

# High-Frequency Chest Wall Oscillation for Airway Clearance in Neuromuscular Dysfunction

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Respiratory complications are a common consequence of neuromuscular dysfunction in inherited and acquired neuromuscular diseases (NMD) affecting motor neurons.<sup>1, 2</sup> These diseases include muscular dystrophies, myotonias, spinal muscular atrophies, amyotrophic lateral sclerosis (ALS), multiple sclerosis, Guillain Barré syndrome, and poliomyelitis. Though not an NMD, cerebral palsy (CP) is a functional neurologic disorder in which respiratory complications arise as a consequence of neuromuscular dysfunction.<sup>3</sup> Neuromuscular dysfunction in CP and NMD affects swallowing and respiratory muscles. Most people with these disorders develop restrictive lung disease. Spinal deformity is also common, which results in reduced chest wall compliance.<sup>4</sup> These people often require daily airway clearance therapy to prevent recurrent respiratory infections that can precipitate atelectasis and respiratory failure. Ineffective cough coupled with aspiration of food particles, saliva and other liquid as a result of dysphagia not only disrupt normal airway clearance but also impair respiratory function and precipitate lung damage,<sup>5</sup> which in turn results in increased secretion production and impaired secretion clearance. Ultimately, respiratory insufficiency is a leading cause of morbidity and mortality in people with severe neuromuscular dysfunction.<sup>1, 2, 6, 7</sup>

In this population, airway clearance problems resulting from impaired or ineffective cough are exacerbated by impaired mobility and subsequent lack of exercise. Under normal circumstances, physical exercise mobilizes lung secretions by increasing airflow throughout the lungs and by activating processes of the endocrine and parasympathetic nervous systems that have important effects on volume and viscosity of secretions.<sup>8, 9</sup> Conversely, immobility can lead to pooled secretions in the lungs,<sup>10</sup> which provide a fertile breeding ground for bacteria, and contribute to a host of pulmonary complications resulting from recurrent lung infection. Atelectasis is a frequent cause of respiratory failure in people with neuromuscular dysfunction,<sup>11</sup> and mucus plugging as a result of insufficient airway clearance is the most common cause of atelectasis in children with neuromuscular disabilities.<sup>5</sup>

Physical limitations of neuromuscular dysfunction pose significant challenges for administering traditional airway clearance therapies. High-frequency chest wall oscillation (HFCWO) delivered via The Vest<sup>®</sup> Airway Clearance System is easier to administer and may be better tolerated in patients with neuromuscular dysfunction. Early studies suggest that The Vest<sup>®</sup> System therapy is effective in mobilizing secretions and, as a result, may help prevent pulmonary complications associated with retained secretions (see Table on page 2).

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## Airway Clearance in Neuromuscular Dysfunction

The feasibility and effectiveness of standard airway clearance modalities are often limited by manifestations of neuromuscular dysfunction. Conventional chest physiotherapy (CPT), consisting of postural drainage, percussion, and vibration, is a long-established standard for airway clearance in cystic

fibrosis (CF) and other disorders where removal of tracheobronchial secretions is impaired.<sup>12</sup> CPT is often supplemented with manually assisted cough techniques intended to assist with removal of secretions mobilized during CPT. The physical requirements of these modalities, however, limit their use in people with neuromuscular dysfunction. The positioning required for CPT is often not feasible for

people with neuromuscular dysfunction due to spinal deformity, presence of a feeding tube, and gastroesophageal reflux (GER), among other factors. Similarly, the effectiveness of manually assisted cough may be diminished by deformity of the thoracic cage or spine,<sup>12</sup> distended abdomen or full stomach, and obesity, all of which occur regularly in patients with neuromuscular dysfunction. Furthermore, people with neuromuscular dysfunction often are not able to generate the coordinated expiration and expiratory pressures required for these therapies to be effective.

A number of alternative therapies are used in place of, or to supplement, CPT and assisted cough. These include forced expiratory technique (huffing) to increase sputum clearance, autogenic drainage, expiratory muscle training to improve peak expiratory pressure, positive expiratory pressure (PEP) devices and oscillatory PEP devices. However, these therapies, too, require coordinated effort that is typically not feasible for patients with more severe neuromuscular dysfunction.<sup>12</sup>

Mechanically assisted cough provided by maximal insufflation-exsufflation devices can be a useful therapy to clear pulmonary secretions that have been mobilized. This therapy can be successfully utilized in

patients without an effective cough. However, many of these patients also require therapy to promote secretion mobilization as mechanical cough assistance alone does not provide optimal treatment.

