MND Association | For professionals

MND in acute, urgent and emergency care

About Motor Neurone Disease

Today, six people

will be told they have MND and six people will die from the disease. There is no cure.

MND is a fatal, rapidly progressing disease.

It affects the nerves in the brain and spinal cord that control muscles.

A third of people will die within a year of diagnosis.

More than half will die within two years.

People may lose movement, speech, swallowing and breathing.

This affects quality of life for them and those around them.

MND doesn’t discriminate.

It affects people from all backgrounds and at all ages.

People with MND may experience changes in thinking and behaviour.

Some develop frontotemporal dementia, with more severe changes.

MND affects everyone differently.

Symptoms progress at different speeds and a different order for everyone.

Contents

Introduction

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.

For more information about MND, contact MND Connect:

Tel: 0808 802 6262
Email: mndconnect@mndassociation.org

The helpline is open to professionals 9am to 4pm Monday to Friday.

On hospital admission of a person with MND, an MND specialist (nurse, neurologist or care centre coordinator) should be alerted within 24 hours.

MND Paramedic's Card for JRCALC

We have produced an information card on MND which slots inside the JRCALC.

Other resources

We have a wide range of information resources on the symptoms mentioned in this booklet, both for professionals and for people living with MND.

See page 31 for details of how to order publications.

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

Service	Contact details
MND specialist nurse, care centre or network	
Respiratory support for people with MND	
Hospital speech and language therapy support	
Hospital dietetics support	
Hospital physiotherapy support	
Hospital occupational therapy support	
Hospital palliative care team	
Community palliative care team	
Local hospice (for out-of-hours advice)	

Key points for acute care of someone with MND

In all acute settings

Oxygen

See caution on oxygen therapy (page 11) before considering this course of action.

Does the person 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. Sometimes also known as an Allow Natural Death order (AND).
- 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 person use a non-invasive ventilator?

If so, use it to manage symptoms of dyspnoea and help prevent or treat worsening hypercapnia.

Does the person have capacity to make key decisions for themselves? See page 20.

In emergency medicine

- Is the person pyrexial and taking riluzole? Check full blood count (FBC) for signs of neutropenia and liver function tests (LFTs) for signs of disturbance, especially alanine transaminase (ALT).
- The person may not be able to lie flat for x-rays or scans due to decline of respiratory function in MND. Propping the person at a 30-45° angle or using NIV if they already have it will help. Additional tubing may be required to facilitate MRI scanning.
- Locate the next of kin and Lasting Power of Attorney (if appropriate). Once located, take account of their insight into the person's condition and care needs.
- Does the person 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 person has swallowing difficulties and no enteral feeding tube.
- Contact your local MND care centre or network for specialist advice and support
- If the person has been admitted with COVID-19, ensure riluzole is stopped until all bloods back to within normal limits.

Communicating with someone with MND

Communication may require significant effort for a person with MND and may be difficult or impossible during an emergency admission. Where communication is possible, it is essential to make every effort to understand the person's 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 any reliable movement, such as eye blinking
- remember that unspoken communication is important
- ensure the person with MND is the focus of communication
- check 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 unless the person with MND has asked for this
- ask complex questions that require long or difficult answers
- use multiple choice questions. Yes or no, or single-word questions are easier to answer.

Respiratory symptoms

Signs and symptoms of respiratory compromise may include:

- shortness of breath
- orthopnoea
- acute panic
- signs of CO_2 retention, including drowsiness, anxiety, confusion, bounding pulse
- increased phlegm production or phlegm which is mucopurulent.
- falling oxygen saturations
- increased respiratory rate
- inability to clear bronchial secretions.

Rarely, people may experience respiratory distress without other symptoms and MND may not yet have been diagnosed.[2]

Investigations:

- full physical examination
- temperature
- O_2 saturation
- bloods: full blood count, urea and electrolytes, liver function tests, arterial blood gas
- chest x-ray.

Check if the person has an ADRT or Lasting Power of Attorney, as this might impact on any treatment you give.

Inappropriate ventilation

Discussions about ventilation should ideally take place in advance between the person with MND and a respiratory specialist, and be documented in an advance care plan.

