Physiological principles of airway clearance techniques used in the physiotherapy management of cystic fibrosis

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Summary Cystic fibrosis (CF) has a reported incidence of one in 2500 live births in the UK and is the most common cause of suppurative lung disease in Caucasian children. Effective management of the respiratory component of this condition relies on optimal antibiotic therapy combined with efficient clearance of retained airway secretions. Physiotherapy management of CF aims to address patient problems associated with the impairment of mucociliary clearance mechanisms. Over the last two decades a number of airway clearance techniques (ACTs) used during physiotherapy treatment have been developed and studied within the CF paediatric population. Many of these ACTs share common physiological principles that are well described and reported. This article gives a brief overview of the most commonly used techniques used in the paediatric CF population.

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KEYWORDS
Cystic fibrosis; Physiotherapy management; Retained secretions; Airway clearance techniques (acts)

Practice points

- **Physiotherapy assessment**: All paediatric patients diagnosed with CF should be assessed by a physiotherapist experienced in the management of this condition
- **Individual patient management plans**: All paediatric CF patients should, following assessment, be provided with individual physiotherapy management plans
- **Airway clearance techniques (ACTs)**: Physiotherapy ACT enhances the clearance of excess bronchial secretions in the paediatric patient with CF resulting in decreased airway obstruction, decreased airway resistance and improved ventilation
- **Treatment evaluation and outcome measures**: Appropriate measurement tools should be used to evaluate physiotherapy treatment interventions

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Research directions

- **Infant treatment**: The development of neonatal screening programmes has resulted in reduced age at diagnosis of CF. Further research is required to determine the effect this will have on early physiotherapy treatment interventions.
- **Combining treatment modalities**: The burden of daily routines of airway clearance techniques (ACTs) and inhalation therapy is significant. Further research could determine how combining these treatment modalities may benefit the paediatric patient with CF.
- **Novel ACTs**: Whilst it is important that paediatric CF patients are offered the most up to date, effective and socially acceptable ACTs available, the introduction of a novel ACT should be properly evaluated.

Introduction

The clinical manifestation of cystic fibrosis (CF) is characterised by chronic pulmonary infection and pancreatic insufficiency, with associated problems such as cystic fibrosis related diabetes (CFRD) and liver disease generally developing as later complications. The main cause of morbidity and mortality remains pulmonary disease and much of the clinical management is aimed at the treatment of a patient’s respiratory problems.¹

Along with antibiotic therapy, physiotherapy management is an important component in the treatment of the respiratory problems seen and has been shown to be of benefit in the removal of excess airway secretions² and in the treatment of sensations of breathlessness (SOB) and decreased exercise activity and tolerance.³

Physiotherapy treatment should be introduced at diagnosis by a clinician experienced in the management of this condition.⁴ In collaboration with the child and their family the physiotherapist should develop individual management plans based on patient assessment. Depending on the patient problems identified, a number of physiotherapy treatment modalities may be introduced.

These include:

- Airway clearance techniques (ACTs), to aid in the removal of retained secretions.
- Adjuncts to ACTs such as aerosol therapy (inhalers and nebulisers).
- Education/advice on exercise activity and exercise programmes.
- Advice on posture and treatment for musculoskeletal pain.
- Oxygen therapy and non-invasive ventilation.
- Advice and treatment of stress incontinence symptoms.

Airway mucus in CF

The presence of excessive airway secretions in CF due to chronic infection and mucosal inflammation and the retention of infected secretions due to bronchoconstriction and impaired mucociliary clearance leads to the pattern of airway obstruction, mucus plugging, atelectasis, airway instability and hyperinflation seen.⁵

Removal of retained secretions using physiotherapy ACTs will result in decreased airway obstruction and resistance, improved ventilation and air flow, decreased shunting and reduced ventilation/perfusion mismatch.⁶ There may also be some benefit in the removal of the bacterial products produced during chronic infection, which have been shown to further disrupt local defence mechanisms.⁷

Airway clearance techniques

In combination with an effective cough, physiotherapy ACTs used in the treatment of paediatric CF can consist of a number of different modalities⁸ including:

- Use of gravity to aid mucus transport—postural drainage (PD).
- External application of forces against the chest wall—percussion, vibrations, shaking, high-frequency chest wall compression (HFCC).
- Breathing techniques—active cycle of breathing techniques (ACBT) and autogenic drainage (AD).
- Devices designed to introduce positive pressure and/or oscillation into the airways—positive expiratory pressure (PEP) masks, flutter, cornet and accapela, intrapulmonary percussive ventilation (IPV).
- Physical activity and prescribed systemic exercise programmes.

