Motor neurone disease: a guide for GPs and primary care teams
# Quick reference

## Responsibilities when MND is suspected or confirmed

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<th><strong>Prompt referral for diagnosis</strong> (page 13)</th>
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<td>Use the Red Flag tool (page 14) to recognise early signs of MND in order to refer to <strong>neurology</strong> in a timely manner.</td>
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<tr>
<th><strong>Assessing needs</strong></th>
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<tr>
<td>Use a person-centred approach to listen to and ascertain the physical, social, emotional and spiritual needs of the person with MND and their carers at each appointment.</td>
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<tr>
<td>Refer to appropriate specialist teams as necessary.</td>
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<th><strong>Monitoring symptoms</strong></th>
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<td>Monitor and assess symptoms, including signs of:</td>
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<td>- <strong>Respiratory involvement</strong> (page 36) – early signs should trigger referral to the <strong>specialist respiratory team</strong>.</td>
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<tr>
<td>- <strong>Cognitive change</strong> (page 44) – this has implications for decision-making and future management.</td>
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<tr>
<td>In collaboration with consultants in <strong>neurology</strong> and <strong>palliative care</strong>, initiate appropriate management and treatment, including anticipatory symptomatic intervention.</td>
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<th><strong>Support and information</strong></th>
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<td>Provide support and information throughout the disease course.</td>
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<td>Advise on the need to inform insurers and DVLA or DVA of diagnosis.</td>
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<td>Complete a DS1500 form to support a benefit application (page 53).</td>
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<th><strong>Repeat prescriptions for riluzole</strong> (page 20)</th>
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<td>Issue repeat prescriptions if the person with MND is prescribed riluzole by their <strong>neurologist</strong>: a shared-care protocol should be agreed.</td>
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<th><strong>Palliative care</strong> (page 55)</th>
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<th><strong>Advance care planning</strong> (page 57)</th>
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<tr>
<td>Help the person with MND to talk through management options, including end of life decisions and Advance Decisions to Refuse Treatment (ADRT), as early as possible.</td>
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Motor neurone disease (MND) is a progressive and ultimately fatal disease that results in degeneration of the motor neurones, or nerves, in the brain and spinal cord.\(^1\)

Around two people in every 100,000 of the general population will develop MND each year,\(^2\) so GPs and others in the primary care team may come across few people living with MND during their professional lifetimes. However, as a member of the primary care team you can make a great difference to the quality of life of those people, with timely care, support and symptom management.

Whether you are a GP, nurse or other member of the primary care team, this booklet is designed to give you information and practical tips on the care of someone with MND, and the role you and your team can play.

### RCGP online module in motor neurone disease

Visit [www.mndassociation.org/gp](http://www.mndassociation.org/gp) to access the RCGP online module on MND, along with other tools and information for GPs.

### How the MND Association can support you

The MND Association supports professionals to care for people affected by MND in a variety of ways, which includes providing a range of information and educational opportunities as well as local support and advice from our staff and volunteers.

See page 69 of this guide for more information about how the MND Association can support you in your role.

### MND Connect

Our helpline offers information and support by telephone and email – on **0808 802 6262** or **mndconnect@mndassociation.org**

### Transforming MND Care

Transforming MND Care is a free, simple to use audit tool developed by the MND Association which helps to improve outcomes and enhance practice. It supports implementation of recommendations in the NICE guideline on MND (NG42).\(^3\)

To find out more email **audittool@mndassociation.org** or call **01604 611770**. Register at [www.mndassociation.org/forprofessionals/transforming-mnd-care](http://www.mndassociation.org/forprofessionals/transforming-mnd-care)
The role of the GP and primary care team

The GP and primary care team can be pivotal to the person with MND maintaining a dignified quality of life throughout the course of the disease.

Working with the specialist team

Where a person with MND is being seen regularly by a neurologist, a shared-care approach may be agreed between the neurologist and GP. This is particularly important when the neurologist is based some distance away and access becomes increasingly difficult as the disease progresses.

The GP is responsible for liaising closely with and, in some cases, co-ordinating the professionals involved in the care of the person with MND. This may involve attending multidisciplinary team meetings.

The GP may work in liaison with an MND care co-ordinator, who may be based in an MND care centre or network, where such a service exists.

Visit our website at www.mndassociation.org/carecentres to find out where your nearest MND care centre or network is located.

Onward referrals

While the primary care team may be able to meet some of the needs of the person with MND and manage many of their symptoms, the input of other health and social care professionals is vital to the care and support of people with MND.

The GP may act as the gatekeeper to other services, referring to appropriate specialists in the multidisciplinary team, including those in the table over the page.
<table>
<thead>
<tr>
<th>Professional</th>
<th>Area of expertise/assistance provided</th>
</tr>
</thead>
<tbody>
<tr>
<td>District/community nurse or community matron</td>
<td>nursing support at home</td>
</tr>
<tr>
<td>Physiotherapist</td>
<td>mobility (walking, balancing etc), exercise and activity, positioning, posture, respiratory, weak cough, difficulty clearing sputum</td>
</tr>
<tr>
<td>Respiratory team</td>
<td>coughing, breathlessness at rest/slight exterion/lying flat, sleep disturbances, daytime sleepiness, morning headaches, tiredness or lethargy, advice on positioning for drooling/thick mucus</td>
</tr>
<tr>
<td>Occupational therapist</td>
<td>mobility (walking, balancing etc), manual handling, positioning, fatigue management, aids/equipment for activities of daily living</td>
</tr>
<tr>
<td>Speech and language therapist</td>
<td>swallowing and diet modification, positioning, coughing, speech and communication, communication aids and equipment, voice banking</td>
</tr>
<tr>
<td>Dietitian</td>
<td>weight loss, lack of appetite, diet modification, gastrostomy</td>
</tr>
<tr>
<td>Palliative care services/hospice</td>
<td>general symptom management, support for individual and family, uncontrolled pain, breathlessness, fear/anxiety/depression, spiritual support</td>
</tr>
<tr>
<td>Social worker</td>
<td>difficulties in activities of daily living, advice on benefits and entitlements, referral to local services, support for individual and family, counselling</td>
</tr>
<tr>
<td>Psychologist/counsellor</td>
<td>fear/anxiety/depression, cognition and frontotemporal dementia</td>
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</tbody>
</table>
# Actions for the GP and primary care team

<table>
<thead>
<tr>
<th>Question</th>
<th>If no:</th>
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</thead>
<tbody>
<tr>
<td>Can you identify the person with MND as having a neurological condition</td>
<td>Use the appropriate coding so your system flags when the person contacts the surgery and the appropriate response can be given.</td>
</tr>
<tr>
<td>on your practice IT system?</td>
<td></td>
</tr>
<tr>
<td>Is the person’s care regularly discussed within the primary care team?</td>
<td>Put the patient’s name on the Supportive Care Register within your practice.</td>
</tr>
<tr>
<td>Is there a member of the primary care team who has been nominated as the</td>
<td>Identify a named person who: (a) acts as the single point of contact within the team and (b) can liaise with all health and social care professionals involved in the person’s care.</td>
</tr>
<tr>
<td>co-ordinator of the patient’s care?</td>
<td></td>
</tr>
<tr>
<td>Are efforts being made to control all symptoms currently experienced by</td>
<td>Use the sections in this booklet to look at options for management. If symptoms are still uncontrolled, refer onwards.</td>
</tr>
<tr>
<td>the person with MND?</td>
<td></td>
</tr>
<tr>
<td>Is it flagged with the out-of-hours service provider that this person</td>
<td>Notify your out-of-hours service provider and ensure access is given to all relevant facts/wishes.</td>
</tr>
<tr>
<td>has MND?</td>
<td></td>
</tr>
<tr>
<td>Are plans in place for emergency care provision if the person’s carer</td>
<td>An assessment of care needs should consider this. Refer to social services.</td>
</tr>
<tr>
<td>cannot support for any reason?</td>
<td></td>
</tr>
<tr>
<td>Question</td>
<td>If no:</td>
</tr>
<tr>
<td>-------------------------------------------------------------------------</td>
<td>-------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Do you know what the patient’s wishes are in relation to end of life?</td>
<td>Use active listening skills to clarify these as the opportunity presents.</td>
</tr>
<tr>
<td>Have end of life wishes been recorded, so other members of the primary</td>
<td>Encourage the patient to record their wishes and make other members of their family and</td>
</tr>
<tr>
<td>care team are also aware?</td>
<td>the primary care team aware.</td>
</tr>
<tr>
<td>Do you have a copy of any existing DNACPR order/ADRT/advance care</td>
<td>Ask to keep a copy in your patient records.</td>
</tr>
<tr>
<td>plan/preferred place of care request in your practice records?</td>
<td></td>
</tr>
<tr>
<td>Is the ambulance service aware of any DNACPR order/ADRT/preferred</td>
<td>Notify the ambulance service so it can be flagged on its system to prevent inappropriate</td>
</tr>
<tr>
<td>place of care request?</td>
<td>care or treatment.</td>
</tr>
<tr>
<td>Are you aware of the name of the main carer for this person?</td>
<td>Record the name of the main carer and their contact details on your practice system.</td>
</tr>
<tr>
<td>If the carer is registered with your practice, is their record</td>
<td>Put a flag on the carer’s record so you are aware of any potential impact on the carer’s</td>
</tr>
<tr>
<td>cross-referenced on your system?</td>
<td>health.</td>
</tr>
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</table>

"Often health professionals don’t realise how swiftly our needs change and how little time we have."  
- A person living with MND
MND is characterised by progressive degeneration of motor neurones:

- anterior horn cells and their axons – resulting in lower motor neurone (LMN) dysfunction, muscle weakness and wasting
- upper motor neurone (UMN) cell bodies and axons of the corticospinal and corticobulbar tracts – resulting in UMN lesions, characterised by weakness and spasticity.

The clinical hallmark is progressive motor weakness in the limbs, trunk or bulbar regions, without sensory disturbance.¹

MND is insidious in its onset and development. The initial stages, the speed and the pattern of progression can all be very variable.

Not all symptoms necessarily happen to everyone, nor do they develop in the same order or progress at the same rate.⁴

Diagnosis is often very difficult (see page 13). Sometimes it is necessary to review a person for some time before a diagnosis becomes relatively certain. This can pose real problems for both the patient and their family.

**Common first symptoms**

MND causes progressive muscular weakness that may present as isolated and unexplained symptoms. These may include:

- stumbling, falls or trips, foot drop
- loss of dexterity, weakened grip
- change in voice quality, such as quiet or slurred speech
- awareness of swallowing changes, such as having to ‘double swallow’
- cramps, fasciculations, muscle wasting.³
First symptoms may progress to:

• loss of function of limbs
• weakness and wasting of trunk and neck muscles
• increased impairment of speech and swallowing, leading to total loss of function.

Many people with MND will eventually be completely dependent on others.\(^5\)

Respiratory muscles are likely to be involved in later disease, leading to breathlessness and symptoms of hypoventilation. Occasionally, these may be the presenting symptoms, with excessive daytime sleepiness, fatigue, early morning headache or shortness of breath on exertion or when lying down.\(^3,6\)

Some people with MND may experience cognitive changes, ranging from mild effects to noticeable impairment. Some will experience frontotemporal dementia (MND-FTD).\(^1\)

Some people will present with behavioural changes, emotional lability (not related to dementia) or FTD, and then go on to display symptoms of MND.

