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MOVEMENT DISORDERS

TOURETTE SYNDROME CLASSIFICATION

A task force (The Tourette Syndrome Classification Study Group), organized by the Tourette Syndrome Association, has expanded the DSM-IIIR definitions of Tourette syndrome and is developing a quantitative rating scale for tics to be used by clinicians in practice and in genetic and other research activities. Each of the four basic DSM definitions (*Tourette syndrome, chronic tic disorder, transient tic disorder, and non-specified tic disorder*) is divided into two categories; "definite," tics witnessed by a reliable observer, and "historical," putative tics not reliably witnessed. Additional tic categories have been included: *chronic single tic disorder; definite tic disorder-diagnosis deferred* (symptoms less than 1 year); and *probable Tourette syndrome* (some criteria for diagnosis absent). (Fahn S et al. Definitions and classification of tic disorders. Arch Neurol Oct 1993;50:1013-1016). (Reprints: Dr Fahn, Neurological Institute, 710 W 168th St, New York, NY 10032).

COMMENT. A similar classification was proposed by a TSA workshop in a previous report (Kurlan R. Neurology Dec 1989;39:1625; see Progress in Pediatric Neurology, Chicago, PNB Publishers, 1991; 230-232).

Most tics are idiopathic but some are symptomatic of head trauma, encephalitis, or other etiologies. Three children with *carbamazepine-induced tics* are reported from the Departments of Pediatrics and Neurology, University of Michigan Medical Center, Ann Arbor, MI (Robertson PL et al. Epilepsia Sept/Oct 1993;34:965-968). The temporal association of repetitive blinking, jaw opening or clenching, and

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tongue movements with the initiation of CBZ treatment for complex partial seizures, and the absence of other etiologies, suggests that CBZ was the likely cause. Additional cases of carbamazepine-related tics and other movement disorders have been reported.

The contingent negative variation (CNV), a slow brain potential which may reflect the level of central dopamine and is a measure of arousal and attention, has been studied in patients with Tourette syndrome (Weate SJ, Drake ME Jr et al. Clin EEG Oct 1993;24:188-191). Patients had higher CNV amplitude and more frequent postimperative negative variation than controls, possibly explained by dopaminergic excess. Medication was not a factor.

BENIGN HEREDITARY CHOREA

The clinical manifestations, differential diagnosis, and treatment of benign hereditary chorea (BHC) are reviewed from the Indiana University and University of Minnesota Medical Schools. The diagnosis of BHC is by exclusion. The most common age of onset is about 1 year and walking is often delayed because of clumsiness and falling. After a short period of progression in severity, movements reach a plateau and may sometimes appear to improve. IQ is normal or low-average. In most reported families, the inheritance is autosomal dominant. Treatment with various drugs, eg. haloperidol, chlorpromazine, and prednisone, is only partially effective, and the chorea is usually refractory. (Wheeler PG, Weaver DD, Dobyns WB. Benign hereditary chorea. Pediatr Neurol Sept/Oct 1993;9:337-340). (Respond: Dr Wheeler, Dept of Medical and Molecular Genetics, IB 130, Indiana University Medical Center, 975 West Walnut St, Indianapolis, IN 46202).

COMMENT. BHC is an uncommon form of childhood chorea that has been mistaken for Huntington disease. The recognition of this benign, nonprogressive form of chorea of early onset and without dementia will avoid unnecessary prolonged hospitalization and the risk of invasive diagnostic procedures.

BILATERAL STRIATAL SYNDROMES

The clinical manifestations and outcome in 13 patients with bilateral basal ganglia lesions and neurological dysfunction are reported from the Child Neurology Unit, Vall D'Hebron University Hospital, Barcelona, Spain. Lesions were demonstrated by CT, MRI, or ultrasound, and in 4 patients who died the pathology was subacute necrotizing encephalomyelopathy (SNE) (2), hypoxic -ischemic encephalopathy (HIE) (1), and intracranial hemorrhage (1). Extrapyramidal signs included dystonia in 9 patients, hypotonia in 2, athetosis 1, and rigidity 1. Consciousness was altered in 5, and seizures occurred in 3.