Painless, progressive weakness
Could this be Motor Neurone Disease?
The importance of early diagnosis

- removal of uncertainty for the person experiencing symptoms
- allowing for care and support to start as early as possible
- enabling the person with MND and their carer to consider and plan for their future
- increasing the window of opportunity to research into, and better understand, the condition
1. Does the patient have one or more of these symptoms?
Limb features
“I’m not in pain but don’t have the strength I used to when doing everyday things – carrying shopping, undoing jars that kind of thing.”
I'm not in pain but don't have the strength I used to when doing everyday things—carrying shopping, undoing jars that kind of thing.

Limb features

- Focal weakness
“It is as if I can’t control my fingers. I’m having difficulty with simple things - unscrewing the petrol cap, doing up zips and buttons.”
It is as if I can't control my fingers. I'm having difficulty with simple things—unscrewing the petrol cap, doing up zips and buttons.

**Limb features**

- Loss of dexterity
“My foot and leg feel heavy and seem to ‘drag’ when I walk – sometimes I fall over because of it.”
My foot and leg feel heavy and seem to 'drag' when I walk – sometimes I fall over because of it.

Limb features

- Falls/trips – from foot drop
“I get cramps in my leg.”
Limb features

- Cramps
70% of patients present with limb symptoms
Bulbar features
“I am finding that I sometimes slur my words – other people are noticing it too.”
Bulbar features

- Dysarthria - slurred or quiet speech often when tired

I am finding that I sometimes slur my words – other people are noticing it too.
“I have difficulty swallowing – it feels like I cannot get my food to go down, almost a choking sensation.”
I have difficulty swallowing – it feels like I cannot get my food to go down, almost a choking sensation.

**Bulbar features**
Swallowing difficulties
- Liquids and/or solids
- Excessive saliva
- Choking sensation especially when lying flat
“My tongue occasionally twitches and ‘flickers’.”
My tongue occasionally twitches and 'flickers'.

Bulbar features
- Tongue fasciculations
25% patients present with bulbar symptoms
Respiratory features
“I feel tired during the day – I’ve not felt like that before. I don’t seem to have any energy.”
I feel tired during the day – I've not felt like that before. I don't seem to have any energy.

Respiratory features
- Excessive daytime sleepiness
- Fatigue
“When I wake up I am still tired and I feel a bit hung-over, my head is ‘muzzy’.”
When I wake up I am still tired and I feel a bit hung-over, my head is 'muzzy'.

Respiratory features
• Early morning headache
“I get quite breathless – for example if I walk fast or run for the bus or train.”
I get quite breathless—for example if I walk fast or run for the bus or train.

Respiratory features
• Shortness of breath on exertion
"When I go to bed and lay down I find it harder to breathe – it is better when I sit upright."
When I go to bed and lay down I find it harder to breathe – it is better when I sit upright.

Respiratory features
• Orthopnoea
Respiratory problems are often a late feature of MND and an unusual presenting feature.

Patients present with features of neuromuscular respiratory failure.
Cognitive features
Frank dementia at presentation is rare
“I find myself crying for no real reason sometimes.”
I find myself crying for no real reason sometimes.

Cognitive features
• Emotional lability
2. Is there progression?
Supporting factors

- Asymmetrical features
- Age – MND can present at any age
- Positive family history of MND or other neurodegenerative disease

Factors NOT supportive of MND diagnosis

- Bladder / bowel involvement
- Prominent sensory symptoms
- Double vision / Ptosis
- Improving symptoms

If the patient has one or more symptoms and there is progression query MND and refer to Neurology.
If you think it might be MND please state explicitly in the referral letter. Common causes of delay are initial referral to ENT or Orthopaedic services.
The ‘Red Flags’ tool
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
  - Swallowing difficulties
  - 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 (core)
  - Behavioural change
  - Emotional lability
    (not related to dementia)
  - Fronto-temporal dementia

2. Is there progression?

- Supporting factors
  - Asymmetrical features
  - Age – MND can present at any age
  - Positive family history of MND or other neurodegenerative disease

- Factors NOT supportive of MND diagnosis
  - Bladder / bowel involvement
  - Prominent sensory symptoms
  - Double vision / Ptosis
  - Improving symptoms

If yes to 1 and 2 query MND and refer to Neurology
If you think it might be MND please state explicitly in the referral letter.
Common causes of delay are initial referral to ENT or Orthopaedic services.
Bulbar features

25% patients present with bulbar symptoms

- Dysarthria
- Quiet, hoarse or altered speech
- Slurring of speech often when tired
- Dysphagia – more often liquids first and later solids. Initially can be sensation of catching in throat or choking when drinking quickly.
- Excessive saliva
- Choking sensation when lying flat
- Weak cough – often not noticed by the patient

Painless progressive dysarthria – consider neurological referral rather than ENT.

Limb features

70% of patients present with limb symptoms

- Focal weakness – painless with preserved sensation
- Distal weakness
  - Falls/trips – from foot drop
  - Loss of dexterity eg problems with zips or buttons
- Muscle wasting – hands and shoulders. Typically asymmetrical
- Muscle twitching/fasciculations
- Cramps

Respiratory features

Respiratory problems are often a late feature of MND and an unusual presenting feature. Patients present with features of neuromuscular respiratory failure

- Shortness of breath on exertion
- Excessive daytime sleepiness
- Fatigue
- Early morning headache. Patients often describe a ‘muzziness’ in the morning, being slow to get going or as if hung over
- Un-refreshing sleep
- Orthopnoea
- Frequent unexplained chest infections
- Weak cough and sniff
- Nocturnal restlessness and/or sweating

Consider MND if investigations for breathlessness do not support a pulmonary or cardiac cause.

Cognitive features

Frank dementia at presentation is rare. Cognitive dysfunction is increasingly recognised, as evidenced by:

- Behavioural change such as apathy or lack of motivation
- Difficulty with complex tasks
- Lack of concentration
- Emotional lability (not related to dementia)

Ask specifically about a family history of these features.
Additional resources:
MND Association downloads and publications at www.mndassociation.org