

# Neurological Diseases and Palliative Care



LEARNING TOGETHER STUDY DAY

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# Dr Zoe Helman

## Motor Neurone Disease – Symptoms, diagnosis and possible treatments

# Motor Neurone Disease

SYMPTOMS, DIAGNOSIS AND POSSIBLE TREATMENTS

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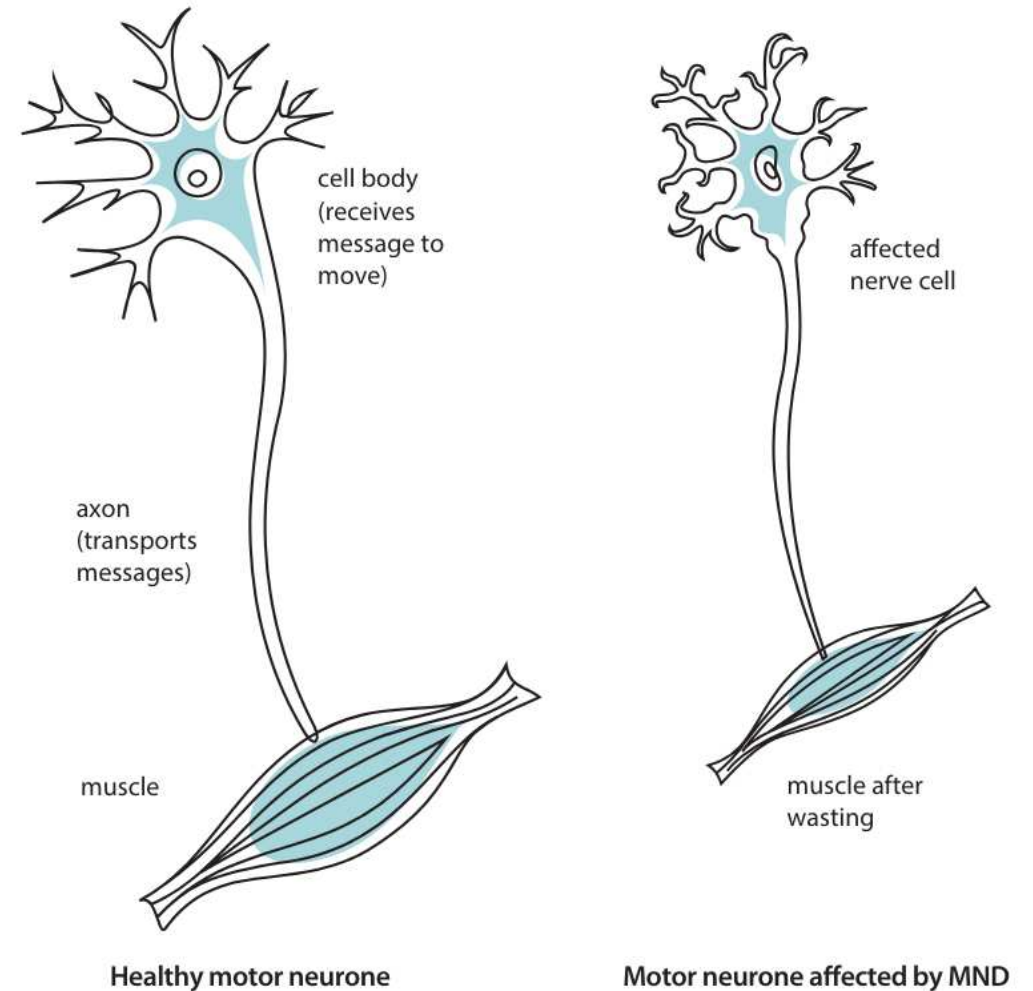
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# Motor Neurone Disease

- Progressive neurodegenerative condition
- Relatively rare
  - Approx 5,000 in the UK
  - 1 in 300 risk across lifetime
  - Six deaths a day in UK
- No cure
- Rarely inherited





# Types of MND

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- Amyotrophic lateral sclerosis (ALS)
- Bulbar onset MND or Progressive bulbar palsy (PBP)
- Progressive muscular atrophy (PMA)
- Primary lateral sclerosis (PLS)

# Symptoms

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- Symptoms are very variable
- Progress at different rates
- Mimic other conditions
  - Most symptoms that are like MND are something else!

