Airway-Clearance Therapy Guidelines and Implementation

Mary K Lester RRT and Patrick A Flume MD

The clearance of secretions from the lungs of patients with cystic fibrosis (CF) is an important component in the fight to preserve their lung function. There is excessive inflammation in the airways of these patients, which is thought to be exaggerated by ineffective mucociliary clearance and bacterial infection. In 2005 the Cystic Fibrosis Foundation formed the Pulmonary Therapies Committee to review all of the medical literature on the various airway-clearance therapies used in treating CF lung disease. The recommendations were: an airway-clearance therapy should be performed by all patients with CF, no form of airway-clearance therapy stood out as being superior to another, and that patients may express a preference of one therapy over another. They also concluded that aerobic exercise is beneficial to patients with CF, as it is to everyone, and that exercise should be a component to the overall health routine of patients with CF. The challenge for respiratory and physical therapists together with the patient/family is to develop a plan of attack through the use of various airway-clearance therapies. The respiratory and physical therapists are integral in helping patients and families develop airway-clearance routines that aid in the removal of the secretions that cause airway obstruction. There is a wide range of airway-clearance therapies that therapists can choose from when they are teaching the patients and family members the strategies of secretion removal. The questions are: What therapy is best for what age or stage of lung disease? What therapies will the patient do? And which therapies will be covered by medical insurance? These are all fundamental questions that must be answered when guiding families in finding therapies that are effective and appropriate for each CF patient's unique situation. *Key words: cystic fibrosis, airway-clearance therapies, secretion removal, airway-clearance teaching, airway-clearance guidelines.* [Respir Care 2009;54(6):733–750. © 2009 Daedalus Enterprises]

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Introduction

Airway-clearance therapies (ACTs) have long been considered an important part of care of the patient with cystic fibrosis (CF). This is based upon our understanding of the pathophysiology of CF lung disease. There is ineffective mucociliary clearance[1] and, from very early in life, there is obstruction of the small airways by mucus.[2] Mucociliary clearance is an important defense mechanism of the airways, and patients are vulnerable to chronic infection of the airways. There is an exaggerated inflammatory response, with the airways containing bacteria, inflammatory cells, and cellular breakdown products (eg, neutrophil-derived deoxyribonucleic acid and filamentous actin (F-actin).[3-6] All of this material increases the tenacity and stickiness of the airway secretions, and the airways become further obstructed.[7,8] Since the mucociliary clearance system isn't working, it makes sense to use other methods to help clear the airway secretions, if only to relieve the obstruction. In addition, these secretions contain bacteria and inflammatory mediators that continue to injure the airways as well as recruit new inflammatory cells. Therefore, clearance of airway secretions does more than merely relieve obstruction; it also reduces the amount of infection and inflammation present in the airways.

What Is Airway Clearance?

ACTs are physical or mechanical means of facilitating the removal of tracheobronchial phlegm through the external and/or internal manipulation of air flow, and the evacuation of phlegm via coughing. One of the earliest forms of therapy, which continues to be a primary method, is chest physiotherapy, or percussion and postural drainage. In 1959, British physiotherapist Jocelyn Reed first reported that "clapping and pressure vibrations, during long expirations, are the most effective forms of mechanical stimulus to elimination of secretions in the treatment of lung abscess, collapsed lobes, and bronchiectasis."[9] ACTs have evolved over the years, using imaginative strategies such as high-frequency chest-wall compression using an inflatable vest connected to an air compressor, hand-held expiratory vibratory devices, and, more recently, acoustic waves to vibrate the mucus from the airway walls. The respiratory therapist has an extensive menu of airway-clearance modalities from which to choose (Table 1), each with advantages and disadvantages. The intent of this paper is not to discuss the techniques for delivering the numerous airway-clearance modalities; however, we have included the instructions that we give to our patients and families on these therapies (Appendix). The intent is to help guide the therapist in choosing an ACT that is appropriate for a CF patient at different stages of life. The challenge for the therapist is to match the therapy with the patient so that there is the most effective clearance of airway phlegm.

Cystic Fibrosis Pulmonary Guidelines

The Cystic Fibrosis Foundation established the Pulmonary Therapies Committee to develop guidelines for medications and therapies to maintain the lung health of patients with CF. Thus far, there are published recommendations for medications used chronically[10] and for ACTs.[11] A systematic review of the published literature was used to develop these recommendations. An important realization in the development of the ACT guidelines was the lack of large, long-term studies of airway clearance. It is not that there isn't any evidence to support ACT for patients with CF; there are, in fact, a large number of studies, but most have small numbers of patients, are performed over short periods of time (some only a single treatment), and many have no comparator. A prior systematic review concluded that ACT increases mucus transport for the short-term, but the authors could not draw conclusions about the long-term effects or benefits.[12] Another review noted that, despite the "dearth of high-level evidence to support any secretion clearance technique," the lack of evidence does not mean lack of benefit.[13] The Pulmonary Therapies Committee felt that the existing evidence supporting ACT compared to no therapies was fair in quality overall. They expressed concern that because ACT is so fundamental to CF treatment, a large controlled trial designed to explicitly prove the benefits of ACT could not be completed. They determined that the overall benefit of ACT was moderate, based upon the various outcomes that had been studied with a variety of therapies, including increased sputum production, increased lung function in the short-term, reduced rate of decline of lung function, and increased exercise tolerance. In the end, they recommended that some form of ACT be performed as a routine in all patients.[11]

There were other important findings in the systematic review that led to specific recommendations by the committee. First, there weren't any data to suggest that one type of ACT was better than any other. Second, patients will express preferences, and it is believed that patients are more likely to use therapies they believe to be beneficial. Finally, aerobic exercise seemed to offer benefits to a regimen of airway clearance in addition to the other well-known benefits of exercise.[14]

Implementing the Airway-Clearance Therapy Guidelines

Now that we have recommendations regarding ACT, we must address how to implement them in our CF clinics. We must find a way to communicate these recommendations to our patients and their families, and help them find