If there is concern about cognition and behaviour, explore these changes with the person and their family members and/or carers as appropriate (see page 44).\(^3\)

**Prognostic factors**

These factors, if present at diagnosis, are associated with shorter survival:

• speech and swallowing problems (bulbar presentation)
• weight loss
• poor respiratory function
• older age
• lower Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS or ALSFRS-R) score
• shorter time from first developing symptoms to time of diagnosis.\(^3\)
Additional symptoms

• fatigue – this is common throughout the disease and can be severe, affecting remaining functional ability\(^7\)

• saliva clearance, tenacious saliva or mucus (see page 29)\(^8\)

• emotional lability – inappropriate or excessive crying or laughter (see page 52)\(^9\)

• anxiety and depression (see page 51)\(^{10}\)

• pain and discomfort from muscle cramps or spasticity (see page 42)\(^{11}\)

• insomnia\(^6\)

• constipation (see page 33).\(^{12}\)

MND rarely affects:

• the cranial nerves that control eye movement and the lower sacral segments of the spinal cord (the latter affect continence), although they may become involved in the very late stages of MND.\(^5, 13\)

Note: While MND rarely affects bowel and bladder function, immobility may affect the ability to get to the toilet on time and the strength required to defecate.

• sexual function – this remains intact in many people with MND.\(^{14}\)

  However, physical impairment, interventions and self-esteem may affect sexual expression and intimacy.

• the senses (sight, hearing, sensation) are generally unimpaired.\(^1\)

Cause of death

This is almost always due to respiratory failure as a consequence of respiratory muscle weakness and/or repeated chest infections.\(^1\)

It is very important to reassure people with MND and carers that death from choking is exceptional.

For some people with MND, death can be very sudden, before an obvious end stage is reached. Others experience a longer final stage, which can last many weeks. For most people with MND, death will be peaceful.\(^{15}\)
Diagnosis of MND

Rapid and accurate diagnosis, performed by a medical doctor experienced in MND care, usually a neurologist, is crucial in ensuring the needs of people living with MND are met from the earliest possible stage.¹

Accurate recognition of the condition, explanation of the likely progression of symptoms within a supportive therapy team, and prompt introduction of appropriate treatments and interventions, all help to improve quality of life for people with MND.

There is no single diagnostic test for MND. Diagnosis is based on features in the clinical history and examination, usually accompanied by electrophysiological tests, including EMG and nerve conduction studies.

Other tests may include:
- MRI scanning of the brain and spinal cord
- various blood tests
- lumbar puncture
- muscle biopsy.

The purpose of these tests is to exclude the presence of other neurological conditions, with particular focus on those with treatments likely to be effective if administered early.

In the early stages of MND, symptoms can be similar to those seen in other conditions, so people may spend months seeing various specialists and undergoing unsuccessful treatments until MND is suspected.

When someone is diagnosed with MND, and during the diagnostic process, it is important that they, and family members/carers, have suitable support. This includes a point of contact to return to with questions and access to appropriate information.

They may take time to adjust and come back with questions some time afterwards. See page 15 for topics people may want to discuss.
Reg Flag tool for diagnosis

The MND Association’s Red Flag tool for GPs, produced in partnership with the Royal College of General Practitioners, is designed to improve timely referrals to neurology and therefore speed up the time to accurate diagnosis.

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.

This tool aims to reduce any delay in referral by prompting GPs to look for additional symptoms and consider the possibility of a neurodegenerative condition. GPs are not expected to be able to make the diagnosis.

Download a copy of the full tool at [www.mndassociation.org/redflag](http://www.mndassociation.org/redflag)
Confirmation of diagnosis

The NICE guideline on MND\(^3\) recommends that, if MND is suspected, the possible diagnosis should be specified in the referral letter. The GP should contact the consultant neurologist directly if they think the person needs to be seen urgently.\(^3\) Be aware that the patient may be copied into this letter.

The NICE guideline also advises that a person’s GP should be informed of a consultant neurologist’s suspected or confirmed diagnosis of MND without delay, and given information about likely prognosis.\(^3\)

The person with MND should be offered a face-to-face, follow-up appointment with a healthcare professional from the multidisciplinary team, to take place within four weeks of diagnosis.\(^3\)

The guideline also recommends that professionals should set aside enough time to discuss the person’s concerns and questions. Topics may include:

- their understanding of MND and how it affects daily living
- the management and treatment of MND, who will be involved and what will happen next with their healthcare
- accepting and coping with the diagnosis and prognosis, including concerns and fears about life expectancy and dying
- their ability to continue with current work and usual activities
- adjusting to changes in their life and their perception of self
- changes in relationships, familial roles and family dynamics
- sexuality and intimacy
- concerns about their family members and/or carers
- how to tell family and friends about MND
- concerns around whether their children will get MND
- decision-making.\(^3\)

See the *psychological support* section on page 48 for more information about talking to people before and after diagnosis of MND.
Precise figures are not certain:
• Incidence is thought to be around 1-2 per 100,000 per year.\(^2\)
• Prevalence is thought to be around 7 per 100,000 of the UK population (from prospective figures calculated by Chio et al 2013).\(^{16}\)
• Based on the size of the UK population, this equates to 4,500 people currently living with MND.\(^{16}\)

The MND Association is currently funding a national population-based register, known as the MND Register for England, Wales and Northern Ireland, to accurately update these numbers.

• MND can affect adults of any age. However, incidence is highest in people aged 55-79, and onset below the age of 40 is uncommon.\(^3\) Onset under the age of 30 is extremely rare.\(^{17}\)
• The male:female ratio is 3:2, although this varies with age and evens out in later years of life.\(^{18}\)
• Clinical presentation is sub-divided in two ways: by the presence of upper motor neurone (UMN) and/or lower motor neurone (LMN) signs, and the anatomical area first affected.\(^{19}\)

Site of onset
• Limb-onset MND affects about two-thirds of people with MND.\(^{19}\)
• Bulbar-onset MND affects about a third of people with MND.\(^{19}\)
• Respiratory-onset is rarer, affecting about 3%.\(^{19}\)

ALS and MND are commonly used to mean the same thing.\(^{20}\) MND is the overarching term used in the UK and Australia to denote various diseases of the motor nerves, classified by whether they affect UMN, LMN or both. There is considerable overlap between the different forms of MND. The variants, or subtypes, are:

Amyotrophic lateral sclerosis (ALS)
• Most common form: represents almost 90% of all MND cases.\(^{20}\)
• Involves UMN and LMN.
Incidence and prevalence of MND

- Characterised by a combination of muscle wasting with spasticity.
- Arm, leg or bulbar (speech and swallowing) onset.
- The bulbar region is usually affected, but may not be prominent in everyone with ALS.
- Wide variation in age of onset: mean age at onset is mid-60s.
- Survival is usually less than five years from symptom onset; where initial onset is in the bulbar region, survival may be shorter (1-2 years).

**Progressive bulbar palsy**\(^4,21\)
- A small group of people with bulbar-onset MND have symptoms relatively confined to the bulbar region for several months (occasionally years), before it moves to involve the limbs.
- Characterised by rapidly progressive speech and swallowing problems, often also with emotional lability. Muscles in the upper limbs, neck and shoulder girdle may become progressively weaker.
- Both UMN and LMN may be involved.
- Overall survival 6 months – 4 years.
- Tends to affect people over 55.

**Progressive muscular atrophy (PMA)**\(^19,22\)
- Accounts for 5-10% of people with MND.
- Characterised by LMN degeneration, with muscle wasting/weakness.
- Limb onset, often with visible fasciculations.
- ‘Flail arm syndrome’ falls within this group, characterised by slowly progressive, symmetrical, and usually proximal upper limb weakness.
- Survival is often longer than seen in ALS, typically more than four years; if someone presents with a single flail arm or leg, survival is often five to 10 years or more from onset.

**Primary lateral sclerosis (PLS)**\(^4,19,23\)
- Accounts for approximately 2% of all people with MND.
- Affects UMN only.
- Characterised by spasticity/brisk reflexes. Balance is often impaired.
- Survival is notably longer (more than 10 years from symptom onset).

In some cases, people present with features of PMA or PLS, but go on to develop more typical ALS.\(^19\) It is well recognised that some individuals live considerably longer than the average survival times given.\(^1\)
Aetiology

Most people with MND have no evident family history of the disease. In these apparently sporadic cases, it is likely that the disease develops due to a combination of genetic susceptibility and environmental factors.¹

The most clearly identified risk factor for MND in people without a family history of the disease is increasing age. Some studies suggest a slight increased risk with cigarette smoking.²⁴

Inherited forms of MND

Inherited MND (sometimes known as familial MND) accounts for approximately 5-10% of all people with MND.¹

Mutations in several different genes can be detected in about three quarters of people with inherited forms of MND.¹

• 45% of inherited cases are caused by gene expansion mutations in the gene C9ORF72.²⁵

• Fewer than 20% are caused by mutations in the superoxide dismutase 1 (SOD1) gene.¹

• Changes in the genes TDP43 and FUS are each responsible for 5% of inherited cases.¹

Research is ongoing to identify further genes involved in inherited MND. The sporadic and inherited forms of MND are clinically indistinguishable.¹³

Genetic testing can be arranged for people with MND who have a family history of the disease. It is not generally undertaken in those without a family history and should only be carried out after extensive genetic counselling, due to the huge psychological and practical implications.
Mechanisms of motor neurone degeneration

The pathogenic processes underlying MND are likely to be multifactorial. Current evidence suggests interplay between several mechanisms including:

- abnormal intracellular protein aggregation
- glutamate-mediated excitotoxicity
- oxidative stress
- mitochondrial dysfunction
- axonal transport dysfunction
- neurotrophic factor dysfunction
- glial cell dysfunction.

Information you can share

Our resources for people affected by MND include:

Research sheet A – *Introduction to MND research*

Research sheet B – *Inherited motor neurone disease*, which is split into three parts:

1 – *Introduction*

2 – *Genetic testing and insurance*

3 – *Options when starting a family*

See page 69 for details of how to order copies or download from our website at [www.mndassociation.org/researchsheets](http://www.mndassociation.org/researchsheets)

You can also contact our research team with any queries on **01604 611880** or [research@mndassociation.org](mailto:research@mndassociation.org)
Management of MND

Riluzole is the only licensed drug available that prolongs life in MND. Clinical trials have shown that, on average, it extends survival by around three to six months if taken for 18 months.\(^{26}\)

Its benefit does not show in tests of improved muscle strength. Instead it seems to reduce, on average, the rate of decline of function in MND. People with MND who take riluzole should be made aware that they will not feel any stronger when they take the drug.

How the drug acts in MND is unknown, but it is thought to modulate the release of glutamate and promote neuronal survival by enhancing production of various neuronal growth factors.\(^{27}\)

**Prescribing riluzole**

Riluzole is recommended for use in MND by NICE.\(^{28}\) It is not suitable for everyone with the condition, so generally a hospital consultant will assess a person’s suitability for riluzole and prescribe the first course of the drug.

