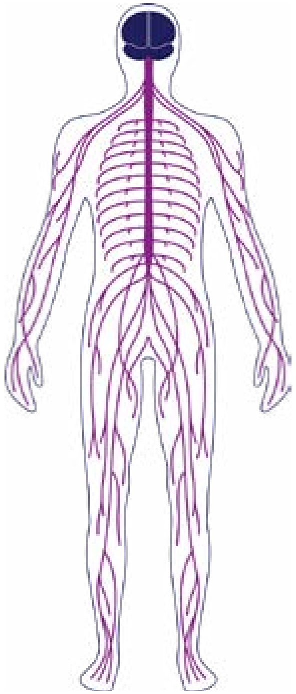
motor neurone disease association

MND Professionals' Community of Practice

A professionals' guide to end of life care in motor neurone disease

About MND

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



Would you like to find out more?

Contact our helpline MND Connect if you have any questions about MND or want more information about anything in this booklet.

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MND Professionals' Community of Practice

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

Introduction

Motor neurone disease (MND) is a progressive terminal disease that results in degeneration of the motor neurones, or nerves, in the brain and spinal cord.[1] There is no cure. A third of people with MND die within a year of diagnosis and more than half die within two years.[2]

This booklet is for all health and social care professionals working with people with MND. It aims to support professionals to discuss advance care planning, death and dying as soon as someone with MND is ready.

By understanding their needs, professionals can support people affected by MND to prepare for end of life.

Further information for you

MND in acute, urgent and emergency care

Information to share with people affected by MND

End of life: a guide for people with MND - This comprehensive and candid guide provides guidance for people with MND on how to plan ahead and communicate their choices to family and professionals.

Information sheet 14A - *Advance Decision to Refuse Treatment (ADRT) and advance care planning*

Information sheet 8C - *Withdrawal of ventilation with MND*

Understanding my needs – A record of someone's needs to help professionals provide appropriate support.

See page 39 for details of how to order our publications.

Talking about end of life

People with MND may have many concerns about the progression of the disease, the process of dying and death itself. They may not express their fears openly.

Choice and control is important. They 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.

Conversations may be more difficult if the person's speech becomes affected, or if they experience cognitive change, but every effort should be made to involve the person with MND as much as possible.

Timing discussions

Be sensitive about the timing of discussions and take into account the person's current communication ability, cognitive status and mental capacity. People with MND may expect a professional to raise the topic, or they may indicate, by the questions they use, when they are ready to have information.

The NICE guideline on MND recommends offering opportunities to discuss preferences and concerns for end of life at trigger points: at diagnosis, if there is a significant change in respiratory function, or if interventions such as gastrostomy or assisted ventilation are needed.[3] Other times may also be appropriate.

Topics to talk about

People with MND often want to talk about their fears and concerns, as well as the management of their symptoms. They may have clear views about how they want the latter stages of their illness managed. This may include decisions about:

- artificial feeding and ventilation
- resuscitation
- use of antibiotics
- place of care/death
- who they want to be involved in their care.

Time is needed for the person to feel comfortable enough to express their worries. It is important that concerns are taken seriously and acknowledged compassionately. Members of the multidisciplinary team should work together and negotiate towards prompt support and/or solutions wherever possible.

It is also important that the language should remain sensitive yet clear. Professionals should not be afraid to use words such as 'death' and 'dying' instead of euphemisms. This can be difficult for some professionals, but if the person with MND, and those close to them, are ready for it, this clarity is vital.

Recording discussions

If discussions are held and decisions are made, they should be clearly documented, ideally in a personalised advance care plan (see pages 10-17), and communicated to all relevant health and social care professionals.

Conversations discussing wishes, preferences and plans for future care should also be registered and shared on local palliative care registers/lists/co-ordination systems. Examples include the Gold Standards Framework, Electronic Palliative Care Co-ordination Systems (EPaCCS) and Recommended Summary Plan for Emergency Care and Treatment (ReSPECT).[4]

The person should be reminded to keep these documents somewhere emergency services can easily access them.

Reviewing choices

People may change their minds about the choices they make. The progression of MND may affect how they feel about potential interventions, so they should have the opportunity to review their wishes and alter their preferences.

Any written statements will need to be signed, dated and distributed to all who hold the existing documents, with previous statements destroyed or crossed through. Minor updates, such as a change of address, do not usually require a new document. An updated review should be sufficient.

Key actions

- The person with MND should have the opportunity to discuss end of life with a trusted healthcare professional. They should also be encouraged to talk with those closest to them.
- The specialist palliative care team (see page 20-23) can advise on the timing of end of life discussions. See page 9 for resources.
- The person's wishes regarding end of life care and preferred place of care/death should be discussed before the need is urgent or the capacity to communicate is limited and tiring. However, where communication and capacity exist, all decisions must be discussed with the person with MND before any action is taken.

Discussing suicide and assisted suicide

Living with MND can create fear about what will happen as the condition progresses. You may be asked questions about suicide and assisted suicide.

