Cystic Fibrosis Trust

Physiotherapy
Keeping your lungs healthy

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Name			
DOB		**Hospital number**	
Physiotherapy recommendations			

In partnership with

ACPCF
Association Of Chartered Physiotherapists In Cystic Fibrosis

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Contents

Introduction

If you have cystic fibrosis (CF), you may produce mucus in your lungs, which is thicker and stickier than the mucus produced by people who don't have CF. This thick mucus needs to be cleared.

This leaflet talks about physiotherapy and airway clearance for children and adults. If you have a young child you may prefer to use our leaflet on **Airway clearance for babies and young children with cystic fibrosis**.

For information on physiotherapy and more, please visit our website at **cysticfibrosis.org.uk/physioleaflets** or contact our Helpline.

Mucus, sputum, or secretions? People use different words to talk about the mucus in their lungs. Some people call it something different if it's in their lungs or if it has been coughed out. You can use the words you like, but here we will use the word mucus.

How are the lungs are affected by CF?

Below are two different ways of explaining how CF affects the lungs.

1. Start here

In CF the mucus in the lungs is thicker and stickier than normal. This makes it difficult to clear and easier for bacteria to get stuck in the mucus.

Bacteria can then build up in the mucus and cause an infection.

Infection causes the lining of the airways to become swollen.

Repeated infections can lead to scarring (fibrosis) of the lungs.

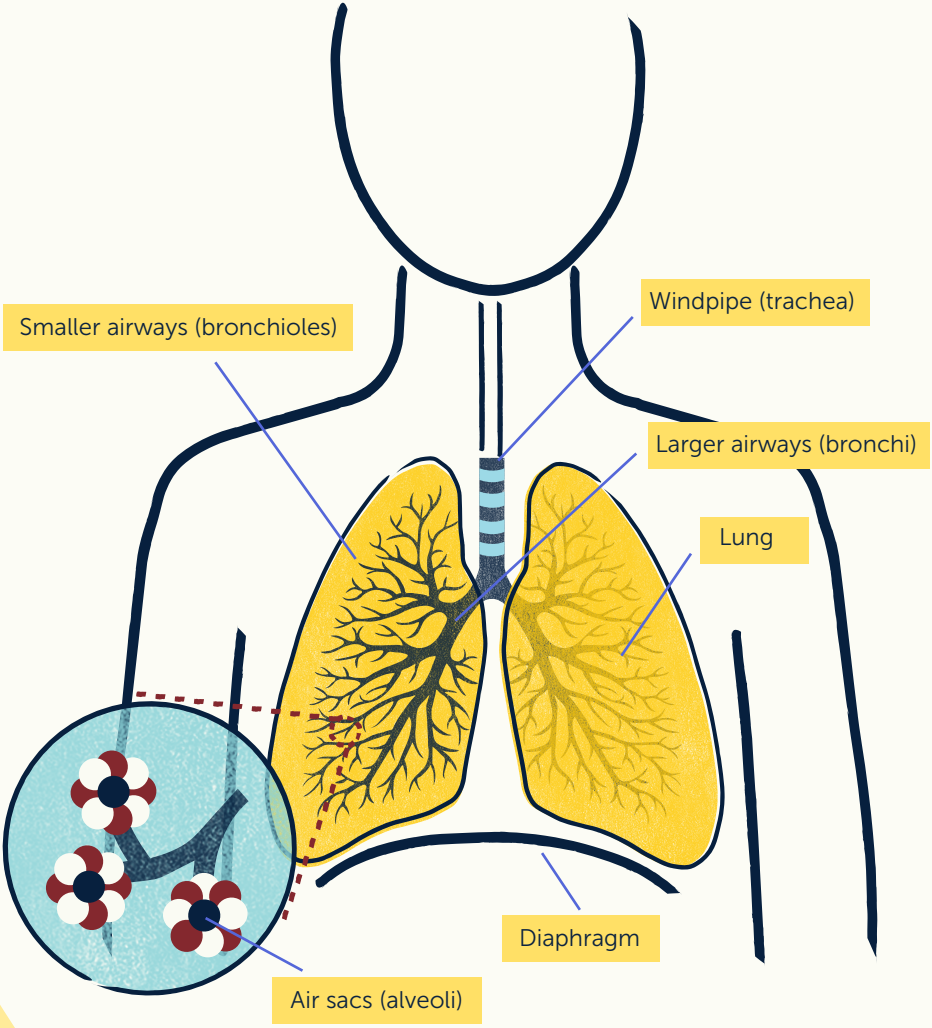
Infection also produces extra sticky mucus.

2.

- Thicker mucus
- Infection
- Swelling of the airways
- More mucus is produced
- Repeated infection

Lung damage

What are the lungs like inside?



