

Airway Clearance Needs in Neuromuscular Disease: An Overview

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Excluding the peripheral neuropathies¹, approximately 400,000 Americans suffer from hereditary or acquired neuromuscular diseases (NMD) affecting motor function.² Neuromuscular diseases of this class are characterized by progressive atrophy and weakness of skeletal muscle, skeletal-spinal deformities, limb contractures, and restrictive lung disease leading to poor respiratory function.³ Among inherited NMDs, the most prevalent include muscular dystrophies (Duchenne, Becker, facioscapulohumeral, limb girdle), the myotonias, and the spinal muscular atrophies. Acquired NMDs include amyotrophic lateral sclerosis, multiple sclerosis, Guillain Barré syndrome, and poliomyelitis. Many NMDs are progressive and incurable. Increased morbidities frequently result in shortened life expectancy.

Among patients with severe NMD, respiratory failure ranks as the leading cause of death.⁴ Pulmonary complications account for 75% to 90% of deaths occurring in patients with Duchenne's muscular dystrophy.^{5, 6} In amyotrophic lateral sclerosis (ALS), nearly all patients die of respiratory failure, usually within two to five years after onset of the disease.⁷ Recurrent pulmonary infection, the leading cause of morbidity among NMD patients, places them at high risk for progressive lung damage and, ultimately, respiratory failure.

The high incidence of pulmonary complications among NMD patients is influenced by a number of interrelated pathophysiological factors. Although the significance of each of these conditions depends upon the particular characteristics of the primary disease, patients with NMD are predisposed to the following life-threatening respiratory complications:

- Restrictive lung disease as a result of respiratory muscle weakness and spinal deformity
- Ineffective cough as a result of weakness and ensuing restrictive lung disease
- Immobility as a result of muscle weakness or dyscoordination
- Atelectasis as a result of secretion retention and restrictive lung disease
- Chronic aspiration as a result of dysphagia and exacerbated by an ineffective cough

Each of these contributing factors will be discussed in detail below.

Restrictive lung disease

Because NMD generally affects the respiratory muscles, most of these patients suffer from restrictive lung disease (RLD). The restrictive component tends to be especially severe in amyotrophic lateral sclerosis, Duchenne's muscular dystrophy, and spinal muscular atrophy types I and II.⁸ Even in the earliest stages of these diseases, when few if any respiratory symptoms have been reported, pulmonary function tests (PFTs) may be abnormal. In evaluating patients with early-stage NMD, Demedts et al found an average vital capacity of 75% of predicted norm, as well as a reduction in maximal transpulmonary and transrespiratory pressures.⁹

The severity of RLD depends not only upon the degree of muscle weakness and the rate at which weakness is progressing, but also on the presence of spinal deformity. Kyphoscoliosis and other abnormalities of the spine, which

typically result from inadequate muscular support of the spinal cord,¹⁰ are extremely common among people with NMDs. This is especially true for those with early onset diseases. Almost 100% of patients with early-onset spinal muscular atrophy, and 50-80% of those with Duchenne's muscular dystrophy (a childhood illness in which the average life expectancy is 20), develop kyphoscoliosis.¹¹ The effect of spinal deformity is reduced chest wall compliance.¹²

This reduced compliance of the chest wall combined with weakness of the diaphragm and other inspiratory muscles reduces maximum inspiratory pressure and limits inspiratory and vital capacities.¹³ Other effects of the restrictive component on lung volumes and capacities vary across specific disease states¹⁴, but in general, maximum expiratory pressure has been found to be more significantly affected than maximum inspiratory pressure, indicating that expiratory muscles may be more

profoundly affected than the diaphragm.¹⁵ In short, both the ability to take a big breath and the ability to generate expiratory force are typically affected; as explained below, both of these impairments, in turn, affect the cough.

Impaired or ineffective cough

Muscle weakness, spinal deformity and consequent RLD result in a weak cough and ineffective airway clearance.

