CYSTIC FIBROSIS – APPROACHES TO MANAGEMENT

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INTRODUCTION

Chest physiotherapy (CPT) forms an integral part of the treatment regimen for Cystic Fibrosis (CF). It is aimed at promoting mucus and mucociliary clearance. The mucus secretions in CF are thick and sticky because of the basic defect which affects chloride and water secretion. Recurrent respiratory infections cause an increase in the amount and density of the secretions. This leads to airway obstruction with subsequent infection and destruction of the airway walls and further impairment of mucociliary clearance. Chest physiotherapy is an attempt to alleviate this process and improve the quality of life for these patients.

Over the past 10 – 15 years a number of alternatives to the original postural drainage (PD) with percussion, have been proposed. The traditional CPT is both time consuming and uncomfortable. Its value is questionable in the milder cases of CF. Compliance with the regimen is generally poor and when the patients reach adolescence it becomes an even greater problem[1]. The newer methods take less time and can be performed by the older child without assistance.

This article presents the different modalities available at present. Although a great many studies have been performed using and comparing the various modalities, no one method has been proved to be the method of choice.

Since CF is an hereditary condition with symptoms frequently presenting from birth, the CPT has to be age appropriate. Most of the techniques discussed are suitable only for the older child and adolescent but it is particularly these age groups that present with compliance problems.

CHEST PHYSIOTHERAPY MODALITIES

Postural Drainage

Postural drainage (PD) is the conventional physiotherapy treatment for CF patients[2]. PD is defined as positioning the patient to allow gravity to assist the drainage of secretions from specific areas of the lungs. Percussion, shaking and vibration (manual and mechanical) are performed on the patient in the required position. Patient compliance with PD is poor because it is time consuming, uncomfortable and sometimes painful[3]. Cystic Fibrosis sputum is thick and tenacious, and the likelihood of it draining through the bronchi is probably minimal. PD in the head-down position increases gastro-oesophageal reflux, and diaphragmatic work is increased in order to push the abdominal contents against gravity on inspiration, thus further compromising respiratory function[4]. PD positions recommended are alternate side lying and sitting only. These positions are applicable to children of less than five to six years of age, or until one of the breathing techniques has been affectively mastered. During respiratory infection exacerbations, PD is the treatment of choice.

A recent study by Lannefors and Wollmers compared PD, Positive Expiratory Pressure (PEP) and exercise. Interestingly, the PD position to clear the right middle lobe actually cleared the left dependent lung. This indicates that gravity has little or no influence on mucus mobilisation and other mechanisms are involved. The effects of PD incorporating the Forced Expiration Technique (FET) was conducted by Webber *et al*[6]. The results showed significant improvement in large airways function and they concluded that the addition of the FET improves clearance of bronchial secretions in PD.

Active cycle of breathing techniques

The active cycle of breathing techniques (ACBT) is the most frequently utilised and recognised method in the UK[7]. The technique incorporates breathing control and relaxation, thoracic expansion exercises and forced expiration. The characteristics of FET can be described as a number of huffs from mid to low lung volumes, followed by a period of relaxed, controlled diaphragmatic breathing[8]. The latter is necessary to allow the airways to return to their resting calibre as they narrow following any forced expiratory manoeuvre. The FET can be explained using the concept of the equal pressure point (EPP)[9], which is the point where the pressure within the airways in equal to the pleural pressure. Downstream of the EPP, towards the mouth, the dynamic squeezing of airways allows secretions to be mobilised and cleared[10]. As lung volume decreases the EPP's move peripherally and a huff to low lung volume clears secretions from the peripheral airways[11]. Secretions mobilised to the upper airways is cleared by a huff from high lung volume.

The ACBT was described by Louise Lannefors at a recent CF Symposium[4]. ACBT should be performed daily, even with minimal chest symptoms. The technique is repeated until the huff becomes dry sounding and non-productive, or a rest interval is needed. Recommended positions for treatment are sitting and/or alternate side lying.

The cycle consists of the following:

- Relaxation and breathing control

(approximately one minute).

- Thoracic expansion exercises (4 or 5 deep breaths, emphasising inspiration, with passive expiration – encourages airflow through the collateral ventilatory system: pores of Kohn and canals of Lambert, increasing airflow in the smaller bronchi helping to mobilise secretions[12].)
- FET (huffs from mid to low lung volume, and breathing control).

Mobilised secretions are then expectorated.

Pryor and Webber[11] evaluated the FET with PD in comparison to PD and percussion in 24 CF patients. The results showed that sputum cleared in less time, and FEV increased after FET. Using the ACBT in PD position, Pryor *et al*[13] measured oxygen saturation, and concluded that there was no evidence of hypoxaemia during the procedure. ACBT offers the patient a means of clearing secretions without the assistance of another person or a mechanical device[14].

