Airway Management and Ventilation

The Role of Airway Clearance Therapy in Reducing Pulmonary Complications in Cystic Fibrosis Patients

A report by

Giovanna Pisi and Alfredo Chetta

1. Senior Registrar, Cystic Fibrosis Unit, Paediatric Department; 2. Professor of Respiratory Medicine, Respiratory Disease Section, Cardiopulmonary Department, University of Parma

A dramatic improvement in cystic fibrosis (CF) survival rates has been witnessed over the past 50 years, and the median survival of children born in the 1990s is estimated to exceed 40 years. Nowadays, more than 90% of the mortality and most of the morbidity from CF is related to the consequences of chronic pulmonary infection, especially by Pseudomonas aeruginosa, and suppurative lung disease. Respiratory exacerbations of CF lung disease have been associated with increased mortality and hospitalisation and reduced quality of life. Consequently, strategies to reduce the frequency and severity of respiratory exacerbations may ensure continuous advances in clinical outcomes. Impaired clearance of abnormally viscous airway secretions is a major feature of lung disease in CF, therefore attempts to improve airway clearance through physiotherapy remain one of the cornerstones of treatment for patients with CF. Airway clearance techniques (ACTs) have been shown to improve mucociliary clearance. Furthermore, ACTs can decrease mucus plugging and aid the removal of secretions containing inflammatory cells and bacteria, thus improving ventilation, reducing airway obstruction and atelectasis, correcting ventilation-perfusion mismatch and decreasing proteolytic activity in the airways.

Airway Clearance Therapy

ACTs currently available to treat CF patients include postural drainage associated with chest percussion and vibration (PD&P) or conventional physiotherapy (CPT), active cycle of breathing techniques (ACBTs), autogenic drainage (AD), positive expiratory pressure (PEP) and its variants and high-frequency chest wall oscillation (HFCWO). PD&P was introduced in the 1950s as a standard part of CF care; however, until now little evidence has been provided to support its use in clinical practice. Reisman and colleagues conducted a three-year prospective study that compared the effects of PD&P on pulmonary function combined with the forced expiratory technique (FET) versus the effect of the FET alone. The authors found that combined therapy was able to reduce the annual rate of decline in respiratory function. In 1995, a meta-analysis of studies comparing PD&P with no physiotherapy demonstrated a significant benefit in favour of PD&P. In contrast, a subsequent study showed that PD&P had detrimental effects on patients by inducing hypoxic episodes. Moreover, patients find PD&P so burdensome that compliance with the prescribed treatment regimen is probably less than 50%. Currently, PD&P (modified to exclude the head-down position) should be used only in young children (less than five years of age) and in patients with extremely severe lung disease who are unable to perform ACTs independently.

In the 1990s, several self-administered ACTs were developed, including the ACBTs and AD. Both of these breathing techniques are based on increasing expiratory airflow rates to move secretions up the airways. In addition, these breathing strategies aim to improve the regularity of ventilation. ACBTs, developed by Pryor and Webber at the Royal Brompton Hospital, London and originally called FET, combine forced expiration (huffing), relaxed tidal volume breathing (or breathing control) and thoracic expansion exercises. Developed in Belgium by Chevalier, AD utilises breathing at different lung volumes to influence the movement of mucus from different parts of the bronchial tree; at the end of the exercise a huff or cough removes the secretions from the upper airways. Some studies suggest that both ACBTs and AD are more effective than PD&P and may offer many advantages over PD&P. In fact, neither ACBTs nor AD causes oxygen desaturation or requires a caregiver, thereby promoting more independence than PD&P.

