

Airway Clearance in Duchenne Muscular Dystrophy

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ABSTRACT

This is a summary of the presentation on airway clearance in neuromuscular disorders presented as part of the program on airway clearance in Duchenne muscular dystrophy at the 30th annual Carrell-Krusen Neuromuscular Symposium on February 20, 2008. *Pediatrics* 2009;123:S231–S235

DUCHENE MUSCULAR DYSTROPHY (DMD) is an X-linked recessive hereditary disease that affects both skeletal and cardiac muscle.^{1–5} The protein dystrophin is made in inadequate amounts, which leads to the break down of muscle cell walls and the cell's ultimate death. This loss of muscle leads to numerous problems including axial skeletal muscle weakness and the loss of ambulation, diaphragmatic weakness with recurrent pneumonias and progressive respiratory failure, and cardiomyopathy with arrhythmias.^{2–5} The combination of cardiopulmonary failure leads to early death, with the mean life span of affected males being ~25 years.^{1,2,4–7}

Although there is currently no cure for DMD, supportive therapies can markedly decrease the morbidity of this condition, thus prolonging the life span of these young men and improving their quality of life. This review will focus on airway clearance and cough augmentation as ways of decreasing the incidence of atelectasis, pneumonia, and respiratory failure associated with this muscular dystrophy.

NORMAL COUGH

Coughing works by helping to clear airway secretions from the lungs.^{8–11} These secretions act as a protective layer covering the bronchi and bronchioles. They trap environmental particulate matter as well as bacteria and viruses that are inhaled during respiration.^{8,11} Cilia protruding from bronchial epithelial cells move this protective layer in a proximal direction.¹¹ Coughing augments the ciliary clearance of these secretions, with the ultimate expectoration or swallowing of this material.⁹

Coughing consists of 3 components: an inspiratory phase; a contraction phase; and an expiratory phase.^{8,10} During the inspiratory phase, the patient inhales, usually to 60% to 90% of total lung capacity.^{2,3,10,12,13} In the contraction phase, the patient's glottis closes and the expiratory muscles begin to contract. This phase lasts only 0.2 seconds; however, during this time, elevated intrathoracic pressures are reached.⁸ During the final (expiratory) phase of a cough, the glottis opens, and there is a sudden expiratory rush of air. It is during this phase of the cough that large shearing forces are generated, cleaving off parts of the secretory lining and carrying with them the trapped foreign material.^{8,10} During this expiratory phase, velocities as high as 12 m/second can be reached, with flow rates of 360 to 1000 L/minute being obtained for a normal cough.^{2,3,8,10,12}

Many conditions can lead to an increase in airway secretions. In pediatrics, 2 of the more common etiologies of increased airway secretions include pneumonia and asthma. These illnesses are associated with an increase in airway inflammation and a resultant increase in secretion volume. One of the functions of the cough in these situations is to clear this increased volume of mucus. If the increased secretions are not expelled, mucus can build up, which can lead to the development of atelectasis and/or pneumonia with resultant ventilation-perfusion mismatch and hypoxia.¹⁴ If the underlying condition progresses unabated and/or the ability of the cough to clear this extra material is overwhelmed, respiratory failure can ensue. If any phase of the cough should be impaired, a vicious cycle may develop, leading to a worsening of the clinical situation.

ALTERED COUGH IN DMD

With DMD, secretions in the lung are normal in quantity and quality. Several factors, however, conspire to diminish the effectiveness of the cough. The neuromuscular weakness affects both the inspiratory and expiratory phases of the cough. Any type of glottic dysfunction will prevent the adequate build up of intrathoracic pressure seen in the second phase of a cough. Finally, scoliosis (frequently seen in DMD) can also diminish the effectiveness of the cough.^{6,12}

Inspiratory muscle weakness prevents the patient from taking a deep breath, thus limiting the volume of air available to be expired during the exhalation phase of the cough.^{1,8,13} The diaphragm is the major muscle of inspiration, and its weakness greatly affects the patient's ability to achieve the adequate lung volume needed for an

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Abbreviations

DMD—Duchenne muscular dystrophy

SaO₂—oxyhemoglobin saturation

CPF—cough peak flow

MEP—maximal expiratory pressure

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effective cough. Furthermore, decreased chest wall compliance is seen in DMD, and this increased chest wall stiffness decreases the available chest wall excursion, further limiting the inspiratory capacity required for an effective cough.^{1,15}

