

mnda

motor neurone disease
association



**A professional's
guide to end
of life care in
motor neurone
disease (MND)**



For health and
social care
professionals



Motor neurone disease (MND) is a progressive and terminal disease that results in degeneration of the motor neurones, or nerves, in the brain and spinal cord.¹

There is no cure for MND. Although the condition itself is unpredictable, the terminal prognosis is not. 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 health and social care professionals who are working with people with MND. The aim of this booklet is to encourage professionals to discuss death, the process of dying and advance care planning, as soon as someone with MND is able to discuss the subject. By understanding their needs, professionals can support people affected by MND to prepare for end of life.

Need to find out more?

The MND Association supports professionals to care for people affected by MND in a variety of ways, which includes providing:

- a range of information and educational opportunities
- local support and advice from our staff and volunteers.

MND Connect

Accredited by the Helplines Standard, our helpline offers information and support by telephone and email – on **0808 802 6262** or **mndconnect@mndassociation.org**

Professionals' forum

Visit **<http://proforum.mndassociation.org>** to share best practice with other health and social care professionals who are caring for people living with MND.

See page 30 for more information about how the MND Association can support you in your role.

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With MND you have to develop courage to talk about things and death is one of them. One professional came and talked to me and my wife about death like she was talking about the price of potatoes. I really appreciated that.

“I’m sure that may not be everybody’s cup of tea, but I wanted someone that I could just talk about death to, without crying, without getting upset.”

A person with MND



Talking about end of life

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

People with MND need to be able to exercise choice and control and should be encouraged to talk through options for their care and their preferences for end of life.³ Conversations may be more difficult if the person's speech becomes affected, or if they experience cognitive change.

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.⁴

Topics to talk about

The overriding topics that people with MND often want to talk about are their fears and concerns, and not necessarily the management of the condition. However, 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, and it is important that concerns are taken seriously and solutions, where possible, are made available. It is also important that the language used, while remaining sensitive, is clear and easy to understand. This means not being afraid to use the words 'death' and 'dying' instead of euphemisms. This can be difficult for some professionals, but if the person with MND is 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 page 9), and communicated to relevant health and social care professionals. Some areas have standard forms that are used for this purpose.

Conversations discussing wishes, preferences and plans for future care should also be registered and shared on local palliative care registers/lists/co-ordination systems, where these exist. Examples include the Gold Standards Framework and Electronic Palliative Care Co-ordination Systems (EPaCCS).

Reviewing choices

People can and will 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.

Key actions

- The person with MND should have the opportunity to discuss end of life with their GP, specialist palliative care team or any other professional with whom they have built a trusting relationship.
- The specialist palliative care team (see page 17) can advise on the timing of end of life discussions.
- The patient'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.

In a 2012 MND Association study into the views of people with MND on death, dying and end of life decision making, some people with MND responded that they would want more information about how to exert choice over the timing of their death, and clarity over the legality of these options.³ It may be that you are 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. If suggestions or solutions to these concerns can be provided, thoughts of suicide may subside. Discussion is therefore crucial in order to explore and understand these issues and concerns.

This information explains what is and isn't allowed within the law (at time of publication).⁵ 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
- refuse life-sustaining treatments, which they feel are no longer 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.

A person with MND can influence how their care will be managed in the later stages of the condition using advance care planning (see page 9). 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.

Medication cannot be used to hasten death, but it may be provided as part of a package of palliative care to reduce pain and other symptoms at end of life.⁶ It is important that these symptoms are treated in order to reduce distress at this stage.

Clinicians should feel confident in addressing these matters and seek support if needed from their local specialist palliative care team.

Resources to support end of life discussions

The MND Association's End of life guide

We produce a comprehensive booklet for people with MND and their families about end of life care and decisions. It may support professionals to open any difficult conversations. The guide covers subjects including:

- how MND progresses
- 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.

Call our MND Connect helpline on **0808 802 6262** for further support and to order a copy of the guide. Alternatively, you can download it from **www.mndassociation.org/publications**, where you can access separate sections, allowing you to focus on specific topics.

