

# Breathing under pressure – non-invasive ventilation for respiratory muscle weakness

People with neuromuscular disorders often require breathing assistance at some stage in their lives. For some, breathing problems develop slowly over months or years. For others, respiratory complications may occur suddenly following an apparently minor event such as a cold or chest infection. In both situations, the role weak respiratory muscles are playing in these problems needs to be recognised and appropriate care put into place in a timely fashion.

Inspiratory muscle weakness makes breathing in fully difficult. Over time this promotes underbreathing or *hypoventilation*. If this is severe enough or occurs over a long period of time it will affect the level of oxygen and carbon dioxide in the blood, causing *respiratory failure*. In addition, shallow breathing limits expansion of the chest wall creating stiffness of the rib cage which then makes taking a normal sized breath more difficult, promoting further underbreathing. The expiratory muscles also play a key role in maintaining lung health. Weakness of the abdominal muscles reduces the ability to build up pressure to create an effective cough. This can severely affect clearing secretions from the lungs leading to respiratory failure during a chest infection.

Abnormalities in breathing first appear during sleep. It is normal to reduce the depth of breathing slightly during sleep. For people with respiratory muscle weakness however, these normal changes in breathing and oxygen levels can become exaggerated, and significant underbreathing can occur. This is referred to as *nocturnal hypoventilation*. Initially people will wake up briefly to limit how much their oxygen and carbon dioxide levels change. However, over time the brain gradually adapts to more abnormal breathing to the point where shallow breathing occurs even when awake. The lack of chest wall expansion further stiffens the rib cage so that more effort has to be put into taking a normal sized breath or the breaths will be smaller, further worsening oxygen and carbon levels. Since these changes in breathing usually occur over months or years, people are often unaware of the problem. Even if symptoms are reported, these may be mistaken as just the natural changes in the neuromuscular disorder rather than a sleep disorder and not investigated further.

Once problems related to either inspiratory or expiratory

muscle weakness are recognised, appropriate respiratory therapy can be offered.

There are a range of devices to assist “breathing in”. Generally, we use a technique known as *non-invasive ventilation* which allows assistance of someone’s breathing without the need for an invasive breathing tube. Basically, a mask fits over the nose (or nose and mouth) and is attached to a small portable device that is able to pump air into the lungs to expand them. Most commonly (and ideally) this is started when someone is otherwise well but shows signs of sleep disordered breathing. However, it can also be started when a person is unwell in hospital and having breathing difficulties, for example during a severe chest infection or pneumonia. A *bilevel* ventilator is the most common device currently used. It provides two levels of pressure: a higher pressure in to inflate the lungs and increase the depth of breathing with a much lower pressure on breathing out to allow carbon dioxide to be cleared from

the system. Additional breaths are usually added to maintain a regular and normal breathing rate. Non-invasive ventilation during sleep is now seen as a standard part of care for people with respiratory muscle weakness.

It is important to identify the appropriate window of time to start night time ventilation. Evidence suggests that starting treatment before respiratory problems are present may actually be harmful. On the other hand, delaying therapy for

too long could lead to people presenting in a crisis situation, limiting treatment options. There are three opportunities for commencing night time ventilation: when awake carbon dioxide levels are found to be above normal (which is why blood gases are taken); when symptoms of underbreathing are reported (which is why a sleep history is taken) or if underbreathing during sleep is detected (which is why a sleep study or other night time monitoring is performed).

Initially, therapy is used at only night, which effectively reverses daytime respiratory failure and symptoms. However, if the underlying muscle disorder is progressive, daytime breathlessness or fatigue can appear months or years after commencing treatment as weakness increases. If this occurs, additional daytime ventilation will be needed. However, bilevel ventilators are not suitable for continuous use and the nose is



Dr Amanda Piper

not meant to have pressure on it constantly. Once ventilation is required more than 16 hours a day, alternative masks and methods of breathing support need to be considered. Most commonly a mouthpiece is used combined with a different type of ventilator that has an internal battery which can operate for 4-6 hours without being connected to the mains power. By using a combination of mask ventilation at night and mouthpiece or nasal plugs during the day, a substantial number of people with little or no spontaneous breathing capacity can have their breathing supported 24 hours a day.

While ventilators deal with the problems of weak inspiratory muscles, it is also important to develop skills in managing chest infections and to maintain good chest wall expansion. This is done by undertaking a program of regular lung inflation using a technique known as "lung volume recruitment" or "breath stacking". With this technique, a series of breaths are "stacked" together to maximum capacity before exhalation. Not only does this help maintain chest wall flexibility, it is also used to

improve the effectiveness of cough by increasing expiratory flow rates. The simplest and most accessible way to achieve this is to use a self-inflating bag to squeeze breaths in via a mouthpiece or mask, filling the lungs with air (see photo).

If secretions in the chest are present and cough remains ineffective despite using breath stacking, a mechanical cough assist device can be used. Both breath stacking and mechanical cough assist are techniques which require training and practice to perform correctly, but are important and valuable additions to the respiratory management of people with respiratory muscle weakness. Being aware of the options for managing the complications arising from respiratory muscle weakness is important so people have the opportunity to discuss and trial different treatment techniques.

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## Medical Seminar

On Saturday 17 November 2012, MDNSW hosted a Medical Seminar at Shepherd's Bay Community Centre, which was attended by about 60 members and their families.

Dr Amanda Piper, from the Department of Respiratory and Sleep Medicine at Royal Prince Alfred Hospital at Camperdown and Dr Nigel Clarke, Clinical Geneticist and Senior Lecturer from the University of Sydney Institute for Neuroscience and Muscle Research, were the guest speakers.

Dr Piper spoke on muscle weakness of the chest wall and the use of non-invasive ventilation and the equipment available.

Dr Clarke informed the group about the major advances in DNA testing and how these advances are now being adopted by diagnostic laboratories.



Victoria Berg and Maryanne Murray



Prof Corbett and Dr Nigel Clarke

After a delicious afternoon tea, our very own Maryanne Murray and Victoria Berg introduced the new online Training Program developed by MDNSW and funded by ADHC. This program is easily accessible on our website and will provide a basic understanding of neuromuscular disorders for people living with a neuromuscular condition as well as health and community professionals.

The afternoon concluded with the opportunity to ask questions of the guest speakers. People were reluctant to leave at the end, wanting to stay around and chat.

Our new offices were officially opened at the conclusion of the Medical Seminar, where members and their families enjoyed inspecting our new larger office!

**Jenny Smith**, Project Assistant