

## Case Report

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# Intensive Management of Chronic Bronchorrhea in a Tracheostomized Duchenne Patient with Bronchiectasis

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## Abstract

Duchenne muscular dystrophy (DMD) is a severe X-linked hereditary neuromuscular disease characterized by progressive muscle degeneration and functional tetraplegia. Advances related to the use of mechanical ventilation and techniques of airway clearance have improved the lifespan of these patients until 30-35 years. In a 27 years old home-ventilated tracheostomized DMD patient affected by bronchiectasis and atelectasis with excessive bronchorrhea (>300ml mucus production/24h), endotracheal suctioning act became so frequent that it severely affected his mental health and that of his family. An intensive programme of airway clearance including ventilation via Intrapulmonary Percussive Ventilation (IPV) and suctioning via mechanical insufflation-exsufflation (MI-E) associated with strong manual chest compression improved mean nocturnal SpO<sub>2</sub> from 91 to 95% and reduced mucus production and suctioning rate by 50%. The perceived physical and emotional health (SF-36) increased in the patient and in his family. In the patient, the general mental health score increased from 16 to 48%, the vitality from 10 to 40% and the general health perception from 10 to 35%. In the patient's father, the social functioning improved from 38 to 63%, the general mental health from 36 to 60% and the vitality from 35 to 60%. In this report, intensive respiratory therapy with IPV in combination with MI-E and chest compression was safe and spectacularly improved the quality of ventilation, the mucus production and the perceived mental health in an adult Duchenne patient with bronchorrhea.

**Keywords:** Chest physiotherapy; Cough-assist; Duchenne; Mechanical insufflation-exsufflation; Tracheostomy; Ventilation

## Introduction

Duchenne muscular dystrophy (DMD) is a severe neuromuscular (NM) disease, considered as the most common and rapidly evolutive muscular dystrophy. The incidence of this X-linked hereditary disease is approximately 1/3000 [1] to 3500 [2] male births. DMD is caused by mutation of the dystrophin gene which leads to the absence of dystrophin in muscle tissue [1]. Missing dystrophin precludes normal muscle contraction and leads to progressive muscle weakness. Muscle weakness typically leads to functional tetraplegia, the need for mechanical ventilation and techniques of airway clearance, and ultimately death.

Clinical diagnosis is mostly made at the age of 3 to 8 years after consideration of history, physical findings, and elevated serum creatine kinase level that reflects muscle degeneration [3]. With increasing age, DMD progressively affects the skeletal, respiratory and cardiac muscles [1]. When ambulation is lost, there is a need for an electronic wheelchair at the age of 7-12 years [4]. Vital capacity (VC) typically declines at a rate of 8.5 percent predicted value per years [5]. Hypercapnic respiratory failure is an inevitable complication of progressive NM diseases. Without ventilatory support and techniques of airway clearance, death is predictable around 20 years [1]. However, recent advances related to the use of respiratory aids have improved the lifespan of the patients with DMD by 10-15 years [6].

In adolescent and young adult DMD patients, overnight noninvasive intermittent positive pressure ventilation (NIPPV) via nasal mask is effective in providing 24h normocapnia during several years but needs to be extended with disease progression when diurnal dyspnoea [7] or ultimately hypercapnia [1] occurs. Non-invasive interfaces may be used for continuous assisted ventilation [8]. Some DMD patients however may require tracheostomy when bulbar function is severely altered. Other patients may undergo tracheostomy in the emergency room after acute respiratory insufficiency. Long-term intermittent positive pressure ventilation via tracheostomy is reported with good

acceptance in 2/3 of the DMD patients [9]. In spite of advantages such as safety and possibility of endotracheal suction, tracheostomy also puts patients in danger of side effects such as excessive mucus production, tracheal stenosis, tracheobronchomalacia, bleeding, chronic bronchitis and bronchiectasis [10,11].

Until now, few studies have assessed the effectiveness of the combination of mechanical ventilation and airway clearance techniques in DMD patients affected by bronchiectasis, a permanent dilatation of the bronchi with destruction of the elastic and muscular component of the bronchial walls. In the present case, a dramatic mucus hyperproduction aggravated by a severely impaired secretion clearance was treated in a 27 yrs old tracheostomized Duchenne patient affected by bronchiectasis. In the current case, bronchiectasis was probably related to prolonged use of invasive ventilation and recurrent aspirations [12]. In addition, impaired clearance of secretions caused chronic respiratory infection resulting in a vicious cycle of bronchial damage, bronchial dilation, impaired clearance of secretions, recurrent infection, and more bronchial damage. The mental health of this patient and that of his family had been destroyed with the challenging management of this bronchorrhea.

