

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 Pédiatrie, 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

cerebral blood flow and oxygen metabolism were abnormal and the cerebral metabolic rate of oxygen and oxygen extraction fraction were low. Impaired oxidated metabolism in Rett syndrome is suggested by this study. (Yoshikawa H et al. J Child Neurol July 1991; 6:237-242).

A significant increase in B-endorphin in the cerebrospinal fluid, an elevation of the CSF/Plasma B-endorphin ratio and a decrease in CSF cortisol are reported in a study of 15 girls with Rett syndrome at the Unite de Neuropediatrie, Centre Gui de Chauliac, Montpellier, France. The plasma cortisol and B-endorphin levels were similar in the patient and control groups (Echenne B et al. J Child Neurol July 1991; 6:257-262). The authors comment that the inconsistent nature of the CSF endorphin increase exclude its use as a biologic marker of Rett syndrome.

## LEARNING AND BEHAVIOR DISORDERS

### **TRIAL OF METHYLPHENIDATE AND THIORIDAZINE**

A double-blind, placebo-controlled, cross-over study of methylphenidate (0.4 mg/kg/day) and thioridazine (1.75 mg/kg/day) in 27 intellectually subaverage children is reported from The Nisonger Center for Mental Retardation and Developmental Disabilities, Ohio State Univ., Columbus, OH, and the University of Auckland, New Zealand. IQs ranged from 30-90 with a mean of 54 of the Stanford Binet, Form L-M. Ages ranged from 4 to 16 years; 22 boys and 5 girls. ADD-H was diagnosed in 21 and the hyperactivity was severe (a score of 19 on the Conners Parent-Teacher Questionnaire). Epilepsy was treated with anticonvulsants in 4. Methylphenidate improved accuracy on a memory task, reduced omission errors on an attentional task, and reduced seat movements on 2 tasks. Thioridazine had no adverse effect on IQ and cognitive-motor performance tests. (Aman MG et al. Methylphenidate and thioridazine in the treatment of intellectually subaverage children: effects on cognitive-motor performance. J Am Acad Child Adolesc Psychiatry September 1991; 30:816-824).

**COMMENT.** Methylphenidate and thioridazine are two of the most commonly used psychotropic drugs in treating mentally retarded children. In this study involving children with mild developmental delays, methylphenidate improved attention and motivation and thioridazine at relatively low dose levels had no adverse effects on learning performance. A combination of methylphenidate AM and Noon and thioridazine PM is sometimes recommended in children with ADHD of normal intelligence who exhibit behavioral problems both during and after school hours. The present study corroborates the probable benefits of this combination therapy.