



The Usage of Positive Expiratory Pressure Devices

Douglas Li, MD, and the CTS Pediatric Committee (DJ Kaley, RN, MSN, CPN, Carmen Reyes, RN, Joann Blessingmoore, MD. Allergy/Pediatric Pulmonology, Gwynne Church, MD. Pediatric Pulmonology/Sleep Medicine Mary Anne Tablizo, MD. Pediatric Pulmonology/Sleep Medicine, Ronald Ferdman, MD. Allergy/Immunology, Daniel Lesser, MD. Pediatric Pulmonology, Cheryl Lew, MD. Pediatric Pulmonology, Matthew Dartt, RCP, RRT-ACCS)

Mechanism of Action

Positive expiratory pressure (PEP) devices create resistance during expiration through usage of a one-way valve (Elkins, 2006). This may improve clearance of mucus by preventing airway collapse during expiration, increasing air volume distal to secretions through collateral ventilation, creating a pressure gradient across mucus, and increasing functional residual capacity (Darbee, 2004). In specific populations with increased sputum viscosity such as cystic fibrosis, there may be an effect on the sputum viscoelasticity (App, 1998). PEP therapy can be combined with high frequency oscillations that are theorized to displace secretions from airways through vibrations and creation shear forces in the airway.

A case report by Ehrlich in 1999 described a single case of an adult male with C3 injury who after using PEP therapy, had a modest improvement in lung function and frequency of respiratory infection (Ehrlich, 1999).

There is not strong evidence to date that PEP improves mucus clearance in patients with severe restrictive lung disease due to neuromuscular conditions, likely due to combined ventilatory muscle weakness and difficulty forming a mouth seal with the device. Insufflation/exsufflation devices remain the mainstay of chest clearance strategies in this population. PEP devices may have benefit for inspiratory muscle training in this population (LoMauro, 2015).

Due to the limited research done in this area, it is not possible to comment on the safety or efficacy of PEP devices in this population.

Asthma

PEP has been studied to a limited extent in patients with asthma and has been hypothesized to prevent distal airway collapse particularly in the outpatient setting. Studies have been small and generally show minimal to no clinical improvement.

When used five times a day, Girard and colleagues demonstrated improved FEV₁, FVC and PEF expiratory flow in the majority of patients with asthma over a 1-month study period (Girard, 1994).

A small study by Navanandan tested 52 children in the emergency department with acute asthma exacerbations (Navanandan, 2017). A positive benefit to short term usage of PEP in this population was not demonstrated.

Due to the limited research done in this area, it is not possible to comment on safety or efficacy of PEP devices in this population.

Device specifics

PEP devices generally have a one-way valve connected to an orifice or an adjustable expiratory

resistor. A manometer is included to measure the expiratory pressure. Tightening the expiratory resistor increases expiratory pressure.

PEP therapy can include an oscillatory device. The transmission of oscillations to the airways decreases the viscoelastic properties of mucus. This in combination with the increased expiratory airflow helps to mobilize secretions. There are PEP multiple devices including the "Flutter" device, "Acapella", "Aerobika", and the "Metaneb" system.

Device Time Cost

Time cost includes approximately 10-15 minutes per treatment excluding other adjunct therapies such as huff cough.

Summary

In summary, PEP therapy seems to be a well-tolerated strategy for improving forced expiratory flow in patients with lung disease. This improved flow translates into variable clinical efficacy depending on the patient population studied. The data most strongly supports regular usage in patients with cystic fibrosis. In patients with cystic fibrosis, PEP devices demonstrated improvement in lung function and decreased pulmonary exacerbations and compared favorably to other mucociliary clearance techniques. In adults with non-CF bronchiectasis, PEP devices have shown to improve cough symptoms and the amount of mucus expectorated. However, data is lacking regarding an impact on health outcomes such as lung function or pulmonary exacerbations. The safety and efficacy of PEP in patients with asthma and neuromuscular weakness is not yet known due to the small sample sizes and small number of trials completed.

Research Supporting Utility

Cystic Fibrosis

The majority of studies in PEP therapy have focused on patients with cystic fibrosis (CF). Generally, PEP therapy in CF has been shown to be effective and safe in children and adults, with likely benefits including improved lung function and reduce pulmonary exacerbations. Several studies of patients with CF compared PEP therapy to other airways clearance techniques.