After this, the GP and specialist should agree a shared-care protocol, under which the GP issues repeat prescriptions. In a few areas however, the prescribing and dispensing of riluzole is undertaken by the specialised centre throughout the patient’s illness.

Normal baseline blood test should be undertaken initially, then monthly full blood count and liver function test for three months, followed by three-monthly for the first year, and annually thereafter.\(^{29}\)

Responsibility for prescribing and monitoring should be made clear in the protocol.

Riluzole is available in tablets in generic or branded form (Rilutek). A liquid form is also available (Teglutik), which may be easier for some people to swallow.

**New treatments**

A number of ongoing studies are testing the efficacy of new drugs in the treatment of MND and existing drugs that are currently used for other conditions. Research with stem cells, in both familial and sporadic MND, is also now underway.
You can track the progress of research at: www.mndassociation.org/treatment-trials

**Symptom management**

Sometimes, when people are told there is no effective curative treatment for MND, they interpret this as meaning that nothing can be done. Some will struggle on without asking for advice and treatment for symptoms, which could alleviate discomfort. They should be encouraged to talk about the symptoms they have and the potential treatments available. There are many other drugs that can be offered for the management of particular symptoms (see pages 22 to 47).

**Free prescriptions**

Prescribed medicines are free for everyone living in Wales and Northern Ireland.

People with MND in England who do not qualify for another exemption may be able to claim free prescriptions by applying for a medical exemption certificate. The current regulations state that people can apply if they have a continuing physical disability which means they can’t go out without the help of another person. A prescription exemption certificate will need to be completed.

**Complementary therapies**

Although they provide no treatment for MND, many people living with the disease use complementary therapies, such as aromatherapy, reflexology, massage, relaxation techniques and acupuncture. There is no controlled trial evidence of the benefits of these therapies in MND, but some people report improvements in wellbeing after having such complementary therapies. It is important the therapist has some knowledge of MND and neither the person with MND nor the therapist has false expectations that the complementary therapy will offer a cure. The treatment being undertaken should also be safe and affordable.

**Unproven treatments**

The MND Association co-funds an international group of scientists and clinicians, collectively known as ALS Untangled, to investigate unproven or alternative treatments for MND. This group looks for the evidence behind these unproven treatments and concludes with their own recommendations. See www.alsuntangled.com
Information you can share
Our resources for people affected by MND include:

• Information sheet 5A – *Riluzole*
• Information sheet 6B – *Complementary therapies*
• Research sheet H – *Accessing unapproved drugs*

See page 69 for details of how to order copies or download from our website at [www.mndassociation.org/publications](http://www.mndassociation.org/publications)

Muscle weakness
Muscle wasting in MND causes weakness and can affect balance and posture, with the risk of falls. General muscle tightness or spasticity (stiffness, rigidity and spasms) can affect mobility, co-ordination of movement and may cause pain or increase the risk of falls.

Consider:

• physiotherapy (including walking aids, orthotics, splints, active and passive exercise, posture management)
• occupational therapy for posture management and equipment for managing activities of daily living
• medication for spasticity (see page 43).

Fasciculations:

• are often among the first signs of MND
• may be felt as a rippling effect in individual muscles, but can be more widespread
• may be so obvious they can be seen by other people
• can seem worse when a person is stressed
• usually ease over time.

Ability to drive
Everyone with MND is required to inform the DVLA or DVA and their insurer immediately after diagnosis. They will usually be permitted to continue driving until weakness and/or cognitive change compromise safety. Judgement of continued driving capacity should be regularly discussed.
Dysarthria
80% of people with MND are affected eventually with slurred, quiet or complete loss of speech.\textsuperscript{33}

Cause
Muscle weakness and/or spasticity leads to a reduced range of movement in the:
• tongue
• lips
• facial muscles
• pharynx and larynx\textsuperscript{,34}
Reduced palatal elevation can lead to hypernasal voice quality.\textsuperscript{34}
Reduced breath support can lead to reduced volume or breathy voice quality.\textsuperscript{34}

Impact
Progressive difficulty with articulation, slurred speech and/or loss of volume.
May rapidly lead to anarthria, even though limb function may be maintained for many months.\textsuperscript{34}

Treatment
Early referral to \textit{speech and language therapist} (SLT) who will:
• examine the patient’s range of movement in their lips, tongue and palate
• give advice on voice banking and strategies for communication.
The SLT can also arrange for assessment and provision of Augmentative and Alternative Communication (AAC).
Options include:

\textbf{Low-tech}
• pen or pencil and paper
• eye-pointing frames, sometimes known as an E-Tran (eye-transfer) frame
• a word/message chart, where useful or frequently used words or phrases can be pointed at by the person with MND or the listener
• an alphabet chart, where the person with MND or the listener can point to letters to spell out words
• a personalised communication book.

**High-tech**
• portable electronic communication aids with a keyboard, symbol or scan screen such as Eye Gaze, and computers or tablet devices with text-to-speech software.

An **occupational therapist (OT)/the neuro rehabilitation service/orthotics service** can assist with advice on:
• seating, positioning, wrist and head supports
• switches and pointers
• mobile arm supports and tables to access communication aids
• environmental controls.

**Dysarthria may exacerbate emotional reactions, including:**
• isolation – communication inadequate or avoided
• frustration – difficult or impossible to be understood; the person needs time that may not be available
• increased fear and anxiety – being unable to discuss these fears and anxieties
• low self-esteem – presumption by others that they are either deaf, drunk or intellectually impaired
• loss of control – because they are misunderstood or their opinion is ignored or not sought
• increased sadness.

Allow time to explore and discuss the above issues.

Be aware that cognitive impairment (see page 44) can cause problems with communication and the ability to learn and use alternative communication methods.34
Communicating with someone with MND

Do:

• find out how the person with MND prefers to communicate and any equipment they like to use

• make sure any necessary communication aid is available and the person with MND is in the best position to access it

• have a pen and paper handy so that the person, if they are able, can write down any appropriate words: this helps to reduce frustration and misunderstanding

• sit face to face and watch the person’s eyes, lips and gestures: unspoken communication is important

• make sure the environment is conducive to communicating: check lighting and reduce background noise where possible

• ensure the person with MND is comfortable and is the focus of communication

• allow the person time and create a relaxed atmosphere

• encourage the person with MND to slow down and over-emphasise words to help clarity

• check back with the person on what you think has been said and admit when you don’t understand

• find out whether the person uses a simple code for ‘yes’ and ‘no’: this can be a movement of any part of the body that can be carried out reliably and does not cause fatigue, for example eye blinking

• make it easier for the person to contact you: if they struggle to speak on the phone, they may prefer to use email or text messaging

• make an audio recording of discussions or appointments so the person can listen back later when they are less fatigued, or if they missed any detail: attending appointments can be exhausting

• encourage the person with MND to make a written list, or store a message on their communication aid, before their appointment, of the specific areas they wish to discuss: this will make the appointment less tiring and will ensure all issues are covered

• remember that speech problems may mask signs of cognitive change (see page 44).
Try not to:

• alter the rate or sound of your speech, unless the person with MND has asked you to, or a speech and language therapist has recommended that people should speak more slowly to the person

• finish the sentences of the person with MND, unless they ask you to, and avoid interrupting them

• use family members or carers as a translator for the person with MND, unless it is clear that this is what the person with MND wants

• use open questions. Instead use closed questions which can be answered with yes, no, or a single word.

Information you can share

Our resources for people affected by MND include:

• **Notepad for people with speech difficulties**

• **Speech card for people with speech difficulties** – people can carry this card to let others know that they have problems speaking but can understand what is being said

• **Information sheet 7C – Speech and communication support**

• **Information sheet 7D – Voice banking**

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*Losing your voice is a big deal and can be very isolating.*  
- A person living with MND
Dysphagia
Swallowing problems affect at least two thirds of all people with MND during the course of their illness.\textsuperscript{35} When a patient presents with this symptom, deterioration is inevitable and referral to appropriate professionals should be made without delay.

There may be times when a person with dysphagia will have episodes of coughing and choking. Even though a choking episode is very distressing for them and their carer at the time, they can be reassured that choking is very rarely a direct cause of death.\textsuperscript{1}

**Cause**
As for dysarthria (see previous section).

**Effect**
Problems swallowing liquids results in coughing when drinking. This may be the first sign of early dysphagia.

**Other signs include:**
- loss of ability to form lip seal, chew food with the tongue and/or form a bolus
- weak or absent swallow reflex
- food not moving to the back of the mouth or spontaneously moving down the throat due to a weak or absent swallow reflex, or an unco-ordinated swallow caused by weak pharyngeal muscles
- muscle spasm
- more time needed for chewing, several swallows needed for each mouthful of food and becoming tired on eating and drinking
- a wet or muffled sounding voice after eating.\textsuperscript{36}

**This can result in:**
- sialorrhoea (drooling due to impaired swallowing of saliva rather than excess production), see page 29
- dehydration and weight loss from not eating enough
- aspiration and recurrent chest infection
- problems swallowing medication
• tiredness and loss of strength and wellbeing
• lengthy mealtimes, leading to food going cold
• constipation, see page 33.

There may be a social impact too; the person with MND may feel unable to eat with other people due to dysphagia. They may join others and take a few spoonfuls, while eating most of their meal before or afterwards.

Treatment

Assess nutritional and fluid intake and weight. Weight loss is often related to dysphagia, but may also be due to upper limb weakness or fatigue affecting the ability to use cutlery or cups independently. Respiratory symptoms and risk of aspiration and/or choking should also be assessed.

A dietitian can suggest management strategies, including altered meal patterns (eg eating little and often). An early referral is beneficial for dietetic advice and support.

A speech and language therapist can assess ability to eat and swallow and can monitor for changes. They may suggest altering the consistency of the diet and/or suggest prescribing thickeners for fluids.

A physiotherapist or occupational therapist can advise on head supports, seating and positioning.

Dysphagia and cognitive change

Additional support will be needed for people with dysphagia and cognitive impairment.

People with more severe cognitive changes may place too much food in their mouth at one time and cram food, while others may eat more food than they need or have a preference for sweet food. It may be more difficult to follow advice on safe swallowing, so repeated reminders and supervised eating may be necessary.
Saliva management

Excess saliva is a commonly reported symptom of MND. The actual amount of saliva produced is normal ie two to three pints a day. Problems with saliva usually result from poor lip seal and/or impaired ability to swallow.8

Patients have either runny, thin saliva that drools out of the mouth or thick, tenacious saliva that is difficult to clear.8

It is crucial to establish the volume and viscosity of saliva, as medication to dry up saliva can worsen the situation if the saliva is already thick.3,8

Medication for thin, runny saliva

If a person with MND has problems with drooling of saliva (sialorrhoea), provide advice on swallowing, diet, posture, positioning and oral care. The NICE guideline on MND3 recommends:

• a trial of antimuscarinic medicine as the first-line treatment for sialorrhoea in people with MND, eg hyoscine (tablets or transdermal patches – be aware of the possible side effect of confusion in the elderly).

• glycopyrrolate as the first-line treatment for sialorrhoea in people with MND who have cognitive impairment, as it has fewer central nervous system side effects.

