Physiotherapy for People with Cystic Fibrosis: from Infant to Adult

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1st edition 1993
2nd edition 1995
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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. Current physiotherapy management of CF is multifaceted, inclusive of 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 collaboration with the patient and family, to tailor an individualized, reasonable, effective and efficient physiotherapy regimen. This should take into consideration all relevant physical and psychosocial factors.

Current physiotherapy management of CF 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 adults is increasing and this trend is likely to continue. Comorbidities 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.

The addition of several new techniques and physiotherapy management approaches in this updated version of the Blue Booklet reflects the ever evolving and advancing role of the physiotherapist in the management of infants children and adult with Cystic Fibrosis.

On behalf of the IPG/CF,

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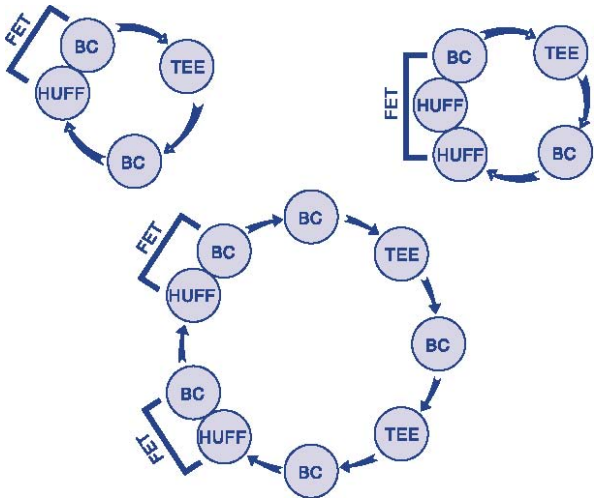
Airway Clearance Techniques

Active Cycle of Breathing Techniques

The active cycle of breathing techniques (ACBT) is a flexible airway clearance regimen which is used to mobilize and clear excess bronchial secretions (Pryor et al 1979). This technique has been used for many years, for a range of ages, respiratory conditions and disease severities. Following suitable training, the technique can be performed independently and in a position of choice.

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

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 is an integral part of the cycle as it regulates airflow and helps to minimise irritation and bronchoconstriction of the airways. Breathing control should be continued until the person feels rested and ready to use the more active components of the technique – the thoracic expansión exercises or 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' allows time for all lung units to expand as air flows more slowly into diseased and obstructed regions than into healthy, unobstructed areas – Pendelluft flow (Mead et al 1970). Up to three thoracic expansion exercises are followed by breathing control and may be combined with chest shaking or percussion. Combining ACBT with manual techniques can be of benefit to some people, but are not required by others.

The Forced Expiration Technique (FET) is the combination of one or two forced expirations (huffs) and periods of breathing control for rest. Huffing in low lung volumes should assist in loosening and mobilising excess bronchial secretions from smaller peripheral airways to larger central airways. When secretions reach the larger larger

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.

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 forced expiration technique should never be violent or cause wheeze. 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). In end stage lung disease, the ACBT can be used in conjunction with non-invasive ventilation (NIV) where settings can be adjusted to increase thoracic expansion (increase in IPAP), support breathing control (usual settings) with some evidence to suggest that offloading the respiratory muscles with NIV during airway clearance can improve tolerance in advanced disease.

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 nonproductive or it is time to rest or stop treatment. 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), oscillating PEP Flutter® (Pryor et al 1994, Pike et al 1999) or mechanical percussion (Pryor et al 1981). 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 Concept of Flow and Breathing Level Modulation"

Introduction

Autogenic Drainage is an airway clearance technique based upon elementary physics, fluid dynamics, pulmonary anatomy, respiratory physiology and breathing mechanics.

The mechanism of mucus clearance rests on two different systems; the ciliary clearance and the effects of shearing forces induced by the airflow. This last phenomenon can be compared to erosion; the higher the velocity of the medium, the stronger the erosive effect on the bronchial walls.

Because the cross-sectional area of the trachea is 180 times smaller than the cross sectional area of all the peripheral airways, the bronchial tree has a funnel shape and function. Therefore, it is mandatory to take into account the specific characteristics of fluid dynamics related to a funnel model.

To create the necessary expiratory shearing forces to clear the bronchi from secretions, it is essential to modulate both 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 obstructive lungs and a back flow of the secretions.

During exhalation the optimal shearing forces, induced by the expiratory linear airflow velocity, must be localised to where the secretions are.

By modulating the breathing level within the vital capacity and the intra-thoracic pressure generated by the expiratory muscles, the optimal airflow will be obtained at the precise level of the bronchial "funnel" where the secretions are.

The realized intra-thoracic pressure may not exceed the stability of the airways and cause a dynamic airway compression which leads to a drop in flow rate and consequently in airflow velocity thus inducing inhomogeneous emptying and possibly increase the trapped gas volume.

Key reflections and considerations:

- In obstructive lungs the distribution of the obstructions is rarely homogeneous
- The main driving force to exhale is the alveolar recoil force
- To localize the secretions the three following feedback signals, auditive, tactile and proprioceptieve, have to be used. A fourth feedback signal, the olfactive one, indicates that an obstructed area has been opened.
- The most important aspect in ACT's is the homogeneous filling of all the lung parts really putting air behind the mucus plugs
- One cannot empty what previously has not been filled
- Use the collateral airways if they are already present

Breathing in Autogenic Drainage

The whole airway clearance process in AD is based upon an active modulation of the air flow and lung volume level breathing.

Before clearing the lower airways it is evident to clear the upper airways to optimize the nasal breathing during the therapy session.

The positioning of the patient and the shape correction of the respiratory pump, to optimize the action of the respiratory muscles, induces more homogeneous ventilation.

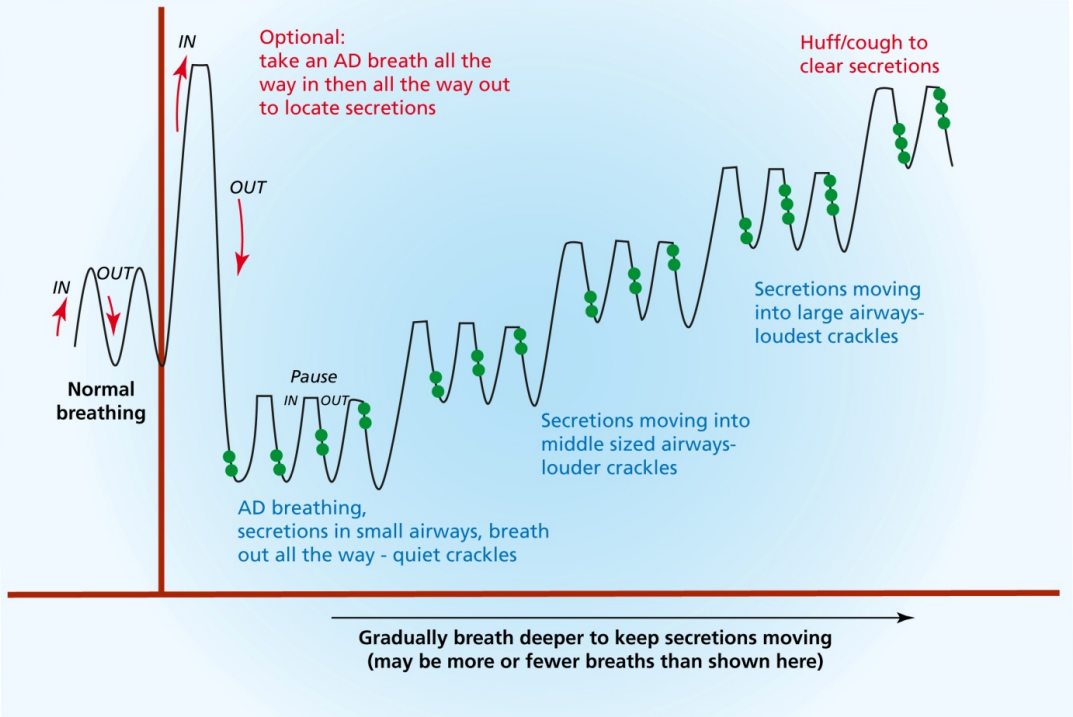
Taking special care of spastic and/or swollen airways is a must in all ACT's. The bronchial resistance must be normalized as much as possible and the secretions must be made easier to remove by means of drugs, or special devices like the oscillating PEP (Flutter). This should be done after correction of the respiratory pump.

Correct dosage of the expiratory force 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 effort and decreases the appearance of paradoxical airflows and breathing movements. For some reason the incentive to cough is less intense which allows one to inhibit and postpone the cough more easily. Postponing cough allows the patient to collect more mucus from the depth of the lungs and makes the migration of the secretions easier.

Modulating the breathing level within the vital capacity makes it possible to avoid an "air jam" in the central airways which lowers the flow rate and consequently the velocity of the airflow in the smaller (peripheral) airways.

When the right modulation of the breathing level and the expiratory force is found, it is easy to hear and feel the secretions migrate centrally. These two feedback signals added to the proprioceptive feedback makes it simple to adjust the modulation of the breathing.

Feedback "leads the way" and is essential to perform a thorough clearance of secretions.



Example of the breathing level modulation in AD. The green dots represent the collection of the secretions migrating from the periphery to the central airways during the exhalations.

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 and to avoid paradoxical movements to put more air behind the mucus 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

7. Breathe OUT the chosen functional tidal volume preferably through the nose.
   If a drop in velocity does occur or, if one wants to hear the bronchial noises more distinctively, breathe OUT through the mouth. In this case always keep the upper airways (glottis, throat, mouth) open.

8. 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.

9. Breathing Out correctly, the mucus (crackles) can be heard easily.Putting the hands on the upper chest, one can also feel the mucus vibrating. The frequency of these vibrations indicates 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. As soon as 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 into 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 properties of the secretions.

Experienced patients clear their lungs more quickly than others. Drainage should always be done thoroughly. Now a days the main thing is not how much secretions have been cleared, but, how much secretions are left in the lungs finishing the session!

The principles of AD can be used in obstructive and restrictive pulmonary diseases. Active participation is preferred but is 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 (inhalation therapy) 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. Bouncing at low amplitude (4-6cm) was used to maximize the relaxation of the infant, avoiding resistance against or crying during treatment and enhancing 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.

AAD reduces significantly the length of hospital stay and some respiratory symptoms of bronchial obstruction compared to no physiotherapy in children with mild to moderate bronchiolitis (1).

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

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Positive Expiratory Pressure (PEP)

The airway clearance technique PEP is a treatment cycle that includes both breathing towards an expiratory resistance and the forced expiration technique (FET). The FET has been described earlier in the ACBT chapter.

Technique

PEP was developed during the late 1970's (Copenhagen, Denmark), using a mask with a one-way valve to which an expiratory resistor is attached (PEP-mask®). A manometer measuring the expiratory pressure achieved is inserted between the expiratory valve and the resistor to ascertain the optimal size of the resistor. The resistor used for treatment is determined for each individual patient, giving a mid-expiratory pressure of 10-20 cm H2O during 12-15 consecutive breaths. This results in a temporary, limited increase of functional residual capacity (FRC). The quality of the technique is assessed by observing and listening to the breathing pattern. The manometer is used by the physiotherapist to find the optimal resistor but is preferably not used during subsequent treatment sessions, as the focus will be on

pressure rather than technique. The aim of the first part of PEP, expiring towards the resistance, is to recruit obstructed and collapsed airways, thus enabling mobilisation of secretions which can then be transported and evacuated by FET during the second phase.

Physiology

The lung tissue elastic recoil is increased while the FRC is temporarily increased. The interdependence between airways and lung tissue recruits obstructed and collapsed airways through ordinary and collateral airways. This results in more homogenous ventilation. During the FET phase the air in the re-opened airways can be utilized for mobilising and transporting secretions to the central airways, and evacuated by coughing, thereby facilitating the mobilisation and evacuation of secretions, Figure 1.

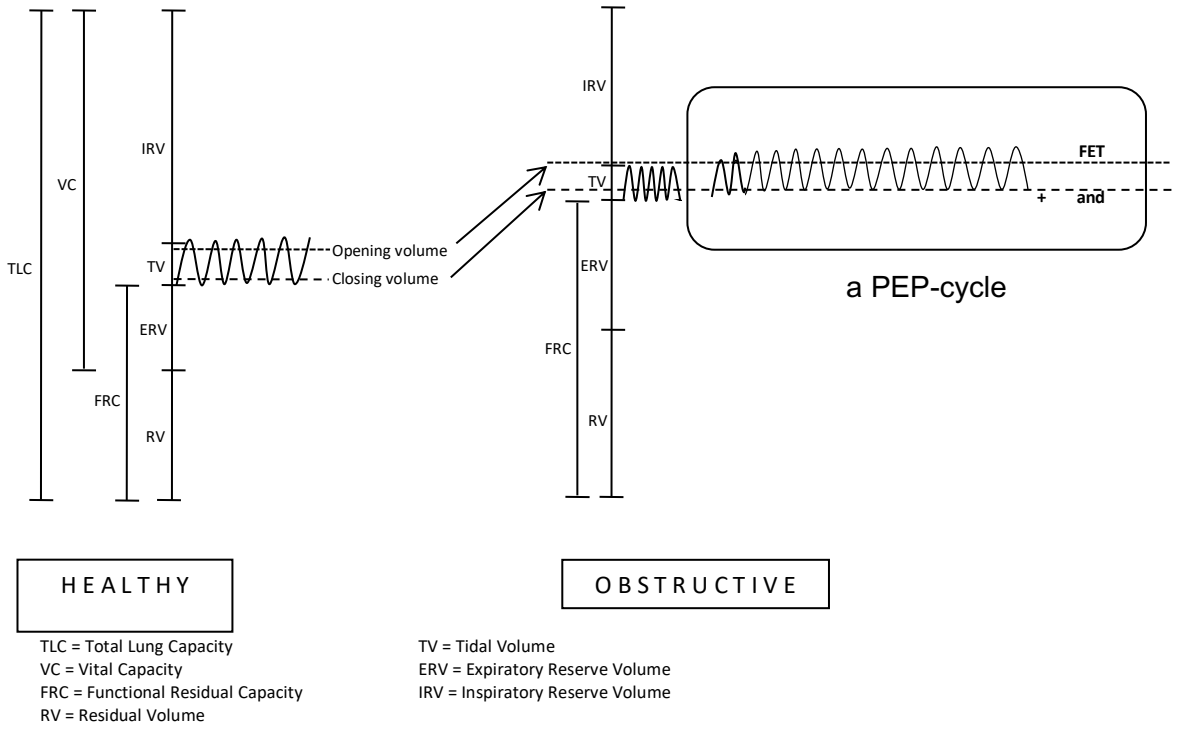

Figure 1. Physiological strategy of the airway clearance technique Positive Expiratory Pressure (PEP).

