EXECUTIVE ABILITY AND DAILY LIVING PERFORMANCE IN PATIENTS WITH ALS

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Amyotrophic lateral sclerosis (ALS) was traditionally thought to affect solely the lower and upper motor neurons. Recent studies suggest that the pathogenic processes of ALS are more extensive, involving a wider dysfunction of cortical grey and white matter with clinical correlates in the impairment of cognitive abilities. Cognitive deterioration has until recently been associated almost exclusively with a subgroup of 3–5% of ALS patients with frontotemporal dementia. However, a number of latest neuropsychological investigations demonstrated selective impairments of the executive and memory functions in nondemented ALS patients (25–75%). The relevance of these results for everyday life, however, still remains in question, thus demonstrating the need for the development and use of an ecologically valid test (1).

The purpose of this preliminary study was to assess the prevalence of cognitive dysfunction in ALS and also dysfunction's impact on patients' daily activities. We evaluated 16 patients with ALS, according to El-Escorial Criteria of "possible", "probable" or "definite" ALS, and with a mean age of M=59.6~(SD=7.3) on a battery of standardized neuropsychological tests as well as a specially designed and ecologically valid test of executive functions called Medication Scheduling task – MST (2). With MST, patients are asked to coordinate multiple rules in order to create a safe daily schedule of medications.

Both, standardized neuropsychological measures as well as the ecologically valid measure of executive functions, revealed important deficits in cognitive performance of ALS patients. Using the cut-off value of the 15th percentile to mark the deficient activity (3), we found that most

patients demonstrated the same kind of impaired activity, i.e. placing incorrectly the pills to the medical schedule and neglecting to follow the rules while executing the MST task. The MST task's high demand for cognitive control, including the ability to coordinate multiple tasks and/or multiple sources of information, the application of strategies to a novel problem and the ability to monitor incoming information for relevance to the task at hand, seems to present great difficulties to patients with ALS and could possibly interfere with the planning and executing of their daily activities. Further explorations of patients' daily living performance are needed. Knowing whether or not an ALS patient has cognitive impairment brings up important considerations regarding the management of the patients, including a higher degree of involvement and supervision on the part of their caretakers.

References:

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