may afford considerable relief when the diagnosis is peritendinitis.in some instances pendular exercises may produce sufficient relaxation to permit effective mobilization. Oral corticosteroids have been used extensively Physical therapy procedures are prescribed to relieve the pain and increase the range of motion.the application of hot packs or cold packs over the deltoid region has become more or less a routine procedure. Frequently it is more desirable to apply the packs over the spastic or contracted pectoral muscles while the shoulder is abducted and in some external rotation rather than adducted and internally rotated. The physical therapist can then intiate the shoulder exercises from a more advantageous position diathermy should be used with considerable caution, particularly in patients having a sensory deficit. Ultrasound therapy has been employed with varying results but may be indicated in treating a persistent localized tendinitis.

The application of a modality to the painful hemiplegic shoulder should always be followed by range-of-motion exercises. The patient must be in a supine position so that the shoulder can be adequately stabilized and the effects of gravity eliminared vigorous stretching should not be devoted to this phase of the treatment to permit the the pectoral and other adductor muscles to relax. Stretching will only elicit a stretch reflex and increase the spasticity in these muscles.

Pulley exercises should not be prescribed unless there is at least 60 to 70 degrees of passive abduction present; otherwise they will only increase the scapulothoracic substitution. The patient must be properly positioned so that the overhead pully is slightly behind him to effectively abduct and externally rotate the shoulder during this exercise.

***4. Vascular complications.*** Thrombophlebitis in the hemiplegic lower extremity occurs not infrequently during the early convalescent period and may be overlooked. this could explain the persistent edema in the lower leg of some patients after they are started on ambulation activities. Simple elastic compression bandage or stocking applied before the patient gets out of bed is quite adequate. Ample provision must be made for the swelling about the ankle when fitting the patient for shoes or a short leg brace. The T strap on the brace must be broader than usual and well padded to avoid any constriction about the ankle. The lower leg should be elevated when the patient is sitting in a wheelchair.

An incipient occlusion of the femoral artery may simulate a thrombophlebitis. Evaluation may be difficult, particularly when the patient is aphasic. Vascular

changes may have been present prior to the onset of the hemiplegia. Pain in the lower limb is persistent, and there may be evidence of an ischemic neuropathy particularly involving the deep branch of the peroneal nerve, which is most sensitive to arterial insufficiency. The foot drops, if present initially, will become more pronounced, the hyperactive tendon reflexs may be lost, and a segmental sensry deficit involving especially the first web space may signal the onset of a major arterial occlusion.prompt surgical intervention is indicated at this stage rather than waiting for the appearance of the cyanosis of the foot and demarcation of the ischemic skin.

***5. Seizure:*** epileptic seizures may occur as a late complication of a cerebral vascular accident.the incidence may be as high as 10%to 15% among those patients who survive beyond the first year. Since the focal signs are localized in the hemiplegic extremities and in the speech mechanism, they often lead to the diagnosis of another stroke. However, these patients recover quite rapidly within the first 24 hours and resume their previous level of activity

***6. Decubiti / Pressure Sore :*** The older hemiplegic patient with peripheral vascular disease must be carefully observed during the early phase of the illness to prevent pressure sores, particularly over the posterior aspect of both heels. Early mobilization helps in preventing it, by improving blood supply.

***7. Trauma.;*** the hemiplegic patient is accident prone, especially when ataxia is the predominant peripheral defect. Fracture of the femoral neck is a frequent mishap, particularly in those ataxic patients who walk with a spastic adducted gait and invariably fall on the hemiplegic side. This injury may be associated with an unrecognized impacted fracture of the surgical neck of the humerus.

Careful consideration should be given to the management of the hip fracture. Replacement of the femoral head with prosthesis, followed by immediate ambulation with full weight bearing, has become the accepted procedure in fractures of the femoral neck in the elderly hemiplegic patient. More recently, consideration is being given to simply pinning the fracture after closed reduction followed by immediate progressive weight bearing.

## Occupational Therapy management of stroke

Stroke survivors face barriers to engaging in productive activity due to imposed long-term disability. To facilitate the client's performance of needed or meaningful occupations within realistic contexts to promote independence, Occupational Therapy practitioners use their expertise in activity analysis and adaptive methods

**Post Stroke Depression -As a hurdle to Rehabilitation**

The most common neuro-psychiatric consequences of stroke are post-stroke Depression. It creates a great impact on rehabilitation. Studies reveal it has an impact on quality of life and can lead to increased mortality, cognitive impairment, increased risk of falls and most importantly has worst outcome.

***Occupational Therapist's strategies:***

- Observe the patient and rule out whether the person requires medical intervention or counselling based on your experience and clinical reasoning
- Insight building- letting the patient know his/her abilities despite of the affectations
- Client centered approach: Therapist can make use of COPM -assessment and treatment planning tool, which will give importance to patient's priorities and help patient to gain self -control to decide for himself/herself.

**Treatment intervention:**

***Improving voluntary control through functional activities:***

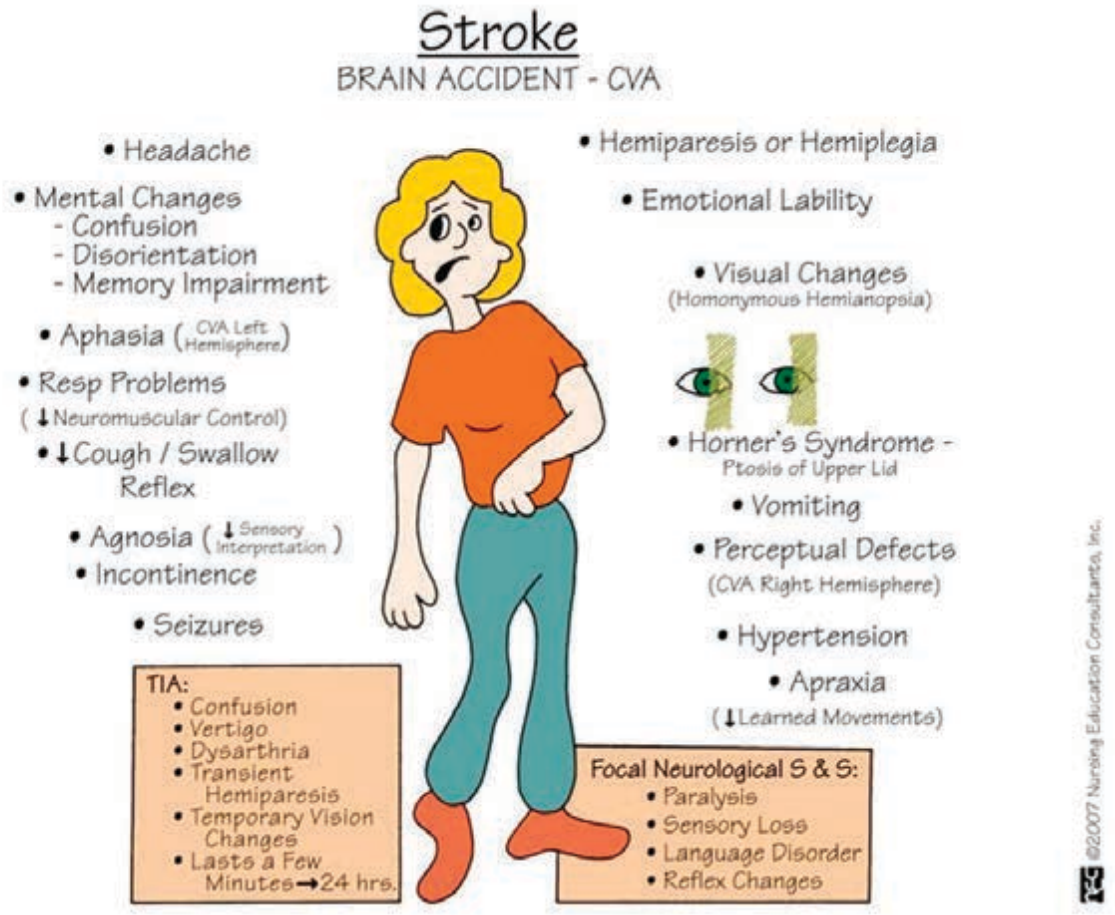



*Figure 12 . depicts the symptoms of stroke and the 1st thing a person will notice is the characteristic posture of a hemiplegic person. The above posture states the Brunnstorm's stage 3- where in the spasticity is at its peak with synergies present in upper extremity (flexor synergy) and lower extremity (extensor synergy) The Brunnstorm's approach thus defines a functional approach to recovery post stoke*

As an occupational therapist we can encourage the functional recovery using Brunnstorm's approach as the evidence suggest its efficacy over other contemporary approaches to rehabilitation. The basic principle to the Brunnstorm's approach is to make the synergy and later break the synergy through incorporating functional movements.

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Figure 13: Depicts the functional movements a patient can incorporate in household activities to break the synergy pattern and at the same time also engage into a functional activity

***ADL training and adaptations:***

*Eating:*

Eating is the most important ADL because it's the need for survival. The patients with their dominant side affected has mental block that they won't be able to eat on their own and gets dependent on the task of feeding. Therapist's thus should make aware of compensating the task of feeding self with the non- dominant hand and stabilizing it with the dominant (affected) hand ( Fig 14 )



Fig 14: Compensating for the lost function with contralateral side

Bathing:

The therapist should encourage the use of un affected hand to apply soap and water(Fig.15) The difficulty to wash back can be sorted by using a long handle scrubber. The person might need help of caregiver to apply soap to the unaffected hand which would be justi-

fied if the affected hand is under Stage 1-stage 3 of voluntary control (according to Brunnstorm's stages of recovery). If movements are possible away from synergy then bathing with the affected hand is encouraged as far as possible. For safe transfers grab bars can be advised. A shower chair can be advised. Non-skid mats are advisable to decrease the risk of falling.



Figure 15: Adapted technique for bathing using contralateral side

*Dressing Technique:*

Fig.15 and Fig. 16 describes how a stroke patient can dress himself/herself independently.

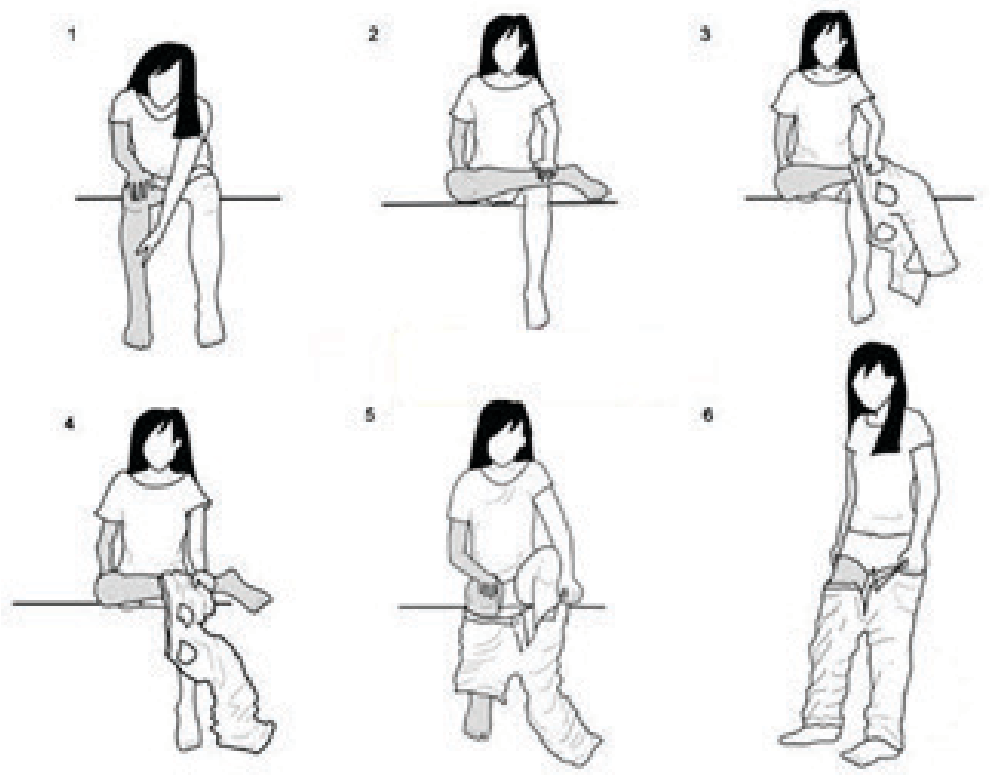



Figure 15 : Adapted dressing technique

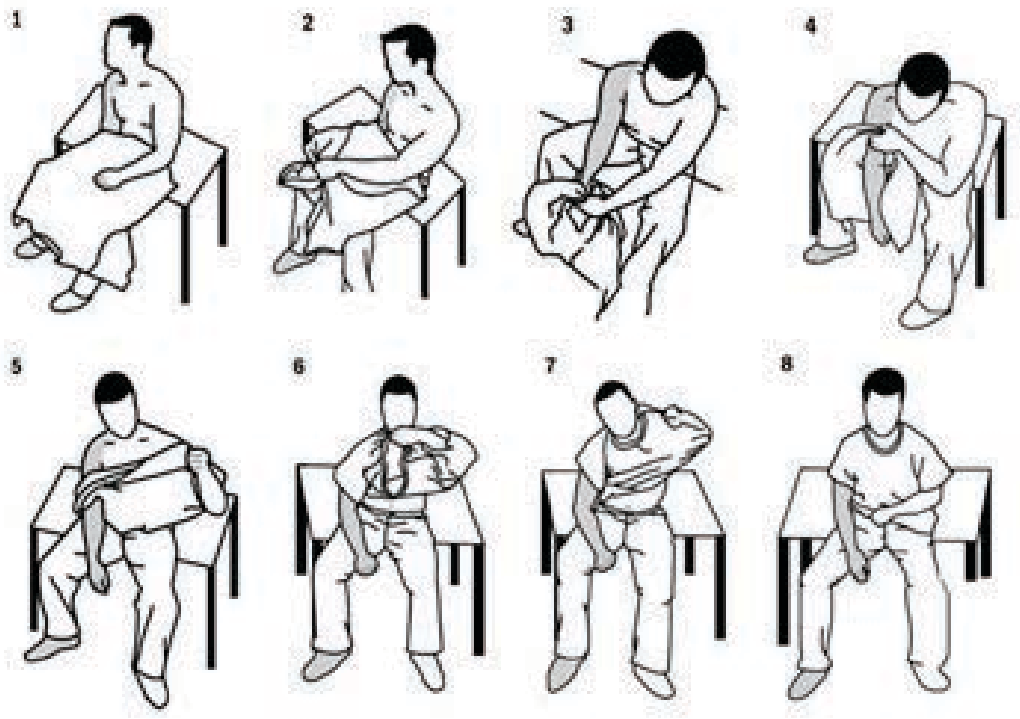



Figure 16: Adapted dressing technique

*Toileting:*

In an Indian style toilet, grab bars are advisable on the unaffected side for support while squatting up-squatting. A cut-out stool or chair becomes a low-cost alternative to western style. In case of a jet spray a short pipe can be attached to the tap inside toilet for maintaining perineal hygiene. The person can be trained to perform perineal hygiene either unilaterally or using the affected hand to hold the pipe and unaffected hand to clean based on stage of voluntary control of arm and hand.

*Grooming:*

A universal cuff can be used to perform multipurpose activities like brushing teeth, combing hair, shaving etc. ( Fig 17) Slippers or Velcro shoes can be advised to avoid laced shoes which needs bilateral co-ordination.



Figure 17: Adapted comb that can be used for grooming activities

Instrumental Activities of Daily Living (IADLs):

Let the client participate in IADLs as per their priorities, the above selfcare activities thus being the most basic and important ones to address. In the following figures, the lady's priority was to make tea for self and the family, hence the therapist trained her to use alternative techniques and encouraged the client to use her ability to her best. Fig 17 to Fig 21



Figure 17



Figure 18



Figure 21



Figure 19



Figure 20

***Addressing contextual components: home and family***

- To assess home environment a home visit is mandatory and necessary home modifications can be advised after discussing with patient and caregiver.
- It is necessary to interview regarding the availability of caregivers (primary, secondary and tertiary) and hence accordingly the caregiver should be taught the home exercise regime and make aware of the patient's abilities and the need and amount of assistance to be provided to patient. In few instances, a role reversal between patient and spouse needs to be considered and counselled accordingly.
- An occupational therapist will be able to provide with aids and adaptive devices in liaison with social services.
- A stroke club/society/day care can be advised for social-reintegration.

In a stroke patient it often happens that the physical difficulties overshadow the cognitive and perceptual dysfunctions which are the effects of pathological changes immediately after cerebrovascular accident. Cognitive -Perceptual dysfunctions are often referred as hidden disability. Therapist's should address these dysfunctions as well while treating a stroke patient. Hence, in

this chapter, cognitive and perceptual dysfunctions with their respective treatment strategies have been discussed in detail

***Cognitive Dysfunctions and Intervention***

Cognition refers to the ability of the brain to Process, store, retrieve and manipulate information. It refers to the acquisition and use of knowledge involving a number of mental components

Let us see the components of cognition: please refer the flowchart below

*Chart 1: Flow chart showing components of cognition*

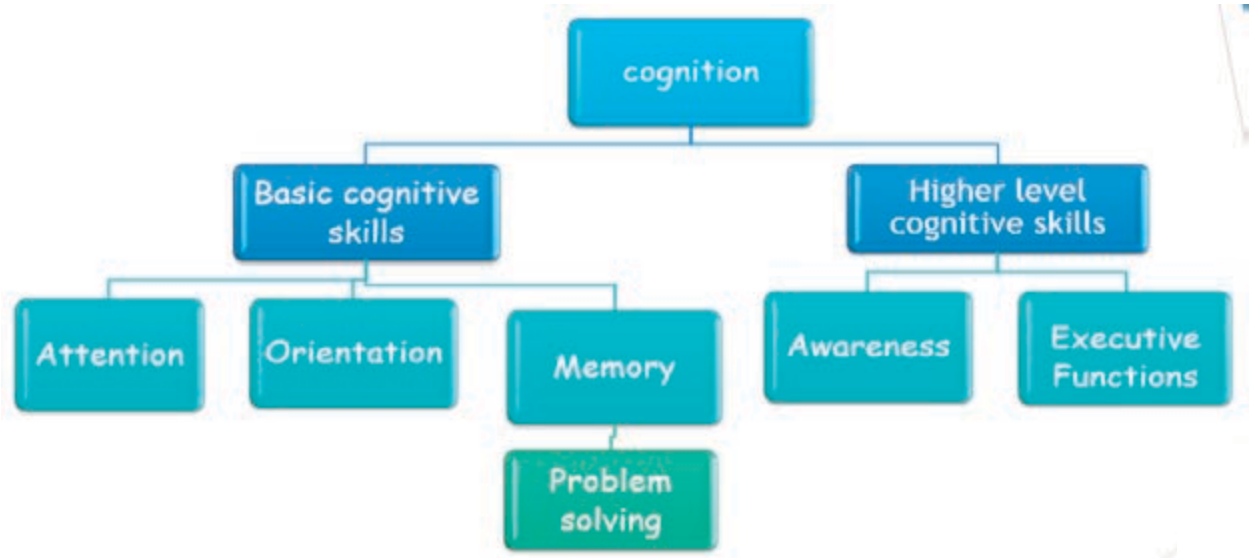



Fig. 22 Depicts The function of brain -lobe wise. The below mentioned figure will help us understand the dysfunction in cognition acoording to the patahology and the area or lobe involved.

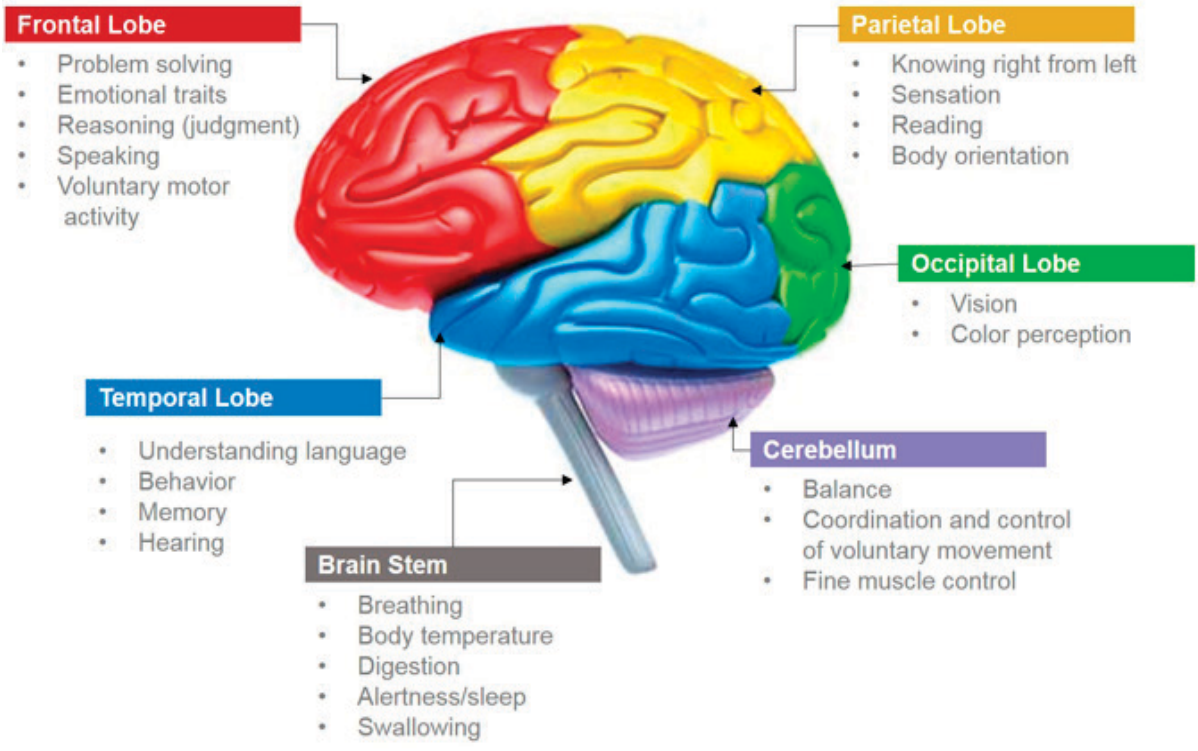



*Figure 22: Lobewise function of the brain*

**Why to assess and intervene Cognition???**

**The ability to SATISFACTORILY and COMPETENTLY engage in our usual ADL tasks :**

- **Go to work or school**
- **Experience leisure**
- **Interpersonal and social relationships**
- **Participate in our treasured occupation**

**……is highly dependent on COGNITION**

Cognition is often a hidden disability as the motor disabilities overshadow the underlying cognitive dysfunction which though leads to difficulty in performing Activities of Daily Living (ADLs)

**Factors that influence the patient's cognitive skills:**

- Age related
- Sensory loss- visual, auditory
- Visuopereception: perception of distance, depth , colour
- Cognition
- Environmental influence
- Physical-quite- more normal-normal
- Cues: external cuing – reduce external cuing- internal cuing

**Assessment Methods :**

- Clinical tests- non standardized and subjective
- Scales/Questionnaires- standardized and objective

**Intervention –Approaches :**

- Remedial approach
- Compensatory approach
- Sensory Integrative approach
- Neurofunctional approach
- Quadraphonic Approach

As mentioned in Chart no.1 there are different components of cognition. Following table describes the components more in detail with its definition and how it affects functionally.

*Table. 10 Functional deficits observed as per affected brain areas*

| Area of Deficit | Definition | Functional Description of deficit |
|---|---|---|
| Attention | Ability to focus on a specific stimulus without being distracted | • Exhibits inability to follow directions or learn<br>• May appear lethargic<br>• May have difficulty performing in group situation |
| Orientation | Refers to knowledge of time, place | • Unable to answer orientation questions<br>• May ask simple questions over and over<br>• May be easily agitated to disorientation |
| Memory | The registration and encoding, consolidation and storage, and recall and retrieval of information. | • Will forget names, schedule, etc.<br>• Decreased ability to learn or to follow direction |
| Problem solving | The ability to manipulate a fund of knowledge and apply this information to new or unfamiliar situations | • Difficulty with self care and households<br>• May be socially inappropriate<br>• Exhibit poor judgement<br>• Difficulty ordering or sequencing information |

*Table courtesy: [The Adult Stroke Patient, Barbara ZOLTON- 2nd edition]*

**Attention:**

***Types of attention***

*Focused attention* : responds only discretely to auditory , visual and tactile stimuli.

*Sustained attention*: ability to maintain attention during continuous and repetitive activity.

*Selective attention***:** ability to focus attention in the presence of distracting stimuli.

*Alternating attention*: ability to shift focus of attention from 1 task to another.

*Divided attention*: ability to respond simultaneously to multiple tasks.

***Assessment:***

- Clinical tests

✓ Digit repetition test

✓ Random letter test

✓ The test

- Clinical observation and activity analysis

***Intervention***

*Remedial approach:*

*Table 11: Therapeutic strategies for different types of attention deficits*

| Type of attention | Therapeutic activity |
|---|---|
| Focused Attention | Initially provide patients with single system sensory cuing and then progress to multi system cues |
| Sustained Attention | The patient reads a list of letter or letters, the patient indicates when he hears the letter b; then every time he hears b or h; then every time he hears each of those letters coming just one after the other |
| Selective Attention | While performing a task a tape is played which is of the individual's interest and thus may be distracting to the patient |
| Alternating Attention- tasks that require flexibility, redirection and reallocation of attention | Big-little task : the word big is and little are written in capitals and small letters; the patient is aske to read what is written as word or size<br>The patient is given a sheet with numbers on it and is asked to cross out even numbers; at some point when he is performing he is asked to stop and strike all the odd numbers |
| Divided Attention | The patient is asked to sort cards by suit and numbers; must also be turned as they are sorted. |

*Compensatory approach*

1. The patient should be alert enough to attend to task
2. Initially intervention should begin in a quiet room (uncluttered environment) then gradually progress to a more normal environment, alleviate chances of internal cues (pain , discomfort, stress and preoccupation)
3. Positive reinforcement in for of token economy can be helpful to improve attention to task
4. Prepare and orient the patient to task-bright colour interesting activity, give verbal as well physical prompts to the patient to make him attend to the task
5. Assist the patient in developing and utilizing internal

strategies-prepares himself to attend the task and vocalize step by step gradually sub vocalization (patient silently vocalizes)

***Orientation:***

Refers to an individual's ongoing attentiveness to situations, the environment , and the passage of time

- Components to be evaluated-time, place and person
- RBD patients found to be more impaired on orientation tests than LBD patient

*Assessment*

- Clinical test: questions related to the components
- Standardized:
  1. The Test of Orientation for rehabilitation Patients (TORP): used in IPD patients, 46 items-person and personal situation, place, time , schedule and temporal continuity
  2. The Galveston Orientation and Amnesia Test (GOAT): measures disorientation and amnesia following closed-head injury

*Intervention*

- Compensatory approach is useful:
  1. Don't let the patient guess…but let the person know Eg; introduce yourself while initiating conversation Eg; tell him where he is and where is he heading to
  2. Use of-cue cards -calenders, diary

***Memory:***

- It is the ability to process, store and retrieve information.
- Relationship between attention and memory- poor attention poor memory

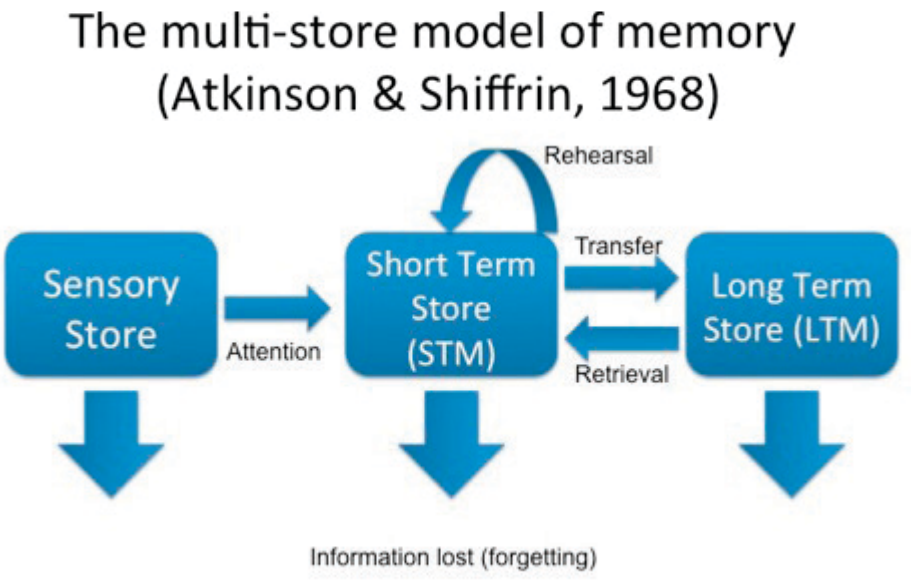



*Figure 23: The multistore model of memory*

*Pic. Courtesy: http://www.psychteacher.co.uk/memory/multistore-model.html*

Studies have been found that long-term memory (temporal lobe-dominant) exhibits the major deficit short term memory because STM is represented equally in both hemispheres and therefore less affected by unilateral lesions

*Assessment:*

- Clinical:
    1. Observation and Activity analysis
    2. 5 words recall-immediate, 5 mins, 10mins and 30 mins, üquestions
    3. stimuli dependent memory
- Standardized:
    1. Wechsler Memory Scale
    2. The Subjective Memory Questionnaire

*Intervention*

Remedial

1. Encoding information (sensory store-STM)
    i. Encourage attention by reducing distracting information
    ii. Simplify the information
    iii. Small amount of information at regular intervals
    iv. Make sure the information is understood
2. Storing information (STM-LTM) :
    i. Rehearsal
    ii. Elaboration
    iii. Visual memory
    iv. Mnemonics
3. Retrieving Information- using different types of cues(LTM-STM)
    i. Semantic cues- eg 1 word was a color
    ii. Phonemic cues- eg. Shoe- for shoelace/shoe maker
    iii. Contextual cues- eg. national animal- tiger
    iv. If the above cues fail- therapist provides the patient with multiple choices and he need to choose the correct one

Compensatory strategies

- Give cues as close as possible to the time the action is required
- Be specific of what is required
- The device should be portable, easy and independent.
- Use of time –tables, diaries, pictures, alarm clocks,
- Labelling doors and colour coding
- Utilize sensory inputs for better functioning

***Problem Solving:***

- It is not a single function, but rather the integration of several cognitive skills
- Problem solving requires both an intact fund of knowledge and the ability to manipulate and apply this information to new or unfamiliar tasks
- Closely related with attention to task, information access (both from environment as well as memory), organization, planning and judgement
- Stages of problem solving

- ✓ Preparation- identifying and understanding the problem
- ✓ Production- generating and executing possible solutions
- ✓ Judgement- evaluating the solutions generated

*Assessment:*

- Structured cognitive activity- copy design- block design, puzzle (2D or 3D)
- Clinical observation and activity analysis- evaluate which stage of problem solving is affected.
- Proverb interpretation test
- Social awareness and judgement
- Similarities and differences
- Conceptual series completion
  - ✓ Eg; 1,4,7,10,….
  - ✓ AZ,BY,CX,D……

*Intervention*

**Remedial:**

- Selection and execution of cognitive plans: listing steps, sequencing, discussion and modification
- Time management: time estimation and scheduling activity
- Self- regulation-increasing awareness of behaviour
- Activities: puzzles, analysis of proverbs, categorization activity or functional problem-solving tasks, computer games

**Compensatory:**

- Monitor how the task or the environment can be altered in order to improve problem solving skills
- Have the patient plan the steps of solution before executing it
- Utilize chaining of activity- breaking it down into functional components
- Vocalization and sub vocalization during the activity
- Group problem solving activity
- Provide external cues-
- Verbal cues-letting him know different strategies, helping him produce as many solutions and apply the same, help him evaluate his errors

***Perceptual dysfunction and intervention:***

*Perception:*

- Perception is the mechanism by which the brain interprets sensory information received from the environment
- Perception is the gateway to cognition
- The perceived information is then further processed by the various cognitive functions and the individual may choose either to respond with a verbal expression or motor act, or simply perceive and think about the observed stimuli

*Factors influencing performance of perceptual tasks:*

- Aphasia and Dysphasia
- Sensory loss
- Vision

*Types of Perception:*

Visual perception - Ability to identify and recognize familiar environment through sight alone

Tactile perception - ability to identify and recognize familiar environment through touch alone

of one's own body, objects in relation to oneself, and relationship between objects in space.

Motor perception – ability to plan and perform purposeful movement when a stimulus is presented

Visual Perception Disorders:

*Agnosia:*

- Loss of ability to recognize familiar objects via visual input with intact visual foundation skills
- Lesion Site: dominant occipital lobe
- AFunctional limitation: if he needs something, he won't be able to tell it to the caregiver, eg. Brush, glass of water.

Assessment:

- Informal, by asking the individual to identify 5 common objects by sight-pen, keys, watch, eyeglasses, comb
- Give cues in form of 3 choice of answers in case of word finding difficulty
- If the patient is aphasic ask him to nod (YES or NO) to the choices
- If unable to name 4/5 objects, visual agnosia is indicated

**Intervention:**

**Remedial**

- Having the client practice identifying familiar objects before and after the task- using intact senses eg.
- Touch to identify brush,
- Auditory to identify keys,
- Smell to identify perfume

**Compensatory**

- Labelling the common objects and keeping them at consistent locations eg.
- Brush in shelf of bathroom,
- Keys on keyholder,
- Perfume on dressing table

Colour agnosia:

- Lesion site: dominant occipitoparietal lobe
- Colour Agnosia- inability to remember and recognize the specific colours for common objects in the environment
- Functional dysfunction: Housewife mistaking the red colour of tomato to green colour of capsicum and buying the same
  - ✓ Assessment: present them with pictures of 2 accurately coloured and 2 inaccurately coloured objects- ask them to identify the inaccurately coloured objects

Colour anomia:

- Inability to name the colour of the objects
- Eg; young girl going for shopping and could not tell the shopkeeper the colour for her desired outfit
- Assessment: ask the individual to name the colour of various objects in their environment, if aphasic ask them to nod (YES or NO) to the choices of colours offered
- Intervention: ask the patient to recognize, identify and name the colour, best if done in familiar environment

Metamorphasia:

- Visual distortion of objects such as the physical properties of size and weight
- Lesion site: non-dominant temporal lobe
- Functional Dysfunction: sit on a chair smaller for oneself, can push a light object with more energy than needed
- Assessment: present the subject with familiar objects like balls of different sizes, glasses filled with water at different levels and ask the individual to arrange them in order of size and weight respectively
- Intervention: Make people identify the objects used in familiar tasks though intact sensory modalities. eg tactile-kinesthetic. Therapist can also provide verbal descriptors of the object when using this approach.

Prosopagnosia:

- Lesion site: non dominant parietal lobe
- Inability to recognize and identify familiar faces –leading to difficulty in social situation

Assessment:

- Standardized test of Facial Recognition: presents a multiple choice of faces presented in a front view and side view, under various lighting conditions
- Informal: presenting photographs of family members on social occasions, photographs of famous faces and ask them to identify the names

Intervention:

**Remedial**: Providing face matching exercises along with naming them

**Compensatory**:

- Providing pictures of family members and famous people with names
- Assist to associate the family member's face with other unique characteristics and features such as weight, height, mannerisms, accents and hairstyles
- Introduce yourself before striking a conversation with that individual

Simultanognosia:

- Refers to inability to recognize and interpret a visual array as a whole
- Able to identify the individual components of a visual scene but unable to recognize and interpret the gestalt of the scene
- Functional Dysfunction

  Eg. A student might read letters but won't perceive the whole word, If a retiree wants to go to a beach- he might give cues take me t the pace where there is water, sand, where we can make sand castles, but will be unable to tell that lets go to a beach

Intervention

- Remedial: assist the individual to construct a meaning of a visual array through verbal cues and therapeutic questions to facilitate abstract reasoning
- Compensatory: if training provided in familiar contexts such as home, work setting, social outing

**Tactile Perception Disorders:**

Astereognosis:

- Lesion site: parietal lobe
- Inability to recognize objects or forms by handling, although tactile function is intact.
- Functional Dysfunction: difficulty finding a pen from handbag

**Assessment**:

individual is asked to identify familiar objects by giving the object in hand to manipulate but with vision occluded

**Intervention:**

Egger's method: allow the patient to manipulate object by seeing and making noise such as tapping- then a pad is put on the table with vision occluded. progress from grossly dissimilar objects-rough and smooth, estimate the quantities through touch, discriminate between large and small, between 2D and 3D, lastly to pick up small object from several others

Farber's method: patient is asked to examine the training object when rotating in examiners hand in unaffected hand- both hands and lastly in affected hand- can place hand in a mirror box, the same is repeated with vision occluded- objects placed in rice-sand or a black bag

Agraphesthesia:

Lesion site: parietal lobe

- Inability to recognize numbers, letters, or forms written on skin with vision occluded
- Assessment: examiner occludes the patient's vision and traces letters, numbers or shapes on the dorsum of hand with a dull-pointed pencil or fingers.
- Intervention: let the patent relate the tactile stimulus with eyes open and then with eyes closed. Gradually progress from letter to numbers to words to shapes.

Body schema Perception Disorder:

- Ability to identify parts on self or others
- Functional dysfunction: when a patient is asked to shift his hand or leg inappropriately i.e. in a train
- Assessment – patient is asked to identify body parts on self and others both separately as well as in combination with R/L tasks.
  - ➢ Intervention
  - ✓ The therapist names a part followed the patient rubbing the area. Eg you rubbed your nose , you touched your legs (C)
  - ✓ The patient verbally identifies the parts of the body as they are touched or moved. Eg .I m moving my hand or you touched my right hand (R)
  - ✓ The patient mimics the therapist, who moves her head and limbs in different positions (awareness of body parts of others) (R)

**Anosognosia:**

- Lesion site: nondominant parietal lobe
- Refers to an unawareness of hemiplegia, can range from an unawareness of weakness, to lack of concern to total denial
- Functional dysfunction: patient will insist on being independent in his ADLs specially bilateral UE activities such as reaching, or insist to drive by himself get up to stand despite of weakness in 1 sided LL and tends to fall

Assessment:

- On draw a man test. He might draw

his left arm and leg to the extreme of page indicating that he believes these limbs are no longer his part

- Interviewing the patient: whether anything wrong? He might dent his illness
- On confrontation that the side is paralyze and will not move in response to his efforts the patient may reply that the limb has a will and purpose of itz own, that it is tired, or that it always was a lazy arm

Intervention:

- If the patient denies he has a problem, little can be done to teach him to compensate,
- As the patient begins to recover, efforts must be made to make the patient aware of his deceit eg. Making him match the demand of the task, his expectations of outcome and the analysis after the performance of the tasK

Unilateral neglect:

- Lesion site: inferior parietal lobe of the right non dominant hemisphere
- It is the inability to respond to orient to stimuli from the left side of the body or the left side if the environment, however the patient is aware of the illness
- Also referred to hemi-inattention, hemi-spatial neglect, and unilateral spatial agnosia
- Functional dysfunction: shave only right side of the face, eats food on right half of the plate, bumps into things on his eft side or constantly turning to his right

Assessment:

- Draw a man, draw a clock
- Copy flower, house
- Human figure or face puzzle- interpretation on time and no. of errors
- Cancellation tasks
- Reading

Intervention:

- Remedial:

1. Provide cutaneous and proprioceptive feedback to the affected side eg rubbing, pressure, ice vibrator, active assisted activities with skate board in front of mirror
2. Use of neurodevelopmental treatment approach- participate in bilateral tasks, Eg. Wand exercises, proprioceptive facilitation techniques through weight bearing activities or tie weighted cuffs while doing activities in later stages
3. Anchoring technique for reading

- Compensatory:

1. Give activities which forces patient to turn to left side, eg. Peg transfer from left to right,
2. Visual guides to look for before reading a line- red line drawn at the left side of the page, have his finger placed at the left side id the page
3. Practice him turn his head or shift his eyes to either side while performing task
4. Provide verbal cues while eating
5. Use mirror while dressing
6. Severe neglect- put food on unaffected side, speak to patient om unaffected side, stand on the unaffected side

Visual-spatial Perception Disorders:

*Figure-ground Discrimination Dysfunction:*

- Lesion Site: non dominant parietal

lobe

- Inability to perceive the foreground from the background
- Functional Dysfunction: inability to identify the white shirt kept on bed with white bedsheets, finding a pen from a cluttered drawer

Assessment:

- LOTCA
- SIPT
- The Figure- Ground Perception Test
- Functional tests

Intervention:

Remedial: ask to identify object of similar colour as background progress from simple to complex visual array

- Compensatory: organize the environment- having only necessary objects around, labelling the objects with similar colors.

*Position in space:*

- Lesion site: non dominant parietal lobe
- Refers to the relative inability to perceive himself in relation to other objects
- Functional dysfunction: when asked to get the basket under the chair, he might not know where to look for it

Assessment:

- Two dimensional drawings
- Positioning blocks
- Functional positioning- component of LOTCA- ask the patient to place the object in different directions in respect to another object

Intervention:

- Felt pictures-2D- eg present a picture with a house and tree on right and give another card with 2 pieces and ask him to position the tree according to the copy
- Using 2 1inch blocks and positioning 1 block in various position in relation to the other block.
- Gradually incorporate the same into functional task, eg. Arranging a book shelf or storing fruits and vegetables un fridge
- Verbalize the positions of the object during the task

*Right-Left Discrimination dysfunction:*

- Lesion site: parietal lobe
- Inability to understand and use the concepts of right and left
- Functional dysfunction: wear socks in opposite foot, difficulty navigating

Assessment:

- Informally, grade by asking the patient to identify his body parts then the therapist's body parts then in combination, initial double uncrossed(R-R) then increase the complexity by double crossed(R-L)
- Benton's standardized 20 item test

Intervention:

- Remedial

1. Give a selected hand extra tactile or proprioceptive stimulation, eg. Tying weighted cuff to right hand, be consistent and do not switch hands
2. Quiz the patients as in the test
3. stress right and left verbally when he is performing a task, later make him verbalize when interacting with their own body, eg I am putting my Right hand into the shirt sleeve or when reacting with their environment eg.i am turning left to the stop

sign

- Compensatory

1. Patient wears his watch on left side to provide him cue to distinguish between right and left side in order to use
2. On clothing, shoes, and so forth, mark the right side different from the left side either coloured tape, a marker or a sticker

*Topographical Disorientation:*

- Lesion site: non dominant occipito-parietal lobe
- Difficulty understanding and remembering relationships of places to one another.
- Functional dysfunction: Difficulty finding his way to home or hospital

Assessment:

- Functional tests- observation and reporting by the caregiver or the patent himself
- Ask the patient to draw a map from his ward to OT department or his home to the grocery shop nearby

Intervention:

Remedial:

An activity where in the therapist walks alongside the patient to reach a target destination in the therapy room (door to toilet) by verbalizing the directions used to reach and later asking the patient to follow the same route by verbalizing by himself

Compensatory:

1. Ask the patient to not go out by yourself, do not drive
2. Use markers like coloured dots to mark a route the patient must travel every day
3. Let the patient always carry an I-card around his neck with basic details in case if they get lost, the fellow pedestrians might help them get back home

➢ Stereopsis: Inability to perceive depth in relation to the self or in relation to various objects in the environment

➢ Functional dysfunction- the patient misjudges depths and distances so that he may have difficulty navigating stairs and architectural barriers eg. Driving and community mobility

Assessment:

- Block test
- Functional test

Intervention:

- Obstacle course
- Put finger in the glass to judge the filling level of water
- Climbing up and down stairs and moving his foot slowly along the different step height
- Mark the steps with bright colour tapes or markers

*Motor Perception Disorders*

❖ Ideational and Ideomotor apraxia:

- Lesion site: Dominant parietal lobe

❖ Ideomotor apraxia:

- Inability to carry out a motor act on verbal command however, the person is able to perform the task correctly when sked to us the actual object
- Functional dysfunction: if the therapist asks the patient to mime the action of brushing but can perform it spontaneously during routine

❖ Ideational apraxia:

- It is a conceptual deficit as inability

to use real objects appropriately

- Functional dysfunction: the person may use the wrong thing for the task or may associate the wrong tool with the object to be acted on, such as by attempting to write with a spoon

Assessment:

- Goodglass test of apraxia
- LOTCA battery

Intervention:

- Treatment is not usually necessary with ideomotor apraxia because the patient is able to use the limbs spontaneously
- Ideational apraxia is treated as follows:
- Interpretation and use of tactile, kinaesthetic and proprioceptive stimulation to influence motor patterns, physical prompt
- Chaining techniques
- Break down the task in small components
- Verbal cues from therapist should be kept minimum and let the patient visualize the movement before carrying out

*Dressing Apraxia:*

- Is an inability to dress oneself.
- Lesion site: nondominant occipital or parietal lobe
- Assessment: functional observation and reporting
- Intervention: positioning the garment
- Colour coding the garments sidesrt lt , front back,
- Start buttoning from down with circling the button with his finger all the way through button loops

*Constructional Apraxia:*

- It is the impairment in producing designs in 2D or 3D, by copying, drawing, or constructing whether upon command or spontaneouslyS
- Functional dysfunction: for an architect it will be difficult for him to draw a layout of building, difficulty making a simple sandwich for a housewife

Assessment:

- Copying designs -2D and 3D
- Matchstick design block
- Block design

Intervention:

- Practice simple copying or construction task -Move from 2D to 3D
- Functional task of making snacks or organizing shelf with verbal and gestural prompts

**Vocational rehabilitation**

It is important that people are able to undertake education, employment, re-training and voluntary work to improve their quality of life, fulfill a role in society, avoid low self-esteem and depression. There is evidence that this can be achieved by offering vocational rehabilitation (VR) to people after a stroke96.

Both paid and unpaid work fulfils a diversity of needs for the individual, and some of the many benefits of work are listed below:

- Increased self-esteem.
- Maintenance of routines and habits.
- Participation in a productive activity.

- Involvement in a socially accepted role that provides value to the community.
- Having 'a reason to get up in the morning'.
- Challenging someone to expand their horizons.

Occupational therapists can still assist the patient in establishing realistic goals and expectations towards work and produce a written joint plan of action.

**Resuming sexual activity**

As rehabilitation of stroke patients constantly improves, many new techniques develop but some activities are often neglected because of lack of research, lack or rehabilitation knowledge or because patients themselves do not wish to raise the subject. One such area is a return to their normal sexual activity. Generally, this is not routinely discussed with patients as part of their rehabilitation, although for many, it plays an important role in their life and discussion should be included in the assessment process.

Research has observed that after stroke there is a decline in sexuality in both genders, and partner dissatisfaction is high. There are many reasons for this decline: physical changes in the brain that may reduce the sexual urge; physical changes to the body that may make it difficult to move; psychological changes that make people not want to have sex. Stroke can cause physical limitations that influence body positioning and movement during sex which can lead to a reduction in sexual activity, misery and frustration. Other physical changes such as incontinence, drooling, emotional liability may be off-putting to partners and again lead to a reduction in sexual activity.

As well as physical difficulties, psychosocial impairments and depression may affect the will to engage in sexual relationships; however, these may not be recorded till late after stroke as patients may be unwilling to talk about sexual difficulties. Mood disorders, such as depression and anxiety are commonly observed after a stroke, frequently affecting sexual relationships and sexual function and conversely, sexual dysfunction may lead to depression.

After a stroke, generally both patients and partners want to know whether resuming sexual activity will cause another stroke or epileptic fit and do not know whom to approach for advice. Although there is only a low risk of stroke from sexual excitement, people are unwilling to have sex as they believe it may cause another stroke. Sexual activity is a subject which is important to both patients and partners, and should be included in stroke rehabilitation.

Occupational therapists are often approached to discuss sexual activity whilst they are dealing with other personal activities of daily living. This chance to contribute to part of patient's lives often ignored, should be embraced with knowledge and understanding.

Patients and partners need to know that returning to their normal sexual activity is considered routinely as an aspect of stroke rehabilitation. They also need to know there is the opportunity to discuss their sexual activity either alone or in couples, at a time which is appropriate to their needs.

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**Speech therapy management of stroke**

Overview:

Stroke, as we know, may involve many areas of the brain; i.e. it could be a posterior region stroke, caused by damage to Posterior Cerebral Artery (PCA), or a temporal and parietal region stroke caused by dam-

age to Middle Cerebral Artery (MCA), or a frontal region stroke caused by damage to the Anterior Cerebral Artery (ACA). There are many more arteries of the brain where damage is possible which may lead to stroke. Damage to specific areas of the brain manifest into Speech- Language problems.

The brain is a highly specialised structure, having specific areas assigned for specific functions; i.e. there is a specific area for walking, talking, eating, writing, thinking, etc. Depending upon the area of brain that is damaged, different Speech- Language- Swallowing symptoms are observed. In order to make a clear understanding of the symptoms, let us understand the regions of the brain that are assigned Speech- Language- Swallowing functions.

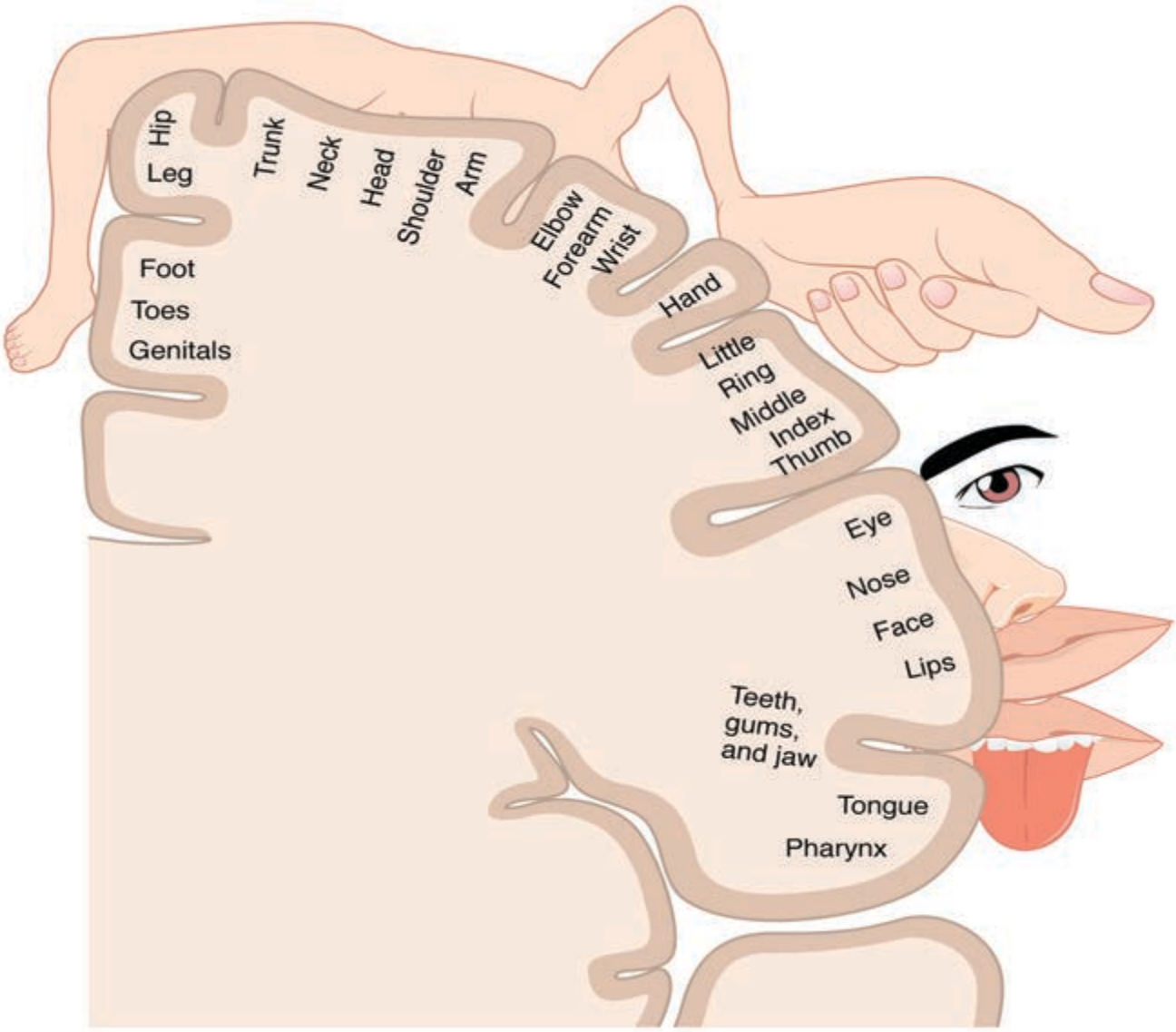



Figure 24: Homunculus of the brain

**Language:** The brain has two hemispheres: left and right. In 90% of the individuals the left hemisphere is specialised for language, i.e. different areas in the left hemisphere, specifically the Broca's area and the Wernicke's area have language blueprints. Therefore, damage to the left hemisphere results in language problems.

**Language dominant: Left Hemisphere**

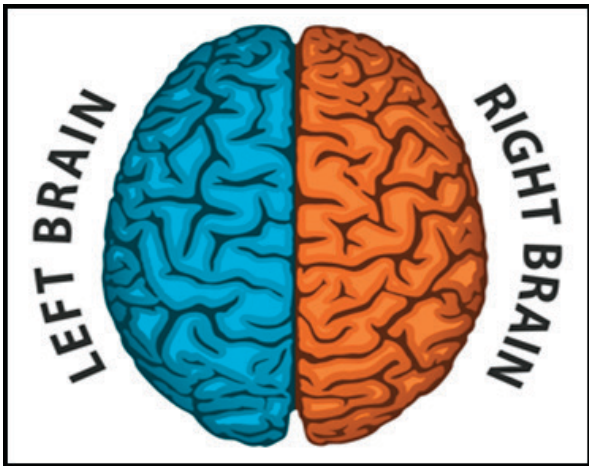



Figure 25: Hemisphere specialized for language

**Speech:** Speech is a motor function. Speech is articulated by the movement of the tongue, lips, vocal cords and other oral articulators. The movement of these articulators comes from the motor strip of the brain which is situated in the left hemisphere. This specialised motor strip is called as the "homunculus".

As seen in the figure above, the homunculus, has specific regions for the movement of hands, legs, lips, tongue, etc. The regions on the homunculus representing motor movements of the tongue, lips, and other oral muscles occupy more space than that of hands and legs. This is because the hands and legs carry out gross motor movements, while the oral muscles carry out fine motor movements, thus requiring a broad representation.

**Swallowing:** Swallowing is a highly complex function. It consists of five phases: anticipatory, oral preparatory, oral, pharyngeal and esophageal. The oral preparatory, oral and pharyngeal phase are under voluntary control while the esophageal phase is under involuntary control. Both the voluntary and involuntary control of swallowing come from the brainstem, mainly pons and medulla.

**Symptomatology:**

**Language Symptoms:** Language problems caused secondary to stroke are clinically called "Aphasia". Depending upon the site and extent of the brain damage, different types of Aphasias are possible. Various authors have given different classifications of Aphasia. The classification given by Davis, 2007 is the most recent and most popularly used classification of aphasia. This classification is based on the Goodglass & Kaplan classification given in 1972. This divides aphasia into 2 types: non-fluent and fluent aphasia.

I. Non-fluent aphasia consists

**Brainstem: Midbrain, Pons, Medulla**

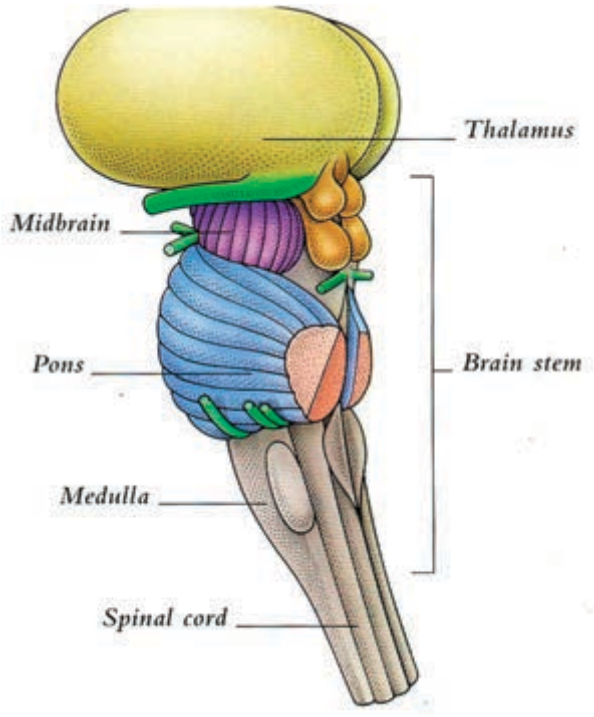



Figure 26: Brainstem

**Classification of Aphasia.**

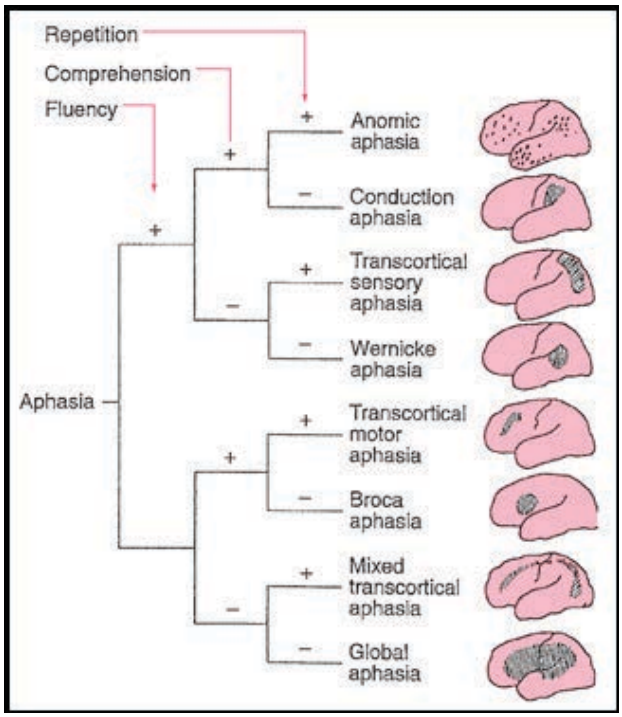



Figure 27: Classification of Aphasia

of: Global aphasia, Broca's aphasia, Transcortical motor aphasia

II. Fluent aphasia consists of: Wernicke's aphasia, Anomic aphasia, Conduction

III. aphasia, Transcortical sensory aphasia.

IV. Some authors also include one more type of aphasia, i.e. Transcortical mixed aphasia.

Symptoms of different types of aphasias can be best understood in the form of a chart given above. This chart is given by American Speech and Hearing Association (ASHA) in 2011. In the description of types of aphasia, ASHA also mentions about "crossed aphasia" where language problems are seen even when the right hemisphere is affected. This is seen in less than 10% of the population where the right hemisphere is specialised for language instead of the left hemisphere. These individuals usually show left handedness.

**Symptoms of different types of Aphasia**

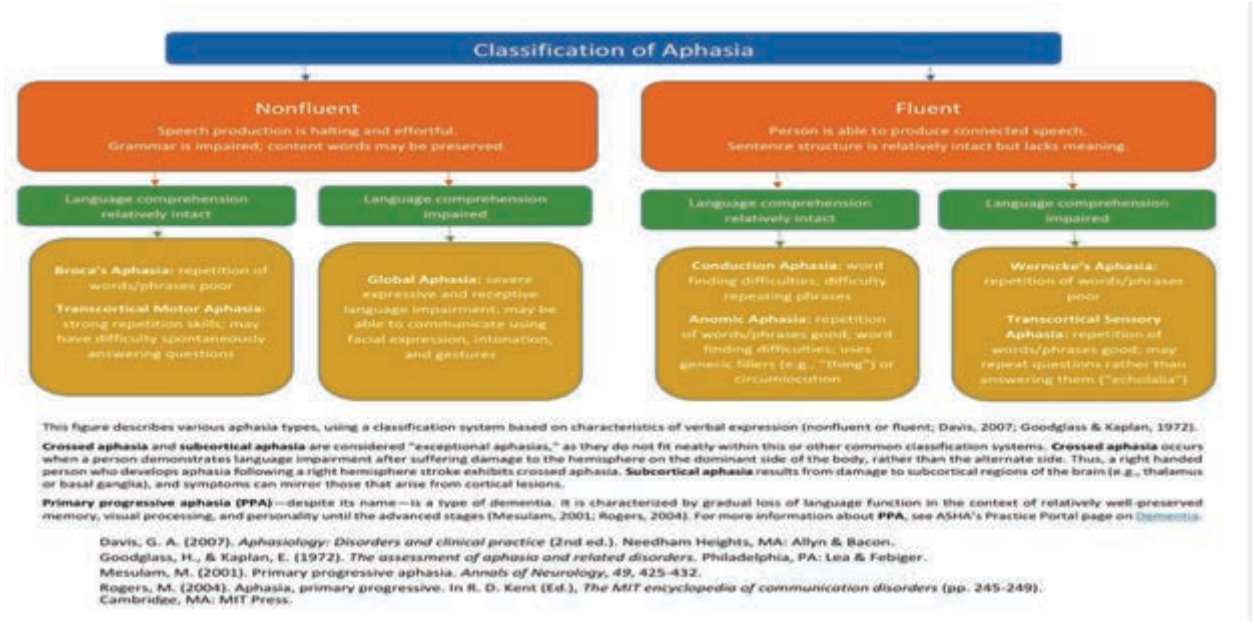



Figure 28: Symptoms of different types of apraxia

**Figure 29: Aphasia Therapy: Orientation**



**Figure 30: Triball**



***Speech Symptoms:*** If the motor strip is affected, it causes a speech disorder called "dysarthria". Dysarthria is a speech disorder manifested by slurring of speech and imprecise articulation. Depending upon the articulators involved in the speech disorder, the types of dysarthrias are classified. There are six types of dysarthrias namely: flaccid, spastic, ataxia, hypokinetic, hyperkinetic, mixed dysarthria.

***Dysphagia Symptoms:*** "Dysphagia" is the clinical term for dysfunction in swallowing. Symptoms of dysphagia are observed when there is a damage to the brainstem. More specifically, a PICA damage causes a swallowing dysfunction, i.e. muscle movement is uncoordinated causing aspiration of food. On the other hand, a MCA damage causes swallow apraxia, i.e. the individual can swallow involuntarily but does not recognise the need to swallow when a bolus is in the mouth.

**Treatment options:**

***Aphasia Treatment:*** In most cases of aphasia, treatment needs to be started with orientation and following simple commands.

Many researchers have contributed to specific therapeutic treatment for aphasia. The most popularly used treatment options for aphasia are Music Induced Therapy (MIT), Schuell's Stimulation Therapy, Phonemic Based Therapy, and many more.

All the above mentioned therapy techniques are used for a specific feature of aphasia, ex. non-fluent speech, poor auditory comprehension, naming, repetition, etc. The Speech-Language Pathologist will use the most suitable approach for a given individual with aphasia, after having considered his/her language deficits.

Many individuals with aphasia have dysarthria as one of the symptom of aphasia. In these cases, both aphasia as well as dysarthria needs to be treated.

***Dysarthria Treatment:*** Individuals with dysarthria present with an affected voice quality, resonance, inappropriate articulation, uncoordinated breathing, affected fluency and affected prosody. All these factors together contribute to slurred speech quality.

***The Speech-*** Language Pathologist will prescribe therapeutic techniques which target specific systems, like the respiratory system, the phonatory system, the prosodic system and the articulatory system,

which in turn brings an improvement in the overall quality of speech. Some of these techniques are:

***Use of Tri-ball:*** This exercise focuses on building breath support required for speech so that the individual does not feel fatigued after speaking.

***Use of Finger Brush:*** This is a massage done by the caregiver on the patient's tongue. It is done in the directions of the tongue muscle in order to strengthen them. Massage with the finger brush is also done over the lips and in the buccal cavity to strengthen the lip and buccal muscles respectively.

***Oromotor exercises:*** These exercises include exercises of the oral articulators. They include the tongue, lips, cheeks and velum. These exercises are to be done by the patient himself unlike the massage with the finger brush.

All the above mentioned are restorative techniques for dysarthria treatment. On the other hand, there are compensatory techniques for dysarthria. These techniques are used when the overall prognosis of speech is poor. A few of them include:

- Talking at slow rate.
- Talking with "Open-mouth-approach".
- Taking conversation turns while talking so that optimum rest is achieved.
- Stressing on every consonant while talking.w

**Dysphagia treatment:** As the pharynx and larynx are close to each other, there is a high chance of food bolus entering into the larynx (wind pipe) due to an uncoordinated swallow. Thus, dysphagia treatment is highly critical and needs be done with utmost care. Slight incoordinance during therapy can cause either choking, or aspiration leading to aspiration pneumonia, both of which are potentially fatal.

In order for the Swallowing Pathologist to understand the patient's swallowing problem, he/she needs to carry out subjective and objective tests. The subjective tests include testing with palpation and auscultation. Objective tests for swallowing involve Modified Barium Swallow (MBS) and Flexible Endoscopic Evaluation of Swallowing (FEES).

Once the exact swallow dysfunction is identified, dysphagia therapy can be initiated. Dysphagia therapy is broadly divided into two: indirect and direct swallowing therapy. Indirect therapy, is done with patients who have a very poor swallow function, i.e. reduced or no hyolaryngeal excursion. In indirect therapy no food is given; swallow function is im-

**Figure 31: Oro-motor exercises**

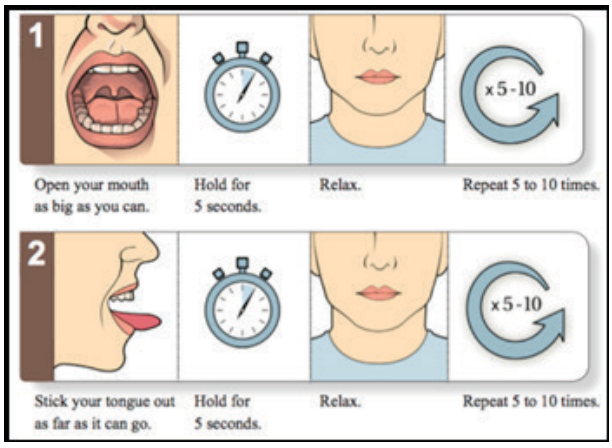
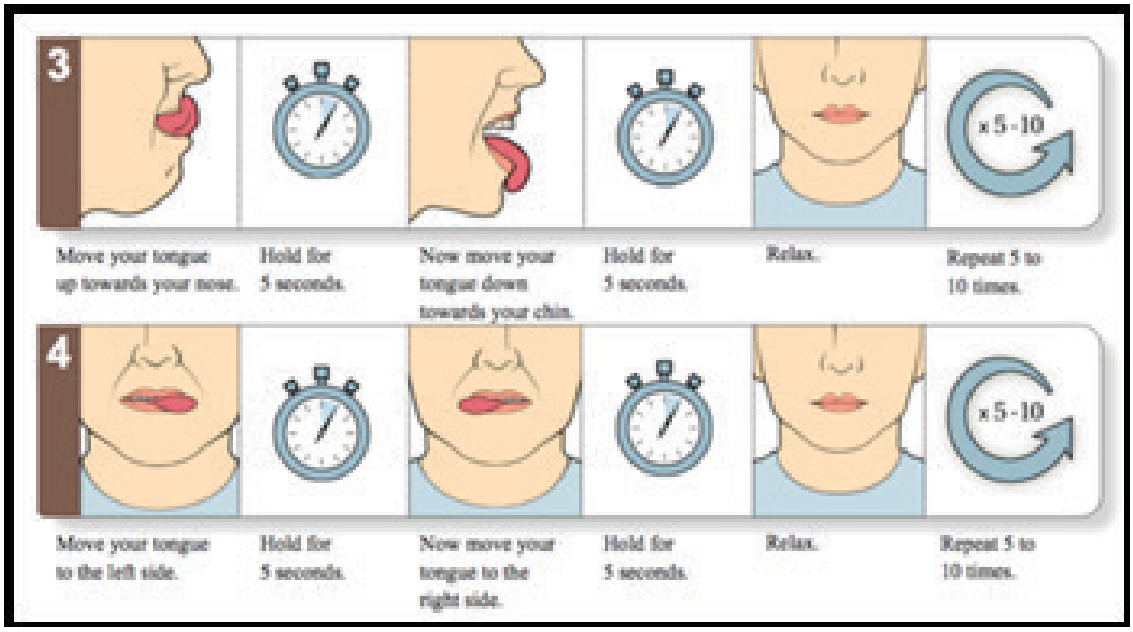



**Figure 32: Finger Brush**






proved using only exercises. On the other hand, direct therapy is when maneuvers and techniques given with the administration of food.

There are a number of exercises, techniques and maneuvers that are involved in swallowing therapy, which are prescribed by the Swallowing Pathologist.

The Swallowing Pathologist gives Trial Feed using suitable maneuvers and techniques and takes a feedback of whether the bolus has entered the oesophagus safely, or has entered into the airway. This feedback is obtained by using palpation or auscultation technique.

If the bolus has entered the airway, coughing out the bolus is the most commonly used technique; but failure of coughing out compels the Swallowing Pathologist to perform other push-out maneuvers which help to dislocate the bolus from the airway. If the bolus safely enters the oesophagus, the techniques and maneuvers used are reinforced and taught to the patient.

Even when the patient is able to tolerate all consistencies, he/she must follow the safe swallow techniques, prescribed by the Swallowing Pathologist to avoid any further risk.

**Psychological management of stroke**

Cognitive, behavioral and adaptive challenges may be faced by children following a stroke. Cognitive problems are seen in many children suffering from ischemic and hemorrhagic stroke. According to research, intelligence scores are found to be skewed to the lower end of the average range and impairment is greater in performance domain as compared to verbal one. Executive functions like attention and processing speed are vulnerable to the effects of pediatric stroke. The indi-

**Figure 33: Flexible Endoscopic Evaluation of Swallowing**



vidual's age at stroke, larger impact size, cortical or sub-cortical lesion location, epilepsy, and co-morbidity of physical deficits lead to greater cognitive deficits. Cognitive rehabilitation for survivors of pediatric stroke include therapeutic activities ranging from teaching introspection and self-monitoring, self-awareness, and self-advocacy skills for conducting executive functions, academic skills, social skills, and activities of daily living. Group counseling or play therapy can be found effective depending on the age of the injured child. Homogeneous groups allow injured individuals to identify with each others' problems, and to receive feedback from each other with safe interpersonal conversation. Group interventions helps in learning to solve a problem or to perform a particular task together and thus participating in the learning process. A holistic rehabilitation program improves children's ability to cope, and increase their self-awareness and cognition leading to general social integration. Children's difficulties with social skills stem from social isolation. Usage of arts-based interventions to increase self-confidence and improve social participation can be of great help. With training, children can be beer at articulating their feelings, reflecting upon them with increased abstract thinking, and participate socially. Because children spend a majority of their time in school, the academic seeing therefore provides the most opportunities for rehabilitation. These interventions for cognitive deficits need to be integrated into children's classroom experience as well as daily life activities. So, caregivers and school authorities need to collaborate concerning the child's abilities and limitations, in order to develop an appropriate academic program. Pediatric stroke also affects the psychological well-being of the family as the identification of the cause and diagnosis of pediatric stroke is usually delayed leading to transmission of misinformation through various sources. This leads to many mothers of children with perinatal stroke to assume that they are somehow responsible for their child's brain injury and its consequences. Misplaced guilt on self and sometimes doctors can have a major effect on their mental health. Psycho-education about causes and the expected effects of a stroke can reduce the feelings of guilt as well as prepare the parents for what to expect. Therefore, parents and family members should be included in goal-setting and developing individualized care plans for each child who has had a stroke, and offered appropriate support throughout the journey of recovery. Children and adolescents who suffered from a pediatric stroke who have cognitive impairments benefit from a holistic and integrated approach to rehabilitation which incorporates developmental, social, and emotional needs, as well as, cognitive rehabilitation techniques. Recovery is believed to depend on stroke severity, timing of treatment and initiation of appropriate therapy.

**Feedback using Cervical Auscultation**



# Chp. 7 Head Injury

**Dr. Joji Joseph, Dr. Arjun K.M., Dr. Monisa Ansari,
Ms. Nikita Khokhani, Dr. Snehal Sontate**

Head injury includes injuries of the scalp, skull, or brain. The most common cause of head injury is trauma which may be road traffic accident, fall or physical assault.

Head injuries can be classified in to closed or open depending on whether the duramater is intact or breached. A closed head injury is one in which duramater remains intact. There may be injury to the scalp or cranial bones. An open head injury occurs when a penetrating object breaches the duramater.



Figure 1: Image showing an example of Head Injury

A head injury may result in only scalp wounds. In more severe cases it may result in skull fracture and direct or indirect injury to the brain. Injury to the brain may be in the form of concussion, contusion, hemorrhage or hematoma.

Patients with head injury may be asymptomatic at the time of injury and may develop symptoms over a few days. Symptomatic patients may present with nausea, vomiting, headache, confusion, drowsiness or seizures. There may a lucid interval during which the patients remain conscious after head injury and deteriorates later. Unconscious after a head injury even for a short period is not normal. The patient may have other symptoms like difficulty concentrating, increased mood swings, lethargy or aggression, and altered sleep habits. These symptoms can occur in concussion type of head injury and can easily be missed.

The physical examination and the history of the exact details of the injury are the first steps in caring for a patient with head injury. Patient should be assessed for symptoms suggestive of severe head injury- severe headaches, fluid draining from nose / mouth / ears, loss / alteration of consciousness, confusion, drowsiness, slurred speech, blurred vision, stiff neck or vomiting, paralysis.

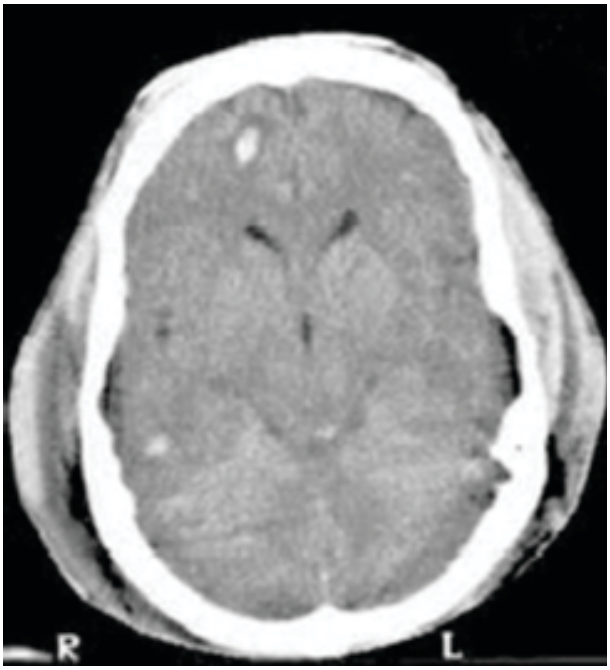



Figure 2: CT scan image of head injury

The Glasgow Coma Scale (GCS) should be used to assess the level of consciousness, severity of head injury and to determine the prognosis.

Minor head injury: GCS 13-15; mortality 0.1%

Moderate head injury: GCS 9-12; mortality 10%

Severe head injury: GCS <9; mortality 40%.

The GCS should be assessed periodically to determine the neurological status of the patient.

Examination of pupil forms a vital part of neurological assessment. The response of pupils to light and their size should be examined. Anisocoria, unequal pupil size, is a sign of serious head injury. Pupil not reacting or reacting sluggishly to light is suggestive of raised intracranial pressure.

Neuroimaging helps in determining the diagnosis, prognosis and in deciding the treatment. Non contrast Computerized tomography (CT) scan of the head can show bleeding and swelling in the brain. It can also evaluate bony injuries to the skull and bleeding in the sinuses of the face associated with basilar skull fractures. CT does not assess brain function, and patients suffering axonal shear injury may be comatose with a normal CT scan of the head.

**Management of head injury**

**Medical management of head injury**

**Treatment:**

If you suspect a skull fracture, do not apply direct pressure to the bleeding site, and do not remove any debris from the wound. Cover the wound with sterile gauze dressing.

***Linear Fracture*:** No Surgical treatment required, analgesics, needs 24 hours observations admission.

***Closed Fractures*:** (No Scalp Defect)

***Depressed Fractures:*** Have to be operated and surgically evaluated, anticonvulsants to be started. These patients have high chances of post injury epilepsy.

***Fractures of Cranial Base:*** Require hospitalization, to be treated with high antibiotics as there might be the case of CSF leak and meningitis.

***Management of CLW's:*** Even if the patient is referred to another hospital, it is always advisable to suture the CLW of the scalp. If patient is not shifted to another hospital then start with broad spectrum antibiotics and con-tinue till suture removal. That is after 10 days.

***Reference:*** **refer the patient urgently to a neurosurgeon.**

***For a moderate to severe head injury, take the following steps:***

- Check the person's airway, breathing, and circulation. If necessary, begin rescue breathing and CPR.
- If the person's breathing and heart rate are normal but the person is uncon-scious, treat as if there is a spinal injury. Stabilize the head and neck by plac-ing your hands on both sides of the person's head, keeping the head in line with the spine and preventing movement.
- Stop any bleeding by only pressing a clean cloth on the wound.If the injury is serious, be careful not to move the person's head. If blood soaks through the cloth, Place another cloth over the stone.

***For a mild head injury***

- No specific treatment may be needed. Advice to, closely watch the patient for any concerning symptoms over the next 24 hours.

***Patient's relatives should watch for***

- Vomiting
- Headache not relieved after a few hours
- Drowsiness
- Irritability
- Seizure
- Unusual or confused behaviour
- Bleeding or discharge from the ear or nose
- Weakness or numbness in arm or leg
- Altered vision

The symptoms of a serious head injury can be delayed. While the person is sleeping, wake him or her every 2 to 3 hours and ask simple questions to check alertness, such as "What is your name?"

Treatment: Analgesics may be used for a mild headache. Do not advice aspirin, because it can increase the risk of bleeding.

***Brain Hemorrhage Can be classifed as three types that is***

- *Extradural:* Extradural hematomas are often secondary to a skull fracture and tear of the meningeal arteries below the site of primary impact.
- *Subdural:* Subdural haemorrhagedevelops immediately at the time of injury, or slowly thus causing symptoms few days after the original head injury also.
- *Intracerebral:* They are uncommon after a head injury except in case of gunshots or penetrating injuries.

In most cases a patient will lose consciousness for a little interval. He may seem lucid on waking up but then as the hematoma develops begins to increasingly complain of headache, irritability, confusion. If these haematomas are large and are causing increase in the ICP they have to be removed surgically by craniotomy.

Hematomas are potentially life threatening and can cause extensive brain injury, including coma, and even death. This is due to the effects of compression and raised ICP. Because of the compression the brain will press the various tentorial compartments, or even be forced down into the foramen magnum at the base of the skull (herniation)thus compressing the brainstem,which can lead to death.

Usually the oculomotor nerve becomes compressed resulting in pupillary dilation, and loss of eye movement. As the brain stem gets compressed, patient begins to experience rapid changes in conscious-awareness, becomes stupor-ous, and may become comatose with irregular respiration. It may result in hemiplegia or decerebrate rigidity (i.e. extension of the extremities).

**Brain Injury**

These are further two types contusions that occur in the outer part of the brain and diffuse axonal injury that occur in the central part of the Brain. These require ICU management with ventilator support for upto 48 hours, and are treated medically with

- Mannitol 100cc IV 8 hrly
- Dexamethasone 4 mg IV 6 hrly
- Eptoin 100 mg IV 8 hrly

**Brain Death Occurs when the brain is irreversibility damaged. These patients can be declared dead for the purpose of organ transplantation. The clinical signs of Brain death are:**

**a**. Deep unconsciousness

b. Fully dilated and non-reacting pupils.

c. Absent doll's eye movements

d. No motor response, to deep central pain.

e. No spontaneous respiration.

**Surgical Management:**

The factors that help in decision making regarding the need for surgery include age, neurological status, mechanism of injury and pupillary reactions. The major questions addressed by the surgeon are:

1) Is the conscious level depressed?
2) Are there other signs of raised ICP (e.g. oculomotor palsy)?
3) Does the scan show a hematoma or contusion with mass effect (midline shift / ventricle effacement / dilatation of contralateral ventricle / loss of basal cisterns / sulcal effacement)?
4) Is expansion of the mass anticipated

(e.g. contusions)?

5) Where is the mass (temporal lobe lesions are considered dangerous due to the association with uncal herniation compressing the mid-brain)?

These questions must be appropriately addressed rapidly and when required the surgery should be performed with an appropriate sense of urgency.

1. **Extradural Hematoma:** The indication of surgery for EDH include-

   a. EDH greater than 30cm3 should be surgically evacuated regardless of patient's GCS.

   b. An EDH less than 30cm3 and with less a 15-mm thickness and with less than a 5- mm midline shift in patients with a GCS score greater than 8 without focal deficit can be managed nonoperatively with serial CT scanning and close neurological observation in a neurosurgical center.

It is strongly recommended that patients with an acute EDH in coma (GCS score < 9) with anisocoria undergo surgical evacuation as soon as possible. There are insufficient data to support one surgical treatment method. However, craniotomy provides a more complete evacuation of haematoma.

2. **Subdural Hematoma:** The indication of surgery for SDH include-

   a. An acute SDH with a thickness > 10 mm or a midline shift > than 5 mm on CT scan should be surgically evacuated, regardless of GCS score.

   b. All patients with acute SDH in coma (GCS score < 9) should undergo ICP monitoring.

   c. A comatose patient with SDH <10-mm thick and a midline shift < than 5 mm should undergo surgical evacuation of the lesion if the GCS score decreased between the time of injury and hospital admission by 2 or more points on the GCS and / or the patient presents with asymmetric or fixed and dilated pupils and / or the ICP exceeds 20 mm Hg.

In patients with acute SDH and indication for surgery, surgical evacuation should be performed as soon as possible. If surgical evacuation of an acute SDH in a comatose patient (GCS < 9) is indicated, it should be performed using a craniotomy with or without bone flap removal and duroplasty. In extreme cases a "burst" lobe may be evident. Such severe hemorrhagic contusions may necessitate resection of brain in the form of a lobectomy. Brain resection requires rapid but careful surgery preserving the middle and anterior cerebral arteries.

3. **Traumatic Parenchymal Lesions:** The indication of surgery for traumatic parenchymal lesions include -

   a. Patients with parenchymal mass lesions and signs of progressive neurological deterioration referable to the lesion, medically refractory intracranial hypertension, or signs of mass effect on CT scan should be treated operatively.

   b. Patients with GCS scores of 6 to 8 with frontal or temporal contusions > 20 cm3 in volume with MLS of at least 5 mm and / or cisternal compression on CT scan, and patients with any lesion greater than 50 cm3 in volume should be treated operatively.

Craniotomy with evacuation of mass lesion is recommended for those patients with focal lesions and the surgical indications listed above. Bifrontal decompressive craniectomy within 48 hours of injury is a treatment option for patients

with diffuse, medically refractory post traumatic cerebral edema and resultant intracranial hypertension. Decompressive procedures, including sub temporal decompression, temporal lobectomy, and hemispheric decompressive craniectomy, are treatment options for patients with refractory intracranial hypertension and diffuse parenchymal injury with clinical and radiographic evidence for impending transtentorial herniation.

4. **Posterior Fossa Mass Lesions:** The indication of surgery for traumatic parenchymal lesions include-

   a. Patients with mass effect on CT scan or with neurological dysfunction or deterioration referable to the lesion should undergo operative intervention. Mass effect on CT scan is defined as distortion, dislocation, or obliteration of the fourth ventricle; compression or loss of visualization of the basal cisterns, or the presence of obstructive hydrocephalus.

   b. Patients with lesions and no significant mass effect on CT scan and without signs of neurological dysfunction may be managed by close observation and serial imaging.

In patients with indications for surgical intervention, evacuation should be performed as soon as possible because these patients can deteriorate rapidly, thus, worsening their prognosis. Suboccipital craniectomy is the predominant method reported for evacuation of posterior fossa mass lesions, and is therefore recommended.

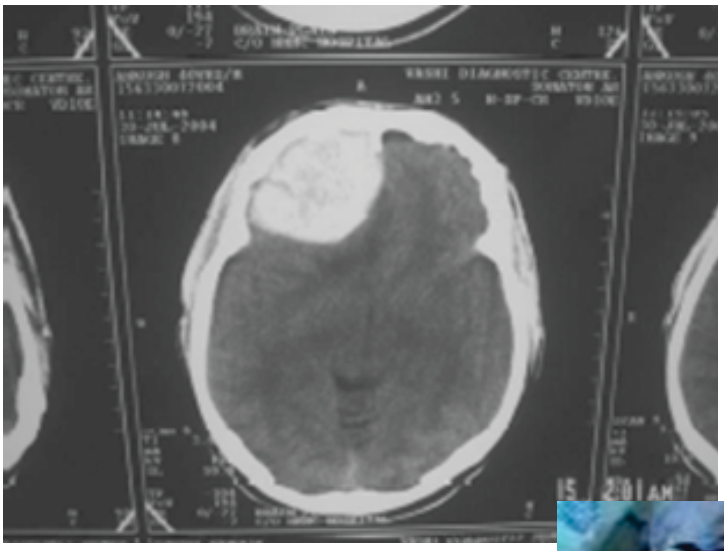


*Figure 3: Computed Tomography brain (axial view) showing a large biconvex hyperdense lesion in the right frontal region suggestive of extradural hematoma.*

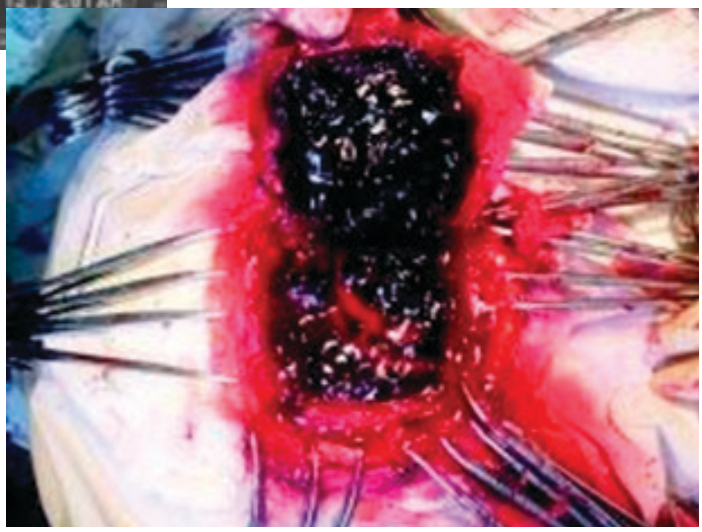


*Figure 4: Intraoperative picture showing large extradural hematoma below the craniotomy flap.*

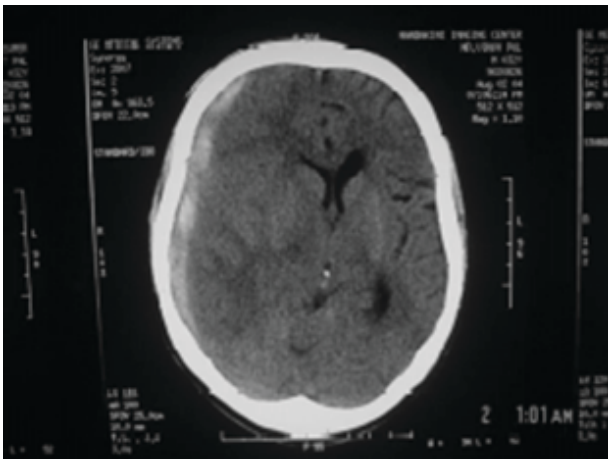


*Figure 5: Computed Tomography brain (axial view) showing a large concavo-convex hyperdense lesion over the right frontoparietal region with mass effect suggestive of subdural hematoma.*

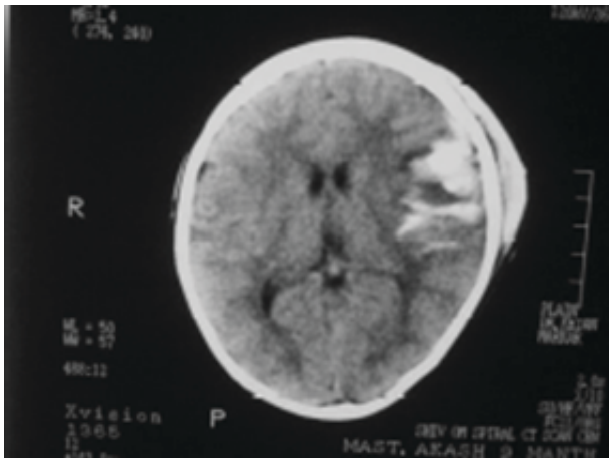


*Figure 6: Computed Tomography brain (axial view) showing a large hyperdense lesion in the left frontal region suggestive of hemorrhagic contusion.*

5. **Cranial Fractures:** The indication of surgery for cranial fractures lesions include-

a. Patients with open (compound) cranial fractures depressed > the thickness of the cranium should undergo operative intervention to prevent infection.

b. Patients with open (compound) depressed cranial fractures may be treated nonoperatively if there is no clinical or radiographic evidence of dural penetration, significant intracranial hematoma, depression greater than 1 cm, frontal sinus involvement, gross cosmetic deformity, wound infection, pneumocephalus, or gross wound contamination.

c. Nonoperative management of closed (simple) depressed cranial fractures is a treatment option.

Early operation is recommended to reduce the incidence of infection. Elevation and debridement is recommended as the surgical method of choice. Primary bone fragment replacement is a surgical option in the absence of wound infection at the time

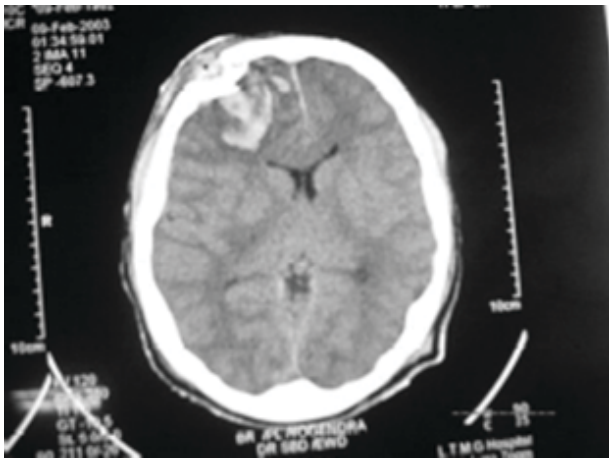


*Figure 7: Computed Tomography brain (axial view) showing a right frontal depressed fracture with underlying contusion.*

of surgery. All management strategies for open (compound) depressed fractures should include antibiotic.

**Speech Affection in Head injury:**

Patients with head injury present with a myriad of problem which include language problems, speech difficulty, cognitive issues and personality disorder depending upon the area of the brain affected. The left hemisphere damage affects language to a greater extent as compared to right hemisphere damage which affects the pragmatics of language.

Individuals with left sided brain damage present with problems of understanding spoken or written language, difficulty in expressive language, alexia, agraphia and naming problems.

An individual with a mild traumatic brain injury may present with poor speech clarity (dysarthria) or even milder being the cognitive issues like affected attention and concentration, difficulty in problem solving issues, and changes in personality which is just a tip of the ice berg.

Management explained in Speech Rehabilitation Chapter.

**Physiotherapy in Head Injury**

A Traumatic Brain Injury (TBI) is a devastating and life changing event for the individual and his or her family. A TBI leads to a much more wider range of conditions than from sustaining any disruption to the vascular supply of the brain making it one of the most challenging conditions for physiotherapists. There is a multitude of issues with these patients as affection for the different parts of brain will be responsible for their own aspects of impairments and secondary impairments. Patients with a moderate to severe TBI require extensive rehabilitation during the different stages of the recovery and physiotherapist will be involved in the most of them depending upon the sensory motor deficit and their recovery. Physical recovery does not always go hand in hand with the cognitive and behavioral recovery. One of the deficits may show very good recovery without any in other during different stages and the durations.

Many traumatic brain injury (TBI) survivors are left with significant disability. The brain is, however, very adaptable and, with the correct physiotherapy input, recovery can take place over a period of years. People often witness a rapid recovery in the first few weeks following a TBI, followed by a slower recovery over the following years.

To gain the maximum recovery, physiotherapy should be continued until patient's full potential has been reached. With the correct physiotherapy input and advice patient has lot of potential to long term improvements regaining as much movement and function over the years.

**Physiotherapy in TBI can help to:**

- prevent or improve the respiratory distress to minimize / prevent secondary brain damage
- prevent the skin, soft tissues, joints dysfunction while unconscious or bed ridden
- reduce muscle spasms, pain and stiffness
- increase strength
- retrain normal patterns of movement
- increase affected arm and leg function
- initiate the ability and do independent ADLs from roll / move in bed / sit to stand and walk
- improve balance and walking
- increase energy levels
- increase independence and quality of life
- decrease risk of falls

**Acute Stage**

The main priority at this stage is to ensure the patient is medically stable. Proactive management at this early stage, of potential secondary complications, will enhance patient outcomes.

At this stage, the goals and anticipated outcomes will be:

- To handle carefully and immediately known or suspected raised ICP
- To treat an unconscious patient as a whole person and not a series of joints and muscles
- To facilitate arousal, increase the level of alertness and physical function
- To reduce the risk of secondary im pairments
- To prevent building up respiratory

secretions and enhance oxygenation of the brain

- To work on the response by stimulating the sensory system
- To work for the normalization of the muscle tone
- To improve joint mobility and con trol
- To provide early family education and support- by means of involvement in aspects of the patient's care, providing time out to discuss the patient's progress and potential outcomes of interventions, advice on support groups and sources of information.

The unconscious patient is best nursed in an intensive care unit as the high levels of care is required. The patient may be on a ventilator, monitoring of ICP may be ongoing; there may be weight bearing precautions and movement restrictions due to musculoskeletal injuries, open wounds or presence of bone fixators. Frequent neurological observations are required to know any deterioration, if any.

**Chest care:**

Hypoxia causing further brain damage is an important cause of neurological deterioration. Cerebral hypoxia may be produced by: airway obstruction from inhalation of blood or vomit or head position; depression of the respiratory drives; associated chest injury, infection or pulmonary edema; and epileptic fits.

Patients who are managing to maintain their own airways and adequate oxygenation may not require intubation and mechanical ventilation. They are nursed in a position which allows the head to be raised at $30^{o}$ and inside lying to prevent inhalation of secretions. Regular turning will prevent consolidation and collapse of lungs and the pressure sores. Positioning and turning may be difficult, if there are associated injuries which require splinting, such as plaster casts or traction.

If the patient is unable to maintain his own airway, a cuffed endotracheal (ET) tube is required. This may be sufficient to obtain sufficient oxygenation and remove any retained secretions to stop any further deterioration. Any evidence of respiratory inadequacy need of artificial ventilation (IPPV) is required.

It is important to keep the chest clear of secretions as patchy collapse with sub clinical hypoxia and hypercarbia is probably more damaging to the brain than any other factor. Due to the traumatic brain a small rise in IC volume may cause a dramatic rise in ICP. Various physiotherapeutic procedures for the chest care such as postural drainage, shaking, bag squeezing, prolonged suction and coughing may precipitate the raise in ICP. Therefore, the patient must be protected during chest physiotherapy; effective treatment can be carried out without provoking large increases in ICP. This can be achieved by drug therapy for sedation, analgesia and muscle paralysis.

This is best carried out to coincide with turns and before bolus feeds, a continuous feed should be switched off. The chest is vibrated on the expiratory phase of breathing. Tipping the patient with his head down is not advisable due to the risk of elevating ICP. When suction is required to remove secretions, nasal suction is contraindicated if CSF Rhinorrhea is present: this indicates a CSF leakage through skull fracture with a risk of ascending infection.

The frequency of treatment depends on the individual patient, but even if there are no obvious lung problems, treatment is given to prevent the unnecessary complications of retained secretions.

Techniques of shaking and vibration are best used while the patient is being ventilated by bag squeezing, i.e. off the ventilator. Careful suction via the ET tube should clear secretions.

Patients having controlled IPPV usually need continuous sedation using anesthetic agents and analgesics. A bolus dose of sedation is given prior to physiotherapy.

Total treatment time is a crucial factor and should be kept at minimum, shorter sessions, more frequently should be given as per the requirement. Suction, in particular, is a relatively dangerous procedure because of its effect on ICP.

**General care:**

Handling these patients is often made more difficult from spasticity with the release of primitive neuromuscular activity such as reflex patterns. Primitive postures may include those associated with decorticate (abnormal flexor response) or decerebrate (abnormal extension response) rigidity.

Patients with TBI are at a high risk of developing contractures owing to prolonged periods of immobilization and abnormal reflexing posturing. Techniques to preserve mobility and inhibit primitive responses are employed as far as possible. These, combined with careful positioning after treatment sessions and every turn, should help reduce the problems to which hypertonicity leads at a later stage. Never assume that these patients are unaware of what is happening around them. Talking to them, telling them what you are doing and why, asking them to try and help, provide important stimuli from other sensory pathways along with the stimuli from movement and handling. As consciousness returns, the clinical features resulting from the head injury are evidently seen, but the predominant problem of spasticity will be evident early on.

Unless there are reasons to delay the start of an active physiotherapy, associated injuries such as limb fractures, chest injuries, patients should be introduced to changes of position out of bed and movement early on. Aim for normal sitting posture, avoiding tipping chairs.

The unconscious patients will also require to be treated for their oromotor impairments for oral hygiene, sensory motor training, chewing, swallowing, as well as speech therapy.

By Passive Range of Motion(PROM) exercises, joint mobility and integrity improvement can be achieved. During PROM exercises of upper extremities, care should be taken to mobilize the scapula, otherwise, impingement of glenoid fossa becomes the cause of the pain in shoulder. PROM exercises when performed, forceful or aggressive movements should be avoided to avoid heterotopic ossification. It is more prone in the proximal joints and becomes the cause of pain and hypomobility. Proper positioning will help in the reduction of secondary impairments such as contractures, bedsores, pneumonia, deep vein thrombosis, which not only hamper the progress but may lead to life threatening consequences. Good positioning will help in preventing skin breakdown, contractures, improve pulmonary hygiene and circulation. In bed, head should be positioned in neutral to prevent the neck contractures and to lessen the effects of tonic neck reflexes. The hips and knees should be slightly flexed. Turning frequently will help in preventing the skin breakdown and pneumonia. Caregivers should be told about the turning the patient every 2 hours when in bed. Specialized air mattresses with electrical / pneumatic pressure relieving system are useful for the alternate weight relieving from the load bearing body parts in the bed and avoiding the pressure sores. Splints help in positioning the head, the legs and the feet in these bedridden patients.

Stimulation of the sensory organs: Sensory stimulation is an intervention used in an attempt to increase the level of arousal and elicit movement in individuals in a coma or persistent vegetative state. It is proposed that, by providing stimulation in a controlled multisensory manner, with a combination of stimulation and rest, the reticular activating system may be stimulated causing a general increase in arousal. The following sensory systems are systematically stimulated: auditory, olfactory, gustatory, visual, tactile, kinesthetic and vestibular. During these types of interventions, the patient must

be closely monitored for subtle responses such as eye movements, facial grimacing, and changes in posture, head turning, or vocalization. Overstimulation may lead to the accommodation making all efforts useless. Patient should be subjected for a single type of stimulus as multi stimuli may lead to the confusion for the patient. During this Coma stimulation, physiotherapist must look for the warning signs like flushing, perspiring, increase in respiratory rate, agitation, eye closure, decreased level of arousal or increase in muscle tone. If any of these symptoms are seen, coma stimulation therapy should be immediately stopped. For the motor response, therapist should communicate with the patient by all the possible means.

The patients who are slow to recover and minimally conscious and who remain in this stage of recovery for a longer time duration are often placed in long term rehabilitation center / nursing home.

Family members and caregivers of patients who have experienced a TBI play a critical role in the recovery process. Family members commonly experience a high level of stress related to worries about the future, less free time, and increased conflict. Family education is an important component of the management. The goal of patient and family education is to teach the family about the stages of recovery and what can be expected in the future. By becoming informed, the family may not be so helpless. The family can also become involved in performing ROM exercises, positioning, and sensory stimulation. Although it is difficult to predict long term outcome at these early levels of recovery, families should be informed of possible outcomes. The therapist should be realistic but provide hope for the family. It is often beneficial to have a medical social worker consult with the family to provide support and guidance. When the patient begins to exhibit improved mobility skills, may lack the insight to recognize that he or she may not yet be safe to ambulate or transfer alone. The patient should be educated in how to best compensate for the residual impairments or disabilities. In severe cases, counseling will be required for the patient and the family members by a medical social worker or a neuropsychologist. Family members should learn how to assist the patient with functional mobility including bed mobility, transfers, ambulation and wheelchair mobility skills. They should be trained for the proper body mechanics when assisting the patients to avoid the injury to themselves or the patient. They should be taught the home exercises for the strengthening and PROM exercises.

**Sub acute stage**

**At this stage, the goals and anticipated outcomes would be:**

- To preserve the integrity of the neuro musculoskeletal system, thereby preventing or minimizing adaptive muscle shortening and contractures
- To manage the effects of abnormal Tone and spasticity
- To provide an appropriate level of sensory stimulation
- To improve the motor control, postural control and functional level

Once the acute phase gets over and the patient with TBI is medically stable, continuation of the care can be in a variety of set ups. Patients with minimally conscious state or in coma may get the therapy in hospital or a long-term care nursing home. Patients who start recovering from coma with moderate to severe cognitive, behavioral, and physical impairments often continue rehabilitation in either in-patient / out-patient rehabilitation center or a multispecialty hospital. The cognitive abilities and behavioral presentation of individuals with TBI will have an impact upon their level of function. It is therefore vital that these elements be assessed and taken into account when agreeing goals for interventions. Access to a neuropsychologist, who can assess and advise on the most effective way to deal with these

factors when planning treatment, is invaluable.

As the patient begins to emerge from coma, he or she often experiences a period of acute post traumatic agitation. The confusion, amnesia and disorientation often result in agitation, aggression, noncompliance and combative behavior. The patient may be markedly agitated and prone to emotional outbursts ranging from verbally acting out to physically attempting to hurt themselves or others or attempt sexually inappropriate behaviors. The patient is always confused and with the short and long term poor memory. Attention span is decreased, and so, get easily distracted.

Examination at this stage would be difficult as the patient is often uncooperative. The therapist must utilize observational skills by seeing the functional mobility, balance both in sitting and standing (if possible), ROM, strength, motor control, tone, sensation and reflexes. Patient's cognitive abilities also have to be examined like orientation, attention span, memory, insight, safety awareness and alertness. Neuropsychologist's help will be required to manage the patient's agitated behavior and help in setting up behavioral modification technique such as positive reinforcement using a reward system, redirection and compliance training which will help in managing inappropriate behaviors and improving in therapy. Severe cases may require medications too.

Due to the confused state of the patient, it is very important to maintain the consistency in the talking as well as in dealing with the inappropriate behaviors by all the members of the team as well as the family members. It is important to maintain the familiarity to the patient by keeping the same timing, place of the treatment and the same therapist. Patient should be frequently provided orientation information.

At this stage, patient may require one to one staff supervision and assistance throughout the day.

Family support required at this stage ranges from the supervision due to cognitive deficits to the physical assistance in all the ADLs due to motor deficits. For discharge from the hospital, this factor of strong family support and ability to help is as important as the health status of the patient.

The therapist will be primarily responsible to improve the motor and postural control which will be a prerequisite to improve the functional level of the patient. Maintaining the normal physiological length of the soft tissues is very important to prevent or minimizing adaptive shortening and contractures. This can be achieved by passive movements and correct positioning of the patient. Utmost care has to be taken to avoid the overstimulation of the tissues and the joints. Taking care of the painful joints, especially the shoulder is very important at this stage as this pain if persists, become the cause of the inability to get the range even passively. If it prolongs for a longer time, to get the motor control over the movements of the upper extremities will be delayed and sometimes terminal ranges are permanently affected. Prophylactic splinting to the hypertonic muscular body parts helps to prevent the shortening or contractures. Worst cases of hypertonicity may require the medications.

Physical and motor problems of abnormal tone in the form of spasticity or rigidity is a common problem throughout the recovery phase or thereafter for many patients with TBI. More than 80% patients with TBI develop contractures due to the spastic hypertonicity. Increased tone may be responsible for the difficulty in personal hygiene, transfers or may be a source of pain or pressure sores. Increased tone in LEs may help the patient in weight bearing and bed transfers or make it easier for the caregiver to transfers.

Physiotherapists use two basic treatment strategies as compensatory (to improve functional skills by compensating for the lost ability) and restorative (to restore the normal use of the affected part) approach.

Many therapeutic skills are available with Physiotherapist to deal with the abnormal tone and in turn to avoid its adverse effects.

Basics of spasticity management are therapeutic stretching-sustained and gentle, strengthening exercises with adjunctive modalities and functional retraining. PROM and selective strengthening of the antagonist muscles can help to decrease spasticity. Positioning is an important adjunct in the management of tone abnormalities. Maintaining the head and neck in a neutral position is important for minimizing the effects that primitive postures may have in increasing tone. Keeping the whole body in proper alignment is also important.

i) Cryotherapy or air splints can also be used to reduce the hypertonia, though their effect is temporary.

ii) Serial casting helps in decreasing hypertonia. Serial casting is often used for plantar flexors or biceps contractures resulting due to either increased tone or prolonged shortening of the muscle. Progressive maximal stretched position is achieved for the cast to maintain for a week before trying to get the new more stretched position is tried for the casting. PROM exercises are continued to have the new increased ROM. There should be constant check on the skin coming in contact with the cast as it can breakdown if the patient has sensory, communication or behavioral impairments.

iii) Various medications either systemic (Baclofen) or local (phenol or Botulinum toxin) can also be used to reduce the hypertonia but their use should be weighed in comparison with the side effects they can produce in a particular patient

iv) Increased tone in lower limbs that requires more than manual techniques to inhibit and maintain range of movement may benefit from the application of below knee weight bearing plasters. This is a skilled task and care must be taken to obtain correct alignment of foot and ankle.

v) Increased tone in lower limbs that requires more than manual techniques to inhibit and maintain range of movement may benefit from the application of below knee weight bearing plasters. This is a skilled task and care must be taken to obtain correct alignment of foot and ankle.

The tilt table can be used for periods of standing in conjunction with standing with necessary assistance and standing transfers. Lack of head control can impede progress to activities in sitting and standing.

Many patients with TBI get affected with ataxia, who will need the coordination and balance therapy. Some of these may require a walking aids either temporary or for a longer time. Sensory disturbance will involve perceptual problems including vision and hearing, apraxia and agnosia. Emotional and intellectual disturbance will involve memory and behavioral disorders. For these patients co treatments will be beneficial if done along with Speech therapy, Occupational therapy and Neuro psychotherapy.

Upright sitting is very important to work towards the treatment goals for the early levels of recovery. Upright position is vital for the proper functioning of many organs like, stimulation of bowel movements and bladder emptying, improved ventilation due to moving down of abdominal contents leading to redistribution of air flow to basal lobes and changing perfusion / ventilation ratio. As soon as medically stable, the patient should be transferred to a sitting position and out of bed to a wheelchair / chair. All precautions should be observed. The head should be properly supported, as the

patient may have very inadequate neck and head control to maintain an upright posture without support. Often, it is beneficial to perform co treatments with an Occupational therapist when first sitting and transferring the patient. Use of a tilt table is very useful as it allows early weight bearing through lower extremities. The upright position in sitting by the side of the bed, in a wheel chair or standing on a tilt table improves overall level of alertness. Sitting training promotes movement and learning by allowing the patient's body in functional position to do the tasks. Assisted movements provide tactile, proprioceptive and kinesthetic stimulation while training to perform a ADL task.

Constant efforts are required by the physiotherapist to improve the motor control and motor learning. Neuro physiotherapeutic techniques can be used to initiate, progress, improve the sensory motor control and finally helping the patient to get back to the as normal life as possible depending upon the severity of the lesion, personal as well as external contextual factors. Due to the newer understandings and the theories of the motor learning and the practices, many new skills are developing but still most of the older skills also hold true today and specifically in the initial and subacute phase, they are most useful. The therapist should work as per the patient's physical level of function and attempt to improve endurance. Progressing to more challenging skills which would require new learning, should be as per the patient's capacity for new learning.

Depending upon the skills and experience of the therapist, clinical conditions and the co-morbidity of the patient any or many of the followings are used.

i) **Developmental sequence:** Developmental postures are utilized to facilitate and help restore movement and functional mobility. Once, the patient gets the fair control over the different parts of the body as per the involvement due to their cerebral or the cerebellar trauma, postural training should be started on the basis of developmental sequence and the biomechanical stability basis. This may start with the supine-side turning as a log first and then by girdle initiation, cephalocaudally or caudocephalic-prone lying- coming on the forearm and then on hands- all four with elbow / forearm loading to hands loading- quadruped position with ability to relieve the load of one or two extremities (crossed)-independent sitting -kneel sitting-half kneel sitting-kneel standing- sitting to standing- kneel standing to standing-squatting -walking and then the training for the dynamic postures. All the static postures first to be trained in assisted and guarded situations. Once active control is gained, then they are made more and more difficult so that in adverse situations they will be mastered. This can be against the resistance or against the smaller base of support or against the unstable base. The gymnastic ball has specific qualities which can be used to gain mobility, stability and postural control. Other injuries, if any, than TBI also should be taken care of during training all these postures. As and when, the patient masters the posture, ability should be used for the related possible activities performance first in the therapeutic area and then in life situations.

ii) **Proprioceptive Neuromuscular Facilitation (PNF):** This is a good technique for the initiation of mass movements of the proximal girdles to the distal components as they are closer to the activities of the daily life. These can be first initiated passively by the therapist with the commands till there is response and then assisted to learn actively by the patient. Full range move-

ments along with the strength and endurance are achieved. All these activities should be trained for the near normal equilibrium responses. Sufficient practice will give this maximum perfection to which the patient can reach a level and with the maximal endurance. Once this is met in the treatment area, this has to work equally good in the actual life situational areas, where patient will have to be multi responsive due to the environmental demands depending upon the crowd, architectural barriers and the vehicular traffic.

iii) **Neuro Developmental Therapy-(NDT):** This is based on Ms. Bobath's core ideas of rigorous observation of posture and movement, careful alignment of body segments and joint position before asking patients to move, constant analysis of how the patient moved throughout the entire treatment session, the contribution of the sensory systems to movement, family involvement in treatment, and home programming. Task oriented approach is currently advocated for this locomotor training. The task is broken down in subtasks and is assessed on the basis of base of support, allignment, sequencing, timing, stability and mobility. Keeping the impairments in mind environment is biased to bring about better recruitment of the affected area.

iv) **Body Weight Support (BWS) and a treadmill:** Suspending the patient in a parachute like overhead harness which allows partial relief / support of the body weight. Therapist needs to support the patient by pelvis / trunk for the weight shifting and the forward advancement of the LEs. This can be done in parallel bars, with walker or with the treadmill. Training as well as corrections can be done in this system in each phase of the gait cycle. Difficulty can be progressively increased by decreasing the harness and physiotherapist's support and increasing the speed of walking or treadmill as the patient's ability improves.

v) **Constraint Induced Movement Therapy(CI/ CIMT):** It involves promoting the use of the most affected UE for up to 90% of waking hours and reducing the use of the lesser affected UE. Intensive, task oriented training is provided for the affected UE for up to 6 hours per day over 2-3 weeks period

vi) **Modified Constraint Induced Movement Therapy(MCIMT):** A mesh is worn to the unaffected hand for gentle restraint of the activities for several hours for 10 weeks period. Training will be same as CIMT.

vii) **Hand Arm Bimanual Intensive Training (HABIT):** This is also a form of intensive task training, but the more emphasis is on the bilateral hand manipulations in ADLs. Unaffected hand may play a major role at the initial phase, but, emphasis should be given on the more and more equal participation of the affected hand during the tasks.

Patient should be started training the simple daily functional activities like bed activities, feeding and the ambulation before expecting him / her to learn new skills. Use of some audiovisual aids like charts / pictures / photos will help the person to learn new skills and the progress each day. Patient will be required to give a recall each day of the previous days teaching as carryover will be difficult in many patients.

It is important for the therapist to perceive the patient's emotions and behavior, so that he / she should be able to make feel the patient safe and secured. Even the family members' and caregiver's attitude towards the patient will have an effect on

the behavior of the patient. Patient will not be in a position to understand the other people's views and most likely egocentricity may be present. Due to the limited attention span of the patient and the easy distractibility, concentration on the activities training is going to be limited for early days of the treatment.

**Chronic stage**

At this stage the main goals of physiotherapy interventions are to:

- Encourage the return of active movement. that carries over into function
- Prevent secondary deformities
- Prevent unnecessary and potentially damaging compensatory movement. strategies
- Maximize respiratory function
- Encourage social and vocational reintegration
- Provide advice to the family, caregivers and other members of the team on aspects of the patient's management
- To educate family members on patient's condition, management goals and outcomes

At this stage, when patient shows the signs of improvement in the consciousness and the awareness or may be only in the sensory motor components with respiratory independence, patient will be most of the times either in half way rehabilitation center or at home and comes for the therapy on outpatient basis.

The examination should be done at this level to access the status on following points:

- attention and cognition
- cranial nerve integrity
- balance
- gait
- joint mobility
- motor control
- muscle strength
- ROM
- Reflexes
- ADL skills and sensory integrity
- Functional abilities of the patients (better in closed environments than an area of multiple distractions)
- Gait is generally examined by Observational Gait Analysis to know the deviations and asymmetries so the impairments can be found to treat the causative factors. Gait speed and the endurance can be measured by 10 meter walk or 6 minute walk test
- Their motor control and functional status including the ability to perform ADL, transfers, bed mobility, stairs, and locomotion can be measured by using Functional Independence Measure (FIM) or Fugl-Meyer Assessment of Motor Recovery (FAM) or Chedoke -MacMaster Stroke Assessment.

For the task performance training, it is to be seen about the balance during the performance, initiation and completion, consistency, energy required, shift of the body weight and maintenance of body alignment.

Those having cognitive and behavioral deficits will have difficulty in reacquisition of the motor skills due to their mental as well as physical fatigue during treatment sessions. That can be known by the signs like increased irritability, decreased attention and concentration, deterioration in the performance of physical skills and delayed initiation. Treatment sessions with sufficient rest periods will avoid the mental and physical fatigue and will maximize the motor relearning.

TBI patients will have many different physical, cognitive and emotional impairments making the therapist to have an individual approach as per the patient's capabilities and the difficulties due to the

uneven recovery of these different aspects of the brain functioning. For every patient, the emphasis is given for the initiating, ability to get it actively and perfecting the motor tasks by making them progressively increasingly difficult and complex to blend them in the daily activities successfully.
TBI patients have a wide variety of impairments and functional limitations, they may be helped to compensate for the lost function by giving them some form of adaptive device. It depends upon the predicted probable residual disability as per the opinion of the rehabilitation team, to decide how much help should be given. Most severe patient who may not be able to make to walk again will require a wheel chair. Depending upon his or her regained cognitive skills, wheel chair can be advanced with the computer-based controls powered one to the simple manual one which may be operated by the patient or the caregiver.

Therapist should have more options of the activities for the training, so after few days, different combinations and the options to the patient can be given for breaking the monotonicity as well as to give choice to the patient.

Prior to discharge from a comprehensive interdisciplinary day treatment setting, cognitive, behavioral, emotional and psychological issues have to be tackled before community re-entry and return to school / work. Skills in judgment, problem solving, planning, self-awareness, health and wellness and social interaction should be taught by the respective professionals with lesser and lesser supervision needs. Once the patient gets the insight into his / her own strength and weaknesses, they should be involved in decision making for their working towards reintegration back to home and community. Group treatment sessions and Support groups are crucial for the patient to learn how to function in society with their present abilities and limitations. Adaptations are required most of the times by the family, work place colleagues or schoolmates to accommodate the patient.

**Occupational Therapy Management**

The rehabilitation team including occupational therapists uses several objective assessments for early detection of neuro medical complications and to monitor changes in function and response to pharmacological, environmental and behavioral interventions.

**RANCHO LOS AMIGOS LEVELS OF COGNITIVE FUNCTIONING SCALE**

The Rancho Los Amigos Levels of Cognitive Function Scale uses behavioral observations to categorize a client's level of cognitive function. It helps clinicians to communicate about a client's level of cognitive function among themselves and with families and to develop appropriate rehabilitation strategies. The first three



Fig 8. Active movements to improve shoulder movements



Fig 9. Turning and rolling with assistance. To teach to do the mobility independently



Fig 10.Prone on elbows: To improve weight bearing through shoulder and to improve head, neck and upper back control



Fig 11. Quadruped. To improve upper trunk, lower trunk, lower extremity and head/neck control, improve shoulder strength, improves trunk control and weight bearing through hips



Fig 12. Kneeling. Improves hip stabilizers strength, improves balance reactions, improve upper trunk, lower trunk, lower extremity and head/neck control



Fig 13. Sitting with elbow splints. To improve upper trunk, lower trunk, lower extremity and head/neck control, sitting balance and weight bearing on hands.



Fig 14 Exercises on swiss ball on prone lying to improve head, neck and back extension



Fig 15 Standing on standing board with full support to improve weight bearing in lower extremities and to improve the physiological benefits(absorption of calcium and other minerals)

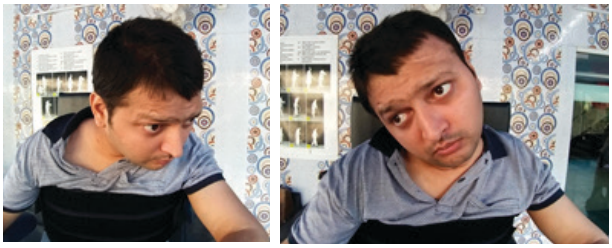


Fig 16 to 17. Active range of motion exercises for the neck, from left to right and back





Figure 18 and 19: Hand activities to improve voluntary control





Figure 20 and 21: Pushups in prone lying: Improves weight bearing in upper extremity and increases muscle strength and sustainance of upper extremity muscles




Figure 22 and 23: Back extension in prone lying to strengthen back extensor muscles





Figure 23, 24 and 25: Walking backward to improve voluntary control of knee flexor muscles




Figure 26 and 27: High stepping to improve hip and knee flexion in standing



Figure 28, 29 and 30: Gait training using obstacle: to improve hip and knee flexion while walking



Figure 31: Exercises to to improve shoulder strength and sustainance





Figure 32 and 33: Pushing activities to improve upper limb strength and sustainance

levels of the Rancho Los Amigos Levels of Cognitive Functioning Scale describe the response to stimulation and the environment of clients emerging from a coma.

**Rancho Los Amigos – Revised**

Level I – No Response: Total Assistance (Complete absence of observable change in behaviour when presented visual, auditory, tactile, proprioceptive, vestibular or painful stimuli).
Level II – Generalized Response: Total Assistance (Demonstrates generalized reflex response, generalized physiological changes, gross body movement and/or not purposeful vocalization)
Level III – Localized Response: Total Assistance (Response directly related to a type of stimulus, but maybe inconsistent or delayed)
Level IV – Confused/Agitated: Maximal Assistance (response heightened, severely confused, maybe aggressive, Absent short-term memory).
Level V – Confused, Inappropriate Non-Agitated: Maximal Assistance (alert, not agitated but may wander randomly, some response to simple commands, but confusion with more complex commands; high level of distractibility)
Level VI – Confused, Appropriate: Moderate Assistance (response more goal-directed but cues necessary, able to attend to highly familiar tasks in a non-distracting environment)
Level VII – Automatic, Appropriate: Minimal Assistance for Daily Living Skills (response robotlike, judgment and problem-solving lacking)
Level VIII – Purposeful, Appropriate: Stand-By Assistance (response adequate to familiar tasks, subtle impairments require standby assistance with acknowledging other people's needs and perspectives, modifying plans)
Level IX – Purposeful, Appropriate: Stand-By Assistance on Request (responds effectively to familiar situations but generally needs cues to anticipate problems and adjust performance; low frustration tolerance possible)
Level X – Purposeful, Appropriate: Modified Independent (responds adequately to multiple tasks but may need more time or periodic breaks; independently employs cognitive compensatory strategies and adjusts tasks as needed)

**BEHAVIOUR-BASED ASSESSMENT INSTRUMENTS FOR ALTERED STATES OF CONSCIOUSNESS**

Coma/Near Coma Scale: Measures small clinical changes in clients with severe traumatic brain injuries (TBI) and non-traumatic brain injuries who function at levels characteristic of near-vegetative or vegetative states; consists of 8 parameters, each with a specified number of stimuli and trials. Based on the numerical scoring system, the client falls into 1 of 5 categories: no coma, near coma, moderate coma, marked coma, extreme coma.

Western Neuro Sensory Stimulation Profile: Assesses cognitive function in severely impaired adults (Rancho levels II-IV) and monitors changes in non-comatose clients who are slow to recover; consists of 32 items related to arousal, attention, response to stimuli, and expressive communication and results in a profile of 6 subscales that summarizes individual patterns of responses.

Coma Recovery Scale-revised: Detects subtle changes in neurobehavioral status; predicts outcome in acute rehabilitation clients with severe alterations in consciousness (Rancho levels I–IV); consists of 23 items in 6 areas: auditory, visual, motor, oromotor, communication, and arousal functions. Responses are criterion-referenced: specific stimuli are administered to elicit specific responses.

Disability Rating Scale (for TBI): Single instrument to provide quantitative information on recovery from severe TBI from coma to community; consists of 8 items in 4 categories: consciousness (eye-opening,

verbal response, motor response); cognitive ability (to handle self-care functions); physical dependence upon others; and psychosocial adaptability for work, housework, or school. Each area of functioning is rated on a scale of 0 to either 3/5 (max. score =29, extreme vegetative state; min. score= 0, no disability) with the highest score representing a higher level of disability.

Agitated Behavior Scale: Allows objective measurement of agitation, particularly as a serial assessment to evaluate the effectiveness of interventions to reduce agitation; consists of 14 items; total score reflects overall agitation, with subscales specific to disinhibition, aggression, and lability.

**INTERVENTION FOR THE INDIVIDUALS WITH SEVERE ALTERATION OF CONSCIOUSNESS**

Rehabilitation for clients with severe alterations in consciousness (Rancho level 1, 2 and 3) aims to foster alertness and behavioural responsiveness and to prevent complications associated with prolonged immobilization. All stimuli should be structured and broken down into simple steps and commands, giving sufficient time for a response due to delayed cognitive processing. Intervention at this stage can be grouped into six areas: sensory stimulation, bed positioning, casting or splinting, wheelchair positioning, management of dysphasia, and emotional and behavioral management, as well as family and caregiver education. Interventions may occur simultaneously to optimize progress. It is important to incorporate close family members and friends into sessions, to enhance response.

**MULTISENSORY STIMULATION:**

Sensory stimulation in a controlled, multisensory manner, with a balance of stimulation and rest, stimulates the reticular activating system and may cause a general increase in arousal. Stimuli are presented in a highly structured, consistent and meaningful manner, typically one sensory modality at a time. During this type of intervention, the client should be closely monitored for any changes in behavior. The client is told what the therapist is doing and often given an instruction specific to the desired response. Minimize competing visual and auditory stimuli by turning off the television, closing the door, or pulling the drape. Give simple, clear verbal feedback on every response elicited.
The most effective types of sensory stimulants are those that have personal meaning to the client.

Here are examples of how various types of stimuli are presented

- Tactile stimulation focuses on the qualities of touch, pressure, and temperature .
- Example: Verbal orientation to body parts being washed or moved during bathing and dressing. Rub the client's skin with items of various textures or temperatures and use a firm or moving touch on the client's limbs with verbal cues to orient the client to his or her body. Performing a passive range of motion provides both tactile and kinesthetic stimulation.
- Provide gentle kinesthetic, proprioceptive, vestibular stimuli by changing body position
- Example: rolling from supine or side-lying to sitting, sitting to side-lying, tilting the bed to sitting, moving head and neck, standing in coma and minimally conscious clients using electric standing frames or tilt tables as the optimal means of enhancing arousal and awareness of the environment. The client's reaction must be closely monitored during and after all stimulation, with careful attention to desired changes in heart rate, intracranial pressure (ICP) and blood pressure. Kinesthetic, proprioceptive, vestib-

ular stimulation may be contraindicated in clients with a tracheostomy, elevated ICP or seizures.

- Stimulate the olfactory sense
- Example: saturated cotton balls or a sniff bottle are held close to but not touching the client's nose for 2–5 seconds. Using noxious or pleasant odours, such as spices, almond, vanilla, banana, lemon, perfume, coffee, or other smells familiar to the client. Avoid odours with fumes, such as ammonia and artificial vanilla, because they irritate cranial nerve V (the Trigeminal nerve). This stimulation may be most effective when it is done before the client is fed. Olfactory stimulation may not elicit a response if injured.
- Auditory stimulation
- Examples: The therapist uses clear, firm speech, often presenting information to the client regarding his or her circumstances and whereabouts. Eliminate other noises as much as possible so that voice commands or selected auditory stimuli for motor responses are the most prevalent auditory input. Tapes of favourite music or familiar voices, bells, loud alerting noises such as clapping hands, direct conversation to the client, verbal commands, explanations, and feedback may be used as an auditory stimulus
- Visual stimulation
- Example: Brightly coloured objects, a mobile over the bed, mirrors, coloured lights, a flashlight in a darkened room, or pictures of family and friends of the client to elicit attention, focus, and visual tracking. Environmental changes from the bed to the therapy room or from indoors to outdoors are important sources of visual stimulation.

## POSITIONING

Occupational therapists collaborate with other members of the rehabilitation team to optimize positioning for normalising muscle tone and ultimately affecting motor performance. Relationships exist between therapeutic positioning, multisystemic stability and prevention of secondary complications after TBI. Some typical abnormal positions seen in clients with severe traumatic brain injuries include abnormally forward head, protracted and forward tipped scapulae with or without elevation, posterior pelvic tilt with unilateral retraction and/or elevation, severe trunk tightness with lack of trunk dissociation from neck and head, hip flexor and adductor tightness, and foot plantarflexion and inversion.

### Bed Positioning

Proper bed positioning is critical in the early stages of TBI as the client spends a lot of time in bed. It is important to prevent pressure sores, facilitate normal muscle tone, and prevent loss of pelvic and trunk mobility. If the client exhibits abnormal tone or posturing, a side-lying or semi-prone position is preferable. This position assists in normalizing tone and providing sensory input. Lying supine triggers an extensor response.

In side-lying, the head should rest on a small pillow and neutral alignment with the trunk; the upper extremity underneath should be in scapular protraction and humeral external rotation, elbow flexed; the top upper extremity should be positioned with scapular protraction, slight shoulder flexion, resting on a pillow to avoid horizontal adduction, elbow extended; wrists should be in extension, and cones should be placed in the hands to decrease spasticity and to maintain thumb web spaces. A pillow should be placed between the knees to decrease hip internal rotation and adduction. The lower leg hip and knee are flexed slightly. Elongation of the lower side of the trunk between the shoulder and

pelvis is desirable. A side-lying trunk position may have to be maintained by a pillow or sandbag behind the back and shoulder. Footboards should be avoided because they elicit extensor thrust.

If the client must be supine, a small pillow is used under the head to flex slightly. Small pillows are placed under the scapulae to protract them, shoulders are positioned in slight abduction and external rotation, elbows are extended, and cones or finger spreaders position the fingers. If the pelvis is retracted on one side, a small folded towel is placed behind it, and that leg is positioned in neutral rotation. A small towel roll placed under the distal thigh just above the knee for slight knee flexion. A semi-recumbent bed positioning is recommended (with head-of-bed angle maintained at 45°) to minimize the risk of aspiration and pneumonia for clients on mechanical ventilators.

**Wheelchair**

Early and correct upright positioning in a wheelchair helps to prevent skin breakdown and joint contractures, facilitate normal muscle tone, facilitate arousal by stimulating the visual and vestibular systems, providing normal proprioceptive input, inhibit primitive reflexes, increase sitting tolerance, enhance respiratory and swallowing function.

Wheelchair positioning should begin at the pelvis. Poor hip placement adversely alters trunk and head alignment and influences tone in the extremities. The pelvis should be in a neutral position or have a slight anterior tilt and should be symmetrical with equal weight bearing on both buttocks. Solid seat and back inserts are necessary to prevent posterior pelvic tilt. If necessary, a small, lumbar roll is placed above the pelvis and below the scapula to facilitate anterior pelvic tilt.

The back of the chair may be reclined 10–15° to position the trunk and head more appropriately. The seat belt should come from the corners of the seat at a 45° angle and fastened over the lower pelvis and hip area to help maintain the anterior pelvic tilt and equal weight bearing through the buttocks.

Hip flexion of 90° can be achieved by a wedge cushion with the high end at the distal thigh. If the seat belt cannot keep the patient from scooting forward in the chair, a removable padded bar may have to be placed close to the pelvis and femurs. The trunk, positioned next, should be symmetrical and in the midline, with shoulders over the pelvis in sagittal, frontal, and horizontal planes. Lateral trunk supports can be used to decrease lateral trunk flexion.

A harness, shoulder straps, or a chest strap may be necessary to prevent forward trunk flexion. An additional seat belt can also be placed horizontally across the anterior superior iliac crests to provide backward pressure. Knees and ankles are flexed to 90°, heels are slightly behind the knees in sitting, feet are in neutral pronation supination and inversion-eversion. The footplate should be large enough to support the whole foot.

Ankle straps are avoided because they encourage plantar flexion in some patients; a foot wedge, heel loops, an insert behind the foot, special shoes, toe guards, or a combination of these may help decrease abnormal tone and in achieving weight bearing on the heel.

The ideal upper extremity position is a neutral scapular elevation or depression with the scapulae in slight protraction, slight shoulder external rotation, and slight flexion and abduction. Elbows are in comfortable flexion, and the forearm is in pronation. The wrists are in neutral flexion-extension and neutral ulnar-radial deviation; the fingers are relaxed, and the thumb is radially abducted. A lapboard is positioned at a proper height to allow good upper extremity weight-bearing and should be large enough to accommodate the whole

arm.

The head should be in the midline, with cervical elongation and the chin tucked in slightly. The position of the patient's shoulders and upper trunk influences head position. Posterior head support must be placed in the suboccipital area to prevent excessive extension, lateral flexion or rotation.

## PASSIVE RANGE OF MOTION

Passive range of motion (PROM) can be difficult when muscle tone is increased. Inhibitory movements are performed slowly, holding the stretch until muscles relax. Sudden stretch and inappropriate stimulation and handling should be avoided. Scapular mobility should be addressed before upper extremity PROM to free the scapula and facilitate normal scapular and humeral movement during the rest of the PROM. PROM within the limits of pain and positioning helps to minimize contractures from heterotopic ossification.

## SPLINTING AND CASTING

Splinting or casting may be used 1) to decrease abnormal tone 2) increase the patient's functional movement (3) to prevent soft tissue contractures. Splints should be carefully monitored when used with severely spastic or posturing patients, as they can aggravate abnormal tone and rapidly create pressure areas. With severe spasticity in the elbow, wrist, knee, ankle, or foot, plaster or fiberglass serial casting may be indicated (16). Casts are usually left on for 1–7 days before being replaced by a new one. A serial casting program is indicated when moderate to severe spasticity cannot be managed by splints.

A resting or functional position splint is worn when the client is not involved in active movements or functional tasks. A schedule for splint-wearing must be established for the rehabilitation team and caregivers to follow. A typical splint schedule during the day requires the client to wear the splint for repeated, alternating 2-hour periods (2 hours on, followed by 2 hours off). The client must be monitored frequently for any skin breakdown or tonal changes that may change the initial fit of the splint. The team and caregivers should be trained in proper application and removal of each splint. Use of anti-spasticity splint positions the hand and wrist in a functional position and also abducts the fingers to further decrease spasticity. Indications for termination of a casting program include achieving functional ROM or plateauing (e.g., the individual has not gained significant improvement in ROM after two consecutive casts).

## Speech therapy management of Traumatic Brain Injury

## ASSESSMENT OF INDIVIDUALS WITH TBI:

- Assessing TBI adults is an evolutionary process. As patients physical, cognitive and behavioral characteristics change with recovery, what happens in testing also changes.

### Levels I, II, and III

- SLP's primary concern with RLA level 1-3 is to determine the patient's level of consciousness. Standard rating scales such as the Glasgow Coma Scale of the Glasgow Assessment Schedule provide a general subjective estimate of these characteristics.
- Patient's responsiveness to speech may be assessed by greeting the patient, and noting whether the patient looks towards the speaker, attempts to speak or respond motorically.
- Responsiveness to visual stimulation may be assessed by introducing lights or brightly colored objects.

- Responsiveness to tactile stimulation may be assessed by lightly touching and stroking the patient's limbs
- Responsiveness to olfactory stimuli may include pleasant odors such as cologne, vanilla extract and unpleasant odors such as ammonia or rubbing alcohol.
- Cognitive and communicative abilities
- Cognitive and communication impairments become a major focus of assessment with RLA's level V and VI patient's which includes:
- Alertness and attention
- There are 2 categories of alertness-
- Tonic alertness - Tonic alertness is an individual's ongoing receptivity to stimulation
- Phasic alertness - phasic alertness is an individual's momentary, rapidly occurring receptivity to stimulation.
- Inattentional impairments are universal consequences of TBI. Attention has been described as:

o Focused attention
o Selective attention
o Sustained attention
o Alternating attention
o Divided attention

- Adults with TBI often are impaired in all, although focused attention typically recovers relatively soon.
- Memory: Memory loss, the most common cognitive impairment among head-injured people, occurs in 20–79% of people with closed head trauma, depending on severity (Hall and Chapman, 2005).

o **Retrospective memory** is memory for the past events, experiences and information previously.

o **Prospective memory** is remembering to do things at specific time i.e. remembering to remember.

o Retention span denotes the number of new bits of information one can hold in immediate memory. Its testing includes a test of immediate retention span, in which the test -taker reproduces the information immediately after it is presented, and a test of short term retention, in which the test-taker must wait for a prescribed time before reproducing the information.

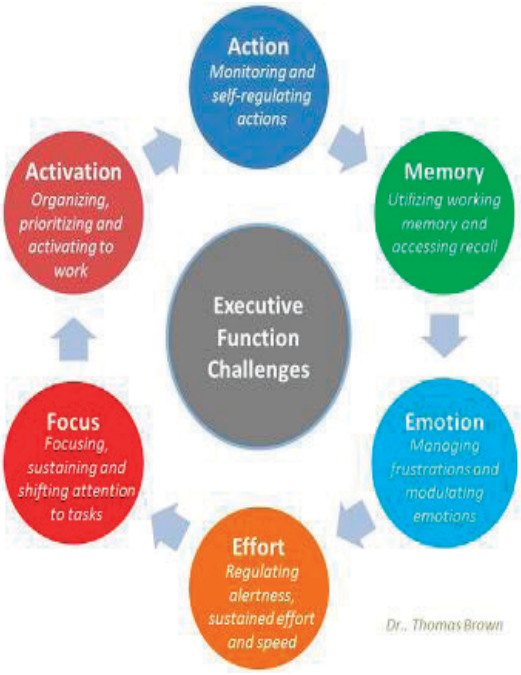
o Recall of new information is tested with lists of unrelated words that are read aloud to the test-taker until he can repeat the list. Then after a delay interval the examiner asks the test-taker to say the words in the list.

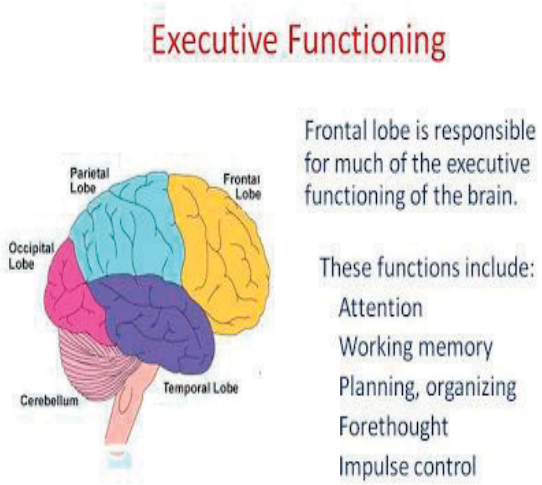
o Retrieval of information from remote memory is usually tested by asking the patient questions related to biographic information such as where and when the patient went to school and where the patient has worked.

o Visual memory is tested by showing the patient cards on which geometric pattern is printed and asking him to draw them from memory.

- Cognitive deficits that can follow TBI include impaired attention, disrupted insight, judgement, and thought, reduced processing speed, distractibility.
- Deficits in executive functions such as abstract reasoning, planning, problem-solving, and multitasking (Arlinghaus and Shoaib, 2005).
- People who have suffered TBI may also have difficulty with understanding or producing spoken or written language, or with more subtle aspects of communication such as body language.

There are various scales used for assessment of speech, language, cognition and swallowing abilities of individuals with TBI. Few of them are listed below:




- Frenchays Dysarthria assessment.
- Western Aphasia Battery (WAB).
- Boston Diagnostic Aphasia Examination (BDAE).
- Cranial nerve examination.
- Dysphagia evaluation.
- The ASHA Functional Assessment of Communication Skills in Adults.
- Communication Activities of Daily Living, 2nd Edition.
- Assessment of Attention and memory skills.

**Speech therapy treatment for individuals with Traumatic Brain Injury**

According to Geschwind, the memory problems exhibited by closed head injury patients are the result of underlying attentional and information processing deficits (executive function). Remediation therefore focuses on attentional and informational deficits and other aspects of executive function including disorientation and confused language.

Because of the smooth interior of the skull near the primary language centers specific aphasia syndromes are relatively infrequent (Ylvisaker and Szekeres, in Chapey, 1994).

- Therapy with Closed Head Injury patients will focus on:
- Improving attention.
- Perceptual abilities.
- Thought organization.
- Performance of higher level cognitive processing
- The techniques listed could be used for any patient who has cognitive problems.

**Symptomatic therapy involves**

- For improving attention, provide an environment in which distractions are minimized.
- As the therapy begins, it may be necessary to limit the length and intensity of sessions.
- Therapy sessions can be longer and tasks can be made more difficult as the patient's tolerance increases.
- It is often helpful to integrate familiar and personally significant items into therapy tasks, especially during initial sessions.
- Sensory stimulation in all modalities can be used to stimulate attention.

- **Improving perception:** According to Henderson and La Pointe (1990), the clinician should identify the sensory modality in which perception is most impaired and begin therapy by presenting stimuli in that modality. The least

affected modality should be used for cueing.

- **Thought organization/information processing:** Patients can be asked to sort items by size, shape, color, etc. also, they might be asked to name the item that does not belong in a particular group.
- **Closure:**

o To improve visual closure, the patient might practice completing pictures with missing parts.
o Tasks like completing sentences or words with phonemes missing can be used to work on auditory closure.

- **Sequencing:** The patient might be asked to order objects from small to large or light to dark. Also, he might practice unscrambling words or sentences.
- To improve visual tracking, the patient could be asked to follow a light with his eyes. To address auditory tracking, ask him to point in the direction of a sound source with his eyes closed.
- **Shape recognition:** Patients could be asked to trace or copy shapes
- **Word recognition:** Read a passage to the patient ask him to raise his hand every time he hears you say a particular target word.
- **Discrimination/matching:** First, the clinician should ask the patient to match two identical objects; next, she/he might match pictures with objects and pictures with other pictures.
- Letters, words, sentences and geometric forms can be incorporate into therapy tasks as the patient's abilities improve.
- As the patient improves, introduce more practical activities like sequencing the steps in daily activities or following complex directions.
- **Executive Function/problem solving:** According to Ylvisaker, Szekeres, and Feeney, 2001, a functional operational definition of the executive system is awareness of strengths and weaknesses and associated understanding of the difficulty level of tasks.
- Based on this awareness, an ability to

√ set reasonable goals,

√ plan and organize behavior designed to achieve the goals

√ flexibly revise plans

√ Strategically solve problems in the event

| | |
|---|---|
| Oral–peripheral stimulation | If a patient is ignoring one side of his mouth, a lollipop could be use to encourage him to move his tongue into that area. |
| Auditory stimulation | Bells or other simple auditory stimuli could be used at first. As the patient's ability to attend improves, present more complicated stimuli like music or speech. |
| Tactile stimulation | Contrasts like hot and cold or soft and rough could be used to stimulate attention. |
| Visual stimulation | Different colors might be used. Also, light might be contrasted with dark. |
| Taste | Sweet versus sour may cause the patient to attend |
| Olfactory stimulation | Start with strong, noxious smells like ammonia, which really command attention, and then present more pleasant scents, such as perfume. |

of difficulty or failure.

- Prognosis:

Prognosis for recovery of language functions in head injured patients is linked to the prognosis of cognitive ability. (Brooks et al., 1980; Hagen, 1984).

Groher (1983) proposed that although Closed Head Injury patients recover basic language skill within first 6 months post injury; they continue to show deficits in analysis and synthesis of expressive and receptive language.

Prevention:

Prevention measures include use of technology to protect those who are in accidents, such as seat belts and sports or motorcycle helmets, as well as efforts to reduce the number of accidents, such as safety education programs and enforcement of traffic laws.

**Psychological management**

Brain injury can lead to mild impairment or complete inability to function that may be temporary or permanent. Injuries to the brain can also affect functioning of other bodily systems including blood pressure, temperature regulation, bladder control, and bowel movements. All those with a moderate to severe brain injury are likely to have some difficulties with the cognitive skills at least in the early stages of recovery. The most common cognitive problems after brain injury involved concentration, memory and general speed of processing information. However, a broad spectrum of cognitive skills may be affected, including perception, language and the awareness of self and others. A set of skills known as executive functions including planning, organization, multitasking, and flexible thinking is also commonly affected and this can be particularly debilitating.

**Memory function:**

Memory problems usually include a period of amnesia surrounding events that took place before and after the injury and/or difficulty in remembering new information.

- ***Retrograde amnesia-*** The loss of memory for events leading up to the injury is known as retrograde amnesia. People with brain injury rarely recall the circumstances of their injuries. Retrograde Amnesia may extend from just a few seconds to minutes, hours, days, weeks or even longer. In addition to the period of total amnesia people frequently complain of incomplete or vague recall of days, weeks or months before their injury. However, they may be able to recall some of this information when prompted by family or friends.
- Post- traumatic Amnesia- The period of confusion and memory loss that happens after the injury or accident is known as post traumatic amnesia (PTA). This covers the period of time from the injury until the person starts to regain consciousness or continuity in their memory. This may range from a few minutes after injury (after a mild injury), to a period of weeks, months or years (after a very severe injury). In post traumatic amnesia, people may be able to hold a sensible conversation about known subjects, such as the weather or something that has just happened, but then forget them afterwards.
- Anterograde Amnesia- The inability to remember new information after the injury is known as anterograde amnesia. The degree of this problem ranges from occasional forgetfulness to an inability to recall basic information, such as personal and family details, current circumstances, and daily routine. Many people with the serious head

injury report problems in learning and remembering names, messages, appointments, routes, and plots of books or television program, etc. People with more severe memory difficulties may also forget where they are going to or what they are doing, and may be dependent on family members to remind them of basic daily activities. In contrast, memory for a long term past maybe retained well.

**Attention and concentration:**

After a head injury, people often find difficulty in attending to or maintaining concentration or interest in activities. While some people may struggle specifically with task that requires divided attention or multi tasking, many others find that their concentration and train of thought is easily disrupted, for example, by noise or interruptions. They may have to start an activity all over again, or ask for information to be repeated. Difficulties with attention and concentration can have far reaching effects on work, leisure, and social activities.

**Speed of information processing:**

People also commonly experience a reduction in the speed in thinking and their responses to external stimuli, even after relatively minor brain injury. They may find that they are unable to take in new information as quickly as they used to. They may often find answers left behind by conversations or television programs, slow to read newspapers or books, sluggish in their reactions, or generally slow at works or in their leisure activities.

**Executive function:**

People with a brain injury often experience difficulties with executive skills like planning, problem solving, reasoning, decision making, and self monitoring. In general conversation they may lose the train of thought and go off topic. Others may be inflexible in their thinking, repeating one rigid opinion and failing to appreciate other people's point of view. This means that conversations can be frustrating and unrewarding for others, particularly when the same conversation is repeated several times. It may be hard for a person with brain injury to identify and analyse problems, think of possible solutions, make decisions and implement coping strategies. They may also be poor at planning, monitoring and evaluating their performance, and organizing their lives effectively.

**Awareness and insight:**

Awareness of oneself and of others can be profoundly affected by brain injury. The people suffering from brain injury maybe unaware of how they are presenting themselves to others. This is particularly common in the early stages of recovery. As people recover, they may gradually regain the self awareness an insight into their more obvious problems (such as mobility, speech or memory), but may never achieve insight into more subtle changes in their skills or behaviour. As a result, they may fail to pick up on hints or social cues and be therefore misjudging social situations, appearing clumsy in their social skills and inappropriate in their behaviour.

**Disinhibition:**

Disinhibition is loss of control over behaviour, resulting in socially inappropriate behaviour. This ranges from a tendency to divulge personal information too freely; to disturbing and unpredictable outbursts of uncontrollable rage.

**Irritability and aggression:**

One of the most common behavioural changes after brain injury is that of increased irritability. People with a brain injury are often impatient, intolerant of others mistakes, and easily irritated by interruptions. They are frequently reported to be short tempered, for example, when things do not work out as expected or where there are differences of opinion with

family or colleagues.

**Apathy and loss of initiative:**

In contrast with those who exhibit poor control over irritability and aggression, some people with the brain injury may become passive, unresponsive and lacking in initiative. Some people may appear unconcerned and even unaware of their problems, especially in the early stages of recovery. Others may appear interested and have good intentions to carry out activities, but are unable to organize themselves and initiate action.

**Egocentricity:**

People may also become egocentric after brain injury, tending to be self-centred and appearing not to consider the feelings or needs of their family and friends. Cognitive impairments can inability to appreciate others point of view, and they may be unaware of the needs of others.

**Liability:**

In parallel with their loss of behavioural control, many people with a brain injury experience loss of emotional control. They may react strongly and unpredictable to events which would not previously have troubled them, and may also show rapid and swings in mood.

**Depression:**

For people with mild injuries depression may be an early reaction to the trauma of the injury and subsequent disability, where as for others it may develop as they struggle helplessly to regain their lost skills. Commonly, depression comes into picture when active treatment is complete and the person returns home, only to be confronted by the realisation that life will probably never be as it was before. The loss of previous physical, cognitive, and social skills, the impact on personal relationships, and the fact that they are unable to return to the previous leisure and social activities or fulfil their work aspirations, can be extremely hard to accept, and can understandably lead to depression.

**Anxiety:**

Many people suffer from anxiety after brain injury, which impact upon their daily lives and may obstruct the rehabilitation. Those with less severe injury may be anxious early in recovery, feeling disturbed by the changes to the cognitive skills and personality. People with a more severe injury may become anxious later on, when they come to appreciate the extent of the long term disability and feel anxious about the future.

**Post traumatic stress disorder (PTSD):**

Some people may experience post traumatic stress disorder (PTSD). This is a serious psychological reaction to a traumatic event. It involves the persistent re-experiencing of the trauma, avoidance of stimuli which remained the person of the event, increased arousal, and a number of emotional responses. As such, PTSD is most commonly experienced after mild brain injuries when memories of the circumstances surrounding the injury are retained. However, some others develop of fear of circumstances similar to that of the injury and a small number of people have disturbing memories of the early stages of their recovery.

Psychological assistance can help people with brain injuries and their families throughout the course of recovery, from acute care and rehabilitation to resettlement and long-term support.

**Cognitive assessment**

As people with brain injury region consciousness, they generally experience period of disorientation or confusion. As the person improves, a formal neuropsychological assessment of cognitive skills is often required. Such an assessment is likely to include tests of general intellectual abilities, memory and learning skills, and may also include tests of

attention, speed of information processing, and executive skills. Assessment of intellectual ability may include basic education test (general knowledge, arithmetic, vocabulary, word reading) and various reasoning task to assess the ability to apply this knowledge to new problems. Assessment of memory function is likely to include recall, learning, and retention of both verbal materials (stories and list of words) and visual materials (geometric shapes, faces, or objects). More specialised tests of other areas of cognitive functions such as perception and language might be carried out according to the inquiry.

**Cognitive rehabilitation**

A formal cognitive assessment should identify the extent of any impairment, as well as areas of strength on which to plan the programme for rehabilitation. A range of treatment strategies may then be adopted, depending on the specific nature of problems and the stage of recovery. This may start with orientation, exercises, and education, and then progress to the development of strategies to manage difficulties with cognitive skills such as a tension, memory, or perception, or to compensate for lost skills through the use of external aids. Reassessment is important in monitoring progress, reviewing treatment strategies and setting new goals taking into consideration improvement and, later, in guiding resettlement plants.

**Behavioural management**

During the initial stages of recovery, people with brain injury often become restless and agitated, for exhibit disinhibited or aggressive behaviour. In some cases a psychiatric opinion and medication may be required to contain agitated, disruptive, or aggressive behaviour. In such situations, a specific behaviour modification programme may be required. This involves staff and relatives working together to systematically discourage disruptive behaviour and actively encourage positive behaviour. In extreme cases, there are few units that specialise in management of very severe behavioural problems.

**Social skills training**

Many people who suffered from head injury experience difficulties with their social skills and relationships due to a combination of cognitive, behavioural, and emotional changes. Social skills training may be of value in raising self-awareness and social awareness, as well as providing a forum for peer support. It can also help people to accept the inappropriateness of some of the conversation and behaviour, and the need to be more controlled and bring about a positive change in the social activities. This can help people to rebuild the confidence, both in maintaining the existing relationship and in making new friendships.

**Psychological counselling**

People with brain injuries benefit from being given feedback and advice about the nature of the psychological difficulties that they will suffer from as a result of the injury. A detailed explanation of the nature of the person's difficulties, and guidance about the likely extent of recovery, can help them again more realistic outlook, and so help them make appropriate plans for the future. This is especially important where there is a tendency for the person to underestimate the extent of the difficulties, and to overestimate future recovery. In the early stages of recovery, this belief may protect people from becoming depressed and serve as a powerful motivating factor. However, continued unrealistic expectations of recovery may impede both progressive rehabilitation and adjustment long term disability.

Psychological counselling may also help in evaluating the employment opportunities and getting back in the social and leisure activities. Relatives usually benefit from detailed feedback about the results of formal assessments, the nature and implications of acquired disability, and some explanation of the rationale and goals of treatment. They often seek advice about management of cognitive and or behaviour problems

and may assume the role of co- therapist in reinforcing treatment strategies at home.

**Individual psychotherapy**

Some people, understandably, feel devastated by the effects of their brain injury. Although psychologist can help working through depression and anxiety, along with related issues, people who are not falling on the clinical spectrum also benefit from psychotherapy. Unsurprisingly, in the confusion of the present and the uncertainty about the future people often cling to the past following the brain injury. Some people attempt to carry on as before, unable to accept the changes that have occurred, and may continue to judge themselves by the pre injury standards that they can no longer meet. For others, the preoccupation with what they have lost may cloud their appreciation of the positive attributes and potential that remains. Psychotherapy can help people to re-appraise their new situation. With support and guidance they can often be help to move forward to explore the new selves, and search for a new direction and purpose to their lives. Cognitive behavioral therapy (CBT), insight oriented therapy, relaxation training, development of social skills, and anger management can also be useful in helping a person understand and accept the injury as well as adjust relationships and roles after the injury, when necessary.

**Family therapy**

Family members face similar process of adjustment following brain injury. As the person improve, against their Independence and resumes former roles, family relationships can actually revert to their previous pattern. However, for those with permanent disability, some changes in family roles and relationships maybe necessary. Many families adjust well towards the changes, and some report feeling closer as a result. Children may also benefit from help in understanding and coping with the changes in their parents.

**Conclusion**

Having a good personal support network is very important for those living with a traumatic brain injury. When someone is affected by a head injury, their entire family is also impacted. Their activities of daily life may also need assistance. Depending on the severity of the injury, a person's interpersonal relationships and social life may is also affected. Family and friends are an important part of the continuous rehabilitation process. Support groups are available for those suffering from head injury as well as their families.

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# Chp. 8 Multiple Sclerosis

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## Overview

Multiple sclerosis is a chronic inflammatory demyelinating disease of the central nervous system affecting younger adults between age 20 to 40 years of age. It is an unpredictable disease affecting the flow of information within central nervous system connecting brain and the body. The CNS is made up of the brain, spinal cord and optic nerves. Dr. Jean Charcot in 1868 who defined the disease by its clinical and pathological characteristics: paralysis and cardinal symptoms of intension tremor, scanning speech and nystagmus, later termed as charcot's triad.

## Epidemiology and Etiology

In the 1980s, the prevalence of MS in India was estimated to be nearly 1/100,000. Evidence collected from hospitals throughout India suggests that the number of MS patients diagnosed annually has nearly doubled. There are no large-scale studies from India on the incidence and prevalence of MS. MS is most predominant in whites of Northern European ancestry and is rare in some races like African blacks and Eskimos. By studying people who move from one area to another, researchers have learned that individual risk changes based on location. They have concluded that some exposure in the environment increases the risk of for MS. The incidence of MS is seen more in places which are farther from the equator, MS occur more prominently over there. Studies have shown that distribution of MS depend on areas of high, medium and low frequency. Other environmental factors associated with the development of MS include cigarette smoking (odds ratio of 1.81), animal fat intake, and deficiency of vitamin D. A multitude of other environmental factors have been suspected to alter the risk for MS (cold climate, precipitation, amount of peat in the soil, exposure to dogs, and consumption of meat, and dairy products), but none has been verified to be an independent risk factor.

## Pathophysiology

In multiple sclerosis, the immune response triggers the production of T-Lymphocytes, macrophages, & immunoglobulin's (antibodies). In turn, a command or trigger protein, the antigen is activated, producing autoimmune cytotoxic effects within the CNS. The blood brain barrier fails and myelin sensitized T - Lymphocyte cells enter and attack the myelin sheath that surrounds the nerve. Myelin serves as an insulator, speeding up the conduction along nerve fibers from one node of Ranvier to another termed salutatory conduction. It also serves to conserve energy for the nerve because depolarization occurs only at the nodes. Disruption of myelin sheath produces active demyelination, slowing neural transmission & causing nerves to fatigue rapidly. With severe disruption, conduction block occurs with resulting disruption of function.

Within the CNS, the immune system causes inflammation that damages myelin — the fatty substance that surrounds and insulates the nerve fibers — as well as the nerve fibers themselves, and the specialized cells that make myelin.

- When myelin or nerve fibers are

damaged or destroyed in MS, messages within the CNS are altered or stopped completely.

- Damage to areas of the CNS may produce a variety of neurological symptoms that will vary among people with MS in type and severity.
- The damaged areas develop scar tissue which gives the disease its name – multiple areas of scarring or multiple sclerosis.
- The cause of MS is yet not known, but it is believed to involve genetic susceptibility, abnormalities in the immune system and environmental factors that combine to trigger the disease.
- People with MS typically experience one of four disease courses (types of MS). There are over a dozen treatments to help modify the MS disease process.

## Clinical Course of Multiple Sclerosis

Course of MS can not be predicted.

Patients, presents with one of the following clinical course:-

**Benign MS:** Benign multiple sclerosis" is often temporary as apparently benign disease often becomes disabling. Few patients get rare relapses of these disease, in which patients' remains functional till 15 years after onset. It affects 20% of MS patients

**Malignant MS:** In this type there is rapid progression of the disease which sometimes cause significant disability or death within short period after the onset

Relapsing/Remitting type: It is the most common type and is seen among 70% of MS patients and occur frequently, one to several times per year.

**Primary Progressive MS:** In the primary progressive form continuous worsening occurs with steady progression but not interrupted by distinct relapses. It affects 10% of MS patients

**Secondary-Progressive MS:** In this type relapsing remitting disease followed by progression with or without occasional relapse, minor remissions and plateaus.

Progressive-relapsing

Progressive disease from onset but with clear, acute relapses that may or may not have some recovery or remission; Periods between relapses are characterized by continued progression.

That mainly seen in people, whose disease onset after 40 years.

## Signs and symptoms

MS usually affects the optic nerve and any tissue within the brain or spinal cord, as explained in pathophysiology so almost any neurological symptom can result. Individual assessments are required to identify these problems present. Even so, the following list constitutes the most common problems encountered by people with MS.

**Fatigue:** Of people with MS, 65% to 97% report fatigue during the course of the disease; as many as 40% of people with MS state that fatigue is their most disabling symptom.15 There are two types of fatigue in people with MS: primary and secondary. Primary fatigue, often called

lassitude, is caused by the effects of the demyelination and axonal destruction and its effect on nerve conduction. Restorative rehabilitation has little effect on primary fatigue from neurodegeneration. Secondary fatigue results from problems such as deconditioning, infections, sleep disturbances, poor nutrition, medication side effects, other medical conditions (such as thyroid disease), and heat intolerance. Clinicians should be extremely careful to separate the types of the fatigue in order to determine the most appropriate interventions.

**Sensory Symptoms:** This is the most common presenting manifestation in MS (21% to 55%) and ultimately develop in nearly all the patients. The symptoms like Hypoesthesia, numbness, Paraesthesia (tingling) Dysesthesias(burning) may occur in one or more limbs, trunk, face or in combinations. The more distinctive sensory relapses of MS consist of the sensory cord syndrome and sensory useless hand syndrome. The sensory symptoms may ascend to the trunk, producing a sensory level, or may involve the upper extremities. In most of the MS patients it has seen that they develop persistent sensory loss, usually consisting of diminished vibratory and position sensations in distal extremities.

**Motor Symptoms:** Motor symptoms like Weakness, Spasticity or Paralysis, Fatigue, Inco-ordination, Intentional Tremor, Impaired Balance, Gait Disturbances occur due to Pyramidal tract dysfunction. It can occur acutely or in a chronic progression with weakness of one or more limbs and facial weakness, leg stiffness that impairs gait and balance, or extensor or flexor spasms. Exercise or heat frequently worsens subtle deficits. Muscle atrophy is usually due to disuse, but lesions of lower motor neuron fibers or of the anterior horn itself can cause pseudo-radiculopathy with segmental weakness, atrophy, and diminished reflexes. Motor symptom manifestations of MS in 32% to 41 % of all cases and their prevalence is higher than 60% in long-standing MS.

**Visual Symptoms:** The initial symptoms of MS include optic neuritis (ON) in 14 % to 23% of patients, and more than 50 % experience a clinical episode of optic neuritis during their lifetime. The most common manifestation is visual loss in one eye that evolves over a few days. Periocular pain, especially with eye movement, usually accompanies and may precede the visual symptoms.

**Cerebellar Symptoms:** Cerebellar pathways are frequently involved during the course of MS, but a predominately cerebellar syndrome is uncommon at onset. The manifestations include dysmetria, dysdiadochokinesia, action tremor with terminal accentuation, dysrhythmia, breakdown of complex motor movements, and loss of balance. Patients with long-standing MS may develop a "jiggling" gait and an ataxic dysarthria with imprecise articulation, scanning speech, or varying inflection, giving it an explosive character.

**Bladder Symptoms:** Urinary urgency, frequency, and urge incontinence (due to detrusor hyper- reflexia or detrusor-sphincter dyssynergia) result from spinal cord lesions and are frequently encountered in MS patients. The combined incidence of bowel and bladder dysfunction in MS is more than 70%. Symptoms of bladder dysfunction may be transient and occur with an exacerbation but are commonly persistent. Impaired vesicular sensation

causes a high capacity bladder and may lead to bladder atonia with thinning and disruption of detrusor muscle. Due to incontinence, there is constant dribbling of urine in this irreversible condition. Interruption of brain stem mictuirition center input sometimes lead to co-contracture of the urinary sphincter and detrusor muscles (detrusor=sphincter dyssnergia). If it is untreated, high pressure may lead to hydronephrosis and chronic failure.

**Bowel Symptoms:** Constipation is a common problem, occurring in 39% to 53% of MS Patients, especially with limited activity and spinal cord involvement. Fecal incontinence is a socially devastating symptom that is often associated with perineal sensory loss in MS patients.

**Sexual Dysfunction:** It is a frequent problem in MS. Nearly two thirds of patients report diminished libido. One third of men have some degree of erectile dysfunction, and a similar percentage of women have deficient vaginal lubrication. Besides direct neurological impairment, sensory loss, physical limitations, depression, and fatigue additionally contribute to sexual difficulties in MS patients and sometimes the partner's attitude and psychological factors dealing with self-image, self-esteem, and fear of rejection may also lead to impotence or loss of libido.

Intense vertigo associated with nausea and emesis is an occasional manifestation of MS relapse.

**Speech and Swallowing:** Dysphagia is often due to impairment of cranial nerves IX, X and XII and generally appears late in the course of some patients.

**Cognitive Symptoms:** Cognitive symptoms are present in 40% to 70% of MS patients. The pattern of cognitive decline is typified by decrease of episodic memory, processing speed, verbal fluency and difficulty with abstract concepts and complex reasoning. To a lesser extent, executive functioning and visual perception, semantic memory and attention span may also decrease. General intelligence is not typically affected.

**Emotional Symptoms:** Anxiety and depression are more frequent in MS patients than in the general population. In long-term studies, the incidence of depression in MS patients is close to 75%. Patients sometimes experience uncontrollable weeping or less commonly laughter incongruent with their mood. Interruption of inhibitory corticobulbar fibers is responsible for these symptoms (pseudobulbar effect).

**Pain:** Intense pain and ipsilateral or crossed sensory symptoms may accompany them. Paroxysmal weakness occurs, but it is uncommon. Paroxysmal sensory symptoms like tingling, prickling, burning, or itching may occur in MS patients. Sharp neuralgic pain is also common. Trigeminal neuralgia may appear in patients with MS. The occurrence of trigeminal neuralgia in a person younger than age 40 is suggestive of MS.

Lhermitte's sign (transient sensory symptoms usually precipitated by neck flexion)- It is an electrical or tingling sensation that travels down the spine or into the extremities. Although quite common in MS, Lhermitte's sign can also occur with a wide variety of other disorders, such as vitamin B12 deficiency, spondylosis.

**Seizures:** It has seen that seizures occur in larger proportion of MS cases compared

to normal control subjects. Cortical and juxtacortical lesions may be responsible for the increased incidence of seizures in MS patients. Focal motor seizures, possibly with secondary generalization, are the most frequent. The occurrence of seizures usually follows one of two patterns. Sometimes focal onset seizures begin early in the course of MS and become a chronic problem that may be difficult to control.

## Rehabilitation Management

Multiple Sclerosis is a chronic neurodegenerative condition, which results in loss of physical and cognitive function due to destruction of the neurons. People with Multiple sclerosis experience physical and cognitive impairments potentially leading to inactivity and resultant deconditioning.

Depending on the severity of the condition; therapist can use compensation or restoration strategies for lost function.

Compensation includes interventions such as wheelchairs or walkers to assist with mobility or braces for inadequate muscle power. By maximizing cardiovascular endurance by increasing maximal oxygen uptake or restoring full range of motion (ROM) is an example of restoration which aims at increasing the capacity of the system. The compensation or restoration should be the goal of therapeutic interventions by rehabilitation professionals.

## Assessment

- Assessing Body System Problems Contributing to Activity Limitations
- Ataxia and In-coordination
- Spasticity
- Fatigue
- Vestibular Dysfunction
- Cognition
- Assessing Activity Performance and Participation
- Balance
- Gait
- Assessing Quality of Life

## Interventions

The rehabilitation team includes nurses, occupational therapists, physical therapists, speech language therapist, neuropsychologists, and physicians. The rehabilitation goals for persons with Multiple sclerosis are to maintain and maximize and function and prevent complications so that they can do all the activities of the daily living independently. The rehabilitation professionals require to be flexible and creative in the variable presentation that patient with Multiple sclerosis can manifest. The impairments, activity limitations and participation restrictions are identified during the assessment & are considered in the goal setting and plan of care developed to manage a patient. The rehabilitation should be a part at all stages of the disease, even in comprehensive health care plan. The neuroprotective effect on the brains of people with neurological insults is shown in the evidence which may be due to increased activity, whether cognitive or physical.

This also implies that rehabilitation, specifically exercise, should occur early in the course of the disease and not only after

clinical disability has occurred.

## Fatigue

Fatigue is one of the most frequent and disabling symptoms associated with Multiple Sclerosis and is best managed with a multidisciplinary team composed of physicians, physical therapists, occupational therapists, and nurses.

The causes of fatigue can be divided into two basic categories: primary and secondary.

1. Primary fatigue related to neurodegeneration and demyelination may have very less options for treatment.

2. Secondary fatigue caused by deconditioning, comorbidities, depression, poor nutrition, heat intolerance, sleep disturbance, and medications may be more easily managed. Interventions for fatigue management include cooling devices, energy conservation and work simplification techniques and education training, exercise, etc.

3. Exercise shows promise as an intervention that can improve fatigue for people with Multiple sclerosis that may improve muscle weakness caused by disuse and deconditioning. Energy conservation is defined as energy effectiveness and includes an analysis of individuals' home, work, and leisure activities and the environments in which they occur in order to develop activity modifications designed to reduce fatigue. This can include a variety of strategies such as reducing energy expenditure through activity and modification, workspace organization and improving efficiency of movements; balancing work and rest periods; delegating tasks; evaluating standards and prioritizing activities; and using assistive technologies that conserve energy usage. Patients may need to be prescribed assistive devices for ADLs. People with Multiple sclerosis who have spasticity has difficulty in walking, hence using wheeled mobility for longer distance outings (to the shopping mall, an extended event, on vacation) can conserve energy and extend the time a person can participate in activities of importance to him or her. However, therapists should be aware that using assistive devices such as walkers or crutches actually increases energy expenditure for elderly people, and therefore the need for improved support must be balanced with the increased energy burden an assistive device might add.

## Spasticity

Several rehabilitation strategies to manage spasticity are available, including Range of motion, stretching, light pressure, stroking and weight bearing exercises. The Multiple Sclerosis Council for Clinical Practice Guidelines recommends, stretching a muscle with spasticity for 60 seconds or longer or using a prolonged stretch, lasting hours, with braces or splints helps in reducing spasticity.

## Balance and Postural Control

Balance is foundational to the ability to stay upright and perform dynamic movements. It is a frequent problem in people with MS and results in a person limiting his or her participation in home, work, and leisure activities. Abnormalities of balance can

increase the risk of falling. Other fall risk factors that have been identified include fear of falling, male sex, poor concentration or forgetfulness, and urinary incontinence. Rehabilitation programs must be based on a thorough understanding of the impairments, personal and environmental factors that may be contributing to the balance dysfunction.

## Mobility

People with Multiple sclerosis rate gait as one of the most important bodily functions; gait is often adversely affected in people with Multiple sclerosis. Lesions in the brain and spinal cord produce a wide variety of potential impairments that can adversely affect gait. Imbalance, fatigue, spasticity, incoordination, muscle weakness, and sensory system impairments were all reported to negatively affect ambulation ability. Therefore, addressing each of these impairments has the potential to improve gait. Treadmill training has been investigated in several small, pilot or case studies with promising results of improved Quality of life, energy expenditure, and gait parameters. Several exercise studies have an association with improved gait. Technique that shows promise for improving mobility in people with MS is an evaluation and intervention approach that uses small amounts of weight placed on the torso in response to identified balance dysfunction. Balance-Based Torso-Weighting (BBTW) is an intervention that uses directional loss of balance in both static and dynamic assessment to determine where small amounts of weight (generally less than 1% to 1.5% of body weight) are placed in a treatment orthotic called Balance Wear. The Balance Wear orthotic can be worn during the performance of activities in therapy or daily for home, work, or leisure activities. Other technologies include functional electrical stimulation (FES), neuro-prostheses, and orthotics. FES is applied to specific muscles or muscle groups to activate weak muscles. Some of these stimulators can be built into a neuro-prostheses that can be set up for use during exercising or walking. Orthotics such as the ankle-foot orthosis (AFO) or hip flexion assist orthosis (HFAO) can compensate for muscle weakness in the lower extremity, improve foot and knee positioning, and reduce energy expenditure.

## Pain and Dysesthesias

The occurrence of pain in people with MS is often underestimated. Pain can be acute, as in optic neuritis or Lhermitte syndrome, or chronic, as in dysesthesias in the limbs or joints or mechanical pain related to abnormal positions or repeated movements that cause abnormal wear and tear on the musculoskeletal system. Physical therapists can address poor body mechanics and weakness and poor movement patterns with retraining, and soft collars may help reduce Lhermitte syndrome. Transcutaneous electrical nerve stimulation has been suggested anecdotally as beneficial for reducing pain. Cognitive behavioral therapy has been researched for managing chronic pain; however, little evidence exists for using it in people with Multiple sclerosis.