## Case Summary

We report here the original experience of aggressive respiratory therapy in a 27 year old patient with Duchenne muscular dystrophy

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presenting bronchiectasis with chronic bronchorrhea. In this patient, nocturnal NIPPV was initiated at 15.5 years. Lung function tests at this time were as follows (in % of predicted value): Vital Capacity (VC) was 1330 ml (25%), Maximal Inspiratory Pressure (MIP) was 39 cmH<sub>2</sub>O (29%) and the peak expiratory flow (PEF) was 136 L/min (19%). Nocturnal NIPPV allowed 24h normocapnia for five years. During an acute chest infection, tracheostomy was placed in the emergency room to provide permanent ventilation and to offer the possibility for airway clearance by endotracheal suctioning. As a consequence of the tracheostomy the patient had recurrent chest infections and was regularly hospitalized. Hypersecretion, aspiration and prolonged use of intermittent positive pressure ventilation via tracheostomy involved in severe bronchiectasis and bronchorrhea. Bronchiectasis was diagnosed *via* a Computed Tomography (CT) scan. This examination showed bronchial wall thickening and luminal dilatation and a complete atelectasis of the right lower lobe with a discrete air bronchogram. A right upper lobe and, to a lesser extent, a right lower lobe bronchiectasis were found with alveolar infiltrates of infectious origin.

The patient was primary treated at home by non instrumental chest physiotherapy and two weeks turning antibiotherapy (Amoxycillin + Clavulanic acid; Clarithromycin; Trimethoprim + Sulmameothoxazol). The patient lost 13 kg (from 42 to 29kg) within 7 years after tracheostomy and pulse oxymetry (SpO<sub>2</sub>) was less than 88% within 14% of the time with ventilation. At the age of 27, the family addressed to our Centre a rescue appeal for helping them in the management of the bronchorrhea. At this moment, no ventilator free time was available, VC was 80 ml (1.5%), MIP was 4 cm H<sub>2</sub>O (3.7%)



**Figure 1:** Use of mechanical insufflation-exsufflation associated with endotracheal suctioning.



**Figure 2:** Active manual chest compression during mechanical insufflation-exsufflation.

and PEF was 15L/min (2.7%). The impact of permanent hypersecretion and recurrent suction every hour largely interfered with the social activities of the parents. Mucus production was abnormally elevated, reaching more than 300 ml per 24h. This largely surpassed the most severe cases we had previously met.

A recent literature overview suggested that Intrapulmonary Percussive Ventilation (IPV) is effective to reduce peripheral airway encumbrance, to improve the quality of ventilation, and to reduce the time of hospitalisation and the quantity of antibiotics used in neuromuscular patients during acute chest infections [13]. Another literature overview suggested that Mechanical Insufflation-Exsufflation (MI-E) is effective to compensate for deficits of cough in patients with low respiratory muscle strength and to enhance mucus transport from the central airways to the throat [14]. In addition, a recent editorial suggested that using MI-E devices is possible *via* artificial airways [15]. Both therapies are deemed as well adapted in patients with poor physical collaboration. For this reason, we proposed an hospitalisation including a respiratory therapy program mixing IPV and MI-E to manage bronchorrhea, accepted by the patient and his family. We used IPV and MI-E as complementary techniques. IPV and MI-E were used to enhance peripheral and proximal airway clearance, respectively.

We started intensive chest physiotherapy during a 5 days program. The program included 9 hours of night-time high frequency percussive ventilation (VDR4®, Percussionaire, Sandpoint, USA). The parameters were as follows: high frequency signal: peak inspiratory pressure = 50 cm H<sub>2</sub>O (37 torr); peak expiratory pressure = 15 cm H<sub>2</sub>O (11 torr); rate = 550 min<sup>-1</sup>; low frequency signal: plateau inspiratory pressure = 50 cmH<sub>2</sub>O (37 torr); convective peak pressure = 78 cm H<sub>2</sub>O (57 torr); oscillatory CPAP = 8 cm H<sub>2</sub>O (6 torr); rate = 21.8 min<sup>-1</sup>. We started night-time high frequency percussive ventilation after a 2 hour monitoring of Transcutaneous CO<sub>2</sub> tension (TcCO<sub>2</sub>) to check that VDR4 did not cause hyperventilation. During the day, airway clearance was achieved by 4 sessions/day, 1 hour each, divided in sub-sessions of 10 minutes separated by 5 minutes rest. Airway clearance was obtained by the combination of 3 techniques, two non-invasive and one invasive at the same time. Mechanical Insufflation-Exsufflation (MI-E) display (Cough-Assist®, Emerson, Cambridge, USA) was directly connected on the canula of tracheostomy at an intermittent negative pressure of -40 cm H<sub>2</sub>O (-29 torr), with inspiratory and expiratory times of 2 seconds. In addition, invasive suction was achieved by a catheter placed into the canula through the ventilation tubing adapted with a suction hall in front of the canula (Figure 1). Initially controlled by endoscopy, the suction catheter was placed 4 cm above the carina avoiding any mechanical traumatism. Invasive suction by catheter was carried out only to avoid mucus invasion in MI-E tubing. This precaution meant that the therapist did not waste time cleaning out secretions in the tubing. Expiratory manual thoracic compression was achieved during the 10 minute sub-session by two strong hand compressions on the right and on the left ribs (Figure 2). Mucus collection surpassed 300 ml/day during the trial. After 5 days hospitalisation, the patient was discharged home with a Cough-Assist® and was asked to prolong therapy. Intrapulmonary percussive ventilation (IPV) was recommended during the day via a portable device for home use (Impulsator®, Percussionaire, Sandpoint, USA) for 3 periods of 15 minutes.