Any dysfunction at the level of the glottis can prevent the generation of adequate intrathoracic pressures needed to develop the sufficient shearing forces required for expectoration of airway secretions.^{8,13} Although most patients with DMD have satisfactory glottic function, other types of neuromuscular weakness, such as spinal muscle atrophy type 1 (Werdnig-Hoffman disease) or amyotrophic lateral sclerosis, include bulbar dysfunction leading to the failure of the vocal cords to fully adduct and maintain apposition during the second component of a cough.¹⁶ Thus, adequate pressures are not reached, and the effectiveness of the cough is diminished. Some patients with DMD may require the placement of a tracheotomy tube to allow for assisted ventilation in treating respiratory failure. This tracheostomy allows for air leakage and prevents achieving adequate intrathoracic pressures, thus confounding the effects of muscle weakness and further diminishing the cough.

Although both the inspiratory and expiratory muscles are weakened in DMD, the expiratory muscles are more affected and provide for a dramatic impact on the quality of a cough.² The exhalation phase is compromised with the patient being unable to generate enough force to properly expel secretions present in the airway.¹³ This limitation is further exacerbated by several factors related to the compromised inspiratory phase.^{8,13} The decreased chest wall compliance has a direct effect on the movement of the chest wall during both inspiration and expiration.^{6,8,15} As mentioned earlier, this increase in chest wall stiffness limits the ease of inspiration, as well as expiration, during a cough. Because the chest wall was not fully inflated at the beginning of the expiratory phase, the muscles of expiration were not stretched to their point of optimal mechanical advantage to maximize their expiratory contraction.⁸ The lungs themselves were not fully inflated as to allow for maximal passive elastic recoil during expiration. These factors further limit the effectiveness of the expiratory muscles. Finally, because the lung was not fully inflated to begin with, the conducting airways were not maximally dilated; thus, their cross-sectional diameter is diminished, increasing airway resistance during exhalation, which further limits flow rates.

Patients with DMD begin to develop scoliosis once they lose ambulation and become confined to a wheelchair. This usually occurs by the second decade of life.^{2,17} This scoliosis has many deleterious effects on breathing including discomfort, the impairment of respiration by placing the muscles of inspiration and expiration at a mechanical disadvantage, further worsening chest wall compliance, and the physical distortion of bronchi leading to diminished air flow. These factors can easily affect the quality of a patient's cough.

Thus, by starting with diminished lung volumes secondary to weakened inspiratory muscles, the potential loss of adequate intrathoracic pressures being achieved

from glottic dysfunction, and inadequate expiration caused by weakened muscles of expiration, confounded by increased chest wall stiffness and airway obstruction from scoliosis, the cough in DMD is ineffective for maintaining proper airway hygiene.

MEASUREMENT OF COUGH EFFECTIVENESS

Although the cough is diminished in DMD, its weakening follows a predictable pattern. There are various ways of measuring a cough's effectiveness that allow the attending physician to know when the optimal time occurs for adding interventions to assist or augment the cough.

Pulmonary function testing should be performed on a routine basis for all patients with DMD.^{17,18} Spirometry in DMD follows a generally predictable course, with values increasing as the patient ages, then plateauing after requiring the permanent use of a wheelchair, followed by gradual deterioration over time as muscle weakness progresses.^{2,19} Although the absolute value (or percent predicted) of the patient's forced vital capacity does not directly correlate with the quality of a patient's cough,² the degree of restrictive lung disease present gives one a good idea of the patient's pulmonary reserve. This can help determine when either cough assisting or respiratory support is needed or, at the least, requires further evaluation. For example, a forced vital capacity of <40% to 50% of predicted or <1 L are associated with nocturnal hypoventilation and the need for non-invasive ventilation, independent of clinical symptoms.^{1,19-21} The use of such support heralds increased risk for diminished airway clearance and the need to augment the patient's weakened cough.