Atropine drops on the tongue may be used for short term relief.38

For drug dosages, refer to British National Formulary (BNF) or Palliative Care Formulary. Take into account the person’s needs and preferences and whether they may have any difficulty swallowing medication.

Botulinum toxin A

If first-line treatment for sialorrhoea is not effective, not tolerated or is contraindicated, consider referral to a specialist service for Botulinum toxin A.3

Be aware that increasing dysphagia may be a side effect of Botulinum toxin A, so some suggest only resorting to this treatment if the person already has an enteral feeding tube in situ. Botulinum toxin A should only be administered by specialist practitioners.
Radiotherapy
Another possible treatment for excessive, watery saliva is the use of radiotherapy to destroy part of the salivary glands.\textsuperscript{8}

Suction
Portable oral suction units are helpful if saliva builds up in the mouth.\textsuperscript{8}

Body position
Attention is needed to seating, general posture and head support\textsuperscript{39} – consult a physiotherapist or occupational therapist.

Medication and strategies for thick, tenacious saliva
If a person with MND has thick, tenacious saliva:

- review all current medication, especially any treatments for sialorrhoea\textsuperscript{3}
- provide advice on swallowing, diet, posture, positioning, suctioning, hydration and pay attention to oral hygiene – consult district nurse\textsuperscript{3}
- consider treatment with humidification, nebulisers and carbocisteine. Carbocisteine is available as capsules or liquid,\textsuperscript{40} which can be administered via a feeding tube.\textsuperscript{3, 40, 41}

For drug dosages, refer to British National Formulary (BNF) or Palliative Care Formulary. Take into account the person’s needs and preferences and whether they may have any difficulty swallowing medication.

Also:

- Suggest pineapple/papaya juices,\textsuperscript{38} or flavoured ice cubes. These juices contain bromelain/papain (respectively). These hygroscopic agents are also available as tablets from health food stores.

See page 35 for information on dealing with weak cough.

Dry mouth\textsuperscript{42}

- Check and change dosage of medications if needed.
- Consider artificial saliva sprays or gels, for example Aquoral, Biotene Oralbalance or Xerotin.
- Pay careful attention to oral hygiene.
- Increase fluid intake, whether orally or through a feeding tube.
Nutrition and enteral feeding

Monitoring nutritional intake and weight is important. Unintentional weight loss and risk of malnutrition can result from:

- dysphagia, hypermetabolism, respiratory insufficiency
- depression, anxiety, cognitive impairment
- constipation, fatigue, social isolation
- physical difficulties buying, preparing and eating foods
- dislike of modified foods, increased time taken to finish meals
- reduced dexterity and strength affecting ability to hold utensils.

Functional consequences of weight loss/malnutrition include:35

- increased muscle wasting due to muscle catabolism
- increased respiratory muscle weakness
- decreased strength and mobility, reduced energy levels
- impaired immune function, which can increase susceptibility to opportunistic infections
- decreased tissue viability and increased risk of pressure sores
- increased discomfort sitting or lying due to weight loss and loss of tissue ‘padding’ over bony protuberances
- decreased morale and quality of life.

Early and ongoing involvement of a speech and language therapist and a dietitian is essential to assess, monitor and review the individual’s swallow function, aspiration risk and nutritional status and to advise on personalised interventions to enable the person’s nutritional needs to be met.35

Advice may include:

- dietary enrichment to optimise nutritional content
- recipe ideas to increase variety and optimise nutritional density
- practical solutions to ease food preparation and physical eating and drinking difficulties (in association with an occupational therapist)
- reduce aspiration risk by using softer foods/drinks thickeners
- recommending prescribed nutritional supplements
- discussing the option of gastrostomy feeding tube placement.
**Enteral feeding**

Alternative feeding methods should be discussed at an early stage and revisited as symptoms which limit oral intake present themselves. Introducing the option of a feeding tube early will give the patient time to make an informed decision.

The following may be offered to ‘top-up’ oral intake, or to meet their full nutritional and hydration needs, in addition to allowing administration of medications: nasogastric tubes (NGT), percutaneous endoscopic gastrostomy (PEG), radiologically inserted gastrostomy (RIG) and per-oral image-guided gastrostomy (PIG).

With RIG, the feeding tube is inserted under x-ray guidance. PIG is a hybrid of PEG and RIG but at present is not widely available.

**Nasogastric tube (NGT) placement**

NGT is usually a short-term option. It may be appropriate for someone who is malnourished or dehydrated while waiting for gastrostomy placement, or where gastrostomy placement is not possible.

**Gastrostomy placement**

PEG is the preferred method of gastrostomy when someone has good respiratory function, or PIG/RIG when there is significant compromise of respiratory function.

The optimal timing of gastrostomy is not entirely clear, but is likely to be when someone has lost around 5% of their body weight (from measurement taken at diagnosis). Earlier placement of a gastrostomy tube is often recommended. It may not be used immediately, but when needed, can improve or help maintain quality of life. Possible risks of a late gastrostomy include continued weight loss, respiratory complications, dehydration, failed insertion attempts, and a higher risk of mortality and procedural complications.

Someone considering a gastrostomy should be made aware of the level of support needed to manage living with this intervention, which includes deciding who will manage the feeds. The person with MND and their carer should be told about what is involved to ensure they can physically manage this. If extra care support is needed, arrangements should to be made at an early stage.
Some people will not want this type of intervention and their decision should be respected. Exploring pros and cons will enable the individual to make an informed choice. Refusal or withdrawal of gastrostomy in specific circumstances may be included in an Advance Decision to Refuse Treatment. When intervention such as gastrostomy is discussed, offer the opportunity to discuss or review any existing ADRT, DNACPR or Lasting Power of Attorney (pages 58 and 59).

A person can still continue to eat and drink orally for pleasure, even after having a gastrostomy. The speech and language therapist should suggest how to manage this safely.

**Constipation**

Sphincter muscles are not normally affected by MND. Changes in bowel function are usually the result of:

- physical inactivity
- reduced peristalsis
- low fluid intake
- reduced fibre intake
- use of analgesics
- weakened breathing
- weakness of pelvic floor/abdominal muscles.

**Treatment:**

- Maintain hydration and assess fibre intake.
- Stool softeners and stimulants may help. Liquid versions are available that can be given via a feeding tube.
- Use of suppositories, enemas or manual evacuation may be necessary on occasions.
- Remedies such as bulking agents and fruits with a high sorbitol content (e.g., prunes) may help and are readily available.

Diarrhoea may be due to constipation with overflow.

**Information you can share**

Our resources for people affected by MND include:

- *Eating and drinking with motor neurone disease (MND)*
- *Information sheet 7A – Swallowing difficulties*
- *Information sheet 7B – Tube feeding*
- *Information sheet 11B – Mouth care*

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Acute dyspnoea

Acute breathlessness is common in the later stages of MND, but may occur earlier.\(^6\) (For chronic respiratory insufficiency, see page 36.)

**Causes of breathlessness**

Weakened respiratory muscles, particularly the diaphragm, characteristically causes orthopnoea.

Acute breathlessness in MND is usually triggered by an acute event such as pneumonia, respiratory tract infection or sputum retention, against a background of previously undetected respiratory muscle weakness.\(^6\) However, other causes of acute breathlessness, such as pulmonary embolus or heart failure, should also be considered.

Once the acute event has been treated, consideration can be given to long-term care, which may include assisted ventilation (see page 38).

**Actions:**

- relieve fear and anxiety with calm reassurance
- request an MND Just in Case kit (see opposite)
- recommend flu and pneumococcal vaccination\(^47\)
- refer to a **consultant in respiratory medicine** or **palliative care team**, as close to the onset of respiratory symptoms as possible, to explore options. Involve family in these discussions as appropriate.

**Note:** The emergence of this symptom has implications for end of life decisions, increased dependency and being unable to communicate.\(^3\) (See sections on *psychological support, palliative care and respiratory management*.)

**Medication to consider:**

- buccal midazolam\(^48\) (liaise with **palliative care team** for dosage)
- sublingual lorazepam\(^48\) (slower acting than midazolam and ineffective in those with excess saliva)
- oral morphine (refer to **palliative care team**).\(^49\)

**Laryngeal spasm**

Impaired swallowing may predispose to this. It is often preceded by gastroesophageal reflux\(^50\) and can be extremely distressing. It can prevent breathing or speaking and can be accompanied by stridor.\(^51\)
**Treatment:**

- Consider lorazepam.\(^{48}\)

**Weak cough**

The NICE guideline on MND\(^3\) recommends the following cough augmentation techniques for people with MND who cannot cough effectively:

- unassisted breath stacking and/or manual assisted cough
- assisted breath stacking (e.g., using a lung volume recruitment bag with a one-way valve) for those with bulbar dysfunction or whose cough is ineffective with unassisted breath stacking
- use of a mechanical insufflation/exsufflation machine (sometimes known by the brand name CoughAssist), if assisted breath stacking is not effective and/or during a respiratory tract infection.

These techniques can enable expectoration of phlegm or mucus from the throat or lungs, thus reducing the likelihood of A&E admission and emergency intubation.\(^6\)

See page 30 for information on addressing thick secretions. For management techniques consult a **physiotherapist**.

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**The MND Just In Case kit**

The *MND Just in Case kit* is designed to hold medication for the relief of anxiety and breathlessness. Its presence in the home provides tangible evidence for people with MND and carers that fears have been addressed and practical help is at hand.

For the GP and district or community nurse it provides guidance on symptom management and storage for the prescribed medications.

Once the need for a kit has been discussed and agreed with the person with MND and their carer:

1. The GP orders a kit from MND Connect (see page 69) for a named person with MND. The kit is supplied free of charge.
2. The GP and district or community nurse agree on a plan to advise and support the carer in the use of the kit.
3. The GP prescribes medication to be supplied with the kit.
4. The GP, or community or district nurse, gives the kit to the person with MND and ensures the carer knows how it can be used.
Respiratory insufficiency management

Respiratory insufficiency arises in most patients late in the course of their disease, due to involvement of the diaphragm and muscles of the chest wall. It is the usual cause of death in MND.\textsuperscript{6}

For some patients, breathlessness, reduced vocal power, poor sleep quality or daytime fatigue may be the initial presenting symptoms. Rarely, patients may present to A&E in respiratory distress without any other symptoms.\textsuperscript{6}

Bulbar weakness contributes to various respiratory complications, including increased risk of aspiration, weak cough and abnormal respiratory patterns.\textsuperscript{50}

A proportion of MND patients have additional ‘central’ brain stem failure of the respiratory drive.\textsuperscript{6} These patients may be particularly sensitive to even small doses of benzodiazepines and opiates.

Respiratory tract infection (RTI) should be treated with early use of antibiotics.\textsuperscript{52}

Monitoring for signs and symptoms

The specialist MND team should monitor for ongoing respiratory symptoms, and the GP also has a role in this. This is important, so that timely interventions can be made to improve/maintain quality of life for as long as possible.\textsuperscript{3}

A baseline respiratory assessment and discussion of future options should be undertaken as soon as possible after diagnosis, so check whether this has been done. Deterioration can happen quickly, so an urgent respiratory referral may be needed.\textsuperscript{3}

The NICE guideline recommends that respiratory function tests, eg FVC (forced vital capacity), and/or SNIP (sniff nasal inspiratory pressure) or MIP (maximal inspiratory pressure) should be performed every two to three months by an appropriately trained professional. These tests monitor signs of respiratory impairment, rather than progression of the condition. \textsuperscript{3} SpO\textsubscript{2} may be used for those with severe bulbar involvement.

