Motor neurone disease (MND) in acute, urgent and emergency care
As most people with MND are cared for at home, if an acute incident occurs, they will need to access crisis intervention services.¹

Whatever your role in acute, urgent or emergency care, this information is designed to give succinct guidance on actions you need to take and things you need to consider when treating someone with the condition.

This information includes actions for both community care/treatment and hospital settings.

For more information about MND, contact MND Connect:

**0808 802 6262**
**mndconnect@mndassociation.org**

The helpline is open to professionals 9am to 5pm Monday to Friday.

**On hospital admission of a person with MND, an MND specialist (nurse, neurologist or care centre coordinator, who may or may not be the person’s usual contact) should be alerted within 24 hours.**

Please use the space below to record local contacts who may provide support in MND emergencies.

<table>
<thead>
<tr>
<th>Service</th>
<th>Contact number/extension</th>
</tr>
</thead>
<tbody>
<tr>
<td>MND specialist nurse, care centre or network</td>
<td></td>
</tr>
<tr>
<td>Respiratory support for people with MND</td>
<td></td>
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<tr>
<td>Hospital speech and language therapy support</td>
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<tr>
<td>Hospital dietetics support</td>
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<td>Hospital physiotherapy support</td>
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<tr>
<td>Hospital occupational therapy support</td>
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<tr>
<td>Hospital palliative care team</td>
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<tr>
<td>Community palliative care team</td>
<td></td>
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<tr>
<td>Local hospice (for out-of-hours advice)</td>
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</tbody>
</table>
People with MND may need emergency care for a number of reasons, related or unrelated to their condition. Particular issues related to MND may include:

**Chest infection**
The leading cause of hospital admission in MND\(^2\) – see page 5 for Respiratory symptoms.

**Acute respiratory distress**
See page 5 for Respiratory symptoms.

**Choking caused by mucus build-up or problems swallowing food, water or saliva**
See page 8 for Dysphagia/choking/enteral feeding tube problems.

**Problems with a blockage in feeding tube**
See page 8 for Dysphagia/choking/enteral feeding tube problems.

**Dehydration due to swallowing difficulties**
See page 8 for Dysphagia/choking/enteral feeding tube problems.

**Collapse of social care package/carer illness**
See page 9 for Social care breakdown/carer illness.

**End of life**
See page 10.

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For drug dosages please refer to the British National Formulary (BNF) or Palliative Care Formulary (PCF), or liaise directly with pharmacy and/or palliative care/local hospice.
Key points when you have a patient with MND

In all acute settings

**Oxygen**
See caution on oxygen therapy (opposite) before considering this course of action.

**Does the patient have a record of these documents?**
- Emergency healthcare plan
- ReSPECT – a summary of recommendations to help make immediate decisions about a person’s care and treatment
- Advance care plan – this includes preferences for care, including place of care and end of life decisions.
- Advance Decision to Refuse Treatment (ADRT) – if so, this must be considered.
- Do Not Attempt Cardiopulmonary Resuscitation (DNACPR) – if so, this must be considered.
- MND Alert Card or Understanding My Needs – These include key contacts and information about their care team and the specialist help they may need.

**Communication** - this can take great effort in MND. The person may use communication aids.

**Does the patient use a non-invasive ventilator?** If airways are clear, reapply NIV to treat breathlessness.

**Does the patient have capacity to make key decisions for themselves?** See page 13.

In emergency medicine

- **Is the patient pyrexial and taking riluzole?**
  Check full blood count (FBC) for signs of neutropenia and liver function tests (LFTs) for sign of disturbance, especially alanine transaminase (ALT).

- **The patient may not be able to lie flat for x-rays/scans** – due to decline of respiratory function in MND.

- **Locate the next of kin** – Once located, take account of their insight into the person’s condition and care needs.

- **Does the patient have an enteral feeding tube?**
  This will affect decisions around administration of medication (including route and type) and fluids.

- **Consider the administration route of any drug you choose to prescribe** – especially if the patient has swallowing difficulties and no enteral feeding tube.

- **Contact your local MND care centre/network for specialist advice and support**
  Where available, contact the specialist team in your hospital or local area.
Respiratory symptoms

Signs/symptoms may include:
• shortness of breath
• orthopnoea
• acute panic
• signs of CO\textsubscript{2} retention, including increased drowsiness, increased/irregular respiratory rate, abdominal paradox, use of accessory muscles of respiration, anxiety, confusion
• signs of respiratory infection
• inability to clear bronchial secretions.

