

Cognitive Dysfunction in ALS

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Neurologists often speak of “cognitive function,” a term referring to memory, organization of thoughts, reasoning, judgment, concentration, and other higher cortical functions which determine the ability of individuals to think and express themselves clearly and logically. ALS is traditionally thought of as a disease which spares cognitive function, leaving the mind clear while causing progressive weakness. However, recent studies have shown that this is often not the case. Individuals with ALS may develop frontotemporal lobar degeneration (FTLD). This may take several forms: In frontotemporal dementia, individuals may exhibit uninhibited behavior, easy distractibility, loss of insight, and deficits in “executive function” such as attention, abstraction, planning, problem solving. In semantic dementia, individuals, persons with ALS may demonstrate difficulties with naming and word comprehension. Progressive nonfluent aphasia causes effortful speech production , grammatical errors, word retrieval difficulties, and problems reading and writing. These types of deficits are much different from those seen in Alzheimer’s disease, because memory appears to be well-preserved. That is most likely the reason that these cognitive problems have been under-recognized for many years.

How common is cognitive dysfunction in ALS? It appears to be common, but not universal. A recent study of patients with ALS (Lomen-Hoerth et al, Neurology 2003;60:1094-1097) demonstrated evidence of FTLD in 52%. This appeared to be more common in patients with bulbar involvement, but was seen in some without bulbar involvement as well. Moreover, there appears to be a range of cognitive dysfunction in ALS, varying from mild to severe. Conversely, a study of patients with frontotemporal dementia without a known diagnosis of ALS revealed that 14% had ALS when they underwent a thorough evaluation (Lomen-Hoerth et al, Neurology 2002;59:1077-1079). Another recent study in our ALS clinic by Dr. Anna Barrett and her assistant, Erin Zimmerman, looked at whether individuals with ALS could perceive emotions properly. 12 of 13 had difficulty with this task.

As you might imagine, impairments of cognitive function have profound implications with respect to the ability of a person with ALS to make critical decisions regarding medications, equipment, feeding tubes, ventilatory support, and a variety of end-of-life issues. Such deficits also have enormous implications for issues such as driving, managing finances, and

determining the need for supervision. It is important for the individual, the family, and health care professionals to be aware of these deficits, and, if they are present, of their severity. This is the basis for our recent testing of cognitive function in ALS clinic by Dr. Claire Flaherty-Craig. Although we have looked at our data for only 20 of our patients so far, it is becoming clear that cognitive dysfunction of some degree, while not present in everyone we have tested, is rather common. If such deficits exist, this impacts greatly on how family, other care providers, and the ALS team should work with the patient to assure safety, provide a supportive environment, and maximize quality of life.

It may be difficult for the individual or the family to acknowledge that these problems are developing. After all, ALS produces so many losses which require attention that it is easy to dismiss changes in behavior as resulting from the stress of the illness or from depression. Alternatively, many family members and caregivers assume that the problem is one of communication for an individual who has lost his or her ability to speak. However, the acknowledgment of such deficits, along with the development of strategies to cope with and manage them, is, I believe, a critical part of providing the best overall care for individuals with ALS. It is becoming increasingly clear that ALS is not a disease just of motor neurons. It is a degenerative neurological condition with other consequences as well. This broader understanding of ALS will help direct our research efforts more effectively. And, as we look for better treatments and eventually for a cure, it is important that we all understand the many faces of this disease, so that we may judge the ability of these treatments to address the varied deficits caused by ALS, all of which impact greatly on the individual with ALS, on family, on caregivers, and on the health care professionals who work with these individuals.