



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

Quick reference

Responsibilities when MND is suspected or confirmed

Prompt referral for diagnosis (page 13)

Use the Red Flag tool (page 13-14) to recognise early signs of MND in order to refer to neurology in a timely manner.

Assessing needs

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. Refer to appropriate specialist teams as necessary.

Monitoring symptoms

Monitor and assess symptoms, including signs of:

- Respiratory involvement (page 40) – early signs should trigger referral to the specialist respiratory team.
- Cognitive or behavioural change (page 54) – this has implications for decision making and future management.

In collaboration with consultants in neurology and palliative care, initiate appropriate management and treatment, including anticipatory symptomatic intervention.

Support and information

Provide support and information throughout the disease course. Advise on the need to inform insurers and DVLA or DVA of diagnosis. Complete a form DS1500 or SR1 to support a benefit application (page 64).

Repeat prescriptions for riluzole (page 19)

Issue repeat prescriptions if the person with MND is prescribed riluzole by their neurologist: a shared-care protocol should be agreed.

Palliative care (page 67)

Include the person on local palliative care registers/lists/co-ordination systems – eg the Gold Standards Framework, Electronic Palliative Care Co-ordination Systems (EPaCCS).

Advance care planning (page 69-71)

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.

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are
clickable

Introduction

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 There is a 1 in 300 risk of being diagnosed with MND, but as the progression can be rapid, fewer people are living with this disease than you might expect. GPs and others in the primary care team may come across few people living with MND during their professional lifetimes. However, as a member of the primary care team you can make a great difference to the quality of life of those people, with timely care, support and symptom management.

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

NICE Guideline on MND

The National Institute for Health and Care Excellence (NICE) Guideline NG42 - *Motor neurone disease: assessment and management* aims to improve care from point of diagnosis through to end of life. It includes topics such as information and support, organisation of care, managing symptoms and preparing for end of life.

More information about the NICE guideline on MND is available from **www.nice.org.uk/guidance/ng42**

RCGP online module in motor neurone disease

Visit **www.mndassociation.org/gp** to access the RCGP online module on MND, along with other tools and information for GPs.

How the MND Association can support you

The MND Association supports professionals to care for people affected by MND in a variety of ways, which includes a range of information and educational opportunities, as well as local support and advice from our staff and volunteers. See page 86 of this guide for more information about how the MND Association can support you.

MND Connect

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

Information for you and your patients

We have an extensive range of information and resources designed to support:

- people with MND
- carers
- families (including children and young people)
- health and social care professionals.

Useful resources are signposted throughout this guide. Please share these sensitively with your patients, particularly resources about end of life care.

Visit **www.mndassociation.org/publications** to view our full range, or contact our MND Connect helpline to order hard copies. Call 0808 802 6262 or email **mndconnect@mndassociation.org**



The role of the GP and primary care team

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

Working with the specialist team

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

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

The GP may work in liaison with an MND care co-ordinator, who may be based in an MND care centre or network, where such a service exists. Visit **www.mndassociation.org/carecentres** to find out where your nearest MND care centre or network is located.

Onward referrals

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

The GP may act as the gatekeeper to other services, referring to appropriate specialists in the multidisciplinary team, including those in the table on the following page.

Professional	Area of expertise/assistance provided
District/ community nurse or community matron	nursing support at home, monitoring of symptoms and arranging/demonstrating nursing equipment
Physio- therapist	mobility (walking, balancing etc), exercise and activity, positioning, posture, respiratory, weak cough, difficulty clearing sputum
Respiratory team	coughing, breathlessness at rest/slight exertion/lying flat, sleep disturbances, daytime sleepiness, morning headaches, tiredness or lethargy, advice on positioning for drooling/thick mucus
Occupational therapist	mobility (walking, balancing etc), manual handling, positioning, fatigue management, aids/equipment for activities of daily living
Speech and language therapist	swallowing and diet modification, positioning, coughing, speech and communication, communication aids and equipment, voice banking
Dietitian	weight loss, lack of appetite, diet modification, hydration, gastrostomy
Palliative care services/ hospice	general symptom management, support for individual and family, uncontrolled pain, breathlessness, fear/anxiety/depression, spiritual support, guidance on getting affairs in order
Social worker	difficulties in activities of daily living, advice on benefits and entitlements, referral to local services, support for individual and family, counselling
Psychologist/ counsellor/ neuro- psychologist	fear/anxiety/depression, cognition or behavioural change and frontotemporal dementia

Actions for the GP and primary care team

Can you identify the person with MND as having a neurological condition on your practice IT system?	If no: Use the appropriate coding so your system flags when the person contacts the surgery and the appropriate response can be given.
Is the person's care regularly discussed within the primary care team?	If no: Put the patient's name on the Supportive Care Register within your practice.
Is there a member of the primary care team who has been nominated as the co-ordinator of the patient's care?	If no: Identify a named person who: <ul style="list-style-type: none"> (a) acts as the single point of contact within the team and (b) can liaise with all health and social care professionals involved in the person's care.
Are efforts being made to control all symptoms currently experienced by the person with MND?	If no: Use the sections in this booklet to look at options for management. If symptoms are still uncontrolled, refer onwards.
Is it flagged with the out-of-hours service provider that this person has MND?	If no: Notify your out-of-hours service provider and ensure access is given to all relevant facts/wishes.
Are plans in place for emergency care provision if the person's carer cannot support for any reason?	If no: An assessment of care needs should consider this. Refer to social services.
Do you know what the patient's wishes are in relation to end of life?	If no: Clarify these as the opportunity presents, and ensure this is recorded.

Have end of life wishes been recorded, so other members of the primary care team are also aware?	If no: Encourage the patient to record their wishes and make other members of their family and the primary care team aware.
Do you have a copy of any existing DNACPR order/ ADRT/advance care plan/ preferred place of care request in your practice records?	If no: Ask to keep a copy in your patient records.
Is the ambulance service aware of any DNACPR order/ADRT/preferred place of care request?	If no: Notify the ambulance service so it can be flagged on its system to prevent inappropriate care or treatment.
Are you aware of the name of the main carer for this person?	If no: Record the name of the main carer and their contact details on your practice system. Add a flag that indicates they are a carer.
If the carer is registered with your practice, is their record cross-referenced on your system?	If no: Put a flag on the carer's record so you are aware of any potential impact on the carer's health.

Presentation

Pathology of MND

MND is characterised by progressive degeneration of motor neurones.

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

MND is insidious in its onset and development. The initial stages, the speed and the pattern of progression can all be very variable. Not all symptoms necessarily happen to everyone, nor do they develop in the same order or progress at the same rate.⁴

Diagnosis of MND can be challenging (see page 13). Neurological review over several months may be necessary before a person receives a firm diagnosis. This delay can pose real problems for both the patient and their family, but is important that the person is not misdiagnosed, and that the MND diagnosis is certain.

Typical first symptoms

MND causes progressive muscular weakness that usually presents in one body region. Depending on which region, symptoms may include:

- foot drop, with trips and other mechanical falls, without sensory change
- loss of dexterity, weakened grip, without sensory change
- change in voice quality, in particular slurred speech - can occasionally be mistaken for myasthenia gravis
- gradual awareness of more effortful tongue movements or swallowing
- frequent muscle cramps, continuous fasciculation, muscle wasting and weight loss.³

First symptoms then progress to:

- loss of function of limbs
- weakness and wasting of muscles
- increasing impairment of speech and swallowing.

Most people with MND will eventually be completely dependent on others for all activities of daily living.⁵

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

Some people with MND may experience cognitive or behavioural changes, ranging from mild effects to noticeable impairment. A minority will experience frontotemporal dementia (MND-FTD) and this may be apparent as the first symptom or soon after the motor weakness presents.¹ 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 54).³



Additional symptoms

- fatigue – this is common throughout the disease and can be severe, affecting remaining functional ability⁷
- reduced saliva clearance and drooling: this can be watery (sialorrhoea) or thick, tenacious saliva and mucus (see page 31)⁸
- emotional lability (emotionality or emotional reflex hypersensitivity) – sudden unexpected crying or laughter that is recognised by the individual with MND to be out of proportion or inappropriate to the stimulus (see page 63)⁹
- anxiety and depression (see page 62)¹⁰
- pain and discomfort from muscle cramps, spasticity or loss of muscle tone around joints (see page 22-24)¹¹
- insomnia⁶
- constipation (see page 38-39).¹²

Cause of death

Progression of symptoms in MND is inevitable. A third of people with MND die within a year of diagnosis, and more than half die within two years. This is almost always due to respiratory failure as a consequence of respiratory muscle weakness and/or repeated chest infections.¹

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

For a very few people with MND, death can be very sudden, before an obvious end stage is reached. Most experience a longer final stage, which can last comfortably for many weeks. For most people with MND, death will be peaceful.¹³

People with MND should be encouraged to talk through options for care and preferences for end of life with those closest to them, as well as with the professionals involved in their care. With good forward planning, their wishes around end of life care (eg place of death) are more likely to be fulfilled. See page 68.

Diagnosis of MND

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

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

There is no diagnostic test for MND. Diagnosis is based on features in the clinical history and examination, usually accompanied by supportive electromyography and other tests to exclude the presence of any plausible alternative 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. If the disease course is slower than average then people may spend months unnecessarily 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.