## Bladder Dysfunction

Urinary incontinence and retention are common and often embarrassing problems for people with MS. Patients may be advised to avoid bladder irritants including caffeine, alcohol, concentrated urine, and infection. Physical therapists

may work with patients to assess the factors contributing to bladder dysfunction by retraining hyperactive or weak pelvic floor muscles using biofeedback techniques and exercise. Nurses may need to teach patients with urinary retention intermittent catheterization.

## Cognition

Strategies for managing cognitive impairments include compensation techniques such as memory notebooks, diaries, calendars, and computer-assisted programs for memory, attention, or other executive functions. Neuropsychologists, speech-language pathologists, and occupational therapists can all direct cognitive rehabilitation programs. Strategies for coping with cognitive impairments are often shared with the other members of the health care team for reinforcement.

## Occupational Therapy in MS:

EVALUATION

1) Physical abilities: Range of motion, muscle strength, balance

2) Sensory function, paresthesia, ataxia, intentional tremors

3) Level of independence in selfcare

4) Behavior and emotional stability and adjustment to disability

5) Psychological and intellectual functions

TEST AND MEASURES

**1) Cognition**

The Mini-Mental State Examination (MMSE) or Folstein test is a 30-point questionnaire that is used extensively in clinical and research settings to measure cognitive impairment. Administration of the test takes between 5 and 10 minutes and examines functions including registration (repeating named prompts), attention and calculation, recall, language ability to follow simple commands and orientation.

**2) Affective and psychosocial function**

Beck depression inventory

**3) Sensation**

McGill pain questionnaire

Dallas pain questionnaire

**4) Visual acuity**

Acuity, tracking and accommodation should be examined

**5) Cranial nerve integrity**

Motor and sensory cranial nerve function should be examined. Optic pain for optic neuritis, oculomotor dyscontrol, trigeminal neuralgia and impaired gag reflex

**6) Range of motion**

Active and passive ROM should be examined.

**7) Muscle performance**

Manual muscle testing for functional strength and dynamometer for grasp and pinch should be examined.

**8) Fatigue**

Modified Fatigue Impact Scale (MFIS): it is self-report consist of 21 item questionnaire. It includes effect of fatigue on cognitive, physical and psychosocial function. It has 5point ordinal scale with 0: never and 4:

almost always.

**9) Motor function**

Spasticity can be examined using Modified Ashworth Scale.

**10) Posture**

Static and dynamic postural control in sitting and standing.

**11) Balance, gait and locomotion**

Berg balance scale, Tinetti Performance Oriented Mobility Examination (POMA).

Timed walk test – 10meter walk test or 6-minute walk test, Dynamic Gait Index and Ambulation Index.

River-mead visual gait examination was developed for MS patients.

**12) Aerobic capacity and endurance**

Rating of perceived exertion scale and Dyspnea scale

**13) Functional status**

Basic activities of daily living (BADL), instrumental activities of daily living (IADL)

Functional independence measure is used for patients who are undergoing rehabilitation.

**14) General health**

Health status questionnaire (SF-36) The Short Form-36 was derived from the General Health Survey of the Medical Outcomes Study by Stewart and colleagues (1988). Administration time is approximately 10 minutes. The SF-36 is a structured, self-report questionnaire that the patient can generally complete with little or no intervention from an interviewer. However, patients with visual or upper extremity impairments may need to have the SF-36 administered as an interview. Interviewers should be trained in basic interviewing skills and in the use of this instrument. The scoring system for the SF-36 is relatively complex and generates subscale scores for physical functioning, role limitations due to physical problems, bodily pain, general health perceptions, vitality, social functioning, role-limitations due to emotional problems, and mental health. Two summary scores can also be derived from the SF-36: the physical component summary and the mental component summary.

**15) Expanded Disability Status Scale (EDSS)**

It is 0-10 rating scale in 0.5-unit increments represent higher level of disability.

1- No disability and 10- death due to MS.

## Occupational Therapy Management

Occupational therapy aims at maintaining the functional independence and avoids fatigue. MS may result in life threatening events because person may have multiple affectations like fatiguability at end of day or in afternoon, diminished vision, decreased sensation, incoordination etc. Possible strategies include restoration, compensation, adaptation and prevention.

## Rehabilitation includes

- Inpatient rehabilitation program
- Outpatient rehabilitation program
- Home based programs

- Vocational rehabilitation program
- Functional mobility programs
- Fatigue management –face to face format and long-distance format
- Health promotion programs

## Management of sensory deficits and skin care

Pressure relieving devices i.e. mattress made up of gel, water or air (fig.1). Cushions of seat of wheelchair should be of foam, air or fluid filled. Soft shoes should be used. Patient is advised to change his position every 2hrs in order to avoid bed sores. Skin should be kept dry and clean. Clothing should not be tight. It should not be pressing any area of skin.



**Fig 1. Pressure relieving mattress**

## Management of pain

Soft collar is used for Lhermitte's sign i.e. the sudden electric sensation felt in the back of your neck or in the spine after bending head towards the chest (fig. 2). Stocking of gloves to relieve pain and feel neutral warmth. Postural retraining and postural correction will help to prevent pain. Stress management techniques, relaxation training and biofeedback are helpful. Modalities like TENS can be used to relieve pain.



**Fig 2. Soft collar**

## Exercise training

Based on FITT principle, strengthening and conditioning exercises are use.

## Management of fatigue

Energy conservation techniques and work simplification refer to completing tasks in the most energy efficient way, in order to have enough energy for the activities you enjoy most (fig.3). The following are some principles of energy conservation and work simplification that will help you plan your activities to conserve your energy, and pace yourself appropriately.




**Fig 3 Energy conservation**

1) Plan, set priorities, and balance your activities when planning your day.
2) Sitting saves 25% of energy so sit whenever you can. Tasks such as folding

laundry, doing dishes, preparing food, bathing, and dressing can all be done while sitting.

3) Rearranging work areas i.e. kitchen, desk, bathroom or sink area -to avoid excessive standing in order to save energy.

4) Take frequent breaks during the activities.

5) Slide objects instead of lifting or carrying them.

6) Avoid all unnecessary bending, reaching, and stretching. Keep all things within reach.

7) Combine details. Cook and serve in the same container

8) Complete your bowel program and then shower since this eliminates the need to undress and dress an extra time.

## Management of spasticity

Splints and orthotic devices are often used in the treatment of spasticity to reduce tone, prevent contractures, and reduce pain. Splints act mechanically by reducing muscle spindles' reaction to stretch and are generally applied to position affected joints in opposition to the tonal pattern. Although clinical observations suggest tone to be reduced with splints, studies have not determined the most effective design. Orthotic devices control joint instability and may alter the loading of a limb to prevent stretch reflex activity in antagonist muscles resulting in tone reduction. Prolonged muscle stretch is provided by both static and dynamic orthoses to prevent or correct early contractures that result from chronic spasticity. Devices such as ankle-foot orthoses (AFO's) (fig 4) may help improve functional abilities, such as ambulation. Plaster casting, an extension of splinting, acts to reduce tone by inhibiting the response of afferent fibers arising in the Golgi tendon organs. Mild soft tissue contractures are managed by prolonged immobilization through serial casting. With serial casting, an extremity is positioned in full stretch and casted in that position; the cast is left in place for 7 to 10 days and is then removed. The patient undergoes range of motion therapy between castings and is then re-casted at the new (reduced) angle; this is repeated 4 to 5 times. After the cast is removed, further therapy and splinting is often needed to maintain the improved range of motion. It is suggested that serial casting is most effective within 6 months of an acute injury, while neurologic recovery is still possible.

PNF pattern, cryotherapy and hydrotherapy is given to reduce spasticity.




**Fig 4 Frenkel's exercises**

Management of coordination and balance deficits

Static control in various position i.e. sitting, quadruped, kneeling, plantigrade and standing. Reachouts, weight shifting

and stepping activities to improve balance. Weighted jackets, boots, weight cuffs to reduce tremors while performing activities. Frenkel's exercise is used to improve coordination (fig.5). Assistive devices like canes, forearm crutch and walker are used. As disease progresses patient may require wheelchair (powered). Patient may require adaptations at home like raised toilet seat, transfer board, grab bars, long handled shoe horns, reachers, button hooks and sock aids.



**Fig 5 AFO**

Management of speech and swallowing

With speech problems, patient may require voice amplification devices and electronic aids. Oromotor exercises and swallowing by icy beverages.

Cognitive training

Use of compensatory strategies like memory book, alarms, timer, sticking directions notes on walls.

## Speech Therapy in MS:

Multiple Sclerosis is an autoimmune Disorder which is caused due to damage in functioning of the central nervous system. Although the pathophysiology seems to evolve but the inflammation to the CNS is the result of Demyelination of Axons and loss of oligodendrocytes. The resulting lesions in the brain, spinal cord, and optic nerves contribute to the clinical features seen in persons with Multiple Sclerosis are Motor impairments & spasticity along with ataxia and tremors, impaired speech and difficulties with swallowing, Weakness and limb in-coordination and other autonomic disturbances. The onset of this disorder is usually seen between the ages of twenty and forty years of age. Multiple sclerosis has a huge impact on the quality of life of an individual.

There different types are classified based on onset and progression of the disease over time are which as the name suggest is characterized by sudden episodes of new or worsening symptoms known as relapses followed by weeks, months or years of remissions. Every relapse is different and unpredictable, and the period of remission is uncertain. This the most common type of MS . Eventually over 50% of the people diagnosed with of Relapsing-Remitting MS (RRMS)within 10 years of being diagnosed usually develop Secondary Progressive Multiple sclerosis (SPMS), most of them overtime notice that there are fewer or no relapses, but disability is increasing. The symptoms seem to be progressively worsening. Only a few individuals are diagnosed with Secondary Progressive Multiple sclerosis from the onset of the disease. Only 10-15% of the persons with Multiple Sclerosis are diagnosed as having Primary Progressive MS in this type of MS its very rare to relapses or remissions the symptoms gradually worsen from the onset in these in these individuals. Overall the course of this disorder is highly unpredictable. Hence the diagnosis and treatment require a more dynamic

approach that incorporates the individual needs and provides a more personalized and customized therapeutic approach.

## Speech, Language & Swallowing in persons with Multiple Sclerosis:

Approximately over 50% Persons with Multiple Sclerosis report of having speech, swallowing and cognitive communication problems.

## Cognition and Language in Multiple Sclerosis

Similar to the other Symptoms cognitive impairment in these individuals does not follow a predictable pattern. Attention, Memory, executive functions, Information Processing and visuospatial skills are the cognitive functions most affected in persons with Multiple Sclerosis.

Executive functioning is inclusive of planning, logical reasoning. Abstraction which requires an individual to form theories about ideas and processes all these cognitive functions are essential for someone to adapt and survive. Problem Solving which is crucial for everyday decision making and strategize to solve problems which seems to be difficult for people with Multiple sclerosis.

Long term memory and working memory are the most frequently affected Memory components in persons with Multiple sclerosis, this usually result in difficulties in retrieving information and storing new information, which makes it difficult for them to instructions and procedures which in turn reduces the information processing speed.

In reference to pragmatics they find it difficulties in initiating and keep a conversation going. They have trouble expressing their opinions and make a judgement. Persons with Multiple Sclerosis find it hard to focus and respond to the communication partner in a noisy environment, also situations which requires them to multitask.

Studies show that persons with Multiple Sclerosis prefer not to participate in activities which require them to perform complex task such has formulate a message and keep track during conversations as result they tend to avoid social gatherings and events. They have problems with attention, information processing and working memory which affects their ability to maintain grammatical coherence and lexical cohesion during a conversation. They tend to rely more on redundant cues from the speaker as its difficult for them to decipher the speakers body language, subtle gesturing and inferential language such as metaphors and sarcasm etc.

## Speech

Speech and swallowing difficulties may appear at any stage of the disease. Initially, the symptoms may be mild and manageable but as the disease progresses the symptoms worsen but the change is more individualistic. In persons with Multiple Sclerosis the most common concerns with regards to speech are slurred speech, changes in loudness, they may have a harsh, hoarse or a breathy voice quality and they lack pitch control. The prosodic aspects such as rhythm, intonation and stress are usually impaired which makes their speech sound more robotic.

Weakness and decrease range of motion in the lips, tongue other muscles of the head, neck and trunk which are essential

for phonation and articulation. Dysarthria is a motor speech disorder which leads to weakness and in coordination of muscles often seen in the advanced stage of the disease

## Swallowing

Lesions along the neuromuscular pathway that originate from the cerebral region terminate at the muscles responsible for deglutition via the nucleus ambiguous and nucleus tractus solitarius are responsible for neurogenic dysphagia in individuals with Multiple Sclerosis. Dysphagia is prevalent in 30-43% of the population with Multiple Sclerosis, the commonly reported abnormalities in swallowing are difficulties in biting chewing due to weakness and in-coordination of muscles, they also have problems with controlling liquids and food getting stuck they experience episodes of coughing and choking and often dribble or drool. The trigger for the pharyngeal swallow is often delayed which could lead to aspiration especially on liquids. The laryngeal elevation during swallow is reduced which contributes to residue after swallow with a risk of aspiration due to weak closure of the airway and impaired closure of the esophagus.

## Treatment

## VOICE

### Sustained "ah"

This exercise not only helps improve vital lung capacity but also increases breath support for speech and vegetative purposes. It helps improve control the vocal volume (loudness) and intelligibility (speech clarity). The patient is asked to take a deep breath and Open their mouth wide and say "Ahh" for as long and loud as they can. To improve Loudness voice amplifiers can be prescribed. The amplifiers can be personal amplifiers worn on the body or placed in a room, such as classroom or conference room. These devices only help improve the loudness and not clarity of speech. Other techniques are the patient is asked to maintain an asked to take a deep breath before starting the vocalize and then speak while exhaling. Make sure that the sentences/phrases that are spoken are not too long. The longer the phrase the more air that is used. If the amount of air runs out, then it will require more physical effort to keep the voice loud. The pitch of the voice is what controls the variation of the highs and lows of the voice. The pitch is controlled by vibration of vocal folds

### Syllable Repetition

The patient is asked to say "ah ah ah" as loudly and clearly as they can. The patient is asked to repeat this exercise. Another technique requires the patient to Hold the vowel "e" and glide smoothly up in pitch. Alternatively, the patient can be asked to say word "whoop" and repeat this 5-10 times then they are asked to Hold the vowel "e" and glide smoothly down in pitch. Or say the word "boom" and repeat this 5-10 times.

### Speech

Pronounced -articulation- The person with multiple sclerosis is asked to over emphasize production of consonants and vowels a greater intensity then the intelligibility and accuracy of speech production can improve. The patient is also asked to open is mouth manner to improve speech intelligibility

Reduce rate of speech. Various strategies

such as pausing, pacing can be introduced in order to reduce the rate of speech. The patient can be asked to slow down in a general context during speech production or furthermore at the multi syllable level these strategies can assist in improving speech articulation in patients with dysarthria.

## Swallowing

As a speech language therapist, we can counsel the patient and their caregivers the importance of the recommend strengthening and other swallowing exercises to ensure efficient mealtimes in these individuals. It constantly becomes a challenge for the caregivers to maintain to implement safer swallowing strategies or modify food/liquid consistencies on a reliable basis, especially if the patient has significant weakness, fatigue, or cognitive disabilities. Hence most of the techniques recommended need to be easy for the caregiver to follow through. When managing dysphagia, the individual's multidisciplinary team aims to team aims to maintain adequate nutrition and supervised medical guidance and also aid in preventing aspiration.

Most commonly recommended strategies to improve swallow and feeding are diet modification which includes the changes with regards to the consistence's of solids and the viscosity of the liquids, compensatory strategies during mealtime such as postural adaptations an positioning.

Maneuvers such as the effortful swallow and Mandelson's helps to get food past the valleculae.and these Specific swallowing techniques change the swallow physiology to reduce the risk of aspiration and voluntarily protect the airway.

More postural and oral motor exercises for effortless swallow, strengthening and facilitating better function are recommended.

## Language and Cognition:

Most of these patients encounter word finding difficulties this often leads to frustration and avoidance. The key strategies to aid word finding are Delay, describe, gesture, initial syllable prompt, Synonym and association. A visual aid might often prove useful as well. It is essential to involve these individuals in conversation to maintain their communication intent, turn taking, rephrasing, reformulating is essential to aids to achieving this. Augmentative and alternate devices (AAC) not only help facilitate better communication but also motivate the speaker to communicate more descriptiveness. These AAC interfaces are inexpensive and are useful for basic verbal interactions or supplement oral speech.

Some progressively complex examples of AAC systems include:

Low tech or Analog communication options include portable resources to facilitate verbal communication, such as word boards, pen & paper, alphabet boards and picture books to help communicate basic concrete information for individuals with more urgent (High-tech AAC systems).

High end apps or devices with software computers, smartphones, and tablets that either synthesizes speech for what the user types into the keyboard or generates a pre-programmed audio message when a specific picture/icon or series of icons is selected on-screen.



Fig 6. AAC Device

## PSYCHOLOGICAL ASPECT IN MULTIPLE SCLEROSIS

Multiple sclerosis is an extremely unpredictable and complex disease as no two patients suffering from the disease have the same psychological effect. Numbness, fatigue, pain, visual disturbance, parenthesis, muscle weakness, muscle spasm - these troubling symptoms are invisible to other people as compared to a cane or a wheelchair. This makes multiple sclerosis like an iceberg with a huge hidden part that often causes intense mental anguish.

The significant stress of being diagnosed with chronic illness can trigger off psychological issues such as depression and anxiety. When behavioural changes are addressed, the changes are described simply as emotional reaction to the life situation or poor adjustment to chronic illness. Patients with multiple sclerosis don't go through a definite orderly set of stages culminating in adjustment. Rather, adjustment is an ongoing, life process that flows through the unpredictable changes brought about by the disease.

### Psychological Process and Problems:

**Emotional Issues:**

Major Depressive Disorder: Depression is the most common psychological symptoms in multiple sclerosis. The incidence rate in multiple sclerosis is three times that of the general population. The 12- month prevalence of major depressive disorder among persons with multiple sclerosis is 15.7%, nearly double the prevalence of major depressive disorder in persons without multiple sclerosis (7.4%). Reports of the lifetime risk for major depressive disorder in multiple sclerosis populations have ranged from 27-54%. It could be very difficult to diagnose multiple sclerosis as there are many symptoms that are seen in both like fatigue, weight loss, etc. Also, suicidal ideations are commonly seen in patients suffering from multiple sclerosis, occurring7.5 times higher than in general in the population. The point prevalence of anxiety in multiple sclerosis ranges from 19% to 34%. In order to slower the progression of multiple sclerosis patients are put on various medications such as Baclofen, Dantrolene, Betaseron and Avonex etc. these could cause changes in mood in either direction. Also, as some of them are on steroids, it could also lead to periods of hyperactivity and euphoria, which could be followed by a low feeling period. Also, certain steroids can cause or worsen depression. Causes of depression in multiple sclerosis patients is due to psychosocial challenges, maladaptive coping, brain lesions resulting from multiple sclerosis disease in process and immune deregulation associated with multiple sclerosis exacerbation.

Studies suggest that 13% patients with multiple sclerosis also have bipolar disorder. Bipolar may be an initial symptom of multiple sclerosis, preceding other neurological symptoms. While manic episodes in multiple sclerosis

may be precipitated by steroid therapy, there is increasing evidence that affective disturbance may be due to organic changes in the brain. Both euphoria and pathologic weeping or laughing in multiple sclerosis patients may be due to lack of emotional control in multiple sclerosis.

Treating depression in MS is often rewarding and drugs in the Specific Serotonin Reuptake Inhibitor (SSRI) group are probably safest. Common side effects, usually transient, include nausea, sexual dysfunction and gastrointestinal disturbance. Withdrawal from these drugs should be gradual, especially with paroxetine (Seroxat).

**Euphoria:** This is a mental condition where there is a feeling of happiness, confidence, or well-being sometimes exaggerated in pathological states as mania. It can often lead to having unrealistic expectations and thought process. It is seen that patients in this state lack insight. Cognitive problems like inattention, disturbed thinking, etc usually trigger euphoria.

**Emotional Liability:** Emotional liability means that the patient may easily burst into tears, both at happy and sad events or might become suddenly very angry or aggressive which may be inappropriate or exaggerated for the situation. This is different from pathological crying or laughter. In patients with multiple sclerosis, the symptom of emotional liability may vary from mild to severe.

**Lack of Insight:** Patients with multiple sclerosis may lack insight about their condition and this could cause a lot of problems. Lack of insight may lead to problems in day-care or rehabilitation. For example, when a patient suffering from multiple sclerosis may try to regain walking, when everybody around would be aware that he or she may not be able to walk again. This is when the caregivers and the therapists may face a problem as they would be in a soup whether to encourage or discourage the patient.

There could be a number of emotional aspects that a patient may go through like anger, isolation, loss of control and dependence, self- doubt and dislike which is discussed in the chapter of psychological rehabilitation.

**Cognitive Deficits:**

As multiple sclerosis is a disease that causes damage to nerve fibers in the brain and spinal cord because of which demyelination occurs in the brains of most patients with multiple sclerosis. A neuropsychological point of view suggests multiple sclerosis often affects higher order cognitive functioning; typically causing deficits in speed of information processing, attention, executive functions, verbal fluency and memory were clearly associated with less brain volume, and with higher lesion loads, in particular at frontal and occipital areas. There has been research that between 45% to 65% of all people, with multiple sclerosis experience problems with memory, attention, problem -solving and other cognitive functions, which may vary from one patient to another.

Problems in neurocognitive functioning in multiple sclerosis are mainly modulated by speed and stability of speed processing; in particular when attention-demanding controlled information processing is required. Cognitive difficulties can be

particularly stressful for people because they are changes that nobody sees. Understanding the type and the source of cognitive problems is the first step for patients and their family members to begin coping with these symptoms effectively. The cognitive issues may vary from patient to patient. The range of cognitive dysfunctions which can be associated with multiple sclerosis:

**Cognitive Fatigue:** Research shows that patients with multiple sclerosis even if they have no other cognitive problems, they can get fatigued from doing tasks that are mentally challenging work, even if they aren't exerting themselves physically. Patients with multiple sclerosis once they are cognitively fatigued, they are more likely to work slowly and make errors than people who do not suffer from multiple sclerosis.

**Attention and Concentration:** Problem arises when patients suffering from multiple sclerosis are asked to focus on more than one thing. Often patients as their disease progresses, they feel that they can work on one thing at a time, in a quiet and distraction free place whereas, before they could multi-task quiet easily.

**Processing Speed:** Cognitive slowing in individuals with multiple sclerosis (MS) has been documented by numerous studies employing explicitly timed measures in which speed of responding is an obvious focus of task performance. Often the patients experience an overall slowing the way they process information as compared to before.

**Memory:** Memory is a complex neuropsychological function. There are two separate systems for memory:

1. Procedural memory is the memory for how to do things. Procedural memory guides the

processes we perform and most frequently resides below the level of conscious awareness. When needed, procedural memories are automatically retrieved and utilized for the execution of the integrated procedures involved in both cognitive and motor skills. In multiple sclerosis, this almost always remains intact and unaffected, so although you may forget to do something, you will not forget how to do it.

2. Semantic memory needs several steps to occur before a memory can be stored or recalled, and interruption at any stage, such as may be caused by the inability to concentrate, can interfere with the ability to store or retrieve information. This memory can be affected in multiple sclerosis. It's often seen that patients with multiple sclerosis have trouble recalling recent everyday events since the diagnosis. Some patients could also experience difficulty recalling things that they see but have no trouble recalling things that they have heard, while others can remember things that they see with clarity but are unable to remember what they have heard. In very advanced cases, memory problems could be as severe as to completely disrupting the learning and recall process.

**Language: In patients with multiple s**clerosis they could be issues related to language fluency. Especially, there could be difficulty in finding words i.e. accidentally saying the wrong word and problems with naming i.e. problems with recalling uncommon words or the names of acquaintances. Patients with multiple sclerosis can also have problems with

dysphagia or dysarthria, which refer to problems coordinating the movements of the lips, mouth, tongue, and throat. They also suffer from slurred speech or physical difficulty saying certain words or syllables, without any cognitive problems at all.

**Problem Solving:** At times, people with multiple sclerosis have difficulty solving problems in new situations or they may have difficulty making decisions because it is hard to identify which of a number of choices is the best.

**Visual Spatial Skills:** Patients may find themselves getting lost more frequently, losing items, or having trouble understanding visual-spatial information like maps, diagrams, and charts.

Evaluating the Cognitive Changes: The first step to dealing with cognitive changes is to recognize that they have been changes. Usually patients with multiple sclerosis do not take these changes quiet seriously. Adequate assessments of cognitive deficits need to be carried out by a psychologist or a neuropsychologist with the help of standardized neuropsychological tests. It is also important to evaluate depression, stress, anxiety which may be a contributing factor to cognitive problems with the help of scales like Beck's Depression Inventory, Hamilton Anxiety Rating Scale, etc. An overall assessment would help the psychologist formulate a good rehabilitation plan like vocational rehabilitation, improved family life, cognitive rehabilitation, etc.

## Treatment for Cognitive Deficits:

There is as such no "pill" which could improve the cognitive functions in multiple sclerosis. However, there is a class of drugs called potassium channel blockers (4-aminopyridine (4-AP and 3, 4 diaminopyridine) have been studied for several years. The potassium channel blockers increase the speed of nerve conduction in demyelinated nerve fibers, suggesting that they might improve neurologic function and cognition. However, these drugs lead to high risk for seizures as they speed up the nerve conduction. A recent study of 69 patients suffering from multiple sclerosis with memory deficits showed that Aricept had modest benefits for verbal memory i.e. the ability to remember a list of words. Five drugs are now approved by FDA of which Avonex has demonstrated to be the most positive benefit on cognition. However, none of the medications have shown dramatic benefits, on slowing the progression of cognitive dysfunctions or reversing the effects.

**Cognitive Rehabilitation:** Cognitive rehabilitation is a systematic program designed to improve cognitive functions. There are a few strategies mentioned which are used during a cognitive rehabilitation session:

- If the patients complain of weak memory then substitute it for an organizer for example maintain a dairy where one needs to write down everything down, or maintain planners and set up sections for appointments, addresses, phone numbers, driving directions, etc.

- Maintain particular place for everything as this would help to avoid misplacing things.

Try to focus more attention on things that are important and keep repeating

it to yourself so that it registers in the long-term memory. Meditate regularly to improve attention and concentration.

- Work slowly and the focus should be on accuracy rather than speed as a patient with multiple sclerosis suffers from poor processing.

**Sexual Issues:**

Sexuality is an integral part for each one of us and it has an impact on our identity, personality, interpersonal relationship and our life span. Multiple sclerosis is a disease which can target any part of the nervous system like if it affects the nerves servicing the reproductive organs, which in turn could lead to changes in sexual functioning. Multiple sclerosis can cause problems for the patients on two aspects: neurologically and psychologically. Neurologically, the sexual excitement and response arises in the central nervous system which is normally affected. While the brain sends messages to the sex organs through spinal cord but due to multiple sclerosis the affected area is the nerve pathways which is caused by demyelination. This can cause impairment in sexual functioning like:

- Loss of libido or sexual interest
- Slower arousal time
- Difficulty masturbating
- Changes in genital sensation
- Decreased vaginal lubrication and muscle tone
- Difficulty in erectile functioning and ejaculation
- Difficulty attaining climax

Psychologically, the patient may go through problems like bowel and bladder incontinence, weakness, fatigue, decreased sexual desire and loss of interest. Also, as physical contact and sexual positioning may become uncomfortable due to sensory changes this could lead to further impact psychologically like:

- Depression, stress or anxiety
- Performance anxiety
- Altered body image or sexual self-image
- Fear and worries about the future of the relationship
- Low self esteem
- Fertility and pregnancy issues
- Genetic factors

## How do couples regain their sex life?

1. Communication: The first step is to accept that there is a problem and that to verbalize it with your partner or the person you are able to confide in. By being open about the situation would reduce the level of embarrassment and doubt about the situation.

2. Consulting a Doctor: Discuss the side effects of medication that one takes for multiple sclerosis and medications taken for muscle relaxants and pain as this could lead to being intimate more readily.

3. Alternatives: Doctors could suggest

various alternatives.

4. Sexual Counseling is necessary as it helps the couple to come in terms with the challenges of multiple sclerosis and also come up with different alternatives. Sexual Counseling is further discussed in the psychological rehabilitation chapter.

**Family Issues:**

Families have their own style of functioning, coping and communicating. If anyone the family suffers from multiple sclerosis eventually the whole family would be involved physically, psychologically and financially. It can lead to a lot of stress and challenge to the coping mechanisms of everyone in the family.

A person who was once there for the family as financial, social and psychological support to the family members, after diagnosis may have to reduce working hours which may lead to reduces family income. Also, the person who may have been very active socially may now avoid social functions and interaction and may also need emotional support as a result of his deteriorating condition. Family members may be expected to take on additional responsibilities of the family member who is suffering from multiple sclerosis. This leads to change in functioning and family role, also people might feel that this change would be temporary which is not true, it would be a lifelong change and responsibility.

Often the family members may stop spending time on leisure activities that they would enjoy doing previously. Family members should spend some time on their hobbies or what they enjoy doing, otherwise this would lead to care-givers burnout.

Multiple sclerosis can be very challenging to a married life as when the patient is diagnosed as multiple sclerosis the couple is usually planning and working towards future goals. The couple needs to have a strong bond which would help them to come out with effective coping mechanisms. As the course, of the disease could be very unpredictable with high chances of relapse. The best possible way to deal with the situation is to forget the old ways of doing things and adapting to the progression of the disease and finding out alternatives to old ways of doing things. Work out a specific plan for determining how practical needs are handled and who will be responsible for what, when. Patience, compromise and enlisting help from other family members could prove to ease out the situation.

Hiding the diagnosis from the children would not be an idle way to deal with the situation as the children would sense something wrong. Also, they would not be able to understand the behavior of the parent suffering from multiple sclerosis like being inactive and unavailable most of the time. It could lead to misunderstanding that the parent is not interested in the child. It's better to inform and educate the children about the disease, and how the children need to cope with the situation. Children may go through a number of emotions like guilt, stress and anxiety. Let the children ask questions and if you don't know the answer, help them find those answers. Children often take pride in being able to contribute in a small way, allow them

to do so and appreciate their concern. What children need the most? Is time, love and attention, so find alternatives as to how this could be done.

**Family Therapy:** Family therapy is based on the belief that the family is a unique social system with its own structure and patterns of communication. These patterns are determined by many factors, including the parents' beliefs and values, the personalities of all family members, and the influence of the extended family. Before getting on with family therapy it is very important to evaluate the strength and weakness of how the family functions.

Psychological Tests and Battery: The below listed are discussed in detail in psychological rehabilitation chapter.

Beck's Depression Inventory - To assess depression Hamilton's Anxiety Rating Scale - To assess anxiety Wechsler's Memory Scale - To assess cognition Psychological Intervention for Multiple Sclerosis:

## The below listed are discussed in detail in psychological rehabilitation chapter.

- Individualized Psychotherapy: This allows the patient with multiple sclerosis to deal with emotional difficulties, lack of motivation, negative impact of the disability and inability to handle change in functional skills typical of this condition.
- Group Therapy: A new study at The University of Nottingham has revealed that offering emotional support to Multiple Sclerosis sufferers through group therapy sessions could improve their quality of life.
- Sexual Counseling
- Cognitive Rehabilitation
- Family Therapy

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# Chp. 9 Parkinson's Disease

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## Introduction

In 1817, a British apothecary James Parkinson published "An Essay on Shaking Palsy". He called it paralysis agitans. Several decades later, Jean – Martin Charcot (1868 – 1881) called it "Parkinson's Disease" It is characterized by the cardinal features of rigidity, brady kinesia, tremor and postural instability.

Parkinson's disease is the second most common progressive neurodegenerative condition (after Alzheimer's disease) that occurs in the elderly. It accounts 3 –8% of neurological disorders with a crude prevalence of 0.4% in population older than 60 years of age in India. In 2006, the prevalence of Parkinson's disease in Mumbai, India was 328 per 100,000 persons.

## Etiology

1. Idiopathic
2. Vascular: Artherosclerotic Parkinsonism
3. Infection: Influenza virus may cause Parkinson-like features. It is commonly called as encephalitis lethargica.
4. Due to toxicity: Exposure to certain toxins such as manganese, carbon disulfide, carbon monoxide, cyanide can cause diffuse brain damage to areas causing parkinsonism
5. Due to drugs: Parkinsonism can be caused as a side-effect due to long standing use of drugs like phenothiazines, butyrophenones, tetrabenazine.
6. Metabolic: An abnormal calcium metabolism can cause disposition of calcium in the basal ganglia causing parkinsonism.
7. Traumatic: Multiple brain damage encountered in boxers can cause punch boxers syndrome.

## Pathophysiology

The presentation of Parkinsonism arises due to disturbance in the basal ganglia. Basal ganglia is divided into substantia nigra, caudate nucleus and putamen. Basal ganglia mainly secrete dopamine, which is a neurotransmitter responsible for transmitting electrical message across a synaptic terminal of a neuron. In Parkinsonism, there is a depletion of dopamine. The cause of depletion is unknown and progressive in nature. The prognosis depends on the rate of progression. In some cases, it is slow; whereas in others, it is rapid.

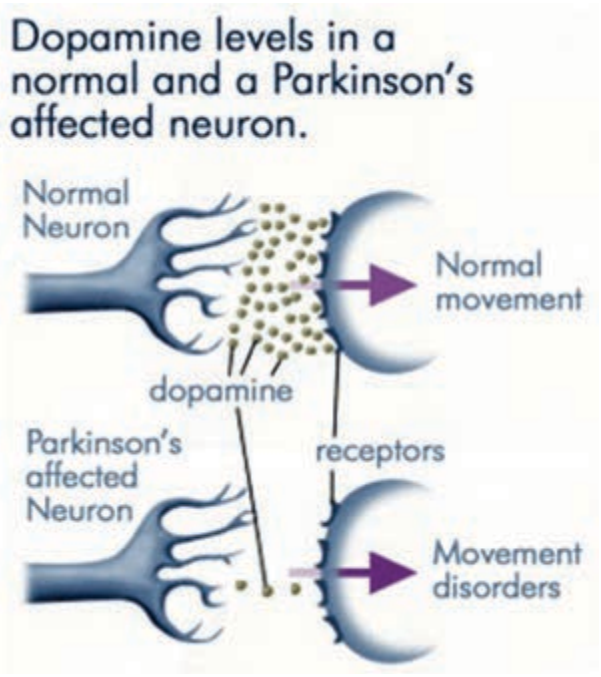



Fig 1 Dopamine levels in a normal and a Parkinson's affected neuron

The Basal Ganglia is composed of the striatum (caudate and putamen), globus pallidus, subthalamic nucleus and substantia nigra. The input from all parts of the cortex is via the striatum and substantia nigra. The output to thalamus and the cortex is via the globus pallidus and substantia nigra. There are both direct and indirect pathways.

The direct pathway disinhibits motor thalamus; and activates thalamo-cortex and motor cortex. Thus, facilitating movement. The indirect pathway inhibits motor thalamus; and inhibits thalamo-cortex and motor cortex. Thus, inhibiting movement.

Loss of dopamine results in an overactive indirect pathway causing akinesia and rigidity. It is also responsible for an underactive direct pathway causing bradykinesia.

## Clinical Presentation

**I Cardinal Features**

1. Rigidity: It may involve all body parts. It maybe in any of the three forms, which are lead pipe (resistance is constant and uniform), cogwheel (resistance is intermittent but uniform) and akinesia (inability to initiate movement)
2. Bradykinesia: Slowness of movement is seen. There is a time lag in person's desire to move and actual movement.
3. Tremor: Pin-rolling type of tremor is commonly seen in them
4. Postural Instability

**II Clinical Manifestation**

1. Posture: They usually have a stooped posture.
2. Gait: Initially, there is only lack of associated movements like swinging of arm like walking. Later, the gait becomes slow, shuffling gait with small steps. This is called as Festinating gait. Patient may also have freezing gait wherein the person walks quite well initially with relatively larger steps but eventually shuffles at the same place without proceeding further. Some may also have difficulty in stopping suddenly without external force.
3. Facial Attitude: They have a masked/expressionless face.
4. Slurred Speech with reduction in pitch / volume of voice
5. Dysphagia
6. Change in handwriting
7. Visual Difficulties: Blurring of vision, decreased blinking of eye may cause eyestrain.
8. Asthenia: Generalized weakness may be seen due to deconditioning and rigidity.
9. Psychological Impairment: Dementia and depression is commonly seen due to overdependency on others.
10. Cardiopulmonary Impairment: Deconditioning of the cardiovascular and respiratory system is common due to decreased activity and kyphotic posture.
11. Osteoporosis

## Clinical Course

The most widely used Scale is the Hoehn and Yahr Classification of Disability. It is useful in knowing the progression of the disease.

It has 5 stages, which are:

| Stage | Character of Disability |
|---|---|
| I | Minimal or absent, unilateral if present. |
| II | Minimal bilateral or midline involvement. Balance not impaired. |

| | |
|---|---|
| **III** | Impaired righting reflexes. Unsteadiness when turning or rising from chair. Some activities are restricted, but patient can live independently and continue some forms of employment. |
| **IV** | All symptoms present and severe. Standing and walking possible only with assistance. |
| **V** | Confined to bed or wheelchair. |

Table 1: Classification of disability

## MEDICAL MANAGEMENT

Treatment of early Parkinson Disease-

- Most effective anti-Parkinsonian drug- Levodopa combined with decarboxylase inhibitor (Carbidopa or Benserazide).
- Long term side effects of Levodopa- Motor fluctuations and dyskinesias.
- In the early stage of the disease, it is beneficial to delay the use of Levodopa and start treatment with other drugs like Selegiline or Amantidine or the newer dopamine receptor agonists (Pramipexole and Ropinirole).
- The golden rule of all anti-Parkinsonian drug is 'Start low and Go slow'.

| **Common drugs used in Parkinson Disease** | |
|---|---|
| Levodopa preparations | • Carbidopa plus Levodopa<br>• Benserazide plus Levodopa<br>• Controlled release Carbidopa plus Levodopa |
| Amantidine | |
| Dopamine agonists | Bromocriptine, Ropinirole, Pramipexole |
| Apomorphine | Subcutaneous injections |
| COMT Inhibitors | Entacapone, Tolcapone |
| MAO-B Inhibitors | Selegiline, Rasagiline |

Table 2: common drugs used in Parkinson Disease

| **Drug treatment of specific symptoms** | |
|---|---|
| Action Tremor | Propanolol, Primidone, Diazepam |
| Orthostatic Hypotension | Avefludrocortisone (mineralocorticoid) |
| Depression | Serotonin reuptake blocker-<br>• Fluxetine<br>• Sertraline |
| Psychosis | Bromocriptine, Amantidine, Anticholinergics, Selegiline, Levodopa |
| Sialorrhoea | Trihexyphenidyl |
| Constipation | • Anticholinergics should be withdrawn.<br>• Dioctylsulphsuccinate<br>• Cisapride<br>• High fiber diet |

Table 3: Drug treatment of specific symptoms

**Treatment of Advanced Parkinson Disease-**

- Advanced PD is characterized by two types of motor fluctuations-
  - ➢ Wearing Off- Duration of benefit of each Levodopa dose becomes progressively shorter.
  - ➢ Dyskinesias- Abnormal involuntary movements of the face, trunk and limbs during the 'ON' phase treatment.
- Some procedures may benefit in Advanced PD-
  - ➢ Surgery- Deep Brain Stimulation
  - ➢ Continuous enteral infusion of Levodopa (Duodopa)
  - ➢ Continuous subcutaneous apomorphine infusion

**SURGERIES IN PARKINSON DISEASE**

- Procedures currently being used are-
  - ➢ Thalamotomy

- Pallidotomy
- Deep Brain Stimulation

- Thalamotomy- It is used for asymmetric, severe, medically uncontrollable tremor.
  - Pallidotomy-
    - Unilateral pallidotomy is used for motor fluctuations, drug induced (unilateral) dyskinesia, severe bradykinesia and rigidity.
    - Bilateral pallidotomy is not indicated because of its side effects such as speech and memory disorders.

- Deep Brain Stimulation (DBS)-
  - DBS of subthalamic nucleus- It is very effective in reducing almost all motor manifestations of PD with a high safety profile.
  - DBS of Ventral Intermediate (VIM) thalamic nucleus- It is an alternative to thalamotomy for severe contralateral tremor and essential tremor.
  - DBS of Globus Pallidus- Unilateral or bilateral stimulation of internal Globus Pallidus showed improvements in bradykinesia, rigidity and dyskinesias.

- Continuous enteral infusion of Levodopa-
  - In patients where DBS is not possible or refused, duodopa can be given.
  - Duodopa is a gel form of Levodopa which is infused completely by a pump at a constant rate directly into the small intestine through a jejunostomy tube.
  - It is advantageous in reducing motor complications of advanced PD.

## PHYSIOTHERAPY MANAGEMENT

Even though pharmacotherapy is the primary management, the role of exercise and physiotherapy should be emphasized at every stage of the disease. Rehabilitation results are better when treatment is started early in the disease process.

'BIG, FUN & NOVEL' are good words to remember when planning treatment. Exercises are more effective if performed in 'ON' phase of medication.

## GOALS

- Patient education
- Improve motor learning
- Normalization of muscle tone
- Reduce resting tremor
- Improve transfers and functional activities
- Correction of postural impairments
- Improve balance and coordination
- Improve locomotion or gait
- Improve speech
- Reduce difficulty in swallowing
- Prevent orthostatic hypotension
- Improve cognition
- Reduce depression
- Improve cardiopulmonary endurance
- Improve patient's efficiency, independence and safety

<u>Patient education-</u>

- Educate the patient and their family members regarding the disease process and importance of exercises.

Motor learning strategies-

- The principles of motor learning should be taken into consideration while planning exercises for patients with Parkinson disease.
- Large number of repetitions is used to develop procedural skills.
- Random or Blocked practice should be used.
- Advantages of blocked practice- Structured instructional set will improve movement speed and consistency. (e.g., walking patterns can be improved with focused instructions like- "swing your arm", "walk fast" or "take large steps")
- Advantages of random practice-

  It enables the patient to learn the correct schema and regulate the extent, speed and direction of the movement.

  ➢ It also helps to improve the ability of shifting attention and to access working memory.

Relaxation exercises-

- Rigidity is common in patients with PD.
- Treatment proceeds better if rigidity is decreased early in the treatment session.
- Movement throughout a full ROM is very important to prevent changes in the properties of muscles.
- Breakdown the activity into simpler sequences to reduce the effects of rigidity.
- Generalized relaxation of excessive muscle tone can be achieved by gentle, slow rocking exercises.
- Rocking exercises includes slow, rhythmical, rotational movements of extremities and trunk. (e.g., lower trunk rotation exercises, side-lying rolling, upper and lower trunk segmental rotation) (Fig 2a & 2b)



Fig 2a: Trunk rotation exercise



Fig 2b: Trunk rotation exercise

- Rocking chair or Bobath ball can be used to give gentle rocking.
- Relaxation is better achieved in sitting or standing positions because rigidity may increase in supine position.
- Relaxation is easier to achieve by following a distal to proximal progression, because the proximal muscles are more involved than the distal muscles.
- PNF techniques like rhythmic initiation can be used with specific breathing patterns for relaxation.



Fig 3: PNF pattern- Rhythmic initiation

- Guided mental imagery training, meditation, gentle yoga and tai-chi exercises are also helpful to induce relaxation.

**Exercises to reduce resting tremor-**

- Tremor is usually present at rest and is frequently reduce or eliminated by voluntary movement.
- Biofeedback and relaxation techniques may be helpful in reducing resting tremor.
- EMG-biofeedback can also be used.
- Tremors in the hand-
  - Grip strengthening exercises with sponge or rubber ball.
  - Wrist strengthening exercises with weight cuffs and dumbbells.
  - Weight bearing activities of hand- Quadruped position, pushups or planks. (Fig 4,5 & 6)



Fig 4: Quadruped exerci



Fig 5: Push up exercise



Fig 6: Plank exercise

**Functional training-**

- Transitional movement is a major problem in patients with PD.
- Bed mobility exercises-
  - Segmental rolling (Isolated upper and lower trunk rotations)
  - Supine to sit- Teach the client to roll on to the sides and lower the legs off the bed which facilitates getting out of bed. (Fig 7a &7b)



Fig 7a: Supine to sit



Fig 7b: Supine to sit

- Pelvic mobility exercises like anterior and posterior tilts, side to side tilts and pelvic clock exercises should be encouraged to facilitate sitting posture.
- In sitting position weight shifts, upper trunk rotations and reach-out activities can also be given. (Fig 8a & 8b)



Fig 8a: Reach out in sitting



Fig 8b: Reach out in sitting

- PNF patterns can be used in sitting position to enhance trunk mobility. (Fig 9 & 10)



Fig 9: PNF pattern- Stabilizing reversal



Fig 10: PNF pattern- Joint approximation

- Sit to stand- (Fig 11)
  - Initial rocking forward and backward should be given to induce relaxation.
  - Verbal cues can be used.
  - Instruct the client to come at the edge of the seat with both the feet touching the ground.
  - Ask the client to bend forward and slightly flex the knee beyond $90^{o}$.
  - Client puts weight on both the feet and attempts to take the seat up.

Fig 11: Sit to stand activity

- Standing position-
  - Standing with upper extremities extended forward on a wall (shoulder flexion to $90^{o}$ and hands weight bearing) improves initial stability in standing.
  - Full upright position with symmetrical weight bearing over the base of support (BOS).
  - Weight shifts and rotational movement of the trunk should be encouraged. (Fig 12)



Fig 12: Weight shifts in standing position

- Movements like step-up and step-down, side stepping should also be practiced.(Fig 13)



Fig 13 a



Fig 13 a



Fig 13 a

Fig 13: Step up and down activity

- Facial muscle training-
  - ➢ Exercises like massage, PNF, manual contact and verbal cueing may enhance facial movements.
  - ➢ Facial movements (e.g., frowning, smiling, lip pursuing, etc.) and tongue movements can be performed in front of a mirror to provide visual feedback.
- Functional training in advanced PD-
  - ➢ If getting up from a chair becomes too difficult, chairs with seat that lift up have been used effectively.
  - ➢ Beds with a head that can be raised electrically may be helpful as the disease progresses.

**Flexibility exercises-**

- In Parkinson disease the contractile elements of flexors become shortened and those of extensor surface become lengthened.
- Exercises should focus on strengthening the weak elongated extensor muscles and lengthen the shortened tight flexor muscles.
- Chin tuck exercise-
  - ➢ This is used to decrease forward head posture.
  - ➢ Instruct the client to sit up as straight as straight as possible and move the head and neck up and back, bringing the ears into alignment over the shoulders.
- Scapular retraction-
  - ➢ This is used to decrease rounding of the shoulders.

- Bend the elbows at the side and then pull them back behind the body. As the arm move back, pinch the shoulder blades together.

- Thoracic extension-
  - The goal of thoracic extension is to decrease rounding of the upper back (kyphosis).
  - To perform this exercise, sit in a chair that has a low back and will act as a hinge point for the spine.
  - Clasp both the hands behind the head and wide open the elbows.
  - Lean up and back over the chair and tilt the head up towards the ceiling.

- Stretching and joint mobilization will reduce tightness of joint capsule or of ligaments around a joint.
- Passive positioning will improve flexibility and stretch the tight muscles and soft tissues. (e.g., position the patient in prone lying)

**Balance and Coordination training-**

- Client should be instructed to practice balance exercise at the early stage of the disease.
- Equilibrium reactions in all planes of movement and under different conditions should be encouraged.
- Techniques to increase dynamic balance control should be encouraged- weight shifts, reach outs, axial rotation of head and trunk.
- Therapist should give appropriate verbal, tactile or proprioceptive cues for postural awareness to facilitate the desired responses. (e.g., Instruct to 'Sit Straight' or 'Stand Straight' or a mirror is used to provide feedback)
- Sitting on inflatable disc or Bobath ball and giving weight shifts or reach outs may improve balance.
  - Challenges to balance in standing can be introduced by-
  - Varying arm positions (arms out to side, arms folded across chest)
  - Varying foot/leg position- feet apart, feet together and tandem standing
  - Adding voluntary movements- Arm clapping, arm overhead, single leg raises, head and trunk rotation
  - Adding stepping or marching place
  - All balance strategies can be addressed in a variety of environmental conditions- (Fig 14)
  - Visual inputs (reduce lighting, eyes closed)
  - Altering the support surface (standing on foam, balance board)
  - Variable open environment



Fig 14: Balance board exercise

- Frenkel exercises can be performed to improve coordination.

- External perturbations are mostly contraindicated for patients with PD because perturbations can increase the postural stiffness and fixation.
- Kitchen sink exercises are also prescribed to improve balance-
  - Toe raises
  - Heel raises
  - Mini- squats
  - Walking on the spot
  - Alternate leg out and in
  - Alternate leg behind
  - Sit to stand
- Some of the programmed exercises to improve balance are-
  - Sensory attention focused exercises (PD SAFEx)
  - Tai-chi exercise
  - Zumba exercise
  - Virtual reality or computerized games

**Locomotor training-** (Fig 15)

- Addition of external sensory cues that are timed with step initiation or step maintenance helps to improve gait pattern in Parkinsons.
- The external sensory cues may be in the form of tactile, auditory or visual modalities.
- Both single and multiple cues can be given. (e.g., Verbal cues- Walk fast, take large steps, walk and swing both the arms, cues with the help of a metronome)
- Cues were provided in a variety of tasks including walking with dual tasks, walking sideways and backward.
- Freezing episodes-
  - Visual stimuli- It includes the use of lines on the floor and stair climbing. Parallel lines are most effective and do not place the line too close together.
  - Rotational stimulation or trick movement; such as, dropping a tissue that the client must step over.
  - Client watches the movement of a person walking beside him, the movement of that person's feet encourages the client to move his/ her own feet.
  - Canes are useful for patients who fall because of freezing.
- Trunk-dissociation exercises
- Strength training
- Compensation strategies for overcoming obstacles- Programs that emphasized whole body movement and trained anticipation obstacles (e.g., hurdle crossing).
- Increasing the magnitude of the step or the amplitude of the movement appears to be the most important component for improvement in gait.



Fig 15a



Fig 15b



Fig 15c

Fig 15: Gait training

- In severe PD gait training can be given using body-weight supported treadmill to improve upright posture, walking speed and stride length. It also increases safety and allows the therapist to control speed of the movement.
- Marching in place.
- Side walking progress to cross-step walking.
- Stair climbing and ramp walking.
- Vary environmental demands-
  - Altering the support surface (e.g., Walking on a tile floor, on carpet, on grass, etc.)
  - Stepping on and off on varied surfaces (e.g., Foam).
- Practicing walking with different stride length, because in the real-world step length and speed must change with environmental demands.
- Walking with performing math problems or walking with a glass of water on a tray will improve attention and cognition.

**Measures to prevent orthostatic hypotension-**

- Plenty of fluids should be taken to keep oneself hydrated and to prevent symptoms of low blood pressure.
- Exercise the calf muscle before sitting up.
- When getting out of the bed, sit on the edge of the bed for a minute before standing.
- Instruct the client to get up slowly.
- Avoid bending at the waist to pick up something from the floor.
- Use of compression stockings and abdominal binders.
- Elevate the head end of the bed.
- Move the legs while standing.
- Tilt table training is useful for the clients with severe orthostasis.

**Cardio-Pulmonary training-**

- Diaphragmatic breathing exercises.
- Chest expansion exercises.

- Trunk extension is important to improve breathing patterns in clients with postural kyphosis.
- Aerobic training program-
  - Warm-up period- Seated activities for upper extremities and combination movements (e.g., Reach with the arms and touch the opposite foot).
  - Exercise period- Standing and marching activities that includes coordinated movement of arms and legs as well as balance and trunk rotations.
  - Cool-down period- It allows the clients to practice fine motor coordination activities of the hands.
  - Training modes can also include-
    - Lower extremity Ergometry
    - Upper extremity Ergometry
    - Walking
  - Intensity of the training should be less than normal or submaximal.
- Post exercise fatigue can be relieved by a gradual and extended cool down period.
- Aquatic therapy and swimming can also improve strength and cardio-pulmonary endurance.

**Adaptive and Supportive devices-**

- Use of ambulatory aids like walker, walking stick, or canes are helpful for the client who is able to walk with a heel-toe gait pattern but lacks postural stability.
- Four-wheel walkers with push down brakes are also helpful for some patients.
- Height of walker or cane should be adjusted to promote trunk extension.
- Equipment such as raised toilet seats and grab bars are also beneficial in making the movement fluid, efficient and safe.
- Other assistive devices include devices for walking, reaching, rising from bed and performing ADL's.
- Patient's home environment should be studied and modified according to the need of the patient to permit improved mobility.

**Home Program-**

- Home program should encourage moderate, consistent exercise as a part of normal daily routine.
- Home exercise program is important to improve the effects of deconditioning by maintaining range of motion, deep breathing and as much activity as possible.

## Occupational Therapy Management in Parkinson's Disease

Management of Triad of Parkinsonism:

**Bradykinesia**: The traditional method to reduce bradykinesia is to give rhythmical gross movements to the patient. The Recent literature review provides us with new approach to address bradykinesia.

**Motor Skills Practice**: Bradykinesia is one of the cardinal signs of Parkinson`s disease. Bradykinesia affects ADL to such a great extent that there is delayed reaction time and because of that ADLs take more time to accomplish than the period before the onset of disease, this is indeed a matter of concern. Motor Skills Practice proves to

be an effective exercise program to reduce reaction time. Motor skills practice can simply mean practice of motor tasks which are skillfully used in ADLs. Motor skills may include gross motor and fine motor task or activities Motor skills practice is practice of the basic motor components of a task.

Various study quotes that making Parkinson patient to perform a task, it helps reduce bradykinesia. Bradykinesia leads to increase in reaction time Hamid Rostami et al Department of Occupational Therapy (2009) conducted a study titled- "Effects of motor skill practice on reaction time and learning retention in Parkinson's Disease''- revealed significant reduction in RT after 1 week of practice. This reveals that practice reduced bradykinesia which in return reduced reaction time. Smiley-Oyen et al conducted a study in 2012- They concluded that short-term practice (one week in our paradigm) is sufficient for decreasing movement time, but more extensive practice is needed to improve consistency of rapid aiming performance for people with mild to moderate PD. Thus, it suggested that a study which includes more practice over a long duration of time is required. Study conducted by Susanna summa et al reveal training induced reduction of bradykinesia. Limitation of these study states that the motor skill included cannot be generalized to real life tasks such as ADLs. Also, another study conducted by platz brown concluded that practice can reduce bradykinesia and suggested to investigate the effect of motor skill practice on real life tasks such as ADL

**LSVT BIG Exercises:** The LSVT BIG program uses specific techniques to make big, whole-body movements over and over. It is created by Lee Silverman Voice Treatment.

Throughout therapy, patients perform exercises like:

Lunging far out to the side

- Sweeping their arms forwards and backwards

Rocking back and forth from heel to toe

- Taking large steps

LSVT BIG exercises increase range of motion and cause patient to use all their muscles, from head to toe.

The intensive therapy can:

- Improve flexibility
- Help patient take bigger steps, speeding up walking
- Strengthen abdominal muscles, which improves balance

This will in turn improve gait, self-care activities and leads to better quality of life.

**Activities to reduce Tremors:** Tremors present in Parkinson's Disease are resting tremors. Very rarely Intentional tremors are present. Weight bearing activities are beneficial to reduce resting tremors. As a compensatory strategy we can suggest the patient to tie weighted cuff to wrist and perform functional activities like reach outs or fine motor activity.



Fig 16: Wearing weighted cuff to wrist and perform functional activities to reduce tremors

**Activities to reduce Rigidity*:***

Passive mobilization to maintain the tone of muscles and prevent them is beneficial. Also, slow rhythmical movements may reduce rigidity.

**Activities for mask face**:
Ask the patient to perform different facial reactions to whatever emotions the therapist will ask him to imitate. This exercise will be beneficial when done in front of a mirror. (Fig 17)




Fig 17 Activities for mask face

**Activities for Monotonous Speech**: Therapist can ask the patient to read aloud books or a small dialogue wherein he is supposed to express each character's emotions through sound.

**Hand writing Training**: Micrographia is another clinical feature of Parkinson's Disease. Handwriting training can start with making big shapes big alphabets by tracing and then gradually and appropriately reducing the size.

**Eye hand co-ordination activities**: These activities include table top as well paper-pencil activities.

## Encouraging Independence in Activities of Daily Living:

**Eating:**

Due to tremors and poor grip patient faces difficulty to feed himself.

1. To stabilize the tremors a weighted cuff at wrist can be added while eating
2. The plate/bowl can be stabilized with the help of a damp napkin placed beneath so that the plate is stabilised and the food does not spill. A non-slip mat or a suction mat can be advised
3. There are more chances of spilling while having liquid food for eg. Water, juices etc. 2-handled plastic mug, angled beaker or a flexible straw can be used.
4. In the presence of swallowing difficulty, a thick consistency food is advisable



Fig 18 Modifications in eating

**Dressing:**

1. Tremors and bradykinesia increase due to performance anxiety hence the caregiver should be advised to let him dress himself at his own pace.
2. To avoid fastening of blouse or shirt-large buttons, or Velcro can be stitched to the garments

5. Loose and large sized clothing is advised for the ease of getting up quickly
6. A slip-on foot wear is advisable with a firm grip material to avoid slipping of footwear and fastening the sandal/ shoes
7. A stitched pleated saree can be used to avoid time taken to pleat the saree as it requires lot of hand manipulation skills



Fig 19 Modification in dressing

**Bathing:**

8. Non skid mats are advisable to prevent anticipatory falls
9. Grab bars can be attached in bathroom can be attached for a better safety in the bathroom
10. Often due to tremors the patient complains that the soap slips down in such a case a liquid soap is advised or a soap mitt, soap chain around the neck makes the task of applying soap to the body a much easier way





Fig 20 Bathroom modifications

**Toilet activities:**

1. Grab bars can be attached in toilet can be attached for a better safety in the toilet
2. A raised toilet seat with support handles is advisable in case of difficulty in getting up from chair
3. A cut- out stool or chair can be advised as it is cost effective if the toilet is Indian style.

4. For bowel/bladder incontinence, adult diapers and retention bags which can be worn inside the pants alongside his length is useful



Fig 21 Toilet modifications

**Grooming:**

Shaving becomes unsafe due to frequent cut on cheeks due to tremors. Hence, an electric razor can be advised, sitting in front of a half-sized mirror and shaving with weighted cuffs tied to wrists is useful to reduce tremors and provide safe shaving

**Kitchen activities:**

1. Open space is required for ease of movement and work surfaces should be planned so that carrying is reduced to a minimum.
2. While cooking potatoes and other vegetables, a wired basket placed inside the pot eliminates the need to drain boiling water/oil.
3. A medium sized stool with castors is very useful in carrying articles and reduce the back and forth trip to the kitchen. The lowest shelf of this modifies trolley can be turned side on and can be used as a table for cutting vegetables.
4. Wall mounted bottle /jar opener is useful
5. A fully automatic washing machine can assist with laundry

**The expression of sexuality**:

Tremors, bradykinesia and rigidity can make even the simplest gestures of affection functionally difficult, tiring and lacking in spontaneity. The occupational therapist can help to promote self esteem by encouraging the patient's interest in his/her own appearance and by adopting a friendly, supportive and respectful manner towards the patient

**Family counselling:**

1. The clinical feature of masked face can be misunderstood as disinterest boredom or lack of understanding on the part of the patient. The family members should be counseled regarding this feature and their cause
2. Periods of short, regular conversation are likely to be effective than lengthy exchanges.
3. Prompt referral for speech therapy should be done in cases of swallowing, speech problem, etc
4. The patient might show features of dementia and other old age problems like osteoporosis, care should be taken by the family members to avoid any bedside falls or slips
5. The medicines can cause depressive features as on of the side effects, the care givers should be counseled o check for the symptoms of depression and give a prompt referral to the treating neurologist, also the family members being supportive and trying to engage him in his leisure activity in is day schedule will help him keep motivated

## Speech Problems in Individuals with Parkinson's Disease

Parkinson's disease may affect one or all components of the speech production mechanism which include respiration, phonation, resonation or articulation.

With regards to communication the most common symptoms observed speech problems are weak, hoarse, nasal or monotonous voice overall a poor voice quality, misarticulation, Increased or Reduced rate of speech and impaired intonation and prosody.

A few of them also experience dysfluencies while speaking. Dysfluencies are often noted at the beginning of an utterances or after a pause. The dysfluencies are characterized by phoneme repetitions.

Individuals with Parkinson's usually present with dysarthria, which can be defined as group of speech disorders that have resulted in the disturbances in the muscular control of the speech mechanism which has been caused due the damage to the central nervous system. The rate of speech widely varies in persons with Parkinson's, some of them may have reduced rate of speech and the people with hypokinetic Dysarthria usually have a speech rate that is faster than normal which could be attributed to imprecise articulation and mono pitch which gives a sense of increased rate of speech.

The most obvious deviance is seen are mono-pitch, mono-loudness, and decreased loudness. Their voice quality usually perceived to be breathy and harsh. Bowing of the vocal folds and incomplete closure during phonation are often observed which leads to unstable phonation and lowered pitch. Patients with Parkinson's have hypernasal voice quality.

Persons with Parkinson's disease have inefficient and variable chest wall movements during speech. They have a lowered lung volume capacity.

## CONGNITION AND LANGUAGE

In addition to the speech problems these individuals also suffer from cognitive impairment which directly effects their language comprehension, problems with spontaneous speech , difficulties in generating verbs , they usually have problems with expressing themselves verbally, word finding difficulties are also very common in these individuals .People with Parkinson's usually tend to use more simple sentence structures , they tend to use more simpler word forms such has nouns & adjectives compared to auxiliaries, preposition and determiners. They frequently pause while speaking and use fillers while speaking.

## Swallowing Problems in Individuals with Parkinson's Disease

Dysphagia can be defined as inadequacy or unsafe transfer of solid, liquids or saliva from the mouth to the stomach. The most complications reported include aspiration pneumonia, malnutrition, and dehydration. It is critical to identify and manage the swallowing abnormalities and minimize the risk of complications since they negatively impact the individual's health and quality of life.

Dysphagia in Parkinson's Disease can be evident or all of the oral, pharyngeal, and esophageal stages of swallowing. Dysphagia occurs from the early stages in these individuals, often asymptomatic

initially, with drooling being their major concern. These patients often report of dry mouth or xerostomia which could attributed to reduced production of Saliva and the Levadopa dosage.

Oral preparatory stage deficits occur most frequently in these people. Tongue pumping which is the repetitive backward and forward rocking motion is characteristic of Parkinson's disease. They also report of food pocketing and are unable to form a proper bolus. The oral preparatory stage seems to be the most affected. Although some of them also have a delayed swallow reflex, the narrowing and delayed opening of the esophageal sphincter. For some individuals drinking and eating could be a challenge since are not able to manipulate the cutlery due to restricted movement of the limbs.

In people with Parkinson's Disease the efficiency of the jaw muscles is reduced and hence in turn it affects their ability to chew and maintain adequate lip closure while swallowing. Additionally, the movements of the tongue are also affected, they find it difficult to propel the bolus to the back of their mouth. They could also experience lingual tremors, repetitive lingual pumping. The often have slow or impaired mastication.

Persons with Parkinson's disease are observed to have a delay in Pharyngeal swallow, this delay may result in aspiration. The delay pharyngeal swallow and reduced retractions of the tongue base may result in residue left over in the valleculae and pyriform sinuses. The decreased laryngeal elevation with reduced pharyngeal wall motion can result in cricopharyngeal dysfunction wherein material is unable to pass into the esophagus.

The treatment varies based on the individual needs. The management of speech and language difficulties is majorly focused on speaker oriented strategies or communication oriented strategies. The Speaker-oriented are focused on treating the underlying deficit in the speech production mechanism or teaching compensatory strategies. As for the communication oriented strategies are focused on aiding or modifying the modes of communication.

The most common program used to improve voice loudness is the Lee Silverman Voice treatment. The major goal of this technique is to help individual with Parkinson's Disease to improve their loudness, but the therapy also results in improved speech intelligibility and also improved breathing .The program has two components the LSVT LOUD and LSVT BIG . The Speech language pathologist are usually the ones that work on the form and the Physical therapists & occupational therapy.

The LSVT has 5 essentials concepts they are

1. The major focus is on voice this concept reinforces the better closure of the vocal folds and also highlights the idea of thinking loud and also attempts to improve the clarity of speech

2. This concept works on high effort, the emphasis is on to replace the slowness of muscles with a new efforts especially as the disease progresses.

3. The program focuses on extensive training sessions which are focused on increased vocal effort.

4. This program helps person with Parkinson's disease to consistently maintain vocal loudness.

5. The key is to motivate the patient and provide feedback about the improvement or the need for more improvement.

**Facial Exercises**

Many individuals with PD request face and mouth exercises to reduce the effects of rigidity of muscles in the face and to increase facial expression. Some people with PD may find exercises helpful.

Try the exercises below.

Start with 10 repetitions of each of the facial movements explained below. Each exercise should be completed with purposeful movements and sustained effort. For instance, if smiling is the exercise, try to smile as wide as you can, and hold each smile for 5–10 seconds. Make sure you continue to breathe throughout the exercises.

Try practicing in front of a mirror, so you can see your muscles work.

1. Smile – hold – relax – repeat.
2. Pucker your lips – hold – relax – repeat.
3. Alternate puckering and smiling. Pucker as tightly and smile as hard as you can. To increase the benefit of this exercise, knit your eyebrows together when you pucker, and raise the brows when you smile.
4. Open your mouth and move the tip of your tongue all around the lips. The tongue should touch every part of the lips: bottom, top and both corners. Movements should be deliberate, not darting.
5. Open your mouth and move the tongue around the gumline. Move your tongue over the back of top and bottom teeth, front of top and bottom teeth and edges of top and bottom teeth.
6. Open your mouth as wide as you can – hold – relax – repeat.
7. Say KA – a prolonged sound – as loud and hard as you can.
8. Say PA-TA-KA as loud and fast as you can.

## Language and Cognition:

Most of these patients encounter word finding difficulties this often leads to frustration and avoidance. The key strategies to aid word finding are delay, describe, gesture, initial syllable prompt, Synonym and association. A visual aid might often prove useful as well. It is essential to involve these individuals in conversation to maintain their communication intent, turn taking , rephrasing , reformulating are essential to aids to achieving this .

Augmentative and alternate devices (AAC) not only help facilitate better communication but also motivate the speaker to communicate more descriptiveness. These AAC interfaces are inexpensive and are useful for basic verbal interactions or supplement oral speech.

### Some progressively complex examples of AAC systems include:

Low tech or Analog communication options: include portable resources to facilitate

verbal communication, such as word boards, pen & paper, alphabet boards and picture books to help communicate basic

concrete information for individuals with more urgent High-tech AAC systems.

High end apps or devices with software computers, smart phones, and tablets that either synthesizes speech for what the user types into the keyboard or generates a pre-programmed audio message when a specific picture/icon or series of icons is selected on-screen.

## Psychological Intervention of clients with Parkinson's diseases

The psychological aspects of Parkinson's disease along with the motoric symptoms reduces the quality of life in patients. The presence of fatigue in Parkinson's disease results in the primarily body discomfort, emotional wellbeing that can be assessed by PDQ 36 form which is specific to the disease. The main contributor to reduced QoL is depression and the occurrence in the community- based samples being 20% to 70%. The progression of the disease lead to multiple motor impairments and disability in functioning of daily routine. There is greater incidence of falls that lead to the vicious cycle of having higher levels of perception of being handicapped, reflection of pessimistic outlook of the future, enhancing depression and its occurrence. In addition to the motoric symptoms and depression there are mental health is affected by decline in intellectual functioning which can be associated to dementia, memory complaints, changes in personality traits, Sensory perception deficits, poor language and decline in processing speed of information received, decline in reaction and motor skill execution. The embarrassment caused due to problems with communication, behavioral Problems leading to loos of social contact.

**Frame Work:**

In figure 22, the multidimensional construct that has been defined the characteristics of the disease as taken from the WHO model of the International classification of functional disability and health as a result of functional consequence of pathological process in the body.

The ICF could be applied in a wide range of scientific and also practical areas, e.g. social security, management of health care, prevention and health promotion on a national, but also on an international level.

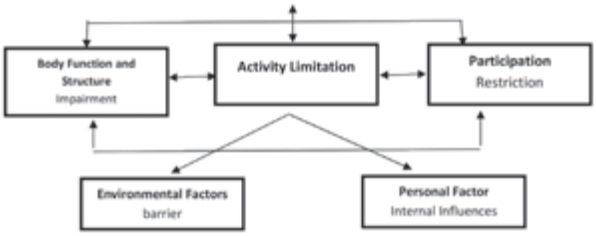



Fig 22: ICF classification

## Psychological Intervention in the treatment of Parkinson's disease:

The Physical aspects of the disease like instability, Slowness of movement, Rigidity, Balance Issues are commonly associated with Parkinson's disease. Cognitive impairment causes disability in every day functioning and cause stress to the individual client and the family. The following are the cognitive problems associated with Parkinson 's disease.

Cognitive changes range from mild problem till dementia. The test for mood, attention, Executive Function, Memory, Language, Visual Spatial Skills, behavior is evaluated for subsequent rehabilitation plan. The following are the intervention strategies used for rehabilitation of patients with Parkinson's disease.

1. Cognitive behavioral Therapy:
2. Memory Training Program
3. Community based Activity
4. Family Counselling
5. Coping Strategies

## Cognitive behavioral Therapy (CBT)

Cognitive behavior therapy is a type of psychotherapy that address behavior and thought processes that contribute to depression. Clients with movement disorders often complain of psychiatric disturbances, including mood disturbances, anxiety, and psychosis. The psychiatric disturbances are often ignored and left untreated affecting the patient's quality of life. Psychotherapies address this issue and the most widely used psychotherapy being Cognitive behavioral therapy. CBT has shown short term benefits in clients with Parkinson's diseases on depression and anxiety.

## Memory Training Program

Parkinson's disease is often associated with dementia. The cognitive decline in PD include impairments in functional skills, Executive function, performing day to activities, working memory, attention, thinking, reasoning, problem solving Various memory training program have been used to improve the cognitive abilities. Training program has proven to enhance the memory performance and reduce the decline rate in the functional skills in the instrumental activity of daily living.

## Community based Activity

Community based activity has its own benefits like socialization, bringing people together, motivation, solving individual problems, understanding the progression of the disease, importance of certain therapy or medication, knowing new trends, adherence to exercises like yoga, activities like meditation, laughter therapy. Although there aren't enough studies that individualized each activity and report their benefits. But a community based tango dance program has shown to improve the participation among individuals with Parkinson's disease.

## Family Counselling

Family play a significant role in support, motivation, maintaining independent living and as acts as a primary caregiver. Its debilitating and a difficult situation for the family to face their loved ones suffering with an incurable diseases. They face stressful and difficult situation quite often. A family counselling that brief the clinical condition and promote acceptance of the condition may lead to better care giver metal heath and reduce burden.

## Coping Strategies

Coping is the conscious effort to reduce stress and the techniques used to is termed as coping strategies. They are leaned from watching or advices taken to avoid stressful situations. A good coping skill can be developed by practice. The usual techniques are to perform progressive relaxation exercises, breathing technique, emotional regulation, diverting oneself etc. coping strategies are used by about 70 % PD patients in compared to other techniques in clients with PD with visual hallucination. The coping strategies is dependent on the stressful symptoms and emotional regulation is the most common strategy used by patients with Parkinson's disease.

## Conclusion:

Psychological intervention can promote heath related quality of life of the client and the family by providing assistance for coping and accepting a long- term disability.

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# Chp.10 Dementia

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## NTRODUCTION:

Dementia is defined as a progressive impairment of cognitive functions occurring in clear consciousness (i.e. in absence of delirium). This is a disease which has a significant global impairment of intellect, manifested as difficulty with memory, attention, thinking and comprehension. Other psychological functions can be included including mood, personality, judgment, and social behavior. There are specific diagnostic criteria that are found for various dementias, such as Alzheimer's disease or vascular dementia, however all dementias have certain common elements that result in significant impairment in social or occupational functioning and cause a significant decline from a previous level of functioning.

The disorder can be progressive or static, permanent or reversible. An underlying cause is always assumed, although in rare cases, it is impossible to determine specific cause. Approximately 15 percent of patients with dementia have reversible illness if treatment is initiated before irreversible damage takes place.

## HISTORY:

Western literature for centuries has described a disease process what we call as "dementia" today. "Dementia is a word which is given by a French physician named Philippe Pinel. In 1801 he was working on a 34-year-old woman who lost her memory, speech, ability to walk or use common objects like a fork or a hairbrush and Pinel called this process "demence". He used the words "demence" to mean an "incoherence" of mental faculties to describe her disease. Today the word dementia indicates a person having cognitive impairment significant enough to interfere with daily functioning and describe one of more than 48 types of these diseases in the brain.



**Figure 1 :French Physician, Philippe Pinel who coined with the term "demence" i.e dementia.**

When Pinel's patient died, he autopsied her brain. Using a primitive microscope, he studied the brain tissue. With his microscope, he was only able to describe two distinct features of her disease.

He wrote that the woman's brain was full of fluid and it had dramatically shrunken in size.

In 1907, a German physician published a paper on a patient of his who exhibited behaviors similar to Pinel's patient. This woman was in her fifties and she appeared to have the same disease Pinel described. The woman suffered a "failure of memory, paranoia, loss of reasoning powers, incomprehension and stupor."

When the German physician looked at her brain however, he had a more advanced microscope, an optical microscope. And in writing a research paper about his patient, Alois Alzheimer described the disease process for which he is known today. Alzheimer described a brain that was (1) shrunken and (2) full of fluid, but also (3) suffered structural damage in the form of neurofibrillary tangles and had bone structures growing in the brain tissues. These are the four hallmark features of a brain with Alzheimer's disease.

Alzheimer sent his paper documenting his findings to his mentor, Dr. Emil Kraepelin. When Dr. Kraepelin published his eighth medical textbook, Alzheimer's paper and research were included and the disease became known as Alzheimer's disease. One of the challenges for us today is the pronunciation of Alzheimer's. Some people have a tendency to pronounce Alzheimer's like the words

"Old-Timers," which continues to enforce the thought that as we age we lose our mental abilities. Alzheimer's is actually a disease and is not considered a part of the normal aging process. Most people do not develop any type of dementia. And most people remain cognizant throughout their lifetimes. Indeed, people learn at the age of eighty at the same rate they learned at the age of twenty. Dementia is truly not a part of normal aging.

## DEFINITION OF DEMENTIA:

The World Health Organization (2007): "a syndrome due to disease of the brain, usually of a chronic or progressive nature, in which there is disturbance of multiple higher cortical functions, including memory, thinking, orientation, comprehension, calculation, learning capacity, language, and judgment. However, consciousness is not clouded. The impairments of cognitive function are commonly accompanied, and occasionally preceded, by deterioration in emotional control, social behavior, or motivation which occurs usually in Alzheimer's disease, in cerebrovascular disease, and in other conditions primarily or secondarily affecting the brain."

## EPIDEMIOLOGY:

The prevalence of dementia is rising, with the growing aging population. The prevalence of moderate to severe dementia:

- 5 percent - general population more than 65 years of age
- 20 to 40 percent - general population more than 85 years of age
- 15 to 20 percent in outpatient general medical practices
- 50 percent in chronic care facilities

Of all the other types of dementia, 50 to 60 percent of patients suffer from the most common type of dementia i.e. the Alzheimer's type. Dementia of the

Alzheimer's type increases in prevalence with increasing age. For persons aged 65 years men have a prevalence rate of 0.6 percent and women of 0.8. At age 90 years, rates are 21 percent. The second most common type of dementia is vascular dementia, which accounts for 15 to 30 percent of all cases. Vascular dementia is most common in persons between the ages 60 and 70 and is more common in men than in women.

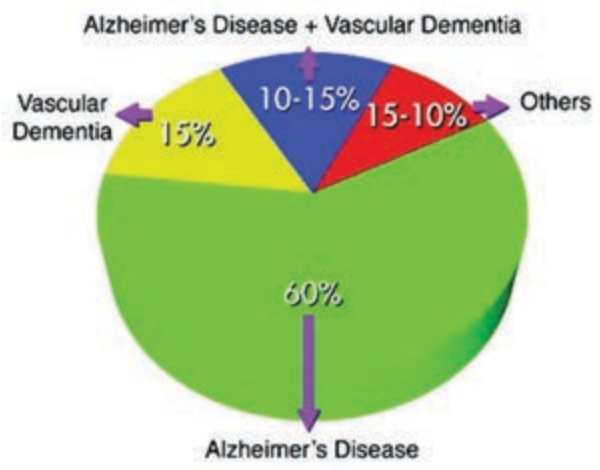



Figure 2: Prevalence rates of different types of dementia

Other common causes of dementia, each representing 1 to 5 percent of all cases, include head trauma, alcohol related dementias and various movement disorder - related dementias, such as Huntington's disease and Parkinson's disease.

## ETIOLOGY:

The most frequent causes of dementia in individuals older than 65 years of age are: (1) Alzheimer's disease (2) Vascular dementia and (3) Mixed vascular and Alzheimer's dementia. Other illness that account for approximately 10 percent include Lewy body dementia, Pick's disease, frontotemporal dementias normal pressure hydrocephalus, alcoholic dementia, infectious dementia, such as human immunodeficiency virus (HIV) or syphilis and Parkinson's disease. Many types of dementias evaluated in clinical settings can be attributable to reversible cases, such as metabolic abnormalities, nutritional deficiencies or dementia syndrome caused by depression.

## DIAGNOSIS:

Dementia is a type of chronic encephalopathy that can have many causes, including irreversible degenerative and potentially reversible nondegenerative causes. The most important diagnostic step in evaluating dementias is to determine whether a chronic encephalopathy results form a degenerative or other potentially reversible causes. Historical clues suggesting a reversible process include fluctuating severity, altered level of consciousness or hypersomnolence, and visual hallucinations. Clues on mental status testing include findings the patient to be inattentive, disoriented and somnolent but not particularly amnestic. Clues on physical examination include a variety of findings that may be common in elderly patients are not part of the typical picture of Alzheimer's disease, such as ataxia, hyper-reflexia and tremulousness. Laboratory evaluation should be directed towards.

## Dementia of The Alzheimer's Type:

In 1907, Alois Alzheimer first described the condition that later assumed his name. He described a 51 year old woman with a 4 ½ year course of progressive dementia. The final diagnosis of Alzheimer's disease requires

a neuropathological examination of the brain; nevertheless, dementia of the Alzheimer's type commonly diagnosed in the clinical setting after other causes of dementia have been excluded from diagnostic consideration. The clinically defined syndrome, Alzheimer's dementia has been considered as the paradigm of a cortical dementia syndrome. The hallmark of, cortical dementia include not only memory loss, which is common to most dementia syndromes but also elements of aphasias, apraxia and agnosia.



**Alois Alzheimer 1864-1915**

***German neuropathologist & psychiatrist who described in 1906 the clinical and neuropathological features of a woman aged 51 years, with atrophied cerebral cortex, senile plaques and neurofibrillary tangles***

Figure 3: Alois Alzheimer after whom the condition assumed his name Alzheimer 's disease.

Most clinical studies are in agreement that the earliest clinical signs of AD are memory loss, which precedes the actual dementia. Abnormal cerebral metabolism, demonstrated by positron emission tomography (PET), may precede even mild memory loss. A few studies have correlated other relative cognitive inefficiencies occurring much earlier in life with the subsequent development of dementia, although interpretation of these findings have been a source of controversy.

Recent memory is easier to access reliably then remote memory and is thought to be disproportionately severely involved. Nonetheless, remote memory is also abnormal, and there is a gradient effect regarding recall over a retrograde time interval: The oldest memories appear to be the best preserved, with proportionately greater forgetting as the retrograde interval shortens. In contrast procedural memory appears to be relatively spared. Alzheimer's patients are able to learn simple skills as easily as normal controls and better than patients with subcortical patterns of dementia or patients with various types of sensorimotor deficits. One very important skill that needs to be addressed clinically is driving. Despite the relative preservation of procedural memory, patients with mild Alzheimer's dementia have a higher rate of collisions and moving violations than age-matched controls, although estimates of risk vary, especially during the first 2 years of the disease. Whether this actually results from impaired procedural memory, attentional factors, other cognitive aspects or a combination is unclear, although visual tracking, memory, and Mini-Mental State score all correlate with a laboratory-based driving score.

Aphasia, apraxia and agnosia are the other categories of cognitive impairment that typically occur in cortical dementia syndromes and particularly in AD. In Alzheimer's dementia, these aspects do not usually dominate the clinical picture, but they can in variant syndromes. In mild to moderate stages of dementia, anomia is prominent and readily detectable with neuropsychological testing using a variety of naming tests. However, patients are fluent and may have relatively good comprehension, so clinical detection is not always easy.

Apraxia can be confused with impaired comprehension in mild to moderate stages of Alzheimer's dementia. Patients have difficulty in learning new procedures such as a new car or performing tasks that they were previously adept at. In moderately advanced stages, patients have difficulty in dressing and performing other activities of daily living.

Anosognosia is the failure to recognize illness, this is a cardinal feature of Alzheimer's dementia and this is typically present, even in mild stages of the disease. In AD, a patient may deny significant memory problems and will actively try to explain away the observations of concerned family members and friends, even to a point of becoming hostile and accusative.

Psychiatric symptoms include both affective and psychotic disturbances. Depression interferes with an individual's functional status and an accurate cognitive assessment, erroneously leading to the diagnosis of dementia; such is the nature of pseudodementia, which can be estimated to account for 4 percent or 5 percent of dementia cases. Psychotic symptoms, most commonly involves paranoid delusions and less commonly hallucination.

Sleep wake cycle disturbances are common and may be present even during relatively mild stages of their illness. They may become more common and more severe during moderately severe stages. There are two aspects to the sleep cycle disturbance. The first is the so called sun-drowning effect, which means that the patient becomes more confused, agitated, and difficult to manage during the evening. The second regards not sleeping at night, waking up during very early hours or going to sleep very early in the evening. As, the disease progresses to more advanced stage, patients may become generally less active and eventually in terminal stages, are bedbound with little apparent conscious activity. Weight loss is common during the latter stages as well. Incontinence is uncommon in mild stages but becomes increasingly frequent as the disease progresses and is universal in late stages.

## Genetic Factors:

Although the cause of dementia of the Alzheimer's type remains unknown, progress has been made in understanding the molecular basis of the amyloid deposits that are a hallmark of the disorders neuropathology. Some studies have indicated that as many as 40 percent of patients have a family history of dementia of the Alzheimer's type thus genetic factors are presumed to play a part in the development of the disorder, at least in some cases.

In several well-documented cases, the disorder has been transmitted in families

through an autosomal dominant gene, although such transmission is rare. Alzheimer's type dementia has shown linkage to chromosomes 1, 14 and 21.

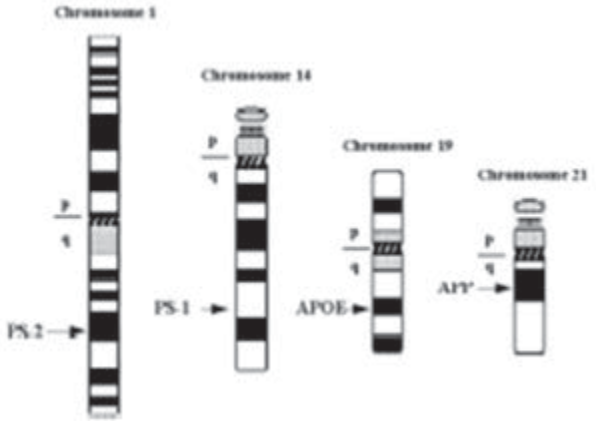



Figure 4: Genetic affect in Alzheimer's dementia

## Amyloid Precursor Protein:

The gene for amyloid precursor protein is on the long arm of chromosome 21. Whether the processing of abnormal amyloid precursor protein is of primary causative significance in Alzheimer's disease is unknown, but many research groups are studying both the normal metabolic processing of amyloid precursor protein and its processing in patients with dementia of Alzheimer's type in an attempt to answer this question.

## Multiple E4 Genes:

One study implicated gene E4 in the origin of Alzheimer's disease. People with one copy of the gene have Alzheimer's disease three times more frequently than do those with no E4gene, and people with two E4 genes have the disease eight times more frequently than do those with no E4 gene. Diagnostic testing for this gene is not currently recommended because it is found in persons without dementia and not found in all cases of dementia.

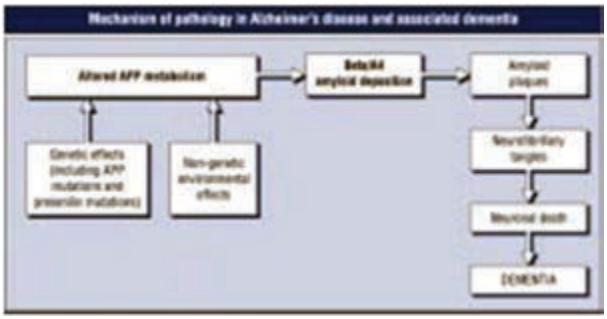



Figure 5: Mechanism of pathology in Alzheimer's disease and associated dementia

## Neuropathology:

The classic gross neuroanatomical observation of a brain from a patient with Alzheimer's disease is diffuse atrophy with flattened cortical sulci and enlarged cerebral ventricles. The classic and pathognomonic microscopic findings are senile plaques, neurofibrillary tangles, neuronal loss (particularly in the cortex and hippocampus), synaptic loss (perhaps as much as 50 percent in the cortex) and granulovascular degeneration of the neurons.

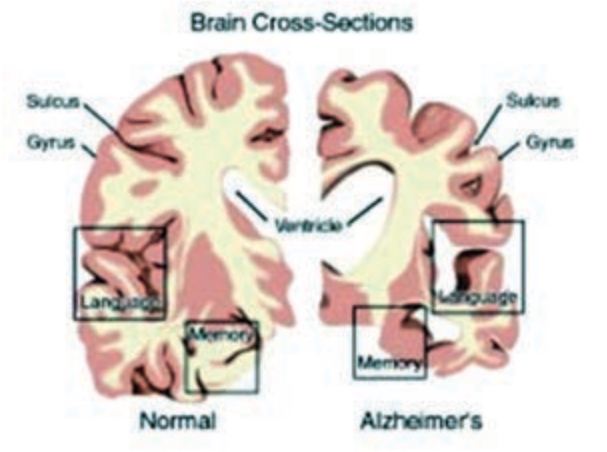



Figure 6: Neuroanatomical Observation diffuse atrophy with flattened cortical sulci and enlarged cerebral ventricles.

Senile plaques also referred to as amyloid plaques more strongly indicate Alzheimer's disease, although they are also seen in

Down syndrome and to, some extent, in normal aging. The number and the density of senile plaques present in postmortem brains have been correlated with severity of the disease that affected the person.

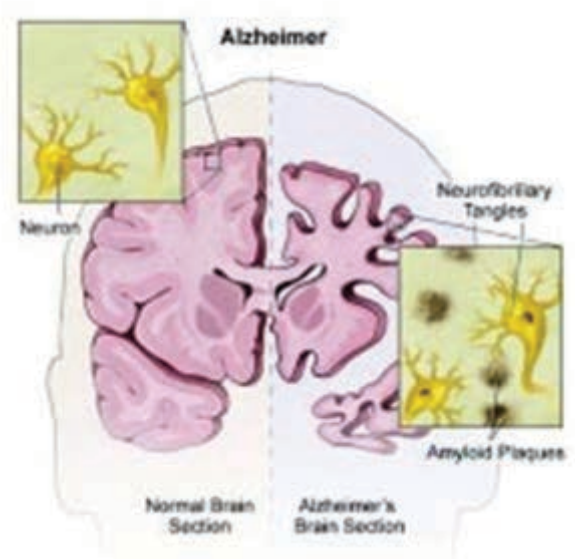



Figure 7: Neurofibrillary tangles and Amyloid Plaques in Alzheimer's patients

## Neurotransmitter:

The neurotransmitters that are most often implicated in the pathophysiological condition of Alzheimer's disease are acetylcholine and norepinephrine both which are hypnotized to be hypoactive in Alzheimer's disease. Decreased norepinephrine activity in Alzheimer's disease is suggested by the decrease in norepinephrine containing neurons in the locus ceruleus found in some pathological examination of brains from persons with Alzheimer's disease. To other neurotransmitter implicate in the pathophysicological condition of Alzheimer's disease are the neuroactive peptides somatostatin and corticotrophin; diseases concentrations of both have been reported in person with Alzheimer's disease.

## Other Causes:

Another theory to explain the development of Alzheimer's disease is that an abnormality in the regulation of membrane phospholipid metabolism results in membranes that are less fluid - that is, more rigid - than normal. Aluminium toxicity has also been hypothized to be causative factor, because high levels of aluminium have been found in the brains of some patients with Alzheimer's disease; but this is no longer considered a significant etiological factors. Excessive stimulation by the transmitter glutamate that may damage neurons is another theory of causation.

Familial Multiple System Taupathy with Presenile Dementia: A recently discovered type of dementia, familial multiple system taupathy, shares some brain abnormalities found in people with Alzheimer's disease. The gene that causes the disorder is thought to be carried on chromosome

17. The symptoms of the disorder include short term memory problems and difficulty in maintaining balance and walking. The onset of the disease occurs in the 40s and 50s and persons with the disease live an average of 11 years after the onset of symptoms.

As, in patients with Alzheimer's disease, tau protein builds up in neurons and glial cells, of persons with familial multiple system taupathy. Eventually, the protein build up kills brain cells. The disorder is not associated with the senile plaque seen with Alzheimer's disease.

## Diagnosis:

**Diagnostic criteria for Dementia of the Alzheimer's Type (cautionary statement).**

A The development of multiple cognitive deficits manifested by both (1) memory impairment (impaired ability to learn new information or to recall previously learned information) (2) one (or more) of the following cognitive disturbances:

a aphasia (language disturbance)

b apraxia (impaired ability to carry out motor activities despite intact motor function)

c agnosia (failure to recognize or identify objects despite intact sensory function)

d disturbance in executive functioning (i.e., planning, organizing, sequencing, abstracting)

B The cognitive deficits in Criteria A1 and A2 each cause significant impairment in social or occupational functioning and represent a significant decline from a previous level of functioning.

C The course is characterized by gradual onset and continuing cognitive decline.

D The cognitive deficits in Criteria A1 and A2 are not due to any of the following:

(1) other central nervous system conditions that cause progressive deficits in memory and cognition (e.g., cerebrovascular disease, Parkinson's disease, Huntington's disease, subdural hematoma, normal-pressure hydroce- phalus, brain tumor)

(2) systemic conditions that are known to cause dementia (e.g., hypothyroidism, vitamin B or folic acid deficiency, niacin deficiency, hypercalcemia, neurosyphilis, HIV infection)

(3) substance-induced conditions

E The deficits do not occur exclusively during the course of a delirium.

F The disturbance is not better accounted for by another Axis I disorder (e.g., Major Depressive Episode, Schizophrenia).

Code based on presence or absence of a clinically significant behavioral disturbance:

294.10 Without Behavioral Disturbance: if the cognitive disturbance is not accompanied by any clinically significant behavioral disturbance.

294.11 With Behavioral Disturbance: if the cognitive disturbance is accompanied by a clinically significant behavioral disturbance. (e.g., wandering, agitation)

***Specify subtype:***

**With Early Onset:** if onset is at age 65 years or below

**With Late Onset:** if onset is after age 65 years

Coding note: Also code 331.0 Alzheimer's disease on Axis III. Indicate other prominent clinical features related to the Alzheimer's disease on Axis I (e.g., 293.83 Mood Disorder Due to Alzheimer's Disease, With Depressive Features, and 310.1 Personality Change Due to Alzheimer's Disease, Aggressive Type).

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## Prognosis:

Alzheimer's disease is a progressive disorder and as the patients are elderly, and susceptible to a variety of other disease processes, AD however is not the common cause of death. However, if no other disease process supervenes, death usually ensues approximately 6 t 10 years after the onset.

## VASCULAR DEMENTIA:

The primary cause of vascular dementia is from impaired blood flow to the brain. Formerly referred to as multi- infarct dementia is presumed to be multiple areas of cerebral vascular disease, resulting in a symptom pattern of dementia. However, the name was changed to vascular dementia to reflect the array of conditions that can impair the blood's ability to circulate to the brain. Vascular dementia often occurs alongside Alzheimer's disease, resulting in mixed dementia between 1% to 4% of people over the age of 65 years have vascular dementia, and the risk of developing it increases dramatically with age. Vascular Dementia most commonly is seen in men especially those with preexisting hypertension or other cardiovascular risk factor.

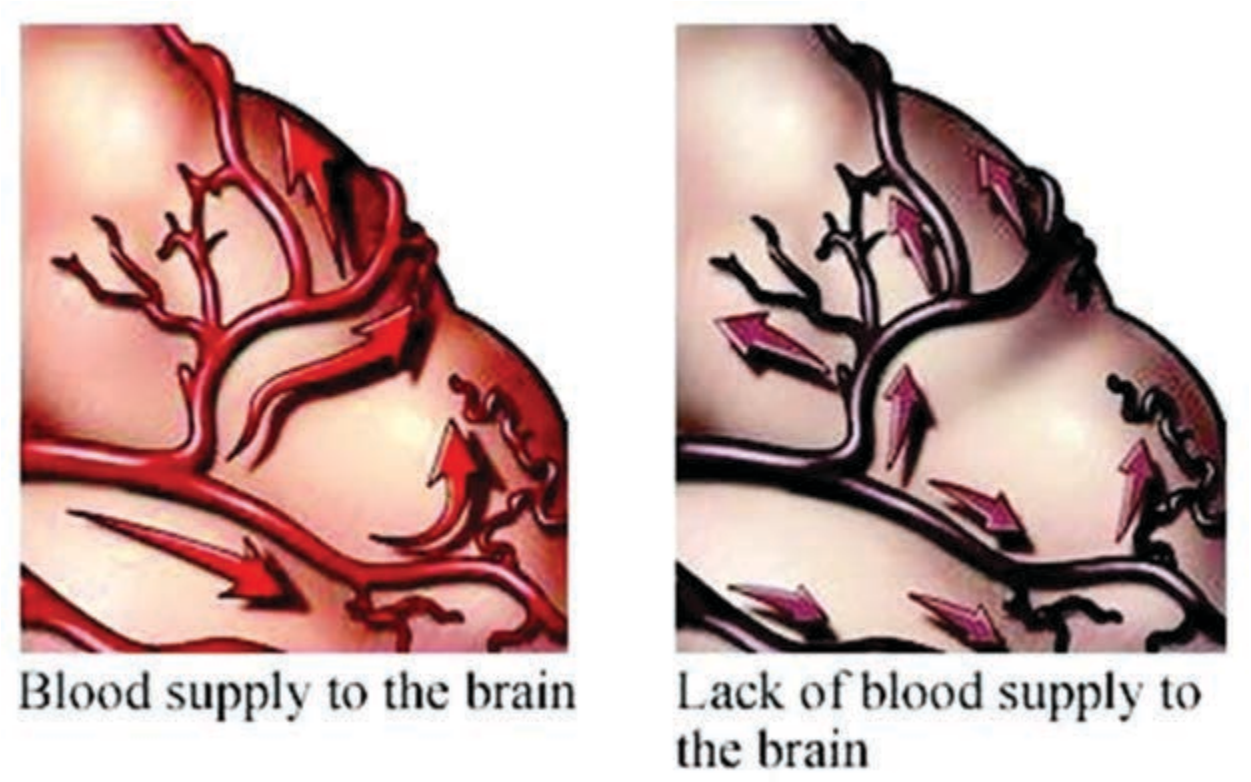



Figure 8: Impaired blood flow in the brain in Vascular Dementia

## Causes of Vascular Dementia:

Vascular dementia can occur either due to narrowing or a complete blockage of blood vessels in the brain, which deprives brain cells from nutrients and oxygen they need to function properly. Vascular dementia often results from several small strokes that occur over time. It can also occur after a single major stroke, which is sometimes referred to as post-stroke dementia. Not all strokes lead to dementia, but up to one-third of those who have a stroke will develop dementia within six months. Conditions like high blood pressure and diabetes that don't block blood vessels, but simply narrow them, can also lead to vascular dementia.

## Epidemiology and Risk Factors for Vascular Dementia:

Incidence and prevalence rates of vascular dementia are affected by the definition used. The prevalence of post-stroke vascular dementia ranged from 11.3 percent using the NINDS- AIREN criteria to 20. 1 percent using the ICD- 10 NA criteria, and the incidence ranged from 2.6% with the ADDTC to 5.2 percent with the ICD - 10 - NA[5]. The incidence rate of AD is higher among individuals with prevalent cardiovascular disease than among those without it.

Individuals who develop vascular dementia often have a history of one or more of the following: heart attack, stroke, high blood pressure, diabetes, or high cholesterol. In particular, if an individual has a history of multiple strokes, the risk of developing vascular dementia increases with the number of strokes experienced over time.

## Clinical Feature and Associated Disorders:

People with vascular dementia often display multiple cognitive problems, including memory impairment and possibly aphasia, apraxia, agnosia, or problems with executive functioning. In maximum cases this makes it difficult for the patients to have a successful career or hold a job, take care of the household activities or socialize. People with vascular dementia also experience neurological symptoms such as exaggerated reflexes, problems with walking and balance, and / or weakness in the limbs, hands, and feet. Depending on the individual and on the cause of the dementia, delusions, confusion, agitation, urinary problems, and / or depression can also accompany vascular dementia.

Patients have a more subcortical pattern of dementia with psychomotor slowing, poor learning curve, and relative preservation of naming and other language skills. The degree of impairment, however, correlates better with the degree of neurofibrillary pathology than withcerebrovascular pathology at autopsy.

Interestingly, memory loss usually occurs later in the disease compared to when it appears in Alzheimer's. In vascular dementia, the first symptoms are often the neurological ones, such as problems with reflexes, walking, and muscle weakness. On the other hand, memory problems and behavioral symptoms are commonly the first issues noticed in Alzheimer's. Additionally, vascular dementia often progresses in a step-wise fashion. For example, the person will seem stable for a period of time, then suddenly get much worse, then continue to alternate

between stable periods and sudden drops in functioning. On the other hand, Alzheimer's disease progresses in a more gradual, downward fashion.

## Diagnosis of Vascular Dementia:

As with Alzheimer's disease, a complete diagnostic workup should be performed in order to rule out other possible causes of the person's symptoms. Vascular dementia is usually identified through imaging procedures, which can reveal strokes and narrowed or blocked arteries. Neuropsychological tests might also be conducted to determine the nature and extent of cognitive impairment.

DSM-IV Criteria for the Diagnosis of Vascular Dementia

A The development of multiple cognitive deficits manifested by both:

1 Memory impairment (impaired ability to learn new information or to recall previously learned information).

2. One or more of the following cognitive disturbances:

   a Aphasia (language disturbance)

   b Apraxia (impaired ability to carry out motor activities despite intact motor function)

   c Agnosia (failure to recognize or identify objects despite intact sensory function)

   d Disturbance in executive functioning (i.e., planning, organizing sequencing, abstracting)

B The cognitive deficits in criteria A1 and A2 each cause significant impairment in social or occupational functioning and represent a significant decline from a previous level of functioning.

C Focal neurological signs and symptoms (e.g., exaggeration of deep tendon reflexes, extensor plantar response, pseudo bulbar palsy, gait abnormalities, weakness of an extremity) or neuroimaging evidence indicative of cerebrovascular disease (e.g., multiple infarctions involving cortex and underlying white matter) that are judged to be etiologically related to the disturbance.

D The deficits do not occur exclusively during the course of delirium.

## Differential Diagnosis:

There is a broad range of cerebrovascular disorders that may result in cognitive impairment. Multiple large artery territory infarction and hemorrhages may result in more extensive physical disability and cortical cognitive signs such as aphasia. Non- atherosclerotic forms of cerebrovascular disease may have their own disease associations that, in turn, may influence cognition. Cerebral vasculitis may present as a rapidly progressive fulminant encephalopathy, a sub-acute course of recurrent cerebral infarction or a more chronic dementia - like course.

## Evaluation:

Considerations are the same as that for AD except that causes of cerebral infarction also need to be included.

## Prognosis for Vascular Dementia

Currently, there is no cure for vascular dementia. If the dementia was caused by multiple strokes, the person may get

worse in a step-wise progression, where stable periods are interrupted by sudden downward episodes. Life expectancy for someone with vascular dementia is highly individual and depends on the nature of the cardiovascular problems that are causing the dementia, along with the person's age and other medical conditions.

## BINSWANGER'S DISEASE:

Binswanger's disease, also known as subcortical arteriosclerosis encephalopathy, is characterized by the presence of many small infarctions of the white matter that spare the cortical regions. Atherosclerosis (commonly known as "hardening of the arteries") is a systemic process that affects blood vessels throughout the body. Although Binswanger's disease was previously considered a rare condition, the advent of sophisticated and powerful imaging techniques, such as magnetic resonance imaging (MRI), has revealed that the condition is more common than previously thought.

### Symptoms of the Binswanger's Disease:

The symptoms associated with Binswanger's disease are related to the disruption of subcortical neural circuits that control what neuroscientists call executive cognitive functioning:

- Short-term memory,
- Organization,
- Mood,
- The regulation of attention,
- The ability to act or make decisions, and
- Appropriate behavior.

The most characteristic feature of Binswanger's disease is psychomotor slowness - an increase in the length of time it takes, for example, for the fingers to turn the thought of a letter into the shape of a letter on a piece of paper.

Other symptoms include:

- Forgetfulness (but not as severe as the forgetfulness of Alzheimer's disease),
- Changes in speech,
- Unsteady gait,
- Clumsiness or frequent falls
- Changes in personality or mood (most likely in the form of apathy, irritability, and depression), and
- Urinary symptoms that aren't caused by urological disease.

### Diagnosis:

Brain imaging, is very essential as it reveals the characteristic brain lesions of Binswanger's disease, and is essential for a positive diagnosis.

### Prognosis:

Binswanger's disease is a progressive disease; there is no cure. Changes may be sudden or gradual and then progress in a stepwise manner. Binswanger's disease can often coexist with Alzheimer's disease. Behaviors that slow the progression of high blood pressure, diabetes, and atherosclerosis -such as eating a healthy diet and keeping healthy wake / sleep schedules, exercising, and not smoking or drinking too much alcohol -- can also slow the progression of Binswanger's disease

## PICK'S DISEASE:

Arnold Pick, was the first one who described

the disease in 1892, he reported that Pick's disease causes an irreversible decline in a person's functioning over a period of years. It is most commonly confused with the much more prevalent Alzheimer's disease; Pick's disease is a rare disorder that causes the frontal and temporal lobes of the brain, which control speech and personality, to slowly atrophy. It is therefore classified as a frontotemporal dementia, or FTD.

Pick's disease is a relatively rare form of dementia that causes a slow shrinking of brain cells due to excess protein build-up. Patients with Pick's initially exhibit marked personality and behavioral changes, and then a decline in the ability to speak coherently.

## Pathogenesis and Pathophysiology:

In Pick's disease the brain tissue changes and loss occurs in focal areas rather than the generalized damage of Alzheimer's. Pick's disease affects the frontal and temporal lobes of the brain. Marked shrinkage, called atrophy, of the frontal lobes of the brain occurs that can be seen on brain scans.

Pick's disease is marked by the presence of abnormalities in brain cells called Pick's bodies. These are found in the affected areas as well as elsewhere in the brain. Pick's bodies are fibers that look very different from the neurofibrillary tangles found in Alzheimer's disease. Pick's bodies are straight rather than paired and helical.

Changes showing in the hippocampus gyrus and ballooned achromatic neurons and pick bodies

## Symptoms of Pick's Disease:

Many of the early symptoms of Pick's disease are frontal lobe symptoms. It is these symptoms that tend to mark out the differences between Pick's dementia and the other types, such as Alzheimer's. In Alzheimer's disease the initial symptoms tend to be memory impairment. In Pick's diseasebecause the frontal lobes of the brain are affected, the first symptoms occur in emotional and social functioning. It is the mood changes, often biased towards euphoria, disinhibition and deterioration in social skills that are so noticeable.

Behavioral Symptoms Include:

- Repetitive or obsessive behavior
- Overeating or drinking to excess (when this was not previously a problem)
- Impulsivity and poor judgment
- Extreme restlessness (early stages)
- Lack of attention to personal hygiene
- Sexual exhibitionism or promiscuity
- Withdrawal or decreased interest in activities of daily living
- Decline in function at work and home Emotional Symptoms Include:
- Unaware of the changes in behavior
- Apathy
- Abrupt mood changes
- Easily distracted; poor attention span
- Rudeness, impatience, or aggression
- Lack of warmth, concern, or empathy Language Problems Include:
- Complete loss of speech
- Difficulty speaking or understanding speech

- Weak, uncoordinated speech sounds
- Repeating words others say
- Decrease in ability to read or write
- Trouble finding the right word
- Loss of vocabulary Physical Symptoms Include:
- Memory loss
- Difficulty moving about
- Lack of coordination
- Increased muscle rigidity or stiffness
- Urinary incontinence
- General weakness

Pick's disease generally occurs between the ages of forty and sixty years of age.

Pick's disease affects slightly more women than men.

## Diagnosis of Pick's Disease:

The diagnosis from a doctor can help to rule out other diseases or conditions which can be treatable or cured. A psychologist can help establish the type of dementia.

## Distinguishing Feature to Diagnose:

There are a few distinguishing feature that Pick's disease has which distinguishes it from other disorders: (i) Onset before 65 years of age, (ii) Lack of inhibition, (iii) Initial personality changes (iv) Loss of normal controls for example: hyper sexuality (v) Roaming behaviour.

## Prognosis in Pick's Disease:

The length of progression varies in individuals from less than 2 years to 10 years in some.

## LEWY BODY DISEASE:

Lewy Body Disease is a dementia that is clinically similar to Alzheimer's disease and often characterized by hallucinations, parkinsonian features and extra pyramidal signs. These degenerative brain diseases are linked to abnormal clumps of a protein called alpha-synuclein. These clumps are called as Lewy bodies, which are found in nerve cells throughout the outer layer of the brain (the cerebral cortex) and deep inside the midbrain and brainstem. These patients usually experience progressive cognitive decline, although there is considerable variability between the symptoms. The common symptoms comprise of problems with movement, visual hallucinations, and fluctuations in thinking skills or attention.

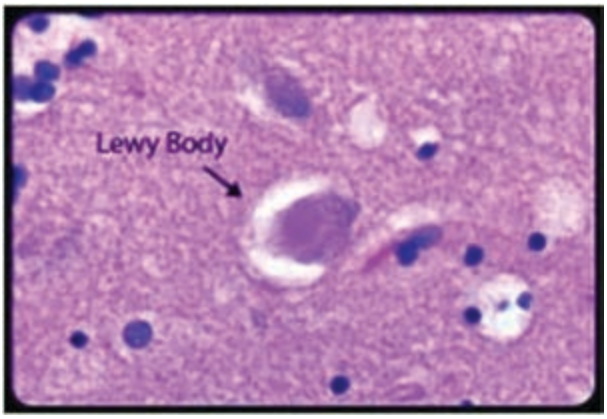



Figure 9: Lewy Bodies found in the midbrain and brain stem

## Causes:

The causes of Lewy body dementia, is unknown in majority of cases. Monogenetic forms of Lewy body disorders, where a patient inherited the disease from one parent, are rare and comprise about 10% of cases. These familial variants are more common in patients with an earlier age of disease onset. The majority of cases are thought to result from complex interactions among different "susceptibility" genes and

environmental risk factors. The primary environmental factors that have been implicated in increasing risk of some cases of Lewy body disorders include pesticide or other chemical exposure and head injury. There are undergoing researches to study the role that genetics and environment play in Lewy body disorders.

The underlying brain changes of Lewy body dementias involve the clumping of a protein in the brain called alpha-synuclein. These clumps are called "Lewy bodies." The regional distribution of these clumps in the brain influences the types of symptoms (e.g., cognitive, motor, psychiatric). Lewy body dementias are also associated with reductions in important brain chemicals called neurotransmitters, including dopamine and acetylcholine.

## Diagnosis:

An extensive neurological and neuropsychological evaluation is essential for Lewy body dementia diagnosis. Structural brain imaging for example MRI Brain and CT scan and laboratory tests are used to rule out other diagnoses. Lewy body dementias can be difficult to diagnose because they can resemble and overlap with other causes of dementia like Alzheimer's disease, other parkinsonian syndromes and vascular dementia.

As in many neurodegenerative diseases, a definitive diagnosis is only available after an autopsy.

## Differential Diagnosis:

Parkinson's disease versus Lewy Bodies: A few patients with Parkinson's disease (PD) experience a subtle cognitive decline and their primary limitation is their motor disorder. Although patients with, Parkinson's disease will develop dementia as a consequence of the disease. When dementia develops after an established motor disorder, we call the disease PDD. In contrast, when the dementia develops prior to or at the same time as the motor disorder, we call the disease DLB. Although the initial sequence of symptoms differs in PDD and DLB, as the disorders progress, the symptoms and the underlying brain changes are much more similar than they are different. As such, many researchers and clinicians think of PDD and DLB as being on a continuum of a similar disease process rather than as two distinct entities.

## Signs and Symptoms:

The parkinsonian motor syndrome is similar in PDD and DLB and can include slowed movement (bradykinesia), rigidity (muscles feel still and resist movement), tremor and gait difficulties. In terms of the cognitive difficulties, both disorders can involve progressive problems involving visual spatial processing, attention, executive dysfunction (e.g., planning and multi-tasking) and memory. Psychiatric symptoms are common and can include anxiety, depression, hallucinations (usually visual) and delusions (false beliefs). Sleep problems including excessive daytime drowsiness and difficulty staying asleep are common. Many patients exhibit REM sleep behavior disorder (RBD) that manifests by the patient acting out his or her dreams.

Many patients experience constipation, repeated falls, syncope and loss of smell. Individuals diagnosed with Lewy body dementias often have adverse

reactions including confusion when taking medications that affect the brain, such as anti- anxiety drugs (examples: Valium®, Ativan®), anticholinergicdrugs(examples: Benadryl, Detrol), and antiparkinson's drugs (examples: Sinemet®, Mirapex). To avoid adverse reactions to medications, physicians should carefully monitor medications, introduce medications one at a time and prescribe minimal doses when possible.

## Disease Progression:

The onset of the disorder usually occurs in geriatric adults, and the disease risk increases with age. There is substantial unpredictability in the course of the illness. In general, symptoms progress over several years as cognitive decline becomes more prominent and psychiatric symptoms emerge or worsen. The progression of symptoms represents increasing pathology in the brain including more widespread Lewy bodies, loss of neurotransmitters (examples: dopamine and acetylcholine) and nerve cell death.

## Prognosis:

The prognosis for Lewy Body Dementia (LBD) can be highly variable. There are no known therapies to stop or slow the progression of. LDB. The average life expectancy following the diagnosis is between five and seven years.

## HUNTINGTON'S DISEASE:

Huntington's disease is classically associated with the development of dementia. The dementia seen in this disease is the subcortical type of dementia, characterized by more motor abnormalities and fewer language abnormalities than in the cortical type of dementia. Grossly, the cortical gyri appear normal to slightly atrophic. Coronal sections reveal striking caudate greater than putamen and pallidum atrophy. Milder degenerative changes occur in the neocortex and thalamus. Evidences exist for excitotoxic damage and mitochondrial DNA mutations and oxidative stress in the pathogenesis of cortical pathology in Huntington's disease.

**Clinical Feature and Associated Disorder:** Lieberman and associates [7] reported that around 90 percent of patients were demented by a mean age of 48.3 years and that dementias preceded the onset of chorea in 24 percent. Whether or not it is the critical determinant of cognitive decline, and cognitive decline correlates to pathologically and radiologically with degree of caudate atrophy and hypometabolism [8]. The area of particular interest in dementia of HD are frontal lobe relate or frontostriatal - related neuropsychiatric deficits, declarative memory impairment, and motor learning difficulties. Regarding cognitive disturbances, prospective study of cognitive decline in early cases has shown that psychomotor slowing is a characteristic early feature and memory impairment although present early on does not deteriorate rapidly, or correlate as well with caudate atrophy, as does psychomotor speed. Huntington's disease causes memory loss but it appears to be less severe as compared with that of Alzheimer's disease at a matched stage of severity, although this factor has been more consistent at milder stages of dementias. Most studies have found that patients with HD have greater difficulty learning a task but benefit more by cued recall or recognition as compared with free or unassisted recall. This has been

thought to reflect greater impairment of retrieval than storage mechanisms and it affects remote memory retrieval as well as more recently learned material. There are many similarities in memory disturbances between HD and PD, suggesting similarities between subcortical dementia illnesses but overlap is imperfect. Evidence shows that the basal ganglia in patients with Huntington 's disease shows a decrease in activity of the mitochondrial pathway, complexII-

**I.** Such deficiencies are often associated with basal ganglia degeneration [9]. This degeneration of striatal neurons projecting to GPe leads to disinhibition of the indirect pathway, increased inhibition of STN, and therefore, reduced output of the basal ganglia [10].

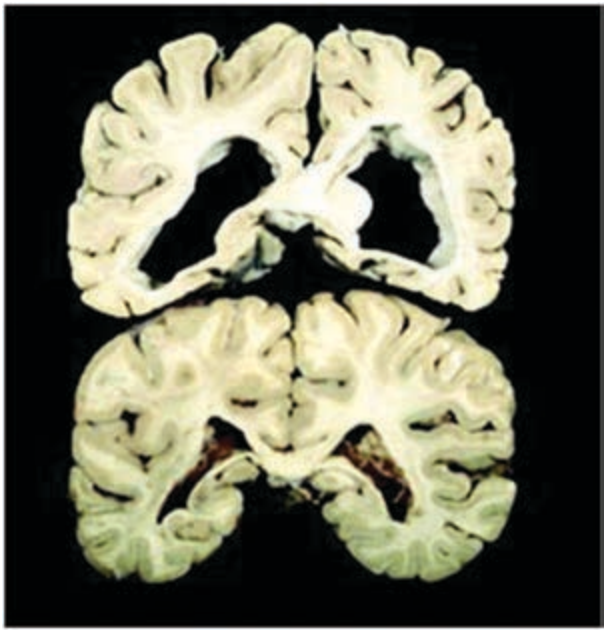


Figure 10: The impact of Huntington Disease on the brain structure in the basal ganglia

## Differential Diagnosis:

In absence of chorea, the family history is critical in reaching a diagnosis. In the absence of both chorea, the family history is critical in reaching a diagnosis. In the absence of chorea and family history, the differential diagnosis is that of other subcortical dementias and chronic progressive encephalopathies.

## Evaluation:

The clinical examination may provide compelling evidence for HD, which can be confirmed genetically. Ancillary tests include MRI and CT to ascertain caudate atrophy and SPECT or PET to ascertain caudate hypoperfusion or metabolism. Neuropsychological assessment is useful to document the nature and severity of cognitive decline, particularly in mild to moderate stages of illness.

## Prognosis and Future Perspectives:

Death occurs within 10 to 20 years after onset, although suicide is more prevalent in at risk and early onset HD patients.

## HIV - RELATED DEMENTIA:

Encephalopathy in HIV infection is associated with dementia and is termed acquired immune deficiency syndrome (AIDS) dementia complex or HIV dementia. The HIV virus will attack the nerve cells in the brain, which will lead to AIDS Dementia. So a person who carries the HIV virus may contract AIDS Dementia. As said earlier even comparatively healthy people with HIV positive status can fall prey to it. At times, the immune system has to grow very weak for the symptoms to surface properly. Patients infected with HIV experience dementia at an annual rate of approximately 14 percent. An estimated 75 percent of patients with AIDS have involvement of the central nervous system (CNS) at the time of autopsy.

The development of dementia in people infected with HIV is often paralleled by the appearance of parenchymal abnormalities in MRI scans. Other infectious dementias are caused by Cryptococcus or Treponema palladium.

Symptoms of HIV Dementia: lack of concentration, withdrawal, forgetfulness, long term and short-term loss of memory, mania, altered personality, irritability, poor coordination, impaired judgment, blurred or hazy vision, reduction in attention span, apathy and loss of bowel and bladder control.

## Diagnosis:

The diagnosis is done on the basis of neurological brain scans, coordination tests and mental activities.

## Head Trauma - Related Dementias:

Dementia can be a sequela of head trauma. The so-called punch- drunk syndrome occurs in boxers after repeated head trauma over many years. It is characterized by emotional liability, dysarthria and impulsivity.

## Dementia Due to Other General Medical Conditions:

The DSM IV TR lists six specific causes of dementia that can be coded directly: HIV disease, head trauma, Parkinson's disease, Huntington's disease, picks disease and Creutzfeldt - Jakob disease. A seventh category allows clinicians to specify other nonpsychiatric medical conditions associated with dementia.

Substance induced persisting dementia: alcohol- related dementia has been estimated to account for, approximately 4% of dementias.

## DIAGNOSIS AND CLINICAL FEATURES:

The dementia diagnosis in DSM-IV-TR dementia of the Alzheimer's type, vascular dementia, dementia due to other general medical condition, substance induced persisting dementia, dementia due to multiple etiologies, and dementia due to not otherwise specified. The most important step in making the diagnosis is to determine whether a chronic encephalopathy results from a degenerative or due to other potentially reversible causes. A reversible process includes fluctuating severity, altered level of consciousness or hypersomnolence and visual hallucinations.

The diagnosis of dementia is based on the clinical examination, including a mental status examination, and information from the patient's family, friends, and employers. Complaints of a personality change in patients older than age 40 suggest that the diagnosis of dementia should be carefully considered.

Clinicians should note patients complain is about intellectual impairment and forgetfulness as well as evidence of patients evasion, denial, or rationalization aimed at concealing cognitive deficits. Excessive orderliness, social withdrawal, or a tendency to relate event in minute detail can be characteristic, and sudden outburst of anger sarcasm can occur. Patients in appearance and behaviour should be observed. Liability of emotions, sloppy gloomy, uninhabited remarks, silly jokes, or a dull, apathetic, or vacuous facial expression and manners suggest the presence of dementia, especially when coupled with memory impairment.

Memory impairment is typically an early

and prominent feature in dementia, especially in dementia involving the contacts, such as dementia of the Alzheimer's type. Early in the course of dementia, memory impairment is mild and usually most marked for recently went; people Forget telephone numbers, conversation, and events of the day. As the course of dementia progress is, memory impairment becomes severe, and only the earliest learned information example of a person's place of birth is retained. In as much as memory is important for orientation two per cent, place, and time, orientation can be progressively effected during the course of a dementing illness. For example patients with dementia may forget how to get back to their room after going to the bathroom. No matter how severe the disorientation seems, however, patients show no impairment in their level of consciousness.

Dementing processes that affect the cortex, primarily dementia off the Alzheimer style and vascular dementia, can affect patients language abilities. DSM-IV-TR includes aphasia as one of the diagnostic criteria. The language difficulties may be characterized by awake, stereo type, imprecise, circumstantial locution and patient may also have difficulty naming objects.

## PSYCHIATRIC AND NEUROLOGICAL CHANGES:

### Personality:

Changes in the personality of a person with dementia are especially disturbing for their families. Preexisting personality traits may be accentuated during the development of a dementia. Patients with dementia may also become introverted and seem to be less concerned than they previously were about the effects of their behaviour on others. Persons with dementia who have paranoid delusions are generally hostile to family members and caretakers. Patients with frontal and temporal involvement are likely to have marked personality changes and may be irritable and explosive.

### Hallucinations and Delusions:

An estimated 20 to 30% of patients with dementia have had hallucination and 30 to 40% have delusions, primarily of paranoid or persecutory and unsystematized nature, although complex sustained and well systematized illusions are also reported by these patients. Physically aggression and other forms of violence are common in demented patients who also have psychotic symptoms.

### Mood:

In addition, to psychosis and personality changes, depression and anxiety major symptoms in an estimated 40 to 50% of patients with dementia, although the full syndrome of depressive disorder may be present in only 10 to 20 per cent. Patients with dementia also may exhibit pathological laughter or crying that it is, extremes of emotions with no apparent provocation.

### Cognitive changes:

In addition to appraise in patients with dementia, apraxia's and agnosia's are common, and they are included as potential diagnostic criteria in DSM IV TR. Other neurological signs that can be associated with dementia seizures, seen in approximately 10% of patients with dementia of the Alzheimer style and in 20%

of patients with vascular dementia and a typical neurological presentation, such as nondominant parietal lobe syndrome. Primitive reflexes, such as the grasp, snout, suck, tonic foot and palmomental reflexes may be present on neurological examination and myoclonic jerks are present in five to 10 percent of the patients.

Patients with vascular dementia may have additional neurological symptoms, such as headaches, dizziness, faintness, weakness, focal neurological signs, and sleep disturbances, possibly attributable to the location of the terrible or vascular disease.

## Behavioral symptoms of dementia:

Patients with dementia may experience behavioral and psychological symptoms and during the course of their illness. These may include:

Behavioral Symptoms: screaming, restlessness, physical aggression, agitation, shaking, screaming, wandering, culturally inappropriate behaviors, sexual disinhibition, hoarding, cursing, and shadowing.

Psychological Symptoms: depression, delusions, hallucinations, delirium and apathy.

## Causes of Behavioral Symptoms:

***Biological:***

- Genetic abnormalities related to the structure of particular parts of the brain.
- Changes of chemicals in the brain.
- Changes in the structure of different parts of the brain due to dementia.

**Medical:**

- Patients with dementia who also suffer from infections, pain, dental problems and arthritis may be unable to articulate the pain they are experiencing and instead may express their distress by vocalizing or becoming aggressive.
- Co-morbid conditions such as delirium, depression, anxiety or psychosis. Individuals with dementia are more susceptible to delirium and the illnesses / environmental stressors that lead to delirium.
  - Side effects caused due to medication can be numerous and may have a significant effect on a person's behavior
  - Hearing or vision impairment that is not well managed (for example, malfunctioning hearing aids may lead to frustration and changed behaviors).
  - Sleep disturbances are common in people with dementia and can cause agitation and restlessness during the day and night.

Environmental and social:

- Changes in social routine (for example, alteration in meal times or introduction of a new care routine can cause confusion and a feeling of loss of control for the person with dementia, contributing to behaviors of concern).
- Change in environment (for example, relocation to a new room or home can increase agitation and disorientation).

## Psychosocial Determinants:

The severity and course of dementia can be affected by psychosocial factors. The

greater a person's premorbid intelligence and education, the better the ability to compensate for intellectual deficits. People who have a rapid onset of dementia use fewer defenses than do those who experience an insidious onset. Anxiety and depression can intensify and aggravate the symptoms. Pseudodementia occurs in depressed people who complain of impaired memory, but in fact are suffering from a depressive disorder. When the depression is treated, the cognitive defects disappear.

## Catastrophic reaction:

Patient with dementia also exhibit a reduced ability to apply what Kurt Goldstein called the abstract attitude. Patients have difficulty generalizing from a single instance, forming concepts and grasping similarities and differences are among concepts. Furthermore, the ability to solve problems, to reason logically and to make sound judgement is compromised. Goldstein also described a catastrophic reaction marked by agitation secondary to the subjective awareness of the intellectual depth received under stressful circumstances. Persons usually attempt to compensate for defects by using strategies to avoid demonstrating failures in intellectual performance; they may change the subject, make jokes or otherwise divert the interviewer.

## Sundrowner Syndrome:

Sundrowner Syndrome is characterized by drowsiness, conclusion, ataxia, and accidental faults. It occurs in older patients who are overly sedated and inpatients with dementia that reacts adversely to even a small dose of psychoactive drug. The syndrome also occurs in demented patients when external stimuli, such as light and interpersonal orientation is cues are diminished.

## GENERAL PATHOLOGY, PHYSICAL FINDINGS AND LABORATORY EXAMINATION:

A comprehensive laboratory workup must be performed when evaluating a patient with dementia. The purposes of the workup are to detect reversible causes of dementia and to provide the patient and family with a definitive diagnosis. The range of possible causes of dementia mandate selected use of laboratory test. The evaluation should follow clinical suspicion, based on the history and physical and mental status examination results. The continued improvements in brain imaging techniques, particularly MRI, have made differentiation between the dementia of the Alzheimer type, and vascular dementia, in some cases somewhat more straightforward then in the past. And active area of research is the use of single photon emission computed tomography (SPECT) to detect patterns of brain metabolism in various types of dementia; the use or off (SPECT) images may soon help in the clinical differential diagnosis of dementing illnesses.

A reasonably recent complete physical evaluation should be obtained in patients who are suspected of suffering from dementia. It may reveal evidence of systematic disease-causing brain dysfunction, such as an enlarged liver, and hepatic encephalopathy, or it may demonstrate systematic disease related to particular CNS processes. Patient should also undergo structural neuroimaging with computed tomography (CT) or magnetic resonance imaging (MRI) of the brain.

When a nondegenerative based chronic progressive encephalopathy is suspected, electroence- phalography (EEG) may be helpful in demonstrating severe dysrhythmic slowing. Cerebrospinal fluid (CSF) examination may show evidence of a chronic meningoencephalitis process such as an elevated IgG index and synthesis rate and occasionally oligoclonal bands, suggesting an intrathecal inflammatory reaction. Ultimately is suspicion remains high based on noninvasive tests for nondegenerative causes, cerebral angiography and meningeal and brain biopsy can be considered, or in the absence of such invasive testing, an empirical therapeutic trial of prednisone could be considered for a possible steroid-responsive inflammatory meningoencephalitis. Focal neurological findings, such asymmetrical hyperreflexia or weaknesses are seen more often in vascular then in degenerative diseases.

## DIFFERENTIAL DIAGNOSIS:

**Dementia of the Alzheimer type versus Vascular Dementia:**

Classically, vascular dementia has been distinguished from dementia of the Alzheimer type by the decremental deterioration that can accompany cerebrovascular disease overtime. Although the discrete, stepwise deterioration may not be apparent in all cases, focal neurological symptoms are more common in vascular dementia of the Alzheimer's type, as are the standard risk factors for cerebrovascular disease.

**Vascular Dementia versus Transient Ischemic Attacks:**

Transient Ischemic Attacks (TIA's) are brief episodes of focal neurological dysfunction lasting less than 24 hours. Although a variety of mechanisms may be responsible, the episodes are frequently the result of microembolization from a proximal intracranial arterial lesion that produces transient brain ischemia, and the episodes usually resolve without significant pathological alteration of the parenchymal tissues. Approximately one third of the person's with untreated TIA's experience a brain infarction later; therefore, recognition of TIA's is an important clinical strategy to prevent brain infarction.

Clinicians should distinguishing episodes involving the vertebrobasilar system from those involving the carotid arterial system. In general, symptoms of vertebrobasilar disease reflect a transient functional disturbance in either the brainstem or the occipital lobe; carotid distribution symptoms reflect unilateral retinal or hemispheric abnormality. Anticoagulant therapy, antiplatelet agglutinating drugs such as aspirin, and extracranial and intra cranial reconstructive vascular surgery are effective in reducing the risk of infarction in patients with TIA's.

## Delirium:

In general, delirium is distinguished by rapid onset, brief duration, cognitive impairment fluctuation during the course of the day, nocturnal exacerbation of symptoms, marked disturbance of the sleep - wake cycle, and prominent disturbance in attention and perception.

## Depression:

Some patients with depression have symptoms of the cognitive impairment difficult to distinguish from symptoms of

dementia. The clinical picture is sometimes referred to as pseudodementia, although the term depression- related cognitive dysfunction is preferable and more descriptive. Patients with depression - related cognitive dysfunction generally have prominent depressive symptoms, more insight into their symptoms than do demented patients, and often a history of depressive episodes.

### Factious Disorder:

Person who attempts to stimulate memory loss, as in factitious disorder, do so in an erratic and inconsistent manner. In true dementia, memory for time and place is lost before memory for persons, and recent memory is lost before remote memory.

### Schizophrenia:

Although schizophrenia can be associated with some acquired intellectual impairment, its symptoms are much less severe than are the related symptoms of psychosis and thought disorder seen in dementia.

### Normal Aging:

Aging is not necessarily associated with any significant cognitive decline, but minor memory problems can occur as a normal part of aging. These normal occurrences are sometimes, referred to as benign senescent forgetfulness or age - associated memory impairment. They are distinguished from dementia by their minor severity and because they do not interfere significantly with a person's social or occupational behaviour.

### Other Disorders:

Mental retardation, which does not include memory impairment, occurs in childhood. Amnestic disorder is characterized by circumscribes loss of memory and no deterioration. Major depression in which memory is impaired responds to antidepressant medication. Malingering and pituitary disorder must be ruled out, but they are unlikely.

## COURSE AND PROGNOSIS:

The classic course of dementia is an onset in the patients 50s or 60s with gradual deterioration over 5 to 10 years, leading eventually to death. The age of onset and the rapidly of deterioration vary among different types of dementia and within individual diagnostic categories. The average survival expectation for patients with dementia of the Alzheimer's type is approximately 8 years, with a range of 1 to 20 years. Data suggest that in persons with an early onset of dementia or with a family history of dementia the disease is likely to have a Rapid course.

The most common course of dementia begins with a number of subtle signs that may, at first, be ignored by both the patient and the people closest to the patient. A gradual onset of symptoms is most commonly associated with dementia of the Alzheimer's type, vascular dementia, endrinopathies, brain tumors and metabolic disorders. Conversely, the onset of dementia resulting from head trauma, cardiac arrest with cerebral hypoxia, or encephalitis can be sudden. Although the symptoms of the early phase of dementia are subtle, they become conspicuous the as the dementia progresses, and family members may then bring a patient to a physician's attention. People with dementia may be sensitive to use of benzodiazepines or alcohol, which

can be precipitate agitated, aggressive or psychotic behaviour. In the terminal stage of dementia, patients become empty shells of their former selves- profoundly disoriented, incoherent, amnestic and incontinent of urine and feces.

With psychosocial and pharmacological treatment possibly because of the self-healing properties of the brain, the symptoms of dementia may progress slowly for a time or may even recede somewhat. Symptoms regression is certainly a possibility in reversible dementias once treatment is initiated. The course of the dementia varies from steady progression to an incrementally worsening dementia to a stable dementia

## Stages of Dementia:

***1- Early stage (developed in 1-2 years):***

The early stage of dementia is often overlooked because the onset of dementia is gradual; it is often difficult to be sure exactly when it begins. The person may for example:

- Individual may have problems talking properly (language problems).
- Individual may have significant memory loss
- particularly for things that have just happened.
- The individual may not be oriented.

**Middle stage (developed in second to fifth year):**

As the disease deteriorates gradually, limitations become clearer and more restricting. The person with dementia has difficulty with day-to-day living and:

- May become very forgetful - especially of recent events and people's names.
- Can no longer manage to live alone without problems.
- Is unable to cook, clean or shop.

***2-*** **Late stage (developed in fifth year or after):**

This stage is one of near total dependence and inactivity. Memory disturbances are serious and the physical side of the disease becomes more obvious. The person may:

- Have difficulty eating.
- Be incapable of communicating.
    - Not recognize relatives, friends and familiar objects.
    - Display inappropriate behavior in public.
- Be confined to a wheel chair or bed.

Morbidity and Mortality associated with Dementia Global Morbidity and Mortality:

- In 2000, age-standardized dementia mortality rate was 6.7 and 7.7 for 100,000 male and female respectively. [10]
- 24.3 million have dementia [11]
- 4.6 million new cases per year [11]
- Worldwide dementia contributes 4.1% of all disability-adjusted life years (DALYs) and 11.3% of years lived with disability and 0.9% of years of life lost.

**India Morbidity and Mortality:**

- Age standardized death rate of 12.1 per 100,000 reference number?
- 1.8 million have dementia in India and South Asia in people >60 years of age. [11]

- 400,000 new cases per year for India + South Asia.[11]
- 1,034 per 100,000 DALYs.

## TREATMENT FOR DEMENTIA:

**PHARMACOTHERAPY:**

Clinicians may prescribe benzodiazepines for insomnia and anxiety, antidepressant for depression and antipsychotic drugs for delusions and hallucinations, but they should be aware of possible distinctive drug effects in older people. In general, drugs with high anticholinergic activity should be avoided.

Donepezil, rivastigmine, galantamine and tacrine and cholinesterase inhibitors used to treat mild to moderate cognitive impairment in Alzheimer's disease. They reduce the inactivation of the neurotransmitter acetylcholine and thus potentiate the cholinergic neurotransmitter, which in turn produces a modest improvement in memory and goal-directed thought. These drugs are most useful for persons with mild to moderate memory loss who have sufficient preservation of their basal forebrain cholinergic neurons to benefit from augmentation of cholinergic neurotransmission.

Donepezil: AriceptTM is the trade name for donepezil. It is a cholinesterase inhibitor, making it a drug from the main class of compounds now used to treat people with Alzheimer's disease. AriceptTM has been used in a large number of clinical trials, including some from Canada that have used a variety of individualized symptom-based tests of its effectiveness. In 2006, the US Food and Drug Administration approved AriceptTM as a treatment for severe dementia associated with Alzheimer's disease.

Dosage: This is taken once a day, with food. The starting dose is 5 mg / day. After a month on the drug, if it is well tolerated, most patients will be increased to the 10 mg / day dose. Usually, at least two months more are needed to know whether the drug is meeting the goals of treatment.

Rivastigmine: ExelonTM and Exelon TMPatch are the trade names for rivastigmine. It is a cholinesterase inhibitor for the treatment of Alzheimer's disease. It is also used to treat people with Lewy body dementia, and with the dementia of Parkinson's disease.

Dosage: The starting dose is 1.5 mg, twice a day. Typically, after one month, this is increased to 3.0 mg, twice a day, which is often the lowest effective dose. Patients who tolerate that dose can have it increased to a dose of 4.5 mg twice a day and later to a dose of 6.0 mg twice a day. These are the recommended doses.

Exelon TMPatch is now available in some countries, including the United States and Canada. It is applied to the back, chest or upper arm and provides continuous delivery of medication through the skin over 24 hours. Treatment is initiated with Exelon TMPatch 5 (9mg / 5cm2) with a release rate of 4.6mg over a 24 hour period to a maximum dosage of Exelon TMPatch 10 ( 18mg / 10cm2) with a release rate of 9.5mg over a 24 hour period.

Galantamine: Reminyl ER is the trade name for galantamine. It is a cholinesterase inhibitor, making it a drug from the main class of compounds now used to treat people with Alzheimer's disease..

In addition to acting like the other cholinesterase inhibitors, it also directly stimulates a class of receptors to which acetylcholine binds.

Dosage: Briefly, galantamine (extended release) is taken once a day, with food. The starting dose is 8 mg / day. After a month on the drug, if it is well tolerated, most patients will be increased to the 16 mg / day dose, and there is also a 24 mg / day dose. Usually, at least two months after starting at the 16 mg / day dose are needed to know whether the drug is meeting the goals of treatment.

Memantine: Memantine goes by several trade names, or brand names, including EbixaTM, NamendaTM (in the United States) and AxuraTM. It is an NMDA, or N-Methyl-D-aspartic acid. (N- methyl-D-aspartate) receptor antagonist, making it distinct from cholinesterase inhibitors, the other class of compounds commonly used to treat people with Alzheimer's disease. The reason to block the NMDA receptor is to block transmission of a brain chemical called glutamate. Glutamate is a neurotransmitter, which is used widely throughout the brain, but too much glutamate is felt to cause calcium overload in brain cells. The theory is called Glutamatergic Excitotoxicity.

Dosage: Briefly, memantine is taken twice a day, with food. The starting dose is 5 mg / day. After a week, this is increased to 5 mg, twice a day. If it is well tolerated, most patients will be increased to the 10 mg / once a day and 5 mg once a day, after which the dose increases to 10 mg twice a day. Usually, after starting at the 10 mg twice a day dose, at least two months are needed to know whether the drug is meeting the goals of treatment.

Donepezil is well tolerated and widely used. Tacrine is rarely used, because of its potential for hepatotoxicity. Fewer clinical data are available for rivastigmine and galantamine, which appears more likely to cause gastrointestinal and neuropsychiatric adverse effects than donepezil. None of these medications prevents the progressive neuronal degeneration of the disorder.

Other Treatment Approaches: Other drugs being tested for cognitive-enhancing activity include general cerebral metabolic enhancers, calcium channel inhibitors, and serotonergic agents. Some studies have shown that selegiline, a selective type B monoamine oxidase inhibitor, may slow the advance of this disease. Ondansetron, a 5 HT 3 receptor antagonist, is under investigation.

Estrogen replacement therapy: Estrogen replacement therapy may reduce the risk of cognitive decline in postmenopausal women; however, more studies are needed to confirm this effect. Complementary and alternative medicine studies of ginkgo biloba and other phytomedicinals to see if they have a positive effect on cognition. Reports have appeared of patients using nonsteroidal anti-inflammatory agents having a lower risk of developing Alzheimer's disease. Vitamin E has not been shown to be of value in preventing the disease.

## Medical Management of Dementia

**Conservative management:**

- Treat the associated conditions that can also affect the memory. For eg.Treat hypertension or depression or sleep disturbances if present.

- Start neuroprotective vitamins. For eg. Vit B12, folic acid, CoQ10, omega3fatty acids

- Start cognitive rehabilitation. Refer to a psychologist for specialized rehabilitation. Studies have shown computer use, playing games, reading books and other intellectual activities may help preserve function and prevent cognitive decline.

- Regular physical exercise may also help prevent or slow cognitive decline.

- Dietary advice:

A diet low in fat and rich in fruits and vegetables

- Social engagement may help to preserve mental function and slow mental Decline.

**Pharmacology:**

1. Cholinesterase inhibitors

Tab.Donepezil 5 to 10 mg at bed time.in 4 to 6 weeks for mild to moderate disease.

Tab.Galantamine ER 8 mg -16mg OD X2WEEKS

Tab.Rivastigmine 1.5 -3mg OD X 2 WEEKS

2. NMDA receptor antagonist

Tab.Memantine 5mg-20mg OD\

Tab.Memantine 7mg OD dose increase weekly upto 28mg

Management of the associated symptoms & non pharmacological treat-ment given in the end of this chapter(pg number )

3. Vascular Dementia

This is also called as multi-infarct dementia,and this occurs because theblood supply to the brain is affected typically due to a series of minorstrokes.Here the features of cognitive decline is associated with atleast onecerebral infarct which is temporary innature.

Investigations:

Radiological findings may show one or more cerebral infarcts of varying sizes,orsevere diffuse white matter abnormalities

- MRI may show global atrophy, diffuse white matter lesions, lacunes and'strategic infarcts' (infarcts in regions that are involved in cognitive func-tion)

- PET CT: Hypometabolism in the areas of infarct

- CT: Multiple lacunar infarcts

Management of vascular dementia

a. Control blood pressure with antihypertensives like tablet amlodepin 5mgod or tablet telmisartan 20 mg od or Tb Lisinopril 5 mg od

b. Control diabetes with antidiabetic medicines like tablet metformin,glyboride

c. Control cholesterol levels by medications like tablet atorvastatin 40 mg atnight, tablet rosuvastatin 20mg at night.

d Anti cholinester ase medicine slike Donepezilisal sousedas cognitive enhancer.

Management of the associated symptoms & non-pharmacological treat-ment given in the end of this chapter.

4. Dementia with Lewy Bodies (DLB)

This is a type of dementia closely associated with Parkinson's disease, this is characterized by presence of lewy bodies which are clumps of alpha-synuclein & ubiquitin proteins in neurons.

Features like Impaired attention, concentration are seen, whereas

visuospatial functioning, memory and naming is preserved.

Parkinsonism, uctuations and visual hallucinations are highly suggestiveof DLB.

Behavioural disturbances worsen with typical antipsychotics. Thus drugslike haloperiodol should not be given in this dementia.

Investigations:

MRI may often be normal or may have hippocampus and cortical

atrophy if coexisting Alzheimer's disease

Management of DLB

Medications such as Tab.Donepezil /Tab.R ivastigmine or Tab.carbidopa or levodopa

Management of the associated symptoms & non pharmacological treatmentgiven in the end of this chapter.

5. Frontotemporal Dementia

This is a type of dementia where the frontotemporal area of brain is

affected.

It may be associated with motor neuron disease.

Prominent features are personality/ behavioural change,apathy or language dysfunction.

Investigations:

MRI -Frontal or/and temporal cortical atrophy

PET CT - Hypometabolism in frontal and temporal lobes

Treatment of FTD

Medications such as tablet donepezil or tablet rivastigmine or Tab.carbidopa or levodopa

Management of the associated symptoms & non pharmacological treatment given

Normal pressure hydrocephalus or NPH ia also and important differential diagnosis which is described in Section A.

Management of Associated Symptoms With Dementia

Sun-downing, Aggressive Behavior

Antipsychotic drugs such as haloperidol or respiridone 0.5-1.0mg at bedtime to be given

Depression:Amitriptyline 25 mg bd, or Nortryptyline 20-50 mg twice a day or serotonin

reuptake inhibitors like sertraline to be given.

Sleep Disturbance

Tablet zolpidem 5 mg qhs, or Tb alprazolam 0.2 mg, or Tb Temazepam 15 mg 0-0-1

Neuroprotective Drugs (Supplements)

As Supplements are very helpful along with the medicines

Tablet CO Q or Tablet Ubiquinol also tablet vitamin c,vitamin e,multivitamin especially B complex including B12 & folic acid.

Other alternating medicines like gingko biloba are also being used.

**PSYCHOLOGICAL INTERVENTION:**

Psychological Intervention is very often required in geriatrics but for individuals who are suffering from serious disorders it is very important that they undergo psychological treatment. Psychological treatment very often involves working in an orchestrated way with several professionals.

Intervention Plan:

- Early testing and diagnosis
- Optimization of physical health, cognition, activity and overall well being.
- Detection and treatment of Behavioural and Psychological Symptoms of Dementia.
- Educating carer and providing long term support to carer.

## PSYCHOLOGICAL ASSESSMENT FOR DEMENTIA:

1. ADAS- Cog: This test is used to measure cognition which consists of 11 tasks measuring the disturbances of memory, language, praxis, attention and other cognitive abilities which are often referred to as the core symptoms of Alzheimer's disease. This is recommended for the second stage on Alzheimer's disease. The estimated evaluation time of this test is 30 to 45 minutes. This test is usually administered by a neuropsychological or a clinical psychologist

2. Quality of Life - Alzheimer's disease Scale (QOL - AD): This test is used to measure the quality of life. This scale consists of 13 domains to measure quality of life.

3. DEMQOL: This test assesses the quality of life in persons with mild to moderate dementia. It takes approximately 10 to 20 minutes for assessment.

4. Modified Mini - Mental Status Examination (3MS): The Modified Mini - Mental Examination is a brief widely used test to measure the cognitive functions with good reliability and validity. The administration time taken is approximately 15 minutes. Qualified health-care professional (at least trained in the Mini Mental) interviews the patient using a standard set of questions. Scoring takes 5 min.

5. Holden Communication Scale: This is a scale which is used to assess the communication changes in the patient. This scale is completed by staff or family caregivers and covers a range of social behaviour and communication variables.

6. Cornell Scale for Depression in Dementia: This scale is used to assess

depression. This scale consists of five broad categories using the information from interviewers, with staff and participants

7. Rating Anxiety in Dementia Scale (RAID): This scale assesses anxiety in the individual suffering from dementia. This scale rates anxiety in four main categories and uses interviews with staff and participants.

8. Neuropsychiatric Inventory (NPI): This test is used to assess behavioral disturbances in patients with dementia. To assess psychopathology in the person with dementia and to help distinguish between the different causes of dementia. Acute, Primary, Community and Residential Care. Includes symptoms known to be rare in Alzheimer's disease, but are characteristic of fronto- temporal dementias. It also assesses the level of caregiver distress engendered by each of the neuropsychiatric disorders. This test assesses 10 behavioral disturbances occurring in dementia patients. It takes around 10 to 20 minutes to administer.

9. Short Form-12 Health Survey (SF-12): This scale is used to measure generic health concepts relevant across age, disease, and treatment groups. The SF-12 includes 8 concepts commonly represented in health surveys. It is a self-administrative measure that provides a comprehensive, psychometrically sound, and efficient way to measure health from the patient's point of view by scoring standardized responses to standard questions. This measure is only used when a family caregiver is available from the community sample participants.

10. Frontal Executive Function: The FAB is a brief tool that can be used at the bedside or in a clinic setting to assist in discriminating between dementias with a frontal dysexecutive phenotype and Dementia of Alzheimer's Type (DAT). The FAB has validity in distinguishing Fronto-temporal type dementia from DAT in mildly demented patients (MMSE > 24). Total score is from a maximum of 18, higher scores indicating better performance. It takes approximately 1o minutes to administer and 5 minutes to scor

11. Behavioural Pathology in Alzheimer's Disease (BEHAVE-AD): The purpose of this test is to measure behavioural and psychological symptoms of dementia in persons with Alzheimer's disease. It takes approximately 20 minutes.

12. Clinical Dementia Rating (CDR): The purpose of this test is to assess the severity of cognitive and functional impairment in dementia. The severity of dementia is rated on the following severity scale:

    0 - No Impairment

    0.5 - Questionable Impairment

    1 - Mild Impairment

    2 - Moderate Impairment

    3 - Severe Impairment

    This is a very comprehensive test and takes approximately 40 to 75 minutes.

13. Clock Drawing Test:

Instructions for the Step 1: Give patient a sheet of paper with a large (relative to the

size of handwritten numbers) pre drawn circle on it. Indicate the top of the page.

Step 2: Instruct patient to draw numbers in the circle to make the circle look like the face of a clock and then draw the hands of the clock to read "10 after 11."

Scoring:

Score the clock based on the following six-point scoring system:

1 Perfect: No errors in the task

2 Minor visuospatial error

3 Inaccurate representation of 10 after 11 when visuospatial organization is perfect or shows only minor deviations.

4 Moderate visuospatial disorganization of times such that accurate denotation of 10 after 11 is impossible.

5 Severe level of disorganization as described in scoring of 4

6 No reasonable representation of a clock

The first step in intervention of dementia is verification of the diagnosis. Accurate diagnosis is imperative, for the progression may be halted or even reversed if appropriate therapy is provided. Preventive measures are important, particularly in vascular dementia. Such measure might include changes in diet, exercise and control of diabetes and hypertension. Pharmacological agents might include antihypertensive, anticoagulant or antiplatelet agents. Blood pressure control aim for the higher end of the normal range, because that has been demonstrated to improve cognitive function in patients with vascular dementia. Blood pressure below the normal range has been demonstrated to further impair cognitive functions in the patient with dementia. Surgical removal of carotid plaques may prevent subsequent vascular events in carefully selected patients. The general treatment approach to patients with dementia is to provide supportive medical care, emotional support for the patients and their families, and pharmacological treatment for specific symptoms, including disruptive behavior.

**Psychological Interventions:**

Dementia being an impairment in cognitive skills and progressive deterioration of daily function, often associated with behavioral disturbances. Psychological intervention assists in coping with self and surrounding along with family counseling's that improve acceptance and reduces stress and other hindrance that could be avoided by planning, organizing and accepting. The following are the strategies that are used to intervene clients with dementia.

**Cognitive stimulation:**

This includes involving the client in the pleasurable activities such as games, puzzles, dance, cooking, music, and gardening, Discussion of pleasurable past events, which are done by trained professional who assist in reality orientation and reduce confusion. This has proven to reduce hospitalizations., Improve working memory, Reduce behavioral disturbance.

**Reminiscence Therapy**

It's therapy involves discussion of the past experiences, events, family members, friends, activities using materials like photographs, tapes, videos, books, newspapers that are familiar to the client. This facilitates sharing with people and also valuing the experiences.

The therapy reduces agitated behavior

**Validation therapy:**

The principle of acceptance of the truth and reality of another person's experience. This assist in resolving the conflicts by using verbal and nonverbal communication and speaking out clearly and by encouraging.

Validation therapy promotes the communication deterioration with family and friend.

**Behavior Management**

The Intervention constitute of the behavior Management that comprise of cognitive behavioral therapy, analysis of specific behavior, Reinforcement strategies, Communication and habit training, progressive muscle relaxation and token economies. The intervention can be implemented on the client or by training the caregivers to implement on the client. Behavior management have shown to improve the behavioral symptoms of dementia. (13,14)

Treatment for Behavioral Issues in Dementia: Treatment strategies for behavioral problems in dementia include:

- Nonpharmacological Approach (e.g., behavior therapy, cognitive stimulation therapy, psychoeducation)
- Pharmacological Approach (e.g., antipsychotics, antidepressants, mood stabilizers, cognitive enhancers)

**Assisted Living**

Clients with dementia do not require nursing when they are at home, rather they require assistance in the ADL activities such as bathing, dressing and eating. Family members could act as the intermittent care givers by providing different types of support for the ADL activities. The behavior management training underwent to improve caregiver management with clients with dementia at home has shown to reduce behavior problems and depression in the clients.

**Residential Care**

This residential care involves assisted living residences and nursing homes. It included facilities for people with dementia who needed significant nursing care. There is significant reduction in behavioral disturbances with clients in residential care.

**Family counselling:**

It is important to care of the family taking responsibility of the client having dementia. The coping strategies, understanding the pathology of the condition by the family and assist their journey towards acceptance and playing a role in the assistance and care of the client is necessary for the limited stress and squeal of depression. Thus, frequent counseling's session by a counsellor promotes healthy lifestyle and better coping as a family member of the client with dementia.

- Communication Strategies for Psychologist: Speaking to Patients with Dementia
- Use short, simple, familiar and precise words, sentences and commands.
- Take on a similar posture to theirs to develop rapport non-verbally be on their level.

- Go to their eye level and be sure that they can see you clearly when you talk and listen.
- Other forms of communicating include music, touch, food and joint activity.
- Treat person as an adult and don't be condescending.
- It is important to remember that the person who is suffering from dementia and has associated problems had at some point of time a life rich with history, experience, relationships, skills, hopes and dreams. In your role, you may have a wealth of information about this person as he / she declines, and it is important to share this information to keep continuity of care and interests alive for the person. Be sure to inquire into the activities that have had meaning to see if they are still being initiated.
- Behavioural Strategies for Dementia Caregivers:
- Families and carers may find the following strategies helpful when responding to the behavioural and psychological symptoms of dementia:
- Every individual has a specific behaviour pattern, so do people suffering from dementia have. One needs to look for behaviour pattern and try to identify any triggers, as certain behaviours are displayed on at certain times or during particular activities. For example: at a very noisy place, or very bright light which may add to the confusion and restlessness.
- Establishing a routine is very helpful as it is helps patients with dementia be oriented, have sequential memory feeling secure. The person with dementia may become upset if they find themselves in a strange situation or among unfamiliar people, and may become confused, anxious, or agitated
- Clear and simple communication is very important i.e. giving specific commands. The person with dementia may become agitated if they do not understand what is expected of them. They may also feel frustrated with their
- inability to make them self understood. It is important to face the person, speak slowly in a calm and reassuring voice and use simple sentences. Be patient and allow extra time. Ensure the person with dementia participates as much as possible in daily activities. Make sure the task isn't too complicated or the person with dementia may become overwhelmed.
- Regular exercise is very important. Simple exercise, like a walk outside or a game, can reduce anxiety and depression and can minimise other behaviours such as wandering and restlessness
- Distraction often helps. If possible, direct the person with dementia away from a particular task or environment that seems to be triggering the behaviour. Suggest a different activity that the person with dementia may like, such as listening to a favourite song, or going outside.
- Avoid punishment. The person with dementia may not remember the event, and therefore may not be able to learn from it. Try to stay positive.
- Be consistent. If there are strategies for modifying behaviours that work, try to

ensure other people who also care for the person with dementia use the same strategies.

- Communication is important. Always explain to the person with dementia what is going to happen. Speak slowly in a calm and reassuring voice, and use simple sentences. Be patient.
- Involve the person with dementia as much as possible. Even if the person with dementia cannot participate verbally, they can still enjoy certain activities, such as reminiscing about their past by looking at old photos, or listening to a favourite song.
- Be sensitive to the reactions of the person with dementia. Therapies that involve remembering about the past, such as reminiscence or music therapy, may prompt happy memories, but may also prompt painful or sad memories.
- Avoid creating discomfort for the person with dementia. Sometimes the person with dementia may not feel comfortable being touched, so therapies such as massage may not be appropriate for these people.

## Occupational Therapy in Dementia

Alzheimer's Disease is one of the types of progressive and irreversible type of Dementia.

**Functional Stages of Cognitive Decline:**

| | |
|---|---|
| Normal | No objective evidence of impairment. Activities of Daily Living (ADLs) |
| Forgetfulness | No Objective evidence impairment, but the person may express concern about forgetting the location of familiar on objects or forgetting name and words. No Problems with ADL |
| Borderline Alzheimer's Disease | Objective evidence is hard to obtain, but the family and associates confirm problems, such as the person getting lost, job performance declining, person forgetting what has just been said and having difficulty finding words in conversation. Individual expresses anxiety about memory problems. |
| Mild Alzheimer's Disease | Objective evidence of cognitive deficit is apparent, such as decreased ability to handle finances or shop for supplies, decreased knowledge of current events, and deficit in memory of personal history and ability to concentrate on a task. No consistent problems with ADLs. |
| Moderate Alzheimer's Disease | Objective evidence of dementia is apparent, such as inability to recall address and telephone number, names of family members or date of graduation from high school. ADLs are beginning to get affected, such as difficulty choosing -roper clothing or remembering to bathe, eating and toileting skills be performed independently |
| Moderately-severe Alzheimer's Disease | Objective evidence of dementia is readily apparent such as difficulty in remembering the name of the primary care giver although the person can distinguish between strangers and friends. The person is generally unaware of current events, is not oriented to time and place and cannot travel alone. The person is semi-dependent in most ADLs, has difficulty putting on cloths, needs assistance in bathing and toileting and may be incontinent, the person may still be independent in eating if food is served on plate |
| Severe Alzheimer's Disease | The brain no longer directs voluntary activities. Verbal and psychomotor skills are severely limited or lost. The person may be in a stupor or coma. The person is totally dependent in all ADLs. |

**Functional Evaluation:**

The assessment of one's ability to function independently in the activities of daily living is an important aspect before planning a treatment program

According to Zolga (1987) there are 7 "Ws" of the functional evaluation as follows:

- What Can the client do?
- What does the client do?
- How does he/she do it?
- Which Part of the task is the client unable to do?
- why is he/she unable to do them?
- where does he/she performs best?
- when does he/she perform best?

Areas of Assessment:

- Gross motor skills and co-ordination
- Fine motor skills
- Postural control and balance
- Mobility
- Perceptual skills
- Attention span
- Orientation
- Learning skills
- Judgement of safety
- Communication skills
- Social Skills
- Daily living skills
- Productivity skills and interests
- Leisure skills and interests

**Objective assessment:**

Following Scales can be used to assess Patient with Dementia

1. The modified Crichton Geriatric Behavior scale: it is designed be R.A.Robinson. It assesses 8 behaviors on a Likert scale of 0-4 where 0- is normal and 4 is majorly affected. The behaviors listed are Mobility, Orientation, Communication, Cooperation, Restlessness, Dressing, Feeding and continence.
2. SAILS: Structural Assessment of Independent Living Skills:

   The Sails is a set of standardized behavioral tasks which were designed to measure activities of Daily Living with Dementia. The test consists of 50 tasks which measure the functional ability of the patient. These tasks are divided into 10 groups of 5 items each which measure fine motor skills, gross motor skills, dressing, eating, expressive language, receptive language, time and orientation, money-related skills, instrumental activities and social interaction.

3. Mini Mental State Examination
4. Short Portable Mental Status

The first and foremost step towards rehabilitation of a patient requires an appropriate

**APPROACH TO THE PATIENT**

- As the patient may be disoriented and may have memory deficits, each session should be considered as a new on, the therapist might have to start each session afresh while considering that the patient might not recollect the events of the previous day

- Try to engage or initiate with activities which he already knows and remembers, the skills he acquired over years out of daily practice or using the habitual memory, this will develop a trust in therapist and decrease anxiety in anticipation before starting a session. Also, the successful completion of activity will be motivating for the patient to co-operate for further session.
- Gradually start with learning of a new task which needs repeated reinforcement, reminders, praise and rehearsal to ensure significant amount of new learning. Therapist should consider the fact that patient has the ability to new task which is not lost but it is slowed down and little difficult.

**Occupational Therapy Intervention:**

- **Orientation**: following strategies will help patient to get themselves oriented to reality and reduce social isolation:
  - The caregivers in contact with the patient will always introduce them before initiating a conversation rather than asking him whether he remembers about the caregiver or not because this will increase anxiety.
  - The caregiver should constantly give information about the time and place and the surrounding environment assuring the individual understands the information
  - Use of Visual Orientation Aids: wall clocks, calendar, name tags, newspapers, etc.
  - The patient should be trained to associate the visual aids, for e.g. We can label the different rooms in house, if patient has difficulty finding a toilet- always keep the light of toilet on and label it -the patient should be then directed to it, asked to read it and then reminded of the association between going to the sign, reading it and finding the toilet.
- **Attention and Concentration:**
  - The therapy/ patient's room should be quiet with less possible distraction, for e.g. Lot of people, background TV, Radio, etc.
  - Get his attention by calling him by his name while maintaining an eye contact
  - Preferably choose an activity which is short term and structured – sorting colours, shapes, vegetables, etc.
  - Visual instructions should be given with frequent repetitions
  - The task to be performed should be broken down into simple and small basic components
  - Transition to another activity only after completion of present activity.
  - Activities of patient's interest can improve his concentration
- **Memory:**
  - Strategies to register information: rehearsal of the information and/ or using mnemonics (coding by initials, rhymes)
  - Strategies to store information: write the information down and maintain a diary

- Strategies for recall: using different types of cues (LTM-STM)

Semantic cues- e.g. 1 word was a colour

Phonemic cues- e.g. Shoe- for shoelace/ shoe maker

Contextual cues- e.g. national animal- tiger

If the above cues fail- therapist provides the patient with multiple choices and he need to choose the correct

- **Language and communication:** To maintain communicative functioning at optimal level

- Environmental Modification to facilitate Communication:
  - Rearranging furniture to allow person to converse more easily without distraction
  - Involving the individual in family discussions, along with the children in the house
- For Mild Dementias:
  - Speak slowly and clearly
  - Provide multimodal input like gestures and facial expressions
  - Talk about the present, giving the patient enough time to understand and respond
  - Keep the environment consistent
- For Moderate Dementias:
  - Read aloud multiple choice
  - Break down sequential commands into steps
  - Remove distracting stimuli
  - Provide positive consequences for good communication such as hugs, smile and touch.
- For Advance Dementias:
  - Get the patient's attention by calling his name and through gentle touch
  - Talk about objects that are visible. E.g. Showing an orange ask the patient, "do you want orange"
  - Maintain eye contact

5. Perceptual Deficits:

- Apraxia:
  - Cues such as labels to tell the front from the back or taking himself through the steps of putting on each item of clothing
  - An audiotape and a specific method of sequencing can improve dressing
- Topographical Orientation:
  - Role practices and memorization of routes
  - As a means of compensation markers to identify a route, use of maps
  - Always keeping a basic ID card with patient including their basic Data such as Name, Contact.NO, Address, blood group.
- Figure Ground Perception:
  - Scanning activities such as finding objects from a random scattering of objects
  - Word search games are also helpful

- keep the environment as simple as possible and marking significant objects with a bright colour

- Behaviour Management:
  - Behaviour is controlled by environmental factors as well as the consequences that follow a behaviour, thus providing positive reinforcement for the behaviour and not the negative one.
  - Any precipitating or exacerbating factor such as patient or caregiver action etc. should be evaluated.
  - Target one behaviour at a time beginning with the one most problematic.
  - Possible intervention techniques for aggressive behaviour include acknowledging anxious fearful feelings, reassure patient using familiar, calming, simple activities, maintaining consistency in the environment and caregiver.
  - Catastrophic reactions are overreactions to simple incidents which could be the results of emotional instability, misinterpretation of communication, reaction to one's own inabilities. Maintaining an eye contact, speaking calmly and simply can help. Avoid quick transactions and maintain structured and consistent environment.
  - Suspiciousness due to memory loss and confusion which can be helped by giving re-assurances and offering help.
  - Wandering behaviour can be due to disturbances in intellect, communications, restlessness and agitation. Strategies for the same are as follows
    - Establish a regular routine for daily activities
    - Provide safe, supervised area and a controlled environment.
    - Anticipate emotional needs and provide opportunities for socialization
    - To avoid boredom, involve the patient in a suitable activity program considering his
    - likes and dislikes at the same time giving him simple activities.

- Activities of Daily Living:

  Training in ADLs in the demented patient's requires simplifying the environment and using multi-modal (visual, auditory, tactile and motor demonstration) cues to compensate for perceptual as well as cognitive, memory and language losses this type of an environment with an emphasis on procedural memory abilities, which are spared , helps the patient to maintain skills and avoid declines for longer period. Following are the components of activities with problem areas in each and strategies for the same:

❖ Eating:

- Forgets to eat:
  - Provide meals at scheduled time: can put alarms for the meal times
  - Provide sensory stimuli: smell of food, high contrast between the table and plate, plate and food to draw visual attention to the food.
  - Provide verbal and/or tactile cuing

for patient to scoop food, to chew, to swallow.

- Can't / won't sit to eat:
  - ➢ Use food the patient can eat while walking such as cut up veggies, rolls, liquid supplements
  - ➢ Feed several meals rather than 3 large meals
  - ➢ Play soothing music
  - ➢ Avoid stimulating activities immediately prior to meals
- Distractible:
  - ➢ Limit auditory stimuli by turning off music or television
  - ➢ Limit visual input by putting dividers between tables
  - ➢ Remove unnecessary utensils
- Messy:
  - ➢ Use adaptive equipment such as built-up weighted utensils, cups with spill guards or plate guards
  - ➢ Use bowls rather than plates; use cups with large handles.

❖ Dressing:

- Lay out a clean out fit
- Eliminate belts, scarves, sweaters, ties and other accessories that are likely to put on wrongly.
- Clothes in the order in which he puts them on may also be useful.
- If the patient refuses to change clothes, avoid arguments, make suggestion again and later.
- Buttons can be replaced with Velcro, using loos-fitting clothes, slip-on shoes
- Select colours with considerable contrast
- Grooming (brushing teeth): tooth brushing maybe broken down into the following steps:
  - Picking up the toothbrush
  - Putting toothpaste on the brush
  - Bringing the toothbrush to one's mouth
  - Brushing
  - Rinsing
  - Spitting out water
    - ➢ Therapist can use forward or backward chaining method to train them for ADLs once the task is broken into small steps.

❖ Toileting:

- Using Scheduling method: putting alarms after every 1 hour to go to toilet and give a praise if they void urine at correct times in the toile
- Many types of incontinent aids are available. Special absorbent pads or pants can be useful for both urinary and faecal incontinence
- Protective bedding can reduce the discomfort, smell and damage of incontinence
- Occasionally an indwelling catheter maybe necessary

❖ Home Modifications:

- ➢ Maintain adequate lighting
- ➢ Provide visual contrast between items and background.

- Keep pathways & patient's room uncluttered and arranged to allow easy access to bed, chair and bathroom.
- Identify room or areas with signs
- Eg. "toilet" on the toilet door, pictures of clothing on drawers on closet door.
- Use a low bed to make transfer easier
- Create a structured environment by maintaining furniture & equipment in the same place and establish a routine of daily activities
- Use a raised chair to assist with standing. Using a low chair, reclines, bean bag or rocking chair to discourage an unstable patient from ambulating without assistance
- Use a raised toilet chair with or without rails. However, remember that additional equipment such as commode rails can increase the patient's confusion

❖ Sensory Integrative Approach:

- Assumption of SI theory in Dementia: people with Dementia had adequate sensory-integrative functioning but, because of progressive, accelerated or chronic degeneration of sub-cortical structures, there has been a consequent loss in SI function.
- Sensory integrative treatment should be a part of an overall treatment programme aimed at restoring competence in occupational performance
- Aims of Sensory Integrative Treatment are:
  - Increase vestibular, proprioceptive and tactile stimulation
  - Facilitation of normal motor function, especially motor planning
  - Emphasis on body awareness to promote body scheme
  - Use of non-competitive pleasurable activities to reduce anxiety
- Activities with inherent S.I. properties:
  - ADLs: feeding, dressing, cooking, housework
  - Crafts/Arts: pottery, envelope making, making and using puppets, making kites, weaving
  - Leisure and games: carrom, playing musical instruments, square dances, horse riding, swimming
  - Horticulture and gardening
- Emphasis should be laid on using reality orientation in conjunction with S.I.T. previously mastered skills that have been lost should be used. Eg: Self-care, cooking, banking, dancing and crafts
- Gustatory and olfactory stimulation should be included

## Physiotherapy in Dementia

Physiotherapy helps in dementia to address the symptoms present due to dementia. e.g. reduced mobility and pain. Regular exercise and an active lifestyle are very effective in the management of the depression component of dementia.

By slowing down the cognitive decline, exercise can also prevent or delay the onset of dementia.

In addition to its cardiovascular benefits, physical activity may also slow cognitive decline and the incidence of dementia. No RCTs have yet demonstrated that regular physical activity prevents dementia, but increasing evidence suggests that an easy to perform physical activity programme can result in a healthy ageing brain. This argument may convince sedentary individuals to change their lifestyle habits. Physical activity can slow the rapid functional decline and improve the quality of life of the patient. This approach should be systematically proposed as a key non-pharmacological treatment for this population.

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## Physiotherapy Interventions

1. To improve physical function (mobility, balance, coordination and strength).
2. To reduce risk of falls - changes in judgment and spatial control contribute to tendency to fall. Exercises improve balance and reduce the fear of falling.
3. To reduce stress and induce relaxation – Exercises help to reduce the incidence of depression, agitation and aggression symptoms that are common with dementia patient
4. To improve general cardiovascular health.
5. To improve sleep - sleep disorders are common in dementia patients. Exercise can help them get into normal sleep routine.
6. To slow mental decline – exercise seems to slow brain atrophy especially in the hippocampus, which influence memory and spatial navigation.

## An exercise routine should be composed of

1. Flexibility exercises for Musculoskeletal System:

Exercises increases joint range of movement and muscle strength to maintain the normal functions of an individual. It includes: I. Both active and passive ROM (Range of Movement) exercises





Figure 11 and 12: Active range of motion exercises.

## 2. Bed mobility activities

The bed mobility exercises are important as the patient ages they might have difficulties in performing the bed activities. This includes turning, rolling, side lying to sitting, supine to sitting, shifting in bed, activities in all fours and knee standing activities



Figure 13



Figure 14



Figure 13, 14, and 15: Turning and rolling



Figure 16



Figure 17



Figure 16 and 17: All fours and back kicking in all fours



Figure 18

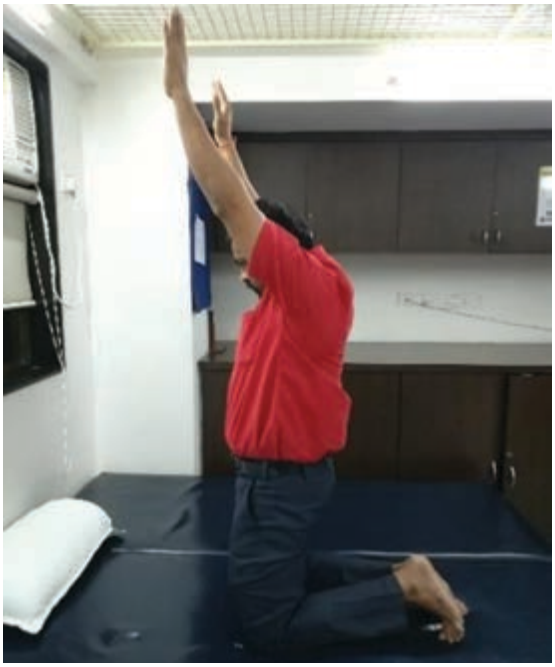


Figure 18 and 19: Kneel standing and hand activities in kneel standing

3. Balance Training: As there is risk of fall increasing with age overall and especially in individuals with dementia, it is important to improve confidence and reduce the risk of falling. As balance is position specific so both standing and sitting balance exercises are encouraged.

Training should begin with weight shifts in both sitting and standing in order to help the patient develop an appreciation of his limits of stability.

Activities include;

- Giving slight push to patient and patient tries to maintain the balance in sitting and standing
- Reaching activities in sitting and standing



Figure 20: Reach outs on balance board

- Heel-toe standing
- Partial wall squats
- Single limb stance



Figure 21: Spot marching

- Marching in place

4. Strength Training: It helps in maintaining the strength of the muscles of upper limb, lower limb and core muscles. Ideally 10-15 repetitions of 8-10 exercises should be performed thrice a week. Resistance may be applied with therabands, weight cuffs, etc.



