BENIGN MYOCLONUS OF EARLY INFANCY

To redefine benign myoclonus of early infancy (BMEI), clinical and neurophysiologic features in 102 infants (60 male) with brief paroxysmal abnormal movements and normal neurologic and psychomotor development were studied at one center in Argentina and two in Italy. Infants with movements occurring only during sleep and those with abnormal EEG were excluded. Age at onset was 1-12 months (median 6.2 months). The nonepileptic paroxysmal motor phenomena included myoclonus in 23, brief tonic contractions and spasms in 38, shuddering in 35, atonia or 'negative' myoclonus in 4, and more than one motor phenomenon in 9. The movements generally involve the head and neck and upper limbs, and lower limbs are spared. EMG recordings of motor phenomena were characteristic of spasms, and associated with normal EEG. Episodes occurred only while awake in 87 (85%), both awake and asleep in 15 (15%), and were repeated several times a day, often (44% cases) in clusters. Except for 2 sisters with BMEI, no familial cases were found. Brain imaging and metabolic studies were normal. Episodes resolved at 6-30 months of age, the majority in the 2nd year. Two cases presented clinical and EEG features of benign focal epilepsy. Language and cognitive development were normal in all cases; fifteen (16%) developed hyperkinetic behavior without learning disorders. (Caraballo RH, Capovilla G, Vigevano F, Beccaria F, Specchio N, Fejerman N. The spectrum of benign myoclonus of early infancy: Clinical and neurophysiologic features in 102 patients. **Epilepsia** June 2009;50:1176-1183). (Respond: Dr Roberto H Caraballo, Neurology Department, Hospital de Pediatria, CP 1245, Buenos Aires, Argentina. E-mail: rhcaraballo@arnet.com.ar).

COMMENT. The authors conclude that the spectrum of the syndrome of BMEI is wide, each paroxysmal motor phenomenon has a characteristic EMG pattern with normal EEG, and the prognosis is benign. The syndrome is similar to that described by Lombroso CT and Fejerman N. (**Ann Neurol** 1977;1:138-143), and should be distinguished from West syndrome. A report of shuddering attacks in a 3-year-old girl found that flexion spasms without loss consciousness were controlled by propranolol (Barron TF, Younkin DP. **Neurology** 1992;42:258-259). A family history of essential tremor is reported in patients with shuddering attacks and some patients have both shuddering and tremor (Vanasse M et al. **Neurology** 1976;26:1027-1030). These authors propose that shuddering is an early manifestation of essential tremor. Intolerance to monosodium glutamate is reported in children with shuddering attacks (Reif-Leahrer L et al. **N Engl J Med** 1975:293:1204).

POST-TRAUMATIC DANCING EPILEPSY

Researchers at Thomas Jefferson University Hospital, Philadelphia, PA, report a case of "dancing epilepsy" in a 39-year-old, right-handed man who developed refractory complex partial seizures following head trauma at 15 years of age. During video-EEG monitoring of an episode of dancing movements with unresponsiveness lasting a few minutes, the ictal EEG was poorly localized, whereas the interictal EEG showed a left anterior temporal sharp wave focus. The MRI showed left frontal and right anterior temporal encephalomalacia and gliosis. The authors comment that dancing movements are a new behavioral manifestation of epilepsy, not typical of temporal lobe epilepsy, and more suggestive of frontal lobe epilepsy. (Bagla R, Khoury JS, Skidmore C. Teaching video neuroimages: dancing epilepsy.

Neurology June 2009;72;e114). (Response and reprints: Dr John Koury, 900 Walnut St, Ste 200, Philadelphia, PA 19107. E-mail: jskhoury@gmail.com).

COMMENT. Dancing as a form of epilepsy is a complex automatism, such as running (epilepsia cursiva) or bicycling movements. Lennox WG, in his book on epilepsy (1960;page 260) refers to episodes of running, spinning round and around, in seizures following head injury.

CHANGING TRENDS IN ANTIEPILEPTIC DRUG USAGE IN GIRLS

Concerns about potential effects on offspring have prompted a gradual change in antiepileptic drug usage in girls of child-bearing age in the last decade, according to a study at the School of Pharmacy and Institute of Child Health, University of London, UK. More females aged 12-18 years are prescribed lamotrigine (LTG) than carbamazepine or sodium valproate, and the 10-fold increase in LTG in females is significantly greater than the 5-fold rise for males. (Ackers R, Besag FMC, Wade A, Murray ML, Wong ICK. **Arch Dis Child** 2009;94:443-447) (Respond: Ian Wong. E-mail: ian.wong@pharmacy.ac.uk).

MOVEMENT DISORDERS

CLINICAL AND GENETIC ANALYSIS OF MYOCLONUS-DYSTONIA

Eighty-six myoclonus-dystonia (M-D) index patients from the Dutch national referral center underwent clinical and genetic evaluation in a study at University of Amsterdam, and other centers in the Netherlands and Belgium. Age of onset was 1 – 18 years in 48 (56%) and during adulthood in the remainder. Based on clinical examination, 24 cases were classified as definite M-D, 23 were probable, and 39 possible cases. According to previously published criteria, definite M-D had early onset and a positive family history. In the definite group, 50% carried an SGCE mutation; in the probable group, 4%; and in the possible cases, none had the mutation. (Ritz K, Gerrits MCF, Foncke EMJ, et al. Myoclonus-dystonia: clinical and genetic evaluation of a large cohort. **J Neurol Neurosurg Psychiatry** June 2009;80:653-658). (Respond: Dr MAJ Tijssen, Department of Neurology, Academic Medical Centre, University of Amsterdam, PO Box 22660, 1100 DD Amsterdam, The Netherlands. E-mail: m.a.tijssen@amc.uva.nl).

COMMENT. Myoclonus-dystonia is a genetically heterogeneous movement disorder with autosomal dominant inheritance. The clinical manifestations are myoclonus and dystonia predominantly in the upper body, and in adults may respond to alcohol. A mild dystonia often presents as cervical dystonia or writer's cramp; the myoclonus is rhythmic or arrhythmic, bilateral, asymmetric, involving mainly the proximal arms and axial muscles. The major gene locus maps to the epsilon-sarcoglycan gene (SGCE, DYT11) on chromosome 7q21-22. Various SGCE mutations are reported in several families and sporadic cases. In 50% cases of M-D, no mutation is identified.