COMMENT. The diagnostic criteria for Rett syndrome proposed by the International Rett Syndrome Association (see Ped Neur Briefs 1988;2:29) in April 1988 were separated into 1) necessary, 2) supportive, and 3) exclusion categories. Normal development through the first 6 or 18 months was regarded as a necessary criterion and intrauterine growth retardation and microcephaly at birth were thought to exclude the diagnosis. By these criteria, 4 of the 8 cases described here would not be accepted as examples of Rett syndrome or, alternatively, a "forme fruste" atypical variety of the syndrome might be recognized. Even the female sex is no longer considered a necessary diagnostic criterion and a less restrictive symptom complex is proposed by some. The occurrence of a similar history and syndrome is boys is not uncommon. A plethora of publications on Rett syndrome has appeared in the last 12 months but none has uncovered a specific cause. The present authors note that pertussis vaccine was considered causative in 7 of 19 girls with Rett syndrome reported from Scotland (Br Med J 1985; 219:579), and the onset of regression heralded by inconsolable screaming attacks had followed recent pertussis immunization in 1 patient in their series of 8.

SPINOCEREBELLAR ATAXIA

The onset below 15 years of age of autosomal dominant spinocerebellar ataxia (SCA) in 6 of 41 affected patients is reported from the Dept of Pediatrics, Baylor College of Medicine, Houston, TX. Linkage analysis was performed on 93 individuals in a seven-generation kindred, and strong evidence for linkage of the SCA to the human leukocyte antigen loci on the short arm of chromosome 6 was documented. Age at onset was 6 to 15 years, and clinical findings included ataxia, dysmetria, dysdiadochokinesia, intellectual deficit, ophthalmoparesis, dysarthria, dysphagia, amyotrophy, and cerebellar atrophy on CT scan. Progression was rapid, 1 patient dying 3 years after the onset of symptoms at age 12 years, and 2 patients aged 15 to 26 years are terminally ill. None had seizures, retinal degeneration, or optic atrophy. Poor intellectual performance preceded other neurological abnormalities in 5 children. Of the 6 patients with juvenile onset, 5 were offspring of affected males. This was the first report of childhood onset in the HLA-linked form of SCA. (Zoghbi HY et al. Spinocerebellar ataxia: variable age of onset and linkage to human leukocyte antigen in a large kindred. Ann Neurol June 1988;23:580-584).

COMMENT. The authors comment that all families with dominantly inherited SCA should undergo genetic studies to determine linkage to HLA. Families with the HLA-linked form of SCA may be advised to have HLA typing for presymptomatic or prenatal diagnosis.

The spinocerebellar ataxias are a heterogeneous group of diseases characterized by a slowly progressive loss of neurons in the cerebellum. Friedrich's ataxia, an autosomal recessive trait, presents in childhood, whereas the autosomal dominant varieties of SCA, described by Marie, are usually distinguished by an onset in adult life.

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