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 tool at www.mndassociation.org/redflag

Painless, progressive weakness – Could this be Motor Neurone Disease?

1. Does the patient have one or more of these symptoms?

Bulbar features

- Dysarthria
 - Slurred or quiet speech often when tired
- Dysphagia
 - Liquids and/or solids
 - Excessive saliva
 - Choking sensation especially when lying flat
 - Tongue fasciculations

Limb features

- Focal weakness
 - Falls/trips – from foot drop
- Loss of dexterity
- Muscle wasting
- Muscle 'twitching' (fasciculations)
- Cramps
- No sensory features

Respiratory features

- Hard to explain respiratory symptoms
- Shortness of breath on exertion
- Excessive daytime sleepiness
- Fatigue
- Early morning headache
- Orthopnoea

Cognitive features (rare)

- Behavioural change
- Emotional lability (not related to dementia)
- Fronto-temporal dementia

2. Is there progression?

Supporting factors

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

Factors NOT supportive of MND diagnosis

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

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

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

Bulbar features
25% of patients present with bulbar symptoms

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

Painless progressive dysarthria – consider neurological referral rather than ENT.

Limb features
70% of patients present with limb symptoms

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

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

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

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

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

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

Ask specifically about a family history of these features.

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Confirmation of diagnosis

The NICE guideline on MND³ recommends that, if MND is suspected, the possible diagnosis should be specified in the referral letter. If so, then the GP should contact the consultant neurologist directly for an urgent review similar to the cancer pathway, so that the person is not left waiting with a very serious diagnosis in question.³

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

See the psychological support section on page 58 for more information about talking to people before and after diagnosis of MND.

ALS and variants of MND

Amyotrophic lateral sclerosis (ALS) and MND essentially mean the same thing. MND is the overarching term, largely only used in the UK and Australia, with ALS more generally used in the USA and other countries. It is also sometimes referred to as Lou Gehrig's disease in the USA.

Historically MND was classified by whether it affected predominantly upper motor neurones (UMN) or lower motor neurones (LMN), or both.

Where there is only UMN or LMN involvement, the rate of disease progression is somewhat slower.^{4, 15, 16} However, there are not really different 'types' of MND, but variable rates of progression across the whole spectrum of the disease.^{17, 18}

Amyotrophic lateral sclerosis (ALS) key features:^{16, 19}

- Wide variation in age of onset: mean age at onset is mid-60s.
- Involves a mixture of upper and lower motor neurone involvement but one may still predominate clinically.
- Characterised by a combination of muscle wasting, hyperreflexia, and sometimes spasticity.
- LMN-predominant cases with marked muscle wasting were sometimes termed progressive muscular atrophy (PMA), but these are now recognised to be hard to distinguish from ALS pathologically and progression is not always slower, so the distinction is less useful. See Progressive muscular atrophy on page 17.
- Arm, leg or bulbar onset.
- The site of symptom onset usually bears the greatest functional loss over time.
- The bulbar region is usually affected to some extent eventually but may not be prominent in all cases.

- A small group of people with bulbar-onset ALS have symptoms relatively confined to the bulbar region for several months (occasionally years), before the limbs are affected. Older females are the most common demographic, often with emotional lability. See Progressive bulbar palsy on the next page.
- Survival is variable across all cases of ALS, but is less than five years from symptom onset for 75%.
- Where initial symptom onset is in the bulbar region, or with dementia, survival is often significantly shorter (1-2 years).
- Another rarer variant involves an isolated site of onset around the shoulders, known as 'Flail arm syndrome'. It overwhelmingly affects men and may be associated with a much slower progression of up to 10 years and sometimes longer.

Primary lateral sclerosis (PLS) key features:^{15, 20}

- Primary lateral sclerosis (PLS) is a rare variant at the upper motor neurone extreme of the spectrum of motor neurone disease.
- Accounts for <3% of all people with MND.
- Age of onset is usually at least a decade earlier (age 40s-50s).
- Affects UMN only, usually with very marked spasticity and may be confused with hereditary spastic paraplegia (HSP).
- Symptom onset is usually in the legs, or more rarely in the bulbar region.
- Balance is often impaired, plus a degree of bladder hypersensitivity.
- Survival is notably longer - PLS is not usually life-shortening as it progresses very slowly and a gastrostomy is rarely needed, even in those with marked dysarthria.
- It can be difficult to distinguish PLS from UMN-predominant forms of ALS in the first few years, which are also slower than average in progression but still life-shortening for most.

In addition to ALS and PLS described above, some professionals may still refer to the historical “types” of MND, and some people may have been given them as a diagnosis. We have included a description of these below, however we recommend viewing MND/ALS as a single disease with variable symptoms, sites of onset and rates of progression.

Progressive bulbar palsy (PBP)^{4, 16}

- 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.

Progressive muscular atrophy (PMA)^{4, 16}

- Accounts for 5-10% of people with MND.
- Characterised by LMN degeneration, with muscle wasting and weakness.
- Limb onset, often with visible fasciculations.
- Includes ‘Flail arm syndrome’, and is characterised by slowly progressive, symmetrical, and usually proximal upper limb weakness.
- Sometimes misdiagnosed as multifocal motor neuropathy.

Information to share

Information sheet 2C - *Primary lateral sclerosis*

Information sheet 2D - *Progressive muscular atrophy*

Download from **www.mndassociation.org/publications** or contact MND Connect to order hard copies. Call 0808 802 6262 or email **mndconnect@mndassociation.org**

Aetiology

Most people with MND have no evident family history of the disease. In these apparently sporadic cases, it is likely that the disease develops due to a combination of genetic susceptibility and environmental factors.¹ The most clearly identified risk factor for MND in people without a family history of the disease is increasing age.²¹

A recent study¹⁴ has also shown some evidence that frequent strenuous exercise is a risk factor for MND. However, as exercise has a huge health benefit to the majority of people, this study should in no way suggest that exercise should be avoided. Further research into which people are more at risk of developing MND through exercise is needed so that appropriate advice can be given in due course.

Inherited forms of MND

Inherited MND (sometimes known as familial MND) accounts for approximately 10-15% of all people with MND.¹ The apparently sporadic and familial forms of MND are clinically indistinguishable.¹³

It is now recognised that taking a family history is unreliable as a way to exclude an inherited form. There are statistically more people with an identifiable genetic cause for their MND who will give a negative family history than among those with a known family history of MND.

Family history needs to ask about possible cases of frontotemporal dementia (FTD - see page 54-57) which may be caused independently by the most common genetic cause of MND. FTD may be misdiagnosed as Alzheimer's disease and so not realised as important.

It is important to stress that carrying a gene associated with MND does not mean the person will definitely get the disease. Concerned family members can be referred for genetic counselling.

Information to share

Research sheet B1 - *Inherited MND: introduction*

Research sheet B2 - *Inherited MND: genetic testing and insurance*

Research sheet B3 - *Inherited MND: options when starting a family*

Download from **www.mndassociation.org/researchsheets**

You can also contact our research team with any queries on

01604 611880 or **research@mndassociation.org**

Management of MND

Riluzole is the only licensed disease-modifying drug available for MND. The pivotal trial showed that, on average, it extended survival by around three months over the 18 month study (so is estimated to have at least a 10% survival benefit overall).²² It is worth noting that not all MND experts support the use of riluzole due to its limited effects and mixed evidence on its benefits.

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 may also promote neuronal survival by enhancing production of various neuronal growth factors.²³

Prescribing riluzole

Riluzole is recommended for use in MND by NICE.²⁴ There is over 20 years of safe use with significant adverse effects (typically liver function test rises) rare.

The GP and specialist should agree a shared-care protocol, under which the GP issues repeat, if not also the first prescription.

Normal baseline blood test should be undertaken initially, then monthly full blood count and liver function test for three months, followed by three-monthly for the first year, and annually thereafter.¹³ Responsibility for prescribing and monitoring should be made clear in the protocol.

Riluzole comes in tablet form, or in a liquid form for people with swallowing difficulties. A film which slowly dissolves on the tongue (orodispersible film) is also available.

Information to share

Information sheet 5A - *Riluzole*

Download from www.mndassociation.org/publications or contact MND Connect to order hard copies. Call 0808 802 6262 or email mndconnect@mndassociation.org

New treatments

Many ongoing studies are testing the efficacy of new and repurposed drugs in the treatment of MND. It is routine for these to be discussed by the neurologist involved in care. You can track the progress of research at www.mndassociation.org/treatment-trials

Symptom management

Sometimes, when people are told there is no effective curative treatment for MND, they interpret this as meaning that nothing can be done. In fact, multidisciplinary care, addressing symptoms and planning ahead to anticipate disability, has been shown to have a very significant effect on overall survival.²⁵ 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 22-54).

Free prescriptions

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

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

Complementary therapies

Although they provide no treatment for MND, many people living with the disease use complementary therapies, such as aromatherapy, reflexology, massage, relaxation techniques and acupuncture. There is no controlled trial evidence of the benefits of these therapies in MND, but some people report improvements in wellbeing.¹³

It is important the therapist has some knowledge of MND and neither the person with MND nor the therapist has false expectations that the complementary therapy will offer a cure.

