

CLINICAL SCENARIO

Cystic Fibrosis

Cystic Fibrosis (CF) is an inherited disease that causes thick, sticky mucus to form in the lungs, pancreas and other organs. When mucus overwhelms the lungs, the airways become blocked, causing lung damage, breathing difficulty and infection. Bronchiectasis, dilated bronchioles, can be a component of CF, creating a vicious cycle of excessive secretions overwhelming the mucociliary clearance system and obstructing the airways, sometimes causing infection. While lung problems associated with CF cannot be cured, use of the appropriate products can ease symptoms and help to maintain quality of life.

	Criteria	Benefits of therapy
acapella® Vibratory PEP System	Chronic secretions or hypersecretions	Vibrations decrease viscosity and loosen or dislodge secretions
	Air trapping secondary to secretions/hypersecretions	PEP therapy stents open airways, provides collateral ventilation
	Medication delivery	Combine PEP and medication delivery
	Age 4 or older	Self-administered by patient, appropriate for home use, high compliance, versatile
	Need for multiple therapies	Taken apart and cleaned per CF Foundation recommendations Dual therapy depending on patient personal needs, with TheraPEP® in between acapella® treatments
TheraPEP® PEP Therapy System	Secretions	Move secretions from distal to central airways, provides pressure distal to secretions
	Age 4 or older	Easy to use, self-administered by patient, appropriate for home use Compact and portable, leading to high patient compliance
	Need for multiple therapies	Adjunct therapy with other airway clearance devices PEP therapy, stents open airways, provides collateral ventilation Passive method of action for those patients who can't complete therapy independently
EzPAP® PAP Therapy System	May be appropriate during periods of exacerbation involving increased work of breathing	Avoid more invasive therapies
Palm Cups® Chest Physiotherapy	Secretions	May loosen secretions
	Age dependent	No age barrier

Targeted therapies, reliable results



acapella® Vibratory PEP System

The acapella® System combines the benefits of PEP therapy with airway vibrations to mobilise the pulmonary secretions that can obstruct airways, decrease lung function and cause infection. With patients free to stand, sit or recline, this therapy is better tolerated than chest physiotherapy (CPT) and can be delivered in about half the time.

Finding:

The oscillation frequencies of PEP therapy and acapella® are within the range demonstrated to improve secretion clearance, providing some evidence for their use during physiotherapy with CF patients.

Study:

McCarren B, Allison JA. **Physiological Effects of Vibration in Subjects with Cystic Fibrosis.** Eur. Respir J. 2006;27:1204-1209

Finding:

Airway clearance therapy is recommended for all patients with CF for clearance of sputum, maintenance of lung function and improved quality of life.

Study:

Flume PA, Robinson KA, O'Sullivan BP, et al. **Cystic fibrosis pulmonary guidelines; airway clearance therapies.** Respir Care. Apr 2009;54(4):522-537



TheraPEP® PEP Therapy System

Designed for PEP therapy and simultaneous aerosol drug delivery, the TheraPEP® System allows air to enter the lungs, get behind the mucus and push it up to the central airways. It is completely portable, and can be used in conjunction with a nebuliser.

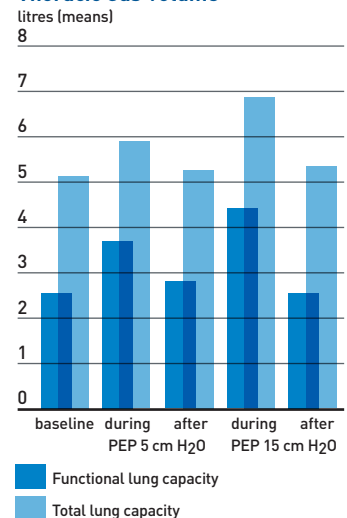
Finding:

In patients with CF, positive expiratory pressure breathing increases lung volumes in relation to the expiratory pressure imposed, opening collateral pathways or small airways in obstructed parts of the lungs.

Study:

Van der Schans CP, van der Mark TW, de Vries G, et al. **Effect of positive expiratory pressure breathing in patients with cystic fibrosis.** Thorax. Apr 1991;46(4): 252-256

Thoracic Gas Volume



Finding:

In patients with CF, PEP is recommended over conventional chest physiotherapy because it is approximately as effective and is inexpensive, safe and can be self-administered.

Study:

McCool FD, Rosen MJ. **Nonpharmacologic Airway Clearance Therapies: ACCP Evidence-Based.** Chest. 2006;129;250-259

Both acapella® and TheraPEP® Systems are designed to:

- Maintain lung function when chronic secretions or hypersecretions are a problem so that patients can better tolerate activities of daily living
- Clear sputum to decrease the chance for infection
- Be well tolerated compared to other therapies
- Contribute to better patient compliance and quality of life
- Be used in conjunction with other therapies

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