# Symptoms and Signs

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Symptoms determined by which nerves affected

- Bulbar
- Non-Bulbar
- Cognitive
- General- Fatigue

# Bulbar

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- Speech
- Swallowing
- Saliva
- Breathing
- Coughing





# Non bulbar

- Muscle weakness
- Muscle stiffness
- Muscle cramps
- Muscle twitching/fasciculations





# Cognitive

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- Emotional lability
- Difficulties with decision making , planning and language
  - Estimates vary 35-50%
  - 15% form of dementia
- The Edinburgh Cognitive and Behavioral ALS Screen (ECAS) can be used

# What is not affected

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- Generally, senses e.g. taste and smell not affected
- Sensory nerves and ability to feel pain
- Bladder and bowel control
  - But other muscle weakness can cause issues

# Symptom Progression

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## Huge spectrum of rates of progression

- Some over months
- Some over many years



@limpbroozkit



@lucylintott1



# Diagnosis

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- Can take time as symptoms can be vague
  - Often only progression raises suspicion
- Allied Health Professionals e.g. Physiotherapy and SALT may be first to raise possibility of a more sinister cause for symptoms
- Need to exclude other causes before final diagnosis

# Differential Diagnoses

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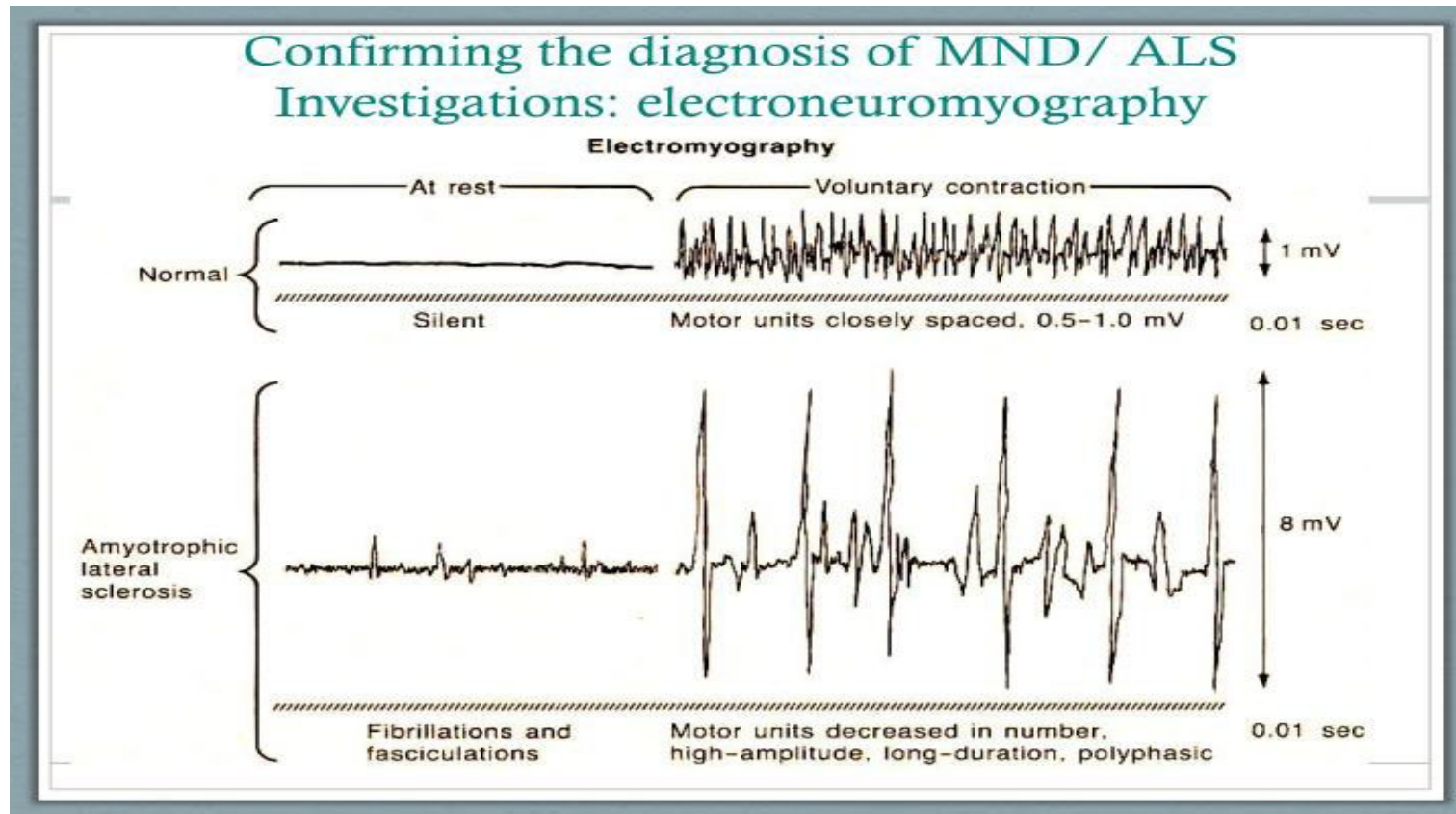
- Other progressive neurological conditions
- Brain tumours
- Nerve injuries/radiculopathy/myelopathy
  - Eg spinal disc disease
- Myositis
- Chronic Inflammatory Demyelinating polyneuropathy
- Vascular disease
- Arthritic disease
- Muscle diseases

