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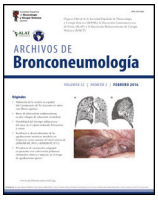

Editorial

Amyotrophic Lateral Sclerosis: The Assessment of Inspiratory Muscle Failure☆

Esclerosis lateral amiotrófica: valoración del fracaso muscular inspiratorio

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Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease which causes progressive muscle weakness due to the involvement of both first and second order motor neurons. The incidence is 1–2 cases per 100 000 inhabitants per year, while prevalence is 4–6 per 100 000 inhabitants. It is diagnosed from a combination of clinical symptoms and electromyographic changes.[1,2] Median survival of ALS patients is 20–48 months, although between 5% and 10% of patients survive more than 10 years.[3] Complications associated with respiratory failure in these patients are among the most common causes of hospitalization and even death.[4]

Respiratory failure in ALS patients is due mainly to respiratory muscle dysfunction. This editorial is a brief review of the methods used to assess inspiratory muscle dysfunction in patients with ALS. Many of these tests can be difficult to perform in ALS patients, mainly due to bulbar compromise and the possibility of associated frontotemporal dementia.[5] Treatment with mechanical ventilation (MV) may be considered for the management of inspiratory muscle dysfunction.[6] Home MV improves patients' quality of life and survival.[6] Therefore, an accurate, early diagnosis of inspiratory muscle dysfunction must be made so that MV can be properly prescribed and disease prognosis can be better determined.

Functional parameters obtained from forced spirometry are currently used in the diagnosis of inspiratory muscle dysfunction. The most frequently used measurement is forced vital capacity (FVC), which has been described as an impressive predictor of disease progression and survival.[7] However, some authors have reported that between 65% and 75% of ALS patients with a normal FVC have inspiratory muscle dysfunction determined by measuring muscle strength.[8] In order to improve the assessment of inspiratory muscle dysfunction using FVC, variations depending on the position of the patient have been studied. For example, changes in FVC between a sitting and supine position have been shown to correlate with diaphragm strength in ALS patients.[9] This test is difficult to perform, as many ALS patients have limited mobility, and forced spirometry maneuvers in patients with advanced disease and/or bulbar involvement are also difficult to perform, thus limiting its validity.[10] In order to avoid these limitations, vital capacity can be determined by a slow maneuver, as this involves fewer technical difficulties for patients than the forced maneuver.[11]

For all these reasons, the best option would be to directly determine the strength of the inspiratory muscles. Diaphragm strength, measured by determining transdiaphragmatic pressure during a maximum inspiration maneuver or with magnetic stimulation, is the best predictor of prognosis and the need for MV in ALS patients.[11] However, this technique is invasive and not available in all centers. A non-invasive method is to measure the strength of the inspiratory muscles in the mouth (maximum inspiratory pressure) and the nose [nasal inspiratory pressure during the maximum inspiration maneuver (SNIP)]. It is difficult to determine maximum inspiratory pressure in patients with bulbar involvement, as they have problems maintaining the mouthpiece in place and coordinating with the technician to perform the maneuver. This practical problem is overcome with the use of SNIP, which is an easier maneuver, the main limitation of which is nasal obstruction. SNIP is an easily performed technique, even in advanced stages of disease or in patients with bulbar involvement.[8] In ALS patients, SNIP correlates with nocturnal hypoventilation,[6] inspiratory muscle strength determined using invasive techniques,[6,11,8] and time to start MV.[11] Only Bauer et al. found that SNIP correlates poorly with disease severity in ALS patients.[7]

However, all the above-mentioned lung function tests require patient collaboration, so ultrasound evaluation of diaphragm function has been introduced in recent years. Diaphragm ultrasound, carried out in the zone of apposition, is useful for assessing movement and thickness of the diaphragm.[12,13] In ALS patients, diaphragm thickness measured by ultrasound correlates with lung function parameters, and even with hypercapnia.[14]

A fundamental factor in the follow-up of these patients is the detection of nocturnal hypoventilation, suggested by changes in nocturnal oximetry.[5] The more technically complex sleep studies would not be currently indicated in routine practice, despite the fact that ALS patients have changes in sleep

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architecture.[15] Transcutaneous capnography, used at night, represents an advance in the non-invasive monitoring of hypercapnia, and in the future it may be one of the procedures that will be added to the battery of tests that help improve the assessment of inspiratory muscle function. The increase in carbon dioxide measured by transcutaneous capnography might correlate inversely with vital capacity[16] in ALS patients, as well as with greater MV compliance.[17] Finally, analysis of arterial blood gases can be used to assess hypercapnia, although when hypercapnia is detected, the disease is already in an advanced stage with significant inspiratory muscle dysfunction.[5]

A group of researchers is currently exploring the combination of different parameters obtained from these functional tests in order to improve the assessment of inspiratory muscle function in patients with ALS.[11]

In conclusion, the correct evaluation of inspiratory muscle function is essential for indicating MV, the treatment that has been proven most effective for the survival of these patients. No single test is ideal for evaluating this function, so we must depend on a combination of several indicators, taking into account the specific dysfunctions of each patient during the course of their disease.

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