

Non-invasive ventilation for neuromuscular disease

 Information for patients



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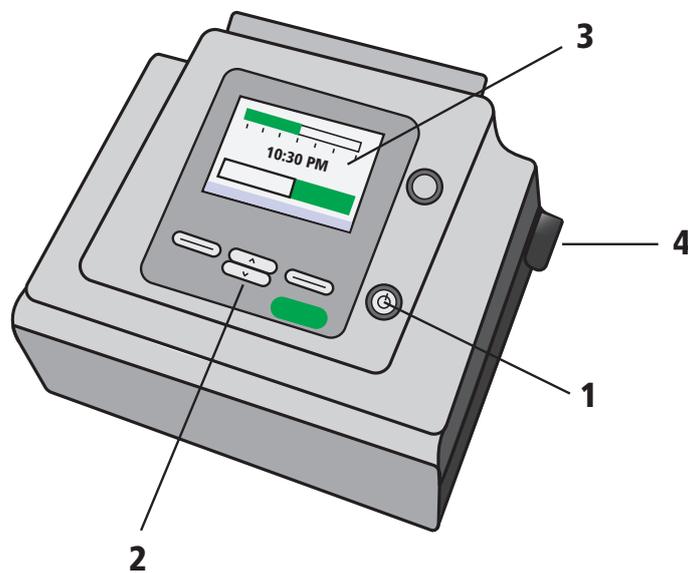
What is non-invasive ventilation?

You might need non-invasive ventilation (NIV) due to weakness in the muscles that you use to breathe, or due to changes in the shape of your chest, or both. These changes mean that you can struggle to get enough oxygen into your lungs, or get the waste gas (carbon dioxide) out.

NIV is usually first needed at night, when your breathing muscles are more relaxed and you breathe less deeply and less often.

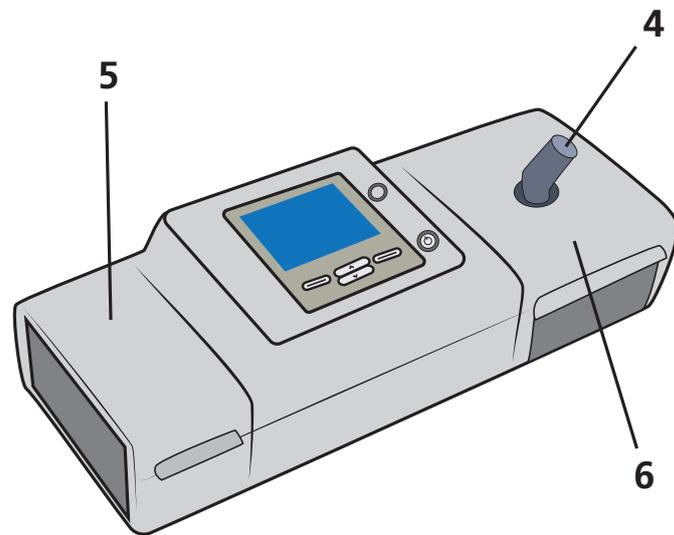
The aim of using NIV is to help to reduce the symptoms associated with weak breathing muscles such as breathlessness, poor sleep, headaches, tiredness, and fatigue during the day.

