Evidence-based update on transcranial Doppler

The AAN practice parameter by Sloan et al. reviews transcranial Doppler (TCD) and transcranial color-coded sonography (TCCS). TCD is of established value in screening children aged 2 to 16 years with sickle cell disease for stroke risk and for detecting and monitoring of angiographic vasospasm after spontaneous subarachnoid hemorrhage. Contrast-enhanced TCD can provide useful information in detection of right-to-left cardiac/extracardiac shunts. TCD and TCCS may have value for detection of intracranial occlusive disease, vasomotor reactivity, detection of cerebral circulatory arrest/brain death, monitoring carotid endarterectomy, and monitoring thrombolysis.

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Risk factors for postherpetic neuralgia

In 965 patients with herpes zoster, Jung et al. found that in older women, the presence of a prodrome, greater rash severity, and acute pain each made an independent contribution to the development of postherpetic neuralgia (PHN).

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Unraveling causes for hypomyelination

In two girls with severe developmental delay, seizures, nystagmus, and CNS hypomyelination, Wolf et al. found markedly elevated levels of *N*-acetylaspartylglutamate in CSF. The underlying genetic defect could not be elucidated.

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Genotype-phenotype correlation in eIF2B-related leukodystrophies

Fogli et al. evaluated MRI-based criteria to help select patients with an undetermined leukodystrophy who tested positive for EIF2B genes mutation. Disease severity from fatal infantile to adult asymptomatic forms correlated with age at onset. Two mutations were significantly associated with milder forms.

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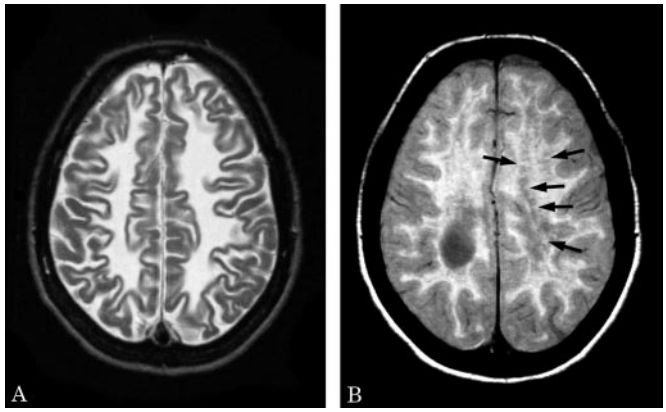
Vanishing white matter disease in adults



T2-weighted (A) and proton density (B) MR images in a 35-year-old patient. The cerebral white matter is diffusely abnormal (A). There is white matter rarefaction and cystic degeneration (B).

Vanishing white matter (VWM) is a well-known leukoencephalopathy in children. A DNA-based diagnosis is now possible. Van der Knaap et al. describe five adult patients homozygous for a particular VWM mutation. VWM should, therefore, also be considered in the differential diagnosis of adult diffuse leukoencephalopathies.

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Ohtake et al. report a 52-year-old woman who developed dementia in her mid-40s presenting with arrests for shoplifting. She had a spastic gait and pathologically brisk reflexes. T2-weighted MRI showed diffuse hypointense lesions in cerebral white matter. Genetic studies showed a homozygous mutation in EIF2B5.

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The editorial accompanying these four articles by Edward Kaye and Hugo Moser notes that the term "leukodystrophy" is not appropriate for many such disorders: they are better termed "leukoencephalopathy" since failure in myelination or hypomyelination rather than a loss of previously acquired myelin is responsible for the disease. The first and most common is vanishing white matter (VWM) or childhood ataxia with cerebral hypomyelination (CACH). VME/CACH is caused by mutations in the one of the five subunits (alpha, beta, gamma, delta, and epsilon) of eukaryotic initiation factor 2B (eIF2B), which appears to be important in the regulation of translation initiation. Some genetic defects, such as the R113H in the eIF2B epsilon region and the E213G substitution in the EIF2B beta region, are associated with a mild clinical course. As the report of Wolf et al. demonstrates, there are still more "new" leukoencephalopathies that remain to be defined.