People with MND may consider suicide for fear of becoming a burden or due to other concerns about independence. Discussion is crucial in order to explore and understand these issues.

It is important to let the person know that thoughts of suicide are not unusual among people with MND.[5,6] Being able to explore the reasons for these thoughts, and knowing they are not alone can help.

If suggestions or solutions to concerns can be provided, thoughts of suicide may subside. Active plans to commit suicide should always be taken seriously. It is important to involve the patient's GP and also consider the use of suicide risk assessment tools to guide you.

This information explains what is and isn't allowed within the law (at time of publication).[7] It is not intended to replace legal advice or act as guidance to take any specific action, but simply to provide the facts.

More information can be found in section 13 of the MND Association's End of life guide (see next page).

It is **legal** for someone to:

- take their own life
- decline or discontinue life-sustaining treatments which they feel are not helpful or have become a burden. This is not assisted dying.

But it is **not legal** for someone else to:

- encourage another person towards suicide (including advising them how to do this)
- assist them with their suicide.

Health and social care professionals found to be assisting someone's suicide could face prosecution and up to 14 years' imprisonment.

A person with MND can influence how their care will be managed in the later stages of the condition using advance care planning (see pages 10-17). Within the law, they are able to record advance decisions to refuse or withdraw treatment (ADRT), in the event they become unable to make or communicate these decisions for themselves. A valid ADRT is legally binding (see page 12).

Medication cannot be used to hasten death. It may be provided as part of a package of palliative care to reduce pain and other symptoms at end of life.[8] It is important that these symptoms are treated in order to reduce distress at this stage.

Do not avoid the discussions or deny the person the chance to talk. If you don't feel able to do it, refer them to someone who can.

Liaise with the person's specialist palliative care team or MND care team for advice and support in managing conversation about suicide and assisted suicide, as they will have experience of this.

In tender situations, a helpful response is to ask, 'What makes you feel that way?' This both acknowledges the person's feelings and opens the door to deeper understanding.

Resources to support end of life discussions

The MND Association's End of life guide is a comprehensive booklet for people with MND and their families about end of life care and decisions. It may also support professionals to open sensitive conversations.

The guide covers subjects including:

- how MND progresses and the benefits of planning ahead
- how to manage end of life discussions
- what is likely to happen at end of life with MND
- advance care planning, advance decisions and the introduction and withdrawal of treatments for MND. See next chapter.

Finding your way with bereavement is a booklet which provides information on finding emotional and practical support when bereaved, and supporting young people who are grieving. See page 39 for details of how to order our publications.

Royal College of Physicians - Talking about dying: How to begin honest conversations about what lies ahead

This report supports doctors in conversations with patients after a progressive or terminal diagnosis. Search "Talking about dying" at **www.rcplondon.ac.uk**

Talk CPR

This website includes useful information and resources to support conversation about CPR with people with life-shortening conditions. Visit **http://talkcpr.wales** to find out more.

Advance care planning

Thinking things through, discussing with family and knowing that wishes have been recorded, gives many people peace of mind.

Information to share

Information sheet 14A - *Advance Decision to Refuse Treatment (ADRT) and advance care planning*

Information for you

Advance care planning for MND booklet

The *Universal Principles for Advance Care Planning*, published by NHS England, sets out six high level principles for advance care planning, with the aim that a consistent approach would enable people, their families, and professionals share the same understanding and expectations of advance care planning.

Download from **www.england.nhs.uk/universal-principles-for-advance-care-planning**

See page 39 for details of how to order our publications.

Advance care plan

An advance care plan can be wide-ranging, covering all aspects of day-to-day care, as well as future plans, for example:

- who should provide personal care and how they should do it
- special requirements for food and drink
- care of dependants or pets in an emergency
- leaving special messages for friends and loved ones
- memory boxes or books for children
- wishes for funeral arrangements.

In some areas, a standard form will be used to record an advance care plan, such as ReSPECT.[4]

An advance care plan may be a much-used document. Although advance care plans and advance statements (see below) are not legally binding, they still give a guide to decisions the person may make in the future.

If the person loses capacity to make decisions, healthcare professionals should take the recorded preferences into account as part of an overall judgement of the person's best interests, and the person's preferences should be honoured where possible.

Advance statement

This is a written statement of a person's preferences, wishes, beliefs and values for future management, medical choices and care. This may include where the person would like to be cared for and where they would 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 decisions or communicate them. Advance statements may be included within an advance care plan, but can also stand alone.

Advance Decision to Refuse Treatment (ADRT)

An ADRT allows a person to identify specific treatments to be refused or withdrawn and the specific circumstances in which this would apply in future. This can include the right to refuse life-sustaining treatment. Any person making an ADRT must be over 18.

The decisions written down in an ADRT are legally binding as long as the document is dated and witnessed, and it is applicable to the situation. It should also include a statement that the specific treatment is to be refused 'even if my life is at risk'. It can only be used if the individual lacks capacity to make that particular decision at the time.

