24: 27).

Partington MW (Am J Med Genet March 1988; 29: 633) describes Rett Syndrome in a pair of monozygotic twin girls, pointing out that their development was delayed from birth with no period of normal progress in infancy and subsequent regression, findings at variance with the necessary diagnostic criteria listed above. He states that the cause is not necessarily genetic but could be explained by prenatal toxic or slow viral factors.

Karet D et al. (J Pediat Orthopaedics March/April 1988; 8: 138) reports scoliosis in eight of 10 females with Rett Syndrome treated at the Alfred I. DuPont Institute, Wilmington, Delaware. Curve progression occurred in four and posterior spinal fusion was performed in five. Scoliosis developed at an average age of 11 years and progression was rapid in adolescence. Early surgery is recommended to arrest curve progression and to obtain correction of the deformity.

PEROXISOMAL DISORDERS

Generalized peroxisomal disorders are classified in three main groups in a review article from the Kennedy Institute and the Departments of Neurology and Pediatrics, Johns Hopkins University, 707 N. Broadway, Baltimore, MD. Group 1 includes Zellweger (cerebro-hepato-renal) syndrome, neonatal adrenoleukodystrophy, infantile Refsum disease, and hyperpipecolic acidemia, all characterized by a reduction in the number of peroxisomes and deficiency of multiple peroxisomal enzymes.

Group 2 contains only one rare disorder, rhizomelic chondrodysplasia punctata, characterized by stippled calcification of hyaline cartilage, dwarfing, cataracts, multiple malformations with contractures, koala bear facies, and sever mental retardation. Peroxisomes are normal in number but functionally impaired.

Group 3 includes Refsum disease, X-linked adrenoleukodystrophy, pseudo-Zellweger syndrome, hyperoxaluria type 1, acatalasemia and an undescribed variant. All have a normal number of peroxisomes and the activity of only one peroxisomal enzyme is reduced.

Peroxisomal disorders are a newly recognized and heterogeneous group of diseases with variable manifestations transmitted as autosomal recessive or sex-linked recessive traits and have in common one or more peroxisomal enzyme defects. The term peroxisome is coined from the hydrogen peroxide-forming enzymes found within the subcellular organelle. More than 40 enzymes have now been localized to the peroxisomes. (Naidu S, Moser AE, Moser HW. Phenotypic and Genotypic Variability of generalized peroxisomal disorders. Pediatr Neurol Jan/Feb 1988; 4: 5-12).

COMMENT. This is an excellent review of the various entities now classified as generalized peroxisomal disorders. See Ped Neuro Briefs (March 1988; 2: 22-23, and Oct 1987; 1:32) for case reports of infantile Refsum and Zellweger syndromes.

SPINO-CEREBELLAR DEGENERATION AND CEROID LIPOFUSCINOSIS

Neuronal ceroid lipofuscinosis (NCL) presenting in two different forms within a family is reported from the New York State Office of Mental Retardation and Developmental Disabilities, Institute for Basic Research,