Measurement of oxygen saturation is also useful.^{6,7,18} A normal oxyhemoglobin saturation (SaO_2) suggests good air exchange and the absence of ventilation-perfusion mismatch. Hypoxia, however, suggests the presence of poor airway clearance and/or hypoventilation with secondary ventilation-perfusion mismatch.^{6,18,20} Thus, a patient who is clinically well with new-onset daytime hypoxia might require assisted ventilation because of hypoventilation, whereas a patient with new-onset hypoxia who is acutely ill with a viral process more likely has a diminished ability to clear airway secretions and would benefit by cough-assisting maneuvers.⁷

Similarly, measurement of CO_2 levels via capnography can aid in the assessment of overall respiratory muscle function. Elevations of end-tidal CO_2 levels suggest hypoventilation, either from inadequate nocturnal ventilation with a spillover effect to the morning or overall inadequate ventilation both while awake and asleep.⁵ This elevation of CO_2 levels heralds worsening muscle strength, which will have an effect on the patient's ability to develop an adequate cough.⁵ In general, equipment for monitoring oxygenation is more readily available in the community, so Sao_2 levels are more closely followed on a day-to-day basis. End-tidal CO_2 levels are usually followed on a more chronic basis. Blood gas measurement (arterial, capillary, or venous) can also help assess for adequacy of gas exchange, although the use of noninvasive measures is preferred.

Two tests that greatly predict the adequacy of a patient's cough are the measurement of the cough peak flow (CPF) and the maximal expiratory pressure (MEP).^{8,12,16,20} CPFs are performed by using a standard peak flow meter with the attachment of a full face mask at the mouthpiece.¹⁸ The patient then coughs into the mask. Someone with no underlying abnormalities should be able to generate a CPF of >360 L/minute.¹² With DMD, patients with a CPF of >270 L/minute are felt to have an adequate cough.^{7,8,16,20} Studies have demonstrated that a CPF of <160 L/minute is associated with diminished cough effectiveness and that cough augmentation is required.^{7,16,20} Patients with a CPF between 160 and 270 L/minute have an adequate cough when they are well.^{8,16} That being said, patients with DMD frequently get weaker with illness.^{12,22,23} As a result, a patient with a baseline CPF in this range when well will often have it drop to <160 L/minute when ill. Therefore, patients with a baseline CPF of <270 L/minute benefit from cough-assisting maneuvers.^{3,7,17,23} Some patients may have trouble performing the CPF maneuver. Measurements of MEP may be substituted for the CPF.^{8,17,20} A MEP of >60 cm H₂O is associated with an effective cough,^{13,17} whereas a MEP of <45 cm H₂O is associated with diminished cough effectiveness and warrants the use of cough-assisting techniques.^{8,17}

AIRWAY CLEARANCE AND COUGH-ASSISTING TECHNIQUES

Once it has been determined that a patient with DMD has an inadequate cough, he or she would benefit from instituting cough assistance. There are a wide variety of techniques available from which to choose, ranging from manual techniques to mechanically assisted maneuvers. Any of the methods used require a combination of improved insufflation of the lungs to achieve sufficient lung volumes for an effective cough (phase 1) in conjunction with adequate forced expiratory techniques to augment the patient's natural, but weakened, cough (phase 3). Ideally, the patient should be able to (briefly) hold his or her breath before exhaling to maximize the precough lung volumes and help generate sufficient intrathoracic pressures (phase 2).

There are a wide variety of ways to maximally insufflate the lungs to an adequate precough volume. Glossopharyngeal breathing (air stacking or "frog breathing") is an effective technique for inflating the lungs one gulp of air at a time.^{14,18,24} This technique has the advantage that no equipment or assistance is required. It can, however, be a difficult technique to master. A simpler method of achieving sufficient lung volumes is with the use of a self-inflating ambu bag.²⁵ Direct feedback to an assistant can be given to help properly inflate the lungs to a sufficient but comfortable volume for the patient.

Mechanical equipment is available to help inflate the lungs. Patients who require the use of a volume-cycle ventilator for treatment of respiratory insufficiency can use the ventilator in assist mode to breath stack.²⁴ The CoughAssist (Respironics Corp, Millersville, PA) machine (the mechanical insufflator-exsufflator) has proven to be a boon to airway clearance in patients with neuromuscular weakness.^{6,12} It provides both an inspiratory phase (to inflate the lungs) and an expiratory phase (for the actual

cough) all in one piece of equipment. Both manual and automatic modes can be used. This equipment has been shown to be effective for children and adults with various etiologies to their muscle weakness.¹² With this machinery, the duration of treatment and pressures used can be preset to maximize the effectiveness of the cough and the patients' comfort. Studies have shown that the CoughAssist machine is well tolerated, with no increased risk for complications such as pneumothorax, gastroesophageal reflux, or pulmonary hemorrhage.¹² It also can be used through various interfaces such as the mouth, a full face mask, or an endotracheal or tracheostomy tube.⁸

