

P6

# Evaluation and management of respiratory symptoms in motor neurone disease

Information for health and social care professionals

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## Introduction

Motor neurone disease (MND) is a progressive and terminal disease that attacks the motor neurones, or nerves, in the brain and spinal cord.<sup>1</sup> Respiratory muscle weakness eventually affects most people with MND, and ventilatory failure is the most common cause of death.<sup>2</sup>

Most commonly, respiratory symptoms occur as the condition progresses. However, very occasionally, respiratory problems are the presenting feature of MND, with a diagnosis made after an acute admission to hospital.<sup>3</sup>

### **Information to share with people with or affected by MND:**

Information sheet 8A – *Support for breathing problems*

Information sheet 8B – *Ventilation for motor neurone disease*

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

Information sheet 8D – *Troubleshooting for non-invasive ventilation (NIV)*

Information sheet 8E – *Air travel and ventilation for motor neurone disease*

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## NICE guideline on MND

The National Institute for Health and Care Excellence (NICE) have published guideline NG42 - *Motor neurone disease: assessment and management*, which aims to improve care for people with MND from the time of diagnosis through to end of life.

The guideline includes specific recommendations about respiratory function and symptoms, cough management and non-invasive ventilation (NIV). This information sheet incorporates the recommendations outlined by NICE. For more information, see [www.nice.org.uk/guidance/ng42](http://www.nice.org.uk/guidance/ng42)

### Information to share with people with or affected by MND:

Information sheet 1A – *NICE Guideline on MND*  
*What you should expect from your care pocket book*

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## Signs and symptoms of potential respiratory impairment

People with MND may not complain much of breathlessness and, as they often lack mobility, signs of breathlessness may be subtle.

The NICE guideline on MND recommends early referral to a respiratory specialist if one or more of these occur:<sup>4</sup>

<b>Symptoms</b>	<ul style="list-style-type: none"><li>• Breathlessness</li><li>• Breathlessness when lying flat (orthopnoea)</li><li>• Recurrent chest infections</li><li>• Disturbed sleep</li><li>• Non-refreshing sleep</li><li>• Nightmares</li><li>• Daytime sleepiness</li></ul>	<ul style="list-style-type: none"><li>• Poor concentration and/or memory</li><li>• Confusion</li><li>• Hallucinations</li><li>• Morning headaches</li><li>• Fatigue</li><li>• Poor appetite</li></ul>
<b>Signs</b>	<ul style="list-style-type: none"><li>• Increased respiratory rate</li><li>• Shallow breathing</li><li>• Weak cough</li><li>• Abdominal paradox (inward movement of the abdomen during inspiration/sniffing)</li></ul>	<ul style="list-style-type: none"><li>• Weak sniff</li><li>• Reduced chest expansion on maximal inspiration</li><li>• Use of accessory muscles of respiration eg pulling shoulders up to aid breath</li></ul>

Additional signs not described by the NICE guideline include:

- difficulty coordinating breathing with swallowing
- breathlessness when bending over
- reduced voice volume or being unable to complete long sentences.

Symptoms during the night are often an early indicator of respiratory inefficiency.<sup>3</sup> Ineffective breathing when lying down (orthopnoea) can lead to an imbalance in blood gases and a build-up of carbon dioxide (CO<sub>2</sub>).

As a result, the person with MND may:

- complain of waking with a headache, feeling unrefreshed or 'hungover' or not having slept well
- fall asleep during the day
- be difficult to rouse from sleep
- have difficulty achieving deep sleep, which may present as more frequent urination at night<sup>24</sup>
- seem confused when they wake up.

The person's partner may have noticed more laboured or shallow breathing patterns or apnoeic episodes during sleep, where they wake up gasping for air or periods where they stop breathing.

**Note:** people with MND may also experience problems with sleep resulting from reduced mobility, muscle cramps, swallowing problems and anxiety.<sup>5</sup>

## Monitoring symptoms

Respiratory function significantly predicts both survival and quality of life in people with MND.<sup>3</sup> Regular monitoring of respiratory function will assist health professionals and the person with MND to reach decisions about its management in a timely way.

