

patterns in children presenting to a pediatric headache clinic. Headache Oct 1993;33:497-500). (Respond: Jack Gladstein MD, University of Maryland School of Medicine, Department of Pediatrics, Rm N5W70, 22 South Greene St, Baltimore, MD 21201).

COMMENT. These findings suggest that the International Headache Society criteria (1988) should be modified to increase their sensitivity to children and adolescents, and criteria at present proposed for the pediatric headache population should be reexamined.

In 100 consecutive children with chronic, recurrent headaches as a presenting complaint seen in a private office during a 2 year period, 42 were diagnosed as migraine, of which 27 (64%) were classified as classic migraine. These were hemicranial, associated with visual phenomena and/or nausea and vomiting, and the family history was positive in the majority. The 15 patients with common migraine had generalized headaches complicated by visual phenomena, nausea, or vomiting. Phenytoin controlled the migraine in 77%; the beneficial response was unrelated to EEG abnormalities. In this study, a unilateral location and family history were considered important criteria in the diagnosis of classic migraine. The character of the pain, throbbing or pulsating, emphasized in some classifications, was considered unreliable in children and was not included in diagnosis. (Millichap JG. Recurrent headaches in 100 children. Electroencephalographic abnormalities and response to phenytoin (Dilantin). Child's Brain 1978;4:95-104).

An uncommon EEG pattern, characterized by diffuse continuous beta activity, is described in an 8-year-old boy with recurrent migraine aura without headache from Ferrara University, Italy. (Soriani S et al. Headache Oct 1993;33:509-511). Epileptiform EEG discharges are not unusual in pediatric migraine patients.

METABOLIC and TOXIC DISORDERS

MENKES DISEASE: COPPER-HISTIDINE THERAPY

The response to subcutaneous copper-histidine treatment (50 - 150 mcg Cu/kgm/daily) in seven children with Menkes disease (Kinky-Hair disease) is reported from the Hospital for Sick Children and University of Toronto, Canada. Two patients, ages 16 and 6 years, whose therapy began within 1 month of birth, did well neurologically but have skeletal deformities and muscle weakness. Both have normal IQs and no seizures. The other 5 patients whose treatment was initiated at 2 to 7 months of age failed to thrive and had progressive neurological deterioration. Treatment with copper-histidine is recommended and should be started before 1 month of age. (Sarkar B, Lingertat-Walsh K, Clarke JTR. Copper-histidine therapy for Menkes disease. J

Pediatr Nov 1993;123:828-830). (Reprints: B Sarkar PhD, Head, Department of Biochemistry Research, Hospital for Sick Children, Toronto, Ontario M5G 1X8, Canada).

COMMENT. In a 13-week-old boy with Menkes disease treated with copper-histidine by daily intramuscular injections, Cu and ceruloplasmin in serum and Cu in CSF became normal after 6 weeks, epileptic discharges and infantile spasms were reduced, muscle tone was improved, motor activities increased, and developmental regression stopped. A marked improvement in osseous changes at 6 months was also noted. (Kreuder J et al. Clinical and biochemical consequences of copper-histidine therapy in Menkes disease. Eur J Pediatr Oct 1993; 152:828-832). This report from Children's Hospital, Justus Liebig University, Giessen, Germany, is the first to demonstrate normalization of dopamine and dopamine/norepinephrine ratio in CSF after copper-histidine therapy.

The recent reports from three independent laboratories of the isolation of a Menkes' gene candidate have narrowed the search for the defective or missing factor in this sex-linked, recessive neurodegenerative disorder. A copper-transporting ATPase may be the important factor in normal copper metabolism. (Harris ED. Menkes' disease: Perspective and update on a fatal copper disorder. Nutrition Reviews Aug 1993;51:235-245).

FETAL VALPROATE SYNDROME

The high frequency of minor abnormalities and major malformations, the withdrawal manifestations, and hypoglycemia in infants born to mothers who received sodium valproate during pregnancy are reported from the Department of Paediatrics, Aalborg Hospital, Denmark. Of 17 mothers, 11 had valproate monotherapy, and 6 had valproate combined with another anticonvulsant, usually carbamazepine. Minor abnormalities (epicanthic folds, hypertelorism, low set ears, etc) affected 9 (53%) infants, of whom 5 also had major malformations, predominantly congenital heart disease, present in 4. The frequency of abnormalities was related to the valproate dosage in the first trimester. Of 6 infants born to mothers receiving >2.5 g daily, 5 had minor abnormalities, and 3 had major malformations. Withdrawal symptoms in 9 (53%) infants included irritability, jitteriness, seizures, and feeding problems; hypoglycemia occurred in 4. At 2.0 - 3.5 year follow up, 3 showed psychomotor retardation. (Thisted E, Ebbesen F. Malformations, withdrawal manifestations, and hypoglycemia after exposure to valproate in utero. Arch Dis Child Sept 1993;69:288-291). (Respond: Dr Ebbe Thisted, Department of Paediatrics, University Hospital of Hvidovre, Kettegards Alle 30, DK-2650 Denmark).