In 1998, App conducted a study of 14 child and adults with CF, comparing flutter vs autogenic drainage over a 1-month period, and found that sputum viscoelasticity was significantly lower ($p < 0.01$) in PEP vs autogenic drainage groups (App, 1998).

In CF infants, Costantini showed that PEP therapy was well tolerated over a 1-year study period (Costantini, 2001).

McIlwaine compared PEP therapy to percussion and postural drainage in 40 children for a study period of 1 year (McIlwaine, 1997). FVC ($p = 0.02$), FEV₁ ($p = 0.04$) had significant improvement in the PEP treatment group (FVC, +6.57; FEV₁, +5.98) while declines were seen in all parameters in the postural drainage group.

In a 2013 study of 107 children and adults with cystic fibrosis by McIlwaine and colleagues, PEP therapy resulted in a significant reduction in number of pulmonary exacerbations and a longer time to the first pulmonary exacerbation in outpatients compared to those who received high frequency chest wall therapy (McIlwaine, 2013).

In 2015, McIlwaine performed a Cochrane Review of 26 studies comparing PEP therapy to

active cycle breathing, autogenic drainage, oscillating PEP, and high frequency chest wall oscillation in children and adults (McIlwaine, 2015). The primary endpoint of FEV1 showed no significant treatment difference between PEP and other techniques over 3 months. However, there was a lower respiratory exacerbation rate in PEP compared to other techniques over the course of 1 year. Participants did report preference for PEP therapy in 10 studies where preference was measured.

Thus, PEP is a safe and effective therapy for children and adults with CF, with likely effects of improving lung function and reducing pulmonary exacerbations. It has also been demonstrated as a safe therapy for infants with CF.

Non-Cystic Fibrosis Bronchiectasis

Non-cystic fibrosis bronchiectasis is a disease process more prevalent in the adult population, and PEP has been studied as a treatment for this diagnosis with benefits mostly being sputum weight expectorated. This data is limited to the adult population.

In 2009, Murray studied 20 adult patients in a crossover trial comparing oscillatory PEP and no chest physiotherapy. Endpoints included the Leicester Cough Questionnaire (LCQ), sputum volume, and measures of lung function (Murray, 2009). There was a statistically significant improvement with modest effect size in the LCQ (1.3 units) and sputum volume (2ml over 24 hour period).

In 2007, Eaton studied 36 adult patients with non-cystic fibrosis bronchiectasis, comparing efficacy and tolerability of oscillatory pep therapy, active cycle of breathing technique, and postural drainage (Eaton, 2007). Sputum wet weight was significantly improved in patients using active cycle breathing over the other 2 techniques.

Neuromuscular Weakness

Literature supporting usage in pediatrics remains minimal, with possible physiologic benefits, but no demonstrated clinical benefit.

Indications

PEP therapy is indicated in patients with cystic fibrosis as part of an airways clearance regimen.

PEP therapy is not yet indicated in patients with asthma and neuromuscular disease due to lack of data supporting improved clinical outcomes in these groups.

PEP therapy has been used in adults with non-cystic fibrosis bronchiectasis but is not supported for usage in children.

Complications/Contraindications

The only reported adverse event was 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.

The only reported adverse event was in a study where infants performing either PEP or postural drain- age with percussion experienced some gastro-oesophageal reflux. This was more severe in the postural drainage with percussion group.

PEP therapy is generally well tolerated with low risk of pulmonary complications.

The only reported adverse event was in a study where infants performing either PEP or postural

drainage with percussion experience; some gastro-oesophageal reflux. This was more severe in the postural drainage with percussion group.

Future Research Needs

- a) Evaluation of PEP therapy in pediatric patients with larger studies non-cystic fibrosis bronchiectasis, asthma, and neuromuscular disease with clinical outcomes. Outcomes suggested include oxygenation, frequency of illnesses/antibiotic usage, respiratory quality of life, and measures of lung function.
- b) Evaluation of PEP therapy for use in hospitalized pediatric patients with bronchiectasis, chronic infections, asthma and neuromuscular disease – include documentation of change in work of breathing indices, oxygenation and or portable pulmonary function tests before and after use, change in chest radiograph findings.