Features of Motor Neuron lesions.

	UMN	LMN
Inspection.	Normal (May be Wasting due to disuse (chronic)	Muscle Wasting Fasciculation.
Tone.	- Increased with clonus.	- Normal or Decreased - No clonus.
Weakness:	Affects extensor in arms, Flexors in legs. Hemiparesis, paraparesis. Tetraparesis.	- Typically Focal, in the distribution of nerve root or peripheral nerve with associated sensory changes.
Deep tendon Reflexes.	Increased.	Decreased / Absent.
Plantar response.	Extensor (Babinski sign)	Flexor.

Motor Neuron Diseases

Primary lateral sclerosis → UMN.

Progressive muscular atrophy → LMN

Progressive Bulbar palsy → Cranial nerves.

Amyotrophic lateral sclerosis → UMN & LMN Mixed.

Amyotrophic Lateral Sclerosis

- Most Frequent MND, also used interchangeably.
- It is neurodegenerative disease, caused by degeneration of the primary motor cortex, brainstem and spinal cord.

Cause :- Genetic → SOD gene mutation.

Clinical Features:

Symptom.

Usually starts as limb onset (Foot drop) or with bulbar symptoms (Dysarthria, Swallowing difficulty).

- Usually ends with respiratory Failure II-

- Cognitive and Behavioural symptoms are common (Fronto temporal dementia).
- Sensory, autonomic and visual impairment does not occur but cramps are very common.

Signs:

Examination reveals combination of UMN and LMN defects.

Inspection → Wasting and Fasciculation of muscles.

Weakness → Limbs, face tongue & Palate.

Tone → Spasticity.

→ Exaggerated reflexes

→ and Extensor responses.

Investigations

- Conduction Nerve Test.
- Electromyography.
- Genetic Testing.
- CT/MRI.

Treatment → Multidisciplinary.

- Speech therapy, Cognitive, occupational.
  Feeding therapy. Ventilatory.

Primary lateral sclerosis

- Predominantly UMN.
- Spasticity and few lower motor neuron sign.
- Gradually progressing.

Progressive Muscular atrophy

- Predominantly LMN signs & symptoms.

Progressive Bulbar Palsy → LMN of Cranial

- Predominantly Cranial Nerves.
- Involvement of Tongue, Palate and
  Pharyngeal muscles.

→ Dysarthria, Dysphagia,

- Wasting, Fasciculation of Tounge muscles
- Reflexes are normal or absent.
- No emotional lability.

Spinal muscular atrophy

Genetic affection of spinal cord and cranial lower motor neurons.