Table 1. Methods of Airway Clearance

Therapy	Age Range	Advantages	Disadvantages
Percussion and postural drainage	All ages	No cost/no equipment Target specific areas of lung	Requires caregiver Tiresome Does not promote independence
Blowing games	18 mo to 5 y	Low cost Prepares child for future spirometry	Patient must have appropriate cognitive ability
Appropriate coughing	≥ 18 mo	Teaches to cover mouth Promotes good hand-washing	None
High-frequency chest compression	≥ 2 y	Promotes independence Portable Administer nebulized medications at same time	Expensive
Huff cough	≥ 3–4 y	Gentle coughing technique	None
Active cycle breathing technique	≥ 4 y	Independent, inconspicuous Can be performed anywhere, anytime	Patient must have appropriate cognitive ability
Autogenic drainage	≥ 8 y	Independent, inconspicuous Can be performed anywhere, anytime	Patient must have appropriate cognitive ability
Handheld devices, PEP, oscillating PEP	≥ 4 y	Low cost Promotes independence Some devices allow nebulized medications	Patient must have appropriate cognitive ability
Intrapulmonary percussive ventilation	≥ 5 y	Can nebulize medications while performing	Expensive

PEP = positive expiratory pressure

a method of ACT that works best for them. Given the third recommendation that patient preference matters, it seems rational to introduce the patients to all of the available therapies. Then they can make an informed decision as to which of the therapies they are most likely to use. However, there are important factors in the choice of therapies. For example, an infant is not capable of performing most of the available therapies and is dependent upon others (ie, parents) to perform the treatment, typically percussion and postural drainage. On the other hand, a person who lives alone will not have a partner to assist with percussion and postural drainage, and so will probably benefit more from a type of ACT that can be performed independently (eg, high-frequency chest compression). Therefore, there are a number of factors to consider regarding recommendations of specific ACT, including independence, effectiveness, ease of use, flexibility, treatment durations, comfort, convenience, and interruption to daily living. It has been our experience that patients will benefit from more than one type of ACT, and so we introduce them to all of the options at an appropriate time of their life (Fig. 1). Before discussing how to introduce various therapies to patients, it is useful to express our other basic tenets of ACT:

1. *The physician prescribes the therapy but does not necessarily know what is best.* The CF team typically includes an expert in ACT, most often a respiratory therapist or a physical therapist. The therapist's role is to work with the patient and family to determine the most appropriate ACT and educate them as to its proper performance.

2. *Airway clearance is an active process.* Most patients will be performing their therapies on themselves in the out-patient setting. Even in the hospital setting, the therapist or parent who is performing ACT on a patient works with the patient to assist clearance of the airway secretions. The patient should play an active role in making the therapy effective by coughing throughout and performing deep breath-holds and forced expiratory maneuvers at appropriate times. No single method of airway clearance is better than another. This is similar to the CF Pulmonary Therapies Committee's second recommendation. Although no therapy has been shown better than the others, this does not mean that one form of therapy won't prove superior to the others for the individual patient. What is also apparent is that one form of ACT may prove useful at one time, but another will be more effective at another time. Similarly, patients may be able to use one form of therapy when they are home (eg, percussion and postural drainage) but may need a different therapy when they are away from home living alone (eg, college).

3. *Airway clearance is boring.* There is no way around this reality. ACT is tedious, monotonous, and takes time to perform effectively. This is another reason for patients to have access to more than one form of therapy.

4. *Be creative.* This has more than one meaning. First, there are a number of fun activities and toys for children that can serve as a method of airway clearance. Take a walk down an aisle of party favors and look at all of the toys that behave like positive-expiratory-pressure (PEP)

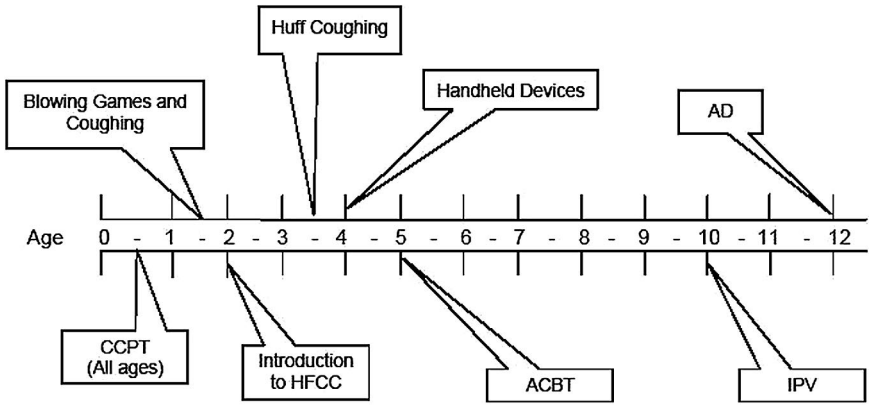

Fig. 1. Not all airway-clearance modalities are appropriate for all ages. We have a strategy of introducing airway-clearance techniques to all patients at our center. We introduce the various airway-clearance techniques based upon the patient's age and ability to perform the therapy. This timeline approximates the appropriate age the therapy can be introduced. AD = autogenic drainage. CCPT = conventional chest physiotherapy. HFCC = high-frequency chest compression. ACBT = active cycle breathing technique. IPV = intrapulmonary percussive ventilation.

devices. Second, we ask our patients to perform lengthy therapies several times daily. We need to work with them to help them figure out a way to get these therapies into their routine. Listen to the patients to learn how they spend their day, either at work or at school, and discuss ways to incorporate ACT into that schedule. We must try to adapt the therapies to the patient's needs and let the therapies be less interruptive to their daily living.

