

Airway Clearance Needs in Duchenne's Muscular Dystrophy: An Overview

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The muscular dystrophies are a group of inherited, progressive neuromuscular disorders classified on the basis of specific phenotypic and genetic characteristics.¹ Among such conditions, Duchenne muscular dystrophy (DMD) is the most common and most debilitating.^{2,3} DMD is a sex-linked recessive disease which results in the absence of dystrophin, a protein found inside the muscle cell membrane. Sometimes called pseudohypertrophic muscular dystrophy because fatty and fibrous tissue may replace certain enlarged muscle groups, DMD is the second most common genetic disorder in humans, affecting one in 3,000 live male births.^{4,5}

Duchenne muscular dystrophy is named for the Italian neurologist who in 1861 recognized the condition as a distinct disease entity.^{6,7} DMD is characterized by progressive atrophy and weakness of skeletal muscle, skeletal-spinal deformities, limb contractures, and restrictive lung disease resulting in life-threatening pulmonary problems. Although present at birth, symptoms of Duchenne typically appear between the ages of three and seven. Early signs of DMD include frequent falls and difficulty in running, jumping, and hopping. By the age of six, contractures of the heel cords and iliotibial bands appear, and are associated with a characteristic waddling gait, toe walking, lordosis, and difficulty in standing up and climbing stairs. Loss of muscle strength is steady and progressive, typically involving the pelvic girdle before the shoulder girdle. Limb contractures and scoliosis develop. By the age of twelve, most boys with DMD are confined to wheelchairs.

Although all forms of muscular dystrophy share deterioration of the respiratory and other skeletal muscles, respiratory insufficiency in Duchenne follows a relentless downhill course. Chest deformity as a result of progressive scoliosis impairs pulmonary function, which is already compromised by muscle weakness. By late adolescence, individuals with DMD experience serious, recurrent pulmonary infections. Most DMD patients die by the age of 20. In DMD, death is most frequently a direct result of chronic ventilatory failure, often with a superimposed acute respiratory insufficiency due to pneumonia, mucus plugging, or atelectasis.^{8,9} Seventy-five to ninety percent of DMD deaths are a direct result of pulmonary complications.^{10,11,12}

The high incidence of illness and death in DMD from respiratory causes is influenced by several interrelated pathophysiological factors. Although the significance of each of the following conditions is related closely to the extent of disease progression, DMD patients are predisposed to the following life-threatening complications:

- Restrictive lung disease as a result of respiratory muscle weakness and spinal deformity
- Ineffective cough as a result of weakness of the muscles of the abdomen and diaphragm
- Immobility as a result of muscle weakness or dyscoordination
- Predisposition to atelectasis as a result of secretion retention and restrictive lung disease
- Chronic aspiration as a result of dysphagia and exacerbated by ineffective cough

Restrictive Lung Disease

The clinical course of DMD is characterized by the development of both spinal deformity and progressive respiratory insufficiency secondary to failure of the breathing apparatus. Without the muscular strength necessary to take regular deep breaths, the lungs and chest wall stiffen, limiting the ability to take in sufficient oxygen

to meet metabolic requirements. Even in the earliest stages of the disease, when few if any respiratory symptoms have been reported, pulmonary functions begin to decline at fairly consistent, age-related rates.¹³ At ages 7-10, percent of predicted forced vital capacity (FVC) declines at an average annual rate of -0.3%; between ages 10-20, -8.5%; and after age 20, -6.2%.¹⁴ Finally, at mean age of 21.53,

percent predicted FVC falls to under 10% and respiratory failure ensues.¹⁵ Decreased airway pressures, most notably maximal expiratory pressure, appear before reductions in FVC, but follow the same pattern.

The severity of restrictive lung disease (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. Scoliosis develops in at least 90% of DMD patients when they become wheelchair bound.¹⁶ More than half of these individuals also develop kyphoscoliosis.¹⁷ One effect of scoliosis and kyphoscoliosis is a reduced chest wall compliance, limiting the ability to take a deep breath.¹⁸ In DMD, 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. Like muscle weakness, spinal deformities are progressive, and decline in pulmonary function in terms of FVC occurs in concert with the rate of curve progression.^{19,20,21}

Effects of the restrictive component on lung volumes and capacities vary from individual to individual, but in general, maximum expiratory pressure is affected more significantly than maximum inspiratory pressure, indicating that in DMD, the expiratory muscles are affected more profoundly than the diaphragm.²² Both the ability to take a big breath and the ability to generate expiratory force are affected. In RLD associated with DMD, there is nothing wrong with the lungs themselves. The problem is not obstruction, but an inability to breathe deeply enough to fill and clear the lungs. These impairments, in turn, affect the cough function.

