to five years of age she had developed an awkward stiff gait, she became socially withdrawn in personality, had frequent nightmares, speech was dysarthric, and she began to fall frequently and to have problems controlling her hands. Swallowing, chewing and speech progressively deteriorated. She was hypertonic and had exaggerated deep tendon reflexes. Her father had Huntington disease. Her EEG showed bilateral epileptic foci but she had no clinical seizures. PET showed marked reduction in cerebral glucose metabolism in the posterior nuclei of the thalamus, a finding that differs from adults with the disease who show normal or increased rates of thalamic glucose metabolism. These metabolic findings were consistent with previously recognized postmortem pathologic differences between juvenile and adult forms of the disease. (Matthews PM et al. Regional cerebral glucose metabolism differs in adult and rigid juvenile forms of Huntington disease. Pediatr Neurol Nov-Dec 1989; 5:353-356).

COMMENT. The juvenile form of Huntington disease has a more rapid progression than the adult form and is manifested by rigidity rather than chorea. In children, the globus pallidus and thalamus reveal marked degeneration and unlike the adult form the cerebellum and cortex are also involved.

MENTAL RETARDATION SYNDROMES

CAUSES OF MENTAL RETARDATION

The mechanisms of mental retardation with relative prevalence in a hospital referral experience are reported from the Developmental Evaluation Clinic, Children's Hospital, Boston, MA. Early alterations of embryonic development (including Down syndrome) account for 32%, unknown causes 30%, environmental problems (psychosocial deprivation, childhood psychosis) 18%, pregnancy and perinatal morbidity 11%, hereditary disorders 5%, and acquired childhood diseases 4%. This classification uses the timing of the putative noxious event. The patients with mental retardation were obtained from over 3000 children referred for general developmental assessment to a tertiary children's medical center. (Crocker AC. The causes of mental retardation. Pediatr Ann October 1989; 18:623-636).

COMMENT. This issue of Pediatric Annals also includes articles concerning community services for children with mental retardation and special needs adoption agencies.

LAURENCE-MOON-BIEDL SYNDROME

Thirty-two patients with a form of Laurence-Moon-Biedl syndrome are reported from the Departments of Medicine, Ophthalmology, Radiology, and Community Medicine, Memorial University, St. John's, Newfoundland, Canada. The patients were located through the registry of the Canadian National Institute of the Blind, as a result of their attendance at an Ocular Genetics Clinic. Fourteen were male and 18

female with an age range of 12-54 (mean, 33 years). The estimated prevalence was 1 per 17500. The patients were distributed all over Newfoundland but were primarily in families of English West Country origin. Consanguinity was documented or presumed in six families. Mental retardation was present in 41% of patients. All patients had severe retinal dystrophy but only two had typical retinitis pigmentosa. Polydactyly occurred in 58%, 96% were overweight, and 48% were grossly obese. The majority were below the 50th percentile for height. Other abnormalities included small testes and genitalia, menstrual irregularities, low serum estrogen levels, diabetes mellitus, and renal abnormalities. (Green JS et al. The cardinal manifestations of Bardet-Biedl syndrome, a form of Laurence-Moon-Biedl syndrome. N Engl J Med October 12, 1989; 321:1002-9).

COMMENT. The patients were classified as Bardet-Biedl syndrome rather than Laurence-Moon-Biedl syndrome. The authors conclude that the characteristic features of Bardet-Biedl syndrome are 1) severe retinal dystrophy, 2) dysmorphic extremities, 3) obesity, 4) renal abnormalities, and 5) hypogenitalism in male patients only. Mental retardation, polydactyly, and hypogonadism in female patients are not necessarily present.

The earliest reference to this condition appears in II Samuel 21:20, in which there is mention of "a man of great stature, that had on every hand six fingers, and on every foot six toes, four and twenty in number; and he also was born to the giant." (See Millichap JG. Proc Roy Soc Med 1951; 44:1063). This man was probably related to Goliath. Although the Hebrew word describing the stature is generally accepted to signify tall, the Aramaic translation uses a word "mashach", which means oily or fat. Pliny, in his Natural History, also refers to a baby with six fingers and six toes. The cardinal signs of the syndrome described by Laurence and Moon (Ophthal Rev 1866; 2:32), in order of their frequency, were obesity, retinitis pigmentosa, mental deficiency, genital dystrophy, familial incidence, and polydactyly. Spastic paraparesis mentioned in the present article was not included and was not present in my own case report. The early retinal changes do not usually conform with the classical picture of retinitis pigmentosa but these may appear in the later stages of the illness. Early retinal abnormalities are nonpigmented degeneration with loss of central vision, minimal peripheral pigmentation of the fundus, optic atrophy and fine retinal vessels. (Lyle DJ. Amer J Ophthal 1946; 29:939). The need to separate patients in the present paper as examples of Bardet-Biedl syndrome seems questionable. Renal abnormalities have been reported in the Laurence-Moon syndrome and spastic paraparesis is not usually a dominant feature of that syndrome. As in most syndromes, individual expressions are variable and one or more of the cardinal features is often absent.