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# Presence and Severity of Language Deficits in Comprehension, Production and Pragmatics in a Group of ALS Patients: Analysis with Demographic and Neuropsychological Data

**Authors :** M. Testa, L. Peotta, S. Giusiano, B. Lazzolino, U. Manera, A. Canosa, M. Grassano, F. Palumbo, A. Bombaci, S. Cabras, F. Di Pede, L. Solero, E. Matteoni, C. Moglia, A. Calvo, A. Chio

**Abstract :** Amyotrophic Lateral Sclerosis (ALS) is a neurodegenerative disease of adulthood, which primarily affects the central nervous system and is characterized by progressive bilateral degeneration of motor neurons. The degeneration processes in ALS extend far beyond the neurons of the motor system, and affects cognition, behaviour and language. To outline the prevalence of language deficits in an ALS cohort and explore their profile along with demographic and neuropsychological data. A full neuropsychological battery and language assessment was administered to 56 ALS patients. Neuropsychological assessment included tests of executive functioning, verbal fluency, social cognition and memory. Language was assessed using tests for verbal comprehension, production and pragmatics. Patients were cognitively classified following the Revised Consensus Criteria and divided in three groups showing different levels of language deficits: group 1 - no language deficit; group 2 - one language deficit; group 3 - two or more language deficits. Chi-square for independence and non-parametric measures to compare groups were applied. Nearly half of ALS-CN patients (48%) reported one language test under the clinical cut-off, and only 13% of patents classified as ALS-CI showed no language deficits, while the rest 87% of ALS-CI reported two or more language deficits. ALS-BI and ALS-CBI cases all reported two or more language deficits. Deficits in production and in comprehension appeared more frequent in ALS-CI patients ($p=0.011$, $p=0.003$ respectively), with a higher percentage of comprehension deficits (83%). Nearly all ALS-CI reported at least one deficit in pragmatic abilities (96%) and all ALS-BI and ALS-CBI patients showed pragmatic deficits. Males showed higher percentage of pragmatic deficits (97%, $p=0.007$). No significant differences in language deficits have been found between bulbar and spinal onset. Months from onset and level of impairment at testing (ALS-FRS total score) were not significantly different between levels and type of language impairment. Age and education were significantly higher for cases showing no deficits in comprehension and pragmatics and in the group showing no language deficits. Comparing performances at neuropsychological tests among the three levels of language deficits, no significant differences in neuropsychological performances were found between group 1 and 2; compared to group 1, group 3 appeared to decay specifically on executive testing, verbal/visuospatial learning, and social cognition. Compared to group 2, group 3 showed worse performances specifically in tests of working memory and attention. Language deficits have found to be spread in our sample, encompassing verbal comprehension, production and pragmatics. Our study reveals that also cognitive intact patients (ALS-CN) showed at least one language deficit in 48% of cases. Pragmatic domain is the most compromised (84% of the total sample), present in nearly all ALS-CI (96%), likely due to the influence of executive impairment. Lower age and higher education seem to preserve comprehension, pragmatics and presence of language deficits. Finally, executive functions, verbal/visuospatial learning and social cognition differentiate the group with no language deficits from the group with a clinical language impairment (group 3), while attention and working memory differentiate the group with one language deficit from the clinical impaired group.

**Keywords :** amyotrophic lateral sclerosis, language assessment, neuropsychological assessment, language deficit
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