Motor neurone disease:
a guide for GPs and primary care teams
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 GPs and others in the primary care team may come across few people living with MND during their professional lifetimes. However, they 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 has been 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 to access the RCGP online module in 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
- local support and advice from our staff and volunteers.

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

See page 69 for more information about how the MND Association can support you in your role.
## Quick reference

### Responsibilities when MND is suspected or confirmed

<table>
<thead>
<tr>
<th><strong>Prompt referral for diagnosis</strong> (page 13)</th>
</tr>
</thead>
<tbody>
<tr>
<td>• use the Red Flag tool to recognise early signs of MND in order to refer to <strong>neurology</strong> in a timely manner.</td>
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<thead>
<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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<td>• refer to appropriate specialist teams as necessary.</td>
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<tr>
<th><strong>Monitoring symptoms</strong></th>
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<tr>
<td>• monitor and assess symptoms, including signs of:</td>
</tr>
<tr>
<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>
</tr>
<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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<tr>
<th><strong>Support and information</strong></th>
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<tr>
<td>• provide support and information before diagnosis and throughout the course of the disease</td>
</tr>
<tr>
<td>• advise on the need to inform the DVLA or DVA of MND diagnosis</td>
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<tr>
<td>• complete a DS1500 form to support a benefit application (page 53).</td>
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</tbody>
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<table>
<thead>
<tr>
<th><strong>Repeat prescriptions for riluzole</strong> (page 20)</th>
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<tbody>
<tr>
<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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<tr>
<th><strong>Palliative care</strong> (page 55)</th>
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<tbody>
<tr>
<td>• include the person with MND on local palliative care registers/lists/co-ordination systems, where these exist – eg the Gold Standards Framework, Electronic Palliative Care Co-ordination Systems (EPaCCS).</td>
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<tr>
<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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</table>
The role of the GP and the primary care team

The GP and primary care team are 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 where 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 currently 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 facility 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 healthcare 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>
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<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) respiratory posture and positioning exercise</td>
</tr>
<tr>
<td>Respiratory team</td>
<td>coughing, breathlessness at rest or 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 aids/equipment for activities of daily living positioning</td>
</tr>
<tr>
<td>Speech and language therapist</td>
<td>swallowing and diet modification positioning speech and communication communication aids and equipment coughing</td>
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<tr>
<td>Dietitian</td>
<td>weight loss lack of appetite diet modification gastrostomy</td>
</tr>
<tr>
<td>Palliative care services/ hospice</td>
<td>support for individual and family uncontrolled pain breathlessness fear/anxiety/depression</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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## Actions for the GP and the primary care team

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

Three things I wish my GP had known about MND:
- the right referral process for diagnosis
- to learn about the condition from the patient and family
- that time is of the essence.”

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) lesions, 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, speed and pattern of progression can be very variable. Not all symptoms necessarily happen to everyone, nor do they develop in the same order or progress at the same rate.³

**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
• cramps
• fasciculations
• change in voice quality, such as quiet or slurred speech
• awareness of swallowing changes, such as having to ‘double swallow’
• muscle wasting⁴

First symptoms may progress to:

• loss of function of limbs

• weakness and wasting of muscles of the trunk and neck

• 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.\textsuperscript{4,6}

Some people with MND may experience cognitive changes, ranging from mild effects to noticeable impairment. Some will experience frontotemporal dementia (MND-FTD).\textsuperscript{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).\textsuperscript{4}

**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.\textsuperscript{4}

**Additional symptoms**

- fatigue – this is common throughout the disease and can be severe, affecting remaining functional ability\textsuperscript{7}
- saliva clearance, tenacious saliva or mucus (see page 29)\textsuperscript{8}
- emotional lability – inappropriate or excessive crying or laughter (see page 51)\textsuperscript{9}
- anxiety and depression (see page 50)\textsuperscript{10}
- pain and discomfort from muscle cramps or spasticity (see page 42)\textsuperscript{11}
• insomnia\textsuperscript{6}
• constipation (see page 33).\textsuperscript{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.\textsuperscript{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.\textsuperscript{14} However, physical impairment, interventions and self-esteem may affect sexual expression and intimacy.

• the senses (sight, hearing, sensation) are generally unimpaired.\textsuperscript{1}

**Cause of death**
This is almost always due to respiratory failure as a consequence of respiratory muscle weakness and/or repeated chest infections.\textsuperscript{1}
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 that 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.

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 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.4

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.4

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.4

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.4

See the psychological support section on page 48 for more information about talking to people before and after diagnosis of MND.
Incidence and prevalence 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).¹⁵
• Based on the size of the UK population, this equates to 4,500 people currently living with MND.¹⁵

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 years is uncommon.⁴
• The male:female ratio is 3:2, although this varies with age and evens out in later years of life.¹⁶

Clinical presentation is sub-divided in two ways: by the presence of upper motor neuron (UMN) and/or lower motor neuron (LMN) signs, and the anatomical area first affected.¹⁷

Site of onset
• Limb-onset MND affects around two-thirds of people with MND.¹⁷
• Bulbar-onset MND affects around a third of people with MND.¹⁷
• Respiratory-onset is rarer, affecting around 3%.¹⁷

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)¹⁸
• Most common form: represents at least 80% of all MND cases.
• Involves UMN and LMN.
• 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**\(^3,19\)

• 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 years old.

