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Notable in *Neurology* this week

This issue features an article that investigates brain regions underlying interictal generalized paroxysmal fast activity in patients with Lennox-Gastaut syndrome; another determines that pathogenic variants of *DYNC2H1*, *KIAA0556*, and *PTPN11* are associated with hypothalamic hamartoma. A featured Views & Reviews article examines the link among *STXBPI* function, early infantile epileptic encephalopathy, and the neurodegenerative synucleinopathies.

Articles

SUDEP in the North American SUDEP Registry: The full spectrum of epilepsies

This study of 237 SUDEPs highlights that all ages and all epilepsy severities are affected, including patients with well-controlled or benign epilepsies. SUDEP is not limited to patients with chronic treatment-resistant epilepsy. Further, most SUDEP cases did not take their last dose of antiseizure medication, suggesting a target for prevention.

Page 98

From editorialist Burneo: "The important take-home message from this study is that from now on, clinicians should start conversations about SUDEP with their patients with epilepsy earlier rather than later."

Page 93

Midlife adiposity predicts cognitive decline in the prospective Multicenter AIDS Cohort Study

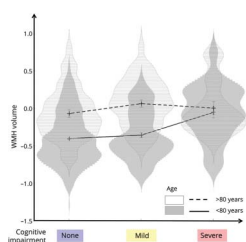
Addressing midlife obesity, a modifiable vascular risk factor, may prevent cognitive decline. In a prospective study of 972 HIV-infected and HIV-uninfected men, higher midlife total and central obesity were related to lower cognition and greater cognitive decline, particularly among HIV-uninfected men. Midlife obesity may predict neurologic outcomes and provide intervention possibilities.

Page 101

From editorialists Cysique et al.: "The ultimate strength of this study is to generate new hypotheses and suggest a more complex story about body weight and brain health connections."

Page 95

Age-dependent association of white matter abnormality with cognition after TIA or minor stroke



White matter hyperintensities (WMH) are frequently reported in elderly patients who undergo MRI. This study demonstrates that WMH are strongly related to cognition in patients with TIA/minor stroke at younger ages, but not at age >80. High WMH loads in patients aged >80 should not overly concern clinicians and patients.

Page 102

MORE ONLINE

Editor's Summary

Audio summary of highlighted articles.

NPub.org/edsum

Continued

Sporadic late-onset nemaline myopathy: Clinical spectrum, survival, and treatment outcomes

This article highlights the clinical characteristics and long-term outcomes of patients with sporadic late-onset nemaline myopathy (SLONM). Seventy-eight percent of patients responded to IV immunoglobulin and 75% to hematologic therapy, with good overall survival regardless of the presence of a monoclonal protein. IV immunoglobulin should be a first-line therapy in all patients with SLONM.

Page 105

NB: “The two lives of neurologist Helmut J. Bauer (1914–2008): Renowned MS specialist and National Socialist,” p. 109. To check out other Historical Neurology articles, point your browser to Neurology.org/N. At the end of the issue, check out the Resident & Fellow Teaching Video NeuroImage illustrating hepatic myelopathy. This week also includes a Resident & Fellow Clinical Reasoning paper titled “A misdiagnosis of atypical trigeminal neuralgia.”

NEW EPISODE



July 16, 2019

CME Opportunity:

Listen to this week's *Neurology* Podcast and earn 0.5 AMA PRA Category 1 CME Credits™ by answering the multiple-choice questions in the online Podcast quiz.

SUDEP in the North American SUDEP Registry: The full spectrum of epilepsies (see p. 98)

1. SUDEP in the North American SUDEP Registry: The full spectrum of epilepsies
2. What's Trending: Long-term effect of thymectomy plus prednisone versus prednisone alone in patients with non-thymomatous myasthenia gravis: 2-year extension of the MGTX randomised trial (article published in *The Lancet: Neurology*)

In the first segment, Dr. Halley Alexander talks with Dr. Orrin Devinsky about his paper on the full spectrum of epilepsies and the North American SUDEP Registry. In the second part of the podcast, Dr. Ted Burns focuses his discussion with Dr. Gil Wolfe on his *Lancet: Neurology* paper on the long-term effect of thymectomy plus prednisone vs prednisone alone in patients with nonthymomatous myasthenia gravis. You can read the paper here: [https://www.thelancet.com/journals/laneur/article/PIIS1474-4422\(18\)30392-2/fulltext](https://www.thelancet.com/journals/laneur/article/PIIS1474-4422(18)30392-2/fulltext).

Disclosures can be found at Neurology.org.

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Spotlight on the July 16 issue

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