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

impairment or normal functional independence. (Debray F-G, Lambert M, Chevalier I et al. Long-term outcome and clinical spectrum of 73 pediatric patients with mitochondrial diseases. **Pediatrics** April 2007;119:722-733). (Respond: Grant A Mitchell MD, Medial Genetics Division, CHU Sainte-Justine, 3175 Cote Sainte-Catherine, Montreal, Quebec, Canada H3T 1C5).

COMMENT. A high level of suspicion for mitochondrial disease (MD) is recommended in the evaluation of patients with unexplained organ dysfunction. "Any age, any symptom, any organ" is an appropriate description of MD (Munnich A et al. 1996; cited by Debray et al). In diagnosis of MD, the above authors propose initial least-invasive techniques such as fibroblast culture, and blood DNA testing for specific etiologies. Muscle and/or liver biopsy should be deferred, pending the results of fibroblast culture and blood DNA. In a large series of MD patients published in 1995 (Jackson MJ et al. **Brain** 1995;118:339-357), the most useful confirmatory diagnostic test was histochemical analysis of muscle. Elevated plasma and CSF lactate are good indicators of MD (an especially high plasma lactate is a predictor of poor outcome), but specific etiologies require molecular diagnosis.

BRAIN TUMORS

OPTIC PATHWAY GLIOMAS IN NEUROFIBROMATOSIS 1 (NF-1)

Advances in the pathophysiology and clinical behavior of NF-1 associated optic pathway gliomas (OPG) made over the past 10 years are examined, and evidence-based recommendations for diagnosis and management are proposed by researchers from Children's Memorial Hospital, Chicago; St Thomas's Hospital, London; Children's Hospital of Philadelphia; University of Pennsylvania School of Medicine, Philadelphia; and Washington University School of Medicine, St Louis, MO, The initial diagnostic and management guidelines proposed by a task force in 1997 (Listernick R et al. **Ann Neurol** 1997;41:143-149) are extended, and unanswered questions are addressed. OPG may arise de novo or progress later in childhood or adulthood. The prevalence rate of OPG with NF-1 is estimated at 15% (between 5 and 25%). Children 6 years and younger are at greatest risk, but older children and adults with NF-1 are susceptible. Visual signs are present at the time of diagnosis in 59% of patients with OPG, including decreased visual acuity, proptosis, and nystagmus. Precocious puberty, manifested initially by accelerated growth, occurred in patients older than 6 years at diagnosis, and in 12-40% of those with chiasmal OPG. Progressive disease leading to treatment occurs in 35-52% cases, but risk factors for progression are not clearly defined. Late presentation at 10 years or older may correlate with progressive disease requiring treatment.

A synopsis of recommendations for diagnosis and management of NF-1 and OPG:

1. Annual complete eye exam for children with NF-1 younger than 8 years.
2. Eye exam every 2 years until 18 years of age, for children >8 years. No role for VEP.
3. Yearly height and weight in all NF-1 children, to detect sign of precocious puberty.
4. MRI of brain and orbits, and repeated eye exams, once abnormal eye exam recorded and OPG diagnosed, and at varying intervals (3 months or longer) thereafter.