Can percussive intrapulmonary ventilation improve the efficacy of physiotherapy in children with cystic fibrosis?

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Abstract

Lung disease in cystic fibrosis (CF) is characterized by reduced mucociliary clearance, airway plugging, recurrent infections, and chronic pulmonary inflammation. Patients who are affected undergo daily respiratory physiotherapy to improve airway clearance. Intrapulmonary percussive ventilation (IPV) is a technique used in clinical practice, but it is not commonly used in CF patients. Evidence for various respiratory pathologies, particularly in children, is still lacking. We present the case of an 11-year-old boy with cystic fibrosis who did not respond to traditional respiratory physiotherapy techniques. We proposed and tested the use of IPV during hospitalization. In this case, the use of IPV in physiotherapy treatment reduced the need for intravenous antibiotics, hospitalization, and improved radiologic features. IPV can be used successfully in CF patients who are resistant to traditional physiotherapy techniques.

Introduction

Cystic fibrosis (CF) is the most common autosomal recessive lethal disorder in the Caucasian population due to mutations in the cystic fibrosis transmembrane conductance regulator gene (*CFTR*). The respiratory system and gastrointestinal tract are primarily involved, but eventually, multiple organs are affected.

Lung disease is characterized by bronchiectasis, reduced mucociliary clearance, airway plugging, chronic bacterial infections, and chronic pulmonary inflammation. Daily physiotherapy aims to compensate for reduced mucociliary clearance to minimize lung disease and preserve lung function, as well as maintain endurance and allow a good quality of life [1].

Physiotherapy treatment improves airway clearance; usually, a combination of techniques targeting various levels of the airways is performed for patients with CF. Intrapulmonary percussive ventilation (IPV) is a ventilatory technique that uses a device to deliver small bursts of high-flow air into the lungs at high rates, superimposed upon the spontaneous breathing pattern. This causes airway pressures to oscillate between 5 and 35 cm of water (cmH_2O) and the airway walls to vibrate in synchrony with these oscillations. A unique sliding Venturi, called phasitron, which is powered by compressed gas at 0.6 to 6 bars, generates these oscillations in the range of 80 to 650 cycles per minute. The high-frequency gas pulses expand the lungs, vibrate and enlarge the airways, and deliver gas into distal lung units over accumulated mucus [1].

Case Report

Our patient was an 11-year-old boy with cystic fibrosis diagnosed by neonatal screening and was heterozygous for the *F508del* and *R347P* mutations in *CFTR*. He was pancreatic sufficient and had no cystic fibrosis-related diabetes. Despite treatment with inhaled antibiotics, he developed *Pseudomonas aeruginosa* chronic infections at the age of 4 years old.

Through inhaled mucolytic therapy, inhalation of dornase a was started at 3 years old and then increased to twice a day in the past 9 years. Hypertonic saline was not tolerated due to bronchospasm.

Airway techniques have been started at the age of 2 months old. The patient performs airway clearance techniques with a positive expiratory pressure (PEP) mask twice a day. He was hospitalized annually from birth until he was 5 years old, and then at least two or three times a year for pulmonary exacerbations. At 7 years old, he started high-frequency chest wall oscillation therapy in addition to standard airway clearance techniques and stopped after 12 months for no morbidity reduction.

The patient's daily pulmonary care included two or three daily sessions of respiratory physiotherapy performed by PEP mask and autogenic drainage, preceded by inhalation of a pressurized meter dose inhaler (pMDI) bronchodilator, two daily inhalations of dornase a, and two daily inhalations of antibiotics (colistin and tobramycin, alternating continuously). In October 2019, at the age of 9, the patient was hospitalized again for pulmonary exacerbations, and intravenous antibiotic therapy was started. On examination, he appeared dyspneic, his oxygen saturation was 95% (breathing in room air), and his forced expiratory volume in 1 second (FEV_1) was 1.16 L (57% predicted). Secretions were very thick and difficult to expectorate, and he got tired during physiotherapy sessions with the PEP mask.

Upon resolution of the exacerbation, chest computed tomography (CT) showed a large amount of cystic and varicose bronchiectasis in the left upper lobe (100% of the lobe), filled with a large amount of mucus plugging, especially in the inferior lingular segment (>66%). In the same lobe, small airways' mucus plugging was visible in the periphery, in the apical part of the apico-dorsal segment. A large amount of cystic and varicose bronchiectasis with mucus plugging (33-66%) was visible in the left lower lobe, superior segment, and dorsal basal segment too, accompanied by several small airways' mucus plugging (33-66%). In the right upper lobe, a small amount of small airways' mucus plugging (<33%) was shown. In the medial segment of the middle lobe, large varicose bronchiectasis were cleared.

During the hospitalization, we proposed and tested various airway clearance techniques, including periodic continuous positive airway pressure, oscillating PEP, and IPV. IPV was the most effective and well tolerated by the patient, allowing it to shorten the treatment's duration. The IPV device has been set to 12-13 cmH_2O/400 bpm, combined with 3% hypertonic saline nebulization. Upon discharge from the hospital, he appeared eupneic, his oxygen saturation was 97% (breathing in room air), and his FEV_1 was 1.29 L (65% predicted). His daily pulmonary care included two daily sessions of chest physiotherapy performed by IPV, preceded by the inhalation of a pMDI bronchodilator, two daily inhalations of dornase a, and two daily inhaled antibiotics. From release for the next 14 months, he was clinically stable. He reported that IPV was effective, simple to use, and improved tolerance to hypertonic saline therapy. He did not need intravenous antibiotics or hospitalization, and no changes to drug therapy were made. Because the patient was under the age of 12, it was not possible to begin cystic fibrosis transmembrane conductance regulator modulator therapies. In January 2021, at the age of 11, he was hospitalized following a pulmonary exacerbation and concomitant COVID-19 positivity. On examination, he appeared eupneic; his oxygen saturation was 97% (breathing room air); and his secretions were few and thick.