If an ADRT is made or updated, copies should be kept with the person's medical records. If local services have a preferred proforma for an ADRT, ensure this is used to avoid confusion.

Before making an ADRT, the person should discuss the contents with an appropriate professional to ensure they are clear about which treatments they wish to decline and that these are recorded correctly.

The person with MND should understand that refusing life-sustaining treatment may mean a natural death will follow, but this may not happen straight away.

Symptoms can be eased with good management, including medication to reduce anxiety, pain or discomfort. The person making an ADRT cannot authorise a doctor to do anything illegal, such as euthanasia, but they can refuse prolongation of life (eg by artificial feeding or antibiotics). In addition, people cannot demand specific interventions within an ADRT, they can only refuse it.

ADRTs are not available in Scotland. People can express their wishes in an advance directive (living will), but this is not legally binding.

Information to share

Our booklet *Advance care planning for MND* includes information and practical strategies to support professionals to effectively facilitate advance care planning for people with MND.

See page 39 for details of how to order our publications.

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. The decision on resuscitation lies with the most senior clinician, and the person with MND cannot demand resuscitation. In England and Wales, refusal of CPR may be included in an ADRT, which is legally binding, with clear instructions about when this should be applied.

Choosing where to die

While for some people with MND death can be sudden, for many, the course of the disease is predictable, and end of life care can be planned. It may be possible, therefore, for the person to be cared for and to die in a place of their choosing.

Encouraging open discussions with those closest to the person with MND helps ensure the person's wishes are clear, potential challenges are recognised, and plans are realistic.

If the person with MND does not die in the place of their choice, it is important to provide reassurance to the family. In some cases, they may feel they have failed, even if the circumstances meant it was not possible to respect the choices of the person with MND.

Health and social care professionals may also experience a similar response and need reassurance that they did not fail the person, and in fact, people may well change their mind over a stated place for care as they come closer to dying. Increasingly, evidence is showing that being well-cared for, feeling safe with good control of symptoms is more important than the place in which the care occurs.[9]

Home – A person with MND may wish to die within the security of familiar surroundings, close to family and usual carers. However, people with advanced MND may need high levels of care, increasing the demands on family carers to provide extra help. This help may involve moving, handling or using complex medical equipment.

Family carers often take on the full responsibility of caring for someone with MND, and their need to be involved must be balanced with their need for respite.

A night sitting service, if available, can be arranged via district nurses, the GP or through fast-track NHS Continuing Healthcare funding. The co-operation and support of the GP and the primary healthcare team, and the inclusion of the person on any available local palliative care registers, is essential. In some areas, Hospice at home services may also be available to provide support. For further information and resources, visit **www.mndassociation.org/chc**

Hospital – Hospital is often not the preferred place to die, but some people may wish to return to a ward where they are known. Timing of admission can be difficult, as many acute hospital beds have restrictions over duration of care. In some areas, there are identified end of life care beds in local community hospitals, which may be available. The hospital palliative care team should be made aware if a person with MND is admitted.

Care or nursing home – Some people may choose to die in a care or nursing home, particularly if it has been their home up until that point. However, some homes will need support to care for someone with MND at end of life.

Hospice – Most hospices provide care and support for people with MND. Early introduction to a hospice and its services, for example home care, day care or physiotherapy, is advised. It is important to be aware that hospices do not usually offer inpatient care for prolonged periods of time, and a bed may not be available when needed.

Many hospices have a community palliative care team who accept referrals for symptom control and other complex palliative care needs. In some areas the community palliative care team may be part of the NHS community or hospital services, but the support they can offer should be similar.

Cognitive change and decision making

Up to half of all people with MND will experience varying degrees of change in thinking, memory and behaviour. This increases to 80% in the final stage of the disease.[7] A small percentage of these will be diagnosed with frontotemporal dementia, which can significantly affect mental capacity.

Where cognitive change is mild, the person may still be able to make reasoned choices on their own behalf.

People with MND and their families may want to talk about potential changes to cognition and decision-making, and should be encouraged to discuss advance care planning before cognitive change affects their capacity.

Mental Capacity Act 2005[10]

The Mental Capacity Act 2005 empowers people to make decisions for themselves wherever possible, and protects people who lack mental capacity to make specific decisions.

Wherever possible, people with MND must be supported to make their own decisions. This may include the use of communication aids.

Information for you

Cognitive change, frontotemporal dementia and MND

Information to share

Changes to thinking and behaviour with MND

See page 39 for details of how to order our publications.

Advance care planning checklist

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

☐ Has the person with MND and their family been reassured that death from choking is exceptional and that death in the majority of cases is peaceful? See page 25.

☐ Are you prepared for questions about suicide and assisted suicide? See pages 7-8.

☐ Does a palliative care clinician or team need to be involved? The terminal and unpredictable prognosis of MND means the principles of palliative care should apply from diagnosis. Early links with palliative care services can provide useful support but should be introduced at a time appropriate to the individual (see pages 20-22).