In acute situations, NIV may be initiated with specialist input for ongoing management.

If intubation occurs, extubation can be difficult. Seek advice from the home ventilation team. Weaning to NIV may be appropriate unless there is significant bulbar weakness. Tracheostomy ventilation is rarely first-line and has major implications for care.

Where ventilation is not used, manage breathlessness with medication, including titrated opioid analgesics.

Oxygen therapy in MND[4]

MND causes respiratory muscle weakness, so oxygen saturations may be lower than expected. Oxygen saturations in a person with MND of between 88% and 92% are acceptable unless other pathology is present. Oxygen does not help breathlessness.

Supplemental oxygen should only be used with extreme caution in MND as excessive oxygen can lead to acidaemic respiratory failure and death. Respiratory problems should be monitored by arterial blood gas analysis. Seek guidance from the specialist palliative care team or a respiratory consultant with links to neurology.

See our video on oxygen use at **bit.ly/MND-Oxygen**

Actions and reminders for all acute settings

Signs of respiratory failure

- Does the person have an ADRT or DNACPR?
- If the person uses non-invasive ventilation, this may help to palliate breathlessness.

Panic and 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.[5,6] Intravenous or subcutaneous routes should be considered when appropriate.
- Some people may experience distressing sensations of choking or mild choking episodes. Reassure that death from choking is rare.

Signs of infection

- Does the person have an ADRT?
- Discuss with person whether they wish to have antibiotic treatment.
- Treat symptoms of infection: pyrexia, discomfort, shortness of breath.
- Morphine salts may help ease sensations of breathlessness.[6]

If the person is 24-hour dependent on NIV and presents with breathlessness

- Treatment may involve changing the settings on the NIV.

- Liaise with the home ventilation team.

Actions and 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 person. 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 person known to the local specialist MND respiratory service?

- If so, liaise with this team.

Dysphagia and enteral feeding issues

Issues may include:

- loss of swallow
- acute choking episodes due to build-up of mucus, food or liquids
- blockage of enteral feeding tube
- excessive saliva
- dehydration.

Investigations:

- full examination and bloods: full blood count, urea and electrolytes
- abnormal levels of electrolytes need to be corrected
- swallow function assessed by SLT
- check patency of enteral feeding tube.

! Acute choking

Actions and reminders for all acute settings

- Clear the blockage (saliva, food or fluids).
- Palliation may be required in case of distress (see suggestions for panic and acute distress).

Actions and reminders in emergency medicine

- Intensive chest physiotherapy may be needed.
- SLT will assess if swallow is safe and advise on risks.

Actions and reminders in emergency medicine

Saliva control problems

- Ask the person whether saliva has been thick, thin or a mixture of secretion types.
- Antimuscarinic medication can be used to dry up saliva. Consider glycopyrronium as the first line treatment.[3]
- Treatment of this symptom may be difficult to address. Dosage must be monitored, as treatment can reduce saliva too much, causing it to become thick and difficult to clear.

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 person may have discussed this with their MND team, but may have delayed the decision or may change their mind when swallow is lost.
- Ask the person 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 or upper GI specialist on call.
- Consider short term nasogastric tube feeding until enteral feeding tube can be inserted.

Actions and reminders in emergency medicine

If the person 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 or 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 person, must only be considered a short-term treatment, particularly to allow them to get home for end of life care.[9]
- The person needs to be aware of this and be able to give informed consent.

Social care breakdown or 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 page 5 for contact details.
- Assess the person's care needs, home circumstances, current care package, and family and carer support.
- Find out what the person with MND and carer would need for discharge to be facilitated.
- Does the person have social care funding or NHS 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.

Pain

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]
- Dosage of muscle relaxants should be adjusted to avoid increased weakness, and reduced mobility.

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]

Neuropathic pain

- tricyclic antidepressants
- gabapentin or pregabalin.[16]

Mechanical stress on joints from muscle weakness

- Simple analgesia, eg long-acting non-steroidal anti-inflammatory drugs (NSAIDs).[12]

Skin sensitivity

- Good skin pressure care is vital.