Impaired or ineffective cough

Coughing is an important component of respiratory health. Forceful expulsion of air from the lungs during a cough loosens, mobilizes, and clears mucus secretions. In DMD, muscle weakness, spinal deformity, and consequent RLD result in a weak cough and ineffective airway clearance. In advanced DMD, the ability to cough is absent.²³

An effective cough generates a force sufficient to clear secretions and provide adequate airway defense. Such a cough requires inspiration or insufflation to 85%-90% of total lung capacity, followed by closure of the glottis for approximately 0.2 seconds. Simultaneous with glottis closure, the expiratory muscles, stretched by the volume of the inspiration, recoil, thus allowing intrapleural pressure to build up in the lungs. As a result of such pressure, when the glottis opens, mucus is sheared from the walls of the airway by expired gas at high linear volumes.²⁴

Duchenne muscular dystrophy can interfere with this mechanism in several ways:

1. Diaphragmic weakness or spinal deformity diminish inspiratory capacity. If the diaphragm is weak, if chest wall compliance is compromised, or if spinal deformity interferes with expansion of the chest, inspiratory volume is insufficient and expiratory flows are diminished. Moreover, expiratory muscles cannot lengthen sufficiently to permit optimum elastic recoil of the lung. Thus, intrapleural pressures are insufficient to produce high peak cough flows.²⁵ Inspiratory volumes at or below 2.5 liters are generally regarded as inadequate for effective cough function.²⁶
2. Bulbar muscle or throat muscle weakness impairs the ability to close the glottis, thus inhibiting the build-up of intrapleural pressures. Without adequate bulbar muscle control, it may be impossible to produce a productive cough.²⁷
3. Weakness of the expiratory (intercostal and abdominal) muscles lessens intrapleural pressures, thereby diminishing gas linear velocities and preventing mucus shearing.²⁸ Normal cough flows are between 6 and 30 liters per minute; with advanced DMD, cough flows may be as low as one liter per minute.²⁹ Abdominal muscles, which weaken progressively in DMD, are even more important than the diaphragm in facilitating the forceful expiration required for effective cough.³⁰ Weak abdominal muscles mean weak cough.

People with DMD may benefit from maximal insufflation (via a ventilator) accompanied by a manual assisted cough.³¹ Certain conditions which occur commonly in DMD, however, including deformity of the thoracic cage or spine, distended abdomen, and obesity, may compromise the effectiveness of assisted cough.³²

Without access to either a spontaneous or assisted cough, the ability to clear secretions is seriously impaired. Mucus clearance is essential to the prevention of infection. In DMD, underventilation and inadequate coughing may persist for some time without causing a respiratory crisis. Typically, respiratory emergencies appear suddenly, often in association with respiratory infections, causing life-threatening situations in which a major airway becomes clogged with retained secretions. Because the ventilatory muscles and cough reflex are too weak to eliminate the mucus, air passages become plugged. Such mucus plugging not only reduces the amount of lung tissue available for gas exchange, but also greatly increases the likelihood of more serious infection.

Immobility

Secretion clearance problems associated with immobility are a recognized aspect of DMD.

Physical exercise is an important component of normal airway clearance, increasing mucus clearance by as much as 41%.³³ For those peripheral airways not effectively cleared by coughing, exercise is the most important component of the clearance mechanism.³⁴

Exercise augments airway clearance in at least three ways: 1) increased airflow throughout the lungs helps mobilize secretions;³⁵ 2) increased activity of the parasympathetic nervous system reduces viscosity of secretions by promoting fluid secretion;³⁶ and 3) circulation of certain endocrines released by exercise results in further changes in the volume and viscosity of secretions.³⁷ Because most boys with DMD are confined to wheelchairs by the age of twelve, immobility contributes progressively to their airway clearance difficulties. The combined effects of immobility and ineffective cough contribute to the accumulation of pooled secretions.³⁸ Pooled secretions 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; and bronchiectasis. In addition, such pooled secretions can contribute to ventilation-perfusion mismatch with consequent hypoxia, tachypnea and increased work of breathing.³⁹

Atelectasis

Atelectasis is a frequent cause of respiratory failure in people with neuromuscular disease, including DMD.⁴⁰ The collapse or closure of alveoli, or the lung units responsible for gas exchange, may involve an entire lung, one or more lobes, or one or more lung segments.⁴¹ Significantly, obstruction of the airway by mucus plugs is the most common cause of atelectasis in children with neuromuscular disorders.⁴² Widespread microatelectasis, which increases as lung compliance declines, is a typical feature of the disease.⁴³ Frequently, such diffuse microatelectasis involves small areas of alveolar collapse too small to appear on chest x-rays.⁴⁴

In DMD, several factors promote the development of atelectasis. As weakened respiratory muscles lose the ability to move large volumes of gas, the ability to inspire large tidal volumes, or sigh, may also be diminished or lost. The adverse effects of breathing only at low tidal volumes include:

- Loss of the alveolar stretching required to ensure adequate production of surfactant, thus allowing alveolar collapse.
- Increasing pleural pressure to levels exceeding airway pressure, resulting in closure of peripheral airways.