☐ Is someone helping the person with MND to develop an advance care plan? Advance care planning should include discussions about preferred place of care/death, Advance Decisions to Refuse Treatment (ADRT) and their wishes regarding burial or cremation.

☐ Has an Advance Decision to Refuse Treatment (ADRT) or Do Not Attempt CPR (DNACPR) been recorded?

☐ Have any advance care planning documents been reviewed with the person with MND, and are up-to-date copies filed with all relevant professionals, including the ambulance service and out-of-hours team?

☐ Is there a contingency or emergency care plan in place, so the family carers know who to contact in any likely scenario, both in and out of hours? Lack of such a plan may lead to the person being admitted to hospital at end of life.

☐ Have patient's current medications been rationalised and reviewed to ensure they are still appropriate?

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

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

☐ Has the local primary care out-of-hours service been informed of any DNACPR or ADRT?

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

☐ Is the person registered on a local palliative care database?

Supporting family and carers

☐ Is the main family carer on the carers' register at the local GP practice? This will allow the carer's own needs, and any impact of the caring role on their own health, to be recognised.

☐ Does the person's family have the support and information they need? See pages 31-33.

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

Putting affairs in order

Power of Attorney[11,12]

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 if they are unable to communicate their wishes or lack the capacity to make decisions themselves.

LPAs can cover decisions regarding financial and/or specified health/care related matters. An EPA allows one or more people to manage financial affairs and property but doesn't cover decisions on health or care. In Scotland, there is a Welfare Power of Attorney and also a Continuing Power of Attorney, which is for finance.

The person with MND must have capacity to understand and make the required decisions at the time they complete their Power of Attorney. The Power of Attorney will not come into force until the person no longer has capacity to make a decision.

Wills, trust funds and guardianship

A will allows instructions to be left about what will happen to money, property and possessions when someone dies. This is essential for ensuring wishes are carried out as expected, especially if there are problems within a family or where partners are not married.

Legal advice should be sought when making a will to ensure its validity. If there are concerns regarding guardianship of children, these will need to be clearly expressed. Some people may wish to set up trust funds to ensure the financial future of their family.

The person should also be encouraged to gather important paperwork, such as information about bank accounts, and to keep these together with the will.

Organ and tissue donation[13]

Donation for transplant is not usually possible if a person dies from MND. The organs of people with MND can be accepted for life-saving transplants if they die in hospital following an accident or from a cause unrelated to MND.

Some people will want to donate brain and spinal cord tissue for MND research. It is not usually possible for someone to donate organs for both transplant and medical research.

Any arrangements for organ or tissue donation should be made well in advance. The person should discuss with their family if they would like their organs or tissue to be donated once they have died. This is important, because even though the person's request to donate may be registered, the family will still be consulted at the time of death.

Although they do not have the legal right to veto or overrule the person's decision to donate, there may be cases where it would be inappropriate to go ahead if it would cause distress to the family.

The decision should ideally be recorded in an advance care plan, and communicated with key members of the healthcare team, funeral directors and, where relevant, the tissue bank.

If someone has registered to donate their tissue for research, their details will be held by a specific tissue bank. Because donation must take place within strict timeframes after death, the process needs to be carefully organised and clearly communicated.

See section 12 of the MND Association's *End of life guide*.

Information to share

Research sheet I - Tissue donation

See page 39 for details of how to order our publications.

Funeral and memorial planning

By discussing wishes with their family, the person with MND can ensure a funeral or memorial will be as they would have wanted.

It saves the people left behind from worrying whether they've made the right choices. Some people choose to organise and pay for their funeral in advance. If religious rites or other rituals need to be observed, people may need to ensure arrangements are made before they die. See section 8 of the MND Association's End of life guide.

Involving palliative care services

Palliative care is the active holistic care of people with life-limiting and life-threatening illness. It involves:

- management of pain and other symptoms
- psychological support
- social support
- spiritual support.

The aim of palliative care is to achieve best quality of life for people with MND, their families and those close or important to them.[14]

It is important to distinguish between palliative and end of life care. Many people believe that palliative care is just offered near the end of life, but the terminal and unpredictable prognosis of MND means it is vital that people with MND are offered access to specialist palliative care as early as possible, so that symptoms can be managed effectively. Many aspects of this type of care are applicable earlier in the course of the illness in conjunction with other treatments.

While early links with specialist palliative care services can provide a useful source of advice and support, introducing this concept may present some difficulties.

This is often because people are unaware of what palliative care is and what it can provide. It is important to explain the role of the palliative care team and the potential benefits they can offer to the person's quality of life. It might help to introduce the service via outpatient appointments, day care services, and access to support such as physiotherapy or occupational therapy.

Information to share

Information sheet 3D - *Hospice and palliative care*

See page 39 for details of how to order our publications.

