

# Multidisciplinary Management of Physical and Cognitive Disability In Children

- A Handbook for Patients & Parents , Therapists & Teachers

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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).

Published by  
**NeuroGen Brain & Spine Institute**

Cover Designed by  
**Dr. Khushboo Bhagawanani**  
**Satish Narayan**

Printed by  
**SUREKHA PRESS**  
A-20, Shalimar Industrial Estate,  
Matunga Labour Camp, Mumbai 400 019.  
Email: surekhapress@gmail.com

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## Grateful Acknowledgments

Dr. V. C. Jacob, Dr. Prerna Badhe, Dr. Joji Joseph, Dr. Hema Biju, Mrs. Vibhuti Bhatt, Dr. Monali Dhanrale, Dr. Hema Sriram, Dr. Richa Bansod, Dr. Jayanti Bhate, Ms. Pooja Kulkarni, Dr. Shruti Shirke, Dr. Jasbinder Saini, Dr. Jidnyasa Koli, Dr. Niranjana Kophrekar, Dr. Jyotsna Gaikwad, Dr. Karishma Rajpal, Dr. Sonali Parmar, Ms. Ridhima Sharma, Ms. Ashly Joseph, Ms. Sonali Nalawade, Mr. John Julius, Dr. Snehal Sontate, Dr. Sushil Kasekar, Dr. Reena Jain, Dr. Kirti Lad, Dr. Abhishek Gupta, Dr. Shailesh Patil, Dr. Shruti Pasi, Dr. Lata Thakur, Dr. Amol Salagre, Dr. Hetal Mori, Dr. Ajaz Khan, Mrs. Ritu Vargese, Mr. Sam Nivins, Ms. Alitta Jose, Ms. Nupur Jha, Ms. Shreya Madali, Ms. Monica Chugh, Ms. Monica Vacchani, Mrs. Hridhika Rajesh, Ms. Larissa Monteiro, Ms. Sharon Kinkar, Dr. Aarushi Bhaumik, Dr. Ummeammara RK, Mr. Aditya Nagaraja, Dr. Umami Khanbande, Mr. Bryan Pinto, Mr. Mandar Thakur, Ms. Monica Vachhani, Mrs. Geeta Arora, Mrs. Kanchan Patil, Mr. Kiran Pawar, Mr. Rajendra Patole, Mrs. Manjula Shete, Mrs. Daisy Devassy, Mr. Sumedh Kedare, Mrs. Yasmeen Shaikh, Mr. Dinesh Desalae, Mr. Abhishek Patil, Mrs. Mani Nair, Mrs. Chandra Bhatt, Mr. Udaykant Mishra, Ms. Anjali Avalusa, Mrs. Amruta Kadam, Mr. Satish Pol, Mr. Roshan Salanki, Mr. Vikram Chikne, Ms. Bhargavi Mamidi, Ms. Minal Vedante, Ms. Jigyasa Chabbria, Ms. Namrata Katke, Mr. Roshan Solanki, Mr. Savio Agviour, Mr. Rohit Padale, Mr. Kevin More, Ms. Sneha Khopkar, Mr. Rohan Awate, Ms. Neeta Thakur, Ms. Prajakta Bhoir, Ms. Nanda Mane, Ms. Shweta Sathe, Mr. Satyavan More



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देवेन्द्र फडणवीस  
मुख्य मंत्री  
महाराष्ट्र



मंत्रालय,  
मुंबई - ४०० ०३२

2.63% of the population of Maharashtra is affected with various forms of physical and intellectual disabilities. These affect not just the patient but put a huge medical and socioeconomic burden on the entire family. It is our responsibility as the government to do all we can to assist such families by ensuring that they have access to the best medical care, appropriate education, suitable jobs and barrier free facilities in public spaces. Our Honourable Prime Minister Shri. Narendra Modi has launched 'Sugamya Bharat Campaign' two years ago to make the physical as well as virtual infrastructure accessible and easy to use for persons with disabilities. He also changed the term 'Persons with disability' i.e. 'Viklang' to 'Persons with divine abilities' i.e. 'Divyang'; to change the attitude towards disability. Under his leadership we have also passed an act 'Rights of Persons with Disabilities Act, 2016'; which entitles and lays specific responsibilities on the appropriate governments to ensure basic rights for persons with disability. The most important part is the act has created a facility for grievance redressal so if these rights are denied our 'Divyang' brothers and sisters can directly contact the government officials and a necessary action will be taken to meet their rights.

We also need to invest our efforts in finding improved medical solutions for the illnesses and to create awareness amongst the families about the need and availability of these treatments. The availability of a simple and easy to understand book would be an important means of creating this awareness.

I would like to thank the Author of this book Dr. Alok Sharma and his team for making such a book available not just in English but also in Marathi so that all the ordinary citizens of Maharashtra can read it. I want to thank him as well as all the other doctors and rehabilitation therapists of Maharashtra who are working hard on a day to day basis to care for people with disabilities. I take immense pride in saying that Maharashtra offers some of the most recent treatments like advanced prosthesis and assistive devices, newer corrective and reconstructive surgeries as well as stem cell therapy etc. to these patients; not only in private but also in the government hospitals so that even the poor patients can benefit. I would like to appreciate 'NeuroGen Brain and Spine Institute' and the work they are doing for our 'Divyang' children. As Chief minister of Maharashtra I assure every family having a 'Divyang' family member that the government will stand strongly with you through all your efforts of overcoming the disability.

देवेन्द्र फडणवीस

Shri. Devendra Fadnavis,  
Chief Minister, Maharashtra

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दूरध्वनी : ०२२-२२०२५१५१ / २२०२५२२२, फॅक्स : ०२२-२२०२९२१४,  
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## Preface

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Children with physical and mental disabilities form weakest, most vulnerable and dependent segment of society. Because they can't live independently and in most cases, cannot have normal education. They need support of Family and society to be able to perform normal activities of life. 2.1% of the population in India consists of persons with disabilities. In Maharashtra 23,60,000 people could be estimated to have various disabilities.

Having a special needs child puts a tremendous financial, social & medical strain on the entire family. From those coming from the low socio-economic background & those from rural areas who do not have access to specialized medical and rehabilitative help, this burden can be devastating.

Government of India passed a land mark bill, 'The rights of persons with disabilities bill' in 2016 which offers huge support and many rights to children and adults with disabilities. We believe that empowered and informed parents and caregivers can make a big difference to the lives of these children. This book is to empower the parents and care givers of children with disability. The book provides information about comprehensive management of physical and cognitive disabilities caused due to various neurological disorders. Disability is dynamic and not a personal attribute. Management of disability therefore requires a multidisciplinary management.

Multidisciplinary team consists of Physiotherapist, Occupational therapists, Speech therapist, Aquatic therapist, Clinical psychologist, Special educators, and Nutritionist along with different medical professionals. The book also empowers caregivers about their legal rights.

The NeuroGen brain and Spine Institute has been created to offer the best medical & rehabilitation treatments from the field of neurosciences & Regenerative Medical.

Stem Cell Therapy is a newly evolved treatment which can help many children with disabilities, especially those with Autism, Cerebral Palsy, Mental Retardation, Down Syndrome, Duchene Muscular Dystrophy and Other neurological disorders recover from their disabilities. Using stem cell therapy many wheelchair bound children of Cerebral Palsy and Duchene Muscular Dystrophy are now able to walk and many children with Autism and Intellectual disability are able to go back into the education system. India is a leader in the field of Stem cell therapy.

This book is being printed in both English & Marathi so that all the people in the state of Maharashtra can benefit from the knowledge and information shared in this book.

## Scientific Publications on Pediatric Neurodevelopmental Disorders

### AUTISM:

1. Alok Sharma, Nandini Gokulchandran, Hemangi Sane, Anjana Nagrajan, Amruta Paranjape, Pooja Kulkarni, Akshata Shetty, Priti Mishra, Mrudula Kali, Hema Biju, Prerna Badhe. Autologous bone marrow mononuclear cell therapy for autism – an open label proof of concept study. **Stem cell international**. 2013 (2013), Article ID 623875, 13 pages.
2. Alok Sharma, Nandini Gokulchandran, Prerna Badhe, Pooja Kulkarni, Priti Mishra, Akshata Shetty and Hemangi Sane. An Improved Case of Autism as Revealed by PET CT Scan in Patient Transplanted with Autologous Bone Marrow Derived Mononuclear Cells. **J Stem Cell Res Ther** 2013, 3:2.
3. Alok Sharma, Nandini Gokulchandran, Akshata Shetty, Hemangi Sane, Pooja Kulkarni and Prerna Badhe. Autologous Bone Marrow Mononuclear Cells may be Explored as a Novel. Potential Therapeutic Option for Autism. **J Clin Case Rep** 2013, 3:7.
4. Alok Sharma, Nandini Gokulchandran, Hemangi Sane, Pooja Kulkarni, Nancy Thomas, Amruta Paranjape, Prerna Badhe. Intrathecal autologous bone marrow mononuclear cell transplantation in a case of adult autism. **Autism open access**. 2013, 3:2.
5. Alok Sharma, Nandini Gokulchandran, Hemangi Sane, Pradnya Bhovad, Hema Biju, Akshata Shetty, Mrudula Kali and Prerna Badhe. Cell therapy effects portrayed on positron emission tomography computerized tomography scan of the brain serve as a new dimension for autism: A case report (2014), **Journal of Paediatric Neurology**, 12:3.
6. Sharma A , Gokulchandran N , Shetty A , Kulkarni P, Sane H , Badhe P. Neuropsychiatric Disorder Tackled by Innovative Cell Therapy-A Case Report in Autism. **J Stem Cell Res Transplant**. 2014;1(1): 4.
7. Alok Sharma, Guneet Chopra, Nandini Gokulchandran, Mamta Lohia, Pooja Kulkarni. Autologous Bone Derived Mononuclear Transplantation in Rett Syndrome. **Asian Journal of Paediatric Practice**. 2011; 15 (1): 22-24.

8. Alok Sharma, Nandini Gokulchandran, Hemangi Sane, Avantika Patil, Akshata Shetty, Hema Biju, Pooja Kulkarni, Prerna Badhe. Amelioration of Autism by Autologous Bone Marrow Mononuclear Cells and Neurorehabilitation: A Case Report. **American Journal of Medical Case Reports**, 2015, Vol. 3, No. 10, 304-309
9. Alok Sharma, Hemangi Sane, Nandini Gokulchandran, Prerna Badhe, Avantika Patil, Pooja Kulkarni, Amruta Paranjape PET- CT scan shows decreased severity of Autism after autologous cellular therapy: A case report. **Autism Open Access** 2016; 6:169.
10. Alok Sharma, Hemangi Sane, Nandini Gokulchandran, Prerna Badhe, Pooja Kulkarni and Suhasini Pai. Stem Cell Therapy in Autism Spectrum Disorders. Recent Advances in Autism. **SMGroup**. (In Press)
11. Alok Sharma Nandini Gokulchandran , Hemangi Sane , Pooja Kulkarni , Suhasini Pai , Vaishali Ganwir ,Prerna Badhe. A case of autism showing clinical improvements after cellular therapy along with PET CT evidence. **Journal of Stem Cell Research & Therapeutics**. April 2017
12. Alok Sharma, Nandini Gokulchandran, Pooja Kulkarni, Sarita Kalburgi, Shruti Kamat, Riddhima Sharma, Samson Nivins, Hemangi Sane, Prerna Badhe. "Improvements in a case of autism spectrum disorder after cell therapy as noted on PET CT brain scan" **SJSC**. May 2017
13. Alok Sharma, Nandini Gokulchandran, Hemangi Sane, Samson Nivins, Amruta Paranjape, Prerna Badhe. The baseline pattern and age-related developmental metabolic changes in the brain of children with autism as measured on positron emission tomography/computed tomography scan. **World J Nucl Med** 2018;17 (In Press)
14. Alok Sharma, Nandini Gokulchandran, Hemangi Sane, Pooja Kulkarni, Sarita Kalburgi, Riddhima Sharma, Prerna Badhe, Samson Nivins. PET CT Scan Brain As A Monitoring Tool To Study The Effects Of Autologous Bone Marrow Mononuclear Cell Transplantation In Autism Spectrum Disorder. **International Journal of Current Advanced Research**. Sep 2017 (In Press)

## INTELLECTUAL DISABILITY

1. Sharma A, Sane H, Pooja K, Akshya N, Nandini G, Akshata S. (2015) Cellular Therapy, a Novel Treatment Option for Intellectual Disability: A Case Report. **J Clin Case Rep** 5:483. doi: 10.4172/2165-7920.1000483.
2. Sharma A, Gokulchandran N, Sane H, Pai S, Kulkarni P, et al. Cognitive Changes after Cellular Therapy in a Case of Intellectual Disability. **J Transplant Stem Cel Biol.** 2017;4(1): 4.

## CEREBRAL PALSY:

1. Alok Sharma, Hemangi Sane, Nandini Gokulchandran, Pooja Kulkarni, Sushant Gandhi, Jyothi Sundaram, Amruta Paranjape, Akshata Shetty, Khushboo Bhagawanani, Hema Biju and Prerna Badhe. A clinical study of autologous bone marrow mononuclear cells for cerebral palsy patients: a new frontier," **Stem Cells International**, Volume 2015, Article ID 905874, 11 pages.
2. Alok Sharma, Hemangi Sane, Amruta Paranjape, Nandini Gokulchandran, Pooja Kulkarni and Anjana Nagrajan, Prerna Badhe. Positron Emission Tomography - Computer Tomography scan used as a monitoring tool following cellular therapy in Cerebral Palsy and Mental Retardation - A Case Report. **Case Reports in Neurological Medicine.** Volume 2013, Article ID 141983, 6 pages.
3. Alok Sharma, Hemangi Sane, Nandini Gokulchandran, Prerna Badhe, Pooja Kulkarni and Amruta Paranjape. Stem Cell Therapy for Cerebral Palsy - A Novel Option. **Cerebral Palsy. Challenges for the future.** 2014: 217-242.
4. Alok Sharma, Hemangi Sane, Pooja Kulkarni, Myola D'sa, Nandini Gokulchandran, Prerna Badhe. Improved Quality of Life in a Case of Cerebral Palsy after bone marrow mononuclear cell transplantation. **Cell J.** 2015; 17(2): 389-394.
5. Dr. Alok Sharma, Ms. Pooja Kulkarni, Dr. Hemangi Sane, Dr. Nandini Gokulchandran, Dr. Prerna Badhe, Dr. Mamta Lohia, Dr. Priti Mishra. Positron Emission Tomography- Computed Tomography scan captures the effects of cellular therapy in a case of cerebral palsy. **Journal of clinical case reports.** 2012 J Clin Case Rep 2:195.
6. Alok Sharma, Tongchao Geng, Hemangi Sane, Pooja Kulkarni. Clinical neurorestorative progresses in cerebral palsy. **Journal of Neuro-restoratology** 2016;4; 1-7

7. Alok Sharma, Hemangi Sane, Suhasini Pai, Pooja Kulkarni, Meenakshi Raichur , Sarita Kalburgi, Sanket Inamdar, Nandini Gokulchandran, Prerna Badhe. Intrathecal administration of autologous bone marrow mononuclear cells in a case of Cerebral Palsy coexisting with autistic features". **Phys Med Rehabil Int.** 2017; 4(1): 1110.
8. Sharma A, Sane H, Kalburgi S, Kulkarni P, Bhagwanani K, et al. Autologous Bone Marrow Mononuclear Cell Transplantation with Neurorehabilitation for Cerebral Palsy. **J Stem Trans Bio** 2017; 2(1): 110
9. Dr. Alok Sharma, Dr. Nandini Gokulchandran Mrs. Suhasini Pai, Ms. Pooja Kulkarni, Dr. Hemangi Sane, Dr. Khushboo Bhagwanani, Dr. Prerna Badhe. Diplegic dystonic Cerebral Palsy treated with cellular therapy: a case report. **Journal- International Journal of Case Studies.** 2017

#### **DUCHENNE MUSCULAR DYSTROPHY:**

1. Alok Sharma, Hemangi Sane, Amruta Paranjape, Khushboo Bhagwanani, Nandini Gokulchandran, Prerna Badhe. Autologous bone marrow mononuclear cell transplantation in Duchenne muscular dystrophy – a case report. *American journal of case reports* 2014;15: 128-134.
2. Dr. A. Sharma, Ms. P. Kulkarni, Dr. G. Chopra, Dr. N. Gokulchandran, Dr. M. Lohia, Dr. P. Badhe. Autologous Bone Marrow Derived Mononuclear Cell Transplantation In Duchenne Muscular Dystrophy-A Case Report. *Indian journal of Clinical Practice* 2012; 23 (3): 169-72.
3. Alok Sharma , Dr. Prerna Badhe, Hemangi Sane, Suhasini Pai , Pooja Kulkarni, Khushboo Bhagwanani, Dr. Nandini Gokulchandran. Halting of functional decline in a case of Duchenne Muscular Dystrophy after cellular therapy. *International Journal of Recent Advances in Multidisciplinary Research (IJRAMR)*, 2017 Jan.
4. Alok Sharma, Hemangi Sane, Vaibhav Lakhnopal, Amruta Paranjape, Pooja Kulkarni, Nandini Gokulchandran, Prerna Badhe. Stabilization of the disease progression in a case of Duchenne Muscular Dystrophy with cellular transplantation. *Stem cell: Advanced research and therapy.* 2017; 2017(3).



## **SPINAL CORD INJURY:**

1. Alok Sharma, Prerna Badhe, Pooja Kulkarni, Nandini Gokulchandran, Guneet Chopra, Mamta Lohia, V.C.Jacob. Autologous Bone marrow Derived mononuclear cells for the treatment of Spinal Cord Injury. *The Journal of Orthopaedics*. 2011;1(1):33-36.
2. Sharma A, Gokulchandran N, Sane H, Badhe P, Kulkarni P, Lohia M, Nagrajan A, Thomas N. Detailed analysis of the clinical effects of cell therapy for thoracolumbar spinal cord injury: an original study. *Journal of Neurorestoratology*. 2013;1:13-22.
3. Sharma A, Sane H, Gokulchandran N, Kulkarni P, Thomas N, et al. (2013) Role of Autologous Bone Marrow Mononuclear Cells in Chronic Cervical Spinal Cord Injury-A Longterm Follow Up Study. *J Neurol Disord* 1: 138.
4. Alok Sharma, Hemangi Sane, Dipti Khopkar, Nandini Gokulchandran, Hema Biju, V C Jacob, Prerna Badhe. Cellular therapy targeting Functional outcome in a case of Cervical Spinal Cord Injury. *Advances in Stem Cells* 2014 (2014)
5. Alok Sharma, Hemangi Sane, Dipti Khopkar, Nandini Gokulchandran, Varghese Chako Jacob, Joji Joseph, Prerna Badhe. Functional recovery in chronic stage of spinal cord injury by Neurorestorative Approach. *Case Reports in Surgery* 2014 Volume 2014, pages 1-4
6. Alok Sharma, Hemangi Sane, Suhasini Pai, Pooja Kulkarni, Amruta Paranjape, V C Jacob, Joji Joseph, Sanket Inamdar, Sarita Kalburgi, Nandini Gokulchandran, Prerna Badhe, Samson Nivins. Functional and symptomatic improvement after cellular therapy in a pediatric case of chronic traumatic incomplete SCI. *J Stem Cell Regen Biol* 2017;3(1):1-7.
7. Alok S, Prerna B, Suhasini , Hemangi S, Samson N, Pooja K, Amruta P, Dhara M, Nandini G. Functional Recovery and Functional Magnetic Resonance Imaging changes Following Cellular Therapy in a Case of Chronic Complete Spinal Cord Injury. *Curr Trends Clin Med Imaging*. 2017; 1(4): 555566.

## **BRAIN STROKE:**

1. Alok Sharma, Hemangi Sane, Anjana Nagrajan, et al., "Autologous Bone Marrow Mononuclear Cells in Ischemic Cerebrovascular Accident Paves Way for Neurorestoration: A Case Report," *Case Reports in Medicine*, vol. 2014, Article ID 530239, 5 pages, 2014. doi:10.1155/2014/530239.
2. Alok Sharma, Hemangi Sane, Nandini Gokulchandran, Dipti Khopkar, Amruta Paranjape, Jyothi Sundaram, Sushant Gandhi, and Prerna Badhe. Autologous Bone Marrow Mononuclear Cells Intrathecal Transplantation in Chronic Stroke *Stroke Research and Treatment*, Volume 2014, pages 1-9.
3. Dr. Alok Sharma, Dr. Hemangi Sane, Dr. Prerna Badhe, Ms. Pooja Kulkarni, Dr. Guneet Chopra, Dr. Mamta Lohia, Dr. Nandini Gokulchandran. Autologous Bone Marrow Stem Cell Therapy shows functional improvement in hemorrhagic stroke- a case study. *Indian Journal of Clinical Practice*, 2012;23(2):100-105.
4. Alok Sharma, Hemangi Sane , Amruta Paranjape, Nandini Gokulchandran , Sushant Gandhi, Prerna Badhe. Benefits of Autologous Bone Marrow Mononuclear Cell Transplantation in Chronic Ischemic Pontine Infarct. *Journal Of Case Reports* 2016;6(1):80-85
5. Alok Sharma, Hemangi Sane, Nandini Gokulchandran, Pooja Kulkarni, Rishabh Sharan, Amruta Paranjape, Prerna Badhe Effect of Cellular Therapy Monitored on Positron Emission Tomography - Computer Tomography Scan in Chronic Hemorrhagic Stroke: A Case Report. *Archiv Neurol Neurosurgery*, 2016 Volume 1(1): 22-25

## **HEAD INJURY:**

1. Alok Sharma, Hemangi Sane, Pooja Kulkarni, Jayanti Yadav, Nandini Gokulchandran, Hema Biju, Prerna Badhe. Cell therapy attempted as a novel approach for chronic traumatic brain injury - a pilot study. *SpringerPlus* (2015) 4:26.

2. Alok Sharma Hemangi Sane Pooja Kulkarni Nandini Gokulchandran Dhanashree Sawant Samson Nivins Prerna Badhe. Effect of Cell Transplantation in a Chronic Case of Traumatic Brain Injury. Transplantation Open. 2016 Volume 1(1): 22-25

**ATAXIA:**

1. Alok Sharma, Hemangi Sane, Amruta Paranjape, Nandini Gokulchandran, Hema Biju, Myola D'sa, Prerna Badhe. Cellular Transplantation May Modulate Disease Progression In Spino-Cerebellar Ataxia – A Case Report. Indian Journal Of Medical Research And Pharmaceutical Sciences. August 2014; 1(3).

**SPINAL MUSCULAR ATROPHY (SMA):**

1. A Sharma, P Kulkarni, N Gokulchandran, P Badhe, VC Jacob, M Lohia, J George Joseph, H Biju, G Chopra. Adult Stem Cells for Spinal Muscular Atrophy. Bangladesh Journal Of Neuroscience. 2009; 25(2): 104- 107.

## Chapters in international books



**CEREBRAL PALSY**  
CHALLENGES FOR THE FUTURE

Edited by **Emira Švraka**

**INTECH**

Chapter 7

**Stem Cell Therapy for Cerebral Palsy – A Novel Option**

Alok Sharma, Hemangi Sane,  
Nandini Gokulchandran, Prema Badhe,  
Pooja Kulkarni and Amruta Paranjape

Additional information is available at the end of the chapter  
<http://dx.doi.org/10.5772/57152>

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**1. Introduction**

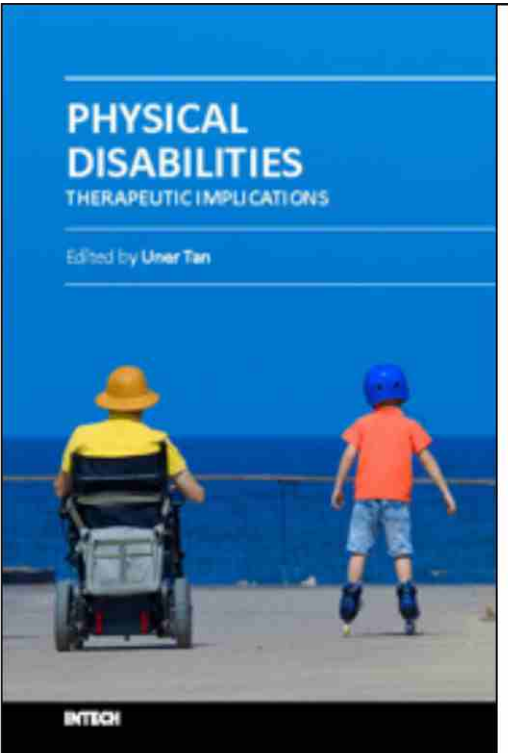
Discovery of stem cells by James Till and Ernest McCulloch in 1961, stands as one of the most remarkable medical research achievements of the 20th century. This discovery provided a foundation for further breakthroughs in the field of stem cells. Sir Martin J. Evans along with Mario R. Capecchi, and Oliver Smithies were jointly awarded a Nobel Prize in 2007 for their contribution in introducing specific gene modifications in mice by the use of embryonic stem cells. Later in 2012, John B. Gurdon and Shinya Yamanaka were also jointly awarded a Nobel Prize for discovering that mature cells can be reprogrammed to become pluripotent cells. [1]

Ramon y Cajal in 1926 stated “Once the development was ended, the founts of growth and regeneration of the axons and dendrites dried up irrevocably. In the adult centers, the nerve paths are something fixed, ended, and immutable. Everything may die, nothing may be regenerated. It is for the science of the future to change, if possible, this harsh decree.” [2]. It was a long-standing belief that cells of the central nervous system once damaged cannot be regenerated. The medical science of stem cells has finally made restoration of CNS possible which has changed the old concept of medicine. Not too long ago, this therapy was hampering by various controversies, ethical and moral issues. But, tremendous progress of research in this field has finally led to its translation from laboratory to innovative clinical therapies.

A variety of cells including embryonic stem cells, adult stem cells, umbilical cord blood cells and induced pluripotent stem cells have been explored as a therapeutic alternative for treating a broad spectrum of neurologic disorders including stroke, Alzheimer’s, Parkinson’s, spinal cord injury, cerebral palsy etc. amongst others. It is essential to select suitable cells depending on the nature and status of neurological dysfunctions to achieve optimal therapeutic efficacy. Along with the selection of cells, the route of administration also plays an important role to

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**PHYSICAL  
DISABILITIES**  
THERAPEUTIC IMPLICATIONS

Edited by **Uner Tan**

**INTECH**

Chapter 8

**Stem Cell Therapy in Pediatric Neurological Disabilities**

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Additional information is available at the end of the chapter  
<http://dx.doi.org/10.5772/57656>

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**Abstract**

Prevalent neurological disorders represent a major part of the disabilities worldwide. In over 10 decades of research to find a cure for these disorders, medical science has not been able to repair the underlying brain injury. This chapter focuses on recent advances in the application of stem cells as a therapeutic tool for some of the common neurodevelopmental disorders (cerebral palsy, autism, intellectual disability and muscular dystrophy). The mechanism of action of stem cells in each disorder has been explained. A review of clinical data has been described giving a clear understanding of current status of stem cell therapy in these disorders. Various factors influencing the selection of stem cell therapy such as different types of cells, different routes of administration and dosage and frequency of transplantation have also been discussed. Our experience of treating these disorders is published in the form of our published data. Use of novel monitoring tools such as MRI/MRS and PET-CT can help to track the changes occurring at cellular level after stem cell therapy are described. We also highlight the importance of a multi-disciplinary approach of combining rehabilitation with stem cell therapy.

**Keywords:** stem cell therapy, autism, cerebral palsy, muscular dystrophy, intellectual disability

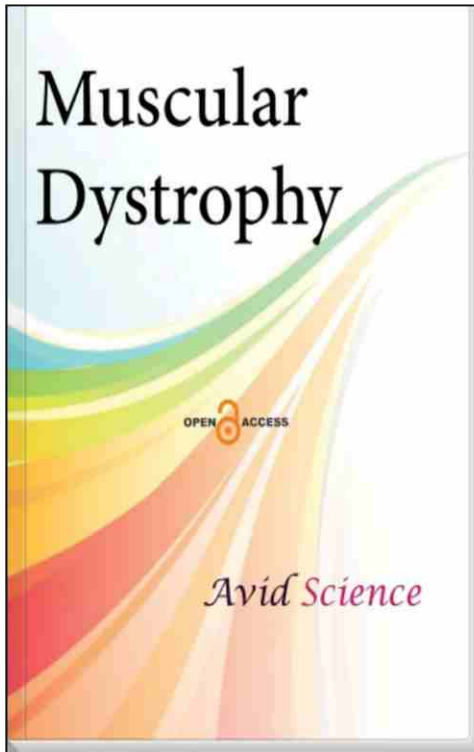
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**1. Introduction**

Neurodevelopmental disorders (NDD) are characterized by an abnormal development of the brain during the early development phase, leading to a myriad of symptoms and diseases, including delayed milestones and deficits in personal and social functioning [1]. The developmental deficits can vary from specific limitations of adaptive, behavioral and

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*Muscular Dystrophy*

## Chapter 2

### Stem Cells as a Therapeutic Modality in Muscular Dystrophy

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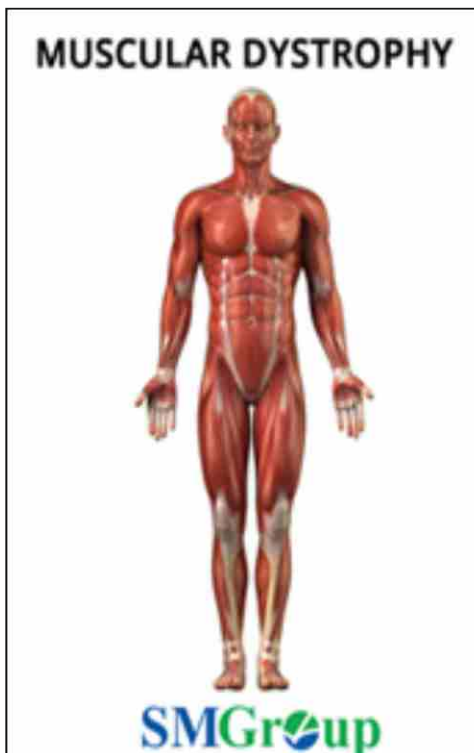
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First Published **June 02, 2017**

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### Role of Stem Cell Therapy in Treatment of Muscular Dystrophy

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**Published Date:** July 23, 2016

#### INTRODUCTION

Muscular dystrophy connotes a group of disorders due to genetic abnormality of the genes that code for various proteins essential for the integrity of muscle cells. Most of these disorders are hereditary but there can be sporadic mutations as well. Different types of muscular dystrophies are identified based on the core genetic abnormality however they are all characterized by progressive muscle degeneration. Some of the most severe forms of muscular dystrophies are Duchenne's muscular dystrophy (**DMD**) and Congenital muscular dystrophy (**CMD**); the onset of which is early in the childhood, with fast progression and considerably reduced lifespan up to second or third decade of life. The other forms like becker's muscular dystrophy (**BMD**), limb girdle muscular dystrophy (**LGMD**), facioscapulohumeral dystrophy (**FSHD**) have their onset ranging from 2nd to 4th decade of life and the lifespan is largely unaltered but quality of life reduces significantly with severe disability in the later part of life.

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## Introduction

Disability is a part of the human condition. Almost everyone of us will be disabled temporarily or permanently at some point in life. With increasing age we notice increased disability and increasing difficulties in independent functioning. Most extended families have a disabled member, and many non-disabled people take responsibility for supporting and caring for their relatives and friends with disabilities. Challenges to a person's health can happen to anyone, at any age and at any time because of different causes. When limitations related to a medical condition arise and begin to have a negative effect on essential life functions, such as walking, talking, seeing, hearing, or working functions often referred to as "Activities of Daily Living" (ADLs), a person is said to have a disability.

Disability is generally equated with incapacity but disability is an evolving concept. Limitation experienced by a disabled person results from a combination of the person's disability with attitudinal and environmental barriers that prevent their full and effective participation in the society. Responses to disability have changed in the late 19<sup>th</sup> century due to large self-organized groups of people with disabilities who have created awareness about disability and have fought for their rights. Now disability is viewed under the domain of human rights. In the recent past the historical attitude of segregating the disabled has changed into a more inclusive approach. This approach has warranted changes in the environment, infrastructure as well as national policies to support people with disability. Knowledge and attitudes are important environmental factors, affecting all areas of service provision and social life of people with disability.

In the current international classification of function disability is identified as the term for encompassing impairments, activity limitations and participation restrictions, and negative aspects of the interaction between an individual with disability and his/her environmental factors. This classification explains disability as a dynamic interaction and does not limit it to its physical or cognitive attributes. ICF highlights disability as an interaction between body functions, body structures, environmental factors and participation in personal or social tasks.

Making disability a concept and not a personal attribute. It is very important to understand this and address disability as multifaceted problem which requires holistic solution.

When disability occurs in a child either acquired after birth or since birth or a progressive disability; it is very important to empower the parents and caregivers about the medical management of the condition as well as the social implications. The aim of this book is to provide information to patients, parents and caregivers to provide best quality care to the children limited from various disabilities.

Disability can be categorized in to two broad categories

**Physical disability:** In which body structure and function is damaged however cognitive capacities are unaltered.

**Cognitive disability:** In which intellectual and cognitive capacities are affected and physical capacities may be affected or preserved. Cognitive disability can be measured by calculating intelligence quotient and on the basis of this it is categorized as

**Mild cognitive disability:** Accounting for around 85% of all cognitive disabilities. children in this category have IQ scores between 55 and 70 and are usually included in the regular classroom.

**Moderate cognitive disability:** Students with this type of disability have IQ scores between 30 and 55.

**Severe cognitive disability:** Kids with severe cognitive disabilities have IQ scores that fall under 30 and will have few communication skills, and will need direct supervision. Of all cognitive disabilities, only about 3 to 4% of children have a severe cognitive disability.

### **What is multidisciplinary management of disability?**

Diseases of the neurological system are the most challenging for the modern medicine. Most of these remain incurable and lead to major neurological deficits. This affects the quality of life of patients and puts significant burden on the caregivers. Patients are completely dependent on the care givers for their day to day activities. Parents of these patients do not have easy access to the guidance required to manage their

disabilities and secondary complications. Most of them find it difficult to follow the rehabilitation routine at home. This leads to higher incidence of secondary complications and increases the financial and care burden on the parents. Therefore, to provide good quality care and improve the functional outcome, long term inpatient multidisciplinary neurorehabilitation is required.

Neurorehabilitation is an educational process that reduces the level of disability experienced by a patient and increases the level of participation. Neurological disorders affect multiple body systems and lead to physical, cognitive and functional disability. Therefore, a team of qualified professionals with expertise in treating neurological disorders is required to work as an integrated unit with a patient centered approach to provide quality care.

### **Multidisciplinary team for neurorehabilitation**

Neurorehabilitation team consists of core participants like neurosurgeon, neurologist, urologist, nurse, physical therapist, aquatic therapist, occupational therapists, speech therapist, psychologist, dietitian and family members or caregivers of the patient. Multiple other professionals like plastic surgeons, orthopedic surgeons, andrologists, cardiologists, ophthalmologist, gastroenterologist and ENT specialists form the secondary team and may be given appropriate referrals when required.

### **Aims of neurorehabilitation**

Process of neurorehabilitation is to reintegrate the patient in community. Therefore, the most important aim of the neurorehabilitation is to facilitate maximum neurological recovery and functional independence. However, some of the other important aspects of neurorehabilitation are to prevent secondary complications, deformity prevention and crisis management. The process of neurorehabilitation educates the patients and caregivers about the prognosis of the condition, makes them aware of the possible complications and helps them to manage the complications in the most effective way. Neurorehabilitation process also includes compassionate therapies and palliative care provided at the terminal stages of the progressive disease.



## **Core participants in neurorehabilitation**

### **1. Physiotherapist:**

Physiotherapists address the physical disability experienced by the patients. They assist in movement restoration, improving range of motion of the joints, increasing muscle strength and endurance. They work towards improving the balance and co-ordination. It also includes retraining for the appropriate gait pattern and independent ambulation. Management of spasticity and prevention of further deformities are some of key aspects of physiotherapy.

### **2. Occupational therapist:**

Occupational therapists work towards functional independence of the patient. Training for activities of daily living and self care activities such as dressing, eating, bathing & personal hygiene is the most important component. It involves extensive patient and family education to make home environment barrier free and accessible. They also teach energy conservation techniques to minimize fatigue and conserve energy. Vocational rehabilitation forms an important part of occupational therapy.

### **3. Aquatic therapist:**

Aquatic therapist makes use of physical properties of water. The treatment is conducted in a swimming pool which allows for early movement restoration, reduction of spasticity, balance training, muscular strengthening, improved range of motion of the joints and greater degree of freedom. The aquatic therapist carefully identifies the problem areas and designs patient specific water based exercise program that slowly translates into land based exercises. Aquatic therapy is important for early therapeutic intervention.

### **4. Speech therapy:**

Role of speech therapy is to improve clarity, loudness and quality of speech (Dysarthria management) and to improve swallowing ability and prevent choking spells and aspiration (Dysphagia management)

### **5. Clinical psychologist:**

Neurological disorders are devastating for the patients and family members. Most often they are unable to cope with their disease and need guidance. Clinical psychologist identifies the problem areas and provides and counseling and guidance for better coping strategies to both the patients and care givers. They also contribute towards the cognitive rehabilitation of the patient.

### **6. Special educator:**

Children with cognitive disability require specialized teaching for academic as well as conceptual learning. Special educators provide this education and training for children and guide their parents for the same.

### **7. Dietician:**

Nutrition is most important for good recovery. A dietitian taken into consideration the nutritional needs and prescribes a feasible diet program to the patients.

Neurological disorders are complex and challenging leading to severe functional deficits. Long term inpatient 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. In the next sections of the book we will look in to detailed management of some of the physical and cognitive disabilities in children caused by neurological disorders.

**Section A:**  
**Management of children**  
**with Cognitive disability**



## Chapter 1

# Autism

## 1.1 About Autism

### What is Autism?

Autism Spectrum Disorder is a Neurodevelopmental disorder which typically appears during early childhood. The term Autism was first used by Leo Kanner in 1943. He described it as a psychological disorder. According to his description people with Autism show “an inability to relate to in an ordinary way to people and situations” (Kanner, 1943, p.217). According to the Centers for Disease Control and Prevention (CDC), the prevalence rate of Autism is 1 in 68 children.

### Symptoms of Autism

Autism Spectrum Disorder involves a large range of symptoms, and level of Impairments such as:

- Impaired social interaction which makes it difficult for them to initiate a conversation with other people.
- Impaired cognition and problem solving skills.
- Not responding to name.
- Difficulty in understanding other's expressions and emotions.

- Difficulty in conveying their needs appropriately.
- Repetitive and restricted patterns of behavior and interest.
- Failure to make and maintain appropriate eye contact with others.

Sensory issues like a:

- Avoids touch or seeks more physical touch, avoids/seeking certain texture of clothing or food.
  - Seeking constant movements of different parts of the body like rocking, swinging, hand flapping, jumping or avoiding movement like fear of climbing stairs, avoids swings, and traveling.
  - Hypersensitive towards loud noise.
  - Licks and tastes certain textures or avoids certain textures or food items.
  - Staring at spinning objects, light objects.
- Hyperactivity in the form of excessive restlessness, physical activity and not sitting at one place.
  - Difficulty in Non-verbal communication and understanding non-verbal cues of others.
  - Self-injurious behavior like self-biting, head banging.
  - Difficulty in Adapting to change.
  - Delayed speech and language skills.
  - Inappropriate emotional responses like laughing and crying without reason.
  - Inappropriate attachment towards objects.
  - Difficulty in carrying out activities of daily living.

### **Causes of Autism**

- Prenatal Factor like advanced age of the mother during pregnancy.
- Health of the mother during pregnancy like diabetes, hypertension, stress, thyroid, use of psychiatric drugs during pregnancy.