It is also important because:

- it can help to identify reversible causes for respiratory changes eg respiratory tract infections or secretion problems
- it may prevent an acute respiratory emergency, or inappropriate action being taken in the event of an emergency
- it will help to ensure that placement of a gastrostomy tube, if needed, is performed in a timely manner, as respiratory evaluation is needed prior to surgical procedures requiring sedation.<sup>6</sup>

Therefore, when monitoring someone with MND, it is important to:

- ask about symptoms of respiratory involvement
- look for signs of weakness in the trunk and respiratory muscles.

### Establishing a baseline of respiratory function

Tests should be performed to establish the baseline respiratory function, as part of the initial assessment to diagnose MND, or soon after diagnosis.<sup>4</sup>

Tests may include:

- pulse oximetry (SpO<sub>2</sub>) – to measure the level of oxygen in the blood (at rest and breathing room air)
- forced vital capacity (FVC) or vital capacity (VC) – to find out the volume of the lungs from a full breath in to a full breath out
- sniff nasal inspiratory pressure (SNIP) or maximal inspiratory pressure (MIP) – to test the strength of the muscles used to breathe in.

These tests should be carried out by a health professional who has the knowledge and experience to perform them, usually the respiratory consultant.

If the person with MND has severe bulbar impairment (weakness in the tongue, mouth, throat) or severe cognitive problems, it may be difficult to choose the right mask or mouthpiece to use for these tests. As a minimum, pulse oximetry should be carried out, though the evidence for using pulse oximetry alone is not strong.

### Frequency of respiratory function testing

Respiratory function tests should be performed every two to three months, depending on:

- whether there are any signs or symptoms of respiratory impairment
- the rate of progression of an individual's MND
- the person's preference and circumstances.

Because respiratory function testing monitors progression, it may be frightening for someone with MND. However, keeping the person with MND and the wider MDT informed can promote discussion of their future care options and potential intervention.<sup>4</sup>

Monitoring of respiratory signs and symptoms can be completed by any member of the team including the patient and carers, leading to early interventions if necessary.

### Assessing cough effectiveness<sup>4</sup>

The NICE guideline on MND (NG42) recommends assessing cough effectiveness using peak cough flow (PCF). A PCF of 160 litres per minute is required to cough effectively. A PCF below 270 litres per minute is the accepted threshold for someone to be taught cough augmentation techniques, as their PCF would be likely to fall lower than 160 litres per minute if they were to get a respiratory infection.

### Referral to the specialist respiratory team

Even mild respiratory symptoms or signs should highlight the need for early referral for a respiratory assessment.<sup>4</sup> The specialist respiratory team may include a respiratory consultant, respiratory physiotherapist and specialist nurses.

Referral should also be made to the specialist palliative care team, who can advise on psychological strategies, medication and advance care planning. The person with MND should also be referred to the specialist respiratory team to perform an arterial or capillary blood gas analysis if the person with MND has percutaneous oxygen saturation (SpO<sub>2</sub>) equal to, or less than 94% (or 92% if they have a known lung disease).<sup>4</sup>

Urgent referral to a specialist respiratory service (to be seen within one week) is indicated when the arterial partial pressure of carbon dioxide (PaCO<sub>2</sub>, measured from a blood sample) is greater than 6 kPa. Explain the reasons for and implications of the urgent referral to the person and (if the person agrees) their family and carers, with support from the respiratory team.<sup>4</sup>

Referral to a specialist respiratory service for continuous nocturnal (overnight) oximetry and/or a limited sleep study is also indicated when:<sup>4</sup>

- a person has sleep-related respiratory symptoms, despite the SpO<sub>2</sub> being within normal limits
- they have symptoms or signs of respiratory impairment, particularly orthopnoea, despite the PaCO<sub>2</sub> equal to or less than 6 kPa
- they have symptoms or signs of respiratory impairment despite a normal overnight pulse oximetry.

No one test is best for monitoring or indicating the ideal time to start NIV. The NICE guideline suggests that discussion with the person with MND and, if appropriate, their main carer/family around respiratory impairment, treatment options and possible referral to the respiratory ventilation service if any of the following results are obtained:

- FVC or VC less than 50% of predicted value
- FVC or VC less than 80% of predicted value plus any symptoms or signs of respiratory impairment, particularly orthopnoea
- SNIP or MIP less than 40cmH<sub>2</sub>O

- SNIP or MIP less than 65cmH<sub>2</sub>O for men or 55cmH<sub>2</sub>O for women plus any symptoms or signs of respiratory impairment, particularly orthopnoea
- repeated regular tests show a rate of decrease of SNIP or MIP of more than 10 cm H<sub>2</sub>O per three months.<sup>4</sup>

For further guidance on assessment, please refer to the NICE guideline on MND ([www.nice.org.uk/ng42](http://www.nice.org.uk/ng42)).

