Why do people with neuromuscular disease develop breathing problems?

People with neuromuscular disease, such as Motor Neurone Disease or Duchenne Muscular Dystrophy, can develop problems with their breathing. This is because the breathing muscles may become weak. These muscles are also important for coughing, sneezing and sniffing and so they can become more difficult than usual.

What problems can weak breathing muscles cause?

You may develop chest infections more easily than other people because it is difficult to clear phlegm (sputum) from your lungs. You may experience other problems such as breathlessness, particularly when lying down, or find you have difficulty sleeping at night.

When should I ask for help?

If you notice any of the following symptoms:

- You cannot lie flat because you get short of breath
- You become breathless doing general everyday activities
- You are having a lot of chest infections
- You are finding it difficult to clear phlegm (sputum)
- If your cough strength measured with a peak flow meter falls below 270L/min (litres per minute)

What other symptoms should I be aware of?

If you notice you are tired during the day, need lots of cat naps, cannot sleep for long periods without waking, have headaches on waking, difficulty swallowing, general malaise, or lack concentration.

Who should I tell?

Let your GP, Nurse, or Physiotherapist know. They can refer you to a specialist who deals in breathing problems.

What can be done if I am having difficulty removing phlegm?

You should be referred to a Physiotherapist who can teach you techniques to help your cough be more effective.

What are these techniques and why do I need them?

When we cough we first take a big breath in. This allows us to have plenty of air to cough out, which makes the cough move the phlegm. When your breathing muscles are weak you may be able to take only a small breath in. If that is the case, you need a way to help you take a bigger breath in before coughing.

You may also, or instead, find your tummy muscles are too weak to cough properly. In this case, you need a method to make your cough stronger.

So, in general, the methods involve either helping you take a bigger breath in, or helping you cough more forcibly. Some can help you do both.

What are the methods to help me take bigger breaths?

We call this achieving ‘maximum insufflation capacity’ or MIC, that is, the largest amount of air you can hold in your lungs.

You may be able to get an extra large breath from your non-invasive ventilator, if you use one. If you don’t use one, then you can get a bigger breath from a special bag, which you or a helper squeeze to fill your lungs, or from a mechanical Insufflator-exsufflator. If these methods don’t give you a big enough breath in one go, then you can use a method called breath stacking.

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What is breath stacking and how does it help?

Breath stacking involves taking several small breaths in without breathing out in between. You do this until you can take no more air in. By breath stacking like this you can reach a big breath which makes your cough stronger. Some people need the help of breath stacking via the special bag or the non-invasive ventilator machine. Alternatively, some people find Glossopharyngeal Breathing a good way to breath stack.

What is Glossopharyngeal Breathing (GPB) and how does it help?

GPB is also known as ‘frog breathing’, because you learn how to gulp air into your lungs like a frog does. This is a way of breath stacking and helps increase the size of your breath. This will make you cough stronger. A specialised physiotherapist will need to teach you how to do this.

What is a non-invasive ventilator and how does it help?

The non-invasive ventilator is a simple machine that helps give you breaths when you want them. It can be used to help with your regular breathing and many people use it on a daily basis, usually at night. It can also be used to give you a bigger breath, to help you cough better.

What are the methods to help me cough more forcibly?

There are three possible ways you can get help to cough more effectively: with a technique called manually assisted coughing, with a mechanical In-exsufflator, or by respiratory muscle training to make your muscles stronger.

What is manually assisted coughing and how does it help?

Very firm pressure is applied to your stomach, or your chest, at the same moment that you cough, making your cough more effective; in effect, helping push the air out more rapidly. Usually, another person applies this pressure. Your carer can be taught to do this. Some people are able to apply this pressure by themselves. Your physiotherapist will help you select the method that is best for you.

What is a mechanical in-exsufflator and how does it help?

The in-exsufflator is a machine that helps you take a bigger breath in by inflating your lungs. It then helps you cough by reversing the airflow and creating a gentle sucking pressure, which help makes your cough stronger.

What is respiratory muscle training and how can it help?

This is an exercise programme to help your respiratory (or breathing) muscles get stronger. It involves breathing in (or out) against a resistance so that the muscles that you breathe in with (or breathe out and cough with) can be exercised. However this will only work for some people. Speak to your physiotherapist to see if you are suitable for this type of training.

Ask for the patient Action Plan to help you and your physiotherapist better manage your condition: Web Appendix 10 on the British Thoracic Society website.

Who should I contact if I do not have a physiotherapist?

Ask your GP or specialist to refer you to a physiotherapist who specialises in neuromuscular disease or breathing problems.

Useful Contacts

Your physiotherapist is __________________________

Association of Chartered Physiotherapists in Respiratory Care
www.acprc.org.uk

British Thoracic Society
www.brit-thoracic.org.uk

British Lung Foundation
www.lunguk.org

Chartered Society of Physiotherapy
www.csp.org.uk

Motor Neurone Disease Association
www.mndassociation.org/

Muscular Dystrophy Campaign
www.muscular-dystrophy.org/

Post Polio Support Group
www.ppsg.ie

NHS free smoking helpline 0800 0224332
www.smokefree.nhs.uk