**Progressive muscular atrophy (PMA)**\(^{17,20}\)

• 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)**\(^3,17,21\)

• 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.\(^{17}\) It is well recognised that some individuals live considerably longer than the average survival times given.\(^1\)
Aetiology

Most people with MND have no apparent family history of the disease. In these 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 factors for MND in people without a family history of the disease are increasing age and cigarette smoking.\(^2\)

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.\(^1\)

- 40% of inherited cases are caused by gene expansion mutations in the gene C9orf72.\(^1\)
- 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:¹

- 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 – Research overview
- Research sheet B – Inherited motor neurone disease, which is split into three parts:
  1 – Introduction to inherited motor neurone disease
  2 – Genetic testing and insurance
  3 – The options available when starting a family

Download from our website at www.mndassociation.org/researchsheets or see page 69 for details of how to order copies.

You can also contact our research team with any queries on 01604 611880 or research@mndassociation.org
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. 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.

Prescribing riluzole
Riluzole is recommended for use in MND by NICE. 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.

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 with MND 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/research
**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 43).

**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 that 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 many people report improvements in wellbeing after having such complementary therapies.  

It is important that the therapist has some knowledge of MND and that neither the person with MND or 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](http://www.alsuntangled.com)
Information you can share
Our resources for people affected by MND include:
• Information sheet 5A – Riluzole
• Information sheet 6B – Complementary therapies

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

Muscle weakness
Muscle wasting in MND causes weakness and can affect balance and posture, with the risk of falls.\(^\text{27}\)

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 42).

Fasciculations
• Fasciculations are often among the first symptoms of MND.\(^\text{28}\)
• Some people feel the rippling effect in individual muscles, but it can be more widespread.
• Fasciculations may be so obvious they can be seen by other people.
• They can seem worse when a person is stressed.
• The problem usually eases over time.

Ability to drive
All people with MND are required to inform the DVLA or DVA immediately after diagnosis, although 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, rarely, complete loss of speech.\textsuperscript{29}

**Cause**
Spasticity, largely resulting in weakness of the:
- tongue
- lips
- facial muscles
- pharynx and larynx.\textsuperscript{30}

Reduced palatal elevation can lead to hypernasal voice quality.\textsuperscript{30}

Reduced breath support can lead to reduced volume or breathy voice quality.\textsuperscript{30}

**Impact**
Progressive difficulty with articulation, slurred speech and/or loss of volume.

May rapidly lead to anarthria, even though limb function is maintained for many months.\textsuperscript{30}

**Treatment**
Early referral to **speech and language therapist** (SLT) who will:
- examine the patient’s range of movement in their lips, tongue and palate
- give advice on strategies for communication.

The SLT can also arrange for assessment and provision of Augmentative and Alternative Communication (AAC).

Options include:

**Low-tech**
- 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, and computers or tablet devices.

An **occupational therapist (OT)**/the **neuro rehabilitation service/orthotics** 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 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.
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 can reduce frustration and misunderstanding

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

• use hand gestures to assist with your own speech where suitable. Over time, this will become a very important tool, for example using the ‘thumbs up’ sign to mean ‘good’ or ‘yes’

• ensure the person with MND is the focus of communication

• allow 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
• ask complex questions that require long or difficult answers that may take up unwelcome time and energy
• use multiple choice questions. Simple questions that can be answered with yes, no or a single word are easier when speech is difficult.

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.

See page 69 for details of how to order copies.

Inability to speak doesn’t mean I can’t think or understand. Be prepared to take time to communicate.”
A person living with MND
Dysphagia

Swallowing problems affect at least two-thirds of all people with MND during the course of their illness. 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 virtually never a direct cause of death.

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
• poor or absent swallow reflex
• food not moving to the back of the mouth or disappearing down the throat due to a poor or absent swallow reflex, or an uncoordinated 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.

This can result in:
• sialorrhoea (drooling due to poor saliva control 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.³²

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.³² 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.⁴

People with more severe changes may place too much food in their mouth at one time and cram food, while others may eat more food than they need. It may be more difficult to follow advice on safe swallowing, so repeated reminders and supervised eating may be necessary.⁴

I think people with MND would find it useful if, before appointments, they could email a brief history of any changes that have occurred and any questions they have. This would save time, remove stress (especially for those who have communication problems) and reassure them that their concerns will be dealt with.”
A person living with MND
Saliva management

Although a normal amount of saliva is produced, around two to three pints every day, excess saliva is a commonly reported symptom of MND. In most patients, saliva problems are the result of poor lip seal and/or impaired ability to swallow.\textsuperscript{8}

Patients have either runny, thin saliva that drools out of the mouth or thick, tenacious saliva that is difficult to clear.\textsuperscript{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.\textsuperscript{4,8}

**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 MND 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) glycopyrronium bromide (glycopyrrolate)\textsuperscript{4}
- glycopyrrolate as the first-line treatment for sialorrhoea in people with MND who have cognitive impairment, because it has fewer central nervous system side effects.\textsuperscript{4}

*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.\textsuperscript{4}

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 enteral feeding 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.\(^8\)

Suction
Portable oral suction units are helpful if saliva builds up in the mouth.\(^8\)

Body position
Attention is needed to head support, seating and general posture\(^{33}\) – 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\(^4\)
- provide advice on swallowing, diet, posture, positioning, suctioning, hydration and pay attention to oral hygiene – consult district nurse\(^4\)
- consider treatment with humidification, nebulisers and carbocisteine. Carbocisteine is available as capsules or liquid, which can be administered via a feeding tube.\(^4, 34, 35\)

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,\(^{36}\) or flavoured ice cubes. These juices contain bromelain/papain (respectively). These hygroscopic agents are also available as tablets from health food stores.