Implementing Airway-Clearance Therapies: A Timeline for the Newly Diagnosed Patient

The infant (birth to 18 months old) is completely dependent upon the caregiver (eg, parent) for performance of ACT. Most of the other therapies are inappropriate for the infant. Therefore, the parents of the newly diagnosed infant with CF should be introduced to percussion and postural drainage. The therapist will meet with the parents initially to teach them the technique, and will meet with them at subsequent clinic visits to evaluate their technique and re-educate as needed.

Toddlers (18 months to 4 years) are often ready to try high-frequency chest compression. There are currently 3 high-frequency chest-compression devices available on the market: The Vest (Hill-Rom, St Paul, Minnesota), InCourage (Respirtech, St Paul, Minnesota), and SmartVest (Electromed, New Prague, Minnesota). These devices are expensive, and so we confirm with the patient's insurance carrier that such durable medical equipment will be covered. We introduce each of the devices to the patient and family at a clinic visit or in the hospital; the parent or caregiver will then be able to make an informed decision on which machine to choose, if it is clear the patient will use the device. Also, at this young age the patient should be taught blowing games (eg, pinwheel, bubbles) to prepare them for the prolonged exhalation maneuvers they will need to master prior to their first spirometric test. Huff cough instruction can also start during this time.

School-age children (5 years to preteen) can begin basic active cycle breathing techniques. Patients can be taught breath-holds and prolonged exhalation maneuvers. The huff cough or forced expiratory technique can be mastered at these ages, and portable devices such as PEP or oscillating PEP (eg, Acapella or Flutter) should be used. Patients need to have access to portable devices for use while away from home, such as at camp, on sleep-overs, or while being driven to school. Patients this age should be encouraged to participate in sports. Exercise needs to be a part of CF patient's daily routine. Not only does exercise improve cardiovascular health and self esteem, it can also act as a method of removing secretions.[15] Some patients report that when they run, bike, or swim, these movements cause a vibration and loosening of the mucus in the chest. There have not been enough controlled trials to demonstrate that exercise is an effective form of airway clearance, but the Pulmonary Guidelines Committee found that exercise should be considered an adjunct to a patient's airway-clearance routine.

Preteen to early teen CF patients express a need to be "not different" from their peers. It is common for parents to complain that their child "will not do anything" when asked about airway clearance. This is a very challenging time in life because teenagers want to be independent and to make their own decisions, and often they choose to skip performance of ACT. But this is the most vulnerable time for patients, and one in which we see a greater rate of decline in their lung function.[16] Patients at this age can improve on the techniques of active cycle breathing technique and huff cough, and learn autogenic drainage. When done properly, these techniques are effective and they can

be done inconspicuously. We must negotiate with the teens to help them establish a routine that is effective, convenient, and causes little interruption in their lifestyle. They should learn that breathing techniques can be done at any time and they do not require an external device or caregiver.

By the time they reach adulthood (age 18 years and up), patients with CF will have been introduced to the entire ACT armamentarium, including percussion and postural drainage, high-frequency chest compression, PEP, oscillating PEP (eg, Acapella), autogenic drainage, active cycle breathing technique, and intrapulmonary percussive ventilation. All adults should have more than one technique available to them, and we must work with them to establish a routine that fits with a busy lifestyle that includes school, work, and family time. As their lung disease reaches a more severe stage, they must realize that they are likely to require increased time spent with ACT.

Implementing Airway-Clearance Therapy: The Patient Who Transfers to Your Center

Like many others, people with CF may move to a different city, and will transfer their care to a different CF center. When we see a new patient for the first time, we try to learn what therapies the patient is currently performing and assess their technique, providing tips for improvement as needed. We also perform a survey of their knowledge of all of the other therapies. Do they have knowledge of the other therapeutic options? Have they tried them? What did they like or not like about those therapies? We will then make plans for a future clinic visit to introduce them to the therapies they have not tried, following the same timeline noted above.

Implementing Airway-Clearance Therapy: The Hospitalized Patient

Patients with CF may experience acute worsening of symptoms, such as increasing cough and sputum production, often called a pulmonary exacerbation. The CF pulmonary guidelines on treatment of a pulmonary exacerbation suggest that ACTs may need to be changed during such an event.[11] In situations where the exacerbation is mild and out-patient therapy is planned, the patient can be advised to increase the time spent with therapy, or to perform the therapies more frequently, or even to consider an alternative therapy. For those who are ill enough to warrant admission to the hospital, there is an opportunity for therapists to become fully engaged. At the start of the hospitalization it is most important to begin with a therapy the patient finds the most comfortable. It is a time to establish early success in clearance of secretions and to establish a trusting relationship between the therapist and the patient. As the patient improves, this is now an opportunity to evaluate the patient's ability to perform airway clearance on their own as well as introduce therapies they have not yet tried.

A Role for Exercise

A question asked frequently in the CF clinic is, "Can my 30 min of running take the place of my ACT?" Indeed, there are many patients with CF who participate in regular exercise activities that are fairly rigorous. The Pulmonary Guidelines Committee recommended aerobic exercise as an adjunctive therapy for airway clearance. Although there was not sufficient evidence to conclude that exercise was as effective as other ACT, there was evidence that exercise does result in clearance of secretions and there is improvement in quality of life.[15] There are additional benefits to aerobic exercise: increased endurance, increased self esteem, and improved cardiovascular health.[14] Patients with CF who have greater fitness have better survival.[17] It is our perception that the patients in our clinic who exercise regularly tend to have better overall health.

Summary

The CF Pulmonary Guidelines Committee came to the same conclusion that most clinicians had already believed: that performance of techniques of airway clearance works for patients with CF. We do not have evidence that any single method of ACT is best for all patients, but we are fortunate to have many therapies from which to choose. The opportunity for the therapist and the entire CF team is to work with the patients and families to find the therapies that work for the individuals, given the myriad of resources they have and the hurdles they face. It is important to listen to the patients as they will help us figure out what works best for them, and they will teach us tricks that will work for other patients as well.

