

Motor Neurone Disease

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Background

About motor neurone disease (MND)

The motor neuron diseases are a group of progressive neurodegenerative disorders of unknown aetiology characterised by degeneration of the motor neurons. Individuals can have variable degeneration of the central motor neurons within the brain, the anterior horn cells in the spinal cord, and other neurons in the frontal and temporal lobes. This results in different phenotypes of disease which include spasticity, progressive muscle atrophy, and in up to 50% of cases cognitive changes which may include in 30%, frontotemporal dementia. The disease is relentlessly progressive, and depending on the involvement of motor neurons is ultimately fatal in majority of cases within 3 to 5 years from respiratory failure and/or lower respiratory tract infection.

- The average age of onset is 58 to 63 (younger in genetic forms).
- Approximately 10% are due to genetic mutations.
- There is currently no cure.
- Management is largely supportive.

MND service support

MND presents insidiously and can be difficult to diagnose in the early stages. Once diagnosed, patients and their families require a lot of support and rapid access to a wide range of services as their condition can change in a short time.

Clinicians involved with patients with MND:

- work in many areas of the health system (e.g., general practitioners, neurologists, respiratory physicians, gastroenterologists, rehabilitation technologists, dietitians, speech pathologists) but collaborate with each other
- provide a prompt service for the rapidly changing needs of this group of patients

Assessment

1. Look for the combination of upper and lower motor neurone signs, particularly in the same distribution, as the key feature of MND.
 - Less common variants include pure involvement of:
 - lower motor neurons, causing muscle wasting, fasciculation, cramping.
 - upper motor neurons, causing spasticity, hyper-reflexia.
 - Bulbar symptoms including swallowing difficulties, slurred speech, and drooling.
 - **Other symptoms.**

Other symptoms

Including:

- emotional lability.
- depression.
- disinhibition.
- frontotemporal cognitive abnormalities.
- constipation.
- cough.
- pain.
- excessive daytime somnolence.
- shortness of breath.

2. Perform an examination, including:

- Full **neurological examination**.

Neurological examination

This includes:

- *focal muscle wasting*
- *fasciculations*
- *muscle tone*
- *power – the pattern of wasting and weakness is important*
- *reflexes and plantar responses*
- *sensation – this should be normal, particularly in the distribution of the weakness, meaning that the deficit is a pure motor abnormality.*
- *coordination testing is normal within the limits of their weakness.*

- If cognitive impairment is suspected, consider using the [Standardised Mini-Mental State Examination \(SMMSE\)](#).

3. If MND is suspected, do not delay requesting neurology opinion.

- There is no simple "test for MND". This can cause a considerable delay from the time of first symptoms to diagnosis.
- Given the very poor prognosis for most patients, it is vital that the diagnosis be correct and all possible alternatives excluded.

4. Arrange **investigations** to exclude **MND mimics**.

Investigations

- *Creatine kinase – often elevated.*
- *MRI – is key to exclude an extrinsic or intrinsic lesion of the spinal cord, nerve roots and/or nerve head.*
- *Nerve conduction studies – to exclude a neuropathy. Search for nerve entrapment and conduction block.*
- *Electromyography (EMG) – necessary to confirm evidence of acute LMN loss.*

MND mimics

- *Spinal cord and/or nerve root compression secondary to degeneration of the vertebrae and discs in the spine.*
 - *Most common mimic*
 - *Can present with painless muscle wasting, weakness, and fasciculation in one or more limbs*
- *Neuropathies*
- *Myopathies*
- *Spinal muscular atrophy*

Management

1. Establish the diagnosis:
 - Diagnosis may take a year or more to establish.
 - Request an [urgent or routine neurology assessment](#) if MND is suspected, while providing support to the patient and their family.
 - While awaiting a diagnosis, consider requesting physiotherapy and occupational therapy assessments to assist with function.
2. Following diagnosis, provide [patient information](#) about the Motor Neurone Disease Association of Victoria, which can help support the patient and their family or carers.
3. Discuss services and supports with the patient and family or carers:

- **Coordination of care**

Coordination of care

The large number of people and organisations involved can be overwhelming and place additional stress on the patient and family. Ensure they are involved in planning their care and understand who is involved and what they do and why.

- **Equipment and mobility care**

Equipment and mobility

- Most people with MND will need a range of adaptive and mobility equipment to enable them to remain at home as long as possible and to assist in the prevention of pressure injury.
- Changes in mobility can occur quickly and general practice teams need to be prepared to advocate for rapid attention from providers.
- Refer to home occupational therapy assessment providers, who prioritise patients with MND and assess equipment needs. If aged:
 - < 65 years refer via adult occupational therapy assessment, the [MND Association of Victoria](#), or [NDIS](#).
 - ≥ 65 years refer via aged care assessment.

- **Psychological care**

Psychological care

- Referral to a counsellor experienced in dealing with MND is often helpful for patients and their families.
- When emotional lability and/or depression is present, an anti-depressant is often useful. The choice depends on the side-effect profile and tolerability:
 - Mirtazapine is often useful if appetite and/or sleep are poor.
 - Escitalopram is often useful when these other effects are not present.
- Provide written information.
- Arrange for the patient to return for more information or with a support person.

- Identify a health professional to provide ongoing support, e.g. [neurology services](#), the general practice team, palliative care.
- Gently discuss the future with respect to planning ahead, have open discussion with the family regarding preference and wishes.

- **Carer respite**

Carer respite

- Respite can be a difficult option for families as the patient's usual care cannot always be maintained in aged residential facilities.
 - Support for families in planning regular breaks and self care is vital.
 - See [Carer Resources and Support Services](#).
- Offer [Advance Care Planning](#), as some MND patients will eventually lose the ability to communicate.
4. Consider early involvement of palliative care specialists, as the terminal phase can be difficult to predict. Palliative care can provide:
 - Important assistance for symptom management during the middle and end of the disease.
 - In-home care during the final phase of life.
 5. Discuss **cognitive changes** and associated problems (e.g. difficulties with work or finances) with the patient and family or carers.

