

Managing respiratory symptoms in motor neurone disease

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

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.³

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 – *Air travel and ventilation for motor neurone disease*

See page 18 for details of how to order our publications.

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.

It includes specific recommendations about respiratory function and symptoms, cough management and ventilation. This information sheet incorporates the recommendations outlined by NICE.

For more information, see 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*

See page 18 for details of how to order our publications.

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).

They should be given sufficient information to give informed consent about any proposed interventions. This can be more challenging if the person with MND is affected by cognitive change or frontotemporal dementia. See page 12.

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
- details of the different types of respiratory function tests
- the different interventions available (types of ventilation, 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 and withdrawal of ventilation)
- any changes to the person’s viewpoint around future care and interventions.

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.⁴

Signs and symptoms of 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:⁴

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

Additional symptoms not specifically described by the NICE guideline include:

- difficulty co-ordinating breathing with swallowing
- breathlessness when bending over
- reduced voice volume or being unable to complete long sentences
- avoidance of task secondary to breathlessness.

Symptoms during the night are often an early indicator of respiratory inefficiency.³ When lying down, breathing can become worse, leading to breathlessness, a fall in oxygen levels and a build up of carbon dioxide in the blood. As a result, the person with MND may:

- complain of waking with a headache, feeling unrefreshed or groggy, or as though they have not 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 night²²
- seem confused when they wake up.

The person's partner may have noticed laboured or shallow breathing patterns or long pauses between breaths (apnoea) during sleep, where the person may wake up gasping for air or periods where they stop breathing.

Monitoring symptoms

Respiratory function significantly predicts both survival and quality of life in people with MND.³ 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.⁶

Therefore, when monitoring someone with MND, it is important to ask about symptoms of respiratory involvement and 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.⁴ Tests should be carried out by a health professional who has the knowledge and experience to perform them.

Generally these tests are carried out by a respiratory physiologist, physiotherapist or nurse, and usually take place in the MND/neurology clinic.

Tests should include:

- pulse oximetry (SpO₂) – 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
- peak cough flow (PCF) - to assess cough effectiveness
- arterial or capillary blood gas analysis if the person's SpO₂ (measured at rest and breathing room air) is less than or equal to 92% if they have known lung disease **or** is less than or equal to 94% if they do not have lung disease.⁴

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. In this case a measurement of blood gas will ascertain if there is daytime respiratory failure (raised CO₂) or under-breathing at night, known as hypoventilation (normal CO₂ and raised bicarbonate).

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, and their preferences 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.⁴

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 effectiveness⁴

The NICE Guideline on MND (NG42) recommends assessing cough effectiveness using peak cough flow (PCF). If PCF is reducing or if the patient is reporting difficulty clearing secretions then cough augmentation techniques such as breath stacking might be sufficient.

Once the PCF falls below or around 270 litres per minute cough augmentation techniques are essential. PCF is likely to fall still further if the patient were to get a respiratory infection. These techniques may help prevent respiratory infection and will be more effective if the patient has learnt them in advance.

Referral to the specialist respiratory team⁴

The person should have an early referral for respiratory assessment. Some services are able to offer this at the point of diagnosis. Even mild respiratory symptoms or signs should highlight the need for early referral for a respiratory assessment.

The specialist respiratory team may include a respiratory consultant, respiratory physiotherapist and specialist nurses. Consider referral 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 peripheral oxygen saturation (SpO₂) equal to, or less than 94% (or 92% if they have a known lung disease), or if the person has symptoms of sleep disordered breathing (regardless of the SpO₂ measurement).

Urgent referral to a specialist respiratory service (to be seen within one week) is indicated when the arterial partial pressure of carbon dioxide (PaCO₂, measured from a blood sample) is greater than 6 kPa. A raised bicarbonate of above 27 mmols/l even in those with normal arterial CO₂ would also be an indication for urgent referral. This indicates nocturnal hypoventilation which could rapidly progress to daytime hypoventilation. 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.

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

- a person has sleep-related respiratory symptoms, despite the SpO₂ being within normal limits
- they have symptoms or signs of respiratory impairment, particularly orthopnoea, despite the PaCO₂ 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 40cmH₂O

- SNIP or MIP less than 65cmH₂O for men or 55cmH₂O 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 cmH₂O per three months.

