



## In Focus

### Spotlight on the January 25 Issue

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Editor-in-Chief, *Neurology*®

#### Olfaction in *Parkin* heterozygotes and compound heterozygotes: The CORE-PD study



The authors administered a smell identification test to 44 probands with Parkinson disease (PD) onset before age 50 years (10 *Parkin* mutation heterozygotes, 9 compound heterozygotes, 25 noncarriers) and 80 family members (18 heterozygotes, 2 compound heterozygotes, 60 noncarriers). Olfaction was reduced among *Parkin* mutation heterozygotes with PD but not among their heterozygous relatives without PD.

See p. 319; Editorial, p. 312

#### Myasthenic syndrome caused by plectinopathy

Defects in plectin cause muscular dystrophy, myasthenic syndrome, and epidermolysis bullosa simplex (EBS). This paper identifies frameshifting mutations of plectin in 2 patients with EBS and traces the myasthenia to degeneration of the postsynaptic folds and the dystrophy to unmooring of fiber organelles and sarcolemmal calcium leaks, as in Duchenne dystrophy.

See p. 327

#### Muscle histology vs MRI in Duchenne muscular dystrophy



Analysis of muscle involvement on MRI was used to diagnose neuromuscular diseases.

The results provide a detailed correlation between muscle histology

and MRI changes in Duchenne dystrophy and demonstrate the value of this MRI technique as a reliable tool for the selection of patients recruited into clinical trials.

See p. 346

#### Enrollment of women and minorities in NINDS trials



The authors assessed enrollment of minorities in 56 NINDS-funded phase III trials. Reporting of minorities in trial publications was poor and Hispanic Americans were underrepresented. To improve the generalization of trial results, enrollment of Hispanic Americans, the largest minority group in the United States, needs to be increased.

See p. 354; Editorial, p. 314

#### $\gamma$ -Secretase-dependent amyloid- $\beta$ is increased in Niemann-Pick type C: A cross-sectional study



The fact that amyloid metabolism is affected in Niemann-Pick type C disease may be important for our understanding of lysosomal storage disorders, where there is increasing evidence for lysosomal dysfunction in the pathogenesis. This paper showed that it is possible to use CSF biomarkers to monitor disease processes in lysosomal storage disorders.

See p. 366

*From editorialist Edward M. Kaye: "The article by Mattsson et al. published in this issue of Neurology® may have added some insights into the neuropathologic processes in NPC by reporting that  $\gamma$ -secretase-dependent amyloid is increased in the CSF of patients with NPC."*

See p. 316

#### Child development following in utero exposure: Levetiracetam vs sodium valproate



The authors recruited children aged <24 months and assessed them using the Griffiths Mental Development Scale. Children exposed to levetiracetam in utero were not at an increased risk of delayed early cognitive development; therefore, levetiracetam may be a preferable drug choice for women with epilepsy and of childbearing age.

See p. 383

#### VIEWS & REVIEWS

##### White matter synapses: Form, function, and dysfunction

This review highlights the evidence for synaptic transmission in white matter, its potential function, and the importance of this new physiology in understanding common neurologic diseases and how it may be the key in translating bench science to bedside.

See p. 397

**NB:** Be sure to check out the next trial issue of *Neurology: Clinical Practice*, which will accompany your February 15 issue of the journal. Please take a few minutes to look it over and provide feedback. Did you know the journal is celebrating 60 years of publishing? Check out the first article, published back in 1951, which was reprinted in the January 4 issue.

Podcasts can be accessed at [www.neurology.org](http://www.neurology.org)

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