

# Painless, progressive weakness. Could this be Motor Neurone Disease?

## 1. Does the patient have one or more of these symptoms?

### Bulbar features

dysarthria

- slurred or quiet speech, often when tired

dysphagia

- liquids and / or solids
- excessive saliva
- choking sensation especially when lying flat
- tongue fasciculations

### Limb features

- focal weakness
- falls/trips – from foot drop
- loss of dexterity
- muscle wasting
- muscle twitching (fasciculations)
- cramps
- no sensory features

### Respiratory features

- hard to explain respiratory symptoms
- shortness of breath on exertion
- excessive daytime sleepiness
- fatigue
- early morning headache
- orthopnoea

### Cognitive features

- behavioural change
- emotional lability (not related to dementia)
- frontotemporal dementia (affects 15%)

## 2. Is there progression?

### Supporting factors ✓

- asymmetrical features
- age (MND can present at any age)
- positive family history of MND or other neurodegenerative disease.

### Not supportive of MND diagnosis ✗

- bladder / bowel involvement
- prominent sensory symptoms
- double vision / ptosis
- improving symptoms.

**If one or more symptoms (section 1) with progression (section 2) are present, consider MND and refer to neurology. If MND is suspected, state this explicitly in the referral letter. Delays often occur due to referral to ENT or orthopaedic services.**

<b>Bulbar features</b>	<b>Limb features</b>
<p><b>25% of patients present with bulbar symptoms.</b></p> <ul style="list-style-type: none"> <li>– dysarthria</li> <li>– quiet, hoarse or altered speech</li> <li>– slurring of speech often when tired</li> <li>– dysphagia, often liquids first then solids, with early sensations of catching in the throat or choking when drinking quickly</li> <li>– excessive saliva</li> <li>– choking sensation when lying flat</li> <li>– weak cough, often not noticed by patient</li> </ul> <p>Painless progressive dysarthria – consider neurological referral rather than ENT.</p>	<p><b>70% of patients present with limb symptoms.</b></p> <ul style="list-style-type: none"> <li>– focal weakness – painless with preserved sensation</li> <li>– distal weakness</li> <li>– falls/trips – from foot drop</li> <li>– loss of dexterity eg problems with zips or buttons</li> <li>– muscle wasting – hands and shoulders Typically asymmetrical</li> <li>– muscle twitching/fasciculations</li> <li>– cramps</li> </ul>
<b>Respiratory features</b>	<b>Cognitive features</b>
<p><b>Respiratory problems are often a late feature of MND and an unusual presenting feature. Patients present with features of neuromuscular respiratory failure.</b></p> <ul style="list-style-type: none"> <li>– shortness of breath on exertion</li> <li>– excessive daytime sleepiness or fatigue</li> <li>– early morning headache. Patients oftendescribe a ‘muzziness’ in the morning, being slow to get going or as if hung over</li> <li>– un-refreshing sleep</li> <li>– orthopnoea</li> <li>– frequent unexplained chest infections</li> <li>– weak cough and sniff</li> <li>– nocturnal restlessness and/or sweating</li> </ul> <p>Consider MND if investigations for breathlessness do not support a pulmonary or cardiac cause.</p>	<p><b>Around 50% are affected by cognitive change. This increases to 20% in the final stage of the disease. Frank dementia at presentation is rare. Cognitive dysfunction is increasingly recognised, as evidenced by:</b></p> <ul style="list-style-type: none"> <li>– behavioural change such as apathy or lack of motivation</li> <li>– difficulty with complex tasks</li> <li>– lack of concentration</li> <li>– emotional lability (not related to dementia).</li> </ul> <p>Ask specifically about a family history of these features.</p>

Additional information and resources available at [mndassociation.org/gp](http://mndassociation.org/gp)

