Relationships between Dyspnea, Respiratory Muscle Strength, and Ventilatory Failure in Patients with Amyotrophic Lateral Sclerosis

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Abstract. The purpose of this study was to investigate the relationships between dyspnea, respiratory muscle strength, and ventilatory failure in patients with amyotrophic lateral sclerosis (ALS). A cross-sectional study, based on patients’ medical records, was performed at a primary care hospital in Japan. Twenty-five patients diagnosed with ALS who received care as outpatients or inpatients were included in this study, and patients’ characteristics, dyspnea on the subscale of the revised ALS Functional Rating Scale, sniff nasal inspiratory pressure (SNIP), an indicator of respiratory muscle strength, and arterial carbon dioxide pressure (PaCO₂) were selected as main outcome measures. All data were collected from patients’ medical records. For patients with dyspnea, SNIP was significantly lower and PaCO₂ tended to be higher than in patients without dyspnea. There were no significant differences in both bulbar symptoms and types of disease onset between the patients with and without dyspnea. In patients with ALS, it is suggested that dyspnea reflects respiratory muscle weakness and ventilatory failure. For the assessment of respiratory function in patients with ALS, it may be important to evaluate dyspnea in addition to SNIP and PaCO₂.

Key words: Amyotrophic lateral sclerosis, Dyspnea, Respiratory function

INTRODUCTION

Amyotrophic lateral sclerosis (ALS), a disease of progressive degeneration in motor neurons, is a disease of uncertain cause. This progressive disease causes weakness not only in the limb and trunk muscles, but also in the respiratory muscles. Respiratory muscle weakness leads to decreased forced vital capacity (FVC) and increased residual volume (RV)1), and finally brings about ventilatory failure. Therefore, because respiratory function considerably affects the prognosis of patients with ALS, its maintenance by respiratory physical therapy plays an important role. Respiratory function may be measured as %FVC, sniff nasal inspiratory pressure (SNIP)2), and dyspnea on a subscale of the revised ALS Functional Rating Scale (ALSFRS-R)3). The %FVC is assumed to be the gold standard4), but its correlation with the degree of dyspnea is reportedly weak3). However, it is reported that dyspnea is associated with diaphragm dysfunction5). Therefore, the relationship between dyspnea and respiratory function in ALS is not yet clear. The purpose of this
The study was to investigate by cross-sectional analysis the relation between dyspnea and respiratory function in patients with ALS.

METHODS

2.1 Patients

Patients with a diagnosis of ALS whose disease progression had been followed at a primary care hospital between September 2005 and April 2006 were included in this study. Patients who had had a tracheostomy and complications, pulmonary disease, dementia, or other neurological diseases, and who were using an invasive positive pressure ventilator (IPPV) were excluded from the data analysis. However, patients who were using a non-invasive positive pressure ventilator (NIPPV) were included.

2.2 Data collection

A subscale of ALSFRS-R \(^3\) was used to assess dyspnea. This is a point distribution scale from 0 to 4 points according to the degree of dyspnea. The distribution point definition is as follows: 0 points is when breathing is significantly difficult, with the patient considering using mechanical respiratory support; 1 points is when dyspnea occurs at rest, with the patient having difficulty breathing when either sitting or lying; 2 points are when dyspnea occurs with at least one activity of daily living such as eating, bathing, or dressing; 3 points are when dyspnea occurs when walking; and 4 points are given for no dyspnea.

In measurement of SNIP, a plug (NPLG00; Micro Medical Ltd. United Kingdom.) made from polyethylene to which a catheter was connected was inserted into the nostril of each subject. Each subject was encouraged to relax and breathe such that the contralateral nostril was not occluded and the mouth was closed; the subject was then instructed to sniff with maximum effort from the functional residual capacity. The pressure generated during sniffing was measured five times by a respiratory pressure meter (RPM01; Micro Medical Ltd. United Kingdom.), with each test separated from the next by 30 seconds. The highest value was selected for later analysis.

Arterial carbon dioxide pressure (PaCO\(_2\)) was analyzed from laboratory reports. All data described above were collected from medical records. The protocol of this study conformed to the Helsinki Declaration.

2.3 Statistical analysis

Statistical analyses were performed using SPSS for Windows, version 10.0J (SPSS Japan Inc. Japan). An unpaired t-test was done for the comparisons of age, disease duration, SNIP, and PaCO\(_2\) between the group with dyspnea (dyspnea score<4) and the group without dyspnea (dyspnea score=4). In addition, onset of the disease and the presence of bulbar symptoms were compared by Fisher’s exact probability test between the two groups. Statistical significance was set at 5%.

RESULTS

This study included 25 patients who satisfied the inclusion criteria and for whom all data on dyspnea, SNIP, and PaCO\(_2\) were obtained. Subject characteristics were an average age of 62.9±10.1 years, average disease duration of 0.3–13.0 years,
and types of disease onset that were bulbar for 5 patients and limbs for 20 patients (see Table 1). When patients were grouped based on dyspnea, the group without dyspnea (dyspnea scale=4) had 11 cases and the group with dyspnea (dyspnea scale<4) had 14 cases. Comparing both groups, there were no significant differences in age, disease duration, types of disease onset, and bulbar symptoms, but in the group with dyspnea, SNIP was significantly lower and PaCO₂ tended to be higher than in the group without dyspnea [SNIP: 41.2 ± 23.7 vs. 23.7 ± 17.8 cmH₂O (p<0.05); PaCO₂: 45.3 ± 8.5 vs. 39.6 ± 4.6 cmH₂O (p<0.06)] (Table 1).

**DISCUSSION**

In this study, there were no significant differences between patients with dyspnea and those without dyspnea regarding age, disease duration, bulbar symptoms and disease onset, but the SNIP was significant lower and the PaCO₂ tended to be higher in patients with dyspnea. Sniff nasal inspiratory pressure was reported to correlate to transdiaphragmatic pressure⁶) and to predict hypercapnia⁷). Therefore, SNIP reflects respiratory muscle strength and PaCO₂ is an indicator of the degree of ventilatory failure. The above results suggest that the existence of dyspnea in patients with ALS is an indicator of respiratory muscle weakness and ventilatory failure. In conclusion, for the assessment of respiratory function in patients with ALS, it may be important to evaluate dyspnea in addition to SNIP and PaCO₂.

**REFERENCES**