included adequate caloric intake, respiratory support, and control of pain, hypertension, hyponatremia, and self-mutilation. Gavage with high carbohydrate, high calorie feeds without phenylalanine or tyrosine were employed. Barbiturates and other agents contraindicated in porphyria were avoided.

JUVENILE MULTIPLE SCLEROSIS

A young girl with recurrent episodes of CNS demyelination associated with defective mitochondrial beta oxidation is reported from the Departments of Pediatrics, Medical Genetics, Oregon Health Sciences University, Portland, and University of Iowa Hospitals, Iowa City, IA. The child was well until age 14 months when she began having episodes of ataxia with slurred speech and extreme irritability lasting hours to days. At 19 months she was admitted in coma and a CT revealed periventricular loss of white matter. She recovered within three weeks after treatment with immunoglobulins, Acyclovir, and corticosteroids. A similar episode occurred at 22 months of age and the MRI had increased signals in the periventricular and frontoparietal areas. Many episodes of ataxia, slurred speech, painful bright red hands and feet, furrowed tongue and extreme irritability occurred from age 22 to 38 months. The episodes lasted from hours to weeks and were associated with an acrid body odor. Urinary sarcosine was elevated and an increase in ethylmalonic acid in the urine pointed to a disorder affecting fatty acid metabolism. Metabolic evaluations and decreased oxidation of palmitate demonstrated defective mitochondrial beta oxidation. The patient was treated and remained stable for 30 months on a low fat high carbohydrate diet, L-Carnitine (100 mg/kg/d), and Riboflavin (20 mg/kg/d). (Powell BR et al. Juvenile multiple sclerosis-like episodes associated with a defect of mitochondrial beta oxidation. Neurology March 1990; 40:487-491).

COMMENT. The present patient appears to represent a unique disorder of beta oxidation producing multiple sclerosis-like episodes. The youngest patient with classic multiple sclerosis previously reported was two years old. Duquette P et al reported the clinical profiles of 125 children with multiple sclerosis (J Pediatr 1987; 111; 359) (see Ped Neur Briefs September 1987; 1:25-26). The diagnosis of MS should be considered especially in girls with initial sensory or visual symptoms that remit completely and later evolve in a relapsing-remitting manner. Oligoclonal bands in the CSF are the best single laboratory test for the presence of abnormal IgG in patients suspected of having MS. MRI is superior to CT in diagnosis.