April 13 Highlights

Practice Parameter: Recurrent stroke and PFO and atrial septal aneurysm

The Messé et al. AAN article reviews the risk of stroke with PFO. The presence of PFO alone does not increase risk of recurrent stroke or death, but PFO with atrial septal aneurysm may increase stroke risk. There are insufficient data to define optimal medical treatment or evaluate PFO closure.

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Enzyme replacement therapy for neuropathy in Fabry disease

Hilz et al. assessed effects of enzyme replacement therapy (ERT) on small fiber neuropathy in 22 patients with Fabry disease. After 18 to 23 months of biweekly IV ERT (1.0 mg/kg α -galsidase A), patients had significantly improved vibratory, cold, and heat-pain perception thresholds demonstrating clinical benefits of high-dose ERT. Lack of recovery in some patients suggests a need for early enzyme replacement.

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Triflusal for prevention of stroke: The TAPRISS study

In a study conducted in Buenos Aires, Culebras et al. compared the antiplatelet agent triflusal with aspirin in the secondary prevention of ischemic stroke. They identified no differences in the prevention of vascular complications after TIA or ischemic stroke, but found a lower risk of hemorrhagic complications with triflusal.

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The accompanying editorial by David Anderson and Larry Goldstein notes that as a pilot study, this trial lacked the statistical power required to conclude with confidence that there was no difference between the treatments. However, if the data from this TAPIRSS trial are combined with data from another trial (TACIP), triflusal is associated with fewer major hemorrhages, but there is no difference between aspirin and triflusal for the other indicated endpoints, and no overall difference when all endpoints are considered together. Further, it is not clear whether the difference in bleeding events would be seen if a dose of aspirin lower than 325 mg were used in these studies because aspirin complications, but not efficacy, are dose-related within the range recommended by the FDA.

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Cannabis extract unhelpful for multiple sclerosis tremor

Fox et al. studied 14 patients in a crossover trial of cannabis extract for the relief of tremor in patients with multiple sclerosis. The study showed no objective improvement in tremor.

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Genetics of temporal lobe epilepsies

Recent work suggests that temporal lobe epilepsies (TLE) can be inherited. Berkovic et al. confirm that mutations in LGI1, a gene of unknown function, occur in some families with partial epilepsy with auditory features. Importantly, LGI1 mutations are shown to be specific to this form of familial TLE.

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LGI1 mutations in ADPEAF

Ottman et al. report that mutations in LGI1 are a common cause of autosomal dominant partial epilepsy with auditory features, found in 50% of families with this syndrome. In addition, evidence from two newly identified families suggests mutations may also cause idiopathic generalized epilepsy.

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Primary trochlear headache

Yangüela et al. describe a distinctive headache emerging from the trochlear region, characterized by chronic pain in absence of radiological or autonomic signs. Locally injected corticosteroids relieved the periorbital pain in 48 hours and also improved concurrent headaches, in patients who did not respond to conventional headache therapy.

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Cerebral infarctions and Alzheimer disease pathology

Using clinical and postmortem data from participants of the Religious Orders Study, Schneider et al. found that cerebral infarctions added to the likelihood that Alzheimer disease pathology will be expressed as dementia; however, there was no evidence of a multiplicative effect (interaction) between the two pathologies.

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Is the NART a valid estimator of premorbid intelligence?

McGurn et al. validate the National Adult Reading Test (NART) as an estimate of prior intelligence in people with dementia. After adjusting for childhood IQ a dementia group had similar NART scores but lower MMSE scores than a large non-demented group. The NART-childhood IQ regression slopes were near-identical in the two groups.

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"It is impossible to estimate the extent of dementia in a patient without knowing how they functioned before they were affected by the dementia. By giving the NART, clinicians can determine the premorbid functioning of a patient."

The accompanying editorial by Douglas K. Detterman notes that having a way of estimating premorbid intelligence is useful both clinically and in clinical investigation. He points out that the basis for the observation that pronunciation is maintained while semantic memory is lost presents an interesting disparity deserving explanation.

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Autoimmune limbic encephalitis

Thieben et al. report a series of patients with seizures and cognitive and behavioral changes associated with autoantibodies against voltage-gated potassium channels. Clinical features and antibody studies identify this potentially reversible autoimmune disorder and differentiate this syndrome from other causes of encephalitis.

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The accompanying editorial by David Beeson notes that this potentially treatable disorder has as its major features episodic memory impairment, confusion, and disorientation. Complex partial and grand mal seizures are common and personality changes such as irritability and aggression and hallucinations are sometimes present. Hyponatremia is frequent. Another recent report suggests that this non-paraneoplastic form of immunemediated encephalitis may be relatively common, but may be

misdiagnosed and treated as herpes simplex encephalitis or Korsakoff's syndrome. It is not obvious why members of the VGKC Kv1 ion channel family are targeted. Antibody-mediated disturbance of function in the Kv1 ion channel family is also seen in acquired peripheral nerve hyperexcitability (APNH) syndromes—neuromyotonia and cramp-fasciculation, and Isaac and Morvan syndromes.

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Risk of stroke after discontinuing antiplatelet drugs

Sibon and Orgogozo report a high frequency of stroke (4.5%) between days 6 and 10 following antiplatelet drugs discontinuation.

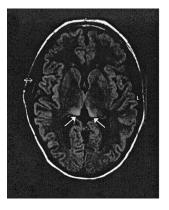
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Induced arterial hypertension in acute ischemic stroke

Marzan et al. reviewed 34 patients with acute ischemic stroke, treated with induced arterial hypertension. Although this approach was safe, clinical improvement was only observed in 27% of patients.

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False-positive pulvinar sign in sporadic Creutzfeldt-Jakob disease



The pulvinar sign on MRI. Bilateral hyperintensities in the pulvinar thalami (arrows) relative to the anterior putamen. Fluidattenuated inversion-recovery (FLAIR) weighted axial image.

The MRI pulvinar sign has been regarded as specific for variant CJD. Petzold et al. report a pulvinar sign occurring in an autopsy-proven case of sporadic CJD.

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