Palliative care settings

Palliative care support may be available in different settings, including at home, in hospital, in MND care centres and networks or in hospices. They may offer day care and inpatient admission for symptom control or end of life care. Some may also offer respite services.

If palliative care includes referral to a hospice, it is important to stress this does not always mean that death is imminent, as many people view hospices purely as places where people die.

What the specialist palliative care team offers

The different disciplines of the specialist palliative care team can:

- offer psychological, spiritual, emotional and bereavement support to the whole family
- refer to other sources of support, such as community faith leaders
- advise on control of symptoms, including pain
- support complex and difficult decision making, including advance care planning and decisions about life-prolonging interventions
- enable access to rapid response services, helping people to be treated in their preferred place of care/death, and to prevent inappropriate hospital admissions
- arrange access, where available, to complementary therapies for the person with MND and their carer
- liaise closely with the local hospice
- liaise closely with and offer advice, support and educational opportunities to health and social care professionals eg advice on introducing the topic of palliative care in conversation.

Emotional and spiritual support

It is important that people's emotional and spiritual needs and/or beliefs are recognised and that they have the opportunity to discuss them if they wish. Support may come from partners and family, friends as well as health and social care professionals.

A person's basic spiritual care needs can be met in many ways, for example someone's spirits may be lifted by a visit from a neighbour or a letter from a friend. Pets can play an important role here too.

Comfort may come not only from spending time with them, but also from the reassurance that their pet is being well cared for.

Be aware of the role you and your colleagues can play. Although spiritual support may be provided by a chaplain, counsellor, psychologist or therapist, the person's need is not always religious and may not need a referral to formal services.

Information to share

Emotional and psychological support

See page 39 for details of how to order our publications.

Caring for a person with MND at the end of life

Because everyone with MND has a different experience of the condition, it can be very difficult to predict when someone with MND will reach the terminal phase.

For some people, death can be very sudden, before an obvious end stage is reached. Others experience a protracted final stage, which can last many weeks. But the most usual clinical picture is of rapid deterioration in respiratory function, often following an upper respiratory tract infection.[1]

Death in the majority of cases is very peaceful, following lengthening periods of sleepiness, gradually resulting in a coma.[15]

Recognition of the dying phase in neurological disease can be difficult. Its onset can be signalled by symptoms that might include:

- breathlessness, caused by reduced chest expansion and use of accessory muscles (if any are still in use), a quietening of breath sounds, and morning headache from CO_2 retention overnight
- increased NIV dependency (if used)
- reports that the NIV "isn't working anymore"
- aspiration pneumonia
- systemic sepsis
- reduced level of consciousness without reversible cause
- pressure sores.

These signs might be noticed by the carer or a member of the multidisciplinary team. Every effort should be made to recognise this final deterioration and to discuss the situation with the person with MND (if possible) and their carers. This will ensure everyone has had the opportunity to understand and prepare for what is happening, so that plans are updated and the right support is put in place.

Withdrawal of ventilation[3,16]

People using ventilation should be reassured that they can request withdrawal at any time and seek help and advice, particularly if fully dependent. Some may record their wishes in an ADRT, for example if they reach a certain level of disability. Others may later feel ventilation is no longer beneficial or becomes burdensome, or may suddenly find the mask claustrophobic and decline its use despite previous acceptance.

A discussion with the person with MND and their carer/family may be needed about whether to continue ventilation and the implications of stopping it. This planned procedure should follow palliative care and organisational guidelines and is usually led by a clinician, such as a palliative care specialist.

If someone reliant on ventilation is approaching end of life, death is likely to happen in a relatively short time following withdrawal of support. Medication can be given to relieve anxiety and distress.

If a person with MND wishes to stop assisted ventilation, they, and those close to them, should have support from professionals with knowledge and expertise in stopping ventilation, medication to control symptoms, legal and ethical frameworks and responsibilities, including up-to-date knowledge of the law regarding the Mental Capacity Act, DNACPR, ADRT orders, and Lasting Power of Attorney.

Support should be available for professionals involved if there is a plan to stop the person's ventilation, including the legal and ethical implications.[3] Guidelines from the Association for Palliative Medicine support professionals through the process of withdrawing ventilation.

Information for you

Information sheet 8C - *Withdrawal of ventilation with MND*

Information to share

information sheet P3 - *Managing respiratory symptoms in MND*

See page 39 for details of how to order our publications.

Choking and breathlessness

Many people with MND fear they will die from choking, but death from choking is extremely rare. Some people may experience choking episodes due to swallowing problems (dysphagia) caused by weak bulbar muscles.[17] Others experience sensations of choking due to spasms in the larynx (laryngospasm).

Careful management with medication can reduce sensations of choking and actual choking episodes.

Consider managing breathlessness with an opioid, a benzodiazepine or a combination of the two. Antimuscarinics, such as glycopyrronium or hyoscine hydrobromide, may be used to reduce saliva and respiratory secretions.[8,18]

The experience of breathlessness can lead to anxiety, which can increase the panic of breathlessness. This panic can spiral out of control when people fear they may die fighting for breath or suffocating. Talking about fears and concerns with the person and their family can help minimise anxiety.

