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,