VASCULAR DISORDERS

ETIOLOGY OF BRAIN ATTACKS IN CHILDREN

Investigators at the Royal Children's Hospital Melbourne, Australia, studied the presenting features, scope, and prevalence of conditions causing brain attack symptoms in children aged 12 month to 18 years presenting to a tertiary pediatric ED. Brain attack is defined as apparently abrupt-onset focal brain dysfunction. Exclusion criteria include epilepsy, hydrocephalus, head trauma, and isolated headache. Of 287 children (46% male) with 301 presentations over 17 months, 35% arrived by ambulance. Median symptom duration before arrival was 6 hours (range 2-28 hrs.); median time from triage to medical assessment was 22 min (range, 6-55 min). Common symptoms included headache, vomiting, focal weakness, numbness, visual disturbance, seizures, and altered consciousness. Common signs included focal weakness, numbness, ataxia, or speech disturbance. CT imaging in 30% was abnormal in 27%, and MRI in 31% was abnormal in 62%. Diagnoses included migraine (28%), seizures (15%), Bell palsy (10%), stroke (7%), and conversion disorders (6%). Relative proportions of conditions in adults (obtained by meta-analysis) and children differed significantly for stroke, migraine, seizures, and conversion disorders. Brain attack etiologies in children differ from those in adults; stroke is a relatively infrequent diagnosis (7%) in children and accounts for 73% of cases in adults. (Mackay MT, Chua ZK, Lee M, et al. Stroke and nonstroke brain attacks in children. **Neurology** 2014 Apr 22;82(16):1434-40).

COMMENTARY. Migraine is the most common stroke mimic in children, accounting for more than one-quarter of cases, whereas in adults it accounts for less than 3% of cases.

Transient ischemic attacks requiring hospitalization in children. Using a Kids' Inpatient Database, TIA was the primary diagnosis for 531 children, and secondary diagnoses and risk factors for TIA included sickle cell disease (20%), congenital heart disease (11%), migraine (12%), moyamoya disease (10%), and stroke (4%). Mean length of hospital stay decreased from 3.0 days in 2003 to 2.3 days in 2009. During the same period, pediatric admissions for ischemic stroke (n=2590) were ~5-fold more common than for TIA; 4.8 children with stroke were admitted for every child with TIA [1].

References.

1. Adil MM, et al. Stroke. 2014 Mar;45(3):887-8.

MELAS, STROKE-LIKE EPISODES AND KETOGENIC DIET

Investigators at University of Toronto and McMaster University, Canada, report a 22-year-old woman with multiple episodes of status epilepticus and migratory cortical stroke-like lesions. Ketogenic diet and magnesium resulted in seizure freedom and decrease in frequency of stroke-like episodes following improvement of mitochondrial dysfunction. Initial mitochondrial genetic testing was negative. Diagnosis was established by muscle biopsy for mitochondrial genome sequencing, demonstrating a mitochondrial DNA disease-causing mutation. (Steriade C, Andrade DM, Faghfoury H, Tarnopolsky

MA, Tai P. Mitochondrial encephalopathy with lactic acidosis and stroke-like episodes (MELAS) may respond to adjunctive ketogenic diet. **Pediatr Neurol** 2014 May;50(5): 498-502).

COMMENTARY. A ketogenic diet should be considered for treatment of intractable seizures and stroke-like episodes related to mitochondrial respiratory chain complex (MRC) defects. In a Korean study of 14 patients with MRC defects and various seizure types (5 with infantile spasms, 4 with Lennox-Gastaut syndrome, 1 with Landau-Kleffner syndrome), 50 – 90% seizure control was obtained with the ketogenic diet [1]. A subsequent study in Korea involving 24 cases of MRC defect with seizures, the ketogenic diet controlled seizures in 75% patients [2].

References.
1. Kang HC, et al. Epilepsia. 2007 Jan;48(1):82-8.
2. Lee YM, et al. Epilepsia. 2008 Apr;49(4):685-90.

HEADACHE DISORDERS

HEADACHE AND MIGRAINE WITH SICKLE CELL DISEASE

Investigators from University of Texas Southwestern Medical Center, Dallas, TX, and other centers in the US and London, UK, studied risk factors for headache and migraine in 872 children, age 5 to 15 years (mean age, 9.1 years), with sickle cell disease (hemoglobinSS or hemoglobinSb-thalassemia) and no history of overt stroke or seizures. Recurrent headaches were reported in 317 (36.4%) and migraines in 132 (15.1%). Both were associated with lower hemoglobin and higher rate of hospitalization for pain events requiring hospitalization for treatment with opioids in the previous 3 years. Only six of 317 (1.9%) children reporting recurrent headaches were receiving medication for headache prophylaxis. The prevalence of silent cerebral infarct, diagnosed by MRI and neurological examination, was similar in patients with recurrent headaches and in those without headaches (32.8% and 29%, respectively; P=0.241). Older age, lower Hgb concentration, and higher pain event rate were associated with recurrent headaches and migraines. (Dowling MM, Noetzel MJ, Rodeghier MJ, et al. Headache and migraine in children with sickle cell disease are associated with lower hemoglobin and higher pain event rates but not silent cerebral infarction. **J Pediatr** 2014 May;164(5):1175-1180.e1).

COMMENTARY. Isolated recurrent headaches or migraine in neurologically normal children with sickle cell disease (SCD) might not necessitate additional evaluation with imaging studies, but new severe headaches presenting acutely warrant further investigation, especially when associated with acute CNS events. In a study of children with SCD who presented acutely with headache, headache was the chief complaint in 3.8% of acute care visits, and acute CNS events occurred in 16.9%. Factors associated with acute CNS events included older age, history of stroke, TIA, or seizure, focal neurological findings, and elevated platelets [1].

References.
1. Hines PC, et al. J Pediatr. 2011 Sep;159(3):472-8.