# Physiotherapy for people with Cystic Fibrosis: from infant to adult

**Supported by the International Physiotherapy Group for Cystic Fibrosis**

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# 1

# Introduction

Dear Reader,

Established cystic fibrosis (CF) lung disease is characterized by reduced mucociliary clearance, airway plugging, recurrent infections and chronic inflammation. Areas not ventilated soon become hypoxic, which allows growth of anaerobic micro-organisms. The progressing airways obstruction results in impaired ventilation distribution, gas exchange and breathing mechanics leading to musculoskeletal complications. Daily physiotherapy aimed at ventilating all parts of the lungs and compensating for impaired mucociliary clearance is essential to minimize lung disease and preserve lung function, to encourage good posture and avoid musculoskeletal complications, and to maintain endurance and allow a good quality of life.

In the past, the primary aim of physiotherapy for people with CF was to clear excessive secretions and thus reduce symptoms. The term ”physiotherapy” is today used in a much wider sense. Modern physiotherapy in CF is a combination of inhalation therapy, airway clearance techniques (ACT 's), physical education/exercise and ongoing education about the disease and its treatment. The physiotherapist should be involved in recording the evaluation of patients, the instructions given to them, quality control and professional development. The role of the physiotherapist is, in co-operation with the patient and family, to tailor an individualized, reasonable, effective and efficient physiotherapy regimen. This should take into account all relevant physical and psychosocial factors. Modern physiotherapy is primarily preventative and has to be incorporated into each patient's daily routine in a lifetime perspective. This can be achieved only by tailoring a time-efficient treatment that places the least possible burden on the patient or his/her family and makes compliance with the treatment possible.

The proportion of people with CF who are middle-aged is increasing and this trend is likely to continue. Many are married and working ,and other ”adult” issues occur. Problems of the older person should be recognised and addressed appropriately.

This booklet aims to be a useful tool and reference document for all physiotherapists involved in the delivery of care to people diagnosed with cystic fibrosis from birth and throughout life. It is based on scientific evidence but where this is not available, a best practice consensus has been outlined.

On behalf of the IPG/CF,

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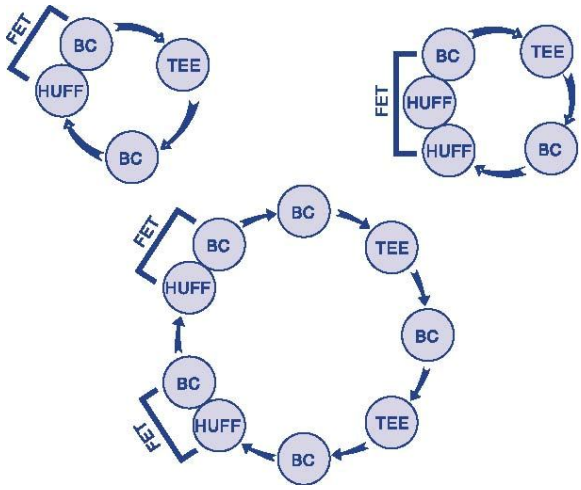
# 2

# Airway Clearance Techniques

## Active Cycle of Breathing Techniques

The active cycle of breathing techniques (ACBT) is used to mobilize and clear excess bronchial secretions (Pryor *et al* 1979). The components of the ACBT are breathing control, thoracic expansion exercises and the forced expiration technique.

**Active Cycle of Breathing Techniques**




Key: BC - *breathing control;* TEE - *thoracic expansion exercise:* FET - *forced expiration technique*

The regimen is flexible, should be adapted to suit the individual and can be used by the young, the elderly, the fit and the sick. It was first documented by Thompson and Thompson in 1968. The ACBT can be used independent of an assistant or with an assistant and either sitting or in a gravity assisted position.

*Breathing Control* (BC) is the resting period between the more active parts of the cycle. It is breathing around tidal volume, at the individual's own rate and depth. The person is encouraged to relax the upper chest and shoulders and to use the lower chest, diaphragmatic pattern of breathing as they are able. This should minimise any bronchoconstriction and should be continued until the person feels ready to use either the thoracic expansion exercises or the huffing of the forced expiration technique.

*Thoracic Expansion Exercises* (TEE) are deep breaths emphasising inspiration. Inspiration is active and usually combined with a three-second, end-inspiratory hold before a passive, relaxed and unforced expiration. With an increase in lung volume the resistance to air flow via the collateral channels (Menkes & Traystman 1977) is reduced. Mobilization of secretions can be facilitated by air passing through these channels and behind secretions. The 'hold' should permit asynchronous ventilation as air flows more slowly into diseased and obstructed regions than into healthy, unobstructed areas – *Pendelluft* flow (Mead *et al* 1970). *'Airflow is essential for airway clearance'* (Lapin 2002). Up to three thoracic expansion exercises are followed by breathing control and may be combined with chest shaking or chest clapping. Chest clapping and shaking appear to be of benefit to some people, but are not required by others.

Upper, middle or lower thoracic expansion may be encouraged (Tucker *et al* 1999).

*The Forced Expiration Technique* (FET) is a combination of one or two forced expirations (huffs) and periods of breathing control. Huffing to low lung volumes should assist in loosening and mobilising excess bronchial secretions from smaller peripheral airways to larger central airways. When secretions reach the upper airways, a huff or cough from a high lung volume can be used to clear them. *'Forced expiratory manoeuvres are probably the most effective part of chest physiotherapy'* (van der Schans 1997). The length of the huff and force of contraction of the muscles of expiration should be altered to optimise clearance of secretions (Pryor & Prasad 2008) by maximising air flow. During a forced expiratory manoeuvre (for example a huff) there is compression of the airway downstream (towards the mouth) of the equal pressure point (West 2004). This squeezing action (which moves peripherally with decreasing lung volume) together with the increase in air speed, as air flows through the narrowed segment, facilitate the movement of secretions along the airway. In addition the airway wall oscillates as the airway narrows (Freitag *et al* 1989).

The ACBT can be introduced as huffing games from the age of about two years, and from the age of eight or nine years the child can begin to take some responsibility for his/her own treatment, gradually becoming independent. The ACBT should never be uncomfortable or exhausting and the huff should never be violent. It can be used in any position according to the requirements of the individual. The sitting position is often effective and adherence to treatment is frequently better than with other positions. In some people, as identified on assessment, other gravity assisted positions may be indicated. It has been shown that the horizontal, side lying position is as effective as the head down tipped position and preferred by individuals (Cecins *et al* 1999).

The flexibility of the regimen (the number of deep breaths, the number of huffs and the length of the periods of breathing control) is demonstrated in the figure. The ACBT is repeated until the huff becomes dry sounding and non-productive or it is time for a rest. If more than one position is needed, two positions are probably enough for one treatment session. The total treatment time is usually between ten and thirty minutes. The physiotherapist and/or patient determine by assessment the most suitable regimen, the position(s) required for treatment, the length of time and the number of treatments in a day. This will change within a treatment, from treatment to treatment and during acute exacerbations of pulmonary infection compared with periods of clinical stability.

Studies using the ACBT have shown it to be an effective and efficient technique for the mobilization and clearance of secretions (Pryor *et al* 1979, Wilson *et al* 1995). It is not further improved by the adjuncts of positive expiratory pressure - PEP (Hofmeyr *et al* 1986), Flutter® (Pryor *et al* 1994, Pike *et al* 1999), mechanical percussion (Pryor *et al* 1981) or high frequency chest wall oscillation (Osman *et al* 2008). An improvement in lung function following the instigation of the ACBT (Webber *et al* 1986) has been demonstrated, and hypoxaemia is neither caused nor increased (Pryor *et al* 1990). In the long term (one year) the ACBT, PEP and oscillating PEP have been shown to be equivalent in airway clearance (Pryor *et al* 2006).

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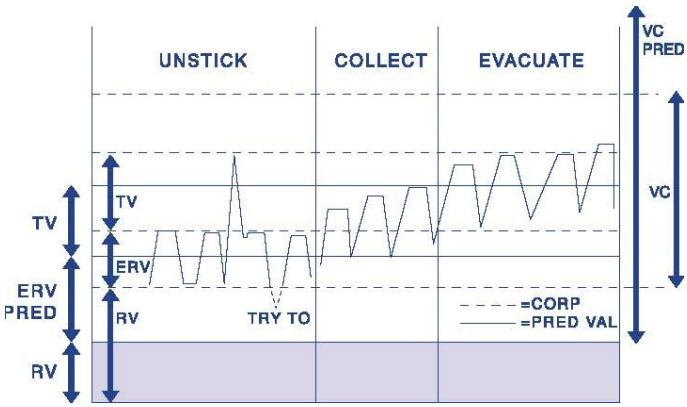
## Autogenic Drainage (AD)

### "The flow and breathing level modulation concept"

Autogenic Drainage is an airway clearance technique based on basic physics, fluid dynamics, pulmonary anatomy, respiratory physiology and breathing mechanics.
The mechanism of mucus clearance rests on two different systems: the effect of the ciliary clearance and the effect of shearing forces induced by the airflow. This last phenomenon can be compared to the effect of erosion. The higher the velocity of the medium, the stronger the erosive effect.
To create the necessary shearing forces to clear the bronchi from secretions, it is essential to modulate the inspiratory and expiratory airflow.
During inspiration, the linear velocity of the airflow may not be too high to avoid an inhomogeneous filling of the lungs and a lead back of the secretions.
During exhalation the optimal shearing forces, induced by the linear airflow velocity, must be localized there where the secretions are. To localize the secretions the three feedback signals (auditive, tactile and proprioceptive) are used. By modulating the breathing level within the vital capacity and the expiratory muscle force, the optimal airflow will be obtained at the precise level of the bronchial "funnel", the targeted airway generations, where the secretions are. The realized intra-thoracic pressure generated by the expiratory muscles may not exceed the stability of the airways.

**Breathing in Autogenic Drainage**




The whole airway clearance process in AD is based upon an active or passive assisted autogenic drainage (AAD) modulation of the air flow and lung volume level breathing.
The positioning of the patient and the shape correction of the respiratory pump can induce an increased regional ventilation to optimize the clearance of particular lung parts.
Taking special care of spastic and/or swollen airways is a must in all ACT's. The bronchial resistance must be normalized if possible and the secretions must be made easier to remove by means of drugs, or special devices.
Before clearing the lower airways it is also evident to clear the upper airways, without causing appreciable increase of the airway resistance. Correct dosage of the expiratory force increases only slightly the bronchial resistance, keeps the alveolar gas compression low thus optimizing the elastic recoil force of the alveoli, and does not compress the airways in an early stage. It also lightens the expiratory efforts and decreases the appearance of paradoxical breathing movements. For some reason the stimulation to cough is less intense which allows one to inhibit and postpone the cough more easily.

