disorders such as obsessive compulsive disorder and attention deficit disorder with hyperactivity occur in 50% of patients and may represent the predominant or only clinical manifestation of the illness. Diagnostic criteria for Tourette syndrome in the DSM-IIIR include 1) multiple motor tics, 2) one or more vocal tics, 3) onset before 21 years of age, and 4)duration more than one year. The Tourette Syndrome Association Workshop participants divided tic disorders into 11 categories: 1) definite Tourette syndrome, 2) Tourette syndrome by history, 3) definite chronic multiple motor or phonic tic disorder, 4) chronic multiple motor or phonic tic disorder by history, 5) chronic single motor or phonic tic disorder by history, 6) definite transient tic disorder, 7) transient tic disorder by history, 8) definite nonspecific tic disorder, 9) nonspecific tic disorder by history, 10) definite tic disorder diagnosis deferred until followed for one year, 11) probable Tourette syndrome. Causes of associated school problems in Tourette syndrome are as follows: 1) primary Tourette syndrome symptoms, 2) obsessive compulsive behaviors, 3) attention deficit hyperactivity disorder, 4) general behavioral disturbances, 5) associated learning disabilities, 6) poor socialization, 7) low self-esteem, and 8) medication side effects. Genetic factors in etiology are recognized and striatal dopamine receptor supersensitivity is suggested as the likely mechanism for tics. Pharmacotherapy should be considered only when symptoms of Tourette syndrome are functionally disabling and not remediable by nondrug interventions. Most patients with Tourette syndrome can probably be managed well without drug therapy and by educating the patients, family members, and school personnel concerning the nature of Tourette syndrome, restructuring the school environment (one on one tutoring) and supportive therapy. Haloperidol is the most commonly prescribed medication for Tourette syndrome but the "reflex" prescribing of this medication at diagnosis of Tourette syndrome should be avoided. (Kurlan R. Tourette's syndrome: Current concepts. Neurology December 1989; 39:1625-1630).

COMMENT. The author correctly notes that the accurate assessment of drug effectiveness in Tourette syndrome is hampered by the natural waxing and waning course of tics and the strong placebo effect of medications. The author's condoning of a combination of haloperidol and methylphenidate in selected patients with attention deficit disorder complicated by tics, a view shared by his colleague from the same institution (Roddy SM. Contemporary Pediatrics. November 1989; 6:22-36) may not receive universal acceptance.

HUNTINGTON'S DISEASE

The positron-emission tomography (PET) findings in a seven year old girl with the juvenile form of Huntington's disease are described from the Department of Neurology and Neurosurgery, Montreal Neurological Institute and Hospital, Montreal, Canada. The birth and early development were normal and at three years of age she could dance and ice skate. By 3½ years she had difficulties in dancing and by four

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