Dry mouth

- Check and change dosage of medications if needed.\(^{37}\)
- Consider artificial saliva sprays or gels, for example Aquoral, Biotene Oralbalance or Xerotin.\(^{37}\)
- Pay careful attention to oral hygiene.\(^{37}\)
- Increase fluid intake, whether orally or through a feeding tube.\(^{37}\)
Nutrition and enteral feeding

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

• dysphagia
• depression
• cognitive impairment
• hypermetabolism
• anxiety
• respiratory insufficiency
• constipation and fatigue
• physical difficulties buying, preparing and eating foods.

Functional consequences of weight loss/malnutrition include: 31

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

Involvement of a speech and language therapist and a dietitian is essential at an early stage to assess, monitor and review the individual’s nutritional intake and to provide practical oral and non-oral dietary advice to enable nutritional needs to be met. 31

Advice may include:

• fortification to improve food quality
• recipe ideas to increase variety
• practical solutions to ease food preparation and physical eating and drinking difficulties (in association with an occupational therapist)
• recommending prescribed nutritional supplements
• discussing gastrostomy.
**Enteral feeding**

Alternative feeding methods should be discussed at an early stage and at regular intervals as swallowing problems progress.

The following may be used to ‘top-up’ oral intake, to meet full nutritional and dehydration needs and to administer medications:\textsuperscript{38}

- Nasogastric tubes (NGT)
- Percutaneous Endoscopic Gastrostomy (PEG)
- Radiologically Inserted Gastrostomy (RIG)
- 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.

A nasogastric tube can be used in the short-term to build up someone who is malnourished or dehydrated, and who wants to proceed with gastrostomy, or it may be used for a longer period. This method may be used in people for whom other types of gastrostomy are not appropriate, or may be preferred by some. It is often considered less comfortable than gastrostomy tube placement.\textsuperscript{31}

**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.\textsuperscript{39}

The optimal timing of gastrostomy 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 recommended, even if it isn’t used straight away as, when needed, it can improve/maintain quality of life.\textsuperscript{39} Possible risks of a late gastrostomy include low critical body mass, respiratory complications, risk of dehydration, different methods of insertion, and a higher risk of mortality and procedural complications.\textsuperscript{4}

Someone considering a gastrostomy needs to be aware of the level of support needed to manage living with this intervention. For example, who will manage the feeds in the community.

If it is a family member, this person will need to understand what is involved and how often feeds should be administered, to ensure they
can physically manage. If care support is needed, arrangements need to be made at an early stage.

Be aware that some people will not want to have this type of intervention and their decision should be respected. Exploring pros and cons will enable the individual to make an informed choice. Gastrostomy 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 (page 58).

**Constipation**

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

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

**Treatment:**

- Maintain hydration and assess fibre intake.
- Use of 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 (eg prunes) are also readily available.

Diarrhoea may be due to constipation with overflow.

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**Information you can share**

Our resources for people affected by MND include:

- *MND Association Recipe Collection: easy to swallow meals*
- *Information sheet 7A – Swallowing difficulties*
- *Information sheet 7B – Tube feeding*
- *Information sheet 11B – Mouth care*

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

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

**Causes of breathlessness**

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

Acute breathlessness is usually caused by an acute event, such as pneumonia, lower respiratory tract infection or sputum retention, against a background of previously undetected respiratory muscle weakness.\(^6\)

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.\(^42\)
- 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 concerns around end of life decisions, increased dependency and being unable to communicate.\(^4\) (See sections on psychological support, palliative care and respiratory management.)

**Medication to consider:**

- buccal midazolam\(^43\) (liaise with palliative care team for dosage)
- lorazepam sublingually\(^43\) (slower acting than midazolam and ineffective in those with excess saliva)
- oral morphine (refer to palliative care team).\(^44\)

**Laryngeal spasm**

- This may be experienced due to impaired swallow.
- It is often preceded by gastroesophageal reflux.\(^45\)
- It can be extremely distressing: it can prevent breathing or speaking.
- It can be accompanied by stridor.\(^46\)
Treatment:
• Consider lorazepam.43

Weak cough
The NICE guideline on MND recommends cough augmentation techniques for people with MND who cannot cough effectively:
• Unassisted breath stacking and/or manual assisted cough.4
• Assisted breath stacking (eg 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.4
• 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.4

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.