Schematic drawing.

Instructions

To perform a PEP cycle, patients are instructed to

- sit comfortably at a table, lean slightly forward with elbows on the table and place the head in the mask

- hold the mask tightly over nose and mouth with both hands
- breathe 12-15 consecutive tidal volume (TV) breaths with slightly active expiration
- remove the mask
- mobilize and transport secretions by adequate number of FET at different lung volume levels
- evacuate by coughing when secretions are collected and transported to the central airways

The lung volume and muscle effort and the number of FETs performed are tailored for each patient. The number of PEP cycles within a treatment session and the treatment frequency are also tailored to individual needs.

PEP is used in babies, toddlers and young children, although not scientifically evaluated. An expiratory pressure of 10-20 cm H2O cannot be expected to be achieved in children too young to follow the instruction to perform a "slightly active expiration". In practice, when using PEP in this age-group, changes in breathing pattern and increase in lung volume have been observed. Breathing towards the resistance in the mask is interspersed with well-coordinated, repeated very gentle facilitated exhalations without the mask. The aim of the facilitated exhalations is to enhance expiratory airflow slightly (but not maximal) to allow secretion mobilization at a temporarily lowered lung volume. Professional expertise and manual skill is mandatory to tailor the treatment, to teach the parents carefully and to frequently check and optimise the technique and the cooperation with the child. If a baby, toddler or child is uncomfortable or very upset, and crying or screaming it is not possible to achieve temporarily lowered lung volume or increased expiratory flow. If facilitated exhalations are not well coordinated and gently performed they should be avoided.

Children can start practising and playing with forced expirations usually at the age of 2½-3 years. Once they have achieved good quality FETs they start to use the "adult" PEP cycle. The length of time it takes to achieve this varies between individuals. The number of treatment cycles per session and sessions per day varies depending on individual needs.

Contraindications:

Contraindications to PEP are undrained pneumothorax and massive haemoptysis. In young children with fragile abdomen, facilitated exhalations are inappropriate.

Considerations:

PEP devices

There are many expiratory resistance devices available on the market and new devices are constantly emerging. The expiratory resistance can be connected to a mask or mouthpiece or come with the choice of both. Using a mouthpiece may result in air leakage nasally during the PEP breaths. Patients can learn to compensate for this but may need a nose clip. A nose clip may be too uncomfortable and some patients may prefer a mask. Which interface is best is decided by the physiotherapist in consultation with the patient, ensuring that the physiological response desired is achieved.

Resistance is either flow or pressure regulated depending on the device. To what extent it influences the achieved pressure and lung volumes must be evaluated for each individual. Although the hole is equally large in two different flow regulated devices, the resistance at a certain flow may differ due to the length of a tube and/or other characteristics. Therefore, devices can´t be interchanged without evaluation.

Pressure and volume

The ability to analyse and correct the immediate response to instructions during treatment is of utmost importance for good results. Achieved expiratory pressure is the most easily measured parameter, which may be why pressure alone has become the guide in choosing resistor size. But, in spontaneously breathing patients the expiratory pressure alone is not sufficient for good results, the parallel expected change of breathing pattern and temporary increase of FRC is equally important. Therefore, it is essential that a physiotherapist evaluates the choice of resistor size and the treatment quality to ensure optimal use and results.

Converse effects on FRC

Breathing towards an expiratory resistance is a tool that can be utilized in different ways for totally different respiratory physiological aims. To decrease dyspnoea and improve ventilation by normalizing FRC and TV in obstructed, pulmonary hyperinflated patients is one such aim utilizing pursed lips breathing. As previously described the correct instruction for the airway clearance technique PEP is to perform TV breaths with "slightly active expirations", achieving a "mid-expiratory pressure of 10-20 cm H2O", parallel to a "temporary increased FRC". An incorrect instruction appearing in discussions and the literature is to perform TV breaths with "a stable mid-expiratory pressure of 10-20 cm H2O maintained during expirations", which will decrease rather than increase FRC. Using the resistance correctly is essential for good treatment quality and making comparisons to other techniques possible. Another airway clearance technique that is based on the use of an expiratory resistance is high-pressure

PEP (HiPEP), which is described in the HiPEP chapter. It is crucial that PEP and HiPEP are not confused, as the instructions to the patients and the physiologic aims are totally different.

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Oscillating PEP – Therapy

Flutter VRP1 and Co

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 [1] The oscillating positive expiratory pressure is reported to prevent premature closure of the bronchi, to loosen secretions and improve mobilisation of sputum (detaching mucus from the airway walls) which may be subsequently cleared by the forced expiration technique, a huff or a cough. The Flutter VRP1, as a form of the oscillating positive expiratory pressure technique, was first developed in Switzerland [2].

The device (Figure 1.1) is made of a mouthpiece (Figure 1a), a plastic cone (Figure 1b), a steel ball (Figure 1c) and a perforated cover (Figure 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 VRP1 or the Acapella device.

The Acapella device uses a counterweighted plug and magnet to create air flow oscillations. The performance of the Acapella is not gravity dependent (i.e. not dependent on device orientation) and may be easier for some patients to use, particularly at low expiratory flows [3].

Further devices are the RC-Cornet, the Shaker or the Aerobika. The Shaker works the same way as the Flutter VRP1 with a steel ball in a circular cone and in addition, it provides the option of exchanging the mouthpiece position [4].

With the RC-Cornet, the positive expiratory pressure and the oscillations are created by blowing out into a curved plastic tube, in which a flexible rubber hose is connected inside to the mouthpiece. The frequency of oscillation, pressure and flow can be adjusted (by turning the rubber hose within the plastic tube). The RC-Cornet creates a higher constant positive expiratory pressure and therefore reduces hyperinflation and decreases residual volume [5].

The Aerobika creates a positive expiratory pressure and oscillations by movable flap in the device which is activated by blowing slowly, steady and firmly into the Aerobika. Due to the fact, that the Aerobika is a fairly new device there are no published data regarding the use for Cystic Fibrosis patients.

With devices such as the Flutter VRP1, 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 repeatedly forced.

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 promotes the clearance of the obstructed airways.

Modulation of the flow and pressure oscillations are obtained as follows, applying the same approach as for Autogenic Drainage: The patient exhales into the Flutter VRP1 device; during exhalation the steel ball inside the device bounces, causing a vibratory obstruction to air flow, which oscillates both the pressure and air flow only during exhalation.

The Flutter VRP1 generates PEP in the range 18 to 35 cm H2O. The angle at which the device is held determines the oscillation frequency (usually between 6 and 26 Hz) [6] 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 manoeuvre for 10-15 breaths (Figure 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:

1. The patient should sit comfortably (Figure 2), hold the Flutter VRP1 horizontally, put the mouthpiece of the device into their mouth, between their teeth and close their lips tightly around the mouthpiece. Take a slight deep breath through the nose, hold their 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), and breathe out normally and deeply (Figure 4), keeping their cheeks flat and hard, using the method of unforced abdominal exhalation while relaxing the muscles of their upper chest.

2. Repeat breathing in through the nose and out again into the Flutter VRP1. On successive attempts, the patient may find it necessary to move the Flutter VRP1 (Figure 3) slightly upwards (higher pressure, oscillation frequency and flow amplitude) [7][8][9] or downwards (lower pressure and oscillation frequency and more of a huff effect) [8] by a few degrees. The aim is to feel the full effects of the vibrations at the abdominal level during the first stage of the exhalation process and ideally feel the mucus moving upwards. The angle of the Flutter VRP1 has to be adjusted to reach this optimum effect for the individual and their current condition [8].

It is not necessary to complete a full exhalation each time when breathing out through the Flutter VRP1 (Figures 4 & 5).

During each 10 to 15 breath cycle coughing should be suppressed until the last exhalation, which should be performed at about twice the speed of a normal exhalation. This should automatically initiate 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.

The Flow-Volume curve (Figure 6) shows the benefits of the Flutter VRP1 device in comparison to a forced expiratory manoeuvre. [10]

The Flutter VRP1 can also be used while lying horizontally on either side or on the back. The Flutter device should therefore be held in a position, so that the steel ball is exposed to gravity and can function properly.

Short-term studies with CF patients have shown the Flutter VRP1 produces similar effects to those of CPT and PEP [11, 12, 13, 14].

A randomised crossover study of patients with stable CF compared 4 weeks of treatment using 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 VRP1. [15]

Tambascio et al [16] demonstrated that the use of the Flutter_VRP1 for a four week period is capable of altering and improving the respiratory secretion transport properties and that this alteration is related to the high frequency oscillation component.

Chicayban et al [17] in a clinical trial with mechanically ventilated patients found an improvement of lung secretion removal mucus production and respiratory mechanics using the Flutter VRP1.

Konstan et al.[11] 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. [18]found that significantly more sputum was produced with the active cycle of breathing techniques (ACBT) than with the Flutter VRP1 in individual supervised sessions but similar sputum weights were produced with both methods over 24 hours.

Two studies compared the Flutter VRP1 with percussion, vibration, and postural drainage carried out 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 showed similar improvement in pulmonary function and exercise tolerance compared to CPT after two weeks of therapy, suggesting that Flutter VRP1 valve therapy is an acceptable alternative to standard CPT during inpatient hospital care of patients with CF [12].The second study demonstrated that the Flutter VRP1 device appears to be safe, efficacious, and cost effective for CF inpatients capable of undertaking this type of therapy.[14]

One long-term study (more than a year) in children with CF compared the Flutter VRP1 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 [19]

Eaton [20],in a randomized prospective study, evaluated the acute efficacy, acceptability and tolerance of three airway clearance techniques in non-cystic fibrosis (non-CF) bronchiectasis: Flutter VRP1, 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. The patients' preferences indicated 16 (44%) for Flutter VRP1, eight (22%) for ACBT and 12 (33%) for ACBT-PD but ACBT-PD proved superior in terms of acute efficacy.

Pereira dos Santos [4] compared, in an experimental model, the effects of the Flutter VRP1, the Acapella and the Shaker. The level of positive expiratory pressure of the three devices was not different. The pressure amplitude produced by the Flutter VRP1 and Shaker were greater at low and high pressure. All three devices produced pressure and oscillations which aid in the transport of respiratory secretions.

Pryor [21] found oscillating positive expiratory pressure devices to be as effective as other airway clearance techniques.

In recommendations for obstructive disorders, Rosière [26] ranks the Flutter VRP1 in Category C.

A randomised crossover study was performed by Thomson [22] 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 concluded that the daily use of the Flutter VRP1 device in the home is as effective as ACBT in patients with non-cystic fibrosis bronchiectasis and has a high level of patient acceptability.

Morrison [27] in its Cochrane Review states no clear evidence that oscillation was a more or less effective intervention than other form of physiotherapy and that additional evidence is needed to evaluate whether oscillating devices combined with other forms of airway clearance is efficacious in people with cystic fibrosis.

Oscillating positive expiratory pressure devices such as the Flutter VRP1 are relatively inexpensive especially when compared to other airway clearance techniques which require assistance from a physiotherapist or machines. [23] Once the technique has been taught by a respiratory physiotherapist and learned by the patient, it can be performed regularly and independently by the individuals themselves. However, regular supervision by a respiratory physiotherapist is recommended and important.

For the time being, it seems best to choose the method that matches the patient's ability and preference in order to improve compliance (or satisfaction [24]) with the physiotherapy regimen. Flume [25] stated that the Airway Clearance Technique should be adapted to patient's individual needs and preferences.

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 choosing any one technique for all patients with cystic fibrosis.