Difficult conversations booklet

The National Council for Palliative Care (NCPC) has produced a booklet with the support of the MND Association, called *Difficult Conversations: making it easier to talk about the end of life with people affected by motor neurone disease*. It has plenty of tips about how to start conversations. See page 32 for NCPC contact details.

Advance care planning

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

Advance care plan

An advance care plan can be wide-ranging, covering all aspects of day-to-day care including, 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.

In some areas, a standard form will be used to record an advance care plan. 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 a patient 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 withdrawn or refused 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.

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 all named on the ADRT. 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. However, any symptoms can be eased with medication that aims 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 intervention within an ADRT, they can only refuse it.

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, where needed, to avoid potential confusion.

Do Not Attempt CPR (DNACPR)

Someone with MND may choose to have a DNACPR. This will be respected in most instances, but is not legally binding. In England and Wales, refusal of CPR may be included on an ADRT, which is legally binding, with clear instructions about when this should be applied.

Information you can share

See Information Sheet 14A – *Advance Decision to Refuse Treatment (ADRT)* and section 9 of the MND Association's *End of life guide*.

See page 30 for details of how to order copies or download this sheet from **www.mndassociation.org/publications**

Further information is also available at **www.adrt.nhs.uk**

Choosing where to die

While for some people with MND death can be sudden, for many, the course of the disease is predictable and palliative care can be planned.⁷ It may be possible, therefore, for the person to be cared for and to die in a place of their choosing.

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 and/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 or the GP. 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 register, is essential.

Hospital

Hospital is often not the preferred place to die, but some people with MND may wish to return to a ward where they are known. Timing of admission to hospital 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.

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 local hospice and its services, for example home care, day care or physiotherapy, is advised. It's important to be aware, however, that hospices do not usually offer inpatient care for prolonged periods of time, and a bed may not be available at the time it is needed.

Many hospices have a community palliative care team who may accept referrals for symptom control. The local palliative care team may alternatively be based elsewhere, for example in the community or a local hospital.

If, for some reason, 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.

Cognitive change and decision making

Up to half of all people with MND will experience varying degrees of change in thinking, memory and behaviour. A small percentage of these will be diagnosed with frontotemporal dementia, which can significantly affect capacity.^{8,9} Where cognitive change is mild, the person may still have capacity to make reasoned choices on their own behalf.

People with MND and their families may want to talk about this and should be encouraged to discuss advance care planning in good time.

Mental Capacity Act 2005

The Mental Capacity Act 2005 empowers people to make decisions for themselves wherever possible, and protects people who lack capacity to make specific decisions.¹⁰ Wherever possible, people with MND must be supported to make their own decisions.

Information for you

- *Cognitive change, frontotemporal dementia and MND* booklet

Information you can share

Our information sheets for people affected by MND include:

9A – *Will the way I think be affected?*

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

9C – *Managing emotions.*

Call MND Connect on **0808 802 6262** to order copies or download from our website at **www.mndassociation.org/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?
- Are you prepared for questions about suicide and assisted suicide? See page 7 for more information.
- 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 page 17).
- 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 and advance decisions to refuse treatment (ADRT).
- 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.

- Are appropriate medications available in the home, to prevent a crisis admission (end-of-life symptom management following local palliative care guidelines)? See below for information about the *MND Just in Case kit*.
- Is the ambulance service aware of any signed DNACPR form, or an ADRT?
- Have you informed your local primary care out-of-hours service of any DNACPR or ADRT?
- Does the district or community nurse know there is someone with MND in their area?
- 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 they need?
- Is there adequate physical and emotional support for the family if the person wishes to die at home?

The MND Just In Case kit

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

For the GP and district or community nurse it provides guidance on symptom management and storage for the prescribed medications. Once the need for a kit has been discussed and agreed with the person with MND and their carer, the GP orders a kit for free from MND Connect (see page 30) and prescribes medication to be supplied within it.

Putting affairs in order

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 if they are unable to communicate their wishes or lack capacity to make decisions.^{11,12}

An LPA 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. 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 patient no longer has capacity to make their own decisions.

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

Donation for transplant is not usually possible after a person dies from MND.