## The AD Technique in Practice

**Breathing IN**

1. Clear the upper airways (nose and throat)
2. Optimize the shape of the respiratory pump
3. Choose a breath-stimulating and airway clearance enhancing position
4. Breathe IN slowly through the nose keeping the upper airway open to optimize the even distribution of air, to avoid paradoxical movements and to get more air behind the mucous plugs
5. Hold the breathing movement for approximately 2 to 4 seconds during which the UAW are kept open, thus improving the even filling of all lung parts. The breathing movement has to be stopped in its 3 dimensions!
6. Depending on where the mucus is, in peripheral, middle-large or large airways, the functional tidal volume needed is ventilated at low-, mid- or high lung volume level.

**Breathing OUT**

1. Breathe the chosen functional tidal volume OUT preferably through the nose. If a drop in velocity does occur or, if one wants to hear the bronchial noises in a better way, breathe OUT through the mouth. In this case always keep the upper airways (glottis, throat, mouth) open.
2. The expiratory force must be modulated in such a way that the expiratory airflow reaches the highest possible velocity without causing an early airway compression.
3. Breathing Out correctly, the mucus can be heard distinctively. Putting a hand on the upper chest, one can also feel the mucus vibrating. The frequency of these vibrations indicate where the mucus is localized in the bronchial tree. This FEEDBACK makes it possible and easy to adjust the breathing pattern and the appropriate expiratory airflow modulation.

**Successive breathing cycles**

1. Repeat the cycle.
2. Continue to use the same breathing pattern until the mucus starts to collect by moving upwards. If this occurs, the level of the functional tidal volume is gradually raised. Thus, the breathing evolves from a lower to a higher lung volume level. Finally, the collected mucus plug arrives in the trachea from where it can be evacuated by a high lung volume huff or a similar cough. Cough must be postponed as long as possible to collect larger mucous pieces which are easier to remove.

**Frequency and indications**

The duration and number of the AD sessions depends on the total amount and the viscosity of the secretions. Experienced patients drain their lungs more quickly than others. Drainage should always be done thoroughly. The principles of AD can be used in obstructive and restrictive pulmonary diseases. Active participation is preferred but not essential. The modulation of the breathing pattern keeps the respiratory pump mobile and the respiratory muscles in a good length-tension ratio.

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## Assisted Autogenic Drainage (AAD)

Assisted Autogenic Drainage is based upon the principles of Autogenic Drainage and used in infants and non-cooperative patients.

By modulating, manually or/and by using elastic straps, the functional breathing level within the vital capacity, the optimal airflow velocity will be obtained at the targeted airway generations, where secretions have been identified. AAD is carried out in a gentle and progressive way, using the patient's breathing pattern and stabilizing the infant's abdominal wall to avoid paradoxical movements.

To guide the breathing of the patient towards the desired lung volume level, striving to find the normal physiological breathing level, a gentle increase of manual pressure on the chest during each **inspiration** is performed. In fact the hands gradually restrict the inspiratory level to stimulate the patient to exhale slightly more than the previous breathing cycle.
During expiration we follow gently the breathing movement of the patient. No thoracic compression or excessive force is performed, which could lead to a resisting response by the patient.
Feedback plays a key-roll, feeling or hearing the secretions move while avoiding any early or abnormal airway compression or closure.
Wait for the spontaneous cough. Patience is a must in this kind of technique!

To optimize the shape of the respiratory pump, allowing the respiratory muscles to function more properly and efficiently, semi-elastic belts will be used in addition of the physiotherapists hands. The positioning of the patient and the shape correction of the respiratory pump can induce an increased regional ventilation to optimize the clearance of particular lung parts.
Before starting any pulmonary treatment the upper airways must be cleared.
"Preparing" the lungs before the airway clearance is very important to lower or normalize the bronchial resistance and to liquefy the secretions.

Assisted Autogenic Drainage can be combined with bouncing, a gentle up-and-down movement on a physio ball, to relax the patient and to enhance the expiratory air velocity. The patient sitting upright is correctly supported, avoiding a slumped sitting position which may in turn predispose to gastro-oesophageal reflux (GOR) during treatment.

No provocation of GOR has been associated with AAD, bouncing or the combination of both.

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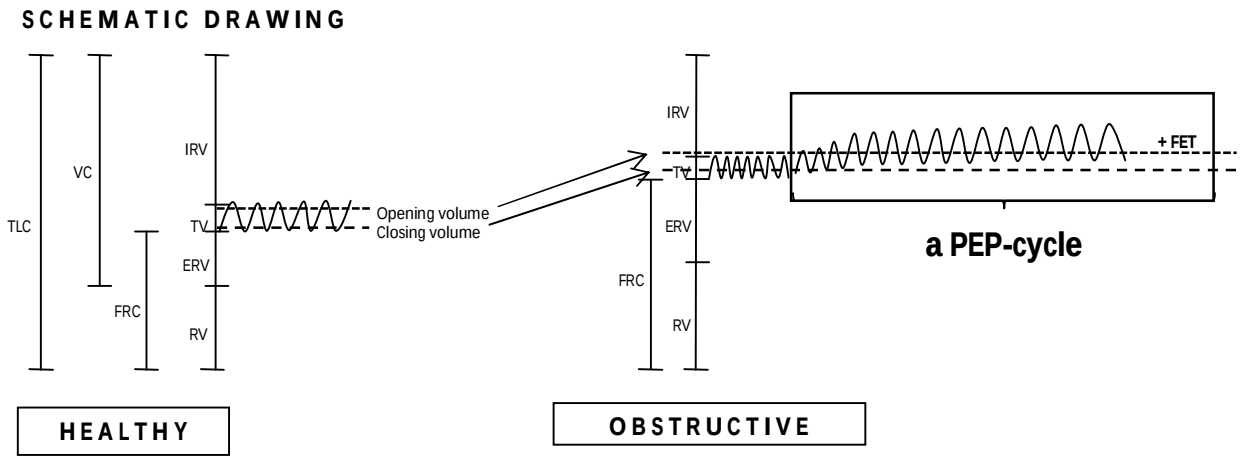
## Positive Expiratory Pressure (PEP)

Expiratory resistance breathing can be used for many different physiological purposes. When the target is to recruit closed or clogged peripheral lung volumes by mobilising, transporting and evacuating secretions in spontaneously breathing patients, it is by definition combined with the forced expiration technique (FET)( see: Active Cycle of Breathing Techniques). This airway clearance technique is called PEP.

**Aim**

To obtain a temporary increased functional residual capacity (FRC), that allows the tidal volume (TV) to reach above the opening volume for the otherwise closed or clogged airways, see the schematic drawing. Due to the interdependence between the airways, the lung parenchyma and the elastic recoil of the lung tissue at this temporarily increased FRC level, closed airways should open and collateral ventilation should increase. The air in the recruited lung volumes can then be used with the help of a technique such as FET to mobilise, transport and evacuate secretions.

**PEP**




**PEP-mask and instructions**

The airway clearance technique PEP was developed during the late 1970's – early 1980's using a mask with a one-way valve to which an expiratory orifice resistor can be attached (Astra Tech). A pressure manometer may be inserted between the valve and the resistor. Patients are instructed to sit comfortably forward leaning with elbows on a table holding the mask tightly over mouth and nose, and to breathe using slightly active TV breaths. Babies do the treatment in a backwards leaning "sitting" position on an arm of a parent, where the baby's head is supported by the upper arm while the other hand holds the mask firmly on the baby's face. The resistor that gives a stable pressure level of 10-20 cm $H_2O$ during the middle of expiration is the one which should be selected. By simultaneous listening and looking at the breathing pattern, the expected change towards an increased FRC level should be perceived. Adequate instructions to obtain the expected aims are essential.

**Treatment**

Each treatment cycle consists of 12-15 breaths with only slightly active TV expirations, followed by one or more cycles of FET, see the schematic drawing above. The number of treatment cycles within a treatment session and the treatment frequency is adapted to individual need. A treatment cycle should end with recruiting lung volumes by breathing towards the resistance to finish with the airways as open as possible.

**Considerations**

There are many expiratory resistance devices on the market, and more will come. In some of the devices resistance is flow regulated, in others it is pressure regulated. This influences the expiratory pressure achieved and the breathing pattern in different ways. A physiologic strategy is essential when using these devices. The ability to analyse the immediate response during treatment is of utmost importance.

If the expiratory pressure achieved is due to a flow regulated resistance as the PEP-mask, the instructions to the patients are of great importance as well as the individual feed-back during treatment. The TV breaths should be only slightly active while achieving a mid-expiratory pressure of 10-20 cm $H_2O$. Patients should be taught how to recognize a temporary increase in FRC. The increase in FRC is of utmost importance, due to the physiological elastic recoil theory that the technique is based on.
If the expired volumes are too big the increased FRC level may not appear, or FRC may even decrease. When using the PEP-mask, a manometer may be inserted between the expiratory valve and the resistor, to measure the mid-expiratory pressure and to identify when a stable mid-expiratory pressure is achieved. The use of the manometer is to find the optimal resistor, there is no reason to use the manometer during each treatment session and the patient does not need to see it at all, as that may affect the breathing pattern in an undesired way (expiration becoming too active).

The size of the resistor selected, to obtain beneficial effects, is dependent on lung volume, breathing frequency and to a certain extent capability to take instructions. The size of the resistor needs to be changed as conditions change.
PEP was originally developed and described to be used in sitting, but can also be used in horizontal positions. PEP can be applied to patients with severe lung disease, who may need assisted ventilation (Bi-level PAP) with high inspiratory pressures to attain the physiological aim to reach above closing volume.

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## Hi-PEP

**Technique**

The High-Pressure technique of PEP-mask physiotherapy employs forced expiratory manoeuvres against the PEP-mask's expiratory resistor for mobilizing and transporting intrabronchial secretions. The instrument used for this technique is the same as the one described in the previous chapter, albeit equipped with another manometer for monitoring higher pressures. Therapy is performed with the patient seated, elbows resting on a table, and shoulders moved close to the neck to cover and support the lung apices. PEP breathing for eight to ten cycles is done using moderately increased tidal breathing, then the patient inhales to total lung capacity and performs a forced expiratory manoeuvre against the stenosis. The consequent mobilization of secretions usually results in coughing from low lung volumes. After expectorating sputum, the same sequence of breathing manoeuvres is repeated until no more sputum is produced. Care must be taken not to terminate these forceful expirations before reaching residual volume; sustained expiratory pressures achieved usually range between 40 and 100 cms of $H_2O$. The dimension of the expiratory resistor and the pressure developed against it is determined individually by a spirometer-assisted method. For this purpose the outlet of the PEP-mask is connected to a spirometer, and the patient performs forced expiratory vital capacity manoeuvres through a series of resistors with different internal diameters. The resistance for daily therapy is chosen on the basis of maximal homogeneity in the expiratory behaviour of different lung units, as determined by the shape of the flow–volume curve.