It is important to note that whilst ACTs used within the paediatric and adult CF populations share common principles, there are important physiological differences between the two groups and rationales for use may differ substantially.
These differences are most apparent in the neonate, but are present throughout infancy and disappear only as growth and development evolves through childhood into adulthood.9

Gravity assisted positions (postural drainage)

One of the earliest and frequently used ACTs in CF, postural drainage, requires the patient to be placed in positions that anatomically favour the gravity directed movement of secretions towards the upper airways, from where secretions can be cleared by coughing. The rationale for this technique assumes that additional gravitational forces on vertically positioned bronchi will enhance impaired mucociliary transport. However in CF, consideration must be given to the fact that airway secretions are hyper viscid and effective drainage times can result in prolonged treatment sessions that may be non-acceptable to children and families. Recent studies have also shown that the prevalence of gastro-oesophageal reflux (GOR) in infants and young children with CF may vary from between 35% to 81%10 and that standard PD positions may not be recommended in these patients.11

Manual techniques (percussion, vibrations and shaking)

Manual techniques such as percussion, vibrations and shaking are generally performed whilst the patient is placed in postural drainage positions to further enhance mucociliary clearance. Percussion consists of a rhythmical clapping on the patient’s chest wall with a cupped hand, whilst the patient is asked to take deep breaths. Vibrations consist of a fine oscillation of the therapist’s hands placed either side of the patient’s chest wall and directed inwards, while shaking is a similar but coarser movement in which the hands rhythmically compress the patient’s chest wall again in an inwards direction. The latter two techniques are performed during exhalation following a maximum inspiration.12

The exact physiological mechanism by which manual techniques aid the removal of secretions is not proven. The hypothesis is that the application of an external force to the chest wall creates intrapleural pressure changes that are transmitted through the thoracic cage and lung tissue, loosening any secretions attached to the airway wall. These secretions then collect in the lumen of the airway from where other mucociliary mechanisms can aid in removal. If correct, then these techniques should be more effective over solid, i.e. consolidated or atelectatic, lung segments than healthy lungs where aerated lung tissue would absorb any pressure changes. In clinical practice such techniques are usually applied directly over areas of lobar or segmental lung disease. In widespread disease pathologies such as CF, they are used with the understanding that forces generated during therapy may be attenuated, thus reducing efficacy.

Care must also be taken when using manual techniques with newborn and very immature infants to ensure that excessive movement of the unsupported head does not occur. Whilst some concerns have been reported in neonatal management13 there have been no reports of adverse effects in CF patients.

Breathing techniques

Active cycle of breathing techniques (ACBT)

The ACBT is the most commonly used ACT in the UK.14 ACBT is a cycle of breathing techniques comprising:

- Breathing control (BC)—relaxed tidal volume breathing using minimal effort by the patient. This aims to decrease airway obstruction and improve airflow by reducing bronchoconstriction and by using collateral ventilation to open up airways and get air behind secretions.5
- Thoracic expansion exercises (TEE)—where the patient is encouraged to inspire to full lung volume. This technique also utilises collateral ventilation and the interdependence mechanism15 to move air into smaller airways obstructed by secretions. During inspiration, expanding alveoli exert a force on adjacent alveoli that may assist in the re-expansion of collapsed ventilatory units; these effects are felt to be more marked during deep breathing.
- The forced expiration technique (FET) or ‘huffing technique’—where the patient performs a combination of one or two forced expirations combined with periods of breathing control.5 The rationale behind the FET is based on the physiological concept of the equal pressure point (EPP), which is described ‘as the point at which pressure within the bronchi (Pbr) equals peribronchial (Ppl) pressure during a forced expiration’.15 During expiration the EPP starts at the mouth and moves peripherally towards the lobar and segmental bronchi but does not extend to the terminal units. Downstream of the EPP, dynamic compression and collapse of the airways

The principles of physiotherapy in CF
occurs because $P_{br}$ becomes less than $P_{pl}$, expiratory flow is compromised, airflow becomes increased and more turbulent thus mobilising secretions.\textsuperscript{5}

In clinical practice the FET can be started at different lung volumes thereby clearing different sized airways. As clearance of secretions using the FET is dependant on patient lung volume, 'huffing' to low lung volume will move more peripherally situated secretions and a 'huff' at high lung volume will clear secretions from more proximal airways. However, using the EPP to mobilise secretions can result in an increase in airway obstruction, particularly in diseased or immature airways which may be anatomically unstable and/or hyper reactive.\textsuperscript{9}

**Autogenic drainage (AD)**

AD is a three phase breathing technique developed in Belgium.\textsuperscript{16} It aims to increase airflow through airways whilst avoiding the early airway closure that occurs in diseased or damaged airways as previously described. AD aims to control expiratory flow at differing lung volumes whilst avoiding the generation of an EPP during expiration, allowing flow rates to last longer, therefore move secretions further during each expiration.\textsuperscript{16} During therapy, tidal volume breathing is maintained at low lung volume (un-stick phase), mid lung volume (collect phase) and high lung volume (evacuate phase) depending on whether the secretions are located within the peripheral, proximal or central airways, respectively. AD has shown to be of particular benefit to patients with significant airway hyper-reactivity\textsuperscript{17} but in clinical practice patients with advanced disease might find sustained breathing at low lung volumes uncomfortable.

