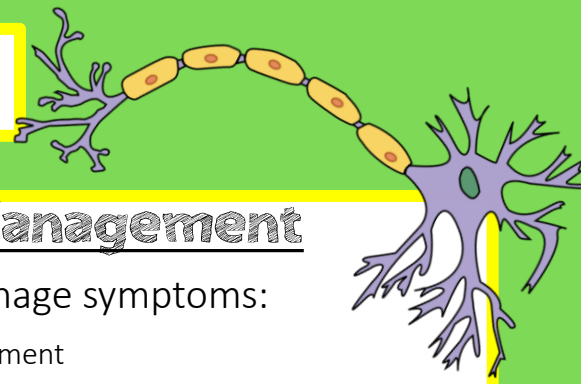
MOTOR NEURONE DISEASE



Progressive disease characterised by **degeneration** of the motor neurons with **cortical, brainstem, and ventral cord** locations

Epidemiology

- Incidence: 1.5-2/100,000
- Mean onset 57 years
- M>F
- Median survival 2-3 years

Pathogenesis

- 10% familial
- Can be UMN/ LMN predominant
- Cause unknown
- Likely combination of **environmental and genetic factors**

	UMN	LMN
Strength	↓	↓
Tone	↑ Spasticity	↓ Hypotonia
Deep tendon reflexes	↑ Brisk	↓ Diminished/ absent
Plantar reflex	Upgoing toes	Down-going toes
Atrophy	No	Yes
Fasciculations	No	Yes

Clinical presentation

Symptoms/ signs **depend on region** involved

Typical presentations:

- *Pseudobulbar palsy w. minimal limb involvement*
- **ALS-** *mixed UMN and LMN signs in limbs, usually starts asymmetrically*

Revised El Escorial Criteria to diagnose

- Need clinical evidence of UMN and widespread LMN dysfunction

Progression is **variable**

Diagnosis of exclusion

- ***Investigations:*** *Conduction studies, MRI of CNS, routine bloods, genetic tests*

Management

No cure, manage symptoms:

- Pain management
- Antimuscarinics to reduce drooling
- Anti-depressants

Riluzole: inhibits glutamate release, slight increase in survival

MDT approach

- Nutrition
- Palliative care
- Speech and language therapy
- Dieticians
- Respiratory care:
  - *Respiratory muscle weakness may accompany bulbar symptoms*
  - *Non-invasive ventilation at night*
  - *Death is usually due to respiratory failure*

Patient and carer **support groups** and **education**

- Advanced directives