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Mini-Symposium: Cardio-Respiratory considerations in CNMD

Airway clearance modalities in neuromuscular disease

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SUMMARY

Airway clearance consists of two linked processes: mucociliary clearance and cough clearance. Patients with neuromuscular weakness are at risk for impaired cough clearance and therefore the development of pneumonia and atelectasis. Aiding airway clearance in the patient with neuromuscular weakness is critical to the maintenance of health and the prevention of significant respiratory morbidity. This can be achieved using both manual and mechanical techniques. This review will discuss the physiology of cough and the mechanics of aiding cough clearance in the patient with neuromuscular weakness. In addition, technologies and techniques used to improve mucociliary clearance will also be discussed. Newer technologies such as mechanical insufflation-exsufflation have gained widespread acceptance in the management of airway clearance in the patient with neuromuscular weakness.

However dangerous diseases of the chest may be, they are, nevertheless, more frequently curable than any other severe internal affection.
RTH Laennec, from *A Treatise on the Diseases of the Chest*, 1821

INTRODUCTION

Airway clearance, also referred to as secretion clearance or mucus clearance, is a critical aspect of respiratory care and when impaired represents a life-threatening aspect of many diseases. Airway clearance consists of two related processes: mucociliary clearance and cough clearance. The mucociliary "escalator" requires functioning cilia on the airway epithelium as well as a normal periciliary fluid layer. Diseases in which there is impairment of mucociliary clearance include cystic fibrosis [CF], primary ciliary dyskinesia [PCD], bronchiectasis and chronic obstructive pulmonary disease [COPD]. In patients with neuromuscular disease there can be impairment of cough clearance, despite the presence of normal mucociliary clearance (Table 1). As a result, there is a disproportionately high degree of respiratory disease in neuromuscular patients who have involvement of the respiratory musculature. This article will review modalities of therapy available to assist airway clearance in the patient with neuromuscular weakness.

PHASES OF COUGH

Coughing is a complex process.[1] It has been described as having 4 phases, although it can be argued that the process has 5 steps. The phases of coughing are as follows (Fig. 1):

1. **Irritation**: The cough is triggered by a chemical or physical irritant in the tracheobronchial tree. There are many irritant sensors in the airway (nasopharynx and tracheobronchial tree).
2. **Inspiratory phase**: irritation triggers inspiration through a widely opened glottis (inhalation volume varies). Length-tension relationship in respiratory muscles is optimized; elastic recoil potential is maximized. Air enters alveoli behind the secretions.
3. **Glottic closure**: necessary for the subsequent phases.
4. **Compressive phase** (200 msec): Expiratory muscles compress thoracic cavity, increasing intrapleural pressure rapidly to as high as 300 mm Hg.
5. **Expulsive phase**: The glottis is opened, releasing pressure, shearing secretions from airway walls and moving secretions cephalad. This may be interrupted by a short series of glottic closures, each with compressive and expulsive phases. When this occurs during a maximal expiratory flow volume curve manoeuvre, a series of "spikes" can be seen, with supramaximal flows over the MEFV curve.[2] These spikes are referred to as flow transients, and probably result from compression of the central airways (Fig. 2). An absence of flow transients has been associated with a loss of an effective cough.[3]

The "equal pressure point" limits flows. In patients with collapsible airways, the equal pressure point can be quite proximal thereby further limiting cough clearance.

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Table 1
Neuromuscular Diseases that Can Result in Respiratory Impairment (adapted from Bach, *Non-invasive Mechanical Ventilation*, Hanley & Belfus, 2002)

MYOPATHIES
The dystrophinopathies (Becker, Duchenne)
Other muscular dystrophies (Congenital, Fascio-scapular humeral dystrophy, limb-girdle, Emery-Dreifuss, myotonic dystrophy)
Other myopathies: acid maltase deficiency, acid alpha glucocidase deficiency, mucopolysaccharidoses, mitochondrial myopathies
Inflammatory myopathies: polymyositis
Diseases of the myoneural junction (myasthenia gravis, congenital myasthenia syndromes, mixed connective tissue disease
Myopathies of systemic disease, medications
NEUROLOGIC DISORDERS
Spinal Muscular Atrophy (Types I, II, III)
Motor neuron diseases (ALS, PLS, PMA)
Poliomyelitis
Charcot-Marie-Tooth
NEUROPATHIES: Charcot-Marie-Tooth, Phrenic nerve palsy/injury, Guillain-Barré, others
Multiple Sclerosis
Spinal cord injury
Spina Bifida
Cerebral palsy, static encephalopathies