Figure 22: Upper abdominal strengthening



Figure 23: Back extensor strengthening



Figure 24



Figure 25

Figure 24 and 25: Hip muscle strengthening using weight cuffs

5. Gait Training: Gait re-education helps in improving mobility and functional ability without support. The major goals are to lengthen stride, broaden BOS (Base of Support), improve stepping, improve heel–toe gait pattern, increase contralateral movement and arm swing and provide a program of regular walking. It would improve the confidence in independent walking and reduce risk of fall

Activities include;

- Weight transfer; standing on single limb.
- High stepping to strengthen the flexors.
- Side stepping or crossed stepping with or without support.
- Normal heel-toe progression
- Walking on toes / walking on heels
- Obstacle crossing
- To overcome shuffling pattern, draw foot marks or parallel lines and ask the patient to walk on it.
- The patient should practice stopping, starting, changing direction.

- Auditory cues can be effective in improving gait. Turning of 180 degree should be practiced first then 360 degree.

6. **Aerobic Exercises**: aerobic training helps in improving cardiovascular health, strength the hormones immune system. As physical activity decreased beta-amyloid proteins leading to decreased amyloid plaque. Neural disruption, hence improving brain health. Aerobic exercise includes jogging, cycling, swimming or any physical activity that rejuvenate the patients pulmonary and cardiac capacity. For maximum benefits 30 minutes session thrice a week is advised.
7. **Managing urinary incontinence**: Urinary incontinence is a common problem in patients with dementia. Physiotherapy includes pelvic floor muscle exercises, bio feedback, and electrical stimulation.

Kegel's exercises: Tighten the muscles as if you are holding in your urine, then let go. You should feel the muscles tighten and move up and down.

- Make sure your bladder is empty, then sit or lie down.
- Tighten your pelvic floor muscles. Hold tight and count 3 to 5 seconds.
- Relax the muscles and count 3 to 5 seconds.
- Repeat 10 times, 3 times a day (morning, afternoon, and night)

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# Chapter 11 : Cerebellar Ataxia

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## Introduction:

Cerebellum is an important structure of the hind brain. It is placed in the posterior cranial fossa. It is also known as 'Little brain'. Controlling movements of the body which requires the co-ordination is the primary function of the cerebellum. The function of the cerebellum is to time, coordinate and integrate motor movements, movements of eyes and speech. so, whenever cerebellum is affected, it ultimately affects the movements controlling co-ordination.

**FunctionalRole of Cerebellum:**

Cerebellum is made up of two main parts i.e. Cerebellar cortex and deep cerebellar nuclei.Cerebellum receives afferent pathways from other parts of the brain. It plays a role of aregulatory centre for controlling the movements. It also plays an important role in maintaining and adjusting the posture by regulating the outputs of descending motor systems which is done by adjusting the size of muscle contraction. Normal motor movements are achieved in human beings with constant neuro-developmental maturation following birth and gets completed when normal postural mechanisms become localized, followed by the ability to maintain balance in different positions and lastly by the formation of muscular coordination. Normal postural tone helps in keeping posture erect in standing against gravity, adaptation to changes on support surface and proximal stabilization. Balance can be defined as postural adaptation to changes in gravitational center with the contribution of normal postural tone. Muscular coordination on the other hand, is the functioning of all muscles active during the voluntary motor movement in appropriate rhythm, velocity and amplitude. A person may perform daily life activities through normal motor movement formed by the above mentioned three components. Nervous system diseases and/or injuries usually affect postural control mechanisms where balance problems are more dominant in some types of diseaseswhile in other types of diseases coordination problems related to extremities are prominent.

Cerebellum plays a key role in adapting to the requirements of the task. Therefore, it plays a key role in reorganizational and adaptive process and motor learning. Motor learning is a task where moving is from nonspecific response to highly selective associations is required. (Brooks 1986).

**Role of cerebellum is in**

1. Skill acquisition and refinement.

2. Skill automation and retrieval.

The word ataxia is derived from the Greek word meaning (Ataxia - not ordered) is used to describe number of abnormal movements occur while doing voluntary movements. People with cerebellar ataxia have problems like inaccuracy in achieving the target, inability in doing movements in constant force and rhythm.

## Parts of cerebellum:

The cerebellum consists of two cerebellar hemispheres They are joined together by vermis (Fig 1).There are three lobes of the cerebellum, it is classified into anatomical or structural classification (anterior lobe, posterior lobe &flocculonodular lobe) & physiological or functional classification (spino-cerebellum, cerebro- cerebellum &vestibulo-cerebellum) as shown in figure 2. If there is damage or dysfunction or atrophy in cerebellum it causes imbalance &impaired eye movements (spino-cerebellum), truncalataxia & difficulty in walking (cerebro- cerebellum), in-co-ordination of planned movements &intentional tremors (vestibulo-cerebellum) depending on the part of cerebellum affected.

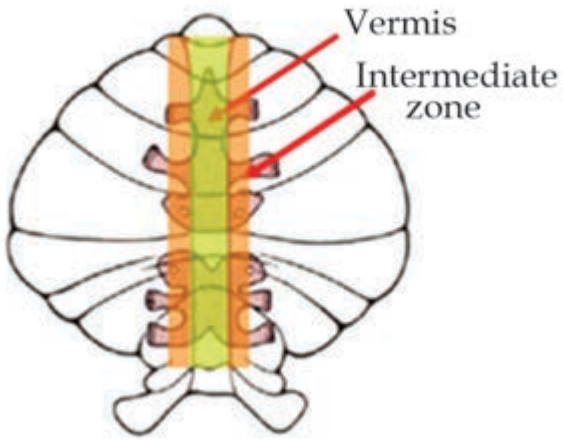



Fig 1: Topographical representation of cerebellum

## Physiological Classification (Function)

- **Spino-cerebellum:** Imbalance & Impaired eye movements
- **Cerebro-cerebellum:** Truncal Ataxia & difficulty in walking
- **Vestibulo-cerebellum:** Inco-ordination of planned movements & Intentional Tremors

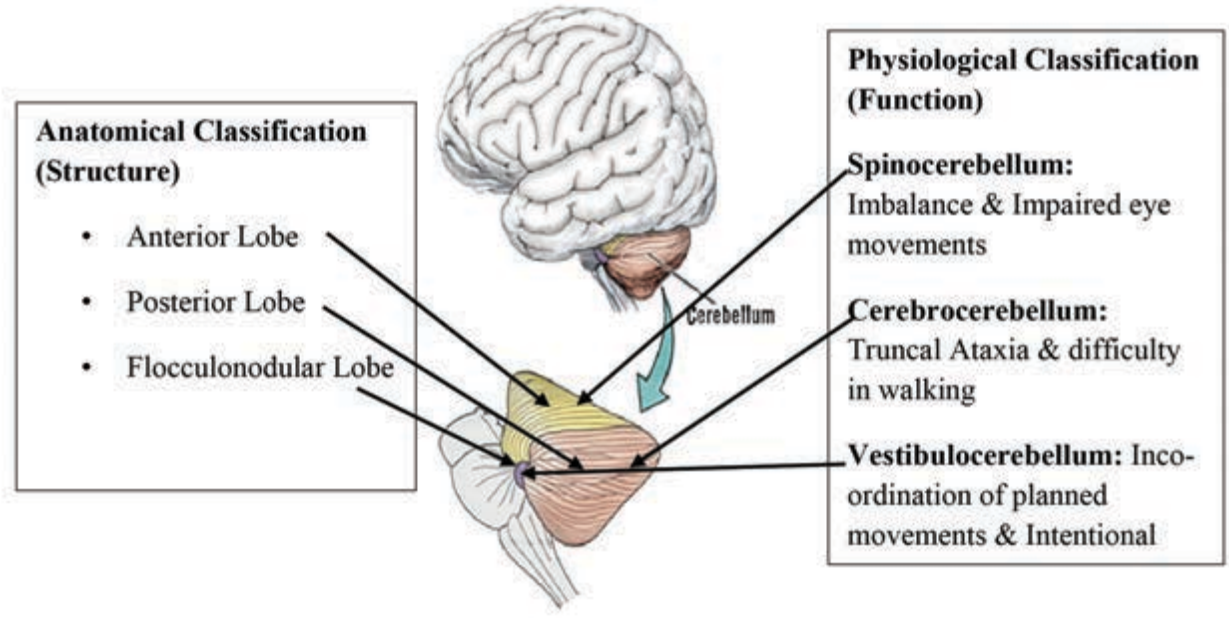



Fig 2: classification of cerebellum

## Anatomical Structures Responsible for Balance and Co-ordination:

Normal motor control is the output of normal postural tone, muscle coordination, and balance working together. Postural control within the normal functioning of the nervous system is maintained by the different sensory-motor subsystems working together within a circular network. The vestibular, visual, somatosensory systems and cerebellum work in a coordinated manner to produce postural orientation depending upon the goal of the movement and environmental conditions. Each subsystem may be dominant at different times. For example, whenever the type or quality of the surface on which the patient is standing changes, the changes will be registered by the somatosensory system whereas when the information in the visual area changes, the visual system will register changes. The subsystems collecting the information at that particularmoment will dominate thereby determining the appropriate motor response (Crutchfield et al. 1989).

### Somatosensory System (proprioceptive andsuperficial senses):

Joints, muscles, ligaments, and tendons have proprioceptors. Information about the muscle strength, position of cervical and lumbar area, joint position is sent to the brain via proprioceptors. Normal motor function is carried out because of contribution of proprioceptive sense of the joint receptors (Sherrington 1907, Bear et al. 2001).The message is transmitted to central nervous system through two paths. The sensory areas of cerebral cortex (area 3,2,1) receives the sensory input form the dorsal column of medial lemniscus. Visual system and proprioception helps to give information about the shape and the size of the movement, which is generated at the motor cortex. Dorsal and ventral spinocerebellar tracts are the second pathways through which proprioceptive input is provided from the spinal cord to the brain. Spino-cerebellum is the area where all the sensory input is gathered. In other words, the sensory input carried by these tracts is the subconscious proprioceptive sense. This sensory input enables the cerebellum to correct the faulty motor commands that the motor cortex may send by informing the cerebrum and cerebellum simultaneously about the size, speed, form and timing of the movement before the movement is performed (Guyton 1976, Ramnani et al. 2001).

**Dysfunction of the dorsal columns of the spinal cord results** in loss of proprioception whichgenerally carries the proprioceptive information tothe brain leading to condition called **Sensory ataxia.** At times it may be due to dysfunction of variousbrain parts which receives theinformation,including the thalamus, and parietal lobes.

**Sensory ataxia shows the following features:** (Fig 3)



Fig 3: Sensory Ataxia

1. High steppage gait and postural instability due to lack of proprioception which cannot be compensated by visual input.
2. Romberg's test is positive when patient's stands with his feet together and eyes closed resulting into instability with wide oscillations and possibility for fall. This finding differentiates sensory ataxia from the other ataxia. (Bannister 1992).
3. Loss of vibration sense in the extremities and loss of deep tendon reflexes.
4. Worsening of the finger-pointing test with the eyes closed.

**Sensory ataxia is generally observed in:**

1. Hereditary types of ataxia such as Friedreich's ataxia and spinocerebellar ataxia.
2. Other diseases like diabetic or alcoholic neuropathy, vitaminB12 inadequacy neuropathy, Tabesdorsalis,
3. Tumoral conditions found in the posterior cord of the medulla spinalis,
4. Multiple sclerosis (Edwards 1996).

## Vestibular System:

Vestibular system has two pathways:

1. Central Pathway.

2. Peripheral Pathway. The central vestibular pathways coordinate and integrate information about head and body movement and use it to control the output of motor neurons that adjust head, eye, and body positions.

The primary vestibular axons from cranial nerve VIII make direct connections to the vestibular nucleus of the same side of the brain stem and to the cerebellum. The vestibular nuclei also receive inputs from the cerebellum, visual and somatic sensory systems, thereby combining incoming vestibular information with data about the motor system and other sensory modalities.

Thevestibular systemis composed of otolithic organs (utricle and saccule) and semicircular canals.

1. **Vestibulo-spinal reflex:** Axons from otolith organs project to the lateral vestibular nucleus, which then projects via the vestibulospinal tract to excite spinal motor neurons controlling leg muscles which helps to maintain posture even on unstable surface.

2. Axons from semicircular canals project to the medial vestibular nucleus, which sends axons

via medial longitudinal fasciculus to excite motor neurons of trunk and neck muscles that orient the head. These pathways help the head to stay straight even if the body moves around. One important function of the vestibular system is to keep eyes fixed in a direction, which is actualized by vestibulo-ocular reflex which works by sensing rotations of the head and immediately commands a compensatory movement of the eyes in the opposite direction. The movement helps to keep gaze fixed on a visual target. (Guyton 1976,Bear et al. 2001).

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## Vestibular ataxia:

This develops as a result of peripheral or central diseases which directly affects the vestibular nuclei or the afferent and efferent connections of the vestibular nuclei.




Fig 4: Vestibular Ataxia

**A patient with vestibular ataxia shows:** (Fig 4)

1. The symptoms of imbalance in standing and sitting. The patient represents stagger, has a broad base support and may tend to lean backwards or towards the side of the lesionwhile walking.

2. Head and trunk motion along with arm swing is often reduced (Borello-France et al. 1994). The patient is limited particularly when crossing the street and shopping at the market.

3. Balance is disrupted when performing a head or eye movement.

4. It may be accompanied by vertigo, nausea, vomiting, blurred vision and nystagmus due to the vestibular system's role in sensing and perceiving self-motion and stabilizing gaze via the vestibulo-ocular reflex (Horak&Shupert 1994). Extremity ataxia is by no means observed in vestibular ataxia.

5. Deep tendon reflexes are normal.

## Vestibular ataxia can be seen in:

1. Central factors such as medullar stroke and multiple sclerosis.

2. Peripheral vestibular diseases such as Menier's, benign paroxysmal vertigo, or vestibular neuronitis.

## Aetiology:

Cerebellar lesions can be developed because of brain injury, birth complications, due to trauma, vascular diseases, hereditary conditions or because of degenerative, metabolic diseases and infections as well.

## Conditions, Disorders which causes Cerebellar Ataxia:

1. **Brain Surgery**
2. **Trauma:**

   Gait instability may persist in following cerebellar, vestibular, brain stem, or frontal lobe injury or with interruption of the fronto- ponto- cerebellar tract.

3. **Mass Lesions:**

   Lhermitte-Duclos disease (dysplastic gangliocytoma) is a benign cerebellar haematoma.

4 **Alcohol abuse**

5. **Drug abuse:**

   It can cause ataxia as they have a depressant effect on central nervous systemfunction. For example, Ethanol can cause reversible cerebellar and vestibular ataxia. Antiepileptic drugs produce cerebellar ataxia as an adverse effect.

6. **Infections** such as Chicken pox.

7. **Exposure to toxic Chemicals:**

Inability to excrete copper from the body is seen in Wilson's Disease which is autosomal-recessive gene disorder. Copper accumulates in the nervous system and liver causing ataxia aswell as other neurological and organ impairments.

8. Demyelinating disorders like **Multiple sclerosis**, direct or indirect trauma caused to brain seen in conditions like **Cerebral palsy** and some other neurological conditions can also lead to Cerebellar Ataxia.

9. **Malformation** of the cerebellum while the baby is still in womb.

10. **Radiation Poisoning:**

Ataxia can be induced as a result of severe acute radiation poisoning with an absorbed dose of more than 30 grays.

11. **Congenital Disorders**

Dandy Walker Syndrome, with ataxia, nystagmus, cranial nerve palsies, apneic episodes, hydrocephalus, and cognitive dysfunction, demonstrates hypoplasia of the cerebellar vermis and a CSF collection that is predominately posterior to the cerebellum but continuous with the fourth ventricle.

**12. Paroxysmal Disorders Associated with Ataxia:**epilepsy, migraine, and high systemic fever.

**What are the common symptoms seen in Cerebellar Ataxia:**(Fig 5)

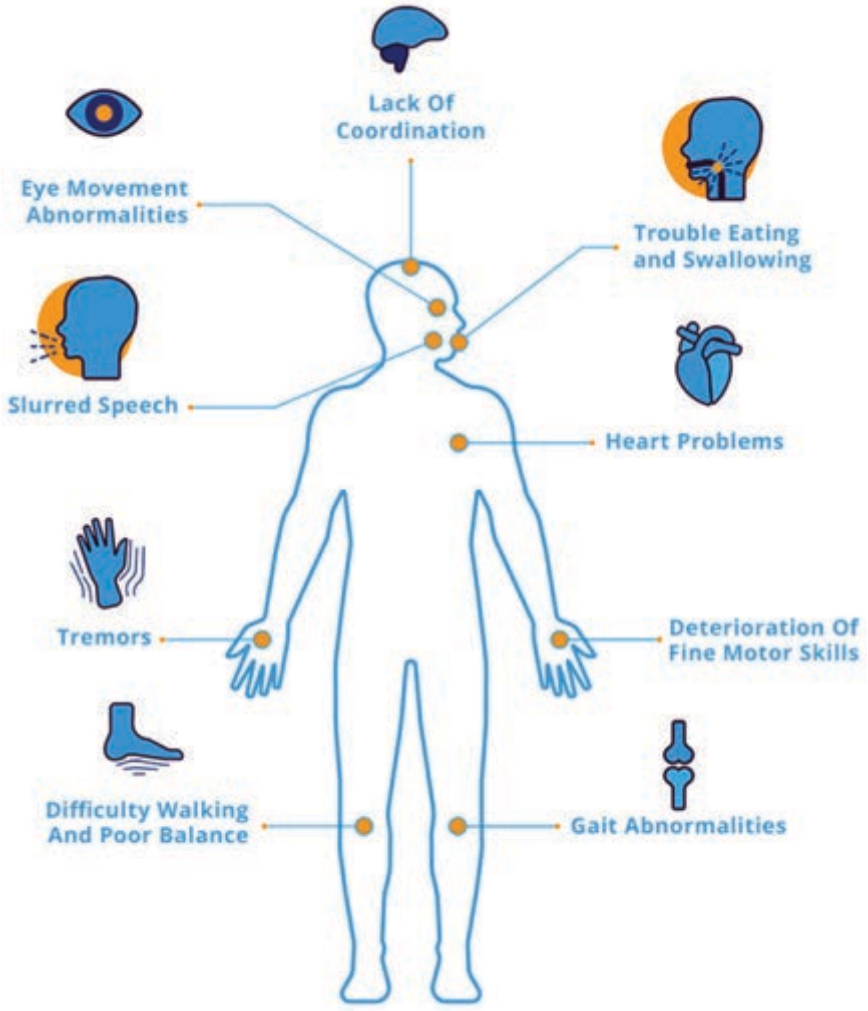



Fig 5: Symptoms of Cerebellar Ataxia

**Ataxia:** Lack of co-ordination in doing voluntary movements (Fig 6)

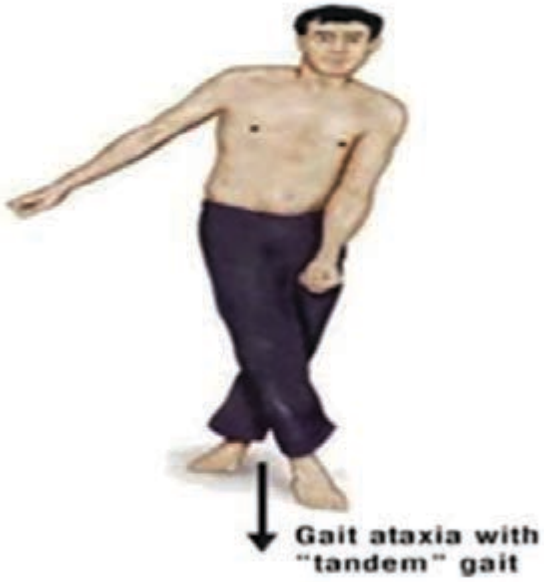



Fig 6: Ataxia while walking

**Hypotonia:** Decreased muscle tone.

**Asthenia:**Generalized weakness

**Dyssynergia:** Movement performed in sequence rather than performing as a whole movement.

**Dysmetria:** Inability to judge distances or range of movements (Fig 7)




Fig 7: Dysmetria

**Dysdiadochokinesia:** Inability to perform rapid alternating movements (Fig 8)




Fig8: Dysdiadochokinesia

**Tremors:** Involuntary, oscillatory movements of the limbs

**Dysarthria:** Difficulty in articulation

**Nystagmus:** Involuntary moments of the eyes (Fig 9)

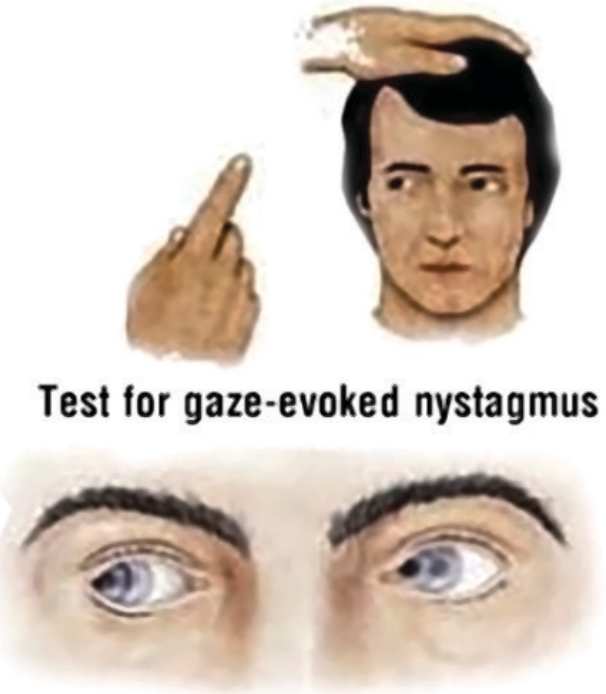



Fig 9: Nystagmus

**Rebound phenomenon:** Inability of the antagonist muscle to contract when the resistance is offered to the certain body movements. (Fig 10)

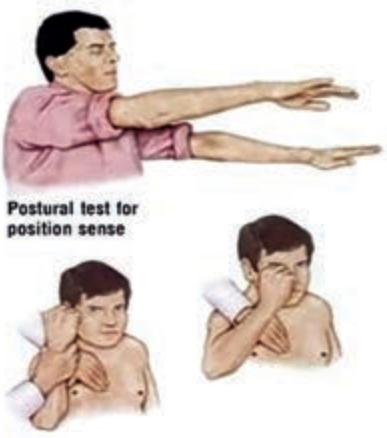



Fig10: Rebound phenomenon

**Pendular knee jerk:** It is demonstrated by irregular knee jerk usually demonstrated by more than **3 oscillations** equidistant from the neutral position of the knee.

**What are the different types of Cerebellar Ataxia?**

Cerebellar ataxia can be divides commonly into two subtypes- **Hereditary ataxia** and **Sporadic ataxia.**

**Hereditary ataxia:** It is caused because of the mutation in the specific gene, which is present right from the birth. This is further divided into two sub categories as autosomal dominant and autosomal recessive type.

**Sporadic ataxia:** This type of ataxias usually seen during adulthood, with no significant positive family history.

**Features of basic types of ataxia** are briefed in Table 1 below

| Type | Cerebellar Ataxia | Sensory Ataxia | Frontal Ataxia | Vestibular Ataxia |
|---|---|---|---|---|
| **Head posture** | Upright and sometimes fixed | Leans forward | Leans forward | Upright and definitely fixed |
| **Trunk posture** | Stooped-leans forward | Stooped-upright | Upright | Upright |
| **Stance** | Wide-based | Wide-based | Wide-based | Wide-based |
| **Initiation of gait** | Normal | Normal-wariness | Start hesitation | Normal |
| **Postural reflexes** | + /- | Intact | May be absent | + /- |
| **Steps** | Stagger-lurching | High-stepping | Small-shuffling | Normal |
| **Stride length** | Irregular | Regular | Short | Normal |
| **Leg movement** | Variable, ataxic | Variable hesitant and slow | Stiff, rigid | Normal |
| **Speed of movement** | Normal-slow | Normal-slow | Very slow | Normal-slow |
| **Arm swing** | Normal, exaggerated | Normal | Exaggerated | Normal |
| **Turning corners** | Veers away | Minimal effect | Freezing - shuffling | Dysequilibrium |
| **Heel-toe test** | Unable | + / - | Unable | Unable |
| **Romberg's test** | + /- | Increased unsteadiness | + /- | – |
| **Heel-shin test** | Usually abnormal | + /- | Normal | Normal |
| **Falls** | Uncommon | Yes | Very common | Common |

Table 1: Types of Ataxia

**Spino-cerebellar Ataxia:**

It is hereditary & autosomal dominant ataxia. It is also degenerative and progressive in nature

- Classification:
  SCA (Type 1 to 29 & 35)
  Signs & symptoms:
  a. Inco-ordination
  - Gait
  - Hand
  - Eye movements
  a. Dysarthria (speech disturbances)

**Genetic-biochemical basis for classification**

Initially, on the basis of anatomical localization of pathological changes, inherited ataxias were classified. In 1993, Harding introduced another classification in which the ataxias were placed into 3 categories:

1. Congenital.
2. Inherited metabolic syndromes with known biochemical defects.
3. Degenerative ataxias of unknown cause.

The last category was subdivided further into early onset (< 25 years) and late-onset types

**In ataxia:**

1. Presentation can be variable (For example: static vs progressive, intermittent vs chronic, early vs delayed).
2. Mode of inheritance also varies such as a utosomal dominant, recessive,and non-mendelian inheritance patterns (where nonmendelian inheritance patterns refers to disorders of inheritance for which the rules of Mendelian genetics do notapply).

Disorders of triplet repeat expansion and certain mitochondrial defects are example of non-mendelian inheritance.

## Functional Difficulties Seen in people with cerebellar Ataxia:

**1. Ataxic Gait:**

It is a typical sign of a person with cerebellar disorders. People with cerebellar disorders walk with wide base of support and irregular stepping. Step length is reduced because of which cadence is also affected. Functionally, Patients with cerebellar dysfunctions have difficulties in walking on uneven surfaces, doing outdoor and indoor mobility activities, climbing stairs.

**2. Postural Adjustments:**

Patients with cerebellar lesions have difficulties in maintaining stationary position especially in standing position. Postural adjustments are required to shift the body mass but at the same time, to ensure that the movement doesnot lead to unwanted fall of the body. People with cerebellar ataxia have difficulties in maintaining the steady posture Most difficulties are seen if they have to perform activities where center of body mass moves beyond the certain radius.

**3. Rising on Tiptoes in standing:**

This is a functional task which require forward shifting of the body mass. Timing of the muscle activations and pattern of force production is the key factor in achieving tis functional task.

**4. Postural Sway:**

It has been found out that, patient who have cerebellar signs have increased postural sway, especially in standing. Comparatively, the swaying is less in patients who have degenerative vermis lesion.

**5. Upper Limb actions:**

Functional problems are seen in performing activities which require goal directed reaching and pointing. More Inco ordination is observed in individuals who try to perform upper limb activities fast as compared to individuals who perform same activities slower.

**How to diagnose Ataxia?**

- Clinical features
- Medical history
- Family history
- Physical examination

The following tests are performed:

1. Magnetic resonance imaging (MRI) or Computerized Tomography (CT) scan to determine whether there is any brain damage.
2. Genetictests: to determine whether the patient has in her itedataxia.
3. Urine tests: Urinalysis may suggest specific abnormal ities that are associated to some types of ataxias.
4. Blood test: to rule out any specific type of ataxia.

**MEDICAL TREATMENT**

Several drugs used to control ataxia including muscle elaxants like diazepam which also reduces muscle spasm.

A detailed neurological assessment or evaluation of an ataxic patient is done which includes:

1. Tone
2. Posture
3. Co-ordination
4. Balance
    i. Sitting
    ii. standing
5. Gait
6. Functional independence
7. Speech
8. Swallowing
9. Fine motor activities
10. Vision
11. Hearing
12. Bladder & Bowel
13. Cognitive assessment

Depending on the above assessment, the treatment protocol is designed by a physical therapist. The treatment protocol can vary depending on the type, characteristics & severity of ataxia. Forinstance, while approaches which improve proprioception and incorporate visual aids are used more commonly in patients with sensory ataxia, stabilization training is more important to reduce truncal and extremity ataxia in patients with cerebellar ataxia. The patient with vestibular ataxia should be given habitation exercises in order to reduce vertigo, and also vestibulo-ocular, vestibulo-spinal reflexes should be stimulated to improve balance.

**The goals can be briefly described as:**

1. Improving balance and postural reactions against external stimuli and gravitational changes.

2. Improving and increasing postural stabilization following the development of joint stabilization.

3. Developing accuracy of limb movements incorporating placement of both upper and lower extremity when performing all functional activities.
4. Developing independent and functional gait including clearance of obstacles,increasing the patient's independence while performing activities of daily living.

**Main principles of training**

1. Whatever be the pathology, treatment requires repetitions of a task and task sequence which will help in execution of both slow and rapid movements.
2. Exercises should be practiced consciously at first, and in later stages should be followed by automatic exercise-activities.
3. Exercises should progress from simple to complex-activities (for example: dual tasks)
4. Exercises should progress from wide base of support to narrow base of support.
5. Activities should be practiced first with the eyes open and later with the eyes closed.
6. After achieving proximal to nus and stabilization, the coordinated movement of the distal segments should betaken into consideration.
7. Activities should be practiced in all planes such as sagittal, frontal & transverse plane [for example: reaching out for object in sitting or standing with the center of gravity (COG) reaching the limits of base of support (BOS) known as the limit of stability (LOS)]
8. Activities should be designed to train anticipatory & compensatory mechanisms.
9. Compensation methods and supportive aids and equipment should be employed whenever necessary.
10. Treatment should be supported by an appropriate home exercise program and sports activities.

**A physical therapy protocol consists of:**

a Bed Mobility Activities

b Strengthening of core & extremities

c Co-ordination exercises

d Balance exercises in sitting & standing

e Gait Training

f Stair climbing

g Visual Rehabilitation

h Respiratory exercises

i Bladder exercises

Intervention slike Proprioceptive Neuromuscular facilitation (PNF), Frenkel' sexercises, dynamic training of postural stability with task and activity focus, gait and balance training; along with strengthening and flexibility can be included to achieve the maximum functional independence. PNF approach uses spiral and diagonal components of movements rather than the traditional movement sin cardinal planes of motion with the goal of facilitating movement pattern that will have more functional relevance than the traditional technique of strengthening individual group of muscles. This is used by resistance with the goal of facilitating "irradiation" of muscles to other parts of the body associated with the primary movement.

The **classification of treatment applications** can be briefly described as follows:

**A Bed Mobility Activities**

**a Turning & Rolling:**

A facilitation of turning & rolling is achieved by recruitment & activation of scapular elevators & depressors, abdominals, back extensors, lateral trunk flexors, hip flexors, hip extensors & hip abductors, knee flexors & extensors.



Fig 11: Turning & rolling

The PNF technique of ulnar thrust with scapular elevation & depression along with visual cues to a patient in order to complete a given task of turning & rolling (Fig 11). A facilitation of anterior depression of scapula along with anterior elevation of pelvis to strengthen lateral trunk flexors to achieve turning from supine position & vice versa.

**b. Bridging & Unilateral Bridging:**

Bridging is usually performed to improve truncal control & to reduce or eliminate truncal ataxia. This is achieved by an activation of back extensors, hip extensors, hip abductors, knee extensors from crook lying position along with an eccentric control of back extensors & hip extensors while lowering from bridging to crook lying position. (Fig 12a)



Fig 12a: Bridging

Once bridging is achieved with proper truncal control, the patient is advised to progress the exercise from bridging to unilateral bridging. (Fig 12b)



Fig 12b: Unilateral Bridging

This helps to activate & strengthen back extensors, unilateral hip extensors, unilateral hip abductors. Initially, patient may require minimal assistance to perform & it can be done in short lever or long lever depending on the truncal control of the patient.

**b. Supine to sit:**

In patients with ataxia due to poor truncal control & poor co-activation of truncal muscles, supine to sit is difficult for these patients. Here, the patient is advised to raise their head & neck into flexion then come on elbows while shifting their body weight on elbow & straightening it. Hence, achieving the supine to sit position. (Fig 13)



Fig 13: Supine to sit

This position is achieved by concentric contraction & sustainability of abdominals with maintaining stability (in shoulder) over mobility (in elbow). Timing for emphasis,a PNFprinciple, on elbow extensors & shoulder rotators can be used in patients with poor elbow control & sustainability. Supine lying from sitting position is achieved by eccentric control of abdominals, this can be activated using combination of isotonic in PNF technique.

**c. Shifting forward, Backward &side-wards:**

Patients with ataxia have difficulty in shifting forward, backward & sideward in bed as they lose balance due to poor truncal control, reciprocal innervation & co- activation of postural muscles.



Fig 14a: Shifting forward in bed

The patient is taught anterior & posterior elevation of pelvis to perform bottom shuffling forward & backward. The patient is also advised forward trunk bending to prevent imbalance while performing shifting. (Fig 14a &14b)



Fig 14b: Shifting backward in bed

**d. Prone on forearm & hand:**

The patient is advised to perform pushups in prone with as shown in the figure 15 in order to strengthen scapular muscles, back extensors, shoulder & elbow extensors in closed kinematic position.



Fig 15: Pushups in prone



Fig 16: Quadruped & quadruped with arm & leg extension

Here, the patient is advised to raise the lower extremity while being in quadruped position without losing balance. This is further progressed by raising alternate upper extremity & lower extremity with sustaining the position for approximately 10 seconds. (Fig 16)

**f. Crawling – Forward & Backward**



Fig 17a: Crawling forward

Crawling forward & backward helps to improve co-ordination, inter limb dissociation, co-activation of postural muscles& activation of core muscles, weight shifting, weight bearing, while maintaining the crawling position without losing balance. (Fig 17a & 17b)



Fig 17b: Crawling backward

**g. Kneel sit to stand:**

The patients with ataxia are advised to perform kneel sit to stand in order to improve truncal control, graded control of trunk over pelvis, co-activation of postural muscles. (Fig 18)



Fig 18: Kneel sit to stand

This can also be achieved by applying combination of isotonic, a PNF technique, to have an eccentric control of hip extensors & knee extensors along with concentric contraction in smaller ranges. This activity can be progressed in kneel standing with overhead activities. (Fig 19)



Fig 19: Kneel standing with overhead activities

**h. Kneel walking- Forward, Backward, Sidewards**

The patients are advised to perform kneel walking forward, backward & sideward in order to improve truncal control, graded control of trunk over pelvis, inter-limb dissociation, co-activation of postural muscles& to improve weight shifting. This activity is trained in parallel bar then progressed on mat to eliminate the assistance & improve level of independence. (Fig 20)



Fig 20: Kneel walking in parallel bar- Forward & sidewards

A patient is trained with anterior elevation of pelvis & strengthening of hip flexors, hip abductors & hip extensors. The anterior elevation of pelvis is achieved by rhythmic initiation & can be progressed to combination of isotonic, a PNF technique, to strengthen quadratus lumborum. The principle of irradiation & re-inforcement can be used to strengthen hip extensors & hip abductors.

**i. Half Kneeling**

The patient is encouraged to perform half kneeling in preparation to standing. (Fig 21)



Fig 21: Half kneeling

### j. Getting up from floor

The patient is advised to come into kneeling position from cross legged sitting, then to half kneeling from kneeling position & later standing. (Fig 22)



Fig 22: Getting up from floor

### A. <u>Strengthening of core & extremities</u>

- **Upper extremity strengthening**

The strengthening of scapular muscles using weight cuffs in prone lying with upper extremity out of the plinth in thumbs down position.

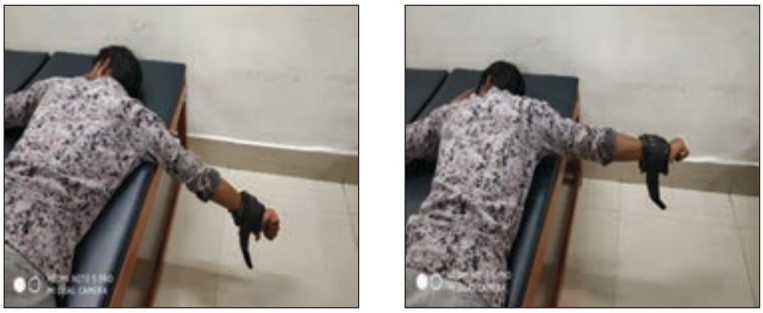


Fig 23: Scapular muscles strength training

Here, the patients are advised to keep their elbow straight position while performing this activity (Fig 23).



Fig 24: Strengthening of scapular muscles, shoulder & elbow muscles

Strength training of shoulder, elbow muscles with scapular muscles using weight cuffs.

- **Trunk Strengthening**

i. Upper Abdominal strengthening

The strengthening of abdominals can be achieved by using chopping pattern of PNF with appropriate visual cues to the patient. Here, the therapist achieves strengthening along with eccentric control of abdominals using combination of isotonic technique in association to replication of the task. (Fig 25)



Fig 25: Upper abdominal strengthening

ii. Lower abdominal strengthening

The patient is advised to tilt the pelvis posteriorly & raise both the lower extremities with hip knee flexion (short lever) then it can be progressed to long lever activity. (Fig 26)



Fig 26: Lower abdominal strengthening

The patient is given command of holding the position for 10seconds & lowering it to neutral position while keeping the pelvis posteriorly. Also, the patient is asked to keep breathing while performing this exercise.

iii. Back extensors strengthening

Depending on the voluntary control of back extensor, strength training is achieved by various grade of exercises. The patient is brought out of the plinth & asked to clasp the hands behind the back (grade 3).



Fig 27a: Grade 4- Back extensor strengthening

Then this exercise is progressed to (grade 4) the patient clasps the hands behind the head & finally to (grade 5) upper extremity in extension (Fig 27a &27b). Here, with change in position of hands makes the activity difficult as the center of gravity (COG) is shifted higher.



Fig 27b: Grade 5- Back extensor strengthening

- **Lower extremity strengthening**

i. Hip& knee musculature strengthening:



Fig 28a: Hip flexor, abductor & extensor strengthening

A combination of isotonic & dynamic reversal techniques are used to strengthen the lower extremity musculature. According to PNF principle, strength training can be further achieved in functional position.



Fig 28b: Hip & knee strengthening using PNF pattern & technique

This pattern shown in (Fig 28b) can be used for training of walking. Using the reinforcement principle, even the other hip & knee muscles can be strengthened with same pattern & technique.

**B. <u>Co-ordination exercises:</u>**

- Finger to Nose exercises



Fig 29: Finger to nose exercise

- Finger to finger exercises
- Bilateral co-ordination exercises



Fig 30: Bilateral co-ordination exercises

- Rapid Alternating hand exercises
- Frenkel's exercises

Frenkel's exercise is performed in a slow manner & practiced repetitively. This improves eye-hand & eye- leg co-ordination in patients with ataxia as the patients are advised to look at their hand or arm & leg movements & correct it accordingly while performing the exercise.

**1. Supine:**

a) Flex and extend one leg, heel sliding down a straight line on table.

b) Abduct and adduct hip smoothly with knee bent, heel on table.

c) Abduct and adduct leg with knee and hip extended, leg sliding on table.

d) Flex and extend hip and knee with heel off table.

e) Place one heel on knee of opposite leg and slide heel smoothly down shin toward ankle and back to knee.

f) Flex and extend both legs together, heels sliding on table.

g) Flex one leg while extending other leg.

h) Flex and extend one leg while abducting and adducting other leg.

**2. Sitting:**

1. Place foot in therapist's hand, which will change position on each trial.
2. Raise leg and put foot on traced footprint on floor. Sit steady for a few minutes.
3. Rise and sit with knees together.

**3. Standing:**

Place foot forward and backward on a straight line. (Fig 31)

1. Walk along a winding strip.
2. Walk between two parallel lines.
3. Walk, placing each foot in a tracing on floor.



Fig 31: Frenkel's exercises in standing

- Bouncing the ball in standing

## C. Balance activities in sitting & standing:

- **Static sitting posture& balance training**

A tactile feedback can be used to correct the slouched sitting posture in patients with ataxia.



Fig 32a: correction of sitting posture

After attaining the erect sitting posture with tactile feedback, it is also advised to keep the feet supported to maintain erect sitting posture. It helps to assist in activating postural muscles to obtain straight & upright posture. (Fig 32a & 32b)



Fig 32b: correction of sitting posture with feet supported

- **Dynamic posture &sitting balance training**

i. Sitting with reaching out for objects overhead

In patients with ataxia due to poor truncal control & poor co-activation of postural muscles, they find difficulty in performing overhead activity (Fig 33). The patient compensates with slouching to maintain their stability while performing overhead activities



Fig 33: overhead activity with slouched posture

The patient is encouraged to perform overhead activity using unilateral or bilateral upper extremity while maintaining the erect posture. (Fig 34)



Fig 34: Sitting with reaching out for objects overhead

This activity is achieved by upper extremity pattern along with approximation (compression in the line of joint) at the end of the movement to improve co- activation of core & postural muscles, and to improve joint proprioception. Performing this activity is encouraged with proper feet support. This also help to improve visual guided movement & visual fixation.

ii. Sitting with pivot turn



Fig 35: Pivot turn in sitting

iii. Sit to stand & stand to sit:

- From same level



Fig 36a: sit to stand from chair in parallel bar

Using rhythmic initiation technique & rocking exercise, the patient is trained to achieve sit to stand. The patient is also trained for weight shifting forward & taking weight on lower extremities. (Fig 36a & 36b).



Fig 37b: stand to sit

Initially, the patient is trained for facilitation of concentric & eccentric control of hip extensors, abductors & knee extensors along with trunk muscles.

- **From lower level**



**Fig38: sit to stand from lower level**

- **Standing posture &balance training**

i. Static Standing balance training

A stabilizing reversal can be used for core activation in patients with ataxia. Then it can be progressed to rhythmic stabilization. (Fig 39)



Fig 39: Stabilizing reversal for core activation

ii. Dynamic standing Training

a) Standing balance training with reach outs using gutter walker

b) Overhead activity using PNF technique

An upper extremity pattern & technique is used to achieve overhead activity in standing along with approximation at the end of the movement to improve core co-activation & joint proprioception. (Fig 40)



Fig 40: Overhead activity in standing

c) Standing with overhead activity & pivot turn

d) Standing balance training on balance board



Fig 41: Balance board exercises in standing

e) Standing with feet together

f) Standing with functional activity

**D. Gait training**

The patient is guided by weight shifting techniques & facilitation of lower extremities. An anterior elevation of pelvis is associated with hip & knee flexion to take a step forward. Then the patient is guided to weight shift on mid stance leg. (Fig 42)



Fig 42: Gait training

In midstance phase, the patient tends to hyper extend the knee due to affected joint proprioception. The technique of stabilizing reversal can be used to improve co- activation of knee muscles in midstance phase of gait cycle. It can be practiced with replication of stepping forward & backward. (Fig 43)



Fig 43- Activation of dorsiflexors using PNF technique in gait training

The patient with ataxia is trained to walk independently with walker or rollator. This is the progression of gait training from parallel bar to walking under supervision initially then to independent walking (Fig 44)



Fig 44- Independent walking with rollator

Also, the patients are advised arm swinging while walking as these patients tend to keep their upper body stiff in order to maintain their balance with poor trunk dissociation over pelvis.

- **Obstacle crossing**



Fig 45: Obstacle crossing

**E. Stair climbing& descending**

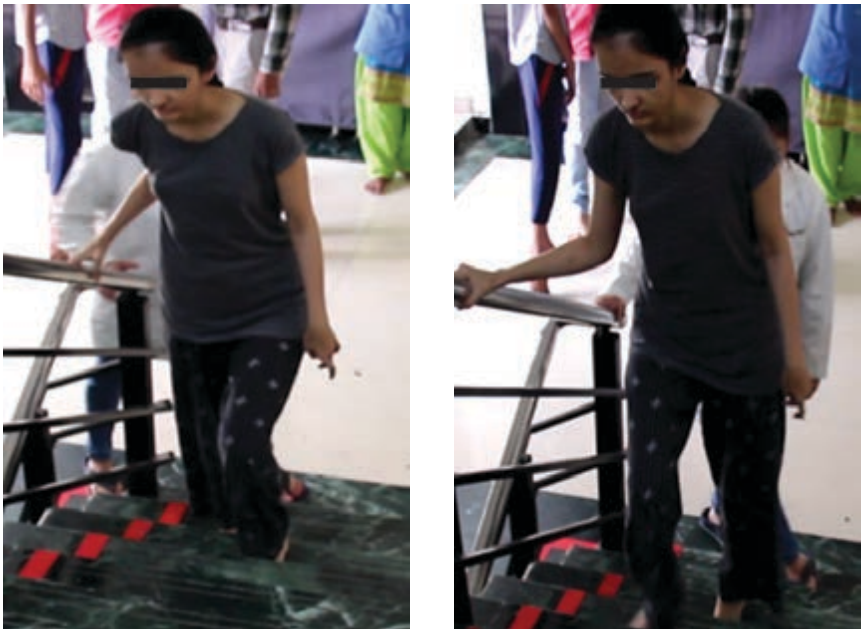


Fig 46: Stair climbing

The patients with ataxia have poor quadriceps control, weak hip extensors & abductors while descending the stairs.



Fig 47: Descending from a stepper

To improve quadriceps eccentric lengthening & strengthen hip abductors & extensors using PNF technique of combination of isotonic. (Fig 47 & 48)



Fig 48: Descending on stairs

**F. Visual rehabilitation**



Fig 49:Fixating eyes at the object with head movement



Fig 50: Focusing the object near & far

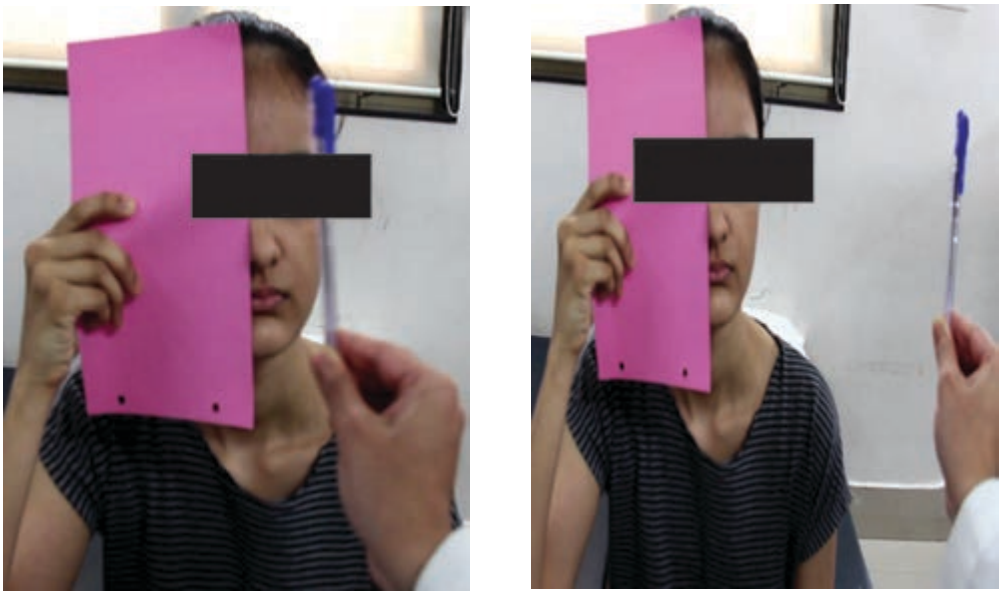


Fig 51: Eye fixation on the moving object without head movement with patch on one eye

## Interventions used to improve vestibular dysfunction:

Since dizziness accompanies balance dysfunction in vestibular problems, repetitive head movement and Cawthorne and Cooksey exercises (Dix 1979) are of great importance. A vestibular exercise program consists of repetitive,progressively more difficult,eye, head and body movement designed to encourage movement and facilitate sensory substitution.

## Management of spasticity:

Most commonly used medications are

i Baclofen (Lioresol), Tinzanidine (Zanaflex)

ii Other drugsare-

- Sodium dantrolene (Dantrium) - induce weakness
- Diazepam (Valium)- Spasms (in Night) Cyclobenzaprine (Flexoril)- Back spasm
- Carbamepazine (Tegretol)- Tonic Spasms

There are possibilities of one side effects with overdose of these drugs including sedation, weakness and fatigue but reduction in spasticity.

a) Apart from oral medications,

   a) Topical cold or hydrotherapy can temporarily reduce spasticity by decreasing tendon reflex excitability and clonus and by slowing conduction of impulses in nerves and muscles.

   b) Intermittent static stretching held for minimum of 30 to 60 seconds be applied ideally for 5 to 10repetitions.

   c) Active and passive ROM exercises

   d) Weight bearing exercises

   e) Aquatic therapy

   f) Serial casting in inhibitive postures has been seen to be effective in tone reduction.

## Occupational therapy management

Occupational therapy (OT) is widely recognized to be a valuable part of the multi-disciplinary care team in ataxia.

The occupational therapist working with people suffering from ataxia will focus mainly upon how the common impairments will have impact on activities of daily living affecting the functional life and occupations and hence provide specific interventions. In addition, an occupational therapist will play an important role for patients with progressive neurological conditions in maintaining independence and quality of life and to enable people to participate in self-care, work and leisure activities that they want or need to perform.

When it is no longer possible to maintain usual activities, Occupational therapist will support people in changing and adapting their relationship with their physical and social environment to develop new valued activities and roles.

The focus of OTs on 'engagement' in activity, rather than the disorder is important in progressive conditions and OT intervention should focus on goals that support the person and carers thereby adding to quality of life.

OTs should use assessment and outcome tools that measure the person's satisfaction with the performance of an activity; use of tools that measure impairment would not demonstrate the effectiveness of OT

intervention. Appropriate tools include, but are not limited to, AMPS (assessment of motor and process skills), GAS (goal attainment scale), COPM (Canadian Occupational Performance Measure), self-efficacy tools and quality of life measures.

## Common issues encountered with Activity and Participation

The effects on a person's occupational performance are not predictable and will depend on the types of activities that the person needs and wants to participate in. It is important to take a client-centered approach to analyzing the problem areas with performance of daily activities and roles.

As most ataxias are progressive an important consideration is proactive planning for future needs.Evidence suggests that people with ataxia may have a lower quality of life in the early and end stages of the condition. It is therefore important to recognize that even at the early stage of the condition difficulties with roles and activity engagement may benefit from support. This can be a difficult situation to deal with in a sensitive manner and OTs must respect the individuals in their own journey of acceptance of this condition. If appropriate, OTs should broach the expectation of decline in the future when considering any major adaptations.

## Common interventions

- **Self care and toileting**

Aims of treatment include minimizing the impact of excessive movement and helping the person to optimize their independence where possible.

Toileting is an area that people with ataxia report as difficult and stressful. Rails can help the person to fix their arms and provide greater stability during transfer. Other problems encountered include dressing and undressing to toilet and perineal hygiene.

## Practical suggestions

- Encourage sitting to bath or shower and consider providing seating with support for the back and arms
- Use of the rmo-regulation devices on taps can be an important safety consideration if the person has difficulty with using taps
- Lever taps may be easier to use than standard taps
- Level access shower can be a useful consideration for a person with ataxia if bath transfers become unsafe or dangerous
- Zippers may be overcome via the use of 'zip pulls' and fastenings could be replaced with Velcro instead
- Button hooks can also be helpful
- An add-on bidet or an automatic washing/drying toilet such as a Closo-mat® (www.clos-o-mat.com) or similar device may help
- Toilet rails around the toilet may be of benefit and wherever possible these should be fixed to minimize risk of accidents
- The height of the toilet seat is to be considered and adapted whenever it is required

- **Eating and drinking**

Feeding needs to be considered due to multiple impairments impacting on safe

and effective eating and drinking. Before commencing any feeding assessment, standard practice would be to ensure that the need for a speech and language therapy assessment is considered. Feeding solutions varies depending on contextual factors and solutions for eating at home varies for social events. Altering positioning and/or seating will maximize posture and support core stability, thus reducing the impact of excessive limb movement.

## Practical suggestions

- Reduce clutter and organize work spaces to optimize the performance.
- Plate guards, and Dycem® non-slip matting can be used as a placemat to limit movement of plate/ cup (see www.dycem.com)
- Use lidded/insulated cups or cups with straws for drinking, especially hot liquids such as tea and coffee
- To assist with drinking, use of travel mug with lid can, will avoid spillage.
- Cups with anti-tremor insert devices can help
- Use of the Neater-eater® device, with its dampening hydraulic mechanism can be very effective in aiding independent spoon or fork feeding (www.neater.co.uk)

## • Food preparation

Preparing food is one of the common concerns in the early stages due to obvious risks. An Occupational Therapists will carry out an activity analysis of food preparation tasks and suggest a variety of methods and aids that may compensate for difficult or unsafe aspects of tasks. This may include completing food preparation tasks in a seated position, having someone else do aspects of the tasks for them, but not the whole task such as cutting vegetables, which could be difficult or use of devices to aid grip and maximize safety. (Fig 52)



Fig 52: Food preparation

## Practical suggestions

- Kettle tipper devices can help making drinks safer
- Waist height ovens; use of full-length oven gloves; sliding food to a level surface (or level trolley) rather than lifting
- A microwave oven can provide a safer alternative to standard ovens
- Chopping boards with an attached cutting blade can be safer than a separate knife
- A food processor can help with slicing or chopping vegetables

## Household management

It is important to identify areas of household management important to the person and to recognize the cognitive and physical elements to these tasks. Most people with ataxia will continue to be able to cognitively manage the home but may have difficulty in physically doing

heavy housework such as vacuuming or heavy laundry. Fatigue management, energy conservation techniques need to be discussed prior to listing the priorities.

### Practical suggestions

- Ensure that people have all the benefits that they are entitled to such as DLA (Disability Living Allowance), which can manage the extra costs of living with a progressive ataxia (for example hiring of a home help to assist with heavy household tasks)

## • Bed, chair and toilet transfers

It is necessary to ensure that the height of the chair is correct for the person to transfer easily on and off. Ensure that the hip and knee angle is at 90 degrees and that the feet are flat on the ground. Armrests greatly enhance the ease of chair transfers and this should be considered when using high-backed dining chairs with arms. The chair should be stable and the armrests should be at a suitable height and position to enable the patient to push up. Educate the patient and carer on sit to stand techniques. Importantly, consider for the carer's safety.

Some people will need hoist provision for transfers and Occupational Therapists should ensure that they are adequately trained to perform this and that it is undertaken by a relevant team member. In particular, where full body tremor presents, slings need to provide the most support possible for safety reasons.

### Practical suggestions

- The height of the bed and location in the room should be considered.
- A bed lever is of benefit which aids to rolling and rising in bed
- Profiling beds, Mattress variators will be of benefit
- The surfaces should be at the optimal height to ensure the most efficient and safest transfers
- Firmer mattresses will aid bed mobility
- Pressure care needs should be considered if mobility is severely restricted

## • Indoor Mobility

Indoor mobility should be assessed in the environments that the person uses. For example, a person with progressive ataxia may walk well at home using walls and rails but be unable to walk independently in a hospital, work or community setting. Mobility should be assessed jointly with the physiotherapist. Occupational therapist should be considering the interaction of the person with their environment and the tasks that the person wants to perform once they have walked/mobilized somewhere. Consider what and how the person plans to carry or transport items while walking. Occupational therapist should consider the use of walking aids in the home and other environments. Use of walking frames and parallel bars may need to be reconsidered in very small areas. A combination of devices may need to be considered to move from one area to another, eg a walking frame to the toilet and then the use of hand rails inside the toilet.

When wheelchairs are required, one should consider the environment including door widths, issues of access, ramps and interaction with furniture/tables to ensure that the person can still access areas they want/need to.

Major home modifications may be required, if not in the first instance, as the condition progresses, and this should be considered earlier, rather than later, with sensitive respect given to the person's psychological adjustment process. Some people may choose what is considered an unconventional solution which helps them navigate their home such as crawling. A compromise between safety and risk management and patient choice may be required.

## Practical suggestions

- Trolleys may help to transport items, especially heavy items, drinks and food at work or in the home and should be discussed and assessed if thought beneficial
- One-handed trays such as the Handitray® can help transportation of items
- Advise removal of items such as loose electrical cables and scatter rugs that may present as risks to mobility in the home environment
- Good lighting will help optimize performance of tasks and ensure that potential hazards in the home are avoided

- **Falls management**

Falls may occur in any area that a person mobilizes. At home, the person should be taught fall recovery techniques and where appropriate, consider the use of community care alarms such as pendant alarms, and techniques to avoid further injury such as pressure sores while waiting for help to arrive.

Occupational therapist should consider joint assessment with or referral to a physiotherapist, and referral to a falls programme/ group locally. If there is a family member or carer involved, the occupational therapist and physiotherapist should consider the safety of the carer.

- **Outdoor and community mobility**

Mobilizing outdoors can often present particular difficulty for the person with ataxia, as it may be an unfamiliar environment. Educate the caregiver and the person with ataxia about resting regularly whilst walking outdoors. A wheelchair for outdoor use can help to reduce fatigue or maximize safety. Motorized wheelchair helps more in management of fatigue.

An assessment for optimal seating position can be useful even in the early stages and is essential in the later stages. Consideration should be given to stable cushions and back supports, as canvas backs/seats in standard wheelchairs do not encourage good posture which may impact on function. A compromise between optimizing function and providing adequate support is important.

## Practical suggestions

- Shop-mobility, taxi card schemes, mobility buses, dial a ride services can be helpful
- Public transport and rail providers offer subsidized fares and can provide a meet and greet service/ access assistance for customers
- Outdoors motorized scooters or wheelchairs can maximize independence

## • Handwriting

This can be an area of difficulty for someone with ataxia that is progressive. If the person is still at school or at university, it is important to work within the provisions of special educational supports such as those provided through support workers. Access to work (AtW) is done a full assessment. This may include an Ability-net assessment for suggestions of alternative technological solutions for handwriting problems. Activity analysis may reveal the need for adaptations such as alternative positioning and/or seating, desks and different pens.





Fig 53:Fine motor activities & writing

**Practical suggestions**

- Ensure the work space and seating are set to maximize support and optimize posture advisable to avoid carrying items while walking
- Use of specialized grippers, weighted pens, weighted wrist cuffs and thick barreled pens may be of help.
- Voice-activated computer or Dictaphones software can be used to compensate for problems with handwriting discussed and assessed if thought beneficial
- Consider the type of pen nib and the pressure applied as some people experience fatigue affecting sustained pen grip

## • Computer use

There are many aids to compensate for ataxia when using a computer. Again, this assessment may require joint assessment with a speech and language therapist in considering whether voice-activated software may be appropriate for overcoming problems with using a keyboard to enter information; however, dysarthria may prevent voice-activated software from being useful. Importantly, Information Technology is a constantly changing area with new devices and solutions becoming available all the time. If the person is still at work, funding for this assessment should be gained thorough AtW referral. Alternatively, Occupational therapist should investigate charitable organizations that may provide funding to access these services and equipment.

### Practical suggestions

- A referral to IT solutions experts (such as Ability-net) is strongly advised
- Free advice is available on Ability-net website about IT adaptations for people with ataxia (www.abilitynet.org.uk)
- Keyboard and mouse modifications can be made to adjust the sensitivity and speed of response
- Alternative mouse such as a Tracker ball can be helpful

- Smaller keyboards or keyguards may help
- The layout and location of equipment should be considered for ease of access

## • Work

Occupational therapist should consider maintenance of the working role for as long as the person wants it to continue and for as long as that is possible.

It is important to provide education on their rights and responsibilities under the 1995 & 2005 Disability Discrimination Act. Support the individual regarding the disclosure of their diagnosis to others and their employers.It should be remembered that the intervention should allow them to develop skills to manage the employer themselves where possible. Where required a work site visit or referral to AtW will ensure a work site visit is undertaken if appropriate.

Occupational therapist may directly intervene by assessing and advising on reasonable adjustments which may include changing work hours, environmental adjustments or assistance with specific tasks or travel which may be supported financially by AtW (for information on AtW see www.jobcentreplus.gov.uk). AtW will help considerably with costs of other aids required such as motorized wheelchairs, ergonomically appropriate seating and desks, and IT devices. When work is no longer possible consider rebalancing the loss of the working role with other activities or help to access relevant benefits.

## • Driving

If they are a driver, the newly diagnosed person with progressive ataxia is legally obliged to inform the DVLA and their insurance company of their diagnosis as soon as it is confirmed. Reporting the diagnosis does not mean cessation of driving. The DVLA will request information from the person and their medical team and may request attendance at a driving assessment centre before making a decision.

Some people will require driving adaptations to allow safe driving to be completed (see www.direct.gov.uk/en/Disabled People / Motoring And Transport). Specialist centres provide assessment for suitable adaptations as well as driving ability (www.mobility-centres.org.uk). Where appropriate, the Motability scheme can assist people who receive the higher mobility rate of DLA with minor adjustments, lessons or funding a vehicle (www.motability.co.uk).

There are some cases when the condition causes such difficulty with driving that it is unsafe for the person to continue with this role. (www.direct.gov.uk/en/DisabledPeople/MotoringAndTransport). When this is true, Occupational therapist should explore alternative community mobility.

**Practical suggestions**

- Educate the person and caregiver on allowing the car door to be opened fully and to consider the height of the transfer being undertaken
- Ensure the person sits their bottom down first before moving their legs into the car
- Try inserting a swivel transfer mat and if the car seat is particularly low a firm foam cushion or blanket in a pillowcase
- Choose a model of car that optimizes transfers, door access and storage space

- **Leisure**

Occupational therapist should bear in mind that if there is loss of other occupational roles, leisure may be an area that can help to re-address this loss in a different capacity. For hobbies such as horticulture, there may be adaptations that can be made to maintain participation, or aspects of tasks can be performed such as visiting gardens locally, planning planting or weeding of raised beds.

Enjoying leisure time with one's family can be continued in modified ways, such as the use of accessible holiday homes, use of a wheelchair when visiting outdoor areas such as parks and galleries, and ensuring social contact continues in the home or other spaces. Due to visual problems and due to difficulty in holding a book, reading can be difficult.

## Practical suggestions

- Book stands can be obtained to hold the book
- Use of elastic bands around the loose pages of books can limit the frustration caused by rustling pages where tremor exists
- Use of page turners can be useful to help turn pages where fine motor coordination is a problem
- Books may be downloaded online and use of text enlarging buttons, magnifiers can help where vision is a problem
- Talking books, Text to speech are also available if preferred.
- Electronic page-turners can be purchased but are costly and take up space

- **Control of the indoor environment**

When communication difficulties present, ensure speech and language therapy involvement and assess jointly if available. Telephone use can present particular difficulty in pressing small buttons where tremor and reduced coordination present. Use of electrical equipment may be difficult if handling of small switches is necessary. As the condition progresses Occupational therapist should consider referral for regional environmental control unit assessment for optimizing control of the indoor environment.

Prioritizing activities for the person to participate in; especially in the palliative stage the focus on meaningful activity can provide immense satisfaction and comfort to the person with ataxia and the family.

## Practical suggestions

- Consider use of 'big buttons' telephones and phones with autodial numbers or voice activation
- Consider use of a telephone with two-way record to save conversation for replay later to help keep messages
- Telephone providers such as Bluetooth have an inclusion phone services policy including a communication solution guide obtainable online
- Light switches should be simple and easily reached from a standing or wheelchair position suitable to the person
- Light sensors can be used, which lit automatically when entered in the room
- To avoid bending, squatting and complex bending, appliance sockets should be located off the ground.

# Chp. 12 Multiple System Atrophy

**Dr. Joji Joseph, Dr. Rohit Das,**
**Dr. Rabab Kothari, Dr. Snehal Sontate**

## INTRODUCTION:

It is defined as an adult-onset, sporadic, rapidly progressive, multisystem, neurodegenerative fatal disease of unknown cause characterized by varying severity of parkinsonian features, cerebellar, autonomic and urogenital dysfunction ; corticospinal disorders. It can affect the autonomic nervous system causing autonomic failure, basal ganglia causing parkinsonism and / or cerebellum causing ataxia in any combination.

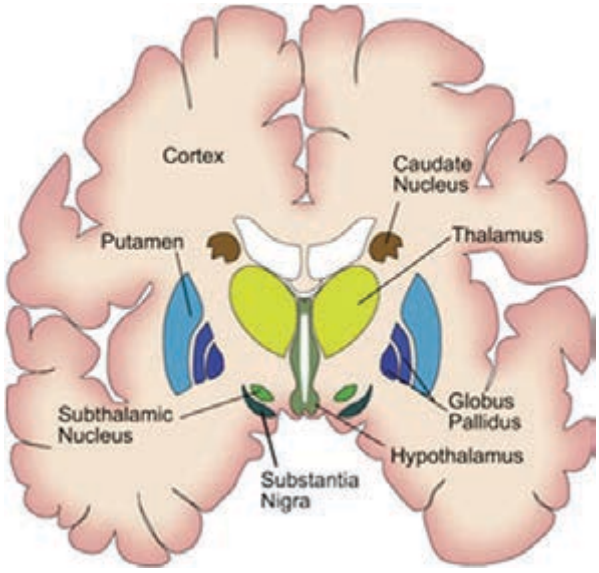



Figure 1

Multiple system atrophy, or MSA, is a rare neurological disorder that impairs your body's involuntary functions, including:

- heart rate
- digestion
- bladder function
- blood pressure

## Pathophysiology

Neuropathological hallmarks of MSA is cell loss in the striatonigral and olivopontocerebellar structures of the brain and spinal cord accompanied by profuse, distinctive glia cytoplasmic inclusions (GCIs) formed by fibrillized alpha-synuclein proteins.

## MSA: Types and symptoms.

Two forms of MSA, are recognized, according to whether Parkinsonian or cerebellar symptoms. Both types involve autonomic dysfunction, or dysautonomia. Autonomic symptoms usually appear first, though either motor symptoms or autonomic symptoms may appear first in the Parkinsonian type.

**MSA-P** produces Parkinson-like symptoms, including a slow, shuffling gait, rigid muscles, slurred speech and lack of facial expression. Patients with MSA-P may also develop a form of tremor known as resting tremor, which occurs when they are still and disappears when they are moving.

In the early stages, this form of MSA may respond to medications used for Parkinson's disease. However, MSA-P tends to progress more rapidly than Parkinsonism and as patients pass through the initial stages they no longer respond to Parkinson's medications. Individual variation with regard to the course of

the disease varies widely. Some MSA-P patients require assistance with daily activities as early as 5 years after diagnosis and others are able to remain independent for much longer.

**MSA-C** is characterized by progressive loss of coordination and balance; functions controlled by the area of the brain known as the cerebellum. MSA-C patients may experience a form of tremor known as action tremor, which occurs when they reach for an object. Muscle weakness associated with MSA-C can lead to slurred speech and problems swallowing. This form of MSA can appear as early as the 40's or not until the 60's1.

Dysautonomia in MSA leads to problems regulating heart rate, blood pressure, breathing, digestion and other internal organ functions. Patients may become dizzy or faint when they sit up or stand up; a condition known as neurogenic orthostatic hypotension. Conversely, blood pressure may be excessively high when lying down. Loss of bladder or bowel control, abnormal sweating, sexual impotence in men and sleep disturbances, including sleep apnea, and flailing movements during sleep also occur.

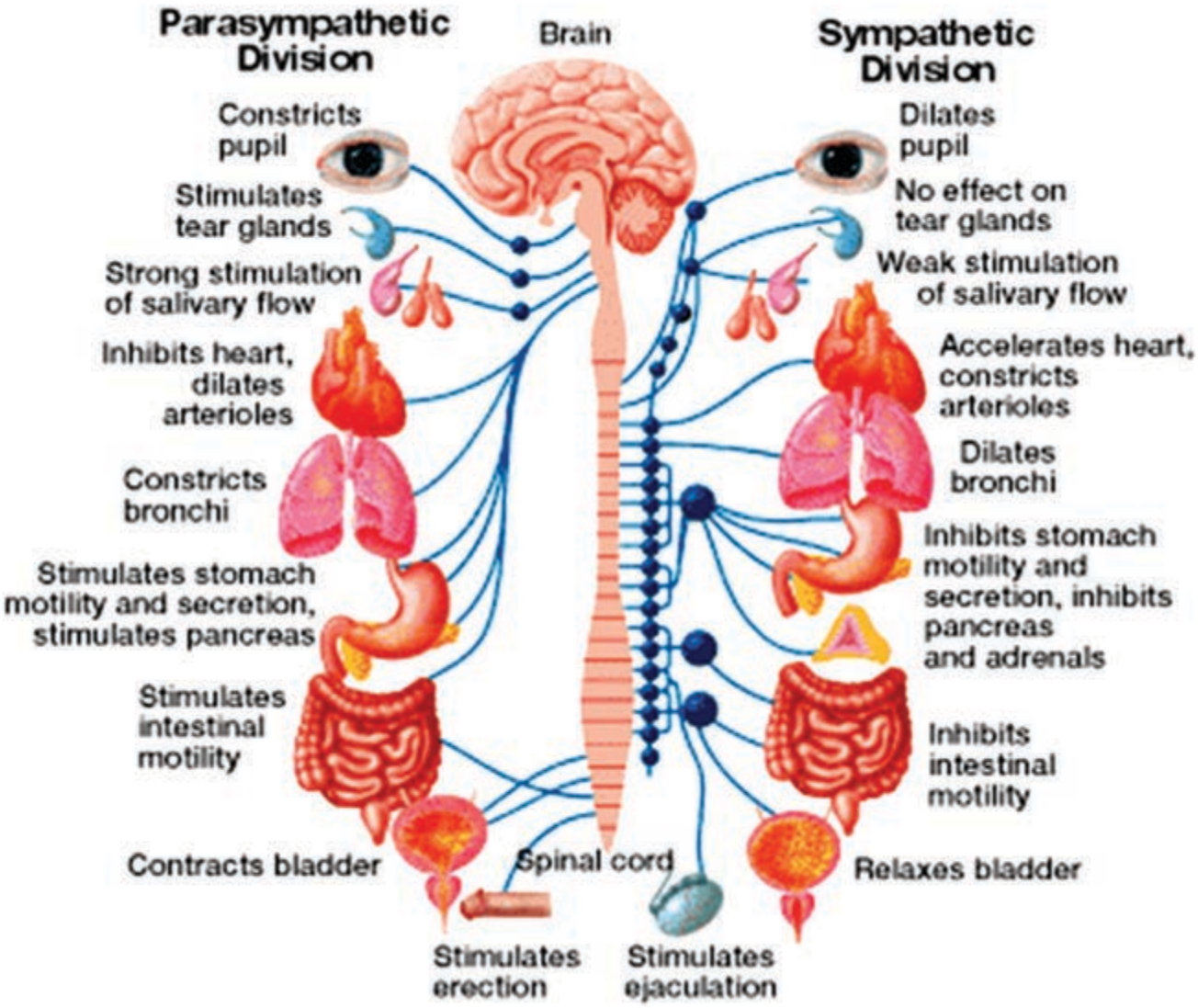



Figure 2

## Diagnostic criteria

Diagnosis of MSA can be challenging because there is no test that can definitively make or confirm the diagnosis in a living patient. A definite diagnosis of MSA can only be made upon autopsy where degenerative changes characterized by cell loss, gliosis, and the presence of cytoplasmic inclusions in the oligodendrocytes are apparent on tissue examination.

Some atrophic changes are visible on MRI. MRI is most useful in detecting MSA-C, as atrophic changes will occur in the cerebellum and pontine regions. However, due to varying data, current research is unsure of this method's ability to differentially diagnose MSA from other parkinsonian disorders.

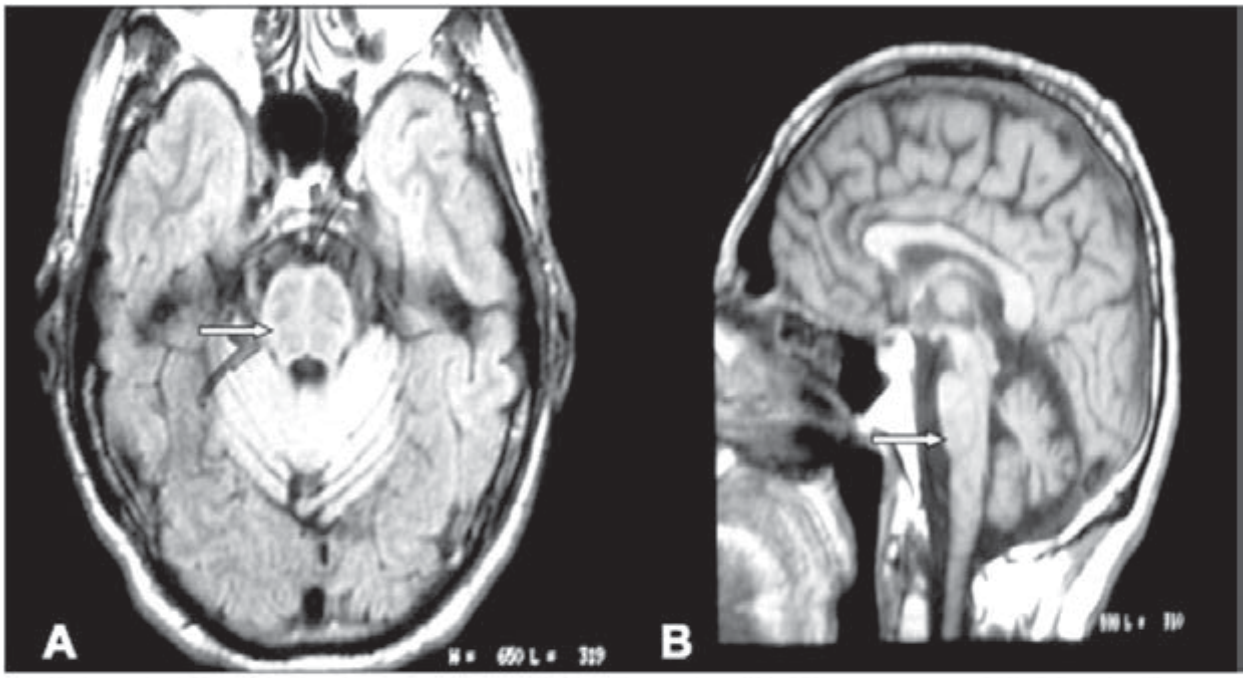



*Fig 1. Case 1. MRI: (A) Pontine atrophy with the "cross sign" in T1 image (arrow). (B) Cerebellar and brainstem atrophy in T1 image (arrow).*

**Figure 3**: MRI showing visible atrophy of the pontine (A) and cerebellar (B) regions.

While MRI and PET scans are of some use, particularly in ruling out other conditions, there is no single clinical feature or investigation that confirms MSA.

Table I defines the diagnostic categories of MSA, which are possible, probable, and definite. Table II displays what clinical features and criterion are required for the diagnosis of each clinical domain. Table III includes exclusion criterion for each clinical domain.

## Table I. Diagnostic categories of MSA

| | |
|---|---|
| **I. Possible MSA** | One criterion plus two features from separate other domains. When the criterion is parkinsonism, a poor levodopa response qualifies as one feature (hence only one additional feature is required) |
| **II. Probable MSA** | Criterion for: autonomic failure/urinary dysfunction plus poorly levodopa-responsive parkinsonism or cerebellar dysfunction |
| **III. Definite MSA** | Pathologically confirmed by the presence of a high density of glial cytoplasmic inclusions in association with a combination of degenerative changes in the nigrostriatal and olivopontocerebellar pathways |

## Table II. Clinical domains, features and criteria used in the diagnosis of MSA.

| Clinical Domain | Feature (Characteristic of the Disease) | Criterion (Defining Feature) |
|---|---|---|
| Autonomic Dysfunction | • Orthostatic hypotension with blood pressure falling by 20 mm Hg systolic and 10 mm Hg diastolic within 3 min of standing<br>• Urinary incontinence or incomplete bladder emptying | Orthostatic hypotension with blood pressure falling by 30 mm Hg systolic and 15 mm Hg diastolic within 3 min of standing and/or urinary incontinence as persistent, involuntary, partial or total bladder emptying, accompanied by erectile dysfunction in men |
| Parkinsonism | • Bradykinesia - Slowness of voluntary movement with progressive reduction in speed and amplitude during repetitive actions<br>• Rigidity<br>• Postural instability not caused by primary visual, vestibular, cerebellar, or proprioceptive dysfunction<br>• Tremor - Postural, resting, or both | Bradykinesia plus at least 1 parkinsonian feature |
| Cerebellar Dysfunction | • Gait ataxia (wide-based stance with steps of irregular length and direction<br>• Ataxic dysarthria<br>• Limb ataxia<br>• Sustained gaze-evoked nystagmus | Gait ataxia plus at least 1 cerebellar feature |
| Corticospinal Tract Dysfunction | • Extensor plantar response with hyperreflexia (pyramidal sign) | Not used as criterion in defining diagnosis of MSA |

## Table III. Exclusion criteria for the diagnosis of MSA

| Procedure | Findings |
|---|---|
| History Taking | • Symptomatic onset at <30 years<br>• Family history of similar disorder<br>• Systemic diseases or other identifiable causes for features listed in Table II<br>• Hallucinations unrelated to medication |
| Physical Examination | • Prominent slowing of vertical saccades or vertical supranuclear gaze palsy<br>• Evidence of focal cortical dysfunction such as aphasia, alien limb syndrome, and parietal dysfunction |
| Laboratory Study | • Metabolic, molecular genetic, and imaging evidence of alternative cause of features listed in Table |

## Complications :

This may include a gradual loss of the ability to walk , gradual loss of the ability to perform Activities of daily living. Injuries from falls etc. MSA can lead to long-term complications such as : dementia ,Confusion, Depression , sleep apnea etc.

## Prognosis :

MSA usually progresses more quickly than Parkinson's disease. There is no remission from the disease. The average remaining lifespan after the onset of symptoms in patients with MSA is 7.9 years. Almost 80% of patients are disabled within five years of onset of the motor symptoms, and only 20% survive past 12 years. Rate of progression differs in every case and speed of decline may vary widely in individual patients.

O'Sullivan and colleagues (2008) identified early autonomic dysfunction to be the most important early clinical prognostic feature regarding survival in MSA. Patients with concomitant motor and autonomic dysfunction within three years of symptom onset had a shorter survival duration, in addition to becoming wheelchair dependent and bed-ridden at an earlier stage than those who developed these symptoms after three years from symptom onset. Their study also showed that when patients with early autonomic dysfunction develop frequent falling, or wheelchair dependence, or severe dysphagia, or require residential care, there is a shorter interval from this point to death.

**Medical Management of Multisystematic atrophy**

There is no known cure for MSA and management is primarily supportive.

Ongoing care from a neurologist specializing in «movement disorders» is recommended as the complex symptoms of MSA are often not familiar to less-specialized health care professionals.

One particularly serious problem, the drop in blood pressure upon standing up (with risk of fainting and thus injury from falling) often responds to fludrocortisone, a synthetic mineralocorticoid..

Another common drug treatment is midodrine (an alpha-agonist).

Non-drug treatments include "head-up tilt" (elevating the head of the whole bed by about 10 degrees), salt tablets or increasing salt in the diet, generous intake of fluids, and pressure (elastic) stockings. Avoidance of triggers of low blood pressure (such as hot weather, alcohol, and dehydration) are crucial.

homecare services can be very useful as disability progresses.

Levodopa (L-Dopa), a drug used in the treatment of Parkinson›s disease, improves parkinsonian symptoms in a small percentage of MSA patients. A recent trial reported that only 1.5% of MSA patients experienced a less than 50% improvement when taking levodopa, and even this was a transient effect lasting less than one year. Poor response to L-Dopa has been suggested as a possible element in the differential diagnosis of MSA from Parkinson›s disease.

**Rehabilitation:** Management by rehabilitation professionals (physiatrists, physiotherapists, occupational therapists, speech therapists, and others) for problems with walking/movement, daily tasks, and speech problems is essential.

Physiotherapy can help to maintain the patient's mobility and will help to prevent contractures. Instructing patients in gait training will help to improve their mobility and decrease their risk of falls. A physiotherapist may also prescribe mobility aids such as a cane or a walker to increase the patient's safety.Other ways a physiotherapist can help to improve the patient's safety are to teach them to move and transfer from sitting to standing slowly to decrease risk of falls and limit the effect of postural hypotension. Instruction in ankle pumping helps to return blood in the legs to the systemic circulation.To further control the postural hypotension, raising the head of the bed by 8 in (20.3 cm) while sleeping may be indicated as well as the use of elastic compression garments.

Speech and language therapists may assist in assessing, treating and supporting speech (dysarthria) and swallowing difficulties (dysphagia). Early intervention of swallowing difficulties is particularly useful to allow for discussion around tube feeding further in the disease progression.{doubtful - citation needed} At some point in the progression of the disease, fluid and food modification may be suggested. Speech changes mean that alternative communication may be needed, for example communication aids or word charts.

Social workers and occupational therapists can also help with coping with disability through the provision of equipment and home adaptations, services for caregivers and access to healthcare services, both for the person with MSA as well as family caregivers.

## Physiotherapy management in multisystem atrophy

### MULTIPLE SYSTEM ATROPHY (MSA)

- Multiple System Atrophy (MSA) is characterized by Parkinson's, Cerebellar and autonomic features.
- There are mainly two types of MSA-
  - Predominant Parkinsonian (MSA-P)
  - Cerebellar (MSA-C)
- Pathology- Degeneration of SNPc, cerebellum, and inferior olivary nucleus coupled with glial cytoplasmic inclusion (GCI).
- Clinical features-
  - Patient presents with Parkinsonism features in conjunction with cerebellar signs and/or early and prominent autonomic dysfunction; usually orthostatic hypotension.
- Investigations-
  - It can show pathologic iron accumulation in the striatum on T2 weighted scans.
  - High signal changes in the region of the external surface of putamen (putaminal rim) in MSA-P.

### PHYSIOTHERAPY MANAGEMENT

- Even though pharmacotherapy is the primary management, the role of exercise and physiotherapy should be emphasized at every stage of the disease.
- Rehabilitation results are better when treatment is started early in the disease process.
- Physiotherapy management in Multi-System-Atrophy (MSA) is based on the

clinical features. Kindly refer chapters of Physiotherapy management in Stroke, Cerebellar ataxia and Parkinson disease

**OCCUPATIONAL THERAPY ROLE IN MSA.**

MSA is a rare progressive neurological disorder that affects adult men and women and leads to premature death. Currently, there is no known cause or cure. MSA causes degeneration or atrophy of nerve cells in several (or multiple) areas of the brain which results in problems with movement, balance and autonomic functions of the body such as swallowing, bowel, bladder and blood pressure control.

Occupational Therapy helps individuals recovering from injury/illness or with disabilities participate as much as possible in everyday activities

They train and/or educate individuals to use adaptive equipment and techniques to improve functional abilities.

They develop UE strength and coordination and provide splints accordingly.

They assist family members with effectively supporting and/or care for their loved ones.

The goals should be according to the need of the patient and caregivers.

The therapist should help the patient by providing external resources and safety measures.

Occupational therapy adds value to the lives of people with MSA by improving self-perceived performance in daily activities. Occupational therapy aims to make day to day life easier by increasing knowledge around symptom management and by supporting decision around the choice and timing of equipment, home adaptations and care until the end of life.

The nature of MSA and its relentless course means occupational therapy intervention needs to be placed within a client centered framework. Although techniques for symptom control in other progressive neurological disorders may prove useful such techniques should always be used within a client centered context, to avoid treating the symptoms rather than the person.

The challenges of coping with a complex, and at times, unpredictable movement disorder means that a person's skills and abilities can vary from day to day. Additionally, as some people have been misdiagnosed for years prior to an accurate diagnosis of MSA, each person's understanding of their disease course and resulting stage of emotional acceptance and adjustment of loss of role and function can vary tremendously.

Given the rarity of MSA and a sometimes fraught journey to accurate diagnosis and a rapidly declining disease course, occupational therapists tend to see people with MSA long after their symptoms have begun.

It is therefore essential that any prior assumptions are set to one side and the process of identifying and meeting unique occupational performance needs is led by the person with MSA and their family

**Access to other services**

During the assessment phase, occupational therapists need to be thinking about

accessing the right people, with the right skills and actively involving health and social care teams at the right time. Joint sessions and shared goals with physiotherapy, speech therapy, dietitians and specialist nursing colleagues are essential to meet a person's needs in the face of rapid functional decline.

Occupational therapists are central to providing information about the role and contribution of other team members and where possible, should do this before they are needed to ensure enough time is given for the person with MSA and their family to understand and accept support from other services. Occupational therapists should also be mindful of identifying and meeting the needs of carers through accessing a full carer assessment and facilitating provision of both physical and psychological support including counselling as required.

When accessing other services, it is essential that this is done swiftly, with information regarding MSA and its prognosis is emphasized. Long waiting lists are frequently encountered when requesting for equipment such as a wheelchair or home adaptations, and people with MSA must be recognized as top priority to avoid any delay.

## Treatment Phase:

Regardless of the stage in the MSA disease, occupational therapy treatment should aim to increase satisfaction in daily tasks.

Skill level intervention to enhance performance. Skill level intervention looks at how the person is able to perform the task along with interventions which aims at reducing the physical or cognitive effort used to enhance the skills.

**Skill level treatment includes:**

| Technique | Example |
|---|---|
| **Hands-on help** | Teaching the carer to physically guide weight transference techniques during sit-to-stand to gain 'nose over toes' |
| **Breaking the task down into smaller steps** | Smaller steps of the task are practiced to slow the task down, with physical help, verbal prompting or equipment added in to reduce task effort |
| **Optimising the environment** | This includes the addition of lighting in dark or troublesome areas of the house<br><br>Securing loose floor surfaces and providing grab rails<br><br>Using hazard tape to draw cognitive attention to the need to change direction or walking speed at door thresholds |
| **External and self cueings** | Cueing technique, such as extrinsic visual cue cards such as 'feet first' and intrinsic such as 'large steps' [16], may be useful for people with MSA-P to gain more control over the flow of movement. |

## Knowledge level intervention to support performance

Supporting performance by increasing knowledge is a powerful intervention. Whilst not aiming to change how the task is done, education and information can help the person understand why the task is broken down so that s/he can make informed choices regarding options to support their performance

**Knowledge level treatment includes**

| Technique | Example |
|---|---|
| **Education about symptom management** | Providing details about how MSA progresses<br>Providing guidance about the use and timing of medication<br>Keeping a fatigue diary and understanding how to proactively manage fatigue |
| **Information about equipment options** | Providing information regarding the choice and use of small pieces of equipment<br>Discussing how to get the process of major adaptations at home underway before the disease progresses<br>Knowing how to hire a shop-mobility scooter whilst shopping |
| **Providing help to stay in work** | Discussing legal rights and available support to stay in work |
| **Providing information about other services** | Introducing other services and the role they play including help at home from carers, support for families, palliative care services, respite and hospice care |

**Attitude level intervention to change performance**

Attitude level intervention is possible when the person with MSA understands the level of emotional and practical acceptance of their disease. This includes knowing what has changed in the body and understanding how this impacts on the quality of performance, combined with correct information to make appropriate choices.

Helping support attitudinal level change requires the occupational therapist to work along with the person with MSA and their family towards a different set of expectations. This may include discussions on the nature of a task. Modifications required to achieve the same goal with less effort.

**Attitude level treatment includes**

| Technique | Example |
|---|---|
| **Changing the way the task is performed** | Buying pre-prepared meals to avoid cooking from scratch Using a scooter to mobilise safely outdoors |
| **Removing the need to do a task** | Ordering shopping on-line and having it delivered Paying for a gardener or cleaner Deciding to accept medical retirement |
| **Accepting more care and support** | Accepting the need for care and equipment at home Understanding when residential or hospice care might be the safest place to live |

**Case studies:**

These case examples aim to illustrate the type of goals people with MSA might articulate and how occupational therapist might plan their treatment, including the role and function of other team members.

**Bob: a 66 year old man with a diagnosis of MSA-P**

| **Goal: to reduce effort and increase safety with bath transfers** | | | | |
|---|---|---|---|---|
| **Activity analysis** | **Skills** | **Knowledge** | **Attitude** | **Referrals made** |
| Problems due to - increasing stiffness and rigidity-reduced balance reactions - increasing episode of 'freezing' leading to - anticipatory anxiety when bathing | Practicing - a step by step way of getting into and out of the bottom of the bath - using visual prompt cards - and verbal cueing from wife to guide movement | Providing information regarding-the need for grab rails by the bath-use of a swivel bather or bath lift-installation of a level access shower -the need to have the bathroom door open outwards in the event of a fall | Discussing options regarding-providing paid for care to assist with morning routine - anticipatory discussion regarding moving tosingle storey living | Physiotherapist for stiffness and rigidity<br>Social Services OT for equipment provision<br>Social Work Team for future care provision |

**Barbara: a 49 year old lady with MSA-C**

| Goal: to reduce fatigue and increase satisfaction with daily energy levels. | | | | |
|---|---|---|---|---|
| **Activity analysis** | **Skills** | **Knowledge** | **Attitude** | **Referrals made** |
| Problems due to - trunkal instability - increasingly unsteady gait - reduced coordination of finer skilled movements | Practising - reducing effort during personal care and domestic routines by sitting when possible to undertake tasks - reducing unwanted excursion of hands by elbow propping during finer skilled tasks | Providing information regarding - fatigue management techniques including the need for proactive rest - using a motorised scooter to get to work rather than struggling to walk outdoors | Discussing options regarding - applying for any benefit entitlement to help pay for domestic support at home or taxis - raising the option of working from home with employer | Physiotherapist for ataxia and core stability<br><br>Disability Employment Advisor for support via the 'Access to Work' scheme<br><br>Citizens Advice Bureau possible regarding benefit entitlement |

Occupational therapy may improve functional abilities in patients with mild to moderate MSA and it depends on symptoms and has a symptomatic approach.

## Signs and Symptoms

- Rigidity
- Action tremor, i.e., irregular, jerky, myoclonic
- Bradykinesia
- Freezing of gait/gait and limb ataxia
- Early postural instability and falls
- Orthostatic hypotension
- Erectile dysfunction
- Bladder dysfunction/constipation
- Speech and swallow difficulties; mixed dysarthria tends to emerge earlier in MSA than PD, is more severe, and deteriorates more rapidly
- REM Behavioral Disorder (RBD)—acting out dreams while sleeping due to lack of atonia
- Nystagmus and jerky pursuit
- Antecollis
- Difficulty with thermoregulation
- Cognitive impairment is typically mild

## Treatment Strategies

- No medications to slow disease progression
- Patient and caregiver education about disease

For bradykinesia and rigidity:

- Levodopa and dopamine agonists may help initially
- Avoid hand flexion/resistive exercise (e.g., Thera Putty®/foam squeezes).
- Encourage hand/wrist extension instead

## For orthostatic hypotension:

- Fludrocortisone, midodrine
- Increase salt in diet
- Rising slowly and resting after position changes
- Support stockings
- Elevate head of bed

## THE ROLE OF OCCUPATIONAL THERAPISTS

- Smaller, more frequent meals
- Avoid increased time in motionless positions
- Avoid warmer temperatures
- Avoid valsalva-provoking maneuvers

Compensatory strategies for self-feeding skills with tremor:

- Use of adaptive devices (adaptive utensils and plate guards)



Figure 4: Use of universal cuff for various everyday tasks.

- Nonskid mat to prevent the plate/ bowl from sliding
- Arm position—anchor elbow on table, then lower arm down to plate/bowl to retrieve food and then back up to mouth (using the arm as a lever, never taking the elbow up/off of the table)



Figure 5: Scoop spoons

- Use deep spoons to assist with scooping
- Elevate the height of the dish bowl

Functional mobility in the home bedroom:

- Use a bedrail to assist with difficulty rolling
- Keep floors clutter free and walkways well lit
- Wear warmer pajamas and use fewer blankets to decrease likelihood of getting tangled up in bed



Figure 6: Buttoning Device



Figure 7: Universal cuff for fork

- Use a satin pillowcase/sheet under the bottom of the patient to assist with improving bed mobility
- Always sit as much as possible when dressing to help decrease risk of falls (hypotension)
- Always dress the most affected limb first



Figure 8: Sock aid



Figure 9: Cup to avoid spillage Figure 10



Figure 11



Figure 12



Figure 13

Figure 11, 12 and 13: Angled devices for ease of daily tasks



Figure 14: Enlarged spoon, knife and fork

Functional mobility in the home bathroom:



Figure 15 and 16 : Grab bars alongside toilet seat



Figure 16



Figure 17: Grab bars in bathroom

- Grab bar/hand-held showerhead installation
- Remove glass partitions in the shower/ tub to make transfers in/out easier and safer (sit and swing technique vs. step in)
- Use a shower bench/tub chair with a back to decrease risk of slipping, promote good posture, and conserve energy
- Always sit as much as possible when bathing; eliminate all turning in the shower
- Avoid extremely hot showers to decrease dizziness/lightheadedness

**Functional mobility in the home kitchen:**

- Never overreach; get as close as you can to the object that you are reaching for (e.g., in cabinets)
- Always support yourself with one hand when reaching (e.g., countertop)
- Always stand to the side of the dishwasher/oven/refrigerator when opening



Figure 18: Long handle scrubber



Figure 19: Shoe aid



Figure 20: Enlarged knob



Figure 21



Figure 22

Figure 21 and 22: Reachers



Figure 23: Table top height according to wheelchair



Figure 24: Adapted keys

- Make U-turns in open spaces around the house (swivel-wheeled rollators with brakes work well)
- Avoid pivot turns or crossing one foot over the other to turn; instead move right foot first to turn right, left to turn left
- Use a wide staggered stance when performing activities (this helps to reduce LOB and retropulsion)

- Side step when working at the counter-space or sink



Figure 25:



Figure 26



Figure 27



Figure 28

Figure 25, 26, 27 and 28: various types of ramps

## Other considerations for home safety

- Installations of ramps
- Decluttering living space
- Widening doorways
- Installing grab bars
- Add lighting and use night lights
- Make lists, use calenders
- Use of alarms to remember medications
- Stair lifts and chair lifts



Figure 29



Figure 30

Figure 29 and 30: Stair and chair lifts

**Strengthening techniques:** MSA might cause following upper extremity deficits:

Slow movements, tremors/impaired coordination, decreased functional ability, impaired handfunctions, weakness/ stiffness and easy fatiguability, contractures or loss of ROM etc.

Active movements should be performed to maintain/improve strength and reduce stiffness. Regressive resistive exercises techniques are used to prevent muscle fatigue. Reduce prolonged static positioning especially in MSA-type P. Various weight bearing exercises to improve/maintain proximal stability/ mobility and trunk strength.

## SPLINTS:

Splints help to provide support to weak joints and relieve pain.

Splints can provide stretching to maintain / increase range of motion. (functional cockup splint , radial bar cockup etc)

Use of splints during night or day depending on purpose.

They can provide support to weak wrist muscles and encourage functional use of hands.




Figure 31



Figure 32



Figure 33

Figure 31, 32 and 33: various hand splints

## Energy conservation and work simplification techniques:

- Use of these techniques will help maintain energy throughout the day.
- Enable to perform the ask with less fatigue.
- Help maintain muscle strength for task performance
- Avoid rushing
- Take appropriate rest intervals
- Plan accordingly
- Organize all necessary equipments at one place
- Sit rather than stand, when possible
- Use good posture and proper body mechanics
- Use strongest joint available for specific task.

## End of Life Care: The occupational therapist's role

Every occupational therapist has an active role to play in preparing the person with MSA and their family for functional decline towards death. Significant and life changing decisions need to be raised and discussed as early as acceptable, so people have time to understand and accept change and decline over time. Major adaptions at home need to be considered and undertaken early and discussions around where the person wants to die raised and plans put in place.

Identifying and meeting the needs of carers is critical, with occupational therapists playing a central role in assessing carer burden with the aim of facilitating additional support to avoid carers reaching breaking point at home. Occupational therapists should discuss accessing family counselling for spouse or partner and children, and keep a frequent dialogue open about the need for formal care, including options for respite, residential care and hospice support. Alongside members of the wider team including equipment providers, MSA specialist nurses, district nursing and palliative care colleagues, occupational therapists should guide decision making with realistic and feasible options to maintain goals where possible, relieve suffering and optimize effective end of life care.

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https://www.healthline.com/health/multiple-system-atrophy

# Chp. 13 Cerebral Palsy

**Dr. Khushboo Bhagwanani, Dr. Jidnyasa Koli, Dr. Myola D'sa, Ms. Nikita Khokhani, Mr. Vishal Ganar, Dr. Abhishek Gupta**

Cerebral palsy (CP) is a non-progressive disorder of movement and posture due to defect or lesion or immature brain. The main motor function problems of children with CP are the delay or arrest of motor development, slow gross and fine motor functioning skills, an abnormal movement pattern and poor endurance for functioning. The motor disorders are often accompanied with poor cognition, sensory system and perception which further hamper their motor functioning.

## History:

Dr. Little theorized that lack of oxygen damaged the sensitive brain tissues, which controls movement. Sigmund Freud disputed Dr. Little's conclusions. He pointed out that most CP children suffer from additional problems like seizures, mental retardation and visual disturbances. This led him to believe that CP might because during the brain's development in the womb many months before birth. However doctors only seriously began investigating CP in 1861 when Dr. William John Little, published a paper describing children with spastic diplegia. Spastic diplegia is still sometimes called Little's Disease.

Mac kathie and Polani in 1959 explained CP as persisting but not changing disorder of movement and posture, appearing in early years of life and due to non-progressive disorder of brain. Later, Mutch et al suggested that it is an umbrella term covering a group of non-progressive but often changing, motor impairment syndromes secondary to lesions or anamolies of the brain arising in the early stages of development. in early year CP was treated mainly with surgical interventions but later by philips intervention it was believed that exercises are equally important for rmotor development. Since Phelp's era various non urgical approaches have evolved, emphasing Neuromuscular training. They include Neuro Muscular Reflex therapy (Fay & Doman), Neuro developmental treatment ( Bobath), Neuro physiological Approach (Rood), Proprioreceptive Neuro Muscular Facilitation (Kabat, knott & Voss) and Sensory Integration. Dr. Rosenbum et al, thus released a report in 2004 that it is a group of disorders of development of movement and posture, causing activity limitation, that are attributed to non progressive disturbances.

## Prevalence & Incidence:

Prevalence rates for CP vary in different countries and regions, based on their economic and nutritional factors and access to good obstetric care. Because of lack of tests for CP, the vagueness and elasticity of even the best definition, inaccurate ascertainment (many cases are never reported to physicians or public health authorities) and lack of agreement between different experts. The confusion results because many cases die in infancy and others may be so mild as to be unrecognized. Prevalence rate for CP presented as the prevalence at a givenageper 1000 live births of the defined population understudy.

Ofevery2000infantsborn,5arebornwith CP. This incidence had remained constant for over 30 years. Although improvement in medical care have decreased the incidence of CP among some children who otherwise would have developed the disorder. An estimate for India is around 45 per 1000, rather more in rural & less in urban populations.5 to 8 percent of premature infants under 1500 grams birth weight, who survive have CP. The marked difference in prevalence creates challenge for Indian Preventive medicine to make the best obstetric and perinatal care available to all communities. Essential keys to progress include improve deducation and nutrition to mother stobe.

## Etiology

### 1) Premature birth

Children born with pre-term delivery have greater risk to develop cerebral palsy, prevalence of cerebral palsy causing due to pre-term is reduced upto (35%) now. Mechanism of pre-term and post-term birth leading to cerebral palsy would be related to the physiological changes that triggered labour along with certain cerebral abnormalities occurring during the time of birth or later in time span. Periventricular leukomalacia (PVL) is a type of brain injury caused due to under developed infant leading to damage to white matter in the brain surrounding the ventricles which is the leading cause of CP seen in preterm infants. Intraventricular hemorrhage (IVH) is mainly associated with bleed in the ventricular system predominatly seen in premature infant caused due to fragile and under developed blood vessels in the infant's brain. IVH may cause PVL or ischemia in other parts of the brain.

| **Antenatal cause (70-80% of causes)** | **Peri-natal cause** | **Post-natal cause** |
|---|---|---|
| Prematurity and low birth weight<br>Infections<br>Multiple gestation<br>Previous maternal complications | Birth asphyxia<br>Quality of neonatal care<br>Complicated labour and mode of delivery<br>Birth weight and APGAR | Head trauma<br>Non-accidental injury<br>Meningitis/ Encephalitis |

**Table: Common antenatal, perinatal and pos tntal causes**

### 2) Infections

Infections can be acquired in utero or certain congenital infections in newborns leading to developmental anomalies, affecting growth and lead to significant neonatal morbidity and mortality. Fetoplacental and uterine infection or inflammation can also lead to CNS injury and CP caused during pre-natal period. Underdeveloped fetal brains are susceptible to inflammation and inflammatory cytokines which are responsible for PVL. Chorioamnionitis, an infection of the chorion and amnion, these are two membranes surrounding the developing fetus frequently associated with maternal infection in CP. TORCH infections comprises of toxoplasmosis, Treponema pallidum, rubella, cytomegalovirus, herpesvirus, hepatitis viruses, human immunodeficiency virus, and other infections, such as varicella, parvovirus B19 and these are associated with approximately 5% of all CP cases .

**3) Multiple gestation**

This increases the risk of antenatal complications, such as preterm labour, growth restriction, low birth weight, and death of a co-twin.Death of co-twin induce neuropathologic changes leading to CP in the surviving twin. Prevalence of CP in the surviving twin was found to be 15 times higher than average.

**Pathophysiology**

Its physiopathology is essential in developing some preventing and curative strategies. Interrupted oxygen supply to fetus or brain asphyxia are considered to be the main factor leading to CP. It is often the result of environmental factors, possibly interacting with genetic vulnerabilities, would be severe enough to cause the destructive injuries seen on standard imaging (i.e., ultra sonographic study or MRI), predominantly in white matter, gray matter and the brainstem nuclei in infants and newborns. They mainly have impact on immature brain and could alter the remarkable series of developmental events. Biochemical factors originate with cell process, loss or cellular death mainly observed in hypoxic-ischemic along with inflammatory conditions, such as excess production of proinflammatory cytokines, oxidative stress, maternal growth factor deprivation, extracellular matrix modifications, and excessive release of glutamate, triggering the excitotoxic cascade .

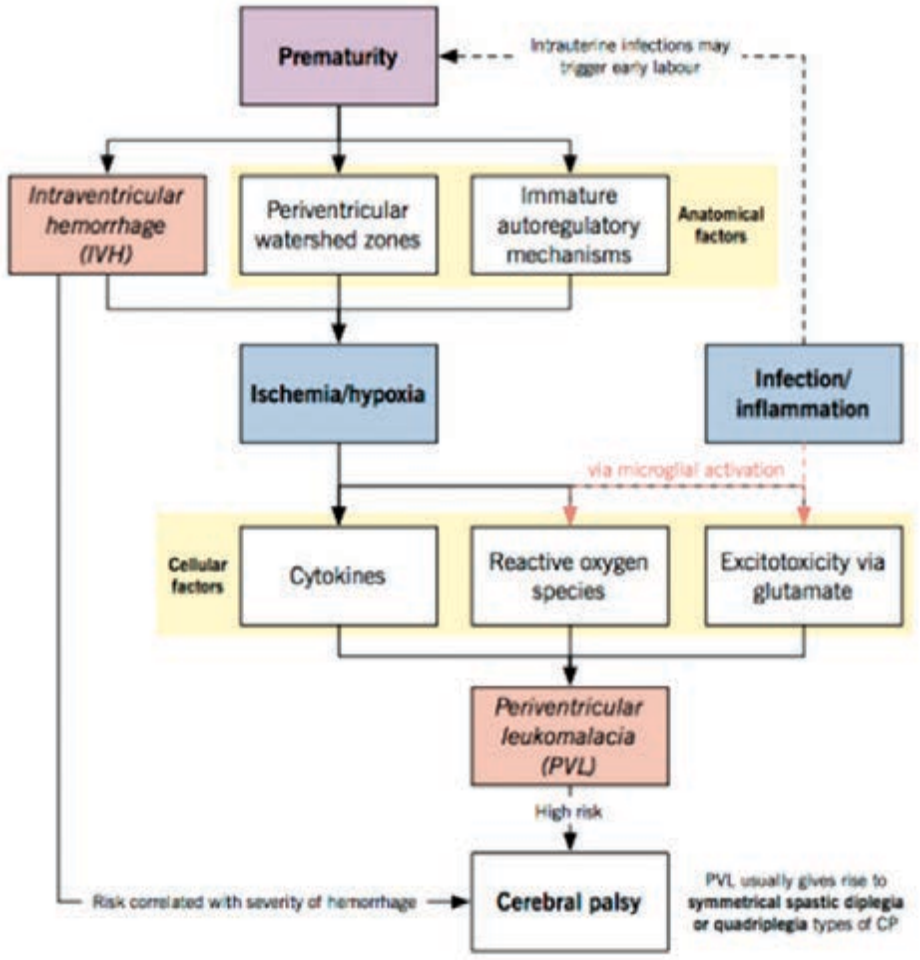



Fig 1 Pathophysiology of CP

**Ischemia/hypoxia**: The periventricular white matter of the neonatal brain is supplied by the distal segments of adjacent cerebral arteries. Although collateral blood flow from two arterial sources protects the area when one artery is blocked (e.g., thromboembolic stroke), this watershed zone is susceptible to damage from cerebral hypoperfusion (i.e., decreased cerebral blood flow in the brain overall). Since preterm and even term neonates have low cerebral blood flow, the periventricular white matter is susceptible to ischemic damage. Autoregulation of cerebral blood flow usually protects the fetal brain from hypoperfusion, however, it is limited in preterm infants due to immature vasoregulatory mechanisms and under development of arteriolar smooth muscles.

**Infection and inflammation**: The process involves microglial (brain macrophage) cell activation and cytokine release, leading to damage to their specific cell type in the developing brain called the oligodendrocyte. The oligodendrocytes are mainly brain cell that surrounds neurons to form the myelin sheath, essential for white matter development. An intrauterine infections is responsible to activate the fetal immune system producing cytokines (e.g., interferon γ and TNF-α) along with free radicals that are toxic to premyelinating oligodendrocytes. They have immature defences against reactive oxygen species (e.g., low production of *glutathione*, an important antioxidant). IVH is hypothesized to cause PVL because iron-rich blood causes iron-mediated conversion of hydrogen peroxide to hydroxyl radical, contributing to oxidative damage.

**Excitotoxicity** is a process where increased extracellular glutamate levels stimulate oligodendrocytes to increase calcium influx, which stimulates reactive oxidative species release. Glutamate is increased because hypoxia causes white matter cells to reduce reuptake of glutamate due to lack of energy to operate glutamate pumps. Glutamate is also released from microglial cells during the inflammatory response.

***Etiopathology:***

The under lying basis of sequelae in CP infant is white matter damage, collectively called periventricular Leukoence-phalopathy. This term encompasses germinal matrix haemorrhage (GMH), Periventricular

Haemorrhage (PVH), Intraventricular Heammorhage (IVH), Periventricular Heammorhagic Infarction, and Periventricular Leukomalacia (PVL) Preterm infants are more prone to GMH, PVH, and IVH, because theircerebral circulation is sensitive to changes of bold pressure (pressure passive) and they lack supporting glia in their germinal matrix.

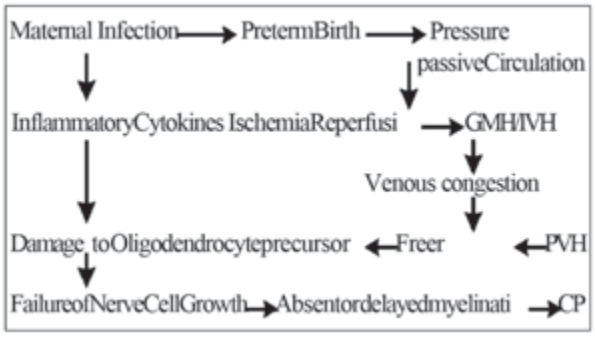



Fig 2. Events leading to CP in preterm Infant.

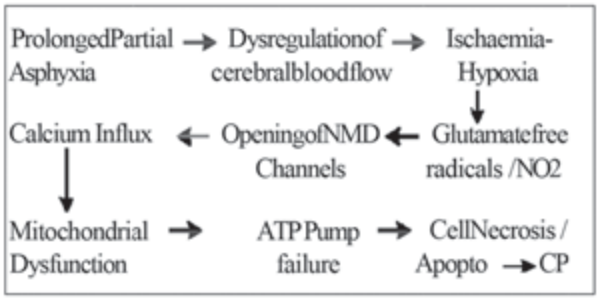



Fig 3.Events leading to Cerebral Palsy in Term Infant

***Classification of CP :***

The diversemotor and nonmotor associated defects in CP, would suggest the difficulties in categorizing this disorder. Therefore children with CP are categorized on the basis of the types of neuro motor deficit and topographic distribution of the deficit

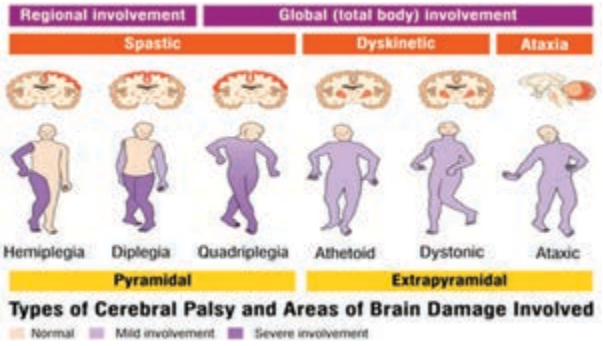



Fig. 4 Types of CP

*Neuromotor Classification by Minear (1956)*

1. **Spastic :** This type is characterized by exaggeration of the stretch reflex and increased deep tendon reflexes in the affected parts.
2. **Athetoid:** The chief characteristics of this type are the slow worm like, involuntary uncontrollable, unpredictable and purposeless motions atrest.
3. **Rigidity :** When the part is moved, there is a continuous resistance inagonistand antagonist muscles.

**Topograhic Distribution Of Neuromotor Involvement**

1. **Paraplegia:** involvement ofthe lower extremities. Patients with paraplegia are practically always of the spastictype.
2. **Hemiplegia:** involvement of an upper and lower extremity on the same side of the body. Patients with Hemiplegia are almost always spastic, but occasionally anathetoid hemiplegic patient may be seen.
3. **Triplegia:** involvement of three, extremities, usually both lower extremities and one arm. The disability is usually of spastic type.
4. **Quadriplegia or tetra plegia :** involvement of all four extremities. The term diplegia is some times used to indicate that the lower extremities are more involved than the upper. Almost all patients of athetosis have all four extremities involved.



Fig 5 Presentation of CP

**Classification based on Severity :**

This type of classification is emphasizes on the quantification of neuro-motor involvement that limits patient's ability to perform Activities of daily living.

1. Mild: The patient needs no treatment, since he has no secondary problems, is able to care for his daily needs and ambulates without the aid or any appliance.

2. Moderate: The patient need treatment, since he is inadequate in self care, ambulation, and had associated problems. Braces and self help appliances are needed.
3. Severe: The patient needs treatment, but the degree of involvement is so severe that the prognosis of self care, ambulation is poor. And also the associated secondary complications are major concern hampering patient's quality of life.




Fig 6. Classification based on severity

The following list presents the medical problems most often associated with CP.

*Neurological Problems:*

1. Mental Retardation.
2. Learning Disabilities.
3. Attention Deficit Hyperactivity Disorder/ Behavior alissues.
4. Seizure Disorder (epilepsy).
5. Visual Impairment.
6. Swallowing Difficulties.
7. Speech Impairment.

*Orthopaedic Problems:*

1. Scoliosis.
2. Hip Dislocation.
3. Contractures ofJoints.
4. Discrepancy in leglength.

**Secondary Effects:**

1. Communication Disorder.
2. Drooling.
3. Poor Nutrition/growth
4. Aspiration Pneumonia and gastro esophageal reflux.
5. Depression.
6. Fragile bones and frequent fractures.
7. Cavities-Tooth decay and gum disease.
8. Constipation or Bladder bowelincontinence.



Fig 7 Quadriplegic CP child with spasticity and poor voluntary control of limbs.

*Primitive Reflex Patterns:*

These are patterns seen in the early stages of development with disappear later on or with advanced age. These reflexes are essential for normal progressive motor development. However not overcoming these primitive reflex patterns at the right time should be definitely considered as abnormal. Initially lower centers such a s spinal cord control these movements but later on higher centres like mid brain and cortex take control over them and dominate the lower ones thus integrating them for various voluntary functional task.

Accordingly we have four levels at which reflexes are regulated:

1. Spinal cord: Flexor with drawal, Extensor thrust, Cross extension, Palmar grip, Plantar grip, Sucking reflex, Rooting reflex, Primitive walking.

2. Brainstem: Symmetrical Tonic neck reflex, Assymmetrical Tonic neck reflex, Tonic Labrinthine reflex, Positive and negative supporting reaction.

3. Midbrain: Optical Righting, Labr in thine reflex, Body on neck, Body on body, Neck righting reflex.

4. Cortical reflex: Equilibrium reactions, which can be in prone, supine, kneeling, sitting and standing. Equilibrium reaction in supine and in prone position in present from 6 months onwards, in quadriped position is present from 8 months, and in sitting starts normally from 10-12 months. Kneeling equilibrium occurs from 15 months onwards and finally in standing occurs from 18 months onwards.

5. Automatic reflexes: Moro's reflex, Gallants trunk incurvatum, Landau's reflexes and parachute reflex.

Following are the positive sign's usually observed in children with CP:

Abnormal primitive signs/ persistent perinatal reflexes/reactions which hampers the motor development and motor milestones of an infant. This results in inability to isolate the efforts to perform specific movements resulting in deficient postural mechanisms leading to sustained holding of certain postures difficult.






Fig 8. TLR and ATNR reflex

## Abnormal Manifestations and Prognosis

1. A child with spinal cord dominance will not be able to walk and carry out ADL and therefore he/ she will bebedridden throughout. This condition leads to a lot of contractures and thus our aim of treatment would be prevention to secondary complications due to lack of ambulation like preventing tightness or contractures, respiratory complications, etc.

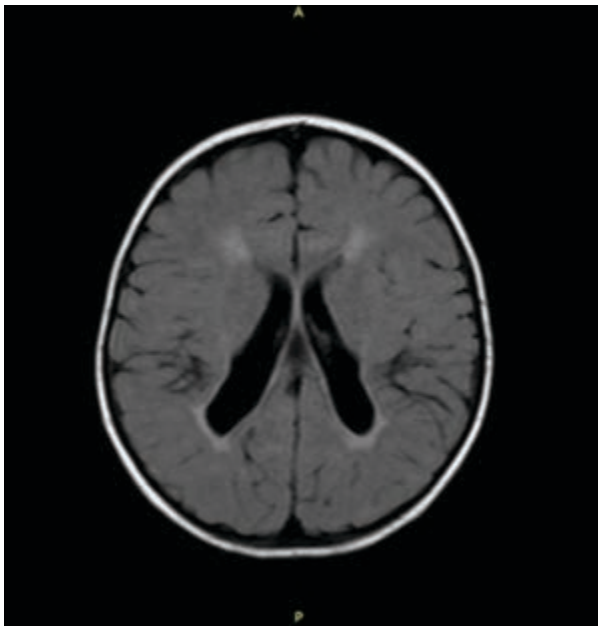
2. Child with brain stem dominance will also be bedridden and unable to walk. treatment would be aiming towards prevention of secondary complications due to lack of ambulation need to be prevented.

3. Midbrain dominance children will have a relatively better prognosis, such a child can ambulate or may have absent equilibrium reactions, somemay even require walkingaids.
4. Cortical Dominance children will have near normal development.

## NeuroImagining:

Doctors diagnose CP by evaluating a child's motor skills and taking a careful and thorough look at medical history. In addition to checking their signs and symptoms characteristic symptoms - slow development,abnormal muscle tone, unusual posture - doctors have to rule out other disorders that could cause similar symptoms. Neuro imaging techniques like MRI can also show a doctor the location and type of brain damage hence, helping them in predicting the severity and type of disorder and prognosis accordingly.

1. Peri-ventricular leukomalacia (Fig 9) (most commonly seen)
2. Ventriculomegaly (Fig 10)



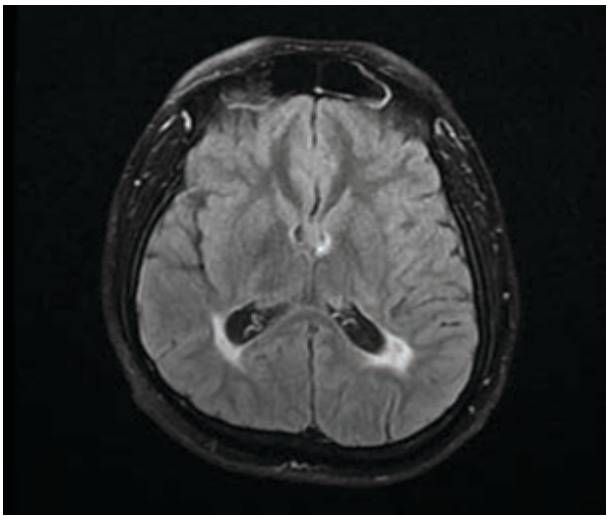
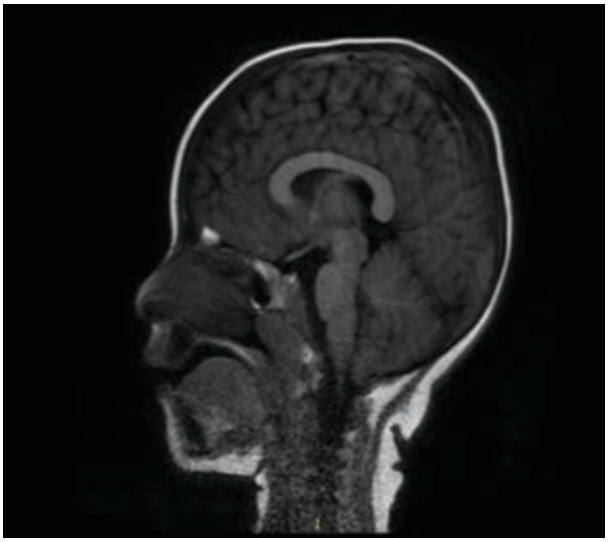


Fig 9 and 10



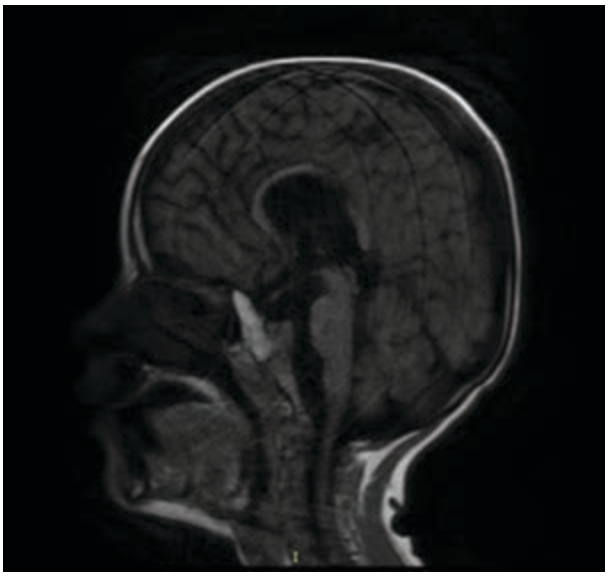


Fig 11. thinning of corpus callosum

Physical Assessment and Developmental Screening Scales:

A large variety of rating / screening instruments have burgeoned within recent years, with varying degrees of accuracy for prediction of

1. Developmental delay
2. Definite developmental abnormality
3. Specific Developmental Diagnosis.

Among those most commonly used at present at the present tiem are the following :

*I ) Developmental Scales:*

1. Amiel -Tison
2. Bayley
3. Brazelton
4. Dubowitz
5. Haataja etal
6. Peabody
7. Prechtl

1. Clinical Adaptive Test (CAT)/ Clinical Linguistic and Auditory Milestone Scale (CLAMS)
2. Knobloch Revised Screening Inventory
3. General Movement Assessment(GMs).

*III) Motor Assessment Instruments:*

1. Alberta Infant Motor Scale(AIMS)
2. Early Motor Pattern Profile(EMPP)
3. Gross Motor Function Measure(GMFM)
4. Movement Assessment Inventory(MAI)
5. Test of Infant Motor Performance(TIMP)

*Developmental Evaluation Procedure*

*1 Developmental History*

- Chief Complaint
- Family and genetic history
  - Pregnancy
  - Labor /Delivery
  - Perinatal /Neonatal
- Developmental Milestones
- Other development al Features.
- Reviews of systems
- Past medical history

*2 Developmental Physical Examination. Developmental Neurological Examination.*

- General Observation.
- Quality of general movements.
- Tone
- Patterns of motor behavior.
  - Primitive
  - Postural
- Sensation
- Cranial nerves.
- Cerebellar Function.
- Dystonia.

Motorsigns

- Uppermotor
- Lowermotor
- **Neurological Soft signs.**

*1 Developmental Screening Instruments.*

**Laboratory Evaluation**

**Medical Management Of Cerebral palsy**

**Diagnosis**

If your family doctor or pediatrician suspects your child has cerebral palsy, he

or she will evaluate your child's signs and symptoms, review your child's medical history, and conduct a physical evaluation. Your doctor may refer you to a specialist trained in treating children with brain and nervous system conditions (pediatric neurologist).

Your doctor will also order a series of tests to make a diagnosis and rule out other possible causes.

Brain scans

Brain-imaging technologies can reveal areas of damage or abnormal development in the brain. These tests may include the following:

**Magnetic resonance imaging (MRI).** An MRI uses radio waves and a magnetic field to produce detailed 3-D or cross-sectional images of your child›s brain. An MRI can often identify any lesions or abnormalities in your child›s brain.

This test is painless, but it's noisy and can take up to an hour to complete. Your child will likely receive a mild sedative beforehand. An MRI is usually the preferred imaging test.

- **Cranial ultrasound.** This can be performed during infancy. A cranial ultrasound uses high-frequency sound waves to obtain images of the brain. An ultrasound doesn›t produce a detailed image, but it may be used because it›s quick and inexpensive, and it can provide a valuable preliminary assessment of the brain.

**PET CT BRAIN: this can be done to know hyper metabolic or hypo metabolic areas of the brain to rule out the functional disorder.**

Electroencephalogram (EEG)

If your child has had seizures, your doctor may order an electroencephalogram (EEG) to determine if he or she has epilepsy, which often occurs in people with cerebral palsy. In an EEG test, a series of electrodes are affixed to your child's scalp.

The EEG records the electrical activity of your child's brain. If he or she has epilepsy, it's common for there to be changes in normal brain wave patterns.

Laboratory tests

Laboratory tests may also screen for genetic or metabolic problems.

Additional tests

If your child is diagnosed with cerebral palsy, you'll likely be referred to specialists for assessments of other conditions often associated with the disorder. These tests may identify:

- Vision impairment
- Hearing impairment
- Speech delays or impairments
- Intellectual disabilities
- Other developmental delays
- Movement disorders
- Treatment

Children and adults with cerebral palsy require long-term care with a medical care team. This team may include:

- **Pediatrician or physiatrist.** A pediatrician oversees the treatment plan and medical care.
- **Pediatric neurologist.** A doctor trained to diagnose and treat children with brain

and nervous system (neurological) disorders may be involved in your child›s care.

- **Orthopedic surgeon.** A doctor trained to treat muscle and bone disorders may be involved to diagnose and treat muscle conditions.
- **Physical therapist.** A physical therapist may help your child improve strength and walking skills, and stretch muscles.
- **Occupational therapist.** An occupational therapist can provide therapy to your child to develop daily skills and to learn to use adaptive products that help with daily activities.
- **Speech-language pathologist.** A doctor trained to diagnose and treat speech and language disorders may work with your child if your child suffers from speech, swallowing or language difficulties.
- **Developmental therapist.** A developmental therapist may provide therapy to help your child develop age-appropriate behaviors, social skills and interpersonal skills.
- **Mental health specialist.** A mental health specialist, such as a psychologist or psychiatrist, may be involved in your child›s care. He or she may help you and your child learn to cope with your child›s disability.
- **Recreation therapist.** Participation in art and cultural programs, sports, and other events that help children expand physical and cognitive skills and abilities. Parents of children often note improvements in a child›s speech, self-esteem and emotional well-being.
- **Social worker.** A social worker may assist your family to find services and plan for care transitions.
- **Special education teacher.** A special education teacher addresses learning disabilities, determines educational needs and identifies appropriate educational resources.

## Medications

Medications that can lessen the tightness of muscles may be used to improve functional abilities, treat pain and manage complications related to spasticity or other cerebral palsy symptoms.

It's important to talk about drug treatment risks with your doctor and discuss whether medical treatment is appropriate for your child's needs. Medication selection depends on whether the problem affects only certain muscles (isolated) or the whole body (generalized). Drug treatments may include the following:

**Isolated spasticity.** When spasticity is isolated to one muscle group, your doctor may recommend on a botulinumtoxin A (Botox) injections directly into the muscle, nerve or both. Botox injections may help to improve drooling. Your child will need injections about every three months.

Side effects may include pain, mild flu-like symptoms, bruising or severe weakness. Other more-serious side effects include difficulty breathing and swallowing.

**Generalized spasticity.** If the whole body is affected, oral muscle relaxants may relax stiff, contracted muscles. These drugs include diazepam (Valium), dantrolene (Dantrium) and baclofen (Gablofen).

Diazepam carries some dependency risk, so it's not recommended for long-term use. Its side effects include drowsiness, weakness and drooling.

Dantrolene side effects include sleepiness, weakness, nausea and diarrhea.

Baclofen side effects include sleepiness, confusion and nausea. Note that baclofen may also be pumped directly into the spinal cord with a tube. The pump is surgically implanted under the skin of the abdomen.

Your child also may be prescribed medications to reduce drooling. Medications such as trihexyphenidyl, scopolamine or glycopyrrolate (Robinul, Robinul Forte) may be helpful, as can Botox injection into the salivary glands.

## Therapies

A variety of nondrug therapies can help a person with cerebral palsy enhance functional abilities:

**Physical therapy.** Muscle training and exercises may help your child›s strength, flexibility, balance, motor development and mobility. You›ll also learn how to safely care for your child›s everyday needs at home, such as bathing and feeding your child.

For the first 1 to 2 years after birth, both physical and occupational therapists provide support with issues such as head and trunk control, rolling, and grasping. Later, both types of therapists are involved in wheelchair assessments.

**Braces or splints** may be recommended for your child. Some of these supports help with function, such as improved walking. Others may stretch stiff muscles to help prevent rigid muscles (contractures).

**Occupational therapy.** Using alternative strategies and adaptive equipment, occupational therapists work to promote your child›s independent participation in daily activities and routines in the home, the school and the community.

**Adaptive equipment** may include walkers, quadrupedal canes, seating systems or electric wheelchairs.

**Speech and language therapy.** Speech-language pathologists can help improve your child›s ability to speak clearly or to communicate using sign language.

**Speech-language pathologists** can also teach your child to use communication devices, such as a computer and voice synthesizer, if communication is difficult.

**Another communication** device may be a board covered with pictures of items and activities your child may see in daily life. Sentences can be constructed by pointing to the pictures.

**Speech therapists** may also address difficulties with muscles used in eating and swallowing.

**Recreational therapy.** Some children may benefit from recreational therapies, such as therapeutic horseback riding. This type of therapy can help improve your child›s motor skills, speech and emotional well-being. Surgical or other procedures. Surgery may be needed to lessen muscle tightness or correct bone abnormalities caused by spasticity. These treatments include:

**Orthopedic surgery.** Children with severe contractures or deformities may need surgery on bones or joints to place their arms, hips or legs in their correct position.

**Surgical procedures** can also lengthen muscles and tendons that are proportionally too short because of severe contractures. These corrections can lessen pain and improve mobility. The procedures may also make it easier to use a walker, braces or crutches.

**Severing nerves.** In some severe cases, when other treatments haven›t helped, surgeons may cut the nerves serving the spastic muscles in a procedure called selective dorsal rhizotomy. This relaxes the muscle and reduces pain, but can also cause numbness.

## Surgery of Cerebral Palsy

Single Event Multilevel Surgery (SEMLS)
&
Percutaneous Apneurotic Slide Surgery (PERCS)



Fig 12. Surgery for Cerebral Palsy



Fig 13 Seven year old boy with spastic diplegia



Fig 14 Six months post surgery - PERCS and standing with orthotics



Fig 15, 16 and 17- Twelve year old boy with typical crouch gait: hip and knee flexed with weak quadriceps



Fig 18, 19 and 20- Post surgery : guarded hamstring lengthening, fractional psoas surgery and patella tendon plication



Fig 21, 22- Fourteen year old boy with severe crouch. After bilateral distal femoral extension deformity, patella tendon placation, foot surgery child ambulatory with crutches.

Crouch gait correction involves muscle rebalancing procedures and require dedicated post operative physiotherapy to achieve optimal results.

Most children with Cerebral diplegia with develop muscle tightness and imbalance during period of rapid growth spurts. This can make standing and ambulation difficult. The muscle tightness and spasticity also consume energy leading to early fatigue and reluctance for the child to stand or walk. Staged surgery and repeated plaster cast treatment cause muscle wasting and weakness. The concept of SEMLS and PERCS were developed with the common goal to achieve the most optimal results with minimal immobilization. The surgery is performed under regional anesthesia and post-operative rehabilitation commences as soon as the child is comfortable.

**Percutaneous aponeurotic slide surgery (PERCS)**

This procedure is indicated for spastic muscles which interfere in positioning and posturing of limb. Involves minimal incisions and can be done at multiple sites. Orthotic support is required after PERCS and rarely plaster casts are required.

**Multidisciplinary Approach to Cerebral Palsy:**

Management of children with cerebral palsy need an integrated approach. Children with Cerebral Palsy have multiple issues: muscle tightness, deformities of limb and spine , speech disorders, epilepsy , poor cognition, visual disturbances and other systemic problems. A multidisciplinary clinic which has a various specialities under one roof will serve the children well so that the treatment plan for each child can be individualized. Before any procedure the child is seen by physiotherpaist, an occuptational therapist A systematic assessment is carried out prior to surgery to identify the goals of SEMLS or PERCS for that particular child and to determine the what is called the “Surgical Dose” required to achieve optimum results.

The surgery is planned after thorough discussion with the parents /caregivers and the treating physiotherapists so that the post operative rehabilitation protocol is tailored to the individual child. Appropriate orthotics are also fabricated

so that plaster immobilization is minimal to prevent muscle disuse. The child is also prescribed appropriate analgesics and muscle relaxants to ease the pain and spasm which can sometimes occur in the post-operative period. Occasionally Botulinum toxin injections are combined with surgery to decrease the need for extensive muscle releases. Also many of these children will need splints and orthotics which help to prevent deformities and to preserve the correction. Please don't hesitate to ask your doctor or physiotherapist whether SEMLS or PERCS Botox or Surgery is appropriate for your child.

## Treatment

Physical therapy (PT) is a branch of rehabilitative health that is considered as important aspect of treating children with Cerebral Palsy. These children experience mobility, function, posture and balance challenges of varying degrees which is benefited by physical therapy – which focuses on basic mobility such as sitting, standing, walking, climbing stairs, reaching or operating a wheelchair.With this book we aim to guide the parents/paramedics with different type of handling techniques. Knowledge about different handling and relaxation techniques will help achieve the best (18)

The main goal of physical therapy is to make everyday movements easier in children's with cerebral palsy. Physical therapy will improve following impairments such as- co-ordination, balance, flexibility, posture, gait, tonal modification, quality of life. With this chapter our main aim is to educate therapist and parents about various treatment procedure which are available and few techniques/ home care exercises which can be implemented by therapist across different professions so that the children can be treated in correct alignment of head and trunk. The treatment of child varies from the severity and type of CP (19)

The Gross Motor Function Classification System (GMFCS) is a five-level classification system that focuses on the voluntary movements of children with cerebral palsy (CP), with specialized focus on walking and sitting. The higher the level in GMFCS, the more severe the CP is. The GMFCS applies to all types of CP and all levels of severity.LEVEL I - Walks without Limitations, LEVEL II - Walks with Limitations, LEVEL III - Walks Using a Hand-Held Mobility Device, LEVEL IV - Self-Mobility with Limitations; May Use Powered Mobility LEVEL V - Transported in a Manual Wheelchair (17, 19)

Treatment for Cerebral Palsy primarily aims to increase mobility and promote physical development such as sitting, crawling and walking. Treatment should be started early and should aim to continue on a regular basis. Every child with Cerebral Palsy presents differently and has different needs. According to detailed assessment every patients therapy is a tailor made treatment program (specific to each child's needs). Treatments aims at- Increase muscle strength and activation, Decrease muscles tightness and spasms through stretching programs and splinting techniques, Encourage physical development through play and everyday activities, Improve mobility with the use of different aids. (19,20)

## Different therapies used by therapist to treat children with cerebral palsy are:

**Physiotherapy**

**NDT (neurodevelopment therapy):** it is a therapeutic approach, which helps in assessment and management of movement problems or dysfunction in children with neurological dysfunction. It aims to maximize child's functional capabilities. It is also known as bobath therapy and was developed in 1940's by Dr and Mrs. Bobath. The treatment aims to make desired movements more possible and prevent undesired movement. It also helps to achieve normal motor milestones in children with cerebral palsy. It is widely used therapeutic approach and gives the best functional output.

*Facilitation:*

The use of primitive or tonic reflexes, quick stretching, tapping, vibration, approximation, and weight bearing may be required to prepare the child for the performance of functional activities. In addition, facilitation can also be in terms of education, mechanical, postural, manual, motivation, etc based on the clinical presentation of the child and goal of therapy.

*Principles of facilitation:*

The most important Kinesiological parameters need to be considered are

1. Base of support
2. Alignment
3. Movement in all three planes(sagittal , frontal and transverse plane)
4. Range of motion

*Inhibition:*

Therapist also uses inhibition to restrict the child's atypical postures and movements that may prevent the development of more selective motor patterns and efficient performance. The therapist achieves inhibition by:

1. Prevention or redirection of components of movements those are unnecessary and interfering with intentional coordinated movement.
2. Constrain the degree of freedom to decrease the amount of force the child uses to stabilize posture.
3. Balance antagonistic muscles
4. Reduce spasticity or excessive muscle stiffness that interferes with moving specific segments of the body

NDT follows the principle of **'Elongation'** first and then **'Activation'.**

**Multiple system impairments:**

1. Base of support: The BOS refers to the area that includes every point of contact that a person makes with the supporting surface. Formation of appropriate BOS is very important as child's postural control emerges by forming required BOS within center of mass. CP children face difficulty in postural control and movement because they are unable to form efficient BOS to work on. Also CP children are unable to maintain COM within their BOS. For example: Level 5 children do not form any BOS. Level 4 and above children either form wide BOS (hypotonic, ataxic) or a narrow BOS (spastic, dystonic). Also look at the extent of base formation, i.e which part of feet or buttocks are in contact with surface. For eg: forming BOS only at feet/ only on medial aspect of feet.




Fig 23- Atypical base of support in sitting

2 Alignment: Term referred to as ability to maintain head, trunk and pelvis in one line. Look for alignment of head, trunk and pelvis in resting postures – supine/prone, sitting unsupported, standing unsupported. Failure to maintain head, trunk, pelvis in one line and tendency to weight bear on one side than equal weight bearing leads to assymetry usually seen in children with CP. Can be caused due to involvement of one side of the body more than the other or it can be as a result of compensation to maintain stability of the body in a particular position.




Fig 24, 25- Improper v/s proper alignment




Fig 26, 27- Improper v/s proper alignment

3 Weight shifts (in all planes): Ability to shift weight of body from one point to other and thus shifting COM relative to BOS in a particular direction. Three planes of weight shifts (chronology of appearance): sagittal, frontal and transverse. Check child's ability to perform weight shift in above planes focusing on maintenance of adequate BOS, overuse/under use of muscles

4 Stability V/s mobility: Stability in a particular position or in a movement, stability is achieved through activation and co activation of postural muscles. CP children face difficulty in maintaining stability as postural system is weak or movement system is over working. Thus, assess how the child is stabilizing himself (wrong postures) and aim for correcting them and facilitating good postures. Ideally when shifting weight mobility is present at trunk pelvis (axial, in transverse plane motion). But children fail to attain trunk pelvis mobility. Rather they move from their upper thorax or just at neck with compensations at other distal joints. Assessing point of mobility is thus important to achieve appropriate weight shift in our treatment

5 Postural control: Allows to maintain certain posture against gravity with minimum effort along with stability and accuracy in the performance of voluntary movements. It is innate and life-preserving ability of postural muscles. Begins with the drive for verticality and facilitates antigravity muscle activity which initiates with extensors activity before flexors activity and Progress from Head to toe, Proximal to distal, Begins with head control. Prepares the body for movement- Maintains alignment of body, Overcomes the effect of gravity in all positions at all times, Preserves joint integrity, Counterbalance movement that changes COM, Maintains COM within BOS

Postural adjustments: Anticipatory postural adjustments-Feed forward mechanism which is the response to counteract the prediction of pertubations and Protects stability balance. Direction specific patterns of postural muscle gets activated to counter respond to perturbations. Distal to proximal activation of muscles with Posterior muscles of trunk and upper leg activated prior to arm flexion as a response. Anterior muscles of trunk and upper leg activated prior to arm extension as a response.

Compensatory postural adjustments: Occur after perturbation. Triggered by sensory feedback signals. Reorganize posture and maintain balance. Postural muscle activity is observed after the body perturbation has already occurred. Pertubation is generally external and unexpected. Fully developed in sitting by 3years. Fully developed in standing by 7 to 10 years

6 Atypical movement patterns: A healthy and harmoniously working postural and movement system results in forming of a good postural synergy in any position. CP children fail to attain any postural synergy/ good postural synergy due to weak postural muscles/ over recruitment of movement muscles. Thus, assessing and knowing how and to what extent child can form postural synergy helps you to move ahead with your treatment

## Single system impairments:

A. **Neuromuscular system**

1. Timing and Sequencing: initiation, sustenance, termination
2. Recruitment of postural and movement muscles
3. Coactivation ability of muscles
4. Graded control of movements

B. **Musculoskeletal system**

1. Tightness
2. Contracture
3. Weakness

C. **Sensory systems:** mention about different sensory systems such as visual, auditory, tactile, vestibular and proprioceptive systems.

D. **Respiratory system:** mention about ribcage mobility, chest expansion.

Chest development of children with significant trunk muscles weakness and/ or tonal imbalances because of abnormal muscle/tone function, the disabled child is incapable of independently counter acting or balancing gravity's influence. Children with trunk muscle dysfunction in CP can present a different thoracic abnormality. Children who demonstrate weak abdominal muscles, but have ability to expand chest laterally expand the lower chest wall, may develop a lateral flaring of ribcage. Due to improper positioning - in case of supine position much of the time, the lower border of the ribcage are likely to be flared out laterally because this motion is in a gravity eliminated plane.





Fig 28- Abnormal chest development

**Abnormal chest development** as seen above refers to as: If a child lacks adequate intercostal muscle support the chest wall may be drawn in towards the lungs due to negative pressure gradient generated during inhalation. The anterior posterior and transverse dimensions of the chest thus decrease rather than increase during inhalation. This appearance looks like intercoastal retraction or in more severe cases as a pectus excavatum deformity. Ribcage positioned high within trunk and decreased mobility in ribcage for expansion with respiration and ribcage or trunk move as a unit

A. **Cardiovascular system:** use of diaphragmatic and accessory muscles of respiration, endurance, fatigue

B. **Gastrointestinal system:**constipation, delayed gastric emptying, chronic reflux

C. **Integumentary system**

**Evaluation :** Prioritize the impairments based on posture and movements problems. List and prioritize the functional activity and limitations and participation restriction. Analyze the relationship of issues for a given individual

## DYSTONIA

Strategies used to work with patients suffering from Dystonic type of CP:

| Multisystem impairments | Single system impairment |
|---|---|
| Decreased upright stability and lock the joints<br>Uses assymetry for stability<br>Uses wide BOS<br>Difficulty in assuming and maintaining posture due to involuntary movement | Neuromuscular: Sustained contraction (decreased ability to terminate muscle contraction) in various parts of the body and atypical timing of muscle activation (off and on) trunk problems with I/S and can see collapse in kids |
| Cant use upper extension to support as they are in end ranges and bear weight on the dorsum of the hand | Decreased recruitment of postural motor unit and over recruitment of multi-joint muscles |
| May develop contractures, deformities, and pain over life span due to subluxation and stress injury | Increased co-contraction of agonist and antagonist because they have sustain activation rather then allowing automatic/ reciprocating movements |
| May be better in standing, then sitting, then prone<br>Decreased anticipatory activation<br>Atypical force production | Stiffness can be either increased or decreased and varies dramatically. It is decreased during sleep and increases with stress and emotion |
| Decreased eccentric control muscle start contract concentrically in burst to the end range but then they hold isometrically so-very limited mid range control is available | Synergies limited, but may vary day to day, massed recruitment within involved body parts, twisting in nature can be either in flexion or extension |
| Dystonia is frequently associated with athetosis or ataxia | Sensory system: Decreased modulation of regulatory system, Atypical arousal, Poor state regulation, Problems with visual motor system, Pursuit movements, convergence, Hearing impairment, Difficulty in modulating of sensory information and grading response, Decreased awareness of position of body segments and their relationships |
| | Musculoskeletal system:Soft tissue length, Hypermobility/ instability, extremities, Hypomobilities: trunk, thoracic spine and connective tissue, Strength. |

Strategies used to work with patients suffering from Dystonic type of CP:

| **Dystonia** | |
|---|---|
| **Multiple and single system impairments** | **Treatment strategies used** |
| Hypo to hyper recruitment of movement muscles | Use of weight bearing and compression resistance to improve postural muscle recruitment |
| No sustained postural holding.<br>Difficulty in assuming and maintaining functional positions due to destabilizing effect of involuntary movement and affected co contraction<br>Locks joints in end ranges | Use distal key points to improve proximal activation and reduce involuntary movements<br>Work to move proximal joints over distal ones<br>Use closed chain transitions and postural activities<br>First work on isometric holding, then eccentric work |
| Wide ranges of movements with no grading | Work on small, mid range graded activity with postural stability |
| No initiation of weight shift from base of support<br>Usually wide BOS | Establish a narrower base of support and initiate activity within it |
| Uses asymmetry for stability | Work towards symmetrical postural control, introduce rotation to prevent 'fixing' |
| Appears to enjoy movement | Repeated learning is important |
| Wide ranges of movements in a variety of patterns | Concern for contractures over a long period of time |

## Spasticity

**Working with a spastic CP baby:** Based on ICF model the treatment goals should be directed according to limitations and particapation

| **Functional activity limitations** | **Participation restriction** |
|---|---|
| • Quadriplegia-<br>Bed and floor mobility and transfers<br>Feeding and swallowing<br>Reaching, grasping, manipulations and manual skills<br>Visual impairments<br>Cannot sit, stand and walk<br>• Diplegia-<br>Independent transfers from floor to standing and walking<br>High level ADL skills<br>Visual, motor and perceptual skills | • Diplegia-<br>High level of ambulatory skills<br>Play with peers<br>Fall behind school work<br>• Quadriplegia<br>ADL- full development<br>No verbal communication and poor nutrition orally<br>Mobility and transfers<br>No binocular vision |

| Multisystem impairments | Single system impairment |
|---|---|
| Poverty of movement | Increased use of superficial two-joint muscles |
| Decreased muscle synergies | May see phasic bursts especially in two joint muscles |
| Stereotypical movements | Difficulty initiating and terminating purposeful muscle activity |
| Difficulty maintaining upright postures | Under recruitment/central weakness of postural muscles |
| Mal-alignment throughout the body | Mass recruitment of extension/ flexion with decreased isolated control between and within the extremities |
| Uses of asymmetry | Increase stiffness distally (extremities) than proximally (trunk and oral ) motor |
| Purposeful movements primarily in sagittal plane | |
| May use narrow BOS | |

**Treatment strategies used for the same:**

1. **Capital flexion:** babies develop capital flexion at 3 to 4 months of age, which helps counterbalance the cervical extension, develop graded ranges of isolated head motion, downward visual gaze.

To facilitate capital flexion, therapist applies sustained traction between her hands to elongate spinal extensors in preparation for antigravity flexor control.

2. **Facilitating posterior pelvic mobility:** the therapist supports the pelvis in an elevated position, and waits for the child to bring his legs closer to his hands and mouth. In this position, the baby brings attention to the lower extremities through tactile, visual or auditory inputs and initiates rolling.

3. **Abdominal control:** abdominal control develops when the baby attains the above position and they begin to roll.

The therapist provides a connection between the lower ribcage and the pelvis as the abdominal would do, by gently traction the lower ribs towards the pelvis.

4. **Anti-gravity flexor control:** The therapist helps get the child his hands to knees and feet together. This position helps activate the neck and trunk flexors and gives baby a sense of midline control.

5. **Ribcage mobility:** infants begin to gain ribcage mobility in prone, supine and other positions throughout the first year of life. Ribcage mobility helps child prepare for appropriate alignment of the spine and ribcage for optimum activation of trunk muscles and postural control.

The therapist places the child on her lap in side lying position and gives diagonal compression to the rib under her fingers in the direction of the opposite hip. This procedure is repeated for the next 3 ribs and repeated on the other side as well.



Fig 29- Ribcage mobility

6 **Prone on forearms:** Prone position is challenging for atypically developing children and in case of respiratory, gastrointestinal problems.



Fig 30- Prone on forearms

7 **Caudal weight shift in prone:** In order to reach for toys in prone on elbows position, the child must be able to shift his weight caudally onto the lower abdomen and thighs which also elongates rectus abdominis and hip flexors.

To assist this weight shift, the therapist elongates the rib cage muscles by moving ribcage towards the pelvis while gently using thumbs over paraspinal musculature to facilitate extension of lumbar spine. Therapist can also perform a lateral weight shift in this position to facilitate work of abdominal obliques.

8 **Prone on hands:**



Fig 31- Prone on hands

9 **Activate postural control and vestibular system on ball(**Fig 32**):** Placing the child prone on forearm on a therapy ball, the therapist places hands around humeri keeping them in neutral rotation, and giving gentle compression towards the weight bearing surface. As the child activates his postural muscles, therapist moves the ball laterally to provide vestibular input and further challenge to postural muscles in frontal plane.



Fig 32

10 **Side lying positioning**(Fig 33)**:** Once a baby can tolerate side lying, therapist places the child in side lying, puts the upper leg in front of the lower leg and externally rotates so that the child experiences weight bearing on that foot. Side lying position helps facilitate hand to mouth activity, improve tactile awareness and also elongation of lateral aspect of trunk, and selective graded control between both the legs.



Fig 33

11. Facilitation of rolling(Fig 34):

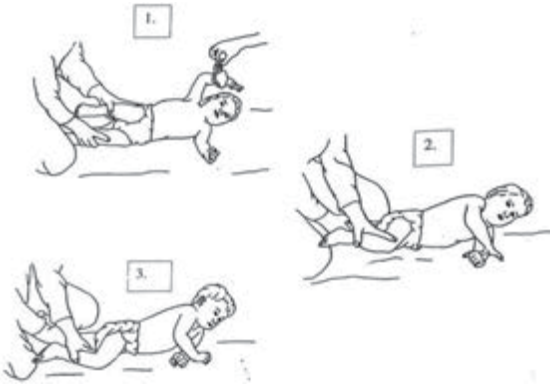



Fig 34

12. Facilitation of head control(Fig 35):

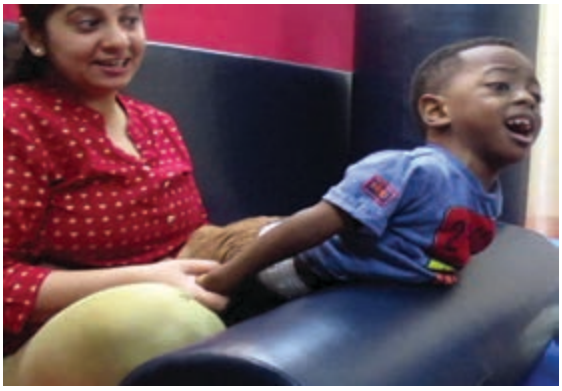


Fig 35

13. Elongation of anterior chest wall musculature(Fig 36, 37 and 38):



Fig 36



Fig 37



Fig 38

14. Activation of postural muscles in sitting(Fig 39):



Fig 39

II. Elongation of hip flexors (Fig 40):



Fig 40

15. Elongation of hamstring muscles (Fig 41):

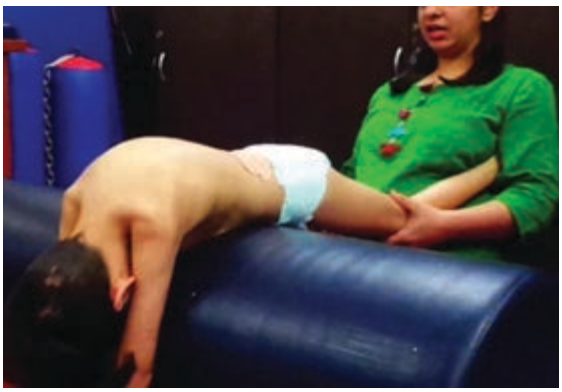


Fig 41

16. Facilitation of rotation and reaching(Fig 42):



Fig 42

17. Establishing base of support and loading of feet(Fig 43,44):



Fig 43



Fig 44

18. Assisting bear position(Fig 45):



Fig 45

19. Facilitating half kneel position(Fig 46, 47):



Fig 46



Fig 47

20. Facilitating unilateral weight bearing and lateral weight shifts (Fig 48, 49 and 50):



Fig 48



Fig 49

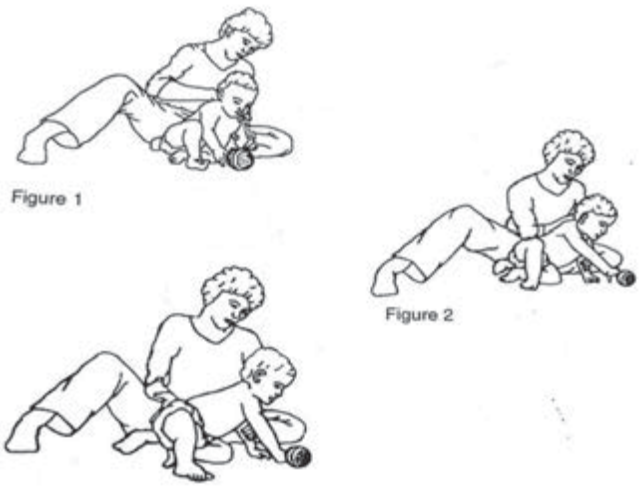



Fig 50

**21. Facilitating sagittal weight shift (for tranasition) (**Fig 51**):**



Fig 51

**Normalizing tone of the muscles:**

In the infant with CP, spasticity is seldom significant in the first 6 months, but sometimes between 6 months to 24 months, it starts becoming apparent. Spasticity is defined as hyper excitability of the stretch reflex, resulting in a velocity dependent increase in muscle tone. The muscles produce more force for a given change in length and therefore feel stiff and resist passive lengthening.

In children with hyper tonicity slow passive movements, sustained stretch, cryotherapy over the muscles for 15 to 20 minutes, stimulation of antagonist movement and vibrations are used. On the contrary, for the cases with hypotonicity weight bearing, joint compression, rhythmic stabilization, vibrations, cryotherapy in brisk manner and tapping is used.

**Developing postural reaction:**

Postural adjustments are essential if the child is to move freely and to adjust to various environmental demands rapidly. Motor milestones can be best achieved by good postural reactions. Postural reaction consists of righting reactions, protective extension and equilibrium reactions. Initially children first develop righting reactions. Righting reaction allow the child to orient his head in space so that the eyes and mouth are horizontal regardless of the position of the body. It also helps in maintaining the proper alignment of the head with respect to the body and vice versa. Once righting reaction develop in a particular position protective extension comes up. Lastly equilibrium reaction emerge which helps the patient to counteract the opposing force and enable them to maintain balance. These reactions are best developed by various exercises on vestibular ball, swings and tilt board.



Fig 52- Developing postural reactions

## Strengthening Programs:

Children who cannot walk are by definition weak. There is no evidence that strengthening increases spasticity. Infact, there is evidence that weakness is a problem in this population of children and strengthening is effective in reducing weakness and improving function. Therapist need to lengthen spastic agonist and strengthen the antagonist. For very young children, weight training may not be appropriate but therapists can incorporate strengthening activities in to child's

therapy program through games and repetition of functional movements. eg: In Partial weight treadmill training child is suspended in a specialized harness and helps strengthening the weak antigravity muscles and facilitate normal gait patterns.

## Stretching and Mobility:

The muscles should be maintained at the appropriate physiological length for normal movement control and normal postural adjustments. In CP because of delay or absence of normal movement muscles are usually in a shortened position hence stretching of the muscles is essential to increase the neuromuscular control. Length of the muscles should be maintained not only through stretching but also through various functional activities. Thus home exercises and activities are very important in additional to the therapy at the pt clinic.

Types of elongation:

1. Active elongation





Fig 53 and 54- Stretching and mobility

2). Passive elongation







Fig 55, 56,57- Passive elongation

## Working with an Ataxic child

Any damage to the cerebellum and associated tracts results in an Ataxic type of presentation.

Common impairments seen in ataxic |cases are

i. abnormal/poor postural stability,
ii. poor coordination,
iii. poor alignment,
iv. hypotonia,
v. difficulty in grading and sustaining movement,
vi. tremors,
vii. vision - nystagmus, upward gaze.

Following are some compensation strategies ataxic children use for maintaining stability:

i. Wide base of support
ii. Fixation with eyes
iii. Rigid holding of trunk, locking knees and elbows in end ranges when weight bearing, clasping of hands
iv. Prefer midline and resist any movement attempt

Thus, treatment strategies related to functional goals should aim to:

i. Decrease joint and spine stiffness by working on ribcage mobility and improve chest expansion as it enhances movements
ii. Reduce fear with slow movements
iii. Achieve downward gaze and saccadic eye movements
iv. Elongate/ Align / Activate
v. Develop sustained muscle activity using closed chain activities
vi. Initiate rotation
vii. Achieve movements away from midline but in gradations
viii. Reinforce base of support and reduce involuntary movements by giving intermittent joint compression

## Working with a hypotonic child:

A hypotonic presentation could be seen in case of ataxia, dyskinesia, transitional phase towards hypertonia, GDD and syndromes such as Down's syndrome, Angelman's syndrome, delayed myelination, metabolic disorders, etc.

Common multisystem impairments seen in a hypotonic child are wide base of support, poor alignment, decreased tone, weakness - difficulty in sustaining muscle activity in mid range, poor postural control and stability, delayed/absent equilibrium reactions.

Common neuromotor impairments include poor recruitment of muscles, poor gradation of activity, insufficient force generation, whereas sensory system impairments include poor registration of proprioceptive/ tactile/ somatosensory inputs.
Therapeutic approaches are:

1) general strengthening-



Fig 58- Forward and back ward kneel walking



Fig 59- wheel barrow walking



Fig 60- Back extensor strengthning

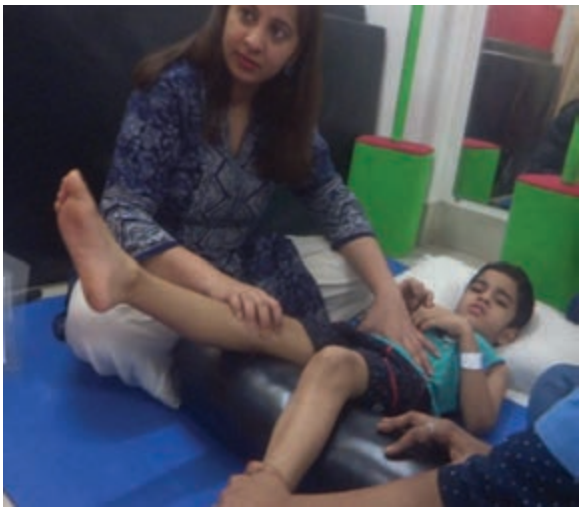




Fig 61, 62- Knee extensors strengthning



Fig 63- Hip extensors strengthening



Fig 64- Bridging

*Sensory integrative therapy:*

Sensory integrative (SI) therapy, was developed by Jean Ayresinthe1970's, is based on promoting the organization and processing of sensory information. Proponents believe that information coming into the body from the proprioceptive, tactile, and vestibular systems is disorganized and not processed well. This lack of cognitive processing is considered a perceptual problem. These processing problems cause difficulties with motor planning and motor control. Therapy focuses on movement and environmental awareness. The use of swings, scooters, and other moving objects helps the child process sensory input and use sensory information to plan movement and gain postural control. Traditionally, occupational therapists are more involved in the use of SI treatment.

Children with CP often have sensory processing dysfunction (SPD) and show sensory based motor dysfunction, Sensory processing evaluates how the brain takes it what it receives through the senses. In SI the rapystate of arousal of child is very important for learning movement. Depending on arousal the rapist will up regulate or down regulate sensory stimulation. Use of varying textures like cotton, rough gauge, deep pressure are used to stimulate kinesthetic and Proprio receptive input.

## Transition Training

### 1) Rolling:

Technique:

Hold the front of the knees, turning the legs out

Encourage your child to go forwards onto their hands, keeping the elbows straight.

When they can do this well, encourage them to reach out for a toy while keeping the as shown in the images in next slide



Fig 65- Rolling

Rolling transition training on moveable surface for assisting a child using distal key point of control over feet (hands below knee) as shown in image



Fig 66- Rolling

Assisting rolling using distal key point of control using hands as shown in image



Fig 67- Rolling

**2) Lying to sit position:**

Lay the child infront of you

Bring the weight backwards over the knees, holding over child's hips

Encourage the child to push up with their hands

Move the child's hips slowly over to one side so the child is now sitting as shown in image



Fig 68- Lying to sit position

**3) Reaching out in sitting:**

Hold onto child's legs to give support

Encourage them to reach up and out sideways for a toy

This helps to develop balance and helps to open up their chest along with eye hand co-ordination

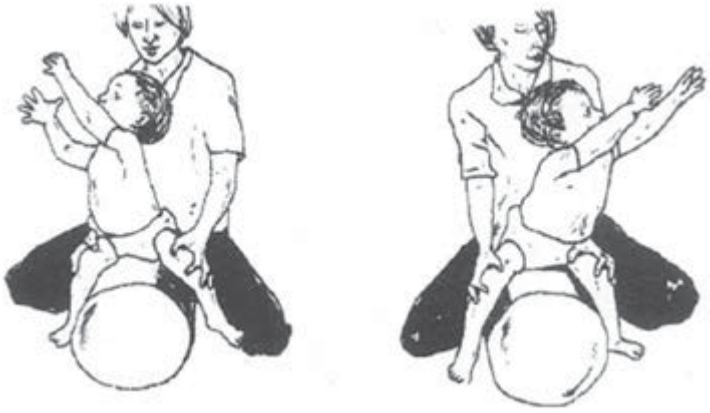


Fig 69- Reaching out

**4) Sit to stand (pull to stand):**

Hold the arms as shown such that their palm face each other

Bring the child's weight forward over their feet and then help them to stand up



Fig 70- Pull to stand

**5) Kneeling down:**

Support your child at the hips

Encourage your child to stay up on their knees, with the weight supported equally by both legs

Kneeling against a table is easier to start with

Later, using a ball is good for developing balance and control over hip



Fig 71- Kneeling down

**5) Encouraging the crawl position:**

Place your child in the crawl position

Support them at the hips

Encourage your child to stay in this position for a short while



Fig 72- Crawl position

**6) Kneeling up on one leg**

Hold your child at the hips.

Make sure their legs are apart.

Play games in this position and encourage your child to keep their balance.



Fig 73- Half kneeling

**7) Encouraging standing: holding the knees:**

Stand your child astride a roll.

Hold the knees, with the child's feet facing forwards.

Encourage balancing in this position and reaching out for toys.



Fig 74- Standing

**8) Standing against a high table or bench:**

Find a surface that your child can use to help them to remain standing up, as shown.

Encourage them to keep their balance in this position



Fig 75- Standing against a high table or bench

**9) Standing with one foot on a step**

Support your child at the hips

Encourage body weight to be forward over the front foot



Fig 76- Standing with one foot on a step

3) Lifting and carrying techniques

Carrying baby curled up

Carry your baby in a curled up position with the shoulders forward and the hips bent up.

Support behind the neck and shoulders, not just the head

Carry your child as shown, with their legs around your hips

Support them under their bottom- carrying baby on hips

Carrying in the 'aeroplane' position

Carry the child with your arm between their legs and your hand under the chest or, if they are bigger, with one arm under the chest and shoulders and the other keeping the legs up

Try to keep their arms and legs turned out

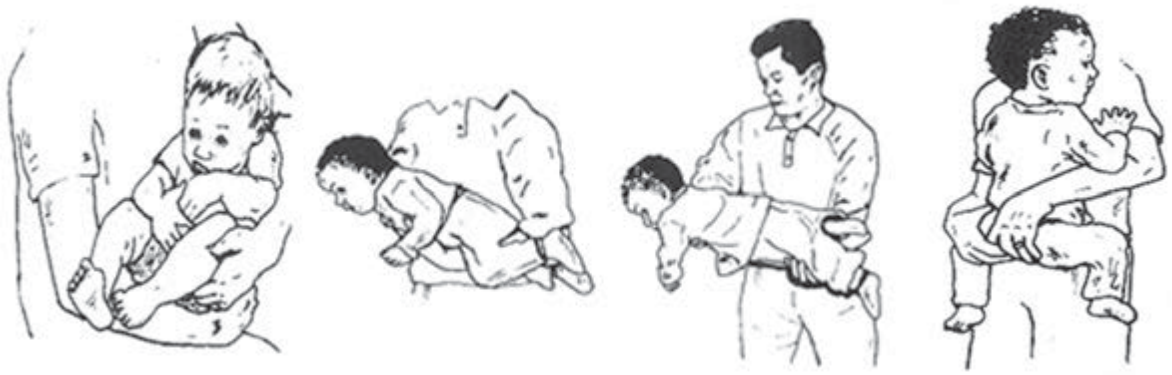


Fig 77- Lifting and carrying techniques

***Pediatric Constraint Induced Therapy:***

Constraint induced therapy is used to improve the use of affected upper extremity in a child with Hemiplegic CP. The normally functioning or stronger upper extremity is immobilized for a variable duration in order to force the use of the affected or weaker upper extremity overtime.

The efficacy of this approach has not been established, and the adverse effects of prolonged immobilization often normally developing upper extremity are a significant concern. (17)

***Rood approach:***

Rood utilized both the sensory and motor systems to facilitate movement with her treatment techniques. She felt that preventing the development of abnormal movement patterns was important. She used sensory input to achieve or relax muscles to promote normal movement. For example, during the rapygentle stroking is used to have relax atoryeffecton the muscles there by used to inhibit spastic muscles. On the other hand brisk stroking, tapping, quick ice, etc, has been found to develop tone in the muscles, hence can be used for cases like flaccidity. (17)

***Vojta's Technique:***

This approach was introduced by Dr. Vaclac Vojta and used afferent sensory stimulation through touch, stretch and pressure for facilitation of movement. Reflex locomotor

patterns and proprioreceptive input are the basis for treatment.

***Theratogs:***

They provide joint stability and increase body awareness. Improves posture, balance, gait and movement skills. More information available on www.theratogs.com. Stabilising Pressure Input Orthosis (SPIOS) are flexible, provide dynamic stability and balance thereby increasing body awareness in space. (27)

## Bracing and Orthosis:

The role of orthoses in CP is :

1. Improve function andefficiency.
2. Improve joint biomechanics andalignment.
3. PreventDeformity.
4. Protection aftersurgery.
5. Encourage a normal motor patterning. The most commonly used Orthoses are:
    - Ground reaction AFO(GRAFO)
    - AFO's Solid andhinged.
    - Supra malleolarOrthoses.

Child made to walk with AFOs of rehabilitation, controlling ankle joint motion produces an effect on the knee during gait and by using an AFO to manipulate the ankle rockers, therapist can increase or decrease the plantar flexion knee extension couple. The correct selection and prescription of lower limb or thos is isessential to maximize a child's rehabilitation. Orthosis for correct period of time gives the bestresults. (26)



Fig 78- Orthoses

Trigger points were used to reflexly to stimulate creeping,rolling, or crawling activities. Proponents of Vojta suggest that very early intervention does not allow the development of abnormal crawling patterns and subsequently promotes normal walking patterns.

**Occupational therapy Intervention**

The goal of Occupational therapy is for the child to participate as actively and fully as possible in all areas of ADL's-self care, play, and school skills. Pediatric Occupational therapists often use a variety of approaches in assessing and treating children, including neuro-developmental treatment (NDT), sensory processing, and motor learning approaches. Therapy is child directed and based on activities that are meaningful and purposeful to that specific child.

Occupational therapists also assess and incorporate various tools and adaptive equipment to increase independence. Examples include specialized feeding utensils, adaptive scissors and writing utensils, splints, and adaptations to clothing such as zipper pulls, button hooks, and reachers.

Occupational therapists may work with the child to develop fine motor tasks such as writing and drawing, self-care activities such as eating, dressing, grooming and toileting, and play and social skills, which can help reduce demands on teacher aids and caregivers and boost the child's independence and self-esteem. Occupational therapy also addresses cognitive and perceptual disabilities especially in the visual motor area. Therapists may also assist with the adaptation of equipment, such as seating, or appropriate technology to allow enhanced participation at school. Additionally, they may be involved with developing organisational skills related to planning, initiating, completing and modifying tasks to ensure and enable completion of routines and problem-solving associated with new activities.

Parental counselling is another important aspect of the occupational therapist with regards to optimizing parental support for improving functional abilities of child with CP. Different approaches to treatment are taken and considered. Since no child is the same, intervention for each child is specific and unique and related to the skills the strengths and limitations of the child.

**Frames of Reference**

Conceptual frameworks and Models are essential for determining "best practice". Best Practice involves imaginative problem solving, creative application of knowledge and research and evaluation of effectiveness of Intervention to bring about improved client outcomes and effect organizational change. Depending upon the impairments in a child a suitable frame of reference or combination of approaches is used Examples of specific occupational therapy Frame of Reference models which are used in evaluation and treatment in CP include the:

- Bio mechanical model
- Canadian Model of Occupational Performance (CMOP)
- Cognitive Disabilities model
- Lifestyle Performance Model
- Model of Human Occupation (MOHO)
- Neuro developmental therapy (NDT)
- Occupational Performance Model (OPM)
- Person Environment Occupation Performance Model (PEOP)
- Sensory Integration model

**Occupational therapy assessment scales**

Occupational therapy services may include comprehensive evaluations of the home and other environments (e.g., school), recommendations for adaptive equipment and training in its use, and guidance and education for family members and caregivers.

OT interventions can be classified into five specific intervention categories,

1. Training of sensorimotor functions including play activities to facilitate motor performance.
2. Training of daily activities skills

including feeding, dressing, bathing, personal hygiene, writing etc.

3. Parental counselling in which parents are educated how to stimulate Independence in their child.
4. Advice and instructions regarding use of assistive devices including provision of mobility aids like wheel chair and bathroom devices.
5. Provision of splints such as hand orthosis to facilitate hand functions.

The above Classification is based on the International Classification of Functioning Disability and Health (ICF) and enables the categorization of all intervention possible in OT

The clinical Occupational therapists must be aware of the complexities faced by a cp child and more than one assessment may be needed to understand the unique skills and behaviors of these children. The following are the occupational therapy tests that can be used specifically in the assessment of Cerebral palsy:

• BRUININKS-OSERETSKY TEST OF MOTOR PROFICIENCY (BOTMP)

**Purpose:** Developmental motor skills

**Age Range:** 4.5 -14.5 years

**Areas Tested:** Balance, strength, coordination, running speed and agility, upper limb coordination (ball skills), dexterity, fine motor control, visual-motor ability.

• CANADIAN OCCUPATIONAL PERFORMANCE MEASURE (COPM)

**Purpose:** To detect changes in parent or child's self-perception of performance over time.

**Age Range:** Any age

**Areas Tested:** Satisfaction and disability rating of daily activities and routines, which are, identified by the child and family as important part of daily life

• FUNCTIONAL INDEPENDENCE MEASURE FOR CHILDREN (WeeFIM)

**Purpose:** To determine the severity of a child's disability, the measurement of caregiver assistance needed in the performance of functional activities, and outcomes of rehabilitation

**Age Range:** Children without disabilities: 6 months to 8 years; Children with developmental disabilities: 6 months to 12 years; Children with developmental disabilities and mental ages less than 7 years

**Areas Tested:** Eighteen items grouped into two major categories of function, motor, and cognition that are divided into six domains divided into sub domains: Motor, Self-care: eating, grooming, bathing, dressing, toileting, Sphincter control: bladder and bowel management, Transfers: chair, wheelchair, toilet, tub, and shower, Locomotion: wheelchair/ crawl, stairs, Cognitive -Communication: comprehension, expression, Social cognition: social interaction, problem solving, and memory.

