Mechanical insufflation–exsufflation for an individual with Duchenne muscular dystrophy and a lower respiratory infection

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Abstract
Duchenne muscular dystrophy (DMD) is an X-linked recessive myopathy associated with progressive muscle weakness and wasting, loss of ambulation, respiratory insufficiency, weak cough, repeated respiratory infections, and ultimately death from respiratory failure in early adulthood. Mechanical insufflation–exsufflation (MI-E) devices, such as the CoughAssist®, are used in individuals with neuromuscular weakness to augment cough and help clear secretions; however, there is no consensus on the optimal treatment regimen. We present the challenging case of a patient with DMD admitted to a tertiary hospital with a severe lower respiratory infection, and discuss how MI-E was used by physiotherapists to assist secretion clearance and prevent further deterioration.

Introduction

Duchenne muscular dystrophy (DMD) is an X-linked recessive myopathy associated with progressive skeletal muscle weakness and wasting, including those that assist respiration. This results in loss of ambulation, thoracic cage abnormalities, respiratory insufficiency, weak cough, repeated respiratory infections, and ultimately death from respiratory failure in early adulthood [1].

Mechanical insufflation–exsufflation (MI-E) devices, such as the CoughAssist® (Emerson, Cambridge, MA, USA), are used in individuals with neuromuscular weakness to increase inspiratory lung volumes prior to cough, and improve peak cough flow (PCF). Manual techniques are often added to further enhance PCF and secretion clearance [2].

Despite the known benefits, there is no consensus regarding the optimal MI-E treatment regimen [3]. This is in part due to the importance of tailoring the treatment to ensure individual tolerance, while still optimizing cough effectiveness. We present the challenging case of a patient with DMD admitted to a tertiary hospital with a severe lower respiratory infection, and discuss how MI-E was used to assist secretion clearance and prevent further deterioration.

Case Report

A 23-year-old male with DMD presented with increasing shortness of breath due to a severe lower respiratory tract infection (Fig. 1). At admission to the respiratory high dependency unit, he had type 2 respiratory failure (arterial blood gas pH 7.26, PaCO2 77.3, PaO2 68.9, HCO3 34.2), tachypnoea (respiratory rate (RR) 40 per minute), a low grade fever (37.6°C), and elevated C-reactive protein (CRP, 130 mg/L). Prior to admission, he was wheelchair-dependent, and used nocturnal non-invasive positive pressure ventilation (NIV) (inspiratory positive airway pressure
(IPAP) 20 cmH2O, expiratory positive airway pressure (EPAP) 10 cmH2O, and a CoughAssist device at home. The CoughAssist insufflation and exsufflation pressures used at home were low (10 and −10 cmH2O) and likely inadequate. This was his first admission for respiratory infection in spite of known pharyngeal dysphagia.

Medical management included intravenous antibiotics (IVAB) and continuous NIV with increased pressure support (IPAP 24 cmH2O and EPAP 10 cmH2O) with humidification, and arterial blood gases normalized over the next 3 days. A speech therapist provided ongoing assessment of the dysphagia. Initial physiotherapy assessment found reduced breath sounds bibasally and coarse inspiratory crackles on auscultation, in conjunction with a rapid, shallow breathing pattern and poor chest expansion bilaterally. The patient’s cough was weak, moist, and ineffective, and he was unable to clear secretions independently. SpO2 was 95% on supplemental O2 3 L/min. While not available in this instance, other measures that could improve clinical assessment include vital capacity, sniff nasal inspiratory pressure, PCF, and baseline cardiac function.

Physiotherapy management involved the use of the CoughAssist between two and four times per day. Our MI-E regimen consisted of three to five cycles of between three and five breaths with insufflation pressure of 25 cmH2O and exsufflation pressure of −30 cmH2O. Insufflation and exsufflation were each 3–4 sec in length and pressures were titrated to the individual’s tolerance. A thoracic compression was combined with the exsufflation-assisted cough. A second therapist often assisted in order to combine manual techniques with MI-E. Rest periods were provided on NIV between cycles for 2–5 min, followed by thoracic expansion exercises (TEE) and expiratory vibrations of the chest wall delivered in combination with NIV. Positioning was limited to upright sitting with tilting to the left and right due to the patient’s multiple lower limb contractures. Treatment duration was usually between 30 and 60 min, guided by the patient’s fatigue and comfort.

Blood pressure was measured before each treatment, and SpO2 and heart rate (HR) were continuously monitored throughout treatments to ensure clinical stability. Auscultation and subjective work of breathing were also used to monitor treatment effectiveness. Copious amounts of secretions were cleared with each MI-E treatment in the first week, and secretion volume decreased throughout the second week. The frequency of treatment sessions were reduced progressively until only one treatment per day was required. By day 15 the patient’s cough was subjectively stronger with minimal secretion production, there was near complete resolution of the acute changes on chest radiography (Fig. 1), and no supplemental oxygen was required. Further inpatient stay was required to reduce NIV dependency, and the patient was discharged home on day 27.

**Discussion**

This case highlights the role of MI-E, in combination with manual techniques, for improving cough and clearing secretions in an individual with DMD admitted to hospital with a severe lower respiratory infection. The challenges in
this presentation also raise ideas on how the regimen could be altered to optimize treatment effectiveness.

Our MI-E regimen consisted of three to five cycles of between three and five breaths, with pressures of 25 cmH2O on insufflation and −30 cmH2O on exsufflation. Number of breaths per set and total sets were adjusted according to clinical presentation, tolerance, and fatigue during each session. Despite receiving between two and four physiotherapy sessions per day, it took 2 weeks for the respiratory infection to resolve. Two possible reasons for this are the lower than optimal MI-E pressures used due to pressure intolerance by the patient and the inability to greatly vary positioning during treatment sessions. Another factor that may have impeded recovery was dysphagia and potential ongoing aspiration early in the admission. While the primary aim of the intervention was to increase inspiratory lung volumes to aid secretion clearance and cough, the maximal insufflation pressure tolerated was not much higher than the IPAP used on NIV at rest. Clinical studies have found insufflation and exsufflation pressures of up to 40 cmH2O and −40 cmH2O to be optimal in adults [4], and optimized pressures in the home setting may have enabled the respiratory infection to be treated effectively prior to admission to hospital. However, the risk of complications associated with high positive airway pressures, such as abdominal distension, aggravation of gastroesophageal reflux, haemoptysis, chest and abdominal discomfort, acute cardiovascular effects, and pneumothorax, should also be considered [5]. As further increases in insufflation pressures were not tolerated, a gradual increase in IPAP on NIV may have further improved secretion clearance.

This report provides physiotherapists with an example of how MI-E can be used and optimized to successfully treat individuals with DMD, presenting with lower respiratory infections.

Disclosure Statements
No conflict of interest declared.
Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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