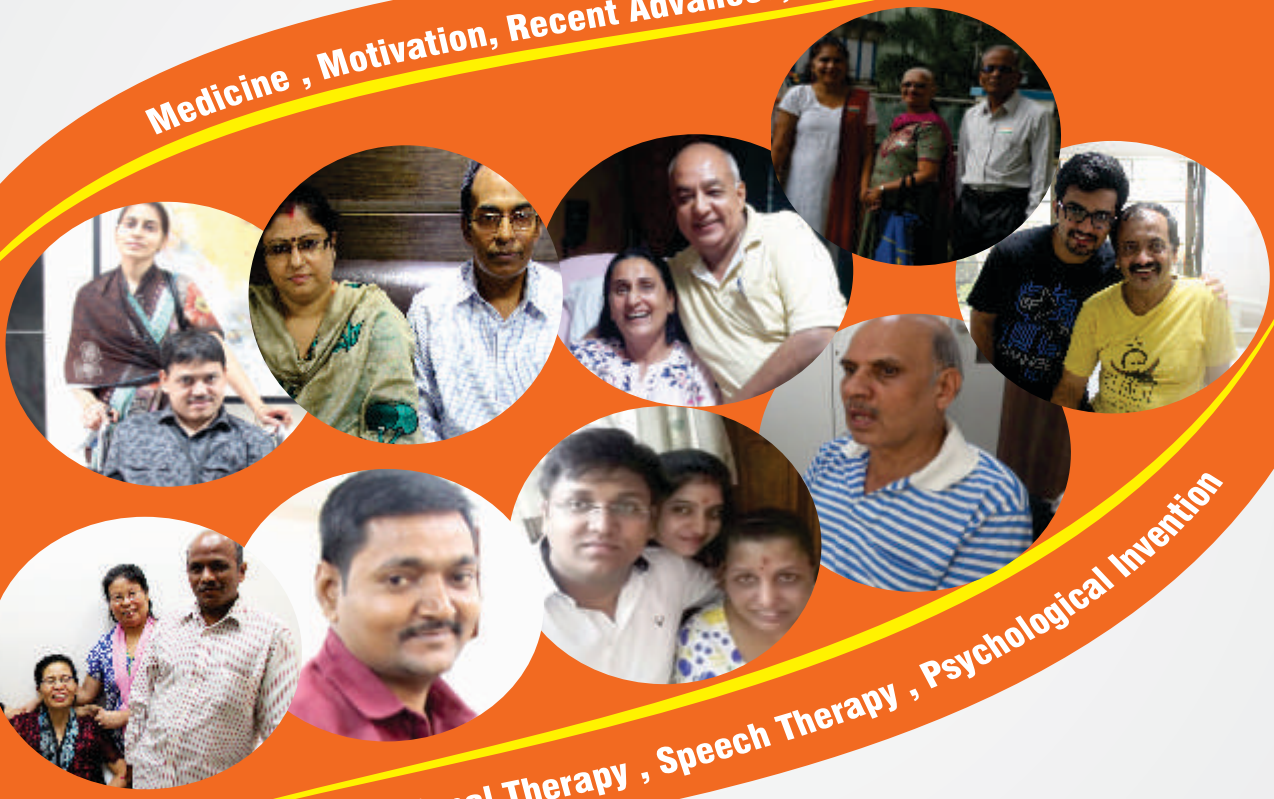
ALS/MND Guide Book

For Patients & Families

Authors:
Dr. Hemangi Sane, MD (USA)
Dr. Alok Sharma, MS.,MCh

Medicine , Motivation, Recent Advances, Family Support



Physiotherapy , Occupational Therapy , Speech Therapy , Psychological Invention

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Asha Ek Hope
Foundation for MND/ALS

Vision- To make ALS/MND a completely reversible and curable condition.

Mission- To build hope and positivity in the lives of people with ALS/MND and their families, helping them to cope with confidence and dignity, supporting them to live a complete life . Also spread awareness about the disease along with supporting research to find cure for ALS/MND .

About Asha Ek Hope foundation

Asha Ek Hope Foundation, a non-profit organization was founded in June 2011 to support patients with ALS/MND. This is the first foundation for MND/ALS in India. This **registered NGO (with Mumbai Charity Commissioner and 80 G Tax exemption**) is founded with the help of family and friends of a doctor, who is living with motor neuron disease since 2004. The foundation is an associate member of the international alliance for ALS/MND. The Alliance is an international forum to share information about different aspects of management of MND/ALS.

The foundation offers various programs like ALS/MND clinic, rehabilitation services, equipment support, Rilutor etc.

Stay involved and make the most of your membership by joining a chapter near you at Mumbai, Bangalore and Kolkata.

We urge you to show your support by lending a helping hand in this humanitarian cause. You can donate funds to help the patients in terms of medication, equipments or fund research to find a cure for ALS/MND . *Come join us and make hope a reality.*

INTERNATIONAL ALLIANCE OF ALS/MND ASSOCIATIONS

ALS/MND Guidebook for Patients and Families

Dedicated to all the brave patients and their courageous caring families

ALS/MND Guidebook for Patients and Families

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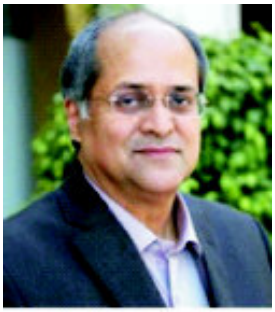
Foreword



My introduction to motor neuron disease was emotionally traumatic one when a very dear friend and colleague, Dr. Sorab Bhabha was diagnosed, suffered, and finally succumbed to the disease. Sorab was associate professor of neurology at Seth G. S. Medical college and KEM Hospital whilst I was doing my neurosurgical residency. The department of neurosurgery and neurology shared the same ward (ward 10). Sorab and I developed a deep friendship as we discussed our clinical work. All of us watched helplessly as the disease slowly crippled and eventually killed Sorab.

Through those years, I wish so often that we had something that could slow down or arrest the progression of MND. It was only several years later that in my quest to find a solution to the devastating neurological consequences of Spinal Cord Injury that I finally discovered a possible solution to combat the devastation caused by MND. I sometimes refer this disease as a "black hole" of neurological diseases and I am convinced that if we do eventually find a definitive solution to motor neuron disease, we will have also found solution to all incurable neurological diseases. We have for the last 7 years been treating patients of MND/ALS with bone marrow derived autologous mononuclear cell therapy also referred to as stem cell therapy. We have compared the outcome of our treatment to the outcome of the patient's that have not taken this treatment and found that we have been able to prolong the survival by about 30 months. This work has been published in the American Journal of stem cells. Work done by Dr. Prabhakar form Chandigrah and published in neurology India also showed similar results.

Publications from Professor Huang and several others have also shown the potential of regenerative medicines in the management of MND. We are still nowhere near a cure for this dreaded disease. But if all through the last century we were in a dark tunnel, now in the last 10 years we have started seeing light at the end of the tunnel. Whilst it is unlikely that any new drug will be discovered for ALS in the next decade or so, it is almost certain that one or the other form of cell therapy will show good results.

Regenerative medicine and stem cell therapy is here to stay. The only thing left to discover is which type of cell therapy will work the best in MND and which would be the best mode of delivering these cells. What we need to do is join hands so that we can discover the best possible way of conquering this disease.

Stem cell therapy is the only half the battle a positive and inspired attitude of the patients and family and a sustained well informed and executed rehabilitation program is equally important. It has been my observation that the 4 pillars to the successful conquest of the disease are:

1. A positively inspired and motivated patient with a determined will to combat disease.
2. Supportive and encouraging family.
3. A regular structured, disciplined, individually designed rehab program, supervised by therapists with an experience in the management of MND.
4. Stem cell therapy along with medical treatment such as Rilutor and Lithium.

This book which is a brain child of Dr. Hemangi Sane, who has seen this disease from all its different perspective including those of a patient, recipient of stem cell therapy, a doctor treating patients of MND, a researcher trying to find solutions for this difficult problem and a founder of non-profit organization to help these patients the most suitable person to write this book.

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Preface

Life was beautiful with good health, a successful career and a happy family. But then my world came crashing down with the diagnosis of ALS/MND in 2004. I had graduated with an MBBS degree from Seth G.S Medical College and then travelled to America for post graduate studies. After acquiring an M.D degree in Internal Medicine from New York Medical College, I was practicing as a physician in New York. Suddenly, I felt weakness in the hands while writing and thought that maybe a nerve is compressed. But investigations revealed that I had developed ALS/MND. With no family history, I did not understand from where this was coming. Being a doctor myself, I knew there was no cause, medicine or cure for this disease. I still remember that day when I burst into tears upon hearing from a neurologist that I had only 3-5 years of life span. I consulted the best of the doctors and hospitals in USA but I didn't get any hope. My weakness kept progressing in the hands, legs with frequent falls, slurred speech, swallowing difficulty and I finally started getting dependant for the activities of daily living. I could no longer continue my job in USA. I returned back to my parents in India.

My life was filled with sadness and no hope. But then, miraculously I met Dr Alok Sharma who offered me hope through stem cell therapy. I saw genuineness in his efforts and concern and so I decided to give it a try. I underwent stem cell therapy in 2009 (autologous bone marrow stem cells) and followed a regular neuro-rehabilitation program. To my surprise, I saw that my disease progression was halting and my speech and stamina were improving. As my condition became stable and improved, I could join my medical profession and started to practice again. I got my life back. I believe that this was possible for me due to strong support from my parents and family. From a life of complete independence to a life of dependence, made me angry and frustrated; but, I realized that it was important to focus on solutions rather than problems. The positive energy of my parents motivated me to keep my will power strong and to face the challenges of life with courage. The positive attitude helped me to take life a day at a time and to live each day to its fullest.

I wish to shine a ray of hope through this guide book and empower the people with ALS/MND. The book is a holistic approach which includes information about the disease, diagnosis and multidisciplinary treatment (medicines, physiotherapy,

occupational therapy, speech therapy, psychological counseling, diet and nutrition management, motivation, assistive devices and advanced care). It takes into account the varied symptoms and stages of the disease which will direct the goals of management. The book can serve as a powerful tool to take best possible care of the patient. The book also compiles the recent advances and ongoing research in the field of ALS/MND. With emerging research in stem cell therapy and gene therapy we are confident that our dream of cure for MND/ALS will soon be a reality.

I have also founded Asha Ek Hope, a non-profitable organisation for ALS / MND patients. Through this organisation, we provide care, guidance and support to patients and their families.

I personally urge all my readers to have a positive attitude, strong will power and self motivation to lead their lives in the best possible way.

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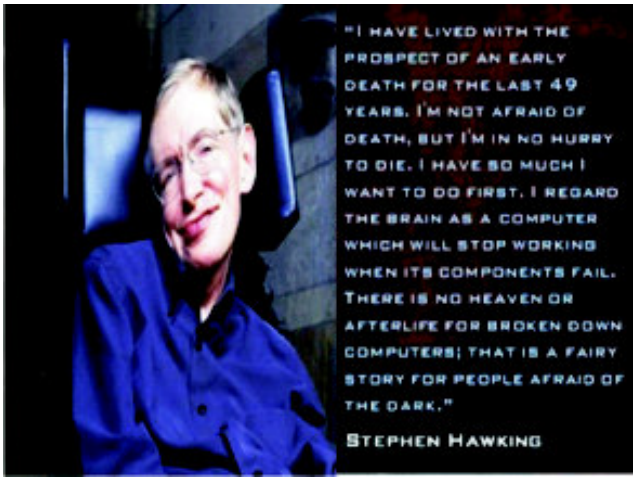
Shining Stars of Motor Neuron Disease

- Success Stories

Motor Neuron Disease is a life - changing condition. Itusually comes with a negative stigma. Because of the progressive nature of the disease, society considers it as an end to the existing life. However, there are some who against all odds have overcome the diverse challenges of life.



Stephen William Hawking is a British theoretical physicist, whose world-renowned scientific career spans over 40 years.One of the most recognizable figures in modern science was diagnosed with ALS, a form of Motor Neuron Disease, shortly after his 21st birthday. Doctors said he would not survive more than two or three years. At the age of 72 years, in spite of being wheelchair bound and dependent on a computerized voice system for communication,he still hopes to make it into space one day.His books and public appearances have made him an academic celebrity and he is an Honorary Fellow of the Royal Society of Arts, a lifetime member of the Pontifical Academy of Sciences, and in 2009 was awarded the Presidential Medal of Freedom, the highest civilian award in the United States. He usually says "However difficult life may seem, there is always something you can do and succeed at! While there is life, there is HOPE!"

Let us shed some light on some of our friends whose spirit and determination;spread joy and hope amongst all.

Dr. Hemangi Sane, working as a DeputyDirector of NeuroGen Brain & Spine Institute and Head of the R&D Department now, Dr. Hemangi Sane was diagnosed to have a Motor Neuron Disease 11 years ago. There was progressive weakness in hands and legs with difficulty in getting in and out of the chair and climbingstairs, severe calf muscle cramps while attempting any activities, balance issues and easy

fatigability. The symptoms worsened and she had to give up her job. Eventually there was difficulty in speaking, swallowing and even coughing.

She underwent Stem Cell Therapy (SCT) 5 years after she was diagnosed with MND. Her condition became more stable and the deterioration halted. Today, she is an inspiration to all of us in the institute. Her zeal for bringing about a change in the patient's life makes all of us work harder.

Besides this, she has been awarded as an "Leading Physician" in the year -2013; the Mayor of Mumbai also felicitated her on International Women's Day. She is the founding president of Asha Ek Hope foundation, the only NGO for MND patients in India. More than 25 published medical research papers and 10 books, she has it all in her kitty!

"So many people walk around with a meaningless life.They seem half asleep, even when they are busy doing things they think are important. This is because they are chasing the wrong things.The way you get meaning into your life is to devote yourself to loving others, devoting yourself to your community around you, and devoting yourself to creating something that gives you purpose and meaning."

- Morrie Schwartz

Purningam Jacob : A nurse by qualification and a housewife at heart, she is a perfect example of an Indian woman working double shift!

She was diagnosed to have Amyotrophic Lateral Sclerosis (ALS) around 3 years ago after the delivery of her 2nd daughter. Weakness in her legs kept on increasing followed by weakness in hands. Eventually her speech and swallowing was also affected.

She underwent SCT, 2 years after the onset of her symptoms. She got a second shot within the next 6 months. Following which not only did the rate of deterioration reduce but also there was tremendous improvement in terms of speech, fine movements of fingers, stamina, ambulation and her overall muscle strength.

Her disciplinary quality of following a rehabilitation routine has made her overcome her difficulties and continue working as a nurse along with taking care of her family.

"She is clothed in STRENGTH And DIGNITY And she LAUGHS Without Fear Of The Future"

Shera Mukherjee : A 40-year-old independent corporate woman was diagnosed to have amyotrophic lateral sclerosis 3 years ago. She complained of weakness in both her lower extremities and eventually in upper extremities. Her symptoms started gradually and progressed to difficulty in ambulation since 2 years and difficulty in speech since 1 year. She came with complains of difficulty in speaking, easy fatigability, and slowness in activities and difficulty in getting up from bed.

After 2 shots of SCT, there was a great improvement in her speech, balance and walking pattern. Her frequency of falls while walking also reduced considerably. She could work for a longer hours because of increase in her stamina. Today, she continues to leave her mark in the corporate world and sets an example to many for her perserverance.

"People are always blaming their circumstances for what they are. I don't believe in circumstances.

The people who get on in this world are the people who get up and look for the circumstances they want, and if they cant find them, they make them."

Mr. Ramesh Yadav : Ramesh Yadav, the most cheerful person one can ever come across, is living with MND since the past 3 years. When he came to us, 2 years ago, his main complains were that of weakness in his upper limbs, which were becoming weaker and weaker with time. After SCT, his deterioration has stopped and he continues to connect people to the real world through his newspapers.

"Strong people always have their life in ORDER. Even with tears in their eyes,

They still manage to say "I am GREAT" with a SMILE!"

CONTENTS

Section D: Recent advances

Section E: Going beyond the limitations of ALS

SECTION A
An Overview

Dr. Jean Marie Charcot

(29 November 1825 - 16 August 1893)

A French neurologist and professor of anatomical pathology known as "the founder of modern neurology", and his name has been associated Charcot-Marie-Tooth disease and Charcot disease (better known as amyotrophic lateral sclerosis, motor neurone disease, or Lou Gehrig disease). Motor neuron disease or ALS was first described by Dr. Jean Marie Charcot, the father of neurology in 1874. MND destroys the motor neurons progressively leaving an active mind in the paralyzed body. The weakness spreads all over the body affecting the walking, hand function, talking, swallowing and ultimately breathing. All this happens rapidly in individuals with ALS the most dreaded type of MND. Before the person can adapt to a weak part of the body another part gets weaker. This progression makes it very difficult for the individual and family to cope with the disease. The varied symptoms and unpredictable course makes it harder. We know this disease over a century but yet we do not know the exact cause or a cure.

Chapter 1

What is Motor Neuron Disease?

Before we get to know what motor neuron disease (MND) is, let us first understand about the body system affected by motor neuron disease. MND is a disease of nerves and thereby affecting the muscles. This chapter will briefly explain the basic structure and function of motor neuron system and how this is altered by MND.

Understanding the nervous system

Any movement in the human body requires synchronized work from musculoskeletal systems (bones and muscles of the body) and nervous system (brain and nerves) in the body. The nervous system is categorized in 3 types of nervous systems (Figure 1.1): Central Nervous System, Peripheral Nervous System, Autonomic Nervous System.

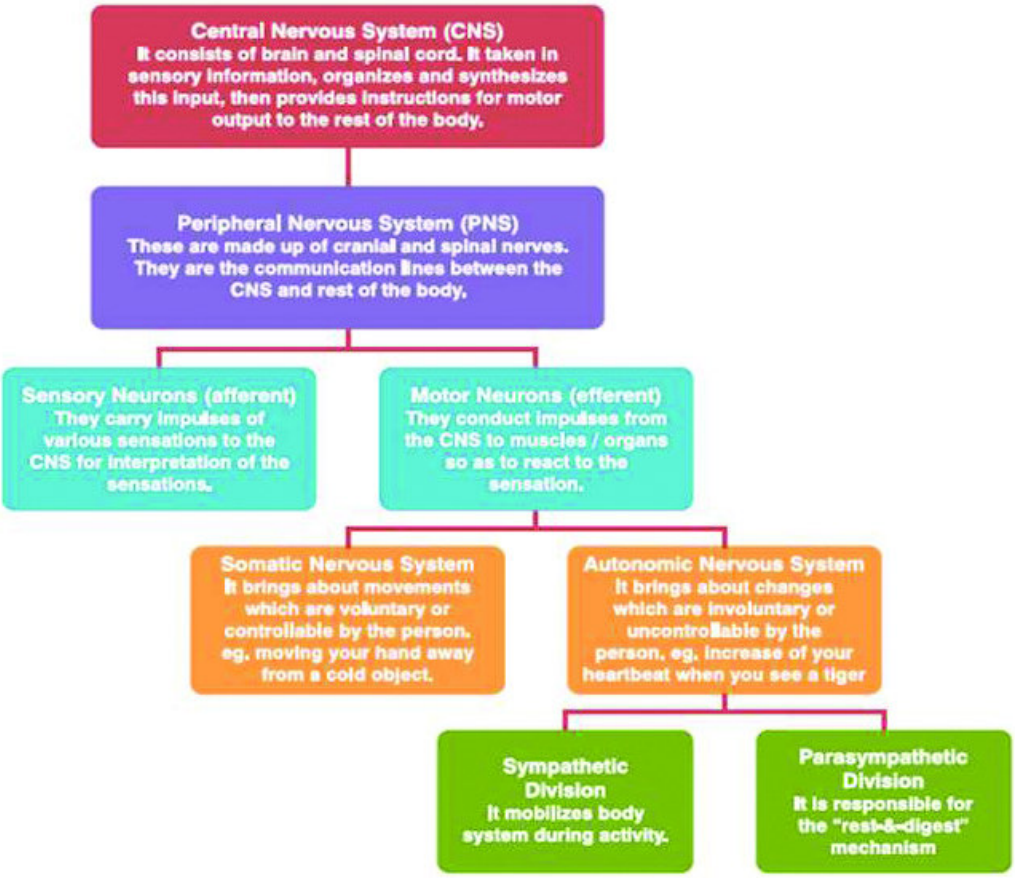


Figure 1.1- Types of Nervous systems

Central nervous system:

Central nervous system is made of brain and spinal cord. There are tracks of fibers that originate from brain and run down across the spine as spinal cord. Central nervous system is the controlling unit of the whole body. This system receives the information from peripheral nervous system and upon analyzing gives commands to bring about required actions.

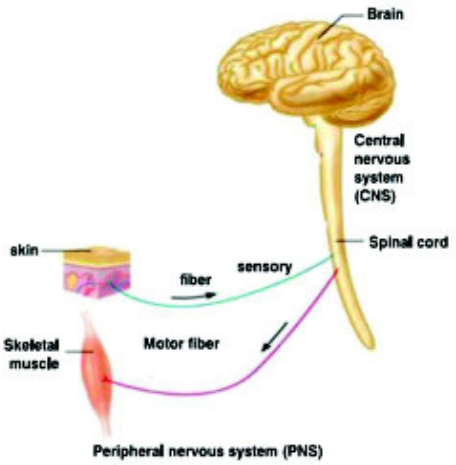


Figure 1.2- Central and peripheral nervous system

Peripheral nervous system:

Peripheral nervous system consists of the nerve fibers that originate from spinal cord and are spread throughout the body to form a web. The function of this system is to receive information from various parts of the body and convey it to the spinal cord and brain as well as to execute the commands sent from brain and spinal cord.

Autonomic Nervous system:

Located along the spinal cord this system is independent of the central nervous system. It has an access to the information gathered by peripheral system and can control the peripheral nervous system. This nervous system controls the functioning of the internal organs. It consists of two divisions, Sympathetic nervous systemand parasympathetic nervous system.

Central and peripheral nervous systems consist of two types of nerve cells (neurons)

1) Sensory neurons - These fibers carry impulses to the brain which recognizes sensations like touch, pain, joint movement, pressure, vibration, tastes etc.
2) Motor neurons - These fibers carry the signals from the brain and bring about movements in the muscles of the body. These are further divided into upper and lower motor neuron. (Figure 1.3)

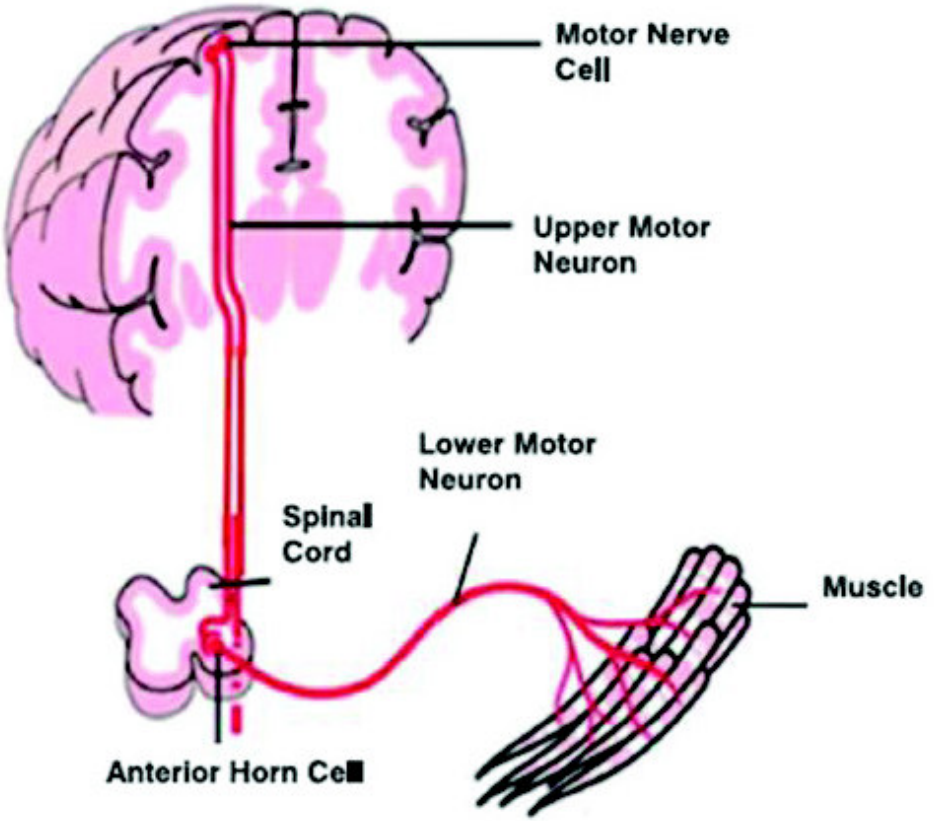


Figure 1.3- Upper and Lower motor neurons

i) Upper motor neurons: Upper motor neurons are the nerve cells that originate in the brainand end at the level of spinal cord

ii) Lower motors neurons:Lower motor neurons are the nerve cells that originate in the spinal cord and are spread all throughout the body

The musculoskeletal system

The lower motor neurons then enter the muscles to stimulate the muscles bring about the muscle contraction. When muscles contract they move bones to bring about movement. Every movement in the body is controlled by motor nerves.

What is motor neuron disease?

In Motor Neuron Diseases (MND), the motor neurons start dying (figure 1.4) and this causes muscle weakness in various parts of the body. Thus the affected nerves stop sending messages to the muscles gradually leading to weakness and thinning (atrophy/ wasting) of those muscles. Motor neuron disease encompasses a group diseases depending upon which of the motor neurons are damaged.

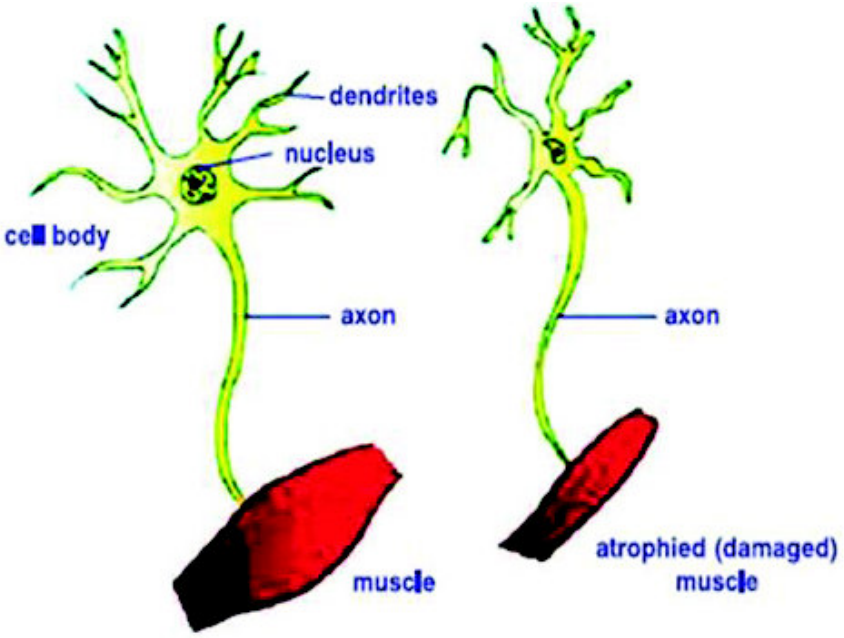


Figure 1.4: Damaged upper motor neuron

Types Of Motor Neuron Disorders

These are classified into three types (Figure 1.5)

1. Upper motor neuron disease
2. Lower motor neuron disease
3. Upper and lower motor neuron disease UMN & LMN Disorders

   In UMN and LMN disorders all the nerve fibers are affected. There are two types of this disease.

1. Amyotrophic Lateral Sclerosis (ALS)

It is the most common form of MND. It occurs due to degeneration of the neurons in brain and in the spinal cord as well as the peripheral neurons.

It is characterized by weakness and wasting of both upper limbs and lower limbs. There could be speech, swallowing and breathing impairment at any stage of the disease. Average life expectancy is 2 - 5 years from onset of symptoms. But this is only a combined average and there are many people who live beyond 10 - 20 years after the diagnosis. One of the greatest examples is Prof. Stephen Hawking, the well-known physicist who was diagnosed with MND more than 35 years ago.

Symptoms of ALS can start with the weakness of muscles of speech and swallowing (bulbar onset) or weakness of the muscles of arms and legs (limb onset).

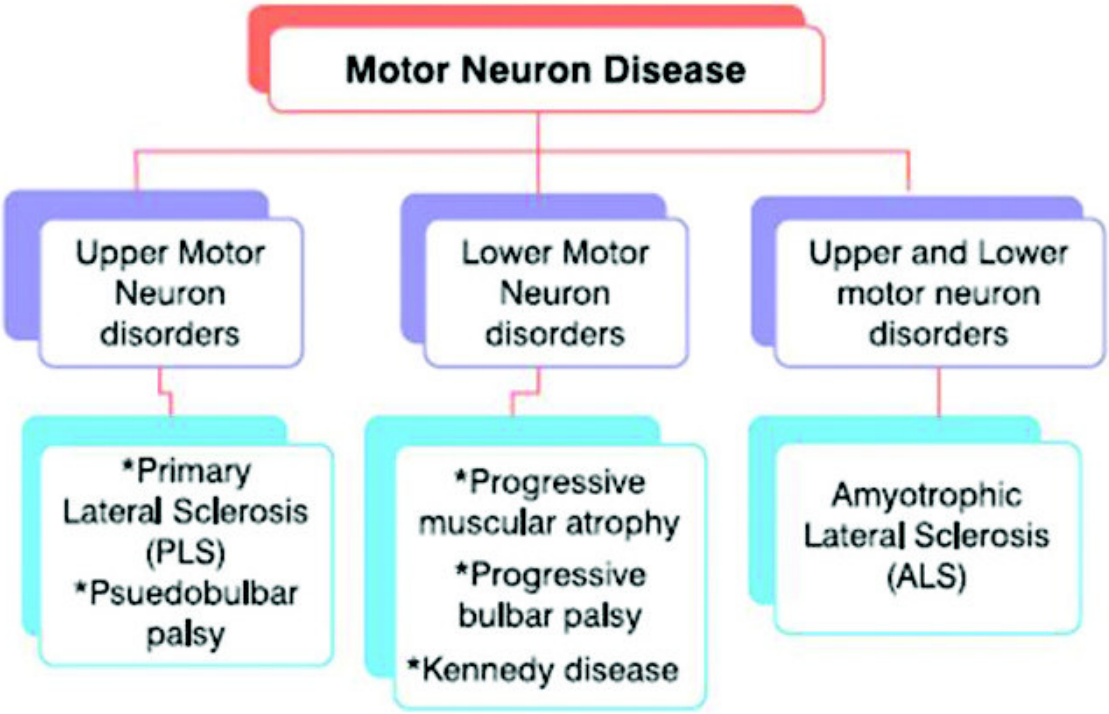


Figure 1.5: Types of motor neuron disease

UMN Disorders

These are the disorders in which the neurons originating from the brain and ending in spinal cord are damaged. The neurons originating from spinal cord, spreading throughout the body are preserved.

1. Primary Lateral Sclerosis (PLS)

This is a rare form of MND and where mainly the legs undergo weakness and in some cases arms. In most of the cases speech, swallowing and breathing remains unaffected till the advanced stage of the disease. Life expectancy in this disorder is near normal.

2. Psuedobulbar palsy

This is a rare form of MND. It can also be an associated symptom of other neurological disorders. It is characterized by progressive weakness of facial muscles, mastication muscles and laryngeal muscles. This causes facial expression, difficulty to chew and swallow and progressive difficulty of speech.

LMN Disorders

In these disorders the nerve originating from spinal cord and spreading though the body are damaged. The neurons originating from the brain and ending in spinal cord are preserved. There 3 different types of LMN disorders

1. Progressive Muscular Atrophy (PMA)

It is a rare form of MND. The common symptoms are weakness of legs and arms. Speech, swallowing and breathing is rarely affected or is affected only at the advanced stage of the disease. The prognosis of this form is better than other forms of LMN disorders.

2. Progressive Bulbar Palsy (PBP)

It is a very rare but most severe form of the disease. It occurs due to degeneration of motor neurons of the nerves that originate in the brain and control speech and swallowing. These nerves are called cranial nerves IX to XII. The disease is characterized by progressive difficulty with chewing, talking, and swallowing. Average life expectancy is between 6 months to 3 years from the onset of the symptoms.

Sometimes bulbar onset of ALS can be misunderstood as progressive bulbar palsy but as the disease progresses with progressive weakness of arms and legs, the diagnosis of ALS becomes evident.

3. Spinal Muscular Atrophy (SMA)

It is a genetic disorder caused due to a defect in either of the genes namely SMN1 (Survival Motor Neuron 1) or SMN2 (Survival Motor Neuron 2). The severity of the condition differs from patient to patient. There is progressive wasting of the proximal muscles (hip and shoulder muscles), and also weakness in those muscles.

There are 4 types of SMA, viz.

- Type I (Werdnig-Hoffman disease) is a severe form evident at birth.
- Type II is seen in chidren after 6 - 12 months.
- Type III (Kugelberg-Welander disease) is a milder form which has its manifestation in early childhood or adolescence.
- Type IV occurs only after 30 years of age.

4. Kennedy Disease

It is a rare form of MND. This form also runs in the family and is an inherited disorder. It is seen more in males and rarely in females. There is weakness in upper and lower extremities. There is more affection of throat and face making it difficult to swallow, talk and chew. Life expectancy is usually near normal.

What causes motor neuron disease?

Even with all the advancements in the field of medicine, the cause of MND is uncertain. Medical science has not been able to identify a definite cause for this disease but some of the risk factors have been identified. These risk factors may contribute to cause the disease but presence of these risk factors does not always lead to the disease.

MND has also been presumed to have a genetic origin where in the disease may be transmitted from one generation to the other. Some of the genes have been identified that may cause MND. When there is a gene involvement it is called as familial MND. However MND may be present without any family history (sporadic). But 90% of the times MND is sporadic and only 10% is of the times a definite genetic cause is identified (Figure 1.6).

Motor neuron disease like symptoms can be mimicked in many of the neurological disorders. Careful clinical assessment and thorough investigations are required to diagnose MND at earliest. It is important to rule out other causes that may lead to similar progressive weakness. How to rule these out is explained in detail in Chapter 3.

Incidence of ALS in USA is 2 per 100000, in Canada it is 2.4 per 100000, in UK it is 2.16 per 100000 however the incidence rate in India has not been identified.

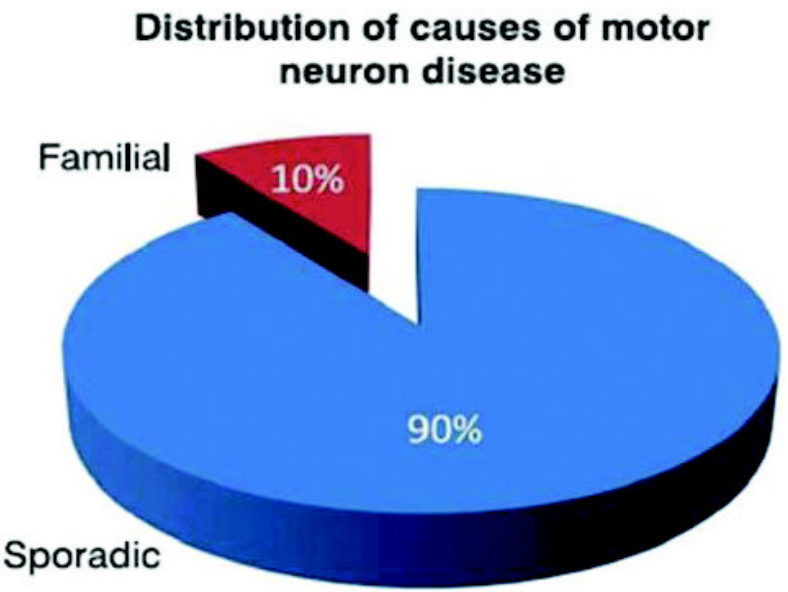


Figure 1.6: Distribution of causes of MND

Risk factors of MND

Some of the risk factors that have been identified (Figure 1.6) and have considerable evidence to show the increased risk of having MND are male gender, age beyond 50 years, presence of family history and mutation of the gene SOD1. It is important to understand these factors increase the risk of having the disease and it does not mean that people who don't exhibit these risk

factors will not get the disease. Simply put males are more prone to get the disease than females. People above the age of 50 years are more prone than younger population and people with SOD1 mutation are more prone to get the disease as compared to the ones without.

Other than these known factors there are several other risk factors. The supporting research is scanty and therefore these are only possible risk factors. Some of the proposed possible risk factors are lifestyle factors like sedentary lifestyle, stress, irregular sleeping hours etc. Some of the environmental factors like chronic exposure to neurotoxins, chronic exposure to air pollutants have also been presumed to be a risk factor. Figure 1.7 summarizes the risk factors for ALS.

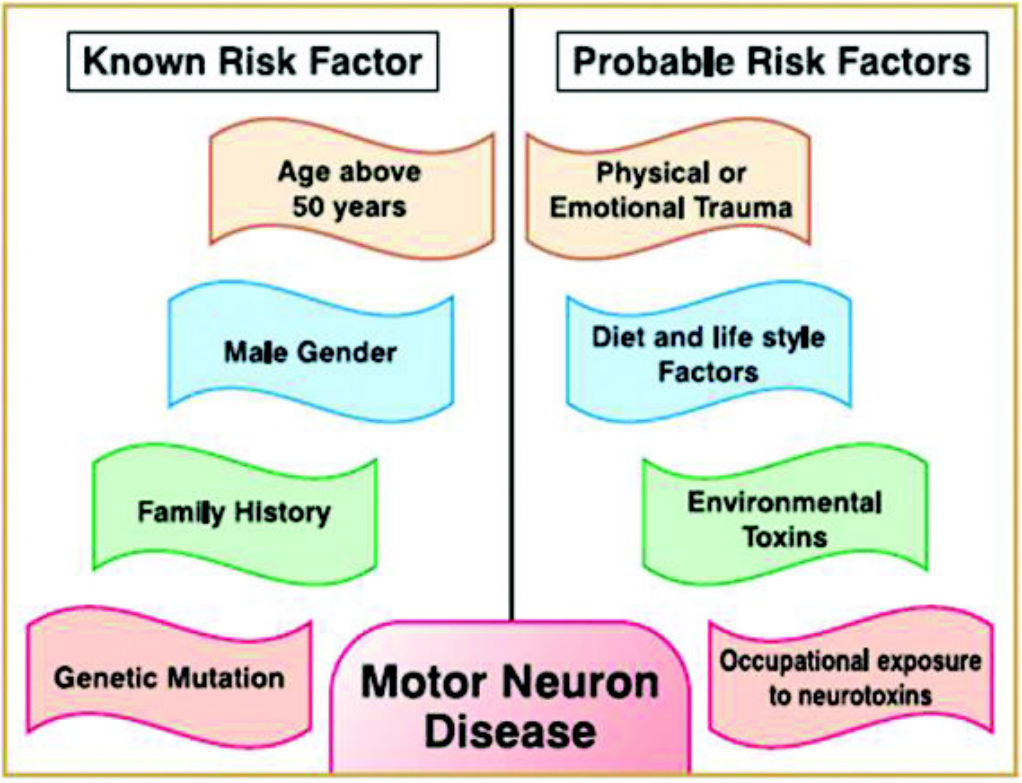


Figure 1.7: Risk factors for ALS

Chapter 2

What happens in Motor Neuron Disease?

As we saw in the last chapter motor neuron disease is the progressive degeneration and death of the nerves that control the movement of the muscles. Let us now understand what happens to these nerves at a local level. The chemical changes that damage these neurons. These mechanisms vary with the type of the disease.

Various chemical processes have been identified that can cause potential damage to the nerve cells in MND. These mechanisms seldom act alone and mostly it is the combination of one or two mechanisms that causes the degeneration. There is no diagnostic test available at the moment that can identify the mechanisms involved and therefore medicines addressing only one of the mechanisms fail to cure or control the disease.

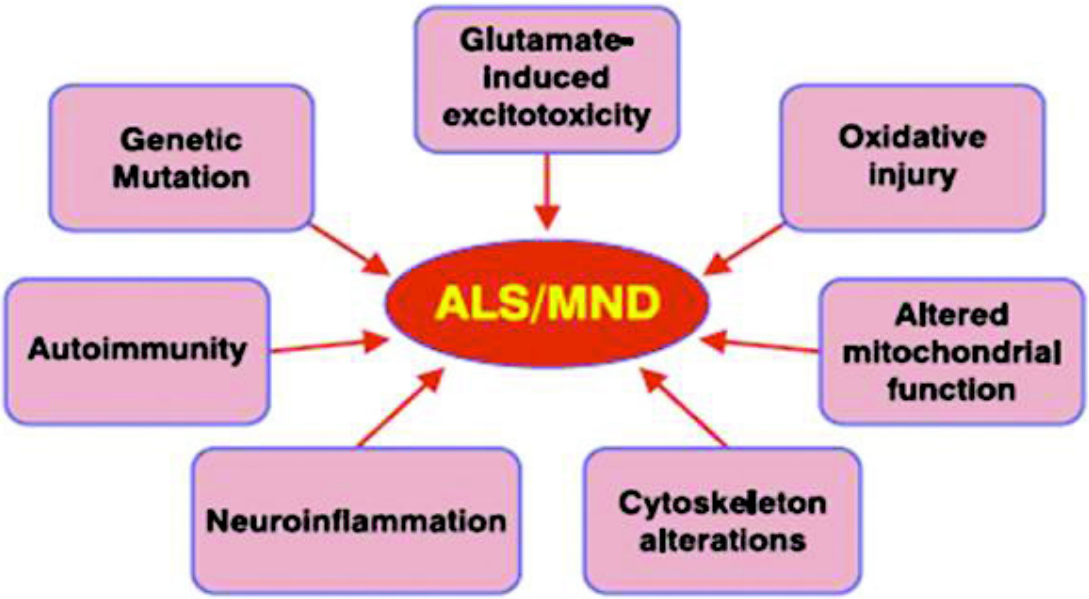


Figure 2.1- Processes causing damage to the nerve cells in ALS/MND

- **Glutamate-induced excitotoxicity:**

  In the nervous system there are chemicals that travel from one nerve cell to another. These chemicals are responsible for activating and inhibiting the function of the nerve cells and are called neurotransmitters. Glutamate is one such chemical which

is responsible for activating or stimulating the nerve fibers. If the concentration of glutamate around the nerve cells increases beyond normal levels then the nerve cell is stimulated excessively. With prolonged and continuous stimulation there are internal chemical imbalances in the cell. These changes damage the cells and resulting in their death. The phenomenon of damage to the cells due over activation is known as excitotoxicity.

- **Oxidative injury:**

  The food we eat is broken down to molecules which are absorbed in the blood and are used to build different tissues. The air we breathe is responsible for facilitating this breakdown. This breaking down and building of the molecules releases energy which is crucial to body functioning. One of the byproducts of this process is reactive oxygen species (ROS). Various enzymes in the body combine these with other molecules to form benign molecule. If this does not occur then the prolonged exposure to ROS can damage the proteins in the body including DNA and RNA. ROS is also useful as these are used by body's defense system to kill pathogens.

  Uncontrolled ROS formation that cannot be controlled by the body systems can cause damage to the junction of nerve and muscles as well as nerve cells. The damage due to increased oxidative stress is proposed as one of the mechanisms of nerve damage in ALS.

- **Altered mitochondrial function:**

  Mitochondria are the power houses of the cells. The ROS produced can alter the functioning of mitochondria. Mitochondrial dysfunction combined with oxidative stress causes abnormal neurodegeneration of motor neurons. Motor neurons are large in size and have higher energy requirements, which make them more susceptible to the injury caused by mitochondrial dysfunction. (1)

- **Cytoskeleton alterations:**

  Cytoskeleton is the skeleton or the wall of the cells in the body. Cytoskeleton is important to hold different organelles of the cells together. ROS can damage the proteins that are the building components of this cytoskeleton. Damaged cytoskeleton results in cell death and motor neuron degeneration.

- **Neuroinflammation:-**

  Inflammation is the response of the body to harmful stimuli like infection and injury. In ALS inflammation of the nervous system is observed even in absence of any identifiable trigger factors. If inflammatory response prolongs it leads to cell death and motor neuron degeneration causing symptoms of ALS.

- **Autoimmunity:**

  There is some evidence suggesting that body's defense system miss understands motor neurons as harmful stimuli. Immune system responds to this by secreting chemicals that damage the motor neuron.

  In patients with ALS all or some of these mechanisms may be responsible for the progressive degeneration of motor neurons.

- **Genetic mutation:-**

  Genes are the smallest functional units of the human body that provide instructions to all the cells in the body. Genes decode for preparation of proteins that build the body. Proteins provide structure and control the functions of the cells. Genes are inherited from the parents. A mutation in the gene is a fault or anomaly that leads to formation of faulty protein. A mutation gene can either make faulty proteins and alter the functionality of the cell or may lead absence of protein formation and subsequent loss of functionality and degeneration.

  How are the genes transferred from one generation to the other?

  Genes can be present as a copy where every parent contributes one gene. In some cases if any one of the two copies is faulty the result is a faulty gene and it can cause disease which is known as 'dominant' pattern of inheritance. Where as in some cases disease can occur only when both the copies are faulty and such pattern is called 'recessive'. Most of the genes known to cause ALS are inherited by dominant pattern.

  In the dominant pattern there is a 50% chance that the gene will be inherited by the child but all the children that receive the gene will have the disease.

  Genes that Cause ALS

  Various genes have been identified to cause ALS. Figure 2.2 illustrates the percentage of ALS patients with different genetic mutation.

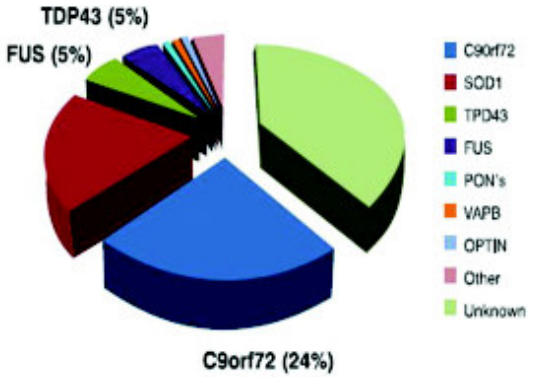


Figure 2.2 The percentage distribution of ALS patients with different genetic mutations

- **C9ORF72**

  Named after its location on the chromosome, this was the most recently found but the most common genetic cause of ALS. It is inherited in the dominant pattern. This gene mutation can cause another neurodegenerative disease fronto-temporal dementia. The symptoms of this disease are progressive memory loss and progressive loss voluntary physical activity. Sometimes the symptoms of both these disorders may overlap.

- **Cu/Zn Superoxide Dismutase 1 (SOD1)**

  This was first gene to be known to cause ALS. Presence of this gene leads to formation of protein clusters inside the cell that lead to cell death. It is unclear whether these clusters are formed inside the motor neuron itself or surrounding cells that necessary for the survival of the motor neuron.

- **TDP-43**

  This gene was also recently found to be associated with ALS pathology. It is inherited in a dominant pattern. Mutation in this gene causes a protein to displace from its actual location in the central part of the cell to the peripheral location. The protein forms clusters in this case as well. These clusters are toxic to cells and may cause cell death. Even in absence of the mutation of the gene such protein clusters are found in ALS patients.

- **FUS**

  Fused in sarcoma (FUS) is a gene that is inherited in a dominant pattern and may have similar effects as that of TDP-43.

- **Ubiquilin-2**

  This gene is found on the X-chromosome. This chromosome is the chromosome that determines female gender. Both males and females are affected by the mutation of this gene. The gene leads to formation of a protein molecule that is required for clearance of toxic proteins from the cell. Mutation of this gene also leads to toxic protein aggregates in the cell and death of cells.

  Other genes that are involved in the pathology of ALS are iVCP (valosin-containing protein), alsin, senataxin, and angiogenin and optineurin.

Chapter 3

Signs and symptoms of Motor Neuron Disease

The signs and symptoms of motor neuron disease (MND) are very diverse which makes diagnosis difficult and often late. The most important thing is that MND causes only muscle weakness but the sensations remain normal. The weakness may start in the limbs or in the mouth. One should immediately get checked by a doctor if any of the below symptoms are seen.

Most common symptom of motor neuron disease is progressive muscle weakness. Muscles weakness is painless and increases gradually. Fatigue, Cramps and fasciculations are some common symptoms of MND other than muscle weakness. Depending on which muscles of the body have undergone weakness the symptoms and progression varies. Although motor neuron disease is a disease of motor nerves these patients may exhibit emotional and behavioral disturbances.

The progression of motor neuron disease cannot be predicted. It is all the more unpredictable in ALS where in the onset is in one region and the disease then progresses to the rest of the body. The symptoms of the disease are therefore discussed in general. The symptoms in the early, progressive and advanced stage of the disease can only be divided based on the region of onset. The symptoms can be divided into bulbar and non-bulbar symptoms.

Bulbar symptoms:

Bulbar symptoms are caused due to weakness of the muscles responsible for speech, swallowing and respiration. Bulbar region is the region of head and neck.

Bulbar symptoms include

1. Increased speech effort: Before slurring person with MND may experience laborious speaking. This means that the person is unable to speak for long duration. The quality of voice may become shallow and soft after sometime and the person may not be able speak loudly and clearly.
2. Slurring of speech: Slurring of speech is characterized by inability to speak fast and clear.
3. Breathlessness upon speaking (Dysponea in speaking): This is a symptom of advanced disease when the person experiences some difficulty in breathing while speaking and has to stop talking to breath.
4. Neck drop: Neck drop occurs due to weakness of the neck extensors this is a symptoms of advanced disease. Persons with MND will find it difficult to hold their neck up against gravity even for a few seconds and neck may flop forward and backward uncontrolled with changes in the trunk position.

5. Fasciculation of the tongue: This is one of the earliest symptoms. Fasciculations are the involuntary twitching movement in the tongue.

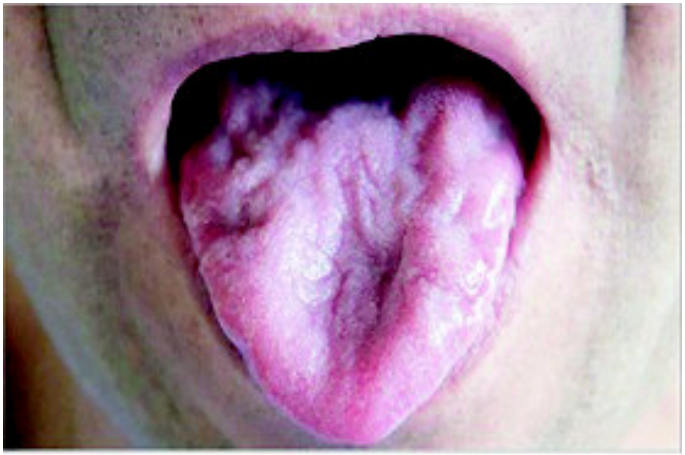


Figure 3.1:Fasciculations of the tongue

6. Drooling: Due to weakness in the facial muscles mouth closure and lip sealing is difficult this gives rise to leaking of saliva and drooling. Initially drooling may occur only at night when voluntary muscle activity is suppressed during sleep. It may then progress to drooling infrequently during day time. As the weakness of swallowing muscles also sets in the saliva that is formed cannot cleared by swallowing and the patient may experience continuous drooling throughout the day.
7. Inability to swallow: Every movement in the body is carried out by muscles. Swallowing food is possible only with muscle activation as the muscle grow weaker persons with ALS may not be able to swallow the food. The initial symptom may infrequent cough and choking while drinking liquids. Liquids and solids are more difficult to swallow where as semi solid food can be swallowed easily.
8. Loss of tongue movements: Tongue is a muscle. Progressive weakness of this can cause inability to move tongue effectively. This can cause inability to speak clearly and inability to clean the food particles from the mouth. In advanced disease the tongue may fall back on the air tract causing a feeling of breathlessness.
9. Inability to perform rapid eye movements: In the advanced stage of the disease weakness of ocular muscles (the muscles that control the eye movements) may be observed and this can cause inability to perform rapid eye movements and slow pursuit reactions.
10. Facial muscle weakness: Facial muscle weakness in the early phases leads to inability to close the mouth, inability to balloon up the cheeks, inability raise eye brows, inability to close eyes tightly and inability to flare the nostrils.

11. Breathlessness upon lying straight on the back (Orthopnoae): Lying supine pushes the abdominal organs on to the diaphragm which is the muscle responsible for breathing this leads to resistance for the muscles action. In the advanced stage of the disease a person is unable to breath against this resistance and therefore may feel out of breath upon lying flat on their back. This is usually relieved in the reclined position and when one turns on to side.
12. Breathlessness with minimal activity (Dyspnoea): This is also a symptoms of advanced disease and person may feel easily tired and out of breath with minimal activities like brushing, eating, bathing etc.
13. Inability to breathe without external assistance: This is a symptoms of advanced bulbar disease where in the respiratory muscles grow so weak that the person is unable to breath unless assisted externally by devices like Bi-Pap or ventilator.

Non-bulbar symptoms:

Non bulbar symptoms are the symptoms observed in other parts of the body.

1. Fasciculations: Fasciculation is uncontrolled twitching of muscles. This may be triggered upon performing a particular activity or at rest and may subside on its own. Fasciculations are not painful.
2. Cramps: Cramps are painful, sustained and uncontrolled muscle contractions. Most often cramps are triggered when a weak muscles is put through performing a stressful activity. Sometimes cramps may be triggered at rest as well. Usually cramps are painful, mostly the pain is bearable. Sometimes severe cramps can lead to severe pain.
3. Foot drop: foot drop is characterized floppy feet that drop down when the leg is lifted up to walk. This downward movement of the feet requires the leg to be lifted higher while walking and causes a slapping action on the ground while stepping.

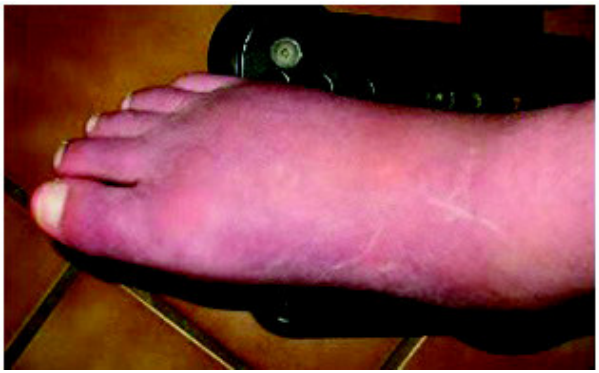


Figure 3.2- Foot drop

Foot drop may be one of the first symptoms of limb onset MND. It usually takes time before the floppyness of the feet is noted. At an earliest stage foot drop may simply manifest as frequent falls and stumbling at obstacles.

4. Wasting of muscles: Wasting of muscles can be identified as unusual thinning of the muscles and prominence of bony surfaces. Muscle wasting is always associated with muscle weakness and inability to perform movements (Figure 3.3 to 3.6)

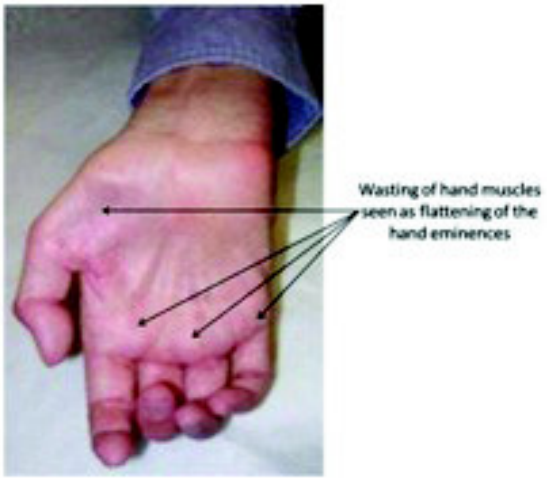


Figure 3.3 Wasting of scapular muscles

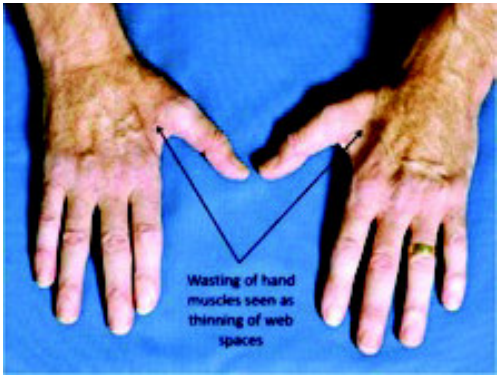


Figure 3.4 Wasting of hands

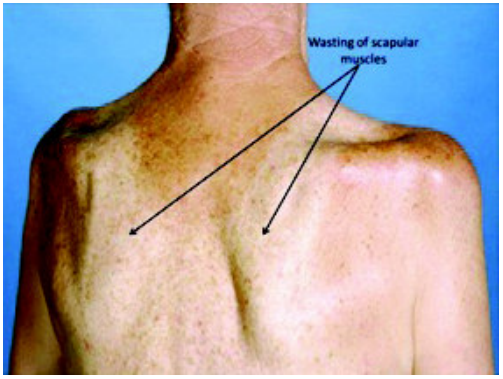


Figure 3.5:Wasting of muscles of palms

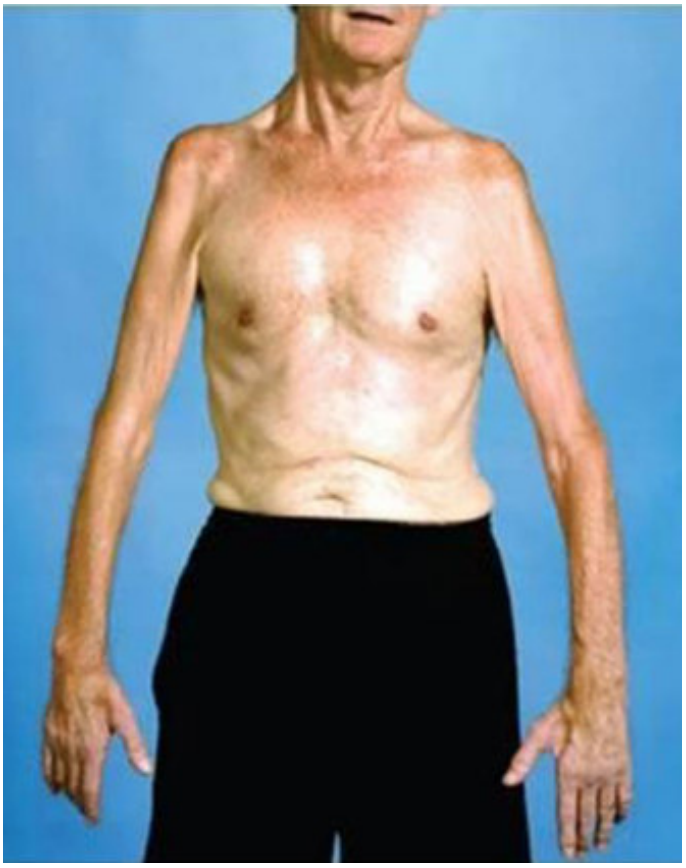


Figure 3.6 Man in the Barrel syndrome

5. Muscles weakness: It is the most common symptom of ALS and is characterized by progressive inability to perform movements. Typically the weakness sets in as loss of endurance where in it is difficult to perform a particular activity for a longer time but a person is able to complete the activity. As the weakness progresses there is more and more restriction and limitation of activities. Initially the limitation is only while performing the activities against gravity but in later stages movement may be difficult in gravity eliminated plane.
    - Usually more pronounced on one side than the other at first.
    - Loss of function is usually more rapid in the legs among people with familial ALS and in the arms among those with sporadic ALS.
    - Legs weakness may first become apparent by an increased frequency of stumbling on uneven surface or an unexplained difficulty climbing stairs.
    - Arm weakness may lead to difficulty grasping and holding a cup, for instance or loss of dexterity in the fingers.

- While initial weakness may be limited to one region, ALS almost always progresses rapidly to involve virtually all the voluntary muscle group in the body.

6. Loss of dexterity (co ordinate movements of the hands): This may be one of the initial symptoms of limb onset MND. Loss of dexterity can manifest as inability to button the shirt, fix the hooks and loops of clothes, hand writing changes, fatigue while writing, inability to handle fine objects spilling food while eating and difficulty in handling cutlery.
7. Slow movements and tightness of body movements: Slowness and tightness of movement in MND usually occurs at an advanced stage when the tone of the muscles increases providing resistance to movement. Slowness can also occur due to progressive weakness when the muscles are unable to carry out a particular movement.
8. Emotional disturbances: As the disease progresses loss neurons in the brain may spread to other regions and some symptoms like uncontrolled laughter, crying spells with minimal triggers, sudden emotional outbursts, anger episodes and frequent episodes depression and hopelessness may be observed.

