eventual clinical severity of younger children with newly diagnosed TS, whereas the number of CT brain abnormalities does not correlate with prognosis. High-signal MRI lesions involving the cerebral cortex are characteristic of TS and correspond to hamartomas and gliotic areas seen pathologically. Periventricular calcific lesions are better visualized with CT than with MRI. (Roach et al Arch Neurol 1987;44:301).

MULTIPLE SULFATASE DEFICIENCY

A 9-year-old girl with a phenotype similar to a mucopolysaccharidosis (MPS) and a clinical history characteristic of late infantile metachromatic leukodystrophy (MLD) is reported from the Department of Neurology, National Defense Medical Center, Taipei, Taiwan, Republic of China; the Developmental and Metabolic Neurology Branch, NIH, Bethesda, Maryland; and Department of Pediatrics (Dr. Horwitz), University of Chicago, Chicago, Illinois. The girl's early history and development were normal up to 18 months of age. Following a high fever with a flu-like illness, her gait became unsteady and broad- based. Gradually her speech became slurred and her vocabulary deteriorated. Examination at 7 1/2 years showed short stature and microcephaly. She was autistic and inattentive, with marked cognitive impairment. She had hyperreflexia, extensor plantar responses, dysmetria, and incoordination. Dysmorphic features suggested MPS but dysostosis multiplex and organomegaly were absent. Funduscopic examination revealed a cherry-red-like spot and yellowish-granular appearance of the retina. Deficient activities of arylsulfatase-A, arylsulfatase-B, iduronate sulfatase, and heparan N-sulfatase in the leukocytes established the diagnosis as MSD. The total urinary content of the glycosaminoglycans was normal, but the concentration of heparan sulfate was increased, stressing the need for qualitative estimations when MSD is suspected. (Soong B-W, Casamassima AC, Fink JK, Constantopoulos G, Horwitz AL. Neurology August 1988;38:1273-75).

COMMENT. Multiple sulfatase deficiency or mucosulfatidosis (MSD) is an autosomal recessive genetic disease affecting the expression of lysosomal sulfatases with consequent accumulation of sulfate-containing glycolipids, glycosaminoglycans, and steroid sulfates in tissues and body tissues. The clinical manifestations represent a combination of 2 diseases: late infantile MLD and MPS. The disorder is rare and the authors cite 20 previous reports of this phenotype.

FRIEDREICH'S ATAXIA AND GLUCOSE METABOLISM

Glucose metabolism was investigated in 21 patients with FA at the Instituto Neurogico, Cattedra di Clinica Medica, Milan, Italy. Abnormalities of glucose tolerance occurred in 5 (23.8%) and 4 were diabetic (19%). By oral glucose tolerance tests, the plasma glucose levels of 5 patients were 140-200 mg/ml 2 hours after glucose ingestion. Plasma insulin levels of glucose-intolerant patients were significantly higher than controls after 180 minutes following glucose ingestion. Plasma glucagon levels of FA patients were higher

than controls, and plasma lactate levels of glucose intolerant FA patients were higher than controls. By intravenous glucose load, glucose responses did not differ significantly between patients and controls. The results showed that FA is associated with insulin resistance, B-cell deficiency, and type I diabetes. The alterations might be genetically linked or metabolically related to the primary defect in FA. (Finocchiaro G et al. Glucose metabolism alterations in Friedreich's ataxia. Neurology August 1988;38: 1292-96).

COMMENT. Friedreich's ataxia, the most common heredodegenerative ataxia, is recessively inherited. The incidence of diabetes in FA patients is high, glucose intolerance during oral glucose tests have been reported previously, but the abnormalities of glucose metabolism have not previously been studied in detail. The abnormalities of glycemic control in FA may be due to the interplay of insulin resistance and B-cell dysfunction or their independent effects.

INFANTILE GANGLIOSIDOSIS

Three sisters with infantile-onset 3 GM1 gangliosidosis are reported from the University of Siens, Italy, and the University of Louvain, Brussels, Belgium. The diagnosis was based on the clinical findings of progressive intellectual deterioration by age 6-8 years, ataxia, spastic tetraparesis, and athetoid-choreiform movements; lysosomal vacuoles in CSF, bone marrow, and conjunctiva; and on decreased activity of serum, leukocyte, and fibroblast B-D-galactosidase and abnormal urinary excretion of oligosaccharides. (Guazzi GC et al. Type 3 (chronic) GM1 gangliosidosis presenting as infanto-choreo-athetotic dementia, without epilepsy, in three sisters. Neurology July 1988;38:1124-27).

COMMENT. Gangliosidosis occurs in 3 forms: 1) infantile, characterized by Hurler's facial features, bony abnormalities, hepatoslenomegaly, cherry-red spot, and progressive neurological signs; 2) late infantile-juvenile, without skeletal changes or marked visceromegaly, but severe intellectual deterioration, ataxia, myoclonic seizures, and retinal degeneration; and 3) dystonic juvenile form. The clinical findings in the present report resembled those in the dystonic form except that the intellectual deterioration was more severe.

TOXIC DISORDERS

FETAL ALCOHOL SYNDROME

Hearing and speech and language development in 14 children with fetal alcohol syndrome were evaluated at the Fetal Alcohol Research Center, Wayne State University School of Medicine, Detroit, Michigan, and the Audiology Division, School of Medicine, University of Colorado Health Sciences Center, Denver, Colorado. Recurrent serous otitis media with hearing loss and speech and language problems occurred in 13, and 4 also had sensorineural hearing loss. The IQ was 70-85 in 7 children and below 69 in 7. The authors conclude that