Neuronal migration disorder as a cause of focal motor status epilepticus or epilepsy partialis continua in a child may not be excluded by the MRI and may require surgical intervention for diagnosis and treatment.

VASCULAR DISORDERS

MOYAMOYA DISEASE AND HEMIPLEGIC MIGRAINE

A 6-year-old girl presenting with a 6-month history of classical familial migraine, complicated by transitory hemiparesis and moyamoya disease diagnosed by MRI, is reported from the Kaiser Permanente Medical Center, Hayward, CA. Headaches occurred each week and were associated with nausea, vomiting, and photophobia. She awoke on 3 occasions with right-sided weakness, numbness, and garbled speech. All symptoms resolved after two hours of sleep. Neurologic exam was normal between attacks. MRI showed bilateral occlusion of the carotid arteries and increased vascularity of collateral vessels in the basal ganglia, consistent with moyamoya. (Bernstein AL. Hemiplegic migraine and moyamoya disease. AJDC July 1993; 147: 718-719).

COMMENT. Children with a diagnosis of hemiplegic migraine may have a vascular malformation or moyamoya disease as the underlying pathology. The clinical course depends on the rapidity and extent of vascular occlusion and the ability to develop a collateral circulation. Early diagnosis and surgical revascularization have been advocated because of the risk of permanent neurologic deficits. (See Ped Neur Briefs July 1987, and Progress in Pediatric Neurology. Chicago, PNB Publ, 1991, pp389-391).

ENDOCRINE DISORDERS

ADRENAL, ALACRIMAL, ACHALASIA SYNDROME

The results of a British and European Paediatric Endocrinology Society, multicenter collaborative, questionnnaire study of the neurological complications of familial glucocorticoid deficiency syndrome are reported from the Hospital for Sick Children, Great Ormond Street, London. Of 20 patients identified, ages 2 to 29 years, all had impaired cortisol secretion, 19 absent tear secretion, 15 achalasia of the cardia, and 17 had neurologic abnormalities including hyperreflexia, hypertonia, Babinski signs, muscle
weakness and/or wasting, ataxia, dysarthria, and sensory impairment. Impaired autonomic function in 11 patients was manifested by postural hypotension and anisocoria. Eleven were intellectually impaired. Ten had hyperkeratosis and fissures of their palms. Siblings were affected in 3 families, and 7 siblings had died in childhood. (Grant DB et al. Neurological and adrenal dysfunction in the adrenal insufficiency/alacrima/achalasia (3A) syndrome. Arch Dis Child 1993; 68: 779-782). (Respond: Dr DB Grant, Hospital for Sick Children, Great Ormond Street, London WC1N 3JH).

COMMENT. The most frequent presenting symptom was hypoglycemia, often associated with addisonian skin pigmentation. In 4 cases the diagnosis of achalasia preceded the recognition of cortisol deficiency. The authors recommend a test of cortisol secretion in children with achalasia, if associated with alacrima or with any of the above neurologic abnormalities.

INFECTION DISORDERS

HEMOPHILUS INFLUENZAE VACCINE AND GUILLAIN-BARRE

A four-year-old girl with Guillain-Barre syndrome that developed 10 days following H influenzae type b conjugate vaccine immunization is reported from Geneva, Switzerland. CSF contained 1 mononuclear cell/cmm and 0.89 g/l protein. Nerve conduction velocities were decreased in upper and lower extremities. Measurements of immunoglobulins against the polysaccharide (PRP) component of the vaccine showed a marked elevation (100 mcg/ml) of the anti-PRP IgM antibody level. Following treatment with two 5-day courses of intravenous immunoglobulins (0.4 g/kg/day) 2 weeks apart, cranial nerve function and muscle strength improved. Apart from absent deep tendon reflexes, the neurological exam was normal at one month follow-up. (Gervaix A et al. Guillain-Barre syndrome following immunization with Hemophilus influenzae type b conjugate vaccine. Eur J Pediatr July 1993; 152: 613-614). (Respond: Dr A Gervaix, Children's Hospital, University Hospital of Geneva, 30 bvld de la cluse, CH-1211 Geneve 4, Switzerland).

COMMENT. A total of five cases of Guillain-Barre syndrome (GBS) following vaccination with the H influenzae type b diphtheria toxoid-conjugate vaccine have been reported. The authors speculate that an excessive anti-PRP IgM antibody response to the vaccine might explain this rare complication.

Treatment of acute GBS is controversial and not without risk, especially in patients with vascular disease. Two adults died from vascular thrombotic complications following gamma globulin therapy for GBS. (McFarland HR. Arch Neurol July 1993; 50: 687).