Introduction

Myotonic dystrophy, or Steinert disease, is an autosomal dominant disorder that develops slowly, with neuromuscular and systemic involvement. The main symptoms are muscular weakness, myotonia, gonadal atrophy, cataracts, cardiac arrhythmias, and restrictive pulmonary impairment. Pulmonary involvement is usually secondary to muscle weakness and develops along with diminished thoracic mobility and respiratory muscle strength.

Respiratory muscle training has been shown to be effective for improving muscle function in some diseases that cause muscle weakness. Training also facilitates ventilator weaning in postoperative patients. Moreover, muscle weakness in general and respiratory muscle weakness in particular often lead to respiratory complications in a variety of circumstances. However, the effectiveness of muscle training has never been investigated in the context of myotonic dystrophy. We report the case of a patient with this disease for whom tailored respiratory muscle training led to clear improvement in symptoms and lung function.

Case Description

A 42-year-old man diagnosed with myotonic dystrophy experienced loss of respiratory muscle strength over a period of 6 months. We report the application of a domiciliary training program targeting both inspiratory and expiratory muscles. Maximal inspiratory and expiratory pressures, forced vital capacity, and forced midexpiratory flow rate were measured 6 months before start of training, just before commencement of the program, and immediately after 12 weeks of training. Adherence to the program was satisfactory. Inspiratory muscle training was efficacious in increasing respiratory muscle strength. Expiratory muscle training, which made use of the Threshold PEP bronchial hygiene device incorporating an adapted flutter valve, was not efficacious in increasing maximal expiratory pressure or halting its loss. However, decreased obstruction of medium-caliber airways was observed with use of the device.

Key words: Myotonic dystrophy. Respiratory muscle training. Maximal respiratory pressures.

Entrenamiento muscular respiratorio domiciliario en la distrofia muscular miotónica

Se describe el caso de un varón de 42 años de edad con diagnóstico de distrofia muscular miotónica e historia de disminución de la fuerza muscular respiratoria en los últimos 6 meses. Relatamos la aplicación de un programa de entrenamiento muscular respiratorio (inspiratorio e expiratorio) domiciliario. En los 6 meses previos al entrenamiento, justo antes del comienzo del programa se observó la disminución de la fuerza muscular respiratoria. El entrenamiento muscular inspiratorio utilizando la válvula adaptada de higiene bronquial Threshold PEP®, adaptada para el entrenamiento, no fue efectivo para aumentar la fuerza muscular respiratoria. El entrenamiento muscular espiratorio utilizando la válvula adaptada de higiene bronquial Threshold PEP® se observó la disminución de la obstrucción de las vías aéreas de medio calibre.

start and end of training. The lung function tests revealed slight restrictive abnormalities and maximum respiratory pressures within reference values. At the time of the visit to start therapy (6 months after the first visit), the lung function study also revealed a slight decline in respiratory muscle strength but no change in basic spirometric parameters (Table).

The patient was advised to undertake a respiratory muscle training program, taught in the hospital and continued at home. That program included the following components: inspiratory muscle training (IMT) with a specific device (Threshold IMT, Respironics, Cedar Grove, New Jersey, USA) which provided an inspiratory resistance between 7 and 60 cm H\(_2\)O and expiratory muscle training with a device able to induce a huff cough. There are no valves available on the market designed expressly for expiratory muscle training. Because the load could not be increased beyond 20 cm H\(_2\)O, it was impossible to halt the loss of expiratory muscle strength. Also important is the fact that training did not lead to improved forced vital capacity or forced expiratory volume in 1 second (FEV\(_1\)_1). From the reduced severity of expiratory muscle weakness, the results would also indicate greater facility in mobilizing secretions and reduce the risk of respiratory infections. In addition, the involvement of inspiratory muscle training eventually causes hypercapnia, which may develop soon or be delayed. In both cases respiratory muscle involvement culminates in respiratory failure.

Although satisfactory results were observed in relation to inspiratory muscles and midexpiratory flow, it must be recognized that the most important limitation in this case was the commercial valve used for expiratory muscle training. Because the load could not be increased beyond 20 cm H\(_2\)O, it was impossible to halt the loss of expiratory muscle strength. Also important is the fact that training did not lead to improved forced vital capacity or forced expiratory volume in 1 second in spite of satisfactory improvement in midexpiratory flow. The reason may be related to the low training load provided by the commercial valve, which was designed to improve bronchial hygiene and induce a huff cough. There are no valves available on the market designed expressly for expiratory muscle training. Finally, another important finding was the improvement in PaCO\(_2\) after 12 weeks of training. This observation suggests that improvement might have been linked to a switch to a more efficient breathing pattern, although we did not measure the relevant variables to confirm this.

Our patient’s increase in inspiratory muscle strength (36%) allows us to predict slower functional decline as well as slower progression to respiratory failure. Perhaps if we had applied loads greater than 60% of maximum inspiratory pressure, the results would also have led to improved forced vital capacity. Improvement in midexpiratory flow is interpreted to indicate greater facility in mobilizing secretions and greater availability of peripheral airflow. Because the program is home based, its simplicity and applicability allow us to propose it for certain patients in order to objectively evaluate the effects of short- and long-term muscle training of this type.

**REFERENCES**


<table>
<thead>
<tr>
<th>TABLE Lung Function During Clinical Follow-up*</th>
</tr>
</thead>
<tbody>
<tr>
<td>G-Muscle Before Training</td>
</tr>
<tr>
<td>-------------------------</td>
</tr>
<tr>
<td>FVC, L (%)</td>
</tr>
<tr>
<td>FEV(_1), L (%)</td>
</tr>
<tr>
<td>FEV(_1)/FVC, %</td>
</tr>
<tr>
<td>FIF(_1)(_25)-%</td>
</tr>
<tr>
<td>FE(_FV(_1))(_25)-75)</td>
</tr>
<tr>
<td>pH</td>
</tr>
<tr>
<td>PaCO(_2), mm Hg</td>
</tr>
</tbody>
</table>

*FIF\(_1\)\(_25\)-75\) indicates forced inspiratory flow; FE\(_FV\(_1\)\)\(_25\)-75\), forced expiratory volume in 1 second; FVC, forced vital capacity; FIF\(_1\)\(_25\)-75\), maximal inspiratory pressure; FIF\(_1\)\(_25\)-75\), maximal expiratory pressure.