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Living Better for Longer: MND Australia Fact Sheet EB7

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

What you should know

- Respiratory muscle weakness usually develops gradually, but can occur suddenly.
- Increasingly, many people with MND are choosing to use non-invasive breathing support, known as non-invasive ventilation (NIV).
- Oxygen does not usually help respiratory problems caused by MND.
- The way you manage your respiratory symptoms may affect how your other symptoms of MND can be managed.

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 get information, have discussions and decide which strategies are right for you.

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. 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.

Other medications

There are a number of medications that help to reduce the symptoms of breathlessness. Small doses of morphine, or similar medications, may be effective in reducing the sensation of breathlessness and help you to feel more comfortable. Anti-anxiety medication may be prescribed to reduce feelings of anxiety. 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 gives you breathing support without the need for the insertion of a tube into your windpipe. It provides relief from symptoms such as fatigue, breathlessness and disturbed sleep patterns, but does not prevent progressive weakening of the respiratory muscles (Andersen et al 2007).

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 motor neurone disease it is not suitable for everyone. The suitability of NIV for you will not be known until you have had a respiratory assessment.

NIV involves wearing a mask 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 but as the respiratory muscles weaken you might use it at times during the day as well. When using NIV for the first few times you may need to try several different types of masks to find the most comfortable one for you. Also, it can take some time to get used to the machine and to feel comfortable wearing the mask when you go to bed.

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 disease 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 greatly 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 therapies 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 the electricity fails?
- How will I be supported to use the NIV machine?

MND research in this area

Several research trials are underway overseas looking at the effectiveness of diaphragmatic pacing, which involves the electrical stimulation of a nerve in the diaphragm to 'pace' breathing. In 2011 the U.S. Food and Drug Administration (FDA) approved the NeuRx Diaphragm Pacing System (DPS)® for treating suitable ALS patients who have respiratory problems. Research is ongoing and at present this therapy is not used as standard treatment for motor neurone disease in Australia or other countries overseas.

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.

References

Andersen et al 2007, 'Good practice in the management of amyotrophic lateral sclerosis: clinical guidelines: an evidence-based review with good practice points. EALSC Working Group.', *Amyotroph Lateral Scler* 8(4), 195-213.

Kiernan MC (ed) 2007, *The motor neurone disease handbook*, MJA books, Australasian Medical Publishing Company, Sydney.

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)
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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.mndaust.asn.au

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MND Australia is indebted to the MND Association UK for permission to adapt their Information Sheet Understanding how MND might affect breathing.