MOVEMENT DISORDERS

GABAPENTIN THERAPY IN HEMICHOREA DUE TO STROKE

A 13-year-old girl with subacute onset hemichorea of presumed ischemic origin was benefited by gabapentin therapy at the University of Massachusetts Memorial Health Care, Worcester, MA. She had presented with left sided jerky movements, worsening over 2 months, and preceded by a mitral valvuloplasty. Mitral valve prolapse was diagnosed at 4 years, and she developed left upper extremity weakness and numbness at age 5. She had also suffered from migraine headaches. Neurologic examination revealed choreiform, athetoid, and ballistic movements of the left side, and dysarthria. MRI showed a small, old ischemic infarct in the right posterior parietal and subcortical region. Following gabapentin, 300 mg 3 x daily gradually increased to 1600 mg 3 x daily (75 mg/kg/d), involuntary movements almost completely disappeared, and attention and school performance improved. When gabapentin was discontinued after 6 months and during a 2-month school break, movements recurred, especially with lack of sleep and when fatigued. (Kothare SV, Pollack P, Kulberg AG, Ravin PD. Gabapentin treatment in a child with delayed-onset hemichorea/hemiballismus. Pediatr Neurol Jan 2000;22:68-71). (Respond: Dr Paula D Ravin, Department of Neurology, University of Massachusetts Memorial Health Care, 55 Lake Ave North, Worcester, MA 01655).

COMMENT. Cardiac embolism resulting from infective endocarditis and mitral valve prolapse was the most likely cause of the subacute onset ischemic stroke in this child who presented some 8 years later with hemichorea. Gabapentin is effective in the treatment of some choreiform movement disorders caused by a static ischemic lesion. Like most antiepileptic drugs, it may rarely induce involuntary movements.

Gabapentin as possible cause of chorea. In direct contrast to the above report, choreoathetotic movements occurred as a possible adverse effect of gabapentin in a 37-year-old man with mental retardation and intractable epilepsy. Choreoathetosis and orofacial dyskinesia developed within 5 days of introducing gabapentin with AED polytherapy for seizures. Movements resolved within 2 days of discontinuing gabapentin. (Buetefisch CM et al. Neurology 1996;46:851-852). See Progress in Pediatric Neurology III, PNB Publ, 1997;p157, for abstract and commentary.

MUSCLE DISEASES

CONGENITAL MUSCULAR DYSTROPHY / RIGID SPINE SYNDROME

A detailed phenotypic description of 4 siblings (3 boys and 1 girl) with congenital muscular dystrophy and rigid spine syndrome is reported from the University of Utah, Salt Lake City, UT. They were offspring of a nonconsanguineous marraige and Northern European-American heritage. All 4 had hypotonia and neck weakness in infancy, early spinal rigidity, and early scoliosis, followed by skeletal deformities and respiratory insufficiency, the 3 oldest requiring noninvasive supportive ventilation. Muscle strength stabilized or slowly declined after initial improvement. Muscle biopsy at 9 months in one child revealed nonspecific myopathic changes, and his sibling at age 14 years had chronic and severe dystrophic changes, with normal dystrophin-glycoprotein staining. MRIs of the thigh muscles had shown selective muscle involvement early and widespread muscle abnormalities in later stages. Genetic studies showed linkage to the chromosome 1p rigid spine locus (RSMD1) at 3 cM. (Flanigan KM,