Inside the ribcage there are two lungs that fill with air. Inside each lung there are thousands of air tubes called airways. These airways move air containing oxygen from the mouth to little air sacs at the end of each airway. The air sacs are called alveoli. Inside the air sacs, oxygen passes into the blood vessels and goes off around the body where it is needed. At the same time, waste gases (including carbon dioxide) are passed through the blood vessels back into the airways to be pushed out as we breathe out.

The airways are larger nearer the mouth, but divide into smaller and smaller tubes until they reach the air sacs.

When we breathe, air is sucked down through the airways to the air sacs.

The diaphragm, which is below the lungs, is the main muscle for breathing.

How the lungs keep clean

The lungs are naturally very good at keeping clean. The airways are lined with cells which have tiny hair-like structures on them called cilia. The cilia are covered with a layer of mucus which keeps the airways moist. Any dust, pollen or bacteria that is breathed in will get trapped in the mucus, and the cilia move back and forwards together to move the trapped particles up the airways and to the throat and mouth, where they can be coughed out or swallowed.

Keeping your lungs clear

What is CF physiotherapy?

Physiotherapy in CF is a name given to a number of different treatments that help keep your lungs healthy. CF physiotherapy includes airway clearance, exercise, and inhaled and nebulised medicines.

Why is airway clearance important?

If you have CF, you may have thick and sticky mucus in the airways in your lungs. It's important to try and clear the mucus because germs can get stuck in the mucus and can cause infections. Infections can cause swelling, block the smaller airways, and lead to more mucus being produced. All of this can stop your lungs from working well and can lead to lung damage. Your lungs can't repair any damage, so it's better to try to prevent the damage from happening.

CFTR modulator medicines and airway clearance

Some people with CF can use CFTR modulator medicines, like Kaftrio®, Kalydeco®, Orkambi®, and Symkevi®. These medicines might help with clearing mucus, but airway clearance may still be recommended by your CF team. Your team might think you need to do your airway clearance every day, or just when you're feeling unwell, to help keep your lungs clear and to keep you healthy. Speak with your CF team to make sure you know how much airway clearance you need to do to keep healthy, especially if modulator medicines are new to you.

Modulator medicines are quite new for treating CF, but some studies are showing they seem to help reduce the amount of sticky mucus, making the mucus easier to clear. Although this can help with keeping your airways clear and reducing the numbers of infections you might get, they can't change any lung damage that's already there.

How much airway clearance do I need?

The amount of airway clearance done, how often, and what type will vary from person to person and may even change on a day-to-day basis for some people. You may need to change your physiotherapy routine if you are unwell or feel you have more mucus, for example by doing more repetitions of breathing exercises or adding an extra session within your day. A physiotherapist can help you learn how to assess your chest, to help you to decide how much airway clearance to do each session.

If you produce mucus daily your CF team will usually recommend you do airway clearance once or twice a day. If you are ill and have an infection, you may need to increase it up to three or four times a day to try and keep on top of the extra mucus your body will produce.

Even if you don't produce much mucus, you may still need to do daily airway clearance to help keep your lungs healthy, to make sure you know how to do it correctly and/or to get into a good routine for when you get ill. You may be told to only do airway clearance when you have a chest infection. Your physiotherapist will advise you on the right amount of airway clearance specifically for you.

The type of airway clearance you use may change over time, as some types are better at different ages and at different stages of CF. You will need regular checks with a physiotherapist to make sure you are doing the right airway clearance for you.

It's vital to start airway clearance as soon as there are signs of a chest infection. This will help clear mucus from your chest, which helps to prevent lung damage.

What types of airway clearance are there?

There are many different types of airway clearance, ranging from using simple breathing exercises to using an airway clearance device (machine or equipment). A physiotherapist will work with you to find a type of airway clearance that is best for you. This will be based on your symptoms, health and lifestyle. It is very important that a physiotherapist checks how you do your airway clearance regularly so that changes can be made as you get older or if there are changes in your lungs.

As part of a physiotherapy routine some people use medicines, such as inhalers and/or nebulisers, with airway clearance. These medicines can help you breathe more easily, or loosen your mucus. There is more about these on page 13.

Please don't try a new type of airway clearance without first talking about it with your physiotherapist.

The next section is a basic guide to the most common types of airway clearance used in the UK, so you know what is available.

Breathing exercises

There are two main types of breathing exercises – the active cycle of breathing techniques (ACBT) and autogenic drainage (AD).

