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,

10560 Forest Hill Rd, Staten Island, NY and the Dept of Neurology, Albert Einstein Coll of Med, Bronx, NY. In the proband, the clinical course was compatible with an atypical juvenile form of NCL, beginning with ataxia and spasticity at 4 to 5 yrs, and followed by blindness with optic atrophy, intractable seizures, dementia, and death at 14 yrs. Areflexia, hypotonia, and ataxia were atypical manifestations, suggesting peripheral nervous system involvement similar to that in her two affected siblings. The illness in the siblings, a brother and a sister, showed a more protracted course, a later age of onset (8.5 and 10.5 yrs), more severe cerebellar and cortico-spinal signs, and sensorimotor neuropathy; seizures, dementia and visual loss were lacking. All 3 siblings had cytoplasmic inclusion bodies characteristic of the juvenile form of NCL and increased excretion of urinary dolichol. The authors propose that either variability of gene expression or two different recessive genes in this consanguinous family may account for the divergent phenotypes in the proband and siblings. (Wisniewski KE et al. Spino-cerebellar degeneration with polyneuropathy associated with ceroid lipofuscinosis in one family. J Child Neurol Jan 1988; 2: 33-41).

COMMENT: The diagnosis of neuronal ceroid lipofuscinosis (Batten's disease, Spielmeyer-Vogt-Sjogren syndrome, Kufs' disease) is based on characteristic clinical manifestations, ultrastructural fingerprint cytoplasmic inclusion bodies in the rectal biopsy, punch skin biopsy, and buffy coat of lymphocytes, and elevated urinary dolichol excretion as a biochemical marker. Although the clinical course and manifestations were atypical, the patients in this study exhibited the cytoplasmic inclusions seen in the juvenile variant of ceroid lipofuscinosis. These cases include an unusual presentation as a spino-cerebellar degeneration.

LEARNING AND BEHAVIOR DISORDERS

TEACHERS AND PSYCHOPHARMACOLOGY

Educators at the UCLA Neuropsychiatric Hospital and Inpatient School, Los Angeles, CA, and the Division of Special Education, University of Iowa, IO, discuss the issues in child psychopharmacology that are of importance to teachers and stress the need for greater interdisciplinary collaboration between the medical profession and the schools. The beneficial and adverse classroom effects of four major classes of psychotropic medication are discussed: 1) CNS stimulants; 2) anticonvulsants; 3) neuroleptics or antipsychotics; and 4) antidepressants.

A so-called "metanalysis" of available research data by special educators involved 135 studies of stimulant medications used for treatment of hyperactivity. Benefits were demonstrated not only in attention and memory but also in academic performance. Children on stimulants gained the equivalent of a 15% rank increase in achievement while those treated with major tranquilizers for severe behavior disorders showed a 20% rank increase on various cognitive measures. Other metanalyses of certain classroom interventions such as perceptual motor training or diet treatments resulted in gains of only 5 or 6 percentile ranks. The authors allude to an antimedication bias and the application of different