### **Involvement of other professionals**

A range of health and social care professionals may be involved in the care of the person with MND, and can offer additional support to manage respiration:

- district nurse – can provide an adjustable bed
- occupational therapists – can provide adjustable chair, postural support, relaxation techniques
- speech and language therapist – support with ways of controlling breathing while eating
- physiotherapist – support with breath stacking, assistance to cough, clearing secretions
- GP, palliative care team or neurologist for symptom management including anxiety, saliva etc
- complementary therapist – for relaxation.

The multidisciplinary team should be led by a healthcare professional with a specific interest in MND. The leader should ensure the person with MND's multidisciplinary care plan is coordinated and communicated to all relevant professionals, including the GP and primary care team, services providing urgent and emergency care, the person themselves and, where appropriate, their carer/family.

## **Strategies for managing respiratory signs and symptoms**

### **Positioning**

It is important to find positions that make it easier for the person with MND to breathe. The ideal position is usually upright or slightly reclined, with the arms supported, rather than lying down.<sup>7</sup>

A semi-reclined position may be achieved by using a riser-recliner chair with good arm support, or a wheelchair with a tilt-in-space mechanism. This position allows the diaphragm to move more freely and efficiently. In bed, it may help to raise the head, perhaps with more pillows or a profiling bed.

### **Relaxation and anxiety management**

Massage and aromatherapy massage may be considered for people with MND who are anxious, short of breath or in the terminal stages of life. Relaxation techniques may be helpful for the relief of anxiety.<sup>8</sup>

### **Use of a fan or open window**

Some people with MND find that movement of air around the face sometimes helps when they feel breathless.

### **Use of a walking frame**

Some people with MND have found the use of a three or four wheeled walking frame helpful. Depending on the level of use retained in the arms and shoulder girdle, the frame can help stabilise posture and balance. With the person's weight supported by the aid, this helps them to make better use of the muscles used to breathe.

## Fatigue management

The aim is for the person to conserve energy so they can do what is important for them. An occupational therapist can advise on adjusting activities to reduce the amount of energy needed to perform them.

### Information to share with people with or affected by MND:

Information sheet 11C – *Equipment and wheelchairs*

Information sheet 11D – *Managing fatigue*

### Information for professionals:

*Occupational therapy for MND*

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## Breathing techniques

A respiratory physiotherapist can give advice on exercises to maximise lung capacity, muscle elasticity and protect against partial lung collapse and infection.<sup>9</sup>

## Cough management

Difficulty in coughing can be due to weakness in inspiratory, expiratory and bulbar muscles. A respiratory physiotherapist can prescribe the most appropriate cough management regime for the person with MND and teach techniques to help them to cough more effectively. See page 7 for more information about cough management.

## Saliva management

Many people with MND experience problems with saliva, which can be difficult to control. All management options should be explored, as it is often a case of trial and error to find the right solution. The NICE guideline recommends a range of treatment options, including medications.

A weak cough may make it difficult to clear respiratory secretions, which can become thick and tenacious. Humidification and nebulisers should be considered. Drinking pineapple or papaya juice may also help. These juices contain proteolytic enzymes, bromelain (pineapple) and papain (papaya), which help break down protein in mucus. The juices can also be applied gently on a sponge as part of a mouth care regimen. Bromelain and papain enzymes are also available as tablets.

### Further information for you:

Information sheet P3 – *Managing saliva*

Information sheet P9 – *Oral suction*

### Information to share with people with or affected by MND:

Information sheet 7A – *Swallowing difficulties*

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## Avoidance of infections

The specialist respiratory team can advise on strategies to avoid infections. People with MND should be offered prophylactic pneumococcal and influenza vaccinations. There may be a place for early use of antibiotics,<sup>13</sup> if this is what the person with MND wants.