However, results of these tests may give false reassurance, so ask about the following symptoms and signs:\textsuperscript{3}
Respiratory insufficiency management

### Onward referral
Presence of one or more of these symptoms should trigger referral to the [specialist respiratory team](#), who can provide a full respiratory assessment and discuss options. A [respiratory physiotherapist](#) may also give advice on positioning, breath stacking, or assisted cough techniques to try to prevent emergency intubation.53

**MND care centres and networks** can also advise on these issues.

**Referral may also be made to:**
- an [occupational therapist](#) or [district nurse](#) for advice on the provision of appropriate equipment, such as a profiling bed
- the [specialist palliative care team](#) for advice on psychological strategies, medication and advance care planning.

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Signs</th>
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<tbody>
<tr>
<td>Breathlessness</td>
<td>Increased respiratory rate</td>
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<tr>
<td>Orthopnoea</td>
<td>Shallow breathing</td>
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<tr>
<td>Recurrent chest infections</td>
<td>Weak cough</td>
</tr>
<tr>
<td>Disturbed or non-refreshing sleep</td>
<td>Weak sniff</td>
</tr>
<tr>
<td>Nightmares</td>
<td>Abdominal paradox</td>
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<td>Daytime sleepiness</td>
<td>Use of accessory muscles of respiration</td>
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<tr>
<td>Poor concentration and/or memory</td>
<td>Reduced chest expansion on maximal inspiration</td>
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<tr>
<td>Confusion/hallucinations</td>
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<tr>
<td>Morning headaches</td>
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<td>Fatigue</td>
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<td>Loss of appetite</td>
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Assisted ventilation

Non-invasive ventilation (NIV)
This method uses a close-fitting mask over the nose and/or mouth. Acceptance and success often depends on the correct choice of mask or nasal cushion. A trial can be offered if the person’s symptoms, signs and respiratory function tests indicate a likely benefit.

Potential benefits of NIV include:
- better sleep, improved appetite, more alert during the day\(^6\)
- initial ability to use selectively, as reliance usually builds gradually\(^6\)
- increased survival\(^6\)
- portability.

Potential disadvantages of NIV include:
- less likely to be effective with significant bulbar involvement\(^6\)
- difficulties speaking or eating with a full-face mask\(^6\) (a nasal interface will also have some impact)
- increased carer burden,\(^6\) especially as the disease progresses, posing problems if the person with MND lives alone, as a carer may need to be available at all times to reposition/remove the mask
- continued risk of aspiration if control of oropharyngeal secretions is poor\(^6\)
- uncomfortable mask interface, inability to synchronise breathing to the rhythm of the ventilator, skin ulceration, claustrophobia\(^6\)
- less effective as MND progresses\(^6\)
- increasing reliance over time, which may have an adverse impact on quality of life, and present end of life withdrawal difficulties.\(^6\)

Invasive ventilation by tracheostomy
Elective tracheostomy for invasive ventilation is controversial and some units may be reluctant to offer this. Open discussion about both the advantages and disadvantages can help the patient, and their carer/family, make an informed decision.

Very occasionally, a person with MND may be invasively ventilated due to an acute episode of respiratory insufficiency, possibly due to initial presentation of MND, and diagnosed in intensive care. If so, it can be difficult to wean the person from invasive ventilation.\(^6\)
Potential benefits of invasive ventilation include:
- prolonged survival and remains effective as MND progresses
- eases respiratory symptoms, as with NIV, and may still be effective even with significant bulbar involvement
- no mask, which some patients cannot tolerate.

Potential disadvantages of invasive ventilation include:
- requires a surgical procedure, after which it may be more difficult to speak or eat
- complex discharge arrangements after surgery may mean a lengthy stay in hospital
- prolonged survival with progressive disability raises questions (e.g., inability to communicate, total dependence and possibility of dementia)
- impact on carers and families, as the patient may need 24-hour care, and specialist assistance (cost and resourcing may require a move into a nursing care home)
- the need for suction of airway secretions
- although it can be used selectively, dependency is likely to increase with disease progression, which may have an adverse impact on quality of life, and present end of life withdrawal difficulties.

Management without assisted ventilation
Not everyone will choose to have assisted ventilation, or can tolerate this intervention. As part of palliative care, symptoms such as breathlessness will need to be managed. See pages 62-63 for medications used to manage these symptoms.

Discontinuation of assisted ventilation
Someone with MND may wish to stop assisted ventilation when they become more dependent or when their symptoms reach a certain point. Stopping assisted ventilation should be discussed well in advance with the person with MND, carer and any professionals involved, preferably during initial discussions over the use of assisted ventilation and as part of advance care planning.
Discussions therefore need to include whether the person may want to make an Advance Decision to Refuse Treatment (ADRT) – see page 58.
The withdrawal process
Withdrawal of assisted ventilation should be carefully planned so that symptoms can be managed to avoid distress or discomfort. Refer to medications for palliative care on pages 62-63.

Withdrawal of ventilation can be very challenging. Palliative care professionals often have experience in managing the process and may be called upon if needed for support and information. Guidelines are available from the Association for Palliative Medicine.54

Oxygen
Respiratory muscle weakness leads to the retention of carbon dioxide (hypercapnia) rather than hypoxia. In this situation, assisted ventilation may be the most appropriate treatment, as use of oxygen can lead to further respiratory depression. However, when oxygen levels are low, oxygen may sometimes be used with caution53 – preferably under guidance of the person’s specialist team.

Although oxygen is sometimes used for palliative or end of life care with MND, opiates may be more effective for terminal dyspnoea.49 For more guidance, speak to your specialist palliative care team or a respiratory consultant with links to the neurology team.

The emergence of respiratory insufficiency should prompt discussion of the following issues:

Preparing for end of life
This symptom may raise concerns about end of life decisions and preparing for end of life, increased dependency and being unable to communicate. Offer people the opportunity to discuss or review any existing ADRT, DNACPR or Lasting Power of Attorney.3
See psychological support (page 48), palliative care (page 55) and advance care planning (page 57).

Alternative feeding
If respiratory function is deteriorating, enteral feeding may be discussed even if there are no immediate problems with swallowing or weight (see pages 31-32).44 If possible, the carer should be involved in discussions as they will share the burden of any interventions.
Information for you

• Respiratory checklist card
• Information sheet P4A – MND Just in Case kit
• Information sheet P6 – Evaluation and management of respiratory symptoms in motor neurone disease (MND)
• NICE Guideline NG42: Motor neurone disease: assessment and management (www.nice.org.uk/ng42)

Information you can share

Our resources for people affected by MND include:

• Information sheet 7A – Swallowing difficulties
• Information sheet 8A – Support for breathing problems
• Information sheet 8B – Ventilation for motor neurone disease
• Information sheet 8C – Withdrawal of ventilation with MND
• Information sheet 8E – Air travel and ventilation for MND

See page 69 for details of how to order copies or download from our website at www.mndassociation.org/publications

"MND patients face immense challenges every day, as do their families."

- A person living with MND
Pain

The disease process causing MND is not itself painful. However, MND leads to stiffness, cramps, reduced mobility and altered posture, all of which can result in pain. The person with MND may not mention their pain so it is important that healthcare professionals ask about it, both at the initial assessment stage and routinely during follow-up visits. Pain may occur at any stage of MND, including early on, with no relationship between pain intensity and length of time since diagnosis. Because it is usually a result of poor mobility, changes in posture, or reactions to changes in muscle tone, MND pain is more frequent in the limbs.

Pain significantly interferes with the quality of life for people with MND because of its impact on activity levels, mood, sleep, relationships, and general enjoyment of life. Anticipatory prescribing is crucial to enable the patient to maintain control.

Be clear that MND pain can sometimes be difficult to treat and it might not be possible to get rid of pain completely.

Physiotherapy is a key treatment strategy for pain in MND, and can prevent contractures, prevent and treat frozen shoulder, and relieve pain resulting from changes in tone or posture. Occupational therapy assessment and the intervention of a wheelchair therapist can reduce pain resulting from postural weakness, or straining weak muscles. Correct management of constipation can reduce abdominal pain.

Medication for pain

For drug dosages, refer to British National Formulary (BNF) or Palliative Care Formulary. Take into account the person’s needs and preferences and whether they may have any difficulty swallowing medication.

For joint pain: simple analgesia, eg long-acting non-steroidal anti-inflammatory drugs (NSAIDs).

For muscle cramps:

- Consider quinine as first-line treatment for muscle cramps in MND.
- If quinine is not effective, not tolerated or contraindicated, consider baclofen instead as a second-line treatment.
- If baclofen is not effective, not tolerated or contraindicated, consider tizanidine, dantrolene or gabapentin.
For muscle stiffness, spasticity or increased tone:\(^3\)
- Consider baclofen, tizanidine, dantrolene or gabapentin to treat muscle stiffness, spasticity or increased tone in people with MND.
- If these treatments are not effective, not tolerated or contraindicated, consider referral to a specialist service for treatment of severe spasticity.

Take care that the dosage of muscle relaxants is carefully adjusted to avoid increased weakness and decreased mobility.\(^{11}\) Also check whether the patient is taking a statin, as muscle soreness and weakness may be side effects.

Opiates (morphine, buprenorphine or fentanyl patches) may be used for pain relief and can also be used for symptomatic treatment of dyspnoea and coughing.\(^{11}\) With careful titration, excessive drowsiness and respiratory depression can be avoided.

For neuropathic pain: tricyclics or gabapentin/pregabalin.\(^{59}\)

Skin sensitivity
- Good skin and pressure care is vital.\(^{60}\) Someone with MND may be aware when they need to turn or move, but may need help to adjust their position. This must be done with extra care.
- Consider equipment for skin sensitivity relief, such as a bed cradle to relieve the weight of bed clothes, lightweight bed clothing and a pressure-relieving mattress and cushions.
- Advise warm socks for cold feet.

Oedema (fluid retention)
- This may largely be related to restricted activity and posture or to an underlying health condition, which should be treated accordingly.\(^{61}\)
- Attention to posture and seating requires regular assessment by an occupational therapist.
- Compression support stockings, effleurage (light massage) and reflexology may be beneficial.
- Diuretics are rarely helpful as they can promote urinary urgency and electrolyte disturbance.\(^{62}\)
- In some areas, referral to the lymphodema service may be an option.
Cognitive change

There is now increased awareness of cognitive and behavioural changes in MND. People with MND may fall into one of the following groups:

• those unaffected by cognitive change - about 50%; this decreases to 20% in the final stage of the disease\textsuperscript{63}

• those with mild cognitive change, with deficits in executive functions, language, behaviour and/or social cognition - about 35\%\textsuperscript{64, 65, 66}

• those with frontotemporal dementia (FTD), either at the same time or after diagnosis of MND - up to 15\%.\textsuperscript{67}

Up to 15\% of those presenting with FTD go on to develop motor impairments, with MND diagnosed after their dementia.