Figures

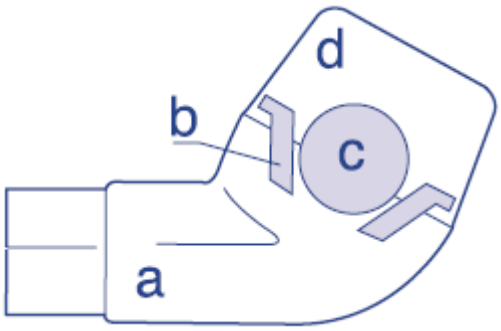

Figure 1: Flutter VRP1 - Detail inside

Figure 1.1.: Flutter VRP1dissasembled and assembled

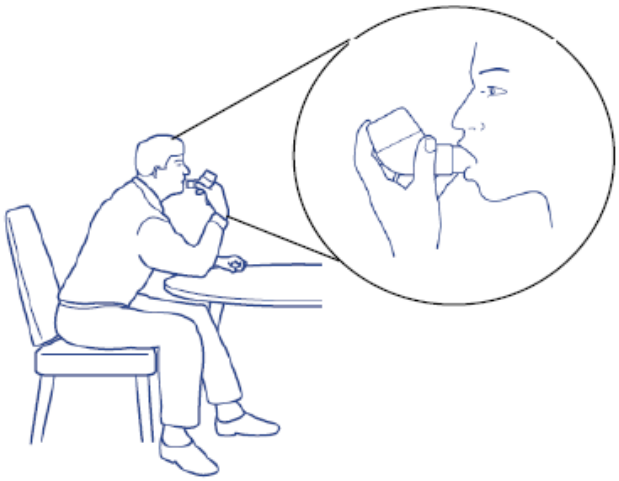

Figure 2: Flutter VRP1 - Position for use

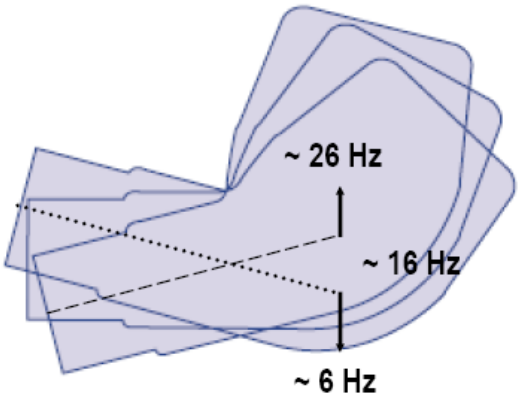

Figure 3: Moving the Flutter VRP1 up or down

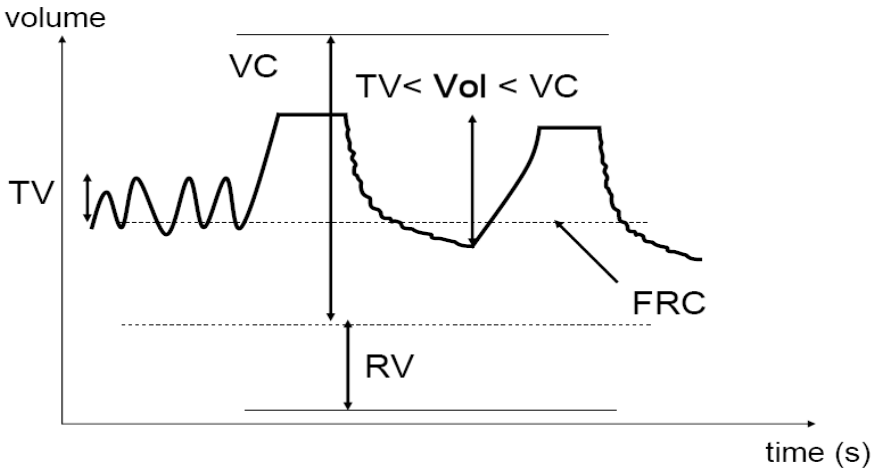

Figure 4: Schematic way of breathing with the Flutter VRP1 (Volume – Time)

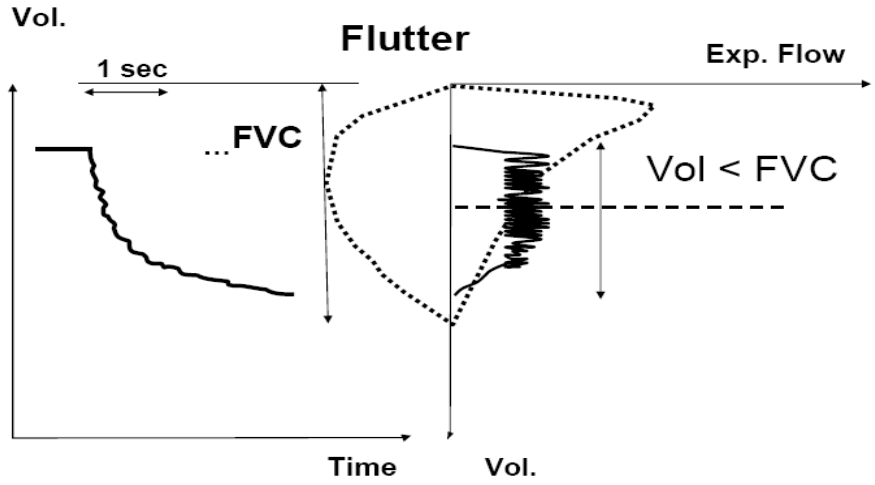

Figure 5: Schematic way of breathing with the Flutter VRP1 (Flow – Volume)

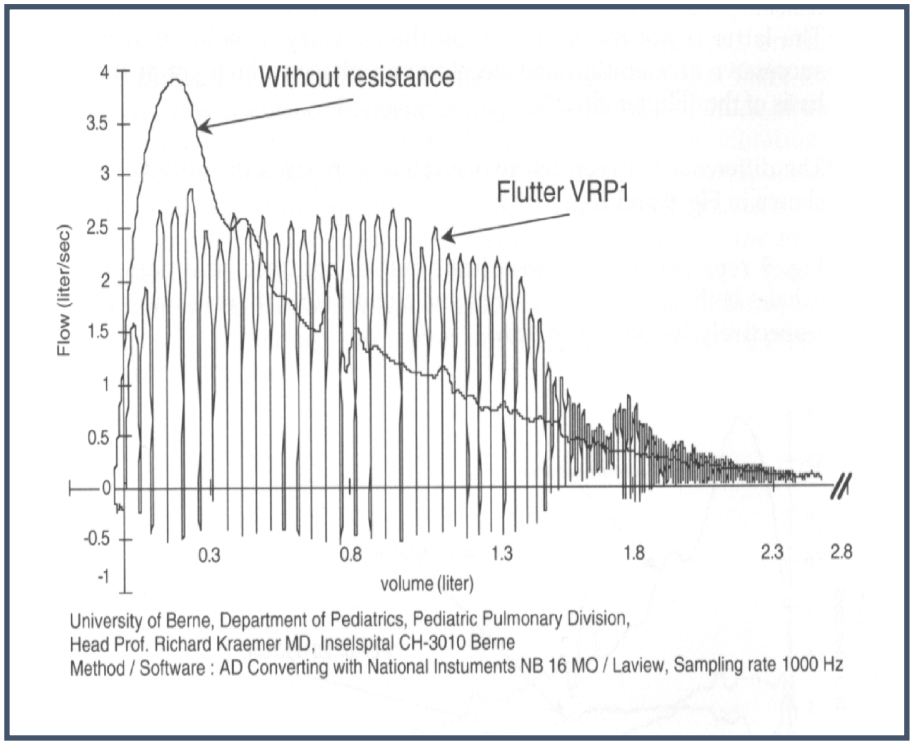

Figure 6: Flow-Volume curve without resistance and with the Flutter VRP1. Published with permission of Prof. Richard Kraemer [28]

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Revised Article with permission from Patrick Althaus CH-1041 Bottens

Bottle PEP

Bottle PEP is a positive expiratory pressure device that can be used to increase resting lung volumes equal to tidal volume or increase lung volume temporarily equivalent to PEP and enhance sputum clearance.

The aims: to recruit obstructed or collapsed airways and under-ventilated lung regions improve gas exchange and mobilize pulmonary secretions using ordinary ventilation route and/or collateral ventilation.

Equipment

- A plastic bottle of at least 1 litre capacity
- A length of tubing 4-5mm in diameter and 20-40 cm long (individualized to each patient's requirements)
- Water column of 4 – 10 cm depending on individual patient's pathophysiology and therapeutic needs
- Pressure manometer and connector to measure pressure (optional)**:** according to aims of techniques
- For children a few squirts of dish liquid and some food colouring make this activity more fun making bubbles
- Commercially available customized equipment is available in some regions.

Indications:

- Sputum retention
- Atelectasis
- Lobar consolidation

Contra-indications / precautions

- Frank haemoptysis
- Undrained pneumothorax
- Extensive / large bullae / cysts
- Post oesophageal and lung surgery
- Ear drum infections / risk of perforation
- Cognitively impaired / acute confusion
- Fluid restriction (with patient likely to suck on the tubing instead of blowing)
- Haemodynamic instability
- Increased intra-cranial pressure
- Severe bronchospasm

Methods

- Select a length of tubing and column of water and place in a bottle (individualized to each patient's pathophysiology and therapeutic requirements)
- Position the tubing close to the bottom of the container during use
- Maintain an open exhalation port to allow air to move out of the bottle during expiration
- Aim for a positive expiratory pressure of 10-20 cmH2O during the middle part of expiration
- Select technique according to aim and age of patient
- Instruct the patient to inhale and exhale the therapeutically selected breathing volume based on each individual's pathophysiology and needs
- Select the number of breaths per cycle and cycles per treatment to achieve the treatment aims
- Other inhalation and exhalation techniques can be used by different physiotherapists with individual patients to achieve their therapeutic aims
- BPEP should always be accompanied by intermittent forced expirations (huffing) and coughing when ready to expectorate sputum

Cleaning and storage

- After each session the bottle and tubing should be emptied and cleaned to local infection control requirements
- Equipment should be left to air dry between treatments
- Patients should be provided with written instructions on use of their device, dosage, treatment plan and cleaning method

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Hi-PEP

Technique

The High-Pressure technique of PEP-mask physiotherapy employs forced expiratory manoeuvres against the PEP-mask's flow-regulated 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 it needs to be equipped with another manometer for monitoring higher pressures, if the pressure is expected to become measured. 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 cm of H2O. The specific level reached is not important – it is highly individual, but an achieved positive expiratory pressure at the end of the expiration is a pre-requisite for reaching a Residual Volume (RV) as low as possible. 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, the size of forced vital capacity and the expiratory airflow velocity at the end of expiration, as determined by the shape of the flow–volume curve (Figure).

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 the first part of Hi-PEP-therapy can be explained by increased interdependence and collateral airflow to underventilated regions; air expired from there should mobilize obstructing secretions. In addition, during the second part, 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.

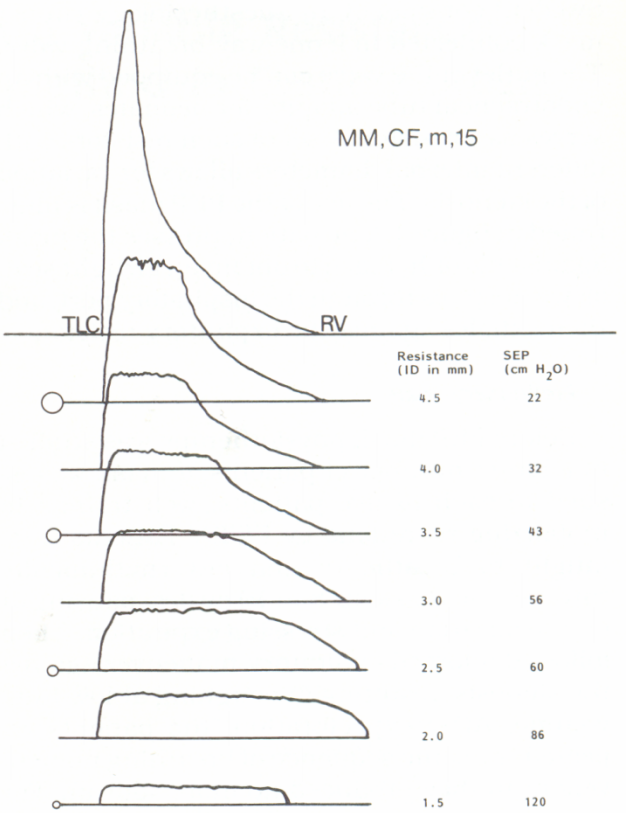

Figure 1: A series of MEFV-curves blown by a CF-patient through differently sized resistors.

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

Figure 1 shows 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 homogenisation 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. And, with optimal resistor, the expiratory airflow velocity actually increases at the end of the expiration, or even better, parts of the lungs that did not participate in the expiration are contributing as the previously trapped gas is exhaled and the RV is decreased. 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!). In this patient case, 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 at the beginning of the expiration. 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 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 clear 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. Full chest mobility is maintained and in- and expiratory muscle training, as a side effect of this technique, comes for 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, very experienced physiotherapists may also apply Hi-PEP in 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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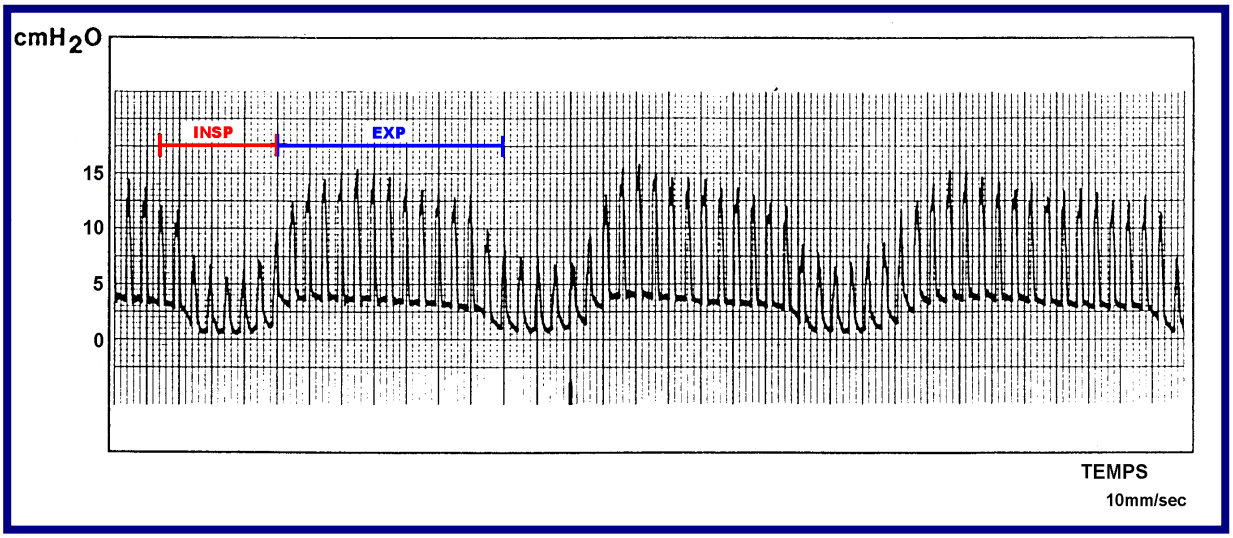
Intrapulmonary Percussive Ventilation