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 accessory muscles. 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 be admitted via 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{45}

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

Upper Respiratory Tract Infection (URTI) should be treated with early use of antibiotics.\textsuperscript{47}

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{4}

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 referral may be needed.\textsuperscript{4}

The \textbf{NICE guideline on MND (NG42)} 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. $\text{SpO}_2$ may be used for those with severe bulbar involvement.\textsuperscript{4}

However, results of these tests may give false reassurance, so ask about the following symptoms and signs:\textsuperscript{4}
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.\(^{48}\)

**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>
</tr>
</thead>
<tbody>
<tr>
<td>Breathlessness</td>
<td>Increased respiratory rate</td>
</tr>
<tr>
<td>Orthopnoea</td>
<td>Shallow breathing</td>
</tr>
<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>
</tr>
<tr>
<td>Daytime sleepiness</td>
<td>Use of accessory muscles of respiration</td>
</tr>
<tr>
<td>Poor concentration and/or memory</td>
<td>Reduced chest expansion on maximal inspiration</td>
</tr>
</tbody>
</table>
Assisted ventilation

Non-invasive ventilation (NIV)
This is a method of providing ventilator support to a person with MND using a close-fitting mask that may cover the nose and/or mouth. A trial of NIV can be offered if the person’s symptoms and signs and the results of respiratory function tests indicate they are likely to benefit from the treatment.

NIV is often best for people with orthopnoea, disturbed sleep, morning headaches and excessive daytime sleepiness.\(^6\)

The acceptance and success of this method often depends on the correct choice of interface (mask or nasal cushion).\(^6\) Patients who derive most benefit are those cases where the respiratory muscles are weak without any bulbar disease.\(^6\)

If a person with MND has very weak upper limbs, they need to have a carer available at all times to allow repositioning or removal of the mask.

Benefits of NIV include:
- improved quality of life and sleep, with less troublesome symptoms during the day\(^6\)
- increased survival.\(^6\)

Disadvantages of NIV include:
- it is less likely to be effective where there is more severe bulbar involvement\(^6\)
- it can be used during the day, but unless a nasal rather than a full-face mask is used, it may be difficult to speak or eat while using it.\(^6\)
- risk of aspiration if control of oropharyngeal secretions is poor\(^6\)
- skin breakdown if the interface is used for long periods\(^6\)
- inability to tolerate the interface or use the machine\(^6\)
- it can become less effective as MND progresses\(^6\)
- increasing dependency over time, which may have an adverse impact on quality of life, and present end of life withdrawal difficulties\(^4\)
• it usually requires assistance of a carer and can increase the carer burden. As such, there may be additional problems if the person with MND lives alone.

Invasive ventilation
The decision to offer a tracheostomy electively, with full ventilation, is controversial and some units are reluctant to offer this as an option. There are huge cost implications for the provision of specialist care and great impact on the carers and families of people with MND. This intervention may mean the person with MND lives longer, but their other disabilities will continue to progress.

Great thought must be put into supported discussions with the person with MND (and, where appropriate, their carer/family) about the long-term implications of invasive ventilation (being unable to communicate, total dependence and what would happen if they were diagnosed with dementia).

Very occasionally, a person with MND will be invasively ventilated due to an acute episode of respiratory insufficiency. This can happen as an initial presentation of the disease with the diagnosis of MND made in intensive care. There can be difficulties in such instances in weaning the person from invasive ventilation.

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 page 62 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 page 62.

Many [hospice doctors](#) have experience in managing the process of withdrawing ventilation and may be called upon if needed for support and information. It has been recognised that withdrawal of ventilation can be very challenging for professionals, and guidelines are available from the Association for Palliative Medicine.

Oxygen

Respiratory muscle weakness leads to the retention of carbon dioxide (hypercapnia) rather than hypoxia. In this situation, NIV 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 caution – preferably under guidance of the person’s specialist team.

Oxygen is not likely to be the first line in treatment of terminal dyspnoea, when opiates may be more effective. For more guidance, speak to your specialist [palliative care team](#) or a [respiratory consultant](#) with links to the neurology team.

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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. See [psychological support](#), [palliative care](#) (page 55) and [advance care planning](#).

- **Alternative feeding**
  
  If respiratory function is deteriorating, enteral feeding may be discussed even if there are no immediate problems with swallowing or weight (page 31). 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)
• *Management of respiratory insufficiency in MND DVD*
• *MND Reading list: Respiratory problems*

*NICE Guideline NG42: Motor neurone disease: assessment and management* ([www.nice.org.uk/ng42](http://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 – *NICE guideline for non-invasive ventilation (NIV)*
• Information sheet 8D – *Troubleshooting for non-invasive ventilation (NIV)*
• Information sheet 8E – *Air travel and ventilation for motor neurone disease*

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

MND is a complicated disease with no two people having identical problems or symptoms. A person living with MND may only have one symptom at a particular time, but things can change either rapidly or slowly.”

