In Focus

Spotlight on the November 15 Issue

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

Incidence and predictors of acute symptomatic seizures after stroke

The authors followed 714 patients with first stroke, looking for seizures during the first week. Seizures presented in 6.3% overall: in 4.2% with ischemic infarctions, 12.5% with hemorrhagic infarctions, and 16.2% with primary intracerebral hemorrhage. The incidence of early seizures in stroke patients was higher than expected, particularly in those with cortical lesions.

See p. 1785; Editorial, p. 1776; see also p. 1794

Early seizures in intracerebral hemorrhage: Incidence, associated factors, and outcome

The authors recruited 562 patients with a spontaneous intracerebral hemorrhage (ICH). Early seizures were defined as occurring within 7 days of stroke onset, and their associated factors were identified with Cox regression. Early seizures were a frequent complication in patients with spontaneous ICH; however, their occurrence did not influence outcome at 6 months.

See p. 1794; Editorial, p. 1776; see also p. 1785

Quantifying physical decline in juvenile neuronal ceroid lipofuscinosis (Batten disease)

Although juvenile neuronal ceroid lipofuscinosis (JNCL) is a rare neurodegenerative disorder, the authors followed 82 patients using the Unified Batten Disease Rating Scale (UBDRS). The data support the use of the UBDRS to quantify the rate of progression of physical impairment in JNCL subjects in clinical trials.

See p. 1801; Editorial, p. 1779; see also p. 1808

Quantitative telemedicine ratings in Batten disease: Implications for rare disease research

Patients who had clinical characteristics of JNCL and mutations in both alleles of *CLN3* were recruited sequentially over a 5-month period. Remote administration of the Unified Batten Disease Rating Scale, an outcomes measurement tool in Batten disease, was reliable and feasible for these patients.

See p. 1808; Editorial, p. 1779; see also p. 1801

Clinical severity and quality of life in children and adolescents with Rett syndrome

Cross-sectional and longitudinal analyses were conducted on data collected from an NIH-sponsored Rett syndrome (RTT) natural history study of more than 200 participants, 5–18 years of age, with classic RTT. As clinical trials in RTT emerge, the CHQ-PF50 may represent one of the important outcome measures.

See p. 1812

From editorialist Susan L. Hyman: "The family-centered, community-based, coordinated approach to care for children with disabilities has been a culture shift within medicine over the past 24 years. It calls for families both to partner in decision-making with their health care providers and be satisfied with the services their children receive."

See p. 1781

Anti-MuSK autoantibodies block binding of collagen Q to MuSK

The authors obtained peripheral blood from 4 MuSK-MG patients and residual plasmapheresis fluid from a patient with limb-girdle muscular dystrophy.

Autoantibodies inhibited binding of collagen Q to MuSK that leads to

acetylcholine receptor (AChR) clustering. AChR deficiency observed in MuSK-MG may result from this mechanism.

See p. 1819; Editorial, p. 1783

Intravenous dihydroergotamine for inpatient management of refractory primary headaches

Inpatient use of dihydroergotamine produces substantial improvement in more than 80% of patients with otherwise medically refractory chronic migraine. Increasing dose through a 5-day course and careful control of nausea were predictors of better outcome.

See p. 1827

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