VASCULAR DISORDERS

NEONATAL CEREBRAL SINOVENOUS THROMBOSIS

The presentation, treatment, and outcome of neonatal cerebral sinovenous thrombosis (SVT) were studied in 42 children, using neurology clinic records (1986-2005) at Indiana University School of Medicine. Gestational/delivery complications occurred in 82%, including preeclampsia/hypertension in 26%, gestational diabetes (26%), and meconium aspiration in 24%. Comorbid risk factors in 62% included dehydration. sepsis, meningitis, and cardiac malformations. Seizures in 57% were the most common presenting symptom. A single sinus was involved in 50%, most commonly the sagittal sinus. Infarcts occurred in 60%, and 64% had received prothrombotic evaluations, testing negative for protein C, protein S, or antithrombin III deficiencies. Three (13%) of 24 tested were heterozygous for factor V Leiden, and 8 (42%) of 19 were positive for other prothrombin gene mutations. Three (7%) were treated with heparin sodium, and all others received only supportive care. One died, and of 41 who survived, 23 (79%) had impairments that included cognitive disorders, cerebral palsy, and epilepsy. (Fitzgerald KC, Williams L S, Garg BP, et al. Cerebral sinovenous thrombosis in the neonate. **Arch Neurol** March 2006;63:405-409).

COMMENT. SVT is a risk factor in neonates following a complicated pregnancy or birth or with acute systemic debilitating illness. Presenting symptoms include seizures, focal or generalized, focal motor deficits, and increased intracranial pressure. Diagnosis is made with MRI and venogram. The outcome is usually poor, with long-term sequelae. Fortunately, due to improved fluid therapy and antibiotics, cerebral venous thrombosis is now uncommon. In earlier times, thrombosis was usually due to sepsis of the mastoid, lip or orbit, and often as a complication of cyanotic congenital heart disease (Byers RK, Hass GM. **Am J Dis Child** 1933;45:1161).

PROGNOSIS OF CEREBRAL ARTERIOPATHY IN STROKE

The evolution of cerebral arteriopathy in 50 children with first arterial ischemic stroke (AIS) was evaluated at Great Ormond Street Hospital for Children, and Neuroscience Unit, Institute of Child Health, London. The median age was 49 months (range 4 mo to 14 yr). Risk factors for AIS included varicella-zoster within 12 months (22 patients), congenital heart disease (4), sickle cell disease 1, and gene mutations in 6. Arteriopathy graded for severity on serial MR angiograms affected 72 arteries in 43 (86%) patients; 5 had clinical recurrence, 12 were progressive, 24 improved, and 7 were stable. Magnetic resonance angiograms were normal in 7. Arteriopathy was transient in 24, chronic in 11, and diagnoses included arterial dissection in 3, moyamoya (3), and vasculitis in 1. In patients with progressive arteriopathy, the hazard of stroke recurrence was increased threefold. After adjusting for age and AIS risk factors, the hazard ratio was 3.1; p=0.27). (Danchaivijitr N, Cox TC, Saunders DE, Ganesan V. Evolution of cerebral arteriopathies in childhood arterial ischemic stroke. **Ann Neurol** April 2006;59:620-626). (Respond: Dr Ganesan, Department

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COMMENT. The authors have employed serial magnetic resonance angiography to distinguish a relatively benign and self-limiting vasculopathy from progressive types such as moyamoya. Risk factors that determine progression of arteriopathies include a history of varicella zoster, congenital heart anomalies, immunodeficiences, and hemolytic anemias, especially sickle cell disease. In addition to varicella, enteroviruses and Borrelia are reported as triggers for arterial ischemic stroke (Sebire G et al. **Ann Neurol** 1999;45:679-680; Kirkham F. **Ann Neurol** 2006;59:580-582).

EEG hyperventilation in sickle cell disease is a preventable risk factor for arterial stroke, not mentioned by the above authors. Three reports and four childhood cases of sickle cell disease (SCD) with stroke precipitated by routine hyperventilation during EEG recordings are cited in the literature. The earliest report (Protass LM. **Ann Intern Med** 1973;79:451) concerned an 11-year-old girl with SCD who developed a left hemiparesis immediately following hyperventilation during an EEG. Recovery was incomplete in all cases. Additional cases have involved hyperventilation secondary to obstructive sleep apnea, or ingestion of toxic doses of aspirin. Hyperventilation-induced hypocapnea leads to arterial constriction, decreased cerebral blood flow, and in patients with SCD, intravascular sickling precipitated by hypoxia and cerebral ischemia or infarction. Seizures occurring in 12-14% of patients with SCD are a precursor to stroke in 10-33% and may be associated with vasculopathy, focal hypoperfusion, and silent infarction. (Prengler M et al et al. **Ann Neurol** 58:290-302). The lack of appreciation of potential dangers of hyperventilation in SCD in some reports prompted a cautionary comment and review of the literature (Millichap JG, **Ann Neurol** 2005;58:972; Millichap JG. **Clin EEG and Neuroscience**, in press). The avoidance of hyperventilation in patients with SCD is recommended.

SEIZURE DISORDERS

ETIOLOGY AND OUTCOME OF NEONATAL SEIZURES

The prognostic value of seizure etiology, neurologic examination, EEG, and neuroimaging in the neurodevelopmental outcome of 89 term infants with neonatal seizures was determined at the Children's Hospital and Harvard Medical School, Boston, MA. The seizure etiologies were global cerebral hypoxia-ischemia (HI) in 40%, focal HI in 18%, intracranial hemorrhage (17%), cerebral dysgenesis (5%), transient hypoglycemia or hypocalcemia (3%), meningitis or encephalitis (3%), and pyridoxine dependency (1%). Neurologic outcome at 1 year was favorable in 72%, and poor in 28%. Neurologic examination was abnormal in 54% (mild in 26% and severe in 22%) with motor impairment in 53%, mental impairment in 48%, and seizures after NICU discharge in 21%. Long-term outcome was poor in 28% of survivors; neonatal mortality was 7%. Risk factors for a poor outcome were seizures associated with cerebral dysgenesis or global HI, an abnormal EEG background activity, and multifocal cortical or deep gray matter neuroimaging lesions in the neonate. A favorable outcome at 12-18 month follow-up was predicted by a normal neurologic examination in the neonatal and early infancy period, and a normal/mildly abnormal neonatal EEG. An abnormal neurologic exam in the neonate was an unreliable