**Physiology**

Expiratory resistive loading effects a progressive homogenization of the expiratory behaviour of different lung units. This is especially important for people with CF with widespread bronchiectasis. Their disseminated bronchial instability lesions will tend to occlude the bronchial airway as soon as it is subjected to any positive expiratory pressure (coughing, some chest physiotherapy techniques, hyperventilation during exercise). This effectively interrupts airflow from the dependent lung units; they will remain inflated by trapped gas while alveolar regions behind less damaged airways will properly contribute to expiratory volumes and flows. From a physiological perspective, the equal pressure point will get stuck for the major part of a forced expiration in the airway instability lesion, while properly moving upstream elsewhere. Consequently, the most diseased airways are hardly incorporated in the compressed downstream segment, thus missing the most effective mechanism for clearance of the more central intrathoracic airways.

This mechanical handicap, which is typical for advanced airway disease in cystic fibrosis, is compensated by exhaling against a correctly dimensioned expiratory resistor. In the first part of a forced expiration, the backpressure from the stenosis effects a completely homogenized slow expiratory evacuation of all lung units. When monitored by the recording of a flow-volume–curve, this effect is expressed by a plateau formation in the expiratory tracing. Lung units behind bronchiectatic lesions evacuate to the same extent as those behind less diseased airways. Finally the loss of lung volume effects a decrease of static-elastic recoil pressure to such an extent, that the plateau formation cannot be maintained any longer; the equal pressure point, previously arrested in the resistor, starts to move upstream via the trachea towards the bronchial periphery. This important terminal phase of the expiratory high-pressure PEP-mask clearance manoeuvre effects a dynamic compression of all bronchial airways. In contrast to an unloaded expiration, however, the compression wave moves over the diseased airway at a much lower local lung volume. This again means less distension by dilated parenchyma; the necessary subtle balance between compression wave and bronchial calibre is effectively re-established, and mucus clearance from the more diseased lung units is possible again.

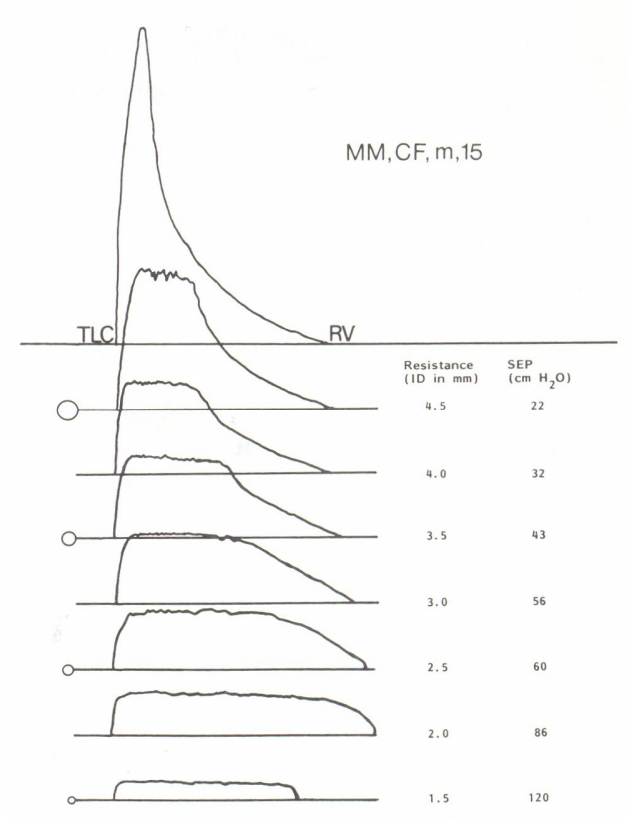
**The manoeuvre consists of two important parts:**

**a) Mobilisation phase**

The effects of High-PEP-mask therapy can be explained by increased collateral airflow to underventilated regions; air expired from there should mobilize obstructing secretions. In addition, a forced expiration against a marked resistive load will squeeze Pendelluft from hyperinflated into unobstructed and atelectatic lung units. Mobilization of mucous plugs is supported by back pressure-effected dilation of airways.

**b) Transportation phase**

A progressive incorporation of the peripheral airways into the compressed downstream segment is a prerequisite for efficacy. Incomplete manoeuvres, either caused by the choice of an inappropriate resistor or by incorrectly performed technique should be avoided.




TLC= total lung capacity; RV=residual volume; ID=internal diameter; SEP=sustained expiratory pressure

Figure : Oberwaldner B, Evans JC, Zach MS. Forced expirations against a variable resistance: a new chest physiotherapy method in cystic fibrosis. Pediatr Pulmonol 1986;2:358-67.

**Figure:**
A series of MEFV-curves blown by a CF-patient through differently sized resistors. Uppermost curve is MEFV-curve without resistor; internal diameters of subsequently used resistors are given on the right side of the curve. Sustained expiratory pressure increases stepwise with raised resistive loads.
Note the gradually decreasing curvilinearity of descending part of MEFV-curve; complete homogeneisation of expiration is achieved by resistors with an ID between 3.0 and 2.0 mm. Also note resistor-effected plateau formation at increasingly low expiratory flow rates. With a resistor of 1.5 mm ID expiratory loading is increased to an extent that patient terminates forced expiration before exhaling to RV (to be avoided!). A resistor with an ID of 2.5 mm is chosen for further Hi-PEP treatment.

The positive effects of the high-pressure PEP-mask therapy, however, do not come for free. One price to pay is reduced expiratory airflow velocity. Even in an unloaded 'free' forced expiration, expiratory airflow velocity decreases rapidly towards the bronchial periphery, as due to the rapid increase of the total bronchial cross-sectional area.

It follows that the reduction of expiratory airspeed that is effected by the resistor, decreases in importance towards the periphery.

The net balance is that shearing forces of the expiratory air flow are traded against the re-established effects of dynamic expiratory bronchial compression. Most likely, the latter is more reliable for peripheral bronchial clearance than the former.

The other price to pay in Hi-PEP is the development of relatively high and sustained expiratory pressures. This calls for a dedicated and energy-consuming muscular effort from the patient.
It follows that this chest physiotherapy technique is not to be recommended for self-treatment in exhausted patients, who find it hard to develop such expiratory pressures. Rather, the technique offers itself for well-trained patients in a good nutritional condition, who aim to clearing their airways effectively in a minimum of time and are willing to invest in this with maximum effort. From a more general care giving perspective, Hi-PEP is thus an important component of a modern CF-management that is characterized by a psychological groundswell of activity and dedication. In- and expiratory muscle training as a side effect of this technique, comes free and contributes to a good body image.

## Hi-PEP as a passive physiotherapy technique

The technique, as described above, is self-applied and thus requires a well trained and actively cooperating person. It follows that Hi-PEP can be taught to patients up from the age of about four years.
With modifications, however, Hi-PEP may also be applied to babies and exhausted patients who are unable to cooperate actively. A forceful expiratory effort of the patient can be replaced by a therapist´s skilfully performed chest compression and the resulting forced expiration may then be modified by a resistor as described above. In older patients, this may require the coordinated efforts of two therapists, but in babies, an experienced therapist can usually manage to compress the chest and hold a PEP-mask in place simultaneously. For treating babies, who will not reliably inspire to TLC and have only low tidal volumes, small PEP-masks with minimal dead space are mandatory.

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## Oscillating PEP

The Flutter VRP1 (VRP1 Desitin/Scandipharm Flutter VarioRaw SA) is a pocket device, approved by the US Food and Drug Administration for use in 1994. It is used to improve pulmonary ventilation and to facilitate expectoration (Althaus *et al* 1989).The oscillating positive expiratory pressure is reported to prevent premature closure of the bronchi, to loosen secretions and improves mobilisation of sputum (detaching mucus from the airway walls) which may be cleared using the forced expiration technique (Thompson *et al* 2002) .
The device comprises a mouthpiece (fig. 1a), a plastic cone (fig. 1b), a steel ball (fig. 1c) and a perforated cover (fig. 1d). During exhalation through the device, the tracheobronchial tree undergoes internal vibrations together with repeated variations of the exhaled airflow against the resistance (positive expiratory pressure PEP) and oscillations of the endobronchial pressure (oscillating pressure). Oscillating PEP is most commonly performed with the Flutter device or the Acapella device.

The Acapella device uses a counterweighted plug and magnet to create air flow oscillation. The performance of the Acapella is not gravity dependent (i.e. dependent on device orientation) and may be easier for some patients to use, particularly at low expiratory flows (Volsko *et al* 2003). With devices such as the Flutter, the individual exhales through the instrument and pressure builds up in the airways until the mechanism in the Flutter moves and gas escapes. The occlusion and opening of the gas pathway produces oscillations in pressure which are transmitted through the tracheobronchial tree.

The Flutter VRP1 has two main characteristics:
1. It generates an automatically controlled oscillating positive pressure. The patient is thus protected against a collapse of the airways, as well as against any prolonged hyper-pressure which could occur should the instructions for use not be followed and exhalations be forced repeatedly.
2. It enables a modulation of the pressure and airflow oscillation frequency. By tuning this frequency to his/her own ventilatory abilities, the patient induces maximal vibrations of the bronchial walls which promote clearance of the obstructed airways.

Modulation of the flow and pressure oscillations are obtained as follows, applying the same approach as for the Autogenic Drainage: The patient exhales into the Flutter device; during exhalation the steel ball inside the device bounces, causing vibratory obstruction to air flow, which oscillates both the pressure and air flow only during exhalation.

The Flutter generates PEP in the range 18 to 35 cm $H_2O$ and the angle at which the device is held determines the oscillation frequency (usually between 6 and 26 Hz) (Gumery *et al* 2002) and the patient's expiratory effort determines the pressure. The combination of PEP and oscillation that forms the basis of the technique is thought to break up and reduce the viscosity of secretions.

As with other PEP techniques, the patient repeats the maneuver for 10 –15 breaths (fig. 4) followed by mucus expectoration, several huffs without the device or when not controlled, coughs. This cycle is repeated 3–4 times, resulting in a 15–20 minute airway clearance session.