HFCWO therapy with The Vest<sup>®</sup> System offers people with neuromuscular dysfunction an airway clearance therapy that does not require positioning, postural drainage, or coordinated expiration, and can be administered with minimal caregiver support. The Vest<sup>®</sup> System, consisting of an inflatable garment attached by hoses to an air pulse generator, produces rapid, gentle compressions and relaxations (oscillations) of the chest wall. These oscillations generate increased airflow velocities, in turn creating brief changes in lung airflow patterns similar to coughing. Research has shown HFCWO therapy to be at least as effective as CPT for secretion clearance in patients with CF,<sup>13-18</sup> as well as an effective means to mitigate decline in pulmonary function.<sup>13, 18</sup> Although the pathologies leading to airway clearance problems in neuromuscular dysfunction differ from those of CF, the need for airway clearance assistance to mobilize and clear secretions is common to both conditions.

### Research in CP and Various NMDs

As described above, inadequate clearance of airway

#### Studies Evaluating Vest Therapy (VT) in Neuromuscular Dysfunction

<i>Diagnosis</i>	<i>N</i>	<i>Mean Age (y)</i>	<i>Treatments</i>	<i>Results</i>
Quadriplegic CP <sup>21</sup>	7	19	VT	Significant decrease in pneumonias and increase in effective suctioning from year prior to VT.
CP and NMD <sup>22</sup>	44	15	VT (home use)	Mean hospitalization days/year significantly reduced from year prior to VT.
CP and NMD <sup>23,24</sup>	25	VT: 13 CPT: 15	VT (n=11) CPT (n=14)	Significantly fewer hospitalizations for respiratory complications in VT vs CPT group. Adherence to prescribed regimen significantly higher in VT group.
ALS <sup>25</sup> (FVC ≥40% predicted)	46	58	VT (n=22) Untreated (n=24)	Significantly less breathlessness and more night-time coughing in VT-treated vs untreated patients.
ALS <sup>26</sup> (FVC 40-47% predicted)	9	VT: 64 Standard Care: 53.5	VT (n=5) Standard Care (n=4)	VT did not slow decline of FVC or increase survival time (11-15 mo) compared with non-invasive ventilatory support/BiPAP alone.
FD <sup>28</sup>	15	19.5	VT	Statistically significant improvements in oxygen saturation, FVC, and PEF rate from baseline to 12-mo follow-up. Significantly fewer pneumonias, hospitalizations, antibiotic courses, antibiotic days, doctor visits, and absentee days compared with year prior to VT.

CP=cerebral palsy; NMD=neuromuscular disorder; CPT=chest physiotherapy; ALS=amyotrophic lateral sclerosis; FD=familial dysautonomia; FVC=forced vital capacity; PEF=peak expiratory flow; Standard Care=non-invasive ventilatory support with BiPAP

secretions and recurrent pneumonias are consequences of neuromuscular dysfunction in children with CP and NMDs. In addition to dysphagia, people with CP or neuromuscular weakness disorders are also prone to aspiration resulting from high incidences of GER<sup>19</sup> and seizures<sup>20</sup> in this population. Three studies evaluating The Vest<sup>®</sup> System therapy in CP and varied NMDs are summarized below.

In a case series of 7 patients with quadriplegic CP and frequent pulmonary infections, investigators compared pulmonary function measures in the 12 month-period prior to initiating HFCWO therapy with the first 12 months using HFCWO therapy.<sup>21</sup> Mean patient age was 19 years (range 7-28 years); 5 patients had a tracheostomy. Episodes of pneumonia decreased from 36 per year prior to therapy to 18 per year during HFCWO therapy (P=0.03). Similarly, hospitalizations for pneumonia decreased from 9 per year to 3 per year, though this difference was not statistically significant (P=0.16). Frequency of effective suctioning of secretions increased in all patients, with a sum total of 4825 before treatment and 10,455 after treatment (P=0.008). There were no significant adverse events related to the therapy; in particular induced emesis, fractures, worsening seizures or GER were not observed. This initial foray into the use of HFCWO therapy for airway clearance in CP patients provided a rationale for larger, longer-term studies on the effects of the therapy for airway clearance in CP and NMD.