Cognitive impairment and/or behavioural change in MND

People with MND may experience cognitive and/or behavioural changes not classified as dementia, ranging from mild to moderate or more severe in some cases.\textsuperscript{68}

Cognitive impairment may manifest as a deficit in verbal fluency. Some people may have primary language problems that manifest as reduced verbal output and failure to initiate conversations. The person may respond in short phrases or single words.\textsuperscript{68}

People may experience problems with:

• learning new tasks, including the use of equipment (such as gastrostomy or NIV)

• impulsivity, which can include repetitive or persistent actions

• a preference for sweet foods and a tendency to cram

• inappropriate social behaviour

• lack of awareness or insight into their problems or their impact

• managing affairs/finances, making decisions, planning for the future

• short term memory loss

• poor concentration

• literacy and language.

Not all of the above may be evident and they can vary in degree. Changes may be subtle and masked by movement and speech problems. Some may be part of the normal ageing process.
**Behavioural changes**

Behavioural impairment is a recognised feature of MND and changes may include egocentric or selfish behaviour, loss of interest, apathy, disinhibition, irritability, aggression and absence of insight into these changes.\(^6\) These behaviours may meet some of the criteria for frontotemporal dementia.\(^6\)

People with MND may also find it difficult to recognise emotions in others and have altered sensitivity to social and emotional cues.\(^6\)

Some people with MND will experience both cognitive change and behavioural impairment, but it is important to note that people with no cognitive changes can have profound behavioural abnormalities.\(^6\)

It is unlikely that someone experiencing these changes will know this is happening or can acknowledge the change. More often, it is the family or carer who sees behavioural change, and this can be very challenging for them. Behavioural change can be difficult for interaction with carers and increases the burden on carers.\(^6\)

Carers should be offered appropriate support, and this may require a conversation in confidence, away from the person they care for.

**Diagnosis of cognitive impairment**

Use of the Edinburgh Cognitive and Behavioural ALS screening tool (ECAS tool – [http://hdl.handle.net/1842/6592](http://hdl.handle.net/1842/6592)) can help determine whether someone may be experiencing cognitive change. The results should be interpreted with the help of a psychologist. Referral should be made to a clinical neuropsychologist for a full neuropsychological assessment and advice on management.

**Ventilatory failure developed during the course of MND can exaggerate cognitive symptoms.**

Non-invasive ventilation has been shown to improve changes in concentration, memory and confused thinking that can result from raised \(\text{CO}_2\) levels.\(^3\) Respiratory impairment should be excluded as a cause of cognitive changes.
Frontotemporal dementia (MND-FTD)
The following explanation defines MND-FTD and the combination of symptoms that may indicate frontotemporal dementia.  

Cognitive change in MND-FTD
- marked executive dysfunction (difficulties with higher order cognitive abilities, planning, decision making, problem solving and poor attention span)
- language changes in some cases; marked aphasia will be mostly expressive (problems communicating what they are trying to say) or sometimes also receptive (problems understanding what is communicated to them)
- memory difficulties; this isn’t a primary symptom, but someone may experience difficulties due to executive dysfunction and ‘taking in’ information, rather than the memory problems associated with Alzheimer’s disease, for example
- visuospatial functions are preserved.

Behaviour change in MND-FTD
- significant personality change
- disinhibition and impulsivity (socially inappropriate behaviour)
- perseveration (continuing to conduct an activity that is no longer appropriate)
- change in eating behaviour (sweet food preference)
- loss of emotional understanding (appearing egocentric/selfish)
- being withdrawn (apathy/failure to initiate)
- stereotyped/ritualistic behaviour
- behaviour change often occurs before cognitive impairment.

One day, on the verge of a breakdown, I called the mental health team telling them I could no longer cope.

- A carer of person living with FTD and MND
Managing cognitive change/MND-FTD

It is important to acknowledge to patients and carers that cognitive change may be part of the clinical picture of MND, which will help them prepare. For advice on managing cognitive change or MND-FTD, speak to local psychological support services.

Your role as a professional

A lack of recognition of this issue by health and social care professionals can lead to extra stress for people close to someone with MND. Learning to recognise the symptoms of cognitive change can enable earlier referral for support.

It is important that people affected by MND have consistency in, and from, the health and social care professionals they see. This can help reduce stress from repeatedly having to explain the issue and its effects. The professionals involved can also have insight into, and ability to monitor, which issues are caused by muscle weakness and which by cognitive change.

Impact on decision-making

Care needs to be taken to ensure informed consent during decision-making. People with MND and FTD may lack mental capacity. Capacity should be assessed where FTD is evident and care provided in line with the Mental Capacity Act 2005.\(^3\)\(^{,68}\)

Ability to drive

Although there may be no physical barrier to driving in early MND, lack of insight into danger is sometimes an issue for a small minority, which may affect their ability to continue to drive safely.\(^{69}\)

**Information for you**

*Cognitive change, frontotemporal dementia and MND booklet*

**Information you can share**

Our resources for people affected by MND include:

Information sheet 9A - *Will the way I think be affected?*

Information sheet 9B - *How do I support someone if the way they think is affected?*

Information sheet 9C - *Managing emotions.*

See page 69 for details of how to order copies or download from our website at [www.mndassociation.org/publications](http://www.mndassociation.org/publications)
Psychological support

Psychological well-being is an important determinant of quality of life. MND has a significant psychological as well as physical impact on people living with MND and those close to them, including children.  

MND is characterised by a series of losses, with associated grief and bereavement. Feelings of anxiety, depression and isolation are common, particularly in the first year after diagnosis.  

A feeling of hopelessness, which is more likely to arise soon after diagnosis, is the most common marker of psychological morbidity in MND.  

The psychological experience of MND will be influenced by age, gender, stage of life, cultural and educational background, spiritual and religious beliefs, and psychosocial circumstances.  

Excellent psychological support is essential to holistic, patient-centred care. It can greatly influence quality of life, reduce risk of anxiety and depression, diminish caregiver burden and deliver significant savings for the healthcare service.  

Before the diagnosis  
Anxiety and low mood may be triggered by:  
• onset of worrying symptoms and problems in identifying their cause  
• a long period of investigation and the specialist’s need to be sure before giving the diagnosis.

After diagnosis is confirmed  
• give sufficient information to allow the person with MND to understand what is happening without overwhelming them  
• provide space for them, their families and carers to express thoughts and feelings openly and without judgement  
• give information that can help them maintain control and make plans for the future
• feelings about the illness may change; allowing time to express thoughts, feelings and concerns regularly can help identify information and support needs
• check preferences for involving family members/carers; offer opportunities for the person with MND, and their partner if relevant, to rehearse how they might tell other family members, particularly children, about the diagnosis and prognosis
• be aware that the type, quantity and timing of information and support required may be different for each individual, for example carers may seek out information earlier, to help them prepare for their role
• determine what the person with MND has understood and retained of the information provided
• where possible, direct them to a specialist psychologist or someone from the local mental health team to provide ongoing support and information; signpost to other sources of support including counselling services, a local hospice, local support groups, the MND Association website and online forums.

See page 69 for details of the MND Association’s care information.

**Support as the disease progresses**

As MND develops, its impact will be influenced not only by how quickly it progresses, but also the area of the body involved. Adapting to the changes in physical function can be particularly challenging for patients with a more rapid rate of progression of the disease as there is less time available for psychological adjustment.

• **Ensure people with MND and their families understand the likelihood and implications of respiratory problems.** This enables everyone to explore the options and likely outcome, thus avoiding uninformed last-minute decisions, eg prolongation of unacceptable quality of life.

• **Consider the psycho-social effects of communication problems and not being able to share thoughts and feelings.**

• **Consider having conversations with an emotional/psychological content from the outset.**
Earliest possible referral to the **palliative care team** is recommended, as they can help provide support on a range of issues, from symptom management to psychological and spiritual support.

We produce resources on end of life planning, both for people affected by MND and health and social care professionals (see page 61).

**Emotional reactions**

Fears may include:
- inability to move and/or communicate
- choking or ‘fighting for breath’
- loss of independence and dignity
- feeling isolated and alone
- increasing dependency and becoming a burden
- inability to cope and loss of control
- the unknown, death and the process of dying.

Allowing time and opportunities to acknowledge and discuss these concerns and fears may help alleviate some of them.

**Denial**

Denial may form part of a coping strategy for some people. Early on this may be a useful means of adapting to difficult circumstances, but if denial persists it can contribute to higher levels of anxiety and depression.\(^7\)

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*I was told I might have a year left, or I might have three. So much was going through my head. I'm just getting round to accepting my MND now, but it's taken nearly a year.*

- A person living with MND

**Hopelessness**

Feelings of hopelessness are common, especially soon after diagnosis and may be linked to the recognition that many of life's expectations will never be realised.\(^7\) Time and a safe environment are required to explore these issues.
Anger
Many people with MND are concerned about how angry they feel as a result of frustration and loss of control. Anger may be displaced onto others and may occur at any stage of the illness. Acknowledging and discussing such feelings may be helpful. If anger is existential in nature, spiritual support may be appropriate if the family has a particular faith or belief system.

Anxiety and depression
Persistent and disabling symptoms of anxiety and depression need to be distinguished from appropriate responses to the difficulties faced by people living with MND. The symptoms of anxiety and depression may mimic some of the physical consequences of MND making identifying them more challenging.

These include sleep disruption, diminished appetite, fatigue or weakness, poor concentration and irritability. Screening for anxiety and depression, using tools such as the hospital anxiety and depression score (HADS), may assist in identifying and/or monitoring symptoms in both patients and carers.

Antidepressants and/or anxiolytics may be beneficial for some people. These include selective serotonin reuptake inhibitors (SSRIs) such as sertraline and citalopram. These may take several weeks to have an effect.

Apathy
This response has been widely reported in some people living with MND. It may be linked to anxiety and depression, or to cognitive change, and may be a barrier to accessing help and support.

Sleep disturbances
Treatment of anxiety and depression can often result in quite rapid improvements in sleep quality. This in turn can improve both physical and psychological well-being. Sleep disruption due to other factors (including pain, cramps, spasm, breathing difficulties and choking) can have a negative impact on quality of life and should also be treated.
**Emotional lability (pseudobulbar affect)**

This may occur as a consequence of upper motor neurone involvement in the corticobulbar tracts in patients with MND, giving rise to a “pseudobulbar” presentation. It may or may not be associated with more widespread cognitive decline\(^7\)\(^9\) and may also be a manifestation of depression (with or without anxiety).

Emotional lability presents as disturbed emotional responses, including inappropriate and uncontrollable laughter and crying.\(^9\) In some cases, this can be extreme. It can be very disabling for patients and difficult for those around them. Some people find understanding the symptom helps them to manage the impact, but others find it limits where they go and what they do.

**Impact on professionals**

MND poses many challenges for professionals who may experience feelings of frustration, powerlessness, inadequacy and sadness. It highlights attitudes to issues related to disability, quality of life and measures taken to prolong life.

Good multidisciplinary teamwork is necessary to provide support and opportunities to discuss concerns and responses to difficult situations.