The patient should sit comfortably (fig. 2), hold the Flutter VRP1 horizontally, then take a slight deep breath, put the mouthpiece of the device into his/her mouth, hold his/her breath for 2 to 3 seconds (permitting the inhaled air to be more evenly distributed in the lungs and behind the mucus in the very small airways), close his/her lips tightly around the mouthpiece and breathe out normally and deeply (fig. 4), keeping his/her cheeks flat and hard, using the method of unforced abdominal exhalation while relaxing the muscles of his/her upper chest.
Repeat breathing in through the nose and out again into the Flutter. On successive attempts, the patient may find it necessary to move the Flutter VRP1 (fig. 3) slightly upwards (higher pressure and frequency, a positive incline and a large airflow result in an increase in expiratory pressure (Brooks *et al* 2002)) or downwards (lower pressure and frequency) by a few degrees until he/she can feel the full effects of the vibrations at the abdominal level during the first stage of the exhalation process. It is not necessary to complete a full exhalation each time when breathing out through the Flutter VRP1 (fig. 4 & 5).
During each 10 to 15 breath cycle coughing should be suppressed until the last exhalation, which should be done at about twice the speed of a normal exhalation. This should automatically bring up a cough followed by mucus expectoration. The frequency and duration of each session should be adapted to the needs (airway clearance efficiency) of each patient.

Short-term studies with CF patients have shown the Flutter to be similar to postural drainage and percussion or PEP ( Konstan *et al* 1994; Gondor *et al* 1999; van Winden *et al* 1998; Homnick *et al* 1998).
A randomised crossover study of patients with stable CF compared 4 weeks of treatment with the Flutter with autogenic drainage. No differences were found in sputum weight or lung function after a single session with either method at the end of the treatment period, but sputum viscoelasticity was significantly reduced with the Flutter (Apps *et al* 1998).
Konstan *et al*. reported that up to three times more sputum was produced with the Flutter than with postural drainage in similar subjects. In contrast, again in patients with stable CF, Pryor *et al* found that significantly more sputum was produced with the active cycle of breathing techniques (ACBT) than with the Flutter in individual supervised sessions, but similar sputum weights were produced with both methods over 24 hours.
Two studies compared the Flutter with percussion, vibration, and postural drainage by a physiotherapist in children with CF admitted to hospital with an acute exacerbation and found no significant differences in lung function or exercise tolerance: The first study demonstrated that patients using the Flutter device had better pulmonary function after one week of therapy and similar improvement in pulmonary function and exercise tolerance compared to CPT after two weeks of therapy, suggesting that Flutter valve therapy is an acceptable alternative to standard CPT during in hospital care of patients with CF (Gondor *et al* 1999).The second study demonstrated that the Flutter device appears to be safe, efficacious, and cost effective for CF inpatients capable of undertaking this type of therapy (Homnick *et al* 1998).
One long-term study (more than a year) in children with CF compared the Flutter with the positive expiratory pressure mask and found a greater decline in forced vital capacity (FVC), increased hospital admissions, and increased antibiotic use with the Flutter (McIlwaine *et al* 2001).
Eaton *et al* in a randomized prospective study evaluated the acute efficacy, acceptability and tolerability of three airway clearance techniques in non-cystic fibrosis (non-CF) bronchiectasis: Flutter, active cycle of breathing techniques (ACBT) and ACBT with postural drainage (ACBT-PD) were evaluated in random order over a week in 36 patients (mean age 62 years, range 33-83), with stable non-CF bronchiectasis. All three techniques were well accepted and tolerated. Patient preference was 16 (44%) for Flutter, eight (22%) ACBT and 12 (33%) for ACBT-PD, but ACBT-PD proved superior in terms of acute efficacy.
A randomised crossover study was performed by Thompson *et al* in 17 stable patients with non-cystic fibrosis bronchiectasis at home, in which four weeks of daily ACBT were compared with four weeks of daily physiotherapy with the Flutter device. He concludes that the daily use of the Flutter device in the home is as effective as ACBT in patients with non-cystic fibrosis bronchiectasis and has a high level of patient acceptability. For the time being, it seems best to choose the method that matches the patient's abilities and preference in order to improve compliance (or satisfaction (Oermann *et al* 2000)) with the physiotherapy regimen. Randomized controlled multicenter investigations, with adequate patient numbers and descriptions of the treatments and measurement tools are needed before we change patient care. The physiotherapist must consider which physiotherapy regimens are more effective for individual patients rather than any one technique being the most effective for all patients with cystic fibrosis.

**Figures**

Fig. 1 : detail inside

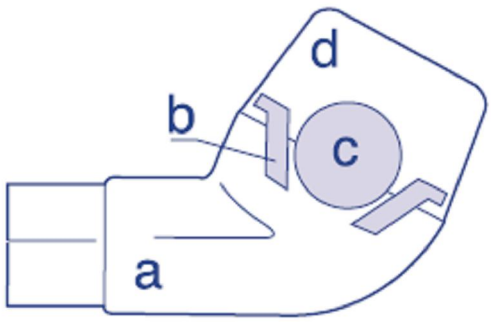


Fig. 2 : position for use

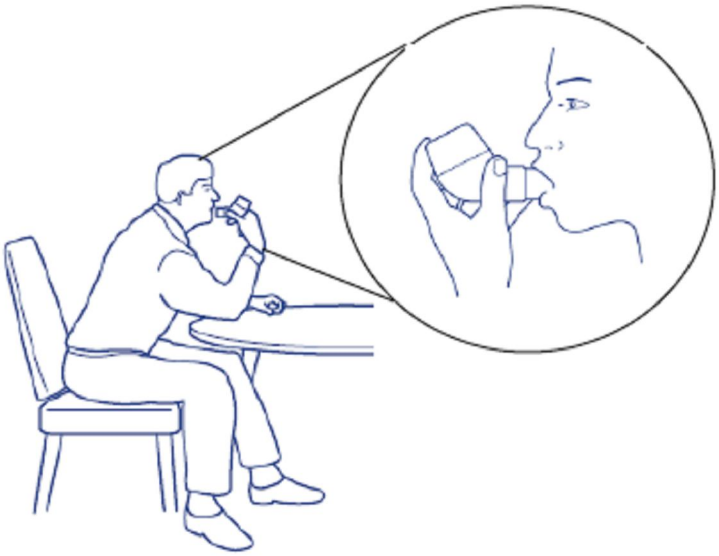


Fig.3 : moving the Flutter up or down

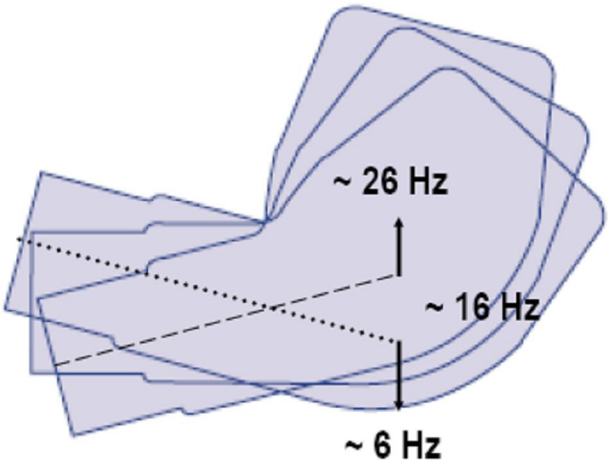


Fig.4 : schematic way of breathing with the Flutter (Volume – Time)

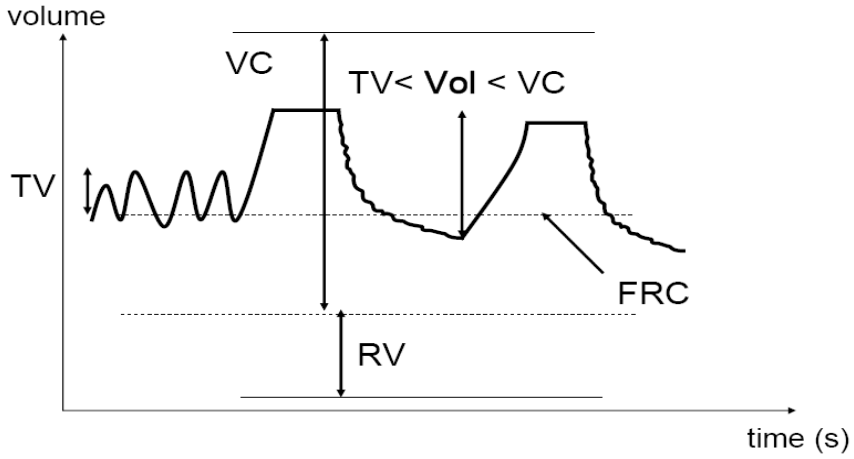
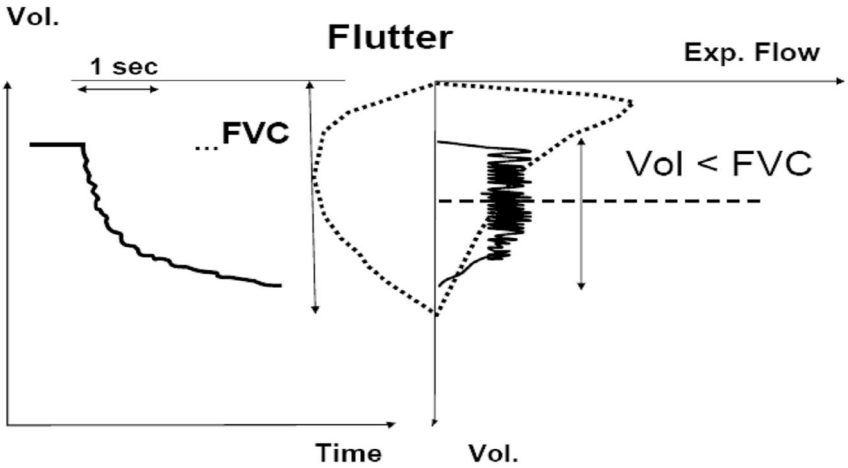



Fig.5 : schematic way of breathing with the Flutter (Flow – Volume)

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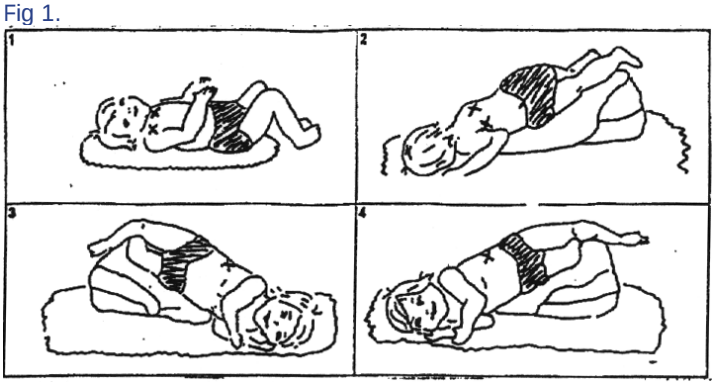
## Postural drainage and Percussion

Postural drainage and percussion was first introduced for the treatment of cystic fibrosis (CF) in the 1950's (Matthews *et al* 1964) and remained the cornerstone of therapy until the 1980's. Traditonally, postural drainage has consisted of placing the patient in a position which allows gravity to assist in draining mucus from the periphery of the lungs centrally. Usually, between six and twelve postural drainage positions are used, depending upon which lobes or segments of the lungs are to be drained (fig. 1). Babies were placed in postural drainage positions over the caregiver's lap. As the child got older, pillows or a postural drainage board were substituted. Percussion is used as an adjunct to postural drainage. While in each postural drainage position, the patient usually has his/her chest percussed for between three to ten minutes. This is followed with deep breathing exercises, vibration on expiration and huffing. Treatment is divided into two or three daily treatment sessions.