However, the NHS Blood and Transplant Authority agrees 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 essential, because even though the person's request to donate organs or tissue may be registered, the family will need to give their permission at the time of death.

The decision should ideally be recorded on an advance care plan, and communicated with key members of the healthcare team, funeral directors and, where relevant, the tissue bank. If the person has registered to donate their tissue for research, their details will be registered with an individual tissue bank.

Information you can share

See section 12 of the MND Association's *End of life guide* (see page 8 for details).

Our other resources for people affected by MND include:

- Research Information Sheet I – *Tissue donation for MND research*

Call MND Connect on **0808 802 6262** to order copies or download from our website at **www.mndassociation.org/researchsheets**

Funeral and memorial planning

By discussing their 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 special arrangements are made before they die.

Involving palliative care services

Palliative care is the active holistic care of people with advanced, progressive 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 and their families.¹³

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.^{14, 15}

While early links with palliative care and specialist services can provide a useful source of advice and support, introducing the concept of palliative care and specialist services 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 day care, respite and complementary therapies.

Palliative care settings

Palliative care support may be available in different settings, including at home, in hospital or in hospices, which may offer day care, respite care and inpatient admission for symptom control or end of life care.

If palliative care includes referral to a hospice, it's important to stress this doesn't 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 specialist palliative care team may include a number of professional disciplines. They will:

- offer support to the whole family
- offer psychological, spiritual, emotional and bereavement support
- refer to other sources of spiritual support, for example community faith leaders and representatives of other beliefs
- advise on control of symptoms, including pain
- 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
- provide educational opportunities
- liaise closely with and offer advice, support and educational opportunities to health and social care professionals who are supporting people with MND. For example, they may give 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. Emotional support may come from partners, family, friends and health and social care professionals.

Spiritual care is not always religious and may be provided by a chaplain or dedicated spiritual care provider or community leader, counsellor, psychologist or therapist.

Caring for a person with MND at 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.¹⁶

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.^{1,7} Death in the majority of cases is very peaceful, following lengthening periods of sleepiness, gradually resulting in a coma.¹⁷

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 CO₂ retention overnight
- systemic sepsis
- reduced level of consciousness without reversible cause
- pressure sores.⁷

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

People using ventilation should be reassured that they can ask for it to be stopped at any time. They can ask for help and advice if they need it, especially if they are fully dependent on ventilation.⁴ Someone may have used an ADRT to specify withdrawal in certain circumstances

(eg when reaching a specific point of disability). Some people may reach a time when they feel their breathing support is no longer helping or has become a burden. They may suddenly feel claustrophobic from wearing the mask and decline ventilation when previously it has been accepted. In such situations, a discussion may take place with the person with MND and their main carer/family about whether it is appropriate to continue using ventilation.⁴

If someone has been reliant on ventilation and 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 should have support from professionals with knowledge and expertise in stopping ventilation, palliative medicines, 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.⁴

Support should be available for healthcare professionals who may be involved if there is a plan to stop non-invasive ventilation, including the legal and ethical implications.⁴ Guidelines from the Association for Palliative Medicine support professionals through the process of withdrawing assisted ventilation.¹⁸

Choking and breathlessness

Many people with MND fear they will die from choking, but death from choking is rare and many people do not experience choking at all.¹ Some people may experience choking due to swallowing problems (dysphagia) caused by weak bulbar muscles.¹⁹

Careful management with medication can reduce choking episodes. Opioids, such as morphine sulphate or diamorphine, can reduce pain and distress.²⁰ Antimuscarinics, such as hyoscine hydrobromide or glycopyrronium, may be used to reduce saliva and respiratory secretions.⁶

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.¹⁶ Talking about fears and concerns with the person and their family is an important aspect of minimising anxiety.

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

- correct positioning
- use of appropriate medication
- cognitive strategies, including how to stay calm and in control.^{21,22}

Strategies to help people with MND and their carers to deal with choking can be found in our care information sheet 7A: *Swallowing difficulties*. See page 30 for how to order resources.