**Positive expiratory pressure (PEP)**

A number of PEP devices have been developed for use with paediatric CF patients. These include low-pressure PEP therapy,\textsuperscript{18} high-pressure PEP therapy\textsuperscript{19} and oscillating PEP therapy.\textsuperscript{20} PEP therapy is based on the physiological principles of collateral ventilation,\textsuperscript{15} which suggests that ventilation and airflow can occur between adjacent lung segments through anatomical channels such as:

- Interbronchiolar channels of Martin
- Bronchiole—alveolar canals of Lambert
- Alveolar pores of Kohn

In children, any reliance on collateral ventilation channels to enhance mucociliary clearance during physiotherapy treatment should recognise that they are poorly developed during infancy but may be present from 2 to 3 years of age.

In healthy adults, resistance to airflow through collateral channels is high in relation to airflow resistance within the conducting airways and therefore does not readily occur. However in disease, when airflow resistance is increased within the conducting airways by the presence of retained secretions or airway collapse, resistance within collateral channels is reduced in relative terms and airflow through collateral channels is more likely to occur.

In clinical practice, PEP therapy allows more air to enter the airways through collateral channels during inspiration, than escapes during expiration. This results in the development of a pressure gradient that builds up behind sputum plugs, moving secretions into more central airways from where it can be more easily removed.\textsuperscript{18} Positive pressure within the airways is also thought to prevent compression-induced collapse, splinting open the airways during expiration and thus further assisting with the clearance of secretions from distal airways.\textsuperscript{19}

A commonly used PEP therapy system consists of a close fitting mask and a one-way valve to which expiratory resistors can be attached. A manometer placed in parallel with the resistor can be used to determine the correct pressure used during therapy. In low-pressure PEP therapy the diameter of the resistor used for treatment is determined, using tidal volume breathing, to give a steady PEP of 10–20 cm H$_2$O during the middle part of expiration. High-pressure PEP therapy uses the same equipment but sustained expiratory pressures achieved usually range between 40 and 100 cm of H$_2$O.

The physiological effects of high pressure PEP in CF can be explained by increased collateral airflow to underventilated lung regions, effective splinting of the damaged airways during a forced expiration manoeuvre and by allowing the patient to expire to a volume greater than his/her usual forced vital capacity (FVC).\textsuperscript{5} This technique can be useful for the paediatric patient with more unstable airway disease but as it requires significant patient effort may not be suitable for those who tire easily.

Devices are now available that offer a combination of variable positive airways pressure and oscillation of air within the airways during expiration. These include the Flutter\textsuperscript{46}, RC-Cornet\textsuperscript{47} and Acapella\textsuperscript{48}. Exhalation through these devices generates both oscillations of positive pressure in the airways and repeated accelerations of expiratory
Sputum clearance. Physiologically this effect can be explained by:

- Oscillating positive pressure causing the diameter of the airways to increase and thus prevent early airway collapse.
- Pressure induced vibrations within the airway walls displacing secretions into the airway lumen.
- Repeated accelerations of expiratory airflow favouring movement of secretions from the distal to the central airways from where expiration can take place.

These devices can be used independently by patients for regular therapy, however, appropriate education and re-education by a physiotherapist is required for effective therapy and attention must be paid to correct cleaning procedures in order to reduce any risk of infection.

Physical activity/exercise

A number of paediatric CF centres worldwide commonly use ACTs based on physical activity. Physical activity is considered socially acceptable and helps normalise the patient’s life rather than accentuate the medical aspects. In children with CF, systemic exercise has been shown to improve ventilation, aid in the mobilisation of secretions and counteract microatelectasis. The development of a healthy lifestyle and improved general fitness in children with CF are to be recommended, with the recognition however that oxygen consumption during maximal exercise appears to be less than that seen in adults and in children the cardiovascular training effect seen may be smaller.

Conclusion

Whilst the physiological principles that underpin the physiotherapy ACTs used in the treatment of paediatric CF are well recognised, recent studies have changed how they are applied in clinical practice. Modified postural drainage positions are now considered to be more suitable for patients at risk of GOR and the routine use of traditional ACTs in the management of the asymptomatic patient is questioned. Paediatric patients are increasingly being educated in the use of independent ACTs and, with few exceptions, physical activity is encouraged for CF patients of all ages.

As novel ACTs are introduced to our paediatric CF populations it is important to ensure that their physiological basis is established and their efficacy examined by prospective research.

References
