In Focus

Spotlight on the August 10 Issue

Robert A. Gross, MD, PhD, FAAN
Editor-in-Chief, *Neurology*®

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REM sleep behavior disorder preceding other aspects of synucleinopathies by up to half a century

The authors used medical records to identify cases meeting the criteria of "idiopathic" REM sleep behavior disorder (RBD) at onset and at least 15 years between RBD diagnosis and the development of other neurodegenerative symptoms. These cases show that the α-synuclein pathogenic process may start decades before the first symptoms of neurodegenerative diseases.

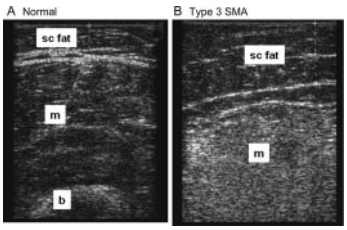
See p. 494; Editorial, p. 488

Mild cognitive impairment in prediagnosed Huntington disease

Using baseline data from 160 non–gene-expanded comparison participants, normative data were established for cognitive tests of episodic memory, processing speed, executive functioning, and visuospatial perception. Consistent with the Alzheimer disease literature, mild cognitive impairment in the prodromal period may occur in pre-Huntington disease.

See p. 500; Editorial, p. 490

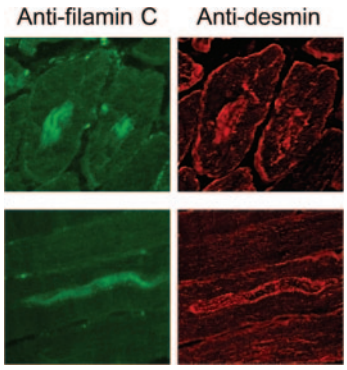
Assessing spinal muscular atrophy with quantitative ultrasound



Twenty-five patients with spinal muscular atrophy were compared to 21 normal subjects after study of the biceps brachii, wrist extensors, quadriceps, and tibialis anterior at their midpoints, in a relaxed posture, on one arbitrarily chosen side. The investigators showed that quantitative ultrasound may be useful in assessing other neuromuscular disorders and deserves further investigation.

See p. 526

Dominant-negative effects of a novel mutation in the filamin myopathy



A liquid chromatographic and mass spectrographic analysis on 6 Japanese patients with dominantly inherited myofibrillar myopathy showed dominant-negative effects of the *FLNC* mutation and the mutation-specific nature of those effects. This likely explains the variation in the clinical phenotypes in filamin myopathy.

See p. 547

SPECIAL ARTICLE

Invited Article: Comparative effectiveness research, evidence-based medicine, and the AAN

Comparative effectiveness research is necessary. However, it is important to understand the relative strengths and potential weaknesses of this type of research and to contribute to the discussion about how it should and should not be done.

See p. 562

Editorial quote from Dorsey and Meltzer: "The challenges (and controversies) with CER are thus not only in performing the research but also in applying its results."

See p. 492

RESIDENT & FELLOW SECTION

Clinical Reasoning: A 16-year-old boy with freezing of gait

Freezing of gait is an uncommon presentation in the setting of generalized dystonia. Here, the authors treated a 16-year-old with freezing of gait with anticholinergics, who had significant improvement of limb and trunk dystonia.

See p. e23

NB: "The neurologist in Charles Dickens. . ." and other Reflections.

See p. 571

Podcasts can be accessed at www.neurology.org

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