Intrapulmonary percussive ventilation (IPV) is a ventilatory technique that uses a device to deliver small bursts of high-flow air into the lungs at high rates, superimposed upon the spontaneous breathing pattern. This causes airway pressures to oscillate between 5 and 35 cm H2O and the airway walls to vibrate in synchrony with these oscillations. A unique sliding venturi, called a phasitron, which is powered by compressed gas at 0.6 to 6 bar, generates these oscillations in the range of 80 to 650 cycles per minute. [1] The high-frequency gas pulses expand the lungs, vibrate and enlarge the airways, and deliver gas into distal lung units, beyond accumulated mucus. [2] IPV can be used with a mouthpiece or a well-fitting mask. Frequency, pressure and inspiratory-expiratory ratio can be adapted during IPV. Adjustment of the IPV variables depends on the patient's pathology. High frequencies (250-300 cycles/min) and low pressures (8-20 cm H2O) are used to mobilize secretions, to open-up non-ventilated territories (atelectasis), to fight against bronchospasm and to improve ventilation and gas exchange. Low frequencies (80-150 cycles/min) and high pressures (40-80 cm H2O) are used to work on the thoraco-pulmonary compliance and to stimulate lung development and growth. Nebulisation of medication with the IPV device has to be avoided. Whole-body deposition of particles is significantly higher with IPV compared to standard jet nebulisation, due to a higher extrapulmonary deposition. The intrapulmonary deposition of nebulized particles is too variable and thus too unpredictable to recommend its use for drug delivery to the lung. [3] IPV improves airway secretion clearance in CF patients [4,5], Duchenne muscular dystrophy [6], adult and pediatric patients with atelectasis [7,8], COPD exacerbation [1,9,10], tracheostomized patients [11] and patients with acute respiratory failure [1]. IPV reduces significantly the length of hospital stay and some respiratory symptoms of bronchial obstruction compared to no physiotherapy in children with mild to moderate bronchiolitis12. The use of IPV should be avoided in patients with a non-drained pneumothorax and with haemoptysis. IPV in upright position at a high frequency of 300 cycles/min. with pressures between 10-12 cm H2O does not induce, nor aggravate gastro-oesophageal reflux (GOR) in infants without and with pathological GOR, respectively. On the contrary, a decreased number of reflux episodes has been found [13].



Pressure/time waveform of a patient treated with IPV®, with spontaneous breathing. Work of breathing is performed by the patient and the device.

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Specific Cough Technique

The Specific Cough Technique (SCT) is a planned and controlled cough used at specific lung volumes to localise and clear mobilised bronchial secretions (Gursli 2005, Gursli et al 2017). SCT was developed in Norway in the second part of the 90s (Gursli 2005). By using SCT patients may clear mobilised mucus with less effort, thus covering quality criteria for treatment defined by GEMS, i.e. Gentle – Efficient – Motivating – Self-supporting (Gursli 2005).

Features of the Specific Cough Technique

The SCT comprises one gentle cough at lower lung-volume, followed by two-three coughs from higher lung-volume to expectorate, and followed by tidal breathing. SCT is characterized by two phases as outlined below (Figure 1). (Gursli 2005, Gursli et al 2017).

1. Localise mucus at lower lung volume

The initial phase starts with a relaxed and prolonged expiration followed by one single and gentle cough at lower lung-volume to localise and collect mobilised mucus. The cough is initiated from the larger bronchi or trachea. It involves a soft glottis closure and is characterised by a low-sound.

2. Clear mucus from higher lung volume

The second phase starts with an inspiration, which typically comprises a mid-sized breath, relative to the subsequent cough-effort. Inspiration is followed by two-three coughs with modest cough-effort from higher lung-volume, i.e. two coughs after the first breath followed by a modest inspiration and one single cough to expectorate, followed by tidal breathing.

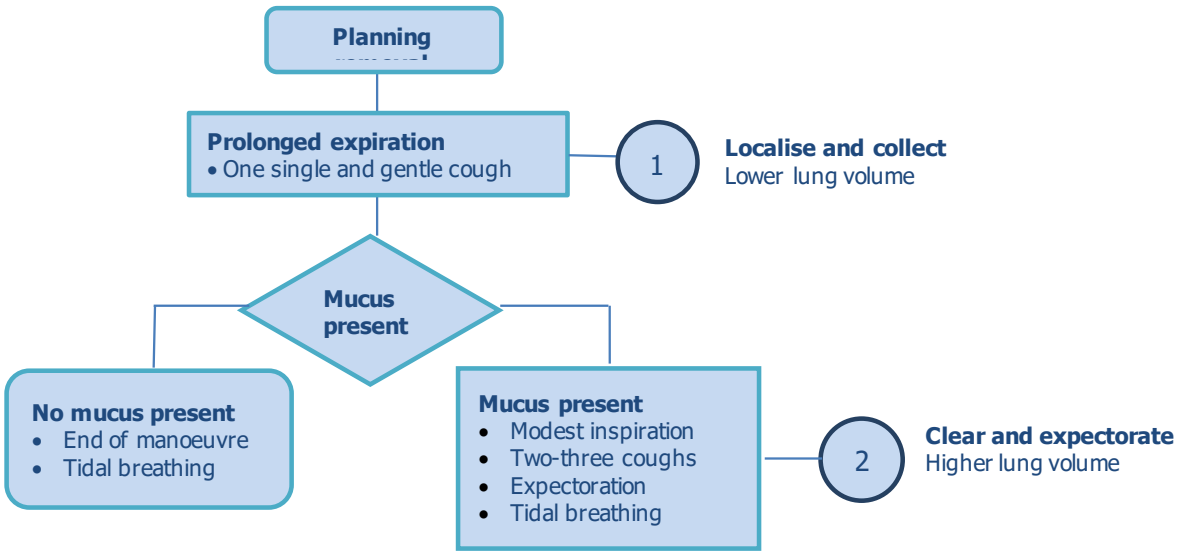

Figure 1. The two phases in Specific Cough Technique for the clearance of bronchial secretions: 1. Localise and 2. Clear

Mucus clearance by Specific Cough Technique

The SCT aims at localising mobilised mucus before expectoration by utilizing the basic cleaning mechanism of cough. The technique allows adaptation and control of the expiratory starting volume, the subsequent inspiratory volume and cough-effort, thus the pressures and flows being developed. Unlike cough in general, SCT starts with a relaxed and prolonged expiration followed by a single and gentle cough characterised by a low expiratory sound (Figure 1). The inspiratory volume typically involves a mid-sized breath aiming to prepare for two-three coughs with modest cough effort. Hence the necessary cough flow velocity is attained to aid sputum removal by cough (McCool 2006). Mucus is cleared from central airways utilizing the cleaning mechanisms of two-phase gas-liquid flow, local dynamic airway compression and increased expiratory airflow velocities, thus enhancing sputum removal (van der Schans et al 1999, Foster 2003, McCool 2006).

SCT is adapted to individual needs with the aim to localise and clear mobilised sputum. Furthermore, the initial cough can be used to check whether mucus is present or not. When mucus is present the manoeuvre continues to the second phase by breathing in slowly preparing for two-three coughs and sputum expectoration, i.e. two coughs after the first breath followed by a modest inspiration and one cough to expectorate. However, some patients may perform one cough after the first breath followed by a modest inspiration and one cough to expectorate. The manoeuvres are repeated according to need. When no mucus or too little mucus is present, or the patient feels it too early for sputum expectoration, the expiratory manoeuvre ends after the first phase and the mobilising part of treatment continues.

SCT may be used as an alternative technique in airway clearance therapy to aid mucus clearance and expectoration. The technique can be used in individually tailored treatments with other techniques and therapies, i.e. as part of an airway clearance strategy; in conjunction with saline inhalation and/or physical activity/exercise, when used as part of treatment (Gursli 2002, Gursli 2005), or as part of an ACT, e.g. Positive Expiratory pressure (PEP) or Oscillating Positive Expiratory Pressure (OPEP).

Aerosolized saline is preceded by bronchodilator when recommended and used in conjunction with or during airway clearance to hydrate and mobilize secretions (Houtmeyers et al 1999, Fahy & Dickey 2010, Pryor 1999, Dentice et al 2012). Breathing - cycles are performed calmly and slowly to increase tidal volume and expand lung tissue, improve ventilation and deposition of inhaled saline and mobilise secretions, alternating with SCT to expectorate (Gursli et al 2017). Horizontal side-lying positions may be used as the preferred treatment position to optimize relaxation and aid sputum expectoration (Gursli 2005). Further, to increase or change ventilation and increase mucociliary clearance (Cecins et al 1999).

Physical activity/exercise is used according to age and preference to improve ventilation and mobilize secretions alternating with SCT according to need (Gursli 2002). More and rapid breathing during exercise has beneficial effect on ventilation and helps dislodging mucus from the airway wall, thus secretions can be moved in central direction (Hebestreit et al 2001, Foster 2003). Children may use both saline inhalation and physical activity as part of treatment with the choice and order based on beneficial response. Most children can check for and/or clear sputum with SCT from about five years, thus being more able to rely on the response. When bronchial secretions are present the frame and setting for the treatment is important to make expectoration possible and desirable (Gursli 2005).

The SCT appears to be a promising technique for the clearance of mobilised secretions. The technique has been shown to be an effective alternative to Forced Expiration Technique (FET) in adult patients with CF, and found to be safe and well perceived by the patients (Gursli et al 2017). Furthermore, evaluation and experience from clinical practice has shown that SCT may be of benefit in other lung diseases with bronchial secretion issues, e.g. primary ciliary dyskinesia (PCD), non-CF Bronchiectasis, immunodeficiency disorders, COPD. SCT involves a planned and controlled performance. By starting with expiration and a single, gentle and low sounded cough to localize bronchial secretions, it appears that patients localise and collect mobilised mucus easier compared to spontaneous cough. Further, it appears that the use of SCT may help avoid increased obstruction in hyper reactive airways.

Considerations

SCT can be used according to individual need and beneficial effect. The technique is developed to balance lung volume, cough effort, pressure and expiratory flow. Furthermore, to work together with other techniques, strategies and therapies to enhance clearance of bronchial secretions. Since patients have different presuppositions and needs, individual adaptations are required, including choice of techniques, strategies and treatment duration. As for treatment in general it is essential that patients learn how to control cough and that the following factors are considered, e.g. presence of any increased cough sensitivity reflex, bronchial hyper responsiveness, gastroesophageal reflux (GER), airway wall instability and the need for expiratory resistance.

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Postural Drainage and Percussion – In Process

Positioning – In Process

High Frequency Chest Wall Oscillation – In Process

3

Inhalation Therapy

Inhalation therapy has become increasingly important in pulmonary medicine. Sufficient intrapulmonary dose is a prerequisite for the expected result, but to what extent it is obtained differs a lot in clinical as well as scientific work. The three different types of aerosol delivery devices; pressurised metered-dose inhaler (pMDI) ±spacer, powder inhaler and nebuliser system, can all be used to deliver the most conventional drugs, while some of the recently developed drugs for inhalation have been introduced only as a powder. Depending on the target for the inhaled drug, the aim is always for even deposition within the lung in order to optimise the chance for success. The achieved deposition pattern varies among patients and inhalation device, but can be influenced.

There are many important factors which influence the results of inhalation therapy. These include:

- choice of device
- inhalation technique
- treatment strategy
- adherence with the treatment

Choice of device

If expected effects from the inhalation therapy are not achieved, the tendency is to increase the prescribed dose. Instead, making sure that the choice of device suits the individual is mandatory and regular check-ups of the inhalation technique should be a routine, as well as when a new drug is prescribed. Some important factors regarding the devices are:

- delivered aerosol quality, preferably for the prescribed drug
- residual volume of the nominal dose (drug left un-aerosolised in the device)
- delivery flow velocity
- proportion of the aerosolized volume accessible for inhalation
- interface used i.e. mouthpiece or mask

All factors will not be discussed in this chapter.

Inhalation technique

Aerosol is transported into the lungs and airways with inspired air to ventilated parts of the lungs. The more diseased the airways are, the more affected the breathing pattern and heterogeneous the ventilation distribution becomes. This individualises the intrapulmonary dose and increasingly deteriorates the deposition pattern. Undesired side effects may appear more frequently from extra-thoracic deposition. Depending on the drug target, it may be that the more impaired the ventilation distribution is, the more significant the impact working with optimising inhalation technique may have.

The total or local intrapulmonary drug dose in clinical and scientific work is difficult to measure. Concentration in evacuated secretion does not inform us about the intrapulmonary dose and deposition pattern. It may even be the opposite – the more concentration in the evacuated secretion, the less reaches more peripheral airways.

The ventilation distribution is dependent on lung volumes and air flow velocity. A low inspiratory flow velocity results in less impaction of aerosol in the throat, larynx and central airway bifurcations. A low velocity also reduces air flow turbulence which minimises impaction of aerosol in destructed airways, bronchiectasis, cysts and around secretion lumps. The inspiratory flow velocity is very high around larynx in a distressed or screaming child. Most of the aerosol deposits there and almost nothing reaches the lungs. The inhalation technique needs to be instructed and trained specifically with each device and every patient. The importance of individualized controlled inhalation (ICI) is frequently discussed with regard to administration of drugs in clinical trials and in clinical routines. Repeated review of technique is required, as technique deteriorates with time.