Raised daytime CO₂ is a red flag and indicates NIV should be started as soon as possible if the patient wants it. For further guidance, see the NICE guideline on MND available at 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 – may be able to help with equipment provision such as mobility and independent living aids, medical equipment such as specialist beds and mattresses
- occupational therapist – can provide adjustable chair, postural support, relaxation techniques
- speech and language therapist – support with ways of controlling breathing while eating
- respiratory physiotherapist – support with breath stacking, assistance to cough, clearing secretions
- GP, palliative care team or neurologist - symptom management including anxiety, saliva etc
- complementary therapist – for relaxation.

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

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.⁷ 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, bed blocks 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.⁸

Use of a fan or open window

Some people find that movement of air around the face sometimes helps when they feel breathless. Hand held fans are particularly beneficial, if the patient is able to hold it.

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 strength of the arms and shoulder girdle, the frame can help stabilise posture and balance. With the person's weight supported by the aid, they can 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*
Personal care guide - includes information about fatigue management

Information for professionals:

Occupational therapy for MND

See page 18 for details of how to order our publications.

Breathing techniques

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

Cough management

Difficulty in coughing can be due to weakness in inspiratory, expiratory and bulbar muscles. If the person has a PCF of less than 270L/min, they may have an ineffective cough and therefore would benefit from cough augmentation.

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 7-8 for more information about cough management.

Saliva management

Many people with MND experience problems with saliva, which can be difficult to control. Difficulty controlling saliva can be distressing for patients and could increase the risk of chest infection, making effective saliva management essential. 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.

Treatment with mucolytic medication such as carbocisteine can help to thin secretions. An oral suction unit may be used to help with saliva and secretion management.¹³

Information for professionals:

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*

See page 18 for details of how to order our publications.

Avoidance of infections

The specialist respiratory team can advise on strategies to avoid infections. People with MND should be offered coronavirus, prophylactic pneumococcal and influenza vaccinations. There may be a place for early use of antibiotics¹⁴ 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.

Cough augmentation to help clear respiratory secretions can help avoid infections.

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

Information for professionals:

Information sheet P12 - *MND information for dental teams*

Information to share with people with or affected by MND:

Information sheet 11B – *Mouth care*

See page 18 for details of how to order our publications.

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.

Further information:

Information sheet P4 - *Just in case kit*

See page 18 for details of how to order our publications.

Oxygen⁹

Oxygen should be used with caution in patients with MND, as it can lead to an increase in carbon dioxide levels and respiratory depression. Oxygen should only be used if advised to do so by the person's specialist team or respiratory consultant with links to neurology, using arterial blood gas analysis.

In a patient with MND who is displaying signs of acute illness, oxygen should be given with caution keeping oxygen saturations between 88-92%

In acute situations, non-invasive ventilation (NIV) may be the most appropriate treatment (see NICE guideline NG42)⁴, as oxygen therapy can lead to further respiratory depression. See pages 9-11 for more information on NIV.

Air travel assessments

Airlines and travel insurance companies may request that patients with MND have an 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. These assessments are usually charged for and are not generally provided by the NHS.

Information to share with people with or affected by MND:

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

See page 18 for details of how to order our publications.

Cough management

A weak cough may make it difficult to clear respiratory secretions, which can become thick and tenacious. 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 regime. Bromelain and papain enzymes are also available as tablets. Artificial saliva can also be useful. Nebulisers can be helpful for people using NIV with thick secretions.

Difficulty in coughing can be due to weakness in inspiratory, expiratory and bulbar muscles. A respiratory specialist 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:⁴

- 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.

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 helps to keep the chest wall flexible. It may also help to improve voice strength.²³

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 an LVR bag, and may ask for advice from their respiratory specialist. 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, however their availability varies. If problems are experienced, contact our MND Connect helpline - email mndconnect@mndassociation.org or call 0808 802 6262.

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. The device supports and enhances the ability of weakened inspiratory and expiratory muscles in order to aid secretion clearance. 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 or augments a natural cough.