- Neo-natal factors such as low birth weight, lack of oxygen of the child during birth, seizures, infections etc.
- Postnatal factors like allergies, high fever, immune system abnormalities, exposure of children to drugs, certain food, or heavy metals etc.
- Gastrointestinal abnormalities in the child etc.

However, there is no substantial evidence to pin point the cause of the disease.

### **How is Autism Diagnosed?**

There are certain early signs of Autism which are noticed by the parents, teachers and caregivers. Many a times, they fail to notice early signs of Autism. Reason behind this is lack of knowledge among parents and teachers, this lead to delay in the child's intervention. Diagnosing Autism in children requires assessment by multiple evaluators, using different assessment tools and methods. This is done by Psychologists, Developmental pediatricians, an Occupational therapist, and speech and language therapists. This team carries detail observation, assessment, tests and pinpointing each and every chief complaint of the patient. This includes:

- Parent's interview by a Psychologist in which parents are asked about the developmental milestones, their habits, abilities, and challenges the child is facing.
- Psychologists take a detailed history of the patient in the form of assessing and observing his/her attention and concentration abilities, understanding level, social interaction and if there is any family history of similar condition.
- Various psychological tests which are designed to evaluate the child are carried out and the diagnosis of the child is confirmed.
- Occupational therapists assess the patient's chief complaints such as fine and gross motor skills, sensory issues, eating, bathing, dressing and toileting.
- They also assess muscle tone, posture and strength of the patient.

- Speech and language therapists observe the patient's receptive and expressive skills, voice quality including use of intonations.
- Pediatricians also recommend auditory tests i.e. BERA, to identify if there is any hearing problem.

## **1.2 Multidisciplinary management of Autism**

### **Treatments Available for Autism**

Treatment of Autism requires involvement of patient's family as well as team of professionals. Recent interventions for Autism Spectrum Disorder includes ABA therapy which involves one-to-one sessions, group sessions, social training sessions as well as parent training. Speech therapy, Occupational Therapy and Special Education. Apart from this, therapies like stem cell therapy, chelation therapy, HBOT, Auditory Integration therapy, animal assisted therapy are also provided for the treatment of Autism. Apart from therapies medicines are commonly used as a treatment for the symptoms of hyperactivity, problem behavior, anxiety and sleep problems.

### **Medical management of Autism**

The most commonly used prescription medicines are different types of antipsychotics, antidepressants, stimulants, mood stabilizers, anticonvulsants, Glutamate antagonists, vitamin and mineral supplements. The medicines don't address the core pathology of the disease but provide symptomatic relief.

Antipsychotics can be given to reduce the irritability, Antidepressants to treat depression, anxiety and obsessive compulsive disorder in autism. Drugs such as methylphenidate, amphetamine, and dextroamphetamine belong to the category of stimulants which act on the dopamine system and are primarily used to treat hyperactivity and inattention. Lithium, lamotrigine, valproic acid, carbamazepine, topiramate, oxcarbazepine and levetiracetam are used as mood stabilizers to control behavioral symptoms. For the patients who suffer from epilepsy anti-epileptics can be given. Drugs like amantadine, memantine lead to improvement in memory, hyperactivity, irritability, language, social behavior and self-stimulatory behavior. Sedatives can be used in case of patients with



disturbed sleep cycles. Apart from the medicines various suppliments like vitamins and anti-oxidants are also prescribed to reduce inflammation and promote brain development.

## **ABA (Applied Behavior Analysis)**

**ABA therapy:** Applied Behavior Analysis is a scientific study of behavior. It is defined as the process of systematically applying different interventions based upon the principles of learning theory. ABA therapy improve social significant behaviors to a meaningful degree. It involves various techniques to increase and decrease a target behavior.

### **ABC MODEL**

It refers to Antecedent, Behavior and Consequence. These are the building blocks of analyzing and changing challenging behaviors.

*Antecedent*-These are events or behavior which precedes the target behavior. It happens right before the behavior has occurred. It might be a request/ command given by the teacher/ therapist, it also refers to any situational event, e.g. presence of another person.

*Behavior*- This refers to the 'Target Behavior'. Everything a person does, say feel or think is a behavior. Any reaction or response from a person is a behavior. In order to increase or decrease the target behavior, it is important to clearly define the behavior.

*Consequence*- This refers to what happens after the behavior occurs. It could be a reaction from a therapist/ teacher/ parent in the form of a tangible reward, verbal praise, or a reprimand.

### **Example of ABC Model**

*Antecedent:* Therapist provides instruction to a child to fix a two piece puzzle.

*Behavior:* Child fixes the puzzle.

*Consequence:* Child receives his/her favorite toy as a reward from the teacher.

## **Different Strategies of Applied Behavior Analysis**

### **1. Discrete Trial Training**

Discrete Trial Teaching (DTT) is a method of teaching skills to a child which involves breaking down a skill to be learnt into small discrete steps and repeated trials are taught one-to-one. For e.g. Therapist teaching colors to a child might start by teaching red color. She would ask the child to point to red and then reward the behavior if the child responds by pointing to the red card. She would then move on to teaching blue by itself, reinforce the skills and ask about both the colors together. DTT have found to be effective in teaching skills to children with Autism.

### **2. Shaping**

The process of shaping involves differentially reinforcing any successive approximations towards a terminal behavior. Differential Reinforcement is a procedure which reinforcement is provided for responses that share a predetermined criterion and reinforcement is withheld for responses that do not match the criterion. Egg: Teaching a child how to wear his eyeglasses by himself involves the following steps:

- Teaching the child to first pick up the glasses by himself and reward is provided immediately.
- Once the child learns to pick up the glasses, rewards is then provided if the child learns to place the glasses up to his face.
- Once first two skills are mastered then the therapist moves onto the final skill i.e. placing the glasses in their proper position and reward is provided for performing the final skills and not the first two skills.

Shaping uses a positive approach to teach new skills. Shaping can also be combined with other established behavior change or behavior building procedures such as the chaining.

### 3. Chaining:

A behavior chain involves breaking down a skills into simple steps or units. Every step performed by a child is reinforced in a behavior chain. E.g. : Teaching a child wearing his jacket by himself would involve the following steps in a behavior chain.

- Obtain the jacket from closet
- Place one arm in sleeve
- Place second arm in sleeve
- Zip up the coat

### 4. Prompting:

Prompts are supplementary antecedent stimuli that are used to occasion a correct response which eventually controls the behavior. It is a physical help, instructions, gestured and demonstrations provided by the therapist to teach a child and eventually increase the responses from the child.

#### Types of Prompts:

- **Physical Prompts:**It is also known as hand over hand prompt. It refers to physically guiding the child to complete a task. E.g., holding a child's hand to teach body parts.
- **Verbal Prompts:**It involves any verbal assistance or a hint that is provided to teach any skill to the child. E.g. "asking the child what is the color of the sky?" and providing prompt like "say Blu."
- **Gestural Prompts:**It involves pointing, touching an object or any other action a child can observe in order to learn. E.g., pointing towards a cup and asking the child "what is something you drink from".
- **Positional Prompts:** Involves using 2D cards or objects on the table. To implement a positional prompt a therapist while teaching object identification to a child between 'Apple' and 'Banana' will place apple card close to the child.
- **Visual Prompt:** It is also known as Model prompt where the child

learns by observing others or looking at videos, flashcards etc. E.g. Teaching a child turn taking by showing videos of kids engaging in turn taking.

- **Auditory Prompt:** It involves any kind of sound a child can hear in order to complete a task. E.g., Mother tells the child “clean up your toys” and prompts the child by setting up a timer to go off in 2 mins as a cue for finishing the clean up task.

However, it is important to fade away these prompts to avoid prompt dependence. For E.g., In order to teach a child how to Zip his jacket, therapist will provide prompts hierarchy in the form of Hand-to- hand, hand to elbow, hand to shoulder and then no physical contact.

### **5. Imitation Training:**

Imitation training is an important aspect in the development of a child. Imitation skill teaches various other skills to the child such as pretend play, social play, language other skills that requires observational learning. Steps to teach imitation skills :

- Sit with the child face to face.
- Make sure the child is giving eye contact.
- Give the instruction to the child as “do this” and show the model for imitation.
- Wait for 3-5 seconds if the child imitates give reward if not then physically prompt the child.

### **6. Self-management Training:**

Self-management training is a useful technique to achieve greater levels of Independence and generalization among people with Disabilities. Self-management involves the following components :

- **Self-Monitoring-** It is a procedure whereby a person observes and responds to, usually by recording, the behavior s/he is trying to change.
- **Self-evaluation-** It involves comparing performance of oneself with a predetermined goal or standard.

- ***Self-monitoring with Reinforcement***-This involves providing reinforcement to oneself for either meeting goals selected for oneself or taught by others. The reinforcer can be self-administered or delivered by a therapist.

Applied Behavior Analysis investigates socially significant behavior with immediate importance to the children. It also entails precise measurement of the actual behavior in need of improvement. It is effective and improves behavior sufficiently to produce practical results for the child. Lastly, it produces behavior change that last over time, appear in other environments and generalize the appropriate learned behavior.

### **Psychological Intervention**

Raising individuals with disabilities can be a challenging process for the caregivers. A Psychologist play an important role in helping people with different disabilities manage their anxiety levels, modify their behavior , help them manage their anger and also help the patients as well as families manage their stress and emotional issues. However, providing complete information and proper knowledge along with techniques to manage the complications makes it possible to reduce these issues. Psychological treatment plays a pivotal role and involves the following techniques:

**Behavior Modification**: This involves focusing on one area at a time rather than dealing with all at once. Narrowing down a behavior will help us understand the function of the occurrence of a behavior so that it becomes easier to change and manage it. Behavior modification replaces undesirable behavior to more desirable ones through the process of Positive and Negative Reinforcement. Various strategies involved in behavioral Modification are :

- **Positive Reinforcement**: It is a process of increasing a desirable behavior by presenting a stimulus to the person after the desirable behavior is exhibited making that behavior more likely to occur in future. E.g., providing edibles, desired items like toys, stars, stickers, providing tokens and even verbal praises are few examples of positive reinforcement.

- **Negative Reinforcement:** It is a process of termination, reduction of a stimulus on the occurrence of a response which leads to an increase in the future occurrence of a response. E.g., Teaching cleaning up the mess to a child in order to avoid solving maths problems which s/he doesn't like. In the future, the child will be more likely to engage in clean up tasks because it was rewarded by removing aversive task of solving maths problems.
- **Positive Punishment:** Refers to presenting a stimulus immediately following a response which decreases the future frequency of that type of behavior in similar conditions. For e.g., Providing a reprimand as “No” Contingent upon occurrence of aggressive behavior. Another example could be that a child is asked to clean up the toys after s/he throws it in the room.
- **Negative Punishment :** It involves removing a desired stimulus after an undesirable behavior is exhibited. Also refers to withdrawal or loss of opportunity to earn reinforcement. For E.g., A child is asked to sit in an alone room after engaging in aggressive behavior with other children removing the opportunity to be with others.
- **Extinction :** This procedure occurs when reinforcement of a previously reinforced behavior is discontinued, as a result the frequency of that behavior decreases in the future. For e.g., Ignoring the behavior of throwing tantrums of a child who engages in this behavior in order to gain teacher's attention. Here, attention was a reinforcement for the child which was put on extinction.

Psychoeducation : Refers to the process of providing information and educating people regarding a better understanding of their illness to them and their family members. Helping people developing a thorough understanding of the difficulties and challenges they are facing as well as providing knowledge of personal coping ability. Psychologists also provide insight into their own areas of strengths, and help them have a greater internal capacity in order to work towards mental and emotional well- being. Psychoeducation can be provided one-to-one as well as in group. People who participate in psychoeducation has a positive impact on their quality of life. When people who have been diagnosed with a

disability/ illness are made to understand what their diagnosis means, they are more likely to view their condition as treatable rather than being ashamed of it. Family Involvement in Psychoeducation increases compliance and confidence of the individual and ensuring that the person is given adequate support.

Psychoeducation can be provided in the following set up :

- Private Clinics
- Hospitals
- Special Schools
- Through Internet
- Rehabilitation Centers

Family Counseling: Presence of a family member with an illness involves attention in the form of time and resources. Psychologists conduct family counseling to help the family members vent out their emotions, help them to cope with situations better and provide a nurturing environment to the patient.

*Family Counseling can help with different issues like :*

Parent's mental health  
Trauma due to family member's illness

Issues like stress, anxiety, depression and grief

Stress due to financial problems

Relationship between the parents

**Psychologist practicing family counseling uses different approaches:**

*Family Counseling using Cognitive Behavioral Therapy* : It attempts to change the ways people think or behave in order to reduce the problem.

*Family Counseling using Psychodynamic techniques* : It tends to look more into the individual's own subconscious minds. It is based on the principle that providing the individuals in the family the real reason behind what is going on, and this will help the people to deal with their difficulties more easily.

***Systematic Family Counseling:*** It attempts to identify different kinds of problems, relationships, ideas, attitudes and thinking of the whole family. Once these areas are discussed with the family members, the counsellor attempts to shift the problems, attitudes, relationships, to a position that is more beneficial, less damaging, or simply make it more realistic.

## **Occupational Therapy Treatment**

### **Goals:**

1. Maintaining the arousal level of the child.
2. To improve eye contact.
3. Improve attention and concentration.
4. Improve social skills.
5. Improve gross and fine motor skills.
6. Improve eye hand co-ordination.
7. Improving motor planning.
8. Improving functional independence by using adaptive devices and assistive technology.
9. Home, Environmental and classroom modifications.

Social skills, affected communication and presence of sensory processing issues affects child's participation in academic activities and affect the daily routine.

Children with autism also have impaired body awareness, they can't use their body parts for doing appropriate and meaningful actions and activities. Which ultimately affects their school performance.

Occupational therapy helps children with autism to organize sensory information and to translate it into meaningful and purposeful activities.

### **Sensory integration therapy:**

SI therapy will help child to get exposed to different sensory stimuli in a structured and repetitive way.



School should have a separate provision of sensory integration unit.

**Improving eye contact:**

- Give direct verbal instructions to the child (Look at me when you are talking).
- Direct nonverbal instructions: Move your finger along with your eyes from my eyes to your eyes.
- Indirect verbal instruction: instruct the child by saying that " I don't think so you are talking to me as you are not looking at me"
- Indirect nonverbal: Putting the sticker on the forehead and asking the child to focus on the sticker while talking to you.

**Improving Visual perceptual skills:**

- Giving visual, auditory cues to improve directional concept Eg. Colored dot or arrow at the beginning of the page to help the child to understand from where to start writing.
- Using the 'alphabet lines' to help the child in maintaining alignment of the letters and words.
- Use of word search puzzles.
- Games like copying 2D and 3D design helps to improve directional concept.
- Identifying letters by touch (writing letters on the back and asking the child to identify the letters).
- Children with autism have good visual processing.
- Use flashcards, charts, pictures for teaching them new skills.

**Gross motor coordination:**

- Improving gross motor coordination by engaging the child in gross motor activities like dancing (Simple steps) and swimming.
- Giving activities like ball throwing, punching the bag, kicking the bag, marching.

- Practicing functional activities like drinking water, dressing.

### **Fine motor activities:**

- Asking the child to pick up small objects of different shapes and sizes. Giving him activities like lacing, stacking small blocks.
- Manipulation games: Picking up sticks and making design.
- Engaging the child in craft activities.
- Making the child to do Self-help skills like buttoning, zipping, opening and closing the lunch box, bottle caps.

### **Social skills:**

- Story telling: Put the pictures on the table. Ask the child to make a story on basis of these pictures.
- Conversation game: By giving a topic ask the child to express his /her opinion on that and start the conversation.
- Ask the child to act out the different emotions by showing them different cards.

### **Adaptive Devices:**

- Children with autism have difficulties in doing daily routine tasks because of low muscle tone, poor proximal stability of shoulders, affected co-ordination.
- Adaptive devices like enlarged handle spoon, modified glass, glass holders to hold the glass, writing devices, adapted buttons, zippers can help children to do ADL activities faster, more easily and independently.
- In school using adaptations like modified paper, stencils, highlighters can help children to excel in the academic skills.

### **Use of assistive technology:**

- Use of key board instead of actual writing.
- Adaptive Switches to say simple 'Yes' and 'No'.

- Use of pencil grippers for children who have difficulties in holding the pen/pencil.
- Using Speech amplifier, voice activated headphones.
- Touch screen tablets, word processor, e reader.

### **Classroom and Environmental Modifications:**

- Cool down rooms: a room where children can go before the child gets aggressive. A school should have provision for the same.
- Colors of the wall should be preferably light and should have a soothing effect on child.
- Don't make the child sit in front of the wall which is painted with bright colors and multiple colors.
- Make sure that there is ample light in the room.

### **Animal Assisted Therapy**

Animal Assisted Therapy (AAT) is a type of therapy which involves animals as a form of treatment and it play a major role in improving a person's social, cognitive and emotional functioning. Animal –assisted therapy can vary from something as simple as bringing a pet into the home like a pet dog, cat or it can also be very structured as horse riding or swimming with dolphins. A particular kind of bond and relationship formed with animals can especially help children with autism a better sense of well- being, confidence and develop sense of empathy. Animals used in AAT are dogs, cat, horses and sometimes even dolphins. It is carried out by a specialist in professional training in Animal Assisted Therapy.

Animal Assisted Therapy is helpful for people with different disabilities in the following way: It helps in social and emotional development.

- It modifies the behavior of children.
- Develops communication skills in children.
- Helps in developing care taking skills.

- Helps to overcome sensory defensiveness among children with sensory issues.
- Increases attention span.
- Enhance problem solving and cognitive skills.
- Helps the child to learn about caring and providing unconditional love.

### **Aquatic therapy**

A new and fast developing area of therapy is aquatic therapy. It is found to be extremely beneficial and exciting for children with autism. The Aquatic Therapy is using water and specifically designed activity by qualified professional to aid in the restoration, extension, maintenance and quality of function for children or adults with acute, transient, or chronic disabilities, syndromes or diseases.

Water immersion is beneficial for memory and cognition, it increases blood supply to brain and heart, activating the parasympathetic nervous system and suppressing the sympathetic nervous system. This helps to reduce anxiety, hyperactivity, repetitive motor mannerisms, stereotypical behavioral and inappropriate emotional responses.

Use of aquatic therapy can improve posture, co-ordination, sensory processing difficulties experienced by the child, attention span, concentration, impulse control, frustration tolerance and ability to follow instructions. Aquatic helps to promote physical activity and regularizes the sleep patterns of the children.

### **Physiotherapy**

A small percentage of patients shows some motor impairments as well like low muscle tone, increased joint flexibility, poor core strength and co-ordination, poor balance etc. A physiotherapist can address these issues. A physiotherapist can also identify the sensory processing deficits that in turn may hinder movements and motor performance and may use sensory integration to facility correct movement patterns.

Despite all the rehabilitative therapies, majority of children are still dependent for all of their daily activities on their caregivers. Although rehabilitative therapies improve quality of life of children with Autism and improved their level independence, it does not alter the core pathology of the disease. There is no medicine available that can cure Autism. Therefore there is a need for a treatment that will address the core problems in the brains of the children with Autism.

### **Stem Cell Therapy**

Stem cell therapy is newer treatment modality available for Autism. Basic underlying principle of stem cell therapy is to replace non-functional cells with functional cells, to improve the function of dysfunctional cells and to protect the cells with preserved function from any damage. Stem cells promote repair of the damaged tissues and replace the unhealthy tissue with healthy tissue.

Therefore, stem cell therapy has the potential to alter the basic problem causing the disease and bring about improvements in the brain function of children with Autism.

There improvements Stem cell therapy brings about significant improvements in:

- Hyperactivity
- Eye contact
- Attention and concentration
- Speech and/or communication
- Command following
- Fine motor activity
- Reduced self- stimulatory behavior
- Social awareness
- Social behavior

### **Improved function of the cells because of stem cell therapy can be seen in the PET-CT scan**

Patients with autism show reduced metabolism in different areas of the brain like Mesial Temporal Lobes, Thalamus, Hippocampus, Parahippocampus, Amygdala, Cerebellum, Caudate Nucleus, Superior And Middle Temporal Pole. Post stem cell therapy there is a significant increase in the metabolism of these areas .

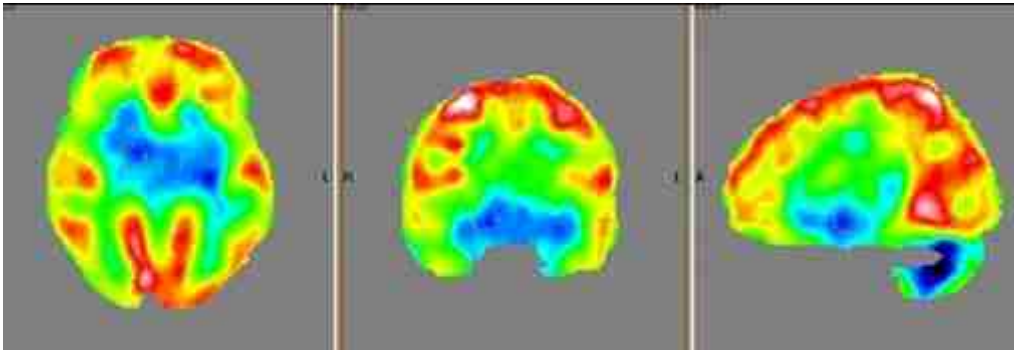


Fig 1.8 PET-CT scan showing areas of brain with reduced metabolism and function showing brain damage in blue colour

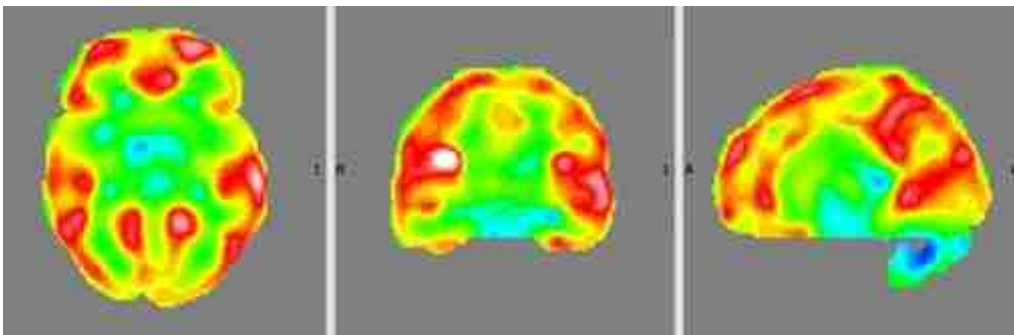


Fig 1.9 PET-CT scan images after stem cell therapy showing reduction in the blue colour i.e improved function post stem cell therapy

### **Scientific publications of NeuroGen in Autism**

1. Sharma A, Gokulchandran N, Shetty A, Kulkarni P, Sane H, Badhe P. Neuropsychiatric Disorder Tackled by Innovative Cell Therapy-A Case Report in Autism. *J Stem Cell Res Transplant*. 2014;1(1): 4.
2. Alok SharAlok Sharma, Nandini Gokulchandran, Hemangi Sane, Anjana Nagrajan, Amruta Paranjape, Pooja Kulkarni, Akshata Shetty, Priti Mishra, Mrudula Kali, Hema Biju, Perna Badhe. Autologous bone marrow mononuclear cell therapy for autism – an open label proof of concept study. *Stem cell international*. 2013 (2013), Article ID 623875, 13 pages.
3. Alok Sharma, Nandini Gokulchandran, Perna Badhe, Pooja Kulkarni, Priti Mishra, Akshata Shetty and Hemangi Sane. An Improved Case of Autism as Revealed by PET CT Scan in Patient Transplanted with Autologous Bone Marrow Derived Mononuclear Cells. *J Stem Cell Res Ther* 2013, 3:2.
4. Alok Sharma, Nandini Gokulchandran, Akshata Shetty, Hemangi Sane, Pooja Kulkarni and Perna Badhe. Autologous Bone Marrow Mononuclear Cells may be Explored as a Novel. Potential Therapeutic Option for Autism. *J Clin Case Rep* 2013, 3:7.
5. Alok Sharma, Nandini Gokulchandran, Hemangi Sane, Pooja Kulkarni, Nancy Thomas, Amruta Paranjape, Perna Badhe. Intrathecal autologous bone marrow mononuclear cell transplantation in a case of adult autism. *Autism open access*. 2013, 3:2.
6. Alok Sharma, Nandini Gokulchandran, Hemangi Sane, Pradnya Bhovad, Hema Biju, Akshata Shetty, Mrudula Kali and Perna Badhe. Cell therapy effects portrayed on positron emission tomography computerized tomography scan of the brain serve as a new dimension for autism: A case report (2014), *Journal of Paediatric Neurology*, 12:3.
7. ma, Guneet Chopra, Nandini Gokulchandran, Mamta Lohia, Pooja Kulkarni. Autologous Bone Derived Mononuclear Transplantation in Rett Syndrome. *Asian Journal of Paediatric Practice*. 2011; 15 (1): 22-24.

8. Alok Sharma, Nandini Gokulchandran, Hemangi Sane, Avantika Patil, Akshata Shetty, Hema Biju, Pooja Kulkarni, Perna Badhe. Amelioration of Autism by Autologous Bone Marrow Mononuclear Cells and Neurorehabilitation: A Case Report. American Journal of Medical Case Reports, 2015, Vol. 3, No. 10, 304-309.
9. Alok Sharma, Hemangi Sane, Nandini Gokulchandran, Perna Badhe, Avantika Patil, Pooja Kulkarni, Amruta Paranjape PET- CT scan shows decreased severity of Autism after autologous cellular therapy: A case report. Autism Open Access 2016; 6:169.
10. Alok Sharma, Hemangi Sane, Nandini Gokulchandran, Perna Badhe, Pooja Kulkarni and Suhasini Pai. Stem Cell Therapy in Autism Spectrum Disorders. Recent Advances in Autism. SMGroup. (In Press).
11. Alok Sharma Nandini Gokulchandran , Hemangi Sane , Pooja Kulkarni , Suhasini Pai , Vaishali Ganwir ,Perna Badhe. A case of autism showing clinical improvements after cellular therapy along with PET CT evidence. Journal of Stem Cell Research & Therapeutics. April 2017.
12. Alok Sharma, Nandini Gokulchandran, Pooja Kulkarni, Sarita Kalburgi, Shruti Kamat, Riddhima Sharma, Samson Nivins, Hemangi Sane, Perna Badhe. "Improvements in a case of autism spectrum disorder after cell therapy as noted on PET CT brain scan" SJSC. May 2017.
13. Sharma A, et al., The baseline pattern and age-related developmental metabolic changes in the brain of children with autism as measured on positron emission tomography/computed tomography scan. World J Nucl Med 2018;17:XX-XX.
14. Alok Sharma, Nandini Gokulchandran, Hemangi Sane, Pooja Kulkarni, Sarita Kalburgi, Ridhima Sharma, Perna Badhe, Samson Nivins. PET CT Scan Brain As A Monitoring Tool To Study The Effects Of Autologous Bone Marrow Mononuclear Cell Transplantation In Autism Spectrum Disorder. International Journal of Current Advanced Research. Sep 2017 (In Press).



## Chapter 2

# Intellectual disability

## 2.1 About Intellectual Disability

### What is intellectual disability?

“The brain, the organ that is responsible for your conscious experience, is the eternal prisoner in the solitary confinement of the skull and must rely on information smuggled into it from the sense the world is what your brain tells you it is, and the limitations of your senses set the boundaries of your conscious experience.(Coren, Porac & Ward, 1984, Sensation and Perception).

What if a certain limitation prevented you from learning something new, remembering what you learned, make a connecting inference, thinking and doing your way forward in a task. What if you find yourself unusually clumsy, always off beat and struggling with your social and daily life despite a good physical health? It is because of the limitation is affecting you in your intellectual functioning and in your adaptive functioning. Now the question is, is this limitation the disability?

Intellectual disability is a disability characterized by significant limitations both in **intellectual functioning** (decision making, conceptual thinking, abstract thinking, memory, reasoning, learning, problem solving etc.) and in **adaptive behaviour**, which covers a range of

everyday social and practical skills. This disability originates before the age of 18.

Previously it was known as Mental Retardation. The term intellectual disability covers the same population of individuals who were diagnosed previously with mental retardation in number, kind, level, type, duration of disability, and the need of people with this disability for individualized services and supports. Furthermore, every individual who is or was eligible for a diagnosis of mental retardation is eligible for a diagnosis of intellectual disability.

**Developmental Disabilities:** "Developmental Disabilities" is an umbrella term that includes intellectual disability but also includes other disabilities that are apparent during childhood. Developmental disabilities are severe chronic disabilities that can be cognitive or physical or both. The disabilities appear before the age of 2 and are likely to be life-long. Some developmental disabilities are largely physical issues, such as cerebral palsy or epilepsy. Some individuals may have a condition that includes a physical and intellectual disability, for example Down syndrome or fetal alcohol syndrome.

Intellectual disability encompasses the "cognitive" part, that is, a disability that is broadly related to thought processes.

IQ alone is not the criteria to diagnose ID: The evaluation and classification of intellectual disability is a complex issue. There are three major criteria for intellectual disability: significant limitations in *intellectual functioning*, significant limitations in adaptive *behaviour* and onset *before the age of 18*.

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

Other tests determine limitations in *adaptive behaviour*, which covers three types of skills:

Conceptual skills—language and literacy; money, time, and number concepts; and self-direction.

Social skills—interpersonal skills, social responsibility, self-esteem, gullibility, naïveté (i.e., wariness), social problem solving, and the ability to follow rules, obey laws, and avoid being victimized.

Practical skills—activities of daily living (personal care), occupational skills, healthcare, travel/transportation, schedules/routines, safety, use of money, use of the telephone.

AAIDD (American association of intellectual and developmental disabilities) publishes the most advanced scientific thinking on this matter in the 11th edition of its manual, *Intellectual Disability: Definition, Classification, and Systems of Supports*. In defining and assessing intellectual disability, AAIDD stresses that, in addition to an assessment of intellectual functioning and adaptive behaviour, professionals must consider such factors as community environment typical of the individual's peers and culture linguistic diversity cultural differences in the way people communicate, move, and behaviour.

### **What causes intellectual disability?**

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 and Heredity.

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 the details of few of the causes of ID.

Pre Natal Causes: Genetic disorders:

We classify the genetic disorder into three major groups

1. Single gene disorder
2. Multi-factorial Inheritance
3. Developmental disorder of brain function

### **1. Single Gene Disorder**

They are caused by mutation of one pair of chromosome, matched with a normal allele on the homologous chromosome or both of the chromosomes of a pair. They are called as single gene disorder or Mendelian disorder. For example, you may have heard of cystic fibrosis, sickle cell disease, Fragile X syndrome, muscular dystrophy, or Huntington disease.

Autosomal Recessive: Recessive autosomal disorder approximately accounts for one third of mendelian disorder. Autosomal recessive disorder has a recurrent risk. eg. *Phenylketonuria (PKU)* if untreated will lead to development disability.

Autosomal Dominant: Appears in every generation having the affected parent. Male and female likely to transmit equally to the offspring causing every generation with the disorder. Eg. Neurofibromatosis.

**X Linked Recessive:** The incidence is much greater in males to the female counterpart. The disorder is never passes from father to son. The genes are spread to affected males via their daughter becoming carriers. Eg. Duchene Muscular dystrophy.

**X linked Dominant:** The incidence with affected male with normal wife wherein all the daughters are affected and no son is affected. Eg. Vit D resistant Rickets.

## **2. Multi-factorial Inheritance.**

It is defined as the factor wherein the genetic and the non-genetic factor, each of it having a small effect. The risk is less in second degree relatives. Eg. Heart disease, obesity and other factors with environment hazards may leads to ID.

### **Genomic Imprinting:**

It challenges the fundamental assumption of Medelian Model. It states that genes may be differently inherited from the mother or father. These can be deletion of maternal contribution or paternal Contribution. Eg. Prader Willi and Angelman Syndrome conditions may be associated with sleep disturbances.

**Inborn Error of Metabolism:** Inborn errors of metabolism are rare genetic (inherited) disorders in which the body cannot properly turn food into energy. The disorders are usually caused by defects in specific proteins (enzymes) that help break down (metabolize) parts of food.

## **3. Development Disorder of Brain Function:**

Neural tube defects consisting of Spina Bifida, Anencephaly, Congenital Malformation caused by the failure in closure during embryogenesis leading to Intellectual disability. One factor that plays a role is folic acid deficiency leading to neural tube defect.

### **Environmental Influence:**

One factor that can be controlled is environmental influence like intrauterine malnutrition, toxic exposure, irradiation during pregnancy.

**Maternal Infection:** There is major medical advances to prevent maternal

infection. TORCH infections are a key to remember and take precautions for the same. TORCH, which includes Toxoplasmosis, Other (syphilis, varicella-zoster, parvovirus B19), Rubella, Cytomegalovirus (CMV), and Herpes infections, are some of the most common infections associated with congenital anomalies.

**Congenital HIV Infection:** Parent affected with HIV often lead to a child with affected nervous system as the HIV lead to the penetration of the central nervous system.

Toxic Substance: Ethanol and Drug exposure lead to teratogenicity and causes Fetal alcohol syndrome and others.

### **Perinatal Causes:**

Infections: Prenatal Infections like TORCH infections accounting for Neuronal Defects.

Problem at delivery: During delivery Asphyxia is the most common factor for brain damage especially in premature infants. Cerebral Hypoxic ischemia is a major cause of acute perinatal brain injury.

Other Perinatal Problems: Extremely low birth weight are at risk of Intracranial Hemorrhage and hypoglycemia that are the consequence of limited hepatic glycogen storage. Such problems result similar to asphyxia.

### **Post Natal Causes:**

Infections: Meningitis and encephalitis from bacterial and viral infection may lead to permanent brain damage with intellectual disability.

Toxic Substance: Lead poisoning is less common now but is important factor leading Intellectual Disability. Without recognition it may lead to ID, Intentional problems, Ataxia, and changes in personality.

Other Postnatal Causes: Road Traffic Accident, Brain Tumors are causes of permanent brain damages.

Psychosocial Risk: Poverty disposes the child to several developmental risks. These include teenage pregnancy, malnutrition, abuse, and deprivation. Physical abuse can lead to head trauma and permanent

physical disability may be association 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.

There are several problems specific to the assessment of ID persons. They are:

1. The persons with ID may have multiple sensory and motor impairments like loss of vision, hearing and deficits in gross and fine motor skills. These can substantially affects test performance and the resultant IQ score.
2. They may have severe delay in language development affecting their expressive and receptive speech as well as all forms of communication- verbal and non-verbal. Their comprehension of test instruction may be limited.
3. The presences of behavior problems like hyperactivity, aggressiveness, social withdrawal etc., make the child difficult to assess according to standard testing procedures.
4. Some of the individuals with ID have poor attention and high distractibility and hence testing will be difficult.
5. They may be poorly motivated and they may not be cooperative.

As there is no single test applicable to all ID persons with or without the associated problems mentioned above, the choice of the right tests becomes very important.

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 important areas of assessment in ID persons.

1. Measurement of the overall level of GENERAL INTELLECTUAL FUNCTIONING (IQ/ DQ/ SQ).
2. Assessment of ADAPTIVE BEHAVIOR.
3. Detailed analysis of INDIVIDUAL ABILITIES AND DEFICITS.

Psychological assessment encompasses assessment of the three major aspects of the mind namely, cognitive, affective and Conative. Maloney and Ward 1976 defined psychological assessment as a process of solving problems or answering questions. Psychological assessment involves understanding not only the causes of the problems but also potential solutions. The purpose of psychological assessment is to evaluate an individual or group of persons relative to a specific issue or problem. These may include intellectual functioning, learning disabilities, special abilities, scholastic achievement, personality functioning, emotional and social areas and questions of normality and abnormality. The Psychologist develops hypotheses based upon information of past behavior, present behavior and prediction for future behavior as defined by given situation incorporated in assessment information.

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 influenced by the adequacy or inadequacy of assessment procedures employed. The two major components of the most widely acknowledged definition of ID are a “significant limitation in both intellectual functions and adaptive functions”. The assessment of persons with ID is useful for as a guide to diagnosis, classification, and eligibility, determination, monitoring and evaluation.

### **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 of mental abilities in infants and pre-school children. It 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 is limited because of the problems of normative and constructive validity since these were concerned and standardized in highly industrialized societies.

### **3. Binet Era**

Modern intelligence testing began with Binet Era when Alfred Binet in collaboration 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.

### **4. 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.

### **5. Paper and Pencil Test**

A well known paper pencil test is Draw –A-Person test by Good enough. It is easy to administer and has shown correlation (.80) with other intelligence test. Pramila Phatak has developed norms for this test for Indian children.

Limitations of these type of drawing test in pre-industrial countries is lack of familiarity of children with paper and pencil, the use of this test with school children enrolled in formal education system is perfectly justified.

### **6. Cultural-Fair Test**

The quest for a culture-free instrument for measuring intelligence has long been there because available tests have limited use in studying cross-cultural intelligence. For this purpose, psychologist devised non-verbal, performance tests and called these as culture-fair test. The best known among the alleged culture-free test are: Raven's Progressive Matrices and Porteus Maze test.

It has now been realized that search for a culture-free test is elusive and not sensible. Culture has its effect on all skills, verbal and non-verbal. Infact, culture free test would mean devising an instrument based on experience common to all humanity and could be applied everywhere under the sun and universally valid.

Actually culture-free is contradiction in terms as intelligence itself is culturally determined. Hence, 'culture-free test means intelligence-free.

### **To Debate**

Does an IQ measure intelligence is often asked questions; but it is an inadmissible question. It is like asking 'Does thermometer measures temperature? By Temperature, We means, the scientific concept that which is measured by thermometer. Similarly, when IQ tests are properly constructed measures intelligence. Indeed in

a real sense, intelligence may be defined that which is measured by IQ tests.

Intelligence tests have been attacked both for general and specific limitations, based on both political and scientific grounds. Particularly, vehement have been the onslaughts against widely the IQ. Actually the opposition to IQ is mostly due to misunderstanding of what is an IQ. IQ, with whatever defects is still the most useful measure of intelligence available to us.

### **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.

Psychological methods: in recent years Psychological methods like use of reaction time and cortical evoked potentials (CEP) are used as objective methods for measuring intelligence. These methods are known to measure innate capacity, the phenotype aspect of intelligence. The basic premise of these methods is that there is a corresponding activity in the central nervous system (CNS) the way in which an individual responds to given stimulus. The CNS activity can be recorded by electrophysiological methods like cortical evoked potentials. These methods provide a new paradigm for assessment of intelligence.

There is a need for new; unconventional appraisal methods of the mentally retarded that would allow the assessment of the mental retardation require the outcomes of measurements to have direct implications for programmed planning.

Most of the existing methods of intelligence testing give only global and amorphous picture of the individual while a precise appraisal of paramount importance. Contemporary assessment methods should involve a different approach than that used a traditional practice. Rather than relying on measures of abilities inferred from the performance on tests, the contemporary approach should emphasize on direct assessment of actual competence. Contemporary methods require a distinction between 'product' and 'process' approach to assessment. In order to identify and measure that actual ability of persons with ID it is necessary to adopt a process approach of assessment which is useful for planning intervention.

**On the basis of IQ assessment ID can be classified as below**

<b>Mild retardation</b>	<b>Moderate retardation</b>	<b>Severe retardation</b>	<b>Profound retardation</b>
75% to 90% of all cases of retardation	~10% to 25% of all cases of retardation	~10% to 25% of all cases of retardation	~10% to 25% of all cases of retardation
Function at one half to two thirds of CA (IQ: 50 to 70)	Function at one third to one half of CA (IQ: 35 to 49)	Function at one fifth to one third of CA (IQ: 20 to 34)	Function at < one fifth of CA (IQ: < 20)
Slow in all areas	Noticeable delays, especially in speech	Marked and obvious delays; may walk late	Marked delays in all areas
May have no unusual physical signs	May have some unusual physical signs	Little or no communication skills but may have some understanding of speech and show some response	Congenital abnormalities often present
Can acquire practical skills	Can learn simple communication	May be taught daily routines and repetitive activities	Need close supervision
Useful reading and math skills up to grades 3 to 6 level	Can learn elementary health and safety habits	May be trained in simple self-care	Often need attendant care
Can conform socially	Can participate in simple activities and self-care	Need direction and supervision	May respond to regular physical activity and social stimulation

**Rehabilitative Management for intellectual disability**

Intellectual disabilities (ID, formerly mental retardation) have many medical causes. They manifest in different ways. The symptoms will vary according the medical cause. Depending on the origins of the disability, medical treatment can be helpful. However, medical treatment alone is inadequate to address the associated impairments and participation restriction.