Healthcare professionals have a role in teaching people with MND and their carers/family members how to manage episodes of choking, sensations of choking and breathlessness. This may include:

- correct positioning
- a handheld fan
- use of appropriate medication
- cognitive strategies, including how to stay calm and in control.[9]

Strategies to help people with MND and their carers deal with choking can be found in our care information sheet 7A: *Swallowing difficulties*. See page 39 for details of how to order our publications.

Anticipatory prescribing[8]

People with MND who are anxious about choking, sensations of choking, breathlessness or inability to clear saliva or secretions may be reassured by having medications to help nearby. These are sometimes known as 'pre-emptives'.

Anticipatory prescribing is essential for ensuring patients can receive prompt symptom relief at home in their final days. By prescribing medications in advance, distressing symptoms such as pain, nausea, and agitation can be managed without delays, particularly outside normal working hours. This approach helps prevent unnecessary suffering and supports patients who wish to die at home with dignity.

A key element is the 'just in case' box, which contains essential medications tailored to the patient's needs. These drugs remain the patient's property and can be administered by attending clinicians when required. While anticipatory prescribing improves care, it must complement, not replace, clinical assessment at the time of use. Clear communication between healthcare teams, including out-of-hours providers, ensures effective coordination and supports high-quality end of life care.

A 'just in case' box is usually prescribed by a GP, palliative care team, or nurse prescriber involved in a patient's end of life care. It is typically arranged following discussions with the patient and their family about symptom management and care preferences.

Once prescribed, the medications and necessary administration equipment are dispensed by a community pharmacy. It's important to ensure that the patient's healthcare providers, including out-of-hours services, are aware that a just in case box is in place, so they can use it if needed. If you are caring for someone who may require one, speak to their GP, district nurse, or palliative care team.

Medications at end of life

If someone is dying, early intervention can prevent symptoms from becoming distressing. The person will become drowsier as part of the dying process. Occasionally, the doses of medication required for symptom control may contribute to drowsiness.

Only medication to control or prevent symptoms is appropriate at this time. Thought should be given to stopping medication not specifically aimed at this purpose.[8]

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

- antimuscarinics, such as hyoscine hydrobromide and glycopyrronium, to reduce saliva and respiratory secretions[3,8]
- medications to manage breathlessness exacerbated by anxiety, and others to reduce anxiety/terminal restlessness, such as midazolam, haloperidol or levomepromazine[3,19,20]
- opioid analgesics, such as morphine sulphate or diamorphine, to reduce cough reflex, relieve dyspnoea (breathlessness), fear and anxiety. They can also control pain[3,8]
- antiemetics, such as levomepromazine or cyclizine, for nausea.[18]

Oxygen

Oxygen therapy must be used with great caution for people with MND. It corrects oxygen saturations, but a rising level of carbon dioxide can lead to symptoms and ultimately death in people with MND.[21]

Oxygen may occasionally be used at end of life in combination with opioids and benzodiazepines to reduce the distress of breathlessness. However, always involve the specialist palliative care team to ensure appropriate symptom control and support for the person with MND.

Pain

As they reach the terminal stage of MND, many people complain of generalised pain and severe discomfort from musculoskeletal causes, such as muscle spasm, or from skin pressure due to immobility.[22] This can often be treated with analgesics.

Carefully titrated opioid analgesics may be necessary, especially for pressure pain, and should not be withheld if needed.[8,22] Regular analgesics should usually be continued until death, even if oral medication is no longer possible due to dysphagia. Alternatives, such as topical, transdermal or parenteral routes should be considered. Parenteral medication may be more conveniently given as a continuous subcutaneous infusion using a syringe pump.[8] Gut function may reduce over the dying phase, so ongoing review of medications is recommended.

Before putting any medication through a gastrostomy tube, check that it will not occlude the tube or affect the drug's action. Information can be obtained from a pharmacist and also available in Palliative Care Formulary (in Chapter 28 - *Drug administration to patients with swallowing difficulties or enteral feeding tubes)*.

It is also important to check whether carers (family or paid workers) are able to administer other forms of medication.

Physiotherapy, including passive exercise, can ease the pain from immobile joints.[23] Some people with MND may find massage helpful for pain and spasticity.

All healthcare professionals have a role in prevention of pressure sores.

Information to share

Information sheet 6C - *Managing pain*

Information for you

Information sheet P11 - *Pain in MND*

See page 39 for details of how to order our publications.

Terminal restlessness

This is the agitation sometimes seen in people just before death, which is usually associated with a reduced level of consciousness. A person may appear unconscious, restless and unsettled. They may seem disorientated, anxious, fidgety and may look scared or distressed. It can happen intermittently or be a persistent feature.

