Evaluation of respiratory muscle dysfunction in patients with Pompe’s disease by optoelectronic plethysmography: a preliminary study

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SUMMARY
Some muscular diseases are characterized by respiratory impairment principally due to diaphragmatic weakness. The respiratory management of these patients is based on a better understanding the impact of respiratory failure on chest wall biomechanics. Optoelectronic plethysmography (OEP), using a three dimensional motion analysis system, can be used to evaluate respiratory impairment from chest wall compartment kinematics using a three-dimensional model and allow quantification of coordination between compartments. The aim of this preliminary study was to use OEP in order to identify early indicators of the diaphragmatic weakness and to characterize ventilatory patterns in neuromuscular patients with Pompe’s disease. Three patients with Pompe’s disease and five healthy subjects were included. Spirometry, ventilatory pattern and compartmental chest wall kinematics were assessed using OEP during spontaneous breathing (SB), slow vital capacity (SVC) and sniff maneuver, in a supine position. The results showed that patients had lower values of tidal volume and vital capacity than healthy subjects. A negative abdominal contribution was observed in patients, during SB, SVC and sniff, indicating a paradoxical movement during inspiration due to paralysis of the diaphragm. In conclusion, the study of rib cage and abdomen compartment coordination during breathing, can allow characterization of the ventilatory pattern and diaphragmatic impairment in patients with neuromuscular disease.

INTRODUCTION
In addition to locomotor disabilities, patients with neuromuscular disease progressively develop chest deformities and abnormalities of lung ventilation due to atrophy and weakness of their respiratory muscles. Involvement of respiratory muscles, with a progressive diaphragmatic weakness, is the most common cause of premature death in patients with neuromuscular disease. Chest wall distortion leads to ineffective action of the respiratory muscles, and a deterioration of their ability to generate sufficient pressure and flow rates during tidal breathing, and during voluntary respiratory maneuvers. In this clinical context, understanding the mechanisms that lead to impairment of coordination of the chest wall compartments during breathing could assist appropriate management of patients. Respiratory inductance plethysmography, a technique that measures tidal volume from rib cage and abdominal motions with two elastic bands into which a coiled wire is sewn, is generally used in clinical routine, but it is little accurate and is based on two-dimensional biomechanics. Using a motion analysis system, optoelectronic plethysmography (OEP), is reliable and accurate for the evaluation of the kinematics of different thoracoabdominal compartments (rib cage, abdomen, and right and left side) and pulmonary ventilation, in three dimensions, based on tracking of reflective markers placed on the chest wall. With OEP, the study of coordination between different compartments of the thoracoabdominal wall during breathing can help to characterize the ventilatory pattern of different neuromuscular diseases and to identify early indicators of diaphragmatic impairment. The aim of this preliminary study was to evaluate the relevance of OEP to identify early indicators of diaphragmatic weakness and to characterize the ventilatory pattern in Pompe’s disease, an autosomal recessive disease.

METHODS
Three patients with Pompe’s disease and five healthy subjects were included. Each patient performed spirometry and OEP recordings in two conditions: 3 min of spontaneous breathing (SB) and slow vital capacity (SVC). In addition, patients performed sniff maneuvers assessed only with OEP. A recording of SB, SVC and sniff were performed in a supine position and for SB and SVC spirometry was carried out using a flow meter attached to a mouthpiece and a nose clip. Simultaneous, chest wall kinematics were recorded by a motion analysis system (100 Hz, Motion Analysis®) consisting of 8 optoelectronic cameras. Based on the biomechanical model developed by Cala [1], 52 reflective markers were placed on specific landmarks of the chest wall (42 anterior and 10 lateral), from the clavicles to anterior superior iliac spines. To enclose the chest wall volume, the posterior part of the chest wall (in contact with bed) was created with virtual markers [2]. The chest wall (CW) was separated into three compartments: pulmonary rib cage apposed to the lungs (RCp), abdominal rib cage apposed to the diaphragm (RCa) and abdomen (AB) [3]. From the recording of the 3D coordinates of points relating to the surface of the CW, the changes in volume of the chest wall and its compartments were computed from a triangulated mesh, created by connecting nodes, represented by markers. This mesh defined a geometrical model describing the entire surface of the chest wall or only a part of it. Changes in
volume of total chest wall (VCW) and each compartment (VRcp, VRca and VAB) were quantified using the Gauss divergence theorem from the triangulation of the closed surface, obtained by the 3D coordinates (X, Y and Z) of the reflective markers [1]. The total VCW was computed as the sum of VRcp, VRca and VAB. From compartmental chest wall volume variations, pulmonary parameters, (tidal volume (VT), vital capacity (VC)), chest wall kinematics and compartmental coordination were considered. Values of patients’ diaphragmatic pressure (Pdi) assessed during routine diaphragm exploration were also noted for comparison with OEP data.