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*We have what we call time out mornings where we talk about patients that have died, what went well and what could have gone better, difficulties people have had or are having. It’s a great way we can offer each other support.*

- MND specialist nurse
Providing evidence for benefit applications (DS1500)

People with MND may apply for disability/employment benefits.

If someone with MND is applying for, or being reassessed for, Attendance Allowance, Personal Independence Payment (including transfer from Disability Living Allowance), Employment and Support Allowance or Universal Credit, their application may be considered for fast-tracking under provisions called ‘Special Rules for Terminal Illness’.

These rules make the application process less burdensome and speed up access to the highest level of financial support.

GPs can help by completing form DS1500 to show the person with MND is eligible for these rules.

If a person with MND is aware that Special Rules exist, they can usually start the process during their initial phone call by telling the call handler that they want their application to be considered as a Special Rules case.

Once they have made it clear they wish to claim under Special Rules, they will be asked whether they have, or are going to get, a DS1500 form, which they will be asked to send in. This can be completed by a GP.

The claim will then be processed by a Department for Work and Pensions (DWP) assessment provider and a decision made by the DWP.

The initial application process for Universal Credit is online in most cases.

It is possible for a DS1500 form to be completed on someone’s behalf without their knowledge. This can be helpful if your patient is not up to dealing with the terminal nature of their condition, if they cannot handle the claims process, or if they have not been told the full nature of their condition.

In England, DS1500s can also be submitted electronically for PIP applications.
How MND qualifies for the Special Rules

In order to satisfy the special rules provisions fully, the person must be terminally ill. Legislation defines that: ‘a person is “terminally ill” at any time if at that time the person suffers from a progressive disease and the person’s death in consequence of that disease can reasonably be expected within six months.’

A third of people with MND die within a year of diagnosis and more than half die within two years. As a result, people with MND will pass the diagnosis element of Special Rules on the DS1500 form. However, the prognosis element can be problematic.

It is important to remember that the course of MND is unpredictable. The DS1500 form only asks if there is a reasonable expectation that their prognosis could be six months. It does not mean that someone is going to die within six months, only that it is a possibility.

The MND Association strongly recommends that a Special Rules application using form DS1500 should be actively considered in all cases involving MND. There may be an exception where MND presents with noticeably slow progression or limited symptoms.

As the six month prognosis is a guideline only, there is no sanction on any health or social care professional who has declared someone terminally ill, should that person live beyond the six month period.

Information for you

Information sheet P5: Providing medical evidence for benefit applications made by people with MND.

Information you can share

Information sheet 10A - Benefits and entitlements

See page 69 for details of how to order copies or download from our website at www.mndassociation.org/publications

Our Benefits Advice Service provides advice for people with MND by phone and e-mail in England, Wales and Northern Ireland.

England and Wales: email mnd@cacv.org.uk or call 0808 801 0620. Northern Ireland: email welfarechanges@adviceni.net or call 0808 802 0020.
People with MND have particularly complex care and support needs in the final stages of life. Because MND can be rapidly progressive, it is important that people with the condition are offered referral to specialist palliative care services at an appropriate time.

The appropriate time for referral will vary, depending on factors such as the availability of services and the speed of progression of symptoms. Establishing early links with palliative care and specialist services can provide a useful source of support. However, introducing the concept of palliative care can present some difficulties, because of the stigma and lack of understanding around what palliative care is and what it can provide.

**Key points**

- Explain the role of the palliative care team and the potential benefits.
- Introduce the service via day care, respite or complementary therapies.
- If palliative care includes referral to a hospice, stress that this doesn’t mean that death is imminent, as many people view hospices purely as places where people die.
- Discussing advance care planning (see page 57) ensures the patient’s wishes are respected at each stage of the disease.
- All people with MND should be placed on local palliative care registers/lists/co-ordination systems, where these exist – eg the Gold Standards Framework, or Electronic Palliative Care Co-ordination Systems (EPaCCS). This will ensure that future care can be planned and co-ordinated, and that guidance and support are given to both the patient and the GP.
Advance care planning

Talking about end of life
People with MND will have many concerns about the progression of the condition and may not express their fears openly.

Time is needed for the person to feel comfortable enough to talk about their worries, and it is important that concerns are taken seriously and solutions, where possible, are made available.

People may have clear views about how they want the latter stages of their illness managed.

Discussions around options for care and preferences for end of life should be held before the need is urgent or the capacity to communicate is limited and tiring.

The NICE guideline on MND\(^3\) recommends offering opportunities to discuss preferences and concerns about care at the end of life at certain trigger points, such as:

- at diagnosis
- if there is a significant change in respiratory function
- when discussing gastrostomy or non-invasive ventilation.

Be sensitive about the timing of discussions and take into account the person’s communication ability, cognitive status and mental capacity.\(^3\)

Having time to think things through and knowing that wishes have been recorded gives many people peace of mind.

Recording discussions
If discussions are held and decisions are made, they should be clearly documented and communicated to relevant health and social care professionals. Ideally this should be brought together in a personalised advance care plan.
**Advance care plan**

An advance care plan can be wide-ranging, covering all aspects of day-to-day care including, as examples, who should provide personal care and how they should do it, special requirements for food and drink, decisions about artificial feeding, ventilation, resuscitation, use of antibiotics, place of care and preferred place of death, and other issues such as care of dependants or pets in an emergency.

Also included in this may be leaving special messages for friends and loved ones, memory boxes or books for children, and digital legacies.

The process for advance care planning, the documentation used and who needs to have this documentation will vary between areas. Many places will have end of life care co-ordinators or facilitators.

**Advance statement**

This is a written statement of a person’s preferences, wishes, beliefs and values for future management, medical choices and care, and may include where the person would like to be cared for and where they want to die. It is designed to guide anyone who might have to make treatment and management decisions if the person has lost the capacity to make or communicate their own decisions. Advance statements may be included within an advance care plan, but can also stand alone.

Even though advance care plans and advance statements are not legally binding, they still give guidance on decisions. Health professionals should take the recorded preferences into account as part of an overall judgement of best interests, and they should be honoured where possible.

**Advance Decision to Refuse Treatment**

An Advance Decision to Refuse Treatment (ADRT) allows people to make decisions to refuse treatments or have treatment withdrawn, including treatments that sustain life, such as assisted ventilation, in specific circumstances in the future.

Everyone involved in the care of a person with MND should have access to the current ADRT. Emergency staff should be able to access it easily.

When completed correctly, an ADRT is legally binding in England and Wales.
Do Not Attempt CPR (DNACPR)
Someone with MND may choose to have a DNACPR. This will be respected in most instances, but is not legally binding.
If included in an ADRT, with clear instructions about when it should be applied, refusal of CPR is legally binding in England and Wales.

Power of Attorney
A Lasting Power of Attorney (LPA – England and Wales) or an Enduring Power of Attorney (EPA – Northern Ireland) is a legal document that allows a trusted person to make decisions on another person’s behalf. An LPA can cover decisions about financial and/or specified health/care related matters, while an EPA covers financial affairs and property but doesn’t cover health or care decisions.
The person with MND must have capacity to understand and make the decisions when they complete the LPA or EPA. It will not normally be used until they do not have capacity or cannot communicate their own decisions.83, 84

Reviewing choices
Any decisions made about advance care should be regularly reviewed, to reflect that people do change their minds. This can be difficult when someone’s condition advances, as problems with communication can make it more problematic to identify any changes in wishes. If changes are made, new copies should be signed and dated and distributed to those who hold the existing paperwork.

“I’ve been lucky. I had a very supportive team who I could discuss advance decisions with, but even this was hard emotionally. Now that I’ve done it I feel better.”
- A person living with MND
Advance care planning checklist

☐ Is someone helping the person with MND to develop an advance care plan?

☐ Has an Advance Decision to Refuse Treatment (ADRT) been recorded?

☐ Has any existing advance care plan or ADRT been reviewed?

☐ Does a palliative care physician need to be involved?

☐ Has the GP or another professional discussed the situation with the person and family, so they are prepared for what is likely to happen?

☐ Has the person with MND and their family been reassured that death by choking or fighting for breath is not the norm?

☐ Are appropriate medications available in the home, to prevent a crisis admission (end of life management of symptomatic breathlessness following local palliative care guidelines)?

See page 35 for information about the MND Just in Case kit.

☐ Is there adequate physical and emotional support for the family if the person wishes to die at home?

☐ Is the ambulance service aware of any signed DNACPR form, or an ADRT?

☐ Have you informed your local primary care out-of-hours service of any DNACPR or ADRT?

☐ Does the district or community nurse know there is someone with MND in their area?
Useful resources

The MND Association’s publication *End of life - a guide for people with motor neurone disease (MND)* contains comprehensive information about this topic for people affected by MND, and may support you with these conversations. The guide covers subjects including:

- how MND progresses
- the benefits of planning ahead
- how to manage end of life discussions with health and social care professionals, family, children and friends
- what is likely to happen at end of life with MND
- advance care planning, advance decisions and the introduction and withdrawal of treatments for MND
- how to put personal affairs in order and what to do when someone dies
- an overview of the law regarding suicide and assisted dying
- the rules around organ and tissue donation with MND.

We also have an ADRT information sheet for people with MND, along with a booklet for health and social care professionals: *A professional’s guide to end of life care in motor neurone disease (MND)*.

Call our MND Connect helpline on 0808 802 6262 for further support and to order a copy of these resources, or download them from [www.mndassociation.org/publications](http://www.mndassociation.org/publications)

In 2012 the National Council for Palliative Care (NCPC) produced a booklet with the support of the MND Association, called *Difficult Conversations: making it easier to talk about the end of life with people affected by motor neurone disease*.

NCPC has since merged with Hospice UK, but their information can still be found at [www.ncpc.org.uk](http://www.ncpc.org.uk)
The most common cause of death in MND is respiratory failure, often with additional chest infection.\textsuperscript{6}

Death in the majority of people with MND is peaceful, following lengthening periods of sleepiness, gradually resulting in unconsciousness and death. Very occasionally, this process may occur suddenly.\textsuperscript{15}

It is very important to reassure people with MND and carers that death from choking is exceptional.\textsuperscript{1}

**Medication at end of life**

Check all symptom control:

- pain – patients may experience severe discomfort and/or distress, particularly in the later stages. Use of opiates may be an option with careful titration to avoid excessive drowsiness and respiratory depression\textsuperscript{85}
- pressure care
- dyspnoea
- dysphagia
- excess oral secretions or dry mouth
- insomnia
- anxiety/depression
- restlessness/agitation
- bowels and bladder
- reassess emotional and practical needs of the carer and family.

The GP, specialist or other appropriate prescriber should consider anticipatory prescribing of a range of medications to address worsening symptoms, including:

- antimuscarinics, such as hyoscine hydrobromide and glycopyrrolate, to reduce saliva and lung secretions\textsuperscript{49}
• benzodiazepines, such as midazolam, to manage breathlessness that is exacerbated by anxiety, and other medications to reduce anxiety/terminal restlessness, such as haloperidol or levomepromazine.\(^3,49,86\)

• opioid analgesics to reduce cough reflex, relieve dyspnoea and therefore fear and anxiety. They can also control pain.\(^11\)

• anti-emetics for nausea.\(^49\)

Regular analgesics should usually be continued until death, even if oral medication is no longer possible because of dysphagia. Alternatives, such as suppositories or parenteral routes should be considered. Parenteral medication may be given as a continuous subcutaneous infusion using a syringe pump.\(^49\)

For drug dosages, refer to British National Formulary (BNF) or Palliative Care Formulary. Take into account the person’s needs and preferences and whether they may have any difficulty swallowing medication.