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Opsoclonus-myoclonus syndrome: CSF lymphocyte markers

Pranzatelli et al. immunophenotyped CSF lymphocytes in 36 children with opsoclonus-myoclonus syndrome. B- and T-cell markers correlated strongly with motor severity, suggesting they may be useful in evaluation of relapses and treatment failures.

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"What is missing here unfortunately is the presence of an antibody that would allow the authors to demonstrate the antigen (if any) responsible for the immune reaction that causes POM."

The accompanying editorial by Jerome Posner notes that despite the logical conclusion that paraneoplastic CNS diseases such as paraneoplastic opsoclonus/myoclonus (POM) associated with neuroblastoma in children is immune-medicated, there has been no antibody consistently identified. It is unclear whether the pathogenesis is mediated by B-cells, T-cells, or both. The high percentage of B-cells suggests that in POM, despite the inability to identify an antibody, one is being produced within the CNS.

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Assessment of cognitive function in collegiate boxers

In a prospective study utilizing computerized neuropsychological testing, Moriarity et al. measured changes in cognitive function during a 7-day collegiate boxing tournament. Boxers participating in multiple bouts displayed no change in cognition immediately after boxing. Mild dysfunction was observed in seven boxers whose match was stopped by the referee.

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The accompanying editorial by Deborah Warden notes that the Moriarity et al. article provides encouraging data that participation in amateur boxing under tournament conditions in young healthy students does not have negative consequences. The stringent and conservative management of the bouts in this study may have contributed to the generally good outcomes, and reinforces the wisdom of more conservative rules in recent years for amateur collegiate boxers. This study says nothing about the risks of longer term participation in boxing. Prospective long-term studies are required to answer questions of cumulative exposure, individual variation, and subsequent outcome.

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Rofecoxib for the acute treatment of migraine

Silberstein et al. demonstrated in a randomized, double-blind, placebo-controlled, multicenter trial in 557 migraine patients that 25 and 50 mg doses of the selective cyclooxygenase-2 inhibitor rofecoxib were effective for the acute treatment of migraine attacks.

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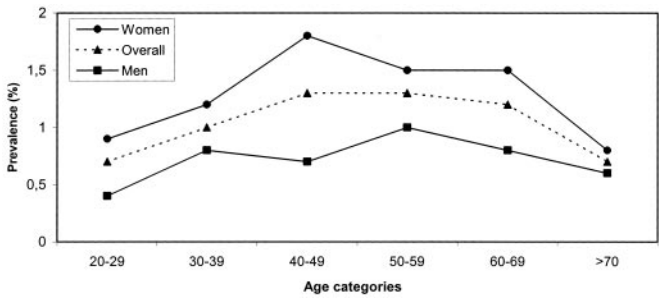
Medication overuse headache: The Head-HUNT study



Prevalence of chronic headache associated with daily/almost daily use of analgesics ≥1 month during the last 12 months.

Zwart et al. showed in a large cross-sectional population-based study in Norway (1995–1997) that the prevalence of chronic headache associated with analgesic overuse was 1% (1.3% for women and 0.7% for men). The association was stronger for chronic migraine than other chronic pain disorders.

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Dietary fat intake and 6-year cognitive change

Morris et al. related fat consumption to 6-year cognitive change in a study of 2,560 older persons with no history of cardiovascular disease. Diets high in saturated or transunsaturated fat or low in non-hydrogenated unsaturated fats were associated with faster rate of cognitive decline.

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Is complex regional pain syndrome type 1 (CRPS1) mediated by the brain?

G. Lorimer Moseley studied a 34-year-old woman with CRPS1 17 months after a wrist fracture. He documented an increase in pain and swelling of the affected hand during imagined movements not accompanied by muscle activity or a sympathetic response.

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May 11 Highlights

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