

Improving Outcomes in Amyotrophic Lateral Sclerosis: Selecting an Airway Clearance Modality

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Author Profile

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Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease that affects both the upper and lower motor neuron systems. Presently, ALS is of unknown etiology and is incurable. There are few effective treatments.¹ Between diagnosis and death, patients experience progressive muscle weakness, increasing functional disability including loss of the ability to speak, chew, swallow, and cough, inability to breath without mechanical assistance, and, eventually, complete motor paralysis.² The physical, emotional, social, and economic burdens of ALS are enormous; disease management demands sustained efforts by patients, families, caregivers, and healthcare professionals.³ Current average annual care costs are estimated at \$200,000 per patient.⁴ These burdens, however, may be mitigated by rational treatment and management decisions.

Early Intervention

To optimize patient outcomes, neurologists specializing in ALS advocate early, aggressive symptomatic treatment to preserve functional independence as long as possible.⁵ Because decline and death in ALS are coupled with the progressive failure of bulbar and respiratory muscles, treatment must focus upon maintenance of pulmonary health.^{6,7} A variety of causes may lead to disruption of normal airway clearance and impaired respiratory function, resulting in *increased* secretion production and *impaired* secretion clearance. Poor secretion clearance affects all components of normal mucus function. Consequent disruption of the pulmonary defense system increases vulnerability to serious, progressive, and ultimately life-threatening lung disease.⁸ Almost always, the immediate cause of death is respiratory failure,⁹ frequently associated with refractory respiratory infections.^{10,11}

Treatment Strategies

Airway clearance therapy is an essential component of any treatment regimen.^{12,13} Strategies to treat ALS include techniques to augment cough, to improve lung volumes, and to support the patient with progressive ventilatory failure.¹⁴ Clinicians recognize that individuals with significant secretion clearance problems require an aggressive regimen of bronchial hygiene including airway clearance therapy to slow the process of progressive lung disease, to preserve pulmonary function, and to avoid or reduce the need for more intensive therapy such as intravenous antibiotics and hospitalization.^{15,16,17} Anticipation and prevention of lung complications by means of effective airway clearance therapy can promote general health, delay the onset of pulmonary dysfunction, costly morbidities, and premature mortality. Quality of life of patient and caregiver is improved.

Selecting an Airway Clearance Modality for ALS

Mobilization of secretions can be accomplished by a variety of techniques and devices. ALS patients, however, do not have the energy or lung capacity for techniques that depend on forced expiration, such as the Flutter® device, active cycle of breathing (ACT) or positive end-expiratory pressure (PEP) masks. Thus, options are limited to percussion and postural drainage (P&PD), aka chest physiotherapy (CPT), and high-frequency chest wall oscillation (HFCWO), administered via The Vest™ Airway Clearance System. For patients with the disabilities associated with ALS, a comparison of the two methods demonstrates that The Vest™ system therapy offers significant, clear advantages over CPT.

Chest physiotherapy

Traditionally, the removal of mucus from the lungs is accomplished by the application of CPT. CPT is an airway clearance technique that combines manual percussion of the chest wall by a caregiver, strategic positioning of the patient for mucus drainage, and cough and breathing techniques. Typically, a treatment session consists of manual percussion for 3-5 minutes on each of nine specific thoracic regions while assuming appropriate drainage postures. Forced expiratory maneuvers are performed between percussion periods. The technique is based on the theory that percussion to various areas of the chest and back transmits shock waves through the chest wall, which loosen secretions in the airways.¹⁸

Although CPT is established in the peer-reviewed literature as an effective method for mobilizing mucus,¹⁹ its benefits are compromised by a number of factors.^{20,21,22,23} CPT is:

- technique-dependent...*effective treatment depends upon the skill and dedication of a trained caregiver*²⁴
- labor-intensive...*caregivers must possess the physical*

*strength and endurance both to administer effective chest percussions and to position patients on a slant board to facilitate gravitational movement and expectoration of mucus*²⁵

- time-consuming...*typical treatment sessions last 30-45 minutes and may be required three or more times daily*²⁶

CPT requires:

- mental and physical cooperation from the patient...*patients who are mentally impaired, physically uncooperative, paralyzed, or severely spastic are difficult to treat efficiently and effectively*²⁷
- patient ability to tolerate treatment...*contraindications arising from anatomical deformity, transient hypoxemia associated with postural drainage, predisposition to gastroesophageal reflux, aspiration, inability to perform breathing techniques, etc., preclude the use of CPT*²⁸
- treatment adherence... *various studies indicate that, in general, CPT has the lowest adherence rate of any component of bronchial hygiene care*^{29,30,31,32,33}

To overcome obstacles that limit effectiveness of CPT for ALS patients, physicians are challenged to identify an alternative that is:

- equal to or more effective than CPT
- effective despite physical problems including:
 - predisposition to aspiration and gastroesophageal reflux
 - physical paralysis; inability to assume therapeutic postures
 - inability to tolerate head-down position
 - inability to perform cough or breathing techniques
 - physical attachment to ventilatory devices, catheters, IV lines, and other encumbrances
- responsive to patient needs and preferences
 - ALS patients may prefer to forego treatments burdensome to their caregivers
 - depressed ALS patients may be unwilling to accept physically demanding therapy
 - responsive to caregiver issues
 - primary caregivers are typically severely overburdened family members; they may lack the energy or skill to administer labor-intensive, time consuming CPT reliably.
 - ALS caregivers are frequently older spouses; they may lack the strength, endurance, or physical ability to percuss the chest effectively or to position an immobile, technology-dependent patient on a slant board
 - responsive to cost issues

- unreliable airway clearance therapy is not cost-effective; medical expenses, premature physical deterioration, and quality of life losses associated with ineffective respiratory health care are incalculable.
- the cost of professionally administered CPT may be prohibitive; average ALS patients survive 2-5 years.