The progression of each symptom through the different stages of the disease is mentioned in Table 3.1.

Table 3.1 Progression of the symptoms

Symptom	Early stage	Progressive stage	Advanced stage
Speech disturbances	• Increased effort of speech • Softening of voice after talking for sometime	• Slurring of speech • Inability to speak for a long time with high fatigability • Inability to speak loudly	• Unintelligible speech • Complete loss of speech
Swallowing difficulty	• Infrequent chocking and coughing after drinking liquids	• More frequent episodes of choking and coughing • Inability to swallow tablets • Inability to swallow solid food, food needs to be mashed up	• Inability to swallow • Considerable aspiration • Continuous drooling of saliva

Symptom	Early stage	Progressive stage	Advanced stage
Breathing difficulty	• Heavy breathing after physical activity • Feeling out of breath upon walking or climbing stairs	• Feeling out of breath while eating, bathing and activities of daily living. • Feeling out of breath upon lying in bed flat • Inability to breath while talking for a longtime • Using neck muscles excessively while breathing	• Inability to breathe without external device support
Drooling	• Minimal drooling at nighttime • Drooling in postures with neck in flexion • General feeling of excess saliva in mouth	• Considerable drool-ing throughout the day • Spitting of saliva while speaking and difficulty to swallow the excess saliva	• Sticky, thick saliva • Inability to swallow the saliva • Continuous drooling
Facial muscle weakness	• Twitching in the cheeks and chin • Spilling of water while gargling due to inability	• Difficulty in opening the mouth • Affected facial expressions	• Facial asymmetry • Minimal opening of the mouth
Neck muscle weakness	• Mild fasciculations in the neck region • Difficulty to the lift the head up from supine	• Difficulty holding the neck in the upright position • Difficulty lifting the head up after flexing the neck	• Inability to look up in sitting posture • Severe neck cramps
Fasciculations	• Uncontrolled, sudden twitching of muscles	• Increased frequ-ency of twitching • Twitching of muscles in more	• Reduced frequency of twitching in the body parts that have undergone

Signs and symptoms in different types of MND

1. Amyotrophic lateral sclerosis

- Progressive muscle weakness in the upper or lower limbs.
- Approximately one fourth of patients presents with dysarthria, followed by progressive dysphagia (bulbar-onset ALS). (1)
- In ALS with SOD1 mutations, extremity onset, particularly in the legs, is much more common than bulbar onset. About 5% of cases present with respiratory weakness without significant limb or bulbar symptoms. (2)
- The flail arm and flail leg variants are initially localized forms with predominantly lower motor neuron (LMN) symptoms (3) These are characterized by progressive weakness and severe wasting of the upper and lower limbs, bilaterally.
- Rectal and bladder sphincters muscles are usually spared.
- Sensory examination is usually unremarkable.
- Approximately 5% of patients with ALS develop dementia of the frontotemporal type, characterized by behavioral changes with or without language dysfunction.

2. Primary lateral sclerosis

- The age of onset for primary lateral sclerosis (PLS) is usually between 40 and 60 years.
- Spasticity in the legs accompanied by hyperactive deep tendon reflexes, clonus, and Babinski sign.

3. Progressive bulbar palsy

- Brown-Vialetto-Van Laere syndrome typically presents with progressive sensorineural hearing loss.
- The age of onset of the initial symptoms reportedly ranges from infancy to the third decade of life. (6)
- The hearing loss is usually followed by other symptoms, including abnormalities of lower CNs VII-XII and LMN signs in the limbs, with an interval of several years. This interval has been reported to be shorter in males (mean, approximately 5 y) than in females (mean, almost 11 y). (6) Abnormalities of CNs II-VI occur much less frequently.
- Fazio-Londe disease manifests itself in childhood as rapidly progressive weakness of the tongue, face, and pharyngeal muscles, as well as progressive upper limb weakness. No hearing impairment is seen.

4. Spinal muscular atrophy

The clinical picture of spinal muscular atrophy (SMA) is highly variable and represents a continuum. The age of onset ranges from before birth to adulthood.

Type I SMA :- This is the most severe form. Patients present with profound hypotonia and generalized weakness ("floppy infants") and never achieve the ability to sit. By definition, all patients present before age 6 months (sometimes with onset in the prenatal period). The diaphragm and the extraocular muscles tend to be spared (in contrast to SMA-plus disease types and severe congenital SMA). (7) No cardiac muscle involvement is seen. Creatine kinase is normal.

Type II SMA :- The onset of type II SMA is before the age of 18 months. There may be some overlap with type I SMA, but the median age of onset is generally later than in type I. The clinical course of type II SMA is marked by periods of apparent arrest in clinical progression. Patients are able to sit independently, but they are never able to walk unaided. Spine deformities and contractures of all major joints often develop.

Type III SMA(Kugelberg-Welander disease):- Type III is a mild form of childhood and juvenile SMA. By definition, the disease onset is usually after the age of 18 months, but it may occur over a wide range, sometimes as late as the third decade of life. Patients gain the ability to walk without support. Spine deformities and contractures are frequent complications.

Type IV SMA:-Type IV is an adult-onset type (age > 30 y) and represents the mildest form within the spectrum of SMA phenotypes. Patients present with pronounced proximal weakness, and the clinical picture is very similar to that of a limb-girdle muscular atrophy.

5. X-linked spinobulbar muscular atrophy (Kennedy disease)

X-linked spinobulbar atrophy (SBMA) is clinically characterized by adult-onset limb and bulbar weakness, muscular atrophy, and fasciculation, with frequent occurrence of endocrine disturbances such as gynecomastia, testicular atrophy, hypercholesterolemia, and diabetes mellitus. Asymptomatic patients are also present.

One clinical study reported that the mean age at first onset of muscle weakness was 41 years and that the most common presenting symptom was muscle cramps, followed by tremors and leg weakness. (8) Also reported was that muscle strength and function correlated directly with serum testosterone levels.

6. Hereditary spastic paraparesis

- In pure (uncomplicated) hereditary spastic paraparesis (HSP), the age of disease onset ranges from infancy to the eighth decade, (4) and the disease severity varies; both reflect marked interfamilial variation.

- The essential clinical findings are slowly progressive and often include severe spasticity, hyperreflexia, and weakness in a pyramidal distribution, noticeably in both lower limbs, with extensor plantar responses.
- Most patients with HSP present with difficulty in walking or a gait disturbance. Upper limb involvement is usually mild. Notably, as many as 25% of affected patients are asymptomatic. (5)
- Sensory impairment is seen in 10-65% of cases and usually consists of diminished vibration sense and, less often, diminished joint-position sense in the lower extremities. (5)
- Urinary sphincter dysfunction occurs in up to 50% of patients, whereas anal sphincter involvement is unusual.[29] Important negative clinical findings include no cranial nerve (CN) involvement and no corticobulbar tract involvement.

7. Hirayama Disease:

Initially begins as progressive muscle weakness usually in the upper extremities. The progressive course later stangnates and the weakness only limits to distal extremities. It is a form of spinal muscular atrophy.

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Chapter 4

How to diagnose ALS/MND?

Some of signs and symptoms of ALS/MND can also be seen in other disorders. So it can be misdiagnosed and the line of treatment can go in a different direction. For eg. , in early stages when there is weakness in the hand muscles, people can get diagnosed as Carpel Tunnel syndrome or cervical spondylosis.

The only diagnostic test available for MND/ALS is Electromyography (EMG) and Nerve conduction Velocity testing (NCV). The other tests like blood tests, CSF studies, MRI of brain/spine are done to rule out other possibilities or differential diagnosis. In case of familial ALS, genetic testing can be done to confirm the diagnosis. But, one has to remember that EMG/NCV can be near normal in the very early stages of the disease. Very often NCV testing may be misleading to carpal tunnel syndrome, cervical spondylosis. A lot of patients have even got operated for CTS and prolapsed disc due to misdiagnosis. Thus, there is no definite diagnostic test for MND/ALS. The diagnosis is often made by a combination of clinical assessment and EMG/NCV testing along with exclusion of other possible causes.

The diagnoses of ALS is thus made possible by

1. History, physical neurological examinations which may show findings suggesting suspected, possible, probable or definite ALS
2. Electrophysiological examination (EMG/NCV) to identify and confirm involvement of motor neurons, the level (i.e. anterior horn cell) and the extent of involvement, and also to exclude other disorders
3. Neuroimaging (CT, MRI scan) examination to exclude other disease processes
4. Laboratory (blood, CSF) examinations to exclude closely related neurological disorders
5. Repetition of clinical and electrophysiological examinations at six months interval to study progression of the disease.

Although no definite diagnostic test exists for MND/ALS except for EMG/NCV and one genetic testing; clinical presentation and various tests such as blood tests, CSF tests, muscle and nerve biopsies, neuroimaging such as MRI support the diagnosis and help rule out other disorders.

Clinical assessment:

Signs and symptoms vary according to the type of MND. They are sometimes peculiar of that type of MND. Therefore, the clinical presentation of the disease and assessment by a physician may reveal the diagnosis of any MND.

Table 1: Shows which type of MND affects which motor neurons

Type	UMN degeneration	LMN degeneration
Amyotrophic lateral sclerosis (ALS)	Present	Present
Primary lateral sclerosis (PLS)	Present	Absent
Progressive muscular atrophy (PMA)	Absent	Present
Progressive bulbar	Absent	Present - bulbar area
palsy (PBP)		
Pseudobulbar palsy	Present - bulbar area	Absent

Abbreviations: UMN - Upper Motor Neuron; LMN - Lower Motor Neuron

Following are signs and symptoms associated with MND depending on motor neurons involved:

In presence of UMN degeneration:

- Spasticity (increased tone of muscles)
- Hyperreflexia (exaggerated deep tendon reflexes,)
- Pathological reflexes (Babinski's sign positive,)
- Muscle weakness
- Muscle spasms (flexor or extensor muscles)
- Dyssynergia (abrupt, uncoordinated voluntary movements)

In presence of LMN degeneration:

- Muscle weakness and atrophy (loss of muscle mass, figure)
- Hypotonia (decreased tone of muscles)
- Hyporeflexia (absent/diminished deep tendon reflexes)
- Muscle cramps
- Fasciculations (flicker of movement caused by involuntary muscle contraction)

In presence of bulbar degeneration:

- Dysphagia (difficulty in swallowing)
- Dysarthria (difficulty speaking)

- Sialorrhea (excessive salivation)
- Dysphonia (difficulty producing sound)
- Pseudobulbar affect (episodes of uncontrollable crying and/or laughing)

In presence of respiratory muscle weakness:

- Breathlessness on exertion
- Orthopnea (breathlessness on lying down flat)
- Nocturnal breathlessness (at night, during sleep)

Diagnosis of ALS requires presence of LMN signs + UMN signs + history of spread of the disease + absence of pathological, electrophysiological and neuroimaging evidence of other disorder.

The El Escorial Revised Criteria for the diagnosis of ALS

Lack of definite diagnostic test and variability of clinical picture makes the diagnosis of ALS difficult. Therefore, The World Federation of Neurology (WFN), in the year 1990, developed a diagnostic algorithm called the El Escorial criteria which was revised in the year 1999.

El Escorial revised criteria are considered standard for the diagnosis of ALS in clinical settings and research purposes.

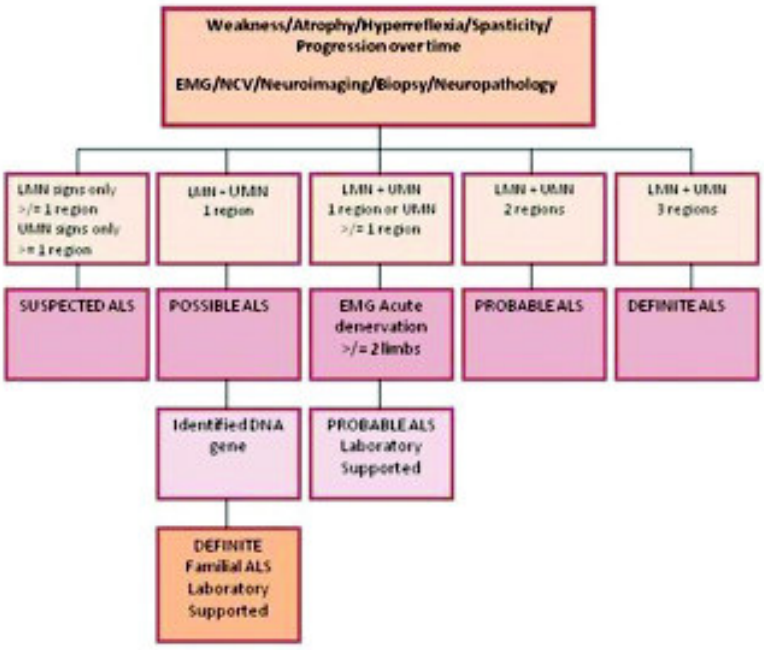

Fig 4.1: The algorithm of El Escorial revised criteria:

Hence, UMN and LMN signs in at least 3 body segments are required to diagnose ALS.

Functional Assessment

As ALS progresses patients' functions and independence diminish. ALS Functional Rating Scale Revised (ALS-FRS R) scale is a standard scale used extensively for the evaluation of

ALS patient's degree of functional impairment. This scale can be scored serially to objectively assess response to any treatment or the progression of disease.

It is a 12 item scale which measures functions of various muscles. Each item is scored from 0 to 4, where 4 stands for no involvement by the disease and 0 stands for maximal involvement. The scores of all 12 items are then summed up to obtain a final score.

ALS-FRS Revised scale

1. **Speech**
   - 4 - Normal Speech processes
   - 3 - Detectable speech with disturbances
   - 2 - Intelligible with repeating
   - 1 - Speech combined with nonvocal communication
   - 0 - Loss of useful speech
2. **Salivation**
   - 4 - Normal
   - 3 - Slight but definite excess of saliva in mouth; may have nighttime drooling
   - 2 - Moderately excessive saliva; may have minimal drooling
   - 1 - Marked excess of saliva with some drooling 0 - Marked drooling; requires constant tissue or handkerchief
3. **Swallowing**
   - 4 - Normal eating habits
   - 3 - Early eating problems - occasional choking
   - 2 - Dietary consistency changes
   - 1 - Needs supplemental tube feeding
   - 0 - NPO (exclusively parenteral or enteral feeding)
4. **Handwriting**
   - 4 - Normal
   - 3 - Slow or sloppy; all words are legible

2 - Not all words are legible

1 - Able to grip pen but unable to write 0 - Unable to grip pen

5. Does subject have gastrostomy?

No- Answer 5a

Yes - Answer 5b

5.a Cutting Food and Handling Utensils (patients without gastrostomy)

4 - Normal

3 - Somewhat slow and clumsy, but no help needed

2 - Can cut most foods, although clumsy and slow; some help needed

1 - Food must be cut by someone, but can still feed slowly

0 - Needs to be fed

5.b Cutting Food and Handling Utensils (alternate scale for patients with gastrostomy)

4 - Normal

3 - Clumsy but able to perform all manipulations independently

2 - Some help needed with closures and fasteners

1 - Provides minimal assistance to caregivers 0 - Unable to perform any aspect of task

6. Dressing and Hygiene

4 - Normal function

3 - Independent and complete self-care with effort or decreased efficiency

2 - Intermittent assistance or substitute methods

1 - Needs attendant for self-care 0 - Total dependence

7. Turning in bed and adjusting bed clothes

4 - Normal

3 - Somewhat slow and clumsy, but no help needed

2 - Can turn alone or adjust sheets, but with great difficulty

1 - Can initiate, but not turn or adjust sheets alone

0 - Helpless

8. Walking

4 - Normal

3 - Early ambulation difficulties

2 - Walks with assistance

1 - Nonambulatory functional movement only 0 - No purposeful leg movement

9. Climbing Stairs

4 - Normal

3 - Slow

2 - Mild unsteadiness or fatigue

1 - Needs assistance 0 - Cannot do

10. Dyspnea

4 - None

3 - Occurs when walking

2 - Occurs with one or more of the following: eating, bathing, dressing

1 - Occurs at rest, difficulty breathing when either sitting or lying

0 - Significant difficulty, considering using mechanical respiratory support

11. Orthopnea

4 - None

3 - Some difficulty sleeping at night due to shortness of breath, does not routinely use more than two pillows

2 - Needs extra pillow in order to sleep (more than two)

1 - Can only sleep sitting up 0 - Unable to sleep

12. Respiratory Insufficiency

4 - None

3 - Intermittent use of NIPPV

2 - Continuous use of NIPPV during the night

1 - Continuous use of NIPPV during the night and day

0 - Invasive mechanical ventilation by intubation or tracheostomy

Total ALS-FRS R score indicates severity of ALS as follows:

- >40 (minimal to mild)
- 39-30 (mild to moderate)
- < 30 (moderate to severe)
- < 20 (advanced disease)

Reduction in the score by 8-10 points may indicate severe implications if the reduction is seen in the respiratory or bulbar components.

Investigations

Laboratory tests:

There is no clinical laboratory test, findings of which rules out the diagnosis of ALS. Whereas depending on clinical judgment by your physician, specific laboratory tests may be ordered, which may include blood and/or CSF (cerebrospinal fluid - fluid found in the spaces of brain and spine) and/or urine tests. When the laboratory abnormalities or associated infection or disease are treated, which results in reversal/stabilization of symptoms, ALS may be ruled out. Whereas, when the appropriate treatment does not result in improvement, ALS may be considered.

Electrophysiological tests:

These tests include Electromyography (EMG), Nerve conduction velocity (NCV).

What is EMG-NCV testing?

EMG is a test used to measure the response of the muscles upon electrical stimulation by the nerves. The test is used to detect abnormalities in the transmission of electrical impulses from the nerves to the muscles. Most commonly EMG is accompanied by the measurement of nerve conduction velocity (NCV). Abnormalities in the speed of nerve conduction help identify the location of the damage to the nerve. Hence EMG-NCV testing helps to identify the exact location of damage. Needle EMG is a commonly used procedure to measure electrical activity of muscles. Needle EMG is an invasive technique as explained below.

The procedure of needle EMG and NCV testing

Preparation for the procedure:

- No special preparation or fasting is required for the test.
- You may be advised to avoid any caffeinated drinks 2 - 3 hours before the test.
- Wear loose clothes that allow exposure of all the regions of the body or that can be easily removed.
- Inform the doctor if you have any metallic implants in the body or pacemakers.
- Do not use any oils or lotions on the day of the examination.

During the procedure:

- You will be advised to remove any clothing, hairpins, eyeglasses, hearing aids, or other metal objects that may interfere with the procedure.
- The test can be performed in sitting or lying down.
- The muscles to be studied will be located. The skin region will be cleansed using an antiseptic solution.

- An electrode to complete the circuit (ground electrode) will be placed under your arm or leg depending on the muscle to be examined.
- Multiple needles may be inserted to record the impulses. There may be mild to moderate pain due to needle insertion. However if pain is severe inform the doctor as it may interfere with the results.
- Once the needle is securely placed in its location, you will be asked to perform a small movement of that muscle and later on a more forceful movement. The electrical activity will be recorded during these contractions. Multiple such muscles will be tested in the regions of arms and legs.
- For NCV testing, two superficial electrodes will be placed on the skin. One of them will be a recording electrode and the other stimulating electrode. A small current will be passed through the stimulating electrode. The time taken to carry this impulse to the recording electrode and the distance between the two electrodes will determine the conduction velocity of the nerve. Multiple nerves may be tested. You may feel mild discomfort as the current is passed through your body but generally it is pain free.

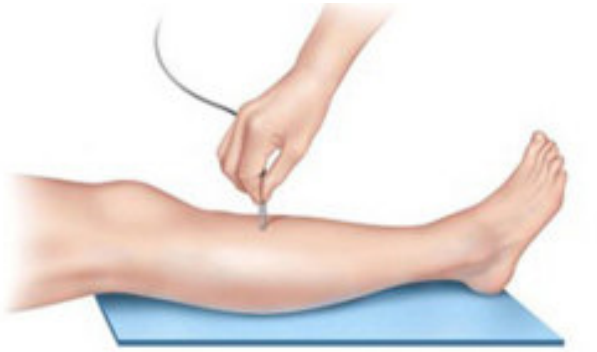

Figure 4.2 Procedure for EMG

Post procedure precautions:

- You may experience dull soreness to moderate pain at the site of needle insertion.
- Notify your doctor immediately in case of any swelling, pus, persistent severe pain and tenderness.

Interpretation of the results:

Following are the EMG findings which show primary LMN degeneration:

- Reduced recruitment
- Reduced interference pattern
- Large amplitude, long duration motor unit action potentials
- Fibrillation potentials

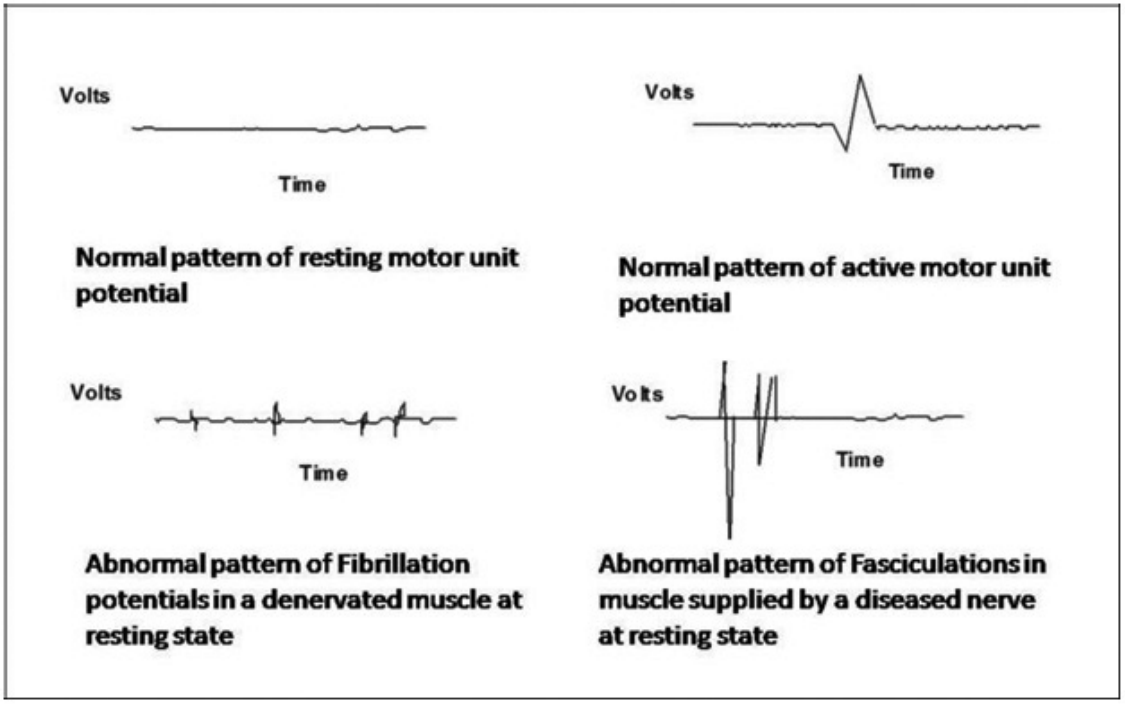

Figure 4.3 Normal and Abnormal EMG patterns

Following are the NCV findings which show UMN degeneration:

- NCV study may show normal motor and sensory conduction
- It may sometimes show increase in central motor conduction velocity also

Evidence of LMN degeneration in at least two muscles of different root or peripheral nerve innervation in two or more of the four (bulbar, cervical, thoracic, lumbosacral) regions confirms the diagnosis of ALS.

What does Neuroimaging include?

There are no neuroimaging tests which confirm the diagnosis of ALS. Whereas, to rule out other causes of weakness, your physician may order an MRI scan (Magnetic Resonance Imaging) of the brain and/or spine. Absence of imaging abnormalities in patients with clinical and/or electrophysiological evidence will help confirm the diagnosis from probable ALS to definite ALS.

What is MRI of brain/spine?

MRI brain/spine is a non-invasive imaging technique used to identify any structural abnormalities in the brain and spinal cord. MRI uses a powerful magnetic field, radio waves and a computer to produce detailed pictures of organs, soft tissues, bone and all

other body structures. MRI does not use X-rays (ionizing radiation). The images obtained are studied on the computer by the radiologist and are printed.

The procedure of MRI

Preparation for the procedure:

- Wear loose clothes that can be easily removed.
- Remove all your jewelry, glasses with metal frames, clothing with metal hooks or any other metal articles on your body.
- Do not carry any electrical devices like mobile phones, tablets, pagers etc. in the examination room
- Fasting and food restrictions if any will be instructed by the MRI center where your appointment is been taken. Unless instructed otherwise you can carry on with your daily routine.
- Inform your radiologist about any allergies if you have, any medication you take and any recent injury, surgeries or metal implants.
- Pregnant women must inform the radiologist about their pregnancy. MRI testing should be avoided during the first trimester. So far no documented ill effects of MRI on pregnant women or fetuses have been noted.

  If you feel anxious or you are fearful of closed spaces (claustrophobic) inform you physician and radiologist in advance. If counseling and reassurance does not relieve your fear, your physician may prescribe mild sedative for the examination. The amount of sedation and anesthesia required depends upon patient's age and the type of MRI. If sedation or anesthesia is required to complete the test, you may be asked to fast for some duration prior to testing.
- Implanted devices like artificial heart valves, pacemakers, metal pins, screws, plates, stents, implanted nerve stimulators etc. can interfere with the MR signal which can lead to faulty scanning, and false and misleading images may be produced. Therefore, you should inform your doctor and radiologist about these implants in advance. It is also advisable to consult the doctor who has fitted the implant for detailed information regarding MRI compatibility and safety of procedure.
- Some tattoo ink contains traces of metal, but most of the tattoos are safe in MRI scanner.
- Family members or attendants who accompany patients into the scanning room also need to remove metal objects and inform the technician of any metal implants or electronic devices they may have.

During the procedure:

- To conduct an MRI scan you will be positioned on a movable table.
- You will then slowly move into a tunnel like magnet of the MRI (Figure 4.3).
- Your radiologist will be sitting outside the MRI room and will be able to see you and communicate with you through a two way intercom.
- The entire examination is usually completed within 30 to 45 minutes.
- You will be asked to remain still during this examination. It is absolutely essential to remain steady while the images are being obtained.
- The part under investigation may feel a bit warm but if it is uncomfortable, painful or you feel any burning sensation inform your radiologist immediately.
- You will hear and feel loud tapping or thumping sounds while the images are being recorded. You may be provided with earplugs or headphones to help reduce the intensity of these sounds.

Post procedure precautions:

- You can resume your activities as soon as the examination is done.
- If any sedation is used you would be taken to the recovery room to rest. As the effect of the sedative wears out you may feel some nausea and vomiting sensation. But it should reduce within a couple of hours.

Interpretation of the results:

MRI scans are analyzed by the radiologists. MRI scans are normal in most people with ALS. However, the findings may show evidence of other disorders that may be causing the symptoms, such as brain/spinal cord tumor, disc herniation that compresses the spinal cord, syringomyelia (a cyst in the spinal cord), or cervical spondylosis (abnormal wear of the cervical spine).

Muscle biopsy:

Muscle biopsy is needed only rarely. But if the clinical presentation of ALS is atypical, then your physician may order a muscle/nerve biopsy.

What is muscle biopsy?

A muscle biopsy is a test which helps to diagnose neuromuscular diseases by viewing the muscle cell structure under microscope. Depending upon where the symptoms are observed a small piece of muscle tissue will be removed by needle or open biopsy. A needle biopsy involves inserting a thick needle into the muscle and removing it along with a small piece of muscle tissue in it. An open biopsy involves making a small cut in the skin up to the muscle and excising small amount of muscle tissue.

The procedure of muscle biopsy

Preparation for the procedure:

- Thorough assessment will be performed by your doctor.
- Your doctor will explain the procedure to you in detail.
- Inform your doctor if you have any allergies to medication or anesthetic agent.
- Inform your doctor of all the medications you are consuming.
- Your doctor may ask you to fast for sometime before the procedure.
- You may also be given mild sedative to help you relax.

During the procedure:

- You will be asked to change in to a hospital gown. The area from where the muscle is to be taken will be exposed and cleansed with an antiseptic solution. The doctor will inject anesthetic to numb the area. As the needle pricks you will feel a stinging and pricking pain for some time.
- Your doctor will insert the biopsy needle or make a small cut on your skin and remove a small piece of muscle. If the cut on the skin is too big your doctor may put some stitches or use adhesive tapes to seal the wound. A sterile bandage and dressing will be applied on the wound. The muscle tissue will be sent to the laboratory to be analyzed.

Post procedure precautions:

- Keep the area of biopsy clean and dry.
- Follow the bathing instructions given by your doctor accurately.
- Stitches will be removed in a few days. The adhesive tape will fall off, on its own. It is important to take care of the wound site to make sure it does not get infected.
- You may feel mild tenderness and pain at the site of biopsy for next 2 to 3 days. If pain is unbearable please talk to your doctor and take appropriate treatment for pain.
- In case of fever, redness, swelling, bleeding, oozing of pus or increased pain around the biopsy site notify your doctor immediately.
- Avoid any excessive physical exercises for 24 to 48 hours after biopsy.

Interpretation of the results:

The muscle biopsy can reveal whether there is presence of a neuropathic disorder (problem with nerves) or a myopathic disorder (problem within the muscle itself). In neuropathic disorders, denervation atrophy occurs with small angular fibers, grouped atrophic fibers. These features are not present in a myopathy. Typical myopathic

abnormalities include central nuclei, both small and large hypertrophic round fibers, split fibers, and degenerating and regenerating fibers.

Therefore, muscle biopsy may help in confirming the diagnosis or in ruling out other disorders.

Genetic testing:

In most of the cases ALS is not inherited. Whereas some of the cases are considered familial ALS (FALS). In these cases, more than one person in the family has ALS. In patients with FALS, symptoms often start at earlier ages than in non-familial ALS. FALS is most often autosomal dominant which means that a mother or a father who has a genetic change (or mutation that causes ALS), has a 50% chance of passing that mutation to each of her or his children. If a child does not inherit the gene mutation for FALS, they cannot pass it onto their children. Also, inheriting a gene for FALS does not guarantee that the child will develop symptoms of ALS. Although not always, but there will be someone in each generation with ALS and/or dementia. FALS can begin at different ages in different family members.

Therefore, if the patient has a family history of ALS, genetic testing may be ordered by your physician to confirm the diagnosis. Genetic testing is not used routinely for diagnosing ALS/MNDs. Genetic counseling is recommended before testing is ordered.

Genetic Counseling:

If there is more than one person with ALS and/or frontotemporal dementia in your family, you may have to undergo genetic counseling. A genetic counselor is an expert in genetic counseling, working closely with health care team who provides information and support to families who have genetic disorders, or are at risk of inherited conditions. During genetic counseling, the counselor takes a detailed family history, evaluates it, and discusses impact of genetic testing. However, genetic counseling does not always lead to genetic testing.

Procedure of testing:

The test involves taking a blood sample at your physician's clinic or in a lab.

Since FALS usually does not usually appear till later in life, genetic testing of children under 18 years of age is not usually recommended.

Interpretation of results:

Genetic testing is done to help determine the cause of FALS in a family. Superoxide dismutase (SOD1) is a gene located on chromosome number 21 and its function is to detoxify free radicals which are harmful to cells. Changes in this gene are responsible for abnormal functioning of this gene and leads to damage to motor neurons. This genetic mutation has been found in 20% of families with FALS.

The findings of the test and the results are analyzed and communicated by the genetic counselor or your doctor. Genetic mutation will be identified in only about 50% of individuals with FALS. However, if genetic mutation is not identified, it does not rule out diagnosis and risk of FALS.

If a mutation has been identified in one family member, genetic testing can be done for other members of family who do not have any symptoms to check if they have also inherited the genetic mutation. This is called predictive testing.

Genetic testing thus:

- explains if there is a genetic cause of ALS in the family.
- allows other family members to be tested to check if they also carry the genetic mutation.
- allows couples to plan on having children and to undergo prenatal testing.

Differential diagnosis:

Sometimes, presentation and clinical picture of some other neurological disorder may look like that of ALS/any type of MND. Therefore, it is necessary that a thorough clinical examination be done and other such disorders be excluded while diagnosing. Following are some conditions that could be considered in the differential diagnosis of various MND.

D/D for ALS:

- Other neurodegenerative disorders - primary lateral sclerosis (PLS), hereditary spastic paraparesis (HSP), spinal muscular atrophy (SMA), and X-linked spinobulbar muscular atrophy (SBMA).
- Peripheral neuropathies - multifocal motor neuropathy, acquired immune-mediated demyelinating neuropathy that responds to treatment with intravenous immunoglobulin (IVIg).
- Myopathies
- Postpolio syndrome (PPS)
- Monomelic amyotrophy (Hirayama disease) - young males in the second and third decades of life are mainly affected, the onset is insidious, and predominantly unilateral upper-extremity weakness and atrophy occur with no sensory or UMN symptoms.

D/D for Primary Lateral Sclerosis:

- Structural spinal cord disorders - cervical spondylotic myelopathy, Arnold-Chiari malformation, or tumors

- Pure, uncomplicated HSP
- Metabolic disorders - vitamin B-12 deficiency
- Viral infections - tropical spastic paraparesis (human T-lymphotropic virus type I) or HIV infection
- Primary progressive multiple sclerosis (PPMS)

D/D for Hereditary Spastic Paresis:

- Dopa-responsive dystonia and Arginase deficiency - both the disorders are treatable.

D/D for Pseudobulbar Palsy:

- Brown-Vialetto-Van Laere syndrome and Fazio-Londe disease
- Madras motor neuron disease (MMND)
- Boltshauser syndrome

D/D for Spinal Muscle Atrophy:

- Congenital muscular dystrophy
- Congenital myopathy
- Congenital neuropathy
- Disorders of carbohydrate metabolism
- Myasthenia gravis
- ALS (especially juvenile forms)
- Limb-girdle muscular dystrophy - For SMA types III and IV

D/D for Post Polio Syndrome:

PPS is a diagnosis of exclusion, i.e complete medical history and clinical examination will help exclude other conditions such as ALS, multiple sclerosis, myasthenia gravis, inclusion body myopathy, infectious myopathy, collagen vascular diseases, and hypothyroidism.

Tests like PET Scan (whole Body), Blood Test, Urine Test', CSF testing may be advised by your doctor for ruling out other diagnoses.

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Chapter 5

What are the common complications of motor neuron disease?

Symptoms of motor neuron disease (MND) give rise to certain secondary complications which aggravate the existing disease condition. The complications are independent of the disease pathology. Most often these complications can be avoided or delayed with adequate care of the patient. It is important for the patients with MND to know the complications of the disease so that these can be prevented. This chapter will also guide the patients and caregivers to detect and treat the complications at an early stage. People with MND experience complications as the disease progresses. The complications are seen in various body systems like Respiratory system, Musculoskeletal system, Nervous system, Gastrointestinal system and cognition and behavior.

Some of the common complications of MND are:

1. Breathing problems (respiratory complications) related to bulbar dysfunction
2. Cognitive and Behavioural changes
3. Thrombophlebitis/ Deep vein thrombosis (blood clot that develops in a vein deep in the body)
4. Contractures
5. Joint pain
6. Joint subluxation / Dislocation
7. Fractures
8. Constipation
9. Anxiety related to difficulty of breathing

Respiratory (breathing) problems related to bulbar dysfunction:

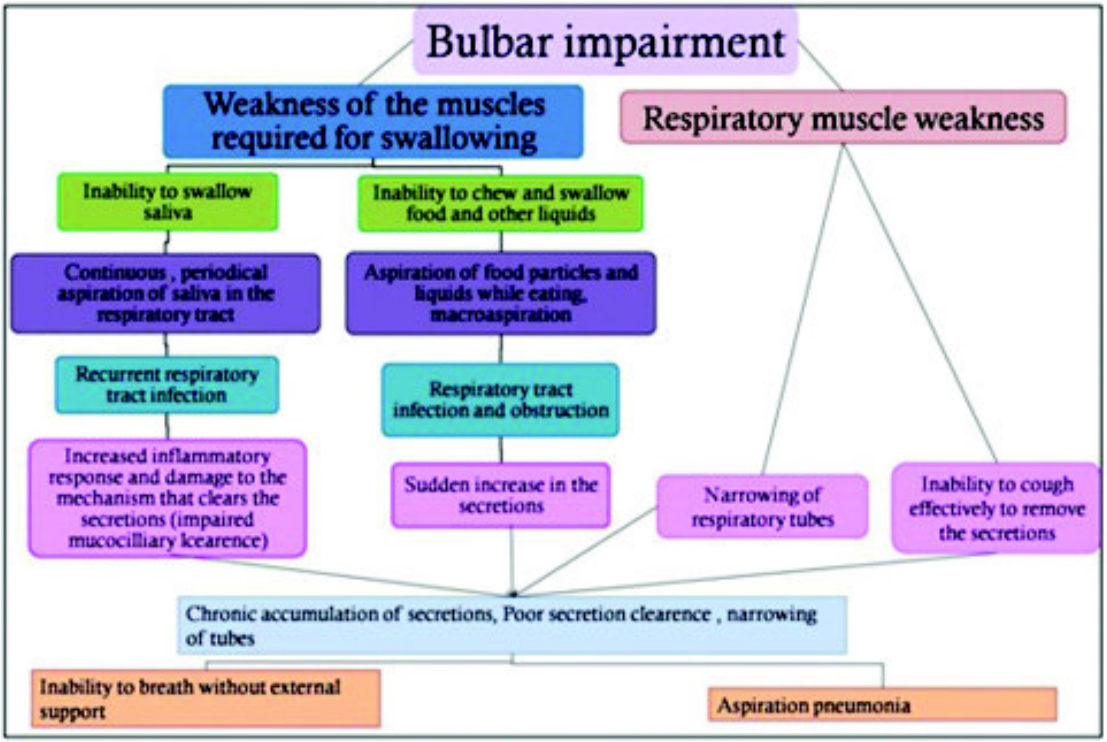


Figure 5.1: Respiratory problems due to muscle weakness

The gradual deterioration of muscles required for breathing results into breathing difficulties. The respiratory muscle weakness may increase the fatigue levels and deplete the energy. The most common complications resulting from respiratory muscle weakness and weakness of the muscles responsible or swallowing and speech (bulbar dysfunction) is respiratory problems, malnutrition and dehydration.

Respiratory problems:

Most common respiratory complications are, Aspiration pneumonia and Respiratory distress.

Aspiration Pneumonia -

When solid food or liquids enter into the upper airway instead of esophagus and stomach it is called aspiration. Such accumulation can lead to upper airway obstruction as well as infection of the respiratory tract. Aspiration over prolonged time can lead to severe infections like Pneumonia which can be fatal. The obstruction and infection causes further decline of breathing capacity and increases respiratory distress.

Respiratory distress:

Respiratory distress is characterized by inability to breathe. There are multiple reasons for developing respiratory distress in MND. These reasons can be categorized as narrowing of the respiratory tubes, accumulation of secretions in the respiratory tubes and respiratory muscles weakness.

Due to bulbar dysfunction, muscles of swallowing and breathing undergo weakness. Due to the weakness of swallowing muscles; patients are unable to swallow the saliva and there is continuous aspiration of small amount of saliva in the respiratory tract, microaspiration. Patients are Also unable to swallow food and liquids during the day and an attempt to do so may lead to sudden aspiration of larger amounts of food and liquids in the respiratory tract, macroaspiration.

Because of prolonged microaspiration there are frequent infections and chronic inflammation of respiratory tract. Due to the inflammation ability of the respiratory tract to clear the secretions reduces. In addition there is weakness of respiratory muscles and therefore the ability to cough and remove the secretion also reduces. With macroaspirations and infection the secretions in the respiratory tract increase. Inability to clear the secretions and increased secretions causes blockage and narrowing of the respiratory tract.

Obstruction of the respiratory tract is also triggered by bulbar dysfunction due to weakness of the muscles of the tract connecting nose and throat (laryngeal muscles) and the tract connecting mouth and throat (Pharyngeal muscles). These tracts are like a tube and the tone of the muscles holds the tube open as the muscles undergo weakness and the tube starts collapsing on its own. This gives rise to a feeling of chocking and breathlessness. The obstruction of the airways makes it difficult for the air to flow through and increases the resistance to airflow. Because of the increased resistance the respiratory muscles have to act harder to perform their function and difficulty of breathing increases even more.

It is a vicious circle of events that eventually leads complete inability to breathe without an external support and causes respiratory distress.

Malnutrition and Dehydration:

MND weakens the muscles of lips, tongue, palate and mastication muscles (bulbar muscle weakness); this can cause chewing and swallowing difficulty (dysphagia) and also nutritional deficiency (malnutrition). Chocking and drooling is common due to inability to swallow saliva and it increases while having food or drinking water. Chocking sensation is caused as the food gets stuck in the food pipe (Oesophagus) and blocks the wind pipe (Trachea). As the disease progresses, the severity of swallowing difficulty increases. Inability to swallow leads to poor food intake and increased drooling upon swallowing causes lack of interest in eating. Poor food intake and liquid intake

causes malnutrition and dehydration. Dehydration can cause symptoms like cramps, dry mouth, headache and malnutrition contributes to loss of energy and body mass.

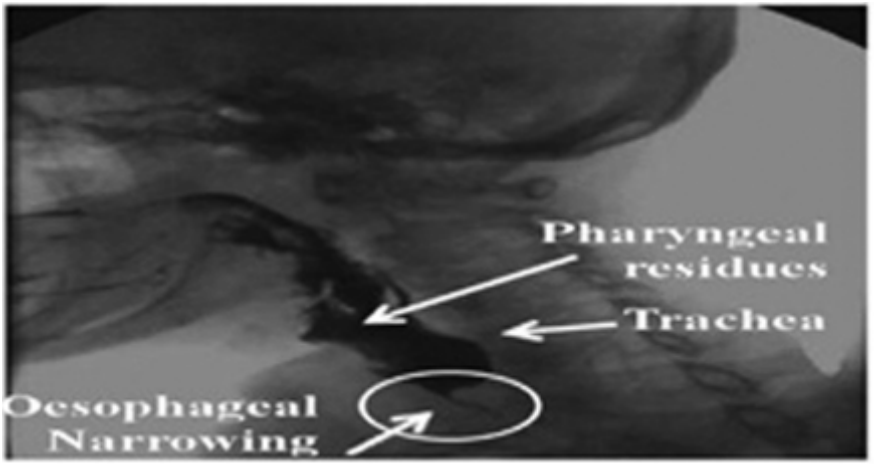


Figure 5.2: Narrowing of oesophageal opening and pharyngeal residues leading to choking

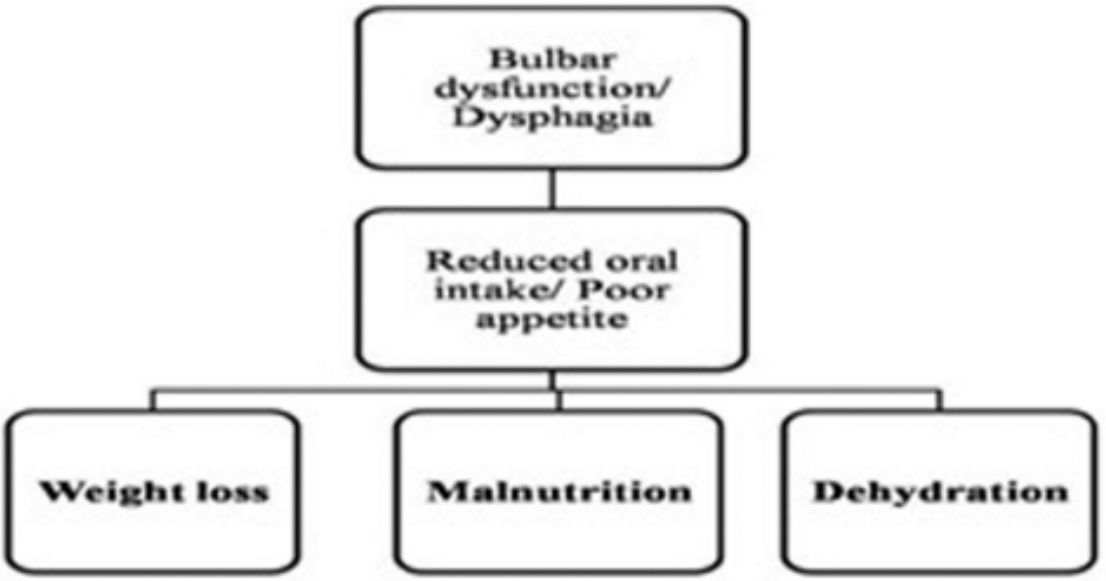


Figure 5.3 Complications of bulbar dysfunction

Cognitive and Behavioural changes:

Patients with MND experience problems with memory and making decisions. Eventually, some patients are diagnosed with the form of dementia (a chronic or persistent disorder

of the mental processes marked by memory disorders, personality changes, and impaired reasoning) which is called as frontotemporal dementia (FTD).

MND patients are also affected with two discrete conditions that are depression and emotional liability (inability to control emotions is uncontrollable crying or laughing or other emotional displays, which is Also called as pseudo bulbar effect). Patients may be depressed after getting diagnosed with MND and facing difficulties to perform activities of day to day life. Depression may be present due to increased difficulty in communicating for their problems as well as physical changes and progression of the disease.

Musculoskeletal complications:

Contractures:

Another most common complication in MND is contractures. Contracture means tightness of muscle that leads to shortening of the muscles and thereby restricts the movement at a joint. Patients with MND are bed ridden, so the full range of joint movement restrictions, mobility limitations, inability to perform ADL's results in contractures. Because of static positioning of the limbs, the muscles form adhesions between other structures and within the muscle fibers as well. The adhesions restrict the movement. If adhesions are present for a prolonged period then those become permanent. Contracture formation is also a consequence of imbalance between muscles.

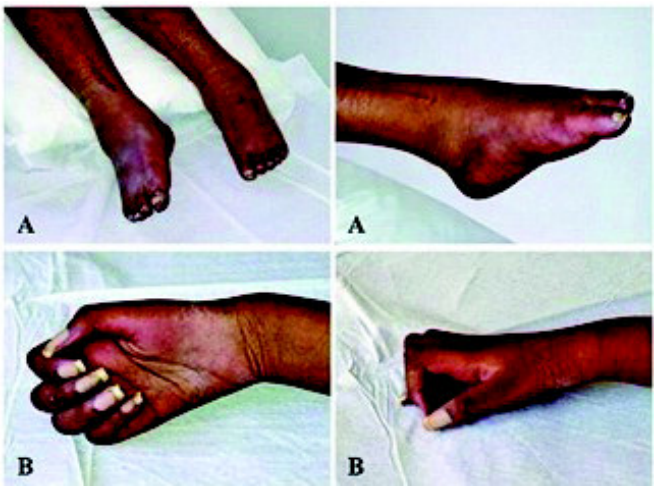


Figure 5.4: contractures

Joint pain:

Lack of active movement and muscle weakness increased the compressive forces on the joints. Active and strong muscles help to hold the surfaces of two bones in a joint apart from each other. In absence of these the surfaces come in close proximity and rub

onto each other causing pain. Regular movement of the joints passively will reduce such pain.

Subluxation:

A joint is covered by a capsule of tissues. This capsule holds the joint in place but is unable to protect the joint from the stress and strain of movement. This is usually the work of muscles. But as the muscles undergo weakness those are unable to maintain the joint integrity. This causes the bones in the joints to displace out of the joint cavity, subluxation. This may lead to pain while performing active as well as passive movements.Most common site of subluxation is shoulder as the gravity exerts a continuous pull on the arms.

Fractures:

Patients of MND are at a risk of frequent falls. Falls can cause fractures of different bones in the body. It is therefore extremely important to prevent such falls and prevent injuries and fractures.

Thrombophlebitis/ Deep vein thrombosis (blood clot that develops in a vein deep in the body):

Thrombophlebitis/ Deep vein thrombosis DVT are probable complications for all immobilized MND patients. During the early stages of DVT, the dangers are particularly high when the blood flow in the veins becomes sluggish due to bed rest, decreased activity and limb paralysis. Patient experience tenderness (a state of unusual sensitivity to touch or pressure), dull ache, rapid onset of one leg swelling. Early diagnosis and treatment of acute DVT are necessary to reduce the risk of fatal pulmonary embolism. If there is any unexplained and sudden swelling of extremities with tenderness it is advisable to contact the doctor immediately.

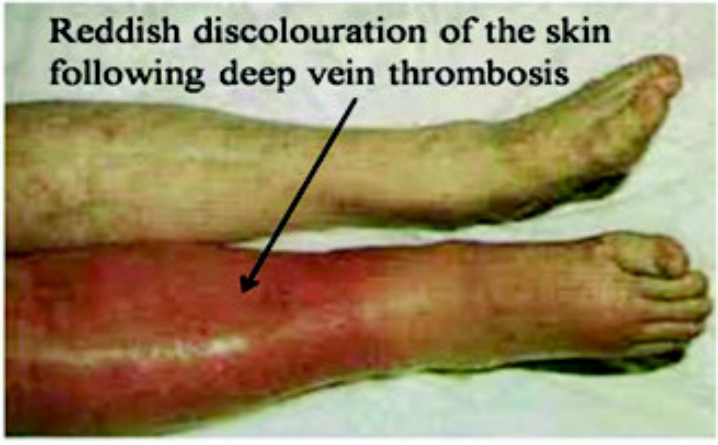


Figure 5.5: Deep vein thrombosis

Chapter 6

How ALS can be managed and who can help?

ALS is a persistent progressive degenerative disease leading to increasing numbers and severity of impairments. ALS leads to multiple changes in the body function like muscle weakness, fasciculations, fatigue, progressive activity limitations, dysphagia and dysarthria.

The individual's ability to perform the activities suddenly gets limited because of the multiple impairments and becomes very difficult to cope up with such situations. In due course of time as the disease progresses the body becomes weak and becomes unable to perform the regular work load. As ALS has no known cure and has minimal effective disease slowing treatments the treatment focuses on restoring the available function and preventing the progression how much ever possible. Due to the varied symptoms of the disease approaching and managing all the symptoms are very much important for the quality care. The progressive nature of the disease and continually changing status of the individual a comprehensive and multidisciplinary approach to care is advantageous.

Currently the management of ALS focuses on

- maintenance of safe mobility and independence in functioning, to include mobility for patient and caregiver;
- restoration of maximal muscle strength and endurance within limits imposed by ALS;
- management of dysarthria (speech problems) and dysphagia (swallowing difficulties);
- eliminating or preventing pain, spasm and fasciculations
- prevention and minimization of secondary complications of the disease;
- energy conservation techniques and respiratory comfort;
- prescription of assistive aids and appliance needed to include mobility, self-help and feeding devices, augmentative communication units, and hygiene equipment that supports both patient and caregiver;
- care at advanced stage for PEG;
- palliative, nursing and nutritional care.

For the foundation of the successful management of the above goals for the individuals with ALS a multidisciplinary team has to work in consistent and complete care approach. (Figure6.1) No one person could possibly have the knowledge and skills necessary to manage all aspects of ALS. A multidisciplinary team approach is essential in providing

the most comprehensive care that will lead to maximize independence and QOL. All individual members in the team collaborate, contributing their expertise in specialized area, thereby enhancing the team's overall effectiveness. The different members in the team work through sharing their skills and finding about the disease status to each others. The multidisciplinary care team includes:

An individual with ALS

The management of the disease should be based on the expertise of the multidisciplinary team as well as should focus the needs and goals of the individual. An individual with ALS at the initial stages will have different needs than at later stages when mobility level is markedly reduced. The individual should also know that the goals will have to be adjusted and plans should be reset as the disease process continues. The goal at all stages is to optimize health and increase the quality of life.

Figure- 6.1 Management of ALS/MND

Family, caregiver andcommunity

The family should be introduced to their role in the course of the disease. Familial role may change as the individual who previously used to take care of the children may be at the receiving end. The individual, family and care giver are the most important member of the team.The caregiver education is essential component of care and it should begin immediately with individual and family. The majority of people are unaware of the disease. It is the job of the multidisciplinary team to explain the affects of the disease with prognosis and how to keep their positive attitude towards the individual.

Physician

Physician is the first health professional whom the individual consults. The role of the physician is to guide you towards the other essential treatment approaches. He can be your family doctor as well and can be one of the best counselors throughout the course of the disease.

Neurologist

Neurologist is expertise in the realm of nervous system and has better knowledge about recovery and impairments through the disease course. He can diagnose the disease and prescribe the best suited drugs to ameliorate the symptoms of the disease.

Physical therapist

A physical therapist's skills lie in field of restoring range of motion, mobility and normalizing muscle tone. They can assist you in choosing devices that can help with mobility. In the advanced stages of the disease, when the patient is bedridden, interventions to effectively control pain, they can manage contracture and respiratory infection development.

Fig 6.2

Occupational therapist

An Occupational therapist will facilitate the diminished ability to perform activities and upper limb functional loss. They train to maintain the activities of daily living and hand function. A valuable management approach is co-treatment of individual with

the physical therapy and occupational therapy, which can be very productive for retraining. They educate the caregiver on the best ways to assist the individual. They can help you in adapting your home environment to meet your varying needs.

Speech pathologist

A speech pathologist can help to make speech and swallowing easier. They can develop and give advice for different/ alternate ways of communication. They can guide the team about the impairment and the alternate way to interact with the individual.

Psychologist / Psychiatrist

A Psychologist / Psychiatrist helps in developing a behavioral management program and can counsel for different emotional and coping response for the disease. They can also help with cognitive training and problem solving approach in the advanced stage of the disease.

Fig 6.3

Physiatrist

A Physiatrist helps in the coordination in the training of physical medicine and he communicates with other team members and ensures that the rehabilitation is effective and being provided is truly team oriented.

Orthotist

Orthotist will help in manufacturing the prescribed orthotics or assistive aids and appliances, which suits the individual and improves his independence maximally.

Nutritionist

A nutritionist helps in providing diet management and counseling throughout the course of the disease. They keep a track of nutritional and hydration status and advice diet modification for dysphagia and Percutaneous endoscopic gastrotomy (PEG).

Nurse

A Nurse will be responsible in dispensing medicines and adequate nursing care in the advanced stage and when patient is in hospital or hospice. The nurse has the maximum communication with the patient and caregiver.

Medical social worker

A medical social worker will support and counsel with you and your family on how to cope with the disease and will identify additional resources to help you make the needed changes. They will also provide information about transitions related to work (vocational rehabilitation and employment).

Intensivist

An intensivist helps the individual with monitoring during the advanced critical care and ventilatory and PEG management.

Associations and Resources

Associations will help the individual to and provide support for individuals and their families. They can provide a stabilizing influence to focus on healthy behaviours, coping skills and effective self care management. Resources can aid in providing the health care facilities, equipment loan programs.

Day care center / Hospice

Day care center can be a very useful for individuals whose family is working. In day care center the individuals can stay from morning to evening when they are alone at house and can receive a multidisciplinary care. This will help to get the best care and a chance to stay with the family, which will help the individual to be emotionally sound.

Hospice care is a type of care that focuses on severely ill patient's pain and symptoms, and attending to their emotional and spiritual needs.

These care centers involve assistance for patients' families to help them cope with the course of the disease and provide care and support to keep the patient at home. Groups of similar patients staying together will motivate the patients and will help in performing rehabilitation easily.

The multidisciplinary care should be delivered to individuals and families in time to make thoughtful decisions rather than just before a time of crisis, such as after a choking

episode or during a respiratory arrest. The team should remember that they should plan on spending enough time with the family to respond to concerns and help with.

Advantages of visiting a multidisciplinary clinic

- Can help people with ALS get the best possible care.
- Possible to live longer and with some relief from symptoms.
- Available treatments can make daily life easier and more comfortable.
- Meet with several specialists in the field of ALS, each of whom will focus on a particular area of care if you eventually need it.
- Several therapies are now available to make daily life easier and more comfortable.
- Many services and resources that can help to meet the needs of people with ALS

It is important and always better to detect and manage the problems as early as possible. Evidence and research stated multidisciplinary approach can improve the quality of life and independence for a longer period of time and prolong their survival.

SECTION B
Multidisciplinary Care

Lou Gehrig

June 19, 1903 - June 2, 1941

Henry Louis **"Lou"** or **"Buster"**Gehrig (Born Heinrich Ludwig Gehrig;) was an American baseball first baseman who played 17 seasons in Major League Baseball (MLB) for the New York Yankees, from 1923 through 1939. Gehrig's streak ended on May 2, 1939, when he voluntarily took himself out of the lineup to stunned fans after his play was hampered by amyotrophic lateral sclerosis (ALS). MND/ ALS came into focus of the world when famous base ball player Lou Gehrig was diagnosed with ALS .The pathos of his farewell from baseball was capped off by his iconic 1939 "Luckiest Man on the Face of the Earth" speech at Yankee Stadium. A monument in Gehrig's honor currently resides in Monument Park at Yankee Stadium. The Lou Gehrig Memorial Award is given annually to the MLB player best exhibiting his integrity and character.

Chapter 7

Muscle weakness

Don't just strengthen your muscles but also your mind.

As the motor neurons stops ending messages to muscles, muscle weakness begins to set in. Gradually muscle group start becoming weaker leading to weakness and inability to move the joints affecting the mobility. As the joint movement is limited associated muscles become stiff and tight.

Once the motor neurons have degenerated which control a particular muscle it is irreversible and muscle cannot be regenerated by exercise reversed but early exercise therapy or intervention can help keep weakened muscles as strong as possible as regular

Exercise can help in maintaining muscle elasticity, range of motion ,prevention of muscle shortening and endurance.

Treatment/Management

The natural course of the disease(MND)can consist of several stages. There is no clear, presentation of how the speed of progression will be and it can vary from patient to patient. To assist rehab specialists to plan treatment programmers' along with thorough assessment of the individual problems this includes MMT (manual muscle testing)and needs of the patient. There is evidence that exercise is beneficial in maintaining function. The overall goals of intervention will vary as the condition progresses.

Exercises are important for

1. To maintain mobility as long as possible.
2. Prevents joints from getting stiff.
3. Helps strengthening muscle groups which are not yet affected as it can help to compensate for those weakened or wasted muscle.
4. Active muscle movement promotes circulation.
5. Improve cardio respiratory endurance.
6. Improves stamina and decrease fatigue level.
7. Decreases pain, cramps and fasciculation.

Maintenance of strength and endurance requires daily activity and repetitive muscle contractions.

Keep patient physically independent as long as possible through pleasurable activities such as walking.

Exercises can be done based on the patient's ability and muscle strength to perform.

