COMMENT. The recognition of the partial epilepsies of childhood is important from the point of view of treatment and prognosis. In children with infrequent seizure occurrence and a diagnosis of benign partial epilepsy the use of potentially toxic anticonvulsant medications may be deferred.

GENETICS OF FEBRILE SEIZURES

Waking and sleep EEGs were recorded in 67 siblings of 52 patients with febrile seizures (FS) at the Epilepsy Centre Bethel, Bielefeld, F.R.G. Epileptic activity was recorded in at least one sibling of 54% of patients. Epileptic discharges were noted in 50% of the 67 siblings; only in waking in 3%, in waking and sleep in 31% and in sleep only in 15%. The greatest number of epileptic discharges were noted in the waking state during hyperventilation (33%) and during sleep stage C (39%). Of nine patients with complicated febrile seizures 56% had at least one sibling with seizure discharges and of 43 patients with simple febrile seizures 54% had a sibling with seizure discharges. Of nine patients with later onset epileptic seizures, 44% had seizure discharges in the EEG. Siblings age six to ten years had the highest rates of activation (67%). Epileptic discharges occurred in 83% of siblings with seizures, but in only 46% of those without seizures. Epileptic activity in patients and in siblings was significantly greater in those who had occipital slow wave activity in the EEG. A multifactorial inheritance of EEG abnormalities was suggested and the genetic factor was important in patients with both simple or complicated FS. Exogenous brain damage may act as an accidental event or catalytic effect leading to the activation of FS, mental retardation, neurologic deficits, behavior disturbances, focal FS, and/or a higher rate of later epileptic seizures (Degen R. et al. A contribution to the genetics of febrile seizures: waking and sleep EEG in siblings. Epilepsia July/August 1991; 32:515-522).

COMMENT. The importance of sleep EEGs and other activating procedures in the prediction of prognosis in patients with febrile seizures is demonstrated by this study. Reports minimizing the value of the EEG in prognosis and later occurrence of epilepsy are often based on EEGs in the waking state only, and without sleep recordings.

A case control study of risk factors for febrile seizures in the People's Republic of China is reported from the Neurological Institute, Sun Yat-Sen University of Medical Sciences, Guangzhou, Beijing Neurosurgical Institute, Beijing, China; National Institutes of Health, Bethesda, MD; and the World Health Organization, Geneva, Switzerland. Information was gathered by door-to-door survey in six major cities in the People's Republic of China (Zhao F et al. Epilepsia July/August 1991; 32:510-514). Family history of febrile seizures, birth and postnatal factors were assessed in 182 subjects and in an equal number of matched controls. The odds ratio for risk of FS in children with a family history of FS in a first-degree relative was 10. Apart from maternal acute respiratory infection during the first

trimester of pregnancy none of the other examined factors was associated with an increased risk. Birth asphyxia was not a significant risk factor.

EPILEPTIC ACQUIRED APHASIA

The syndrome of acquired aphasia, dementia and behavior disorder in a child with partial complex epilepsy and continuous spike and waves during sleep is reported from the Service de Pediatrie, et Service de Neurologie, CHUV, Lausanne, Switzerland. The child was observed from age three to 18 years and deterioration in behavior and language occurred between the ages of 3-1/2 and 5 years, coinciding with the period of maximal EEG activity and the presence of almost continuous spike waves during sleep (CSWS). Improvement coincided with the disappearance of CSWS, between the ages of 5-1/2 and 6-1/2 years, and the onset of a unilateral focus that persisted throughout the following years. A definite correlation between the paroxysmal EEG abnormalities and the neuropsychological deterioration and improvement was established retrospectively. (Roulet E, Deonna T et al. Acquired aphasia, dementia, and behavior disorder with epilepsy and continuous spike and waves during sleep in a child. Epilepsia July/August 1991; 32:495-503).

COMMENT. Complex partial seizures with recurrence in adolescence and the persistence of an epileptic focus on the EEG are atypical features of the Landau-Kleffner syndrome but this diagnosis could not be definitely excluded.

RETT SYNDROME

CEREBRAL LIPIDS IN RETT SYNDROME

The lipid membrane composition of cerebral tissue from five patients, age 12-30 years, and from 14 age-matched controls was studied at the Department of Psychiatry and Neurochemistry and Pediatrics, University of Goteborg, Goteborg, Sweden. A selective loss of myelin-associated lipids and an enrichment of gangliosides was demonstrated in the temporal white matter. The ganglioside pattern showed an increase of astroglial cell-associated gangliosides and reduced proportions of gangliosides GD1a and GT1b. The fatty acid compositions of ethanolamine phosphoglyceride, choline phosphoglyceride, and galactosylceramide were normal. (Lekman AY, Hagberg BA, Svennerholm LT. Membrane cerebral lipids in Rett syndrome. Pediatr Neurol May/June 1991; 7:186-190).

COMMENT: There is as yet no specific biochemical marker for Rett syndrome. These changes in lipid membranes are thought to be secondary, confirming the neuropathologic finding of slight demyelination in Rett syndrome. PET studies performed on six patients with Rett syndrome at the Division of Child Neurology, National Center Hospital for Mental Disorders, Kodaira, Tokyo, Japan showed that