Information to share

Information sheet 6B – *Complementary therapies*

Download from **www.mndassociation.org/publications** or contact MND Connect to order hard copies. Call 0808 802 6262 or email **mndconnect@mndassociation.org**

Unproven treatments

Many people with MND consider and try unproven treatments for the disease. This may lead to false hope, wasting time and money and possible adverse effects.

The MND Association co-funds an international group of scientists and clinicians, collectively known as ALS Untangled, to investigate unproven or alternative treatments for MND. This group looks for the evidence behind these unproven treatments and concludes with their own recommendations. See **www.alsuntangled.com**

Information to share

Research sheet H – *Unproven treatments in MND*

Download from **www.mndassociation.org/researchsheets**

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



Muscle weakness

MND causes muscle wasting and weakness, and can affect balance and posture, with the risk of falls.²⁶ General muscle tightness or frank spasticity (stiffness and spasms) can affect mobility, co-ordination of movement and may cause pain or increase the risk of falls.

Information to share

Information sheet 6A – *Physiotherapy*
Information sheet 6C – *Managing pain*
Information sheet 11C – *Equipment and wheelchairs*
Information sheet 11E – *Environmental controls*
Getting around booklet
Personal care booklet
Sex and relationships booklet

Information for you

Occupational therapy for MND
Information sheet P1 – Head supports for people with MND
Information sheet P2 – Wheelchairs for people with MND
Information sheet P11 – Pain in MND

Download from **www.mndassociation.org/publications** or
contact MND Connect to order hard copies. Call 0808 802 6262 or
email **mndconnect@mndassociation.org**

Consider:

- physiotherapy (including walking aids, orthotics, splints, active and manually-assisted exercise, posture management eg head supports, mobile arm supports etc)
- occupational therapy for posture management and equipment for managing activities of daily living
- medication for spasticity - although this may not always be helpful, as it can indirectly exacerbate weakness and affect alertness (see page 52).

Fasciculation:

- may be among the first signs of MND though frequently not noticed by the individual until pointed out²⁷
- may be felt as a rippling effect in individual muscles, but can be more widespread
- may be so obvious they can be seen by other people
- can seem worse when a person is stressed
- usually ease over the course of MND.

Ability to drive

Everyone with MND is required to inform the DVLA or DVA and their insurer immediately after diagnosis. They will usually be permitted to continue driving until weakness and/or cognitive change compromise safety. Judgement of continued driving capacity should be regularly discussed.



Dysarthria

80% of people with MND experience changes to their speech, which may become slurred or quieter. Complete loss of all speech is less common.²⁸

Muscle weakness and/or spasticity leads to a reduced range of movement in the:

- tongue
- lips
- facial muscles
- pharynx and larynx.²⁹

Reduced palatal elevation can lead to hypernasal voice quality.²⁹

Reduced breath support can lead to reduced volume or breathy voice quality.²⁹

The person should be referred to a speech and language therapist for an assessment and to access support.

Information to share

Notepad for people with speech difficulties

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

Information sheet 7C – *Speech and communication support*

Information sheet 7D – *Voice banking*

Telling people about MND

Information for you

Communication, speech and language support booklet

Information sheet P10 - *Voice banking*

Download from **www.mndassociation.org/publications** or contact MND Connect to order hard copies. Call 0808 802 6262 or email **mndconnect@mndassociation.org**

Impact

- Progressive difficulty with articulation, slurred speech and/or loss of volume.
- May rapidly lead to anarthria, even though limb function may be maintained for many months.²⁹

Support

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 voice banking and strategies for communication
- arrange for assessment and provision of Augmentative and Alternative Communication (AAC).

Low-tech:

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

High-tech:

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

An OT/neuro rehabilitation service/orthotics service can assist with advice on:

- seating, positioning, wrist and head supports
- switches and pointers
- mobile arm supports and tables to access communication aids
- environmental controls.

Dysarthria may exacerbate emotional reactions, including:

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

Allow time to explore and discuss the above issues.

Be aware that cognitive impairment (see page 54) can cause problems with communication and the ability to learn and use alternative communication methods.²⁹

Communicating with someone with MND

Do:

- find out how the person with MND prefers to communicate and any equipment they like to use
- 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 or thumbs up/down
- make sure any necessary communication aid is available and the person with MND is in the best position to access it
- have a pen and paper handy so that the person, if they are able, can write down any appropriate words: this helps to reduce frustration and misunderstanding
- sit face to face and watch the person's eyes, lips and gestures: unspoken communication is important
- make sure the environment is conducive to communicating: check lighting and reduce background noise where possible

- ensure the person with MND is comfortable and is the focus of communication
- allow the person time and create a relaxed atmosphere – give them time to process what is being said, and to formulate their reply
- 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
- 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 24).

Try not to:

- alter the rate or sound of your speech, unless the person with MND has asked you to, or a speech and language therapist has recommended that people should speak more slowly to the person
- finish the sentences of the person with MND, unless they ask you to, and avoid interrupting them
- use family members or carers as a translator for the person with MND, unless it is clear that this is what the person with MND wants
- use open questions. Instead use closed questions which can be answered with yes, no, or a single word.

Dysphagia

Swallowing problems affect at least two thirds of all people with MND during the course of the disease.³⁰ When a patient presents with this symptom, deterioration is inevitable and referral to a dietitian and speech and language therapist 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 almost never a direct cause of death.¹

Information to share

Information sheet 7A – *Swallowing difficulties*

Information sheet 7B – *Tube feeding*

Eating and drinking with MND – information on how to adapt food and drink, as well as easy-swallow recipes.

Information for you

Information sheet P8 – *Dysphagia in MND*

Download from **www.mndassociation.org/publications** or contact MND Connect to order hard copies. Call 0808 802 6262 or email **mndconnect@mndassociation.org**

Cause

Muscle weakness and/or spasticity leads to a reduced range of movement in the:

- tongue
- lips
- facial muscles
- pharynx and larynx.²⁹

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 and/or form a bolus with the tongue
- weak or absent swallow reflex
- food not moving to the back of the mouth or spontaneously moving down the throat due to a weak or absent swallow reflex, or an 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.³¹

This can result in:

- sialorrhoea (drooling due to impaired swallowing of saliva rather than excess production), see page 31-34
- 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
- social impact - the person may feel unable to eat with other people due to dysphagia or drooling
- constipation, see page 38-39.

Note: Weight loss can also occur from wasting muscles and managing weight in a timely manner may be important in relation to other interventions eg gastrostomy placement. See page 34-39.

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 fatigue³¹ affecting the ability to use cutlery or cups independently. Respiratory symptoms and risk of aspiration and/or choking should also be assessed.

A dietitian can suggest management strategies, including altered meal patterns (eg eating little and often) and high calorie intake to help maintain weight. 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.

Speech and language therapists and dietitians can advise on gastrostomy feeding to help avoid excessive aspiration, used as a top up or to replace oral intake.

Where the patient is aware of risk, they may choose to still have small amounts orally for flavour and enjoyment. See *Enteral feeding* on page 36.

A physiotherapist or occupational therapist can advise on head and/or arm supports, seating and other equipment for positioning.

Dysphagia and cognitive change

Additional support will be needed for people with dysphagia and cognitive impairment.³

People with more severe cognitive changes may place too much in their mouth at one time and cram food, while others may eat more food than they need or have a preference for a particular and narrowed range of food, often sweet.³²

It may be more difficult for someone with cognitive change to follow advice on safe swallowing, so repeated reminders and supervised eating may be necessary.³ See page 54-58 for further information on cognitive change.

Saliva management

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

Patients can have either runny, thin saliva that drools out of the mouth or thick, tenacious saliva that is difficult to clear.⁸ It is crucial to establish the volume and viscosity of saliva, as medication to dry up saliva can worsen the situation if the saliva is already thick.^{3,8}

Information to share

Information sheet 7A – *Swallowing difficulties*

Information for you

Information sheet P3 – *Managing saliva problems in MND*

Information sheet P9 – *Oral suction*

Download from **www.mndassociation.org/publications** or contact MND Connect to order hard copies. Call 0808 802 6262 or email **mndconnect@mndassociation.org**



Medication for thin, runny saliva

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

- a trial of antimuscarinic medicine as the first-line treatment for sialorrhoea, eg hyoscine (tablets or transdermal patches – be aware of the possible side effect of confusion in the elderly)
- glycopyrrolate as the first-line treatment for sialorrhoea in people with MND who have cognitive impairment, as it has fewer central nervous system side effects
- atropine drops on the tongue may be used for short term relief.¹³

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

Botulinum toxin A

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

Be aware that increasing dysphagia may be a side effect of Botulinum toxin A, so some suggest only resorting to this treatment if the person already has an enteral feeding tube in situ. Botulinum toxin A should only be administered by specialist practitioners.

Suction

Portable oral suction units are helpful if saliva builds up in the mouth.⁸

Body position

Attention is needed to seating, general posture and head support³³ – 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 sialorrhoea³

- provide advice on swallowing, diet, posture, positioning, suctioning, hydration and oral hygiene – consult district nurse³
- consider treatment with humidification, nebulisers and carbocisteine. Carbocisteine is available as capsules or liquid, which can be administered via a feeding tube.^{3, 34, 35}

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

It can also be useful to suggest pineapple/papaya juices,¹³ or flavoured ice cubes. These juices contain bromelain/papain (respectively). These hygroscopic agents are also available as tablets from health food stores.

