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

standards to drug studies compared to other classroom intervention techniques. Little systematic data on side-effects of medication were available in the studies analyzed.

Studies of antidepressants were limited and few had valid educational variables or measures suited to the classroom performance. One study demonstrated the importance of interpretation of behavior of children on medication in light of behavior of other untreated children in the classroom. What appeared to be significant effects of medication was actually a reflection of the overall disruptive or inattentive behavior of the whole classroom. (Forness SR, Kavale KA. Psychopharmacologic treatment. A note on classroom effects. J Learn Disabilities March 1988; 21: 144-147).

COMMENT. The pressures of clinical practice sometimes preclude physicians' visits to schools and close collaboration with patients' teachers. The point of this report is that as pediatricians, neurologists and psychiatrists increase their use of psychopharmacological agents, teachers must have increased access to information about the classroom effects and side-effects of these medications. Medical researchers are not always aware of the possible problems that drug treatments may present in classroom situations, and regular reports from teachers can provide valuable information regarding their overall effects.

METHYLPHENIDATE IN HYPERACTIVE AUTISTIC CHILDREN

Nine children, eight boys and one girl, ages 4 to 16 yrs, with a diagnosis of autism, were treated with methylphenidate (10-50 mg/day) as outpatients at Columbia Presbyterian Medical Center, Babies' Hospital Pediatric Psychiatric Clinic, New York. All were hyperactive, impulsive and mentally retarded. All showed significant improvement on the Conner's Teacher and Parent Questionnaire scores during treatment with the stimulant. No significant side-effects were noted or worsening of stereotyped movements. (Birmaher B et al. Methylphenidate treatment of hyperactive autistic children. J Am Acad Child Adolesc Psychiatry March 1988; 27: 248-251).

COMMENT. The beneficial effect of methylphenidate on the behavior of autistic children, at variance with many previous reports, is confirmed in a randomized trial of the drug in a 6-year-old autistic boy reported in the same journal from the Western Psychiatric Institute, Pittsburgh, PA (Strayhorn Jr. JM et al. J Am Acad Child Adolesc Psychiatry March 1988; 27: 244). Negative effects on mood and tantrums were outweighed by positive effects on attention and activity, destructive behavior and stereotyped movements. These reports fail to support previous statements that stimulants are ineffective and contraindicated in hyperactive autistic children. Rutter M (J Child Psychol Psychiatry 1985; 26: 193) writing on treatment refers to a basic cognitive deficit which underlies language and behavior problems in autistic children. Provided that methylphenidate does not exacerbate psychotic behavior, its known effects in promoting cognitive development could be beneficial.