Acute atelectasis, however, is believed to result most frequently from respiratory infection.⁴⁵ The upper respiratory

tract infections associated with DMD often exacerbate lower respiratory tract secretion retention, leading to pneumonia, mucus plugging, pulmonary atelectasis, and respiratory failure.⁴⁶ Restrictive lung disease (RLD) also contributes to the development of acute atelectasis. The shallow breathing characteristic of RLD limits the distribution of ventilation throughout the lobes of the lungs, increasing the risk of collapse of the peripheral airways. Even in persons with normal lungs, periodic hyperinflation of the lungs is required to prevent closure of lung units. In DMD, prolonged shallow breathing, coupled with the inability to take occasional deep breaths, can result in the rapid development of microatelectasis, sometimes in as little as one hour.⁴⁷ Chronic microatelectasis in children may result in the loss of or underdevelopment of lung tissue. Such a development worsens the restrictive component of the respiratory pathology by increasing the elastic tension of the chest wall, thus creating increased work of breathing. This increased work of breathing, in turn, contributes to hypoinflation and to further atelectasis in a true vicious cycle.⁴⁸

When unresolved respiratory infection occurs in DMD patients with RLD, the vicious cycle promoting atelectasis is exacerbated by hyperproduction of secretions, mucus plugging and the eventual development of scar tissue that reduces further chest wall compliance.⁴⁹

Aspiration

Dysphagia, or difficulty in swallowing, is a consequence of anatomical abnormalities or weakness of the muscles associated with swallowing. Gastroesophageal reflux occurs when a defective lower esophageal sphincter allows stomach contents to surge into the esophagus. Patients suffering from either disorder are at risk for aspiration. Patients with DMD present esophageal motor disorders in both striated and smooth muscle, as well as upper and lower gastrointestinal symptoms.⁵⁰ By the time boys with DMD become wheelchair bound, nearly all suffer from dysphagia.⁵¹ In cases where dysphagia is severe, gastrostomy may be required.

In addition to underlying muscle pathology, including bulbar muscle weakness, the prevalence of dysphagia and consequent aspiration in DMD patients is a function of certain anatomical abnormalities including cricopharyngeal bars, which appear when the esophageal sphincter fails to relax to permit food to pass from the pharynx into the esophagus, and the dysfunctions leading to ineffective cough.^{52,53} These individuals often swallow very slowly, increasing further the likelihood of aspiration. In patients who routinely take five or more seconds to complete a swallow, the incidence of aspiration pneumonia is 90%.⁵⁴ The abnormalities which cause dysphagia place DMD patients at significant risk for serious pulmonary infection.⁵⁵ The probability of infection resulting from aspiration depends on

the frequency and volume of aspirate as well as its contents. Because saliva contains, on the average, 10^6 - 10^8 bacteria per milliliter, aspiration of a moderate quantity of saliva introduces a large number of bacteria into sterile airways, increasing the risk of acute pneumonia,⁵⁶ tracheitis and bronchitis,⁵⁷ and chronic lung disease.⁵⁸ Respiratory infection secondary to aspiration results in mucus hypersecretion. Because the ability to clear secretions is impaired in people with DMD, their risk for acute decompensation is increased significantly.⁵⁹

Indications for airway clearance

The primary respiratory pathology in DMD is mechanical in nature, consisting chiefly of weak or absent cough and hypoventilation secondary to restrictive lung disease. As a result of these deficits, people with DMD are prone to interrelated pulmonary complications including atelectasis, respiratory infection and respiratory failure. In DMD, acute respiratory failure is precipitated most frequently by otherwise uncomplicated upper respiratory tract infections.⁶⁰ During such infections, existing pulmonary deficits are worsened by bronchial mucus plugging and further weakening and fatigue of respiratory muscles. Such illnesses initiate a downward spiral involving recurrent pneumonias, hospitalizations, tracheal intubations, and eventually, permanent assisted ventilatory support or death.⁶¹ Because respiratory infections precipitated by secretion retention pose a serious threat to vulnerable patients with muscle weakness and poor cough, airway clearance therapy is now recognized as an essential component in the standard respiratory care regimen for DMD.^{62,63,64,65}

Traditionally, the removal of mucus from the lungs is accomplished using a technique called chest physiotherapy (CPT). Chest physiotherapy 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. The technique is sometimes called percussion and postural drainage (P&PD). CPT is based on the theory that percussion to various areas of the chest and back transmits shock waves through the chest wall, loosening secretions in the airways.

