Dr Mohamad Mikati, Department of Neurology, Division of Neurophysiology, Hunnewell 2, Children's Hospital, Boston, MA 02115).

COMMENT. This is the first description of truly familial alternating hemiplegia, a syndrome of unknown etiology. The authors favor neurovascular or metabolic, rather than epileptic mechanisms.

Sakuragawa N reports 23 cases in an excellent review article from the National Institute of Neuroscience, Ogawa Higashi-cho, Kodaira-shi, Tokyo 187, Japan (Brain Dev Sept 1992; 14: 283-8). The classification of alternating hemiplegia as a migraine variant is discussed.

HEMIPLEGIA WITH SUPERIOR SAGITTAL SINUS THROMBOSIS

A 2-year-old boy who developed a thrombosis of the superior sagittal sinus due to inherited protein S deficiency is reported from the Hospital de Cruces, Vizcaya, Spain. While recovering from pneumonia, the patient had symptoms of an acute disseminated intravascular coagulation disorder associated with sinus thrombosis, left hemiparesis, focal seizures, and thrombosis of the left femoral vein. The paresis resolved within two months. (Prats JM et al. Superior sagittal sinus thrombosis in a child with protein S deficiency. Neurology Dec 1992; 42: 2303-5). (Reprints: Dr Jose Maria Prats, Neuropediatric Unit, Hospital de Cruces, 48903 Baracaldo, Vizcaya, Spain).

COMMENT. Homozygous protein S deficiency, an autosomal dominant trait with partial penetrance, usually presents with purpura fulminans in the newborn period. Heterozygous cases are responsible for recurrent peripheral thromboembolisms mainly in adults. Cerebral venous thrombosis is a rare complication of protein S deficiency, but should be considered in the etiology of acute hemiparesis in children.

RASMUSSEN'S SYNDROME AND UVEITIS

Two children with Rasmussen's syndrome complicated by asymptomatic uveitis are reported from the Royal Children's Hospital, Melbourne, Australia, and Montreal Neurological Institute, Montreal, Canada. A 10-year-old girl developed somatosensorimotor seizures affected her right leg and left sided acute uveitis. Epilepsia partialis continua, with onset 9 months later, was associated with right hemiparesis and cortical sensory loss, and dysphasia. EEG showed epileptic discharges in the left central region. CT revealed atrophy of the left hemisphere. Left frontal corticectomy failed to control seizures. The cortex showed chronic encephalitis. The second patient, a 5-year-old girl, had complex partial status epilepticus which responded to IV gammaglobulin, left facial weakness, and right sided uveitis. Right anterior temporal lobectomy revealed leptomeningeal inflammation. Seizures became intractable, and a

dense left hemiparesis developed. She died 2 years 7 months after the onset of seizures. (Harvey AS, Andermann F et al. Chronic encephalitis (Rasmussen's syndrome) and ipsilateral uveitis. Ann Neurol Dec 1992; 32: 826-829). (Correspondence: Dr Harvey, Dept of Neurology, Royal Children's Hospital, Flemington Rd, Parkville, Victoria 3052, Australia).

COMMENT. A viral cause seemed likely but was not confirmed by serology or tissue culture. Slit-lamp examination of the eye should be included in the evaluation of children with Rasmussen's syndrome.

ENCEPHALOPATHIES

ACUTE ENCEPHALOPATHY OF OBSCURE ORIGIN

Six previously healthy children who developed an acute encephalopathy several days after a prodromal illness are reported from the Hopital Bicetre, and Hopital Necker-Enfants Malades, Paris, France. Prodromal illnesses consisted of upper respiratory infection with fever, and headache and vomiting. Coma was the initial symptom in 4 patients. Abnormal movements included gesticulation, chewing, swallowing, orofacial dyskinesia, limb dystonia, and choreoathetosis. Rigidity was constant in 5 patients and intermittent in one. Seizures occurred in 2 patients. Recovery extended for several weeks and was characterized by a rapid return of motor function and persistent behavioral and cognitive disturbances. Four patients recovered fully, and two had mild sequelae. (Sebire G et al. Coma associated with intense bursts of abnormal movements and long-lasting cognitive disturbances: An acute encephalopathy of obscure origin. J Pediatr Dec 1992; 121: 845-51). (Reprints: G Sebire MD, Service de Neurologie, Departement de Pediatre, Hopital Bicetre, 78 rue du general Leclerc, 94275, Le Kremlin Bicetre Cedex, France).

COMMENT. These cases with a favorable outcome were thought to represent a different syndrome from that described by Lyon, Dodge, and Adams, whose 16 patients died from an acute encephalopathy of obscure origin. Attempts at viral isolation and antibody detection were negative.

DEGENERATIVE DISEASES

HEPATOCEREBRAL DEGENERATION OR VALPROATE TOXICITY

Six children with refractory seizures and focal neuronal damage who died of liver failure are reported from the Washington University School of Medicine, St Louis, MO. Four were treated with valproic acid (VPA) and developed liver failure within 30 - 68 days. Two of these children each had one sibling who was not exposed to VPA and developed the same clinical picture, but liver failure was delayed. Siblings receiving VPA survived only 3 and 5