- **ACBT** is made up of deep breathing exercises, relaxed breathing, and huffs. People use deep breathing to move the mucus, rest in between deep breaths and then clear the mucus with a huff. Young children can be taught to do ACBT. It can be done with an airway clearance device.
- **AD** is a series of breathing exercises which uses high flows of air to move mucus from the smaller airways up into the larger airways so it can be cleared with as little effort as possible. AD can be taught to adults and children, and a parent or carer can be taught how to support their baby to do AD too.

Leaflets about '**Active cycle of breathing techniques**' and '**Autogenic drainage**' are available from our **Helpline** and at **cysticfibrosis.org.uk/physioleaflets**

Positioning or postural drainage

Positioning and postural drainage involves lying or sitting in different positions to use gravity to drain mucus from the lungs. Your physiotherapist may tell you to use different positions during your airway clearance, particularly if you have an area of your lungs which has an infection or collection of mucus. Postural drainage can be used at any age and is usually done with breathing exercises or with percussion.

Please note: Traditional postural drainage included some positions where a person would lay with their head tipped down below their body. Research showed that these positions could increase reflux of food from the stomach, so now physiotherapists do not routinely include these positions.

Percussion and vibrations

Percussion and vibrations are done to help loosen the mucus. Percussion or chest clapping involves using a cupped hand to clap the chest rhythmically. Vibrations (also called chest shaking) is done by giving several short rhythmic squeezes to your chest as you breathe out. These will be taught by a physiotherapist for safety, and you can do them on your own or a parent or carer can be taught how to do them for the person they care for. Percussion and vibrations are usually done with breathing exercises and/or with postural drainage.

More about positioning and percussion can be found in '**Airway clearance for babies and young children**' with cystic fibrosis. Available from our **Helpline** and at **cysticfibrosis.org.uk/physioleaflets**

Positive expiratory pressure (PEP)

Using a PEP device helps to open up your airways and get air behind the mucus to help it move higher up in your airway. Once it gets into the back of your throat, you can huff and cough the mucus out.

- A number of different PEP devices are available.
- PEP can be used at any age.
- Your PEP device will need to be checked regularly by your physiotherapist, as the resistance setting may need to be changed.

Bubble PEP is another type of PEP and can be useful for young children as it is more like playing. It uses exactly the same action as the PEP device described above does. Instead of using a mask or mouthpiece, a child will blow out through a plastic tube which is put into soapy water. Blowing into the tube in the water makes the pressure. A child will blows lots of bubbles and it can be a fun way of getting them to do their PEP.

Bubble PEP can be taught to a child when they are old enough to understand the difference between blowing and sucking. That might start at around two and a half years old.

We have leaflets about '**How to use a PEP mask**', '**Bubble PEP**' and '**PARI PEP®**' available from our **Helpline** and at **cysticfibrosis.org.uk/physioleaflets**

Oscillating PEP

An Oscillating PEP device is similar but as you breathe out and get the resistance or back pressure in your airways, it also vibrates your airways. These vibrations can help loosen the mucus away from the airway walls, allowing it to be cleared from your lungs. Different devices are available and your physiotherapist will help to decide which one is best for you.

The devices include:

- The Flutter®
- Aerobika®
- The Acapella®
- TheraPEP®
- The RC-Cornet®
- SoloPEP®

'**The Acapella choice**' is a leaflet available from our **Helpline** and at **cysticfibrosis.org.uk/physioleaflets**

Mechanical devices

There are a few mechanical devices, including high frequency chest wall oscillation (HFCWO) also known as 'the Vest', the Metaneb®, and the Physio-Assist Simeox®. This equipment is available in the UK but it is not used much by people with CF at home. CF centres may have these machines for people to use while they are in hospital, but they are only used if a physiotherapist sees a specific benefit for someone. This equipment can be expensive to buy and needs to be serviced regularly. There's also not much evidence to say that one type of physiotherapy is better than another. The ACPCF recommends that 'the Vest' is not used as the only form of physiotherapy, especially in children. You can read more about 'the Vest' here at **cysticfibrosis.org.uk/what-is-cystic-fibrosis/cystic-fibrosis-care/physiotherapy/vest-statement**

Non-invasive ventilation (NIV)

Non-invasive ventilation is often recommended overnight for some people with CF. It's normally used when there is a build-up of carbon dioxide (waste gas) and the person's oxygen levels are lower than they should be. NIV can be used with a mouthpiece, nose or face mask. Sometimes NIV will be used to help with airway clearance, as it can help people take a deep breath with little effort. If you're going to use NIV, it should be started with your CF team, and the machine settings need to be checked regularly.

Can I count exercise as my airway clearance session?

Both airway clearance and exercise are important parts of your physiotherapy routine. There is currently no evidence to say that just doing exercise is enough to keep your lungs clear of mucus.