Depending on how the exercises are performed they are divided in 3 types:

1. Active:

Where the person performs the exercise on his or her own which includes activities like cycling, jogging, swimming, bed mobility, activities of daily living and general range of motion and strengthening exercises.(Fig 7.1- 7.42)

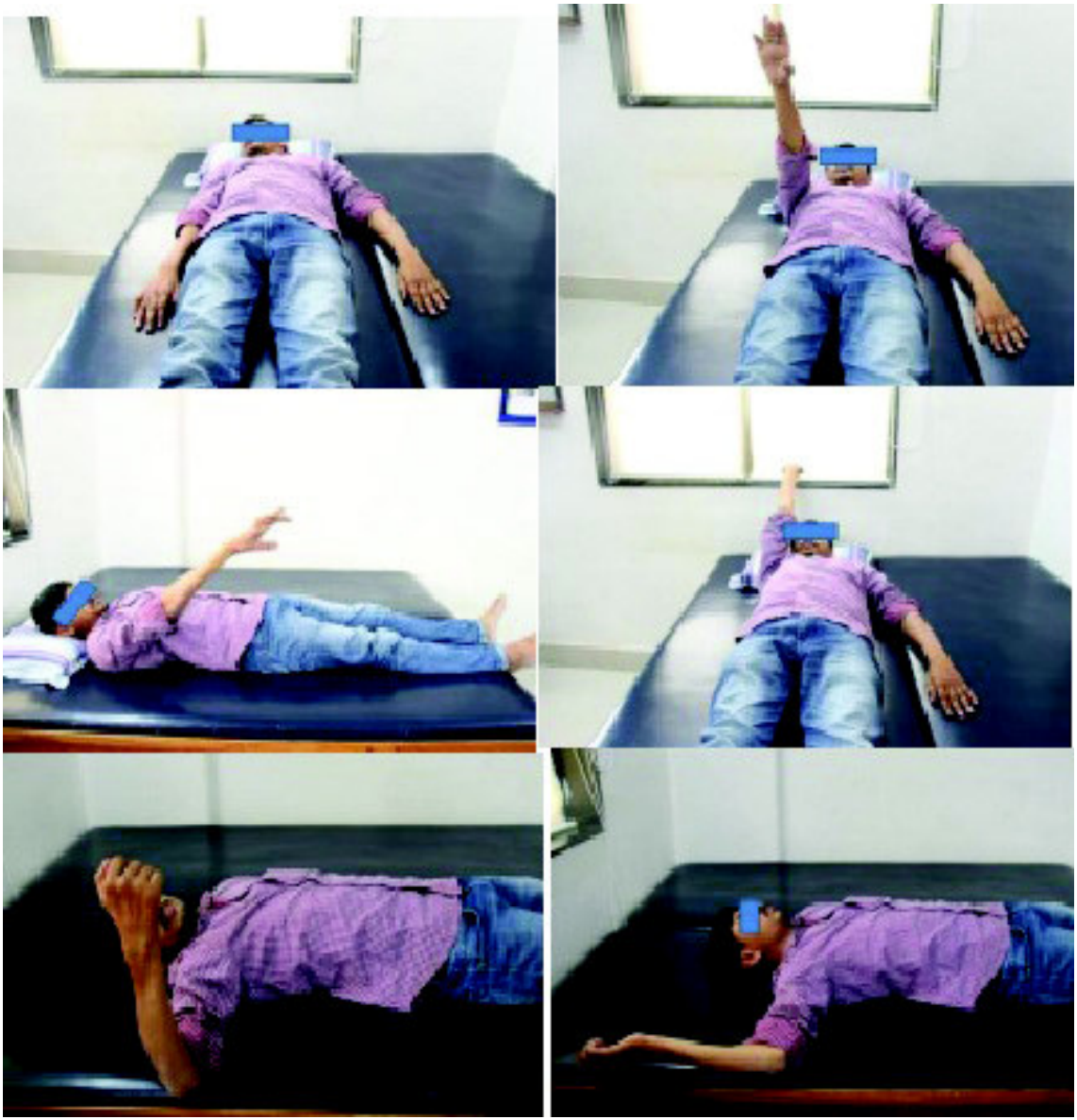


Fig7.1-7.12: Active exercises for upper extremities

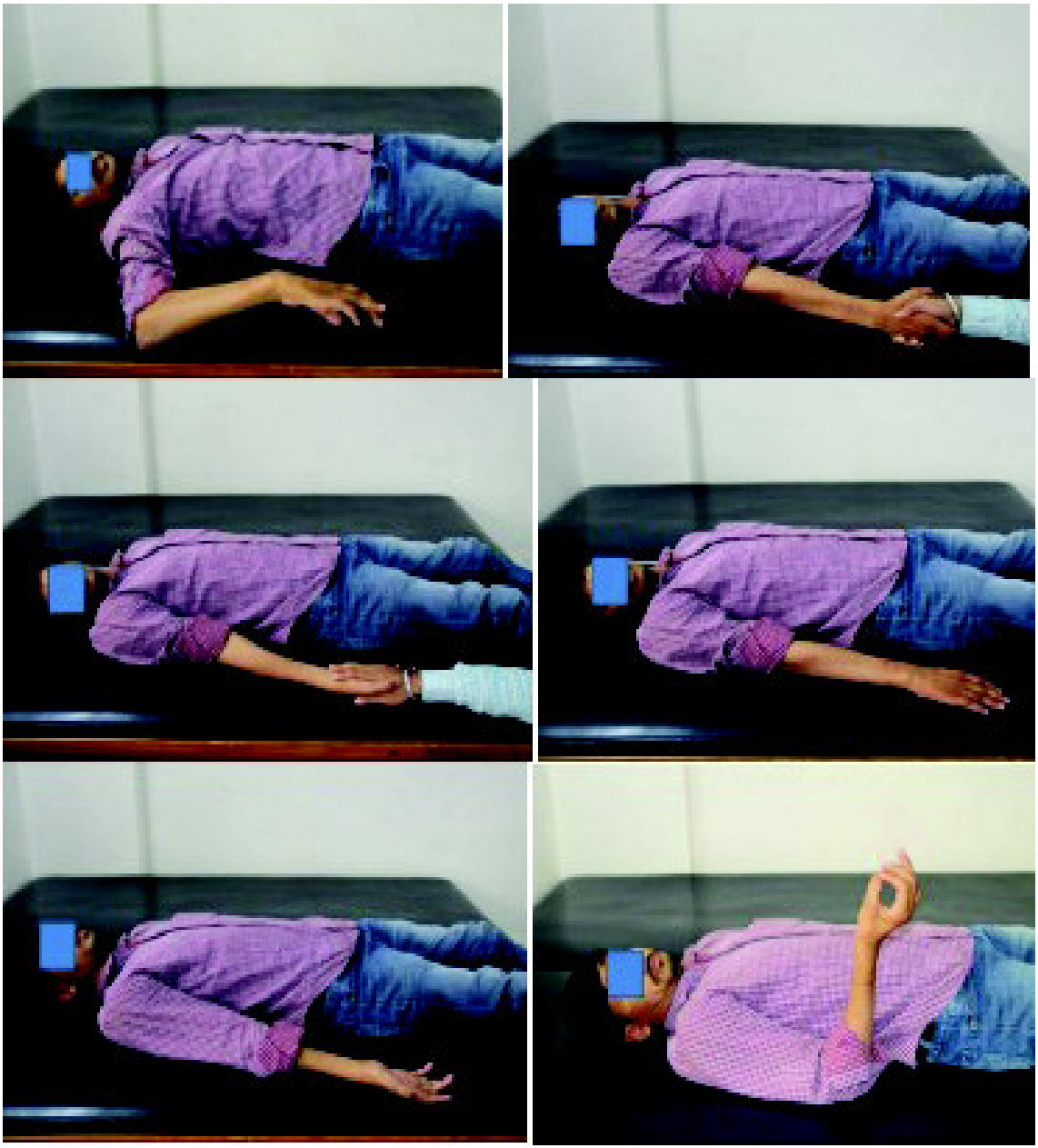


Fig7.1-7.12: Active exercises for upper extremities

Fig7.13-7.28: Active exercises for lower extremities

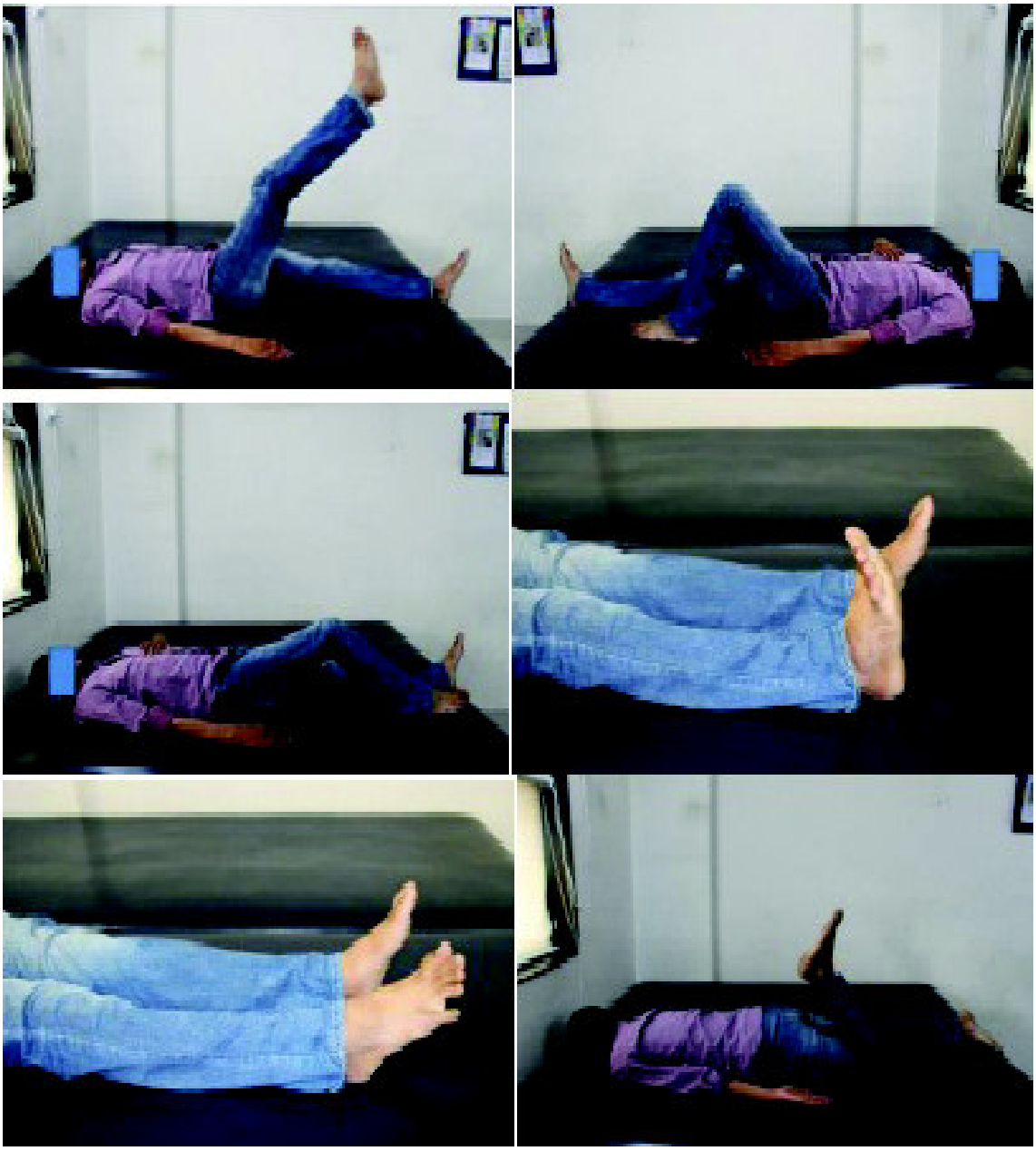


Fig7.13-7.28: Active exercises for lower extremities

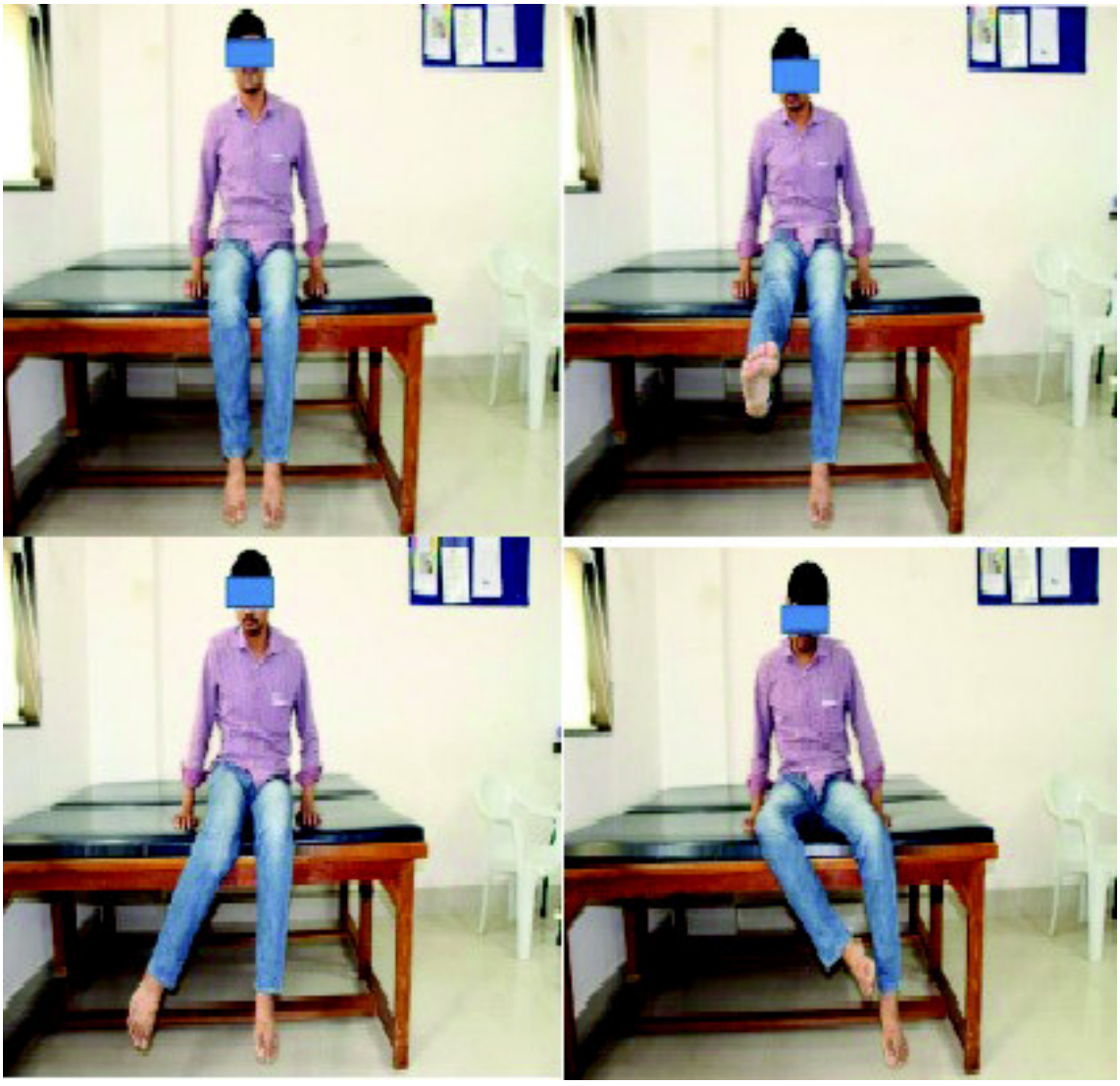


Fig7.13-7.28: Active exercises for lower extremities

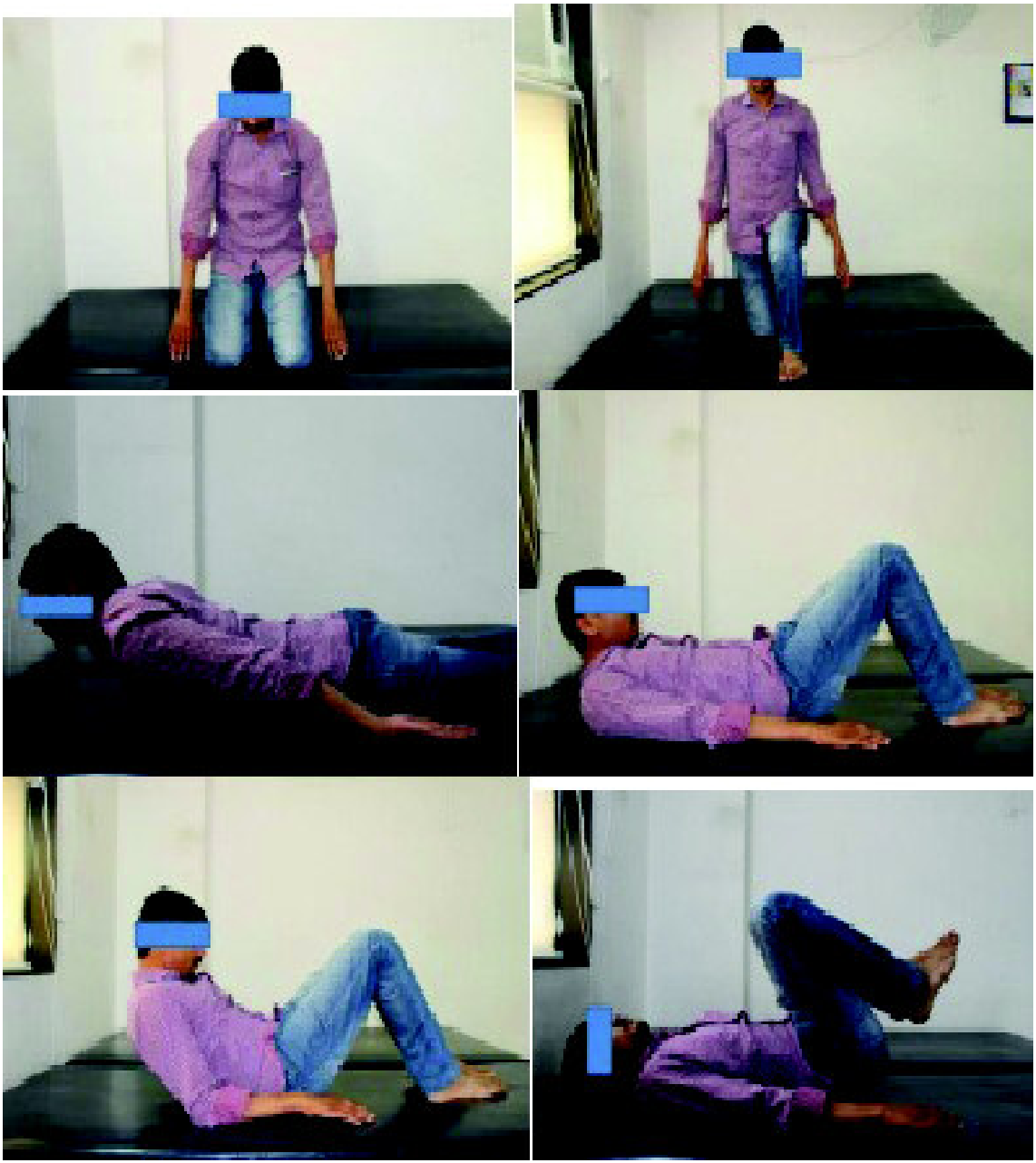


Fig7.29-7.37: Active exercises for trunk

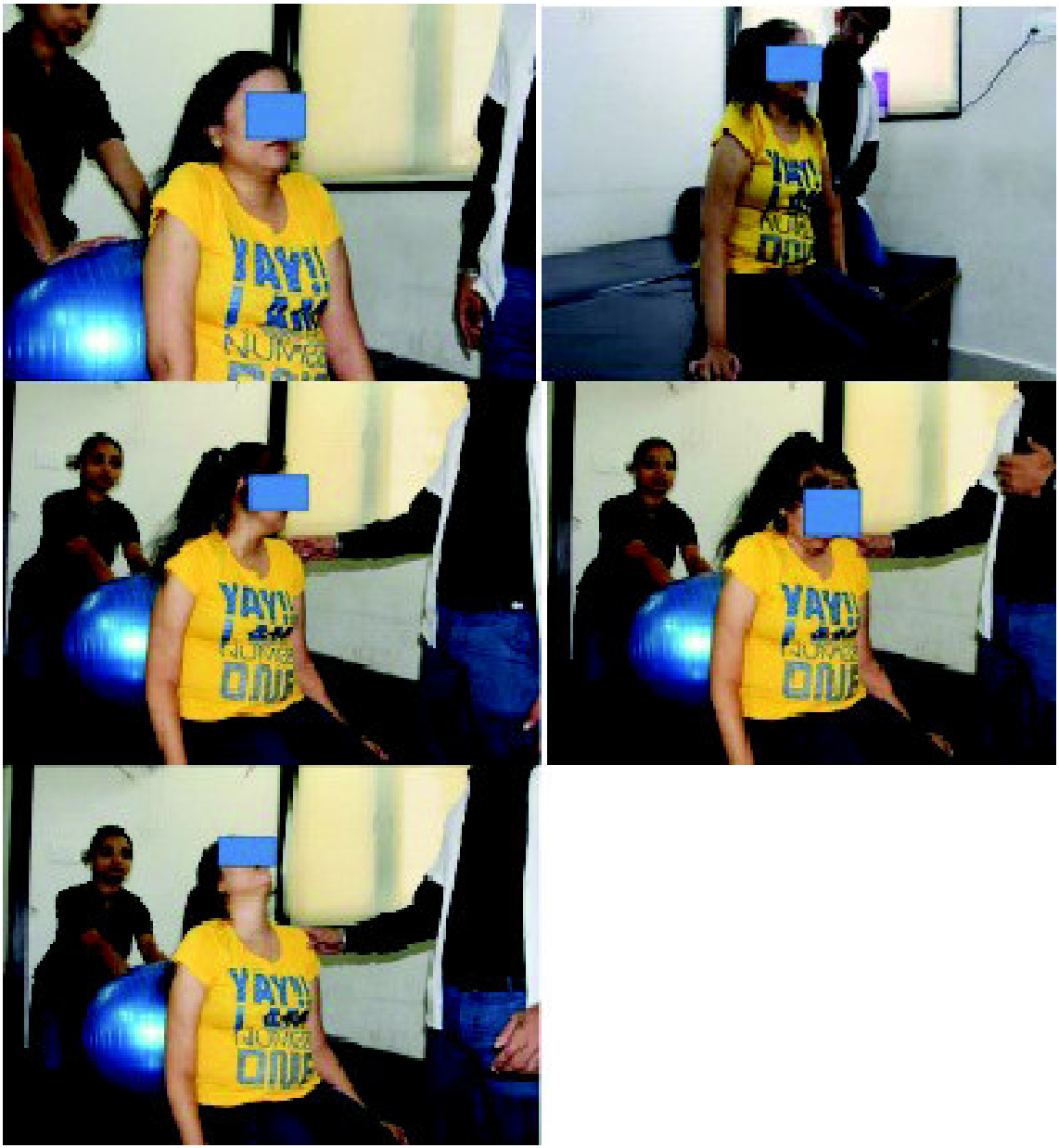


Fig7.38-7.42: Active exercises for neck

2. Active assisted:

Where the person performs some of the exercise by his own but with the help from others which includes assistance by the therapist or caregiver, using suspension or mechanical aids like assistive device etc. Suspension therapy can be of great use in improving muscle weakness as it provides assistance which will help to complete the ROM.(Fig 7.43- 7.53)

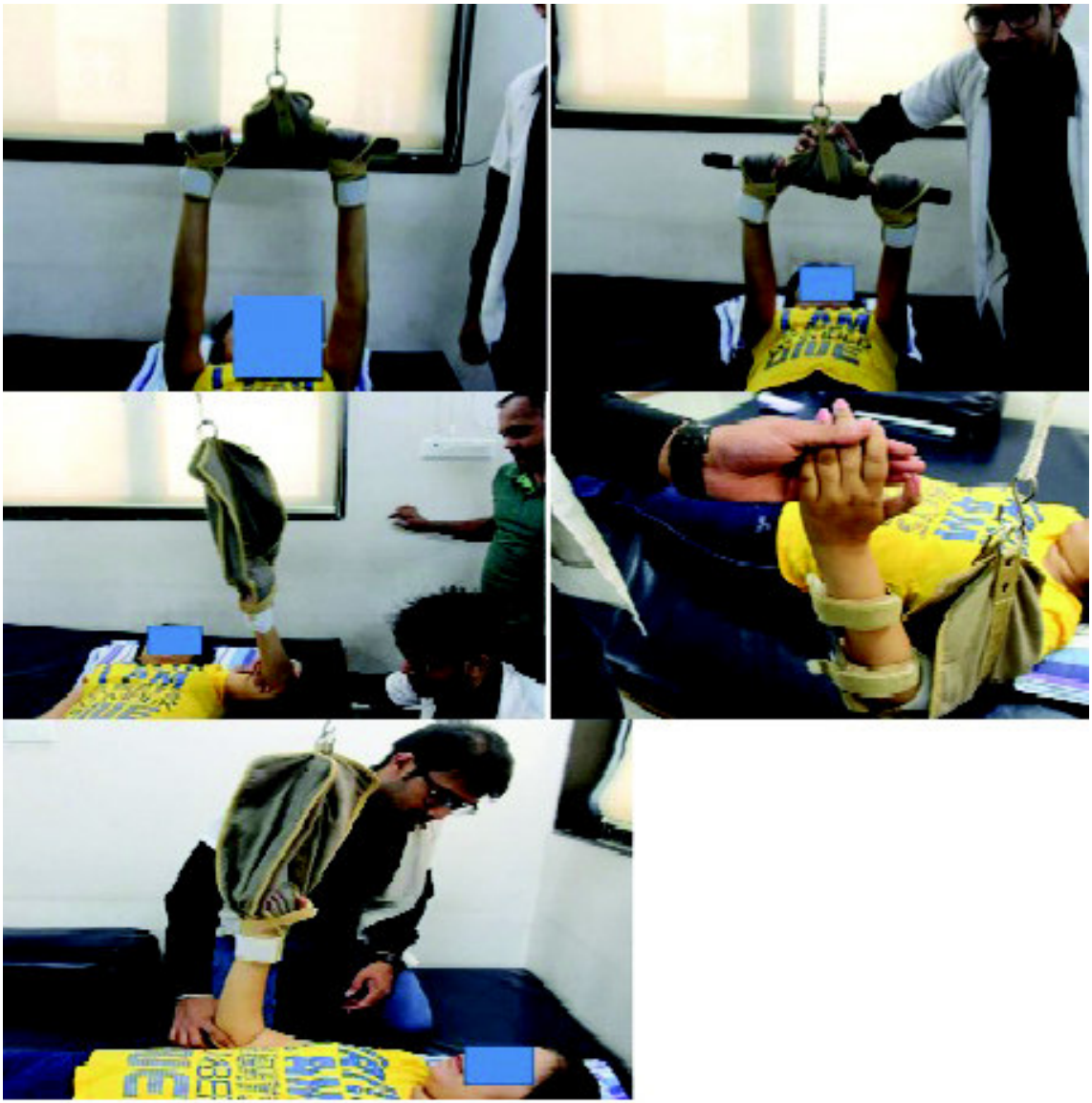


Fig7.43-7.53: Active assistedexercises using suspensionandspring

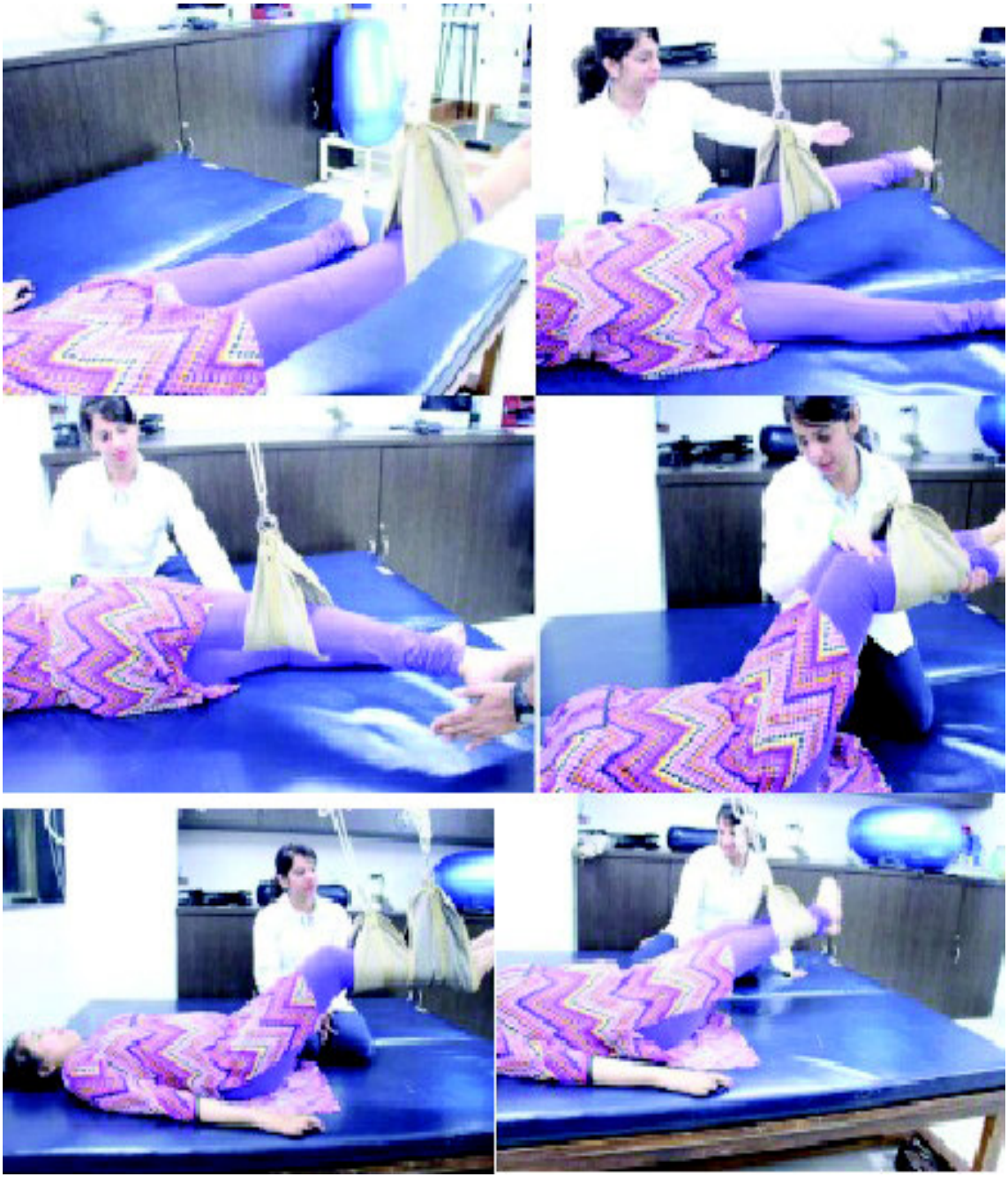


Fig7.43-7.53: Active assistedexercises using suspensionandspring

3. Passive:

Where the exercise is entirely done by some other person or using a modality. The main aim of passive exercises is to maintain ROM and joint flexibility. It improves the blood circulation and avoids tightness and contracture of muscles. It can be performed when it is difficult for the individual to do it by them. The caregiver or the family can perform and learn to perform these exercises by a physical therapist.(Fig 7.54-7.65)

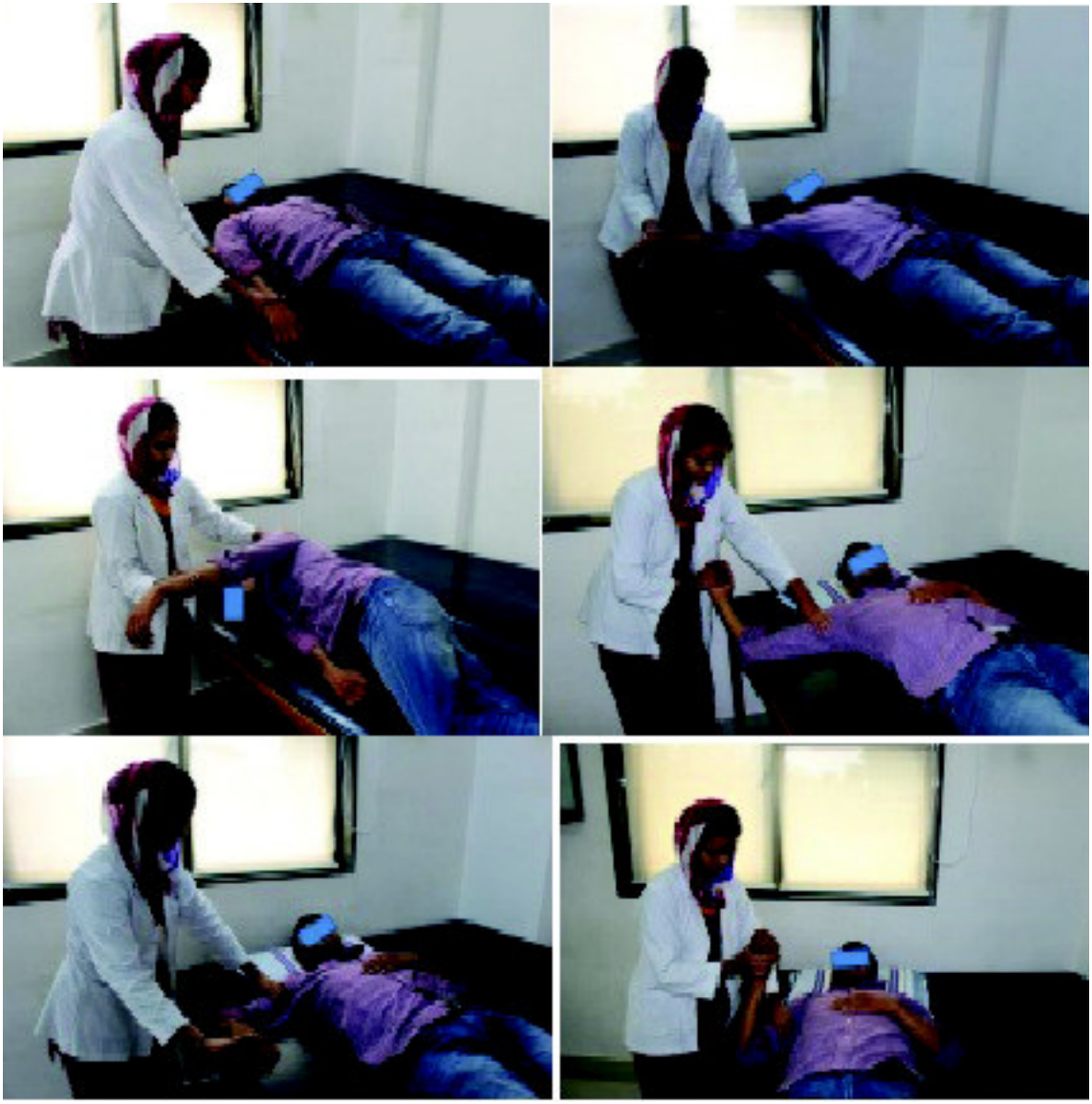


Fig7.54-7.65: Passive exercises

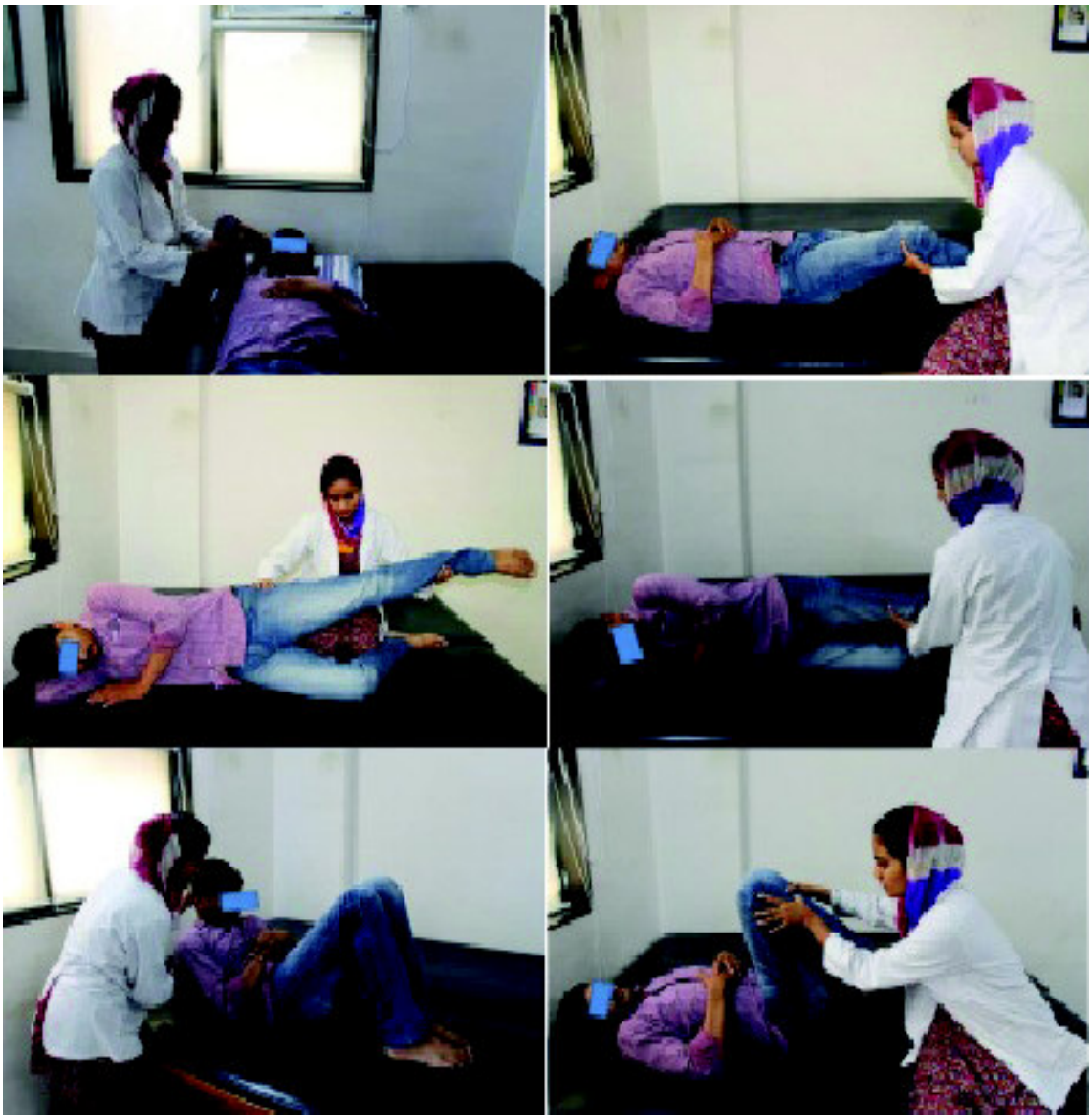


Fig7.54-7.65: Passive exercises

Mild resistance exercises

Mild resistance exercises using theraband, theratubes, springs, pulleys can be used if the patient has normal strength.

Proprioceptive neuromuscular facilitation exercises

PNF can be used for relieving spasticity as well as increasing the range of motion. It consists of diagonal movement of extremity and can help to treat whole body.

It can facilitate as well as inhibit the tone. The exercises can be reinforced with verbal commands, stretch, timing of emphasis etc.

Exercises should be

- Continue normal activities or increase activities if sedentary to prevent disuse atrophy.
- Begin program of range-of-motion (ROM) exercises.
- Add strengthening program of gentle resistance exercises to all musculature with caution not to cause over work fatigue.
- Continue cautious strengthening of muscles based on manual muscle testing(MMT) grades

Patients should exercise for short periods several times a day rather than attempting to exercise all muscle groups in one session.

Group activities

Group activities for strengthening can be highly advantageous as it will motivate the patients with all the peer members and the patient will never be bored of doing similar exercises.

All the above exercises can be taught to the patient by the physical therapist and can prescribe them the do sage according to the needs. After which it can be practiced by the patient at home.

It is important to know that exercise will not strengthen muscles that have already been weakened by MND. Once the supply of motor neurons that control a particular muscle has degenerated, it cannot be regenerated by exercise.

Chapter 8

Alteration in muscle tone

Loosen your tight muscles and tighten your loose muscle!!!

Tone is the resistance of muscle to passive movement of any joint. Alteration in muscle tone is functionally limiting and contributes to the development of other complications such as tigthness, contracture, postural deformities and pressure sores. The management of muscle tone can be a key to remaining mobile for as long as possible. The tone may alter based on the lesion.

Lower motor neuron lesion

- flaccidity / hypotonia / decreased toneinthe muscle
- muscleswill berelaxedandloose
- passive limb movements are veryeasy

Initial muscle weakness usually due to decrease tone occurs in isolated muscles most often in the hands and foot followed by proximal weakness. Muscle weakness leads to decrease ROM , predisposing to joint subluxation, muscle and tendon shortening, joint contracture. It also may lead to postural abnormalities, de-conditioning, balance instability and walking difficulties.

Treatment

As the weakness gradually progresses , the tone of the muscles start decreasing and the joint becomes hypermobile. To alleviate these symptoms a regular management to normalize the tone is required before starting the exercises. The below techniques should be followed.

- Quickstretch to the muscles whichhave lowtone(fig 8.1,8.2)

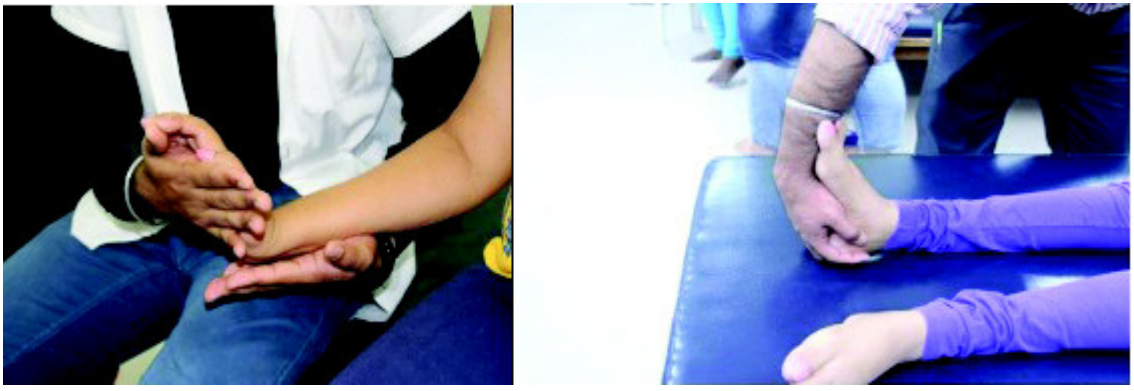


Fig8.1& 8.2: Quick stretch to the muscles whichhave low tone

- Weight bearing on the jointcan help to buildthe tone in these positions (fig 8.3)
  - prone on elbows
  - sitting
  - quadruped
  - kneeling
  - standing

Fig 8.3: Weightbearingonthe jointto developtone

- Joint compressions to all joint can facilitate the muscle tone(fig 8.4-6)

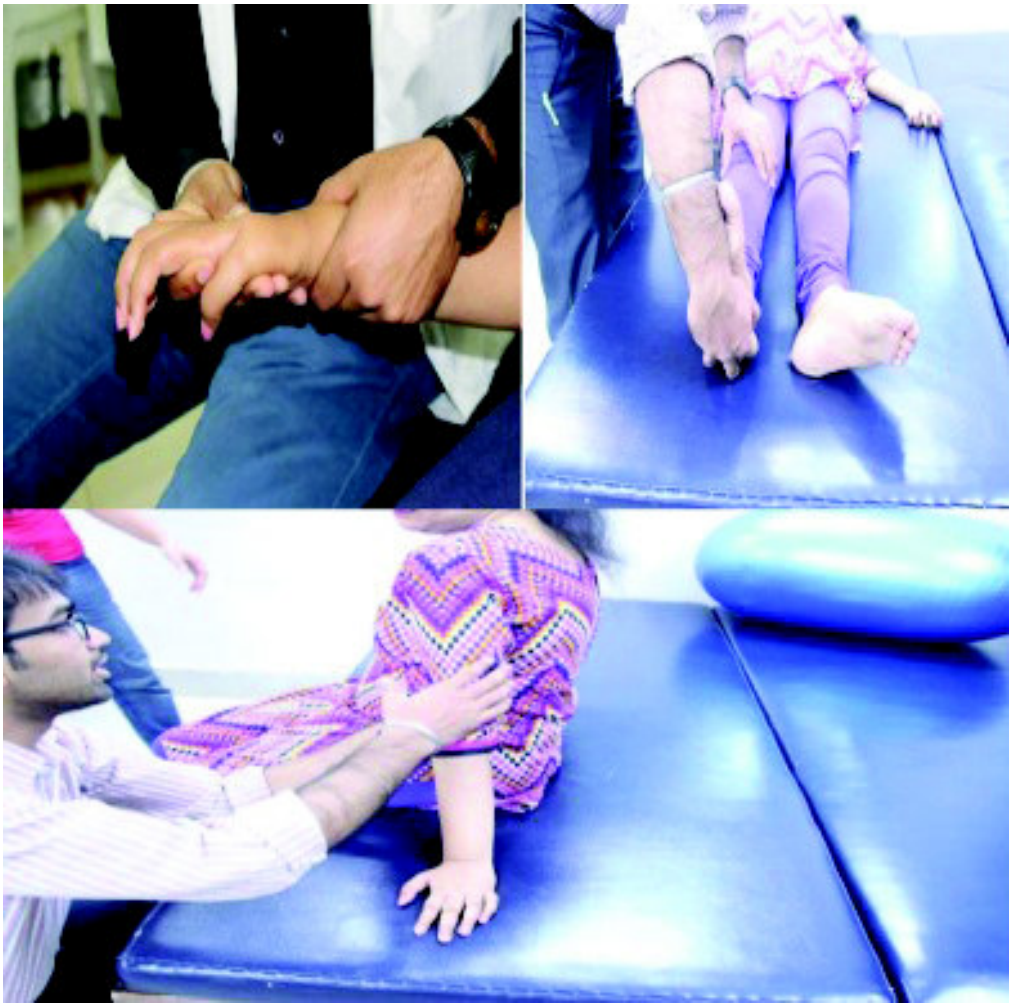


Fig 8.4-8.6 Joint compressions

- tapping on the muscle belly and asking the patient to try to perform the action of the muscles (fig8.7)

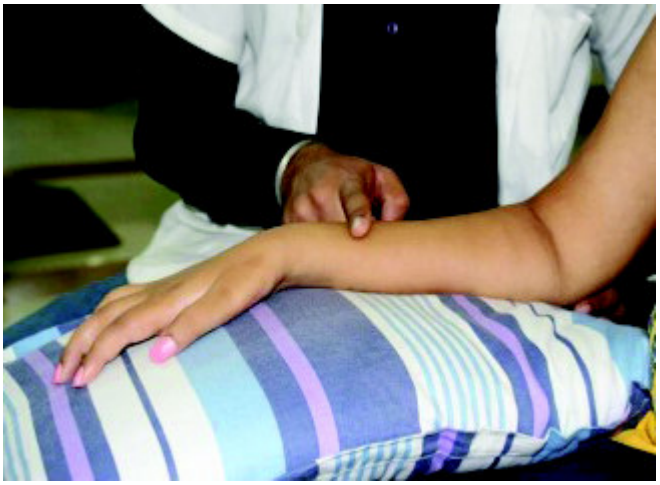


Fig 8.7- Tapping on the muscle belly

- Verbal commands reinforcement during the movement may help the patient to contract the muscle during the movement
- Resistance during the exercise will also lead to developing the tone but it should be very minimal.(fig8.8)

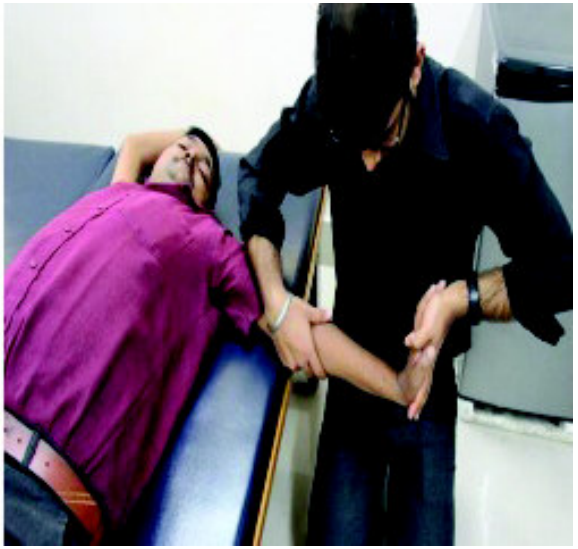


Fig 8.8&8.9: Resistance during the exercise using theratube

After this the regular exercise can be performed.

Upper motor neuron lesion

- spasticity/ hypertone/increased tone in the muscle
- muscles will be contracted and taught
- high resistance felt during passive limb movements

Increased tone can lead to altered movement patterns of upper and lower limb, abnormal timing of movement, loss of hand function and fatigue. This causes motor control issues as the movements are not voluntarily controlled by an individual. This leads in walking problems while taking the step and swinging the leg to place on the floor. The patient may loose balance during movements because of the altered tone and requires more effort and energy to move the legs. Spasticity can also lead to muscle weakness, joint contracture and deformities.

Treatment

Physical therapy

- Sustained stretch to end range of the muscles which are shortened
  - calf (fig8.10)
  - quadriceps
  - hipadductors
  - trunklateral flexors
  - long finger flexors (fig8.11)
- Prolonged weight bearing for more than a minute may help in different positions. (fig 8.12)

Figure 8.12- Prolonged weight bearing in different positions

- Positioning: for limited functional mobility positioning of the individual in normal position is very important. Positioning can be very important during sitting on wheelchair and lying on the bed. A positioning schedule of the individual should be made. Mechanical positioning devices can be used like ankle splint ,toe spreader, finger spreader.
- Proprioceptive neuromuscular facilitation exercise to break the abnormal movement patterns.(fig8.13&8.14)

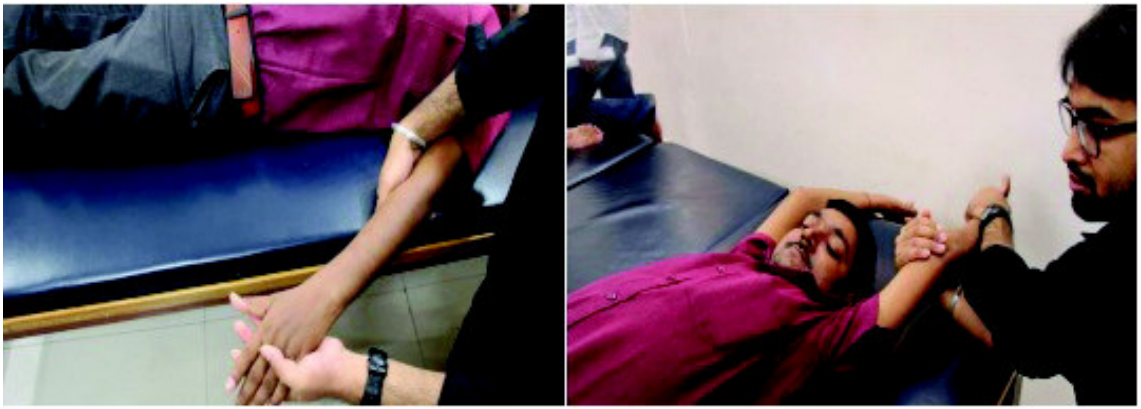


Figure 8.13, 8.14-Proprioceptive neuromuscular facilitation exercise

- Gentle rockingon the ball too and forth.(fig 8.15)

Figure-8.15- Gentle rockingon the ball

- Cold/ ice wraps
- Soothing verbal commands
- Relaxation ofthe muscles
- Inflatable pressure splint

Pharmacological management

Anti spastic agents

- muscle relaxants
- Carbamazepine
- Benzodiazepines (Diazepam, chlorazepam, lorazepam)
- Phenytoin

The medication should always be prescribed by the doctor and should be taken care for the dosage and side effects depending on the patient's status.

Chapter 9

Early fatigue

There's a name for people who save energy…. SMART

Fatigue can be defined as the distinct experiences of tiredness, exhaustion, weakness, and weariness. Fatigue is unpleasant symptoms which include total body feelings ranging from tiredness to exhaustion creating an insistent overall condition which hinders an individuals' ability to function to the normal capacity.

Patients' conceptualization of fatigue is a greater understanding of their subjective symptom, and is a gate way to develop a successful management. Fatigue can be primarily experienced in two ways by patients with MND, and could be experience as use-dependent reversible muscle weakness or feelings of whole- body tiredness, or a combination of the two. It is essential to understand this dichotomy in MND fatigue and understand these issues involved with fatigue, especially that individuals may interchangeably use words such as "fatigue" and "tiredness" to describe essentially different experiences of fatigue. The greater muscle fatigue in ALS patients results from activation impairment, due in part to alterations distal to the muscle membrane. (Fig: 9.1)

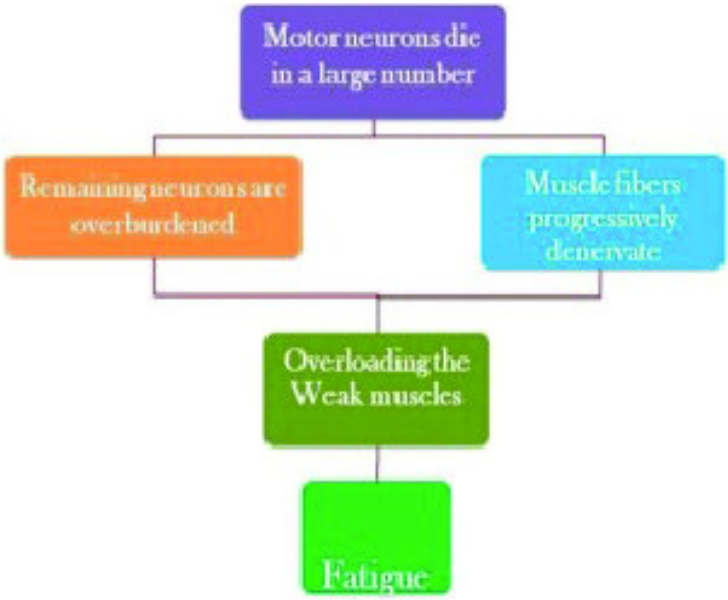


Fig: 9.1: Cause of fatigue

In Bulbar weakness and breathing difficulties fatigue can be more severe than the other form. Fatigue may worsen with symptoms of breathlessness. ALS related fatigue is an independent factor, which deserves individualized approach and treatment. But it is

always important to know these factors and there relation to fatigue can be related to functional limitations, cognitive effects, fatigue development, social effects, negative effect, learned responses and relieving factors and can be used as are lieving factor for its management. Regardless of how fatigue presented its elfin the patients, all patients adopted adaptive strategies.

As well as reconceptualising the difficulty tasks, patients adapted by adopting strategies of budgeting daily energy expenditure and planning around fatigue. Prophylactic rest can be commonly employed by patients who are expecting to expend more energy than they usually would.

Budgeting energy is an adaptation strategy of that was used on a daily basis. Because of the inevitable fatigue that would occur in the afternoons, many patients try to do all of their daily activities in the morning, and are cautious not to "overspend" their daily budget.

The amount of effort required to accomplish such tasks impacted on patient motivation in a number of ways. Patients sometimes avoided actions, like picking up a cup of coffee because a sip of a drink could not justify the inevitable fatigue that would follow.

Interestingly, descriptions of reduced motivation to do task that would lead to fatigue were followed by explanations of how patients felt more motivated to integrate socially with friends and in particular reported that worry and stress contributed tofatigue. Feelings of frustration were often described when patients were unable to do what they wanted to do, because doing so would leave them feeling fatigued.

Treatment

Anti-Fatigue strategies:

1. Learn methods of making every task easier.
2. Use assistive devices when needed.
3. See an occupational therapist for determining what is best for your needs. If you have trouble walking, don't resist get in ganassistive device. The device cans pare you from the exertion of manually doing the activities and spending more energy.
4. Advising more frequent intervals of rest including and possible
5. Pace yourself. Perform activities slowly and easily. Stop and rest often and take a few breathes before you start again. If you become breathless during a task, stop the task.
6. Alternate activities with periods of rest. Schedule regular rest periods each day, perhaps every4hourly.Rest before going away.
7. Plan your activities and gather everything you need before you start. Schedule heavier task during predicted times of higher energy.
8. Don't stand when you can sit. Utilize possible shortcuts. Obtain assistance in Completing tasks if you need help.

9. Maintain a good posture and try avoid faulty posture like slouched sitting.
10. Always allow enough energy to enjoy at least one valued experience each day.
11. Try to establish a regular sleeping pattern. If you have problems waking up at night, determine why and what to do about it.
12. Avoid prolonged bathing in warm water, as it may worsen muscle fatigue. Be cautious of extreme outdoor temperatures.
13. Maintain your nutritional requirements each day, and prevent unnecessary weight loss.
14. Avoid stressful situations as much as possible.
15. Inform your doctor know if you feel notice ably weaker or have difficulty breathing after taking a medication. Perhaps medication can be substituted for another one or the dosage altered.
16. Make your living environment accessible for daily activities, and promote energy conservation. Moving abed to another location or relocating personal items are some examples.

Energy conservation technique

1. Problem solving -Figure out the activities which require causes you fatigue, pain or discomfort. If a task causes you a problem, think of getting do fit or do it differently. You can seek some ones help.
2. Planning-Create a plan of the things you want to achieve during the day or over the week. Plan how and when you're going to do certain tasks, and make sure that demanding jobs are spaced out during each day or week. Spread them out wherever possible over a number of days
3. Prioritizing-List your tasks according to priority and necessity, you can put them in order of importance and can remove, delay the t ask which is not important. Thin khow this can be done, how to be done, I have to do or someone else can help.
4. Pacing-Break tasks down into achievable parts and spread them throughout the day or week, and take short, regular rest breaks. Change your position and activity regularly. Avoid rushing and unnecessary motion. Arrange your work center and use proper working conditions.

Breathing exercises, relaxation exercises and Meditation can also be useful to limit fatigue. Managing fatigue can result in unnecessary suffering, social isolation and rapid physical deterioration. Effective management will maximize wellness and abilities, giving you the desire to keep living and the strength to carry on. Although in ALS we can't seize away fatigue, we can learn to overcome fatigue and not let it overcome us.

Chapter 10

Pain, Cramps & Fasciculations

Limit pain, cramps and fasciculations but not your activities.

Pain, cramps and fasciculations are very common symptoms seen in all the motor neuron disease but they get unnoticed in the early stage by the patient as well as the caregiver. The symptoms are only expressed when these verity increases. It is fore most important to notice the see changes at a very early stage and plan a care to alleviate these symptoms before it limits the functional level of an individual.

The commonest cause (fig. 10.1) of pain, cramps and fasciculations are:

- Motor neurons die in a large number
- Remaining neurons are overburdened
- Muscle fibers progressively denervate
- Resulting in decrease muscle volume
- Hyper excitability of reserved motor nerves
- Weak muscle work at maximal strength to perform same activities

Individuals with MND frequently develop musculo skeletal pain syndromes such as frozen shoulder low back pain, and neck pain due to muscle weakness and in ability to change positions.

The pain causes restriction in range of motion (ROM) and prevents activities. Low back pain can be trigger edbyanuncom for tables eating position. Neck pain associated with head drop is one of the most difficult musculoskeletal pain issues to remedy.

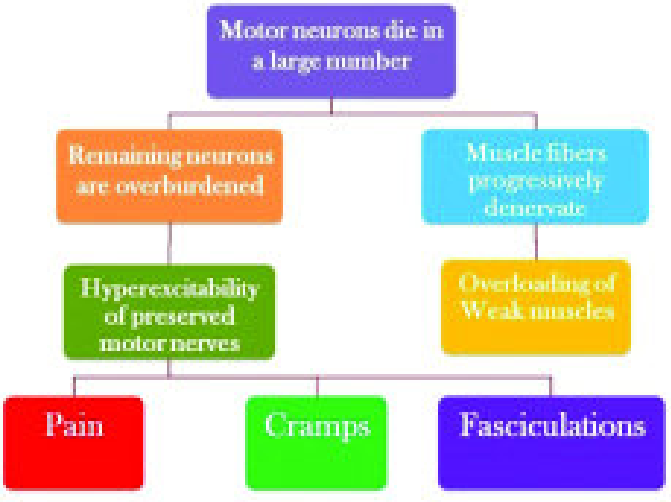


Fig10.1: Cause of pain, cramps and fasciculations.

Treatment

1. Exercises

- ROM exercises
- Joint mobilization: mobilization can be performed to improve joint integrity and flexibility of the issues and joint. A physical therapist can help the patient or the care giver in teaching mobilization.
- Maintaining correct postural alignment: sitting and standing with a erect posture may help.
- Assistance : supporting the arm as much as possible an allowing it to dangle at the side may avoid shoulder pain. A lumbar support for the wheelchair, a good cushion on a solid seat, encouraging frequent weight shifts, and are clining back or tilt-in-space wheel chair. A variety of cervical collars ranging froma soft foam to a semi open to a hard plastic collar may be tried. As weakness progresses, the soft collars, which are mostcom for table, provide in adequate support. The hard collars are usually uncomfortable and poorly tolerated. A head support on the wheelchair or are clining lounge chair may be more comfort able than a collar. When a head droph as developed, the patient should be assisted with neck ROM exercises to prevent contract ure of the anterior neck muscles.
- Stretching exercise: for tight and spastic muscles.
- Relaxation exercises: releasing the muscle tension by slowly relaxing the muscle of every joint one by one.
- Therapeutic massage
- Yoga
- Meditation

2. Physical modalities

- Application of Hot
- Application of cold
- Hydrotherapy
- Ultrasound

  Physical the rapist can guide you for the best suited treatment option depending on the pain.

3. Pharmacological measure

- acetaminophen,
- non-steroidal anti-inflammatory drugs (NSAIDs),or opioids.

  As much as possible these medicines should be avoided. Careful administration of medications is useful for patients because each has a different action and side effects, the medications must be prescribed for the right dosage and combination.

Cramps

Musclecramps and spasms can be experienced due to altered muscle tone. It may occur during any activity done beyond the capacity of the weak muscle.