People with MND who are anxious about choking, breathlessness or inability to clear saliva or secretions may be reassured by having medications to help nearby, in an MND Just in Case kit (page 14).²³

Medications at end of life

If someone is dying, early intervention can prevent symptoms from becoming distressing. As people are dying, they will become more drowsy 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.⁶

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

- antimuscarinics, such as hyoscine hydrobromide and glycopyrrolate, to reduce saliva and respiratory secretions^{4,6}
- medications to manage breathlessness exacerbated by anxiety, and other medications to reduce anxiety/terminal restlessness, such as midazolam, haloperidol or levomepromazine.^{4,24,25}
- opioid analgesics, such as morphine sulphate or diamorphine, to reduce cough reflex, relieve dyspnoea (breathlessness), fear and anxiety. They can also control pain^{4,26}
- antiemetics, such as levomepromazine or cyclizine, for nausea.⁶

Oxygen

Supplementary oxygen therapy is generally not very helpful for people with MND. It corrects oxygen saturations, but it is a rising level of carbon dioxide that can lead to symptoms and ultimately death in people with MND.²¹ However, oxygen may be used at end of life in combination with opiates and benzodiazepines to reduce the distress of breathlessness.²⁷ The involvement of a specialist palliative care team can ensure symptom control and support for the person with MND.

Pain

People with MND rarely die a painful death, although some people with MND do have pain from musculoskeletal causes, such as muscle spasm, or from skin pressure due to immobility.²⁶

As they reach the terminal stage of MND, many people complain of generalised pain and severe discomfort. This can often be treated with painkillers. Carefully titrated opioid analgesics may be necessary, especially for pressure pain, and should not be withheld if needed.^{26, 28}

Regular analgesics should usually be continued until death, even if oral medication is no longer possible due to dysphagia. Alternatives, such as suppositories or parenteral routes should be considered. Parenteral medication may be more conveniently given as a continuous subcutaneous infusion using a syringe pump.²⁹ Before putting any medication through a gastrostomy tube, check with a pharmacist that it will not harden and clog the tube or affect the drug's action.³⁰ 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.³¹ Some people with MND may find massage helpful for pain and spasticity.³²

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

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.³³

This situation can be distressing for families and carers as they can feel a lack of control over the situation. It's 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 packed rectum.³³
- Provide calm reassurance and spend time with agitated people and their family in a soothing environment.
- If no reversible cause can be found, medication may be required to manage the agitation. The use of antipsychotic medication and benzodiazepines (such as midazolam), either separately or in combination, can usually control the distress. But these medications may result in the person being more sleepy.^{25, 33}
- In all cases where repeated doses of medication are needed, the use of a syringe pump may be a preferred method of administration.³⁴

Nutrition and hydration

While most people stop eating and drinking in the final days of life due to reduced appetite and consciousness, distress should be lessened where possible by continuing to offer hydration. Support the dying person to drink if they wish to and are able to, but check for any swallowing problems and the risk of aspiration.³⁵

Hydration may be given by feeding tube. If a feeding tube is not available and there are concerns a patient is thirsty, subcutaneous fluids can be considered as a trial and then reviewed, although this may be at a reduced level.³⁵ It is important to explain the situation carefully to relatives or carers, who might fear the person with MND may die from starvation or dehydration.³⁵

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.²⁵ The mouth should be kept clean and moist using foam stick applicators; some people find crushed ice refreshing to suck. Relatives can be shown and encouraged to provide this aspect of care. It is also acceptable to use one of the person's favourite drinks, whatever it is.

Key points on care at end of life

- Every effort should be made to recognise the end stage of MND, so that this can be discussed with the person with MND and their carer/family members, if relevant, and the appropriate support can be put in place.
- Ensure medication is reviewed to enable adequate symptom control.
- Anticipatory prescribing should consider analgesics, anxiolytics, antiemetics and anticholinergics (see page 21).
- Pain management is crucial and may involve prescription of opioids.
- Hydration should be continued where possible (see page 23).
- The person's mouth should be kept clean and moist.
- There should be adequate physical and emotional support for the family and main carer.