The young child

Inhalation therapy should always be delivered via a mouthpiece where possible, therefore gentle use of a mouthpiece should always be tried as a first alternative. Small children can accept using a mouthpiece early in life. The baby or toddler sits on the adult's lap, leaning back with the head supported. Parents are instructed and trained how to carefully hold the child´s head, the mouthpiece in the mouth and the index finger gently under the nose to close the nostrils. If working cautiously and with respect, the little child often cooperates better inhaling through a mouthpiece since they are used to having something in the mouth. The obstruction of the child's view and the risk for aerosol deposition in the eyes is less and the child does not need to defend itself. However, closing

the nostrils cautiously is crucial, otherwise they will suck what is in the mouth. When using a mouthpiece, the aerosol flowing out of most nebulisers is interrupted during inhalation through the mouth. This is a clear indication of inhalation through the mouth when evaluating the inhalation therapy and it gives good feed-back during the training.

When using a mask is inevitable, it is mandatory to find one that fits well, is of adequate shape and size and provides a good seal to the child´s face. This is critical for the intrapulmonary dose and deposition pattern.

However, leakage is common which is frustrating for the child since it obstructs the child's visual field and there is a risk of aerosol deposition in the eyes which can cause side effects. However, forcing a mask on the child´s face is uncomfortable and scary for the child. In a wriggling and screaming child the breathing pattern and inspiratory flow velocity are affected, resulting in even poorer intrapulmonary dose and deposition pattern. The wriggling increases the leakage which augments forcing the mask even further and a vicious circle is started.

When inhaling through a mask aerosol is deposited around the mouth and nose which many find uncomfortable, and inhalation may merely be through the nose. Therefore, a lot of the inhaled aerosol is deposited on the nasal mucous membrane, especially when the droplets are relatively big and/or the inspiratory flow velocity is high. Evaluating the effect of the therapy is difficult since the inhalation technique may be the reason for failed response, rather than the drug itself. Inhaling through a mouthpiece with a laminar flow allows the aerosol to reach intrapulmonary airways to a greater extent and must be used as early as possible.

Handling the device

The patient and/or parents have to learn how to handle the chosen device, to be instructed and given the opportunity to practice. In the open care clinic and if possible also when admitted to hospital, patients preferably bring their own device or use the same kind of device as at home. Hospital specific infection control guidelines regarding inhalation devices must be adhered to. They are asked to handle it and inhale as they usually do at home and adherence and the inhalation technique are evaluated. If needed, we optimise the technique or change the device to another type, if possible. Bringing their own device also gives an indication as to their cleaning procedure and an opportunity to discuss cleaning routines. There are many important factors to consider for the different types of inhalation devices.

Pressurised Metered Dose Inhaler with spacer

There are several different kinds of pMDI, most popular in general is still the pMDI that is manually actuated at the start of a slow, deep inspiration. Successful inhalation includes many steps that can go wrong. Most of the aerosol will deposit in the oral cavity if the patient can´t manage to coordinate actuating the dose with having started to inspire and to continue the slow inspiratory flow. The breath actuated pMDIs obviate the coordination problem. However, the other errors are still there. The pMDIs deliver aerosol with a high flow velocity and a big amount of the aerosol is deposited in the mouth and pharynx. Another alternative is a spacer that is added to the pMDI. The spacer withdraws the coordination problem and acts as a reservoir from which the patient can inspire slowly, which facilitates the intrapulmonary deposition. The droplets will start to sediment immediately after having been released into the spacer, a short delay between actuating and inhaling is important. However, the biggest droplets that would not reach the airways, rather depositing on the tongue where they cause side effects, still are deposited in the spacer instead. Among the different spacers on the market, the valved holding chamber is the most frequently recommended. Most pMDIs are not equipped with dose counting.

Instructions for using a pMDI with a valved holding chamber:

1) sit upright or stand
2) take the cap off the pMDI
3) shake the device
4) attach the pMDI to the spacer
5) put the mouthpiece between the teeth, lips around
6) put the tongue underneath the mouthpiece, if possible
7) actuate the pMDI once
8) expire more than usual
9) inspire slowly, deep
10) hold the breath for 5-10 seconds, if possible
11) relax and expire
12) inspire through the device a second time to ensure the dose is inhaled

If the prescribed dose consists of more than one actuation of the pMDI at each treatment occasion, repeat 3-12. When finished, separate the pMDI from the spacer and put the cap back on the pMDI. When new, the plastic spacer is often electrostatic due to the plastic. The electrostatic charge is minimised by cleaning and priming the spacer with a household detergent. Instruct the patients as recommended in the guideline that comes with the device. Due to the spacer being bulky and difficult to carry, it works best with drugs inhaled routinely at home.

There are those who advocate using a spacer for all inhalation of corticosteroids in order to reduce the side effects in the mouth.

A pMDI with extra-fine droplets results in a bigger lung dose for small children. A little child does not hold the breath, but takes several breaths in a row. A small volume valved holding chamber takes less time to empty and suits the little child better due to small tidal volume and no ability to inspire deep on instruction. The valve must be close to the mouth to ensure that each breath contains as little "dead volume" (without aerosol) as possible.

Dry Powder Inhaler (DPI)

There are two different types of DPI. One type needs to be loaded with a capsule to be punctured once before each administration, the other is preloaded with up to 200 doses where each dose is prepared immediately before being inhaled. There are many different DPIs on the market and each has its own specific preparation procedure. Most are equipped with dose counting and a sign when close to empty.

Instructions for using a DPI:

1) sit upright or stand
2) take the cap off the DPI
3) prepare the device (in detail for the given device)
4) expire more than usual, away from the DPI
5) hold the breath while placing the mouthpiece in between the teeth, lips around
6) put the tongue underneath the mouthpiece, if possible
7) inspire immediately resolute and deep
8) take the DPI out of the mouth immediately, turn it away
9) hold the breath for 5-10 seconds, if possible
10) relax and expire
11) inspire through the device a second time, re-start from 4) to ensure the dose is inhaled

If the prescribed dose consists of more than one preparation at each administration occasion, repeat 3-11. When finished, put the cap back on the DPI. A dose can consist of up to 10 capsules. Some DPIs have a mouthpiece with a shape that makes it impossible to put the tongue underneath, perhaps even to put it properly between the teeth. It is not possible to expire through a DPI due to the moisture it causes inside the device, which will capture the powder drug before it leaves the device during the following inspiration and it will never reach the airways. The DPIs are breath-actuated, there is no coordination problem between inspire and actuate the dose. However, some patients have great difficulties to differ between inspiration and expiration. Although able to learn how to expire away from the device it may still be difficult for them to hold the breath while placing the mouthpiece in the mouth, which means they will start exhaling in the device anyway.

A resolute inspiration through the device is required when using a DPI to free the powder and to de-aggregate it into optimal aerosol quality for intrapulmonary deposition. The high inspiratory flow may mean increased deposition in pharynx. How resolute the inspiration needs to be varies between devices due to design and inspiratory resistance. High resistance need lower inspiratory flow than devices with low resistance. A device is available on the market to check the inspiratory flow for each type of DPI, a good help when choosing device and for training the technique. Patients who have difficulties to follow instructions, have too poor lung function, pain or dysfunctional inspiratory muscles may not generate an as resolute inspiration as needed and a DPI is not suitable, as well as for small children. Although discussed, we still don´t know to what extent a sufficient but weak inspiratory flow and/or short breath affects the residual volume or the de-aggregating of the powder into optimal aerosol quality. To what extent an inspiration can be too resolute and thereby become negative for the intrapulmonary deposition is even less discussed. Not the least if the effect would be equal in all the DPIs. But, the inspiratory flow may become turbulent in the opening of the mouthpiece, resulting in a lot of the dose being deposited already in the oral cavity. Most DPIs should not be cleaned. They must be protected from damp. If the inside become moist and it is stored with cap off to dry, humid environment must be avoided.

Nebuliser system

There are three different types of a nebuliser system; jet, ultrasonic and mesh. The delivered aerosol quality and inhalable proportion of the nominal dose differs a lot between different capacity of the devices and different design of the nebuliser. Many nebuliser systems are either time efficient or can produce a good aerosol quality, while some can do both. Usually the better capacity, design and the more reliable the more expensive they are. Some nebuliser systems are equipped with a chip that contains information about the use (adherence) of it. One component of a nebuliser system must not be replaced with one from another model since that will influence its performance and quality of aerosol. The nebuliser part of the system should be replaced regularly, often annually or more often if time for aerosolizing clearly increases. Some drugs are more difficult to aerosolize than others and require a device with more capacity.

Instructions for inhalation with a nebuliser system:

1) assemble and prepare the nebuliser (specific for each model)

2) load the nebuliser with the drug
3) sit upright elbows on a table or backward leaning with support for back and shoulders
4) put the mouthpiece between the teeth, lips around
5) put the tongue underneath the mouthpiece, if possible
6) breathe normal, abdominal breaths
7) slow inspirations
8) relaxed expirations

Patients can be instructed to expire and inspire a little deeper occasionally, if possible. However, there is less use for that in some nebuliser systems, since they are equipped with a chip and a programme that individualize inhalation to only the beginning of each inspiration. This is based upon information about the size of the TV at the start of the treatment or on a mean of the size of the last 4 breaths. The inhaled proportion of the nominal dose may be bigger, but some devices stop aerosolizing before emptied to make sure the dose is not increased.

Most nebulisers produce aerosol continuously and a big proportion of the nominal dose is lost into the room or a filter during expiration. Some nebulisers are breath enhanced and less volume is lost, there are breath actuated, then aerosolizing a certain volume takes longer and the inhaled dose may be bigger. The residual volume (left non-aerosolised in the device) varies a lot (≤0,3 ml - ≥1,5 ml) between nebulisers depending on design. Patients are instructed to clean the nebuliser part after each treatment session and to disinfect the nebuliser once a day. Follow guidelines that come with the device. Cleaning and disinfection takes a lot of time every day.

Treatment strategy

The expected effect of an inhaled drug is dependent on to what extent it reaches the target. The target varies between the drugs.

- The bronchodilators are meant to treat or prevent bronchoconstriction. If deposited in the central airways it is easily absorbed and transported to the blood supplied target. If prescribed, it is inhaled before the airway clearance therapy, before inhaling another drug that may be bronchi constructive or before physical exercise.
- Most drugs meant to affect mucus viscosity or facilitate mucus clearance are inhaled before the airway clearance to make the therapy easier. But the aerosol is deposited on the top layer of the secretion and in patients with big amounts most of the inhaled drug is immediately evacuated. To evacuate the easiest secretion lumps before starting on the inhalation can be useful. If possible the optimal would be to inhale during pauses in the airway clearance therapy, to gradually affect mucus further peripheral. Hypertonic saline that has a short time for onset seems to be possible to alternate with airway clearance therapy during a session in these patients. But due to the need of long time for onset this is not the case for all the drugs. Often rhDNAse that has a time for onset on ≥30 minutes is inhaled after airway clearance therapy during every-day life, in order to utilize its effect on the secretions left in the lungs.
- Anti-inflammatory agents need to be inhaled after airway clearance therapy. Although the target is blood supplied, the drug is not absorbed by the blood and not transported. It acts mainly where it deposits.
- The target for the anti-microbial drugs is the micro-organisms in the airways. Anti-microbial drugs must be inhaled after airway clearance therapy to deposit peripherally and with an as good deposition pattern as possible, in order to reach the micro-organisms still harboured.

Adherence

Adherence with treatment is essential. Prescriptions and recommendations that actually are possible to perform during every-day life are basic. Experience say that individual education and instruction, practising, follow-up, listening to the patient and parents and making agreements will all influence adherence. It is all regularly repeated as needs and time change.

Inhalation therapy and positive expiratory pressure

In different airway clearance techniques positive expiratory pressure is a tool that is employed with various physiologic aims expected to influence lung volumes, flow velocity and breathing pattern differently. A positive expiratory pressure can also be utilized by obstructed patients to handle dyspnoea and their hyperinflated chest, or in a completely different way in patients with atelectasis. Thereby depending on pressure level achieved with precise instructions the positive expiratory pressure can be used for various purposes. Utilized as a tool in an airway clearance technique in combination with inhalation therapy, it will temporarily give either a positive or negative effect on the intrapulmonary dose and deposition pattern for the single individual. Due to increased expiration towards the resistance, the tidal volume can be bigger with a better deposition pattern. But should the expiration become too slow, the intrapulmonary dose may decrease due to an increased inspiratory flow velocity. And, a temporarily increased functional residual capacity during breathing towards the positive expiratory pressure can decrease the deposition pattern due to decreased ventilation distribution. That is, for whom to combine the inhalation therapy and the positive expiratory pressure should be well thought-out. Combining the two as a routine cannot be recommended.

Inhalation therapy during NIV and mechanical ventilation

If patients are in need of non-invasive ventilation (NIV) or mechanical ventilation, aerosol can be administered with the help of a nebuliser while having the treatment. Where to put the nebuliser in the circle, close to the patient or not, has been a subject for discussion. The most recent results from studies on NIV seem to be that placing the aerosol device close to the turned off humidifier would be the best, to utilize the tubing as a kind of "spacer". A low inspiratory flow if possible, is always to be preferred during the inhalation.

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4

Physical Exercise

Physical exercise is known to be beneficial in CF with improved and maintained physical capacity and feeling of well-being reported, as well as decreased rate of progression of the pulmonary disease. Less musculoskeletal complications have been shown in the aging CF population if physically active. The European CF Society (ECFS) has published guidelines about physical exercise testing as it is a sensitive tool to identify early progress of the disease.

