

- Identify the cause of fever;
- Consider meningitis in the differential diagnosis;
- Perform LP if the child is ill-appearing or there are clinical signs or symptoms of concern;
- LP is an option in any infant 6-12 months of age who presents with a seizure and fever and who has not received immunization against *Haemophilus influenzae* type b or *Streptococcus pneumoniae*, or when immunization status cannot be determined;
- LP is an option for children pretreated with antibiotics;
- In general, further evaluation is not usually required, specifically EEG, blood studies or neuroimaging.

(AAP subcommittee on febrile seizures. Clinical practice guideline—neurodiagnostic evaluation of the child with a simple febrile seizure. **Pediatrics** Feb 2011;127:389-394)

COMMENT. In this revised guideline, compared to that published in 1999, specific indications for LP based primarily on age are modified. LP recommended in a child who is “ill-appearing” addresses the importance of clinical acumen of the treating physician, an indication omitted in the previous guidelines for children <18 months of age. It should be emphasized that these guidelines do not apply to children with complex febrile seizures.

HEADACHE DISORDERS

ALICE IN WONDERLAND SYNDROME ASSOCIATED WITH TOPIRAMATE FOR ADOLESCENT MIGRAINE PREVENTION

Neuroscientists at the University Medical Centre Hamburg, Germany, report the case of a 17-year-old girl with migraine without aura who developed an intermittent “Alice in Wonderland Syndrome” associated with prophylactic treatment of migraine headaches with topiramate. She presented with a 7-year history of migraine and headaches occurring on 5-10 days/month. Neurological examination and MRI were unremarkable. Topiramate 50 mg each night was associated with depressive aggressive symptoms and mood swings. Also, she developed paresthesias of finger tips, toes and lips, and alopecia. With continued headaches 3-4 days/month, the dose of topiramate was increased to 75 mg/nightly. When sleep was delayed, she described intermittent nocturnal distortions of her body image: her head and one hand grew bigger, while her body and other hand shrank in size. Within 2 weeks of decreasing the dose of topiramate to 50 mg/night, these nocturnal phenomena ceased. EEG between attacks was normal. A rechallenge with 75 mg daily dose was associated with recurrence of body distortions within 2 weeks. The distortions stopped within 1 week after reduction of dose to 50 mg/day, and none was reported at 5-month follow-up. Other potential pathophysiologies, including migraine aura and complex partial seizure, were considered unlikely. (Juergens TP, Ihle K, Stork J-H, May A. “Alice in Wonderland syndrome” associated with topiramate for migraine prevention. **J Neurol Neurosurg Psychiatry** Feb 2011;82:228-

229). (Respond: Dr Tim P Juergens, University Medical Center Hamburg-Eppendorf, Martinistr. 52, Hamburg D-20246, Germany. E-mail: t.juergens@uke.de).

COMMENT. The authors cite only one other case-report of topiramate-associated body image distortion: a 31-year-old female migraine patient had taken 25 mg/daily for 1 week. Several trials, some randomized, double-blind, and placebo-controlled have demonstrated the efficacy and tolerability of topiramate in prevention of migraine in children and adolescents. The frequency of side effects varied among studies, and included dizziness, anorexia, abdominal pain, difficulty concentrating, sedation and paresthesia. (Ferraro D et al. **J Headache Pain** 2008;9(3):147-150). Overall, topiramate was safe and well tolerated. (Lewis D et al. **Pediatrics** 2009;123(3):924-934). A dose of 100 mg/day was required for an optimal beneficial effect.

NEUROMUSCULAR DISEASES

CHARCOT-MARIE-TOOTH DISEASE SUBTYPES AND GENETICS

Researchers at Wayne State University School of Medicine, Detroit, MI, identified distinguishing clinical and physiological features of subtypes of Charcot-Marie-Tooth (CMT) disease among 787 patients that could be used to direct genetic testing. A total of 527 patients with CMT (67%) received a genetic subtype, while 260 had no identifiable mutation. The most common CMT subtypes were CMT1A, CMT1X, hereditary neuropathy with pressure palsies (HNPP), CMT1B, and CMT2A. Other subtypes accounted for <1% each. Eleven patients had >1 genetically identified subtype of CMT. Genetically identified CMT patients were separable into specific groups based on age of onset and degree of slowing of motor nerve conduction velocities. A focused approach based on phenotype, physiology and prevalence is proposed for genotyping. With a genetic diagnosis made in a patient, other family members can be identified by clinical and neurophysiology evaluation, and costly genetic tests may be unnecessary. (Saporta ASD, Sottile SL, Miller LJ, Feely SME, Siskind CE, Shy ME. Charcot-Marie-Tooth disease subtypes and genetic testing strategies. **Ann Neurol** Jan 2011;69:22-33). (Respond: Dr Shy, Department of Neurology, Wayne State University School of Medicine, 421 Ea Canfield, Elliman 3217, Detroit, MI 48201. E-mail: m.shy@wayne.edu).

COMMENT. CMT affecting 1 in 2500 population is the most common inherited neurological disorder. Generally autosomal dominant in inheritance, some cases are X-linked or autosomal recessive inheritance. Demyelinating neuropathy is most common, but one-third are primary axonal disorders. (Harding AE, Thomas PK. **Brain** 1980; **J Med Genet** 1980). CMT is a heterogeneous disorder, and more than 30 genes have been identified that cause various clinical and physiological subtypes. The focused approach to diagnosis outlined by the above authors should facilitate family planning, prognosis, and treatment.

X-linked CMT disease in childhood. A retrospective review of 17 children with CMTX at children's hospitals in Melbourne and Sydney, Australia, showed that 15 were