

to five years of age she had developed an awkward stiff gait, she became socially withdrawn in personality, had frequent nightmares, speech was dysarthric, and she began to fall frequently and to have problems controlling her hands.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