Use of MI:E has been shown to reduce the incidence of chest infections and the likelihood of emergency hospital admissions.¹⁰ However, the benefits may be limited for someone with severe bulbar impairment.⁹ Spasms in the larynx (laryngospasm) may obstruct the airflow. Individually customised settings for pressure and flow can improve and extend the use of MI-E.¹¹

MI:E may be started once a person has experienced repeated respiratory infections.¹² Once set up, people are advised to use the device twice daily to get maximum benefit and prevent further respiratory infections. MI:E is used with either a face mask, a mouthpiece, tracheostomy or endotracheal tube (usually only used in an acute/ICU setting).

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 or 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 our MND Connect helpline - email mndconnect@mndassociation.org or call 0808 802 6262.

Assisted ventilation

As respiratory symptoms progress, the person with MND may benefit from assisted ventilation. The two types of ventilation are:

Non-invasive ventilation (NIV): the ventilator provides extra air through a mask over the nose or mouth (see page 11).

Tracheostomy ventilation: the ventilator supports breathing via a tube inserted into the windpipe through the neck (see page 12).

Before starting ventilation, the following should be discussed with the person with MND, and if appropriate, their carer/family:

- what assisted ventilation is and what it can achieve
- that ventilation won't stop the progression of MND, and they may become more dependent on breathing support eg during the day as well as overnight
- the long-term support offered, with arrangements in place for device maintenance and emergency clinical and technical support
- possible problems using ventilation with other equipment, eg old eye gaze systems
- how and when respiratory function will be monitored and ventilation reviewed
- 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
- all possible treatment and management options, including the right to choose to stop treatments when they are no longer beneficial, including withdrawal of ventilation. See page 13.

Some people with MND may wish to refuse ventilation, or other treatments in advance. See *Advance care planning* on page 14.

Information to share with people with or affected by MND:

Our information sheet 8B – *Ventilation for MND* can support someone with MND to consider their options when making decisions about assisted ventilation.

See page 18 for details of how to order our publications.

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 the ventilator and interfaces
- emergency procedures
- using the equipment with a wheelchair or other mobility aids
- secretion management
- palliative care support
- therapy to assist coughing.⁴

Involvement of family and carers

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 need support to use assisted ventilation, eg unable to manage the machine or adjust the NIV mask independently.

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

- their ability and willingness to assist with ventilation, including giving support during the night
- their training needs, alongside the wider care team
- the opportunity to discuss any concerns they may have.⁴

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.

It is important to use the most appropriate interface (mask) based on the person's needs, lifestyle factors and safety, and trialling different masks may be needed.

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.

Potential benefits of NIV:

- it can be stopped or withdrawn at any time even when the device is being used to completely support breathing,
- it can ease symptoms caused by weakened breathing muscles, such as shortness of breath, disturbed sleep, daytime sleepiness or fatigue and early morning headaches
- it can increase survival time
- it is portable and can be used in many places, including at home and on the move, if powered by battery
- surgery is not needed for NIV
- although the person is likely to need more support over time, NIV care is generally quite easy to manage in the earlier stages of MND
- a range of available masks means the person can try out different options to establish what suits them best
- in some cases, using NIV can add volume to the person's voice if this has become weak.

For someone who has severe bulbar impairment, NIV may have limited benefits.³ 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.⁴

Potential issues with NIV:

- it can cause discomfort from air flow leaking around the mask and irritating the eyes
- pressure of the mask on the skin can cause sores

- wearing a mask can cause claustrophobia, which in some cases may lead to anxiety or panic
- it can cause a blocked nose, a runny nose or dry mouth
- the person may need support with eating or drinking while using the equipment, as the flow of air may cause an increased risk of aspiration
- it can cause the person to swallow more air than usual, which can make their stomach feel full and uncomfortable, meaning it may take more effort to breathe
- it can take a while to get used to the feel of the equipment and flow of air
- care needs can be complex while using NIV in the later stages of the disease
- the flow of air can disturb the rhythm of natural speech
- although NIV machines are generally quiet, the noise can take some getting used to and may keep the person awake at night at first, and anyone they share a room with.

Mouth piece ventilation

Some of the issues above can be alleviated with mouth piece ventilation (MPV). For people who are becoming more breathless in the daytime, MPV can be added in to their management plan, in addition NIV via a mask.