After one month therapy at home, the amount of secretions diminished to 150 ml/day, and mean nocturnal SpO<sub>2</sub> improved from 91% to 95% (SpO<sub>2</sub> mini from 82 to 94%). Measured at the time of the trial and 4 weeks later, the quality of perceived physical and emotional

health was assessed by the short-form health survey questionnaire (SF-36) for the patient and for his father who mostly looks after him. In the patient, the general mental health score increased from 16 to 48%, the vitality from 10 to 40% and the general health perception from 10 to 35%. In the patient's father, the social functioning improved from 38 to 63%, the general mental health from 36 to 60% and the vitality from 35 to 60%. The father found that the outcome of the situation had improved very satisfactorily.

## Discussion

In this study, the mucus production of an adult Duchenne affected by bronchiectasis diminished by 50%. In addition, the mean nocturnal SpO<sub>2</sub> improved from 91% to 95%. Finally, the general mental health and the vitality improved in both the patient and his father.

We report here an original, safe and effective way of MI-E use in a weak, hypersecretive and tracheostomized ventilated Duchenne patient. The goal of the present trial was to increase the time between two successive suctionings. The need for suction interfered so much with the father's social activities that he reported having actually no more social life out of the house, as evidenced by his low social functioning score before treatment.

Excessive mucus production is reported in neuromuscular patients receiving long-term ventilation with uncuffed tracheostomies [10]. Hypersecretion is considered as a common side effect of the tracheostomy [11] and is generally defined as a mucus production above 30ml per day. The present case had a singular massive mucus production. In the literature, no case of such a massive hypersecretion was previously reported as surpassing 300ml/day in neuromuscular disorders. We hypothesized that atelectasis and bronchiectasis, confirmed by CT scan, had probably structured and fed a vicious circle of mucus hypersecretion and stagnation. We logically aimed at breaking this vicious circle by (1) improving the quality of ventilation and (2) achieving intensive and effective mucus clearance. Techniques of intrapulmonary percussive ventilation (IPV) [16], mechanical insufflation-exsufflation (MI-E) [17] or chest compression [18] are reported to help in secretion clearance in neuromuscular disorders. MI-E without chest or abdominal compression was already described in invasive ventilation and MI-E seemed more effective in clearing out secretions than conventional tracheal suction [19]. The three techniques (IPV, MI-E, chest compression), however, were not previously associated.

As a result of the present trial, the quality of ventilation improved as assessed by SpO<sub>2</sub> improvement. Mucus production diminished by 50%. The general mental health improved both in the patient and in his father. According with a previous report [20], the delay between 2 suctionings rose from 1-2 hours to 5-9 hours which was deemed very satisfactory by the patient and his family. After initiation of the therapy, the father told that he could now spend complete nights without suctioning secretions for the first time in 7 years.

When airway clearance techniques are mixed, it is difficult to assess which part of the efficiency can be attributed to each technique. In the present case, we assume that the combination of MI-E and the thoracic compression both brought a significant part of the efficiency in clearing out secretions. In patients receiving mechanical ventilation, mechanical rib-cage compression during the expiratory phase is reported effective in airway-secretion removal [18]. Clearly, the efficiency of strong thoracic compression was here visually observed in real time by an immediate higher mucus collection. Moreover, mixing MI-E and chest compression was very well tolerated by the patient who felt it very soft. To our surprise, MI-E was useful in ventilating the patient during the respiratory care sessions.

In this report, intensive respiratory therapy achieved by non-invasive suctioning was safe, improved the quality of ventilation and reduced mucus production in a Duchenne patient with bronchorrhea. Moreover, suction frequency substantially diminished. This treatment improved the perceived mental health of the patient and that of his family.

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