CSF WBC count (21 cells per mm3), 48 hours of antibiotics may be warranted, followed by in-hospital observation or outpatient clinic visit at 48-72 hours after discharge.

CLINICAL DIAGNOSIS OF SYDENHAM'S CHOREA IN AREA ENDEMIC FOR ACUTE RHEUMATIC FEVER

A retrospective chart review to determine the causes of childhood chorea was conducted in the Division of Allergy, Immunology and Infectious Disease at the Children's Hospital of Pittsburgh, PA. Of 144 patients with a diagnosis of chorea between 1980 and 2004, 82 had new-onset chorea; 79 (96%) were Sydenham's chorea (SC), 1 a postoperative cerebral ischemia, and 2 had basal ganglia infarcts. SC patients were female in 71%, the mean age at presentation was 9.8 years (range 5-14 yrs), chorea was unilateral in 30%, and 30% had a family history of acute rheumatic fever (ARF). Symptoms included dysarthria (67%), abnormal gait (51%), behavior change (46%), dysgraphia (29%), and headache (11%). Carditis was present in 44%, arthritis (11%), and erythema marginatum (3%). An elevated antistreptolysin O titer was documented in 99% of 53 tested, anti-deoxyribonuclease B titer in 7, positive streptozyme in 53, and acute throat infection with Streptococcus pyogenes in 19 tested. In patients with ARF and SC, brain MRI was abnormal in 8 of 32, and CT in 1 of 20 patients tested; abnormalities were not considered diagnostic of chorea and included petrous bone anomalies, medulloblastoma and macrocephaly, punctate lesions in parietal and frontal lobes, increased signal in right globus pallidus, and Arnold Chiari I. The 3 patients with new-onset chorea without ARF had MRI evidence of basal ganglia ischemic lesions; histories were considered atypical for SC. Neuroimaging should be reserved for patients with atypical presentation, including hemichorea. (Zomorrodi A, Wald ER. Sydenham's chorea in Western Pennsylvania. Pediatrics April 2006;117:675-679). (Respond: Ellen R Wald MD, Division of Allergy, Immunology and Infectious Diseases, Children's Hospital of Pittsburgh, 3705 Fifth Ave, Pittsburgh, PA 15213).

COMMENT. Although the majority of cases of new-onset chorea in childhood are related to streptococcal infection and are rheumatic in origin, the diagnosis based only on clinical findings is not invariably correct. Typically, Sydenham's chorea (SC) has an acute or gradual onset, and is a self-limiting disease. In patients with a protracted course, pathology other than ARF should be considered, and a complete neurologic evaluation is essential, including an MRI. A 6-year-old boy who developed hemichorea after scarlet fever and had been treated for a year as SC was subsequently referred and diagnosed with a Grade 3 astrocytoma of the right thalamus (Millichap et al. JAMA 1962;179:589-593). Of 300 childhood intracranial tumors treated at the Mayo Clinic 1950-1960, 4% involved the basal ganglia and less than 1% presented with involuntary movements. The differential diagnosis also includes tics, chorea secondary to perinatal anoxia or infarction, drugs, metabolic disorders, paroxysmal choreoathetosis, familial benign choreoathetosis, and Huntington's chorea. Methlphenidate-induced chorea is reported during convalescence (Nausieda PA et al. Neurology 1983;33:750).

EEG abnormalities in SC are reported in 55%-87% cases (Johnson DA et al. Arch Neurol 1964;10:21-27). Although not specific for SC, paroxysmal bursts of 3-5 Hz/sec and 2-3/sec slow waves in the posterior regions following eye closure are characteristic and may be helpful in diagnosis.