Home use of The Vest<sup>®</sup> System in pediatric patients with non-CF diagnoses at risk for pneumonia/atelectasis was studied in a retrospective chart review of 44 patients (mean age: 15±8 years) over a 7-year period.<sup>22</sup> Diagnoses included muscular dystrophy (n=20), spinal muscular atrophy (n=10), CP (n=8), bronchopulmonary dysplasia (n=2), and other NMD (n=4). The Vest<sup>®</sup> System was prescribed for atelectasis in 12 patients, pneumonia in 15 patients, and preventive use in 17 patients. During a mean 37.8±15.5 months of follow-up, there were no reports of pneumothorax, aspirations, hemoptysis or rib fracture. Based on compliance data (6±2 weeks), mean daily therapy was 23±8.7 minutes, with 65% of patients averaging a minimum of 15 minutes per day. Mean days of hospitalization per year were reduced from 10.5 days in the one-year period before initiating HFCWO to 4.1 days in the first year of therapy use (P<0.01).

In a small, randomized, controlled trial of 25 patients (mean age: 14 years), investigators compared The Vest<sup>®</sup> System with standard CPT in pediatric patients with CP (n=9) or NMDs, including Duchenne and other muscular dystrophies (n=8), mitochondrial or other myopathies (n=5), and other NMDs (n=3).<sup>23</sup> The mean treatment durations of 4.5 months in the HFCWO group and 5.3 months in the CPT group were not statistically different. Significantly fewer hospitalizations for respiratory complications occurred in the HFCWO group (0/11) compared with the CPT group (5/14) (P=0.046). Fewer antibiotic courses were required in the HFCWO group (total: 7, pneumonia: 0) compared with the CPT group (total: 10, pneumonia: 3), although these differences did not reach statistical significance. No differences were observed between groups on chest radiographs (modified Brasfield score), maximum inspiratory and expiratory pressures, and polysomnography. These investigators also assessed adherence to therapy.<sup>24</sup> Patients and caregivers were instructed to perform HFCWO therapy or CPT thrice daily for 12 minutes over a 3-month period. At study end, patients and caregivers were asked how many times per day they performed the assigned therapy, for how many minutes per session, and how many days per week. Adherence to the prescribed regimen was higher in patients using The Vest<sup>®</sup> System (283±142 minutes/week) compared with the CPT group (86±98 minutes/week) (P<0.001). Taken together, these findings suggest that HFCWO with The Vest<sup>®</sup> System may be both more effective and easier to adhere to than CPT for pediatric patients with CP or NMD and their caregivers.

### Research in ALS

The relentless progression of neuromuscular weakness in ALS results in respiratory insufficiency and ultimately respiratory failure, the most common cause of death in ALS patients, typically occurring within 2 to 5 years of diagnosis.<sup>6</sup> Though lung disease is not the primary problem in ALS, respiratory muscle weakness, ineffective cough, bulbar dysfunction, recurrent aspiration, and progressive immobility make these patients highly susceptible to pulmonary infection, and frequent pneumonias are common. Noninvasive positive pressure ventilation (NIPPV) is the current standard of care in ALS patients with forced vital capacity (FVC) ≤ 50% of predicted.<sup>6</sup> This helps to delay respiratory failure, but most patients still develop problems clearing respiratory secretions. Two

studies have explored the use of The Vest<sup>®</sup> Airway Clearance System in ALS patients.

A randomized, controlled trial of HFCWO with The Vest<sup>®</sup> System was conducted in 46 ALS patients (mean age 58 years) with ALS Functional Rating Scale respiratory subscale scores  $\geq 5$  and  $\leq 11$  and FVC  $\geq 40\%$  predicted.<sup>25</sup> Changes from baseline to 12-week follow-up were compared between patients treated with HFCWO and untreated control patients. Although the study was underpowered to detect differences between treatment groups for pulmonary complications, hospitalizations, and mortality, patients using HFCWO therapy reported significantly less breathlessness ( $P=0.02$ ) and more night-time coughing ( $+0.67$  vs  $-0.65$ ,  $P=0.048$ ) at 12 weeks than untreated patients. HFCWO patients also reported a significant decline in breathlessness from baseline to 12 weeks ( $-1.28$ ,  $P=0.048$ ), whereas untreated patients reported an increase in breathlessness ( $+0.84$ ,  $P=0.20$ ). No significant differences were observed between groups for changes in FVC, peak expiratory flow (PEF), capnography, oxygen saturation, fatigue, or transitional dyspnea index. A secondary analysis of 21 patients with FVC 40% to 70% predicted was completed. It focused on patients with moderate pulmonary dysfunction, excluding those with minimal pulmonary dysfunction. In these patients with mean baseline FVC of 54% predicted in control and 59% predicted in the HFCWO therapy group, there was a significant mean decrease in FVC in the control group but not in HFCWO treated patients. Additionally, while the changes in capnography were not significant between the groups, the changes were in opposite directions, increasing in the control group and decreasing in HFCWO group. This analysis suggests that The Vest<sup>®</sup> System therapy may be of benefit for moderate but not minimal ALS-associated respiratory dysfunction.

