Physiotherapy treatment in cystic fibrosis: airway clearance techniques
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Introduction

Looking after the chest and keeping the lungs clear is an extremely important part of the care of cystic fibrosis (CF). Over the years many different treatment techniques have been developed in order to help with this. This factsheet explains why it is important to keep the lungs clear and the various airway clearance techniques available. There are many other important aspects of physiotherapy in cystic fibrosis and a number of other factsheets are available from the Cystic Fibrosis Trust which deal with issues such as exercise, and physiotherapy in babies and children.

Written by S. Ammani Prasad, MCSP, Tamara Orska, MCSP, Kate Ferguson, MCSP, Penny Agent, MCSP and Mary Dodd, FCSP on behalf of the Association of Chartered Physiotherapists in Cystic Fibrosis. Updated by Elaine Dhouieb MCSP and Alison Gates MCSP.

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# Contents

How the lungs work 4  
How CF can affect the lungs 4  
Keeping the lungs clear 4  
How much airway clearance is needed? 5  

## Airway clearance techniques 5  
- Active cycle of breathing techniques 5  
- Postural drainage and percussion 6  
- Autogenic drainage 7  
- High frequency chest wall oscillation 8  
- Positive expiratory pressure (PEP) 8  
- Oscillating PEP devices 9  

## Other important aspects of chest care in CF 9  
- Physical activity 9  
- Inhalation therapy 10  
- Oxygen therapy 11  
- Non-invasive ventilation 11  
- Infection control 11  

Further information 12
How the lungs work

- The purpose of the lungs is to transfer oxygen from air we breathe into our blood. Our blood then transports the oxygen to the tissues and organs of our bodies, which need it to function.
- The lungs start at the back of the throat with the windpipe (trachea), which then divides into two airways (tubes), one going in to each lung.
- In each lung, the airways branch in a tree-like manner and get smaller in diameter. Eventually, they form small air sacs called alveoli at the edge of the chest.
- Intertwined with the smallest airways and air sacs are lots of tiny blood vessels called capillaries. Oxygen from the air we breathe in is transferred from the air sacs into the blood in the capillaries. At the same time waste gases are transferred from the blood into the airways and expelled as we breathe out.
- The lining of the airways continually produces mucus, to keep the airways moist and to trap dust and debris that may enter the lungs. The airways are also lined with microscopic hairs (cilia) which move the mucus up to the throat, allowing it to be cleared.

How cystic fibrosis can affect the lungs

- In cystic fibrosis the mucus produced by the lungs is stickier than normal, and CF lungs are more prone to infection. Infection in the lungs also leads to swelling of the airways, which makes the airway tubes narrower. The airways react to the swelling by producing more mucus.
- All of these factors can make it harder for people with cystic fibrosis to clear mucus from their lungs. Repeated infections cause irreversible damage to the lung tissue.

Keeping the lungs clear

Chest physiotherapy uses airway clearance techniques to help clear excess thick sticky mucus from the lungs. It is important to try and clear these secretions because they increase the problems with infection / inflammation and can block the smaller airways, which can result in the lungs not being able to work effectively. In addition to chest physiotherapy, antibiotics and inhaled medicines that help to open the airways and thin the mucus (mucolytics) are also used to help keep the chest clear.
How much airway clearance is needed?
The amount of treatment needed depends very much on the individual. The physiotherapist will help to decide how much treatment is needed and how often.

- Once the lungs become productive, airway clearance treatment is usually needed on a daily basis, and may be required up to three-four times a day if there is active infection. However, when well treatment will be needed less frequently – once or twice a day.
- The length of each treatment session will again vary according to need. Sessions may only be 10-15 minutes when there are only small amounts of secretions, however longer treatments will be necessary if there is a lot of sputum to be cleared.
- The type of treatment will alter over time, as various techniques are more suitable for different ages and at different stages of the condition process. The physiotherapist will continue to monitor this and change treatment as necessary. It is therefore very important that everyone with cystic fibrosis is regularly reviewed by a specialist physiotherapist.