Aims of physical activity/exercise

The aims of physical exercise are to

- educate parents in physical handling of the child
- reduce rate of progression of the pulmonary disease
- stay physically fit; working capacity, muscular strength, mobility, posture
- maintain normal bone mineral density (BMD)
- preserve physical capacity in patients with impaired lung function
- develop body awareness and physical reference frames
- avoid musculoskeletal complication and pain in the aging population
- utilize effects on muco-ciliary clearance

Habitual physical activity – the base

Normal habitual physical activity allows physical fitness and muscular condition to develop if the rest of the treatment is sufficient. Therefore, patients should be actively encouraged to participate in physical activity from the beginning. Avoiding adding treatment time to the treatment burden is a major aim and the reason to engage in habitual physical activity together with parents and siblings, in school or as leisure activities with friends. To achieve normal habitual physical activity some factors are essential in the early and continuous contact between parents/patient and physiotherapist;

- parents need to be introduced to handling their child physically at diagnosis if possible, since a natural reflex may be to protect a diseased child from effort
- physical activity/exercise is practised at the CF centre from the beginning and regularly, continuously - not only discussed and recommended
- participating in physical activity/sports in school may need to be introduced
- parents and patients learn to differentiate between breathlessness and dyspnoea, and to manage it
- patients learn to manage cough

Carrying out physical exercise and/or sports some days per week equates to living a normal life in many cultures. When necessary, an experienced physiotherapist adapts physical activities to the physical condition. If desired, alternative physical activities and sport are worked out with the help of the physiotherapist.

Content of physical activity/exercise

Physical activity/exercise in pulmonary disease includes different components, either as prevention of dysfunction or rehabilitation of impaired physical capacity:

- **Working capacity training** includes moderately intense interval and endurance training and is performed in the same way as in the healthy population, although the first choice would be weight bearing exercises in order to achieve a simultaneous effect on **BMD**.
- **Muscle strength training** is performed as in healthy, but always includes exercises targeting postural muscles. Inspiratory muscle training is a natural part of general physical exercise training. But isolated inspiratory muscle training is beneficial in the less generally active patients.

- **Mobility training** in patients with pulmonary disease targets chest and back specifically, focusing on muscles involved in posture, auxiliary breathing and coughing. If including phases of breathing at low lung volume (decreasing FRC and lowering RV) in the airway clearance therapy, the mobility training of the chest wall is performed simultaneously.

Physical exercise – in general

Preserve

Physical exercise aiming to maintain normal physical fitness is easily performed and less time demanding than trying to regain what has been lost. Exercise can be made fun for young children, enjoyable and acceptable for most adolescents and adults, when educated and motivated. Physical exercise training 3 times per week is considered sufficient and sport may constitute a good base. Young children carry out the physical exercise or training at home and or out-door. For the older children/adolescents and adults this is to a greater extent dependent on what surroundings allow. Performing physical exercise in a gym or fitness centre suit some and mean all advanced equipment/tools/devices can be used and educated staff are available. Although having severe pulmonary disease, patients may maintain reasonable physical fitness with physical exercise training. If patients desaturate the training may need to be carried out in an out-patient clinic with supplemental oxygen and/or non-invasive ventilation under supervision of an experienced physiotherapist.

Regain

Rehabilitation can regain muscle strength, endurance and working capacity to a great extent, but chest wall mobility that has been lost is more difficult to regain. A decreasing BMD may be possible to stabilise but difficult to regain in the adult population. Physical rehabilitation does not necessarily improve a decreased lung function, although the progress rate often slows down. But improved physical fitness is vital in daily life.

The training programme is preferably worked out together with the patient/parents and becomes a result of working collaboratively. The programme is up-dated as required, depending on adherence or change in physical condition.

Physical activity/exercise for airway clearance

Physical activity/exercise is reported by many to have a mucus mobilising effect. This can be due to different factors such as:

- breathing pattern reaching closer to TLC due to increased TV in mild pulmonary disease and temporarily hyperinflated lungs, increased FRC in more obstructed patients during physical exercise may re-open closed airways, "get air behind mucus", allowing mobilisation of secretions from otherwise not reachable airways.
- reduced Sodium reabsorption from the airways is reported to preserve water on the airway wall mucosa during physical exercise in CF. Thereby muco-ciliary clearance is facilitated, with mucus loosening more easily from mucosa being utilized as the basis in the airway clearance therapy.

Most often physical activity/exercise alone is not a sufficient airway clearance technique. But it can work for many patients when interspersed with a technique transporting mucus to the central airways followed by controlled coughing for evacuation.

Carrying out physical exercise as the basis in an airway clearance technique is time efficient since it comprises two parts of treatment simultaneously. For infants, toddlers and young children it may be fun and stimulating, exercises used are easily varied.

Physical exercise test

Regular physical exercise tests are recommended as a more sensitive tool to assess progress of the disease than lung function tests at rest. The ECFS has produced guidelines for different kinds of tests, depending on what resources are available. It is essential that the same test is performed regularly, in order to be able to compare changes over time. Patients start to do the test when ≥10 years old, even with limited resources, but without chronic or recurrent pain. Osteoporosis influences individual choice of test. Patients can start younger to familiarize and get motivated. The tests discussed are either

- cardiopulmonary test (CPET) such as aerobic exercise capacity (cycle ergometer or treadmill) with a progressive protocol (ramp, each step 1 min), measuring exercise ventilation characteristics and exercise circulation characteristics, or
- "field tests", such as

- 6-minute walking test (6MWT)
- 6-minute walking distance (6MWD)
- 3-minute step test
- shuttle tests

All tests have disadvantages. The CPET is expensive, requires expert supervision and expert interpretation, while the 6-minute tests are not standardized, the 6- and 3-minute tests are not necessarily completed and the shuttle tests appear in many versions.

Considerations:

- Physical exercise should be looked upon as preventative care, starting early in life rather than rehabilitation. Malnourished patients should not do working capacity and muscle strengthening training until the energy balance has been corrected. Nor should patients during an exacerbation or those with hay fever.
- mobility exercises can always be carried out
- if needed patients pre-medicate with a bronchodilator
- drinking adequate amount of water with salt and minerals in connection to physical activity/exercise is essential
- patients who desaturate to SpO2 ≤90% during moderate physical exercise should do the exercise with supplemental oxygen to avoid negative effects on the heart. Often SpO_2 92% is considered sufficient during exercise, more absolute terms may be recommended at each specific CF centre
- patients with CF related diabetes learn to control glucose levels in conjunction to physical exercise
- patients with gout or arthritis are recommended to perform physical exercise but to respect the pain when training. Mobility training is always possible to a certain extent

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Musculoskeletal Screening and Assessment Tools for Patients with Chronic Respiratory Conditions

Introduction

Patients with chronic respiratory conditions sometimes report musculoskeletal (MSK) problems during Respiratory Outpatient visits and hospital admissions that may cause difficulties with the ability to carry out airway clearance therapy and regular physical exercise. Commonly these may occur because of sprains and strains during physical activity; sub-optimal postural alignment; deconditioning resulting from lack of regular physical exercise; loss of muscle mass/ imbalance during exacerbations; or issues related to frequent/prolonged/forceful coughing.

A MSK screening tool and a MSK assessment tool have been developed to facilitate a physiotherapist to identify and appropriately manage MSK complications.

Manchester Musculoskeletal Screening Tool: developed for use in cystic fibrosis (Jane Ashbrook and Julia Taylor)

The aims of developing a musculoskeletal screening tool were to provide a quick and easy tool to collect baseline data annually on patient reported pain, urinary incontinence and postural problems. To address patient under reporting of CF related MSK disorders. To proactively identify MSK problems facilitating early intervention. (4) To enable patients to make informed decisions regarding the management and prevention of CF related MSK disorders (in a patient population that has a high burden of care). To provide a matrix to signpost appropriate care pathways including those who need MSK specialist assessment and individualised management plans.

After appropriate training it is a quick and easy assessment tool for any physiotherapist to use irrespective of any specialist MSK training.

It incorporates validated outcome measures for pain and urinary incontinence i.e.; Visual analogue scale, Short Form McGill pain questionnaire and the International consultation in incontinence questionnaire.

Increased thoracic kyphosis is common in the CF population and the tool includes a thoracic kyphosis and thoracic movement screen to quickly identify any problems.

The management matrix for identified problems provides a framework to ensure that there are evidence-based care pathways for commonly identified problems and identifies those who need referral for MSK specialist assessment and treatment.

Copies of the form can be requested from: Manchester Adult CF Centre, Manchester, England. Julia.Taylor2@UHSM.nhs.uk; SMU-TR.CFPhysiotherapyTeam@nhs.net

The Alfred Musculoskeletal Assessment Tool for Patients with Chronic Respiratory Conditions (Brenda M. Button, Lisa M. Wilson, Mary Massery)

The aims of developing a musculoskeletal assessment tool are to provide a framework to systematically measure musculoskeletal (MSK) and postural function; to identify MSK problems; to educate patients about normal posture and physical function and the importance of preventing problems; to instigate early treatment to normalize function and prevent chronic changes and pain.

The form includes an anterior and posterior body pain chart and demographic information. This is followed by sections on: postural alignment, shoulder and scapula positions, thoracic, lumbar and hip mobility, angle of pelvic tilt, hamstring and calf length and measurements of upper, middle and lower chest expansion and breathing patterns. Lastly there is a question about whether the presenting MSK problem limits participation in regular physical exercise.

Many of the items are rated as yes or no; changes are graded as nil, mild, moderate, or severe; and some items require the use of a tape measure. The form is completed with a summary of the main problems and an individualized treatment plan. An explanatory sheet accompanies the assessment form on "how to" carry out the measurements in a standardized way.

The short form is useful in a busy Outpatient Clinic visit when a patient reports musculoskeletal problems. After a thorough history and completion of the short assessment form the problems and likely causes are documented as

a main problem list and the treatment plan is established. This takes approximately twenty minutes. The longer full assessment form includes additional measurements and assessments providing further information and takes around 45 minutes.

Copies of the form can be requested from The Alfred Physiotherapy Department. Melbourne, Australia from b.button@alfred.org.au; l.wilson@alfred.org.au

Treatment

Musculoskeletal problems should be managed as part of the CF multi-disciplinary team and specialist MSK input should be incorporated into the care plan of all CF patients. That may be screening, prevention, assessment, treatment or any combination. Intervention should be specific to the needs and priorities of each individual patient at any given time.

People with CF should be offered a proactive management plan in an attempt to prevent postural problems and common CF related MSK disorders. This may include stretching and strengthening, taking part in regular sporting activities and using cognitive learning to develop postural awareness and encourage lifelong good postural habits.

When problems are reported or identified, an individualised assessment should be offered to take account of the many contributing factors. There is no consensus on the most effective treatment strategies however, manual techniques, specific exercises and ergonomic advice should be considered. A stretching programme may be useful to achieve changes in posture; however, this should not be considered in isolation from functional movement and muscle activation. If necessary, early referral to a musculoskeletal specialist is recommended to provide optimal and individually tailored management.

When acute injuries occur, the patient should receive prompt assessment and treatment to enable a timely return to function and respiratory physiotherapy treatments, including airway clearance, sport and exercise.

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Non-Invasive Ventilation

Non-invasive ventilation is an important component of the care of patients with advanced lung disease. Its potential is not only in the management of respiratory failure and bridging to lung transplantation but extends to enhancing airway clearance regimes, promoting independence and physical activity programmes and can be pivotal to symptom control at end of life. As lung disease advances, the emergence of uncomfortable symptoms such as poor sleep quality, breathlessness and fatigue can significantly compromise quality of life.

Who might benefit from NIV?

- Type II respiratory failure (acute or chronic) – NIV might help to reverse or stabilise hypercapnia and hypoxaemia (Hodson et al, 1991)

- Nocturnal hypercapnia – NIV can improve sleep and reduce symptoms of respiratory failure such as headache and fatigue (Gozal, 1997)

- Increased work of breathing and fatigue during airway clearance – NIV can offload the respiratory muscles and reduce hypoxia if used as an adjunct to airway clearance (Dwyer et al, 2010; Holland et al, 2003)

- Waiting for transplant – time from listing to transplant is hard to predict, NIV can help stabilise and 'bridge' a patient to transplantation (Madden et al, 2002) or assist the rescue from a severe pulmonary exacerbation where endotracheal intubation is not in the patients interest

- Severely limited exercise capacity – NIV can support the breathing and reduce the sensation of breathlessness during exercise and activities of daily living

NIV for airway clearance

Effective airway clearance is often difficult to achieve with advanced lung disease due to increased ventilatory demands, hypoxia, airway irritability or instability and fatigue. NIV can unload the respiratory muscles during airway clearance techniques this, combined with improved alveolar ventilation, reduces dyspnoea and improves oxygen saturations (Cecins et al, 1999; Dwyer et al, 2010; Holland et al, 2003). There is Little evidence to suggest that the volume of sputum cleared is improved with the addition of NIV however, there is theoretically greater potential for longer or more frequent airway clearance sessions to be tolerated.