Unfortunately, CPT is fraught with intrinsic and extrinsic difficulties. Under the best of circumstances, CPT is a time consuming, labor-intensive treatment requiring significant skill and strength on the part of the therapist and the mental and physical cooperation of the patient. Typical treatment sessions last 20-30 minutes and may be required up to three times daily. During postural drainage, the patient is placed in various positions on a slant board to facilitate gravitational movement and expectoration of mucus. Such manipulations

are difficult or impossible to achieve in patients who are medically complex, anatomically deformed, immobile, dependent on ventilators or other pieces of cumbersome medical equipment, or who are physically large. Neither can postural drainage be accomplished effectively in patients who are mentally impaired, uncooperative, or severely spastic. People with DMD typically manifest several of these contraindications.⁶⁶ Such individuals may also be unable to tolerate the transient hypoxemia associated with postural drainage, to coordinate expiration sufficiently, or to generate the expiratory pressures required for effective CPT.

For patients unable to tolerate traditional CPT, The Vest™ airway clearance system provides an alternative therapeutic modality that delivers consistent, hospital-quality airway clearance. Developed initially for the treatment of cystic fibrosis, The Vest™ system is appropriate for any patients with excessive mucus production and difficulty clearing secretions.⁶⁷ Its method, high-frequency chest wall oscillation, achieves the outcomes for airway clearance therapy published by the American Association for Respiratory Care.⁶⁸ 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.

Assisted ventilation

Although DMD remains incurable, it must not be regarded as untreatable. In recent years, physicians have recognized the benefits of a variety of interventions designed to modify the progress of the disease.⁶⁹ Chief among these interventions are incremental introduction of airway clearance therapy; measures to control spinal deformity; non-invasive ventilation, used initially during the night⁷⁰ and eventually to provide 24 hour ventilatory support; and finally invasive assisted ventilation.⁷¹

Non-invasive ventilatory support

The clinical course of Duchenne muscular dystrophy is characterized by respiratory insufficiency secondary to failure of the breathing apparatus. Until recently, patients who so elected then received life support by means of mechanical ventilation via tracheostomy. Although eventually all surviving DMD patients require full-time invasive ventilation, a variety of non-invasive methods have been introduced, most notably nasal intermittent positive pressure ventilation (NIPPV), which can maintain patients for several years. Initially, NIPPV is used to manage nocturnal hypoventilation. As respiratory decline progresses, NIPPV may be used 24 hours daily.⁷² In addition to quality of life advantages, studies demonstrate significant reductions in morbidity and hospitalization rates and impressive increases in survival among patients receiving non-invasive ventilatory support.^{73,74,75} However, for non-invasive NIPPV to be effective, adequate airway clearance is essential. Retained

secretions and recurrent infections exacerbate existing poor gas exchange and sleep-related hypoxemia.⁷⁶ Airway clearance is of critical importance during episodes of upper respiratory tract infection.

Assisted ventilation via tracheostomy

Unfortunately, despite non-invasive ventilatory support, DMD patients experience a progressive decline in pulmonary function and bulbar muscle strength, resulting in recurrent aspiration and other problems.⁷⁷ When individuals are unable to clear secretions or obstructions from the respiratory tract sufficiently to maintain open airways, tracheostomy is required. However, although artificial airways are intended to maintain airway patency, their presence may also contribute to ineffective airway clearance as a result of their adverse effect on normal airway clearance mechanisms.⁷⁸ Consequently, regular airway clearance therapy plays a vital role in the maintenance of tracheostomized ventilator-dependent patients.⁷⁹ Many patients who elect ventilator dependence via artificial airway experience both increased survival and, on the whole, a satisfactory quality of life.^{80,81,82} In a study of 23 DMD patients using ventilators with and without tracheostomy, the overall length of survival was increased from 19.9 years to 25.9 years.⁸³

Because the decision to rely upon full-time mechanical ventilation via tracheostomy raises both ethical questions and practical problems concerning maintenance and nursing care, the issue has generated considerable debate among DMD clinic directors.^{84,85} Advocates point to comparable treatment instituted routinely for individuals suffering high cervical lesion spinal cord injury. Others stress the importance of preserving maximal physical capacity and function in anticipation of the development of specific treatments, including gene therapy.^{86,87} Although presently there is no cure for DMD, meaningful supportive care is available with the incremental use of:

- active physiotherapy to delay the development of muscle contractures and deformities
- scoliosis management
- routine airway clearance therapy to prevent or reduce the pulmonary morbidities associated with secretion retention
- noninvasive ventilation
- assisted ventilation delivered via tracheostomy

With the assistance of well-managed medical care, many boys and young men with DMD are able to adapt to their physical disabilities and live emotionally, intellectually, and spiritually satisfying lives.

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⁷¹ Ibid.

⁷² Bach JR, op. cite (n. 63).

⁷³ Ibid.

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