See page 41-42 for information on dealing with weak cough.

Dry mouth³⁶

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



Nutritional management

Monitoring nutritional intake and weight is important. People living with MND can face many challenges to taking adequate oral diet and fluids including:

- dysphagia
- respiratory insufficiency
- depression and/or anxiety
- cognitive impairment
- constipation
- fatigue
- social isolation
- physical difficulties buying, preparing and eating foods
- dislike of modified texture diet and fluids
- increased time taken to eat meals due to dysphagia and difficulties using cutlery
- reduced dexterity and strength affecting ability to hold utensils.



Around 50% of people living with MND are hypermetabolic,³⁷ with a resting energy expenditure estimated to be 10% higher than people without MND.³⁸ Raised energy requirements, coupled with the challenges people with MND face taking adequate diet, are the main causes of the high prevalence of malnutrition.

Functional consequences of weight loss/malnutrition include:³⁰

- body mass losses due to muscle wasting
- increased respiratory muscle weakness
- decreased strength and mobility,
- reduced energy levels
- impaired immune function, which can increase susceptibility to opportunistic infections
- decreased tissue viability and increased risk of pressure sores
- increased discomfort sitting or lying due to weight loss and loss of tissue 'padding' over bony protuberances
- decreased morale and increased anxiety for the person with MND and their family members
- reduced quality of life.

There is evidence that having a lower body mass index (BMI) at the time of diagnosis with MND,³⁹ and losing weight during the disease course, is associated with a poorer prognosis.⁴⁰ Close monitoring of a person's nutritional status is vital to ensure timely nutrition support interventions.

The entire multidisciplinary team can contribute to identifying the risk factors and treating malnutrition risk in people with MND:

- Early and ongoing involvement of a speech and language therapist to assess swallow function and to advise on strategies to reduce the risk of aspiration and choking on oral diet. Strategies include modifying the texture of the diet and fluids.
- A physiotherapist can facilitate the person to be in an optimal body and head position to take oral diet safely and efficiently.
- An occupational therapist can advise on adapted cutlery, plates and cups to help maintain a person's independence at mealtimes.

- A dietitian can assess, monitor and review the individual's nutritional status and to advise on personalised oral and artificial nutrition support interventions to enable the person's nutritional needs to be met.³⁰
- Psychologists can help address low mood or any difficulties the person is having adapting to the diagnosis or disease progression.
- Social workers and neurology case managers can support with health and social care, or to access benefits.
- The supporting clinicians (e.g. GP, neurologist, palliative care) can advise on interventions to manage the symptoms associated with MND that can present a risk to the person's nutritional status, such as sialorrhea, low mood or pain.

Enteral feeding

The placement of an enteral feeding tube is an option for people with MND who are unable to meet their nutritional requirements via the oral route.

The two main routes of enteral feeding are via the placement of a nasogastric (NG) tube or a gastrostomy feeding tube (see next headings).

Enteral feeding may be used to 'top-up' oral intake, or to meet the person's full nutritional and hydration needs. They can also be used for administration of medications.⁴¹

Nasogastric (NG) tube placement

NG tube placement may be used when enteral feeding is required immediately eg following an acute deterioration of dysphagia, to bridge the gap to the placement of a gastrostomy feeding tube, or when gastrostomy feeding tube placement is not possible. It is usually used as a temporary measure.

Additional considerations are the increased risk of tube displacement and blockage; and the need to confirm gastric position by aspirating gastric contents prior to administering any feed, water or medications via the tube.

Gastrostomy placement

Where placement is possible, long-term home enteral feeding is generally better tolerated via a gastrostomy feeding tube.

It is important that gastrostomy placement is discussed at an early stage, and revisited as issues present themselves because:

- respiratory insufficiency in people with MND can make the placement of a gastrostomy tube more complicated and risky, so this should be taken into account when considering the optimal timing of gastrostomy feeding tube placement
- prognostic outcomes improve if gastrostomy tube feeding is initiated before the person with MND has lost less than 5% of their body weight (from measurement taken at diagnosis).

Declining respiratory function or increased aspiration risk may be the main trigger to recommending an earlier placement of a gastrostomy feeding tube.



It is important to remind people with MND considering gastrostomy feeding tube placement that they can still continue to eat and drink orally, even after having a gastrostomy, if their swallow is safe and/or if they wish to. The speech and language therapist can support people with MND in making informed decisions about continuing oral diet, even if this involves accepting a degree of risk that may be associated with aspiration.

Where the risks associated with the gastrostomy procedure become too high and the option may no longer be open to the patient, discussions will need to be had with the person regarding the option to either place a NG tube or continue to eat and drink with accepted risks of aspiration, with input from a speech and language therapist.

The person with MND and their carers should be made aware of the practical care required when living at home on enteral feeding, including who will administer the enteral feed and care for the tube.

Some people will not want to have an enteral feeding tube, and their informed decision should be respected.³ Exploring the risks and benefits of the procedure will help the individual to make an informed choice.

Refusal or withdrawal of gastrostomy in specific circumstances may be included in an Advance Decision to Refuse Treatment. When intervention such as gastrostomy is discussed, offer the opportunity to discuss or review any existing ADRT, DNACPR or Lasting Power of Attorney (page 69-71).³

Constipation

Sphincter muscles are not normally affected by MND.⁴³ Changes in bowel function are usually the result of:

- physical inactivity
- reduced peristalsis
- low fluid intake
- reduced fibre intake
- use of analgesics
- weakened breathing
- weakness of pelvic floor/abdominal muscles.⁴⁴

Diarrhoea may be due to constipation with overflow.

Treatment

- Maintain hydration and assess fibre intake.⁴⁴
- Stool softeners and stimulants may help.⁴⁴ 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) may help and are readily available.⁴⁴

Information for you

Information sheet P8 – Dysphagia in MND

Information to share

Information sheet 7A – Swallowing difficulties

Information sheet 7B – Tube feeding

Eating and drinking with MND – information on how to adapt food and drink, as well as easy-swallow recipes.

Download from **www.mndassociation.org/publications** or contact MND Connect to order hard copies. Call 0808 802 6262 or email **mndconnect@mndassociation.org**

myTube - <https://mytube.mymnd.org.uk>

Developed by SITraN – the Sheffield Institute for Translational Neuroscience. This website includes information and video content on gastrostomy to support people with MND to make decisions on whether the intervention is right for them.

Respiratory symptoms

Information for you

Information sheet P5 - Managing respiratory symptoms in MND

Information to share

Information sheet 8A – *Support for breathing problems*

Information sheet 8B – *Ventilation for MND*

Download from **www.mndassociation.org/publications** or contact MND Connect to order hard copies. Call 0808 802 6262 or email **mndconnect@mndassociation.org**

myBreathing - <http://mybreathing.mymnd.org.uk>

Developed by SITraN – the Sheffield Institute for Translational Neuroscience. This website includes information and video content on assisted ventilation to support people with MND to make decisions on whether the intervention is right for them.

Exertional dyspnoea

Breathlessness, particularly on exertion is common in the later stages of MND, but may occur earlier.⁶ (For chronic respiratory insufficiency, see page 43).

Causes of breathlessness

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

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

Once the acute event has been treated, consideration can be given to long-term care, which may include assisted ventilation (see page 45).

Actions:

- relieve fear and anxiety with calm reassurance
- request an MND Just in Case kit (see page 42)
- recommend coronavirus, flu and pneumococcal vaccination⁴⁵
- refer to a consultant in respiratory medicine or palliative care team as close to the onset of respiratory symptoms as possible to explore options. Involve family in these discussions as appropriate.

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

Medication to consider:

- buccal midazolam⁴⁶ (liaise with palliative care team for dosage)
- sublingual lorazepam⁴⁶ (slower acting than midazolam and ineffective in those with excess saliva)
- oral morphine.⁴⁷

Laryngeal spasm

Impaired swallowing may predispose to this. It is often preceded by gastroesophageal reflux⁴⁸ and can be extremely distressing. It can prevent breathing or speaking and can be accompanied by stridor.⁴⁹ The person may feel like they are choking. This symptom usually settles over time.

Consider prescribing lorazepam for acute episodes.⁴⁶ A speech and language therapist can offer advice on managing laryngeal spasm.

Weak cough

Peak Cough Flow (PCF) is an objective measure of a weak cough. PCF > 270 is normal. PCF < 160 means the cough is ineffective and adjuncts are needed to raise the PCF over the 270 threshold and make the cough effective. A PCF between 160-270 indicates that a patient should be referred to a specialist respiratory physiotherapist for review.

The NICE guideline on MND³ recommends the following cough augmentation techniques for people with MND who cannot cough effectively:

- unassisted breath stacking and/or manual assisted cough
- assisted breath stacking (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
- use of a mechanical insufflation/exsufflation machine (sometimes known by the brand name CoughAssist), if assisted breath stacking is not effective and/or during a respiratory tract infection.

These techniques can enable expectoration of phlegm or mucus from the throat or lungs, thus reducing the likelihood of A&E admission and emergency intubation.⁶ See page 32-33 for information on addressing thick secretions. For management techniques consult a respiratory 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 can help reassure 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 5) for a named person with MND. The kit is supplied free of charge.
2. The GP and district or community nurse agree on a plan to advise and support the carer in the use of the kit.
3. The GP prescribes medication to be supplied with the kit.
4. The GP, or community or district nurse, gives the kit to the person with MND and ensures the carer knows how it can be used.

