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 (rare)

- 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.

Additional resources:
MND Association downloads and publications at www.mndassociation.org/gp

Published February 2017

Bulbar features

25% of 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.

Development group for this resource:

RCGP (L Davies, R Pizzaro-Duhart, I Rafi) **MND Association** (J Bedford, H Fairfield)
Neurology (P Callagher, C McDermott, K Morrison, R Orrell, A Radunovic, S Weatherby, A Wills) **Palliative Medicine** (I Baker)

Published February 2017

Monthly headache diary

St George's Healthcare NHS
NHS Trust

Name: DOB:
Month: **Year:**

Date	**Day**	**Time**	**Severity** (1-10)	**Duration** (min / hrs)	**Nausea (N) / Vomiting (V)**	**Painkillers** (Name / Dose)	**Notes** (e.g. triggers, period, changes in preventatives, side effects etc.)
1							
2							
3							
4							
5							
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Diagnosis of tension-type headache, migraine and cluster headache

APPENDIX 3

NHS National Institute for Health and Clinical Excellence

Headache feature	*Tension-type headache*		*Migraine (with or without aura)*		*Cluster headache*	
Pain location[1]	Bilateral		Unilateral or bilateral		Unilateral (around the eye, above the eye and along the side of the head/face)	
Pain quality	Pressing/tightening (non-pulsating)		Pulsating (throbbing or banging in young people aged 12–17 years)		Variable (can be sharp, boring, burning, throbbing or tightening)	
Pain intensity	Mild or moderate		Moderate or severe		Severe or very severe	
Effect on activities	Not aggravated by routine activities of daily living		Aggravated by, or causes avoidance of, routine activities of daily living		Restlessness or agitation	
Other symptoms	None		• Unusual sensitivity to light and/or sound or nausea and/or vomiting. • Aura: symptoms can occur with or without headache and; are fully reversible, develop over at least 5 minutes, last 5 - 60 minutes. Typical aura symptoms include visual symptoms such as flickering lights, spots or lines and/or partial loss of vision; sensory symptoms such as numbness and/or pins and needles; and/or speech disturbance.		On the same side as the headache: • Red and/or watery eye • Nasal congestion and/or runny nose • Swollen eyelid • Forehead and facial sweating • Constricted pupil and/or drooping eyelid	
Duration of headache	30 minutes–continuous		4–72 hours in adults 1–72 hours in young people aged 12–17 years		15–180 minutes	
Frequency of headache	< 15 days per month	≥ 15 days per month for more than 3 months	< 15 days per month	≥ 15 days per month for more than 3 months	1 every other day to 8 per day[3], with remission[4] >1 month	1 every other day to 8 per day[3] with a continuous remission[4] <1 month in a 12-month period
Diagnosis	**Episodic tension-type headache**	**Chronic tension-type headache**[2]	**Episodic migraine (with or without aura)**	**Chronic migraine (with or without aura)**	**Episodic cluster headache**	**Chronic cluster headache**

[1] Headache pain can be felt in the head, face or neck. [2] Chronic migraine and chronic tension-type headache commonly overlap. If there are any features of migraine, diagnose chronic migraine. [3] Frequency of recurrent headaches during a cluster headache bout. [4] The pain-free period between cluster headache bouts.

Adapted from 'Headaches' (NICE clinical guideline 150), available from www.nice.org.uk/CG150

Published February 2017

APPENDIX 4. Hoover's sign, Medication Overuse Headache, and further reading

Hoover's sign (Stone)

Hoover's sign (1908) is the most useful test for functional weakness and the only one that has been found in studies to have good sensitivity and specificity. It is a simple, repeatable test, which does not require skilled or surreptitious observation. The test relies on the principle that we involuntarily extend our hip when flexing our contralateral hip against resistance (you can test this out on yourself):

- voluntary hip extension — ask the patient, lying down, to extend their left hip – i.e. push their heel down onto the bed. In functional weakness there is often weak voluntary extension – the patient "pretends" not to be able to extend the hip
- now test for involuntary hip extension by asking them to flex the right hip against resistance. Keep your hand under the left heel. In functional weakness there will now be normal downward pressure in the left leg – the patient cannot prevent this extension, no matter how hard they try.

Medication Overuse Headache

The expert reviewer of this module felt we should put in more detail about the definition of this condition, which can be hard to diagnose. Diagnostic criteria are:

- headache occurring on 15 or more days per month in a patient with a pre-existing headache disorder
- regular overuse for more than three months of one or more drugs that can be taken for acute and/or symptomatic treatment of headache.

In detail, this means regular intake, for ≥10 days per month for >3 months, of

- ergotamines, triptans, opioids, or combination analgesics, or
- any combination of ergotamines, triptans, simple analgesics, nonsteroidal anti-inflammatory drugs (NSAID) and/or opioids without overuse of any single drug or drug class alone, or
- when the pattern of overuse cannot be reliably established.

It also means regular intake, for ≥15 days per month for >3 months, of simple analgesics (i.e. acetaminophen, aspirin, or NSAID).

Further reading / Resources

Guillan Barre
Guillain Barre and Associated Neuropathies UK
http://www.gaincharity.org.uk/

MND
MND Association / RCGP Motor Neurone Disease : a guide for GPs and primary care teams
http://www.mndassociation.org/wp-content/uploads/PX016-Motor-neurone-disease-a-guide-for-GPs-and-primary-care-teams.pdf

MND Scotland
http://www.mndscotland.org.uk/

Headache
British Association for the Study of Headache Guidelines for All Healthcare Professionals in the Diagnosis and Management of Migraine , Tension-Type headache , Cluster headache and Medication- Overuse Headache (2010)
http://www.bash.org.uk/wp-content/uploads/2012/07/10102-BASH-Guidelines-update-2_v5-1-indd.pdf
(due to be updated Dec 2016)

Headache Pathway for adult patients presenting with headache
http://www.nottsapc.nhs.uk/media/1040/adult-headache-guideline.pdf