



Stapedial Reflex in a Cohort of ALS Patients



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INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is a rare neurodegenerative disease, characterized by unknown etiology, different phenotypes and no effective treatment. About two thirds of cases have a spinal onset, while 30% of patients have a bulbar onset.

Several prognostic factors have been identified in ALS, but there is still a lack of clinical prognostic markers useful for care management and for clinical trials.

To our knowledge, the only study that analyzed impairment of the stapedius motor neurons in ALS reported that there may be a close relation between bulbar involvement and changes in the acoustic reflex in ALS.

The aim of this study was to investigate acoustic reflex in ALS patients and to found differences between patients with bulbar and spinal symptoms.

METHODS

- 51 ALS patients, selected from those followed-up in the Center for ALS of Molinette's Hospital in Turin, and 10 sex- and age-matched control subjects were recruited. Patients were further divided in two groups: ALS-B (38 cases, with bulbar signs at the time of evaluation) and ALS-S (13 cases, without bulbar signs at the time of evaluation).

- All patients underwent otoscopy, pure tone audiometry and tympanometry to exclude middle ear disease. The acoustic reflex was elicited in each ear by presenting a contralateral pure tone stimulus at 500–1000–2000–3000–4000 Hz, broadband noise (BBN) from 250 to 4000 Hz and low-pass noise (LPN) from 250 to 1800 Hz. Amplitude, latency and rise time of stapedial reflex were analyzed basing on graphics obtained.

- Statistical analysis was performed by Wilcoxon test and the level of significance was set at 5%.

- The acoustic reflex was measured using an impedance audiometer (Amplaid A724-A728, Amplifon, Milan, Italy).

RESULTS

Stapedial reflex was present in all patients. There was a statistically significant difference in the mean amplitude, latency and rise time between the ALS patients as compared with the controls. Amplitude was lower in both the ALS-B and the ALS-S patients than in the controls ($p < 0.05$) and rise time was longer in both patient groups compared with the controls ($p < 0.05$).

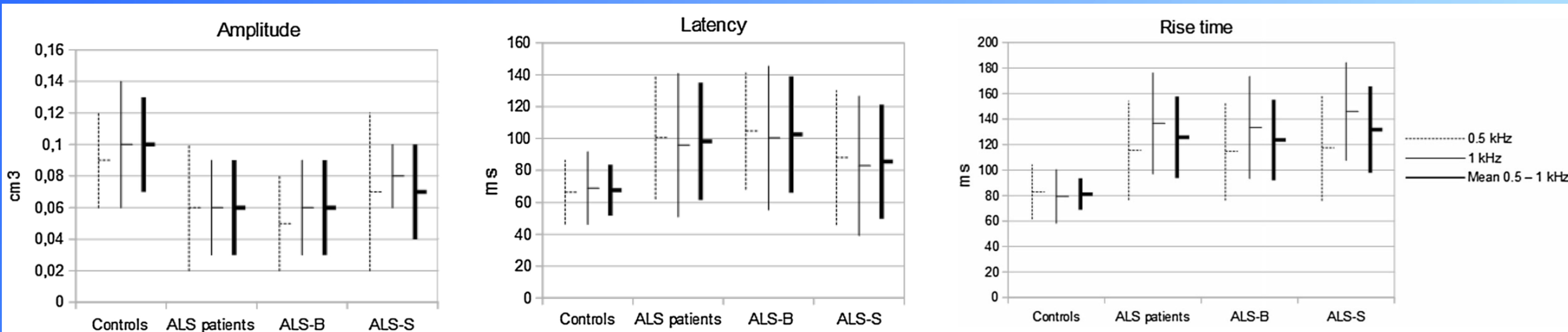


Fig. 1 Amplitude at single 0.5–1 kHz and at mean 0.5–1 kHz in the control group, ALS group, bulbar type (ALS-B) and spinal type (ALS-S); $p < 0.05$, except the difference at 1 kHz between ALS-S and controls ($p = 0.30$)

Fig. 1 Latency at single 0.5–1 kHz and at mean 0.5–1 kHz in the control group, ALS group, ALS-B and ALS-S group; $p < 0.05$, except the difference between ALS-S and control at 0.5–1 kHz and mean 0.5–1 ($p = 0.11$; $p = 0.44$; $p = 0.23$)

Fig. 3 Rise time at single 0.5–1 kHz and at mean 0.5–1 kHz in the control group, ALS group, ALS-B and ALS-S group; $p < 0.05$ except the difference at 0.5 kHz between ALS-S and controls ($p = 0.07$)

CONCLUSION

These results confirm the presence of abnormal acoustic reflex patterns in ALS cases with bulbar signs and, moreover, suggest that its alteration could show a possible subclinical involvement of the stapedial motor neuron even in ALS-S patients. Therefore this reflex could be precociously altered before the appearance of bulbar clinical signs. If this results will be confirmed in a future longitudinal study the stapedial reflex test could be used as a prognostic marker for progression of disease to the bulbar district in ALS-S patients.

DISCUSSION

In this paper authors observed a significant reduction in amplitude, a prolonged of latency and significantly longer contraction and relaxation times in bulbar ALS patients vs controls at 500 Hz and 1000 Hz. Besides looking at ALS-S patients we observed that there were statistically significant differences with the controls only in the amplitude of the reflex at 500 Hz and in its rise time at 1000 Hz. This suggests that the an involvement of the stapedius motor neurons or of the supranuclear stapedius motor system might be responsible for the abnormalities of the stapedial reflex in ALS. Moreover It seems to be partially altered in ALS-S patients who will develop bulbar sign shortly afterwards.

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