Muscle cramps may occur in any part of the body:

- Tongue
- Jaw
- Neck
- Abdomen
- Arm, hands
- Thigh or calf

Treatment

- Pharmacological treatment like anti-cramping drugs
- Muscle relaxants
- Carbamazepine
- Benzodiazepines (Diazepam,chlorazepam,lorazepam)
- Phenytoin
- Exercises
- Muscle stretching
- Adequate hydration and nutrition
- Relaxation
- Stretching and flexibility exercise
- Therapeutic massage to increase the blood flow of the area.
  Application of heat can also increase the blood circulation and may relief the cramp.

Fasciculations

- Fasciculations are often visible through the skin and are involuntary contraction of any of the muscle. It can be present with musclecramps or spasm also. It is seen particularly in calf muscles, hand, feet and eyes.

Treatment

- Patient with brisk wide spread fasciculations can avoid or minimize caffeine(coffee)
- andnicotine (avoid smoking).
- Lorazepam can also help to reduce fasciculations.
- Relaxation techniques

Chapter 11

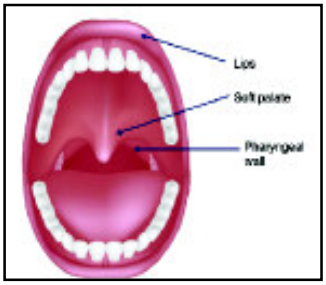
Speech Difficulties

Communication - the essence of human connection!

The speech disorder most commonly seen in ALS is clinically known as "Mixed (Spastic+Flaccid) Dysarthria". This type of speech disorder is mainly caused due to the weakness, slowness, or sometimes paralysis of muscles, that is encountered in Motor Neuron Diseases; especially ALS.

WHAT CAUSES SPEECH PROBLEMS IN MND/ALS?

The tongue, lips, jaw, soft palate and larynx along with their muscles, together help us to speak. When these set of oral muscles are affected (weakened / paralysed), the range of movement, speed of movement and accuracy with which these muscles move is lost. Thus the overall precision of producing accurate sounds is lost. Hence, the speech produced is considered to be out of the normative range and is unclear/muffled/non-intelligible.



Respiration also gets affected in many cases. Change of breathing pattern often leads to a change in the voice quality, making the voice breathy in quality. Weakness of soft palate also influences the voice quality making it nasal in nature.

All the above changes causes the patient to "sound" completely different, making the patient unrecognizable by voice print.

CLINICAL SYMPTOMS

Symptoms seen in initial stages:

- Loss of precision of certain sounds
- Slurring of speech
- Worsening of speech with long hours of speaking (fatigue) Muffled speech quality
- Breathy/hoarse voice
- Symptoms seen in severe stages:
- Inability to speak loudly
- Inability to change pitch of voice
- Slow rate of speaking
- Nasal voice quality Breathlessness while speaking

MANAGEMENT

A Speech Language Pathologist will provide exercises of oral muscles. These exercises help in improving speech intelligibility, and also prevent the oral muscles from getting fatigued during speech acts, thus helping effective expression.

They include:

Oro-motor exercises: Strength and rapid mobility of oral muscles is a requisite for clear speech. Oro-motor exercises involve exercises of the lips, tongue, jaw and soft palate in order to build strength and the necessary mobility in these muscles.

The oro-motor exercises include:

- Lip retraction spreading exercises
- Lip pursing, puffing exercises
- Tongue protrusion, retraction, elevation, depression and lateralization exercises
- Tongue rotation exercises
- Blowing and sucking velar exercises

The SLP will make the use of an Oral Motor Kit (as shown below) in order to aid exercises of the oral muscles.

Proprioceptive Neuromuscular Facilitation (PNF): It is a rehabilitative technique used to stimulate the neuromuscular system to excite specific muscles in the effort to bring about desired movements. The SLP will use a Finger Brush, and other assistive materials in order to give taste, temperature and tactile stimulation to the oral muscles to assist the movement of those muscles.

Intelligibility Drills: Once the oral muscles start gaining increasing strength and mobility, these muscles have to be integrated with speech movements. The SLP will bring about this integration by prescribing the patient with intelligibility drills. This will help improve intelligibility in daily conversations, helping patients regain interest and confidence to speak.

Communication Strategies: Communication strategies may be prescribed by the SLP (if required) in order to fill the intelligibility gap that may exist even after speech production strategies are given.

The SLP may also serve the patient with an alternative/ augmentative communication (AAC) aid/ device to bring about effective communication, in patients where speech production may not be the best communication option.

Some AAC option are:

1) **AAC Board:** This board is costume made for each patient and may consist of pictures/ words. The SLP trains the patient to use this board to communicate using either finger or laser pointing.

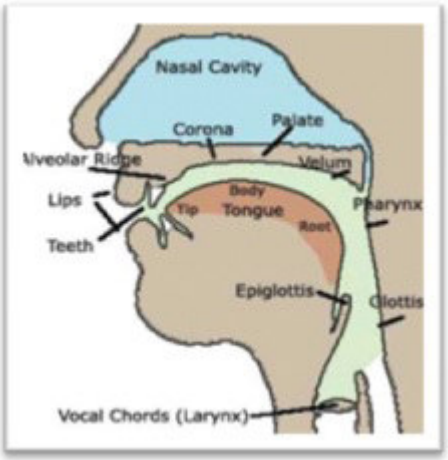
2) **Speech Generating Devices:** These devices produce speech either in written or spoken format and hence help in aiding communication.

3) **AAC Apps:** There are a number of Android and ions AAC Applications. The SLP will make the patient avail to these Apps and will also train the patient to use them effectively for quick communication.

ARTICULATION THERAPY

Patient should make conscious effort to produce the below sound several times a day. Take time to make each sound as clearly as possible put emphasis on each letter sound. Say each letter as loudly as possible.

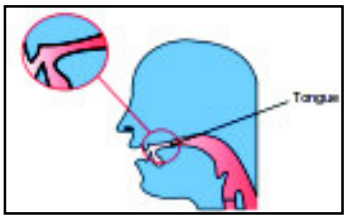
Articulation therapy includes exercises and/or techniques, given in order to increase the clarity of sounds in speech. The SLP targets the improvement of sounds by mainly giving the feedback of how these sounds are actually produced, so that the patient can consciously make an effort to produce them accurately.



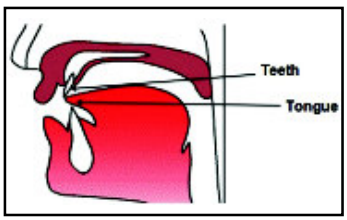
The diagram given below shows the various articulators which help to produce speech sounds. Patient should make conscious effort to produce the below sound several times in a day. Take time to make each sound as clearly as possible. Put emphasis on each latter sound.Say each letter as loudly as possible.

Now that we are a little oriented to the clinical names of different oral structures, let us have a lot as to how we produce sounds.

Bilabial sounds: They are the sounds that are produced by bringing the two lips together; sounds like /p-u-h/, /b-u-h/, /m-u-h/.

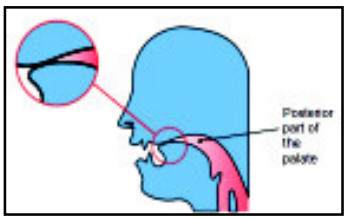


Labio-dental sounds : They are sounds that are produced by touching the the teeth to the lips; sounds like /f-u-h/, /v-u-h/.

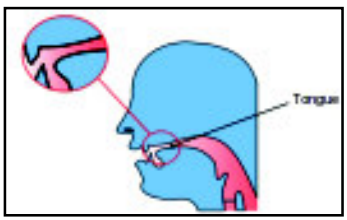


Lingua-dental sounds : They are sounds that are produced by touching the tongue to the teeth; sounds like /d-u-h/, /t-u-h/.

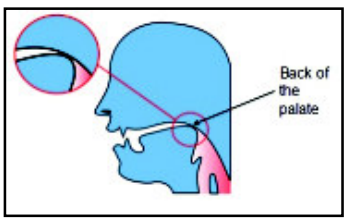
Alveolar sounds : Sounds that are produced by touching the tongue to the alveolus; like /l-u-h/, /r-u-h/.



Palatal sounds : Sounds that are produced by touching the tongue to the palate; sounds like /t-u-h/, /d-u-h/.



Velar sounds: Sounds that are produced by touching the back of the tongue to the back of the palate; sounds like /k-u-h/, /g-u-h/.



Chapter 12

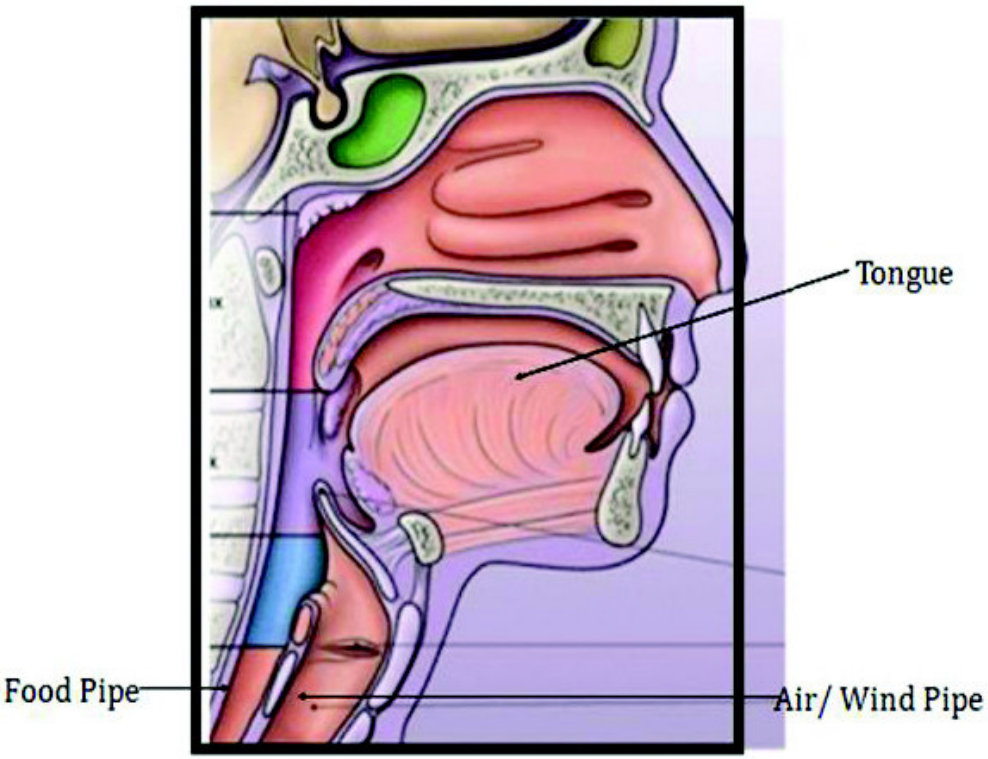
Swallowing Difficulties

Humans love and live to eat!

Swallowing is one of the very first acts that a human child elicits in its lifespan. Since swallowing is acquired very early and easily, it is taken for granted many a times. But very few know that a highly complex neural network is triggered in order to carry out a single swallow.

The lips, tongue, jaw, soft palate and pharyngeal wall are used to bring about a swallow. Undeniably, these are the same set of muscles used while speaking. Hence it is very commonly seen that speech and swallowing functions both are hampered as a sequel of one another.

As discussed under the title of "Speech Disorder" in MND/ALS, oral muscles are affected. As an effect, the oral muscles (specially tongue) are unable to push the food posteriorly into the food pipe, hence creating a risk for food to enter the airway. Food entering the airway (lungs) is clinically known as "aspiration".



CLINICAL FEATURES

Some of the signs of swallowing problems are:

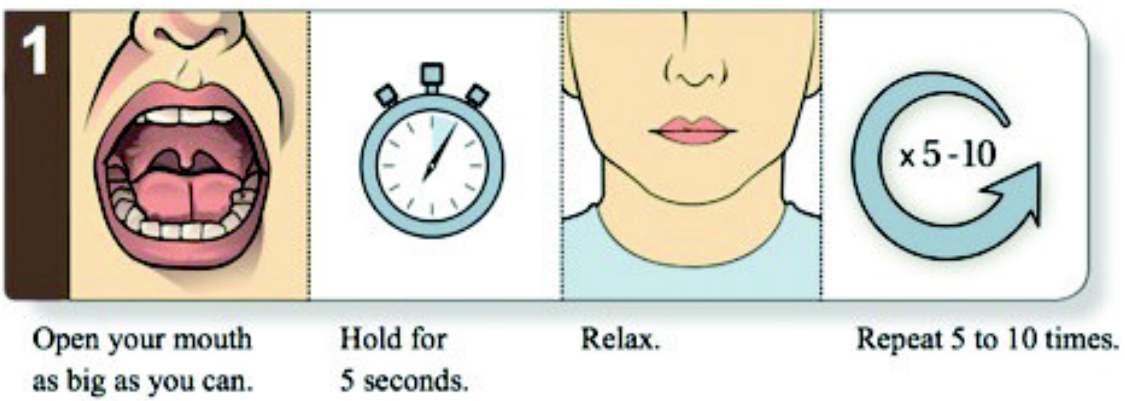
- Inability to chew food properly
- Frequent cough while eating
- Eye watering/ eye reddening while eating
- Feeling of breathlessness while eating
- Effortful swallow
- Increased meal time with reduced meal intake
- Sudden weight loss
- Feeling of something stuck in throat Spilling out of food from corners of mouth Wet voice quality

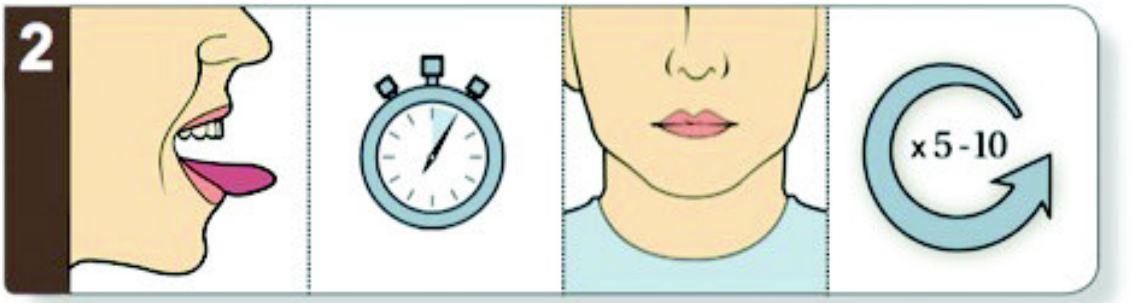
MANAGEMENT

After a detailed swallow examination, the Speech Language Pathologist (SLP) will prescribe the patient with certain patient-specific techniques, manoeuvres and exercises and may also suggest an alteration in the type of consistency that is safe for the patient.

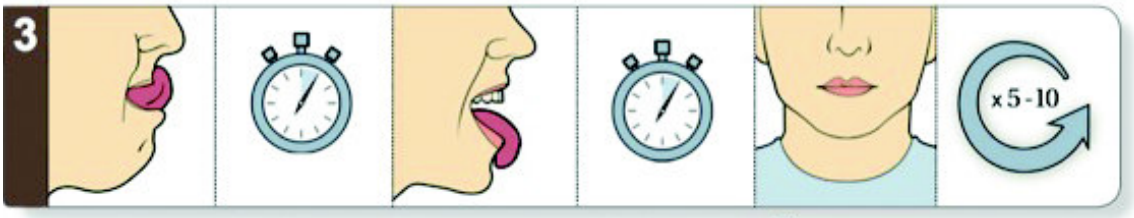
Some of the therapeutic management are:

1) **Oro-motor exercises :** These exercises work on
   - Jaw opening
   - Tongue protrusion
   - Tongue elevation and depression
   - Tongue lateralisation





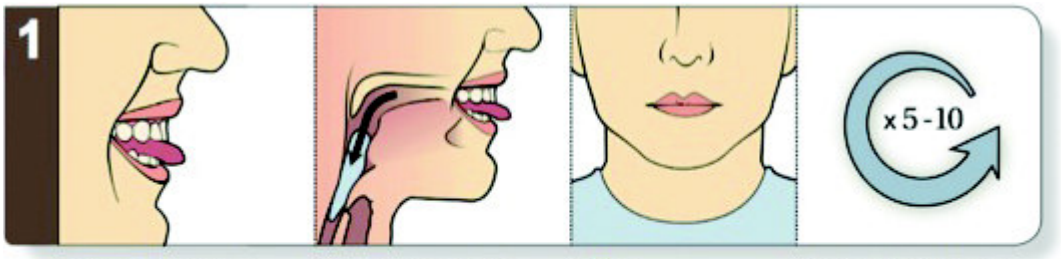
Stick your tongue out as far as it can go. Hold for 5 seconds. Relax. Repeat 5 to 10 times.



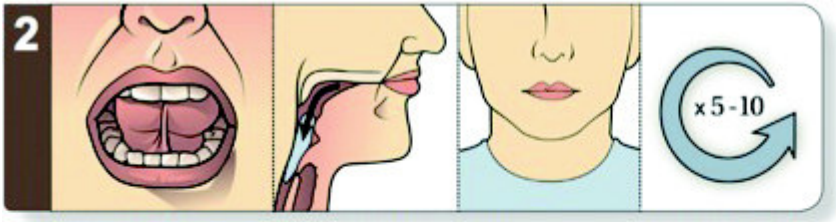
Move your tongue up towards your nose. Hold for 5 seconds. Now move your tongue down towards your chin. Hold for 5 seconds. Relax. Repeat 5 to 10 times.

Move your tongue to the left side. Hold for 5 seconds. Now move your tongue to the right side. Hold for 5 seconds. Relax. Repeat 5 to 10 times.

2) Manoeuvres to improve swallowing



Stick out the tip of your tongue. Hold it between your teeth or lips. Now, try to swallow your spit with your tongue in that position. Relax. Repeat 5 to 10 times.



Press your tongue against the roof of your mouth as hard as you can.

With your mouth closed, swallow your spit (saliva) as hard as you can.

Relax.

Repeat 5 to 10 times.

Try swallowing some water now

Take a small sip of water and swallow it. You may need to swallow it a few times to get it all down. Clear your throat or cough if you need to.

3) **Techniques to reduce risk of aspiration :** The SLP will prescribe patient specific "Safe Swallow Techniques" in order to reduce risk of aspiration.

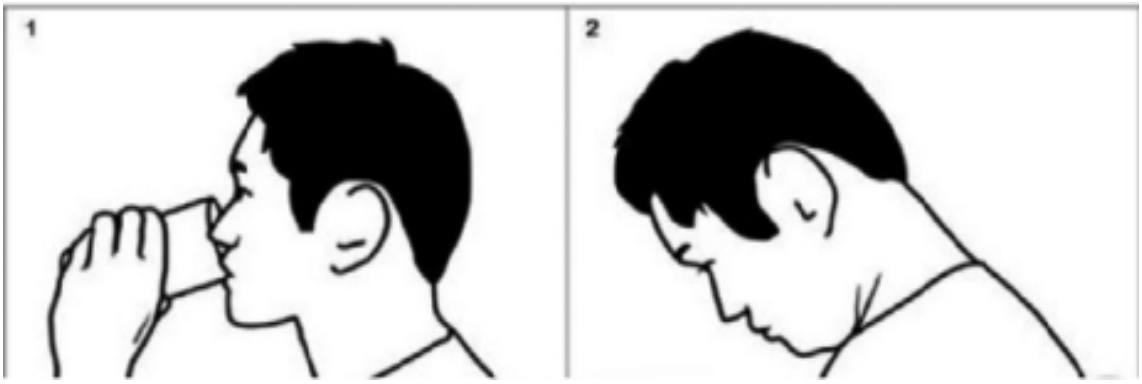
Swallowing therapy is mainly divided into two parts:

a) **Restorative exercises :** These exercises mainly help to restore the function of the muscles which may have been weakened.

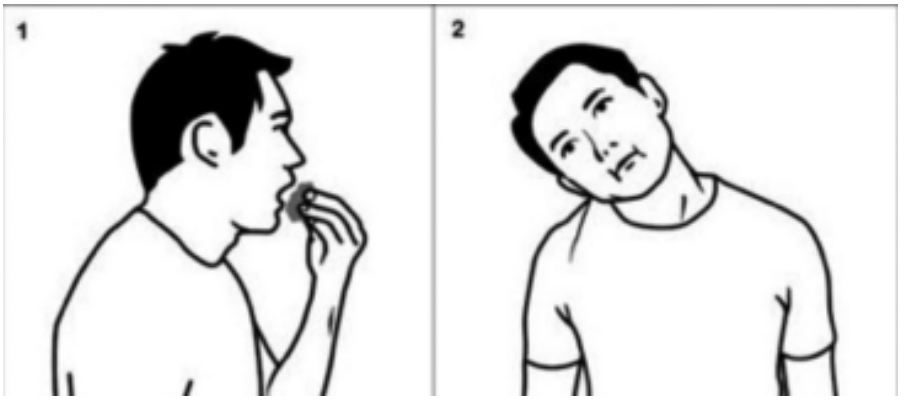
b) **Compensatory techniques :** The SLP recommends these techniques when the muscles have reached the maximum recovery stage and/or the further recovery may be very slow. These techniques thus help the patient to start having oral feeds safely and comfortably even while the muscles are in the process of recovery. While eating or drinking the patient should have position of 900 upright sitting with the head low, chin tuck and slightly kneeling forward.

Compensatory techniques

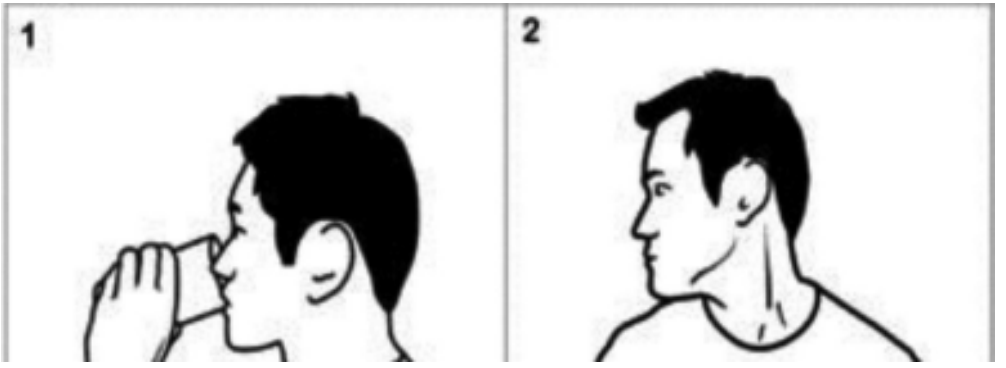
a) Chin-Tuck Technique



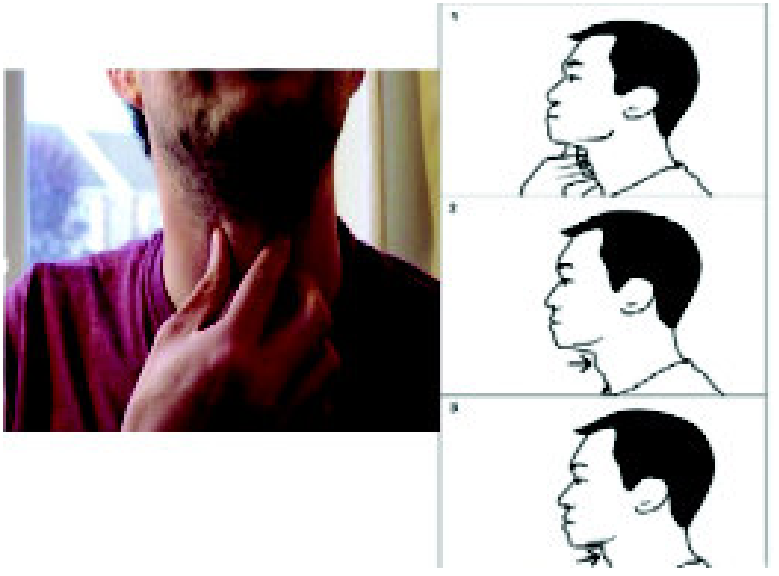
b) Head Tilt Technique



c) Head-Turn

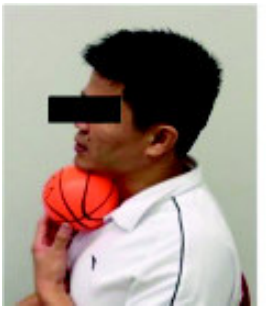
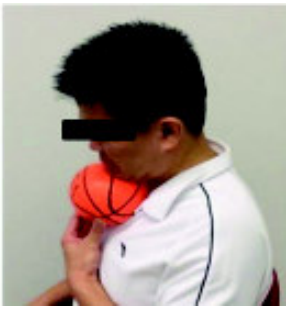


d) Mendelsohn's Maneuver

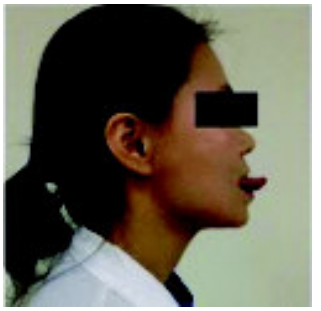


EXERCISES TO BE DONE

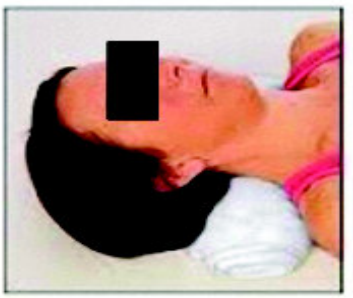
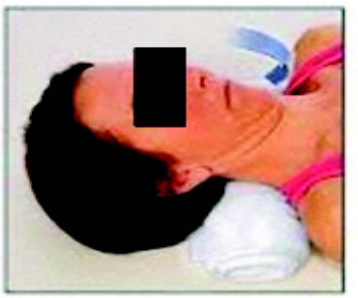
a) Chin-Tuck Against Resistance

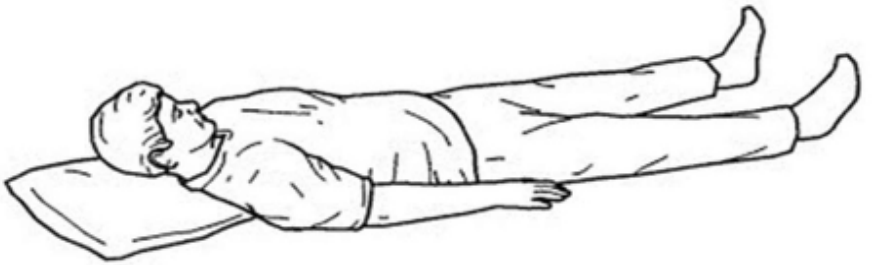
b) Masako Maneuver



c) Shaker's exercise

d) Passive Shaker's exercise



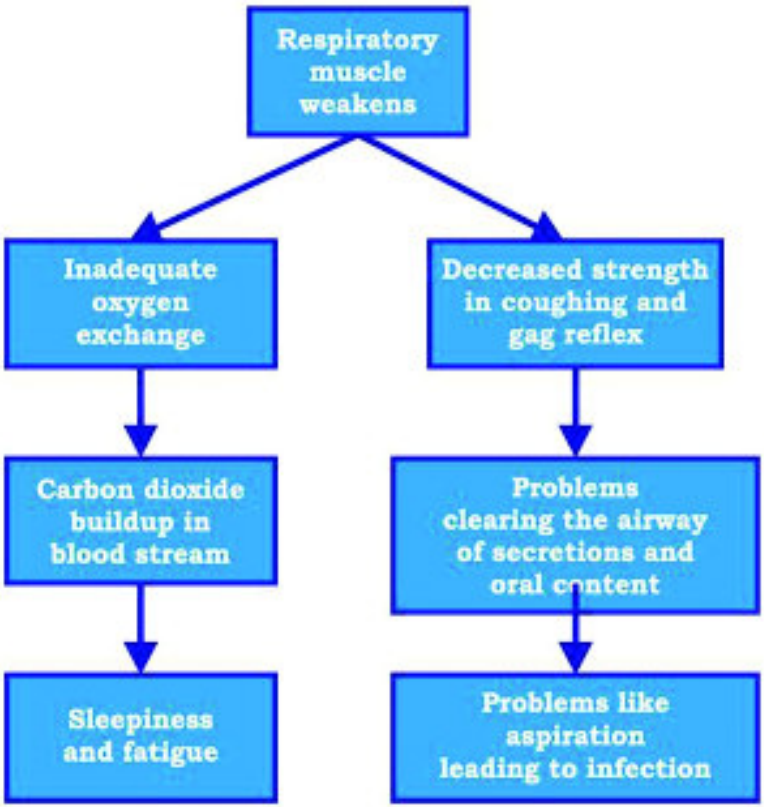
4) Alteration of type of consistency: The SLP along with the Dietician will prescribe the patient with an alteration of food consistency if required in order to preserve the safety of the airway. At all times, care will be taken that the patient's nutrition and hydration is maintained even through with prescribed alternative diet.

5) Active monitoring of weight: Any weight loss encountered by the patient will be targeted by both the SLP and Dietician. The SLP will provide the patient with swallowing strategies which will reduce fatiguability caused to the patient while eating and hence increase food intake.

Chapter 13

Breathing Issues

Respiratory problems are, unarguably, the most serious of medical complications in ALS. Breathing difficulties occur from the gradual deterioration of breathing muscles, i.e. the diaphragm and the intercostals. The course is as shown below.



SIGNS OF RESPIRATORY INSUFFICIENCY

1. General fatigue, drowsiness, lethargy.
2. Sleep disturbances such as nightmares, night terrors, sleep apnoea (interrupted breathing during sleep) or sudden awakening.
3. Morning headaches.
4. Daytime sleepiness.
5. Confusion, disorientation, anxiety.
6. Poor appetite, weight loss.
7. Excessive yawning or hiccups.

8. More laboured breathing, especially when lying down.
9. Rapid, shallow respirations with increased heart rate.
10. Weakened or softened voice; speaking in short phrases; inability to sing or shout.
11. Difficulty coughing and clearing the throat (weakened abdominal and throat muscles also contribute to this)

Monitoring for respiratory problems

1) Pulmonary function test (PFT)
2) Regular monitor of SPO2 (Normal SPO2 should be more than 98%)
3) Spirometer reading

Managing Breathing Problems

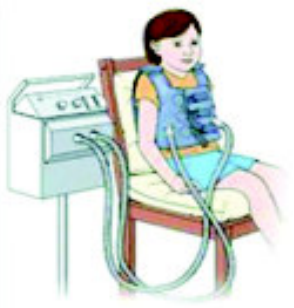
1) Breathing Exercises:
   - It is very important to start breathing exercises regularly even before the symptoms start.
   - This prevents fatigue and delays muscle weakness.
   - Deep breathing exercises are recommended to improve the lung capacity. Yoga can be very beneficial for eg: (Anuloma Viloma) breathing exercises.
   - Spirometer is a device that can be used for breathing exercise

2) Medications

- **Bronchodilators :** Dilate the airway to help breathing, but otherwise ineffective due to muscle weakness. This can be used by nebulization.
- **Expectorants :** To thin out the secretions and make them easier to cough out.
- **Saliva management :** Weakened breathing muscles and weaker muscles of the mouth can cause drooling. Medications can be prescribed by the physician to control drooling (glycopyrolate).
- **Supportive medications :** The physician may prescribe other drugs for symptomatic relief such as for pain, depression, sleep disturbances, constipation, etc.

3) Chest physiotherapy:

- Chest physiotherapy can be provided by a specialized trained therapist.
- They may teach breathing maneuvers to prevent atelectasis (lung collapse) and aspiration.
- They can also teach breathing exercise to improve the



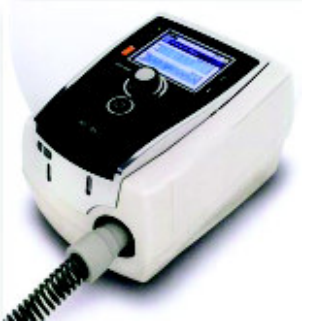
lung capacity. Chest physiotherapy can help to clear secretion from the base of the lungs. It can help to cough out when the patient has weak cough.

4) Cough Assist Device:

- Automatic Mechanical cough assist devices such as the Philips Cough assist or Hillrom Vital cough helps those with an ineffective cough by breathing through a mouthpiece or masks
- The device gradually applies positive pressure to insure a deep breath, then shifts to negative pressure to assist with pulling secretions upward, simulating a deep natural cough.
- High frequency chest wall oscillation involves an inflatable vest that is attached to a machine.
- The machine mechanically vibrates at a high frequency. This helps to loosen and thin mucus and clear the airway.

5) Noninvasive Breathing Support

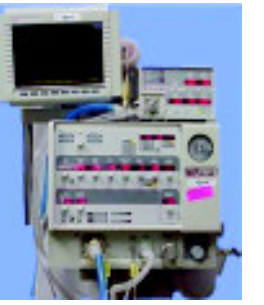
A BiPAP Machine, is often prescribed in ALS.



- BiPAP is short form of Bilevel Positive Airway Pressure and delivers air at two pressures, one for inspiration and one for expiration (inhalation and exhalation).
- A number of nasal or face masks and attachments are available, can be customized for the best fit.

C Pap Machine continuous pressure assisted. Positive ventilation.

Pressure-cycled vent machines, which deliver air at a set pressure level with a variable volume of air on a timed cycle.



Advanced respiratory care in ALS

- As ALS progresses, the person living with the disease may become increasingly dependent on ventilation and ultimately, will require invasive ventilation with tracheostomy. This will provide more efficient ventilation and better control of the upper airway and secretions
- Ventilation through tracheostomy
- A tracheostomy is a surgically created hole in the trachea (windpipe) through which air is forced. The tube through which the air is delivered also is called a tracheostomy (trach) tube.

Caring for the Tracheostomy

- Its normal for the patient to have small amount of mucus around the tube
- Hole in the neck should be pink and painless.
- It's always beneficial to carry an extra tube in case of plugging of the tube.
- Hold a tissue or cloth to catch the mucus coming from the tube.
- Consult a doctor to know how to prevent the plugs in the tube as the patients' nose will no longer keep the air moist.
- One of the strategies to keep the breathing air moist is to put a wet gauze or cloth outside the tube.
- A few drops of salt water (saline) will loosen a plug of thick mucus. Putting a few drops in the tube and windpipe, then taking a deep breath and coughing will help bring up the mucus.
- Protect the hole in the neck with a cloth or tracheostomy cover when outside. These covers can also help make the breathing sounds quieter.
- During showers, cover the hole with a tracheostomy cover. No swimming is allowed with a tracheostomy tube.
- To speak, the patient will need to cover the hole with a finger, a cap, or a speaking valve.
- Once the hole in the neck is not sore from the surgery, clean the hole with a cotton swab or a cotton ball at least once a day to prevent infection.
- The bandage (gauze dressing) between the tube and neck helps catch mucus. It also keeps the tube from rubbing on the neck. Change the bandage when it is dirty, at least once a day.
- Change the ribbons (trach ties) that keep the tube in place if they get dirty. Make sure the tube is held in place while changing the ribbon. Two fingers should be fit under the ribbon to make sure it is not too tight.

LTV® 1150 ventilator

- Provide your adult and pediatric patients pressure control and pressure support without a high pressure oxygen source
- The LTV 1150 ventilator provides portable, advanced ventilation for adult and pediatric patients at home or a post-acute care facility
- At only 14 lb, the ventilator provides a wide range of ventilation therapies to meet demanding patients needs, including volume control, pressure control, pressure support and spontaneous breath types.

Advantages of invasive ventilation:

1. It provides for much longer survival
2. It provides a secure connection directly to the airway for suctioning secretions.
3. It leaves the face free, without headgear, straps, and skin pressure problems on the pace.

Decision making regarding invasive care

The decision to choose invasive mechanical ventilation is a very personal one. People with ALS who choose Invasive Ventilation can live for years. Some of them are cared for in nursing homes while others are able to remain at home.

So, in making the decision, one should consider his family support, level of independence and financial resources.

Chapter 14

Ambulation Issues

Ambulation in a MND patient depends on the amount of movement one can perform. The training protocol in these people differs accordingly. If the patient has adequate movement then therapists incorporate accommodative and restorative approach. On the other hand if the patient has relatively less movement then the therapists incorporate restorative and compensatory approaches in their therapy.

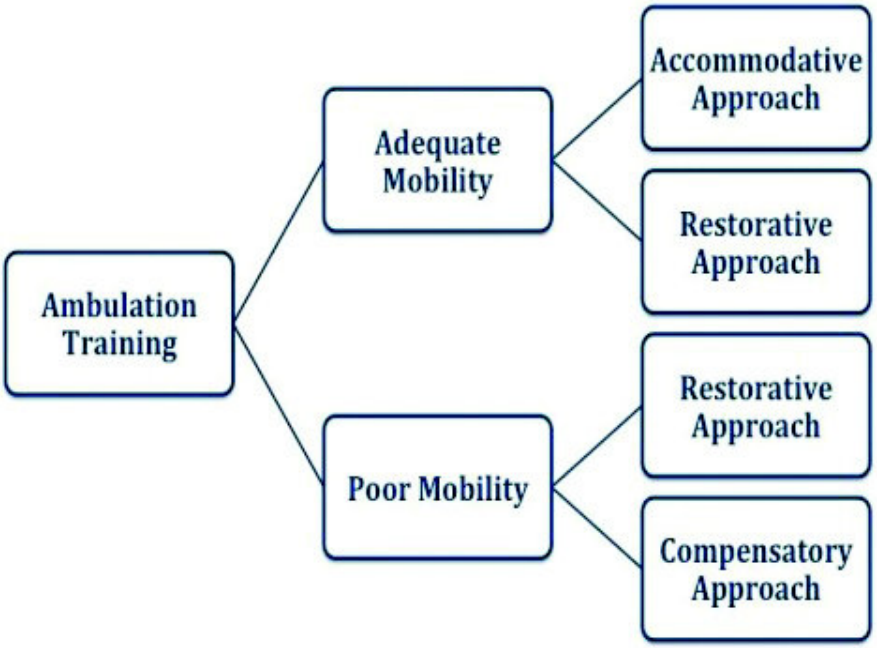


Fig 14.1: Approaches incorporated for ambulation depending on the available movement

Adequate Mobility

Patients with adequate mobility are those who are relatively independent in their daily activities. Their walking capabilities can be more enhanced by making use of the following approaches.

1. Accommodative Approach This approach includes making use of external devices so as to compensate for the lack of movement.

i. Walkers

- They provide more support than the canes and crutches.
- They could be with or without wheels depending on the stability of the patient.

Fig 14.2: Types of Walkers

ii. Canes

- They provide the least amount of support.
- They could be given in early stages when upper limb strength is good.
- It is given in the hand opposite to the weakest lower limb.

Fig 14.3: Types of Walking Canes

iii. Crutches

- They are rarely recommended.
- They are given to the patients with excellent upper limb strength and good trunk control.

Fig 14.4: Types of Crutches

iv. High boots

- In these shoes, there is no movement at the ankle joint. It is like a fixed Ankle Foot Orthosis.
- Most of the patients have a foot drop due to muscle weakness, thus it is a must to use these boots so as to clear the ground.
- It is given in patients who have lack of medial lateral stability along with poor knee control.

Fig 14.5: High Boots

v. Dynamic Ankle Foot Orthosis

- They have a hinged joint incorporated between the foot and calf.
- For this kind of an orthosis, the patient requires good knee control.
- Climbing stairs and sit-to-stand becomes easier with this AFO as compared to a high boot.

Fig 14.6: Dynamic AFO

2. Restorative Approach In this, we work on the weakened aspect and put in continued effort to improve weak muscles and make normal muscles supernormal.

i. Stretching Exercises

- Flexibility at a joint affects the movement at the joint resulting in affection of the walking pattern.
- Regular daily stretches help to maintain muscle length and keep joints mobile to prevent any deformities.
- Thus stretching plays a very important part in the exercise protocol.
- However, some amount of tightness or spasticity is helpful as it helps in maintaining postures.
- Thus "selective" stretching is a must.
- Hold each stretch for 30 seconds and repeat 3-4 times.
- These stretches should be performed at least 5 times in a day.
- If the range is not complete, try to stretch it slowly and gently; just a little more each time.

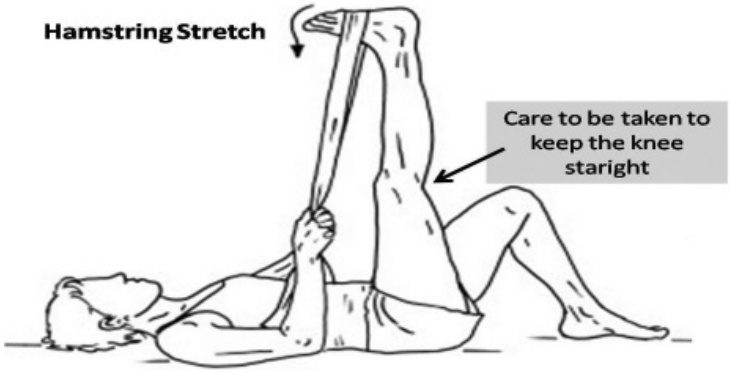


Fig 14.7: Hamstring Stretch

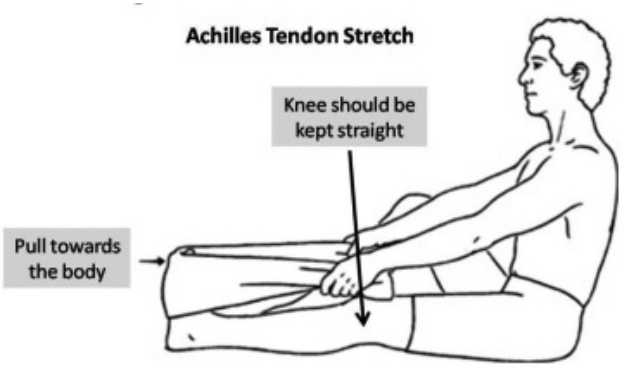


Fig 14.8: Achilles tendon Stretch

ii. Exercises to improve your stamina

- It includes exercises of minimal to moderate resistance but higher repetitions.
- It mainly includes building up of stamina.
- These improve the capacity of the muscles to perform an activity for a longer time.
- It also helps in improving physical functioning of the patient.
- It involves activities like walking, cycling, and swimming.

iii. Exercises to improve your strength

- Greater the strength in muscles more is the gait speed and vice-versa.
- Strength training can be given by making use of weight cuffs, therabands, springs, and theratubes.

iv. Exercises to improve your balance

- Standing sway exercises

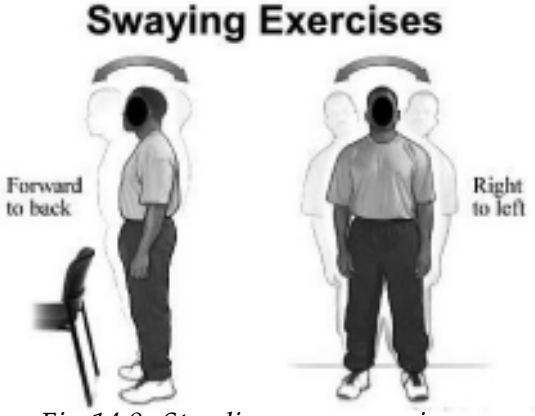


Fig 14.9: Standing sway exercises

- Tandem standing

Fig 14.10: Tandem Stance

- Stand with feet together
- Standing on one leg

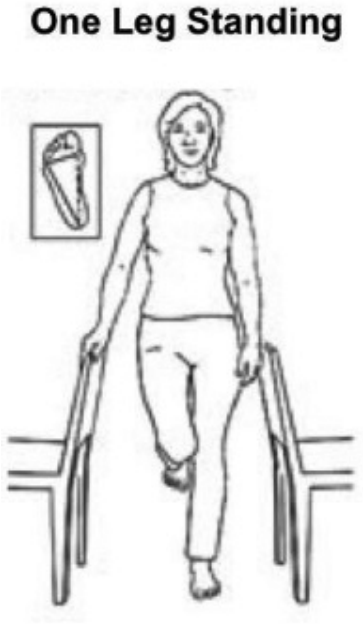


Fig 14.11: One Leg Standing

- All the above-mentioned exercises on a foam pillow so as to increase the difficulty level.

Fig 14.12: Tandem Stance on a Foam Pillow

- Balancing on a balance board

Fig 14.13: Balance Training on a Balance Board

Poor Mobility

Patients with poor mobility are those who are dependent on others for most of their activities of daily living. Their ambulatory capabilities can be more enhanced by making use of the following approaches.

1. Restorative Approach
   i. Stretching Exercises
   ii. Exercises to improve your stamina
   iii. Exercises to improve your strength

2. Compensatory Approach

i. Wheelchairs

- As the disease progresses, walking becomes more and more energy consuming and thus more dependent on wheelchair for ambulation.
- It usually happens in middle to late stages of MND.
- Depending on the patient's condition, the patient either uses manual or a motorized wheelchair.

Fig 14.14: Manual Wheelchair

Fig 14.15: Motorized Wheelchair

Exercise Program For Gait Training

1. Forward Walking
2. Backward Walking
3. Sideways Walking

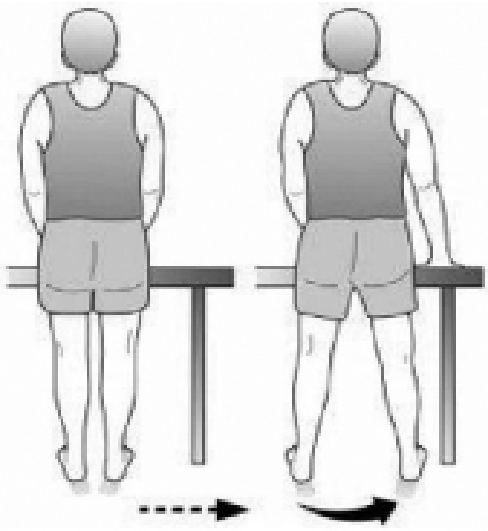


Fig 14.16: Sideways Walking

4. Tandem Walking

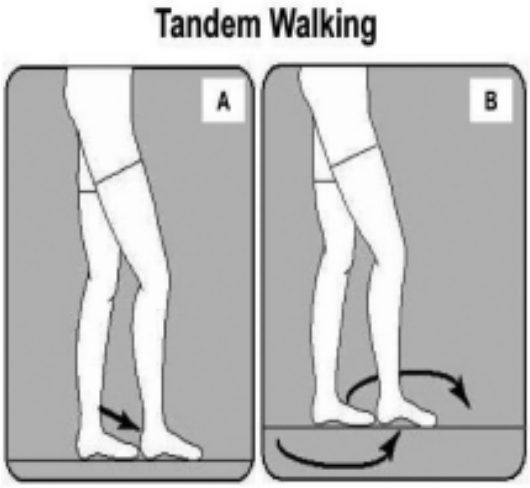


Fig 14.17: Tandem Walking

5. Walking On Toes & Walking On Heels

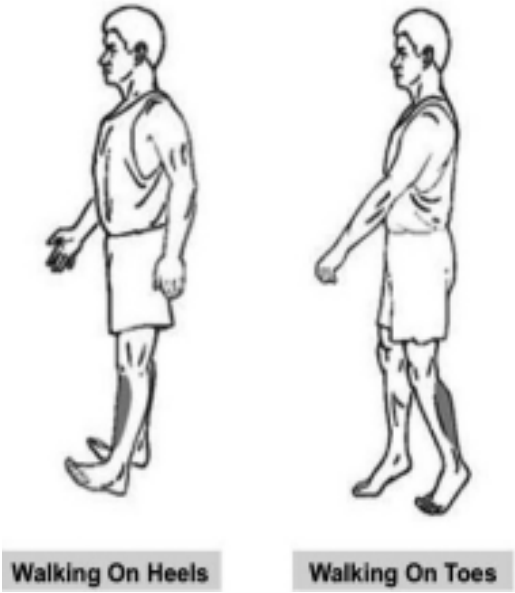


Fig 14.18 : Walking On Heels and Walking On Toes

6. Stepping Over Obstacles - Forward
7. Stepping Over Obstacles - Sideways
8. Practice turning while walking in large circles and gradually making smaller and smaller turns

Fig 14.19: Gait Training in MND

Chapter 15

Activity limitation

Impairment in the activities of daily living (ADL) in motor neuron disease (MND) has been little investigated and focused upon. Activity limitation is dependent on both the behavioral and motor changes that occur throughout the course of the disease; also it is dependent upon the progressive nature of MND. Functional disability is commonly measured using specific scales of ADL. These scales are generally subdivided into basic ADL (BADL) and instrumental ADL (IADL), depending on their complexity.

1) BADL are those related to everyday core tasks such as eating, hygiene, dressing
2) Instrumental activities comprise more complex ones such as meal preparation, shopping, housework and managing finances.

Activity limitation is mainly due to

1) functional impairment: upper limb dysfunction is a major determinant of ADL given their dependence on hand function
2) behavioral changes

   Home assessment plays a vital role in improving the functional independence.

Factors to consider when doing a home evaluation:

1) Focus on the immediate need of the individual and their family with MND regarding functional independence.
2) Cultural and family values
3) Safety factors.

Note: the therapist should make it clear with the individual's family that the needs of the individual will change as the disease progresses and that recurrent evaluations will be needed.

Points to consider when planning a goal for ADL modification:

1) The stage of the disease
2) Value of the individual: computer access and home mobility may be more important for an individual in the progressive stage as compared with seeking assistance for BADL and IADL.
3) Difficulty: The perceived ease and the projected difficulty with which an individual completes an activity should also be discussed.
4) Fatigue and Dyspnoea : Each and every activity which the individual does daily should be analyzed with respect to fatigue level and energy expenditure

5) Safety: individual should be able to analyze the risk factors and find out an appropriate plan for managing it.

6) Pain: pain factors should in considered when selecting an appropriate intervention approach.

Different Approaches used:

- Modify / Compensatory
- Establish / Restore

Modify / Compensatory approach:

Compensatory strategies are used to improve activity performance. If restoration of the previous function is not possible then this approach can be used.

Figure 15.1 Universal cuff

1) Change the way in which the task is performed: E.g. With difficulty in bathing (unable to get in / out of the tub) one can substitute the washing part by doing the activity at sink.

2) Adapt the task objects or prescribe adaptive devices: E.g. Use of enlarge handle spoons or universal cuff for those clients who have poor hand functions (Fig 15.1 Universal Cuff).

   "Individuals satisfaction level should be taken into consideration as sometimes use of an adaptive device decreases the satisfaction with the task performance level of the individual."

3) Modify the task environment: Environmental modification is used to enhance the task performance. E.g. installing ramp for the wheelchair mobility (Fig 15.2 ramp for wheelchairs), Installing grab bars and transfer seat so that client can remain seated while performing bathing activity (Fig 15.3Grab Bars)

Figure 15.2 Ramp for wqheelchair

Figure 15.3 Grab bars

Establish / Restoration approach:

This approach basically focuses on the restoring the lost functions of people with disabilities.

E.g. restoring the functions like muscle strength, endurance, range of motion.

ADLs for individuals with MND

Individuals with MND in the early stage can do ADL activities same as normal individuals; except the fact that they have affected fine motor functions and have poor-fair endurance and they need more time to perform their ADLS. In the progressive stage they may need various adaptive devices or may need minimal to moderate assistance to perform their ADLs and in the advanced stage they will be completely dependent for all their ADLs.

Dressing: It is always advisable to put underwear and trousers when the individual is still in the bed, because of the energy expenditure.

Lower body dressing:

Donning the Pants:

- Lay out your skirt, pants, or underwear as you normally would.
- Lower the garment to the floor. First, slip it over the affected (weaker) leg. Then slip it over the other (stronger) leg.

Figure 15.4 Reacher

- Use the reacher to pull the garment up and over your knees (Fig 15.4 reacher).
- Stand up, with your walker in front of you. Be sure to keep your balance.
- Pull the garment over your hips.
- Sit down to button or zip the garment.

Tips: Gather your clothes, dressing aids, and walker. Place them within easy reach.
Doffing the Pants:

- Sit down to unbutton or unzip your garment.
- Stand up, with your walker in front of you. Be sure to keep your balance
- Pull the garment down over your hips.
- Then push the garment down and over your knees.
- Sit down.
- Lower the garment to the floor. Slip it over the weaker leg first. Then slip it over your stronger leg.
- Use the reacher to pinch the waist of the garment. Then remove it completely.

Note: Your therapist may tell you other ways to dress and undress, based on your needs. Also it will vary in individuals with upper limb and lower limb weakness.

Upper Body dressing:

Donning the shirt:

Figure 15.5 Button Hook

- Keep the shirt across the thighs with back side up.
- Place both arms under the shirt and push the sleeves up.
- By using wrist extensors and shoulder adductors and external rotators along with elbow flexion, pass the shirt over the head.

Once it reaches over the head, keep the shoulders and wrist relaxed.

- Shoulder shrugs and leaning forward along with elbow flexion and wrist extension will help to move shirt down over the body.

- Adaptive devices like button hook (Fig 15.5 Button Hook) or wrist driven flexor hinge splint can be used for doing buttoning. Else individuals with affected hand functions can use fasteners to avoid buttoning.
- By keeping the palms on mattress on either side, maintain the balance of your body, back support is needed for those who have poor balance.

Doffing the shirt:

- Lean forward, duck the head and pull the shirt over the head.
- Remove the sleeve first from supporting arm and then from working arm.

Eating: Different options available for performing eating activity are:

- Use of mobile arm support or externally powered splint (Fig 15.6 mobile arm support) is recommended for individuals in advanced stage who have lost shoulder, elbow and forearm movements
- Use of wrist splint along with the universal cuff (Fig 15.7 wrist splint with universal cuff) in individuals whom grasp is completely gone
- Use of nonskid mat and a plate with a plate guard in the early phase (Fig 15.8 Plate with a plate guard)
- Bilateral or unilateral cup holder (Fig 15.9 unilateral and bilateral cup holders).
- Electric self feeder for individuals in advanced stages of the disease.
- Long plastic straw with a straw clip to stabilize it in the cup (Fig 15.10 plastic straw with straw clips).
- Use of wheelchairs with desk arm (Fig 15.11 wheelchairs with desk arm).
- Use of swing away footrests so that the person can sit close to the table (Fig 15.12 Swing away foot rest).

(Fig 15.6 mobile arm support)

(Fig 15.7 wrist splint with universal cuff)

(Fig 15.8 Plate with a plate guard)

(Fig 15.9 unilateral and bilateral cup holders)

Fig 15.10 plastic straw with straw clips

Fig 15.11 wheelchairs with desk arm

Fig 15.12 Swing away foot rest

Hygiene and Grooming:

- Use of shower and bath tub seat along with the transfer boards (Fig 15.13 bath tub seat).
- Use of long handle scrubber and reacher (Fig 15.14 long handled scrubber).
- Use of bathing mitts for clients who have poor hand muscle strength.
- Universal cuff for doing combing, brushing (Fig 15.15 universal cuff).
- Wall mounted hair dryer (Fig 15.16 wall mounted dryer).
- Use of hand held light weight shower (Fig 15.17 hand held shower).
- Install the grab bars in the bathroom which will increase the safety.
- Use of non skid mat (Fig 15.18 non skid mat).
- Replace doors of the bathroom by shower curtains (Fig 15.19 bathroom shower curtains).

Fig 15.13 bath tub seat (2)

Fig 15.14 long handled scrubber

Fig 15.15 universal cuff

Fig 15.16 wall mounted dryer

Fig 15.17 hand held shower

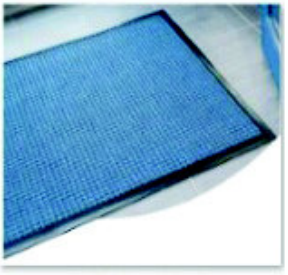


Fig 15.18 non skid mat

Fig 15.19 bathroom shower curtains

Fig 15.20 electronic page turner

Fig 15.21 typing stick

Fig 15.22 specialized mouse

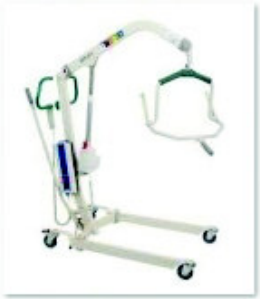


Fig 15.23 hoists

Fig 15.24 sip and puff wheelchair

Communication and Environmental adaptations: for individuals in advanced stages who have lost the upper limb functions completely

- Electronic page turner for turning pages (Fig 15.20 electronic page turner).
- Typing stick for doing typing activity (Fig 15.21 typing stick).
- Use of specialized mouse for computer use in individuals who have few finger movements (Fig 15.22 specialized mouse).
- Use of environmental controls to run tv, radio, telephones.

Mobility and transfers For individuals in advanced stage, hoists are used for doing the transfers (Fig 15.23 hoists), sip and puff wheelchair (Fig 15.24 sip and puff wheelchair), powered wheelchair (Fig 15.25 powered wheelchair)is advisable.

Mobility can be further be enhanced by using electric wheelchairs operated by hand, chin or pneumatic controls.

Home management activities:

Individuals in the early stage can perform light home making tasks by using adaptations and environmental modifications. Energy Conservation Techniques, Work Modification Techniques and Fall Prevention Measures are needed to be focused upon.

a) ECT: Energy conservation techniques include

- Pacing the activities like alternating energy consuming and less energy consuming activities so that there is less of fatigue.

- planning and listing the activities in advance so that the individual is mentally and physically prepared
- finding out short ways to perform an activity
- making unnecessary trips to and fro, avoiding to stand for long , instead sit
- Taking frequent rest periods in between the activities
- Establishing a regular sleep pattern
- Focus on working postures

b) WST: Work simplification techniques include using electrical gadgets and labor saving devices to ease the task. Making the environment friendly and easy to move in. Following are the 3 classes of change that can improve the method of work in individuals with MND.

1) Changes in Hand and Body Motions are done by eliminating unnecessary movements, improving the sequence of work, developing skill in work, comfortable posture. E.g: Carrying several things at once to the kitchen or up and down stairs by the help of trays and baskets reduces unnecessary movements. Working with good posture reduces the expenditure of energy. Poor posture can cause backache, increase fatigue, tension and a lower efficiency. Doing a task with efficient way means saving both time and energy e.g. in bending to do certain tasks, it is easier to put one foot slightly forward and bend through the knee and ankle joint.

2) Changes in works and storage space and equipments includes changes in equipment, changes in the work surface, changes in the storage space.

   E.g.: pressure cooker, mixer, grinder, well sharpened knives, peelers, rice cookers, egg Beaters, chapatti makers, dough mixers and other time and energy saving equipment. The height of kitchen work surfaces should be given careful attention.

3) Changes in the product. E.g.: The use of paper napkins instead of hand towels to minimize the effort for laundering. E.g.: use of washing machine instead of washing the clothes by hand, using an elevator instead of stairs.

c) FPT: Fall Prevention Techniques include both indoor and outdoor modifications: Indoors:

- Keep rooms free of clutter, especially on floors.
- Keep floor surfaces smooth but not slippery or nonskid flooring at home.
- Wear supportive, low-heeled shoes even at home.
- Be careful on highly polished floors that become slick and dangerous when wet.

- Avoid walking in socks, stockings, or slippers.
- Be sure carpets and area rugs have skid-proof backing or are tacked to the floor.
- Be sure stairwells are well lit and that stairs have handrails on both sides.
- Install grab bars on bathroom walls near the tub, shower, and toilet.
- Use a rubber bath mat in the shower or tub.
- Keep a flashlight with fresh batteries beside your bed.
- If using a step stool for hard-to-reach areas, use a sturdy one with a handrail and wide steps.
- Add ceiling fixtures to rooms lit by lamps.
- Consider purchasing a cordless phone so that you don't have to rush to answer it or so that you can call for help if you do fall.

Fig 15.25 powered wheelchair

Fig 15.26 cane

Fig 15.27 walker

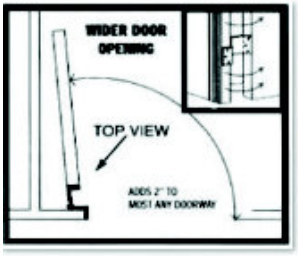


Fig 15.28offset hinges

Fig 15.29 drop leaf board

Fig 15.30 front loading washer

Outdoors:

- Use a cane or walker for added stability (Fig 15.26 cane, Fig 15.27 walker).
- Wear rubber-soled shoes for traction.
- Walk on grass when sidewalks are slippery.

Communication:

- Use a cord less speaker phone or a touch screen mobile.
- Use of short handle reacher to grasp the receiver for individuals who have affected hand functions.