Several self-administered devices have been used in clinical settings to aid airway clearance in CF patients, including PEP masks and HFCWO. The PEP mask, developed in Denmark in the late 1970s as an alternative to PD&P, is the simplest and least expensive of the airway clearance devices. Using a face mask or mouth-piece, the patient exhales through an expiratory resistor, which generates positive pressure in the airways, which stabilises the peripheral airways, while air is pushed through collateral ventilation pathways into distal lung units beyond retained secretions. A recent Cochrane review of PEP masks concluded that there was no clear evidence that this device was better than other forms of physiotherapy, although patients tended to prefer PEP masks over PD&P.
Recent reports in large groups of CF patients showed that one- and three-year treatment programmes with HFCWO were equivalent in terms of an increase in respiratory function and sputum weight. In contrast, in a recent cross-over trial Klutt et al. demonstrated that significantly more sputum was expectorated during HFCWO than during CPT, as determined by both the wet and dry measurements. In contrast, in a recent study Phillips et al. found that HFCWO was less effective than ACBT in terms of an increase in respiratory function and sputum weight. Interestingly, a long-term non-randomised study showed that the rate of decline in pulmonary function was significantly decreased during the HFCWO treatment period compared with CPT.

To date, several studies have been published on the acute and long-term effects of HFCWO in CF patients. As a whole, these studies suggest that HFCWO is at least as effective as CPT in clearing secretions from the airways of patients with CF. Two short-term randomised controlled trials comparing HFCWO, CPT and PEP mask in CF patients hospitalised for a pulmonary exacerbation showed no differences in pulmonary function and sputum production outcomes. Moreover, in a random cross-over trial Klutt et al. demonstrated that significantly more sputum was expectorated during HFCWO than during CPT, as determined by both the wet and dry measurements. In contrast, in a recent study Phillips et al. found that HFCWO was less effective than ACBT in terms of an increase in respiratory function and sputum weight. Interestingly, a long-term non-randomised study showed that the rate of decline in pulmonary function was significantly decreased during the HFCWO treatment period compared with CPT.

Recent reports in large groups of CF patients showed that one- and three-year treatment programmes with HFCWO were equivalent in terms of clinical outcomes compared to CPT. Taken together, these studies indicate that HFCWO therapy can be a reliable alternative to CPT. Moreover, this treatment is usually well tolerated, although some patients—especially those with end-stage lung disease—may complain of discomfort or pain from the inflated vest. With this in mind, a study was specifically addressed to compare patient acceptance of HFCWO, administered via the Vest System, with either CPT or flutter valve (which is a combination of the PEP mask and airway oscillation). In this study, patient satisfaction and compliance were significantly higher with the Vest System compared with either CPT or flutter. In fact, the major problem regarding use of the Vest System remains the high cost of the device ($US15,000); however, a retrospective record-based analysis of the impact of HFCWO on total healthcare expenditure showed a decrease in total direct cost for 23 CF patients after HFCWO treatment.

Among the self-administered techniques, ACBTs, PEP and AD require continuous active participation by the patient, while HFCWO using the Vest System allows the patient to be passive.

Conclusions

Treatments to enhance the clearance of airway secretions are crucial to the management of CF lung disease. ACTs can facilitate the expectoration of tenacious secretions that would otherwise accumulate in the airways. There is evidence from short-term, but not long-term, trials about the benefit of ACT over no treatment. However, there is no consensus about which ACT is the most effective.

Traditionally, chest physiotherapy relied on postural drainage combined with percussion and forced expirations, although there is evidence that PD&P is at least as effective as other forms of ACT. However, this kind of chest physiotherapy is time-consuming and sometimes uncomfortable for patients, who tend to prefer self-administered treatments. Among the self-administered techniques, ACBTs, PEP and AD require continuous active participation by the patient, while HFCWO using the Vest System allows the patient to be passive. Therefore, the Vest System may be useful both in fatigued patients and in patients without a care-giver.

In conclusion, CF is a multisystemic disease with a high degree of variability in lung impairment and it is likely that specific airway clearance regimens may be required. Therefore, ACTs need to be individually and continuously adapted to suit the needs of patients, families and care-givers. Randomised clinical studies are needed to examine the long-term effects of ACTs on exercise tolerance, quality of life and survival of CF patients.