MPV involves the use of a straw or alternative interface which fits comfortably in the mouth for each breath. The ventilator is usually set at higher pressure, with a longer breath length, than the usual NIV settings. This ensures maximal inspiratory capacity is achieved and can help to reduce breathlessness. MPV can help to reduce pressure ulcers and also act as aid to increase compliance with NIV where patients are finding this difficult. As breathing worsens, and need for ventilation increases in the daytime, some patients use MPV all day to support their breathing adequately alongside NIV. NIV via a mask continues at night for patients using MPV by day.

Tracheostomy ventilation^{24, 25, 26}

Tracheostomy ventilation is where a tube is inserted into the person's windpipe through an opening in the neck. This is attached to a ventilator which is triggered by the person's breathing. As breathing muscles weaken, the ventilator can completely take over the person's breathing.

Tracheostomy ventilation is not common in MND²⁵ and is not routinely offered to those who wish to have the procedure electively.²⁴ Local guidelines and criteria differ, but the respiratory team is most likely to consider an elective tracheostomy if:

- NIV is ineffective
- the mouth and throat muscles are weak and there are a lot of secretions
- more extensive suctioning is needed during a serious chest infection
- if breathing becomes more difficult during a period of acute illness and non-invasive ventilation does not provide sufficient support
- there is difficulty protecting the upper airway due to severe bulbar weakness.

Although it is occasional, the most frequent cause for tracheostomy ventilation is acute episode of ventilatory failure. 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.³ There can be difficulties weaning the person from the ventilator long-term, and they may become dependent on it, particularly if they have severe bulbar weakness.

Some people may wish to refuse this treatment in advance, and making decisions about future care and recording their wishes should be encouraged before acute deterioration occurs. See Advance care planning on page 14.

Potential benefits of tracheostomy ventilation:

- it can improve symptoms caused by weakened breathing muscles, such as shortness of breath, disturbed sleep, daytime sleepiness or fatigue and early morning headaches
- it may increase survival time
- it can be used where NIV cannot successfully support the person's breathing
- depending on the type of tracheostomy, it can help to protect the airway from aspiration and reduce the risk of lung infections
- it can be used to help with control of secretions
- a mask is not used, so mask related issues, such as claustrophobia, can be avoided.

Potential issues with tracheostomy ventilation:

Great thought must be put into supported discussions with the person with MND (and, where appropriate, their carer/family). It is important to explain:

- the impact on the family and the need to provide specialist care in the future, which may not be possible to deliver at home
- the need for carers in the home
- the lengthy stay in hospital following surgery
- care packages can take a long time to arrange, as staff need to be appropriately trained
- complete loss of voice or more difficulty speaking
- it can increase secretions, which will require regular suction.

Those services who do not offer elective tracheostomies to people with MND should be prepared for requests for the procedure, and handle these conversations with great sensitivity. Being refused may have a strong emotional impact on the person with MND and their family, as they may perceive that tracheostomy will increase their lifespan compared to NIV. Current research to support this is not strong, and no controlled trial has yet taken place to establish objectively whether tracheostomy directly extends life in MND compared to NIV.²⁴

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.³ 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.⁴

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, and then seek advice about longer 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, and if the person has not refused this treatment in advance. See page 14.

Cognition and mental 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.²¹

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 interfere with the equipment eg pulling off mask, changing settings
- 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 Advanced Decision to Refuse Treatment (ADRT) has been recorded. See next heading.

Information for professionals

Cognitive change, frontotemporal dementia and MND

Information to share with people with or affected by MND:

Changes to thinking and behaviour with MND

See page 18 for details of how to order our publications.

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.

The progressive nature of MND means the person will continue to deteriorate physically, despite ventilatory support.³ The treatment may be started to help with night-time symptoms but this can, over time, lead to 24-hour dependence.³ 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) regarding use of ventilation (see below)
- what to do if NIV fails because of either an acute, but potentially reversible, deterioration in health, or 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.

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 more 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)*

End of life guide

See page 18 for details of how to order our publications.

