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Persistent Pulmonary Atelectasis in Duchenne Muscular Dystrophy Treated with High Frequency Chest Wall Oscillation

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Abstract

Patients with neuromuscular disease that affects the respiratory pump can experience mild to profound limitation in both ventilation and cough. They frequently present recurrent pulmonary infections, respiratory failure, hypoventilation and atelectasis. We describe the case of a 22-years-old patient with Duchenne Muscular Dystrophy, he was referred to our hospital with persisten fever (>38°C), dyspnea and respiratory failure. Chest CT showed parenchymal consolidation. The patient was treated with antibiotics and rehabilitation therapy, bronchial endoscopy, cough assist, but only with the use of High Frequency Chest Wall Oscillation followed by mechanical in-exsufflation (MI-E) for cough assistance, we observed clinical and radiological improvement. We suggest that High Frequency Chest Wall Oscillation can be helpful for neuromuscular patient, particularly in cases of difficult clinical management hospedalizations.

Keywords: Oscillation; Ventilator; Neuromuscular; Hospedalizations

Introduction

Chronic and progressive neuromuscular disease can result in various degrees of ventilatory impairment, atelectasis and cough inefficiency. The effectiveness of cough clearance depends on the coordinated neural sequence of phases involved in the cough maneuver and the inspiratory, expiratory, and glottic muscle functioning necessary to produce sufficient intrathoracic pressure and expiratory gas velocity. In the inspiratory phase, inhalation to VC will produce the highest intrathoracic pressure, cough volume, and cough velocity. When neuromuscular-induced inspiratory muscle weakness limits the volume of air that can be inspired, expiratory muscle length tension and chest-wall recoil forces are limited, which limits intrathoracic pressure and expiratory flow and volume, which limits the airway linear airflow velocity, which limits secretion clearance in the expiratory phase [1].

Respiratory complications in neuromuscular diseases with ineffective cough are represented by the onset of pneumonia and impaired gas exchange with acute respiratory failure. Acute episodes of respiratory infection determine, also, an increase of bronchial secretions in patients with reduced muscle strength. These alterations can cause reduction in vital capacity and peak flow, which urgently need manual or mechanical assistance to cough [2].

Case Presentation

22 year old boy with to Duchenne Muscular Dystrophy (DMD), was referred to our hospital with persisten fever (>38°C), dyspnea, and greenish expectoration, after 30 days and two previous hospedalizations. He was in treatment with non invasive home mechanical ventilation with nasal mask, and had no history of recurrent pneumonia or increased frequency of upper airway infections. The patient had convex right kyphoscoliosis, with a Cobb angle of about 65° [3]; he had also dilated cardiomyopathy (DCM) with ejection fraction of 35%. The patient had weight loss of about 2 kg last month. The spirometry showed a Vital Capacity (VC) of 0, 46 lt (10% of theoretical). The peak cough flow was 140 L/min. Peripheral O_2 saturation was 82%. Arterial blood gas analysis showed normocapnia with hypoxemia and acid-base balance was normal. The microbiological examination of sputum showed a Staphilococco Aureus infection. The Chest CT showed "parenchymal consolidation of the lower lobe, in right lung" (Figure 1). Bronchoscopy with a bronchial toilet had a positive Staphilococco Aureus 1,000,000 colony forming unit, multidrug resistent. The search for Koch's bacillus was negative. Blood cultures were negative.

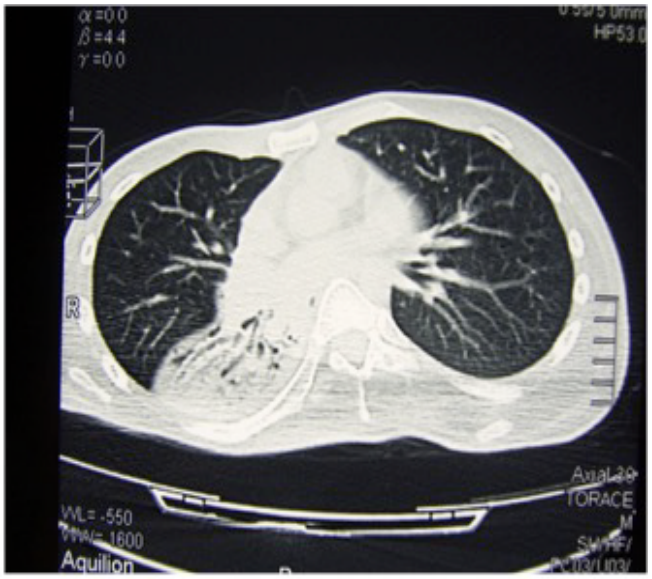

Figure 1: Chest TC before treatment.