Repeated aspiration and poor lung function can leave a person prone to chest infections and pneumonia. It is important to have contact with a speech and language therapist who can assess and advise about swallowing ability.

Poor mouth care can also lead to an increased risk of respiratory infections, so it is important to maintain oral hygiene.

**Information for professionals:**

*Motor neurone disease for dental professionals*

**Information to share with people with or affected by MND:**

Information sheet 11B – *Mouth care*

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**MND Just in Case kit**

The MND Just in Case kit is designed to hold medication for the relief of choking, breathlessness or related anxiety. 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 medication and storage for the prescribed medications. The MND Just in Case kit should be ordered from MND Connect by a GP for a named patient.

**Oxygen**

Oxygen should be used with caution in the presence of respiratory muscle weakness, because this weakness can lead to the retention of carbon dioxide (hypercapnia) rather than hypoxia (inadequate levels of oxygen in the body).<sup>9</sup>

In this situation, NIV may be the most appropriate treatment (see NICE guideline NG42)<sup>4</sup>, as oxygen therapy can lead to further respiratory depression. However, when oxygen levels are low, oxygen may sometimes be used with caution – preferably under guidance of the person's specialist team.

**Fitness to fly assessments**

Airlines and travel insurance companies may request that patients with MND have a fitness to fly assessment to ensure safety during air travel. These can be arranged at respiratory departments and involve breathing a gas mixture simulating cabin air and determining if the resulting blood oxygen level is adequate and safe, or if supplementary oxygen is required during the flight.

**Information to share with people with or affected by MND:**

Information sheet 8E – *Air travel and ventilation for motor neurone disease*

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## Cough management

Difficulty in coughing can be due to weakness in inspiratory, expiratory and bulbar muscles. A respiratory physiotherapist can prescribe the most appropriate cough management regime for the person with MND and teach techniques to help them to cough more effectively.

The NICE guideline on MND recommends:<sup>4</sup>

- unassisted breath stacking and/or manual assisted cough as first-line treatment. Breath stacking involves adding additional air to that already in the lungs to add force to a cough
- assisted breath stacking (eg using a lung volume recruitment (LVR) bag with a one-way valve) for those with bulbar dysfunction or whose cough is ineffective with unassisted breath stacking
- if available, use of a mechanical insufflation:exsufflation (MI:E) machine, if assisted breath stacking is not effective and/or during a respiratory tract infection.

## Manual assisted cough

This technique works by creating increased pressure beneath the diaphragm. This mimics the movement of the diaphragm using the hands or a towel. This is usually performed by an assistant, but some people can learn to perform the technique on themselves. A respiratory physiotherapist can advise on the best technique for an individual and train their carer to assist them.

## Mechanical insufflation:exsufflation (MI:E) machine

MI:E is often referred to as Cough Assist. In the UK, there are two main MI:E devices used: the NIPPY Clearway (made by B&D Electromedical) and the E70 (made by Philips Respironics). Some UK centres may also still use the Emerson Cough Assist device (also made by Philips Respironics).

An MI:E device clears secretions by gradually applying a positive pressure to the airway, then quickly shifting to negative pressure. This rapid change in pressure simulates a natural cough. The device supports and enhances the ability of weakened inspiratory and expiratory muscles in order to aid secretion clearance.

Use of MI:E has been shown to reduce the incidence of chest infections and the likelihood of emergency hospital admissions.<sup>10</sup> However, the benefits may be limited for someone who has severe bulbar impairment.<sup>9</sup>

MI:E may be started once a person has experienced repeated respiratory infections.<sup>11</sup> Once MI:E has been set up, people are advised to use the device on a daily basis to get maximum benefit and to prevent the onset of further respiratory infections. MI:E is used with either a tight-fitting face mask, a mouthpiece, tracheostomy or endotracheal tube.

MI:E can be used in different modes to improve synchronisation between the patient and the machine, improving patient comfort and therefore treatment effectiveness. A respiratory specialist will be able to prescribe the exact treatment regime needed. Full training should be provided to the person with MND and any family/carers who may operate the device.

The provision or loan of MI:E machines varies from region to region and sometimes servicing and maintenance can be problematic. If problems are experienced, contact the local MND Association regional care development adviser (RCDA – see page 16) who may be able to help.