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 dependent on ventilation.⁴ Someone may have 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 level of disability or breathing support has become a burden or respiratory muscles fail to such an extent that ventilation no longer alleviates symptoms or corrects blood gas abnormalities.³ In such situations, a discussion may take place with the person with MND and their carer/family about whether it is appropriate to continue using ventilation.⁴

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 will be given to relieve anxiety and distress.⁴

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.⁴

It has been recognised that withdrawal of ventilation can be very challenging for professionals and the Association for Palliative Medicine has produced guidance.¹⁵ Support should be available for healthcare professionals who may be involved in the process of withdrawing NIV.⁴ 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

Information for professionals

A professional's guide to end of life care in MND

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

See page 18 for details of how to order our 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.^{3,4}

Increasing doses of medication and increased levels of sedation may be required to control symptoms at the end of life.¹⁶ If dysphagia prevents the use of oral medication, parenteral medication may be given as a continuous subcutaneous infusion using a syringe pump.²⁰

Medications used in palliation of symptoms include:

- antimuscarinics, such as hyoscine hydrobromide and glycopyrrolate, to reduce saliva and respiratory secretions^{4,17}
- medications to manage breathlessness exacerbated by anxiety, and other medications to reduce anxiety and terminal restlessness, such as midazolam, haloperidol or levomepromazine.^{4,16,18}
- 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 pain¹⁹
- anti-emetics, such as levomepromazine or cyclizine, for nausea.¹⁷

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

Information to share with people with or affected by MND:

End of life guide

Information for professionals

A professional's guide to end of life care in MND

See page 18 for details of how to order our publications.

MND Respiratory Pathway

The MND Respiratory Pathway below is not intended to be exhaustive, but to provide a quick reference to support professionals to ensure they have considered all areas of need. This pathway is interactive: if you're viewing online, click **find out more** to view further details.

Signs and symptoms

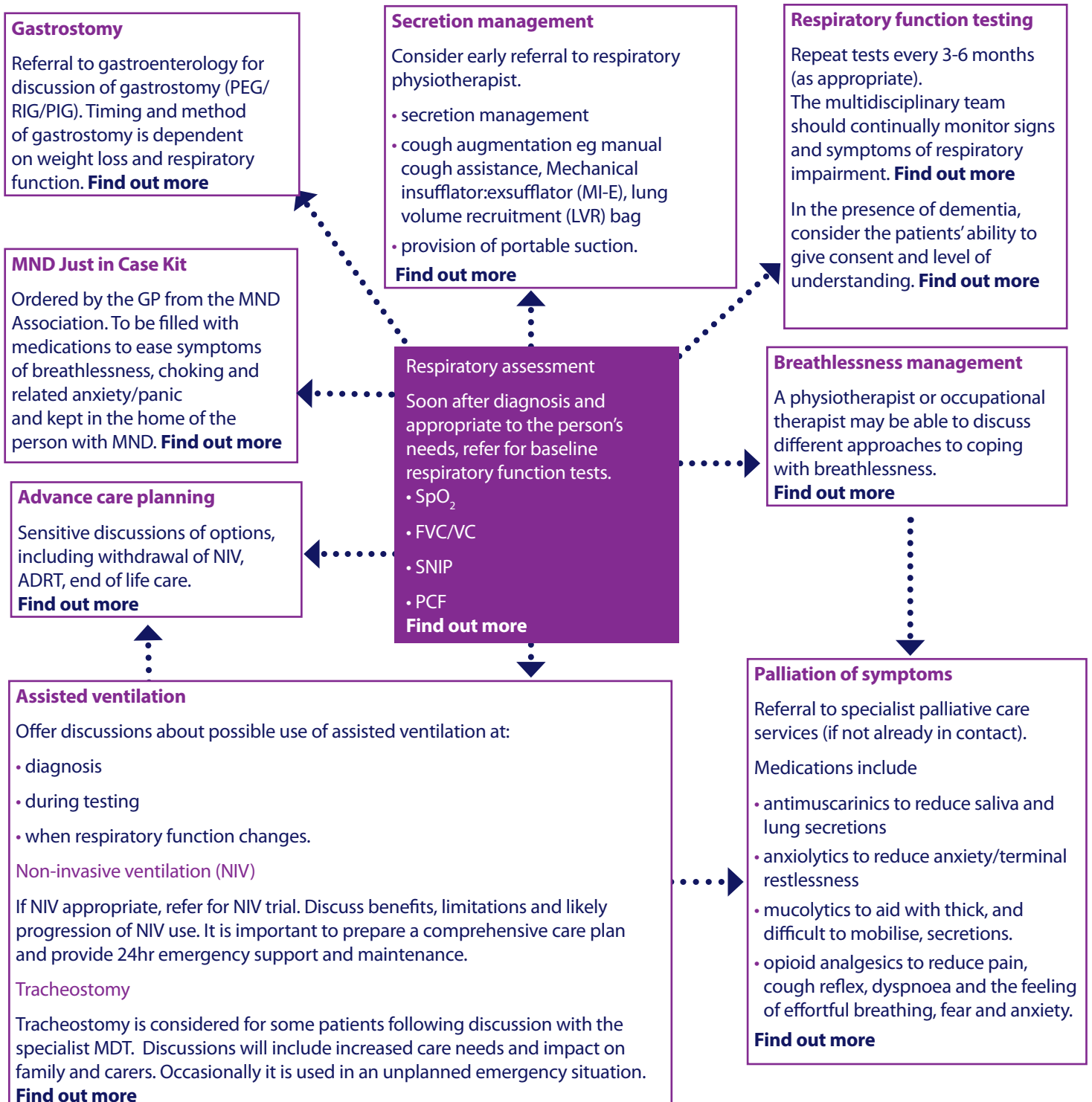
Signs and symptoms of respiratory impairment include (but are not limited to):

