non progressive condition of the upper limbs. J Neurol Neurosurg Psychiatry April 1993; 56: 365-368). (Respond: Dr G Hageman, Department of Neurology, Medical Spectrum Twente, Hospital Enschede, PO Box 50000, 7500 KA Enschede, The Netherlands).

COMMENT. The absence of sensory deficits and normal motor nerve conduction velocities point to involvement of the anterior horn cells. The etiology of the prenatal pathology with loss of anterior horn cells in cases of neurogenic arthrogryposis is unknown, except for one case cited of rubella exposure. The limitation to the upper limbs is unusual.

METABOLIC AND TOXIC DISORDERS

SYNDROMES OF 3-METHYLGLUTACONIC ACIDURIA

The most common clinical syndromes associated with 3-methylglutaconic (MGC) aciduria are reviewed by researchers from various centers; Courtwright and Summers Metabolic Disease Center and Baylor Research Institute, Dallas, TX; Shaare Zedek Medical Center, Jerusalem; Free University of Amsterdam; Loewenstein Hospital, Tel-Aviv Univ, Raanana, Israel; and Kennedy Krieger Institute, Baltimore, MD. Three distinct syndromes are described: *Type I 3-MG-CoA Hydratase Deficiency.*- (autosomal recessive) 3 patients had delayed speech and macrocephaly, increased urinary excretion of 3-MGC acid, 3-M Glutaric(MGR) and 3-hydroxyisovaleric acids, elevated CPK and serum carnitine, and hypoglycemia. Urinary 3-MGC excretion is decreased by restriction of L-leucine intake. *Type II Barth Syndrome.*- (X-linked) dilated cardiomyopathy, recurrent infection, neutropenia, growth retardation. Improves with age. Increased urinary excretion of 3-MGC, 3-MGR, fumaric, and 2-ethylhydracrylic acids. *Type III Costeff Optic Atrophy Syndrome.*- (autosomal recessive) A Behr-like syndrome described in 39 patients of Iraqi-Jewish origin living in Israel. Optic atrophy, choreoathetosis, spastic paraparesis, cerebellar ataxia. Nonprogressive. 3-MGC and 3-MGR aciduria. An *Unclassified 3-MGC aciduria* includes patients presenting in the first year of life with neurologic impairment, seizures, retinal pathology, and a fatal course. Cardiomyopathy, hepatic dysfunction, hypoglycemia, lactic acidosis, and dysmorphia are associated manifestations. Defects of the mitochondrial respiratory chain are suggested. (Gibson KM et al. Multiple syndromes of 3-methylglutaconic aciduria. Pediatr Neurol March/April 1993; 9: 120-123). (Respond: Dr Gibson, Baylor Research Institute, 3812 Elm Street, Dallas, TX 75226).

COMMENT. This review will assist the neurologist in the differential diagnosis of patients with 3-MGC aciduria. Some cases are multisystemic and progressive, with onset from birth to several years, some are primarily neurologic and non-progressive, while others involve the heart muscle and may improve with age.