The settings you choose for airway clearance assistance should take into consideration the following:

- The interface of choice – you will notice less leak with a full face mask however, it is often much easier to expectorate during an airway clearance session by using a mouth piece or nasal mask. Alternatively, if using PEEP, it can be more comfortable for some to cough with a face mask insitu to assist airway stability

- Additional humidification – high ventilatory flows and supplementary oxygen can be very drying to the airways and therefore additional humidification should be considered for those established on NIV (Holland et al, 2007)
- Increasing the IPAP (inspiratory positive airways pressure) by at least 2cmH2O (from resting settings) in order to support a deeper breath and encourage increased thoracic expansion

- Reducing the back up rate and adjusting the inspiratory flow (if possible) to allow time for adequate inspiration and to allow an inspiratory hold perhaps and therefore encourage the expansion of slow to fill lung units

- Combining with airway clearance techniques – the ACBT can be very effectively mimicked using NIV and may improve tolerance to airway clearance in advanced disease. Consider increasing the IPAP for the TEE section, encourage adequate rest in between the active parts (BC) and FET can be performed on or off the NIV interface. Alternatively, if using a different airway clearance techniques such as AD or PEP, the NIV can be used for recovery post coughing.

Exercise and NIV

Where treatment and symptom burden is high, maintaining functional independence, muscle bulk and exercise tolerance can be challenging. In clinical practice, where significant dyspnoea on exertion limits functional and exercise capacity, NIV can support the lungs to respond to the demands of exercise by enabling an increase in minute ventilation.

NIV settings are likely to need adaping during exercise to allow the patient to breathe more quickly, deeply and comfortably. Exercise sessions should be closely observed and adaptations in IPAP, inspiratory trigger, inspiratory time, rate and entrained oxygen may be required. A variety of interfaces can be considered for use during exercise with patient confort and clinical stability a priority.

NIV at end of life

NIV is a useful modality for managing the symptoms of end stage lung disease (Philip et al, 2008), considerations should include:

- Frequent but short airway clearance sessions using NIV may be required to maxmise symptom relief and minimise treatment burden
- Poor tolerance of NIV as a result of advancing disease or acute exacerbation will likely require adjustment of NIV settings and/or the use of a volume assured mode to maximise ventilation potential in the presence of severe airways obstruction
- In the case of NIV dependence, consider setting the NIV on a trolley to allow ease of use for activities of daily living and mobilisation and in addition consider:
    - Delivering essential nebulised medicines through the NIV circuit via a t-piece connector
    - Providing a range of interfaces to avoid pressure areas and enable plenty of opportunities for eating, drinking and communication
    - Providing two NIV machines which can be rotated to ensure adequate function and reduce the risk of equipment failure

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Critical Care

Admission to the critical care or intensive care unit is associated with a poor prognosis in CF. Factors associated with a poor outcome include prior colonisation with Burkholderia cepacia complex, rapid decline in FEV_1, and severe exacerbation (Ellafi, 2005). Positive outcomes are associated with potentially reversible conditions such as the acute management of haemoptysis or pneumothorax (Sood et all, 2001) and post-operative management. Endotracheal intubation (mechanical ventilation) is associated with a poor prognosis (Texereau et al, 2001; aeration, 2010). However, the outcome of treatment with non-invasive ventilation (NIV) is good (Efrati et al, 2010; Vedam et al, 2014) and many centres may manage NIV in high dependency or ward areas. Extracorporeal membrane oxygenation (ECMO) is used on critical care as a salvage strategy in CF patients with respiratory failure and is being increasingly used as a bridge to lung transplantation (Reid & Bell, 2013; Shafii et al, 2012). The use of ECMO has emerged as a promising intervention that can avoid invasive ventilation, and allows patients to eat, ambulate and undertake airway clearance while awaiting lung transplantation (Rehder et al, 2011; Rehder et Al, 2012; Jones et al, 2013).

There are no published studies of physiotherapy management of the intubated and ventilated patient with CF. However, the following should be considered (NICE, 2009):

- Ensure regular airway clearance is continued, and optimise humidification
- Ensure good positioning for optimal ventilation and drainage of secretions
- During the patient's critical care stay and as early as clinically possible, perform a short clinical assessment to determine the patient's risk of developing physical and non-physical morbidity
- For patients at risk, agree short-term and medium-term rehabilitation goals, based on the comprehensive clinical assessment. The patient's family and/or carer should also be involved
- For patients at risk, start rehabilitation as early as clinically possible, based on the comprehensive clinical assessment and the rehabilitation goals. Rehabilitation should include:
- Measures to prevent avoidable physical and non-physical morbidity, including a review of previous and current medication
- An individualised, structured rehabilitation programme with frequent follow-up reviews. The details of the structured rehabilitation programme and the reviews should be collated and documented in the patient's clinical records
- For patients on ECMO ambulation and rehabilitation should be completed as able by physiotherapists trained in managing patients on ECMO
- To ensure optimal management there needs to be excellent communication and liaison between both the critical care and CF physiotherapy teams and the wider MDT'
- The use of timetables to protect airway clearance and rehabilitation time for patients may be beneficial.

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Cystic Fibrosis and Lung Transplantation

Bilateral sequential lung transplantation (BSLTX) is a valid therapeutic option for patients with cystic fibrosis (CF). The International Society for Heart Lung Transplantation (ISHLT) data shows that CF is the 3rd most common diagnosis for lung transplantation (LTX) (16.2%) and has the best survival rate (60% at 5 years). The aim of LTX is to improve both survival and quality of life.

One of the dilemmas surrounding LTX for CF patients is the time to refer to a lung transplantation centre. Lung transplantation performed as an elective procedure has superior outcomes than as a lifesaving procedure.

ISHLT candidate selection guidelines state that LTX should be considered for CF patients who have a 2 –year predicted survival of<50% and have a NYHA class III or IV. In addition, a fall in FEV1 to 30% or a rapidly declining FEV1 in a patient with non-tuberculous mycobacterial(NTM)infection or *B cepacia complex,* the development of pulmonary hypertension (defined by PAP>35mm Hg) or an episode of acute respiratory failure requiring NIV should stimulate a referral to a transplant centre.

In order to be accepted onto a waiting list for LTX, CF candidates must demonstrate compliance with their medical, pharmaceutical & exercise regimes. Abstinence from smoking and illicit drug usage is also a requirement.

Surgery

The surgical technique involves removing the diseased lungs sequentially via one of the two incisions. A traditional clam shell incision involves 2 large anterior thoracotomies plus a transverse incision of the sternum at the level of 5th rib. This has the advantage of giving a superior view to the mediastinum but has increased pain and deformity post LTX. The less invasive incision of 2 large anterior thoracotomies utilises the 5th rib space but can cause thoracic spine pain. Both these operative approaches can take up to 12 hours for completion. ECMO can be used as a bridge to transplantation in some centres.

Immunosuppression

Lifelong immunosuppression therapy usually consists of a calcineurin inhibitor, cell cycle inhibitor and a corticosteroid. Dosages of the immunosuppressant regimen are patient tailored. Surveillance for rejection is performed at regular intervals particularly during the first year via bronchoscopies. Episodes of acute rejection are treated with increased and/or additional immunosuppression. Antimicrobials are also commonly prescribed. Continuance of other CF related drugs should be continued post LTX.

The side effects of these medications can include osteoporosis, and increased risk of fractures. Other problems associated with LTX include early onset of lactic acid threshold, persistent changes in lung function and muscle weakness. Careful monitoring of the immunosuppression drugs will optimise renal and hepatic function.

Physiotherapy Role

1. Assessment

Physiotherapy assessment includes a full medical and social history including a compliance history with exercise & medical regimes. Functional exercise capacity should be assessed with six-minute walk test. Assessing 6MWD at 3 monthly intervals whist waiting for LTX is recommended as a 6MWD < 400m has been associated with poorer prognosis. Assessment of airway clearance regimes, O2 therapy will assist in the development of an exercise rehabilitation plan for LTX candidates.

2. Pre LTX

Whilst on the waiting list, physiotherapy should aim to optimise physical function and offer ongoing support. CF patients should continue all usual ACT and physical exercise programs. Maintenance of lean muscle mass can impart a survival advantage post LTX. Physio therapists can provide education about life after LTX as it is important to allay fears for both patients and carers. Any musculoskeletal problems that could limit post transplantation rehabilitation potential need to be fully assessed and treated prior to transplant.
Due to size matching, some smaller CF patients may wait longer for matched organs.

3. Post LTX

Both the ATS and ERS recommend that that training must begin within 24-48 hours after surgery, and focus on lung expansion, permeabilization of the airway, improvement in breathing pattern efficiency, recovery of both upper and lower limb range of movement, strength and ensure independent walking is achieved.
There are similarities to all post cardiothoracic surgery with the post LTX physiotherapy management.
Mobilisation commences in the ICU setting and the exercise program can commence while the patient is an inpatient.

Acute post LTX rehabilitation programs vary in duration and composition worldwide. These can range from in patient to outpatient or home-based programs. Exercise programs are transplant centre specific and are also dependent on each country's health care model. These programs are highly valued by the patients.
Post LTX rehabilitation programs vary in duration and structure world- wide but are recommended by ISHLT guidelines. These programs should include aerobic training and strength training components for both upper and lower limbs. Particular emphasis should also include postural re-education and core strength training.
The goal of rehabilitation should be to return patients to peer matched normal life activities including return to school, work, sport or community work.
Long term, the maintenance of muscular & bone health by a regular exercise regime & sport is essential to the recipients' quality of life over their long-term survival. Physiotherapists should aim to support and encourage these physical activity regimes.
4. Re transplantation
Re-transplantation is an option for those LTX recipients who develop chronic rejection. ISHLT registry statistics state that 20% of all re-transplants occur between 5-10 years post the first transplantation. Assessment for suitability and acceptance is centre dependent for re- transplantation.

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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 FEV1 less than 50% predicted (Penketh et al 1982, Flume et al 2005). Recurrence rates of 41% ipsilaterally (MacDuff et al 2010) and 46% contralaterally (Rich et al 1978) are reported.

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 sputum retention, which may delay re-expansion of the collapsed lung (Henry et al 2003). A recurrent pneumothorax requires more aggressive management including either a partial pleurectomy or a talc pleurodesis (Henry et al 2003). Pleurodesis can make transplantation more difficult as it may impede lung removal, however it is not an absolute contraindication to transplantation (Noyes and Orenstein 1992, Schidlow et al 1993, De Abreu e Silva 1996, Henry et al 2003).

The physiotherapy management of pneumothorax

There are no published data regarding physiotherapy management of patients with pneumothorax.

The following recommendations are based on expert opinion (Flume et al 2010) and clinician consensus.

Small Pneumothorax

Monitor respiratory status including shortness of breath; and cease PEP and other forms of positive pressure therapy. Review use of nebulised hypertonic saline and other mucolytic agents especially if they stimulate increased coughing. Ensure adequate humidification for ease of sputum expectoration. If the patient regularly uses Dornase Alfa, continue inhalation. Encourage effective huffing and gentle coughing. Reduce exercise intensity and avoid upper limb resistance exercises.

Large Pneumothorax

If the pneumothorax is undrained, cease physiotherapy treatment and liaise with the medical team. If the pneumothorax is drained: if patient uses PEP or non-invasive ventilation, consider cessation or reduction in pressures while draining and for at least 48 hours afterwards to avoid pleural fistula and risk of recurrence. Review use of nebulised hypertonic saline and other mucolytic agents especially if they cause unnecessary coughing. 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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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 (Brinson et al. 1998). Mild haemoptysis affects approximately 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). Increasing age is the greatest risk factor for massive haemoptysis (Flume et al. 2005, Brinson et al. 1998). Other risk factors include moderate to severe lung function impairment, concurrent infection with Staphylococcus aureus, vitamin K deficiency and diabetes (Hurt and Simmonds 2012).

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, 2004). For massive haemoptysis, the vessel may need to be occluded using bronchial artery embolisation (King et al. 1989, De Abreu e Silva 1996, 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

The CF Foundation Pulmonary Therapies Committee (USA) have developed guidelines for the management of haemoptysis based on expert opinion using a Delphi process (Flume et al. 2010).

Scant (<5mL) haemoptysis, first episode: seek medical review, continue with inhaled therapies (Flume et al. 2010). Modify airway clearance techniques: reduce the force of coughing, cease highPEP. If active bleeding, position in high side lying with the bleeding side down (Thomas 2003). Provide reassurance and education and do not withold non-invasive ventilation if warranted.

Scant haemoptysis, recurrent episode: continue with normal airway clearance regimen and inhaled therapies and normal exercise routine.

Mild to Moderate Haemoptysis (<250mls/ 24 hrs)

Seek medical review, cease percussion, vibrations, oscillatory PEP techniques and head down tilted positions. Consider active cycle of breathing, autogenic drainage and gentle huffing and coughing. Continue inhaled therapies in most situations (Flume et al. 2010). Ensure adequate humidification to ease sputum expectoration. Cease vigorous exercise and instead encourage low intensity exercise such as walking or cycling.

Massive Haemoptysis (>250mls/ 24 hrs)

Seek medical review (Flume et al 2010). If active bleeding occurs position the patient in high side lying with the bleeding side down (Thomas 2003). If the patient cannot tell where the bleeding is, position in upright supported sitting. Cease airway clearance therapy, hypertonic saline and exercise until the active bleeding has resolved. Cease other inhaled therapies if they are considered to provoke bleeding. Cease non-invasive ventilation. Once the active bleeding is contained, continue as per moderate haemoptysis.

Following bronchial artery embolisation

Ensure adequate analgesia and humidification. Gentle mobilisation following the surgeon / radiologist's advice, then gradually increase intensity of exercise. Airway clearance using the active cycle of breathing techniques or autogenic drainage are initially appropriate followed by the gradual re-introduction of the usual airway clearance regimen and more vigorous physical exercise.

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Physiotherapy during Pregnancy, Labour and the Post-Natal Period

Women with chronic lung disease such as bronchiectasis and cystic fibrosis have the same desire to be mothers as healthy women in the community. 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. There are few published data that relate to the management of pregnancy in women with chronic lung disease. 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, Lau et al. 2011) together with descriptions relating to clinical practice (Johannesson et al 1998, 2002). Women can have successful pregnancies following lung transplantation but the risks of organ rejection and death are high (Whitty 2010).