Home management activities:

- Remove the cabinet doors and keep the frequently needed items in the front.
- Use of offset hinges to replace standard door hinges. Which increase the door width by 2 inches (Fig 15.28 offset hinges).
- Use of wheelchair cushion to increase the user's height. Use of lapboard which can serve as a work surface for doing different ADL tasks.
- Use of drop leaf board or a side out board to provide a work surface (Fig 15.29 drop leaf board).
- Use of front loading washers and dryers (Fig 15.30 front loading washer).

To summarize, the ultimate goal of ADL training and ADL modification is to make the individual independent in basic and instrumental activities of daily living leading to successful participation in desired activities and roles. However, in advanced stage, an individual with MND will need a full time help to perform self care activities, for individual's safety should be considered most important. Privacy in all areas of personal care is important and personal space of the individual should be respected. If individual is able to carry out tasks for themselves and do not fatigue easily, this can improve morale and dignity. The support a person receives from their professional team can help them to maintain dignity.

Chapter 16

Hand Rehabilitation in MND

In MND, motor nerves become damaged and eventually stop working affecting the muscles of hands, forearm and elbows. The muscles that the damaged nerves supply gradually lose their strength. The sensory nerves are preserved with intact sensation in the body. The main feature of ALS-MND is muscle weakness which is mild at first, but gradually becomes worse. The first symptoms commonly develop with atrophy and weakness in the hands and progress toward the arms, the shoulders , and the legs, and finally are generalized.

Symptoms to look for:

- Clumsiness while handling objects
- Slipping of objects from hand
- Weak grasp and grip strength (Fig 16.1)

Fig 16.1 Showing weak grasp

- Difficulty in opening bottle tops ,turn keys, buttoning and unbuttoning, etc.
- One may also notice that the muscles in the hands(especially at the base of thumbs) become flatter with time. There is wasting seen in thenar, hypothenar web space and interossei area of the hand (Fig16.2).

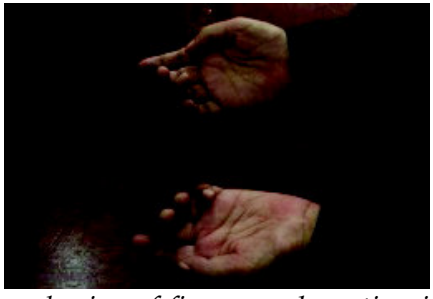


Fig 16.2 Showing clawing of fingers and wasting in hand muscles

- Clawing tendency of fingers
- Wrist drop
- Ape/ Simian Hand

With the progressive weakness and wasting of hand muscles, it is difficult for individuals to perform their regular ADLs. One manner in which Occupational Therapists (OTs) assist patients with MND are planning exercise programs and hand splinting to improve the activities of daily living(ADLs)and improving function. It is important for Occupational Therapist to be aware of the rapid progression of the disease process, to identify the individual's needs and provide efficient interventions.

Wrist, Hand and Finger exercises:

These exercises vary with the various stage of the disease from resistive exercises to active to active assistive exercises: Resistive exercises are performed using the rubber bands, theraputty, clay and manual resistance. Therapist should keep in mind to look for the various trick movements that can mimic the actual movement.

Early Stage: Spring, theraputty and rubber band exercises to improve the strength of the wrist flexors, extensors, finger flexors, interrosei, lumbrical muscles and muscles of the thumb. Also finger flickering exercises to improve on the thumb and finger extensor strength

- Wrist extension (Fig 16.3)

Fig 16.3 Showing wrist extension exercises

- Wristflexion
- Finger bends (Fig16.4).

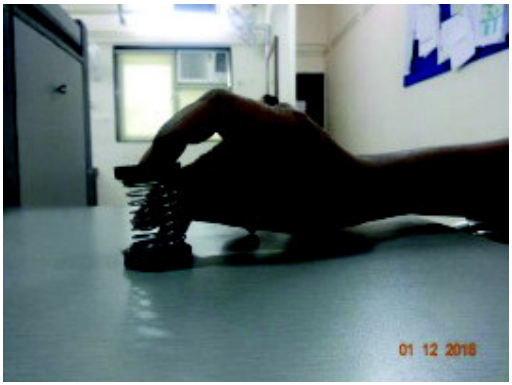


Fig 16.4 Showing finger bending exercises using springs

- Finger spreads (Fig 16.5).

Fig 16.5 Showing finger spread exercises using rubber bands

- Finger squeeze (Fig 16.6)

Fig 16.6 Showing finger squeeze

- Finger-to-thumb touches(Fig 16.7).

Fig 16.7 Showing finger to thumb Spring exercises

- Thumb-to-index finger (Fig 16.8)

Fig 16.8 Thumb to index finger press

- Gripper to improve the grip strength(Fig16.9).

Fig 16.9 Showinggripper to improve the grip strength

- Finger extension exercises (Fig 16.10).

Fig 16.10 Showingfinger extension exercises

- Thumb circles. Use the thumb to make wide circles.
- Thumb towards palm (Fig 16.11)

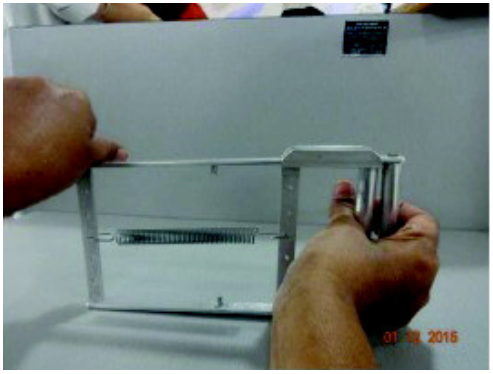


Fig 16.11 Showingthumb towards palm exercises

- Making a fist (Fig 16.12).

Fig 16.12 Making a fist

- Finger towards the palm (Fig 16.13).

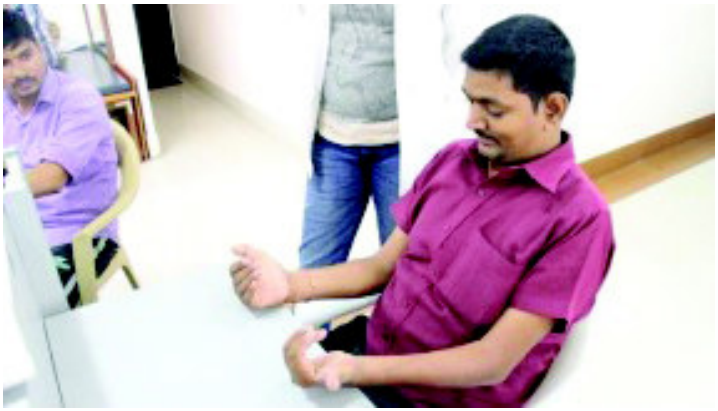


Fig 16.13 Showing finger towards palm exercises

Progressive Stage : Active assistive exercises for the wrist, finger and thumb muscles and splinting to prevent the trick movements and prevent from development of contractures. All the above exercises to be done using assistance from the therapist, family members.

Advanced Phase: Complete Passive wrist, finger and thumb exercises are given to prevent contracture and joint stiffness.

Splinting:

Splinting is done to assist the individual to achieve optimal performance in ADLs

Goals of Splinting:

- To assist in functional activities
- To prevent contractures
- To maintain alignment.

Various Splints required:

1) Oval8 o accustom fabricated circumferential orthosis for weak finger extension and weak hand intrinsic muscles(Fig 16.15)

Fig 16.15- Accustom fabricated circumferential orthosis for weak finger extension

2) Resting forearm based orthosis in intrinsic plus position for night splinting to preventfinger clawing (Fig 16.16).

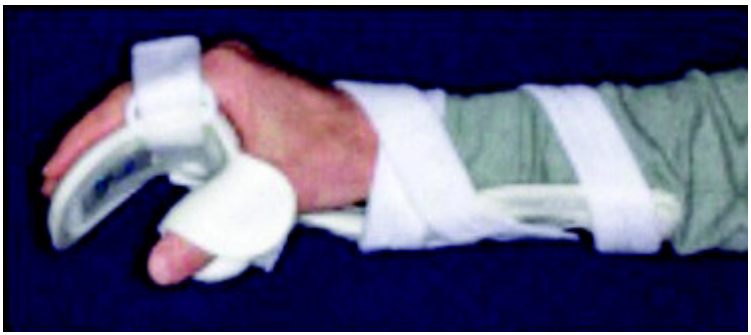


Fig 16.16 Resting forearm based orthosis

3) A light weight Dorsal wrist orthosis to support wrist, is designed to leave the volar surface free for sensory input (Fig 16.17).

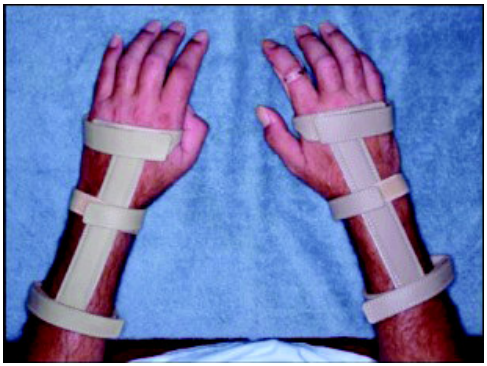


Fig 16.17- Light weight Dorsal wrist orthosis

4) Resting finger extension piece added to a dorsal wrist orthosis(Fig 16.18).

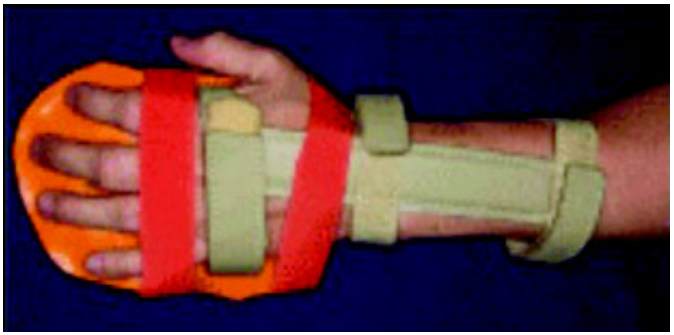


Fig 16.18 - Resting finger extension piece

5) A combined wrist and thumb orthosis(Fig 16.19).

Fig 16.19 combined wrist and thumb orthosis

6) A figure of 8 style orthosis for stabilizing the fingers with weak intrinsic muscles (Fig 16.20).

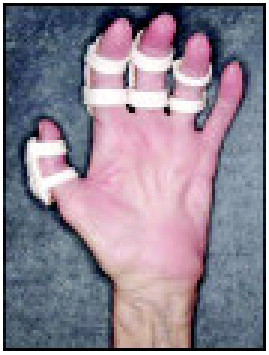


Fig 16.20- Figure of 8 style orthosis for stabilizing the fingers

7) A hand based thumb orthosis with a free IP joint for allowing opposition (Fig 16.21).

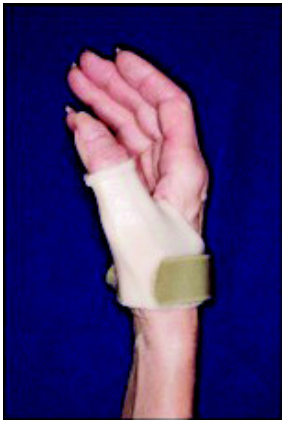


Fig 16.21- A hand based thumb orthosis

8) A hand based thumb spica splint(Fig 16.22).

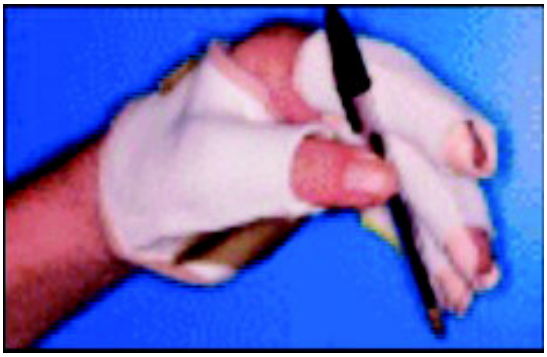


Fig 16.22 - hand based thumb spica splint

Chapter 17

Nutritional management

Amyotrophic lateral sclerosis(ALS) is a progressive neurological disease with high risk of malnutrition. Due to changing disease status diet modification may need to be done frequently. To maintain energy level good nutrition plays vital role. Patient can become deprived of proteins, calories and vitamins very quickly because of feeding difficulties. Early nutritional assessment and intervention is important to prevent malnutrition and enhance quality of life. A Nutritionist can be of a great help in planning a balanced diet & suggest modification according to status of swallowing& other complications.

The symptoms of ALS can affect Nutrition status of patients by two ways:

- The Upper extremity weakness limits patients movements so that cutting food and feeding can be difficult.
- The onset of dysphagia can impairs the swallowing of food.

Five stages of Dysphgia

1. Normal Eating Habits(ALS Severity scale rating- 10-9)
2. Early eating problem (ALS Severity scale rating-8-7)
3. Dietary consistency changes (ALS Severity scale rating-6-5)
4. Tube feeding (ALS Severity scale rating-4-3)
5. Nothing by Mouth (ALS Severity scale rating-2 -1)

1. Normal Eating Habits

- Making early and frequent nutrition assessment and intervention is essential.
- It is right time to start educating patient before development of speech and swallowing difficulties.
- Assessing pattern of normal chewing and swallowing is important.
- It is important to maintain balanced diet and proper hydration.

2. Early eating problem

- Due to masticatory muscle weakness difficulty in chewing and swallowing starts at this stage.
- Coughing while eating results in unusually long meal times.
- Dietary Intervention:
  - ¢ Modification like smaller bite size can be helpful.
  - ¢ Avoiding thin liquids.
  - ¢ Inclusion of foods those are easier to chew and swallow will help.

3. Dietary consistency changes

- As symptoms progress eating dry food becomes difficult.
- Also patients have difficulty in drinking thin liquids especially water which can lead to dehydration.
- Dietary Intervention:
  - ¢ Mechanically soft (foods mashed with hands, finely chopped, or soaked in gravies/curry)food can be given e.g. Roti(Indian bread)dip-in-dal (lentil) gravy or mashed finely chopped vegetables.
  - ¢ Thick liquids can be given e.g. milkshakes, custards, thick fruit puree.
  - ¢ Liquids can be thickened with corn starch e.g. Soups.
  - ¢ Cold foods may be better tolerated as cold temperature facilitates swallowing mechanism.
  - ¢ If difficulty in having mechanically soft food then try with soft cooked food e.g. Soft rice with curry or dal, soft cooked khichdi, soft boiled vegetables, soft cooked chicken.
  - ¢ As difficulty increases pureed foods can be incorporated e.g. rice, lentils and vegetables pureed together, porridge
  - ¢ Small and frequent meals can help increase calorie intake
  - ¢ Vitamin and mineral supplementation may be needed
  - ¢ Fiber can be added along with fluids for relieving constipation
  - ¢ High calorie protein supplement can be included if it's difficult to meet nutritional needs with food
- Special considerations:
  - ¢ Sitting in upright position with head in chin down position will help safe swallowing
  - ¢ Avoid environmental distractions and conversation during mealtime

4. Tube feeding (ALS Severity scale rating-4-3)

- Feeding tube placement
  - ¢ As dysphagia and feeding problems progress providing adequate nutrition orally becomes difficult.
  - ¢ PEG (Percutaneous endoscopic gastrostomy) Tube placement can be considered at this stage.
  - ¢ It is better to put PEG tube once signs of weightloss and dehydration seen than putting it later when malnutrition and respiratory distress progress.
  - ¢ Patients can continue to eat orally even after tube is placed.
  - ¢ Through tube nutritional support can be provided.

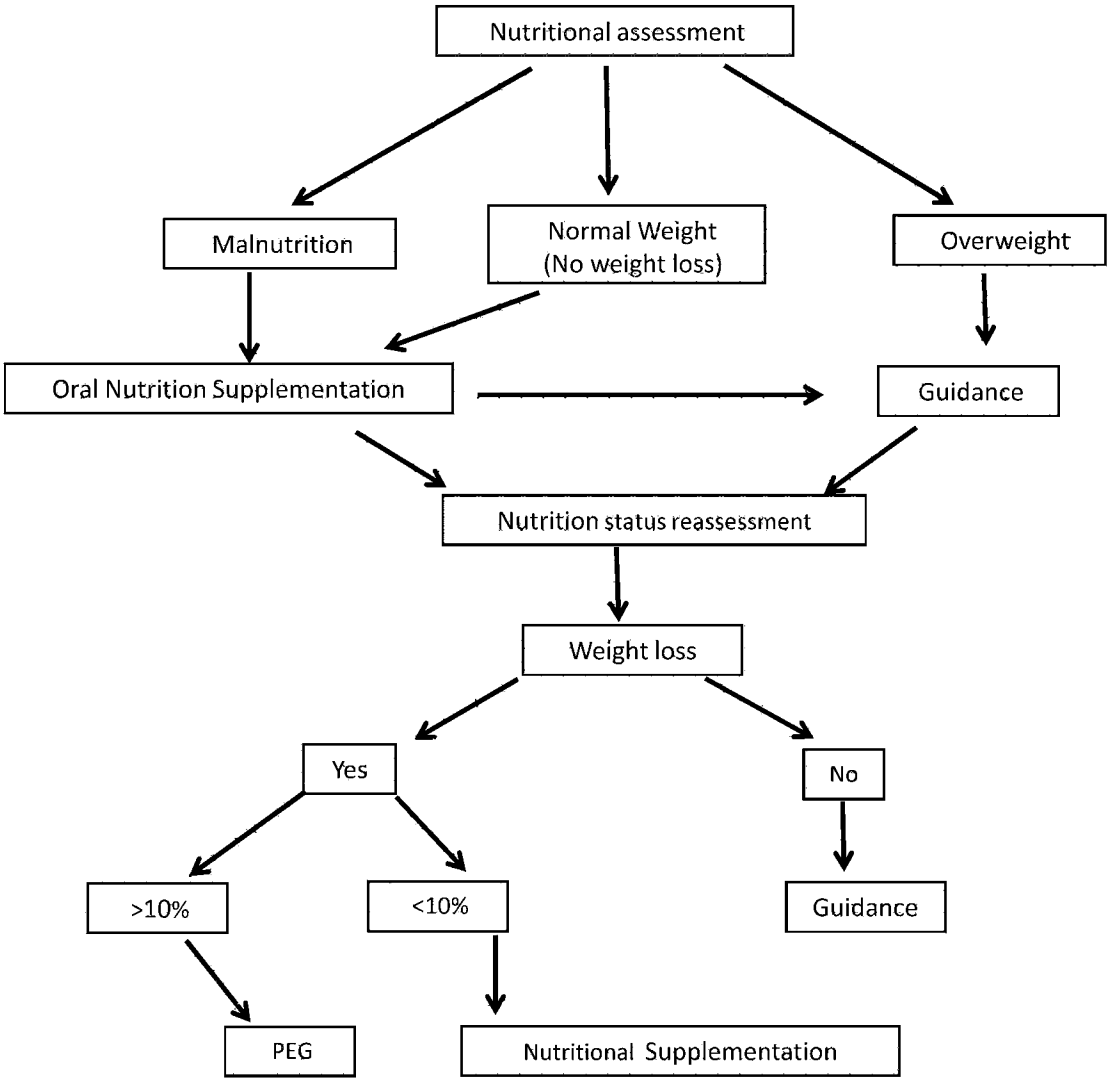
5. **Nothing by Mouth (ALS Severity scale rating-2-1)**

- When final level of dysphagia is reached patients cannot eat anything orally
- Feeding tube will help meet nutritional need at this stage.
- Tube feeding is permanent at this stage

Table 17.1. Correlation of dysphagiaintervention with ALS Severity scale

NORMAL EATINGHABITS
10-Normal Swallowing - Patient denies any difficulty chewing or swallowing. Examination demonstrates no abnormality.
9-NominalAbnormality-Onlypatientnoticesslightindicatorssuchasfoodlodging inthe recesses of the mouth or sticking in the throat.
EARLYEATINGPROBLEMS
8-Minor Swallowing Problems - Complains of some swallowing difficulties. Maintains essentially a regular diet. Isolated choking episodes.
7-Prolonged Time or Smaller BiteSize - Meal time hassignificantlyincreasedand smaller bite sizes are necessary. Must concentrate on swallowing liquids.
DIETARYCONSISTENCYCHANGES
6-SoftDiet-Dietis limited primarily to soft foods. Requires some special meal preparation.
5 -Liquefied Diet - Intake by mouth is adequate. Nutrition limited primarily to liquefied diet. Adequate thin liquid intake is usually a problem. May force self to eat.
NEEDSTUBE FEEDING
4-Supplemental Tube Feedings - PO intake alone is no longer adequate. Patient uses or needs a tube to supplement intake. Patient continues to take significant(greater than50%) of nutrition PO.
3-Tube Feeding With Occasional PO Nutrition- Primary nutrition and hydration accomplished by tube. Receives less than 50% of nutrition PO.
NOTHING BY MOUTH
2-Secretions Managed with Aspirator/Medication-Cannot safely manage any PO intake. Secretions managed by an aspirator and/or medications. Swallows reflexively.
1-Aspiration of Secretions-Secretions cannot be managed non invasively. Rarely swallows.

Yorkston, Strand, Miller, Hillel,& Smith,1993;andYorkston,Miller,&Strand,1995The Swallowing Scale from the ALS Severity Scale.



It is important to meet Dietitian for regular Nutritional assessment and Intervention.

Nutrition Therapy

- Aim of nutrition therapy is:
  - ¢ To supply nutritional needs for all stages of disease progression.
  - ¢ Minimize protein catabolism.
  - ¢ Ensure oral feeding and indicate early nutritional support.

- Diet should be High calorie, high protein ,normal lipids, high fiber and balanced in all nutrients.
- Energy / calories:
  - ¢ Meals should be calorie dense.
  - ¢ Include sheera, custards, puddings, milkshakes, smoothies, fruit pulps, creamy soups (figure 17.1)

Figure17.1 caloriedense food

  - ¢ Make sure cereal pulse combination along with some vegetable is given at mealtime e.g. Mix vegetable Khichdi , Soft rice with lentil & vegetable stew.
- Protein:
  - ¢ Proteins are needed for tissue growth and minimize catabolism.
  - ¢ Eating difficulties lead to poor oral intake and hence less protein intake.
  - ¢ High biological value protein sources should be included like meat and Chicken, eggs, fish, sprouts, dal, oil seeds, soybean, and nuts.
  - ¢ Condensed milk can be included addition of powdered dry fruits will make it calorie dense.
  - ¢ Protein supplements can be added whenever needed(seek advice from dietician before addition of any supplement)
- Lipids:
  - ¢ Lipids will add calories to diet.
  - ¢ 25-35% daily energy requirement can be provided by lipids.
  - ¢ Inclusion of omega 3and Omega 6 is important.

- ¢ Sources include almond, walnuts, olive oil, mustard oil,canola oil,flaxseeds, cod liver oil/fish oil.

- Fiber:
  - ¢ Because of worsening dysphagia fiber intake is restricted.
  - ¢ It is important to add modified consistency fiber rich food to patient's diet.
  - ¢ Include cereals in porridges.
  - ¢ Include vegetables, pulses in boiled and mashed form.
  - ¢ Include fruit puree like papaya ,oranges.
  - ¢ Whenitbecomesdifficulttomeetfiberrequirementorallyfibersupplement can be added.
- Water
  - ¢ There is risk of dehydration in ALS patients as with progressiveDysphagia it is difficult to drink thin liquids.
  - ¢ Foods with high liquid content can be included like fruit juice, fruit and vegetable puree, smoothies (Figure 17.2)

Figure 17.2Thickliquids (fruit juice vegetable puree)

- Vitamins and Minerals:
  - ¢ Accumulation of Free radicals and oxidative stress has been proposed as factor that contributes to progression of disease.
  - ¢ Supplementation of vitamins and minerals therefore important.
  - ¢ Vitamin D & calcium
- Clinical research findings supported a neuroprotective role for vitamin D and a role as a reliable prognostic marker of ALS.
- Studies show lower level of Vitamin D in ALS patients.
- Vitamin D is generated within the body in response to adequate sunlight. Hence exposure to sunlight is essential
- Vitamin D rich food like egg yolk, liver, oily fish can be included
- Vitamin D Supplementation may be helpful.

- Calcium rich food should be included like milk, cheese, yogurt, milk pudding, and sesame seed, rajgira seeds.
- All fresh fruits and vegetables & nuts are rich sources vitamins and minerals and therefore important to include in diet.
    - ¢ Vitamin B12
        - O Scientists postulated that Vitamin B12 could help regenerate nerves.
        - O Food sources of Vitamin B12 include milk & milk products, meats, cheese, eggs, fish etc
    - ¢ VitaminE
        - O VitaminEis potent antioxidant
        - o Deficiency of Vitamin E is associated with progressive neurological deterioration.
        - O Several studies describe improvement in ALS patients with Vitamin E supplementation
        - o Sources of Vitamin E includes almonds, Hazel nuts, sunflower seeds, sunflower oil, safflower oil
    - ¢ VitaminC
        - O It plays important role in transmission of signals between neurons.
        - o Vitamin C supplementation may help protect respiratory dysfunction.
        - O Sources include orange, sweet lime, guava, papaya, amla.

Special considerations

- Try to include food from all food groups.
- Try and modify consistency of food as per patient's tolerance.
- Give enough time to chew and swallow then only feed next bite.
- Make sure patient is well hydrated.
- Consult your dietitian at intervals.
- Nutritional assessment at intervals is necessary to check for any malnutrition and need for tube placement.
- Give small and frequent meals.

Special considerations for PEG feeding

- Always feed the patient in propped up position(Figure17.3).
- Do not give very hot or very cold feeds.
- Strain each feed twice before giving

- Do not mix feed and medication together
- Flush the tube with water after every feed.
- Do not give very thick consistency feeds it may block the tube.
- Give Nutrition supplement as per dietician's instructions.

Figure 17.3 PEG Tube feeding

Sample dietchart (Softdiet) MENU PLAN FOR INDIANS (NONVEG)

Meal pattern	Food group	Units	Sample Menu
MORNING:	Milk	1/4unit	Tea/Milk/coffee
BREAKFAST:	Cereal	2units	Rawa (semolina) porridge 1Bowl
	Milk	1/2unit	Tea/Milk/coffee
	Fat	1unit	Forcooking,1tsp
	Lean meat	2unit	Omlette,1(2 Egg Whites)
MID MORNING:	Fruit	1unit	Apple (small pieces)
LUNCH:			Soup
	Cereal	3units	Roti, 2 no (Dipped in curry)
			Soft rice Rice,1/2cup
	Vegetable	2units	a) Peas bhaji (soft cooked)
			b) Grated boiled carrot
	Lean meat	3units	Small pieces Grill fish, 3pc/fish curry
	Fat	1unit	For cooking, 1 tsp
MID AFTERNOON:	Milk	1/4unit	Tea/Milk/coffee
	Nuts	1unit	2 almonds, walnuts (powdered)
EVENING:			Watermelon small piece
DINNER:	Cereal	2units	Soft Rice,1cup
	Pulse	1unit	Dal, med,1cup
	Vegetable	2 units	Palak bhajee (puree)
			Boiled beet salad
	Lean meat	1 unit	1 cup chicken curry
	Fat	1 unit	For cooking, 1tsp
BEDTIME:	Milk	1 unit	Cold milk 1 cup

Sample dietchart(Softdiet)MENU PLAN FOR INDIANS(VEG)

Meal pattern	Food group	Units	Sample Menu
MORNING:	Milk	1/4unit	Tea/Milk/coffee
BREAKFAST:	Cereal	2units	2Idli
	Pulse	1units	sambhar
	Milk	1/2unit	Tea/Milk/coffee
	Fat	1unit	For cooking,1tsp
MID MORNING:	Fruit	1unit	Apple (small pieces)
LUNCH:	Cereal	3units	Roti, 2 no (Dipped in curry)
			Soft rice Rice,1/2cup
	Vegetable	2units	a) peas bhaji (soft cooked)
			b) grated boiled carrot
	Pulse	1units	Dal fry
	Fat	1unit	For cooking,1tsp
MID AFTERNOON:	Milk	1/4unit	Tea/Milk/coffee
	Nuts		2 no almonds, walnuts (powdered)
	Pulse	1 unit	1 cup soft cooked sprout bhel
DINNER:	Cereal	2units	Soft Rice,1cup
	Pulse	1unit	Dal,med,1cup
	Vegetable	2units	a) Palak bhaji
	Pulse	1unit	1 cup dal tadka
	Fat	1unit	For cooking,1tsp
BEDTIME:	Milk	1unit	Cold milk 1 cup

Chapter 18

Psychological impairments

Motor Neuron Disease is a progressive neurological condition which affects the individual not only physically, but emotionally and psychologically as well. The fact that the patient cannot perform his daily activities as he/she used to before, creates a significant impact on the patient's mental well-being. The first and fore most emotion an individual experiences after receiving the diagnosis is severe anxiety and depression. There are numerous questions going on in the patient's mind, with the most frequent thought being-Why Me? There is anxiety and fear about the future and feelings of helplessness and hopelessness. Many patients also undergo behavioural changes such as apathy, disinhibition and impulsivity which can lead to considerable care giver burden as well. Therefore, it is extremely essential to take into consideration the patient's and his family's psychological state of be in gat every step of the treatment. Family, friends, caregiver, the rapists and a Psychologist can playa very important role in helping the patient achieve maximum recovery and emotional stability in the process of rehabilitation.

Initial Reactions: Depression is usually experienced in reaction to the diagnosis or challenges of the disease. A diagnosis of alifethreatening disease such as MND can be devastating for anybody. Therefore, it is important for the concerned physician or Neurologist to break the news with as much sensitivity as possible. Mentioning the diagnosis to the family, instead of the patient in the beginning might help. It has been observed that many patients tend to give upon life immediately after getting to know about their condition from their physician. The family can break the news with utmost are and emotional support for the patient. His initial reaction of shock and anger can be handled by the close family members or a psychologist, if need be.

For some patients, it might also be a sense of relief after a long period of uncertainty. It is not uncommon to feel arrange of mixed emotions as one can experience more than one emotion at a time. The patient can feel:

- Shock
- Disbelief about the diagnosis and denial
- Fear of the future
- Anxiety related to family, spouse and children
- Anger at the physician or any family member; blame game
- Intense sadness, withdrawal and apathy
- Guilt for feeling like a burden to the family
- Feeling overwhelmed.

Emotional ability / Pseudobulbar affect: Emotional ability is one of the symptoms of MND where in the patient may find it difficult to have a control over his emotions. He may laugh un control ably or feel extremely sad and start crying. This can become quite embarrassing for the family members. Some patients might also lose their temper quickly. However, it is vital to realize that the patient is not doing this on purpose. Giving some time and space will help the patient deal with this condition in a more effective manner. In many cases, this is a temporary condition and reduces with time. Not all patients with MND experience emotional liability.

Depression: As the initial reaction so shock and anger subside, the patient may gradually tend to feel sad and experience depression. The patient might stop communicating, keep himself withdrawn, lose appetite, experience difficulty in sleeping and lose interest in daily life activities. It is vital for the family to notice the see changes and offer help. Talking about the fears and concerns with a closed one or a professional counsellor and receiving counselling can goal on way in dealing with depression at times, it is also important to give the patient some space and allow him to deal with his own emotions. Constantly trying to talk and asking questions may backfire and lead to anger outbursts in the patient.

Coping Strategies for Depression:

- Stays focused and have small goals
- Talk it out
- Challenge negative thinking
- Listen to happy music
- Practice relaxation techniques
- Meditate every day for10 minutes in morning and 10minutes before going to sleep
- Spend some time in nature
- Take professional counselling

Intimacy and Sexuality: Physical intimacy forms a very important part of any relationship between couples. If one of the partners is affected with MND, it creates assign if cant impact on the aspects of intimacy and closeness in the relationship. MND does not affect sexual function, but the difficulty and inability in the physical movements end to make sexual expression difficult. Especially patients with arm or leg weakness find it difficult to enjoy physical contact and sex. Therefore, the most important thing in such a situation is having an open discussion with the partner. Intimacy does not necessarily mean having inter course. But gentle touch and being close to the partner can play a significant role in showing affection. It is essential not to focus on areas that are associated with sexual pleasure, but to learn about the whole body, each other's preferences, likes and dislikes. As the disease progresses in the patient, the partner

might need to take a more active role sexually. It is helpful to talk about this in advance, and explore different sexual positions or activities such as mutual masturbation, massage, oral sex or using sexual aids. While trying something new, it is important to have an open mind, patience and a willingness to laugh together to keep the intimacy intact.

Rethinking and Planning: Every individual has some goals and ambitions in life and a plan to accomplish them. However, a diagnosis of MND can suddenly alter the direction of a patient's life. He is expected to make the necessary changes and come up with new plans. It will be helpful to become aware and informed about then a true of the disease, and plan things accordingly. This way the patient can help himself have a better quality of life. The more a patient is in denial about the disease and its impact, the more he will delay the process of taking control and managing the illness. MND is always unpredictable and for some patients, the progression can be quite rapid. Therefore, it is important to take decisions as early as possible. Preparing oneself for the future, forex: anticipating the use of a wheel chair or a walker or using compensatory mechanisms taking into consideration the course of the disease will goal on away in helping the patient continue as normal life as possible. Being more proactive and changing questions like "Why Me" into "What can I do" will help the patient cope up better in accommodating life style changes.

"We cannot direct the wind but we can adjust the sails."

Speech and Communication : One of the most important function affected in MND is Speech. Due to weakness in the muscles of the tongue and lips, the patient finds it difficult to speak. The speech becomes slurred, slow, unclear and indistinct making it difficult for the family to comprehend. As a result, the patient tends to become frustrated and loses his motivation to talk. He then withdraws himself from having any communication and interaction. In order to deal with this situation, the family and the clinician must encourage the patient to make use of technology. Other ways of communicating can be as follows:

- Speak slowly
- Use short sentences to save energy
- Use of gestures and hand movements
- Avoid back ground noise
- Pause more often while speaking
- Use more of keywords

As the speech problems tend to increase and the patient loses his ability to talk, the above ways of communicating will not be possible. Therefore, the patient needs to:

- Work out some hand signals and actions for frequently used interactions or sentences.
- Respond in Yes / No through blinking of eyes or raising 1 finger or hand.

- Compile at least of regular questions and requests and ask the family to indicate on the list.

Advice for Family /Care give : As the patient's speech starts deteriorating, the family or the caregiver of the patient also finds it difficult to communicate with him/her. However, the below mentioned points need to be considered while making an effort to communicate or understand what the patient is trying to say:

- Keep the tone, speed and volume of the speech same.
- Impaired speech does not mean impaired understanding. Therefore, the family needs to be very careful while speaking in front of the patient.
- Face one another and notice the patient's face and lip movements carefully.
- Keep a pencil and paper handy.
- If the patient gets fatigued easily, give him time to pause, take rest and then speak.
- This will require a lot of patience and understanding.
- Use questions that can be answered only in Yes/No.

Personal Factors: As mall clinical study was done on the identification of Personal factors in MND in the year 2011 in Australia, by the Department of Medicine,

Royal Melbourne hospital. Rates of depression and anxiety are reported to be 0-44% and 0-30%, respectively, in persons with MND. It has been previously reported that QoL appears to be more dependent on "psychological and existential issues, social support and spirituality" rather than physical factors. These findings were supported by Chiòetal who found that the main determinants of quality of life in MND were social support, depression, religiosity, and

Socio economic status. The importance of coping strategies in the experience of MND is further supported by Gallagher and Monroe who surmised that MND is not a static disease but a progressive disorder that required different coping strategies at different stages of the disease. Matuzetal also found that the be stpredictors for the severity of depressive symptoms in MND were perceived social support (especially a supportive marital relationship) and coping potential (information seeking and strategies of emotional avoidance behavior). For example, a combination of confronting and avoiding coping strategies might be useful for MND patients, because search of information and support may help them to initiate actions that ensure optimal future care. On the other hand, emotional avoidance behaviour (e.g., choosing isolation or denial) could protect them from psychological distress and despair. However, as the disease progresses, avoidance is no longer an adaptive strategy, as it prevents patients from taking appropriate measure to cope further with the illness.

"I am not what happened to me. I am what I chose to become"- Carl Gustav Jung

The pre-morbid personality of a patient also plays a significant role in the experience of MND. Personality traits such as optimism, flexibility and humor play an important role in helping the patient cope up well with the disease. Certain belief systems such as fate having a control over one' she all have also been found to be prominent. A strong will and determination can goal on way in helping the patient cope up well with the illness on a day to day basis. Preparing health goals for the future will give a new direction to the patient and he will have something to look forward to in life.

"Our greatest weakness lies in giving up. The most certain way to succeed is to try just one more time"

Palliative Care: According to the WHO definition of Palliative Care, the aims of care for people with MND should be to reduce the effects of the disease on the patient and their family, maintain the patient's ability for as long as possible, and enable the patient and family to live life as fully as possible Ideally, palliative care should begin at the time of diagnosis or after a few months of the diagnosis when the patient has come to terms with the illness. It is important to establish the patient's wishes in regard to end of life care and preferred place of death while they are still able to communicate easily. Communication may become extremely difficult but eye pointing or single response answers to closed end questions can be maintained.

Chapter 19

Pharmacological Management

Pop the pills only with prescription.

Riluzole: Disease modifying drug

Disease modifying drugs are drugs which may modify the progression or the course of the disease by all eviating the symptoms. As presently there is node finitive cure for ALS modifying the disese course can be the best possible treatment provided to an individual. Riluzole is known to modify the course of the disease and increase the quality of life in individuals with ALS. It is an europrotective drug. Only riluzole has been approved for treatment of ALS. Riluzole provides improvement in both bulbar and limb function, but not in actual strength of muscles. It delays the onset of ventilator- dependence or tracheostomy in selected patients and may increases revival by approximately two to three months.

Mechanism of action

Riluzole can suppressglutamates action (a chemical messenger in the central nervous system).Glutamate is released during nerve impulse transmissions. Excessglutamate has been shown to cause brain and spinal cord nerve damage.

Dosage

Thestandarddoseis50mgtabletstwotimesaday.Precautionsshould betakenforlow weight individuals.Please consult your doctor and take the medicine on his prescription.

Side effects

Liver toxicity, abnormal skin sensations around the mouth; diarrhea; dizziness; drowsiness; headache; loss of appetite; muscle weakness; nausea; stomach pain; vomiting; weakness. Liver function, Blood and creatinine should be tested before starting riluzole therapy.

Contraindications:

- known prior hypersensitivity to riluzoleor any of the excipients inside the preparations,
- liverdisease,
- pregnancyorlactation

Availability

The drug is freely available from superpharma on doctor's prescription.

A shaek hope foundation can help the patients to provide aprescriptionde pending on their need of the drug.

Vitamin E

Vitamin E supplements in ALS as an additive to riluzole treatment or as adjunctive therapy can show better neuro protective effects.

Dosage: 400mg

Vitamin C

Vitamin C has antioxidant effect and can help to increase the immune level.

Dosage: 500 mg

Vitamin B 12

Vitamin B 12 should be taken in high dosage as a supplementary nutrition .

Dosage: 1000 mg

Calcium with Vitamin D

Calcium with Vitamin D is required to prevent secondary complication of osteoporosis as the individual less exposed to sun because of his activity limitation and is less mobile in compare to normal.

Multivitamins

Multivitamins should be taken as a supplementary nutrition as the food uptake is reduced which leads to deprivation of many nutrient factors.

Dosage: 1 tablet daily

Coenzyme Q 10

Co Q 10 is a critical component of the electron transport chain of mitochondria. It exhibits antioxidant properties .

Omega 3 fatty acids

They are antioxidant which can be beneficial.

Protein Supplementation

Proteins are required for muscle building. Therefore is ALS protein supplementation by high protein powder is recommended to improve energy level and reduce fatigue.

Symptomatic management

1. Muscle relaxant

   Carisoprodol, metaxalone, and cyclobenzaprine are the muscle relaxant which

can be used to relax the muscles in case of excessive spasm.

Side effects: confusion, lethargy, dry mouth, fatigue, light headedness, constipation or blurred vision.

2. Anti spastic agents

   Quinine, baclofen, carbamazepine Benzodiazepines (Diazepam, chlorazepam, lorazepam) Phenytoin can be used. This also help store life spasm and spasticity in the muscles. In some patients with severe cramping, botulinum toxin injections might be helpful, but they must be carefully administered to prevent further weakness.

3. Painkillers

   Analgesics include paracetamol (acetaminophen), tramsdol, non-steroidalanti-inflammatory drugs (NSAIDs) such as the salicylates (asprin) can be used to decrease the pain.

4. Antianxiety drugs

   Benzodiazepines like Chloradezepoxide, Diazepam, chlorazepate, florazepam can be prescribed for anxiety and panic attack.

5. Antidepressants

   Patients with depression and insomnia (decreased sleep) tricyclicantide pressant can be used. Fluoxetineandsertra line can also be prescribed.

6. Drooling

   Glycocorpyrrolate, benztropine, atropine and trihexyphenidyl hydrochloride can decrease the saliva production.

7. Respiratory management

   Aspiration pneumonia can be a very common problem because of swallowing issues. Antibiotics Ceftriax one plus azithromycin, levofloxacin, ormoxifloxacin are appropriate choices Nebulisation with bronchodilators can help to loosen the secretions. Salbutamol and steroids can be used. Cough suppressants (codeine, dextromethorphan, and noscapine). cough suppressants, are substances which suppress the coughing itself Expectorants (acetylcysteine and guaifenesin). Decongestants (ephedrine) relieve nasal congestion. Steam in halation is very useful to loosen the secretion and relax the bronchial tract.

8. Gastrointestinal management

   Antacids are acid-reducing drugs like H2-receptor antagonists (ranitidine) orprotonpumpinhibitors (omaprazole, pentaprazole) which can be used to decrease the acid level of the stomach.

   Vomiting, nausea : Domperidone, dimenhydrinate and meclizine hydrochloride

For Constipation stool soften like, laxatives (duflac), suppositories (delocolax, glycerine) can be used. High fiber diet and hot water or milk can be used.

Swallowing difficulties: Percutaneous endoscopic gastrotomy is the best option when the bulbar muscles become weak. You should always try to avoid rhyl's tube for swallowing deformality and is not the permanent option for that.

9. Urinary urgency

Don't hold the urine for long hour sand maintain a habit of urinating every four hours. Urine potsorbed pancanbeused during limited mobility and in advanced stages andc an save the energy in case of urgency and maintains the hygiene also. Maintain proper hygiene and if the urine is leaked or passed please change the dress or bed sheets immediately to avoid infections and sores. Check for infections. Urinary tract infection can be treated by lots of fluid intake and a correct dosage and type of antibodies.

Chapter 20

Yoga

Yoga- a way to health, energy and peace

Yoga is an ancient Indian mind body technique, which is becoming increasingly popular throughout the world because of its several health benefits. Yoga is an integrated system of self culture which aims at harmonious development of body, mind and covers all aspects of human life that lead to physical well being, mental harmony culminating into positive thinking, happiness and peace.

Pranayama practice in the yoga schedule might have a role in reducing the oxidative stress. As an example, diaphragmatic breathing has been reported to reduce oxidative stress by decreasing cortisol that which inhibits enzymes responsible for the antioxidant activity of cells and by increasing melatonin , which is a strong antioxidant.

Many published research studies indicate that patients who undergo yoga therapy show significant improvement in oxidative stress and inflammatory markers. Continued efforts should be made by researchers and clinicians to educate regarding the benefit of yoga therapy in order to improve TAOS, reduce ALSA, RER and inflammatory markers.

Yoga Therapy

As we have seen in this book, that management of debilitating and degenerative diseases like ALS involves multifold aspects. It is generally a combination of conventional medical treatment with rehabilitation techniques. The importance of rehabilitation techniques such as physiotherapy, occupational therapy and psychological rehabilitation cannot be over emphasized. All of the above therapies prove to be significant in managing the conditions arising out of the underlying neurodegenerative disease such as ALS. But as with all forms of treatment, there is always room for improvement. This therapeutic gap can be filled by an ancient Indian wisdom for treatment of various conditions of the mind and body, namely Yoga Therapy.

Yoga for ALS

Inclusion of a therapeutic practice like yoga, in tandem with physiotherapy and occupational therapy can help mitigate the conditions of ALS and greatly improve the quality of life. Yoga therapy stretching and relaxation of various muscles, in combination with deep breathing techniques to improve muscle tone and reduce pain. Research shows that the benefits of yoga for movement disorders include improved strength, flexibility, balance, overall fitness and quality of life.

Each individual has a different degree to which ALS has caused physical and neurological degeneration. Muscle spasms, atrophy and rigidity associated with

movement disorders often restrict balance and range of motion. Limits to balance and range of motion restrict the ability of individuals with movement disorders from practicing traditional yoga poses in a way that is beneficial to them. Yoga for movement disorders is marked by a practice that addresses the needs particular to people living with movement disorders. Yoga therapy needs to be customized according to the patient after studying his medical history in detail. Each asana has a specific purpose and helps specific areas of the body.

Thus not every asana can be prescribed to every patient in general. This is one of the main reasons that parents must help the individual suffering from ALS in practicing Yoga on a regular basis. Also, it is of absolute importance to consult a yoga expert to evaluate which asanas can be recommended to the individual, in order to facilitate his/her existing physiotherapy.

While physiotherapy is important to increase muscle strength, the entire exercise can be tiring and painful. But Yoga helps in reducing the physical and mental stress. Deep breathing techniques help in infusing more oxygen into the blood as opposed to regular shallow breathing. This improves blood circulation, strengthens the weakening muscles and helps in the removal of toxins that accumulate in the body. Yoga also addresses the mental and emotional damage caused by the illness. It helps to infuse positive energy into the individuals and thus help them to increase their inner strength to fight off depression & anxiety that characterize such illnesses. It inducts a surge of hope into the individuals.

Under modern conditions of physical stress, multiplying distractions and mental strain, what is equally ,if not more essential than exercise for the health of the nervous system is recreation, relaxation and sleep. This is because it is a fact that every bodily power needs rest after exertion. It's also known that even machines cannot operate without rest. The heart rests between beats, and the muscles relax after every contraption. All bodily movements and for the matter even the mental activities involve same form of essential or subtle muscular contractions.

During waking hours, thus, we constantly pass through a series of complicated muscular contractions, which result from multifarious nerve stimuli. The high tension of modern living further adds to the nervous strain and invariably devitalizes both the nervous and muscular systems. Besides recreation and sleep ,therefore the power to relax whenever fatigue is experienced is probably only most important safeguard one can possess to keep these systems in their normally healthy conditions.

We will illustrate here some of the simple yet powerful asanas that help in ALS.

Pranayama

Prana means Life and Ayam means control. Pranayam means control of the inner force of humanlife. The breath we breathe in and out is regarded as Prana which means

bioenergy that endows man with ultimate potential for self-development. It is the vital/ lifeforce. But man must suitably control and channelize the prana or use it for right end. Yoga prescribes various practices of Pranayama or control of Prana popularly referred to as breathe control. The yogic breathing itself becomes a prayer, a satisfying spiritual experience in which one is aware of the living presence of God.

Benefits of Pranayama

1) Better blood circulation.
2) More oxygenation.
3) Longeveity i.e .full healthy life.
4) Mental concentration.
5) Increase in lung elasticity and capacity.
6) Purifies blood.
7) Emotional control.
8) Cheerfulness.
9) Prevention to diseases (many cases cured also).

Life is a series of activities. We conclude one activity only to start another. A preparedness kriya allows a person to be mentally prepared for an activity.

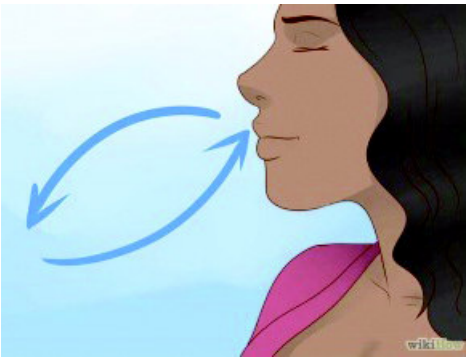
There are 3 types of Pranayama which we describe here:

Pranayama (I): Equalization of inhalation& exhalation; Technique:

- Sit firmly and comfortably.
- Breathe in out for equal counts.
- Breathe 10 times.

Benefits:

- Augments pleasant feelings throughout whole body.
- Helps to calm the mind.



Pranayam (II): Intercostal Breathing

Technique:

- Hands on the side of the chest.
- Make chest rise up as you breathe in for 3 seconds and fall as you breathe out for 3 seconds.
- Repeat 10 times.

Benefits:

- Activates the abdominal organs, provides a gentle massage, releases flatus and reduces fat in the abdominal region (best for ALS because fat can be reduced).
- Helps in respiration and relaxation.
- Useful in insomnia.

Pranayama (III) - Anulom Vilom

Technique:

- Sit in an asana for preparedness.
- Use the little finger to block the other nostril. Breathe out from the open nostril.
- Do same for the other nostril.
- Repeat 10 times.

Benefits:

- Sedative effect on the tone and rhythm of the heart & brain.

Stretching

In ALS limbs are always weak, if one cannot to do the asanas properly, then patients have to stretch only to the extent they can. But stretching is compulsory for patients with ALS. In Yoga, slow stretching is very beneficial. Patients need to do the stretching regularly.

Asanas for ALS

1. Sukhansana

   Sukhansana is one of the many asanas which quieten the mind. This asana is named for preparedness or conditioning.

Technique:

- Spread the legs forward.
- Fold the left leg and place the feet near the right thigh muscles.
- Similarly, bend the right leg and push it in the space between the left thigh and calf muscles.
- Now you will find the two feet between the thighs and calves of the legs.
- Breathe normally.

Note: ALS patients have to sit in a comfortable pose. If patients are notable to sit, they can do the asana by laying down.

Benefits:

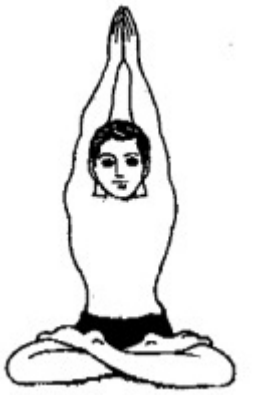
- Amplifying your state of serenity and tranquility.
- Broadensyourcollarbonesandchest.
- Calming your brain.
- Eliminating stress and anxiety.
- Improving alignment.
- Reducing fatigue.
- Strengthening your back.
- Stretching your ankles and knees.

2. Parvatasana

Technique:

- Sit in comfortable pose.
- Both the hand should be on both sides of the body.

- Inhale for 3 seconds ,raise both the hands simultaneously upward and above the head, palms facing upward.
- Keep the elbows straight & joint the palms.
- Pull the abdomens lightly inside. Hold breath.
- Exhale for 3 seconds, bring the arms down.



Benefits:

- Stretches all the abdominal and pelvic muscles.
- Loosens the hip joints.
- Helps to reduce the flatty and flabby abdomen.
- Improves the shape of body. (It is very beneficial for ALS).

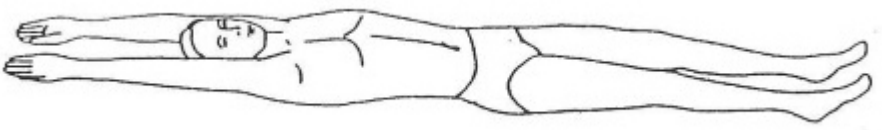
3. Yastikasana

Technique:

- Lie on back legs fully extended and arms extended at the side.
- Be relaxed, inhale and raise arms above the head, rest them on the floor and stretch.
- Holding breath, slowly stretch the body at full length, the toes and fingers pointing outward, as if trying to reach out. (Any attempt at maximum stretching of the body should be only during retention of breath).
- Repeat 3 to 4 times with in-between pose.

Benefits:

- Corrects faulty posture.
- Tenses the usually relaxed abdominal and pelvic muscles.
- Offers relaxation, when maintaining an on-stretch, passive attitude.
- Removes spasm.



4. Shavasana:

Shavasana is recommended for complete relaxation of the body and mind.

Technique:

- Lie supine on the floor. Consciously relax different areas of the body starting with the toes

- Next relax the ankles
- The knees
- The thighs and fingers together
- The genital area
- The anus
- The abdominal area
- The chest region, the shoulders
- The neck
- The chin
- The mouth
- The tip of the nose
- The space between the eye brows
- The forehead
- The mind

Benefits:

It is intended to revitalize and rejuvenate the body, mind, and spirit.

There are 3 main benefits of Shavasana which are described in detail below:-

Relaxation

Post the exertions of physiotherapy or other asanas, shavasana allows the body a chance to reset itself. Yoga asanas stretch, contract, twist and even invert all the muscle so the entire body in a well balanced and ergonomic way. Shavasana which is normally done as the last asana helps the deepest muscles to let go and shed their regular habits.

The physiological benefit of deep relaxation can be numerous and may include the following.

- a decrease in heart rate and the rate of respiration
- a decrease in blood pressure
- a decrease in muscle tension
- a decrease in metabolic rate and the consumption of oxygen
- a reduction in the number and frequency of anxiety attacks
- increase in energy levels and productivity in general
- improvement in memory
- increase in focus and concentration

- reduction of fatigue
- deeper and sounder sleep
- higher self-confidence and better morale. Revitalisation &Resetting of the nervous system

A good yoga practice will feed the nervous system with a host of new neuromuscular information. Shavasana gives the nervous system a chance to integrate that internally before it is forced once again to deal with all the usual stresses of daily life. Think of it like a brief pause that the nervous system takes to process all the information.

Higher Concentration and Inner focus

Shavasana is the beginning of deeper, meditative yogic practices. During the performance of this asana the practitioner's awareness is turned inwards and purified of sensory distraction. In state of sensory withdrawal it becomes easier to be aware of the breath and of the state of the mind itself.

Shavasana is a good way to reduce stress and tension. On the other hand, yoga-nidra ("yogic sleep") meditation is often practiced in a lying position. Drowsiness or restlessness of the mind while in shavasana may be counteracted by increasing the rate and depth of breathing.

||??|| *[Patanjali Yoga Sutras]Tatrasthitauyatno'bhy?sa?*||*13*||

Abhy?saorpractice(abhy?sa?)istheeffort(yatna?)toattaintothat(tatra)Sthitior mental peace (sthitau)||13||

Face Yoga

Be the change you wish to see in the world." Mahatma Gandhi ji

- AiyanaFaceYoga":Dynamic Muscle Resistance program. A special technique of Asana/ Exercises that increase blood circulation to the 57 muscles of the Face, Neck and Scalp. Act on and stimulate the central nervous system, brain and endocrine glands through facial muscles.

Purpose:

1. Increases Circulation to the Neck, Face and Scalp.

2. Applied rehabilitationusing Facial muscles.
3. Increase cell Nutrient and Oxygenation.
4. Isolating themuscles that control the mouth with various exercises can have a positive effect on overall speech quality for those suffering from a disease that weakens the muscles. Utilizing various facial exercises help build muscle tone.
5. Stress management and anxiety control.

Facial Exercises

The "O"

Open your mouth wide, for man "O" with your mouth. Fold your lips over your teeth. Hold

Lips tight and smile. Hold 15 counts.Relax and repeat 4- times.

Benefit : The Round muscles around the mouth comprise the Foundation of half of your Face. Tightening, toning and strengthening is "The O". Lifts and firms the cheeks, jaw and round muscles of the mouth.

The "Sour-Kitten"

Close your lips hard,tighten the corners of your mouth. Hold 10 counts.

Smile, stretch your lips (sour smile), imagine pulling the corners of your mouth towards your ears . Hold the position for 15-20counts.repeat 5 times.

Benefits : Tones and strengthens round muscles of the mouth, cheek, lip and jaw muscles.

The Balloon"3 Finger

Fill air in the cheeks(like blowing a balloon), place your fingers on your mouth. Using your facial muscles, forcefully try to displace the fingers covering our mouth. Breathing , Hold for 10count.Repeat.

Benefit : Strengthening muscles used for laughing, cheeks, chin and the jaw area.

Breathing and Toning with Flexion and Extension

Sit straight ,exhale forcefully. Touch the chin to your collar bone. Now slowly stretch your neck to the ceiling, inhaling as you commence stretching. Completely, fill your lungs with air. Hold 5 counts. Now commence exhaling as you start bring your chin

downwards to touch your collar bone once again. Hold the position 2counts.Repeat the entire set (upwards and downwards), 7 repetitions.

Vizulisation

Sit straight, close your eyes. Imagine a golden light flowing into your face.Now it is filling your cheeks, your chin, your neck. As it fills you can swallow easily, the saliva is not drooling. You can hear yourself speak easily, your muscles of your mouth are moving easily. You sit still and feel the clouds of gold flowing in and out of your face. Slowly when you are ready, open your eyes.

Note : Consult a Face Yoga expert prior to using the exercises.

SECTION C
Care at Advanced Stage

Henry Agard Wallace

(October 7, 1888 - November 18, 1965)

Henry Agard Wallace was the 33rd Vice President of the United States (1941-45), the Secretary of Ariculture (1933-40), and the Secretary of Commerce (1945-46).

When Wallace became ill with ALS he kept a detailed report of his deteriorating condition in hopes it would help scientists' understand the disease. He first experienced the onsets of amyotrophic lateral sclerosis in 1964. He died in Danbury, Connecticut, on November 18, 1965. He remained a scientist to the end of his life.

Wallace left a lasting influence on American agriculture as a scientist, an agriculturalist, a journalist, a cabinet member, an elected politician, an author, an economist and a statesman. In December 1999 the Des Moines Register named Henry A. Wallace the "Most Influential Iowan of the 20th Century." In 1966, one year after his death, he received the Iowa Award. This is the highest honor the state of Iowa bestows on a citizen who has been an outstanding leader and made outstanding contributions to their field throughout their life and career.

Chapter 21

Advanced respiratory care in ALS

As ALS progresses, the person living with the disease may become increasingly dependent on ventilation and ultimately, will require invasive ventilation with tracheostomy. This will provide more efficient ventilation and better control of the upper airway and secretions.

Many people with ALS ultimately need invasive ventilation because of the weakness of the mouth and throat (bulbar) muscles. Invasive ventilation is thought to be a more reliable means of delivering air to the lungs when the disease is advanced. Not everyone with ALS will need or choose to have a tracheostomy, but there may come a time when it's necessary for continued breathing.

Ventilation through tracheostomy

A tracheostomy is a surgically created hole in the trachea (windpipe) through which air is forced. The tube through which the air is delivered also is called a tracheostomy (trach) tube. A ventilator delivers air on a timed cycle through the trach, and ensures that the patient will take a minimum number of breaths per minute. Many ventilators can then be adjusted to respond to the person's own efforts to breathe, or to completely override these efforts.

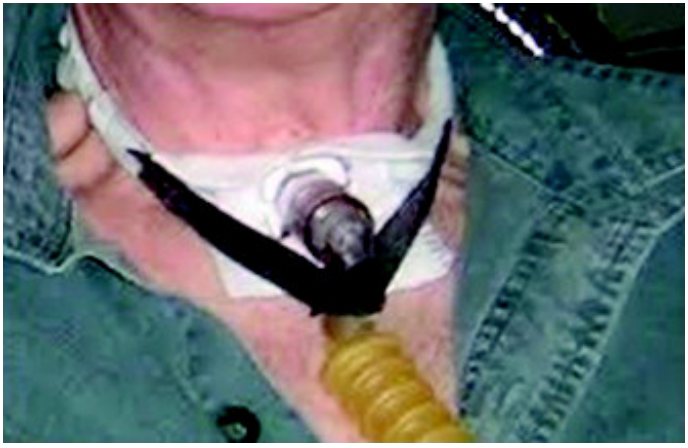


Figure 21.1-Tracheostomy tube

Advantages of invasive ventilation:

1. It provides for much longer survival
2. It provides a secure connection directly to the airway for suctioning secretions.

3. It leaves the face free,without headgear, straps, and skin pressure problems on the pace.

Disadvantages of invasive ventilation:

1. The patient must have specially-trained individuals (reliable family / friends) able to handle advanced life support equipment, or skilled nurses (LPN or RN-level), in very close attendance, at all times: 24x7.
2. Continuous suctioning is required as secretions can block off the tracheostomy tube, often at night as well as during the day.
3. Most insurances do not cover this kind of expensive nursing care at home.
4. Patients on Invasive Ventilation often lose the ability to eat or drink, and require a feeding tube inserted in the stomach for their nutrition and hydration.
5. Patients on Invasive Ventilation often lose the ability to speak and will have to use assistive technology to communicate.
6. Patients usually require a motorized wheelchair with a special tray to hold their respiratory equipment and require a wheelchair-accessible van to travel.
7. Weakness continues to progress in the rest of the body, with eventual paralysis of all voluntary muscles. At somepoint, people with ALS will progress to a totally "locked- in" state, unable to communicate with the outside world in any way.
8. Family members / friend's lives can become disrupted and deferred. Quality of life for caregivers may decline significantly.
9. Personal and family financial resources can be consumed.