• GROSS MOTOR FUNCTION MEASURE(GMFM)

**Purpose:** To evaluate change in gross motor function in children with cerebral

palsy, describe a child's current level of motor function, and determine treatment goals.

**Age Range:** No specific age range is recommended by the authors; however, the test has been validated on children between 5 months and 16 years. Seems best suited for children two to five years

**Areas Tested**: Eighty-eight items of gross motor function divided into five dimensions: -Lying and rolling-Sitting Crawling and kneeling-Standing-Walking, running, and jumping. Items were selected to represent those typically performed by children by age five

• GROSS MOTOR FUNCTION CLASSIFICATION SYSTEM (GMFCS)

**Purpose:** To classify a child's present gross motor function.

**Age Range:** 12 months to 12 years

**Areas Tested:** based on self-initiated movement, with emphasis on sitting, transfers, and mobility

• HOME OBSERVATION FOR MEASUREMENT OF THE ENVIRONMENT (HOME)

**Purpose:** A screening tool to identify the quality and quantity of social, emotional and cognitive supports available to the child in the home environment

**Age Range:** Infant and toddlers version birth to three years of age.

**Areas Tested:** Infant and toddlers version: forty-five items clustered into six subscales: Parental responsivity, acceptance of child-Organization of the environment-Play materials-Parental involvement with the child.

• ORAL MOTOR/FEEDING RATING SCALE

**Purpose:** To document oral motor/feeding patterns and feeding function

**Age Range:** One year through adulthood

**Areas Tested:** Two major areas of oral motor/ feeding behavior: Oral motor/ feeding patterns lip/cheek movement, tongue movement, jaw movement. Related areas of feeding function: self-feeding, adaptive feeding equipment, diet adaptation, position, sensitivity, food retention, swallowing, oral-facial structures.

• PEDIATRIC EVALUATION OF DISABILITY INVENTORY (PEDI)

**Purpose:** To determine functional capabilities and performance, monitor progress in functional skill performance, and evaluate therapeutic or rehabilitative program outcome in children with disabilities

**Age Range:** Six months to seven years, six months

**Areas Tested:** Two hundred seventy-one items divided into three subtests in the Functional Skill Scale:-Self care: eating, grooming, dressing, bathing, toileting-Mobility: transfers, indoors and outdoors mobility-Social function: communication, social interaction, household and community tasks. Also environmental

modification and amount of caregiver assistance is systematically recorded in Modification Scale and Caregiver Assistance Scale.

• SENSORY INTEGRATION AND PRAXIS TEST (SIPT)

**Purpose:** Measures sensory systems contributions to balance and motor coordination

**Age Range:** 4-8 yrs 11 months

**Areas Tested:** Numerous tests of postural control, motor coordination & planning, fine and gross motor function, & sensory integration.

• THE ALBERTA INFANT MOTOR SCALE (AIMS)

**Purpose:** To identify motor delay and to evaluate maturation over time. Fifty-eight items related to posture, movement, and weight bearing in prone, supine, sitting, and standing . In addition, the AIMS may also be useful in designing and monitoring a treatment program

**Age Range:** Used during first year of life
**Type of Test:** 58 item, performance-based, norm-referenced, observational too

**Areas Tested:** gross motor maturation, to trace motor retardation and to identify infants that might benefit from early intervention.

• TEST OF INFANT MOTOR PERFORMANCE (TIMP)
The TIMP is a test of functional motor behavior in infants

**Purpose:** a criterion-referenced measure designed to evaluate motor control and organization of posture and movement for functional activities in infants

**Age Range:** 32 weeks gestational age to age 4 months

It predicts 12-month motor performance with sensitivity 92% and specificity 76% and preschool motor performance with sensitivity 72% and specificity 91% at 3 months of age

**Areas Tested:** 27 observed behaviors and 26 elicited behaviors assessing the ability to orient and stabilize the head in space and in response to auditory and visual stimulation in supine, prone, side lying, upright, and during transitions from one position to another, body alignment when the head is manipulated, distal selective control of the fingers, wrists, hands, and ankles, antigravity control of arm and leg movement

• SCHOOL FUNCTION ASSESSMENT (SFA)

Designed to facilitate collaborative program planning for students with a variety of disabling conditions.

**Purpose:** Used to measure a student's performance of functional tasks that support his or her participation in the academic and social aspects of an elementary school program.

**Type of Test:** criterion-referenced assessment

**Areas tested:** Three parts: Participation in school activity settings; Task supports; Activity Performance. Includes physical and cognitive/ behavioural tasks.

**Occupational therapy strategies**

In cases of Cerebral palsy, Occupational therapists work on the following :-

**1. Development of Postural Control**

Postural control is the ability to maintain balance and alignment while upright in space. The child with CP has poor postural control due to delayed or incomplete motor development, decreased voluntary control, lack of stability, difficulty in antigravity movements, lack of dissociative movements, exhibition of stereotypical movements with compensatory mechanisms. The development of postural control is also affected in presence of deficits in the organization of sensory inputs. Therefore Interventions are aimed at:-

- Facilitation and development of antigravity movements, e.g. lowering a child from a sitting to supine position to facilitate neck flexion against gravity. Neck and trunk extension can be promoted by placing the child in prone positions over a wedge or large therapy ball



Fig 79- Development of Postural Control



Fig 80- Development of Postural Control

- Facilitation of automatic reactions like Righting, equilibrium and protective reactions, e.g. placing the child on unstable surfaces and moving the surface using variations in speed, ranges and rhythms. Reach out activities while sitting on ball, bolster or balance board.



Fig 81- Facilitation of automatic reactions

- Facilitation of sensory organization. E.g. walking on different texture surfaces with various visual conditions like closed eyes or dim lights. Reaching out while swinging on different types of swings




Fig 82- Facilitation of sensory organization

- Facilitation of anticipatory control. e.g. catching and throwing a ball. Kicking a ball.

Occupational therapists use Neurodevelopmental Treatment (NDT) Approach which is aimed at facilitating and normalizing hyper- or hypotonic muscle reflexes using facilitationmechanisms; reaction and movement patterns; and managing the specific reactions to the treatment of equilibrium. A solid understanding of typical developmental sequences

is essential for the treating therapist to facilitate postural activities at higher developmental levels for improved learning. E,g To achieve good sitting posture and balance the child needs practice moving over the base of support and coming in and out of sitting, and control of coming to stand from sitting.

**2. Sensory Integration (SI)**

SI is the ability of the CNS to process information to make adaptive response to the environment. The CP child may have sensory integration problem which are:

- Sensory modulation problems
- Sensory defensiveness
- Sensory dormancy/Sensory registration problems
- Adaptive movement response problems
- Vestibular processing problems
- Sensory discrimination and perceptual problems
- Tactile discrimination
- Proprioceptive perception
- Visual perceptual
- Other sensory problems
- Praxis problems

Occupational therapists work with children who have sensory processing disorders, by engaging them in activities that promote sensory integration. SI therapy in CP enables the child to make purposeful adaptive responses to sensory input in the environment. It refers to a therapeutic intervention which uses strong kinesthetic and proprioceptive stimulation to attempt to organize the CNS.

**Guiding principles in SI:**

- Sensory input can be used systematically to elicit an adaptive response
- Registration of meaningful sensory input is necessary before an adaptive response can be made

- An adaptive response contributes to development of sensory integration
- Better organization of adaptive responses enhances the child's general behavioral organization
- More mature and complex patterns of behavior are composed of consolidations of more primitive behaviors
- The more inner- directed a child's activities are the greater the potential of the activities for improving neural organization.

Intervention techniques should address the underlying sensory deficit and not the behavior. SI uses all senses but focuses primarily on the Vestibular, Proprioceptive and Tactile Senses together known as the power senses. A child's brain organizes sensory stimulation from touch and movement in order to learn and respond successfully to the environment. Vestibular stimulation includes linear, circular and rotatory movements like lying on the top of a large exercise ball while receiving rocking or bouncing movements and sitting inside an inner tube swing. Proprioceptive stimulation includes compression and traction like weight bearing activities like wheel barrow walking and jumping on trampoline and hanging from a trapeze. Tactile stimulation includes different textures stimulation like sand playing, brushing painting activities. Different types of stimuli influence a child's muscle tone and normalizing muscle tone enables a child to move and function well. Children with increased muscle tone typically benefit from calming sensory stimuli such as slow movement, rhythmic music, dim lights and a quiet environment. Children with low muscle tone typically benefit from erratic music, bright lights and irregular movement such as jumping.

Sensory Integration and Neurodevelopmental treatment can be merged to improve the quality of not only the motor control and skills but also the child's affect, emotional responses and ability to integrate and synthesize information from his body and his environment to make better adaptive responses. Using a holistic approach can facilitate not only the postural control, coordination and motor output, as well as improve the sensory processing, which is supposed to be the base for developing core behavioral and emotional regulation; develop the ability to regulate one's arousal and attention and improve motor planning needed to be effective in daily life. Both approaches however need specialized training which will help therapists stepping out of your comfort zone; learning new theoretical points of view, and acquiring new skills and training to benefit the child with cerebral palsy

## 3. Development of Hand Skills

Hand function involves the following components like reach, grasp, carry voluntary release, in hand manipulations, bilateral hand use. Often the CP child has impaired hand function skills due to problems in isolation of movements and tending to use total patterns, insufficient force application, improper timing of movements, limitations in trunk control and inability to bring hands in midline. Intervention aims at

- Proper positioning of the child. E.g. first consider the optimal position for eliciting the skills desired. Certain body

positions can be used to elicit specific hand skills like Supine position for arm movements and visual regard of hands, prone position with forearm weight bearing for shoulder stability, side lying position for unilateral movements. While sitting at table for fine motor tasks the child should be seated at an appropriate table height on a stable chair with foot supports, with arms on table surface without elevating shoulders.

• Improvement of postural tone and control E.g. (refer to development of postural control

above) and inhibition of tone using weight bearing or slow movement activities

• Development of hand skills by:-

Promoting isolated arm and hand movements e.g. stabilizing proximal muscles (trunk and shoulder) to promote opportunity for the isolated hand movements. Enhancement of reach, grasp, carry voluntary release, in hand manipulations, bilateral hand use Facilitation of neck flexion while going from sit to supine e.g. using pegs, beads or marbles of various sizes and shapes, clay activities, stringing and stacking activities

## 4. Development of Visual Perceptual Skills

Visual Perception is defined as the total process responsible for the reception (sensory functions) and cognition (specific mental functions).

**Visual receptive Functions**

- Acuity
- Accommodation
- Binocular fusion
- Steropsis- Binocular depth perception or three dimensional vision
- Convergence and divergence

**Visual Cognitive Functions**

- Visual Attention - alertness, selective attention, visual vigilance and divided or shared attention
- Visual memory- short term and long term
- Visual discrimination
- Object (form) perception- form constancy, visual closure and figure ground
- Spatial perception-position in space, depth perception and topographical orientation
- Visual Imagery

CP children with Visual Perceptual problems demonstrate many problems like in ADLs e.g. difficulty in combing, tying laces, matching clothes or in Play e.g. difficulty in sports, cutting, constructing, doing puzzles. Incorporating vision as an integral part of a therapy session will not only improve the child's orientation in space and spatial orientation but will also help with his or her ability to scan the environment while learning to move through space.

Intervention focuses on visual perceptual training involving Developmental, Neurophysiologic, Sensory Integration and Compensatory approaches which helps in improving skills that limit function and also compensate for the limitation.

## 5. Positioning and seating

Proper positioning can be achieved by use of handling techniques, pillows/ wedges, bracing, inserts etc to minimize the effects of abnormal tone and reduce secondary complications. Evidence supports that children with CP should be fitted for wheelchairs that place them in a functionally safe position, which includes; a hip belt, an AO, footrests, and a cut out tray, with the addition of a sloped forward seat to improve upper extremity function.

**Adaptive Devices**

- Nonslip surface on chair to prevent slipping (e.g. Dycem)
- Bolster, rolled towel, or blocks for feet
- Adapted or alternate chair, stander
- Custom fitted wheel chair or insert



Fig 87, 88, 89, 90, 91, 92, 93, 94, 95- Examples of different seating devices to promote proper alignment and symmetry



Fig. 96- Neck pillow for neck stability



Fig 98 Adapted toilet seat



Fig 97 Adaptive device to promote mobility

## 6. Activities of daily living (ADLs)

Activities of Daily Living involves taking care of one's body such as toileting, bowel and bladder management, bathing personal hygiene and grooming, eating and feeding, dressing, functional mobility sleep and rest. Approaches to improve Performance of ADL involve Establishing, restoration, maintenance and Modification or Adaptation and Prevention and education. Occupational therapists use many Behavioral techniques like chaining (backward and forward), shaping and reinforcements to teach ADL skills.

The following are the issues to be considered while applying the functional approach (Activities of Daily Living-ADL) in a combined intervention of child-centered activity and structural developmental intervention respectively:

1. Developing a feeling of interest in performing daily activities and motivation
2. Experiencing learned functions by using daily life devices
3. Practicing self-care skills
4. Developing visual-spatial skills within the environmental setting
5. Developing creative self-expression through play, artistic activity, movement and other activities
6. Developing perceptual and visual-motor functions which are necessary for learning
7. Developing more complex play levels

• **Feeding**

Evaluation should encompass the patient's visual, perceptual and cognitive skills, physical control of head, trunk and extremities, oral structures and ability to suck, masticate, and swallow. Also face and mouth sensitiveness, ability to sense temperature, facial muscles, interference of primitive reflexes like rooting, bite, sucking. Impaired oral reflexes like gag coughing reflexes, Outer oral motor assessment ( facial expression, lip control and jaw control) and inner oral motor assessments (palatal functioning, tongue musculature and motor control) are conducted. The family's desires and expectations in regard to the child's feeding capacities should also be considered. Intervention in feeding involves positioning, handling and compensatory strategies. Proper positioning should be emphasized while feeding to promote oral motor function.

**Appropriate positioning for feeding:**

- Neutral pelvic alignment of trunk. Pelvic alignment is facilitated when the childs well supported against a flat back, on a flat seat and square on the buttocks with hip and knee in 90° of flexion.
- Good head, neck and shoulder alignment in slight neck flexion or in neutral.
- Chin tuck with the back of the neck elongated
- Providing the child with external postural stability enhances stability, good alignment and easy feeding.

**After positioning the child, the therapist can use handling techniques to aid oral movements:**

- Tapping or quick stretch, vibration to increase tone
- Oral motor stimulation to improve oral motor control

**Adaptive devices**

- Non slip mats, wet towels, suction cups are used to stabilize eating utensils
- Adapted cutlery may be more suitable, e.g. rocker knife, extended handle cutlery (e.g. foam handle on utensil), Adaptive drinking devices (e.g. cup with cut out rim)
- Adapted chairs, tray attached to table





Fig 99, 100, 101- Adaptive devices

**Practical tips**

- Normalize child's tone as much as possible before beginning the feeding process
- Observe the child while he/she is eating.
- Make sure the child can see the plate, the food which is on it and the spoon bringing the food from the plate to her/ his mouth.
- Talk to her/him about the process and let her/ him see, feel and smell the food, feel the plate and the spoon
- The feeding person should be seated directly in front in order to maintain proper position
- Table should be positioned at axilla height and close to chest so distance from plate to mouth is reduced
- Chewing may need to be encouraged slowly and patiently by very gradually increasing the density of texture and later on the 'lumpiness' of the food offered.
- Deep and firm pressure to reduce tone
- The therapist can also use her/his hands and fingers to provide external support e.g. using one finger to promote chin tuck and the other under jaw for support from the side or from front
- Encourage her/him to pick up food (such as cakes, biscuits, bread or fruit) in her hands and bring it to her/his mouth.
- Use different food temperatures and textures to increase child's awareness of what is in his mouth.
- Firm pressure is more acceptable to the child than light touch
- Placing a mirror in front of the child during mealtimes helps his feeding skills.
- Give the child the drink he likes best allowing him to make a choice when you teach him to drink
- Therapist can guide the extremity in the correct pattern if required

## • Dressing

Learning and participation in dressing is a major step in step in achieving independence. Even with limited motor and sensory skills dressing can be made easy by motivating the child to actively participate and reducing demands placed on the child.

**Adaptive clothing**

- Loose clothing free from unnecessary fastenings which should be limited to few layers
- Use of Velcro and elastic band clothing instead of buttons or zippers
- Front openings, Pullovers, large buttons, Stretchy clothing




Fig 102- Examples of boards to practice different fine motor skills involved in dressing

**Practical tips**

- Work on undressing first, as it is an easier skill than dressing.
- Always put the clothes on the most affected part of the body first.
- Positioning child on floor is safer than on a chair.
- Techniques that inhibit spastic postures will facilitate the movements in and out of clothing, If her legs are bent before putting on socks and shoes it may help ease any stiffness in her ankles and feet and her toes are less likely to curl under.
- Encourage to stand up and hold on to furniture.

**• Bathing**

Achieving cleanliness is essential for maintaining good hygiene. Bathing can be made fun and a special bonding time with the caregiver. Occupational therapists can use bathing therapeutically to enhance motor and sensory skills.

**Adaptive devices**

**Hand held Faucet**

- Soap on a string or soap tied in a lofah or liquid soap
- Railing, grab bars
- Adaptive chairs
- Semi-inflated inner tube can be used as a positioning device
- Nonskid mats
- Hydraulic grab bars

**Practical tips**

- Positioning the child in a such way that normalizes his muscle tone as much as possible providing a sense of security, rather than challenge his sense of balance.
- Bath sponge wedge can be used to bathe an infant in both sitting and lying positions.
- Bathing can be time which can be used to teach various concepts for example, arm in, out, through: tub full, empty, wet, warm, cold and working on body parts identification. Thus bathing provides the child with many opportunities to learn cause-and-effect relationships, spatial relationships and tolerance of different sensations.
- The time following a bath can also be very productive. For example, a fast run with a terry towel can help the child learn to process sensations and can lead to better body awareness.

• Toileting

Independent toileting is very important in achieving self maintenance. Toilet training can be quite challenge in a CP child as the child must be temperamentally and physically ready to accept toilet training as well as be able to understand the process in order to have any success.

Adaptive devices

- Use of Toilet paper
- Hand held jet sprays
- Soap on a string or soap tied in a lofah or liquid soap
- Railing, grab bars

- Adaptive commodes or chair with hole placed over commode
- Footstools to support feet
- Use of straps to maintain safe and proper posture
- Nonskid mats

**Practical tips**

- A fairly casual, non confrontational introduction to the process will help reduce any stress related to toilet training.
- Visual break down of task on wall
- Habit training to go at same times to toilet
- Reinforce them for their success or for sitting patiently on the toilet and trying.
- Instruct caregivers to follow same routines at school

**Mobility**

Being mobile enhances a person's ability to learn, interact with others and participate in the community. For children with mobility impairments, a variety of mobility aids and devices are available to provide support, motion and access, as well as to enable them to lead active and fulfilling lives.

Mobility aids include:

- Canes
- Crutches
- Walkers
- Manual wheelchair
- Powered wheelchair with joystick, head switch, or sip/puff controls
- Grab rails and Railings

## 7. Play Skills

Play can be an important part of the learning experience and development of motor skills for a child with cerebral palsy. Selecting an activity should incorporate the child's interests and the skill he possess to participate in safe playing.

- Appropriate adapted equipment, such as wedges, bolsters, bean bags, CP chairs, may be used.
- Make certain your child changes positions frequently. Children should be encouraged to play on different surfaces and at safe heights
- Position child with both arms forward when playing with toys.
- Make certain that the child can see what is happening.
- Talk to the child at the child's eye level.
- Give the child ample time to respond
- Maintain a good balance between noisy, active play and quieter, less strenuous activities.
- Present toys that encourage your child to reach and grasp with the hand that is more difficult to use, but allow the child to use whichever hand he chooses.
- When teaching dressing to the child, put the more affected arm or leg into the clothing first.
- When interacting with your child, take the more affected hand.
- Encourage bilateral activities such as rolling clay or throwing a large ball,
- Provide multi sensory input toys that have interesting things to see, hear and feel.
- Avoid too noisy small play items

- Grade level of activities, gradually increasing its complexity
- A mirror can be a great aid in playing so that the child can get visual feedback
- Toys that have enlarged handles or knobs to grasp

**Age Appropriate toys used to enhance play behaviors**

0 -2 Years

- Play mat and frame with dangling toys
- Rocking and bouncing games
- Making lots of babbling and cooing noises
- Tickling games
- Building blocks
- Story books - with voice
- Peek a Boo games
- Banging on musical drums
- Imitation and turn-taking
- Playing with mirrors, press toys
- Unwrapping toys
- Lentils, rice and pasta in tubs to sit in, put your hands in or just throw about
-
- 1-3 Years
-
- Finger puppets
- Tunnel games
- Surprise bags full of toys and interesting objects for your child to find
- Pulling and pushing
- Object identification
- Pretend games with dollies and teddies
- Painting, using fingers or brushes
- Cars and trains
- Paper tearing and crumpling

**2-4 Years**

- Story books
- Clay modeling
- Ball games
- Obstacle courses
- Messy play (i.e. sand castle, finger painting)
- Sticking
- Stamping (leaves, crumpled paper prints etc.)
- Spot the difference
- Shape and color matching
- Make believe
- 'Simon says'-imitation games
- Imaginative Play e.g. Turning boxes into toys (such as castles, cars or space ships)
- Action rhymes
- Listening games
- Making music with home-made instruments
- Games involving "spotting the difference", or pointing out which part of a picture or drawing does not belong, help the child's neurological development.

## 8. Reflex integration

In healthy infants reflexes appear at ordered times. Some reflexes appear near or at birth and some appear at 2 months, 6 months etc. therefore, what is considered

"normal" in reflex evaluations depends on the age of the child being observed. Each reflex has a specific stimulus and consistent motor response that continues to be active for a definite period of time. The repetition of the same motor response gives the baby an opportunity to learn a movement to achieve voluntary movement.

"Integration" of a reflex indicates that a connection network has been made between different levels and areas of the brain. The reflex achieves its function and becomes inactive, becoming secondary to higher brain activity unless called upon in an emergency situation (e.g. brain injury, PTSD or stress). It is suspected that voluntary motor skills learned without the reflex foundation are not as strong and can be much more easily overwhelmed by stresses and environmental factors.

If a child is not demonstrating typical development due to atypical brain working or difficulties in learning, a reflex may present itself in a pathologically powerful way, overtaking control of the child's nervous system and replacing typical responses. In extreme cases, the pathological reflex responds to any stimulus.

The child with CP may have primitive reflexes that persist such as asymmetrical tonic neck reflex, symmetrical tonic neck reflex, tonic labyrinthine reflex, moro's , grasp and positive supporting reflex. Retained reflexes may cause muscle tone and movement difficulties in the limbs. Also there may be immaturation of the postural reactions like righting, protective and equilibrium reactions.

Different reflex integration programs like Masgutova Neurosensorimotor Reflex Integration (MNRI®), Quantum Reflex Integration (QRI), Rhythmic Movement Training (RMT), Institute for Neuro-Physiological Psychology (INPP) training etc are available and therapists can be furthered trained in them.

## 9. Hand writing Skills

Writing is a fine motor task required to compose stories, complete written examinations, copy numbers etc. It is a complex task which requires synthesis and integration of memory retrieval, organization, problem solving, and language and reading ability, ideation and graphomotor function.

The building blocks of early handwriting are:

- Spatial and body awareness
- Postural control
- Visual perception
- Fine motor control
- Directionality
- Handwriting

Intervention focuses on :-

*Neurodevelopmental approach:* postural and limb preparation activities like jumping on a trampoline, pushups, bear walks etc to modulate tone

*Biomechanical approach:* sitting posture should be with feet firmly planted on the floor, height of table such that it should be 2inches above flexed elbow, paper position should be slanted so that it is parallel to the forearm of the writing hand, pencil grip should ideally be a dynamic tripod grasp, pencil

with a wide diameter, various writing paper (lined, unlined, double lined, textured, margins).

*Sensorimotor approach:* involves controlling sensory input through selected activities to enhance sensory systems. An inclined, vertical or horizontal writing surface, writing tools like crayons, sketch pens, vibratory pens, and using different textures like sand, shaving cream, talcum powder

**Adaptive writing equipment**

Pencil Grippers and weighted pencils and handiwriters (soft elastic looped around the writing utensil and wrist that keeps the utensil in the appropriate space of the hand)

Adaptive writing paper that is color-coded, sticky or rough on one side or has larger lined area









Fig 103, 104, 105- examples of different grippers to assist in pencil grasp

## 10. Use of Assistive technology

The cerebral palsy assistive technology includes hardware and software which helps maximize their abilities to access information and services. It has the potential to increase abilities of person with disabilities and can lead to independent living.

- Typing Master Software
- Text to Speech Software
- Voice recognition computer applications
- On - Screen Keyboard setting, Magnifier setting, Narrator setting, Mouse Keys Utilization and setting.

The objective is to see how the technology can be used by a person with disabilities to enable him or her lead a more purposive life with some skills to do work and at the same time too have time for relaxing and enjoying life.

**Some Assistive Technology used in writing**

- Pencil or pen with adaptive grip
- Adapted paper (e.g. raised lines, highlighted lines, and so on)
- Slantboard
- Type writer
- Portable word processor
- Computer
- Text to Speech Software
- Voice recognition computer applications
- On - Screen Keyboard setting, Magnifier setting, Narrator setting, Mouse Keys Utilization and setting.

**Alternate Computer Access**

- Hand-held pointer (e.g., dowel, stylus, or pencil)
- Light pointer worn on head or held in hand
- Mouse or mouse emulator (e.g., HeadMaster Plus, HeadMouse®, trackball, track pad)
- Proportional or video pointing devices (e.g., joystick) that are controlled by the hand, chin, or tongue
- Keyguard
- Arm support (e.g. ergorest)
- Track ball, track pad, joystick with onscreen keyboard
- Alternate keyboard:- Expanded/ Miniature/ Light pointer/ Onscreen keyboards (e.g. Intellikeys, Discover Board, TASH, Head Master Plus, HeadMouse®, and Tracker 2000)
- Mouth stick or head pointer with standard or alternate keyboard
- Switch with scanning
- Voice recognition software
- Word prediction (e.g. Co:Writer) to reduce keystrokes

**Composing Written Material**

- Word cards, word book, or word wall
- Pocket dictionary or thesaurus
- Electronic or talking electronic dictionary, thesaurus, or spell checker (e.g. Franklin Bookman)
- Word processor with spelling and grammar checker
- Word processor with word prediction (e.g. Co:Writer) to facilitate spelling and sentence construction
- Talking word processor for multisensory typing
- Voice recognition software
- Multimedia software for expression of ideas (assignments)

**Communication**

- Augmentative and Alternative Communication (AAC)
  - Communication board or book with pictures, objects, letters, or words
  - Eye gaze board (Eye gaze communication)
  - Simple voice output device (e.g. Big Mack, Cheap Talk, Voice-in-a-Box, Micro Voice, Talking Picture Frame, or Hawk)
  - Device with speech synthesis for typing (e.g. Cannon Communicator, Link, Write:Out Loud with laptop computer)

**Academics**

**Reading**

- Changes in text size, spacing, color, or background color
- Use of pictures with text (e.g. Picture It, Writing with Symbols)
- Book adapted for page turning (e.g. page fluffers, 3-ring binder, cardboard in page protector)
- Talking electronic device to pronounce challenging words (e.g. Franklin Bookman)
- Scanner with talking word processor
- Electronic books
- Print or picture schedule
- Low tech aids to find materials (i.e., index tabs, color coded folders)
- Highlight text (e.g. markers, highlight tape, ruler)
- Software for manipulation of objects or concept development (e.g. Blocks in Motion, Toy Store). Consider alternate input device (e.g. switch or touch window)
- Software for organization of ideas and studying (e.g. Inspiration, Claris Works Outline, PowerPoint)
- Recorded material (books on tape, taped lectures with number coded index)
- Key guard to go over keyboard to help select the right keys and forearm supports to help stabilize upper extremity.

**Maths**

- Abacus or math line
- Calculator, with or without print out
- Talking calculator
- Calculator with large keys or large LCD print out
- On screen calculator
- Software with templates for math computation (consider adapted input methods)
- Tactile or voice output measuring devices (e.g.
- clock, ruler)

**Recreation and leisure**

- Adapted toys and games (e.g. toy with adaptive handle)
- Use of battery interrupter and switch to operate a toy
- Adaptive sporting equipment (e.g. lighted or bell ball, Velcro mitt)
- Universal cuff to hold crayons, markers, or paint brush
- Modified utensils (e.g. rollers, stampers, scissors)
- Ergo Rest to support arm for drawing or painting
- Drawing or graphic program on computer (e.g. Kid Pix, Blocks in Motion)
- Playing games on the computer
- Music software on computer

**Environmental control**

- Light switch extension
- Use of Powerlink and switch to turn on electrical appliances (e.g. radio, fan, blender, and so on)

- Radio or ultrasound remote controlled appliances

**Vision**

- Eye glasses
- Magnifier
- Large print books
- Screen magnifier (mounted over screen)
- Screen color contrast (e.g. Close-View)
- Screen magnification software (e.g. Closeview, Zoom Text)
- Screen reader (e.g. OutSpoken, Jaws)
- Braille Keyboard and Note taker (e.g. Braille N Speak)
- Braille Translation Software

**Hearing:**

- Hearing aid
- Classroom amplification
- Captioning
- Signaling device (e.g. vibrating pager)
- Screen flash for alert signals on computer

## 11. Environment modifications

To ensure safe mobility and increase independence of the CP child, the physical environment in which he functions may need modifications in the form of:

- *Accessible Entrance/Exit* - includes adding ramps, widening doorways, making entrance locks and door handles accessible and providing an emergency exit, considering lightweight doors or automatic door openers
- *Accessible Interior* - includes widening hallways or interior doors, moving electrical switches and outlets related to door widening, reinforced wall for grab bars, railings
- *Accessible Bedroom* - including widening doorways, making the closet accessible (lowered shelves and hanging rods), relocating electrical switches and outlets
- includes modifying design of commode, sink *Accessible Bathroom* - and cabinets, tub or shower, widening entrance, moving switches and outlets, faucet hardware
- *Other modifications*- includes reinforced ceiling if need a lift, roll under sink in kitchen and bath, assigning workspace in close proximity to school supplies and equipment, modifying workspace or desk design and height

## 12. Splinting

The Occupational Therapist evaluates and recommends use of Upper extremity splinting to improve, maintain and prevent contractures and deformities or to improve functional movements. Hand splints to improve thumb abduction, wrist extension and functional positioning

of digits are generally prescribed. E.g. resting pan splint is one which keeps the wrist in 20° to 30° extension, the metacarpophalangeal joints in 60° flexion and the interphalangeal joints in extension. This type of splint is used at night and during periods of inactivity with the hope of preventing deformity. An example of a functional splint is an opponens splint to bring the thumb out of the palm of the hand, allowing for better grasp. This type of splint is used in everyday activities. A thumb abduction splint may improve use and manual function of the affected hand in children with hemiplegia.



Fig 106- Splint for thumb abduction

**Precautions during splint use**

The caregivers should be educated about the wearing of the splints which includes donning and doffing of the splint, wearing schedule and care of skin and splint. Non verbal children having poor sensations may not be able to report sensory problems occurring during wearing of the splint, so a thorough skin inspection should be taught to the caregivers Starting with wearing of the splint for few minutes the wearing time should be gradually be increased to about 8 hours a day for static splints. Use of dynamic splints can be increased for additional more hours according to the child's tolerance. Hand orthoses may inhibit the active use of the extremity. Hence it is important for the child to spend a certain amount of time in between the wearing schedule without the splints. Gentle range-of-motion exercises to be done on removal of splints

## 13. OT intervention in school for CP children

OT in school focuses on child's ability to participate in functional school activities. A problem solving approach is used in identify the difficulties the child faces and to identify intervention strategies. Intellectual disabilities and learning disabilities are some of the problems which the CP child may have to face. Parents may require counseling regarding the type of school which best suits the child. The Occupational therapist is one of the members of a child's IEP (Individualized Education Program) team.

Strategies used in the school:-

- Reframe teacher's perspective. e.g. by explaining the issues and the underlying deficit the child seems to be facing
- Improve child's skills. e.g. use of practice worksheets
- Adapt the task e.g. use of keyboard to take down notes
- Adapt the environment. e.g. keeping visual distractions to a minimum
- Adapt the routine. e.g. extra time to complete worksheets.

**Adaptive aids**

- Adaptive chair (CP chair)
- Adaptive writing devices, aids and paper
- Railings along staircase, classrooms and toilets
- Adaptive commodes
- Schedule boards, checklists timers, calendars

## 14. Vocational and prevocational Rehabilitation:

Vocational and prevocational activities are ones that help to prepare students for a future job they may pursue as they get older.

*Pre-Vocational skills training:*

Activities are developed that will prepare an individual for employment. It includes support and training in behaviors related to following directions, attending to task, task completion, problem solving, and safety and assisting the person to adjust to the productive and social relationship demands of a work place.

In the classroom it is important to give children a chance to experience what sorts of tasks they may encounter in the workplace. Some examples of these tasks are: sorting, putting objects together, alphabetizing, filing, data entry, and packaging items.

*Vocational skills training:*

Prior to vocational skills training a Transition Programme can be conducted, in which the skills learnt already in the pre vocational training are transferred to vocational training. Occupational therapists specialized in Vocational rehabilitation, conduct workshops to give training in a variety of occupations like tailoring, greeting card making and pot painting exercise, soap and phenyl, pickle, card making, craft works etc.

**Management of associated problems in CP**

1) **Seizures Management:** Many children with cerebral palsy experience seizures.

Practical Things to remember in event of any child having a seizure are:

- Do not attempt to hold the child still or to prevent physical movement; instead, make the environment safe so that she cannot be physically hurt.
- If the child is in a sitting position, or standing or walking when you observe the seizure, help her to lie down, so that she will not fall and get hurt. If at all possible, place the child on a blanket or protected surface. Position the child on her side, supporting the head. Move all sharp objects out of the way, as well as any furniture. Pad any sharp objects that cannot be moved, to prevent the child from getting hurt. Loosen clothing, especially in the chest and abdominal area.
- Do not put anything in the child's mouth. Do not interfere with the seizure or try to stop it.
- Allow the seizure to continue without interruption. Check the child for breathing. If the child has stopped breathing, clear the airway and perform mouth-to-mouth breathing.

- Turn the child on her side so the saliva can flow out of her mouth.
- After the seizure has run its course, let the child rest and be supportive.
- Although, medications are very effective in preventing or reducing seizures when given regularly, they may also produce a variety of side effects. For example, they may cause hyperactivity behavior, irritability, sleep problems, lethargy, depression, or sedation which may affect therapy.

**2) Sexuality issues**

Adolescents with cerebral palsy have delayed and prolonged puberty. They may develop precocious puberty as well. The therapist can guide the parents in preparing the child about puberty and the bodily and behavioral changes he may experience through books or pictures. Try and recognize the timing of sexual maturation and educating parents to provide age-appropriate sexual education. Pose questions about sexuality privately, using normalizing statements and open-ended questions helps the adolescent open up to discussions.

**Speech therapy Management in CP**

Having a child with severe developmental disabilities which are long-standing can be a severe challenge to parents whose entire lives are transformed into being caregivers, proxy clinicians and watchful nurses tending to their children. Their lives are half lived in hospitals, nursing homes and doctors' clinics for the many medical ailments suffered by their children and in the therapy clinics of rehabilitation professionals which they need to visit regularly for consultations and therapy. The tremendous strain -physical, mental, emotional, financial - on them is consistent and unrelenting!

In spite of all this, their courage, commitment and endless striving for the betterment of their children is exemplary and commendable! Parents are all the while searching for any course, any new treatment modality being researched, any bit of new information that comes their way which might help their child with Cerebral Palsy and they try their level best to make it available for their child. In a country like India where, the available resources for children with disabilities are very few and more likely to be in metropolitan cities and larger towns, it is the need of the hour to have ready and easily applicable clinical literature at hand, so that the caregivers of these children may work towards enabling their children towards better health and functioning. This may help them to reduce the participation barriers of their children and enhance their functioning in their respective environments.

Parental/ caregivers' role in the management is paramount and the most important determiner of the success of the program as it is only with their support and carry- over of the goals and activities into extra-clinical settings, that any therapy will prove to be effective. They need to be active participants in the therapy program, should be given extensive demonstration sessions and detailed home programs. This chapter is meant to be a 'ready reckoner' for caregivers of children with Cerebral Palsy to help them deal with the manifold problems dealing with hearing, feeding and communication. The brief overview of the problems faced by these children has been presented earlier and in this

chapter easy -to -apply strategies, clinical tips and recommendations are presented.

The Audiologist and Speech Language Pathologist (ASLP) is responsible for the effective assessment, diagnosis and management of the disorders related to swallowing and feeding, hearing and communication, affecting children with CP. Management by an SLP will vary according to the age of onset of the pathology and age at which the child reports to the SLP, affected domains, severity, pathophysiology, and associated problems. An important guideline is that for the assessment of deficits, the child's chronological age needs to be considered and for treatment planning, his developmental age. It then becomes clear to the caregiver as to at what level the child is, at present and what is the lag between present and the level he needs to achieve within the constraints of his impairment. In all these endeavors, it is important that neither the caregivers nor the professionals lose the sight of the child as a whole. When all the team members and the parents work in coordination, it will result in faster and better progress in all the affected domains of the child's development. The purpose of this endeavor is not to replace sessions with the rehabilitation professionals, but provide a support to caregivers of children with CP, where such help is not available, or until such sessions can be arranged. It is hoped that this venture is found useful and practical for them. The following section deals with the management of primary disorders- breathing disorders (from the SLP's perspective); swallowing/ feeding disorders; hearing disorders- and secondary disorders overlaid on the primary ones- communication and speech.

## MANAGEMENT OF BREATHING DISORDERS FOR IMPROVING VEGETATIVE BREATHING

Relaxation and Breathing Exercises

These are meant to relax the muscle tone and reduce rigidity of the respiratory musculature and are to be primarily done by the Physiotherapist. It is essential that these need to be also done by the SLP in consultation with the PTist owing to its relevance in swallowing and speech production. The exercises need to be demonstrated in a clinical set up and then to be implemented in the daily regimen by caregivers. Since PTists trained in Neurodevelopment Treatment (NDT) principles and treatment can help a great deal in this regard, this exercise regimen has to be planned in coordination with the physiotherapist trained in NDT. NDT is the best treatment approach for enhancing neuromuscular control and breathing capacity which is imperative for the child's general health and also for his later speech development.

Some exercises that may be of help.

1. For enhancing Inhalation and Exhalation:- The caregiver makes the child lie on his back (supine position) and just places his/her palm for a few moments on the child's abdomen without any overt pressure. This seemingly simplistic positioning actually facilitates the breathing and steadily you see the abdomen rise due to the inspiration occurring. The caregiver can then flex the child's legs at the knees and then press his bent legs towards his abdomen, thus aiding in expiration. This sequence could be repeated 5-6 times.

2. For reducing rigidity and hypertonicity of ribcage and respiratory muscles:-

   The child is made to lie on his stomach (prone position) and taking the child's spine from the nape of the neck as the reference position, the caregiver supports the child's trunk with one hand and with the side of palm placed on the outer side of shoulder of opposite side, exerts deep pressure and moves in a transverse position towards the spine. Then the same is done on the other side and ending the transverse movement a few cm below on the spine in an almost V shape. This sequence is repeated until the lower end of the ribcage. Then again start the whole exercise. This is done 5-10 times each time.

   Pics showing exercise to reduce hypertonicity of ribcage

3. For relaxing the ribcage:- The child is placed in the prone position and the caregiver with the right hand held taut with fingers together and thumb at right angles, places the hand on the child's left side just below the armpit, with the thumb below and fingers above and held taut thus, with the contact region on the child's body being the space between the thumb and fingers, the caregiver exerts deep pressure and as swiftly as possible moves it downwards in a vibratory mode till the base of the ribcage, then again similarly 5 times and then for another 5 times on the right side.

   These exercises should be done 5-6 times each time and about 4 times each day along with the others for speech facilitation. These are very helpful to relax the respiratory musculature and ribcage which improves the respiration and thus has a positive impact on speech as well.

## MANAGEMENT OF HEARING IMPAIRMENT (HI)

It is a challenge to work with a child with multiple impairments- CP and HI- and treatment of HI needs to be planned in consultation with the ENT surgeon and the ASLP. Referral to the ENT surgeon is imperative for treatment of conditions that can be medically or surgically treated. Once the best of those options have been tried and the child has irreversible sensorineural hearing loss, then the Audiologist assesses the nature and degree of HI through a battery of audiological tests, such as:-

- Behavioral Observation Audiometry (BOA) which gives an idea of the functioning of the better ear with the help of sounds presented from sound makers, ranging across different frequencies and intensities, and speech sounds presented at varying distances from child. Physiological responses such as eye blink, changes in breathing, startle responses are considered as responses to sound stimuli when these occur immediately after sound presentation. BOA is particularly helpful in these children.
- Visual Response Audiometry (VRA) wherein the child is conditioned

to look at a visual stimulus on the presentation of the sound stimulus in a free field setting.

- This may be difficult in a child with CP who may not be able to localize his head towards sound source due to poor neck control, but if eye tracking is present, that could be considered to judge a child's response.

- Pure Tone Audiometry (PTA) which measures the behavioral thresholds of hearing sensitivity in the two ears and the type of hearing loss and is a standard test of choice

- Electrophysiological testing such as Brainstem Evoked Response Audiometry (BERA), which is an objective test to obtain the hearing thresholds to click and tone burst stimuli, and is considered a test of choice in children with CP as the child's active participation is not required. BERA can be done in natural sleep or under sedation .

- Impedance Audiometry or Immittance Audiometry is used which tests middle ear functioning and stapedial refex which can help in diagnosing middle ear conditions such as middle ear fluid, ossicular chain fixation/discontinuity etc. It also helps to estimate degree of hearing loss based on stapedial reflex threshold and helps in differential diagnosis between cochlear and retrocochlear site of lesion.

- Other tests such as Speech Audiometry, Otoacoustic Emissions (OAE), also may be used.

Based on the type and degree of the hearing loss, the child may be fitted with suitable amplification (Hearing Aid fitting) in case of sensorineural and mixed type of hearing impairments. In a child who has HI, fitting with suitable hearing aids can be time consuming and challenging. This just gets all the more challenging when there are multiple impairments that is, CP and HI and even more so if such a child is also having cognitive impairment as may be seen in children with post- Rubella syndrome. It is crucial that the child is fitted with a hearing aid most suited to his HI for best acceptance. If the hearing status is not a true estimate, there is a risk that the child may be fitted with a hearing aid too powerful which the child would understandably reject, and if it is less powerful it not meet his needs. Hence it is after many sessions and painstaking efforts of an Audiologist that hearing aid fitting may be successfully completed. It is necessary that Hearing Aid trials are done with the child's customized earmoulds. Behind-the-ear (BTE) type hearing aids may be mostly considered as best suited for these children.

In recent times, there is the additional option of cochlear implant (CI) surgery. It is a very important role of an audiologist to do a pre-CI evaluation and determine candidacy for CI surgery. Only when the hearing loss indicates a cochlear loss of a permanent nature and there is not a sufficient degree of improvement with amplification tried with at least 6 months of Hearing Aid fitting, is this surgery considered. After fitting with suitable hearing aids or with CI, the Audiologist & SLP begins with rehabilitation and initiates intensive speech and language therapy.

Audiological testing and rehabilitation is a very specialized domain and is beyond the scope of this chapter to give more

information here. Caregivers are advised to consult an Audiologist for details related to the above mentioned domains.

## MANAGEMENT OF FEEDING AND SWALLOWING DISORDERS

Oral motor evaluation of the pre speaking child can be best achieved through modified feeding. Love, Hagerman and Tiami (1980) have presented an experimental clinical dysphagia scale wherein 5 feeding tasks are used for assessment:

- Biting
- Sucking
- Swallowing
- Chewing soft food
- Chewing hard food

The children are observed while having assorted foods such as spoon foods, chewables and drinks. Their positioning and any use of adaptive equipment are also observed. Based on the deficits seen, the SLP as the swallowing therapist can plan the therapy.

Swallowing therapy is indicated when the actual stages of swallowing, the triggering of swallow reflex and the oral and pharyngeal transit time is worked upon, whilst at all times focusing upon airway safety and ensuring safe swallowing. Hence, this requires the active involvement of the SLP in the capacity of a swallowing therapist. The first priority is to focus on ensuring airway safety. Once safe swallow is ensured, the nutritional and hydrational requirements are considered for which, intensive swallowing therapy is necessitated.

## THERAPY TO IMPROVE FEEDING

Attempts to improve feeding will include the caregiver helping the child with CP with positioning of food in the mouth, manipulating food in the mouth with tongue, chewing bolus of varying consistencies, re-collecting bolus into a cohesive mass prior to initiation of oral stage of swallow and organizing lingual action to propel the bolus posteriorly. (Arvedson,2002)

Pic of child placing bolus posteriorly to facilitate chewing

See picture 4 shows some adaptive feeding equipment and 5 shows a child with oral stage dysphagia being fed using a head back posture to position soft food.

The approach using 5 T.'s as suggested by Arvedson (2002) is especially relevant in this context:-

- Taste:- A variety of foods ranging in sweet, salty, sour and a little spicy has to be introduced one by one over time
- Textures of foodstuffs of varying consistencies need to be graded from soft solid to semisolid, to thick liquid to thin liquid in order of increasing difficulty for child
- Temperature :- Cold temperature is known to reduce spasticity and help in mobility of tongue for swallowing. More details given where PNF is explained.
- Total intake:- The child with CP, by and large is a poor eater and the total volume of food per meal has to increase gradually. For example, a child has half a katori of khichdi which needs

to increase over time as suited to his nutritional requirements.

- Time :- The time taken to swallow a bolus as well as the time taken for having the whole meal is measured and noted per meal, in a small notebook or diary and attempt is made to reduce these times with the swallowing therapy. Reduction of time can be used to measure swallow therapy outcomes.

- All the above points need to be carefully monitored by maintaining a diet chart tabulating the quantity and nature of intake, and response of the child during feeding.

- Inclusion of a Dietician and an Occupational Therapist in the team along with the SLP /Swallowing therapist is very important.

Presented below are some specific techniques that may be used for improving swallowing / feeding in children with CP and which help to lay the ground for speech production.

Exercises to prepare the oropharyngeal musculature for swallowing & speech production :- (Compiled by Sanghi,(2007), drawn from basic inputs Crickmay, ….and adapted with some additions based from clinical experience, used for clinical teaching but hitherto, unpublished)

A) ***For normalization of muscle tone-***

- Put the child in an appropriate posture in consultation with the Physiotherapist. These postures or positions are conducive to speech and likely to help the child with CP to be most relaxed

B) ***To facilitate independent movements of head, shoulders & neck-***

- Put child on a cot on his back, arms down by side hips flexed, put your arms under his spine and encourage him to let his head fall back over your arms in loose and relaxed way. Flex the shoulder forward and let neck extend.

- Place the child in a supine position (lying on back) with legs hanging over end of bed, and extend his shoulder backward and head flexed forward.

- Place the child in a supine position, passively move first shoulder, then other independent of each other. With child in same position, tilt/ move the child's head first to one side and then to other.

- Hold child's shoulders back and encourage making small independent movements of head.

C) ***To desensitize oral musculature***

- Hold child firmly in a comfortable posture and gently touch and move

  hypersensitive parts starting from periphery , that is upper part of face, then cheeks gradually moving towards mouth.

  i) PIC Light pressure on sub-mandibular region and thyroid region to facilitate swallowing

As a rule of thumb, if a reflex is absent for the particular age, stimulate. If reflex is persisting beyond a certain age, inhibit.

D) ***To stimulate age appropriate feeding reflexes in infants and older children.***

i) To stimulate Rooting reflex

- Stroking outwards firmly from the corner of the mouth along the cheek

ii) To stimulate tongue groove and suckling in an infant

- Stroking the lower lip and tongue tip in and out of the mouth
- Firm tapping of the corners of the mouth

iii) To stimulate *sucking reflex*

- A medicine dropper can be filled with the drink the child likes. One or two drops can be put into the oral cavity and the lips and tongue can be stroked. If done frequently, the child may develop a weak-sucking behavior

*Later straw can be used.*

i) To stimulate *biting reflex*

- Touching the teeth of the child and with the other hand encouraging rhythmic up and down movements of the jaw

ii) To stimulate *chewing reflex*

- Facilitate tongue movement manually without and then with resistance
- Placing food, such as a wedge of Parle G biscuit between the side teeth or gum ridges. Consistency safe for child to be chosen.
- Massaging the cheek from outside when food has been placed in the cheek pouch
- Important to introduce solid foods in his daily diet to give him more and more of chewing of harder solids in a graded manner, initially starting with soft foods like boiled potato, boiled egg, banana, then moving towards peeled apple, partially cooked carrot sticks.

Note: When swallowing reflex is sluggish, discoordinated or absent, it is imperative to consult an SLP/ swallowing therapist to guide the caregiver about a safe swallow. Choking on food may be a complication if oral feeds are tried by caregivers prior to ensuring safe swallow, hence extreme precautions are to be taken

E) ***To inhibit primitive reflexes in infants and older children.***

i) To inhibit tongue thrust

- Placing a spoon/finger on the middle of the tongue and making downward vibratory movements a number of times
- Activities for jaw stability given in the earlier section

ii) To desensitize hyperactive gag

- Achieving chin-tuck posture quickly and leaning him forward. Maintaining the posture till the child deals with the bolus placed in mouth that caused the gag

iii) To inhibit suckling reflex

- Touching his lips with finger or straw and with other hand prevent them from moving into sucking position particularly when child is thirsty

**iv) To *inhibit biting reflex***

- Gently but firmly holding the child's mouth closed while at the same time stimulating reflex either by bringing his/ her fingers close to his mouth or by touching his teeth

**v) To *inhibit jaw clenching***

- Encouraging child to put his fingers in his mouth which encourages licking, sucking and relaxes mouth and jaw

**F) Teaching normal mouth positions**

- Help child to align his teeth and close his mouth when jaw position is normal
- Place one hand gently under chin and hold in this position
- With other hand stroke cheeks and lips for relaxed expression of face
- Gradually release pressure under chin by removing hand
- Child takes control over himself for few seconds/ minutes
- New sensation of closed mouth- done repeatedly brings about longer periods of control

**G) To improve lip seal and reduce drooling**

- Facilitation of lip closure by bringing the upper lip and lower lip together by holding them between your two fingers. To be done for a few seconds but to be done as frequently as possible.
- Iced candies can be used. It relaxes spastic muscle and activates flaccid muscle. It improves lip closure and tongue movements. Encouraging use of mirror for imitation. When chilled foods are to be avoided, lollipops can be used.

**H) To achieve jaw and lip closure. These also help in preventing tongue thrust and facilitate swallowing.**

- Clinician / caregiver sits facing the child and holding the child's jaw below the lower lip with the thumb and supporting the lower part of the jaw with the index finger on the cheek and remaining fingers below the jaw.
- Clinician / caregiver stands behind the child and holding the jaw with the index finger placed below the lower lip and thumb on the TM joint.

**i) To enhance oral sensory awareness**

To desensitize hypersensitivity of face and mouth – Increase stroking gently,

starting at the face. With a cotton face -towel covering your fingers, stroke gently from the base of child's earline towards lips, below right ear towards right side of lips, and below left ear towards left side of lips together in a clean sweeping movement. Do it around 5 times. This helps to relax the child's facial muscles and may reduce spasticity.

Proprioceptive neuromuscular Facilitation :Stimulation in form of Icing and Brushing is done to increase oral sensory awareness and stimulate movement.

Icing can be done five seconds each, using ice cubes kept in sterile surgical glove or using iced cotton wool buds. Effects of icing are immediate. Two methods of icing may be doneSlow icing –to reduce spasticity. Fast icing –to stimulate muscle tone

Brushing –using "Florite"brush ( infant gum and tongue cleaner) dipped in honey and lime solution to add taste and to serve as a lubricant to stroke along the appropriate muscle such as tongue, prior to the required activity. Useful to stimulate flaccid muscles. Time lag of 20-30 minutes is expected between brushing and getting optimal results

Tapping – Light but firm tapping needs to be done on the cheeks from the earline to the lips, from either side. Then this process is to be done around the lips in a circular fashion and then inside the mouth, on the anterior surface of the tongue. At each level, it should be done for 2-3 minutes each.

Initially, a demonstration needs to be given by an SLP and under her/his guidance, the caregiver needs to be trained who then needs to follow a regular regimen of doing this 10 mins each time spread over 5-6 times each day. Intraoral PNF should be avoided after meals as they may trigger nausea and vomiting. Some of these sessions of PNF may be done prior to mealtimes as this stimulation may facilitate swallowing/ feeding initially and later result in better speech development too.

**J) To strengthen oral musculature**

i) This can be demonstrated by asking children to imitate caregiver and do lateral movements and protrusion and retraction, elevation and depression of the tongue when feasible and then against resistance.

ii) When tongue is too rigid, or too weak, the caregiver can gently hold it with sterile gauze piece and move in the required directions.

iii) A piece of chocolate or jaggery can be stuck on the upper lip or alveolar ridge (region behind the upper teeth). The child will elevate tongue in his attempts to lick it.

iv) Similar with lateral stretching of lips, pursing them and rounding them and later the same to be done against resistance.

v) Jaw movements up/down and lateral movements to be done and then against resistance from caregiver. Exercises for jaw mobility have been explained in H) above.

vi) Blowing exercises by puffing cheeks and holding pressure on a count of 1- 3 or 1-5, or by blowing candle or blowing off pieces of paper, can be demonstrated which help in improving lip seal, increase intraoral pressure and reduce drooling. This further improves swallowing and later help in building intraoral pressure as required for speech.

**K) Some general guidelines during feeding and drinking**

i) Approximately 15-20 minutes before meal time, use pressure stimulation of the face, around the lips, stretching the top and bottom lip from mid-line outwards and stroking downwards on the top lip and upwards on the bottom lip with fingers or fluorite brush.

ii) Present small mouthfuls, slowly and rhythmically.

iii) Feeding with the spoon should be done horizontally without angling the spoon upwards. Assist him in drawing the food from the bowl of the spoon by manually facilitating jaw and lip closure as discussed earlier. Alternatively, transferring the spoon horizontally between the side teeth and scraping there.

iv) Do not tip the child's head back when giving drinks. Use a cup with a cut out section to prevent the need to do so.

v) Adaptive feeding equipment may be used at home to assist in feeding.

PIC-5 Adaptive feeding equipment

vii) Initially feed to the preferred side in the child's mouth or the side where he shows most reaction to food.

## MANAGEMENT OF COMMUNICATION DISORDERS IN A NON- VERBAL/ MINIMALLY VERBAL CHILD

In assessing communication, it is imperative for the ASLP to do detailed audiological evaluations to rule out hearing impairment and if confirmed, to do the necessary intervention. More about this is given in the section on management of hearing disorders.

With the help of a psychologist, the child's cognitive status needs to be understood and performance IQ or SQ needs to be calculated. These evaluations are imperative for the SLP to assess communication and language development of the children with CP.

Non-verbal communication can be assessed from gestures, facial expressions and vocalisations in response to age-appropriate questions asked, and verbal communication comprising of speech and language needs to be assessed using formal and informal assessment procedures. There are assessment protocols which are designed for nonverbal children like the Picture Test of Receptive Language by Kaul (1999) which tests receptive language uptil 4 years of age. In case of non-verbal children, where it is difficult to gauge their comprehension formally, or as an adjunct to formal methods, an attempt may be made to do so by informal methods.

This could be done by attempting to understand their comprehension of humour by telling them funny stories and jokes appropriate to their chronological/ developmental age, and observing whether the child smiles or laughs at the appropriate junctures. The SLP or caregivers could also observe the expression in the child's eyes which may change with the content of speech. This may prove to be an important indicator of verbal comprehension. An effective way to train parents / caregivers can be to demonstrate language therapy sessions by an experienced SLP and train them to emulate the clinician in extra clinical settings and thus be proxy clinicians. Herewith is presented some tips as to language and speech stimulation by caregivers.

**Intensive language stimulation**

- Use chid-directed speech- While speaking to the child with CP, the caregiver needs to use short phrases, constant repetition, using varied intonation patterns, recasting utterances.
- For example, 'Who has come? Papa has come. Rohan's Papa has come. What has Papa got for Rohan? Did you ask Papa what he got for you?'
- Self Talk- The caregiver tries to capitalise the child's attention on him/ her, by speaking about the activity that the caregiver is engaged in, at that moment.
- For example, ' Do you know what I am doing? I am making food for you' or 'See what I am doing. I am building this tower...Would you like to topple it down?'
- Parallel Talk- The care-giver speaks about the activity that the child is engaged in, at the moment.
- For example, ' Oh, you are going out with Dadaji! So you have worn your shoes and taken your bag. Where are you going?'
- Labelling objects around the house- Each time the child is using objects, the caregiver should name them, thus familiarising the child with the labels. For example, ' This is Rohan's toothbrush.' 'Let's take Rohan's plate and sipper and keep it ready.'
- Use of gestures- The caregiver needs to speak using a lot of gestures so that the child begins to associate the spoken words with the gesture. Care needs to be taken to speak first while engaging the child's attention and then immediately after to use the gesture so that the child gets to process speech first and the gesture is used later to facilitate his comprehension.

## ALTERNATIVE AND AUGMENTATIVE COMMUNICATION APPROACH

Once speech and language stimulation has been tried intensively for a sufficiently

long period and the child still does not show relative progress, then the SLP needs to work towards using use of low tech or high tech devices and strategies to further augment speech efforts or switch to a non-verbal approach or an alternative approach.

Thus, for the child who is severely disabled, augmentative and alternative communication (AAC) modes (low tech) may be used such as customised communication boards/ books. Caregivers could click pictures of the child's routine such as the child eating, dressing, playing etc as also the objects involved in his activities of daily living such as his clothes, shoes, plate, cup, bag, special chair etc. Pictures of child and his close family members could be taken too. These pictures could then be printed and laminated to an appropriate size and used to compile communication books for different situations such as for mealtimes, schooltime, playtime, etc. These communication books could be kelp at hand so that the child could communicate his needs in the situation he is in, with the help of that respective communication book.

There are assistive devices which have pre-recorded messages and can be operated by a thump or customised to the motor response feasible for the child. The child could be taught Bliss symbolics

wherein the child is taught the system and expresses through pointing with hand/ finger or head pointer. Presently, there are many high tech devices available which are computer aided wherein the child is taught to type on keyboard or operate customised light touch switches or any other means suited to child's motor abilities. There are text-to speech options wherein the child may type words but that are synthesized into speech output.

## MANAGEMENT OF BREATHING DISORDERS FOR IMPROVING SPEECH BREATHING

The exercises which enhance breathing in general also help in improving speech breathing and should be continued for this purpose as well. Some other exercises may help in this aspect too and are given below:-

1. **Blowing exercises :-** The child should be encouraged to blow a lit candle from a few inches from child's face and then to do so with gradually increasing distance.
2. The same can be done with blowing off bits of paper and the caregiver could increase the level of difficulty by increasing the distance of paper bits from child or increase thickness of paper bits. Another variation of the same function to maintain child's interest is to blow bubbles and the caregiver could compete with child wherein he/she could demonstrate what is expected from the child by blowing for a longer time thus shown by the increased number of bubbles and the child is motivated to imitate that model.
3. **Free Phonation :-** The caregiver should encourage the child to say 'a.....' for as long as he can and keep count of how many seconds the child could phonate. The child should be encouraged for his effort and further motivated to continue longer if he can. His efforts need to be reinforced with a reinforcer such as a sweet, smile or a hug or verbal praise which will motivate him to try harder.
4. **Counting :-** The caregiver could make a verbal child count whilst maintaining his intelligibility and the same level of loudness and pitch. So if initially he can do so till a count of 5, the child is encouraged to continue till 7 in the next effort and then till 10 and so on. The child could say days of the week and months of the year too in a similar manner and the word count is documented.
5. **Increasing length of utterance:-** In case of the verbal child, the caregiver should encourage the child to have an increased length of utterance while maintaining the same quality of speech in terms of loudness and pitch.
6. **Biofeedback:-** Whenever feasible, the child could be given biofeedback in terms of loudness and pitch characteristics such as with a recorded sample played back to child wherein the child is made to identify his fluctuations in voice.

## MANAGEMENT OF VERBAL COMMUNICATION/SPEECH DISORDER:

The objectives of therapy are to familiarize the caregivers to the concepts of intelligibility, naturalness of speech and comprehensibility.

Intelligibility means that the child's speech has to be clear enough for it to be

understood by his listeners. Once that is achieved, the SLP needs to focus on the naturalness of child's speech which may sound strained, strangled, soft or too loud, hypernasal or with misarticulations and distortions in speech. The SLP needs to ensure that his speech sounds as natural as possible within the constraints of his impairment, in terms of the respiratory effort, voice, resonance, articulation and prosody (rate, stress, intonation).

Another focus is comprehensibility. This means that the onus for understanding does not lie on the child alone but also on the listener to understand as best as he/ she can from the child's speech attempts, by training his own listening and guessing skills as to content of the child's speech even if it is not very intelligible. The caregiver could also improve the medium of communication such as by going closer to the child, or by reducing ambient noise in the room by turning off the TV while the child is speaking or encouraging the child to use an augmentative device such as a microphone to amplify his speech in case of limited loudness of voice. The listener could also give the child feedback about message being unintelligible and ask related questions to facilitate understanding.

**To improve voice quality/reduce strained vocalizations**

- Gently massage the laryngeal area.
- If possible use blowing instruments to produce sound,
- Use appropriate posture to facilitate voice production.
- Encourage the child to laugh. Gentle stroking or tickling over the ribs will produce reflex laughing.
- Imitate child's vocalizations and encourage him to continue.
- Record child's vocalizations and play them back to reinforce his sound making efforts.

Encourage babbling- The child can be encouraged to repeat Consonant Vowel (CV) sequences after the caregiver for example, 'Ba ba ba ba' or 'Da da da da'. This may help to facilitate babbling which is a milestone in speech development a child typically goes through between 6-9 months of age.

Encourage imitation of speech sounds- When the child can imitate CV sequences, the caregiver can modify them to include different speech sounds like 'ka ka ga ga', or 'ma ma da da'. This could be followed by imitation of animal sounds, like ' Meow meow' or vehicle sounds, like, 'Drrr Peep peep Pom Pom'

Imitation of simple words- The caregiver could encourage the child to imitate simple words like Mama Papa, Bikki, Tata...comprising a functional vocabulary consisting of kinship terms, edibles, objects of his need, play and interest, objects around the house, names of favourite cartoon characters, etc.

Encourage two word phrases- once child is able to communicate at word level the caregiver could combine two words to form short phrases such as, 'Rohan's mummy'(Agent object), ' Rohan come (Agent action)' ' Throw ball(Action object)'

Encourage small sentences- Once child starts using small phrases, the caregiver can facilitate simple sentence structure like,' Rohan wants to eat', 'Doggie says bow bow'.

Many children with CP may have severe expressive disorders. The techniques such as Modelling, Shaping, Chaining, Prompting and fading etc. could be used too.

Many may be able to comprehend spoken language age appropriately or with some lag from that of peers. Others may benefit from intensive therapy and learn to speak or use communication devices. As in all cases, early intervention is the key to success along with concerted efforts from rehabilitation professionals and caregivers to bring out the best participation of children with CP in their environment by improving functioning and reducing participation barriers.

## Psychology

Education and vocational preparation come into the foreground by school age. During this crucial period, concerns about the physical disability should not distract attention from the emotional and social needs of childhood and adolescence. Disabled youngsters need the same variety of life experiences as all other children, to develop emotional flexibility, personal determination, and social skills. As a child with cerebral palsy grows older, the need for different types of therapies and support services will also change. Psychological interventions for children with cerebral palsy are aimed at obtaining early diagnosis of co-morbidities and therapies.

**Diagnosis of psycho-motor and psycho-social development:**

- Assessment of the actual development in terms of deficits of fine motor skills, perceptive development, social development and independence
- Assessment of interaction between mother and child aged 0 to 3 years of age
- Assessment of the level of the deficit manifestation in terms of neurological diagnosis

**Psychological assessment:**

- Cognitive processes: attention span, memory, thinking, intellect
- Assessment of fine motor skills
- Assessment for school maturity
- Assessment of brain functions - gnosis, praxis, speech
- Assessment of the emotional and behavioural state of the child
- Personality assessment

The assessment process differs from an individual to individual and the nature and severity of the case. The assessment process usually lasts for about 45 minutes and if needed then it is held in several sessions. The final medical report includes the test results and other relevant observations made during the examination. Note: The methods and length of assessment are based on the psychological age and capabilities of the child.

Therapeutic Interventions: Below mentioned are list of therapies for individuals with cerebral palsy:

1 ***Neuro-cognitive therapy:***

This is a new approach to treating individuals with cerebral palsy. It is based upon two proven principles:

ii) **Neural Plasticity:** The brain is capable of altering its own structure and

functioning to meet the demands of any particular environment. Consequently if the child is provided with an appropriate neurological environment, he will have the best chance of making progress.

iii) **Learning:** Lev Vygotsky proposed that children's learning is a social activity, which is achieved by interaction with more skilled members of society. Counseling and behaviour therapy, for emotional and psychological challenges may be needed at any age, but is often most crucial during adolescence.

2 ***Cognitive Rehabilitation Therapy*** *(CRT)(Fig 107- 110):*

Children with cognitive impairments face tremendous levels of uncertainty and frustration, thus in need of additional support. As a parent it is very important that you should learn the best ways to interact with your child as this can immensely beneficial as the child grows up.

Cognitive rehabilitation is a program of guided therapy to learn (or relearn) ways to concentrate, remember and solve problems after an illness or injury affecting the brain. It is a structured set of therapeutic activities designed to retrain an individual's ability to think, use judgment and make decisions.

The focus is on improving deficits in memory, attention, perception, learning, planning and judgment. There are many intervention strategies and techniques used to help children reduce, manage or cope with cognitive deficits. The desired outcome of cognitive rehabilitation is an improved quality of life or an improved ability to function in home and community life.

Cognitive rehabilitation techniques are restorative training or re-training the brain in normal function or compensatory ways. This technique teaches the child to make use of tools to help make up for the deficits.

The goal of cognitive rehabilitation is to improve cognitive abilities in order to obtain as much independent functioning as possible. Some of the specific benefits of cognitive rehabilitation include improvements in the following deficits:

- Attention span
- Memory
- Problem - solving
- Visual - spatial relations
- Learning functional tasks such as feeding, dressing, etc.



Figure 107



Figure 108



Figure 109



Figure 110

Cognitive rehabilitation may be performed in conjunction with or as part of other therapies, such as behavior, occupational or speech therapy.

Cognitive rehabilitation may be performed by a variety of professionals, such as neuropsychologists or psychologists. The nature of the condition and the specific cognitive deficits determines what kind of specialist would be needed.

**There are two treatment approaches to cognitive rehabilitation:**

i) **Restorative approach:** This is based on the theory that repetitive exercise can restore lost or under developed functions. Some methods and techniques include:

- Auditory, visual and verbal stimulation and practice
- Number manipulation
- Computer - assisted stimulation and practice
- Behavior modification

i) **Compensatory approach:** This method strives to develop external assistance for cognitive deficits. Compensatory cognitive rehabilitation may use the following techniques:

- Employing visual cues, written instructions, memory notebooks, watches, beepers, computers and other electronic devices to trigger functional behaviors.
- Simplifying complex tasks, capturing the patient's attention, minimizing distractions and teaching self-monitoring procedures.

The compensatory approach to cognitive rehabilitation has been more widely accepted than the restorative approach, but these approaches are not mutually exclusive. Many therapeutic programs employ elements of both.

A few cognitive rehabilitation therapy strategies for children with cerebral palsy that can be practiced at home by parents are mentioned below:

- Many children with cerebral palsy have cognitive impairments and they have difficulty maintaining their attention span on the activity or when they are being spoken to.

Designate a separate room or a part of a room that is his special area. Avoid brilliant colors or complex patterns in decor. Simplicity, solid colors, minimal clutter, not near a window, proper lighting and a worktable facing a blank wall away from distractions will help to increase attention span and concentration.

- Make use of visual cues like flashcards to teach as this would help the child have clear concept formation.
- Due to physical limitations many children have difficulty writing and producing good, legible handwriting and repeated practice would only frustrate your child. Try and make writing enjoyable by making use of paints, crayons, sand, etc. You can also make use to computers, magical pens and white boards to improve the handwriting and make the activity more interesting.
- There is wide range of educational audio visual CD's available in the market which can be used to teach the child concepts like colours, shapes, fruits, body parts, daily activities of living, etc.
- Modify the homework and class work assignments for the child, according to his or her functioning and the possible barriers which could restrict his development.
- Expose the child to new learning(Fig 106- 110)
- Left untreated, a child may become isolated, have counter-productive interactions, and experience peer rejection. These can lead to lower academic performance, social- emotional deficiencies, and attention deficits. If a child exhibits, the above mentioned signs then its best to consult a psychologist.

### 3 Behaviour therapy:

This therapy is often used to enhance child's ability and discourage destructive behaviours. As symptoms of cerebral palsy can cause behavioural and emotional problems, many children benefit from counseling or behavior therapy. Behavioural therapy utilizes psychological techniques to improve physical, mental, and communicative skills. The activities used may vary greatly according to age and disability of the child. Some techniques will be used to discourage destructive behaviour whereas the others to encourage self-sufficiency. Ultimately, a behavioural therapist will act as a coach to the patient and family, by suggesting ways to improve behaviour, as most of the work will be done at home.

Behaviour therapy can complement physical therapy, as employing psychological techniques that encourage the mastery of tasks can promote muscular and motor development. Behavioral therapy can help alleviate depression, mood swings, sadness, loss, anger and frustration by allowing previous negative outcomes to be replaced with a more positive perspective. Praise, positive reinforcement, and small rewards can encourage a child to learn to use weak limbs, overcome speech deficits, and stop negative behaviors like hair pulling,

throwing temper tantrum, etc.

Living with a disability, no matter what the severity, can feel like a limitation to the disabled person. It is for this reason that individuals, especially children who are subjected to the limitations of a disability from early on in life, can at times develop negative behavioral traits that may further affect the person's ability to live life fruitfully and independently. It is in such conditions, in which the person's quality of life is being affected or they are affecting the environment around them that behavioural therapy can be so valuable. Even in situations where no such personality traits have surfaced, behavioural therapy is an important part of a child's ongoing therapeutic schedule.

In cases involving behavioural therapy, the treatment is aimed at helping them not only become more independent and productive, but also more able to function in everyday life. For instance, in physical therapy the therapist will help the child to learn to walk with more ease by walking with them, or pick things up with more ease while helping their muscles develop, while in behavioral therapy the therapist might put an object of interest, perhaps a toy or a piece of candy, into a box. They will then request that the child with cerebral palsy to reach into the box with his or her weaker hand. It is the reward aspect of coaching the child to use his or her weaker hand that connects with his or her mind.

A few behaviour therapy strategies for children with cerebral palsy that can be practiced at home by parents are mentioned below:

1. First develop a consistent behavioral management plan to use with your child when appropriate or inappropriate behaviors are exhibited. This could mean allowing the child to play with his favorite toy when he displays desired behaviour or giving a time –out when the child displays inappropriate behaviour.
2. Use a reward system to provide motivation to your child for displaying good behaviors and tasks completed. Set up a clear system of rewards like sticker charts, gold stars, non-edible treat such as toys so that the child knows what to expect when they complete a task or refine their behavior.
3. Praise your child for good behaviors rather than only giving time-out or shouting, when he /she displays bad behaviours. This will help your child develop feelings of self-worth and will build upon his / her confidence level.
4. The child should be taught constructive and acceptable ways to vent out physical or verbal aggression. For example: either drawing, painting, playing a musical instrument like drums, practicing breathing exercise when he or she is angry or frustrated.
5. Many children with cerebral palsy become over stimulated in a crowded, noisy environment, which can lead to meltdowns. Learn to recognize what external triggers lead to undesirable behavior in your child, and remove her from those situations. You can help the child by removing the child from the over –stimulated environment or teach the child to calm himself or herself down in such situations.
6. As children with cerebral palsy have limited exposure to socializing due to their physical condition they need to be taught skills to socialize. Techniques such as modeling, role playing and

skills training can be conducted with the child as this will give the child a clear ideas as to what constitutes in socially adaptive behavior.

7. Re-evaluate your family's communication methods if your child seems to be unusually frustrated by the inability to express himself/ herself. Perhaps purchasing a communication board that allows the child to type out his thoughts or point to words can help.
8. Get your child into therapy with a behavioral specialist if you have tried other options and problem behaviors still exist. A professional therapist can help your child sort through his feelings, become better at communicating his wishes and feel better about himself/ herself.

**4. Counseling or Talk Therapy:**

As they get older, children with cerebral palsy will likely feel disliked by their peers, isolated from friendships, embarrassed by body image and/or frustrated with treatment goals. This may be a good time to introduce them to "counseling" or "talk therapy," where they can talk about the things that are bothering them and learn to put them in perspective. It differs from behavioral therapy in that the goals are more along the lines of learning to accept and embrace one's individuality rather than of raw behavior modification.

Even the most "normal" young person faces daily psychological and emotional challenges. Your hopes for your child at this age won't differ much from that of any other parent. You want your child to cultivate a healthy attitude toward his or her challenges and to acquire the basic tools that will carry them into adulthood. Many adolescents and teens see therapists to help process these complicated years.

The counselor will talk with his or her patient to help work through the emotional stresses that cerebral palsy can place on a child. Sometimes children with cerebral palsy can become violent or aggressive, resorting to things such as biting or hair-pulling to help release their anger. It is the job of the counselor to help the child find new ways to release their aggression and frustrations, either vocally or, if the child is able to control his or her hand enough, perhaps drawing or writing, with the aid of new computer technology that makes it possible for people with cerebral palsy to type and have full use of a computer. A counselor will be able to help your child see his or her situation in the best light, and hopefully help to alleviate many of the stresses that a child with cerebral palsy goes through. The right therapist can work wonders.

**5. Aversion therapy:**

This therapy works on the principle to reward rather than punish negative consequences. This can help enhance self – esteem and the confidence level of the individual. For example, if a child is throwing temper tantrums and isn't cooperative during the therapy sessions then the parent or there therapist should not shout or get angry at the child. Rather the child should be rewarded if he or she moves in a positive direction for example; in spite of the temper tantrums if he is exercising then he or she should be praised and rewarded.

**Group Therapy:**

Group psychotherapy is a process of psychological influence by verbal and non-

verbal techniques in which the psychologist uses the interactions created in specially organized small groups of patients (mainly interactions of emotional character), in order to reduce the patients' body and nervous discomfort and to improve psycho-social functioning. Individuals with physical or cognitive limitations often face real, and sometimes self-imposed, challenges in building relationships. Difficulties in communicating, fitting in or feeling accepted can lead to delays in social, emotional and even physical development.

The group therapy is viewed as an important factor for individualization and socialization of children with cerebral palsy. The purpose of this method is to relieve patients and to develop their skills to solve problems as well to overcome noxious personal and behavioural stereotypes. Through group therapy children are able to acquire new knowledge, skills and good behaviour. The group rehabilitation and therapy depends on the specificity and psychological development of children within the group.

**6. Caring for Caregivers:**

The family is considered to be a system of continuous process with different behavioural models. Not only does cerebral palsy alter the emotional dynamics between your child and you, but it also stresses the emotional dialogues between you and your family. We believe parents and the family members have to be emotionally prepared to accept the diagnosis and to carry their child through this "special" journey. You must understand and see the "special" needs of their child. Devoted support from caregivers (you and the family members) is crucial for an improved quality of life and attempting to normalize this "given" situation.

**Strategies for parents to cope with the situation:**

i) Ask for help if it's difficult caring for your child. For example: Hiring a maid makes more sense than wasting time into household work, rather you could invest that time into teaching your child or may be time for yourself like exercising.

ii) Make use of the school hours for meeting your relatives, family members or friends while the child is away. Plan quality time together with your spouse for example; lunch with him when your child is at the school.

iii) Do not feel guilty about the break and try to remind yourself that it will help you to be renewed for the things you need to do when you get back. Try and schedule your time in such a way that you are left with sometime for yourself. If possible, take time off and go for shopping or to the parlor, visit a friend, etc.

iv) It is very important that you talk about your feelings and emotions that you are going through. It is important to vent out your thoughts at regular interval of time. Especially someone who just listens could be a great source of strength.

v) If you feel that you are unable to cope up with things around and your stress or anxiety levels have increased or you are starting to show depressive symptoms (like crying spells, change in sleep or appetite, loss of interest, etc) seek help of a professional. Counseling session for you or your spouse or the entire family may help.

vi) You could join support groups which are for children with autism and their

parents. It may be helpful to listen or talk to people who have been or are going through a similar situation. Also, support groups could be a great source of information about the services available in your area or about the new possible treatment options available.

The principles of the family therapy are used during consultations with patients. The work is done with trained family therapists. The family will be informed about the confidentiality of the process.

7 **Expressive therapies:** These are usually used with people who have difficulty verbalizing their feelings such as art, music, poetry, etc which could help freeing and empowering oneself. Sometimes children with cerebral palsy can become violent or aggressive, resorting to things such as biting or hair pulling to they can be helped to release their aggression and frustrations, by either being vocal or, if the child is able to control his or her hand enough, perhaps drawing or writing.

To conclude….

Regardless of advancements in technology, a psychologist can have a profound impact on your child's life. Because cerebral palsy can so greatly affect a child's physical ability, the psychological health and development of the child is of least concern. However, a positive state of mind and outlook can lead to an improved quality of life and functioning.

# Chp. 14 Autism

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**Definition:**

- The Diagnostic and Statistical Manual of Mental Disorders, DSM-V defines autism spectrum disorders as pervasive developmental disorders characterized by:
  - Qualitative impairment in social interaction
  - Qualitative impairment in communication
  - Restricted, repetitive and stereotypic patterns of behavior, interests and activities.
- Autism is diagnosed when a child or adult has abnormalities in a 'triad' of behavioral domains: social development, communication, and repetitive behavior/ obsessive interests.
- Autism Society of America (ASA)defined autism as: "Autism is a complex developmental disability that typically appears during the first three years of life and is the result of a neurological disorder that affects the normal functioning of the brain impacting the development in the areas of social interaction and communication skills. Both children and adults with autism typically show difficulties in verbal and non-verbal communication, social interactions and leisure or play activities. Autism is one of the five disorders that fall under the Pervasive Developmental Disorder (PDD), a category of neurological disorders characterized by "severe and pervasive impairment in several areas of development".

**Epidemiology:**

**Prevalence:**

Autistic disorder is believed to occur at a rate of about 8 cases per 10,000 children (0.08percent) and it continues to grow at an alarming rate however, because of inadequate data, these numbers may underestimate autisms true prevalence. By definition, the onset of autistic disorder is before the age of 3 years, although in many cases it noticed at a much older age.

**Sex Distribution:**

It is seen that autistic disorder is four to five times more common in boys than in girls. However, girls with autistic disorder are more probable to have more severe mental retardation as compared to boys.

**Socio economic Status:**

Over the past 25 years, no epidemiological studies have demonstrated an association between autistic disorder and any socio economic status.

**Etiology and Pathogenesis:**

**1.Genetic Factors:**

Current evidence supports a genetic basis for the development of autistic disorder in most cases. Family studies have demonstrated a 50 to 200 times increase in the rate of autism in siblings of an index child with autistic disorder. Studies suggest that even if the siblings are not affected with autism they are an increased risk for a variety of

developmental disorders which are often related to communication and social skills. Linkage analyses have demonstrated that regions of chromosomes 7, 2, 4, 14 and 19 are likely to contribute to the genetic basis of autism.

The concordance rate of autistic disorder in the two largest twin studies was 36 percent in monozygotic pairs versus 0 percent in dizygotic pairs in one study and about 96 percent in monozygotic pairs versus about 27 percent in dizygotic pairs in the second study. Approximately 1 percent of children with autistic disorder also have fragile X syndrome, who tend to show gross motor and fine motor difficulties as well as relatively poorer expressive language compared with children with autism without fragile X syndrome.

Recent research was conducted where, the DNA of more than 150 pairs of siblings with autism was analyzed and they found extremely strong evidence that two regions on chromosome 2 and 7 contain genes involved with autism. The likely locations for autism - related genes were found on chromosome 16 and 17, although the strength of correlation was somewhat weaker.

**2.Biological Factors:**

Approximately 70 percent of children diagnosed with autistic disorder have mental retardation. About one third of children with autistic disorder have mild to moderate and close to half of these children are severely or profoundly mentally retarded. Children with co-morbid autistic disorder and mental retardation display marked deficits in social understanding, abstract reasoning and verbal tasks than in performance tasks, such as digit recall and block design.

Of persons with autism, 4 to 32 percent have grand mal seizures at sometime and about 20 to 25 percent show ventricular enlargement on computed tomography (CT) scans. Various electroencephalogram (EEG) abnormalities are found in 10 to 83 percent autistic children and although no EEG finding is specific in autistic disorder, there is some indication of failed cerebral lateralization.

**3.Immunological Factors:**

The lymphocytes of some autistic children react with maternal antibodies, which raise the possibility that embryonic neural or extra embryonic tissues may be damaged during gestation.

**4.Perinatal Factors:**

A higher than expected incidence of perinatal complications seems to occur in infants who are later diagnosed with autistic disorder. Maternal bleeding after the first trimester and meconium in the amniotic fluid has been reported in the histories of autistic children than the normal population. It is seen that in the neonatal period, autistic children have a high incidence of respiratory distress syndrome and neonatal anemia. Males with autism, as a group have been found to be the products of longer gestational age and were heavier babies than babies in the general population. On the other hand, females with autism are more likely to be the product of post term pregnancies than babies in the general population.

**5.Neuroanatomical Factors:**

The neuroanatomical basis of autism remains unknown; however, recent evidence suggests that enlargement of the gray and white matter cerebral volumes, but not at 2 years of age. It is seen that the head circumference appears normal at birth; however, the head circumference growth appears to emerge at about 12 months of age. MRI studies, where the brain volume of the autistic subjects and normal controls was compared the results revealed that the brain volume was larger in those with autism,

however it seen that autistic children with severe mental retardation generally have smaller heads. The greatest average percentage increase in size occurred in the occipital lobe, parietal lobe and temporal lobe, however no difference was found in the frontal lobe. The increased volume can occur from three different possible mechanisms: increased neurogenesis, decreased neuronal death and increased production of non-neuronal brain tissue, such as glial cells or blood vessels. Brain has been suggested as possible biological marker for autistic disorder. The temporal lobe damage is reported to be one of the critical areas of the brain abnormality in autistic disorders as also animal studies reveal that when the temporal region of the animals is damaged, normal social behavior is lost, there is presence of repetitive motor behavior and restlessness. Some brains of autistic individuals exhibit a decrease in cerebellar Purkinje's cells, which is believed to account potentially for abnormalities of attention, arousal and sensory processes.

An interesting study, which reveals the differences between male and female brains suggests the capacity to predict and respond to feelings and behavior of others by inferring their emotional states, is a stronger trait in females than in male's area population level i.e. empathizing. On the other hand, male population is stronger at systemizing, that is, inferring rules that govern "cause and effect" relationships of behavior. People with pervasive developmental disorders are characterized by deficits in empathizing and those with high intellectual capacity have been reported to have relative strengths in rule bound thinking.

A neural basis of empathy or social intelligence was first proposed by Brothers . She suggested from animal lesion studies, single cell recording studies, and neurological studies that social intelligence was a function of three regions: the amygdala, the orbito-frontal cortex (OFC), and the superior temporal sulcus and gyrus (STG). Together, she called these the "social brain".

**The amygdale:**

There are four lines of evidence for an amygdala deficit in autism

(a) Post-mortem evidence:

A neuroanatomical study of adults with autism at post-mortem found microscopic pathology (in the form of increased cell density) in the amygdala, in the presence of normal amygdala volume.

(b) Similarities between autism and patients following amygdalotom:

Patients with amygdala lesions showimpairments in social judgement that have been likened to "acquired autism". People diagnosed with autism tend to show a similar pattern of deficits to those seen in patients with amygdala lesions.

(c) Structural neuroimaging:

A recent structural magnetic resonance imaging study of autism reported reduced amygdala volume. This is not the only structural abnormality in the brain(see below),but the amygdala abnormality has some potential relevance to the social symptoms observed.It Is not yet known why this difference occurs.

(d) Functional neuroimaging:

Using single photon emissioncomputed tomography (SPECT), patients with autism spectrum conditions show significant reductions in temporal lobe blood flow. This is not simply an effect of temporal lobe epilepsy. Inarecent functional magnetic resonance imaging (fMRI) study,adults with High Functioning Autism(HFA) or Asperger Syndrome (AS) showed significantly less amygdala activation during a mentalizing

task (Reading the Mind in the Eyes task), compared to normal.

**Other brain areas that might be abnormal in autism:**

Reduced neuron size and increased cell-packing density has also been found in the limbic system, specificallythe hippocampus, subiculum, entorhinal cortex, amygdala, mammillary bodies, anterior cingulate, and septum in autism.

6. Biochemical Factors:

A number of studies report that about one thirdof patients with autistic disorder have high plasma serotonin concentrations, however, this is not specific to autistic disorder as children with mental retardation without autistic disorder also display this trait. In some autistic children, a high concentration of homovanillic acid in the cerebrospinal fluid(CSF)is associated with increased withdrawal and stereotypes. Some evidence indicated that symptom severity decreases as the ratio of 5-hydroxyindoleacetic acid (5 - HIAA, metabolite of serotonin) to homovanillin acid in CSF increases. The 5-HIAA concentration in CSF may be inversely proportional to blood serotonin concentration, which is increased in one third of autistic disorder patients, anon-specific finding that also occurs in mentally retarded persons.

7. Psychosocial and FamilyFactors:

Studies comparing parents of autistic children with parents of normal children have shown no significant differences in child- rearing skills. It is seen that children with autistic disorder respond like children with other disorders respond to exacerbated symptoms like family discord, the birth of a new sibling or a family move,etc. Some autistic children may be extremely sensitive to even small changes in the families and immediate environment. While making the diagnosis the evaluators should consider prenatal history, perinatal history, birth complications, developmental milestones and outlining family history via interviews with the parent or relevant caregivers with the help of formal and informal assessment. Autistic disorder can be diagnosed by a physician, a psychologist, or a psychiatrist. Specific tools can be used to check for ASD.

**Diagnosis and Clinical Features:**

Autistic disorder has been diagnosed using the Diagnostic Stastical Manual - IV th Edition - Text Revision, this is a manual with diagnostic categories approved by the American Psychological Association and International Statistical Classification of Diseases and Related Health Problems, 10th revision(ICD-10) published by The World Health Organization(WHO). In1994,when the fourth edition of DSM was published, five categories appeared under the Pervasive Developmental Disorder (PDD) heading: Autism, PDD-NOS, Asperger's syndrome, Rett's syndrome, and childhood disintegrative disorder. PDD is an umbrella term for disorders that involve impairments in reciprocal social interaction skills and communication skills, and the presence of stereotypical behaviours, interests and activities. The term Autistic Spectrum Disorder is used to represent the fact that while these individuals share common characteristics, how these characteristics are manifested will differ with each individual. As a result, no two individuals are the same. It is sometimes complicated diagnosing autistic disorder because there are no medical tests, blood tests or any radiological images which will definitively indicate or diagnose a person with autistic disorder. Also, it could be difficult as the person has other issues related to medical, emotional, sensory or learning difficulties. An accurate diagnosis is made on the basis of specific intellectual, social and behavioural characteristics that are listed in the DSM - IV - TR Manual. The diagnosis can be made by a

physician, a psychologist, or a psychiatrist.

The DSM - IV - TR diagnostic criteria for autistic disorder are as follows:

Diagnostic code according to DSM - IV - TR: 299.00

A. A total of six(or more)items from(1),(2)and (3), with at least two from (1) and one each from (2) and(3):

(1) Qualitative impairment in social interaction as manifested by at least two of the following:

a) Marked impairments in the use of multiple nonverbal behaviors such as eye-to-eye gaze, facial expression, body posture, and gestures to regulate social interaction

b) Failure to develop peer relationships appropriate to developmental level

c) A lack of spontaneous seeking to share enjoyment, interests, or achievements with other people, (e.g; by a lack of showing, bringing, or pointing out objects of interest)

d) Lack of social or emotional reciprocity

(2) Qualitative impairments in communication as manifested by at least one of the following:

a) Delay in or total lack of, the development of spoken language (not accompanied by an attempt to compensate through alternative modes of communication such as gesture or mime)

b) In individuals with adequate speech, marked impairment in the ability to initiate or sustain a conversation with others

c) Stereotyped and repetitive use of language or idiosyncratic language

d) Lack of varied, spontaneous make-believe play or social imitative play appropriate to developmental level

(3) Restricted repetitive and stereotyped patterns of behavior, interests and activities, as manifested by at least two of the following:

a) Encompassing preoccupation with one or more stereotyped and restricted patterns of interest that is abnormal either in intensity or focus

b) Apparently inflexible adherence to specific, nonfunctional routines or rituals

c) Stereotyped and repetitive motor mannerisms(e.g.hand or finger flapping or twisting, or complex whole-body movements)

d) Persistent preoccupation with parts of objects

B. Delays or abnormal functioning in atleast one of the following areas, with onset prior to age 3years:

1) Social interaction

2) Language as used insocial communication

3) symbolic or imaginative play

C. The disturbance is not better accounted for by Rett's Disorder or Childhood Disintegrative Disorder

**The International Statistical Classification of Diseases and Related Health Problems, tenth revision (ICD-10) diagnostic criteria for autistic disorder are as follows:**

Diagnostic code according to ICD - 10: F84.0

A type of pervasive developmental disorder that is defined by: (a) the presence of abnormal or impaired development that is manifest before the age of three years,and

(b) the characteristic type of abnormal functioning in all the three areas of psychopathology: reciprocal social

interaction, communication, and restricted, stereotyped, repetitive behaviour. In addition to these specific diagnostic features, a range of other nonspecific problems are common, such as phobias, sleeping and eating disturbances, temper tantrums, and (self-directed) aggression.

Autistic disorder Infantile:

- Autism
- Psychosis Kanner's syndrome
- Excl.: autistic psychopathy (F84.5)

**Atypical autism: F84.1**

A type of pervasive developmental disorder that differs from childhood autism either in age of onset or in failing to fulfill all three sets of diagnostic criteria. This subcategory should be used when there is abnormal and impaired development that is present only after age three years, and a lack of sufficient demonstrable abnormalities in one or two of the three areas of psychopathology required for the diagnosis of autism (namely, reciprocal social interactions, communication, and restricted, stereotyped, repetitive behaviour) in spite of characteristic abnormalities in the other area(s). Atypical autism arises most often in profoundly retarded individuals and in individuals with a severe specific developmental disorder of receptive language.

Atypical childhood psychosis

Mental retardation with autistic features

Use additional code(F70-F79),if desired,to identify mental retardation.

**Other Pervasive Development Disorders:**

The diagnostic category known as Pervasive Developmental Disorders includes: Autistic Disorder, Asperger's Disorder, Rett's Disorder, Childhood Disintegrative Disorder and Pervasive Developmental Disorder Not Otherwise Specified. All of these disorders share common features.

However, there are differences in some areas,such as the number of symptoms, age of onset, developmental pattern and level of cognitive functioning.

**Physical Characteristics:**

Children with autistic disorder do not show any physical signs indicating the disorder on the first glance.A greater than expected number of children do not show lateralization and remain ambidextrous at an age when the cerebral dominance is established in mostchildren.

**Seizures:**

It is estimated that 25% of autistic individuals also develop seizures, some in early childhood and others as they go through puberty (changes in hormone levels may trigger seizures). These seizures can range from mild(e.g;gazing into space for a few seconds) to severe, grand malseizures.

Many autistic individuals have subclinical seizures which are not easily noticeable but can significantly affect mental function. A short one- or two-hour EEG may not be able to detect any abnormal activity, so a 24-hour EEG may be necessary. Although drugs can be used to reduce seizure activity, the child's health must be checked regularly because these drugs can beharmful.

**Sleep Problems:**

Many autistic individuals have sleep problems. Night waking maybe due to reflux of stomach acid into the oesophagus.

**Pica:**

30% of children with autism have moderate to severe pica. Pica refers to eating non-food items such as paint, sand, dirt, paper, etc.Pica can expose the child to heavy metal poisoning, especially if there is lead in the paint or in the soil.

**Low Muscle Tone:**

A study conducted by the first author found that 30% of autistic children have moderate to severe loss of muscle tone, and this can limit their gross and fine motor skills. That study found that these children tend to have low potassium levels.

Chronic Constipation and/or Diarrhoea: Analyses of the ARI's autism database of thousands of casess howover 50% of autistic children have chronic constipation and / or diarrhoea. Diarrhoea may actually be due to constipation-i.e; only liquid is able to leak past a constipated stool mass in the intestine. Manual probing often fails to find an impaction. An endoscopy may be the only way to check for this problem. Consultation with a paediatric gastroenterologist is required.

**Behavioural Characteristics:**

**Difficulties in Social Skills:**

Have you ever made a mistake in social situations? Or To do a joke that did not fit the event? Or misread the intentions of others? Then it is easier to understand why individuals with autism have difficulty in social situations. Social situation is very demanding as they need us to make quick judgements, respond to unpredictable events, read facial expressions and body gestures of others. For individuals with autism, these skills are missing or are functioning at a deficit.

Social difficulties are the primary that individuals with autistic disorder face challenge being employed or in employment. As when young they usually have minimal interest in playing or interacting with peers, hence when social engagement occurs it is typically on their own terms or it gets very awkward. These responses reflect a lack of skill in knowing how to interact rather than a lack of desire in socialization. Also, as they stick to their ritualistic routine, they have difficulty in changing or alternating routine for which they need additional support and transitioning time to new and unfamiliar experiences. Autistic children do not display the expected level of subtle reciprocal social skills that demonstrate relatedness to parents and peers. Children with autistic disorder may lack social smile and may have anticipatory posture as an adult- approaches. They may also have less frequent or poor eye contact. Autistic children display impaired social developmen ti.e. they may display poor attachment behaviour towards parents or peers. They may not be able to differentiate between the most important people in their lives like their parents, siblings, and teachers.



*Fig 1: Anxiety experience by autistic child*

Autistic children strictly adhere to their routine and when disrupted they may display extreme anxiety and irritability. When autistic children have reached schoolage, their withdrawal may have diminished and may be less obvious, particularly in higher- functioning children. A prominent deficit is seen in ability to play with peers and to make friends; their social behavior is awkward and may be inappropriate. Cognitively, children with autistic disorder are more skilled in visual-spatial tasks requiring skill in verbal reasoning. The cognitive style of children with autism is that they cannot infer the feelings or mental state of others around them.That is,they cannot make attributions

about the motivation or intentions of others and thus, cannot develop empathy. This lack of a "theory of mind" leaves them unable to interpret the social behaviour of other s and leads to lack of social reciprocation.

Autistic person in late adolescence, often desire friendship, but their difficulties in responding to another's interests,emotions and feelings are major obstacles in developing them. They are often shunned by peers because of their awkward behaviour that alienate them form others. Autistic adolescents and adults experience sexual feelings, but their lack of social competence and skills prevent many of them from developing sexual relationships.

**Disturbance of Communication and Language:**

Autistic children are not simply reluctant to speak and their speech abnormalities do not result from lack of motivation. Language deviance, as language delay, is characteristic of autistic disorder. In contrast to normal and mentally retarded children, autistic children have significant difficulty putting meaningful sentences together even when they have large vocabularies. In the first year of life, an autistic child's pattern of babbling maybe minimal or abnormal. Some children emit noises like clicks, screeches and nonsense syllables-in a stereotyped fashion, without a seeming intent of communication.

Autistic children with delays in learning to talk and use language may not compensate by using extensive gestures and pointing. Instead young children with this disorder may put an adult's hand on what they want, pull the adult over to the object they want to access, or engage in problematic behaviors to express their message. Individuals with underdeveloped speech may grunt, point a finger, pull an adult to an area,or use a picturecue and some may never develop a meaningful speech and may need to use singing and/or augmentative communication devices. Others begin their journey towards developing oral communication skills by using echolalia. Some have a very restricted repertoire of use of their communication skills and need direct instruction and support to expand their skills. Even those who develop oral skills often have other language/communication problems in social and academic situations.

Individuals with autistic disorder are very concrete in their understanding of the world and can have significant comprehension problems and significant gaps in their store of background knowledge; hence they may see jokes and sarcasm as lies and then mistrust the speakers.

**Stereotypes Behaviour:**

Restrictive, repetitive and stereotypical behaviour may vary with individuals, circumstance, and age and by the level of awareness about others. In an autistic child's first few years of life, the expected spontaneous exploratory play is absent. The toys and objects are often manipulated in a ritualistic manner, with few symbolic features. Autistic children generally do not show imitative play or use abstract pantomime. The play and other activities of these children are often rigid, repetitive and monotonous. The ritualistic and compulsive phenomenaare common in infants and adolescentsyears. Autistic children often use in animate objects in a very vague manner like spinning bottles, banging toys and may exhibit attachment to particular inanimate objects like wires, rods, etc. Autistic children generally show resistance to transition and change like moving to a new house, moving furniture in a room, or a change, such as having breakfast before a bath when the reverse was routine was the routine may evoke panic, fear or temper tantrums.

Rocking one's body for example is a repetitive behaviour which occurs when the child is anxious, agitated at home, school and

social situations. Repetitive behaviours might include lining things up, ensuring that all cupboard doors are closed, aligning chairs in a certain fashion, and making certain noises. Stereotyped movements might include hand flapping, rocking, spinning, jumping, and other patterns. Restrictive behavior involves having a narrow set of interests. Some become walking encyclopedias of facts on certain topics.

**Instability of Mood and Affect:**

It is very common for autistic children to exhibit mood changes, with bursts of laughing or crying spells for no apparent reason. It is however, a difficult task to learn about these episodes if the child cannot express the thoughts to the effects.

**Response to Sensory Stimuli:**

Our senses are conditioned to organize and interact with the world around us. It is important to realize, understand, and accept that some individuals on the autism spectrum may actually feel, hear, see, smell, and taste at an extreme level. They may be hypersensitive (overreactive) or hyposensitive (underreactive) for example to sound and pain.

**Touch Sensation:**

Touch is an important sense that is needed for social interactions with loved ones or to show care for a person. However, some with autistic disorder don't like being touch and respond aggressively on doing so, whereas on the other hand some have a high tolerance for pain and may not realize a shoulder is broken until it is swollen for several days.

**Auditory Senses:**

Imagine how it would feel to hear a fire alarm, vacuum cleaner, or a room full of children at a birthday party at a magnified level at the same time within the environment. This is what autistic children go though and they have difficulty in blocking and auditory senses. An individual may have difficulty discriminating between sounds, remembering directions, paying attention to a voice, and / or reading aloud.

**Vestibular sensations:**

Vestibular sensations occur in our inner ear. The inner ear receptors register every movement we make and every change in head position. This encompasses messages from our neck, eyes, and body. Rotary movements involve moving in circles (e.g; spinning). Linear movements (i.e; back and forth, side to side, up and down), especially when rapid (i.e;rocking in a chair, swaying, swingingon atire), may cause most to become dizzy, nauseated, or to get a headache. Individuals with autistic disorder may actually crave these sorts of movements and this means that they need a lot of vigorous activity in order to get started. Some become distressed and show anxiety due to a fear of falling, being pickedup, standing up, or sledding down a hill.

Visual Senses:

Individual with autistic disorder maybe distracted with objects hanging from the ceiling; they may feel blinded by the sun or maybe unable to focus in the presence of florescent lighting.Whereas others may have their eyes glued on to spinning or bouncing objects or reflection ofobjects.

Smell and Taste Sensation:

Some individuals with autistic disorder may be highly agitated by perfumes ,the odour of foods or animal or hand lotions and may avoid people, places or foods so that they are not around that odour.Whereas, individuals hyposensitive to smell may crave odours or tastes and may lick or taste inedible objects(e.g;clay, chalk)or prefer spicy,hot, or sour foods.

Proprioceptive feedback helps us position our bodies and move through the environment. Proprioceptors exist in our

muscles, joints, ligaments, tendons, and connective tissue. Proprioceptors work closely with tactile and vestibular systems through body awareness, motor control, planning, and postural stability. Autistic children may need to keep their eyes open inorder to know how their own body is moving. Motor control or planning involves coordinating one's gross and fine motor skills within the environment.Those who have difficulty in this area may bump into people or obstacles or frequently fall or trip. Some cannot regulate how much pressure to exert when grasping an object. As a result, they hold pencils too tightly, and their written work is messy. Individuals on the autism spectrum may have poor posture, lean their head on their hands, and be unable to balance on one foot. In all of these areas, individuals may feel emotionally insecure due to feeling uncomfortable or inadequate, rigid, or intimidated by the environment. It is imperative to accommodate individuals with sensory needs throughout the day to help them calm their nervous systems so they can function in home, community, or work. These breaks should not be considered a reward but as a necessary requirement for a person's day. Many autistic children reportedly enjoy music, where they frequently hum a tune or sing a song or commercial jingle before saying words or using speech.

**Associated Behavioral Symptoms:**



*Fig 2: Self Injurious Behaviour*

Hyperkinesis is a common behaviour problem in young autistic children whereas hypokinesis is less frequent; but when present it often alternates with hyperactivity. Aggression and tempertantrums are observed, often prompted by change or demands. Self-injurious behaviors include head banging, biting themselves or others, picking their scabs, scratching, and hair pulling. Others may react to their environment by being physically hurting their siblings, peers or family members, and such behaviors may escalate during the adolescent years due to the hormonal changes. Some children may flap their arms,brush their hands against their face repeatedly, or may hum as a self -stimulatory behaviour.Short attention, poor ability to focus on task, insomnia, feeding and eating problems and enuresis are also common among children with autism.




*Fig 3: Head Banging behaviour*

**Difficulties with Executive Function:**

Individuals with autism have difficulty in multi-tasking, completing assignments and organizing. They are better at carrying out one task at a time. Executive functions also include deficits in the area of making transitions and organizing oneself.

**Intellectual Functioning:**

Approximately 70 to 75 percent of children with autistic disorder fall in the mentally retarded range of intellectual function.

About 30 percent of children function in the mild to moderate range, and about 45 to 50 percent are severely to profoundly mentally retarded. Epidemiological and clinical studies show that the risk of autistic disorder increases as the IQ decreases. About 1/5 of all autistic children have a normal, non-verbal intelligence. The Intelligence Quotient scores of autistic children tend to reflect most served problems with verbal sequencing and extraction still, with relative strength in visuo- spatial or rote memory skill. This finding suggests the importance of the effects in language related functions.

Unusual order precocious cognitive or visuo-motor abilities occur in some autistic children. The abilities, which may exist even in the overall retarded functioning, are referred to as splinter functions or islets of precocity. Perhaps, the most striking example is autistic savants, who have exceptional rote memories or calculating abilities, usually beyond the capabilities of their normal peers. Other gifted abilities in young autistic children include hyperlexia, an early ability to read well (although they cannot understand what they read), memorizing and reciting and musical abilities (singing or playing tunes or recognizing musical pieces). When an individualis gifted in specific areas, he or she is often referred to as being savant. These individuals may have exceptional talents in calculating numbers,playing a musical instrument, or drawing.

**Early Diagnosis:**

Although autism is usually seen to occur during the early gestational abnormalities of brain development, but for the parent it may seem to begin after the first year of life.The babies are often "too good", very quiet and are passive or are too irritable and intense ;the eye contact may be absent from the start and the infants may seem to be unresponsive and different as compared to the other children. The symptoms of autism in a two-year-old are very different as compared to a four year old; hence it is extremely important to make the diagnosis depending on theage. assessment process to begin. Some children maybe seen to be mildly affected with this disorder but eventually may lead to full blown syndrome. Early diagnosis may help to formulate an intervention plan which would significantly help improving the quality of life and functioning of the child.

**Differential Diagnosis:**

Autism must be differentiated from one of the other pervasive developmental disorder such as Asperger's disorder and pervasive developmental disorder not otherwise specified. Further it must be differentiated from other developmental disorders, including mental retardation syndromes and developmental language disorders. It is sometimes difficult to make the diagnos is of autism because of over- lapping symptoms specified below:

Schizophrenia with Childhood Onset: Although a wealth of literature on autistic disorder is available, few data exists on children underage 12 who meet the diagnostic criteria for schizophrenia.

**Mental Retardation with Behavioral Symptoms:**

Approximately 40 percent of autistic children are moderately, severely or profoundly retarded and retarded children may have behavioral symptoms that include autistic features. When both disorders are comorbid, both should be diagnosed. The prominent distinguishing feature between autistic children and mentally retarded children are that mentally retarded children usually relate to adults and other children in accordance with their mental age, use the language they do have to communicate with others, and exhibitarelatively even profile of impairments without splinter functions.

Mixed Receptive - Expressive Language

**Disorder:**

Some children with mixed receptive - expressive language disorder have mild autistic like features and may present a diagnostic problem.

**Acquired Aphasia with Convulsion:**

Acquired aphasia with convulsion is a rare condition that is sometimes difficult to differentiate from autistic disorder and childhood disintegrative disorder. Children with this condition are normal for several years before losing their receptive and expressive speech.

**Speech:**

Delayed speech and language milestones must be taken very seriously and form the most common presenting symptoms in the autistic spectrum. Hearing impairment should definitely be ruled out with the help of tests. It is often seen that autistic children usually lack "communicative intent" and do not seem to "listen" to adult conversations. Absolute indication of preliminary assessment includes absent babbling at 12 months of age, no single word at 12 months, no phrase at 24 months and at eventually may either lead to developing a restricted speech or absence of speech.

**Social Interaction:**

Poor eye contact and delay in social smiling within the first few months of life may require urgent consideration. The one year old who ignores gestures, does not shake or nod the head appropriately during conversations should raise suspicion. Pointing is a fundamental communicative act which is usually seen when a child is around 15 months of age but if there is a failure to point to objects at 18 months of age; this would be a strong suspicion of autism. However, absence of pointing may be seen in children who are mentally retarded or with visual impairment. They may also lack interest in imaginative play or social interaction and if play is present it could be present in the form of repetitive behavior, rejection of new toys, or bizarre use oftoys.The critical step in early diagnosis is to recognize the grounds for suspicion and to arrange for their expressive language over a period of weeks or months. A profound language comprehension disorder then follows, characterized by deviant speech pattern and impairment. Some children recover, but with considerable residual language impairment.

**Congenital Deafness or Severe Hearing Impairment:**

Autistic children are often mute or show selective disinterest in spoken language, they are often thought to be deaf. The prominent distinguishing factors are as follows:

- Autistic infants may babble once in a while, whereas deaf infants have a history of relatively normal babbling that then gradually tapers off and may stop at 6 months to 1 year of age.
- Deaf children respond only to loud sounds, where as autistic children may ignore the loud or normal sounds and respond to soft or low sounds.
- Most importantly audiogram or auditory evoked potentials indicates ignificant hearing loss in deaf children.
- Unlike autistic children, deaf children usually relate to their parents, seek affection and enjoy being held as infants.

**Psychosocial Deprivation:**

Severe disturbance in the physical and emotional environment(e.g. maternal deprivation, psychosocial dwarfism, hospitalism,and failure to thrive) can cause children to appear apathetic, withdrawn and alienated. Language and motor skills can be delayed. Children with these signs almost always improve rapidly when placed in a favorable and enriched psychosocial

environment, but such improvement is not the case with autistic children.

**Course and Prognosis:**

Autism is generally a lifelong disorder with the guarded prognosis. Autistic children who have intelligence quotient about 70 and those who use communicative language by ages five to seven years tend to have the best to prognosis. It is seen that symptoms such as repetitive and ritualistic behavior did not seem to improve overtime. Studies on the outcome of children with autistic disorder reported that about two third of autistic adult's remains severely handicapped and live in complete dependence or semi-dependency with their caregivers or relatives. Only about 2 percent are able to lead a normal, independent life with employment and about 5 to 20 percent of them are able to achieve a borderline status.

The prognosis of children is believed to improve if the environment or home is supportive and capable of meeting the extensive need of such a child. The symptoms may decrease in some cases as the age increase but with others severe self-mutilation and aggressiveness and aggression have been seen to increase. About 4 to 32 per cent have generalized, tonic–clonic seizures in late childhood or adolescence, and seizures adversely affect the prognosis.

**Treatment for Autism:**

Intervention for autism is a very intensive, comprehensive and one which involves the child's entire family and a qualified team of professionals. Based on the issues that the child with autism has and a comprehensive evaluation, the therapies and therapists are narrowed down for the child. Treatment programs may combine therapies for both core symptoms and associated symptoms.

**Medical Management:**

Medical management makes an important contribution to the overall wellbeing and outcome in individuals with autism and to their family members, even though the primary intervention is psychological and educational in nature. The physician is usually the only resource for the families in search of up- to -date and accurate information on the biology and neuroscience of autism.

**Bowel functions (diarrhea and constipation):**

This occurs with the same frequency in children with autism and other neurological disorders such as cerebral palsy. Constipation usually reflects low muscle tone and immobility and deserves very energetic treatment along conventional lines in order to prevent fissures and pain. Family history of severe constipation should lead to the exclusion of Hirschsprungs disease. Good hydration is essential in the management of both constipation and diarrhea.

**Seizures:**

There is substantial evidence that certain nutritional supplements, especially vitamin B6 and dimethylglycine (DMG), can provide a safer and more effective alternative to drugs, for many individuals.

**Sleep:**

Sleep is a frequent concern for parents of autistic children and can be very difficult to treat. A strict going to sleep routine is even more important for these children than for others, and here their strong need for routine and ritual may be used to help them. Stimulating foods and drinks should be avoided. With the exception of melatonin, no drugs are recommended for insomnia in children, unless a simple antihistamine such as Benadryl is given for a few nights to give the parents a break from incessant crying or hyperactivity. Some truly hyperactive children diagnosed with autism and ADHD

maybe helped to sleep by methylphenidate (Ritalin) but most will be kept awake even longer. Placing bricks under the head of the bed may help keep stomach acid from rising and provide better sleep. Melatonin has been very useful in helping many autistic individuals fall asleep. Other popular interventions include using 5-HTP and implementing a behavior modification program designed to induce sleep. Vigorous exercise will help a child sleep, and other sleep aids are a weighted blanket or tight-fitting mummy-type sleeping bag.

**Dental Treatment:**

As some autistic children are non-verbal they may communicate the pain of dental cavities or issues by being self-abusive, hyperactive or otherwise by a very difficult behaviour.

**Nutrition and Diet:**

It is often observed that autistic children are very fussy feeders who incorporate their obsessions and need for the sameness into their feeding behaviour. Regular measurement of weight, height and head circumference will allow calorie deficiencies to be detected early along with clinical evaluation for signs of malnutrition. As sub-optimal nutrition with micro-deficiencies in many vitamins and mineral supplements should be recommended for all but those whose meal-time behavior approaches normal. Zinc deficiency may be associated with sensory blunting, poor appetite, and disturbed bowel functions and perhaps with aggressive behavior so the supplementation should often be considered. Autistic children may have low plasma fatty acids and supplementation with evening primrose oil or fish oil can do no harm. Non-specific or placebo like benefits seem to occur with almost any dietary improvement or modification.

Gluten Free, Casein Free Diet (GFCF): A very popular dietary treatment for symptoms of autism is removal of gluten (a protein found in barley, rye, oats, and wheat) and casein (a protein found in dairy products),in what is known as a Gluten Free, Casein Free diet, or GFCF. This is based on the hypothesis that these proteins are absorbed differently in children with autism spectrum disorders and act like false opiate-like chemicals in the brain. There are ongoing studies to prove the effectiveness of this dietary intervention. However, many families report that dietary elimination of gluten and casein has helped to control bowel habits, sleep activity, habitual behavior and enhance the overall progress in their children.

**Pharmacotherapy in Autism:**

Current psychopharmacologic trials are underway to investigate efficacy of a variety of classes of agents on promoting social interactions and reducing disruptive behaviour in children and adolescents with autism and other pervasive development disorders. Currently, no specific medications with proved efficacy in the treatment of the core symptoms of autistic disorders are available; however, medications have been shown to be promising in reducing hyperactivity, obsessions and compulsive behaviour, irritability, aggression and self - injurious behaviour.

The administration of antipsychotic medication has been shown to be efficacious in the reduction of aggressive and self-injurious behaviour. One early study indicated that haloperidol reduced the behavioural symptoms such as hyperactivity, stereotypes, withdrawal, fidgetiness, irritability and labile affect and accelerated learning. Given its potentially serious adverse effects, haloperidol is no longer the antipsychotic agent of choice in the treatment of self-injurious behaviour in children with autistic disorders.

The atypical antipsychotic agents are known to have a lower risk of causing extra pyramidal adverse effects, although some

sensitive individuals cannot tolerate the extra pyramidal or anticholinergic adverse effects of the atypical antipsychotic agents. The atypical antipsychotic agents include risperidone, olanzapine, quetiapine, clozapine and ziprasidone.

Risperidone, a high-potency antipsychotic with combined dopamine D2 and serotonin 5 - HT2 receptor antagonist properties, has been used to subdue aggressive or self - injurious behaviour. Several reports have suggested that risperidone is effective in diminishing aggressiveness, hyperactivity and self-injurious behavior in children with autistic disorder. In some cases, it reportedly encouraged socially acceptable behaviour. The U.S. Food and Drug Administration has approved Risperdal as autism medication to treat irritability in autistic children and adolescents. This is the first time the FDA approved a drug to treat behavior-related problems associated with autism in children. The drug can be used to treat aggression, deliberate self-injury and temper tantrums. Risperdal is considered an atypical antipsychotic drug manufactured by Janssen Pharmaceutica N.V.in Beerse, Belgium. For autism, lower dosages ranging from 0.5 to 4 mg per day are used in clinical practice. Extra pyramidal effects and akathisia have been reported adverse effects, as well as sedation, dizziness and weight gain. Drooling was reported more in the risperidone group compared with the placebo group. In this sample, extrapyramidal symptoms were not reported more commonly in the risperidone group. The side effects that caused the most concern were somnolence and weight gain. Olanzapine specifically blocks 5- HT 2A and D2 receptors and also blocks muscarinic receptors. No studies provide specific guidelines regarding the use of olanzapine in children with autism. Dosages that have been used clinically to target aggression and self-injurious behaviour range from 2.5to about 10 mg per day. Among olanzapine's most common adverse effects are sedation, orthostatic hypotension, and weight gain.

Clozapine has a hetrocyclic chemical structure that is related to certain conventional antipsychotics, such as loxapine, although clozapine carried lower risk of extrapyramidal symptoms. It is generally used in treatment of aggression and self-injurious behaviour unless those behaviour coexist with psychotic symptoms. It's most serious adverse effects are agranulocytosis, which necessitates monitoring white blood cells count weekly during clozapine's use. Its use is generally limited to treatment-resistant psychotic patients. Lithium can be administered in the treatment of aggressive or self - injurious behaviour when anti-psychotic medications fail. Psychotropic Medication: Psychotropic medication is commonly used to treat disruptive behaviors, agitation, in attention and hyper-activity in children with ASD.

| Class | Medication (Brand name) | Level of Evidence | Target Symptoms | Significant Potential Side Effects | Studies |
|---|---|---|---|---|---|
| Antipsychotics | Risperidone (Risperidal) | Established Evidence | Irritability, hyper-activity, and stereotypy | Weight gain, drooling, dizziness, fatigue, involuntary muscle movement | (Jesner, Aref- Adib, &Coren, 2007; McDougle, et al; 2005; Miral, et al; 2008; RUPP, 2002) |
| | Haloperidol (Haldol) | Established Evidence | Aggression | Tardive dyskinesia, sedation, irritability | (Anderson, et al; 1989; Anderson,et al; 1984) |

| | | | | | |
|---|---|---|---|---|---|
| Stimulants | Methylpheni-date (Ri-talin) | Established Evidence | Hyperactiviy | Social withdrawal, irritability, agitation, stereotypy | (Handen, Johnson, &Lubetsky, 2000; Quitana, et al;1995) |
| Norepineph-rine Reuptake Inhibitor | Atomoxetine HCI (Strattera) | Preliminary Evidence | Attention deficit, hyperactivity | None | (Arnold, et al., 2006) |
| Alpha 2 Agonist | Clonidine (Catapres) | Preliminary Evidence | Hyperactivity, irritability, inappropriate speech, stereotypy, oppositionality | Drowsiness, low blood pressure, irritability | (Jaselskis, Cook, Fletcher, & Leventhal, 1992) |
| | Guanfacine (Tenex) | Insufficient Evidence | Hyperactivity, inattention, impulsivity, aggression | Transient sedation | (Posey, Puntney, Sasher, Kem, &Mcougle, 2004) |
| Selective Serotonin Reuptake Inhibitors (SSRIs) | Fluoxetine (Prozac) & Citalopram (Celexa) | Insufficient Evidence (conflicting results) | Repetitive behavior | Celexa: Hyperactivity, insomnia, inattention, impulsivity, diarrhea, dry skin | (Hollander, et al., 2005; King, et al., 2009) |
| Others | Clomipramin e (Anafranil) | Preliminary Evidence | Stereotypy, ritualistic behavior, social behavior | Insomnia, constipation, twitching, tremors | (Gordon, State, Nelson, Hamburger, & Rapoport, 1993) |
| | Valproic Acid (Depakote) | Insufficient evidence | N / A | Rash, weight gain,hair loss, fatigue | (Heillings, et al., 2005; Hollander, et al; 2006) |
| | Naltrexone (Revia) | Insufficient evidence | N / A | Increased stereotypy | (Willemsen-Swinkels, Buitelaar, Weijnen& van Engeland, 1995) |

**Medical Management of Autism**

The established therapies for autistic disorder are nonpharmacologic and may include individual intensive interventions.

Individuals with autism spectrum disorder and unspecified pervasive developmental disorder typically benefit from behaviorally oriented therapeutic programs developed specifically for this population.

Autistic children should be placed in these specialized programs as soon as the diagnosis is suspected.

**Nonpharmacologic therapy**

- Intensive individual special education
- Speech, behavioral, occupational, and physical therapies (e.g., assisted communication, auditory integration training, sensory integration therapy,

exercise/physical therapy)

- Social skills training in some children with autism spectrum disorder, including those with comorbid anxiety disorders; children with autism spectrum disorder and comorbid ADHD may benefit less from social skills training

**Pharmacotherapy**

No pharmacologic agent is effective in the treatment of the core behavioral manifestations of autistic disorder, but drugs may be effective in treating associated behavioral problems and comorbid disorders (e.g., self-injurious behaviors, movement disorders). The possible benefits from pharmacotherapy must be balanced against the likely adverse effects on a case-by-case basis (e.g., venlafaxine may increase high-intensity aggression in some adolescents with autism

Medications used in managing related behavioral problems and comorbid conditions in children with autism include the following:

- Second-generation antipsychotics (e.g., risperidone, aripiprazole, ziprasidone)
- SSRI antidepressants (e.g., fluoxetine, citalopram, escitalopram)
- Stimulants (e.g., methylphenidate)

**Occupational therapy management**

Occupational therapy intervention revolves around analysis of behavior and performance in context to the occupation the person engages in. It is a person-centered approach in which the person has the necessary resources for adaptation, growth and change and needs a therapist – person guided interaction and treatment. In children and adults with autism, an occupational therapist works on remediation of sensory dysfunction, compensatory techniques to avoid activity or participation difficulties and prevention of secondary problems. Treatment is based on the approaches of sensory integration, occupational role and functioning, neurodevelopmental therapy, sensory diet approach, social communications approach, behavior modification, auditory integration training and play therapy.

**Assessments**

A holistic view of the person's abilities and needs are required so there is a need for combination of standardized evaluations and observations of behavior to be used. It is also necessary to observe clients in variety of settings and over many sessions in order to learn about how the person perceives sensory input and his/her individual sensory preferences. Videotaping the child across different environments like at school, home, garden etc. can be used to allow these observations.

**Main Areas to assess:**

**Sensory skills:**

- Sensory processing
- Registration
- Modulation
- Discrimination

**Motor skills:**

- Muscle tone
- Gross and fine motor skills
- Postural control
- Balance
- Motor planning/ Praxis

**Occupational skills:**

- Activities of daily living like grooming, dressing, eating etc.
- Play and learning
- Presence of stereotyped behaviors which interfere with daily tasks

**Cognitive skills:**

- Visual perceptual skills
- Problem solving
- Reasoning
- Attention and concentration
- Academic skills
-



*Fig 4. Supine flexion*



*Fig 5: Prone extension*



*Fig 6: One leg standing*

**Assessments:**

**Commonly used assessments for sensory processing:**

1. SIPT (sensory Integration and Praxis Test): It is developed to identify sensory integration problems. Test is standardized for use in children 4 –8: 11 years of age. SIPT assesses sensory and neurological process, which leads to behavioural, learning, language and praxis problems. It consists of 17 subtests, which assess tactile, vestibular proprioceptive processing; form and space perception, visual-motor coordination, praxis, bilateral integration and sequencing, and it takes nearly 90 minutes to complete the test.
2. Sensory Profile: It is a caregiver questionnaire, which measures children's responses to sensory events in everyday life for children 3-10 years of age. 5-point likert scale (nearly never, seldom, occasionally, frequently, almost always) is used for assessment.
3. Sensory rating scale: It wasdeveloped as a parent report measure that can be used to identify and quantify sensory responsiveness, including sensory defensive behaviours, in the birth to 3-year-old population.
4. Southern California Sensory Integration Test (SCSIT): It is developed by Ayres for use in physically and mentally normal children with specific learning disorder. It is standardized for children 4-8:11 years of age. It takes nearly 75-90 minutes to complete the test.
5. Sensory Processing measure: It provides a complete picture of children's sensory processing difficulties at school and at home. SPM-P extends the popular SPM down to age 2, making early intervention possible. Meant for 5 to 12 years for SPM; 2 to 5 years for SPM-P.

6.Evaluation of sensory processing: This questionnaire is intended to identify behaviors thought to be indicative of sensory processing problems.

A child with autism has qualitative social interaction and communication impairments, demonstrating low tolerance to sensory stimuli, or be distracted despite use of reinforcers, we need to supplement assessments with clinical observations.

**Treatment approaches**

**Sensory integration**

Sensory and motor dysfunction has been widely reported in persons with autism. Sensory processing problems are reported in more than 80% children with autism. The theory of sensory integration is an important and most widely used frame of reference in occupational therapy. It was developed by A Jean Ayres, an occupational therapist with advanced training in neuroscience and educational psychology. The sensory integration theory explains the relationship between the deficits in interpreting the stimulus from the body and environment and difficulties with learning.

Sensory integration is a process of organizing sensory information in brain in order to create an adaptive response. The aim of sensory integration therapy is to provide controlled and meaningful sensory experiences so that the child can spontaneously and appropriately form responses that require integration of those sensations

**Sensory integration therapy is driven by four key principles:**

- The child must be able to successfully meet the challenges that are presented through playful activities (Just Right Challenge);
- The child adapts his/her behavior with new and useful strategies in response to the challenges presented (Adaptive Response);
- The child will want to participate because the activities are fun (Active Engagement); and
- The child's preferences are used to initiate therapeutic experiences within the session (Child Directed)

Sensory integration therapy is based on the assumption that the child sensory systems are either over stimulated or under stimulated by the environment. It's reported that sensory processing problems observed in individuals with autism are associated with behavioral and/or functional performance problems and specifically stereotypical or repetitive are associated with self-calm or sensory seeking. Different sensory modulation problems such as hypo-responsivity, hyper-responsivity, sensory avoiding or sensory seeking are reported in children with autism. The vestibular, proprioceptive and tactile sensory systems integrate the incoming and outgoing information to the whole brain and helps define the world. Intervention is directed depending upon whether the child is hypersensitive or hyposensitive to a particular stimulus.

- **Vestibular system:**

The sensory system that interprets body position and spatial orientation depending on the position of the head. It maintains the body's balance against gravity.

All head movements and positions (spinning, swinging, rolling, jumping upand down, running) are instrumented by the vestibular system. Stimulus from these systems may not be properly received, synchronized, accurately interpreted or modulated. Hence the child might lack proper spatial orientation or feeling secure as if he is going to fall down, experience light-headedness/dizziness, feel dazed or

disoriented, or have low muscle tone.

Even certain autism-related behaviour that are repetitive or stereotyped in appearance,like spinning,rocking or toe-walking, may actually be that brain's way of trying to modulate the body.So difficulties in processing vestibular input may cause a child to feel gravitational insecure (having a low tolerance for movement, difficulty changing direction and speed, maintaining any other position other than being standing with feet on floor), difficulty in copying from the board or lose their place when reading, may have to move entire body to look around rather than turn only head when called.

Strategies will involve vestibular stimulation in linear, angular, rotatory and orbital planes. Slow rhythmic linear movement within limited ranges are for calming. Eg: Slow rocking, swinging, car seat in moving car, slow bouncing on a ball. Fast dysrhythmic angular, orbital, and rotatory movement within wide ranges are for alerting Eg: Jumping on a trampoline, fast Swinging on frog swing.

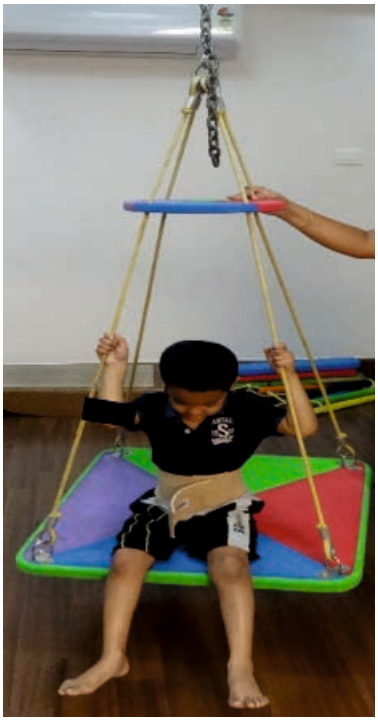


*Fig 7: Swinging*



*Fig 8: Propelling a scooter board*

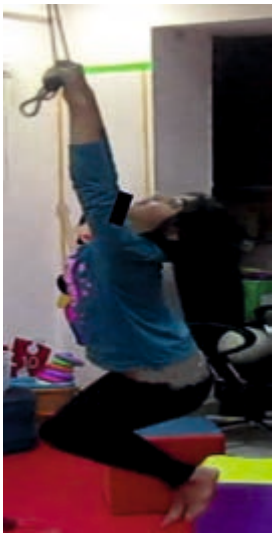


*Fig 9: Hanging from a trapeze*



*Fig 8 Balancing on a balance board*



*Fig 9 Jumping on trampoline*

**Examples of vestibular activities**

- **Proprioceptive system:**

It is the system continuously working with the vestibular, tactile and visual systems, telling the brain where the body is and limbs are, what they are doing, what is needed and why. Both vestibular and proprioceptive systems are internal monitoring systems receiving and transmitting sensory information from different systems to the brain. Muscles and joints are the main receptors of the proprioceptive system. Its functions help increase motor control and planning, moving body in an efficient manner and organization of all sensations. Difficulties in processing of proprioception result in poor body awareness, being uncoordinated /clumsy, problems with balance, slapping feet while walking, chewing on inedible objects, difficulty in grading pressure so either using too much or too little pressure.

Moderate to deep pressure to whole body or proximal joints will help in organizing. Eg. Swaddling in a bed sheet, sustained joint compressions, sandwich between pillow/mattresses, pull and push activities, lifting and carrying groceries.




*Fig 10 Examples of deep pressure (Proprioceptive activities)*

Tactile: Touch stimulus is processed through two main channels: the protection sense and discrimination working together in order for the body to interact in a functional, calm, safe and adaptive manner (music). Efficient processing of the tactile sense is necessary for motor planning, body awareness, academic learning, emotional security and social skills (21). If the child is not able to process tactile information it may result in oversensitivity to sudden / light touch/certain food textures / temperatures, flight/fight response to harmless touch, seeks increased touch sensations like touching people and objects, chewing on inedible objects, physically hurting other people or pets, not understanding their pain. A child may have trouble with fine motor skills needed for feeding, dressing, writing etc.

Slow, moderate to deep skin pressure, on back, dorsolateral surfaces, face will have calming effect like slow massage, stroking in one direction.

Rapid moving, light touch, over ventromedial surfaces of extremities, periorbital area causes alerting or excitatory response. Eg. Playing in a ball pool, light brushing to periorbital area.



*Fig 11 a. sand play, Fig 11.b Grain play -examples of tactile activities*

- **Visual:**

Vision is needed for looking at sights, identifying them and perceiving what is approaching and taking an appropriate

response. It contributes to occulomtor skills and with other senses helping visual spatial skills develop.

Difficulties in processing result in reading, failure to visualize when reading, confusion between left/right, up/down, before/after, turning of head when reading across a page, squint/cover eyes, complaints of seeing double etc. Vision therapy or visual training will help integrate visual information with input from other systems.

Low illumination in pastel colors, in small spaces, with reduced clutter helps when the child is excited with increased visual input. Eg. Playing in a small room tent, bare or while colored walls.

Moderate lighting, bright colors, open spaces and lot of visual effects are excitatory or arousing. Eg. Working in a bright, sunny area, bright color, animation on walls

- **Auditory:**

The hearing system located in the inner hear provides information about sounds in the environment e.g. How loud/soft/high/low/near/far the sound is. Difficulties in processing auditory information may appear like closing ears to certain sounds (like mixer, flush, whistle of pressure cooker, being unaware of the source of sounds , distressed with loud sounds, difficulty in reading aloud, problems in articulation etc. Auditory integration therapy is a process of providing sound stimulation designed to help with improved listening, filtering and processing sounds

Slow rhythmic, music has an inhibitory or calming effect. Eg. music with heartbeat rhythms, nursery rhymes, instrumental music. Fast arrhythmic loud lyrical music tends to be arousing. E.g. loud driving beat music, drums.

- **Olfactory and Gustatory:**

Smell and taste are closely related. Taste perception depends on smell processing. Difficulties in processing smell and taste may present as avoidance in eating of certain textures of food, smelling food before eating and refuse to eat, resist brushing his teeth or going to the dentist, dislike sticky foods or ones with lumps.

Sweet, mild, bland flavors, warm temperatures, smooth textures, sucking, blowing and biting will help for calming. E.g. sucking thick liquids through a straw, pacifier, chewy tube, munch on carrots, potato chips. Sour, spicy, bitter, Smokey flavors, cold temperatures, hard/chewy textures, chewing or licking can be used for alerting. E.g. lemonade, sour candy, Popsicle/lollypops. Crunchy and chewy foods encourage concentration. NUK knobby toothbrush, water pick, electric toothbrush, etc. can be used.

**Praxis:**

Occupational therapists using a sensory integration frame of reference identify dyspraxia, or a deficit in the process of praxis, as the underlying mechanism for the motor performance problem. Jean Ayres explained motor performance as requiring an idea and the organization of a plan of action for executing a motor action. Praxis is composed of:

- Ideation (ability to conceptualize and identify a motor goal and some idea of how to achieve the goal);
- Planning (ability to plan and organize a series of intentional motor actions in response to environmental demands), and
- Execution (the ability to perform motor responses with precision)

It helps with the ability to produce an adaptive response to environmental demands. It combines new actions using knowledge of past experiences and the sensations that accompany them to assist in optimum learning in a child with praxis

difficulties. Unusual play behaviours seen in children with autism may be related to deficits in praxis and executive control.

**Commonly seen difficulties in a child with dyspraxia are:-**

- Poor eye hand coordination
- Unable to learn new tasks
- Having trouble planning ,organising and sequencing body movements like when riding a bicycle
- Poor gross motor control for climbing, jumping , running etc.

**Strategies to improve praxis:**

- Obstacle course
- Animal walks
- Hopscotch
- Skipping - With ropes and hoops



*Fig 12 - Obstacle course*



*Fig 13 - Hopping*



*Fig 14 - Transfer*



*Fig 15 - Coming out of a barrel*

**General precautions to be followed:**

- Monitor for signs of sensory overload as overstimulation can cause disorganized behavior. Pairing proprioception with vestibular input will improve organization.
- Keep therapy rooms free of sharp corners and unsafe objects
- Keep mattresses below suspended equipment
- Ensure suspended equipment is properly fastened into walls or ceiling
- Use only specifically designed oral toys to be put in the mouth
- Use edible stuff for fine motor tasks like cereal pieces if the child has a habit of putting things in the mouth

- Avoid brushing or light touch stimulus over ventral trunk area
- Avoid high stimulus intensities that can cause autonomic shutdown

Neurodevelopmental therapy (NDT) is needed for individual with postural and tonal difficulties such as low tone, unusual weight bearing patterns, clumsiness, inadequate postural control and balance. (quick) Combining Sensory integration and Neurodevelopmental therapy can be combine and used in children with autism as both are based on principles of normal development and neural functioning. Both recognize the plasticity of brain and emphasize the use of sensory input to elicit changes in motor output.

**Sensory diet**

Sensory diet approach will help for meeting arousal needs. An occupational therapist will assist in providing a variety of activities and adaptations based on a person's individual sensory needs. It will vary according to the person's goals, preferences, resources and limitations. The therapist has to understand the sensory difficulties the person has and how it is affecting his/her daily activities and educate the caregivers/parents. The activities have to be well-matched with the person's and family's beliefs and can be adjusted into their routines. It can be used as an addition to therapy sessions or used independently. The therapist should suggest cost effective equipment that parents can make or buy to use at home to provide adequate stimulation to the child.

**The benefits of a sensory diet are:**

- Reduce sensory deprivation, self stimulatory behaviors or over stimulation
- Decrease sensory defensiveness
- Improve self regulation
- Increased postural control
- Enhanced body awareness and schema
- Improve mood and arousal level for increased participation in daily activities

The sensory diet is based on the theory that enhanced inputs provide profound adaptive functioning.

**Play:**

Play is the primary occupation of children which promotes social participation. It needs to be encouraged when not already developed. Many children with autism struggle in the areas of engagement with persons and objects, pretend play, turn taking, following rules and paying attention.

For occupational therapists, play is a therapeutic medium for evaluation and intervention. An interview with caregivers/parents gives the therapist an idea about the child's play development and interests. Test of Playfulness , Knox Preschool Play Scale, Play History and Observations of Factors Influencing Playfulness Form can be used by occupational therapists to plan their interventions.

Intervention involves: Age and skill appropriate toys needs to be selected. However developmental age appropriate toys are better suited than chronological age ones as they may be advanced for some children with autism. The area selected for the child to play should make him/her feel safe and comfortable. Working on imitation and praxis helps build the foundation for engagement in play. Modeling what is expected of the child helps in understanding the outcome and reduces anxiety in new activities. Toys can be modified or carefully selected when there is a mismatch in skills and preference for a particular toy. Simple language needs to be used to reduce the demand on the child. Using schedules to arrange a sequence of activities to ensure predictability and easy transitions are

helpful. Group therapy sessions facilitate turn taking, waiting for turns, and increase in comfort level being around other kids, sharing and copying others. Having play routines and play rituals.

**Auditory integration training:**

Auditory sensory difficulties and sensitivities are often mentioned by parents of children with Autism Spectrum Disorder (ASD), and in research involving children with autism. Auditory Integration Training (AIT), The Tomatis Method, Therapeutic Listening (TL), and the Listening Program (TLP) are some of the treatment programs available.

Some auditory treatment methods are delivered using specialized equipment in a clinical setting (such as Auditory Integration Training (AIT)) and other treatments are delivered via specially-engineered compact-discs that can be listened to through high-quality headphones in the home (such as Therapeutic Listening (TL)).

These programs attempt to address discrepancy in environmental awareness, body awareness and communication by providing stimulation to the physical and neurological structures of the ear. In theory, this will enhance sound awareness to affect environmental awareness; sound discrimination and sound processing to improve speech and language processing; and alertness and modulation of brain activity to increased body awareness. These techniques use electronically-altered sounds and music to accentuate various frequencies and encourage the muscles of the ear to control the dampening mechanisms to receive better and improved sound.

This is believed to: adjust physical and neurological structures of the ear and brain to frequencies and harmonics that have been disregarded (e.g. one who does not attend to another person's vocal pitch), desensitize the ear to frequencies one finds unbearable (e.g. one who has auditory defensiveness), and/or to increase the ear and brain's awareness of sound localization, thus increasing one's environmental awareness and ability to learn.

Therapeutic Listening involves specially modulated music with specific frequencies filtered out. The patients listen to this twice daily through special headphones which allow them to hear ambient sound and perform other task. It can be used as a home-based program also, allowing for optimum use of the protocol.

A Therapeutic Listening program is set up by a specially trained occupational therapist and typically lasts 8- 12 weeks. This may be done in either the clinic or home settings. Auditory information from the specialized music provides input to both the vestibular and the auditory portions of the vestibular-cochlear continuum. Individuals with autism, sensory integration dysfunction, and learning difficulties may benefit from this intervention. Studies have shown that children who are involved in the Therapeutic Listening program in addition to a traditional sensory diet have better outcomes in standardized testing performance. As with all other treatments for the human nervous system, results will be individualized. Neurodevelopmental improvements have been documented with this therapy modality. Increased self regulation, motor coordination, praxis, sleep wake patterns and sensory defensiveness are just a few of many outcomes that have been seen with this modality. Handwriting abilities and academic scores have also been noted to improve for individuals with specific learning disabilities.

**Assistive technology:-**

According to the Assistive Technology- Act of 1988, an assistive technology means any item, piece of equipment, or product system, whether acquired commercially, off-the-

shelf, modified or customized, that is used to increase, maintain, or improve functional capabilities of individuals with disabilities. Assistive technology service is any service that directly assists an individual with a disability in the selection, acquisition, or use of an assistive technology device.

It is divided into:-

- **"Low" Technology**

It includes visual support strategies which do not involve any type of electronic or battery operated device - typically low cost, and easy to use equipment. Example: dry erase boards, clipboards, 3-ring binders, manila file folders, photo albums, laminated PCS/ photographs, highlight tape, etc.

- **"Mid" Technology**

Battery operated devices or "simple" electronic devices requiring limited advancements in technology. Example: tape recorder, Language Master, overhead projector, timers, calculators, and simple voice output devices.

- **"High" Technology**

Complex technological support strategies - typically "high" cost equipment. Example: video cameras, computers and adaptive hardware, complex voice output devices.

The use of assistive technology should always be individualized to meet the unique needs of any child with autism. Most importantly, use of these varied modes of technology will greatly increase the child's independent functioning skills by decreasing the amount of direct support needed from another person.

**Activities of daily living:**

Performing personal activities of daily living (like bathing, toileting, dressing, eating, and functional mobility) are some of the most important skills children learn as they grow. Not only taking care of their own personal care needs and general health and well-being, but they are required to perform self-care activities in ways that are constant with societal and cultural norms, In addition to that, children develop and perform home management, life skills and community living skills in preparation for adulthood. These are referred to as instrumental activities of daily living.

Individuals with autism generally have the risk of limited engagement in activities. Studies show that such individuals engage in less activities less frequently than others and have weaker daily life skills when compared with individuals with other developmental disorders or with typical development.

**The following scales are used for assessment purpose in finding out deficits in daily activities:-**

- The Functional Independence Measure for Children (WEFIM),
- Waisman Activities of Daily Living (W-ADL) Scale,
- The Pediatric Evaluation of Disability Inventory (PEDI),
- The Vineland Adaptive Behaviour Scales,
- The Assessment of Motor and Process Skills (AMPS),

In the initial stage of intervention, therapists use activity analysis to define which steps of activity process are problem areas and define behavioral and educative methods to be used. Behavioral approaches have been used, often in combination with other approaches, to obtain a baseline of the child's behaviors and as a way of establishing clear goals. Behavioral methods like reinforcement and token systems are useful in attainment and sustainment of skills in individuals with autism. Prompts are important to improve the quality of performance and to brief individual on what to do in the next step. As individuals with

autism may experience difficulties in understanding natural cues and verbal communication, context or activity demands such as use of prompting can be modified.

One of the important training methods used in ADL training is chaining. Training of complex skills generally requires breaking down the steps and linking the steps of the task. Three chaining options are available for functional task training:

- Backward chaining in which the last step of the task is trained first, followed by the second to last step and the last step, and so on, progressing backward through the chain,
- Forward chaining in which the first step of the chain is trained first, followed by the first and second step, and so on, progressing forward through the chain
- Whole task method in which each step of the chain is trained on each presentation.

Repetitions and development of habits and routines are required to sustain the skill after it is learned and to reduce environmental supports. Furthermore, generalization of skill in different environments should be emphasized.

Compensatory strategies may include modification of task or task method, use of assistive technology or modification of environment.

**Reflex integration:**

Reflexes are our genetically determined motor-behavioural patterns that are normally integrated by every child into consciously controlled sensory-motor abilities and skills during early childhood.

These integrated reflexes then support the neurodevelopment of a child assuring an increase in processing sensory input, programming, and control of motor and behavioural actions, and enhanced learning and memory, language and communication skills.

People diagnosed with ASD have a lack of sensory motor integration and delay of performance concerning the early motor development. They show a wide range of immature reflex patterns such as Hands Pulling, Hands Supporting, Hands Grasp, Crawling, Asymmetrical Tonic Neck Reflex, Symmetrical Tonic Neck Reflex, Babkin Palmomental, Ocular-Vestibular, and other patterns. Further training in reflex integration programs will equip the occupational therapist with working with integration of primitive reflexes.

**School based Occupational therapy:**

Occupational therapists work in schools to develop educational skills of the student with autism. The occupational therapist evaluates, assesses and accommodates functional abilities of the student with autism in school classrooms, hallways and other related education areas. The occupational therapist works with teachers to help students acquire functional abilities necessary to access and use educational materials and be independent in the school. Occupational therapists work with the students with autism on adapting or modifying school equipment/materials to help them function better in classrooms, the canteen, washrooms and in recess breaks. Other assistance includes helping students with autism to be sensory steady and participate in activities outside of the school through mobility on outdoor trips, sports events, on playgrounds and within the neighborhood. Students with autism face a demanding environment at school. Presentation methods for educational materials must be modified to meet the challenges of students with autism, such as their ability to communicate, view and manipulate educational materials, and move about the school.

Occupational therapists work closely with

teachers to promote the highest level of function possible for a child with autism pursuing educational goals like fine and gross motor skills and attention skills.

Occupational therapists can use direct or indirect services for students with autism in school settings. Occupational therapy interventions purposes are to help students with autism to gain

independence in daily living activities, feeding and oral functions, play skills, task organization and completion, written communication skills, hand function, sensory integration (processing), visual perception, school mobility, participating on a regular and timely basis, using tools and supplies, participating in activities throughout school settings. Also when skill and strength cannot be developed or improved, occupational therapy offers creative modifications and adaptations for carrying out development-appropriate activities. In school practice, occupational therapists work with students, teachers, families and schools. Occupational therapists are the experts who will identify ways to engage students with autism in educational activities and supporting them to develop competence in their roles as students.



*Fig 17 Form boards*



*Fig 18 Puzzle*



*Fig 19 3 D to 3D design copying*



*Fig 20 2D to 2D design copying*

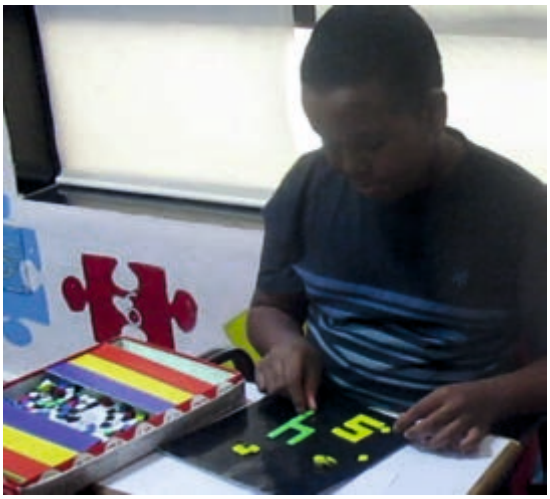


*Fig 21 Magnetic puzzles (Examples of visual perception activities)*



*Fig. 22 cutting paper*

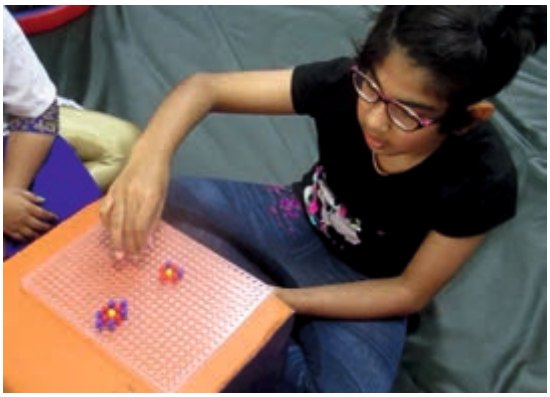


*Fig. 23 Mosaic pegs*



*Fig. 24 Lacing*



*Fig. 25 Painting*

**Vocational training:**

Occupational therapists have varied roles in assessing, recruitment and training, work development and supportive employment process stages of employment process of individuals with autism. During the initial stage of assessment occupational therapists assess sensory and motor skills, professional interests, communication and transportation, daily living activities, cognitive, social and community skills, emergency response and safety in workplace and self-assessment skills.

During the next stage of recruitment and training occupational therapists work on training and adaptations about work task

and settings, identification and training on secondary equipment and technology, training on or compensation of basic cognitive skills like attention, visual perception etc., training of helpers to assist with training of individuals.

During the work development stage occupational therapist work for increasing environmental support and reducing barriers, doing task analysis, adjusting improvement of compliance between skills of the individual and expectations of the employer and work development in line with interests and skills of individual.

In the supportive employment process stage occupational therapist interested in training of work behaviours, training on complex cognitive skills such as problem solving, time management, advocating rights of the individual against the employer and other colleagues and cooperation with interdisciplinary team.

**Role of Psychology in Autism:**

Diagnostic and Assessment Instruments For Children with Autistic Spectrum Disorder:

The below listed scales and tool are used by psychologists, educators and researchers to assess children suspected of or previously diagnosed with a pervasive developmental disorder. The instruments listed below are used to measure specific dimensions of a child's development in different domains of functioning, which would help develop a proper intervention plan for the child.

**Diagnostic Assessment:**

Autism Diagnostic Interview-Revised: This is a semi-structured, investigator-based interview which is useful for diagnosing autism, having an intervention plan and distinguishing autism from other developmental disorders. The ADI-R has proven particularly effective in assessing syndrome boundaries, identifying new subgroups, quantifying Autistic symptomatology, and determining the clinical needs of groups in which a high rate of Autism Spectrum Disorders might be expected, such as people with severe language impairments. It is conducted on children and adults with a mental age of above 2 years. It takes approximately 1 1 / 2 to 2 1 / 2 hours, including scoring.

Prelinguistic Autism Diagnostic Observation Schedule: The Prelinguistic Autism Diagnostic Observation Schedule(PL-ADOS) is a semi-structured observation scale for diagnosing children whose speech is not yet developed and who are suspected of having autism. This instrument provides an opportunity to observe specific aspects of the child's social behavior, such as joint attention, imitation, and sharing of affect with the examiner and parent.PL- ADOS scores are reported to discriminate between children with autism and children without autistic developmental disabilities.

Childhood Autism Rating Scale Second Edition (CARS- 2): This scale helps to distinguish children from developmentally handicapped children who are not autistic. It helps distinguish between mid to moderate to severe autism. Standard Version Rating Booklet (CARS2-ST) is equivalent to the original CARS; for use with individuals younger than 6 years of age and those with communication difficulties or below-average estimated IQs. High- Functioning Version Rating Booklet(CARS2-HF) is an alternative for assessing verbally fluent individuals,6yearsofageandolder,withIQscores above 80. Questionnaire for Parents or Caregivers (CARS2-QPC) is an unscored scale that gathers information for use in making CARS2-ST andCARS2-HFratings.TheCARS2-STandCARS2-HF each include 15 items addressing the following functional areas like: relating to people, social and emotional understanding, imitation, body use, visual responses, verbal communication, taste, smell, and touch

response and use, etc. It is conducted on children 2 years and older and the approximate administration time is 5 to 10 minutes.

Checklist for Autism in Toddlers(CHAT): This is a screening instrument designed to detect core autistic features to enable treatment as early as 18 months. CHAT offers psychologists a means of diagnosing autism in infancy so that educational programs can be started months or even years before most symptoms become obvious. It is a test which aims to look at behaviors which, if absent at 18 months, put a child at risk for a social-communication disorder. These behaviors are (a) joint attention, including pointing to show and gaze-monitoring (e.g. looking to where a parent is pointing), and (b) pretend play (e.g. pretending to pour tea from a toy teapot).

Gilliam Autism Rating Scale - Second Edition (GARS - 2): GARS-2, a revision of the popular Gilliam Autism Rating Scale, assists teachers, parents, and clinicians inidentifying and diagnosing autism in individuals ages 3 through 22. It also helps estimate the severity of the child's disorder. Items on the GARS-2 are based on the definitions of autism adopted by the Autism Society of America and the Diagnostic and Statistical Manual of Mental Disorders: Fourth Edition-Text Revision (DSM-IV-TR). The instrument consists of 42 clearly state ditems describing the characteristic behaviors of persons with autism. The items are grouped into three subscales: Stereotyped Behaviors, Communication and Social Interaction. Using objective, frequency-based ratings, the entire scale can be completed and scored in 5 to 10 minutes. A structured interview form is included for gathering diagnostically important information from the child's parents.

**Developmental Assessment:**

Psycho- educational Profile-Revised (PEP-R): This offers a developmental approach to the assessment of children with autism or related developmental disorders.

It is an inventory of behaviors and skills designed to identify uneven and idiosyncratic learning patterns. The test is most appropriately used with children functioning at or below the preschool range and within the chronological age range of six months to seven years. The PEP-R provides information on developmental functioning in imitation, perception, fine motor, gross motor, eye-hand integration, cognitive performance, and cognitive verbal areas. The PEP-Ralso identifies degrees of behavioral abnormality in relating and affect (cooperation and human interest), play and interest in materials, sensory responses, and language.

**Adaptive Assessment:**

Vineland Adaptive Behavior Scales - Second Edition (Vineland-II): This scale comes in three forms varying in degree of detail and proposed setting. The VABS is administered by interviewing the child's parents, teachers, or care-providers. The scales range in age from birth to nineteen years. It takes approximately 20 to 60 minutes for the conduction and scoring of the test.

**Standardized Tests of Intelligence:**

Wechsler Intelligence Scale for Children(3rded.): This is used to assess intelligence as a global but multifaceted entity that can be inferred from a child's performance on a series of tasks. It is valuable for psycho-educational assessment, diagnosis, placement, and planning. WISC-III can be used to diagnose exceptionality among school- aged children and has a strong place in clinical and neuropsychological assessment and in research. Like the WPPSI-R, the WISC-III is widely used and generally regarded as the best standardized measure of intelligence.

Differential Ability Scales: The Differential Ability Scales (DAS) (Elliott, 1990) [16] measures overall cognitive ability and specific abilities in children and adolescents. It is better suited for intellectually higher-functioning children with autism. The DAS assesses multi-dimensional abilities in children ages two years and six months to seventeen years and eleven months. It is administered individually and takes 45 to 65 minutes for the full cognitive battery. The achievement test takes 15 to 25 minutes to administer. The seventeen cognitive and three achievement subtests yield an overall cognitive ability score and achievement scores. The three achievement subtests are Basic Number Skills, Spelling, and Word Reading.

Stanford-Binet Intelligence Scale (4th ed.): The Stanford-Binet Intelligence Scale(4thed.) (SBIS-IV) (Thorndike, Hagen, &Sattler, 1986) is for individual's ages two years to adult. It provides scores in four areas: Verbal Reasoning, Abstract and Visual Reasoning, Quantitative Reasoning, and Short-Term Memory; and a Composite Score that is equivalent to the Wechsler Scales Full Scale IQ.

**Academic Screening:**

Wide Range Achievement Test 3 (WRAT3): The Wide Range Achievement Test 3 (WRAT3) measures reading, spelling, and arithmetic in persons from five to seventy-four years old. Two equivalent forms make pre- and post testing possible. The test takes 10 to 15 minutes to administer. The WRAT3 provides a good method for measuring basic academic skills in children who perform below their peers.

**Behavior Assessment:**

Achenbach Child Behavior Checklist: The Achenbach Child Behavior Checklist(ACBC) is for children four to eighteen years old and is completed by an adult informant. It has two major scales - externalizing and internalizing behaviors - each of which has four subscales. It has been used as a follow-up measure. The child's primary caregiver (in most cases, the client's mother) serves as the informant.

Analysis of Sensory Behavior Inventory (Rev.ed.): The Analysis of Sensory Behavior Inventory (Rev. ed.)(ASBI-R)(Morton & Wolford, 1994) is designed to collect information about an individual's behaviors as they are related to sensory stimuli. Six sensory modalities are assessed: vestibular, tactile, proprioceptive, auditory, visual, and gustatory- olfactory. Ratings can be made about both sensory- avoidance and sensory-seeking behaviors within each modality. Information obtained from this tool may be helpful in completing a functional analysis of behavior and in designing effective intervention strategies, including accommodations and reinforces for the individual.

**Psychological Intervention for Autism:**

The goals of psychological intervention for children with autism is to eliminate or reduce the undesirable behaviours and target the behaviours that will help the child integrate into schools, develop and meaningful social communication with peers and family members and increase the likelihood of maintaining an independent living as an adult.

To do this, treatment intervention aim to increase socially acceptable and pro-socially behaviour, to decrease or behavioural symptoms, and to improve verbal and non-verbal communication. It basically aims at increasing the level of independence and increasing the quality of life. Both language and academic remediation often required. In addition, treatment goal generally include a reduction of disruptive behaviour that may be exacerbated especially during translation and in school. Children with mental retardation need intellectual appropriate

behaviour intervention to be reinforcing socially acceptable behaviour and encourage self-careskills. In addition, parents, often distraught, need support and counseling. Insight oriented individual psychotherapy has proved ineffective. Educational and behavioural interventions are currently considered the treatment of choice. Structured classroom training, in combination with behavioural method, is the most effective treatment for many autistic children.

Well controlled studies indicate that the gain in the area of language and cognition and diseases in the maladaptive behaviour are achieved by consistent behavioural programs. Careful training of parents in the concept and skills of behaviour modification and resolution of the parents' concerns me yield considerable gain in children's language, and cognitive and social area of behaviour. These training programs, however, are rigorous and require much parental time. And autistic child requires as much structure as possible, and the daily program for as many hours as feasible is desirable.

**Behaviour Therapy:**

**Behaviour Therapy Plan:**

Identification: The behaviours which are undesirable and which need to be either replaced with a desirable behavior or reduced needs to be identified and noted down including the level of severity in it.

Gathering Information: Information need to be gathered from all the family members or caregivers, therapists and teachers who are taking care of the child, teaching or treating the child to understand the behaviours, the pattern of the behavior and the situations where and when the undesirable behaviours occur.

Observation: The child's behaviour needs to be observed in different situation as this helps to understand the response of the child in various situations and the level of acceptability of the behavioural plan.

Making a Behavioural Plan and Implementing: A behavioral plan needs to be formulated, where also the strategies that are to be used are to be listed down and it should be implemented. Along with it a close track needs to be kept on whether, the plan benefit in the client or is it further accelerating the client's undesirable behaviours. If it is accelerating the client's undesirable behaviors then, the techniques should be replaced with one which will help reducing the undesirable behaviors.

Reward System: A reward system needs to be set up, depending on what the child considers as a reward and something which in the long run would not lead to another issue. Examples of rewards which are safe and encouraging are praise, star on the hand, food items, etc.

BEHAVIOUR MANAGEMENT PLAN FOR AUTISM

Name of the patient: Date:

Age: Session Number:

Define the behaviour problem, severity and obstacles caused by it:

Hypothesis regarding the behaviour problem:

Identified skills to be taught to reduce the behavioral problems:

Antecedent strategies used to put in place to prevent the behaviours:

Hierarchy of consequential strategies to use when behaviours occur:

Reward system for good behavior:

*Fig 26: Behavior Management Plan*

**Traditional Approaches to Deal with Poor Behaviour:**

Aversives: Aversives are defined as the use of negative reinforcers to interrupt or stop problem behaviours as and when they occur.

Aversives are response to inappropriate behaviours and not a preventive measure for inappropriate behaviours. For example: When a child is being very hyperactive and is disturbing the class the teacher would direct the child to stand outside the class for 10minutes.

**Overcorrection:** This a type of aversive technique which requires the child to complete the appropriate behaviour several times because he did not do it when asked, or because he continued with the negative behaviour in spite of being asked to stop. For example:

Restraints: Restraints are defined as techniques that are used to "hold back" a child from doing some act which is harmful or dangerous to him. For example: If an autistic child has a self- injurious behaviour of banging his head hard on the table or the ground, then this behaviour can be prevented by having a custom made helmet for the child. This would either stop the child from banging his head or at least prevent from inflicting injury to himself.

**Behavior Modification:**

Behaviour Modification is the use of a collection of techniques that are compiled together to increase the number of desired behaviours and to extinguish or reduce the number of undesirable behaviours. Behaviour modification takes account of only those behaviours which are observable and measureable. The way to change an undesirable behaviour is to "target the behaviour" and then break them down into tiny pieces and steps. After which the tiny pieces of behaviors need to be practiced and rewarded till it is not mastered. Punishments like withdrawal of privileges or timeouts are used to reduce the number of undesirable behaviours. It is again important to remember that behaviour modification is a commonly used technique for handling behavioural problems and that it does not take into consideration the reasons for such behaviours. For example if an autistic child is exhibiting self-stimulatory behaviour because he is stressed, then the application of over correction may lead to replacing oneself-stimulatorybehaviourto another.



*Fig 27: Behaviour Therapy*

**Behaviour Strategies: Social Language Difficulty:**

Autistic children have social language inadequacy which due to which they may not be able to initiate a conversation or sustain it and it may lead to not having peers or peer group. Social skills deficits must be addressed in individualized educational plans, as they are the core symptoms of the autistic spectrum disorder.

The following are few listed strategies to improve social language:

1. Individualized training of facial expression and behaving in a socially acceptable manner would help grasp and understand better. Gradually then exposure to practicing this in a group would be really helpful.
2. Usage of drama and role playing to teach the voice inflection, modulation and facial expression is seen to be very helpful. It helps to reduce the fear of speaking in public and helps boosting the child's confidence.
3. Use videotapes and flashcards to imitate and practice facial expression. This

would also help to improve eye contact and help increase attention and concentration. For example: Expose the child to various expressions on a flash card and ask him to imitate the expression.

4. Peers and siblings can be trained to promote help the client maintain a naturalistic kid friendly conversation.
5. Teach the child how to initiate a conversation, respond and how to sustain it.
6. Also, a very important aspect that is poor in autistic children is maintaining eye contact. Help the child understand the importance and practice this by reminding the child to maintain eye contact while role-playing.

**Behaviour Strategies: Social Behaviour Difficulty:**

Autistic children have difficulty in displaying socially appropriate behaviours due to which they are usually bullied or may face rejection for people around or they themselves prefer to stay in isolation.



*Fig 28: Group Social Interaction*

**The following are few listed strategies to improve social behaviour:**

1. Arrange the situation for the child in such a way where, he needs to find clues to as certain how someone is feeling or predict how they will respond. He or she needs to be taught what various facial expression, emotions and how one's sounds in different emotions.
2. Use videotapes or television to reinforce learning about facial expressions and sounds of emotions.
3. Use role play, group therapy and practice scripts to respond to moments of teasing or being bullied.
4. Teach the child, the social rules and regulations which would help them behave appropriately to a situation.
5. Use stories with a moral and explain to the client the story step by step which would help him / her understand the behaviours that are acceptable socially.

For example: An autistic patient would have trouble invading someone's personal space, by positioning themselves very close to the individual they are listening to, which would place the speaker to be very uncomfortable with the close proximity. To avoid this vocal reminders need to be given and in spite of this if there are no changes then, a strategy to solve this would be to tie a rope around the child's waist, with leaving a 3 foot tail behind. Practice this with peers holding the tail and demonstrating to the client the meaning of 3 feet difference.

**Behaviour Strategies: Difficulties with Transitions and Changes:**

Autistic children yearn for sameness and routine. If there is a change in the ir routine, then they would try their best to stick to their routine or would throw temper tantrums. It's seen that children with autism work better and learn better with people who are more structured in their daily routine.

The following are few listed strategies to help autistic children to accept and adjust to transitions:

1. Teach children to adjust and accept

transitions by making small changes in their routine as and when they get very comfortable with it.

2. Dim the lights of the room, when a new activity is being presented to them.
3. Give prior explanation of the transition as getting them prepared before hand and understanding why a particular change is taking place would help the be prepared rather than having a flaring temper after the change has occurred.
4. Make the transition as enjoyable and ass tress free as possible because otherwise the child's autistic feature will shoot up.

**Behaviour Strategies: Problems with Self-Esteem and/or Depression:**

Often children with autism have a poor self-concept and lack confidence. This is normally seen to be present by the child having a poor eye contact, difficulty socializing or stammering. This is a very common feature in autism as these children undergo a lot of rejection from peers, they hear people around speaking about their behaviour and problems and being bullied.

**The following are few listed strategies to help boost self -esteem in autistic children:**

1. Keep a track of the child's behavioural achievements and encourage it by rewarding the child in front of the family members.
2. If the child is demonstrating poor or undesirable behaviours then do not take away his previously earned rewards.
3. Reward him / her for minor changes in the behaviour.
4. Be specific about the behaviours that are liked and encourage them with a reward. Behaviours that are undesirable need to be changed by desirable ones.
5. Phrases and words like "good boy" or "bad boy" should not be used rather "You followed the direction well" should be used to help increase the confidence.
6. The child's strengths should be motivated and ways to enhance it should be used. For example: A child who enjoys music should be encouraged to learn an instrument and excelling at playing it.
7. Encourage the client to maintain a photo book of his achievement or good behaviours. For example: If he / she won some competition or made some clay model, take a picture and keep it in front of the child.
8. When an autistic child is using negative words, phrases or sentences referring to himself encourage the child to make changes in his / her dialogues to a more positive view about himself /herself.
9. The child's grades, health concerns and short comings should be kept confidential and not publicized about.

Behaviour Strategies: Managing Stress and Anxiety:

Helping children with autism managing stress and anxiety is extremely important in preventing undesirable behaviours.

1. Maintain a time-table for all the daily activities and keep a reward system for it.
2. Teach the child breathing exercises, help him understand when to use it and practice it, so that the child is aware that he has to use it when he is stressed and anxious.
3. Exercise and play session is extremely important for the child, as it would help the client relieve his /her stress.
4. Self-talk method should be taught to the client and he should be also taught that this should be only practiced when he is alone in the room.

5. The client should have some hobby which he or she enjoys and feels relaxed when taking it up.

For example: An autistic patient would have trouble invading someone's personal space, by positioning themselves very close to the individual they are listening to, which would place the speaker to be very uncomfortable with the close proximity. To avoid this vocal reminders need to be given and in spite of this if there are no changes then, a strategy to solve this would be to tie a rope around the child's waist, with leaving a 3 foot tail behind. Practice this with peers holding the tail and demonstrating to the client the meaning of 3 feet difference.

**Behaviour Strategies: Difficulties with Transitions and Changes:**

Autistic children yearn for sameness and routine. If there is a change in the ir routine, then they would try their best to stick to their routine or would throw temper tantrums. It's seen that children with autism work better and learn better with people who are more structured in their daily routine.

The following are few listed strategies to help autistic children to accept and adjust to transitions:

1. Teach children to adjust and accept transitions by making small changes in their routine as and when they get very comfortable with it.
2. Dim the lights of the room, when a new activity is being presented to them.
3. Give prior explanation of the transition as getting them prepared before hand and understanding why a particular change is taking place would help the be prepared rather than having a flaring temper after the change has occurred.
4. Make the transition as enjoyable and ass tress free as possible because otherwise the child's autistic feature will shoot up.

**Behaviour Strategies: Problems with Self-Esteem and/or Depression:**

Often children with autism have a poor self-concept and lack confidence. This is normally seen to be present by the child having a poor eye contact, difficulty socializing or stammering. This is a very common feature in autism as these children undergo a lot of rejection from peers, they hear people around speaking about their behaviour and problems and being bullied.

**The following are few listed strategies to help boost self -esteem in autistic children:**

1. Keep a track of the child's behavioural achievements and encourage it by rewarding the child in front of the family members.
2. If the child is demonstrating poor or undesirable behaviours then do not take away his previously earned rewards.
3. Reward him / her for minor changes in the behaviour.
4. Be specific about the behaviours that are liked and encourage them with a reward. Behaviours that are undesirable need to be changed by desirable ones.
5. Phrases and words like "good boy" or "bad boy" should not be used rather "You followed the direction well" should be used to help increase the confidence.
6. The child's strengths should be motivated and ways to enhance it should be used. For example: A child who enjoys music should be encouraged to learn an instrument and excelling at playing it.
7. Encourage the client to maintain a photo book of his achievement or good behaviours. For example: If he / she won some competition or made some clay model, take a picture and keep it in front of the child.

8. When an autistic child is using negative words, phrases or sentences referring to himself encourage the child to make changes in his / her dialogues to a more positive view about himself / herself.
9. The child's grades, health concerns and short comings should be kept confidential and not publicized about.

**Behaviour Strategies: Managing Stress and Anxiety:**

Helping children with autism managing stress and anxiety is extremely important in preventing undesirable behaviours.

1. Maintain a time-table for all the daily activities and keep a reward system for it.
2. Teach the child breathing exercises, help him understand when to use it and practice it, so that the child is aware that he has to use it when he is stressed and anxious.
3. Exercise and play session is extremely important for the child, as it would help the client relieve his / her stress.
4. Self-talk method should be taught to the client and he should be also taught that this should be only practiced when he is alone in the room.
5. The client should have some hobby which he or she enjoys and feels relaxed w

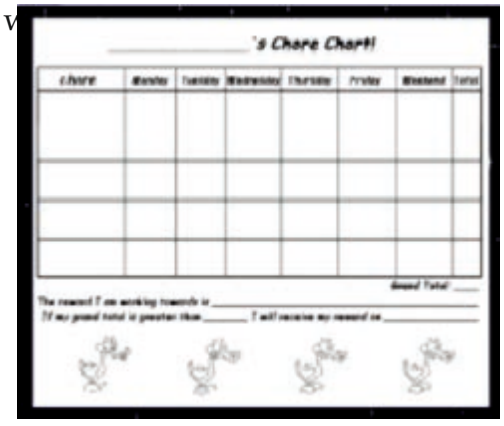



*Fig 29: Cores Chart for managing stress*

**Behaviour Strategies: Attention Problems:**

Attention problem scan lead to poor behavior. It is often assumed that a child is non-compliant when he or she does not pay attention to directions or is non-focused when a task is being performed.

1. The desk on which the child sits to study should be dark in colour and should have very few items on the desk. It is seen that dark colour desks are said to be able to maintain attention for a longer time.
2. A separate room should be allotted to the child, to sit and study. The room should have less furniture and the room should not be of a very bright colour. While the child is studying the windows should be closed as most of the autistic children love to stare outside the window.
3. Demonstrate to the client how one's boy should look when one is paying attention i.e. modeling technique should be used.
4. The child's interests should be known in term of activities with high interest and low interests. The low interest activities should be separated with breaks.
5. Arrange for tests and difficult assignments to be completed in quiet rooms away from distractions.

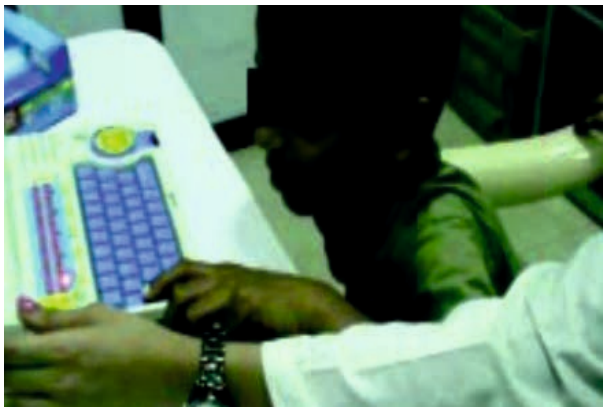


*Fig 30: Cognitive Rehabilitation to increase attention span and level of comprehension.*

**Strategies: Improve Academic Skills**

Autistic children have academic problems especially if they fall into the category of mental retardation or if they have learning difficulties.

1. Modify the homework and class work assignments for the child.
2. Use different modalities to teach in a way which will help the child grasp and understand better like audio and visual modality i.e playing nursery rhymes with video.
3. Create motivators and rewards that are clearly established and known to the child.
4. Have individualized and personalized session with the client to make the child understand concepts in a better way.
5. If the child is comfortable with a certain special educator or a teacher see to it that he / she is there with the client throughout the academic year. Changing teachers or educators would confuse and upset the child as autistic children if they get attached to a particular educator they can be very high chances of improvement in them.