Physiological changes during pregnancy

The pregnancy hormones of relaxin, progesterone, oestrogen and cortisols result in laxity of ligaments in preparation for the birth process. These effects are greater in mutigravidae than primagravidae women. It takes approximately 3-6 months for the body to return to the pre-pregnant state after the birth (Beischer 1989). The maternal centre of gravity shifts posteriorly during pregnancy to accommodate the increased abdominal size resulting in changes in postural alignment and gait in later pregnancy. As thoracic and lumbar curves increase, so does the strain on the vertebral joints. Back pain is common affecting more than 50% of women during pregnancy. Back pain is sometimes accentuated in women with CF and other chronic lung diseases especially during acute lung exacerbations and increased coughing. Cardiac output increases by approximately 40% by 20 weeks gestation. Blood volume increases by approximately 40%, with a peak at 30 weeks gestation, secondary to new placental circulation. General vasodilatation occurs. This results in increased cardiac work during pregnancy (De Swiet et al. 1991). Progesterone stimulates the respiratory centres of the brain to produce hyperventilation early in pregnancy. In late pregnancy secondary to hormonal changes relaxation of smooth muscle occurs in the tracheo-bronchial tree leading to a decrease in total pulmonary resistance. This may be beneficial to women with more obstructive lung disease. However, at the end of pregnancy, residual volume decreases secondary to elevation of the diaphragm. In the third trimester, especially with multiple foetuses, the enlarged uterus pushes upward and outward. The high abdominal wall tension raises the intra-abdominal pressure, even when the woman is upright. The diaphragm does not descend appreciably, and therefore the FRC remains reduced. Expiratory reserve volume and FRC decrease by 15%. This may contribute to difficulty in removing respiratory secretions.

Oedema in the lower limbs is common in pregnancy and is caused by the effects of progesterone. When standing for long periods, gravity causes venous engorgement further exacerbating the problem. Carpal tunnel syndrome is caused by oedema in arms and hands compressing the distal segments of the median and ulnar nerves – this generally occurs later in pregnancy but sometimes occurs as early as 16 weeks.

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 and other chronic suppurative lung diseases. 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 chronic suppurative lung diseases 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 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).

Airway Clearance Therapy during pregnancy in patients with chronic suppurative lung diseases

Head-down tilted postural drainage is not recommended during pregnancy because of the high prevalence of symptomatic and clinically silent gastro-oesophageal reflux (GOR) in adults with CF (Button et al. 2005). This is further compounded by the hormonal effects of progesterone during pregnancy resulting in a hypotonic lower

oesophageal sphincter together with the growing weight of the developing foetus pressing against the stomach. Techniques that exacerbate nausea should be avoided.

Airway clearance techniques suitable for use during pregnancy include:

- Active Cycle of Breathing Technique
- Autogenic drainage
- Positive expiratory pressure (PEP) therapy
- Oscillating positive expiratorypPressure therapy (OscPEP)
- Intrapulmonary Percussive Ventilation (IPV) including the Metaneb® Device
- Physical exercise as airway clearance therapy
- Effective huffing from different lung volumes avoiding dynamic collapse.

Mucolytic agents are commonly used as adjuncts to airway clearance therapy and their continued use during pregnancy should be reviewed with the woman's physician. Hypertonic and isotonic saline are used to optimize airway clearance therapy and lung health in women with chronic suppurative lung disease with viscous secretions. Most pregnant women with CF continue to use RhDNase (Pulmozyme) during pregnancy.

Positioning during Airway Clearance Therapy

Because of the physiological changes of pregnancy, upright sitting is usually the most comfortable position for airway clearance. Consideration should be given to positioning during ACTs to maintain a neutral lumbar spine for prevention and/or minimisation of urinary incontinence during treatment (see Chapter 9). Some women find left and right side lying horizontal or slightly head up to be more effective during ACT. The supine horizontal position should be avoided during the 2nd and 3rd trimesters because of the pressure of the foetus on the inferior vena cava which may decrease venous return and cardiac output.

Exercise during pregnancy

Pregnant women are advised to modify their physical exercise program. 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.

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 (Edenborough et al. 2008).

Physiotherapy during labour in CF and other chronic suppurative lung diseases

Pain, shortness of breath and low oxygen saturation during labour are reported in healthy women (De Swiet 1991). Thus women with chronic suppurative lung disease 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 and other chronic suppurative lung diseases with a normal vaginal delivery being highly desirable in order to minimize post-delivery complications (Johannesson 2002).

Physiotherapy after Caesarian section in chronic suppurative lung disease

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

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

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Physiotherapy for the Prevention and Management of Urinary Incontinence

Urinary incontinence is the involuntary leakage of urine. There are two types of urinary incontinence, stress incontinence and urge incontinence. The reported prevalence of urinary incontinence in girls with CF and women with CF and COPD ranges from 22% to 74% (White et al. 2000; Cornacchia et al. 2001; Orr et al. 2001; Nixon et al. 2002; Moran et al. 2003; Button et al. 2004; Prasad et al. 2006; Vella et al. 2009) in comparison with 13% in healthy women aged 18–24 years (Chiarelli P et al. 1999). There is limited literature in adult males with CF and COPD, with the prevalence reported to be from 8–15% compared to 7.5% in healthy men (Gumery et al. 2002; Browne et al. 2009, Burge, Holland, Button et al. 2011). It is not known whether the cause of urinary incontinence in CF and COPD is habitual chronic cough, increased coughing during acute exacerbations, demands placed on the pelvic floor during airway clearance therapy and physical exercise (all essential elements of recommended daily physiotherapy treatment) or underlying structural differences (Button et al. 2005). Age has been reported to have a strong positive correlation with the severity of stress urinary incontinence (SUI) symptoms (Vella et al. 2009). Studies reporting on the incidence of SUI in people with CF and COPD also report increased anxiety, depression and a negative impact on quality of life (Gumery et al. 2002; Nankivell et al. 2010, Burge et al. 2011).

It has been demonstrated that treatment of urinary incontinence in women with CF and COPD by a qualified continence physiotherapist with exercise, electrical stimulation, biofeedback and bladder training resulted in significant improvements in pelvic floor strength, reduction in leakage and improvement in quality of life which were sustained for at least three months after the completion of treatment (McVean et al. 2003; Button et al. 2005). Care should be taken when teaching pelvic floor exercises as evidence suggests 40% of women with incontinence incorrectly perform a pelvic floor contraction with verbal education alone (Thompson et al. 2003).

It is not appropriate in the paediatric setting to use invasive assessment procedures. Trans-abdominal real-time ultrasound has been used successfully to determine whether adolescent females have correctly learnt pelvic floor exercises without reported embarassment. (Depiazzi et al. 2008). Positive outcomes have also been demonstrated with surgical correction of severe SUI in women with CF (Helm, Dodd, Webb et al. 2008).

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).

Screening for urinary incontinence in the clinical setting

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 and COPD and other respiratory conditions characterized with coughing 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 and may require referral to a gynaecologist or urologist.

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Extracorporeal Membrane Oxygenation (ECMO) for Patients with Respiratory Conditions

Veno-venous (VV) extra corporeal membrane oxygenation (ECMO) is a growing area in the life support of patients with respiratory failure. VV ECMO is sometimes used as a bridge to recovery in patients with acute respiratory distress syndrome and as a bridge to lung transplantation in those with chronic lung diseases such as cystic fibrosis, bronchiectasis, pulmonary hypertension and interstitial lung disease. The availability of a dual lumen single ECMO cannulas (Avalon and Novalung Twinport) in the cervical region placed in the Internal Jugular Vein has revolutionized 'lung bypass' as it allows patients to be awake and able to participate in physiotherapy. Patients are able to carry out airway clearance therapy and physical exercise and are able to be upright and walking while waiting for lung transplant. Preserving muscle mass and general physical condition has been associated with better long terms outcomes after lung transplant.

How does ECMO work?

ECMO involves gas exchange and oxygenation of blood outside the body and can provide complete or partial support of the lungs and/or heart for patients who without this treatment are not likely to survive. An ECMO system consists of cannulas to take off venous blood from the body and to return oxygenated blood to the circulation. Outside the body the blood circulates through a pump, a gas exchange and temperature control device Life-threatening complications can occur while on ECMO including bleeding, thrombus formation, recirculation and sepsis (Lindstrom, Pellegrino & Butt 2009).

Awake ECMO

Until relatively recently patients treated on ECMO were sedated and paralyzed in the intensive care unit, breathing with the assistance of a mechanical ventilator, tube fed and unable to participate in physical activity. This approach led to the loss of muscle mass, strength, bone density and generally debilitation resulting in poor outcomes after lung transplant (Fan et al. 2009). Awake ECMO may lead to better outcomes after lung transplantation compared to traditional sedation and mechanical ventilation (Fuehner, Kuehn, Hadem et al. 2012). The published peer reviewed literature and conference presentations have been mostly based on descriptions of local experiences in relatively small case series. Today, many ECMO units aim for safe early mobilization when all patient clinical measures are deemed to be acceptable (Hodgson, Stiller, Needham et al. 2014). Generally, ECMO cannula placement in the upper body is preferred when considering mobilizing patients out of bed, standing and walking. No randomized controlled trials have tested this assertion to date

The aims of physiotherapy for patients on awake VV ECMO are: (1) to be safe in every aspect of treatment; (2) to clear lung secretions in those with chronic suppurative lung disease; (3) to preserve muscle mass and strength while patients progress to recovery or bridge to lung transplantation.

Contra-indications and precautions with mobilization out of bed

Patients should not be mobilized out of bed if ECMO flows and oxygenation are sub-optimal, if patients are cardio-vascularly unstable, if there is bleeding from the respiratory tract or thrombus formation.

Safety considerations when mobilizing patients on ECMO

All clinical parameters such as vasoactive agents, fraction of inspired oxygen (<0.6), oxygen saturations (>90), respiratory rate (<30/minute), blood pressure, heart rate, level of sedation and other considerations such as ECMO cannula placement, lines and other co-morbidities need to be considered prior to active mobilization (Hodgson, Stiller, Needham et al 2014). Consultation with the medical and nursing team are essential before mobilizing patients.

Airway clearance techniques for use in patients with awake ECMO

In researching physiotherapy practice as seven internationally recognized centres (Hannover, Germany; Paris (La Pitie Salpetriere and Hopital Foch), France; Duke University Hospital, North Carolina, USA; New York Presbyterian, New York and Toronto General Hospital, Canada and The Alfred Hospital, Melbourne, Australia) choice of airway clearance techniques (ACT) varied widely. If patients were intubated or had a tracheostomy then airway suction was employed to remove secretions. Sometimes broncho-alveolar lavage was carried out by members of the medical team for airway clearance and combined with airway suction. In patients not on ventilators, the range of ACT included: positive expiratory pressure therapy (PEP) with a range of devices available, oscillating positive

expiratory pressure therapy (OscPEP) using numerous devices, intrapulmonary percussive ventilation (IPV); assisted autogenic drainage and exercise as airway clearance therapy.

Mucolytic agents used in conjunction with airway clearance therapy

Patients on ECMO often have thick and viscous secretions. Adjunctive mucolytic agents such as saline (0.9%), inhaled mannitol, hypertonic saline (3-7%) and dornase alpha are sometimes used in conjunction with airway clearance therapy.

Planning and assessment of physical function before mobilizing on ECMO

Early mobilization is a high priority in all of the international units previously mentioned. Strong multi-disciplinary teams have been developed in these institutions to support safe and effective early physical activity in patients who are awake and able to co-operate. Adequate resources including well-trained staff and equipment to assist patients to be upright and active while on awake ECMO are required. Before safe mobilization a muscle strength assessment is necessary to ensure the patient is capable of mobilizing out of bed. The MRC and IMS mobility scores assist in determining whether the patient is suitable for mobilization out of bed.

Mobilization while on ECMO

Many patients have been critically ill for a period before being placed on ECMO and may already have lost muscle mass and strength. The patient may need a period of regular passive, assisted or active bed exercises in preparation for getting up. Some units use electrical muscle stimulation to preserve muscle mass and strength.

Sitting upright

Sitting up dangling legs over the side of the bed is followed by sitting out of bed in a chair for up to four or more hours a day. This incorporates all the benefits of being upright including lung expansion into the daily routine. Leg exercises are encouraged while sitting out of bed in the chair.

Standing

Standing with the assistance of a tilt table often precedes supported standing out of bed. While standing upright patients may be encouraged to do supported squats and heel raises as part of the leg strengthening program. Walking frames are often used to achieve safe exercise in standing.

Walking

A team of well-trained staff with one person coordinating the activity is necessary for safe mobilization. It is important that the ECMO cannula is supported by a trained professional to avoid movement and ensure optimal flows and oxygenation. Supported marching on the spot at the bedside is often the first walking exercise. When a portable treadmill is available this is placed at right angles to the patient's bed. The safe achievement of this activity requires on average 5 well-trained staff. The patient is assisted into sitting with legs dangling over the edge of the bed and then assisted into standing on the treadmill using the handrails for support and walks at a slow steady pace aiming for up to 20-30 minutes per session.

Walking out of the room requires an extra person to bring a wheel chair along in case the patient needs to sit down or be transported back to bed. All equipment needs to be battery powered. For safe mobilization each member of the team needs to be well trained and is required to concentrate exclusively on the equipment and parts of the activity for which they are responsible. The patient is usually supported with a walking frame.

Research to date consists mainly of outcomes in regional case series and retrospective reviews of patient charts. There is a lack of robust randomized controlled trials to determine safety, dosage, effectiveness and long-term outcomes relative to airway clearance therapy to prevent sepsis and exercise to prevent loss of muscle mass and general condition.

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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.