Decision making regarding invasive care

The decision to choose invasive mechanical ventilation is a very personal one. People with ALS who choose Invasive Ventilation can live for years. Some of them are cared for in nursing homes while others are able to remain at home. As a general rule, it takes 4 to 5 people committing to "full-time"(40+hours/week) to provide care to someone on Invasive Ventilation at home.

So, in making the decision, a patient living with ALS must consider his family support, level of independence, insurance coverage and financial resources.

Tracheostomy and its care

A tracheostomy is a surgical procedure to create an opening through the neck into the trachea (windpipe). A tube is usually placed through this opening to provide an airway and to remove secretions from the lungs. This tube is called a tracheostomy tube or trach tube.

Self-care

The patient will have a small amount of mucus around the tube. This is normal.The hole in the neck should be pink and painless. It is important to keep the tube free of thick mucus. The patient should always carry an extra tube in case the tube gets plugged. During coughing, it is better to have a tissue or cloth ready to catch the mucus coming from the tube. The patient's nose will no longer keep the air moist. Consulting with the doctor about how to keep the breathing air moist and how to prevent plugs in your tube. One of the most common ways to keep the breathing air moist is to put a wet gauze or cloth over the outside of the tube to keep it moist. A few drops of salt water (saline) will loosen a plug of thick mucus. Putting a few drops in the tube and windpipe, then taking a deep breath and coughing will help bring up the mucus.

Protect the hole in the neck with a cloth or tracheostomy cover when outside. These covers can also help make the breathing sounds quieter.

During showers, cover the hole with a tracheostomy cover. No swimming is allowed with a tracheostomy tube.

To speak, the patient will need to cover the hole with a finger, a cap, or a speaking valve.

Caring for the Tracheostomy

Once the hole in the neck is not sore from the surgery, clean the hole with a cotton swab or a cotton ball atleast once a day to prevent infection.

The bandage (gauze dressing) between the tube and neck helps catch mucus. It also keeps the tube from rubbing on the neck. Change the bandage when it is dirty, atleast once a day.

Change the ribbons (trach ties) that keep the tube in place if they get dirty. Make sure the tube is held in place while changing the ribbon. Two fingers should be fit under the ribbon to make sure it is not too tight.

When to Call the Doctor

Medical attention is warranted if the patient has:

- Fever or chills.
- Redness,swelling,or pain that is getting worse.
- Bleeding or drainage from the hole.
- Cough or shortness of breath, even after suctioning the tube.
- Nausea or vomiting.
- Any new or unusual symptoms.

LTV® 1150 ventilator

- Provide your adult and pediatric patients pressure control and pressure support without a high pressure oxygen source
- The LTV 1150 ventilator provides portable, advanced ventilation for adult and pediatric patients at home or a post-acute care facility
- At only 14 lb, the ventilator provides a wide range of ventilation therapies to meet demanding patients needs, including volume control, pressure control, pressure support and spontaneous breath types

Withdrawal of care

For termination of ventilator support, a careful, thoughtful approach is necessary. There needs to be a complete discussion of the decision and its pros and cons. Extensive counselling is to be done to ensure that the patient understands the ramifications of the decision. Depression, if diagnosed, should be treated accordingly.

People using Invasive Ventilation may elect to discontinue this medical intervention, going back to their natural state as if they had not used it. This is done in a supported (usually hospital) environment to keep them comfortable. Reducing and discontinuing this support means the person will pass away, due to the effects of ALS on the body, usually within hours.

Chapter 22

Percutaneous Endoscopic Gastrotomy (PEG) management

What is PEG?

Percutaneous endoscopic gastrostomy (PEG)is a method of placing a tube into the stomach across the skin and subcutaneous tissues aided by an endoscope. It provides a reliable route for nutrition and hydration in ALS patients with dysphagia (difficulty in swallowing).

PEG is also used to establish an enteral access for medication and supplements, and for decompressing the bloated gut.

How is it done?

Different variations of the technique used by gastroenterologists include the popular 'pull' method, and the 'push' method. The patient is kept fasting for atleast 4 hours. A mild sedative is given intravenously. A check endoscopy is done. The stomach is then filled with air via the endoscope. Once a good spot is selected on abdominal wall, the skin is sterilized and draped. The procedure of placement of the tube then follows after a local anesthetic is injected in skin. The skin procedure is then followed by the endoscopic procedure and the selected tube is introduced and secured on both sides of the stomach and abdomen with bumpers.The internal bumper is checked via the endoscope.The external bumper is kept 1 to 2 cms away from the skin.The feeding adaptor is then fixed to the tube outside. Feeding of the patient can start 4 hours after procedure.The entire securing device of the feeding tube is covered with gauze. The gauze is never inserted between device and skin. The feeding tube itself is looped and taped to the abdominal wall. When not, feeding tube is kept closed.The PEG kit contains all the necessary items and is commerciallyavailable.

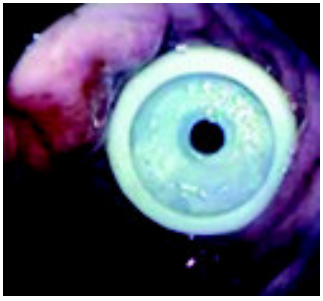


Figure 22.1 Endoscopic view of internal bumper

Figure 22.3 The PEG tube insitu

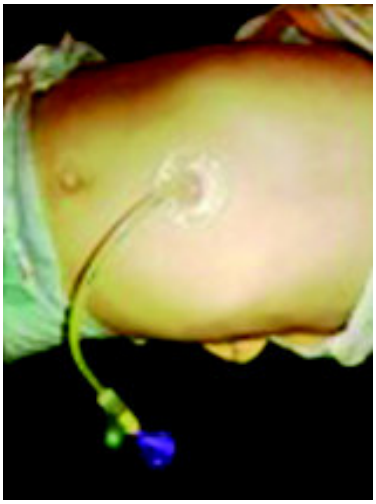


Figure 22.2 - Covered with gauze

Figure 22.4 - Feeding through tube

When do doctors avoid doing the procedure?

The contraindications are: if there is a skin infection at the site selected for PEG, if there is an obstruction in the food-pipe or if the patient has had multiple abdominal surgeries done in the past for fear of adhesions, or if the patient is too sick for any procedure.

What are the precautions you have to take after PEG is done?

The PEG insertion site must be cleaned daily with soap and water and then covered with gauze.The feeding tube must be flushed and aspirated after every meal. An appropriate syringe is used for feeding. No tugging on the feeding tube should be allowed. When in doubt about either the placement or patency of the feeding tube call your doctor. Remember always to feed in a propped up position.

What can happen after PEG procedure?

Immediate complications are aspiration or allergic reactions or bleeding at the site of insertion. These rarely happen because the doctors are very careful.

Late complications are peristomal leakage, if the covering gauze keeps getting soggy call your stoma care nurse or doctor. Do the same if there is an obvious infection at the insertion site.Sometimes the external bumper may get buried in the abdominal wall, show it to your doctor.

What usually scares all patients is the thought of an inadvertent removal of the feeding tube, if that does happen don't panic, call your doctor or stoma care nurse. Don't try to fix things yourself!

Why is there a necessity to have a PEG done at all?

It has been proved that early management of nutrition in patients of chronic neurological disorders helps alleviate symptoms. Very often patients have continuous or intermittent difficulty in swallowing leading to poor nutritional status and generalized weakness. Placement of PEG tube is better than insertion of a nasogastric tube for a long period of time as the risk of aspiration is high with the latter. Many patients and their families sometimes have very decided views about the placement and use of feeding tubes; theyare worried or overwhelmed about the care needed of the stoma created, the care of the device and of preparing the right consistency meals. Remember, doctors usually counsel patients to follow a particular form of therapy for the over all good of the patient, and to give him a better quality of life.

Chapter 23

Home and nursing care at advanced stages of ALS

Chronic diseases like ALS requires care, long-term monitoring of patients as they are dependent for activities of daily living. Since it is not possible to carry out monitoring and care in hospital environment, they should be carried out at home. For this reason, there is a need for quality home care services.

I. Skin care

- Range-of-Motion (ROM) exercises to prevent contracture and pain in joints 3-4x per day; first Active ROM, then passive.

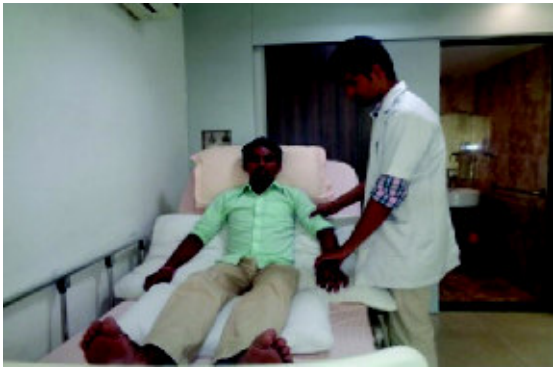


Figure 23.1 Exercises should be done to prevent tightness of the joint

- Use as many different positions as possible when in bed. Change positions every two hours, or on skin tolerance. After each change of position, check for redness over bony prominences to prevent ulcers.

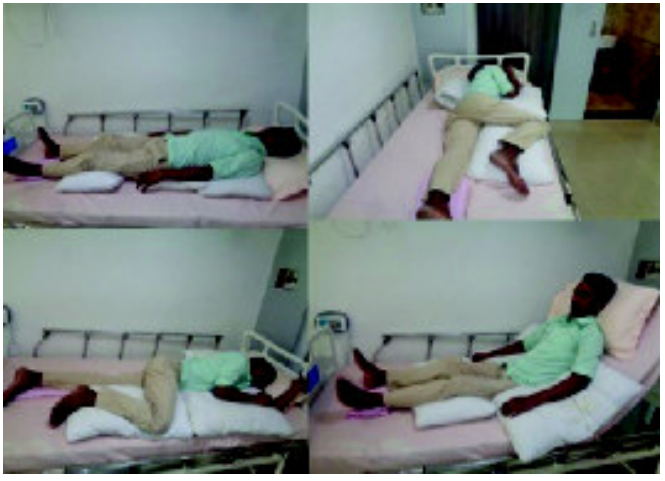


Figure- 23.2 To prevent bedsores, patient positioning in the bed should be changed every 2 hours

- Repositioning in the wheelchair based on the patient's skin tolerance. Use of a wheelchair cushion to prevent skin breakdown
- Promote adequate nutritional intake.
- Keep skin dry.
- Lotion dry areas.
- Air flotation or egg crate mattress to bed.
- Elastic stockings, if ordered, especially when patient out of bed.

II. Urinary care and bowl management

- To prevent the urinary tract infection following encourage daily fluid intake of 2,500 cc's per day unless contraindicated because of swallowing ability.
- Catheterize or when necessary
- Wash skin and dry well after each bowel movement or urination (especially females).
- Consult physician if there are signs of urinary tract infection
- For bowl management encourage patient to keep daily record of bowel movements (include consistency and amount), particularly if there has been change to a more constipated or less frequent stool.

  Provide dietary instruction regarding the importance of fiber, use of bran cereals, prune juices, etc., unless contraindicated because of swallowing ability.

  Daily use of stool softeners and Metamucil are often recommended. Laxatives may be used, but chronic use may be harsh on the bowel. Mineral oil should not be used because of the danger of aspiration.

Figure 23.3 -Urine Bag

Figure 23.4- Catheter

III. Respiratory care

1. Maintain adequate ventilation
   a) Deep breathing and coughing exercises and use of incentive spirometer to encourage lung expansion.
   b) Chest physiotherapy which include assisted cough and postural drainage when indicated.
   c) Oxygen, when ordered.
   d) Elevation of the head of the bed with a foam wedge or hospital bed to ease shortness of breath and provide comfort during sleep.
   e) Auscultate chest frequently to assess total airway states.
   f) Help patient to cough and deep breathe at least every two hours.

Figure 23.5 - Portable home ventilator

2. Maintain airway patency

a) Use of aspirator to suction secretions and prevent choking.

b) Use of medications (i.e., Elvail to reduce secretions).

c) If the patient has a tracheostomy, suctioning and "trach care" should be done to prevent buildup of secretions, possible obstruction and infection. The caregiver or family members need to be trained for monitoring and suctioning if needed.

3. Provide information and opportunities to discuss options for mechanical ventilation. Generally, if there are early signs of dysphagia, dyspnea, or after the patient has

had time to adjust to his diagnosis, the patient and family should be assisted in considering options regarding life support systems. The important thing to remember is the patient should be thinking about these decisions before he/she needs them.

IV. Nutritional care- Nutritional Management of the Dysphagic Patient

1. Promotion of optimum weight
   a) Smaller, more frequent meals.
   b) Foods that are "easy-to-eat", i.e., adjusted mechanical soft diet.
   c) Increased daily activity with non-fatiguing exercise.
   d) Liquid supplement between meals.
   e) High protein, high CHO diet
   f) Soft, mechanical soft or pureed diet as needed.
2. Oral feeding may become a high risk activity due to increased risk of obstruction of the airway, aspiration and inability to meet nutritional requirements.
   a) Percutaneous endoscopic gastrostomy (PEG) has become the intervention of choice when eating becomes exhausting, difficult or nutritional goals are not being met because oral intake is time-consuming and onerous.

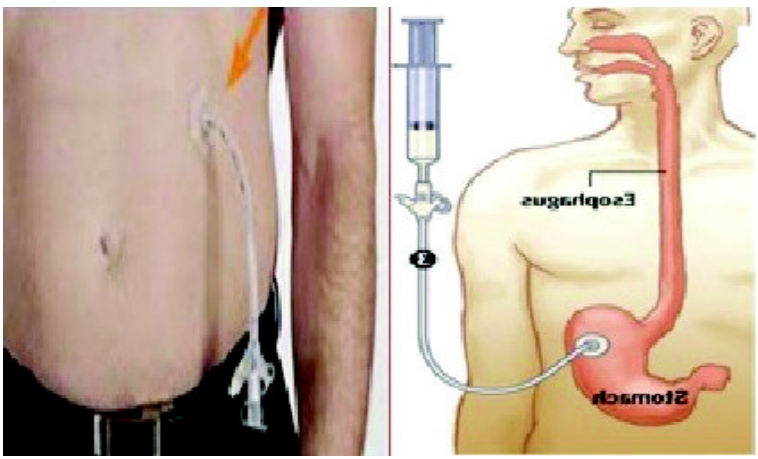


Figure 23.6 PEG tube and feeding

SECTION D
Recent Advances

Peter Frates

(December 28, 1984-)

Peter Frates is a former Boston College baseball player who was diagnosed with amyotrophic lateral sclerosis (ALS) in 2012. He is credited as the creator of the Ice Bucket Challenge, an activity involving dumping a bucket of ice water on one's head to promote awareness of ALS that went viral on social media during the summer of 2014. Upon diagnosis, he immediately began advocacy and fundraising work for the disease.

The Frates ALS Research and support Fund was recently founded by Pete and his family to provide funding for targeted ALS research and to help support those afflicted with ALS who have not been fortunate to receive the support that Pete has. The fund was created to carry on his and Team Frate Train's mission to raise awareness and assist in discovering a treatment and a cure for ALS.

Chapter 24

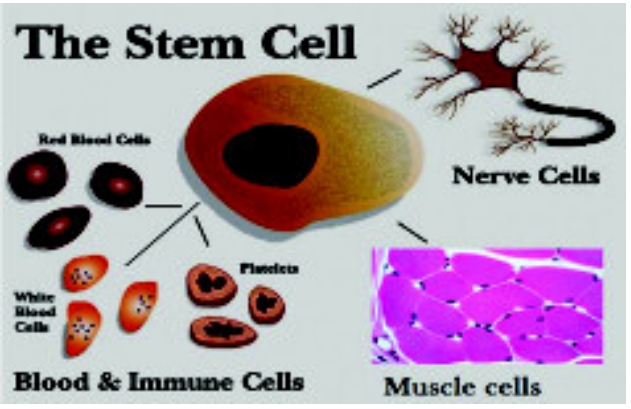
Stem cell therapy in Motor Neuron Disease

Introduction

We all know that currently there is no cure available for MND/ALS. But we have to make efforts to at least slow down or halt the progression of the disease. Attempts need to be made to improve the symptomatic problems. Unfortunately, we do not have any medicine which can do this. But recently stem cell therapy has brought hope in the treatment of ALS/MND.

What are Stem Cells?

Stem cell therapy is being explored extensively for MND/ALS. Experimental studies in animals have shown promising results. Some of the clinical studies in human beings have shown preliminary results of beneficial effect in the disease progression.



Stem cells are unique specialized cells that help to repair, regenerate and replace damaged cells. Stem cell therapy (also called cell therapy/ regenerative medicine) works on the principle of using healthy cells to repair damaged cells. Stem cells are considered to be "blank slates", which means they are programmable.

Types of stem cells

Stem cells are divided into two broad groups.

1. Allogenic (Stem cells taken from outside)
2. Autologous (Stem cells taken from the patient itself)

Allogenic stem cells are further divided into: a) Embryonic Stem Cells; b) Umbilical Cord Stem Cells.

Embryonic stem cells have various ethical and safety issues. Therefore they are not being used in the majority of centres.

Autologous stem cells are divided into: a) bone marrow derived stem cells; b) adipose (fat) derived stem cells; c) peripheral blood stem cells.

Autologous stem cells are extremely safe to use because they are from the same patient.

They are easily available and have no ethical issues.

Induced Pluripotent Stem Cells (iPSC) : These cells can be autologous or allogenic. These are genetically reprogrammed cells. They are called pluripotent as they can differentiate into many types of cells. Currently they are used only in laboratory due to risk of side effects.

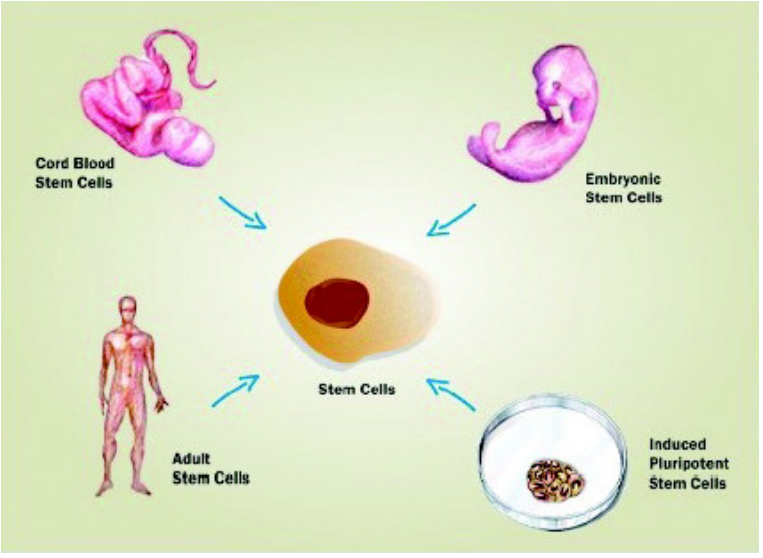


Figure 24.1: types of stem cells.

Mechanism of action of stem cells.

Stem cells act by neuroprotection (Protect the existing motor neurons) and neuroregeneration(making new neurons and other cells). They do this by the following mechanisms:

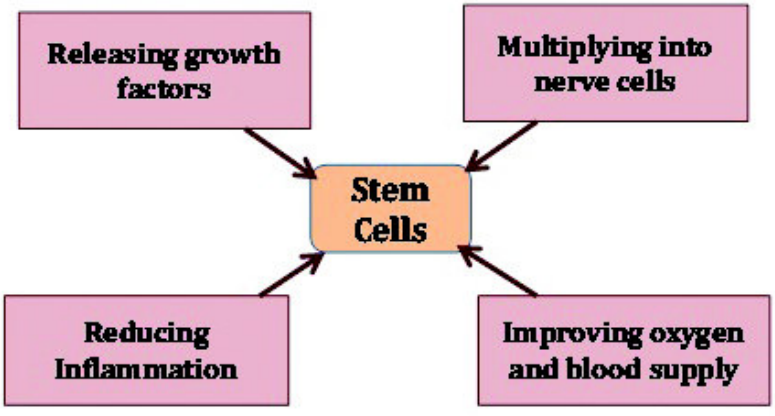


Figure 24.2:mechanism of stem cells

- Releasing certain positive chemicals known as nerve growth factors.
- Improving the oxygen and blood supply to the damaged parts of the nervous system by a process called angiogenesis (making new blood vessels).
- Multiplying and differentiating into nerve cells.
- Producing chemicals that decrease the inflammation and modulate the immune system.

Processing of stem cells

Any stem cell, irrespective of the source from where they have been obtained, has to be processed before using them for intervention. There are different protocols of stem cell therapy; some may require only separation ; some may require separation and culturing. The processing can be carried out in following steps:

1. Separation: Separating stem cells from its source is one of the most important steps in the process of stem cell therapy as it is crucial to gain optimum benefit from the cells. The protocol for processing stem cells is varied.

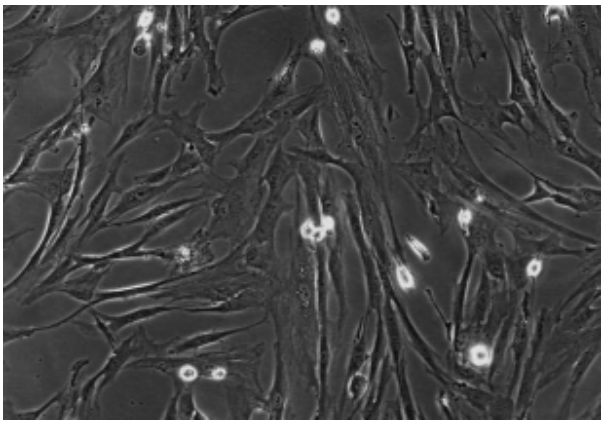


Figure- Microscopic picture showing bone marrow stem cells growing into neurons

2. Culturing: After separation, the cells are cultured in laboratory to increase the quantity of cells. Different media are used to culture different cells. However, purification of these cells is important and has to be done cautiously. Also, there is a risk of infection while culturing the stem cells.

   Although some may prefer injecting the cells directly after separation. In this procedure, the risk of impurities being present is minimal.

Routes of administration of stem cells

Stem cells can be administered through various routes such as intrathecal, intramuscular and intravenous.

1) Intrathecal route: Injection of stem cells into the spinal fluid through an injection at a lower back level (L4-L5space.) This method is minimally invasive and is the safest targeted mode of transplantation.

2) Intramuscular route: Injections of stem cells directly into the affected muscles. This creates "local depots" of implanted cells with increased local action. To inject the stem cells, motor points are marked on the muscles. (Motor point is the junction where the nerve enters the muscle).

3) Intravenous route: Is by a simple injection of stem cells into the veins like any other drug injection. It is one of the safest, minimally invasive and most widely used routes of administration. But this is a very generalized method where very few cells reach the targeted motor neurons in the spinal cord. So this is not a very effective mode of transplantation.

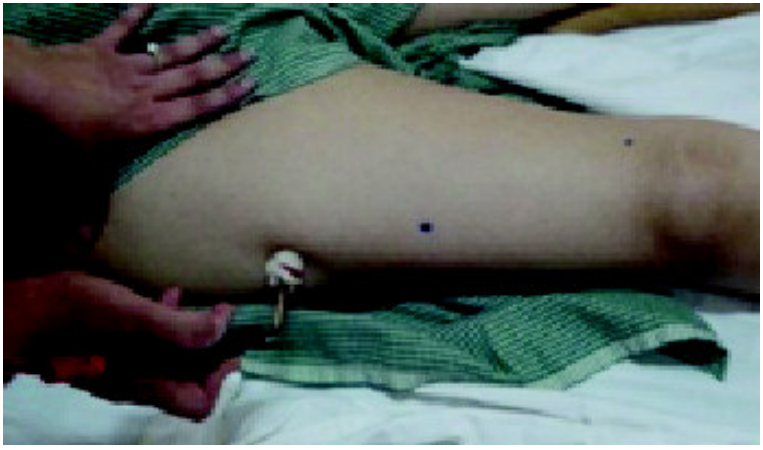


Figure 24.3: plotting of motor points

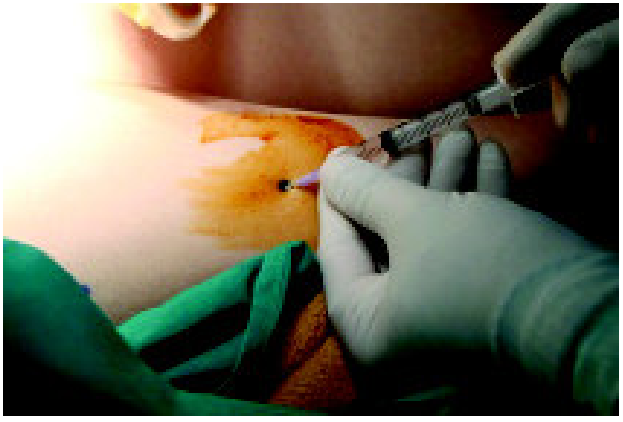


Figure 24.4: intramuscular injection into the quadriceps muscle.

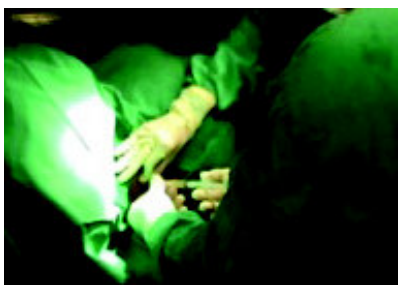
Stem cell therapy at NeuroGen Brain and Spine Institute

We have treated more than 100 patients of MND at NGBSI. The cells are injected via intrathecal route of delivery. The procedure for stem cell transplantation at NGBSI is minimally invasive, with simple steps or processes. There is no major surgery or incisions required.

The procedure is carried out in 3 steps:

a) **Bone marrow aspiration:** 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.

b) **Separation of stem cells:** On the same day, within 3-5 hours, stem cells are separated from the bone marrow and purified in the laboratory.



c) **Stem Cell Injection:** Stem cells are then immediately injected into L3-4 or L4-5 space. An 18G Toughy needle is inserted into the spinal fluid (intrathecal space). After establishing a free flow of CSF, an epidural catheter is inserted into the space and the stem cells are injected into the cerebrospinal fluid (fluid which flows around the brain and spine).

Safety of 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 related to the cells. Some procedure related side effects like nausea, vomiting, fatigue, spinal headache may be experienced. The procedure is more safe in the early stages and non-bulbar stages of MND/ALS.

Role of rehabilitation after stem cell therapy.

Following the stem cell transplantation, from the very next day, patient undergoes an intensive rehabilitation process, consisting of physiotherapy, occupational therapy, speech therapy, psychological therapy, positive reinforcement processes, yoga therapy, etc. It is important that stem cell therapy be followed by a proper exercise regime to gain maximum benefits.

Rehabilitation augments the effects of stem cell therapy. Scientific evidence supporting this theory is also available. Our long term follow-up reveals that patients who participate in a regular rehabilitation program do overall better than those that don't. The availability of the transplanted stem cells makes the rehabilitation process more effective and efficient.

Need for stem cell therapy

Current management strategies for MND

As multiple body systems are involved, the best approach is a multidisciplinary holistic management of the disease. Currently MND is treated with a combination of medical, surgical and rehabilitative treatments [7].

Pharmacological management

Mainstay of pharmacological management is Riluzole. Riluzole inhibits the release and modulates the activity of Glutamate which is found to cause excitotoxicity in MND. It is the only medicine that has shown significant effect on the survival duration in MND [8,9,10]. Other medicines like Talampanel, Memantine, Cephalosporins, antioxidants, agents modulating apoptosis, anti-inflammatory medicines like Thaliomide and Celastol and Autophagy regulators like rapamycin and lithium are being tested with no conclusive results [11].

Non Pharmacological management

With no pharmacological means of altering the disease process current management of MND is widely palliative and symptom oriented. Regular moderate intensity physical exercise [7], and in the later stages of the disease artificial respiratory support [12] dysphagia management and percutaneous endoscopic gastrotomy [PEG] [13] are routinely used.

Research evidence for stem cell therapy in MND

Worldwide research is done using different types of stem cells and routes of administration. They have shown neuroprotective effect in MND/ALS. Studies have been conducted be various researchers which have shown safety and preliminary effectiveness. [14-23]

Recently, Moura et al conducted a systematic review and meta-analysis of studies that showed the safety and efficacy of stem cell therapy in preclinical and clinical studies. Our published study was included in this meta-analysis which had largest number of patients and survival benefits.

In India, Prabhakar et.al has published a pilot study of autologous bone marrow derived stem cell in patients with ALS in the journal of Neurology India. This study included 10 ALS patients and outcome was measured on ALSFRS-R score. After stem cell therapy

Authors	Year	N (years)	Disease (month)	Follow-up	Outcome	Survival	ALSFRS-R	Adverse events
Glass et al [33]	2012	12	4.4	1.5	ALSFRS-R: Adverse events	Not related	Stable	12
Karussis et al [31]	2010	19	2.9	25	Adverse events	Not related	Fall light	Fever: 11
Blanquer et al [37]	2012	11	1.8	6	Adverse events/ motoneurons	Not related	Stable	Pain and paresthesia: 11
Prabhakar et al [38]	2012	10	1.5	24	Survival	Not related	Decline after 3 months	No
Gamez et al [30]	2010	12	2.24	47.2	Survival/ trachea/ gastric tube	48 months	Declining equal to control	Fever, impaired conscious-ness:1
Martinez et al [27]	2009	10	2	19	Survival/ trachea/ gastric tube	66 months	Improvement in 1-2 months and at 6 months	No
Riley et al [35]	2012	12	3.1	18	Adverse events	Not related	Fall	Hemorrhage: 2
Mazzini et al [28]	2012	19	1.9	108	Adverse events/mune	Not related	Fall	Pain: 7 Loss sensitive: 6
Deda et al [29]	2009	13	2.6	12	Adverse events	Not related	Not ascertained	No
Tarella et al [32]	2010	24	1	12	Adverse events	Not related	Not ascertained	Prolactinoma: 1 TVPO: 1
Riley et al [35]	2012	12	3.1	18	Adverse events	Not related	Not ascertained	Hemorrhage: 2 reoperated
Mazzini et al [28]	2012	19	1.9	108	Adverse events/ mortality	52.5 months	Six patients for 74 months stabilised	No
Sharma et al [36]	2015	37	Not related	60	Survival	Improved 87.76(10.45)* 57.38(5.31)	Fall	Minor side effects

Table-Worldwide studies in ALS patients with Stem cell therapy (2009-2015)

(Courtesy: Moura, Mirian Conceicao et al. "Efficacy of Stem Cell Therapy in Amyotrophic Lateral Sclerosis: A Systematic Review and Meta-Analysis." Journal of Clinical Medicine Research 8.4 (2016): 317-324. PMC. Web. 15 June 2016.)

the results showed no significant deterioration in ALSFRS-R score from base line over the follow-up of 1 year. No major adverse events were noted. This pilot study concluded autologous bone marrow derived stem cell therapy is safe and feasible and suggested stabilization of disease in ALS patients .

Our results

Published results

Sharma et al published a controlled study of 57 patients [24]. Out of these, 37 patients underwent autologous bone marrow mononuclear cell transplantation in addition to standard rehabilitation , Riluzole and Lithium. Patients with respiratory distress were excluded. The survival duration since the onset of the disease of this intervention group was compared with a control group who did not receive cell transplantation. The survival duration of patients in intervention group was 87.76 months which was higher than the control group mean survival duration of 57.38 months (Figure 1) (Table 1). So overall stem cell therapy group survived 30 months longer. This study showed higher survival in younger age, limb onset and Lithium group. The result showed that the disease had slowed down in people undergoing stem cell therapy in comparison to the control group.

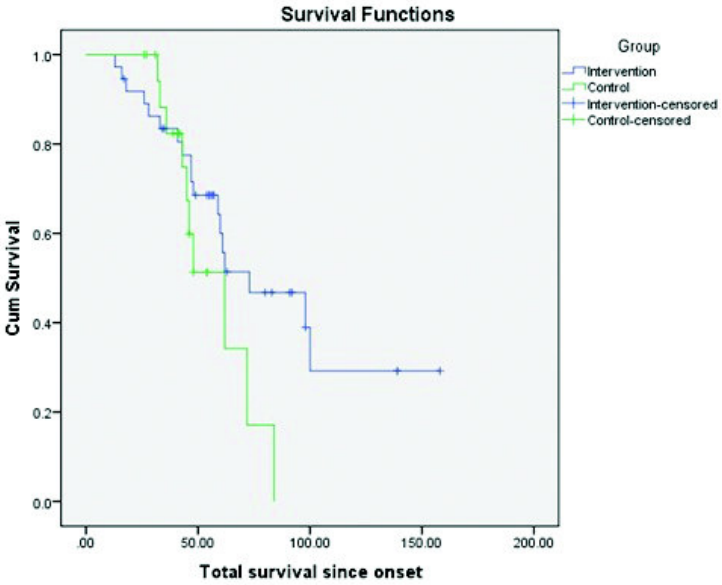


Figure 1 Kaplan Meier survival analysis comparing the mean survival duration of the intervention and control group

Table 2: Survival analysis

Survival analysis	Intervention group	Control group
Total mortality	48.64%	50.00%
Range of survival duration (months)	13 - 158	26-84
Mean survival duration (months)	87.76 (10.45)	57.38(5.31)

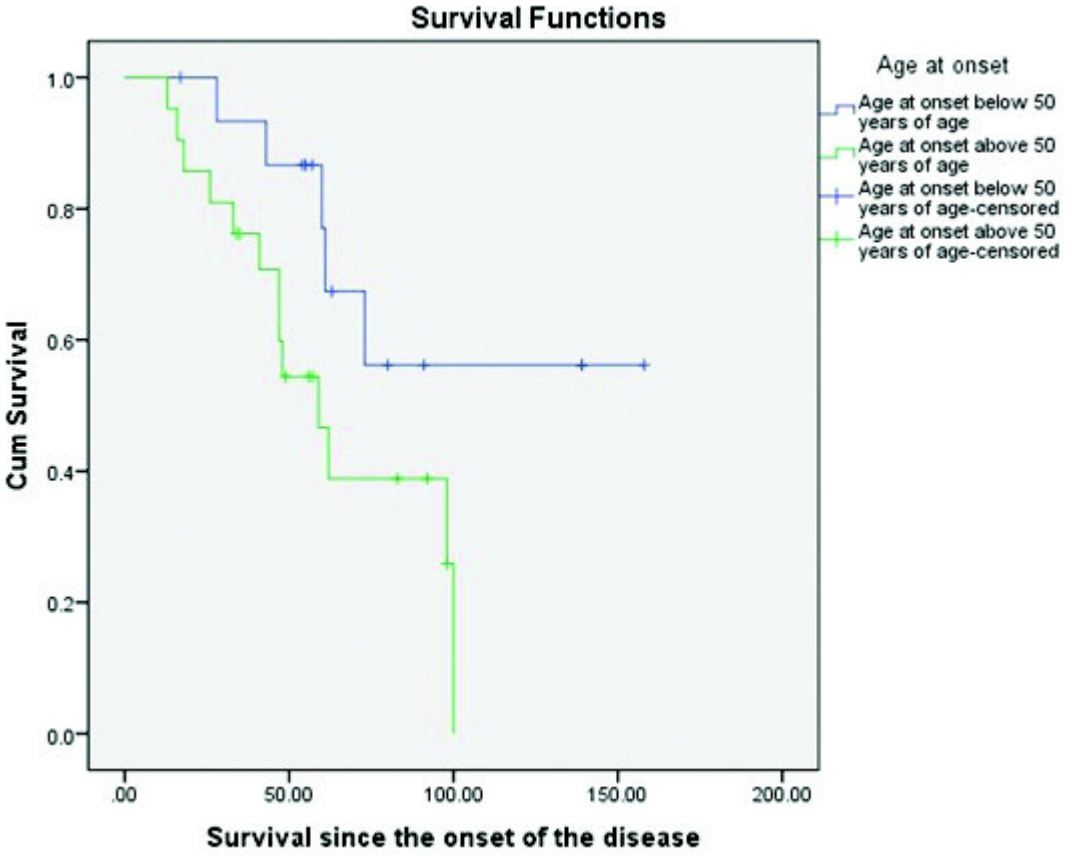


Figure 2: Kaplan-Meier Survival comparison for the patients in intervention group below and above 50 years of age at the onset

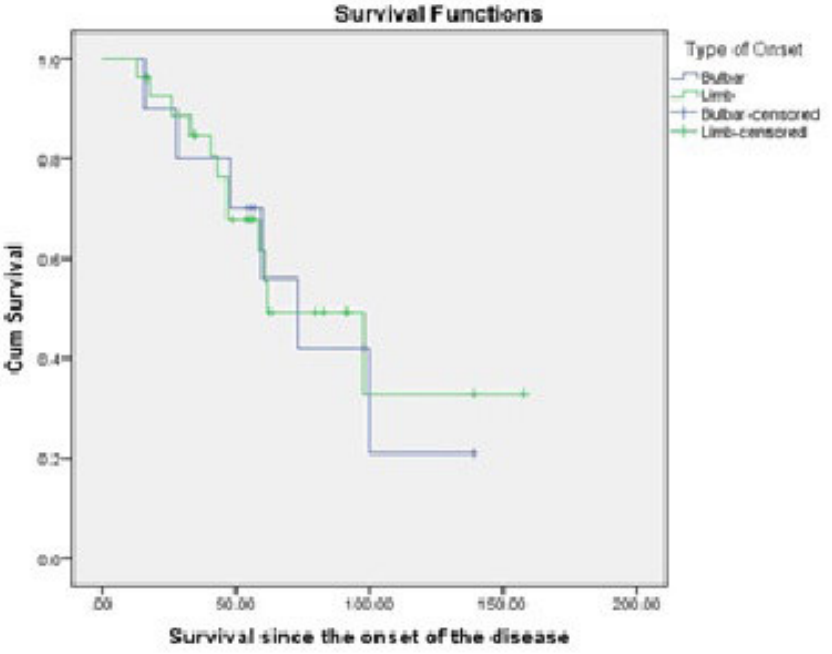


Figure 3: Kaplan-Meier Survival comparison for the patients in intervention group with bulbar onset and limb onset

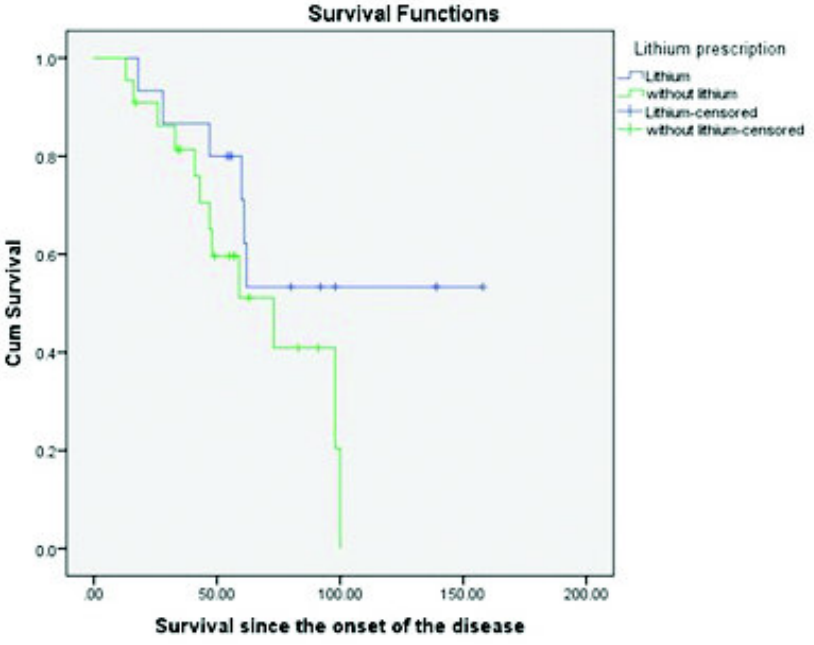


Figure 4.Kaplan-Meier Survival comparison for the patients in intervention group with or without Lithium prescription.

Female hormones show neuroprotective effect in ALS

The fact that ALS occurs more in males than females suggests that female hormones have some protective effect against this disease. Animal studies have shown that estrogen and progesterone have a positive protective effect on motor neurons. These hormones are required for normal functioning of motor neurons.

This neuroprotective effect of female hormones was also seen in a study conducted by Sharma et al. In that study, a total 40 patients (Males below 50 years =16, Males above 50 years= 12, Premenopausal females=7, Post menopausal females= 5) of ALS were treated with autologous bone marrow stem cell therapy. The results showed that the progression of the disease was significantly slower in premenopausal females than men below 50 years of age. Also premenopausal group showed the lowest mortality. Female reproductive hormones may have an additional neuroprotective benefit when combined with stem cell transplantation.

Table : Percentage mortality in all 4 interventional groups in ALS

Group	Mean Pre ALS-FRSr	Mean Post ALS-FRSr	Difference	Avg FU	Percentage mortality
Pre-menopausal women (7)	26	23	3	20	0
Post-menopausal women (5)	21	14	7	14	40
Men below the age of 50 (16)	25	20	5	10	25
Men above the age of 50 (12)	32	25	7	13	55

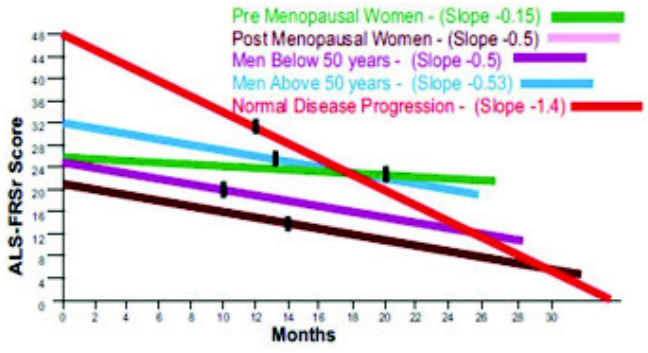


Figure - Graph showing disease progression on ALS- FRSr in all 4 groups and compared with natural rate of disease progression

Unpublished Data

The survival duration of the MND patients treated with intrathecal autologous bone marrow mononuclear cells transplantation since August 2008 till August 2015 was analyzed. There was a comparison made between the survival duration of the patients that underwent stem cell transplantation to those that did not. There were total 84 patients in the intervention group and 20 patients in the control group. Comparison of the survival duration suggested that the mean survival duration of the patients treated with stem cell therapy was longer than those who were not treated [Table 1] [Fig.1]. The mean survival duration of the patients who received treatment was 91 months and those who did not was 57. A clinically significant difference of 34 months in the survival duration suggests the potential benefit of stem cell therapy in the treatment of MND. It also shows that early stage patients and young premenopausal women benefit the most.

Some patients showed improvements in speech, stamina, muscle strength, muscle tightness, fasciculations and cramps. Patients with slow progression respond better than fast progression.

Future directions

Nur-Own cells transplantation

Recently brain storm cell technologies have developed Nur-Own cells. These are adult autologous mesenchymal cells harvested from bone marrow which are differentiated into specialized neuron supporting cells using the technology developed by Brain Storm Cell Therapeutics. Currently a Phase IIa trial is being conducted with 12 participants using intramuscular and intrathecal transplantation of the Nur-Own cells. Preliminary published results have shown slow down of disease progression. Confirmatory final results are awaited.

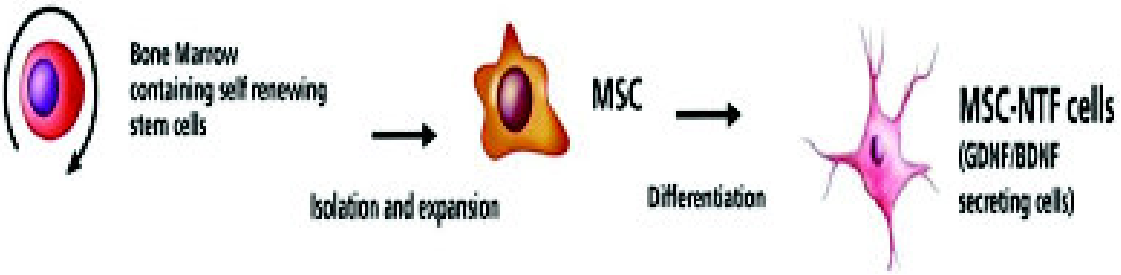


Figure- Nur-Own cells transplantation

Neural Stem

Neuralstem ALS trials used neural stem cells injected into the spinal cord by a novel device. This study showed preliminary safety and beneficial effects. Larger control trials are expected to begin soon.

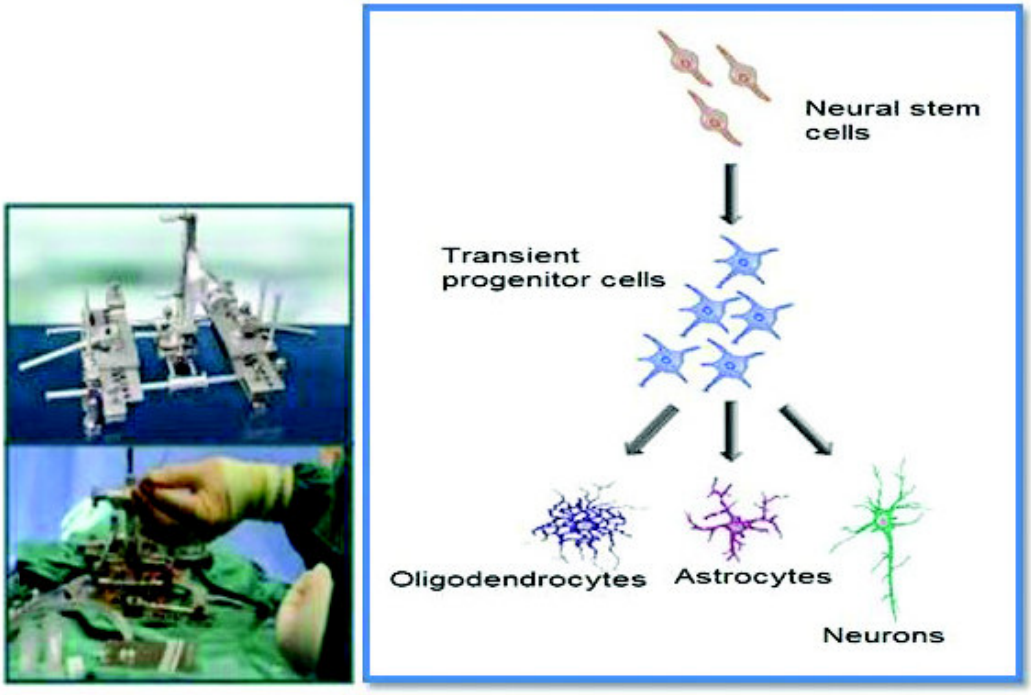


Figure- Neural stem cells

Gene therapy

Suspected genetic causality of MND and some evidence to support the genetic alterations in MND has led to emergence of gene therapy as a future management strategy for MND. A clinical trial using Antisense Oligonucleotides to reduce the toxic protein aggregates in MND is currently being undertaken [29].

Conclusion

At present, there is no proven treatment for ALS/MND. The only available treatment is Riluzole which has 3 to 6 months of survival benefit. Therefore, there is a significant unmet medical need. Stem cell therapy offers a definite hope in the treatment of ALS/MND. Our study shows that a combination of stem cell therapy and Lithium in addition to standard treatment with Riluzole and rehabilitation offers a survival benefit of 30 months. In the treated population, premenopausal women have shown 0% mortality and slowest disease progression. The published results of Prabhakar, Deda, Martinez

and Meamar as well as the recent review by Moura also shows the beneficial effects of cell therapy in ALS.

These results highlight the potential of stem cell therapy in the treatment of ALS/MND. However, there are many unanswered questions pertaining to stem cell therapy. Further research needs to be focused on exploring different routes of administration, types and dosage of cells for better clinical outcome.

It is likely that a definitive treatment and maybe even a cure will come from a combination of treatments which will include cell therapy, medications like Riluzole and Lithium and an aggressive rehabilitation program and may be adjuvant treatments like hormone replacement therapy.

Despite all of the above for an individual patient there are just two questions in their minds

- Will stem cell therapy help to improve my medical condition?
- And where should I get stem cell therapy done?

Based on our own clinical experience we believe that the following patients are likely to get greater clinical benefit with stem cell therapy

1. Younger patients below the age of 50 years
2. Women, especially premenopausal women
3. Patients in the early stage of the disease
4. Patients who have still not developed swallowing, breathing and speech problems

The second question is where the therapy should be done, to determine this we recommend that the patients should choose a centre after considering the following:

1. Use of safer types of cells like autologous cells or minimally manipulated cells
2. Published clinical results in international peer reviewed journals
3. Significant clinical experience in treating ALS patients and specialization in the field of neurological disorders
4. Holistic treatment approach that combines cell therapy with medication, rehabilitation and adjuvant therapies

Despite there being 12 published articles in peer reviewed scientific medical journals, by the standards of evidence based medicine, stem cell therapy is still considered by many as an unproven treatment for ALS/MND. Although stem cell therapy is still unproven, it is a safe treatment option available at present which could possibly alter the course of ALS/MND. The ethical basis for considering it as a treatment option is based on world medical association's Declaration of Helsinki regarding Ethical Principles for Medical Research Involving Human Subjects, paragraph 37 which clearly states that, 'In the treatment of an individual patient, where proven interventions do not exist

or other known interventions have been ineffective, the physician, after seeking expert advice, with informed consent from the patient or a legally authorised representative, may use an unproven intervention if in the physician's judgement it offers hope of saving life, re-establishing health or alleviating suffering. This intervention should subsequently be made the object of research, designed to evaluate its safety and efficacy. In all cases, new information must be recorded and, where appropriate, made publicly available.' Based on this we believe that in view of the near certain neurological deterioration and mortality that lies ahead of MND patients and in absence of any other proven treatment; stem cell therapy along with combination of pharmacological treatment, rehabilitation and adjuvant therapies should be made available to patients with ALS/MND.

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Chapter 25

New Drug Trials

1. Nuedexta

What is Nuedexta ?

Nuedexta is an oral medication, made up of dextromethorphan hydrobromide and quinidine sulfate. Dextromethorphan hydrobromide is the pharmacologically active ingredient that acts on the central nervous system, while quinidine sulfate increase effect of dextromethorphan. The FDA has approved Neudexta to be safe and effective drug for ALS in October 2010.

For what it is used?

Nuedexta is specifically indicated for the treatment of pseudobulbar affect. It means sudden and uncontrolled involuntary crying or laughing with or without any trigger or event. Up to 50% of ALS patients has pseudobulbar affect. It is thought to result from disruptions of neural networks that control the generation and regulation of emotions.

What is the recommended dosage?

The recommended starting dose for the treatment of PBA is dextromethorphan 20 mg/ quinidine 10 mg orally once daily for 7 days. The frequency is then increased to every 12 hours. It is important to reassess patients periodically for spontaneous improvement of PBA and the need to continue the treatment. It is very important that every patient have a thorough discussion with their primary physician or neurologist before taking the medicine.

What are the contraindications and adverse effects of Nuedexta?

The medicine is contraindicated in patients with a history of atrioventricular (AV) block, heart failure. Its adverse effects includes diarrhea, Dizziness, Cough, Vomiting, Asthenia/ weakness, swelling over feet, Urinary tract infection, fever.

Further Research

A clinical trial will begin in US to determine the effectiveness of Nuedexta in bulbar symptoms like impaired speech, swallowing, and excessive salivation.

2. Edaravone

What is Edaravone ?

The Edaravone is a drug that was approved in Japan for the treatment of stroke in 2002. It clear away harmful free radicals and protect neurons in the brain following brain damage. Abe and colleagues wondered if it might protect motor neurons in ALS, too.

How it works in ALS?

In animal studies beneficial effects of Edaravone have been reported as, it slowed disease progression and preserves motor neurons in mice. Oxidative stress has been considered to be involved in the onset and progression of ALS. Oxidative stress is an imbalance between production of free radicals and body's ability to detoxify their harmful effects through neutralization by antioxidants. Edaravone probably eliminates free radicals and might protect motor neurons from oxidative stress and damage in ALS patients.

Contraindications of Edaravone

This is contraindicated in patients with kidney disease, heart problems and liver failure and edaravone allergy.

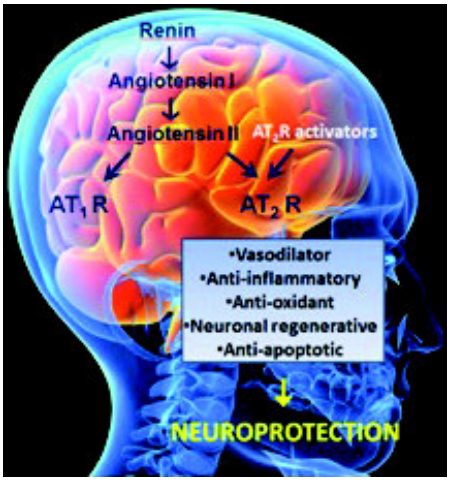
What research says?

Abe and colleagues conducted a 36-week confirmatory study to confirm the efficacy and safety of Edaravone in amyotrophic lateral sclerosis (ALS) patients and they identified that in a subgroup of mildly symptomatic participants progression of disease was seemed to be slowed down on Edaravone but efficacy of edaravone for treatment of all ALS patients was not demonstrated. This trial completed in 2014, led to regulatory approval in June of 2015.

Further studies have designed to conduct a clinical trial to evaluate the efficacy and safety of Edaravone for ALS patients in Japan.

3. Cerebrolysin

Cerebrolysin is derived from porcine brain proteins. Its main mechanism of action is to make the neurons in the brain function more efficiently and more in sync with each other. Cerebrolysin also have neuroprotective properties that reduce the degeneration of neurons and also stimulates the growth of new neurons.



It has been used in treating stroke and traumatic brain injury. Studies are needed to determine the effectiveness of cerebrolysin in ALS patients.

4. Genervon GM604

Cytoplasmic neuronal aggregations formed by TDP-43 DNA binding Protein is one of the major pathological features observed in post-mortem tissue from patients with ALS. This protein spreads out from the nucleus to the cytoplasm, forms aggregation and becomes toxic, causing the death of motor neurons. Dr. Inoue of Kyoto University has demonstrated that as TDP-43 mRNA levels decreased, the death of neurons was prevented. Therefore, a slowing of ALS disease progression is expected. According to the results of Genervon's Phase 2A clinical trial of GM604 in ALS patients indicated that GM604 has the ability to reduce the formation of random protein aggregates, to modulate protein biomarkers and can bring the TDP-43 protein back to their normal ranges, thus restoring homeostasis. GM604 significantly reduced the slope of plasma TDP-43 concentration at 12 weeks when compared with placebo ($p=0.0078$) and demonstrated clinical improvements in the heterogeneous ALS patient population.

5. MN-166 (ibudilast)

The FDA (U.S. Food and Drug Administration) has approved the protocol of a new clinical trial in ALS patients, this study will measure the effects of MN-166 on reducing brain microglial activation utilizing a biomarker in 15 subjects with ALS. This trial is expected to commence shortly. ALS subjects will be recruited and will be treated with ibudilast 50 mg twice daily for 36 weeks. The objectives of trial are to measure the effects of ibudilast on reducing brain microglial activation measured by [[11] C]-PBR28-PET (biomarker) and evaluation of the safety, tolerability, and clinical outcomes (ALS functional rating scale [ALSFRS-R], slow vital capacity [SVC], and muscle strength [measured by HHD - hand-held dynamometry) over 36 weeks in ALS patients. MN-166 (ibudilast) attenuates activated glia cells and its anti-neuroinflammatory and

neuroprotective actions have been demonstrated in preclinical and clinical study.

6. Retigabine

Harvard stem cell scientists have discovered that a recently approved medication Retigabine for epilepsy might be a meaningful treatment for (ALS). The researchers are now collaborating with Massachusetts General Hospital (MGH) to design an initial clinical trial testing the safety of the treatment in ALS patients. Eggan and Clifford Woolf have found that the many independent mutations that cause ALS may be linked by their ability to trigger abnormally high activity in motor neurons. Using neurons derived from stem cells made from ALS patient skin cells, conducted clinical trials of the anti-epilepsy medication on neurons in laboratory, finding that it reduced the cells' hyperexcitability. Eggan's described that possessed ALS mutations had a sporadic increase in neuron firing, while the healthy neurons were quiet unless stimulated in some way. This overexcitable ALS neurons generate more abnormally folded proteins, further increasing their excitability. The strain of this cycle seems to put the neurons in a vulnerable state, where they are more likely to die. MGH neurologist Merit Cudkowicz, with Wainger, will be running the clinical trials, which will first test for side effects in ALS patients treated with the drug.

7. Tocilizumab

ALS Association and Massachusetts General Hospital are currently recruiting the participant for study. This research study is being done to find out if tocilizumab, also known as Actemra™, can help patient with ALS. Although not the initial cause of ALS, the immune system plays a role in the death of motor neurons. The immune cells that participate in this process are stimulated by a substance called interleukin-6 (IL-6) whose effect is blocked by tocilizumab and thus, may slow the death of motor neurons and slow the disease. The investigators also want to find out if tocilizumab is safe to take without causing too many side effects.

8. Mexiletine

University of California, Davis and ALS Association recruiting the participant to determine if mexiletine is effective for the treatment of muscle cramps in ALS patients. Mexiletine is a medication taken orally stops the type of sodium currents that are thought to cause muscle cramps. Mexiletine is a relatively older medication that has been extensively studied in humans. It has been shown to reduce the electrical measures of muscle cramps for other disease conditions.

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Chapter 26

Respiratory Therapy

In Amyotrophic Lateral Sclerosis (ALS) breathing difficulties can start early or much later. Because of this patients prognosis will be poor. So the non-invasive and invasive mechanical ventilation have been used for patient's care. Following are the alternative approaches to treat the respiratory problems.

1. Cough Assist

Coughing is very important to clear material from the airway. Causes of poor coughing mechanism

- Inability to take a deep breath due to weak respiratory and swallowing muscles.
- Reduced physical mobility or decreased fluid intake may also results in shallow breathing and thickened mucus

Cough Assist Device

Automatic Mechanical cough assist devices such as the Philips CoughAssist™ or Hill-Rom Vital Cough™ helps those with an ineffective cough by breathing through a mouthpiece or mask, the device gradually applies positive pressure to insure a deep breath, then shifts to negative pressure to assist with pulling secretions upward, simulating a deep natural cough.

Figure 26.1: Cough Assist Device

2. Chest vibration jacket /High-Frequency Chest Wall Oscillation (the Vest)

High-frequency chest wall oscillation involves an inflatable vest that is attached to a machine. The machine mechanically vibrates at a high frequency. This helps to loosen and thin mucu

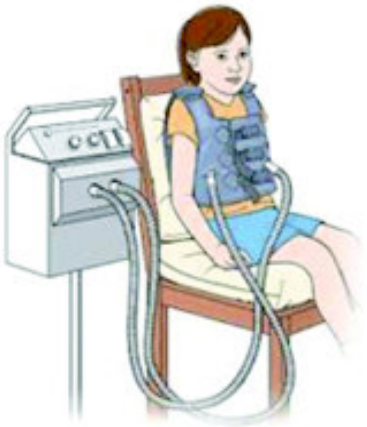

Figure 26.2: Chest Vibration Jacket

3. Clinical trials of diaphragmatic pacing

What is diaphragmatic pacing?

It's like a cardiac pacemaker, which provides rhythmic application of electrical stimulus to the diaphragm (most important muscle of respiration) for ventilatory support. Electrodes are placed in the diaphragm muscle via a laparoscopic surgical procedure.

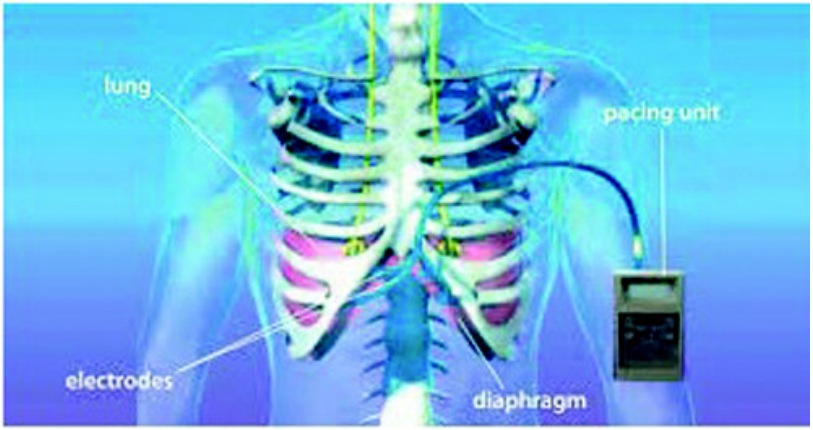

Figure 26.3: Diaphragmatic Pacing System (DPS)

Recent studies about diaphragmatic pacing in ALS patients

1. In 2011 after FDA approval a study was designed to determine the probable benefit and risk from use of DPS for ALS patients. 60 patients were involved into study and out of that 90% patients were had stimulable diaphragm and had successful laparoscopic placement of the electrodes. Adverse events like external electrode wire breaks and infections at the percutaneous wire exit sites were noted. These events were resolved and patients continued therapy. The study has been amended to enroll additional patients.
2. Mcdermott and colleagues conducted a study by using NEURX RA/4 diaphragm pacing system at seven ALS centers in the UK with 74 patients, who were treated either with noninvasive ventilation alone or to this standard of care plus diaphragm pacing. The primary outcome was overall survival. The study was stopped after 2 years because researchers found that survival was shorter when diaphragmatic pacing was added to the mix.

