repetitive movements and mannerisms with grimacing and tics were most conspicuous. Seizures and growth retardation were mentioned in later reports. The cause was unknown but a suspected organic lesion was confirmed in cases examined at necropsy. Diffuse lipoid cell degeneration of the cortical neurons, atrophy of the brain, small disorganized cortical neurons lacking in Nissl bodies, and marginal gliosis have been described, and abnormalities in plasma lipids. Heller dementia is recognized as a syndrome in text books of pediatric neurology and child psychiatry although some have suggested that some examples of the syndrome may have been confused with childhood schizophrenia, and other degenerative brain diseases. Heller dementia occurs in both boys and girls and unlike Rett syndrome, the disorder was not restricted to girls. It is surprising that in publications on Rett syndrome Heller dementia seems to have been overlooked in the differential diagnosis. Drs. Rett and Olsson have now corrected this omission (Dev Med Child Neurol 1987; 29:834) but believe that research will show Rett syndrome to be an independent disorder. They admit that the differentiation may be difficult in older children and that the range of variations of both syndromes cannot be determined until the etiology is known. The suggestion that most cases of Heller dementia were disintegrative psychoses (Stephenson JBP, Kerr AM. Lancet March 28, 1987; 1:741) does not fit with published neuropathological findings, and the introduction of the term "pervasive disintegrative disorder, Heller type and Rett type", based on two patients in the present report, is probably not of value in the classification of these dementias.

NEUROCUTANEOUS DISORDERS

LEOPARD SYNDROME

A 12 year old boy with multiple lentigines (Leopard) syndrome in association with Gerstmann syndrome and CT abnormalities is reported from the Department of Pediatrics and Pediatric Neurology, Beilinson Medical Center, Petah Tikva and Tel Aviv University, Sackler School of Medicine, Petah Tikva, Israel. Learning difficulties were first observed at five years of age. He had a single simple febrile convulsion at three years of age. Several hundred hyperpigmented skin lesions over the face, trunk, and extremities had appeared gradually after birth and had increased in number and size. Mild dysmorphic features included hypertelorism, epicanthal folds, cubitus valgus, and pterygium colli. A systolic cardiac murmur was indicative of pulmonic stenosis and was accompanied by EKG changes. The signs of Gerstmann syndrome included dyscalculia, left/right disorientation, finger agnosia, and dysgraphia. His IQ score was 86. Cranial CT showed dilatation of the left lateral ventricle especially in the occipital horn and mild atrophy of the left parietal lobe. (Garty B-Z et al. Gertsmann tetrad in Leopard syndrome. Pediatr Neurol Nov-Dec 1989; 5:391-392).

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