**Rewards and Motivators:**

A very crucial aspect of any behavior modification/management plan is to have reinforcements listed down which would encourage a child to have a desirable behaviour and eliminate or reduce undesirable behavior. Punishment teaches children with autism that they have done something wrong and it does not teach them the right behaviour but instead has a bad impact on their self-esteem and confidence.

**Rewards can include:**

Edible reinforcers: fruits, biscuits, chocolates, etc.

Material reinforcers: toys, stickers, star on the hand, etc.

Social reinforcers: smiling, patting, praising, etc. Activity reinforcers: play time, or T.V. time, etc.



*Fig 31: Reward system*

**Reward Systems:**

Token Economy: When a child completes a task or activity assigned to him successful he is awarded with a token for example a poker chip, which he can exchange for various things like toys or T.V time.

Points earned versus points lost: This is where the child earns points on daily basis i.e. if a child's behaviour is good then he receives a point but if his behaviour is unacceptable then a point is subtracted. The list of acceptable and unacceptable behaviours should be specified to the child.

Sticker Card: The child here is presented with a sticker card consisting of 20 grid rectangle. If the child demonstrates good behaviour then he receives a sticker and after the completion of card he can trade it for a reward.

Positive notes: A positive word communicated to an autistic child is very encouraging but autistic children are visual learners and if the same words are put on papers then it would help boost the child's

confidence.

Above mentioned are a few reward systems for autistic children, it is very important to understand that a meaningful reward would encourage a child to have a good behavior and maintain it. Initially the child may be very excited about the reward but eventually may lose out interest in the reward, at this time the rewards should be revised. It is important that the child understands the reward system.

**Consequences:**

Like rewards are an important factor in contributing to the child's good behaviour, in the same way consequence is equally important for the child to understand that if not behaved in an undesirable manner then he will be punished.

**Types of Consequences:**

- Vocal chastise (no hitting).
- Loss of pleasure activities (loss of T.V time).
- Verbal threats or warnings (If you do that again, you will be made to sit in the timeout zone).
- Time-out (the child should be made to sit in the corner of the room, without making any noise for a decided time limit and should think about his behaviour and why he was punished? If he argues then his time limit would increase).

**APPLIED BEHAVIOUR ANALYSIS:**

Applied Behaviour Analysis (ABA) is based on the theory that influencing a response associated with a particular behaviour may cause that behaviour to be shaped and controlled. The behavioural methods in ABA are used to measure behaviour, teach functional skills, and evaluate progress. There are various studies which suggest the success of ABA technique in treating deficits in the behaviours of children with Autism Spectrum Disorder (ASD) at any level.

**Approaches of Applied Behaviour Analysis:**

1. Discrete trial training (DTT)
2. Pivotal Response Training (PRT)
3. Picture Exchange Communication System (PECS)
4. Self-Management
5. Social skills training techniques

The goal of the session is to find ways to motivate the child with the help of a number of strategies and to ensure that the session is enjoyable and productive. The purpose of the ABA program is to improve the language skills, play, and socialization and to eliminate or reduce self-injurious or ritualistic behaviours that interfere with the learning process. It also focuses on to improve the eye contact and encourage on the desire to learn. Even if the child does not achieve a "best outcome" result of normal functioning levels in all areas, nearly all autistic children benefit from intensive ABA programs.

Specific targets of the interventions are chosen based on the child's individual issues and disorder. In an intensive behavioural intervention program, goals are set and progress is continuously monitored and evaluated. If there are any positive changes in the child's behaviour then the goals are eventually changed, and the focus is shifted on to improving other disruptive behaviours.

**Basic Principles of Behavioural and Educational Intervention Approaches:**

Behavioural therapies incorporate specific approaches to help the individual acquire or modify behaviours. The basic theory that ABA is based on is operant conditioning which involves presenting a stimulus (request) to a child, and then providing a

consequence (a "reinforcer" or a "punisher") based on the child's response.

Reinforcement: A reinforcer is anything, which when presented as a consequence of a response, increases the probability or frequency of that response being repeated. Examples of possible reinforcers for young children may include verbal praise, or offering the child a desired toy.

Punishment: A punisher is a consequence that decreases the probability or frequency of that response. Possible punishers for young children may include verbal disapproval or withholding a desired object or activity. The term "punisher" is a technical term used in behavioural therapy and does not imply the use of punishment (physical abuse such as hitting, slapping, spanking, or pinching).

It is necessary to keep in mind that, reinforcers and punishers are different for each child. Many different specific behavioural and educational techniques have been used as part of interventions for individual children with autism. These techniques are effective in a wide body of research based on a common set of behavioural and learning principles. Behavioural interventions involve the therapist controlling the activity and / or consequences to shape the child's responses.

**Discrete Trial Teaching (DTT):**

This is an ABA based therapy which was developed in the 1970's by psychologists Ivar Lovaas, and Robert Koegel, at the University of California at Los Angeles (UCLA). This principle makes use of the idea that when children with autism are rewarded for desired behaviors, they are likely to repeat that behaviour. For example: the therapist gives the child a motivating cue, such as a request to stand still or maintain eye contact, along with a correct response. The therapist then rewards the child by a praise, toys or food to reward the child for completing the task.

Discrete Trial Teaching consists of a series of distinct repeated lessons or trials taught one-to - one. Each trial consists of a prior, a "directive" or request for the individual to perform an action; a behaviour, or "response" from the person; and a consequence, a "reaction" from the therapist based upon the response of the person. Positive reinforcers are selected by evaluating the individual's preferences.

Many children initially respond to, recognizable or concrete reinforcers such as food items. These concrete rewards are faded as fast as possible and replaced with rewards such as praise and hugs. Early intensive behavioural intervention such as the Lovaas program is usually implemented when the person is young, before the age of six. Parent training is a necessary part of an effective ABA- based program. A maintenance schedule allows for periodic checking so the person does not regress in mastered skills. Discrete trial training is a technique that can be an important element of a comprehensive educational program for the individual with autism spectrum disorder. In some cases, a much less intensive, informal approach of discrete trial training may be provided by a knowledgeable professional to teach specific skills such as sitting and attending.

**Pivotal response therapy (PRT):**

This therapy is also referred to as pivotal response treatment or pivotal response training, is a behavioural intervention therapy for autism. The main principle of this therapy suggests that behaviour links primarily on two 'pivotal' behavioural skills, i.e. motivation and the ability to respond to multiple cues, and that development of these skills will result in overall behavioural improvements.

**The primary pivotal areas of pivotal response therapy involve:**

1. Motivation

2. Initiation of activities
3. Self - Management
4. Feeling to respond to multiple cues
5. Ability to respond to multiple cues

The PRT training is child - directed: where the child makes the choice in which direction to direct the therapy session. Emphasis is also placed upon the role of parents as primary intervention agents. This training focuses on increasing a child's motivation to participate in learning new skills.

**Pivotal response training involves specific strategies such as**

- Clear instructions and questions presented by the therapist
- Child choice of stimuli (based on choices offered by the therapist)
- Intervals of maintenance tasks (previously mastered tasks)
- Direct reinforcement (the chosen stimuli is the reinforce)
- Reinforcement of reason for purposeful attempts at correct respond
- Turn taking to allow modeling and appropriate pace of interaction

Pivotal response training has a naturalistic training method that is structured enough to help children learn simple through complex play skills, while still flexible enough to allow children to remain creative in their play. The child can be reinforced for single or multiple step play. The therapist has the opportunity to model more complex play and provide new play ideas on his / her turn.

**Reciprocal Imitation Training:**

Reciprocal imitation training (RIT) is a variation on the pivotal response training procedure for teaching play skills. This training was developed to teach spontaneous imitation skills to young children with autism in a play environment; however, this intervention technique has also been shown to increase pretend play actions. This procedure includes unexpected simulation in which the therapist imitates actions and vocalizations of the child.

**Self-Management Training:**

This is an additional approach for teaching children with autism to maximize in independence and generalization without increased reliance on a teacher or parent. Self-management typically involves some or all of the following components: self-evaluation of performance, self-monitoring, and self-delivery of reinforcement. Ideally, it includes teaching the child to monitor his / her own behaviour in the absence of an adult. This therapy uses a self-management treatment package to train school-age children with autism to engage in increased levels of appropriate play. Self- monitoring procedures have also been used to increase social initiations while reducing disruptive behaviour and to increase independent interactions with typical peers.

In a study children displayed very little independent appropriate play before training, and typically engaged in inappropriate or self- stimulatory behaviour when left on their own. With the introduction of the self-management training package, the children increased their appropriate play in both supervised and unsupervised settings, and across generalization settings and toys. Decreases in self-stimulatory and disruptive behaviours were maintained in the unsupervised environments.

**Video Modeling:**

This is like vivo modeling which uses predictable and repeated presentations of target behaviours; however, these behaviours are presented in video format, thus reducing variations in model performance. Studies suggest that video modeling improves various skills in

individuals with autism, including conversational speech like verbal responding, helping behaviours and purchasing skills. This medium has also been claimed to increase vocabulary, emotional understanding, attribute acquisition, and daily living skills.

**Video modeling interventions can be used in 2 forms:**

1. Self-as-model: In this individuals act as their own models and the video is edited so that only desired behaviours are shown.
2. Other-as-model methods: This employs taping other individuals, typically adults or siblings, performing target behaviours

Applications of video modeling as an intervention technique are now being extended to teaching and increasing play in children with autism.

**Therapeutic Use of Applied Behavioural Analysis:**

It is extremely important to have a trained ABA therapist and to give adequate training to the parents or caregivers. ABA is the best method to manage the undesirable and anomalous autistic behaviours such as: self-injurious, repetitive, ritualistic, aggressive and disruptive behaviours. This approach has been seen and studies to extinguish or reduce these behaviours and promote alternative pro-social behaviours simultaneously. The process of ABA is successful as it breaks complex tasks into smaller parts making them less daunting for the child. ABA can also be used to train a child to learn a new adaptive behaviour, such as dressing and toileting and to promote functional communication.

**ABA whether they are observed or taught they function on 3 aspects:**

- Antecedent (A) - what triggered a behaviour or what happens before the behaviour,
- Behaviour (B) - the behaviour itself, and
- Consequence (C) - what happens after the behaviour.

The consequence is whatever the behaviour accomplishes, for example it can be getting attention (negative or positive) or relief of stress. The consequence is not always obvious, especially in the case of unusual behaviour (odd behaviours the child does such as arm flapping or repetitive actions), which is why keeping data is helpful to identify what the function of the behaviours are, as well as what triggers them.

**Applied Behavior Analysis assessment focuses on:**

- Exactly what behaviours are performed by the child
- When these behaviours are performed
- At what rate the behaviours are occurring
- What happens before and after the behaviours
- What purpose does the behaviour serves

Skills that are to be promoted are broken down into small sequential steps. The ABC principles of behaviour intervention are used to teach the child each step:

- A (antecedent): Each instruction is given clearly, in as few words as possible. Assistance is provided; for example prompting through demonstration or physically guiding.
- B (behaviour): An appropriate behaviour is observed.
- C (consequence): A consequence is an outcome that will reward the child and increase the likelihood that the behavior will be repeated again in the future, also called a positive reinforcer.

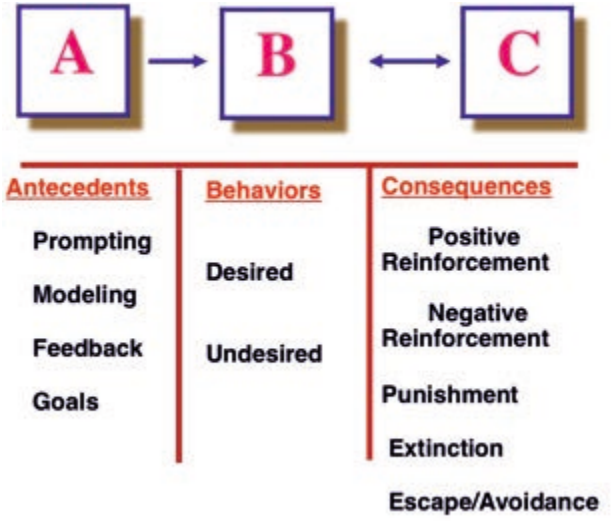



*Fig. 32: ABC Model*

This "ABC" process is repeated f for each behaviour both in structured teaching situations and in the course of everyday activities using PRT techniques.

I. Instructions are given to emphasize metacognition (learning how to learn). In this case learning how to listen, to watch, to imitate, to ask and do.

ii. As the child's learned behaviours improve, the structured guidance is systematically reduced and the prompts are used less frequently and eventually faded out.

**Teaching Procedure:**

This is so that the child learns to perform the trained behaviour independently.

iii. It is also important to change the context of teaching (different people giving the antecedent, more people around, different situations, etc.) in order to generalize the learned behaviours.

iv. When each behaviour consisting of single sequential steps are acquired, the person is taught to combine them to produce more complex behaviours.

v. Problematic behaviours are not reinforced; instead the child is consistently redirected to engage in appropriate behavior.

vi. The child's responses during each step are meticulously recorded. The information is later used to determine if the child is progressing at an acceptable rate. If progress is not satisfactory, the learning steps are analyzed for possible flaws and the program modified.

**Applied Behaviour Analysis: Example of the teaching procedure:**

Objective: The child will be able to make a chutney cheese sandwich.

Materials Required: Bread slices, butter, chutney, cheese, knife and plate.

Number of trials in set: Up to 5

| Step | Presentation | Response | Consequen |
|---|---|---|---|
| 1. | Mother making a sandwich | | |
| 2. | Tell the child to make the sandwich one at a time:<br>1. Get the plate and the knife and assist him.<br>2. Get the chutney, butter and cheese from the fridge and then assist him. | Child performs part of each step with assistance.<br>3 consecutive trial | Correct = Praise<br>Incorrect = Error interruption, re-administer with increased prompts |

| | | | |
|---|---|---|---|
| 2. | 3. Get the bread out of the bag and assist him.<br>4. Open the butter packet and take cheese slices and assist him.<br>5. Spread the butter and assist him<br>6. Spread the chutney and cheese slice and assist him. | | |
| | | | No response= System of most prompts<br>Re- administer with increased prompts |
| 3. | Tell the child to make the sandwich Model the steps " 2 - 5" | The child is able to perform step 1 without any prompt. | Same |
| 4. | Tell the child to make the sandwich Model the steps " 3 - 5" | The child is able to perform step 1 and 2 without any prompt. | Same |
| 5. | Tell the child to make the sandwich Model the steps " 4 - 5" | The child is able to perform step 1, 2 and 3 without any prompt. | Same |
| 6. | Tell the child to make the sandwich Model step " 5" | The child is able to perform step 1, 2, 3 and 4 without any prompt. | Same |
| 7. | Tell the child to make the sandwich | Child performs step 1 - 5 without any prompts. | Same |

**Social Stories:** Social stories are used to teach social skills through the use of accurate information about the situations that the child with autism may find difficult or confusing. Social stories can be used for different purposes like to prepare the child for an upcoming routine, or learn to communicate appropriately in social situations. The concept behind this form of therapy is that the child rehearses the story ahead of time with an adult, which would help the child to act in an appropriate socially acceptable way when the situation arrives.

Social Stories use different types of sentences for example:

1. Descriptive Sentence: This explains the, who, what, where and why about the situation, which helps the child recognize the situation as and when it occurs. For example: There will be many shoes to choose from.

2. Directive Sentences: This suggests the appropriate social response in the situation. For example: When I decide about the shoes, I will tell the grown-up.
3. Perspective Sentences: This describes the possible feelings or the response. For example: I might not know which shoes I like.
4. Affirmative Sentences: This describes the laws or rules that are commonly shared. For Example: That is okay with everyone.
5. Cooperative Sentences: Describe how other people will help out in the given situation. For example: The grown-up will go get the shoes for me.
6. Control Sentences: This helps the child remember strategies that work for him or her. For Example: I can hold onto my string while I decide.

**Theory behind Social Story:**

Autistic individuals have difficulty with reciprocal social interaction and this impairment might result from unusual activity or functioning of certain areas of the brain that are used for social skills. Some researchers believe that individuals with autism have trouble understanding what others believe, know, or don't know. This difficulty is sometimes called the theory of mind deficit in autism. Recent research studies show that social stories can help reduce problem behaviours, increase social awareness, and / or teach new skills. In some cases, the new behaviours were maintained and generalized to other situations, even after the story was faded out. Social stories are most useful for children who have basic language skills.

**Floortime (DIR):**

This therapeutic technique which is based on the Developmental Individual Difference Relationship Model (DIR) was formulated by Dr. Stanley Greenspan in 1980s. The premise of floortime is that an adult can help a child increase his circles of communication by meeting him at his developmental level and building on his strengths. Therapy is often incorporated into play activities on the floor.

**There are six developmental milestones that contribute to emotional and intellectual growth:**

1. Emotional Ideas
2. Emotional Thinking
3. Complex communication
4. Two-way communication
5. Self -regulation and interest
6. Intimacy or a special love for the world of human relations

The therapist or parent engages the child at a level where the child enjoys the activities and the therapist then follows the child's lead. The parent is instructed how to move the child toward more increasingly complex interactions, a process known as opening and closing circles of communication. Floortime does not separate and focus on speech, motor, or cognitive skills but rather addresses these areas through a synthesized emphasis on emotional development. The intervention is called Floortime because the parent gets down on the floor with the child to engage him at his level. Floortime is considered an alternative to and is sometimes delivered in combination with behavioural therapies. Floortime is usually delivered in a low stimulus environment, ranging from two to five hours a day by a psychologist or a special educator.

**Speech Management:**

Speech-Language symptoms of individuals with Autism are diverse. They may vary from being completely verbal to completely non-verbal. This partially depends on the intellectual power of the individual and

partially upon the depth of autism traits they carry. Many of us feel that there is a need to build a language system for these individuals, but this is not called for. These individuals already have a language system. We need to simply work on this existing system to make it more accessible.

Before going to the Speech- Language symptoms and treatment options in individuals with Autism, we must understand that we do not need to teach these individuals how to communicate. The manner in which they are already communicating needs to be understood. Every individual with autism has a unique form of communication. The uniqueness is added to their communication behaviour because of their sensory system which does not allow them to learn language the way normal individuals do. Thus, their manner of communication is deviant and may range from simply crying when he/she needs something to gesturing to vocalizing to verbalizing his/her needs. Deviant form of communication is a trait of their personality.

To understand this better we can compare an individual with Autism to an introvert individual who does not prefer to socialise. We usually do not try to modify an introvert's socializing behaviour, as their behaviour is accepted to be within the range of "normal deviation". On the other hand, the moment an individual is given the diagnosis of "Autism", we begin to teach them and to rebuild their way of communication. But actually all that requires to be done is to give a universal route their unique form of communication so that everybody can understand them. Their unique language which is understood only by a few, needs to be modified to a more universal form, so that everybody can understand them. This may be through words, it may be through a picture exchange communication (PECS), it may be through a speaking application or any other method of communication.

Besides the speech and language deviations, individuals with Autism also show picky eating habits. Some individuals just refuse take in any food orally and have to be put on a feeding tube to sustain nutrition. These behaviours too can be overcome by understanding the underlying sensory problem which is the cause of picky eating behaviours.

Once we understand the sensory system of an individual with Autism, it becomes easier to break the iceberg. But we must always remember, there exists immense variation between two individuals with Autism who may be showing the same symptoms. Like it is rightly said by Dr. Stephan Shore:

"If you have met one child with Autism….you have met one child with Autism."

**SYMPTOMATOLOGY:**

**Speech-Language Symptoms:**

Individuals with Autism experience a number of sensory issues which makes learning of language difficult. Thus, they often rely on crying, pointing, and vocalising to express needs and wants. Their sensory issues and body defensiveness may make it difficult for them to make eye contact and play with peer mates; thus showing poor eye contact, poor social smile, solitary play behaviour and irrelevant speech patterns. Consequently, the overall speech-language profile presents with delayed speech and langue development, severe social and pragmatic issues and behavioural problems may range from severe comprehension issues to inability to express needs and wants.

Some individuals with Autism develop good language skills, but show problems learning the phonology of a language and thus may show misarticulated speech. These individuals are usually also show pragmatic

deficits, i.e. they do not master to use the different modes of language in different situations and around different people.

**Feeding Symptoms:**

Individuals with Autism also often show feeding difficulties where the "anticipatory phase" or/and "oral phase" of swallowing is affected with an intact pharyngeal phase. These problems too are contributed by the altered sensory system of these individuals. They may show symptoms like prolong feeding hours, fear of trying new food, pocketing of food in cheeks for hours, fear of having hard or soft food, etc. These behaviours have be presentin any severity in a given individual, which may range from having mild eating problems to have severe fear of food such that a nasogastric tube has to be in-situ to sustain their nutrition.

**TREATMENT OPTIONS:**

About three decade from now, the world was in a state of learning about and coping with Autism. Everyone was into forming newer devices to overcome the various hurdles for individuals with Autism. Today, because of the consistent efforts made by many dedicated therapists towards Autism, we have many therapeutic treatment options available for individuals with Autism. One must remember that these treatment options are for modifying the communication patterns of the individuals with Autism to a more universal form and not to overcome/cure Autism. It is important to understand that Autism is not a disease that needs to be cured; it is a trait that an individual is born with, which needs to be accepted.

**Sensory Needs:**

At the very beginning is a must to make the individual comfortable by satisfying his/her sensory needs. Only when the individual's sensory needs are satisfied will he/she pay attention and be able to learn new language.

Some of the common sensory needs of individuals with autism are:

**1) Need to chew onto something:**

This is often manifested as biting behaviour, chewing onto new toys, chewing on T-shirt or any other piece of cloth. This sensory need can be fulfilled by providing him/her a chewy tube.

These chewy tubes are

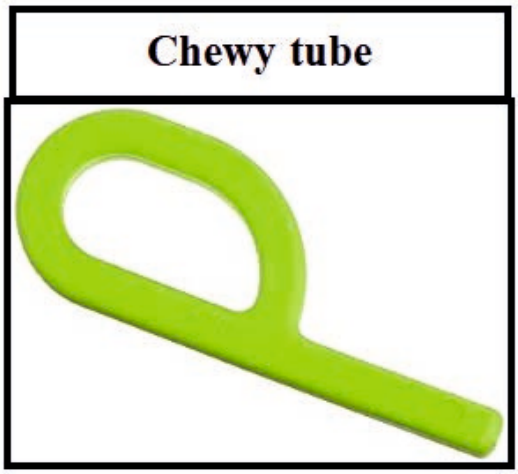



*Fig 33 : Chewy Tube*

Available in different textures &toughness. The speech therapist can help to choose the most suitable chewy tube for a particular individual.

**2) Need for tactile sensory inputs:**

This is often manifested as self-hitting behaviour, banging head, hand against the wall, jumping from heights onto knees, pressing very hard on the head and so on. These are behaviours for seeking increased pressure and tactile sensory inputs, which make it difficult for the individual to sit in one place; this in turn affects their sitting compliance. This need can fulfilled by using a special cushion which provides the sensory inputs which helps them sit in one place.

**3) Aversion of food:**

Many individuals with Autism are picky eaters, but this behaviour gets missed by

parents due to other more evident behaviours. They show some typical food behaviours like wanting to eat the same kind of food, fear of trying new food, fear ofeating certain consistencies of food, taking a prolonged time to have meals, pocketing of food in cheeks for long hours and so on. These behaviours are associated to the oral sensory defensiveness; i.e. their oral cavity may be hyper or hypo sensitive; which makes them a picky eater. A Z-vibe brush can be used to desensitise the oral cavity and gradually starting new food types. This brush is a vibrating brush with bumpy spikes on it which give the oral cavity the required stimulation. This also helps the movement of tongue and other parts of the oral cavity which helps in eating.




*Fig 34 : Cushion with bumpy spikes*




*Fig 35. Z-vibe brush*

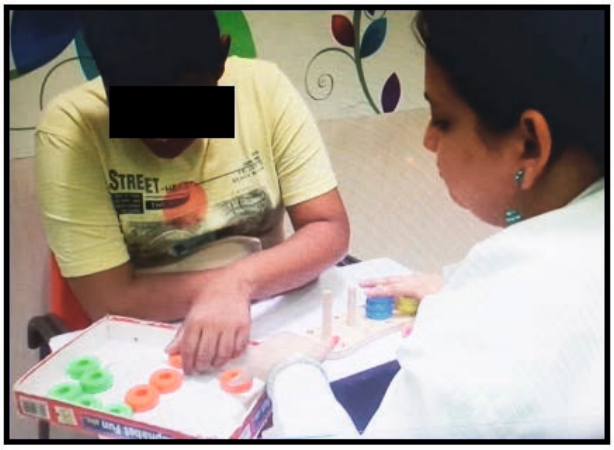


*Fig 36 : wood coins for colourmatching*

This may be manifested as covering ears every time there is a slight noise, or if anyone is taking to the individual with Autism. This can further manifest into irritation and anger. This can be fulfilled by playing pleasant music in the background, which can be of waves, or sea or water. This music can also be played in the background during therapy which helps to reduce melt downs and increase calm behaviour.

Applications for such soothing music are easily available on the PlayStore or Apple Store and can be downloaded for everyday use.

**Therapeutic Treatment:**

All the above, mentioned techniques are required toclam the individual with autism and acquire maximum attention to learn new language skills. Once the individual is settled, speech and language therapeutic methods need to be started as soon aspossible to universalise their language. Language stimulation done in play manner yields maximum results, thus, all therapeutic techniques are advised to be done in a play manner.




*Fig 37 : Cards Matching*

**1) To strengthen pre-requisites of language:**

If the child is completely non-verbal, the pre-requisites of language need to be developed in order to develop language. This requires working on matching, imitation, following directions and functional play.

Matching can be done using game such as puzzles matching, colour matching

card matching etc. Many more games can be using to improve matching skills which can be suggested by your Speech-Language Pathologist.

Simple imitation is the core aspect of acquiring proto-words. Imitation tasks start with whole body imitation, like hands up, touch your nose, etc., go on to sound imitation, like /baba/, /papa/ and then to meaningful words. Once imitation skills are developed, the child is ready for expressing his/her first words.

An important aspect of receptive language is following directions, i.e. a child needs to understand simple commands well. The child needs to be able to identify action, item, and agent, in order to follow a simple one stage command correctly. Once this is built, listening skills of the child are worked on and the command is increased to two orthree stages.

Last, but the most important is focussing on the play skills of children with Autism. Because these children are self-involved, they fail to play with toys the way normal children play. They tend to focus on the smaller bits the toys instead of understanding how to play the toy as a whole; e.x. the child may play with the tyres of a car instead of driving the car. Functional play skills need to be developed in these children, as play is a very important aspect of language.

**2) To strengthen language skills:**

This can be done through verbal language or Picture Exchange Communication System (PECS), or any other language system. Whichever language system is used for the child, the goal in this step is to increase the length of sentences during communication. Sentences can be verbal sentences, the number of pictures forming a sentence in PECS, or any other language system.

This can be done by increasing the vocabulary of the child; i.e. by introducing items from various categories. Remember, one should increase the number of categories instead of taking too many items in a single category. Ex. Different categories like: Clothing, Things around the house, Animals at the farm, Eating Utensils, Things that keep ourself tidy, should be taken instead of introducing a lot of items only in Animals category.

Speech-language therapy for Autism involves much more than the above mentioned techniques. The above, are only primary treatment options for universalizing their language. Every individual with Autism is different and thus the therapy techniques used for all individuals are different. One must consult the individual's Speech- Language Pathologist in order to get more therapy techniques.

## PHYSIOTHERAPY IN AUTISM CHILDREN

Children with Autism will experience difficulties with their motor skills and capabilities in addition to behavioural, sensory and emotional difficulties. The role of a physiotherapist is to help children who have difficulty in functional movements, poor balance and challenges moving through environment successfully. Some children with ASD may have low muscle tone, poor balance and co-ordination. The Physiotherapist after detailed assessment helps them in such situations by implementing a treatment plan to improve the child's overall functioning in daily living.

As children grow older, physical therapists should work more on functional skills such as skipping, kicking, Ball throwing and catching. These skills are important for physical development, social engagement in sports, recess and general play.

Physical therapists start by evaluating the developmental level and abilities of the child. After examination, they design activities that target those areas.

PT might include assisted movement, various forms of exercise, and orthopedic equipment. Additionally, therapists will work with families and caregivers to provide intervention according to the individual's needs. Caregivers can incorporate practice of skills developed in therapy into daily activities at home and in the community. The success of therapy depends on how much the child practices these new skills between therapy sessions.

Using the Bruininks- oseretsky test of motor proficiency, it was found that children with ASD have problems with running speed and agility, motor planning, bilateral coordination, visual motor coordination, limb strength, static and dynamic balance. One in every five children suffering from ASD is found to have generalized hyper-mobility of the joints and low muscle tone. These motor impairments can lead to great difficulty in adjusting to their physical environment, fine motor skills like writing, tying shoe lace or buttoning. Motor deficits in these children may only be detected late in their lives or go undetected.

Physiotherapy uses goal orientated exercises, fun games, to help to increase muscle fatiguability meaning that a child is able to sit at his desk at school more easily for a greater amount of time which in turn leads to less disruption to completing school work.1

**Factors contributing to difficulties with motor activity in ASD :**

- A delay in the development of their movement skills
- Poor coordination – they might seem clumsy, find it hard to coordinate both sides of their body to do things like catch a ball, or find it hard to do things where they have to use their hands to grasp onto something.
- Deficient postural control, which means they find it hard to hold themselves upright, or might lose their balance easily
- Impaired imitation skills – imitation is when a child copies the people around them and for young children it is an important part of how they learn, for example, copying their parent clap their hands.
- Low muscle tone –Children with ASD often have low muscle tone, which means that they require to use more strength and energy to move, and also alters the sensory feedback a child receives through their muscles when they move. Children are clumsy, they tend to avoid physical activity
- Motor planning difficulties – this affects the child's ability to optimally time,

sequence and execute a movement. This is often seen as clumsiness, frequent falls, poor balance, or poor coordination.

- Poor core strength – as a result of their low muscle tone and motor planning difficulties, children with ASD often also have poor core strength. Children with ASD have difficulty recruiting the correct muscles in their core, and lack the strength and endurance to enable them to engage these muscles for functional activities.
- Poor posture – develop because of poor strength and low muscle tone, which impacts on a child's ability to master more sophisticated motor skills, fine motor skills, and can also impact their attention and participation in the classroom or at school. They prefer to sit W- sitting.



*Fig 38 Poor posture*



*Fig. 39 W sitting*



*Fig 40 Poor fine motor skills*

- Gait :Children with Autism tend to walk with flat feet, a reduced stride length, increased step width and therefore wider BOS, and increased time in the stance phase. Also, they have reduced ROM at the ankle and knee during gait, children with ASD may have weakness around these joints. Children with ASD have altered gait patterns to healthy controls, widened base of support, and reduced range of motion. Most of the children with ASD walk on the balls of their feet (Toe Walking).
- Delayed achievement of poor or inefficient balance reactions – these include the child's automatic reactions to keep themselves upright when they start to lose their balance, and the automatic protective reactions to protect themselves from injuries when they fall.

**Intervention/Plan for Autism**

For kids with autism, research paper show that activity for more than 20 minutes can help decrease stereotypical behaviors, hyperactivity, and aggression. Exercise not only helps children with autism better engage in the environment, but it also helps promote weight loss and leads to better overall health.

**Strategies that may be used as part of a physiotherapy intervention plan include:**

- Specific exercises or activities for muscle strength, posture, endurance, motor planning and balance reactions
- Sensory Integration strategies to improve a child's awareness of their body posture and movements
- Recommendations for orthosis, and/or other bracing, taping or positioning strategies and equipment to enhance the child's body alignment, posture and body awareness during functional or physical activities
- Recommendations and support to participate in community based physical activity programs that will support their physical skills and endurance.





*Fig 41 A and B*





*Fig 41 C and D*

*Fig41 A to D.:Exercises to improve motor planning*

**Exercises for Autistic Children**

1. Exercises to improve muscle tone
   - Strengthening exercise using theraband & weight cuff
   - Weight bearing exercises
2. Strengthening abdominal muscles
   - Leg raise
   - Curl ups
   - Core muscle strengthening exercises (Physio ball exercises).
3. Exercises to improve balance and co-ordination
   - Ball throwing
   - Balance board
   - Ladder exercise

- Visual Exercises

4. Exercises to reduce obesity and to improve cardiovascular fitness

- Cycling
- Treadmill
- Floor exercises

5. Exercises to reduce toe walking

- Stretching
- Walking on different Textures
- Modified shoes



*Fig 42: Walking on different textures*



*Fig. 43:Weight bearing exercises*



*Fig 44: Core strengthening exercises*



*Fig 45: Wheel barrow walking*



*Fig. 46: Crawling through tunnel*



*Fig 47: Back extensor strengthening*



*Fig 48 A and B: Lower limb strengthening exercises using weight cuffs*



*Fig. 50: Scootor board - crawling*

**Benefits of physical Therapy in autism are listed below:**

- Maximize sensory processing and organization skills to put into controlled motor planning.
- It will help to improve motor skills, essential for weight management, improve social skills, improve mental health and decrease stereotypical behaviour.6
- Improve self-regulation
- Decrease aggression
- Increase academic responses
- Appropriate motor behaviour
- Reduce Clumsiness
- Improve postural control to increase stability during fine motor, gross motor, and self-care activities. Also able to perform the ideation, sequencing, timing and execution components of motor planning.Improve static/dynamic balance to improve motor control and attention and decrease impulsivity.

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# Chp. 15 Attention Deficit Hyperactivity Disorder

**Mr. Vishal Ganar, Dr. Nishigandha Doke**

## Introduction

Most children experience difficulties in paying attention and concentrating at some point or the other. However, a diagnosis of Attention deficit Hyperactivity disorder is only warranted if these symptoms of difficulties in attention or impulsiveness affects the life of an individual affecting his/ her functioning and development. ADHD is characterised by developmentally inappropriate levels of (i) inattention, (ii) Hyperactivity, and (iii) impulsivity. Although the clinical manifestations in children vary, symptoms of ADHD are chronic (present for at least 6 months), cross situational in presentation and developmental in origin. It is noteworthy to emphasis that the said symptoms are pervasive, they occur in more than one setting (eg home and School). These symptoms also have to be long standing; symptoms have to be present from at least before the age of 12 years.

Attention deficit/Hyperactivity Disorder is a very common mental disorder among the children with over 10 million reported cases each year in India. The core symptoms of ADHD are age-inappropriate inattention, impulsivity and hyperactivity. These core issues manifest in different ways in different aspects of an individual's life affecting the social, occupational or academic functioning and development. American Psychiatric Association's Diagnostic and Statistical Manual-5 classifies ADHD as Predominantly inattentive presentation, predominantly impulsivity/ hyperactivity presentation and combined presentation.

## Definition

Attention-Deficit/ Hyperactivity Disorder is a neurobehavioral disorder characterized by a combination of inattentiveness, distractibility, hyperactivity, and impulsive behaviour. Symptoms include difficulties with attention to tasks, being forgetful in daily activities, difficulties in organizing, and following through. Hyperactivity symptoms can include being fidgety, restless, talking or interrupting others excessively and being impulsive. (Psychology Today)

## History

ADHD has long been studied and attempts have been made to manage the symptoms. Way back in 1798, Sir Alexander Crichton, in his book 'An inquiry into the nature and origin of mental derangement' described 'mental restlessness.' It was not until 1902 that ADHD was clearly described by George Still.

The evolution of our understanding of ADHD is clearly reflected in the changes seen in DSM criteria for ADHD. DSM I published in 1952 termed ADHD as 'minimal brain dysfunction.' DSM-II (1968) used the term hyperkinetic reaction of childhood. DSM-III (1980) called it Attention-deficit disorder (ADD) with or without hyperactivity. DSM-III-R, the revised version of DSM-III, published in 1984 changed it to ADHD while DSM 4 in 1994 split the diagnosis into 3 subtypes which were retained in DSM-5 in 2013

With passage of time, the way we treat ADHD also changed. Use of stimulants to treat ADHD was first described in 1937. In 1934, Benzedrine became the first amphetamine medication approved for use in USA. Methylphenidate was introduced in the 1950s and enantiopuredextroamphetamine in 1970s. Similarly, our understanding of ADHD can be reflected in the goals of therapies. For instance, rather than controlling core symptoms, therapies are aimed at improving functionality and managing the behavioural issues associated with inattention, impulsivity and hyperactivity.

## Epidemiology

***Prevalence:***

ADHD is estimated to affect about 6-7% of people aged 18 and under when DSM-IV criteria are followed. As per DSM 5 the general prevalence rate of ADHD is 5% in children and 2.5% in adults. When diagnosed via ICD 10 criteria, reported prevalence among individuals 18 years or younger is 1-2%. Prevalence rate across different countries differ, with higher rates in North America than in Africa or Middle East. This difference is generally believed to be a result of differing methods of diagnosis rather than some underlying factors. According to Froehlich et al (2007), prevalence of ADHD is 8.7%. The increasing rates of ADHD can be attributed to the changes in how the condition is diagnosed and how willing are people to seek treatment. Some argue that ADHD is over-diagnosed while some debate that it is the contrary. More research in this area is required to conclusively say which opinion is more accurately.

***Sex differences:***

ADHD is more frequently diagnosed in males as compared to females, as much as three times. This difference is cited to be either due to susceptibility or due to difference in diagnosis and gender norms.

***Age:***

The prevalence rate in children is twice as much as adults. Similar to how hyperactivity and inattention reduces with age in a typical development, hyperactivity for an individual will not be manifested as difficulties in remaining seated but as being restless when sitting in one place, underestimating the prevalence of ADHD among adults. These differences in manifestation could leave out many individuals from being diagnosed and more so from seeking help. Some studies also suggest that symptoms of ADHD remits with age.

## Theories of ADHD:

***1. ADHD as a disorder of Self Regulation and Executive Functioning:***

Clinical researchers like Virginia Douglas, are of the opinion that ADHD involves serious deficits in self regulation and many researches have even pointed at it being self-regulation deficit disorder. Self Regulation theory is a system of conscious personal management that involves the process of guiding one's own thoughts, behaviors, and feelings to reach goals (Roy Baumeister). Self regulation and Executive Functioning deficits leads to poor control on mental activities like self awareness, inhibition, executive attention or attentional management, working memory, resistance to distraction, emotional self control, problem solving and self-motivation. These deficits lead to lack of actions that people otherwise use as a guide to direct themselves to alter their behavior which would lead to further improvisation of a futuristic goal or attainment of a goal. Someone with

ADHD would appear to have age inappropriate abilities to engage in self regulation, for the attainment of specific goals and future actions. Interventions or treatment for ADHD should thus be directed at helping them to overpass these delays or atleast at finding alternate strategies to help them engage in these mental activities.

Research has led to the formation of the Executive Dysfunction Theory. According to this theory, ADHD showcases deficits in higher order functioning like planning, sequencing, reasoning, holding attention to a task, working memory, inhibition of inappropriate and selection of appropriate behaviors. These functions of higher order in turn facilitate lower order processes like language, learning, perception, explicit memory and action. Due to the deregulation of these higher order processes, the secondary processes are also affected. Neuropsychological tests that are used to tap on the workings of executive functioning system, have been administered on children with ADHD. The results obtained post the administration portray physiological, anatomical, and biochemical dysfunctions in the frontal cortex, the fronto-parietal and the fronto-striatal circuits. This hypothesis best explains the impulsivity and inattention symptoms involved in ADHD, but fails to account for the overall complexity or hyperactivity symptom of the disorder. A neurological marker called response inhibition has been conceived as a major deficit area that leads to impairments in other executive functions. Response inhibition has been investigated using neuropsychological tasks such as the go/no-go and stop signal in children and adults with ADHD and different forms of attentional dysfunction in ADHD have also been investigated, by using varied forms of the continuous performance task (CPT) to assess sustained attention deficits in children and adults with ADHD. The results on this task have pointed out to deficits in the executive functioning circuitry.

Deficits have been pointed out mainly in working memory, internalization of speech, self-regulation of affect, motivation. arousal and novel goal-directed action, resulting in decreased motor control of internally represented information and action.

### *2. State Regulation Model*

This theory aims at explaining the observed deficits in performance which is attributed to the disequilibrium in the state of energy. Performing a particular task is seen as a structural computational information process. The availability of these processes is dependent on the arousal and activation levels of the subject. Arousal, here is defined as a time-locked phasic physiological response to input, whereas activation refers to a long-lasting voluntary readiness for action. According to the Cognitive Energetic Model of Sanders, it is important to through elementary stages and have an optimal energy distribution to perform a task consistently. The stages are as follows: stimulus encoding, memory search, binary decision, and motor preparation. The efficiency of tasks not only depends on these stages but also on the effort that is channelized to meet task demands and to compensate for a sub-optimal state of arousal and/or activation by either activating or inhibiting the arousal and activation levels. This effort system functions on the basis of an evaluation mechanism, which regulates the momentary state of the arousal and activation levels. The state regulation theory states that children with ADHD have difficulty in keeping an optimal activation state, possibly due to inef-

ficient extra effort allocation.

## Etiology

Nature vs. Nurture has been a subject long debated when it comes to study of Human behaviour. The idea that genetic information and biological systems of an individual affects the individual's behaviour would support nature as the predominant determinant of a behaviour. Conversely, the idea that a behaviour is influenced by the prenatal, post-natal and societal factors in an individual's environment would be imply that 'nurture' is the predominant determinant of a behaviour. Various studies have been done to study determine which influence behaviour more than the other. The conclusion of these studies has been that both Nature and Nurture influence not only behaviours but also each other in a dynamic manner.

### *Genetic Factors*

A number a genes have been identified to play some role in ADHD. Meta analysis of genetic studies (based on twins and adoption studies) have identified several genes including Dopamine transporter gene (DAT1), Dopamine D4 Receptor gene (DRD4), Dopamine D5 Receptor gene (DRD5), Serotonin transporter gene (5-HTT), Serotonin 1B Receptor gene (HTR1B), and Synaptosomal-Protein associated 25 gene (SNAP25).

### *Biological Factors*

Research using MRI compared male adolescents with matched controls found decreased activation of action-attentional system (Broadman's area 46, 39 and 40) along with superior parietal and middle frontal areas (Broadman's areas 7 and 10 respectively). Increased activation was seen in posterior midline attentional system. Research done by NIMH compared children with and without ADHD found that children with ADHD have 3-4 percent reduction in brain volumes in areas like frontal lobe, temporal gray matter, caudate nucleus, and cerebellum. These areas are important in problem solving, planning, understanding behaviour of others and inhibition or restraining impulses.

### *Environmental Factors:*

People often wonder if ADHD is a result of the fast-paced lifestyles; the technology that has infused in our children's lives. Way back in 1845, Heinrich Hoffman, a German Psychiatrist authored Der Struwwelpeter, a children's book which talks of a character called fidgety Philip who keeps rocking in the chair, who is unable to stay still and consecutively spills the food. He is also described to interrupt his parents and being loud at home. In a society widely different from ours, with no IPads and TVs, we find a character with typical ADHD symptoms.

A probable reason for increasing the number of diagnosis made per year is the fact that our contexts have made these symptoms pronounced. For instance, children are required to sit in a classroom for 6 hours with as many as 50 other kids with little individual attention.

There are no known causes for ADHD. It is also important to clear out that bad parenting or bad teachers do not cause ADHD. Similarly sugars, food dye or diet do not cause ADHD though some kids can be sensitive to certain elements of their diet. These kinds of food do not affect behaviours in ways to cause ADHD. Research has also shown support for the idea that

Computer, TV and video-games also do not cause ADHD. While we know that most of the substantiated causes appear to fall in the realm of neurobiology and genetics, environmental factors may affect the severity of the symptoms and the degree of impairment and suffering experienced.

There is a correlation between the use of cigarettes and alcohol use during pregnancy and risk for ADHD in the child. Exposure to high levels of lead is also considered as a risk for ADHD.

## Diagnosis and Clinical features:

A child with behaviour characteristic of ADHD are not referred for impulsivity or inattention but with complains of poor academic performance or disruptive behaviours in class. The goal of a diagnosis is not to give a label and send the child home but to plan and start intervention. It is necessary to align diagnosis and assessment with treatment and intervention.

The first and the most important aspect during diagnosis is to ascertain the areas of impairment in daily life functioning.

Guidelines for diagnosis are clearly given in DSM 5. Based on the cluster of symptoms, an individual can be said to have inattention presentation, hyperactivity/impulsive presentation or combined presentation (have symptoms from both groups). The level of severity can be marked from mild, moderate to severe. Impairment in functioning can be marked in different settings; home, class, sports, unstructured peer relationship activities etc.

Co-morbidities, like learning disorders, anxiety or mood disorders should also be evaluated as a good number of individuals with ADHD are reported to have these co-morbidities. A twin study with 2082 pairs found that conduct problems and ADHD share common genetic etiology.

### *DSM 5 Criterion:*

Attention-Deficit/Hyperactivity Disorder (ADHD) 314.0X (F90.X)

A. A persistent pattern of inattention and/or hyperactivity-impulsivity that interferes with functioning or development, as characterized by (1) and/or (2):

1. Inattention: Six (or more) of the following symptoms have persisted for at least 6 months to a degree that is inconsistent with developmental level and that negatively impacts directly on social and academic/occupational activities

Note: The symptoms are not solely a manifestation of oppositional behavior, defiance, hostility, or failure to understand tasks or instructions. For older adolescents and adults (age 17 and older), at least five symptoms are required.

a. Often fails to give close attention to details or makes careless mistakes in schoolwork, at work, or during other activities (e.g., overlooks or misses details, work is inaccurate).

b. Often has difficulty sustaining attention in tasks or play activities (e.g., has difficulty remaining focused during lectures, conversations, or lengthy reading).

c. Often does not seem to listen when spoken to directly (e.g., mind seems else-

where, even in the absence of any obvious distraction).

d. Often does not follow through on instructions and fails to finish schoolwork, chores, or duties in the workplace (e.g., starts tasks but quickly loses focus and is easily sidetracked).

e. Often has difficulty organizing tasks and activities (e.g., difficulty managing sequential tasks; difficulty keeping materials and belongings in order; messy, disorganized work; has poor time management; fails to meet deadlines).

f. Often avoids, dislikes, or is reluctant to engage in tasks that require sustained mental effort (e.g., schoolwork or homework; for older adolescents and adults, preparing reports, completing forms, reviewing lengthy papers).

g. Often loses things necessary for tasks or activities (e.g., school materials, pencils, books, tools, wallets, keys, paperwork, eyeglasses, mobile telephones).

h. Is often easily distracted by extraneous stimuli (for older adolescents and adults, may include unrelated thoughts).

i. Is often forgetful in daily activities (e.g., doing chores, running errands; for older adolescents and adults, returning calls, paying bills, keeping appointments).

2. Hyperactivity and impulsivity: Six (or more) of the following symptoms have persisted for at least 6 months to a degree that is inconsistent with developmental level and that negatively impacts directly on social and academic/occupational activities:

Note: The symptoms are not solely a manifestation of oppositional behavior, defiance, hostility, or a failure to understand tasks or instructions. For older adolescents and adults (age 17 and older), at least five symptoms are required.

a. Often fidgets with or taps hands or feet or squirms in seat.

b. Often leaves seat in situations when remaining seated is expected (e.g., leaves his or her place

in the classroom, in the office or other workplace, or in other situations that require remaining in place).

c. Often runs about or climbs in situations where it is inappropriate. (Note: In adolescents or adults, may be limited to feeling restless.)

d. Often unable to play or engage in leisure activities quietly.

e. Is often "on the go," acting as if "driven by a motor" (e.g., is unable to be or uncomfortable being still for extended time, as in restaurants, meetings; may be experienced by others as being restless or difficult to keep up with).

f. Often talks excessively.

g. Often blurts out an answer before a question has been completed (e.g., completes people's sentences; cannot wait for turn in conversation).

h. Often has difficulty waiting his or her

turn (e.g., while waiting in line).

i. Often interrupts or intrudes on others (e.g., butts into conversations, games, or activities; may start using other people's things without asking or receiving permission; for adolescents and adults, may intrude into or take over what others are doing).

B. Several inattentive or hyperactive-impulsive symptoms were present prior to age 12 years.

C. Several inattentive or hyperactive-impulsive symptoms are present in two or more settings (e.g., at home, school, or work; with friends or relatives; in other activities).

D. There is clear evidence that the symptoms interfere with, or reduce the quality of, social, academic, or occupational functioning.

E. The symptoms do not occur exclusively during the course of schizophrenia or another psychotic disorder and are not better explained by another mental disorder (e.g., mood disorder, anxiety disorder, dissociative disorder, personality disorder, substance intoxication or withdrawal).

Specify whether:

314.01 (F90.2) Combined presentation: If both Criterion A1 (inattention) and Criterion A2 (hyperactivity-impulsivity) are met for the past 6 months.

314.00 (F90.0) Predominantly inattentive presentation: If Criterion A1 (inattention) is met but Criterion A2 (hyperactivity-impulsivity) is not met for the past 6 months.

314.01 (F90.1) Predominantly hyperactive/impulsive presentation: If Criterion A2 (hyperactivity-impulsivity) is met but Criterion A1 (inattention) is not met over the past 6 months.

Specify if:

In partial remission: When full criteria were previously met, fewer than the full criteria have been met for the past 6 months, and the symptoms still result in impairment in social, academic, or occupational functioning.

Specify current severity:

Mild: Few, if any, symptoms in excess of those required to make the diagnosis are present, and symptoms result in only minor functional impairments.

Moderate: Symptoms or functional impairment between "mild" and "severe" are present.

Severe: Many symptoms in excess of those required to make the diagnosis, or several symptoms that are particularly severe, are present, or the symptoms result in marked impairment in social or occupational functioning.

## Evidence Based Assessments

Unfortunately, there are no medical tests of brain scanning techniques to diagnose ADHD. We also cannot rely on a child's description about their symptoms as most of the children do not have the understanding of how the said symptoms affect their functioning. Therefore, we have to rely on

parent and teacher's reports of the child's observed behaviours to evaluate the extent to which symptoms are present, how frequently they are present and the severity of the impairment in a child's functioning.

Some of the evidence based scales for ADHD are SNAP rating scale, ADHD rating scale and Vanderbilt rating scale. They have the same format; they list down DSM symptoms and ask the rater (parent/ teacher) to rate the symptoms on a likert scale (eg. not present, present sometimes, present most of the time, always present). There are other various and more comprehensive scales like Behavioural checklist, Behavioural assessment for children, and Conner's rating scale.

## Treatment

The symptoms themselves per se are typically not the things that trigger treatment efforts. parents and teachers don't refer children due to distractibility and difficulties in paying attention. These difficulties result in behaviours that cause impairment in daily life function; for instance, it makes it hard for a teacher to manage his class and/or cause frustration to parents because the child requires multiple reminders to do their everyday activities like getting ready for school and/or cause problems in peer relationships where the child doesn't get invited to birthday parties. These occur due to child's inattention, hyperactivity and impulsivity.

Presently, the treatments available for ADHD, are medications, psychotherapy, education, or training, or a combination of treatments. Pharmacological treatment of ADHD has been found to be positively associated with improved academic achievement in elementary school children, and improved health-related quality of life in children and adolescents, and improved brain dysfunction. The most commonly used psychotropic drugs are first line pharmacological agents approved by Food and Drug Administration Department. Stimulants like methylphenidate and amphetamines help in increasing levels of norepinephrine and dopamine. This release is especially facilitated in the prefrontal cortex. Methylphenidate binds to the dopamine transporter and blocks the reuptake of dopamine from the synaptic cleft, whereas amphetamines increase the availability of norepinephrine and dopamine at the synaptic cleft by displacing them from the presynaptic terminal storage sites and by blocking the action of a degradative enzyme, catechol-o-methyltransferase. (Anna Shier, Journal of Central Nervous System Disease. 2013). These medications can have side effects ranging from appetite suppression, stomach ache, insomnia, headache. tics, emotional lability, irritability, and increases in heart rate and blood pressure. In such cases non stimulants may be prescribed by the medical professional.

## Psychotherapy

Psychotherapy is yet another modality to treat ADHD. This can help patients and their families to cope better with everyday problems. A type of psychotherapy, that targets change in a person's behavior helps the child to organize his tasks, complete his schoolwork, resolve emotionally taxing events, monitor his/her behavior and help replace undesirable behaviors with desirable ones.

Social Skills Training: Children with ADHD often have difficulties in peer relationships. Interventions focusing on social

skills training are thus an important part of treatment for children with ADHD. They experience difficulties with social incompetence, and often show disruptive and rule-violating behaviours.

Social-skills training helps to improve and maintain the individual's social skills. The children are taught appropriate verbal and nonverbal behaviour in their social interactions (Storebø et. al, 2011). The focus is on teaching the children to understand cues in social interaction, such as turn taking, understanding when to change topics during a conversation, and being able to recognise the emotional expressions of others and the tone of voice. Learning to share toys, asking for help, and how to appropriately respond to teasing are also important aspects of social skills training. It also involves teaching children to understand social rules and expectations (Liberman, 1988). Social skills training can be done by using the techniques of role play, modelling, feedback, rewards and consequences, exercises and games, homework, and may include parents and sometimes teachers.

Social skills training is usually done in groups. It consists of an 8 to 12 week programme. The duration of each group session is between 50 to 90 minutes. The frequency of treatment sessions can vary from a couple of times a month to several times a week. Most social skills training program focus on improving problem solving, control of emotions, and verbal and non-verbal communication (Storebø et. al, 2011).

## Behavior Modification for Attention deficit hyperactivitydisorder

Behavior therapy goal is to encourage positive behaviors and discourage unwanted or problem behaviors. Good treatment plans will include monitoring improvement closely and adjusting the therapy as needed along the way. Children with ADHD can have behavioral problems at home, and school. Behavior modification is an evidence based treatment for helping them through academic problems and in improving their family relationships.

Behavior modification involves the use of reinforcement and punishment to shape behavior. Behavior modification for children with ADHD children is based on the principles of social learning theory. According to this theory behavior modification helps to understand the continuous two way interaction between individuals, their behavior, and the environment, and the further goal attached to it is to identify the environmental and intrapersonal correlates of problematic behaviors. This observed correlation helps to manage and manipulate those correlated events and change the behavior. Behavior modification involves working with the ABCs of a behavior. It targets on changing the antecedents i.e. A (triggers), behaviors i.e. B (target behaviors that cause problems), and consequences i.e. C (responses). Caregivers are trained to record these problematic target behaviors, explain them in clear and simple terms, identify the context in which they occur, and use strategies to decrease their occurrence. These strategies include working on prevention of problematic events (modifying triggers), or working on reaction or consequences attached to the problematic situation. Another step is to inculcate in the child alternate behaviors that help in achieving his/her goal.

## Cognitive behavioral therapy

The symptoms of ADHD persist in adult-

hood and hence structured CBT programs can help the patient through issues, not of primary concerns but also of primary concerns but also the accompanying secondary issues like behavioral problems associated with use of drugs, self esteem issues, improving peer and familial relationships. Most ADHD, CBT based programs range from 8-12 sessions and can be conducted on individual or group basis. . CBT helps in understanding that how one's negative experiences may lead to disturbances in one's functioning and lead to increased emotional disturbances in adults with ADHD. People with ADHD experience failure and underachievement in varied areas of life due to the associated neurobiological deficits. These repeated negative experiences leads to frustration, poor self concept and low self esteem, negative beliefs about the self, which, in turn, leads on towards the expression of negative emotions such as depression and anxiety. CBT focuses on changing these negative experiences using cognitive and behavioral strategies

Cognitive behavioral therapy can also teach a person mindfulness techniques, or meditation. A person learns how to be aware and accepting of one's own thoughts and feelings to improve focus and concentration. The therapist also encourages the person with ADHD to adjust to the life changes that come with treatment, such as thinking before acting, or resisting the urge to take unnecessary risks.

Family therapy can also be helpful. Before the child is diagnosed, family members may have formed their own opinions about the cause of the child's behaviour. Family therapy can help reduce the tensions that arise at home and between family members while raising a child with ADHD. It can also provide a safe environment for family members and the children to discuss issues and problems in a calm and rational manner. Family and marital therapy can help family members and spouses find better ways to handle disruptive behaviors, to encourage behavior changes, and improve interactions with the patient. It also helps the family to come together and work as a team.

## Education and Training

In order for children and adults with ADHD to reach their full potential they require both, guidance and understanding from their parents, families, and teachers.. Because of the symptoms manifested in ADHD, frustration, blame, and anger is often present within the family environment before a child is even diagnosed. Parents and children might require additional help and counselling to overcome such negative feelings. Therapists need to educate parents about the symptoms of ADHD and how it affects the family as a whole. This will help the parents to not only have a better understanding of the difficulties experienced by their child, but will also help them to learn new skills and develop new and more positive attitudes. This in turn will help the parent and child relate better to each other.

Parent training programs program focuses on training parents and helps them to gather a detailed account of behavioral problems, trying to understand when and in what situations the undesirable behavior occurs. It also helps the associated adult to understand the consequences of his/her reaction to the undesirable behaviors. In short it tries to focus on the social contingencies that would be responsible

for cueing, aggravating, and maintaining the problematic behavior. Parents and family members are trained to give positive and negative feedback for certain behaviors. They are trained to establish a clear routine for the child, and to deliver clear instructions regarding the behaviors the child is expected to show. Parents are trained to work with the child by providing them required assistance considering the child's proximal zone of development. This technique is known as Scaffolding. Parents, teachers, and family members can provide positive or negative feedback based on behaviors shown. This feedback will help them to establish rules and other structured routines which will lead to better control of their behavior.

Parenting skills training helps to teach parents the skills they need to encourage their children to modify maladaptive behaviours and adopt more positive behaviours. It teaches parents the correct way of using the system of rewards and consequences to change problematic and disruptive behaviours displayed by the child. They are taught to be consistent in their behaviours, to reward and give positive feedback to behaviours they want to encourage and are taught ways of discouraging negative behaviours such as ignoring and redirecting negative behaviours. They are also taught how to structure situations in ways that support the desired behavior.

Parents also benefit from learning stress management techniques. This helps them to deal with their frustrations in adaptive ways so that they can respond to their child's behaviour in a calm manner.

Support groups for parents and families also help them to connect with others who have similar problems and concerns. Groups often meet regularly to share frustrations and successes, to exchange information about recommended specialists and strategies, and to talk with experts.

## Role of Occupational Therapy in Attention Deficit Hyperactive Disorder

Children with attention deficit hyperactive disorder may have difficulty performing tasks in environments that are moderately or highly stimulating, such as the typical classroom or Household. When trying to engage in activities, the child may have to work at continuously redirecting her attention from extraneous sounds and sights and back to the targeted task. Reading comprehension and listening skills are often affected. Impulsiveness may result in acting before thinking, leading to Errors on schoolwork or inappropriate social behavior. They may have difficulty following through on commitments due to disorganization and forgetfulness. They are less able to organize their time and materials than most of their peers. They may have difficulty sustaining effort on tasks, quickly become "bored" and leave tasks unfinished.

Occupational therapy for children with attention deficits should address the personal, environmental, and occupational factors. Children and teens can be taught to understand that they have a neurologically based problem. They can then learn ways to manage this through adherence to medication routines and attending and responding to their sensory and emotional states. Framing the child's difficulty as a set of solvable problems, rather than as deliberate misbehavior, Can help all parties feel better and behave more productively. Teachers, parents, and clients can

learn ways to create and maintain physical environments that minimize auditory and visual distractions during tasks that require concentration and to modify tasks to allow well-timed breaks and opportunities for feedback. They can learn to structure and support a well-regulated routine of daily activities, with established and balanced patterns of sleeping, eating, work, and play.

Occupational therapists most frequently use standardized tests as in-depth assessments of various areas of occupation and performance skills.

Standardized tests are used for four main purposes:

1. To assist in the determination of a medical or educational Diagnosis
2. To document a child's developmental and functional status
3. To aid the planning of an intervention program
4. To measure variables in research studies

following are the assessment scales used in ADHD

## Social Skills, Home Life Routine And Participation in Household Tasks in Family

- Child Routines Inventory(CRI)
- Home Situations Questionnaire(HSQ)
- Perceived Efficacy and Goal Setting System(PEGS)
- Social Skills Rating System(SSRS)

## Academic Functioning

- Academic Performance Rating Scale
- School Function Assessment(SFA)
- School Situations Questionnaire(SSQ)

## Behavior

- Behavior Assessment System For Children
- Child Behavior Checklist
- Adolescent Behavior Checklist

## Sensory and sensory motor processing

- Sensory Integration and Praxis Tests
- Sensory Profile

## Visual motor integration

- Test of Visual Perceptual Skills(TVPS)- To determine the visual perceptual strengths and weaknesses of students.

It includes 18 items in each of the following seven perceptual areas:

1. Visual Discrimination: the individual is asked to find the one image, in a field of five similar images, that exactly matches the presented target image.
2. Visual Memory: the individual is presented with a target image for five seconds, is asked to remember it and then find the image, in a field of four images, on the following page.
3. Spatial Relationships: the individual is asked to find the one image, in a field of five images, that is different from the rest.
4. Form Constancy: the individual is asked to find the one image, in a field of four or five images, that matches the presented target image. The matching image may be larger, smaller, rotated, and/or embedded within a larger design.
5. Sequential Memory: the individual is presented with an image of a sequence of elements for five seconds, is asked to remember it and then find the image with the same sequence, in a field of four images, on the following page.
6. Visual Figure-Ground: the individual is asked to find a target image that is embedded in one of a field of four complex designs.

7. Visual Closure: the individual is asked to match an incomplete target image to the correctly completed image in a field of four.

## Occupational therapy intervention in patients with ADHD

### *Behavioral approaches-*

Behavioral approaches represents a broad set of a specific interventions that have the common goal of modifying the physical and social environment to alter or change behavior (AAP,2001). They are used in the treatment of ADHD to provide structure for the child and to reinforce appropriate behavior. Types of behavioral approaches include behavioral training for parents and teachers( in which the parent and/or teacher is taught child management skills), a systematic program of contingency management(e.g. Positive reinforcement," time outs", response cost, and token economy), clinical behavioral therapy(training in problem solving and social skills), and cognitive-behavioral therapy(e.g. Self monitoring, verbal self instruction, development of problem solving strategies, self-reinforcement)these approaches are designed to use direct teaching and reinforcement strategies for positive behaviors and direct consequences for inappropriate behavior.

### *Behavior modification*

Behavior modification has proven effective in children and adolescents with ADHD for improving both social and academic behaviors. Behavior modification includes strategies such as rewarding positive behavior changes; parents and teachers then use the behavioral skills in their daily interactions with their children resulting in improvement in the children's functioning.

In behavior modification, reinforcement is often used and detailed records of the child's responses are kept. Often a positive or desired behavior needs to be reinforced, or a negative or undesirable behavior (e.g., eye poking, uncontrollable tantrums) is ignored, discouraged, shaped, or extinguished, depending on the intensity and potential for harm to self or others.

### *Cognitive Strategy training*

Cognitive strategy training is one of the cognitive approaches that is a top-down or occupation based approach It consist of five problem solving steps- task analysis, anticipation of the difficulties, exploration and selection of task specific strategies, application of a strategy to the task, and evaluation of its improvement this training teaches the child to identify, develop and use cognitive strategies to perform daily occupation effectively

### *Cognitive- behavioral therapy*

Cognitive behavioral therapy(CBT) integrates the cognitive strategy training with the behavioral modification techniques. It is an action oriented form of therapy that assumes that maladaptive or faulty thinking patterns cause maladaptive behavior and negative emotions
Instructional strategies that are used in this kind of therapy include modeling, role playing, and self instruction. Cognitive Behavioral Training attempts to increase self control and problem solving abilities by helping children with ADHD

### *Sensory Integration*

Children's with ADHD may have sensory modulation challenges. Sensory modulation difficulties are often the basis for interventions for children with ADHD, especially in addressing attention within classroom.

It enhances their ability to process low level sensory information related to alertness, body movement and position, and touch. Intervention may help children pay more attention to high level sense of hearing and vision.

sensory integration approach help child with ADHD to use self-regulation strategies(sensory based activities or sensory diet) and increase their school performance such as in seated behaviors, ability to listen and attend, finishing class work, and improvements in regulation of aggressive behavior

### *Social Skills Training*

Occupational therapist working with ADHD often use social skills groups to promote coping effectiveness and occupational performance. Intervention approaches may also involve education and counseling(individual, couple and family) consultation with professionals in schools and community programs, peer support and support groups, and development of supportive social network

Social skills training can help children or adults with ADHD to learn new behaviors, problem solving strategies and methods of dealing with their interpersonal relationships, such as using appropriate communication skills.

Following are some suggestions for practices that may be helpful for parents with a child with ADHD

## Tips for home

Caring for children with ADHD may be challenging, but it is important to remember that these children can learn successfully. It is critical that parents remember that some of child's disruptive behavior is a manifestation of the disability and that the challenge is finding ways to help their child change the inappropriate behavior. key to this is remembering to focus on the need for structure and routine for your child's daily schedule and thereby reinforcing the importance of learning self control and self regulation. the following are suggestions for parents

- focus on discrete rewards and consequences for appropriate and inappropriate behavior:

1. tangible rewards and treats
2. movie night for a good week at school
3. removal of privileges
4. time out from reinforcing activities; the child is essentially removed from situations that foster inappropriate behavior

- set a daily routine and stick to it. Bedtime and preparation for school are much easier if there is a structure already in place
- have a tangible reminders

1. a big clock in a bedroom
2. charts for chores
3. assignment pad to record homework and a specific folder to put work upon completion and
4. gain the child's attention before speaking to him or her. have the child repeat back directions for things that are really important

- avoid the following

1. repeating patterns of inappropriate behavior followed by ineffective punishment
2. administering consequences without prior warning or without the child understanding why he or she is receiving them

3. responding inconsistently to inappropriate behavior

## Tips for school

A student with ADHD can present unique challenges in the classroom. Inattention, hyperactivity, or impulsivity can be a source of frustration, but there are ways teachers can help students with ADHD to improve educational experience and control the symptoms of the disorder. It is important for teachers to be aware of coexisting conditions like learning disabilities as well as reinforcing the importance of classroom and instructional structure.

The following are tips for teachers:

- Work on the most difficult concepts early in the day
- Give directions to one assignment at a time instead of directions to multiple tasks all at once
- Vary the pace and type of activity to maximize the students attention and
- Structure the students environment to accommodate his or her special needs. For example, the student can be seated away from potentially distracting areas(such as doors, windows, and computers) or seated near another student who is working on a shared assignment.

Providing supervision and discipline:

- Remain calm, state the infraction of the rule, and avoid debating or arguing with the student
- Enforce classroom rules consistently
- Avoid ridicule and criticism
- Avoid publicly reminding students on medication to take their medicine

Providing encouragement

- Reward more than you punish, in order to build self esteem
- Praise immediately any and all good behavior and performance
- Change rewards if they are not effective in motivating behavioral change.
- Find ways to encourage the child

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# Chp. 16 Learning Disability

**Mr. Vishal Ganar, Dr. Nishigandha Doke**

## History

Shortly after World War II educators began to notice that increasing number of children with average or above average intelligence experienced difficulty in learning. Origins of research studies related to learning disabilities are traced back to understanding of brain pathology in adults. Many researchers including Franz Joseph Gall, Pierre Paul Broca, Carl Wernicke, etc studied human brain- living and dead to understand various cognitive effects caused by damages caused to different brain parts. Adolph Kussmaul, in 1877, coined the terms "word- blindness" and "word-deafness" after he encountered intelligent adult people who had difficulty in reading or naming things but could write fluently with dictation or spontaneously. It was John Hinshelwood along with Morgan who was a general practitioner in England who first noted that learning disability can be inherited and primarily manifested in males.

Morgan published the first case of dyslexia, a word derived from the Latin word "dys", which translates to 'difficult', and the Greek word "lexia", which translates to 'words'. Dyslexia,thus, literally means, "difficulty with words".

## Definition

In 1980s, National Joint Committee on Learning Disabilities (NJCLD) defined the term learning disability as:

"a heterogeneous group of disorders manifested by significant difficulties in the acquisition and use of listening, speaking, reading, writing, reasoning or mathematical abilities. These disorders are intrinsic to the individual and presumed to be due to Central Nervous System Dysfunction. Even though a learning disability may occur concomitantly with other handicapping conditions

(e.g. sensory impairment, intellectual disability, social and emotional disturbance) or environmental influences (e.g. cultural differences, insufficient/inappropriate instruction, psychogenic factors) it is not the direct result of those conditions or influences."

The following Federal definition by the U.S. Government in Public Law 94-142 of Learning Disabilities has been adopted in India.

"Specific Learning Disabilities means a disorder in one or more of the basic psychological processes involved in understanding or in using language, spoken or written,

which may manifest itself in an imperfect ability to listen, speak, read, spell or to do mathematical calculations. The term includes such conditions as perceptual handicaps, brain injury, minimal brain dysfunction, dyslexia and developmental aphasia. The term does not include children who have learning problems which are primarily the result of visual, hearing or motor handicaps, or mental retardation, emotional disturbance or environmental, cultural

or economic disadvantages." (Federal Register,1977, p. 65083) (Karanth, 2002).

According to the ICD DSR (International Statistical Classification of Diseases and Health Related Problems), learning disorders (LD) refer to a significant deficit in learning due to a person's inability to interpret what is seen and heard, or to link information from different parts of the brain (GEON, 2005).

## Learning disorders can be classified into three major types:

1. Disorder Of Written Expression (DWE)
2. Reading Disorder (RD)
3. Mathematics Disorder (MD).

## Epidemiology

### *Prevalence*

In the eastern world, Learning Disability was earlier considered a problem of countries where English is spoken. As the reporting of these difficulties was comparatively lower in Asia, there was a lack of concern about LD. On the other hand, Western scholars attributed the low prevalence of LD in Asian countries to the general lack of awareness and sensitivity among educationists, overcrowded classrooms, etc. The high prevalence of LD in the West was attributed to the complex nature of alphabetic writing systems such as English from the point of view of the Asians. (Karanth, 2002).

However, during the last decade or two, there has been an increasing awareness and identification of children with LD in India. Despite this growing interest, we still have no clear idea about the incidence and prevalence of LD in India.

### *Sex distribution*

Sex differences in Learning disabilities show that there is a ratio of 5:1 within boys and girls as seen in researches. But theories also state that these differences in data can be caused due to different factors like test bias, referral bias or even biological predisposition in the test sample.

### *Social status*

Social status of children with LD cannot be correlated with the disability but lack of appropriate screening, early intervention and prevention can cause the illusion of high prevalence of LD in children of low socioeconomic status. Also, due to lower literacy rates in the families belonging to the below poverty line, education is given lesser importance and LD is not screened, prevented or intervened for.

## Early diagnosis

Identifying learning disabilities early can help the children get support early and find ways of interventions that will help them overcome the obstacles in a better manner thus reducing the built-up stress which is developed in the later part of life. The purpose of early identification is to determine which children have developmental problems that may be obstacles to learning or identifying children who are at a risk. Development in infants, toddlers, and preschoolers vary in terms of rates and patterns of maturation. For some children, differences and delays in abilities are temporary and are resolved during the normal course of development. For other children, delays may persist in different domains of functioning which makes it necessary to screen and assess them for any developmental disorders and prevent stress caused due to it.

## Social interaction

Samuel T Orton, in his study of emotional aspects of learning disability, found that the majority of preschoolers with learning disability are happy and well adjusted. Their emotional problems begin to develop when early reading instruction does not match their learning style. Over the years, the frustration mounts as classmates surpass these students in reading skills. The frustration of children with LD often centers on their inability to meet expectations. Their parents and teachers see a bright, enthusiastic child who is not learning to read and write. The pain of failing to meet other people's expectations and inability to achieve their own goals cause them more stress. This is particularly true of those who develop perfectionistic expectations in order to deal with their anxiety. They grow up believing that it is "terrible" to make a mistake. they grow up to develop anxiety, depression, self image and problems in family and peer groups.

## Diagnosis and Clinical features

### *There are four criteria prescribed for diagnosis as per DSM 5*

Criteria A: Specific learning disorder is persistent difficulties learning key-stone academic skills with onset during the years of formal schooling. Key academic skills include reading of single words accurately and fluently, reading comprehension, written expression and spelling, arithmetic calculation, and mathematical reasoning. Academic skills unlike other developmental skills like walking, talking have to be taught explicitly (DSM 5, 2013). As discussed, reading is a language based skill and the specific linguist skills affected in a person with SLD are the meta phonological skill.

Assessment and remediation of metaphonological skills comes under the domain of a speech language pathologist. The need to target metaphonological skills are not mentioned in the DSM 5. Word reading involves connecting written expressions to corresponding sounds (phonemes/ words /sentences) which again depends on how well the reader knows about the sounds of the language, the rules by which the sounds and the corresponding written forms are connected while standing alone and when they form words and sentences. There is no instance where a person can develop reading and writing without knowing to speak the language. Neither reading nor writing can be used to teach oral language. The learning difficulties are persistent and not transitory in children and adolescents, meaning that the individual is not catching up with peers for at least six months inspite of extra help at home or school. Evidence of persistent SLD is suggested to be derived from previous cumulative school reports, portfolios of the child's work or clinical interview. (DSM 5, 2013).As many as 75-80% of children with SLD exhibit a language disorder and speech disorder (National Institute of health, 2002) The DSM 5criteria do not suggest of having a language evaluation done.

Criteria B: The individual's performance of the affected academic skills is well below average for age or average performance that is sustainable by extraordinary high levels of support. In children, the low academic skills cause significant interference in school performance (reported by teachers and teacher's grades or ratings). In adults, there is avoidance of activities that require the academic skills. On the basis of clinical judgment a more lenient threshold may be used when SLD is supported by

converging evidence from clinical assessment, academic history, school reports or test scores, as standardized tests are not available in all languages the diagnosis may then be based in part on clinical judgment of scores on available test measures. (DSM 5, 2013).

Criteria C: A third core feature is that the learning difficulties are readily apparent in the early school years in most individuals. However, the learning difficulties may not manifest fully until later school years, by which time learning demands have increased and exceed the individual's limited capacities (DSM 5, 2013). This indicate the scope of identifying SLD quiet early during initiation schooling (around 4-5 years of age), most of the tests which can be used at this stage would be tests which are language dependent hence the speech therapist needs to be involved at this point.

Criteria D: Another key diagnostic feature is that the learning difficulties are considered "specific, "for four reasons. That is they are not attributed to following reasons including, Intellectual disabilities; global developmental delay; hearing or vision disorders, or neurological or motor disorders. (Diagnostic and Statistical Manual of Mental Disorders,2013)

## Causes:

### *Genetic or Heredity Factors*

Studies that show that learning disabilities run in families give evidence that learning disabilities are genetically determined with genetic traits manifesting themselves in the neuroanatomy and neurophysiology of the child. The concordance rate is higher for identical than fraternal twins for reading disorder. Some findings suggest that genes on chromosome 15 or 6 may cause reading disorder for a minority of individuals (Pennington, 1991). Critics of research conducted to find genetic base mention that high rates of learning disabilities within families could be a result of family environment as much as genetics.

### *Neurological Factors*

Neurological theories state that learning disabilities is a reflection of structural damage or improper development of the nervous system. Such problems could occur during prenatal or postnatal period, as the nervous system is developing which can be caused due to head injury, lack of oxygen, exposure to toxins, seizures and nutritional deficiencies or any other causes that may contribute to central nervous system damage. Studies have shown EEG abnormalities in some children with learning disability. Various neuropsychological deficits are associated with learning disabilities, such as in visuospatial perception, auditory perception, semantic memory and phonemic discrimination. Children with reading disorders have left-hemisphere deficits and autonomic disorders such as eczema, allergies. Children with mathematical disorders have deficits in neuropsychological functions indicative of right hemisphere damage (Rao, 2003).

### *Environmental Factors*

Learning disabilities can be a direct result of environmental factors. Research shows that factors such as malnutrition, prematurity, poor prenatal and postnatal health care, stress, poor parenting and teaching can have a negative impact on learning by creating conditions in which the function of the brain is hampered. Substance abuse such as alcohol and other drugs, inappropriate learning models, emotional distur-

bances, social and cultural deprivation can contribute to learning disabilities.
Summarizing the findings of several research studies, Silver and Hagin (2002), concluded that the evidence points to the immense influence of poverty and inappropriate and/or inadequate stimulation on the development of learning disorders in children.

## *The Nature and Symptoms of Learning Disabilities:*

Learning disabilities are defined based on the input-integration-memory-output model (Silver,1991, 1996). The first task in learning is to receive the information and record it in the brain (input). Once recorded, this information must be handled in such a way that it can be understood (integration).
The third process is storage and retrieval (memory). Finally, information must be communicated from the brain (output).

## *Input Disabilities*

Learning disability involves perception. A person might have a visual perception disability or an auditory perception disability. Visual perception disabilities involve difficulty in distinguishing subtle differences in shapes. For example: misperceiving "d" and "b" or "p" and "q" or 6 and 9. The person might not be able to focus on the relevant stimulus in the field of vision. Thus, they have difficulty in visual figure-grounding tasks. Some children have trouble with visual depth perception like they may fall off chairs or bump into things.
Auditory perception disabilities might be in the area of distinguishing subtle differences in sounds, leading to misunderstanding of what is being said. Some words sound similar such as "hair" and "air", "ball" and "bell". Some children might have difficulty with auditory figure-ground, confusing what sounds to listen to when there is more than one source. Some children cannot process sound inputs quickly. Some children have auditory lag. They have to think about what is heard before understanding what is said.

## *Integration Disabilities*

Brain processes an information in three basic steps. A single stimuli has to be (1) sequenced correctly and then (2) understood in the relevant context (abstraction) and then (3) organized with all other stimuli into a concept. One can have an integrative disability in each of the areas of sequencing, abstraction or organization. The disability might be for either or both visual or auditory inputs.

Sequencing disabilities can result in confusing inputs e.g. writing 21 rather than 12, reading "was" rather than "saw". Such children can memorize the months of the year but be unable to say what comes after any month without starting with January and working their way up.

Abstraction difficulties result in trouble picking up the subtle meanings of words. They might miss the meanings of jokes, puns, or idioms. They find it difficult to understand the underlying meaning of the conversation or even the situation.

Organization disabilities can result in difficulty pulling multiple parts of information into a full or complete concept. Such children can learn individual facts but may not be able to integrate them into a full concept. Such children also find it difficult to deduce moral from a story. They show this in other aspects of life as well like a disorganized room, managing time or making

plans might be difficult.

### *Memory Disabilities*

Children with learning disabilities usually have excellent long-term memory but they might have difficulty, however, in short-term memory, i.e. the ability to concentrate on recent information and store it. As soon as they stop attending to the information in front of them, they will forget it. These students need many more repetitions to process the information into long-term memory than the average person. They can learn, but they will need more time and effort to do so. One might have a visual-short-term memory disability or an auditory-short-term memory disability.

### *Output Disabilities*

People with learning disability may experience difficulty getting information out of the brain through oral communication (a language disability) and/or through the use of muscles (a motor disability).
Most children have little difficulty with spontaneous language, which refers to self-initiated talking where they find it difficult to organize thoughts and find words before speaking. Children might have difficulty with demand language, which refers to situations where the child must respond without pre-planning. Such children might speak with ease when they initiate the conversation, however, they might not be able to respond when asked a question or requested to speak.

Some children with learning disabilities will have trouble coordinating the use of large muscle groups, i.e., gross motor disability characterized by clumsiness or difficulties with activities like running, climbing, swimming. Others may have difficulty coordinating small muscle groups, i.e., fine motor disability which is characterized by poor handwriting, difficulty with written language tasks such as spelling, spacing and punctuation.

## Treatment:

The primary mode of treatment for children with learning disabilities is special educational planning and other educational services. Early intervention can be used not only to intervene with learning disabilities sooner, but also to possibly prevent disabilities in young children at risk for developing them.

### *Reading*

Current remediation interventions for children with reading disabilities aim to improve the child's sight reading and phonics.

Sight reading (memorizing words by sight) may be addressed by vocabulary-building exercises, sight reading exercises, learning word roots, prefixes and suffixes.

Programs to improve phonics (recognizing and remembering the association between letters and sounds) such as providing the child with extra experience that integrates several senses into phonemic awareness. For example, the child selects a word, which is written in large letters in a flash-card. The child might trace the word as he says the word sound and looks at a picture of the word (tactile, auditory and visual senses).

### *Mathematics*

Treatments for mathematics disabilities combine teaching mathematics concepts with continuous practice in solving math problems. Concrete objects, pictorial rep-

resentations such as graphs, analogies and logical explanations are used to make abstract concepts meaningful. Strategies for translating word problems into arithmetic problems can be directly taught (e.g., the words followed by "and", "together", etc usually mean that things will be added e.g., how many pencils did Ram "and" Shyam have together). Learning math rules (e.g.,BODMAS; the digits in multiples of 9 always add up to 9 or a multiple of 9), math games, computer games where the focus is on problem solving activities, including word problems rather than only computation.

## *Writing*

Treatment for writing disabilities includes handwriting practice, direct practice in spellings and sentence writing as well as a review of grammatical rules. Spelling problems frequently involve deficits in phonemic awareness and word recognition, therefore interventions focus on mostly spelling errors. Spelling skills can be promoted by teaching the child spelling rules e.g., "i" before "e" except after spelling of word roots, as well as prefixes and suffixes, and mnemonic strategies for remembering the spelling of specific irregular words. Strategies to make spelling game, like for example, spelling bee, finding the hidden word; and to promote repetition learning are used for children with spelling deficits.

When there is a deficit in written expression such as communicating ideas in writing, strategies such as sentence combining, maintaining a diary, letter writing, writing invitations or gift lists and other naturalistic writing exercises are often used. Learning to write an outline before writing a story is used to promote logical, sequential, comprehensive flow in writing. For children with good verbal expression and poor written expression a verbal-to-writing intervention can be used. First the child dictates to an adult who writes what the child says. Next the child dictates to a tape recorder and later transposes own words into writing. Next, the child dictates only a few sentences into the tape recorder and writes the sentences before dictating a few more and so on. After this step is mastered the tape recorder is removed and the child says the words out loud, pausing to write them periodically. Finally the child gradually says the words more and more silently until the child is writing while thinking (but not saying the words).

## *Behavioural Interventions*

Most of the times behavioural problems accompany learning disability. Behavioural characteristics such as inattention and hyperactivity have social consequences, putting a strain on interpersonal relationships and lead to negative self-evaluation. Biofeedback and relaxation training have been identified as relatively non-intrusive methods for dealing with these problems. A number of studies have demonstrated that biofeedback and relaxation can improve the behaviour and emotional well being of children with learning disabilities. (Amerikaner & Summerlin, 1982; Carter & Russell,1985; Loffredo et al., 1984).

Learning disabled children have been found to have an external rather than internal locus of control (Short & Weissberg-Benchell,1989) and this is likely to hamper their academic endeavours. In order to combat this locus of control problem, several researchers have recommended that students with learning disabilities be subjects to attribution training in which they learn to attribute success in school work to

such internally controlled factors as study time and effort (Hoy, 1986; Tollefson, Tracy, Johnson, & Chatman,1986).

### *Social Skills Training*

Several procedures have been used to improve the social skills performance of students with learning disabilities. Pray, Hall and Markley recommend more emphasis on interpersonal skills for students with learning disabilities. To foster peer interaction social skills, curriculum needs to focus on four main areas: conversation skills(e.g., introducing oneself, asking and answering questions),friendship skills (making friends, joining group activities, giving help), skills for difficult situations (accepting and giving criticism, resisting peer pressure) and problem solving skills (negotiating, persuading, asking for feedback).

### *Peer Tutoring*

Several studies have focused on the effectiveness of peer tutoring (an instructional arrangement in which the teacher pairs two students in a tutor-tutee relationship to promote learning of academic skills) for students with learning disabilities. Peer tutoring has been found to improve academic skills, foster self esteem, develop appropriate behaviours and promote positive relationships and
co-operation among peers (Mercer, 1997).

### *Family Interventions*

It is very important for a family to identify and accept a child's disability in learning to avoid various other psychological trauma that is associated with denial and disappointment in the child as well as parents. Sharma (1993), found that parents of learning disabled children were having very poor orlow expectation of their child's academic achievement. These parents also mostly perceived their children as socially incompetent with several problems. Parental counselling and family therapy would help to improve patterns of communication within the family members, between the parents and the child, thereby contributing to the development of a positive self concept in the learning disabled child.

## Role of an occupational therapist in learning disability

**The occupational therapy evaluation process among people with learning disabilities:**

The purpose of an evaluation is to identify the areas of strengths and weakness of individuals with learning disabilities in performing various activities and occupations and analyzing the tasks these individuals need to perform. The process of evaluation starts with an interview with the person and /or his/her family members in order to gather information about the person's occupational history, wishes and needs, routines and habits, areas of interest and values. The occupational therapist then chooses a standardized assessment tool to assess the underlying causes of the individual's difficulties and the various demands of the environment in which he/she functions. The source of the difficulties may lie in various deficits in sensory, motor, cognitive functions, as well as other areas. The process is completed by integrating the findings of the interview, observations and standardized assessments to form a comprehensive picture of the individual's capacities, the source of his/her difficulties and the manner in which the person copes with the environmental demands.

**Following are the standardized tests used in evaluating specific areas**

1. Visual-motor integration(VMI)
2. Miller assessment for preschoolers
3. Sensory integration and praxis test(SIPT)
4. Test of visual motor skills(TVMS)
5. Handwriting tests
6. Reading achievement tests
7. Mathematical achievement
8. Tests of motor proficiency
9. Peabody development motor
10. Bruinin ksostretsky test of motor proficiency

**Intervention of occupational therapy among individuals with learning disabilities:**

Occupational therapy intervention approaches and methods for individuals with learning disabilities take into account the person him/herself with respect to the his/her abilities and disabilities and learning style, as well as to the environment in which the intervention is carried out. In addition, the approaches are adapted with respect to the occupations that individuals want and need to perform in the environments in which they functions. On the basis of an analysis of the factors mentioned, occupational therapists suggest an intervention program that focuses on the person, the task to be performed and the environment in which it is to be performed. Occupational therapy intervention may be carried out as direct, individual therapy; through a group process (group therapy); and / or by consulting with the individuals, their families and the other people in their environment (educational teams, work teams, etc.)The intervention is directed towards adapting the activities and / or the environment of the individual in order to enable him/her to realize his/her full potential.

The occupational therapy evaluation and intervention are carried out in various environments, such as in a clinic and/or in the person's natural environment (at home, class, playground, work place, etc.).There are various approaches to treatment of learning disorders. Focus is on development of functional competence. Therapy is directed at following areas-

A. Development of efficient motor behavior. Gross motor skills like e.g. Jumping, ball throwing, fine motor skills like tying shoelaces, spatial body awareness
B. Development of efficient sensory perceptual skill integration- directionally, tactile discrimination, laterality, balance and body spatial organization, time orientation
C. Development of conceptual and body awareness skills
D. Development of mature social behavior

**Occupational therapy areas of focus with people with learning disabilities**

The occupational therapy evaluation and intervention process for individuals with learning disabilities occurs throughout the life cycle - from early childhood until late adulthood - in a variety of occupational areas:

- **Independence and participation in activities of daily living:**
  Evaluating and improving the person's ability to perform self-care (e.g.,dressing, eating and personal hygiene), as well as to acquire the habits and skills required for instrumental activities of daily life (IADLs) within the home and community environment (such as the efficient use of means of communication devices, household

management and the use of public transportation or driving).

- **Learning:**
  Evaluating and improving the individual's ability to cope with the demands of different academic settings through efficient information processing, improving organizational abilities, developing strategies, improving writing skills, and so forth.
- **Work:**
  Evaluation and improvement of work skills and habits to allow for integration into the work place (e.g., punctuality, work relationships, complying with the rules and norms of the work place), consultation regarding selecting a suitable occupation and recommendations regarding the accessibility of the work environment.
- **Play and leisure:**
  The evaluation of play skills and leisure patterns, the cultivation and development of leisure opportunities, fostering internal control and satisfaction from play and leisure activities (e.g., guidance regarding adapting activities/ clubs to suit the needs and strengths of the individual).
- **Social participation:**
  Evaluation and development of social skills (e.g.,teaching strategies for self-expression and personal empowerment, helping improve interpersonal interactions as well as in identifying andcomplying with accepted social codes and norms). Development and improvement of basic skills that constitute a basis forlearning

**Perceptual motor approach**

Following things child should develop to move effectively in the environment

1. **Posture:** for an individual to be able to explore and learn effectively posture must be well developed
2. **Laterality:** it permits a child to recognize difference between b & d
3. **Directionality:** child must be able to coordinate his visual signals with his hand movements.
4. **Body image:** child must be able to maintain his own body posture, laterality, directionality with reference to his body and its position in space. Inability to move one part of the body independently of other parts is also due to poor body image

Kephart developed an evaluation of perceptual motor skills and designed remedial exercises for each of the areas

- ***Categories of training included-***

1. Exercise to develop balance, posture and laterality
2. Perceptual motor matching in which eye hand activities are used
3. Ocular training like visual tracking
4. Training in form perception such as matching forms or visual patterns

- ***Remediation suggested-***

1. Walking board, Balance board, Trampoline
2. Games and rhythmic activities
3. Training for ocular control includes-visual fixation, ocular pursuits, chalkboard games etc

**Occupational therapy areas of focus with people with learning disabilities**

Following are the manifestations and therapist approach used in child with learning disability

1. ***Eye movements-*** lose their place while reading and keep their fingers on page,

eye movements are jerky, clumsy
***Treatment-*** Therapist can tell the student to follow objects or to attend to stimuli in specific direction e.g. telling a child to follow a lit object in left to right orientation. Flash cards can be used, for dev. Eye movements' control

2. ***Eye hand coordination-*** child is unable to accurately duplicate drawings or to position objects. Children have a problem batting a ball or catching a ball
***Treatment-*** Any type of activity involving hands and vision. Right left, up down and clockwise can be introduced and involves child's entire body. Chalkboard routine-drawings horizontal lines, vertical lines follow dots will improve eye hand skills

3. ***Visual form perception-*** child experiences poor memory or inability to attend to details. Better form sequence reversal in comparison of shapes and forms and they see lines as jumble of words
***Treatment-*** Flash cards, space writing-quick visual presentation of symbols students draw in air, drawing geometric puzzles, erase and then rewrite from memory

4. ***Visual perception-*** reversal/inversion: poor left right discrimination, poor eye handcoordination, confuseddominance of eye hand
***Treatment-*** Tracing of letters, words, give tactile stimulation of various types, recollecting from memory, teaching to concentrate on letters
5. ***Form constancy:*** child cannot recognize similarities between shapes, sizes and colors. Inability to transfer learned skills to new material
***Treatment-*** Use of 3D objects. In reading care should be taken to be consistent in the form used.
6. ***Figure ground perception and distractibility-*** child cannot keep his attention to a fixed point and is unable to focus on specific item at will. Child appears clumsy and inconsistent. While reading loose place
***Treatment-*** Written material should be free of confusing background. Writing should be extra-large. Child should be told to hold line he is reading with plain card. Activity should be divided into stages and accordingly the instruction should be given. In severe cases, highlight the outline of figures in dark color
7. ***Problems with spatial relations:*** relation between two objects and child fails in sequential tasks, cannot string beads. He sees symbols as jumbled, reversed or in random order.
***Treatment-*** Practice comparing large 3D and self-single dimension small objects. Start writing with semi abstractions, forms, and shapes and then to writing. Maze type e.g. Joining dots. Memory practice by producing forms from memory
8. ***Position in space-*** right left discrimination problem so difficulty in placing the objects

**Development and improvement of basic skills that constitute a basis for learning:**

***Cognitive skills:*** Training and improvement of attention, perceptual and memory processes, information processing as well as training and improvement of ability to classify and generalize.

***Meta-cognitive skills and self-management functions (Executive Functions):*** Enhancing awareness and acquisition of

knowledge about the individual's personal learning and thinking processes; knowledge relating to the task's requirements and strategies to cope with different tasks and teaching him/her to apply them. In addition, teaching methods needed to anticipate, plan and use feedback when executing various actions. All of these help individuals improve their ability to solve problems. One point of emphasis of occupational therapists in this sphere is the organization and self-management in time and space, such as planning and maintaining a daily schedule while balancing between the various activities, organization in preparations for travel, managing the routine organization of personalbelongings and spatial orientation in different locations.

***Development of specific skills necessary for learning:*** These include the development and improvement of writing skills – developing the ability to write within a reasonable amount of time without investing unnecessary energy, such that the written document is legible and comprehensible. Writing skills are needed for writing tasks that include copying, dictation, preparing homework, and the like. Intervention includes direct intervention as well as recommendations for adaptation to the school's requirements and of various accessories (e.g., type of pencil, notebook, laptop computer and accompanying accessories), as well as teaching keyboarding skills.

***Development of compensatory techniques:*** Occupational therapists are involved in developing compensatory techniques to support the function of individuals with LD, including : The use of assistive devices and technology - such as a computer, an adapted keyboard, an electronic calendar, time-tables (for use at home, in school and at the work place), a slanted writing board, wide pencil grips and others.
Adaptation of the task - such as reducing the length of written tasks to suit a student's endurance, adapting the time required for learning/working according to the person's attention span, adapting the instructions for an activity and adapting the rules of a game.

***Adaptation of the environment -*** such as structuring and designing the study and work environment in accordance with ergonomic principles, which relate to matching a person's characteristics to the demands of his/her work environment. Examples include adjusting the height of the chair and the table, the type of pen, the placement of the student's seat in the classroom and atthe workplace, adjusting his/her computer set up, etc.

**Intervention for co-occurring deficits that accompany learning disabilities**

Research findings report that in about 50% of the population coping with learning disabilities, there are accompanying deficits such as attention deficit hyperactivity disorder (ADHD), developmental coordination disorder (DCD)and sensory modulation disorder (SMD). In the therapeutic intervention of individuals with learning disabilities, occupational therapists place great importance on simultaneously addressing additional co-occurring deficits.

***Attention deficit hyperactive disorder (ADHD) –***
occupational therapists help by increasing the awareness of individuals and their significant others to their difficulties in attention and the manner in which this influences their function. Therapists also

guide the individual in developing strategies for regulating their level of arousal, modulating their responses and applying self-control to suit the task's requirements. Occupational therapists may also recommend technological adaptations in order to improve function (such as adapted computer programs), the use of assistive devices (such as weighted vests to reduce "out of task", irrelevant behaviors), and/or assist in the adaptation of the environment or task according the type of attention deficit(for example, adapting an occupation that requires transitioning between many activities and/or minimizing external stimuli).

Developmental coordination disorders (DCD) – occupational therapists assist a person with deficits in performing motor and perceptual-motor skills. Examples include improving postural control, coordination and power regulation. Such difficulties may be manifest in the performance of leisure activities, driving, sports, writing, playing an instrument, as well as in the manipulation of various objects and tools (such as a pen, ruler, eraser, keyboard, lab accessories, eating utensils, operating appliances, etc.).Intervention also focuses on improving movement planning and execution of movements in time and in space.

***Deficits in sensory processing and modulation –***

occupational therapists help a person with deficits in hyper- or hyposensitivity to touch, movement, visual and auditory stimuli and/or smells. These are sometimes expressed as avoidance of certain materials used for crafts or for learning, avoidance of participating in games on the playground, or discomfort in a noisy environment, and so forth. For example, occupational therapists may guide the person, and if needed, his/her family members and significant others in his/her environment, and/or in adapting the environment or the task, such that it will suit the individual's sensory needs

**Conclusion**

Learning disability, an educational problem, forms an important cause of failure in school in otherwise capable children. The Central Board for Secondary Education (CBSE) and the Indian Council for Secondary Education (ICSE) recognize learning disabilities as a disability. As a result, specific learning disabled children have been formally granted with the benefit of availing certain provisions (e.g., extra time during written exams, dropping algebra, geometry for lower grade of mathematics and work experience) from standard I to XII. These provisions are meant to help the child to cope up in a regular mainstream school.

Learning disabilities are often accompanied by problems of attention and concentration, organization, mood disturbances, anxiety and depression, low self-esteem and deficits in social skills. Failure and frustration may appear early in the child's school experience where reactive behaviours of avoidance, depression, aggression and disobedience set the stage for subsequent emotional and behavioural problems. Parental reactions of non-acceptance, depression, guilt, all undermine support for the child (Silver & Hagin, 2002). Teachers may see the child as dull and lazy and may not see the child's special needs. The emotional and behavioural problems of the learning disabled child need to be given prompt attention with appropriate psychological interventions and parental counselling.

The multifaceted nature of learning dis-

abilities, the large number of children involved and limited school resources makes it imperative that trained teachers, special educators, psychologists and social workers collaborate together in the education of these children. Remedial education is the basis for management.

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# Chp. 17 Intellectual Disability

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## Introduction:

"Developmental Disabilities" is a broad term that includes intellectual disability and other disabilities that are apparent during childhood. Developmental disabilities are long standing cognitive or physical or both that are chronic in nature. Developmental disabilities can be largely physical issues, such as cerebral palsy or can be physical and intellectual disability, for example Down syndrome.Intellectual disability comprise of the "cognitive" part of this definition, that is, a disability that is broadly related to thought mental processes.

## Causes:

There are a number of causes. Our understanding of the causes of intellectual disability focuses on the types of risk factors (biomedical, social, behavioural, and educational) and the timing of exposure (prenatal, perinatal, and postnatal) to those factors.

Multiple causes do exist leading to intellectual disability. There is a complex relationship with cause and the one leading to intellectual disability being Genetic predisposition, Environmental insult, Developmental vulnerability, Heredity and environment.

Genetic Factors and Intelligence: Genes play a role in transmission of intelligence; the term general intelligence is the cognitive ability that assists in learning and memory, and is among the heritable genetic trait.

Environmental factors and Intelligence: Environment and intelligence studies the impact of environment on intelligence. Neuronal network are undifferentiated in babies and these Neurons make connections with neighbouring neurons, and these become more complex and more idiosyncratic as the child ages. The process continues as new experiences turn up and new network open their gate. It is likely that the growth in neuronal connections is largely due to an interaction with the environment, as there is not even enough genetic material to code for all the possible neural connections.

Classification of Intellectual disability can be done in various domains. The following are furnished with details of few of the causes of ID.

## How is ID Diagnosed?

The two Main criteria given by the American Association on Intellectual and Developmental Disabilities (AAIDD) to define and diagnosis Intellectual Disabilities are intellectual functioning and adaptive behavior. Psychological testing therefore becomes crucial in diagnosing the presence and degree of Intellectual disability.

The intellectual functioning and adaptive behavior of Intellectual Disabled persons cannot be assessed by using a single test or scale. More than one test is needed for providing a reliable and fair evaluation of person with ID.

In addition to the diagnostic function, the uses of psychological testing in ID are:-

1. It provides a profile of abilities and disabilities of the ID person which helps in training.
2. It can be used to evaluate the effects of therapeutic intervention by measuring change over time.
3. It provides prognostic information about the potentialities of the ID persons.
4. Aptitude test, and inters inventories combined with intelligence test are useful aids in vocational guidance.

There are three impotent areas of assessment in ID persons.

1. Measurement of the overall level of GENERAL INTELLECTUAL FUNCTIONING

1. IQ
2. DQ
3. SQ

Assessment of ID refers to a dynamic process determining the quantitative and qualitative needs of persons with disability. Ideally, this includes an appraisal of the client's mental, social and vocational abilities and typical behaviors.

The definition of ID has little significance or utility outside the context of assessment. The diagnosis of ID cannot be made without assessment; and provision of services is greatly influence by the adequacy or inadequacy of assessment procedures employed.

## Methods of assessment of intelligence

The existing methods for assessment of intellectual development in infants include mostly developmental scales as:

1. **Gesell Development Schedules** was the first assessment tool developed in 1925 for evaluation if mental abilities in infants and pre-school children. In provides norms for ages 4 weeks to 72 months and assessment in 4 major development areas namely, motor development, language development, adaptive behavior and personal social development. It provides an estimate of Development Age (DA) and developmental quotient (DQ).It is a useful tool in development diagnosis, identification of mental retardation and behavioral abnormalities.

2. **Bayley Scale of Infant Development** (1933-revised in 1969) provides motor and mental development indices. It has norms at half intervals from 2-5 months, and at one month interval from 6-30 months. It also includes infant behavior record. Indian norms (commonly known as Baroda Norms) have been constructed for this scales from 0-15 months for rural children and up to 30 months for urban children by Pramila Patak in Baroda.

   The other commonly used assessment tools are Griffith scale of mental development and Cattell's Scales of infant intelligence. Einstein's Scale of Sensory motor Intelligence based on Piaget's framework of cognitive development has appeared in the recent years.

   The use of these scales in developing countries in limited because of the problems of normative and constructive validity since these were concerned and standardized in highly industrialized societies.

## Binet Era

Modern intelligence testing began with Binet Era when Alfred Binet in collabora-

tion with Simon developed a fully fledged intelligence scale in 1905 for identification of mentally retarded children. It is an age scale and has norms from 3 years to adult level. There have been many revisions of this scale.

The first attempt at Indian adaptation of this test was made by Dr.C.H.Rice of Lahore in 1922 that developed Hindustani Binet Performance Scale for Indian population.

Another well known adaptation is by Dr.V.V.Kamat, who standardized it for measuring intelligence of Indian children. It gives an estimate of IQ on specific cognitive functions like, comprehension, memory and the like.

## Wechsler Era

A new era in intelligence testing was ushered by Wechsler tests. These are point scales and provide Verbal IQ. The well known scales are:

Wechsler Bellevue Scale (1939) has norms from adolescence to senescence.Wechsler Intelligence Scale for children (1947, revised in 1974). This test has been adapted for Indian children by Malin. It consists of two scales, namely, verbal scale and performance scale.Wechsler Pre-school and Primary scale of intelligence was developed in 1967 and can be used preschool children.Wechsler Adult Intelligence Scale has form 16 year onwards. Dr. (Mrs.) Ramalingaswamy has adapted this test fir Indian population.

The advantages of Binet and Wechsler scales are that have proven excellent predictors of scholastic achievement and late IQ. Rehabilites and validities are substantial. Limitations of these tests are that they require highly trained examiner, long time consumed in test administration and some test items are not suitable for developing countries.

## Newer methods of measuring intelligence

Information Processing Approach: A complementary to psychometric approach is information processing approach for the measurement of intelligence. Information processing approach is an attempt to understand human intelligence in terms of mental processes that contributed to cognitive task performance. While psychometric approach emphasizes on static mental structures, the information process approach analyzes how people solve difficult mental tasks and models are constructed on the setups of how these tasks are solved. The most commonly used examples are simple and choice reaction time tests.

The IQ test is a major tool in measuring intellectual functioning, which is the mental capacity for learning, reasoning, problem solving, and so on. A test score below or around 70—indicates a limitation in intellectual functioning. Mostly classified into four categories as per IQ level as given below:

Mild Intellectual Disability: 70-50
Moderate Intellectual Disability: 49-35
Severe Intellectual Disability: 34-20
Profound Intellectual Disability: Below 20

## Psychological Intervention Strategies:

**Psychosocial needs of children with ID:**

Psychologists in a rehabilitation team at-

tend to the adaptation process and developmental needs in children with ID. It is important to identify the strength and the disorder specific issues as a child progresses through developmental stages. Children with ID go through the developmental stresses. The game changing component that aid in less stress on the child but thrown to the parents and siblings is the presence of intellectual disability. Long standing disability may interfere with normal developmental tasks and skill acquisitions due to boredom or unhappy with the fact there is no complete cure. The cause of permanent or temporary discontinuation of services may lead to a major impact on functioning devoid of the intelligence.

**Needs through the developmental stages:**

**Preschool:** Less demanding and more ignorable delay. There is a high motivation of parents probing for recovery rather than accepting the disorder. The parents go through various emotional stages from denial to depression. Family members and society may prove to be a hurdle or a great source of information that can be a boon to the family at large. Child is in the golden stage of improvement. Appropriate coping strategies for family and proper guidance for the child are key to success.

**Entry to school:** School entry is often a crisis point because it may highlight differences with peers. From gaining admission, to executing the daily needs of the child and proving excellence require a lot of support system from family to rehabilitation professionals. Psychological wellbeing of the child should not be the only stand point, rather the entire family require time to time advice to being over pampering a child leading to a child with behaviour issues to being tough on the child developing a low self esteem. The right decision to where the child should be going and the requirement of the child and the support system is best explained by a psychologist expertise in the field of rehabilitation.

Adolescence: Adolescence is the next difficult phase posing challenges in way of their ability to resolve issues of independence from parents, form strong connections with the peer group, dealing with the ongoing changes in the body, as well as master romantic and sexual issues. Demand is higher than mere acceptance. Manipulation and gaining sympathy could be a lifelong strategy for some and others would like to compete with peers. The needs are as normal as it could get minus the presence of disability. Personality development and other disability needs to be attended with caution. Peer interaction attendance is only when demanded as overprotection is no solution.

## Therapeutic Interventions:

Intervention strategies are set based on the assessment and most suitable intervention strategy the child could respond.

### 1. Neuro-cognitive therapy:

This is a new approach to treating individuals with ID. It is based upon two proven principles:

Neural Plasticity: The brain is capable of altering its own structure and functioning to meet the demands of any particular environment. Consequently if the child is provided with an appropriate neurological

environment, he will have the best chance of making progress.

Learning: Lev Vygotsky proposed that children's learning is a social activity, which is achieved by interaction with more skilled members of society. Counseling and behavior therapy, for emotional and psychological challenges may be needed at any age, but is often most crucial during adolescence.

## 2. Cognitive Rehabilitation Therapy (CRT):

Cognitive rehabilitation is a program of guided therapy to learn (or relearn) ways to concentrate, remember and solve problems after an illness or injury affecting the brain. It is a structured set of therapeutic activities designed to retrain an individual's ability to think, use judgment and make decisions.

The focus is on improving deficits in memory, attention, perception, learning, planning and judgment. There are many intervention strategies and techniques used to help children reduce, manage or cope with cognitive deficits. The desired outcome of cognitive rehabilitation is an improved quality of life or an improved ability to function in home and community life.

Cognitive rehabilitation techniques are restorative training or re-training the brain in normal function or compensatory ways. This technique teaches the child to make use of tools to help make up for the deficits.

The goal of cognitive rehabilitation is to improve cognitive abilities in order to obtain as much independent functioning as possible. Some of the specific benefits of cognitive rehabilitation include improvements in the following deficits:

- Attention span
- Memory
- Problem -solving
- Visual – spatial relations
- Learning functional tasks such as feeding, dressing, etc.

Cognitive rehabilitation may be performed in conjunction with or as part of other therapies, such as behaviour, occupational or speech therapy.

Cognitive rehabilitation may be performed by a variety of professionals, such as neuropsychologists or psychologists. The nature of the condition and the specific cognitive deficits determines what kind of specialist would be needed.

There are two treatment approaches to cognitive rehabilitation:

i) Restorative approach: This is based on the theory that repetitive exercise can restore lost or underdeveloped functions. Some methods and techniques include:
- Auditory, visual and verbal stimulation and practice
- Number manipulation
- Computer - assisted stimulation and practice
- Behavior modification

ii) Compensatory approach: This method strives to develop external assistance for cognitive deficits. Compensatory cognitive rehabilitation may use the following techniques:
- Employing visual cues, written instructions, memory notebooks, watches, beepers, computers and other electronic devices to trigger functional behaviors.
- Simplifying complex tasks, capturing the patient's attention, minimizing dis-

tractions and teaching self-monitoring procedures.

The compensatory approach to cognitive rehabilitation has been more widely accepted than the restorative approach, but these approaches are not mutually exclusive. Many therapeutic programs employ element of both.

- Make use of visual cues like flashcards to teach as this would help the child have clear concept formation.
- Due to physical limitations many children have difficulty writing and producing good, legible handwriting and repeated practice would only frustrate your child. Try and make writing enjoyable by making use of paints, crayons, sand, etc. You can also make use to computers, magical pens and white boards to improve the hand writing and make the activity more interesting.
- There is wide range of educational audio visual CD's available in the market which can be used to teach the child concepts like colors, shapes, fruits, body parts, daily activities of living, etc.
- Modify the homework and class work assignments for the child, according to his or her functioning and the possible barriers which could restrict his development.

## 3. Behaviour therapy:

This therapy is often used to enhance child's ability and discourage destructive behaviors.

A few behavior therapy strategies for children with ID that can be practiced at home by parents are mentioned below:

1. First develop a consistent behavioral management plan to use with your child when appropriate or inappropriate behaviors are exhibited. This could mean allowing the child to play with his favorite toy when he displays desired behavior or giving a time–out when the child displays inappropriate behavior.

2. Use a reward system to provide motivation to your child for displaying good behaviors and tasks completed. Set up a clear system of rewards like sticker charts, gold stars, non-edible treat such as toys so that the child knows what to expect when they complete a task or refine their behavior.

3. Praise your child for good behaviors rather than only giving time-out or shouting, when he/she displays bad behaviors. This will help your child develop feelings of self-worth and will build up on his/her confidence level.

4. The child should be taught constructive and acceptable ways to vent out physical or verbal aggression. For example: either drawing, painting, playing a musical instrument like drums, practicing breathing exercise when he or she is angry or frustrated.

5. Many children with ID become over stimulated in a crowded, noisy environment, which can lead to meltdowns. Learn to recognize what external triggers lead to undesirable behavior in your child, and remove her from those situations. You can help the child by removing the child from the over–stimulated environment or teach the child to calm him or herself down in such situations.

6. As children with ID have limited exposure to socializing due to their physical condition they need to be taught skills

to socialize. Techniques such as modeling, role playing and skills training can be conducted with the child as this will give the child a clear ideas as to what constitutes in socially adaptive behavior.

7. Re-evaluate your family's communication methods if your child seems to be unusually frustrated by the inability to express him/ her. Perhaps purchasing a communication board that allows the child to type out his thoughts or point to words can help.
8. Get your child into therapy with a behavioral specialist if you have tried other options and problem behaviors still exist. A professional therapist can help your child sort through his feelings, become better at communicating his wishes and feel better about him/ her.

## 4. Counselling:

As they get older, children with ID will likely feel disliked by their peers, isolated from friendships, embarrassed by body image and/or frustrated with treatment goals. This may be a good time to introduce them to "counseling" or "talk therapy," where they can talk about the things that are bothering them and learn to put them in perspective. It differs from behavioral therapy in that the goals are more along the lines of learning to accept and embrace one's individuality rather than of raw behavior modification.

## 5. Group Therapy:

Group psychotherapy is a process of psychological influence by verbal and non-verbal techniques in which the psychologist uses the interactions created in specially organized small groups of patients (mainly interactions of emotional character), in order to reduce the patients' body and nervous discomfort and to improve psycho-social functioning development.

## Caring for care givers:

The importance of care giving cannot be more succinctly expressed than by the following quote by lady Rosalynn Carter, "There are four kinds of people in this world: Those who have been caregivers, those who currently are caregivers, those who will be caregivers, and those who will need caregivers." This quote hints at the pervasive nature of being a caregiver, knowing a caregiver, or needing a caregiver.

The family is considered to be a system of continuous process with different behavioral models. Not only does ID alter the emotional dynamics between your child and you, but it also stresses the emotional dialogues between you and your family. We believe parents and the family members have to be emotionally prepared to accept the diagnosis and to carry their child through this "special" journey. You must understand and see the "special" needs of their child. Devoted support from caregivers (you and the family members) is crucial for an improved quality of life and attempting to normalize this "given" situation.

## Strategies for parents to cope with the situation:

i) Ask for help if it's difficult caring for your child. For example: Hiring a maid makes more sense than wasting time into household work, rather you could invest that time into teaching your

child or may be time for yourself like exercising.

ii) Make use of the school hours for meeting your relatives, family members or friends while the child is away. Plan quality time together with your spouse for example; lunch with him when your child is at the school.

iii) Do not feel guilty about the break and try to remind yourself that it will help you to be renewed for the things you need to do when you get back. Try and schedule your time in such a way that you are left with sometime for yourself. If possible, take time off and go for shopping or to the parlor, visit a friend, etc.

iv) It is very important that you talk about your feelings and emotions that you are going through. It is important to vent out your thoughts at regular interval of time. Especially someone who just listens could be a great source of strength.

v) If you feel that you are unable to cope up with things around and your stress or anxiety levels have increased or you are starting to show depressive symptoms (like crying spells, change in sleep or appetite, loss of interest, etc) seek help of a professional. Counseling session for you or your spouse or the entire family may help.

vi) You could join support groups which are for children with autism and their parents. It may be helpful to listen or talk to people who have been or are going through a similar situation. Also, support groups could be a great source of information about the services available in your area or about the new possible treatment options available.

The principles of the family therapy are used during consultations with patients. The work is done with trained family therapists. The family will be informed about the confidentiality of the process.

## 6. Expressive therapies:

These are usually used with people who have difficulty verbalizing their feelings such as art, music, poetry, etc which could help freeing and empowering oneself. Sometimes children with ID can become violent or aggressive, resorting to things such as biting or hair pulling to they can be helped to release their aggression and frustrations, by either being vocal or, if the child is able to control his or her hand enough, perhaps drawing or writing.

It is important to acknowledge that psychological assessment and treatment address the needs of children and families, their abilities to participate in the rehabilitation process and their abilities to cope with stress depending on the process of their adjustment to a disability. When providing psychological care, it should not be assumed that a child with disabilities will be a marker for psychopathology. Families with children with special needs may function in unique ways without being pathological. The goal of psychological care is to optimize child and family adaptation to physical and psychological problems of everyday living in the context of good psychological health and good quality, to enhance the capacity of families as well as to build their confidence and resilience. Such care must be available throughout the whole rehabilitation process in the rehabilitation institution as well as in the broader environment.

## Occupational Therapy Manage-

## ment

Occupational therapists help people "engage in their daily tasks or occupations that they want and need to do in a manner that supports health and participation" (AOTA,2008, p. 626). People with developmental disabilities often need support with occupations, assistive technology and home modifications (Hammel et al., 2002), and community participation.

## Assessment:

In occupational therapy, the assessment process provides the baseline for intervention and often focuses on identifying and defining an individual's priorities and the factors that enable or limit his/her occupational performance.

The assessment process always is ongoing, and should preferably take place in the person's natural environment (e.g. home, school, and workplace). Rapport building is an important part of the process, and involves gathering of information from family members and other professionals. In addition to that, the evaluations may be based on information out of observation, questionnaires, and appropriate standardized and non standardised assessments. Observation, however plays a major part in, and is sometimes the only way in which the needed information is obtained. This process is based upon observing actual performance in different environments. As the occupational therapist observes the person's performance and abilities, the therapist attends primarily to aspects of performance skills, such as motor skills (position, stability or mobility), process skills (attention, initiative, choice or organization in time and space), and communications skills (establishing eye contact, cooperation or methods of expression that are utilized).

## Scales:

- **The Canadian Occupational Performance Measure** (COPM):- a standardized Tool designed to be used with clients with a variety of disabilities and across all developmental stages. It is used as an outcome measure.
- **The Jacobs Inventory of Functional Skills** (JIFS): An assessment tool and treatment guide designed for the Developmentally Delayed population of older children and adults consisting of a fairly comprehensive Life Skills Section that covers Self-Help, Home Management, Work Readiness/Cognition, and Community Tasks. The Foundation Skills section covering: Sensory Integration, Neuromotor Status, Fine Motor Skills and Oral Motor Skills.
- **The Sensory Integration Inventory – Revised, for Individuals with Developmental Disabilities.** It's a screening Tool for people who have Sensory processing issures. Describes behaviors and responses to everyday activities which may be indicators of Sensory Dysfunction.
- **The Adolescent/Adult Sensory Profile:** It's a standard method for professionals and individuals to measure the effect of sensory processing on functional performance. Appropriate for individuals aged from 11-65+.

## Interventions:

People with a disability have the right to receive services in a manner, which results in the least reasonable restriction of their rights and opportunities. Occupational therapists' need to focus on the welfare and rights of the users of their services and

interventions should be practiced so as to be as free as possible from restrictive treatment practices.

Similar to the assessment process, occupational therapy intervention for people with intellectual disability is also an ongoing process that is both gradual and dynamic. Treatment is provided throughout life in accordance with the person's changing needs, requirements and likings in all areas of occupation.

Intervention often requires repetitions and practice to achieve internalization and learning, and performance in a variety of contexts to enable generalization. As is the case with respect to assessment, the intervention is preferably carried out in the person's various daily environments. This enables and encourages the person's active participation in the many contexts of his/her life.

Occupational therapy interventions for people with intellectual disabilities are specifically adapted to the person with respect to the degree and type of support needed as well as the context. Interventions may include direct treatment as well as adaptation or modification in environment, guidance to teachers and caregivers, monitoring and counselling (including of the family, the educational and clinical staff, employers and others).

An occupational therapist will work in the following main areas:-

- **Activities of daily living:**

It includes activities directed to the person's care of his/her personal needs (ADL) such as, eating, dressing, toileting and maintaining hygiene and instrumental activities of daily life (IADL) such as cooking, doing the laundry, shopping, or managing finances. This area represents a central focus of intervention in occupational therapy. For example, with respect to activities related to dressing, the intervention can range from adapting the clothing, choosing preferred clothing, to teaching more advanced skills such as organizing cupboards, shopping for clothes, and washing clothes.

- **Learning/Studies:**

It is essential part to be a student and to participate in a learning place which includes academic and non-academic activities. Intervention here covers a variety of educational settings like day care centers for young children, kindergartens, preschools and special education schools (ages 3-21 years), regular school and professional training units. The intervention plan is varied and may focus on gaining basic to complex learning-skills, such as understanding cause and effect processes and object permanence, or on more complicated skills, such as preparation for learning and writing, organization in time, in space and with accessories, adaptation to different learning environments, the use of information technologies and computers and gaining learning strategies. In addition, the intervention can include adapting various learning environments.

- **Work:**

These are productive vocational activities, whether for compensation or not, that include preparing for work, producing a product and providing services. People with intellectual disabilities may have difficulty in pursing regular vocations as a result of an continuous need for ongoing support and limited understanding of how they express choice and engagement in occupation.2

Intervention in this area will include different work areas like: pre- vocational and vocational training centres in which students receive training before entering enter work forces, youth rehabilitation centers, adult sheltered-work facilities, and other supportive community work systems, and placement-services. Intervention varies depending upon the capabilities and strengths of an individual and may include basic work skills training (behavior norms, work routines), developing and practicing basic cognitive abilities, practicing motor skills, exposure to varied work opportunities and situations, support and advice for exploration and developing areas of interest, identifying abilities and choosing suitable occupations, analyzing occupations and adapting them as needed, as well as supporting and assisting placement in various work sights in the community.

- **Play:**

These are activities that are generally internally motivated and provide pleasure, entertainment and learning. Play-intervention, as an occupational therapy goal in this population, is directed towards the most basic experiencing of play as a source of pleasure, as well as providing the person with an opportunity to participate in play activities. The intervention includes training in basic skills such as the use of equipment, recognizing rules and agreed-upon behavior patterns, role playing or choosing suitable play activities. In addition, play helps as an intervention method for learning and practicing a variety of social, motor and functional skills. For adults, physical activities/sports should be included in occupational therapy programs as it provides considerable functional benefits and improvement in the quality of life for intellectually disabled people.

- **Leisure:**

These are non-obligatory activities that are internally motivated and are performed at times that are not devoted to work, studies, self-care or sleep. Research reveals that people within this population have a relatively large amount of time to devote to leisure, whereas their participation in leisure activities is minimal (Buttimer & Teirney, 2005). Therefore, coping with leisure within this population is a central topic. Intervention in this area may focus on exploration of varied leisure opportunities, identification and choice of areas of interest, planning leisure time and participation in activities that lead to a perception of capability, pleasure, control and satisfaction.. Musical games and specific body expressing activities in the therapeutic program for developing self-esteem, self-confidence, applying personalized intervention programs to mentally disabled children, encouraging their permanent appreciation too.3

- **Social participation:**

These are activities related to agreed-upon behaviour patterns expected of an individual within a given social system (e.g. community, family or with friends). The intervention within occupational therapy encourages the person to gain skills in the various areas of occupation and thus supports and strengthens social participation. For this population, an emphasis is places upon understanding acceptable social norms and as well as learning and practicing activities that lead to satisfactory social interactions. Art, crafts, drama, dance, music and games can be used to improve self-image.

## • Accessibility and Environmental Modification:

Occupational therapy practice relates to the person, the occupation and the environment. The occupational therapist's broad knowledge base in the areas of function and limitation enables him/her to identify, through performance analysis in the different areas of occupation, environments and/or tasks that should be modified. The various limitations that characterize the population of people with intellectual disabilities require both general and person-specific environmental modifications to ensure accessibility. The characteristic difficulty in problem-solving, initiative and coping with unfamiliar situations, amplifies the need for accessibility modifications for this population. These accessibility modifications include changes in the environment (as in widening passageways, modifying playgrounds or adding symbol signs), in the equipment (such as adapting seating systems of adapting feeding aids), or of the task (such as changing the complexity of instructions or dividing a task into sub-stages).

## • Assistive technology :

It is one of the methods used to adapt the environment and includes modifications of hardware, software and various combinations thereof (such as a virtual keyboard, a touch screen, a motorized wheelchair, switch systems, computer programs and internet sites, adapted content amount, or voice output devices). Thus, for example, a switch can be modified to be activated through the person's head or hand. Other modifications of the switch may include size, color, texture, or sensitivity (such as speed or pressure- response). Assistive technology promotes a variety of functions related to the individual, the occupation and the environment. In addition, it allows for the modification of an individual's environment in the manner in which his/her requires, by relating to his personal abilities, wants, areas of interest and specific limitations and difficulties.

Environmental modification is likely to significantly improve a person's ability to participate in all areas of occupation, his or her level of independence and the degree of supports required.

In summary, the occupational therapist, as part of a therapeutic, rehabilitative and educational profession play a central role within the support system available to people with intellectual and developmental disabilities, throughout the life cycle. As such, occupational therapists hold key positions as leaders in this area. Working with people with intellectual and developmental disabilities requires consideration of function, independence and participation in the various areas of occupation, which enables the occupational therapist to utilize all the areas of knowledge and expertise included in the practice of occupational therapy.

Intellectual disability is a condition diagnosed in early developmental ages, responsible for impaired learning abilities, dependent in functional activity, poor gross motor skills and reduced adaptive behaviour. This is highly influenced by their sensory system function and their ability to interpret it. Normally, the various senses work together in unison which helps us to navigate in our surroundings. However, some children with intellectual disabilities have difficulty interpreting, integrating, and coordinating the sensory input.Some children have hyperactive behaviour (underactive system) where they seek more and more movement, enjoys being on swing and are restless whereas few

children have overactive system resulting into gravitational insecurity and motion sickness due to which children may seek assistance with seemingly simple tasks like walking or want to be carried instead of walking. They may also resist activities like climbing or descending stairs.

The above mentioned sensory issues can be treated with sensory integration techniques or therapy. Sensory integration activities address children's sensory needs by either lessening or amplifying the intensity of the sensory stimulation they receive. .Most sensory integration activities work with children's vestibular, proprioceptive, and tactile sensory systems. The vestibular sensory system enables us to stand and coordinate movement; it also involves input from vision and from vestibular centre in middle ear.

## Physiotherapy

A physical therapist works with children in case of underactive /overactive sensory system by engaging them in structured movement exercises which meets their sensory needs within socially acceptable boundaries.Similarly, proprioceptive sensory system provides information about the body's positioning and provide them sense of their presence in environment (helps in developing self awareness) and is also beneficial to develop their motor planning. Motor skills are large movements of the body such as standing, walking, running and jumping, which are typically attained before the pre-primary schooling which is before 6 years of age. Deficits in this gross motor skills may occur due to certain congenital conditions such as down's syndrome or cerebral palsy which is secondary to their intellectual disability along with altered neuromuscular co-ordination and tone. Levels of physical activity may be decreased in children with ID due to reasons such as difficulty moving freely without the support carer input or facilitative inputThis is responsible for gross motor skill deficits for which physical therapy interventions play important role.

Physical therapy may be conducted in schools, homes, or institutions. Sometimes children go to specialized physical therapy facilities. Physical therapy increases flexibility and stimulates learning abilities through sensory integration activities. Physiotherapy can assist people of various ages to attain and improve their gross motor skills and also be able to regularly practice these skills in familiar contexts. Such practice enhances the integration of the skills into daily life and also makes it more likely that they will be successfully used in less familiar settings when required.

Physiotherapy interventions designed to improve GMS deficits may focus on the ICF domains of activity (such as task-specific training interventions for example gait training) or on processes categorized within the domain of body structure and function (such as muscle strengthening, or balance exercises.For physiotherapy interventions that aim to improve levels of physical activity, these same ICF domains may be considered as well as the ICF domain of participation (for example, involvement in team sports).

Motor skill and learning performance depend on healthy connections between different parts of the brain that link the following sensory information from the body through skin, joints, muscles, vestibular system, and also through sensory information from the environment using vision and hearing, intentions, goals and feelings. Regular exercising can improve motor skills in children with ID.Not all children with intellectual disabilities/ require physical therapy.

The underlying condition that caused the intellectual disability determines whether physical therapy is needed. For example, the Fragile X syndrome is often accompanied by sensory challenge. People with Fragile X are easily distracted by noises or smells. People with Cockayne syndrome may need physical therapy to limit joint contractures that interfere with walking. People with Prader-Willie syndrome have poor muscle tone; physical therapy helps to correct this.

Physiotherapy interventions are targeted at improving specific physical impairments (such as weakness) or activity limitations (such as decreased balance or impaired gait). Examples of types of physiotherapy interventions for specific physical issues include improving muscle strength and function through exercise and rehabilitation (including strengtheningand stretching exercisesand aquatic therapy),training of specific Gross motor skillsincluding gait education,and improving a client's balance strategieswhich may help to decrease the risk of falls. Exercises may also be prescribed to aid reduction in pain,or recovery of physical function following surgery.

**Below are few exercises which can be done as therapeutic exercises:**







**STRENGTHENING EXERCISES**



**WEIGHT TRAINING**

# Ch. 18 Neuropathies

**Dr. Joji Joseph, Dr. Shruti Shirke, Dr. Ragini Sharma, Dr. Reena Jain**

"Neuropathy" is a broad, generic term in which one or more nerves are affected. Peripheral neuropathies are caused by damage to the structure and there by function of peripheral motor, sensory and autonomic neurons. This clinically appears as either a mononeuropathy, mononeuritis multiplex (multiple non contiguous nerves) or a polyneuropathy.

## 1. Mononeuropathy:

It refers to involvement of single major nerves, which is usually caused by trauma or compression. Diagnosis will be based on the bedside neurological examination, sensory and motor deficit as per the nerve course, further confirmed by electromyography (EMG).

## 2. Polyneuropathy:

This is the disorder of multiple nerves, involving both major and small, unnamed nerves, or branches. Mostly there will be symmetrical involvement of multiple peripheral nerves with sensory symptoms (numbness, tingling paresthesias, sensory ataxia) or distal weakness or both. Often all types of axons (motor, sensory, autonomic) are involved. Usually distal nerves are more involved than the proximal. since their metabolic maintenance and axoplasmic flow are more susceptible to neurotoxic factors.

## Pathology:

Broadly polyneuropathies can be divided into axonal (80-90%) and demylinating (10-20%) with or without conduction block.

## 1. Demyelinating polyneuropathies

It is chracterised by a mononeuropathy resulting from localized compression,for example carpal tunnel syndrome,where the median nerve is compressed at the wrist. In acute polyneuropathy such as Guillain-Barre syndrome demyelination is also the primary process. In either case, if the neuropathy progresses over time or is initially severe, axonal degeneration may also occur, with less chance for recovery. If there is mild focal trauma to a nerve, recovery may occur sooner and more completely once any swelling has resolved or any remyelination of intact axons has occurred.

Demyelinating Polyneuropathy

1. Immune mediated: AIDP, CIDP

2. Infective: Diphtheria

3. Paraprotein associated: IgM MGUS

4. Hereditary: CMT I, III and X, Refsum's disease,

5. Leukodystrophy

## 2. Axonal polyneuropathies

With a more severe crush or penetrating focal nerve injury, axonal loss occurs via Wallerian degeneration. Axons and myelin degenerate distal to the point of nerve injury.Recovery takes longer and may be incomplete, depending on whether any remaining perineurium can "guide" regrowing or sprouting axons to their intended targets. Without such "support scaffolding" of perineurium, resprouting axons

pile up in a bulbous neuroma, which is often painful.(fig.1)

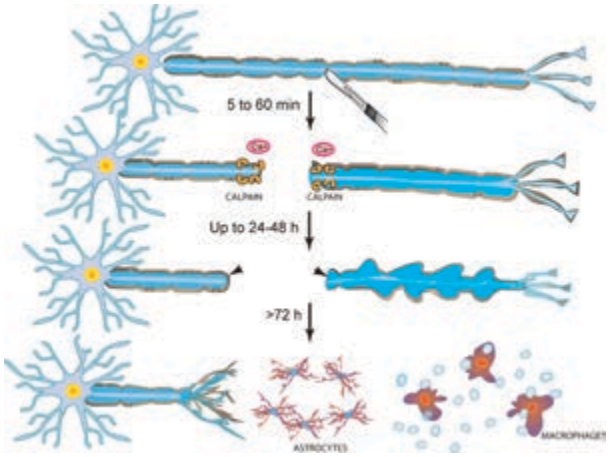



Fig.1 Axonal polyneuropathies

## Causes of Axonal Neuropathies

1. Endocrine: Diabetes, hypothyroidism
2. Metabolic: Chronic renal failure, liver disease, etc.
3. Infections: HIV, HTLV, leprosy, Lyme disease
4. Connective tissue diseases: SLE, rheumatoid arthritis, Sjögren's syndrome.
5. Vitamin deficiency: Vitamin B12, Vitamin E
6. Drugs and toxins:

Alcohol, chemotherapeutic agents: Vincristine, cisplatinum

Phenytoin, statins, antibiotics: Metronidazole, dapsone, etc.

Antiretroviral medications, etc

7. Paraneoplastic
8. Paraproteinemia: Myeloma, Waldenstrom's macro-globulinemia, MGUS, amyloidosis
9. Inherited: Charcot-Marie-Tooth (CMT) type II,porphyria, HSAN

According to time course the polyneuropthies can be divided as acute and chronic polyneuropathies

**A. Acute (days to week):**

1. Acute polyradiculitis
2. Critical illness
3. Polyarteritis nodosa
4. Toxic polyneuropathies
5. Porphyria
6. GBS

**B. Chronic (months to year)**

1. HMSN
2. Diabetes

**C. Relapsing**

Chronic polyradiculitis

**Epidemology**

Prevalence can be as high as 2.4 % in general population2 and 8% in person 55 years and older3

**Symptoms**

*Sensory symptoms*

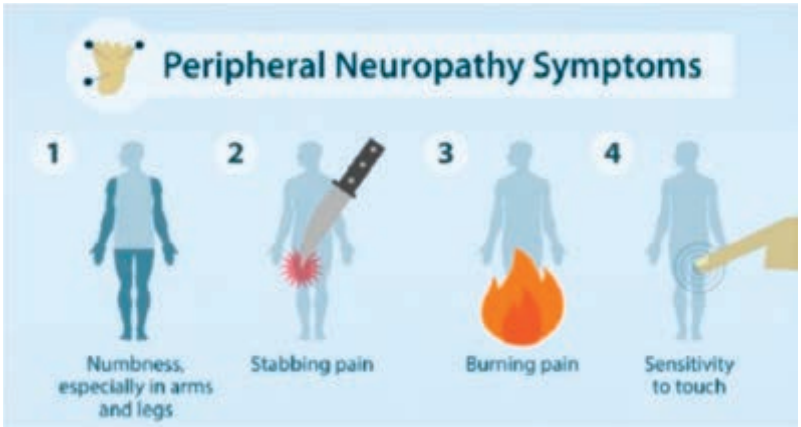



Fig 2. Sensory symptoms of Peripheral Neuropathy

- Tingling, Numbness- usually begins distally in the toes and feet, later affecting the fingers and hands
- Paresthesia, a spontaneous tingling, "pins and needles" sensation
- Dysesthesia, an unpleasant sensation from a non-noxious stimulus
- Burning
- Pricking, stabbing, or shock-like pain
- Muscle cramping
- Sensitivity to touch

*Motor symptoms*

Over the years, nerve damage symptoms may worsen later motor symptoms which includes:

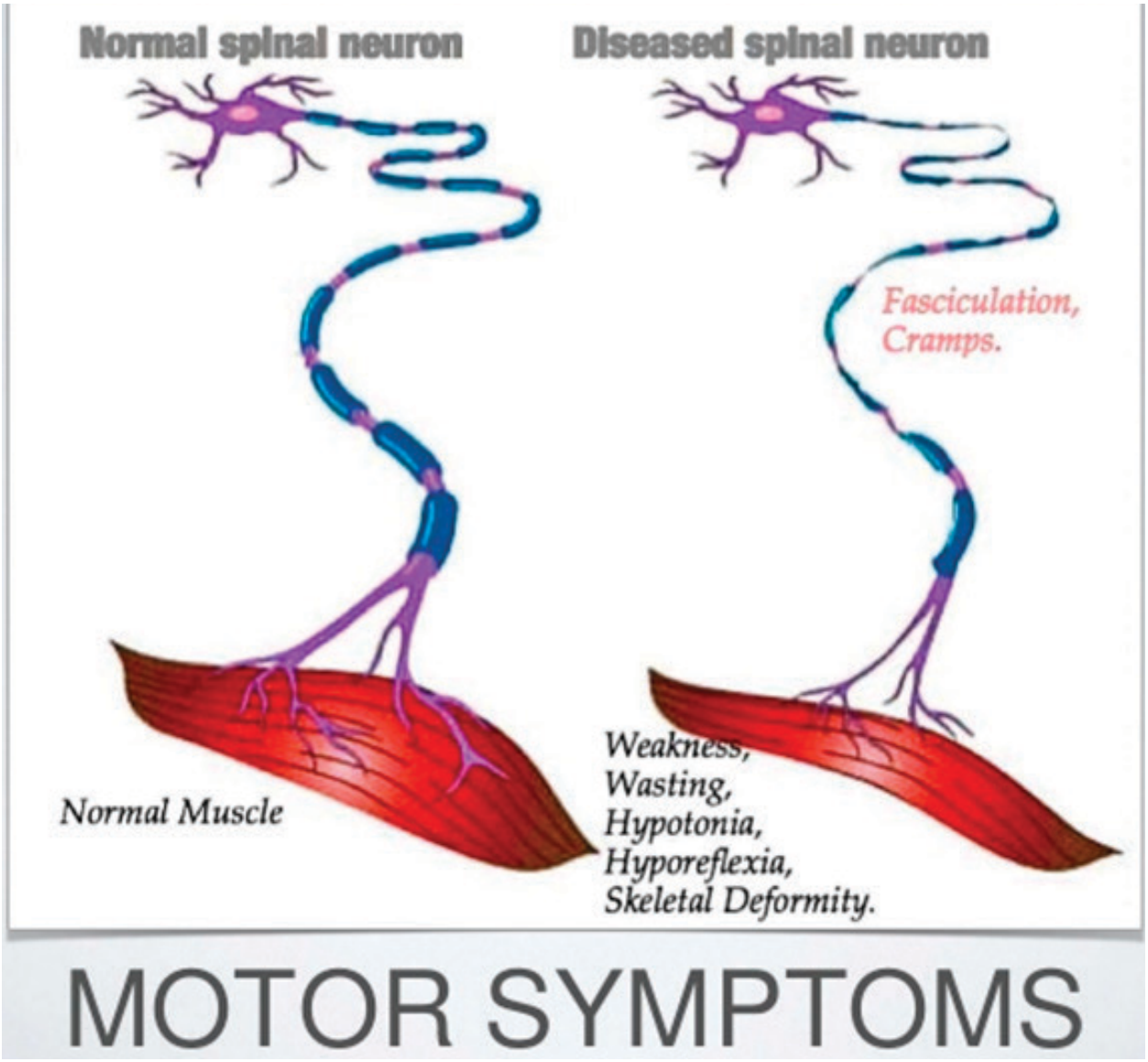



Fig 3. Motor symptoms of Peripheral Neuropathya

- Weakness and muscle atrophy also begins or predominates in the **distal** limbs.
- There is **early loss or decrease of reflexes.**

*Autonomic symptoms*

- Orthostatic hypotension,
- Incontinence
- Sweating abnormalities.




Fig 4. Areas affected in Polyneuropathy

- Digestive problems
- Dizziness
- Balance and walking problems
- Increased risks of ulcers or infections in the feet

## Diagnostic Testing

I. Complete blood count, comprehensive metabolic profile, and measurement of erythrocyte sedimentation rate and fasting blood glucose, vitamin B12, and thyroid-stimulating hormone levels

II. Cerebrospinal fluid (CSF) analysis to evaluate for chronic inflammatory demyelinating neuropathy antisulfatide antibodies to evaluate for auto-immune polyneuropathy; and genetic testing if hereditary peripheral neuropathy is suspected

III. Lumbar puncture and CSF analysis may be helpful in diagnosing Guillain-Barré syn-drome and chronic inflammatory demyelinating neuropathy; CSF protein levels may be elevated in patients with these conditions.

## Electro diagnostic studies

Electro diagnostic studies are recommended if the diagnosis remains unclear after initial diagnostic testing and a careful history and physical examination. Electro diagnostic studies can help determine whether the neuropathy is the result of damage to the axons (axonal neuropathy) or the myelin (demyelinating neuropathy), or both (mixed).

There are two primary types of electro diagnostic studies

1. Nerve conduction studies: Nerve conduction studies assess the shape, amplitude, latency, and conduction velocity of an electrical signal conducted over the tested nerve.

<u>Axonal Neuropathy</u>

Reduced motor and sensory amplitude

Conduction velocity normal or slightly reduced (Median motor >40 m/s)

Distal Latency normal or slightly prolonged

No decay

Demyelinating Polyneuropathy

Conduction velocity reduced >30% (Median nerve <40m/s)

Distal latency > 7ms

Normal or reduced amplitudes

**2.** *Electromyography (EMG).* EMG can detect active axonal damage, as evidenced by the presence of spontaneous muscle fiber activity at rest resulting from the absence of neuroregulation (denervation). The motor unit action potential on voluntary muscle contraction also is assessed.

Normal nerve conduction studies and needle EMG significantly decrease the likelihood of peripheral neuropathy, whereas abnormal nerve conduction findings confirm the diagnosis.

**3.** *Nerve biopsy:* Nerve biopsy should be considered when the diagnosis remains uncertain after laboratory and electrodiagnostic testing, or when confirmation of the diagnosis is needed before initiating aggressive treatment (e.g.,in cases of vasculitis when steroids or chemotherapy is used). Sural and superficial peroneal nerves are preferred for biopsy.Epidermal skin biopsy can be performed in patients with burning,numbness and pain and in whom small unmyelinated nervve fibres are suspected to be the cause.

## Prognosis

Complete recovery from polyneuropathy can occur if a correct cause is found out and treated and if the damage does not affect the entire nerve cell.

The amount of disability varies. Some people have no disability. Others have partial or complete loss of movement, function, or feeling. Nerve pain may be uncomfortable and may last for a long time.

Occasionally sensorimotor polyneuropathy causes severe, life-threatening symptoms

## Guillian Barre Syndrome (GBS)

Guillian Barre Syndrome (GBS) is, an autoimmune disease affecting the peripheral nervous system, an acute inflammatory demyelinating poly- radiculoneuropathy (AIDP).The French neurologists Georges Guillian, Jean- Alexandre Barre and Andre´ Strohl reported the combination of increased protein concentration with a normal cell count in the CSF, or albumin-cytological dissociation, which differentiated the condition from polymyelitis. Hence, it became known as Gullian- Barre syndrome. The hallmark for clinical diagnosis of GBS is the combination of rapidly progressive symmetrical weakness in the extremities with or without sensory involvement, areflexia or hyporeflexia, in the absence of a CSF cellular reaction as there are no specific diagnostic tests.

*Epidemiology*

The world-wide incidence of GBS is 0.6–4 cases per 100,000 populations. The incidence increases with age from 1 per 100 000 in those aged below 30 years, to about 4 cases per 100000 in those older than 75 years. GBS incidence increases by 20% per 10-year increase in age.The highest rates have been reported in adults, especially those aged over 75 years. GBS affects men 1.5 times more commonly than women.

*Etiology and Pathophysiology*

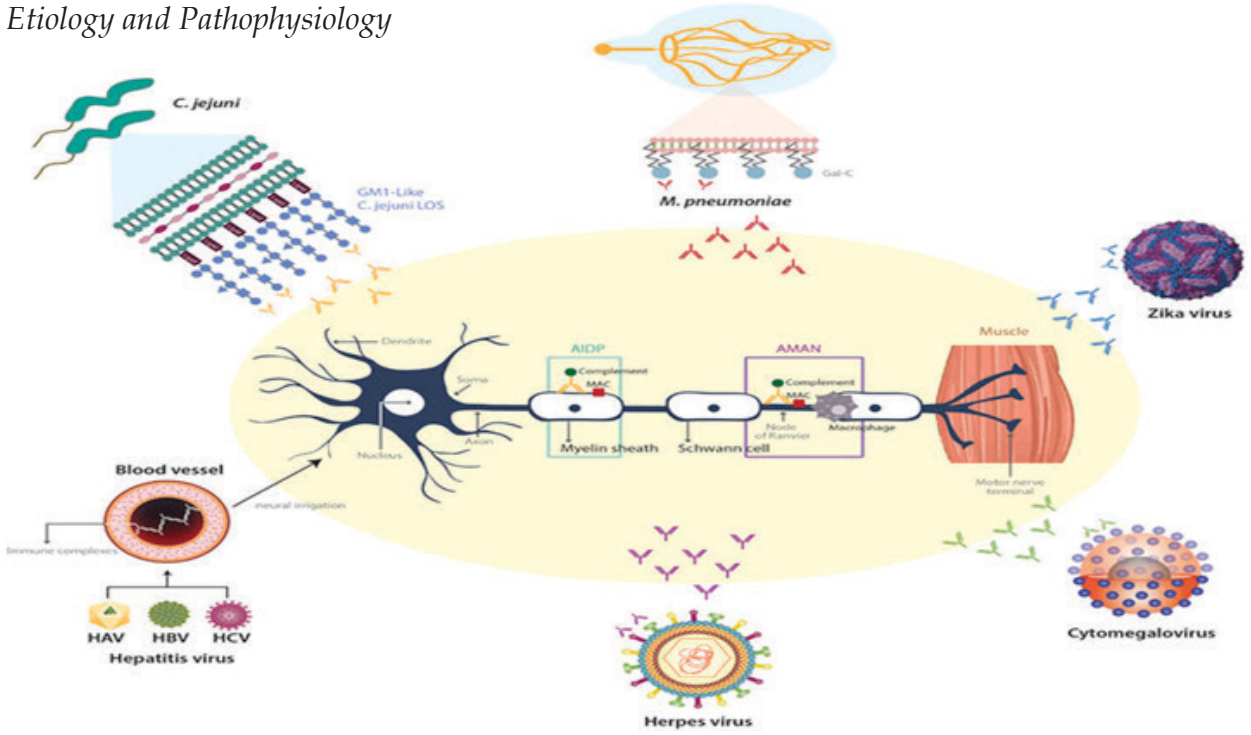



Fig 5: Etiology and Pathophysiology

It is a post- infectious disease and immune- mediated disease affecting peripheral nerves.

Bacterial or viral illness, respiratory and gastro- intestinal infections, administration of certain vaccinations and other systemic illnesses are reported prior to the onset of neurologic symptoms

*Bacterial or viral infections*

C jejuni

It is the most commonly isolated pathogen in GBS. Patients infected with C jejuni have reported gastrointestinal and upper respiratory tract symptoms. C jejuni infections can also have a subclinical course as patients have not reported infectious symptoms before the development of GBS. Patients who develop GBS following an antecedent C jejuni infection often have a more severe course, with rapid progression and a prolonged, incomplete recovery.

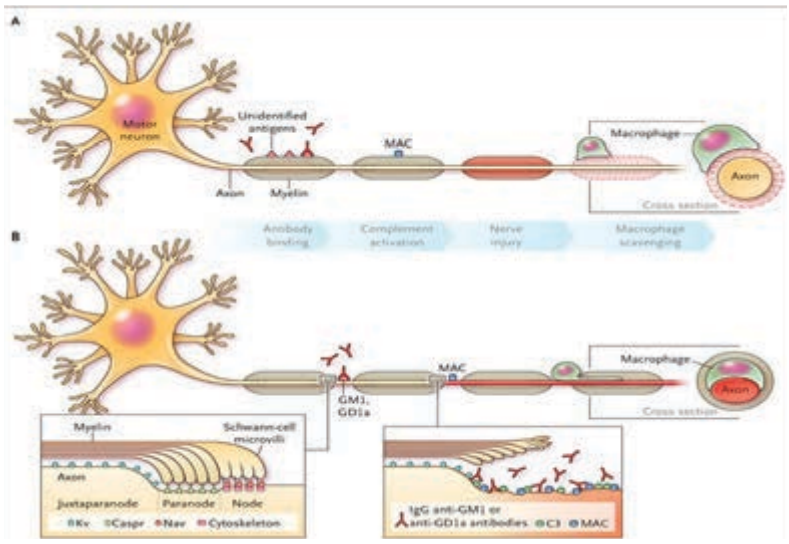



Fig 6: Molecular mimicry

C jejuni infections and the pure motor and axonal forms of GBS have a strong clinical association. The virulence of C jejuni results from the presence of specific antigens in its capsule that are shared with nerves. An antibodies are produced from the immune responses directed against capsular lipopolysaccharides that cross-react with myelin, resulting in immunologic damage to the peripheral nervous system, to cause demyelination. This process has been termed molecular mimicry. [12] Lymphocytic infiltration of spinal roots and peripheral nerves followed by macrophage-mediated, multifocal stripping of myelin are the pathologic findings in GBS. This phenomenon results in defective propagation of electrical nerve impulses with profound delay in conduction or eventual absence that causes flaccid paralysis. Recovery is typically associated with remyelination.

Anti-ganglioside antibodies are also generated by C jejuni infections which includes gangliosides GM1, GD1a, GalNac-GD1a, and GD1b—that are commonly found in patients with AMAN and AMSAN, the axonal subtypes of GBS.

CMV

Cytomegalovirus (CMV) infections are the second most commonly reported infections preceding GBS. It is the most common viral trigger of GBS. Upper respiratory tract infections, nonspecific flulike illnesses, and pneumonias are presented in CMV infections. There is prominent involvement of the sensory and cranial nerves in GBS patients with preceding CMV infections. CMV infections are significantly associated with antibodies against the ganglioside GM2.

Other infections

Other significant infectious agents in GBS patients include Epstein-Barr virus (EBV), Mycoplasma pneumoniae, and varicella-zoster virus. Patients with GBS have demonstrated infections with Haemophilus influenzae, Borrelia burgdorferi, parainfluenza virus type 1, influenza A virus, influenza B virus, adenovirus, and herpes simplex virus. The Zika virus can cause GBS but there is currently no commercially available test for Zika virus .

*Vaccines*

In GBS, vaccinations are linked by temporal association. However, there is no definite causal relation has been established between vaccines and GBS.

*Medications*

Penicillin and antimotility drug use were more frequently and oral contraceptive use were less frequently reported in patients with GBS. Antibiotic therapy with fluoroquinolones also is associated with the development of GBS.

*Other associations*

The possible triggers of GBS are bariatric and other gastric surgeries, renal transplantation, and epidural anesthesia, anecdotal associations include systemic lupus erythematosus, lymphoma, sarcoidosis, and snakebite, trauma, and pregnancy. Many autoimmune and inflammatory diseases are associated with tumor necrosis factor–alpha polymorphisms with increased expression that may increase susceptibility to axonal GBS subtypes.

*Classification of GBS:*

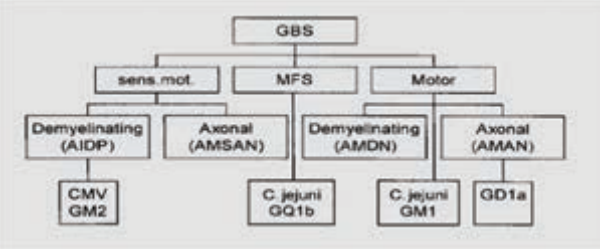



Fig 7: Classification of GBS

GBS can be divided into at least 4 main subtypes of disease: acute inflammatory demyelinating polyradiculoneuropathy (AIDP), the axonal subtypes, i.e. acute motor axonal neuropathy (AMAN) and acute motor and sensory axonal neuropathy (AMSAN), and Miller Fisher

Syndrome.

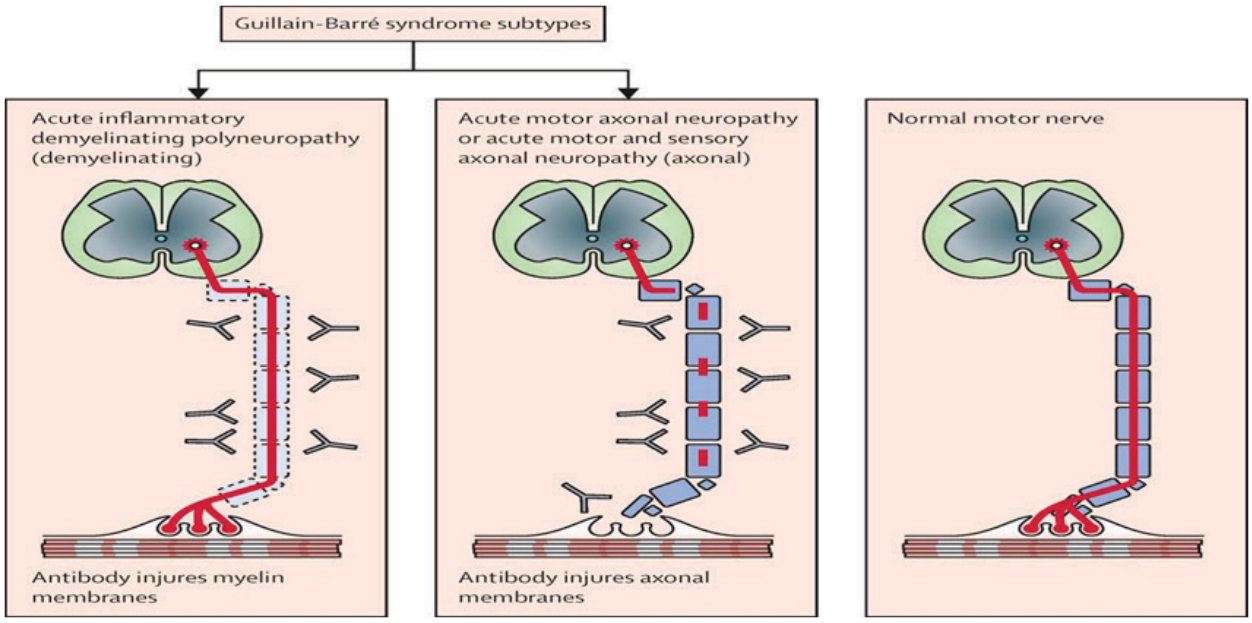



Fig 8: GBS sub types

## Acute inflammatory demyelinating polyneuropathy:

The most commonly identified form in the United States is the acute inflammatory demyelinating polyneuropathy (AIDP) subtype. It is generally preceded by a bacterial or viral infection. The 40 percent of patients with AIDP are seropositive for C jejuni. Macrophage-mediated peripheral nerve demyelination and lymphocytic infiltration is present. Symptoms generally resolve with remyelination.

## Acute motor axonal neuropathy:

The acute motor axonal neuropathy (AMAN) subtype is a purely motor disorder. The prevalence of this subtype is more in pediatric age groups. It is generally characterized by rapidly progressive symmetrical weakness and ensuing respiratory failure.The majority of cases of AMAN are associated with preceding C jejuni diarrhea. The 70-75% of AMAN patients are seropositive for Campylobacter. The disruption of the blood-CNS barrier is due to inflammation of the spinal anterior roots. Wallerian degeneration is showed on biopsies without significant lymphocytic inflammation. The recovery for many patients is rapid while severely disabled patients with AMAN may show improvement over a period of years. On examination, hyper- reflexia is present in one third of patients with AMAN may be due to dysfunction of the inhibitory system via

spinal interneurons that may increase motor neuron excitability although the mechanism for this hyperreflexia is unclear. Hyperreflexia is significantly associated with the presence of anti-GM1 antibodies.

## Acute motor-sensory axonal neuropathy

It is affects sensory nerves and roots. It is also a severe acute illness differing from AMAN and adults are mainly affected. AMSAN presents as rapid with severe motor and sensory dysfunction, marked muscle wasting with poor recovery. It is also associated with preceding C jejuni diarrhea as AMAN. Severe axonal degeneration of motor and sensory nerve fibers with little demyelination is shown in pathological findings.

## Miller-Fisher syndrome

It is observed in about 5% of all cases of GBS. It presents as a triad of ataxia, areflexia, and ophthalmoplegia. The cardinal feature is an acute onset of external ophthalmoplegia. Patients with MFS may present with mild limb weakness, ptosis, facial palsy, or bulbar palsy. Pathological findings show reduced or absent sensory nerve action potentials and absent tibial H reflex. The prominence of anti-GQ1b antibodies are found in MFS. The relationship between anti- GQ1b antibodies and opthalmoplegia may be explained as dense concentrations of GQ1b ganglioside are found in the oculomotor, trochlear, and abducens nerves. Anti-GQ1b/GT1a IgG antibodies are carried by patients with acute oropharyngeal palsy. Within 1-3 months, recovery occurs.

## Acute panautonomic neuropathy

It is the rarest GBS variant which involves the sympathetic and parasympathetic nervous systems. Patients with acute panautonomic neuropathy presents as severe postural hypotension, anhydrosis, bowel and bladder retention, papillary abnormalities and decreased salivation and lacrimation. . Cardiovascular involvement is common, and dysrhythmias are a significant source of mortality. Significant sensory or motor involvement is lacking. Recovery is gradual and often incomplete.

## Pure sensory GBS

It is typified by an areflexia in a symmetrical and widespread pattern and a rapid onset of sensory loss, sensory ataxia. The results from electromyography (EMG) show characteristic signs of a demyelinating process in the peripheral nerves and lumbar puncture studies show albuminocytologic dissociation in the CSF. Patients with severe disease or slow recovery can be given immunotherapies such as plasma exchange and the administration of IVIGs.

## Other variants

The pharyngeal-cervical-brachial (PCB) variant of GBS is distinguished by isolated facial, oropharyngeal, cervical, and upper limb weakness without lower limb involvement. There can be combinations of any of the above subtypes, and any combination of nerve injury. There are mild cases that cause temporary symptoms, improve spontaneously, and never get definitively diagnosed.

## Bickerstaff's brainstem encephalitis (BBE)

Bickerstaff's brainstem encephalitis (BBE) is an uncommon central nervous system (CNS) disease characterized by cerebellar ataxia with disturbances of consciousness & pyramidal signs and acute external opthtalmoplegia. Patients with BBE show

brisk reflexes, extensor plantar responses, drowsiness, hemisensory disturbance, or CSF pleocytosis. The etiology remains unknown but BBE has an autoimmune origin because of an antecedent illness and frequently the presence of anti-GQ1b antibodies.

*Clinical features*

The typical presentation of patient with GBS is having a relatively benign respiratory or gastrointestinal illness with complaints of finger dysesthesias and proximal muscle weakness of the lower extremities in 2- 4 weeks.

Weakness

The classic pattern of weakness is ascending and symmetrical in nature. The lower extremities are usually involved before the upper extremities. Proximal muscles may be involved earlier than the distal muscles. The progression of weakness may be over hours to days that involve the arms, truncal muscles, cranial nerves, and muscles of respiration.

Cranial nerve involvement

Associated common complaints with cranial nerve involvement in GBS are facial droop (may mimic Bell palsy), dysarthria, dysphagia, diplopias, pupillary disturbances, and ophthalmoplegia.

Sensory changes

Paresthesias, numbness, or similar sensory changes are complaints of most GBS patients. Paresthesias usually begin distally (toes and fingertips) progressing upward but generally not extending beyond the wrists or ankles. Deep sensations like vibration, proprioception and superficial sensations like touch, and pain are absent.

Pain

The mechanism of pain is uncertain. Direct nerve injury or the paralysis and prolonged immobilization can result in pain. It is most severe in the shoulder girdle, back, buttocks, and thighs and may occur with even the slightest movements described as throbbing or aching in nature.

Autonomic changes

The autonomic changes in GBS patients are observed as tachycardia, bradycardia, paroxysmal hypertension, facial flushing, orthostatic hypotension, anhidrosis and/or diaphoresis and urinary retention.

Respiratory involvement

Due to respiratory or oropharyngeal weakness, patients with GBS complains of dyspnea on exertion, difficulty swallowing, shortness of breath and slurred speech. Patients with ventilatory failure require respiratory support at some time during the course of their disease.

*Diagnosis*

GBS is generally diagnosed on clinical grounds. There are few diagnostic criterias as follows:

- Biochemical screening such as electrolyte levels, creatine phosphokinase (CPK) level, Liver function tests (LFTs), Erythrocyte sedimentation rate (ESR).
- Needle EMG and nerve conduction studies
- Cerebrospinal fluid studies
- Pulmonary function tests

*Medical Management*

- Intensive care unit

Patients with a forced vital capacity of less than 20 mL/kg, dysautonomia, or severe bulbar palsy are considered to be kept in

ICU. Intensive care includes respiratory therapy, cardiac monitoring, safe nutritional supplementation and monitoring for infectious complications (eg, pneumonia, urinary tract infections, septicemia). Prophylaxis of for prevention of deep vein thrombosis (DVT).

- Immunomodulation

Intravenous immunoglobulin (IVIG) and plasma exchange

- Prevention of infection
- Prevention of thromboses, pressure sores, and contractures

*Complications:*

- Breathing difficulties due to respiratory muscle weakness
- Residual numbness or other sensations.
- Abnormal and irregular Heart rhythms and blood pressure problems
- Pain managed by medications
- Sluggish bowel function and urine retention may result from Guillain-Barre syndrome.
- Deep vein thrombosis
- Pressure sores
- Contractures and deformities

## Hereditary Motor Sensory Neuropathy

Hereditary Motor sensory neuropathy (HMSN) or Charcot-Marie-Tooth (CMT) or peroneal muscular atrophy is a group of inherited, slowly progressive disorders affecting motor and sensory peripheral nerves resulting in distal muscle weakness, muscle atrophy, sensory loss with demyelination, axonal degeneration, or both.This is a large group of disorders in which the onset of symptoms is insidious and progression is very slow over years or decades. It has an autosomal dominant pattern of inheritance. It is divided in following groups.

1. CMT 1 (dominantly inherited hypertrophic demyelinating neuropathies) also called as HMSN types 1A and 1B. it is primarily demyelinating neuropathy noted in nearly half of all CMT cases.

2. CMT2 also called as HMSN type 2 (dominantly inherited neuronal neuropathies) primarily results in axonal involvement and occurs in about one fifth of all CMT cases.

3. CMTX = X-linked, most with both demyelinating and axonal features. CMTX1, due to GJB1 (gap junction B1, formerly known as connexin 32) alterations, comprises 10-12% of all CMT cases and is by far the most common in this group.

4. Dejerine-Sottas variety or HMSN type 3 (hypertrophic neuropathy of infancy results into nerve hypertrophy;also known as CMT3 for axonal autosomal recessive diseases.

5. Refsum or HMSN type 4 (hypertrophic neuropathy) its associated with phytanic acid excess) autosomal recessive, typically more severe, early onset, may be demyelinating, intermediate, or axonal

*Incidence and prevalence*

Overall prevalence of HSMN is approximately 1/2500 and incidence is 15/100,000 in the general population.

*Risk factor*

Family history for autosomal dominant or X-linked forms, and consanguinity for autosomal recessive ones. Primary prevention is genetic counseling.

*Mortality/Morbidity*

Most people with CMT have a normal life

expectancy, the exceptions being patients with respiratory involvement or severe disability. disability varies greatly both between and within families, and can range from asymptomatic with minimal examination findings to severe disability

*Race*

No racial predilection is recognized in the common forms of CMT. Rare, recessively inherited forms (CMT4) cluster in particular ethnic groups.

*Sex*

As expected, CMTX affects males earlier, more frequently, and more severely than females. Other forms of CMT do not show predilection for either sex.

*Age*

Symptoms of Charcot-Marie-Tooth disease typically appear in adolescence or early adulthood, but may also develop in midlife. Some patients experience very slow progression of mild, lifelong symptoms, and may seek medical attention only late in life.
There are patients and families in which CMT has a late onset, as late as the fifth or sixth decade in some mutations, and even as late as the seventh decade in CMT2.

*Symptoms*

In cases of early onset severe forms patients may have infant hypotonia with delayed motor milestones, toe walking and even arthrogryposis; most present before age 20, with clumsiness and abnormal gait. Later onset cases may mimic slowly progressive acquired neuropathies. Fine motor performance is more affected, with cramping pain in distal extremities, and visible atrophy.

Musculoskeletal system

- Pes cavus, hammer or claw toes, quadriceps weakness develops , with increased falls due to fatigue and proprioceptive loss.
- Fine motor performance is more affected, with cramping pain in distal extremities and muscle atrophy.
- Atrophy of the calves, results into an inverted "champagne-bottle" appearance to the lower extremities.
- Peripheral nerves are often palpably enlarged.
- Large fiber sensory loss is present, with a marked reduction in vibratory perception and proprioception.
- Ankle jerk reflexes are lost. Since this is a demyelinating polyneuropathy, nerve conduction velocity measurements are characteristically slow, at approximately 50% of normal values

Pulmonary system

- Laryngeal dysfunction is characteristic of CMT2c due to alterations in TRPV4 causes sleep apnea, restrictive disease and vocal cord paralysis can also occur in some CMT2 and CMT4. Cardiac function is usually unaffected4..

Auditory system

- Impairment characteristic of some types, varied onset and severity; audiologic screening should always be done in CMTX1.

Visual impairments

- Vision impairment is uncommon; optic atrophy can occur in CMT 2a and CMTX5, and glaucoma and cataracts in some CMT4.

Intellectual disability

- Some CMT4, CMTX4 (Cowchock) and CMTX5 (Rosenberg-Chutorian/Arts). Learning differences and cerebral white matter changes may be seen in CMTX1 and others.



Fig 9 A to F: Deformities in HSMN

- Awkward or higher than normal step (high steppage gait)
- Patients may have sensory gait ataxia, and a Romberg test usually yields positive results.

*Pathophysiology*

The most common form of CMT1, CMT1A, was associated with a 1.4 Mb duplication at chromosome 17p11.2. CMT1B, linked to chromo-some 1, is caused by mutations in the gene of myelin protein zero (MPZ) glycoprotein.CMT1a has overproduction of peripheral myelin protein; histologically, onion bulbs, focal folds and/or swelling occur. Axonal forms may have abnormal mitochondrial fusion or fission, or other defects in axonal transport and intracellular membrane trafficking.

X-linked neuropathy, CMTX1, is due to mutations in the gap junction (GJB1) on the X chromosome encoding the gap junction pro-tein, connexin 32.Dejerine-Sottas disease (DSD), a severe infantile neuropathy, is associated with mutations or deletions in peripheral myelin protein 22 (PMP22). With early onset, palpable nerves, and marked onion-bulb formation.

*Assesment*

- Family history
- On observation: attitude of limbs,spine and deformities
- On examination

A. Signs and symptoms

B. Muscle tone

C. Reflexes

D. Sensory examination

E. Gait

*Diagnosis*

- A complete clinical evaluation of muscle atrophy, weakness, and sensory responses, and should include a detailed patient history.
- EMGs and motor nerve conduction test help diagnose the type of HMSN as well as measure its severity.
- Specialized DNA blood tests are helpful in some cases.7

*Complications*

- patients with CMT disease are susceptible to skin breakdown or burns, due tooss of protective sensation distally in all four limb
- nonhealing foot ulcers,
- in severe cases, bony deformities of bilateral feet.
- orthoses are required for treatment of foot drop or to accommodate bony foot deformities. If not fitted properly, the orthoses themselves become a source of skin breakdown secondary to associated distal sensory impairment.

*Prognosis*

- CMT is nearly always slowly progressive. A longitudinal study showed steady progression in nerve conduction velocity and disability, and severity could be predicted based on nerve conduction velocity abnormalities in childhood.
- CMT disease is not a fatal disease as it does not affect normal life expectancy. However, significant inconvenience can occur as 'normal' lifestyles will be affected. Most patients suffering from CMT disease continue to work. Wheelchair confinement is rare.

## Physiotherapy Management

Physical Therapy assessment includes:

1. History:

This includes history of present illness, medical and surgical history, family history oranyexposuretoanyotherdrugsshouldbe assessed.

History of present illness:

The history of the presenting illness or chief complaint should include the following information:

- Symptom onset (acute, subacute, chronic, insidious)
- Course of the condition (static, progressive, or relapsing and remitting)
- Associated symptoms, such as pain, headache, nausea, vomiting, vertigo, numbness, weakness, and seizures
- Duration

Medical History:

It includes history of diabetes, hypertension, cholesterol, asthma or any other comorbidities to be assessed.

Surgical History:

The history of surgery been done of tendon transfer like tight heel cords, hammer toe straightening and arthrodesis of ankle.

Family History:

Majority have a family history of the condition, or having neuropathy in family history. Presentation of pes cavus or abnormal gait represents of having some inherited disorder like CMT. Family tree tracking both sides of the family for 3 generations to rule out who was affected and what mode of inheritance is present whether anybody had trouble with walking, balance, tripping, falling, or with their hands or sensations should be find out.

An autosomal dominant type will have people affected in all generation and male-to-male transmission can be seen thus males are severely affected. Whereas autosomal recessive pedigrees may have only one person or sibling affected with no family history

- At times with no family history does not preclude the diagnosis, as new mutations are relatively caused.

**2. Observation:** Analysis of movement, observation of the condition and shape of muscle contours should be noted. Functional difficulty like clumsiness in gripping a cup or difficulty in manipulating small items such as bottle tops because of wasting of the intrinsic hand muscles. Even muscle imbalances will lead to shortening of unopposed muscle groups resulting in contractures and deformities.

**3. CranialNerveExamination:** Generally cranial nerves are normal at times abnormalities in papillary constriction and in optic nerve. In some types of poly-neuropathy, the cranial nerves are affected. Therefore, it determines the prognosis & severity of the disease.

**4. Sensory Examination:** In conditions such as predominant sensory neuropathy like diabetic neuropathy may be grossly affected. Pinprick, light touch, proprioception, vibration, graphesthesia and temperature should be assessed in terms of extent and pattern of involvement. Both small and large sensory nerve fibers show decreased or absent sensation. Reduction in pin prick and vibration sensation is seen and more pronounced at the toes than in more proximal muscles with sensory ataxia (i.e., imbalance and in- coordination) due to loss of proprioception may also be seen.

**5. ROM testing:** Assessment of both active & passive range of motion to be done & tightness and contracture to also be assessed.

**6. Muscle Strength:** should be assessed with MMT where decreased strength is noted in the distal muscles of the arms and legs like intrinsic hand and foot muscles and tibialis anterior as distal muscles degenerate (axonal loss) first while maintaining strength in proximal muscles and in the gastrocnemius muscle. They are also associated with atrophy and weakened foot eversion. Hand weakness is also seen in terms of poor finger control, poor hand writing, and difficulty using zippers and buttons, and clumsiness in manipulating small objects. A hand held myometer has been shown sensitive and reliable in assessing neuropathies. Weakness in hand muscles can be assessed by using a grip dynamometer which is used in distal neuropathies.

**7. Reflex Testing:** should be tested as deep tendon reflexes are diffusely absent (a reflexia) or reduced (hyporeflexia) and can occur in any condition involving nerve damage.

**8. Girthmeasurement:** is done in order to detect the degree of wasting or atrophy of the muscles.

**9. Deformities:** The deformities such as pes cavus are common and are associated with a reflexia; give the likelihood of having CMT. It results from muscle imbalance as anterior tibialis and the intrinsic foot muscles are affected with sparing of the gastrocnemius muscle thus the stronger pull of gastrocnemius overcomes the weaker pull of anterior tibialis, leading to structural foot deformities. In upper limb, claw hand deformities can be seen.

**10. Gait:** Assessment of the gait reveals difficulty in walking, twisting of ankles, slapping of the feet, or loss of a heel-to-toe pattern, and the patient may walk with a high- steppage gait (lifting legs up excessively to clear the toes) which is due to tight heel cord, and weakness in tibialis anterior leading to inadequate strength to pull the foot up during ambulation. Frac-

tures are common. At times may require bilateral aids, such as ankle-foot orthoses, to ambulate. If quadriceps muscle weakens, hyper extension occurs at the knees to produce a rigid structure when weight bearing whereas when proximal muscles in lower limbs like hip abductors weakness is seen it results in positive Trendelenberg's sign positive.

**11. Autonomic Dysfunction:** Sweating due to poor vasomotor control leading to cold feet with blotching or pallor of the skin of the feet is noticed.