As intellectual disabilities are not a disease, there are no curative treatments. Instead, interventions focus on identifying abilities and limitations. The goal is to provide sufficient supports that maximize each person's functioning and improve their participation. In this respect, we may speak of rehabilitation, rather than of treatment. In this section, we discuss the strategies used in supportive rehabilitation.

Developing an Individualized Support Plan (ISP):

The treatment plan is designed to correct, or limit, the harmful symptoms. However, intellectual disability (ID, formerly mental retardation) is not an illness. Therefore, a different type of plan is developed. This plan is called an *individualized support plan (ISP)*.

The goal of the ISP is

1. To assess the individual needs and abilities of each person.
2. To identify a strategic approach that maximizes functioning. This is accomplished by using strengths to offset limitations.

The main objective of the ISP is to optimize functioning and life satisfaction. The success of the ISP rests upon a collaborative alliance between the team, the person with the disability, and caregivers. The team members like the psychologists, social workers, nurses, doctors, speech, occupational and physical therapists, teachers, and service coordinators are just a few of the professionals who develop and execute the ISP.

Early intervention is crucial. This ensures the optimal development of children with ID. Some early intervention programs begin at birth. These early intervention programs serve multiple functions. Information to the caregivers about early child development, helping children grow and learn by working with their strengths and limitations and the role of the families and educating them with specialized techniques to help children function to the best of their abilities. Settings for these programs may include home, school, and community.

### **Individualized support plans (ISPs): Intellectual functioning & education**

The diagnosis of intellectual functioning (ID) is determined by a variety of

tests. These tests assess intellectual and functional limitations. It evaluates the abilities and limitations of the person. Based on this evaluation, the ISP identifies what additional supports are needed to ensure optimal functioning. Furthermore, the ISP determines the best method to provide these supports.

We discuss below in brief the rehabilitative management and the rehabilitation strategies based on the intellectual quotient, associated problem and the need for familial involvement for a success story to emerge.

## **2.2 Multidisciplinary management of ID**

Early intervention is key to a better tomorrow. Starting early intervention requires skill and identification requires expertise. Multidisciplinary management of ID consists of involvement of different professionals with a common goal.

### **Medical professional:**

Although there is no medical cure for ID, they deal with the underlying medical problems if any. Time to time scrutiny and monitoring the general health is a key for success in rehabilitation sciences. Better health status and prompt medical and surgical intervention is a must. Supplementary medicines like neuroprotective medicines and vitamins can be prescribed.

### **Rehabilitation Psychologist:**

Assess the developmental and social quotient and give guidance for better qualitative life. They assist parents and care givers to cope up with the ongoing disability and its rehabilitation. Motivation, Acceptance and prevention of stress and burnout all are essence to run the marathon of disability rehabilitation. They also play a key role in behaviour modification of the child, Cognitive enhancement, motivation, thinking, reasoning and problem solving, personality development in the child with intellectual disability. They also deal with sensitive and cultural restricted domains, Sexual education and management of hormonal based changed behaviour. They guide the parents to cope up with the ongoing changes. Assessment of Intelligence Quotient and guiding them

in participation, education and employment is one of the major roles which psychologist is a part.

### **Physical Therapist:**

They deal with motor and physical disability by assessing the motor and executive functions of the child and plan in accordance with what the child can achieve. They use various techniques to educate the muscles and provide assistance in physical independence. They also work on planning and providing assistive aid with other rehabilitation team members. They also play a key role in dealing with Neurological dysfunction in Perception, cognition, apraxia, and incoordination in the motor executive function like ataxia, dystonia, choreas etc. They use neurodevelopment treatment strategies, play based therapy, Strengthening, stretching, cognitive play, ergonomics, functional training a few to name to execute their goals. They play a key role in the preparation of the child in sports related training and participation in various social events.

### **Occupational Therapist:**

Assessment of the activity of daily living and routine functioning of the child, dealing with academic related functional problems are performed by the occupational therapist. They assist in training for difficulty in writing, eating and dressing skills. They improvise the participation of the child based on their school, home, play environment. Sensory integration, play therapy, use of splints and customising them, Cognitive play etc. assist in better participation. Skill training for better vocational status, Identification of hazards and proposed injurious activity are dealt by training and modification of the environment.

### **Speech and language pathologist:**

They deal with speech and language related problem by identifying the core impairment. The problem with hearing, Speech execution, Oromotor apraxia, Syntax error are few to name are dealt by them. Oromotor training, Muscle strengthening, sensory training, Post cochlear implant training etc are few which assist in better communication.



### **Orthotics and Prosthetics:**

Assistive aid like orthosis and prosthesis helps in maintenance and improvement in motor and functional tasks. It promotes independence in school and challenging environments. It is custom designed based on the need of the child and economic strength.

### **Special Educator:**

School education and preparation for work are performed by special educator in collaboration with the psychologist. Teaching the child using appropriate techniques and identification of their strength and weakness in education is must for integration. Inculcating self learning, teaching academic concepts using various educational methods, identification of teaching material appropriate to the child proves to achieve varied skills in short duration of time.

### **Vocational Instructor:**

They play a role to assist in vocational training and identification of jobs for placement. They train the children with ID with the skills that are prerequisite to get placed in an employment. Technology based assistance is also provided for independence.

### **Social Worker:**

They assist in sensitization of social problem by working at the grass root level. Identification and assistance play a major part in social awareness and rendering services.

### **Important Members:**

The rehabilitation field does not stop with the list above. Disability leading to ability is a societal issue rather than a personal problem. Assistance can lead to independence by creativity to easy accomplishment. From kitchen to walking out of the house technology plays a major role to provide comfort. The knowledge of medical field and expertise is not must to be a part of a rehabilitation team, it is rather being sensitised to social problems giving comfort to all results to the best solution. Today's advancement from dot watch to assistive talk technology all are results of a sensitised individuals.

## **Special Education in Intellectual Disability**

Assessments, Classification, Nomenclature and tagging should not be at the heart of any disability, Our goal is to understand the condition and to extend a helping hand to a fellow human to help him attain his full potential in life . It is only to get customised, tailor made, {deficit} specific action plans {IEP} for a needy individual. Hence the helping hand we offer to intellectually disabled child in Special Education is an IEP.

Special Education is “specially designed instruction, to meet the unique needs of a child with a disability. Every child is unique just because He/she is having a special need does not mar him of the right to education. Hence based on his/her degree of disability and the domain of limitation an individualised education plan is crafted customized for the child.

### **Why Special Education?**

In a regular set up the Core concept to be taught is the epicentre of the teaching learning process. However, in a Special education set up it is the Learner who is the Epicentre of the process. The Core content, Presentation, Product and even the evaluation is handpicked , based on the learner .

The Content is sliced into bite sized information bits to be chewed on rather than providing learning content in a big chunk. A Child With Special Need is dealing with an issue of inattentiveness and the Teaching Learning Material that are multi-sensory and varied in experience are more engaging than regular study materials and hence provides a better chance of an effective learning. Interesting use of pictures and ICT ensures enhanced learning.

When we take a close look at the common traits of C.W.I.D

1. There's a delay in reaching the developmental milestones
2. Difficulty in remembering things
3. Trouble in comprehending accepted social behaviour
4. Understanding the consequences to actions
5. Poor problem solving skills

The above traits challenge C.W.I.D. education. "They WILL learn, but it will take longer time" Special educator explores the therapeutic effect all modalities of life for learning both as a medium and a mode. Literature can... form the overall domain of Academics, binding both reading & writing and imagination. Music unfolds the domain of Listening, speaking and relaxing. Painting can guide on the important aspect of self-expression, on Fine & Gross Motor abilities. Acting can help child emote, express and communicate to others.

Dance movements in any form can... improve gait, posture issues and body balance. Hence the evolved ability of a special education Teacher makes him/her to break the fragments of an art form for example drama to various domains like, self expression, voice modulation, timings, interpreting, sequencing etc and provide fun learning opportunities.

Child with Intellectual Disability still continues to fight for basic rights, Disabled or typically developed, its every human's right and responsibility to make society more inclusive for everyone. All professionals working in the field of disability have very important goal of sensitizing global citizens the need to accept, appreciate and celebrate individual difference across degrees of disability and geographical barriers.

To explain more, let's take a simple example. Consider a child with moderate Intellectual disability, he or she may have problem with eating, dressing, lacing, toileting. Like may find difficulty in wearing shoe, difficulty in eating or expressing the need to go to toilet. When a child has problem with coping up his daily living activities there's no point of teaching regular academics.

Thus all these activities are focussed in special education. The main aim or target of special education is "Independent living" i.e to make the P.W.I.D has much as independent as possible.

Special education has this in mind and educates the children in various domains to improve their gross motor, fine motor, toileting, eating, dressing, functional academics, language including receptive and expressive, prevocational skills. Special education focuses on Functional academics which includes basic reading, writing, arithmetic, money transaction.

Based on the severity of ID, capability of the child the training is given appropriately. Each individual is unique and this is so true in P.W.I.D.

Example a child with mild ID may be performing all the daily living activities independently. So a special educator may focus on the functional academics for the child.

Whereas if the child is diagnosed with severe ID, then the necessary training needed for the child is in dialy activities.

Special education helps the child or the student or person with ID to use his or her image skills to improve their standard of living. When not given any special education the child's limited abilities present may gradually disappear as time proceeds.

Thus inorder to have an independent living and to improve the standard of living for a child with intellectual fisability, the child or adult should be trained in all the domains with patience, love, care and determination according to his or her own pace and that's is possible only through special education.

### **Psychological Intervention Strategies:**

Psychosocial needs of children with ID:

Psychologists in a rehabilitation team attend 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.

### **Diagnosis of psycho-motor and psycho-social development**

Assessment of normal development considering deficits in fine motor skills, perception social independence, interaction between mother and child aged 0 to 3 years of age, Neuropsychological needs (Neurological deficits assessed and attended).

### **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 behavioral state of the child
- Personality assessment
- Vocational assessment
- Behavioral assessments

The assessment processes differ from individual to individual based on the nature and severity of each child. The assessment process usually lasts for about 45 minutes and may demand multiple sessions. The final medical report includes the test results and relevant observations made during the examination.

### **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:***

- 1) *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 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 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.

Conventional treatments for ID like symptomatic medical management, surgical treatment and rehabilitation address only the symptoms but do not address the fundamental pathology of the disease. Children are still dependent of parents for almost all of their daily activities. As these children grow up they are institutionalized as it is difficult to care for them at home. Once these children stop the training and rehabilitation they may regress and lose all the improvements achieved with rehabilitation. Therefore there is a need for a treatment that can have an effect on the underlying disease physiology. So far, the disease pathology was considered to be a structural damage or underdevelopment, however with the advanced neuroimaging technology it has now been noticed on the PET-CT scan imaging that the metabolism of some parts of the brain is low causing intellectual disability.

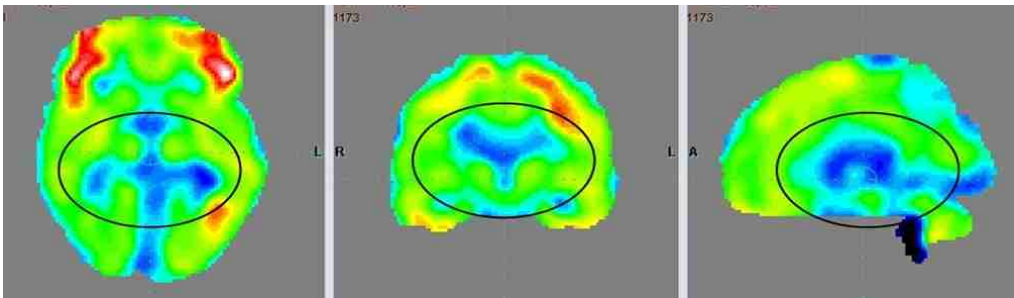


Fig. 2.1 PET-CT scan image showing blue areas of reduced metabolism and function in ID

This low metabolism is synonymous low function of that brain.

### **Stem cell therapy for Intellectual disability**

Because of the above mentioned unmet medical need, it is essential to develop a treatment that can alter the brain function on a cellular level. Stem cell therapy addresses the root problem within the brain in ID. Stem cells initiate the process of repair and regeneration by converting into brain cells and repairing the damaged areas. They release paracrine molecules such as growth factors, cytokines, chemokines, reduce inflammation and promote proliferation of existing stem cells. They also help to alter the immune responses of the body and protect nerve cells from damage. They promote formation of new blood vessels increasing the blood and oxygen supply to the brain. Stem cell therapy in combination with conventional treatment gives much better results as it

enhances the effect of conventional therapies and support the therapies by bringing about positive changes in brain tissue.

Stem cell therapy can improve following symptoms of Intellectual Disability

1. Low IQ
2. Social interaction
3. Cognition
4. Comman following
5. Attention and concentration
6. Recognition of family members

Improved function of the brain cells post stem cell therapy can be seen in the PET-CT scan. These changes often correlate to the functional improvements.

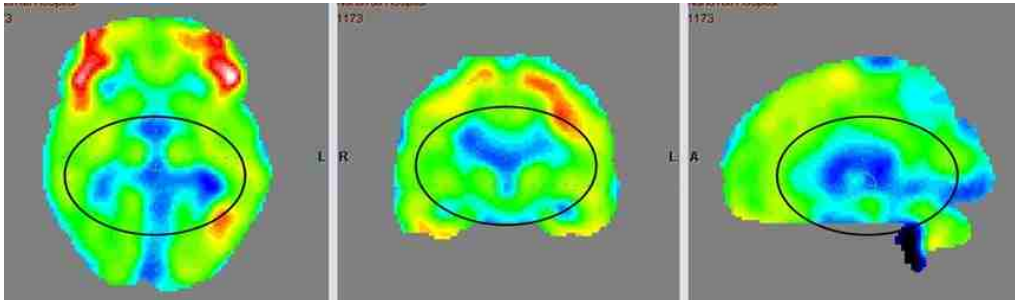


Fig. 2.6 PET-CT scan showing areas of reduced brain function in blue before Stem Cell Therapy

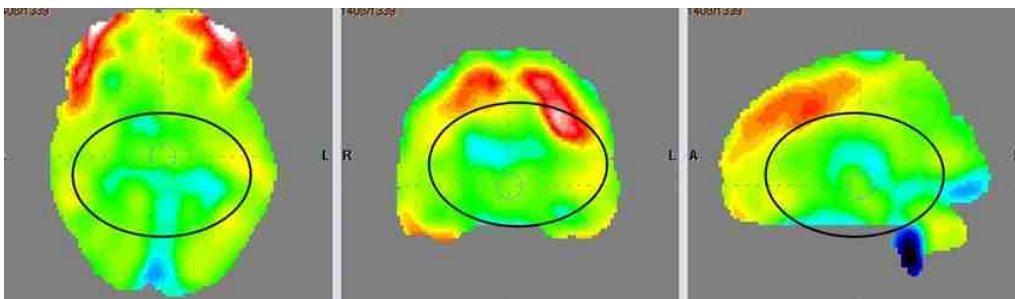


Fig. 2.7 PET-CT scan showing reduction in blue areas suggesting improved brain function after Stem Cell Therapy

## Chapter 3

# Learning Disability

Learning is a process that is consistent with a child's growth and development. When there is a deficit in learning, it calls the attention of parents and teachers. These are the children with Learning disabilities and it is one of the main causes for school failures in majority of children nowadays.

The term "Learning disability" was first used and defined by Dr. Samuel Kirk in 1963 in order to describe children who have serious learning problems in schools but do not fall under other categories of handicap. It is a neurologically based processing problem which can interfere with learning basic skills such as reading, writing, and/or math. They can also have difficulties in performing higher mental functions such as organization, time management, abstract thinking, attention and memory. Learning disability will not only affect a child's academics but also his personal and social life.

Learning disabilities is an umbrella term used to describe a set of impairments in the areas of acquisition and use of listening, speaking, reading, spelling, writing and reasoning skills.

**The different types of Learning disabilities include:**

- Dyslexia (difficulty in reading)
- Dysgraphia (difficulty in writing)

- Dyscalculia (difficulty in numbers and mathematical concepts)
- Dysnomia (difficulty in naming)
- Dysphasia (expressive language difficulty)

**Children with LD will display one or more of the following characteristics:**

- Perceptual problems
- Limited ability to abstract and generalize
- Difficulties with memory and attention
- Slow speech and language development
- Limited social skills
- Inappropriate/immature personal behaviour
- Limited attention span and poor retention ability
- Decreased motivation, Poor self-confidence and Low self esteem
- General clumsiness
- Lack of coordination of gross and fine motor skills
- Emotional disturbances

Multidisciplinary rehabilitation combined with newer treatments like stem cell therapy may help in treating the disorder at a cellular level. The rehabilitation techniques will manage the symptoms while stem cell therapy will address the core neuropathology of learning disability.

**Speech and Language therapy:**

A child with learning disability may have trouble in speech and writing. Problems can occur in the production, comprehension, and awareness of language at the sound, syllable, word, sentence, and discourse levels. Individuals with reading and writing problems may also experience difficulties in using language strategically to communicate, think, and learn. Thus, speech and language intervention should target both spoken language needs along with writing issues. The therapy activities used for a child with LD are always linked to the school work. Therefore, materials for treatment are taken from or are directly related to content that has been taught in class (e.g., school language textbooks for reading activities, essay writing or school assignments for writing activities). Social interactional skills and other pragmatic skills like topic initiation and topic maintenance also taught which will help the child in communicating with others more confidently.

## **Psychological Intervention**

Intervention for children with learning disability involves a review of all the aspects of child's functioning rather than one individual aspect. Therefore, the assessment involves cognitive, emotional, inter-personal, intra-personal, personal independence and self-care skills together. Typically, psychological interventions involve the development of Individual Educational plans and Individual Care plans. Psychologists also provide family support which involves working with the parents, teachers, guardians and siblings and it also involves enabling families to understand and provide for their affected family member's emotional and social needs and acknowledge the child's developmental level. It can be helpful to reduce the isolation that can sometimes occur with having a family member with a learning disability.

## **Special education**

Children with learning disabilities have limited ability to acquire, store, organize and use skills and knowledge. Their memory, auditory, visual, and linguistic information processing is also affected. Special educators help these children by introducing special teaching techniques, visual and memory aids, classroom modifications and use of technology. This may help the children in improving their learning.

## **Physiotherapy**

Children with LD may have other comorbid conditions in which their motor activities might be affected. Physical exercises may help in addressing these issues. Exercise may also help these children to stay calm and concentrate in their activities.

## **Occupational Therapy**

Occupational therapist is usually a part of interdisciplinary team and are involved in helping children with learning disability to adjust in different roles across life span. The intervention involves the patient, the environment in which he/she needs to perform occupations, their family members and significant others. It can be an individualized therapy, or in a group setting or can be in the same environment where he/she needs to perform the task. Occupational therapists mainly focus on ADLs,

learning, work, play and leisure, social participation, developing cognitive-perceptual skills, developing adaptive techniques and devices to improve learning and addressing other associated problems.

## **Nutrition**

Most children with learning disability have greater health needs than the rest of the population

### *Overweight*

It is generally observed that occurrence of obesity in adults with learning disability is higher than general population. Obesity is serious health problem it can lead to diabetes , high blood pressure. It is very important to maintain healthy weight.

Providing small and frequent meals with high fiber diet can help maintain weight

### *Underweight*

People with learning disability are also at risk of being underweight because of inability to ask for food if hungry, slow feeding, swallowing difficulties.

Calorie dense small and frequent meals can help gain weight.

### *Swallowing problems*

Some patients with learning ability and physical disability have swallowing issues.

Diet should be modified as per chewing and swallowing abilities.

Modified consistency diet like soft or mashed food should be included.

If swallowing soft food is also difficult then pureed food should be included.

- Avoiding artificial dyes help increase brains ability to focus
- Read Label carefully before purchasing any product.
- Also limiting intake of sugar will be helpful as sugar increases insulin levels and instant energy followed by drop in insulin level and dramatic



decrease in energy. This fluctuations can lead to hyperactivity and ability to focus.

- Include diet rich in fiber, protein and healthy fats.

### **Stem Cell Therapy**

As discussed above, LD is a neurological disorder. In simple words, the wiring of the brain is disturbed. Defect in myelination is also noted in LD, which affects the signalling and information processing in these children. Stem cell therapy helps in restoration of the myelin sheath and hence improving the information processing. This further helps in functional improvement.

## Chapter 4

# Down Syndrome

Down Syndrome is a genetic disorder resulting from an extra full or partial copy of chromosome 21. This gives rise to multiple systemic complications as part of the syndrome.

### **Symptoms:**

The syndrome is characterized by:

- short stature,
- a typical flat, broad facial structure, with smaller than normal low-set nose, small ears and upward slanting eyes,
- broad, short hands, and feet
- relatively short fingers
- decreased muscle tone
- overly-flexible joints and
- intellectual disability.

In addition, children with Down Syndrome may have a variety of birth defects such as:

- About half of the children may be born with a heart defect.
- Recurrent respiratory tract infections
- There may be in some children with Down syndrome, medical conditions such as gastroesophageal reflux and celiac disease in which the immune system is abnormally sensitive to gluten.
- About 15 % affected children have an underactive thyroid gland leading to hypothyroidism.
- There is also an increased risk of hearing and vision problems
- Delayed and slower development of speech and language
- Behavioral issues include attention problems, obsessive/compulsive behavior and stubbornness or tantrums
- A small percentage of children with Down Syndrome are also diagnosed with autism spectrum disorders with abnormalities in communication and social interaction.

**Diagnosis:**

- The presence of typical features as described earlier
- Chromosomal analysis to confirm diagnosis.

**Treatment:**

There are no medical treatments for intellectual disability associated with Down syndrome but, early intervention for infants and children can enhance their quality of life. Each child is unique and treatment depends on individual needs. Associated conditions must be monitored periodically as the child grows older. Depending on a child's needs, a multidisciplinary medical care may be needed. This may involve inputs from specialists including:

- Pediatrician to provide routine childhood care
- Child psychologist- to provide psychometric evaluations and interventions

- Cardiologist- Early examination is necessary for diagnosis and treatment of congenital heart defects.
- Gastroenterologist
- Endocrinologist
- Pediatric neurologist
- Ear, nose and throat specialist
- Ophthalmologist
- Speech therapist
- Physical therapist and Occupational therapist.

### **Occupational therapy**

Occupational therapist helps children with Down syndrome to master skills required across life span. They help them to become independent in selfcare skills, improve fine and gross motor skills, preparing for school and improving participation in play and leisure activities. They also help their families to develop goals that are appropriate to the child's capability. Occupational therapy is usually implemented immediately after the diagnosis has been made.

### **Occupational therapy intervention across lifespan**

- **Infancy:** occupational therapist may help the mothers by addressing the feeding difficulties secondary to weak muscles, low tone, lack of tongue movements etc. Occupational therapist may also provide training in using appropriate positioning techniques and may provide some devices to improving child's positioning.
- **Early childhood:** An occupational therapist may work on improving on his muscular strength, develop appropriate milestones, develop age appropriate participation in self-care activities, develop appropriate cognitive skills to help them initiate schooling at right age, etc. We may also help to address the sensory processing difficulties if any using sensory integrative techniques.
- **School aged children:** occupational therapist may help in improving

gross and fine motor skills, achieving normal motor milestones, improving participation in selfcare skills, developing necessary skills for school and if required providing them adaptations that will facilitate their learning. Occupational therapist can also be a part of school where they develop healthy routines, improving social interaction, develop peer interaction and other skills required to be independent in school setting. Occupational therapist also help in developing appropriate cognitive-perceptual skills and if required providing them with adaptive devices such as reading aids, writing aids, memory aids, etc.

- **Adults with Down syndrome:** Occupational therapists help them to develop skills required for prevocational or vocational training. They also help to do a comprehensive assessment to identify their strengths, weaknesses and their ability to perform task independently. They also do work simulation to identify which work environment is best suitable for them, whether they will need any supervision or can perform in unsupervised environments and in developing safe work environments.

### **Physiotherapy**

The main goal of physiotherapy for children with Down syndrome is to minimize the development of compensatory movement patterns. It is observed that children with downs syndrome compensate with hypotonia , joint laxity , reduced muscle strength and short limbs which if not addressed at the right time and allowed to persist leads to orthopaedic and functional problems.

1. Low muscle tone : hypotonia is very easily observed in children with downs syndrome. A child with downs syndrome appears floppy or like a rag-doll when picked up. Due to low muscle tone and reduced muscle strength leads to difficulty in learning gross motor skills .
2. Joint laxity : the ligaments holding bones together in the joints are longer than normal in children with downs syndrome which results into increased flexibility in joints, which is noticed primarily in hips, feet and shoulders.
3. Reduced muscle strength: reduced muscle strength is commonly seen

in children with Down syndrome, especially in muscles of trunk (abdominal and back extensors and hip muscles (abductors and glutes). Weakness of core muscles leads to use of compensatory movements. for example: when the child with Down syndrome is learning to sit they typically learn to sit with a posterior pelvic tilt, trunk rounded and the head resting back on the shoulders due to poor trunk muscle strength leading to kyphotic posture.

4. Short arms and legs: the length of the arms and legs is shorter than their trunk which makes it harder for children with Down syndrome to learn certain gross motor skills with correct recruitment of muscles. for example: short legs leads to difficulty in stair climbing.

Children with Down syndrome also have secondary health issues which may affect their physical skills as well:

1. Heart problems
2. Chronic upper respiratory tract infections
3. Ear infections
4. Stomach problems.

Physiotherapy goals: In our experience, children with Down Syndrome learn better with the use of visual learning (observation of somebody else doing or moving an activity) and kinaesthetic learning (someone guiding their arms, legs and body through the movement pattern or activity). Children will learn better if they practice frequently, for short periods.

*Visual/sight learning assistance*

- demonstrate specific movement or an activity
- reinforce your demonstration with clear instructions
- use picture cards or videos demonstrating similar movement/skill you are trying to teach

*Kinaesthetic/hands-on guidance learning assistance*

- use your hands to guide the child's body movements
- be careful while moving the child's hands/legs/arms in any awkward pattern -because you may reinforce wrong patterns.

### ***What can be done?***

#### **Babies and toddlers:**

Encourage achievement of gross motor skills such as sitting, crawling and standing.

- encourage independence in functional activities.
- improve muscle strength, balance and posture.
- prevent the risk of secondary joint problems in later life as a result of lax ligaments achieve early walking by " treadmill training"

#### **Older Children**

- Encourage more complex motor skills i.e. bike riding through inclusion in our bike riding groups
- Aquatic therapy also benefits to gain strength and endurance
- Work on more complex tasks to achieve motor planning

#### **Speech Therapy**

Children with Down syndrome have challenges in development of communication skills, including receptive language skills ex: understanding and expressive language skills Ex: speaking and composing sentences and reading and also may affect hearing, respiration, voice and articulation.

The SLP can help develop a comprehensive treatment plan to address all of the areas in which the child may be experiencing difficulty, including semantics (vocabulary), syntax (grammar), pragmatics (uses of language and social and conversational skills) classroom language skills, speech, oral motor planning and oral motor strengthening.

SLPs can work with families and teachers to design and implement an effective school, home and community program to help children develop stronger communication skills.

#### **Psychological Intervention**

Children with Down Syndrome have cognitive difficulties as well. As mentioned above, intervention involves a psychological assessment of the individual. Cognitive rehabilitation therapy is conducted which

attempts to enhance the functioning and Independence of people with Down syndrome. Psychologist conduct group therapy sessions which teaches then appropriate socialization skills. It would also provide a support system for people with down's syndrome. Another group training sessions can be conducted for the parents as this would provide them with an interactive forum where they could seek advice from professionals and other parents. These children are also given vocational training which prepares them for future jobs that are based on manual or practical activities. This training is non-academic in nature and prepares the individual to be completely independent and is totally related to a specific occupation or vocation.

### **Nutrition**

- Many children are mouth breathers due to smaller nasal passages, and may have difficulties coordinating sucking, swallowing and breathing while feeding.
- Excessive weight gain is a problem for many older children and adults with Down syndrome, mental retardation.
- lower resting metabolic rate of 10-15% further predispose weight gain.
- Deficiencies of vitamin A, vitamin B12, Vitamin B1 & B2 and vitamin C, zinc, selenium in individuals with Down syndrome & MR have been reported.

### **Common problems Associated with Downs Syndrome**

1. Thyroid Disorder
2. Diabetes
3. Infection
4. Celiac Disease
5. Constipation

#### *Gastro esophageal Reflux Disease (GERD)*

- Following are the things help reduce GERD symptoms
- Eat small, frequent meals
- Wait at least an hour after a meal to exercise
- Drink before or after meals, not during
- Do not lie down immediately after meals



### *Underweight*

For underweight Down's syndrome patient following things need to be included:

- Children require small, regular, calorie-rich meals
- Using calorie-rich household ingredients to add to foods will provide additional calories e.g. oil, butter, full fat spreads, cream, cream cheese, grated cheese, sugar
- Some children may require the support of a suitable pediatric nutritional supplement

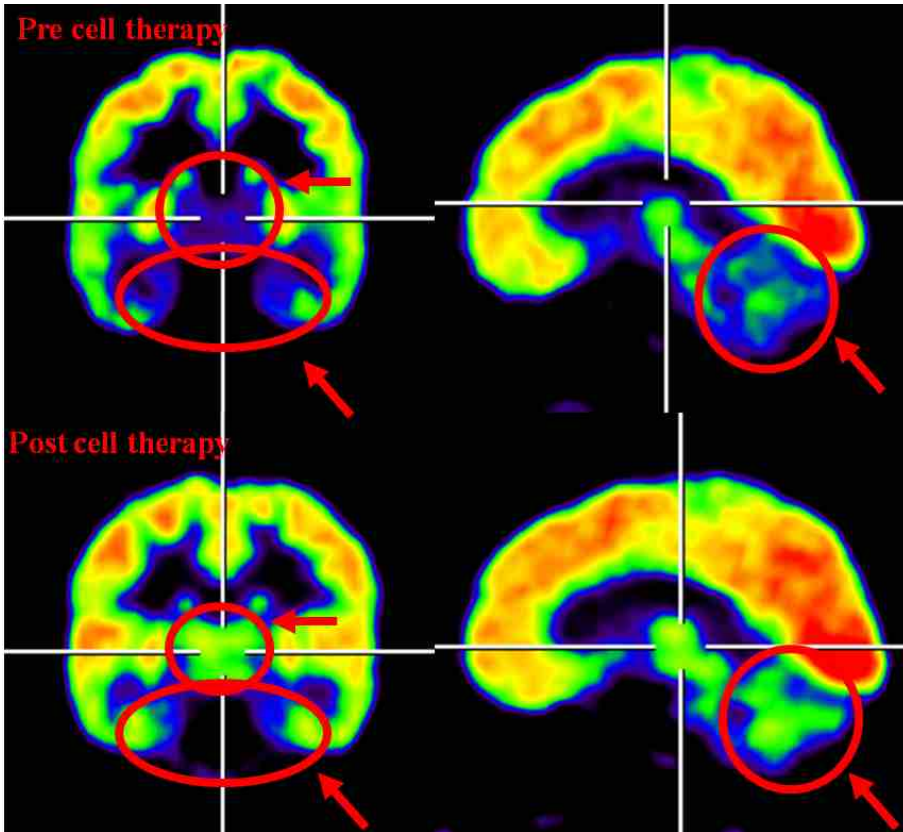
### *Obesity*

For obese patients with Down's Syndrome following things need to be included:

- Have small and frequent meals.
- Drink Plenty of Water Daily more than 2 liters.
- Include more fruits and Vegetables in your diet.
- Do not skip your meal.
- Chew food properly before swallowing.
- If having difficulty in chewing make food soft and give.
- Include protein rich foods like milk, curd, low fat cow's milk paneer, Whole pulses, soybean, sprouts, dal, almonds, and walnuts, chicken, fish.
- Include fruits in form of milkshakes, fruit salads.
- Include vegetables in form of veg parathas, mix veg khichdi, colorful vegetable salads, vegetable soup
- Plenty of fluids (soups, lime juice, buttermilk, coconut water) throughout the day to relieve constipation.

### **Stem cell therapy**

Stem cell therapy is a new therapeutic approach for Down syndrome. It addresses the underlying pathology of the disorder. Administration of stem cells may help in neurogenesis, angiogenesis and synaptogenesis which further helps in restoring the brain damage. It may help in improving the lost connectivity in the brain of children with Down syndrome, thus improving the information processing.



Representative Brain FDG-PET Coronal and sagittal images of a patient with Down syndrome. Before Stem cell therapy, hypometabolism is noted in the bilateral thalamus, medial temporal cortex and cerebellum. One year after Stem cell therapy, improved brain activity is noted in the bilateral thalamus, medial temporal cortex and cerebellum.

## Chapter 5

# Epilepsy

Epilepsy is a central nervous system disorder that causes unprovoked, recurrent seizures. A seizure is the outward manifestation of a sudden rush of synchronous and excessive electrical activity in the brain.

There are 2 main types of seizures:

- Generalized seizures affecting the whole brain. Depending on the subtype of generalized seizures, affected individual may present with staring, muscle stiffness, jerking motion of legs, arms or neck, or decrease in muscle tone causing the individual to fall, or loss of consciousness.
- Simple or Complex focal (partial) seizures affecting a part of the brain. This may present with jerking of body parts, emotional changes and changes in the way a person perceives light, sound and touch.

The most common seizures in children are partial complex seizures originating from the temporal lobe.

The cause of epilepsy is unknown. The possible causes include:

- Infectious diseases,
- Head trauma,
- Side effects of certain drugs and

- Genetic cause is suspected as well.

### **Diagnosis:**

Attacks of epilepsy in children are usually of short duration, which may complicate a timely diagnosis and referral to a specialist. Diagnosis may involve a series of tests including:

- A thorough evaluation including medical history by a neurologist.
- Electroencephalography (EEG) to study the electrical activity of the brain.
- Computed Tomography (CT) of the brain to detect tumors as well as effects of head trauma.
- Magnetic resonance imaging (MRI) and functional magnetic resonance imaging (fMRI).
- Positron emission tomography (PET) and single photon emission computed tomography (SPECT) that assess metabolic activity of the brain.

### **Treatment**

Medications can often prevent seizures from recurring. If, however, medications fail to control seizures, other options include vagal nerve stimulation and a ketogenic diet (high fat, low carbohydrate diet).

Most children with epilepsy have intelligence and abilities like other children. Some children may however have learning disabilities. This is normally due to a coexisting condition such as underlying brain abnormality or may also occur in children with frequent seizures. If a learning disability is identified, educational strategies must be implemented.

Behavioral problems may occur in which case discussing the issue with teachers and epilepsy counsellors may be helpful.

### **Physiotherapy**

Physical therapy is services provided by physical therapists to individuals and populations to develop, maintain and restore maximum movement and functional ability throughout the lifespan.

Physical therapy programs provide both physiological and psychological benefits for people with epilepsy. Various studies have proven the beneficial effects of exercise in epilepsy such as reduction in frequency of seizures in individuals who exercise compared to those who are physically inactive. A most accepted explanation to the neural benefit of physical activity can be attributed to neuroprotection. Animal studies have shown that exercise can reduce brain cell loss or neuronal damage in several animal models of brain damage.

Physical therapy helps in improving the quality of movement and physical performance by working on sensory awareness, response, improving strength, mobility, balance and coordination. Physical exercise also benefits individuals with epilepsy by increasing aerobic capacity, reducing heart rate, weight reduction and thereby reducing risk factors such as diabetes, hypertension, coronary heart disease, obesity, and osteoarthritis.

The psychological benefits of exercise include reduction in anxiety and depression, improvement in mental state, and better psychosocial adjustment. These benefits are attributed to the regulation of the neurotransmitter systems involved in depression pathophysiology and improvement in other comorbidities that impact depression.

Thus, depending on individual's specific deficits and challenges a physical therapy program may include -

1. **Stretching:** to lengthen and relax the muscles which have become tight due to continuous spasms during seizure activity, anxiety or generalized fatigue.
2. **Warm up:** sudden high/moderate intensity exercises may harm/injure the muscles, thus, a light warm up exercise program helps improve blood flow, flexibility, and prevents muscles injury.
3. **Muscle strengthening:** strengthening of weak muscles by active assisted exercises or graded resistance training depending on severity of weakness.
4. **Balance and coordination training:** A suitable training program devised by a therapist will help improve balance and

coordination, which could be impaired due to muscle weakness, impaired functioning of brain areas responsible for balance and coordination, lack of confidence, visual perceptual difficulties, fear of falls, lack of understanding, etc.

5. **Motor planning:** Motor planning is the ability to conceive, plan, and carry out a skilled, non-habitual motor act in the correct sequence from beginning to end, which tends to get affected in individuals with epilepsy. Thus a therapist may work on improving the same so as to maximize mobility indoors/outdoors, improve function, and increase independence.
6. **Gait training:** walking may get affected due to muscle weakness, loss of balance and coordination, fear of falls, etc; which may have to be corrected by training & retraining, building confidence, recommending appropriate aids/orthosis, etc
7. **Cardiorespiratory endurance training:** this may include aerobic activities, walking, running, challenging activities, etc
8. **Caregiver education:** regarding precautions such as safe, supervised driving, climbing stairs/slopes, kitchen work, handling power operated equipments, bathing, swimming, etc

Thus, physical therapy plays a vital role in treatment of manifestations and comorbidities in individuals suffering from epilepsy.

### **Occupational Therapy**

Epilepsy and its various types of seizures affects the physical, social, emotional, vocational and leisure activities of a person and their families. Its prognosis is not predictive and so may make a person with epilepsy anxious about his life and wellbeing. Occupational therapy helps a person to adapt, modify or improve on their skills to perform functional activities.

#### **Areas of concern for Occupational Therapist are:**

1. Basic Activities of daily living
2. Instrumental activities of daily living
3. Leisure

4. Roles
5. Routines
6. Habits
7. Physical issues
8. Sensory issues
9. Cognitive-perceptual issues
10. Education and vocational choices
11. Quality of life
12. Family or caregiver guidance

**Basic activities of daily living:** people with epilepsy usually become dependent for self-care activities due to risk of fall, or families may become over protective. In such conditions, patient and their caregivers can be motivated to allow them to participate in basic self-care activities in safe environments such as:

- Well ventilated bathrooms
- Maintain water temperature used for bathing (not too warm or too cold)
- If possible pad the bathrooms to avoid any sharp edges
- Put anti-skid mattresses in the bathroom.
- If required provide adaptive tools to accomplish the task of bathing, dressing etc. such as long-handled scrubber, soap on rope, modified handled brush to help grasp the brush well etc.
- The bathroom should be well lit

**Instrumental activities of daily living:** Due to fear of seizure they usually avoid getting involved in other activities of daily life such as cleaning, grocery shopping, etc. which may increase burden over their family members or caregivers. They may also feel dependent as they cannot get involved in such activities. In such a case following things can be done to improve their participation:

- The therapist may discuss the areas that patient wants to get involved in and can find out which are safe for them to perform.

- Therapist may also suggest performing some of the activities under supervision.
- Therapist may suggest modified ways to perform these tasks or with some type of adaptations if required.
- Some of the activities the patient may not be able to perform or may not be safe for him to perform, so they may have explore alternate activities for improving participation.

