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Cognitive Impairment Appears to Be Common in ALS Patients

CHICAGO, I.L. -- MARCH 17, 2006 -- In a study of 40 patients with amyotrophic lateral sclerosis (ALS), about one-third showed evidence of cognitive impairment, but these deficits did not appear to be related to survival, according to a study in the March issue of Archives of Neurology, one of the *JAMA*/Archives journals.

ALS, commonly referred to as Lou Gehrig disease, is a progressive disorder characterized by the loss of muscle function and the atrophy (wasting away) of muscle tissue. ALS is primarily a disorder involving the motor neurons, which control muscles and movement in the body, but new evidence suggests it also may have an impact on cognition (thinking, learning and memory), according to background information in the article. Previous research has estimated that anywhere from 2 to 52% of patients with ALS also experience cognitive impairment.

Gregory A. Rippon, MD, MS, and colleagues at the Columbia University College of Physicians and Surgeons, New York, analyzed 40 consecutive patients with ALS who were evaluated at neurologists' offices between August 1991 and August 1992. Participants underwent examinations and testing to gauge their cognitive functioning and verify the diagnosis and history of their disease, including whether symptoms were first detected in muscles of the throat, jaw, tongue or face (bulbar onset) or those in the arms (limb onset). The researchers selected a control group of 80 individuals without ALS, matched to the ALS patients by age, gender and education, from a series of patients referred to a memory disorder clinic from 1992 to 2003.

Of the 40 patients with ALS, 12 (30%) showed evidence of cognitive impairment, including nine (23%) who met criteria for dementia. There were no significant differences between ALS patients who had dementia and those who did not in terms of age, sex, education, site of onset, memory loss, emotional stability, severity of the disease or family history. ALS patients and control participants had similar results on cognitive tests, although patients with more severe ALS showed a decline in verbal skills beyond what would be associated with motor difficulties affecting speech muscles. Survival data from public and medical records were available in January 2004 for 38 of 40 patients with ALS, who lived an average of 3.4 years after testing. Cognitive impairment and dementia did not appear to be associated with survival.

"In conclusion, using a conventional test battery, 30% of a consecutive series of patients with ALS demonstrated cognitive impairment, and nearly a quarter qualified for a neuropsychologic diagnosis of dementia," the authors write. "Free recall, executive function and naming were most impaired in ALS patients with dementia." Future studies using testing and diagnostic criteria specific to frontotemporal lobar dementia, the type believed to be associated with ALS and other motor neuron diseases, may find that the percentage of ALS patients with cognitive impairment or dementia is even higher, they conclude.

This study was supported in part by Ruth L. Kirschstein National Research Service Award from the National Institutes of Health, Bethesda, Md.

Editorial: A New Understanding of ALS

Even though ALS was previously thought to affect only the motor system, physicians are increasingly recognizing that the symptoms are much broader, writes Michael J. Strong, MD, FRCPC, University Hospital, London, Ontario, Canada, in an accompanying editorial.

These revelations lead to questions about whether information about cognitive impairment is clinically relevant to patients with ALS and also raises controversy about the biological mechanisms involved in the disease, Dr. Strong writes. "It behooves us to understand the nature of this process in detail to truly understand the nature of this aspect of ALS, its influence not only on survival but also on the quality of life of patients with ALS and for their caregivers, and also to shed light on the biological nature of this process," he concludes.

Arch Neurol. 2006;63:345-352.
Arch Neurol. 2006;63:319-320.
SOURCE: American Medical Association