

# **Muscular Dystrophy and Congenital Myopathy with Respiratory Insufficiency and Pneumonia: A Case Study**

**Tom Hall RN, RCP**

**Employed by Hill-Rom**

## **Author Profile**

Tom Hall, as Account Manager for Hill-Rom, has contributed to a variety of research endeavors and developed white papers, case studies and other supporting materials.

# Case Study:

## Muscular Dystrophy and Congenital Myopathy with Respiratory Insufficiency and Pneumonia

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Pulmonary complications comprise the number one cause of death in patients with Duchenne's muscular dystrophy (DMD) and other severe myopathies.<sup>1,2</sup> The high incidence of such sequelae likely stems from compromised pulmonary function secondary to progressive weakness of the respiratory muscles, a predisposition toward atelectasis and dysphagic aspiration, and impaired secretion clearance due to ineffective cough.

In order to prevent or delay the onset of life-threatening pulmonary complications, people with muscular dystrophy and other myopathies frequently require daily airway clearance therapy.<sup>3,4</sup> For over 40 years, chest physical therapy (CPT) has been the standard modality for airway clearance. In recent years, however, a number of other methods of effective secretion mobilization have been developed. Whether the use of any of these alternate therapeutic modalities might enhance the pulmonary health and quality of life among patients with muscular dystrophy or other myopathies is a question important to clinicians, case managers, and patients alike.

The following is an account of a critically ill child with muscular dystrophy of uncertain type, congenital myopathy, and pneumonia. During acute hospitalization, standard CPT treatments yielded little improvement. Seeking a more effective means to mobilize her secretions and help resolve the pneumonia, her pulmonologist prescribed high-frequency chest wall oscillation via a device called The Vest™ Airway Clearance System.

The Vest™ system, manufactured by Hill-Rom, consists of an air-pulse generator attached to an inflatable vest that covers the entire thorax. When the generator is turned on, the pressure in the vest increases and decreases between five and 25 times per second, thereby applying rapid oscillations to the patient's chest wall. This procedure is called high-frequency chest wall oscillation, or HFCWO. HFCWO has been shown to produce transient increases in airflow at low lung volumes, cough-like shear forces, alterations in the physical properties of mucus, and consequent increases in mucus mobilization. Because The Vest™ system mechanism of action is not reliant on gravity, The Vest™ system provides effective airway clearance independent of postural drainage.

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### Case Summary

A 15-month old Hispanic female was brought to the emergency room in severe respiratory distress. At the age of six months, she had been diagnosed with muscular dystrophy and congenital myopathy. Diagnosis was based on muscle biopsies, neuromuscular weakness, respiratory insufficiency and a consequent inability to clear secretions. A tracheostomy was placed two months prior to this admission and the patient was being maintained on nocturnal ventilation.

In the emergency room, the child exhibited increased work of breathing (+1 to +2 sternal retractions), a low grade temperature (99.9°F), thick yellow-green secretions, tachypnea of 50-60 breaths per minute, and tachycardia of 200 beats per minute. A chest x-ray showed increased density on the left and atelectasis in the left-lower lobe. The patient was admitted to the pediatric ward with a diagnosis of tracheitis vs. pneumonitis. In order to maintain SaO<sub>2</sub> at or above 95%, she was placed on 2-6 lpm of oxygen. Antibiotic therapy was initiated using gentamycin and ceftazidime along with 2.5 mg nebulized albuterol every four hours.

Two days later the patient's condition worsened, and she was transferred to intensive care. Airway patency was maintained through frequent suctioning. Secretions remained yellow-green. A chest x-ray revealed diffuse opacification bilaterally with decreased volume in the right-lower lobe and obliteration of the right cardiac border. Conventional CPT was added to the treatment regimen.

The next day the patient continued to exhibit tachycardia and tachypnea with a low-grade fever. The chest x-ray showed bilateral pulmonary infiltrates with bilateral consolidation or atelectasis. A capillary blood gas on 5 lpm via tracheostomy mask showed a pH of 7.41, paCO<sub>2</sub> of 50 mm Hg and paO<sub>2</sub> of 60 mm Hg.

In an effort to clear the patient's secretions more effectively, CPT was replaced by The Vest™ therapy.

### Method

The patient received The Vest™ system treatments every four hours at a pressure setting of 5.5 and a frequency of 12 Hz. Initially, she tolerated the treatments without problem for eight minutes; treatment time was gradually

increased to ten minutes with an eventual goal of 15 minutes. Although The Vest™ system treatments often last 20-30 minutes, in this case, because the patient was receiving six treatments with The Vest™ system throughout the day, abbreviated sessions were not considered to diminish therapeutic effectiveness.

## Results

A few days later, the chest x-ray revealed persistent loss of volume in the right-lower lobe and a retro-cardiac shadow on the left.

The next day the patient's temperature was stable around 98.6° F and her secretions were thinning and clearing.