A cough, if it is to clear secretions optimally and provide adequate airway defense, requires inspiration or insufflation to 85%-90% of total lung capacity, followed by closure of the glottis for approximately .2 seconds. Simultaneous with glottis closure, the expiratory muscles, stretched by the volume of the inspiration, recoil, creating intrapleural pressure to build up in the lungs. As a result of this pressure, opening the glottis results in high gas linear velocities that shear mucus from the walls of the airway.¹⁶

Neuromuscular disorders can interfere with this mechanism in several ways:

1. Diaphragm weakness¹⁷ or spinal deformity results in diminished inspiratory capacity. When inspiratory volume is insufficient, expiratory flows are diminished. Moreover, the expiratory muscles are not lengthened sufficiently to create optimum elastic recoil of the lung. Thus, intrapleural pressures are insufficient to create high peak cough flows.¹⁸ The point at which inspiratory volume is insufficient is believed to be approximately 2.5 liters or less.¹⁹

2. Bulbar muscle weakness impairs the patient's ability to close the glottis, which likewise prevents intrapleural pressures from building up.²⁰

3. Weakness of the expiratory (intercostal and abdominal) muscles lessens intrapleural pressures, thereby diminishing gas linear velocities and preventing mucus shearing.²¹

People with NMDs can sometimes benefit from maximal insufflation (via a ventilator) accompanied by a manual assisted cough. The effectiveness of the assisted cough, however, is diminished by any deformity of the thoracic cage or spine, by a distended abdomen or full stomach, and by obesity,²² all of which occur regularly among NMD patients.

Without access to either a spontaneous or an assisted cough, these patients' ability to clear their secretions is seriously impaired.

Immobility

Secretion clearance is further impeded by the immobility

common in this patient population. This is true because physical exercise is an important component of normal airway clearance, increasing mucus clearance by as much as 41%.²³ Exercise augments airway clearance in at least three ways: 1) increased air flow throughout the lungs helps mobilize secretions;²⁴ 2) increased activity of the parasympathetic nervous system reduces viscosity of secretions by promoting fluid secretion;²⁵ and 3) circulation of certain endocrines released by exercise results in further changes in the volume and viscosity of secretions.²⁶ Moreover, exercise has the most significant clearance effects on the peripheral airways that are not effectively cleared by coughing.²⁷

Because many people with NMDs suffer from impaired mobility coupled with an impaired or ineffective cough, they often experience pooled secretions²⁸ that provide fertile breeding ground for bacteria and contribute to a host of pulmonary complications. These complications include recurrent infection leading to tracheitis, bronchitis and pneumonia; impaired gas exchange; tissue damage; bronchiectasis. NMD patients are especially susceptible to hypostatic pneumonia;²⁹ in fact, a study of death certificates in Michigan revealed that people with Parkinson's disease, a form of NMD, are three to four times as likely to die of pneumonia/influenza than people in the general population.³⁰ Similar consequences have been observed in other NMD states.

In addition, pooled secretions as a result of immobility can result in ventilation-perfusion mismatch with consequent hypoxia, tachypnea and increased work of breathing.³¹ Mucus plugging as a result of insufficient airway clearance can lead to atelectasis. In fact, obstruction of the airway by mucus plugs is the most common cause of atelectasis in children with neuromuscular disabilities.³²

Atelectasis

Atelectasis is a frequent cause of respiratory failure in people with NMD.^{33,34} When Schmidt-Nowara and Altman systematically reviewed chest roentgenograms of 20 such patients hospitalized for respiratory failure, the roentgenograms showed atelectasis concurrent with respiratory failure in 17 of them. While it is impossible to determine definitively from the tests whether or not atelectasis actually precipitated the failure, it seems likely based on the high incidence.³⁵ Two independent studies have also revealed atelectasis in 10 of 13 Guillain Barré patients who required positive pressure ventilation³⁶ and in six out of 18 patients with respiratory failure secondary to myasthenia gravis.³⁷

