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