The patient was treated with antibiotics (amikacin, vancomycin, piperacillin tazobactam, fluoroquinolones) and chest physiotherapy. He was also treated with manually assisted cough and mechanical in-exsufflation therapy. After another 26 days the patient was treated with antibiotics with no benefit: the fever and purulent sputum persisted. It was decided to undertake therapy with extrathoracic system to High-Frequency Oscillation (HFCWO) at pressure of 5 cm H_2O and a

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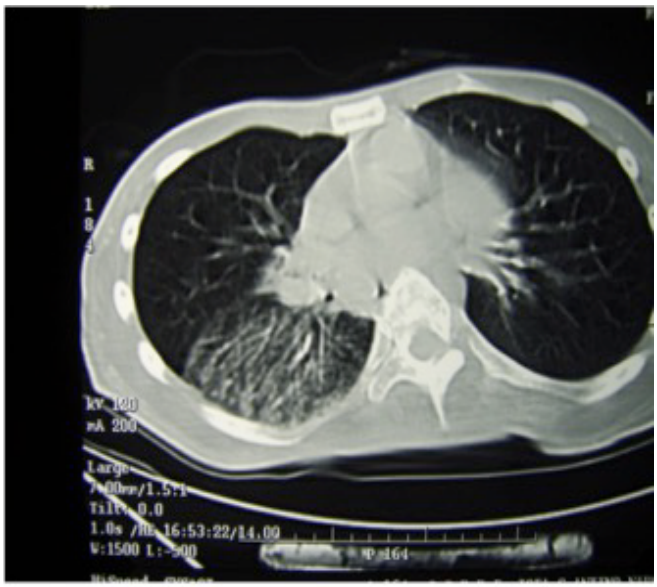

Figure 2: Chest TC after 20 days of treatment.

frequency of 10 Hz. The therapy was applied for 15 minute five times/day. The treatment was well tolerated and followed by five sessions of MI-E for cough assistance with an in-exsufflator at a pressure of +40/ -40 cm H_2O delivered respectively over 3 and over 2 seconds.

After 5 days of treatment there was evidence of an improvement of the clinical data and after 15 days chest CT (Figure 2) showed an almost complete resolution of parenchymal consolidation area. After a month of treatment was obtained a complete regression of the symptoms and an improvement of lung function. Last spirometry practiced after about 30 days of treatment showed a variation of VC from 0.46 lt (10% of theoretical) to 0.87 lt (23 % of theoretical); peripheral O2 saturation was 91 %. The peak cough flow was 180 L/min.

Discussion

Developing cough weakness in patients with neuromuscular disease will usually go unnoticed, as they have little or no need to cough, since the mucociliary system clears the normal daily mucus volume. When a respiratory infection occurs, a weak cough becomes the limiting factor in the patient's ability to maintain secretion clearance, and pulmonary congestion develops as the patient fatigues with the increased need to clear secretions. Despite the therapeutic support with chest therapy, manually and mechanical cough assist, non invasive mechanical ventilation and drug therapy, our patient did not show any improvement. After five days of treatment with high-frequency oscillations the patient presented improvement of the clinical data, resolution of symptoms and, after 30 days of treatment, a significant improvement with resolution of parenchymal lung atelectasis. Interventions for clearing the airway and improving ventilation in DMD are very important and pharmacological measures are often insufficient to improve health status. Similarly, mechanical in-exsufflators alone can be insufficient if secretions are difficult to mobilize. Chest physiotherapy with postural drainage can be often impractical for individuals with limited mobility and significant skeletal deformity, [4] as it requires assuming a posture that patients with severe scoliosis can hardly maintain. HFCWO assist devices generate either positive or negative trans-respiratory pressure excursions to produce high-frequency, small-volume oscillations in the airways. HFCWO can lead to changes in volume of 15-57 ml and in flow up to 1.6 L/s, which generate minimal coughing to mobilize secretions. The typical treatment lasts 20-30 minutes, and consists of short periods of compression at different frequencies, separated by coughing. HFCWO is becoming the method of choice for individuals with many types of disabilities [5]. It is applied through an inflatable vest attached by hoses to an air-pulse generator. Small volumes of gas are rapidly injected into and withdrawn from the vest, which pressurizes and releases the chest at frequencies from 5 to 25 Hz. This brings the patient to cough gently and to clear loosened secretions. HFCWO has been shown to be effective in clearing secretions from the lungs of patients with bronchiectasis, asthma, cystic fibrosis, primary ciliary dyskinesia, emphysema, COPD [6] and, in some recent study, in patients with Amyotrophic Lateral Sclerosis (ALS) [5,7]. There are few data yet about the different methods of drainage of secretions in neuromuscular patients with acute respiratory disease, and surely the treatment with High Frequency Chest Wall Oscillation can be a solution in cases of difficult clinical management.

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