A carer of someone with MND
Pain

Pain may be caused by:
- muscle cramps or spasticity
- mechanical stress on joints from muscle weakness
- skin sensitivity
- immobility
- oedema
- constipation (see page 33)
- occasionally, neuropathic pain.\textsuperscript{11}

The need for anticipatory prescribing is crucial to enable the patient to maintain control.\textsuperscript{50}

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).\textsuperscript{11}

Medication for muscle cramps

- Consider quinine as first-line treatment for muscle cramps in MND.\textsuperscript{4}
- If quinine is not effective, not tolerated or contraindicated, consider baclofen instead as a second-line treatment.\textsuperscript{4}
- If baclofen is not effective, not tolerated or contraindicated, consider tizanidine, dantrolene or gabapentin.\textsuperscript{4}

Medication for muscle stiffness, spasticity or increased tone

- Consider baclofen, tizanidine, dantrolene or gabapentin to treat muscle stiffness, spasticity or increased tone in people with MND.\textsuperscript{4}
- If these treatments are not effective, not tolerated or contraindicated, consider referral to a specialist service for treatment of severe spasticity.\textsuperscript{4}

Notes:
- Dosage of muscle relaxants should be carefully adjusted to avoid increased weakness and decreased mobility.\textsuperscript{11}
- Check whether the patient is taking a statin and consider this being discontinued, as muscle cramps are a frequent side effect.
Opiates may be used for pain relief and can also be used for symptomatic treatment of dyspnoea and coughing. With careful titration, excessive drowsiness and respiratory depression can be avoided.

- morphine
- buprenorphine or fentanyl patches.

**For neuropathic pain:**
- tricyclics or gabapentin/pregabalin.

**Physiotherapy**
- careful positioning to relieve discomfort
- passive exercise programme
- prevention of contractures
- maintenance of joint mobility
- regular review of posture.

**Skin sensitivity**
- Good skin and pressure care is vital. Someone with MND may be aware when they need to turn or move, but may need help to adjust their position.
- 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.

**Immobility**
Extra care is needed when lifting, handling and positioning.

**Oedema (fluid retention)**
- This may largely be related to restricted activity and posture or to intercurrent disease, which should be treated accordingly.
- 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.
- 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, and people with MND may fall into one of four groups:

• around 50% are unaffected by cognitive change

• around 35% experience mild cognitive change, with specific deficits in executive functions, language, behaviour and/or social cognition

• up to 15% develop frontotemporal dementia (FTD), either at the same time or after diagnosis of MND

• up to 15% of patients with FTD go on to develop motor impairments where MND is diagnosed after 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.

Cognitive impairment may manifest as early deficits on neuropsychological tests of executive functions (verbal fluency). Some people may have primary language problems that may manifest as reduced verbal output and failure to initiate conversations. The person may respond in short phrases or single words.

People may experience problems with:

• learning new tasks, including the use of equipment for treatment (such as gastrostomy and NIV)
• short-term memory loss
• lack of awareness or insight into their problems or their impact
• poor concentration
• inappropriate social behaviour
• impulsivity, which can include repetitive or persistent actions and a tendency towards sweet foods
• literacy and language
• making decisions
• planning for the future
• managing affairs/finances.

Not all of the above may be evident and they can vary in degree. Changes can be subtle and may be masked by movement and speech problems. Some may be part of the normal ageing process.
Carers may be unaware of mild impairment, as increasing physical disability results in loss of autonomy and greater reliance on others.\textsuperscript{61}

**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.\textsuperscript{60} These behaviours may partially meet the criteria for frontotemporal dementia.\textsuperscript{60}

People with MND may also find it difficult to recognise emotions in others and have altered sensitivity to social and emotional cues.\textsuperscript{60}

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.\textsuperscript{60}

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 see 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.\textsuperscript{60}

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.\textsuperscript{4} Respiratory muscle weakness as a cause of cognitive changes should be excluded.
Frontotemporal dementia (MND-FTD)

The following explanation defines MND-FTD and the combination of symptoms that may indicate frontotemporal dementia.\(^{60}\)

**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, 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.

Listen to carers when monitoring for change. For example, if a carer says, ‘My partner is different, he just doesn’t love me anymore’ – this may well be a change in the relationship since diagnosis, but it could also indicate something deeper in terms of behaviour and lack of empathy.” A carer whose husband had MND-FTD
Managing cognitive change/MND-FTD

To help explain the need for assessment should changes occur, it is important to acknowledge to patients and carers that cognitive change may be part of the clinical picture of MND. 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 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 MND 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 frontotemporal dementia may lack mental capacity. Capacity should be assessed where FTD is evident and care provided in line with the Mental Capacity Act 2005.4,60

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 equally affect their ability to continue to drive safely.61

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.

Download from our website at www.mndassociation.org/publications or see page 69 for details of how to order copies.
Psychological support

People with MND and those close to them, which may include children, often experience considerable psychological and emotional distress.\(^{10}\)

MND is characterised by a series of losses with the accompanying issues of grief and bereavement. The major challenges are coping with loss and living with change.

The impact will be influenced by other factors, including the person’s age, personality, gender and life stage. Spiritual beliefs, current life stresses, cultural background and the help and support they receive will have a bearing on the person’s ability to cope with their illness.

Much can be done to alleviate this distress, help people to adjust and make the most of their coping skills.