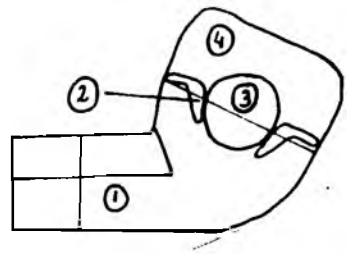
Positive Expiratory Pressure

The rationale for the use of positive expiratory pressure (PEP) is that it is a method which will open up and recruit the obstructed lung periphery[15]. PEP increases the airway transmural pressure in the lung - centrally and peripherally, which results in dilation of the bronchial tree. This promotes inflow of air behind obstructions through the bronchial route or collateral channels, reaching obstructed or collapsed lung periphery[15,16].

The treatment modality used is the PEP mask. It consists of a face mask and a one-way valve to which expiratory resistors are attached. The resistance is measured using a manometer to enable the individual to maintain a PEP of 10 to 20cm H_2O during mid expiration. The smallest diameter resistor which the patient can use comfortably for two minutes while simultaneously maintaining the correct PEP level is the suitable one. Treatment is performed in the sitting position[17]. The mask must be airtight over the nose and mouth. The patient inspires slowly and deeply through the mask. Expiration is slightly activated but not forced. This cycle is continued for 10-15 respirations, the patient then removes the mask and does FET to expectorate mobilised secretions[11]. Since some patients found the mask claustrophobic, a mouthpiece PEP device was designed and used by teenagers at the Red Cross Children's Hospital[18].

Falk *et al*[16] compared the techniques of A: PD and percussion, B: PD and PEP mask, C: PEP in sitting position and D: FET in sitting position. Sputum production was greatest in B and C, and skin oxygen tension was increased following treatment C. Falk concluded that PEP should be incorporated into physiotherapy regimens. In a similar study, Tyrell *et al*[19] showed that the PEP mask alone did not clear the chest adequately and PEP should be used in conjunction with FET. The effect of PEP breathing in CF was conducted by van der Schans *et al*[20]. They studied mucus transport in the lung using a radioactive aerosol tracer technique. Three methods were evaluated, namely: coughing, PEP with pressure of 5cm and 15cm H_2O. The results showed that PEP temporarily increases lung volume, but it did not improve mucus transport. The PEP mask is a useful adjunct method of chest physiotherapy which should be used in conjunction with other techniques[21].

4. Flutter VRP1



Legend: 1. Mouthpiece. 2. Circular cone. 3. High density stainless steel ball. 4. Perforated protective cover

The flutter is a small, simple handheld device which generates a controlled oscillating positive pressure whose frequency can be modulated[22]. The oscillating pulsed during expiration increase airway patency and expiratory flow. The vibratory effect enhances mucus clearance.

As the patient exhales, the ball is displaced and then rolls back into place. The fluctuations in pressure result in oscillations of positive expiratory pressure and airflow[22]. The bronchi are dilated up to the peripheral bronchioles. This positive expiratory pressure vibrates the airway walls to loosen mucus, decreases the collapsibility of the airways and accelerates airflow. The frequency of oscillations can be regulated by changing the inclination of the device slightly up or down from the horizontal position.

Use of the flutter

In a sitting position, the patient must relax and take a few normal breaths. The patient then takes a deep breath, inserts the device into the mouth and closes the lips firmly around it. Keeping the cheeks flat, the patient breathes out normally and deeply through the flutter. This cycle is repeated 10-15 times and mobilised secretions are then expectorated.

The only contraindication for use of the flutter is a pneumothorax. However, hyperventilation could result in dizziness and should this occur, frequent short interruptions every 5–10 cycles must be encouraged[22].

A few studies have investigated the efficacy of the Flutter. Konstan *et al*[23] evaluated the amount of sputum expectorated comparing the techniques of the Flutter, vigorous voluntary coughing and PD with percussion. The results showed that subjects using the flutter expectorated three times the amount of sputum compared to the other techniques. No lung function tests were recorded but a later study by Casaulta[24] evaluated lung function. It compared PEP-mask versus the flutter. After use of the flutter; VC, FEV_1, and MEF_{50} increased significantly compared to the PEP mask.

The advantage of the flutter is that

it is a relatively inexpensive device - which is also small, easily transportable, and allows for independent physiotherapy.

Autogenic Drainage

Autogenic drainage (AD) was devised in 1967 by Jean Chevaillier in Belgium[24]. Chevaillier observed that mucus clearance was enhanced during forced expiratory manoeuvres, playing, breathing activities and laughing. Chevaillier described AD by the principle of reaching the greatest possible airflow in different regions of bronchi by controlled breathing, put into practice by three phase breathing exercise[26].

At the beginning of AD, inspiration is performed slowly through the nose to provide optimal humidification. Expiration is done through the open glottis and open mouth, without pursed lips.

- Phase 1 : peripheral loosening of mucus

This pase begins with a directed increased inspiration followed by deep expiration. Simultaneously, the mid tidal volume is lowered in the range of normal expiratory reserve volume (ERV). Compression of the peripheral alveolar ducts mobilises secretions from the peripheral lung regions[26].