   So the patient selection for diaphragmatic pacing should be done very carefully depending upon patient's respiratory capacity, any diaphragm abnormality or any heart or lung disease. It is very important to consult your treating neurologist or family doctor before taking any decision regarding diaphragmatic pacing.

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Chapter 27

Improving communication in ALS

In ALS speech and voice changes are also present along with other physical symptoms. A person's speech may be slurred and voice may be weak. Difficulty increases especially after talking for an extended period of time and/or when tired later in the day. Following devices might help the person with ALS to communicate to the world in better way.

1. Eye Speak glasses

The eye tracking or eye speak glass will provide the possibility of communicating through the eyes of the users, irrespective of the orientation and position of the user's head.

Figure 27.1- Eye speak glasses

How it works?

The system consists of a pair of glasses that will have a keyboard onto the patient's field of view. There will be a micro camera, which will track his eye movement to understand which key is being selected. As the patient finishes writing his set of words, those words can be heard through the speaker. There is also some pre installed list of sentences like 'I am hungry' or 'I want to get up'. Patient selects those sentences through eye movements and can convey his massage. So patient will be able to communicate with his relative, friends and family members in better way.

2. Voice banking

Voice banking is the process of storing our own voice. So when a communication device is used, the voice you hear is the voice of the person who previously recorded their voice.

Method A: in this method massages are recorded. These messages can be programmed into a speech generating devices (SGD) and used when needed for communication. With this method, only recorded messages will use the person's natural voice. Newly created messages using spelling, will use the SGD's artificial voice.

Method B: This method of voice banking allows for both recorded messages and newly created messages, using spelling, to be spoken using the person's natural voice. It requires the potential user to record a large sample of words and phrases using a specific software program. Model Talker is a speech synthesis system designed specifically for users of SGD's.

3. Robot

Japan ALS association has designed a Robot to aid the communication of PALS with their family members.

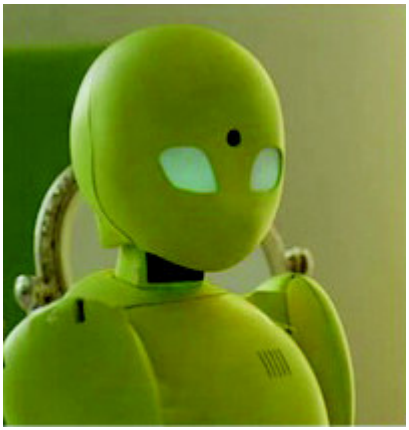

Figure 27.2:- Robot for communication

How it works?

If the patient is hospitalized or away from his home and family members the Robot can relay voice messages with arm gestures to the patients and Family members of patients ALS. There are web cameras in the eyes that can help patients to see their house and family members on a screen.

All these communication devices need to be examined for their possible advantages and disadvantages for easier and faster communication of ALS patients.

References

- www.modeltalker.com
- www.asel.udel.edu/speech/ModelTalker.html

Chapter 28

Biomarkers
PET scan for ALS

Amyotrophic lateral sclerosis results in damage to the brain tissues. The MRI scan gives us idea about only the damaged areas due to disease. However the PET scan tells about how your brain and its tissues work.

What is PET and how it works?

A brain positron emission tomography (PET) scan is an imaging test of the brain. A dye containing radiotracer is injected into blood stream. It will enter the brain. Dye will be taken up and metabolized by brain tissues. The radiotracer will break and emit rays which are detected by scanner to form the brain image.

Dr. Nazem Atassi, used PET imaging to successfully scan the first person living with ALS to measure inflammation in the brain and to develop biomarkers that is a measurable indicator of condition, that would help with diagnosis of ALS and measuring the progression of the disease.

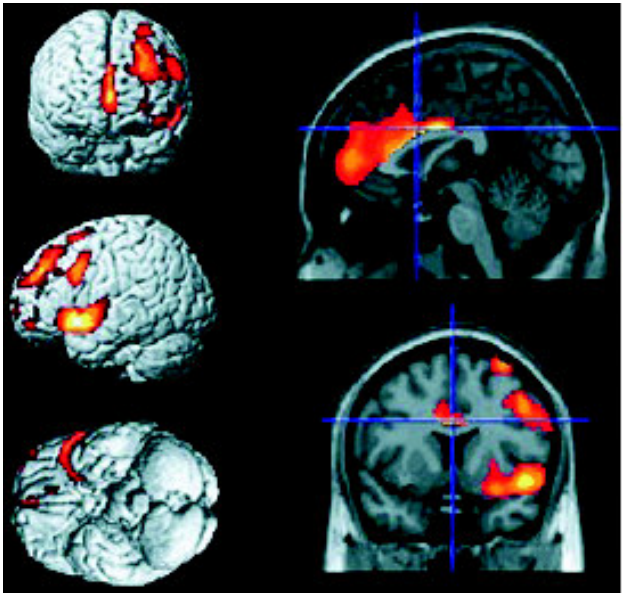

Figure 28.1- PET scan

An investigational PET radiotracer was designed to bind to the support cells of the nervous system) and serve as a marker of inflammation and track changes in the brain glial cells. A study done by using similar tracer show increased uptake in the motor

cortex and brain stem in people living with ALS that is strongly correlated with clinical measures. Inflammation is an important target for ALS drug development, and imaging inflammation may allow to design and conduct more efficient ALS clinical trials and will increase the speed of ALS drug discovery.

Future Research Projects

Zucher N, Lawson R et all are conducting a large biomarkers study that includes imaging, whole genome sequencing, biofluid inflammatory biomarkers and induced pluripotent stem cell generation. Whole genome sequencing will tell about the complete DNA make up of individual so the gene variants can be found much more easily. Biofluid inflammatory biomarkes will give the measureable indicator about the presence of inflammation in brain tissues. Induced pluripotent stem cells are adult cells that have been genetically reprogrammed to an embryonic stem cell. These cells will give rise to other cell type in the body like neurons in the brain which will replace the damaged cells in ALS.

The overall study goal is to identify these new imaging markers of ALS to form and establish the basis for new understandings of the disease and new therapeutic strategies. The TRACK ALS project is funded through the ALS ACT initiativeReference:

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Non invasive EMG for ALS

What is EMG?

Electromyography (EMG) is a diagnostic procedure used to record the electrical activity of muscles. When muscles are active, they produce an electrical current. This current is usually proportional to the level of the muscle activity. An EMG translates this into graphs, sounds or numerical values that a specialist interprets. An EMG uses tiny devices called electrodes to transmit or detect electrical signals.

Why is an EMG test done?

An EMG is often performed when patients have unexplained muscle weakness. The EMG can be used to detect abnormal electrical activity of muscle that can occur in many diseases and conditions, including muscular dystrophy, inflammation of muscles, peripheral nerve damage (damage to nerves in the arms and legs), amyotrophic lateral sclerosis (ALS), myasthenia gravis.

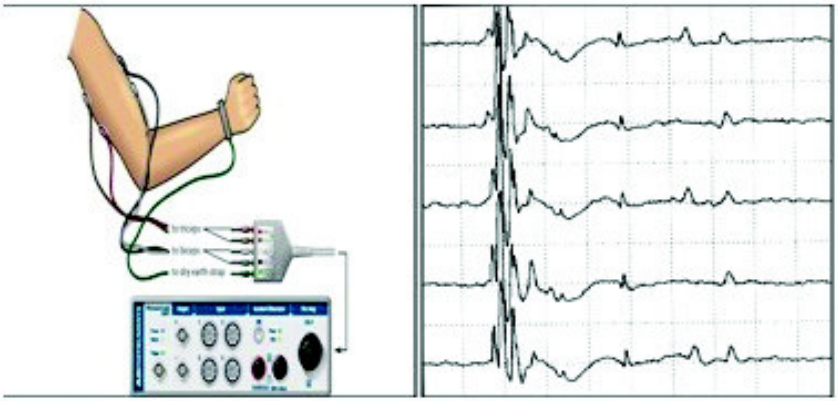


Figure28.2- EMG procedure and the waveforms obtained.

What is invasive and non invasive EMG?

During an invasive EMG, a needle electrode inserted directly into a muscle that records the electrical activity in that muscle.

In non invasive surface EMG electrodes are placed on (not into) the skin overlying a muscle to detect the electrical activity of the muscle. This method can be used for supporting diagnosis of amyotrophic lateral sclerosis (ALS).

Advantages of non invasive EMG

- Not painful as surface electrodes are used to detect electrical activity of muscles
- Can be performed repeatedly to measure disease progression
- Chances of infection are less

Research done

In a study by Zhang et al different surface EMG patterns were used to record the muscle activity during different levels of voluntary muscle contraction. The method was tested in 10 ALS subjects and 11 neurologically intact subjects. Combination of the three surface EMG markers achieved 90% diagnostic sensitivity and 100% diagnostic specificity.

Furthermore confirmatory studies will be required so that non invasive EMG can be used more practically and repeatedly to monitor disease progression in ALS.

References

Xu Zhang Paul E. Barkhaus ; William Zev Rymer ; Ping Zhou Machine Learning for Supporting Diagnosis of Amyotrophic Lateral Sclerosis Using Surface Electromyogram

Chapter 29

Genetic origin of ALS

What is Sporadic and Familial ALS?

Most cases of ALS are "sporadic," meaning they are due to no known cause. Approximately 5-10% of cases are due to known genetic mutations. In most of those cases, another family member also has ALS or a related disease. Such cases are called "familial." Cases of familial ALS (FALS) are presumed to be due to genes, although the gene may not be known in all cases.

Genes that Cause ALS

To date mutations in thirteen genes are known to cause dominantly-inherited adult-onset ALS (SOD1, C9ORF72, TARDBP, FUS, OPTN, VCP, UBQLN2, SQSTM1, PFN1, MATRIN3, CHCD5, TUBA4A and TBK1).

C9ORF72

This gene, discovered in 2011, is the most common genetic cause of ALS. Mutations in this gene account for between 25% and 40% of all familial ALS cases (depending on the population), and also approximately 4% to 6% of sporadic cases. This gene also causes another neurodegenerative disease, called frontotemporal dementia (FTD) this means significant impairment of thinking or behavior that results from progressive deterioration of the frontal and temporal lobes of the brain.

Some people with this gene will develop symptoms only of ALS, some only of FTD, and some will have symptoms of both disorders. How this gene causes ALS is unknown, and is the subject of a great deal of intense research.

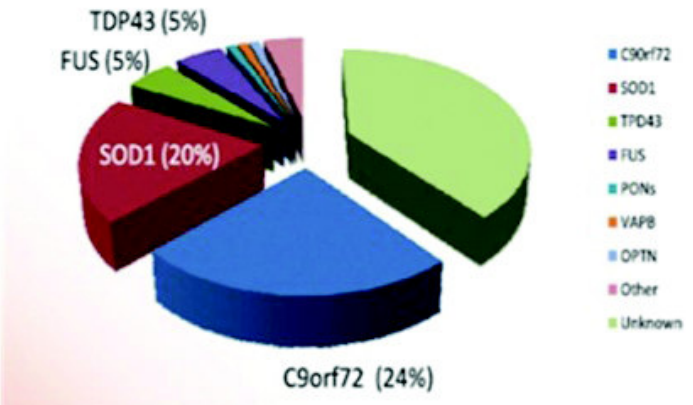


Figure - 29.1 The percentage distribution of ALS patients with different genetic mutations

Cu/Zn Superoxide Dismutase 1 (SOD1)

Mutations in SOD1 were first described in 1993, and SOD1 was the first gene known for ALS. It accounts for about 10% of familial ALS, or 1.5% to 2% of all ALS. How SOD1 mutations cause ALS is unknown. This gene appears to take on some new toxic function, possibly related to an increase in the tendency of mutant SOD1 molecules to aggregate and form clumps in motor neurons. It is also possible that SOD1 causes ALS through actions in nearby cells called astrocytes, not in motor neurons themselves. Astrocytes help maintain motor neurons, and SOD1 mutation may impair their ability to do so.

TDP-43 and FUS

TAR DNA binding protein 43 (TDP-43) and fused in sarcoma (FUS) are RNA-binding protein was linked to ALS in 2008. Mutations in the TDP-43 and FUS gene cause protein to mislocalize away from the nucleus where it is normally found, and into the cytoplasm (the material surrounding the nucleus). Protein will aggregate into cytoplasm to form clumps that can be seen under the microscope. This eventually will cause neuronal cell death and brain damage.

Ubiquilin-2

Ubiquilin-2 was linked to ALS in 2011. The normal function of the protein is to help destroy damaged or defective proteins in the cell. It is likely that mutations in the gene interfere with this function, and may lead to accumulation of harmful material within the cell which leads to cell death in ALS.

Gene Therapy Trials

What is gene therapy?

Gene therapy can be simply a means genetic repair of the damaged brain cell to deliver a beneficial cell protein, which will stop the disease progression and help to repair the dying nerve cells.

What are challenges for gene therapy in ALS?

In ALS, a few percent of patients have a known defect in a gene. But in other cases of ALS it is very important to find out the abnormalities in other genes also in order to design gene therapy for that particular defect.

What is encouraging is that the knowledge of the SOD1 mutations has produced a vast body of evidence for what does go wrong in other cases of ALS. And that evidence is pointing to a general strategy that might be successfully implemented through gene therapy.

Current development in gene therapy for ALS

1. In a study adult ALS mice were treated by silencing the SOD1 gene to find out its therapeutic effects. The study demonstrated profound delay in both disease onset and death in mice. Also significant preservation muscle strength, motor and respiratory functions in mice were noted.

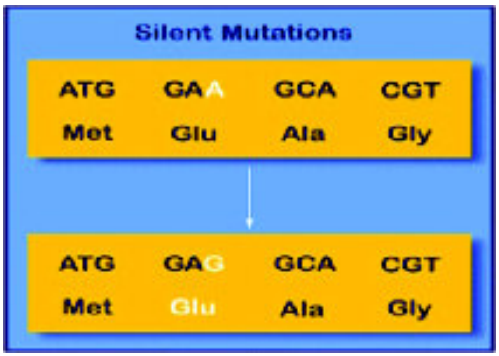


Figure- 29.2 Silent mutation

2. According to a paper in the Proceedings of the National Academy of Sciences USA online a dose of up-frame shift protein 1 (UPF1) prevented degeneration in models of sporadic ALS and in models of ALS due to TDP-43 or FUS gene mutations.

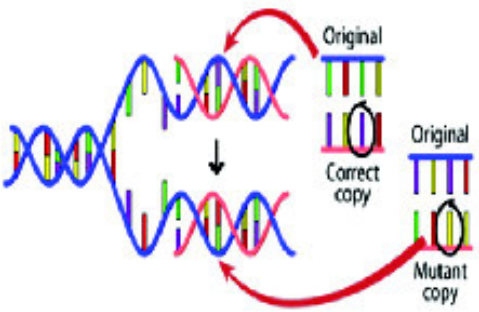


Figure 29.3 - Frameshift mutation

3. Kaspar and colleagues have successfully slowed disease in ALS mice by forcing motor neurons to produce insulin-like growth factor 1 (IGF-1). IGF-1 may protect deteriorating motor nerves and promote growth and regeneration of motor nerve. This may provide real hope for an effective treatment of ALS after conducting human trials.

References

1. Florie Borel et al. Therapeutic rAAVrh10 Mediated SOD1 Silencing in Adult SOD1G93A Mice and Nonhuman Primates.
2. Séverine Boillée, Don W. Clevel Gene therapy for ALS delivers

Ongoing Clinical trials

Sr. No.	Title of the research	Status of the trial	Organization conducting the research
1.	Ibudilast (MN-166) in Subjects With Amyotrophic Lateral Sclerosis (ALS)	Recruiting	Carolinas Healthcare System, Dept. of Neurology Charlotte, North Carolina, United States,
2.	Olanzapine for the Treatment of Appetite Loss in Amyotrophic Lateral Sclerosis (ALS)	Recruiting	Charite University, Berlin, Germany
3.	Mexiletine for the Treatment of Muscle Cramps in ALS	Recruiting	University of California, Davis ALS Association
4.	Intravenous Injection of Adipose Derived Mesenchymal Stem Cell for ALS	Recruiting	Royan Institute, Iran, Islamic Republic
5.	Evaluation of Metabolomic Analysis in Early Diagnosis of ALS (METABALS)	Recruiting	University Hospital, Tours
6.	A Pilot Study of Inosine in ALS	Recruiting	Massachusetts General Hospital, United States, Massachusetts
7.	A Clinical Trial of Safety and Efficacy of Fasudil in Subjects With Amyotrophic Lateral Sclerosis [ALS]	Recruiting	Peking University Third Hospital, Beijing, China
8.	F 18 T807 Tau PET Imaging in Familial Amyotrophic Lateral Sclerosis (T807 ALS)	Recruiting	Washington University School of Medicine United States, Missouri
9.	The Objective is to Compare the Efficacy and Safety of Masitinib in Combination With Riluzole in the Treatment of Patients Suffering From Amyotrophic Lateral Sclerosis [ALS]	Recruiting	Madrid, Spain
10.	A Phase 3, Multi-National, Double-Blind, Randomized, Placebo-Controlled, Stratified, Parallel Group, Study to Evaluate the Safety, Tolerability and Efficacy of	Recruiting	Carol and Frank Morsini Centre for Advanced Health Care - University of South Florida

	Tirasemtiv in Patients With Amyotrophic Lateral Sclerosis (ALS)		
11.	A Pilot Study of RNS60 in Amyotrophic Lateral Sclerosis (ALS)	Recruiting	Massachusetts General Hospital, Neurological Clinical Research Institute, Boston, UNITED STATES
12.	Developing a Discrimination Model to Diagnose ALS Using Advanced MRI Techniques	Recruiting	University of Michigan Hospital, Ann Arbor, Michigan, UNITED STATES
13.	Paired Stimulation to Increase Cortical Transmission to Hand Muscles: Pilot Study	Recruiting	J. Peters VA Medical Center, Bronx, New York, UNITED STATES
14.	Initiation of Long-term Home Non-invasive Ventilation in ALS Using the iVAPS Mode During a Daytime Trial	Recruiting	Montreal Chest Institute, Montreal, Quebe, CANADA
15.	A Phase 2 Pharmacodynamic Study of Ezogabine on Neuronal Excitability in Amyotrophic Lateral Sclerosis	Recruiting	Massachusetts General Hospital - Neurological Clinical Research Institute (MGH-NCRI)
16.	Human Spinal Cord Derived Neural Stem Cell Transplantation for the Treatment of Amyotrophic Lateral Sclerosis (ALS)	Ongoing, not currently recruiting	Neuralstem Inc.
17.	Phase 2, Randomized, Double Blind, Placebo Controlled Multicenter Study of Autologous MSC-NTF Cells in Patients with ALS (NurOwn)	Completed	Brainstorm-Cell Therapeutics
18.	Effect of stem cell therapy on disease progression in ALS	Recruiting	NeuroGen Brain and Spine Institute

SECTION E

Going beyond the limitations of ALS/MND

Walter Lane Smith III

(April 29, 1936 - June 13, 2005)

Walter Lane Smith III, known as Lane Smith was an American actor. Smith was diagnosed with amyotrophic lateral sclerosis (also known as ALS, or Lou Gehrig's disease) in April 2005. He died of the disease at his home in Northridge, California June 13, 2005 at the age of 69.

Some of his well nown roles included portraying collaborator entrepreneur Nathan Bates in the NBC television series V, and American President Richard Nixon in The Final Days, for which he received a Golden Globe award nomination.

Chapter 31

Motivation

The most important and difficult aspect in living with MND is to keep the motivation levels intact. It is understood that even if the patient wants to get better, the body does not support in adequate movements and normal functioning in day to day life. Undoubtedly, it is difficult to keep a positive attitude when one is facing with a life threatening disease, but doing so may mean the difference between living and dying.

There are two types of motivation: intrinsic and extrinsic.

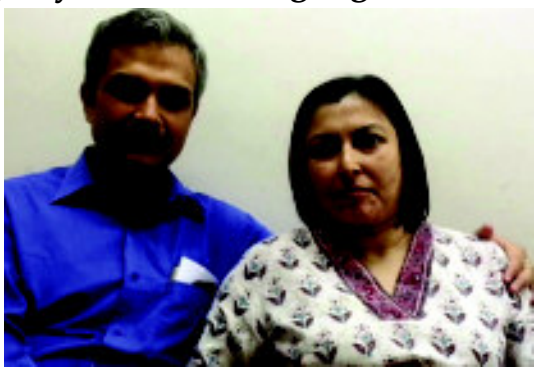
Intrinsic motivation is something that comes from within. It is the desire to seek new things, improve one's skills and abilities and is driven by internal rewards which are in accordance with his belief systems. Therefore, it is very important for a patient with MND to feel from within to get better. The intense desire to recover and improve physically will determine the course of his progression. The will to not give up and a strong determination to fight with the challenges of the illness every day will help the patient lead a more meaningful life.

"Motivation is a fire from within. If someone else tries to light that fire for you, chances are it will burn very briefly".

Extrinsic motivation is something that comes from influences that are outside the individual. For ex: money, reward, bonuses etc.

"Motivation gets you going and habit gets you there." - Zig Ziglar

1. Positive Attitude - It is difficult but definitely possible. Positive attitude attracts positive energy which helps to face MND with a strong mind demeaning its ill-effects. Surround yourself with positive people, enthusiastic songs, inspiring movies, books etc. Make your environment pleasant, energetic and peaceful. Focus



on the half full glass and not the half empty portion. Pay attention on how we can keep on doing the activities that are possible for the longest period. Forget disappointments and think about your dreams. Positive thinking brings about favorable changes as the universal forces always responds in the direction of the mind's signals. The real power lies not in the muscles but in the mind.

"Among the things you can give and still keep are your word, a smile and a grateful heart". -Zig Ziglar

2. Affirmations: Use positive affirmations every day. Affirmations refer to the practice of positive thinking and self-empowerment fostering a positive mental attitude. Affirmations strengthen you by helping you believe in the potential of an action you desire to manifest. They basically help in self-improvement because of their ability to rewire the brain. Some of the affirmations for good health can be:

   - " I love myself and I am perfectly healthy"
   - "Every day and in every way, I am getting better and healthier"
   - "Every day is a new day full f hope, happiness and health"
   - "Today I am brimming with energy and overflowing with joy"

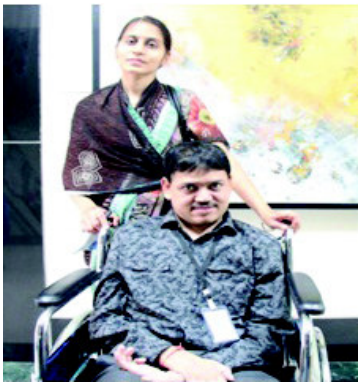
Go as far as you can see and you will see further." - Zig Ziglar

3. Importance to life - Remember 'Life is beautiful, no matter what'. Look at innocent smiling faces of children, delicate peaceful newborn, beautiful mother nature - rivers, mountains, green trees, chirping birds, refreshing flowers, vibrant rainbow and open sky. May be in busy life these things were ignored so take this chance to enjoy and appreciate the beauty of life. Give importance to living and not the disease.

4. Acceptance - It is difficult in the beginning but sooner we accept that we have to cope, we will find ways to work around the situation earlier. Explain the mind

"It's not where you start or even what happens to you along the way that's important. What is important is that you persevere and never give up on yourself." - Zig Ziglar

5. Get back to work & to-do list of life - Try to get back into your profession or any work you enjoy. Get involved in a cause close to your heart or social work. Take a helper along and actively participates in tasks you like e.g. cooking, household work, work on computer or telephone, become an advisor or counselor, teaching etc. There are many ways you can contribute and always remember you are needed. Make a to-do list of things which you always dreamt of doing and start working on it to fulfill it.



"Duty makes us do things well, but love makes us do them beautifully." - Zig Ziglar

6. Socialize - Have no fear and face the world [people] with confidence and dignity. Join online communities, school or college groups. Get out of home even if it means taking your helper along. Meet friends, relatives, neighbors etc. Go for movies, eat at restaurants, go to mall, and visit places you enjoy. Don't bother about people staring at you; just think they are appreciating your high spirits.

"Make today worth remembering." - Zig Ziglar

7. Share feelings - Find some people in your life [family or friends or acquaintance on internet] with whom you can share your feelings and express your emotions. Sharing can be verbal face to face or on phone and can also be written on paper or email. Basically, you should take out your emotions as it acts like a catharsis which calms down your body and mind. Appreciate and acknowledge supportive people around.

You never know when a moment and a few sincere words can have an impact on a life." - Zig Ziglar

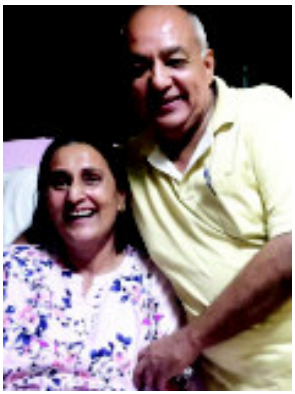
8. Self-entertainment - Enjoy life and remember it's your responsibility to entertain yourself. Play games with family, friends and if nobody is available play on the computer. Hobbies have to be retained. If the hobby is something that you are unable to do then watch someone else doing it. As it will give you immense pleasure to be connected with your hobby.

"It does not matter how slowly you go, as long as you don't stop"

9. Peace of mind - Forgive all and don't blame others or yourself for the problem. Everyone has problems in life and this disease can happen to anybody at anytime. You have done nothing wrong. The renowned scientist Stephen Hawking is also living with MND. There are many doctors, nurses, engineers, and farmers etc who develop this disease. Try to achieve peaceful mind which will increase your inner strength. Meditation, prayers, religious books, enchanting 'OM' and inspirational stories are some of the ways which offer peace of mind.

Do it now. Sometimes Later becomes Never.

10. Keep smiling - 'Laughter is the best medicine'. Smile and the whole world smiles back at you. Watch comedy movies, shows, funny videos, cartoons or read funny books. Laughter stimulates positive chemicals in our body which relieves stress. Always keep smiling.

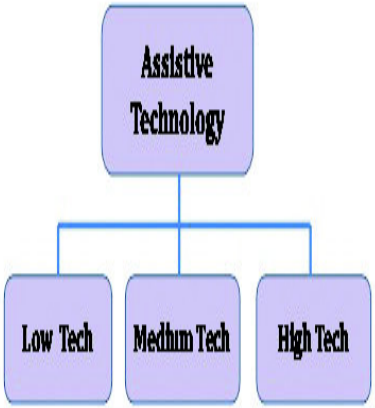


"Laughter is an Instant Vacation" - Milton Berle

Chapter 32

Mind Power

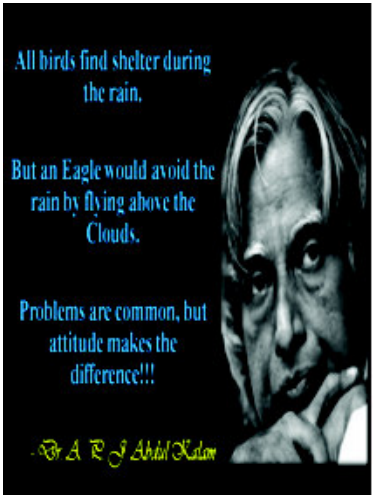
"Mind Nurtures Dynamic U So U Nurture Mind with Positive View"



To understand the power of the Mind, it is essential to first know how the mind actually works. The mind works at two levels - conscious and subconscious. Your conscious mind works according to your thinking; whereas the sub-conscious mind is the seat of your emotions, which is the creative mind. Whatever is thought habitually, it tends to sink into the subconscious mind. The law of the subconscious mind works for good and bad ideas alike. Your sub conscious mind is quite sensitive to your thoughts. When this law is applied in a negative manner, it brings about failure, frustration and unhappiness. However, when your thinking is more positive, harmonious and constructive, you experience success, prosperity and perfect health. Whatever you claim mentally, the sub conscious mind will accept and bring forth into experience. The only thing necessary is to get the sub conscious mind to accept the idea or whatever thoughts are conveyed to the sub conscious mind.

How the sub conscious controls all functions of the body

When you are awake or asleep, the tireless action of the sub conscious mind controls all the vital functions of the body. For ex: when you are asleep, the heart continues to beat in rhythm, the process of inhalation and exhalation continues just as the same way as you are awake. However, the conscious mind interferes with the functions of the body such as heart rate, lungs, stomach and intestine by way of worry, anxiety, fear and depression. These repetitive patterns of thought interfere with the harmonious functioning of the sub conscious mind. Therefore, speaking to the sub conscious mind, giving it suggestions and commands will allow the functions of the body to get back to normal again. Whatever thoughts are conveyed to the sub conscious mind, impressions are formed in the brain cells. As soon as the sub-conscious mind accepts any idea, it starts to put it into effect immediately.



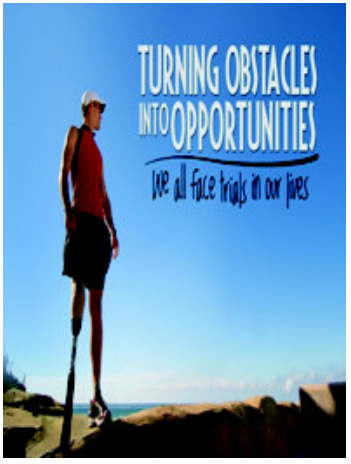
How to get the sub-conscious mind working:

1. ***Through Creative Visualization technique***

   "A picture is worth a thousand words". The easiest way to formulate any idea is to visualize it mentally, in the mind's eye as vividly as possible. What you form in your imagination is as real as any part of your body. This process of visualization involves creating images that convey a strong positive message. The image of what you want to see in yourself. For ex: Imagining walking on your own or writing on your own without any support. Imagining you are in good health and prosperity. Imagining that you are stable and not progressing in your illness. Imagining that your health is improving every single day, that you are stable and not deteriorating in your illness will help you to restore your health.

2. ***Positive Affirmations***

   Affirmations simply mean choosing your thoughts consciously to create positive results in the future. These are statements that you say out loud or quietly repeat to yourself. According to the research on brain functions, saying these affirmations affect the Reticular Activating system, the part of the brain which tells us what to pay attention to and what not to. It is an effective technique to reprogram the mind in a more positive way. Some of the affirmations for health are described as follows:



   - Every day in every way I am getting healthier and feeling better and better.
   - Every cell in my body is health conscious.
   - Every passing day, my body becomes more energetic and healthy.
   - I breathe deeply, exercise regularly and feed only nutritious food.
   - I am perfectly healthy in body, mind and spirit.
   - My mind and body are in perfect unison.

3. **Goal Setting**

   Setting goals gives a direction to our efforts. It is important to decide what health goals you want to achieve and in what period of time. And setting of goals should be SMART.

- SPECIFIC - Each and every small goal should be clear and be specified. For ex: You want your hands to start moving so that you can perform upper head activities.

- MEASURBLE - A goal that can be measured after a certain period of time. For ex: you can walk with the walker for 10 minutes now. So the goal can be - You want to walk with the walker for 30 minutes in the next 3 months.
- ATTAINABLE - The goal that is set should be within your reach.
- RELEVANT - It should be relevant to your current physical condition.
- TIME - It is extremely important to set a time limit for the goals - both short-term and long-term goals. For ex: "You want to start walking with the walker so that you can go back to your work in the next 1 year."

Chapter 33

Assistive Aids and Appliances

Introduction:

ALS is a progressive and degenerative condition. The role of assistive devices in ALS differs in different stages of the disease. In the initial phase of the disease, the role of the assistive devices is to prevent the functional decline whereas in the later stage, assistive devices help the person to restore the functional independence.

Assistive technologies may be defined as "any item, device, or piece of equipment that is used to increase, maintain, or improve the functional abilities of persons with disabilities"

For people without disabilities, technology makes things easier. And for people with disabilities, technology makes things possible.

Classification of Assistive Devices:

National Classification System for Assistive Technology Devices and Services divides assistive technology into the following classes:

- architectural elements
- sensory elements
- computers
- controls
- independent living,
- mobility,
- orthotics/prosthetics
- recreation/leisure/sports
- Modified furniture/furnishings
- Services

The assistive technology continuum, low tech tools, medium tech tools and high tech tools.

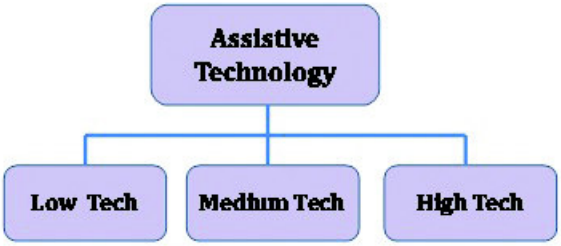


Fig 33.1. Continuum of assistive technology

Stages of ALS and use of Assistive Devices and Technology:

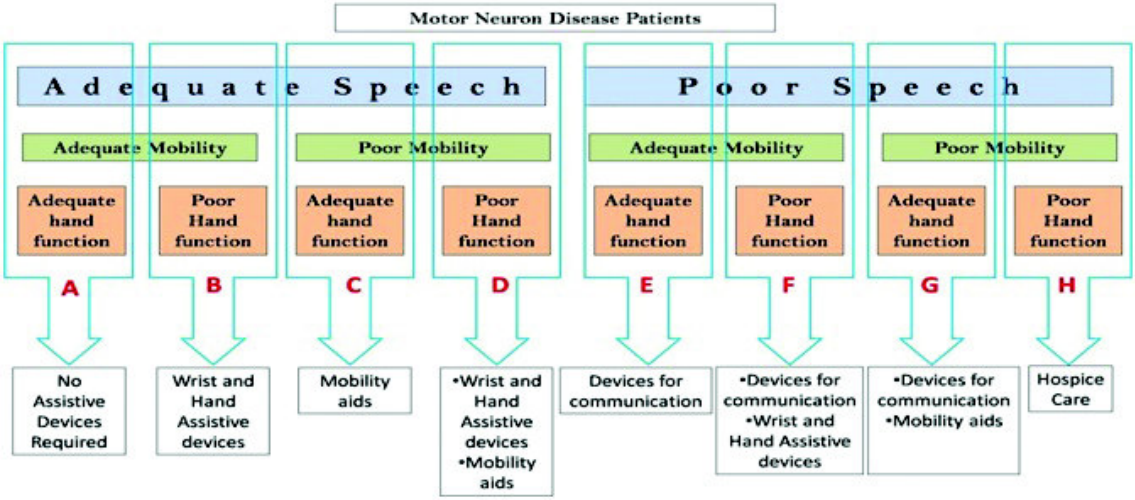


Fig 33.2. Stages of ALS and use of Assistive Devices

Assistive Devices For affected Hand Functions:

Fig33. 3. Assistive Devices for Hand

Universal Cufff : It is a device to be worn on the hand to hold objects like comb, toothbrush, spoon.

Page Turner : This device can be used for people who have hand muscle weakness.

Modified Pen grippers : By enlarging the size of the pen / pencil, this device helps in improving the writing of those individuals who have an intrinsic muscle weakness of the hand. Modified grippers help in improving the quality of the handwriting.

Writing Device : It's a customised device to which a pen / pencil can be attached, to facilitate writing activity. Its is advised for those who have difficulty in writing, signing on the cheques.

Modified nail Cutter : Holding the nail cutter requires good strength of lumarical and interossei muscles of the hand. Modified nail cutter is given for people who have poor hand functions. In this, the nail cutter is mounted on a small rectangular platform and it can be operated with the help of the palm of the other hand.

Swivel Spoon : It is given for those who have a limited hand to mouth movement because of muscle weakness.

Fig 33.4. Assistive Devices for hand

Plate guard: It reduces the chances of spilling food.

Jar Opener: It makes the function of opening the jar a single handed function. People with a weak grip also can open the jar with the help of jar opener.

Long Handle scrubber: It can be given for the patients who are not able to do over-head activities.

Fig. 33.5 Assistive Devices for hand

Long handle hair brush: Grooming activity can be made easy for those who have proximal shoulder muscle weakness by giving this adaptive device. It reduces the distance between hand and head.

Long loop scissor: Cutting can be made easy by pressing the scissor with the help of the palm. It is recommended for those who have minor muscle weakness of the hand.

Built-up Handle spoon: Activity of eating can be made easy for those who have poor hand functions by providing built up handle spoon.

Built up handles increase the girth of the spoon / fork / knife and make it easy to hold.

Cup holder: It provides more stability and helps to save energy.

Modified steering wheel: A knob is provided on the steering wheel for those who have difficulty in holding the steering wheel. Additional modification in the form of a keypad with additional functions can be added for those who have limited hand movement secondary to muscle weakness.

Fig.33. 6 Assistive Devices for hand

Zipper: Adding a loop to the zip can reduce the patients' assistance level.

Bottle Opener: It is recommended for persons with poor grip and difficulty in doing hand rotation movements. It stabilizes the bottle and makes the activity of opening the bottle one hand activity.

Modified Door Knob: Enlarged door knobs or long lever door knobs are used for patients with hand muscle weakness.

Dressing Stick: It is used for those who have affected over -head functions.

Tap opener: Patients who have difficulty with grip and rotating the wrist can use tap openers.

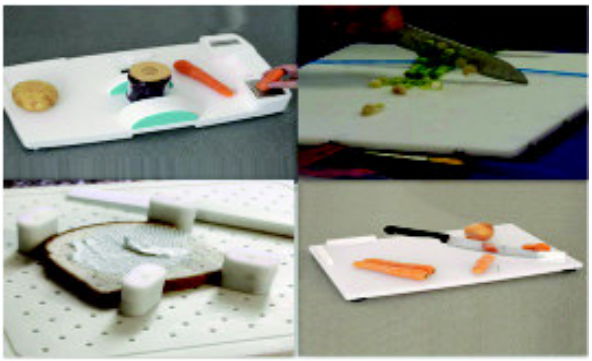


Fig. 33.7 Assistive Devices used in Kitchen

Pivot Knife: It has one end fixed therefore the cutting activity with the help of a single hand is possible. Vegetables / fruits can be cut by using palm for people who have a weak grip.

Stabilizer: It helps to stabilize the objects like bread slice, fruits.

Enlarged handle knives: These can be used by people with reduced muscle strength in hand.

Fig. 33.8 Utensil Holder

Fig. 33.9 Jar Holder

Fig.33. 10 Peeler

Fig. 33.11 Modified Shredder

Modified Knives

Fig 33.12. Modified Knives

Utensil holder: Although a person might have poor hand muscles, he can hold the utensil with the help of 'Utensil Holder'. Side supports of this device rest on the forearm to hold the utensil.

Cup / Jar Stabilizer: It minimizes the excessive energy expenditure by stabilizing the cup / jar. Isometric contraction of the muscle is avoided that saves energy.

Looped peeler: This is recommended for those who have difficulty in holding the peeler because of poor grip. The semicircular loop rests on the upper part of the fingers and makes the peeling activity easier.

Modified Knives : It makes the activities like cutting vegetables / fruits, mixing the vegetables / fruits easier. People with poor hand grip can hold the modified knives as they have enlarged handles.

Assistive Devices For affected Transfers and Mobility:

Different types of transfer devices and mobility aids can be used in different stages of the disease.

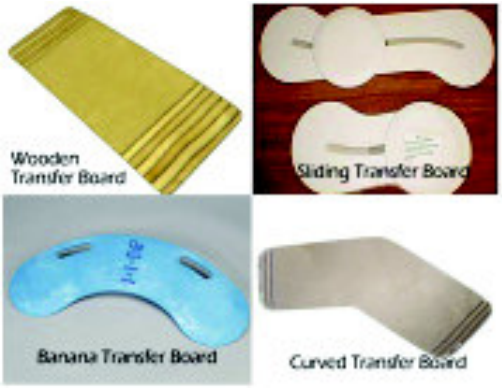


Fig. 32.13 Transfer Boards

Wooden Transfer Board: It is cost effective and can be made from local resources.

Sliding Board: It helps to save energy as it provides smooth sliding transfers.

Curved Transfer Boards: The board can be placed either curve forward or backward depending on the transfer situation and surrounding obstacles (eg. Wheel of the wheelchair). Its Low friction upper surface allows the user to do smooth transfers.

Fig 33.14. Assistive Technology for Transfers

Hoists: Hoists provide a way to transfer somebody with limited mobility without putting unnecessary strain on the caregiver or the person being moved.

Different types of hoists are as follows:

Mobile hoist

A hoist with wheels that can be moved along the floor - used for lifting a client inside a sling or on a stretcher designed for use with hoists.

Standing hoist

A specific type of mobile hoist designed to assist people between sitting and standing positions. Standing hoists are designed to fit under and around chairs.

Ceiling hoist

A hoist attached to a permanently mounted ceiling track that moves a client inside a sling. Gantry hoists have overhead tracks mounted on wheeled frames.

Mobility Aids:

Fig. 33.15 Mobility Aids

- The choice of mobility aid depends upon whether the person has adequate mobility, whether the person has adequate hand functions and the needs of the user.
- Persons having affected mobility but good hand functions can use a simple walker or a manually operated wheelchair.
- Persons with affected mobility and affected hand functions can use electric wheelchair.
- Electric scooters can be used for outdoor mobility.
- Electric mobility aids help to save energy and reduce fatigue.
- For persons with poor mobility, poor hand functions but good oromotor strength, sip and puff type wheelchair can be advised.

Assistive Devices for Affected Speech

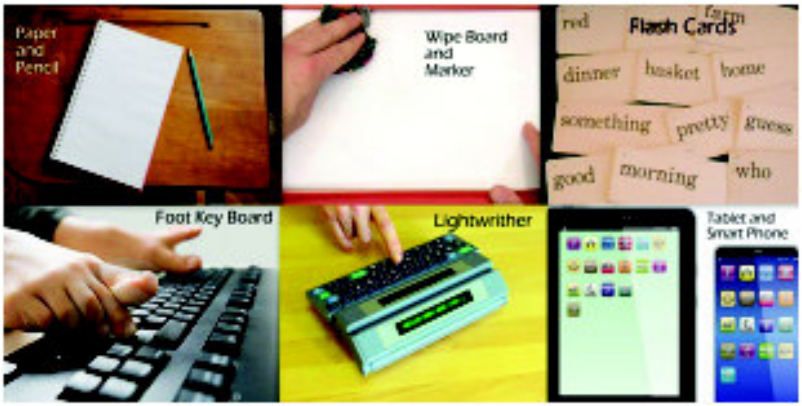


Fig 33.16. Assistive Devices for affected speech

Fig 33.17. Assistive Devices for affected speech and communication

Light writer: It is a text-to-speech device given for patients with poor or no speech. The person types the message on the key board which is displayed on two displays one facing the user and other facing the person sitting in front of the user.

Foot Keyboard: It is given for the patients who have poor hand functions but good lower extremity functions.

Laser Operated Head Mouse:

The sensor is placed vertically below the computer screen and is operated by a small laser pointer mounted on a hat or headband.

Eyegaze operated Communication Device (Mega Bee):

The caregiver tracks patient's eye movement and uses eyegaze as the method to determine letter locations, which are then displayed on the LCD screen below. It is fully portable and does not require a computer for its basic operation.

Speech Synthesizer: Artificial production of human speech can be done with the help of a computer system called speech synthesizer and it can be implemented into the software or hardware products.

Hands free Mouse (Smart Nav)

It is a hands-free mouse alternative that allows complete control of a computer by naturally moving the head in different directions.

Mouse Alternatives

Fig. 33.18 Track ball

Fig. 33.19 Switches

Trackball is a pointing device consisting of a ball held by a socket containing sensors to detect a rotation of the ball about two axes-like an upside-down mouse with an exposed potruding ball. The user rolls the ball with the thumb, fingers, or the palm of the hand to move a pointer.

Switches

They are available in various sizes, shapes, colours, materials and sensitivity. They are easy to activate and use. It requires no software setup.

Plug and play switch interface is also available.

Keyboard alternatives (Adaptive keyboard):

Fig. 33.20 Clevy Keyboard

It has bigger keys and bigger characters.

It has different coloured keys for different functional areas. It has a key repeat on and off function.

Virtual Keyboard

Virtual Keyboard allows people with mobility impairments to type data by using a pointing device.

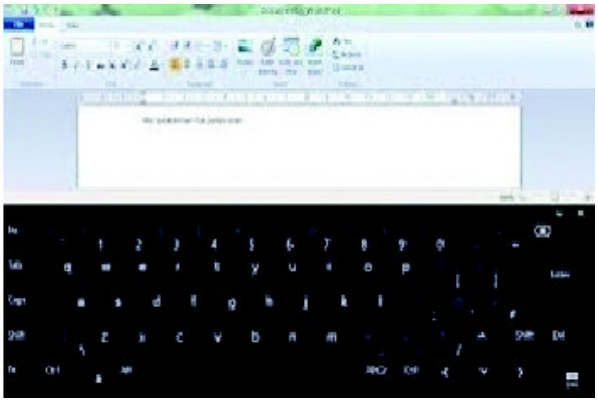


Fig. 33. 21 Virtual Keyboard

Google Voice :

It provides call forwarding and voicemail services, voice and text messaging, The service was launched byGoogle[2] on March 11, 2009

Fig. 33.22 Google Voice

Assistive Devices for Breathing Difficulties:

For patients with breathing difficulties, assisted ventilation is required. Assisted ventilation can be divided into two categories.

1. Non invasive Ventilation

- Pressure cycle ventilator machines (CPAP, BiPAP)
- Volume cycle Ventilators
- Negative pressure Ventilators

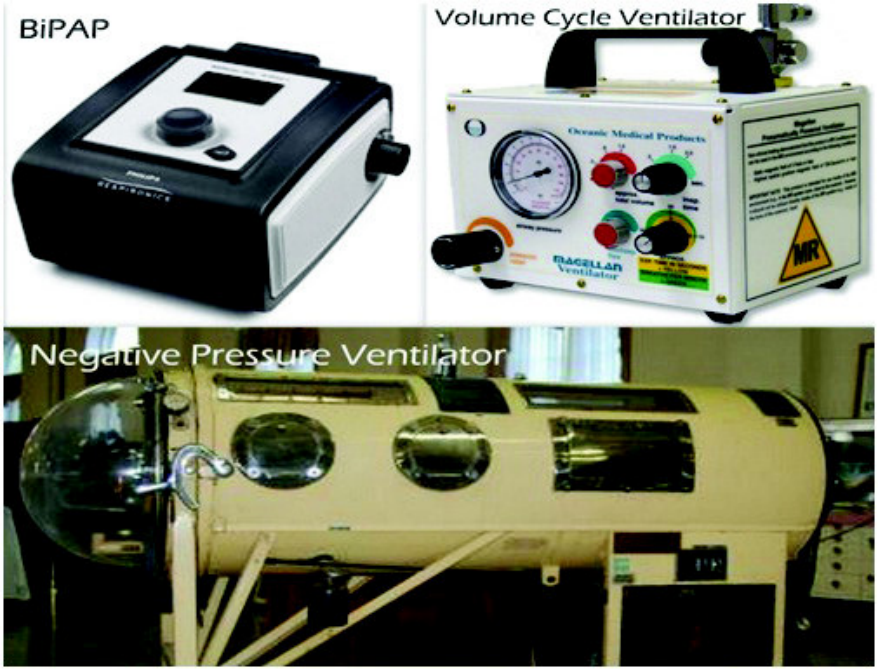


Fig.33. 23 Assistive Devices for Breathing Difficulties

2. Invasive ventilation

Tracheotomy is carried out for invasive ventilation. It is the surgical procedure carried out for invasive ventilation in which a hole is created in the wind pipe through which air in a regulated amount is forced in.

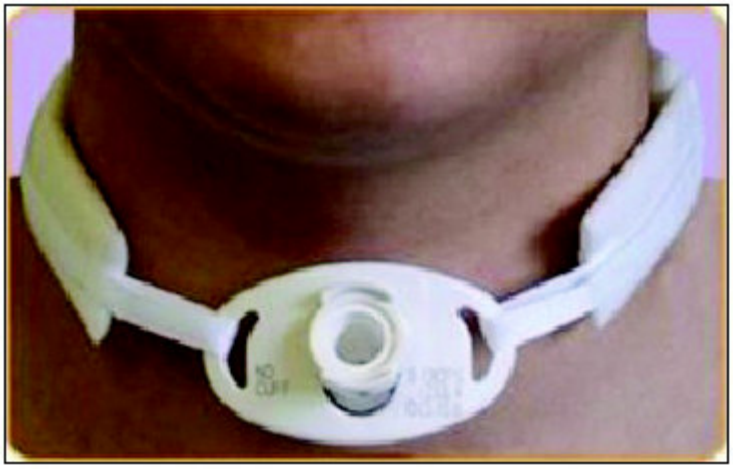


Fig. 33.24 Tracheotomy

Assistive Technology for Safety:

Personal emergency response systems (PERS), is an assistive technology developed to help caregivers and individuals who are at risk to stay independent at home. It includes an alarm system, to which electronic sensors are attached. Sensor detects the alert level. The alerts can be customized according to the individuals' risk levels. If the alert is triggered, an automatic message is sent to the caregivers or the nearby centre working with an individual.

Eg. Fall detection devices, SPO2 detectors.

Advantages of Assistive Devices and Technology:

1. Improves functional independence.
2. Improves quality of life.
3. Saves money by reducing caregiver time.
4. Maintains physical, social, emotional well being of the patient.

Disadvantages of Assistive Devices

1. The mismatch between the person and the assistive technology device.
2. Mismatches result in difficulty with using a device, having the device perform differently than expected, frustration, and even stopping use of the device.
3. A mismatch of technology can be costly and time consuming.
4. Inadequate training of the person with the device is also a barrier to proper use and acceptance of the device.
5. Certain assistive devices may not be as effective for people with multiple impairments.

Role of Caregivers in using Assistive devices

- Caregivers should understand the use of assistive devices and learn how to use the assistive devices.
- Training the person who is going to use the assistive device is an important responsibility of the caregiver.
- Finding out the resource on the assistive devices and receiving information on assistive devices and technology.
- Finding the rehabilitation specialist and seek knowledge from them to overcome the barriers in using the assistive devices.

The use of assistive devices differs from person to person. It depends on the type and stage of the disease. Assistive devices can help in reducing the need of human assistance but they can't completely replace the human assistance.

Training of the caregivers who are dealing with the handling of these assistive devices is essential. Therefore caregiver education should be carried out simultaneously along with patient education.

To get more detailed information about the assistive devices and technology for independent living refer to the following websites:

www.activeforever.com, www.pattersonmedical.com http://www.rehabmart.com

Chapter 34

Home and Environmental Modification

"My disability exists not because I use a wheelchair, but because the broader environment is not accessible"

Introduction:

- Home modification and environmental modification plays a vital role in the successful rehabilitation of the person with ALS. Environment is a place where the individual has to carry out his ADL activities or work includes natural as well as artificial objects. So, the environment includes individuals home, workplace, neighborhood, community.
- Access is defined as the means or opportunity of the individual to approach or enter the place.
- Having a good access to the home and surrounding environment is the first step to be taken into consideration.

Need of home and environmental modification in MND

- Home and environment plays a vital role in maintaining functional independence.
- Falls can be prevented by doing necessary home and environmental modifications right from the early stage of the disease
- Home modification makes life more productive.
- Thinking ahead to plan and doing the necessary modification before its needed is important.
- Home modification can be classified into categories like Early modifications and Late modifications.
- Early modification leads to improved functional independence and successful and independent participation of an individual in a desired meaningful activity.
- Modifications which are done in a later stage of disease helps to improve quality of life of the person.
- Another way of classifying home modification is : Simple modifications and

Elaborate modifications.

- Simple Modifications are as simple as re arranging the furniture to modifying the door way opening . whereas elaborate modification consists of re modeling the room, installing high tech equipment in the room.
- Environment is a setting where occupational behavior of the person takes place.

- So it can be home, community, workplace, garden , mall, auditorium.
- Environmental modification can be succcssfullly done if the caregiver understands the physical and spatial properties of the caregiver.
- Proper communication between patient , caregiver and the therapist is an important aspect environmental modification

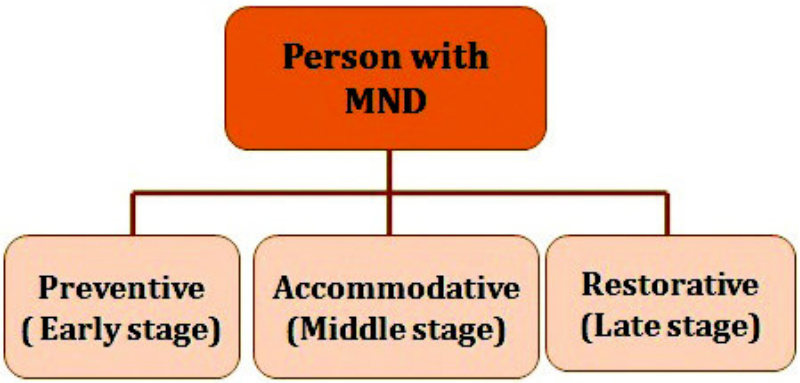


Figure 34.1 : Approaches Used

Different Approaches used :

Preventive : Maintenance of current health status and prevention of further decline.

Eg. Putting grab bars in the bathroom for fall prevention.

Accommodative : Use of compensatory strategies to overcome the limitations.

Eg. Using uplift seat or swivel cushion for making it easier to get up from chair.

Restorative : Rehabilitative techniques used to regain maximum functions.

Eg. Using a soft collar for neck muscle weakness.

Creating Disabled friendly Home and Environment

Disabled Friendly Home

Flooring

- Non skid tile flooring.
- Use non skid mats
- Avoid thresholds

Figure 34. 2 : Non skid Tile and Non Skid Mat

Doors

- Door width should be wide enough
- Automatic opening doors, sliding doors ,avoid revolving doors.
- Sliding door is advisable in narrow space.
- Modified door handles are advised for user who have hand affectations.
- Electronic door opening system can be used.

Corridors

- Narrower hallways restrict the accessibility of an individual who is using walker
- For one way traffic the width of the corridor should be 1.2 meters.
- For two way traffic corridor, the width should be 1.5 meters.
- For a corridor connecting to another perpendicular passage, should be 0.90 meters
- Door placement should be at least 0.55 meters away from the end of the corridor.
- Sufficient space must be kept for taking 'U' Turn . 1.2 Meter Width should be maintained

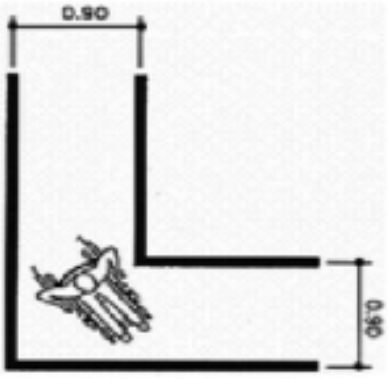


Figure 33.3

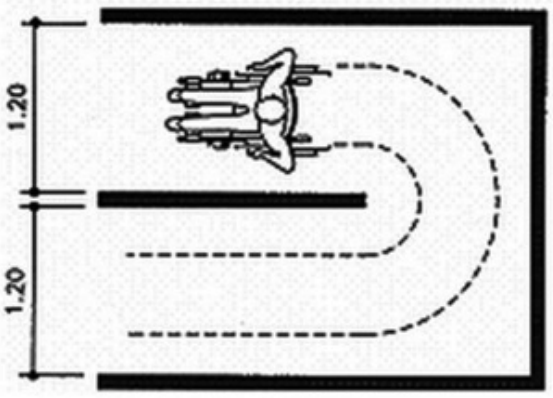


Figure 33.4

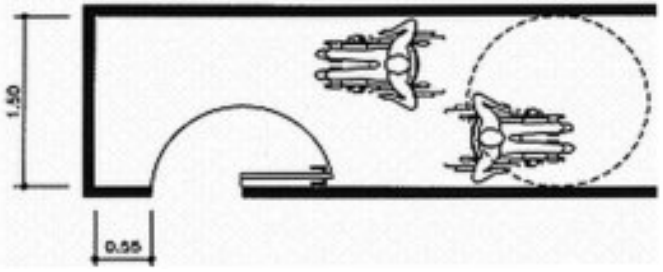


Figure 34.5 Corridors for Wheelchair Accessibility

Stairs:

- Staircases should not be too steep
- The height of steps should be uniform, in case of variation of the height it should be highlighted.
- There should be grab bars on either side of the stairs.
- Staircase should be wide enough to allow for two people stand side by side
- Spiral staircases are not recommended
- There should be appropriate landing (at least 1.2 mts along the entire width of the staircase) at the end of the flight of stairs, marked by tactile markings
- The width should be 0.9 mts for one way traffic and 1.5 mts for two way traffic
- Flooring should non-slippary
- The grab rails should be continuous even around the circumference of the landing, if the staircase if wider than 3 mts an intermediate hand rail should be present and it should be at the height between 0.9 mts to 1.4 mts.
- For every staircase an alternate elevator, ramp or lift should be available.

Railings and Handrails:

- All the hazardous areas, corners, open areas with a risk of slipping must have railings.
- Handrails should not obstruct the path of travel
- The height of the handrails should be between 0.85 to 0.95 mts above the finished floor levels
- For the benefit of wheelchair users a second handrail at the height of 0.70 and 0.75 mts should be places
- Railings should mounted on the wall with proper support structures to withstand heavy load
- At the end the railings should bend and blend into wall rather than leaving them open ended

Kitchen and Bedroom

- Using modified Kitchen platform
- Use of assistive devices like adapted knives, spoons, forks (eg. Built up handle spoons, Swivel spoon, forks, long handle spoons)
- Universal cuff for holding equipments used for cooking.
- Using Plate with plate guard.

- Firm chairs with arm and back support
- Furniture modification
- Smoothening of the sharp edges of furniture
- Repositioning the electrical switches and boards at an accessible height according to the users convenience.
- Using fabricated / wooden blocks to increase height of the chair and bed
- Use of Uplift seat
- Use of Transfer boards and Assistive equipments for transfers
- Use of chairs with mobile arm support
- Chairs with adjustable back support

Figure 34.6 Kitchen Modification

Figure 34. 7 Furniture Extenders

Toilet and Bathroom

- Insufficient space inside the rest room is the most common problem
- Rest rooms must have sufficient space at the entrance and inside to maneuver the wheelchair
- Approach to the restroom should be parallel to the toilet.
- Use of electronic devices like battery operated brush, shaver.
- Use of soap mitt for bathing
- Removing the door thresholds makes it easier for the wheelchair access.
- Installing grab bars in toilet and bathroom
- Use of plastic non skid chairs with arm support.
- Use of wider opening door hinges
- Shelves and basins should be fixed at an optimal height of the users.