Airway clearance techniques
The following section is a basic guide to the most commonly used physiotherapy techniques in the UK. Which technique is used for any one person depends on many factors and the physiotherapist will help to decide on the most suitable technique. It is very important that your airway clearance programme is monitored regularly by the physiotherapist. It is likely that you will use more than one method of treatment over time, as you get older and if there are changes to the way your lungs are affected. The techniques below are not discussed in any particular order, and no single technique has been shown to be more effective than the others.

Active cycle of breathing techniques
The active cycle of breathing techniques (ACBT) consists of a combination of:

Breathing control (BC)
This is a period of relaxed breathing. It is a very important part of this technique as it allows pauses for rest and helps avoid any tightening of the airways, which can make it difficult to clear secretions.

- The upper chest should remain relaxed with most of the movement occurring in the lower chest.
- The length of time spent on breathing control depends on the individual. The physiotherapist will advise you on timings.

Thoracic expansion exercises (TEE)
These are also known as deep breathing exercises. They help the lungs to expand more effectively and allow air to get behind any secretions so that they can then be “pushed” up the airways towards the mouth.

- The breath in should be slow and deep.
- At the end of the breath in, the breath is held for a few seconds.
- Breathing out is relaxed and "quiet".
- TEE are sometimes accompanied by percussion or vibrations (see below) but this may not be necessary.
The forced expiration technique (FET)
This technique consists of huffing and must be combined with breathing control. It helps to move secretions from the smaller to the larger airways from where they can be cleared more easily.

- A medium sized breath in is followed by a forced, but not violent, breath out (often called a huff). It should be made by squeezing the abdominal/tummy muscles while keeping the mouth and throat open – just as if trying to steam up a mirror with your breath.
- Huffing at different levels can move secretions in different parts of the lungs, your physiotherapist will teach you about this.
- One or two huffs are followed by a period of breathing control.
- Often a huff is not sufficient to clear the secretions completely and therefore it is necessary to cough afterwards. Coughing should not be very forced or go on for a long time. If secretions are not cleared after one or two coughs, then the ACBT should be resumed until they are higher up and can be cleared more easily.

The ACBT can be performed while sitting or in positions as described later. The cycle of different breathing techniques can be repeated until the chest is clear or a period of rest is needed. The physiotherapist will give advice regarding how many cycles are needed and the length of treatment.

Positioning
Your physiotherapist may advise using different positions or postural drainage during airway clearance.

Postural drainage
Gravity assisted positioning, often called postural drainage (PD), uses different body positions to help the drainage of secretions from particular areas of the lungs and also helps to increase the air movement or ventilation to different parts of the lungs. It is often used in conjunction with other techniques e.g. ACBT or percussion, and can be used in adults or children to treat more specific areas of the lung if this is necessary.

- In babies, your physiotherapist may suggest routine daily treatment consisting of about five minutes in each position, this may be one session in all four positions, or two sessions with two positions per session. In the presence of infection or increased secretions more frequent and longer treatment may be required. For those babies with no chest problems, routine daily treatment may not be necessary, but treatment will always be required whenever the baby has any signs of increased cough or infection.
- In older children and adults more specific postural drainage positions may be used. Again, the physiotherapist will teach and advise which of these are necessary for an individual using this technique. In some cases, “tipping” may be advised, which involves lying with the head lower than the chest in a tipped position. Some people find tipping uncomfortable as it makes them breathless, and some do not find it a particularly useful addition to their treatment.
There is some concern that using a head-down tipped position might increase the reflux of food from the stomach (particularly in babies). In view of this, many CF centres now use a modified type of PD where the head-down tip is not used. The specialist physiotherapist at your centre will advise you whether or not to use a head-down tip for physiotherapy.

If PD is being used it should always be undertaken at least one hour after food, to reduce the risk of reflux of stomach contents.

