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Temporary positive expiratory pressure (TPEP) as an alternative approach in the treatment of persistent atelectasis in a patient with Steinert disease: a case report

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We describe the clinical case of a patient affected by Steinert disease with persistent dyspnea complicated by a complete obstructive atelectasis of left lower lung lobe. The atelectasis has been successfully treated using the TPEP machine, with resolution of radiological pattern and improvement of the symptoms.

Introduction

Patients with neuromuscular pathologies may present difficulties in the management of secretions and sometimes dysphagia may contribute to the stagnation of secretions, especially in the sloping regions of the chest [1,2]. Respiratory complications in neuromuscular diseases with ineffective cough are represented by the onset of pneumonia and impaired gas exchange, with acute respiratory failure. In patients with muscular dystrophy, the altered relationship of tension in muscle length and the reduction in muscle elasticity and plasticity result in impaired inspiratory muscle function and a decrease in vital capacity. In addition, loss of deep breathing further increases the risk of alveolar collapse, hypoventilation and atelectasis, while acute episodes of respiratory infection determine an increase in bronchial secretions. These alterations can cause reduced values of vital capacity and peak flow, which urgently need manual or mechanical assistance to cough. A personalized rehabilitation treatment, carried out by an experienced team, can lead to clinical resolution.

Case description

A 44-year-old man, never smoker, affected by Steinert disease was admitted to the respiratory care unit of Monaldi hospital for persistent dyspnea.

The patient reported fever about one month earlier and worsening of dyspnea in last time. Admitted to the emergency room, the nasopharyngeal swab was negative for SARS-CoV-2. Chest x-ray revealed left low-

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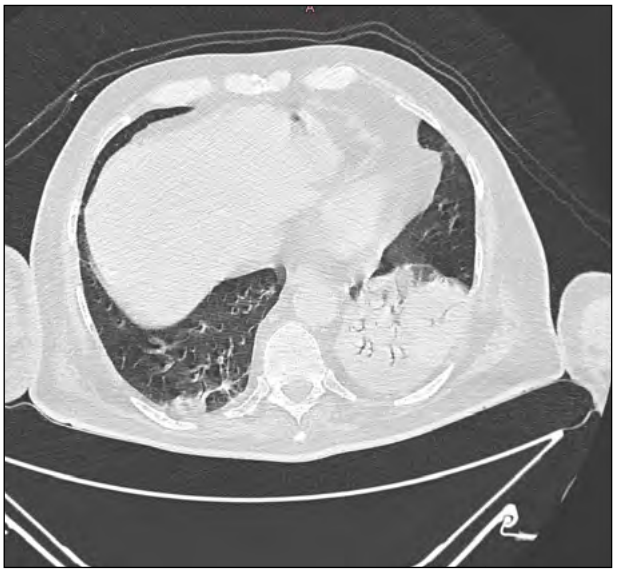

Figure 1. Persistent pulmonary atelectasis.

er lobe atelectasis. CT scan confirmed this finding and homolateral hemidiaphragm elevation (Fig. 1). Lung ultrasound confirmed the presence of a left pulmonary atelectasis which appeared as an area of pulmonary parenchyma with a tissue-like pattern and abolished lung sliding in the presence of lung pulse. Blood gas analysis revealed mild hypoxemia with a normal acid-basis balance. Functional respiratory tests showed a mild restrictive syndrome. Cough peak flow was 270 L/sec. Sputum microbiological examination, carried out on several samples, was negative.

The patient had been treated with various antibiotics (amikacin, vancomycin, piperacillin tazobactam, fluoroquinolones) and chest physiotherapy. It was decided a different respiratory physiotherapy approach, with a recruitment maneuver using the insufflation phase of a cough machine, associated with manually assisted cough and mechanical in-exsufflation therapy. Despite further 20 days of antibiotic therapy, the patient had no benefit: low grade fever (37.5°C max) and dyspnea persisted. As the patient and his family did not give consensus to a bronchoscopy to remove secretions, we opted for an active breathing assisted technique through the Temporary Positive Expiratory Pressure (TPEP®) device, twice daily for seven days. Using visual targets, inspiratory and expiratory resistors, the "I/E mode" program guide the patient to gradually inhale more deeply, to hold the breath for a while and to exhale slowly with an open glottis and an oscillating expiratory counter-flow.

After a brief training session, the initial settings included an inspiratory target of 4 cmH_2O sustained for a mean of 5 seconds, a 3 sec tele-inspiratory pause and an expiratory target of 10 cmH_2O sustained for about 6 sec. This program lasted at least 20 minutes every session, in the right and left lateral position.

In order to not increasing the work of breathing, flow-dependent resistances were gradually adjusted to make the patient's work as easy as possible. Every session ended with 10 minutes of breathing with the "TPEP mode" program, that allows a guided slow and deep expiration with an open glottis while providing a temporized oscillating PEP of 1 cmH_2O during most of the expiration. This second phase was aimed to enhance the expiratory flows and secretion removal. After trials for two days, no symptoms or atelectasis extension were recorded.