Within 48 hours of initiating The Vest™ therapy, chest x-rays showed marked improvement, with resolution of the consolidation in the right-lower lobe, improved aeration and reduced infiltrates bilaterally. Secretions had resolved from green-yellow to white-clear, and acetylcysteine, a mucolytic agent, was added to the nebulized therapy.

A serious desaturation event occurred during the patient's bath. Her tracheostomy was found to be plugged with mucus secretions and was changed. She recovered quickly and blood gas levels returned to baseline. Throughout the incident, the child remained active and alert. Her white blood cell count had increased to a level of 20.3. This leucocytosis was believed to be a physiologic stress response to the desaturation event. In a discussion of this event with the respiratory therapist and pediatric pulmonologist, the possibility was raised that the use of bubble-humidified oxygen via a trach-t may well have dried the sol layer of mucus, leading to increased mucus viscosity and plugging of the tracheostomy tube. The patient was switched to a heated aerosol at .28-.35 FIO<sub>2</sub> and subsequently maintained a saturation above 95%. No further plugging occurred.

The patient was discharged to her group home to continue on nebulized therapy with beta-agonists, heated aerosol with FIO<sub>2</sub> up to .35 as necessary to maintain SaO<sub>2</sub> greater than 95%. She was continued on nocturnal ventilation for 4-8 hours per night and HFCWO with The Vest™ system as often as every four hours to ensure adequate airway clearance.

During the nine month interval following discharge from the hospital, this patient has been weaned completely from nocturnal mechanical ventilation. She has experienced a single episode of tracheitis (p. aeuriginosa was cultured from the stoma) which was treated with

antibiotics. Presently, she requires only low flow oxygen therapy at night. She has required no subsequent hospitalizations. During the day, she is an active child and frequently is able to travel outside the group home without oxygen support. She has been weaned to a smaller tracheostomy and will be evaluated for its complete removal in the spring. Although she still has a g-tube in place, she is able to take both food and liquids by mouth without evidence of dysphagia or aspiration. Interestingly, this child has demonstrated increasing muscular strength and coordination, leading to the speculation that, as a neuromuscular disease patient, her early history of ventilatory insufficiency and subsequent respiratory failure may have been exacerbated by concomitant infection with a Guillain-Barré-like syndrome.

Since the resolution of her acute illness, this child has made impressive developmental progress. Now almost two years old, she is bright, alert, and physically active. Speech therapy is planned for the near future to correct the deficit resulting from tracheostomy. Her mother and grandmother have begun to visit more regularly and are discussing the possibility of caring for her at home. During daytime hours, nebulized therapy and HFCWO have been continued every four hours, and she has experienced no further respiratory infections or complications. For the past several months, her oxygen saturation levels have remained above 95% on room air. The patient has suffered no adverse effects associated with The Vest™ therapy, and, in the judgment of both her physicians and caregivers, HFCWO therapy has been an essential component in the maintenance of this child's respiratory health.

## Discussion

Patients with neuromuscular diseases including muscular dystrophy typically suffer from a host of pulmonary complications secondary to neuromuscular weakness. Common among such complications are restrictive lung disease, chronic atelectasis, dysphagia, aspiration, gastroesophageal reflux, and the inability to generate a cough force sufficient to clear secretions, all of which result in secretion retention and progressive damage to the mucociliary clearance system. Consequences include recurrent bouts with pneumonia, bacterial colonization and, finally, respiratory failure. In patients with progressive neuromuscular disease, aggressive bronchial hygiene, including daily airway clearance therapy, is recognized as an essential component in managing and minimizing the adverse effects of secretion retention. For such patients, effective airway clearance therapy plays a significant role in reducing medical care costs, morbidity and mortality, and in enhancing quality of life.

Because this was not a controlled study, it is difficult to assess quantitatively the contribution of HFCWO to this patient's rapid recovery and subsequent good health following initiation of The Vest™ therapy. Certainly, antibiotics and broncho-dilators were important factors in the resolution of the pneumonia. Still, the progressive improvement experienced in this critically ill patient following the addition of The Vest™ therapy to her regimen suggests that further research into the therapeutic impact of HFCWO treatment for muscular dystrophy and other neuromuscular disease patients is warranted. Under normal circumstances, The Vest™ system is not prescribed for use in children under the age of two. In the present case, the child was critically ill and no alternatives were available. The Vest™ therapy was tried as a desperate measure. The successful outcome suggests that, additionally, the usefulness of The Vest™ therapy in 1) patients under two years of age and 2) those with tracheostomies in place should be studied.

## References

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<sup>2</sup> Smith PEM, Edwards RHT, Evans GA, Campbell EJM. Practical problems in the respiratory care of patients with muscular dystrophy. *New Engl J Med* 1987; 316: 1197-1205.

<sup>3</sup> Kim WD. Lung mucus: A clinician's view. *Eur Respir J* 1997; 10: 1914-1917.

<sup>4</sup> Hardy KA, Anderson BD. Noninvasive clearance of airway secretions. *Respir Care Clin N Am* 1996; 2 (2): 323-345.