



In Focus

Spotlight on the March 19 Issue

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Novel brain expression of CIC-1 chloride channels and enrichment of CLCN1 variants in epilepsy

The chloride channel responsible for myotonia, long believed to reside only in muscle, is also found in brain. This study illustrates how exome screenings lead to the discovery of shared CNS expression of voltage-gated ion channels in disparate tissues, pointing to interesting and sometimes occult comorbid neurologic syndromes.

See p. 1078

From editorialists Berkovic & Kapur: "Perhaps patients with myotonia should be carefully screened for epilepsy. In addition, perhaps patients with idiopathic generalized epilepsy should be screened for subtle myotonia to look for those carrying CLCN1 gene mutation."

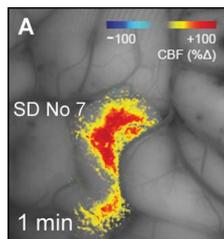
See p. 1074

One-year safety and tolerability profile of pridopidine in patients with Huntington disease

In this trial, patients received pridopidine 45 mg/day for 4 weeks then pridopidine 90 mg/day for 22 weeks; adverse events were recorded. Pridopidine (≤ 90 mg/day) was generally safe and well-tolerated in patients with Huntington disease for up to 1 year.

See p. 1086

Propagation of cortical spreading depolarization in the human cortex after malignant stroke



In 7 of 20 patients, 19 blood flow changes typical of cortical spreading depolarizations occurred during a 20-minute period.

Thirteen events were characterized by increase, 2 by biphasic response, and 4 by decrease of blood flow. In patients with focal ischemia, cortical spreading depolarizations were associated with both unfavorable and

protective hemodynamic responses.

See p. 1095

Vestibular compensation in acute unilateral medullary infarction: FDG-PET study

Twelve patients with circumscribed unilateral medullary brainstem infarctions (6 right, 6 left) causing acute vestibular imbalance underwent resting-state ¹⁸F-FDG-PET. Different compensation strategies seem to be apparent: after vestibular nucleus lesions, compensation occurs preferably in brainstem-cerebellar loops; after peripheral lesions, it occurs at the cortical level.

See p. 1103

Bumetanide prevents transient decreases in muscle force in murine hypokalemic periodic paralysis

Abortive or preventive therapies for transient weakness in periodic paralysis have limited effectiveness and potential side effects. Na-K-2Cl transporter inhibition with bumetanide stabilized muscle resting potential and prevented loss of force, promoting recovery of force in a mouse model of hypokalemic periodic paralysis.

See p. 1110

Cortical atrophy in ALS is critically associated with neuropsychiatric and cognitive changes

This study characterized the patterns of brain atrophy in 22 patients with amyotrophic lateral sclerosis (ALS), 17 patients with ALS-frontotemporal dementia (FTD), and 18 controls, following current ALS and FTD criteria. Cortical atrophy was only apparent in patients with ALS patients with neuropsychiatric and cognitive changes; however, observed atrophy was not as widespread as in ALS-FTD.

See p. 1117

Prediction of Alzheimer disease in subjects with amnestic and nonamnestic MCI

This study compared the predictive accuracy of β -amyloid 1-42 and tau in CSF, hippocampal volume, and APOE genotype for Alzheimer disease (AD)-type dementia in patients with amnestic and nonamnestic mild cognitive impairment (MCI). The findings suggest that AD biomarkers were useful predictors of AD-type dementia in both MCI subgroups.

See p. 1124

Autoimmune chorea in adults

Chorea in adults frequently has a neurodegenerative cause. This study demonstrated autoimmune chorea was half as common as chorea in Huntington disease, and was often treatable. Subgroup comparisons revealed that patients with paraneoplastic chorea were more frequently older, male, and with severe weight loss and coexisting peripheral neuropathy than patients with idiopathic autoimmune chorea.

See p. 1133

NB: "Should clinicians care about preclinical animal research?" see p. 1072. To check out this editorial, point your browser to <http://www.neurology.org>.

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