The Vest™ system: a practical approach to secretion clearance in ALS

High-frequency chest wall oscillation (HFCWO)

HFCWO technology works on the principle that rapid compression and relaxation (oscillations) of the chest wall generates increased airflow velocities, thus creating brief changes in lung airflow patterns similar to coughing. The percussive effects of chest wall oscillation also thin viscous secretions, making them easier to clear.^{34,35}

The Vest™ system administers effective HFCWO therapy by means of an inflatable vest attached by hoses to an air pulse generator. The vest inflates and deflates rapidly, gently compressing and releasing the chest wall 5-20 times per second. The oscillations can be adjusted to different frequencies and pressures. During The Vest™ system therapy, oscillating compressive forces are distributed evenly over the thorax surrounding the area of the lungs. This extensive contact area results in a large total oscillating force applied to the thorax. The Vest™ system does not require positioning or postural drainage; it is not technique-dependent; and it can be administered without a caregiver or with minimal caregiver supervision.

The Vest™ system:

- received FDA clearance in 1988
- is prescribed as an alternative to CPT
- is used in conformity to *Clinical Practice Guidelines for Postural Drainage* established by the American Association for Respiratory Care (AARC)³⁶
- has been prescribed for more than 40,000 individuals
- is associated with cost savings^{37,38}
- is associated with increased treatment adherence^{39,40}
- is associated with quality of life gains⁴¹
- is supported by evidence from more than 70 clinical trials conducted at over 69 clinical centers

The Vest™ system: An evidence-based airway clearance alternative

The primary short-term goal of airway clearance therapy is to remove pulmonary secretions effectively and efficiently. A secondary goal is to achieve improvement in pulmonary health and function. Clinical trials of The Vest™ system and HFCWO published in the peer-reviewed

literature demonstrate its equivalence or superiority to CPT for outcomes including volume of secretions cleared and PFT scores:

- Scherer (1998): Using HFCWO and two forms of CPT, stable CF patients achieved *equal but significantly enhanced secretion expectoration with all three methods*⁴²
- Kluft (1996): *Significantly more mucus was cleared using HFCWO* in contrast to CPT in a study including stable CF patients⁴³
- Braggion (1995): In a study comparing CPT, PEP, and HFCWO in CF patients, *all three methods resulted in equal but significantly increased sputum clearance*⁴⁴
- Arens (1994): *Equal but important improvements in pulmonary function and clinical status* were achieved in acutely ill CF patients receiving *either CPT or HFCWO*⁴⁵
- Warwick (1991): In a long term retrospective-prospective study comparing CPT and HFCWO in CF patients, *patients receiving HFCWO obtained impressive stability in PFT scores and, in many cases, significant clinical improvement*⁴⁶
- Hansen (1990): *Significantly more mucus was cleared using HFCWO* in contrast to CPT in a study including CF patients⁴⁷

Additional evidence published as *Abstracts* and in professional journals reinforces peer-reviewed studies.
48,49,50,51,52,53,54

In clinical trials testing the safety and efficacy of HFCWO as an airway clearance modality, *HFCWO consistently performed as well as or significantly better than comparison methods*. Moreover, in those studies comparing HFCWO with CPT, the latter was administered with a standard of rigor not practiced consistently in the outpatient setting. Thus, research results may overstate the efficacy of CPT.

Improving outcomes for individuals with ALS

The clinical course in ALS varies markedly from case to case. However, regardless of the pace and severity of symptoms, patients with the “best” outcomes are those who, together with their physicians and families, find ways to continue to live as fully as possible. The key to optimal ALS care includes thorough patient/family education, early therapeutic interventions, and rational advance planning.⁵⁵ Because many respiratory complications associated with ALS are *preventable or treatable*, therapy must center upon preservation of pulmonary health.⁵⁶ The justification

for focused and aggressive management of pulmonary disease in ALS is the prevention of the chronic respiratory illness that results in both increased morbidity and cost of medical care. Stabilization of respiratory function can result in an improved quality of life.^{57,58} Healthcare economists emphasize correlation between early, sustained supportive care and cost containment.^{59,60} Effective airway clearance therapy is associated with:

- decreased morbidity and mortality
- increased clinical stability
- reduced incidence of hospitalization
- reduced auxiliary medical care costs
- enhanced quality of life

The Vest™ Airway Clearance System is established as a safe, effective, and efficient airway clearance modality. Because the method has few or none of the disadvantages and limitations associated with CPT, it is an ideal modality for the treatment of individuals with ALS.

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