This situation can be distressing for families and carers as they can feel a lack of control over the situation. It is important to keep them informed at all times. The following may help:

- Ensure there is no physical cause for the agitation, such as pain or discomfort from poor positioning, a full bladder or rectum.
- Provide calm reassurance and spend time with the person and their family in a soothing environment.
- If no reversible cause is found, agitation may be managed with antipsychotics and/or benzodiazepines (e.g. midazolam), which usually relieve distress but may increase drowsiness.[19]
- In all cases where repeated doses of medication are needed, the use of a syringe pump may be a preferred method of administration.[8]
- Consider screening the person for delirium which may be caused by infections, dehydration etc.

Nutrition and hydration[18]

Most people stop eating and drinking in the final days of life due to reduced desire for food and drink and reduced consciousness. It is important to support the dying person to drink if they wish to and are able to, but monitor for swallowing problems and minimise risk of aspiration.

Hydration may be given by feeding tube. If a feeding tube is not available and there are concerns the person is thirsty, subcutaneous fluids can be considered as a trial and then reviewed, although this may be at a reduced level. If subcutaneous or intravenous fluids are necessary, an inpatient admission might be required, though this is rarely needed. Ensure thorough and regular mouth care is given (see next heading) as this can impact on the perception of thirst.

A person with MND may choose to reduce or withdraw their previous established feeding regime. It is important to explain the situation carefully and sensitively to relatives or carers, who might fear the person with MND may die from starvation or dehydration.

Mouth care

In the final days of life (for many conditions, not only MND), mouth breathing and minimal fluid intake can cause the mouth to become dry and make lips more likely to crack.

The mouth should be kept clean and moist, for example using a soft baby toothbrush. Relatives can be shown and encouraged to provide this aspect of care. It may also be acceptable to use one of the person's favourite drinks.

Looking after yourself

MND creates many challenges for professionals and can arouse strong emotions. These can include frustration, powerlessness, inadequacy and sadness. It highlights attitudes to issues related to disability, quality of life and measures taken to prolong life.

Compassion fatigue is a state of physical and emotional exhaustion that results from prolonged exposure to the suffering or trauma of others. It can reduce a professional's capacity for empathy and diminish their sense of purpose, and is a recognised risk for those caring for people with MND.[24]

Caring for someone over time can make their death distressing, which is a normal response. Looking after yourself and building resilience is important. Good multidisciplinary teamwork, regular meetings, and supervision provide support and a space to discuss challenges.

Key points on care at end of life

- Every effort should be made to recognise the end stage of MND early, discuss with the person and family, and put support in place.
- Review medication to ensure good symptom control.
- Use anticipatory prescribing (analgesics, anxiolytics, antiemetics, anticholinergics – see p.26).
- Manage pain effectively, often with opioids.
- Continue hydration where appropriate (see p.29)
- Keep the mouth clean and moist.
- Provide physical and emotional support for family and carers.

Communication

- Always try to maintain communication, even in the final stages. Use closed questions that can be answered with a word, eye movement, blink, or hand squeeze.
- Picture/alphabet boards or other aids may also help.
- Hearing and touch are thought to be the last senses retained, so gentle conversation and touch can offer reassurance and compassion.

Supporting family and carers

MND is unpredictable and may progress rapidly, with death occurring more quickly than anticipated. Carers and family may need practical and emotional support.

Advice should be given to carers and family members about appropriate care and management in certain situations. This can prevent them inappropriately calling the emergency services.

Concerns about finance and paying for care can cause anxiety to carers. At some point, the person with MND may meet the criteria for NHS Continuing Healthcare funding. People who may be entering a terminal phase may require 'fast tracking'. For further information and resources, visit **www.mndassociation.org/chc**

Professionals should provide support for carers as MND becomes more complex. Early end of life discussions can help families reflect, prioritise what matters, and address unfinished business.

Care plans and information must be shared by all members of the care team and adequate care maintained.

Early and sustained symptom control is essential in helping people live as well as possible and to have a peaceful and dignified death.

If they wish to, the person with MND should remain in control of end of life issues as much as possible.

Carers' Alert Thermometer

The Carers' Alert Thermometer is a tool to help professionals identify and support the needs of unpaid carers providing end of life care at home. Access the tools at **www.edgehill.ac.uk/carers**

Information to share with carers

Caring and MND: support for you - Comprehensive information designed to support individual needs during the caring role.

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

See page 39 for details of how to order our publications.

Preparing the family for end of life

As the end of life approaches, families often need to talk about what is happening. The person dying should feel able to share their concerns, whether or not with family. Some families may also benefit from specialist support, such as a social worker or hospice team.

Some families may need encouragement to talk things through sooner rather than later, especially if the person with MND appears to be deteriorating rapidly.

It will be important to reassure relatives that sometimes symptoms at the very end of life, such as noisy breathing, are more distressing to the family than the person with MND.

Support for children

Children also need time to prepare themselves and may have things they want to do or fears they need to express. Equally important, they may need time to themselves. Be ready to identify the need for specialist support for children, which can include palliative care social workers or family support workers.