Rarely, patients may experience respiratory distress without other symptoms and MND may not yet have been diagnosed.\textsuperscript{2}

Investigations:
• full physical examination
• temperature
• O\textsubscript{2} saturation
• bloods: FBC, U&Es, LFTs, arterial blood gas (ABG)
• chest x-ray.

Check if the patient has an ADRT, as this will impact on any treatment you give.

CAUTION: Inappropriate ventilation/against patient wishes
• Ideally, discussions around ventilation will have taken place between the patient and respiratory specialist and recorded on an advance care plan.\textsuperscript{3}
• Where appropriate, a respiratory specialist should explore options for assisted ventilation. In an acute situation, an appropriate professional should make the call on whether to introduce NIV, then seek advice about long-term management.
• In case of intubation, extubation may be problematic: seek advice from the home ventilation team. The course of action may be to wean on to NIV, unless there is significant bulbar weakness. Ventilation via tracheostomy has implications for future care and is rarely the first choice of action.\textsuperscript{2}
• Where ventilation is not used, medication (including titrated opioid analgesics) may be used to palliate breathlessness.

CAUTION: Oxygen therapy in MND
Low O\textsubscript{2} saturation in MND is frequently due to neuromuscular weakness causing rapid CO\textsubscript{2} retention.\textsuperscript{4} Non invasive or invasive support is more useful than supplementary oxygen, as they are a high risk of hypercapnic respiratory failure.

Respiratory problems should be monitored by arterial blood gas analysis. O\textsubscript{2} therapy (unless as a palliative measure) should be used with great caution in patients with MND-related respiratory problems.\textsuperscript{25}

Seek guidance from the specialist palliative care team or a respiratory consultant with links to neurology.
**Actions/reminders for all acute settings**

**Signs of respiratory failure**
- Does the patient have an ADRT or DNACPR?
- If the patient uses non-invasive ventilation, this may help to palliate breathlessness.

**Panic/acute distress**
- This may be a physiological response to breathing problems. It is vital to treat the symptom.
- Lorazepam and morphine salts (Oramorph) may help. In acute situations, buccal midazolam may be indicated.\(^5\)
- Reassure that death from choking is rare.

**Signs of infection**
- Does the patient have an ADRT?
- Discuss with patient whether they wish to have antibiotic treatment.
- Treat symptoms of infection: pyrexia, discomfort, shortness of breath.
- Morphine salts may help ease the sensation of breathlessness.\(^6\)

**If the patient is 24-hour dependent on NIV and presents with breathlessness**
- Treatment may involve changes to NIV settings.
- Liaise with the home ventilation team.

**Actions/reminders in emergency medicine**

**Signs of respiratory failure**
- Contact the on-call respiratory specialist for urgent review.
- Is non-invasive ventilation an option? The respiratory specialist will explore this, if appropriate, with the patient. People may not want this option as it may prolong life.\(^3\)

**Difficulty clearing thick bronchial secretions**
- Ensure the person is hydrated.
- Consider carbocisteine (liquid preparation for those with swallowing problems. Contraindicated in those with gastric ulceration).\(^7\)
- Humidification or a saline nebuliser (driven by pressurised air, not O\(_2\) ) can moisten secretions.\(^8\)
- Contact the on-call physiotherapist for techniques to clear secretions. Ongoing support may be needed from the respiratory team to improve cough through breath stacking, manual assisted cough or mechanical insufflation-exsufflation.\(^2\)
- Liaise with respiratory specialist around referral to specialist teams.

**Is the patient known to the local specialist MND respiratory service?**
- If so, liaise with this team.
Dysphagia/choking/enteral feeding tube problems

Signs/symptoms may include:
• loss of swallow
• acute choking episodes due to build up of mucus/food/liquids, which may lead to aspiration pneumonia
• blockage of enteral feeding tube
• excessive saliva
• dehydration.

Investigations:
• full examination and bloods: FBC, U&Es
• abnormal levels of electrolytes need to be corrected, eg with calcium gluconate or phosphate
• swallow function assessed by SLT
• check patency of enteral feeding tube.

Acute choking

Actions/reminders for all acute settings
• Clear the blockage (saliva, food or fluids).
• Palliation may be required in case of distress (see suggestions for panic/acute distress).

