The outcome was poor in the majority, with motor sequelae and/or cognitive impairment. (Roig M et al. Bilateral striatal lesions in childhood. Pediatr Neurol Sept/Oct 1993;9:349-358). (Respond: Dr Roig, Child Neurology Unit, Hospital Materno-Infantil Vall D'Hebron, Paseo del Vall D'Hebron s/n, Barcelona, Spain).

COMMENT. Based on a review of the literature, the authors tabulate the causes of infantile bilateral striatal syndrome (IBSS) under acute and subacute/chronic categories. Acute causes include SNE, HIE, trauma, hemolytic-uremic syndrome, infections, and MELAS. Chronic causes include SNE, Huntington disease, Wilson disease, acanthocytosis, SSPE, glutaric aciduria, and familial metabolic disorders. The pathological findings were necrosis and neuronal loss. CT demonstrated hypodensities and the MRI showed hyperintense T2 weighted images in the basal ganglia.

The physiology of basal ganglia disorders is reviewed and an hypothesis for basal ganglia function is proposed by Hallett M, Clinical Director, NINDS, NIH, Bethesda (Can J Neurol Sci Aug 1993;20:177-183). A direct path linking the putamen and globus pallidus is a positive feedback circuit that selects specific motor synergies to carry out a desired action whereas an indirect path inhibits these synergies, eg. dystonia results from overactivity of the putamen and the direct pathway; chorea is explained by underactivity of the indirect pathway.

HEADACHE DISORDERS

MIGRAINE DIAGNOSES AND SYMPTOM PATTERNS

The diagnostic rates based on various criteria and symptom complexes of children and adolescents with chronic headache were evaluated in 88 consecutive patients who attended the Pediatric Headache Clinic at the University of Maryland School of Medicine, Baltimore, 1989-1991. The male/female ratio was 1 (43:45). The mean age 12.5 years; 44 Caucasian and 40 African-American. Clinical migraine was diagnosed in 63 (74%), of which only 14% were classic in type. Migraine versus non-migraine diagnoses were not related to age, gender, or race. When International, Prensky, Vahlquist, and the author's (Gladstein) criteria for migraine were used, the diagnosis was applied in only 50%. Prevalence of migraine diagnosis by the International criteria differed from the other classifications as a function of race, favoring Caucasians over African-American, 61% to 35%. Minority children were less likely to present with nausea, vomiting, lateralized pain, or food as a precipitant of headache. Children older than 12 years complained more often of radiating pain, scalp tenderness, and abdominal pain. Adolescent males reported higher rates of vomiting and decreased appetite than in females, while adolescent females found stress a more frequent precipitant. Older children and adolescents, especially females, used aspirin more frequently than those under 12 years of age. (Gladstein J et al. Diagnoses and symptom

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