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

symptomatic before 5 years of age. The CMT typical phenotype in some was complicated by delayed motor development, sensorineural hearing loss, tremor, and pathological fractures. Axonal loss affected all patients. An X-linked dominant inheritance and carrier females with abnormal exam correlated with a connexin 32 mutation in all but 2 pedigrees. The clinical phenotype for CMTX1 is broader than previously recorded. (Yiu EM et al. **Neurology** Feb 2011;76:461-466).

SERUM TRANSAMINASE IN DUCHENNE DYSTROPHY

Researchers at Children's Hospital Boston, MA, have shown a linear relationship between serum CPK and serum ALT and AST and a logarithmic relationship between serum enzyme levels and age for boys with Duchenne (n=46) or Becker (n=9) muscular dystrophy (DMD or BMD). A mathematical model to predict serum ALT and AST levels with known CPK and age was developed to provide reassurance that elevated transaminase may be indicative of muscle disease, avoiding unnecessary tests of liver dysfunction. Serum transaminase was highest in ambulant boys with DMD (1220 U/L ALT and 801 U/L AST). These levels were 22 and 12 times higher than upper limit nomal levels for ALT and AST, respectively. The study was prompted by the observed reluctance of clinicians to attribute high transaminase levels to muscle disease, leading to delayed diagnosis and extensive tests for liver dysfunction. (McMillan HJ, Gregas M, Darras BT, Kang PB. Serum transaminase levels in boys with Duchenne and Becker muscular dystrophy. **Pediatrics** Jan 2011;127:e132-e136). (Respond: Peter B Kang MD, Department of Neurology, Children's Hospital Boston, 300 Longwood Ave, Boston, MA 02115. E-mail: peter.kang@childrens.harvard.edu).

COMMENT. High serum transaminase levels should alert clinicians to possible muscle disease and prompt serum CPK estimation when clinically indicated. Unnecessary liver function tests and withdrawal of drug therapy may be avoided.

MRI DEFINITION OF INVOLVED MUSCLE IN DUCHENNE MUSCULAR DYSTROPHY

The degree of muscle involvement of lower leg muscles of 34 patients with DMD >8 years, using muscle MRI, was estimated in a multicenter study at the Institute of Child Health and other centers in London, in the UK, and 1 in Rome, Italy. Muscle MRI findings in a subgroup of 15 patients were correlated with the histology of open biopsies of extensor digitorum brevis (EDB). A gradient of muscle involvement in the lower leg was documented in all patients, and the posterior compartment (gastrocnemius > soleus) was most severely affected. The anterior compartment (tibialis anterior/posterior, popliteus, extensor digitorum) was least affected. Muscle MRI/EDB involvement correlated with the patient's age (p=0.055). MRI correlated with EDB histopathologic changes in 10/15 patients. Abnormal MRI grades 3-4 (range 0-4) were associated with more severe fibro-adipose tissue replacement. Muscle MRI showed a progressive involvement of the EDB, more obvious in older patients and those nonambulant for a longer time. (Kinali V, Arechavala-Gomeza V, Cirak S, et al. Muscle histology vs MRI in Duchenne muscular dystrophy. **Neurology** Jan 25, 2011;76:346-353). (Respond: Dr