Actions/reminders in emergency medicine
• Intensive chest physiotherapy may be needed.
• SLT will assess whether swallow is safe and if it is not, will advise food texture to minimise risk.

Saliva control problems
• Ask the patient whether saliva has been thick, thin or a mixture of secretion types.
• Treatment of this symptom may be difficult to address. Dosage must be monitored, as treatment can dry the saliva too much so it sticks in the throat.
• Antimuscarinic medication can be used to dry up saliva. Consider glycopyrrrolate for people with cognitive impairment.

Loss of swallow
• This will have been developing for some time and is unlikely to be a sudden change.
• Does the advance care plan include a decision about enteral feeding? The patient may have discussed this with their community speech and language therapist, dietitian and local care team, but may have delayed the decision or may change their mind when swallow is lost.
• Ask the patient if they want to have enteral feeding. If they agree and a respiratory function assessment makes them suitable, contact the enteral feed dietitian/nutrition team/upper GI specialist on call.
Social care breakdown/carer illness

People with MND living at home may have or need large packages of care and a lot of on-going support. Because MND is progressive, the care package needs to be regularly reviewed and upgraded when needed.

Unfortunately, packages of care can break down, or family members become unwell or cease to cope. Sudden change can lead to acute admission.

- Let the MND care coordinator know about the admission: see inside front cover for contact details.
- Determine the patient’s care needs, home situation, current care package and family/carer situation.
- Find out what the patient and carer would need for discharge to be facilitated.
- Does the patient have social care funding or continuing healthcare funding?
- What short-term care placements are available? For example: hospital admission (if there is medical need), local GP hospital unit/cottage hospital, nursing home, residential care, hospice, care in a family member or friend’s home etc. The hospital discharge team may be able to increase the care package or arrange a placement. The person’s palliative care team may be able to arrange hospice or nursing care cover.

Actions/reminders in emergency medicine

If the patient does not want enteral feeding
- Contact the hospital palliative care team for ongoing support during office hours or speak to the local hospice out of hours.

Enteral feeding tube blockage
- Call the nutrition team/upper GI specialist to address.

Dehydration
- This can be a problem for people who do not want enteral feeding or fluids.
- Any decision to treat this with IV fluids, in line with the wishes of the patient, must only be considered a short-term treatment, particularly to allow the patient to get home for end of life care.9
- The patient needs to be aware of this and be able to give informed consent.

MND Association professional information
- Dysphagia information sheet
- Managing saliva problems information sheet
See page 16 for how to order resources.
# End of life

Even if someone with MND has recorded a wish to die elsewhere, for example at home, they may be seen in emergency medicine as end of life approaches.

- The aim in this situation is to facilitate where the person's preferred place of care is. They may feel, once no more can be done, that they want to get home as soon as possible.
- The patient may be scared and think there is something more that can be done to prolong their life.
- The carer may feel unable to cope.
- They may wish to be an in-patient, or be transferred to a hospice.
- Some patients with MND may die in an acute setting. In those cases, the aim is to ensure the best possible care is achieved.

## Assess all symptoms:  

<table>
<thead>
<tr>
<th>Pain – patients may experience severe discomfort and/or distress, particularly in the later stages.</th>
<th>Medication to consider</th>
</tr>
</thead>
</table>
| • analgesics to manage pain, including carefully titrated opiates (morphine salts, diamorphine)\(^{10, 11, 12}\)  
• sedatives to manage distress (lorazepam, midazolam)\(^{13}\) | |

<table>
<thead>
<tr>
<th>Nausea – may be due to enteral feeding, or some medications.</th>
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<tbody>
<tr>
<td>• anti-emetics (levomepromazine)(^{14})</td>
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<tr>
<th>Salivation</th>
<th></th>
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</thead>
<tbody>
<tr>
<td>• antimuscarinics (glycopyrronium bromide/hyoscine hydrobromide) for excessive respiratory secretions(^{14})</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Anxiety/depression</th>
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<tbody>
<tr>
<td>• sedatives such as diazepam, midazolam and lorazepam to reduce anxiety.(^{13}) Use with some caution, as they may adversely affect respiratory drive. However, this should not be the sole reason to avoid treating symptoms in end of life care.</td>
<td></td>
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<table>
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<tr>
<th>Dyspnoea/orthopnoea</th>
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<tbody>
<tr>
<td>• opioid analgesics (morphine salts, diamorphine) to reduce cough reflex, control pain, relieve dyspnoea (breathlessness), and therefore fear and anxiety(^{12})</td>
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</tbody>
</table>
Death in MND
The most common cause of death in MND is respiratory failure, often with additional chest infection. Death in the majority of cases is peaceful, following lengthening periods of sleepiness, gradually resulting in unconsciousness and death. Very occasionally, this process may occur suddenly.