**Leisure:** it is one of the important area where an occupational therapist looks into. Most of the people with epilepsy avoid getting involved in different leisure pursuits as it poses the risk of increasing the seizures frequency. Occupational therapist discusses about different leisure interests and advices them about the safest activities to get involved in how they can get involved in leisure pursuits without harming them.

**Roles:** we also help to participate in safe and skillfully way in their different life roles such as a mother, student, homemaker, etc.

**Routines:** Most of the people with epilepsy have disturbed routine patterns. They may get involved more into non-productive activities (like resting) and reduce their productivity in life. We help them to develop a productive routine and improve their participation in meaningful activities for example a student need to attend school regularly, has to study on regular basis, should have appropriate sleep rest pattern, they may be advised to avoid getting involved in physical games, etc.

**Habits:** it is always advised to have good dietary intake, have god amount of sleep, avoid exertion or being exhausted, avoid activities that require a lot of physical activity, avoid substance use etc. It is important that a person with epilepsy is aware of these and importance of adhering to these healthy habits.

**Physical, sensory and cognitive perceptual issues:** People with epilepsy are always at risk of having post-epileptic encephalopathy, which can affect the different cortical functions. An occupational therapist will then advice different therapeutic activities to improve a person's physical, sensory and cognitive perceptual functions based on their detailed assessment. They may also advise use of some orthotic device or may fabricate one to help improve function or prevent secondary effects.



**Educational and vocational choices:** it is also important people with epilepsy lead a meaningful life and they have some aspirations in life. Occupational therapist may help assess their abilities and skills to help them make appropriate and safe career choices.

**Quality of life:** Irrespective of the condition, it is important that the person and their family members lead a quality of life. Occupational therapist may advice certain modifications or provides adaptations that may reduce the burden on the caregivers and the person with epilepsy thus improving their satisfaction and quality of life.

**Family and caregiver guidance:** It is very important that the families are aware about the condition and its implications. It is especially important in Indian scenario that the families are counseled and guided well at each step. They should be explained about the importance of taking regular medicines and at regular intervals. At times, the families may become over protective which may further hamper the development of the person with epilepsy. Therefore, an occupational therapist may help educate the families about the seizures and how to manage a family member with epilepsy

### **Psychological intervention**

Increasingly, epilepsy is being viewed as a disorder that carries a high risk of comorbidities and cognitive and psychosocial ramifications. Children with epilepsy are at increased risk of cognitive deficits or regression, neurodevelopmental disorders, anxiety and depression, of experiencing low self-esteem, of suffering from the stigma attached to having epilepsy and academic underachievement. Focal epilepsy affecting the dominant hemisphere may impede the development of language-related skills such as reading and spelling. Memory deficits may lead to a loss of confidence in social settings and feelings of inadequacy. The unpredictability of seizures and feelings of lack of control and helplessness may invoke anxiety. Disrupted sleep patterns may result in lowered energy while defensiveness can lead to a need to conceal, anger and bitterness. Diagnosis of epilepsy in their child also leads to stress in parents. Research studies have found lower parent-child relationship quality, higher rates of depression in mothers and problems with family functioning. Parents may be overprotective through fear of injury or

death. Families may harbour misconceptions about epilepsy and may become socially isolated due to concerns about adverse public reactions. To cope with epilepsy, in case of cognitive impairments, cognitive retraining, memory rehabilitation and setting of realistic educational goals may help children deal with the cognitive difficulties. Medication and behavioral interventions also help like encouraging a positive self-image. In addition, some children may benefit from cognitive behavioural therapy (CBT) and other interventions aimed at reducing social anxiety and developing social skills. Psychotherapy has been used to alleviate strong emotional feelings and stigma associated with having epilepsy and to assist children in dealing with psychosocial stresses and emotional conflicts. Identifying factors that might precipitate seizures, learning how to avoid these, trying to interrupt seizures in the early stages of their occurrence and to practicing relaxation and breathing exercises may assuage the psychosocial impairments caused by epilepsy. The attitudes and understanding of families should be a prominent part of epilepsy management as it will greatly influence how children cope. It can be accomplished by establishing good communication channels between the families. Information and psychological and social support for the families may go a long way in allaying fears. Greater recognition of the wider impact of epilepsy and the accurate assessment and treatment of cognitive and behavioral problems is needed to enable the development and targeting of appropriate services and support and enhance the quality of life in childhood epilepsy.

### **Speech-language Therapy**

Children with epilepsy have delays with both speech and language skills but more impairment is noted in speech as compared to language.

The Speech and language symptoms seen in children with epilepsy are as follows:

- They start communicating in words at a later age.
- Delay in formulation of sentences to communicate.
- Difficulty in following commands/ instructions as compared to their peers.
- Need more time and effort to learn new concepts and words.

- Time taken to develop literacy skills is more as compared to peers.
- Difficulty in articulation/ correct acquisition of speech sounds.

### **Speech - language therapy:**

Children with epilepsy have Oromotor weakness or difficulties. Thus, to build oral sensory awareness and improve strength of oral musculature the use of Oromotor / facial exercises and PNF exercises are used which include the following:

- **Oro motor exercises** include, massaging the cheek muscles and muscles around the lips with the help of fingers.
- **PNF exercises** without the use of vibrator which includes massaging the inner oral cavity i.e. the inner cheek muscles and tongue with a finger brush/ Z vibe without vibrator/ NUK brush and also improve oral sensory awareness.
- **Blowing Exercises** – Blowing candles, paper bits, bubbles and whistle to train the facial muscles to produce certain sounds and to improve residual lung volume capacity/ breathe support and breathe control.
- **Breathing Exercises** – Facilitate abdominal breathing by doing inhalation and exhalation exercise to strengthen the diaphragmatic muscles and improve breath support for speaking longer phrases.
- **Jaw Exercises** – Use of chewy tubes {knob shaped, P and Q (XT)} to improve jaw mobilization and eating foods that require extra chewing, like apples and carrots, to strengthen jaw muscles; practicing opening and shutting their mouth using only the jaw muscles while someone else holds their chin.
- **Lip Exercises** – Using a lollipop, squeezing their lips around it to increase strength and lip closure, pursing their lips to kiss a lollipop to improve lip extension.
- **Tongue Exercises** – Strengthening the tongue by sticking it out and pushing it against a tongue depressor/ ice cream stick/ spoon for seconds at a time.

**To improve articulation skills of the child through Articulation Therapy** using a mirror and phonetic placement technique is used i.e. use of flash cards and word lists to help focus on specific sounds and encouraging children to make sounds while looking in the mirror to help them understand how to place their tongue at specific positions in the oral cavity to produce specific sounds.

**Language therapy:**

- At first, children with epilepsy need intensive speech and language stimulation at home as well as other environments. They slowly learn to observe their surroundings and attain speech and language skills.
- Their receptive and expressive language skills are delayed as compared to their peers and thus they cannot acquire them naturally as children of their age do. To improve their expressive language skills one needs to work upon the receptive language skills first i.e. understanding or comprehension skills.
- To work upon that one needs to identify their areas of deficit and their exact language age. Children mostly need to be worked upon with the help of Play therapy.
- To improve receptive language skills the receptive vocabulary of the child needs to be increased. Receptive vocabulary includes identification of all common objects in the environment, lexical categories such as fruits, vehicles, animals and vegetables, verbs, prepositions and other grammatical forms necessary to develop communication skills. It also includes following commands and instructions
- Once the child starts identifying the words included in the receptive vocabulary, the clinician can work towards producing those learnt words verbally first through speech imitation and later through spontaneous speech.
- Once the child learns speech imitation and starts speaking one word spontaneously the clinician needs to work upon increasing the length of utterance to phrases/ sentences. This can be done with the

help of showing action verb cards or using other games for language development. Gradually we increase the length of utterance from 2 word phrases to 4 - 5 word phrases and later on to complex sentences. If the child does not develop higher language skills at a later stage then the clinician can work upon them.

- They also need special attention at school to develop literacy skills.
- There may be a lot of episodes of regression in speech - language skills due to epilepsy at regular intervals but that depend on the severity and extent of the epileptic attack. So in such a case the clinician or parent will have to begin the training again from the core.

## **Nutrition**

- Nutrition plays role in Epilepsy patients especially in uncontrolled seizures.
- The most specialized diet given to these patients is Ketogenic diet.
- The classic ketogenic diet is high fat low carbohydrate diet which help to control seizures in some patients.
- This diet produces ketones in body as fat is utilized by body as energy source instead of carbohydrate.
- A ketogenic diet "ratio" is the ratio of fat to carbohydrate and protein grams combined.
- A 4:1 ratio is more strict than a 3:1 ratio and is typically used for most children.
- A 3:1 ratio is typically used for infants, adolescents, and children who require higher amounts of protein or carbohydrate for some other reason.
- Some foods that provide fat for the ketogenic diet are butter, heavy whipping cream, mayonnaise, and oils (e.g., canola or olive).
- Amount of carbohydrate and protein in the diet have to be restricted therefore it is very important to prepare meals carefully. No other sources of carbohydrates can be eaten.

- The ketogenic diet is supervised by
  - a. a dietician who monitors the child's nutrition and can teach parents and the child what can and cannot be eaten
  - b. a neurologist who monitors medications and overall benefits
- This diet strictly should always be followed under supervision.

**Section B:**  
**Management of children**  
**with Physical disability**





## Chapter 6

# Cerebral Palsy

### 3.1 About Cerebral Palsy

#### **What is Cerebral Palsy (CP)?**

Cerebral Palsy (CP) is a leading cause of childhood disabilities in India. 3 in every 1000 births are diagnosed with CP. Most of the children with CP have severe disabilities. The parents may find themselves hopeless and helpless after finding out the prognosis of the condition. CP does not affect the child alone, it affects the whole family and therefore whole family needs to participate in the care and rehabilitation of these children. CP requires a family centered care, not just a patient centered care. The primary focus of this chapter is creating awareness and guide parents and caretakers of children with CP. Understanding about CP by the parents and caregivers will empower them to provide best care to their child. Knowing the underlying causes or risk factors may help them to prevent further complications.

CP is a movement disorder causing inability to perform movements due to lack of muscle strength, muscle co-ordination or excessive tightness of the muscles. The muscle impairments and movement limitation is due to poor development of brain in the womb or damage to the brain tissues because of lack of oxygen or blood at birth or trauma or infection within 1 year of birth. CP mainly affects movement and development of motor

tasks like use of hands, sitting, walking etc. but in many cases it can affect the understanding and cognitive abilities of the child as well depending upon the areas of the brain damaged.

CP is not progressive, meaning the brain damage does not increase over time but it is permanent. It cannot spontaneously heal itself. Symptoms of CP can worsen as time lapses because of secondary factors like outgrowth of bones and body in comparison to the muscle development and movement control. Therefore management of symptoms depends largely on how well a child receives medical, nursing and rehabilitative care. To provide best care, it is important to understand symptoms of CP and their implications. Although there are broad categories of symptoms of CP, no two patients are the same. There is a different combination of symptoms and their intensities in each child. The impairments experienced by these children range from normal intelligence but mild difficulty in walking or balancing to severe cognitive and physical debility leading to complete dependence for all activities of life. Every child with CP needs tailor made management program that changes as the child grows. The earlier this program starts the better is the outcome.

### Types of CP

Because there is a wide spectrum of symptoms in CP, CP is divided in different types based different body parts involved and difference in the movement abnormality.

#### *Based on body parts involved there are 5 types*

1. Quadriplegic where all 4 limbs are affected, this is the most common and most severe form often accompanied with cognitive impairment as well.
2. Diplegic both the legs affected.
3. Triplegic in which both the legs and any one arm affected, this is a rare type of CP.
4. Hemiplegic in which arm and leg on one side are affected.
5. Monoplegic in which any one limb affected it is a rare and mild form of CP.

### ***Types of cerebral palsy based on the area of the brain damaged***

Different areas of the brain not only control different body parts but also different aspects of movement. Some areas are responsible for causing appropriate tension in the muscles so that we can move a body part, whereas some are responsible for balancing the tension in different muscles groups for timely smooth movement. Some areas are responsible for co-ordination of the body movements with visual, auditory and other sensory cues.

#### **1. Spastic (Increased stiffness in the muscles)**

The word spasticity relates to increased tension in the muscles. The muscles need a certain amount of tension to work efficiently. If this tension increases then it causes the parts of the body to be rigid and stiff and difficult to move. Children that exhibit such stiffness in their body parts are categorized into spastic cerebral palsy. These children are prone for deformities and care should be taken to prevent the same.

#### **2. Dyskinetic (Abnormal movements of the body)**

In this form the brain is unable to co-ordinate the actions of different muscle groups and therefore the tension and force in the muscles during movements keep changing. This leads to various abnormal, uncontrollable movements in the body. These abnormal movements are of different types based on the quality of the movement and tension in the muscles.

#### **3. Ataxic (Clumsy movements and poor balance)**

This type is characterized by in-coordinated movements and loss of balance. It is difficult to perform precision movement especially with fingers. The balance while walking is poor, sometimes sitting balance is also poor. There could be continuous shaking of head. Speech could be slurred.

#### **4. Hypotonic (Floppy muscles)**

Hypotonia is opposite of spasticity. In spasticity there is increased tension in the muscles but in hypotonia there is reduced tension in the muscles. The muscles are flail. Movements are sluggish. This could resemble

muscle weakness. Child may find it difficult to perform a movement and hold a posture. There is chance of severe spine deformities and compromise of joint integrity in this form due to flail muscles that can be overstretched and therefore fail to provide stability.

## **5. Mixed**

Mixed cerebral palsy is when the symptoms of two or more of the above types co- exist. This form of CP is rarely observed and is difficult to diagnose. The management can also be complicated.

### **Symptoms of cerebral palsy**

Cerebral palsy is a disorder with a diverse clinical picture the signs and symptoms vary depending on the type and severity of the brain damage. The signs of cerebral palsy are the effects that can be observed and tested by the clinician. These signs are observed right from the early childhood and infancy.

#### ***Early signs of cerebral palsy***

- At birth a baby with cerebral palsy may seem flaccid and floppy.
- Child may not cry immediately after birth and his skin may turn pale and blue.
- The child may develop difficulties in feeding, sucking and swallowing.
- Child may choke or gag often. These difficulties may persist for months.
- The child may cry excessively and may seem very irritable or not cry at all and seem abnormally quiet.
- The child may show slower or delayed development of standing, walking etc. compared to other children.
- Child's body may stiffen when carried, dressed, washed, or during play.
- It may be difficult to carry the child as the body is too flail or stiff.
- The child may not be able to hold the head properly and it may fall

behind or on the sides.

- The arm and leg movements of the child are diminished and the child may find it difficult to grip the toes or his hands.
- Arms and legs may twitch uncontrollably.
- The child may show some signs of impaired communication. The child may not smile upon looking at the mother. The child may not respond when his name is called.
- The child may not be able to use both his hands together in a coordinated manner e.g use both the hands to grip toys etc.
- There may be excess floppiness or stiffness, or lack of arm gestures, or control of face muscles. The child may not be able to point at things that he needs due to these impairments.

In older age variety of symptoms like spasticity in limbs, joint deformities and muscle contractures, lax joints, inability to sit, stand or walk, poor walking balance, poor sitting balance, abnormal posture, abnormal movement patterns, abnormal gait patterns, cognitive deficits, inability to control bowel or bladder, poor appetite, constipation, poor vision, squint, poor hearing capacity, slurred speech, inability to speak etc. may be seen.

### **What causes of Cerebral Palsy?**

CP can be caused due to many factors that may affect development of the child in womb before birth, brain damage due to complications during birth and trauma to brain after birth. In addition to some of the definite causes that have been identified, there are some risk factors meaning the child is at a risk of developing CP but may or may not develop CP in presence of these.

### **Prenatal causes and risk factors (Before Birth)**

CP can be caused due to abnormal brain formation and underdevelopment of brain structures or abnormal chromosomes causing genetic defects. Infection to the mother, Intra-uterine infection to the foetus, certain medicines administered to the mother, ingestion of substances that can cause birth defects, injury to fetal brain.

### **Perinatal causes and risk factors (At birth)**

Trauma to the brain during delivery, lack of oxygen supply after birth, lack of blood supply, prematurity, Low birth weight, delayed or no birth cry, multiple births, infection to the brain.

### **Post natal (After birth) causes and risk factors**

Traumatic brain injury, infections, epilepsy i.e. seizures or fits, jaundice, hydrocephalus, brain tumors may cause CP.

### **How is Cerebral Palsy diagnosed?**

The diagnosis of cerebral palsy is confirmed based on the signs and symptoms exhibited by the child and results of various diagnostic imaging techniques like Magnetic Resonance Imaging (MRI), Computed Tomography (CT) and Electroencephalography (EEG).

The most common basis of diagnosing cerebral palsy is delayed development of normal physical and cognitive function. This is determined on the basis of observations by parents, birth history and developmental history.

In addition a detailed physical examination is carried out by the doctor to evaluate ability to perform various motor tasks, variations in the tone of muscles, impaired coordination and presence of abnormal involuntary movements. Clinician will also check for the social smile, response to name and may test some other aspects to understand cognitive growth.

### **Special tests:**

MRI and CT scan

If there are any abnormalities identified in the clinical examination and history clinician will then ask for special investigations to identify the cause and the area of brain damage. These investigations include MRI and CT scan. These scans help to look at the brain to identify or rule out any injury to the brain or structural anomalies of the brain. This is important for diagnosis as well as to decide further course of action, if the child needs any surgical intervention to correct the structural anomalies.

## PET-CT scan

Positron emission tomography- computed tomography scan is a recent development. It is a nuclear medicine technique which shows function of brain cells and therefore helps to identify parts of brain that are not functioning optimally.

## **3.2 Multidisciplinary management of Cerebral Palsy**

### **Medical management of Cerebral Palsy**

Medical management of CP is limited to the secondary complications and symptomatic management for the muscle tone. There is not curative medicine or medicine that promotes brain growth in CP. Medication can be used to alter the tone of the muscles and reduce associated spasms. The options are oral baclofen, tolperisone, tizanidine, clonidine, benzodiazepines, chlorzoxazone. These medications are used either single or in combination to achieve a good control. L-Dopa (and other dopaminergic drugs) may be of additive value to lessen spasms in selected cases. Whenever a drug appears to be useful, it is worth tapering the dosage down from time to time so that the patient can help reassess continued benefits. Some children may not tolerate particular drug. In these situations, changing drug or reduction in doses needs to be considered. For other abnormal body movements associated with spasticity drugs like tetrabenazine, trihexiphenydy, clonazepam can be added. Side effects to the medications may be excessive floppiness, sedation, drooling or excess salivation, vomiting, hypotension etc and should be reported to your doctor. In addition to these medicines, boltulinum toxin (Botox) injections can be given to specific muscles but their effect may be short term and the injections must be accompanied by rehabilitation for optimum benefit. In addition to these vitamin and protein supplements can be prescribed if poor nutrition of the child is noted due oro-motor impairment.

### **Surgical management of CP**

Surgical management of CP consists of Neurosurgical management and Orthopedic surgical management.

**Neurosurgical management:** In the immediate post-natal period in case of any malformations or structural abnormalities of the brain, neurosurgical correction may be required. One of the common cause of CP is hydrocephalus or increased fluid accumulation in the brain. This fluid accumulation need to be corrected with a surgery which inserts a shunt in the brain draining this fluid into other parts of the body. In the later stages neurosurgery can be used for correcting spasticity.

**Orthopedic management:** In the later stages of CP where bony deformities and postural deviations are evident, orthopedic surgeries are used to correct the bony alignment and to prevent muscle tightness. These surgeries are crucial to the function of a child with CP. Tendon release surgeries which are used most commonly to avoid contractures post spasticity can be performed. For ankle joint calf muscle tendon release is performed to correct foot posture, for knee joint hamstring muscle tendon release is performed to prevent knee flexion contracture giving the ability to stand erect, for hip joint adductor muscles are released to prevent indrawing of the legs and provides the ability to stand with legs apart. Different tendon release surgeries can be performed for upper extremity in order to prevent contractures of wrist, forearm and elbow. The surgeries must be accompanied with a long term regular rehabilitation to give maximum benefit and functional improvement.

## **Rehabilitative management of Cerebral Palsy**

### **Physiotherapy**

Physiotherapy plays an important role in managing a child with cerebral palsy. Physiotherapy mainly addresses motor issues. It is a key therapy to develop motor milestones of sitting, standing and walking etc. Physiotherapy exercises are aimed and stretching the tight muscles, reducing spasticity, improving posture and co-ordination to achieve independent movements. The aim of physiotherapy exercises varies based on the different types of CP.



**Physiotherapy goals is different for different types of cp:**

**1. Spastic:**

- Relax stiff muscles
- Prevent deformities
- Inhibit compensatory positions
- Inhibit over use of movement muscles

**2. Dyskinetic child:**

- To control the abnormal or unwanted movements.
- Control of abnormal posture.
- Improve coordination
- inhibit hyperrecruitment/ over recruitment of muscles
- Inhibit burst of movements

**3. Ataxic:**

- Improve balance
- Improve trunk control
- Reduce wide base of support
- Develop transitions/disassociation
- Help walk and stand steady
- Control unsteady movements

**4. Hypotonic CP:**

- Provide support in good position
- Strengthen muscles
- Achieve neck control
- Co-activate postural muscles

With this book, we aim to guide the reader with different type of handling techniques. Knowledge about different handling and relaxation techniques will help achieve the best alignment in the child. These techniques will improve the alignment of different joints and will prevent joint deformities and contractures. We aim to guide parents and empower them to prevent the secondary complications arising form CP.

**Physiotherapy treatment aims to:**

- Encourage normal movement as much as possible.
- Follow developmental stages.
- Encourage use of both sides of body.
- Improve posture.
- Strengthen muscles.
- Improve range of motion of all the joints.
- Stretch tight muscles.
- Improve exercises tolerance and endurance.
- Reduce secretions through chest physiotherapy to help keep lungs clear and strengthen respiratory muscles.
- Improve balance in both sitting and standing.
- Teach or develop transitions, example: supine/lying to get up, sit to stand, half kneel to stand etc.
- Teach usage of adaptive devices.

**Different therapies used by therapist to treat children with cerebral palsy are:**

1. 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.

2. Patterning: it is a concept based on the theory that typical brain development can be facilitated in the brain injured child by passively repeating the sequential steps of typical development. Failure to normally complete one stage of development impairs the development of subsequent stage. This approach is laborious and also requires multiple sessions every day. Parents and caregivers are trained to carry the procedure at home also.
3. Constraint-induced therapy: this therapy aims to improve the use of affected hand in child with hemiplegia (paralysis of one side of the body). The normal hand is plastered for couple of days, to improve the use of affected hand.
4. Hippotherapy: It is a technique of horseback riding which has shown improvements in muscle tone, posture and balance.
5. Pediasuit: This technique involves a number of different elastic cords tied to specific points on one end of patient's body to different points on the other end. This device helps to move the patient move independently while controlling their movement as well as strengthening different parts of the body.

### **Demonstration of some of the physiotherapy techniques**



Fig 3.1 Elongation: to stretch the tight muscles



Fig 3.2 Alignment: to correct the wrong posture



Fig 3.3 Activation: to strengthen the correct muscle required for performing functional activity

**Home exercises which can be easily carried out by parents, illustrated with pictures**

**Relaxation techniques**



Fig 3.4 Relaxing child on lap

- Lay your child on your lap and Make sure that the arms are forward and not curled up under the body.
- With your hands gently press the bottom.
- This technique helps to relax child's body, and is always a good exercise to start with



Fig 3.5 Relaxing the child

- Hold the child on their side with their back against your stomach.
- Put one hand under the shoulder, one on the hips.
- Hold the shoulders and slowly twist the lower part of the body away from and towards you



Fig 3.6 Relaxing the child on the swiss ball

- Keep the child's arms forward and place your hand on their legs to steady the child on the ball. Keep the legs apart.
- Gently stretch one leg out and then the other.
- Carry on until you feel child is relaxed.



Fig 3.7 Relaxing in the sitting position

- Make your child sit on your lap/bolster facing away from you.
- Give support to both the arms to maintain alignment.
- This will help to develop base of support on feet.

### Lifting and Carrying techniques



Fig. 3.8 Carrying child from hips

- Carry your child, with their legs around your hips.
- Support them under their hip.



Fig 3.9: Carrying child in an aeroplane position

- Carry the child with your arm between their legs and your hand under the chest .
- Keep their arms and legs turned out.

### Prone activities



Fig. 3.10: Lying on upper extremity

- Encourage your child to lie on their front to play and to take weight on their upper extremity.



Fig. 3.11: Lying on upper extremity with head lifted up

- Encourage your child to lie on their stomach.
- You can place a toy in front to encourage your child to lift-up their head.



Fig. 3.12: C4 Encourage Weight Shift Using Play

- Encourage child to shift weight n take object.

### Sitting activities



Fig. 3.13 Encouraging head control when sitting

- Sit with your child on your lap facing you.
- Place your hands behind their head and shoulder .
- Encourage your child to look at you , encourage to Talk and play.
- 



Fig. 3.14 Encouraging activity in sitting position

- Encourage child to reach for toys to either side and to keep their balance.
- This exercise helps to improve sitting balance and strengthens stomach and back muscles.





Fig. 3.15 Encouraging upright sitting

- Encourage child to sit on CP, in upright position.
- Make sure the hip and knee are in 90 degree position, the hands are well supported.
- The head also should be well supported, if the child has poor head and neck control.



Fig. 3.16 Encourage forward weight bearing

- Support your child at their shoulders.
- Bring their body weight forward over their feet.

**Encouraging standing:**



Fig, 3.17 Encouraging activity in sitting position

- Encourage them to take as much of their weight as possible and to balance themselves by providing good base of support.
- Encourage your child to stand up against a rod or table.
- The height should allow your child to take weight through their hands.

### **Positioning**

Sitting: Child with delayed sitting will need more support. They will require support for a longer time than other children. They may require a special chair to help them to sit in a good position, so that they can use their hands better and chew properly. After the child learns to sit, you should progress to standing.

*Correct position to sit:*



Fig. 3.18: Correct position to sit

- Head should be slightly forward.
- Back should be straight and child should not lean on either side.
- The bottom should be against the back of chair.
- The knees and foot should be at the same level.
- Feet should be well rested or supported on the footrest.

### Positions to discourage while sitting:



Fig. 3.19: Incorrect position to sit

In the above two diagrams the position the hip isn't correct. You cannot initially sit strap the child at the thigh and back.

### 2. Standing:

Children with delayed standing should benefit from standing supported in a good position. Standing helps to encourage muscles that hold the body up to work. This position is the best to prevent contractures and also helps to make the leg muscles strong.

- The body should be in straight line and feet flat with equal weight on both the legs. The body hips, knees and feet should be in same alignment.
- If the child cannot stand and the body bends or he cannot keep heels on ground, you can use a forward lean stander.



Fig. 3.20: Correct position to stand

### **Stretching:**

- It is important to have Child's cooperation for stretching.
- Position the child in such a manner so that you can avoid abnormal position or movement.
- Hold the limb in stretched position and stretch very gently for a count of 20.
- Repeat the stretch and you can stretch a little more if the child is relax.
- Do stretching twice a day, once in morning and other in the evening.
- Stretch very slowly and gently, without causing any pain.
- Do not move the joints to and fro, stretch slowly, this may increase the spasticity , you have to allow the child's joints to relax.
- Avoid stretching during sudden stiffening or uncontrolled involuntary movements.
- Do not over stretch the joints.

### **Common stretches which you can do at home**



Fig. 3.21 Stretching of hip adductor

- Keep the child in straight lying position
- Bend both the knees
- Hold the inner part of the thigh gently, as shown in fig L2 and L3



Fig. 3.22 Stretching of knee flexors



Fig. 3.23 Stretching of knee flexors

- There are two ways of performing this stretch.
- Keep the child on their stomach, with both the knees outside the bed , gently pull one foot downwards , as shown in figure.
- Keep the child in straight lying position, ask someone to hold one leg, raise the other leg which is to be stretched a little high , keep one hand at the knee and with other hand gently raise the foot above , as shown in figure L2.3.

Stretching of foot muscle:



Fig. 3.24 stretching of calf muscles

- Figure shows foot position, when there is foot deformity.
- Keep child in straight lying position, hold the foot at the heels and gently stretch the foot upwards.

Important things to remember for the parents

- Do regular rehabilitation.
- Correct posture while feeding.

- Body alignment in sitting
- Body alignment in carrying by parents
- Correct wheelchair selection and positioning
- Hip X-ray after every 6 months
- Screening for visual/hearing deficit atleast once
- Regular dental checkup

### **Occupational therapy**

#### Occupational Therapy Goals

- Teaching handling techniques to the parents
- Positioning of the child
- Normalization of muscle tone
- Prevention of contractures and deformities
- Use of splints and adaptive devices
- To improve cognitive functions of the child
- Facilitating normal development of the child
- Developing fine motor and functional skills and play behavior
- Improving strength of the upper body and shoulders
- Home and environmental modifications

The common troubles usually observed in children with CP are spasticity; it can lead to contractures secondary to long standing spasticity. Dystonic children have difficulties in regulating their body tone. In children with ataxic CP, difficulties are seen in doing coordinated movements and maintaining the balance that affects the daily routine and their school performance.

Spasticity can lead to contractures of fingers. The common problems observed are cortical thumb (Thumb in hand), difficulty in holding and releasing the objects and handwriting difficulties.

OT intervention for children with CP focuses mainly on the following aspects.

### **Normalization of tone**

Various facilitatory and inhibitory techniques can be used for normalization of the tone. It includes joint compression, weight bearing, rocking, stroking and prolonged stretching.

### **Splints**

Splints can be given for preventing, positioning and functional use.

### **Improving muscle strength and improving Proximal stability**

Improving the strength of the UL and core muscles can help to improve the functional performance of the child.

Activities like walking on hands, crawling, overhead activities, ball throwing, ball catching, reaching activities in kneeling and half kneeling can help to improve the strength of the upper limbs and their core muscles.

### **Bilateral Hand Co-ordination**

Facilitating meaningful and focused activities by using both hands of the body.

Eg. Rolling the clay and drumming.

### **Fine motor skills**

Teaching the child to hold small objects and giving the child an opportunity to hold objects of different shapes and sizes.

It is very important to develop the hand functions like grip, grasp and pinch.

### **Activities to facilitate midline crossing and trunk dissociation**

Because of spasticity and dystonia, children can have difficulties in performing activities that involve 'crossing of midline of the body'. It affects the daily routine activities like eating, bathing, dressing and self-care. During therapy sessions, activities like transferring pegs from left side to the right side, drawing figure of eight in the air can be given to facilitate the midline crossing.

### **Children with sensory issues:**

Few children with cerebral palsy may also have sensory issues. In order to tackle the sensory issues, sensory integration therapy can play a very key role.

### **Cognitive training:**

- It can be started with improving the basic cognitive skills.
- Improving the self and body awareness through play activities like body part puzzles.
- Improving the orientation of the child to time, place and person through play and meaningful activities.
- Use of watch (to improve orientation with respect to time), flashcards/pictures of different places (to improve orientation with respect to place), family photographs (to improve orientation with respect to persons) to improve the basic cognitive skills.
- Making a daily schedule to improve awareness about the routine tasks.
- Giving activities through play and other functional activities to improve skills such as decision making and problem solving.

### **ADL modifications:**

Use of Adaptive devices can improve the functional independence and quality of life. Some of the examples of such devices are as follows:

- Adapted pencil grips
- Modified seating devices
- Modified toilet seats
- Feeding devices
- Bathing devices – Modified shower, lever operated taps.
- Modified toothbrush

### **Home and environmental modifications:**

- The house should have good accessibility to a wheelchair.



- Increasing the width of the doorways
- Removing the passages and keeping enough space for the turning radius of wheelchair near the toilet and washbasin.
- Adjusting the height of the seat as per the height of the wheelchair.
- Designing the furniture as per the height and reach of the person using the furniture.
- Installing handrails near the staircase.
- Installing grab bars on the toilet wall.

### **Use of assistive technology**

For children with CP, assistive technology like electric wheelchairs, voice amplifiers, modified key board, adapted switch board, electric shaving machine and electronic toothbrush and communication devices like iPad, tablets and computers can make a lot of difference in improving the quality of life of these children.

### **Aquatic therapy**

Aquatic therapy is defined 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” by The Aquatic Therapy and Rehabilitation Institute. In simple terms aquatic therapy means making use of different properties of water to facilitate functional recovery and independence of children with CP.

### **Beneficial effects of water immersion**

#### **Heart and lungs**

Water exerts compressive pressure on the blood vessels and pumps up the blood from limbs to the heart. In children with cerebral palsy because of inactivity or muscle weakness or low muscle tone the blood circulation may be sluggish. This causes the blood to be pooled in the lower 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, as the tone of the muscles alters this relaxation is difficult. Incomplete relaxation leads to exhalation of the air from only the upper parts of the lungs and accumulation of air 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.

### **Muscles and bone**

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. 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 of the muscles. Joint compression combined with increased blood supply help in reducing the tone of the muscles. Viscosity of water helps in strengthening the muscles. Improved circulation helps improve flexibility and pliability of the muscles.

### **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 nerve signals that are responsible for increased tone of the muscles. 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 in cerebral palsy.

### **Benefits of aquatic therapy in cerebral palsy**

- Sustained reduction in spasticity
- Improved muscle and movement co-ordination
- Improved oromotor control
- Improved respiratory capacity
- Better flexibility of the muscles
- Improved walking patterns
- Improved eye – hand co-ordination
- Improved muscle strength
- Improved cardiovascular endurance
- Regularization of the sleep patterns
- Reduction in the abnormal involuntary movements
- Reduction in the sudden spasms of the muscles

### **Speech therapy**

Children with CP exhibit variety of oro-motor impairments that lead to difficulty in speech, swallowing and communication. Oral motor functioning depends on an intricate process of sending and receiving messages to various facial, throat and neck muscles to coordinate breathing, talking, chewing, swallowing and digestion.

Speech and language pathologists improve communication and speech, but also improve swallowing and digestion.

Speech and language therapy aims to improve, and make possible, a child with Cerebral Palsy to effectively communicate their thoughts and ideas to the world. Communication skills are a vitally important aspect of interacting with others, developing relationships, learning, and working.

Speech therapy increases a child's potential for independence and positively impacts their quality of life.

CP is of two types depending on the site of lesion/ brain injury:

- Pyramidal (Spastic)
- Extrapyramidal (Non spastic): it further includes Ataxic CP and Dyskinetic CP

Dyskinetic is further divided into: Athetoid and Dystonia.

The intervention strategies used to improve speech and language skills of children with CP will vary based on the type of CP and whether any additional impairment is present e.g. Hearing loss.

**Speech and Language difficulties faced by children with Cerebral Palsy (CP):**

- Children with cerebral palsy often have communication difficulties.
- They have delayed speech and language development.
- They have Dysarthria (a motor speech disorder) that makes it difficult for them to speak clearly due to poor muscle tone and lack of in-coordination in oral facial, neck and throat muscles.
- They also have difficulty articulating sounds correctly due to muscle weakness or spasticity.
- Some children might also have hearing loss, which can further complicate communication and contribute to speech and language delays.
- They also have poor respiration and speech coordination due to reduced breath support/ reduced residual lung volume and weakness/ spasticity of the thoracic and abdominal muscles.
- They have all the oral peripheral mechanism (OPM) functions affected which include both vegetative (blowing, sucking, chewing, biting, swallowing, etc.) and non-vegetative (speech) functions.
- Due to hyper/ hypo sensitivity of the oral structure they have inappropriate oral sensory awareness and due to these issues of drooling, tongue thrust, etc arise.

The role of an SLP is to help children speak clearly, communicate effectively, and control the muscles involved in speaking, eating, drinking, and swallowing. They further have the responsibility of building a child's vocabulary, listening skills, interpretation, or capacity to communicate through non-verbal means.

## **Psychology**

Many people with Cerebral Palsy struggle with psychological issues especially if the person is unable to ask for assistance, socialize like others or has difficulty in communicating their needs. Psychologists implement Cognitive- Behavioral Approach which teaches a child to interact with their environment and gain control over their emotions. Once a person learns to communicate appropriately s/her is able to enjoy daily life activities, involve more with family and peers and this increases their quality of life. They use adaptive equipment and communication devices which is especially designed to meet their specific needs. Psychologists also work with the physical therapists to encourage the person to exercise by rewarding desired outcomes and discouraging negative behaviors and thoughts. Individual counseling sessions are provided by a Psychologist to help them overcome with feelings of Inadequacy and meaninglessness. These feeling are substituted by building their own values, self-worth and accomplishment. Apart from this, Psychologists actively work with the caregivers and family members and guide them regarding how to manage their emotional and aggressive issues if any, and how much level of exposure is required to increase their quality of life and overall development.

## **Nutrition and Cerebral Palsy**

Many of the children with CP are found to be under nourished or mal-nourished. Oromotor impairment like facial muscle weakness and lack of control may lead to poor nutritional status, growth failure, low weight for stature, chronic aspiration, esophagi is and respiratory infections in children with CP.

Across the cerebral palsy spectrum, poor nutritional status is caused by distinct pathways ranging from following:

1. Inadequate intake, i.e. inability to eat enough food this could be due to inability to chew and swallow or lack of appetite in view of immobility.
2. Most common gastrointestinal problems are Gastroesophageal reflux and constipation.
3. Children with CP can have sensory difficulties that make feeding difficult. They may be overly sensitive to touch in and around their mouth and face. The child may avoid the touch of food, bottle teat, as spoon or even hand unpleasant. This may cause the child to bite down, turn away, refuse to open the mouth or even gagging or vomiting.
4. Some children with oromotor difficulties and gastroesophageal reflux are at risk for recurrent aspiration, which can lead to pulmonary disease.
5. These children are unable to feed themselves and may require extensive use of assisted technology and are dependent on others to feed them. Care giver burden is a significant concern as the feeding process may require considerable time and may be associated with stress and fatigue which in turn may affect feeding of the child.
6. Lengthy and prolonged bottle feedings of milk and juice promote the decay of the primary upper front teeth and molars.
7. Children may find it very difficult to swallow especially liquids

Most of the children that receive multidisciplinary rehabilitation show functional improvements. These improvements are due to prevention or alteration of the secondary effects. Rehabilitation therapies make use of the plasticity of the brain to improve functional independence and quality of life but have no effect on the damaged parts of the brain. The brain damage persists because of which once the children stop rehabilitation they may regress in their improvements and develop secondary complications like muscle contractures, postural deviations, increased spasticity and loss of functional independence. Therefore, there is a need for a treatment that can repair the damaged brain.

### **Stem cell therapy**

Stem cell therapy replaces the damaged brain tissue with healthy tissue, reduces inflammation, improves blood supply to the brain, protects the

existing brain cells and improves the metabolism of non-functional cells.

Following stem cell therapy, children with cerebral palsy show holistic improvement in various areas of neurodeficit.

The first and the most prominent improvement is seen in the balance and trunk control. Children who do not have neck control or sitting balance generally improve within 3 months of stem therapy and intensive rehabilitation. Improvement in muscle tone, such as reduction in the spasticity of the upper limb and lower limb, with improved voluntary control is noted. This helps to improve the child's over all functioning and ability to do various activities such as neck holding, siting, kneeling, standing and walking.

Improvement in oromotor activities is also observed as the child can swallow better and is able to move his tongue as well as jaws better. This leads to improvement in chewing and reduction in drooling. Improvement in articulation (speech clarity) and vocabulary is also seen. Overall improvement in cognition, understating and comprehension adds up to the holistic improvement in the child. After stem cell therapy, neuroimaging studies such as PET CT scan show the improvement in the function of the brain. Brain metabolism improves significantly after stem cell therapy which also correlates with clinical improvements.

