was made between two forms of utilization behavior: 1) an incidental form, as exhibited by the patient; and 2) an induced form, occurring with Lhermitte's procedure where the examiner stimulates the palm and fingers of the patient's hands with the object. (Shallice T et al. The origins of utilization behavior. Brain Dec 1989; 112:1587-1598).

COMMENT. Utilization behavior investigated in this adult patient might also be evaluated in children with learning and memory disorders and may assist in the neuroanatomical localization of lesions in the frontal lobe. Lhermitte's neuroanatomical account of utilization behavior is based on the theories of Denny-Brown and a possible imbalance between the activities of frontal and parietal lobes. Visual stimuli activate parietal lobe systems which in turn initiate actions normally inhibited by frontal lobe systems. Damage to the frontal lobes leading to unmodulated effects of parietal systems may result in utilization behavior. Children with minimal brain dysfunction and hyperactivity have an increased tendency to touch and toy with articles within their reach.

METABOLIC DISORDERS

PERIPHERAL NEUROPATHY AND HEREDITARY TYROSINEMIA

Neurologic crises in 48 children with tyrosinemia identified on neonatal screening since 1970 are described from the Departments of Genetics, Hopital Sainte Justine, Hopital de Chicoutimi; and Universite Laval, Quebec, Canada. Neurologic crises had occurred in 20 (42%) and began at a mean age of one year leading to 104 hospital admissions. Abrupt episodes of peripheral neuropathy were characterized by severe pain with extensor hypertonia (75%), vomiting or paralytic ileus (69%), muscle weakness (29%), and self-mutilation (8%). Fourteen patients died due to complications of respiratory insufficiency or mechanical ventilation. In 5 patients undergoing hepatic transplantation none had neurologic crises. Urinary excretion of gamma aminolevulinic acid, an intermediate of porphyrin biosynthesis, was elevated during crises and during asymptomatic periods. Axonal degeneration and demyelination were demonstrated by EMG, NCS and neuromuscular biopsies. (Mitchell G et al. Neurologic crises in hereditary tyrosinemia. N Engl J Med Feb 15, 1990; 322:432-437).

COMMENT. This study indicates that episodes of acute severe peripheral neuropathy are common in hereditary tyrosinemia and resemble crises of the neuropathic porphyrias. Hepatic transplantation eliminated the neurologic crisis, the major cause of mortality in these patients.

Abnormal porphyrin metabolism appears to underlie the neurologic crisis of tyrosinemia and is associated with elevated gamma aminolevulinic acid excretion. Therapy for the neurologic crises

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.