

cerebellar involvement in behavioral disorders is unknown. (Gaffney GR et al. Cerebellar 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 METBOLIC 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