# Diagnosis

- History and examination
- Bloods
- Electromyography-EMG
- MRI
- Nerve conduction tests
- Transcranial magnetic stimulation
- Lumbar puncture



# Electromyography- EMG



# Treatments

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## Supportive

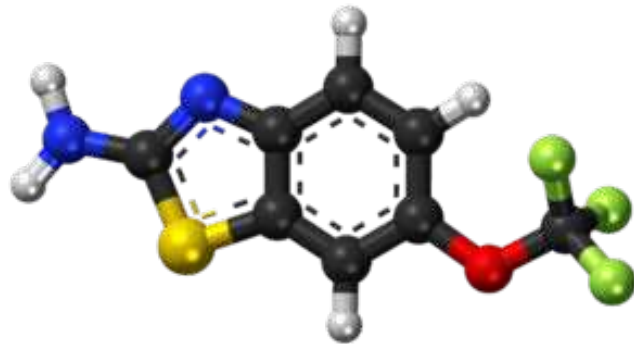
- Treatment for symptoms
- Treatment for complications eg with breathing and swallowing

## Disease modifying

- Maybe

# Riluzole

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- Only approved treatment in the UK
- Increases survival
- Ten percent have significant GI side effects

# Edavarone

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- Not licenced in the UK
- Used in USA and Japan
- IV and Oral
- Mechanism of action – thought to reduce oxidative stress



# Research

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- Genetic Markers/targeted therapy
- Stem cell research
- Other drugs
  - Masitinib-Tyrosine Kinase inhibitor
  - Arimoclomol-Heat shock protein amplifier
- Drugs targeting neuroinflammation
- Drugs promoting neuroprotection
  - Antioxidant and drugs enhancing cellular repair

# References

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- <https://www.mndassociation.org/sites/default/files/2023-02/Living%20with%20MND%20Interactive.pdf>
- <https://www.nice.org.uk/guidance/ng42/chapter/Recommendations>
- Images: Google images
- Motor Neurone Disease Association