enterovirus 71 infection. **N Engl J Med** March 22, 2007;356:1226-1234). (Reprints: Dr Chang, Department of Pediatrics, National Taiwan University Hospital, College of Medicine, National Taiwan University, No 7, Chung-Shan South Rd, Taipei, Taiwan).

COMMENT. Enterovirus, an RNA nonpoliovirus, causes significant and frequent illnesses in infants and children. Clinical manifestations of EV17 are protean and include hand-foot-and mouth disease, brainstem encephalitis and polio-like paralysis. Isolation of the virus in cell culture is the standard diagnostic method, and stool and throat specimens produce the highest yield (AAP Redbook, 27th ed, 2006). The above report shows that neurologic sequelae are frequent, especially in patients with cardiopulmonary failure. Behavior and learning problems present later on attending school, and 13% are diagnosed and treated for ADHD.

OPSOCLONUS-MYOCLONUS AND STREPTOCOCCAL INFECTION

A 9-year-old Nepalese boy living in the UK presented with opsoclonus-myoclonus syndrome associated with group A streptococcal infection, and is reported from St Mary's Hospital, London; and University of Southampton, UK. The onset was acute with headache, random eye movements, vomiting, dizziness, photophobia, and jerking of all four limbs. On examination, he had rapid, chaotic eye movements, myoclonus affecting limbs and head, and ataxia. He was empirically treated with ceftriaxone, acyclovir and azithromycin. Neuroblastoma was excluded. Bacterial cultures of blood, urine, and throat swab were negative. CSF contained 18 lymphocytes/ml and 0 neurophils, 0.26 g/L protein, and normal glucose and lactate. CSF culture, pcr, and viral antibodies were negative. Serum was negative for viral antibodies. Antistreptolysin 0 was 640 units/ml initially and 1600 units/ml at 4 months follow-up. Anti-DNase B was 2880 units/ml on day 1, 1920 u/ml on day 2, and 360 u/ml on day 22. After 2 weeks his eye movements, myoclonus and ataxia had improved. After 4 months he had recovered completely without sequelae. (Jones, CE, Smyth DPL, Faust SN. Opsoclonus-myoclonus syndrome associated with group A streptococcal infection. **Pediatr Infect Dis J** April 2007;26:358-359). (Respond: Dr Saul N Faust, Wellcome Trust Clinical Research Facility, University of Southampton, Mailpoint 218, C Level, West Wing, Southampton General Hospital, Tremona Rd, Southampton, SO16 6YD, UK).

COMMENT. Chorea, tics, and obsessive compulsive disorder (PANDAS) have been associated with streptococcal infections. Dyskinesias and associated psychiatric disorders following streptococcal infections are reported in 40 patients in the UK, and opsoclonus or myoclonus was present in 3 (Dale RC et al. **Arch Dis Child** 2004;89:604-610). Opsoclonus-myoclonus is parainfectious or a paraneoplastic disorder complicating neuroblastoma.

NEUROLOGIC FINDINGS IN SYDENHAM'S CHOREA

The relationship between cardiac and neurologic findings and long-term prognosis of 40 patients with Sydenham chorea were investigated at Istanbul University, Turkey. Patients were predominantly female (70%), and mean age was 11.3 +/- 2.5 years (range 4-16 yrs). Of 304 patients with rheumatic fever, 45 (14.8%) had chorea during the first attack. Duration of chorea was 5.3 +/- 3.1 months (range 1-12 months). Chorea was mild in 30 (75%), moderate

in 9 (22%), and severe in 1 patient (2.5%). Echocardiograms showed evidence of carditis in 28 cases (70%); the mitral valve was affected most often (35%). EEG performed in 20 patients showed nonspecific abnormalities in 2. Cranial MRI in 21 patients showed no abnormality. Benzathine penicillin prophylaxis was given to all patients, and haloperidol for chorea. At a mean of 54 +/- 18 month follow-up for 32 patients, 28 with carditis, 2 (6%) had a recurrence of chorea. (Kilie A, Unuvar E, That B et al. Neurologic and cardiac findings in children with Sydenham chorea. **Pediatr Neurol** March 2007;36:159-164). (Respond: Dr Kilie, Feyzullah Mah, Sehit Hikamet Alp Cad, Adatepe Sitesi, B-1 Blok D 19, Maltepe, 34843 Istanbul, Turkey).

COMMENT. No MRI or significant EEG abnormalties were reported in this series of patients with Sydenham's chorea. Abnormalties in the caudate nucleus have been described in 16% of relapsing cases (Faustino PC et al. **Neuroradiology** 2003;45:456-462); and the caudate, putamen, and globus pallidus were increased in volume in 24 patients compared to controls studied at the NIH (Giedd JN et al. **Neurology** 1995;45:2199-2202).

EEG in Sydenham's chorea. Several studies have found abnormalities. Of 31 patients with EEGs at the Mayo Clinic, 55% had abnormal records (Johnson DA, Klass DW, Millichap JG. **Arch Neurol** 1964;10:21-27). The most prominent abnormality in our study consisted of short trains of bisynchronous waves of 2-3 cycles/sec in posterior head regions following eye closure. The changes were not considered pathognomonic for Sydenham's chorea but may be helpful in differential diagnosis of choreiform movement disorders.

METABOLIC DISORDERS

LONG-TERM OUTCOME OF MITOCHONDRIAL DISEASES

The clinical spectrum and long-term outcome of 73 children diagnosed with mitochondrial diseases between 1985 and 2005 were investigated at the Universities of Montreal and Toronto, Canada. Phenotypic categories included neonatal-onset lactic acidosis (10%), Leigh syndrome (18%), nonspecific encephalopathy (32%), mitochondrial encephalomyopathy (19%), visceral (11%), and Leber hereditary optic neuropathy (5%). Age at onset was a median of 7 months (range, prenatal to 16 years). Presenting symptoms were neurologic in 22%, including seizures, ataxia, extrapyramidal movement disorders, muscle weakness, ptosis, and headache. Neurologic presentation was acute in 35%, with stroke-like episodes, intermittent ataxia, episodic peripheral weakness, and recurrent muscle cramps. MRI showed basal ganglia hyperintensities in 46%, cerebral atrophy in 47%, brainstem lesions in 34%, and cortical infarcts in 10%. One third of the cohort developed acute acidotic crises, usually associated with benign infectious disease and usually in infancy. At follow-up, 66 patients (90%) showed clinical signs of cerebral involvement, and 29% had visceral involvement. Molecular diagnoses were established in 81%, and a mitochondrial DNA mutation was found in 20%. Mortality was 46% at a median age of 13 months, 80% <3 years of age. Patients with first symptoms before age 6 months had a tenfold increased risk of mortality, and age at first symptoms was an independent predictor of mortality. Cardiac or visceral involvement and neurologic crises were not independent outcome factors. Of 32 patients with disease onset >5 years, 62% had a favorable outcome, with only mild