A South West Neuromuscular Network Publication

Need to Know

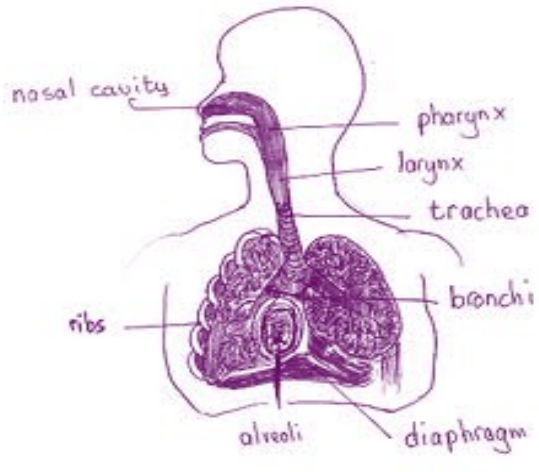
Nice to Know

ABOUT BREATHING

Guidance on breathing and non-invasive ventilation (NIV) for children from the age of 2 years living with a neuromuscular weakness

This leaflet has been designed to answer some of the questions you may have about how your breathing may be affected by a neuromuscular condition, and about breathing support delivered by a mask and ventilator (or 'non-invasive ventilation').

How Do The Lungs Work?



The main job of your lungs is to collect Oxygen from the air that you breathe in and deliver it through your blood to the rest of your body. The lungs then collect Carbon Dioxide, which is a waste gas, from your body and remove it through the air that you breath out again. The inside of your lungs look like a giant sponge. It is a mass of fine tubes, the smallest of which end in tiny air sacs called the alveoli. There are around 3 million alveoli or tiny air sacs., and it's through their walls that the oxygen and carbon di-oxide are exchanged between the lung and blood.

How Might My Neuromuscular Condition Affect My Breathing?

You might find that, because of the way a condition affects your breathing, you feel short of breath when you exercise, or you are getting repeated chest infections. Breathing problems are sometimes more predictable with some types of neuromuscular condition, but everyone is unique. Some children are born with breathing problems. But mostly, breathing problems can develop because of the following reasons:

- If you have a smaller lung volume where the chest is more difficult to expand due to respiratory muscle weakness or a recent loss of ability to walk.
- The respiratory muscle weakness may also result in your cough not being powerful enough to clear secretions
- You have associated swallowing problems due to weak bulbar muscles which means food might be going down the wrong way
- Some other physical change such as 'Scoliosis' (a progressive curvature of the spine) can sometimes affect your chest cavity size and shape, meaning that your lungs can't grow and expand normally.

It is nobody's fault that they have a condition causing muscle weakness, or develop breathing problems because of it. ***More often breathing problems can be helped.***

Need to know...

A persistent cough, or repeated chest infections can often be managed with antibiotics and airways clearance techniques to help mobilise (or shift off) secretions that are difficult to clear from your chest or trachea. You may also need to increase your calorie and fluid intake in order to improve your energy levels and keep the lungs healthy.

How Might My Neuromuscular Condition Affect My Sleeping?

During the night we all breathe less. Your breathing condition could mean that this may become a problem and cause the oxygen level in your blood to fall, and the level of the waste gas, Carbon Dioxide to build up. It may cause you to wake up and interrupt your sleep. If this happens you may be sleepy the next day. In addition, a high Carbon Dioxide level in the blood causes the blood vessels in the brain to dilate, which can lead to a morning headache. The medical term for this is known as 'sleep disordered breathing'.

Need to know...

There are some tell-tale signs of 'sleep disordered breathing' which you will need to let your respiratory team know about, including: snoring, sweating, morning headaches, poor concentration in the day, irritability, lack of appetite at breakfast or morning sickness, restlessness or wakefulness at night, increased breathlessness, snuffly or laboured breathing, recurrent chest infections and blue tinged tongue or lips.