- early morning headaches
- daytime sleepiness
- orthopnoea
- repeat chest infections
- disturbed sleep.

Oxygen

Oxygen should be used with caution in those with MND.

Find out more



References

- 1 Bäumer D, Talbot K and Turner MR. *Advances in motor neurone disease*. Journal of the Royal Society of Medicine. 2014; 107:14.
- 2 Radunovic A et al. *Mechanical ventilation for amyotrophic lateral sclerosis/motor neuron disease*. Cochrane Database of Systematic Reviews 2013.
- 3 Rafiq MK et al. *Respiratory management of motor neurone disease: a review of current practice and new developments*. Practical Neurology. 2012;12(3):166-76.
- 4 NICE guideline NG42. *Motor neurone disease: assessment and management*. 2016.
- 5 Lo Coco D and La Bella V. *Fatigue, sleep, and nocturnal complaints in patients with amyotrophic lateral sclerosis*. European journal of neurology. 2012 May;19(5):760-3.
- 6 Greenwood DI. *Nutrition management of amyotrophic lateral sclerosis*. Nutrition in Clinical Practice. 2013;28(3):392-9.
- 7 Rolfe J. *Postural management in motor neurone disease: a guide to help maximise comfort and function in sitting and lying*. Oxford MND Care and Research Centre: 2013.
- 8 Nyatanga B et al. *A prospective research study to investigate the impact of complementary therapies on patient well-being in palliative care*. Complementary therapies in clinical practice. 2018 May 1;31:118-25.
- 9 Troosters T and Bott J. *Respiratory physiotherapy*. ERS Handbook of Respiratory Medicine. 2019 Sep 1:232.
- 10 Mahede T et al. *Use of mechanical airway clearance devices in the home by people with neuromuscular disorders: effects on health service use and lifestyle benefits*. Orphanet journal of rare diseases. 2015 Dec;10(1):1-7.
- 11 Andersen T et al. *Laryngeal response patterns influence the efficacy of mechanical assisted cough in amyotrophic lateral sclerosis*. Thorax 2017;72:221-229.
- 12 Morrow B et al. *Mechanical insufflation-exsufflation for people with neuromuscular disorders*. Cochrane Database of Systematic Reviews 2013, Issue 12.
- 13 Young CA et al. *Treatment for sialorrhoea (excessive saliva) in people with motor neuron disease/amyotrophic lateral sclerosis*. Cochrane Database of Systematic Reviews 2011; (5).
- 14 NICE Clinical Guideline 69. *Prescribing of antibiotics for self-limiting respiratory tract infections in adults and children in primary care*. 2008.
- 15 Association for Palliative Medicine of Great Britain and Ireland. *Withdrawal of Assisted Ventilation at the Request of a Patient with Motor Neurone Disease: Guidance for Professionals*. November 2015.
- 16 Palliative Care Formulary. <https://about.medicinescomplete.com/publication/palliative-care-formulary> Accessed February 2021
- 17 British National Formulary. *Prescribing in palliative care*. <https://bnf.nice.org.uk/guidance/prescribing-in-palliative-care.html> Accessed February 2021.
- 18 British National Formulary. *Hypnotics and anxiolytics*. <https://bnf.nice.org.uk/treatment-summary/hypnotics-and-anxiolytics.html> Accessed February 2021.
- 19 Brettschneider J et al. *Drug therapy for pain in amyotrophic lateral sclerosis or motor neuron disease*. Cochrane Database of Systematic Reviews 2013.
- 20 British National Formulary. <https://bnf.nice.org.uk> Accessed February 2021.
- 21 Goldstein LH, Abrahams S. *Changes in cognition and behaviour in amyotrophic lateral sclerosis: nature of impairment and implications for assessment*. Lancet Neurol. 2013;12:368-80.
- 22 Dani H, Esdaille A, Weiss JP. *Nocturia: aetiology and treatment in adults*. Nature reviews Urology. 2016 Oct;13(10):573.
- 23 Nerurkar, NK et al. *Endocrine and neurological disorders*. Textbook of Laryngology, p288. 2017
- 24 Turner MR et al. *Tracheostomy in motor neurone disease*. Practical neurology. 2019 Dec 1;19(6):467-75.
- 25 Palmer J et al. *Tracheostomy ventilation in motor neurone disease: a snapshot of UK practice*. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration. 2022 Jan 2;23(1-2):35-41.
- 26 Everitt E. *Tracheostomy 3: care of patients with permanent tracheostomy*. Nursing Times. 2016;112:21-3.

