COMMENT. The VER is proposed as a useful test in the diagnosis of migraine in children. The test may be especially valuable in the differentiation and diagnosis of cases of periodic or cyclical vomiting when a migrainous etiology is unclear. (Millichap JG. Arch Fr Pediatr 1987; 44:231; Pediatrics 1955; 15:705).

SEIZURE DISORDERS

CORTICAL DYSGENESIS AND INFANTILE SPASMS: PET STUDIES

The identification of focal cortical dysgenesis by positron emission tomography (PET) in 5 of 13 children with cryptogenic infantile spasms is reported from the Departments of Neurology and Pediatrics and Division of Neurosurgery, UCLA School of Medicine, Los Angeles, CA. There was unilateral hypometabolism of cerebral glucose involving the parieto-occipito-temporal region. Neuropathological examination of resected tissue in four infants showed microscopic cortical dysplasia. The CT was normal in all infants and the MRI showed a subtle abnormality only in one. The EEG showed hypsarrhythmia and at times, a localized abnormality corresponding to areas of PET hypometabolism. PET may identify unsuspected focal cortical dysplasia in infants with cryptogenic spasms and resective surgery offers improved prognosis. (Chugani HT, Shields WD et al. Infantile spasms: I. PET identifies focal cortical dysgenesis in cryptogenic cases for surgical treatment. Ann Neurol April 1990; 27:406-413).

COMMENT. Early studies showed that infantile spasms were cryptogenic in about 40% of patients (Millichap et al. Epilepsia 1962; 3:188) whereas more recent studies have demonstrated that this figure has diminished to 9-14%. The PET studies have uncovered further symptomatic cases previously not identified by CT and MRI. The same authors report lenticular nuclei hypermetabolism in 12 of 25 infants with spasms of cryptogenic or symptomatic types. They suggest that the lenticular nuclei may contribute to the pathogenesis of infantile spasms. (Chugani HT et al. Neurology April 1990; 40:suppl 1:407).

TUBEROUS SCLEROSIS AND INFANTILE SPASMS

The short- and long-term outcome of 24 children with infantile spasms and tuberous sclerosis was studied at the Department of Pediatrics, University of Turku, Finland and at the Children's Hospital, University of Helsinki. They comprised 10% of all cases of infantile spasms treated in the two hospitals between 1964 and 1985. CT showed brain calcifications in 20 patients examined at an early age. Three of 14 patients tested by renal ultrasound had large polycystic kidneys and severe arterial hypertension. Early diagnosis and the avoidance of ACTH therapy could have prevented hypertensive crises secondary to ACTH injections. One child developed severe myocardial hypertrophy during ACTH therapy and two had rhabdomyomas demonstrated by cardiac ultrasound and angiography at age one week. Short-term

outcome was good but relapses were frequent and the long term outcome was disappointing. All were mentally retarded, only 4% were seizure free at follow-up and 42% had behavioral problems. When examined at 2½ to 19 years of age, 58% had partial or focal, often secondary generalized seizures, and 37% had myoclonic astatic or Lennox-Gastaut syndrome. The dose of ACTH was 20-40 IU daily for six weeks in eight children and 80-140 IU daily in 14 children. Arterial hypertension occurred in ten, two developed cardiac failure, three had fluid retention in the cysts of polycystic kidneys and developed hypertensive crises during therapy. Infections (otitis, gastroenteritis, pneumonia) occurred in four. (Riikonen R, Simell O. Tuberous sclerosis and infantile spasms. Dev Med Child Neurol March 1990; 32:203-209).

COMMENT. The demonstration of cerebral calcifications by CT in all patients with tuberous sclerosis at an early age is of interest. The necessity for abdominal ultrasound in diagnosis and before ACTH therapy is indicated by the study. The dosage of ACTH used was exceptionally large and would have accounted for the unusually high incidence of side effects and frequency of arterial hypertension. Many authorities are content with much smaller doses, 10 and at the most 20 IU of ACTH daily given for shorter periods (three weeks) and repeated at intervals when necessary. Early treatment with ACTH is important in terms of the response of infantile spasms to therapy and possibly in relation to subsequent development (Gordon N. Dev Med Child Neurol April 1990, 32:363). In the present study of patients with tuberous sclerosis and infantile spasms, early treatment with large doses of ACTH provided an initial good response but the long-term outcome was poor despite prolonged administration.

CARBAMAZEPINE TOXICITY

Three children who developed acute liver failure while taking carbmazepine are reported from the Department of Child Health, King's College Hospital, Denmark Hill, London, England. A girl aged 11 developed a severe maculopapular rash, intermittent fever, arthralgia, anemia, and vomiting four weeks after starting carbamazepine. The blood concentration was 32 mcmol/l (therapeutic range 16-50). She developed jaundice two weeks later and on admission six days later she had a generalized exfoliative rash, periorbital edema, generalized lymphadenopathy, and hepatomegaly. Her platelets and differential white count were normal. Concentrations of IgG, IgM, and IgE were increased. Liver biopsy showed acute hepatitis. Steroid treatment with prednisolone (0.7 mg/kg/24 hours) caused a dramatic symptomatic and biochemical improvement. She was discharged one week later after complete recovery and prednisolone was stopped after 18 days. The second child, aged 7, again presented with fever, generalized maculopapular rash, arthralgia, and lymphadenopathy four weeks after starting carbamazepine. She developed jaundice, ascites, and generalized edema 17 days later. Total bilirubin concentration was 236 mcmol/l. Hepatic encephalopathy developed four days after admission and the child died of infectious complications three months after a liver transplantation. The third patient, a 3 year old child,