- Phase 2 : collection of mucus in large airways

This second phase is achieved by deepening inspiration and expiration. Mid tidal volume is changed gradually from ERV into the inspiratory reserve volume (IRV). The velocity of flow must be controlled to avoid high flow peaks which result in spasm of the collapsible segments at the equal pressure point[26].

- Phase 3 : transport of mucus from the large airways to the mouth

In the third phase, the patient increases respiratory flow starting from a level at about the middle of the inspiratory reserve capacity. Mucus is expectorated by a small burst of coughing or huffing.

The German physiotherapists have simplified the Belgian method of AD, as they observed that respiration in the ERV range seldom occurred because patients were uncomfortable breathing at low mid tidal volume[25]. The patient begins by varying the mid tidal volume and adapts the process to the individual need without excessive force. After every inspiration, the patient holds his breath for 2-3 seconds, followed by a passive but fast expiration to normal expiratory level, followed by further active expiration to low ERV.

AD is a difficult technique for the physiotherapist to learn and equally so in teaching the patient, therefore AD should be learned under the supervision of a trained physiotherapist from one of the limited number of centres in Europe. The patient is initially guided by tactile and auditory assistance of the physiotherapist, and gradually takes over and uses his/her proprioceptive sensations for detecting moving secretions. AD requires considerable patient cooperation and modification where indicated.

Schoni[26] registered flow volume curves of CF patients during AD. This study showed that during forced expiration, compression occurs at low lung volumes, whereas, during AD, higher flow occurs momentarily with the same low lung volumes without bronchial collapse. The techniques of AD and ACBT were compared in a study by Miller *et al*[27]. A significant difference was that AD improved FEF more often than ACBT and mucus clearance was greater on the AD treatment days. They concluded that AD is as effective as ACBT.

Exercise

Physical activity plays an important role in daily life. Different forms of physical activity such as exercise, games and sport promote well being - physically, mentally and socially. Exercise therapy can range from 10 minutes to vigorous sessions of hour-long activities including swimming, jogging, gymnastics, cycling and skipping[28]. The mini-trampoline has been included in physiotherapy treatment of CF patients to avoid monotony in training[29]. Research has not defined which physical activities are optimal for mobilising secretions. However, the choice of exercise should be determined by the patient in relation to the severity of their lung disease, their environment and motivation[30]. Physical activities selected for enjoyment generate increased motivation and enthusiasm, and encourage compliance. Patients with mild or moderate lung disease may not find their exercise capacity severely limited, in comparison to those with severe lung disease who may experience difficulty with physical exercise and require regular supervision[31]. Some patients will participate in sporting activities while others will be satisfied to climb stairs with minimal breathlessness. Many first referral patients are unaware of the importance of exercise[32]. The physiotherapist should introduce exercise to all CF patients, especially at an early age. It is important to encourage children to participate in physical activities at home and at school. The patient and the physiotherapist should be aware of dangers such as the possibility of infective exacerbations, exercise induced asthma, injury, exercise induced arterial oxygen desaturation and the risk of salt depletion after strenuous exercise in hot weather[33].

A recent study of Lannefors and Wollmer[5] showed that mucociliary clearance of radiolabelled particles by physical exercise, PEP and PD and percussion was not statistically different. Exercise, was however, associated with the lowest clearance. Cerny[33] conducted a study on bronchial drainage and exercise for in-hospital treatment of CF patients. The study showed significant improved spirometry. Orenstein *et al*[34] showed no improvement in FEV^1 after exercise, but FEV^1 in their control chest physiotherapy groups had sig-

nificantly deteriorated. Salh *et al*[38] studied the role of exercise and conventional physiotherapy in aiding sputum expectoration. The results indicated no improved amounts of sputum expectoration with exercise, and concluded that exercise should not be considered as a replacement, but rather an adjunct to physiotherapy.

CONCLUSION

There are numerous physiotherapy modalities available for CF patients. In addition to conventional PD several new techniques have been developed during the past 10-15 years. The physiotherapy modality must be effective, less time-consuming, appealing to the patient and allow for greater independence. Physiotherapists should be flexible in their approach to the treatment of CF patients. Parts of one modality can be applied and integrated in other methods and a combination can be beneficial. The question of which modality to use or which one is more effective often arises. A modality which suits the individual needs of the patient and at which he/she is competent is optimal. Patients should be involved in this decision making which will enhance compliance with in-hospital and home physiotherapy. Physiotherapy should interfere minimally with daily living.

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- Following publication of the paper in the proceedings of the Congress attended, the paper will be published in the SA Society of Physiotherapy Journal. (Candidates must therefore ensure that permission is obtained for the paper to be published in the SASP Journal, from the Congress Secretariat).
- Applications from any physiotherapist who will be presenting a paper at a congress, must be submitted to the Head Office of the SASP, together with a short CV and motivation by 31 March every year (the notice for 1995 appeared in the Journal in August 1994).

The South African Society of Physiotherapy is very grateful for this generous gesture by Smith and Nephew, which will be of particular value in these difficult economic times.