measures fail. Fludrocortisone is the initial choice, and a beta blocker is added if attacks are unresponsive. Neither drug has been tested by randomized clinical trial. Iron therapy may be helpful in toddlers with neurocardiogenic syncope associated with iron deficiency. Atropine reduces the severity and frequency of syncope, but side effects are common. For older children with frequent syncope unresponsive to drug therapy, a cardiac pacemaker may prevent an associated bradycardia or asystole. (McLeod KA. Syncope in childhood. Arch Dis Child 2003;88:350-353). (Respond: Dr KA McLeod, Consultant in Paediatric Cardiology, Department of Cardiology, Royal Hospital for Sick Children, Yorkhill NHS Trust, Glasgow G3 85J, UK).

COMMENT. The cardiologist's classification of syncope differs from that of the neurologist, who usually reserves the term simple neurocardiogenic syncope for episodes of fainting that occur in older children and are precipitated by pain, fear, excitement, and extended periods of standing, particularly in a warm environment. (Haslem RHA. Conditions that mimic seizures. In Nelson Textbook of Pediatrics. 16th ed, Philadelphia, WB Saunders, 2000;p1830). Simple syncope results from vasovagal stimulation and is characterized by a prodrome of nausea, pallor, diaphoresis, and blurred vision. It is uncommon before 10-12 years of age, and is prevalent in adolescent females. Simple syncope is differentiated from an epileptiform seizure by the complete orientation soon after the event, and the absence of seizure discharges in the EEG during an attack, but the distinction is sometimes difficult. In toddlers, syncope is described under breath-holding spells, especially the pallid variety. Breath-holding differs from simple syncope in the age of onset, precipitation by a startle or painful injury, and the sudden interruption of breathing that precedes loss of consciousness. *Cough syncope* results from an increase in intrapleural pressure, most common in asthmatic children. A coughing spell usually during sleep is accompanied by loss of consciousness, hypotonia, vertical eye-rolling, and clonic convulsive movements of short duration.

Role of the Tilt Test. In patients with a history consistent with neurocardiogenic (vasovagal) syncope, an extensive workup that currently often includes EEG, ECG, MRI, echocardiogram, etc, is not routinely indicated. In those with recurrent syncope, the tilt table testing performed early will increase the frequency of a definitive diagnosis, and avoids the inconvenience and expense of further investigations. In a retrospective analysis of 54 consecutive patients with recurrent syncope, evaluated with or without tilt table testing, 27 patients without tilt table tests received a greater number of neurology consultations, EEGs, and CTs, but a positive diagnosis was made in only 5 (18%). In contrast, a diagnosis was made early in all of 27 patients tested by tilt table, 25 having neurocardiogenic syncope and 2 conversion reactions (Strieper MJ et al. Pediatrics 1994;93:660-662; see Progress in Pediatric Neurology III, PNB Publishers, 1997;pp464-465).

MOVEMENT DISORDERS

PATHOGENESIS OF OPSOCLONUS MYOCLONUS

The proceedings of the 2nd International Opsoclonus Myoclonus Ataxia Syndrome (OMAS) Symposium (Feb 6-8, 2003; Abingdon, Oxfordshire, UK) are summarized from

the Neurosciences Unit, Institute of Child Health, University College, London, UK. OMAS is a rare disorder, only 10 new cases reported each year in the UK. Patients present between 6 and 36 months of age. Opsoclonus and myoclonus, are prerequisite symptoms for diagnosis. Irritability and sleep disturbance are equally disturbing accompanying symptoms, and delayed motor development, and cognitive, language and behavioral disorders are frequent sequelae. The neuroanatomical localization of OMAS is undetermined because of lack of post-mortem studies, but involvement of the brainstem and cerebellum is likely. MRI is normal in the acute stage but later may show cerebellar atrophy. A neural-crest tumor (commonly neuroblastoma) is identified in 50% of cases, but is not a presenting feature. Survival at 1 year for OMAS with neuroblastoma is better than that for patients with neuroblastoma alone. The underlying disease mechanism is probably immune mediated, but no common antibody-antigen complexes have been identified. Treatment with steroids and other immunomodulatory therapies is of some benefit, and relapse is usually precipitated by infections and reduction in steroid dose. Genetic factors in etiology need further study, in addition to environmental exposure to viral infection and the relation to neoplastic disorders. (Dale RC. Childhood opsoclonus myoclonus. Lancet Neurology May 1, 2003;2:270-272). (Respond: Dr Russell C Dale, Neurosciences Unit, Institute of Child Health, University College, London, UK).

COMMENT. An autoantigen diversity in the OMA syndrome is identified in a study of the sera of 21 patients at the University of Arkansas for Medical Sciences, Little Rock, AR (Bataller L, Rosenfeld MR, Graus F, et al. Ann Neurol March 2003;53:347-353). Ten adult patients had idiopathic OMAS, 5 adults had small cell lung cancer, and 6 pediatric patients had neuroblastoma. A brainstem cDNA library was probed to isolate target neuronal antigens, and 37 clones coding for 25 proteins were isolated. Two groups of antigens emerged: 1) proteins localized in or associated to the postsynaptic density (PSD), notably the adenomatous polyposis coli, a colorectal tumor suppressor protein highly expressed in the brainstem and cerebellum, and 2) proteins with expression or function restricted to neurons, including RNA or DNA-binding proteins and zinc-finger proteins. Frequent and heterogeneous immunity to neuronal antigens occur without a specific antibody marker of OMAS. PSD is a frequent source of novel autoantigens, and several proteins of this complex are targeted by antibodies of OMAS patients.

A cross-sectional study of known paraneoplastic antibodies in 59 children referred from various centers with moderate-to-severe OMAS and relapses, including 18 with neuroblastoma, showed that all were seronegative for anti_Hu, anti-Ri, and anti-Yo, the three antibodies most often associated with OPAS in adults. The study emphasizes the distinction between childhood OMAS and that in adults. (Pranzatelli MR, et al. Pediatr Neurol Nov 2002;27:384-387). (Respond: Dr Pranzatelli, National Pediatric Myoclonus Center, SIU School of Medicine, Division of Child and Adolescent Medicine, PO Box 19658, Springfield, IL 62702). For further articles on OMAS (dancing-eye syndrome), see Progress in Pediatric Neurology III, PNB Publishers, 1997;pp332-336.