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COMMENT. CMT1A is a demyelinating neuropathy characterized by progressive muscle weakness and atrophy. The peroneal muscles are involved first, causing a striking stork-like gait, and weakness and atrophy of the upper extremities is initially limited to the intrinsic muscles of the hands. The authors comment that the hand involvement is frequently under-recognized in the early stages.

## HEARING LOSS IN FACIOSCAPULOHUMERAL DYSTROPHY

The clinical presentation of facioscapulohumeral dystrophy (FSHD) with unusual large 4q35 deletions was studied with attention to hearing loss. Hearing function was examined by otoscopy, audiometry and auditory-evoked brainstem responses. Data obtained from 6 patients with EcoRI 4q35 fragment size, ranging from 10 to 13 kb, were compared with those of 28 similar subjects reported in the literature. Sensorineural hearing loss occurred in 4 patients who had an infantile-onset dystrophic phenotype. Hearing loss was associated with mental retardation in 3 and epilepsy in 2. Hearing was mildly impaired in the remaining 2 of 6 patients. When the data from 28 similar cases reported in the literature were combined with that from the 6 patients examined, 68% had auditory impairment. Hearing loss is a characteristic feature of FSHD patients with a large 4q35 deletion. When considering only cases with 10-11 kb fragment size, FSHD is associated with early-onset dystrophic phenotype, mental retardation in 92% and epilepsy in 58%. (Trevisan CP, Pastorello E, Tomelleri G et al. Facioscapulohumeral muscular dystrophy: hearing loss and other atypical features of patients with large 4q35 deletions. *Eur J Neurol* Dec 2008;15:1353-1358 (Abstract)).

COMMENT. Facioscapulohumeral dystrophy with sensorineural hearing loss and Coats' syndrome was described by Taylor DA et al (*Ann Neurol* 1982;12:395). Coats' syndrome includes congenital retinal dysgenesis with telangiectasia and retinal detachment. A PubMed search of the literature found 8 reports of FSHD and sensorineural deafness, dating from 2008 to 1985. One case with epilepsy was complicated by infantile spasms at 6 months of age, the dystrophy presenting at 3 years, and sensorineural deafness noted later (Akiyama C et al. *No To Hattatsu* 1991;23:395-399). All 6 patients reported with facial diplegia in the first year of life and subsequent development of FSHD had sensorineural deafness (Korf BR et al. *Ann Neurol* 1985;17:513-516).

## NEONATAL DISORDERS

### HIPPOCAMPAL VOLUMES IN PRETERM INFANTS

The relation between neonatal regional brain volumes and working memory deficits at age 2 years was investigated in 156 very preterm children born at the Royal Women's Hospital, Melbourne, Australia, prior to 30 weeks gestation or weighing <1250g. Very preterm children who perseverated on the working memory task had significantly smaller