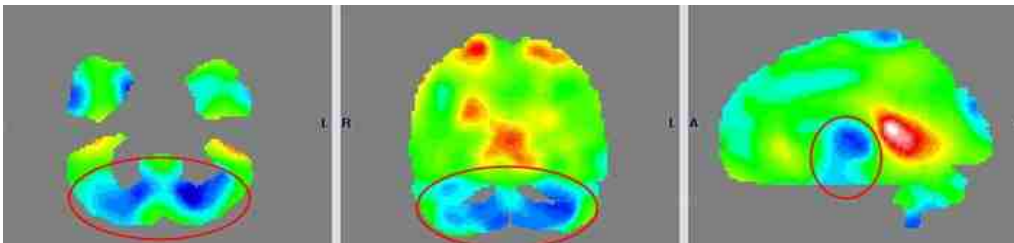


Fig. 3.29 PET-CT scan showing areas of reduced brain function in blue before Stem Cell Therapy

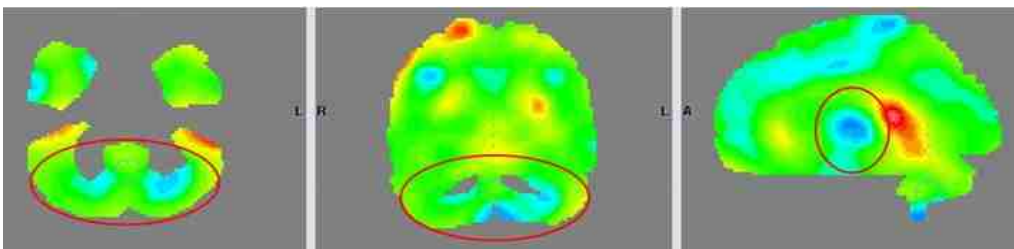


Fig. 3.30 PET-CT scan showing reduction in blue areas suggesting improved brain function after Stem Cell Therapy

### **List of Publications by NeuroGen BSI in cerebral palsy**

1. Alok Sharma, Hemangi Sane, Nandini Gokulchandran, Pooja Kulkarni, Sushant Gandhi, Jyothi Sundaram, Amruta Paranjape, Akshata Shetty, Khushboo Bhagawanani, Hema Biju and Prerna Badhe. A clinical study of autologous bone marrow mononuclear cells for cerebral palsy patients: a new frontier," Stem Cells International, Volume 2015, Article ID 905874, 11 pages.
2. Alok Sharma, Hemangi Sane, Amruta Paranjape, Nandini Gokulchandran, Pooja Kulkarni and Anjana Nagrajan, Prerna Badhe. Positron Emission Tomography – Computer Tomography scan used as a monitoring tool following cellular therapy in Cerebral Palsy and Mental Retardation – A Case Report. Case Reports in Neurological Medicine. Volume 2013, Article ID 141983, 6 pages.
3. Alok Sharma, Hemangi Sane, Nandini Gokulchandran, Prerna Badhe, Pooja Kulkarni and Amruta Paranjape. Stem Cell Therapy for Cerebral Palsy – A Novel Option. Cerebral Palsy. Challenges for the future. 2014: 217-242.
4. Alok Sharma, Hemangi Sane, Pooja Kulkarni, Myola D'sa, Nandini Gokulchandran, Prerna Badhe. Improved Quality of Life in a Case of Cerebral Palsy after bone marrow mononuclear cell transplantation. Cell J. 2015; 17(2):389-394.
5. Dr. Alok Sharma, Ms. Pooja Kulkarni, Dr. Hemangi Sane, Dr. Nandini Gokulchandran, Dr. Prerna Badhe, Dr. Mamta Lohia, Dr. Priti Mishra. Positron Emission Tomography- Computed Tomography scan captures the effects of cellular therapy in a case of cerebral palsy. Journal of clinical case reports. 2012 J Clin Case Rep 2:195.
6. Alok Sharma, Tongchao Geng, Hemangi Sane, Pooja Kulkarni. Clinical neurorestorative progresses in cerebral palsy. Journal of Neurorestoratology 2016:4; 1-7
7. Alok Sharma, Hemangi Sane, Suhasini Pai, Pooja Kulkarni, Meenakshi Raichur, Sarita Kalburgi, Sanket Inamdar, Nandini



- Gokulchandran, Prerna Badhe. Intrathecal administration of autologous bone marrow mononuclear cells in a case of Cerebral Palsy coexisting with autistic features". *Phys Med Rehabil Int.* 2017;4(1):1110.
8. Sharma A, Sane H, Kalburgi S, Kulkarni P, Bhagwanani K, et al. Autologous Bone Marrow Mononuclear Cell Transplantation with Neurorehabilitation for Cerebral Palsy. *J Stem Trans Bio* 2017; 2(1): 110
  9. Dr. Alok Sharma, Dr. Nandini Gokulchandran Mrs. Suhasini Pai , Ms. Pooja Kulkarni , Dr. Hemangi Sane , Dr. Khushboo Bhagwanani ,Dr. Prerna Badhe. Diplegic dystonic Cerebral Palsy treated with cellular therapy: a case report. *Journal- International Journal of case studies* 2017

## Chapter 7

# Duchenne Muscular Dystrophy

### 4.1 About Duchenne Muscular Dystrophy

#### **What is Duchenne Muscular Dystrophy (DMD)?**

Duchenne Muscular Dystrophy (DMD) is a progressive disease in which muscles gradually weaken making it difficult to perform daily activities. The disease is caused due to absence of protein 'Dystrophin' from the muscles. Dystrophin is important to strengthen the cell wall of muscle cell and protect the muscle cell from the contractile stresses. In absence of this protein the muscle cells are vulnerable and easily breakdown during muscle contractions used while performing daily activities. Once the muscles cells are damaged those are unable to heal and therefore are replaced by non-contractile scar tissue or fat. Because of this muscles lose their ability to contract and become weak. It is a fatal disease in which life span of the children is significantly reduced.

Usually the symptoms are noticed between the ages of 3 to 5 as difficulty of getting up from the floor. However symptoms like mild delay in achieving the milestone of walking, swollen calves, and inability to match the physical performance as that of peers are often missed by parents.

When the stem cells i.e. cells repairing damaged muscle cells and forming new muscle cells are exhausted muscle weakness becomes apparent and symptoms are visible. Muscle weakness usually sets in first in the larger muscles of lower extremity followed by smaller muscles of the lower extremity, neck muscles, trunk muscles, upper extremity muscles and finally respiratory muscles. Weakness of respiratory muscles is the cause of breathing difficulty and inability to breath which is the cause of death in most of the children with DMD.

Symptoms vary based on the age and stage of the disease.

Symptoms of Duchenne Muscular Dystrophy as the disease progresses are,

1. Difficulty getting up from the floor, positive Gower's maneuver. (Figure1)
2. Toe walking
3. Walking with increased lumbar lordosis
4. Frequent falls while walking
5. Swollen appearance of calves
6. Progressive deformities of ankle, knee, hip, forearm, elbow and spine
7. Difficulty of getting up from the chair
8. Difficulty climbing stairs
9. Inability to stand or walk
10. Inability to perform overhead activities
11. Inability to bend elbow
12. Easy fatiguability
13. Inability to lift up the neck while lying supine
14. Inability to sit up from supine position
15. Poor balance in sitting
16. Inability to sit without support
17. Spine scoliosis
18. Difficulty of breathing

### **What causes of Duchenne Muscular Dystrophy?**

DMD is genetic disorder. It is caused due to absence or mutation of dystrophin gene. This gene is responsible for production of protein

dystrophin which strengthens muscle cell wall. What causes absence of this gene is not known.

Gene abnormality can be inherited from one generation to other. Males are affected by this disease. Females can carry a faulty but are not affected by the disease therefore are called as carriers. There is a 50% chance that son of carrier female develops the disease and daughters inherit the faulty gene to become carriers. Sometimes this genetic deletion or mutation can occur spontaneously without mother being a carrier.

### **How is Duchenne Muscular Dystrophy diagnosed?**

DMD is diagnosed on the basis of clinical symptoms and some special investigations like serum levels of Creatine Phospho Kinase (CPK), Electromyography(EMG), Musculo-skeletal Magnetic Resonance Imaging (MRI-MSK), Muscle biopsy and Genetic testing. The accurate and confirmatory diagnosis is provided by genetic testing.

**Serum CPK testing:** CPK is an enzyme that is released in blood when there is muscle injury. In DMD lot of muscles are injured and therefore the levels of this enzyme in blood are very high. These can be checked with a simple blood test. The test is only indicative of muscle damage and cannot determine the type of muscular dystrophy.

**Electromyography:** EMG is a test that assesses the response of the muscles to nerve stimuli. It is conducted by inserting needles in various muscles and passing electric signals. If the muscles are damaged they generate a characteristic pattern. The test can only identify dystrophic pattern but can accurately diagnose the type of muscular dystrophy.

**Muscle biopsy:** In this test a small piece of muscle is taken using a needle under anaesthesia and it is then analyzed under microscope. The microscopic features of dystrophic muscles, presence of fat and scar tissue in muscles can identify muscular dystrophy but cannot accurately diagnose the type of muscular dystrophy.

**Musculoskeletal Magnetic Resonance Imaging:** 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 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, muscular dystrophy can be diagnosed and severity of the disease is determined.

Genetic testing: It is a specialized blood test. Blood is drawn and then a genetic testing is performed to check for 72 exons in the dystrophin gene, deletion of a single or cluster of these exons can confirm the diagnosis of DMD.

## **4.2 Multidisciplinary management of Muscular Dystrophy**

### **Medical management of Duchenne Muscular Dystrophy**

Although DMD has been identified as disease for over a century and its pathophysiology and cause has been identified, there is no medical cure for the disease till today. Being a genetic disorder chemical molecules used as medicine are not effective and are unable to correct the underlying pathology. The cure for DMD lies in correction of the genetic defect that can be achieved with various techniques of genetics. There has been limited success to these techniques and several factors limit their efficacy. The medicines that are available at the moment target mainly the after effects of the genetic defect and help in either preventing the complications or delaying the disease progression.

#### **Medical treatment aims at:**

- 1) Slowing down the disease progression
- 2) Increasing the period of independent walking
- 3) Symptomatic improvement
- 4) Functional independence while performing daily activities
- 5) Preventing complications, contractures and deformities
- 6) Preserving cardio-respiratory function

#### **The available medications for MD are:**

##### **Eteplirsen:**

Eteplirsen is a drug that was approved by Food and Drug administration

of United States America in late 2016. The drug is not a cure for the disease but slows the progression of the disease in about 13% of the DMD children afflicted with a specific mutation or deletion in dystrophin gene. It does not correct the mutation but to some extent can correct the process of protein formation. This method can prompt a faulty dystrophin gene with a specific deletion or mutation of exon 51 to produce partially functional dystrophin and therefore can slow down the progression. At the moment this drug is only available in USA.

### **Corticosteroids:**

These are considered to be gold standard for management of DMD. These medicines mimic the function of hormones secreted by adrenal cortex. The corticosteroids prescribed in muscular dystrophy are analogous with glucocorticoids. The use of steroids is not very popular in India, though short term usage has found its place in MD treatment regimens. In other countries like USA, Canada, Australia, etc. steroids have become an integral part of the treatment regime for MD.

Glucocorticoids prevent muscle protein breakdown, stimulate of formation of new muscle cells, stabilize muscle fiber membranes, increase muscle repair, reduce inflammation and other immune responses that expedite muscle damage. This helps to maintain muscle strength for a longer time therefore has a moderate effect on the disease progression. These medicines can lengthen the period of independent walking by 2 to 3 years, they can preserve cardio-respiratory function in the advanced stages of the disease.

However, higher dosage and long term usage of these medicines can cause various side effects like osteoporosis, increased weight, increased body hair, cushingoid symptoms, acne, blurred vision, cataracts or glaucoma, easy bruising, sleep disturbances, increased blood pressure, increased appetite. These side effects are noticed more with prednisolone as compared to deflazacort. These medicines should therefore be prescribed strictly under the supervision of a doctor and under supplementation of calcium and vitamin D along with dietary modifications. Regular bone mineral density measurement, routine blood pressure, weight and eye checkup should be carried out. Physiotherapy rehabilitation especially stretching of deformities and

weight bearing activities like standing with splints should be conducted with precaution. If the child is ambulatory, falls should be prevented as bones may be so porous that these can be fractured easily.

### **Vitamins**

Various vitamins like Coenzyme Q10 (CoQ10) having antioxidant property, Omega 3 fatty acids having an anti-inflammatory and Vitamin E having both anti-oxidant and anti-inflammatory effect are prescribed to improve muscle repair.

In addition to these cardioprotective drugs and respiratory drugs are given as and when symptoms arise but do not alter disease pathology. Muscular dystrophy is still a devastating disease with no definite cure. Existing drugs only slow down progression through addressing secondary effects of the disease pathology but none of these drugs can prevent degeneration or promote regeneration and therefore have temporary effect.

### **Surgical management of Duchenne Muscular Dystrophy**

Surgical management in DMD is to prevent development of contractures or deformities of limbs and spine to preserve maximum function. Along with surgery regular physiotherapy and orthotics prescription is essential. Once muscle contracture develops and walking becomes increasingly difficult, soft tissue surgery is suggested to maintain the limb alignment and joint position. Surgery improves the walking balance & prolongs the ability of the child to walk. Surgeries recommended for DMD, based on stage of the patient:

1. Before contracture begins : Release or opening up of muscles of the hip, thigh and ankle to prevent restriction of joint range of motion
2. When difficulty in walking starts due to tightness of muscles: Release of only ankle and knee muscles. This is called Moderate ambulatory approach.
3. Correction of toe walking (equinus gait) - Minimum Ambulatory approach

4. When the child ceases to walk: Aim of surgery is to re-establish walking. This is

### **Rehabilitative approach.**

5. The Palliative approach is the one in which the child is wheel-chair bound and surgery is only done for relief of pain, comfort, ease of nursing care & for shoe wear.

The surgical management of DMD varies based on symptoms of the patient and phase of the disease. It is important that timely opinion of an orthopedic surgeon is sought in presence of severe contractures and deformities.

### **Rehabilitative management of Duchenne Muscular Dystrophy**

#### **Physiotherapy**

Physiotherapy uses physical means and exercises to reduce pain and improve function in DMD. Physiotherapy systematically and scientifically facilitates and maintains body movements. In DMD physiotherapy plays an important role to prevent most debilitating complications like contractures, deformities, postural deviations and scoliosis of the spine. Physiotherapy is aimed at maximizing the physical performance and improving the quality of life of patients. Restorative functions of physiotherapy in muscular dystrophy are functional enhancement, correcting postural deviations and gait patterns and maintain the integrity of joints using various orthoses.

#### ***Aims of Physiotherapy***

- To assess muscle strength, joint mobility, functional abilities and susceptibility for deformities
- To determine the causes of deformities and prevent deformities
- To suggest supportive physical aids when required to prevent deformities and preserve function
- To educate the patients, parents/caregivers regarding the disease, its outcome and how physiotherapy can benefit



- 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 prescribe exercises such that muscle recruitment can be enhanced and muscle function can be preserved for a longer time

DMD are making use of the muscle plasticity, reducing the resistance to accommodate for the progressive loss of strength, facilitating less harmful muscle contractions, avoiding disuse of the muscles, preventing muscle fibrosis and contractures. Different exercises are prescribed with or without special aids for this purpose.

In the early stages of the disease mainstay of physiotherapy treatment in active exercises and functional activities that facilitate muscle strengthening and also improve muscle endurance. Higher repetitions and moderate to low resistance is the key in DMD. If the resistance is too much it can cause more damage to muscles. Hence there was a myth that exercise is harmful in DMD, but that is incorrect. Moderate exercise has several benefits in DMD.

Different types of exercises that are beneficial in muscular dystrophy are

### **1. Muscle strengthening exercises**

Exercises that make use of resistance bands, weights or individual's body weight to increase the strength of the muscles. However the resistance used in these exercises must be moderate to low.

Basic principles behind prescribing physiotherapy for children with

### **2. Muscle endurance exercises**

These are the exercises that consist of higher repetitions and maintaining a particular position for a longer time. These improve the day to day function.

### **3. Cardio-respiratory endurance exercises**

These 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.

### **4. Stretching exercises**

These are the exercises that are performed to improve the length of the muscles so that all the joints can be used through their complete range of motion. In the early phase of the disease a child can be asked to do these stretches on his own however in the later phase it can be performed passively by a care giver.

### **5. Range of motion (ROM) exercises**

These are the exercises that focus on simple day to day movements of all joints to the extent that is possible to keep the joints flexible. In the early disease stage a child can move his body on his own to achieve this however as weakness increases he could be assisted to perform these movements or the movements could be performed passively by the care giver.

### **Beneficial effects of physiotherapy in DMD**

- Increased bone growth and density of the bone minerals
- Maintained joint lubrication
- Maintenance and replacement of lubricating fluid in the joints
- Increased number and size of muscle fibers
- Improved blood supply to the muscles
- Reduced tiredness of muscles
- Cardioprotection
- Improved respiratory capacity and endurance
- Improved cognition
- Anti-inflammatory benefits

- 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
- Improved blood circulation

**Stretching exercises:**



*Fig. 4.1 Stretching of calf muscles*



*Fig. 4.2 Stretching of hip-flexors*

**Strengthening exercises:**





Fig 4.3 Trunk strengthening exercises



Fig 4.4 Lower extremity strengthening exercises



Fig. 4.5 Upper extremity strengthening exercises

**Balance training exercises:**



Fig 4.6 One leg standing



Fig 4.7 Reaching out exercises on the balance board

**Suspension exercises:**



Fig 4.8 Suspension exercises for lower extremity



Fig 4.9 Suspension exercises for upper extremity

## Gait training



Fig 4.10 Gait training in parallel bar and walker

## Respiratory exercises



Fig. 4.11 Inspiratory spirometer to improve respiratory endurance

## Orthotic and assistive devices used in Physiotherapy

When it becomes difficult for the children to walk some assistive devices can be given that help provide greater stability to the body or to assist or prevent any movement. These devices are called orthoses.

### Lower extremity orthoses

#### 1. Push knee splints

Push knee splints are used to support knees and prevents them from bending while standing and walking. Unfortunately the orthosis prevents knee bending completely and therefore the boys need to be retrained for walking with knee straight but use of this orthosis can delay loss of functional ambulation.



Fig. 4.12 Push knee splint

## 2. High Boots with steel shanks

These are used when the foot muscles are weak and the boys walk with feet slapping on the ground and cannot therefore walk over uneven surfaces or obstacles and are prone to frequent falls. These shoes provide stability to the ankle joint.



Fig. 4. 13High boots with steel shanks

## 3. Below knee braces with outer T- strap

These are special type of shoes that not only provide stability to the ankle joints but also stretch the tight calf muscles that turn the feet inside. Correction of this tightness facilitates correct weight bearing for the children.



Fig. 4.14 Below knee braces with outer T-straps

#### 4. Elbow splints

These splints prevent bending of elbows when weight bearing on upper extremity. These are mainly used while performing upper extremity and trunk exercises and rarely uses for any functional activity. Sometimes they may be used for walking.



Fig. 4.15: Elbow splints

#### 5. Close contact brace:

Close contact brace is a tailor made brace that is a snug fit for the trunk and it prevents unwanted excessive curvature of the spine. Usually this brace is used at the later stages of the disease to prevent the spine curvature while sitting and standing.



Fig. 4.16: Close contact brace

### Occupational Therapy

While physiotherapy deals with the physical impairment, occupational therapy mainly deals with functional aspect. The aim of Occupational therapy is to improve physical performance, make the boys functionally as independent as possible and increase their social participation. Occupational therapists have a unique ongoing role in supporting and working with patients with muscular dystrophy as the patient's needs



and the needs of their care givers are constantly changing. They need to assess and evaluate an individual's physical, psychological and social needs and focus on maximising skills, promoting and enabling independence, as well as improving the quality of life of the affected individual and his family to improve social participation and enhance quality of life.

Occupational therapists assess the physical capabilities of children with DMD like muscle strength, tightness, balance etc. In addition they also assess the independence in performing day to day functional activities and difficulties faced by them. They also assess the play and recreational activities and school performance. Occupational therapy assessment consists of not only the child and his capabilities but environmental and social factors that may limit their function and increase dependence in performing daily activities.

Occupational therapy is multidimensional and includes exercises prescription, cognitive training, prescription of assistive and adaptive devices, prescription of environmental modifications, caregiver counseling and patient counseling.

In the early stage of the disease caregiver counseling, prevention of possible muscle tightness and advice regarding possible environmental changes anticipating future functional dependence. Occupational therapists at this stage make use play behavior, hobbies and sports for the training of children with DMD

In the later stages the therapy consists of prescription of balance exercises, strengthening exercises and stretching exercises in order to limit the physical impairment and provide functional independence. Once maximum physical performance is achieved alternate techniques for performing activities daily living or use adaptive and assistive devices and environmental modifications are prescribed to increase functional independence.

In the advance stage of the disease when the child becomes wheelchair bound, need for the wheelchair and prescription of wheelchair to maximize independence in ambulation, functional activities, activities at school as well as social participation is also done by the occupational

therapists after assessing physical and environmental limitations.

### **Training of activities of daily living**

Basic activities of daily living are Eating, Bathing, Dressing, Toileting, Transferring, Maintaining continence of bowel and bladder.

In the early phase of the disease till about 7 years none of these are affected. Once the child starts experiencing difficulty to get up from the floor toileting activities may be difficult in case of indian toilets, in case of western toilets this independence can maintained for a few years more.

Beyond the age of 10 the child needs some assistance for all of the activities of daily living. Different assistive devices that can be used are as follows.

#### *Eating*



Two handled mug can be used when the muscle weakness of elbows increases and it is difficult to lift the mug or cup with one hand. This provides more strength and stability .

Fig. 4.17 two handled mug



Plate guard is used when dexterity of fingers reduces or wrist muscles weaken limiting the movement and food spillage increases.

Fig.4.18 Plate guard



Anti-slip mat can be used so that spillage is prevented and it is easy to pick up food with spoon as plate is fixed. This is used for boys with poor grip.

Fig. 4.19 Anti slip mat



Mobile arm support is useful for boys who are unable to perform shoulder movements and therefore cannot lift their arms to feed themselves.

Fig.4. 20 Mobile arm support

## Grooming

Grooming includes day to day tasks like brushing, combing hair, tasks for personal care and hygiene.



Fig. 4.21 long handle comb

## Bathing



Fig. 4.22 Soap in a mitten

## Dressing



Fig. 4.23 Velcro clothing



Fig. 4.24 Zip Puller

## Toileting



Fig. 4.25 Raised toilette seat

## Aquatic therapy

Aquatic therapy is one of the 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".

Aquatic therapy is provided by immersing the patients in water to various depths. Different types of exercises are performed in water either actively by the child or with assistance from the therapist or performed

passively by the therapist. The type of exercises performed in water could be categorized into stretching, strengthening and endurance exercises.

The Children who are unable to swim can also undergo aquatic therapy. Boys with advanced disease who have developed severe contractures or severe deformities of spine may not be eligible for this therapy. Also boys with smaller respiratory capacity (<250ml) also cannot undergo aquatic therapy, as it may be difficult for them to breath when immersed in water.

### **Beneficial effects of aquatic therapy in DMD**

The exercises are performed in environment with reduced gravitation effect and therefor the work of the muscles is reduced, harmful eccentric contractions of the muscles are prevented thereby preventing accelerated 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 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 advanced stages of the disease the freedom of movement and three dimensional access to the body can help in performing better stretches and therefore can prevent deformities. Water immersion also has a pain reducing effect and therefore the stretches are better tolerated by the patients.
- Aquatic therapy also improves respiratory capacity and cardiovascular endurance.
- 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.

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

Aquatic therapy is a useful tool to maintain a high functional level of children with DMD. It can be a good tool for rehabilitation especially in

the advanced stages of the disease where it is difficult to perform any movements on land.

Fig 4.26 Aquatic therapy pool in NGBSI

### **Speech therapy**

Boys with DMD may have some speech difficulties in the advanced stages of the disease due to hypertrophy or swollen appearance of the tongue. Speech therapy therefore plays an important role in identifying this impairment and then prescribing exercises to reduce speech disturbances. Tongue hypertrophy also affects their breathing at night and swallowing. Difficulties in swallowing can lead to aspiration i.e fluid or food particles entering lungs in small quantities and not eliciting any cough reflex. Such aspiration is called micro-aspiration and can lead to frequent chest infections. Impaired swallowing can also cause macro-aspiration compromising breathing. Speech therapy includes identification of impaired swallowing and teaching correct swallowing techniques and techniques to avoid aspiration.

Speech therapy exercises comprise of strengthening of facial muscles, strengthening of tongue muscles, improving respiration, improving respiratory control and slowing the pace of speech. For management of swallowing difficulties level of aspiration is considered. Devices like Ryle's tube that helps feed through nose and percutaneous endoscopic gastrostomy can be performed to feed directly through stomach.

### **Do's and Don'ts to prevent aspiration**

- Sit in an upright while eating and drinking; always feed the child and give water to the child in an upright posture.
- Take small bites
- Keep food moist
- Eat slowly
- Eat from the stronger side of the mouth
- Check for food accumulation in the mouth after eating

- Maintain high oral hygiene
- Drink the water by closing the lips on the glass or bottle, use straw if and when prescribed by your therapist
- Turn head down, tuck your chin towards the chest and lean forward to eat this helps to prevent the food from entering the wind pipe
- Eat in a relaxed comfortable environment
- Don't sleep immediately after eating
- Don't talk or laugh while eating
- Don't eat in a distracted environment
- Don't drink immediately after physical exercise, if you must drink take few small sips

## **Psychology**

DMD is a physically debilitating disease but it also has humongous psychological effect on the child as well as the parents, family members and other care givers. It is important to address these issues in the early stage to avoid psychological and behavioral disorders. How child feels, has an effect of his physical wellbeing and therefore psychological care is very important.

In a small percentage of boys with muscular dystrophy concomitant intellectual deficits may be noted. These children may also have subnormal Intelligence Quotient (IQ). These deficits can be identified as difficulty in expressing his needs or may have difficulty in remembering a lot of information, may have problems with maintaining attention or concentrating, however these issues may be overlooked by their parents due to their physical condition. It is therefore important to have regular psychological evaluation to address these deficits in time and take rehabilitative action for the same.

Apart from cognitive deficits which are part of the disease, boys with DMD mainly show lot of behavioral problems owing to high physical limitations. Children with muscular dystrophy show more impulsive



behavior and lack of emotional control. Some of these could be a side effect of the steroids that they take. They have poor capacity to adjust to surroundings. Their tolerance and co-operation levels reduce as the age progresses due to increasing limitation and muscle weakness. As the boys stop walking they may become more adamant, aggressive, uncooperative as the frustration of the disability sets in. Some of them may show exactly opposite signs where they become introverts, isolated and socially withdrawn. The signs of chronic sadness and depression must be identified in time to prevent its effects. As the disease progresses even further fear of death sets in and need for psychological support is the greatest.

Unfortunately, the disease does not cripple the boys alone but it also cripples the parents and care givers. They are limited in their social participation and sometime vocational activities as well. They are also emotionally burdened as the helplessness sets in with advancing disease. Parents themselves may require psychological support and counseling.

Below are some strategies to Improve attention, listening and memory problems:

- Sit close to the child in a calm room with less opportunity for distraction and explain the task to him in simple commands in a timely manner
- 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.

There are few areas of functioning in DMD in which psychosocial adjustment may be required:

1. Relationship with friends
2. Dependence on family members/caregivers
3. Helplessness in performing daily activities
4. Fear of being alone

How to deal with emotional problems?

- The first step to handling the emotional problems is to identify the emotion being felt like anger, sadness, frustration etc.
- Make sure to assure the boys that they are being heard and it is normal to feel these emotions
- It is very important to teach the boys to express his emotions in a constructive manner
- If the boy is having frequent outbursts, negative effects of the emotions, temper tantrums, chronic depression then a psychologist must be consulted.

**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

### **Nutrition in DMD**

Nutritional therapy can aim in improving the overall quality of the life of the patient. Nutritional care in the early stages of the condition can prevent or rather delay onset of osteoporosis and other deficiencies like Calcium and Vitamin D. The body replaces the muscle tissue with fat and connective tissue. Altering the fat intake is therefore essential.

Research has demonstrated inadequate nutrient intake of protein, energy, vitamins (water and fat soluble), and minerals (calcium and magnesium). Significant correlations were found between measures of strength and certain individual nutrients (e.g., copper and water-soluble vitamins).

A complete balanced diet keeping in mind the nutritional requirement and the current nutritional status of the patient is essential in DMD.

Nutritional care for different stages:

Stages of MD	Intervention
Presymptomatic and early ambulatory stage	Monitoring a normal weight gain forage Nutritional assessment for over/underweight
Late ambulatory	Restriction in energy in take due to decreased physical activity Monitoring high protein needs Monitor for nutritional efficiencies specially Vitamin D and calcium and others
Early non ambulatory	Assess the nutritional intake.  Treating constipation (infrequent or difficult bowel movements) by increasing the intake of fiber and fluid or usage of laxatives.

	<p>Monitoring GERD (gastro esophageal reflux), to reduce acidity and heart burn.</p> <p>Monitoring for nutritional changes in specific disease state like heart complications and hypertension</p> <p>Supplementation of nutrients required as they are prone to osteoporosis and fractures, due to steroid intake</p>
<p>Late non ambulatory</p>	<p>Problems of dysphagia to be monitored due to poor oral intake, and the need to consider tube feeding.</p>

**Why is it important to maintain adequate weight?**

- To reduce the burden on weakened muscles
- To maintain mobility and flexibility of the body
- To maintain range of motion of the joints of the body
- To reduce the strain on respiratory muscles and delay onset of heart complications
- To reduce the risk of scoliosis
- To reduce difficulty of transfers, especially for care takers if individual is immobile
- To reduce fatigue
- To minimize the effect on patient's ability to walk

**Simple to follow instructions in case of muscular dystrophy**

- Small frequent meals must be consumed every 2 hourly.

- A balanced meal includes all the nutrients required for the day.
- Breakfast is the most important meal of the day.
- Avoid too much carbohydrate rich foods in the second half of the day. All cereals are rich in carbs - rice, wheat, poha (rice flakes), rawa (semolina), maida (refined wheat flour), breads etc.
- Plenty of protein rich foods to be consumed all throughout the day. All pulses, sprouts, soya bean, paneer (cottage cheese), nonvegetarian foods such as eggs, fish, chicken, lean meats, milk and milk products are good sources of proteins.
- Additional supplements for omega 3 fattyacids can help a great deal after stem cell therapy. Some food sources of omega 3 fattyacids are almonds(badam), walnuts (akrod), olive oil, flaxseeds, eggs. These foods must be consumed on regular basis.
- Including foods rich in zinc and selenium also proves useful as patients are seen having a deficiency of the same.
- Reduction in the total amount of fat ie: oil, butter, ghee, cheese, coconut, groundnuts, selected dryfruits like cashewnuts, kismish is required.
- Simple sugars and sweets can add to a lot of calories in the daily diet hence must be avoided as far as possible.
- Fresh vegetables and fruits not only add fiber to the diet but also lower in calories compared to carbohydrate rich foods or foods high in fat.
- Whole cereals like jowar, bajra, ragi, oats must be encouraged as they have a higher fiber content.
- Any fried food if desired must be eaten for breakfast in small amounts as it can be digested throughout the day.
- Consumption of egg whites increases the protein intake as well as keeping the calories low.
- Chicken and fish also can be included in the daily diet as long as the preparation does not involve too much of fat content.

- Cooking methods such as boiling, grilling, steaming can help reduce oil and fat content in the foods.
- Whole pulses and sprouts must be consumed on daily basis to increase the protein, fiber and micronutrients especially for vegetarian patients.
- Green tea is found to be very rich in antioxidants that prevent the degeneration of the muscles hence must be consumed daily. It also aids in digestion and helps in weight loss. At least 2 to 3 cups of green tea must be consumed per day.
- Make eating a pleasurable one for the patient.
- Adding lot of colourful vegetables and fruits to the plate will make eating healthy foods an enjoyable one.
- Choice of foods that the parents make is very essential as the child looks upon them.

### **Additional matter for DMD chapter**

Despite the multidisciplinary rehabilitation, the disease progresses over time and the children with DMD lose their ability to walk by the age of 8 to 11 years, once they become wheel chair bound they also lose the ability to use their arms and eventually succumb to death because of the inability to breathe. There is no medicine or rehabilitation technique that can cure this disease, children continue to grow weaker and weaker each day. This is mainly because none of the treatment available today can correct the basic genetic flaw or replenish the exhausted pool of stem cells. Therefore, newer treatment strategies must focus on either one of these aspects or both these aspects. Stem cell therapy has a potential to replace and regenerate damaged tissue and therefore can be a promising treatment.

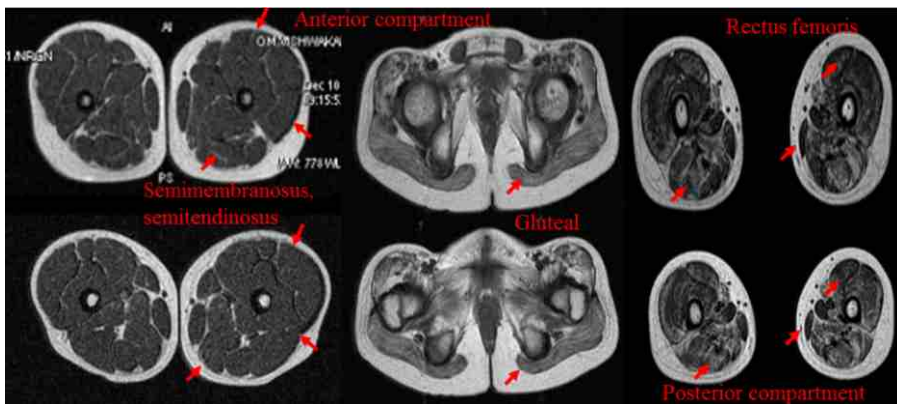
### **Stem cell therapy**

Duchenne Muscular Dystrophy is characterized by progressive loss of muscle cells. The symptoms are visible only when the stem cells that repair the muscles are exhausted. Therefore, it is important to replace these stem cells. Stem cell transplantation in different muscles replenishes this stem cell pool. These cells also have a protective effect on existing muscle cells. Therefore, stem cells can prolong the progression of disease and benefit in DMD.

Stem cell therapy addresses the root problem in DMD. Stem cells initiate the process of repair and regeneration by converting into muscle cells and stimulating local stem cells. They release paracrine molecules such as growth factors, cytokines, and chemokines that reduce inflammation, promote proliferation of existing stem cells, they also help to alter the immune responses of the body and reduce scarring and fatty infiltration of the muscles. They promote formation of new blood vessels increasing the blood and oxygen supply to the muscles. Exercise has these similar effects on the muscles but in absence of stem cells the exercise has very limited potential to form new muscle cells, therefore combination of exercise and stem cells give exponentially greater benefit.

### Improvements noted post stem cell therapy

- Improved muscle strength
- Improved balance
- Reduced frequency of falls
- Improved trunk balance
- Reduced deformities
- Reduced difficulty of walking
- Improved standing
- Improved breathing
- Improved cardiovascular endurance



*MRI-MSK after Stem cell therapy demonstrating slowing or halting of the progression of pathology in Gluteal, and Mid thigh muscles*

• **List of publications by NeuroGen BSI in DMD**

1. Alok Sharma, Hemangi Sane, Amruta Paranjape, Khushboo Bhagwanani, Nandini Gokulchandran, Prerna Badhe. Autologous bone marrow mononuclear cell transplantation in Duchenne muscular dystrophy – a case report. American journal of case reports 2014;15: 128-134.
2. Dr. A. Sharma, Ms. P. Kulkarni, Dr. G. Chopra, Dr. N. Gokulchandran, Dr. M. Lohia, Dr. P. Badhe. Autologous Bone Marrow Derived Mononuclear Cell Transplantation In Duchenne Muscular Dystrophy-A Case Report. Indian journal of Clinical Practice 2012; 23 (3): 169-72.
3. Alok Sharma , Dr. Prerna Badhe, Hemangi Sane, Suhasini Pai , Pooja Kulkarni, Khushboo Bhagwanani, Dr. Nandini Gokulchandran. Halting of functional decline in a case of Duchenne Muscular Dystrophy after cellular therapy. International Journal of Recent Advances in Multidisciplinary Research (IJRAMR), 2017 Jan
4. Alok Sharma, Hemangi Sane, Vaibhav Lakhanpal, Amruta Paranjape, Pooja Kulkarni, Nandini Gokulchandran, Prerna Badhe. Stabilization of the disease progression in a case of Duchenne Muscular Dystrophy with cellular transplantation. Stem cell: Advanced research and therapy. 2017; 2017(3)
5. Alok Sharma, Amruta Paranjape, Ritu Varghese, Hemangi Sane, Nandini Gokulchandran, Jasbinder Kaur, Prerna Badhe. Functional improvements and Musculoskeletal Magnetic Resonance Imaging with Spectroscopy changes following cell therapy in a case of Limb Girdle Muscular Dystrophy. International Journal of Cell Science and Molecular Biology.
6. Alok Sharma, Nandini Gokulchandran, Amruta Paranjape, Hemangi Sane, Dr. Prerna Badh. Stem cells as a therapeutic modality in Muscular Dystrophy. Chapter 2. Muscular Dystrophy. Avid Sciences. India. 2017
7. Alok Sharma, Hemangi Sane, Amruta Paranjape, Khushboo Bhagwanani, Nandini Gokulchandran, Prerna Badhe. Autologous



bone marrow mononuclear cell transplantation in Duchenne muscular dystrophy – a case report. American journal of case reports 2014;15: 128-134.

8. Dr. A. Sharma, Ms. P. Kulkarni, Dr. G. Chopra, Dr. N. Gokulchandran, Dr. M. Lohia, Dr. P. Badhe. Autologous Bone Marrow Derived Mononuclear Cell Transplantation In Duchenne Muscular Dystrophy-A Case Report. Indian journal of Clinical Practice 2012; 23 (3): 169-72.
9. Alok Sharma , Dr. Prerna Badhe, Hemangi Sane, Suhasini Pai , Pooja Kulkarni, Khushboo Bhagwanani, Dr. Nandini Gokulchandran. Halting of functional decline in a case of Duchenne Muscular Dystrophy after cellular therapy. International Journal of Recent Advances in Multidisciplinary Research (IJRAMR), 2017 Jan
10. Alok Sharma, Hemangi Sane, Vaibhav Lakhanpal, Amruta Paranjape, Pooja Kulkarni, Nandini Gokulchandran, Prerna Badhe. Stabilization of the disease progression in a case of Duchenne Muscular Dystrophy with cellular transplantation. Stem cell: Advanced research and therapy. 2017;2017(3)

## Chapter 8

# Ataxia

Ataxia in children is a sign of different disorders involving the nervous system and is associated with an impairment of coordination of movement and balance. It is an impairment of direction, rate and strength of voluntary movements resulting in an inability to perform precise movements and a loss of balance.

**Ataxia is a symptom of underlying disease. Ataxia can be classified into:**

- Sensory ataxia: This results from loss of sensory input arising from diseases in nerves or posterior column of spinal cord.
- Cerebellar ataxia: Resulting from disease or injury within the cerebellum (a part of the brain which controls coordination and balance) or its neuronal connections.

**Causes of ataxia include:**

- Excessive drug ingestion, drug intoxications. Antiepileptic drugs may cause ataxia
- Head trauma
- Cerebral palsy
- Tumor: A growth in the brain can damage the cerebellum

- Infections: It can be a complication of chicken pox and other viral infections
- Low intake of vitamins thiamine, cobalamin, vitamin E, zinc and folate in neglected children or in children affected by intestinal disorders may result in symptoms of ataxia
- Genetic

### **Symptoms:**

- Lack of coordination
- Unsteady walk
- Difficulty with fine motor tasks such as eating, writing or buttoning
- tremors
- Change in speech
- Difficulty swallowing
- Involuntary eye movements

### **Diagnosis:**

Recognizing ataxia in children may be challenging. It may be overlooked in very young children. Diagnosis involves:

- Neurological examination including a detailed history, checking balance, coordination and reflexes
- Computed Tomography (CT) Scan or magnetic resonance imaging (MRI) to determine the potential cause such as a tumor.
- Lumbar puncture for cerebrospinal fluid testing
- Genetic testing: To determine hereditary ataxic conditions

### **Treatment:**

There is no treatment specifically for ataxia. In some cases, treating the underlying cause resolves the ataxia. Ataxia caused by conditions such as cerebral palsy and genetic ataxia may not be treatable. In such cases treatment will include:

- Physical therapy to improve coordination and enhance mobility
- Occupational therapy to provide help in activities of daily living and adaptive devices.
- Speech therapy to improve speech.

