vomiting, hypotonia, dystonia, and aphasia. Deep tendon reflexes were exaggerated and plantar responses extensor. The abnormal neurologic signs and basal ganglia lesions gradually improved with corticosteroid treatment and almost disappeared after one year. The right optic fundus showed chorioretinitis. A stereotactic brain biopsy performed to exclude a neoplasm revealed proliferation of astrocytes of undetermined origin, either reactive or low-grade astrocytoma. The dystonia subsided after one year but peculiar mouthing behavior persisted. A CNS primary lymphoma could not be excluded. (Hirabayashi S et al. Basal ganglia mass lesions in juvenile rheumatoid arthritis. Pediatr Neurol March/April 1991; 7:141-3).

COMMENT. A chronic inflammatory process involving cerebral vessels was suspected in this patient but angiography failed to demonstrate a cerebral vasculitis.

Other rheumatic diseases with CNS complications include lupus erythematosus, polyarteritis nodosa and rheumatic fever. Seizures are a common presenting sign of lupus erythematosus.

## SEIZURE DISORDERS

### BENIGN FAMILIAL NEONATAL CONVULSIONS

Linkage studies with the chromosome 20 markers D20S19 and D20S20 were performed in two families with benign familial neonatal convulsions at the Department of Pediatrics, The University of Texas Health Science Center, San Antonio, TX. In the first family with 14 affected, none had seizures after two months of age. In the second family with 13 affected, seizures did not remit until 6 to 24 months; febrile convulsions occurred in two, and one had refractory epilepsy until late adolescence. In family one, the odds were greater than 20,000:1 against linkage at 10% recombination; whereas the data from family two favored linkage with a maximum odds ratio of 45:1 at 6% recombination. It was concluded that this autosomal dominant primary epilepsy of infancy is clinically and genetically heterogeneous. (Ryan SG et al. Benign familial neonatal convulsions: Evidence for clinical and genetic heterogeneity. Ann Neurol May 1991; 29:469-473).

COMMENT. These data based on large family pedigrees suggest two distinct genetic loci for benign familial neonatal convulsions. The subtype linked to chromosome 20q may be associated with delayed remission and a higher risk for the development of epilepsy. The authors suggest that absence and benign rolandic epilepsy might also show genetic heterogeneity.

### EPIDEMIOLOGY OF ABSENCE EPILEPSY

A population based electroencephalographic study of absence epilepsy in 97 children is reported from the Departments of Neurophysiology and Pediatrics, Goteborg University, Sweden. All patients had regular bilaterally synchronous and symmetrical 2-4 Hz spike-and-slow wave discharges and absences with or without generalized tonic-clonic

seizures (GTCS). Patients with absence seizures not complicated by GTCS had long episodes of 2-4 Hz spike-and-slow wave discharges (more than 10 seconds). Posterior delta rhythms occurred only in patients with pure uncomplicated absence whereas focal abnormalities were predictive of GTCS complication. There was no correlation between poly spike and slow wave and the development of GTCS. Brief episodes of 2-4 Hz spike-and-slow wave (less than 10 seconds) were predictive of a two-fold increased risk of GTCS. GTCS appeared in spite of a normalized EEG in seven of 14 patients. Favorable seizure control was correlated with the normalization of EEG but a normal EEG was no guarantee that GTCS would not develop. (Hedstrom A, Olsson I. Epidemiology of absence epilepsy: EEG findings and their predictive value. Pediatr Neurol March/April 1991; 7:100-4).

COMMENT. This study demonstrates the predictive value of the EEG in prognosis of absence epilepsy. Posterior delta rhythm and long episodes of spike-and-wave with clinical correlates favor a good prognosis whereas brief spike-and-wave discharges without clinical correlates increase the risk of future generalized tonic-clonic seizures.

## LEARNING DISABILITIES

### ATTENTION DEFICIT DISORDER (ADD): METHYLPHENIDATE TREATMENT

The clinical response to three dose levels of methylphenidate (5, 10, and 15 mg BID) in 23 children with ADD+H and 17 children with ADD-H is reported from the Department of Psychiatry, University of Massachusetts Medical Center, Worcester, MA. Both groups of children with ADD showed significant improvements in behavior, inattention, self-control, and academic performance. A low dose was as effective as the moderate or high doses in changing the behavior at school but moderate to high doses were needed to produce improved behavior at home and better task performance on the clinic assessment battery. The greatest effect of medication on the ADD-H group occurred at the low dose. In contrast, the drug effect for the ADD+H group was linear, improving with each dose increase. In subsequent clinical recommendations drug treatment was prescribed more often in the hyperactive group (71% of children) than in those without hyperactivity (24%). Children with ADD+H were rated as having more pervasive behavioral problems than the children with ADD-H. Children with ADD+H were impaired in behavioral inhibition and vigilance whereas children with ADD-H were more impaired in the retrieval of verbally learned material. (Barkley RA et al. Attention deficit disorder with and without hyperactivity: Clinical response to three dose levels of methylphenidate. Pediatrics April 1991; 87:519-531).

COMMENT. This study confirms previous reports that the most active children with ADDH respond better to methylphenidate than those with minimal hyperactivity. (Millichap JG. Learning Disabilities and Related Disorders. Yearbook Publishers, Chicago. 1977).