**12. Neuropathic pain (burning, tingling, shooting):** may also occur as a result of the neuropathy, while bone and joint pain may result from pressure on the feet. Muscle cramps and restless legs is also been noticed. Neuropathic pain can be treated with gabapentin, pregabalin, duloxetine, and amitriptyline. Topical lidocaine patches may help localized pain. Narcotic analgesics need to be carefully considered. Joint, bone, and muscle pains require different approaches and should be treated appropriately.

**13. Respiratory System:** is examined in terms of rate of respiration, chest expansion and vital capacity as in acute phase of polyneuropathy especially in GBS, respiration is compromised and at times the patient may need ventilator for respiratory assistance.

**Fatigue testing:** Fatigue can be seen in any activities such as getting tired while writing letters or walking distances. Fatigue severity scale has been demonstrated to be reliable measure in neuropathies. The patient is asked to maintain a diary documentation activities and length of time performing each one and the symptoms of fatigue. Later it is to be analyzed to try and develop a structure to the patients activities that prevent further fatigue and also to check whether this is not leading to disuse by cutting out all activity that cause any degree of fatigue.

**14. Functional Assessment:** is also measured to any change in performance level. For e.g. ability to rise from a chair and standing on heels in case of diabetic neuropathy.

**15. Electrophysiological tests:** is done to differentiate between a demyelinating and an axonal process. It serves as a prognostic factor in determining whether there are any axonal changes as seen in GBS where once axonopathy is permanent, there are no changes left for repairing the axon.

**16. Gene testing and counseling:** Genes for inherited neuropathies can be identified by deoxyribonucleic acid (DNA) testing. Once a patient is shown to have the gene, counseling can be offered and the implications for other family members are discussed. Prenatal testing can be identify a fetus at risk of developing an inherited neuropathy and give the parents the option of termination.

## Management:

*Physiotherapy Interventions:*

Acute Neuropathies:

Physical management in acute neuropathies includes prevention of contractures, control of pain and respiratory care. It includes Guillain- Barre- Strohl (GBS) syndrome.

**1.** Chest Physiotherapy:

In initial stages, the respiratory care should be taken in patients with GBS.[4]

a. To prevent respiratory complications

b. To maintain clear airways

c. To drain out the secretion -continuous change of position

d. To facilitate air entry to all the lobes of the lung

e. Proper & periodic suctioning to be done

f. Vibrations, percussions, manual mobilization to help clear the secretions

g. Nebulization to be used in respiratory infection.

When there is facial muscle weakness, care must be taken to ensure there is lip seal around the mouth piece when measuring vital capacity. Where the autonomic system is affected, disturbed blood pressure is seen when attempting suction or making early sitting.

**2.** Positioning:

Frequent position changes are recommended in bed ridden patients to prevent selective muscle shortening and pressure sores. Truncal weakness can cause the patient to lie with scapulae retracted, unless a wedge is positioned under the thoracic region as well as the head and shoulders. The pelvis should be supported in a position of posterior tilt with the hip flexors stretched, to prevent shortening. Every two hourly turning helps in preventing pressure sores. If the sore is developed, ultra-violet radiation is given to enhance the healing process. The patient's position is maintained with the help of sand bags, pillows etc. If splint is given, then frequent checking is maintained to avoid the skin breakdown around the application area.

**3.** Maintenance of Range of Motions at all joints:

The passive range of motion exercises to be done to maintain the range of joints. This passive ROM exercises to be given minimum of 3 times/day with 5-10 repetitions of each movement of all the joints. Passive exercises help in maintaining the circulation.

**4.** Stretching to prevent contractures:

To prevent tightness of the soft tissue structures, sustained gradual stretching is to be done.

**5.** Splinting to the limbs:

To prevent contracture as well as to immobilize the part of the limb from any unwanted damage to the muscles or joints, the patients are advised for static splints. To maintain the part of the limb, especially hand & foot, the splints are commonly used.

**6.** Pain:

Pain is an early symptom in acute neuropathy such as GBS due to spontaneous discharge in demyelinated sensory neuropathies. Freeman suggested that the reason why patient with acute GBS enjoy large amplitude mid range movements is because of their pain relieving properties.

**7.** Muscle properties maintenance:

In GBS, muscle atrophy is very rapid. Functional Electrical Stimulation (FES) or interrupted direct current to the functional muscles may be used in order to maintain the muscle property [4].

**8.** Prevention & treatment of pressure sores:

This can be prevented by continuous change in position, to check the areas of the skin on a regular basis where the sensations are compromised, gentle massage of the pressure prone areas. Ice cube massages around the wound can be given in case of pressure sores. Ultraviolet radia-

tion (UVR), LASER. Infrared may accelerate wound healing through preventing infection and keeping ulcer surface sterile and dehydrating it [5].

**9.** Strength Training:

The aim is to maintain the strength of weak muscles which can be achieved by the following techniques:

1. Free active exercises.
2. Proprioceptive Neuromuscular Facilitation (PNF).
3. Progressive resisted exercises.
4. Suspension therapy exercises.
5. Equilibrium and righting reactions.



Fig 10: Suspension therapy exercise

*Chronic Neuropathies:*

Physiotherapy plays a major role in chronic neuropathies which help in maintaining ambulation and prevention of contractures. It helps in facilitating functional recovery in bedridden patients of neuropathies thereby preventing complications and improving the quality of life of patients with chronic neuropathies.

Physiotherapy interventions in chronic neuropathies include:

1. Stretching to prevent contractures

Neuropathy may result in severe disability making the patient totally bed-ridden. When the muscles are not stretched adequately but are left in a shortened position, structural changes, involving loss of sarcomeres, occur and compromises potential recovery. The therapist should ensure that all structures including the nervous system are moved through their full range. Adverse neural tension signs can occur if neural tissues are not stretched. Gentle stretches for muscle groups particularly in HMSN, are done to the muscles that are liable to shorten thereby preventing muscle tightness and loss of strength. Tightness and contracture affect the extensibility of muscle and surrounding connective tissues.



Fig 11: Iliopsoas muscle stretching

Tightening of posterior crural muscle group develops rapidly. Regular stretching of 15 to 30 seconds trice a day can prevent or reduce joint deformities that may result from uneven pulling of muscle on bones. Serial night casting for 4 weeks induced a small increase in ankle dorsi flexion range in children and young adults with CMT.

A recent case report suggested that when triceps surae muscles are weakened, stance

and gait gets affected however range of motion is affected.



Fig 12: Stretching of Tendo- achilles

Continuous passive motion machine has been suggested as a mean to maintain range of movement. Even passive ROM is given 2-3 times in a day. Able patients should be taught self -stretches in a weight-bearing position. For bed ridden patients, foot drop splints should be provided and gentle stretches are performed. Thus, muscles which crosses two or more joints must be stretched to its full length.

2. Strength Training:

The aim is to maintain the strength of weak muscles which can be achieved by the following techniques:

a. Free active exercises.
b. Proprioceptive Neuromuscular Facilitation (PNF).
c. Progressive resisted exercises.
d. Suspension therapy exercises.
e. Equilibrium and righting reactions.

Low-intensity to moderate intensity strength training programs in patients with neuromuscular disease, including Charcot Marie Tooth disease (CMT), show varying results.

Kilmer et al.1994 found that higher resistance exercise was no better than low resistance exercises for strengthening muscle and there was some evidence of damage when tested eccentrically. Endurance training can improve cardio pulmonary fitness seen in established neuropathies. Six weeks home programmed of strengthening, stretching and aerobic conditioning increases strength and quality of life.

Concentrating on the hip and knee helps in compensating and producing a more stable gait pattern. The additional AFO would also increase stability by allowing the development of plantar flexion movement despite very weak muscles, as well as preventing tripping. Abnormally prolonged muscle ache following exercise or sudden loss of functional ability indicates damage due to over use and modification of exercise to be tailored made in order to allow recovery.

In one randomized study where CMT patients underwent home-based strength training were evaluated using outcome measures such as isokinetic knee extension and flexion; maximal voluntary contraction; endurance at 80% MVC; ability to descend and climb stairs and stand up from a chair or from lying supine; and time to walk 6 m at a comfortable pace or 50 m quickly showed a significant reduction in the time taken for a 6- meter walk at 24 weeks after starting the exercise with no other significant improvements in time-scored functional activities

Both studies observed limited improvement in upper-body and lower body strength, but Kilmer and coauthors observed a high number of injuries in their patients and concluded that increases in training frequency, volume, and especially intensity may put patients with neuromuscular disease at increased risk of training-induced injury.

Recently 24-week cycling program using a cyclo-ergometer in limited number of pa-

tients (4 with CMT1A and 4 with CMT2) proved to be effective in improving exercise tolerance and functional ability in the absence of significant changes in fatigue resistance. Another study tested the effects of a 12-week, home-based resistance exercise program on strength, body composition, and activities of daily living (ADLs) in 9 men and 11 women with CMT1A (n=18) and CMT2 (n=2). Strength and ADLs improved equally in men and women, and there were no differences between CMT1AandCMT2.Therefore, similar rehabilitative approaches may be useful in CMT, but with the safety and efficacy. Also, overloading of the weakened muscles can cause fatigue and weakness thus overload principle needs to be considered.

3. Re-education of sensory awareness: The sensory system can be stimulated by the cutaneous stimulation which can be given by the different material, textures, shapes and weights. Equilibrium and righting reactions help in reeducating the proprioception of sensory system. Vision can also be used as an alternative system in reeducating the sensory awareness.

4. Postural kinesiotherapy: May be helpful in reducing the need to control joints from three joints (hip, knee, and ankle) to one joint(hip), and Proprioceptive kinetic SI therapy may help to improve coordination.

5. Balance Training: Balance training plays major role as weakness and deformities at foot reduces balance thereby increasing walking in stability. Thus, it should be emphasize by including the following exercises:

1. Standing with feet apart in comfortable posture.



Fig 13: Standing with support of parallel bar

2. Standing with feet apart and keeping the arms in different positions for support.
3. Standing with close feet



Fig 14: Standing with feet together

4. One leg standing.



Fig 15 and 16: one leg standing

5. Repeat all the exercises with eyes closed.
6. Balance boards exercises



Fig 17: Balance board exercises in standing

7. Quadruped side sitting.
8. Hip extension in quadruped
9. Bilateral arm and leg raise in quadruped.
10. Weight shifts in forward, backward and sideways direction in quadruped.
11. Weight shifts in standing & reaching out for objects



Fig 18: Reaching out for an object and pivot turn in standing

12. Tandem walking. Walking sideways and backward.
13. Walking and stopping alternately.
14. Ask the patient to walk in circles.
15. Walk on toes.
16. Walk on heels.
17. Intrinsic strengthening.

6. <u>Functional and mobility aids used for normalizing the gait:</u>

Aids used for function and mobility include a range of orthotics and wheelchairs.

Gait and balance problems are important risk factors for falls in CMT as well as in other peripheral neuropathies. Where ever there is an abnormal gait pattern, mainly due to the weakness of the dorsi flexors and/or the intrinsic muscles of the feet, the use of orthosis should be advised. It has been seen that foot deformities have become irreversible and insoles are necessary to allow the foot to sit comfortably in a standard shoe and to redistribute the weight across the total surface of the sole. When foot drop exists, light polypropylene splint should be advised.

It is essential to discuss skin care with the patient as most of it has sensory deficit also.

- Ankle-foot orthoses (AFOs) may be useful in treating patients with CMT. AFOs compensate for weakness and correct foot drop, can offer a control of the foot, can help control unwanted inward rotation of the foot, and facilitate a more normal gait pattern.

Prescription of AFO for a patient with CMT can enhance physiological performance and perceived exertion at sub maximal activity levels.

- Falls can also be prevented by paying attention to the floor, avoiding uneven ground, being wary of rugs and carpets, avoiding dark places, using a handrail when going up and down the stairs, avoiding haste, and staying slim. When falls are frequent, a walking stick, one or two crutches, or a walking frame is required; it can also be advisable to use a wheelchair outside of the house.
- Wheelchairs: In some of cases, the neuropathy progresses to render the patient dependent on a wheelchair. For e.g. in the early stages of rehabilitation, a patient with GBS, a reclining wheelchair is valuable to coping up possible fluctuations in blood pressure and to allow gradual accommodation in the upright position.

7. Manual dexterity training:

Hand function is also compromised through weakness or paralysis of intrinsic muscles of the thumb and fingers causing difficulty in pincher gripping and finger movement. Normal daily activities such as dressing, bathing, holding cutlery, or writing becomes difficult. Tripod pinch strength and thumb opposition are major determinants of manual dexterity in CMT and should therefore be the focus of intervention strategies that aim to preserve or enhance manual dexterity in CMT. Simple thumb opposition splint may allow a patient to produce legible writing for a longer time, or to grip a cup or knife. At times when there is thenar eminence weakness and wasting is being seen, a night splint cast in a functional position will help prevent severe contracture. Use of putty and rubber bands of different strengths can be used to improve strength of hand and forearm muscles.

8. Adaptive devices:

Special tools modification is given at times in order to avoid overwork and weakness thereby improving performance. Hand or forearm splinting may be advised later once wasting of the hand muscles sets in. Adaptive equipment may be prescribed to compensate for the hand deformities, sensory loss, and weakness which include a button-hook, along- handled shoehorn, and elastic shoe laces.

9. Neuropathic Pain Management:

Malalignment of joints due to muscle imbalance often leads to pain. In chronic neuropathies, neuropathic pain is seen which is caused by damage or dysfunction in the nervous system, which includes the spinal cord. It can generally be described as a sharp, shooting, or burning pain,

Management of neuropathic pain becomes very important as it hinders to the day-to-day activities of patients. Pain management can be divided into physiotherapy, behavioral and cognitive interventions.

A. Physiotherapy interventions in Neuropathic Pain include:

1. Thermotherapy - can reduce muscle

spasm thereby relaxing the muscle.

2. Cryotherapy- can be used in case if there is any swelling at some distal joints.
3. Massage - can reduce muscle tension by gentle touch thereby increasing local blood circulation and relaxing muscles and soft tissues.
4. Transcutaneous Electrical Nerve Stimulation (TENS): is proved to be useful in neuropathic pain. Low rate TENS or Acupuncture Like TENS acts on small diameter C fibers and acts on mechanisms of endogenous opiates thereby blocking pain gate mechanisms.
5. Ultrasound: has also been used in peripheral neuropathy as it reduce muscle spasm and pain there by acting on both large diameter A fibers and small diameter C fibers.

B. Cognitive Strategies: are used in treating together the body, mind and soul.

C. Behavioral interventions: Distraction – listening to music or using imagery techniques can be helpful during brief episodes of pain or painful procedures. Music therapy has been used successfully to reduce disruptive behavior or aggression attitudes.

10. Aerobic Exercises:

Patients with chronic neuropathies show reduced peak oxygen consumption and decreased functional aerobic capacity, and studies shows aerobic exercise improve functional ability and aerobic capacity. Aerobic walking has been used in neuro muscular disorders and was effective in ameliorating peak power output and peak oxygen intake, walking ability, and metabolic changes.

11. Aquatic therapy: is advised as it increases the muscle power, improve flexibility of shortened soft tissues, coordination and balance of the patient. Care is taken in case of flaccid joint. Pain which is also a feature of polyneuropathy can be relieved by hydrotherapy as water has pain relieving property.

## Occupational therapy management

Occupational Profile: The occupational therapy evaluation begins with an interview that gathers information about the client's relevant history of disease, valued roles and occupations, and screens for specific concerns or problems. This interview must be sensitive to the client's changing function and fear of the unknown as well as illuminate problem areas that may not be spontaneously discussed such as fatigue, depression, sexual function, and cognitive concerns that impact their occupations and social network. With this information, an occupational profile assists to direct further assessment.

Occupational Analysis Based on the occupational profile, the occupational therapist will select assessments and measurement tools that will provide in-depth information on occupational areas of concern. For persons with neurodegenerative disease, the content of these measures may be limited to a few or cover a broad spectrum of occupational performance areas (such as ADLs, IADLs, and work) or symptoms (such as sensation, spasticity, weakness, memory, and fatigue). Occupational Therapy Intervention Process Goal Setting The primary goals of intervention are to reduce the effects of disability resulting from disease impairments and to maintain or promote independence and quality of life; that is, safe, functional, and comfortable participation in chosen tasks and roles.

Goal setting is a collaborative process that includes the client, occupational therapist, and as appropriate, family and other significant persons. The unplanned and profound impact of neurodegenerative disease frequently makes realistic goal setting complex, and requires sensitivity, flexibility, and negotiation skills of the occupational therapist. The therapist frequently helps the client modify behaviors and assumptions of a lifetime while assisting the person and significant others to move toward realistic new goals.

Treatment goals should be in sync with the client's priorities and grounded in evidence-based research. Given the progressive nature of many neurodegenerative diseases, regular reassessment and reordering of goals and priorities may be necessary. Intervention Implementation Due to the complexity of disability, intervening in one area may affect several other factors, such as fatigue, pain, and weakness. Interventions are varied and frequently involve significant others, especially as the disease progresses.

The following is an overview of ways occupational therapy optimizes performance in key occupational roles for persons with neurodegenerative diseases. Self-Maintenance Roles In basic and instrumental ADL, the occupational therapist assists the client to attain safety, maintain independence in priority activities, use specialized equipment, and participate using energy-conserving methods.

Treatment strategies include the following:

- Priority setting
- Education in areas such as the disease process affecting motor or cognitive changes
- Environmental modifications at home and work to promote safety and independence
- Exercise to prevent loss of range of motion and strength needed to dress
- Behavior modification such as use of energy conservation and time management techniques
- Identifying and obtaining equipment to compensate for weakness or other symptoms
- Balancing independence with assistance from others Self-Advancement Roles

The onset of neurodegenerative diseases affects employment in or outside of the home. Contrary to popular expectations, however, many individuals can and should continue to work rather than claim disability. Medical treatments affecting disease progression and legislation mean that modification of employment is frequently more appropriate than an immediate disability claim. Continued employment has implications for financial independence, health care choices, socialization, identity, and self-esteem.

Occupational therapists critically review job expectations and, as required, make modifications to the environment, recommend behavioral changes, prescribe equipment, identify resources, and make recommendations to the individual or the employer. Nonprofit groups such as the National Multiple Sclerosis Society (NMSS) provide resources and are excellent sources of information for both patients and therapists (Resources 40-1). Self-Enhancement Roles maintaining leisure pursuits may be a high priority for individuals with neurodegenerative diseases but are often the first roles to be abandoned. Modifying activities and proper equipment often en-

able the individual to continue to carry out these important meaningful occupations.

*Maintain the quality of life through following strategies:*

- Developing communication tools: such as sign or picture board
- Ensuring access to the nurse call button, TV, and lights by remote control, as appropriate
- Modifying the telephone for hands-free use
- Modifying lying and sitting positions for optimal function and comfort
- Positioning for trunk, head, and upper extremity stability

Teaching about GBS and strategies to reduce anxiety Recovery-phase interventions are oriented to the resumption of activities and roles. Examples of occupational therapy interventions include:

- Providing activities and dynamic splints to maintain range of motion, particularly of wrists, fingers, and ankle (hinged drop-foot orthoses)
- Instructing in safe mobility and independent transfers
- Training in modified self-care techniques and adapting other daily activities
- Adapting modes of communication according to the person's priorities
- Encouraging access to the community
- Modifying and encouraging reengagement in routine activities, as appropriate
- Adapting equipment and modifying behavior in home, leisure, and work activities
- Instructing in energy conservation and fatigue management strategies
- Modifying employment roles, tasks, and environment, as indicated
- Fine motor program to enhance strength, coordination, and sensation
- Home assessment and modifications, as appropriate, to facilitate return to home

## Psychological management

Neuropathic pain is characterized by sensory and affective disturbances, supporting the idea that pain and mood disorders share common pathogenic mechanisms. Biological and neuro-imaging data show that common brain areas are involved in the modulation of painful and emotional experiences. So, an improved understanding of the molecular mechanisms underlying the role of neuro-inflammation in regulation of affective behavior in neuropathic pain is important for the development of novel therapeutic strategies. Psychological issues are a factor influencing treatment and outcome in patients with neuropathic pain. It is important, therefore, to treat an affective disorder that coexists with pain. The affective disturbances associated with pain include working memory dysfunction, impaired cognition, decreased appetite, depression, anhedonia, disruptions to sleep cycles, and impaired familial and social interactions.

There is a strong overlap between neuropathy pain, anxiety, and depression. Each of the three can raise the risk for having the other two. 30-60% of patients with chronic pain develop feelings of depression and anxiety. Pain and depression co-occur 30-50% of the time and each can cause the other.

Anxiety is to be expected in the face of be-

ing diagnosed with a chronic illness. Anxiety can take many forms. There is a difference between anxiety and chronic anxiety. It may be specific worries about the illness such as: Fear of relapse or worsening, disability, isolation, stigma, and pain

*Assessment:*

Assessments are directed towards understanding the severity and the levels of accompanied distress. This distress might be present in the individual in the form of depression, anxiety or even changes in social abilities. If the neuropathy is seen to affect cognitively, assessment may include tests of general intellectual abilities, memory and learning skills, and may also include tests of attention, speed of information processing, and executive skills. Assessment of intellectual ability may include basic education test (general knowledge, arithmetic, vocabulary, word reading) and various reasoning task to assess the ability to apply this knowledge to new problems. Assessment of memory function is likely to include recall, learning, and retention of both verbal materials (stories and list of words) and visual materials (geometric shapes, faces, or objects). More specialised tests of other areas of cognitive functions such as perception and language might be carried out according to the inquiry.

*Treatment:*

Social skills training:

As some neuropathies can cause cognitive impairments, mild neuropathies can cause issues related to body image, contributing to difficulty in communication and social functioning. Thus, the patient may experience low self esteem, lowered confidence in approaching others and social withdrawal as a result. Many people experience difficulties with their social skills and relationships due to a combination of cognitive and emotional changes.

Social skills training may be of value in raising self-awareness and social awareness, as well as providing a forum for peer support. It can also help people to accept their limitations and bring about a positive change in the social activities. This can help people to rebuild the confidence, both in maintaining the existing relationship and in making new friendships.

Psychological counselling

People with neuropathies benefit from feedback and advice about the nature of the psychological difficulties that they will suffer from as a result of the diagnosis. A detailed explanation of the nature of the person's difficulties, and guidance about the likely extent of recovery, can help them again more realistic outlook, and so help them make appropriate plans for the future.

This is especially important where there is a tendency for the person to underestimate the extent of the difficulties, and to overestimate future recovery. In the early stages of recovery, this belief may protect people from becoming depressed and serve as a powerful motivating factor. However, continued unrealistic expectations of recovery may impede both progressive rehabilitation and adjustment long term disability.

Psychological counselling may also help in evaluating the employment opportunities and getting back in the social and leisure activities. Relatives usually benefit from detailed feedback about the results of formal assessments, the nature and implications of acquired disability, and some explanation of the rationale and goals of treatment. They often seek advice about management of cognitive and or behaviour problems and may assume the role of

co- therapist in reinforcing treatment strategies at home.

*Individual psychotherapy*

Some people, understandably, feel devastated by the effects of their neuropathy. Although psychologist can help working through clinical depression and anxiety, along with other related issues, people who are not falling on the clinical spectrum also benefit from psychotherapy. Some people are unable to accept the changes that have occurred, and may continue to judge themselves by the previous standards that they cannot meet any longer. For others, the preoccupation with what they have lost may cloud their appreciation of the existing positive attributes and potential.

Psychotherapy can help people to re-appraise their new situation. With support and guidance they can often be help to move forward to explore the new selves, and search for a new direction and purpose to their lives. Cognitive behavioral therapy (CBT), insight oriented therapy, relaxation training, development of social skills, and anger management can also be useful in helping a person understand and accept the injury as well as adjust relationships and roles after the injury, when necessary.

Individual psychotherapy may also help to deal with the secondary problems that may arise with neuropathic pain. They can be alcoholism or any other substance abuse which has its roots in trials to cope with the distress. A better coping mechanism can be developed by the person with the help of the psychologist and deal with the pain more appropriately.

*Family therapy*

Family members face similar process of adjustment following a neuropathy. As the person improve, against their Independence and resumes former roles, family relationships can actually revert to their previous pattern. However, for those with permanent disability, some changes in family roles and relationships maybe necessary. Many families adjust well towards the changes, and some report feeling closer as a result. Children may also benefit from help in understanding and coping with the changes in their parents.

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# Ch. 19 Peripheral Nerve Injury

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A peripheral nerve is a connection between the brain and spinal cord and the rest of the body. They are an extension of the neuronal bodies placed in the spinal cord or dorsal root ganglion and also the brain.

**Structure of the Peripheral Nerve**

A single peripheral nerve is made of a number of axons arranged together in a group called fascicle. The connective tissue covering each axon is called the endoneurium. Each fascicle is covered by perineurium. The interfascicular connective tissue that separates one fascicle from the other is called as the epineurium. These connective tissues help in protecting and providing nutrition to these fibers.

The neural structure is comprised of the axon and its accompanying Schwann cells. The Schwann cells are placed along the longitudinal extent of the axon. In between the Schwann cells, there is a node of Ranvier which is responsible for the salutatory conduction of impulse.

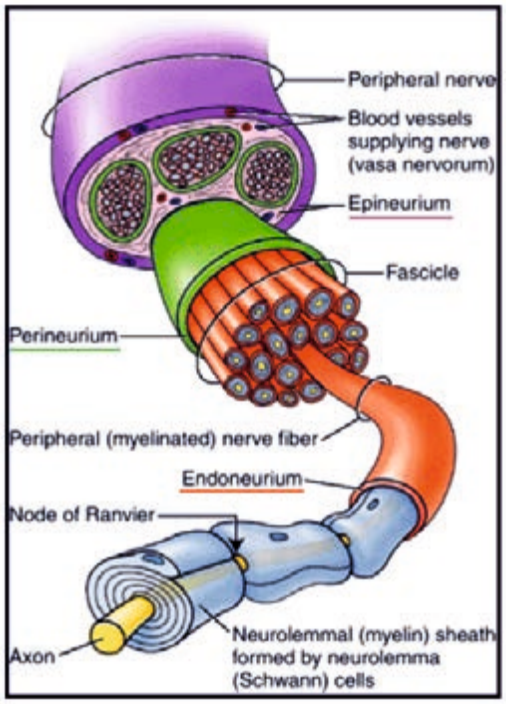



*Fig 1: Structure of Peripheral Nerve*

**Classification of Peripheral Nerve Injury**

| | Sunderland Classification | Seddon Classification | Injury Description | Recovery Duration |
|---|---|---|---|---|
| Mildest | I | Neuropraxia | Conduction Block | Less than 3 months |
| | II | Axonotmesis | Axonal discontinuity | 1 inch per month |
| | III | | Axonal and endoneurial disruption | Less than 1 inch per month |

| | | | | |
|---|---|---|---|---|
| | IV | | Fascicle and perineurial disrupture | Recovery requires surgical intervention |
| Most Severe | V | Neurotmesis | Complete transection of nerve trunk | Recovery requires surgical intervention |

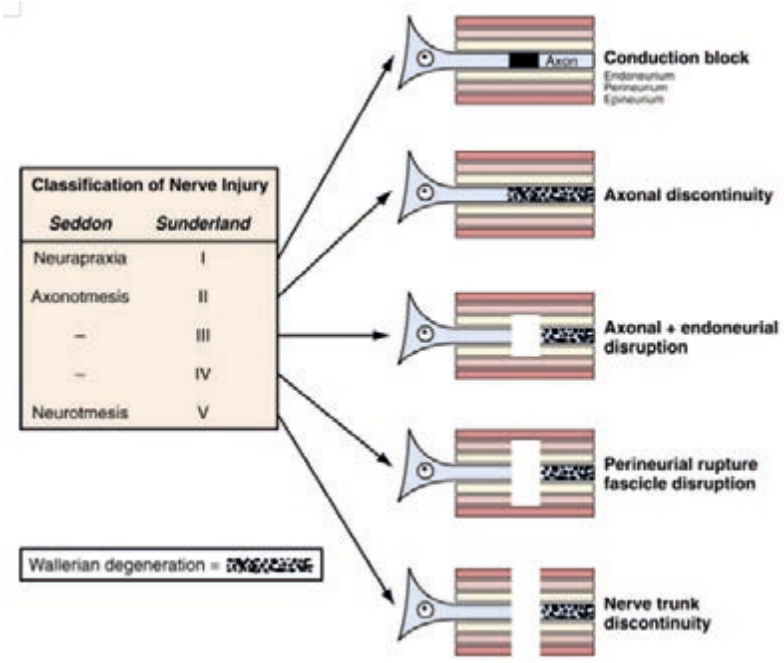



*Fig 2: Classification of Peripheral Nerve Injury*

**Brachial Plexus Injury**

- Commonly occuring palsy: Erb's Palsy and Klumpke's Palsy

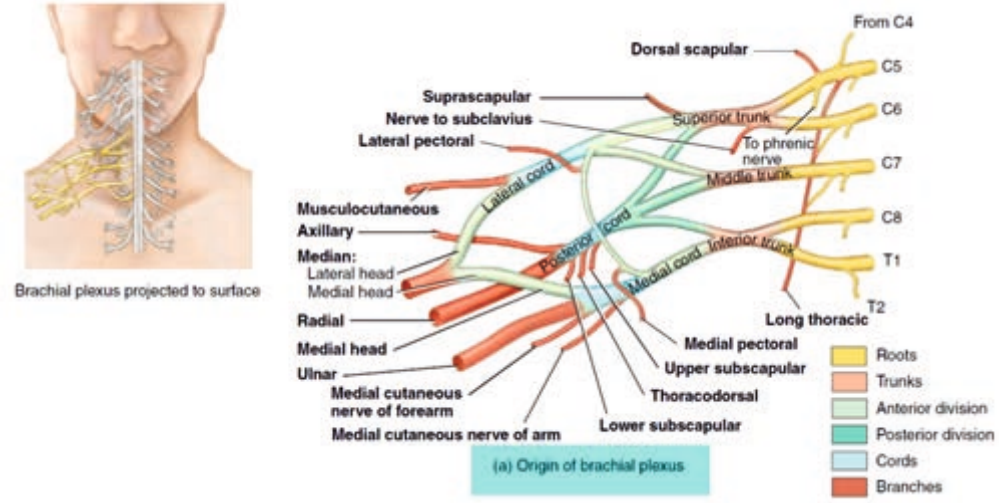



*Fig 3: Origin of brachial plexus*

**Erb's Palsy:** Injury to Upper trunk originating from C5-C6 nerve roots

- Common Causes: Vacuum delivery, pressure on supraclavicular injury
- Sensory Loss: Over deltoid insertion and lateral aspect of forearm and hand
- Motor Deficit: Paralysis of shoulder lateral rotators, shoulder flexors, shoulder abductors, elbow flexors and supinator. There might be weakness in triceps, latissimus dorsi, serratus anterior, pectoralis major and extensor carpi radialis.
- Deformity: Waiter's tip wherein shoulder is in extension, adduction and internal rotation; elbow is in extension; forearm in pronation; wrist and fingers in flexion.
- Functional Disability: Difficulty in activity of daily living like eating, combing, dressing, bathing.

**Klumpke's Palsy:** Injury to lower trunk originating from C8-T1 nerve roots

- Common Causes: Breech delivery, traction in abducted arm, enlarged cervical rib.
- Sensory Loss: Over medial aspect of arm, forearm, hand and hypothenar eminence.
- Motor Deficit: Paralysis of all intrinsic muscles of hand alongwith wrist and finger flexors.
- Deformity: Claw Hand Deformity
- Functional Disability: Inefficient power grip

**Axillary Nerve Injury**

- Common Causes: Fracture of neck of humerus, penetrating injury, compression injury
- Sensory Loss: Sergeant's patch over lower deltoid region.
- Motor Deficit: Weakness of shoulder abductors (15o – 90o), shoulder flexors, shoulder extensors, shoulder external rotators.
- Deformity: Wasting of deltoid muscle

**Musculocutaneous Nerve Injury**

- Common Causes: Penetrating injury, compression injury.
- Sensory Loss: Lateral aspect of forearm
- Motor Deficit: Weakness of elbow flexors and forearm supinators.
- Deformity: Wasting of biceps muscles.

**Median Nerve Injury**

- Common Causes: Supracondylar humerus fracture, penetrating injuries, compression injury
- Sensory Loss: Over volar aspect of lateral 3.5 fingers upto distal phalanx on the dorsal side, thenar eminence, pulp of the thumb.
- Motor Deficit: Weakness of pronator teres, pronator quadratus, flexor carpi radialis, palmaris longus, flexor digitorum superficialis, flexor digitorum profundus (lateral half), flexor pollicis longus, thenar muscles, 1st and 2nd lumbricals.
- Deformity: Ape hand or partial claw hand or poiniting index finger
- Functional Disability: Difficulty in holding things.

**Ulnar Nerve Injury**

- Common Causes: Supracondylar humerus fracture, medial epicondylar fracture, penetrating injuries, compression injury.
- Sensory Loss: Over hypothenar eminence or ulnar distribution of hand.
- Motor Deficit: Weakness of flexor carpi ulnaris, flexor digitorum profundus (medial half), hypothenar muscles, 3rd

and 4th lumbricals, adductor pollicis and interossei.

- Deformity: Claw hand
- Functional Disability: Lack of lumbrical and spherical grip, reduction in pinch strength.

**Radial Nerve Injury**

- Common Causes: Humeral shaft fracture, penetrating injuries, compression injury.
- Sensory Loss: Posterior aspect of arm and forearm, radial distribution of hand.
- Motor Deficit: Weakness of triceps, wrist extensors and finger extensors.
- Deformity: Wrist Drop
- Functional Disability: Poor grip

**Obturator Nerve Injury**

- Common Causes: Dislocation of hip joint, prolonged difficult labor, pelvic fracture, hernia through the obturator foramen, compression injury.
- Sensory Loss: Over medial aspect of knee and distal end of the thigh (medial aspect).
- Motor Deficit: Weakness of hip adductor muscles, obturator externus, gracilis and pectineus.
- Deformity: Hip flexion-abduction deformity

**Femoral Nerve Injury**

- Common Causes: Penetrating injury, pelvis fracture, femus fracture, dilocation of hip joint, inguinal hernia, prolapsed intervertebral disc, lumbar spondylosis, lumbar canal stenosis, complication of spinal anaestesia
- Sensory Loss: The anterior division causes sensory loss over anteromedial aspect of the thigh. The posterior division causes sensory loss over medial aspect of leg and foot up to the ball of the foot.
- Motor Deficit: Weakness of quadriceps muscles
- Deformity: Genu recurvatum

**Sciatic Nerve Injury**

- Common Causes: Penetrating injury, pelvis fracture, femus fracture, dilocation of hip joint, incorrectly given intramuscular injection, compression injury, tight piriformis muscle causing nerve entrapment.
- Sensory Loss: Complete loss of sensation below knee except for the area supplied by saphenous nerve (medial aspect of leg and foot up to the ball of the foot).
- Motor Deficit: Weakness of hamstring muscles and all muscles supplied by tibial nerve and common peroneal nerve.
- Deformity: Flail leg with foot drop. There maybe clawing of toes with trophic ulceration.

**Common Peroneal Nerve Injury**

- Common Causes: Compression injury, fracture of fibula neck, fracture of fibula head, Hansen's disease
- Sensory Loss: Deep peroneal nerve palsy causes sensory loss over web space between great toe and second toe, lateral aspect of the dorsum of the great toe, medial aspect of the dorsum of the second toe. Superficial peroneal nerve palsy causes sensory loss over anterolateral aspect of the leg, dorsum of the foot and toes except a small wedge shape area in the web space between great toe and second toe.
- Motor Deficit: Weakness of tibialis anterior, peroneous tertius, extensor digitorum longus, extensor digitorum brevis, extensor hallucis longus.
- Deformity: Equinovarus deformity of the foot.

**Tibial Nerve Injury**

- Common Causes: Injection palsy, dislocation of the knee, tarsal tunnel injury, penetrating injury to knee and upper half of the leg.
- Sensory Loss: Sole of the foot, medial aspect of the heel.
- Motor Deficit: Weakness of gastro-soleous, tibialis posterior, flexor digitorum, flexor hallucis longus and popliteus muscles.
- Deformity: Talipes calcaneovalgus deformity

**PT management in peripheral nerve injury**

- **Motor Retraining:**

Passive movement : Passive movements are movements that are given by an external force during muscular inactivity or when range of motion is reduced for any cause carried out either by the therapist or a machine. Passive movements are helpful in decreasing complication of immobilization, improving circulation through pumping action, prevent adhesion and contractures.



*Fig. 4: Passive movements for upper extremity*

**B. Facilitation technique:** Facilitation is the process of intervention, which uses the improved postural tone in goal- directed activity.

Tapping on the muscles: Tapping is the use of a mild force applied manually over a tendon or muscle belly to facilitate a voluntary contraction. Reflex activity with normal response or brisk response can be tested by tapping technique.



*Fig.5 Tapping on the biceps muscle*

- Quick stretch: Quick and spontaneous stretch can activates agonist to contract and antagonist to stretch which will inhibit its action with reciprocal innervation effect which further activates synergists.



*Fig 6. Quick stretch*

- Quick icing( sensation must intact): There are four types of techniques which can be used such as; ice chips, ice wraps, ice pack and immersion in cold water.



*Figure 7. Quick icing*

- PNF technique- Proprioceptive Neuromuscular Facilitation (PNF) is a group of stretching techniques commonly used in clinical environments to enhance both active and passive range of motion in order to improve motor performance. With the help of quick stretch, distal muscles can be facilitate by resisting strong proximal muscles.



*Figure 8: PNF techniques for upper limb*

C. **Electrical stimulation:** Electrical stimulation of peripheral nerve tissue has been shown to accelerate axonal regeneration. Faradic for innervated muscles and galvanic for denervated muscles, used for muscle reeducation and nerve stimulation.



*Figure 9: Electrical stimulator*

- **Splinting:** A splint is defined as a rigid or flexible device that maintains in position a displaced or movable part; also used to keep in place and protect an injured part.2. Splinting Prevents stiffness and contractures of joints; supports residual nerve functionality and re-innervation.

There are various types of splint used in various peripheral nerve injury condition according to need.

**A. Aeroplane splint:** A complex splint that holds the arm in abduction with the forearm at approximately 90 degrees of flexion. Mainly use in brachial plexus injury. The research suggests that the management of the axillary nerve damage following shoulder dislocation is treated in the same manner as treating an isolated dislocation, with strengthening and stimulation of the deltoids and teres minor muscles.



*Figure 10: Aeroplane splint*

**B. Shoulder sling-** To protect shoulder joint in peripheral nerve injuries as in axillary nerve injury.



*Fig 11: Shoulder sling*

**C. Cock-Up splint-** Radial nerve injuries are associated with the path it travels close to the humerus. Mechanisms of injury can be humerus fracture, a direct blow or sustained pressure (i.e. from incorrect use of a crutch). Motor function of the triceps are usually preserved as they are innervated superiorly whereas wrist and digit extensor are often paralyse leading to the dropped wrist deformity. To prevent such deformity wrist drop splint can be used.



*Fig 12a:Wrist drop from radial nerve injury*



*Fig 12b:Cock-up splint to prevent radial nerve injury*

**D. Ankle foot orthosis-** Common peroneal nerve injury is often seen in people who are; very thin; predisposed to autoimmune conditions such as rheumatoid arthritis; nerve damage secondary to chronic health issue eg Diabetes, Alcoholism; Charcot - Marie Tooth disease, or have a significant knee valgus or varus. Damage to this nerve is followed by foot drop (due to paralysis of the ankle and foot extensors) and inversion of the foot due to paralysis of the peroneal muscles with unopposed action of the foot flexors and invertors). There are methods to improve the foot drop condition such as: use of splinting in a solid ankle-foot orthoses or foot-up splint. These work to increase the amount of dorsiflexion the foot is held in during gait and can prevent falls as the toes do not get caught on the floor.



*Fig 13: Ankle foot orthosis to prevent foot drop*

**E. Finger splint-** The ulnar nerve is most susceptible to injury at the elbow7. and the wrist resulting in varying degrees of motor and sensory loss. Mechanism of injury includes local trauma, nerve impingement, lacerations at the wrist and Guyon canal cyst. Characteristic signs of injury to the ulnar nerve is the inability to grip paper between the fingers with long-term cases resulting in ulnar claw hand deformity. In ulnar nerve lesion to correct hyperextension of MC joint and correct flexion of IP joints finger splints can be given.

- Pain control: Pain is one of the most common and annoying consequences of nerve damage.

➢ Electro therapy: Electric stimulation plays an important role in the treatment of various neuromuscular dysfunctions. There are various modalities use for pain management which as follows:

A. TENS method (20 min) : Research says that Therapy with transcutaneous electrical nerve stimulation (TENS) gives both sufficient analgesia and a noticeable improvement of the functional situation.

Mild electrical impulses transmitted through the skin which causes body to release endorphins, the body own pain reliving hormones. These positive signal to brain blocks slow moving pain messages at spinal level (pain gate theory).




*Fig 14: TENS method*

**B. Continuous ultrasound:** Transcutaneous ultrasound applied to an axonotmetic injury accelerates recovery for proximal affection. Ultrasound therapy gives symptomatic relief in case of carpel tunnel syndrome.




*Fig 15: Continuous ultrasound*

**C. Deep cold laser:** The most popular form of laser therapy used for neuropathic pain is cold laser treatment. The method works by stimulating blood circulation around the affected areas. This is beneficial because of the lack of blood supply around the distal parts of the body, as a result of increased glucose levels. In doing this, the nerve fibers that have been damaged can receive sufficient nutrition and oxygen, thus helping repair and optimize their functionality. The end result is decreased neuropathic pain and discomfort.



*Fig 16:Deep cold laser*

**D. IFT:** It appears the use of pre-modulated interferential stimulation therapy will increase the temperature by increasing the blood flow when in a prone setting of the extremity for patients diagnosed with peripheral neuropathy of the calves and feet.12.



*Fig 17: IFT*

**E. Hot packs and infrared -** To maintain skin visibility ( superficial sensation must intact)

- Sensory Reeducation:
  - Protection of desensitized area to avoid burn and injuries.
  - By using different materials texture such as cotton, silk etc.
  - Various task such as recognizing

different object or quantity of object by touch with blind folded or vision occluded.



*Fig 18: Sensory Reeducation for hands*

## Occupational therapy Management

### Management by phase

Postinjury management can be organized into three phases of recovery: phase one refers to early healing when immobilization is required; phase two is the period of reinnervation when remobilization, sensory reeducation, and active motor control is emphasized ; and phase three stresses strengthening and functional performance. Rehabilitation methods often include orthotic positioning, active range of motion, biofeedback, neuro muscular electrical nerve stimulation, and functional tasks. Adjunctive methods include massage, edema management, and sensory reeducation.

Phase 1:

- Custom made orthosis or cast is used to protect the repair by limiting tension on the repair site for minimum of 3 weeks. Static orthosis provides appropriate biomechanical position to prevent over stretching of denervated muscles and deformity of associated structures.
- Management of pain, edema, and inflammation can be addressed with anti-inflammatory medications and physical agent modalities such as ultrasound, phonophoresis, and cryotherapy.
- Education is often provided to:

1. Aid in edema reduction via elevation and active movement of non-involved joints.
2. Protect areas of diminished or absent sensation.
3. Review home programs.

- Performance of ADL is addressed using adapted methods and assistive devices.



*Fig 19: ADL training with adaptive device for hand*

Phase 2:

- Active range of motion exercises to foster muscle activation and movement. Active range of motion is initially encouraged through a protected range with gravity eliminated then progressed to full motion against gravity.
- Aquatic therapy can be beneficial, as the buoyancy of the water can provide assistive active range of motion.
- Therapeutic activities focus more on motor relearning and adaptive strategies. Biofeedback and neuro muscular electric stimulation assist with motor learning and control. Biofeedback provides visual or auditory feedback (or both) when the targeted muscle contracts, aiding relearning of muscle activation or relaxation. NMES augments muscle activation when there is evidence if innervation, but also provides proprioceptive and tactile input to enhance muscle activation and awareness.

- The sensory glove provides auditory feedback to substitute for tactile input during fine motor tasks after PNI.
- Mirror visual feedback , which uses mirror image to provide an illusion of movement in the involved hand, is another useful technique that's being practiced. In this the individual is instructed to observe a movement being completed by the non involved hand in a mirror image located at the midline (involved hand is behind the mirror).

Phase 3:

- Strengthening is started once full active range of motion is obtained with gravity eliminated and against gravity.
- Resistance can be in the form of theraputty, theraband, free weights, cuff weights, exercise machines, work simulators, and computer games (e.g. biometrics device).

**Psychological Management**

Disorders involving the peripheral nervous system can have devastating impacts on patients' daily functions and routines. Research studies provide evidence that pain associated with nerve injuries can be severe and are associated with the level of disability. In addition, both increased pain and disability levels are predictive factors for poor quality of life. Similarly, studies of patients with either upper or lower extremity peripheral nerve disorders have shown that patient-reported quality of life is significantly worse when compared to healthy control subjects.

Stress and ability to cope with stress are also important yet under-reported factors in the people suffering from nerve injury. Individuals are at increased risk for chronic pain and disability when "catastrophizing" is used as a coping strategy. Pain catastrophizing is a maladaptive cognitive-affective response to pain that involves negative thinking regarding the pain experience. In 2010, Cederlund et al. reported that patients with a low capacity to cope in a stressful situation, were significantly less satisfied in daily occupations, had lower quality of life scores, and reported more pain.

Psychosocial factors, such as depression, coping, and anxiety, depend on many common patient outcomes, such as levels of pain, patient satisfaction, and disability. According to studies, those who consume alcohol reported significantly lower pain, greater ability to cope with stress at home, and lower work stress levels. Previous work has shown that coping is a frequent motive for the consumption of alcohol.

**Post traumatic stress disorder (PTSD)**

Some people may experience post traumatic stress disorder (PTSD). This is a serious psychological reaction to a traumatic event/ experience. It involves the persistent re-experiencing of the trauma, avoidance of stimuli which remained the person of the event, increased arousal, and a number of emotional responses. As such, PTSD is most commonly experienced after mild accidents when memories of the circumstances surrounding the injury are retained. However, some others develop of fear of circumstances similar to that of the injury and a small number of people have disturbing memories of the early stages of their recovery.Traumatic upper limb PNI may result in lifelong cosmetic and functional deficits to the hands. In the majority of these patients, peripheral nerve injury of the forearm is accompanied by early post traumatic psychological stress, especially in female adults who suffered from combined nerve injuries.

**Assessment:**

Assessments are directed towards understanding the severity and the levels of accompanied distress. This distress might be present in the individual in the form of depression, anxiety, PTSD or even changes in

social abilities.

**Treatment:**

**Social skills training:**

The upper limbs and specifically the hands are a significant part of perceived body image, contributing to communication and function. Thus, the patient may experience low self esteem, lowered confidence in approaching others and social withdrawal as a result. Many people who suffered from PNI experience difficulties with their social skills and relationships due to a combination of physical and emotional changes. Social skills training may be of value in raising self-awareness and social awareness, as well as providing a forum for peer support. It can also help people to accept their limitations and bring about a positive change in the social activities. This can help people to rebuild the confidence, both in maintaining the existing relationship and in making new friendships.

**Psychological counselling**

People with peripheral nerve injuries benefit from being given feedback and advice about the nature of the psychological difficulties that they will suffer from as a result of the injury. A detailed explanation of the nature of the person's difficulties, and guidance about the likely extent of recovery, can help them again more realistic outlook, and so help them make appropriate plans for the future. This is especially important where there is a tendency for the person to underestimate the extent of the difficulties, and to overestimate future recovery. In the early stages of recovery, this belief may protect people from becoming depressed and serve as a powerful motivating factor. However, continued unrealistic expectations of recovery may impede both progressive rehabilitation and adjustment long term disability.

Psychological counselling may also help in evaluating the employment opportunities and getting back in the social and leisure activities. Relatives usually benefit from detailed feedback about the results of formal assessments, the nature and implications of acquired disability, and some explanation of the rationale and goals of treatment. They often seek advice about management of cognitive and or behaviour problems and may assume the role of co- therapist in reinforcing treatment strategies at home.

**Individual psychotherapy**

Some people, understandably, feel devastated by the effects of their nerve injury. Although psychologist can help working through clinical depression and anxiety, along with related issues, people who are not falling on the clinical spectrum also benefit from psychotherapy. Some people attempt to carry on as before, unable to accept the changes that have occurred, and may continue to judge themselves by the pre injury standards that they can no longer meet. For others, the preoccupation with what they have lost may cloud their appreciation of the positive attributes and potential that remains. Psychotherapy can help people to re-appraise their new situation. With support and guidance they can often be help to move forward to explore the new selves, and search for a new direction and purpose to their lives. Cognitive behavioral therapy (CBT), insight oriented therapy, relaxation training, development of social skills, and anger management can also be useful in helping a person understand and accept the injury as well as adjust relationships and roles after the injury, when necessary.

Individual psychotherapy may also help to deal with the secondary problems that may arise with PNI. They can be alcoholism or any other substance abuse which has its roots in trials to cope with the distress or even PTSD. Treatment of PTSD needs a systematic approach to uproot the causal memories of

fear so that the person is functional and achieves a better quality of life.

**Family therapy**

Family members face similar process of adjustment following a nerve injury. As the person improve, against their Independence and resumes former roles, family relationships can actually revert to their previous pattern. However, for those with permanent disability, some changes in family roles and relationships maybe necessary. Many families adjust well towards the changes, and some report feeling closer as a result. Children may also benefit from help in understanding and coping with the changes in their parents.

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# Section 5
# Neuromuscular Disorders

# Ch. 20 Muscular Dystrophy

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Muscular dystrophy is a condition with progressive muscle weakness. Weakness of the muscles in muscular dystrophy is not uniform. This creates imbalance between the strength of various muscle groups and changes the alignment of the bones that they move. In turn, the muscles spend more energy in moving these mal-aligned bones. Muscles may also become shorter in the length and lose their flexibility. Loss of extensibility leads to structural deformities which are the disease progresses and cause severe limitation of the function and movement. This limitation leads to further imbalance in the muscle weakness and the vicious cycle continues.

Individuals with muscular dystrophy may also present with cardiovascular, respiratory, orthopedic, neurological and gastrointestinal symptoms. It is these symptoms that considerably limit the span of the individuals.

**What are types of muscular dystrophy?**

Different genes are responsible for different protein components of muscle cells. Muscular dystrophies are divided into various groups of disorders depending upon the type of gene affected, the protein it produces, symptoms of the disease and progression of the disease.

More common forms of muscular dystrophy are Duchenne Muscular Dystrophy (DMD), Becker's Muscular Dystrophy (BMD) and Limb Girdle muscular dystrophy (LGMD). The less common forms are Facioscapulo Humeral dystrophy (FSHD), Congenital muscular dystrophy (CMD), Myotonic muscular dystrophy, Occulopharyngeal muscular dystrophy, distal muscular dystrophy, and Emery Dreiufuss muscular dystrophy.

**What are the causes?**

1.Fault in the genes

Muscular dystrophies are caused due to faults in various genes that make proteins required for the muscles. These proteins are part of the structure of the cell wall of the muscles. Fault in the genes causes production of faulty genes and weak walls of the muscles cells. Cells therefore are prone to damage very easily. Day to day activities can also damage the cells and cause progressive muscle weakness. Different proteins cause different muscular dystrophies.

| Type of muscular dystrophy | Gene involved | Protein formed |
|---|---|---|
| Duchenne muscle dystrophy | Dystrophin | Dystrophin |
| Becker Muscular Dystrophy | Dystrophin | Dystrophin |

| | | |
|---|---|---|
| Limb Girdle muscular dystrophy | CAPN3 (15q15.1), DYSF (2p13.2), SGCG (13q12.12), SGCA (17q21.33), SGCB (4q12), SGCD (5q33.3), TCAP (17q12), TRIM32 (9q33.1) etc | Dysferlins, Sarcoglycans, Calpains, |
| Facioscapulo Humeral dystrophy | Gene 4q35.2, 10qter, 18p11.32 (Dominant) | Double homeobox protein 4 (DUX4), D4Z4 |
| Congenital muscular dystrophy | lamininalpha-2,CollagenIV,POMT1,POMT2, FKTN,FKRP,LARGE, POMGNT1, andISPD | Merosin, Dystroglycan, Collagen |
| Myotonic muscular dystrophy | Type 1 - DMPK, Type 2 - ZNF9/CNBP | Myotonin protein kinase |
| Occulopharyngeal muscular dystrophy | 14q11.2 | Polyadenylate-binding protein |
| Emery Dreiufuss muscular dystrophy | Emerin Xq28 | Emerin |
| Myotonic Muscular Dystrophy | DMPK gene (Type 1), CNBP gene (Type 2) | Myotonic dystrophy protein kinase |

**2.Exhaustion of stem cells**

Although the muscles are damaged since birth body has a mechanism of repairing these damaged muscles. This repair is done through stem cells. Stem cells are the younger cells that are stored in muscles and can be used whenever new muscles cells are required. These are known as satellite cells. There is a limited source of these cells in the body. In the normal aging process these cells last longer and the weakness only sets in at an older age. But in muscular dystrophy the balance of damage and repair is lost and the cells are exhausted much earlier. Once these stem cells are completely exhausted the damaged muscle cannot be repaired further and that is when the symptoms become visible. Therefore although the disease is caused by faulty genes it progresses further due to lack of stem cells. Muscular dystrophy is therefore a stem cell disease.

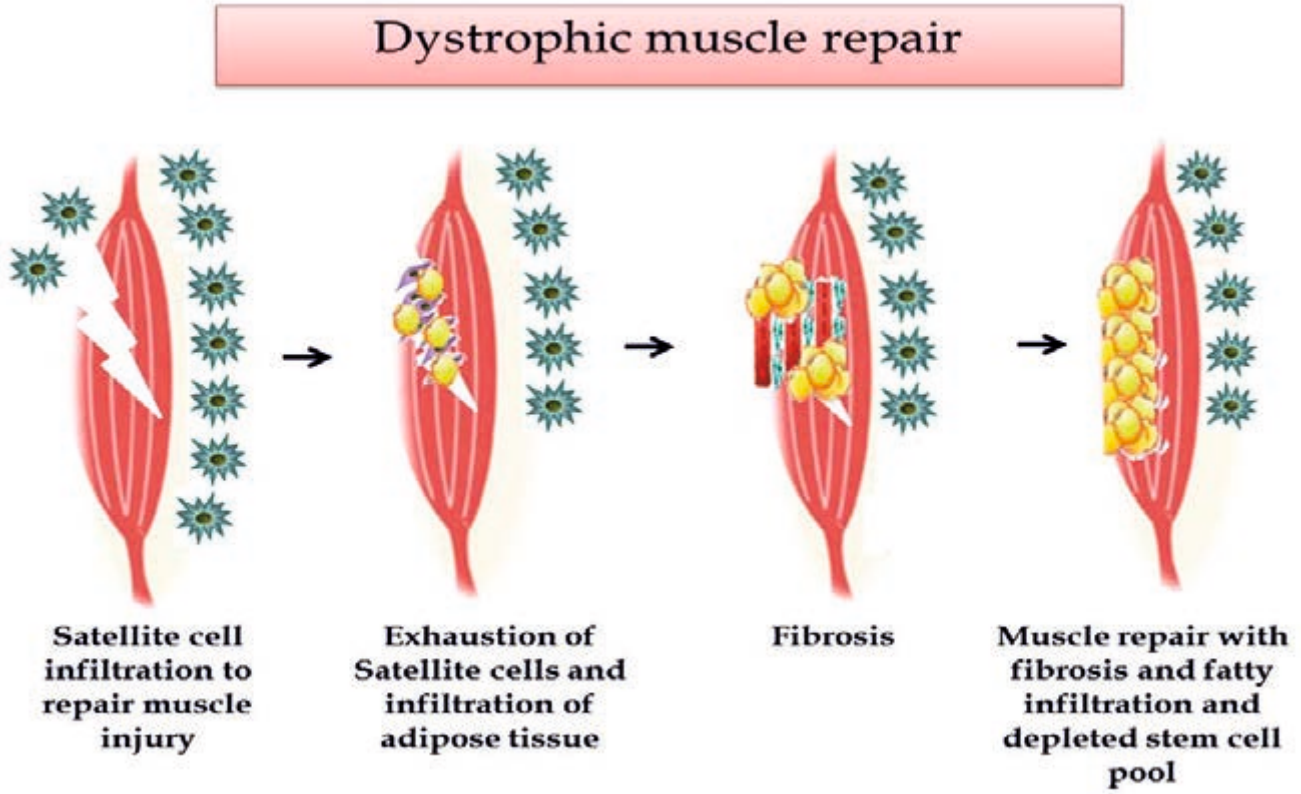



*Figure 1: Stem cell exhaustion causing muscular dystrophy*

**3. Effect of faulty proteins and muscle degeneration on different tissues in the body**

The faulty proteins are present form part of the muscles as well other tissues of the body like kidney, nerve cells, covering of nerve cells, retina etc. therefore these structures may also be damaged.

When the muscles are damaged due to lack of stem cells the faulty repair process gives rise to scar tissue. Therefore muscles are replaced with fibrous and fat tissue. Muscles therefore look big but have no strength. There are muscles in blood vessels also and therefore blood supply to the muscles is also poor in muscular dystrophies. Constant cell death leads to chronic inflammation and therefore the nerves supplying muscles also are damaged. Lack of activity leads to poor bone health. Heart is also a muscle and therefore is affected in this disease as well the heart function reduces as disease progresses and sometime may be the first to be damaged like that in Becker Muscular Dystrophy. Respiratory system is affected and therefore breathing is affected as well. Gastrointestinal system muscles are also affected causing problems like constipation due to poor movement of the gut muscles. In some types of dystrophies eye tissue is also impaired leading to symptoms like cataract.

**How is muscular dystrophy propagated from one generation to other?**

Muscular dystrophy can be caused by spontaneous mutations in the genes. Many times though these defects are hereditary i.e. inherited from family members and ancestors. There are fixed patterns in which the disease is carried forward from one generation to the other. Some dystrophies are caused due to fault in the genes that are present in the chromosomes which determine the gender of the person these are called X - linked inheritance patterns; while others are caused by genes from other chromosomes which are known as autosomal patterns of inheritance. In the process of inheritance some people develop

the disease where as some carry the faulty gene but don't develop the symptoms such individuals are known as carriers. Carries can propagate the disease to their children but don't develop the disease themselves.

| Type of muscular dystrophy | Inheritance pattern |
|---|---|
| Duchenne muscle dystrophy | X – linked recessive |
| Becker muscle dystrophy | X – linked recessive |
| Limb girdle muscle dystrophy (Type 2 A – Q) | Autosomal recessive |
| Limb girdle muscle dystrophy (Type 1 A – H) | Autosomal dominant |
| Facioscapulo humeral dystrophy | Autosomal dominant (some types) and recessive both (some types) |
| Congenital muscular dystrophy | Autosomal recessive |
| Myotonic muscular dystrophy | Autosomal dominant |
| Occulopharyngeal muscular dystrophy | Autosomal recessive (some types) and dominant (some types) |
| Emery Dreiufuss muscular dystrophy | X – linked recessive (some types) and Autosomal recessive (some types) |
| Distal muscular dystrophy | Autosomal recessive |
| Myotonic dystrophy | Autosomal dominant |

the disease where as some carry the faulty gene but don't develop the symptoms such individuals are known as carriers. Carries can propagate the disease to their children but don't develop the disease themselves.

**X – Linked pattern of inheritance:**

1.Recessive inheritance pattern

The gene responsible in this is located on the X-chromosome. Recessive pattern means the gene will cause the disease in absence of the normal gene counterpart. Females have two X-chromosomes whereas males have One X chromosome and one Y chromosome. Therefore the faulty gene can be present in both boys and girls. However boys develop the disease but girls don't. Girls may however carry the abnormal gene and be a carrier therefore their sons may be at the risk of developing this disease.

**Autosomal pattern of inheritance**

Autosomal patterns of inheritance are independent of the sex chromosomes. Therefore, both males and females are at an equal risk of being affected with the disease. There are two chromosomes inherited from each parent. The genes on either of these chromosomes are identically placed. For any

gene therefore, there is a counterpart on the other chromosomes. Based on this there are two different patterns in autosomal inheritance.

Autosomal recessive: Refers to an inheritance pattern where the faulty gene cannot express itself on its own in presence of a normal gene counterpart from the other chromosome.

Autosomal dominant:Refers to an inheritance pattern where the faulty gene expresses itself irrespective of its counterpart from the other chromosome.

**Symptoms of muscular dystrophy**

Muscular dystrophy is a group of disorders that can be seen in patients of varies age groups ranging from infants to adults. It is seen both in males as well as females. All the muscular dystrophies are progressive in nature. The most characteristic symptom of the disease is muscle weakness. The pattern of muscle weakness and rate of disease progression varies depending on the type of muscular dystrophy.

The most common of the muscular dystrophies is Duchenne Muscular Dystrophy (DMD) which occurs in the early childhood and significantly reduces the lifespan of the children.

Symptoms of DMD can be divided in 5 stages of the disease, which are based on the ability of the children to walk. The 5 stages are as follows:

- Stage 1: Pre-symptomatic (0-4 years)
- Stage 2: Early ambulatory stage (4-8 years)
- Stage 3: Late ambulatory stage (8 - 13 years)
- Stage 4: Early non- ambulatory stage (13 to 16 years)
- Stage 5: Late non- ambulatory stage (16 years onwards)

**Pre-symptomatic (0-4 years)**

There are no obvious symptoms present. Sometimes parents may notice swollen calves, some slowness in performing physical activities as compared to other children, the motor milestones like standing and walking may be delayed slightly and serum level of enzyme creatine phsopho kinase (CPK) may be increased in these children.

**Early ambulatory stage (4-8 years)**

–Frequent falls while walking, running or jumping

- Calf psuedohypertrophy
- Child gets tired soon, and cannot perform physical activities as well as or as long as his peer
- Difficulty to get up from the floor, positive gower's sign
- Child need to hold the railing while climbing up the stairs
- There may be over straightening of the knees observed while walking down the slope or climbing down the stairs
- Poor balance in standing and walking
- Waddling
- Toe walking
- Increased arching of the lower back or posterior trunk lean to keep maintain balance while walking
- Some children may also have low IQ and some intellectual disability

**Late ambulatory stage**

- Increased falls due twisting of ankle and buckling of the knee while standing or walking.
- Inability to rise from the floor with or without support
- Inability to climb stairs with or without

support

- Inability to get up from the chair with or without support
- Poor balance while standing
- Child gets tired easily while performing the overhead tasks with upper limbs.
- Child is able to walk only with support or with extreme arching of the back, walking up on his toes and keeping the legs wide apart.
- Child may lurch on one side while standing, walking and sitting. This lurch or curvature may disappear on lying down.
- Increased contracture keeping the limbs in a fixed position even when moved passively by others
- Palpitations
- Shortness of breath while performing routine tasks
- Constipation and lack of appetite

**Early non- ambulatory stage**

- Inability to walk
- Poor sitting balance
- Inability to perform overhead activities
- Limited upper extremity movement
- Inability to come to sitting from lying on the back
- Inability lift the head up while lying on the back
- Inability to do upper limb activities while sitting unsupported
- Leaning on one side for stability (Figure 2.7).
- Leaning on the opposite side during upper limb function to substitute for shoulder girdle weakness in arm lifting
- Leaning backward while lifting the arms up
- Leading with head to compensate for trunk weakness
- Catching fingers in the mouth for overhead activities
- Tightness develops in
- Elbow flexors and pronators (Figure 2.8)
- Wrist and finger flexors (Figure 2.9)
- Curved spine (scoliosis) ( Figure 2.10)
- Constipation
- Severe lack of appetite and reduced food intake
- Difficulty of breathing while performing physical activities
- Left ventricular dysfunction

**Late non- ambulatory stage**

- Inability to use the arms for any functional activity
- Inability to sit
- Increased curvature of the spine
- Severe deformities
- Severe heart dysfunction
- Difficulty to breath even at rest
- Severe constipation
- Loss of appetite, minimal food intake
- Difficulty in swallowing
- Poor ability to cough
- Requirement of nasal tube for feeding (Figure 2.11)
- Requirement of external ventilatory assistance (Figure 2.12)

| **Becker Muscular Dystrophy** | | |
|---|---|---|
| **What It Is** | **Common Symptoms** | **How It Develops** |
| Also caused by a deficiency of dystrophin, and with symptoms similar to those of DMD, Becker can progress slowly or quickly. | Patients with Becker MD may:<br>• Walk on their tiptoes<br>• Fall down a lot<br>• Have difficulty rising from the floor<br>• Have cramping in their muscles<br>• They exhibit cardiac involvement earlier in the disease with presence of cardiomyopathy | Becker MD appears primarily in males between ages 11 and 25. Some people may never need to use a wheelchair, while others lose the ability to walk during their teens, mid-30s, or later. |
| **Myotonic** | | |
| **What It Is** | **Common Symptoms** | **How It Develops** |
| The most common adult form of MD, myotonic MD appears in two forms, type 1 and type 2. Type 1 is more common | Myotonic MD causes an inability to relax muscles following a sudden contraction. Other symptoms include<br>• Long, thin face and neck<br>• Swallowing difficulties<br>• Drooping eyelids, cataracts, and other vision problems | Myotonic MD affects both men and women between ages 20 and 30. |

| | | |
|---|---|---|
| | • Baldness at the front of the scalp<br>• Weight loss<br>• Increased sweating<br>• Drowsiness<br>• Heart problems that may lead to death during the 30s or 40s<br>• Irregular menstrual periods<br>• Infertility<br>• Impotence | |
| **Congenital** | | |
| **What It Is** | **Common Symptoms** | **How It Develops** |
| Majority of the cases with congenital MD are caused by a defect in the protein merosin, which surrounds muscle fibers. When caused by defects in other proteins, this type of MD may also affect the central nervous system. | People with congenital MD may:<br>• Have problems with motor function and muscle control that appear at birth or during infancy<br>• Develop chronic shortening of muscles or tendons around joints, which prevents joints from moving freely<br>• Develop scoliosis (curvature of the spine)<br>• Have trouble breathing and swallowing<br>• Have foot deformities<br>• Have intellectual disabilities | This form of MD appears at birth or by age 2. Congenital means "present from birth." Congenital MD affects both boys and girls, who often require support to sit or stand and may never learn to walk. Some patients die in infancy, but others live into adulthood with only mild disability. |
| **Emery-Dreifuss** | | |
| **What It Is** | **Common Symptoms** | **How It Develops** |
| Affecting boys primarily, the two forms of Emery-Dreifuss MD are caused by defects in the proteins that surround the nucleus in cells. | Weakness begins in the upper arm and lower leg muscles. People with this form may also<br>• Develop chronic shortening of muscles around joints (preventing them from moving freely), in the spine, ankles, knees, elbows, and back of the neck<br>• Have elbows locked in a flexed position<br>• Develop shoulder deterioration<br>• Have a rigid spine<br>• Walk on their toes<br>• Experience mild weakness in their facial muscles | Symptoms usually begin by age 10 but can appear in patients up to their mid-20s. People with this form often develop heart problems by age 30, and they may die in mid-adulthood from progressive pulmonary or cardiac failure. |

| Facioscapulohumeral | | |
|---|---|---|
| What It Is | Common Symptoms | How It Develops |
| FSHD refers to the areas affected: the face (facio), the shoulders (scapulo), and the upper arms (humeral). Researchers don't know what gene causes FSHD. They do know where the defect occurs and that it affects specific muscle groups.6 | FSHD MD often appears first in the eyes (difficulty in opening and shutting) and mouth (inability to smile or pucker).7 Other symptoms may include<br>• Muscle wasting that causes shoulders to appear slanted and shoulder blades to appear "winged"<br>• Impaired reflexes only at the biceps and triceps<br>• Trouble swallowing, chewing, or speaking<br>• Hearing problems<br>• Swayback curve in the spine, called lordosis | FSHD affects teen boys and girls typically but may occur as late as age 40. Most individuals have a normal life span, but symptoms can vary from mild to severely disabling. |
| **Limb-girdle** | | |
| **What It Is** | **Common Symptoms** | **How It Develops** |
| Affecting both males and females, different types of limb-girdle are caused by different gene mutations. Patients with limb-girdle inherit a defective gene from either parent, or, in the more severe form, the same defective gene from both parents. | Patients with limb-girdle MD may:<br>• First develop weakness around the hips, which then spreads to the shoulders, legs, and neck<br>• Fall down a lot<br>• Have trouble rising from chairs, climbing stairs, or carrying things<br>• Waddle when they walk<br>• Have a rigid spine | This form of MD can appear in childhood but most often appears in adolescence or young adulthood. Limb-girdle can progress quickly or slowly, but most patients become severely disabled (with muscle damage and inability to walk) within 20 years of developing the disease. |

| **Distal** | | |
|---|---|---|
| **What It Is** | **Common Symptoms** | **How It Develops** |
| Distal MDs refer to a group of diseases that affect the muscles of the forearms, hands, lower legs, and feet. They are caused by defects in the protein dysferlin5 and can occur in both men and women. | Distal MD may cause:<br>• Inability to perform hand movements<br>• Difficulty extending fingers<br>• Trouble walking and climbing stairs<br>• Inability to hop or stand on the heels | This form typically appears between ages 40 and 60. Distal MD is less severe and progresses more slowly than other forms of MD, but it can spread to other muscles. Patients may eventually need a ventilator. |
| **Oculopharyngeal** | | |
| **What It Is** | **Common Symptoms** | **How It Develops** |
| This form occurs in both men and women, and it can be mild or severe. It is caused by a defect in a protein that binds to molecules that help make other proteins. It is common among Americans of French-Canadian descent, Jewish Ashkenazi, and Hispanics from the Southwest region.8 | Oculopharyngeal MD may cause:<br>• Drooping eyelids and other vision problems<br>• Swallowing problems<br>• Muscle wasting and weakness in the neck, shoulders, and sometimes limbs<br>• Heart problems | This form of MD typically appears in a person's 40s or 50s. Some people will eventually lose their ability to walk. |

**How to diagnose Muscular Dystrophy?**

Diagnosis of muscular dystrophy is done on the basis of clinical features, blood tests, electromyogram and confirmed by muscle biopsy, Musculoskeletal MRI or genetic testing. Genetic testing helps to give the most accurate diagnosis of the type of muscular dystrophy.

**Clinical examination:**

Clinician takes into consideration the history and onset of symptoms. Enquires about presence of the disease in family members or ancestors. A thorough clinical examination is

carried out to check the muscle strength of all the muscles, shortness of muscles and loss of function of the child. Cardiac and respiratory abilities are evaluated. Based on the performance of these tasks the clinician refers for further investigative tests like Serum CPK testing, muscle biopsy, EMG, musculoskeletal MRI, ECG and 2D-echo cardiography.

**Serum CPK testing:**

CPK is an enzyme found in muscles, brain and heart. This enzyme is released in blood when the muscles are injured. Therefore the levels of this enzyme in the blood of patients of muscular dystrophy are increased.

**Normal range of total serum CPK:**

- Male: 38 - 174 units/L
- Female: 96 - 140 units/L

**Muscle biopsy**

A muscle biopsy is a tool to diagnose neuromuscular diseases by viewing the muscle cell structure under microscope. Depending upon where the symptoms are observed a needle is inserted or a small piece of muscle is excised. The muscles will be observed under miscroscope for specific features suggestive dystrophy. Your neurologist and pathologist will give a report to confirm presence of muscular dystrophy.

**Electromyogram (EMG) and Nerve conduction velocity (NCV) testing**

For the diagnosis of muscular dystrophy Needle EMG is required. Needle EMG is an invasive technique.A careful clinical evaluation along with findings from other tests is required in case of muscular dystrophy. The graph on the paper will show the muscle recruitment which can be interpreted by a neurologist to differentiate between normal and dystrophic pattern.

**Musculoskeletal Magnetic Resonance Imaging (MRI-MSK)**

MRI-MSK is a non-invasive imaging technique used to identify any structural abnormalities in the body. MRI uses a powerful magnetic field, radio frequency pulses and a computer to produce detailed pictures of organs, soft tissues, bone and virtually all other internal body structures. MRI does not use any X-ray (Ionizing radiation).

MRI images help identify the amount of fibrous tissue and muscles in a given body part. Based on this proportion and distribution and appearance of muscles and fibrous tissue, radiologists will be able to assess the severity of the disease.

Advantages: It is a non-invasive and painless investigation. It reveals a detailed muscle involvement with regards to number of muscles affected, extent and severity of damage. It also gives information about the amount of fatty infiltration in the muscles. It can also be used as a monitoring tool.

**Electrocardiograph (EKG/ECG)**

An electrocardiogram measures the electrical activity of the heart muscles and is one of simplest and fastest ways of evaluating heart muscles. EKG is not a diagnostic test in MD however it is essential to prevent and treat any heart related complications.

**2D-Echocariography**

2D-echcardiography like ECG is not a diagnostic test in MD however it is essential to prevent any heart related complications.

**Genetic testing**

Genetic testing is the gold standard to identify MDs. Genetic testing identifies the faulty genes. It is performed by drawing blood which is then analyzed by array of tests to find out the faulty gene.

## Physiotherapy management of muscular dystrophy

### Aims of Physiotherapy

- To assess muscle strength, joint mobility, deformities and functional abilities.
- To determine the causes of deformities and prevent deformities.
- To suggest supportive physical aids when required.
- To correct the physical impairments to the best of the abilities of the patient.
- To educate the patients, parents / caregivers regarding the disease and its outcome.
- To guide the patients and parents regarding care and prevention of the deformities.
- To maintain independent walking as long as possible.
- To encourage daily exercises.
- To prevent frequent falls, fractures, pressure sores and stiff joints.
- To improve cardio-respiratory endurance of the patient.
- To motivate them to overcome complications and improve their quality of life.
- To plan an appropriate treatment program in co-ordination with other professionals of the rehabilitation team.

### Basic principles of exercise prescription in muscular dystrophy

Exercise prescription in muscular dystrophy differs greatly as compared to exercise prescription in typically developing individuals. Muscular dystrophy is a primary muscular pathology with progressive degeneration. This degeneration is triggered even during the regular muscle contractions. However, not using these muscles does not preserve them longer. The muscles unless used appropriately undergo atrophy (reduction in the total number of muscle fibers) in addition to the dystrophic (reduction in the number of muscle fibers and replacement with fibrous non-contractile tissue) changes caused by the disease. Therefore regular low intensity graded and supervised exercise is required to be performed by individuals with muscular dystrophy. This exercise is prescribed on the basis of following principles.

### Muscle plasticity

There are two types of muscles in our body, one type of muscle fibers are responsible for producing burst of strong contraction very quickly. These muscle fibers are more prone to damage during contraction. The other type of muscle fiber produces a slow contraction of smaller amplitude but can sustain it for a long time (Type I). These muscle fibers are less prone to damage during contraction (Type II). There is certain percentage of both types of fibers in any muscle of our body. Depending upon the function of the muscle, the proportion of fibers varies in different muscles of the body. This proportion also varies depending on how the muscles are trained. Although a muscle needs both type of fibers, with targeted training proportion of one type can be increased. This is called as muscle plasticity.

### Regressive resistive exercises

For the treatment of individuals with muscular dystrophy regressive overload is used. Regressive overload means over a period of time resistance used to train the muscles is reduced. If the resistance is kept the same then the muscles will undergo more damage. Therefore it is reduced as there is progressive muscle weakness. Principle of overload means that the muscle should be stimulated to perform beyond its current

capacity. This can be achieved using increased resistance provided to the muscle or higher number of repetitions with the same or lesser resistance. In muscular dystrophy higher repetitions are performed. Higher repetitions facilitate the growth of Type II fibers.

**Facilitating concentric contractions and preventing eccentric contractions**

Eccentric contractions are the contractions where the muscles shorten as they contract. These contractions are used to bring about movement in the body and are less damaging to the muscle. Eccentric (lengthening) contractions are the contractions where the muscle is contracting but lengthens while doing so. These are useful to control the speed and extent of the movement but bring about more damage to the muscles. Eccentric contraction of the muscles is unavoidable in day to day activities as these are the contractions that are required to control any body movement performed against gravity. However the exercise training program should be designed to prevent such contractions and facilitate more of concentric (shortening) contractions. The main principle of training in muscular dystrophy is optimizing muscle strength and endurance.

**Preventing disuse**

Disuse is characterized by the difference between the physical activity that can be performed and the actual physical activities of the individual. It is the difference between the physical capabilities and physical performance. Due to the muscle weakness, increased effort to carry out an activity and fear of falls, individuals with muscular dystrophy are physically less active than others. This early decline in the physical activity speeds the process of muscle wasting and atrophy, thereby muscle weakness. This decline is physical activity must be prevented with the help of use of timely consistent tailored physical exercises.

**Preventing Fibrosis**

One of the most important principles in muscular dystrophy is to prevent fibrosis. Exercises aims at moving the joints through complete range, stretching the muscles as well as other tissues in the body are important in preventing the fibrosis. Active exercises also help prevent fibrosis in the muscles. A combination of regular moderate intensity active exercises within the limits of fatigue and passive stretching should be prescribed in muscular dystrophy.

**Physiological benefits of exercise**

All of these exercises have different effects of different body systems as explained below

*Musculoskeletal system*

- Increased density of the bone minerals,
- Improved absorption of calcium and phosphate in the bones
- Bone growth facilitation
- Maintenance and replacement of lubricating fluid in the joints
- Increased number of muscle fibers
- Increased size of the muscle fibers
- Muscle growth stimulation
- Improved blood supply to the bones and the muscles
- Reduced muscle fatigue

*Cardio-respiratory*

- Increased blood pressure as immediate response exercises improving the blood circulation
- Reduction in the blood pressure as a late response to exercises reducing the cardiac stress
- Improved capacity of the heart to pump blood

- Improved return of the blood from extremities to heart preventing stagnation of the blood in extremities
- Improved capacity of the lungs to breath in the air
- Improved oxygenation of the blood
- Improved clearance of carbon dioxide from the blood
- Ability to breath air out more forcefully

*Nervous system*

- Improved blood supply to the brain
- Improved memory and other complex cognitive functions
- Stimulation to form new blood vessels
- Antidepressant effects preventing depression, anxiety and other negative psychological responses

*Effects on Fibrosis:*

- Reduction of fibrotic processes by up to 50% improving pliability, extensibility thereby contractile strength of the skeletal muscles; reducing the joint stiffness and cardiac muscle fibrosis.
- Blood vessels and blood circulation:
- Formation of new blood vessels
- Improved connections and network of blood vessels
- Improved circulation to the deep muscles of the body, brain, lungs, liver and kidney
- Improved clearance of toxic waste substances from the blood

**Exercises prescription in different stages of the disease**

Stage 1: Pre-symptomatic (0-4 years)

In the pre-symptomatic stage very rarely the muscles undergo weakness. The main aim of physiotherapy is to facilitate motor learning and strengthening muscles with endurance exercises to achieve greater muscle plasticity.

*Type of Exercise:*

*Moderate intensity resisted muscle strengthening exercises*

*Moderate intensity cardiovascular endurance training exercises*

*Muscle endurance training exercises*

**Stage 2: Early ambulatory stage (4-8 years)**

In the early ambulatory phase children are still able to walk and can carry out all the physical activities independently with difficulty or with support of external objects. Functional deficits are minimal and the compliance of patients and caregivers to exercise may be limited because of the same. However exercise in this phase is very important. Parents and care givers must make sure that they comply with the instructions and prescription of exercise as suggested by their rehabilitation professionals. Table1. Summarizes the muscles that undergo weakness, tightness and resulting postural compensations.

| Muscles that undergo weakness | Muscles that undergo tightness | Postural compensations |
|---|---|---|
| Hip extensors | Tendoachilis (Calf muscles) | Increased arching of the back while standing and walking (lumbar lordosis) |
| Ankle dorsiflexors | Hip Flexors | Walking on toes |
| Hip abductors | Lower abdominals | Stabilizing the knee with hands while getting up from the chair and floor |

| | | |
|---|---|---|
| Hip adductors | | Unable to control descent while sitting down |
| Abdominal muscles | | |
| Neck flexors | | |
| Shoulder flexors | | |
| Shoulder abductors | | |
| Shoulder extensors and depressors | | |
| Elbow extensors | | |

**Goals of Physiotherapy:**

- To prolong independent walking as long as possible.
- To maintain muscle strength.
- To prevent frequent falls and fractures.
- To prevent bed rest for long period of time.
- To continue exercises (shown in chapter 5).
- Type of exercises:
- Active exercise with moderate resistance and higher repetitions
- Active and passive stretching for the calves, abdominal muscles and hip flexors
- Movement and postural corrective exercises
- Moderate intensity cardiorespiratory endurance training exercises

**Stretching exercises:**

Calf stretching Passive



*Figure 2*



Place one hand under the knee and hold the heel in your other hand. Gently bend the ankle upward by pushing against the sole of the foot with the forearm.

**Hip flexor stretching Passive**

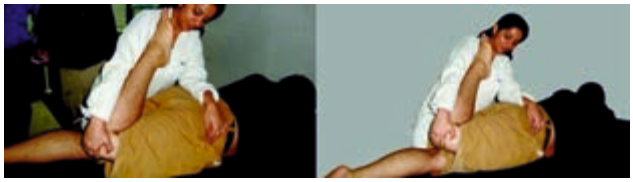


*Figure 3 : Passive stretching of the hip flexors*



*Fig. 4: Passive stretching of the hip flexors*

Alternatively to stretch both the hips joints together, ask the individual to lie on his/ her tummy. Keep a below both the knees to lift knees above the pelvic height. With hip flexion tightness the buttocks will raise up. Push it down gently keeping the knees bent till gentle stretch is felt.

Place one hand on one knee and with the other hand bend the hip and knee of the other leg as if trying to touch chest. Press down the knee of the other leg.



*Figure 5: Passive stretching of the hip flexors*

**Strengthening exercises:**

It is important to remember that the principle of strengthening in Muscular Dystrophies is regressive resistive exercises. Which means, that the resistance should be graded down as the weakness progresses. Therefore early in the disease resistive and active exercises can be performed however as the disease progresses exercises change to active assisted, facilitatory, restorative or compensatory and eventually passive exercises. While applying strengthening exercises it is important to note that the resistance should be mild to moderate with higher repetitions. Avoid eccentric contractions and facilitation concentric, isometric contractions. Figures given below are examples of how the exercises can be graded down.



*Fig. 5 Active exercises for hip strengthening*



*Figure 6: Active assisted exercises for abdominal muscles*

Apart from strengthening other components need to be also incorporated like balance training



*Figure 7: Balance training exercises*



*Figure 8: Balance training exercises*

**Stage 3: Late ambulatory phase**

In the late ambulatory phase the postural compensations are more enhanced and the tightness of the muscles may also accelerate. Children walk with severe arching of the back, waddling from side to side due to weakness of hip muscles and pronounced toe walking. The stability in standing and walking is very poor and they may fall even with a slightest push. Table 4 summarizes the muscle weakness, tightness and postural compensations in this phase.

**Goals of Physiotherapy:**

- To prevent any falls or fractures.
- To maintain walking with long leg calipers.
- To maintain muscle length.
- To prevent any chest infection.
- To conserve energy.
- To continue daily activities using special equipments like use of wheelchair for covering long distances for saving energy

Table 4: Table describing the muscles undergoing weakness, tightness and resultant postural compensations in Stage III

| **Muscles that undergo weakness** | **Muscles that undergo tightness** | **Postural compensations** |
|---|---|---|
| Neck flexors | Tendoachilis (Calf muscles) | Severe arching of the back while standing and walking (lumbar lordosis) |
| Abdominals | Hip Flexors | Walking on toes and minimal waddling |
| Shoulder flexors | Lower abdominals | Stabilizing the knee with hands while getting up from the chair and floor |
| Shoulder abductors | Hamstrings | Unable to control descent while sitting down |
| Shoulder depressers | Ilio-tibial band | Foot turned inwards and downwards |
| Shoulder extensors | | Slouched back while sitting |
| Elbow extensors | | Walking with feet apart |
| Hip extensors spine | | Minimal sideways curvature of the |
| Hip abductors | | |

| | | |
|---|---|---|
| Hip adductors | | |
| Ankle dorsiflexors | | |
| Ankle everters | | |
| Knee extensors | | |

**Type of exercise:**

- Active exercise with moderate resistance and higher repetitions (against gravity)
- Gait retraining with splints
- Active and passive stretching for the calves, abdominal muscles, hamstings, iliotibial band and hip flexors
- Movement and postural corrections
- Moderate intensity cardio-respiratory endurance training exercises
- Stretching exercises:
- Hamstrings stretching
- Iliotibial band stretching

**Strengthening exercises:**

In the late ambulatory phase the exercises as explained above should be continued. Some of these exercises may require assistance. Gait re-training is the key in this stage in.

Due to weakness of knee extensors, quadriceps, children tend to buckle in their knees while walking. This causes frequent falls and fear avoidance of walking. Therefore depending upon the weakness and tightness of the muscles, assistive devices like knee or ankle splints and modified footwear may be prescribed by your physiotherapist.

Some examples of the splints and shoes are:

Push knee splint: It is a simple splint that prevents knee from buckling and bending forward. (Figure 9)

High boots with posterior steel shanks: These are stiff boots that prevent in turning and down turning of the feet. ( figure 10)

Below knee brace with an outer T-strap: If there is severe tightness of the muscles causing severe in turning of the feet which cannot be stretched to neutral position, then a special shoes with a an above ankle strap and outer strap at the level of ankle bones is prescribed. (Figure 11)

Elbow splint: Elbow splint is like push knee splint that prevents bucking of the elbow when putting weight on the arms. This can be used while performing bed exercises as well as walking. (Figure 12)

For Neck, Shoulder, Elbow, Wrist and hand perform the active exercises as described earlier. Hip exercises can be performed using a suspension device in the gravity eliminated plane that provides assistance. In the late ambulatory phase muscles are still stronger than required for a movement in gravity eliminated plane. Your therapist may add resistance using therapeutic resistance bands.



*Figure 9: Push knee orthosis*



*Figure 10: High boots*



*Figure 11: Below knee braces*



*Figure 11: Elbow splints*

In this stage the weakness is progressed and active assisted exercises in the form of suspension therapy can be given.



*Figure 12: Suspension exercises for the hip*



*Figure 13: Suspesion exercises with resistance*

At the same time functional exercises like bed mobility training and weight bearing exercises like crawling and standing should be facilitated.



*Figure 14: Weight bearing exercises*

**Gait retraining**

Gait training is an important component in this phase as the children are likely to lose their ability to walk or stand due to muscle weakness mainly quadriceps and gluteus. It is important to train the children with compensatory strategies using splints and other supportive devices.



*Figure 15: Gait training exercises*



*Figure 16: Progression of gait training*

**Stage 4: Early Non-ambulatory stage**

In the non-ambulatory stage most of the muscles of the big joints like shoulder and hips may show muscle strength below functional level i.e. inability to perform movements against gravity. The exercises in this stage are targeted at the maintenance of muscle strength. Exercises with mild resistance in gravity eliminate plane and active assisted exercises against gravity will be prescribed. Children also show presence of severe postural deviations and start developing scoliosis and spinal deviations. Therefore physiotherapists may prescribe spinal jacket to prevent further deterioration. Cardio-respiratory complications are prevalent in this stage, therefore attention needs to be given to the cardio-respiratory endurance training.

**Goals:**

- To prevent deterioration of muscle by exercising.
- To prevent contractures and deformities.
- Deep breathing exercises and use of incentive spirometer for improving chest volumes.
- To prevent scoliosis and if it develops, prescription of spinal jacket or spinal support.
- To assess requirement of assistive breathing such as Bi-Pap.
- To continue doing daily activities using special equipments like assistive devices.
- Type of exercise:
- Active assisted exercises with higher repetitions (against gravity)
- Suspension exercises (in gravity eliminated plane) with moderate resistance and higher repetitions
- Active and passive stretching for the calves, abdominal muscles, hamstings, iliotibial band, hip flexors, elbow flexors, pronators, and wrist and finger flexors.
- Movement and postural corrections with supportive devices
- Moderate intensity cardiorespiratory endurance training exercises

Table V: Showing Muscles that undergo weakness, tightness, Postural compensations

| **Muscles that undergo weakness** | **Muscles that undergo tightness** | **Postural compensations** |
|---|---|---|
| Neck flexors | Tendoachilis (Calf muscles) | Severe arching of the back while standing and walking (lumbar lordosis) |
| Abdominals | Hip Flexors | Foot turned inwards and downwards |
| Shoulder flexors | Lower abdominals | Bent knee attitude / tightness / contracture |

| | | |
|---|---|---|
| Shoulder abductors | Hamstrings | Legs fall outwards when lying straight on the back |
| Shoulder depressers | Ilio-tibial band | Hips are bent and rise up when an individual is lying facing down |
| Shoulder extensors | Pyriformis | Severe slouching while sitting |
| Elbow extensors | Elbow flexors | Prominent sideways curvature of the spine |
| Hip extensors | Pronators | Legs fall out in lying position with knees bent |
| Hip abductors | Wrist flexors | The alignment of shin bones and thighs is changed and shin bones are turned outwards |
| Hip adductors | Finger flexors | |
| Knee Extensors | Hip external rotators and abductors | |
| Ankle dorsiflexors | | |
| Ankle everters | | |

**Stretching exercises:**

*Elbow Flexors stretching:*

Passive: Rest the arm of the patient on the pillow. The stretching can performed either in sitting or lying down. Gently straighten the elbow till a firm resistance is felt maintain. Maintain the position for 30 seconds.

**Pronator stretching:**

In the same position as hat of elbow stretching turn the arm to face the palm upwards, stretch till you feel a distinct resistance. Hold for 30 seconds.

**Wrist and finger flexors stretching:**

Rest the arm on pillow keeping the elbow and finger straight, push the wrist palm of the patient upwards. Hold for 30 seconds. If the muscles are very tight the same can be performed with fists closed.

**Passive positioning:**

The most important therapeutic component in this stage is passive positioning. As the children lose their walking ability, weight bearing on the lower extremity reduces which can enhance the process of thinning of bones and bone porosity. Porous bones become brittle and are at risk of fracturing with minimal trauma. Sometimes while lifting the child or transferring the child from one place to another the bones can suffer fracture (stress fracture). With use of steroids the bone density reduces and such bones are at a greater risk of fractures. Increased body weight is another contributing factor for such fractures. Therefore passive positioning and weight bearing is required to facilitate increase in the bone density.

Weight bearing on the lower extremities can be achieved in various positions like kneeling on the knees, quadruped and standing. All these positions can be achieved with varying degrees of support, with the help of vestibular ball while performing quadruped and kneeling (Figure 10.97). For standing suspended standing can be performed or a child can be support on standing board with the help chest,

abdominal and knee straps (Figure 10.98).

**Facial Exercises**

As the muscles of the face and swallowing also undergo weakness in this stage, facial exercises for facial muscles, tongue execises are recommended

**Stage 5: Late non-ambulatory**

In late ambulatory stage the physiotherapy care is more palliative. Most of the skeletal muscles have poor muscle strength and many joints may have undergone contractures.

**Goals:**

To continue exercises to maintain muscle strength.

To prevent worsening of the contractures and deformities.

Deep breathing exercises and use of incentive spirometer for improving chest volume.

Prescription of spinal jacket to improve posture and help in breathing.

To assess requirement of assistive breathing such as BiPap.

To continue doing daily activities using special equipment.

To ensure proper to prevent pressure sores.

To clear the chest of secretions.

**Types of exercises:**

Passive stretching

Passive positioning

Suspension exercises (Active assisted / passive)

Cardio-respiratory endurance

Secretion clearance exercises

It is of immense importance to perform regular physiotherapy exercises from early stages in muscular dystrophy to prevent rapid deterioration and maintain muscle function for as long as possible. Rehabilitation of muscular dystrophy is a multidisciplinary approach where the family members perform a very important role. Awareness of the family members towards rehabilitation ensures good and timely rehab for the patients. In later stages when the exercises cannot be performed actively passive exercises are required to be performed passively by the family members and caretakers.

## Aquatic therapy management of muscular dystrophy

Aquatic therapy is one of the most widely used rehabilitative techniques for muscular dystrophy. Aquatic therapy means making use of different physical and chemical properties of water to facilitate functional recovery, independence, prevent complications and slow down the damage to the muscles in individuals with muscular dystrophy. Aquatic therapy is defined by The Aquatic Therapy and Rehabilitation Institute as "The use of water and specifically designed activity by qualified personnel to aid in the restoration, extension, maintenance and quality of function for persons with acute, transient, or chronic disabilities, syndromes or diseases".

**Benefits of aquatic therapy in Muscular dystrophy**

- Reduce the work of the muscles thereby preventing accelerated muscle damage.
- It prevents the eccentric contractions of the muscles while performing various movements and therefore helps reduce the muscle damage.
- It improves the blood supply to the deeper muscles and helps to slow down the fibrotic processes.
- It provides a higher degree of freedom of

movement which helps in boosting the morale, move the joints through their full range preventing secondary complications like muscle tightness, contractures and cardio-respiratory complications.

- It has a distinct physiological mood enhancing effect which may prevent negative emotional responses in individuals with muscular dystrophy.
- There is also improved blood supply to the brain improving the cognition in some cases of muscular dystrophy with co-morbid intellectual disability.
- In the later stages of the disease when the spinal deformities set in water environment provides freedom to unload and stretch the spine preventing some of the detrimental effects of the same.
- Aquatic therapy also improves respiratory capacity and cardiovascular endurance.
- Aquatic therapy and warm water exercises have pain reducing effect while stretching the muscle contractures, it also makes the muscles more pliable and easy to stretch.
- Increased blood supply, hydrostatic pressure of the water, improved endorphins and serotonin levels also helps to improve appetite and bowel movements. In the later stages of the disease it can prevent complications like severe constipation.
- Aquatic exercises help reduce sleep disturbances.

**Properties of water**

*Buoyancy*

Buoyancy is the upward push that water exerts on the immersed body. It is responsible for the weightlessness experienced in water. Deeper the immersion greater is the weightlessness. If the child is immersed till umbilical level then the effective weight on the legs is reduced by 50% and when immersed chest deep it is reduced by 75%.

Weightlessness is a very important phenomenon for therapeutic benefit. Weight bearing joints like ankle, knee and hip joints can be offloaded in water which helps reducing the stiffness of the muscles. Therapeutically this can be used to relieve the compressive forces on the joints and reduce the lengthening contractions (eccentric work) of the muscles. These lengthening contractions bring about accelerated muscle damage and therefore the exercises performed in water can give strength benefits without the harmful muscle contractions.

*Hydrostatic Pressure*

Is the pressure exerted by water and depends upon the density as well as the depth of immersion. The deeper we immerse the child the greater is the force. This is particularly helpful for reducing the swelling and providing passive relaxation through deep pressure. Hydrostatic pressure is also important to push the blood from the legs and thighs up increasing the blood returning to the heart. This also helps in increasing blood supply to the brain and therefore cognitive functions. The increased blood supply to heart is important for good cardiovascular health.

*Density*

Water is thicker or denser than the air. The density of water is more than the air and almost similar to human body. The density of water is important therapeutically as it is this quality of water that supports the child once immersed in water and also exerts an upward force. Because of the difference in the densities of various tissues in the body, a leaner child with lesser fat tissue will tend to sink more in the water whereas individuals with more fat tissue will tend to float.

*Viscosity*

Viscosity of water is the amount of friction generated with a movement in water. The friction is significantly higher in water than air which provides resistance to any movement of the body. This makes water an excellent strengthening tool. The resistance provided by the water is dependent upon the speed of the movement and direction of the movements which are patient dependent. In painful conditions if the patients stops the movement the resistance drops to zero and therefore the strengthening activities can be performed within the limits of patient's tolerance.

These unique properties of water provide desirable environment for the individuals with muscular dystrophy. Immersion in water brings about beneficial effects in various body systems of a child with muscular dystrophy.

**Beneficial effects of water immersion**

*Musculoskeletal system*

Immersion in the water increases the blood returning to the heart and in turn the blood delivered to all the organs. Most of this blood is supplied to skin and muscle tissue. Blood supply to the deep muscles increases nearly threefold during chest level immersion. Improved blood supply helps slow down the pro-fibrotic processes in the muscles. In addition to this as the muscles don't have to perform lengthening contractions for movements in water, the damage to the muscles is minimized and therefore the fibrotic processes are further slowed down. Immersion offloads the joints facilitating relaxation of the muscles and smooth movements. The resistance provided by viscosity of water for any kind of the movement helps in stabilizing the tone and strengthening the muscles. Improved circulation helps improve flexibility and pliability of the muscles. Reduced fibrotic processes , improved pliability and strength of the muscles, prevents the secondary tightness and contractures.

Most of the individuals with muscular dystrophy develop osteopenia (reduced density of bones) or osteoporosis (brittle bones) in the later stage of the disease due to not bearing weight on their extremities. Most of the individuals are on regular steroids which cause osteoporosis after prolonged consumption. The bone density therefore is a concern. Aquatic exercise helps to increase the bone density.

*Cardiorespiratory system*

Water exerts compressive pressure on the blood vessels and pumps up the blood from limbs to the heart. In individuals with muscular dystrophy because of inactivity, muscle weakness and secondary contractures and tightness the blood circulation is sluggish. This causes the blood to be pooled in the extremities and toxic waste to be accumulated. Immersion in water helps clear the toxic wastes. Improved blood pumping in the heart also provides heart with more blood to pump out improving the blood supply to the lungs and better oxygenation of the blood.

Compressive forces of the water provide resistance to the respiratory muscles (muscles required for breathing) and help strengthening these muscles. Exhalation or breathing out is passive rebound compression of the rib cage. In muscular dystrophy due to tightness of muscles and deformities there is incomplete exhalation of the air. Air is exhaled from only the upper parts of the lungs but accumulates in lower parts. Such accumulation could have various detrimental effects on the body. Immersion in water compresses the rib cage helping in better exhalation and lesser accumulation.

*Brain and Nervous system*

Increased blood supply to the brain leads to improvement in memory and other cognitive

symptoms. The child is more attentive in the water. Water immersion facilitates stimulation of para-sympathetic nervous system which facilitates relaxation of the body and suppression of the sympathetic nervous system that is responsible for the responses of anxiety. Immersion in water therefore facilitates relaxation and further suppression of the fibrotic processes.

These beneficial effects of water immersion are used by for therapeutic benefit by aquatic professionals. It is important to understand that although immersion in water is beneficial, goal oriented and targeted exercises are required for optimum recovery and maintenance of the individuals with muscular dystrophy.

Aquatic exercises increase serum endorphin (hormone responsible for mood elevation) levels and levels of neurotransmitter serotonin which are responsible for reducing pain and controlling many functions like sleep, appetite but mainly mood. Aquatic exercises have a psychological mood elevating effect.

*Is aquatic therapy an alternative to land based therapy?*

No, aquatic therapy is not an alternative but a conjunct to it. Land based rehabilitation and aquatic rehabilitation needs to be performed together. Neither is alternative to the other. Although there are some advantages of aquatic rehabilitation as compared to the land based rehabilitation both are essential for optimum recovery.

**Benefits of aquatic therapy over land based therapy**

- There is less weight on the joints, the child is well supported and joints are not under stress like on land. Therefore aquatic therapy helps achieve the benefits of land based therapy without causing any harm to the joints.
- As the individuals are able to perform the tasks in water much easier than on land, their confidence and activity participation increases.
- Activities in water are more fun and interesting for the individuals therefore there is better engagement of the individuals in a session and better adherence to therapy than on land.
- For individuals with severe movement restriction on land. Aquatic environment provides some freedom for movement.
- The risk of fall significantly reduces in aquatic environment
- As the frequency for lengthening contractions in the water environment is much lesser than on land the damage to the muscles while exercising is lesser
- It improves blood circulation to the deeper muscles better than on land and improves pliability of the muscles.
- The fibrotic changes in the muscles are lesser than on land

Therefore aquatic therapy may be preferred for some activities and during some stages of the disease. It provides an excellent medium to train the individual and improve their motor impairments. It is fun and enjoyable ensuring long term adherence. It helps to maintain various cardio-respiratory health parameters. It is safe and very effective in improving the quality of life of individuals with muscular dystrophy. However aquatic therapy alone is not sufficient and must be incorporated in the multidisciplinary rehabilitation program including land based exercises.

**Cardio-respiratory Rehabilitation**

Although muscular dystrophy is mainly a disease of the skeletal muscles, the pathology also has its effects on the breathing (respiratory) muscles and heart (cardiac) muscles. This causes direct impairment of the function of the lungs and heart (cardio-

respiratory function). In addition to this due to increasing movement limitation and skeletal deformities there is indirect impairment of the cardio-respiratory function. Therefore early cardio-respiratory endurance training exercises are recommended in case of muscular dystrophy. These exercises have many beneficial effects on the respiratory system and cardiac system which prevent rapid deterioration and secondary complications. Cardio-respiratory endurance exercises are the exercises that are performed to improve the capacity of the heart to circulate blood and capacity of the lungs to improve oxygenation of the blood during increasing physical effort.

**Benefits of cardio-respiratory endurance exercises:**

- Increased blood pressure as immediate response exercises improving the blood circulation
- Reduction in the blood pressure as a late response to exercises reducing the cardiac stress
- Improved capacity of the heart to pump blood
- Improved return of the blood from extremities to heart preventing stagnation of the blood in extremities
- Improved capacity of the lungs to breath in the air
- Improved oxygenation of the blood
- Improved clearance of carbon dioxide from the blood
- Ability to breath air out more forcefully
- Prevention of aspiration and secondary respiratory infection
- Improved ability to clear secretions by coughing, preventing secondary respiratory infection
- Blood vessels and blood circulation
- Formation of new blood vessels
- Improved connections and network of blood vessels
- Improved circulation to the deep muscles of the body, brain, lungs, liver and kidney
- Improved clearance of toxic waste substances from the blood

**Types of exercises are:**

- Pursed Lip Breathing
- Diaphragmatic strengthening / Deep breathing exercises
- Diaphragm is the most important muscle responsible for breathing. When it does not perform optimally muscles of shoulder and neck are used for breathing hindering their primary work of joint movement and stability. Therefore total muscle work responsible for breathing increases a lot and muscles get fatigues sooner. It is therefore important to strengthen the diaphragm and practice diaphragmatic breathing to improve breathing efficiency.
- Inspiratory Spirometry
- Huffing and coughing
- Forced exhalation
- Active cycle of breathing technique
- Postural drainage

**Benefits:**

- Strengthens respiratory muscles
- Prevents rapid respiratory deterioration
- Improves efficiency of breathing
- Facilitates deep inhalation
- Circulation of air in deeper parts of the lungs
- Uptake of oxygen and exhalation of carbon di-oxide is better

**Important considerations:**

- A doctor should be consulted at least once in a year and forced vital capacity test should be done.
- Flu and pneumonia should be not considered lightly.
- Regular breathing exercises should be done under the supervision of a physical therapist.

Cardio-respiratory rehabilitation is a very important area in the management of muscular dystrophy and it should not be neglected. Parents and caregivers must make sure that adequate care and training is provided to improve cardiac and respiratory health and maintain it for a longer time with fewer complications.

## Speech Therapy management

Parents of DMD/BMD children have noticed that as their age increases and the muscle deterioration progresses, speech also seems to get affected. This in most cases is due to hypertrophy of the tongue muscles. Slowly, swallowing and respiratory problems also ensues. Hence, it is important to identify speech (respiratory/ voice), swallowing, language, and cognitive-linguistic difficulties at the earliest, so that timely intervention can be done.

An attempt has been made herein to present the clinical profiles seen in children and adults with Muscular Dystrophy and the intervention program as suited for that profile. The authors do not promise infallible profiles and their accurate intervention programs as there may be a lot of variability within the same level and same type of MD. Hence an attempt is made to give clinical profiles likely to be seen at each level along with suggestions for therapy at each level. This may hold true for only about 40% of persons with Duchenne MD and Becker MD.. Thus it is only meant to give basic guidelines to care givers and allied rehab professionals or even beginning ASLP professionals who would be working with children and adults with MD.

Attention should be paid to the following points and the level of difficulty should be noted:

Did the child cry immediately after birth. • Did neck holding, turning, sitting, crawling, standing with support, walking, saying /ma/, /pa/, /ba/, /a/, /e/, / u/ and uttering meaningful first words occur as per age or was there any delay.

Does the child have poor attention, thinking, reasoning [inquisitiveness].

Is there any difficulty in moving the tongue adequately within the oral cavity, rolling on lips, touching palate, flapping to say /r/, lifting posterior to say /k/ or coughing, swallowing. Does the tongue appear thick and protrude out.

Is there any difficulty in puffing cheeks, blowing, sucking, chewing, and biting?

Do the lips remain wet always & need to be wiped periodically.

Is breathing from the nose more laborious? Are there complaints of lack of stamina and breathlessness?

Is there any difficulty in lifting things?

Is there a voice change, more towards a nasal tone?

Does the face lack expressions and appear mask like.

Does the child complain about reading, writing [errors in spelling & identifying letters] and complex language abilities like creating a detailed story on any topic etc?

Is the child's speech difficult for a listener to understand? Here are some simple exercises that can be done at home to reduce above mentioned

In case of those persons with other types of muscular dystrophy, at levels of early and advanced levels, the former may match with the first three levels (Levels I,II and

and the latter with the Levels IV and V given for DMD and BMD. Hence no separate profiles have been given for the other types of MD. The concerned professional could match a particular profile with the ones given below and apply the therapy regimen given for the profile it matches best.

The classification followed is as given below:- Level I Pre-symptomatic

Level II Early ambulatory Level III Late Ambulatory

Level IV Early Non-ambulatory Level V Late Non- ambulatory

LEVEL I- Pre-symptomatic

May not need any active therapy at this stage of the disease which may be in the period close to the diagnosis of the disease if it is identified earlier in the day.

**Speech**

There appears to be a slight change in finer aspects of motor development and behavior and hence speech being a very coordinated accuracy driven function may show some deviations.

Some may show less number of words per breath or run out of breath for last words of breath. In terms of voice and voice control, a person with Muscular Dystrophy (PMD) may not have a very wide pitch range and may occasionally have pitch breaks. There may be loudness fluctuations and especially after physical activity, their voice may sound very low and they have reduced loudness. Resonance may not be affected. Speech intelligibility may not be affected and occasionally there may be errors of articulation (pronunciation) such as cluster reduction eg Ice cream sounds like 'Ikkeem' or playtime sounds like 'peytime'. These errors too may not be noticed at single word production but in connected speech.

What may be noticed is the change in naturalness of speech where at times, again when they are physically fatigued, their speech may sound a bit sluggish or prolonged as though spoken with more effort and struggle than otherwise.

**Management**

An intensive therapy regimen is not indicated in children or adults with DMD/BMD at this level. A combination of oromotor strengthening exercises along with relaxation and breathing exercises would enhance their speech. These are given in detail at the management of Speech section at Level II.

**Swallowing**

Only if the child or adult with DMD/BMD, is eating in a hurry, eating mixed consistencies which also involves complex eating like hot soup which has vegetables and noodles apert from the thin liquid, or pani poori, he may have coughing due to entry of food particles into his airway. Or if he is fatigued he may not swallow in time and before the tongue moves posteriorly to guard the airway, food may trickle down in the airway leading to severe coughing. This may happen once in a week or a fortnight. This is not severe enough to cause weight loss but may lead to some discomfort when it happens. Sometimes a PMD may have food preferences such as based on previous experiences, he may avoid eating those foodstuffs where he had coughing or it took too much time to eat like pizza bread or naan / tandoori roti etc.

**Management**

The SLP/Swallowing therapist could guide these individuals to take some basic precautions, such as:-

Slowing the pace while eating,

Reducing the volume per bolus

To take repeat swallows

Pause between two mouthfuls

To avoid having mixed consistencies

To avoid distractions while eating

To maintain good oral hygiene and rinse mouth prior to and after each meal

**Language**

The PMD at this stage is not commonly seen to have any language deficits. In children with MD if the onset is around 5 years, and the child is having delay due to other factors such as reduced speech and language stimulation due to experience deprivation or the child has autistic features or hearing impairment, then there may be a chance that the child may show language developmental delays and deficits.

**Management**

For children with MD associated with other disorders, the SLP should guide the parents to manage the problems holistically with the help of the entire team as required.

Occasionally the child may be able to name people and objects and animals, but may not be able to use the language in communication. Intensive speech and language stimulation and therapy may be indicated in such children. However, if there are no associated disorders, in children with DMD/BMD, there are no indications for language deficits and hence no indications for management.

**Cognition**

As mentioned above, unless associated with other disorders, a child with MD at this stage is not seen to have cognitive deficits.

LEVEL II Early Ambulatory

At this stage, the child or adult with MD may have deficits of a mild to moderate severity in more than one domain. Speech and swallowing may be more affected as if the distal (limb) weakness has affected normal mobility then functions requiring fine motor coordination of muscle groups would understandably be affected.

**Speech**

**Respiration**

Words per breath:- May be reduced due to short breath supply and weakness of respiratory muscles. This may improve with rest.

Phonation Duration:- May be reduced and this contributes to reduced words per breath, low intensity of voice and reduced intelligibility due to very soft spoken speech. However insight may be good and PMD may learn strategies to stop, pick up listener cues and speak louder to reach to their listener.

**Management**

For breathing problem and unclear speech:

Do deep breathing exercises. Inhale as much as possible from nose & exhale through mouth. This will help to relax neck, shoulder muscles.

Do deep breathing followed by phonation of /a/,/e/,/u/ as much as possible.

Mechanical Aids to help breathing: These devices are simple and portable, which give better ventilation to Duchenne muscular dystrophy children. One of the methods is called intermittent positive pressure breathing [IPPB]. This requires a mouthpiece, which is connected to airway. If facial muscles are weak then facemask can also be used. This improves breathing during the day as it can be easily placed under the wheelchair. But if introduced early then respiratory functions are preserved better. Volume ventilator is another device, which can be used during nighttime because it is bigger than IPPB. This device reduces the difficulty in breathing when the child is

sleeping. Facemask or nasal mask is connected to the airway. These devices can be used as and when needed to improve ventilation.

In advanced stages of muscular dystrophy where breathing difficulties increase, another surgical option that can help reduce difficulty is tracheostomy. Here a small hole is made below the neck, which is covered with a mesh and is connected directly to the windpipe. Take proper consultation before choosing this option.

**Phonation/Voicing**

- Pitch range:- May not be affected greatly but with fatigue PMD may have pitch breaks.
- Loudness:- End of utterances are characterised by reduced loudness which is due to reduced respiratory pressure than a problem of the vocal mechanism
- Quality/Timbre:- May not be affected but there may be the effortful speech or fatigue in muscles of speech which may affect the quality of voice. Voice being the so called 'thermometer to portray emotional well being' when the PMD is depressed or upset, the quality and timbre of voice will understandably be affected.

**Management**

A good breathing pattern would be the base for a good voice. Easy onset of voicing needs to be established and relaxed phonation is an important step. The SLP needs to focus on increasing the pitch range and loudness of voive which would also improve the overall quality of voice.

Resonance

Normal

Articulation

Sounds in isolation: Better at sounds and word level than at phrase and sentence level. Intelligibilty of speech may be affected at sentence and conversation level. Most words with clusters would be simplified and produced by omitting the cluster…eg gammar for grammar; Adi for Aditya

**Management**

Motor planning & programming may also be affected: This is different from muscle weakness. Because of planning and programming errors, the child appears clumsy, uncoordinated with poor eye hand coordination. For eg. Picking up a matchbox, taking out the matchstick and lighting it. This task needs signals from brain to hold the target, which is received by eyes and hands. Then the concerned body parts and muscles carry out the task. But if coordination between the muscles and body parts is inadequate, initiation of right body part is affected which results in uncoordinated eye hand movements. To reduce this difficulty, a verbal clue to carry out tasks and help in reaching the target can improve the difficulty.

**Oral motor exercises:**

- For poor lip closure: 1. Hold upper and lower lips together with little pressure and repeat 10 times in a day. 2. Ask the child to hold an ice-cream stick, whistle or spoon between the lips. 3. Ask him to close lips together by himself by using a labial seal with his fingers.
- For inadequate tongue movements: 1. Protrude tongue and elevate, 2. Move from left to right. 3. Roll on lips, on teeth, palate, cheeks from inside. 4. Try to lick candies, lollipops, and ice cream. 5. Try to remove food particle [dough, chocolate] from the palate with force. 6. Do back and forth movement of tongue on palate. 7. Try to cough as an exercise, say /k/ as an exercise. 8. Say /pa/, / ta/, /ka/ as fast as possible maintaining clarity, speech, loudness.
- For inadequate sucking: 1. Try to suck

liquids from spoon. 2. Try to suck on lollipops, ice, tangy candies etc. 3. Suck on gauze piece dipped in honey, chocolate sauce etc.

- For inadequate blowing: 1. Blow whistles, soap bubbles, thermocol balls, bits of paper, candles etc. 2. Use respirometer [instrument to reinforce blowing & sucking].

Difficulties might occur in phonological processing: The child needs to recognize every letter and needs to associate it to its sound. This is done by the brain. The child may find it confusing to remember letter and sound. This problem again supports the problem with short-term verbal memory. Do not scold the child for making mistakes; rather give more practice with words along with pictures and letters. If not rehearsed regularly, the child would have problem. Practice sight words so that reading becomes easy. Consult a speech language pathologist for further detailed child oriented therapy.

Multi sensory approach: This involves integration of auditory, visual, tactile and kinesthetic approach. For eg: while teaching the child /p/ and /b/, the major difference between the two sounds is understood by listening because visually both look the same. Similarly for teaching /ka/ child can be explained it's the sound which can be produced by coughing hence child remembers the association with the action of coughing. When asked why is he coughing he hears the sound /ka/ once again and remembers it. Child will also know he has to produce /ka/ by lifting tongue from back and touching it to the roof of mouth. Kinesthetics means actually feeling the movement of tongue while producing the sound, like feeling how the tongue touches the roof of the mouth to produce /ka/ and with how much force tongue is pulled down back while saying /ka/.

**Prosody**

Overall the rate of speech would be slow and sluggish enough to attract attention to itself. The pauses in words may be awkward as when the PMD would run out of breath supply in the middle of a long word such as Borivli said as Boriv....ali with the pause taken to allow breathing.Intonation may not be markedly affected.

Speech Naturalness may begin to compromise from this level of severity onwards. However it should be borne in mind that the speech patterns in these individuals would be very variable. In some PMD at this level, voice may be a little low pitched, rate may be prolonged and speech sounds as if drawled and he may be at this level of difficulty for a long time with good intelligible speech.

**Management**

Speak slowly by prolonging the initial syllable post deep inhalation. . Exhale after every word. Ex. Count 1 to 10 slowly exhaling after every count. Practice omkar after deep inhalation. This will help to maintain good coordination between deep breathing, exhaling and speaking and reduce effort while speaking.

**Swallowing**

The clinical picture of swallowing may be as described above in Level I and in some PMD may be more severe. Delayed oral phase, difficulty to masticate food, control the bolus while mastication, difficulty to propel tongue posteriorly and inadvertent spillage of liquids into the airway, at this time leading to coughing may be signs in the oral preparatory phase of swallowing. Reflex may be triggered normally or may be a little delayed. If the PMD has pharyngeal weakness, there may be discoordination while swallowing leading to severe coughing for thin liquids such as water and milk. Overall, the PMD at this level would be comfortably managing on oral feeds.

**Management**

For difficulty in swallowing: 1. Reduce the size of the bolus. 2. Masticate slowly. 3. Push behind masticated bolus towards pharynx [throat] slowly. 4. Drink liquids slowly sipping and then pushing behind. Try chin tuck towards collar bone while swallowing. This will close airway passage and reduce aspirations. 5. If coughing starts, consult a speech language pathologist immediately.

**Language**

A PMD would at this level rarely manifest any language deficits but the use of language in a communicative context may be affected owing to the speech difficulties such as the person may not be able to sustain respiratory control long enough to complete the sentence and hence may not like to answer in class or describe an event amongst friends thus beginning to avoid speaking situations. He may participate less in conversations with people and take fewer turns and speak less when they talk.

**Management**

For delay in speech since childhood: 1. Talk as much as possible with the child so that the receptive vocabulary increases. 1. Talk as much as possible with the child so that the receptive vocabulary increases. 2. Use lots of pictures & actual objects to teach the child. 3. Talk slowly using open mouth approach at the level of child's eyes. 4. Speak loudly and let child observe your lip movements and tongue movements. 5. Encourage child's every attempt to communicate verbally. 6. Make it mandatory for child to use sounds to ask for his needs. Let him say / a/, /e/, /u/, pa/, /ma/, /ba/ etc whatever possible. 7. Consult a speech language pathologist for detailed child oriented goal plan to improve his speech and language. • For difficulty in hearing: 1. Consult an audiologist. • For reduced expressions/ smile: 1. Massage the cheeks in circular motion and consult a speech language pathologist [speech therapist]. 2. Stretch the lips and hold smile. 3. Round the lip and hold.

Difficulties doing mathematics [Dyscalculia]: Remembering signs, relative values [greater than, lesser than], abstract or symbolic concepts like ten's place, hundred's place, money, fraction, etc. is difficult. Memorization of tables, automatic calculations are difficult. For this, give plenty of practical examples for additions, subtractions, greater than lesser than. Multiplications and division exercises need to be understood by the child first. Ex. 2x2=4 [here explain 2 comes 2 times i.e. twice] 2+2=4 [here 2 are added i.e. the original value is increased by 2] 2x3=6 [here 2 comes thrice 2+2+2, in multiplication the second value is the number of times the first value is added]. 2+3=5 [here 3 is added in first number i.e. 2, in addition, the second value itself is added in original value]. 2-2=0 [here again second value is decreased from first value. Remember always that the second value should be lesser than first value]. 4/2= 2 [Ask the child to make 4 balls and then ask him to make two parts ensuring both the parts have equal balls. This way, explain the concept of division. Do not go by the regular methods used in school because abstract thinking is difficult for children with dyscalculia. Learning tables will also be easier with the method mentioned above. Even though it is lengthier, it is more practical. Most of the reading, writing and mathematical errors go unnoticed because of more enlarged problem of muscle weakness.

**Cognition**

This would largely be within normal limits and a PMD at this level would even be continuing school, college or vocation at this stage.

**Management**

Other problems experienced, especially in Duchenne muscular dystrophy: 1. Poor short term verbal memory: Inability to retain new

information in brain at a time. For this, try to use lots of pictures to introduce new information. Chunk it into small segments, so that remembering becomes easy. Try to associate it with the known information. eg. For remembering new address: use lots of pictures for landmarks.

Complex language processing might be difficult: This problem is not noticed easily as the child's day to day communication is not affected. But detailed description of any event or abstract reasoning might be difficult. This can be improved by giving topics to the child to construct 10 sentences. eg. Ask the child to do mind mapping. This increases thinking and reasoning abilities.

Mind mapping : Look at the illustration with a flow chart. One word is given to the child, say sun (centre) and it has five arrows where the child has to fill in words associated to sun e.g. solar energy, heat, plants need sunlight, sun is required to survive i.e. life. Again five words related to life; child has to mention words like oxygen, food, water, etc. This will improve association, reasoning and vocabulary as well. This also helps in improving memory and retention.

Some of the Duchenne muscular dystrophy children might show signs and symptoms of autistic spectrum disorder. These may occur at milder level, but if tackled adequately and early, it can help the child in better language and social development. Use extensive language stimulation using multi sensory approach so that child learns fast. If child tries to remain aloof, don't ignore it regarding it as muscular weakness. Consult a speech language pathologist if any kind of delay or deviance is noticed in child's speech and language abilities.

LEVEL III Late Ambulatory Speech

This group would be showing the similar clinical picture as seen in Level II however the severity would be more than that. One may see a moderately severe degree of difficulty in this group in all domains of speech and swallowing with lesser impact on language and cognition.

**Respiration**

There may be a breathless quality to speech with increase in weakness of the respiratory muscles however the primary modality used by an PMD for expression of one's thoughts would be through speech. There may be shortness of breath as the regulation of air supply for speech may be affected which requires excellent coordination of the ribcage muscles known as external and internal intercostals muscles apart from some action of the diaphragm. Hence words per breath would be reduced and the words spoken may be uttered in a soft voice with reduced intensity.

**Management**

The child or adult with MD may need guidance as to how to do steady breathing then to combine this with speech at phonation level then counting numbers and then slowly to introduce single words such as numbers and then days of week, months of the year and then go on to word completion tasks and so on to steadily increase the length of utterance while maintaining the breathing pattern.

**Phonation**

Again pitch range would be limited. Voice quality would be low pitched, soft, husky, hoarse and breathy (however not in all PMD) if associated with unilateral or bilateral vocal cord paresis. In persons with Oculopharyngeal dystrophy, the impact on voice would be much more.

**Management**

In case the PMD has hoarseness, the SLP can guide him into doing vocal adduction exercises wherein he places one palm over other while holding arms folded across chest

and pressing palms tightly over one another says /a/ in a steady voice. Phonation is likely to improve if the vocal adduction is good.

In case there is weakness of the upper limbs of these individuals, they could be guided to push against the hand of the SLP placed on the forehead which again helps in bringing the vocal cords together. This exercise done regularly may improve voice quality. Care needs to be taken to give them adequate rest periods to avoid muscle fatigue.

Resonance

Normal

Articulation

Intelligibility of speech may be affected for phrases and sentences as well as for single words especially in case of proper nouns like own name or address or names of family members. So, if the names are unfamiliar and without any help from caregivers, it may severely affect intelligibility of speech. it would be difficult for strangers and people not in the PMD's inner circle to understand him.

**Management**

The exercise regimen given at Level II can be used more intensively for children and adults with MD at this stage.

Consonant exaggeration as given by Darley, Aronson and Brown (1969,1975) could be given wherein, the person is taught to highlight and exaggerate the production of each consonant in the word eg in the production of / Saturday/ the consonants /s/,/t/ and /d/ are exaggerated. This is done with the intention to reduce speed and focus on speech sounds thus enhancing speech intelligibility.

**Prosody**

The rhythm and rate of speech would be compromised to a moderately severe degree and although the PMD may be communicating his thoughts and ideas through speech, speech would sound very disordered in all domains of speech. Rate of speech would be very slow and sluggish. Prolongations may be so long as to cause a break in the word. Eg 'M...yy...na..... me is...Ro ... han.'

This occurs as the speech muscle movements are very slowed down and by the time the PMD is completing the syllable, owing to prolongation, his breath supply is depleted and hence he has to begin again or complete the broken word in the next breath group. This severely affects the naturalness of speech. There is significant trade off in naturalness of speech in the PMD's attempt to preserve speech intelligibility. The PMD child or adult is likely to be very self-conscious of the awkward manner of their speech and may avoid speaking in new environs and to strangers and especially in group situations.

The PMD may have started using some adjuncts in speech on his own even without training. For example, he may use more gestures to support his speech or use exaggerated mouth movements to aid in clarity or if he sees confusion on the face of his listener, may offer to write or type a message or a significant word to clarify to his communication partner. A child with MD would look at his parent or therapist to ask for help if his communication partner is asking for repetitions.

**Management**

The exercises for fluency enhancement given at Level II may help if intensively followed. The speech efforts during therapy may be recorded and played back to show the desired speed and also the ease of production. This is encouraged to be followed even beyond therapy sessions and again, speech efforts need to be played back.

The techniques used by PMD which they may have developed on their own may be

fine tuned in therapy sessions and if they are helpful, the person is encouraged to use it. Effective communication is to be emphasized by the SLP rather than precision of speech and intelligible speech is to be attempted even if speech may not sound natural.

**Swallowing**

As in case of speech, so too in the case of swallowing, the clinical profile would be a more severe form than seen in Level II.

In persons with Oculopharyngeal dystrophy(Level III), the impact on voice and swallowing would be much more than in other types of MD at same level.

**Management**

Intervention by a swallowing therapist would be imperative.

Some children may have been advised to be on parenteral methods of feeding such as Ryle's Tube(RT) feeds (Nasogastric tube feeds) or even on gastrostomy (PEG) feeds considering the chronic and severe and progressive nature of the swallowing and feeding disorder. Even if the child or adult with MD is able to tolerate oral feeds, it may be easier to have semisolids and soft solids. Liquids, both thin and thick, may well lead to coughing so it would be a good practice to have liquids through RT or PEG.

The PMD could be taken for swallowing therapy and initially rehabilitative strategies could be used and later compensatory strategies could be used. At all times care needs to be taken to check for airway safety and then therapeutic feeds could be tried. If the PMD could have them with least discomfort, the volume of the feed can be increased and the RT feeds could be complemented with oral feeds. A diet chart can be maintained by caregivers in terms of the nature of food given, the quantity and the reaction it triggered. This would enable the SLP as swallowing therapist and the dietician to decide if RT can be removed or at a later stage in therapy, whether RT needs to be replaced by a PEG for long term care.

**Language**

As explained in PMD at Level II, there may not be any language deficits in these children and adults with MD at an implicit level unless associated with any other disorder. Problems are faced in communication as explained earlier owing to severe difficulties in speech and owing to poor intelligibility of speech. They may take less turns in speaking, depend on caregivers and close family members to fill in for them when not able to be intelligible. Avoidance in speaking situations may be frequently encountered or resorting to alternative means of communication may be seen. The load of conversational exchanges may be on their conversational partners and even if they are caring, the PMD may experience increasing amounts of frustration owing to their physical limitations as well as their communicative and feeding problems. From Level III to Level V, the quality of life (QOL) of these children and adults would be severely affected.

**Management**

The International Classification of Functioning, Disability and Impairment for Children and Youth (ICF-CY) gives very intensive considerations as to how these problems need to be looked at to reduce the barriers to communication, enhance the participation and reduce the environmental limitations for these children and adults with MD.

Cognition

**Management**

In Level III as well as in Levels IV and V, cognition may not be affected but the means to express it, that is speech, writing, signing, gestures, may be severely compromised and hence the PMD may not be able to express his thoughts, ideas effectively. In Levels IV and

V, it may be difficult to express his basic needs even and only close family members who are the caregivers would be able to interpret from his body language, slight movements of eyes, hands, fingers, eyebrows and the like.

The SLP can work on planning AAC devices so that the barriers to participation and communication may be reduced.

LEVEL IV: Early Non-ambulatory

**Speech**

There may be a variability in the speech of persons at this level of functioning. Some may show the similar impact of the severity of their physical limitations on speech and some may not be so severely impaired in speech and communication. There will always be a range within which their complaints would fall. Persons with MD even if belonging to same type of MD may still manifest different clinical profiles. So, although at Level IV a person may manifest speech, swallowing, language profiles ranging from those described for Levels II, III, IV and V. As is said, there are no constants in biological disciplines and a caregiver or rehab professional would have to identify their PMD's respective profile from these and work towards intervention accordingly.

**Management**

Speech may be increasingly difficult at this level and at Level V, and if seen may be largely unintelligible. All processes of speech may be affected to a severe degree and the PMD may have been trained by now, to use Alternative and Augmentative Communication (AAC) devices. Communication may be limited to here and now, to express concrete ideas more related to physiological needs of the person. In view of communication intentions, the child or adult with MD, may only use communication to regulate their environment for instrumental needs such as "Do this' 'Give me' ' Call my mother' etc.

Swallowing

**Management**

At this level of functioning as also at Level V, the child or adult with MD may have been placed on RT feeds or PEG feeds and may be having very little oral feeds in small amounts of safe consistencies. Or the PMD may be off oral feeds completely and only given hydration off and on to rinse the mouth or suck at wet gauze to maintain oral hygiene. Care needs to be taken that the PMD does not lose wait owing to swallowing and feeding issues.

**Language**

The profile for language may be as given in Level Iii and the person may be on AAC devices and facing severe limitations in using nonverbal means of communication as well.

**Management**

Communication oriented strategies need to be given whereas the speaker's role in producing intelligible speech is supplemented by efforts made to enhance the medium of communication such as to have better light on speaker's face, use of an amplifier to improve loudness and use better guessing strategies by conversational partners.

Cognition

Same as given in Level III

LEVEL V: Late Non-ambulatory

There may not be marked differences in the clinical profiles from Level IV and Level V except that in the latter the severity would be further affected. The functioning in all domains may be severely compromised.

**Speech**

Same as given in Level IV.

**Management**

**Swallowing**

Same as given in Level IV. Weight loss may be significant if the PMD has been on RT or PEG feeds for a prolonged period. Despite no oral feeds, the PMD is likely to have microaspiration of his own oral secretions which may eventually lead to aspiration pneumonia and may be one of the contributing factor in the mortality of these persons.

Some of these individuals may be on life support at this stage and may be on the ventilator frequently whenever they have severe respiratory difficulties.

**Management**

This would be similar to the management done for PMD at Level IV only the intensity of therapy regimen needs to be increased further to cater to the severity of the swallowing disorders. As in all persons with dysphagia, the focus would be primarily on airway safety and then on improving nutritional intake.

**Language**

Same as given in Level IV

**Management**

If the PMD is physiologically stable then may communicate with use of advanced AAC. Those with high tech devices such as seen in use by the illustrious Mr. Stephen Hawking, they could convey even complex thoughts and be a significant contributor to science and society.

**Cognition**

Same as given in Level IV and as explained above.

These are some of the guidelines that could be followed by rehab professionals and caregivers, family members and significant other persons (SOPs) of children and adults with MD. However it is by no means exhaustive. One may begin from here but may need to refer to more intensive treatment programmes. Also, this program in no way can replace the therapy given by the SLP and swallowing therapist. A treatment regimen would be more effective if the SLP could advise it and the therapy regimen could be carried out by the caregivers of the child/adult with Muscular Dystrophy.

## Occupational Therapy management

The role of the occupational therapist in muscular dystrophy:

An occupational therapist will evaluate and suggests ways that the patient can maintain better health and function as independently as possible. By helping them use self-care as well as work- and play-related activities. The occupational therapist will also train the patient for using any devices and mobility aids that might be required.

**OT evaluation framework**

Occupational therapists evaluate client's occupational performance (social participation), performance components (personal factors), and performance contexts (environmental factors). OT evaluation defines occupational problems of concern to the client 1.

Evaluation of Occupational Performance (Social Participation)

**Interview procedures:**

There are instrumentsthat are designed to assess participation from the individual's perspective, such as the Impact on Participation and Autonomy Questionnaire (IPA)2, the Late Life Function and Disability Instrument (Late-Life FDI)3 and the Assessment of Life Habits (LIFE-H)4. The Canadian Occupational Performance Measure,is often used in clinical practice. Only the LIFE-H has defined metrological properties with a muscular dystrophy population (reliability between evaluation

and between assessors). The LIFE-H documents the way people carry out activities of daily living and social roles.

**Evaluation of Performance Components (personal factors)**

Evaluation of performance components is necessary as muscular dystrophy is a progressive disorder. The client should be evaluated within a functional state of mind and strongly related to occupational performance priority area as identified by the person with muscular dystrophy.

**Evaluation of sensory and neuromuscular performance components:**

Muscular dystrophy is a slow, progressive neurological condition, and in this the other symptoms often precedes the decrease in the muscle strength. The pattern is usually distal to proximal involvement. There is also facial muscle weakness, ptosis, weakness of neck muscles, hypertrophy, foot dorsi- flexors and long finger flexors.5 Myotonia is a frequent presenting symptom (36 -75.9%) 5,6. Upper extremity range of motion is often affected in relation to decreased muscle strength which leads to decrease endurance, gross coordination, postural control, fine coordination and dexterity.

**Evaluation of perception and cognition:**

The neuropsychologist will usually evaluate for difficulties in perception and cognition. The Occupational therapist can provide a unique contribution in evaluating the effect of cognitive-perceptual impairments on participation in daily activities and social roles1. Special attention should be given to fatigue, hyper somnolence, executive function and apathy in patients suffering from muscular dystrophy.

**Evaluation of psychosocial skills and psychological components:**

Evaluation of psychosocial skills and psychological components includes the ability to interact in society and to process emotions2. It is necessary to gain knowledge about these components in order to help the patients with muscular dystrophy to maximize function. It includes psychological skills (interest, valuesand self-concept), social skills (role performance, social conduct, interpersonal skills, and self-expression), and self-management (coping skills, time management and self-control).

**Evaluation of Performance Contexts (environmental factors)**

The role of environmental factors and especially the implementation of community services (home services, meal preparation, nursing at home, budget management, etc.) should not be underrated as these are most probably effective measures for alleviating some of the consequences and burden imposed by the disease7, as muscular dystrophy is a progressive disorder. As a group, muscular dystrophy patients show poor academic achievement, high unemployment,low family income, and high reliance on social assistance compared with the general reference population thus confirming a socioeconomic disadvantage8,9.

People who receive less emotional and social support from others are more likely to experience more depression, less well-being, and higher levels of disability from chronic diseases10. The perception of negative attitude and support of family and friends is an explanatory factor for level of participation in work, leisure and mobility. The perception of obstacles related to access and use of technology and government services is also related to level of participation11.

**OT intervention area**

Recommendations are usually based on clinical practice because there are very few studies in occupational therapy. Based on the findings from qualitative studies, a recent

review of the literature recommends a client-centered approach that includes the following aspects:

Patient education is important about the disease because education plays an important role in his or her understanding of the need to implement adaptive strategies (Jönsson et al., 1999; Nätterlund&Ahlström, 1999; Young, 1989); evaluating the perception of the patient of his or her life history, personal values, goals, and problems (Jönsson et al., 1999); adaptive strategies to be explained to the patient, which can be used in occupational therapy to empower the patient to make changes in his or her occupational performance (Jönsson et al., 1999).

Occupational therapy interventions include training of activities of daily living, skills training (fine motor skills), advice and instruction in the use of assistive devices, provision of splints and slings, counselling on energy conservation and work simplification strategies, educating patients, families, and caregivers or a combination of the above.

## Occupational performance treatment

The purpose of Occupational Therapy treatment is to help the individuals learn or relearn occupational performance that they need to live as independently as possible. Treatment strategy is mostly geared toward compensation and education. Three options can be explored for the compensation approach:

1) Alter the task method;

2) Prescribe assistive devices or;

3) Adapt the task environment.

Education can be a real challenge with muscular dystrophy in relation to cognitive functions.

Personal Care: Specific problems that are often encountered may relate to upper limb function (such as washing hair, picking up a cup, buttoning, wiping oneself after going to the toilet,), to lower limb function (such as standing in the toilet, walking to the toilet,) or both (such as getting in or out of the bath, putting on trousers).

An occupational therapist will recommend modifications for easy and safe access at home, school, or work, especially when using a wheelchair or a walker.

They will also suggest ways that daily living activities such as Dressing, eating, bathing, toileting, shopping, and doing household chores can be performed by the patient on his or her own. Using of assistive devices such as long handle scrubber (Fig 18), universal cuff (Fig 19), writing device (Fig 20) etc. can be altered according to the patients' needs and can be used.



*Fig 18*



*Fig 19*

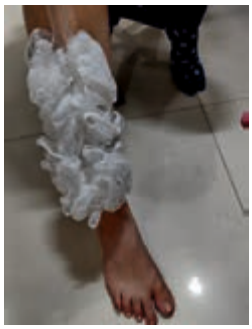


*Fig 20*

They will help in recommend appropriate equipment for self-care tasks, such as commode chairs, shower chairs, beds, and mobile arm supports.

The occupational therapist will help in recommend communication aids and devices such as computers to assist with school and work-related activities as well as social networking.

Multisystemic complications such as anal incontinence, diarrhea, and dysphagia, may add to these - so considering remodeling them can assist in personal care. These aspects are usually approached in a problem-oriented way by suggesting adaptive techniques, specialized equipment and/or community services.

Mobility: Some patients have an early and severe involvement of the knee muscle weakness usually the extensors, due to which they complain of multiple falls and are rapidly wheelchair dependent. Patients who require wheelchairs will often have moderate to severe proximal and truncal weakness. Therefore, when the wheelchair is prescribed, attention must be paid to stability and posture while seated, the ability to stand from the chair and the ability to transfer12. The need of an electric wheelchair, power wheelchair (Fig 4), or a four-wheel scooter can also be explored, although their use is less frequent.



*Fig 21*

**B) Performance components treatment**

*Muscle strength:* Muscle strengthening includes focusing on muscle groups which have muscle grade 3 or more (that could be used against gravity or muscles needed for stabilization).

*Range of motion:* Muscular dystrophy can restrict the flexibility and mobility of joints. Range of motion exercises usually help to keep joints as flexible as possible.

*Exercise:* Low- impact aerobic exercise, such as walking, swimming, can help and maintain the strength, mobility and general health.

*Orthoses:* Teach the use of arm slings or orthotic devices to support weak muscles in the wrists, arms, ankles, and shoulders to improve their function. Suggest the use of a corset or a body jacket to provide better support and balance to the spine in the case of scoliosis (abnormal curvature of the spine).

**C) Performance context environment**

Provision of education and information has been stressed as a mean of assisting patients and families to cope with neuromuscular diseases. Supportive relationships, whether in the form of practical help, emotional support, or provision of information, may facilitate health-promoting behaviors in patients with muscular dystrophy and should be encouraged.

An occupational therapist will teach energy conservation techniques and work simplification techniques so that body movements are more efficient, which can help with reducing fatigue.

An occupational therapist willidentify ways to promote creativity and social activities that can boost the patient's confidence, to promote social interaction at the community level that will help them boost the confidence.

To design facilities which helps in improving

the comfort in patients with muscular dystrophy.

**Psychological Management**

Consideration of psychological aspects of the child is important to be taken into perspective, since it has an impact on the overall physical wellbeing of the child too. Cognitive and behavioral changes that happen need to be understood and handled accordingly.

What are the cognitive and behavioural changes or problems associated with muscular dystrophy at different ages?

**Childhood:**

*Cognitive Changes:*

During this period, the parents may notice language developmental issues like the child may have difficulty in expressing his/ her needs or may have difficulty in remembering a lot of information. This is especially seen in boys with Duchenne's muscular dystrophy because of which they are usually diagnosed as having learning disability especially dyslexia. Children with Duchenne muscular dystrophy may have problems with maintaining attention or concentrating, however these issues may be overlooked by their parents due to their physical condition.

*Behavioural Changes:*

Usually children at this age display problematic behaviour but children with muscular dystrophy display more impulsive behaviour and poor emotional control. Many of these children are on steroids, which usually have side effects that affect their behaviour. Some side-effects seen are that these children are more rigid, emotionally low and may not be cooperative. Eventually as their condition progresses, boys aged 8 to 10 years start having difficulty in adjusting as they stop walking and start using the wheelchair.

**Teenage:**

*Cognitive Changes:*

When young men with muscular dystrophy grow older their cognitive problems increase like difficulty in planning, organizing or completing tasks. As their educational level or work increases, their problems also increase.

*Behavioural Changes:*

Teenage or adolescence is normally known to be a difficult age, as at this point of time there are many hormonal and emotional changes in the body. Along with these, there could also be a possibility of having a limitation in the physical development. This may be a result of certain medical treatment for example: an individual may have small height or may have delayed puberty which could be an effect of puberty. This could make dealing with the changes of adolescence more difficult. Teenagers could have a difficult time adjusting as they might not get the freedom or independence since their condition will require more care and assistance as compared to others. As muscle weakness progresses, they are at risk of becoming more isolated or socially withdrawn. Parents should look for signs of chronic sadness, depression or anxiety.

**Adults:**

*Cognitive Changes:*

Adults with muscular dystrophy may adjust with their cognitive issues and may find out ways of dealing with them.

*Behavioural Changes:*

As individuals with muscular dystrophy progress from teenage to adult hood, they may undergo mood changes and emotional changes which would eventually lead to changes in their behaviour. Adults with Duchenne muscular dystrophy would be bed ridden and this would result in loss of independence. This would eventually lead to increased anger, irritability & frustration and

may even show signs of depression.

Whereas, adults with limb girdle muscular dystrophy or Becker's muscular dystrophy would develop difficulty in functioning, which could lead to decreased socialization, low self esteem & self image. Also they would have multiple questions as to whether they should get married or if married, whether they should have children.

**What is the importance of psychological assessment in muscular dystrophy?**

A person with muscular dystrophy is at an increased risk of cognitive and emotional problems; hence early diagnosis and treatment is of great importance.

Cognitive Assessments: Especially a DMD child with learning disabilities or autism should undergo testing for IQ, memory, attention span, problem solving, etc.

Emotional and Behavioural Problems: A patient with muscular dystrophy should go to a psychologist for emotional status examination either every 6 months or annually. As many of the patients go into depression or suffer anxiety disorders, which may in turn worsen their physical condition.

**Learning and Cognitive Skills in Muscular Dystrophy:**

Intelligence is defined as "an individual's ability to adapt and constructively solve

problems in the environment" as mentioned by David Wechsler, a well known American psychologist.

Intelligence can be assessed by the means of an intelligence test. There are many intelligence tests available but an accurate IQ can be gained when the IQ test is for the Indian population, for e.g. Malin's Intelligence Scale for Indian Children (MISIC). This IQ test usually has 2 parts i.e. verbal sub-test and performance sub-test.

The verbal subtest includes questions regarding language, for e.g. how are the piano and guitar similar. Whereas performance sub-test assesses visual thinking and motor performance, for e.g. a subtest on block design, where you have to copy a block design. The time taken to conduct an IQ test is 30 minutes to 2 hours.

It is often seen that boys with Duchenne muscular dystrophy usually are at a higher risk of delays in walking, running and sitting. In a similar way, it is seen that the IQ of these children ranges from above average to below average. However, they are at an increased risk for having low IQ or some learning disability.

**What is the cause of cognitive weakness?**

Most probably there is a relation to brain functions. It is known that dystrophin is normally present in other tissues as well as muscle tissue, including the central nervous system. The lack of brain dystrophin might therefore play a role in the cognitive functioning of the boys. The dystrophin isoforms Dp427 - C and Dp427 - P would normally be distributed to the hippocampus and purkinje cells in the cerebellum respectively, contributing to greater post-synaptic density. The cerebellum and hippocampus are part of an integrated network that includes connections with the frontal brain regions. This altered postsynaptic plasticity may hinder efficient memory, automation and planning/organization.

**Areas of Cognitive Weakness:**

- Difficulty in finding words.
- Difficulty in short term memory.
- Difficulty in concentrating.
- Difficulty in switching from one activity to another.
- Difficulty in completing tasks.
- Difficulty in multi-tasking i.e. performing

multiple tasks at the same time.

Brain Areas in which Dystrophin has been found:

| Brain Area | Function of the Area |
|---|---|
| Hippocampus | Memory |
| Cerebellum | Automatization |
| Frontal Lobe | Planning & Organization |

Research is still under process about the role of dystrophin in the brain.

**Attention, Listening and Memory:**

It is seen that patients with muscular dystrophy have problems remembering many things or assimilating information together, have difficulty in following directions or may not seem to listen. Also, there is a high chance for problems with the ability to focus on a task and with dividing the attention between many activities at a time. Their level of distraction may be high. Individuals with high IQ could also have difficulties dividing their attention.

*Strategies to Improve attention, listening and memory problems:*

- Sit close to the child and explain to him about the task or the problem to be solved to avoid him getting distracted.
- Break down the instructions and information into simple and specific statements.
- Check if the child has understood what he has been asked to do.
- If the child has difficulty dividing attention between many things at a time then give him one activity to complete at a time this would also help eliminate stress.
- If the child has an attention problem or has reading difficulty, then underline important points so that he does not miss out on important information.
- While teaching the child, use small time durations i.e. instead of a 1 hour long period use short 20 minute periods.
- Either make a to-do list or let your child himself make a list of activities which will help him remember the activities that he is supposed to complete.
- Arrange for extra time to be given to the child for him to complete an examination, as his physical condition might be a barrier or he might have concentration/memory problems.
- If the child is not able to write his examination because of weakness in his hand, arrange for someone else to write the exam for him.

**Psychosocial Adjustment to Muscular Dystrophy:**

Psychosocial adjustment means the way in which one adjusts to difficult and stressful events associated with muscular dystrophy.

*Psychosocial Adjustment has six areas of functioning:*

1. Relationship with friends.
2. Dependence on family members / caregivers.
3. Opposition to things or situation around.
4. Productivity.
5. Anxiety or depression.
6. Withdrawal.

Undergoing emotional and behavioral problems is a normal and a healthy way of socio-emotional development. However, research has shown that individuals with physical disability have an increased risk of developing behavioral and emotional problems. This is usually one of the reasons why individuals with muscular dystrophy adapt to the consequences of behavioral and emotional problems. It is usually seen that individuals with muscular dystrophy

become more capable of coping with their disability.

**Learned Helplessness in Individuals with muscular dystrophy:**

Learned Helplessness is when an individual thinks and feels that he or she has no control over the situation or whatever is happening to him or her. Learned helplessness can lead to many emotional reactions and behavioral problems.

*For example:*

Negative Chain: An individual with muscular dystrophy who is regularly undergoing physiotherapy, despite which he is deteriorating and is wheelchair bound feels sad and disappointed because he cannot walk independently.

Positive Chain: An individual with muscular dystrophy who can move around in his wheelchair feels that he is self-reliant and independent. He is still happy about the things that he can do by himself.

**How to deal with emotional problems?**

Everybody undergoes emotional problems but individuals with muscular dystrophy undergo a lot of stress and emotional changes. These changes are related to the different stressful situations arising because of their disease like hospitalization, being wheelchair bound, dependency, etc. Also when individuals take steroids, it could have side effects like mood swings and frequent changes in emotions.

*Stages of acceptance:*

Phase 1: Denial: "No not me"

Phase 2: Anger: "Why me, it's not fair"

Phase 3: Bargaining: "Yes me, but I still want to"

Phase 4: Depression: "I'm so sad, why bother with anything" Phase 5: Acceptance: "I am going to have a worthwhile life"

**Strategies to handle emotional problems:**

The first step to handling the emotional problems is to understand or give a name to the emotion that he or she is experiencing, for e.g. whether the person is experiencing anger, sadness or fear.

It is important to make the individual feel that he is being heard. Hence, he should have support in the form of a good listener.

It is very important to teach the person to express his emotions in a constructive manner.

The individual could maintain a personal diary where he writes the thoughts & emotions that he is experiencing.

If an individual with muscular dystrophy is having frequent mood changes which are having negative effects on his health and routine, he should consult a psychologist.

**Temper Tantrums:**

Temper tantrums are normal issues associated with the process of growing up. The issue of temper tantrums is usually seen in boys with Duchenne's muscular dystrophy. This is their way of drawing attention to themselves, be it positive or negative attention. Dealing with temper tantrums becomes a tricky situation. If you shout at the child for throwing a tantrum, it may lead to an increase in the tantrums, whereas if you show the child that you are okay with the tantrums then it would lead to encouragement of such a behaviour.

**Strategies to handle temper tantrums:**

The best way to deal with this problem is to ignore when the child is throwing temper tantrums. However this technique cannot be always used.

Time out: This does not mean punishing the child; it means that we are allowing the child to calm down.

For e.g.: we make the child sit in the corner of

the room where there is no possible means of entertainment and where the child feels bored. He should be made to sit there for at least 5 to 10 minutes and he should be asked to think why he was given timeout. This would help him avoid being given a timeout in the future.

However, when temper tantrums worsen or may be out of your control then you should consider consulting a child psychologist.

When to consult a Psychologist?

You feel helpless in dealing with the temper tantrums.

It leads to stress and negative feelings within the family.

You are not comfortable with the response after the temper tantrums.

The child causes some sort of harm to himself.

**Coping with Duchenne muscular dystrophy:**

Usually it is seen that persons with muscular dystrophy accept their disorder and adapt to it quite well. However, many a times, due to stressful situations or inability to function socially or physically, they may undergo sadness, anger, frustration or guilt.

**Strategies to Maximize Coping Skills:**

Be available and open to talk to the affected person.

Try to identify the problems that the person is undergoing, which are stopping him from functioning up to his maximum potential.

Allow the individual to be as independent as possible.

**Quality of Life:**

Quality of life is more important than the quantity of life. This is also true for individuals with muscular dystrophy as they have a whole life with possibilities ahead of them. Hence, it is important to understand that rather than aiming for the quantity of life we should aim to improve the quality of life.

Strategies to improve quality of life:

Take each day as it comes rather than planning for the future.

Help the individual find happiness in small things.

Don't make them feel different or disabled. Treat the individual in the same way as others.

Safety is very important, but that does not equate to binding him within the house. In fact, he should be given the opportunity to experience situations and activities like other people.

Help him socialize and keep in touch with other people. Socializing is very important for his well-being.

Help the individual develop a hobby which keeps him engaged and helps vent out his feelings and emotions.

Help him develop a positive self-image and self-esteem.

Let him be as independent as possible.

Like other people go out and have vacations or small trips, even individuals with muscular dystrophy should be able to take short trips. Make proper arrangements keeping in mind their needs for e.g., a wheelchair accessible hotel.

**Strategies for Caregivers:**

*Ways to manage aggressive & difficult behaviour:*

Develop a weekly routine for the patient, explain it to him and help him stick to it. Incorporate rewards for tasks well achieved, as children are usually noncompliant after exercising every day.

Incorporate recreational activities in the time table as it is very important to have

something to look forward to during the day. This helps to motivate the patient and does not let him get depressed about his condition. For example: Painting or playing board games with siblings or watching television.

Explain the situation to the patient, if there are any changes made or if he is taken for some therapy that he denies to undergo, for example: If the patient does not want to exercise on a particular day, explain to him the situation, how doing so would have repercussions in the long run and set up a reward system if necessary.

Try to keep calm when a child is misbehaving. Angry parents and teachers tend to escalate the situation. This would worsen the situation as the child would feel neglected and feel that nobody understands him.

**Focus on the positives:** Strategies that only focus on punishment do not promote positive behaviours. Increase motivation, or change attitudes. Rewarding/ praising/encouraging good behaviour is more effective in the long run. Look for opportunities to say "yes" instead of "no." ("Yes, you can have a cookie, after you…").

**Psychotherapy and drug interventions:**

Well-known techniques exist which help in various areas. These include training for parents to cope with bad behaviour and conflicts, individual or family therapy and behavioural interventions. Applied behaviour analysis may help with certain behavioural issues related to autism.

Some children & adults may benefit from the use of prescribed medicines which help with emotional or behavioural problems. These medicines can be used under specialized supervision and monitoring for depression, aggression, obsessive compulsive disorder (OCD) or attention deficit hyperactivity disorder (ADHD), when these problems have been specifically diagnosed by a doctor.

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# Chp. 21 Spinal Muscular Atrophy

**Dr. Amruta Paranjape, Dr. Shruti Shirke, Dr. Rabab Kothari, Dr. Abhishek Gupta**

Spinal muscular atrophy (SMA) is an inherited severe neuromuscular disease. It is characterized by degeneration of the anterior motor neurons which leads to progressive muscle weakness and paralysis.Sensation is not affected as it originates from the posterior horn cells of the spinal cord. Several muscles are spared, including the diaphragm, the involuntary muscles of the gastrointestinal system, the heart, and the sphincters. It is an autosomal recessive genetic disorder. (Figure 1). SMA is caused by mutations in the survival motor neuron 1 (SMN1) gene. The location of SMN1 gene is in complex region of 5q13 which contains SMN2. SMN2 is a homologous pseudogene of SMN1.[1] Muscle weakness and atrophy are the predominant clinical features, where proximal muscles are more affected than distal muscles.[1]There are 5 types of SMA of which type 0,1 and 2 is the severe most.

In children SMA is the second most common disease inherited in an autosomal recessive pattern that affects children after cystic fibrosis.(2)

SMA is the leading cause of infantile mortality, and the carrier frequency of SMA is 1:35 with an incidence of 1 in 6000 live births.[3]

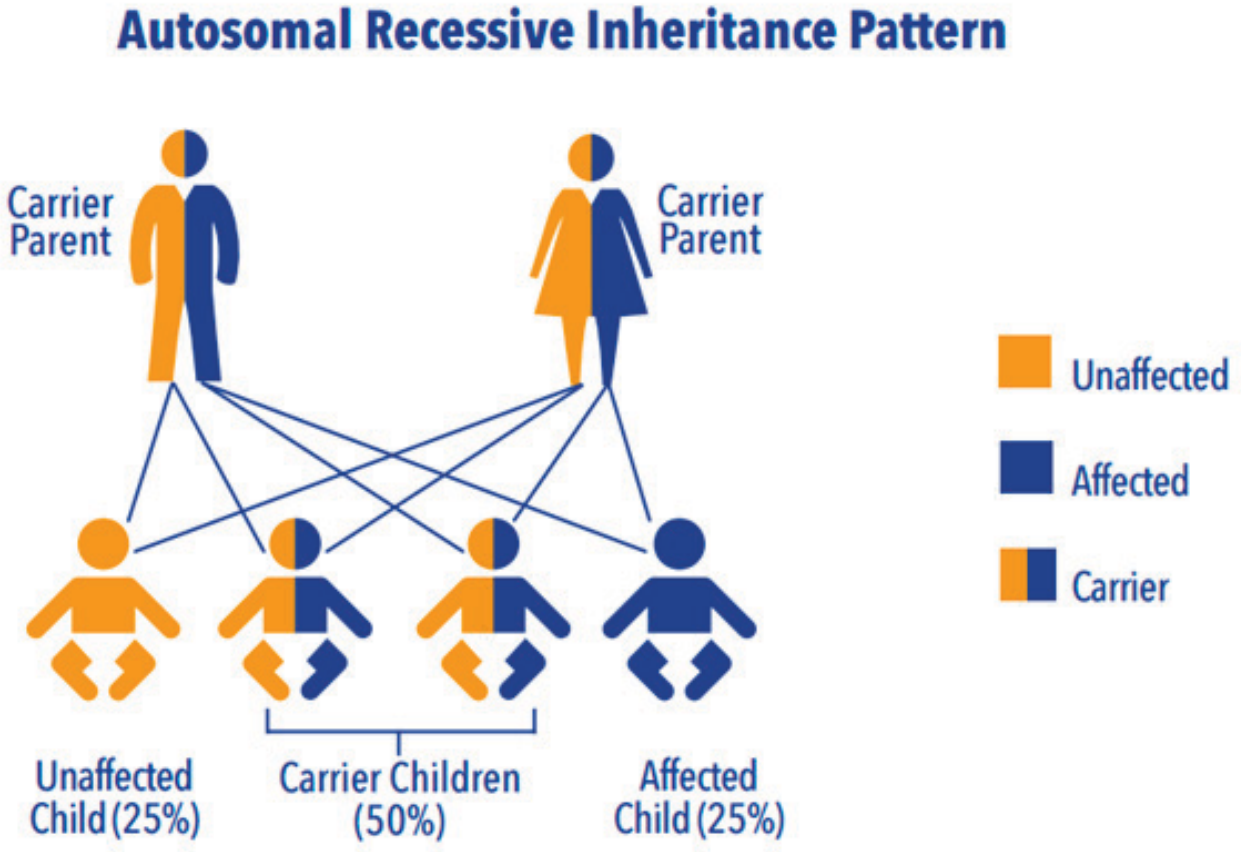



Figure 1: Inheritance pattern of SMA

## Pathophysiology

SMA is caused by a mutation in the survival motor neuron (SMN) gene. If the gene is absent, it results in programmed cell death. (4)

1. **During gestation period gene is inactive and allow normal apoptosis in developing fetus**
2. **Gene is active in matured fetus**
3. **It stabilizes neuronal population.**

**In normal fetus**

4. **Gene produces protein that is required for the functioning of the nerves which control muscles.**

**The severity of symptoms is related to SMN2 genes, when SMN1 gene fails to produce protiens SMN2 gene comes in action and produces few full length and functional protein. A normal person carries 2 SMN2 gene copies, person with**

**In SMA**

4. **Fails to produce the protein hence muscles do not function properly leading to severe muscle weakness.**
5. **The exact mechanism or timing of abnormal motor neuron death is unknown.(5,6)**

**SMA can have anything between 1-4. Therefore SMN2 gene is the maximum in SMA type 4 and less SMN2 gene copies in SMA type 1.(8)**

## SMA Classification with clinical features:

SMA is classified in 5 types; type 1,2,3 are based on the highest level of motor function (i.e sitting or standing) and age of onset while type 4 is for adult onset cases & type 0 is for patients with prenatal onset & death within weeks.[1]

| Type | Age of Onset | Clinical features | Complications | Natural Age of Death |
|---|---|---|---|---|
| 0 | Prenatal | 1. Poor or absent head control (Figure 2)<br>2. Hypotonia<br>3. Areflexia<br>4. Severe weakness<br>5. Facial diplegia | 1) Atrial septal defects<br>2) Joint contractures<br>3) Aspiration & Respiratory failure | <6 month |
| **1 (also known as Werdnig-Hoffman disease)** | 0 – 6 months | Same as type 0 & as follows:<br>1. Inability to sit without support<br>2. Intercostal muscle weakness (relative sparing of diaphragm) and "belly breathing" (pattern of paradoxical breathing)<br>3. Weakness of tongue muscles and tongue fasciculations<br>4. Difficulty in swallowing.<br>5. Never achieve standing, sitting or walking.<br>6. Hypotonic | Same as type 0 | <2 yrs. |
| **2 (also known as Dubowitz Disease)** | < 18 months | Same as type 0 & type 1; but in this type<br>1. Sitting without support is achieved at some point during their development but dependent in walking (Figure 3)<br>2. Proximal leg weakness is more than weakness in the proximal arm muscles<br>3. Survival strictly depends on the care and rehabilitation given. | Same as type 0 & as follows:<br>1. Progressive scoliosis (secondary to muscle weakness)<br>2. Ankylosis of mandible<br>3. Scoliosis & Intercostal muscle weakness together leads to restrictive lung disease (Figure 4) | >2 yrs. |
| **3 (also known as Kugelberg-Welander disease)** | > 12 months to >3years | 1. Walk independently at some point during their lifetime<br>2. Leg weakness leads to the need of a wheelchair at some point<br>3. Difficulty in stair climbing | 1. Progressive scoliosis<br>2. Sometimes, respiratory failure | Adult |
| **4 (Mildest form of disease)** | >21 years | Similar to type 3 | | Adult |

There are many more motor neuron diseases that are caused by mutations in genes other than the SMN1 gene and are called as non 5q SMA disease. A person affected with non- 5q-SMA also have muscle weakness but featuresdiffer from 5q-SMA.

These features can include early deformities, distal muscle involvement more than proximal muscles, diaphragmatic paralysis with early respiratory failure and cerebellar degeneration too. It can be diagnosed with DNA diagnostic tests.

A list of some non-5q-SMA diseases are mentioned in the table below.(7)

| Name | Mode of inheritence |
|---|---|
| Spinal and bulbar muscular atrophy | X-linked recessive |
| Arthrogryposis multiplex congenita | X-linked recessive |
| Spinal muscular atrophy distal | X-linked recessive |
| Motor neuropathy, distal with vocal cord paralysis | Autosomal Dominant |
| Arthrogryposis congenital, neurogenic type | Autosomal recessive |
| SMA distal type 5 | Autosomal dominant |
| SMA with respiratory distress | Autosomal recessive |
| SMA congenital, scapuloperoneal amyotrophy | Autosomal dominant |
| SMA proximal, adult | Autosomal dominant |

***Table 1- Non 5q SMA diseases**

**Diagnosis-**

**Preimplantation testing**

During in-vitro fertilisation It can be used to screen for SMA-affected embryos.

**Prenatal testing**

It is done through chorionic villus sampling, cell-free fetal DNA analysis and othermethods.

**Carrier testing**

People who are at risk of being carriers of SMN1 deletion can undergo carrier analysis using a blood or saliva sample.

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Figure2: Activation of neck muscles & abdominal muscles



Figure 3: Sitting without support



Figure 2: poor or absent head control



Figure 4: Scoliosis

## Physiotherapy Management:

The cure for SMA is not available currently. The supportive treatment should be aimed at improving the patients' quality of life and minimizing disability, particularly in patients with slow progression. The goals are to maximize the patient's independence and quality of life at each stage of the disease.

Care & support in the form of treatment can be given as follows:

1. Respiratory care: Chest Physiotherapy-

The chief cause of morbidity and mortality in SMA patients are the respiratory complications due to respiratory muscular weakness and fatigue, as well as thoracic cage deformity due to the complication of kyphoscoliosis.The work of breathing increases in restrictive lungs caused by SMA. Respiratory failure can result from factors that either directly or indirectly increase work of breathing, including (i) respiratory muscle weakness and fatigue, (ii) impairment of the central control of respiration.



Fig 5: Spiromete

An adequate peak expiratory flow is unable to achieve in SMA patientswith progression of disease which results in impaired cough resulting from inspiratory and expiratory muscle weakness.The primary airway clearance therapy is best administered with the combination of cough assist for all SMA patients with respiratory illness.To improve efficacyof cough mechanism and promote cough mechanism by breathing exercises including spirometry (Fig 1), vibration (Fig 2), percussion (Fig 3), active cycle breathing techniques, coughing techniques, sometimes by suctioning. Suctioning should be used in all SMA patients with excessive secretions or in those with an ineffective cough & is a critical part of the treatment.



Fig 6: Vibration technique



Fig 7: Percussion technique

Breathing exercises such as apical breathing, diaphragmatic breathing & lateral costal breathing, spirometry exercises and expiratory exercises such as blowing bubbles, blowing candles to maintain & strengthen the respiratory muscles (Fig 8a & 8b)

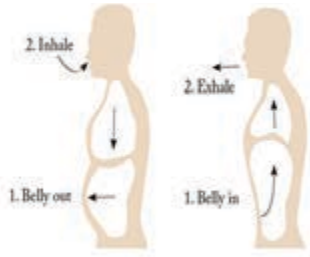



Fig 8a: Diaphragmatic breathing exercises



Fig 8b: Blowing exercises

Breathing exercises are to improve endurance & function of patients' during activities of daily living. Stretching of shortened muscles of lateral trunk & improve breathing in order to prevent restrictive lung disease, secondary to scoliosis.[3]Incaseofrespiratoryfailureorto prevent chest wall distortion, non-invasive positive pressure ventilation (NIV)should be used.NIV improvesthoracoabdominal coordination and sleep breathing parameters during sleep in patients with thoracoabdominal asynchrony.[4] When NIV is not tolerated or in the treatment of chronic respiratory failure, continuous positive airway pressure (CPAP) should only be used.

2. Neuromuscular & Musculoskeletal Physiotherapy:

The goal of active exercises is to maintain range of motion, increase muscle flexibility, and prevent contractures but it should be non- fatiguing exercises.



Fig 9a: Stretching of hamstrings using calipers



Fig 9b: Stretching of hamstrings

An elongation of shortened muscles, such as tightness of ilio- psoas muscle, hamstring muscle, tendoachilles due to improper positioning & weakness of antagonists, is done by stretching with calipers (when there is excessive tightness) & without calipers.(Fig 9 & 10)



Fig 9 & 10: Stretching of Tendo- achilles

The joint integrity & mobility is maintained along with strength training by Passive range of motion (ROM), Active- assisted ROM, Active ROM & Resisted ROM.



Fig 11a: Poor or absent head control



Fig 11b: Activation of neck muscles & abdominal muscles

Exercises to recruit, activate & strengthen weakened neck muscles due to poor or absent head & neck control and poor neck & abdominal muscles strength (Fig 11a & 11b).

Active assisted exercises include suspension therapy of upper & lower extremity muscles with strength below functional level (less than grade 3) & spring exercises for upper extremity as well as lower extremity (Fig 12 & 13).



Fig 12:Suspension therapy for lower extremity using calipers



Fig 13: Upper extremity exercises with springs

For the muscles above functional level (more than grade 3), active exercises of weak muscles are designed. Followed by active exercises regime, a progressive resistance training (PRT) exercise program is provided in order to increase strength and improve motor function in children and young adults with SMA. The type of resistance applied to the muscles is in the form of body weight, resistance bands, free weights which involves a systematic increase in resistance training parameters to improve an individual's ability to exert force.[5]

Bed mobility activities are advised to improve the level of independence. It includes supine to side lying, side lying to sit, quadruped, kneeling, sit to stand.



Fig 14: Supine to side lying with or without partial assistance

The facilitation of supine to side lying & side lying to supine is achieved by strengthening of lateral trunk flexors, abdominals, back extensors & assisting the dissociation of trunk over pelvis. (Fig 14)



Fig 14: Training of side lying to sit with partial assistance

Strengthening of triceps, shoulder rotators, concentric contraction & eccentric control of lateral trunk flexors on contralateral & ipsilateral side respectively along with strengthening of hip abductors & extensors to achieve side lying to sit & vice versa.

The development of poor sitting posture or postural scoliosis of spine (curvature of spine) is due to poor co- activation in truncal muscles. (Fig 15)



Fig15: Scoliosis

To facilitate erect posture or sitting without support or with self-assistance, strengthening of abdominals & back extensors, trunk lateral flexors, stretching of shortened trunk lateral flexors due to scoliosis is advised.(Fig 16). The co-contraction of abdominals & back extensors is achieved by stabilizing reversal of PNF technique in sitting to improve sitting posture.



Fig 16: Stretching of lateral trunk muscles to reduce scoliosis

The facilitation of truncal control is also achieved by attaining the quadruped & kneeling position. Exercises in quadruped such as cat & camel exercises to facilitate, activate & strengthenscapular muscles, abdominal & back extensor muscles. (Fig 17) The co- activation of abdominals&back extensors, hip abductors & extensors, co-activation of knee flexors & extensors is achieved to attain the kneeling position.

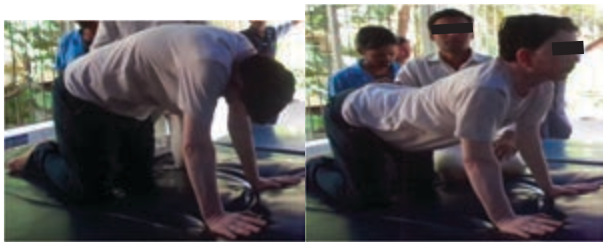


Fig 17: Cat & Camel exercise in Quadruped position

Muscle weakness in SMA will eventually result in spinal deformity, joint contracture, increased pain. In order to prevent further increase in the spinal deformity (scoliosis), spinal jacket is also advised. (Fig 18)



Fig18: Spinal Jacket to prevent scoliosis

In SMA patients, bone density is compromised due to osteoporosis. Facilitation of weight bearing on lower extremity is to maintain the bone density, to prevent joint contractures & to facilitate joint proprioception. Therefore, this is achieved by standing. In non- ambulatory patients, standing is achieved by standing on standing frame or tilt table. (Fig 19)



Fig 19: standing in standing frame or tilt table using spinal jacket with walker

In patients with late ambulatory phase, gait training is achieved in parallel bar using calipers (KAFO) if needed (Fig 20).



Fig 20: Gait training in parallel bar

Here, the patient is taught to eliminate the compensatory mechanisms used while walking. The correct pattern is encouraged with proper facilitation & activation of postural muscles. If the muscle power is appropriate, then the patient is encouraged to walk with the help of walker.

In case of non- ambulatory patients, apart from standing, patients are encouraged for wheelchair mobility in & out of the house (Fig 21).Along with this, patients are advised about energy conservation techniques in order to reduce fatiguability during the activities or exercises.



Fig 21: Wheel-chair mobility

Therefore, patients with SMA are encouraged & motivated to keep themselves active in the form of physical exercises which will help them to improve their level of independence.

## Occupational Therapy

The occupational therapist plays an important role in addressing the individual needs of patients with spinal muscular atrophy. An occupational therapist will help the individual to be independent in his or her activities of daily living. Fine motor skills are usually affected and it may be due to fatigue. School-age patients that are affected, may benefit from an occupational therapy consultation. They will help in improving the keyboarding skills and other ways to avoid fatigue from upper extremity activities in the classroom.

Multiple assistive devices are available but the patients may eventually require the use of a wheelchair on a full-time basis. It will help to maintain the higher level of independence.

## Therapeutic exercise

The role of exercises in spinal muscular atrophy is to assist in improving function, independence, flexibility and quality of life. People with SMA can exercise by practicing the movements and tasks they want to perform throughout the day. This type of practice is called as functional exercise. It can consist of any movement or position one would like to strengthen in order to improve their activities of daily living, for example reaching, sitting, rolling or may be even standing or walking. These activities are dependent amount of neuromuscular involvement, developmental stage and age of the person. Excessive exercising may lead to fatigue and damage. Energy conservation and work simplification techniques are to be addressed prior to setting of fatigue.

## Range of motion

Range of motion (ROM) is the of movement of our joints. We maintain our ROM though stretching and exercise. To avoid our muscles and soft tissues to get tight, occasional ROM exercises are to be repeated. If the tightness becomes permanent and movement becomes limited, it can lead to contractures.

Contractures, can cause discomfort, decreased mobility, and impaired flexibility of the associated body part. It hampers the normal functioning of the joint.

To prevent contractures, hand splinting can be used. Functional hand splint, resting hand splint, knuckle bender are few of the examples of commonly used hand splints to prevent contractures. The use of splints and braces are recommended to maintain joint alignment and prevent tightness and contractures.