### **Physiotherapy Treatment**

- Therapeutic goals include improving balance and posture against outside stimuli, increasing joint stabilization, developing independent, functional gait to promote independence.
- Therapy is used to improve balance and increase the independence of the patient using techniques focusing on balance, posture and increasing coordination.
- Training principles include progressing from simple to complex exercises, practicing exercises with eyes open and closed and providing support with home exercise and sports activities.
- Lack of balance is one of the main symptoms of cerebellar ataxia and physical therapy is used to help improve balance. Stabilizing the trunk and proximal muscles should begin with mat activities such as moving onto the forearms from lying face down; crawling; and moving onto the knees and into a sitting position.
- Balance training over any moving surface (ball/bolster) to improve core muscle strength and balance
- Children with ataxia have poor dissociations and stabilize with upward visual gaze fixation, therapy should focus on developing rotations( frontal plane weight shifts) and reducing the upward gaze.
- Gait training should also be done, as it is an excellent indicator of stabilization and balance. Use of SPIO, to stabilize the trunk.

### **Occupational Therapy for children with Cerebellar Ataxia**

#### ***Goals for children with Ataxia :***

- Activities to improve Gross motor Co ordination
- Activities for improving fine motor co ordination

- Activities to improve Handwriting
- Improving ADLs
- Energy conservation techniques
- Work Simplification techniques
- Home and Environmental modifications
- Use of adaptive devices and assistive technology for improving functional independence

Occupational Therapy Treatment includes following strategies:

***Activities to improve proximal stability:***

- Crawling
- Pushing and pulling activities
- Tug of war
- Hand push activity, chair push activity in the classroom

***Improving Self-care:***

- Using long handle comb
- Using trimmer with broad base to hold instead of razor
- Use of adapted nail cutters
- Use of Electric toothbrush instead of normal brush

***How to improve Eating:***

- Use of weighted spoon, utensils
- Use a bigger plate and spoon with broad and deep base.
- Use of plate guard
- Use of non-skid mat below the plate
- Use of sipper instead of glass

***Improving Bathing:***

- Using the shower chair to sit with proper arm and back support
- Placing grab bars on the wall to hold

- Use of lever taps instead of revolving taps
- Soap with ropes
- Use of hair/body dryer instead of drying body with towel

***Improving Dressing:***

- Using loose fitting clothes
- Use of easy to wear shirt and pants (T shirts and pant with elastic)
- Using dressing stick
- Using larger buttons instead of standard small buttons
- Zips with loops to hold

***Toileting:***

- Height of the toilet seat should match Grab bars
- Using jet spray for cleaning
- Keeping tissue roll at a reachable distance
- Use of power flushing system
- Grab bars to get up easily from toilet seat

***Improving Ambulation:***

- Use of walking aid
- Use of weighted cuffs while walking. (attaching them to the distal end of the leg)
- Using nonskid / anti-skid mats, carpets
- Asking the child to wear shoes with good grip
- Preventing fall by installing grab bars

***Improving Handwriting:***

- Modified grippers
- Weighted pens
- Use of ink / gel pen instead of ball pen (which will reduce the friction)
- Using ruled papers

- Practicing signatures, long strokes on the board.

### ***Home and Environmental Modifications:***

- There should be sufficient light in the room
- Remove door thresholds
- Door width should be increased
- Place hand rails near the staircase
- Use of bright colored tape / tiles near the stair case
- Using chairs with arm rests
- Smoothing the edges of the home furniture
- Replace the soft cushion of the sofa by firm cushion.
- Fix an emergency bell near the bed (in case of emergency)

### **Speech Therapy**

Speech and language Pathologist (SLP) can help people with cerebellar ataxia in the management of communication and swallowing issues.

The most evident communication difficulty experienced in an individual with cerebellar ataxia is that of dysarthria, a motor speech disorder resulting in altered voice quality, speech clarity and intelligibility. Communication may also be affected in case of cognitive impairment impacting on language processing. In addition, any difficulties with executive functions also may result in communication behaviour.

Speech therapy is one of the most recommended treatment options for individual with cerebellar ataxia.

Speech therapy delves much deeper than just basic language training. It includes exercises and techniques.

Some of the most commonly recommended speech therapy techniques include:

- Augmentative and Alternative Communication (AAC) Devices: An AAC device is anything that is an alternative to oral communication. AAC methods are divided into two categories:

1. Unaided: Relies on body language – gestures, sign language, etc.
  2. Aided: Uses tools to help improve communication, such as paper and pencil, computers, speech generating devices, pictures or symbols, or communication boards/books.
- Articulation Therapy: Uses language cards to identify and focus on specific sounds. Often includes having children make sounds in the mirror so they can see how their mouths move.
  - Blowing exercises to improve mouth muscles.
  - Breathing exercises to strengthen the diaphragm.
  - Jaw exercises to strengthen muscles.
  - Lip exercises to strengthen muscles and improve lip extension.

Swallowing is a common symptom of ataxia, particularly as the disease advances. Depending on the path physiology of the disease, swallow dysfunction (dysphasia) may occur at the oral, pharyngeal and/or esophageal stage of swallowing. For example, when there is cerebellar involvement, dysphasia may be characterised by reduced coordination of the oro-pharyngeal muscles involved in swallowing food and drink.

### *Dysphasia Management Techniques.*

- Modification of consistency of food or drink.
- Introduction of safe swallow strategies including use of a chin tuck position, double swallow, throat clear.
- Advice regarding sitting posture and set up for oral intake.
- Introduction of care-initiated prompts to maintain safety e.g. slow rate, small sips, avoiding talking with food or drink in mouth.
- Advice on oral hygiene care.

### **Psychological Intervention**

Children with Ataxia often face difficulties in carrying out their day to day activities which generates feeling of frustration and depression. Talking to a counselor can help them cope up with such feelings. Patients



with Ataxia also suffer from depression and anxiety issues, in such cases, Neuropsychologists work with Psychiatrists who prescribe them anti-Psychotic medications. A Neuropsychologist can evaluate and treat their degree of Impairment using some of the scales like Beck Depression Inventory, Anxiety Inventory and Hopelessness scale, Cognitive assessment screening instrument (CASI), Dean-Woodcock Neuropsychological assessment system (DWNAS) etc. These tests have specifically designed tasks which offers an estimate of a peak level of a person's cognitive performance. Neuropsychological tests are typically administered in an environment which is quiet and free of distractions. Neuropsychologists help family develop a treatment plan by understanding how the brain functions and how that functioning relates to behavior. Treatment plans may include a combination of medications and rehabilitation therapy.

### **Nutrition**

- Inclusion of complex carbohydrate like legumes, starchy vegetables, fruits in diet help this patients feel fuller and have more energy.
- High protein low fat meat, dairy products help in meeting protein requirements of these patients.
- Inclusion of fresh fruits and vegetables is important to keep bowel movement regular.
- Inclusion of plenty of water is important.
- Foods with simple carbohydrate should be limited.
- Food with added preservatives should also be avoided.
- Certain foods which can cause dizziness and lack of balance like aspartame, monosodium glutamate should be avoided.
- Supplementation of multivitamin with zinc and omega 3 is helpful.

### **Stem cell therapy**

All the above-mentioned treatment options focus on symptomatic management. None of the treatment approaches address the underlying pathology of the disorder. A therapeutic strategy is required to stop the

degeneration, repair the damaged areas and protect the unaffected areas.

Stem Cell Therapy has opened new avenues for the treatment of cerebellar ataxia. Studies have shown that stem cells migrate to the site of injury from the site of injection. They enhance angiogenesis and contribute to neovascularisation by producing signalling molecules such as vascular endothelial growth factor (VEGF) and fibroblast growth factor (FGF2). They impart immunomodulatory and anti-inflammatory effects. The various mechanisms along with paracrine effects of cellular transplantation may help reverse the disease pathology.

## Chapter 9

# Brain Stroke

Stroke is brain damage that occurs secondary to the blockage or breakage of blood vessels in the brain. The causes of stroke in children are many and often not well understood. Stroke can happen to anyone at any time, including teenagers, children, newborns, and unborn babies. The risk of stroke in children is greatest in the first year of life and during the period of right before birth to right after birth. Stroke remains among the top 10 causes of death in children.

Stroke happens in about 1 in 4,000 live births. The risk of stroke from birth through age 18 is almost 11 in 100,000 children per year. Strokes are slightly more common in children under age 2.

Common risk factors for stroke in children include:

- Congenital heart defects
- Sickle-cell disease
- Immune disorders
- Diseases of the arteries
- Abnormal blood clotting
- Head or neck trauma
- Maternal history of infertility

- Maternal infection in the fluid surrounding an unborn baby
- Premature rupture of membrane during pregnancy
- Pregnancy related high blood pressure in the mother

The common stroke warning signs used to identify stroke in adults can also be used to identify strokes in children. But, there are some specific symptoms that should be looked for in children depending on their age.

### **In newborns and infants**

- Seizures.
- Extreme sleepiness.
- Tendency to use only one side of their body.

### **In children and teens**

- Severe headaches
- Vomiting
- Sleepiness
- Dizziness
- Loss of balance or coordination

However, the biggest challenge with newborns and infants is that they cannot communicate symptoms that are not readily visible.

Diagnosis requires careful clinical examination combined with brain imaging.

### **Treatment**

Children usually recover better from strokes than adults do because their brains are still growing. Early treatment focuses on protecting the brain and keeping blood vessels open to prevent more strokes. However, most children experience neurologic deficits with long-term treatments focused on physical, developmental, and psychosocial complications.

Treatment for pediatric stroke depends on the specific cause. Some common treatments used with adults such as tPA, or tissue plasminogen

activator, a drug that dissolves blood clots are not appropriate for young children and infants.

Current treatments for pediatric stroke include:

- Supportive care to maintain normal body temperature, proper hydration, and normal blood sugar levels.
- Controlling high blood pressure.
- Detecting and treating seizures with EEG monitoring and anticonvulsant medication.
- Managing intracranial pressure.
- Blood transfusions for children with sickle-cell disease.
- Antithrombotic therapy, which refers to medications used to prevent blood clots from forming or growing, is used in children but generally not infants.
- Surgery related to hemorrhagic stroke (and less commonly, ischemic stroke) is sometimes performed to relieve pressure on the brain.
- Rehabilitation

### **Physiotherapy**

Pediatric patients with brain injury due to a cerebrovascular lesion (stroke) to consider for rehabilitation are based on age and presentation:

- Neonates/Perinatal: 22 weeks pregnant- 1 month old
- Children: 1 month to 18 years

Rehabilitation is key to help with long term outcome. As the child grows their needs for therapy and services may change. Children who survive a stroke, about 60% will have permanent neurologic deficits. These deficits may include:

- Hemiplegia
- Hemiparesis
- Learning and memory problems

- Difficulty with speech and language
- Visual problems
- Behavior or personality changes
- Epilepsy
- Swallowing and eating problems

Infants and toddlers (Ages 1 month to 3 years):

- Make daily stretches/ exercises a part of child's everyday routine starting at a young age.
- Focus should be given on gross motor skills: sitting, crawling, walking, jumping, and running
- Neurodevelopment Therapy (NDT)
- Strategies to normalize the muscle tone.
- Increase movement on the affected side through playful and recreational activities.
- Special splints and braces.

Early childhood (Ages 4 years to 12 years):

- Neurodevelopment Therapy (NDT)
- Strategies to normalize the muscle tone
- Strengthening exercises
- Stretching exercises
- Bed mobility exercises
- Increase movement on the affected side through playful and recreational activities
- Balance training
- Gait training
- Special splints and braces

- Constraint Induced Movement Therapy (CIMT)
  - During this therapy, the unaffected arm/hand is restrained, so that the child is encouraged to use the affected arm/hand
- Spider web therapy
- Animal therapy (Hippotherapy)

Teenagers (Ages 13 to 18 years):

- Strengthening exercises
- Bed mobility exercises like rolling, supine to sit, shifting in bed forward and backward, shifting sideways, quadruped, kneeling and half kneeling positions
- Improve voluntary control through Neuro Development Techniques (NDT), Motor Relearning Program, Proprioceptive Neuromuscular Facilitation techniques (PNF), Constraint Induced Movement Therapy (CIMT)
- Functional Reeducation.
- Balance and postural control training
- Gait training
- Special splints and braces
- Mirror therapy
- Mental Imagery
- Animal Therapy
- Recent advances like virtual reality, robot-assisted therapy.

### **Occupational Therapy**

Multidisciplinary approach is required for the rehabilitation of the stroke patient. Occupational therapist is one of the key members of the multidisciplinary team. Role of occupational therapist is to do the accurate assessment of affected motor functions, cognitive functions, visual perceptual skills, coordination which collectively affect the daily

functions of the individual suffering from stroke. Occupational therapy intervention focuses on individualized goal setting, skillful adaptation in the task and doing environmental modification for the patient to make him independent in their daily routine.

***Affected Motor Functions:***

Because of physical limitations like spasticity, contractures, muscle tightness, deformity, muscle weakness, functional activities get compromised. occupational therapy intervention includes use of Inhibitory techniques, PNF techniques for normalization of tone and transition towards improving the functional independence. Constraint induced therapy involves limiting the movement of the non-affected or stronger arm and instead using the affected or weaker arm more frequently and intensely for doing motor and functional activities.

***Affected Cognitive Functions:***

Cognitive skills like memory, judgment, planning and organization are required for performing daily living activities, work and leisure activities. Fronto- Parietal lobe affectation leads to affected cognitive skills. Accommodative, restorative and compensatory approach by using different therapeutic activities, functional tasks are used for regaining the cognitive functions

Cognitive training works on the fact of Neuroplasticity. Repetition of task and task specific practice to is effective. Patient should be engaged into meaningful activities and Functional training has to be task specific.

***Affected Visual perceptual skills:***

After stroke, occupational therapists commonly do screening of visual-perceptual impairments which affects ADLs such as agnosia; visuospatial relations problems, eg, figure-ground, body scheme disorders, depth perception, and unilateral neglects, and impairments in constructional skills.

Treatment intervention commonly includes activities like Identifying objects from the cluttered environment, Finding a specific image from the group of images, Improving directional concepts through games like dot joining, building blocks. Completing the pictures which are partially



drawn, Tracing the design, Puzzle games for improving sequencing abilities.

### ***Splints:***

Long standing Spasticity leads to contractures. Prolonged progressive stretch helps to reduce spasticity and helps to reduce the degree of contractures. Use of night splint is advised for providing prolonged stretch. Common examples of splint are use of corrective splint for foot for correcting calf muscle contracture post stroke. Using a palm based splint with thumb in abduction for correcting finger contracture.

### ***ADL modifications :***

ADL modifications and Adaptive devices help to improve functional independence. Use of modified spoon, glass holder helps the patient to do eating activity more independently. Devices like long handle shower, jet spray, modified toilet seat makes bathing and toileting easier.

### ***Home and Environmental modifications :***

Occupational therapist plays a vital role in doing home assessment. Occupational therapist. Occupational therapist has to make sure that the patient is successfully able to perform his activities by removing the physical barriers in the home environment. Eg. Making sure that patient is easily able to perform transfer activity from bed to wheelchair by doing the home assessment.

Ultimately transition of the patient occurs from Therapeutic activities to functional tasks and finally resuming back to their work leading to complete functional independence.

### **Speech Therapy**

Common speech disabilities found in patients of pediatric stroke are aphasia, dysarthria and apraxia.

**Aphasia** is a language impairment that affects the production or comprehension of speech and the ability to read or write.

There are two general categories of therapies provided:

- Impairment-based therapies are aimed at improving language

functions and consist of procedures in which the clinician directly stimulates specific listening, speaking, reading and writing skills.

- Communication-based (consequence-based) therapies are intended to enhance communication by any means and encourage support from caregivers. These therapies often consist of more natural interactions involving real life challenges in communication.

**Dysarthria** occurs when areas of the brain controlling our muscles for speech are damaged. A speech pathologist can help with strengthening muscles, increasing movement of mouth and tongue and breathing exercises. Common techniques are focusing on slow clear speech with frequent pauses, and starting a topic with a single word first and then checking whether the other person has understood.

**Apraxia** occurs when the speech muscles are unaffected but the brain has trouble sending the signals to the muscles responsible for speech. In mild cases, therapy involves saying individual sounds and thinking about how the lips and tongue should be placed, or speaking while clapping to improve timing. In severe cases, alternative systems such as gestures, facial expressions, written communication or pre-printed cards are used.

### **Psychological Intervention**

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 individual'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 betterment of 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 better 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 setting 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.

### **Nutrition**

- It is very important to keep watch on nutrition intake of stroke patient as they have difficulty in eating by them self and some may have swallowing chewing issue.
- For the patients with chewing difficulty mechanically soft food can be included.
- Food should be high in fiber with lot of different colored fruits and vegetables. Fiber in food help lower cholesterol.
- Antioxidants in fruits and vegetable also help also help lower blood vessel damage. potassium in fruits and vegetables help maintain blood pressure.
- Low fat protein sources should be included like low fat dairy products, lean chicken, fish, eggs, legumes, pulses.
- Processed foods should be avoided.
- Maintaining low salt intake will help control blood pressure. Also, it is important to read labels to check low salt foods.

### **Stem Cell Therapy**

With the current treatment approaches, medical, surgical or rehabilitative, the pathophysiological processes and the resultant damage occurring at the microcellular level in stroke cannot be reversed. This permanent change in the structure of Central nervous system leads to long lasting physical impairments, seen as residual problems, which translate gradually into activity limitation and restricts these children to participate in the community.

Stem cells mobilize to the injured areas of the brain and initiates the process of neurorestoration. These stem cells secrete various growth factors like VEGF, bFGF and BDNF which support and amplify angiogenesis, neurogenesis and synaptic plasticity at the penumbral region. Along with the above neuroreparative processes, the stem cells

also decrease the glial scar formation and promote glial-axonal remodeling which is seen in chronic stroke. Paracrine molecules carry out neuroprotection by halting further damage of the surrounding cells, reduce inflammation and regulate the immune cells. They also help in improving the blood and oxygen supply to the brain by forming new blood vessels (angiogenesis).

## Chapter 10

# Head Injury

Head injuries occur commonly in childhood and adolescence. Most head injuries are mild and not associated with brain injury or long-term complications. Very rarely, children with more significant injuries may develop serious complications.

Falls are the most common cause of minor head injury in children and adolescents, followed by motor vehicle crashes, pedestrian and bicycle accidents, sports-related trauma, and child abuse. The risk of brain injury varies with the severity of the trauma.

A child's behavior and symptoms after a head injury depend upon the type and extent of the injury. The most common signs and symptoms include:

- Scalp injury
- Skull fracture (eg, basilar skull fracture)
- Scalp swelling
- Loss of consciousness
- Headache
- Vomiting
- Seizures

- Concussion
- Contusion
- Intracranial and/or subarachnoid hemorrhage
- Epidural and/or subdural hematoma
- Intraventricular hemorrhage
- Penetrating injuries
- Diffuse axonal injury

While the symptoms of a brain injury in children are similar to the symptoms experienced by adults, the functional impact can be very different. The cognitive impairments of children may not be immediately obvious after the injury but may become apparent as the child gets older and faces increased cognitive and social expectations for new learning and more complex, socially appropriate behavior. These delayed effects can create lifetime challenges for living and learning for children, their families, schools and communities. Some children may also have lifelong physical challenges.

### **Treatment**

The goal of medical care of pediatric patients with head trauma is to recognize and treat life-threatening conditions and to eliminate or minimize the role of secondary injury.

Treatment of children with severe head injury includes management of the following:

- Airway
- Cardiovascular and circulatory status
- Intracranial pressure and cerebral perfusion
- Bleeding
- Seizure(s)
- Temperature

- Analgesia, sedation, and neuromuscular blockade

Surgical intervention in pediatric patients with head trauma may also be required and includes the following:

Pharmacologic therapy in patients with head trauma is directed at controlling intracranial pressure through the administration of sedatives and neuromuscular blockers, diuretics, and anticonvulsants.

### **Physiotherapy**

Physiotherapy helps in retaining muscle strength and flexibility, enhance coordination, reduce spasticity, regain greater control over bladder and bowel function, and increase joint movement. It can also help to reduce the likelihood of pressure sores developing in immobilized areas. Individuals are also taught to use assistive devices such as wheelchairs, canes, or braces as effectively as possible.

### **Occupational Therapy**

Children with traumatic brain injury can primarily have three types of problems

*Physical problems:* Spasticity, Rigidity, Hypotonicity, contractures, Involuntary movements, visual impairment, balance issues,

*Cognitive perceptual problems:* Forgetfulness, affected short term and long term memory poor attention span, affected cognition, higher cognition, difficulties in reading, writing and calculation

*Emotional Regulation Problems:* Mood swings, anxiety, depression, restlessness, agitation.

### **Occupational therapy Goals:**

**Improving Gross motor functioning:** Teaching the child to do activities using large joints e.g. Reaching out for objects, overhead activities

**Improving fine motor functioning:** Developing skills which are required to hold and manipulate small objects E.g. Writing, picking up coin, picking up beads, putting the clip.

**Improving performance in Basic ADLs:** Improving child's participation in activities like eating, self-care dressing, grooming

**Improving performance in Instrumental ADL:** Improving child's ability



to function in different environments like home, public places, school.

**Personal and social development:** Improving child's routine, habits and role identification.

**Occupational therapy Intervention:**

1. Occupational therapist work with multi-disciplinary team to help the child to excel in his areas of occupation.
2. During therapy, occupational therapists help in developing skills and to translate those skills into daily routine activities.
3. The abilities and their difficulties are measured and depending on that specific adaptive techniques are taught. These adaptive techniques can be specific to a environment or can be applicable in all environments.
4. When due to some physical or cognitive difficulties children find it difficult to apply adaptive techniques, simple low skill adaptive devices are provided and training in same are also given.
5. Children are also prepared to develop specific skills required at school such as pre-writing skills, reading skills, interaction, peer relation etc.
6. They are also evaluated for their ability to attend normal school and if required are referred to special or integrated schools. The occupational therapist is also responsible to communicate the school authorities about the child's abilities and difficulties. They may also help school authorities to develop a program that will benefit the child to excel in academics.

**Speech-language therapy**

Speech and language intervention in Children with TBI often depends on the state of arousal of the child after TBI and it also depends on the severity as well as number of years since the TBI has occurred. Speech and language problems persist even years after the TBI has occurred.

Paediatric traumatic brain injury (TBI) may result in long-lasting language impairments along with dysarthria.

TBI in childhood has been found to cause chronic linguistic impairments as a result of cognitive impairments thus the children can be diagnosed as

having Cognitive-Linguistic impairments.

*Speech and Language characteristics of children with TBI:*

- Childhood acquired language disorders may be characterised by deficits in any or all linguistic domains of **vocabulary i.e. pragmatics, syntax, morphology or semantics** (Feldman & Messick, 2008).
- Communication deficits are often characterized by:
  - ✓ difficulty in understanding or producing speech correctly (aphasia),
  - ✓ slurred speech consequent to muscle weakness (dysarthria)
  - ✓ difficulty in programming oral muscles for speech production (apraxia).
- They have difficulty to understand both written and spoken messages.
- They may also have impairments in spelling, writing, and reading.
- They may also have difficulty in social communication, such as difficulty in turn taking during conversation and problems in topic maintenance and initiation.
- They are often unaware of the inappropriate behaviours.
- Higher-order' language is particularly impaired in paediatric TBI, including discourse skills (Lê, Coelho, Mozeiko, Krueger, & Grafman, 2012; Marini et al., 2011) and understanding of irony or non-literal concepts (Angeleri et al., 2008; Dennis, Purvis, Barnes, Wilkinson, & Winner, 2001).
- Dysarthria, a motor-speech disorder, is also a common occurrence after TBI and may affect the intelligibility of the speaker (Morgan, Mageandran, & Mei, 2010). It can result from a combination of respiratory, phonatory, articulatory, and/or resonatory impairments (Cahill, Murdoch, & Theodoros, 2002).
- Many children also experience memory deficits which most of the time includes both short term as well as long term memory.
- Swallowing deficits (dysphagia) may also result due to weakness

and/or in coordination of muscles in the mouth and throat.

***Intervention:***

A treatment plan is developed after the complete evaluation of the child. The treatment program will vary depending on the stage of recovery, but it will always focus on increasing independence in everyday life.

In the early stages of recovery (e.g., during coma), treatment focuses on:

- Getting general responses to sensory stimulation like tactile, auditory, noxious and visual stimulus.
- Guiding family members on how to increase the responsivity of the child and use the most effective sensory stimulus.

As the child becomes more aware, treatment focuses on:

- Developing attention-concentration for basic daily life activities,
- Reduce confusion and agitation.
- To improve orientation of the child to the date, time, place and situation.

Later on in recovery, treatment focuses on:

- Use of specific strategies to improve memory skills of the child (e.g., using a memory log);
- Teaching specific strategies to develop problem solving, logical reasoning, organizational and abstract thinking skills.
- Help the child to develop social skills in small groups in his/her family or locality and later on in school.
- To improve self-monitoring skills.
- Working with an educational rehabilitation specialist to help the child get back to school.

***Language Intervention***

- Language intervention for children with TBI takes into account the impairments in processing speed, working memory, and executive

function which may contribute to deficits in language (Ewing-Cobbs & Barnes, 2002).

- Language intervention also depends on the child's developmental level at the time of injury and the pattern of deficits that require intervention.
- For younger children, intervention tends to emphasize:
  - ✓ following directions,
  - ✓ phonological awareness,
  - ✓ vocabulary development, and
  - ✓ word fluency for early literacy skills.
- For older children and adolescents, the emphasis is often on inferencing, higher-level comprehension, narrative and discourse processes, and academic or vocational literacy (e.g., summarizing text, taking notes).

### ***Speech Intervention***

- Intervention for speech problems may focus on the individual speech subsystems of respiration, phonation, articulation, and velopharyngeal function or, more globally, on overall speech intelligibility, using behavioural and instrumental treatments, prosthetics, compensatory strategies, AAC, and/or environmental modifications (McDonald et al., 2014; Morgan et al., 2010; Morgan & Vogel, 2008).
- Children who are unable to use natural intelligible speech for communication (e.g., due to severe dysarthria or a voice disorder) may need long-term AAC (Doyle & Fager, 2011).
- In some children with TBI, a speaking valve may be used to facilitate voicing in children who are unable to speak because they have had a tracheostomy.

### **Psychological Intervention**

Cognitive issues in people with brain injury includes problems with attention and concentration, impaired learning, affected cognition and

problem solving skills. Apart from this, emotional and behavioral issues are also common which causes depression and anxiety, anger issues, inappropriate emotional responses and difficulty in controlling their emotions. Psychological Intervention for people with Traumatic Brain Injury is comprehensive focusing on the whole individual and not their specific condition. It involves an active participation of a Neuropsychologist who addresses the cognitive and emotional issues of the person by conducting tests which provides a clarification as to how the injury has changed brain's ability to process information and it also explains specific changes in behavior due to the injury. The results of these tests regulate rehabilitation needs and appropriate treatment guidance. Cognitive exercise are taught which emphasizes on compensatory strategies focusing on each individual's unique strengths to balance their deficits. Psychotherapy also plays an important role which focus largely on teaching them to adjust and accept their disability. Intervention is provided either one-to-one or in a group.

### **Nutrition**

- It is very important to closely monitor nutrition status of Traumatic brain injury patient
- If a patient with TBI is not able to swallow food nutrition support is initiated through tube which is inserted through nose (Ryles tube) or Stomach (PEG- percutaneous endoscopic gastrostomy tube)
- There are two ways in which feeding is done through tube:
- 1.continuous feeding- in this liquid feeding formula is given trough continuous feeding for 12-24 hours depending on patients tolerance using feeding pump.
- 2.Bolus feeding- In this liquid feeding formula is given via syringe at intervals of 2-3 hours.at time not more than 300ml of feed given including flushing.
- As the patient recovers slowly oral liquids are started and gradually soft food as per patient's tolerance.
- Nutrition is also important during rehabilitation phase. If necessarily addition of nutrition supplement is important.

## **Stem Cell Therapy**

The standard treatment modalities for brain injury involves medications, physical and behaviour therapy, Hyperbaric Oxygen therapy(HBOT), and medical management of associated conditions aims at improving the functional abilities and restoring the patient's daily life. However, these strategies have failed to translate into a successful treatment strategy that can address the core neurological damage. Among the numerous barriers to finding effective interventions to improve outcomes after TBI is the severity and heterogeneity of the injury . In chronic TBI, there is high prevalence of residual neurodeficits rendering the patient dependent and many a times bed-ridden. In case of mild TBI, physical recovery is witnessed with the standard treatments, but the memory and behavioural issues can cause distress at home and workplace. The available pharmacological modalities manage these disabilities, but their effect wears off gradually.

Due to the brain's limited capacity to regenerate the damaged neurons, the intervention should aim at halting the degeneration and replacing the lost and damaged neurons. Stem cells migrate towards the damaged areas of the brain and initiate the repair process. They promote angiogenesis, axonal remodeling, neurogenesis and synaptogenesis, which may help reverse the pathology of TBI. These cells differentiate into various cells including neural cells, oligodendrocytes, etc. In TBI, there is loss of myelin which disrupts the signal transduction and damages the axons. The oligodendrocytes help in remyelination of the damaged axons and repair the disrupted neural connections. Bone marrow cells also produce various growth factors and neurotrophic factors such as brain-derived neurotrophic factor (BDNF), nerve growth factor (NGF), vascular endothelial growth factor (VEGF), basic fibroblast growth factor (bFGF), which stimulate the endogenous neuroprotection and repair.

## Chapter 11

# Post Encephalitic Cerebral Damage

Encephalitis is an inflammation of the brain tissue. Commonest cause of encephalitis is viral infection with viruses like herpes simplex, mumps, Epstein-barr virus, Human immunodeficiency virus, cytomegalovirus. Infection due to chickenpox, measles and rubella virus can be prevented with timely vaccination. It could occur due to bacterial and fungal infection as well.

Primary encephalitis: It is caused due infection of brain and spinal cord tissue.

Secondary encephalitis: It is caused when the infection from other parts of the body is transmitted to the brain

Encephalitis can be fatal and cause death or permanent disability therefore it is important to seek immediate attention of the doctor.

Children below the age of 1 year and with poor immunity, malnutrition, chronic-gastrointestinal disturbances and premature babies are at a higher risk of encephalitis.

## **What are the symptoms of encephalitis?**

### **Mild symptoms include:**

- fever
- headache
- vomiting
- stiff neck
- lethargy (exhaustion)

### **Severe symptoms include:**

- High grade fever above 103 degrees F.
- confusion
- drowsiness
- hallucinations
- slower movements
- coma
- seizures
- irritability
- sensitivity to light
- unconsciousness
- Continuous crying
- Vomiting
- Poor appetite
- Body stiffness

## **How to diagnose encephalitis?**

With the help of clinical symptoms and special tests like spinal tap or lumbar puncture to find out any infection to the brain, Computed tomography (CT) scan and Magnetic resonance imaging (MRI) to show which areas are damaged and Positron emission tomography scan (PET – CT) scan to find out the extent of brain damage. Blood test and brain biopsy to detect the type of virus causing infection.

## **Post encephalitic cerebral damage**

Due to the inflammation to the brain there is direct injury to brain tissue. As the brain tissue inflames and exudates accumulate inside the rigid cavity of skull these tissue undergo secondary injury due edema,



ischemia and hypoxia. Such injury can cause short term or long term neurological symptoms.

This damage can cause several symptoms that may resolve in few months or persist long term or may be irreversible. These symptoms are

- Loss of memory
- Behavioral/personality changes
- Epilepsy
- Fatigue
- Physical weakness
- Intellectual disability
- Lack of muscle coordination
- Vision problems
- Hearing problems
- Speaking issues
- Coma
- Difficulty breathing
- Death
- Spasticity
- Altered sleep pattern
- Impulsive emotional behavior

## **Physiotherapy**

### **1. Prevent chest complications-**

- Breathing exercises which includes strengthening of respiratory muscles with the help of spirometer, cervical and thoracic mobility exercises.
- Postural drainage and chest percussion - In postural drainage and chest percussion, the patient is rotated to facilitate drainage of secretions from a specific lung lobe or segment while being clapped with cupped hands to loosen and mobilize retained secretions that can then be expectorated or drained. The procedure is somewhat uncomfortable and tiring for the patient.
- Suction- Suctioning clears mucus from the tracheostomy tube

and is essential for proper breathing.

**2. Prevent DVT (Deep vein thrombosis)-**

- Limb elevation, active limb exercises and bed mobility exercises help in preventing DVT.

**3. Correct Deformity-**

- Positioning of the limbs play an important role in preventing deformity which can be achieved by splinting and casting.
- Sustained stretch of the tightened muscle and strengthening of opposite group of muscle help reduce the deformity.

**4. Improve vital function-**

- Positioning the patient to increase the air intake to targeted lobes.
- Keeping the neck in slight flexion improves respiratory capacity.
- Facilitate swallowing with positioning and proper oromotor stimulation.

**5. Normalize tone-**

- Facilitatory and inhibitory techniques are commonly used to normalise tone.
- Facilitatory technique includes vibration, stroking, joint approximation, quick icing and quick stretch.
- Inhibitory technique includes sustained stretch, joint traction and prolonged iceing.

**6. Improve voluntary movement-**

- Practicing bed mobility activities such as rolling, sitting, quadruped, kneeling, half kneeling, crawling and getting up and sit.
- Neuro development techniques (NDT) and Proprioceptive Neuromuscular Facilitation Technique (PNF) to improve their

functional abilities.

- Balance training focusing on sitting and standing balance to help maximise mobility.
- Stretching and strengthening program to relieve stiffness and increase muscle strength which will make everyday tasks much easier.
- Exercise to improve weight shifts and all the dissociations.
- Open kinematic chain exercise to improve mobility and closed kinematic chain exercise to improve stability.
- Problem solving task to challenge their functional ability.
- Gait training- walking with walking aids.

#### **7. Sensory integration**

- Stimulation by combined proprioceptive, auditory and visual input by training in different environment, Motor Relearning Programs and Sensory Re-education.

#### **8. Educating parents-**

- Manual handling training sessions for carers and families which will include therapeutic handling, transfers and positioning techniques to increase comfort and promote postural alignment.

### **Occupational Therapy**

Encephalopathy secondary to any infection may affect the overall cerebral function. These may affect an individual physically, cognitively, educational, vocational, may become dependent for self-care etc. Its prognosis is not predictive and so may make a person and their family members anxious about their life and wellbeing. They may also have various secondary complications like seizures, contractures/deformity, bed sores etc. Occupational therapist always has a holistic approach towards managing a person with any disease condition. It helps a person to adapt, modify or improve on their skills to perform functional activities.

**Areas of concern for Occupational Therapist are:**

1. Sensorium
2. Physical issues
3. Sensory issues
4. Cognitive-perceptual issues
5. Behavioral issues
6. Prevention of secondary changes
7. Orthosis/Splints
8. Adaptations/ Adaptive techniques
9. Basic Activities of daily living
10. Instrumental activities of daily living
11. Leisure
12. Roles
13. Routines
14. Habits
15. Education and vocational choices
16. Family or caregiver guidance

Sensorium: most of the patients post encephalopathy may remain in altered sensorium or may not be aware about what is happening around them. Occupational therapist may use different environmental stimulation techniques to improve their awareness about self and environment.

Physical issues: due to diffuse cerebral damage, there movements and voluntary control may get affected. It can be only weakness or can be related to tonal changes. Occupational therapist then may use different techniques to facilitate voluntary control and some movements.

Sensory issues: they may at times have sensory disturbances which can be related to sensory processing or sensory perception. Occupational therapist may manage such a issues through sensory re-education or through sensory integration.

Cognitive-perceptual issues: most of them may deteriorate in terms of their cognitive functions, so our role is to retrain them in cognitive-perceptual functions. We may also provide some aids to help with cognitive functions and to function well in day to day activities.

Behavioral issues: these patients may also demonstrate lot of behavioral issues in terms of aggression, mood swings or tantrums etc. this is usually managed using different behavioral and cognitive behavioral approaches.

Prevention of secondary changes such as contracture or deformity and bed sore through use of appropriate positioning and use of some orthotic devices if required. Also, movements of all joints at regular intervals are very important.

Orthosis and splints: are prescribed to prevent or correct a contracture. It can also be given improve a function at a limb.

Adaptations/ Adaptive techniques: these can be in terms of environmental modifications or adaptations to improve function in day to day activities.

Basic activities of daily living: people with encephalopathy usually become dependent for self-care activities due to risk of fall, or families may become over protective. In such conditions, patient and their caregivers can be motivated to allow them to participate in basic self-care activities in safe environments and with cues or with assistance.

Instrumental activities of daily living: Due to physical limitation family members may not allow them to participate them in other day to day activities due to fear of fall or some other injury. So, the family members should be advised to help them improve their participation according to their capabilities and to help them wherever required. Therapist may suggest modified ways to perform these tasks or with some type of adaptations if required. Some of the activities the patient may not be able to perform or may not be safe for him to perform, so they may have explore alternate activities for improving participation.

Leisure: it is one of the important area where an occupational therapist considers. Most of the people avoid getting involved in different leisure pursuits as it poses the risk of some injury. Occupational therapist discusses about different leisure interests and advices them about the safest activities to get involved in how they can get involved in leisure pursuits without harming them.

**Roles:** we also help them to participate in their different life roles such as a mother, student, homemaker, etc.

**Routines:** Most of the people have disturbed routine patterns. They may get involved more into non-productive activities (like resting) and reduce their productivity in life. We help them to develop a productive routine and improve their participation in meaningful activities for example a student need to attend school regularly, has to study on regular basis, should have appropriate sleep rest pattern, etc.

**Habits:** it is always advised to have good dietary intake, have good amount of sleep, avoid exertion or being exhausted, avoid substance use etc. As this may pose risk of some other problems like seizures.

**Educational and vocational choices:** it is also important that they lead a meaningful life and they have some aspirations in life. Occupational therapist may help assess their abilities and skills to help them make appropriate and safe career choices.

**Quality of life:** Irrespective of the condition, it is important that the person and their family members lead a quality of life. Occupational therapist may advice certain modifications or provides adaptations that may reduce the burden on the caregivers and the person thus improving their satisfaction and quality of life.

**Family and caregiver guidance:** It is very important that the families are aware about the condition and its implications. It is especially important in Indian scenario that the families are counseled and guided well at each step. They should be explained about the importance of taking regular medicines and at regular intervals. At times, the families may become over protective which may further hamper the development of the person. Therefore, an occupational therapist may help educate the families about the condition and how to manage a family member with encephalopathy.

## **Speech Therapy**

Common speech disabilities found in PECD are aphasia, dysarthria and apraxia.

**Aphasia** is a language impairment that affects the production or

comprehension of speech and the ability to read or write.

There are two general categories of therapies provided:

- Impairment-based therapies are aimed at improving language functions and consist of procedures in which the clinician directly stimulates specific listening, speaking, reading and writing skills.
- Communication-based (consequence-based) therapies are intended to enhance communication by any means and encourage support from caregivers. These therapies often consist of more natural interactions involving real life challenges in communication.

**Dysarthria** occurs when areas of the brain controlling our muscles for speech are damaged. A speech pathologist can help with strengthening muscles, increasing movement of mouth and tongue and breathing exercises. Common techniques are focusing on slow clear speech with frequent pauses, and starting a topic with a single word first and then checking whether the other person has understood.

