

Editorial Response to The Article:

High Frequency Chest Wall Oscillation for Individuals With Chronic Neuromuscular Weakness

Louie Boitano MSc, RRT
Pulmonary Clinic, Northwest Assisted Breathing Center
Department of Respiratory Care
University of Washington Medical Center
Seattle, WA

The administration of an airway clearance modality should be based upon the understanding of the disease pathophysiology and how the particular therapy can benefit the patient. The author has indicated that the use of high frequency chest wall oscillation (HFCWO) can be beneficial to the respiratory health of neuromuscular patients with respiratory insufficiency by maintaining airway clearance and reducing the potential for morbidity and mortality associated with respiratory infection. Airway clearance therapy includes a variety of therapies that mobilize airway secretions, cough augmentation therapies that clear secretions from the airways and airway suctioning to clear secretions from artificial airways. Intrinsic lung diseases that affect airway muco-ciliary clearance function can result in pulmonary congestion and chronic respiratory infection. Secretion mobilization therapies can be very beneficial in augmenting the clearance of secretions in airway diseases including cystic fibrosis, bronchiectasis, ciliary dyskinesia, and other hyper-secretive pathologies where normal muco-ciliary clearance is encumbered. HFCWO has been found to be at least as good as other secretion mobilization therapies. Patients with intrinsic lung disease that affects normal muco-ciliary clearance generally do not have insufficient cough strength to clear secretions and therefore cough augmentation therapy would not be indicated.

The pathophysiology of neuromuscular induced restrictive lung disease is quite different from that of intrinsic lung diseases. The signs and symptoms of neuromuscular respiratory insufficiency are chronic, progressive hypoventilation and associated lower lung field atelectasis, and insufficient cough strength secondary to diaphragmatic, chest wall and abdominal wall muscle weakness. Neuromuscular patients with respiratory insufficiency generally have no intrinsic pulmonary co-morbidity that affects airway muco-ciliary function. The limiting factor in the clearance of their pulmonary secretions is cough strength. Without adequate cough strength to clear secretions, neuromuscular patients are susceptible to pulmonary congestion. When cough strength is insufficient (peak

cough flow <160 L/min) cough augmentation therapy can support adequate cough clearance to maintain pulmonary health¹. Mechanical in-exsufflation has been found to be the most effective means of cough augmentation therapy for this patient population²⁻⁴. While secretion mobilization therapy has been commonly prescribed as maintenance therapy for neuromuscular patients with respiratory insufficiency, the evidence for this practice is limited. A global respiratory management approach that monitors ventilation sufficiency, corrects inadequate ventilation and supports adequate cough clearance, addresses the respiratory limitations associated with neuromuscular respiratory insufficiency⁵⁻⁸. Secretion mobilization therapy would be indicated if pulmonary congestion develops secondary to respiratory infection and in the subset of neuromuscular patients with airway disease that encumbers muco-ciliary function. Cough augmentation should be used in conjunction with secretion mobilization to clear secretions in these patients. Mobilizing secretions without supporting adequate cough clearance may potentially result in asphyxiation. As the author indicates, in ALS and other neuromuscular diseases where progressive dysphagia develops secondary to bulbar dysfunction, there is a significant increase in the potential for aspiration related respiratory infection. The timely placement of a gastrostomy tube for nutrition, hydration and medication intake will be more effective in reducing the potential for aspiration related infection than a maintenance regimen of secretion mobilization therapy.

The author cites several studies showing both the effectiveness and acceptance of HFCWO compared to other secretion mobilization therapies in patients with intrinsic lung disease, most notably cystic fibrosis. While HFCWO has been shown to be an effective means of secretion mobilization for airway diseases that affect muco-ciliary transport, these studies alone do not provide substantiation to employ HFCWO for a completely different pathophysiology where muco-ciliary function is not encumbered. The author describes HFCWO as producing chest wall compression that creates a burst of air throughout the patient's airways resulting in a brief "cough-like" response that has been referred to as a "staccato cough". While the HFCWO induced rapid movement of air through the airway is designed to produce an airway shear force that mobilizes secretions, the characterization of this therapy as affecting a cough action may not be accurate. The author further states that HFCWO moves the mobilized secretions up through the airways where they can be coughed or suctioned. While HFCWO and other forms of chest physiotherapy may help a small number of neuromuscular patients with airway disease that encumbers secretion clearance, it does not support cough clearance which is the primary airway clearance limitation in this population.

The author describes mechanical in-exsufflation as a therapy that attempts to reproduce a cough maneuver by insufflation that inflates the lungs followed by immediate exsufflation that clears secretions from the lungs. She describes insufflation as having a potentially negative "muzzle loading" affect by blowing secretions into the distal respiratory tract. This conceptual idea is not supported

by any reported clinical observations. If the “muzzle loading” of secretions did occur with insufflation one would expect to find atelectasis in the lung fields distal to the impacted airways. Insufflation induced secretion impacted airways would potentially result in ventilation-perfusion abnormalities as well as an increased incidence of respiratory infection. We have prescribed mechanical in-exsufflation therapy for more than fifty neuromuscular patients with insufficient cough strength, as measured by peak cough flow, and have never observed iatrogenic related clinical symptoms associated with the use of this therapy. We have found no published reports of respiratory infection or ventilation-perfusion abnormalities associated with mechanical insufflation induced airway secretion impaction.

The author also states that patients with ALS often require mechanical ventilation, thereby rendering all of the airway clearance techniques, except HFCWO, unrealistic choices for regular periodic airway clearance. Mechanical in-exsufflation therapy is successfully used via tracheostomy for both ventilated and non-ventilated neuromuscular patients in a number of hospital rehabilitation centers. Our outpatient neuromuscular respiratory clinic program has four ALS patients on long term home mechanical ventilation who use mechanical in-exsufflation. The patients all prefer mechanical in-exsufflation compared to airway catheter suctioning for both comfort and effectiveness in clearing airway secretions.

We appreciate the author’s effort to support airway clearance therapy as a means of decreasing morbidity and mortality in neuromuscular patients with respiratory insufficiency. Secretion mobilization can be a beneficial respiratory therapy in addition to cough augmentation for those neuromuscular patients with airway disease related co-morbidity that results in retained secretions and for neuromuscular patients hospitalized for pulmonary congestion.

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