Respiratory insufficiency management

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

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

Bulbar weakness contributes to various respiratory complications, including increased risk of aspiration, weak cough and abnormal respiratory patterns.⁴⁸

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

Respiratory tract infection should be treated with early use of antibiotics.⁵⁰

Monitoring for signs and symptoms

The specialist MND team should monitor for ongoing respiratory symptoms so that timely interventions can be made to improve/maintain quality of life for as long as possible.³

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

The NICE guideline recommends that respiratory function tests, eg FVC (forced vital capacity), and/or SNIP (sniff nasal inspiratory pressure) or MIP (maximal inspiratory pressure) should be performed every two to three months by an appropriately trained professional. These tests monitor signs of respiratory impairment, rather than progression of the condition. SpO₂ may be used for those with severe bulbar involvement.

Results of these tests may give false reassurance, so ask about the following symptoms and signs.³

Symptoms	<ul style="list-style-type: none">• Breathlessness• Orthopnoea• Recurrent chest infections• Disturbed or non-refreshing sleep• Nightmares	<ul style="list-style-type: none">• Daytime sleepiness• Poor concentration and/or memory• Confusion/hallucinations• Morning headaches• Fatigue• Loss of appetite
Signs	<ul style="list-style-type: none">• Increased respiratory rate• Shallow breathing• Weak cough• Weak sniff	<ul style="list-style-type: none">• Abdominal paradox• Use of accessory muscles of respiration• Reduced chest expansion on maximal inspiration

Presence of one or more of these symptoms or signs above 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.⁵¹ 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.

Assisted ventilation

Information for you

Information sheet P5 - *Managing respiratory symptoms in MND*

Information to share

Information sheet 8A – *Support for breathing problems*

Information sheet 8B – *Ventilation for MND*

Information sheet 8C - *Withdrawal of ventilation*

Download from **www.mndassociation.org/publications** or contact MND Connect to order hard copies. Call 0808 802 6262 or email **mndconnect@mndassociation.org**

myBreathing - **<http://mybreathing.mymnd.org.uk>**

Developed by SITraN – the Sheffield Institute for Translational Neuroscience. This website includes information and video content on assisted ventilation to support people with MND to make decisions on whether the intervention is right for them.

Non-invasive ventilation (NIV)

This is a method of providing ventilatory support to a person with MND using a close-fitting mask that covers the nose and/or mouth. The ventilator detects when the person with MND tries to take a breath and delivers an extra flow of air to increase the volume inhaled. The machine can also be set on different modes, for example, to give extra breaths.

It is important to use the most appropriate interface (mask or mouthpiece) based on the person's needs, lifestyle factors and safety, and experimentation may be needed.

When starting NIV, ideally there should be gradual acclimatisation during the day when the person is awake. Regular treatment at night will then usually begin. The time spent using the machine will build up as necessary.

Potential benefits of NIV:

- it can ease symptoms caused by weakened breathing muscles, such as shortness of breath, disturbed sleep, daytime sleepiness or fatigue and early morning headaches – however, if the patient stops using the machine, the symptoms will return
- it can increase survival time
- it is portable and can be used in many places, including at home and on the move, if powered by battery
- surgery is not needed for NIV
- although the person is likely to need more support over time, NIV care is generally quite easy to manage in the earlier stages of MND
- a range of available masks means the person can try out different options to establish what suits them best
- in some cases, using NIV can add volume to the person's voice if this has become weak.



For someone who has severe bulbar impairment, NIV may have limited benefits.⁶ If a person has severe bulbar impairment or cognitive problems that may be related to respiratory impairment, a trial of NIV may only be considered if they may benefit from an improvement in sleep-related symptoms, such as quality of life during the day due to sleepiness, or correction of hypoventilation.³

Potential issues with NIV:

- it can cause discomfort from air flow leaking around the mask and irritating the eyes
- pressure of the mask on the skin can cause sores
- wearing a mask can cause claustrophobia, which in some cases may lead to anxiety or panic
- it can cause a blocked nose, a runny nose or dry mouth
- the person may need support with eating or drinking while using the equipment, as the flow of air may cause an increased risk of aspiration
- it can cause the person to swallow more air than usual, which can make their stomach feel full and uncomfortable, meaning it may take more effort to breathe
- it can take a while to get used to the feel of the equipment and flow of air
- care needs can be complex while using NIV in the later stages of the disease – the person may be unable to fit or remove the NIV mask themselves, and a team of night carers may be needed
- communication - the flow of air can disturb the rhythm of natural speech and the mask may make it harder to hear what the person is saying, particularly if they have a weak voice
- although NIV machines are generally quiet, the noise can take some getting used to and may keep the person awake at night at first, and anyone they share a room with.

Tracheostomy ventilation^{52, 53, 54}

Tracheostomy ventilation is where a tube is inserted into the person's windpipe through an opening in the neck. This is attached to a ventilator which is triggered by the person's breathing. As breathing muscles weaken, the ventilator will deliver air if the person's breath is not strong enough to trigger it for a set amount of time.

Tracheostomy ventilation is not a common choice in MND⁵³ and may be logistically very challenging to undertake electively. However, the sense of being refused an elective tracheostomy may have a strong emotional impact on the person with MND and their family, as they may perceive that tracheostomy will increase their lifespan compared to NIV. Current research to support this is not strong, and no controlled trial has yet taken place to establish objectively whether tracheostomy directly extends life in MND compared to NIV. It is important to talk through an individual's expectations in case they are misguided.

Although it is occasional, the most frequent cause for tracheostomy ventilation is an unplanned procedure during an acute episode of ventilatory failure. This can happen as an initial presentation of the disease, with a diagnosis of MND being made in intensive care or other high dependency settings. There can be difficulties weaning the person from the ventilator long-term, and they may become dependent on it, particularly if they have severe bulbar weakness.

Some people may wish to specifically refuse this treatment in advance. Making decisions about future care and recording their wishes should be encouraged before acute deterioration occurs. See *Advance care planning* on page 69-71.

Potential benefits of tracheostomy ventilation:

- it can improve symptoms caused by weakened breathing muscles, such as shortness of breath, disturbed sleep, daytime sleepiness or fatigue and early morning headaches
- it may increase survival time but this is not always straight-forward
- it can be used where NIV cannot successfully support the person's breathing

- depending on the type of tracheostomy, it can help to protect the airway from aspiration and reduce the risk of lung infections
- it can be used to help with control of secretions
- a mask is not used, so mask related issues, such as claustrophobia, can be avoided.

Potential issues with tracheostomy ventilation:

Great thought must be put into supported discussions with the person with MND (and, where appropriate, their carer/family). It is important to explain:

- the impact on the family and the need to provide specialist care in the future, which may not be possible to deliver at home
- the possibility of requiring 24-hour care workers in the home
- the possibility of a lengthy stay in hospital following surgery
- care packages can take a long time to arrange, as staff need to be appropriately trained
- some types of tracheostomy will mean the person will no longer be able to speak
- it can increase secretions, which will require regular suction and can cause acute and distressing complications if the tube becomes blocked.

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 72-73 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. This is their legal right and enabling this is not the same as assisted dying, which is illegal. Stopping assisted ventilation should be discussed well in advance with the person with MND, family and any professionals involved, preferably during initial discussions about ventilation and as part of advance care planning – see page 69-71.³

Discussions therefore need to include whether the person may want to make an Advance Decision to Refuse Treatment (ADRT) – see page 69-71.

Withdrawal of assisted ventilation should be carefully planned so that symptoms can be managed to avoid distress or discomfort. Refer to medications for palliative care on pages 72-73.

Withdrawal of ventilation can be very challenging and it is important that symptoms and distress are minimised. Palliative care professionals often have experience in managing the process and may be called upon if needed for support and information. Guidelines are available from the Association for Palliative Medicine.⁵⁵

Oxygen

Respiratory muscle weakness leads to the retention of carbon dioxide (hypercapnia) rather than hypoxia. In this situation, assisted ventilation may be the most appropriate treatment, as use of oxygen can lead to further respiratory depression.⁵¹ Oxygen should not be used as a treatment for breathlessness alone.

Although oxygen is sometimes used for palliative or end of life care with MND, opiates may be more effective for terminal dyspnoea.⁴⁷ For more guidance, speak to your specialist palliative care team or a respiratory consultant with links to the neurology team.

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

Preparing for end of life - This symptom may raise concerns about end of life decisions and preparing for end of life, increased dependency and being unable to communicate. Offer people the opportunity to discuss or review any existing ADRT, DNACPR or Lasting Power of Attorney.³ See psychological support (page 58), palliative care (page 67) and advance care planning (page 69-71).

Alternative feeding - If respiratory function is deteriorating, enteral feeding may be discussed even if there are no immediate problems with swallowing or weight (see page 36-38).⁴² If possible, the carer should be involved in discussions as they will share the burden of any interventions.

Pain

People with MND usually feel surprisingly well in themselves despite their progressive weakness. The underlying disease process is not itself painful. However, MND leads to stiffness, cramps, reduced mobility and altered posture, all of which can result in significant secondary pain. The person with MND may not mention their pain⁵⁶ so it is important that healthcare professionals ask about it, both at the initial assessment stage and routinely during follow-up visits.