**Positioning**

Your physiotherapist may advise lying on either side, your front or back and sitting rather than specific postural drainage positions.

**Percussion and vibration**

**Percussion**

This technique is also known as chest clapping, and is used to help loosen secretions. It is usually combined with modified PD or PD and is also sometimes used during the Thoracic Expansion Exercise phase of the ACBT.

- To perform percussion a cupped hand is used to clap the chest firmly and rhythmically (over a layer of clothing or a towel). This is usually undertaken by a parent or carer although some people do it themselves.
- Parents are often taught this technique (combined with modified PD) when their baby is first diagnosed with cystic fibrosis. Many babies quite enjoy having percussion and it is not painful or uncomfortable.

**Vibrations/ Chest shaking**

This technique consists of several short rhythmical squeezes to the chest wall as the child/adult breathes out. Some people find this to be helpful in mobilising secretions. Like percussion, vibrations/chest shaking is sometimes used with TEE (in ACBT).

**Autogenic drainage**

Autogenic drainage (AD) is a series of breathing exercises which aims to use high flows of air to move secretions from the smaller airways, up into the larger airways so they can be cleared with as little effort as possible. AD consists of three different phases of breathing:

1. A mobilising phase
2. A collecting phase
3. A clearing phase

- By breathing at different levels, secretions can be moved from different parts of the airways. Secretions lower down in the airways are mobilised by breathing at a lower lung volumes (i.e. with only a small amount of air in the lungs). By then breathing at higher lung volumes (with a larger amount of air in the lungs), secretions which have mobilised higher up into the airways can then be moved and cleared.
- The breathing technique (whichever lung volume is being used) consists of a slow breath in followed by a pause, where the breath is then held for a few seconds. This allows air to fill the lungs more effectively improving ventilation).
The breath out is active but in a “sighing manner” and should not be forced. As secretions move higher, crackles should be heard or felt.

Once the secretions have been moved to a higher level they can be cleared with a gentle huff.

The technique can then be continued until the chest feels clear or as clear as possible.

This technique should be taught by a physiotherapist who has been trained in AD.

AD can be used in children and adults, some physiotherapists also use an assisted form of AD in babies.

High Frequency Chest Wall Oscillation

HFCWO (sometimes know as Vest therapy) has been used in the USA for many years. It is now available to purchase in the UK.

The equipment consists of an electric air compressor which is connected to an inflatable jacket.

The compressor sends pulses of air into the jacket, which vibrates the chest wall and lungs within. These vibrations help to move sputum within the lungs.

Although this equipment is now available in the UK, it is generally not widely used. Vest therapy is a useful technique in some patients, however it has not been shown to be of significantly more benefit than any other techniques.

Positive Expiratory Pressure

Positive expiratory pressure (PEP) is a technique that applies a “back pressure” to the airways during the breath out. This helps to open up airways and get air behind secretions to help move them higher up the airway.

A number of PEP devices are available, some have a mask and some a mouthpiece.

The PEP device gives a small degree of resistance to the breath out and this resistance splints open the airways.

Treatment is usually done in sitting with the elbows supported, however with some of the devices a postural drainage position may also be incorporated.

After 8-12 breaths through the PEP device, 1 or 2 huffs or a huff and a cough, with breathing control, are used to clear the secretions.

The PEP cycle is repeated until the chest feels clear.

The PEP system needs to be reviewed regularly at clinic visits, as the amount of resistance used may need to be altered from time to time to ensure that an adequate pressure is being used.

Sometimes a slightly different technique of PEP (high pressure PEP) is used where the last breath out through the device is forced and long to produce a greater back pressure. This is a specialist technique which needs careful assessment and teaching.