## Breath stacking

Breath stacking is a technique to help maintain the size of breath a person with MND can take, and also helps to clear secretions. The technique involves taking additional breaths on top of each other without exhaling. Breath stacking opens the small air sacs in the lungs, and help to keep the chest wall flexible. It may also help to improve voice strength.<sup>25</sup>

Depending on the progression of respiratory symptoms, a lung volume recruitment (LVR) bag may be used to increase the volume of air inhaled. This is known as assisted breath stacking. This technique should only be used once the patient has been trained by a physiotherapist or MND clinical specialist. They will carry out an assessment to make sure the person is medically fit to use LVR bag, and may ask for advice from their GP, consultant respiratory physician or specialist respiratory physiotherapist.

Some people can use the LVR bag unassisted, and some need help to squeeze the bag or hold on the mask or mouthpiece. LVR bags are inexpensive, and may be more easily obtained than an MI:E, which costs a great deal more.

# Assisted ventilation

## Non-invasive ventilation (NIV)

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



A trial of NIV can be offered if the person's symptoms and the results of respiratory function tests indicate they are likely to benefit from the treatment. It is important that the benefits and limitations of the intervention are discussed with the person with MND (and, where appropriate, their carer/family).

They should:

- understand what NIV is and what it can achieve,
- be given advice about how and when to use it
- recognise the need for regular review
- have information about all possible treatment and management options
- understand possible problems using ventilation with other equipment, such as older models of eye gaze systems.

For someone who has severe bulbar impairment, NIV may have limited benefits.<sup>3</sup> If a person has severe bulbar impairment or cognitive problems that may be related to respiratory impairment, a trial of NIV may only be considered if they may benefit from an improvement in sleep-related symptoms, such as quality of life during the day due to sleepiness, or correction of hypoventilation.<sup>4</sup>

### **Before starting NIV**

The following issues should be discussed with the person with MND (and, where appropriate, their carer/family):

- that NIV won't stop the progression of MND
- the most appropriate interface (mask) based on the person's needs, lifestyle factors and safety (experimentation may be needed)
- the long-term support offered, with arrangements in place for device maintenance and 24-hour emergency clinical and technical support
- how and when respiratory function will be monitored
- the person's tolerance of the treatment
- the risk and possible consequences of ventilator failure
- power supply, including battery back-up
- how easily the person can get to hospital
- risks associated with travelling away from home (especially abroad)
- use of a humidifier
- issues relating to secretion management
- availability of carers
- the right to choose to have NIV withdrawn at any time.

### **Training and support**

Ongoing emotional and psychological support should be provided for the person, their family and carers, along with training on, for example:

- using NIV and interfaces
- emergency procedures
- using the equipment with a wheelchair or other mobility aids
- secretion management
- palliative care support

- therapy to assist coughing.<sup>4</sup>

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

### **Involvement of carer/family**

The involvement of the main carer/family is usually vital. It would only be in exceptional circumstances (eg if the person with MND does not consent to the main carer/family being involved) where this would not happen, as most management strategies will impact upon the carer/family. This is particularly relevant if the person's hand function is compromised and they would be unable to manage the NIV machine or adjust the mask independently.

If the person decides to use ventilation, the main carer/family should be assessed, including:

- their ability and willingness to assist in providing NIV, including giving support during the night
- their training needs
- the opportunity to discuss any concerns they may have.<sup>4</sup>

### **Invasive ventilation**

Very occasionally, a person with MND will be fully ventilated via a tracheostomy due to an acute episode of respiratory insufficiency. This can happen as an initial presentation of the disease, with a diagnosis of MND being made in intensive care or other high dependency settings.<sup>3</sup>

There can be difficulties weaning the person from the ventilator long-term. Some units will be reluctant to offer a tracheostomy with full ventilation electively.<sup>3</sup> This is because of the impact on the family and the need to provide specialist care in the future, which may not be possible to deliver at home. Great thought must be put into supported discussions with the person with MND (and, where appropriate, their carer/family).

### **Ventilation in an acute situation**

Acute breathlessness is usually caused by a specific event such as pneumonia, lower respiratory tract infection or sputum retention, against a background of previously undetected respiratory muscle weakness.<sup>3</sup> Once the acute event has been treated, consideration can be given to long term care, which may include assisted ventilation.