Acknowledgements

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

Rebecca Hall, NIV/ Respiratory Physiotherapist, University Hospitals Bristol NHS Foundation Trust

Charlotte Hardaker, Physiotherapist, Home Ventilation Team, Bristol Royal Infirmary

Jonathan Palmer, Nurse Consultant in Domiciliary Ventilation, Plymouth Hospitals Trust

Thank you to the following people for their contributions to earlier versions of this information:

Alison Armstrong, Nurse Consultant, North-East Assisted Ventilation Service, Newcastle upon Tyne Hospitals NHS Foundation Trust

Dr William Kinnear, Consultant Respiratory Physician, Nottingham University Hospitals NHS Trust

Stephanie Mansell, Consultant Respiratory Physiotherapist, Royal Free London NHS Foundation Trust

Dr Naveed Mustfa, Consultant Respiratory Physician, University Hospital of North Staffordshire

Joanna Smith, Ventilation Specialist Physiotherapist, University Hospitals of Leicester

Ema Swingwood, Respiratory Pathway Lead/Physiotherapist, University Hospitals Bristol NHS Foundation Trust

Dianne Fraser, Clinical Specialist for MND, Aberdeen Royal Infirmary

How to order publications

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

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

How we can support you and your team

MND Connect

Our helpline offers help, information and support, and signposting to people living with MND, carers, family and health and social care professionals.

Email: mndconnect@mndassociation.org

Phone: 0808 802 6262

Information resources

We produce high quality information resources for people living with MND, carers, family members and health and social care professionals.

www.mndassociation.org/publications

MND Association website

We have a wide range of information to support health and social care professionals working with people affected by MND.

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.

www.mndassociation.org/education

Support grants and equipment loan

Where statutory provision is not available, we may be able to offer a support grant or loan equipment.

www.mndassociation.org/getting-support

Research into MND

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

www.mndassociation.org/research

MND register

The MND Register of England, Wales and Northern Ireland aims to collect information about every person living with MND to help plan care and discover more about the cause of the disease.

www.mndregister.ac.uk

Regional staff

We have a network of regional staff with specialist knowledge of MND. They work closely with local statutory services and community care providers. Contact MND Connect for further information.

Email: mndconnect@mndassociation.org

Phone: 0808 802 6262

MND care centres and networks

We fund and develop care centres and networks across England, Wales, and Northern Ireland, which offer specialist multidisciplinary care for people with MND.

www.mndassociation.org/care-centres

Branches and groups

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

www.mndassociation.org/branchesandgroups

Association visitors (AVs)

AVs are trained volunteers who provide one-to-one local support to people affected by MND. They can support people affected by MND in person, by telephone or by email or through support groups.

www.mndassociation.org/associationvisitors

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 www.smartsurvey.co.uk/s/mndprofessionals or email your comments to infofeedback@mndassociation.org

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