Checking That Everything's Working

The medical staff known as the "respiratory team" will look after your breathing. The team may include a doctor and/or physiotherapist and/or a nurse who is trained in breathing care. Breathing problems are very individual, so in order for the team to fully assess how well you can already breathe, you may be asked to do some simple and painless tests in clinic, like blowing or sniffing.

You may be asked to have a light probe temporarily clipped onto a finger or toe to check how much oxygen is in your blood (or oxygen saturation levels). Sometimes, it may be necessary for the team to check the oxygen and carbon dioxide levels in your blood more accurately, by taking a small blood sample from your wrist or ear-lobe (whichever you prefer). They may also carry out a sleep study in which the oxygen level in your blood is measured using a clip on your ear. The respiratory team will always take into consideration your thoughts and feelings about these tests.

Depending on the results of these tests, you may need to be shown some breathing exercises and airways clearance techniques to try at home.

Nice to know...

Your respiratory team are there to help you —by offering you some good advice about things that may really help, but also by listening to you about your feelings and choices. Your respiratory team recognises that respiratory care needs to be as individual as you.

Need to know...

There are occasions when some patients need supplementary oxygen to assist their breathing, particularly during acute episodes when they are unwell. Oxygen treatment alone is not a cure for breathing problems associated with muscle weakness and if the respiratory or medical team treating you thinks that oxygen is needed, they will regularly measure your oxygen saturation levels to check that the right amount of oxygen is being safely given to help you breathe more easily .

Getting On Top Of Breathing Problems:

More often, breathing problems can be helped and your respiratory team can suggest things to help you feel a bit better and get you back to doing the things you enjoy. Eating a good and nutritious diet, maintaining an ideal body weight, keeping a good posture, being active, and recognizing the symptoms of a chest infection early so that it can be quickly treated, are all things that you can do yourself to help you try to improve your breathing.

If you have a change in your physical ability, or you are due to have surgery, it is always a good idea to let someone in your respiratory team know this. This will help your team anticipate and try to prevent any breathing problems before they occur. If you are having surgery, the team will work with your surgical team and help to look after your care before and after the operation, with the aim of keeping your breathing function safe and stable.

Your respiratory team may also suggest you see other health professionals such as a speech and language therapist, or a dietician. A speech and language therapist can give advice as to how to prevent foods and liquids going down the wrong way by changing their consistency or by helping to improve your method of eating. A dietician can help to optimise your calorie and nutrition intake.

Need to know...

It's a good idea to try to keep up to date with immunisations, including the annual flu vaccination and pneumococcal immunisation which may also help. Ask your GP or respiratory team for details.

What Is Non-Invasive Ventilation?

If your respiratory team believes that you may benefit from some help with your breathing using a ventilator. The ventilator is a simple device that acts like a pair of bellows to help you improve your breathing. You will wear a mask over your nose, or nose and mouth. The ventilator delivers a gentle pressure through the mask which helps your lungs to expand.

When you first try this treatment, it may feel very strange and will take time to get used to the sensation of having some help with your breathing. Someone from your team may need to spend time with you to work out what sorts of speeds and pressures suit you best. When you use the ventilator it helps to keep the oxygen level in your blood at the right level and helps to get rid of the waste gas (Carbon Dioxide) which you breathe out. This should help you to get a better night's sleep and feel more refreshed on waking in the morning.

It is important that you are comfortable with your mask and that it is a correct fit. There are lots of masks available. Your team will follow up to make sure that the equipment is working and making you feel better.

Nice to know...

"Using a ventilator started out feeling a little weird and it took me a while to get used to wearing a mask all night. Now that I use it, I feel much better in myself and have more energy in the day".
Jamie, aged 16

Taking Equipment Home

When you take equipment home, it should be fully covered by a 'service and maintenance' contract, with yearly servicing being arranged for you by your respiratory team, so you shouldn't need to worry about it. But it's a good idea to discuss the practicalities of servicing and maintaining this equipment with your team, for example, who you will need to inform if it breaks down; how you will be supplied with a replacement ventilator, masks and tubing; and who you should contact for on going advice, so that you have all the information you will need to use and take care of your equipment at home.