Ideally, discussions around ventilation and use of NIV will have taken place at an early stage between the person with MND and respiratory specialist and recorded on an advance care plan.<sup>4</sup>

Where possible, a respiratory specialist should explore options for assisted ventilation with the person with MND before action is taken. In an acute situation where this is not possible, an appropriate professional should make the decision on whether to introduce NIV, then seek advice about long-term management.

If someone presents in hospital with rapid onset ventilatory failure, the NICE guideline on MND recommends urgent introduction of NIV. Unplanned intubation or tracheostomy may be carried out if necessary.

### **Advance care planning**

Regular opportunities should be given to the person with MND (and, where appropriate, their carer/family) to discuss, in a sensitive way, decisions for end of life. This should take place whether or not someone decides to have assisted ventilation. If someone is considering assisted ventilation, discussions need to take place in relation to continuing or withdrawing this intervention (see below).

The progressive nature of MND means the person will continue to deteriorate physically, despite ventilatory support.<sup>3</sup> The treatment may be started to help with night-time symptoms but this can, over time, lead to 24-hour dependence.<sup>3</sup> The person with MND may have strong views on this.

Discussions should include:

- the amount of support that will be required, including during the night
- planning end of life care
- considering whether to record an Advance Decision to Refuse Treatment (ADRT) in certain circumstances in the future (see below)
- considering what to do if NIV fails because of either
  - (a) an acute, but potentially reversible, deterioration in health, or
  - (b) irreversible disease progression
- strategies to withdraw assisted ventilation if the person with MND wishes.

People may find early discussions about these issues difficult and some may disengage completely. It is the health professional's role to show willingness to discuss these topics, provide guidance and make referrals to the appropriate services. If discussions have not taken place at that point, for whatever reason, they should be instigated by an appropriate health professional as the person with MND becomes more dependent on ventilator support.

### **Withdrawal of assisted 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.<sup>4</sup> 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 has become a burden. They may suddenly feel claustrophobic from wearing the mask and decline ventilation when previously it has been accepted. Alternatively, there may come a time when respiratory muscles fail to such an extent that ventilation is insufficient to alleviate symptoms or to correct blood gas abnormalities.<sup>3</sup> 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.<sup>4</sup>

Withdrawal of assisted ventilation should be carefully planned so that symptoms can be managed to avoid distress or discomfort. 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.<sup>4</sup>

If a person with MND wishes to stop assisted ventilation, they should have support from professionals with knowledge and experience 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.<sup>4</sup>

It has been recognised that withdrawal of ventilation can be very challenging for professionals and the Association for Palliative Medicine has produced guidance.<sup>14</sup> Support should be available for healthcare professionals who may be involved in the process of withdrawing NIV.<sup>4</sup> Many hospice doctors have experience in managing the process and may be called upon if needed for support and information.

#### **Information to share with people with or affected by MND:**

Information sheet 8C - *Withdrawal of ventilation*  
*End of life guide*

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#### **Information for professionals**

*Withdrawal of Assisted Ventilation at the Request of a Patient with Motor Neurone Disease* - Association of Palliative Medicine available from <http://apmonline.org/publications>

## Palliation of symptoms

Not everyone will choose to have assisted ventilation, or can tolerate this intervention. As part of palliative care, symptoms such as breathlessness will need to be managed. They may be needed whether or not someone is using NIV, and will be necessary if the person with MND requests withdrawal of ventilation.<sup>3,4</sup>

Increasing doses of medication and increased levels of sedation may be required to control symptoms at the end of life.<sup>15</sup>

Medications used in palliation of symptoms include:

- antimuscarinics, such as hyoscine hydrobromide and glycopyrrolate, to reduce saliva and respiratory secretions<sup>4,16</sup>
- medications to manage breathlessness exacerbated by anxiety, and other medications to reduce anxiety and terminal restlessness, such as midazolam, haloperidol or levomepromazine.<sup>4,15,17</sup>
- opioid analgesics, such as morphine sulphate or diamorphine, to reduce cough reflex, relieve dyspnoea and the feeling of effortful breathing, and therefore fear and anxiety. They can also control pain<sup>18</sup>
- anti-emetics, such as levomepromazine or cyclizine, for nausea<sup>16</sup>

If dysphagia prevents the use of oral medication, parenteral medication may be given as a continuous subcutaneous infusion using a syringe pump.<sup>19</sup>

For drug dosages, refer to British National Formulary or Palliative Care Formulary.

