



**South West**  
Neuromuscular Network

**NHS**

South of England  
Specialised Commissioning Group

## Need to Know

*Nice to Know*

# About Breathing

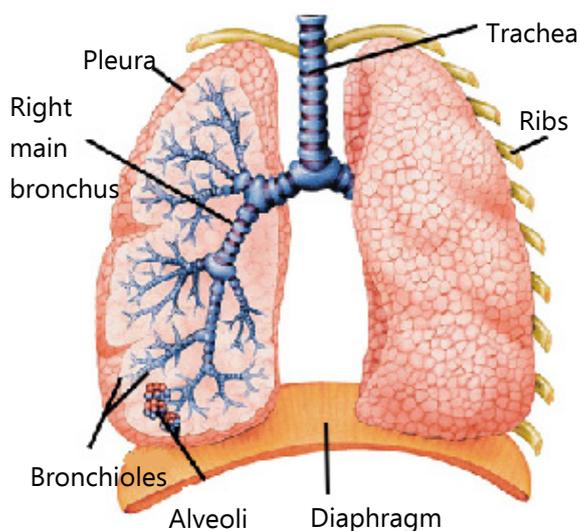
**Guidance on breathing and non-invasive ventilation (NIV) for children from the age of two years old living with a neuromuscular condition**



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 (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 (CO<sub>2</sub>), which is a waste gas, from your body and remove it through the air that you breathe out again. The inside of your lungs looks 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 three million alveoli, or tiny air sacs, and it's through their walls that the oxygen and CO<sub>2</sub> are exchanged between the lung and blood.

## How might my neuromuscular condition affect my breathing?

You might find that because of the way your 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 your 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 throat muscles which means food might be going down the wrong way
- ◆ Some other physical change such as scoliosis (a progressive curving 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 than not 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 secretions that are difficult to clear from your chest or trachea (windpipe). You may also need to increase your calorie and fluid intake in order to improve your energy levels and keep your lungs healthy.

## Getting on top of your breathing problems

Breathing problems can often be improved. Your respiratory team can suggest things to help you feel a bit better and get you back to doing the things you enjoy. Some of their suggestions may include:

- ◆ Eating a good nutritious diet
- ◆ Maintaining an ideal body weight
- ◆ Maintaining a good posture
- ◆ Keeping active
- ◆ Learning to recognise the symptoms of a chest infection early so that it can be quickly treated.



These 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 will help your team anticipate and try to prevent any breathing problems before they occur. If you are having surgery, the respiratory 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 on 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 nutritional intake.

### Nice to know ...

If you think that your breathing is affected by your neuromuscular condition, and you are not currently seen by a member of a specialist respiratory team, you should talk to your neuromuscular specialist or your GP. They may decide to refer you to a respiratory specialist for further advice.

### 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.

## Checking that everything is working

Your respiratory team will look after your breathing. The team may include a doctor, physiotherapist or 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 small probe put on your finger or toe to check your oxygen saturation (how much oxygen is in your blood).



Sometimes it may be necessary for the team to check the oxygen and CO<sub>2</sub> levels in your blood more accurately by taking a small blood sample from your wrist or earlobe, 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.

### 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.

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. It may also be recommended that you use non-invasive ventilation (NIV) to help control your symptoms.

### Need to know ...

There are occasions when some people need extra oxygen to assist their breathing, particularly during episodes when they are unwell. Oxygen treatment alone is not a cure for breathing problems associated with muscle weakness. If your 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.

## What is non-invasive ventilation (NIV)?

If your respiratory team believes that you may benefit from some help with your breathing they may suggest using a ventilator. A 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 may take time to get used to the sensation of having some help with your breathing.

### Nice to know ...

When you first start trying out your 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 it 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 advise how long you need to aim to use the ventilator machine for.

### 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 and have more energy in the day".

*Jamie, aged 16*

### Nice to know ...

"It took a little while to get used to wearing a mask and being ventilated. But as soon as I started to feel the benefits I really didn't mind anymore. Now it's just second nature".

*Ceri, aged 11*

Someone from your respiratory team may need to spend time with you to work out what sort 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 (CO<sub>2</sub>) which you breathe out. This should help you to get a better night's sleep and help you to 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 respiratory team will follow up to make sure that the equipment is working and is making you feel better.

## Why might I need to use non-invasive ventilation (NIV)?

During the night we all breathe less. This is because the muscles that help us breathe are more relaxed during sleep and breathing requires more effort when we are lying down.

Your neuromuscular condition could mean that this may become more of a problem for you and can cause sleep disordered breathing. Sleep disordered breathing is when the oxygen level in your blood falls and the level of the waste gas (CO<sub>2</sub>) builds up overnight.



The more common symptoms of sleep disordered breathing include morning headaches or loss of appetite at breakfast. Additionally, it can sometimes cause you to wake up a lot through the night and have interrupted sleep causing you to feel sleepy or lacking in energy the next day.

### Tell-Tale signs of sleep disordered breathing

- ◆ Snoring
- ◆ Sweating
- ◆ Morning headaches
- ◆ Poor concentration during 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
- ◆ Blue tinged tongue or lips.

Always let your respiratory team know if you have any of these symptoms.

If a diagnosis of sleep disordered breathing has been made, you are likely to benefit from the extra support provided by ventilation and get a much better night's sleep.

