In Focus

Spotlight on the January 28 Issue

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C9orf72 expansions are the most common genetic cause of Huntington disease phenocopies

The *C9orf72* expansion mutation was identified in 10 of 514 patients (1.95%) with Huntington disease (HD) phenocopy presentations. This suggests that neurologists should consider and test for *C9orf72* expansions in those with HD-like disorders, and, more broadly, demonstrates that *C9orf72* has a more extensive phenotype than previously identified, including movement disorders.

See p. 292; Editorial, p. 286

Withdrawing amantadine in dyskinetic patients with Parkinson disease: The AMANDYSK trial

This study assessed the long-term efficacy of chronic treatment with amantadine in 57 dyskinetic patients with Parkinson disease (PD). Unified Parkinson's Disease Rating Scale dyskinesia subscore (items 32 and 33) deteriorated more in patients switched to placebo compared with those maintained on amantadine. Washing out amantadine in dyskinetic patients with PD worsened levodopa-induced dyskinesia (LID) but had no effect on motor parkinsonian symptoms.

See p. 300

From editorialists Rodnitzky & Narayanan: "…these results should help put to rest any long-standing concerns about a uniformly short duration of amantadine's effect on LID and help inform practitioners not to preemptively and unnecessarily discontinue the drug."

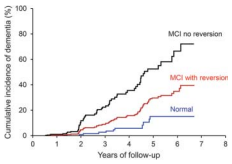
See p. 288

Characterizing mild cognitive impairment in incident Parkinson disease: The ICICLE-PD Study

Two hundred nineteen patients with Parkinson disease (PD) and 99 controls underwent structural MRI, lumbar puncture, and genotyping for common variants of *COMT*, *MAPT*, *SNCA*, *BuChE*, *EGF*, and *APOE*. CSF β-amyloid 1-42 and β-amyloid 1-40 levels were lower in those with PD-mild cognitive impairment and may serve as a biomarker of cognitive decline.

See p. 308

Higher risk of progression to dementia in mild cognitive impairment cases who revert to normal



Five hundred thirty-four participants were evaluated at baseline and every 15 months to identify incident mild cognitive impairment (MCI) or dementia, with 153 progressing to dementia after a median of 5.1 years. Persons with MCI, including those who reverted to cognitively normal, have a high risk of progressing to dementia, suggesting that diagnosis of MCI has prognostic value.

See p. 317; Editorial, p. 290

Alcohol consumption and cognitive decline in early old age

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The authors showed that consuming ≥36 g of alcohol (1 drink = 10 g) per day was associated with accelerated cognitive decline in men, based on 7,000 adults assessed 3 times over 10 years. This study provides robust evidence of the adverse effects of heavy alcohol consumption on cognitive aging.

See p. 332

Eighteen-year trends in stroke mortality and the prognostic influence of comorbidity

The authors identified all 219,354 patients with a first-time hospitalization for stroke in Denmark during 1994–2011, computing standardized 30-day, 1-year, and 5-year mortality by sex. Stroke mortality improved considerably between 1994 and 2011 for both ischemic stroke and intracerebral hemorrhage, but comorbidity remained a strong prognostic factor.

See p. 340

High-dose midazolam infusion for refractory status epilepticus

Status epilepticus frequently does not respond to conventional antiepileptic treatments, which is associated with poor outcomes. This cohort study demonstrates that high doses of midazolam infusions (up to 2.9 mg/kg/h) can safely be administered in an intensive care unit setting with good efficacy and possibly better outcome.

See p. 359

NB: "A 63-year-old man with progressive proximal pain and weakness," see p. e26. To check out other Resident & Fellow Mystery Cases, point your browser to www.neurology.org and click on the link to the Resident & Fellow Section.

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