Abstracts

Articles appearing in the December 2017 issue

Progression and effect of cognitive-behavioral changes in patients with amyotrophic lateral sclerosis

Background To prospectively evaluate the progression of cognitive-behavioral function in amyotrophic lateral sclerosis (ALS) and examine the association of cognitive-behavioral deficits with disease progression, patient quality of life (QOL), and caregiver burden.

Methods We evaluated cognitive-behavioral function using the Amyotrophic Lateral Sclerosis Cognitive Behavioral Screen at enrollment and after 7 months in a cohort of patients with ALS. Paired *t* tests were used to evaluate the change in the 2 assessments. Linear regression and Kruskal-Wallis tests were applied to investigate how initial cognitive or behavioral status related to outcomes.

Results The mean test-retest interval was 6.8 months (SD 1.6). Cognitive status of the study population (*n* = 49) overall did not change over the study period (*p* = 0.06) despite progression of motor weakness (*p* < 0.001), though small subsets of the sample demonstrate cognitive change. Patients initially classified as behaviorally normal showed increased behavioral problems over time (*t* = -2.8, *p* = 0.009). Decline in cognitive (β = -1.3, *p* = 0.03) and behavioral (β = -0.76, *p* = 0.002) status predicted increasing caregiver burden. Behavioral abnormalities predicted decline in forced vital capacity and ALS Functional Rating Scale-Revised score (*p* = 0.008, 0.012) in the study population and patient QOL in the most severely affected group (*t* = 4.3, *p* = 0.003).

Conclusions Cognitive-behavioral change is a key aspect of disease heterogeneity in ALS. Executive function in ALS overall remains stable over 7 months as detected by an administered screening tool. However, patients may develop caregiver-reported behavioral symptoms in that time period. Screening for caregiver-reported symptoms has a particular utility in predicting future clinical decline, increased caregiver burden, and worsening patient QOL.

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“Think metabolic” in adults with diagnostic challenges: Biotinidase deficiency as a paradigm disorder

Neurologists should consider the possibility of an inherited metabolic disorder in adults with neurologic symptoms that may or may not mimic those seen in affected children, such as in the case of biotinidase deficiency. Because many of these disorders are treatable, they must be included in the differential diagnosis. Technologies, such as specific biochemical analysis and whole exomic sequencing, can assist the clinician by leading to the appropriate diagnosis and treatment. Whole exomic sequencing can identify known and putative mutations in a patient's genome. The neurologist must “think metabolic” in sorting out complex and difficult cases.

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Practice Current

The survey on our latest topic, “When do you suspect autoimmune encephalitis and what is the role of antibody testing?”, has received over 850 responses from over 80 countries to date. Explore this topic and others on our newly redesigned website; compare your practice with peers and see real-time results displayed on an interactive world map.

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