Information to share

Information sheet 6C – *Managing pain*

Information for you

Information sheet P11 – *Pain in MND*

Download from **www.mndassociation.org/publications** or contact MND Connect to order hard copies. Call 0808 802 6262 or email **mndconnect@mndassociation.org**

Pain may occur at any stage of MND, including early on, with no relationship between pain intensity and length of time since diagnosis.^{56, 57} Because it is usually a result of poor mobility, changes in posture, or reactions to changes in muscle tone, MND pain is more frequent in the limbs.

Pain significantly interferes with the quality of life for people with MND because of its impact on activity levels, mood, sleep, relationships, and general enjoyment of life.^{56, 58} Anticipatory prescribing is crucial to enable the patient to maintain control.⁵⁹

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

Physiotherapy is a key treatment strategy for pain in MND, and can prevent contractures, prevent and treat frozen shoulder, and relieve pain resulting from changes in tone or posture.^{3, 56} Occupational therapy assessment and the intervention of a wheelchair therapist can reduce pain resulting from postural weakness, or straining weak muscles.^{3, 56}

Medication for pain

For drug dosages, refer to British National Formulary 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).¹¹

For muscle cramps:³

- Consider quinine as first-line treatment for muscle cramps in MND.
- If quinine is not effective, not tolerated or contraindicated, consider a trial of low-dose baclofen instead as a second-line treatment.
- If baclofen is not effective, not tolerated or contraindicated, consider gabapentin.

For muscle stiffness, spasticity or increased tone:³

- Consider baclofen, tizanidine or gabapentin to treat severe spasticity in people with MND.
- Take care that the dosage of muscle relaxants is carefully adjusted to avoid increased weakness and decreased mobility due to the loss of the splinting effect of stiff muscles.¹¹ Sometimes there may also be unwanted sedation, so consider prioritising the overnight period for these treatments with no (or lower) daytime dosing.
- If these treatments are not effective, not tolerated or contraindicated, consider referral to a specialist service for treatment of severe spasticity.
- Check whether the patient is taking a statin, and whether this can be stopped, as muscle soreness and weakness may be side effects.

Opiates may be used for pain relief and can also be used for symptomatic treatment of dyspnoea and coughing.¹¹ With careful titration, excessive drowsiness and respiratory depression can be avoided.

Skin sensitivity

- Good skin and pressure care is vital.⁶¹ Someone with MND may be aware when they need to turn or move, but may need help to adjust their position. This must be done with extra care.
- Consider equipment for skin sensitivity relief, such as a bed cradle to relieve the weight of bed clothes, lightweight bed clothing and a pressure-relieving mattress and cushions.
- Advise warm socks for cold feet.
- Itching can be a problem for some people with MND and may respond to emollients or antihistamines.

Oedema (fluid retention)

- This may largely be related to restricted activity and posture or to an underlying health condition, which should be treated accordingly.⁶²
- 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.¹³
- In some areas, referral to the lymphodema service may be an option.

Cognitive change and frontotemporal dementia

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

- those unaffected by cognitive change - about 50%; this proportion decreases to 20% in the more advanced stages of the disease⁶³
- those with mild cognitive or behavioural change, typically deficits in executive function, language, motivation (apathy or impulsivity) and loss of social cognition - about 35%^{64, 65, 66}
- those with frontotemporal dementia (FTD), either at the same time or soon after the motor features of MND - up to 15%.⁶⁷

It is important to exclude other possible causes of cognitive and behavioural change, such as infections, medication side effects, pain, respiratory problems or mental health difficulties.

Information to share

Changes to thinking and behaviour with MND

Information for you

Cognitive change, frontotemporal dementia and MND

Download from **www.mndassociation.org/publications** or contact MND Connect to order hard copies. Call 0808 802 6262 or email **mndconnect@mndassociation.org**

Cognitive impairment in MND

People with MND may experience cognitive changes not classified as dementia, ranging from mild to moderate or more severe in some cases.⁶⁸ Typically they will lack awareness or insight into their problems or their impact, so engaging with their carers is important. Emotional lability is not a sign of cognitive impairment but should be considered a motor pathway dysfunction.

Cognitive impairment may manifest as a deficit in verbal fluency.

Some people may have primary language problems that manifest as reduced verbal output and failure to initiate conversations. The person may respond in short phrases or single words.⁶⁸

People may experience problems with:

- learning new tasks, including the use of equipment (such as gastrostomy or NIV)
- making decisions, planning for the future
- concentration.

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

Behavioural changes⁶⁸

Behavioural impairment is an increasingly recognised feature of MND and changes may include apathy, egocentric or selfish behaviour, loss of interest, disinhibition, irritability, aggression but with absence of insight into these changes. There may be a loss of food repertoire with a preference for sweet foods and a tendency to cram. These behaviours may meet some of the criteria for frontotemporal dementia.

People with MND may also find it difficult to recognise emotions in others (social cognition) and have altered sensitivity to social and emotional cues. This can be particularly distressing for carers who may find it hard to motivate the individual or worry that they are not performing well enough due to lack of positive feedback.

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. It is unlikely that someone experiencing these changes will know this is happening or can acknowledge the change. More often, it is the family or carer who sees behavioural change, and this can be very challenging for them. Behavioural change can affect family relationships and increases the burden on carers.

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) 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. The ECAS tool can be found at <http://hdl.handle.net/1842/6592>

Frontotemporal dementia (MND-FTD)

The following explanation defines MND-FTD and the combination of symptoms that may indicate frontotemporal dementia.⁶⁸

Cognitive change in MND-FTD is characterised by:

- 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 difficulty 'taking in' information, rather than the memory problems associated with Alzheimer's disease, for example
- visuospatial functions are preserved.

Behavioural change in MND-FTD

More severe changes in behaviour associated with FTD may occur before cognitive impairment. The person may experience:

- significant personality change
- disinhibition and impulsivity (socially inappropriate behaviour)
- perseveration (continuing to conduct an activity that is no longer appropriate)
- change in eating behaviour (fixation with single, often sweet foods)
- marked loss of emotional understanding (appearing egocentric/selfish)

- becoming withdrawn (apathy/failure to initiate)
- stereotyped/ritualistic behaviour.

Managing cognitive change/MND-FTD

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

Your role as a professional

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

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

Impact on decision-making

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

Ability to drive

Although there may be no physical barrier to driving in early MND, lack of insight into danger is sometimes an issue for a small minority, which may affect their ability to continue to drive safely.¹³

Psychological support

Psychological wellbeing is an important determinant of quality of life. MND has a significant psychological impact on people living with the disease and those close to them, including children.¹⁰

MND is characterised by a series of losses, with associated grief and bereavement. Feelings of anxiety, depression and isolation are common,^{10, 69, 70} particularly in the first year after diagnosis.⁷¹

A feeling of hopelessness, which is more likely to arise soon after diagnosis, is the most common marker of psychological morbidity in MND.⁷² The psychological experience of MND will be influenced by age, gender, stage of life, cultural and educational background,^{73, 74} spiritual and religious beliefs, and psychosocial circumstances.

Excellent psychological support is essential to holistic, patient-centred care. It can greatly influence quality of life, reduce risk of anxiety and depression, diminish caregiver burden and deliver significant savings for the healthcare service.⁷⁵

Information to share

Information sheet 6B – *Complementary therapies*

Emotional and psychological support: for people with or affected by MND

Making the most of life with MND – includes information to help people continue activities, interests and hobbies to maintain quality of life

Telling people about MND – information on how to open conversations about an MND diagnosis with family, children, friends, colleagues and professionals

Download from **www.mndassociation.org/publications** or contact MND Connect to order hard copies. Call 0808 802 6262 or email **mndconnect@mndassociation.org**

Before the diagnosis

Anxiety and low mood may be triggered by:

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

After diagnosis is confirmed

- give sufficient information to allow the person with MND to understand what is happening without overwhelming them
- provide space for them, their families and carers to express thoughts and feelings openly and without judgement
- give information that can help them maintain control and make plans for the future
- feelings about the illness may change; allowing time to express thoughts, feelings and concerns regularly can help identify information and support needs
- check preferences for involving family members/carers; offer opportunities for the person with MND, and their partner if relevant, to rehearse how they might tell other family members, particularly children, about the diagnosis and prognosis
- be aware that the type, quantity and timing of information and support required may be different for each individual, for example carers may seek out information earlier, to help them prepare for their role
- determine what the person with MND has understood and retained of the information provided
- where possible, direct them to a specialist psychologist or someone from the local mental health team to provide ongoing support and information; signpost to other sources of support including counselling services, a local hospice, local support groups, the MND Association website and online forums. See page 86 for details of the MND Association's care information.

Support as the disease progresses

As MND develops, its impact will be influenced not only by how quickly it progresses, but also the area of the body involved.¹⁰ Adapting to the changes in physical function can be particularly challenging for patients with a more rapid rate of progression of the disease as there is less time available for psychological adjustment.

- Ensure people with MND and their families understand the likelihood and implications of respiratory problems. This enables everyone to explore the options and likely outcome, thus avoiding uninformed last-minute decisions, eg prolongation of unacceptable quality of life.
- Consider the psychosocial effects of communication problems and not being able to share thoughts and feelings.
- Consider having conversations with about the emotional/ psychological impact of MND from the outset.