**Apraxia** occurs when the speech muscles are unaffected but the brain has trouble sending the signals to the muscles responsible for speech. In mild cases, therapy involves saying individual sounds and thinking about how the lips and tongue should be placed, or speaking while clapping to improve timing. In severe cases, alternative systems such as gestures, facial expressions, written communication or pre-printed cards are used.

### **Psychological Intervention**

The literature strongly suggests that after cerebral damage, children are at an increased risk of adverse cognitive and behavioral outcomes. Long-term physical, cognitive, and behavioral impairments are the factors that most commonly limit a child's reintegration into the community. Neurocognitive problems related to PECD often involve deficits in attention, processing speed, anterograde memory, emotional recognition and executive functions. Emotional concerns following PECD tend to include emergence of internalizing and externalizing problems, aggression, impulsivity and hyperactivity, disinhibition, poor social judgment, apathy, withdrawal, anxiety and depression. Families are quite directly impacted by one of its members suffering from PECD. In

particular, family members and caregivers experience their own symptoms of increased stress, emotional suffering, and strained coping as a result of PECD.

Psychological management for children with PECD involves cognitive, behavioral and other directive methods. Strategies that may be employed when working with a child impacted by PECD and his/her family involve: cognitive retraining, enhancing effective communication and social skills, managing and reducing problematic behaviors while augmenting more adaptive behaviors, strategizing for optimal academic performance, building skills and creating organizational strategies to compensate for persistent deficits in memory, attention, and working memory. Helping the family understand the impact and expected effects of PECD with psycho-education appropriate for the age and cognitive development of the child affected may dispel the fear along with expanding the social support and perhaps encouraging participation in support groups. Caregivers should be encouraged to provide positive environments and parenting styles, which may help reduce chronic behavioral problems after brain damage. In addition, structured problem-solving family techniques may be helpful to address the chronic stress and unique barriers faced by families. The helpful effects of these approaches may positively impact not only the children themselves, but also their families.

### **Nutrition**

- Well balanced diet with inclusion of antioxidants is important.
- Some patients may have swallowing difficulties so diet needs to be modified as per patients tolerance.
- Mechanically soft diet or thick puree diet can be included.
- If oral feeding is difficult in some patients tube feeding can be initiated.
- Inclusion of seasonal fruits and vegetables is very important.
- Protein rich food like chicken , fish, pulses, almonds, walnuts should be included in diet.
- Processed and preserved food should be limited.



- If patient is underweight additional supplementation can be added.

### **Stem Cell Therapy**

Conventional treatments for PECD do not address the underlying core neuropathology of the disease. In PECD, there is loss of brain tissue cells leading to neuronal deficits. Due to the ability of stem cells to self-renew, multiply and differentiate into neuronal cells, it is able to work at cellular level and replace the lost and damaged cells. It restores the neuronal connectivity and thereby neuronal functions by remyelination, synaptogenesis and neurogenesis. It reduces the inflammation and regulates the immune system. It reverses hypoxia by promoting angiogenesis and improving blood and oxygen supply to the brain. Through paracrine mechanisms, stem cells also halt further cellular damage.

## Chapter 12

# Poliomyelitis

Poliomyelitis, also known as Polio is a contagious disease caused by poliovirus. The poliovirus affects the anterior horn, motor neurons of the spinal cord and brain stem, causing a rapid development of paralysis. Children younger than 5 years are more likely to contract the virus.

Transmission of the virus may occur by:

- fecal-oral contact resulting from poor hand washing or through ingestion of contaminated food and water.
- Contact with infected respiratory secretions.

The disease can manifest in 4 different forms:

- 95% of people that contract the poliovirus remain asymptomatic. This is called as *subclinical polio*. However, even though asymptomatic, people so infected by the virus may spread the virus and cause infection to others.
- Approximately 4-8% of people that have contracted the virus, may demonstrate flu-like symptoms including fever, sore throat and fever, nausea, abdominal pain or constipation lasting between 1-10 days. This is called *non-paralytic* or *abortive polio*.
- In 1-2% of infections, *non-paralytic aseptic meningitis* is produced, with symptoms of stiffness in the neck, back and/or legs. These last 2 to

10 days and recover completely.

- About 1% of those affected, develop into *paralytic polio*. Symptoms include, loss of reflexes, severe muscle aches and muscle spasms in the arms, legs or back. In children, the initial symptoms may fade before paralysis appears.

### **Problems associated with paralytic polio:**

- Paralysis may affect any muscle of the body, but is most common in the legs.
- Affected limb may develop contractures with restriction of joint mobility
- Growth retardation of the affected limb
- Paralysis does not worsen with time, however, curvature of the backbone and dislocations may occur
- Urinary tract infections

### **Diagnosis:**

- Recovery of virus from cultures of throat washings, stools or cerebrospinal fluid.
- Test for increasing levels of antibodies to the poliovirus.
- Increased white blood cell count which is a general response to an infection.

### **Treatment:**

Polio is completely preventable by vaccination. There are 2 polio vaccines that are available; inactivated poliovirus vaccine and oral poliovirus vaccine.

For those that have contracted the virus, there is no specific treatment for this viral infection. Children severely affected, may need lifesaving measures, especially help with breathing. Treatment for paralytic polio is symptomatic and may include antibiotics for urinary tract infections and, physiotherapy and occupational therapy that provide strengthening exercises and assistive devices for walking.

## **Physiotherapy**

The clinical Course of Poliomyelitis can be divided into three stages:

1. Acute Stage
2. Convalescent Stage
3. Stage of Residual Paralysis

### ***Acute Stage***

The acute stage generally lasts for 4-8 weeks. The symptoms usually include fever, diarrhea, nausea, vomiting, irritability, joint pain and muscle tenderness.

The treatment strategies in this stage includes:

- i. Rest: Minimal handling will help in decreasing the chances of aggravation of symptoms.
- ii. Isolation: The urine, stools and droplets are highly contaminated and contagious. The other children should be kept protected from coming in contact with it.
- iii. Booster Dose: The other children in the house should be given a booster dose vaccination.
- iv. Nutrition: Protein – rich diet is recommended.
- v. Correct Handling Technique: The child should be carried in such a way that hip should be in extension and no abduction. Avoid lifting the child by his hands.
- vi. Splintage and correct positioning: Splinting of lower limbs will help in preventing muscle damage. It also decreases pain. Contractures can also be prevented.
- vii. Sister Kenny's Bath: In this technique, towels are dipped in hot water and wringed well. These towels are placed on lower limbs and spine. This is a form of wet and moist heat, which helps in resolving inflammation to some extent.
- viii. Gentle Passive Movement: These should be done 2-3 times a day depending on the child's tolerance. However, this should be avoided if there is a lot of muscle tenderness.

### *Convalescent Stage*

The convalescent stage starts from 2 months and continues for 2 years. This is the stage of true or actual paralysis. It is further divided into 2 stages viz. early convalescent stage (up to 3 months) and late convalescent stage (up to 2 years)

The treatment strategies in the early convalescent stage includes:

- i. Continuous Splinting: L – splint can be given so as to avoid knee flexion deformity and equinus deformity. A lumbar corset can be given so as to prevent abdominal hernia.
- ii. Positioning: Carrying the child in Indian position should be avoided so as to avoid IT band contracture. Also while sleeping, putting the child in prone position helps to prevent hip contracture. If the upper limb is affected, then the shoulder should be kept abducted. Axilla rolls can be used to prevent subluxation of shoulder.
- iii. Changing of position: A severely paralyzed patient should be turned every 2-3 hours so as to prevent bedsores and keep the area dry.
- iv. Stretching of contractures: The tightened and contracted soft tissues should be constantly and gently stretched.
- v. Stimulation and facilitation techniques: Interrupted Galvanic stimulation may be given so as to maintain the muscle integrity. Brisk stroking can also be given so as to facilitate the affected muscles.

The treatment strategies in the late convalescent stage mainly includes strengthening of the weak muscles. The muscles can be strengthened to its maximum capacity up to 2 years post onset of the disease. The already active muscle may however show some improvement even after 2 years. It is the paralyzed muscles, which become difficult to activate after 2 years. Some of the techniques used to strengthen the paralyzed muscles are:

- I. Resisted exercises using pulleys, springs, therabands, weight cuffs.

- ii. Suspension therapy
- iii. Hydrotherapy
- iv. Play therapy
- v. Sensory Integration

### ***Stage of Residual Paralysis***

The paralysis that persists after 2 years is usually permanent. The extent of residual paralysis may range from mild insignificant local weakness to gross paralysis of trunk and limbs muscles attributing to severe disability and functional dependency.

The goals of physiotherapy in this stage are:

- i. Strengthening of all the innervated muscles using pulleys, springs, therabands, weight cuffs.
- ii. Preventing contractures and deformities by doing regular stretching exercises and splints.
- iii. Prescribing appropriate orthosis to improve the gait pattern.
- iv. Making the patient as independent as possible.
- v. Emotional and psychological support.

### **Occupational Therapy**

Poliomyelitis, often called polio or infantile paralysis, is an infectious disease caused by the poliovirus. The weakness most often involves the legs but may less commonly involve the muscles of the head, neck and diaphragm.

Treatment of polio often requires long-term rehabilitation, including occupational therapy, physical therapy, braces, and corrective shoes and, in some cases, orthopedic surgery. Occupational therapists assess the impact of polio on muscle strength, joint stability, posture and cognition on a person's capacity to manage daily life tasks. Intervention improves participation in meaningful roles, tasks, and activities; remediates deficits; minimizes secondary complications; and provides education and support to the patient and caregivers. Occupational therapists focus

on independence and function, individual goal-setting, and their specialist skills in task adaptation and environmental modification.

Once polio is diagnosed, occupational therapists set the goals as follows:

### **Short term goals**

- Patient education:
- Educating the patient about the prognosis of illness is the first and foremost goal as this enables the patient as well as the therapist to set realistic occupational goals in therapy.
- Postural correction:
- Providing a proper seating arrangement is essential. Use of braces and orthoses to prevent and correct deformities of spine like kyphosis, scoliosis.
- Correction of posture enhances the participation of the person in daily activities.
- Use of splints, braces for further prevention of contractures and deformities.
- To maintain and improve vital capacity
- Incentive spirometry, Activities like blowing candles and bubbles can be given.
- To Maintain the available muscle power and range of motion through functional, therapeutic and play and leisure activities.
- Use of activities like inclined sanding,
- To Facilitate and improve motor control and hand function in weak muscles

### **Long Term Goals**

#### ***ADL training:***

Self-care activities :

Bathing - for clients with difficulty in standing , a chair can be advised

along with a hand-held shower. Grab bars to prevent falls. Anti-skid mats

Toileting – Use of commode, Providing jet spray.

Grooming- if the hand functions are affected, a universal cuff and built up handles for comb etc can be advised

Eating - built-up handled spoons, swivel spoons and forks, Glass holder.

***Work activities:***

Providing specially designed scooters and cars can be used for community mobility

Ergonomic chairs to accommodate for postural deformities

Adaptations at work place like obstacle-free environment to prevent falls and shelves at low height to accommodate for reduced range of motion and to prepare the home and work environment according to the patient's requirements.

***Play and leisure activities: -***

Specially designed wheelchairs for participation in sports like football , tennis.

Occupational therapists work collaboratively with the patient to establish the impact of polio on the performance of daily tasks, including personal cares, domestic tasks, and work and leisure activities, and develop a goal-focused program to develop the required skills for participation in daily life.

**Psychological intervention**

Psychological intervention is not something that comes to mind when we think of polio. It's a medical condition that basically involves physical and ambulatory difficulties. However, the term disability involves psycho-social connotation. Two persons with the very same medical condition have differing levels of perceived disability. One may be employed and a contributing member of the society while the other may be unemployed and even dependent for his activities of daily living. The difference can be due to a variety of factors, some of these are:



- Socio-economic status
- Educational back-ground
- Social support
- Psychological factors like
  - A) Motivational level
  - B) Positive vs negative outlook
  - C) Locus of control
  - D) Personality traits (for eg neuroticism, openness, agreeableness etc)

A person who is high on emotional stability (i.e low on neuroticism) will be able to cope better with a disability as compared to someone who is not. Similarly, a person who believes that the source of an event is internal (internal locus of control) will be less likely to get hopeless and will work to bring about changes s/he would like to see.

As a child grows up with limited physical abilities, a number of secondary issues emerge. The child may develop low self-esteem, low confidence levels and a negative approach towards his/her environment. Such children are often bullied, which is also a major contributing factor to the aforementioned emotional issues. This in turn leads to difficulties in social interactions and reciprocity. Quite a few times, such children, or adults need to be taught assertiveness through assertiveness training and appropriate social interactions through modeling. Person centered therapy can be provided to deal with major issues in self image, esteem and confidence. 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 better at articulating their feelings, reflecting upon them with increased abstract thinking, and participate socially.

Many children with disabilities discontinue schooling as climbing stairs are extremely difficult for them. The right of every child to education is violated in such a situation, despite having a simple and easy solution I.e. arranging a classroom on ground floor. Sometimes, due to low self-esteem and bullying, the child develops a disinterest towards academics.

Over a period of time, these emerge as major deficits in scholastic performance. This can be indicative of slowed cognitive development and corrective measures in form of cognitive exercises may be required.

As is the case of any disability, a positive out look is very important. Especially in case of illness like polio which is not only *acquired* but is also easily preventable in today's age, the question of 'why me?' is sure to arise. The answer to this question doesn't help in alleviating the difficulties associated with polio, the question 'why me?' doesn't deserve the energy and time we waste in finding an answer to it, for an answer to this question, doesn't really exist. This energy is better utilized by making accommodations to increase the functioning and efficiency of the person.

The second dimension of this is the anger that one feels for his/her parents with regards to negligence about vaccination. It creates a lot of conflict as such emotions feel unacceptable towards one's parents. Suppressing such emotions doesn't help much. However painful it is, it needs to be accepted that parents too are humans, and they can make mistakes, unintentional mistakes that may have a debilitating consequence. Often, talking about it, the pain and the anger with one's parents can be helpful. They too have been carrying the burden of guilt for years and probably want to relieve themselves as well. Occasionally, a counselor's help may be required to facilitate communication. Many counselors prefer to use family counseling and family systems therapy.

Many of us have difficulties in accepting ourselves. There are many things we dislike about ourselves. Sometimes, these dislikes transcends to a dislike for ourselves as a whole. We keep ruminating about those very aspects forgetting the strengths we possess. Same applies to people who have polio or for that matter any disability. The physical limitations consume so much of our time and energy, our focus remains on things we can't do, or things we could do if ...'; we forget our strengths or most importantly the fact that the said disability is just a facet of the multitude of factors that contributes to who I am. And I am, the best me I could be. I deserve to be accepted unconditionally-the goal of person centered therapy.

## Speech Therapy

Patients suffering from Post Polio Syndrome (PPS) with a bulbar involvement often show symptoms related to speech (mainly voice or phonation) and swallowing but at a later stage.

### Symptoms:

If there is a bulbar involvement in PPS then surely the swallowing and phonation functions will be compromised. Other processes of Resonation and Articulation are also affected. Overall, All functions related to Speech and swallowing will be affected which include:

- ✓ Phonation: Reduced vocal loudness Vocal fatigue Inability to modify/ change/ alter pitch and loudness qualities of voice.

Voice quality might be hoarse or more breathy

- ✓ Resonation: Hyper-nasal voice due to weak/ or totally absent movements of the Soft palate.
- ✓ Articulation: Difficulty in producing sounds correctly (Misarticulation) due to structural abnormalities in the oral cavity.
- ✓ Respiration: Weakness of the muscles involved in respiration which include the thoracic muscles as well as abdominal muscles which leads to in coordinated breathing during speech and indirectly affects the Speech Intelligibility of the individual.

To improve the swallowing functions as well as preserve the muscles involved in swallowing, phonation and respiration from degeneration a Speech Therapist needs to be involved early to perform an evaluation of the integrity of the muscles involved in speech and swallowing as well as safety of swallowing.

Safety of swallowing should be looked into as serious matter because if there is a swallowing dysfunction known as Dysphagia, it could lead to aspiration during swallowing and later on cause Aspiration Pneumonia which could result in choking during feeding or choking on his/ her own saliva.

## **Management of speech and swallowing difficulties in Polio:**

- **Swallowing:** The treatment of swallowing issues or Dysphagia is a crucial part in the management of individuals with PPS. The best way to treat a swallowing disorder, is to swallow! By refraining from swallowing or put as NPO for an extended amount of time, the swallowing muscles can atrophy and weaken. To improve swallowing functions, maintain airway safety and ensure a safe swallow , various techniques can be used.

*Management of swallowing disorders is mainly divided into 2 parts:*

**Direct strategies** - refers to treatment that involves food

**Indirect strategies** - refers to an exercise regimen performed without a food bolus.

Direct techniques include modifications of food consistency.

Indirect techniques include stimulation of the oropharyngeal structures and the adoption of behavioral techniques, such as those involving postural changes or the swallow maneuver.

### ➤ **Compensatory Strategies**

Diet Modifications based on what the patient can manage safely.

- Thin/Thickened Liquids: nectar thick, honey thick, and pudding thick.
- Thin/Thick puree food
- Regular diet
- Chopped/Mechanical soft diet.
- Alternative texture/temperature.
- Alternate liquids and solids: to eliminate residue.
- Throat clearing: to eliminate residue.
- Tube Feedings.

**Consider environmental/stress factors.**

- When/where/how the patient is eating/being fed
- Consider patients position during meals
- Most importantly, consider patients etiology and how it may be affecting PO intake

➤ **Behavioral Approaches for management of Dysphagia**

- Alternate small bites with small sips.
- Small bites/small sips
- Liquid wash to clear oral, pharyngeal, valleculae, etc. residue.
- Dry swallow - clear residue
- Multiple swallows - to clear residue

➤ **Postural Changes**

Postural strategies are used to help change the way bolus flows through the swallowing mechanism.

Specific postures are used to compensate for particular types of Dysphagia by changing the way that the food moves through the pharynx. It is a good idea to have the patient try using these postures during the VFFS/MBS; this way you can get an idea of how well or what will really work or not work for that patient.

- ✓ **Head Tilt:** Move the head to better side, bolus is redirected through oral cavity and oral bolus transport is improved.
- ✓ **Head Rotation:** Twist head to weaker side, so weaker side is closed off and bolus travels to stronger side. Avoids pocketing as well.
- ✓ **Chin Tuck:** Put chin down to move bolus anterior. It prevents premature spillage and widens the valleculae so spillage hesitates there giving more time for Vocal Fold's to close thereby reducing the risk of aspiration.
- ✓ **Head Back:** Bypass oral stage by utilizing gravity to clear oral cavity.

➤ **Therapeutic Dysphagia Strategies**

- **Sensory Strategies:** Sensory strategies include changing volume, texture, temperature, or taste therefore changing the sensory feedback provided by the bolus

- **Thermal Stimulation.**

- **Suck-swallow technique (Simulation)**

➤ **Motoric Coordination**

- Various exercises can be done to improve the range of motion (ROM) of the lips, tongue, and jaw, to improve coordination, to improve vocal fold adduction, laryngeal elevation, or tongue base retraction.

**Range of Motion exercises** assist with structural/tissue damage

**Resistance exercises** strengthen muscles

**Resistance Exercises** involve pushing against tongue depressor or spoon to create "resistance"

**Falsetto Exercises** used to increase laryngeal elevation. Involve repetitive /i/ or /ng/.

➤ **Swallowing Maneuvers**

Various swallowing maneuvers are used to change the swallow physiology. *Each swallow maneuver is utilized for very specific diagnoses and should only be used after discussing with your Speech Language Pathologist.*

- Supraglottic Swallow
- Super-Supraglottic Swallow
- Shaker Manuever
- Masako
- Effortful Swallow
- Mendelsohn Maneuver
- Jaw Opening Exercise

➤ **Instrumental**

There are different types of instruments to provide immediate feedback such as Surface Electromyography (SEMG), tongue bulbs and/or Vital Stimulation or EStim. Most of the time Instrumental treatment is done along with a Compensatory Strategy, Postural Change, or some sort of Stimulation therapy.

- ✓ **BioFeedback:** SEMG - electrodes placed on thyroid lamina while patient swallows and can visually see how strong/weak their swallow may be.

- ✓ **Muscle Stimulation:** Newer techniques such as NMES and Vital stim have been introduced and are more widely being used. Recent studies suggest that NMES is most efficient when utilized in conjunction with therapeutic Dysphagia strategies.
- **Phonation:**
  - ✓ To improve phonation use vocal relaxation exercises and Vocal function exercises (VFE)
  - ✓ Techniques to alter vocal loudness and pitch.
  - ✓ Humming with a forward focus.
  - ✓ To maintain vocal hygiene.
  - ✓ Follow modified vocal rest.
  - ✓ No vocal abuse or misuse of voice.
  - ✓ Reduce muscle strain on the neck muscles use of slow neck rotation exercises, shoulder roll technique can be used.
- **Resonation:**
  - ✓ Speak with an open mouth approach i.e. open your mouth as wide as possible.
  - ✓ Speak in a louder voice.
  - ✓ Practice more of activities involving blowing and sucking functions like whistle blowing, bubble blowing, sucking juices with a straw, etc
- **Respiration:**
  - ✓ Work on the thoracic and abdominal muscles for its strengthening.
  - ✓ Practice deep inhalation and exhalation exercises daily: Inhalation through mouth and exhalation through nose in lying down position.
  - ✓ Use of a Modified Respiratory Muscle Training Program.

## **Nutrition**

- It is very important to maintain well balanced diet for polio patient.
- Inclusion of protein rich food like chicken, fish, milk and milk products, pulses is important.
- Inclusion of lot of vegetables with green leafy vegetables is important.
- Also, inclusion of seasonal fruits is important.
- Healthy fats like coconut oil, sunflower oil is important limited amounts.
- Limiting processed food with sugary, starchy food is also essential.

## Chapter 13

# Spinal Muscular Atrophies

Spinal muscular atrophies (SMAs) comprise a group of autosomal-recessive disorders caused by a deficiency of a protein called SMN. Every individual has 2 SMN genes, SMN 1 and SMN 2. All patients with SMA have a homozygous disruption in the SMN1 gene on chromosome 5q. Deficiency of SMN protein causes progressive destruction of lower motor neurons in the brain stem and spinal cord that control muscle activity essential for speaking, walking, breathing and swallowing. The symptoms range from severe to mild depending on the amount of SMN protein there is. The later in life symptoms begin, the milder the course of the disease.

SMA is classified based on the age of onset:

- SMA type I: Onset is from birth to 6 months.

Symptoms include:

- Swallowing and feeding difficulties
- Impaired breathing
- Decreased muscle tone
- Diminished or absent tendon reflexes
- Diminished limb movements
- Fasciculations
- Tremors
- Affected children never achieve sitting or standing milestones.



- SMA type II: Onset is between 6 and 18 months.

Symptoms include:

- Children may be able to sit unsupported but unable to stand or walk unaided.
- Affected children may have respiratory difficulties
- Increased risk of respiratory infections

- SMA type II: Onset is after 18 months between 2 and 17 years of age.

Symptoms include:

- Increased risk of respiratory infections
- Abnormal gait
- Difficulty in running and climbing stairs and/or rising from chair.
- Tremor of fingers
- Joint contractures due to abnormal muscle tone and muscle weakness
- Curvature of spine called scoliosis

- SMA type IV is adult onset occurring at a mean age of mid 30s.

SMA Type I affects approximately 1 in 10,000 births; types II and III affect 1 per 24,000 births. SMA type II accounts for half of all the cases of SMA.

### **Diagnosis:**

Diagnostic tests include:

- Genetic test
- Electromyography (EMG) and Nerve Conduction Velocity Test (NCV) studies
- Muscle Biopsy

### **Treatment:**

- There is no cure for SMA. The mortality and morbidity associated with the disease is inversely correlated with the age at onset. In patients with SMA type I, survival is around 7 months with a mortality rate of 95% by 18 months of age. Death in type I and II are most often due to respiratory complications.

Treatment should be initiated as early as possible and consists of managing the symptoms and preventing complications. Muscle relaxants may reduce spasticity. Physical Therapy and Occupational therapy may help to improve joint mobility, posture. Therapy involves stretching and strengthening exercises and assistive devices such as braces, orthotics and wheelchairs to help retain independence.

## **Physiotherapy**

### **1. Chest Physiotherapy-**

- To maintain & strengthen the respiratory muscles.
- To remove secretion & clear airways by percussion, active cycle breathing techniques, breathing exercises including spirometry, coughing techniques, sometimes by suctioning.
- To prevent restrictive lung disease, secondary to scoliosis, stretching of shortened muscles of lateral trunk & improve breathing.
- A special machine that provides air through a mask or tube in most severe cases.
- Few patients may also need for supplemental oxygen & support of nocturnal non-invasive ventilation.

### **2. Neuromuscular & Musculoskeletal Physiotherapy:**

- To elongate shortened muscles by stretching
- To maintain joint mobility (passive ROM, Active- assisted ROM, Active ROM)
- Active assisted activities include suspension therapy & spring exercises for upper extremity as well as lower extremity.
- To recruit, activate & strengthen weakened muscles
- Gait training
- To prevent pressure sores & contractures/ deformities
- Wheelchair mobility
- To prevent further increase in scoliosis, spinal jacket is advised

## **Occupational Therapy**

Occupational therapist plays an important role in helping children with SMA to continue their work and schooling by using different approaches like preventive, adaptive approach and compensatory approach.

### ***Common Occupational Therapy goals focus on :***

1. To maintain vital capacity
2. Prevention of further contractures and deformities
3. Maintaining Range of motion and Muscle power
4. Maintaining / Improving functional independence
5. Maintaining the ambulation and mobility of the patient.
6. Energy conservation and Work simplifications
7. Home and environmental modifications

### ***Early Stage:***

- This is the phase in which patient can walk and able to do most of his ADL activities.
- Aim of Occupational Therapist is to maintain whatever function person has.
- Maintaining / Improving strength of UL by doing sling activities.
- Encourage the child to play all physical activities and outdoor games and encouraging the adult with SMA to participate in his daily routine tasks.
- Letting the person to take sufficient rest periods in between two physical activities.
- Encourage the child to do activities through play to improve balance and mobility.
- Prevention of fall by modifying the environment (Using nonskid mats, Shoes with good grips).

### ***Transitional Stage:***

- Giving functional activities especially overhead tasks as a part of therapy to maintain / improve functional independence.

- Focusing on giving activities to maintain /improve strength of UL
- Continuing sling activities with upper limbs
- Provide splints and assistive devices to make child/ Adult stand.

***Loss of ambulation:***

- Improving access to the classroom and home
- Providing ramps for easy maneuvering of the wheelchair
- Installing grab bars and handrails in and around the classroom and at home.
- Providing modifications like (footrest to the bench on which child is sitting), Use of assistive device like mobile arm support so that child can write and an adult person can perform work like typing, deskwork and do other functional activities.
- Providing the shelf to keep things. (height of the shelf should be below the shoulder level)
- Providing platform lift If the classroom is not on the ground floor.
- Child should not compromise his play activities so encourage the child to play indoor activities.
- Allowing the patient to participate in social activities
- Wheelchair modifications: Using light weight wheelchairs, use of appropriate cushions, electric wheelchair

***Late / Advanced Stage:***

- Patient becomes bed ridden in this stage.
- It becomes difficult for the child to attend school and attending the work for an adult person once the patient reaches this stage.
- It is important to continue education for the child. So, it can be done by providing the shadow teacher.
- Providing education online for children and providing work from home facility for adults keep the individuals engaged in their normal routine.

## Speech Therapy

Speech therapy is one of the most recommended treatment options for individual with spinal muscular atrophy as it delves much deeper than just basic language training. It includes various exercises and techniques.

Some of the most commonly recommended speech therapy techniques include:

- Augmentative and Alternative Communication (AAC) Devices: An AAC device is anything that is an alternative to oral communication. AAC methods are divided into two categories:
  - Unaided: Relies on body language  
Eg: gestures, sign language, etc.
  - Aided: Uses tools to help improve communication.  
Eg: paper and pencil, computers, speech generating devices, pictures or symbols, or communication boards/books.
- Articulation Therapy: Uses language cards to identify and focus on specific sounds. Often includes having children make sounds in front of the mirror so they can see how their mouths move.
- Blowing exercises to improve mouth muscles
- Breathing exercises to strengthen the diaphragm
- Jaw exercises to strengthen muscles
- Lip exercises to improve lip extension and strengthen muscles

Swallowing is another symptom of spinal muscular atrophy. Depending on the pathophysiology of the disease, swallow dysfunction (dysphagia) may occur at the oral, pharyngeal and/or oesophageal stage of swallowing. For example, when there is cerebellar involvement, dysphagia may be characterized by reduced coordination of the oropharyngeal muscles involved in swallowing food and drink.

Dyshagia management techniques:

- Modification of consistency of food or drink

- Introduction of safe swallow strategies including use of a chin tuck position, double swallow, throat clear
- Advice regarding sitting posture and set up for oral intake
- Introduction of care-taker-initiated prompts to maintain safety eg: slow rate, small sips, avoiding talking with food or drink in mouth
- Caring about oral hygiene

### **Psychological Intervention**

Children suffering from SMA require an adequate rehabilitation program that can help maintain their quality of life and maximize their physical and psychosocial functioning. Psychosocial functioning involves emotional, behavioural and social aspects and are believed to be central to an individual's quality of life. Psychologists make plans that are aimed at increasing motivation and will power. Psychological intervention can contribute in the areas of disruptive and obsessive-compulsive behaviors that may be present. Psychologists also conduct anxiety and depression scales to assess severity of distress faced by these children and provide intervention where needed. Family support groups are created to help parents share and learn from other parents and caregivers which in turn can help them to embrace their child's life. In very severe cases, anti-psychotic medications to treat co-morbid mental illnesses such as schizophrenia, bipolar disorder, depression, mania and severe anxiety are advised.

### **Nutrition**

- Patient with SMA have some gastrointestinal and nutritional complications due to overall muscle weakness.
- Feeding and swallowing problems, gastrointestinal dysfunction such as constipation, delay stomach emptying, sever reflex are also seen in these patients.
- Semisolid diet with thickened liquids will help reducing the risk of aspiration as comported to thin liquids.
- Children with SMA are less active, have less muscle mass and hence use less energy and calorie requirement is lesser.

- Adequate nutrition is also important to avoid cold and viruses that could turn to potential respiratory track infection.

### **Stem Cell Therapy**

All the conventional treatments for SMA only help in managing the symptoms of the disease to a certain extent. However, they do not address the underlying neuropathology. Stem cells have the ability to migrate towards the damaged areas and initiate the repair process. In SMA, replacement of damaged motor neurons is the ultimate goal of stem cell transplantation therapy. Stem cells protect the existing motor neurons and bring about regeneration and repair in the damaged motor neurons. They further demonstrate immunomodulatory, anti-inflammatory and cytoprotective properties. The factors secreted by these cells bring about neoangiogenesis. These paracrine effects lead to neuroprotection and subsequent alteration in the disease course and progression.

## Chapter 14

# Spinal Cord Injury

Spinal cord injury is a devastating event that occurs suddenly and whose consequences range from minimal symptomatic pain to a tragic quadriplegia. If cervical spinal damage is severe, it leads to quadriplegia, whereas an injury to the thoracic or lumbar spine leads to paraplegia.

In children, SCI is relatively a rare condition as compared to its prevalence in the adults. Nevertheless, up to 5% of spinal cord injuries occur in children. Diagnosis and treatment of SCI in children is challenging due to their age and behavioral differences. The type of SCI in children are different from that of adults as the anatomy and the mechanics of the spine varies until the child reaches 8-10 years of age. The primary causes of injury are birth related injuries, child abuse, falls or motor vehicle collisions. Young children also seem more vulnerable to post infectious or inflammatory cervical spine issues.

There could be complete disruption or contusion, compression or penetration of the spinal cord leading to necrosis, demyelination, axonal loss and glial scarring. The demyelination of axons may lead to a permanent loss of sensorimotor functions affecting the quality of life of these children. Complete recovery of the damaged spinal cord is very difficult, as it does not have the ability to regenerate lost or damaged neurons and re-establish the neural connections. The scar also consists of axonal growth inhibitors which further limit the repair and regeneration



process. As a result, there is no cure for SCI available presently.

The current treatment for SCI includes surgical interventions, medicines and rehabilitation. Their main goal is to stabilize the spine and prevent any secondary complications.

### **Treatment**

The acute management of the child with spinal cord injury requires rapid restoration of airway, breathing, and circulation. The child should be immobilized supine in a hard collar and on a fracture board. A full spine survey should be performed to eliminate possibilities of fracture and or dislocation. If there are no fractures or dislocation, a CT is performed to rule out an occult fracture, followed by an MRI. Good quality flexion-extension films are also obtained to rule out overt ligamentous instability. Children with severe spinal cord injuries diagnosed within 8 h of injury are administered a 24-hour course of methylprednisolone.

Early surgical intervention is required with definite spinal cord compression and progressive worsening of neurological deficits secondary to a fracture, epidural hematoma or extruded disc causing compression. Delayed or a planned intervention is advised for correction of spinal instability or kyphosis/scoliosis. Reduction of the fracture segments and fixation using spinal instrumentation is then performed according to the type of lesion.

Despite the surgery, many patients are left with neurological deficits, which may recover to some extent with regular rehabilitation.

### **Physiotherapy**

- Physical therapy can help in retaining muscle strength and flexibility, enhance coordination, reduce spasticity, regain greater control over bladder and bowel function, and increase joint movement. It can also help to reduce the likelihood of pressure sores developing in immobilized areas. Individuals are also taught to use assistive devices such as wheelchairs, canes, or braces as effectively as possible.
- **Reduce risk of pressure ulcers-** Prevention like frequent positioning, use of water/air mattress, use of roho/ air filled

cushions in sitting position, deep manipulation over the pressure sites should be prescribed. Patient should be encouraged to inspect the pressure site at regular intervals.

- **Pain reduction/ management-** Electrotherapeutic modalities like TENS, IFT can be use to alleviate pain. Relaxation exercises also help to reduce pain in patients with transverse myelitis.
- **Stretching programme-** Contractures are a common complication of neurological conditions, and are characterized by a reduction in joint mobility. Stretching is widely used for the treatment and prevention of contractures. Spasticity is also a major concern in transverse myelitis; Prolonged stretching helps in reduction of spasticity.
- **Strengthening exercises-** Weak muscles can be strengthened with suspension therapy, Push up bars, theraband exercises and weight training.
- **Bed mobility exercises-** Exercises like rolling, supine to sit, shifting in the bed forward and backward, quadruped and kneeling positions should be given.
- **Transfers-** Training of same level, high level and low level transfer is important to make the patient functionally independent and improve quality of life.
- **Balance training-** Due to muscular imbalances or loss of control below the level of lesion, sitting and standing balance is poorly affected. Strategies to improve balance in sitting and standing should be given
- **Gait training-** Ambulation training depends on the level of lesion. Higher level lesion can be trained in parallel bars with use of external appliances. Lower level lesion can be trained in walker with use of external appliances and can be progressed to crutches, cane and in the later stages patients should be trained for unsupported walking.
- **Wheelchair training-** Mobility is very important for our daily living. Non-ambulatory patients should be trained for wheel chair

transfers and wheelchair mobility.

## **Occupational Therapy**

With the help of occupational therapy, children with SCI can become more independent in their daily living activities.

Some of the modifications advised for these children are as follow:

**Eating:** Wheelchairs with lap board for eating (especially for children with trunk muscle weakness)

**Bating:** Use of Long handle scrubber for bathing

**Toileting:** Use of inspection mirror while doing toileting activities

**Dressing:** Dressing stick

Using easy to wear shirt and pants, using a loose T shirt, Pants with elastic

**Transferring:** Use of transfer boards while transferring from bed to wheelchair, From wheelchair to classroom bench.

**Mobility:** Use of mobility aid like, crutch, cane, walker as per child's need.

**Home and Classroom modifications:** As per use of the wheelchair. Classroom should be accessible for wheelchair

Lap board can be used as a work surface in the school.

As these patients, have affected bowel and bladder functions, as they have to do procedures like changing the diapers and catheters; Position of the child in the classroom should be near to the washroom. Teachers should be aware of these problems.

## **Psychological Intervention:**

Physical disabilities can be seen as cases of medical complications along with psycho-social problems as severe changes emerge in an individual's life post disability. One of the most prevalent disabilities is Spinal Cord injury. Since many individuals acquire a Spinal cord injury, it is important to look into medical as well as psychological implications in the treatment of this condition.

A Psychologist working in the field of physical rehabilitation can contribute to these conditions by looking into the remediation of behavioral issues like depression, substance abuse, pain or grief that occurs in co morbidity with the physical disability. Physical disability like Spinal Cord Injury may stem from the impairment of physical functioning but the way individuals react to the impairment in relation to the perceived social and environmental context that they are placed in decides the degree of disability attached to the injury. A psychologist can, thus help the individual to function with the new identity that the person has to develop to adapt to the changes post SCI by using client centered therapy. This would help the individual to positively accept self and others irrespective of the changed circumstances. This acceptance and adaptation to the new role will help the individual to alleviate his/her self esteem and confidence. A psychologist can thus help the individual to alter his perception towards the injury, further leading to the development of effective coping mechanisms. Research has shown that an individual's coping with the physical impairment as well as the psycho-social factors, enables the individual to garner to opportunities that help to reduce the extent to which physical injury results in disability.

A psychologist plays a major role in helping the individual to deal with Post Traumatic Stress Disorder that would otherwise keep the individual stuck in the time when the injury was acquired. An individual who has restricted physical abilities and is dependent for ADLs post injury might isolate, hesitate, and cut away from the society, it is here when the psychologist can intervene and encourage the client to be a part of the society leading to his/her well-being. A Psycho-social intervention is important in physical injury cases because of the mind body connection that exists. For example: A dancer who has acquired a SCI might not only lose his/her physical functioning but her personality will also be affected (i.e. she might start feeling inferior). Thus, it is important to treat an individual in a comprehensive set up. Physical injuries can also lead an individual into developing a negative body image and negative self image. These individuals can be taught to alter these images using Cognitive-behavioral therapy and Rational Emotive Behavioral therapy.

Care giving responsibility is another important aspect that comes with SCI. Familial support is very important and on the other hand acceptance

of the provided assistance is also important. The Psychologist can facilitate this process by engaging in family counseling. Physical Disability triggers a chain of psychological reactions that correspond to eight phases of responses to physical disability, Phases 1 to 6 include shock, anxiety, denial, depression, internalized anger, externalized anger and the last two phases are the adaptation phases which would help the client to understand the meaning and purpose of his life irrespective of the physical disability. The psychologist helps the client to follow through these stages and accept self in the changed environment (Livneh and Antonak, 2005).

### **Nutrition**

Good Nutrition will help decrease possible complication like Bowel issue, urinary tract infection, and pressure wounds in patients with spinal cord injury.

Eating balance meal will help maintain patient their weight and will also protects skin.

- Including small and frequent meals help maintain weight and also good for digestion.
- High fiber diet like salads green leafy vegetables will help regulate bowel movements.
- Including good liquid intake will also help clear constipation, limits chances of kidney or bladder stones.
- Drinking 2 to 2.5 liter water in a day help keep your skin hydrated, which help prevent skin tear.
- For overweight and underweight patients their skin is at higher risk of pressure wounds therefore it is very important to maintain healthy weight
- It is important to maintain proper bone health for patients with SCI. Including Vitamin D foods like cod liver oil ,fatty fish like salmon, tuna, mackerel, low fat milk products, egg yolk is important with exposure to morning sunlight
- Including calcium rich foods like low fat milk and milk products,

fortified grains, beans like rajma, sesame seeds, ragi etc.. Is important.