There has also been one small randomized trial comparing HFCWO coupled with non-invasive ventilatory support/BiPAP versus ventilatory support/BiPAP alone in 9 ALS patients with mean FVC 40% to 47% predicted.<sup>26</sup> In this study, the therapy did not increase survival time compared with non-invasive ventilatory support/BiPAP alone, and in contrast to the subgroup analysis from the aforementioned study, HFCWO therapy did not slow the decline of FVC. These findings should be

considered in light of the severity of ALS in this population with a mean survival of only 11 to 15 months and the very small sample size that was likely insufficient to identify a therapeutic difference. The rapid and relentlessly progressive nature of ALS poses unique challenges for evaluating respiratory therapies and makes it unlikely that airway clearance will make a difference in survival outcomes in this population.

### **Research in Familial Dysautonomia**

Familial dysautonomia (FD) is primarily a neurologic disorder affecting autonomic and sensory functions. FD patients are susceptible to secretion retention and recurrent pneumonia as a consequence of numerous clinical manifestations of their disease, including uncoordinated swallowing, esophageal dysmotility, depressed cough and gag reflexes, hypotonia, poorly coordinated breathing, and spinal curvature.<sup>27</sup> Recurrent lung disease in these patients is a result of aspiration resulting from misdirected swallows or GER and ineffective cough due to hypotonia.<sup>27</sup> Traditional CPT, consisting of postural drainage and inhaled bronchodilators, is not always feasible and is often poorly adhered to in this population for several reasons. The Trendelenburg position required for postural drainage is not well tolerated due to postural hypotension without compensatory tachycardia, supine hypertension, and spinal curvature when present. Additionally, for FD patients without a fully functional fundoplication, the head-down position can exacerbate GER thus leading to esophagitis and risk of aspiration.

A prospective clinical study of HFCWO therapy with The Vest<sup>®</sup> System in 15 FD patients aged 11 to 33 years looked at pulmonary function measures from baseline to 12-month follow-up and compared health outcomes between the 12-month period prior to starting The Vest<sup>®</sup> System therapy and the first 12-months of using the therapy.<sup>28</sup> Sustained improvements from baseline to follow-up were observed for all pulmonary function measures. These improvements were statistically significant for oxygen saturation (increased from 94% to 98%,  $P=0.004$ ), FVC (77% improvement,  $P=0.02$ ), and PEF rate (73% improvement,  $P=0.03$ ). All measured health outcomes improved significantly from the period preceding HFCWO therapy to the HFCWO therapy period: pneumonias (3 vs 1,  $P=0.02$ ), hospitalizations (1 vs 0,  $P=0.02$ ), antibiotic courses (8 vs 4,  $P=0.0005$ ), antibiotic days (85 vs 34,  $P=0.0002$ ), doctor visits (11

vs 5, P=0.0005), and absentee days (32 vs 9, P=0.0002).

### Summary

There are currently several airway clearance modalities for mobilizing secretions and assisting cough, though most are not supported by rigorous long-term clinical studies.<sup>2, 12</sup> Furthermore, the most well-established airway clearance therapies (CPT and manual-assisted cough) are often not physically feasible in people with neuromuscular weakness. Thus, there exists a clear need to identify airway clearance therapies tailored to the specific challenges of patients with neuromuscular dysfunction. The optimal therapeutic regimen for this population may well require modalities for both mobilizing secretions (i.e. oscillatory devices) and mechanically assisting cough.

The advantage of The Vest<sup>®</sup> System in neuromuscular weakness disorders is that it offers consistent, effective airway clearance that does not require positioning or coordination that is challenging or unfeasible for people with neuromuscular dysfunction. Furthermore, while caregiver supervision is often required in these populations, the physical burden is significantly less than that required for manual therapy. In addition, the ability to deliver therapy in any position increases tolerance and ameliorates some of the risks associated with positioning required for standard CPT.

Early research on the feasibility, efficacy, and safety of The Vest<sup>®</sup> Airway Clearance System in people with neuromuscular dysfunction requiring airway clearance therapy indicates that this high-frequency chest wall oscillation system may provide valuable airway clearance benefits for patients with respiratory problems associated with neuromuscular dysfunction.

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