Nice to know...

When you first try starting on a ventilator, try to use the mask and machine for an hour or so in the afternoon or early evening while watching TV or doing something you enjoy. This will help you get used to wearing the mask and also what if feels like having some help with your breathing. When you go to bed, try to put the mask on and fall asleep with it on. The aim is to use the machine all night every night but you can build up to that. The team should a advise how long you need to aim to use the ventilator machine for.

Travelling Abroad

The ventilator machine is portable, meaning that if you have one, you can take it abroad with you to use on holiday. Remember to take an electric plug adaptor with you if you are going abroad.

It is a good idea to give your respiratory team plenty of warning if you intend to fly. This will allow them to carry out a special test to help decide whether you need extra oxygen to breathe during the flight. They can also write a letter to the airline you are flying with to ask them to arrange this, and to enable you to take a ventilator or other respiratory equipment onto the plane.

Taking Care Of Your Breathing In The Long Term

If your breathing needs keeping an eye on as you grow older, once you reach an appropriate age to move to adult services (around the age of 16years +) your care will be 'transitioned' to an adult respiratory team or specialist. This process will ideally happen at a joint clinic where both the adult and child respiratory care teams will be present to discuss things with you.

As you get older and start to make decisions for yourself, it is important for you to have an advanced care plan so that any personal wishes about your future treatment and care are clearly known by those caring for you.

Need to know...

Your respiratory team are there to help you —by offering you some good advice on things that may really help you get on top of breathing problems, while listening to you about your feelings and choices. If you think that your breathing is affected by your neuromuscular condition, and you're not currently seen by a member of the specialist respiratory team, please ask the doctor who you normally see about your neuromuscular condition ,or your GP, for a referral.

Our 'Need to Know/Nice to Know' Quick Checklist:

Speak To A Respiratory Doctor or Physiotherapist If:

- ✓ You are worried that your breathing has changed for the worse
- ✓ You get recurrent chest infections or have a persistent cough
- ✓ You think you have developed symptoms of sleep disordered breathing (see page 2 of this leaflet)
- ✓ You are using a wheelchair more than before
- ✓ You have been told by a doctor that your heart needs regular monitoring
- ✓ You have been told by a doctor that you have scoliosis (curvature of the spine)
- ✓ You are due to have an operation
- ✓ You are about to book a holiday and wish to fly

Our Respiratory Team Is Here To Help You By:

- ✓ Assessing and taking care of your breathing so you can get back to normal
- ✓ Teaching you airways clearance and management techniques
- ✓ Planning with you, and other medical professionals, about your long-term care
- ✓ Teaching you about any new equipment before you take it home to use
- ✓ Referring you to other specialists as needed (dietician, speech and language)
- ✓ Answering any questions or concerns you may have about your breathing