**Before the diagnosis**

Anxiety in someone before a diagnosis of MND can increase as a result of:

- onset of worrying symptoms
- problems in identifying their cause
- a long period of investigation and the specialist’s need to be sure before giving the diagnosis.\(^1\)

**After diagnosis is confirmed**

- Give information that can help the person with MND and their relatives make sense of what is happening to them, maintain control and make plans for the future.
- Allow the person to express thoughts, feelings and concerns: this can help to identify information and support needs.
- The information needs of carers and any children in the family will vary and may be different from those of the person with MND. Carers may seek out information earlier, to help them prepare for their role.
• Too much information at the time of diagnosis may be overwhelming, but feelings about the illness may change, so it is important to check individual information needs on a regular basis, give opportunities for people to return for more detail, and check their preferences for involving family members/carers.
• It is important to check the information given, and the language used, is understood and inaccuracies are discussed.
• Information may need to be given a number of times, as anxiety may limit someone’s ability to absorb information.
• Identify an individual, such as someone from the local mental health or psychological support team, to provide ongoing support and information.
• Offer the person with MND information about sources of emotional and psychological support. This may include MND Association local groups or the online forum. If needed, refer to counselling or psychology services for specialist assessment and support.
• Ensure that people know how to source reliable information: see page 69 for details of the MND Association’s care information.
• 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.

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.\(^\text{10}\) Adjustment to life with the disease is made more difficult by the rate of deterioration and the accompanying changes and losses.

• 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. Dysarthria and other communication problems will affect the ability of someone with MND to share their 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 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:

• fear of ‘choking to death’ or ‘fighting for breath’
• loss of independence and dignity
• 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 these.

**Denial**

Denial is a coping mechanism that sometimes operates alongside awareness of the condition and its implications. Sustained denial is less helpful and can contribute to higher levels of anxiety and depression. The strength of denial can often be assessed by using past or future orientated questions.

**Anger**

Many people with MND are concerned about how angry they feel as a result of frustration and loss of control. It may be displaced on to others, including health and social care professionals as well as family. Feelings of anger may occur at any stage of the illness and need to be acknowledged. If anger is existential in nature, spiritual support may be appropriate if the family has a particular faith or belief system.

**Hopelessness**

Feelings of hopelessness may be linked to the recognition that many of life’s expectations will never be realised. Time is required to explore these issues.

**Anxiety and depression**

It may not be easy to diagnose depression, or differentiate it from
appropriate sadness, because the symptoms of depression can be very similar to the symptoms of MND. These include weakness, diminished appetite, irritability, hopelessness, early morning waking, insomnia, feeling worthless and loss of concentration.\textsuperscript{10}

Consider formal screening for anxiety and depression in people with MND and especially in carers (depression is common in carers of people with MND)\textsuperscript{10} particularly where persistent low mood or hopelessness are expressed.

Selective serotonin reuptake inhibitors (SSRIs), such as sertraline or citalopram, may be considered. These may take several weeks to have an effect.\textsuperscript{63}

**Sleep disturbances**

Improvement in sleep is usually the first benefit of antidepressant medication.\textsuperscript{63} Cramps, spasms, fear, anxiety and respiratory problems should also be treated to improve quality of sleep.

**Emotional lability (pseudo-bulbar affect)**

Associated with UMN involvement in the corticobulbar pathways, emotional lability presents as disturbed emotional responses, including inappropriate and uncontrollable laughter and crying.\textsuperscript{9} In some cases, this can be extreme. Emotional lability can be distressing for both the person with MND and those around them.

It is a common problem and can occur in people with cognitive change, but it is not necessarily a sign of wider cognitive changes.\textsuperscript{64}

Some people find that understanding the symptom helps them to manage the impact; but others find it limits where they go and what they do. Medication, such as tricyclic antidepressants or SSRIs, may alleviate the symptoms, although this isn’t always successful.\textsuperscript{9}

**Impact on professionals**

MND creates many challenges for professionals and can arouse strong emotions. These can include 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.
Providing medical evidence for benefit applications (DS1500)

People with motor neurone disease (MND) may apply for disability/employment benefits.

If someone with MND is applying for Attendance Allowance, Personal Independence Payment, Employment and Support Allowance or for a reassessment of Disability Living Allowance, they may be fast-tracked and considered for benefits under the ‘Special Rules for terminal illness’ provisions.

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

The person with MND may have their application fast-tracked under the Special Rules during the initial phone call they make to enquire about Personal Independence Payment. They should tell the call handler at this stage that they want this to be considered as a Special Rules case. They will be asked whether they already have or are going to get a DS1500 form, and will be asked to send it in.

The claim will then be completed.

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.’

MND is a terminal, rapidly progressive disease. As a result, people with MND will pass the diagnosis element of Special Rules on the DS1500 form, as it is a terminal condition. However, the prognosis element can be problematic.
It is important to remember that 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.

A claimant who successfully applies through Special Rules can usually continue to claim for up to three years before reassessment.

A third of people with MND die within a year of diagnosis and more than half die within two years.65

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. Delays mean that someone may be living without any kind of financial support for many months.

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

**Information you can share**
Our resources for people affected by MND include:
- Information sheet 10A - *Benefits and entitlements*

Download from our website at [www.mndassociation.org/publications](http://www.mndassociation.org/publications) or see page 69 for details of how to order copies.
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 local MND care centre or network services and the speed of progression of symptoms.66

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 that 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, 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 recommends offering the person with MND the opportunity to discuss their preferences and concerns about care at the end of life at 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.