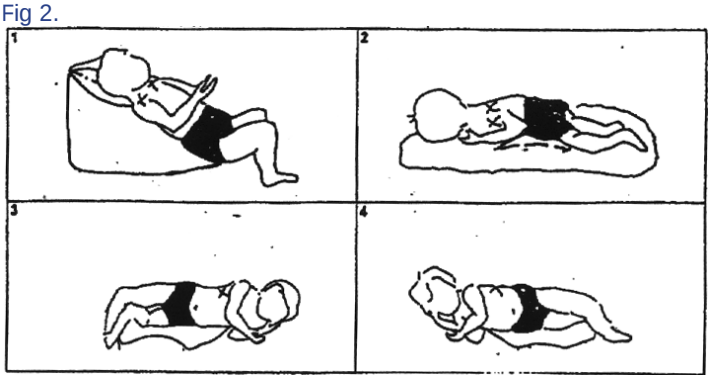
To-day, in many countries, modified postural drainage positions are the accepted method of treatment Modifications include, the elimination of head down positions with the lower lobes being drained in a horizontal plane or the use of a slight tip only (Fig 2.)

Fig 1.




Standard postural drainage positions

Fig 2.




Modified postural drainage positions

**Effectiveness**

Early studies have shown postural drainage and percussion to be an effective means of clearing excessivebronchial secretions in patients with CF (Desmond *et al* 1983; Reisman *et al* 1988). However, it is time consuming, often requiring the assistance of a second person and is uncomfortable for the patient. As a result adherence with this treatment regimen is very low (Passero *et al* 1981). The use of modified non-tipped positions was first introduced after Button found that in patients who have gastroesophageal reflux (GER), the reflux was often aggravated by placing them in a head down postural drainage position, and could potentially lead to aspiration (Button *et al* 1997) . Research, also by Button *et al* suggest that the use of modified postural drainage positions in the baby has no long-term detrimental effects on the patient and may even improve long-term outcomes (Button *et al* 2003). In addition, patients with moderate or severe lung disease often experience oxygen desaturation while receiving postural drainage with percussion (Mc Donnell *et al* 1986).

**Modifications**

As the awareness of the GER in CF patients has increased, many CF centres are now advocating the use of modified postural drainage positions either for all their patients or for those with demonstrated GER.

Due to the adverse effects and poor adherence with postural drainage and percussion, it has largely been replaced in many countries by other modalities of physiotherapy outlined in this booklet.

Today, it is primarily used in the treatment of babies with CF who are unable to co-operate fully enough to perform other types of physiotherapy. Some countries prefer to use modified Autogenic Drainage or PEP Mask with babies. However, more often the child is changed over to one of these other modalities as she/he has learned how to huff effectively.

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# 3

# Physical Exercise

Physical exercise has, justifiably, become an important part of the physiotherapy treatment. Whether physical exercise is to be used in order to maintain function, prevent dysfunction or rehabilitate what has been lost must be an active decision in the treatment approach for all physiotherapists in each CF-center.

### Aims

The aims of including physical exercise from the very beginning are to; develop age adequate fitness and to stay physically fit, to maintain working capacity, endurance, muscular strength and mobility. A well preserved chest wall mobility is a prerequisite for effective airway clearance therapy. A normally developed and retained bone mineral density (BMD) and a good posture are necessary as patients get older to avoid vertebral and fragility fractures and musculoskeletal pain. An immediate aim for the young patient is to maintain a similar level of exercise to that of his/her peers and friends. This is likely to influence self esteem and the type of every-day life activities. The aim of rehabilitating dysfunction is to strive for regaining what has been lost.

### In general

Physical exercise does not necessarily improve a decreased lung function, but with the help of physical exercise fitness can be maintained even though lung function is poor. Preserving physical fitness can be great fun for small children and at least enjoyable/bearable for most adolescents and adults. Intense physical rehabilitation programs limited to a couple of weeks can give very good results, at least temporarily, but often lack follow-up. Such rehabilitation programs or camps can be scheduled at certain intervals or repeated when fitness has been lost, but a lot of time is tied up during these intense rehabilitation programs and they may be difficult for patients to carry out themselves. Such programs can be used to regain muscle strength, endurance and working capacity to a great extent, but lost mobility is more difficult to retrieve. Maintaining is easier than trying to regain what has been lost. However, whether to choose maintenance or rehabilitation programs depends on many factors, not least individual and cultural.

The way working capacity and endurance training is carried out is the same as for healthy people without CF. The first choice would be weight bearing exercises in order to influence BMD at the same time. Physical exercise in CF is not only working capacity and endurance training, although this is the focus in most studies. Muscle strengthening exercises are of great interest, especially the postural muscles such as back extensors and scapulae fixators. Mobility exercises ought to emphasize the spine, neck and shoulders, but also involve other parts of the body, as in people without CF. The very important mobility exercises for costosternal and costovertebral joints may be included every day, preferably as a part of the airway clearance therapy involving lung volumes between total lung capacity and residual volume.

The physical exercise programs and types of exercises used should be individually tailored, taking into account age, nutritional condition, personality, interests, surroundings, physical capability and current pulmonary condition. The programs should be up-dated when necessary, depending on change in physical capacity, surroundings, interests and adherence. The exercises used can vary from one session to another, or be the same depending on what suits the individual. In order to increase adherence, the program is designed and changes are made together with the patient, as an agreement rather than a prescription.

**Preventative physical exercise can be carried out either**

1. As the basis of the daily airway clearance therapy

   The physiological effects from physical exercise on lung volumes, flows and breathing pattern will recruite otherwise closed or clogged airways and an enhanced respiratory pump may be utilized. Cycles of moderately physical exercises can be utilized as the basis of airway clearance therapy, if interspersed with FET or AD, assisted AD for infants and toddlers (FET, AD, assisted AD, see previous paragraphs). When exercises are carried out in different positions (such as upright, side lying, supine or prone) the effects of gravity on regional functional residual capacity and regional ventilation can be utilized. Some patients prefer to carry out physical exercise before airway clearance therapy, due to their experience that secretions loosen more easily when doing it that way. A treatment session should always end with physical exercise or at least thoracic expansion exercises, to finish the session with the airways as open as possible.

   Advantages: Carrying out physical exercise as the basis of the daily airway clearance therapy constitutes time efficient therapy. For infants, toddlers and for many youngsters it is a stimulating airway clearance alternative that allows siblings, parents and friends to take an active part. The different exercises are easily replaced with new, when needed or for variety. The exercises used during each treatment cycle, morning or afternoon treatment sessions, should be varied and this should help to maintain adherence to the treatment.

2. In addition to the airway clearance therapy

   Physical exercise programs added to airway clearance therapy should contain working capacity and endurance training, muscle strengthening and mobility exercises, according to individual aims. Working capacity, endurance and muscle strengthening exercises are preferably carried out 2-3 times/week, in accordance with general recommendations. Mobility exercises are preferably carried out daily (designed as a physiologically structured part of the airway clearance therapy).

   Most CF-centers recommend that severely ill patients need supplemental oxygen during physical exercise to maintain normal $SpO_2$.

   Advantages: Adding the physical exercise some days/week may be easier compared with performing it as a part of the daily airway clearance therapy, especially if there is lack of space at home or in close surroundings. In the more severely ill patients the airway clearance sessions may be less time efficient if based on physical exercise, due to the longer pauses needed to catch up with breath.

**Considerations**

Patients who are malnourished or have decreasing weight trends should not do working capacity, endurance or muscle strengthening exercises until the nutritional situation is under control. This also applies to patients with fever or an acute pulmonary exacerbation of infection. However, mobility exercises can always be undertaken.

Some patients need pre-medication with a bronchodilator before carrying out working capacity and endurance training. However, patients with instable airways may demonstrate an increase in airflow obstruction with a bronchodilator (reference). Spirometry before and after physical exercise with and without bronchodilator and/or physical exercise tests with and without pre-medication should be used to assess the response.

Most CF centres will have a policy for when supplemental oxygen should be offered to patients who desaturate during physical exercise. Based on clinical experience and/ or evidence, the use of oxygen is meant to minimise potential negative effects on the heart and/or to maximize the effects of muscle strengthening exercises.

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# 4

# Inhalation Therapy

Inhalation therapy is often an important component of the treatment in CF. Under optimal conditions it is the physiotherapist who administers the inhalations in conjunction with airway clearance therapy. A sufficient intrapulmonary deposited dose and an even deposition pattern should be the aim. Although we know more about influencing factors nowadays, we still know too little about how to achieve optimal results for the single individual, especially in the very young ones and the severely ill.All the dependent factors can differ to such a great extent. Many results in publised studies are based upon experimental studies including results from a "robot- lung" doing 12 breaths/minute with a tidal volume of 500-800 ml and a inspiratory-expiratory ratio of 1:1. Care must be taken in extrapolating these results to the person with CF.

When about to start inhalation therapy there are important things to consider:

- Different drugs and inhalation strategy
- Optimal mode of aerosol delivery device for a specific drug and each inidividual, dose of drug, aerosol quality and deposition pattern
- Inhalation technique, positioning?