Earliest possible referral to the palliative care team is recommended, as they can help provide support on a range of issues, from symptom management to psychological and spiritual support.

Often patients associate palliative care with immediate death and this is a cause of anxiety initially. However, the benefits to the patient and their family are immense and palliative care teams are vital to supporting the patient /family through the process from diagnosis to death.

We produce resources on end of life planning, both for people affected by MND and health and social care professionals (see page 72). Information on end of life care should be shared sensitively, as it can be upsetting for the person and their loved ones. However, fear of the unknown and the confusion that can happen if end of life arrangements are not in place as preferred, can lead to greater distress.

Emotional reactions

Fears may include:

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

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

Denial

Denial may form part of a coping strategy for some people. Early on this may be a useful means of adapting to difficult circumstances, but if denial persists it can contribute to higher levels of anxiety and depression.⁷⁶ Sometimes the individual will pursue multiple neurological opinions.



Hopelessness

Feelings of hopelessness are common, especially soon after diagnosis and may be linked to the recognition that many of life's expectations will never be realised.⁷⁶ Time and a safe environment are required to explore these issues.

Anger

Many people with MND are concerned about how angry they feel as a result of frustration and loss of control. Anger may be displaced onto others and may occur at any stage of the illness. Acknowledging and discussing such feelings may be helpful. If anger is existential in nature, spiritual support may be appropriate if the family has a particular faith or belief system.

Anxiety and depression

Persistent and disabling symptoms of anxiety and depression need to be distinguished from appropriate responses to the difficulties faced by people living with MND.

The symptoms of anxiety and depression may mimic some of the physical consequences of MND making identifying them more challenging. These include sleep disruption, diminished appetite, fatigue or weakness, poor concentration and irritability.¹⁰

Screening for anxiety and depression, using tools such as the Hospital Anxiety and Depression Score (HADS), may assist in identifying and/or monitoring symptoms in both patients and carers.

Antidepressants and/or anxiolytics may be beneficial for some people. These include selective serotonin reuptake inhibitors (SSRIs) such as sertraline and citalopram. These may take several weeks to have an effect.⁷⁷

Apathy

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

Sleep disturbances

Treatment of anxiety and depression can often result in quite rapid improvements in sleep quality.⁷⁷ This in turn can improve both physical and psychological wellbeing. Sleep disruption due to other factors (including pain, cramps, spasm, breathing difficulties and choking) can have a negative impact on quality of life and should also be treated.

Emotional lability (pseudobulbar affect)

Emotional lability (emotionality or emotional reflex hypersensitivity) presents as unwanted crying or laughing of acute onset that is recognised by the individual as inappropriate or disproportionate to the stimulus.⁹ In some cases, this can be extreme, although it often stops as quickly as it starts. It can nonetheless be very disabling for patients and difficult for those around them to witness. Some people find understanding the symptom helps them to manage the impact, but others find it limits where they go and what they do.

This occurs as a consequence of upper motor neurone involvement in the corticobulbar tracts in patients with MND. It is independent of depression or cognitive decline⁷⁸.

The aim of treatment for emotional lability is to reduce the frequency and severity of episodes. Treatment may include the off-label use of antidepressants which target the neurotransmitters noradrenaline, serotonin and glutamate. Tricyclic antidepressants such as amitriptyline, and selective serotonin reuptake inhibitors (SSRIs) including fluoxetine, citalopram and sertraline may be considered. The doses used tend to be lower than those used for depression.

Impact on professionals

MND poses many challenges for professionals who may experience feelings of frustration, powerlessness, inadequacy and sadness. It highlights attitudes to issues related to disability, quality of life and measures taken to prolong life. Good multidisciplinary teamwork is necessary to provide support and opportunities to discuss concerns and responses to difficult situations.

Providing evidence for benefit applications

People with MND may apply for disability and/or employment benefits.

If someone with MND is applying for, or being reassessed for, Attendance Allowance, Personal Independence Payment (including transfer from Disability Living Allowance), Employment and Support Allowance or Universal Credit, their application may be considered for fast-tracking under provisions called 'Special Rules'. These rules make the application process less burdensome and speed up access to the highest level of financial support.

Information to share

Information sheet 10A - *Benefits and entitlements*

Information sheet 10G - *Support for families with children*

Information for you

Information sheet P5 - *Providing medical evidence for benefit applications*

Download from **www.mndassociation.org/publications** or contact MND Connect to order hard copies. Call 0808 802 6262 or email **mndconnect@mndassociation.org**

MND Association Benefits Advice Service

Our trained advisers can help identify benefits a person may be able to claim if living with MND or a carer. The service is available for people living in England, Wales or Northern Ireland. Visit **www.mndassociation.org/benefitsadvice** for further details.

GPs can help by completing form DS1500 or SR1 to show the person with MND is eligible for these rules:

Form DS1500: To be used if applying for Attendance Allowance, Personal Independence Payment, or for a reassessment of Disability Living Allowance. This form refers to 'Special Rules for Terminal Illness'.

Form SR1: To be used if applying for Employment and Support Allowance or Universal Credit. This form refers to 'Special Rules for End of Life'.

If a person with MND is aware that Special Rules exist, they can usually start the process during their initial phone call by telling the call handler that they want their application to be considered as a Special Rules case. Once they have made it clear they wish to claim under Special Rules, they will be asked whether they have, or are going to get, a DS1500 or SR1 form, which they will be asked to send in. This can be completed by a GP. The claim will then be processed by a Department for Work and Pensions (DWP) assessment provider and a decision made by the DWP.

It is possible for a DS1500 or SR1 form to be completed on someone's behalf without their knowledge. This can be helpful if your patient is not up to dealing with the terminal nature of their condition, if they cannot handle the claims process, or if they have not yet been told the full nature of their condition.

How MND qualifies for the Special Rules

MND is a terminal, rapidly progressive disease. As a result, people with MND will pass the diagnosis element of Special Rules on the DS1500 or SR1 form, as it is a terminal condition. However, the prognosis element can be problematic.

It is important to remember that MND is unpredictable. The forms only ask if there is a reasonable expectation that their prognosis could be:

- six months if applying for Personal Independence Payment, Disability Living Allowance or Attendance Allowance - use form DS1500
- 12 months if applying for Employment and Support Allowance or Universal Credit - use form SR1.

Note: In Northern Ireland, the prognosis timescale is 12 months for all applicable benefits. All new applications are on the new SR1 form but there will be a transition period where DS1500s are still accepted while the process is fully rolled out.

The guidance for professionals completing form SR1 is still in development at the time of writing. The diagnosis criteria does not use the word “terminal”, but this is implied. The rules state that the following indicators may suggest that a patient is eligible under the Special Rules:

- advanced, progressive illness
- worsening symptoms despite optimal treatment or management
- (severe) degenerative condition
- deterioration of incurable condition
- rapid decline
- metastatic disease
- inoperable cancer
- severe frailty
- death is imminent, death is inevitable
- a high risk of sudden death with an underlying deteriorating condition.

Completing an application using the Special Rules does not mean that someone is going to die within the specified timeframe, 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.

As a third of people with MND die within a year of diagnosis and more than half die within two years, a Special Rules application should be actively considered for all people diagnosed with MND. There may be an exception where MND presents with noticeably slow progression or limited symptoms.

As the prognosis timescales are 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 specified period. Delays mean that someone may be living without any kind of financial support for many months.

Palliative care

People with MND have particularly complex care and support needs in the final stages of life. Because MND can be rapidly progressive, it is important that people with the condition are offered referral to specialist palliative care services at an appropriate time. The appropriate time for referral will vary, depending on factors such as the availability of services and the speed of progression of symptoms.⁸⁰

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.

Information to share

End of life guide

Information sheet 3D – *Hospice and palliative care*

Information sheet 8D – *Withdrawal of assisted ventilation*

Information sheet 14A – *Advance decision to refuse treatment (ADRT) and advance care planning*

Information for you

A professional's guide to end of life care in MND

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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 does not mean that death is imminent, as many people view hospices purely as places where people die.

- Discussing advance care planning (see page 69-71) ensures the patient's wishes are respected at each stage of the disease.
- All people with MND should be placed on local palliative care registers/ lists/co-ordination systems, where these exist – eg the Gold Standards Framework, or Electronic Palliative Care Co-ordination Systems (EPaCCS). This will ensure that future care can be planned and co-ordinated, and that guidance and support are given to both the patient and the GP.

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.

The NICE guideline on MND³ recommends offering opportunities to discuss preferences and concerns about care at the end of life at certain trigger points, such as:

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

Be sensitive about the timing of discussions and consider the person's communication ability, cognitive status and mental capacity.³

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

Recording discussions

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

Advance care plan

An advance care plan can be wide-ranging, covering all aspects of day-to-day care including, as examples, who should provide personal care and how they should do it, special requirements for food and drink, decisions about artificial feeding, ventilation, resuscitation, use of antibiotics, place of care and preferred place of death, and other issues such as care of dependants or pets in an emergency. Also included in this may be leaving special messages for friends and loved ones, memory boxes or books for children, and digital legacies.

