the criteria for diagnosis of Rett syndrome, head circumference at birth, perinatal period, and the first 6 months development may not be normal. (Leonard H, Bower C. Is the girl with Rett syndrome normal at birth? Dev Med Child Neurol Feb 1998;40:115-121).

HEREDO-DEGENERATIVE DISEASES

CLINICAL, MRI, AND GENETIC FINDINGS IN BATTEN DISEASE

The correlation of clinical, MRI and genetic factors in 36 patients with Batten disease (juvenile-onset neuronal ceroid lipofuscinosis) followed up for 25 years is reported from the Department of Paediatric Neurology, University of Helsinki, Finland, and other centers. Twenty seven patients were homozygous and 9 were heterozygous for the major mutation, a 1.02-kb deletion. All patients had vacuolated lymphocytes and positive rectal biopsy findings on the electron microscope. Visual failure occurred at a mean age of 5.8 years (range, 4-10 years) in all patients, and blindness developed between 6 and 20 years of age. Epilepsy began at 8 to 13 years in 92% of homozygous and 55% of heterozygous patients. Mental decline occurred slightly earlier (before 10 years in 50%) in homozygous patients than in heterozygous (mean age 12). Neurological and MRI changes were milder in heterozygotes. Parkinsonian signs were noted in 30% of homozygous patients between 12 and 15 years, and in 22% between 17 and 29 years. Extrapyramidal signs developed in only one heterozygous patient at 19 years. Speech failure correlated with onset of parkinsonism. Ataxia occurred in both homozygous and heterozygous patients before 15 years. Behavioral symptoms, aggression and depression, developed in 52% of homoygotes and 33% of heterozygotes. Death of homozygotes occurred at a mean age of 24 years (range, 10-28 years). MRI abnormalities in homozygotes, cerebral atrophy and gray/white matter ratio changes, developed after 10 years of age in homozygotes. The 1.02-kb deletion in homozygous patients was always associated with mental and neurological handicaps, whereas the heterozygous phenotype was often benign and without intellectual deterioration. Whereas progression of visual impairment and epilepsy was highly concordant, progression of motor and mental deterioration was variable. Environmental and therapeutic factors, and modifying genes might influence the phenotype of Batten disease. (Jarvela I, Autti T, Lamminranta S, Aberg L, Raininko R, Santavuori P. Clinical and magnetic resonance imaging findings in Batten disease: Analysis of the major mutation (1.02-kb deletion). Ann Neurol Nov 1997;42:799-802). (Respond: Dr Jarvela, Laboratory of Human Molecular Genetics, Mannerheimintie 166, 00300 Helsinki, Finland).

COMMENT. Homozygous patients with Batten disease, having the major 1.02-kb deletion, have mental and neurological handicap and a poor prognosis, whereas the heterozygous phenotype may be benign and intellectually normal. The diagnostic DNA test is of value in determining the severity and prognosis of the disease. MRI changes indicative of atrophy and progression of pathology may develop at and after 10 years of age.

INFECTIOUS DISORDERS

MEASLES VACCINE AND ENCEPHALOPATHY

The relationship between acute encephalopathy followed by permanent brain injury or death associated with further attenuated measles vaccine was evaluated in 48 children, ages 10 to 49 months, reported to the National Vaccine