**1. Different drugs for use, inhalation strategy**

**a. Bronchodilator**: A number of people with CF have an asthmatic component which may be an indication to use a bronchodilator, especially at certain times of the year. Otherwise bronchodilators may be needed as pre-medication to physiotherapy, to other kinds of inhaled drugs or to physical exercise that may cause a broncho-constriction. The purpose is to avoid broncho-constriction. The target tissue, the bronchial muscles, has a blood supply and bronchodilators are easily absorbed. This means that the drug, when absorbed into the blood vessels in the bigger airways, can be transported to the smaller ones by the vessels supplying the airway tree. Appropriate reversibility tests should be carried out regularly, as response is likely to change with a multifactorial progressive disease and new drugs become available. Various bronchodilators can be used, most often a beta-2 agonists, but if patients have a tendency to suffer from lower limb muscle spasm (often in gastrocnemius and soleus during physical activity), an anti-cholinergic bronchodilator should be tried.

**b. Inhaled steroids:** The indication is to decrease or control mucosal inflammation. The target tissue, the mucosa, has a blood supply, but steroids are not easily absorbed. An even deposition in all airway generations is the aim, to maximise the effect of treatment. The effect is not immediate and daily treatment, over a period of time, is necessary to achieve maximal benefit. Whether steroids should be inhaled before or after airway clearance therapy is not clear The aim of treatment and amount of secretions may give an indication. If treatment is to prevent mucosal inflammation in a patient with small amounts of secretions, the steroid may be inhaled before airway clearance therapy. If bronchial secretions are copious, clearance of secretions first may lead to a more even distribution of the aerosol and improve peripheral deposition. The possibility to inhale the dose over more than one breath should facilitate deposition, especially with severel ventilationabnormalities.
The mouth should be rinsed thoroughly, with water, after the inhalation to minimise oral side effects.

**c. Antimicrobials:** The indication is to eradicate bacteria in the airway lumen and/or to control chronic infection in all parts and generations of the airway tree. The target is the mucus within the airway lumen and on the airway walls and it does not have a blood supply. Therefore an even deposition pattern is strived for to achieve maximal effect of the treatment. Whether antibiotics should be inhaled before or after airway clearance therapy is not clear. The amount of secretions and the type of airway clearance technique may govern the strategy. An improved condition for a more peripheral and even aerosol distribution may be achieved when large amounts of secretions are cleared first. Antibiotics are and will be available pulverulent, to be administered with the help of a powder inhaler.

The possibility to inhale the dosage in more than one breath may facilitate intrapulmonary, peripheral and a more even deposition, especially in small children and the severely ill. Some antibiotics that are easily absorbed may give undesired side effects. The systemic concentration should be monitored by blood analysis and the individual dose adjusted as necessary.
Gargle the throat, with water, after the inhalation to minimise local side effects.

**d. Agents to decrease mucus viscosity or "unstick" mucus from the mucosa:** These range from isotonic saline to CF specific mucolytics. Some act immediately while others have a delayed effect (30-60 minutes). Slow acting agents make the timing of inhalation and airway clearance more difficult, especially when used every day at home. Fast acting mucolytics or unstickers can be used in conjunction or interspersed with the airway clearance cycles.

**2. Optimal mode of aerosol delivery device for a specific drug and each individua,l definition of dosage, aerosol quality and deposition pattern**

The 'dose' delivered is often thought to be that of the dose prescribed, but the amount of the medication that actually reaches the lungs and peripheral airways is very different. If the expected effects do not result, the tendency is to increase the prescribed dose which may lead only to increased side-effects. Instead, the mode of aerosol delivery should be questioned and the inhalation technique evaluated and optimised. There are several important factors which affect the intrapulmonary deposited proportion of the nominal dose and the deposition pattern within the respiratory tract. Some of the more important factors are 1) proportion of the nominal dose that leaves the inhalation device as aerosol, 2) aerosol quality expressed as mass median aerodynamic diameter (MMAD), 3) proportion of aerosolised volume that is accessible during the inspiratory phase of the breathing cycle, 4) delivery flow velocity, 5) patient´s inspiratory volumes, 6) ventilation distribution, and 7) inspiratory flow velocity. When recommending or prescribing a devic,e it is important to consider these factors and the needs of the individual patient. To achieve the best results from the therapy, each patient needs to learn how to handle the device and an optimal inhalation technique. The physiotherapist needs to spend time educating the patients not only at the start of the therapy, but also at the time of follow-ups. During follow-ups the physiotherapist should evaluate whether the prescribed mode of aerosol delivery is still optimal for the individual, or if a better result may be achieved with another type of device. When patients are seen in open care clinic or when admitted to hospital the patient should bring their device, or use the same kind of device, and should assemble and inhale as at home. This is to allow the physiotherapist to evaluate the patient's handling skills and inhalation technique, in addition to assessing and advising the cleaning routine for the device.

**a. pressurised Metered Dose Inhaler (pMDI)**:
There are several different kinds of pMDI´s on the market. Most pMDI´s are activated manually at the start of the inspiration. This coordination is difficult for many patients. Flow-triggered pMDI´s have been introduced, but the trigger may results in a delay of the delivery of the aerosol, of importance for patients with small inspiratory vital capacities. The pMDI´s have a high aerosol delivery flow velocity, with the majority of aerosol being deposited in the pharynx. A spacer can be added to the manually activated pMDI´s, which improves the intrapulmonary deposition significantly since it reduces the coordination problem and the reservoir decreases the problem with the high delivery flow velocity. Some of the spacers are very bulky and difficult to transport, many have an electro-static inside which absorbs a lot of the delivered dose, some have valves placed unfavourably resulting in big dead volumes, some are difficult to clean while others are not supposed to be cleaned. New more user-friendly models appear on the market continuously.

**b. Dry Powder Inhaler (DPI):**
There are several different kinds of DPI´s on the market. Some need to be loaded with e.g. a capsule which is punctured before each dose is administered, while others are preloaded with up to 200 doses or more. With DPI´s an optimal inspiratory flow (inspiratory muscle capacity and sufficient inspiratory vital capacity) is required to set the powder free for inhalation. Loading or preparing the DPI for inhalation may be complicated and even impossible for some patients. The most difficult part of the inhalation process is to distinguish between inspiration and expiration. Breathing out through the device is not recommended as it would cause condensation in the thin passages. Patients need to learn to breathe out beside the DPI, hold the breath while it is placed into the mouth, then to breathe in with an optimal flow velocity, hold the breath while taking the device out of the mouth and then to breathe out beside the DPI. Some patients, although having practised a lot, have difficulty distinguishing between inspiration and expiration.

**c. Nebuliser system:**

There are three groups of nebuliser systems based on different technologies: the jet, the ultrasonic and the mesh. A nebuliser system should be considered as a nebuliser with a driving source. This is especially important with jet nebulisers as the nebuliser performance is dependent on the working pressure delivered from the compressor while aerosolising. The manufacturer should be able to answer questions on the optimal working pressure needed for their nebuliser system (and for in hospital use, the gas flow required to drive the nebuliser) to achieve the specified MMAD for a specific drug. If one component of the nebuliser system is changed, the performance and overall quality of drug delivery also changes. The jet technique is the oldest. The jet nebuliser systems are still the most frequently used and often the cheapest on the market.. Disposable jet nebulisers are not recommended for long-term home use. In ultrasonic nebulisers the aerosol quality is dependent on the frequency of crystal oscillation. Unfortunately the oscillating crystal also generates warmth and this may influence the stability of some drugs,if the crystal is not isolated. The mesh technique is not new, but is the most recent. The aerosolising part is more fragile than expected, at least in some of the mesh devices, and probably dependent on the type and quality of metal, the drugs inhaled, cleaning routines and on the calcium content of the water in which the mesh is cleaned. When new the mesh nebuliser is very fast. The mesh plate requires careful cleaning to maintain this good performance. The mesh plate can be replaced, but a cost is incurred.

Many nebuliser systems are available on the market. All have advantages and disadvantages and many are complicated to handle. Trends point towards the development of new drugs prescribed with a specific nebuliser system, in order to guarantee optimal delivery of the intrapulmonary dose. However, for patients prescribed several drugs for nebulisation this can be very complicated in every day use.

When using a nebuliser system for inhalation therapy, a lot of time each day needs to be spent on handling and cleaning the nebuliser. A dirty nebuliser will decrease the performance of the nebuliser and is a source of infection. Studies have indicated that micro-organisms can lodge in nebulisers, providing a potential source of contamination of the airways. Adequate written cleaning and disinfection guidelines for home nebulisation need to be discussed with and given to each patient, but they must be realistic.

**3. Inhalation technique, positioning?**

The achievement of an optimally even intrapulmonary deposition pattern is of great importance especially when inhaling drugs that are not easily absorbed and aimed at targets within the airway lumen without a blood supply. An effective inhalation technique is essential to achieve an optimal deposition pattern. One of the more important factors is a low inspiratory flow velocity in order to avoid impaction in the pharynx and central airways. During inhalation therapy, the patient should be sitting upright and use relaxed abdominal breathing. Using pMDI's, preferably with a spacer, requires a slow deep inspiration with a breath hold, repeated a couple of times especially if the inspiratory vital capacity is small. Using DPI's requires a deep inspiration with a high inspiratory flow velocity to free the dosage. Using a nebuliser system offers a chance to vary the breathing pattern during inhalation, inspire from different lung volume levels and to utilise the effects of gravity on regional ventilation by inhaling in side-lying or supine, which some nebulisers allow.

Inspiring from different lung volume levels and utilising the effects of gravity is not possible when each dose is released and administered by only one inspiration. But when using DPI´s and pMDI´s a dosage may be administered by more than one release. However, positioning makes the inhalation sessions more complicated and the patient must be motivated to do it, otherwise recommending it is not defensible since it may have a negative influence on adherence.

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# 5

# Non-invasive Ventilation

**Non-Invasive Ventilation in Cystic Fibrosis**

The management of severe lung disease is an important component of care for patients with CF. With life expectancy now extending well into adulthood and many patients choosing to be listed for lung transplantation, maintaining optimal function in patients with end-stage lung disease is crucial. Non-invasive ventilation (NIV) may be used to enhance airway clearance and exercise in patients with severe disease and has an expanding place in the management of acute and chronic respiratory failure.

**Non-invasive ventilation as an adjunct to airway clearance**

Effective performance of airway clearance techniques may be challenging for some patients with advanced disease due to increased ventilatory demand, alterations in gas exchange and dyspnoea (Holland *et al* 2003; Cecins *et al* 1999). Non-invasive ventilation unloads the respiratory muscles during airway clearance in both adults and children with CF, resulting in decreased dyspnea and improved oxygen saturation during treatment. However, the amount of sputum expectorated is not different to a session performed without NIV( Holland *et al* 2003; Fauroux *et al* 1999; Placidi *et al* 2006). The use of NIV to assist airway clearance has been described in both acutely unwell and stable patients with CF (Holland *et al* 2003: Fauroux *et al* 1999) .