My A40 NIV machine



My A40 NIV machine

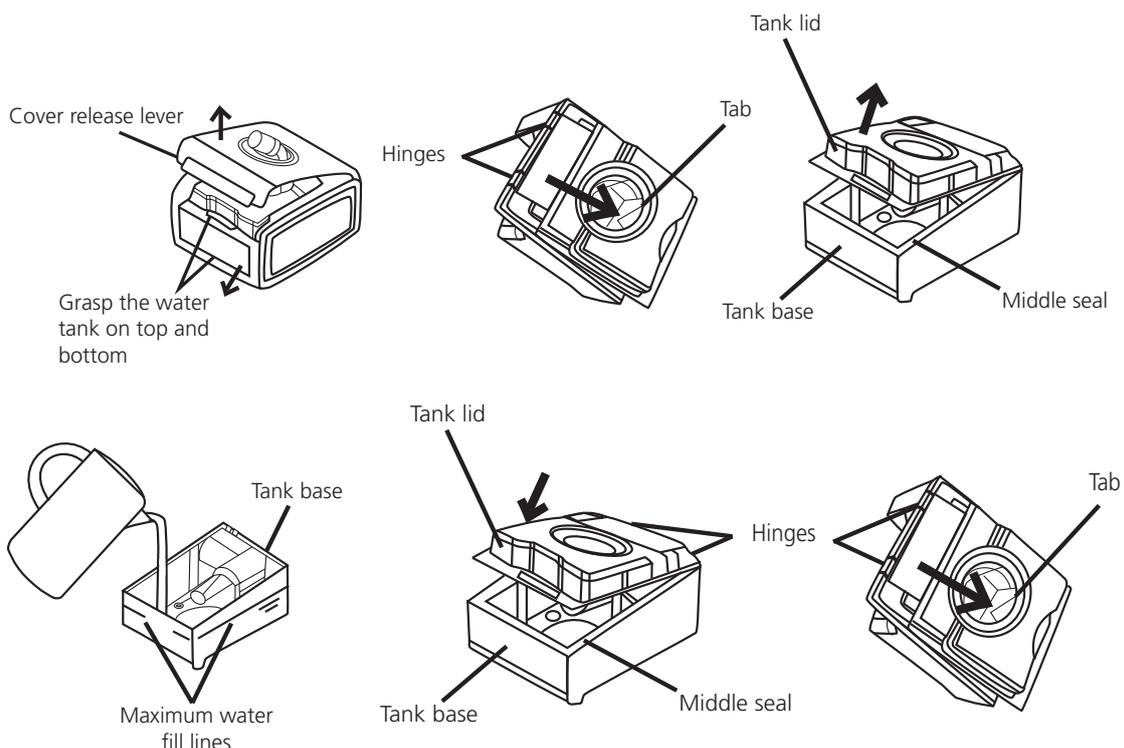
1. **Power button.** Turns the NIV machine on and off.
2. **Navigational buttons.** Allow you to start and end treatment as well as change settings.
3. **Main screen.** Where you navigate the menus and view the settings.
4. **Circuit connector.** Connect the NIV circuit here.
5. **Battery compartment.** Where the battery is stored (this is optional)
6. **Humidifier.** A water chamber to help prevent the NIV from causing a dry mouth and nose (this is optional).



NIV machine with optional battery and humidifier

Using a humidifier

- Some people find that using NIV causes dryness to the nose and mouth. This can be uncomfortable. If you suffer from dryness, a humidifier can be added to the NIV to help prevent this. The humidifier attaches to the side of the machine. It moistens the air that you are breathing in by heating water in the chamber that the air passes over.
- You should keep the chamber topped up with either cooled, boiled water, or sterile water. Fill the chamber up to the line on the front. A full chamber should last all night.
- You can set the humidifier between 1 (the lowest and coolest setting) and 5 (the highest and warmest setting) depending on how dry your mouth feels and how warm you like the air. Setting the humidifier to 0 turns it off.

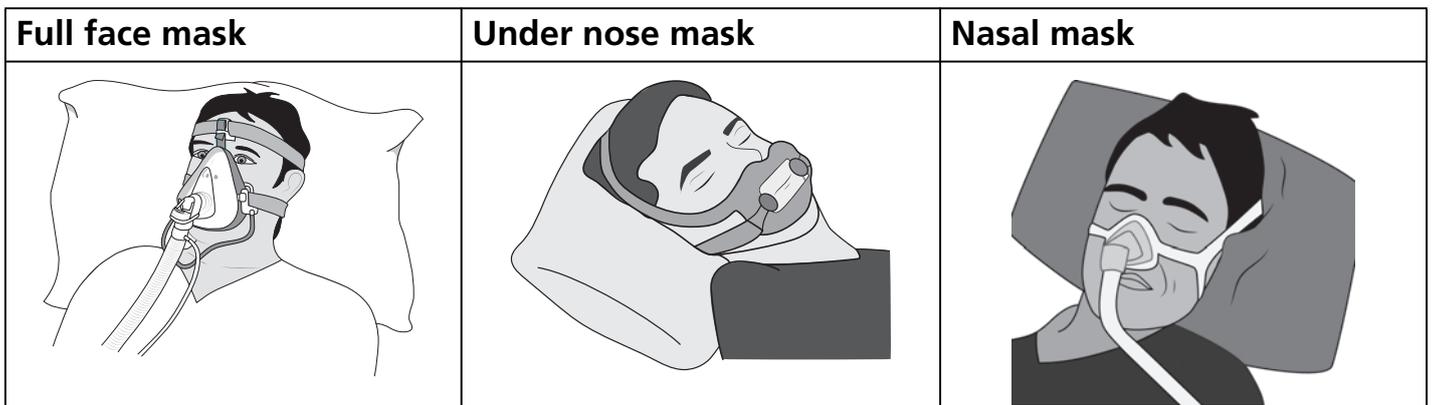


Using a battery

- If you need the NIV machine when you are not at home or close to a power supply, a battery can be added. The battery will give around 3-4 hours' use of NIV without having to plug it in.
- The battery attaches onto the left side of the machine. The battery will automatically charge when the machine is plugged in to a mains power supply.
- The amount of charge in the battery is displayed on the main screen.

