

Treatment of tics

Two articles and an accompanying editorial consider new treatment strategies for tics. The Singer et al. (p. 599) controlled trial of baclofen for tics in Tourette syndrome showed significant benefit to tic severity, although the effect was greatest on symptoms associated with tics. ♦ The Marras et al. (p. 605) randomized double-blind controlled trial of botulinum toxin found a significant benefit to motor tic frequency and the associated urge to tic. ♦ The accompanying editorial by Kurlan (p. 580) notes that Tourette syndrome is common (3% in children) and reviews these new treatments in the context of other available treatments: alpha adrenergic agonists (e.g., clonidine) remain first-line treatment.

Rett syndrome (RS): Clinical and molecular features

Auranen et al. (p. 611) report *MECP2* gene sequence analysis in 39 patients with classic RS, 1 with RS with preserved speech, and 12 with only some features of RS (generally lacking the characteristic hand dysfunction). All patients with classic RS had *MECP2* mutations; the patient with RS and preserved speech had a previously unreported mutation. None of the patients with atypical RS had mutations. ♦ The accompanying editorial by Singer and Naidu (p. 582) reviews the expanding phenotype and the many variants of RS, considers what is known about its genetic cause, and focuses on the unanswered questions concerning this relatively common disease.

Suspected low-grade glioma: To treat or to wait?

Reijneveld et al. (p. 618) investigated this question by

comparing cognition and quality of life in 24 patients in whom low-grade glioma was suspected with 24 carefully matched patients where diagnosis was established and treatment instituted. Both groups had impaired cognition and reduced quality of life but unoperated patients fared better than those on whom an operation was performed. The data defend a “wait and see” policy for such patients.

CADASIL: Radiologic features useful in diagnosis

Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) is not common but because it causes migraine, recurrent strokes, and dementia, it is in the differential diagnosis of a large number of patients. O’Sullivan et al. (p. 628) compared MRI from 20 documented patients with CADASIL with 20 patients with sporadic leukoaraiosis. Patients with CADASIL (19/20) had temporal pole hyperintensity not seen in those with leukoaraiosis. ♦ Auer et al. (p. 635) found striking MRS abnormalities in both abnormal and normal-appearing white matter in 20 patients with CADASIL. The extent of the MRS abnormality correlated with clinical disability.

Screening for calpain-3 and dysferlin deficiency in limb-girdle (LG) and distal dystrophy

Fanin et al. (p. 660) studied a large referral base of muscle biopsies for calpain-3 and dysferlin. They found that about 25% of biopsies where dystrophin and sarcoglycan were normal were either calpain-3 or

dysferlin deficient. Of LG cases, 28% were calpain-3 deficient.

Hypotensive aphasia?

Hillis et al. (p. 670) obtained sequential imaging data in a man with a left frontotemporal stroke while documenting changes in language. Raising the blood pressure improved and lowering it worsened the language deficit, correlating with changes in MR perfusion.

MRI white matter hyperintensities (WMH) and aldosterone synthase

Verpillat et al. (p. 673) studied 829 elderly subjects for MRI WMH and polymorphisms of the aldosterone synthase gene. (Aldosterone synthase is one of five genes related to hypertension—The others are angiotensin converting enzyme, angiotensinogen, endothelin-1, and NO synthase.) The T allele was associated with a marked increase in the risk for WMH— independent of hypertension.

Intractable temporal lobe epilepsy (TLE) from Rasmussen’s syndrome?

Hennessy et al. (p. 678) describe two patients with intractable temporal lobe seizures in whom temporal lobe surgical specimens documented encephalitis. Seizures persisted after surgery but there was no progression of deficits. Chronic, nonprogressive encephalitis is a cause of TLE.

“Catathrenia”—nocturnal groaning

Vetrungo et al. (p. 681) describe four subjects (age 15 to 25) with expiratory groaning during sleep since age 5 to 16. The groaning was only of concern to observers—not to the subjects themselves.

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