Cognitive and Behavioral Impairments in ALS – An Update

Summer 2007

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A great deal of attention has been paid lately to the issues of cognitive and behavioral dysfunction in ALS. The research on this has been fascinating, but patients and families often ask what this means for them. A brief review of this topic may help.

A 1998 paper (Neary D, et al. Neurology 1998;51:1546-1554) defined 3 subtypes of frontotemporal lobar degeneration (FTLD). The most common is frontotemporal dementia (FTD), manifested by a decline in social conduct, resulting in poor manners, poor social graces, and more passive or more outgoing/aggressive behavior. These changes, referred to as behavioral impairment, may be accompanied by cognitive impairment or "executive dysfunction," resulting in difficulty with abstraction, planning, and problem solving, and with a lack insight into deficits, so that the persons affected are unaware or unconcerned about these changes. The other two types of FTLD result in cognitive impairment manifested by language problems. Progressive nonfluent aphasia results in nonfluent speech containing grammatical errors, brief phrases in place of sentences, word-finding difficulties, and phonemic paraphasic errors, such as "parrot" for "carrot" or "robber" for "robin." Semantic dementia leaves the affected individual with speech which is fluent and grammatically correct, but with increasingly empty content because of progressively poorer ability to understand word meanings. Individuals often make semantic paraphasic errors, such as "sock" for "glove" or "cat" for "tiger." They may also develop difficulty recognizing familiar objects or faces.

It is important to understand that more than one of these subtypes often is present in an affected individual. Also, many individuals with ALS do not develop full-blown FTLD as defined by the Neary criteria, but have a lesser degree of cognitive or behavioral impairment which can be identified on special testing, but which may not be readily apparent otherwise. A paper in 2003 (Lomen-Hoerth C, et al, Neurology 2003;60:1094-1097) found that deficits of executive function are present in half of ALS patients, but that fewer have FTLD as defined in the 1998 paper.

We have recently developed a questionnaire which we administer in ALS clinic to screen individuals for cognitive impairment (Flaherty-Craig C, et al. Neurology 2006;67:2070-2072). Patients are asked to name or to write as many things as they can beginning with a particular letter within a specified period of time. These tests of verbal fluency are abnormal in many individuals with ALS. Patients are also asked questions which assess their abstract reasoning and judgment. All these are measures of executive function. When testing 110 patients with ALS, we found that deficiencies in verbal fluency, abstract reasoning, and judgment were found in 20.0%, 18.6%, and 35.7% of limb-onset patients, and in 37.5%, 25.0%, and 60.0% of bulbar-onset patients. This shows that a strikingly high number of individuals with ALS have deficits in problem-solving abilities as measured by abstract reasoning and judgment. In another study, we found that most individuals (59.2%) with ALS and

Simmons ALS Newsletter Column Summer 2007 deficient verbal fluency were deficient in measures of problem solving, and that most of those (72.4%) with intact verbal fluency scored average or above in measures of problem solving.

Tests of verbal fluency, abstract reasoning, and judgment are excellent ways to assess cognitive function, but what about behavior? We administer an instrument known as the Frontal Behavioral Inventory to caregivers of individuals with ALS. This instrument asks a series of questions to identify whether the individual with ALS is demonstrating behaviors of the type described by the Neary criteria. If the deviations from normal are severe enough, FTD can be diagnosed.

If individuals with ALS demonstrate abnormalities on our screens for cognitive or behavioral dysfunction, we offer them the opportunity to participate in more comprehensive testing. This includes a detailed assessment of language skills. Deficits of language can be striking in some individuals with ALS, demonstrating features of progressive nonfluent aphasia or semantic dementia, unrelated to their motor skills.

Why are these assessments important? After all, individuals with ALS and their caregivers often are struggling to cope with a host of physical limitations which impair independence, mobility, nutrition, and communication, and which require walkers, wheelchairs, adaptive equipment for activities of daily living, augmentative communication devices, and modifications in diet. My personal view is that cognitive and behavioral assessments are essential. Individuals and their families are asked to make critical decisions regarding their health care, including decisions about feeding tubes, non-invasive ventilation, tracheostomy/mechanical ventilation, and advance directives. If cognitive impairment is relatively common in ALS, then it is important to take this into account as these important decisions are made. We do not yet know the best way to help individuals with ALS and their caregivers manage important decisions in the face of such cognitive and behavioral changes, but we are designing approaches which we hope will be helpful and successful in achieving the best possible quality of life for these individuals and their caregivers.