My mask (interface)

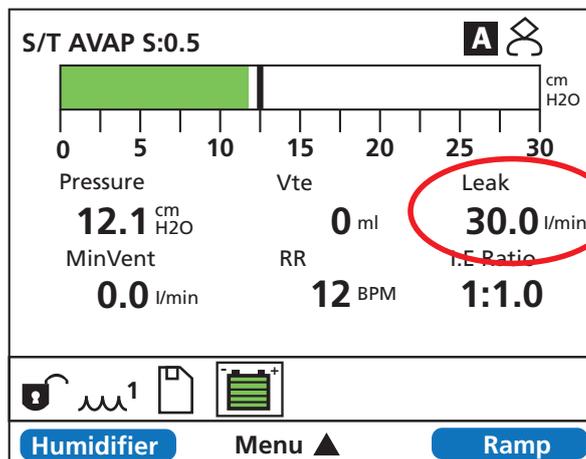
There are many different types of masks that can be used with NIV. Some of the commonly used mask types are pictured below. Your healthcare professional will advise on the best mask for you.



It is important to make sure that your mask fits well and doesn't leak too much. The mask should be snug on your face and shouldn't feel too tight. Once the velcro straps have been adjusted, most people find that the mask can be put on and taken off using just the clips or magnets.

If the straps do need adjusting, it is best to adjust both sides at the same time. Adjust the top two straps together and then the bottom two straps together. It might be easier to ask someone else to do this for you.

Once the mask is fitting well and the NIV machine is working, you can check the amount of leak on the main screen. Try to ensure the leak shown on the main screen is less than 60l/min.



My mask(s)

Date	Mask Type	Mask Name

Starting to use NIV

Most people usually first need NIV at night. Because NIV takes some getting used to, it is normal to start using it for short periods in the day at first. This helps you get used to the feeling of the mask and the pressures. You should try to gradually increase the amount of time that you can keep the NIV on. When you are comfortable, you should try to use the machine at night while you are asleep, as this will usually give you the most benefit. If you find the mask or pressure uncomfortable you should contact your healthcare professional. It might be possible to try different masks, or alter the pressures to make the NIV more comfortable for you.

Set up and use

Plug the NIV machine into the mains power using the power cord supplied.

Set up and connect your circuit to the circuit connector on the side of the machine (or on top of the humidifier if you have one).

Place the NIV machine on a flat, stable surface. If you use a humidifier, try to position the NIV machine so that it is lower than you when you are asleep. This will help to stop water collecting in the tube.

Turning the machine on

1. To turn the machine on, press the green power button. The machine can either be turned on before or after fitting the mask, depending which you prefer.
2. Press the right hand navigation button that says Therapy above it. The machine will start after a few seconds.

Turning the machine off

Press the green power button. Then press the right hand navigation button to confirm that you want to switch the NIV machine off. This puts the NIV machine into standby.

If you put the machine on standby you only need to press Therapy to start it again.

To turn the NIV machine off completely, press the green power button again then press the right hand navigation button to confirm that you want to switch the NIV machine off completely.

My A40 NIV prescription

Your settings will be documented below

Date	Mode	IPAP	EPAP	Rise time	Backup RR	Ti

Note for health professionals: these settings may not be kept up to date. Up to date settings and further clinical information can be accessed via the 'Respiratory Physiology Results' page on the STH intranet (for STH staff only) or by contacting the staff on the back of this booklet.

Starting NIV treatment

When you are first using the NIV, try to relax and breathe as normally as you can. It often helps to do something that will distract you from the NIV, like watching television or reading.

It takes time and effort to get used to the NIV but most people find that they can get used to it if they keep trying.

Time on NIV

Most people with neuromuscular disease will need to use NIV at night time when they are asleep. You should try to keep the NIV on all night to get the most benefit from it. You should also use the NIV if you have a sleep during the day.

If you are struggling to cope with wearing the NIV at night, speak to your healthcare professional. You might be able to try a different mask, or the NIV pressures might be able to be adjusted to make it more comfortable.

You can also use the NIV during the day to help with breathlessness.

Using NIV to help with chest infections

If you think that you have a chest infection you should contact your GP or the respiratory team.

Many people find that they can manage chest infections at home. During a chest infection you will probably find that you need to use NIV more often. You should use it more often if you feel more short of breath, or if you feel more tired and / or sleepy during the day. People who normally use NIV at night often find that they need to use it during the day as well while they have a chest infection.