RESULTS AND DISCUSSION

The preliminary results showed high correlation between chest wall volume variations recorded with OEP and spirometry, during SB and SVC ($r = 0.98$, $p < 0.0001$ and $r = 0.95$, $p < 0.0001$, respectively). The discrepancy between the values of the two measures was less than 5% and the Bland and Altman test indicated a good agreement between the two measures, during SB and SVC. Ventilatory parameters assessed between patients and healthy subjects are presented in Table 1. The results of pulmonary function evaluated with OEP showed that patients had lower values of VT and VC than healthy subjects: 0.34 vs. 0.61 L and 0.54 vs. 4.68 L, respectively. In the supine position, patients had a negative abdominal contribution to chest wall volume variations indicating a paradoxical movement during inspiration, due to paralysis of the diaphragm. The ratios between volume variation of abdominal and rib cage compartment ($\Delta V_{AB}/\Delta V_{RC}$), were -36%, -34% and -26%, during SB, SVC and sniff maneuver respectively. Theses results indicate an asynchronous movement between RC and AB compartments with a positive contribution of the rib cage. Using the Gilbert index [4], which quantifies diaphragm impairment, a preliminary linear relation was found between diaphragmatic pressure values ($\Delta P_{gas}/\Delta P_{di}$) in patients and the ratio $\Delta V_{AB}/\Delta V_{RC}$ during SB.

CONCLUSIONS

Optoelectronic plethysmography is a suitable and accurate method to quantify distortion of different chest wall compartments. In neuromuscular disease, paralysis of the diaphragm leads to a decoupling between motion of the rib cage and abdominal compartments, with a paradoxical movement of the abdomen during inspiration. The magnitude of the diaphragmatic impairment can be appreciated by quantification of the abdominal contribution. The results of this study may help to show a relationship between diaphragmatic pressure data measured during pulmonary function tests and data from OEP, thus dispensing with the necessity of pressure sensors to measure the performance of the diaphragm. Further studies should be conducted in order to show that OEP can be used in clinical routine for respiratory muscle dysfunction evaluation, without using invasive methods.

REFERENCES


Table 1: Pulmonary function and chest wall kinematics parameters assessed by optoelectronic plethysmography during SB.

<table>
<thead>
<tr>
<th>SB condition</th>
<th>VT (L)</th>
<th>$\Delta V_{RC}$ (%)</th>
<th>$\Delta V_{AB}$ (%)</th>
<th>$\Delta V_{AB}/\Delta V_{RC}$ (%)</th>
<th>$\Delta P_{gas}/\Delta P_{di}$</th>
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<tr>
<td>Healthy subjects (n=5)</td>
<td>0.60</td>
<td>49</td>
<td>51</td>
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<td>Patients (n=3)</td>
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<td></td>
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<tr>
<td>Patient #1</td>
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<td>Patient #3</td>
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<td>180</td>
<td>-80</td>
<td>-44</td>
<td>0</td>
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