The process for advance care planning, the documentation used and who needs to have this documentation will vary between areas.

Advance statement

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

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

Advance Decision to Refuse Treatment (ADRT)

An ADRT allows people to make decisions to refuse treatments or have treatment withdrawn, including treatments that sustain life, such as assisted ventilation, hospital admission or antibiotics in specific circumstances in the future.

Everyone involved in the care of a person with MND should have access to the current ADRT. Emergency staff should be able to access it easily. When completed correctly, an ADRT is legally binding in England and Wales.⁸¹

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. They should understand that it prevents cardiopulmonary resuscitation and is not a refusal of care more widely. If included in an ADRT, with clear instructions about when it should be applied, refusal of CPR is legally binding in England and Wales.

Power of Attorney

A Lasting Power of Attorney (LPA – England and Wales) or an Enduring Power of Attorney (EPA – Northern Ireland) is a legal document that allows a trusted person to make decisions on another person's behalf. An LPA can cover decisions about financial and/or specified health/care related matters, while an EPA covers financial affairs and property but doesn't cover health or care decisions.

The person with MND must have capacity to understand and make the decisions when they complete the LPA or EPA. It will not normally be used until they do not have capacity or cannot communicate their own decisions.^{82, 83}



While someone can complete their own LPA or EPA, it can be complex, especially if the individual has detailed finances, taxation, dependent children etc, in which case legal support is recommended.

Decisions outlined in LPAs and EPAs override any previous ADRT, but a new ADRT can be written once the LPA/EPA is in place.

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.

Advance care planning checklist

- ☐ Is someone helping the person with MND to develop an advance care plan?
- ☐ Has an 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 the future?
- ☐ 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 42 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 DNACPR or 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?

End of life

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.¹³

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

Information to share

End of life guide

Information sheet 3D – *Hospice and palliative care*

Information sheet 8D – *Withdrawal of assisted ventilation*

Information sheet 14A – *Advance decision to refuse treatment (ADRT) and advance care planning*

Information for you

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Medication at end of life

Check all symptom control:

- pain – patients may experience severe discomfort and/or distress, particularly in the later stages. Use of opiates may be an option with careful titration to avoid excessive drowsiness and respiratory depression⁸⁴
- pressure care
- respiratory
- dysphagia
- excess oral secretions or dry mouth

- insomnia
- anxiety/depression
- restlessness/agitation
- bowels and bladder
- reassess emotional and practical needs of the carer and family.

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

- antimuscarinics, such as hyoscine hydrobromide and glycopyrrolate, to reduce saliva and lung secretions⁴⁷
- benzodiazepines, such as midazolam, to manage breathlessness that is exacerbated by anxiety, and other medications to reduce anxiety/ terminal restlessness, such as haloperidol or levomepromazine^{3, 47, 85}
- opioid analgesics to reduce cough reflex, relieve feelings of breathlessness, and therefore fear and anxiety. They can also control pain¹¹
- anti-emetics for nausea.⁴⁷

Carers and family should be kept fully informed of how medications might affect the individual. For example, if sedation is likely to prevent further effective communication, the family may wish to have certain conversations as soon as possible.

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.⁴⁷

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

Communicating with the person with MND

This may become extremely difficult as the person reaches end of life, but even if they are unresponsive, every attempt should be made to maintain communication, particularly if assessing mental capacity and ability to make decisions. Eye movements, single response answers to closed questions, picture/alphabet boards or other communication aids may be used as appropriate (see pages 24-28).²⁹

Supporting family and carers at this stage

MND is unpredictable and may progress rapidly, with death occurring more quickly than anticipated.¹³ 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 sensitively discuss end of life with the family as early as possible can be positive, as it allows them to look at their lives together, achieve things that are important to them and tie up loose ends.



Supporting families and carers

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

Information for you

Supporting children and young people close to someone with MND

Information to share

Caring and MND: support for you

Finding your way with bereavement

Sex and relationships

Telling people about MND

Information sheet 10B – *What is social care?*

Information sheet 10G – *Support for families with children*

Information for children and young people

So what is MND anyway? - A comprehensive guide about MND for young people, including young carers. Also available as a web app.

When someone close has MND - An interactive workbook to help a trusted adult communicate about MND with children aged between four and ten years.

Why are things changing? - Our storybook about families affected by MND for children aged between four and ten.

MND Buddies – Our activity hub for children aged four to ten. Visit www.mndbuddies.org

Download from **www.mndassociation.org/publications** or contact MND Connect to order hard copies. Call 0808 802 6262 or email **mndconnect@mndassociation.org**

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

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

Not everyone will have a carer

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

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

Psychological support for carers and family members

The carers and families of those living with MND often experience considerable psychological and emotional distress.¹⁰ 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.¹⁰

- Allow carers time to talk about the impact MND has on them and their family. It may be more appropriate to talk to the carer separately from the person with MND, so they can be more open about their needs.
- Let them express and explore their feelings. By asking questions, you will find out how safe it is to talk about particular issues. A carer may not discuss these feelings and fears spontaneously.
- Reassure the carer that it is natural to have intense and conflicting emotions about the situation they are in.
- MND involves many losses: carers need time to grieve for the past and how things were, the present situation and their loss of a future with the person they care for.

- Discuss with the carer/family what type and frequency of support would help reduce anxiety and fear. Consider referral for additional psychological support. There may be phases of the illness where this needs to be reviewed, particularly when significant changes happen.
- Carers may benefit from being in contact with other carers of people with MND. The MND Association can sometimes provide a volunteer Association visitor. Volunteer-led MND branches and groups run local support meetings for carers and families. It might help to talk to other carers through our online forum at **<https://forum.mndassociation.org>**
- Children and young people in the family may benefit from a referral to a specialist service such as the school nurse or Child & Adolescent Mental Health Service (CAMHS) in order to access a range of appropriate therapies, such as emotional or psychological support or practical help with their education.

MND forces changes in roles and relationships. Consider:

- mapping the social support system of the family using a genogram
- meeting the various information needs of all family members
- counteracting isolation of individuals and promoting awareness of each other's needs
- offering early opportunities for short periods of respite to prevent over-dependence on a single carer
- other concurrent life events, such as moving house, children leaving home, financial difficulties etc and linking the family with supportive services if appropriate
- exploring the way in which illness changes relationships and acknowledge not only the strain of this, but also the ways in which they stay the same or are strengthened
- creating opportunities for people to express negative feelings without feeling guilty
- how carers may experience exhaustion from the caring role, exacerbated by powerlessness to prevent suffering and further deterioration.

Carer's assessments

Carers who provide a substantial amount of care on a regular basis have a legal right to an assessment of their needs, carried out by their local social services or Health and Social Care Trust. The assessment should anticipate need, so that crisis points can be avoided. The results of an assessment must be outcome-based and reviewed on a regular basis.

Respite

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

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



Young carers

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

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

Bereavement

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

Our online bereavement hub includes guidance on practical and emotional bereavement support available from the MND Association and other organisations. Visit **www.mndassociation.org/bereavement** for further information.

Action points for GPs

- Make sure that a person's role as a carer is noted on the record of the person with MND, and on their own GP practice record (if relevant).
- Has the person had a carer's assessment? Has a young carer had an Early Help Assessment or a young carer's needs assessment?
- Does the carer have the support and knowledge they need to carry out a caring role?
- Has a referral been made to an appropriate specialist agency such as school nurse or CAMHS, to support any children or young people?
- Have the needs of the whole family been considered?

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This guide has been endorsed by the
MND Professionals' Community of Practice.

How we can support you

MND Connect

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

Email: mndconnect@mndassociation.org

Phone: 0808 802 6262

Benefits Advice Service

Qualified advisers can help identify benefits the person with MND and carers may be entitled to, and advise on how to claim. This service is confidential, impartial and free.

www.mndassociation.org/benefits-advice

Information resources

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

www.mndassociation.org/publications

MND Association website

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

www.mndassociation.org/professionals

Education

Our education programme is designed to improve standards of care and quality of life for people living with and affected by MND.

www.mndassociation.org/education

MND Professionals' Community of Practice

A peer led group of health and social care professionals encouraging and supporting the development of good care for people living with, or affected by, MND.

www.mndassociation.org/cop

Support grants and equipment loan

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

www.mndassociation.org/getting-support

Research into MND

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

www.mndassociation.org/research

MND Register

The MND Register aims to collect information about every person with MND to help researchers focus their knowledge and expertise in the right areas.

www.mndregister.ac.uk

Regional staff

We have a network of regional staff with specialist knowledge of MND. They work closely with local statutory services and community care providers. Contact MND Connect for further information.

Email: mndconnect@mndassociation.org

Phone: 0808 802 6262

MND care centres and networks

We fund and develop care centres and networks across England, Wales, and Northern Ireland, which offer specialist MND care.

www.mndassociation.org/care-centres

Branches and groups

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

www.mndassociation.org/branchesandgroups

Association visitors (AVs)

AVs are trained volunteers who provide one-to-one local support to people affected by MND.

www.mndassociation.org/associationvisitors

We value your feedback

We would greatly appreciate your feedback on this guide. Please visit **www.smartsurvey.co.uk/s/mndprofessionals** or email your comments to **infofeedback@mndassociation.org**

Visit our webpages for health and social care professionals:
www.mndassociation.org/professionals

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