Cognitive change

- *Up to a third of patients with MND have cognitive deficits without dementia.*
 - *Approximately 20% develop diagnosable dementia (usually frontotemporal lobar dementia), which commonly causes problems with:*
 - *decision making.*
 - *financial planning.*
 - *maintaining personal relationships.*
 - *personality change.*
 - *empathy for loved ones.*
 - *responding appropriately to environmental changes.*
 - *Early discussion of these issues is suggested.*
6. Ensure adequate nutrition.
 - Where bulbar function is impeding swallowing and weight is dropping, request urgent dietitian assessment and speech pathology assessment.
 - The decision to have a percutaneous endoscopic gastrostomy (PEG)/radiologically inserted gastrostomy (RIG) is individual, and many patients choose not to have enteral feeding.
 - If weight is dropping in the absence of bulbar dysfunction, look for underlying depression.
 7. Consider medications:

- **Disease-modifying treatment**

Disease-modifying treatment

Riluzole 50 mg one tablet orally twice a day is the only proven disease modifying medication approved in Australia:

- It is well tolerated, but can affect LFTs and FBE, so monitoring is essential, particularly in the first few months.
- Patients require respiratory function tests with a forced vital capacity (FVC) > 60% to access the medication under the [PBS authority](#) regulations.

- **Medications to manage symptoms**

Medications to manage symptoms

- **Depression/emotional lability**
 - Escitalopram 10 mg orally per day.
 - Mirtazapine 15 to 45 mg orally at night – helpful when insomnia/anorexia are present.
- **Cramps/fasciculation**
 - Magnesium can be trialled first-line.
 - Baclofen – titrate dose up from 5 mg orally per day to 10 mg three times per day.
 - Gabapentin or carbamazepine are alternatives.
- **Pain**
 - *Pain is commonly a complication of immobility.*
 - *Physiotherapy, massage, and stretching can help.*
 - *Aids for comfort and positioning in the car, chair, or bed and to assist with daily living and conserve energy.*
 - *Paracetamol and NSAIDS/COXI are first-line.*
 - *Local injections for local bursitis/capsulitis.*
 - *Buprenorphine patch/Targin often second-line.*
- **Hypersalivation**
 - *Thick sticky secretions can be helped by increasing hydration. Medication which dries the mouth may reduce runny saliva and drooling but may make throat secretions dry and difficult to clear due to reduced swallowing. Caution with medication as secretion management advice from a speech therapist can be very helpful as first-line intervention.*
 - *Consider other options including dark grape juice sipped regularly to help clear thick throat secretions. Pineapple juice and papaya tablets can also be used.*
 - *Atropine eye drops – 0.5% one drop sublingual up to 4 times a day, as required.*

- Avoid overdose or use in elderly patients, as anticholinergic medications can cause confusion, or urinary retention.
 - Ensure that the use of oral atropine drops is closely supervised to avoid the risk of overdose.
 - Use with caution in patients on antipsychotics.
- Donnatal or probantheline – 15 mg one tablet orally, 3 times a day.
 - Amitriptyline – titrate dose up from 12.5 mg orally at night to 25 mg, maximal dose 50 mg.
 - When severe, subcutaneous glycopyrrolate – titrate dose up from 25 micrograms to maximal dose 100 micrograms, 3 times a day.
- **Constipation**
 - Check patient is hydrated
 - Stool softeners such as Coloxyl first-line
 - Bowel stimulants such as senna or Duro lax for slow bowel transit
 - Movicol if able to swallow
 - **Spasticity**
 - Baclofen – titrate dose up from 5 mg orally, twice per day, to 25 mg three times per day, but beware of sedation
 - Dantrolene – 25 mg orally daily, titrate up to 200 mg daily
 - **Breathlessness**
 - Non-invasive ventilation may improve ventilation, but ensure patient has a PEG/RIG if using during the day as well, to ensure adequate nutrition.
 - Oral liquid morphine – titrate up from 2 mg/mL, 0.5 to 2 mL orally, as required.
8. Consider sleep medicine specialist assessment to determine if non-invasive ventilation is appropriate.
9. Seek advice from a pharmacist on best formulations for patients with difficulty swallowing, as rationalisation and [deprescribing](#) of medications often needs to occur as the disease progresses.
10. If oedema (often associated with immobility and dependent position):
- Compression stockings
 - Elevated position
 - Massage

Referral

- If suspected or confirmed motor neurone disease, request a [urgent or routine neurology assessment](#).
- While awaiting a diagnosis, consider requesting community physiotherapy and occupational therapy assessments support for the patient and their family.

- If symptoms of sleep-disordered breathing, e.g. excessive daytime somnolence, dry mouth, early morning headaches, request sleep medicine specialist assessment for consideration of non-invasive ventilation.
- Request home occupational therapy assessment for equipment needs for patients aged:
 - < 65 years via adult occupational therapy assessment, the [MND Association of Victoria](#), or [NDIS](#).
 - ≥ 65 years via aged care assessment.
- Where bulbar function is impeding swallowing and weight is dropping, request urgent dietitian assessment and speech pathology assessment.
- For difficult symptoms, end of life decision-making, guidance on [Advance Care Planning](#), and in advanced disease, consider requesting palliative care services.

Information

For health professionals

Further information

- Better Health Channel – [Motor Neurone Disease \(MND\)](#)
- [MND Australia](#)
- [MND Victoria](#)

For patients

[MND Victoria](#)

References

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3. MNDcare. ACT: MND Australia; [Constipation](#). 2014.

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