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Appendix

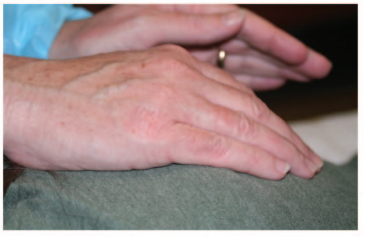
Chest Physiotherapy

What is chest physiotherapy (CPT)?

CPT is a way to help get secretions out of the airways in order to improve airflow inside the lungs. This is achieved most successfully by applying percussion and vibration to the chest wall in positions that allow for proper drainage. There are 3 main components of CPT, including percussion, vibration, and postural drainage.

Percussion

Percussion is a rhythmic clapping on the exterior chest wall. The proper technique involves cupping of the hands (or fingers), working from the wrists, and "clapping" the chest wall over a specific area. Percussion is done over one area and position at a time and for at least 1 minute. The duration, clapping frequency, and pressure of the percussion are dependent upon the age, need and tolerance of the patient.



Vibration

Vibration immediately follows percussion of each area and position. Lay one of your hands over the other hand and place them on the chest over the area that was percussed (alternatively you may place your hands on either side of the chest wall). Tense up (or contract) your shoulder muscles and exert some pressure to cause your hands to move rapidly (vibrate). Instruct the child to take in a deep breath and then apply the vibrations while they are exhaling. Effective vibration will transmit through the chest wall, help in loosening secretions, and promote a spontaneous cough.

Postural Drainage

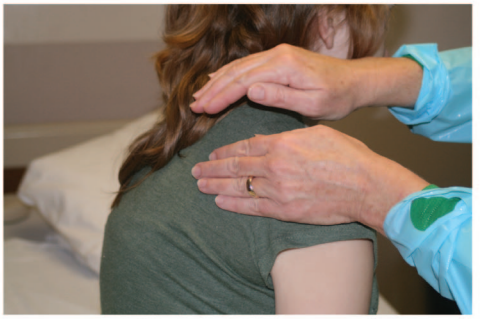
Placing your child in specific positions allows for gravity to assist in the movement of secretions to the large airways (main bronchi), where they can then be coughed out of the lungs. We discourage positions that include head-down positioning as this can contribute to gastroesophageal reflux (GER), which is the regurgitation of stomach contents back up the esophagus.

CPT can be done before meals or at least 1 hour after eating. Spend 2 minutes on each segment, or longer in a specific area of the lung if it is problematic. The entire therapy session should last 30 minutes. Patient participation during the session can include deep breath holds while applying percussion to a certain area, then a forced exhalation can aid in dislodging the secretions.

Chest Physiotherapy

Upper lobes

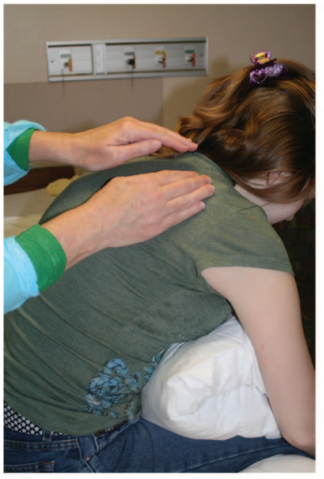
Apical segments: Have the child sit upright and percuss over the shoulder area on each side. This may be done from the front (Panel A) or from the back (Panel B), whichever is more comfortable.



Panel A

Panel B

Posterior segments: Have the child lean forward (Panel C). Percuss over upper back on both sides. Avoid percussing over the spine.



Panel C

Anterior segments: Have the child lay flat on back. Percuss just under the collarbone (Panel D). Avoid percussing over the sternum.

Panel D

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Chest Physiotherapy

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Right middle lobe and left lingula

Have the child lying on her left side with head supported by a pillow. Lift the arm above the head and percuss over the middle area of the right chest, just under the armpit (Panel E).

Panel E

The child lies on her right side and the left arm is placed behind her (Panel F). Percuss over the middle area of the ribs.

Panel F

Chest Physiotherapy

Lower lobes

Superior segments: Have the child lie flat on her stomach. Prop pillows under the chest (Panel G). Percuss over the shoulder blade area on either side of spine. Then prop pillows under the hips (Panel H) and percuss in the same area.

Basal segments: Place the child on her side supported by a pillow and have her lean forward (Panel I). Percuss over the back and side. Then have the child move the pillow to support her head (Panel J) and percuss in the same area. Repeat on both sides.

Panel G

Panel H

Panel I

Panel J

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Blowing Therapies

What are blowing therapies?

Blowing or breathing games encourage toddlers to take deep breaths and manipulate exhalation. This helps with clearing secretions from the airways, but also helps the child to become more aware of his or her breathing and prepare for future pulmonary function testing.

Examples of blowing therapies

Blowing Bubbles – have the child take a deep breath and gently exhale. As the child masters bubble blowing, this will encourage a longer exhalation time.

Bubble Water – Place a straw in a glass of water. Have the child blow into the straw and make lots of bubbles. This produces positive expiratory pressure (PEP) and encourages a long exhalation time.

Toys that promote deep breaths and long exhalation:
- Harmonica
- Pin Wheel
- Hunting Bird Whistles

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High Frequency Chest Compression

What is high frequency chest compression (HFCC)?

HFCC uses an inflatable vest that covers the chest and attaches with hoses to an air-pulse generator. The generator inflates and deflates the vest at different frequencies (Hz). These oscillations compress and release the chest wall, thus manipulating airflow to dislodge secretions and moving them towards the larger airways where the secretions can be coughed out.

Selecting a vest

Vest therapy promotes a more independent form of airway clearance but can be introduced when the child is around the age of 2 years. There are 3 HFCCs currently on the market (see below). We will arrange for a trial of each of the vests so that you may decide if this is a preferred therapy. If you choose HFCC, we will place the order with the vest company that you have selected. They will contact you to verify insurance coverage and your address, and to schedule delivery of the device. A representative of the vest company will call to arrange a training session. This training should be done in person and before you begin to use the device regularly.

