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SÉRIE DE CASOS

High frequency chest wall oscillation plus Mechanical In-Exsufflation in Duchenne muscular dystrophy with respiratory complications related to pandemic Influenza A/H1N1

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Abstract

Two young boys with Duchenne muscular dystrophy, who had contracted 2009 pandemic influenza A/H1N1 (pH1N1), had been treated with antibiotics and steroids without significant improvement. One of them showed severe scoliosis. After hospitalization chest CT scan revealed extensive pulmonary bilateral segmental atelectasis. Their clinical and radiological findings rapidly improved when a sequential respiratory physiotherapy protocol was adopted that consisted of the application of multiple sessions of high-frequency chest wall oscillations, each one followed by mechanically assisted coughing manoeuvres. The protocol was well tolerated, effective, easy to apply and special positioning was not required. Fifteen days after treatment initiation both patients clinically recovered. This treatment can be very helpful for neuromuscular patients, particularly when scoliosis prevents conventional respiratory physiotherapy.

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Oscilação de alta frequência da parede torácica associado a in-exsuflação mecânica na distrofia muscular de Duchenne com complicações respiratórias relacionadas com o vírus pandêmico da gripe A/H1N1

Resumo

Duas crianças do sexo masculino com distrofia muscular de Duchenne que contraíram o vírus da gripe pandêmica A/H1N1(pH1N1) de 2009 foram tratados com antibióticos e esteroides sem melhoria significativa.
Um deles revelou escoliose severa. Depois da hospitalização, um TAC ao peito revelou uma atelectasia pulmonar segmentar bilateral extensa. Os seus resultados clínicos e radiológicos melhoraram rapidamente quando foi adoptado um tratamento de fisioterapia respiratória sequencial, consistente na aplicação de múltiplas sessões de oscilações torácicas de alta frequência, cada uma seguida por exercícios de tosse mecanicamente assistidos. O tratamento foi bem tolerado, eficaz e fácil de aplicar, sendo que não foi necessário um posicionamento especial. Quinze dias depois do início do tratamento, ambos os pacientes se encontravam clinicamente recuperados. Este tratamento pode ser muito útil em pacientes com doenças neuromusculares, particularmente quando a escoliose impede a fisioterapia respiratória convencional.

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Background

Patients with Duchenne muscular dystrophy (DMD) are at high risk of experiencing life-threatening complications from viral infections due to their reduced respiratory muscle function. Therefore, they have been recommended vaccination against seasonal influenza and recently even against 2009 pandemic influenza A/H1N1 (pH1N1). Early reports of pH1N1 influenza described a more pathogenic agent than the seasonal influenza virus, causing severe complications in young individuals. So far there are no reports on pH1N1 complications in neuromuscular patients and on their management. Viral respiratory infection can cause secretion encumbrance, reductions in vital capacity and peak cough flow, which urgently need manual or mechanical assistance to cough. We report two cases of boys affected by DMD who developed acute respiratory failure caused by bilateral pulmonary segmental atelectasis after pH1N1 infection and who were successfully treated with a new protocol that included high-frequency chest wall oscillation (HFCWO) followed by mechanical in-exsufflation (MI-E) for cough assistance.

Case presentations

Case 1

A 15 year old boy with DMD was referred to our hospital in November 2009 with productive cough and fever (38.5 °C). He had received spinal arthrodesis at 12 years of age and had a history of progressive weakness, but not of recurrent pneumonia or increased frequency of upper airway infections. The patient had a frequent cough upon admission, but appeared unable to effectively clear airway secretions. Clinical exam revealed tachypnea (40 cycles/min) and tachycardia (140 beats/min). Lung auscultation revealed bilateral ronchi and discrete crackles in both lung fields. Peripheral O₂ saturation was 80% on oximetry. Blood gas analysis demonstrated normocapnia. Virus infection by pH1N1 (H1N1 RT- PCR test in nasopharyngeal swabs) was demonstrated. He was already on the sixth day of antibiotic and steroid therapy. Bilateral atelectasis was suspected and a chest CT scan was performed (fig. 1A).
The pulmonologist prescribed HFCWO (The Vest Airway Clearance System, Hill-Rom St. Paul, MN) at a pressure setting of 5 cm H2O and a frequency of 12 Hz for sessions of 20 minutes, followed by five or six sessions of Mi-E for cough assistance with an in-exsufflator at a pressure of +40/+45 cm H2O delivered respectively over 3 and over 2 seconds (Cough-Assist®, Philips Respironics, Murrysville, PA, USA). Oseltamivir was administered for five days and broad-spectrum antibiotics were administered intravenously for 10 days. A respiratory therapist monitored the patient with a pulse oximeter and acted as a liaison with the pulmonologist. The sequential protocol (HFCWO + Mi-E) was applied for 15 minutes three times/day until clinical and radiological recovery was obtained (fig. 1B). Few minutes after its initiation the patient’s cough rate increased and a large volume of secretions was returned. The patient tolerated well this protocol. Functional respiratory tests before viral infection and at hospital discharge are shown in table 1.