Every attempt should be made to maintain communication with the person with MND, even in the final stages of life. Eye movements and questions that only need a single word or yes/no answer may be used, alongside picture/alphabet boards or other communication aids as appropriate.³⁶ It is believed that hearing and touch are the last senses we are aware of, so talking reassuringly and touching someone is a natural and human expression of compassion.

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 – emotional, physical and spiritual exhaustion from witnessing and absorbing the problems of others – can impact on professionals caring for those with MND.³⁷

When you work with a person over a period of time, you will get to know them and it may be distressing when they die. This is not unusual: many professionals feel this way after building a relationship with a person. It is important to look after yourself and work on building resilience. Good multidisciplinary teamwork, including regular meetings and/or supervisions, is necessary to provide support and opportunities to discuss concerns and responses to difficult situations.

Supporting family and carers

MND is unpredictable and may progress rapidly, with death occurring more quickly than anticipated.⁷

- Carers and family may need practical and emotional support.¹⁴ Offer information about sources of support, including support groups, online forums and counselling or psychology services.
- Advice should be given to carers and family members about the appropriate care and management in certain situations. This can avoid 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 is likely to meet the criteria for continuing healthcare funding and health professionals are in a position to complete the initial screening tool for this.
- Professionals can ensure adequate support for the carer when MND becomes more complex.
- Taking time to discuss end of life with the family as early as possible can be positive, as it allows them to look at their lives together, achieve things that are important to them and tie up loose ends.
- Care plans and information must be shared by all members of the care team and adequate nursing cover maintained.
- Early and sustained symptom control is essential in the management of a peaceful and dignified death.⁷
- Ideally, 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 to identify and support the needs of unpaid carers providing end of life care at home. Access the tools at www.edgehill.ac.uk/carers

Preparing the family for end life

When someone is nearing the end of life, families often need to talk about what is happening. The person who is dying should feel able to discuss their concerns and fears, which may or may not be shared with the family. Some families may need the specialist support of a family support worker, perhaps from the local hospice.

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.

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 you can share

Our resources for young people affected by MND include:

- *When someone close has MND*: an interactive workbook designed to promote coping strategies for children aged four to 10 years.
- *So what is MND anyway?* a booklet designed to introduce MND to young people, including a section on being a young carer.

Call MND Connect on **0808 802 6262** to order copies.



Carers are just as important as the person with MND. Their fears are for themselves as much as the person they are caring for. They need their own support.”

A person with MND

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 financial viability and there are many sources of support.

See our care information sheet 10A – *Benefits and entitlements*. Details of how to order can be found on page 30.

Concerns about inherited MND

Inherited MND (sometimes known as familial MND) accounts for approximately 5-10% of all people with MND.³⁸ The neurologist may refer those affected on to genetic counselling.

You may want to see our Research sheet B – *Inherited motor neurone disease*, which is split into three parts:

- 1 – *Introduction to inherited motor neurone disease*
- 2 – *Genetic testing and insurance*
- 3 – *The options available when starting a family.*

Download from our website at www.mndassociation.org/researchsheets or see page 30 for details of how to order copies. You can also contact our research team with any queries on **01604 611880** or research@mndassociation.org



My main concerns are my wife and my son ... it's more of a concern for them really than myself ... How my wife is going to be in the future is obviously a great concern to me.” A person with MND

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Further reading

Difficult conversations: Making it easier to talk about end of life with people affected by motor neurone disease. National Council for Palliative Care, with support from the MND Association. £5. ISBN 978-1-898915-94-2.

End of life care in long term neurological conditions: a framework for implementation. National End of Life Care Programme 2010.

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One chance to get it right: improving people's experience of care in the last few days and hours of life. Leadership Alliance for the Care of Dying People. June 2014.

How the MND Association can support you

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

MND Connect

Accredited by the Helplines Standard, MND Connect offers information and support, and signposting to other services and agencies.

