Motor Neuron Disease: Early Diagnosis and Multidisciplinary Management Strategies

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Objectives

1. Define Motor Neuron Disease (MND).
2. Describe the types and causes of MND.
3. Explain the pathophysiology of MND.
4. Identify the common clinical features and symptoms.
5. Discuss the diagnostic investigations used in MND.

Objectives(Cont)

6. Explain the principles of management and treatment.
7. Complications and prognosis of MND.
8. Understand the importance of multidisciplinary and palliative care.

Definition

- Motor neuron disease is a progressive neurodegenerative disorder affecting upper and lower motor neurons.
- Initially neuronopathy followed by axonopathy

Epidemiology

- Annual incidence: about **1–3 cases per 100,000 population**
- Prevalence: approximately **5–8 cases per 100,000 population**
- Usually affects adults between 40–70 years.
- More common in males.
- Amyotrophic lateral sclerosis (ALS) is the most common type.

Terminology

- Motor neuron disease/ALS
- Motor neuron diseases
- Motor neuron disorder- A broader term that refer to any disorder involving motor neurons. It may be degenerative or secondary.

Classification of Motor neuron diseases

- Amyotrophic Lateral Sclerosis (ALS)/MND
- Primary Lateral Sclerosis (PLS)
- Progressive Muscular Atrophy (PMA)
- Progressive Bulbar Palsy (PBP)

Classification of ALS

1. Sporadic or acquired ALS
2. Familial ALS
3. FTD-ALS overlap

Classification of ALS According to Onset

- Classic (spinal-onset)
- Mills hemiplegic variant
- Pseudoneuritic presentation
- Monomelic presentation
- UMN onset
- LMN onset
- Bulbar onset

Etiology and Risk Factors

- Etiology – Idiopathic/ Genetic
- Mostly sporadic- (90-95)%
- Genetic- (5-10)%
- Oxidative stress and glutamate excitotoxicity
- Environmental factors may contribute

Pathophysiology

- Degeneration of upper and lower motor neurons
- Loss of muscle innervation
- Progressive muscle weakness and atrophy

Clinical Features

- Muscle weakness
- Muscle wasting and fasciculations
- Spasticity and hyperreflexia
- Dysarthria and dysphagia in bulbar involvement

Upper Motor Neuron Signs

- Increased muscle tone/Spasticity
- Hyperreflexia
- Extensor plantar response

Lower Motor Neuron Signs

- Muscle atrophy
- Fasciculations
- Hypotonia
- Reduced reflexes

There are certain NO's in MND

- No sensory involvement
- No autonomic/ sphincter involvement
- No cognitive impairment
- No ocular involvement
- No cerebellar/ extra pyramidal involvement

Diagnosis

- Clinical History and examination
- Nerve conduction studies
- Electromyography (EMG)
- MRI to exclude other structural causes
- Investigation to exclude others D/D

Diagnostic Criteria

Gold Coast criteria

- Either (1) upper and lower motor findings in one body region; or (2) lower motor findings in two body regions – without an alternative demonstrable cause.

Body regions

- Bulbar
- Cervical
- Thoracic
- Lumbosacral

Differential Diagnosis

- Cervical myeloradiculopathy
- Myeloneuropathy
- Multiple sclerosis
- Paraneoplastic Motor neuron syndrome

Management

1. Disease-Modifying Treatment

- Riluzole
- Edaravone
- Tofersen(SOD1 gene mutation–associated ALS) Antisense oligonucleotide that reduces abnormal SOD1 protein production.

2. Symptomatic Treatment

TABLE 98.5 Symptomatic Treatment in Amyotrophic Lateral Sclerosis

Symptoms	Pharmacotherapy
Fatigue	Pyridostigmine bromide Antidepressants Methylphenidate Amantadine Modafinil
Spasticity	Baclofen Tizanidine Dantrolene sodium Diazepam
Jaw clenching	Benzodiazepines
Cramps	Quinine sulfate Baclofen Vitamin E Clonazepam
Fasciculations	Carbamazepine
Sialorrhea	Hyoscyamine sulphate Diphenhydramine Scopolamine patch Glycopyrrolate Atropine TCA
Pseudobulbar laughing or crying	TCAs SSRIs L-Dopa/carbidopa Lithium Mirtazapine Venlafaxine Quinidine/dextromethorphan

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Thick phlegm	Guaifenesin Nebulized *N*-acetylcysteine Nebulized saline Propranolol
Aspiration	Cisapride
Joint pains	Antiinflammatory drugs Analgesics
Depression	TCAs SSRIs, venlafaxine, mirtazapine, bupropion
Insomnia	Zolpidem tartrate Lorazepam Opioids TCAs
Laryngospasm	Sublingual lorazepam
Respiratory failure	Bronchodilators Morphine sulfate
Constipation	Increase oral liquid Metamucil Dulcolax suppositories Lactulose and other laxative

Management(cont)

3. Supportive care and rehabilitation

- Physiotherapy
- Occupational Therapy
- Speech therapy
- Nutritional support
- Respiratory support

Nutritional support

- PEG (Percutaneous Endoscopic Gastrostomy)

Respiratory support

Respiratory failure is the leading cause of death in MND.

Non-Invasive Ventilation (NIV)

- BiPAP commonly used
- Improves sleep quality and survival

Indications

- Orthopnea
- Morning headache
- Daytime somnolence
- Reduced vital capacity

4. Multidisciplinary Care

- A multidisciplinary approach improves survival and quality of life.

Multidisciplinary Care

Team Members

- Neurologist
- Physiotherapist
- Speech therapist
- Respiratory physician
- Dietitian
- Occupational therapist
- Psychologist
- Palliative care specialist

Complications

- Respiratory failure
- Aspiration pneumonia
- Malnutrition
- Psychological distress

Prognosis

- Median survival in Spinal onset ALS: 3–5 years after diagnosis
- Bulbar onset ALS: 6 month to 1 year
- PMA: 10-15 years
- PLS: 15-20 years

Conclusion

- MND/ ALS is a serious progressive neurological disorder.
- Early diagnosis and multidisciplinary care improve quality of life survival.

Thank You