### Case 2

The second patient with DMD was a 16 year old boy with severe scoliosis, pelvic distortion and severe reduction of respiratory muscle strength and lung volumes. In December 2009 he developed dyspnea and persistent cough and fever and was treated with antibiotics and steroids for 10 days without any improvement. Later in the same month he was admitted to our department. He showed severe left convex scoliosis with a mean Cobb angle of over 70°. Clinical examination revealed tachypnea (50 cycles/min), tachycardia (140 beats/min), bilateral ronchi and high-pitched expiratory wheezes at lung auscultation. Peripheral O₂ saturation was 86 %. Blood gas analysis showed normocapnia. A nasopharyngeal swab was positive for pH1N1. Chest radiographs did not explain the clinical picture and chest CT scan was urgently performed. Bilateral pulmonary atelectasis was observed, similar to the other boy (fig. 2A). He was started on assisted pressure controlled ventilation (Idea Ultra SAIIME) with a pressure support of 16 cm H₂O and 4 cm H₂O of PEEP. The same sequential protocol with HFCWO followed by Mi-E was prescribed and administered with the same setting until clinical and radiological improvement (fig. 2B). Oseltamivir and broad-spectrum antibiotics were also administered. Despite the severe scoliosis, the protocol was very well tolerated, effective and easy to apply. The patient was

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Respiratory function in Case 1 before and after treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>FVC (ml)</td>
</tr>
<tr>
<td>Baseline</td>
<td>1.68</td>
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<tr>
<td>1st week</td>
<td>1.58</td>
</tr>
<tr>
<td>2nd week</td>
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</table>

FVC: forced vital capacity; MIP: maximal inspiratory pressure; MEP: maximal expiratory pressure; PCF: peak cough flow (unassisted); PaO₂: arterial partial pressure of oxygen; PaCO₂: arterial partial pressure of carbon dioxide in blood; HCO₃⁻: bicarbonate; BEb: base excess; SaO₂: arterial oxygen saturation.

Figure 2  Panel A: Case 2: Atelectasis of left lower lobe and lingula; lower right lobe is also partially involved. Panel B: Same patient: almost complete resolution after treatment.
Cough assistance in Duchenne muscular dystrophy with pandemic Influenza

Table 2  Respiratory function in Case 2 before and after treatment

<table>
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<tr>
<th></th>
<th>Baseline</th>
<th>1st week</th>
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<tbody>
<tr>
<td>FVC (ml)</td>
<td>0.79</td>
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<tr>
<td>MIP (cmH2O)</td>
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<tr>
<td>MEP (cmH2O)</td>
<td>17</td>
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<td>PCF (L/min)</td>
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<tr>
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<td>PaO2 (mmHg)</td>
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<td>82</td>
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<td>PaCO2 (mmHg)</td>
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</tr>
<tr>
<td>HCO3 (mmol/L)</td>
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<td>30</td>
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<tr>
<td>BEb (mmol/L)</td>
<td>4.2</td>
<td>6.6</td>
<td>4.2</td>
</tr>
<tr>
<td>SaO2 (%)</td>
<td>86</td>
<td>96</td>
<td>97</td>
</tr>
</tbody>
</table>

FVC: forced vital capacity; MIP: maximal inspiratory pressure; MEP: maximal expiratory pressure; PCF: cough expiratory peak (unassisted); PaO₂, arterial partial pressure of oxygen; PaCO₂, arterial partial pressure of carbon dioxide in blood; HCO₃⁻, bicarbonate; BEb: base excess; SaO₂: arterial oxygen saturation.

discharged clinically recovered on nocturnal mechanical ventilation after two weeks. Table 2 shows functional respiratory tests and blood gas analysis before and after treatment.

Discussion

Influenza can have devastating effects in neuromuscular patients. Dystrophic children are highly likely to develop severe complications with influenza, including atelectasis,² as a result of acute retention of secretions. A direct consequence of atelectasis may be acute hypoxemia.

Interventions for clearing the airway and improving ventilation in DMD are vital. Pharmacological measures are often insufficient to improve health status. Similarly, mechanical in-exsufflators alone can be insufficient if secretions are very tenacious or difficult to mobilize. Chest physiotherapy with postural drainage is now obsolete. It can be uncomfortable and often impractical for individuals with limited mobility and significant skeletal deformity,³ as it requires assuming a posture that patients with severe scoliosis can hardly maintain. Moreover it may have side effects that reduce its effectiveness. Fiberoptic bronchoscopy has been used in the management of proximal airway obstruction to aspirate secretions and has successfully resolved atelectasis in some paediatric intensive care patients.⁴ However, in a randomised controlled trial on adult patients with atelectasis this method did not improve the rate of resolution of volume loss better than chest physiotherapy and was occasionally followed by adverse effects on intracranial pressure.⁵ In neuromuscular patients it has not been of proven benefit and should be considered only after all non-invasive airway clearance techniques have proven unsuccessful.⁶ Nebulised bronchodilators are traditionally recommended for the management of atelectasis, but there is no experience with their use in dystrophic patients. In patients with acute bronchoconstriction, a bronchodilator may improve secretion clearance, but there are no published studies evaluating its use in the management of atelectasis in non-asthmatic patients.⁷ HFNCWO is becoming the method of choice for individuals with many types of disabilities.⁸ 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. Treatment generally lasts up to 30 minutes. This technique was designed for in-home use and can treat all lung segments simultaneously. Special positioning and breathing techniques are not required.

Acknowledgment

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References