Palliative care services should be involved as early as possible to support people nearing end of life.

Also assess for:

- pressure area care
- dysphagia
- insomnia
- restlessness/agitation (consider urine retention and constipation as causes)
- effective elimination of bowels and bladder
- reassessment of emotional/spiritual and practical needs of patient, carer and family.

Delivery of suitable medications via a syringe driver may maximise symptom control.2

Oxygen should only be given to people with MND in specific circumstances (see page 5) but may be given along with sedatives at end of life.2

Communicating with the person with MND
This may become extremely difficult as the patient with MND reaches end of life, but even if they are unresponsive, every attempt should be made to maintain communication (see page 15).15

Withdrawal of non-invasive ventilation (NIV)
As respiratory muscle weakness progresses, there will be a point when NIV is no longer effective. Some people may continue to use NIV for palliation of symptoms of breathlessness.

Some may have specified within their advance care plan when they would want the treatment to be withdrawn. This may be when symptoms reach a particular point. In some cases, NIV will be withdrawn by the medical team due to its ineffectiveness.

If the time comes for NIV to be withdrawn, it should be done with supervision from a palliative care doctor, pre-medication and support for the person with MND and their family.2

Seek advice from the respiratory team on call.

The MND Association’s End of life guide
This booklet for people with MND may answer some of your questions. See page 16 for how to order resources.
# Pain

<table>
<thead>
<tr>
<th>Pain may be caused by:</th>
<th>Medication options:</th>
</tr>
</thead>
</table>
| **Muscle cramps**      | Consider quinine as first-line treatment. Quinine sulphate is not associated with the problems of sedation seen with benzodiazepines.³  
|                        | If quinine is not effective, not tolerated or contraindicated, consider baclofen instead as a second-line treatment.³  
|                        | If baclofen is not effective, not tolerated or contraindicated, consider tizanidine, dantrolene or gabapentin.³  
|                        | Warning: dosage of muscle relaxants should be carefully adjusted to avoid increased weakness, and decreased mobility.¹² |
| **Muscle stiffness, spasticity or increased tone** | Consider baclofen, tizanidine, dantrolene or gabapentin.³  
|                        | If these treatments are not effective, not tolerated or contraindicated, consider referral to a specialist service for the treatment of severe spasticity.³ |
| **Mechanical stress on joints from muscle weakness** | For joint pain: simple analgesia, eg long-acting non-steroidal anti-inflammatory drugs (NSAIDs).¹² |
| **Occasionally, neuropathic pain** | tricyclic antidepressants  
|                        | gabapentin/pregabalin.¹⁶ |

Other potential causes:  
- **skin sensitivity** – good skin pressure care is vital  
- **immobility** – care should be taken when positioning, handling and lifting  
- **oedema** – diuretics are rarely helpful, as they can promote urinary urgency and electrolyte disturbance  
- **constipation**.
Cognitive change and decision-making

There is now increased awareness of cognitive and behavioural changes in MND, and people with MND may fall into one of four groups:

• around 50% are unaffected by cognitive change
• around 35% experience mild cognitive and/or behavioural change, with specific deficits in executive functions, language and/or social cognition\(^{17,18,19}\)
• up to 15% develop frontotemporal dementia (FTD), either at the same time or after diagnosis of MND\(^{20}\)
• up to 15% of patients with FTD go on to develop motor impairments where MND is diagnosed after dementia.

Mild cognitive impairment and/or behavioural change tends to include subtle changes that may have little impact on daily life.\(^{21}\)

Those with MND and frontotemporal dementia (MND-FTD) will show marked cognitive change. Some will exhibit challenging behaviour.\(^{21}\)

Cognitive change can have implications for decision-making.\(^{23}\) Ideally, advance care planning will have taken place and documentation will exist of the patient’s wishes for care and treatment.