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.
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, and memory boxes or books for children.

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, including 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 guide on advance 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. When appropriately recorded as part of advance care planning, an ADRT is legally binding, and these wishes should be followed.
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.

In England and Wales, refusal of CPR may be included on an ADRT, which is legally binding, with clear instructions about when this should be applied.

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 be used until they do not have capacity or cannot communicate their own decisions.67,68

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 *End of life guide* has plenty of 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.

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)

The National Council for Palliative Care (NCPC) has 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*. Visit [www.ncpc.org.uk](http://www.ncpc.org.uk) for more information.
The most common cause of death in MND is respiratory failure, often with additional chest infection.\(^6\)

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.\(^6^9\)

It is very important to reassure people with MND and carers that death from choking is exceptional.\(^1\)

**Medication at end of life**

Check all symptom control:

- pain – patients may experience severe discomfort and/or distress, particularly in the later stages. Opiates may be an option. Careful titration will avoid excessive drowsiness and respiratory depression\(^7^0\)
  - pressure care
  - dyspnoea
  - dysphagia
  - salivation
  - insomnia
  - anxiety/depression
  - restlessness/agitation
  - bowels and bladder
  - re-assess 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\(^4^4\)
• 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.\textsuperscript{4,44,71}
• opioid analgesics to reduce cough reflex, relieve dyspnoea and therefore fear and anxiety. They can also control pain\textsuperscript{11}
• anti-emetics for nausea.\textsuperscript{44}

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.\textsuperscript{44}

*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. Eye movements and single response answers to closed questions may be used, or picture/alphabet boards or other communication aids as appropriate (page 23).\textsuperscript{30}

**Supporting family and carers at this stage**

MND is unpredictable and may progress rapidly, with death occurring more quickly than anticipated.\textsuperscript{69} 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 will be able to 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.

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.
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.

- 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.
- Negotiate with the carer/family what type and frequency of support would help reduce anxiety and fear. Consider referral for additional psychological support if appropriate. 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. MND Association visitors and local branch or group meetings can be a valuable source of support, as can chaplaincy services, which can be accessed via MND care centres and hospices.
- Children in the family may benefit from being referred to the school nurse. They may need support for mental wellbeing and any educational difficulties experienced due to anticipatory grief or school absence for caring duties.

**MND forces changes in roles and relationships. Consider:**
- mapping the social support system of the family using a genogram
- meeting the varying 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 link 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 on 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 carer’s assessment of their needs. You may also need to liaise with their school to ensure they are receiving the support they need. An early help assessment 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 the school nurse to support any children?

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 at a speed that feels appropriate.

These resources can be ordered from MND Connect on 0808 802 6262.

So what is MND anyway? can also be downloaded from our website at www.mndassociation.org/ypinfo

Other information you can share
Our resources for people affected by MND include:
• Caring and MND: quick guide
• Caring and MND: support for you
• Information sheet 14B – Bereavement support

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 information, support and signposting to other services and agencies.
Telephone: **0808 802 6262**
Email: [mndconnect@mndassociation.org](mailto:mndconnect@mndassociation.org)

**Information resources**
We produce high quality information resources for health and social care professionals who work with people with MND. We also have a wide range of resources for people living with and affected by MND. Downloads of all our information sheets and most of our publications are available from our website at [www.mndassociation.org/publications](http://www.mndassociation.org/publications) or you can order our publications directly from the MND Connect team.

**MND Association website**
Access information for health and social care professionals on our website at [www.mndassociation.org/professionals](http://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, such as conferences and masterclasses. Find out more at [www.mndassociation.org/education](http://www.mndassociation.org/education)
**Wheelchair service**

If you are seeking information on wheelchairs, or if there are delays in assessment or provision of an appropriate wheelchair for someone with MND, our wheelchair service may be able to help, through training, joint assessments, advice or support. Call MND Connect on **0808 802 6262** or email **wheelchairs@mndassociation.org**

**Communication aids service**

This service helps people affected by MND and health and social care professionals with queries about communication aids. It provides limited financial support for communication aids or some items on loan, if unavailable or delayed through health and social care services. Our aim is to improve provision and information on a local and national level, through collaboration with professionals. Call MND Connect on **0808 802 6262** or email **communicationaids@mndassociation.org**

**MND support grants and equipment loan**

Where statutory funding or provision has been explored and is not available, we may be able to provide a support grant or equipment loan. Our support grant service consists of providing care and quality of life grants for people with MND. This service is supported by MND Association branch and group funds, and by the Association’s central fund.

Our equipment loan service is focused on three core items:

- riser-recliner chairs
- specialist communication aids
- portable suction units.

For suction units, a small charge is made to statutory services for carriage, maintenance and cleaning.

Referrals for support grants or equipment loan need to be made by a relevant health or social care professional.