***Clinical considerations for NIV during airway clearance***

**Patient selection:** The benefits of NIV during airway clearance are greatest in those with poor respiratory muscle strength or severe airflow obstruction (Holland *et al* 2003: Fauroux *et al* 1999). The therapist's assessment of such patients should include careful observation of respiratory work at rest, the level of resting dyspnoea and effectiveness of forced expirations. Where it is evident that dyspnoea and fatigue are limiting the effectiveness of a patient's usual airway clearance technique, NIV-assisted airway clearance can be considered. The clinical characteristics of patients who may benefit from this technique are in Table 1.

**Mode of ventilation**: Pressure-cycled modes of ventilation (inspiratory pressure support and bilevel ventilation) are effective modes of NIV for use during airway clearance. A back-up rate should not be used during airway clearance sessions as it may interfere with expectoration. Volume-cycled modes, where a pre-set tidal volume is used, are less leak-tolerant and therefore unlikely to prove suitable for use during airway clearance, where leaks will inevitably occur during expectoration.

**Maximise pressure support**: The aim of NIV during airway clearance is to provide respiratory muscle unloading. The goal should be to provide as much pressure support as possible by increasing IPAP as tolerated. Delivered inspiratory positive airway pressure (IPAP) has been reported as ranging from 10 to 20 cm $H_2O$, with more severely hyperinflated patients requiring higher levels of pressure support (Holland *et al* 2003).

**Maximise expiratory flow**: High levels of expiratory pressure may interfere with forced expirations and coughing. Therefore if expiratory positive airway pressure (EPAP) is used, it should be kept at relatively low levels during these manoeuvres, with reported values of 4 to 5 cm $H_2O$.

**Expectoration and NIV**: Expectoration should ideally occur without removing the mask in order to maintain positive pressure in the airway. For this reason a nasal mask is usually preferred. However, some patients report that forced expirations and coughing are easier with an oronasal mask, presumably due to splinting of collapsible airways by positive pressure during expiration. Patients using an oronasal mask may require more assistance with expectoration.

**Use of oxygen**: In hypoxic patients, NIV increases oxygenation due to improvements in alveolar ventilation. However, some patients may still require entrainment of oxygen to achieve acceptable oxyhaemoglobin saturation. During airway clearance the preferred site of entrainment is close to the ventilator, as attachments at mask level are frequently dislodged during expectoration. Supplemental oxygen flow rates should be titrated according to oximeter readings.

**Airway clearance techniques under NIV**: The well-established principles of the active cycle of breathing techniques or forced expiration technique can be used whilst NIV is in place. Coaching will be required so that the patient is able to perform forced expirations, cough and expectorate independently, even in patients who are accomplished at performing standard airway clearance.

**Exercise and non-invasive ventilation**

Maintaining and improving exercise capacity poses significant challenges in patients with advanced lung disease, where high work of breathing may limit exercise duration or patients may be NIV-dependent.

Continuous positive airway pressure (CPAP) has been shown to improve exercise endurance, reduce dyspnoea and improve oxygenation during exercise in patients with severe lung disease and hyperinflation ( Henke *et al* 1993). These data indicate that NIV may have a place in reducing the work of breathing during exercise in patients with advanced lung disease.

In clinical practice, NIV is used to assist exercise training in patients with CF where severe dyspnoea limits training duration, or in patients who are severely unwell and bridging to transplantation. In NIV-dependent patients training should not be started until arterial blood gases have stabilised. It is important to allow time for the patient to acclimatise to NIV at rest before beginning exercise training, particularly if NIV has not previously been used. The aim of NIV during exercise is to reduce work of breathing and therefore the patient should be given as much pressure support as possible by increasing IPAP as tolerated. This will often require titration during exercise. A full face mask may be required during exercise to prevent mouth leak.

**Non-invasive ventilation for respiratory failure – physiotherapy considerations**

Non-invasive ventilation may be used to stabilise patients with CF who have acute or chronic respiratory failure. In acutely unwell patients with severe lung disease, NIV results in reduced $PaCO_2$, respiratory rate and dyspnoea. Longer-term outcomes of NIV for chronic respiratory failure may include improvements in daytime $PaCO_2$, reduction in the number of days spent in hospital and improvement in symptoms. Although NIV does not reverse the respiratory deterioration inherent in end-stage disease, it may allow the patient to be stabilised for long enough for donor lungs to become available for transplantation. Physiotherapy treatment regimens may require modification during this time, especially in the NIV-dependent patient.

***Airway clearance****:* effective airway clearance remains an important priority in the NIV-dependent patient. Close liaison with the care team is required to determine whether patients are stable enough to take breaks from NIV to perform airway clearance, or whether airway clearance should be performed under NIV as previously described in this chapter.

***Humidification:*** Non-invasive ventilation delivers air at high flow rates and with low relative humidity, which may overwhelm the capacity of the upper airway mucosa to warm and humidify inspired air ( Holland *et al* 2007). Due to the risks associated with drying of secretions and sputum retention, heated humidification is recommended when NIV is used in CF.

***Inhaled therapies:*** Consideration must therefore be given to the route of administration of these therapies in the patient using NIV. For some patients, breaks from NIV may be appropriate, at which time inhaled therapies can be given via their usual route. For NIV-dependent patients however, administration of inhaled therapies during NIV will be required. Connectors for metered dose inhalers (MDIs) are available; alternatively, a T-piece connector for nebulisation can be used.

| Table 1. Clinical characteristics of patients likely to benefit from NIV-assisted airway clearance |
|---|
| Reduced inspiratory muscle strength (PImax < 80 cm $H_20$) |
| Severe bronchial obstruction ($FEV_1$ < 40% predicted) |
| Reduced body mass index (BMI < 20 $kg.m^{-2}$) |
| Dyspnoeic at rest |
| Severe pulmonary hyperinflation |
| Low resting oxyhaemoglobin saturation |

Based on Holland et al (2003) and Fauroux et al (1999).

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# 6

# Physiotherapy management during pneumothorax

A pneumothorax is defined as the presence of air within the pleural space. In CF, it is generally termed a secondary pneumothorax as it occurs as a result of underlying lung disease. A pneumothorax may occur in CF as a result of rupture of sub-pleural blebs on the visceral pleura (ACPCF 2002, Rich 1978) or, less commonly, as a result of misplacement of a central line (ACPCF 2002). A pneumothorax can present a major problem in the person with CF as the collapsed lung can be stiff and take longer to re-expand (Henry *et al* 2003). The overall incidence of pneumothorax in patients with CF is 3.4% to 6.4% (Flume *et al* 2005, Rich *et al* 1978, Luck *et al* 1977). There is an equal risk for men and women (Flume *et al* 2005). Pneumothoraces occur more frequently in patients with more advanced disease (Flume 2003), and hence the incidence increases to 18-20% in adults (Penketh *et al* 1982, Penketh *et al* 1987, Schidlow *et al* 1993). The vast majority of people with CF, with a spontaneous pneumothorax, have an $FEV_1$ less than 50% predicted normal (Penketh *et al* 1982, Flume *et al* 2005).

For a small pneumothorax in an asymptomatic patient, medical management usually involves observation and/ or aspiration (Henry *et al* 2003). A large pneumothorax requires intercostal drainage (Henry et al 2003). Intravenous antibiotics should be started at the same time to prevent infection and consequent sputum retention, which may delay re-expansion of the collapsed lung (Henry *et al* 2003). A recurrent pneumothorax requires more aggressive management, and hence a partial pleurectomy may be performed if the patient is fit to undergo surgery (Henry *et al* 2003). If the patient is too unwell to undergo surgery, a talc pleurodesis is recommended (Henry *et al* 2003). Pleurodesis is controversial in patients waiting for a lung transplant (Noyes and Orenstein 1992, De Abreu e Silva 1996, Henry *et al* 2003) but is not an absolute contraindication to lung transplantation (Schidlow *et al* 1993).

**The physiotherapy management of pneumothorax**

There are no scientific studies to provide evidence on which to base physiotherapy in this condition. The following recommendations are based on consensus of expert opinion among physiotherapists experienced in treating patients with CF.

Pneumothoraces are a complication which impact greatly upon the airway clearance treatment of patients with CF. In general, physiotherapists need to ensure adequate airway clearance continues whilst minimising the amount of positive pressure generated inside the patient's lungs. Gentle exercise should continue.

**Small Pneumothorax**

Monitor respiratory status and cease PEP and other forms of positive pressure therapy. Ensure adequate humidification for ease of sputum expectoration. Encourage effective huffing and gentle coughing. Reduce exercise intensity and avoid upper limb resistance exercises.

**Large Pneumothorax**

If undrained, cease physiotherapy treatment and liaise with medical team. If drained
cease all forms of positive pressure therapy and only resume once the pneumothrax has resolved in consultation with the medical team. Ensure adequate analgesia and humidification and encourage gentle huffing and coughing with chest support during airway clearance therapy. Gentle graduated exercise with walking or cycling should be encouraged while maintaining shoulder range of movement and avoiding upper limb resistance exercises

**Pleurodesis**

Ensure adequate analgesia. Regular nebulised humidification and / or mucolytic therapy to decrease the viscosity of secretions and improve the ease of sputum clearance. The active cycle of breathing and autogenic drainage, with gentle huffing and coughing, are appropriate forms of airway clearance therapy. Early mobilisation should be encouraged.

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# 7

# Physiotherapy management of haemoptysis

Haemoptysis is defined as the expectoration of blood from the lungs or bronchial tubes as a result of pulmonary or bronchial haemorrhage (Hensyl, 1990). Whilst the presence of occasional mild haemoptysis is common in cystic fibrosis (CF) and not life-threatening, severe haemoptysis can lead to asphyxiation, airway destruction, shock and exsanguination, and should be treated promptly (Brinson *et al* 1998). Mild haemoptysis is common in CF, affecting approx 62% of all patients with CF (Penketh *et al* 1987). The overall incidence of massive haemoptysis in CF has been reported as approx 1% in children (Barben *et al* 2003), and between 4% and 10% in adults (Flume *et al* 2005, Penketh *et al* 1987). Research has found haemoptysis is unrelated to the severity of lung disease in children (Barben 2003). However massive haemoptysis is more prevalent in adults with severe lung disease, with approximately 60% of patients having an $FEV_1$ <40% predicted (Flume *et al* 2005, Brinson *et al* 1998). Medical management of mild haemoptysis consists of observation combined with antibiotic therapy to treat underlying infection (Schidlow and Varlotta, 1997) and the use of tranexamic acid (Wilson and Kotsimbos, 2000). For severe haemoptysis, the vessel may need to be occluded using bronchial artery embolisation (De Abreu e Silva 1996, King *et al* 1989, Schidlow and Varlotta 1997). Surgical ligation or excision of the affected segment/ lobe is recommended if embolisation is not successful (De Abreu e Silva 1996).