Consenting to treatment in an emergency

In an emergency situation, it’s not always possible to find out a patient’s wishes in terms of treatment. As with any emergency, treatment can be carried out without consent if it is immediately necessary to save their life or prevent a serious deterioration of their condition.\(^{22}\)

However:

• someone with MND may have expressed a wish to refuse treatment that could potentially prolong life
• interventions such as invasive ventilation via tracheostomy, for example, have serious implications for future care and survival for people with MND.\(^{2}\)

Where possible, it is essential to support and encourage patients to be involved, as far as possible, in decisions about their care.

MND Association professional information

• **Cognitive change, frontotemporal dementia and MND** booklet

We also have three information sheets for people affected by MND on cognitive change and managing emotions (9A-9C). See page 16 for how to order.
Communicating with someone with MND

It may take great effort for a person with MND to communicate and, in the case of an emergency admission, this may be even more difficult, if not impossible.

Where it is possible to communicate with the person with MND, it is vital to make the effort to do this in order to understand their individual needs.

Do:
• find out how the person with MND prefers to communicate and any equipment they like to use
• find out whether the person uses a simple code for “yes” and “no”. This can be a movement of any part of the body that can be carried out reliably, such as eye blinking
• remember that unspoken communication is important
• ensure the person with MND is the focus of communication
• check back with the person on what you think has been said and admit when you don’t understand.

Try not to:
• alter the rate or sound of your speech, unless the person with MND has asked you to
• finish the sentences of the person with MND, unless they ask you to, and avoid interrupting them
• use a family member or carer as a translator for the person with MND, unless it is clear that this is what the person with MND wants
• ask complex questions that require long or difficult answers and take up unwelcome time and energy
• use multiple choice questions. Simple questions that can be answered with yes, no, or a single word are easier when speech is difficult.
How the MND Association can support you and your team

The MND Association is the only national charity in England, Wales and Northern Ireland focused on MND care, research and campaigning.

We support health and social care professionals to provide the best possible care for people living with MND, their carers and families. Here are some of the ways we do this:

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

The service is for people living with MND, carers, family members and health and social care professionals.

Freephone **0808 802 6262**
Email mndconnect@mndassociation.org

Please contact MND Connect if you have any questions about the information in this publication.

**Education and training**
Training in specific areas of care for people with MND may be provided by relevant members of the specialist MND team.

Regional staff from the MND Association can also offer awareness training about MND and the Association’s services, as well as more detailed study days. Visit [www.mndassociation.org/education](http://www.mndassociation.org/education)

**Information resources**
We produce high quality information resources for health and social care professionals who work with people with MND and have a wide range of resources for people living with and affected by MND.

Downloads of all our information sheets and most of our publications are available from our website at [www.mndassociation.org/publications](http://www.mndassociation.org/publications)

You can also order our publications direct from the MND Connect team.
Research into MND
We fund and promote research that leads to new understanding and treatment and brings us closer to a cure. Find out more at www.mndassociation.org/research

Local support

Regional Care Development Advisers
We have a network of regional care development advisers (RCDAs) covering England, Wales and Northern Ireland.

RCDAs have specialist knowledge on the care and management of MND. They work closely with local statutory services and community care providers to ensure effective support for people affected by MND. RCDAs provide education on MND, and are champions at influencing care services in their respective areas.

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

Care centres are not physical buildings, but teams of professionals who are specialists in MND.

Association visitors (AVs)
Association visitors provide one-to-one local support to people affected by MND. Managed by regional care development advisers, they are volunteers with experience of MND who can visit people affected by MND within their homes or contact them by telephone, email or through a local support group. They can also provide a link with care centres and other health and social care professionals.

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


18 Crockford C et al. ALS-specific cognitive and behaviour changes associated with advancing disease stage in ALS. *Neurology*. 2018; 91(15):e1370-e1380.


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Jasmine Harwood, Paramedic, South East Coast Ambulance Service.
Sue Jones, Advanced Paramedic, Member of the College of Paramedics.

If you’d like to help us by reviewing future versions of this or other resources, please email us at infofeedback@mndassociation.org
MND at a glance

• Motor neurone disease (MND) is a progressive, terminal, neurological disease that attacks the motor neurones in the brain and spinal cord.\(^2\)

• Around two people in every 100,000 of the general population will develop MND each year.\(^2\)

• MND can affect how people walk, talk, eat, drink and breathe.

• Not all symptoms happen to everyone and it is unlikely they will all develop at the same time, or in any specific order.

• It affects people from all communities.

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

• There is no cure for MND, but symptoms can be managed to help improve quality of life.\(^2\)

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.