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Mechanical insufflation–exsufflation for airway clearance in adults with cystic fibrosis

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Abstract

In cystic fibrosis (CF), acute exacerbations can decrease the effectiveness of patients' usual airway clearance techniques (ACT). In order to maintain effective airway clearance and preserve lung function, these ACT must be adapted to prevent further dyspnoea and fatigue and improve ease of expectoration. Mechanical insufflation–exsufflation (MI-E) is widely used in neuromuscular disorders to facilitate airway clearance and augment cough but has rarely been used in CF despite potential indications. The NIPPY Clearway, an airway clearance device with multiple modes including MI-E, can be set to deliver multiple insufflations prior to a single exsufflation. We present two cases where this modified version of MI-E was used as an adjunct to traditional ACT in adults during an acute exacerbation of CF.

Introduction

Cystic fibrosis (CF) is a life-limiting autosomal recessive disease, with the major cause of morbidity and mortality being severe airway obstruction secondary to bronchiectasis. Traditional airway clearance techniques (ACT) during an acute exacerbation, while essential, have been associated with increased energy expenditure, oxygen desaturation, respiratory muscle fatigue, and dyspnoea [1,2].

Mechanical insufflation–exsufflation (MI-E) is a treatment widely used for cough augmentation and airway clearance in neuromuscular conditions due to severe respiratory muscle weakness and reduced peak expiratory flow (PEF). Despite potential indications for MI-E in CF, its use is limited.

The NIPPY Clearway (B & D Electromedical, England) is a novel airway clearance device with multiple modes, including MI-E. The patient-triggered Programmable Auto Mode can be set to deliver multiple insufflations prior to a single exsufflation, repeated in cycles. We present two cases where this modified version of MI-E was used as an adjunct to usual ACT in adults during an acute exacerbation of CF.

Case Report

A 29-year-old male with CF presented with an acute exacerbation and a percent-predicted forced expiratory volume in 1 s ($ppFEV_1$) of 44, reduced from his recent best of 81. At admission, his oxygen saturation (SpO_2) was 94% on room air (RA), and his chest X-ray (CXR) showed diffuse bilateral bronchiectatic changes and left middle and lower zone infiltrates. He was poorly compliant with home treatment, demonstrated an ineffective forced expiratory technique, had an irritable cough, and found it difficult to expectorate sputum.

To facilitate airway clearance, we used modified MI-E with a mouthpiece interface. A nebulizer was incorporated into the circuit to allow for the inhalation of 6% hypertonic saline (HTS) during treatment. The NIPPY Clearway was set to deliver five 2.5-s insufflations prior to a single 3-s exsufflation. This was repeated five times consecutively for a total cycle time of approximately 3 min. Pressures were increased according to comfort, reaching a final insufflation pressure of 20 cmH_2O and an exsufflation pressure of −30 cmH_2O. The insufflation time was set to the slowest ramp over the longest time tolerable to

promote collateral ventilation. During the exsufflation time, the patient was instructed to gently huff into the mouthpiece. On completion, he would expectorate, rest for 2 min, and repeat the modified MI-E cycles until he felt clear – which typically took about 30 min. During treatment, SpO_2 and heart rate were monitored to ensure clinical stability. After using modified MI-E daily during his 14-day admission, his $ppFEV_1$ improved to a new 12-month best of 87.

The same modified MI-E technique was trialled in a patient with more severe respiratory disease. She was a 38-year-old woman with CF admitted with an acute exacerbation, with a $ppFEV_1$ of 25, reduced from a baseline of 35. She reported marked increased dyspnoea at rest, myalgia, and increased sputum and was unable to perform her usual ACT effectively. Her SpO_2 was 90% on RA, and CXR revealed advanced bronchiectatic changes.

Modified MI-E was used with the aim of reducing dyspnoea and increasing ease of expectoration. Pressures were titrated from initial low pressures (insufflation of 6 cmH_2O and exsufflation of −4 cmH_2O) to a final insufflation pressure of 12 cmH_2O and an exsufflation pressure of −12 cmH_2O. She completed seven 2-s insufflations followed by a single 1.8-s exsufflation, during which she would huff into the mouthpiece. This cycle was repeated twice for a total cycle time of approximately 1 min before expectorating. After 1 min of rest, she would complete a set of her usual ACT (Flutter® Aptalis Pharma, Bridgewater, NJ, USA with a mask) to continue to clear secretions, prior to repeating modified MI-E. Subjectively, she reported increased ease of expectoration, reduced fatigue, and less dyspnoea when completing ACT. No adverse events occurred during or following modified MI-E, and at the time of her discharge, her $ppFEV_1$ improved to 38. She has continued using modified MI-E as an adjunct to usual ACT during all subsequent admissions.

Discussion

In CF, acute exacerbations can decrease the effectiveness of usual ACT due to fatigue, dyspnoea, or a high sputum load [3]. It is therefore necessary to adapt techniques to maintain effective airway clearance and preserve $ppFEV_1$. As an established adjunct to traditional ACT in moderate to severe CF, non-invasive ventilation (NIV) has been demonstrated to improve respiratory muscle strength and oxygen saturation while reducing dyspnoea [4]; however, NIV is unable to provide exsufflation.

MI-E is indicated in any patient with sputum retention who is unable to cough or clear secretions effectively due to respiratory muscle weakness or reduced PEF [5]. During CF exacerbations, a high sputum load is coupled with dyspnoea and hyperinflation, compromising effective respiratory muscle function, leading to reduced PEF [3]. Modified MI-E has the potential to overcome these barriers and assist airway clearance.

Modified MI-E facilitates airway clearance by providing multiple insufflations at pressures high enough to increase inspiratory lung volumes, promoting collateral ventilation by overcoming collapsed or blocked small airways. These insufflations were delivered over the longest time tolerable to compensate for asynchronous ventilation caused by sputum retention or atelectasis. The addition of a huff during exsufflation facilitates the movement of secretions towards central airways for expectoration. During this time, each patient was monitored for signs of dynamic compression (indicated by wheeze), airway closure or obstruction, paroxysmal coughing, and oxygen desaturation as measured by SpO_2.

Insufflation and exsufflation pressures of up to 40 cmH_2O and −40 cmH_2O have previously been established as optimal in adults with normal lung compliance and airway resistance and in neuromuscular disorders [5]. However, previous studies have also reported good outcomes with low pressures. We felt that higher pressures may not be tolerated in these patients; therefore, lower pressures were trialled and increased according to comfort. High positive pressures may also be associated with an increased risk of barotrauma and haemoptysis.

The use of a mouthpiece enabled greater patient control over the treatment and allowed each exsufflation to be combined with a huff. Both patients preferred using modified MI-E over NIV, finding it an energy-efficient and effective ACT.

The presented cases discuss a novel treatment not previously reported in CF. MI-E can be considered an adjunct to traditional ACT for adults with an acute exacerbation of CF.

Disclosure Statement

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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