COMMENT. The term Leopard is a mnemonic acronym for the features of the syndrome which may include: L - lentigines, E - EKG abnormalities, O - ocular hypertelorism, P - pulmonary stenosis, A - abnormal genitalia, R - retardation of growth, D - deafness. Mild mental retardation has been reported in patients with Leopard syndrome. Perhaps the "A" in the mnemonic should stand for acalculia in place of "abnormal genitalia". Some neuroradiologists would report the mild asymmetry of ventricles on the CT as a variant of normal and, in the absence of abnormalities of the white and gray matter, the diagnosis of parietal lobe atrophy may be questionable. As the author suggests, Gerstmann syndrome in children may be more common than indicated in the literature. In my experience this syndrome is not infrequent in children presenting with attention deficit disorders and normal CT scans are not unusual. It is possible that the MRI may be more revealing of associated cortical defects.

NEUROFIBROMATOSIS AND THE MRI

The MRI was abnormal in seven of ten children with clinically proved neurofibromatosis reported from the Department of Radiology, the Oregon Health Sciences University, Portland, and the Departments of Neurology, Pediatrics and Radiology, University of Miami School of Medicine. Clinical diagnosis was based on six or more cafe-au-lait spots at least 1.5 cm in size. MRI was indicated because of mental retardation (5 patients), bilateral optic nerve tumors (1), shunt malfunction (1), learning disability (1), and possible brain tumor (2). The MRIs showed increased signal intensity on the T2-weighted images in the globus pallidus, brain stem, and cerebellum. The abnormalities most likely represented hamartomas. (Goldstein SM et al. A new sign of neurofibromatosis on magnetic resonance imaging of children. Arch Neurol November 1989; 46:1222-1224).

COMMENT. The MRI in this study was more revealing than the CT scan which was normal in all except one of the patients studied. The neurologic and developmental examinations showed no correlation with the MRI findings.

## MOVEMENT DISORDERS

TOURETTE SYNDROME

The current concepts of Tourette syndrome, including research diagnostic criteria formulated by a workshop sponsored by the Tourette Syndrome Association, are reviewed from the Department of Neurology, University of Rochester School of Medicine, Rochester, NY. The author concludes that Tourette syndrome is a common, hereditary, neurobehavioral disorder with heterogeneous clinical manifestations. Chronic multiple motor or phonic tic disorder and transient tic disorder represent milder variants of the same illness. Behavioral

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