the risk was greatest in the first year. Factors related by multivariate analysis to relapse were neurologic abnormalities and organic etiology, mental retardation, seizure type (infantile spasms, absence seizures), and appearance or persistence of EEG abnormalities during the course of the illness and before discontinuation of the drugs. (Matricardi M et al. Outcome after discontinuation of antiepileptic drug therapy in children with epilepsy. Epilepsia October 1989; 30:582-589).

COMMENT. The authors believe that drug withdrawal can be attempted in patients with well controlled idiopathic epilepsy, without signs of brain damage and without persistent EEG abnormalities. They stress that predictive factors must be considered to individualize the risk of relapse for each patient.

METHYSERGIDE AND INFANTILE SPASMS

A trial of antiadrenergic and antiseretonergic drugs in the treatment of 24 newly diagnosed and previously untreated infantile spasm patients is reported from the Epilepsy Research Center, Section of Neurophysiology, Department of Neurology, Baylor College of Medicine; the Methodist Hospital; and Texas Children's Hospital; Houston, TX. Response to therapy was determined with 24 hour polygraphic/video monitoring techniques and was defined as complete control of spasms and disappearance of the hypsarrhythmic EEG pattern. Two of 12 patients treated with alpha-methylparatyrasene and one of 12 treated with methysergide showed a response. (Hrachovy RA et al. Treatment of infantile spasms with methysergide and alpha-methylparatyrasene. Epilepsia October 1989; 30:607-610).

COMMENT. The hypothesis that infantile spasms may result from a dysfunction of monoaminonergic neurotransmitter systems is not exactly confirmed by the results of this study. However, the authors were impressed that the patients responding to treatment were not in spontaneous remission. Methysergide cannot displace ACTH as the treatment of choice in infantile spasms.

NUTRITIONAL DEFICIENCIES AND SEIZURES

BIOTIN RESPONSIVE ENCEPHALOPATHY

A case of biotin responsive infantile encephalopathy is reported from the Department of Pediatrics and Child Neuropsychiatry, University of Verona, Italy; and Hopital des Enfants Malades, Paris, France. At one month of age the infant developed dermatitis of the ears. At two months she began to have tonic clonic seizures occurring several times a day and refractory to treatment with carbamazepine, phenobarbital, phenytoin, clonazepam, nitrazepam, ACTH, and hydrocortisone. Seizure frequency increased up to ten per day. At three months, she became hypotonic and a CT scan showed enlargement of cortical sulci and lateral ventricles. At four months she was very lethargic and floppy, reflexes were hyperactive, and plantar responses were extensor. Her behavior was autistic-like and her scalp hair was sparse. The urine examination

showed an increased excretion of 2-ketoglutaric acid and 3-hydroxy-isovaleric acid. Serum biotinidase activity was 0.15 nmol min^{-1} ml^{-1} (normal range 5.2). Father's biotinidase activity was 0.31 (8% of normal) and the mother's 0.42 (10% of normal). An electroencephalogram showed frequent independent spikes of variable amplitude prominent in the left posterior temporal region and numerous EEG seizures. Within 36 hours of starting biotin therapy 5 mg BD there was dramatic clinical improvement; the infant became responsive to surrounding stimuli, seizures were controlled and antiepileptic treatment was reduced to only phenobarbital 15 mg BD. After ten days of treatment the urinary examination was normal and the EEG showed a well organized background activity and no paroxysmal abnormalities. At two years four months the neurological exam was normal, the CT scan and EEG normal, and the dose of biotin was at 7.5 mg a day. The authors suggest a therapeutic trial of biotin in all drug resistant infantile seizures. (Colamaria V et al. Biotin-responsive infantile encephalopathy: EEG-polygraphic study of a case. Epilepsia October 1989; 30:573-578).

COMMENT. Two forms of biotin responsive encephalopathy are reported. 1) neonatal holocarboxalase synthetase deficiency (HCSD) and 2) late onset infantile or juvenile biotinidase deficiency (BD). HCSD patients have vomiting, lethargy, and hypotonia associated with metabolic ketoacidosis, hyperammonemia, and organic acidemia. BD infants present with seizures, ataxia, skin rash and alopecia. Seizures are reported in two of five HCSD cases and in 15 of 28 BD cases. The authors stress that the epileptic symptomatology may be the first clinical feature in BD cases. Myoclonias, auditory myoclonus, and repetitive startles documented in the present case were thought to be nonepileptic in nature.

LANGUAGE DISORDERS AND EPILEPSY

OROMOTOR DYSPRAXIA IN BENIGN CHILDHOOD EPILEPSY

A six year old right handed boy with prolonged intermittent drooling, oromotor dyspraxia, and benign childhood epilepsy with centrotemporal spikes is reported from the Departments of Pediatrics and Neurology, Centre Hospitalier, Universitaire Vaudois, Lausanne, Switzerland. Seizures began on the third day of life and were controlled with phenobarbital. Febrile seizures began at eight months and recurred 12 times up to six years of age. At first the seizures were generalized but after four years of age they were partial motor, involving the face and sometimes the right arm. The drooling probably resulted from hypersalivation and oromotor dyspraxia. The fluctuant course of the symptoms and correlation with intensity of the paroxysmal discharges on EEG were consistent with an epileptic dysfunction located in the lower rolandic fissure. No lesion was demonstrated by MRI. (Roulet E, Deonna T, Despland PA. Prolonged intermittent drooling and oromotor dyspraxia in benign childhood epilepsy with centrotemporal spikes. Epilepsia October 1989, 30:564-568).