Physical activity and exercise are essential parts of living with cystic fibrosis. It's great for your:

- Fitness
- Muscles
- Bone structure
- Mood
- Posture

Exercise and physical activity can also help you to keep your chest clear. **It's important to know that exercise can be good at loosening the mucus in your lungs, but it may not clear it.** When you are exercising, make sure you add in huffing and coughing to clear the mucus.

Some people with CF have good lung health, don't produce any mucus and do lots of exercise. If this sounds like you and you want to try counting an exercise session with huffs and coughs as a daily physiotherapy session, speak with your physiotherapist about it before making any changes to the physiotherapy you have been prescribed.

It is still very important you know how to do at least one type of airway clearance because there may be times when you can't do exercise, or the exercise is not enough to clear your chest. For example, you might:

- have an illness or infection
- have a sports injury
- be too tired to exercise.

Inhaled medicines with airway clearance

There are a few different types of medicines which can be breathed in (inhaled). They do different things.

- Medicines that open your airways are called bronchodilators. An example of a bronchodilator is Salbutamol (Ventolin®).
- Medicines that thin the mucus are called mucolytics. An example of a mucolytic is DNase (Pulmozyme®).
- Medicines that loosen and help clear mucus are called expectorants. Examples of expectorants are hypertonic saline or Mannitol®.
- Medicines that reduce swelling in your airways are called inhaled steroids. An example of an inhaled steroid is Beclomethasone.
- Medicines that fight infections are antibiotics.

You can take inhaled medicines through an inhaler or with a spacer device. There are also medicines that are given through a nebuliser device, which turns a liquid into a mist and the mist is then breathed in. All these medicines need to be taken at the right time, so speak to your CF team if you are not sure when to take your inhaled medicines.

There is more information in '**Inhaled therapies for people with cystic fibrosis**', available from our **Helpline** and at **cysticfibrosis.org.uk/physioleaflets**

Cleaning your equipment

You must wash and dry the removable parts of the physiotherapy devices, including airway clearance devices and nebuliser equipment, after every use. Germs can live and grow in any part of the device that is not clean and dry. Each device may have slightly different ways to clean it, so make sure you read the instructions or speak to your CF physiotherapist to find out how to keep your device and equipment clean. Cleaning your physiotherapy equipment in hospital might be a little different to the way you do it at home. Speak with your physiotherapist or the ward staff to find out the best way to clean it while you are in hospital.

Cross-infection

You probably know that when you go to clinic or you are in hospital, you will be kept separate from other people with CF. This is because people with CF can have germs in their lungs that can grow into an infection. Although the bugs are not harmful to people who don't have CF, they can easily be passed from one person with CF to another person with CF and this can make the other person ill. Passing bacteria between people with CF is called cross-infection.

Cross-infection at hospital

Every CF centre will have their own policy to reduce the risks of cross-infection between their patients. This will help to keep you safe while in hospital. You will not have face-to-face contact or be in the same room at the same time as someone who also has CF.

Good hygiene can also help to prevent cross infection, such as:

- good handwashing
- covering your nose and mouth when coughing and sneezing
- putting tissues and sputum pots in the correct bins
- not sharing any physiotherapy or sports equipment.

You can find out more about cross-infection at **cysticfibrosis.org.uk/life-with-cystic-fibrosis/cross-infection**

Summary

There are many types of physiotherapy you can do to help clear your lungs. The one that suits you will depend on many things such as your age, lifestyle, the amount of mucus produced, and lung health. Teamwork and communication between you and your physiotherapist will help you to find the best airway clearance technique for you.

Further information

Find more information resources about living with cystic fibrosis at **cysticfibrosis.org.uk/information**.

Our Helpline is open 10am – 4pm Monday to Friday. It's available to anyone looking for information or support with any part of cystic fibrosis, a listening ear, or just to talk things through.

You can contact our friendly team by:

- phoning **0300 373 1000** or **020 3795 2184**

If you are worried about the cost of the call please let us know and we'll call you back.

- emailing **helpline@cysticfibrosis.org.uk**
- reaching out on all our social media channels

Visit **cysticfibrosis.org.uk/helpline** for more information.

We welcome your feedback on our resources.

You can also ask for this resource in large print or as a text file. Email **infoteam@cysticfibrosis.org.uk.**

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Cystic Fibrosis Trust

Cystic Fibrosis Trust is the charity uniting people to stop cystic fibrosis. Our community will improve care, speak out, support each other and fund vital research as we race towards effective treatments for all. We won't stop until everyone can live without the limits of cystic fibrosis.

Since 1964, we've supported people with cystic fibrosis to live longer, healthier lives – and we won't stop until everyone can live without limits imposed by CF.

cysticfibrosis.org.uk

Uniting for a life unlimited