Telephone: **0808 802 6262**

Email: **mndconnect@mndassociation.org**

Information resources

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

MND Association website

Access further information at **www.mndassociation.org/professionals**

MND Association membership

Join us now and help fight MND. Membership costs £12 for individuals. It is free for people with MND and for carers, spouses or partners of people living with MND. Call **01604 611855** or email **membership@mndassociation.org**

MND support grants and equipment loan

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

Our equipment loan service is focused on three core items:

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

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

Referrals for support grants or equipment loan need to be made by a relevant health or social care professional. Call the Support Services team on **01604 611802**, email support.services@mndassociation.org or visit www.mndassociation.org/getting-support

Research into MND

We fund and promote research that leads to new understanding and treatment and brings us closer to a cure.

Contact the Research Development team on **01604 611880** or research@mndassociation.org. Alternatively, visit www.mndassociation.org/research

For the latest research news, visit our research blog at www.mndresearch.wordpress.com

Our peer-to-peer research and care community blog (RECCOB) has a number of reporters who write updates on MND-related workshops and events at www.reccob.wordpress.com

International Symposium on ALS/MND

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

Regional care development advisers

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

MND care centres and networks

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

Branches and groups

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

Association visitors (AVs)

Association visitors are volunteers with experience of MND who provide one-to-one local support to people affected by MND.

Other organisations

Childhood Bereavement Network

Provides guidance, information and directions to support services.

020 7843 6309 or **www.childhoodbereavementnetwork.org.uk**

Cruse Bereavement Care

Cruse is the largest bereavement charity in the UK, offering information and support. **0844 477 9400** or **www.cruse.org.uk**

Dying Matters

This organisation offers a wide range of resources to help people start conversations more easily about death, dying and bereavement.

0800 021 4466 or **www.dyingmatters.org**

The Gold Standards Framework

A standard for the improvement of end of life care.

www.goldstandardsframework.org.uk

The National Council for Palliative Care

A charity supporting those involved in palliative, end of life and hospice care in England, Northern Ireland and Wales.

020 7697 1520 or **www.ncpc.org.uk**

Feedback form

Please send us your feedback on *A professional's guide to end of life care in motor neurone disease (MND)*

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

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

What is your profession or specialism?

Did you find this resource useful?

Yes Somewhat Not really No

Please explain your answer

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

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

Continued overleaf

Feedback form continued

Were there any particular topics that were useful to you?

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

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

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

Yes (please include your email address below) No

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

Yes (please include your email address below) No

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

Name:

Email:

Acknowledgements

Thank you to the following people for their valuable contributions to this booklet:

**Dr Emma Husbands, Consultant in Palliative Medicine,
Gloucestershire Hospitals NHS Foundation Trust**

**Katy Harrison, End of Life Care Facilitator, Cambridgeshire and
Peterborough NHS Foundation Trust**

**Gabrielle Tilley, Community Education and Development Manager,
Great Western Hospitals NHS Foundation Trust**

Thank you also to staff from LOROS Hospice, Leicester, for their input.

We are grateful to the following for their work on a previous version of this booklet:

**Dr David Oliver, Honorary Reader, Centre for Professional
Practice, University of Kent, UK and Locum Consultant in Palliative
Medicine, Wisdom Hospice, Rochester, UK**

If you'd like to help us by reviewing future versions of this or other resources, please email us on **infofeedback@mndassociation.org**

About us

The MND Association was founded in 1979 by a group of volunteers with experience of living with or caring for someone with MND. Since then, we have grown significantly, with an ever-increasing community of volunteers, supporters and staff, all sharing the same goal – to support people with MND and everyone who cares for them, both now and in the future.

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

Our mission

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

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

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

MND Association

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@mndeducation



/mndassociation



About MND

- MND is a fatal, rapidly progressing disease that affects the brain and spinal cord.
- It attacks the nerves that control movement so muscles no longer work. MND does not usually affect the senses such as sight, sound and feeling.
- It can leave people locked in a failing body, unable to move, talk and eventually breathe.
- It affects people from all communities.
- Some people may experience changes in thinking and behaviour, with a proportion experiencing a rare form of dementia.
- MND kills a third of people within a year and more than half within two years of diagnosis.
- A person's lifetime risk of developing MND is up to 1 in 300.
- Six people per day are diagnosed with MND in the UK.
- MND kills six people per day in the UK.
- It has no cure.

mndconnect
0808 802 6262
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