- Hill Rom Vest @ www.thevest.com
- InCourage System @ www.respirtech.com
- Smartvest @ www.electromed.com

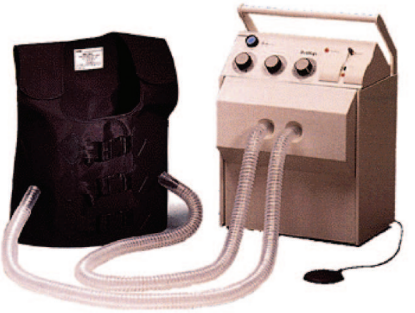
The basics of HFCC

1. An inflatable vest is attached by tubing to an air – pulse generator.
2. Vest size is determined by measuring chest circumference at the nipple line. The sizes range from 16-60 inches.
3. When ordering the device, talk to the company representative to determine vest style options.
4. The vest inflates and deflates using high frequency oscillations, which vibrate the airflow in the airways to loosen secretions.
5. The basic controls on the vest models are:
   a. Frequency of vibration in Hz (hertz)
   b. Inflation pressure of the vest
   c. Timer
6. Vest therapy has evolved from using a single vibration or Hz setting to using various Hz settings, recognizing that changing the Hz frequency and vest pressure amplitude can optimize the efficiency of vest therapy.
7. Vest therapy sessions were originally thought to be a passive therapy; that is, patients sat through a 30-minute session while inhaling nebulized medications. Current vest therapy protocol should include:
   a. Changing the frequencies.
   b. Pausing the therapy session for active coughing.
   c. Delivering inhaled medication throughout the therapy session.
   d. Inhaled antibiotics and corticosteroids should be taken after completion of vest therapy session.

High Frequency Chest Compression

Example of Ramping Session using the Hill Rom Vest

Frequency	Pressure	Time
6, 8, 10 Hz	10	• Five minutes at each Hz setting • Pause machine and cough 3 times • Resume session
16, 18, 20 Hz	6	• Five minutes at each Hz setting • Pause machine and cough 3 times • Resume session

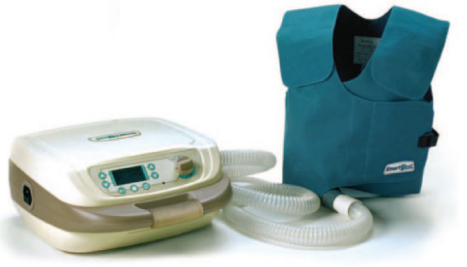


Therapy Session with InCourage System

- Push QuickStart button to initiate pre-programmed 30 minutes automatic ramping session
- Pause button may be pushed at any time to allow for cough
- Push Run button to resume therapy from point that therapy was previously paused
- Pressure can be increased or decreased during therapy session

Standard Protocol for SmartVest

Frequency	Duration
10 Hz	10 minutes, then huff and cough
12 Hz	10 minutes, then huff and cough
14 Hz	10 minutes, then huff and cough



Active Cycle Breathing Technique

What is active cycle breathing technique (ACBT)?

ACBT is a breathing technique that utilizes breathing control, thoracic expansion (deep breathing), and forced expiratory techniques (huff coughing) to loosen airway secretions. By alternating deep breaths with breath holds and forced exhalations, the secretions are moved from small airways to large airways where they can be coughed out.

Patients can perform this technique in different positions, but for training purposes it is best to be sitting straight up and relaxed in a chair.

The basic cycle is as follows:

1. **Breathing control or gentle diaphragmatic breathing**: relax the shoulders and breathe in through the nose and exhale through the mouth with 3-4 normal sized breaths.
2. **Thoracic expansion (deep breath):** make a full inspiration followed by a 3-4 second breath hold, then a relaxed expiration
3. **Back to breathing control** for 3-4 breaths**.**
4. **Thoracic expansion and breath hold** for 3-4 breaths.
5. **Forced exhalation, or huff**, for 2-3 exhalations to cough up secretions.
6. **Repeat** these steps for several cycles until airway secretions have cleared.

Huff Coughing: Normal coughing may cause bronchial closure, use of excessive energy, and little sputum production. Huff coughing is a gentle coughing technique that includes 3 mini huff coughs to loosen secretions from the peripheral small airways and a final forced huff to expectorate the sputum.

1. Deflation Breaths - have patient deflate excess air from their lungs by slowing breathing rate down and performing prolonged exhalation (4-5 seconds) for 3-4 breaths.
2. Take a slow deep breath but not a maximum breath.
3. Shape your mouth like an “O” .
4. Do a mini / short cough by contracting the upper abdominal muscles. This can be described as pushing a tennis ball out of an open mouth while making a “huff” noise.
5. Take in a quick partial breath and repeat “huff” a second time with this smaller breath.
6. Take in an even smaller breath and “huff” for a third time. (The patient is now at a very low lung volume that helps loosen the secretions from the lungs.)
7. Take a forced full breath but not a maximum breath.
8. Give a forced, hard “huff” and cough out the sputum.
9. Repeat the steps if airway secretions are not cleared.

Autogenic Drainage

What is Autogenic Drainage?