While acute atelectasis is believed to result most frequently from infectious processes³⁸ and consequent secretion

retention and mucus plugging, it can also develop from RLD. Restrictive lung disease results in shallow breathing, which limits the distribution of ventilation throughout the lobes of the lungs, potentially leading to collapse of the peripheral airways. According to J. R. Bach, prolonged shallow breathing coupled with inability to take occasional deep breaths can lead to microatelectasis in approximately one hour. Chronic microatelectasis results in loss of or underdevelopment (in children) of lung tissue, which worsens the restrictive component by decreasing the elasticity of the chest wall, creating increased work of breathing. This increased work of breathing, in turn, contributes to hypoinflation and further atelectasis in a true vicious circle.³⁹

When unresolved respiratory infection occurs in a patient with RLD, this vicious circle is augmented by hyperproduction of secretions, mucus plugging and the eventual formation of scar tissue that further reduces chest wall elasticity.⁴⁰

Aspiration

A major source of chronic unresolved infection is aspiration of food particles, saliva and other liquid as a result of dysphagia. A large percentage of people with NMD suffer from dysphagia, including:

- almost 100% of those with amyotrophic lateral sclerosis
- up to 51% of those with poliomyelitis
- 44% of those with myasthenia gravis
- 38% of those with limb-girdle syndrome
- 33% of those with myotonic dystrophy
- 32% of those with spinal muscular atrophy
- 20% of those with Duchenne's muscular dystrophy
- 6% of those with facioscapulohumeral muscular dystrophy.⁴¹

In some patients, dysphagia is severe enough to require gastrostomy.

The prevalence of dysphagia and consequent aspiration among these patients is a function of several anatomical abnormalities: bulbar (throat) muscle weakness⁴²; cricopharyngeal bars, which occur when the esophageal sphincter fails to relax in order to allow food to pass from the pharynx into the esophagus⁴³; and poor airway defense mechanisms, i.e. an ineffective cough. Since the likelihood of infection resulting from aspiration depends upon the frequency and volume of the aspirate as well as its contents, these abnormalities put NMD patients at considerably greater risk than those in the general population.⁴⁴ Moreover, these patients often swallow very slowly, which also increases the likelihood of aspiration

and ensuing infection; in patients who routinely take five or more seconds to complete a swallow, incidence of aspiration pneumonia is 90%.⁴⁵

Because saliva contains, on average, 10^6 - 10^8 bacteria per milliliter,⁴⁶ aspiration of a moderate quantity of saliva introduces a large number of bacteria into the sterile airways and can lead to acute pneumonia,⁴⁷ tracheitis, bronchitis⁴⁸ and chronic lung disease.⁴⁹

Respiratory infection in these patients results in increased secretion production and puts them at risk for acute decompensation.⁵⁰

Consequent need for airway clearance

Because people with NMDs are prone to respiratory infections due to frequent aspiration and impaired airway clearance, and because respiratory infection frequently precipitates atelectasis and respiratory failure, these people often require daily airway clearance.

Traditionally, the removal of mucus from the lungs is accomplished using a technique called chest physical therapy (CPT). Under any circumstance, CPT is time-consuming and labor intensive. Typical treatments last 20 to 30 minutes and may need to be administered up to three times per day. Throughout this time, a caregiver must administer aggressive percussion to the patient's chest wall while having the patient lie in a number of different positions. CPT thus requires the presence of a dedicated, physically-able caregiver with time to administer adequate CPT treatments. Moreover, people with NMDs often exhibit structural abnormalities such as scoliosis that make it difficult or impossible for them to assume the positions required for effective postural drainage. Positioning is likewise impossible for those patients with feeding tubes in place. Moreover, NMD patients may be unable to coordinate expiration sufficiently to use technique-dependent airway clearance modalities, or to generate the expiratory pressures they necessitate.

For these patients, The Vest™ Airway Clearance System provides an alternative therapeutic modality that provides consistent, quality airway clearance. 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.

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- patients with acquired peripheral neuropathies demonstrate disease-related pulmonary dysfunction. Neuromuscular Disease Database: UC-Davis School of Medicine, Physical Medicine and Rehabilitation. Wysiwyg://18/http://disability.ucdavis.edu/Clearinghouse/VirtualLibrary
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