To find out more about MND support grants or equipment loan, please visit [www.mndassociation.org/getting-support](http://www.mndassociation.org/getting-support), email support.services@mndassociation.org or call MND Connect on **0808 802 6262**.
Research into MND
We fund and promote research that leads to new understanding and treatment and brings us closer to a cure. For more information go to www.mndassociation.org/research, call 01604 611880 or email research@mndassociation.org
For the latest research news, visit our research blog at www.mndresearch.blog
For updated information on clinical trials, visit www.mndassociation.org/treatment-trials
Our peer-to-peer research and care community blog (RECCOB) has a number of reporters who write updates on MND-related workshops and events at www.reccob.wordpress.com

International Symposium on ALS/MND
Each year we organise the world’s largest clinical and biomedical research conference on MND. It is the premier event in the MND research calendar for discussion on the latest advances in research and clinical management. Visit www.mndassociation.org/symposium

Local support
Regional care development advisers
Our network of regional care development advisers (RCDAs) have specialist knowledge of the care and management of MND. They work closely with local services and care providers to ensure effective support for people affected by MND, provide education on MND, and are champions at influencing care services.

MND care centres and networks
We fund and develop care centres and networks across England, Wales, and Northern Ireland, which offer specialist clinical expertise from diagnosis onwards.

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

Association visitors (AVs)
Association visitors are volunteers with experience of MND who provide one-to-one local support to people affected by MND.
Other organisations and resources

You can find links to other useful websites at
www.mndassociation.org/usefulorgs

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)

Carers

**Caring for Carers Hub**
An e-learning course from the RCGP to help primary care staff to support carers. [http://www.rcgp.org.uk](http://www.rcgp.org.uk)

**Caring for carers (Northern Ireland) 2006**
[www.tinyurl.com/caringforcarersni](http://www.tinyurl.com/caringforcarersni)

**The Carers Strategy for Wales 2013**
[www.tinyurl./carersstrategyforwales](http://www.tinyurl./carersstrategyforwales)

**Recognised, valued and supported: next steps for the Carers Strategy (England) 2010**
[www.tinyurl.com/carersstrategynextsteps](http://www.tinyurl.com/carersstrategynextsteps)

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)**
Information, advice and social opportunities for people affected by inherited forms of FTD. [www.ftdsupport.org](http://www.ftdsupport.org)
End of life care

Dying Matters
A coalition of organisations promoting public awareness of dying, death and bereavement. [http://dyingmatters.org](http://dyingmatters.org)

End of Life Care Strategy: promoting high quality care for adults at the end of their life 2008
[www.tinyurl.com/endoflifecarestrategy](http://www.tinyurl.com/endoflifecarestrategy)

GMC’s ethical guidance on end-of-life care

Welsh Government End of Life Care Delivery Plan 2013
[www.tinyurl.com/endoflifecarewales](http://www.tinyurl.com/endoflifecarewales)

Palliative care

National Council for Palliative Care
An umbrella charity for all those involved in palliative, end of life and hospice care in England, Wales and Northern Ireland
[www.ncpc.org.uk](http://www.ncpc.org.uk)

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](http://www.pallcare.info)

Cicely Saunders Institute of Palliative Care, Policy and Rehabilitation
Research and resources on palliative care. [www.kcl.ac.uk/palliative](http://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](http://www.goldstandardsframework.org.uk)
Reference List


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


Thank you for taking the time to provide your feedback on one of our information resources.

This questionnaire can be accessed online if preferred, using the following link: www.surveymonkey.com/s/gpbooklet

What is your profession or specialism?

Did you find this resource useful?

☐ Yes    ☐ Somewhat    ☐ Not really    ☐ No

Please explain your answer

Will this information resource help you to provide people affected by MND with any of the following? (tick all that apply)

☐ an increased understanding of their symptoms
☐ an increased understanding of their condition
☐ more independence
☐ an increased ability to raise awareness of their needs
☐ more confidence
☐ improved quality of life
☐ a greater ability to maintain dignity

Continued overleaf
Feedback form continued

Were there any particular topics that were useful to you?

Was there any information that you didn’t find useful or relevant?

Are there any other MND-related topics you would like more information about?

Would you be happy to help us improve our information by becoming an expert reviewer?

☐ Yes (please include your email address below)  ☐ No

Do you have any experiences of working with people with MND you could share as an anonymous quote or case study for future resources?

☐ Yes (please include your email address below)  ☐ No

Please return your completed form to:
Education and information team
MND Association
PO Box 246
Northampton NN1 2PR

Name:

Email:
Acknowledgements
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The MND Association is indebted to the authors of and contributors to the previous version of this booklet, *Motor Neurone Disease: A problem solving approach.*

If you’d like to help us by reviewing future versions of this or other resources, please email us on infofeedback@mndassociation.org
About us
The MND Association was founded in 1979 by a group of volunteers with experience of living with or caring for someone with MND. Since then, we have grown significantly, with an ever-increasing community of volunteers, supporters and staff, all sharing the same goal – to support people with MND and everyone who cares for them, both now and in the future.

We are the only national charity in England, Wales and Northern Ireland focused on MND care, research and campaigning.

Our mission
We improve care and support for people with MND, their families and carers.

We fund and promote research that leads to new understanding and treatments, and brings us closer to a cure for MND.

We campaign and raise awareness so the needs of people with MND and everyone who cares for them are recognised and addressed by wider society.

MND Association
PO Box 246, Northampton NN1 2PR
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www.mndassociation.org

@mndeducation
/mndassociation

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. MND does not usually affect the senses such as sight, sound and feeling.
• 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 a proportion 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 up to 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.