Autogenic drainage was first described by Jean Chevaillier in 1967. It is a breathing technique that helps the patient drain the lungs from within (autogenic) by breathing at different lung volumes in 3 phases:

Phase 1 – unsticking / loosening the phlegm in the small airways
Phase 2 – collecting / moving phlegm to the middle airways
Phase 2 – evacuating / moving the phlegm out by huffing

Unsticking

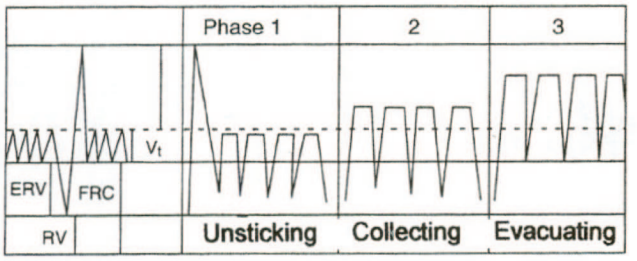
1. Sit in a relaxed position; clear your nose and throat of secretions.
2. Start the session with breathing control, breathing in through the nose and out through the mouth.
3. Inhale through the nose and take in a deep breath and hold for 3-4 seconds; then exhale air out of the mouth for as long as you can to reach low lung volume.
4. Inhale through the nose a small breath and hold 3 seconds.
5. Exhale through the mouth and squeeze out as much air as you can.
6. Repeat this low lung volume breathing at least 3 times or until you:
   hear the secretions crackle while exhaling
   feel the secretions moving, or
   feel an urge to cough.

Collecting

1. Take a deep breath and hold for 3-4 seconds and exhale (but not as low as in the unsticking phase).
2. Inhale through the nose a slightly larger breath and hold for 3-4 seconds.
3. Exhale through the mouth a normal sized exhalation.
4. Repeat normal-sized breaths (with breath hold after inspiration and a normal exhalation) for 3 breaths.

Evacuating

1. Take a deep breath and hold for 3-4 seconds.
2. Exhale forcefully with a huff cough.



Autogenic Drainage

Each phase should take approximately 2-3 minutes. Coughing should be avoided until the evacuation phase, but if patient must cough, they should do 2-3 huff coughs. As patient becomes more comfortable with autogenic drainage, they will probably learn to move secretions by developing their own personal method.

Huff Coughing: Normal coughing may cause bronchial closure, use of excessive energy, and little sputum production. Huff coughing is a gentle coughing technique that includes 3 mini huff coughs to loosen secretions from the peripheral small airways and a final forced huff to expectorate the sputum.

1. Deflation Breaths - have patient deflate excess air from their lungs by slowing breathing rate down and performing prolonged exhalation (4-5 seconds) for 3-4 breaths.
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6. Take in an even smaller breath and "huff" for a third time. (The patient is now at a very low lung volume that helps loosen the secretions from the lungs.)
7. Take a forced full breath but not a maximum breath.
8. Give a forced, hard "huff" and cough out the sputum.
9. Repeat the steps if airway secretions are not cleared.

PEP Therapy

What is Positive Expiratory Pressure (PEP) therapy?

Hand-held Positive Expiratory Pressure (PEP) devices are used as a form of airway clearance by having the patient exhale through an expiratory resistor, which creates positive pressure or back pressure in the lungs. This positive pressure stents the airways open and helps get air behind phlegm and expectorated by coughing. The Acapella, Flutter, and Quake devices also have an oscillating component that causes the airway walls to vibrate during exhalation thus loosening the secretions.

Technique

1. Have patient in a sitting position.
2. Do a couple of relaxed breaths – air in through the nose and out through the mouth.
3. Take a slightly deeper than normal breath and hold for 3 seconds.
4. Exhale through the device with lips tight on the mouthpiece and cheeks tight for 3-4 seconds.
5. Repeat 10-12 breaths.
6. Perform 3-4 Huff coughs.
7. Repeat the cycle 5-6 times for approximately 15-20 minutes.

Huff Coughing: Normal coughing may cause bronchial closure, use of excessive energy, and little sputum production. Huff coughing is a gentle coughing technique that includes 3 mini huff coughs to loosen secretions from the peripheral small airways and a final forced huff to expectorate the sputum.

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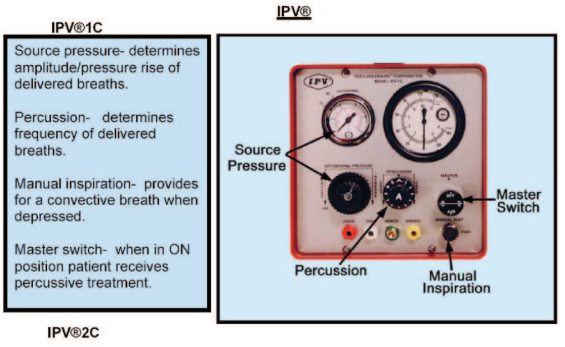
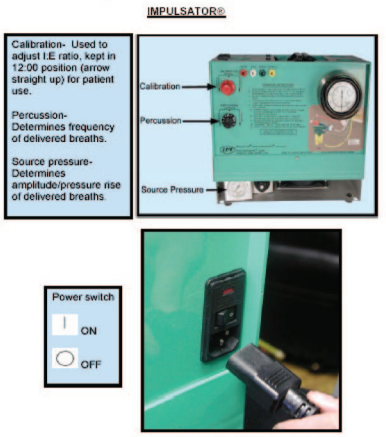
Intrapulmonary Percussive Ventilation

What is intrapulmonary percussive ventilation (IPV)?

IPV is a form of airway clearance that uses a pneumatic device (Phasitron) to deliver high flow, mini bursts of air to the lungs at rates of 100 to 300 per minute. An aerosol mist of 1 mL per minute is delivered during the percussion, which loosens the retained secretions. IPV is to be used on patients who have increased sputum and are unable to clear secretions effectively on their own.

IPV is a hospital-based unit that runs off of a 50 psi gas source (left figure)
The Impulsator is the home-based unit that runs off of electricity (115V / 60Hz) (right figure)





Therapy Session

- Have the patient sit in an upright position.
- Assemble the circuit in accordance with manufacturer's directions.
- Fill the nebulizer with prescribed medications (1 mL per minute of therapy time).
- Start the frequency at the "Easy" position which is 300 Hz.
- The beginning operating pressure should be 20-25 psi, or until chest wall movement is seen and patient remains comfortable.
- Have the patient breathe through the circuit for 1-2 minutes before depressing the Phasitron button, which will trigger percussions.
- Have the patient inhale and exhale through the percussions, keeping their lips sealed on the mouthpiece and cheeks tight.
- The patient can rest between percussion sessions by releasing the percussion button and can continue breathing the aerosolized medication.
- The frequency can be decreased by 50 Hz every 5 minutes, working to 100 Hz, which would be the "Hard" position.
- Huff coughing should be performed every 5 minutes or as necessary.