For the expiratory phase of an assisted cough, manual cough-assisting techniques are simple but effective maneuvers for aiding in airway clearance. They require the use of an assistant trained in this technique. It can be as simple as a properly placed abdominal thrust (similar to a Heimlich maneuver).^{6,8} From a mechanical perspective, the CoughAssist machine (as discussed above) provides for a timed, forced exhalation maneuver that effectively mimics a cough. For patients who lack the ability to expectorate any produced sputum, having suction equipment available will allow these secretions to be easily removed from the mouth rather than have the patient swallow them.^{18,23}

Once assisted coughing is recommended, it should ideally be used not just when the patient is ill but also when he or she is well to allow the patient and caregivers to remain well versed in its use for times when its use is more urgently required.¹² Once- to twice-per-day maintenance therapy should provide for optimal ongoing training in this airway-clearance technique.

The use of chest physiotherapy in patients with DMD is more controversial and not fully established.¹⁷ Although manual chest physiotherapy, intrapulmonary percussive ventilation, and high-frequency chest wall oscillation (the Vest Airway Clearance System [Hill-Rom Services, Inc, St Paul, MN]) are effective at aiding airway clearance in conditions of highly viscous secretions (such as with cystic fibrosis), the underlying problem with DMD is not so much hyperviscous secretions but, rather, the inability to clear the normal secretions and/or increased volume of secretions associated with infection.¹⁴ Certainly, in cases of DMD with focal atelectasis from mucus plugging, these techniques may prove useful.^{9,15} At this time, however, there are not enough adequate studies available to recommend its routine use; rather, a case-by-case approach will be required. Of importance, optimal airway clearance may require some type of positioning to be entailed.¹⁵ This may prove problematic in a less-than-fully mobile patient. Also, the head below the body position may transiently decrease the already diminished lung volume (from the abdominal contents pushing up against the diaphragm), so care must be taken to prevent further respiratory dysfunction during the technique.

The use of airway-secretion-thinning agents has also not yet been fully established.⁸ Dornase α (Pulmozyme [Genentech, Inc, South San Francisco, CA]) and *N*-acetylcysteine (Mucomyst [Apothecon Inc, Princeton, NJ]) have not been formally studied for DMD, so their use cannot be

routinely recommended. In cases of isolated atelectasis secondary to mucus plugging, their use (in combination with chest physiotherapy or high-frequency chest wall oscillation) can be considered. Drying agents, such as glycopyrrolate (Robinul [Baxter Pharmaceutical, Deerfield, IL])^{26,27} might be useful for patients with increased oral secretions, but overuse might lead to the drying of lower airways secretions, thus making for a more ineffective cough by thickening the secretions and making it more difficult to clear them from the airways. If used, one should closely observe the patient for this complication. Injections of botulinum toxin (Botox [Allergan Inc, Irvine, CA]) offer the advantage of not being a general drying agent, but its use with DMD has not been established.

Finally, any factor that contributes to increased airway secretions and/or altered airway clearance needs to be addressed. Many patients with DMD may be obese secondary to inadequate energy expenditure in light of a normal caloric intake.²⁸ The increase in chest wall mass from obesity further increases the load against which the already weakened respiratory muscles must work, leading to further muscle fatigue and a worsened cough.^{5,29} Chest wall compliance is further altered by obesity, making for a more ineffective cough, as described earlier. On the other extreme, malnutrition secondary to inadequate caloric intake decreases muscle mass and leads to a diminished cough.^{5,17,28} Aspiration (of normal oral secretions, food, and/or liquids) is frequently seen secondary to impaired swallowing^{27,29} and can further worsen any underlying lung disease by adding to the burden of a weakened cough.²⁷ Gastroesophageal reflux can lead to increased risk for aspiration²⁹ and can be worsened by obesity as well as scoliosis. Thus, the physician caring for the patient with DMD needs to help the patient maintain adequate caloric intake in a safe-as-possible manner (by either mouth and/or gastrostomy tube feedings) while treating any gastroesophageal reflux or impaired swallowing of oral secretions that may be present.