Bubble PEP is another modification of PEP and is very useful in young children. It uses exactly the same action as PEP described above, but instead of using a mask or mouthpiece the child blows out through a length of plastic tubing placed in a bottle of water and soap to produce lots of bubbles. This is a fun way of getting small children to do PEP therapy.
Oscillating positive expiratory pressure devices (Oscillating PEP)
These devices combine vibration of the airways with PEP. Several different devices are available for this type of treatment and the physiotherapist will help to decide which one is most suitable for each patient. The devices include:

- The Flutter
- The Acapella
- The Cornet

The Flutter
The Flutter is a small pipe shaped device which contains a metal ball in a cone. During the breath out the ball moves up and down in the cone. This interrupts the flow of air and gives an intermittent “back pressure” to the airways as well as causing them to vibrate.

- Flutter treatment is usually done in sitting.
- The angle at which the Flutter is held will change the area where the vibrations are felt and so it should be held at an angle where maximum vibrations are felt within the chest.
- The physiotherapist will advice on the best way to use the flutter, but usually the treatment combines cycles of normal breaths followed by one or two big and active breaths out through the device.

The Cornet
The Cornet consists of a curved hard plastic tube within which sits a soft flexible rubber tube. It works in a very similar way as the flutter, producing a vibration and PEP effect in the airways. The degree of PEP and vibration can be altered by changing the twist in the rubber tube.

- The Cornet can be used in any treatment position and like the flutter a combination of breathing techniques are used to help to move and clear secretions.

The Acapella
The Acapella device also works in a very similar way to the flutter. It consists of a plastic outer shell, inside which is a lever and magnets. The lever action and the attraction between the magnets during the breath out provide the vibration and PEP. The degree of PEP and vibration can be altered by a dial at the end of the device.

- Treatment can take place in any position and is very similar to that of the Flutter.

Thorough, daily cleaning and drying is vital for all devices as unwashed devices can harbour bacteria. There will be individual cleaning instructions for different devices.

Other important aspects of chest care in cystic fibrosis
Physical activity
Physical exercise is an essential part of the management of cystic fibrosis. As well as having beneficial effects on fitness levels, muscles, bone structure and posture it also complements airway clearance and helps to keep the chest clear. A separate factsheet that offers advice and explains
the benefits of exercise is available from the Cystic Fibrosis Trust (see back page for contact details). Some children and adults use a combination of exercise and airway clearance for their physiotherapy.

Inhalation therapy
A range of inhaled drugs may be prescribed as part of the daily management of cystic fibrosis. These may include bronchodilators (to open the airways), nebulised antibiotics (to help fight infection), mucolytics (to thin sputum e.g. DNase [Pulmozyme]), expectorants (to clear sputum e.g. hypertonic saline) and inhaled steroids (see the Cystic Fibrosis Trust factsheet Nebuliser therapy in cystic fibrosis). The order in which these medications are taken is usually assessed on an individual basis and the physiotherapist will advise you about this. Bronchodilators are usually taken before airway clearance and antibiotics after airway clearance. The exact timing for taking DNase (Pulmozyme) does vary but in general, it should always be taken at least 30 minutes before physiotherapy (but preferably longer, sometimes it is taken just after the morning physiotherapy session so that it can act in the lungs for a longer period of time). However it is recommended that DNase is NOT taken within 30 minutes of an inhaled antibiotic, to prevent the medication being inactivated.

It is important that any medications that are inhaled are able to get in to the airways in order to be effective. These medicines are given using either an inhaler or via a nebuliser system.

Inhaler devices
Various devices are available for a range of drugs. Some devices are combined with a spacer to improve delivery. The drugs commonly given via inhaler are bronchodilators and inhaled steroids. However, in future antibiotics will be available via this method.

Nebuliser systems
Traditional nebuliser systems consist of an air compressor with a separate nebuliser unit in which the liquid drug is placed. Different types of drug require separate nebuliser chambers. There have been many developments in nebuliser systems and there are now a wide variety of devices available. The latest of these are small, hand held, battery driven, efficient in terms of delivery time and quiet. These devices often combine an air compressor and nebuliser system in one, resulting in a single hand-held device. Drugs commonly nebulised are bronchodilators, antibiotics, DNase and other mucolytics (to thin sputum).

