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,

England. The results showed that complex partial seizures, with or without secondary generalization, responded best with more than 1/2 achieving a greater than 50% reduction. Generalized tonic-clonic seizures also improved. Absence or myoclonic seizures were not benefited. Only 4 patients (10%) were seizure free on monotherapy with vigabatrin (Gibbs JM et al. Vigabatrin in intractable childhood epilepsy: a retrospective study. Pediatr Neurol Sept/Oct 1992; 8:338-40).

In a study of 45 patients ages 2-21 years treated with vigabatrin in Paris, France, 20 (44%) showed no benefit and 24% were well controlled. Partial seizures were most responsive.

ANTICONVULSANTS AND SERUM CHOLESTEROL

The effect of antiepileptic drugs (AED) on total cholesterol, high density lipoprotein (HDL) cholesterol and triglycerides was studied in 208 epileptic children compared with 175 normal children at the University of Bologna, Italy. In epileptic patients the mean total cholesterol plasma level was higher (195 mg/dl) than that of controls (184 mg/dl). HDL cholesterol and triglyceride levels were not different in the two groups. Carbamazepine and phenobarbital treated patients had significant elevations of mean cholesterol values and valproate treated patients had a significant lowering of cholesterol. An increase in cholesterol in the primidone and phenytoin treated patients was not significant. In a group of 32 patients studied before and during AED therapy the total cholesterol level before therapy was 168 mg/dl and after therapy 203 mg/dl. The highest level (313 mg/dl) of total cholesterol was observed in a boy receiving carbamazepine therapy (Franzoni E et al. Total cholesterol, high-density lipoprotein cholesterol, and triglycerides in children receiving antiepileptic drugs. Epilepsia Sept/Oct 1992; 33:932-935). (Reprints: Professor E. Franzoni, Centro di Neurologia Pediatrica, Universita di Bologna, Via Massarenti 11, 4013B Bologna, Italy.)

COMMENT. Carbamazepine treated patients with a family history of hypercholesterolemia or cardiac infarction should be followed carefully for elevated levels of total cholesterol. The effect of antiepileptic drugs on cholesterol levels during treatment with the ketogenic diet should be carefully monitored, especially when treatment is combined with carbamazepine or phenobarbital.

Carbamazepine appeared to be responsible for a nonconvulsive status epilepticus in a 12 year old epileptic girl, in a report from Washington University School of Medicine, St. Louis, MO (Callahan DJ, Noetzel MJ. Neurology Nov 1992; 42:2198). CBZ level was 7.1 mcg/ml on admission. The absence status was resistant to multiple AEDs and was associated with increased intracranial pressure and transiet MRI abnormalities. CBZ should be avoided in patients with generalized, synchronous, spike-and-wave EEG patterns.