In a clinical review of 20 childhood cases of Friedreich's ataxia at the Dept of Child Neurology, Aegean University, Bornova, Izmir, Turkey (Ulku A et al. Acta Neurol Scand June 1988;77:493-7), the mean age at onset was 6.1 years, a positive family history was present in 8 cases, ataxia was the main presenting symptom, and reflexes were depressed or absent in all cases. Electrophysiological studies, especially depressed or absent sensory nerve conduction velocities, were confirmatory of the diagnosis in 9 of 10 patients tested. The EKG was abnormal in 5 (25%).

DEVELOPMENTAL DISORDERS

NEURAL TUBE DEFECTS: NON-CLOSURE V. EARLY CLOSURE

Non-closure of open neural tube defects above L2 in 105 infants born between 1978 and 1985 resulted in a significantly lower incidence ($p<0.001$) of hydocephalus, shunt insertions, and ventriculitis during the first few months of life, and mortality was not increased throughout the first year, in a study reported from the Royal Belfast Hospital for Sick Children, Belfast, Northern Ireland. This non-closure or deferred-closure group was compared with 109 infants born between 1964 and 1971 whose open neural tube defects were treated by early closure. Hydrocephalus correlated with the occurrence of ventriculitis ($p<0.001$) during the first year of life in both non-closure and early-closure groups; 37 of 72 infants with hydrocephalus developed ventriculitis compared with 6 of 37 without hydrocephalus in those whose defect was not closed, and results were similar in those who received early closure. The authors conclude that non-closure of neural tube defects is associated with a better prognosis and a reduction in the number of shunt operations and revisions. (Deans GT, Boston VE. Is surgical closure of the back lesion in open neural tube defects necessary? Br Med J May 21 1988; 296:1441-2).

COMMENT. A rate of infection of 20% or higher is reported with the operative treatment of hydrocephalus (see Ped Neur Briefs, Sept 1987;1(4):28), and patients with myelomeningocele are most susceptible. In those shunted at 1 week of age or earlier, the rate of infection was 48% but when shunting was performed at 2 weeks or later, the incidence of infection was lower. Since non-closure of myelomeningocele appears to be safe and reduces the necessity for shunt procedures, this method of management should be preferred. However, I am sure that other pediatric neurosurgeons have opposing opinions.

CONGENITAL CALLOSAL DEFECTS

A lethal and previously undescribed syndrome in 3 siblings with hypoplasia of the corpus callosum is described from the Istituto Materno-Infantil de Pernambuco, and Laboratorio de Genetica, Universidade Federal de Pernambuco, Recife, PE, Brazil. The combination of anomalies, probably inherited as an autosomal recessive trait, included corpus callosum hypoplasia, microcephaly, severe mental retardation, preauricular skin tag, campodactyly (fixed flexion of one or more fingers), growth retardation, and recurrent

bronchopneumonia. The children lived for 10, 23 and 32 months. At autopsy in 2 patients, the brain weighed approx 500 gms and showed marked hypoplasia of the corpus callosum, aqueductal stenosis, enlarged 3rd and 4th ventricles, widened cavum septi pellucidi, and diffuse degenerative changes with astrocytosis. The literature on anomalies associated with callosal defects is reviewed. (da-Silva EO. Callosal defect, microcephaly, severe mental retardation, and other anomalies in three sibs. Am J Med Genet April 1988;29: 837-843).

COMMENT. Congenital callosal defects occur alone or with other brain anomalies, e.g. septum pellucidum defect, hydrocephalus, porencephaly, polymicrogyria, and cerebellar hypoplasia. Mental retardation, seizures, and failure to thrive are commonly associated, but the callosal defect itself may be asymptomatic. At least 4 distinct syndromes include callosal agenesis as a major component: Aicardi (with infantile spasms, hypsarrhythmia, chorioretinal lacunae and coloboma), Schinzel acrocallosal syndrome (with macrocephaly, and polydactyly), Anderman's syndrome (with anterior horn cell disease), and Shapiro syndrome (with recurrent hypothermia). (See Ped Neur Briefs, March 1988;2:17).

HYPOMELANOSIS OF ITO

The neurological complications in 34 Spanish children with hypomelanosis of Ito are described from the Paediatric Neurology Service, Hospital Infantil La Paz, and La Universidad Autonoma, Madrid, Spain. Most were referred because of mental retardation (65%) and seizures (53%), and the ages at time of the first visit were 2 months to 10 years. Skin lesions, observed within the first year of life in 70% of patients, consisted of hypomelanotic depigmented patches, cafe-au-lait spots, and angiomatous nevi; changes in hair color and alopecia also occurred. Noncutaneous abnormalities, observed in 94%, included macrocephaly, microcephaly, hemihypertrophy, kyphoscoliosis, coarse facial features, and hypertelorism. Autosomal dominant inheritance was demonstrated in some. (Pascual-Castroviejo I et al. Hypomelanosis of Ito. Neurological complications in 34 cases. Can J Neurol Sci May 1988; 15:124-129).

COMMENT. The incidence of this disease was estimated at 1 per 1000 new patients consulting a pediatric neurology service, or 1 per 10,000 unselected patients in a children's hospital. It affects all races, but fair-skinned individuals may require a Woods lamp examination to detect the cutaneous lesions.

BRAIN LESIONS IN THE NEWBORN

NEUROPATHOLOGY OF PRENATAL BRAIN DAMAGE

Autopsy results of 89 infants who died at 7 days of age or less were analysed in the Depts of Pathology and Pediatrics, University of California School of Medicine, Davis, CA. Twenty-two (25%) showed evidence of prenatal brain lesions; 10 (16%) were preterm and 12 (48%) were term infants. Term infants were affected more often than