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,
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