cerebellar involvement in behavioral disorders is unknown. (Gaffney GR et al. Cerbellar structure in autism. AJDE 1987; 141:1330-1332).

COMMENT. The recognition of an organically based dysfunction of the brain as a major causative factor in infantile autism is gaining favor. It may be necessary to redefine the syndrome when the causal mechanisms are better understood and when primary and secondary symptoms have been distinguished (Rutter M. J Autism Child Schizophr 1978;8:162).

DEGENERATIVE AND METHBOLIC DISORDERS

EEG IN RETT'S SYNDROME

The electroencephalographic (EEG) characteristics of Rett's syndrome were studied in 17 girls between the ages of 1 and 16 yrs at the Sections of Neurophysiology and Pediatric Neurology, Baylor College of Medicine and The Methodist Hospital, Houston, TX. The criteria for the diagnosis of Rett's syndrome included: (1) normal prenatal and perinatal development (2) normal neurological development for the first 7 to 18 mths; (3) cessation of development between 1 and 4 yrs of age and subsequent regression; (4) dementia and autistic features, (5) loss of purposeful use of the hands and development of stereotypical movements; (6) ataxia; (7) acquired microcephaly (8) seizures.

A specific diagnostic EEG pattern was not seen but serial records were characterized by a progressive deterioration: (1) slowing; (2) loss of vertex transients and spindles in sleep; (3) multifocal epileptiform abnormalities; and (4) almost continuous generalized slow spike-and-wave activity. These EEG patterns appeared to correlate with the clinical stages: (1) early onset stagnation; (2) rapid destructive; (3) pseudo-stationary; and (4) late motor deterioration (see Am J Med Genet 1986 (suppl). The EEG of 3 patients was not typical and the average age at onset of their symptoms was later than usual. The authors suggest that the EEG may help to identify variants or atypical cases of Rett's syndrome (Glaze DG et al. Rett's syndrome. Correlation of EEG abnormalities with clinical staging. Arch Neurol 1987;44:1053-1056).

COMMENT. Others have described similar age-related changes in the EEG's of patients with Rett's syndrome. This EEG classification correlated with clinical stages may be useful in diagnosis and prognosis. The imprecise nature of the clinical-EEG correlation, admitted by the authors, may be explained by the nonspecific character and frequency of atypical cases of Rett's syndrome of undetermined etiology.

ATAXIAS

CEREBELLAR ATAXIA, OPSOCLONUS, AND NEUROBLASTOMA

A 20 month-old girl with cerebellar ataxia and opsoclonus associated with neuroblastoma is reported from the Pediatric Neurology

Unit, Tel Aviv Medical Center, Israel. Ataxia, present since 1 year of age, and irregular, "jerky" eye movements, noted on admission, became worse over a 2 month observation period. An abdominal mass found at 20 months and removed at operation was a ganglioneuroblastoma. Following surgery, steroids for 3 weeks, and chemotherapy 1 year, blood pressure returned to normal immediately and the ataxia and opsoclonus disappeared within 6-7 weeks. At a 2 year follow-up, the neurological and general examinations were normal. (Harel S et al. Cerebellar ataxia and opsoclonus as the initial manifestations of myoclonic encephalopathy associated with neuroblastoma. Child's Nerv Syst 1987;3:245-247).

COMMENT. Opsomyoclonus or "dancing eye syndrome", also known as myoclonic encephalopathy of infancy, is frequently of undetermined etiology. It may follow viral infection and it is sometimes associated with occult malignancies, notably neuroblastoma. Normal urinary catecholamines do not exclude the presence of tumor and repeat evaluations including radiographs of abdomen and chest are indicated. The acute stage of the dancing eye syndrome usually responds best to ACTH followed after a few weeks by prednisone. Steroids may need to be continued for several months.

DEGENERATIVE ATAXIC DISORDERS

Harding AE at the Institute of Neurology, London, author of the Hereditary Ataxias and Related Disorders (Edinburgh, Churchill Livingstone, 1984) reviews the classification, causes, clinical characteristics and treatment of degenerative ataxias. A combination of genetic and environmental factors is the most common origin for this complex group of over 50 distinct diseases, subdivided according to clinical and genetic features. Metabolic defects such as arylsulfatase-A in metachromatic leukodystrophy are recognizable but untreatable but some deficiency diseases (e.g. Vitamin E) are amenable to treatment with supplements. The cause of Friedreich's ataxia, an autosomal recessive disorder, is unknown and reported deficiencies of pyruvate dehydrogenase and mitochondrial malic enzymes have not been confirmed. Similarly, in olivoponto-cerebellar atrophy, a late onset ataxia, recent studies have not confirmed an earlier report of reduced leucocyte glutamate dehydrogenase activity. Most attempts at treatment of degenerative ataxias have been disappointing but promising results using thyrotropin releasing hormone have been reported from Japan. (Harding A. Degenerative ataxic disorders: still perplexing. BMJ 1987; 295:1223-4).

COMMENT. Degenerative ataxias resembling Friedreich's ataxia that may be amenable to treatment include Vitamin E, B12, folate and biotin deficiencies and Refsum's disease, responsive to a diet low in phytol and phytanic acid.