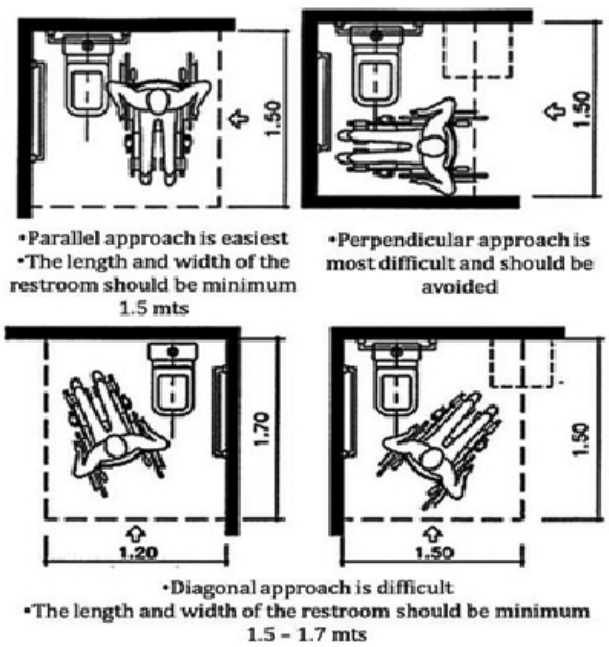


Fig. 34.8 Approach to the washroom

Some types of flooring may not hold up under the weight of the power wheelchair. Flooringchanges like installation of nonskid mats can be done

- Recoating floors, pressure-sensitive abrasive strips can further improve safety and reduce risk of falling. In addition, resilient, non-slippery flooring prevents or reduces foot fatigue

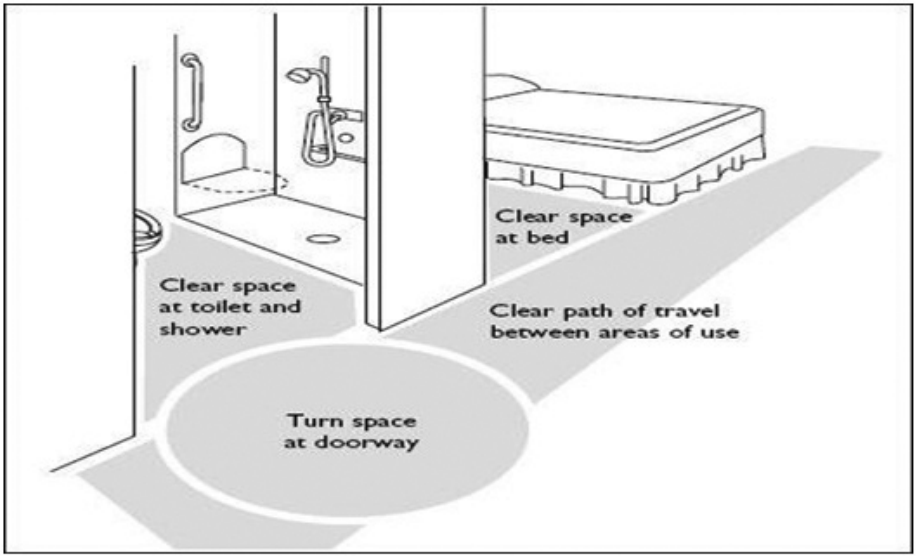


Figure 34.9: Bathroom and Toilet Modification

Figure 34.10 : Toilet modifications

Figure 34.11 : Bathroom modifications

Fall Prevention techniques:

You can reduce the risk of slipping on wet flooring by:

- taking your time and paying attention to the floor surface and floor height
- adjusting your stride to a pace that is suitable for the walking surface and the tasks you are doing
- walking with a slightly broader BOS
- making wide turns at corners

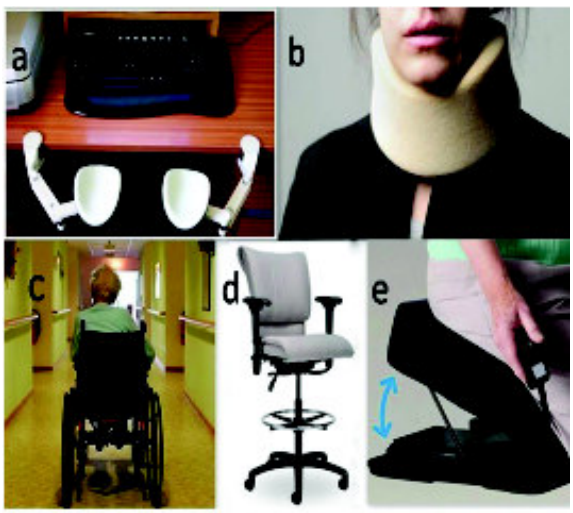
You can reduce the risk of tripping by:

- keeping walking areas clear from clutter or obstructions
- keeping flooring in good condition

- always using installed light sources that provide sufficient light for your tasks
- using a flashlight if you enter a dark room where there is no light
- ensuring that things you are carrying or pushing do not prevent you from seeing any obstructions, spills, etc.
- Both slips and trips result from some a kind of unintended or unexpected change in the contact between the feet and the ground or walking surface. This shows that good housekeeping, quality of walking surfaces (flooring), selection of proper footwear, and appropriate pace of walking are critical for preventing fall accidents.

Work

- Professions which require sitting activities and supervision work are most suitable vocations.
- Use raised chairs, turning chairs for energy conservation and work simplification.
- Grab bars and hand rails should be installed thru out the work place including toilet and bathroom for prevention of falls .
- Use of 'neck collar' for maintaining upright position of neck while working for person who are having neck muscle weakness.
- Use of assistive devices and technology is always advisable. Eg. Use of writing device, computer for writing.
- Key board modifications, mouse modifications enhance the quality of work and helps in saving energy and reduces easy fatigability.
- Lifts/ Elevators should be used if available instead of climbing stairs
- Chairs with adjustable arm rests
- Shelves should be placed at a reachable height and distance.



a. Mobile arm Support
b. Neck Collar
c. Handrails at work place
d. Rotating and Raised Chair
e. Seat uplifter

Disabled Friendly Transport

- Car with ramp, Bus with ramp
- Modified steering wheel
- Automobiles without gear
- Battery operated four wheeler
- Platform height should match the train height for easy accessibility.
- Putting symbols / markings on the platform at the position of the doorway
- Train doorway modification
- Electronic door opening system
- Providing separate sitting arrangement in the bus / Trains for person with MND.
- Trained staff should be assigned at railway station, bus depot to help person with MND and caregivers.

Figure 34.13 : Disabled friendly transport a. Doorways for metro railway access b. Adjusting the height of railway and platform c. and d. Ramp for bus access

Figure 34.14 car and two wheeler modification

Parking :-

For parking of vehicles of handicapped people the following provisions shall be made:

- Surface parking for two care spaces shall be provided near entrance for the physically handicapped persons with maximum travel distance of 33 M from building entrance.
- The width of parking bay shall be minimum 3.60 Meter.
- The information stating that the space is reserved for wheel chair users shall be conspicuously displayed.

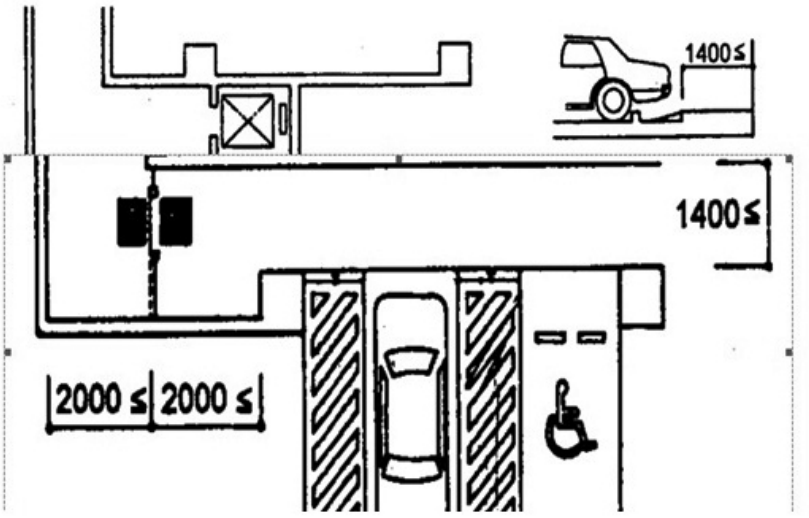


Fig 33.15 parking modification

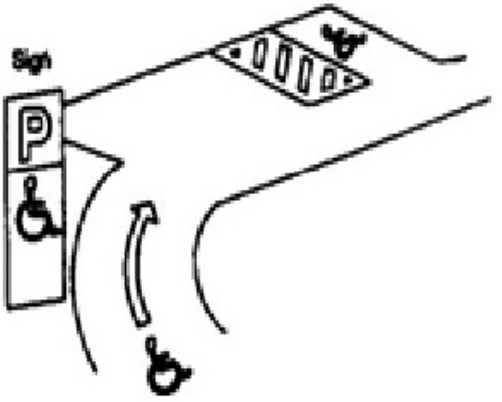


Fig 34.16 Access aile

An access aisle 1.20 m wide can be located between two ordinary parking spaces (fig. 18).

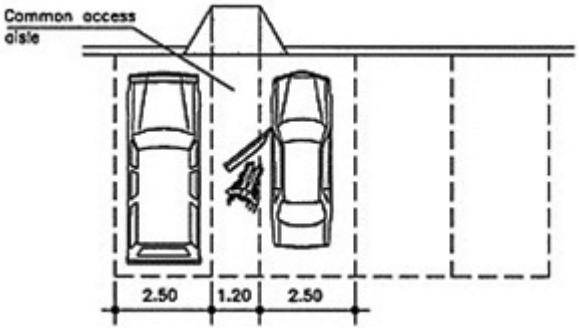


Fig 34.17 Indoor Parking Lot

Where parking spaces are angled, the extra space at the end of a row can be used as a parking aisle for disabled person.

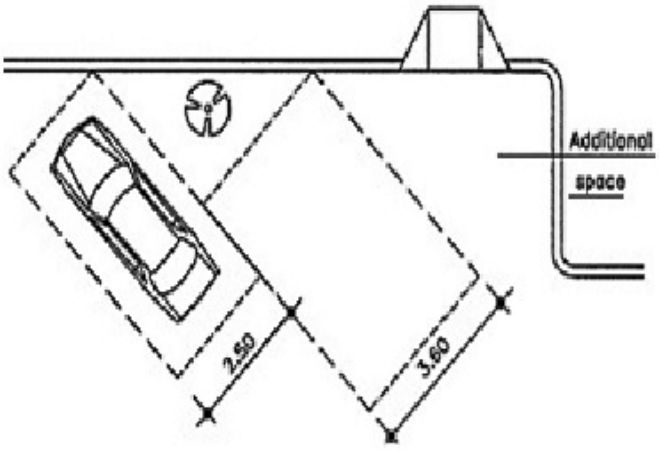


Fig 34.18 Parking curb

Disabled Friendly Public Places

- Spacious elevators and lifts
- Providing sitting arrangements (chairs, benches) near the waiting area of the mall, theatre.
- Accessible washrooms, toilets for wheelchair users.
- Separate cue for persons with MND at mall, park, theatres.
- Auditorium seats should be near to the exit door
- Ramps at the entrance of the park, malls, theatre
- Hand rails on the ramp.
- Curbs on the footpath

- Separate tracks for wheelchair in the park
- Trained staff for taking care of person with MND
- Accessibility at public places like temples, stadiums, musiums makes the lives of isabled person more fulfilling

For People with adequate mobility :

- Walk straight across the road - don't jay-walk.
- Keep checking in both directions to make sure the way is clear.
- Try not to cross the road from between parked cars or near trees and bushes as drivers may not see you.
- Avoid crossing near a bend or crest in the road. Give yourself a good chance to see vehicles coming from both directions.
- Use the zebra crossing for crossing the road

For People without adequate Mobility :

- Using portable curb place assists in better mobility and functioning at public place.
- Ramps at public places allows the better access for wheelchair bound patient to the public places.

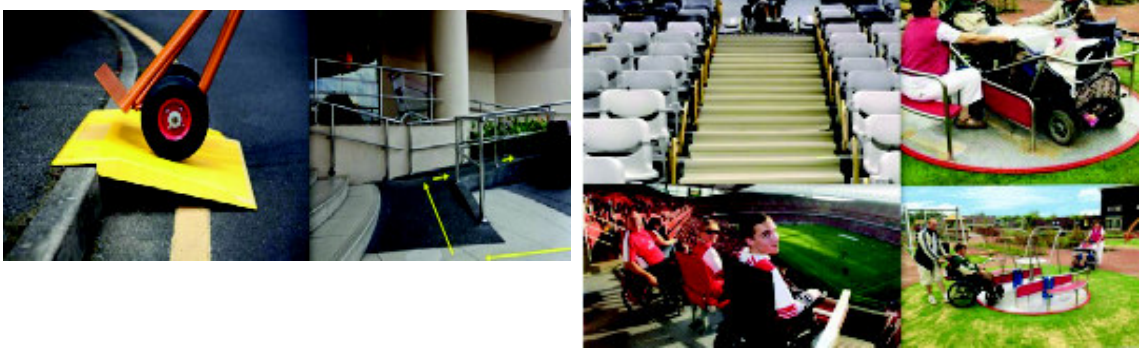


Figure 34.19, 20 : Disabled friendly public places

Other Places : Counters

To make a counter easily accessible for a wheel chair user, allow a space about

700mm high and 350mm deep under the counter.

Water Fountains (Drinking)

Allow sufficient space around the water fountain to make it easily accessible for wheel chair users. Depending on the type of water fountain allow a space about 700 mm high and 350 mm deep under the fountain.

Telephones

Allow a space about 700 mm high and 350 mm deep under the telephone stand. The telephone receiver must be placed at a height of 1 IO cm or less.

Mailboxes

The mail slot must be located at a height of 1200 mm or less.

Vending Machines

The coin slot must be located at a height of 1200 mm or less.

Taxi Stand

For wheelchair users to be able to approach a taxi easily, sudden level differences from the taxi stand to the road need to be eliminated

Taxi Interior

It is recommended that taxis be adapted to allow passengers to get in and them while remaining seated in their wheelchairs.

Fig 33.21 Taxi Modification

Auditorium

Applies to wheelchair spaces in auditoria, assembly halls, theatres and similar facilities.

* Accessible seating space should be provided in a variety of locations to give persons with physical disabilities a choice.

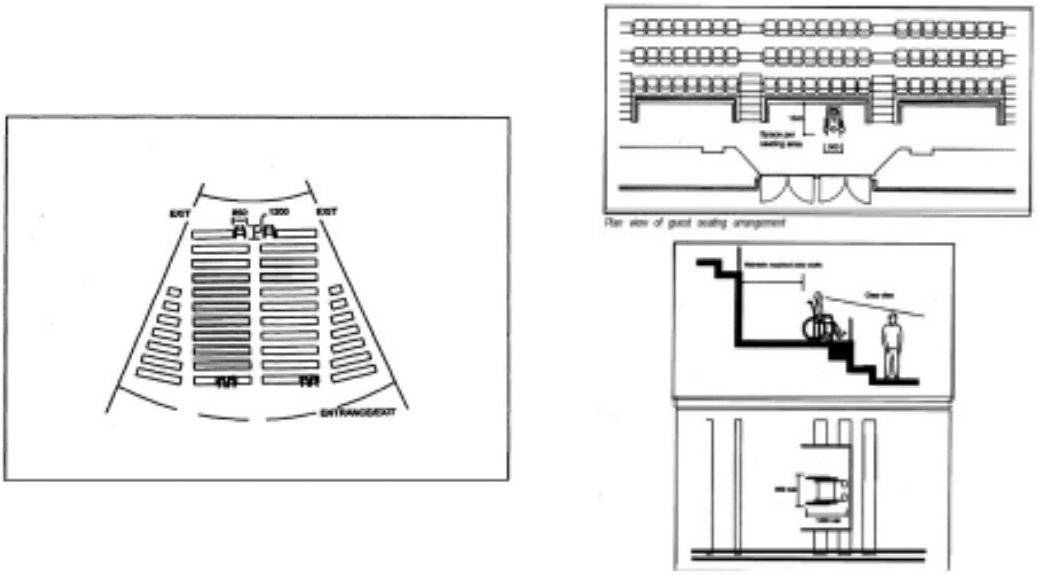


Fig 34.22 Design of an auditorium for wheelchair access

"However difficult life may seem, there is always something you can do and succeed at… Stephen Hawking

Following picture shows , the public places in India which are made accessible for persons with disabilities,

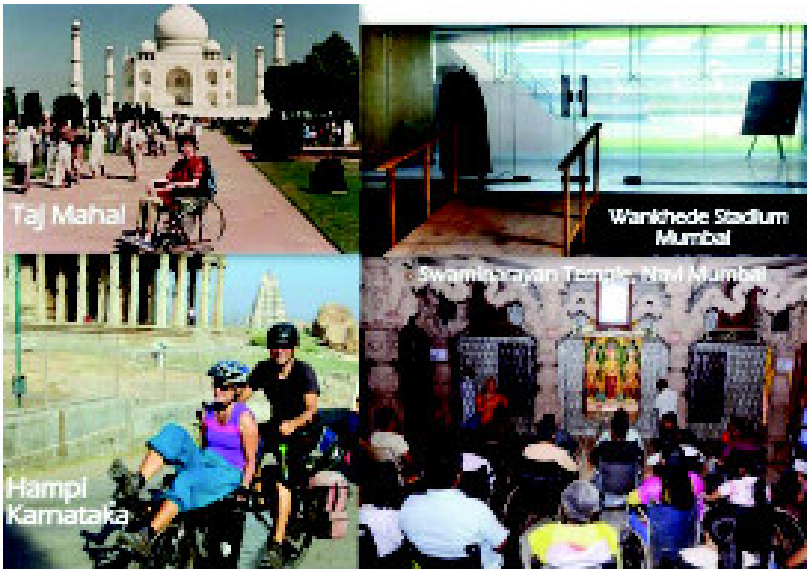


Figure 34.23 Disabled friendly public places a. Taj mahal, b. Wankhede stadium, c. Hampi Karnataka, d. Swaminarayan Temple Navi Mumbai

Summary :

In short, creating the disabled friendly home and environment by using adaptation ,modification and compensatory techniques helps in improving quality of life and leads to meaningful and successful life.

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2. Physical Rehabilitation 5th Edition Susan O Sullivan Chapter 20 Amyotrophic lateral Sclerosis

3. Willard and spackman's occupational Therapy 11 th Edition

4. Willard and spackman's occupational Therapy 9th th Edition

5. MDA ALS caregiver's Guide

6. www.barrierfreehome.com

7. http://www.un.org/

Chapter 35

Support from family and friends

Living with MND can be the most challenging aspect of a patient's life. Therefore, an active role played by his family and friends can help the patient cope up with his difficulties and lead as normal life as possible. The family may have to go an extra mile to make things easy and comfortable for the patient. Some of the ways are as follows:

- **Home Visits** -Friends, cousins or extended family can pay regular home visits to the patient so that he/she does not feel isolated. They will have someone to talk to, share their feelings, worries and concerns or talk about things in general. It could only be a meeting for 2 hours in a week, but it will make a lot of difference to the patient's emotional and psychological well-being.
- **Parties** - Friends or family can plan their birthday parties at home or celebration of festivals with the patient. This way, the patient will have a change from his regular monotonous life of only exercising and being at home. Celebrating festivals together will also help bring people together and make the patient feel wanted and cared for.

"There are no traffic jams on the extra mile." - Zig Ziglar

- **Outings** - Regular outings such as a small drive or shopping to disable friendly areas can be planned so that the patient gets an opportunity to step out of his house. This will help the patient feel fresh, meet new people, see around and not feel that he/she is missing out on something.
- **Past Achievements:** Family or friends can have a discussion about the patient's past achievements in his life. This will boost his confidence in the present and make him feel good about himself. Reminisce about the happy moments spent together in childhood and with friends to make the patient feel that he has lived a meaningful life.

"Appreciate the small achievements. Reward yourself from time to time".

- **Decision-making:** The patient must be included in all the important decisions taken in the family. His opinion, feedback and suggestions hold the same value and respect as any other family member's opinion.
- **Humor** - Plan for a movie once in a while that is funny or humorous in nature.

Watch funny videos, cartoons, comedy shows to lighten up the mood. This will act as a stress buster and give him a chance to laugh and be happy.

- **Working together** - If the patient is working, assist the patient whenever necessary. Provide help, inputs and guidance. The use of assistive technology can play a very important role in helping the patient work from home. For example, people with limited hand function may use a keyboard with large keys or a special mouse to operate a computer, people who are blind may use software that reads text on the screen in a computer-generated voice, people with low vision may use software that enlarges screen content, people who are deaf may use a TTY (text telephone), or people with speech impairments may use a device that speaks out loud as they enter text via a keyboard .

- **Don't overprotect :** Provide help and support whenever necessary but do not over sympathize or over protect the patient. This may lead to over dependency on the part of the patient and will prevent him from being independent. Respect the patient's limitations as well as his strengths.

Chapter 36

Community Care and Vocational Rehablitation

What is Community Care?

Community care is a group of programs that is already established as a support system for an individual with disability, frail old people and the carers of both the groups to allow the individual remain in the community and lives in their own homes, as an alternative to institutional or residential care and helps them to live more independently.

Why in MND:

Community care is required for MND patients because of the ongoing progressive nature of the disease and the changing need s over the disease course. Also with the increasing number of cases it is posing a challenge on the health care providers to meet the demands of the individuals with MND at the institution level. Community Care providers help to provide with interventions that will improve the survival and quality of life for individuals living with MND and also for their carers. Also community care help bridge a gap between hospital and community. Unfortunately in India, there are no such active support groups available for individuals suffering from MND, or if available it is done at an individual level.

What do the Community Care Providers do?

The care providers conduct regular assessments, to find out about an individuals needs, to determine the need of support services & gather information required for the delivery of supports services.

Who all are involved…??

The list of community care providers is varied for an individual with MND. It is a multidisciplinary team effort and likely to include the following professionals:

1. General Practioner (GP): A GP will provide you with information regarding the diagnosis and guide you throughout the course of the disease. He/she is able to prescribe treatment for the control of symptoms and can help for giving referrals to an appropriate specialist, other consultants and members of the multi-disciplinary team.
2. Consultant: It usually a neurologist or a consultant in respiratory medicine, who will provide with assessment, diagnosis and symptom management. Depending

on your symptoms you may be referred to other consultants, such as palliative care consultant.

3. Occupational Therapist (OT): An Occupational Therapist will advise you about everyday activities as well as moving and handling, hand functions, splinting, adaptations at home and at working environment, wheelchair prescription and modification, to increase the participation and improving the QOL.

Figure 36.1- Occupational therapist

4. **Physiotherapist:** A physiotherapist will provide with a range of interventions including, advice regarding mobility, falls prevention, positioning and specialist equipment and respiratory support. An Occupational therapist and a Physiotherapist both work very closely with each other.
5. **Community Nurses:** They offer help with nursing care and equipment provision
6. **Specialist Neurology Nurses:** A specialist nurse will give specific advice and support related to neurological conditions.
7. **Speech and Language Therapist:** They can give advice on need of communication aids if speech becomes a problem. They work closely with the dietitian and offer advice regarding compensating for swallowing difficulties and recommends safe consistencies of diet and fluids, if the throat muscles make eating and drinking a problem.
8. **Dietitian:** In consultation with the speech and language therapist the dietitian can advise on a diet which will help to maximize energy levels and reduce weight loss.

Figure 36.2 Dietician

9. Pharmacist: they advise on the best type of medication for symptom control, for example, many medicines can be dispensed in liquid form for ease of swallowing.
10. Specialist Palliative Care: Specialist palliative care services include consultants in palliative medicine; specialist community nurses and hospice teams and provide symptom management, support, information and counseling, to assist the person living with MND and their carers and family from the point of diagnosis.
11. Psychologist and Counselors: they provide with emotional and psychological support. They help assist with understanding and managing situations and relationships associated with the individuals need for care.

Figure 36.3- Clinical psychologist

12. Social Worker: A social worker will be able to offer advice regarding a range of services including financial support, help for carers and children and, if needed bereavement support.
13. House work and Home maintenance: assistance with housework and chores and assistance with home and repair to ensure it is a safe place to live.
14. Personal care assistants for daily self-care tasks (eg. eating, bathing, toileting, dressing, grooming, getting in/out of bed & moving about the house; may also include monitoring medication).
15. Transport: Assistance with travel to/from essential appointments & social activities, either directly (in a car or bus) or indirectly (through a taxi subsidy or voucher); for groups or individuals (eg. medical transport, non-medical transport, shopping buses, social groups).
16. Social Support: Assistance in the home or community to meet an individuals need for social contact & help him/her participate in community life (eg. visiting & telephone monitoring, letter-writing).

Apart from this community care providers can help in raising an awareness regarding MND condition, prevent, support and help in fund raising to help families of individuals with MND.

Vocational Counselling / Vocational Rehabilitation:

WORK is the most important occupations in which an adult engages. Work is important for individuals with disability to:

- To become financially independent and contribute as an earning member of family
- Receive health insurance and other benefits
- It gives an opportunity for them to meet and interact with other people
- It boosts their psychological well being and self esteem and overall life satisfaction. This is also true for individuals suffering from MND. Barton stated "the purpose of work is to divert the mind, exercise the body, relieve the monotony and boredom of illness."

What is Vocational Rehabilitation?

Chan et al., (1997) defined vocational rehabilitation as:

"A dynamic process consisting of a series actions and activities that follow a logical, sequential progression of services related to the total needs of a person with a disability. The process begins with the initial case finding or referral, and ends with the successful placement of the individual in employment. Many activities and developments occur concurrently and in overlapping time frames during this process".

Although individuals with SCI go on to have active work lives and successful careers, they have more barriers to overcome than those without disabilities. Vocational rehabilitation programs help individuals with disabilities to

- Acquire relevant education and training
- Find new employment opportunities and apply for jobs
- Overcome accessibility issues that act as barrier in post injury gainful employment
- Additional training may be required by many individuals to sustain the employment
- Also VR programs help with work site assessments and onsite work task analysis at clients workplace

An Occupational therapist should

- Interview the individual during the inpatient rehabilitation regarding the previous jobs for the purpose of future planning and sharing information regarding the

vocation with the rehabilitation team, so that vocational goals can be incorporated early in the rehabilitation plan.

- A work site assessment and onsite work task analysis performed at the client's workplace as well as examination of the worker and the work environment will provide the therapist valuable insight about the present skills of the patient.
- If a patient is considering returning to work an Occupational Therapist can assist him by assessing his work abilities in a simulated work environment.
- Patient will have build up his strength and stamina and both he and the staff will have a clearer idea of his employment capabilities.
- The individual should be taught proper body mechanisms and energy conservation to perform work in a safe manner.

Types of accommodations/ Getting support at work

Job accommodations can include modifying work schedules, tasks or the work environment. Solutions can range from simple to complex. Examples:

- Removing a desk drawer or raising the height of a desk with four wood blocks so a wheelchair can fit underneath.
- Special software and hardware so a person with no hand function or speech problems can work on a computer.
- Shifting work hours to a later start and end to the work day to accommodate a worker's lengthy morning care needs.
- A person with MND may need an additional support in the form of extra supervision or personal care assistance during the work day that will help with positioning or personal care assistance during the work day.
- Also there should be facility for resting area to lie down for a short period at the workplace.

Many individuals suffering from MND have differences due to considerable functional limitation acquired during the disease the process. Sometimes it is not possible to return to work after MND, especially in individuals who show rapid deterioration e.g. bulbar onset ALS who have difficulty in speech and swallowing and breathing difficulties, in such cases a change of job to light duty or work from home options or voluntary retirement can be considered. Return to gainful employment is seen in only those individuals who are limb onset disease progression or in whom only the upper limb or lower limbs are involved. After diagnosis some individuals prefer to continue on with the pre injury employment or they prefer to work part time, and early fatigability are the major obstacles an individual faces that prevent them for returning to work.

Following are the contributing factors that influence the employment after MND:

- Easy fatigability
- Breathing and Swallowing complications
- Rapid progression and nature of the disease
- Inadequate educational opportunities appropriate to the individuals abilities causes job discontent and job dissatisfaction
- Insufficient health care insurance
- physical barriers
- and inability to access effective employment training and placement services
- Inaccessibility of the work places and lack of enabling technology to allow competative work output.
- No accessible transportation to and from work.
- Inconsistent attendant care that affects the work attendance
- Negative attitude about the employers and prospective employers about the abilities and inabilities of a diseased individual.
- Poor self image, fear of failure, lack of working peer role models and low self expectations off potential workers.

Role of Computer Technology in Vocational Rehabilitation and Return to Work

Computer use and training is of special benefit to individuals with MND because computer technology may help lessen the impact of mobility limitations that are inherent with this condition. The use of assistive devices such as hands free telephones, environmental control systems and mouth sticks also play an important role. However, the utilization of these assistive technologies is one of the major challenges that influence the employment success.

Chapter 37

Care for Caregiver

MND is a group of disorders that affects not just the patient, but his entire family. It has been seen that the burden of care giving is found to be highest in the spouse, as he/she is the primary caregiver. Major stressors identified by the caregiver include not leaving the patient by themselves for a longer period of time, physical and mental exhaustion, financial issues, an underlying guilt of not doing enough and feelings of helplessness. As the patient progresses in the disease, he gradually becomes dependent on the caregiver for physical needs, psychological support and basic household chores leading to increased burden on the caregiver. Therefore, support for the caregiver also has to start sooner along with the treatment of the patient.

It has been found that caregivers spend a lot of time during the day caring for the patient, almost up to 11 hours in a day. Those who believe that they cannot leave the patient for long tend to be the most distressed. They tend to neglect or not pay attention to their own physical health issues such as back ache, head ache, knee pain etc. Also, their emotional needs are not met as there is no support received most of the time. There are sudden changes happening to which the caregiver has to keep getting adjusted without getting the time to prepare oneself. Changes might be taking up a new job to take care of the finances or taking up additional household chores such as cooking, cleaning, dusting etc. As a result, their own personal and social life gets affected. They cannot find time for themselves and sometimes miss the normal life they used to spend earlier with the patient. Children of patients with ALS also undergo significant psychological and physical distress. They either tend to feel frustrated with the additional responsibilities on them or feel helpless as they cannot do anything to help the patient get better. Their quality of life is affected; they do not get privacy for themselves, cannot focus on their goals and ambitions due to constant interruptions in the family and there is fear of future with respect to finances and impending death of the parent.

Therefore it is highly essential for the caregiver to equally take care of their own needs as well:

- The caregiver should take some time out for themselves, may be once a week or once in 15 days. It will help if he/she distributes the responsibility of taking care of the patient with some other family member or a close friend.

- It is also important for the caregiver to tap his/her own social support system. It could be as simple as meeting a close friend or relative and venting out his concerns, fears and repressed emotions.
- It is extremely important for the caregiver to feel that whatever he/she is doing in his role of a caregiver is to the best of his ability. There could have been no other or better way of taking care of the patient. They need to feel positive about their role and find meaning in their act of care giving.
- If needed, the caregiver can take the help of a professional and undergo counseling sessions to deal with the depression and anxiety. Medications to treat depression can also be used if necessary.
- The caregiver can realize that if this role brings with it high costs, it also tends to give high rewards as well. These rewards can be emotional, psychological and spiritual such as growing ability in one's confidence, feelings of personal satisfaction and increased family closeness.
- The caregiver can also join some faith based groups to seek support and help.
- The family/ or the caregiver can also join caregiver support groups in the community wherein they can share their every day struggles and problems and feel that they are not alone.

Chapter 38

Supporting the Children

A young carer may be the child, brother, sister, other relative or friend of the person with MND. They may live with the person who has MND or live nearby and visit. Finding a balance between life and care can be difficult when young people are involved. Depending on the type of care needed, young carers may get involved in:

- helping younger brothers and sisters in their school activities and assignments.
- house work or laundry
- shopping for food and supplies, taking turns
- making meals
- helping the person with MND to eat and drink
- giving medication on time
- helping the person with MND to get washed and dressed
- Helping the person with MND to exercise
- providing emotional support and company.

It is important then at this point of time, a young person's needs assessment must be arranged for. The assessment will consider the impact on their wellbeing, school work or employment, social life and interests. A care support plan will be developed in agreement with the young person and their parents or guardian.

What can their school do?

If a young carer is struggling to keep up with schoolwork or has missed any school as a result of caring, it is best to keep the school informed. Services in schools provide support for a range of practical, emotional and behavioral needs. This type of support is often provided by teachers, or even other pupils who act as mentors. Ask the school what they offer and how this might help. Once aware of challenges the young person may face, they may provide a pastoral care plan (following discussion with the young person's parents or guardian - which may include the family). This can help the young person feel more secure at school and gives them a point of contact if they feel worried or upset about anything. Teachers can help by:

- allowing extra time for homework or during exams
- arranging support services through the school, such as counseling sessions
- providing information about external support services in the local area.

- introducing a young carer to other young carers in the school for peer support
- advising about school commitments and homework, especially if an emergency happens at home.
- A 2 day or 3 day retreat can be organized for the children. It is an opportunity to have fun, to be challenged through team building exercises and to meet other children who are in the same situation.
- Children can volunteer with other children of parents with ALS. They can form a small social circle of their own and organize events for themselves, such as assisting each other with their school assignments, going for small outings together so that they get a break from their duties and responsibilities.

Figure 38.1 Counselling

- Children can undergo timely counseling sessions with a professional therapist or a Psychologist wherein they can vent out their painful feelings and emotions. The therapy session can provide them with a safe environment where they can talk freely, ask questions and feel comfortable. Children can be prepared for the future in terms of their daily activities, additional roles and responsibilities and a sense of belief and pride in their abilities that they are doing the best that they can at every moment. Young adults or adult children of patients with ALS may also undergo feelings of guilt - for ex: constant feeling of not doing enough for the parent. It is important then to address such feelings and emotions so that they can take care of their additional responsibilities with a strong and positive attitude towards the future.

What can employers do?

If a young carer is employed, in either par-time or full-time work, they may worry about their job being affected by the demands of caring. In most cases, employers are willing and able to help. They may be able to:

- adjust working hours to suit the caring role
- allow flexible working, so hours can change when appropriate
- provide some form of carer's leave (this may be unpaid, but could be useful in urgent situations).

Chapter 39

Disability and Rights for the patients

Introduction:

Disability is an umbrella term, covering impairments, activity limitations, and participation restrictions. An impairment is a problem in body function or structure; an activity limitation is a difficulty encountered by an individual in executing a task or action; while a participation restriction is a problem experienced by an individual in involvement in life situations. International Classification of Functioning, Disability and Health (ICF Model) is the WHO framework for measuring health and disability at both individual and population levels. ICF was officially endorsed by all 191 WHO Member States in the Fifty- fourth World Health Assembly on 22 May 2001. It provides a unified and standard language and a framework for the description of health and health-related states.

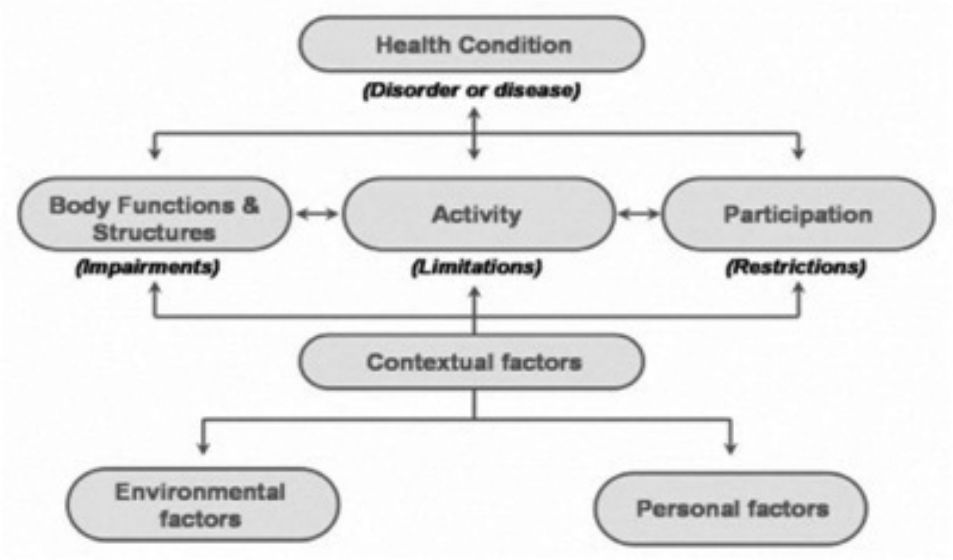

Fig. 39.1 ICF Model

Generally the chronic progressive degenerative neurological disorders are incurable. Family members of these persons are the secondary sufferers.

The impact of the disability on the family members or society of persons suffering from such disorders is not given the due importance. Economic burden due to these chronic neurological disorders on the society is gaining more attention. the likelihood of the

person with progressive degenerative disorders to go back to their previous job is very minimal. So, specifying the precise deficit for the job requirement of the person with disability is very essential.

For example a person suffering from pure bulbar weakness may have considerable disability as a receptionist but may not have much disability with an office job involving typing or other hand functions. The Disability Division in the Ministry of Social Justice & Empowerment facilitates empowerment of the persons with disabilities,

The next part of this chapter will provide the information about different legislative acts and policies implemented in india for persons with ALS / MND and other chronic neurological disorders

Legislative Acts in India for person with Disabilities :

1. The Person's with Disabilities (Equal Opportunities, Protection of Rights and Full Participation) Act 1995.

Function of the act -

a) develop a national policy to address issues faced by individuals with disability

b) Take steps to ensure barrier free environment in public places work places utilities schools etc.

c) monitor and evaluate the impact of policies program designed for achieving equality full participation of with disabilities

d) design schemes, projects, national plan for the individuals with disability

This act extends to the whole of India except the State of Jammu and Kashmir.

2. The right of persons with disabilities Bill, 2014

- The Bill replaces the Persons with Disabilities (Equal Opportunities, Protection of Rights and Full Participation) Act, 1995. Instead of seven disabilities specified in the Act, the Bill covers 19 conditions.
- For those who have 40% of a disability are entitled for educational benefits, employment, preference in government schemes, etc.
- The Bill confers several rights and entitlements to disabled persons. These include disabled friendly access to all public buildings, hospitals, modes of transport, polling stations, etc.
- Violation of any provision of the Act is punishable with imprisonment up to six months, and/or fine of Rs 10,000. Subsequent violations carry a higher penalty.

3. The National Policy for Persons with Disability, announced in February 2006

It provodes a framework for ensuring a life with dignity for the persons with disabilities and their caregivers.

Following things are included in this policy :

- Extending rehabilitation services to rural areas
- Increasing trained personnel to meet needs
- Emphasizing education and training
- Increasing employment opportunities
- Improving access to public services
- Encouraging state governments to develop a comprehensive social security policy
- Ensuring equal opportunities in sports, recreation and cultural activities
- Increasing the role of civil society organizations as service-providers to persons with disability and their families.

4. Guidelines and Space Standards for Barrier free built environment for disabled elderly persons
    - The aim is to provide an environment that enhances independent functioning for disabled individuals so that they can perform activities .
    - Barrier free buildings / places / transportation systems for public use will be made.
    - Central Public Works Department under the Ministry of Urban Development has issued guidelines in this regard.
    - The therapist plays an important role as one of the member for disability evaluation and for rehabilitation
    - It is mandatory on the part of the therapist to have expertise and skills in clinical evaluation along with knowledge of rules, legislative laws and policies for individuals with disabilities

Use of Disability Certification:

The disa bility certificat ion helps in dividua l with disabilit y to get followi ng facili ties

- reservation of jobs in Government Sector
- travel concession
- loan for entrepreneurship development
- scholarship
- income tax exemption
- age relaxation in employment

Welfare Schemes for Physically Challenged Persons:

The schemes implemented by the Government for the welfare of the persons with disabilities:-

- Assistance to Disabled Persons for Purchase/Fitting of Aids and Appliances (ADIP):- Under the scheme, aids/appliances are distributed to the needy persons with disabilities
- The National Handicapped Finance & Development Corporation provides concessional credit to persons with disabilities for setting up income generating activities for self employment
- Scheme for Implementation of Persons with Disabilities (Equal Opportunities, Protection of Rights and Full Participation) Act, 1995 (SIPDA):- Under this Scheme, assistance is provided for setting up of District Disability Rehabilitation Centres, Regional Rehabilitation Centres, creating barrier free environment in public buildings, awareness generation etc.
- Government of I ndia has introduc ed several ot her schemes t o promote employment/self-employment among disabled population - Government provides assistance to Voluntary Organization for training and Sheltered workshops. Ba nk s Provide loans a t low int erest ra t es t o prom ot e Self- employment. Certain Categories of handicapped are allotted public telephone booths and other types of shops such as tea stalls.
- Residential houses are allotted to the handicapped persons who are in Govt. service on a priority basis. The Delhi Development Authority has reserved 5%.of shops, 10% residential plots and 1% flats in each housing scheme for the disabled persons
- Ministry of Welfare provides assistance to disabled persons for purchase and fitting of aids and appliances for their physical rehabilitation in order to increase their capacity to participate in economic activities.
- There are several national bodies that are looking after the training and service programs for the handicapped e.g. All India Institute of Physical Medicine and Rehabilitation, Bombay
- Institution that are importing equipment and apparatus for education and training of the handicapped are exempted from Custom duty including the Braille wrist watches.

References :

http://www.who.int/

http://www.prsindia.org/

Chapter 40

Resources and Organizations for ALS and MND

The aim of including this section is to provide information regarding the centers and organizations working internationally and nationally for individuals with ALS and MND.

International Alliance of ALS/ MND Associations

http://www.alsmndalliance.org

The International Alliance of ALS/MND Associations was founded in 1992 to provide an international community for individual ALS/MND associations from around the world. Our vision is to engage with our members, prospective members and other organisations to share resources globally, advance awareness and support people with ALS/MND worldwide.

Asha Ek Hope Foundation

Asha Ek Hope foundation is a non-profit organization supporting people with motor neuron disease [MND] and their families. We are committed to stand by every step of the journey of people with this devastating disease and their families. We strive to infuse hope and positivity in their life, which will empower them to overcome the illness. This is the first foundation for MND in India. This registered NGO is founded with the help of family and friends of a doctor, who is living with motor neuron disease since 2004.

Mission is the organization is to build hope and positivity in the lives of people with MND and their families; while helping them to cope with confidence and dignity; supporting them to live a complete life and also spread awareness of the disease.

Physical , psychological and social support and rehabilitation services with compassion

to improve the quality of life of people with motor neuron disease are provided by the organization.

It provides people with motor neuron disease continue their daily activities by providing advanced mechanical aids and facilitates access to high quality, consistent and modern technology support services.

The organization arranges social activities for people with motor neuron disease and their families, along with a holistic approach to boost their moral and spiritual health.

The organization raises awareness about motor neuron disease and impart proper knowledge about it in the community and keep them updated about recent advances in treatment of motor neuron disease.

www.ashaekhope.org

Neurogen Brain and Spine Institute and Research Centre

http://www.neurogen.in

Diseases resulting from degenerative changes in the nervous system markedly impact the lives of millions and pose growing public health challenges. The prevention and treatment of these neurodegenerative disorders represents critical goals of medical research today and is the mission of the NeuroGen Brain and Spine Institute. At NeuroGen Brain and Spine Institute we provide a ray of hope to people who have none and thereby increase the quality of life. NeuroGen Brain and Spine Institute is an institution which not just wants to give treatment to the patients but also take into consideration the overall well being of the patient.

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ALS India

http://www.alsindia.org

https://www.facebook.com/ALSinIndia

It is a platform for people across India to connect and help fight Amyotrophic Lateral Sclerosis (ALS) to help find the cause and cure for ALS and help people suffering from ALS in India to have access to information, support and updates.

The mission of ALS India association is to help find the cause and cure for ALS and help people suffering from ALS in India to have access to information, support and updates.

Paraplegic Foundation Sion, Mumbai

The paraplegic foundation was started in 1968 to rehabilitate and provide total health care facilities for parap;egics and severly orthopedically disabled.

The foundation also provides services for the rehabilitation of persons with chronic degenerative neurological disorders.

Address : T1 Old Barrack of LTMG Hospital (Sion Hospital),
Next to VGP Showroom, Sion Mumbai 400 022,
Tel : 24071671 / 24033669 • Mobile : 9223549042

Muscular Dystrophy Association India

V. J. Clinic, New No. 6 (Old No. 21), 4th cross Road,
Sastri Nagar, Adyar, Chennai 6000020, Tamilnadu, India
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Centers Providing Assistive Devices and Technology

Innovative Vastunirman Pvt. Ltd.

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Phone : +91-80-26492222 / 26597337 / 26491386 - Ext-9 (Reception)
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Email : e-mail@mobility-india.org

Relief Orthotics

Mr. Kishore Kumar P. P. (Proprietor) No. 83, 10th Cross, 20th Main BTM,
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+(91)-9945666515, +(91)-8197832353
Phone: +(91)-(80)-26781490

HARYANA (Ambala)

Rajas Enterprises (India) Rajas Enterprises (India) Mr. Sanjay Sharma
Nanhera Chowk, Nanhera, Post Office Kuldeep Nagar, Ambala-133 004,(India)
E-mail : rajasenterprises@hotmail.com, rajasenterprises@rediffmail.com
Phone : +(91)-(171)-2611337/2610397 • Fax : +(91)-(171)-2610404

ATICO Medical Pvt. Ltd

Mr. Varun Chopra (General Manager) ATICO House, 5309, Grain Market,
Near B. D. Senior Secondary School Ambala - 133001, Haryana, India
Call Us: 08376808154 • Mobile: +(91)-9896793832, +(91)-9034949132
Phone: +(91)-(171)-4004736
Email: aticomedical@gmail.com , info@aticomedical.com , exports@aticomedical.com

UTTAR PRADESH (Kanpur)

Artificial Limbs Manufac uring Corporation of India

G.T. Road, Kanpur - 209217
Phone: 91-512-2770172, 2770897, 2770817
Fax: 91-512-2770137, 2770870, 2770172 •Toll Free No. 1800-180-5129
Web : http://www.artlimbs.com • Email: alimco_hq@vsnl.net

Technical Automotives

8/3 Old Ganga Bridge, Unnao 209 001
Uttar Pradesh, Or, Post Box No. 460, Kanpur 208 001
Phone: (0512) 2825158, 2825539

GUJARAT (Surat)

Evolution Health Care Private Limited

Mr. Dillip Kumar Sahoo (Managing Director) Plot No. 51, Diwali Baug Society,
In Front Of Suman School, Near Diwali Bag Circle, Opposite Rusabh Tower,
Adajan, Surat - 395009, Gujarat, India
Call Us:09953353389 • Mobile: +(91)-9321112216
Phone: +(91)-(261)-2768132 • Fax : +(91)-(261)-2768132

Navchetan Orthopaedic Appliances (Jamnagar)

HEAD OFFICE : Opp. S.T. DEPOT, JAMNAGAR - 361 005.
GUJARAT (INDIA)
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E-mail : info@navchetan.com

Bonny Orthotech Pvt Ltd (Ahmedabad)
Bonny Orthotech Private Limited Ellisbridge,
Near V.S.hospital,
Ahmedabad Gujarat 380006
Phone : +91 9825034936, 079 26577446, 079 26577497

MADHYA PRADESH (Indore)

Dinesh Industrial Corporation (Indore)
Mr. Nain Batra / Dinesh Batra (Proprietor) No. 584, 585 & 586, Bhagirathpura
Indore - 452003, Madhya Pradesh, India
Call Us:09953355110 • Mobile: +(91)-9302103772
+(91)-9826014409, +(91)-9826022004
Phone: +(91)-(731)-2423404, +(91)-(731)-2420321, +(91)-(731)-2422260
Fax: +(91)-(11)-43852260
Email: navin@activeforall.co.in dineshind@hotmail.com

WHEELCHAIRS INDIA (Indore)

584/3, Bhagirath Pura Near Sai Baba Mandir, Bapu Market
Indore - 452001, Madhya Pradesh, India
Phone : +(91)-(731)-2424083 • Website: http://www.wheelchairindia.com

Handicapped Products (India) Pvt Ltd (Indore)

Handicapped Products (India) Pvt. Ltd.
FH- 392, Scheme No. 54, Indore (M.P.), India
Phone : . 91-731-2571845, 5004377 • Fax: 91-731-2422260
Email: dineshind@hotmail.com

Saify Traders Madhya (Indore)

Mr. Juzer Saify (Proprietor)
Akabar Villa, 14/1, RNT Marg, Street No. 1,
Behind Dawa Bazar, Indore - 452001, Madhya Pradesh, India
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Phone : +(91)-(731)-2704152 , +(91)-(731)-2704481 • Fax: +(91)-(731)-4042439

Barod Basic Appliances & Research Center (Indore)

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Main Road Indore - 452010, Madhya Pradesh, India Call Us: 09953353369
Mobile: +(91)-9826219869, +(91)-9425056486 • Phone : +(91)-(731)-4030397
Fax: +(91)-(731)-2554507

RAJASTHAN

International Fabrication (Jaipur) Mr. Mitangshu Dutta /
Mr. Mayur Dutta (Proprietor)
B- 79, Ramnagarriya Scheme, Behind SKIT College, Jagatpura, Jaipur - 302017,
Rajasthan, India Call Us: 08586968462
Mobile: +(91)-7891699462 , 9529899118

Shrinath Hitech Prosthetic & Orthotic Centre (Udaypur)

Mr. Manas Ranjan Sahoo, No. 4/AV-23, RHB Colony, Goverdhan Villas,
Behind Saras Dairy Udaipur - 313 001, Rajasthan , India
Mobile: +(91)-9828056345

Nisha Foot Centre (Jaipur)

Mr. Vinod Kumar Sharma (Director) A-34, Vijay Nagar,
1st Kartarpura, Jaipur - 302 006, Rajasthan, India Call Us: 09953352359
Mobile: +(91)-9414042603, +(91)-9460000153
Phone: +(91)-(141)-2500009 • Fax: +(91)-(141)-2500009

WEST BENGAL

Karma Healthcare Ltd. (Kollkata)

GD-347, Salt Lake, Sector-III, Kolkata 700106, India
Phone : +91-033-6450-7062
Email : service@karmahealthcare.com www.karmahealthcare.com

Nevedac Prosthetic Centre (Chandigarh) Company

Website: http://www.nevedacprostheticcentre.com/
Contact Address: 104, sector 11-A, Chandigarh - 160011, India

Nautiyal Artificial Limb Center (Uttarakhand -Dehradun)

Mr. Vijay Kumar Nautiyal Shastri Nagar, Haridwar Road,
Near Vidanshabha Chowck, Dehradun - 248 001,Uttarakhand, India
Mobile: +(91)-9837224144 • Phone : +(91)-(135)-2669425

International Associations working for people with MND China

BEIJING ORIENTAL RAIN ALS CARE CENTER (ORACC)
http://www.jiandongren.org

SHAANXI ALS ASSOCIATION
http://www.sxals.cn

HONG KONG NEURO-MUSCULAR DISEASE ASSOCIATION
http://www.hknmda.org.hk/index.php

JAPAN ALS ASSOCIATION (JALSA)
http://www.alsjapan.org

KOREAN ALS ASSOCIATION
MND MALAYSIA (PERSATUAN PENYAKIT MOTOR NEURON MALAYSIA)
http://www.mnd.org.my

MONGOLIAN ALS ASSOCIATION

TAIWAN MOTOR NEURON DISEASE ASSOCIATION
http://www.mnda.org.tw

North America

ALS HOPE FOUNDATION
http://www.alshopefoundation.org

ALS THERAPY DEVELOPMENT INSTITUTE
http://www.als.net

FORBES NORRIS MDA/ALS RESEARCH CENTER
http://www.cpmc.org/services/als

HOPE LOVES COMPANY
http://www.hopelovescompany.com

KICK ALS!
http://www.kickals.org

LES TURNER ALS FOUNDATION
http://www.lesturnerals.org

PRIZE4LIFE, INC, USA
http://www.prize4life.org

TEAM GLEASON
http://teamgleason.org

THE ALS ASSOCIATION
http://www.alsa.org

27001 Agoura Road, Suit 150
Calabasas Hill , CA 91301 - 5104 http://www.alsa.org

The Muscular Dystrophy Association

3300, East Sunrise Drive
Tucson, AZ 85718-3208
http://www.mdausa.org

SOUTH AMERICA

ASOCIACIÓN COLOMBIANA DE ESCLEROSIS LATERAL AMIOTRÓFICA
http://www.acelaweb.org

ASOCIACIÓN DISTROFIA MUSCULAR
http://www.adm.org.ar

ASOCIACIÓN ELA ARGENTINA (ALS ASSOCIATION OF ARGENTINA)
http://www.asociacionela.org.ar

ASSOCIAÇÃO BRASILEIRA DE ESCLEROSE LATERAL AMIOTRÓFICA (ABRELA)
http://www.abrela.com.br

ASSOCIAÇÃO PRÓ-CURA DA ELA (ALS PRO-CURA ASSOCIATION)
http://procuradaela.org.br

INSTITUTO PAULO GONTIJO
http://www.ipg.org.br

CANADA and CENTRAL AMERICA

ALS CANADA
http://www.als.ca

CUBAN NEUROLOGY AND NEUROSURGERY INSTITUTE

UNITED KINGDOM

MND SCOTLAND
http://www.mndscotland.org.uk

MOTOR NEURONE DISEASE ASSOCIATION
http://www.mndassociation.org

ALS FOUNDATION OF THE NEDERLANDS (STICHTING ALS NEDERLAND)
http://www.als.nl

ALS LIGA BELGIË (THE ALS LEAGUE OF BELGIUM)
http://www.ALSLIGA.be

ALS SCHWEIZ (SWITZERLAND)
http://www.als-schweiz.ch

ALS-MNH DERNEGI (ALS-MND ASSOCIATION) (TURKEY)
http://www.als.org.tr

ASOCIACIÓN ELA/EMN (SPAIN)
http://elaemn.es

ASOCIACIÓN ESPAÑOLA DE ESCLEROSIS LATERAL AMIOTRÓFICA (ADELA) (SPAIN)
http://www.adelaweb.com

ASOCIATIA HANDICAPATILOR NEUROMOTOR DIN ROMÂNIA (ROMANIA)
http://www.ahnr.ro/

ASSOCIATION OF MUSCLE DISORDERS (TURKEY)
http://www.kashastaliklari.org.tr

ASSOCIATION POUR LA RECHERCHE SUR LA SCLÉROSE LATÉRALE AMYOTROPHIQUE (ARSLA)
http://www.arsla.org

ASSOCIATION SUISSE ROMANDE ET ITALIENNE CONTRE LES MYOPATHIES (ASRIM) (SWITZERLAND)
http://www.asrim.ch

ASSOCIAZIONE ITALIANA SCLEROSI LATERALE AMIOTROFIA (AISLA ONLUS) (ITALY)
http://www.aisla.it

CYPRUS INSTITUTE OF NEUROLOGY AND GENETICS (CRYPTUS)
http://www.cing.ac.cy

DEUTSCHE GESELLSCHAFT FUR MUSKELKRANKE E.V. (GERMANY)
http://www.dgm.org

DIGNITAS DOLENTIUM (POLAND)
http://www.mnd.pl

DRUSTVO DISTROFIKOV SLOVENIJE (MUSCULAR DYSTROPHY ASSOCIATION OF SLOVENIA) Slovenia
http://drustvo-distrofikov.si

FUNDACION ESPANOLA PARA EL FOMENTO DE LA INVESTIGACION DE LA ESCLEROSIS LATERAL AMIOTROFICA (FUNDELA)
http://www.fundela.info

IRISH MOTOR NEURONE DISEASE ASSOCIATION
http://www.imnda.ie

MARTHA-MARY MEDICAL CENTER "MILOSERDIE"
http://www.mndfund.ru

MND ASSOCIATION OF ICELAND
http://www.mnd.is

MIDDLE EAST

ALS-MNH DERNEGI (ALS-MND ASSOCIATION) (TURKEY)
http://www.als.org.tr

ASSOCIATION OF MUSCLE DISORDERS (TURKEY)
http://www.kashastaliklari.org.tr

ISRALS (THE ISRAELI ALS ASSOCIATION) (ISRAEL)
http://www.israls.org.il

STICHTING ALS IRAN-NEDERLAND (SAIN) (IRAN)
http://www.alsiran.com | http://alsiran.blogspot.nl

AFRICA

MOTOR NEURONE DISEASE ASSOCIATION OF SOUTH AFRICA
http://www.mnda.org.za

AUSTRALIA and NEW ZEALAND

MOTOR NEURONE DISEASE AUSTRALIA
http://www.mndaust.asn.au | http://www.mndcare.net.au

MOTOR NEURONE DISEASE ASSOCIATION OF NZ
http://mnda.org.nz

Chapter 41

Frequently asked questions

1. What is MND?

Motor neurone disease (MND) is the name given to a group of diseases in which the nerve cells - neurones - controlling the muscles that enable us to move around, speak, breathe and swallow, fail to work normally. With no nerves to activate them, muscles gradually weaken and waste. The patterns of weakness vary from person to person.

2. How many people have ALS in world?

In world there is estimated at around 450,000. These numbers are hard to articulate accurately because there is no standard measure; the disease takes time to diagnose. General someone is diagnosed with ALS every 90 minutes. The life-time incident rate for an average person is often estimated at between 1 in 400 to 1 in 600 people.

3. What are reasons to gets ALS and why?

Only 5- 10% of all cases are of family history of ALS. There are no clear cause known for remaining 90% of cases (known as sporadic ALS). ALS seems to affect males and females at the same rate, and there is no difference based on ethnicity or other demographic variables. Familial ALS and sporadic ALS are clinically similar from one another.

4. Is there a connection to military service?

Harvard School of Public Health broadened the case for ALS's military relevance. Its epidemiological study found that men with a history of any military service in the last century were nearly 60% more likely to die of ALS than men in the general population.

5. How to diagnosis of ALS?

Diagnosis requires an expert neurological opinion and can be assisted through a range of tests, including some which eliminate other conditions. Often an Electromyograph (EMG) is used, in which a needle is inserted into various muscles to measure their electrical activity.

6. What are the symptoms of MND?

Early symptoms are mild, and include stumbling due to weakness of the leg muscles, difficulty holding objects due to weakness of the hand muscles, slurring of speech or swallowing difficulties due to weakness of the tongue and throat muscles. In some instances people experience changes in their behaviour.

7. What is life expectancy after ALS?

Progression varies and up to 10% are estimated to survive 10 years or longer. For some cases its 3 to 5 years from onset of symptoms. Survival can also be extended if a patient opts for mechanical ventilation. However initial symptoms, rate and pattern of progression, and survival time after diagnosis - vary significantly from person to person. Many of the patients with MND survive longer than 10 years and some may survive upto a natural life span with advanced life support equipment. Stephen hawking is an expiring example of how wrong the estimates of survival are in MND.

8. What is the treatment for MND?

There is no cure for motor neurone disease (MND), but a medication has been approved for the treatment of amyotrophic lateral sclerosis (ALS) - the most common form of MND.Riluzole, the first effective treatment for people with ALS.

It may:

- Slow disease progression
- Keep people in the milder stages of disease for longer, thus improves the quality of life.
- People started on riluzole soon after diagnosis show the greatest benefits.

9. What remains intact in ALS?

In most instances the intellect and memory are not affected. The bowels and bladder are not affected by the disease, although diet and exercise should be carefully monitored. In the PLS (Primary Lateral Sclerosis) form of MND some people experience urinary urgency.

10. Where to get support to ALS/ MND patients?

Patient can get support from family, friends also from general practitioners, neurologists, occupational therapists, physiotherapists, speech pathologists, psychologists, dietitians, home care nurses, social workers and MND Associations in each state and territory. MND Associations can provide information about resources, equipment, psychological support, and counseling to patient and family members.

11. Is there a relation to Primary Lateral Sclerosis (PLS) or other similar diseases?

PLS neurodegeneration is restricted to only upper motor neuron, while ALS diagnosis requires loss of both upper and lower motor neurons.

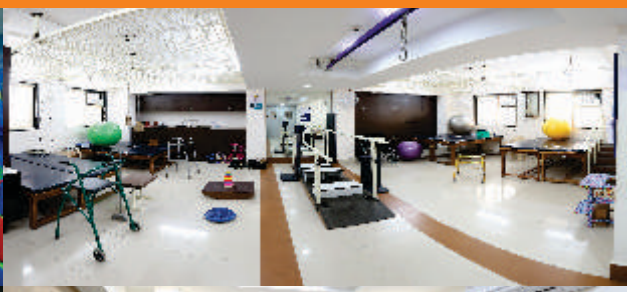
12. Is there a treatment for ALS?

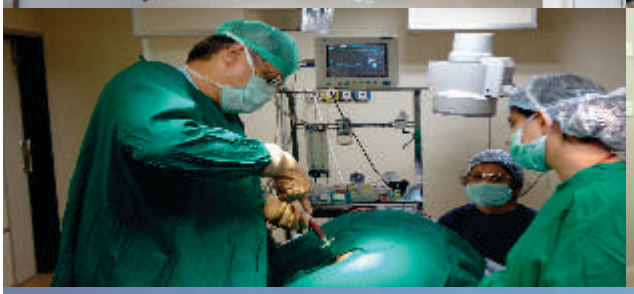
Currently, there is no effective therapeutic approved for use by the FDA that has been identified to stop the disease's progression or prevent onset. Rilutek is a drug that has been approved for use in the treatment of ALS.

13. Can I have children after being detected with MND?

Yes both men and women can have children even after the diagnosis. Women if pregnant must take care as the weakness may progress during pregnancy and child birth. The doctors may advice you to undergo a C-section delivery.

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 3500 patients from 36 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 45 scientific papers in international and national journals. 12 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 9001:2008, 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.

Asha Ek Hope
Foundation for MND/ALS

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Dadar (W), Mumbai- 400028

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