TREATMENT PLAN FOR ACUTE ILLNESS

When a person is infected with a viral illness, increased airway secretions develop as part of the body's innate defense system to treat this infection. For patients with an intact cough, this increased volume of secretions is easily handled and cleared. For patients with neuromuscular weakness and a resultant diminished cough, however, this increased volume of mucus can quickly overwhelm the host's ability to clear the secretions. This can then lead to the development of airway obstruction, atelectasis, ventilation-perfusion mismatch, secondary bacterial superinfection, muscle fatigue, and, ultimately, respiratory failure.²² Therefore, it is imperative to help the patient clear this increased airway burden as quickly as possible.

Assisted coughing maneuvers have been demonstrated to be an effective aid to clearing secretions. Tzeng and Bach²³ developed a protocol using pulse oximetry in combination with the CoughAssist machine to maintain airway patency and prevent respiratory compromise. Use of the CoughAssist machine when the patient is asymptomatic and has a normal CPF and/or MEP is not

routinely required; however, for patients in whom the CPF is <270 L/minute and/or the MEP is <60 cm H₂O at baseline, maintenance use of the CoughAssist machine on an at least once- to twice-daily basis would be a reasonable intervention to promote good skill levels for when increased therapy is warranted. For patients with acute viral illness, increased secretions, and normal oximetry readings, using the CoughAssist machine as needed, on the order of every 3 to 4 hours, would represent an appropriate treatment plan. If the symptoms increase and/or the SaO₂ decreases to <95%, increasing the frequency of assisted coughing is then necessary to maintain airway clearance. The CoughAssist machine may be used as often as every 10 to 15 minutes, as the patient tolerates. Once oxygen levels normalize and the patient clinically improves, the frequency of assisted coughing can be decreased. At no time should supplemental O₂ be used in the outpatient setting to maintain normal oxygenation; supplemental O₂ should only be used in the inpatient setting under close supervision.²³ Good hydration should be maintained to prevent drying of secretions (which may then prove more difficult to remove). Suctioning may be required to help the patient expectorate any produced sputum.^{18,23}

Patients with DMD also may have impaired ventilation, especially when they are acutely ill.^{6,7} Assisted ventilation, preferably by a noninvasive route, may be necessary during an acute illness.^{6,30} This might be just while asleep or, in more severe cases, both while awake and asleep. Patients already using noninvasive ventilation when asleep may transiently require increased nocturnal support as their condition initially worsens. Hopefully, with adequate airway clearance, the patient will return to his or her baseline state once the illness passes.

Bronchodilators such as albuterol or levalbuterol may be used in situations where airway hyperreactivity is present, but their efficacy for routine use has not been confirmed. Chest physical therapy can also be used in conjunction with bronchodilators and the CoughAssist machine on a case-by-case basis. The use of routine antibiotics is controversial and should be tailored to the needs of the individual patient.

When patients with significant neuromuscular weakness have an acute illness, their swallowing function can often transiently decompensate.²⁷ Many patients with DMD have altered swallowing function to begin with, but through compensatory measures they are able to take in adequate calories for weight maintenance without aspiration. When their work of breathing increases with illness, however, these compensatory measures may fail, putting the patient at increased risk for aspiration. Close observation of their ability to handle nutritional and fluid requirements needs to be undertaken until they return to baseline.

CONCLUSIONS

Patients with DMD, as well as other etiologies of neuromuscular weakness, are at risk for both acute and, ultimately, chronic respiratory failure from a combination of hypoventilation (brought on by muscle weakness) and ineffective airway-secretion clearance (caused by an

inadequate cough). Although there is not yet a cure for DMD, there are a wide variety of supportive interventions available that can aid with the clearance of airway secretions. It is important to have appropriate equipment in the home and have the caregivers acclimated to its proper use so that when an acute illness develops, they will be able to quickly intervene to prevent respiratory decompensation. Although the equipment chosen should be tailored to the needs of the individual family, a pulse oximeter and the CoughAssist machine would represent a useful starting point.

Early intervention at the first sign of respiratory illness can help prevent worsening of the patient's clinical status and prevent hospitalizations. It is important that patients with DMD be followed by physicians who have familiarity with this equipment and are knowledgeable in their use to provide the patient with the optimal resources for care to improve both the quantity and quality of their lives.

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