Immobility

- Care should be taken when positioning, handling and lifting. Passive limb exercises should be carried out to avoid pain from immobility. Contact physiotherapy for advice.

Oedema

- Diuretics are rarely helpful, as they can promote urinary urgency and electrolyte disturbance.

Constipation

- Avoid laxatives that need large volumes of water, such as Laxido or Movecol, as person with end stage MND is unlikely to be able to tolerate it and may feel more nauseous.

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 the following groups:

- around 50% are unaffected by cognitive change
- Around 35% experience mild cognitive change, behavioural change, or a combination of both, with specific deficits in executive function, language or social cognition.[17,18]
- up to 15% develop frontotemporal dementia (FTD), either at the same time or after diagnosis of MND.[19]

Mild cognitive impairment or behavioural change tends to include subtle changes that may have little impact on daily life.[20]

Those with MND and frontotemporal dementia (MND-FTD) will show marked cognitive change. Some will exhibit challenging behaviour.[20]

Cognitive change can have implications for decision-making.[22] Ideally, advance care planning will have taken place and documentation will exist of the person's wishes for care and treatment.

Consenting to treatment in an emergency

In an emergency situation, it's not always possible to find out the person'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.[21]

However:

- someone with MND may have expressed a wish to refuse treatment that could potentially prolong life so check for an ADRT
- 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 the person to be involved, as far as possible, in decisions about their care.

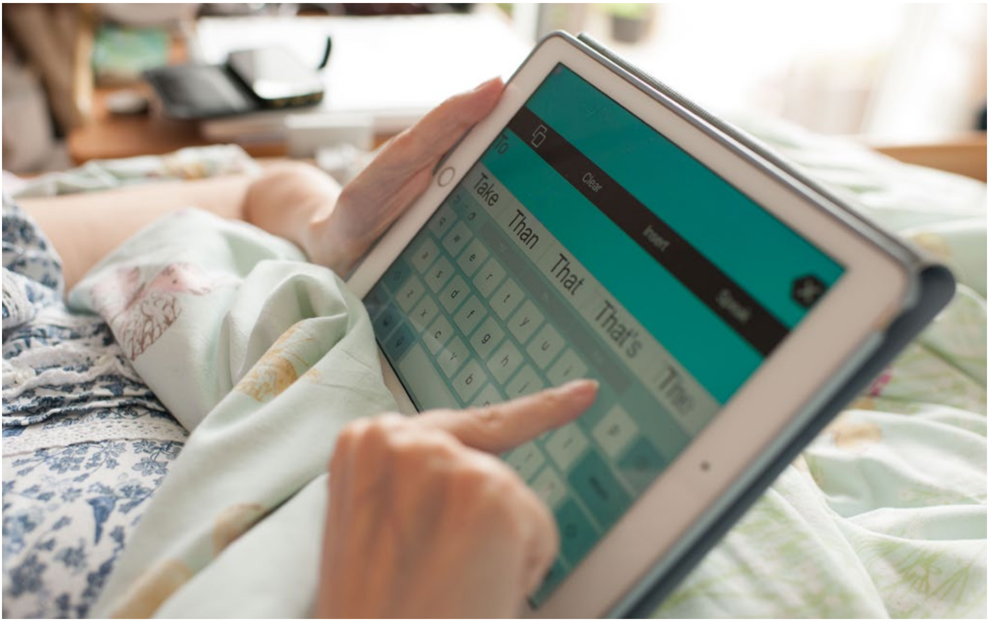
End of life

Even if a person with MND has recorded a wish to die elsewhere, such as at home, they may be seen in emergency medicine as end of life approaches. The aim is to facilitate the person's preferred place of care where possible.

Some people may feel frightened or believe there is more that can be done to prolong life, while carers may feel unable to cope. The person may wish to remain as an in-patient or be transferred to a hospice. Some people with MND will die in an acute setting. The priority is to ensure the best possible care is provided.

Communication

Communication may become extremely difficult as end of life approaches. However, even if the person appears unresponsive, every effort should be made to maintain communication (see page 9).[15]



Death in MND

The most common cause of death in MND is respiratory failure, often with additional chest infection.[2] Death is usually peaceful, with increasing sleepiness leading gradually to unconsciousness, although it may occasionally occur suddenly.

