In Focus

Spotlight on the November 19 Issue

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Episodic weakness due to mitochondrial DNA MT-ATP6/8 mutations

This article shows that acute episodes of limb weakness mimicking periodic paralysis that dramatically respond to acetazolamide may be due to homoplasmic mitochondrial DNA MT-ATP6/8 mutations. It is important for diagnosis because of the maternal inheritance of these diseases. Further, the findings illustrate mitochondrial effect on plasma membrane ion transport.

See p. 1810

From editorialists Ruff & Cannon: "...increasing inward rectifier potassium channel current may be the mechanism of action of acetazolamide-induced improvement of symptoms in hypokalemic periodic paralysis."

See p. 1806

Fibroblast growth factor 21 is a sensitive biomarker of mitochondrial disease

The authors recruited 54 adult patients with mitochondrial disease, 20 patients with nonmitochondrial neuromuscular disease, and 66 controls. Serum fibroblast growth factor 21 levels are a sensitive biomarker and predictor of mitochondrial disease when compared to serum levels of classical indicators: creatine kinase, lactate, pyruvate, and the lactate to pyruvate ratio.

See p. 1819; Editorial, p. 1808

Residual fatigue in Guillain-Barré syndrome is related to axonal loss

Thirty-nine patients diagnosed with Guillain-Barré syndrome (GBS) were examined and divided into subgroups based on the presence of fatigue. All patients underwent standard nerve conduction studies and motor unit number estimation. Severe fatigue after GBS was related to greater axonal loss, represented by lower motor unit number estimations and lower sensory nerve action potentials.

See p. 1827

Classification and pathology of primary progressive aphasia

The authors determined clinicopathologic relationships in primary progressive aphasia (PPA) using clinical records from a cohort of autopsy-confirmed cases and raters blinded to postmortem diagnosis.

Meeting criteria for the semantic variant of PPA predicted TDP-43 type A pathology, but other PPA subtypes did not permit accurate pathologic prediction.

See p. 1832

Patterns of subregional mesiotemporal disease progression in temporal lobe epilepsy

The progression of mesiotemporal atrophy highlights the importance of early surgery in patients with drug-resistant temporal lobe epilepsy. Mapping the regional distribution of atrophy in single patients may help decide the extent of resection, have value as a biomarker for postsurgical outcome, and provide rates of contralateral atrophy to predict seizure relapse.

See p. 1840

Clinically meaningful performance benchmarks in MS: Timed 25-Foot Walk and the real world

Candidate Timed 25-Foot Walk (T25FW) benchmarks were validated in 95 participants using 13 measures of ambulation and cognition, patient-reported outcomes, and optical coherence tomography, with 2 clinically meaningful benchmarks identified. However, larger studies are needed to confirm clinical relevance and to decide whether there could be additional benchmarks in the lower and higher ranges of performance.

See p. 1856

Spinal cord injury and type 2 diabetes: Results from a population health survey

Data were obtained on 60,678 respondents from the Statistics Canada 2010 Cycle of the cross-sectional Canadian Community Health Survey. Analyses adjusting for both sex and age revealed a 2-fold increased odds of type 2 diabetes among individuals with spinal cord injury, suggesting the need for targeted interventions and prevention strategies.

See p. 1864

NB: "See-saw nystagmus," see p. e159. To check out other Resident & Fellow Teaching Video NeurolImages, point your browser to www.neurology.org and click on the link to the Resident & Fellow Section.

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