## Information, decision-making and patient choice

Effective management of the respiratory symptoms of MND requires some understanding of the person's wishes for care, including end of life care. Clear explanations of all procedures and the rationale behind them should be given to the person with MND (and, where appropriate, their carer/family). The person with MND should be given sufficient information to give informed consent about any proposed interventions.

Discussions should take place at an appropriate time and in a sensitive manner, for example:

- soon after MND is first diagnosed
- when monitoring respiratory function
- when respiratory function deteriorates
- if the person with MND asks for information.

The ability of the health professional to assess the willingness and capacity of the person with MND to absorb information at any particular stage of the disease is absolutely crucial.

The topics that are likely to require discussion at some point are:

- possible symptoms and signs of respiratory impairment
- a description of all the different types of respiratory function tests, how and when these are done and why
- the different interventions available (NIV, alternative palliative strategies), including the advantages and disadvantages of each
- how respiratory function will change as the disease progresses and decisions that may need to be made about changes in management towards end of life (including increasing dependence on NIV and withdrawal of NIV)
- any changes to the person's viewpoint around future care and interventions. Any decisions that have been made and recorded should be reviewed at regular intervals.

Key decisions reached with the person with MND (and, where appropriate, their family and carers) should be recorded in the person's emergency care summary and relayed to all other relevant health and social care professionals, including services that may see the person in a crisis situation, such as the GP and services providing emergency and urgent care.<sup>4</sup>

### **Cognition/capacity**

Research suggests that up to half of all people with MND may experience a degree of cognitive impairment. Up to 15% of these will develop frontotemporal dementia.<sup>20, 21, 22, 23</sup>

For people with limited cognitive function or mental capacity, careful consideration needs to be given to the appropriate management of any respiratory impairment. The individual's ability to weigh up treatment options and give informed consent will be important.

The key points to consider are:

- the person's capacity to make decisions and to give consent to any treatment or intervention
- the impact on their family and carers
- the severity of cognitive problems
- whether the person is likely to understand and tolerate respiratory function tests and their willingness to undertake them
- whether the person is willing to accept NIV, if indicated
- whether the person is likely to achieve improvements in sleep or behavioural symptoms.

In these situations, the involvement of the next of kin, key family members, an advocate or a person who has been given Lasting Power of Attorney (relevant in England and Wales) is clearly indicated. It is also important to establish whether an Advance Decision to Refuse Treatment has been recorded.

#### **Further information for you:**

*Cognitive change, frontotemporal dementia and MND*

#### **Information to share with people with or affected by MND:**

Information sheet 9A – Will the way I think be affected?

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

Download from [www.mndassociation.org/publications](http://www.mndassociation.org/publications) or contact MND Connect to order hard copies. Call 0808 802 6262 or email [mndconnect@mndassociation.org](mailto:mndconnect@mndassociation.org)

### **Advance Decision to Refuse Treatment (ADRT)**

An ADRT allows people to make decisions to refuse treatments or have treatment withdrawn, including those treatments that sustain life.

When appropriately recorded as part of advance care planning, a decision by someone with MND to discontinue the use of ventilation is legally binding, and these wishes should be followed.

Advance care plans and ADRTs may be reviewed over time. It is important to check back with the person with MND whether their wishes have changed, and that up-to-date copies are kept with patient records and with relevant professionals.

For information on ADRTs, contact your local palliative care service.

#### **Information to share with people with or affected by MND:**

Information sheet 14A – *Advance Decision to Refuse Treatment (ADRT)*

**See page 15 for order information.**

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## How the MND Association can support you and your team

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 Helpline

Our helpline offers help, information and support, and signposting to other services and agencies. The service is for people living with MND, carers, family members, health and social care professionals and Association staff and volunteers who directly support people with MND. Please contact MND Connect if you have any questions about the information in this publication. Phone 0808 802 6262 or email [mndconnect@mndassociation.org](mailto: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, accredited by the Information Standard. Downloads of 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 directly from the MND Connect team by phoning 0808 802 6262 or email [mndconnect@mndassociation.org](mailto:mndconnect@mndassociation.org)