Discussion

Geller: It's great to hear somebody talk about the practicalities of how to get people to do airway clearance, which is neglected by a lot of people, and it's really fundamental. You mentioned Rule 4, airway clearance is boring, and Rule 5, think outside the box. I ask my patients, "Have you figured out a way to make airway clearance fun?" As of yet, I have not had a single person give me an answer other than, "I sit in front of the computer or TV," and that's their exercise too: using the remote control.

Some of my healthiest patients play wind instruments: trumpet, trombone, saxophone, or clarinet, but not flute: instruments that create some resistance. With wind instruments, they're basically performing high-pressure PEP, and it's a wonderful airway-clearance technique. Usually a practice session lasts about 30 minutes to an hour a day, and if they like the instrument, then it's fun. That's one way to make airway clearance fun.

Exercise is an adjunct to therapy, and when they get old enough, they can join the marching band and play and march and get exercise at the same time. That's the only thing that I've found, besides putting a vest in the car, that might actually be fun, at least for some patients. Unfortunately, when they graduate high school, they usually drop the instrument and don't play any more, and then lung function deteriorates because they never learned any other technique. But it was fun while it lasted.

Lester: On YouTube an airway-clearance video showed a duck-calling contest, and the 3 finalists were CF patients. They exhaled through a vibrating duck whistle. They had fun with it and probably didn't know that they were doing airway clearance. Vest therapy can be a passive therapy, and when I went to Minnesota last year for adult benchmarking, they used the Minnesota Protocol,[1] in which they increase the frequency throughout the treatment, and every 5 minutes they pop a hose and do some coughing. It adds time to the therapy session, but it makes the session more active, because they're doing true airway clearance by stopping to cough. I've had some good response with that treatment plan. Some patients really take on the Minnesota Protocol, and they do their nebulizer treatments at the same time. That makes vest therapy more active than passive.

REFERENCE

1. Kempainen RR, Williams CB, Hazelwood A, Rubin BK, Milla CE. Comparison of high-frequency chest wall oscillation with differing waveforms for airway clearance in cystic fibrosis. Chest 2007;132(4):1227-1232.

Geller: After vest therapy, how important is the addition of postural drainage? They're usually sitting upright during vest therapy, so how important is it to add postural drainage, for someone who has ineffective cough?

Lester: I've never suggested that. Sometimes at the hospital we might add CPT [chest physiotherapy] and postural drainage to target specific problem areas of the lung.

O'Malley: If you want to adversely impact adherence, asking them to do 2 consecutive treatments would probably be the way. I wouldn't ask a patient to do 30 minutes of vest therapy and then spend more time doing *another* therapy, at least not in home management.

Lester: We had a 5-year-old CF patient recently, and her main therapy was vest therapy. While she was an in-patient, we thought it was a good time to try different therapies. She was so sick and had a lot of secretions, and the therapist thought vest therapy was not being effective, so they tried chest physiotherapy and postural drainage and got a lot of mucus out. If one therapy is not working, try another. I have not had any patients who felt like doing drainage positions after vest therapy.

Flume: Or during.

Lester: Sure, or during.

Ratjen: The guidelines reflect our lack of data on whether one airway-clearance therapy is better than another, and I'm a bit frustrated by that, because we've been doing airway clearance techniques in CF for decades now, and the evidence we have comes from rather small studies. On many questions we've done large, well-powered studies that provided good evidence, but we haven't done that for airway clearance. Should we just give up on that, or, rather than saying, "One method is probably as good as the other, but we don't really know," should we try to get some more evidence?

Our physiotherapists are getting together to compare PEP and vest therapy, and to determine whether it's going to be feasible and whether you can power the study to find an outcome measure that really shows a difference. I still think that having stronger evidence than we have now would help, and maybe some of the more sensitive techniques, such as the lung-clearance index, will help us sort this out and give us better outcome measures. The current evidence is as you presented it, but I still feel a little uncomfortable about this, because in many areas we say we'll build our guidelines on the data, but lack the data for airway clearance.

Lester: It's an *n*-of-1 study sometimes. What works for one patient may not work for another. If vest therapy is the best way to do it, but the patient won't do vest therapy, then we have to use other therapies.

Flume: The guidelines committee did ask whether we should do a definitive study of airway clearance. The first question was to compare airway clearance to no airway clearance. As much as we all know the importance of airway clearance, the evidence to support it was only level B. The guidelines[1] state that, although we might want more robust data, the chance of a study being completed successfully is probably nil, because I don't think anybody would enroll a patient in such a controlled trial.

As for the studies of devices, the rigor of the approval studies for a device is far less than that for a medication, and that's why device studies have few subjects. We would like more rigorous evaluation of new devices, but the challenge is, what would we like to see measured? As Bruce said, what might seem to be the most intuitive outcome measure (sputum volume) may not be very useful.

REFERENCE

1. Flume PA, Robinson KA, O'Sullivan BP, Finder JD, Vender RL, Wiley-Courand DB, et al. Cystic fibrosis pulmonary guidelines: airway clearance therapies. Respir Care 2009;54(4):522-537.

Newton: Would you put the patient in the head-down position after vest therapy?

Geller: I'm not the right person to ask that question, you are. I asked that question because before the advent of vest therapy we had hands-on CPT followed by postural drainage. With vest therapy there's no postural drainage recommended, so the question is, did that do any good in addition to CPT, or did we just do it because it made sense that gravity might help? And maybe it did help in some people, maybe it didn't in others, but it was just the standard regimen: CPT and postural drainage. I'm not sure it would have an effect. I don't know that there's any evidence.