Going in to hospital

If you need to go into hospital you should take your NIV machine with you. You should also take this booklet with you to help the team looking after you know about your NIV settings, and know who to contact if they have any questions about your NIV.

Becoming more dependent on NIV

The amount of time that you use NIV is a personal choice. You should discuss your options with the people involved in looking after your NIV. The main aim of using NIV is always to help manage your symptoms and help to improve your quality of life.

If you need non-invasive ventilation for more than 16 hours per day you will be offered a second machine with a battery back-up.

You should make sure that your non-invasive ventilator is accessible at all times, including when you are out of the house, in case of a sudden increase in your symptoms, such as breathlessness.

You should also let your electricity supplier know that you rely on medical equipment that requires electricity. This means that you will receive additional services in case of a power cut.

If you have a metered electricity supply you should also contact your electricity company to let them know that you rely on NIV.

If you live in Yorkshire you should contact Northern Powergrid Priority Services Membership on **0800 169 2996** or register on the Northern Powergrid website; **www.northernpowergrid.com** (scan the QR code below):



Cleaning

Before cleaning, **disconnect the NIV machine from the power supply**. Do not immerse the machine in water or use any cleaning agents on the device. **Wipe the machine with a clean, damp cloth and then dry with a soft cloth**. Once the machine is completely dry then reconnect to the power supply.

Mask (daily care)

It is important to wipe the mask every day with a damp cloth. The more you look after your mask the longer it will last.

Mask (weekly care)

The mask needs to be washed every week. Separate the mask from all attachments; gently wash in warm, soapy water (mild detergent). Rinse with clean, warm water and allow to air dry before using again.

Tubing (weekly care)

The same procedure as with the mask, ensure all excess water is shaken off and hang the tubing vertically to allow the water to drain out.

Filter (weekly care)

Remove the sponge filter from the back of your NIV machine, this should be washed in a mild detergent every week and allowed to air dry before placing back in the machine.

Humidifier

If you have a humidifier you should **rinse the chamber out and change the water every day**. Once a week you should **wash the chamber in warm soapy water** (mild detergent) or put it in the dishwasher.

Frequently asked Questions (FAQs)

When I start to use NIV it feels quite claustrophobic, what should I do?

It might take you a little while to get used to wearing a mask on your face. You can start getting used to the sensation of the mask on your face by holding it in place, if you are able. Try to build up the time you can hold it in place until you feel ready to put the straps on.

Try to start using NIV for short periods of time and build up the amount of time gradually. It might help to do things that will distract you while you are using the NIV, such as watching television or reading.

When I start to use NIV the pressure feels too high, what should I do?

It might take you a little while to get used to the pressure when you start using NIV. It is normal for the pressure to feel strange to begin with, but most people get used to it quite quickly.

Try to start using NIV for short periods of time and build up the amount of time gradually. It might help to do things that will distract you while you are using the NIV, such as watching television or reading.

If you find that you are unable to get used to the pressure, you should speak to your healthcare professional. It might be possible to turn the pressure down, or to use a ramp setting that will slowly increase the pressure over the first 20-30 minutes of use.

The mask is leaking a lot, making strange noises while I am trying to sleep, or blowing air into my eyes, what should I do?

If too much air is leaking around the mask it might be uncomfortable or noisy. It might also stop the NIV from working effectively. Check the amount of leak on the screen and try to always keep the leak below 60.

You should try to adjust your mask to reduce the leak. Try moving the mask on your face to get it into a better position. If this doesn't work you might need to adjust the velcro straps. If this doesn't work you might need to remove the mask and try it again.

If you are unable to reduce the leak you should speak to your healthcare provider. They might suggest trying a different mask.

My skin is getting sore from the mask, what should I do?

It is important to keep a close eye on your skin when you start to use NIV. Your skin might be a bit red where the mask touches it, but this should disappear quite quickly once you remove the mask.

The mask should be snug on your face but shouldn't be sore. Keeping the mask clean will help prevent your skin becoming irritated and sore. If your skin is very red or sore you should contact your healthcare professional. It might be necessary to change your mask to one that fits better.

Useful contact details

My Neurology Consultant is

My Respiratory Consultant is

My Respiratory Physiotherapist is

I need replacement equipment, what should I do?

If you are worried that any of your NIV equipment is damaged or broken, you should contact the Pulmonary Function Unit (PFU) at the Northern General Hospital:

- **0114 226 9605**
Monday - Thursday: 7.30am - 6.00pm
Friday: 8.00am - 4.00pm

If you have problems outside these hours during the week or at a weekend and you need replacement equipment urgently you should contact Robert Hadfield 5 ward at the Northern General Hospital on:

- **0114 271 5092**



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