Lung CT showed a better detersion than in the previous CT of the varicose and cystic bronchiectasis of the left upper lobe (33-66%) and of the left lower lobe, where the big airways' mucus plugging was reduced (<33%) and the small airways mucus plugging persisted but less than before (<33%), as in the basal segment of the right upper lobe. In the medial segment of the middle lobe, there was mucus plugging of the large bronchiectasis mentioned (33-66%), while in the last CT, they were cleared.

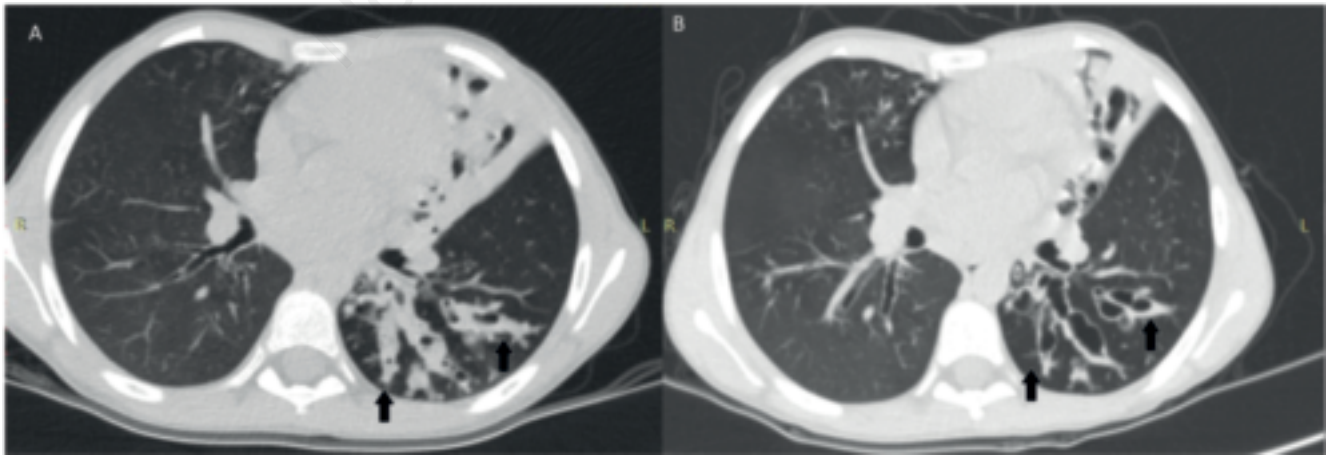

Figure 1. A) Axial image of the inspiratory computed tomography scan, we can see the mucus obstructing varicose bronchiectasis in the superior segment of the left lower lobe and atelectasis of the lower segment of the lingual lobe, with bronchiectasis filled with mucus; B) axial image of the inspiratory computed tomography scan of the same patient, at the same level as A, we can see cleansed, mucus-free varicose bronchiectasis in the superior segment of the left lower lobe and better cleansing of the bronchiectasis of the lower lingular segment.

Discussion

Intrapulmonary percussive ventilation is frequently used in clinical practice to increase airway clearance and lung recruitment in patients with respiratory disease. However, the evidence in different respiratory pathologies, especially as regards children, is still lacking [1].

A recent review suggests that the systematic use of IPV as an airway clearance technique in chronic obstructive airway diseases is not supported by sufficiently strong evidence to recommend routine use in this patient population [2]. However, many authors propose IPV as an airway clearance technique that may offer valuable assistance in the treatment of severe respiratory diseases refractory to conventional respiratory therapy techniques [3,4]. One study in cystic fibrosis patients demonstrated that IPV at high frequency increased FEV_1 and forced vital capacity compared to other techniques [5].

In this subject, the introduction of IPV in physiotherapy management contributed to i) reducing the frequency of intravenous antibiotic use: zero times/years *versus* three times/years; ii) reducing the need to be hospitalized: zero times/years *versus* three times/years; iii) improve radiologic features (Figure 1); iv) reducing big airways mucus plugging of the left upper lobe (33-66% *versus* >66%); v) reduction of big and small airways mucus plugging of the left lower lobe (<33% *versus* 33-66%).

In our clinical practice, IPV is widely used in the pediatric intensive care unit; for at-home treatment, this device is used only in selected patients when other techniques are not efficacious. The complexity, size, and difficulty of adjusting the equipment must be considered for daily use. It is also not possible to set a standard setting because the system is dynamic and changes in relation to pulmonary resistance in the same session.

Patients with chronic respiratory disease, such as CF, generally prefer devices that encourage independence, are easily transportable, and are easy to clean, such as positive pressure devices.

Conclusions

Intrapulmonary percussive ventilation, in our experience, can be used in CF patients who are resistant to traditional physiotherapy techniques, particularly those who are not candidates for treatment with new *CFTR* modulators.

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