That said, if an individual patient is not improving with vest therapy and they don't have an effective cough, Pulmozyme's not going to work—nothing will work—you have to be able to cough it out. If they don't have an effective cough, they might jiggle the secretions around, but they won't get them out. So the question is, would postural drainage help? I don't know the answer.

Volsko: We haven't paired vest therapy with postural drainage, but we have paired postural drainage with oscillating PEP devices, such as Flutter or Acapella, and that seemed to help.

Mary, you brought up a really good point about thinking outside the box. We use the Acapella with a mask in very young patients who can't perform all the postural-drainage positions because of reflux, and that works nicely, as it does with patients at end of life, who have very low expiratory flow. We get a good cough and some expectoration from those patients as well.

Rubin: If I can address this: at the airway clearance journal conference a year ago[1,2] there was a full review about airway clearance physical therapy, and Cees van der Schans, who does the Cochrane reviews, looked at the role of postural drainage.[3] It was clear that from the studies that were done, which were few, there's no advantage with postural drainage at all, under any circumstances, other than the fact that it prolonged physical therapy. There are some risks for increased intracranial pressure in small babies, increased reflux, and discomfort in a head-down position. So in our center, for example, we do not use postural drainage.

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1. Airway clearance: physiology, pharmacology, techniques, and practice. Respir Care 2007;52(9):1070-1238.
2. Airway clearance: physiology, pharmacology, techniques, and practice. Respir Care 2007;52(10):1239-1405.
3. van der Schans CP. Bronchial mucus transport. Respir Care 2007;52(9):1150-1156; discussion 1156-1158.

McFall:* Do you find that patients have a big resistance to airway clearance, or is it just that they do not understand the reasons for this therapy? Or is it the time required? Patients don't necessarily realize that they're needing to do their therapy. If I don't take my enzymes, I notice within 12 hours, but patients adapt very well to feeling short of breath; they get used to feeling congested, and they don't necessarily make the connection that if they did their airway clearance, it would be beneficial. How do you address that?

Lester: It's different at different phases in life. With parents of a newly diagnosed and asymptomatic infant, it can be difficult to get them to buy in and do the therapies twice a day. With 5-to-8-year-olds who are starting to become symptomatic and have had their first *Pseudomonas*-positive culture, usually the family will buy in at that point. Teenagers don't want to do their therapies; they want to be inconspicuous and fit in with their peers, so therapies stop. When they become adults and the lung disease gets more severe, then they're on board, and usually very adherent to therapy. At our center we teach all the therapies by the time they're 12 years old. Life circumstances get in the way a lot of times.

Flume: Folks with CF are not different from the rest of us. Most of us aren't going to get on the treadmill today, even though we know it has health benefits; it's boring and it's hard to get into that routine.

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* Kristin McFall, Hill-Rom, St Paul, Minnesota.

Lester: And it'd be hard to fit in today. Every day is different. I don't think it's a lack of knowledge of the clearance techniques; it's just, "what am I going to do at this particular time?" That's why if you can incorporate sports or wind instruments into activities of daily living, the patient will get health benefits and be doing normal activities without necessarily realizing how much benefit they're getting, and they'll buy into it more.

Marshall: I'm always struck by how complicated it is for pediatricians with the developmental stages and dealing with all those little intricacies, as opposed to internists, for whom it's a little more straightforward. I'm curious about differences in what airway clearance techniques are prescribed and how exercise is approached in Europe and Canada versus the United States.

Davies: Something that struck me in a couple of the talks today is that in the United Kingdom I think we use fewer devices, we certainly use vest therapy very much less commonly than what you're describing. In my center, and at Great Ormond Street Hospital, where I've also worked, we very much more rely on the parents and the child to administer the straightforward techniques, as opposed to using the devices that you've mentioned. We have them available, and we use them in difficult cases, and we depend on our physiotherapy colleagues to advise us on what they think is best for the child, but in general I suspect that we use rather fewer devices, and certainly relatively fewer vests. Part of that might be cost. At my institution there are studies underway on whether vest therapy is as effective as other techniques.

Lester: Do you emphasize physical exercise?

Davies: We emphasize physical exercise, but not as a substitute for airway clearance. We try to promote that as a healthy lifestyle, but certainly in the pediatric clinic we're always saying, "That's not *instead* of your physiotherapy; that's *as well as*."

Ratjen: In Canada the primary form of physiotherapy is PEP, based on the Toronto study of PEP versus postural drainage.[1] It was a small study but it looked rather positive, and it used a relatively simple technique, so this is the primary form of physiotherapy. The cost of vest therapy is one of the major concerns, and there's a lot of interest in looking at that.

We helped our physiotherapy colleagues Maggie McIlwaine and Jennifer Agnew design a study to compare PEP to vest therapy in a cohort of CF patients, with pulmonary exacerbation as a primary outcome.[2] I think looking at lung function is probably not going to be very useful, and if I use the analogy from hypertonic saline, then I expect that pulmonary exacerbation is probably the outcome we have to look at. It's going to be a one-year study. The problem is that there is no placebo arm, and ideally you would like to do that study with a placebo arm, but I don't think that's feasible.

In various countries throughout the world there's a lot of belief in certain techniques. In Belgium, Chevalier introduced autogenic drainage and just about every patient is going to use that technique, and he goes from center to center teaching those techniques. The problem I have is that there's a lot of *belief* in the field, but a lack of data. We strongly encourage exercise and we have a long-term exercise study going on in Toronto. One interesting thing we see in the data from more than 6 years is a correlation between daily physical activity and lung-function decline in CF patients, across the whole population [unpublished data], so we strongly believe that exercise should be a focus. The only problem is that we have the ideal intervention to promote that as a long-term goal, because exercise protocols are difficult to follow, so you have to find ways to promote regular exercise; the data support that.

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