In Focus

Spotlight on the December 5 issue

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Notable in *Neurology*

This issue features an article that identifies *CTNND2* as the causative gene in familial cortical myoclonic tremor and epilepsy and another that describes acute illness-associated weakness in patients with dystroglycanopathy, compared to patients with Duchenne-Becker muscular dystrophy. A featured Views & Reviews article summarizes the 2016 Alzheimer's Disease-Related Dementias Summit.

ARTICLES

Population-based study of ischemic stroke risk after trauma in children and young adults

In this study, children and young adults who experienced arterial ischemic stroke within 4 weeks of trauma were identified and compared to controls. Ischemic stroke incidence was 4 strokes per 100,000 during the first 15 days. Stroke onset was frequently delayed after trauma, providing a time window for stroke prevention.

See p. 2310

From editorialists Jordan & Bhattacharya: "This work represents an excellent first step toward understanding stroke incidence and timing after trauma as well as the limitations of acute cerebrovascular imaging."

See p. 2306

Lower cardiac index levels relate to lower cerebral blood flow in older adults

Autoregulatory mechanisms may preserve cerebral blood flow in the presence of cardiac output fluctuations. However, the authors found lower cardiac index values related to lower cerebral blood flow, as measured using arterial spin labeling. Autoregulation may be compromised in aging adults with a lifetime burden of vascular risk factor exposure.

See p. 2327

SNPs in A β clearance proteins: Lower CSF A β ₁₋₄₂ levels and earlier onset of dementia in PD

The authors investigated the influence of β -amyloid (A β) clearance genetic variations on dementia in patients with Parkinson disease. *APOE* and *cystatin C3* single nucleotide polymorphisms were independently associated with clinical and biochemical dementia markers, suggesting that A β clearance mechanisms play a role in the pathogenesis of Parkinson dementia.

See p. 2335; Comment p. 2339

Long-term benefit of enzyme replacement therapy in Pompe disease: A 5-year prospective study

Data are lacking on the long-term efficacy of enzyme replacement therapy in adult patients with Pompe disease. This study showed that long-term treatment positively affects muscle strength, pulmonary function, and daily-life activities, irrespective of sex, age, or disease duration. Individual patient outcomes differed; elucidation of treatment-modifying factors is still needed.

See p. 2365

NB: "Teaching Neurolmages: A rare case of giant perivascular spaces in the midbrain manifesting as atypical parkinsonism," p. e272. To check out other Resident & Fellow Teaching Neurolmages, point your browser to Neurology.org and click on the link to the Resident & Fellow Section. At the end of the issue, check out the Clinical/Scientific Note discussing mutation of the *HTRA1* gene in 3 CARASIL families. This week also includes a Neurolmage titled "Visualization of a spinal capillary telangiectasia with 2D DSA and cone-beam CT."

Podcasts can be accessed at Neurology.org

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