Most people only ever need to use NIV at night while they are sleeping. However, there are other times when it can be very helpful, for example, during routine airways clearance therapy and chest physiotherapy. Using your NIV will give you bigger breaths, open up your airways more effectively and really help to clear any secretions in your lungs.

Some people may also find it helpful to use NIV for periods during the day if feeling unwell or more tired than usual. Please talk to your respiratory team for more help and advice or if you are concerned that you are using NIV more regularly and for longer periods during the day than usual.

## Taking your non-invasive ventilator home

When you are given equipment to take home it should be fully covered by a service and maintenance contract with yearly servicing arranged for you by your respiratory team —so you don't need to worry about it.



It is a good idea to discuss the practicalities of servicing and maintaining this equipment with your respiratory team. Make sure you know:

- ◆ Who you will need to inform if it breaks down
- ◆ How you will be supplied with a replacement ventilator, masks and tubing
- ◆ Who you should contact for on-going advice.

Make sure you have all the information you need to use and take care of your equipment at home.

**Use this space to write down important contact numbers and details regarding the maintenance of your ventilator equipment:**

Respiratory Team: .....

Equipment Supplier: .....

Machine Model: .....

Serial Number: .....

Machine Settings: .....

Mask Name / Size: .....

Other supplies you may need to order (ie, filters, tubing, etc):

## Taking care of your breathing in the long term



It is possible that your breathing will need to be monitored as you grow older. Therefore, once you reach an appropriate age to move to adult services, around the age of 16 years plus, your care will be transitioned (moved) 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.

### Nice to know ...

The NIV ventilator machine is portable, therefore, you can take it with you to use on holiday, or use it as you would do normally during travel. Please ask your respiratory team for portable batteries to power your ventilator when you don't have access to a power socket. **Remember to take an electric plug adaptor if you are travelling abroad.**

## Travelling 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.

Your respiratory team can write a letter to the airline you are flying with to ask them to arrange for extra oxygen during your flight if they think it is necessary. This letter would also make it easier for you to carry a ventilator or other respiratory equipment with you in your hand luggage.



Having to use a ventilator should not stop you from travelling abroad, but you will need to notify the airline company about it when you book.

## Don't forget to have fun too



Here are some suggestions for making breathing exercises fun. Why not try some of them, as your breathing condition allows, or make up some of your own? If you can, try to be active every day.

### Blow Football

Take two straws, a light feather, a scrunched up piece of tissue or a ping pong ball. Place the feather, tissue or ball on

the centre of a flat surface and use the straws to blow it towards your opponent. You can even set up mini-goals! If you have difficulty blowing or sealing your lips around the straw, experiment using shorter, thinner and wider straws for the most effective result.



### Looney-Ballooney

The object of the game is for you to try to kick, punch or pat a balloon through a doorway (or "the goal"), while the other person blocks the doorway to prevent it from getting through. LED light-up balloons are slightly weightier and more accurate to direct.

**Singing, whispering and shouting** are very effective exercises for your lungs. Try to think how you can build some voice exercises into everyday life. You could try singing along to You Tube, karaoke, nursery rhymes or songs on the radio.

**Learning to play a wind instrument** will really help you to expand your lungs — whether it's a harmonica, recorder, flute, trumpet, saxophone or trombone. If none of these appeal, why not simply blow a whistle?



**Exercising in water** gives a good cardiovascular (heart and lung) workout. Warm water is nicer to exercise in if you have limited mobility. You can just splash about or blow a ping pong ball across the surface of the water or try cycling on a wobble float. **Note to parents/carers:** Children should be supervised in water at all times and use appropriate buoyancy support as necessary.

**If in doubt about what you can do, ask your respiratory team or physiotherapist for advice.**

## Your Need to Know / Nice to Know 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 5)
- ◆ 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.

### Your 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 therapist)
- ◆ 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 yourself 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 ...

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

**Always dial 999 if someone stops breathing**

## **My Notes/Questions:**

## Need more information?

### South West Neuromuscular Network

Tel: 0117 330 2597 / email: [swscg.neuromuscular@nhs.net](mailto:swscg.neuromuscular@nhs.net)

Web: <http://www.swscg.org.uk/networks/Neuromuscular/Neuromuscular.htm>

### Acknowledgements

The South West Neuromuscular Network would like to thank Wendy Hughes, Myotubular Trust, Dr Anne Prendiville, Consultant Paediatrician, Royal Cornwall Hospital NHS Trust and Nicola Doran, Specialist Neuromuscular Physiotherapist, North Bristol Hospital NHS Trust for writing this booklet, and Professor Anita Simonds, NIHR Respiratory Biomedical Research Unit, Royal Brompton Hospital, London.

Thanks to the Network's Patient Reference Group for their contribution. This group includes representation from:

- ◆ Myotubular Trust (<http://www.myotubulartrust.org>)
- ◆ Myotonic Dystrophy Support Group (<http://www.myotonicdystrophysupportgroup.org/>)
- ◆ Association for Glycogen Storage Disease (AGSD-UK) (<http://www.agsd.org.uk>)
- ◆ Myasthenia Gravis Association (<http://www.mga-charity.org>)
- ◆ CMT UK (<http://www.cmt.org.uk/>)
- ◆ Muscular Dystrophy Campaign (<http://www.muscular-dystrophy.org>)
- ◆ Action Duchenne (<http://www.actionduchenne.org>)

If you would like further copies of this booklet or in another format, please contact the South West Neuromuscular Network on 0117 330 2597