### **Speech therapy**

Few children SCI may have difficulty in speech and swallowing. They may suffer from a motor speech disorder that makes it difficult for them to speak clearly due to poor muscle tone and lack of in coordination in oral facial, neck and throat muscles. The role of a speech therapist is to help these children speak clearly, communicate effectively, and control the muscles involved in speaking, eating, drinking, and swallowing. Speech therapy can improve oral-facial muscle tone and coordination of articulators for improving speech intelligibility/ clarity as well as improving oral sensory awareness, reduce drooling and solve the problem of tongue thrust.

### **Stem Cell Therapy**

The available treatments for SCI fail to repair the underlying pathology completely, leaving behind some neurological deficits. Presently, all modalities aim at repairing the vertebral column but no surgery or medication repairs the spinal cord. None of the treatments help in neuronal or axonal regeneration. Due to loss of functions, the SCI patients have a high level of dependency on the care taker. Rehabilitation and assistive devices are used to improve functions like ambulation and hand functions but while functional improvements are seen in the patients, residual disability always remains. Affected sensations, loss of bladder bowel control and altered muscle tone, are some of the major complications observed in SCI. The currently available treatment modalities fail to improve these complications in cases of severe injuries.

Stem cell therapy is a potential treatment for spinal cord injury. It mainly focuses on replacing the lost or damaged cells and promoting axonal growth and remyelination of axons. The cells migrate to the site of injury and initiate the repair process. They release trophic factors to stop neuronal degeneration and stimulate angiogenesis. These factors also activate the quiescent cells and recruit them to the injured site. Experimental models have demonstrated the formation of functional neuronal circuits promoting functional recovery.

# **Section C: Stem Cell Therapy**





## Chapter 15

# What is Stem Cell Therapy?

*"An idea whose time has come"*

Stem cell therapy, a revolutionary concept in the field of medicine. The highest award for inventions in science the 'Nobel Prize' has been awarded for stem cell research multiple times. In **1990 to Dr. E Thomas received a Nobel Prize for 'Demonstration of stem cells in bone marrow'; in 2007 to Sir Martin Evans for "Isolation of embryonic stem cell in mice' and most recently in 2012 to Dr. John B. Gurdon and Shinya Yamanaka, for 'Reprogramming mature specialized cells into immature cells capable of developing into all tissues of the body'.**

This chapter endeavors to capture the essence of what stem cells are, how can they be of use in neurological and neuromuscular disorders.

Stem cells are cells that help to repair, regenerate and replace damaged cells. Stem cell therapy (also called cell therapy / regenerative medicine) works on the principal of using healthy cells to repair damaged cells, thereby bringing about a biological healing.

### **What are different type of Stem Cells?**

Based on whom the cells are procured from they are classified as:

*Autologous cells* - If cells procured from recipients own body then they are categorized as **autologous** cells

*Allogenic cells* - If stem cells are procured from a different host they are categorized as **allogenic** cells

Based on their ability to differentiate they are categorized as

*Totipotent cells* – The cells that are capable for differentiating to form any tissue including embryonic tissue, e.g. cells of the morula.

*Pluripotent stem cells* - The cells that are can also differentiate to form any cell except the embryonic tissue and placenta. Recently some of the multipotent cells have been genetically reprogrammed to achieve pluripotency. These cells are known as **induced pluripotentstem cells** (i-PSCs).

*Multipotent stem cells* - The cells that are can produce cells from more than one lineage but have limited differentiation capability.

*Unipotent stem cells* - The cells that are highly specialized stem cells that are committed to a single lineage and can only differentiate into one cell type.

Based on the type of tissue that the cells are procured from they are classified as

*Embryonic stem cells* – These cells procured form human embryo i.e. cluster of cells that develops into entire human body. The safety of these cells has not been studied extensively, there are several ethical issues for the use of these cells and these cells may cause tumors.

*Umbilical cord stem cells* – These are the cells that are taken from the placental tissue.

*Adult stem cells* - Adult stem cells are the stem cells that are present throughout the human body and can be procured from various sources such as bone marrow, adipose tissue, dental pulp and peripheral blood. These cells are safe to use and don't have any side effects.

### How can stem cells be transplanted?

Different routes of administration

Stem cell transplantation has been explored through various routes of administration, Most commonly used routes in neuromuscular disorders

are Systemic i.e. Intra-venous and Intra- arterial, Intracerebral, Intrathecal and Intramuscular.

### 1. *Systemic*

Cells are administered through systemic routes i.e intravenous and intraarterial. However, there are several disadvantages of using these routes. The cells administered are diluted and therefore large numbers of cells need to be administered for a positive outcome. The cells are entrapped in liver, lungs and spleen and therefore a very small fraction of injected cells has a potential to reach the target organ.

### 2. *Intrathecal*

Cells are administered in the cerebrospinal fluid through lumbar puncture procedure. This approach is simple, minimally invasive, cells are not diluted in other organs and it allows focused delivery.

### 3. *Intramuscular*

The cells can be injected in several points in the muscle alternatively they can be injected in the motor point of the muscle.

#### Motor point

A motor point is the point at which the motor branch of the innervating nerve enters the muscle (Figure 5.1). It is the point with the highest concentration of motor endplates and myoneural synapses.

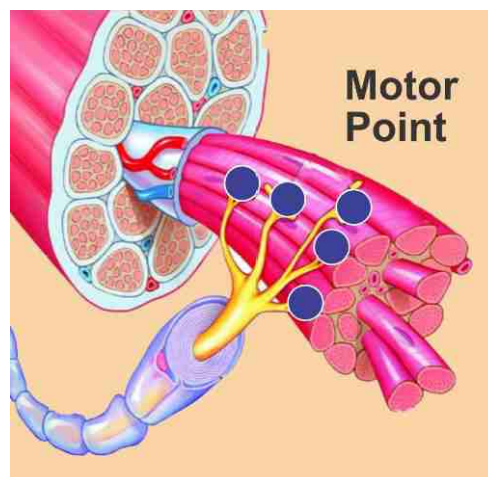


Figure 5.1 Motor point

### **How stem cells repair the damaged tissue?**

- a) By Releasing positive chemicals known as nerve growth factors.
- b) By Improving the blood supply to the damaged parts by a process called angiogenesis (formation of new blood vessels)
- c) By multiplying and differentiating into nerve cells and muscle cells.
- d) By reducing inflammation
- e) By modulating the immune system

At NeuroGen Brain and Spine Institute we use adult stem cells, procured from patients own bone marrow (autologous) and are transplanted intrathecally and intramuscularly depending upon the diagnosis of the patient.

## Chapter 16

# Adult Stem Cell Therapy & Results

### How is stem cell therapy done?

This is a one day procedure done in 3 simple steps with only 2 injections. There is no surgery or stitches.



### **Step1: Bone marrow 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. The bone marrow is aspirated from inside the bone which takes about 20 minutes.



*Stem Cell Separation*

**Step 2: Stem cell separation: (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.



*Stem Cell Intrathecal Injection*



*Stem Cell Intramuscular Injection*

**Step 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. For patients with neuromuscular disorders like muscular dystrophy cells are injected intramuscularly as well. By dividing the procured cells into multiple smaller portions for intramuscular injections at the motor points of the muscles.

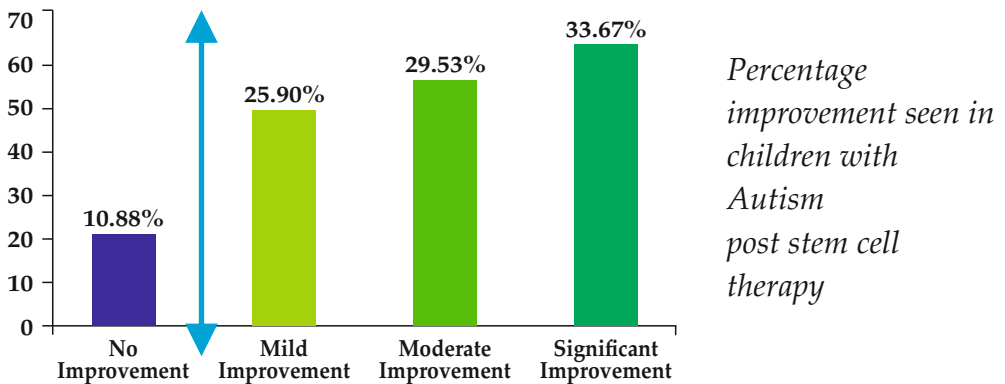
### How safe is stem cell therapy?

When the type of stem cell therapy done is the autologous type (cells taken from/injected into the same patient) then there is no risk of any major irreversible side effects or complications. There is no possibility of the patients becoming worse in anyway after the treatment. At NeuroGen BSI we only use this safest form of autologous bone marrow derived stem cell therapy.

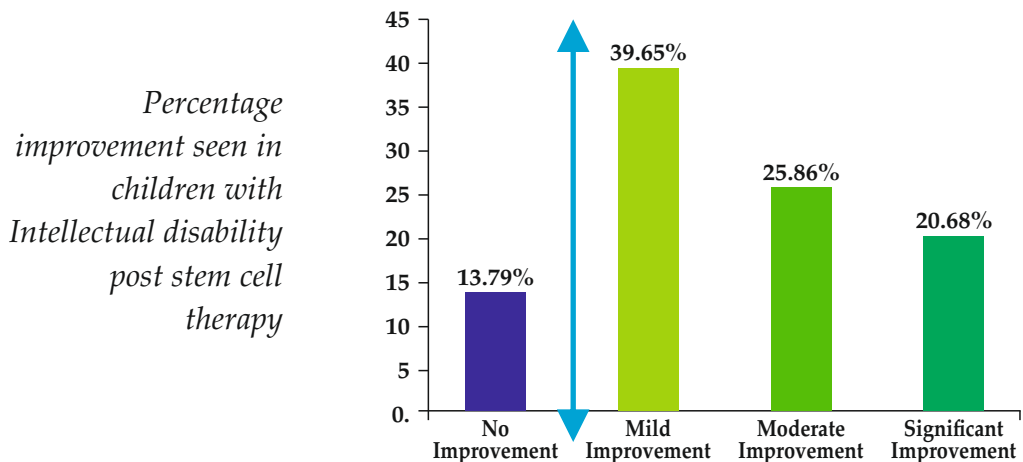
### What is the success rate of stem cell therapy in different neurological disorders?

At NeuroGen BSI the success rate of stem cell therapy in autism is 90% in intellectual disability it is 84% in cerebral palsy it is 91% and muscular dystrophy it is 85%.

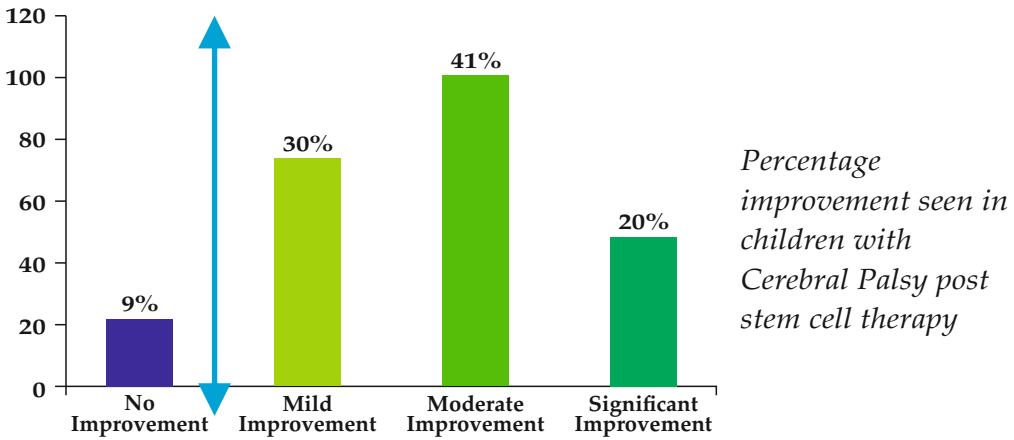
#### Improvements in Autism After Stem Cell Therapy (90% success rate)



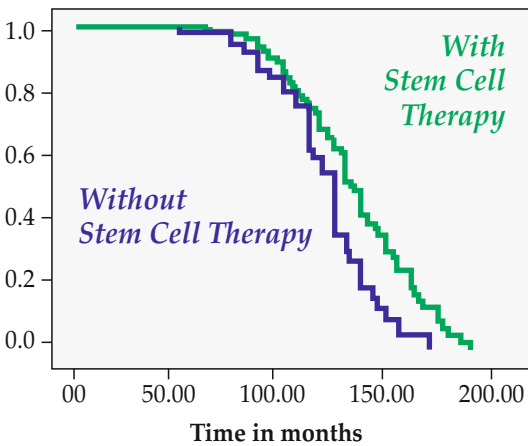
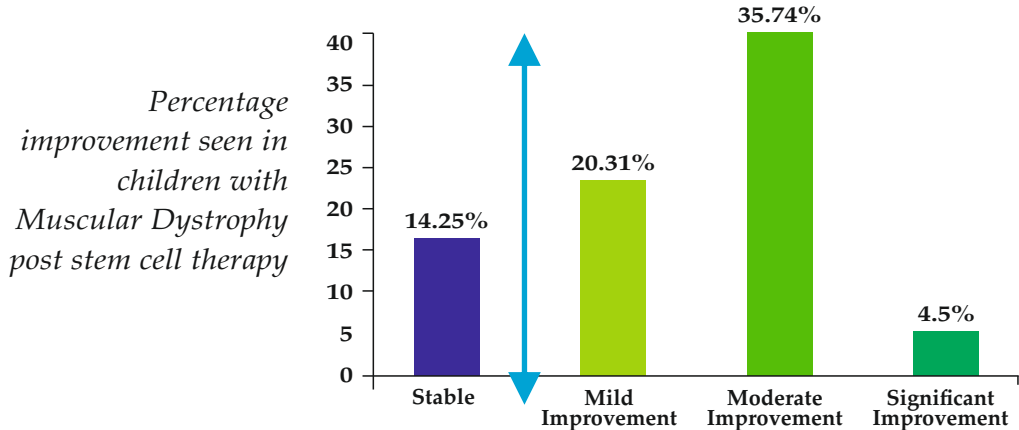
#### Improvements in Intellectual Disability After Stem Cell Therapy (84% success rate)



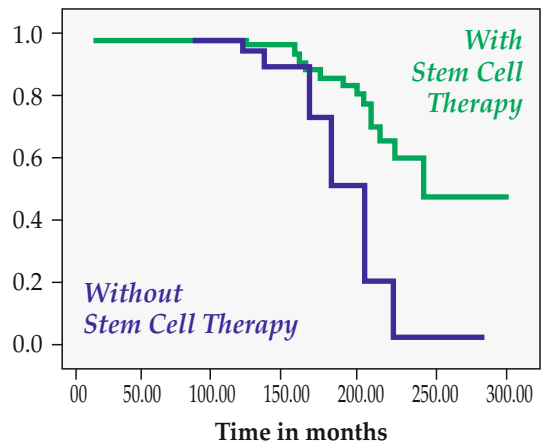
**Improvements in Cerebral Palsy After Stem Cell Therapy (91% success rate)**



**Improvements in Muscular Dystrophy After Stem Cell Therapy (85% success rate)**



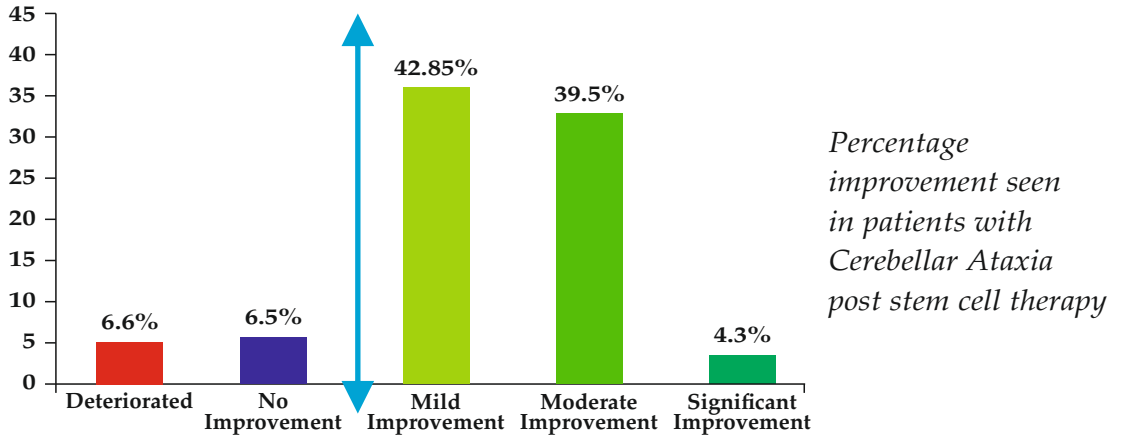
Children with Muscular Dystrophy who underwent stem cell therapy could walk 13 months more than children who did not take stem cell therapy



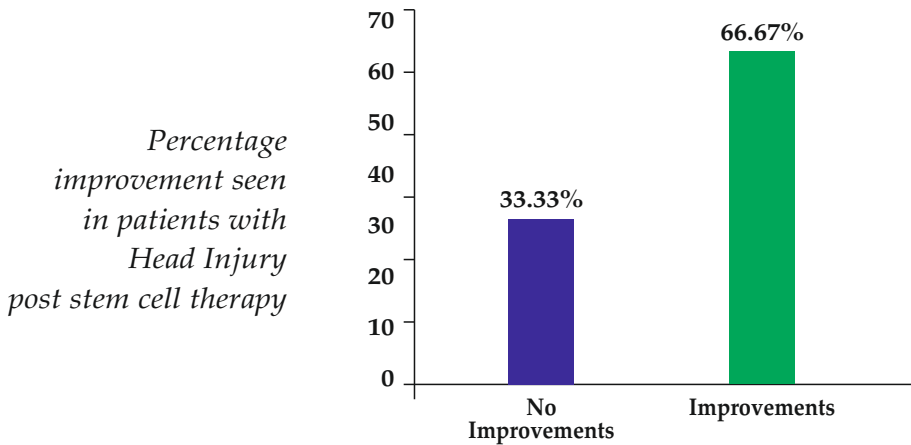
Children with Muscular Dystrophy who underwent stem cell therapy lived 3 years longer than children who did not take stem cell therapy.



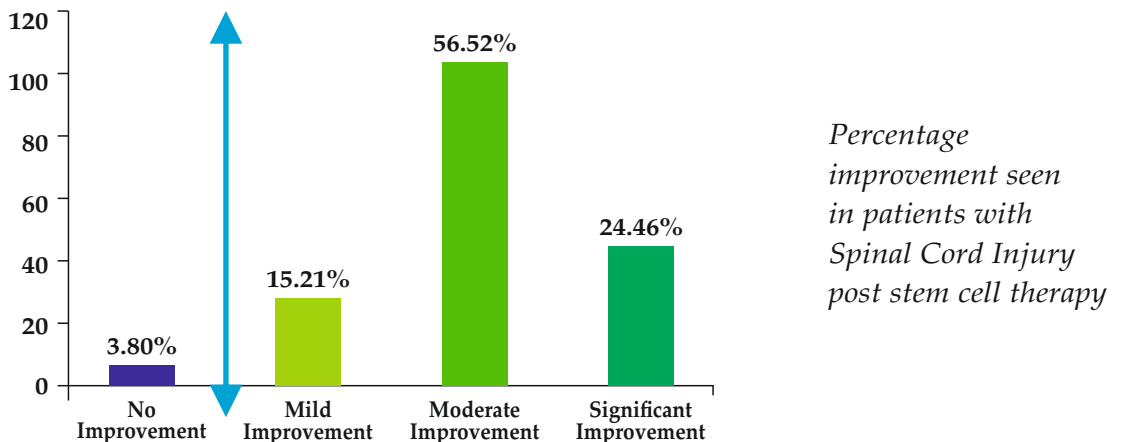
**Improvements in Cerebellar Ataxia After Stem Cell Therapy (87% success rate)**



**Improvements in Head Injury After Stem Cell Therapy (67% success rate)**



**Improvements in Spinal Cord Injury After Stem Cell Therapy (96% success rate)**



### **What is the scientific evidence that stem cell therapy works in neurological disorders?**

Our clinical results showing safety and efficacy of stem cell therapy in autism, cerebral palsy, intellectual disability and muscular dystrophy have been documented in scientific papers published in international and national medical journals. The full text of these articles can be reviewed at [www.stemcellspublications.com](http://www.stemcellspublications.com). The list of these papers is given at the end of the chapter

### **How many patients of have been treated at NeuroGen BSI?**

We have treated over 6000 patients from 60 different countries. Out of these there are over 800 patients of Autism, over 300 patients of Intellectual Disability, over 700 patients of Cerebral palsy, over 1200 patients of Muscular Dystrophy, over 130 patients of Ataxia, over 75 patients of Head Injury and over 500 patients of Spinal Cord Injury.

### **What is the assurance of quality of Stem Cell Therapy services at NeuroGen BSI?**

- a) NeuroGen BSI is ISO 9001:2008 certified
- b) The Stem Cell laboratory at NeuroGen BSI has both the following certifications:
  - (i) GLP (good laboratory practice)
  - (ii) GMP (good manufacturing practice).

### **How should I choose a good center for stem cell therapy for my child?**

The following criteria should be kept in mind:

- Choose a center that uses safe forms of cells like adult stem cells and autologous stem cells.
- Choose a center that has successfully treated patients before without any major or irreversible adverse events.
- Choose a center that has published clinical results in peer reviewed scientific journal in their country and internationally.

**Section D:**  
**Legal rights for the  
people with disabilities**



## Chapter 17

# Legal rights of Persons with Disability

India is one of the largest democracies of the world and equality before law is firmly put forth by the Constitution of India in its various articles. The law professes through all these articles that every person irrespective of age, caste, physical status, race etc. has equal rights in every known form as codified in the law and in every unknown form to be uniformly accepted as told.

The Constitution of India came into force on the 26th of January 1950. It declares through the Chapters on Fundamental Rights and Directive Principles of State Policy, the rights for all persons the essence of which is Equality. The United Nations has also passed a Bill – Universal Declaration of Human Rights, 1948 pertaining to equality and equal treatment of all persons. This bill describes the rights of all persons including those with disability and the laws for the same.

This Chapter aims to put light on the aspects in relation to equality and equal rights in India for differently abled children with physical and cognitive impairment.

The Government of India during its various parliamentary sessions has evolved the following laws and statutory bodies in all areas of concern in relation to differently abled persons –

- Rights of persons with disabilities, 2016
- The National Trust for Welfare of Persons with Autism, Cerebral Palsy, Mental Retardation, and Multiple Disabilities Act, 1999
- The Person with Disabilities Act, 1995
- The Rehabilitation Council of India, 1992
- The Mental Health Act, 1987
- Declaration on The Rights of Mentally Retarded Persons, 1971
- Declaration on the Rights of The Disabled Persons, 1975

### Constitutional rights of persons with disabilities

The Constitution of India has firmly laid and guaranteed to protect the rights of disabled people. The Preamble of the Constitution which is the foundation stone of all laws of India embodies the concept of equality of status and opportunity to all the people of India irrespective of diversities in any form.

Article 14 & 16 of the Constitution guarantee equality of opportunity to every citizen of India. Article 38 of the Constitution requires the State to promote the welfare of the people by securing a social order in which the State is required to make efforts to eliminate inequalities in status, facilities and opportunities amongst individuals.

Constitutionally disabled/differently abled citizens have the same rights as other citizens to a decent standard of living, right to access, communication and economic security through right to work, education and employment.

Some of the specific mentions pertaining to the fundamental rights as mentioned in the Constitution of India are as below-

1. The Constitution secures to the citizens including the disabled, a right of justice, liberty of thought, expression, belief, faith and worship, equality of status and of opportunity and for the promotion of fraternity.

2. Article 15 (1) enjoins the Government not to discriminate against any citizen of India on the ground of religion, race, caste, sex, physical abilities or place of birth.
3. Article 15 (2) States that no citizen (including the disabled) shall be subjected to any disability, liability, restriction or condition on any of the above grounds in the matter of their access to shops, public restaurants, hotels and places of public entertainment or in the use of wells, tanks, bathing ghats, roads and places of public resort maintained wholly or partly out of government funds or dedicated to the use of the general public.
4. There shall be equality of opportunity for all citizens (including the disabled) in matters relating to employment or appointment to any office under the State.
5. Every person including the disabled has his life and liberty guaranteed under Article 21 of the Constitution.
6. Article 25 guarantees to every citizen (including the disabled) the right to freedom of religion. Every disabled person (like the non-disabled) has the freedom of conscience to practice and propagate his religion subject to proper order, morality and health.
7. No Disabled person will be deprived of the right to the language, script or culture which he has or to which he belongs.
8. Every disabled person can move the Supreme Court of India to enforce his fundamental rights and the rights to move the Supreme Court is itself guaranteed by Article 32.
9. No disabled person owning property (like the non-disabled) can be deprived of his property except by authority of law; though right to property is not a fundamental right. Any unauthorized deprivation of property can be challenged in the court.
10. Every disabled person (like the non-disabled) on attainment of 18 years of age becomes eligible for inclusion of his name in the general electoral roll for the territorial constituency to which he belongs.

## **Education Law for the Disabled**

### ***Constitutional rights***

1. The right to education is available to all citizens including the disabled. Article 29(2) of the Constitution provides that no citizen shall be denied admission into any educational institution maintained by the State or receiving aid out of State funds on the ground of religion, race, caste or language.
2. Article 45 of the Constitution directs the State to provide free and compulsory education for all children (including the disabled) until they attain the age of 14 years. No child can be denied admission into any education institution maintained by the State or receiving aid out of State funds on the ground of religion, race, caste or language.

### ***The Persons with Disabilities Act of 1995***

The Persons with Disabilities Act of 1995 imbibes and professes the following rights for education. It is a significant step which ensures equal opportunities for the people with disabilities and their full participation in the nation building. The Act defines the responsibilities of the Central and State government with regards to the services for disabled persons. Provisions have been made in this Act for the protection of rights, provision of medical care, education, training, employment and rehabilitation of disabled persons. The Act also recommends to create a barrier free environment by removing all type of discrimination against persons with disabilities where they can share the development benefits which a normal person enjoys.

1. Every Child with disability shall have the rights to free education till the age of 18 years in all integrated schools or special schools.
2. Appropriate transportation, removal of architectural barriers and restructuring of modifications in the examination system shall be ensured for the benefit of children with disabilities.
3. Special Schools for children with disabilities shall be equipped with vocational training facilities.



4. Non-formal education shall be promoted for children with disabilities.
5. Teachers' Training Institutions shall be established to develop requisite manpower.
6. Parents may move to an appropriate forum for the redressal of grievances regarding the placement of their children with disabilities.

### **Other rights for people with disabilities**

1. 3% of vacancies in government employment shall be reserved for people with disabilities
2. Health and Safety measures and creation of a non- handicapping, environment in places where persons with disabilities are employed
3. Government Educational Institutes and other Educational Institutes receiving grant from Government shall reserve at least 3% seats for people with disabilities.
4. No employee can be sacked or demoted if they become disabled during service, although they can be moved to another post with the same pay and condition. No promotion can be denied because of impairment.
5. A specific act has been enacted namely- The national trust for welfare of persons with autism, cerebral palsy, mental retardation and multiple disabilities act, 1999 to provide security in every form for persons covered under this act. The Act gives direction for the care and protection of persons with these disabilities in the event of death of their parents, procedures of appointment of guardians and trustees for persons in need of such protection and to provide need-based services in times of crisis to the families of the disabled.
6. The Central Government has the obligation to set up, in accordance with this Act and for the purpose of the benefit of the disabled, the National Trust for Welfare of Persons with Autism, Cerebral Palsy, Mental Retardation and Multiple Disability at New Delhi.

7. The National Trust created by the Central Government has to ensure that the objects for which it has been set up as enshrined in Section 10 of this Act have to be fulfilled.
8. It is an obligation on part of the Board of Trustees of the National Trust to make arrangements for an adequate standard of living of any beneficiary named in any request received by it, and to provide financial assistance to the registered organizations for carrying out any approved program for the benefit of disabled.
9. Disabled persons have the right to be placed under guardianship appointed by the 'Local Level Committees' in accordance with the provisions of the Act. The guardians so appointed will have the obligation to be responsible for the disabled person and their property and required to be accountable for the same.
10. A disabled person has the right to have his guardian removed under certain conditions. These include an abuse or neglect of the disabled, or neglect or misappropriation of the property under care.
11. Whenever the Board of Trustees are unable to perform or have persistently made default in their performance of duties, a registered organization for the disabled can complain to the central government to have the Board of Trustees superseded and/or reconstituted.
12. The National Trust shall be bound by the provisions of this Act regarding its accountability, monitoring finance, accounts and audit.

**A derivation of the Rights of Persons with Disabilities Bill - 2016**  
**Passed by the Parliament in December 2016**

1. Responsibility has been cast upon the appropriate governments to take effective measures to ensure that the persons with disabilities enjoy their rights equally with others.
2. Additional benefits such as reservation in higher education, government jobs, reservation in allocation of land, poverty alleviation schemes etc. have been provided for persons with benchmark disabilities and those with high support needs. (Benchmark disabilities are where the percentage of disability exceeds 40%)
3. Every child with benchmark disability between the age group of 6 and 18 years shall have the right to free education (Benchmark disabilities are where the percentage of disability exceeds 40%)
4. Government funded educational institutions as well as the government recognized institutions will have to provide inclusive education to the children with disabilities.
5. Office of Chief Commissioner of Persons with Disabilities has been strengthened who will now be assisted by 2 Commissioners and an Advisory Committee comprising of not more than 11 members drawn from experts in various disabilities.
6. Similarly, the office of State Commissioners of Disabilities has been strengthened who will be assisted by an Advisory Committee comprising of not more than 5 members drawn from experts in various disabilities.
7. The Chief Commissioner for Persons with Disabilities and the State Commissioners will act as regulatory bodies and Grievance Redressal agencies and also monitor implementation of the Act.
8. National and State Fund will be created to provide financial support to the persons with disabilities. The existing National Fund for Persons with Disabilities and the Trust Fund for Empowerment of Persons with Disabilities will be subsumed with the National Fund.

9. The Bill provides for penalties for offences committed against persons with disabilities and also violation of the provisions of the new law.
10. Special Courts will be designated in each district to handle cases concerning violation of rights of Persons with Disabilities

### **Where to register a complaint?**

In case of discrimination or if any of the above mentioned rights are declined then you can complain to the office of state commissioner for persons with disabilities.

#### **State of Maharashtra**

**Commissioner for persons with disabilities** - Shri Nitin Patil

**Office address** - Commissioner, disabilities, maharashtra state, govt. of maharashtra, 3, church road. Pune - 411 001

**Contact no.** - (020)26122061, (020) 26111590

**Fax no.** – (020) 26126698

### **How to register a complaint?**

The complaint should be filed along with disability certificate and supporting documents. It can be sent by post or by hand with following details.

1. *The name, description and the address of the complainant;*
2. *The name, description and the address of the opposite party or parties, as the case may be, so far as they can be ascertained;*
3. *The facts relating to complaint and when and where it arose;*
4. *Documents in support of the allegations contained in the complaint;*
5. *The relief that the complainant claims*

### **Procedure once the complaint is filed:**

- The chief commissioner on receipt of a complaint shall refer a copy of the complaint to the opposite party/parties mentioned in the complaint directing him to give his version of the case within a period of thirty days or such extended period not exceeding fifteen days as may be granted by the chief commissioner.

- On the date of hearing or any other date to which hearing could be adjourned, it shall be obligatory on the parties or their agents to appear before the chief commissioner.
- Where the complainant or his agent fails to appear before the chief commissioner on such days, the chief commissioner may in his discretion either dismiss the complaint on default or decide it on merits.
- Where the opposite party or his agent fails to appear on the date of hearing the chief commissioner may take such necessary action under section 63 of the act as he deems fit for summoning and enforcing the attendance of the opposite party.
- The chief commissioner may dispose of the complaint ex parte, if necessary.
- The chief commissioner may on such terms as he deems fit and at any stage of the proceedings, adjourn the hearing of the complaint.
- The complaint shall be decided, as far as possible, within a period of three months from the date of notice received by the opposite party.
- In cases of complaint regarding discrimination relating to central government, a complaint can be registered directly with the Chief commissioner for persons with disabilities

**Chief commissioner for persons with disabilities - Dr. Kamlesh Kumar Pandey**

**Office address** - Sarojini House, 6 Bhagwan Dass Road,  
New Delhi 110001

**Contact no.** - 91-011 - 23386154, 23386054

**Email:** ccpd@nic.in

**Fax:** 91-011-23386006

## **Maharashtra State Disability Pension scheme (Niradhar Anudan Yojna)**

The scheme provides financial assistance. Each applicant will get Rs. 600/- per month and Family with more than one beneficiary will get Rs .900/- per month.

### ***Eligibility–***

1. Destitute persons, blind, disabled, orphan children, persons suffering from major illnesses, divorced women, abandoned women, women freed from prostitution, outraged women, Transgender, etc.
2. Minimum 40% of disability
3. Below age of 65 years
4. Annual family income upto Rs 21,000/-
5. Resident of the state

### ***Documents Required -***

Certificate of Residence

Certificate of Age

Certificate of Income /Proof of Below Poverty Line family

Certificate of Incapacity /Disease - issued by the Civil Surgeon and Medical Superintendent of the Government Hospital.

### ***Procedure for application -***

Visit Talathi /Tahsildar of the respective village

Contact the district social welfare officer of respective district

### ***Mode of payment -***

The amount will be credited to the saving bank account or account in post office of the applicant.

### **Special schemes run by the state government of Maharashtra for the persons with disability**

1. Special Education and Vocational Training through Government institutions
2. Non-Governmental Aided organization (Special Schools and Vocational Training Centre) for Disable.
3. Homes for Intellectually Impaired Persons
4. State Post-metric Scholarship for Disabled
5. State Pre-metric Scholarship for Disabled
6. Merit Awards
7. Financial Assistance to disabled for self-employment
8. Financial Assistance to Self-Employment for Trained Disabled Persons
9. Financial Assistance for Aids and Appliances for Disabled Persons
10. The Scheme of State Award for Disabled
11. Matrimonial Incentives

### **International Initiatives**

All human rights instruments affirm fundamental and inalienable rights to all persons who are physically disabled. Article 1 of the Universal Declaration of Human Rights, 1948, states that, "all human beings are born free and equal in dignity and rights. They are endowed with reason and conscience and should act towards one another in a spirit of brotherhood".

In 1971 the General assembly adopted the Declaration on the Rights of Mentally Retarded Persons, taking into account the necessity of providing help to mentally retarded persons in order to enable them to develop their abilities and promoting their integration in the normal life. The Declaration recommends a frame work within which national and international actions should initiated for the advancement of rights such as medical care, education, training, rehabilitation, economic security,

right to have qualified guardian, protection from exploitation,

Another declaration was adopted by the general assembly for mentally retarded persons in the year 1975, keeping in view, “the necessity of preventing physical and mental disabilities and of assisting disabled persons to develop their abilities in the most varied fields of life”.

India is an active member of the United Nations Organization and was the prime state to the Proclamation on the Full Participation and Equality of People with Disabilities in the Asian and Pacific region, which was adopted at the meeting to launch the Asian & Pacific Decade of Disabled persons 1993-2002 convened by the Economic and social Commission for Asia and Pacific at Beijing in 1992.



# NeuroGen Brain & Spine Institute

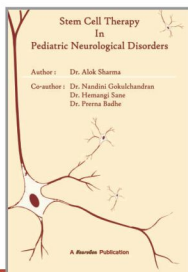


The NeuroGen Brain & Spine Institute is an International center of excellence for Neurological disorders. Founded by Dr. Alok Sharma it is India's First dedicated Hospital for Stem Cell Therapy and Comprehensive Neurorehabilitation. Located adjacent to the Arabian sea on the scenic Palm beach road in Navi Mumbai, this center has a multidisciplinary team of expert and experienced medical professionals that provide holistic care using the latest technological advances in the world. It has treated over 5000 patients from 50 different countries. The care offered here is very professional yet very caring.

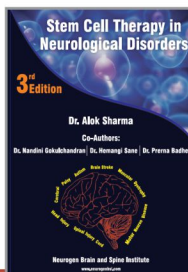
A separate pediatric neurorehabilitation facility and other play areas makes it very child friendly. The institute is very scientific and academic in its approach and to date has published 73 scientific papers in international and national journals. 14 books have also been published and chapters contributed to several international textbooks. NeuroGen also has many international tie ups with leading organizations from America and other countries for research and treatment collaborations. The institute is very quality conscious and has several certifications (1. ISO 9001:2015, 2. GLP & 3. GMP certification). Despite all the international partnerships and treatments offered to patients from all over the world the institute is very socially conscious and through the Stemcare foundation financially supports patients from the lower socioeconomic strata to be able to avail of the treatments that are needed. It is a policy of the institute that no patient should be deprived of any treatment due to financial reasons. NeuroGen doctors conduct free medical camps all over the country. Conferences, workshops and CME's are regularly conducted to impart knowledge to doctors, therapists as well as patient families. Cutting edge research, pioneering new treatments, the best medical professionals, comprehensive treatment facilities all under one roof and a caring holistic approach and make the NeuroGen Brain and Spine institute a unique and special facility for patients with Neurological problems.

# Other books published by NeuroGen Brain and Spine Institute

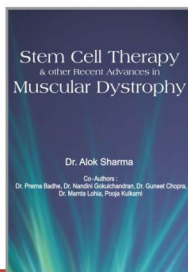
## Stem Cell Therapy In Pediatric Neurological Disorders



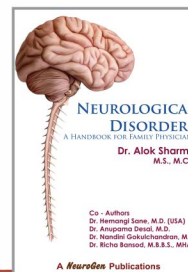
## Stem Cell Therapy in Neurological Disorders 3rd Edition



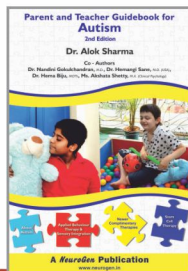
## Stem Cell Therapy & Other Recent Advances in Muscular Dystrophy



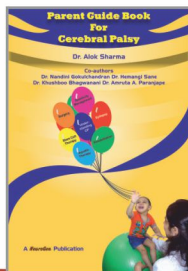
## Neurological Disorders A Handbook for Family Physicians



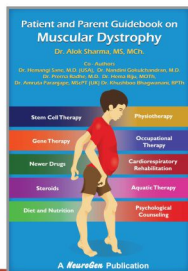
## Parent & Teacher Guide Book for Autism 2nd Edition



## Patient Guide Book for Cerebral Palsy



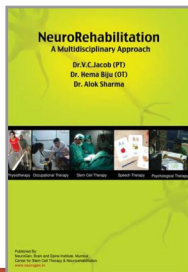
## Patient & Parent Guidebook on Muscular Dystrophy



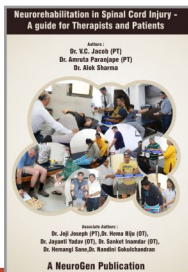
## પેશન્ટ અને પેરન્ટ માર્ગદર્શિકા મસ્ક્યુલર ડિસ્ટ્રોફી વિશે



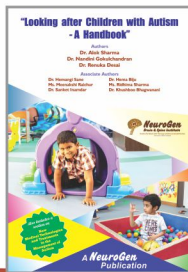
## NeuroRehabilitation- A Multidisciplinary Approach



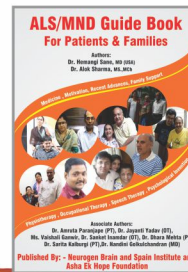
## Neurorehabilitation in Spinal Cord Injury A guide for Therapists and Patients



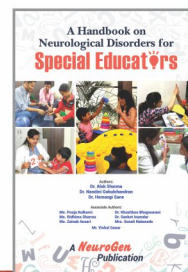
## Looking after children with autism - A handbook



## ALS / MND Guide Book For Patients & Families



## A Handbook on Neurological Disorders for Special Educators



[www.neurogenbsi.com](http://www.neurogenbsi.com)



**NeuroGen**  
Brain & Spine Institute

Centre for Stem Cell Therapy and Neurorehabilitation  
ISO 9001:2015 Certified

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