## Weight bearing

Weight bearing is important for the health of our bones and muscles. It improves bone strength, respiratory and bowel function. This is even for children who are unable to bear weight independently. Standing frame helps the children maintain the standing position.

There are several different types of standing frames. They can address both prone position and in supine position. Supine standing frames are recommended for children without adequate head control.

In addition to the weight bearing benefits, standing frames often allow children to be on eye level with their peers, which has psychosocial advantages. Some standing frames can accommodate for the development of tightness in the leg by transitioning from sitting to standing to position.

1. Supine standing frame

2. Prone standing frame

3. Sit-to-stand standing frame

In order to determine which assistive device is most appropriate, one must consider the individual's size, amount of assistance needed, and space available in the home and at school.

## Walkers

Walkers can provide balance and arm support.For children who are able to walk when given external support, the use of assistive devices may improve their ability and reduce the dependence from the caregivers.Walker with wheels (rolling walkers) assist the children with mobility. The type of walker can be either posterior or anterior. A gait trainer can provide bodyweight support with assisted walking. The therapists usually assist with deciding

on the appropriate type of walker for the child.

## Orthotics: Splints/Braces

Orthotics are usually designed to support and align the foot. Devices vary depending on the amount of support a child needs.

Orthotics are modified and typically custom-fit as the child grows and/ or progresses. Children with SMA can be prescribed a SMO (supramalleolarorthosis) or an AFO (ankle-footorthosis). These braces are lightweight plastic designs that fit inside a shoe and help maintain foot alignment when the child is standing or walking. KAFO (knee-ankle-foot orthosis), HKAFO (hip-knee-ankle-foot orthosis), and RGO (reciprocating gait orthosis) can be used to help with optimal positioning and alignment in standing.

## MOBILITY

## Wheelchair

A wheelchair is a mobile chair. There are many types and brands available. Wheelchairs can either be power or manual. Power chairs are wheelchairs that are controlled electronically, typically by a joystick. The joystick is usually located at the dominant hand, if muscle strength is not up to the mark, the controls can be changed or modified. Power wheelchairs may offer children, who cannot manipulate their chair with their arms, the ability to move without help from others. Manual chairs can be manipulated by the individual in the chair or pushed by a caregiver. The burden on the care givers increases if the wheelchairs are manual.

## Adaptive tricycles

Adaptive tricycles may give your child the freedom to move and engage in age appropriate play activities with peers. It can also be used as a fun form of exercise!

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# Chp. 22 Motor Neuron Disease

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MND is also known as Charcot's disease, and Lou Gehrig's disease or Amyotrophic lateral Sclerosis. It is characterized by progressive degeneration of anterior horn cells of the spinal cord causing lower motor neuron type of weakness; the corticospinal tracts causing upper motor neuron symptoms; and certain motor nuclei of the brain stem leading to bulbar symptoms. It is one of the most devastating types of neurological disorder, with no known cause.

**Prevalence :**

The prevalence of this disorder is 2 per 1,00,000 and there is a male to female ratio of 2:1

In about 10% of cases, ALS is caused by a genetic defect. In the remaining cases, the cause is unknown.

There are no known risk factors, except for having a family member who has a hereditary form of the disease.

**MND manifests in different forms like:**

1. Amyotrophic lateral sclerosis (ALS)
2. Progressive muscular atrophy (PMA)
3. Progressive bulbar palsy (PBP)
4. Primary lateral sclerosis (PLS)

Amyotrophic lateral sclerosis (ALS) can be defined as a neurodegenerative disorder characterized by progressive muscular paralysis reflecting degeneration of motor neurons in the primary motor cortex, brainstem and spinal cord. It usually begins at the age of 40 years of age however, the prevalence is now shifting in the early ages as well. "Amyotrophy" refers to the atrophy of muscle fibers, which are dennervated as their corresponding anterior horn cells degenerate, leading to weakness of affected muscles and visible fasciculations. "Lateral sclerosis" refers to hardening of the anterior and lateral corticospinal tracts as motor neurons in these areas degenerate and are replaced by gliosis. Thus ALS exhibits mixed presentation of UMN and LMN symptoms which is the diagnostic criteria.

The syndrome of progressive muscular atrophy (PMA) accounts for 5-10% of patients with MND, and indicates a pure lower motor neuron syndrome without accompanying upper motor neuron signs. There is gross wasting and weakness, along with fasciculations in small muscles of all four limbs, leading to foot drop and reduced grip strength. Unlike ALS, this disease progresses very gradually upto 10-15 years.

Primary lateral sclerosis (PLS) is a clinically progressive pure upper motor symptoms that cannot be attributed to any other disease process. Patient may have involvement of both corticospinal and corticobulbar paths. Pathologically there is loss of pyramidal cells in the precentral gyrus and cerebral cortex, with degeneration of corticobulbar and corticospinal tracts.

Progressive Bulbar Palsy is a condition with involvement of motor nuclei in the lower brain stem. There is primary involvement of the bulbar muscles like the jaw muscles, facial muscles, tongue, larynx and pharynx. Hence patient suffers from dysphagia and dysarthria. As muscles of

mastication and deglutition are affected the food particles get stuck to one corner of the mouth Weakness of the pharyngeal muscles causes improper pushing of food particles into oesophagus giving rise to choking. Fibrillation of tongue is a common symptom. It is a fast progressive disorder leading to death with in a span of 2 years of beginning of symptoms and mainly due to respiratory failure.

| Subtype | Nervous System Pathology |
|---|---|
| Amyotrophic Lateral Sclerosis | Degeneration of the corticospinal tracts, neurons in the motor cortex & brain stem & anterior horn cells in the spinal cord. |
| Primary Lateral Sclerosis | Degeneration of upper motor neurons |
| Progressive Bulbar Palsy | Degeneration of motor neurons of cranial nerves IX to XII |
| Progressive muscular atrophy | Loss or chromatolysis of motor neurons of the spinal cord & brainstem. |

(Reference: Rowland, LP:Diverse forms of Motor neuron disease.Adv Neurol 36:1, 1982.)

**Clinical Features:**

The features of ALS were first clearly described as a clinico-pathological entity by Jean Martin Charcot in 1869 Approximately two thirds of patients with typical ALS have a spinal form of the disease (classical 'Charcot ALS'). They present with symptoms related to focal muscle weakness where the symptoms may start either distally or proximally in the upper limbs and lower limbs. Patients may have noticed fasciculations (noticed as involuntary muscle twitching) or cramps preceding the onset of weakness or wasting for some months (or years). The weakness is usually of insidious onset, and patients may notice that symptoms are exacerbated by cold weather. Although it is usually asymmetrical at onset, the other limbs develop weakness and wasting sooner or later, and most patients go on to develop bulbar symptoms and eventually respiratory symptoms.



Fiure 1: weakness and wasting of thenar and hypothenar muscles.

Patients with bulbar onset ALS usually present with dysarthria. Rarely, patients may present with dysphagia for solid or liquids before noticing speech disturbances. Limbs symptoms can develop almost simultaneously with bulbar symptoms and in the vast majority of cases will occur within 1-2 years.

**Common Impairments Associated with MND**

Almost all patients with bulbar symptoms develop sialorrhoea (excessive drooling) due to difficulty in swallowing saliva and mild UMN type bilateral facial weakness which affects the lower part of the face. 'Pseudobulbar' symptoms such as emotional lability and excessive yawning are seen in a significant number of cases.

| Type of Impairment / Location | Clinical Manifestation of Pathology |
|---|---|
| Impairments related to LMN pathology | Muscle weakness, hyporeflexia, hypotonicity, atrophy, muscle cramps, fasciculations. |
| Impairments related to UMN pathology | Spasticity, Pathological reflexes, hyperreflexia, muscle weakness. |
| Impairments related to bulbar pathology | Dysphagia,dysarthria,sialorrhea,peudobulbar affect. |
| Respiratory Impairments | Exertional dyspnea, nocturnal respiratory difficulty, orthopnea,hypoventi lation. |
| Other Impairments | Dementia, Cognitive Impairments |
| Rare Impairments | Ocular Palsy, bowel & bladder dysfunction. |
| Indirect & Composite Impairments | Fatigue, weight loss,cachexia,decreased ROM,pain,gait disturbance,depression,anxiety,-deconditioning,balance & postural control impairments |

On examining the cranial nerves, the jaw jerk may be brisk, especially in bulbar-onset disease. An upper motor neurone type facial weakness affects the lower half of the face causing difficulty with lip seal and blowing cheeks, but often varying degrees of UMN and LMN facial weakness coexist. The gag reflex is preserved and is often brisk while the soft palate may be weak. Patients develop fasciculations and wasting of the tongue, and tongue movements are slowed due to spasticity. Sensory examination is almost always normal. Respiratory failure and other pulmonary complications are the usual cause of death in ALS. Summary of Revised El Escorial Research Diagnostic Criteria for ALS (Brooks et al., 2000)

**The diagnosis of ALS requires:**

1. Evidence of LMN degeneration by clinical, electrophysiological or neuropathological examination;
2. Evidence of UMN degeneration by clinical examination, and
3. Progressive spread of symptoms or signs within a region or to other regions, as determined by history or examination,

**Together with the absence of:**

1. Electrophysiological and pathological evidence of other disease that might

explain the signs of LMN and / or UMN degeneration, and

2. Neuroimaging evidence of other disease processes that might explain the observed clinical and electrophysiological signs

UMN signs: Clonus, Babinski sign, absent abdominal skin reflexes, hypertonia, loss of dexterity.

LMN signs: atrophy, weakness. If only fasciculation: search with EMG for active denervation. Regions reflect neuronal pools: bulbar, cervical, thoracic and lumbosacral.

**Pathogenesis of motor neuron degeneration in MND**

Most ALS cases are sporadic but 5-10% of cases are familial, and of these 20% have a mutation of the SOD1 gene and about 2-5% have mutations of the TARDBP (TDP-43) gene. Two percent of apparently sporadic patients have SOD1 mutations, and TARDBP mutations also occur in sporadic cases. The diagnosis is based on clinical history, examination, electromyography, and exclusion of 'ALS-mimics' (e.g. cervical spondylotic myelopathies, multifocal motor neuropathy, Kennedy's disease) by appropriate investigations. The pathological hallmarks comprise loss of motor neurons with intraneuronal ubiquitin- immunoreactive inclusions in upper motor neurons and TDP-43 immunoreactive inclusions in degenerating lower motor neurons. Signs of upper motor neuron and lower motor neuron damage not explained by any other disease process are suggestive of ALS. The cell mechanisms that lead to degeneration include glutamate excitotoxicity, Oxygen reactive species toxicity.

The clinical diagnosis is confirmed by Electromyography (EMG) and muscle biopsy. On EMG, the motor neuron conduction velocity is normal until late in the disease, but the amplitude is reduced. On needling, the mechanical stimulation of the needle causes fibrillation potentials, and spontaneous fibrillation and fasciculation potential are seen when the needle is stationary in relaxed muscle, with greater duration and amplitude of the action potentials. When the muscle contracts there is marked decrease in the number of spikes leading to incomplete interference pattern.

**Examination:**

1. Joint Integrity, Range of Motion and Muscle length: is examined using standard tools.
2. Pain : seen in ALS is assessed by Visual Analog Scale (VAS)
3. Muscle Performance: is examined by Manual Muscle Testing (MMT), isokinetic muscle strength testing or hand -held dynamometer.
4. Motor Function: Due to Spasticity, and weakness of muscles there could be many manifestations like Impairments in dexterity, incoordination of both gross and fine movements as well as loss of motor control. Therefore Functional assessment of both upper and lower extremities should be done.
5. Tone and Reflexes: Tone can be assessed by Modified Ashworth Scale and reflexes by deep tendon reflexes.
6. Cranial Nerve involvement should be assessed. Pseudo Bulbar and Progressive Bulbar varieties of MND only show involvement of cranial nerves.
7. Postural malalignment and imbalance are seen which can be assessed by Tests like Tinetti Performance Oriented

Mobility Assessment (POMA), Berg Balance Scale, Timed Up andGo Test and Functional Reach Test

8. Gait: Deviations due to muscle imbalance should be assessed, so also endurance.
9. Respiratory Function: There could be involvement of respiratory muscles resulting into breathlessness, Low vital capacity and lack of cough effectiveness. Therefore Respiratory Function evaluation should be done in detail by using a hand-held spirometer. Aerobic capacity and cardiovascular pulmonary endurance should also be tested to evaluate aerobic conditioning.
10. Because of being in bed for long time without mobility there are chances of getting trophic ulcers: periodic skin inspection should be done.
11. Functional Status: Functional Independence Measure (FIM) can be used to document functional status.
12. Environment Barriers: should be considered for easy accessibility and safety.
13. Fatigue: Fatigue Severity Scale to be used.
14. ALS Functional Rating Scale (ALSFRS): The functional status of ALS patients can be rated by ALS Functional Rating Scale (ALSFRS) and revised version ALSFRS-R It correlates with muscle strength of both upper and lower limbs. ALSFRS- R includes respiratory muscles measures also.

**Conventional therapies:**

The management of ALS / MND has considerably changed over the past two decades, with an emphasis on coordinated multidisciplinary care between specialist, like physician, speech therapist occupational therapist, physical therapist, dietician and nurses. Advanced directives are on end of life care, respiratory and nutritional management during late stages of life are focused on.

Riluzole is the only approved drug that has been shown to have a modest effect on prolonging life in ALS patients. The mechanism of action of riluzole is not entirely certain but is thought to include interference with N-methyl-D-aspartate (NMDA) receptor mediated responses, stabilisation of the

inactivated state of voltage-dependent sodium channels, inhibition of glutamate release from pre- synaptic terminals, and increasing of extracellular glutamate uptake, thereby preventing degeneration of motor neurons.

Treatments to control symptoms are also helpful:

- Baclofen or diazepam may be used to control spasticity that interferes with daily activities.
- Trihexyphenidyl or amitriptyline may be prescribed for patients with problems in swallowing their own saliva.
- Choking is a common symptom. Patients may decide to have a tube placed into their stomach for feeding. This is called a gastrostomy or Percutaneous Endoscopic Gastrostomy (PEG). A nutritionist's help is very important for them as they tend to lose weight. The illness itself increases the need for food and calories. At the same time, problems with swallowing make it

difficult to eat enough.

**Physiotherapy management**

**The efficacy of therapeutic interventions is related to:**

1. Timing of interventions,
2. Motivation and persistence of patient in carrying out the program.
3. Support from family members.

Rehabilitation intervention plan depends on the following:

1. The rate of progress of the disease
2. Presence of spasticity, bulbar involvement, respiratory involvement causing hypoxia and fatigue.
3. Phase of Disease. Exercises are to prescribed according to level of impairment, functional limitation and level of disability

**Phase I :** Patient is independent in all ADL, is ambulatory, except for mild weakness and clumsiness in movements.

In case of mild weakness advice is to continue normal activities. In case of clumsiness, stretching exercises like Yoga In case of ambulatory patients, gentle resisted exercises with caution not to fatigue or over work.

**Stage 2:** Patient presents with moderate selective weakness in ankles, wrist and hand

**Advice :** Stretching exercises to avoid contractures.

In case of decreased independence in ADLs like climbing, overhead activities and difficulty in buttoning etc, strengthening exercises to be prescribed avoiding fatigue.

In case of difficulty in Ambulation, Orthotic devices like AFO, hand splints to be considered.

**Stage 3:** Patient presents with fatigability in long distance ambulation, needs supervision in ADL.

**Advice :** Continue stage 2 and emphasize on deep breathing exercises to be added.

In case of non-ambulatory patients, consider wheelchair, (standard or motorized).

**Stage 4:** Patient may present with additional pain and edema in hand and feet,

**Advice :** Consider modalities like massage, elevation and active exercises.

In case of severe weakness in extremities, caution is to be taken to support the joints while doing rotations.

In case of Fatigability in ADLs, encourage isometric upto level of tolerance and to consider slings or arm support, motorized chairs etc.

**Stage 5:** Patient presents with severe lower extremity weakness,

**Advice:** Emphasize on teaching family members proper techniques of transfer and positioning of patients limbs.In case of severe upper extremity weakness, consider modifications at home like high dinning table to facilitate eating etc.

**Stage 6 :** In case of totally bedridden patients with dysphagia, consider suction, soft diet, tube feeding, PEG feeding etc.In case of severe breathing difficulty, frequent clearing of airways, tracheotomy and respiratory support if needed.

**The research evidence suggests:**

1. Overuse weakness does not occur in muscles with MMT grade 3(fair) or greater out of 5(normal).
2. Moderate resistance exercises can increase strength in muscles with a MMT grade3 or greater out of 5.
3. Strength gains are proportional to initial muscle strength.
4. Heavy eccentric exercise should be avoided.
5. Exercises may produce functional benefits.
6. Psychological benefits have yet to be determined.

**Patients with severe respiratory and bulbar complications may not benefit from active exercise programs. The goal in end stage is to optimize health and increase quality of life.**

- Secondary complications in the form of pressure sore and contractures should be avoided and when the patient goes into respiratory failure, chest physiotherapy becomes necessary. The therapist should make all efforts to maintain the chest expansion of the patient by teaching and supervising breathing exercises in the form of incentive spirometer.
- Proper positioning of patient is necessary because once the limbs become weaker and wasting progresses, it becomes increasingly difficult to maintain upright position for the patient. This difficulty is noted while standing, sitting in wheel chair or on bed, hence proper positioning is necessary. At times patients also develop neck drop. In order to minimize the effect of gravity on the body, it is advised to incline the patient back from vertical position, as in this position the line of gravity passes in front of head and neck through thorax. It also causes relief of pressure of thorax on abdomen, allowing diaphragm to work efficiently and aid in breathing with ease, as their breathing is restricted due to intercostal muscle weakness.

**Occupational therapy management**

Occupational therapists work with people with MND and their families providing advice, support and guidance during their journey. The role of an occupational therapist is to work with clients to maximize their level of independence in day-to-day activities 2

**When a condition such as MND impacts their independence, occupational therapists give them hope through:**

- Energy conservation, activity pacing and workplace modifications are advised to retain employment as long as possible
- Fatigue management is explained to conserve energy, prioritize work according to levels
- Ensuring the correct seating posture and a wheelchair is provided to maximize function and independence
- Support with providing a source for the right equipment to help with difficulties as they arise, such as beds or chairs
- To preserve independence as long as possible, an occupational therapist works with a client to identify the most suitable environmental controls for them i.e. the control of functions in the home such as opening curtains, turning on lights or adjusting music or TV settings to be controlled by the client

from their wheelchair, armchair or bed

- Recommendations given to ensure that the home environment will meet both current and future needs are given 2
- Coping with difficulties in standing or maintaining posture for a sufficient length of time
- Stairlifts or through-floor lifts can be used to conserve energy (Figure 1)
- Positioning aids and cushions to maximize comfort and relieve pressure 2

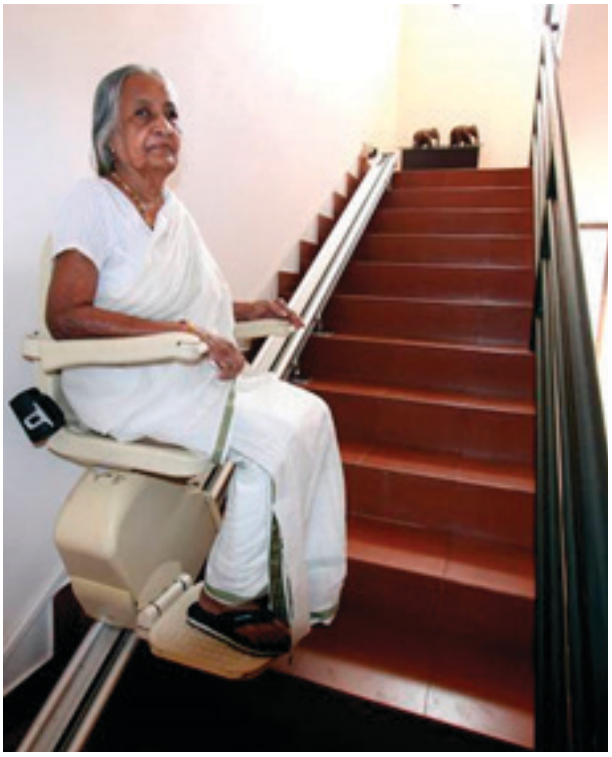


Figure 1: Staircase lift

**Assessment and outcome measures**

Occupational therapists assess the strength of the patient, their skills and requirements to carry out their occupations and the environment in which activity takes place.3

**Standardized Assessment Instruments**

A. Revised ALS functional rating scale (ALS-FRS R)

This is a questionnaire based scale for clients with ALS to measure disease progression. It has 4 domains:

1) Bulbar function i.e speech, salivation and swallowing
2) Fine motor tasks i.e writing and feeding
3) Gross motor tasks i.e.walking, climbing stairs and bed mobility
4) Respiratory functions

It has good inter rater reliability and validity.3

**B) Quality of life assessment in ALS**

It consists of general or disease specific health status questionnaires. It is used due to its practical feasibility and has high internal validity and consistency in reliability.4,5

C) Functional Independence measure (FIM) is used for patients who are undergoing rehabilitation.

D) Mini mental status examination for cognitive impairment6

E) Tinetti Performance Oriented Mobility Assessment (POMA), Berg balance scale and functional reach test for postural alignment, control and balance6

F) Dynamometer is used to measure grip strength and pinchometer to measure pinch

**Non standardized assessments**

1) Range of motion
2) Manual muscle testing
3) Informal Activities of daily living (ADL) and instrumental activities of daily living (IADL) assessments
4) Assessment of activity tolerance and limitations due to fatigue

As the disease progresses re-evaluations are necessary to see if there is any functional

decline in clients.

Early-stage goals for patients with ALS are

- Optimize strength and range of motion
- Decrease pain and fatigue
- Use joint protection and pain management strategies

**Later stage goals include**

- Positioning
- Ensure safety
- Environmental modifications
- Maintain skin integrity

**Management of MND**

**Posture**

Due to the progression of the disease and increasing weakness, it becomes difficult for people to sit erect against gravity. This may lead to poor posture, difficulty in shifting positions leading to pressure sores. Muscles of neck and trunk get affected which leads to kyphosis. Wheelchair with arm support, adjustable back, neck and head support, pressure-relieving seat and armrest is required.

**Orthosis for people with MND**

Upper extremity orthosis

Wristcock-uporthosis,restinghandorthosis, short and long thumb spica, neoprene orthosis or simple web spacer may be used .7

**Lower limb orthosis**

foot up orthosis for wearing along with shoes

spring leaf AFO

**Mobility**

Active and/or passive exercises maintain good posture and mobility in patients with MND. In initial phases, people with MND can use mobility aids such as sticks, walkers and rollators to support walking. Grab bars can be installed in a home where sticks or walkers cannot be used Figure 2.



Figure 2: Grab bars instead of a walker



Figure 3: Vehicle modifications

But as the disease progresses people may have difficulty moving around using walking aids as well, so they may require a wheelchair. In the presence of upper extremity weakness, powered or electric wheelchair can be used to conserve energy. Floor to ceiling pole and mechanical lift can also be used.

For outdoor mobility, adaptations are also required in vehicles Figure 3.

**Pressure sore management**

Pressure-relieving mattresses that are gel, water or air-filled should be used. Avoid using memory foam as the person may find it difficult to move in it. An electric blanket in winter may be used.

**Head and neck support**

Weakness in neck, shoulder and back extensor muscles leads to neck drop and difficulty in lifting neck up for eating, speaking and breathing. Wheelchair with recliner and propelling bed is used to keep head and trunk in proper alignment. Proper arm support is required to avoid forward curvature of the spine and helps in opening chest cavity while breathing.



Figure 4: Neck support

**Transfer**

Rails and bars should be installed in the home and toilet to help in transfers. Static or powered pillow lift should be used.

**Respiratory function**

There is an increased risk of aspiration, weak cough and abnormal respiratory patterns due to bulbar involvement. In later stages of the disease, diaphragm and accessory muscles may be affected. This is the main cause of death in MND. Patients may have poor sleep, day time fatigability, breathlessness or reduced vocal power. Breathing exercises like pursed lip breathing and deep breathing is adviced.

**Eating**

Non-skid mats to avoid slippage of plates, modified spoons and knives for grip and control, modified cups, mugs, plates and bowls should be used. Proper head and neck support for ease in swallowing should be used. To clear mouth debris, portable suction can be used.

**Toileting**

Non-skid mats in bathroom or bathtub to avoid falls, bath seat that has a built-in tilt-in-space shower seat, grab bars for holding, manual or powered hoist to transfer in and out of the bath, raised commode seat or chair should be use 8 fig 5.3 d).

**Dressing and grooming**

Hand muscles weakness leads to reduced grip strength and poor grasp. Dressing stick, button hooks or velcros and elastic to ease dressing can be used. Lightweight fabrics and loose-fitting clothes are used to reduce frustration while dressing and undressing clothes. Shoe horns and sock aids and leg lifters may be used. Shoes should be wide if the person has a foot drop, to accommodate anti foot drop splint.

**Safety at home**

Mobile phone or smart watch with mobile functions should be placed close to the person, environmental controls such as alarm functions or with door entry and lights should be installed at home.

Social and psychological support is needed as the condition can lead to depression, anxiety, stress and fear. So coping skills

based on problems can be taught.

Sleep Hygiene: Guidance regarding sleep patterns is given. Patient should take bath before bed time for better sleep.

**Client and Caregiver education**

Educate patient and caregiver regarding positioning, transfer, dressing, bowel and bladder management, handling techniques, proper body mechanics to avoid deformities.

## Psychological Interventions in Motor Neuron Diseases

Motor neuron diseases is a progressive disease with an impact on physical and psychological wellbeing. The self-reported measures completed by MND clients reported that every day functioning account for the incidence of depression, low self-esteem. (1) Motor Neuron diseases causes psychological distress and demands emotional support as a priority. The IHF Strategic Plan ensures continues support and high-quality care for people with life limiting illness. (2)

**The psychological impacts of Motor Neuron Diseases:**

Motor Neuron disease possess serial emotional changes that occur as per the difficulties encountered while executing daily duties. The serial emotional changes occur on daily basis of which 0 to 30% are affected by anxiety and 9 to 11% are affected with depression (3), 20 to 49 % possess emotional liability (4), felling of hopelessness with greater suicidal considerations being linked to their sufferings (5,6,7).

Existential Shock ie losing of confidence in their bodies, recognizing their physical vulnerability, scared of facing death (8) are the result of initial stages of acceptance. Demoralization could occur by interacting with people with similar disease and greater knowledge of the disease consequences. (9) Hence episodes of hopelessness, demoralization, anger are argued to be more felling of depression (2).

Therapy Pre- Requisites:

Time frame: Scheduling the client to allow extra time when working with persons with MND due to fatigue, Communication impairment etc.

Environment: Providing a quiet space with appropriate access or therapy at home due to limitation of mobility should be considered.

Knowledge of MND:Understating the clinical condition allow future prediction and better clientele approach and better interventional plan.

## Therapeutic Approaches:

**Promote Self Realization:**

The understanding of who they really are and understanding their needs and need of fulfillment is self-realization. The term is more philosophical and psychological theories based that promote mental health.

**Promote Self Direction:**

Self-direction follows a principle of recovery, independence, self-sufficiency and choice. It recognizes that all individuals can achieve goals, basic human needs for fulfillment as interests of preference. For eg people with self-direction would involve in funded projects, purchasing good and providing services.

**Pacing:**

Pacing is an energy management strategy to manage exertion while performing activities. There are two forms: symptom-contingent pacing, where the decision to stop is determined by an awareness of an exacerbation of symptoms. Thus the principle behind pacing is to avoid over-exertion and an exacerbation of symptoms. It is not aimed at treating the illness but management of energy for task execution.

**Coping Strategies:**

Coping is the conscious effort to reduce stress and the techniques used to is termed as coping strategies. They are leaned from watching or advices taken to avoid stressful situations. A good coping skill can be developed by practice. The usual techniques are to perform progressive relaxation exercises, breathing technique, emotional regulation, diverting oneself etc.

**Family Counselling:**

Family play a significant role in support, motivation, maintaining independent living and as acts as a primary caregiver. Its debilitating and a difficult situation for the family to face their loved ones suffering with an incurable diseases. They face stressful and difficult situation quite often. A family counselling that brief the clinical condition and promote acceptance of the condition may lead to better care giver metal heath and reduce burden.

**Conclusion:**

Psychological intervention can promote heath related quality of life of the client and the family by providing assistance for coping and accepting a long term disability.

Speech and swallowing therapy management for Motor neuron disease

**SWALLOWING DISORDERS IN PERSONS WITH MND (more specific to ALS)**

**Persons with ALS may complain of :-**

- Difficulties with chewing and swallowing which generally follow speech impairments by several months
- Swallowing disorders which may occur first either with solids or liquids
- Reduced tolerance to certain foods leading to strong avoidance of some foods and preference for others
- Reduced intake of foods due to reduced choices as well as increased effort in eating and swallowing
- Longer time to complete meals than others or in comparison to his own time prior to onset of disease
- Significant loss of weight
- Reduced pleasure to severe anxiety associated with mealtimes

**Signs for Oral preparatory stage of dysphagia as seen by the SLP**

- Poor lip closure
- Drooling
- Dribbling of saliva and /or food
- Poor food manipulation
- Difficulty using a straw
- Pocketing of food in the cheeks
- Poor mastication of food
- Partially chewed food or liquid going into airway by gravity prior to initiation of swallow (Premature spillage)

- Poor propulsion of food due to weakness of tongue

Signs for oral stage dysphagia :

- Absence of /or delayed triggering of swallow reflex
- Poor propulsion of food into the pharyngeal inlet

Signs for pharyngeal stage dysphagia:

- Incoordinated swallow
- Nasal reflux
- Delayed pharyngeal reflex
- Incomplete opening of UES
- Entry of food into laryngeal inlet
- Coughing due to penetration or aspiration of food/ liquid particles
- Reddening of eyes
- Watering of eyes
- Spiked fever
- Mouth breathing
- Choking
- Weight loss more than 5% of average bodyweight in one month without any other reason accounting for it
- H/o Aspiration pneumonia

Other factors influencing feeding/ swallowing

- Loss of hand function or reduced use reduces self-feeding abilities which may reduce volume of meals
- Fear of choking and depression may further interfere with oral intake (Slowie,Paige and Antel, 1983; Kasarskis, Neville,1998 as cited in Francis and others,1999)

(Logemann, 1983) (Robbins, 1987 as cited in Francis and others,1999)

Nutrition in ALS

- All patients with ALS eventually experience difficulty maintaining adequate nutrition and hydration thus not meeting their energy needs (Slowie,Paige and Antel, 1983; Kasarskis, Neville,1998 as cited in Francis and others,1999)
- Dehydration contributes to malaise, excessive fatigue, and thickened secretions (Francis and others, 1999)
- When swallowing is affected due to respiratory issues, noninvasive methods of assisted ventilation can allay dysphagia and delay resort to gastrostomy
- Weight loss of more than or 5% of body weight may indicate need for percutaneous endoscopic gastrostomy (PEG) or jejunostomy with former having greater risk of regurgitation and aspiration, needing greater attention to positioning during and after feeding and latter having increased risk for cramps and diarrhea

## MANAGEMENT OF SWALLOWING DISORDERS

- Patients with MND should be encouraged to eat where they feel relaxed and comfortable, as slowness of eating and dribbling during eating may cause severe embarrassment to them.
- It may be possible to work on improving the head posture and head support using a neck collar as well as lip closure.
- Application of ice in mouth may reduce spasticity of the tongue.
- Avoidance of food and drink which precipitate coughing and choking such

as spicy foods or thin liquids.

- Avoidance of dairy foods or proprietary thickeners as they may increase volume and tenacity of mouth secretions (Enderby, 1987 as cited in Leigh and Ray-Chaudhuri,1994) and could be avoided.

Management of oral stage dysphagia

- Lip ROM exercises in patients at early stage of disease to improve labial seal
- Tongue ROM exercises in patients at early stage of disease to improve tongue movements for mastication and propulsion of food
- Posture could be sitting upright with head erect position or slightly bent forward during feeds and in later stages as disease advances, posture can be leaning back but neck needs to be supported by caregiver and kept as erect as possible to prevent penetration /aspiration of food into airway
- Giving softer foods reducing effort for mastication
- Small morsels, slow and repeated chewing to facilitate cohesive bolus formation
- Repeat swallows to clear the oral cavity of food
- Submandibular massage in an anterior to posterior direction, to facilitate initiation of swallow

Management of pharyngeal stage dysphagia

- To primarily ensure healthy respiratory status prior to giving trial or therapeutic feeds
- Until then, the swallowing therapist to work on indirect swallowing therapy by improving posture, and mobility of structures required in swallowing and training patient to use airway safety maneuvers such as supraglottic swallow, Mendelsohn's maneuver etc.
- Posture same as given earlier for management of oral stage dysphagia
- Head to be tilted forwards while swallowing for increasing safety of swallow
- Semisolids and thick liquids for oral intake as thin liquids may cause aspiration
- Water and liquids given with thickeners used with advice of the swallowing therapist
- Patients taught to cough voluntarily after swallow to prevent aspiration of food particles in mouth and throat into airway
- When coughing may be difficult, to phonate /ah/ as loudly as possible for same result
- To continue with feeds if none of the following features are seen:- frequent coughs, discomfort, facial grimaces, watering from eyes, changes in breathing pattern, delayed cough, persistent wet gurgly voice quality
- Small frequent meals of the consistency best tolerated by patient, safest being semisolid consistency

The caregivers of the person with MND/ ALS needs to ensure safe swallowing till as best as they can through oral feeds. When swallowing is no longer safe through oral means, they should gradually help the person with MND/ALS accept PEG as a means for ensuring nutrition and hydration. There needs to be due monitoring for, maintenance of safe airway ; weight of the patient; regular suctioning

of secretions, so that he/she may have an optimal QOL as best as can be assured, in the circumstances.

In conclusion, as expressed best by Francis, Bach and DeLisa, (1999), ' The rehabilitation of patients with MND can be as rewarding as any other difficult mission in medicine if health care providers will regard these patients as individuals living with , rather than dying from, their disease.

**NON-SPEECH CLINICAL FEATURES OF THE ORAL MECHANISM**

**Features of spasticity: Those persons with spasticity show hypertonicity in some of the classic features given below:-**

- Jaw jerk
- Sucking reflexes- These are pathological when seen beyond 2 years of age.
- Hyperactive gag reflex
- Slow orofacial movements
- Pseudobulbar affect- This is characteristic of emotional lability where the person may have bouts of laughter or fits of crying or strong emotional outbursts which are characteristic of bilateral lesions in the upper motor neuron (UMN)
- Features of flaccidity (weakness) are as follows:-
- Gag reflex may be reduced or hypoactive
- Weak cough
- Reduced tone in facial muscles
- Lingual fasciculations which are wormlike movements on the surface of the tongue and are visible to the naked eye and is a classic sign in persons with ALS.
- Atrophy or wastage or shrinking of muscles due to the disease progress.
- Fasciculations in chin and corner of lips
- Dysphagia
- Audible reflexive dry swallow (Duffy, 1995)
- Tongue atrophy and dysphagia were the two most common signs in ALS patients with reduced intelligibility. This may be due to the fact that, atrophy means fewer muscle fibers available for contraction and force of movement is reduced in all directions. Hence the muscles are weak to do a function such as speaking which requires the accurate and skilled movements of a 100 muscles for any speech sound to be produced.

These non-speech characteristics add to the severity of the underlying condition and need to be addressed while planning therapy for the speech characteristics. We may now proceed to have a brief look at the speech and communication disorders.

**Speech disorders with persons with MND**

Dysarthria may be the first manifestation of ALS (Gubbay and others,1985 as cited in Duffy,1995 p244). When dysarthria and dysphagia are the initial symptoms of ALS, they tend to remain the most functionally limiting symptoms as the disease progresses (Yorkston and others, 1993 ibid p 244). 47% of those with ALS in hospice care had reduced speech intelligibility , 25% became anarthric and only 25% were intelligible just prior to death (Saunders, Walsh, and Smith ,1981ibid p 244). Dysarthria may not be perceived as mixed at all points during the disease. It may present as either flaccid

or spastic dysarthria; when xed, one type may predominate. (Duffy, 1995)

The dysarthria of ALS may be flaccid, spastic, or most often, mixed flaccid-spastic. The overall pattern of the mixed form, as given by Duffy, (1995) is one of :-

- Labored, slowly produced speech
- Short phrases
- Grossly defective articulation
- Vowel distortion
- Hypernasality
- Nasal emission
- Strained-strangled and groaning voice quality
- Vocal harshness which has a 'wet' or 'gurgly' qualityowing to oral secretions trickling into larynx and on to the mucosal layer of the vocal folds
- Monopitch and monoloudness.

**According to Putnam and Hixon, (1984), low lung volumes may have contributed to:-**

- Reduced loudness and stress contrasts
- Short phrase
- Reduced power for coughing in patients with respiratory weakness.

**Management of speech disorders**

Francis, Bach and DeLisa (1999) have mentioned about therapy for speech in persons with ALS and Duffy, (1995) has given very succinctly about speaker oriented and communication approaches as given as an additional resource.

In Speaker Oriented approaches, the onus and responsibility for communicating effectively rests with the speaker who has to compensate for his speech disorders and modify his speech using strategies such as :-

- Slowing rate of speech
- Exaggerating articulation
- Improving respiratory efficiency through phrasing, as it is seen that if the SLP instructs the person with ALS to effectively use breath groups to accommodate spoken words, the intelligibility of speech and the quality of speech both improve
- Tongue exercises (Dworkin and Hartmann, 1979) and diaphragmatic exercises but these are to be used with caution as efficacy of exercise in neuromuscular disease is controversial

Instrumental approach can further facilitate the speaker oriented strategies by giving a device which could enhance speech intelligibility.An example of this would be the palatal lift to reduce hypernasal speech (Dworkin and Hartmann, 1979).

Communication Oriented Approachesare resorted to when the speaker can no longer speak intelligibly and needs support to augment his speech or use an alternative for speech. Here there is focus on enhancing the speech of the speaker with ALS, or by improving the medium of transmission between the speaker with ALS and his caregiver. Here a remedy such as fitting the spouse with a Hearing Aid (Hillel,1989) or by using a communication or an alphabet board, could be tried by improving the medium. Also in this approach, the listener could try various strategies such as guessing the theme of discussion chosen by the speaker of ALS, identifying topic and words used by the speaker of ALS and filling in for his communication breakdowns by stepping

in to cover awkward silences.

AAC devices, which come under this third approach, would be ranging from simple paper and pencil; alphabet / word communication boards to high tech keyboard based and computerised instruments. Switches to operate both a communication device and an environmental control unit. These devices could be designed to be activated by finger, fist, forehead or toe based on that which retains adequate movement. There could be devices allowing eye-gaze input for patients with very advanced disease (Kazandjian, 1997). Those who required augmentative communicative devices needed them an average of 3 years following diagnosis, and that such devices were used for an avg of 3 years (Silver and Kratt,1982)

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# Section 6
# Other Aspects of Rehabilitation

# Chp. 23 Spasticity

**Dr. Alok Sharma, Dr. Vishal Chafale, Dr. Amruta Paranjape, Dr. Arjun K.M.**

Lance in 1980 gave the definition of spasticity as 'Spasticity is a motor disorder characterized by a velocity-dependent increase in tonic stretch reflexes (muscle tone) with exaggerated tendon jerks, resulting from hyperexcitability of the stretch reflex, as one component of the upper motoneuron syndrome.

Upper motor neurons (UMN) originate in the brain and brain stem and project to lower motor neurons (LMN) within the brain stem and spinal cord. The lower motor neurons are of two types, both of which originate in the ventral horn of the spinal cord: (1) alpha motor neurons project to extrafusal skeletal fibers and (2) gamma motor neurons project to intrafusal muscle fibers within the muscle spindle. Due to central nervous system lesion there is a disruption of the signals sent from upper motor neurons to lower motor neurons and other interneurons.

Spasticity is one of features of Upper Motor Neuron Syndrome (UMNS) are muscle paresis or weakness caused by inability of the muscle to generate enough force, impaired motor control hindering the speed, accuracy and dexterity of movements, decreased muscle endurance and brisk or exaggerated deep tendon reflexes including spasticity, spasms and clonus. Paresis generally affects distal muscles, with loss of the ability to perform fractionated movements of the digits. The upper motor neuron syndrome results from damage to descending motor pathways at cortical, brainstem, or spinal cord levels and spasticity evolves in the days and weeks after injury. When the injury that leads to spasticity is acute, muscle tone is flaccid with hyporeflexia before the appearance of spasticity. The interval between injury and the appearance of spasticity varies from days to months according to the level of the lesion.

The stretch reflex arc is the most basic neural circuit contributing to spasticity. When a muscle is stretched, an impulse is generated in the muscle spindle and is transmitted via the sensory neuron (Ia fibers) to the grey matter of the spinal cord. Here the sensory neuron synapses with the motor neuron (alpha motor neuron), which then transmits an impulse for muscle contraction. While agonist muscles contract in response to stretching, antagonist muscles must relax. Their relaxation is brought about via an inhibitory neuron within the spinal cord. The alpha motor neuron and the muscle comprise the final common pathway in the expression of motor functions, including spasticity. Failure of this arc results in inability of inhibiting the antagonist muscles causing spasticity.

There are numerous excitatory and inhibitory modulatory synaptic influences on this pathway. An imbalance in these influences results in hyperexcitability of the stretch reflex arc, which is thought to be the basis for spasticity. Some of the factors that play a role in suppressing hyperactivity of the final common pathway include cerebral inhibitory pathways (from the brain) and spinal mechanisms such as nonreciprocal Ib inhibition (from golgi tendon organ receptors in tendons), presynaptic inhibition

of the Ia terminal (at the axoaxonic synapse between 2 axons), reciprocal Ia inhibition (inhibition of antagonistic muscles) and recurrent Renshaw inhibition (inhibitory feedback of the alpha motor neuron cell body by the inhibitory interneuron).

**Pathophysiology**

Spasticity arises from prolonged disinhibition of spinal reflexes as a result of UMN lesion. These spinal reflexes include stretch, flexor and extensor reflexes and are under supraspinal control by inhibitory and excitatory descending pathways. Stretch reflexes are proprioceptive reflexes and are either phasic or tonic. The tonic stretch reflex arises from a sustained muscle stretch and is the cause of spasticity. Stretch reflex is dependent on tendon lengthening and excitatory post synaptic potentials (EPSPs) carried by Ia afferents but Inhibitory post synaptic potentials (IPSPs) arising from antagonistic muscle spindles, oligosynaptic and polysynaptic pathways also have an important role in the maintenance of tone.

Damage to pyramidal tracts alone does not result in spasticity. It occurs only when the lesion involves premotor and supplementary motor areas. Selective damage to area 4 in the cerebral cortex of primates produces paresis that improves with time but increases in muscle tone are not a prominent feature. Lesions involving area 6 cause impairment of postural control in the contralateral limbs. Combined lesions of areas 4 and 6 cause both paresis and spasticity to develop. Physiologic evidence suggests that interruption of reticulospinal projections is important in the genesis of spasticity. In spinal cord lesions, bilateral damage to the pyramidal and reticulospinal pathways can produce severe spasticity and flexor spasms, reflecting increased tone in flexor muscle groups and weakness of extensor muscles.

Spastic dystonia is primarily due to abnormal supraspinal descending drive, which causes a failure of muscle relaxation and is sensitive to the degree of tonic stretch imposed on that muscle. There is inappropriate recruitment of antagonist muscles in spastic co-contraction upon triggering of the agonist under volitional command. This occurs in the absence of phasic stretch and is sensitive to the degree of tonic stretch of the co-contracting antagonist. For instance, triceps will be recruited during volitional action of biceps and will lead to elbow stiffness. Associated reactions are found in muscles that are not particularly stretch sensitive. They include, when there is extra-segmental co-contraction due to cutaneous or nociceptive stimuli, or inappropriate muscle recruitment during autonomic or reflex activities, such as yawning.

The resultant pattern is determined by the age, size and location of the lesion. Supra-bulbar lesions present predominantly with flexor patterns of spasticity, whereas spinal cord lesions produce extensor patterns predominately. Patients with partial lesions, where sensation is intact or partially intact, are typically bombarded by nociceptive inputs and display greatly increased ?-motor neuron activity. Different patterns emerge early on after the neurological insult and later, when patients may find themselves in a rehabilitation unit. The following figure shows the effects of the different scenarios.

Immediately after injury, a period of neuronal shock occurs and spinal reflexes are lost, which include stretch reflexes. A flaccid weakness in seen, but even during this, the positive features of hypertonia can start to be seen. Limbs are not sufficiently stretched and may be immobilized in shortened positions. Rheological changes occur within muscles in the form of loss of proteins and sarcomeres and accumulation of connective tissue and fibroblasts. Unless treated, tendon and soft

tissue contracture and limb deformity are established. Altered sensory inputs such as pain, recurrent infection and poor posture, maintain a further stimulus to lead to yet further shortening, and this cycle is difficult to break.

Spasticity is set up later on, as plastic rearrangement occurs within the brain, spinal cord and muscles. This attempt at restoration of function through new neuronal circuitry creates movement patterns based on existing damaged pathways. Neuronal sprouting occurs at many levels with inter-neuronal endings moving into unconnected circuits from decreased supraspinal command through the vestibular, rubrospinal and reticulospinal tracts. The end-effect is muscle over activity and exaggerated reflex responses to peripheral stimulation . This process occurs at any time but is usually seen between one and six weeks after the insult. Muscle over activity declines over time and the following are suggested as possible causes:

- Structural and functional changes due to plastic rearrangement
- Axonal sprouting
- Increased receptor density

**Intrinsic tonic spasticity**

Intrinsic tonic spasticity (increased muscle tone) as that component of spasticity resulting from an exaggeration of the tonic component of the stretch reflex. Briefly, the stretch reflex is a monosynaptic reflex pathway that originates in the muscle spindles embedded parallel to the muscle fibers and travels via a Ia afferent to the spinal cord, where it synapses either first with interneurons or directly with an alpha motor neuron innervating the muscle from which the stimulus originated. The tonic component of the stretch reflex associated with increased muscle tone results from a maintained stretch of the central region of the muscle fibers and the reflex is polysynaptic. Upon a sustained stretch, both type Ia and type II afferents (from secondary spindle endings) synapse with interneurons within the ventral horn of the spinal cord. Synapses of the interneurons with alpha motor neurons facilitate contraction in the muscle being stretched.

It is the hyperexcitability of this tonic stretch reflex that is commonly thought to result in increased muscle tone in response to passive stretch following SCI. This hypertonia is velocity-dependent, with faster stretching velocities being associated with greater amounts of reflex activity. The development of tonic stretch reflex hyperexcitability could be due to a lower threshold, an increased gain of the stretch reflex, or a combination of the two. The resultant increase in muscle tone is thought to be due to a combination of increased denervation hypersensitivity and changed muscle properties. Denervation leads to an initial downregulation of neuronal membrane receptors, followed by an upregulation, with enhanced sensitivity to neurotransmitters. Gradual changes in muscle properties also occur following SCI, such as fibrosis, atrophy of muscle fibers, decrease in the elastic properties, decrease in the number of sarcomeres, accumulation of connective tissue, and alteration of contractile properties toward tonic muscle characteristics, which likely contribute to the increased passive tension.

**Intrinsic phasic spasticity**

Intrinsic phasic spasticity encapsulates symptoms such as tendon hyper-reflexia and clonus, and is due to exaggeration of the phasic component of the stretch reflex. Tendon hyper-reflexia is identified as an exaggerated muscle response to an externally applied tap of deep tendons. Reduced presynaptic Ia inhibition is thought to play an important role in this hyper-reflexia, as the occurrence of reduced

presynaptic inhibition of group Ia fibers appears to correlate with the excitability of tendon reflexes.

Clonus has been defined as 'involuntary rhythmic muscle contraction that can result in distal joint oscillation and most often occurs at the ankle. Clonus is elicited by a sudden rapid stretch of a muscle. The prevailing theory explaining the underlying mechanism responsible for clonus is that of recurrent activation of stretch reflexes. According to this theory, dorsiflexing the ankle causes activation of the Ia muscle spindle afferents and induces a reflex of the triceps surae, resulting in plantar flexion of the ankle. This reflex contraction is brief, essentially phasic, and ceases rapidly. The muscle then relaxes, causing the ankle to be dorsiflexed once again, due either gravity or the stretch being sustained by an examiner. The result is a new stretch reflex, etc. Ultimately, it is the disinhibition of the stretch reflex due to interruption of descending influences with SCI, that is thought to cause exaggeration of the phasic stretch reflex pathway and, hence, clonus.

The second theory is that clonus is the result of activity of a central oscillator or generator within the spinal cord, which rhythmically activates alpha motor neurons in response to peripheral events. Beres-Jones et al outline observations that they feel support such a hypothesis: (1) reports of similar frequencies of clonus among ankle, knee, and wrist muscles, (2) observations that the clonus frequency is not entrained by the input frequency, suggesting that clonus cannot be solely stretch-mediated, (3) the finding that stimuli other than stretch evoke clonus, and (4) the observation of a refractory period following the clonic EMG burst where tendon tap, H-reflex stimulation, and vibration fail to elicit an efferent response. Therefore, whereas reduced presynaptic inhibition of group Ia fibers appears to be among the contributing factors to tendon hyper-reflexia, the underlying mechanism of clonus has not been clearly elucidated.

**Extrinsic spasticity**

In addition to the various intrinsic factors that contribute to symptoms of spasticity, involuntary muscle spasms can also occur in response to a perceived noxious stimulus originating extrinsic to the muscle: extrinsic spasticity. Flexion spasms are the most common form of extrinsic spasticity, triggered by afferent input from skin, muscle, subcutaneous tissues, and joints (collectively referred to as 'flexor reflex afferents'). These flexor reflex afferents mediate the polysynaptic reflexes involved in the flexion withdrawal reflex. SCI can interrupt the inhibition of these reflexes by supraspinal pathways, making them hyperexcitable. In other words, whereas flexor withdrawal reflexes occur normally in individuals without SCI, upon disruption of normal descending influences, the threshold for the flexor withdrawal reflex may become lowered, the gain of the system may become raised, or both may occur together. A recent study has provided evidence to implicate plateau potentials in the spinal interneuronal and motoneuronal circuitry in the hyperexcitability of flexion withdrawal reflexes in individuals with chronic SCI. Intrasegmental polysynaptic connections cause the flexor reflex initiated by a localized stimulus to generate a widespread flexor spasm, which can appear as a coordinated flexion of all joints of the leg.

Spasticity can have a negative impact on quality of life through restricting activities of daily living (ADL), inhibiting effective walking and self-care, causing pain and fatigue, disturbing sleep, compromising safety, contributing to the development of contractures, pressure ulcers, infections, negative self-image, complicating the role of the caretaker, and impeding rehabilitation efforts.

But it has been suggested it also has beneficial effect as symptoms of spasticity may increase stability in sitting and standing, facilitate the performance of some ADL and transfers, increase muscle bulk and strength of spastic muscles (thereby helping prevent osteopenia), and increase venous return (possibly diminishing the incidence of deep vein thrombosis). This potential for a beneficial effect of spasticity on QOL has a large impact upon decisions regarding its management.

**Aetiology**

Increased tone (Spasticity / Rigidity) typically occurs in patients following:

1. Stroke,
2. Brain injury (trauma and other causes, e.g. Anoxia, post-neurosurgery)
3. Spinal cord injury,
4. Multiple sclerosis and
5. Cerebral palsy
6. Syringomyelia
7. Myelomalacia
8. Transverse Myelitis
9. Parkinson's disease
10. Disorders of brain and spinal cord

**Measurement of Spasticity:**

Spasticity depends on several factors like presence of noxious stimuli, the patient's physical and mental status and the position of the body. Therefore, it is difficult to measure spasticity because of its multifactorial nature. Different methods are available for measurement as measurement is essential to assess the response to treatment but none of them is precise and reliable enough to quantify the severity of spasticity clinically.

Ashworth Scale

This scale is based on the assessment of resistance to stretch when a limb is passively moved. It was originally validated for patients with multiple sclerosis and was validated by Ashworth (1964. Its reliability is questioned by the subjectivity required by the observer to carry out the test and by the fact that it measures multiple aspects of limb stretch. However, it is in general use and has good inter-and intra-rater reliability. The original Ashworth scale is only validated for measuring spasticity in the lower limb. In addition, it does not distinguish between increased neurogenic muscle tone and mechanical limb stiffness. The major modification (Modified Ashworth Scale) was proposed to differentiate between mild and moderate spasticity, as discrepancies appeared in clinical judgement at the lower end of the original scale. Bohannon validated the scale in elbow flexion in post-stroke patients and attempts have been made to widen the validity. A grade 1+ was added and the top of the scale was reduced from 5 to 4.

**Tardieu Scale**

The angle at the point of resistance is noted by stretching a limb passively. This is performed during as slow a movement as possible (V1), under gravitational pull (V2) and at a fast rate (V3). The examiner will feel a catch in a muscle under the influence of an overactive stretch reflex. Five levels have been described at the point of this catch to capture the quality of the muscular reaction. In essence the scale assesses dynamic and static muscle length as well as joint range of motion. The inter and intra-rater reliability is generally good.

**Stretch Velocity**

1. V1: Slow as possible
2. V2:Speed of limb falling under gravity
3. V3: Fast as possible

**Y Angle (Dynamic Range of Motion)**

1. R1 Fast Velocity: Movement through full range of motion.
2. R2 Slow Velocity: Passive joint range of motion or muscle length.

**Quality of Muscle Reaction**

**Course of Passive Movement**

0 No resistance

1. Slight resistance
2. Clear catch at precise angle, interrupting the passive movement, followed by release
3. Fatiguable clonus at precise angle, interrupting the passive movement, followed by release
4. Unfatiguable clonus at precise angle, (less than 10 sec when maintaining the pressure) occurring at a precise angle, followed by release.
5. Rigid limb and joint

**Wartenberg Pendulum Test**

In this, the leg moves under gravity and the observer measures the pendular activity of a spastic limb as it relaxes. It is best carried out on the lower limb, for it is not so reliable for other limb segments.

Other methods for evaluating or assessing spasticity include:

1. Muscle grading,
2. Deep tendon reflexes
3. Range of Motion measuring,
4. Bilateral adductor tone score,
5. Visual analogue scale,
6. Spasm frequency score.
7. Torque devices
8. Electrophysiological studies (including dynamic multichannel EMG, tonic vibratory reflexes and electrical tests related to the H reflex and F wave).

**Most of these methods are:**

1. Time consuming,
2. Expensive,
3. Require specialized equipment and
4. Mainly used in research.

**Treating Spasticity:**

Spasticity can be disabling in itself and, if left untreated, may lead to consequences, such as:

- muscle shortening,
- contractures (leading to abnormal body segment loading and sensory change),
- limb deformity and altered body mechanics,
- altered body image,
- the need for special wheelchairs and seating and pressure-relieving equipment,
- loading on pressure points,
- pressure sores,
- difficulty in the management of pressure sores,
- pain from muscle spasms,
- degenerative joint disease,
- loss of function, and
- mood problems and inability to participate in rehabilitation.

| Score | Ashworth (Ashworth 1964) | Modified Ashworth (Bohannon and Smith 1987) |
|---|---|---|
| 0 | No increase in tone | No increase in tone |
| 1 | Slight increase in tone giving a catch when the limb is moved in flexion / extension | Slight increase in tone giving a catch, release and minimal resistance at the end of range of motion (ROM) when the limb is moved in flexion / extension |
| 1+ | | Slight increase in tone giving a catch, release and minimal resistance throughout the remainder (less than half) of ROM |
| 2 | More marked increase in tone, but the limb is easily moved through its full ROM | More marked increased in tone through most of the ROM, but limb is easily moved |
| 3 | Considerable increase in tone - passive movement difficult and ROM decreased | Considerable increase in tone - passive movement difficult |
| 4 | Limb rigid in flexion and extension | Limb rigid in flexion and extension |

Complications will prevent patients from achieving their optimal functioning and deconditioning from ill-health and pain will have a negative effect and patients and their caretakers may find reduced quality of life. Complications that may result due to spasticity are interference with function, nursing care and hygiene, pain, deformity and disfigurement, contractures, joint subluxation and dislocation, peripheral neuropathy and pressure ulcers. Although associated with complications, spasticity is beneficial to some patients. It may help to transfer, stand and ambulate, maintain muscle bulk, prevent deep vein thrombosis and osteoporosis.

**Indications of Antispastic Treatment**

Non-ambulatory patients who have moderate to severe weakness, hyperflexia, clonus and painful flexor spasms which interferes with their ADLS usually require treatment of spasticity. Patients may fulfill more than one indication, e.g. pain relief and care management.

1. To improve the functional improvement in terms of mobility by enhancing the speed and endurance of person in gait or wheelchair propulsion thereby improving transfers. It also improves hand functions in dexterity and in reach outs. It eases the person while performing sexual acts.
2. It helps in relieving pain and muscle spasms thereby keeping the legs in anticontracture postures enabling the person to wear the splints or orthosis.
3. It enhances the body image thereby improving posture.
4. It decreases the burden of caretakers in terms of dressing, positioning the patients for feeding and in personal hygiene.
5. It helps in carryout the rehabilitative therapies thereby delaying or preventing

surgery.

**Principles of Management**

The main goal of therapy is to increase functional capacity, relieve symptoms and decrease caretaker burden. This should be clear to the physician, the patient and the care giver. The consequence of reduction of spasticity should be assessed. If spasticity offers stability to a joint, its reduction may decrease the patient's function. But, if there is minimal weakness with significant spasticity, treatment will result in considerable improvement in the patient's function.

Spasticity requires treatment when it is causing harm, and this is the sole indication. Some patients early on after their stroke or brain injury are helped by their spasticity. For example, patients may start to support their weight by using their spastic lower limb when the degree of weakness in the leg would not allow it. Physical management (good nursing care, physiotherapy, occupational therapy) through postural management, exercise, stretching and strengthening of limbs, splinting and pain relief is the basis of spasticity management.

The aim of treatment is to reduce abnormal sensory inputs, in order to decrease excessive a-motor neuron activity. All pharmacological interventions are adjunctive to a programme of physical intervention. Stretching plays an important part in physical management but needs to be applied for several hours per day. Limb casting has been developed in this field to provide a prolonged stretch. Some studies have suggested that task-specific training might be more effective.

**Patient Assessment**

Spasticity is a movement disorder and patients cannot be adequately assessed unless they are observed during movement and function. Physiotherapists and occupational therapists contribute to the observation and examination process, but some patients with complex movement patterns need assessing in a gait laboratory. The assessment process highlights the differences in patterns of limb posture and movement following an upper motor neuron lesion. Where there is no movement, the assessment process is fairly straightforward, but where there is loss of motor control rather than a spastic dystonia, one has to attempt to identify the different aspects of motor impairment. Patients with longstanding problems also develop compensatory movements, which may or may not require treatment and the clinician has to be clear about the underlying pathophysiological processes.

One can then identify how function is impaired and whether the problem is generalized, focal, or more regional. This will then point to the options for treatment. The indication for pharmacological treatment therefore is when spasticity is causing the patient harm. Some patients early on in their rehabilitation following a stroke or brain injury use their spasticity to walk on, when their weakness would otherwise not allow it. Clearly, treating the spasticity here would not be helpful and physical measures to utilize the developing movement patterns would be the treatment of choice, but where the spasticity gives rise to problems for either the patient or the caretaker, then treatment is required.

It is sometimes quite difficult to distinguish between severe spasticity and contracture formation, but it is important to do so. The clinicians and the patient / caretaker can then know what anti- spastic treatment can or cannot achieve and realistic expectations can then be identified. Severe, inadequately treated spasticity will go on to develop a limb contracture through shortening the muscle and tendons. A contracture may be fixed and will require serial splinting or surgery to

correct it, but before it becomes fixed, the spasticity contributes to a dynamic contracture and treating the underlying spasticity may allow easier treatment of the contracture. One way to do that is examination under sedation. It is advisable to use a general anaesthetic for children. This relaxes spastic muscles and allows the range of passive joint movement to be assessed. One particular use is in assessing patients, who externally rotate their leg during walking. The adductor muscles can compensate for weak hip flexors and the patient rotates the leg accordingly. Blocking the obturator nerve reduces the function of the adductors and it is then possible to see the degree of hip flexor weakness, so that a programme of muscle strengthening can be started rather than of BTX injections to weaken the adductors.

**Medical Management of spasticity:**

Spasticity is a symptom and not a disease. Management of spasticity begins with the assessment of the underlying disorder.

**Medical Management:**

**Treatment options:**

A. Oral Drug Therapy: Number of drugs are available for the treatment of spasticity. These drugs reduce muscle tone and painful spasms, however their use is often limited by their side effects. The efficacy of oral antispastic drugs is small and evaluation of the effect on patient's quality of life is lacking from the available studies. 23

1. Baclofen: Baclofen is a GABA (?-aminobutyric acid) agonist and acts by inhibiting both monosynaptic and polysynaptic spinal cord reflexes.24The oral dose of baclofen used to treat spasticity ranges from 30-100 mg / d in two to three divided doses. Oral baclofen may cause considerable side effects such as sedation, respiration problems and muscular weakness in higher doses. 25Baclofen must be tapered slowly to prevent withdrawal effects like increase in spasticity, fever, altered mental status, seizures, malignant hyperthermia.

2. Benzodiazepines: (Diazepam and clonazepam) Benzodiazepines act by increasing the affinity of GABA receptors for endogenous GABA.26Diazepam can be started at 5 mg at bedtime, and if daytime therapy is indicated, the dosage can be increased slowly to 60 mg / d in divided doses. Clonazepam can be started at 0.5 mg at night and slowly increased to a maximum of 20 mg / d in 3 divided doses. The side effect includes sedation, confusion, habituation and tachyphylaxis.

3. Dantrolene: Dantrolene is more useful for spasticity of cerebral origin. Dantrolene interferes directly with the excitation-coupling reaction. It acts at the level of the muscle fiber, affecting the release of calcium from the sarcoplasmic reticulum of skeletal muscle and thus reducing muscle contraction.]27 Dantrolene is given in a dose of 0.5-3.0 mg / kg / d. Dantrolene may cause side effects like muscle weakness and hepatotoxicity.

4. Tizanidine: Tizanidine decreases the excitability of and motor neurons in the spinal cord by reducing the release of excitatory neurotransmitters in the spinal cord and decreasing the action of these excitatory neurotransmitters at their receptors. 28 Tizanidine also acts by inhibiting the release of substance P from small sensory afferent nerve fibers as well as slowing the firing of the locus ceruleus. 29 Tizanidine should be started at a low dose, 2-4 mg, preferably at bedtime. The average maintenance dosage of tizanidine is 18-24 mg / d. The

maximum recommended dosage is 36 mg / d. The side effects of tizanidine are sedation, dizziness, dry mouth, and hypotension. Tizanidine does not cause any muscle weakness. 30 Therefore, it might be preferable over other antispasticity medications that do cause weakness, such as baclofen and dantrolene, especially in patients whose strength is already compromised by neurological disease.



*Fig.1 : Intrathecal Baclofen pump*

5. Intrathecal Baclofen pump: The poor penetrataion of blood-brain barrier and significant side effects of oral baclofan can be minimized by intrathecal administration (directly into CSF) via a programmable pump. Considerably lower doses are required in intrathecal injection and it is without the development of tolerance. It should be considered in patients unresponsive to oral pharmacotherapy and a severity of 3 on the Ashworth scale for at least 12 months. A test dose should first be given intrathecally before the pump is implanted. The dose range is 12- 2000 mcg / d and should be fine tuned according to the severity of symptoms and response to therapy. The complications of intrathecal baclofen pump implantation are relatively few and usually are limited to mechanical failures of the pump or the catheter. Adverse drug effects are usually temporary and can be managed by reducing the rate of infusion.



*Fig 2. Intrathecal Baclofen pump*

Neurolysis with Neurotoxins: Botulinum toxin is a neurotoxin produced by the clostridium botulinum bacterium. It is the same toxin that also causes food poisoning which causes stomach and intestinal problems. Botox is a brand name and not the name of the drug. Botulinum toxin acts by causing reversible block of neuromuscular transmission by inhibiting acetylcholine release. Botulinum toxin is an "Exotoxin" i.e., it is secreted / produced outside the cell and on injection in the muscle causes blockage of Acetylcholine release (ACH) at the neuromuscular junction. This Ach causes the muscles to contract. By blocking the release of the Ach, Ach receptors cannot generate an impulse and thus the spastic muscles undergoes partial paralysis. There are seven different serotypes of botulinum toxins (A,B,C,D,E,F,G). Botulinum toxin is injected intramuscularly into the spastic muscles using a very fine needle. Injections should be targeted to spastic muscles responsible to functional loss using EMG guidance. The effect of botulinum toxin starts within two weeks after injection. The clinical effect appears 4-7 days after injection, reaches a maximum after about 2 months thereafter

the effect tapers off. Ideally it is best for classical clasp-knife type spasticity due to periventricular leukomalacia and cortical lesions and less effective for dystonia, lead – pipe type rigidity and athetosis. Patients who have severe spasticity may require a cast for 2 weeks. Delayed casting is now advocated as muscle movement is supposed to enhance the efficacy of the drug. Exercise both active and assisted is required to get the maximum benefit after injection. The effect of botulinum toxin is not permanent and lasts for 3-4 months. Therefore role of physiotherapy is more important after the injection as one can work on the muscles to improve voluntary control and strength of the muscles which in turn will prevent reoccurrence of spasticity.

Botulinum injections are very safe provided certain dosage guidelines are adhered to. The technique and site of injection require knowledge and training. In experienced individuals the side effects are minimal which includes mild fever or muscle pain. In children who have respiratory compromise the injection should be used with caution especially in children with quadriplegia.

Based on the amount of spasticity the common muscles injected are calf for equines deformity or toe walking, hamstring for knee flexor tightness, hip adductors for crossing of leg or scissoring, rectus or quadriceps for knee stiffness in extension and also the strap muscles around the foot to control foot inversion or eversion.

In the upper limb, the injection is used in the biceps tendon, long flexors of the fingers, wrist flexors, thumb adductors to correct various deformities.

Splints or orthosis are very important after the injection as this maintain the correct attitude of the joint for day-day function.

**Side effects of Botox:**

1. Local Weakness
2. Systemic effects: bowel and bladder control, generalized weakness, dysphasia, dysphasia, dysphagia
3. Effect on local and distant musculature

**Advantages of Botox?**

1. Relatively painless
2. Ease of administration (even an orthopedic surgeon can do it)
3. Diffuses readily
4. Transient effects
5. No known cumulative effects

**What are the disadvantages of Botox?**

1. Variable dosing
2. Transient effect
3. Expensive
4. Possibility of antibody formation
5. Long term effect on muscle not known
6. Chemical neurolysis (Phenol or alcohol Injections): A peripheral nerve innervating the spastic muscles is injected with phenol or alcohol solutions, which destroys myelin. It is used in patients with focal spasticity. The effectiveness of the injection diminishes over time and repeated injections are needed.

**Alcohol Blocks (Ethyl alcohol (>10%)**

1. Pharmacology: injected at the motor point and destroys tissue and imparts circulatory damage to denervate
2. Administration: intramuscularly or intraneurally
3. Cost: drug itself is inexpensive, may require anesthesia or sedation for children
4. Side-effects: pain at injection site, chronic dysesthesia, pain, vascular complications

5. Advantages : effects last 2-36 hours
6. Disadvantages: may have side effects, painful

**Phenol Blocks (>3%)**

1. Pharmacology: tissue destruction and circulatory damage
2. Administration: intramusculary with stimulation of the motor point.
3. Cost: inexpensive, but may require anesthesia
4. Side-effect: pain at infection, chronic dysesthesia and pain, permanent nerves palsy
5. Advantages: long lasting (2-36 months), inexpensive
6. Disadvantages: difficulty locating motor point, side effect, muscle fibrosis
7. Electrical Stimulation: Electrical Stimulation of muscles, peripheral nerves and spinal cord has been used in the management of spasticity. Surface electrical stimulation of spastic muscles cause reduction of spasticity by stimulating cutaneous afferents and suppressing motoneuronal excitability. 40 Electrical stimulation of peripheral nerves reduces spasticity by inducing complete and reversible conduction block.41 Electrical stimulation of the dorsal columns of the spinal cord through epidurally placed electrodes may reduce spasticity. 42 Direct stimulation of spinal cord suppressess excitability of spinal motoneurons and cause reduction of spasticty.
8. Transcranial magnetic stimulation: Repetitive high-frequency (5 Hz) and low-frequency (1 Hz) transcranial magnetic stimulation may improve spasticity.

**Surgical Management:**

Surgery can play a very important role in the treatment of chronic spasticity or to allow more normal bone and muscle growth. Surgical treatment of spasticity involves neurosurgery and orthopaedic surgery. Neurosurgical treatment for spasticity is reserved for severe cases in which medical management has been ineffective or has lost its effectiveness. Neurosurgical procedures for spasticity include Selective Posterior (Dorsal) Rhizotomy (SDR), microsurgical DREZotomy, Peripheral Neurotomy, Longitudinal Myelotomy and Neurectomy.

Posterior (Dorsal) Rhizotomy (SDR) involves cutting of the dorsal nerve roots that lie just outside the vertebral column and transmit nerve impulses to and from the spinal cord. SDR is primarily indicated in conditions exhibiting severe spasticity that interferes with mobility or positioning. 43 Microsurgical DREZotomy (Dorsal Root Entry Zone-otomy) is a type of selective rhizotomy. It involves cutting the nerve fibers at the entry zone and suppresses afferent discharges to the spinal cord. It is more effective in the treatment of severe spasticity limited to the upper or lower limbs. Peripheral neurotomy involves cutting of peripheral nerves at the point at which they enter the muscle. It is indicated in the treatment of spastic neck, elbow, hand, hip, and foot. 45Longitudinal Myelotomy involves longitudinal division of the spinal cord to sever crossing sensory fibers and produce localized analgesia. It used to be performed earlier for severe and painful spasticity. 46 Neurectomy involves the cutting of the nerve branches as they enter the targeted muscle. It is indicated in patients with focal spasticity refractory to botulinum toxin.

Orthopaedic surgical procedures for the management of spasticity include muscle / tendon surgery and bone surgery. Muscle / tendon surgery involves lengthening or release of muscles and tendons. In

contracture release surgery, contractured muscle is cut and the joint is then positioned at a more normal angle allowing regrowth of the tendon to this new length. Tendon transfer involves moving the attachment point of a spastic muscle to allow improved active function or retention of only passive function. Bone surgery can be osteotomy or arthrodesis. Osteotomy involves removal of a small wedge of bone to allow it to be repositioned or reshaped. Arthrodesis is fusion of bones that normally move independently and thereby limiting the spastic muscle to pull the joint into an abnormal position.

**Physical Therapy Management:**

Spasticity sets in gradually after the initial insult to the central nervous system. It becomes noticeable in the first few months, but the timing varies depending on the extent of underlying neurologic insult. Once recovery from the neurologic deficit stabilizes, the spasticity also tends to stabilize. Spasticity is not always detrimental as a weak flaccid limb can interfere with such daily activities as transferring, dressing, grooming and perineal care, spasticity provides posture and tone to a limb that can assist with weight bearing even if the patient cannot walk. However, excessive tone may interfere with these activities. Thus, it is only when spasticity interferes with function or puts the individual at risk of hurting himself or herself, it needs to be treated.

Physical interventions helps in minimizing the viscoelastic properties of the connective tissue; altering the neural patterns of spasticity or spasms and maintaining levels of function for the individual.

**The goal of physiotherapy in the management of spasticity should include** :

1. Reduction of excessive tone.
2. Facilitate normal movement patterns.
3. To give a sense of normal position and normal movement.

**1) Body Positioning:**

It is advisable to inhibit the unwanted activity of the released reflex mechanisms by positioning the head and neck of the patient as they have tendency to elicit strong postural reflex mechanisms. Proper head and neck positions can inhibit these reflexes thereby

preventing the rest of the body from going into the reflex pattern. Side lying with pillow support is most preferred position as it avoids stimulation of the tonic labyrinthine reflex and the asymmetrical tonic neck reflexes. Rhythmical trunk rotations can be easily given in both passive and active assisted exercises which helps in reduction of spasticity.

**Equipment that Improves Positioning**

Controlling the position of a child's pelvis, knees, and ankles can help decrease arching, increased head control and shoulder stability, and improve functional hand use. Children may need different types of equipment at different times during their growth and development. Again, the goal is to increase control of movement and manipulation of objects in the environment. Some examples of equipment that can help improve positioning are:

1. Adapted strollers
2. Wheelchairs
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**1) Rotatory Movements:**

Trunk rotation produces lower limb to extend, abduct and externally rotate which is

the position opposite to reflex pattern. Limb rotations are also very effective in normalizing the muscle tone of the patient.

**2) Pressure over undersurface of Foot:**

When the pressure is applied to the ball of the foot it stimulates an extensor reflex which is a pathological pattern of extension, adduction, and medial rotation of hip with plantar flexion of the foot, which is undesirable event in case of spasticity. When the pressure is applied under the heel of the foot, purposeful contraction of muscle occurs giving a normal supporting pattern. One should facilitate normal movements Patterns thereby avoiding triggering factors. Due to reflex release, some motoneurone pools are already in an excitatory state and any volitional effort will act as a trigger to those motoneurone pools resulting in muscle contraction in the spastic pattern. In such patients strong volitional effort should be discouraged as this facilitates the movement in the spastic pattern.

Other factors such as quick movements, abruptly performed, noisy surroundings, anxiety, excitement, over exertion should also be avoided as it may increase spasticity.

Neurolysis with Neurotoxins: Botulinum toxin is a neurotoxin produced by the clostridium botulinum bacterium. It is the same toxin that also causes food poisoning which causes stomach and intestinal problems. Botox is a brand name and not the name of the drug. Botulinum toxin acts by causing reversible block of neuromuscular transmission by inhibiting acetylcholine release. Botulinum toxin is an "Exotoxin" i.e., it is secreted / produced outside the cell and on injection in the muscle causes blockage of Acetylcholine release (ACH) at the neuromuscular junction. This Ach causes the muscles to contract. By blocking the release of the Ach, Ach receptors cannot generate an impulse and thus the spastic muscles undergoes partial paralysis. There are seven different serotypes of botulinum toxins (A,B,C,D,E,F,G). Botulinum toxin is injected intramuscularly into the spastic muscles using a very fine needle. Injections should be targeted to spastic muscles responsible to functional loss using EMG guidance. The effect of botulinum toxin starts within two weeks after injection. The clinical effect appears 4-7 days after injection, reaches a maximum after about 2 months thereafter the effect tapers off. Ideally it is best for classical clasp-knife type spasticity due to periventricular leukomalacia and cortical lesions and less effective for dystonia, lead – pipe type rigidity and athetosis. Patients who have severe spasticity may require a cast for 2 weeks. Delayed casting is now advocated as muscle movement is supposed to enhance the efficacy of the drug. Exercise both active and assisted is required to get the maximum benefit after injection. The effect of botulinum toxin is not permanent and lasts for 3-4 months. Therefore role of physiotherapy is more important after the injection as one can work on the muscles to improve voluntary control and strength of the muscles which in turn will prevent reoccurrence of spasticity.

Botulinum injections are very safe provided certain dosage guidelines are adhered to. The technique and site of injection require knowledge and training. In experienced individuals the side effects are minimal which includes mild fever or muscle pain. In children who have respiratory compromise the injection should be used with caution especially in children with quadriplegia.

Based on the amount of spasticity the common muscles injected are calf for equines deformity or toe walking, hamstring for knee flexor tightness, hip adductors for crossing of leg or scissoring, rectus or quadriceps for knee stiffness in extension and also the strap muscles around the foot to control foot inversion or eversion.

In the upper limb, the injection is used in the biceps tendon, long flexors of the fingers, wrist flexors, thumb adductors to correct various deformities.

Splints or orthosis are very important after the injection as this maintain the correct attitude of the joint for day-day function.

**Side effects of Botox:**

1. Local Weakness
2. Systemic effects: bowel and bladder control, generalized weakness, dysphasia, dysphasia, dysphagia
3. Effect on local and distant musculature

**Advantages of Botox?**

1. Relatively painless
2. Ease of administration (even an orthopedic surgeon can do it)
3. Diffuses readily
4. Transient effects
5. No known cumulative effects

**What are the disadvantages of Botox?**

1. Variable dosing
2. Transient effect
3. Expensive
4. Possibility of antibody formation
5. Long term effect on muscle not known
6. Chemical neurolysis (Phenol or alcohol Injections): A peripheral nerve innervating the spastic muscles is injected with phenol or alcohol solutions, which destroys myelin. It is used in patients with focal spasticity. The effectiveness of the injection diminishes over time and repeated injections are needed.

**Alcohol Blocks (Ethyl alcohol (>10%)**

1. Pharmacology: injected at the motor point and destroys tissue and imparts circulatory damage to denervate
2. Administration: intramuscularly or intraneurally
3. Cost: drug itself is inexpensive, may require anesthesia or sedation for children
4. Side-effects: pain at injection site, chronic dysesthesia, pain, vascular complications
5. Advantages : effects last 2-36 hours
6. Disadvantages: may have side effects, painful

**Phenol Blocks (>3%)**

1. Pharmacology: tissue destruction and circulatory damage
2. Administration: intramusculary with stimulation of the motor point.
3. Cost: inexpensive, but may require anesthesia
4. Side-effect: pain at infection, chronic dysesthesia and pain, permanent nerves palsy
5. Advantages: long lasting (2-36 months), inexpensive
6. Disadvantages: difficulty locating motor point, side effect, muscle fibrosis
7. Electrical Stimulation: Electrical Stimulation of muscles, peripheral nerves and spinal cord has been used in the management of spasticity. Surface electrical stimulation of spastic muscles cause reduction of spasticity by stimulating cutaneous afferents and suppressing motoneuronal excitability. 40 Electrical stimulation of peripheral nerves reduces spasticity by inducing complete and reversible conduction block.41 Electrical stimulation of the dorsal columns of the spinal cord through epidurally placed electrodes

may reduce spasticity. 42 Direct stimulation of spinal cord suppressess excitability of spinal motoneurons and cause reduction of spasticty.

8. Transcranial magnetic stimulation: Repetitive high-frequency (5 Hz) and low-frequency (1 Hz) transcranial magnetic stimulation may improve spasticity.

**Surgical Management:**

Surgery can play a very important role in the treatment of chronic spasticity or to allow more normal bone and muscle growth. Surgical treatment of spasticity involves neurosurgery and orthopaedic surgery. Neurosurgical treatment for spasticity is reserved for severe cases in which medical management has been ineffective or has lost its effectiveness. Neurosurgical procedures for spasticity include Selective Posterior (Dorsal) Rhizotomy (SDR), microsurgical DREZotomy, Peripheral Neurotomy, Longitudinal Myelotomy and Neurectomy.

Posterior (Dorsal) Rhizotomy (SDR) involves cutting of the dorsal nerve roots that lie just outside the vertebral column and transmit nerve impulses to and from the spinal cord. SDR is primarily indicated in conditions exhibiting severe spasticity that interferes with mobility or positioning. 43 Microsurgical DREZotomy (Dorsal Root Entry Zone-otomy) is a type of selective rhizotomy. It involves cutting the nerve fibers at the entry zone and suppresses afferent discharges to the spinal cord. It is more effective in the treatment of severe spasticity limited to the upper or lower limbs. Peripheral neurotomy involves cutting of peripheral nerves at the point at which they enter the muscle. It is indicated in the treatment of spastic neck, elbow, hand, hip, and foot. 45Longitudinal Myelotomy involves longitudinal division of the spinal cord to sever crossing sensory fibers and produce localized analgesia. It used to be performed earlier for severe and painful

spasticity. 46 Neurectomy involves the cutting of the nerve branches as they enter the targeted muscle. It is indicated in patients with focal spasticity refractory to botulinum toxin.

Orthopaedic surgical procedures for the management of spasticity include muscle / tendon surgery and bone surgery. Muscle / tendon surgery involves lengthening or release of muscles and tendons. In contracture release surgery, contractured muscle is cut and the joint is then positioned at a more normal angle allowing regrowth of the tendon to this new length. Tendon transfer involves moving the attachment point of a spastic muscle to allow improved active function or retention of only passive function. Bone surgery can be osteotomy or arthrodesis. Osteotomy involves removal of a small wedge of bone to allow it to be repositioned or reshaped. Arthrodesis is fusion of bones that normally move independently and thereby limiting the spastic muscle to pull the joint into an abnormal position.

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3)Slow Sustained Stretching:

Stretching forms, the basis of spasticity treatment. Stretching helps to maintain the full range of motion of a joint, and helps prevent contracture, or permanent muscle shortening as it activates muscle spindles (Ia & II endings), golgi tendon organs (Ib

endings) which are sensitive to length changes, inhibits or dampens muscle contraction and tone due largely to peripheral reflex effects. It is effective in extensor muscles than flexors due to the added effects of II inhibition. Stretching when done forcefully against severe spasticity, it increases hyperexcitable stretch reflex more strongly damaging periosteum of bone when excessive tension is applied to the tendons of the stretched muscles.

**Techniques used to maintain slow sustained stretching are as:**

1. Manual contacts
2. Inhibitory casting or splinting
3. Reflex-inhibiting patterns
4. Mechanical low-load weights

**4) Prolonged Cold Application:**

Application of cold packs to spastic muscles (usually for 10 minutes or longer) may improve muscle tone. While the effect is not long lasting, it helps to improve function for a short period of time, or to ease pain by activating thermoreceptors thereby decreasing neural, muscle spindle firing leading to inhibition of muscle tone.

Techniques used

1. Immersion in cold water; ice chips
2. Ice towel wraps
3. Ice packs
4. Ice massage
5. Ice application with exercises

**5) Neutral Warmth:**

Retention of body heat stimulates thermoreceptors, autonomic nervous system mainly parasympathetics, which produces generalized inhibition of tone, calming effect, relaxation and decreases pain. It should be applied for about 10 to 20 minutes. Overheating should be avoided as it might increase arousal or tone.

**Techniques used are as:**

1. Wrapping body or body parts: ace wraps, towel wraps
2. Application of snug fitting clothing (gloves, socks, tights) or air splints

**6) Relaxed Passive Movements:**

Rhythmical, passive movements performed slowly throughout the range of motion in normal pattern may help in reducing spasticity.

**7) Deep Rhythmical Massage (Tendon Rolling):**

Deep rhythmical massage with pressure over the muscle insertions can be given to reduce spasticity.

**8) Inhibitory Pressure (Weight-Bearing):**

Prolonged pressure to long tendons inhibits the hypertonicity of a muscle as it activates muscle receptors (muscle spindles, golgi tendon organ) and tactile receptors. It can be applied manually or by body weight. Weight bearing postures provides inhibitory pressure, such as quadruped or kneeling postures can be used to promote inhibition of quadriceps and long finger flexors and sitting, with hands open, elbow extended, and upper extremity supporting body weight can be used to promote inhibition of long finger flexors.

**9) Biofeedback:**

Biofeedback is the use of an electrical monitor that creates a signal usually a sound as a spastic muscle relaxes. Thus, the person with spasticity may be able to train himself to reduce muscle tone consciously.

**10) Functional Electrical Stimulation:**

Electrical stimulation may be used to stimulate a weak muscle to oppose the activity of a stronger, spastic one thereby improving motor activity in agonistic

muscles and reducing the tone in antagonistic muscles. Once stimulation has been stopped, the effect last for less than 1 hour probably because of neurotransmitter modulation within reflex arc. It improves standing, walking, and exercise training as well as decreases upper extremity contractures.

**12) Tone Reducing Orthosis:**

Orthotics are designed to help provide support to weak muscles and minimize the risk of joint deformity. There are a variety of orthotics made from a number of different materials. The goal is to use an orthotic which can give support depending on a child's pattern of movement, avoid skin breakdown, and be comfortable. If areas of the skin become red, this indicates that the orthotic may not be fitting appropriately, especially if the redness lasts more than half an hour after removing the orthotic.

1. Ankle foot orthoses (AFO's): These are plastic AFO's in which foot plate and broad upright are designed to modify reflex hypertonicity by applying constant pressure to the plantarflexors and invertors. Foot plate may be modified which maintains the toes in an extended or hyperextended position, thus assisting individual to walk with better foot and knee control thereby preventing the foot to go into equinovarus position.
2. Leg braces or casts
3. Hand splints
4. Soft body jackets

**13) Serial Casts to Gain Range of Motion**

Serial casts can either be used for the arms or the legs. The goal is to maintain or increase range of motion of a muscle, tendon, or joint. The casts provide a sustained stretch across the joint. Prolonged stretch can help muscles relax. The cast may also help "soften" tendons. Serial casts are normally changed one time per week. The number of total weeks of casting varies depending on the need of each child. The goal is to slowly gain more joint range without causing significant discomfort to the child.

**14) Inhibitive Casts to Improve Function**

Inhibitive casts are used to increase function more than to improve range of motion. Sometimes, inhibitive casts will be used to "give more information" (proprioception) to the ankle joint or the foot in order to prevent a child from using "reflex patterns" of movement. They are often utilized when a child with muscle imbalance is learning to walk. They can give better proprioceptive input to the foot when compared to the AFO since they are heavier and can have special features built in to them. For example, inhibitive foot plates can be built into the base of each cast. The inhibitive foot plates apply pressure to different areas of the foot to give better proprioceptive input to the joint which inhibits reflex patterns of movement. Often times, we will use inhibitive casts prior to prescribing orthotics, depending on a child's function.

**15) Slow Maintained Vestibular Stimulation: Low-intensity vestibular stimulation such as slow rocking produces generalized inhibition**

of tone as it facilitates primarily otolith organs (tonic receptors); less effects on semicircular canals (phasic receptors). Slow rolling movements assisted rocking in a weight- bearing position or rocking with equipments like rocking chair, Swiss ball, equilibrium board, Hammock can help in reducing the tone.

**16) Proprioceptive Neuromuscular Facilitation Techniques :**

Proprioceptive neuromuscular facilitation is a neuromuscular treatment that uses repetitive stretches where one muscle is

contracted while another muscle is relaxed at the same time. There are several techniques performed in proprioceptive neuromuscular facilitation which increases range of motion, improves range of stretch, increase strength and develop healthy muscle tissue.

**Techniques used in reducing spasticity are as:**

1. Rhythmic Initiation - Voluntary relaxation followed by passive movements through increments in range, followed by active movements progressing to resisted movements using tracking resistance to isotonic contractions.
2. Rhythmic Rotation - Voluntary relaxation combined with slow, passive, rhythmic rotation of the body or body part around a longitudinal axis, followed by passive movement into the antagonist range.
3. Contract Relax Active Contraction - Isotonic movement in rotation is performed followed by isometric hold of the range limiting muscles in the antagonist pattern against slowly increasing resistance followed by voluntary relaxation and active movement into the new range of the agonist pattern.

**17) Manipulating Key Points:**

Spasticity can be reduced by manipulating the key points in the upper limb and lower limb like the movements with thumb in abduction helps in reducing the spasticity in upper limb and in lower limb in sitting position, placing one hand over the lower back and other near the xiphoid process and moving the patient in the figure of 8 pattern forwards and backwards thus manipulating the pelvis helps in reducing spasticity.

**18) Examples of different therapy models include:**

1. Neurodevelopmental therapy (NDT),
2. Conductive education.
3. Manual therapy (myofascial release and craniosacral therapy).
4. Hippotherapy, or horseback riding, can help a child increase balance and improve trunk control.
5. Aquatic therapy can help promote more typical patterns of movement that are often affected by gravity.

**19) Alternative Treatment**

Alternative and complementary therapies include approaches that are considered to be outside the mainstream of traditional health care. Techniques that reduce stress, such as yoga, Tai Chi, meditation, deep breathing exercises, guided imagery, and relaxation training, may be helpful to induce relaxation and manage spasticity. Acupuncture and biofeedback training also may help induce relaxation.

**Occupational Therapy management in Spasticity**

Intervention for hypertonicity and spasticity is necessary when it interferes with ADL's, gait, sleep or wheelchair positioning or when it causes severe pain and limits hygiene (for eg. the client is unable to wash hand or axilla) or when spasticity leads to contractures or pressure sores.

**Spasticity, if not managed can lead to secondary problems such as:**

- Deformity of the limbs, specially the distal upper limb (elbow to digits)
- Maceration of the palm tissue. Possible masking of underlying motor control.
- Pain syndromes resulting from loss of normal kinematics
- Impaired ability to manage BADL tasks ( especially upper extremity dressing and bathing of the affected hand and axilla

when flexor positioning is present in a patient with stroke).

- Loss of reciprocal arm swing during gait activities.

However before treating hypertonicity, the therapist should closely evaluate the function of the tone. Hypertonicity can have beneficial effects such as aiding him in standing and transfers, maintaining muscle bulk and prevent deep vein thrombosis, osteoporosis and edema.

A uniformly acceptable, reliable and practical measure of spasticity continues to elude the therapists. Objective assessment of muscle tone in clients with hypertonia and spasticity is difficult. The postural reflex mechanism, the position of body and head in space, the position of head in relation to body and stereotypical reflexes and associated reactions all influence the degree and distribution of abnormal muscle tone.

**Manual rating scale for hypertonicity and spasticity:**

The Ashworth Scale and the Modified Ashworth Scale are 2 most widely used scales to manually rate spasticity. There is a controversy in literature about the validity and reliability of these scales.

Brashear et al concluded that in upper limb spasticity, the Ashworth Scale had good intra and inter - rater reliability when used by trained medical professionals.

Tardieu Scale: Modified Tardieu Scale and the Tardieu Scale both measure spasticity. Modified Tardieu Scale has an interrater reliability coefficient of 7 and was shown to be more reliable than Modified Ashworth Scale .

Mild-Moderate-Severe Spasticity Scale and Preston's Hypertonicity Scale: Some therapists find it easier to use these scales.

**MILD-MODERATE-SEVERE SPASTICITY SCALE:**

Mild: The stretch reflex (palpable catch) occurs at the muscles end range (i.e. the muscle is in a lengthened position).

Moderate: The stretch reflex (palpable catch) occurs in mid range.

Severe: The stretch reflex (palpable catch) occurs when the muscle is in a shortened range.

**PRESTON'S HYPERTONICITY SCALE:**

0: No abnormal tone detected during slow passive movement.

1. Mild: First tone or resistance is felt when the muscle is in a lengthened position during slow passive movement.

2. Moderate: First tone or resistance is felt in the mid range of the muscle during slow passive movement.

3. Severe: First tone or resistance occurs when the muscle is in the shortened range during slow and passive movement.

**MECHANICAL AND COMPUTER RATING SYSTEMS FOR HYPERTONIA AND SPASTICITY**

Mechanically determined parameters are more reliable than the manual methods. McCrea et al, concluded that using a linear spring damper model to assess the hypertonic elbow was reliable and valid 62. However, it is time consuming and certain muscle groups are difficult to assess with it.

Leonard et al investigated the construct validity of myotonometer, a newly developed computerized, electronic device. The authors concluded that the myotonometer could provide objective data about the tone reducing efficacy of various tone reducing procedures .

In the past, sensorimotor approaches were used to treat patient's with abnormal skeletal muscle activity. These approaches were developed by Rood, Bobath, Knorr and Voss

(PNF) and Brunnstorm and are based on an understanding of CNS dysfunction. Although these interventions are commonly used, their effectiveness is being challenged as occupational therapist move towards models of evidence-based practice64.

For years, occupational therapists have been using weight bearing activities to reduce hyper-tonicity and to remediate paresis in patients with upper motor neuron lesions. At present there is a limited amount of research to support these neurofacilitation approaches. Brouwer and Ambury concluded that cortico-spinal facilitation of motor units occurred during weight bearing. They believed that afferent input from weight bearing increased motor cortical excitability.

Chakerain and Larson studied the effects of upper extremity weight bearing on hand opening and prehension in children with spastic cerebral palsy. Computer calculation of the patient's hand surface area were measured and it was found that there was an increase in surface area after weight bearing and an increase in the maturity of movement components needed for prehension.

McIllroy and Maki demonstrated that if the affected arm is used when weight bearing, postural responses occur throughout the weight bearing extremity and occur during other perturbations of Posture.67Although, not many studies have documented how and why weight bearing works physiologically, it certainly is a requirement for improving performance.

An important occupational therapy objective is to have client manage muscle tone to engage in and complete basic and instrumental activities of daily living. Positioning and movement in patterns opposite to hypertonic or synergistic patterns are important to expand the motor repertoire and develop movement that is as close to normal as possible.

The client should also be taught how to incorporate the affected extremity as much as possible into all ADLs. ADLs, crafts, games, and work activities can be used to teach incorporation of the extremities for a total approach to treatment Casting in inhibitive postures has been shown to be effective in tone reduction. The beneficial effect of casting on hypertonia is well documented in literature. Serial casting is effective in presence of a contracture and its use should cease when desired position is achieved and tone is manageable using a splint.

Multiple commercially available spasticity reducing splints are used to keep hand and wrists in inhibitive postures. The client and family need to be educated in continuing to incorporate the extremities in occupations and to bear weight on extremities as much as possible; to retain the ROM gain achieved during casting.

Physical agent modality such as cold, superficial heat, ultrasound and Neuromuscular Electric Stimulation can be used as a preparation for or in conjunction with purposeful activity and muscle reeducation, provided the therapist has the appropriate training.

The Functional Tone management (FTM) Arm Training Program is based on distal activation model focusing on the key points of early initiation of upper extremity movements that incorporate grasp and release. In order to incorporate the hand into FTM arm training, a dynamic orthosis for the hand called the Saebo Flex is used. This orthosis assists an individual who exhibits hypertonia in the hand to place the hand in an open functional position and also to produce a graded muscle contraction of the finger flexors in order to grasp an object. The FTM program combines high repetition grasp and release with task specific arm training drills to progress the client towards a functional goal. A significant body of

research support the FTM arm training program. Clinically observed improvements with FTM arm training Program include increased AROM at the shoulder, elbow and wrist, improved upper extremity Fugal-Meyers Scores and decreased modified Ashworth Scores.

In conclusion, the impact of spasticity can be far- reaching on an individual's lifestyle and that of family and caretakers. Health care professionals can provide vital care support to individuals and their families and refer them appropriately towards other professionals and organizations for required intervention and support.

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# Chp. 24 Aquatic Therapy

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Aquatic therapy is one of the widely used treatment modalities. Although, it was relatively new in India, the awareness for the same is increasing. Aquatic therapy in simple terms means making use of different physical and chemical properties of water to facilitate functional recovery, independence, prevent complications and slow down the damage to the muscles in individuals with muscular dystrophy. Aquatic Therapy network of India defines aquatic therapy as "The aquatic therapy is the use of water and its chemical and physical properties to perform specifically designed activity given by qualified personnel to aid in the restoration, extension, maintenance and quality of function for persons with acute, transient, or chronic disabilities, syndromes or diseases." In aquatic therapy along with aquatic environment qualified personnel are equally important in bringing about the outcome. Therefore as therapists, it is important for us to understand how aquatic therapy can be beneficial in achieving our therapeutic goals. Aquatic therapy is being practiced by Physiotherapists and Occupational therapists currently, other disciplines of rehabilitation like psychologists, speech therapists and special educators are yet to explore this concept but water offers unlimited opportunities. The aim of this chapter is to offer information about properties of water, physiological effects of immersion and different disciplines of aquatic therapy as well as application of aquatic therapy in Neurological disorders.

**Properties of water**

Water has various chemical as well as physical properties and physical principles that guide movement of object in water. It is important for an aquatic therapist to understand these principles in order to understand their own movement in water and movement of their patients in water as well.

**Chemical properties of water**

These do not directly interfere with the therapy and therapeutic handling of the patient but may have a physiological effect on the patients. Some of important chemical properties are that water is colourless, odourless, a good conductor of heat and a good solvent. Many of the salts can be dissolved in water, this property is important from the point of view of water quality maintenance in the therapeutic pool; as the impurities may get dissolved in water and may change the chemical composition of water. One property that warrants caution is that water is a good conductor of electricity and therefore electrical therapeutic appliances must be kept well out or reach in aquatic therapy set up.

**Physical properties of water**

These are very important from the therapeutic perspective and an aquatic therapist must have a very good understanding of these properties. Water has not shape and takes the shape of the container in which it is kept, it is usually liquid at room temperature in most of the parts of the world. For therapeutic use of water, water should be heated to be 30 to 34

degrees celcius.

**Some of the important physical properties of water are:**

**1.Buoyancy**

It is an upward force exerted by a fluid that opposes the weight of an immersed object. In a column of fluid, pressure increases with depth as a result of the weight of the overlying fluid. Property of water, that contributes to the weightless experienced by the patients and therapists. It is the force that counters the downward pull of gravity.

**2.Specific gravity**

Specific gravity, is also known as relative density, is the ratio of the density of a substance to the density of a reference substance; equivalently, it is the ratio of the mass of a substance to the mass of a reference substance for the same given volume. Apparent specific gravity is the ratio of the weight of a volume of the substance to the weight of an equal volume of the reference substance. The reference substance for liquids is nearly always water at its densest (at 4 °C or 39.2 °F). So the specific gravity of water is 1. The specific gravity of solids to be immersed in the water is computed taking water as reference substance. If the specific gravity of a substance is less than 1 it is less dense in comparison with water and therefore floats on the water where as if it more than 1 it sinks in the water. This is especially important for the therapists as the specific gravity of different body tissue is different and it changes based on various factors like injury, fibrosis, aging etc. therefore the therapist should be aware of the changes expected in the response of the body to water based on these changes. In a typical state, muscles and bones are heavier and sink in water where as fat and connective tissues are lighter and float on water.

**3.Adhesion**

It is the attraction between the molecules of substance. It is the property that keeps the substance together and resists deformation. This is the property that offers resistance for movement in water.

**4.Cohesion**

It is the attraction between the molecules of substance with another substance. It is the property that keeps two substances together and resists separation.

**5.Surface tension**

At liquid–air interfaces, surface tension results from the greater attraction of liquid molecules to each other (due to cohesion) than to the molecules in the air (due to adhesion). The net effect is an inward force at its surface that causes the liquid to behave as if its surface were covered with a stretched elastic membrane. Thus, the surface comes under tension from the imbalanced forces, which is probably where the term "surface tension" came from. Because of the relatively high attraction of water molecules to each other through a web of hydrogen bonds, water has a higher surface tension (72.8 millinewtons per meter at 20 °C) than most other liquids. This is the property of water that makes it difficult to move out of the water and enter in the water. This is the property that can be used to advantage by therapists while planning strengthening activities.

**6.Viscosity**

The viscosity of a fluid is a measure of its resistance to deformation at a given rate. For liquids, it corresponds to the informal concept of "thickness". Viscosity can be conceptualized as the frictional force that arises between adjacent layers of fluid that are in relative motion. The viscosity of water also provides resistance to the movement in water.

**7.Refraction**

Refraction is the change in direction of a wave passing from one medium to another or from a gradual change in the medium. For the discussion of therapeutic usage the refraction is the change in the direction of light waves as they enter water. This makes the object look crooked in water. This is important to understand that while assessing the alignment of the body refraction should be accounted for.

**8.Compressibility**

Compressibility is a measure of the relative volume change of a fluid or solid as a response to a pressure change. Water is incompressible; this property also contributes to the resistance offered by water.

**9.Hydrostatic pressure**

Hydrostatic pressure is the inward force exerted by water on an object that is immersed in the water. It is exerted equally in all directions. This is an important property to bring about many of the physiological effects of immersion in water.

Other important aspects and principles: of fluid dynamics

**Center of buoyancy:**

The Center of Buoyancy is the point through which the forces of buoyancy act on the body. As the density of different parts of the body is different the center of gravity and buoyancy are not located at the same point. This difference creates a rotational force in the body. When the volume of the immersed body is equal to the volume of the water displaced the object floats. To achieve flotation force of gravity and buoyancy must be equal.

Center of buoyancy in human body is located at the level of S1.

**Metacenter:**

Metacenter is point of insertion of straight lines drawn vertically through center of buoyancy at different positions. It is not a fixed point and keeps changing with moving object.

**Metacentric effect:**

Metacentric effect is seen when a body moves in water. As the body moves the position of metacenter in relation to center of gravity shifts. As the vertical distance between the metacenter and center of gravity reduces the body becomes more and more unstable. Human beings want to be aligned in a perfect symmetry to achieve floatation in order to orient the body in space. For this the metacenter should directly above the center of gravity the height of the metacenter from center of gravity determines the initial stability of the object. Greater the height better the stability. The reactive contractions created in the body to align the metacenter and the center of gravity is known as metacentric effect. These reactive contractions can be used by the therapist for training the patients and achieving therapeutic goals.

**Drag force:**

A drag force is the resistance encountered by moving object in water. It is in the direction opposite to the movement. Drag force increases in turbulent flow and reduces in laminar flow of water.

Other important principles are Archimedes principle, capillary action and Pascal's law.

**Physiological effects of immersion in water**

When immersed in chest deep water the hydrostatic pressure increases venous and lymphatic compression increasing the venous return and thereby stroke volume. Increased stroke volume reduces the heart rate. The hydrostatic pressure provides resistance for the rib cage movement and increases work of breathing. Several other mechanisms contribute to increased work of breathing. Work of breathing is increased by

60% at this immersion level. Therefore, therapists should be careful about deeper immersion in patients with respiratory muscle weakness and restrictive pulmonary disorders. Immersion reduces the loading of joints at chest level immersion 80% unloading is experienced. This also reduces the proprioceptive inputs at rest. However, propriceptive inputs can be increased by movement as the muscles experience greater resistance as compared to movement in the air. Immersion has effects of hormones and secretion of other chemicals like cytokines and chemokines. The cumulative effect of these hormonal changes brings about anti-inflmmatory and neuroprotective effect. Immersion also increased glomerular filration rate and therefore urine output. Also the loss of water through osmosis increases this can lead to dehydration if we do not rehydrate timely. These are some of the most important physiological factors that influence therapy however physiological effects of immersion change as per the depth of immersion, composition of water, temperature of water and duration of immersion.

**Benefits of aquatic therapy in Neurological Disorders**

- Improves sensorimotor stimulation
- Helps to improve balance
- Improved vestibular stimulation
- Reduction in the gravitational forces therefore reduced joint loading in awkward postures
- It improves the blood supply to the deeper muscles and helps to slow down the fibrotic processes.
- Reduce the work of the muscles thereby preventing accelerated muscle damage in neuromuscular disorders
- It provides a higher degree of freedom of movement
- It has a distinct physiological mood enhancing effect which may prevent negative emotional responses.
- Improved blood supply to the brain improving the cognition
- In case of musculoskeletal deformities, aquatic environment provides freedom to unload and stretch
- Aquatic therapy also improves respiratory capacity and cardiovascular endurance.
- Analgesic and anti-inflammatory effect
- Increased blood supply, hydrostatic pressure of the water, improved endorphins and serotonin levels also helps to improve appetite and bowel movements.
- Aquatic exercises help reduce sleep disturbances.

**Aquatic therapy intervention**

Aquatic therapy does not mean just swimming in water. Aquatic therapy includes purposeful therapeutic movements or exercises performed in order to achieve optimum benefits. There are various techniques in aquatic therapy. An exercise session will consist of a combination of these techniques and approaches like Water specific therapy ®, Bad-Ragaz ring method ®, Clinical Ai-Chi ®, Aquatic exercises, Aqua aerobics, myofascial mobilization and Passive relaxation or Watsu®. Mostly exercise session will be conducted one - on - one by the therapist but group sessions may also be conducted to improve participation and peer interaction.

In the beginning the exercise session will emphasize on adaptation to water environment and being comfortable in water as well achieving breathing control in water. This is to ensure safety of the client in the new aquatic environment. Therapist may choose to engage the patient in various play

activities on the surface of the water. This may be followed by respiratory and oromotor control in water and slow introduction to under water environment facilitating better breath control and greater adaptation to the new environment. This will be followed by various rigorous goal oriented activities.

**Aquatic therapy set up**

Aquatic therapy is performed in a therapeutic pool. Pool area of 2 meters x 2.25 meters is recommended for the therapy of one patient and therapist couple. Larger area may be required for some of the techniques like Bad Ragaz Ring Method ® and Watsu ®. The pool should be temperature controlled and most of the disorders are treated in a heated pool but some may be required to be treated in cold temperatures. There should ease of accessibility to accommodate for patients impairments and limitations. Usually a ramp into the pool or a pool lift can be used. There should be facilities of toilet and shower in immediate vicinity of the pool that are easily accessible to the patients. The therapists should be trained in Basic Life Support techniques and emergency water evacuation techniques.

**Contraindications and relative contraindications for performing aquatic therapy**

Open wounds that cannot be covered satisfactorily with water proof bandaging, active infections, contagious disorders, active skin allergies and infections, low LVEF, respiratory insufficiency, cardiovascular insufficiency and pyrexia are some of the contraindications. Therapists should be precautious when the patients are using external devices like catheters and riles tube. Precaution should be taken in case of hypertension and cardiovascular disorders to monitor the cardiorespiratory response during the session. Patients who are hydrophobic should be allowed time for safe entry in water. Precaution should be taken in case of reactions and allergies due to chemical disinfectants used in water and immediate medical attention should be sought.

**Is aquatic therapy an alternative to land based therapy?**

No, aquatic therapy is not an alternative but an adjunct to it. Land based rehabilitation and aquatic rehabilitation needs to be performed together. Neither is alternative to the other. There are different advantages and applications of both the disciplines in neurorehabilitation.

**Benefits of aquatic therapy over land based therapy**

- There is less weight on the joints, the client is well supported and joints are not under stress like on land. Therefore aquatic therapy helps achieve the benefits of land based therapy without causing any harm to the joints.
- As the individuals are able to perform some of the tasks in water much easier than on land, their confidence and activity participation increases.
- Activities in water are more fun and interesting for the individuals therefore there is better engagement of the individuals in a session and better adherence to therapy than on land.
- For individuals with severe movement restriction on land. Aquatic environment provides some freedom for movement.
- The risk of fall significantly reduces in aquatic environment
- As the frequency for lengthening contractions in the water environment is much lesser than on land the damage to the muscles while exercising is lesser in case of neuromuscular disorders. However, different therapeutic strategies can facilitate lengthening contractions

when required in neurological conditions.

- It improves blood circulation to the deeper muscles better than on land and improves pliability of the muscles.
- The fibrotic changes in the muscles are lesser than on land
- The reactive behaviors help to improve activation of muscles

Therefore aquatic therapy may be preferred during some stages of the disease. It provides an excellent medium to train the individual and improve their motor and cognitive impairments. It is fun and enjoyable ensuring long term adherence. It helps to maintain various cardio-respiratory health parameters. It is safe and very effective in improving the quality of life of individuals with neurological disorders. However aquatic therapy alone is not sufficient and must be incorporated in the multidisciplinary rehabilitation program including land based exercises.

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# Chp. 25 Orthotics and Prosthetic Management

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## LOWER LIMB ORTHOSIS

A lower limb orthosis is an external device applied or attached to a lower extremity to improve function by controlling motion, providing support, reducing pain through transferring of the load to another area, correcting flexible deformities, preventing the deformities getting worsened.

Orthosis is the medical terminology for brace or splint.

- **AFO:** Ankle Foot Orthosis.
- **KAFO:** Knee Ankle Foot Orthosis.
- **HKAFO:** Hip Knee Ankle Foot Orthosis.
- **THKAFO:** Trunk Hip Knee Foot Orthosis.

**Orthosis are of two types:**

1. **Static:** Support the weakened joint.
2. **Dynamic:** Allows joints movements during locomotion.

**Materials used are of:**

1. Metal
2. Plastic
3. Leather
4. Synthetic fabrics.

Plastic materials such as thermosetting materials / thermoplastic are commonly used. They can be easily molded after heating and hardened after cooling. Low temperature thermoplastics can be fabricated easily.

**Principles:**

In all orthosis

1. 3-point pressure is essential.
2. Orthotic joints should be at the anatomical joint level
3. Should be light weight, strong, durable and cosmetically acceptable.
4. Static or dynamic stabilization.
5. Flexible material. Tissue tolerance to compressive and shear force.
6. Thermoplastics (polypropylene) requires high temperature (150 degrees Celsius) for molding and it is used mainly for high stress activities.
7. Rubber is used for padding
8. Metals are used for joint components and uprights.

**External Shoe Modifications:**

1. **Cushion heel:** to minimize impact on heel, encourage plantarflexion and minimize knee flexion.
2. **Heel wedge:**
   a) Medial wedge to promote inversion (for flat feet).
   b) Lateral wedge to promote eversion (for varus foot)
3. **Crooked and elongated heels:** Medially (for flat feet) Reverse crooked and elongated heels (C / E Heels) i.e. elongated lateral part of heel (for varus foot)
4. **Heel elevation:** to compensate fixed

equinus deformity or length discrepancy.

5. **Compensation platform:** for length discrepancy

**External Shoe Modifications:**

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9. Heel elevation: to compensate fixed equinus deformity or length discrepancy.
10. Compensation platform: for length discrepancy

**Sole Modifications**

1. Metatarsal bar at posterior metatarsal head to shift the weight bearing on metatarsal heads, commonly used in Diabetes Mellitus and anesthetic foot.
2. **Sole wedge:** Medial wedge to promote supination of the foot

Lateral wedge to promote pronation.

**Internal Shoe Modifications:**

1. Cushion Heel: Soft pad with scooped out central part under the painful points.
2. Heel wedge: When given medially, can promote inversion of hind foot.

   When given laterally, can promote eversion of hind foot.
3. Metatarsal pad: shifts stress from metatarsal head to meta tarsal shaft.
4. Innersole excavation: at one or more Metatarsal heads.
5. Valgus pad or medial arch support. (for flat feet)

**Foot Orthosis:**

1. **Plastic insert:** This is a rigid plastic insole for manual correction. It has rigid medial, lateral or posterior walls.
2. **Heel Cup:** Plastic cup under the heel with rigid wall posterior and medial and lateral aspect of the heel (for pronated heel)

**Custom made orthosis are always better than ready made devices.**

Ankle Foot Orthosis (AFO):

- Prescribed for weakness of

  1) ankle dorsiflexors

  2) plantar-flexors

  3) inverters

  4) evertors
- They are used for maintaining the feet in the neutral position while walking.
- They are also used to prevent deformities.
- In a case with an UMN weakness of the foot with spasticity in the inverters and plantar-flexors, which would give an equino- varus tendency of the foot, it is prescribed to maintain the foot in corrected position and also prevent foot drop. This would help in the gait training
- The position of foot affects the stability of knee indirectly.

1) Equinus foot with spasticity could lead to hyper extension of knee.
2) Calcaneal foot may lead to knee flexion.
3) Varus foot may cause Genu varum
4) Valgus foot could lead to Genu valgum

- An AFO can reduce energy consumption in Diplegics, Hemiplegics and in LMN foot drop.

**TYPES OF AFO**

**1. Fixed Ankle:**

a) Plastic insert

b) High boots with steel shank.

2. **Hinged AFO:** Adjustable ankle hinges with desired ankle plantarflexion or dorsiflexion.

Conventional below knee braces with ankle or without ankle joints, along with medial T-strape (for valgus foot) or lateral T-strape (for varus foot)



*Fig 1: High boots with*



*Fig 2: Plastic AFO steel shank*



*Fig 3: Plastic AFO*

**KAFO (Knee Ankle Foot Orthosis)**

KAFO is prescribed whenever there is an inability to lock the knees due to weak muscles around the knees (Quadriceps and Hamstrings). There is a tendency for buckling of the knee.

**Types of KAFO**

1. With Knee Joint
2. Without Knee Joint
3. Ischial Weight Bearing
4. With Free Knee Joint

- Knee Joint is provided for the patient to wear a brace and fold the knee.
- Ischial Weight Bearing KAFO is prescribed when the weight bearing has to be on the ischium (in cases of fractures of the weight bearing bones like femur or tibia)
- KAFO with free knee joint is prescribed whenever there is strong quadriceps but the knee joint has tendency for hyperextension.

Many a times, trial splints like Push Knee Splints are prescribed to see whether the patient can manage locking the knee and walking. (Provided the foot has got proper control)



*Fig 4: Knee Ankle Foot orthosis*



*Fig 5: Push knee splints*

**Hip Knee Ankle Foot Orthosis (HKAFO)**

HKAFO is prescribed whenever there is a lack of control of hip and knee. A pelvic band is provided in order to give Hip stability. Hip is controlled mainly by hip extensors and abductors, in the absence of which, the hip

becomes wobbly and with bilateral involvement walking is very unstable. Thus patient may require use of a pair of crutches or a walker



*Fig 6: Hip Knee Ankle Foot Orthosis*

**SPINAL ORTHOSIS**

Spinal Orthosis are prescribed for

1. Unstable Spine due to weak musculature
2. Unhealed Fractures of the spine
3. Correction of deformities like Scoliosis, Kyphosis, etc
4. Spinal Dysraphysm e.g. Meningo-myocoele, Spina Bifida
5. Post surgical conditions like Laminectomy

**Types of spinal orthosis**

1. Lumbosacral belt or corset
2. Thoracolumbar Corset
3. Close Contact Spinal Orthosis
4. Milwaukee Brace
5. Taylor's Brace
6. Minerva Jacket
7. Anterior Hyperextension Brace

By far, a close contact spinal jackets can give the best support for scoliosis and other deformed spine.



*Fig 7: Close Contact Spinal Orthosis*



*Fig 8: Lumbosacral belt or corset*



*Fig 9: Taylor's Brace*



*Fig 10: Anterior Hyperextension Brace*

**CONCLUSION:**

Various types of orthotic devices are useful for preventing deformities getting worse, maintaining corrected position of the lower extremities and also improving the gait pattern as much as possible. They compensate for the weak muscles for stability. Prescribing the correct orthotic

devices is one of the most essential part of Rehabilitation, especially in Neurological conditions

**UPPER LIMB ORTHOSIS**

**Definition**

An orthosis is a device that is applied to the body in order to protect and stabilize body parts, to prevent or correct scarring and deformities, or to aid in performance of certain functions. Upper limb orthoses are applied to the shoulder, elbow, arm, wrist, or hand. These devices may be called orthoses, orthotic devices, or splints.

Upper extremity orthoses are used frequently on patients who have

- musculoskeletal problems like those resulting from trauma, sports, and work-related injuries.
- neurologic problems, such as stroke, traumatic brain injury(TBI), multiple sclerosis (MS), cerebral palsy (CP), spinal cord injury (SCI), and peripheral nerve injury.
- arthritic conditions

**Purpose of Splinting**

Upper limb orthoses can be used for a wide variety of purposes. Some of the more common uses include:

- Stabilizing fractures or unstable joints
- Immobilizing joints to promote healing
- Preventing or correcting joint contractures
- Protect weak muscles from overstretch
- Correcting subluxation of joints or improper alignment of tendons
- Helping to provide enhanced function
- Serving as an attachment for assistive devices
- Blocking unwanted movement of a joint
- Maintaining correct joint alignment
- Assisting movement of joints
- Reducing muscle tone in spastic muscles
- To reduce weight bearing forces for a particular purpose
- To otherwise correct the shape and / or function of the body, to provide easier movement capability or reduce pain

**Team Approach**

Creating and employing upper limb orthoses often involves a team approach, especially in rehabilitation settings. Custom-made upper extremity orthoses may be fabricated by physical, occupational, and hand therapists. The therapist may help the patient learn to use the orthosis but can also recommend ready-made devices or refer the patient to an orthotist.

Health care professionals who create and fit upper limb orthoses must have a good understanding of the anatomy and physiology of the upper limbs. They must also understand the mechanics and forces involved in making various body movements, and they must be familiar with the materials and tools involved in constructing orthoses. Certified orthotists are specialists who focus exclusively on fitting and building orthoses.

The professional relationship between the physical / occupational / hand therapist and orthotist is very important in identifying patient functional goals and a variety of predictable outcome.

**Basic Principles in Splinting:**

I. Mechanical Principles: Use of mechanical principles helps make splints comfortable, durable, effective and diminishes of additional injury secondary to splint application.

Mechanically, splints may be grouped into two categories:

1. Those that apply three point pressure: Three point pressure splints function through a series of reciprocal forces with the middle force directed opposite to the two end forces.
2. Those that pull adjacent articulated bony segments together through circumferential pressure. In the circumferential pressure splint, the middle reciprocal force is absent.

Pressure and shear can be reduced by increasing the surface area of the splint. Short narrow splints or components are often problematic since they apply pressure to a small area creating pressure necrosis of underlying soft tissues. Longer splint designs increase mechanical advantage and make splints less susceptible to causing pressure problems. (Willard)

Other Mechanical principles that need to be taken into consideration are

**1. Use Optimal Rotational Force**

- Rotational element- To produce joint rotation
- Translational element- To produce joint distraction or compression

**2. Consider the Torque Effect**

- Increase torque by increasing the distance between the joint axis and the point of attachment of the dynamic assist

3. Consider the Relative Degree of Passive Mobility of Successive Joints
4. Consider the Effects of the Reciprocal Parallel Forces

- Use first -class lever system
- If Mechanical Advantage is increased, the middle opposing force is decreased

5. Provide Contours

- Eliminate Frictions by proper padding, forces, stockinet, etc

**ii. General Principles of Design**

1. Consider individual patient factors such as age, intelligence, motivation, body size, activity level, socio economic status etc.
2. Consider the length of time the splint in to be used

- Shorter the anticipated need of splint, simpler its design, material type and construction should be.

3. Strive for simplicity and pleasing appearance
4. Allow for optimum function of the extremity

- Avoid needless immobility of normal joints

5. Allow for optimum sensation

- Palmar tactile surface areas should be free

6. Allow for quick, low cost and efficient construction and fit.

- Provide for ease of application and removal
- Avoid dependence on others for assistance

7. Design for several functions into one splint to avoid confusion and simplify the wearing and exercise routines.

**iii.Specific Principles of Design**

1. Identify the key joints to be splinted by careful evaluation.
2. Review purpose to immobilize, to increase passive motion or to substitute for active motion by re-evaluating for functional objective of a given splint.
3. Decide whether to employ Static or Dynamic forces.
4. The surface of the extremity on which the splint will be based is determined and secondary joints that need to be controlled or positioned are noted.

5. Identify area of diminished sensibility to avoid pressure necrosis.
6. Consider kinetic effects: Since application of the splint often alters internal and external forces to proximal or distal joints, the kinetic effects of the splint must be considered including what will be the forces on un- splinted joints and what will be the ramifications to extrinsic and intrinsic musculo-tendinous structures.
7. Choose the most appropriate materials and adapt to general properties of the selected splint materials.

**IV.Principles of Construction**

1. Use fabricating equipment appropriate to material.
2. Use type of heat and temperature appropriate to the material
   - Over heating or under heating can disrupt the material.
3. Strive for good cosmetic effect.
4. Round corners and smoothen edges
   - For increased strength, durability, cosmesis and comfort
5. Stabilize Joined Surfaces
   - By using one or two rivets as per the requirement
6. Rivets should be flush with the material surface or apply tape / moleskin over internal metal rivet to prevent rusting.
7. Provide small ventilation holes to improve air circulation
8. Perforation should be made from inside out to give a smoother inner surface
9. Secure Padding

- Allow ease of splint application and removal
- Apply extra padding to the edges, to allow curl around splint edges

10. Secure straps to provide splint stability by gluing or riveting and the strap ends should be rounded and smoothed to prevent fraying.

**V. Principles of Fit**

a. Mechanical Considerations

**1. Use Principles of mechanics**

- The mobilizing force should always be directed perpendicular to both t2he segment being moved and to the rotational axis of the joint
- The leverage system applied (to the placement of straps, finger cuffs and fingernail hooks) should help in dissemination of the applied force and reduce pressure and elimination of friction.

**b. Anatomic Considerations**

Contiguous fit of the splint to the extremity reduces pressure on bony prominences as well as soft tissue.

**1. Bone**

- Accommodate bony prominences

- To avoid tissue necrosis

- Incorporate Dual Obliquity concepts

- The progressive decrease of length of the metacarpals from the radial to ulnar aspects of the hand

- The immobility of the second and third metacarpals as compared to the mobile first, fourth, and fifth metacarpals

**2. Ligaments**

- Consider ligament stress

- Preserve the ligament structure in correct position and tension to prevent inflammation, attenuation and disruption of ligament tissue

**3. Arches**

- Maintain the skeletal arches (proximal transverse, distal transverse and longitudinal arches)

**4. Joints**

- Align splint axis with anatomic axis
- Use optimum rotational force in 90 degree angle of pull

**5. Skin**

- Use skin creases as boundaries

Kinesiologic Considerations

1. Allow for Kinematic changes when fingers and thumb are in flexion or in extension
2. Employ kinetic concepts

**Technical Considerations**

1. Develop patient rapport and trust

- Give instructions and explanations regarding the importance and function of the splint

2. Work efficiently
3. Change method according to properties of material used

Prefabricated or Custom fabricated Orthoses
Prefabricated orthoses are easily available in a cost effective and timely manner. As a general rule,

prefabricated orthoses should be considered for patients having a normal anatomy, and or who will require the orthosis for a short period of time. Though these prefabricated orthoses are easy to apply, the therapists need to understand the

indications, contra indications and limitations of these devices.

Custom made orthoses are generally used when orthotic device is required for extremities or when the spine have deformities or in case of unusual sizes and / or when they have to be used indefinitely

The advantages of custom fabricated orthotics are:

- Designed to patient specification
- Easily adjusted if uncomfortable
- Any position requested can be achieved with the low-temperature plastic
- Able to clean as needed
- Can get wet
- Adjustable as swelling decreases or as able to get into a better position

**Materials and construction**

Although ready-made orthoses are available for some applications, many are custom made to fit the specific needs of each patient. The material used in orthotic devices typically includes low-temperature thermoplastics that are readily shaped for fit and function. Other materials used include casting, metal, straps, and hook-and-loop. Most orthoses have employed lightweight thermoplastic materials, which are plastics that become pliable when they are heated and retain their shape once they cool. The thermoplastic sheets can be molded to fit body parts exactly, and some can be reshaped repeatedly as the treated body part changes shape. The resulting orthotic device is lightweight and relatively easy to use and maintain.

Thermoplastic materials are usually classified into high- and low-temperature types, based on the temperature at which they become pliable. High- temperature thermoplastic materials must be molded at a temperature that is too high to come in contact with human skin. These materials must be molded over a plaster model of the body part, but have the advantage of being stronger and more durable than low-temperature thermoplastics. They are used in situations where the orthosis will undergo a lot of stress or will be used for a long time. High-temperature thermoplastics

require special tools for cutting and shaping, and orthoses made from these materials are usually constructed by an orthotist, a technician who specializes in making these devices.

Many upper limb orthoses are constructed of low- temperature thermoplastics. These material becomes pliable below 180°F (80°C), and can be molded directly against the body. It is relatively easy to cut and shape, and many therapists construct orthoses using these materials. Precut shells made from low-temperature thermoplastics are also available. The therapist can use a precut thermoplastic shell as the base for a device and then modify it to fit by trimming and adding pads and straps. Orthoses made from low-temperature thermoplastics are commonly used in situations in which the orthosis will receive relatively little stress or is intended for temporary use. These orthoses are especially important when a device is needed quickly, such as in postsurgical or trauma treatment.

Both high- and low-temperature orthoses must be attached to the body. Most modern orthoses use straps made of hook-and-loop tape for this purpose. This material is lightweight, durable, and readily adjustable, and comes in a variety of widths and colors. Orthoses can also include padding to cushion sensitive areas, as well as specialized linings. Patients often use a separate interface that absorbs perspiration and protects the skin, and which can be washed or replaced as needed.

**Types of orthoses**

The upper limbs comprise a complex system of muscles, joints, ligaments, and tendons, which are capable of a number of distinct movements. For this reason, a wide variety of upper limb orthoses have come into existence. These devices often go by multiple names, reflecting the name of the manufacturer, the name of the person who developed the device, or the anatomy and function it serves.

In the 1970s, a group of American professionals involved in the field of orthotics-put forth an effort to classify orthoses by their function and acronyms describing the joints that are encumbered by the orthoses. From this effort sprung the current nomenclature : AFO- ankle foot orthosis, TLSO- thoraco- lumbo- sacral orthosis, WHO- wrist hand orthosis, etc. However, the nomenclature based on the function of the orthosis; such as assist dorsiflexion at ankle, limit wrist flexion to 10 degrees, resist thoracolumbar rotation has not found wide spread acceptance.

**Orthoses are usually classified under the following three categories:**

-static,

-serial progressive

-dynamic or functional

Static orthoses hold a body part in a fixed position and do not allow joint movement. Some static orthoses do not contain joints, as with fracture orthoses that stabilize the long bones of the arm after a fracture. Most others simply maintain the joint at a particular angle, providing support and proper positioning. For example, a static wrist orthosis can be used to hold the wrist in a neutral position to promote healing and prevent injury during activities. Sometimes static orthoses include attachments that help patients perform functional activities. For example, a hand-wrist orthosis may include an attachment for pens or eating utensils. Static orthoses sometimes serve the function of promoting eventual joint movement.

**Complications associated with use of static orthoses**

1. Skin breakdown
2. Contractures

3. Infection

Serial or progressive orthoses loosen joints that have become frozen due to contractures or arthritis. Serial orthoses involve several similar devices used in a series, with each successive device gradually increasing the range of motion of the affected joint by providing a gentle stretching action.

Progressive orthoses accomplish similar goals, but do so by allowing adjustments in the device so that it gradually increases the amount of stretch created in the joint.

Serial and progressive orthoses must be designed and used carefully to provide the correct amount of stretching in the joint. Excessive stretching can damage the tissues, and inadequate stretching will be ineffective.

Dynamic / functional orthoses allow or create joint movement. These devices hold the joint in the proper position while assisting movement using springs, rubber bands, or other mechanical features. Dynamic orthoses are useful for patients who have weakened muscles or limited neuromuscular control, because they allow the patient to perform actions that would be difficult or impossible without assistance. These devices promote independence in patients who have handicapping conditions, and they are common in rehabilitation settings. Since no single device can perform all the movements that the human hand can perform, the patient may need to use several different dynamic devices in order to carry out activities of daily living. Some dynamic splints have a dual or bilateral tension-providing mechanism that can safely accommodate moments of spasm and thus potentially limit or prevent soft-tissue injuries.

One study indicated that patients who have sustained a stroke can be aided by a gravity-compensating arm orthosis that helps to loosen the grip of the affected hand and help stroke patients gain greater functional independence.

However, a systematic review that examined 4 trials including 126 patients with stroke or other nonprogressive brain lesions found that upper limb orthoses showed no effect on upper limb function; ROM at the wrist, fingers, or thumb; or pain.

**Types of upper-limb orthoses**

- Upper-arm orthoses
- Clavicular and shoulder orthoses
- Arm orthoses
- Functional arm orthoses
- Elbow orthoses
- Forearm-wrist orthoses
- Forearm-wrist-thumb orthoses
- Forearm-wrist-hand orthoses
- Hand orthoses
- Upper-extremity orthoses (with special functions)

**Clavicular and shoulder orthoses include the following:**

- Figure-8 harness/clavicular brace - This is used to restrict motion in patients with clavicular fractures so as to allow tissue healing and bone remodeling
- Shoulder sling - This is used to restrict motion in subluxated shoulders by providing humeral cuff and chest straps to keep the humeral head in the glenoid cavity
- Overhead sling suspension - This is used for patients with proximal arm weakness or paralysis to allow hand or arm use when the muscles are at least antigravity in strength
- Hemiplegic arm sling - This is used for immobilization of the hemiplegic shoulder, which helps to decrease pain

and subluxation (Fig 11)

Functional arm orthosis are used primarily in patients with proximal arm weakness involving the shoulder and arm, such as that resulting from spinal cord injury (SCI) or peripheral nerve lesions. They include a shoulder saddle from which a proximal forearm cuff is suspended by means of straps or a Bowden cable.

**Elbow orthoses include the following:**

- Posterior elbow splint - This is used particularly for elbow immobilization in patients
- Serial cast - This is used for preventing or correcting contractures through promotion of soft-tissue stretch and passive range of motion (ROM)
- Air splint - This is a form of circumferential inflatable sleeve, used to maintain or increase elbow extension and also used for contractures and elbow immobilization
- Dynamic elbow splints, static progressive elbow splints, turnbuckle elbow splints- gently elongate the soft tissues over a long period to attempt to reverse joint malalignment (contractures, burns, and late phase of fracture). They are not used in spastic muscles as they may further increase tone.
- Dorsal elbow-extension mobilization orthosis - extend the elbow as well as provide mediolateral elbow stability and rotational forearm stability
- Dorsal elbow - flexion mobilization orthosis

  - flex the elbow and provide mediolateral stability and rotational forearm stability

**Forearm orthoses**

Balanced forearm orthosis (BFO) - This supports the weight of the forearm and arm against gravity and is used primarily in patients with high-level tetraplegia or severe proximal arm weakness or paralysis; it may be attached to a wheelchair or table; patients may be able to perform tabletop activities; prerequisites for its use include a power source, such as neck or trunk muscles (to shift the trunk center of gravity) or adequate scapular movement

**Requirements**

- Some residual muscle strength (MMT at least poor or grade 2) and coordination of elbow flexion (can be used for C5 quad)
- Adequate trunk stability and balance
- Adequate endurance in a sitting position
- Preserved ROM of the shoulder and elbow joints

Other uses: they also may be used in spastic patients to allow them to self-feed by dampening muscle tone through a friction device

Forearm mobilization (corrective) orthosis- dynamic supination / pronation splints used to increase supination or pronation in forearm rotational contracture, or to increase passive or active-assisted ROM in spinal cord injury

Shoulder-elbow-wrist-hand orthosis (SEWHO) consists of a forearm trough (attached by a hinge joint to a ball-bearing swivel mechanism) and a mount (which can be mounted on the WC, on a table or working surface, or onto the body jacket). It helps support the forearm and arm against gravity and allows patients with weak shoulder and elbow muscles to move the arm horizontally and flex the elbow to bring the hand to the mouth (e.g., patients with spinal cord injury, Guillain-Barré Syndrome, polio, muscular dystrophy, and brachial plexus injury).

Forearm-Wrist Orthoses: Forearm-wrist orthoses may be either volar or dorsal and either gutter- based or circumferential. Ideally, the wrist should be positioned in 15-

30° of dorsiflexion (wrist extension), except in carpal tunnel syndrome. The volar type should allow for metacarpophalangeal (MP) flexion by ending before the distal palmar crease.

**Examples of forearm-wrist orthoses include the following:**

- Wrist cock-up splint
- Wrist extension splint
- Ulnar gutter splint

**Volar wrist-hand stabilizers/wrist cock-up splint**

- Wrist-hand orthosis (WHO) extending from distal two-thirds inch of forearm to about one- quarter proximal to the distal palmar crease to allow full metacarpophalangeal flexion while maintaining the functional position of the wrist and hand
- Uses include resting wrist and hand in acute arthritis (RA), wrist sprain / contusion, flexor / extensor tendinitis, carpal tunnel syndrome, postsurgical wrist extensor tendon repair, wrist fusion, and skin grafting, contractures prevention, reduce pain, reduce spasticity, prevent ulnar / radial deviation wrist / hand (e.g., RA)

**Dorsal wrist-hand stabilizers**

WHO are used to provide the same functions of the volar WHO as well as greater stabilization because of rigid dorsal hand section

- More difficult to fabricate and fit than the volar WHO



*Fig 11: Hemiplegic arm cuff / sling*



*Fig 12: Resting hand splint*



*Fig 13: Basic Opponens Splint*



*Fig 14: Long opponens splint*



*Fig 15: 'C' bar splint*



*Fig 16: Ulnar deviation correction splint*

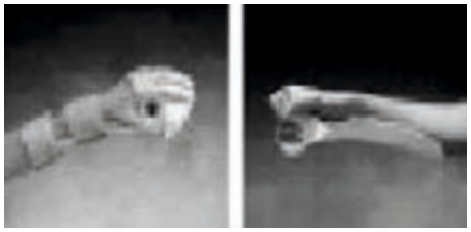


*Fig 17: (A) Antispasticity "Ball" splint (B) Hand Cone splint*

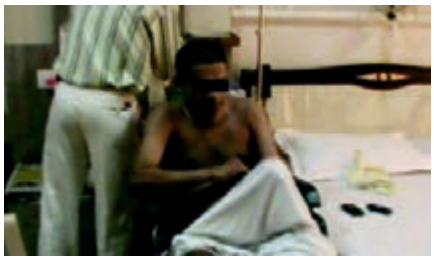


*Fig 18: Universal Cuff*

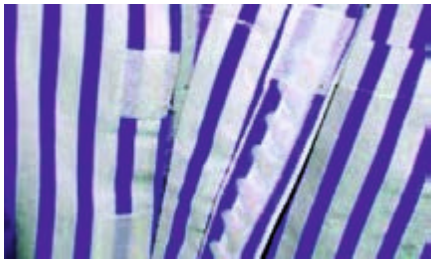


*Fig 19: Patient Using U-cuff for brushing teeth*

Forearm-Wrist-Hand Orthoses: The basic types of forearm-wrist-hand orthoses are the resting hand splint (see the image below), the functional resting splint, and the static hand splint. Other types include the burn splint and the weight-bearing splint.

Placement may be dorsal, volar, or circumferential and extends from the tips of the fingers to cover two thirds of the distal forearm. The dorsal type of splint is particularly useful in patients who demonstrate palmar hypersensitivity and grasp reflex. The position of the wrist is neutral or in slight dorsiflexion. Immobilization in this position preferred because metacarpophalangeal joint and Interphalangeal collateral ligaments are kept stretched, minimizing future joint capsule contractures. In addition, it provides functional thumb position for opposition and three-jaw chuck pinch.

**Functions of forearm-wrist-hand orthoses include the following:**

- Immobilization in patients who have hand flexor and extensor tendinitis or who are undergoing tendon, nerve, or fracture repair.
- Maintenance of passive range of motion (ROM) in patients with upper motor neuron lesions and contractures

**Wrist Orthoses**

**1.Wrist control orthosis**

Promotes slight extension of the wrist or prevent wrist flexion, thus assisting weak grasp (via tenodesis effect)

a. Volar wrist-flexion control orthosis (cock-up splints)

- Wrist-hand orthoses (WHOs) in which the palmar section is extended (usually 20°. They are used to tighten finger flexors (via tenodesis effect) and prevent wrist flexion contracture in patients with radial neuropathy.

b. Wire wrist-extension assist orthosis (Oppenheimer splint)

Prefabricated from spring steel wire and padded steel bands to assist wrist extension by tensing the steel wire, thus aiding finger flexion through tenodesis effect

**Wrist-driven prehension orthosis (tenodesis orthosis, flexor hinge splint)**

Used in C6 complete tetraplegia (in which no muscles to flex or extend fingers remain innervated but wrist extension, through the extensor carpi radialis muscle, is intact) to provide prehension trough tenodesis action and maintain flexibility of the hand, wrist, and elbow.

- Wrist extensors should be 3+ or better to use body-powered tenodesis
- May interfere with manual WC propulsion.
- Rarely accepted by C7 and C8 tetraplegics who prefer to use their residual motor power or utensil holders.

**RIC tenodesis splint**

- Orthosis made of low-temperature

thermoplastics in three separate pieces (wristlet, short opponens, and dorsal plate over index and middle finger)

- Easily and quickly fabricated; made as a training and evaluation splint for patients; light weight.
- Uses a cord / string running from the wrist piece, across the palm and up between the index and ring fingers. The string is lax when the wrist is flexed and tightens with wrist extension, bringing the fingers close to the immobilized thumb, accomplishing three-jaw chuck prehension.

**Wrist driven prehension orthosis.**

i. Wrist extension: three jaw chuck

ii. Wrist flexion: release

**Hand Orthoses**

-Wrist, Hand, and Finger Static Orthoses

**Positional orthoses**

Opponens orthoses : primarily used to immobilize the thumb to promote tissue healing and / or protection or for positioning of the weak thumb in opposition to other fingers to facilitate three-jaw- chuck pinch. Examples: short opponens splints, C- bar splints, cone splints, static thumb splints.

**Opponens orthoses with wrist control attachments**

- Ex: long opponens splints and thumb spica splints
- In addition to the benefits already mentioned for opponens orthoses (stabilizes first MCP), forearm bar maintains wrist in extension and prevent radial and ulnar deviation deformities

**Opponens orthoses with lumbrical bar**

- Finger orthosis that prevents metacarpal phalangeal joint hyperextension but allows full MCP flexion
- Prevents claw hand deformity (in addition to the benefits already mentioned for opponens orthoses)

**Opponens orthoses with finger extension assist assembly**

- Similar to basic opponens orthoses plus finger orthosis that assist proximal interphalangeal and distal interphalangeal extension
- Used for interphalangeal flex contracture, boutonnière deformity, or postsurgical release of Dupuytren's contracture.

**Utensil holders/universal cuff (splints)/ADL splints**

- Consist of a handcuff with palmar pocket onto which a utensil can be inserted.

Protective orthoses: used to protect wrist, hand, and / or fingers from potential deformity or damage by restricting active function / limiting motion

**Digital stabilizers:**

**1.Finger stabilizers/static finger orthoses (FOs):**

Interphalangeal stabilizers (DIP, PIP, and DIP+PIP gutter splints, static finger splints, stax or stack splints, egg-shell finger casts, etc.)

- FOs used to restrict motions at the PIP and DIP
- Generally, IPs maintained in full extension to keep the collateral ligaments stretched and to prevent IP flex contracture (unless condition dictates otherwise)
- Used to promote healing (e.g., phalanx fx, PIP / DIP dislocation, etc.) and to provide prolonged finger stretch (e.g., burns and contractures)

**2. Finger mobilization orthosis**

**a. Interphalangeal extension-mobilization orthoses-passively extend the PIP joints**

- Uses: Finger IP flexion contracture, Boutonnière deformity, and postsurgical release of Dupuytren's contracture
- Examples: Dynamic IP extension splints, reverse finger knuckle benders, Capener splints, safety-pin splints, spring coil assist, eggshell finger extension casts, buddy splints

**b. Interphalangeal flexion mobilization orthoses-passively flex PIP joints**

- Use: Finger IP extension contracture
- Examples: Dynamic IP flexion splints, finger-knuckle benders, fingernail book orthoses, buddy splints

**Ring Stabilizers:**

- Swan neck ring-FO that prevents hyperextension of the PIP joint (via three point pressure system) but allows full IP flexion
- Boutonnière ring-FO that immobilizes the PIP in extension (prevents flexion) through a three point pressure system.

**Metacarpophalangeal ulnar-deviation restriction orthosis:**

- FO used to limit ulnar deviation of the MCP with unrestricted (if possible) MCP flex / extension in arthritic patients with ulnar deviation at the MCPs.

**Metacarpophalangeal (MCP) mobilization orthoses**

a. MCP- extension mobilization orthoses-

- Uses: extend MCP joints in MCP-flexion contractures, burns, and post ORIF of metacarpal fracture, patients with weak finger extension (e.g., radial nerve lesion and brachial plexus lesion).
- Examples: Reverse MCP knuckle benders, dynamic MCP extension splints with dorsal outrigger, MCP extension assists; radial nerve splints

b. MCP-flexion mobilization orthoses

- Uses: used to flex MCP joints in MCP collateral ligament contractures, extensor tendon shortening, median / ulnar lesion, claw hand, post capsulotomy, post ORIF of metacarpal fracture.
- Examples: MCP knuckle benders, dynamic MCP flexion splints with volar outrigger and fingernail hooks, MCP flexion assists

**Thumb stabilizers**

**Thumb carpometacarpal stabilizers/thumb posts**

- Thumb orthosis that stabilizes the first CMC and MCP joints in neutral position to protect the thumb form inadvertent motion. Thumb- web space stabilizers / thenar web spacers / c- bar splint.
- FO that consists of a rigid C-shaped splint held firmly in the thumb and index finger web space.
- Function: increases or maintains the thenar space and prevents web-space contractures
- Uses: Burns, postsurgical revision of scar, web- space contractures

**Thumb mobilization orthoses**

a. Thumb extension-mobilization orthosis-dynamic thumb IP extension splints. Use: thumb IP flexion contracture.

b. Thumb flexion-mobilization orthosis-

dynamic IP flexion splints

Use: Thumb IP flexion contractures.

c. Thumb abduction-mobilization orthosis-

dynamic thumb abduction splint.

Use: Thumb-adduction contracture.

**Upper Extremity Orthoses With Special Functions**

Tone-Reducing Orthosis

Theoretical basis for tone-reducing orthosis

- Inhibition of reflexes
- Pressure over muscle insertions
- Active and static prolonged stretch
- Ortho-kinetics

**Inhibition of reflexes**

- A reflex consists of a motor act that is elicited by some specific sensory input
- Primitive reflexes appear at birth and become integrated once more complicated movements emerge
- When the CNS is damaged, primitive reflexes reemerge and again dominate motor activity

**Pressure over muscle insertion**

- Farber reported in 1974 that continuous firm pressure at point of insertion reduces tone

**Active and static prolonged stretch**

- Decrease reflex tone by providing mechanical stabilization of the joint and altering properties of the muscle spindle

**Ortho-kinetics**

- Originally developed in 1927 by Julius Fuchs, an orthopedic surgeon
- Focuses on physical effects to materials placed over muscle bellies
- Passive field materials (those that are cool, rigid, and smooth) produce inhibitory effect
- Active field materials (those, warm, expansive, and textured) produce facilitatory effect

**Examples of Upper Extremity Tone-Reduction Orthoses are**

A. Anti-spasticity "Ball" Splint.

B. Hand Cone Splint.

- They can be either hand-based wrist-hand orthoses (e.g., hand-cone splints) or forearm- based wrist-hand-finger orthoses (e.g., anti-spasticity ball splints and Snook splints).
- They can be volar based, dorsal based, or circumferential
- Typically worn two hours on and two hours off throughout the day
- Forearm based splints usually are more effective because of the extension positioning of the extrinsic finger flexors

Rationales of efficacy of tone-reduction orthoses include:

**• Reflex-inhibiting positioning-NDT**

technique approach (Bobath)

**• Firm pressure into volar surface (PALM)-**

Rood (sensorimotor) approach

- **Dorsal-based splints (eg, Snook):** facilitation of muscle contraction by direct contact-it is theorized that stimulation of extensor surface might produce extensor muscle contraction and balance muscle tone and / or avoid increase flexor tone.
- **Functions:** flexor tone reduction, prevent skin breakdown / maceration of palm by fingernails, increase passive range of motion via low-load, prolonged stretch (serial static splinting)
- **Indications:** spasticity-upper motor neuron lesions (cerebral vascular accident, HI, multiple sclerosis, cerebral palsy)

**Evaluation**

The therapist must understand the complex and intricate interrelations of normal

anatomic structures, their kinesio- logic functions and their biomechanical and physiologic ramifications before attempting to interpret, define and treat the abnormalities that accompany upper extremity problems.

Prior to evaluation, gathering patient history is important and can be used to establish a rapport with the patient and the family members. Information that are required from the patient, caretakers and health care professionals include details / cause of initial injury, previous medical care, reason for seeking additional care and desired outcomes of new treatments based on the wide range of specific functions a patient performs daily.

The evaluation of upper extremities requires inputs regarding strength, ROM, condition of soft tissues and sensation. In addition, ambulatory status, bilateral or unilateral condition, status of vision, and condition of the spine and head need to be considered before deciding on the appropriate orthotic device for the patient.

Since minor changes in strength dramatically change the orthotic need, many critical muscle tests must be made in patients with the upper extremity dysfunction. A patient with unilateral involvement can be provided with a typical prefabricated positional wrist hand orthosis (WHO) to prevent contracture and injury and a supporting shoulder orthosis for shoulder subluxation. On the other hand for a patient with bilateral involvement, conservation for grooming, feeding mobility, etc must be considered and the patient may be needed to be fitted with a more complex and customized orthoses.

There are similarities in the orthotic management of orthopedic and neurologically impaired patients. However, the neurologically impaired patients additional factors that challenge prescription criteria for the rehabilitation team. Lack of proprioception, sensation, (hyper or hyposensitivity) and spasticity represent some of these special considerations and possible problems with communication add to these patient management complications.

Evaluation of the neurologically impaired patient must be comprehensive. The therapist must assess the client or family motivation, client ability to tolerate and or function with the orthosis and risks associated with orthotic intervention. Further a total evaluation of the patient environment is important in developing the patient plan.

The medical diagnosis should alert the evaluator to the patterns associated with identified impairments and should be used to confirm potential findings. In addition to ROM evaluations, muscle power testing, assessment of sensation, skin sensitivity or lack, integrity of the affected limb or spine the therapist must assess what limitations initiating the orthotic care may have on other important functions.

**Goals of Orthotic Intervention**

The clinical experience of the therapist and patient evaluation must be used to create a plan of treatment. Only a well thought out plan that is thoroughly communicated to all the participants can ensure success of the intervention. Several other key factors that have an impact on the success of intervention are:

- Need to address the major complaint of the patient.
- Need to increase function without complication or patient risk.
- Need to establish a baseline of function so that results of intervention are measurable.
- Ability of the patient to independently donn and doff the orthotic device.

In some cases, concentrated instructions,

orthotic modifications and time are required before improved function can be observed. It is essential that the orthotic intervention be as simple as possible and to ensure that the least amount will help achieve the goals of intervention. The treatment goals need to be realistic, manageable and well discussed with the patient and care takers to be successful.

**Operation**

Although there is a wide variety of upper limb orthoses, most of these devices operate on similar principles. The general goal of most orthoses is to provide stability and support while allowing as much motion as possible. Immobilizing joints for long periods has proven deleterious for most patients since muscles atrophy, joints stiffen, skin tightens, and the healing process is ultimately slowed. By allowing movement while restricting motion that would create stress on joints, muscles, or tendons, orthotic devices allow healing and preserve range of motion and function.

Exact fit is a key element for many upper limb orthoses. In order to work properly, the orthosis must hold the body part in an exact position since if the orthosis does not fit exactly, it may not work and may actually cause harm. This can become a problem in situations where the patient has experienced swelling and may require a new fitting for the orthosis once the swelling has resolved. Poor fit can also lead to discomfort and the development of pressure sores.

Dynamic orthoses usually operate with the aid of attached outriggers. These provide a place to attach rubber bands, springs, or other materials that assist motion. They also provide leverage and help to ensure that the joint stays in proper position during movement. These devices require exact fit, as well as adjustment to ensure that the device works properly.

Many upper limb orthoses require a period of training for the patient to learn how to use the device properly. This is especially true with devices that assist motion, because the patient must initiate the motion properly in order for the orthosis to work. Patients with a long history of paralysis or immobilization may require considerable time in order to learn how to use the device. Orthotic treatment must maximize what little muscle strength and ROM the patients have. Further, orthoses used during the day for functional purposes might have to be replaced with positional orthoses to preserve gain and to prevent decline.

It is very important to consider the patient's motivation and attitude toward the orthosis as part of the treatment plan. Since most upper limb orthoses are removable, patients can choose whether or not to use these devices. Patients may object to orthoses because of discomfort, unattractive appearance, or restrictiveness of the device. Health care professionals must work closely with the patient to ensure that the patient will accept the orthosis and use it properly.

Exercise: Splints and exercises must be carefully integrated to allow the patients to achieve their full rehabilitation potential. Splints are used to improve passive motion, substitute for weakened or lack of active motion and are infrequently fabricated to provide resistive exercise. Evaluation measures provide the guidelines from which splinting and exercise programs are coordinated. Because patients with identical diagnosis may respond differently to therapeutic interventions, it is insufficient and often detrimental to adhere to rigid pre determined protocols and the exercise and splinting programs will require constant alterations to keep pace with the changing requirements. Patients are generally instructed to repeat a splint wearing and exercise routine every two hourly through out the day. This involves wearing the splint for one hour and forty five minutes and then

removing the splint and exercising for 15 minutes. Conversely if stiffness is a problem, exercise may be prescribed more often. for e.g. every 45 minutes. Written instructions for splint wearing and exercise routine should be used to augment verbal instructions.

**The patients and the family should also be instructed regarding**

- Donning and doffing
- Wearability
- Precautions

Patients must be taught to monitor the status of their splinted extremities. The presence of pain, reddened areas, blisters, swelling, rashes, or other problems associated with wearing a splint should be noted immediately and the use of splint discontinued until the appropriate corrective measures have been taken to rectify the problem.

Patients should be cautioned about exposing their splints to warm or hot temperatures such as hot water heaters or stoves or leaving the splints in hot car interiors since thermoplastic splints may lose or alter their configurations in these conditions.

**Maintenance**

Many upper limb orthoses require little or no maintenance. This is especially true for static orthoses and for those intended for temporary use. The plastic shell can be wiped clean, and materials worn underneath the orthosis can be washed or replaced. The patient may need to be checked periodically to ensure that the orthosis fits. Dynamic orthoses may require adjustments and replacement of worn springs, rubber bands, and the like Splinting in stroke patients.

Within the literature there is conflicting evidence and opinions on whether to use splinting as a form of intervention for stroke patients. There is a lengthy debate on the theoretical basis for splinting within neurology with two conflicting theories of biomechanical and neurophysiological approaches (Copley and Kuipers, 1999). Biomechanical rationale argues that splinting is used to prevent and manage length-associated changes in muscles and connective tissues. The neurophysiological rationale recommends that splinting is used to inhibit reflexive contracture of the muscle. The decision on whether to splint must be made on sound clinical reasoning. There are a wide variety of materials and types of splints and all have different qualities. Splints should not be considered when there is active movement that would be restricted if a splint should be provided.

Lannin and Herbert (2003) found a lack of evidence for splinting following stroke. In 2007, Lannin et al. conducted a randomized control trial and found no evidence to support the provision of a splint to prevent contracture in the acute phase for spasticity as a method of prevention of contracture. Regular passive range of movement and stretching is recommended and it is important to provide teaching to the patient and carer to perform this programme. The type of splint chosen will vary depending on the ideal position, but may include cones or resting splints in volar, dorsal or mid prone.

**Reduction of pain**

Following a stroke, pain can occur in various joints although wrist pain is a common complaint among patients. This can be due to the wrist being in a prolonged flexed position due to spasticity or flaccidity, leading to overstretching and / or shortening of muscles. Provision of a volar resting splint may be beneficial to prevent further harm and provide support. This must be alongside regular range of movement.

**Maintaining joint alignment**

Within the early stages of low tone, the hand may lose the curvatures due to prolonged

resting in a flat position. A resting splint may be considered to maintain the hand's natural curves and prevent secondary complications developing.

**Functional aims of splinting in stroke patients**

- To improve grasp in functional activities.
- To increase range of movement to open hand easily to enable daily hygiene.
- To use the upper limb pain free within activities.

**Orthoses for SCI patients:** Though the evaluation and rationale for determining the appropriate splint per injury level are agreed upon, the time of providing the splint and whether permanent orthoses enhance long term functions is debated. However early splinting for positioning to prevent deformity has proven to be effective at all injury levels.

Clients with C1 to C4 tetraplegia require resting hand splints to assist with proper positioning and maintain the support of the wrist and web space

Clients with Tetraplegia at C5 level can be independent with communication, feeding and hygiene only with the assistance of an orthosis. They must have joint stability and support at the wrist the hand to perform these skills. Static splints such as wrist cockup, long opponence and resting hand splints can be given. A universal cuff can be provided which include a slot for a spoon, fork, comb, tooth brush or hair brush so that a patient can effectively feed or groom himself.

Clients with injury at the C6 level can use their wrist for a tenodesis grasp. Clients who are not strong enough to use their wrist for tenodesis may require splinting to support their wrist until they can use their wrist against gravity. Long opponens splint can be used to position the thumb for function but support the weak wrist. Once the wrist strengthens, the long opponens splint can be cut down to a hand based short opponens splint to maintain proper web space and thumb positioning and maximize tenodesis.

Clients with C8 to T1 injuries or clients who have incomplete injuries may experience "clawing". A metacarpo-phalangeal block splint to block the MCP joints and promote weak muscle function can be given. Finally educating the client on the splint wearing schedules, skin checks and splint care are very important to prevent skin breakdown.

Orthoses for patients with cerebral palsy: In children with cerebral palsy, orthotic intervention is used in combination with an active task oriented therapy program. The new and wide variety of thermoplastic materials available to fabricate customized orthotics provides many options for upper and lower extremities as well as for trunk. The least restrictive devices, soft splints can be constructed from webbing, neoprene, hook and loop fastener material and other substances. Soft splints do not limit dynamic mobility and sensory feedback as much as do thermoplastics. For children with moderate degree of spasticity stronger molded thermoplastic materials are necessary to provide stability. They are lighter in weight than the original metal and leather braces and are easily cleaned. Some of the low temperature plastics can be altered as growth or change occurs. OT have found that upper extremity casting which provides prolonged and gentle stretch to spastic or contracted muscles is an effective adjunct to therapeutic techniques. Results of upper extremity casting have been significant with increased strength, control, and spontaneous use of impaired arm as well as bilateral hand use during play and transitional movements. The casting program is always integrated into the therapy program with the same functional goals.

**Summary:**

Therapist's clinical expertise along with a sound understanding of orthotic principles, knowledge of multiple orthotic options available are vital to reach the desired outcome of the orthotic intervention. Since every patients' need for an orthotic intervention is unique, it is essential to develop an individualized splint wearing and exercise schedule with periodic reassessments. Coordination between individual team members such as Orthotist, therapist (occupational physical or hand therapist) patient and family (especially in case of children) is essential for successful orthotic intervention.

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# Chp. 26 Visual Rehabilitation

**Dr. Hema Biju, Dr. Khushboo Bhagwanani, Dr. Chandali Mehta**

Vision is the special sense by which the qualities of the environment such as color of a fruit, contours of a garden and facial features of a person are perceived through the physiological process wherein light rays entering the eye are transformed by the retina into electrical impulses that are transmitted to the brain. The brain then perceives the image and understands its characteristics and contexts. Vision is man's most highly developed sense. It was extremely important primitively to find food and resources, and to stay safe and away from dangers. Visual functioning is important for learning, using tools and devices, moving from one place to another and enjoying human interaction. Thus, vision is vital to everyday activities.

**Anatomy and Physiology of the Visual System**

The eye is an extremely complex organ comprising myriad structures that work together to provide the sense of sight. An understanding of the anatomy and physiology of the visual system provides eye care professionals and staff with a valuable tool when meeting their patients' needs and expectations.

- Embryology: Eyes develop in the fetus during the second week of pregnancy. The formation of the eye is primarily a product of different cell growth rates, as well as cellular specialization as development takes place. The optic cup predetermines the size and shape of the orbit. Postnatal growth of the orbit coincides with growth of the globe. Eyelids start out fused together and separate between the fifth and the seventh month. Most babies are hyperopic, or farsighted, at birth.
- Dimensions, shape, and position: The eyes are situated in a bony orbit whose functions are to provide protection, support, and attachment for the muscles that control eyeball movement. Eyelids are the flexible and mobile multilayer structures that cover the eye anteriorly. When the eyelids are closed, they cover the globe. The primary function of the lids is to protect the eye from bright lights and foreign objects. The blinking action also serves to protect the cornea from drying. The tear film is spread over the surface of the eye at the rate of five times per minute. Blinking also aids in the tear flow from the lacrimal glands and the aspiration of tears into the nasolacrimal system.
- The anterior 1/6th-inch of the surface of the eyeball, known as the window of the eye, is the clear, dome-shaped cornea. The remaining 5/6-inch is the white, opaque, fibrous sclera. The anterior segment is the front third of the eye and includes all structures in front of the vitreous: the cornea, iris, ciliary body and lens.
- The eye consists of three layers: the fibrous, vascular, and nerve layers, and additionally, the ocular media. The fibrous layer is the outer layer of the eye, consisting of two parts, the cornea and the sclera. Light enters through the cornea, the transparent outer covering of the eye. The eyeball is rounded, so the cornea acts as a lens and bends or refracts light. The fluid

beneath the cornea called the aqueous humor helps to shape the cornea and provides nourishment to the eye. The iris and the pupil assist with allowing light into the eye. Pupil is the aperture in the eye, the size of which is determined by the iris, the contractile ring that is associated with eye color as well. As the pupil dilates (gets bigger), more light enters the eye. The lens allows the eye to focus on either near or distant objects. Ciliary muscles surround the lens, relaxing to flatten it to image distant objects and contracting to thicken the lens to image close-up objects. The vitreous humor is a transparent watery gel that supports the eye and allows for this distance.

- The retina contains the cells that sense light (photoreceptors) and the blood vessels that nourish them. The most sensitive part of the retina is a small area called the macula, which has millions of tightly packed photoreceptors (the type called cones).There are 120 million rods and 6 million cones. Cones are densest in the macula, responsible for color and central vision, work best in bright light, capture detail and color, and require direct stimulation. Rods are more numerous in the periphery of the retina, function best in dim illumination, detect general form rather than details, and provide background information. Each photoreceptor is linked to a nerve fiber. The nerve fibers from the photoreceptors are bundled together to form the optic nerve. The optic disk, the first part of the optic nerve, is at the back of the eye.

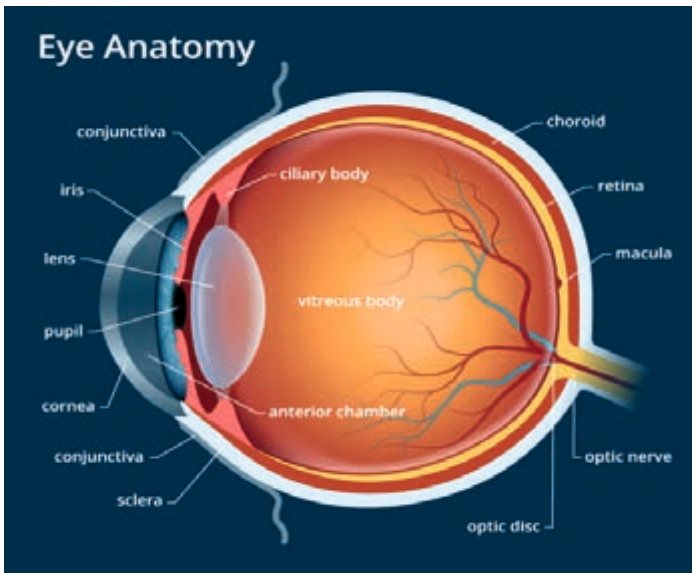



*Fig 1: Anatomy of eye*

- Functions: Visual acuity is the level of detail with which a person can see objects. Visual field is the total area one sees in a single view without turning the head or eyes where the central visual field includes the spot on which the person focuses and the area immediately surrounding the spot on which the person is focusing is called the peripheral field of vision. Contrast sensitivity is the capacity to distinguish between similar shades of light and dark. Light modulation is the capacity to regulate light control glare and adapt to changing light condition like light to dark and dark to light. Visual perception and interpretation are concerned with correctly processing and interpreting the information the brain receives from the eyes via the optical athway.

- Visual Pathway: Lee Ann Remington (2012) explains the visual pathway as follows: The visual pathway consists of the series of cells and synapses that carry visual information from the environment to the brain for processing. It includes the retina, optic nerve, optic chiasm, optic tract, lateral geniculate nucleus (LGN), optic radiations, and striate cortex. The first cell in the pathway are the photoreceptors that convert light energy into a neuronal signal that is passed to the bipolar cell and the amacrine cell and then to the ganglion cell. All these cells and synapses lie within the retina. The axons of the ganglion cells exit the retina via the optic nerve, with the nasal fibers from each eye crossing in the optic chiasm and terminating in the opposite side of the brain. The optic tract carries these fibers from the chiasm to the lateral geniculate nucleus (LGN), where the next synapse occurs. The fibers leave the LGN as the optic radiations that terminate in the visual cortex of the occipital lobe. From various points in this pathway, information about the visual environment is transferred to related neurologic centers and to visual association areas in the brain.

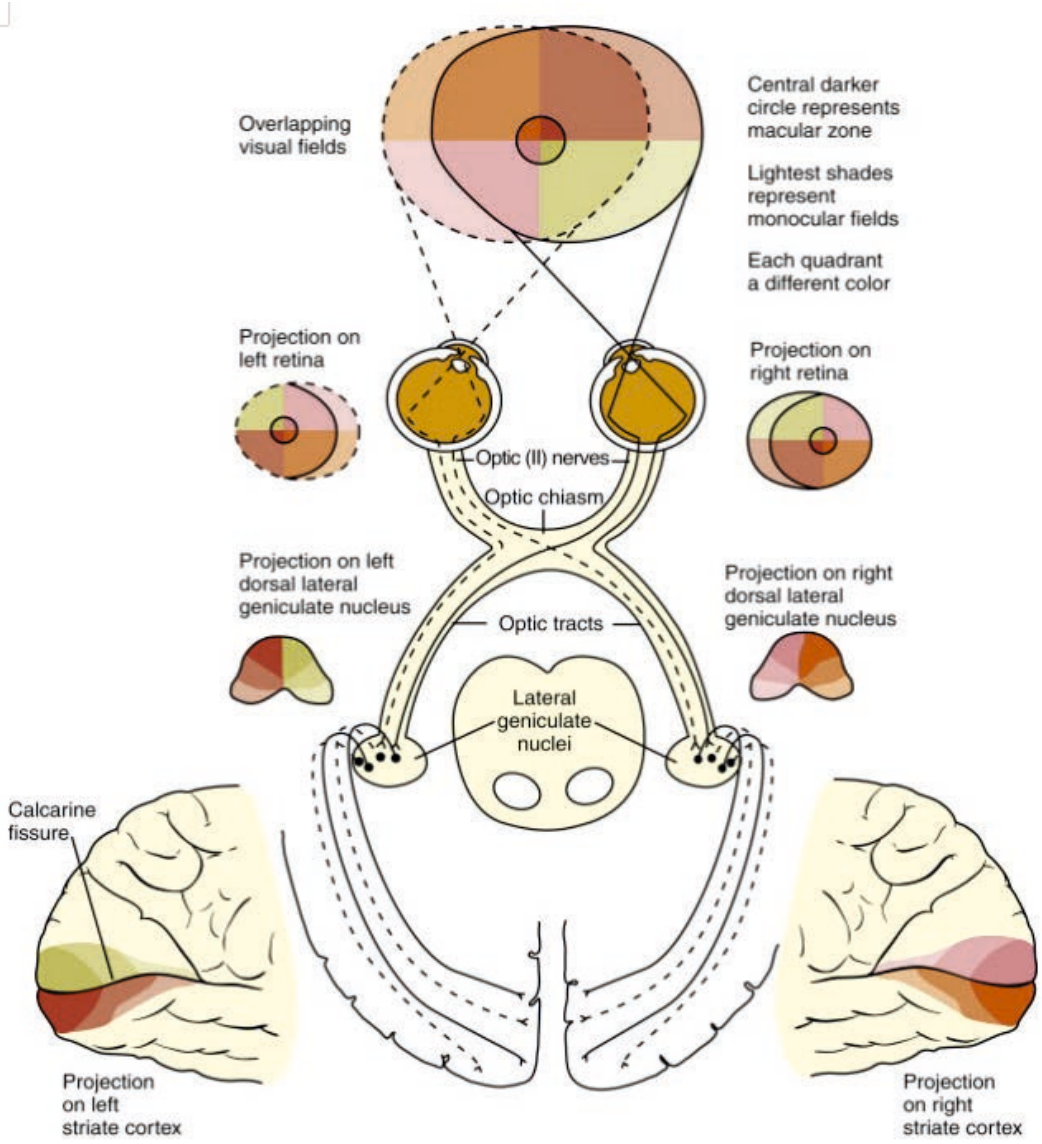



*Fig 2: Visual Pathway*

**Vision development:**

Child's vision development begins before birth. It takes several months for your child's vision to develop fully after birth. In the first week of life, babies don't see much detail. Their first view of the world is indistinct and only in shades of gray.

| | |
|---|---|
| Birth | • Corneal reflex present<br>• Eyes are sometimes uncoordinated, may look crossed-eyed<br>• Able to stare at object if held 8 to 10 inches away<br>• Initially fixes eyes on a face or light then begins to follow a moving object |
| 1 month | • Looks at faces and pictures with contrasting black and white images<br>• Can follow an object up to 90 degrees<br>• Watches parent closely<br>• Tears begin to work |
| 2 to 3 months | • Begins to be able to see an object as one image<br>• Looks at hands<br>• Follows light, faces, and objects |
| 3 to 5 months | • Will look at self in mirror<br>• Will look at own hand |
| 5 to 7 months | • Has full color vision, able to see at longer distances<br>• Can pick up a toy that is dropped<br>• Will turn head to see an object<br>• Likes certain colors<br>• Will touch image of self in mirror |
| 7 to 11 months | • Can stare at small objects<br>• Begins to have depth perception<br>• Plays peek-a-boo |
| 11 to 12 months | • Can watch objects that are moving fast |
| 12 to 14 months | • Able to place shapes in proper holes<br>• Becomes interested in pictures<br>• Points and gestures for objects and actions<br>• Recognizes own face in mirror |
| 18 to 24 months | • Able to focus on objects near and far<br>• Scribbles with crayon or pencil<br>• Can point to body parts (nose, hair, and eyes) when asked |
| 36 to 38 months | • Can copy shapes<br>• Vision is nearing 20/20<br>• Able to identify colors |
| 48 to 72 months (4 to 6 years) | • Recognizes and recites the alphabet<br>• Ready to begin reading<br>• depth perception |

**Types of Visual Problems**

Visual impairment is defined by the World Health Organisation (WHO) as vision of <3/60, that is being unable to see at 3 m what someone with typical vision can see at 60 m within a <5° field.

There are many conditions that can affect part of the eye ranging from temporary issues like conjunctivitis to permanent damage to optic nerve from multiple sclerosis. Depending on the structure affected, visual functioning is compromised leading to temporary or permanent visual loss. Functional visual loss can result from congenital deficits like amblyopia, acquired injuries like stabbing, systemic disorders like diabetic retinopathy or age related conditions like macular degeneration. However any visual problems that can impair a person's ability to observe and perceive the surrounding limit successful completion of everyday tasks, work or leisure activities fall under the purview of the occupational therapist. Therapists may often see clients with primarily physical impairments such as hip and knee injuries or stroke but having co-morbidities related to vision that affect their functioning. Hence it is important for therapists to understand the visual system as well as interventions that can help individuals overcome or compensate for the visual deficits.

