

Physical therapy

Positive expiratory pressure in cystic fibrosis

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Background

Positive expiratory pressure (PEP) devices provide constant back pressure to the airways during expiration. This may improve clearance by building up gas behind mucus via collateral ventilation. There is no consensus regarding optimal treatment frequency and number of cycles included in each treatment session, so their use must be individualized ([Mcllwaine M et al. 2017](#)). Given the widespread use of PEP devices as self-administered therapy compared to conventional physiotherapy, there is a need to determine the evidence for their effect.

Issues

To determine

1. effectiveness of PEP device compared to:

- postural drainage with percussion;
- oscillating PEP (Flutter);
- forced expiratory technique (FET);
- non-invasive bi-level ventilatory support (nBVS);
- different pressures at 5cmH₂O or >20cmH₂O

as a mean of improving mucus clearance and other outcomes including

1. side effects;
2. acceptability.

What is known

A CDSR ([Mcllwaine M. 2019](#)) evaluated the PEP technique for airways clearance in CF patients. A total of 28 studies (involving 788 children and adults) were included in the review; 18 studies involving 296 participants were cross-over in design. The evidence provided by this review is of variable quality, but suggests that all techniques and described devices may have a role in the clinical treatment of people with CF. Following meta-analyses of the effects of PEP versus other airway clearance techniques on lung function and patient preference, this Cochrane Review demonstrated that there was a high-quality evidence that showed a significant reduction in pulmonary exacerbations when PEP using a mask was compared with HFCWO. Airway clearance techniques should be individualised throughout life according to developmental stages, patient preferences, pulmonary symptoms and lung function. This also applies as conditions vary between baseline function and pulmonary exacerbations.

1 CDSR ([Wilson LM. 2018](#)) included six reviews that compared any airway clearance technique, either as a single technique or as a combination of techniques, to no intervention, to coughing, or to another airway clearance technique. The quality of the body of evidence comparing different airway clearance techniques on different outcomes was either low or very low. The Authors concluded that patients with CF should choose the airway clearance technique that best meets their needs, after considering comfort, convenience, flexibility, practicality, cost, or some other factor. More long-term, high-quality randomised controlled trials comparing different airway clearance techniques among people with CF are needed.

In one CDSR ([Mcllwaine M et al. 2015](#)) a total of 26 studies (involving 733 participants) was included. Eighteen studies involving 296 participants were cross-over in design. These studies compared PEP to active cycle of breathing techniques (ACBT), autogenic drainage (AD), oral oscillating PEP devices, high frequency chest wall oscillation (HFCWO) and Bi level PEP devices (BiPaP) and exercise. Through the studies FEV1% predicted for age and sex, number of respiratory exacerbations per year, number of days of intravenous antibiotics per year, adherence and participants preference were evaluated as primary outcomes. As secondary outcomes radio labelled aerosol clearance after a single treatment of PEP, volume of expectorated sputum, total lung capacity, FEF25-75%, FVC%, blood oxygen levels, nutritional status, radiological imaging were evaluated both in short-term (up to seven days) or long-term (more than seven days) studies. Mainly gastro-oesophageal reflux was evaluated as a potential side effect. FEV1 was the primary and the most frequently reported outcome in the studies. No significant difference in effect between PEP and other methods of airway clearance were reported on this outcome, both when evaluating single interventions and series of treatments that continued for up to three months. Long-term studies had conflicting results regarding the effect on this outcome. A second primary outcome was the number of respiratory exacerbations. There was a lower exacerbation rate in participants using PEP compared to other techniques when used with a mask for at least one year. Participant preference was reported in ten studies; and in all studies with an intervention period of at least one month, this outcome was in favour of PEP. No sufficient details are available for the remaining outcome measures in different papers. The only reported adverse event was registered in a study where infants performing either PEP or postural drainage with percussion experienced some gastro-oesophageal reflux. This was more severe in the postural drainage with percussion group.

It is still debated whether PEP is more effective for secretion clearance during an infective exacerbation or for maintenance therapy. In general PEP is used in combination with various other interventions (e.g. pharmacological therapies, other physical therapy techniques, or modification to the technique known as high pressure PEP).

A retrospective study ([Orlik T et al. 2015](#)) was performed to evaluate the application of the PEP system for inhalations with hypertonic saline or saline and additionally rhDNase in 29 patients with CF, showing the greatest improvement in the values of MEF 75%50%25% after a 18 months observation.

A clinical trial ([NCT02303808](#)) has been completed in 2015 in order to analyze whether the inhalation of hypertonic saline combined with a positive expiratory pressure (PEP) device (Acapella Duet) increases the amount of sputum expectorated during the chest physiotherapy session (inhalation + bronchial drainage) in subjects with CF 16 years and older. Data have not been published.

