PAEDIATRIC RESPIRATORY REVIEWS (2007) **8**, 8–16

ELSEVIER

Paediatric Respiratory Reviews

MINI-SYMPOSIUM: AIRWAY CLEARANCE IN CYSTIC FIBROSIS

Chest physical therapy, breathing techniques and exercise in children with CF

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KEYWORDS
cystic fibrosis;
chest physiotherapy;
postural drainage and percussion;
active cycle of breathing;
forced expiration;
autogenic drainage;
exercise;
airway clearance techniques

Summary Chest physiotherapy in the form of airway clearance techniques and exercise has played an important role in the treatment of cystic fibrosis. Until the 1990s the primary airway clearance technique used was postural drainage combined with percussion and vibration (PD&P). It was introduced into the treatment of CF with little evidence to support its efficacy and once established, it has been difficult ethically to perform a study comparing PD&P to no treatment. A common question, yet unanswered is when should it be commenced, especially for the newly diagnosed asymptomatic CF patient? Recently, the technique of PD&P has been modified to include only non-dependant head-down positioning due to the detrimental effects of placing a person in a Trendelenburg position. In the 1990s other airway clearance techniques gained popularity, in that they could be performed independently, in a sitting position and avoided many of the detrimental effects of PD&P. These techniques include the Active cycle of breathing technique, formally called the Forced expiration technique and Autogenic drainage. Both these breathing techniques aim at using expiratory airflow to mobilize secretions up the airways and incorporate breathing strategies to assist in the homogeneity of ventilation. Studies suggest that both these techniques are as effective if not more effective than as PD&P and offer many advantages over PD&P. It has been suggested that exercise can be used as an airway clearance technique; however the literature does not support this. Rather, when exercise is used in addition to an airway clearance technique there is enhanced secretion removal and an overall benefit to the patient. Further research needs to be directed at assessing the effects of an airway clearance technique on the individual patient using appropriate outcome measures.
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Chest physiotherapy (CPT) is an ambiguous term which refers to a variety of physiotherapy modalities used in the treatment of patients with underlying cardio-respiratory pathology. This includes airway clearance techniques (ACT), exercise, thoracic mobility exercises, positioning, breathing exercises, and inhalation therapy. Mistakenly, the term, 'CPT' has been used synonymously with the terms postural drainage and percussion with the latter being the traditional method of airway clearance used to treat cystic fibrosis patients until the early 1990s. In order to clarify terminology, the International Physiotherapy Group for Cystic Fibrosis (IPG/CF) has defined chest physiotherapy in the former term.

CPT should now only refer to, 'Cardio-pulmonary Physical Therapy' encompassing a more comprehensive approach to the cardiopulmonary system. Individual techniques are classified under subheadings such as airway

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doi:10.1016/j.prrv.2007.02.013

clearance techniques. The airway clearance techniques currently available to treat patients with cystic fibrosis (CF) include postural drainage and percussion (PD&P), Active Cycle of Breathing Technique (ACBT), Autogenic Drainage (AD), Positive Expiratory Pressure (PEP), High Pressure PEP, Oscillating PEP, High Frequency Chest Wall Oscillation (HFCWO) and Exercise.[1] This review will focus on the techniques of PD&P, ACBT, AD and the use of exercise in the treatment of cystic fibrosis.

The goals of airway clearance techniques in CF are to assist in the removal of airway secretions, thus improving ventilation, reducing airway resistance, correcting ventilation-perfusion mismatch and decreasing the proteolytic activity in the airway. As the mean life expectancy of CF patients increases due to better treatment, the goals of ACTs must also include long-term outcomes such as retarding lung disease and preserving physical function in order to improve quality of life.

COMMENCEMENT OF PHYSIOTHERAPY

Prior to newborn screening, the diagnosis of CF was made as a result of symptoms, either nutritional and/or respiratory. Physiotherapy was initiated immediately as it was considered the mainstay of therapy. However, over the past decade, the number of infants diagnosed with CF through newborn screening has increased worldwide. Often these infants are diagnosed before any recognizable respiratory symptoms appear. Now some CF centres are questioning when to begin physiotherapy in these infants, especially in ones who are apparently asymptomatic.[2]

The question arises as to whether these infants are truly asymptomatic or whether the present clinical tools used to assess the degree of respiratory involvement are inadequate to detect underlying lung pathology. Broncho-alveolar lavage has shown that airway inflammation and infection are present in the CF infant as early as 4 weeks of age.[3–5] Ranganathan[6] found that infants, who were judged asymptomatic on clinical examination, had diminished full and partial forced expiratory maneuvers. Tepper *et al.*[7] states, 'In the absence of respiratory symptoms, the clinical exam and radiological evidence may understate the degree of lung disease'. He concluded that it is not possible to assess a patient as being asymptomatic on the basis of clinical examination alone. One argument for commencing physiotherapy at diagnosis comes from Connett *et al.*[8] They compared the outcomes at 1, 3, 6, and 10 years of 73 CF patients with a delayed diagnosis of CF. The control was a group of CF patients diagnosed within 4 months of age who were commenced on treatment at diagnosis. Treatment consisted of physiotherapy, digestive enzymes, antibiotics and counseling. At 10 years the delayed diagnosis group required increased treatment to maintain their health and had significantly more radiological changes.

Although there is a question of when mucociliary transport becomes impaired in CF patients, we know that CF lung disease begins early in life. For this reason, many members of the multidisciplinary care team believe that physiotherapy should be commenced at diagnosis with the aim of preventing lung disease. This also lays a foundation for infants to accept physiotherapy as part of their daily life. Their parents learn to differentiate between the normal baseline for their infants and any early changes and when they need to communicate with the CF team.

In advocating for physiotherapy to commence at diagnosis, one must carefully balance the burden that is placed on the family to perform this treatment against the gain from the treatment as it must be acknowledged that there is no direct evidence showing the benefit in lung function of commencing ACT at diagnosis. In North America, the ACT most often initiated with newly diagnosed CF infants is a modified form of postural drainage and percussion.

POSTURAL DRAINAGE AND PERCUSSION (PD&P)

In the 1950s, postural drainage and percussion was introduced as a standard part of the CF care,[9] despite any large randomized controlled trials to assess its effectiveness. It includes placing a patient in a gravity dependent position and percussing the chest wall over the area being drained, for typically between 3–5 minutes. The patient is then asked to inhale deeply 3–4 times and on exhalation the chest wall is vibrated; this is followed by directed coughing.[10] There are 12 different postural drainage positions used, based on the nomenclature of the bronchopulmonary anatomy.[11] The physiological rationale for the use of postural drainage to assist in the clearance of secretions is based on the use of gravity to assist with the mucociliary action. In healthy individuals ciliary action moves secretions up the airways at a rate of 3–5 mm/min.[12,13] In people with CF where the mucociliary action is impeded, Wannemaker *et al.* found airway secretions moved slowly in the opposite direction towards the periphery of the lung, but when placed in a head-down gravity dependent position, secretions moved up the airways at the normal rate of 3–5 mm per minute.[13] This study suggests that gravity can assist in the removal of secretions from the periphery of the lungs. However, taking this study one step further, for PD to be effective in moving secretions for the basal segments of the lungs to the bifurcation of the right and left main stem bronchus, (approximately a distance of 30 cm in an adult) a patient would have to be placed in a head down position for between 60–100 minutes. MacKenzie *et al.*[14] in a study on 42 ventilated patients, demonstrated an increase in total lung compliance following a session of postural drainage, percussion and vibration

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