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