**Communicating with the person with MND**

This may become extremely difficult as the person reaches end of life, but even if they are unresponsive, every attempt should be made to maintain communication, particularly if assessing mental capacity and ability to make decisions. Eye movements, single response answers to closed questions, picture/alphabet boards or other communication aids may be used as appropriate (see pages 23-24).\(^34\)

**Supporting family and carers at this stage**

MND is unpredictable and may progress rapidly, with death occurring more quickly than anticipated.\(^15\) Carers and family members, including children and young people, will need practical and emotional support (see next section). Care plans and information must be shared by all members of the care team and adequate nursing cover maintained.

Early and sustained good symptom control is essential in the management of a peaceful and dignified death. It is important to avoid the implication that the family or carer are making the decision to end the life of the person with MND. Ideally the person with the disease should remain in control of end of life issues as much as possible. Taking time to discuss end of life with the family as early as possible can be positive, as it allows them to look at their lives together, achieve things that are important to them and tie up loose ends.
Supporting families and carers

Many people who are providing unpaid care for someone with MND will not recognise themselves as a carer. They may not be aware that, if they are identified as a carer, they will be able to access specific support and apply for particular benefits.

It is therefore crucial that GPs identify unpaid carers and young carers, so appropriate support and services can be offered. Carers can experience pressures on emotional and physical health, as well as financial pressures. It may not be possible for you to provide solutions to every problem, but much can be achieved by listening to carers and their experiences.

Carers of people with MND need advice, support and information to help them make sense of what is happening, to enable them to continue in their caring role and to help them plan for the future. Good quality and timely care and support can have a positive effect on emotional wellbeing and can prevent a crisis situation.

Not everyone will have a carer

Not every person with MND will have family or friends to support them. These people may have additional practical and emotional needs that cannot be provided without external support.

Where a relationship was poor before the MND diagnosis, it can sometimes improve, but it may also get worse. Professionals should be aware that, in some cases, this can lead to a relationship breakdown. The person may need external care if family or friends cannot help and should be referred as promptly as possible to social care services.

Psychological support for carers and family members

The carers and families of those living with MND often experience considerable psychological and emotional distress. Assumptions, hopes, plans and expectations for the future may have to be reviewed. Caring for someone with MND can be physically and emotionally challenging and often leads to feelings of isolation.
Allow carers time to talk about the impact MND has on them and their family. It may be more appropriate to talk to the carer separately from the person with MND, so they can be more open about their needs.

Let them express and explore their feelings. By asking questions, you will find out how safe it is to talk about particular issues. A carer may not discuss these feelings and fears spontaneously.

Reassure the carer that it is natural to have intense and conflicting emotions about the situation they are in.

MND involves many losses: carers need time to grieve for the past and how things were, the present situation and their loss of a future with the person they care for.

Discuss with the carer/family what type and frequency of support would help reduce anxiety and fear. Consider referral for additional psychological support. There may be phases of the illness where this needs to be reviewed, particularly when significant changes happen.

Carers may benefit from being in contact with other carers of people with MND. The MND Association can sometimes provide a volunteer ‘Association visitor’. Volunteer-led MND branches and groups run local support meetings for carers and families. It might help to talk to other carers through our online forum at www.mndassociation.org

Children and young people in the family may benefit from a referral to a specialist service such as School Nurse or Child & Adolescent Mental Health Service (CAMHS) in order to access a range of appropriate therapies, such as emotional or psychological support or practical help with their education.

**MND forces changes in roles and relationships. Consider:**

- mapping the social support system of the family using a genogram
- meeting the various information needs of all family members
- counteracting isolation of individuals and promoting awareness of each other’s needs
- offering early opportunities for short periods of respite to prevent over-dependence on a single carer
- other concurrent life events, such as moving house, children leaving home, financial difficulties etc and linking the family with supportive services if appropriate
• exploring the way in which illness changes relationships and acknowledge not only the strain of this, but also the ways in which they stay the same or are strengthened

• creating opportunities for people to express negative feelings without feeling guilty

• how carers may experience exhaustion from the caring role, exacerbated by powerlessness to prevent suffering and further deterioration.

**Carer’s assessments**

Carers who provide a substantial amount of care on a regular basis have a legal right to an assessment of their needs, carried out by their local social services or health and social care trust.

The assessment should anticipate need, so that crisis points can be avoided. The results of an assessment must be outcome-based and reviewed on a regular basis.

**Respite**

Carers of people living with MND may need regular, planned breaks from caring. As a key feature of the disease is the speed of progression, many carers find themselves continually trying to meet unrelenting demands. Without breaks, carers will be less able to perform their caring role and their own health will suffer.

Respite may involve care at home, in a day centre or hospice day centre, or a residential setting. It can allow the carer to go out and retain social contact.

**Young carers**

Young people who are caring for someone with MND are also entitled to a young carer’s needs assessment. You may need to liaise with their school to ensure they are receiving the support they need.

An Early Help Assessment (EHA) for children who are not providing care within the family will enable them to access targeted support, including emotional support.

**Bereavement**

Family members, including children, may benefit from ongoing bereavement support. Consider referring to **palliative care services** or local bereavement organisations.
Action points for GPs

• Make sure that a person’s role as a carer is noted on the record of the person with MND, and on their own GP practice record (if relevant).

• Has the person had a carer’s assessment? Has a young carer had an Early Help Assessment or a young carer’s needs assessment?

• Does the carer have the support and knowledge they need to carry out a caring role?

• Has a referral been made to an appropriate specialist agency such as school nurse or CAMHS, to support any children or young people?

• Have the needs of the whole family been considered?

Information for you

Booklet PX031 Supporting children and young people close to someone with MND.

Information for young people

The MND Association has two resources for young people:

So what is MND anyway? is a booklet about MND for young people aged 11-18. It includes a section on being a young carer.

When someone close has MND is an interactive workbook for 4-10 year olds, which enables a trusted adult to communicate with a young child about MND in an age appropriate way.

Other information you can share

Our resources for people affected by MND include:

Caring and MND: quick guide
Caring and MND: support for you
Finding your way with bereavement
Information sheet 4A – Communicating about MND with children and young people
Information sheet 10G – Support for families with children

See page 69 for details of how to order copies or download from our website at www.mndassociation.org/publications
How the MND Association can support you

We support health and social care professionals to provide the best possible care for people living with MND, their carers and families. We do this in a number of ways:

**MND Connect**
Our helpline offers help, information and support, and signposting to people living with MND, carers, family and health and social care professionals.

Email: mndconnect@mndassociation.org
Phone: 0808 802 6262

**Information resources**
We produce high quality information resources people living with MND, carers, family members and health and social care professionals.

www.mndassociation.org/publications

**MND Association website**
We have a wide range of information to support health and social care professionals working with people affected by MND.

www.mndassociation.org/professionals

**Education**
Our education programme is designed to improve standards of care and quality of life for people living with and affected by MND. Opportunities include online modules and face-to-face training.

www.mndassociation.org/education

**Support grants and equipment loan**
Where statutory provision is not available, we may be able to offer a support grant or loan equipment.

www.mndassociation.org/getting-support
Research into MND
We fund and promote research that leads to new understanding and treatment and brings us closer to a cure.
www.mndassociation.org/research

MND register
The MND Register of England, Wales and Northern Ireland aims to collect information about every person living with MND to help plan the care and discover more about the cause of the disease.
www.mndregister.ac.uk

Regional staff
We have a network of regional staff with specialist knowledge of MND. They work closely with local statutory services and community care providers. Contact MND Connect for further information.
Email: mndconnect@mndassociation.org
Phone: 0808 802 6262

MND care centres and networks
We fund and develop care centres and networks across England, Wales, and Northern Ireland, which offer specialist MND multidisciplinary care.
www.mndassociation.org/care-centres

Branches and groups
We have volunteer-led branches and groups nationwide providing local support and practical help to people with MND and their carers.
www.mndassociation.org/branchesandgroups

Association visitors (AVs)
AVs are trained volunteers who provide one-to-one local support to people affected by MND. They can support people affected by MND in person, by telephone or by email or through support groups.
www.mndassociation.org/associationvisitors
Visit www.mndassociation.org/usefulorgs for other useful sites.

**Bereavement support**

**Cruse Bereavement Care**
Bereavement support charity. [www.cruse.org.uk](http://www.cruse.org.uk)

**Winston’s Wish**
Charity for bereaved children. [www.winstonswish.org.uk](http://www.winstonswish.org.uk)

**Cognitive change**

**The Frontotemporal Dementia Support Group**
For carers who are coping with behavioural changes in a partner, family member or friend as a result of frontotemporal dementia. [www.ftdsg.org](http://www.ftdsg.org)

**The familial Frontotemporal Dementia Support Group (fFTDSG)**
Part of the same group, provides information, advice and social opportunities for people affected by inherited forms of FTD. [www.ftdsupport.org](http://www.ftdsupport.org)

**Palliative care**

**National Council for Palliative Care**

**Palliative Care Formulary**
Includes information on administration of drugs via feeding tubes. [www.palliativedrugs.com](http://www.palliativedrugs.com)
Palliative Care Matters
A website for healthcare professionals working in palliative care. www.pallcare.info

Cicely Saunders Institute of Palliative Care, Policy and Rehabilitation
Research and resources on palliative care. www.kcl.ac.uk/palliative

Standards of care
Gold Standards Framework
Enables a ‘gold standard’ of care for those nearing the end of life. www.goldstandardsframework.org.uk
References

20 Sheffield MND Care and Research Centre. What’s the difference between MND and ALS? www.sheffieldmndcentre.group.shef.ac.uk/differencebetweenmndals.html Accessed September 2018.


We value your feedback

Your feedback helps improve our information for the benefit of people living with MND and those who care for them.

Visit [www.smartsurvey.co.uk/s/mndprofessionals](http://www.smartsurvey.co.uk/s/mndprofessionals) or email your comments to [infofeedback@mndassociation.org](mailto:infofeedback@mndassociation.org)

If you would like to help us by reviewing future versions of our resources, please email [infofeedback@mndassociation.org](mailto:infofeedback@mndassociation.org)

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80 SEALS Registry (for background information on SEALS see *Neuroepidemiology*. 2007; 29:44-8).


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About MND

• MND is a fatal, rapidly progressing disease that affects the brain and spinal cord.
• It attacks the nerves that control movement so muscles no longer work.
• It can leave people locked in a failing body, unable to move, talk and eventually breathe.
• It affects people from all communities.
• Some people may experience changes in thinking and behaviour, with some experiencing a rare form of dementia.
• MND kills a third of people within a year and more than half within two years of diagnosis.
• A person’s lifetime risk of developing MND is 1 in 300.
• Six people per day are diagnosed with MND in the UK.
• MND kills six people per day in the UK.
• It has no cure.