All 5 steps of coughing can be affected by neuromuscular disease. Patients with profound encephalopathy, for example, may lack irritant receptors and have an absence of coughing and airway protective mechanisms. Patients with isolated weak inspiratory muscles (diaphragm paresis, palsy) as well as those with diffuse muscle weakness can have impaired inspiratory phase. Glottic closure can be impaired in bulbar involvement with motor neuron disease, late in Duchenne muscular dystrophy (DMD), and prevented in patients with tracheotomies. Impairment of the compressive phase is present in those patients who cannot close the glottis as well as those with weak accessory muscles of expiration (chiefly these are the rectus abdominus, obliques, and internal intercostals). The expulsive phase is affected by all the above factors.

IMPROVING MUCOCILIARY CLEARANCE

It should be first stated that most patients with neuromuscular disease have normal mucociliary clearance at baseline. Occasional patients with neuromuscular disease (NMD) will have chronic aspiration, which can lead to a chronic mucopurulent bronchitis state with impaired cilia. Chronic retention of secretions can also damage cilia secondary to the inflammatory cascade induced by white blood cell products. It is for these reasons that aiding mucociliary clearance can at times be helpful in the setting of acute disease in the NMD patient. Secretion clearance can be aided by traditional, manual chest physiotherapy (also called percussion and postural drainage). Other means of secretion mobilization include high frequency chest wall compression (The Vest and others) and intrapulmonary percussive ventilation (IPV). This latter device uses small bursts of air at 200 to 300 cycles per minute along with entrained aerosols delivered through a mouthpiece. Both IPV and high frequency chest wall oscillation are techniques that are effort independent. Most airway oscillation devices (Acapella, Flutter, and others), which deliver oscillating PEP (positive airway pressure) are effort dependent and as such are generally ineffective in the very weak neuromuscular patient.

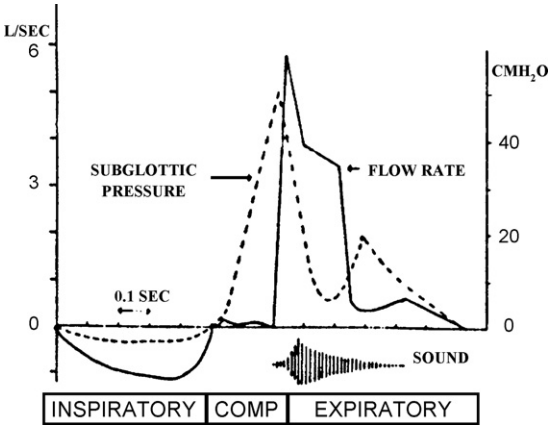

Figure 1. Phases of cough. (From McCool, 2006).

IPV has been studied for years in various disease states including CF and in the neuromuscularly weak patient, especially in the setting of atelectasis. Birnkrant et al (1996) demonstrated resolution of atelectasis with the use of IPV in a small case series.[4] They found that the IPV treatment resulted in clinical and radiographic improvement in three of four patients; although one patient experienced a third-degree atrioventricular block and hypoxemia. In a larger case series, Deakins and Chatburn (2002) demonstrated that IPV could be used safely in paediatric patients and was efficacious in treating atelectasis.[5] Reardon et al. (2005) published a randomized, prospective, controlled trial demonstrating a significant benefit with IPV use in patients with NMD at a residential facility (ages 11-19).[6] They showed decreased antibiotic use, school absenteeism and rates of hospitalization. The control group used incentive spirometry, which may not be an appropriate therapy for a patient with weak muscles, however.

High frequency chest compression (HFCC) (The Vest™ and others) has been widely advocated as an adjunctive therapy for patients with neuromuscular disease. At this time there is a dearth of literature in support of this therapy for patients with NMD, despite a wide acceptance for use of this device in cystic fibrosis. A single report on its use in patients with amyotrophic lateral

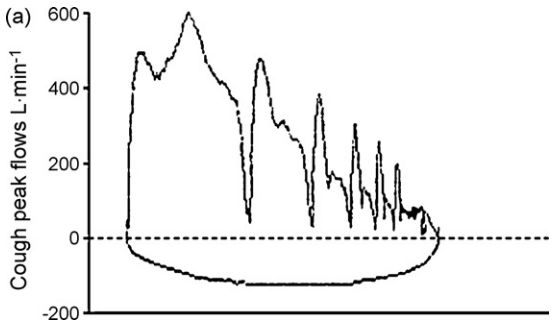

Figure 2. Flow transients on spirometry: "cough spikes" (from Chaudri, Eur Respir J 2002;19:434–438).