**Physiotherapy management of haemoptysis**

There are no scientific studies on which to base physiotherapy practice. The following recommendations are based on consensus of expert opinion of physiotherapists with significant clinical experience in CF.

Blood-streaked sputum: if the first episode, provide reassurance and education and continue with the usual airway clearance therapy and exercise routine, avoiding paroxysms of coughing.
Moderate Haemoptysis (<250mls/ 24 hrs)
Seek medical review, cease percussion, vibrations, oscillatory PEP techniques and head down tilted positions. Minimise coughing by encouraging breathing exercises and huffing to mobilize secretions to the upper airways for gentle expectoration. Some units cease the use of hypertonic saline particularly if it results in increased coughing in individual patients. Encourage gentle walking or cycling without markedly increasing heart rate and pulmonary artery blood flow. Cease vigorous exercise.
Severe Haemoptysis (>250mls/ 24 hrs)
Seek medical review. If active bleeding occurs, and the patient can feel where the haemoptysis is, position the patient in high side lying with bleeding side down. If the patient cannot tell where the bleeding is, position in upright supported sitting. Cease airway clearance therapy and exercise until the active bleeding has resolved, then continue as per moderate haemoptysis with consultation with the medical team.

**Following bronchial artery embolisation**

Ensure adequate analgesia and humidification. Gentle mobilisation following surgeon's/ radiologist's advice, then gradually increase intensity of exercise. The active cycle of breathing techniques or autogenic drainage initially appropriate followed by the gradual re-introduction of the usual airway clearance therapy routine. Resume physical exercise progressively.

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# 8

# Physiotherapy during pregnancy, labour and the post-natal period

The improvement in health, quality of life and longevity in CF has increased the desire and possibility of women to become mothers. In the 1980s, pregnancy was thought to be too risky for women with CF. Today, CF care teams can give better advice about the risks and management of pregnancy. Most of the published data report the negative and positive outcomes of relatively small case series (Fiel *et al* 1997, Frangolias *et al* 1997, Gilljam 2000, Jankelson *et al* 1998) together with descriptions relating to clinical practice (Johannesson et al 1998, 2002).

**Pre-pregnancy planning**

It is widely recognized that a planned pregnancy is likely to result in fewer problems than an unplanned pregnancy. The multi-disciplinary team involved in the care of pregnant women should at least consist of a respiratory physician, obstetrician, physiotherapist, nutritionist and psychosocial practitioner, all experienced with CF. Inhaled, oral and intravenous medication and their potential for iatrogenic effects together with optimal nutrition and dietary supplementation should be reviewed (Johannesson *et al* 1998). All women with CF are advised to approach pregnancy with an optimally effective regular airway clearance therapy routine which should be developed before pregnancy. Modifications to physical exercise should also be planned. Domestic support during pregnancy and afterwards together with child care support once the baby has arrived are necessary so that the mother has enough time and energy to carry out regular airway clearance therapy, adjunctive inhalations and exercise (Johannesson 2002).

**Physiotherapy during pregnancy**

The following airway clearance techniques are suitable for use during pregnancy in CF:

- Active cycle of breathing techniques
- Autogenic drainage
- Positive expiratory pressure (PEP) therapy
- Oscillating positive expiratorypPressure therapy (OscPEP)
- Physical exercise as airway clearance therapy
- Effective huffing from different lung volumes avoiding dynamic collapse

Pregnant women are advised to modify their physical exercise program during pregnancy. Contact sports should be avoided. Walking and swimming are appropriate forms of exercise. Women should avoid overheating and dehydration during exercise and should ensure adequate hydration and electrolyte replacement. Postural awareness, ergonomic advice, strengthening, mobilizing and stability exercises and sometimes a lumbar sacral support belt assist in managing the normal changes of pregnancy.

Postural drainage in head down tilted positions is contra-indicated during pregnancy because of the hormonal effects of progesterone resulting in hypotonia of the lower oesophageal sphincter together with the growing weight of the developing foetus applying pressure against the stomach. All of these changes predispose women to gastro-oesophageal reflux during pregnancy. Further, there is a high prevalence of symptomatic and clinically silent gastro-oesophageal reflux in adults with CF (Button *et al* 2005). In clinical practice, pregnant women report that the use of manual techniques such as percussion exacerbates nausea during pregnancy and should be avoided.

Mucolytic agents are commonly used as adjuncts to airway clearance therapy and their continued use during pregnancy should be reviewed with the CF physician. Most pregnant women with CF continue to use RhDNase(Pulmozyme) and hypertonic saline to optimize airway clearance therapy and lung health.

Pregnant women with CF should be referred to a women's health physiotherapist during pregnancy for education relating to the normal changes of pregnancy, labour and the post-partum period and to help manage the common musculoskeletal changes of pregnancy that may cause pain and discomfort.

**Physiotherapy during labour in CF**

Pain, shortness of breath and low oxygen saturation during labour are reported in healthy women (De Swiet 1991). Thus women with CF may experience these symptoms during labour. Oxygen therapy should be provided if necessary to maintain normal saturation. Bronchodilator therapy and assistance with sputum clearance may be required by some women during labour. Conservation of energy strategies should be employed. Adequate pain relief during labour is a high priority for women with CF with a normal vaginal delivery being highly desirable in order to minimize post-delivery complications (Johannesson 2002).

**Physiotherapy after Caesarian section in CF**

Adequate post-operative pain relief, oxygen therapy (if required), appropriate inhalation therapy in the form of bronchodilators and mucolytic agents together with optimal airway clearance therapy and early mobilisation are a priority after a Caesarian section.

**Physiotherapy in the post-natal period in CF**

Physical support for the mother is a priority after birth. She needs to have time and energy to carry out appropriate airway clearance therapy, inhalational therapy and post-natal exercises to ensure her future long term health.

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# 9

# Physiotherapy for the prevention and treatment of urinary incontinence

Urinary incontinence is the involuntary leakage of urine. There are two types of urinary incontinence, stress incontinence and urge incontinence. Studies of urinary incontinence in women with CF have reported prevalence rates ranging between 30% and 68% (White *et al* 2000, Cornacchia *et al* 2001, Orr *et al* 2001 and Moran *et al* 2003, Button *et al* 2004). In two studies investigating urinary incontinence in girls and adolescents with CF, the prevalence rate was between 33% and 47% (Nixon *et al* 2002, Prasad *et al* 2006). An increase in prevalence of urinary incontinence in men with CF has also been reported (Gumery *et al* 2002). It is postulated that urinary incontinence in CF is exacerbated by chronic cough, increased demands placed on the pelvic floor during airway clearance therapy and physical exercise and may be more problematic during acute exacerbations (Button *et al* 2005). Physiotherapy treatment of the urinary incontinence resulted in improved outcomes and increased continence (McVean et al 2003, Button *et al* 2005).

**Prevention of urinary incontinence in CF**

CF and continence physiotherapists have met to review current evidence and based on expert opinion have devised the following recommendations:

(1) Patients should be taught “the knack”, a contraction of the pelvic floor prior to and during any activity that increases the load to the pelvic floor (such as coughing, huffing, sneezing, laughing) to prevent leakage. This should become a lifelong habit (Miller *et al* 1998).

(2) Patients should be taught strength and endurance training of the pelvic floor and lower abdominal muscles for prevention of leakage during all activities that apply force to the pelvic floor such as physical exercise, airway clearance, huffing and coughing. The patient should be taught to draw the pelvic floor upwards towards the diaphragm, hold the contraction for 3-5 seconds and then to superimpose three quick contractions pulling each one higher up. The dosage recommended by the Women’s Health Group is three sets of ten per day (Button *et al* 2005).

(3) Patients should be taught optimal positioning during airway clearance therapy in upright sitting that enhance pelvic floor function (Sapsford *et al*, 2006). Airway clearance therapy in sitting should be carried out with feet flat on the floor with a 90° angle at hips and knees, the lumbar spine should be held in a neutral or extended position. If leakage feels imminent, the patient should apply manual pressure over the pelvic floor region or cross the legs if in a standing position to maintain bladder control.

Trampoline jumping, a commonly prescribed form of physical exercise and airway clearance therapy is appropriate until the age of puberty. Thereafter, jogging on the trampoline is more appropriate to avoid excessive downward pressure on the pelvic floor (Sherburn *et al* 2005). Patients are embarrassed about incontinence and will seldom raise the topic with the health care team. However, if asked as part of routine assessment patients value the opportunity to discuss the problem and learn strategies to prevent and / or resolve the problem. All physiotherapists working with people with CF should ask whether they experience incontinence and teach preventative /rehabilitative strategies as part of routine care. If the problem persists the patient should be referred to a specialist continence physiotherapist for assessment and treatment.

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# 10

# International Physiotherapy Group for Cystic Fibrosis

The International Physiotherapy Group for Cystic Fibrosis is an international working committee founded in 1986. IPG/CF consists of a national contact person in each member country and a committee. All countries are welcome to become members.

**The objectives of the IPG/CF are:**

1. To encourage high standards of physiotherapy practice in the treatment of patients with cystic fibrosis (CF).
2. To promote rigorous research in physiotherapy for people with CF.
3. To disseminate information and knowledge of physiotherapy practice in the treatment of people with CF.
4. To promote communication with and among Contact Persons and respiratory interest groups in countries throughout the world.
5. To advance the knowledge and understanding of CF among both medical and related professionals and lay people.

**The duties for the national contact person are:**

1. To where possible fulfil the objectives of the IPC/CF within their own country.
2. To disseminate, as appropriate, information from the Committee to interested persons within their own countries.
3. To present, in writing, an annual report to the Committee at the Annual General Meeting of the IPG/CF, for presentation and printing in the Newsletter.
4. To submit annually the recommended subscription for Contact Persons, or a donation, to the Treasurer of the Committee by 31st March of each year.
5. To receive correspondence from the Committee, such as the Newsletter twice a year.

A list of national contact persons can be downloaded from the website www.cfww.org/ipg-cf/

**This booklet is available online on www.cfww.org/ipg-cf/**