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