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Review article

Airway clearance techniques in neuromuscular disorders: A state of the art review

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ABSTRACT

This is a unique state of the art review written by a group of 21 international recognized experts in the field that gathered during a meeting organized by the European Neuromuscular Centre (ENMC) in Naarden, March 2017. It systematically reports the entire evidence base for airway clearance techniques (ACTs) in both adults and children with neuromuscular disorders (NMD). We not only report randomised controlled trials, which in other systematic reviews conclude that there is a lack of evidence base to give an opinion, but also include case series and retrospective reviews of practice. For this review, we have classified ACTs as either proximal (cough augmentation) or peripheral (secretion mobilization). The review presents descriptions; standard definitions; the supporting evidence for and limitations of proximal and peripheral ACTs that are used in patients with NMD; as well as providing recommendations for objective measurements of efficacy, specifically for proximal ACTs. This state of the art review also highlights how ACTs may be adapted or modified for specific contexts (e.g. in people

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with bulbar insufficiency; children and infants) and recommends when and how each technique should be applied.

1. Introduction

In healthy individuals, mucociliary clearance and cough mechanisms are effective and efficient in defending against secretion encumbrance, but these mechanisms may become ineffective if the systems malfunction and/or in the presence of excessive bronchial secretions. Mucus is transported under normal circumstances from the lower respiratory tract into the pharynx by cephalad-bias airflow and the mucociliary escalator mechanism [1].

An effective cough is essential to clear airway secretions from the more proximal airways [2]. For an effective cough one needs firstly to take a sufficiently deep breath in; the glottis needs to close briefly to allow an increase in intrathoracic pressure; followed by expulsive glottic opening together with abdominal contraction, which results in air being forcibly expelled [3]. This cough expiratory airflow can be measured and is known as peak cough flow (PCF). Individuals with weak or impaired inspiratory and/or expiratory muscles, with or without glottis closure issues (bulbar insufficiency, tracheostomy), will have decreased PCF.

Weakness of the inspiratory muscles leads to a progressive decrease in vital capacity (VC), but the lung volume changes that appear in some patients with neuromuscular disorders (NMD) are attributable to a combination of muscle weakness and alterations of the mechanical properties of the lungs and chest wall [4]. Reduced ability to cough leads to secretion retention, predisposing to progressive respiratory morbidity. Severe bulbar dysfunction and glottic dysfunction most commonly occurs in patients with amyotrophic lateral sclerosis (ALS), spinal muscle atrophy (SMA) type 1, other rarer neuromuscular disorders such as x-linked myotubular myopathy and pseudobulbar palsy of central nervous system etiology [5]. Inability to close the glottis and vocal cords results in complete loss of the ability to cough and swallow. Difficulty swallowing liquids may result in pooling of saliva and mucus in the pharynx, especially in the valleculae and the pyriform sinuses. This results in the perception of excessive pharyngeal secretions, similar to post-nasal drip [6].

Alterations in alveolar ventilation, atelectasis, mucus plugging, and recurrent respiratory tract infections (RTI's), as a consequence of an

ineffective cough; together with severe bulbar dysfunction, are the main causes of morbidity and mortality in patients with NMD [7–9]. Recurrent RTI's lead to further respiratory muscle weakness, with a resulting vicious cycle of respiratory disease [10,11]. Hypoventilation and managing secretions are amongst the most important problems from patients' perspective [12] and present the respiratory physiotherapist with unique management challenges in the care of people with NMD. Despite the clear implications, the problem of managing secretions has received little attention in the care of patients with NMD.

Patients with NMD's are living longer [13–15]; and consequently we are seeing more complex ventilator dependent and independent patients. Respiratory physiotherapy is an essential part of the multi-disciplinary management of these individuals, but owing to the inherent heterogeneity of the condition; the growing number of available airway clearance techniques (ACTs) and associated technological developments, it is challenging for physiotherapists to understand what assessments are required and what treatment options are available and appropriate for people with NMD.

As in other chronic disorders, the home organization of patients with chronic respiratory disorders is challenging and time consuming. The cost and availability of respiratory experts in primary care, the geographical location of patients, lack of engagement of general practitioners (GP) and care coordination may lead to poor care quality and organization. Project “Leonardo” investigated the impact of a new care organization that included a partnership between patients, considered here as key members of their own health team, their GP and their dedicated care coordinator. This study suggested positive effects in terms of increasing patient health knowledge and autonomy, improved care collaboration, appropriate resource utilization and readiness to make changes in health behaviours. A similar project worth's investigation in the respiratory care of patients with NMDs [16].

In this state of the art review written by an expert group during the 228th European Neuromuscular Centre (ENMC) international workshop on ACT's in NMD, we aim to define ACTs using simple, common language to help patients and all members of their care team. ACTs will be classified into proximal (cough augmentation) and peripheral (secretion mobilizing) ACTs (Fig. 1). We further aim to provide standard

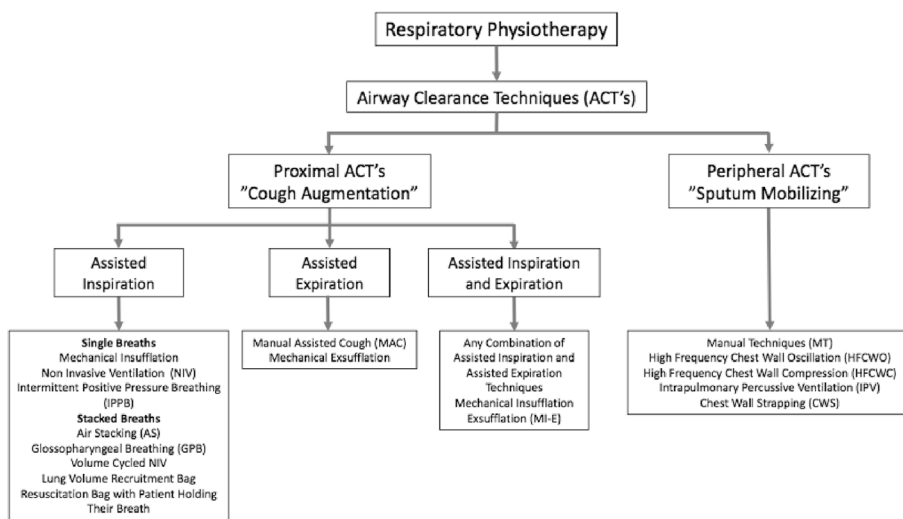


Fig. 1. Shows the classification of airway clearance techniques that are used in individuals with neuromuscular disease. Single breaths are defined as a breath given to the individual by the device followed by them exhaling or coughing. Stacked breaths are defined as multiple breaths in without the individual breathing out after the inspiration. Once the individual is close to their total inspiratory capacity they can actively or passively exhale, or cough. Mechanical insufflation is defined as an insufflation only provided by a positive pressure device. Mechanical exsufflation is defined as an exsufflation only provided by a mechanical insufflation-exsufflation (MI-E) device.

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