

Breathing and motor neurone disease: medications and non-invasive ventilation

What you should know

- Respiratory muscle weakness usually develops gradually, but can occur suddenly, and make breathing difficult.
- Increasingly, many people with MND are choosing to use non-invasive breathing support, known as non-invasive ventilation (NIV).
- NIV is a type of assisted ventilation, where a machine boosts air flow into your lungs, normally through a mask held on by straps around the head
- Oxygen, as opposed to natural air flow, should be used with caution and appropriate monitoring because it does not usually help respiratory problems caused by MND.
- Medications can help reduce the symptoms of breathlessness and anxiety.
- The way you manage respiratory symptoms may affect your experience of other MND symptoms.

What you can do

Seek advice about respiratory management soon after your diagnosis with motor neurone disease, even if you have not noticed any changes in your breathing. This will give you more time to obtain information, have discussions and decide which strategies are right for you. If you think your respiratory muscles are becoming weaker, let your doctor or clinic know. Reversible causes of respiratory impairment such as a respiratory tract infection should be treated first¹.

Using medications

Oxygen

Oxygen does not usually help respiratory problems caused by motor neurone disease, but is sometimes prescribed if you have an underlying lung problem unrelated to motor neurone disease. Caution is required in the use of oxygen. Any decisions regarding oxygen for home use should be discussed with your neurologist, palliative care or respiratory physician. High levels of oxygen in the blood can affect the natural drive to breathe.

Opioids, benzodiazepines and other medications

There are a number of medications that help to reduce the symptoms of breathlessness. Small doses of morphine, or similar opioid medications, may be effective in reducing the sensation of breathlessness and help you to feel more comfortable¹. Benzodiazepines may be prescribed to reduce feelings of anxiety^{1,3}. Anticholinergics can address excess saliva¹.

Your neurologist, general practitioner, palliative care or respiratory physician can advise you about medications.

Using non-invasive ventilation

Increasingly, many people with motor neurone disease are choosing to use non-invasive breathing support, known as non-invasive ventilation (NIV). NIV can prolong and improve quality of life, and provide relief from symptoms of poor sleep, the build up of carbon dioxide in the body and shortness of breath^{1,2,3}. Surgery is not required. However, NIV does not prevent progressive weakening of the respiratory muscles¹.

Over time, NIV will be less effective in helping you to control your respiratory symptoms, because your motor neurone disease will continue to progress. While NIV is suitable for many people with the disease, it is not suitable for everyone. A respiratory physician is required to assess how suitable NIV is for you. Discussions regarding the use of NIV should also be held with family and others involved in your care for motor neurone disease³.

A range of available masks means you can try one that suits best. The mask is connected to a small pump that creates just the right pressure to keep your airways open so that room air can easily come in and out of your lungs when you breathe. People with motor neurone disease most commonly use variable positive airway

pressure (VPAP) or bi-level positive airway pressure (BIPAP) machines. This is because these types of NIV machines can provide a lower level of pressure when you breathe out. In addition, these machines can be adjusted to provide increased respiratory support if needed.

The NIV machine is usually used at night. As the respiratory muscles weaken, you might also use NIV at times during the day. You might take some time to get used to the machine and feel comfortable wearing the mask when you go to bed. Use of NIV may also lead to irritation of the eyes, sores on the skin, claustrophobia and other issues. Care needs can also be complex while using NIV in the later stages of the disease.

What about invasive ventilation?

Life-support, or invasive positive pressure ventilation, maintains a person's breathing when their respiratory muscles have failed. This type of ventilation requires a tracheostomy, which involves the insertion of a permanent tube into a person's windpipe through an incision in their neck. The tube is attached to a machine that supplies air for the lungs. A person on invasive ventilation requires constant, 24 hour care.

Invasive ventilation is not commonly used in Australia for a person with motor neurone disease² as their respiratory function will not recover and their disease will continue to progress.

Points to think about

The way you manage your respiratory symptoms may affect how your other symptoms of motor neurone disease can be managed. For example, if in the future you need liquid feed and fluids through a percutaneous endoscopic gastrostomy (PEG) tube, your respiratory function needs to be at or above a certain level to ensure safe insertion of the PEG tube (see below, *Living Better for Longer* fact sheets).

While strategies and treatments for respiratory management can reduce respiratory symptoms, they do not prevent progressive weakening of the respiratory muscles in a person with motor neurone disease.

Eventually, motor neurone disease affects the respiratory muscles so significantly that there is not enough respiratory muscle function to sustain life. Early advice will provide you with time to think about a respiratory and palliative care plan that:

- best meets your personal preferences
- provides you with comfort in the final stages of the disease.

Remember though, your preferences may change over time.

It can be difficult to discuss your thoughts and feelings about respiratory management with your doctor, family and others close to you, but it is only by talking to them that they will understand how you feel. It will also help them provide support for your preferences.

Some questions to ask

- What other options are available if NIV is not suitable for me?
- How do I get out and about with a NIV machine?
- How much does NIV cost?
- Who provides the machines and the masks?
- Does the government or my state MND Association provide assistance with cost?
- What happens if I am using NIV and there is a power failure?
- How will I be supported to use the NIV machine?
- What happens if I decide that I want to stop using NIV?
- How will my carer be trained and supported to supervise my use of NIV?

More information

For more information about respiratory management contact your general practitioner, neurologist, respiratory physician, palliative care team, MND clinic or service or your MND Association. The online MND decision tool can also help you learn more about your options for assisted ventilation: https://mnddecisiontools.com/public/3/decision_tool

References

1. National Institute for Health and Care Excellence (NICE) 2016, Motor neurone disease: assessment and management. Retrieved from <https://www.nice.org.uk/guidance/NG42/chapter/Recommendations#managing-symptoms>.
2. Berlowitz et al. 2016, 'Identifying who will benefit from non-invasive ventilation in amyotrophic lateral sclerosis/motor neurone disease in a clinical cohort', *J Neurol Neurosurg Psychiatry*, 87(3), 280-86.
3. Rafiq et al. 2012, 'Respiratory management of motor neurone disease: a review of current practice and new developments', *Pract Neurol* 12, 166-76.

Living Better for Longer: MND Australia evidence-based fact sheets

MND evidence-based interventions - an overview (EB1)

Multidisciplinary care (EB2)

Multidisciplinary care team (EB3)

Riluzole (EB4)

Breathing and motor neurone disease: an introduction (EB5)

Breathing and motor neurone disease: what you can do (EB6)

► **Breathing and motor neurone disease: medications and non-invasive ventilation (EB7)**

Considering gastrostomy - PEG and RIG (EB8)

To find out about motor neurone disease and other fact sheets in this series contact the MND Association in your state or territory
ph. 1800 777 175 or visit www.mndaustralia.org.au