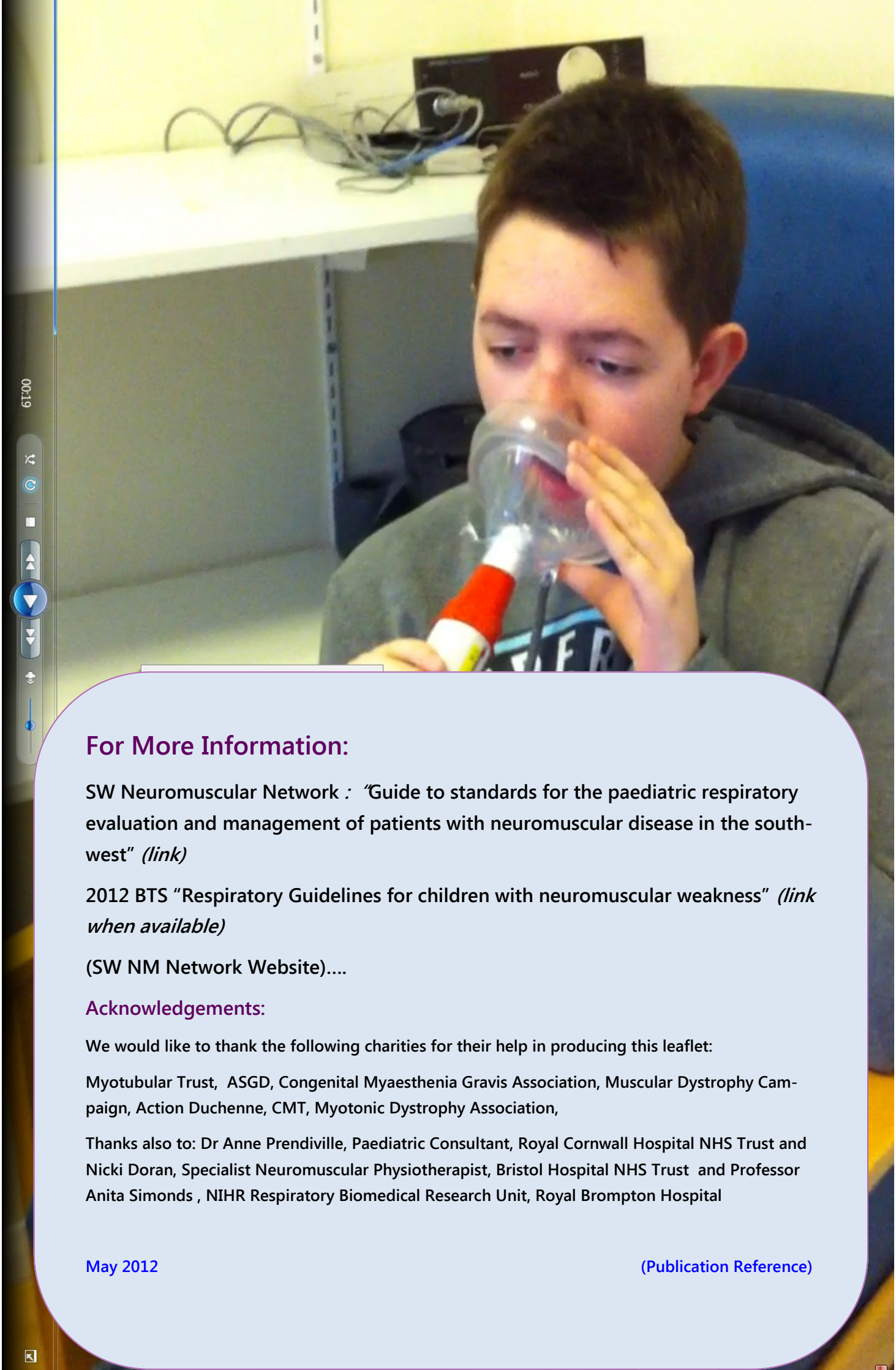
You Can Help Yourself By:

- ✓ Eating a healthy and nutritious diet
- ✓ Drinking plenty of fluids to keep you hydrated
- ✓ Trying to do some gentle exercise, as your condition allows
- ✓ Maintaining a healthy body weight
- ✓ Keeping up to date with immunisations
- ✓ Getting chest infections treated early with antibiotics

Need to know...

CPR is a good life skill for anyone to learn, please ask your respiratory team for advice.

Always dial 999 if someone stops breathing.



For More Information:

SW Neuromuscular Network *: "*Guide to standards for the paediatric respiratory evaluation and management of patients with neuromuscular disease in the south-west" *(link)*

2012 BTS "Respiratory Guidelines for children with neuromuscular weakness" *(link when available)*

(SW NM Network Website)….

Acknowledgements:

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