Whichever device is prescribed, the CF nurse or physiotherapist should teach you how to use it properly. However, some general points to remember are:

- The order in which medications are taken needs to be assessed individually.
- Babies and small children may need to use masks with devices, although mouthpieces should be introduced as early as possible. A mask may need to be used for nebulised antibiotics if there is severe sinus disease or after lung/heart-lung transplant.
- Thorough, daily cleaning and drying is vital for all devices as unwashed devices can harbour bacteria or the device may become blocked and not work effectively.
- Different nebuliser chambers may be needed for different drugs.
Oxygen therapy
As lung problems worsen and the lungs become less efficient at taking in enough oxygen, it may be necessary for some people to use oxygen therapy. This will happen at different times for different people, and at first oxygen may only be required during an infection, during exercise or for use at night time. Later it may be needed for longer periods. Oxygen is an extremely helpful therapy and it can make a huge difference to how well you feel and how much you can do during the day.

The CF team will monitor things very closely and use various tests to decide if and when oxygen needs to be started (these include exercise tests, and sleep studies). Oxygen can be given via a variety of masks or nasal cannulae. Small oxygen cylinders and other portable oxygen devices can be provided so that it is still relatively easy to get out and about. In the home, an oxygen concentrator machine may be installed to provide a regular supply of oxygen.

Non-invasive ventilation
With worsening lung disease, some people may need to use non-invasive ventilation (NIV) to help with their breathing. This is normally used when there is a build up of carbon dioxide (waste gas) and oxygen levels are low. Sometimes NIV is used overnight or at other times it is used to assist with airway clearance. In a very few cases, people need to breath with the assistance of NIV for the majority of the day and night. NIV can be used with a mouthpiece, nose or face-mask. Whenever it is started, a full explanation is essential and the machine settings need to be checked regularly. Even when using NIV, it is possible to talk, eat and drink although care should be taken when eating and drinking while using the machine.

Infection control
Every specialist CF centre and clinic will have their own Infection Control Policy to minimize the risk of cross infection between you and others with cystic fibrosis and to prevent the spread of CF organisms. Some centres and clinics segregate their outpatient clinics. This might mean that you are not moved around the clinic to see the various members of the team but are shown to one room where all the team will come to see you, or the clinic may segregate according to the organism grown in your sputum.

Special attention should be paid at all times by you and your physiotherapist when handling sputum during airway clearance and exercise. These treatments will always be performed in a separate area away from other people with cystic fibrosis. Personal hygiene e.g. handwashing, covering your nose and mouth when coughing and sneezing and disposing of tissues and sputum pots in appropriate waste bins, all help to prevent cross infection. Any equipment given to you will be for your own personal use and should not be shared with others. The CF Trust also provides guidelines about the potential risks away from the hospital and these are available directly from the Trust. If you have any concerns or worries you should speak to your CF consultant, specialist nurse and physiotherapist.
Further information

The Cystic Fibrosis Trust provides information about cystic fibrosis through our factsheets, leaflets and other publications.

Most of our publications are available through our helpline and can be downloaded from our website or ordered using our online publications order form. Visit cysticfibrosis.org.uk/publications.

The Cystic Fibrosis Trust helpline can help you with a range of issues, no matter how big or small. Our trained staff can provide a listening ear, practical advice, welfare/benefits information or direct you to other sources of support. The helpline can be contacted on 0300 373 1000 or helpline@cysticfibrosis.org.uk and is open Monday to Friday, 9am – 5pm.

Calls to 0300 numbers cost no more than 5p per minute from a standard BT residential landline. Charges from other landlines and mobile networks may vary, but will be no more than a standard geographic call and are included in all inclusive minutes and discount schemes. If you are worried about the cost of the call please let us know and we’ll call you back.

You can also find more information at our website cysticfibrosis.org.uk.

Cystic Fibrosis Trust
2nd Floor One Aldgate
London
EC3N 1RE
020 3795 1555

cysticfibrosis.org.uk

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