ketonuria. Treatment with carnitine and biotin was followed by clinical improvement and normal urine organic acids. One patient in the early onset group of the London study was treated with biotin and lived to 6 years of age. The outcome for patients with propionic acidemia is generally poor, although there is a wide variation.

Propionic acidemia (ketotic hyperglycinemia) should be distinguished from non-ketotic hyperglycinemia which is characterized by hypotonia, lethargy and seizures beginning on the first day of life. Some infants succumb within a few weeks, whereas others develop mental retardation and extrapyramidal signs (Menkes JH. Textbook of Child Neurology 3rd Ed., Philadelphia, Lea & Febiger 1981).

LEIGH ENCEPHALOPATHY

Histologic and biochemical analyses of muscle biopsies from 33 patients with Leigh encephalopathy were performed at the National Institute of Neuroscience, Tokyo and Tokushima University School of Medicine, Japan. Cytochrome c oxidase activity was decreased or absent in 7 patients (21%), 10 patients (30%) had biochemical defects including 2 with pyruvate dehydrogenase complex, 4 with cytochrome c oxidase, 1 with NADH-cytochrome c reductase and 3 with multiple complex deficiencies. None had DNA deletions in the muscle mitochondria. (Nagai T et al. Leigh encephalopathy: histologic and biochemical analyses of muscle biopsies. Pediatr Neurol Sept/Oct 1992; 8:328-332.) (Communications: Dr. Nagai, Division of Ultrastructural Research, National Institute of Neuroscience, NCNP, Kodaira, Tokyo 187, Japan.)

COMMENT. A mitochondrial DNA mutation in the ATPase 6 gene was reported in 7 of 40 patients with neuropathologically or MRI defined Leigh syndrome but no known biochemical defect (Santorelli FM et al. Ann Neurol Sept 1992; 32:467). A high abundance of the "NARP" mutation (neuropathy, ataxia, and retinitis pigmentosa) can cause Leigh syndrome and should be looked for in patients without biochemical defects. In another report, a 7 year old girl presented with a partial pyruvate carboxylase deficiency and basal ganglia lesions compatible with Leigh's disease (Schrank WI et al. Ann Neurol Sept 1992; 32:468). The biochemical defects in Leigh encephalopathy are probably heterogeneous.

NEUROPATHIES

HMSN I: EARLY DIAGNOSIS

The value of nerve conduction velocities in the detection of hereditary motor sensory neuropathy type I (HMSN I) in children at risk was determined in 36 children under 10 years of age at the University of Western Ontario, London, Ontario, Canada. Clinical signs and slowed motor nerve conduction velocities were found in 17 of the 36 children who had 1 parent with

HMSN 1. Four children had slowed conduction velocities at 1 year of age or less. Clinical signs were subtle and included pes planus, distal foot wasting, weakness of ankle eversion and dorsiflexion, and areflexia. In all but 1 of the 17 affected patients the motor nerve conduction velocities were less than 40 m/s. Sensory potentials were abnormal in 7 children with HMSN I at ages 6-7 years. (Feasby TE et al. Hereditary motor sensory neuropathy type I in childhood. J Neurol Neurosurg Psychiatry Oct 1992; 55:895-897). (Reprints: Dr. T.E. Feasby, Department of Clinical Neurosciences, University of Calgary, Foothills Hospital, 1403-29th Street NW, Calgary, Alberta T2N 2T9, Canada.)

COMMENT. These data show that HMSN I can be detected in early childhood. Even at 1 year of age or less the motor conduction velocity is significantly slowed. Abnormal physical findings are found in all children who have slow conduction velocities. The signs may be subtle and not accompanied by disability. Pes planus was the most common foot abnormality in this series. Nerve hypertrophy was uncommon.

Dr. Sghirlanzoni et al., Milan, Italy, report two siblings with HMSN III (Dejerine-Sottas disease) whose parents were both affected with autosomal dominant axonal HMSN II. This family and others cited show the existence of an HMSN III phenotype resulting from the homozygous expressions of HMSN I and II genes. (Neurology Nov 1992; 42:2201).

A 4-year-old child with severe hypertrophic peripheral neuropathy had antibodies to myelin glycoprotein of peripheral nerve (Jelloun-Dellagi SB et al. Ann Neurol Nov 1992; 32:700). This anti-Po glycoprotein activity may have a role in pathogenesis of the neuropathy.

ANTICONVULSANT DRUGS

VIGABATRIN INTRACTABLE EPILEPSY

The relationship between vigabatrin dosage and plasma concentrations, platelet GABA-transaminase inhibition and seizure reduction in 16 children with refractory epilepsy was studied at the University of Cantabria, Santander, Spain. Vigabatrin dosages of 57 mg/kg/day and plasma concentrations of 8 mg/L reduced the GABA-T activity from 13.9 to 5.1 and seizures were reduced from 51 to 22 per month. Seizure reduction was correlated with the dosage but not with the plasma concentration or with platelet GABA inhibition. The initial dose of vigabatrin recommended was 50 mg/kg/day and was increased to 75 or 100 mg/kg/day (Arteaga R et al. Vinyl GABA (vigabatrin): Relationship between dosage, plasma concentrations, platelet GABA-transaminase inhibition, and seizure reduction in epileptic children. Epilepsia Sept/Oct; 33:923-931). (Reprints: Dr. J.A. Armijo at Farmacologia Clinica, Hospital "M. de Valdecilla," E-39008 Santander, Spain.)

COMMENT. A study of vigabatrin in 43 children with intractable epilepsy is reported from the Royal Liverpool Children's Hospital,