Kazlow PG. Acute hepatic failure in a child treated with lamotrigine. Pediatr Neurol April 1998;18:251-252). (Respond: Dr Philip G Kazlow, Department of Pediatrics, Columbia University, 630 West 168th Street, New York, NY 10032).

COMMENT. This appears to be the first case of hepatic failure caused by lamotrigine in a child. The authors cite two previous reports in adults, both after adding lamotrigine to polytherapy, including carbamazepine and/or valproic acid. The previously administered valproic acid in the present case might possibly have contributed to the lamotrigine high blood level and toxicity. Glucuronic acid metabolism is the metabolic pathway of lamotrigine, and valproic acid blocks the elimination of lamotrigine. Careful monitoring of liver function is recommended in patients treated with lamotrigine.

Outcome of children with cerebral edema caused by fulminant hepatic failure is reviewed from the Children's Hospital of Pittsburgh, PA (Alper G, Jarjour IT, Reyes JD et al. Pediatr Neurol April 1998;18:299-304). Neurologic complications of encephalopathy and cerebral edema were major contributors to mortality in 14 (70%) of 20 children with FHF due to various causes. Orthotopic liver transplantation is recommended with severe and worsening encephalopathy before radiographic evidence of cerebral edema develops.

Effect of L-carnitine on valproic acid concentrations in rat serum, brain, and liver was studied at the Department of Pediatrics, Akita University School of Medicine, and Yuri Kumiai General Hospital; Akita, Japan (Sakemi K, Takada G. Pediatr Neurol April 1998;18:331-333). Supplements of carnitine increased free valproic acid concentrations in the brain and may be expected to potentiate the anticonvulsant effects of valproic acid treatment in humans.

HEREDO-DEGENERATIVE DISORDERS

GENETICS OF CHARCOT-MARIE-TOOTH DISEASE

The nomenclature, classification, and genetic basis of Charcot-Marie-Tooth (CMT) disease are reviewed from the Department of Molecular and Cell Biology, University of Aberdeen Medical School, Scotland. Electrophysiological examination with nerve conduction studies and nerve pathology allow subdivision of CMT disease into two major types, CMT type 1 and type 2. CMT 1, also called hereditary motor and sensory neuropathy (HMSNI) is most common and is characterized by slow nerve conduction velocities (NCV), and "onion bulbs" in nerve biopsy due to demyelination and remyelination. CMT2 (HMSNII) has normal NCVs and myelination but marked axonal degeneration. CMT is both clinically and genetically heterogeneous. The duplication of a 1.5 Mb DNA fragment on the PMP22 gene of chromosome 17p11.2 is associated with more than 70% of CMT1 cases. Point mutations of the PMP22 gene are also associated with CMT 1 phenotype. PMP22 duplication testing is the initial screen in CMT disease diagnosis, accounting for 70% of CMT1 cases. Cx32 screening will uncover the second most common genetic defect. (Bell C, Haites N. Genetic aspects of Charcot-Marie-Tooth disease. Arch Dis Child April 1998;78:296-300). (Respond: Drs Bell and Haites, Medical Genetics, University of Aberdeen Medical School, Foresterhill, Aberdeen AB25 2ZD, Scotland).

COMMENT. **A longitudinal clinical and electophysiologic study of Charcot-Marie-Tooth disease type 1A** with 17p duplication in infancy and early childhood is described from the University Hospital "Marques de Valdecilla," Santander, Spain (Garcia A, Combarros O, Calleja J, Berciano J. Neurology April 1998;50:1061-1067). Twenty at-risk children from 6 unrelated CMT-1A families were examined in the first 5 years of life, and 12 were affected. Initially 2 had symptoms, and 5 were symptomatic at the last exam. MCV and SCV were abnormal in 50% at the beginning and in 83% at conclusion of study. After 2 years of age, all affected children had abnormal MCV, SVC, and F-waves. Serial electophysiologic studies can detect the CMT-1A gene carrier in infancy.

CLINICAL AND GENETIC DIAGNOSIS OF FRIEDREICH'S ATAXIA

The clinical diagnostic criteria and genetic testing for Friedreich's ataxia are reviewed from the National Hospital for Neurology and Neurosurgery, Queen Square, London, UK. The essential clinical diagnostic criteria, after Harding, are onset before 25 years, progressive ataxia, absent tendon reflexes, axonopathy, and dysarthria. Additional criteria include scoliosis, cardiomyopathy, optic atrophy, pes cavus, and diabetes. The gene for Friedreich's ataxia is mapped to chromosome 9q13, with an X25 transcript and GAA mutation repeat in intron 1 of the frataxin gene. The repeat length is correlated with the age at onset and the presence of cardiomyopathy. The frataxin protein may be an iron transporter within the mitochondria. (Wood NW. Diagnosing Friedreich's ataxia. Arch Dis Child March 1998;78:204-207). (Respond: Dr Nicholas W Wood, Institute of Neurology, Queen Square, London WC1N 3BG, UK).

COMMENT. A direct genetic test permits diagnosis of Friedreich's ataxia in forme fruste cases, including those with retained reflexes or onset later than 25 years. The author postulates that the clinical manifestations of Friedreich's ataxia coupled with the nature of the frataxin protein have the hallmarks of a mitochondrial disease.

INFECTIOUS DISORDERS

ACUTE HEMIPLEGIA WITH CHICKENPOX

A case of an 18-month-old girl who developed a right hemiplegia 10 days after onset of varicella infection is reported from the Division of Pediatric Neurology, Istanbul University, Turkey. The child was admitted with hemiparesis following a focal clonic seizure involving the arm and leg. She was afebrile and had healed varicella lesions on the trunk. CT and MRI showed infarction of the left putamen and internal capsule. MR angiography was normal. A mild hemiparesis had persisted at 7-month follow-up. (Yilmaz K, Caliskan M, Akdeniz C et al. Acute childhood hemiplegia associated with chickenpox. Pediatr Neurol April 1998;18:256-261). (Respond: Dr Yilmaz, Bulbuldere Cad, No 5/2 Uskudar, 81130 Istanbul, Turkey).

COMMENT. The authors' review of the literature cites 21 previous reports of hemiparesis and infarct following varicella in children. The interval between the rash and hemiparesis was 10 days to 4 months (mean 9 weeks). EEGs showed focal abnormalities in one half the cases. Angiography had revealed arterial stenoses involving the middle cerebral. Occasional cases of hemiparesis presented before the rash appeared. Pre-eruptive varicella encephalitis with cerebellar ataxia is also reported in a child treated at the Mayo Clinic (Goldston EC, Millichap JG,