Symptom management at end of life

Pain may be severe in the later stages of MND, and may be managed with analgesics, including carefully titrated opiates such as morphine salts or diamorphine.[10,11,12] Sedatives such as lorazepam or midazolam may be used to manage distress.[13]

Nausea may be related to enteral feeding or medication and may be treated with anti-emetics such as levomepromazine.[14]

Salivation and excessive respiratory secretions may be managed with antimuscarinics, such as glycopyrronium bromide or hyoscine hydrobromide.[14]

Anxiety and depression are common at end of life. Sedatives such as diazepam, midazolam or lorazepam may reduce anxiety and should be used with caution, but not withheld at end of life.

Dyspnoea and orthopnoea may cause significant distress. Opioid analgesics, such as morphine salts or diamorphine, may reduce cough reflex, relieve pain and breathlessness, and reduce associated fear and anxiety.[12]

Clinicians should also assess pressure care, dysphagia, insomnia, and restlessness or agitation, including checking for bladder or bowel issues. The emotional, spiritual and practical needs of the person with MND, their carer and family, should be considered.

Oxygen should only be given to people with MND in specific circumstances (see page 11) but may be given along with sedatives at end of life.

Withdrawal of non-invasive ventilation (NIV)

As respiratory muscle weakness progresses, NIV may no longer be effective. Some people may continue to use NIV to relieve breathlessness, while others may have recorded wishes in an advance care plan for when treatment should be withdrawn. In some cases, NIV is withdrawn by the medical team due to lack of benefit.

When NIV is withdrawn, this should be done with supervision from a palliative care doctor, appropriate pre-medication, and support for the person with MND and their family.[2] Seek advice from the respiratory team on call.

The Association of Palliative Medicine provides guidance, 'Withdrawal of assisted ventilation at the request of a patient with MND'. This is available on their website. Visit **apmonline.org** and search 'MND'.

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How we can support you

Our MND Connect helpline offers practical and emotional support, information and signposting to people with MND, carers, family and professionals. They can also provide information about our services mentioned below.
Email: mndconnect@mndassociation.org
Tel: 0808 802 6262

MND Association website

Our website offers supporting information on MND, our work, services, and how to get involved.
mndassociation.org/professionals

Stay updated on events, publications and opportunities for health and social care professionals.
mndassociation.org/educationupdate
X: **mndeducation**
Bluesky: **mndeducation.bsky.social**

Information resources

We produce high quality information for people with MND, carers, families and professionals, available in multiple formats and languages.
mndassociation.org/pro-info-finder
mndassociation.org/careinfofinder

Education

Our education programme aims to improve standards of care and quality of life. Opportunities include webinars and face to face equipment training.
mndassociation.org/education

MND Professionals’ Community of Practice

A peer led group supporting cross disciplinary learning in MND care. Membership can contribute to CPD and offers access to networking and learning events.
mndassociation.org/cop

Local support

We offer online and local peer support, plus trained volunteers who provide practical help by phone, email or visits.
mndassociation.org/local-support

We fund and develop specialist care centres and networks across England, Wales and Northern Ireland, offering multidisciplinary care.
mndassociation.org/care-centres

Financial support

We offer a range of grants to support people living with MND, their families and unpaid carers. These are not in place of any statutory funding that should be available.
mndassociation.org/getting-support

MND register

The Register aims to collect information about everyone with MND in England, Wales and Northern Ireland to support care planning and research.
mndregister.ac.uk

Research into MND

We fund and promote research that leads to new understanding and treatments, and brings us closer to a cure for MND. We also produce information sheets on research for people with or affected by MND.
mndassociation.org/research

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We value your feedback

Your feedback helps improve our information for the benefit of people living with MND and those who care for them. Visit **smartsurvey.co.uk/s/mndprofessionals** or email your comments to **education@mndassociation.org**.

If you would like to help us by reviewing future versions of our information resources, please email us at **education@mndassociation.org**.

How to order our publications

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

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

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**Every day we support people affected by Motor Neurone Disease.
Because with MND, every day matters.**