As part of planning for the future, someone with MND may think about leaving special messages for friends and loved ones, and memory boxes or books for children. Video or sound recordings can also help, but this may need to be encouraged in the early stages of MND, in case speech and communication deteriorate.

Information for children and young people

When someone close has MND, an interactive workbook, is designed to promote coping strategies for children aged four to ten.

So what is MND anyway? is a booklet designed to introduce MND to young people aged 13 to 18, including young carers.

Information to share

Our booklet *Finding your way with bereavement* offers guidance on emotional and practical support, including how to help grieving children and young people.

See page 39 for details of how to order our publications.

Cultural diversity

Family reactions to death and dying can be diverse and sensitivity is needed towards cultural variation. Careful exploration is advised.

Worries about the future

Many families will have fears and concerns about coping in the future. These concerns may be linked to their financial situation. There are many sources of support to which you can signpost them. See our care information sheet 10A – *Benefits and entitlements.* See page 39 for details of how to order publications. Visit w**ww.mndassociation.org/benefits-advice** for information about our specialist benefits service.

Concerns about inherited MND

Inherited MND (sometimes known as familial MND) accounts for approximately 5-10% of all people with MND. The neurologist may refer those affected for genetic counselling.

Information to share

Our booklet *Inherited MND and genetic counselling* can help people with MND, or those with a family history of the condition, find guidance about genetic counselling and genetic testing.

See page 39 for details of how to order our publications.

Useful resources

Gold Standards Framework

A practical systematic, evidence-based approach to optimizing care for all people nearing the end of life, given by generalist front-line care providers. It includes a range of useful resources and tools.
www.goldstandardsframework.org.uk/

Health Education England - E-learning programme for End of Life Care (e-ELCA)

This e-learning programme aims to enhance the training of the health and social care workforce so that well-informed high-quality care can be delivered. It includes over 170 highly interactive sessions which are grouped into nine modules.
www.e-lfh.org.uk/programmes/end-of-life-care/

Royal College of Nursing - End of Life Resources

Includes a range of useful guidance and resources, as well as extensive links to other useful resources.
www.rcn.org.uk/clinical-topics/end-of-life-care/professional-resources

Royal College of Physicians - Talking about dying report

This report offers advice and support to any doctor holding conversations with patients after the diagnosis of a progressive or terminal condition. Search for "Talking about dying" at **www.rcplondon.ac.uk**

Talk CPR

This website includes useful information and resources to support conversation about CPR with people with life-shortening conditions. Visit **http://talkcpr.wales** to find out more.

Royal College of GPs - Palliative and End of Life Care Toolkit

a collection of tools, knowledge, and current guidance for healthcare professionals to support patients nearing the end of life to live well until they die. Visit **www.rcgp.org.uk** and search for end of life toolkit.

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How we can support you

MND affects people from all backgrounds, with varied and complex needs. Health and social care professionals should tailor support, share relevant information, and work closely with specialist services.

The MND Association provides resources and services to help you do this. Find out more through:

MND Connect

Our helpline offers help to people living with MND, carers, family and health and social care professionals. Our advisers can provide emotional and practical support, including social and financial information, and refer to local support and MND Association's services.

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

MND Association website

Our website offers supporting information on MND and our work, including the research we fund, the services we provide and how to get involved.

www.mndassociation.org/professionals - discover all resources for health and social care professionals

www.mndassociation.org/support-and-information - discover all help available for people with or affected by MND

Support for health and social care professionals

Professional information resources

We have a wide range of information resources available to support professionals from different disciplines, including booklets, infographics and online courses. Our professional information finder can help navigate them according to profession and topic.

www.mndassociation.org/pro-info-finder

MND Professionals' Community of Practice

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

www.mndassociation.org/cop

Education

Our education programme is designed to improve standards of care and quality of life for people living with and affected by MND. We regularly organise webinars, workshops, international lectures and networking events.

www.mndassociation.org/education

Support for people with or affected by MND

Care information resources

We produce high quality information resources for people living with MND, carers and family members. Our publications, available in different formats and languages, can help people understand MND, plan ahead, and talk about MND to other people, including children. Professionals can obtain this information to supply people they support, or to open conversations.

www.mndassociation.org/careinfofinder

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

Financial support grants and equipment loan

Where statutory provision is not available, we may be able to offer financial support.

www.mndassociation.org/our-services

Online forum

Our online forum is accessible to anyone and can offer first-hand experience of MND, and practical and emotional support.

forum.mndassociation.org/

Local support

The MND Association offers a range of options for support in your local area:

- Specialist MND care centres and networks across England, Wales and Northern Ireland
- Nearly 150 online and face-to-face support groups
- Trained volunteers and volunteer-led groups offering practical help by phone, email, or home visits

www.mndassociation.org/local-support

How to order publications

Our publications are free of charge to people living with or affected by MND, or Kennedy's disease. Health and social care professionals can also order items for themselves or on behalf of someone with or affected by MND or Kennedy's disease.

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

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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