COMMENT. HARP syndrome is distinguished from Hallervorden-Spatz disease (HSD) by acanthocytosis and the abnormality of lipoprotein. The authors note that all cases of HARP syndrome have been sporadic and lack the autosomal recessive feature of HSD. For further reports of HARP syndrome, see Progress in Pediatric Neurology II, PNB Publ, 1994, p477.

FRIEDREICH'S ATAXIA WITH RETAINED REFLEXES

Genetic linkage analyses in 11 patients from 6 families with Friedreich's ataxia (FA) phenotype, including cardiomyopathy, but retained reflexes (FARR), are reported from the University of Naples and C Besta Neurological Institute, Milan, Italy; and La Fe University Hospital, Spain. Mean age of onset was 13.5 years. Inheritance was autosomal recessive. All patients had progressive ataxia, dysarthria, dysmetria, scoliosis and pes cavus. FARR mapped to the FA locus on chromosome 9q13-21.1, suggesting that FARR is a variant phenotype of FA. (Palau F et al. Early-onset ataxia with cardiomyopathy and retained tendon reflexes maps to the Friedreich's ataxia locus on chromosome 9q. Ann Neurol March 1995;37:359-362). (Respond: Prof Filla, Clinica Neurologica, Universita Frederico II, via Pansini 5, 80131 Napoli, Italy).

COMMENT. The diagnosis of FARR, a variant of Friedreich's ataxia, should be considered in patients with early onset cerebellar ataxia, cardiomyopathy, and sensory neuropathy. Barbeau found absence of deep tendon reflexes to be a required criterion in the diagnosis of FA (Can J Neurol Sci 1978a;5:57-59), whereas Bell and Carmichael allowed hyperactive reflexes in some cases (Treas Hum Inherit 1939;4:141-281). (Bala V Manyam, personal communication).

INFANTILE LEUKOENCEPHALOPATHY WITH MILD COURSE

Eight children, including 2 siblings, with infantile onset cerebral leukoencephalopathy and megalencephaly, and mild neurological signs and symptoms, are reported from Free University Hospital, and Academic Medical Center, Amsterdam, The Netherlands. Ataxia and spasticity were slowly progressive, while intellectual functioning was preserved for a few years. MRI showed swelling of supratentorial hemispheral white matter, subcortical cysts, and sparing of corpus callosum and internal capsule. Metabolic studies were negative. (van der Knaap MS, Barth PG et al. Leukoencephalopathy with swelling and a discrepantly mild clinical course in eight children. Ann Neurol March 1995;37:324-334). (Respond: Dr MS van der Knaap, Department of Child Neurology, Free University Hospital, PO Box 7057. 1007 MB Amsterdam, The Netherlands).

COMMENT. This type of infantile leukoencephalopathy is distinguished from Canavan and Alexander diseases by an MRI showing severe white matter abnormalities which contrasted with a slow clinical progressive course. Lysosomal and other metabolic white matter disorders characterized by megalencephaly were also ruled out biochemically and clinically.

LATE ONSET KRABBE'S DISEASE WITH PRESERVED INTELLECT in a 24-year-old Swedish male patient is reported from the County Hospital of Jonkoping, and the University of Goteborg, Sweden. (Arvidsson J,