At the seventh day, we observed an improvement of gas exchange and the resolution of atelectasis at lung ultrasound. Patient underwent a thoracic TC scan that confirmed the resolution of the atelectasis (Fig. 2).

Discussion

Lobar atelectasis is a common problem caused by a variety of mechanisms including resorption, airway obstruction, hypoventilation, and compressive atelectasis from abdominal distension and adhesive atelectasis due to increased surface tension.

In literature there is a lack of evidence-based studies to guide the management of this common problem. Treatment modalities that have been described include chest physiotherapy, bronchodilators, and the use of positive end-expiratory pressure [3].

Chest physiotherapy is the first-line therapy for atelectasis due to airway obstruction. If physiotherapy fails, further radiological examination of the chest may be helpful to identify airway obstruction and to determine whether proximal lobar or distal bronchi are involved. Bronchoscopy to aspire secretions is useful in the management of atelectasis when less invasive solutions fail [4].

In this case report, the patient had a lower left lobe atelectasis mainly due to presence of blood cot and se-

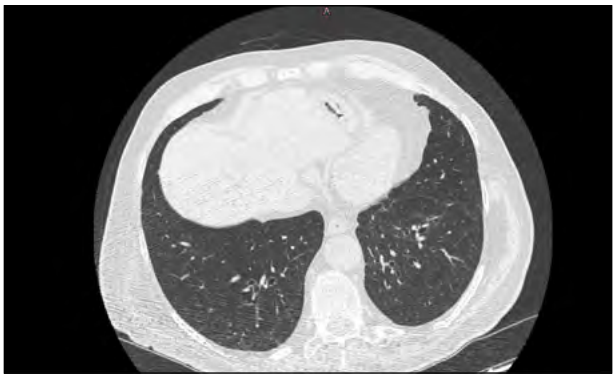

Figure 2. After TPEP therapy.

cretion retention. As the first approach with mechanical assisted lung recruitment fails, we decide for a bronchoscopy aimed to reduce the obstruction, but the patient refused it. Thus, we opted for an active device-assisted recruitment.

Temporary Positive Expiratory Pressure (TPEP) is a well-known patented technology to mechanically deliver a low positive expiratory pressure at the mouth during spontaneous breathing and it is usually used to assist patients with chronic obstructive pulmonary disease for airway clearance. The I/E mode was recently added to allow the treatment of deep lung in cooperative patients. The use of positive expiratory pressure to improve lung volumes is well documented in the literature; in general the physiotherapists use this strategy in patients with low lung volumes to improve ventilation and gas exchanges.

The use of PEP and T-PEP also improve dyspnea and quality of life and speed up the improvement of bronchial encumbrance [3,4]. Preliminary results show that an expiratory pressure ≤ 1cm H_2O applied for a fraction of the expiratory phase, may improve the distribution of alveolar ventilation [5].

The use of TPEP is documented in chronic respiratory diseases, including COPD, asthma and Cystic Fibrosis; it has been shown that symptoms and pulmonary function testing (including reduction in air-trapping) improved after 2 weeks of treatment [6]. Currently, no studies are available about the use of this device for the treatment of massive atelectasis, neither in the treatment of Steinert patients.

Nevertheless, we know that lung alterations in neuromuscular diseases are unusual and require extra-caution with the use of positive pressure. TPEP uses positive expiratory pressures several times lower than the commonly used devices and, when used in the I/E mode, pressures are used as a target driving the patient breath by breath, thus preventing excessive mechanical stress on the bronchial tree and lung parenchyma.

This clinical case was challenging because no results were obtained with the usual recruitment strategies (long-term low pressure mechanical insufflation), while with the active "I/E mode" strategy we obtained a complete resolution of a massive atelectasis, with improvement of gas exchanges and resolution of hypoxemia.

This experience encourages the use of the TPEP device in neuromuscular patients with persistent lung involvement. Clinical diffusion of the I/E mode is still limited, although there is a growing interest in identifying the potential benefits of this technology in a wide range of respiratory conditions, and its role in pulmonary rehabilitation programs for neuromuscular diseases.

Conclusions

The multidisciplinary team is necessary to guarantee a comprehensive diagnostic approach, early recognition of complications, and personalized therapy for these patients with multiple frailties. In this clinical case we have successfully used TPEP® and I/E mode for the treatment of low left lobe obstructive atelectasis in a patient affected by Steinert disease, achieving rapid resolution of atelectasis. This experience encourages the use of this device in the treatment of atelectasis as a viable option for other passive or invasive methods; moreover its role in the rehabilitation of lung inhomogeneities in neuromuscular patients.

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Conflict of interest statement

The Authors declare no conflict of interest.

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Authors' contributions

AC: conceptualization, data collection, draft writing, final version writing; AA: conceptualization, reading, final version writing; SM: data collection; ES: data collection; GF: supervision

Ethical consideration

This study was approved by the Institutional Ethics Committee of University of study of Campania "L. Vanvitelli" (protocol number 372/2019). The research was conducted ethically, with all study procedures being performed in accordance with the requirements of the World Medical Association's Declaration of Helsinki. Written informed consent was obtained from patient for study participation and data publication.

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