### MND Association website

Access information for health and social care professionals on our website at [www.mndassociation.org/professionals](http://www.mndassociation.org/professionals)

### Education

Our education programme is designed to improve standards of care and quality of life for people living with and affected by MND. Opportunities include online modules and face-to-face training, such as conferences and masterclasses. We also have a bursary scheme to support health and social care professionals undertaking training relevant to MND. Find out more at [www.mndassociation.org/education](http://www.mndassociation.org/education)

### Transforming MND Care Audit

Our free, simple to use audit tool has been specifically designed for health and social care professionals working with people with MND. Its standards are based on the NICE guideline on MND. Visit [www.mndassociation.org/transformcare](http://www.mndassociation.org/transformcare) for more information and to register your team or organisation for the audit.

### 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. Referrals for support grants or equipment loan need to be made by a relevant health or social care professional. Contact MND Connect on 0808 802 6262 or email [mndconnect@mndassociation.org](mailto:mndconnect@mndassociation.org)

### Wheelchair service

If you are seeking information on wheelchairs, or if there are delays in assessment or provision of an appropriate wheelchair for someone with MND, our wheelchair service may be able to help, through training, joint assessments, advice or support. Call MND Connect on 0808 802 6262 or email [wheelchairs@mndassociation.org](mailto:wheelchairs@mndassociation.org)

### Communication aids service

This service helps people affected by MND and health and social care professionals with queries about communication aids. The service provides limited financial support for communication aids or some items on loan, if unavailable or delayed through health and social care services. Our aim is to improve provision and information on a local and national level, through collaboration with professionals. Call MND Connect on 0808 802 6262 or email [communicationaids@mndassociation.org](mailto:communicationaids@mndassociation.org)

### Research into MND

We fund and promote research that leads to new understanding and treatment and brings us closer to a cure. Visit [www.mndassociation.org/research](http://www.mndassociation.org/research) or contact the Research Development team on 01604 611880 or [research@mndassociation.org](mailto:research@mndassociation.org)

For the latest research news, visit our research blog at [www.mndresearch.wordpress.com](http://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 around the world. Subscribe for email alerts at [www.reccob.wordpress.com](http://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, attracting more than 800 delegates from at least 30 countries. Find out more at [www.mndassociation.org/symposium](http://www.mndassociation.org/symposium)

### **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](mailto:membership@mndassociation.org)

## **Local support**

### **Regional care development advisers (RCDAs)**

We have a network of regional care development advisers (RCDAs) covering England, Wales and Northern Ireland. RCDAs have specialist knowledge of 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 for health and social care professionals in MND, and are champions at influencing care services in their local areas.

### **MND care centres and networks**

Care centres and networks are teams of professionals who are specialists in MND. 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 provide one-to-one support to people affected by MND. Managed by our RCDAs, 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 local support groups. They can also provide a link with health and social care professionals. You may be in contact with Association visitors following up on queries from the families they support.



Please send us your feedback on  
**Evaluation and management of respiratory symptoms  
in motor neurone disease**

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/professionalinformation](http://www.surveymonkey.com/s/professionalinformation)

What is your profession or specialism?

---

Did you find this resource useful?

Yes    Somewhat    Not really    No   Please explain your answer

---

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

- |                                                                                 |                                                                |
|---------------------------------------------------------------------------------|----------------------------------------------------------------|
| <input type="checkbox"/> an increased understanding of their symptoms           | <input type="checkbox"/> more confidence                       |
| <input type="checkbox"/> an increased understanding of their condition          | <input type="checkbox"/> improved quality of life              |
| <input type="checkbox"/> more independence                                      | <input type="checkbox"/> a greater ability to maintain dignity |
| <input type="checkbox"/> an increased ability to raise awareness of their needs |                                                                |

Were there any particular topics that were useful to you?

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

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Are there any other MND-related topics that you'd like more information about?

---

Do you have any other feedback about this resource or our other information for professionals?

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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

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## MND Association

PO Box 246, Northampton NN1 2PR  
Tel: 01604 250505  
Email: [enquiries@mndassociation.org](mailto:enquiries@mndassociation.org)  
[www.mndassociation.org](http://www.mndassociation.org)

 @mndeducation

 /mndassociation

## 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.

## 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  
[mndconnect@mndassociation.org](mailto:mndconnect@mndassociation.org)