

Non-invasive ventilation (NIV) for cystic fibrosis

Information for patients

This leaflet answers some of the questions you may have about using non-invasive ventilation (NIV) for cystic fibrosis. It explains when to use NIV, how to use it for physiotherapy and how to clean your machine. It also has servicing details and a section for recording your own machine settings.

We will show you how to use your machine before you take it home. If you have any questions or concerns, please speak to the physiotherapists caring for you.

What is non-invasive ventilation (NIV)?

Non-invasive ventilation (NIV) is a treatment to help with your breathing. It involves you using a mask or mouthpiece connected to a machine which makes your breathing easier by reducing how hard your breathing muscles have to work. Each machine is set up to suit your individual needs.

Why do I need NIV?

The main role of your lungs and breathing muscles is to get rid of waste gas (carbon dioxide) and make sure that there is plenty of oxygen in your blood. If your lungs and breathing muscles aren't working correctly, carbon dioxide will build up in your blood which can lead to drowsiness, headaches and twitchiness. You may breathe less effectively due to carbon dioxide and not get enough oxygen so this can be a serious problem. NIV works to support your lungs and breathing muscles, helping to get rid of carbon dioxide. This is the main reason people need to use NIV.

NIV can also be used:

- as part of your physiotherapy treatment sessions to help clear phlegm
- for a short time if you have a bad chest infection to support your breathing muscles
- to support you during exercise to help you do more.

What are the benefits?

- It can ease the symptoms of increased carbon dioxide, such as morning headaches, drowsiness and twitching
- It can help clear phlegm
- It can improve your ability to exercise
- It may improve your sleep quality and energy levels in the day.

What are the risks or side effects?

- Pneumothorax, where air leaks and becomes trapped between your lung and chest wall. You may get a sudden sharp chest pain followed by pains when you breathe in. You may also become breathless. You will need to attend your local A&E urgently if this happens as you may need the air draining.
- Some people have reported trapped wind/stomach bloating. If you find you suffer with this, you may need your machine settings altering to reduce it.
- Some people find they may get sore skin where the mask sits on their face. If you find this is a problem, you may need to try different style masks or liners to find a comfortable fit.
- If you are using NIV overnight, you may find you get a dry nose or mouth. If this is the case, your physiotherapist can provide you with a humidifier to minimise this. They will explain how to use this if you require one.

Are there any alternatives?

There are no alternatives if you are using it to improve the symptoms of too much carbon dioxide in your body.

There are alternatives if you use NIV to help clear phlegm or for exercise. To clear phlegm, alternatives may be a different machine called The Bird, which helps you take a better breath in, but this can only be used in hospital or various breathing exercises. Your physiotherapist can explain the alternatives and advise on which would be the most useful for you. An alternative to using NIV for exercise may be to use oxygen for exercise instead.

Some alternatives may not be suitable for you to use and may be less effective than NIV. Your CF team or CF physiotherapist can advise you on this.

Your NIV machine

Machine model:

Serial number:

Settings	Physio	Rest
IPAP		
EPAP		
Ti		
Mode		
Oxygen		
Date		
Signature		

Your physiotherapist will fill in this table, so you know what settings to use at home. If you need to use the NIV machine with oxygen, a humidifier or a nebuliser your physiotherapist will advise

you how to do this and will advise in what order you should do these, for example, nebulisers before clearing phlegm.

You will need to make sure you keep your machine near where you will need to use it, so by your bed if you use it overnight or where you do clearance if you use it for physio only. Your physiotherapist will advise you on how long you should try to use the machine for.

Do I need to take my machine with me everywhere?

If you are using it to rest or sleep then yes, you do need to take it with you wherever you are going to be sleeping. This includes staying at friend's houses or going on holiday. Always bring it into hospital if you are admitted and make sure the staff know you use NIV.

Cleaning your NIV machine

Keep the machine clean and dust free. Mouthpieces and masks can be cleaned with warm soapy water and then be left to dry. We recommend you clean them daily. Your tubing and any headgear you may have should be cleaned weekly with warm soapy water and left to dry. The filter at the back of the machine will become dusty over time as you use the machine.

We recommend you take out the filter every week to remove excess dust and change it every 3 months. Your physiotherapist or Chest Unit can provide you with new ones.

If you use a humidifier with your NIV machine the water should be changed daily and the chamber cleaned in warm soapy water weekly. If lime scale builds up in the chamber you can clean in by using a small amount of lemon juice in water and leaving it in the chamber for half an hour. Clean the chamber after this with warm soapy water and allow to dry before filling with water again to reuse. We recommend using previously boiled and cooled water in the humidifier to reduce any bacteria that may be in the tap water.

Your physiotherapist can show you how to clean all the different parts of your machine and can give you any new/replacement parts you may need.

Servicing your NIV machine

Your machine needs to be serviced every year. The Chest Unit will contact you when it is due to be serviced and they will give you a replacement machine.



Examples of a NIPPY 3+ machine and a Quattro FX full face mask

Who can I contact with queries and concerns?

If you have any chest pain or shortness of breath while using your NIV machine, stop using it and contact one of the numbers below.

Monday to Friday, 8.30am to 5pm

Cystic fibrosis physiotherapist, tel: switchboard on 020 3299 9000 and ask them to page KH0119 or bleep 737188.

Out of hours

Adult cystic fibrosis ward on Lonsdale, tel: 020 3299 8652 and the on-call consultant will be contacted for you.

If you have problems with the machine or parts, such as masks and humidifiers, contact the Chest Unit, tel: 020 3299 2075, Monday to Friday, 8.30am to 5pm.

Care provided by students

We provide clinical training where our students get practical experience by treating patients. Please tell your doctor or nurse if you do not want students to be involved in your care. Your treatment will not be affected by your decision.

PALS

The Patient Advice and Liaison Service (PALS) is a service that offers support, information and assistance to patients, relatives and visitors. They can also provide help and advice if you have a concern or complaint that staff have not been able to resolve for you. They can also pass on praise or thanks to our teams.

PALS at King's College Hospital, Denmark Hill, London SE5 9RS

Tel: 020 3299 3601

Email: kch-tr.palsdh@nhs.net

PALS at Princess Royal University Hospital, Farnborough Common, Orpington, Kent BR6 8ND

Tel: 01689 863252

Email: kch-tr.palspruh@nhs.net

If you would like the information in this leaflet in a different language or format, please contact our Communications and Interpreting telephone line on 020 3299 4826 or email kch-tr.accessibility@nhs.net