Visual deficits are mainly explained by terms like 'Low vision' or 'Blindness'. However their differences are not necessarily always clear. Blindness is the inability to see light (no light perception) or to form or use functional vision for any activities. The National Eye Institute (NEI) defines Low Vision as a condition that cannot be corrected by medical or surgical intervention or by eye glasses; it allows some usable vision but is severe enough to interfere in completing daily activities. The US Government coined the term legal blindness in the 1930s to enlist criteria that qualify individuals for benefits and services- a legally blind person has best corrected visual acuity of 20/200 in the better eye or a visual field of 20 degrees or less in the better eye. WHO has divided visual capabilities into 7 categories based on visual acuity. The international Council of Ophthalmology 2002 recommended that the term low vision be used when the person can use magnification or other vision enhancement devices to complete activities and that blindness be referred for use only when the person has complete vision loss and must rely on vision substitution.

| Sr. No. | Structures | Functions | Injury | Testing | Treatment |
|---|---|---|---|---|---|
| 1 | Bony Orbit | Houses and Protects: Eyeball, Optic nerve, Extraocular muscles, Other nerves, Blood vessels, Lacrimal gland, Fat, Connective tissue | Orbital blow out fracture | | Surgery |

| | | | | | |
|---|---|---|---|---|---|
| 2 | Eyelids and Eyelashes | • Protect eye from foreign bodies<br>• Help limit light into the eye<br>• Eyelids also function as part of the lacrimal system<br>• Blinking squeezes tears which fill in uneven surfaces of cornea and nourishes and protect it | Entropion (inward turned eyelids)<br>Ectropion<br>Ptosis<br><br>Trichotillomania<br>Atrichia (congenital absence of hair)<br>Trichiasis (inward growth of eye lashes) | Observation | Surgery |
| 3 | Eyeball 3a) Outer layer: Conjunctiva | • Thin transparent membrane covering sclera and inner eyelid<br>• Provides protection and moisture<br>• Many blood vessels<br>• Few pain fibers | Conjunctivitis | | Eye drops |
| | Sclera/<br><br><br><br>Cornea | • Tough opaque white elastic fibrous layer<br>• Encloses eyeball except for cornea<br>• Extension of the dura mater of CNS<br>• Protects inner contents of eye<br>• Helps maintain shape of the eye<br>• Extra ocular muscles attach to its surface<br>• Three layer: epithelium, stroma, endothelium<br>• Refracts light and helps focus on fovea | Scleritis | | Eye drops |

| | | | | | |
|---|---|---|---|---|---|
| | 3b) Middle layer:<br>Uveal tract: Choroid, ciliary body and iris<br><br>Aqueous/ lens/ Vitreous | • Choroid: vascular supply<br>• Ciliary body: ciliary muscle controls the lens and the ciliary process secretes aqueous<br>• Iris: controls the pupil (dilator/constrictor muscle) and protects retina from UV light<br>• Reduces scatter of light<br><br>Maintains shape and pressure of eye/ helps refract appropriately/ balloons the retina | Uveitis<br><br>Presbyopia<br>Cataracts (congenital, youth-heredity, injury, topical steroids, age) | | Steroids: drops (iris only), injections and oral meds<br><br>Intra Ocular Lens Replacement |
| | 3c) Inner layer:<br>Retina | | (H)<br>Diabetic retinopathy (increases incidence of glaucoma/ cataracts)<br>BDR: fluid, hemorrhage<br>PDR: neovascualarization<br>Traction retinal detachment | Myopia<br>Anisometropia (diff nearsight in 2 eyes) | Prevention: Nutrition, Exercise, Smoking, maintenance & Mx of diabetes: check and insulin admin<br>LASER by photocoagulation<br>Injection: Vitreous steroids, anti-VEGF<br>Intraocular Sx: vitrectomy, laser |

| | | | | | |
|---|---|---|---|---|---|
| | | | Retinitis Pigmentosa (night blindness, hereditary)<br><br>Retinal degeneration: Best's AD, Stargardt's AR but AD | HIV, Virus, Vision<br><br>Opthalmoscope<br>Fluroscein angio<br>OCT<br><br>Mainly central but may cause both or only peripheral | Cryotherapy<br>Laser<br>Full term babies<br><br>Anti-retroviral however toxic<br><br>Anti-VEGF: eliminate existing vessel and block new ones, need repeats<br>Hot laser, photodynamic therapy/ cool laser<br>Wet; Macular translocation surgery<br><br>AREDS: (25000 IU beta carotenes for non-smokers, 500 mg Vit C, 400 mg Vit E, 80 mg Zn, 2 mg Cu)<br>Prevention: no smoking, sunglasses (blue), green leafy veggies, omega 3, no packaged foods, trans fat & maintain ex/BP. |

| | | | | | |
|---|---|---|---|---|---|
| 4 | Optic Nerve | | (AA)Glaucoma: closed angle- access blocked, traumatic-damaged, open angle-not draining Optic neuropathy, MS | Eye Check: intraocular pressure, field of vision, optic nerve health | Not preventive: drug regimens color coded by patient Trabeculectomy (bleb), trabeculoplasty |
| 5 | Optic Pathway | Optic Chiasm Optic tract Lateral geniculate nucleus Superior colliculus | Pituitary tumors Infection MS | Eye check, perimetry, CT/MRI | Neuro-surgical/ pharmacological |
| 6 | Brain | Calcerine fissure | Head injury/Stroke Infection | Eye check, perimetry, CT/MRI | Neuro-surgical/ pharmacological |

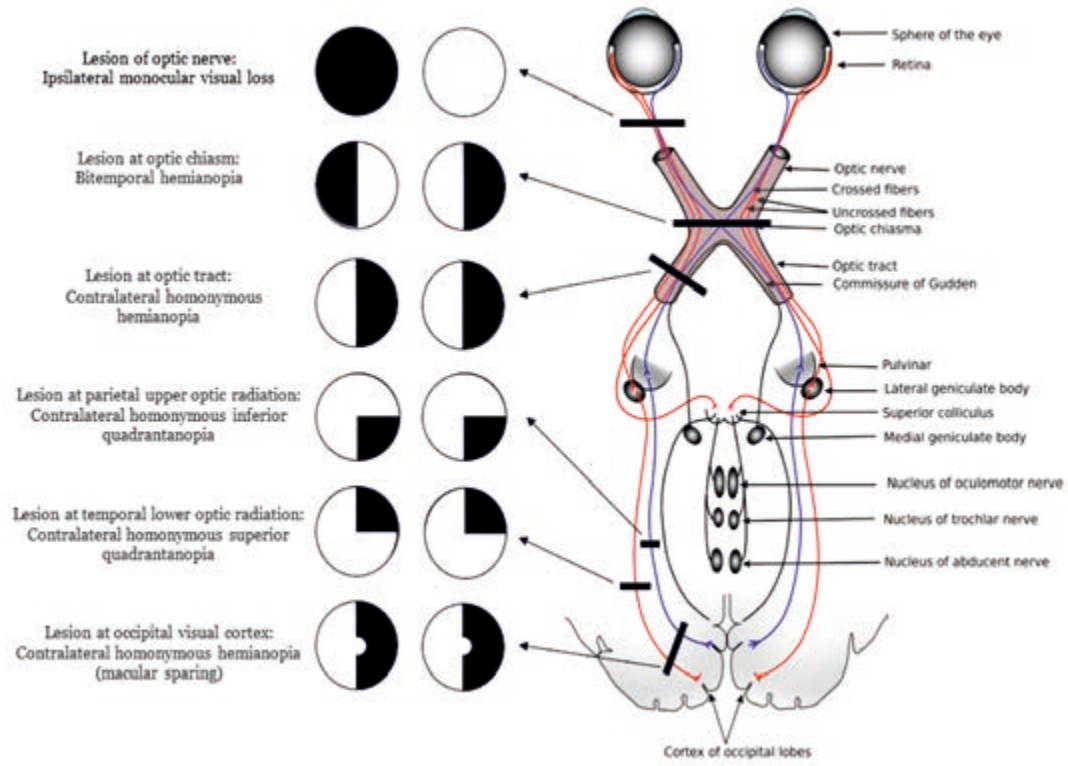



*Fig 3: Field deficits post disruption of visual pathway*

**THEORIES**

The two main theories in visual perception are that of psychologist Richard Gregory who argued that perception is a constructive process which involves a top-down approach were information from past experiences and stored knowledge is used to make inferences about what we visualize. Gregory estimated that about 90% of information that reaches the eye is lost on its way to the brain. Constructivists like Gregory use the example of form constancy to support their theories i.e. we can judge the size of an object like a tree even if the image of the object on the retina is very small (the tree is far away) or very large (the tree is very close). However other researchers countered that newborn infants often showed form constancy and preferred normal features to scrambled features even as soon as they were born. One such researcher was Gibson who contested that perception is direct and not subject to any hypothesis testing by the individual consciously or unconsciously. He proposed the bottom-up process where the sensory information is solely analyzed based on the environmental cues such as optical arrays, relative brightness, texture gradients etc. However Gibson's theory could not explain illusions such as staring at a moving object and then transferring the gaze to a stationary object gives the stationary object an illusion of moving.

The field of visual perception was also influenced by other theories which supported either the top-down or the bottom-up approach to understanding visual processing.

1. Gestalt theory: The word gestalt means configuration in German. This theory emerged in the early 1900s out of psychology and Ehrenfels, Wetheimer, Koffka and Kohler have made noteworthy contributions to it. Gestalt explained the brains tendency to group things- why some images belong and others don't. Dr. Graham explains the gestalt as it refers to structure, configuration or layout that is unified and has specific properties greater than the simple sum of the individual parts. For example, a person reading a text perceives each word first as a complete word and its meaning rather than seeing individual letter forms. Similarly each frame in the movie may be considered separately however its rapid projection across times forms the perception of movement and the narration continues.
2. Brunswick probabilistic functionalism- The Theory of Probabilistic Functionalism (TPF) was developed for visual perception and certain cognitive processes by Hungarian born US psychologist Egon Brunswik. According to TPF perception involves the selection of environmental cues that are most useful or functioning in responding which means individuals pick up those visual cues that are conducive for them to create a response hence the validity of all perception and beliefs being necessary probabilistic rather than certain. For example, when we see a flower bouquet with our eyes the elements from the visual system are combined with remembered information from the viewer's personal history, motives and other sensations.
3. Cognitive constructivism- According to this theory perceptual processing involves inductive or intelligent problem solving. Under this theory similar to the probabilistic functionalism the perceptual processing is assumed to be unconscious, similar to those involved with problem solving but are dependent the features of the environment observed as well as previous general and context-specific knowledge. This theory explained the cognitive processes

involved in understanding of visual information and reiterated the influence of higher centers and cognitive processes in visual perception.

4. Modified information processing theory- Under the information processing theory the first or input stage of processing is a affected by the integrity of the optical system or eye health as well as the intact functioning of the accommodative and convergence systems, adequate occulomotor control as well as corrected refractive errors as these primary systems are important for progressing the visual information to visual cortex accurately for interpretation. Occular motor skills dependent on feedback from areas that monitor head and body orientation and movement as well as those areas that monitor feedback from the ocular motor driver at the primary visual cortex. This is how binocular vision occurs and visual input is processed as color, contour, contrast, and depth. During the second or integrative stage of processing, sensory and proprioceptive information are mixed. This blending or combination of information results in a concept or plan that will serve as a guide to an action or response organization of space and motion. Form perception, object recognition and visual awareness are developed at this stage. Visual integration is dependent on intact communication within visual processing areas and pathways between these processing areas as well as intact reception in addition pathways mediating other sensory and motor areas.

5. Mary Warrens hierarchical model

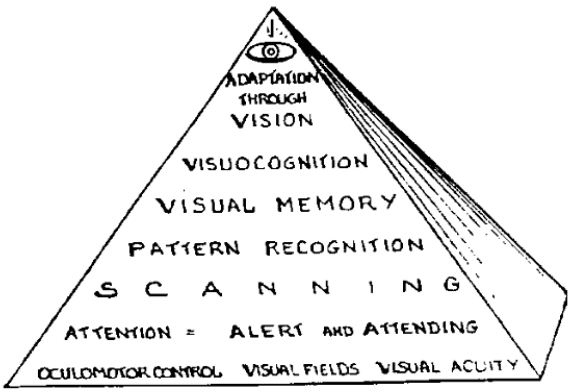



*Fig 4. Mary Warrens hierarchical model*

**Occupational therapy management in Visual deficits**

Occupational therapists use the Warren model for understanding and treating visual dysfunctions as it tends to be based on the Occupational Therapy Practice Framework (OTPF) and assists in evaluating and remediating visual dysfunction using functional activities to achieve functional goals. She explains visual functioning as a hierarchy of skills where the higher skills are disrupted when the basic visual functions such as visual fields and occulomotor control are affected. Warren used a bottoms-up approach for individuals with brain injuries by remediating lower level skills in order to achieve integration of higher level visual-perceptual functioning. She also created an evaluation tool called the biVABA (which provides therapists a framework to assess and treat individuals with visual processing disorders.

**Intervention planning based on functional impairments**

1. Reduced Visual Acuity

Occupational therapists do not issues glasses or corrective lenses. Therefore, it is important to refer the patient to appropriate eye care specialist (ophthalmologist/ optometrist) in order to determine if visual acuity can be restored with corrective lenses. If vision cannot be restored and acuity less than 20/100, then the OT goals focus on improved independence using compensatory strategies. Characteristics of environment and task that need to be considered are as follows:

1) Illumination: increase illumination without increasing glare using halogen, fluorescent, full spectrum lighting.

2) Enlargement: Use enlarged font for calendars, medication bottles, on computers and phones

3) Background contrast: change background color to differ from object, provide markers such as bright tape for step or curb, changes in surface

4) Background pattern: Preferably remove throw rugs and small carpets, eliminate pattern designs, use solid colors (rugs, bedspreads, carpet, and placemats)

5) Organization and Decluttering: Establish routines that will reduce the number of steps especially those that involve work with small items such as use pre-cut vegetables instead of cutting, organize living space by eliminating multiples and arranging the environment so that only essential items are placed where needed (closets, drawers, shelves, countertops, fridge).

6) Resources and community services: Use assistance of family/friends, paid help and/or community resources when able. Be aware of organizations and services available in your locality addressing needs of individuals with low vision.

2. Reduced Contrast Sensitivity

Therapists cannot improve contrast sensitivity function however strategies to be used are similar to those used to improve acuity with emphasis on using stark contrasts such as milk in black cup and coffee in a white cup, tactile compensatory strategies such as use of rubber bands on sugar or salt containers to distinguish, bright fluorescent tapes for safely managing stairs and curbs, improved illumination using stick on lights inside closets and cabinets.

3. Visual Field Deficit

Visual field deficits affect the ability of the individual to visualize the entire surrounding and can be a safety concern. Clients often will run into objects or miss them when working on a task, if they fall into the field with the deficit.

**General search strategy:**

Compensation of a visual field deficit requires a conscious, cognitive strategy of using head movement to broaden the visual field for example, if the person has a left visual field deficit ask him to look at his left shoulder and then straight up, this is where scanning for the environment should begin.

Educating the patient about the importance of being cautious is also important as the CNS performs perceptual completion and hence the visual input on the affected side cannot always be trusted. In order to improve the effectiveness of the search pattern, client will have to focus on speed (slow) and scope (method) of search pattern. It is very important to initiate of a wide head turn towards the blind side, increase in the number of head and eye movements toward the blind field, execute an organized and efficient search pattern that begins on the blind side, attend to and detect visual field detail on the blind side, and check repeatedly

until it becomes a habit.

The therapist can practice these strategies in addition to work on the client's ability to quickly shift attention and search between the central and peripheral visual field on the blind side and foresee possible visual cues on the blind side using activities such as ball pass, balloon volley, using a laser pointer on a white wall in multiple locations, using 'post it' notes with letters or numbers widely scattered on a wall and the Dynavision 2000.

The client needs to practice different types of search strategies such as linear, circular, in both static and dynamic environments such as finding clothes in closet, locating items while walking in a crowded area, locating items in rooms or cabinets, playing games such as solitaire and checkers, search and match games, etc.

Mobility:

Therapists also focus on functional mobility by practicing these search strategies in dynamic and unfamiliar environments. The client is educated to be observant and predictive of obstacles in the environment (curbs, curb cuts, changes in surface), use objects/features in the environment as landmarks (picture on the wall, railing of the staircase, etc.) and also count steps or distances when needed. Search tasks, scanning tasks and 'I spy' games during mobility in different environments can be practiced. To make things more difficult attention to moving objects in the blind side like the patient is asked to relay the color or number plate of a moving vehicle that passes on the affected side can be worked on.

**Reading and writing:**

Patients usually have difficulty locating and maintaining the correct line of print we reading and accurately identifying words and numbers since they may omit the beginning of the words and numbers. Patients may also have difficulty locating the left margin of reading material and often lose place in a busy book or with small print.

Therapist can have the client anchor their vision to the left margin of the reading or writing material using a red line down left margin or a rubber band at the left end of the book that may also serve as a page marker or any other visual cue (used on right for right visual field deficit to indicate end of line). A card or ruler can be used to move from line to line. Table top activities such as Sudoku, crosswords, jumble, word and number searches, pre-reading and pre-writing exercises (e.g. Warren or LUV reading series) can be used.

To ensure the client stays on the line while writing they should monitor pen tip as hand moves across the page and use tasks such as tracing, complete blank checks or diaries. Templates for writing or checks are available for use as a tactile cue if needed

**Compensatory strategies:**

It is important to organize and declutter work space for office or school tasks. Reduce 'visual noise', eliminate and simplify using solid colors (eliminate patterns), add color and contrast to key structures needed for orientation or for things that need to stand out, contrast in writing material (black pen and bold lined paper) assists in patient being able to monitor handwriting and increase illumination assists with reading and finding subject matter easily. The client needs to be vigilant and methodical to safeguard against perceptual completion for e.g. with financial figures or important documents.

**4. Oculomotor Dysfunction**

The aim of occupational therapy is to reduce or eliminate oculomotor dysfunction which may reduce the speed and precision of eye movements resulting in slower and less accurate visual processing. Dysfunctions like diplopia cause perceptual distortion which may also affect postural control and cause

visual stress. Occasionally extreme fatigue and agitation may arise. Interventions for occulomotor dysfunction include: Occlusion to eliminate diplopia, eye exercises, application of prism and surgery. Occupational therapists address only occlusion and implementation of an eye exercise program (This is not allowed in the US as the eye exercise program is directed by an optometrist). Ophthalmologists may address the issue with prism and/or surgery and management of everyday tasks with the use of prisms or post-surgery falls under the occupational therapy domain.

Diplopia must be eliminated in order for the patient to fully benefit from therapy. Two types of occlusion are undertaken:

- Complete occlusion using an opaque tape, eye patch, or an eye occluder. This method is difficult to tolerate for long periods of time. It is recommended that the eye patch or occluder be alternated to prevent eye muscle strain. This method is cheap and easy to apply however it eliminates peripheral visual input, disrupting normal CNS mechanisms for control of balance and orientation to space and often causes the patient to feel off balance and disoriented. Therefore is has poor compliance, especially if the affected eye is the dominant eye.
- Partial Occlusion using a strip of opaque tape applied to lens to block visual stimulation of central field. The peripheral field is not occluded which means only the nasal portion of the lens of the non-dominant eye is taped. If patient does not regularly wear glasses non-prescription glasses can be used. This method is more comfortable hence has better compliance.

Daily range of motion exercises

Patient covers the unaffected eye and moves the affected eye towards the direction of the paresis and then repeats the range of motion exercises using both eyes together in all cardinal directions of gaze. Performed 4 to 5 minutes several times per day

5. Visual Inattention

Visual inattention hampers the ability to learn and memorize visual information in the surroundings. The therapist has to educate the patient and provide compensatory strategies to re-organize the visual search pattern. Patients are asked to adopt3 strategies:

- Left to right linear: for reading and inspection of small visual detail, for structured personal space
- Circular (clockwise or counterclockwise): for viewing unstructured and extra personal space
- Scanning IN from end range rather than scanning OUT

These strategies can be practiced using therapeutic activities like card match (ensure left to right, top to bottom search pattern), solitaire, dominoes, checkers, large puzzles (100-500 pieces), enlarged word or number searches, etc.

Examples of therapeutic activities for the occupational therapist to use for specific visual deficits:

1. Vision tracking: letter cancellation activities using paragraphs or letters (cancel Q)
2. Vision and body movements: use vision target at eye level in front of patient and a 6 feet line of masking tape on floor. Have patient walk forward and backward on tape while maintaining fixation on target
3. Vision directions: focusing on left, right, up and down using words and arrows. Therapist holds word or arrow card and patient verbalizes direction and moves (B)UEs in the correct direction

4. Vision ball catching: Throwing and catching the ball underhanded and then over handed with (B) hands, (R) hand only, (L) hand only
5. Vision and hand coordination: tape 'x' target at eye level, stand 12 inches away. Patient's hands should make circles with hands moving clockwise (simultaneously), counterclockwise (simultaneously), then opposite directions (simultaneously) while the eyes stay on target.
6. Hand and vision ball: Have a small ball hanging from ceiling at waist level. Hold dowel horizontally with (B) hands, gently bump ball. Upgrade task by having 3 pieces of tape wrapped around dowel. Bump ball with each piece of tape
7. Vision ball: Have a small ball hanging from ceiling (eye level) with letters or number on it. Gently swing ball, having patient focus on letter or number using eyes ONLY
8. Vision shapes: Tracing shapes circle, square, rectangle, diamond then identifying them at a distance and tracing using arms while focusing on the shape
9. Vision lines: Following the curvy line from the letter to the number using eyes only
10. Vision movements: The patient focuses on a target in front of him and two other targets are placed in the visual field. Patient is asked to look at them individually and then back at the central target using eyes only
11. Vision accuracy: Having the patient read alternate letters placed at eye level in front of patient and on table in front of patient
12. Vision copying: Having the patient create a design using shapes and have patient replicate design
13. Vision recognition: matching sequences of 3 letters or numbers or shapes in exact sequence
14. Vision memory: show sequences of 3 letters or numbers or objects for 3 seconds. Have patient recall 3 numbers (verbally or by writing)

**Evaluation of Visual Functions**

Basic Visual Functions

Oculomotor Functions

| | | | |
|---|---|---|---|
| Ocular alignment | C/o of diplopia<br>Head turning<br>Vertigo, motion sickness | Hirschberg technique<br>Scheiman Alignment screening | Referral to optometrist<br>Eye exercises<br>Occlusion<br>Activities to obtain fusion |
| Visual fields | Running into objects<br>Inability to see steps<br>Inability to see signage<br>Difficulty reading, writing | Confrontation test<br>Damato campimeter<br>Humprey visual field analyzer<br>Kinetic two point test<br>Tangent screen<br>SLO<br>Amsler grid testing | Training in yoked prisms or mirrors<br>Visual retraining: restorative approach (e.g. search strategies) & adaptive approach (environmental changes) |

| | | | |
|---|---|---|---|
| Oculomotor control | Visual fixation<br><br>Saccadic eye movements<br><br><br><br>Smooth pursuits<br><br><br>Organized scanning<br><br><br><br>Convergence and accommodation | Fixation test<br><br>Northeastern State University College of Optometry Developmental eye movement test<br>Pepper visual skills for reading test<br>Direction of gaze<br>Pursuits : direct observation method<br>Scanboard test<br>Scan course<br>Warren visual search subtests<br>Dynamic object search test<br>Scheiman: aptitude of accommodation<br>Padula eval of convergence<br>Near point convergence<br>biVABA | Fixation with paper pencil task<br>Fixation with ambulation task<br>Remedial- encourage saccades<br>Adaptive- anchoring, decrease density<br><br><br>Remedial- encourage smooth pursuits<br>Adaptive- anchoring, decrease density<br><br>Remedial- encourage scanning<br>Adaptive- anchoring, decrease density<br><br><br>Warm up exercises- lemon on spoon |
| Eye movements | Strabismus | Eye movements in different planes | Refer to optometry<br>Exercises and training in use of prisms |
| Diplopia | C/o double vision<br>Squinting or head position to focus | Cover/Uncover<br>Alternate cover/uncover | Prisms, partial occlusion, full occlusion, vision training exercises |
| Visual inattention | | Behavioral inattention test<br>Conventional test-<br>Behavioral test-<br>Alternating simultaneous stimuli<br>BiVABA<br>Dynamic object search | |

**Physiotherapy management in Visual deficits**

Children with cerebral palsy will experience vision problems, but it is recommended that all children be screened so that interventions can be put into place as early as possible; impaired vision can be roadblock that interferes with developmental growth. Approximately 50% of children with cerebral palsy have some form of visual impairment. The severity of vision loss tends to increase with the severity of cerebral palsy, as measured by the GMFCS score, and with the number of limbs affected. In general, visual impairments are thought to be more common in children with spastic cerebral palsy than those with athetoid or ataxic types.

Individuals with ASD see and process the world differently, Autistic people may also have problems coordinating their central and peripheral vision. For example, when asked to follow an object with their eyes, they usually do not look directly at the object. Instead, they will look to the side of the object. Eye movement disorders like strabismus and crossed eyes are common in older autistic children (7–19 years).Many autistic people are visually defensive. Visually defensive persons avoid contact with specific visual input and might have hypersensitive vision. They have difficulty with visually holding still and frequently rely on a constant scanning of visual information in an attempt to gain meaning. Often, the signs of these vision problems can be masked by the behaviors that autistic individuals use to cope with the sensory overload of the world around them.

**Types of Vision Impairment Associated with Cerebral Palsy**

- Visual acuity is the acuteness, or the clearness, or a person's vision. Abnormal condition is commonly referred to as nearsightedness or farsightedness. Nearsightedness (myopia) is a common vision condition in which you can see objects near to you clearly, but objects farther away are blurry. It occurs when the shape of your eye causes light rays to bend (refract) incorrectly, focusing images in front of your retina instead of on your retina. Farsightedness (Hypermetropia), the person can see far objects clearly, but cannot see near objects clearly. presbyopia (a condition of the lens that can cause blur vision); astigmatism (an inability see objects that are very close or far away); and refractive errors (insufficient or misplaced light in the retina).
- This is the most common set of vision issues, and the ones most likely to be corrected by wearing eyeglasses or contact lenses. Glasses are very important for a child who has CP and acuity loss. Glasses can greatly improve incidental learning or learning that takes place by observation.
- Field loss – If a person cannot see 180 degrees in either direction, he or she may have field loss. Different types of field loss include hemianopia (loss to the right or left, or upper or lower field); central loss (a spot in the center of one's field of view); and peripheral loss (sight loss at the edges of the vision field). scotomas (isolated spots are missing from your vision) islands of vision: (you can see only scattered spots)
- Oculomotor concerns – These are conditions that affect the movement of the eye, as well as depth perception. Types of oculomotor conditions include esotropia (eye turns inward); exotopia (eye turns outward); hypertropia (eye turns upward); hypotropia (eye turns downward); strabismus (eye misalignment); and amblyopia (a lazy eye).
- Cortical Visual Impairment – This is a

condition that exists in the brain, not in the eyes. Sometimes called cortical blindness, many children can see but can't interpret images.




*Fig 5:Hyperopia*



*Fig.6:Strasbismus*

**Physical signs of visual impairment in cerebral palsy include:**

- Complaints of frequent headaches
- Covering an eye while reading
- Messy Handwriting
- Holding reading materials or picture books very close
- Moving his or her head – versus the eyes – back and forth to read
- Poor hand-eye coordination
- Squinting while focusing on objects
- Turning his or her head to use one eye
- Underdeveloped speech and vocabulary
- Rubs eyes frequently
- Able to read for only a short time
- Poor reading comprehension
- Trouble visualizing what is read- Poor reading comprehension
- Trouble with mathematical concepts
- Poor recall of visually presented material
- Trouble learning right and left
- Reverses letters and words

The most common vision problems in children with cerebral palsy are strabismus (which makes up 55.7 percent of vision problems associated with CP). All children and young adults with CP, regardless of their neurologic status, should be considered at risk of significant refractive errors which, if uncorrected, may exaggerate present condition. Furthermore, those with non spastic types of CP demonstrate the most extreme errors of refraction.

Studies describing behaviour in ASD commonly report hypo or hyper sensitivity to certain stimuli, situations or environments, and up to 95 per cent of parents report an atypical sensory behaviour in their child with ASD. This exhibits in avoidance or seeking out types of visual stimulation, aversion to smells or colours, and seeming indifference to pain.

**Physical signs of visual impairment in autism include:**

- Gaze aversion
- Turning head, looking out of corner of eye
- Being attracted to shiny surfaces or mirrors
- Prolonged fixating on light patterns, windows
- Hyperfixating on one object while ignoring other objects in the room
- Always prefers/avoids a particular color
- Shows distorted body postures or orientation including arching back, hyperextension of neck
- Toe walking
- Touches all surfaces (walls, furniture, etc) when in an unfamiliar environment
- Anxiety or avoidance associated with fast

moving objects or animals

- Spinning objects close to face
- Intense light sensitivity
- Poor attention to one's surroundings as well as a lack of interest in ones environment
- Poor eye contact
- Excessively lines up toys or other objects
- Fidgets with objects repetitively
- Maintains interest in spinning objects for periods greater than a couple of minutes
- Does not follow where someone else is looking
- Flicking fingers or hands near face
- Stares at nothing with no apparent purpose

**Visual symptoms highly associated with ASD**

Hyper:

(1) focusing on tiny pieces of dust/particles,

(2) dislike of the dark and bright lights

(3) dislike of sharp flashes of light,(4)looking down most of the time,(5)covering/closing eyes at bright lights.

Hypo:

(1) attracted to light,

(2) looking intensely at objects or people,

(3) moving fingers or objects in front of the eyes,

(4) fascination with reflections and/or brightly coloured objects,

(5) running hands around the edges of objects.6

**Treatment to Improve Quality of Life**

After a comprehensive exam has identified whether vision problems are present, treatment can start. The goals of treatment may be to help the autistic patient to: organize visual space

- Gain peripheral stability
- Improve peripheral & central vision
- Gain more efficient eye coordination
- Improve visual information processing.

Achieving these goals can help the autistic& cerebral palsy person to feel less overwhelmed by visual stimuli and to interact with the world more comfortably.

**Neuro-Visual Postural Therapy**

NVPT provides a rehabilitative intervention for persons who have had a neurological impairment affecting vision and its function/process. The therapy is directed to affect spatial visual processing dysfunction and its relationship to the proprioceptive base of support (BOS). Following a neurological event the spatial visual process can become compromised which can lead to functional binocular vision difficulties as well as problems affecting balance and posture increasing risk of fall or affecting postural tone. The therapy works with the spatial visual process and the BOS to re-establish the integration of these processes.

**Visual Therapy**

1. Preparation:

- Referring patient to opthalmologist.
- Treatment should take place in a quiet area in order to minimise distractions.
- Perform a comprehensive subjective and objective assessment with the caregivers.

2. Postioning:

A child with CP has to overcome many factors to maintain a stable position. The child should be supported in order to focus

on visual input only. Supported sitting is therefore a good position to start visual stimulation. Different positions can also be implemented if child is comfortable. If specific one eye is more weak.,patching should be done to good eye.

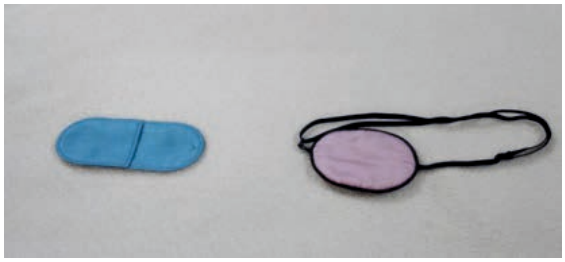


*Fig. 7: Eye patch*

**3.Introduction to visual stimulus:**

Attention should be given to the caregivers' observation. They will often note that children tend to stare at lights (as observed in 60% of children with CVI), respond to high contrast colours or objects illuminated by light in a darkroom and they will often be aware in which area of the visual field the child responds best to. The following low-cost objects can be used during treatment:

**Contrast**



*Fig.8: Contrast*

**Visual Guided Toys and Reflective Object**



*Figure 9 and 10: Visual Guided Toys and Reflective Object*

**Using light to illuminate objects**



*Fig.11: Using light to illuminate objects*

4. Use tactile and verbal stimulation to assist with the location and identification of objects.
5. Improve co-ordination (hand-eye, leg-eye):Kick a ball against a wall and retrieve it; Dribble a ball along a straight line and in and out of cones, jumping through obstacles.
6. Start with shorter treatment sessions as children may become overstimulated and irritated due to fatigue and/or frustration. (Recommended duration is less than 25 minutes, twice per day, 5 days a week for one year.)
7. Give appropriate education and home programme. This may be in the form of an individualised family service plan or programme. The family centered approach is essential.

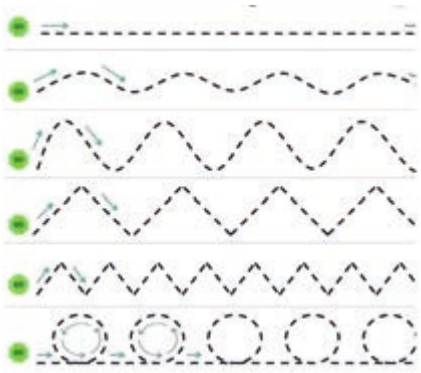


*Fig 12: Visual motor co-ordination Exercises*

**Home Programme**

- Comply with the exercises as prescribed above.
- Try to incorporate stimulation into activities of daily living (ADLs)
- Try to create a quiet and calm & dark environment for the child when doing vision stimulation exercises. Don't leave the radio or television on throughout the day as background noise can overstimulate and irritate them.
- Eye hand coordination activities (color sorting, puzzle solving, shape matching) with patching of either of the eyes for 10-15 minutes.
- Tracking with eyes/ finger of different patterns with help of laser light or flash light in the dark room.
- Bate's method (keeping two objects equidistant from the eyes and moving one object in different direction while keeping the other object stable)

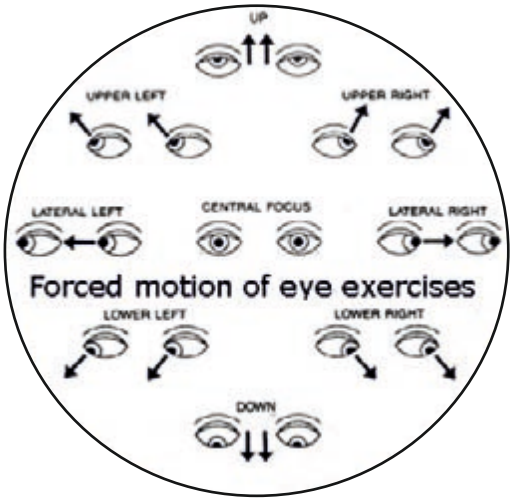



*Fig 13. Bate's method*



*Fig.14: Bate's method*

- Visual tracking of laser light or flash light on the wall of the dark room to recognize basic numbers or letters.
- Focusing on different visual guided toys., tracking horizontal,vertical.






*Fig 15 and 16: Astigmatism exercises*



*Fig 17:Hand eye Co-ordination*



*Fig 18: Hand eye Co-ordination*

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# Section 7
# Stem Cell Therapy

# Chp. 27 Overview on Stem Cells and Their Therapeutic Potential

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Regenerative medicine is an evolving branch of modern medicine that has the potential to heal or replace tissues and organs damaged by age, birth, disease, or trauma. It encompasses various strategies including cell-based therapy. Cell based therapy involves use of stem cells which can restore or replace lost or damaged cells with healthy ones. This could therefore potentially hold the key for treating incurable neurological disorders such as spinal cord injury, cerebral palsy, autism, brain stroke, muscular dystrophy, Alzheimer's disease, motor neuron disease, multiple sclerosis, etc which cannot be completely cured with standard treatment strategies.

The central nervous system is like the central processing unit of the animal body. In humans, it is more evolved and specialized. Since, disorders and injuries affecting the nervous system lead to irreparable damage and disability, this area has become a major focus point in the arena of regenerative medicine. By combining plasticity of the nervous system with regenerative potential of stem cells, it would be possible to evolve definitive treatments for degenerative and traumatic disorders of the nervous system.

**What Are Stem Cells?**

Stem cells are immature cells that exist in most tissues of the body at all stages of development, from early embryogenesis all the way throughout adult life of an organism. These cells have a prolonged self-renewal capability and can differentiate into more cells of same type or different. Stem cells may replicate through two types of cell divisions: (i) symmetric: both daughter cells are identical as parent stem cells and continues to contribute to the original stem cell line, (ii) asymmetric: one of two cells is identical to parent stem cells and the second differentiates into more specialized cell lineage. They also secrete paracrine molecules which indirectly help in neuroprotection and repairing the damage via paracrine mechanisms.

Stem cells can be classified depending on their potency or the origin. These cells can be totipotent, pluripotent, multipotent or unipotent. Stem cells also can be classified based on their origin like embryonic stem cells, adult stem cells, umbilical cord blood cells and induced pluripotent stem cells.

**Potency of stem cells:**

There exists a hierarchy in the stem cell compartment, depending on their 'potency' or fate restriction.

1) Totipotent cells have ability to differentiate into cell lineages from all three germ layers: mesoderm, endoderm, and ectoderm including placental cell. This means, it has the capacity to form whole of the embryo.
2) Pluripotent stem cells have the capacity to give rise to cells of all the three germ layers of the embryo, i.e., endoderm, mesoderm and the ectoderm. Pluripotent stem cells are cells from the inner cell mass of the blastocyst (ICM), epiblast (EPSC) and SC obtained as immortalized cell lines - blastocyst derived embryonic stem cells (ES) and

Primordial Germ Cell-derived embryonic germ cells (EG).

3) Multipotent stem cells give rise to cells of one of the germ cell layers only, either ecto-, meso- or endoderm. Sources range from 8 day old embryo to adult bone marrow.

4) Unipotent stem cells are tissue-committed stem cells that give rise to cells of one lineage, e.g., hematopoietic stem cells, epidermal stem cells, intestinal epithelium stem cells, neural stem cells, liver stem cells or skeletal muscle stem cells.

Though the above classification has evolved over decades, understanding of the potency of these cells are everchanging. Many of these cells, which were earlier considered to be multipotent, have shown limited pluripotent properties. Also, transdifferentation of monopotent / unipotent cells by external stimulation or manipulation have shown that these classifications, based on fate restriction or potency, are fast becoming redundant.

*Table 1: Potency of stem cells*

| Cell potency | Characteristics |
|---|---|
| Unipotent | Capability to produce cells of their own type |
| Multipotent | Capability to differentiate into few types of cells from one germ layer |
| Pluripotent | Capability to differentiate into cells from all three germ layers except for placenta |
| Totipotent | Capability to differentiate into cell linages from all three germ layers: mesoderm, endoderm, and ectoderm including placental cell |

**Types of Stem Cells**

Stem cells can also be classified according to their origin as embryonic stem cells, adult stem cells, umbilical cord stem cells and induced pluripotent stem cells.

**(A) Embryonic Stem cells:**

ESCs are pluripotent cells isolated from the inner cell mass of the blastocyst. They present two distinctive properties: they are able to differentiate into all derivatives of three primary germ layers, and they are capable of propagating themselves indefinitely, under defined conditions.

The potential to replicate indefinitely and differentiate into germ layers allows embryonic stem cells to be a useful tool for both research and regenerative medicine but has instigated debates regarding ethics and legalities. There is also risk of tumorogenicity involved in use of ESCs. To realize the potential of ESCs, it is essential to be able to control its differentiation and to direct the development of these cells along specific pathways.

**(B) Adult Stem Cells**

Adult stem cells are self-renewing and have ability to differentiate into the mature cell of it resident environment and also, may have transdifferentiating abilities. They are found in specific microenvironment called stem cell niche such as liver, brain, bone marrow, adipose tissue, heart, etc. However, their quantity and quality decreases with the age. These are a preferred source of stem cells as they do not involve any ethical, moral or legal issues as compared to the use of embryonic stem cells.

**1. Bone Marrow**

Bone marrow is the most accessible and most studied source of adult stem cells. It consists

of different types of stem cells which differ in their potential to differentiate. Initially, the bone marrow was thought to contain only haematopoietic stem cells. However, evidence suggests presence of heterogenous population of cells having varying plasticity.

**(a) Mononuclear Cells:**

Bone marrow mononuclear cells are a heterogeneous population that includes hematopoietic lineage cells such as lymphocytes, monocytes, stem cells and progenitor cells as well as mesenchymal stromal cells, along with endothelial progenitor cells (EPCs) and very small embryonic like (VSELs) stem cells. Mononuclear cells are isolated from human adult bone marrow, peripheral blood and umbilical cord. This mixture of cells has shown promising therapeutic potential in various neurological conditions

**(b) Mesenchymal Stem Cells (Mesenchymal Stromal Cells):**

Mesenchymal stem cells (MSC) are nonhematopoietic multipotent cells. They have the potential to differentiate into multiple lineages including bone, cartilage, muscle, tendon, ligament fat and a variety of other connective tissues. Indeed, marrow-derived cells seem to retain a remarkable plasticity, since they have much wider differentiation potential than previously thought. Marrow cells have been reported to contribute to angiogenesis, somatic muscle development, liver regeneration, and the formation of central nervous system cell types. It is likely that MSC may be accompanied by other populations of primitive non-hematopoietic stem cells.

**(c) Multipotent Adult Progenitor Cells (MAPC):**

MAPC are isolated from BM as well from various adult organs as a population of CD45 GPA-A- adherent cells and they display a similar fibroblastic morphology to MSC. Interestingly MAPC are the only population of BM derived stem cells that have been reported to contribute to all three germ layers after injection into a developing blastocyst, indicating their pluripotency. The contribution of MAPC to blastocyst development, however, requires confirmation by other, independent laboratories

**(d) Marrow-isolated adult multilineage inducible (MIAMI) cells:**

This population of cells was isolated from human adult BM by culturing BM MNC in low oxygen tension conditions on fibronectin. MIAMI cells were isolated from the BM of people ranging from 3- to 72-years old. Colonies derived from MIAMI cells expressed several markers for cells from all three germ layers, suggesting that, at least as determined by in vitro assays, they are endowed with pluripotency. However, these cells have not been tested so far for their ability to complete blastocyst development. The potential relationship of these cells to MSC and MAPC is not clear, although it is possible that these are overlapping populations of cells identified by slightly different isolation / expansion strategies

**(e) Multipotent Adult Stem Cells (MACS):**

These cells express pluripotent-state-specific transcription factors (Oct-4, Nanog and Rex1) and were cloned from human liver, heart and BM- isolated mononuclear cells. MACS display a high telomerase activity and exhibit a wide range of differentiation potential. Again the potential relationship of these cells to MSC, MAPC and MIAMI described above is not clear, although it is possible that these are overlapping populations of cells identified by slightly different isolation / expansion strategies.

**(f) Very Small Embryonic Like (VSEL) Stem Cells:**

Recently, a homogenous population of rare

(~0.01% of BM MNC) Sca-1+ lin- CD45- cells was identified in murine BM. They express (as determined by RQ- PCR and immunhistochemistry) markers of pluripotent stem cells such as SSEA-1, Oct-4, Nanog and Rex-1 and Rif-1 telomerase protein. Direct electron microscopical analysis revealed that VSEL (2-4 μm in diameter) display several features typical for embryonic stem cells such as i) a large nucleus surrounded by a narrow rim of cytoplasm, and ii) open-type chromatin (euchromatin). Interestingly, these cells despite their small size possess diploid DNA and contain numerous mitochondria. VSEL, however, do not express MHC-1 and HLA-DR antigens and are CD90- CD105- CD29.

**(2) Central Nervous system (CNS)**

Adult CNS is a potential niche for isolation of neural stem cells (NSCs) Adult-derived neural progenitor and stem cells have been transplanted in animal models, and shown functional engraftment, supporting their potential use for therapy. Stem cell niches have now been identified in adult mammalian forebrain, a) in the subventricular zone (SVZ), subgranular zone (SGZ) and b) dental gyrus of the hippocampus. The most active NSC compartment is found in SVZ. Two main cell types are found in the SVZ: migratory, proliferating neuroblasts and astrocytes.

**(3) Nose:**

it contains cells with considerable regeneration potential, including neural cells, progenitor/ stem cells, and olfactory ensheathing cells. OECs can promote axonal regeneration by producing insulating myelin sheaths around growing and damaged axons, secreting growth factors, and generating structural and matrix macromolecules that lay the tracks for axonal elongation.

**(4) Skin:**

The skin harbors a distinct population of stem cells such as melanoblasts and epidermal Stem cells. These cells can generate both neural and mesodermal cell types and that most of the neural cells generated by them have characteristics of peripheral neurons and Schwann cells

**(5) Adipose tissue:**

The adipose tissue is a highly complex tissue and consists of mature adipocytes, preadipocytes, fibroblasts, vascular smooth muscle cells, endothelial cells, resident monocytes/macrophages and lymphocytes. Hence, this tissue compartment provides a rich source of pluripotent adipose tissue-derived stromal cells. It has been demonstrated that AT contains stem cells similar to BM-MSCs, which are termed processed lipoaspirate (PLA) cells. Exhibiting a neuronal-like morphology and expressing several proteins consistent with the neuronal phenotype.

**(6) Peripheral Nervous system (PNS):**

Schwann cells are the supporting cells of the PNS. Like oligodendrocyte, Schwann cells wrap themselves around nerve axons, but the difference is that a single Schwann cell makes up a single segment of an axon's myelin sheath. Schwann cells originating from dorsal and ventral roots are one of the cellular components that migrate to the site of tissue damage after spinal cord injury. The remyelinating capability of Schwann cells has been demonstrated in a number of studies and the functioning status of this myelin in conduction of neural impulses has confirmed.

**(7). Eye stem cells**

Stem cells have been identified in the adult mouse eye. Single pigmented ciliary margin cells were shown to clonally proliferate in vitro to form sphere colonies of cells that can differentiate into retinal-specific cell types,

including rod photoreceptors, bipolar neurons and Muller glia. The adult retinal stem cells were localized to the pigmentary ciliary margin and not to the central and peripheral retinal pigmented epithelium.

**(8) Dental Stem Cells:**

Different types of dental stem cells have been isolated from mature and immature teeth, dental pulp, exfoliated deciduous teeth, periodontal ligament, apical papilla and dental follicle. Dental stem cells are rich source of mesenchymal stem cells and neural cells. They are multipotent stem cells which are being widely explored for its potential in treatment of neurodegenerative and ischemic diseases.

**(9) Muscular Stem Cells:**

The progenitor/stem cells are also found in skeletal muscles which are also known as satellite cells and side progenitor (SP) cells. These stem cells are involved in repair of regular wear and tear of muscle fibers. These cells help to regenerate the damaged muscles.

**(C) Umbilical Cord Stem Cells**

Umbilical cord blood stem cells can be obtained from the umbilical cord immediately after birth. Cord blood stem cells have shown to be multipotent by being able to differentiate into neurons and liver cells. Umbilical cord blood is another rich source of hematopoietic stem cells and progenitor cells and is used as an alternative allogeneic donor source to treat various disorders. It is also a source of non-hematopoietic stem cells such as MSCs, endothelial cells, and unrestricted somatic stem cells (USSC).

There have been reports that matrix cells (wharton's jelly) from the umbilical cord also contain potentially useful stem cells. Wharton's jelly has been a source for isolation of mesenchymal stem cells. These cells express typical stem cell markers such as c-kit and high telomerase activity and can be induced to differentiate in vitro into neurons.

**Advantages of using cord blood as a source of stem cells are as follows:**

1) It is a non-invasive source and can be obtained from the umbilical cord immediately after birth.
2) Available in vast abundance; thousands of babies are born each day and the umbilical cord and placenta are discarded as waste.
3) High proliferative capacity
4) Despite its high content of immune cells, it does not produce strong graft-versus-host disease possibly due to high interleukin-10 levels produced by the cells and / or decreased expression of the beta-2-microglobulin.
5) Therefore, cord blood grafts do not need to be as rigorously matched to a recipient as other allogenic grafts.

Hence, cord blood has recently emerged as an alternative source of hematopoietic stem cells for treatment of leukemia and other blood disorders.

**However, there are a few disadvantages:**

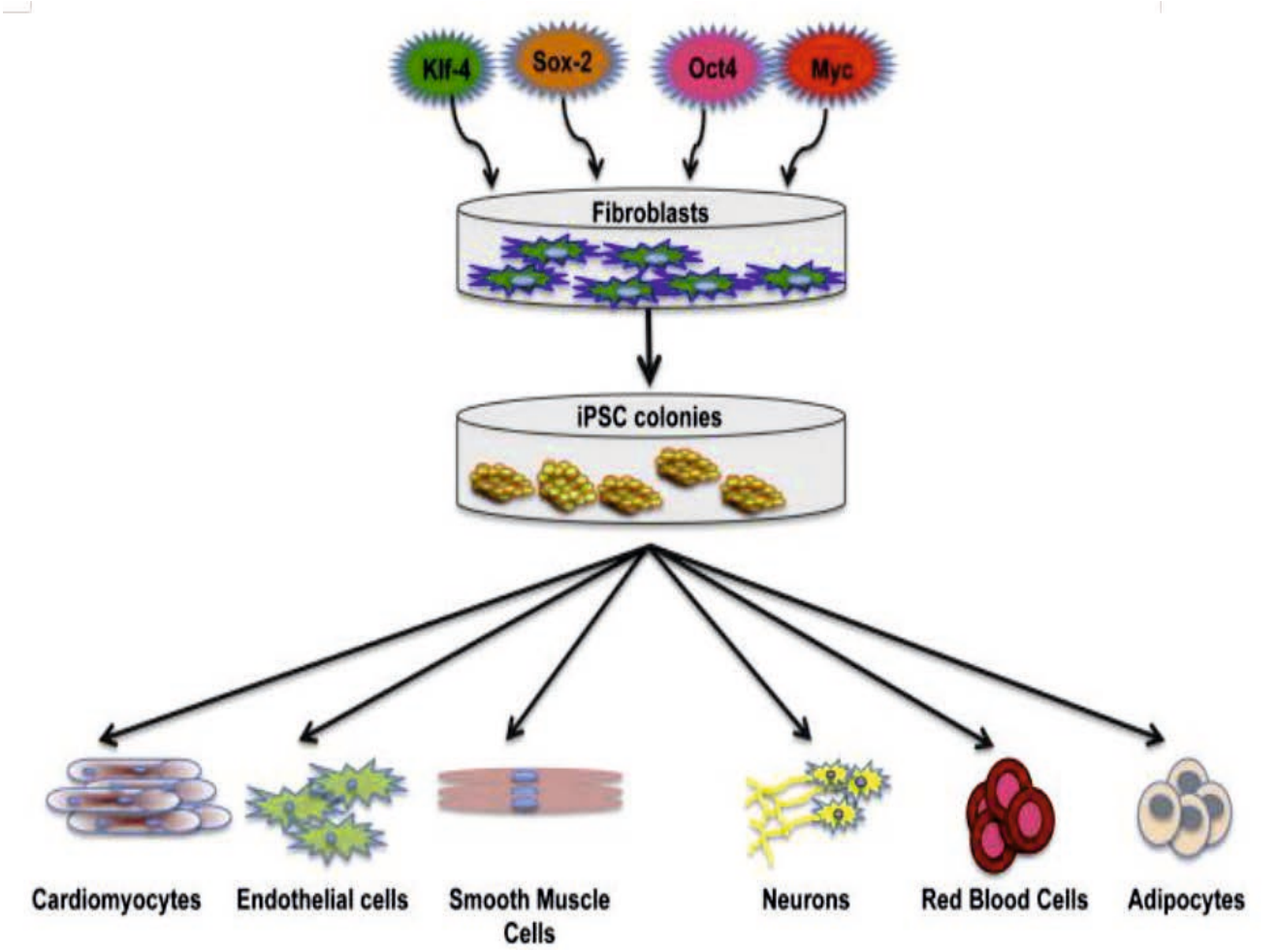
1. Slow engraftment
2. Limited cell dose- small volume of unit, additional cell dose unavailable
3. Autologous donation- limited benefit owing to hereditary disorders
4. Storage issues - unknown length of long term storage, Cost related to long term storage,
5. Quality control

All over the world, innumerable cord blood banks have been established for storage of umbilical cord stem cells. These are generally either pure public banks or private banks.

There are certain banks which offer both types of banking (mixed type). Umbilical cord stem cells banks also differ in the type of biological material that they store. Some banks only store the cord blood (from the umbilical vein) which predominantly carries the haematopoietic stem cells. Increasingly, banks have started storing pieces of the placenta and cord, which are a rich source of mesenchymal stem cells.

**(D) Induced Pluripotent Stem Cells:**

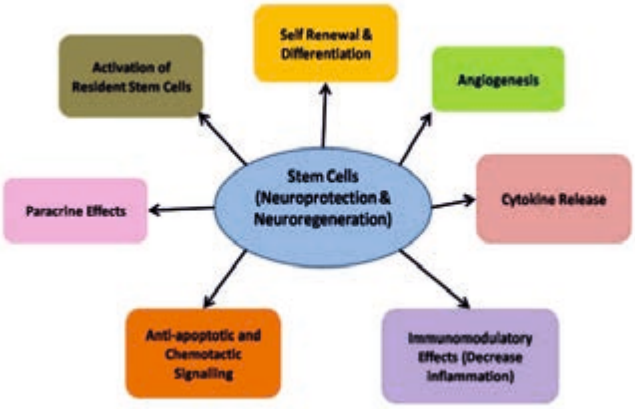
One of the emerging areas in laboratory investigations of stem cells is the attempt to induce differentiated somatic stem cells into pluripotent stem cells by inducing certain factors which will initiate cellular reprogramming. The induced pluripotent human stem cells have normal karyotypes, express telomerase activity, express cell surface markers and genes that characterize human ES cells, and maintain the developmental potential to differentiate into advanced derivatives of all three primary germ layers. These IPSCs sidesteps the ethical issues that have limited the use of embryonic stem cells, as they can be generated without the use of oocytes or cell from the preimplantation embryo. These cells can be autologous, thereby surmounting the problem of immune reaction. Thus, development of IPS cell technology can add to the sources of autologous cells for transplantation therapy.




*Courtesy: Nsair, Ali, and W. Robb MacLellan. "Induced pluripotent stem cells for regenerative cardiovascular therapies and biomedical discovery." Advanced drug delivery reviews 63.4 (2011): 324-330.*

The progression of Adult Stem Cells to Induced Pluripotent Stem Cells (IPSCs) is already a dynamic area of research in stem cell therapy. However, recent work has exhibited strong evidence that the adult somatic cells can be reprogrammed into mature neurons, without the in-between transition into IPSCs (41-43). There are recent reports which provide us sufficient evidence that transcription-mediated reprogramming of human fibroblasts into subtype specific neurons can be achieved without undergoing the proliferative progenitor stage. In one of the studies, the authors reported that the fibroblasts were reprogrammed into motor neurons, by forced expression of select transcription factors .

**Mechanism of Action**




The naturally occurring stem cells in the organs constantly repair the daily wear and tear of tissues through multitudes of mechanisms. In various disease models, the mechanism of action of stem cells has been studied in great depths. The pathways through which they act have also been studied in vitro. The micro cellular environment plays a crucial role in deciding the fate of stem cells. Stem cells carry out the repair process by neuromodulation, neuroprotection, axon sprouting, neural circuit reconstruction, neurogenesis, neuroregeneration, neurorepair, neuroreplacement and muscle regeneration. Mechanism of stem cells is divided into two categories- Direct cell replacement and indirect repair via paracrine mechanisms.

**1. Direct Cell Replacement:**

**Homing of the stem cells**

Stem cells are known to migrate to injured/ damaged sites on transplantation. This property is attributed to the expression of growth factors, chemokine and extracellular matrix receptors on the surface of cells. Evidence also exists that both endogenous and exogenous stem cells are able to migrate into the area of injury from the site of injection or infusion. MSC in the bone marrow can be mobilized, target the areas of infarction, and differentiate into target tissue type. Granulocyte colony-stimulating factor (G-CSF) has been studied widely and promotes the mobilization of bone marrow-derived stem cells in the setting of acute injury. This homing mechanism may also depend on expression of stromal cell-derived factor 1 (SDF-1), monocyte chemoattractant protein-3 (MCP-3), stem cell factor (SCF), and / or IL-8.

SDF-1 represents the major chemokine for initiating stem cell migration. The majority of cytokines that mediate stem cell migration do so via modulation either of SDF-1 or of its receptor, CXCR4. Thus the SDF-1/CXCR4 axis is central for stem cell mobilization. Post damage/ injury, injured tissue releases SDF-1 which stimulates the mobilization and homing of cells. These cells get recruited to the damaged/injured tissue and carry out further repair.

These cells self renew and differentiate into host cells and replace the damaged or dead cells. Their ability to differentiate into various other cell types is known as stem cell plasticity.

**Stem Cell Plasticity**

Stem cell plasticity refers to the capacity of the tissue derived stem cells to give rise to cell types, other than that of the resident tissues.

One of the mechanisms explaining stem cell plasticity is "cell fusion". An undifferentiated stem cell fuses directly with a more differentiated cell resulting in a single cell expressing properties of either cells. However, many reports have excluded this mechanism as experimental studies have not shown any evidence of cell fusion. Another mechanism justifying plasticity of stem cells is "transdifferentiation" or "dedifferentiation" wherein cells may differentiate from one cell type into another within the same tissue or develop into a completely different tissue without acquiring an intermediate recognizable, undifferentiated progenitor state. Also, there is a possibility of true pluripotent stem cells existing in various tissues of the body making it the most appealing explanation for the plasticity of stem cells.

Adult stem cells like bone marrow cells have been reported to generate a wide spectrum of different cell types, including hepatocytes, endothelial, myocardial, neuronal, and glial cells. In 1998, Ferrari et al. reported that mouse bone-marrow-derived cells give rise to skeletal muscle cells when transplanted into damaged mouse muscle. Hematopoietic cells can differentiate into cardiac myocytes and endothelial cells, functional hepatocytes and epithelial cells of the liver, gut, lung, and skin. Mesenchymal stromal cells (MSC) of the bone marrow can generate brain astrocytes . Enriched stem cells from adult mouse skeletal muscle were shown to produce blood. cells. In most of these plasticity studies, genetically marked cells from one organ of an adult mouse apparently gave rise to cell type characteristics of other organs following transplantation, which suggest that even cell types are plastic in their developmental potential.

A critical observation of adult stem cell plasticity is that in order for plasticity to occur, cell injury is necessary(16), thus micro-environmental exposure to the products of injured cells may play a key role in determining the differentiated expression of marrow stem cells.

**2. The Paracrine Effect**

Stem cells transplanted into injured tissue express paracrine signaling factors including cytokines and other growth factors, which are involved in orchestrating the stem cell-driven repair process through neuroprotection, increasing angiogenesis, decreasing inflammation, preventing apoptosis, releasing chemotactic factors, assisting in extracellular matrix tissue remodeling and activation of resident/satellite cells which is discussed further in details.

**Neuroprotection**

Stem cells secrete a vast array of neuroprotective growth factors including BDNF, nerve growth factor (NGF), neurotrophin-3 (NT-3), glial cell line-derived neurotrophic factor (GDNF), fibroblast growth factor-2, and insulin-like growth factor type 1. These growth factors activate a number of signalling pathways and help in survival of cells. Brain-derived neurotrophic factor (BDNF) is one of the most important growth factors. It has shown to enhance differentiation, survival of neurons and maintain neuronal functions.

**Increased Angiogenesis**

Stem cells produce local signaling molecules like vascular endothelial growth factor (VEGF), hepatocyte growth factor (HGF), and basic fibroblast growth factor (FGF2) that may improve perfusion and enhance angiogenesis to chronically ischemic tissue.

Chen et al. have recently shown that treatment with bone marrow stromal cells enhances angiogenesis by increasing endogenous levels of VEGF and VEGFR2. They previously demonstrated that administration of recombinant human VEGF165 to rats 48 h after stroke significantly

increased angiogenesis in the penumbra and improved functional recovery. Hepatic Growth Factor (HGF) exerts beneficial effects on neovascularization and tissue remodeling, while FGF2 is involved intimately with endothelial cell proliferation and may be a more potent angiogenic factor than VEGF.

When exposed to either insult or stress, mesenchymal stem cells (MSC) in cell culture and in vivo significantly increase release of VEGF, HGF, and FGF2, which may improve regional blood flow and promote autocrine self survival. Increased perfusion due to the production of stem cell angiogenic growth factor has also been associated with improved end organ function. Thus, VEGF, HGF, and FGF2 may be important paracrine signaling molecules in stem cell-mediated angiogenesis, protection and survival.

**Immunomodulation**

Stem cells appear to attenuate infarct size and injury by modulating local inflammation. When transplanted into injured tissue, the stem cell faces a hostile, nutrient-deficient, inflammatory environment and may release substances which limit local inflammation in order to enhance its survival. Modulation of local tissue levels of pro-inflammatory cytokines by anti-inflammatory paracrine factors released by stem cells (such as IL-10 and TGF-β) is important in conferring improved outcome after stem cell therapy.

**Anti-Apoptosis**

Stem cells in a third pathway promote salvage of tenuous or malfunctioning cell types at the infarct border zone. Injection of MSC into a cryo-induced infarct reduces myocardial scar width 10 weeks later. MSCs appear to activate an anti-apoptosis signaling system which effectively protects ischemia-threatened cell types from apoptosis. Furthermore, expression profiling of adult progenitor cells reveals characteristic expression of genes associated with enhanced DNA repair, upregulated anti-oxidant enzymes, and increased detoxifier systems. HGF has been observed to improve cell growth and to reduce cell apoptosis.

**Beneficial Remodeling of the Extracellular Matrix**

Stem cell transplantation alters extracellular matrix, resulting in post-infarct remodeling, strengthening of the infarct scar, and prevention of deterioration in organ function. MSCs improved this function by increasing the cellularity and decreasing production of extracellular matrix proteins such as collagen type I, collagen type III, and TIMP-1 which result in positive remodeling and function.

**Activation of Neighboring Resident Stem Cells**

Recent work demonstrates the existence of endogenous, stem cell-like populations in adult hearts, liver, brain, and kidney. These resident stem cells may possess growth factor receptors that can be activated to induce their migration and proliferation and promote both the restoration of dead tissue and the improved function in damaged tissue. Mesenchymal stem cells have also released HGF and IGF-1 in response to injury which when transplanted into ischemic myocardial tissue may activate subsequently the resident cardiac stem cells.

To sum up, although the definitive mechanisms for protection via stem cells remains unclear, stem cells mediate enhanced angiogenesis, suppression of inflammation, and improved function via paracrine actions on injured cells, neighboring resident stem cells, the extracellular matrix, and the infarct zone. Improved understanding of these paracrine mechanisms may allow earlier and more effective clinical therapies

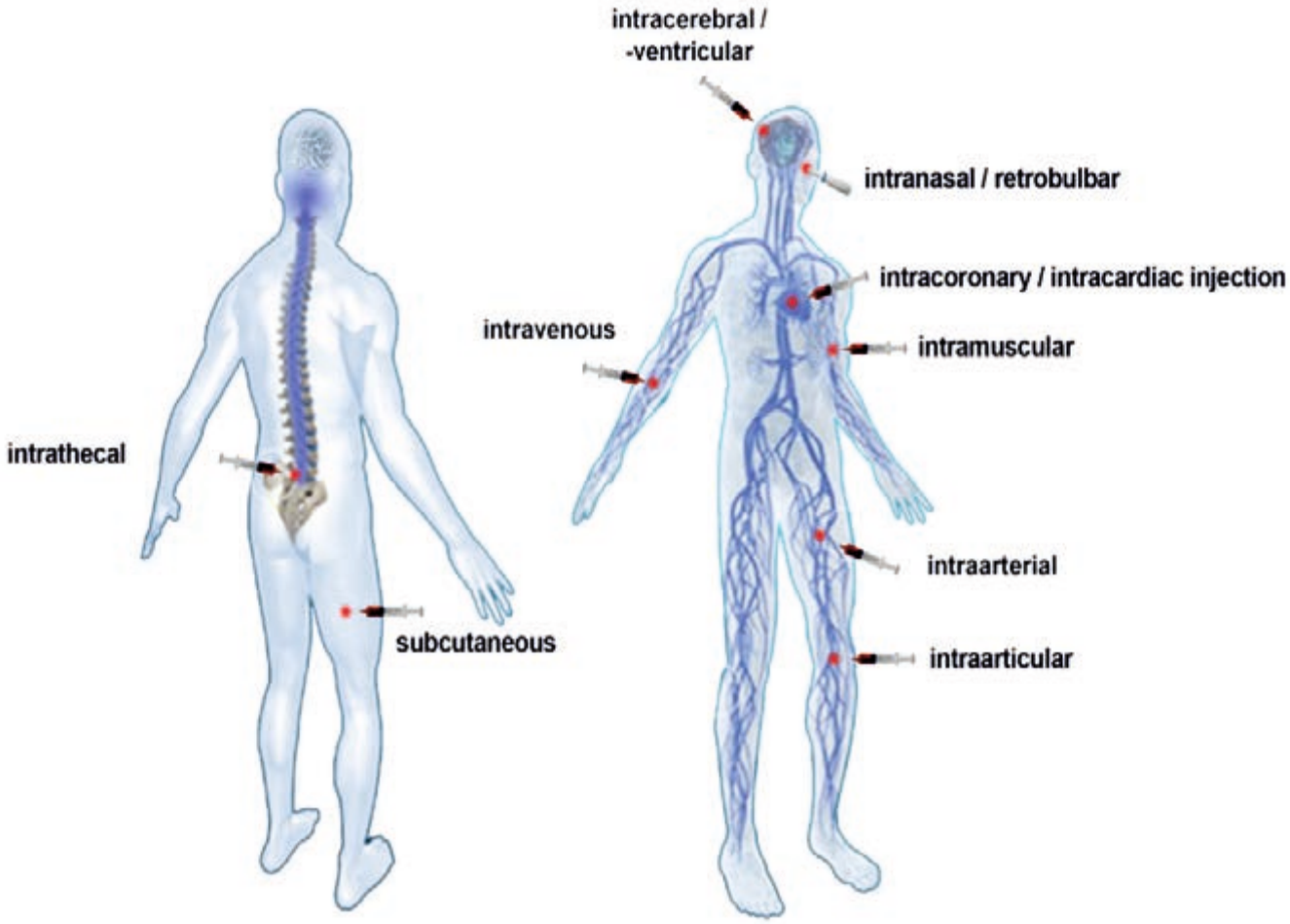
**Remyelination**

Remyelination is the phenomenon by which new myelin sheaths are generated around axons in the adult central nervous system. Previous attempts aimed at regenerating myelin-forming cells have been successful but limited by the multifocal nature of the lesions and the inability to produce large numbers of myelin- producing cells in culture. Stem cell-based therapy can overcome these limitations to some extent and may prove useful in the future treatment of demyelinating diseases.

Recent studies have shown that remyelination can be accomplished by supplying demyelinated regions with cells like Schwann cells, oligodendrocyte lineage cells lines, Olfactory ensheathing cells (OECs), embryonic stem cells and neural stem cells , Adult bone marrow derived stem cells. The remyelinating effect of these cells may be via one or more mechanisms, including: the stem cells act as an immunomodulator by producing soluble factors; they carry out direct cell replacement by differentiating into neural and glial cells in the lesion; and promote differentiation of endogenous cells. Interactions with viable axons and supportive astrocytic responses are required for endogenous immature cells to fulfill their potential remyelinating capacity.

Contrary to the general expectations that stem cells would primarily contribute to formation of tissue cells for repair, other mechanisms such as paracrine effects and remyelinations appear to be important ways via which stem cells seem to exert their effect. More Basic research to understand these mechanisms is underway throughout the world.

**Route of Administration of Stem cells**



**1. Intrathecal delivery**

The patient is positioned in the lateral decubitus position, in the curled up "foetal ball" position. Occasionally, the patient is made to sit, leaning over a table- top. Both these maneuvers help open up the spinous processes. The back is painted and draped and local anaesthetic is injected into the L4- 5 or L3-4 space. An 18G Touhy needle is inserted into the sub-arachnoid space. After ascertaining free flow of CSF, an epidural catheter is inserted into the space, far enough to keep 8-10 of the catheter in the space. The stem cells are then injected slowly through the catheter, keeping a close watch on the hemodynamics of the patient. The cells are flushed in with CSF. The catheter is removed and a benzoin seal followed by a tight compressive dressing is given. This procedure is usually done under local anesthesia. General anesthesia is given to children.

A spinal needle instead of a catheter is preferred in patients with cardiac problems, where excessive intravenous infusion is to be avoided, in patients on anti-coagulant or anti-platelet drugs so as to avoid bleeding into the sub-arachnoid space, in case where the spine is scoliotic which happens often in patients with muscular dystrophy and in some previously operated cases of lumbar spine surgery.

Sometimes in patients with severe spinal deformities such as scoliosis it is very difficult to get the needle intrathecally and at times assistance has to be taken of the C arm to exactly locate the point and direction of needle placement.

Callera et al (2007) demonstrated for the first time that autologous bone marrow CD 34+ cells labelled with magnetic nanoparticles delivered into the spinal cord via lumbar puncture (LP) technique migrates into the injured site in patients with spinal cord injury. They conducted the trial on 16 patients with chronic SCI. 10 of them were injected intrathecally with labelled autologous CD 34+ cells and the others received an injection containing magnetic beads without stem cells. Magnetic resonance images were obtained before and 20 and 35 days after the transplantation. Magnetically labelled CD 34+ cells were visible at the lesion site as hypointense signals in five patients, which were not visible in the control group.

**2. Intraspinal transplantation**

Direct implantation into the spinal cord may be done in one of many ways:-

a) Through a complete laminectomy from one level above to one level below the injury site so that there is sufficient access to the transplantation site. The dura is incised, sparing the arachnoid, which is subsequently opened separately with microscissors. The dorsal surface of the contusion site is located under high-power microscopic magnification. After exposure of sufficient surface in the contusion site, 300μL aliquots of cell paste (total volume, 1.8 mL) are injected into six separate points surrounding the margin of the contusion site. To avoid direct cord injury, 2 × 108 cells are delivered at a rate of 30 μL / min, using a 27-gauge needle attached to a 1-mL syringe. The depth of the injection site is 5 mm from the dorsal surface. To prevent cell leakage through the injection track, the injection needle is left in position for 5 min after completing the injection, after which the dura and arachnoid are closed. The muscle and skin are closed in layers.

(b) Though a minilaminectomy and exposure of the spinal cord. The dura is opened and a 27 gauge scalp vein is used by cutting one of the wings. The other wing is held by a hemostat and inserted at a 45 degree angle into the dorsal root entry zone. It is inserted 3mm deep into

the spinal cord. Two injections are made on either side above the injury site and two injections are made below the injury site. In China, surgeons are injecting 35 μL of stem cells. In his planned trials, Wise Young is intending to inject an escalating dose of 4 μL, 8 μL and 16 μL.

**3. Intra-arterial injection**

Following revascularization surgery such as Carotid endarterectomy or Superficial Temporal artery to Middle Cerebral artery bypass, stem cells could be injected directly intra-arterially immediately after the completion of the revascularization procedure. The advantage of this approach is that the stem cells would go directly to the ischemic brain and also that since the artery is already exposed no separate procedure needs to be done for the stem cell injection. The other method of direct intra-arterial injection would be via the endovascular interventional route. This is done by making a puncture in the femoral artery and negotiating a catheter to the arteries supplying the brain. The advantage of this is that it is a relatively non invasive procedure and the limitations of intravenous injection are avoided.

**4. Stereotactic implantation into the brain**

Cell transplantation for neurological conditions started with stereotactic implantation of fetal cells for Parkinson's disease. However, after a randomized trial done by Freed et al showed that the clinical outcomes were not significantly different from non transplanted patients this has now been given up. There are many stereo tactic systems available all over the world however the two most popular ones are the Leksell Stereotactic system and the CRW Stereotactic system. The Leksell system involves fixing the frame on the patients head and then getting a MRI done with the frame on. The area where the tissue is to be transplanted is identified on the MRI scan and then using the MRI software the X, Y and Z coordinates are obtained. The patient is now shifted to the operating room where a small burr hole is drilled into the skull and then through this the cells to be transplanted and inserted at the desired location using the X, Y and Z coordinates. The entire procedure is done under local anesthesia.

**5. Intramuscular injection**

In certain disorders, especially Muscular dystrophy, cells are also transplanted into the muscle. The points at which these have to be injected are termed as the "motor points. At these motor points, the area is cleaned with povidone iodine. The cells diluted in CSF are injected with the 26G needle going into the muscle at an angle (approx. 45 degrees).The piston / plunger of the syringe is slightly withdrawn to verify the needle is not inside a blood vessel. The cells are then injected, the needle removed and the site immediately sealed with a benzoin seal.

Stem cell transplantation, in its various forms, has been performed successfully in various neurological disorders. World wide reports reveal the use of bone marrow derived mononuclear cells and mesenchymal cells, umbilical cord blood stem cells, mesangioblasts, myoblasts, neural stem cells for various incurable and intractable neurological disorders. We have discussed the clinical results in detail in Chapter 28.

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# Chp. 28 Outcomes of Stem Cell Therapy In Neurological and Neuromuscular Disorder

**Dr. Alok Sharma, Dr. Nandini Gokulchandran, Dr. Hemangi Sane, Dr. Prerna Badhe, Ms. Pooja Kulkarni**

All over the world, cell therapy has been performed successfully in patients with various incurable neurological disorders. In this chapter, we have compiled and reviewed the clinical results of all the available studies. Along with which we have in detail described the results of the clinical studies published by NeuroGen Brain and Spine Institute.

At NeuroGen Brain and Spine Institute, Intrathecal Transplantation of autologous bone marrow mononuclear cells was carried out on various neurological disorders like Autism, Cerebral palsy, stroke, Muscular dystrophy, Spinal cord injury, Intellectual disability, Motor neuron disease, and other rare incurable neurological and neuromuscular diseases based on the Helsinki Declaration. Till date over 10000 patients with incurable neurological disorders have been administered Stem Cell Therapy.

The stem cell therapy process using autologous bone marrow derived stem cells consists broadly of 3 stages at NeuroGen Brain and Spine Institute.

**1. Aspiration:** (done in the operation theatre)

This is done by putting a needle into the hip bone, after making the area numb with local anesthetic, so that the patient does not experience pain. 100-120 ml of bone marrow is aspirated from inside the bone which takes about 20 minutes.

**2. Purification and Isolation:** (done in the stem cell laboratory)

The bone marrow removed from the patient is taken to the Stem cell laboratory, where the stem cells are separated from the remaining cells of the bone marrow by the density gradient method. This takes about 3 hours.

**3. Stem cell injection:** (done in the operation theatre)

Injection of stem cells into the spinal fluid is done by a very thin needle at lower back level (L4-5 space) after giving local anesthesia. The stem cells are injected this way which takes about 20 minutes.

In special cases, such as patients with muscular dystrophy, stem cells are also injected intramuscularly at motor points. Motor point is a point at which the motor branch of the innervating nerve enters the muscle. It is the point with the highest concentration of motor endplates, myoneural synapses and neuromuscular junctions.



*Fig 50.4: Aspiration of cells from the bone marrow*



*Fig 50.5: Separation and purification of stem cells*



*Fig 50.6: Intrathecal administration of stem cells*



*Figure 50.7: Intramuscular Injection*

Stem Cell Therapy is followed by an extensive personalized neurorehabilitation program.

Herein, we have described the clinical results of Stem Cell Therapy in incurable neurological conditions mainly Autism Spectrum Disorder, Cerebral Palsy, Intellectual Disability, Muscular Dystrophy, Spinal Cord Injury, Brain Stroke, Head Injury, Motor Neuron Disease and Ataxia.

**Autism Spectrum Disorder:**

**Worldwide studies:**

18 studies have been published including 6 clinical studies and 12 case reports demonstrating the effect of Stem Cell Therapy in ASD. Different types of cells were used namely autologous bone marrow mononuclear cells, fetal stem cells and umbilical cord blood cells.

In 2013, world's first clinical study showing safety and efficacy of stem cell therapy in autism was published by Sharma et al from Neurogen Brain and Spine Institute. It was an open label proof of concept study including 32 patients of autism who were administered autologous bone marrow mononuclear cells intrathecally. Following which, in 2019, another study was published in American Journal of Stem Cells which included a larger sample population of 254 patients. The results of these study have been described in detail below.

The next clinical study was published by Yong-Tao Lv et al, wherein they studied use of human cord blood MNCs and MSCs in 37 patients. CARS, CGI and ABC scales were used to assess the therapeutic efficacy in this study. A positive outcome was obtained in the treatment group.

In 2014, Bradstreet et al published their study using fetal stem cells including 45 children with autism. On follow up after 6 months and 12 months, there was a significant change in Autism Treatment Evaluation Checklist (ATEC) test and Aberrant Behavior Checklist (ABC) scores. Improvement was also seen in behavior, eye contact, appetite, etc.

In 2017, Dawson et al published their Phase 1 safety and tolerability study wherein they administered a single intravenous infusion

of autologous cord blood cells in 25 children with ASD. The infusions were safe, with no serious adverse events and occasional allergic reactions and irritability reported. Improvements were observed on caregiver-completed measures (Vineland Adaptive Behavior Scales Second Edition (VABS-II) and Pervasive Developmental Disorder Behavior Inventory (PDDBI)), clinician assessment (CGI-I) and computerized eye tracking assessments. Positive changes were seen in social communication skills, receptive/expressive language, repetitive behavior and sensory sensitivities six months post infusion and maintained at 12 months.

In 2018, Chez et al assessed the safety and clinical effects of autologous umbilical cord blood (AUCB) infusion in 29 children with ASD and observed a trend towards improvement, particularly in socialization and no adverse events were recorded.

**Our Published Data:**

**Publication 1:**

NeuroGen Brain and Spine Institute published world's first study on stem cell therapy in Autism in "Stem Cell International" in 2013. It was an open label proof of concept study conducted on 32 patients diagnosed with autism. Autologous bone marrow mononuclear cells (BMMNCs) were intrathecally administered in the patients, followed by neurorehabilitation. All patients were followed up for 26 months (Mean 12.7) Outcome measures used were ISAA, CGI and FIM/ Wee-FIM scales. Positron Emission Tomography Computed Tomography (PET-CT) scan recorded objective changes. Out of 32 patients, a total of 29 (91%) patients improved on total ISAA scores, 20 patients (62%) showed decreased severity on CGI-I and 29 (90.6%) patients showed improvements in the domain of Social relationships and reciprocity. Improved emotional responsiveness was observed in 18 (56%) patients and 25 (78%) patients improved in speech- language and communication domain. Behaviour patterns of 21(66%) patients improved. Hyperactivity or restlessness (71%) and engaging in stereotype and repetitive motor mannerisms (65%) decreased significantly. Sensory aspects improved in 14 patients (44%). (Figure:1) Cognitively they showed improved consistency in attention and concentration and response time. 71% patients showed better attention and concentration, 45% patients showed reduction in the delay in responding. Statistically significant difference between pre and post scores was observed ($p < 0.001$) on Wilcoxon Signed Rank Test. (table:1) On CGI-II 96% of patients showed global improvement. (Figure 2) The efficacy was measured on CGI-III efficacy index. (Figure 3) Functional neuroimaging in the form of PET - CT scan in eight patients, documented changes in brain metabolism which correlated with clinical improvements. (Figure 4) Few adverse events including seizures were controlled with medications in three patients. The encouraging results of this leading clinical study provide future directions for application of cellular therapy in autism.

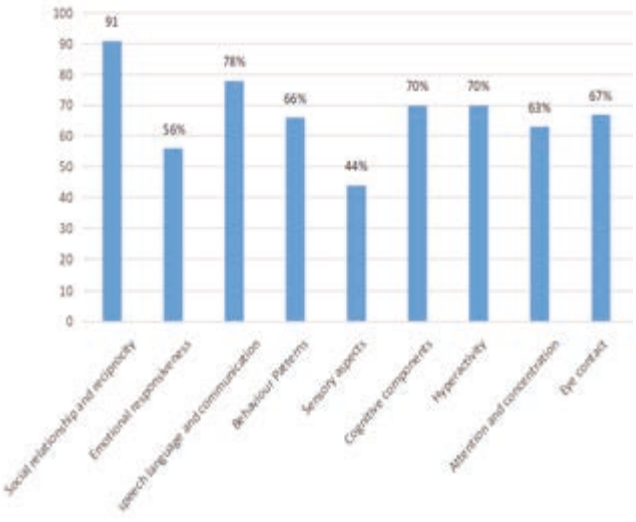



***Figure1: Symptomatic Improvements after Stem Cell Therapy***

Table 1: Change in the scores of CGI and ISAA before and after intervention.

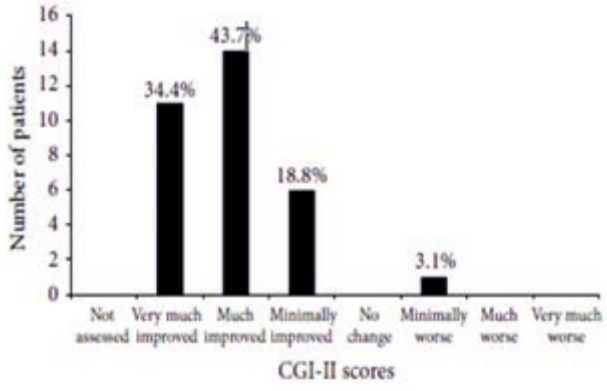



*Figure 2: Frequency distribution of participants on CGI-II scale*

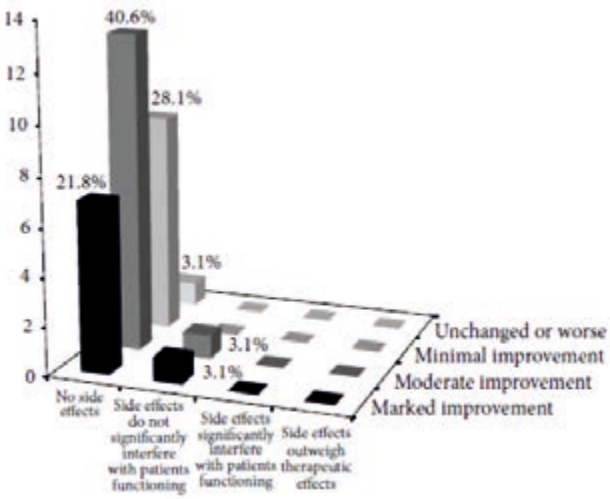



*Figure 3: Frequency distribution of participants on CGI-III scale*

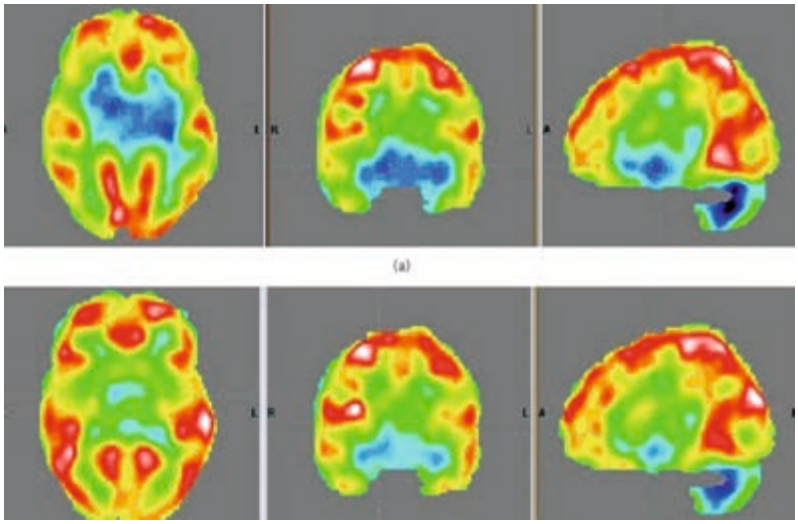



*Figure 4: Findings in PET-CT scan before and after cellular therapy. (a) PET-CT scan before intervention showing reduced FDG uptake in the areas of frontal lobe, cerebellum, amygdala, hippocampus, parahippocampus, and mesial temporal lobe. (b) PET-CT scan six months after intervention comparison shows increased FDG uptake in the areas of frontal lobe, cerebellum, amygdala, hippocampus, parahippocampus, and mesial temporal lobe.*

**Publication 2:**

In continuation to the first study, another study was published in "American Journal of Stem Cells" in 2019 which included a larger population of 254 patients of ASD showing benefits of Stem Cell Therapy. On mean follow up of 7.50 months, improvements were observed in eye contact, attention and concentration, hyperactivity, sitting tolerance, social interaction, stereotypical behavior, aggressiveness, communication, speech, command following and self-stimulatory behavior. Statistically significant improvement was observed in scores of ISAA and CARS after intervention.

On subgroup analysis, significantly better outcome of the intervention was found in patients at younger age and with shorter duration of disease [<5 years from time of diagnosis]. 86 patients who underwent a repeat PET CT scan showed improved brain metabolism after intervention in areas which correlated to the symptomatic changes. No major procedure related adverse events were recorded. However, 5 patients, with history of seizure and abnormal EEG, had an episode of seizure which was managed using medications. Outcome of intervention in these patients was not affected by seizures as improvements were observed in them.

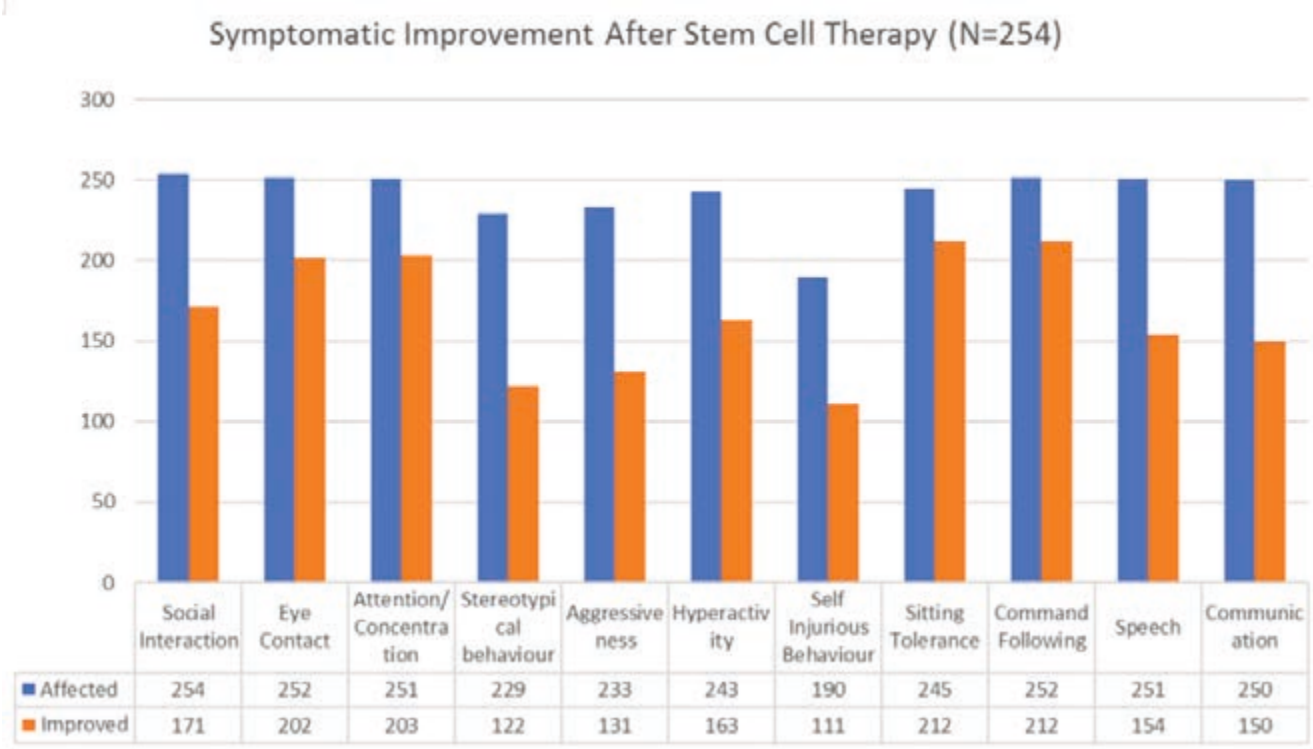



*Figure 5. Graph representing improvements in symptoms after intrathecal transplantation of autologous BMMNCs.*

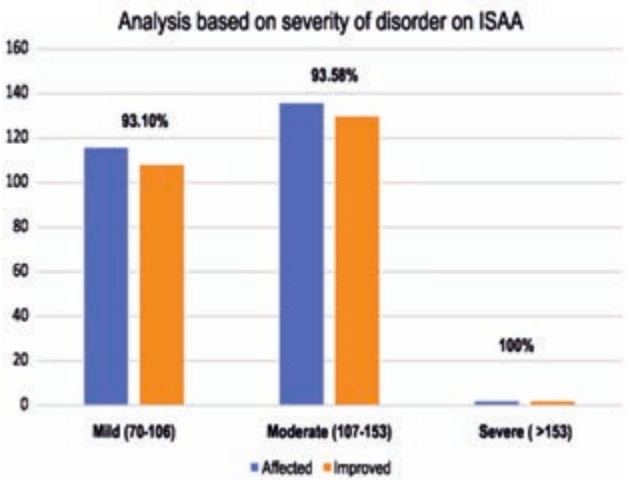



*Figure 6. Graph representing results based on severity of disorder on ISAA.*

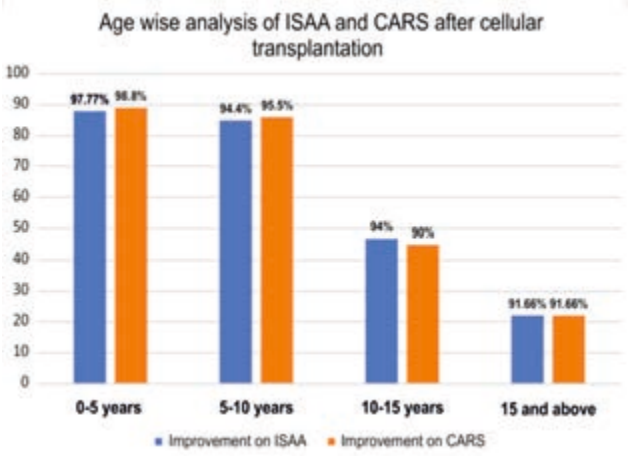



*Figure 7. Graph representing post Stem Cell Therapy results of ISAA and CARS based on age of patients.*

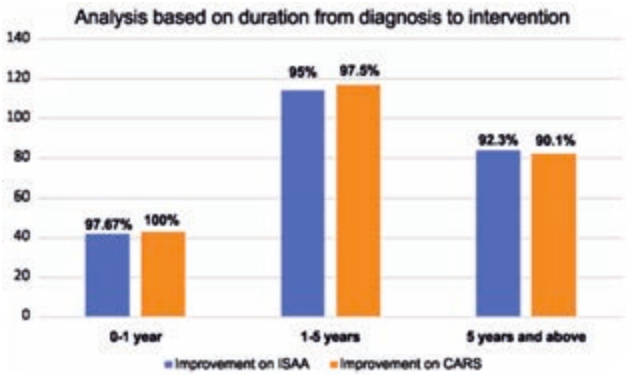



*Figure 8. Graph representing post Stem Cell Therapy results of ISAA and CARS based on duration from diagnosis to intervention*

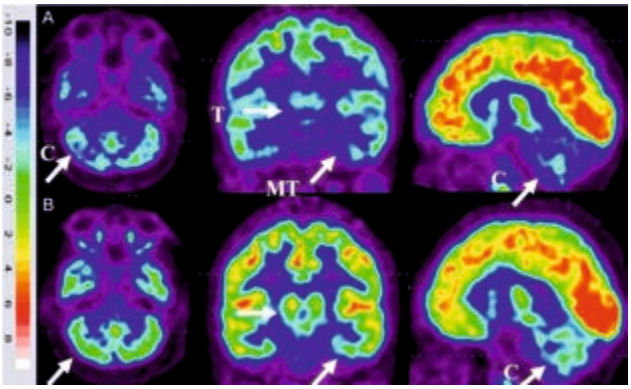



*Figure 9. Representative images of FDG-PET/CT scan brain of ASD patients performed before and 6 months after Stem Cell Therapy. A. Top Row: blue areas marked with arrows demonstrate hypometabolism. B. Below Row: green areas marked with arrows demonstrate improved metabolism following Stem Cell Therapy. Thalamus [T], Medial temporal cortex [MT] and Cerebellum [C].*

**Cerebral Palsy:**

**Worldwide studies:**

30 studies which include 17 clinical studies and 13 case reports have been published to demonstrate the effects of various types of stem cells in cerebral palsy. These studies used different cells such bone marrow derived stem cells, umbilical cord blood cells, Olfactory Ensheathing cells, embryonic cells and neural cells.

Many studies used bone marrow derived stem cells such as bone marrow mononuclear cells, mesenchymal cells, enriched bone marrow progenitor cells and total nucleated cells. These cells were administered either intravenously or intrathecally All the authors reported these cells to be safe and efficacious. Neurological and functional improvements were observed in all these studies. Umbilical cord blood cells have also shown a positive outcome after intravenous and intrathecal route of transplantation. No major adverse events were recorded in any study. Shroff et al, administered human embryonic stem cells intravenously in 91 patients. 63 patients showed improvement however, seizures were recorded as a side effect of the intervention. Seledtsov et al carried out a controlled study injecting a cell suspension from immature nervous and haematopoietic tissues. Their findings suggested that Stem Cell Therapy was an effective, safe and immunologically justified method of therapy for patients with cerebral palsy. Chen et al published the results of a randomized controlled study of intracranial administration of olfactory ensheathing cells. They reported these cells to be safe and efficacious. Similarly, Luan et al and Chen et al, administered neural stem cells and neural stem cell like cells intracranially and intrathecally in a large number of cases in China and reported it to be an effective treatment for CP. Whereas, Rah et al, demonstrated the neuroregenerative potential of intravenous G-CSF and autologous peripheral blood stem cells in CP, through a randomized, double-blind, cross-over study.

**Our Published Data:**

In 2015, Sharma et al published a study in "Stem cell international" which consisted of 40 patients who were administered with autologous bone marrow mononuclear cells intrathecally followed by neurorehabilitation to evaluate the benefits of stem cell therapy in cerebral palsy. Patients were followed up at 1

week, 3 months and 6 months after intervention. On follow up patients showed improvements in various symptoms. One week after the intervention, out of 40 CP patients, 6 had improvement in oromotor activities, 3 in neck control, 8 in sitting balance, 5 in standing and walking balance, and 6 in speech. Three months after intervention, 14 patients had improvement in oromotor activities, 11 in neck control, 17 in sitting balance, 15 in standing balance, 9 in walking balance, and 12 in speech. Overall, at six months, 38 out of 40 (95%) patients showed improvements. The total population was further divided into diplegic (Figure 5), quadriplegic (Figure 6) and miscellaneous group (Figure 7) of cerebral palsy. On statistical analysis using McNemar's test, a significant association was established between the symptomatic improvements and stem cell therapy in diplegic and quadriplegic cerebral palsy. PET-CT scan done in 6 patients showed metabolic improvements in areas of the brain correlating to clinical improvements.

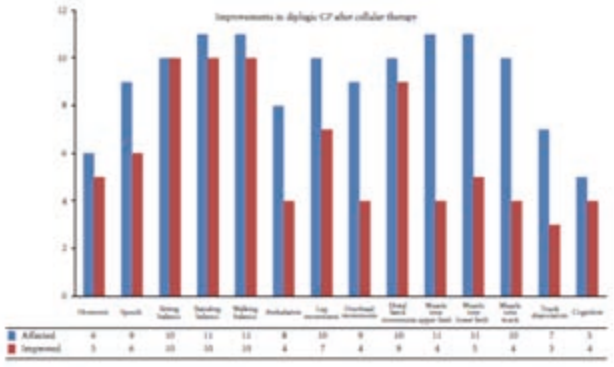


*Figure 10: Symptomatic Improvements after Stem Cell Therapy in Diplegic CP*

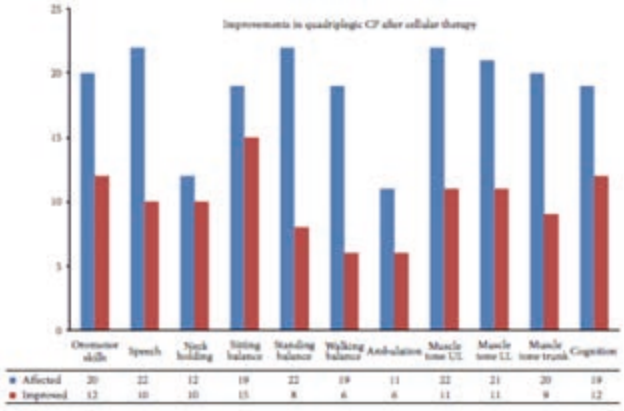



*Figure 11: Symptomatic Improvements after Stem Cell Therapy in Quadriplegic CP*

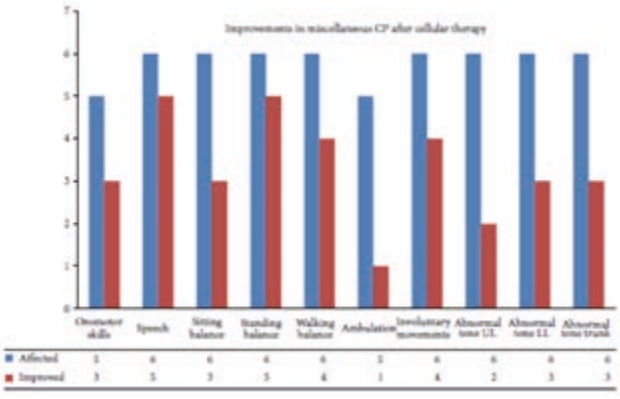



*Figure 12: Symptomatic Improvements after Stem Cell Therapy in miscellaneous CP*

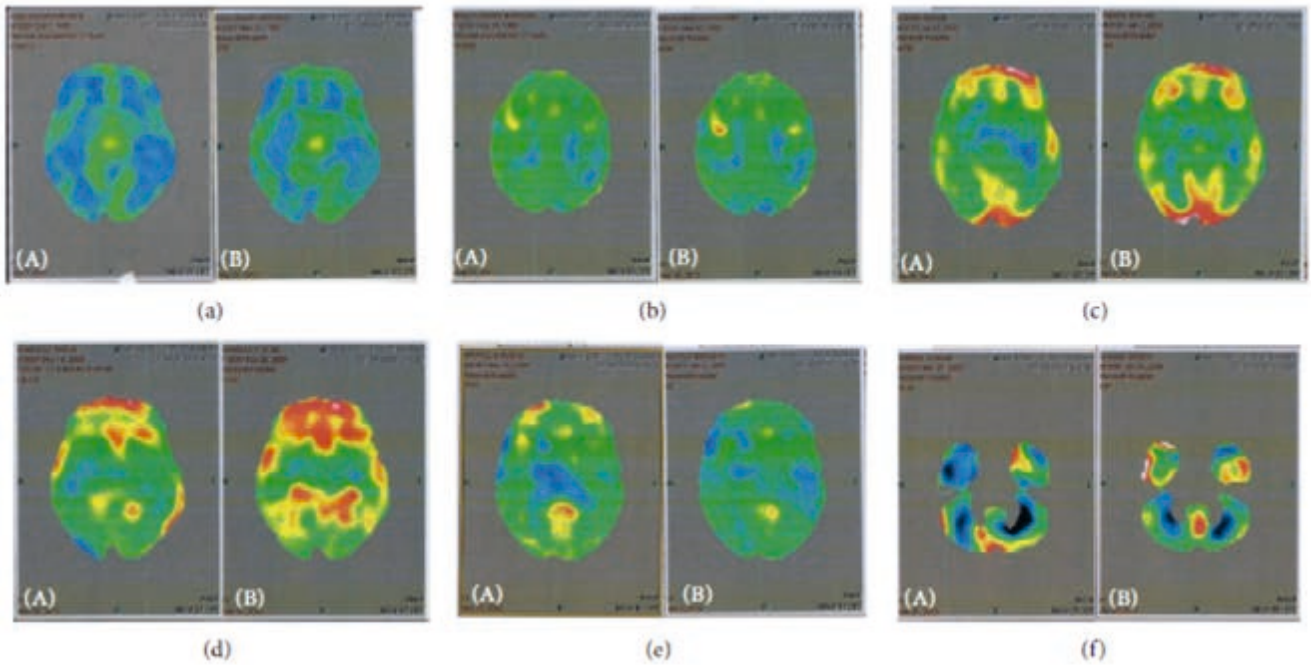



*Figure 13: Improvements in PET-CT scan brain: PET-CT scan images of (A) pre- and (B) postintervention showing increased metabolic activity in various areas. Blue areas indicate hypometabolism, green areas indicate normal metabolism, yellow areas indicate slightly high metabolism, and red areas indicate high metabolism.*

**Stroke:**

**Worldwide studies:**

Over 33 studies have been published demonstrating the benefits of Stem Cell Therapy in stroke. Different cell types studied are Autologous bone marrow mononuclear cells (BMMNCs), Autologous and allogenic mesenchymal cells (MSCs), Fetal porcine cells, Neural stem cells (NSCs), Immature neurons and hematopoietic cells, Olfactory ensheathing cells (OECs).

All these studies unanimously suggested that the transplantation of various types of stem cells was safe using various routes of administration. Although some adverse events were noted in these studies none of them were related to Stem Cell Therapy. Many studies showed that there was a statistically significant difference in the pre and post measures of Barthel Index, Modified Rankin Scale, National Institute of Health Stroke Score, Functional Independence Measure and Frugal Myer scale after stem cell therapy. Along with the functional improvements there were improvements in the metabolism as measured on PET-CT scan and some studies showed reduction in the lesion size on MRI.

**Our published data:**

In 2014, Sharma et al published a study in Stroke Research and Treatment which included 24 patients diagnosed with chronic stroke who were administered stem cell therapy, followed by multidisciplinary neurorehabilitation. They were assessed on Functional Independence Measure (FIM) objectively, along with assessment of standing and walking balance, ambulation and hand function. Out of 24 patients, 12 improved in ambulation, 10 in hand functions, 6 in standing balance, and 9 in walking balance. Statistically significant difference ($p < 0.05$) was seen in FIM scores before and after the Stem Cell Therapy. Patients of younger group (<60 years) showed higher percentage of improvement

in all the areas such as ambulation, hand functions, and sitting and standing balance and FIM scores (figure 10). Better improvements were observed in patients who underwent stem cell therapy within 2 years after the stroke. Patients with ischemic type of stroke had better outcomes as compared to those with haemorrhagic stroke. Also, patients with right brain involvement showed higher percentage of improvement as compared to the left brain. These results suggest that stem cell therapy may be a safe and effective treatment approach in improving the functional outcomes in the chronic stage of stroke.




*Figure 14: Symptom-wise improvements seen in patients of stroke after intrathecal administration of autologous BMMNCs.*

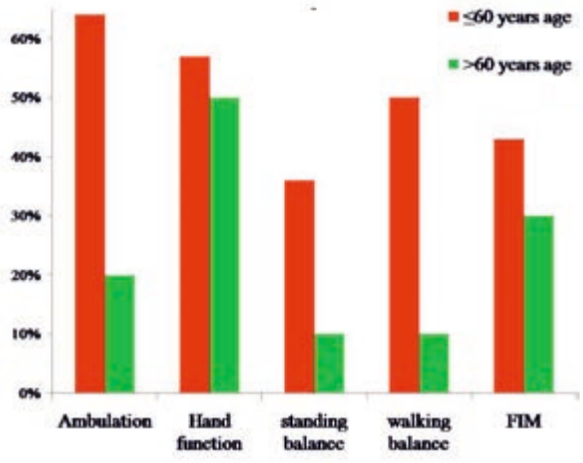



*Figure 15: Comparison of symptomatic improvements in patients according to duration since onset of stroke.*

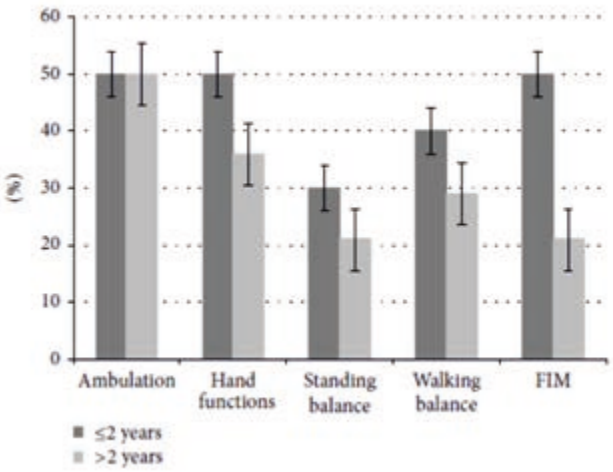



*Figure 16: Age-wise comparison of symptomatic improvements in patients of stroke*

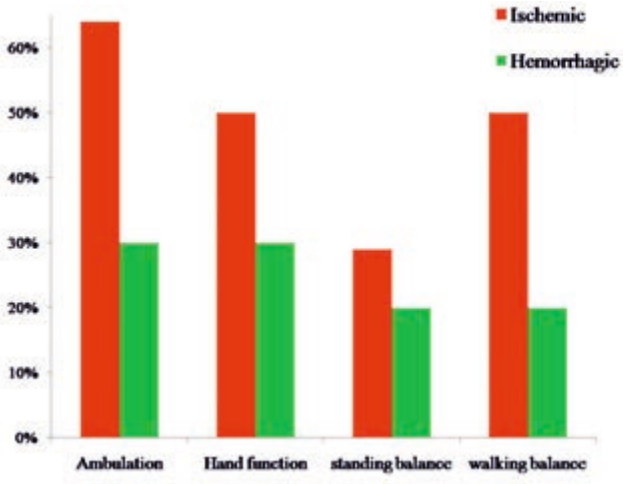



*Figure 17: Comparison of symptomatic improvements according to the type of stroke*

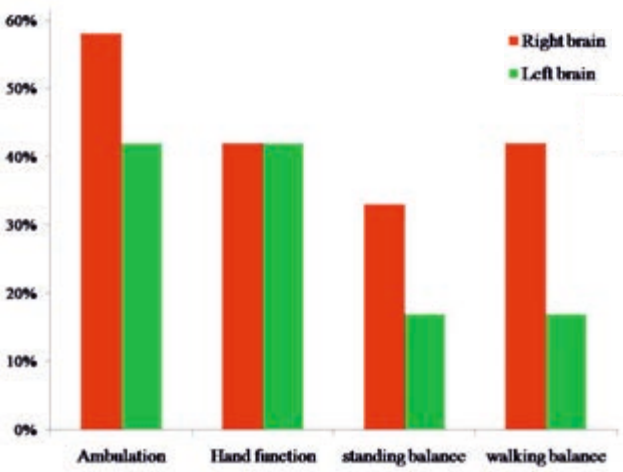



*Figure18: Comparison of symptomatic improvements according to side of the brain involved.*

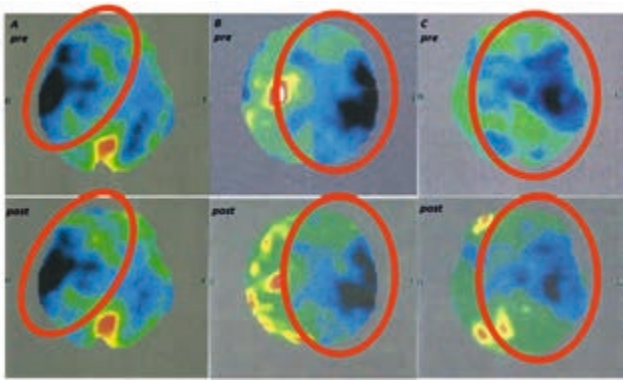


*Figure 19: 14 Increased FDG uptake seen in 3 patients after cell therapy. Blue areas representing hypometabolism have turned green after stem cell therapy which represents normal metabolism*

**Muscular Dystrophy:**

**Worldwide studies:**

30 studies which include 17 clinical studies and 13 case reports have been published to demonstrate the effects of various types of stem cells in cerebral palsy. These studies used different cells such bone marrow derived stem cells, umbilical cord blood cells, Olfactory Ensheathing cells, embryonic cells and neural cells.

Many studies used bone marrow derived stem cells such as bone marrow mononuclear cells, mesenchymal cells, enriched bone marrow progenitor cells and total nucleated cells. These cells were administered either intravenously or intrathecally All the authors reported these cells to be safe and efficacious. Neurological and functional improvements were observed in all these studies. Umbilical cord blood cells have also shown a positive outcome after intravenous and intrathecal route of transplantation. No major adverse events were recorded in any study. Shroff et al, administered human embryonic stem cells intravenously in 91 patients. 63 patients showed improvement however, seizures were recorded as a side effect of the intervention. Seledtsov et al carried out a controlled study injecting a cell suspension from immature nervous and haematopoietic tissues. Their findings suggested that Stem Cell Therapy was an effective, safe and immunologically justified method of therapy for patients with cerebral palsy. Chen et al published the results of a randomized controlled study of intracranial administration of olfactory ensheathing cells. They reported these cells to be safe and efficacious. Similarly, Luan et al and Chen et al, administered neural stem cells and neural stem cell like cells intracranially and intrathecally in a large number of cases in China and reported it to be an effective treatment for CP. Whereas, Rah et al, demonstrated the neuroregenerative potential of intravenous G-CSF and autologous peripheral blood stem cells in CP, through a randomized, double-blind, cross-over study.

**Our published Data:**

**Publication 1:**

A study including 150 patients of muscular dystrophy was published in "Cell Transplantation" by Sharma et al. The patients were administered autologous bone marrow-derived mononuclear cells intrathecally and intramuscularly at the motor points of the antigravity weak muscles followed by vigorous rehabilitation therapy. On a mean follow-up of 12 ± 1 months, overall 86.67% cases showed symptomatic and functional improvements, with six patients showing changes with respect to muscle regeneration and a decrease in fatty infiltration on musculoskeletal magnetic resonance imaging and nine showing improved muscle electrical activity on electromyography. 50% of the cases showed an increase in trunk muscle strength, 48% showed an increase in upper limb strength, 59% showed an increase in lower limb strength, and approximately 10% showed improved gait. Symptomatic improvements were also analysed in DMD, LGMD and BMD patients. These data were statistically

analyzed using Student's paired t test and found to be significant. A favourable shift on assessment scales such as the Functional Independence Measure and the Brooke and Vignos Scales was observed. No significant adverse events were noted. Furthermore, imaging and electrophysiological studies also showed significant changes in selective cases. The results show that this treatment is safe and efficacious and improves the quality of life of patients having muscular dystrophy.

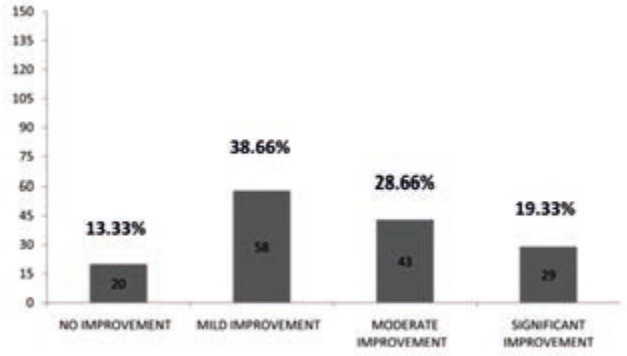



*Figure 20: Graph showing improvements in muscular dystrophy patients after stem cell therapy*

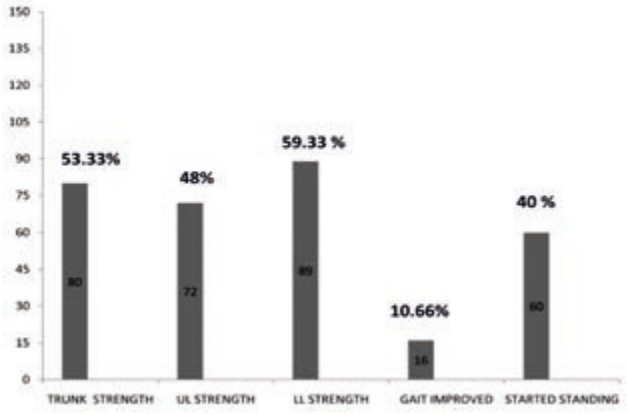



*Figure 21: Graph showing symptomwise improvements in muscular dystrophy patients after stem cell therapy. Number of patients showing improvements with respect to trunk strength, upper limb (UL) strength, lower limb (LL) strength, gait, and standing are shown.*

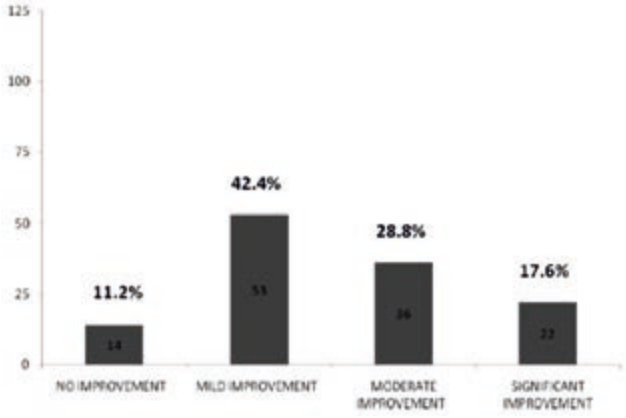



*Figure 22: Graph showing improvements in Duchenne muscular dystrophy (DMD) patients after stem cell therapy.*

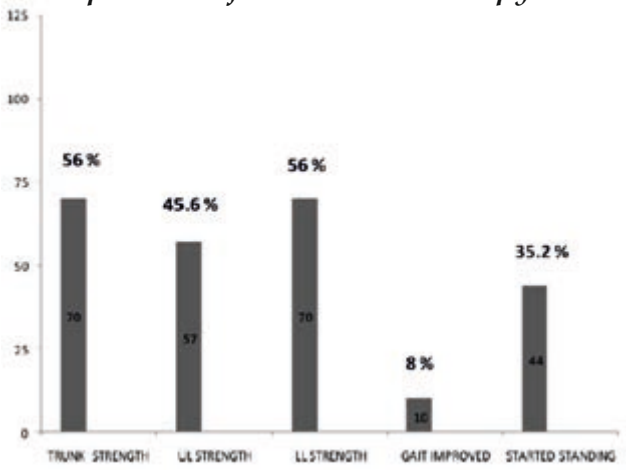



*Figure 23: Graph showing symptomwise improvements in DMD patients after stem cell therapy. Number of patients showing improvements with respect to trunk strength, upper limb (UL) strength, lower limb (LL) strength, gait, and standing are shown*

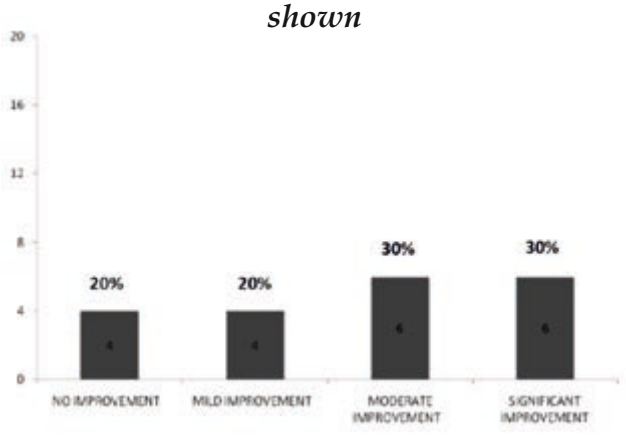



*Figure 24: Graph showing improvements in limb-girdle muscular dystrophy (LGMD) patients after stem cell therapy. y-axis = number of patients (n = 20).*

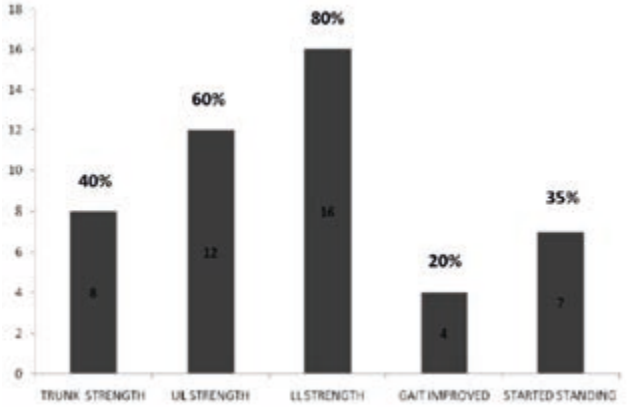



*Figure 25: Graph showing symptomwise improvements in LGMD patients after stem cell therapy. Number of patients showing improvements with respect to trunk strength, upper limb (UL) strength, lower limb (LL) strength, gait, and standing are shown.*

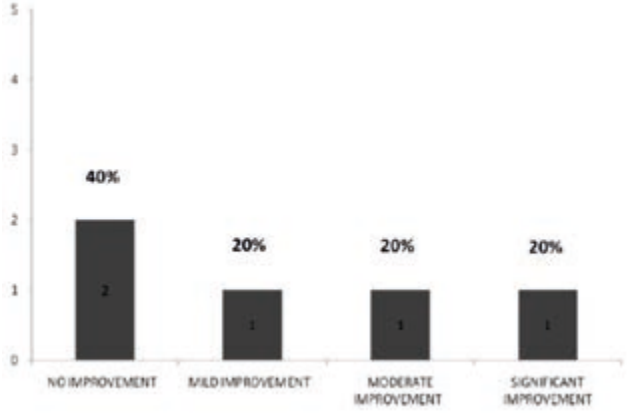



*Figure 26: Graph showing improvements in Becker muscular dystrophy (BMD) patients after stem cell therapy.*

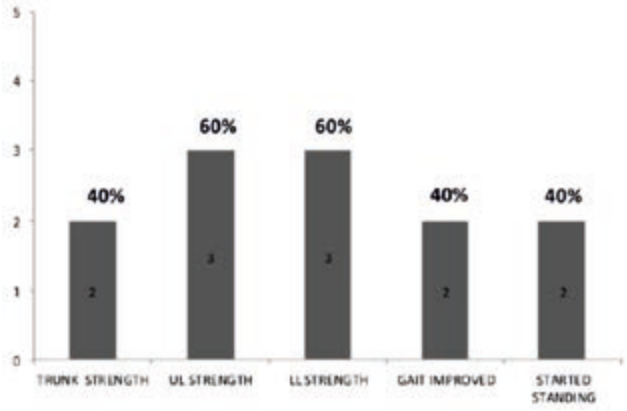



*Figure 27: Graph showing symptom wise improvements in BMD patients after stem cell therapy.*

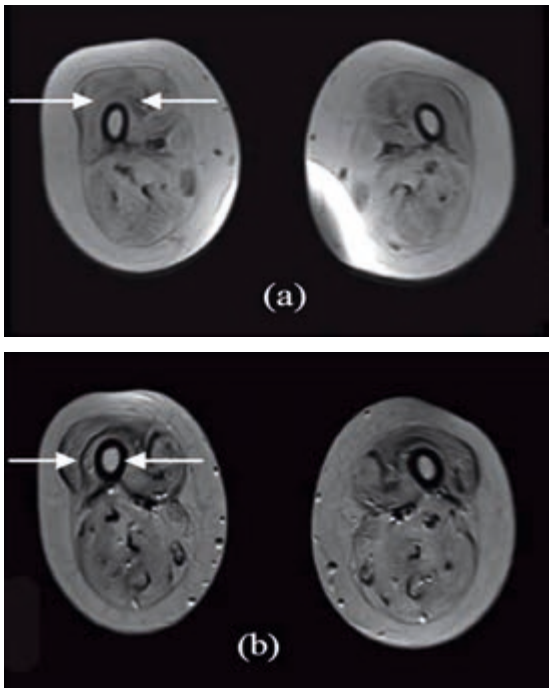



*Figure 28: Figures a & b shows the Musculoskeletal MRI images before and after stem cell therapy, respectively. Figure b shows regeneration of muscle in vastus medialis and vastus lateralis*

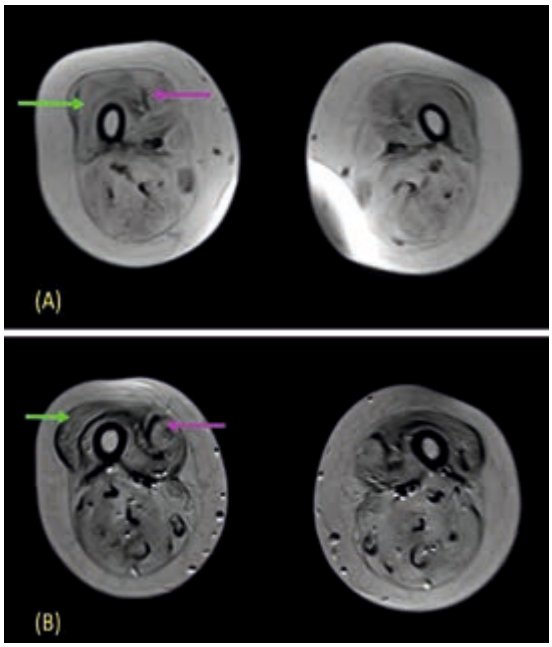



*Figure 29: Figures, A & B show MRI Musculoskeletal images before and after stem cell therapy. Regeneration of muscles is seen in Fig B.*

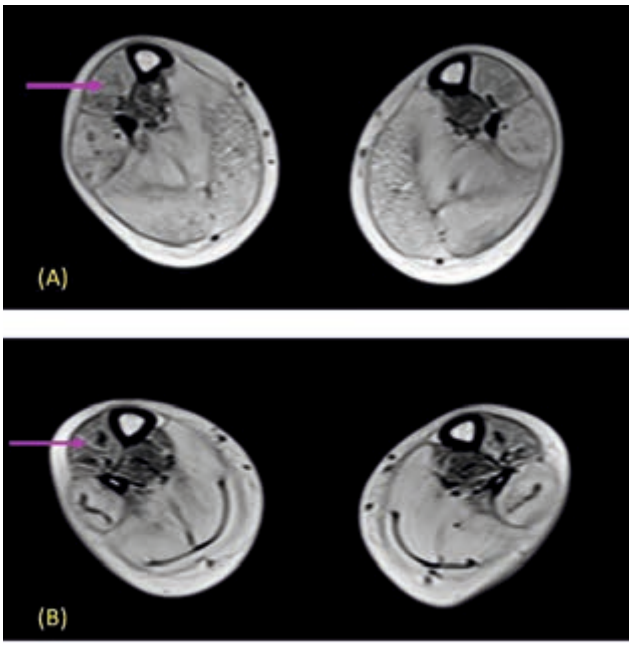



*Figure 30: The figures A & B show MRI Musculoskeletal images before and after stem cell therapy, respectively. Regeneration of muscles is seen in Figure B.*

**Publication 2:**

**Limb Girdle Muscular Dystrophy (LGMD)**

In 2015, Sharma et al published a study demonstrating effect of stem cell therapy in LGMD patients in the journal "Degenerative Neurological and Neuromuscular Disease". 65 patients underwent autologous bone marrow mononuclear cell transplantation intrathecally and intramuscularly to evaluate the benefits of stem cell therapy. Out of 65 patients, 59 cases completed the study and 6 were lost to follow up. Patients were divided into 3 groups depending on the number of transplantations administered. Group 1 included the cases who underwent one stem cell therapy (N=31). Group 2 included cases who underwent two transplantations (N=24). Group 3 included cases who underwent 3 transplantations (N=4). In group 1, 97% of the patients showed improved function. The total percentage of strength improvement ranged from 84% to 100% in all the muscles. In group 2, 96% of the patients showed an improved function. The total percentage of strength improvement ranged from 90% to 100% in all the muscles. In group 3, of the four patients, one patient deteriorated in his FIM score, whereas two patients improved, and one maintained functional status. Most of the patients had maintained muscle strength.

**Spinal cord injury:**

**Worldwide studies:**

So far 60 studies have been conducted demonstrating a beneficial effect of stem cell therapy in more than 1000 patients. Stem cells from different lineages and sub-types have been used in these studies which include autologous bone marrow derived mononuclear stem cells, bone marrow derived mesenchymal stem cells, bone marrow derived stromal cells, adipose derived mesenchymal stem cells, hematopoietic progenitor cells, peripheral hematopoietic cells, activated Schwann cells, olfactory ensheathing cells and umbilical cord mesenchymal stem cells. Improvements have been observed clinically in the form of improved functions and objectively in the form of improved ASIA scores and MRI changes. Post therapeutic adverse events were observed in a few patients such as fever, headache, nausea and vomiting, tingling sensations, spasms, neuropathic sensory symptoms including burning and pain sensations.

**Our published data:**

**1) Paraplegia:**

110 thoracolumbar SCI patients who underwent intrathecal administration of autologous bone marrow derived mononuclear cells followed by neurorehabilitation, were assessed. The outcome measures were Functional Independence Measure (FIM) score, American Spinal Injury Association scale (ASIA) and detailed neurological assessment. On a mean follow up of 2 years ±

1-month, overall improvement was seen in 91% of patients, including reduction in spasticity, partial sensory recovery, and improvement in trunk control, postural hypotension, bladder management, mobility, activities of daily living, and functional independence. A statistically significant association of these symptomatic improvements with the Stem Cell Therapy intervention was established using McNemar's Test. On electrophysiological studies, 2 showed improvement and 1 showed change in functional MRI. No major side effects were noted.

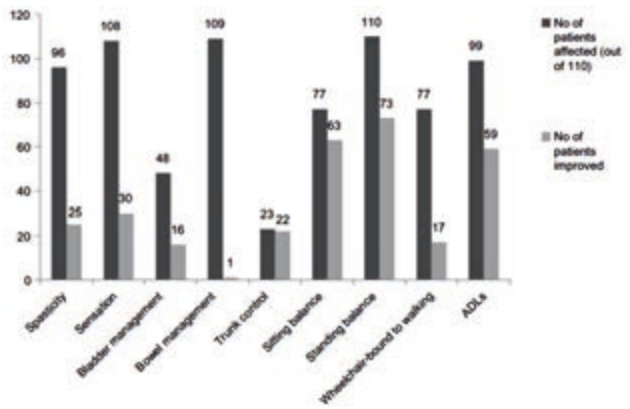



*Figure 31: Symptomatic improvements in patients with spinal cord injury after stem cell therapy.*

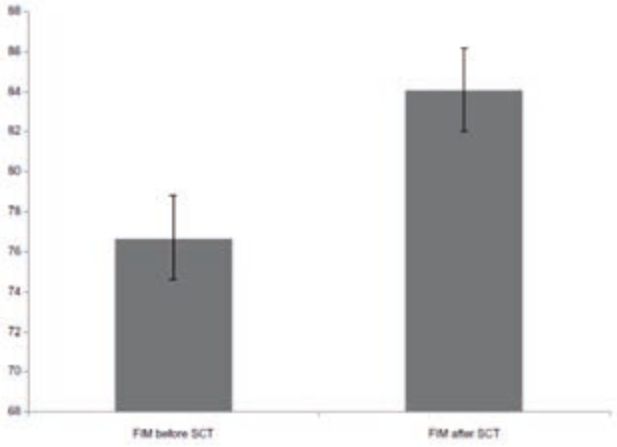



*Figure 32: Effect of stem cell therapy on mean FIM scores after stem cell therapy.*

| Symptom/function | Affected patients (n of 110) | Patients improved (n) | Chi-square value† | P value* |
|---|---|---|---|---|
| Spasticity | 96 | 25 | 23.04 | <0.0001 |
| Sensation | 108 | 30 | 28.033 | <0.0001 |
| Bladder management | 48 | 16 | 14.06 | 0.0002 |
| Bowel management | 109 | 1 | 0 | 1.000# |
| Trunk control | 23 | 22 | 20.045 | <0.0001 |
| Sitting balance | 77 | 63 | 61.016 | <0.0001 |
| Standing balance | 110 | 73 | 70.014 | <0.0001 |
| Wheelchair-bound to walking | 77 | 17 | 15.059 | 0.0001 |
| ADLs | 99 | 59 | 57.017 | <0.0001 |

**Notes:** *Significant at $P \leq 0.05$; †Chi-square value at one degree of freedom; #P value insignificant for improvement in bowel management.
**Abbreviation:** ADLs, activities of daily living.

*Table 2: Statistical significance for each symptomatic/functional change using McNemar's test*

| (A) | Nerve/sites | Amplitude 2–4 mV (before) | Amplitude 2–4 mV (after) |
|---|---|---|---|
| Patient 1 | R tibial (knee)-AH ankle | 3.5 | 5.4 |
| | R tibial (knee)-AH knee | 2.7 | 5.1 |
| | L tibial (knee)-AH ankle | 4.1 | 5.7 |
| | R tibial (knee)-gastrocnemius knee | 7.2 | 14.8 |
| | L tibial (knee)-gastrocnemius knee | 10.2 | 11.7 |
| Patient 2 | L common peroneal-EDB ankle | 0.8 | 3.0 |
| | R common peroneal – tibialis anterior, fibular head | 1.6 | 3.4 |
| | L common peroneal – tibialis anterior, fibular head | 1.8 | 4.3 |
| | R tibial (knee)-AH ankle | 7.0 | 8.0 |
| | L tibial (knee)-AH ankle | 7.9 | 8.3 |
| | R tibial (knee)-gastrocnemius knee | 6.2 | 18.7 |
| | L tibial (knee)-gastrocnemius knee | 2.5 | 17.2 |
| **(B)** | **Functional MRI (before)** | **Functional MRI (after)** | |
| Patient 1 | No activity in the pre and post central gyri | Activation in the right precentral gyrus | |

Abbreviations: AH, abductor hallucis; EDB, extensor digitorum brevis; MRI, magnetic resonance imaging; R, right; L, left.

*Table 3: Objective improvements evident on electromyography (A) and functional magnetic resonance imaging (B) after stem cell therapy in selected patients*

**2) Quadriplegia:**

A detailed study was conducted on 56 chronic cervical SCI patients. These patients underwent intrathecal administration of autologous bone marrow mononuclear cells followed by Neurorehabilitation. The outcome was recorded at a mean follow up of 2 years ±1 month. The outcome measures were Functional Independence Measure (FIM) score, American Spinal Injury Association scale (ASIA) and detailed neurological assessment. symptoms such as trunk stability, sitting balance, trunk muscle strength, upper limb strength, standing balance, deep touch sensation, bladder sensation, spasticity and walking balance demonstrated improvements. The improvement was statistically significant using Wilcoxon Signed Rank test. A detailed analysis of factors such as age, cause of injury, chronicity of injury and rehabilitation before the intervention was performed. No major side effects were noted in the duration of 2 years in both the studies. A better outcome was observed in thoracolumbar injury as compared to the cervical injury suggesting that the level of SCI greatly influences the recovery of the patient. Both studies demonstrated statistically significant clinical and functional outcome.

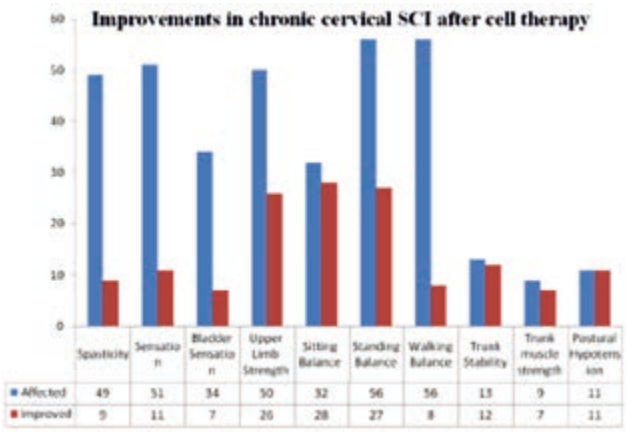



*Figure 32: Graph demonstrating symptomatic improvements in chronic cervical SCI patients after Stem Cell Therapy*

| Symptom | No. of patients affected | No. of patients improved | McNemars test value | P value |
|---|---|---|---|---|
| Spasticity | 49 | 9 | 7.11111 | *0.00766 |
| Sensation | 51 | 11 | 9.09091 | *0.00257 |
| Bladder Sensation | 34 | 7 | 5.14286 | *0.02334 |
| Upper Limb Strength | 50 | 26 | 24.03846 | *<0.000001 |
| Sitting Balance | 32 | 28 | 26.03571 | *<0.000001 |
| Standing Balance | 56 | 27 | 25.03704 | *<0.000001 |
| Walking Balance | 56 | 8 | 6.125 | *0.01333 |
| Trunk Stability | 13 | 12 | 10.08333 | *0.0015 |
| Trunk muscle strength | 9 | 7 | 5.14286 | *0.02334 |
| Postural Hypotension | 11 | 11 | 9.09091 | *0.00257 |

*significant at p value ≤ 0.05

***Table 4: Statistical analysis for each symptomatic improvement in cervical SCI using McNemar's test***

| Factors | | Percentage improvements |
|---|---|---|
| Age | <18 yrs | 100% |
| | 18-35 yrs | 41% |
| | >35 yrs | 42% |
| Cause of Trauma | RTA | 37.20% |
| | Non-RTA | 30% |
| Chronicity | 1-3 yrs | 47.82% |
| | 3-5 yrs | 33.33% |
| | >5 yrs | 44.44% |
| Rehabilitation | Done | 36.84% |
| | Not Done | 55.55% |

***Table 5: Percentage analysis of improvements: Table demonstrating a detailed analysis of various factors and the improvements***

| Symptoms improved | Cervical SCI | Thoracolumbar SCI |
|---|---|---|
| Spasticity | 18.37% | 26% |
| Sensation | 21.57% | 28% |
| Bladder Sensation | 20.59% | 33% |
| Bowel Sensation | 5.66% | 0.9% |
| Sitting Balance | 87.50% | 81.81% |
| Standing Balance | 48.21% | 66.36 |
| Trunk Stability | 92.31% | 95.65% |
| Postural Hypotension | 100.00% | 100% |

***Table 6: Comparison between Cervical SCI and Thoracolumbar SCI: Table comparing the outcome of Stem Cell Therapy in cervical SCI and Thoracolumbar SCI.***

**Intellectual Disability (ID):**

Worldwide not many studies have been published demonstrating efficacy of Stem Cell Therapy in ID. Sharma et al, have published world's first and only publication in "Stem Cell Research & Therapy" in 2018 which was an open-label proof-of-concept study including 58 patients of ID. These patients were divided into two groups: intervention group (n = 29) and rehabilitation group (n = 29). The intervention group underwent stem cell therapy and standard neurorehabilitation while, the rehabilitation group underwent only standard neurorehabilitation. The results of the symptomatic outcomes were compared between the two groups. The outcome measures used were the intelligence quotient (IQ) and the Wee Functional Independence Measure (Wee-FIM). To compare the pre-intervention and post-intervention results, statistical analysis was done using Wilcoxon's matched-pairs test for Wee-FIM scores and McNemar's test for symptomatic improvements and IQ. The effect of age and severity of the disorder were assessed for their impact on the outcome of intervention. A significantly better outcome of the intervention was found in the paediatric age group (<18 years) and patients with milder severity of ID. PET CT brain scan was used as a monitoring tool to study effects of the intervention. Repeat PET-CT scan in three patients of the intervention group showed improved metabolism in the frontal, parietal cortex, thalamus, mesial temporal structures and cerebellum. No major adverse events were witnessed.

On symptomatic analysis, greater improvements were seen in the intervention group as compared to the rehabilitation group. In the intervention group, the symptomatic improvements, IQ and Wee-FIM were statistically significant.




***Figure 33: Graph demonstrating a comparison of overall percentage improvements in ID between the intervention group and rehabilitation group***

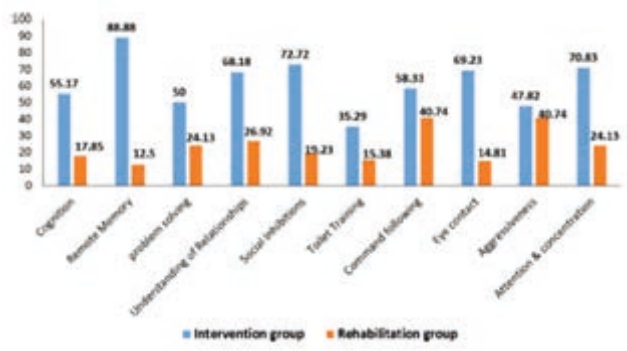



*Figure 34: Comparison of overall percentage improvements in the symptoms of ID between the intervention group and the rehabilitation group*

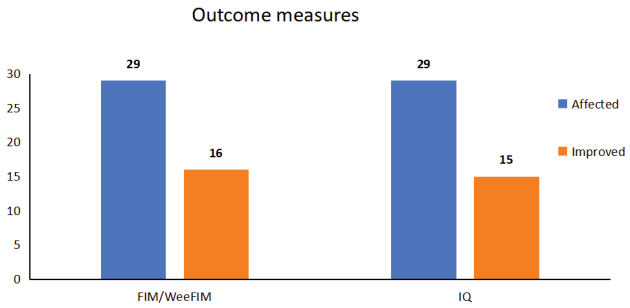



*Figure 35: Graph demonstrating improvements in the outcome measures (FIM/ WeeFIM and IQ) in patients with ID in the intervention group, six months after cellular therapy*

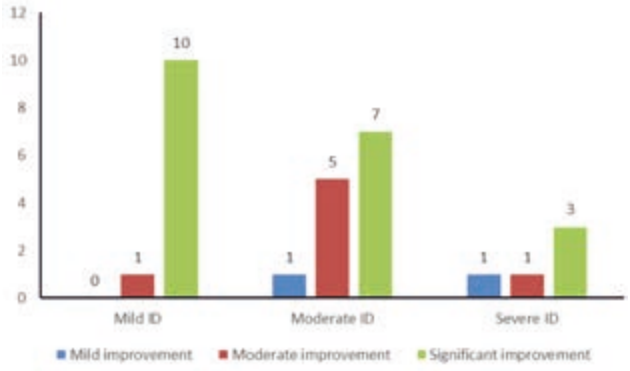



*Figure 36: Graph showing the comparison of improvement in patients in the intervention group with severity of ID.*

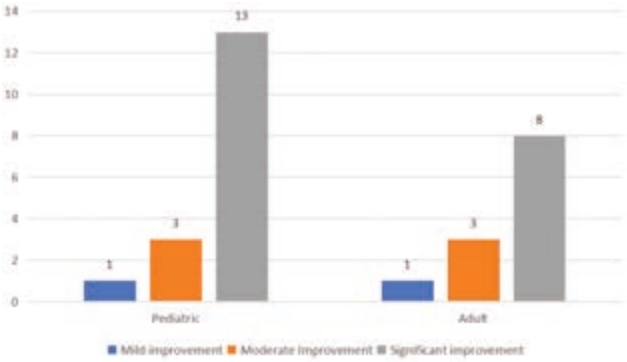



*Figure 37: Graph showing the comparison of improvement in patients in the pediatric and adult population of intervention group.*

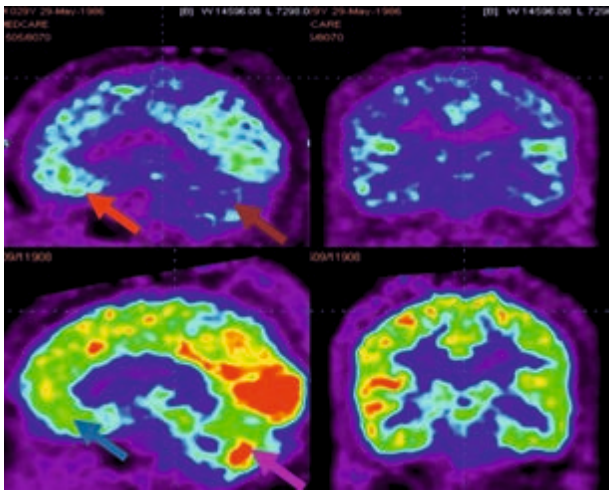


*Figure 38: The top row indicates the PET CT Scan image before stem cell therapy showing reduced metabolism in prefrontal, frontal (red arrows), cerebellum (brown arrow); Below row indicates improved 18F-FDG metabolism after intervention in the prefrontal, frontal (blue arrows), and cerebellum (pink arrow).*

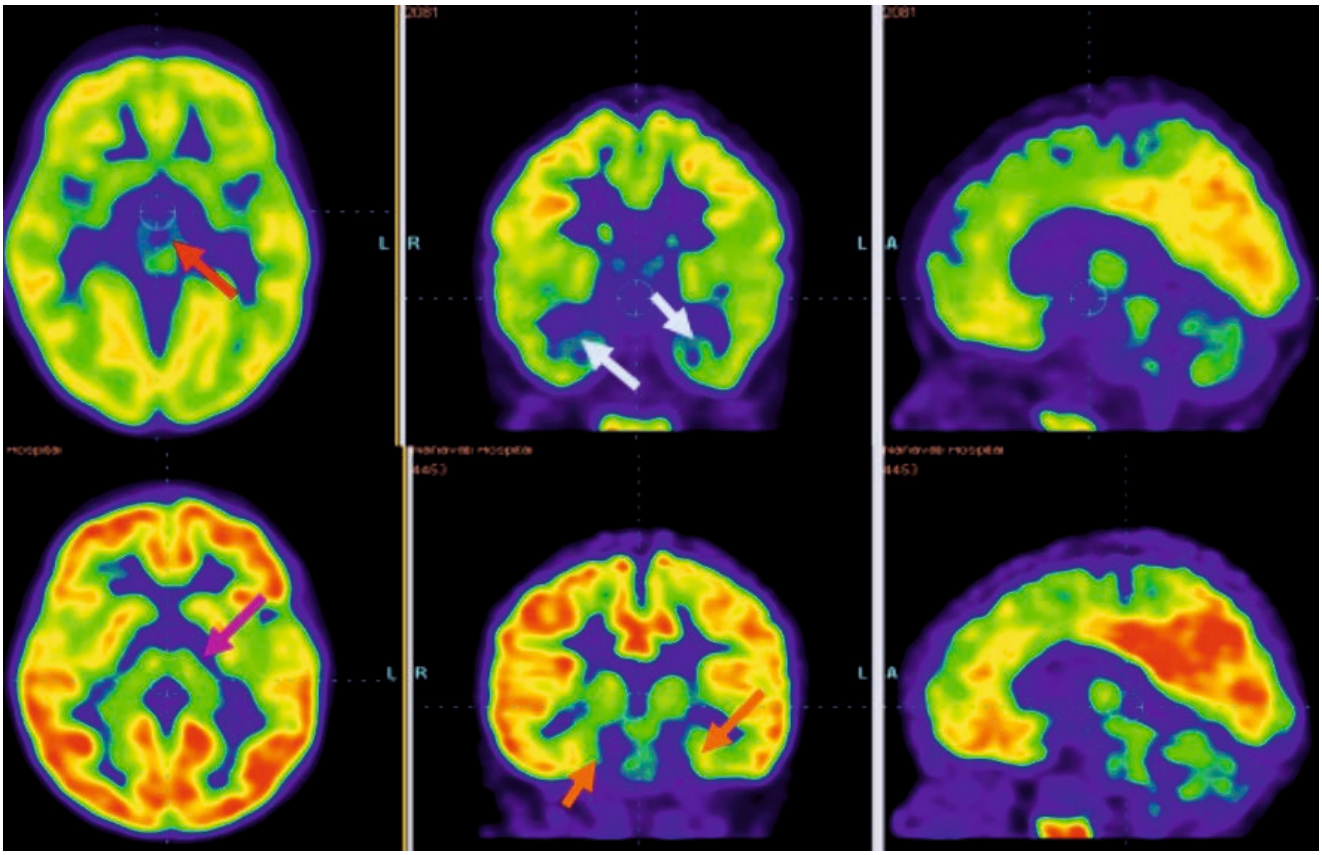


*Figure 39: The top row indicates the PET CT scan brain image before stem cell therapy showing reduced metabolism in thalamus (red arrow), mesial temporal structures (white arrow); Below row indicates improved 18F-FDG metabolism after therapy in thalamus (pink arrow), mesial temporal structures (orange arrow).*

**Traumatic Brain Injury (TBI):**

**Worldwide studies:**

Not many clinical studies have been conducted on human subjects of TBI showing effects of stem cell therapy. One of the first studies was published in 2008 by Zhang et al. They assessed the safety and feasibility of a combined procedure to deliver autologous mesenchymal stromal cells in 7 patients with traumatic brain injury. They found that neurological functions improved significantly at 6 months after Stem Cell Therapy. In 2011, Cox et al, performed a phase1 clinical trial to demonstrate the safety and feasibility of autologous BMMNCs in children with severe traumatic brain injury. 10 children were included in the study. Dichotomized Glasgow Outcome Score at 6 months showed 70% with good outcomes and 30% with moderate to severe disability. Followed by this, they published the results of their phase 2 clinical trial in 2017 which included 25 patients; 15 in the intervention group and 10 in the control group. They recorded positive functional and neurological outcome and also down regulated Inflammatory biomarkers. Similar studies were published by Sharma et al and Liao et al in 2015 demonstrating the positive effect of BMMNCs. Wang et al, in 2013 conducted a study on patients with sequelae of TBI. They administered 40 patients with umbilical cord mesenchymal stem cells and observed improved neurological functions and self-care in these patients as compared to the controls. Vaquero et al in 2017, showed improvement and progressive increase in brain glucose metabolism after intrathecal

administration of autologous mesenchymal stromal cells using 18 FDG PET CT, in 3 patients. In 2017, Wang Z et al demonstrated the safety and feasibility of neural stem cell transplantation in patients with severe TBI. They observed that majority of patients experienced improved neurological function on follow up along with increased serum levels of nerve growth factor and brain-derived neurotrophic factor.

**Published Study**

A pilot study was published in "SpringerPlus" in 2015 which was conducted on 14 chronic TBI patients. These Patients underwent stem cell therapy and were regularly followed up at 1 week, 3 months and 6 months. The outcome measures utilized were Functional Independence Measure (FIM) scale, SF-8 Health Survey Scoring and Disability rating scale. At the end of 6 months, these scales showed a positive shift in scores. Improvements were observed in various symptoms along with activities of daily living. Improvement in PET CT scan performed before and 6 months after the intervention in 3 patients corresponded to the clinical and functional improvements observed in these patients. The results of this study suggest that stem cell therapy may promote functional recovery leading to an improved quality of life in patients suffering from chronic TBI.

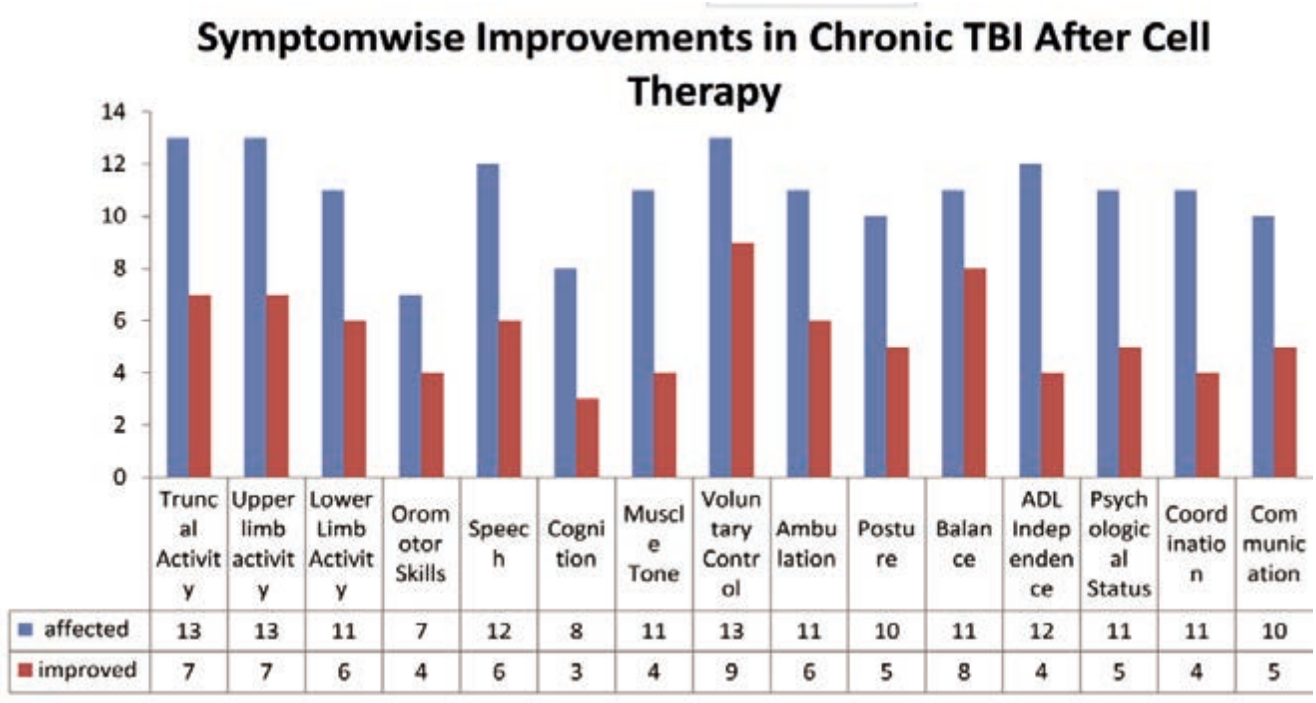



*Figure 40: Symptomatic improvements after Stem Cell Therapy*

| Patient | Areas of the brain | Functions improved |
|---|---|---|
| Patient 1 | Parieto-occipital areas | cognition, speech, sensation, orientation and visual perception |
| Patient 2 | Cingulate gyri | Emotion, attention, cognition, memory |
| | Amygdala | Emotional responses, memory, attention |
| | Frontal | Planning, Long term memory, emotions, speech, problem solving |
| | Temporal lobes | Speech, memory |

| | | |
|---|---|---|
| Patient 3 | Amygdala | Emotional responses, memory, attention |
| | Cerebellum | Coordination, balance |
| | Cingulate gyri | Emotion, attention, cognition, memory |
| | Basal ganglia | Voluntary motor control, learning,cognition. |
| | Occipital lobes | Vision and perception |
| | Parietal lobe | Movement, orientation |
| | Temporal lobes | Speech, memory |
| | Frontal lobes | Planning, Long term memory, emotions, speech, problem solving |
| | Thalamus | Motor control, sensory functions |

***Table 7: Areas of the brain showing improved metabolism in PET CT scan and their correlation to the clinical function improvement***

**Motor Neuron Disease (MND) / Amyotrophic Lateral Sclerosis**

In a study which published a Phase I clinical trial, human spinal cord-derived neural stem cells were transplanted into the spinal cord of 12 late- to mid-stage ALS patients. There were no long-term surgical complications and importantly, ALS patients tolerated the procedure, gave no indications that the stem cells were injurious to the spinal cord and showed no disease acceleration due to injections. Second part of this Phase 1 trial determined the safety of injecting cells into the C3–C5 cervical region of the spinal cord. Three new ALS patients and three patients that had previously received lumbar injections received human spinal cord-derived neural stem cells. There were some positive effects including slowing of rate of progression, and one patient even showed improvement in clinical status. In an expansion of the study, Mazzini et al. transplanted human fetal brain tissues into the anterior horns of the spinal cord and additionally used a much higher cell dosage. The authors concluded that the procedure was well tolerated and safe

In 2003, Mazzini and colleagues injected variable numbers of mesenchymal stem cells (MSCs) (7–152 × 106 cells) into the thoracic spinal cord of seven ALS patients. Although there was no functional improvement, the transplantation of these cell suspensions into the human ALS spinal cord was safe and well tolerated. Their follow-up studies, lasting more than 4 years post-surgery, showed no signs of toxicity or abnormal cell growth. Four patients showed a significant slowing down of the linear decline of the forced vital capacity and of the ALS-FRS score. A phase I/II clinical trial by Karussis and colleagues showed that intrathecal and intravenous administration of autologous bone-marrow-derived MSCs into ALS patients is feasible and safe. Although this study was not designed to detect therapeutic efficacy, encouragingly, it induced immediate immunomodulatory effects in ALS patient and ALSFRS scores remained stable for up to 6 months following treatment.

Although allogeneic hematopoietic stem cells (HSCs) transplanted intravenously in six ALS patients did not provide any clinical benefit, donor-derived cells were found to localize at the sites of pathology, rendering these cells to be particularly suitable for delivering therapeutic molecules.

Olfactory ensheathing cells extracted from human fetal olfactory bulbs were injected into the bilateral corona radiata in 15 ALS patients who were compared to 20 untreated controls. Over a 4-month follow-up period,

positive and beneficial results were observed as a five-point difference in the ALSFRS-R. In another study, 327 patients received injection of olfactory ensheathing cells into the spinal cord, the bilateral corona radiata, or both. They reported improved ALS functioning rating scale and normalized electromyographical findings 4 weeks after transplantation, with no differences between the three groups.

Studies have demonstrated safety of intraspinal infusion of autologous bone marrow mononuclear cells. Deda et al, demonstrated reinnervation in 7 of 13 patients on electro neuro myography following intraspinal infusion of autologous bone marrow mononuclear cells, 1-year post treatment. Clinical improvements were seen in 10 patients. Intrathecal transplantation of autologous bone marrow mononuclear cells was found to be safe and effective in slowing down disease progression in 5 studies.

**Our Published Data:**

In 2015, Sharma et al published a retrospective controlled cohort study of 57 patients. Out of these, 37 patients underwent autologous bone marrow mononuclear cell transplantation in addition to standard rehabilitation and Riluzole. Control group consisted of 20 patients who did not receive cell transplantation. Survival duration was computed using a Kaplan-Meier Survival analysis and compared using log-rank test. Various Factors such as effect of age at onset, type of onset and lithium on survival duration in the intervention group was analyzed. Mean survival duration of patients in intervention group was 87.76 months which was higher than the control group mean survival duration of 57.38 months. Patients with the onset of the disease below 50 years of age showed significantly higher change in survival duration (p=0.039) (Figure 38). Limb onset and lithium also showed positive influence on the survival duration (Figure 39,40).

*Table 7: Survival analysis*

| Survival analysis | Intervention group | Control group |
|---|---|---|
| Total mortality | 48.64% | 50.00% |
| Range of survival duration (months) | 13 – 158 | 26-84 |
| Mean survival duration (months) | 87.76 (10.45) | 57.38(5.31) |

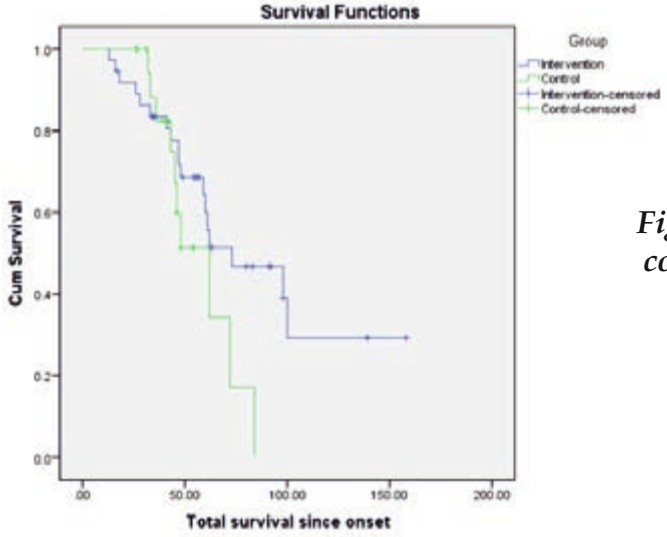



*Figure 41: Kaplan Meier survival analysis comparing the mean survival duration of the intervention and control group*

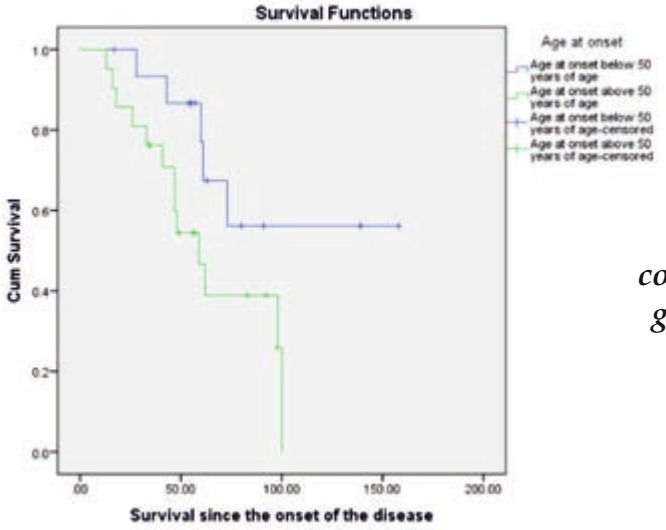



*Figure 42: Kaplan-Meier Survival comparison for the patients in intervention group below and above 50 years of age at the onset.*

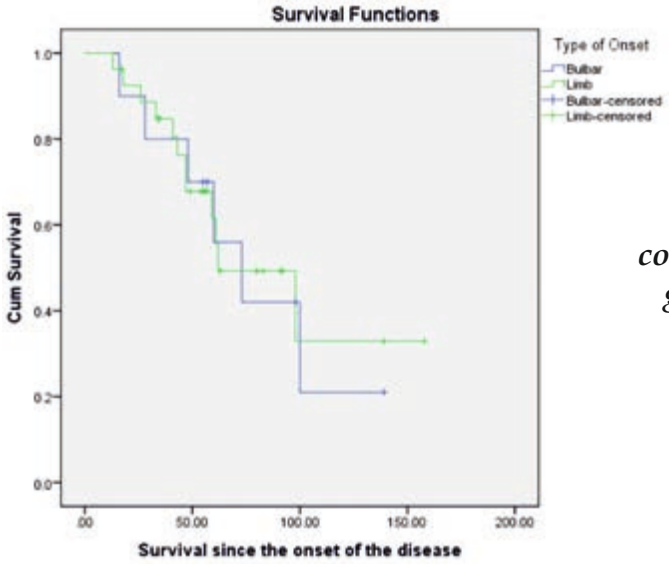



*Figure 43: Kaplan-Meier Survival comparison for the patients in intervention group with bulbar onset and limb onset*

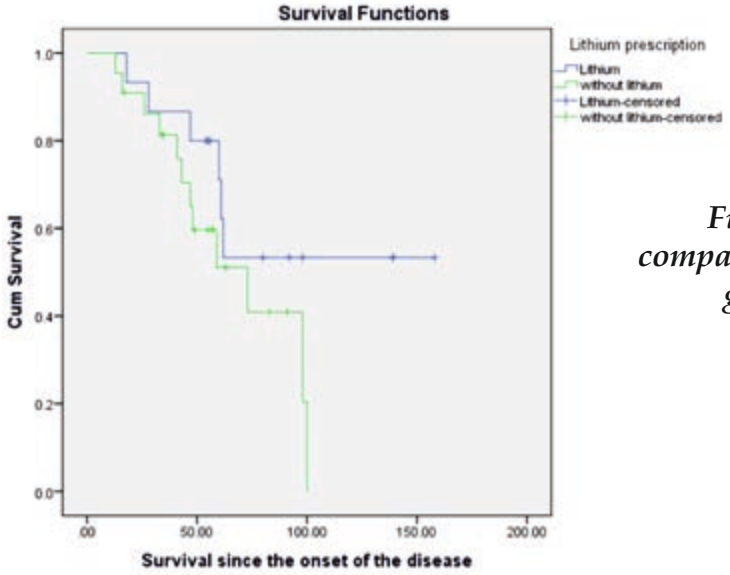



*Figure 44: Kaplan-Meier Survival comparison for the patients in intervention group with or without Lithium*

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# Chp. 29 Role of Neuro-Rehabilitation in Stem Cell Therapy

**Dr. Alok Sharma, Dr. Prerna Badhe, Dr. V. C. Jacob, Dr. Hema Biju, Dr. Nandini Gokulchandran, Dr. Hemangi Sane, Dr. Amruta Paranjape**

Neuroplasticity is defined as brain's ability to adapt or use cellular adaptations to learn or relearn functions which are previously lost as result of cellular death by trauma or disease at any age. Neuronal sprouting is thought to be primary mechanism, allowing injured neurons, to reconnect in new ways and allowing intact undamaged neurons to form new connection and enhance function. Motor learning will continue throughout life as long as environment asks for change and CNS has pliability and desire to learn. The rehabilitation team promotes this learning and facilitates neural plasticity. The philosophic foundation of rehabilitation team is to promote purposeful activity thereby preventing dysfunction and eliciting maximum adaptation. These goal-oriented activities are meant to be culturally meaningful and important to the needs of patient and their families. Activities include daily life and work skills, exercise, recreation and crafts.

Exercise tasks in animal models; have shown that specifically skilled type of exercises lead to increased angiogenesis in damaged cortical areas whereas unskilled activities did not show this positive change. It is believed that in humans too rehabilitation techniques would enhance neuroplastic changes.

Various studies have suggested that regular physical activity and exercise leads to neoangiogenesis, anti-inflammatory effect on the local as well as global systemic environment, anti apoptotic effects and effects modulating the immune system. But the most important advantage of exercises is the mobilization of the local stem cells for repair and regeneration of the tissue.

Exercise also secretes various growth factors that stimulate the resident stem cells to grow into the tissue cells. It is therefore of utmost importance that stem cell therapy should be followed by the rigorous rehabilitation (3).

**NeuroRehabilitation as an adjunct with stem cell therapy**

The concept of combining rehabilitation with regenerative medicine promotes a multidisciplinary and holistic approach to bring about recovery of neural function with a close integration of Neuro regenerative (including stem cell therapy), Neuro protective (medications) and neurorehabilitative therapies (physical / occupational / speech / psychology / aquatic ). Thus, it combines the best neurobiological repair technologies and neurorestorative techniques. The rehabilitation protocol is then individualized to the specific requirements of each patient emphasizing on functional recovery.

Survival of the transplanted stem cells forms the basis for successful stem cell transplantation. Externally applied motor or sensory stimuli are said to promote the proliferation and survival of stem cells. Attention to integrating cell based therapies with rehabilitation was first introduced by Fabrisia Ambrosio. Muscle derived stem cells were transplanted into the injured hind limbs of mice. The mice in the experimental group were made to run on the treadmill for

five weeks. The active mice developed more new muscle fibers as compared to the sedentary controls. Exercise stimulates the transplanted stem cells and the local stem cell pool to secrete neurotrophins which in turn can promote plasticity through their impact on dendrites and increase in vascularity. Exercise also impacts the synaptic and receptor protein expression. A long term, low-intensitystrengthening and low-load weight-bearing exercise program may cause a shift in type II fibers to type I muscle fibers which are less prone to degeneration in muscular dystrophy. Physical rehabilitative exercises also promote secretion of vascular endothelial growth factor (VEGF) which is known to stimulate satellite cell proliferation and migration thus promoting muscle fiber regeneration. Exercise causes intense muscular activities and metabolic changes in peripheral organs which in turn cause an increase in the peripheral production of insulin-like growth factor-1 (IGF-1). The IGF-1 can reach the CNS by crossing the blood-cerebrospinal fluid barrier. The IGF-1 has various benefits including neural cell and oligodendrocytedifferentiation and in cognitive functions. Exercise also helps improve respiratory and cardiac functions which are affected in many of the neurological and neuromuscular disorders and require attention.

Studies have shown that exercise enhances the effect of injected stem cells by inducing mobility in the injected stem cells, by activation and proliferation of the local stem cells, muscle angiogenesis and release of cytokines and nerve growth factors, thereby enhancing the achievable outcomes. Hence, neurorehabilitation appears to work complimentarily with stem cells therapy.

In ischemic or traumatic disorders of central nervous and spine, regenerative medicine opens the window of neural recovery and rehabilitation helps to maximize the potential for recovery. Once the tissue is recovered it is important to unmask the old pathways and abandon the new compensatory pathways that have been learnt by the brain. Therefore to translate the cellular and tissue level changes brought about by stem cell therapy into function rehabilitation is important. It is important to perform rigorous rehabilitation as repetition is the key. It requires nearly 500 repetitions per day of a particular skill for at least 7-8 days so that a neural pattern can be imprinted. In progressive conditions, exercises helps to promote survival of transplanted cells and recruitment of local cells preventing the rapid loss of tissue / function as is usually seen in these disorders.

Rehabilitation also prevents secondary complications of the disease. The secondary complications like cardiovascular disorders are known to have a negative effect on brain recovery. Whereas, other complications like fibrosis, bed sores, contractures etc. can change tissue quality and hinder movement as well representation of the affected areas in the brain. In progressive conditions these secondary complications can develop have fatal consequences. Rehabilitation prevents this and facilitates recovery.

Rehabilitation also has a positive psychological effect. Socialization associated with rehabilitation helps patients to be engaged in the recovery process actively.

It is crucial that the rehabilitation is started early after transplantation, it is rigorous in the intensity and duration, and it is performed under professional supervision. This ensure optimum outcome after stem cell therapy.

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At KLS Wellness Institute modern medical technology is available that helps in slowing down and reversing the ageing process. Oxygen supply to the tissues is enhanced by using Hyperbaric Oxygen Therapy and Ozone Therapy. Circulation to tissues is improved by Enhanced External Counter Pulsation (EECP) and by Cyrotherapy. Detoxification of organs is done by using Chelation therapy and Antioxidants. Rejuvenation and repair of tissue damage is done by Cellular therapy and Hormone therapy. Weight loss is achieved with the help of Cyrolipolysis and Cool Sculpting. Other treatment modalities like Infrared, laser, hydrotherapy etc help in reducing the discomforts of age-related disorders. When these modern technologies are combined with the traditional systems of medicine like Yoga and Naturopathy, they help in revitalizing and rejuvenating the body & mind.

The Integrative therapies at KLS Wellness institute help to control and reverse Diabetes Mellitus. The Weight loss program at KLS Wellness helps in target driven weight loss and body contouring. Cardiac patients who are unfit or unwilling for invasive cardiac treatments are helped with the supportive cardiac care. Women going through Menopause are given symptomatic relief through the Menopause Management Program. The progression of Osteoarthritis and spondylosis is halted and the patients given symptomatic relief. Psychosomatic stress is relieved holistically. Thyroid and endocrine imbalances are corrected. Advanced Cosmetology procedures assist in enhancing skin health and appearance. Both Male & Female Sexual dysfunction is corrected through a multi pronged approach. Patients with Neurological disorders who have already exhausted all treatment options get benefit from the comprehensive integrative therapies.

The scientific benefits of these treatments on a cellular level for reversing the ageing process have been shown in various scientific medical publications. What makes these treatments appealing is that they are non-invasive, very safe and with no major side effects.

# NeuroGen Brain and Spine Institute

Centre for Cell Therapy and Neurorehabilitation



NeuroGen Brain & Spine Institute in Navi Mumbai, India is an International center of excellence for Neurological disorders which has been set up specifically to treat incurable neurological conditions through a combination of Cell Therapy and comprehensive NeuroRehabilitation. It was founded on 17th December 2008, by renowned Neurosurgeon, Neuroscientist and Professor Dr. Alok Sharma.

A unique aspect of the treatment Neurogen offers is a comprehensive treatment protocol for Autism, Cerebral Palsy and other Neurodevelopmental disorders. This includes a detailed clinical and Neuro - radiological evaluation, followed by cell therapy that repairs the brain's malfunctioning along with a comprehensive rehabilitation program which include Physiotherapy, Occupational Therapy, Speech therapy & ABA Therapy etc. With this holistic care it has been possible to reverse Autism and give relief to Cerebral Palsy Children in a manner that was not possible earlier. In fatal neurological conditions like Muscular Dystrophy and Motor neuron disease, Neurogen has saved hundreds of lives using Cell therapy. Paralysis due to Stroke, Spinal cord injury and Head Injury has also been benefited through the innovative treatment protocol.

Since, its inception in 2008, NeuroGen has successfully treated 10000+ patients from 70+ different countries. The institute have been accredited by ISO 9001-2015 and a GLP (Good laboratory practice) and GMP (Good Manufacturing Practice) certified Cell laboratory. At NeuroGen, along with treatment there is a strong emphasis on clinical research. The clinical results of this work are documented in several international and national scientific papers and chapters in medical textbook. The world's first scientific paper on the role of cell therapy in Autism has been published by Neurogen in the journal "Stem Cell international". A detailed analysis of the clinical data is carried out on a regular basis and this is published in various scientific journals. Currently, 100+ scientific papers have been published by our team in various international and national peer reviewed medical journals. We also have 10+ chapters in International textbooks and our team has also published 20+ books on various neurological conditions for parents/caretakers as well as professionals.

NeuroGen also has received 15+ National and International awards like "The Best Stem Cell Therapy Centre in India" by National Healthcare Excellence in 2016, "The Rose of Paracelsus Award" by European Medical Association in 2016. "The Best Stem Cell therapy center in Maharashtra " by Navbharat Healthcare Excellence in 2017, "European Awards for best Practices 2018" by European society of quality research in 2018 and many more. NeuroGen has tied up with leading international organizations from America, Africa, United Kingdom, Middle east and other countries for research and treatment collaborations. Cutting edge research, pioneering new treatments, the best medical professionals, comprehensive treatment facilities all under one roof and a caring holistic approach makes the NeuroGen Brain and Spine institute a unique and special facility for patients with Neurological problems.

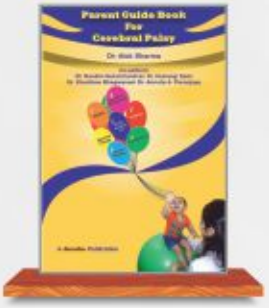
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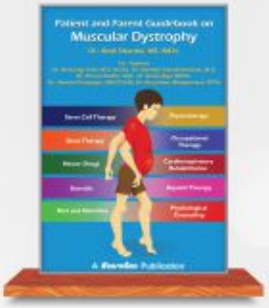
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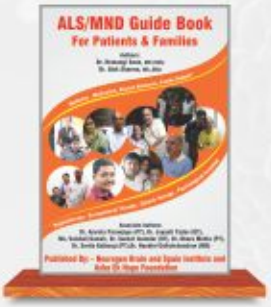

પેશન્ટ અને પેરન્ટ માર્ગદર્શિકા મસ્ક્યુલર ડિસ્ટ્રોફી વિશે



Looking after children with autism - A handbook



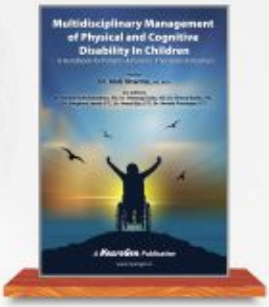

ALS / MND Guide Book For Patients & Families




A Handbook on Neurological Disorders for Special Educators




Multidisciplinary Management of Physical and Cognitive Disability In Children

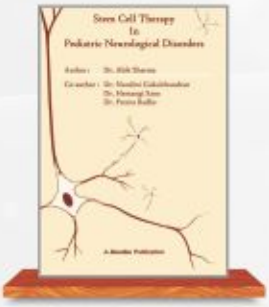



## Other Medical Books

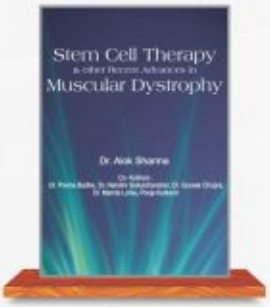
Stem Cell Therapy in Neurological Disorders 4th Edition



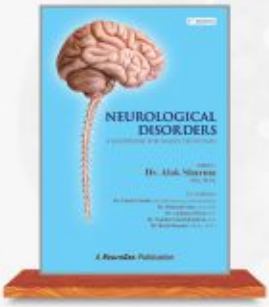

Stem Cell Therapy In Pediatric Neurological Disorders 2nd Edition




Stem Cell Therapy & Other Recent Advances in Muscular Dystrophy




Neurological Disorders A Handbook for Family Physicians 2nd Edition




**All the books can be downloaded free of cost from www.neurogen.in**

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