Data from a prospective, 3-month randomized trial comparing standard PEP to NIV in 32 subjects 31 years of age with a mean FEV1 of 47% (± 14) and mean FVC of 69% (± 13) ([Rodriguez Hortal MC et al. 2016](#)) showed a significant reduction in lung clearance index (LCI) following NIV compared with PEP ($p = 0.01$). NIV could be a good alternative to PEP in chest physiotherapy for severely ill patients with CF.

An Australian randomized controlled trial for parallel group to evaluate the exercise alone versus exercise and positive expiratory pressure as a form of airway secretion clearance in adults with mild cystic fibrosis-related respiratory disease has been completed ([ACTRN12615001361594](#)). Data have been published ([Ward N et al. 2018](#)). The number of recruited adults was so little to achieve firm conclusions on the effect of PEP mask plus exercise vs the exercise-only group.

One RCT ([Dwyer TJ. 2019](#)) investigated the effects of treadmill exercise vs resting breathing and PEP therapy on mucus clearance in 14 adults with mild to severe CF lung disease. Mucus clearance was measured using the radioaerosol technique and gamma camera imaging. Treadmill exercise alone was less effective than PEP therapy (mean difference -7%, 95% CI -6- -8). There were no significant differences in mucus clearance from the intermediate and peripheral lung regions, but significantly less clearance from the central lung region.

One retrospective case-control study at a single CF care center ([Byrwa DJ. 2023](#)) enrolling 50 CF patients (age < 18 years) evaluated lung function and measures of healthcare utilization in a case group (n=14), who used high-frequency chest wall oscillation or positive expiratory pressure devices at school for at least 1 year after self-reported or physician identified inadequate use at home, and a matched control group by age and gender (n=36), composed of subjects with self-reported adequate use of ACT at home. In the case group paired t-tests showed that after initiation of ACT at school, there were significant reductions in PEx requiring IV or PO abx ($P = 0.010$), total days of abx ($P = 0.032$), and visits to the CF care center ($P = 0.037$). There was no change in these outcomes in the matched control group. This is the first known study to highlight an initiative between a CF care center and schools which utilized airway clearance devices at school to ensure pediatric CF patients completed ACT. Through increased adherence, this relationship was associated with improved health outcomes. Use of alternative strategies may help patients with CF sustain adequate airway clearance.

One randomized, sham-controlled crossover trial ([Gambazza S. 2024](#)) investigated short-term effects of positive expiratory pressure mask on ventilation inhomogeneity in children with CF. The study sample was 19 cwCF (ten girls), aged 11.4 (2.7) years. The adjusted $S(acin) \cdot VT$ mean difference between the standard and the sham procedure was -0.015 (90% confidence interval [CI]: -? to 0.025) $L(-1)$. There was no statistically significant difference in $S(cond) \cdot VT$ and lung clearance index between the two procedures: -0.005 (95% CI: -0.019 to 0.01) $L(-1)$ and 0.49 (95% CI: -0.05 to 1.03) turnovers, respectively. Findings do not support evidence for an immediate effect of PEP mask physiotherapy on $S(acin) \cdot VT$ with pressure range 10-15 cmH₂O. Measurement with the N(2)MBW and the crossover design were found to be time-consuming and unsuitable for a short-term study of airway clearance techniques.

A clinical trial ([ACTRN12613000929707](#)), including participants from 4 years age maximum 17 years, had to evaluate improving adherence with PEP chest physiotherapy in children with CF through the use of an electronic device, the PEP PT (personal trainer), a device that can measure the pressure associated with each blow using an electronic sensor contained within the mask. The study has been ceased.

Unresolved questions

There is no clear evidence that PEP was more or less effective intervention than other forms of physiotherapy as HFCC, Acapella, Flutter. There is some evidence to recommend PEP as a more acceptable intervention than other forms of physiotherapy for people with CF. However, the evidence that PEP is preferred over other techniques comes from studies which are generally of low quality.

Reports regarding the use of PEP in CF did not find any significant difference in efficacy when compared to alternative different treatments. Cross-over studies are not considered a good design for clinical studies in CF due to the unstable nature of the disease. More parallel, randomized clinical studies comparing PEP with other airway clearance modalities are needed. Multicentre studies should be adequately approached to improve the power of results. Otherwise, studies have to be planned focusing on clinical stability of eligible patients in order to avoid potential bias.

In the era of HEMT several bias exclude to plan RCTs on this topic.

Keywords

Airway clearance technique; Artificial Ventilation; Chest physiotherapy - Devices; Ventilators;