

Institute for Neuromuscular Research, The Children's Hospital at Westmead, Locked Bag 4001, Westmead, NSW 2145, Australia. E-mail: joshuab2@chw.edu.au.

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

hippocampal volumes than controls with intact working memory. (Beauchamp MH, Thompson DK, Howard K, et al. Preterm infant hippocampal volumes correlate with later working memory deficits. *Brain* Nov 2008;131:2986-2994). (Respond: Dr Peter J Anderson, School of Behavioural Science, The University of Melbourne, Melbourne, VIC 3010, Australia. E-mail: peterja@unimelb.edu.au).

COMMENT. Children born preterm have smaller hippocampal volumes that correlate with working memory deficits measured at 2 years of age. Further research will determine the longterm effect of altered hippocampal volumes on cognitive function of premature infants.

NEUROGLIAL HETEROTOPIA AND NASOPHARYNGEAL OBSTRUCTION IN A NEONATE

The clinical presentation, imaging, treatment, and pathology of a case of neuroglial heterotopia in the nasopharynx causing airway obstruction in a newborn are reported from Columbus Children's Hospital, OH. MRI and CT showed a cystic mass filling the nasopharynx with a midline bony defect in the sphenoid bone above the clivus. Posterior nasal endoscopy visualized the cystic lesion prior to surgical removal. Connection with CSF and subarachnoid space was excluded. At 6-month follow-up, developmental milestones were normal, and repeat CT showed no evidence of recurrence of the mass. Histopathology of the lesion showed choroid plexus, glial, and respiratory-like epithelial cells. (Husein OF, Collins M, Kang DR. Neuroglial heterotopia causing neonatal airway obstruction: presentation, management, and literature review. *Eur J Pediatr* Dec 2008;167:1351-1355). (Respond: OF Husein, 1414 N Houk Rd, Suite 208, Spokane, WA 99216. E-mail: tiffhusein@yahoo.com).

COMMENT. Reviewing the literature, the authors found reports of 30 cases of pharyngeal neuroglial heterotopia. Both CT and MRI are recommended in the assessment of nasopharyngeal masses. CT visualizes any bony deformities of the skull base, and MRI detects intracranial connections through the skull defect. Encephalocele has a similar histology but differs from neuroglial heterotopia by maintaining a connection to the subarachnoid space.

ATTENTION DEFICIT DISORDERS

NEUROANATOMICAL ABNORMALITIES IN ADOLESCENT ADHD

Twenty-four adolescents with familial ADHD and 10 control youths underwent high-resolution structural MRI, and frontal lobe gyri and caudate were compared in a study at Stanford University and other US centers. Youths with ADHD had larger right caudate and right inferior frontal lobe volumes than control subjects. An increase in left caudate volume in a subgroup of ADHD youths was correlated with decreasing functional activation in this region. The findings were thought to reflect neurodevelopmental changes specific to late adolescence in familial ADHD. (Garrett A, Penniman L, Epstein JN et al. Neuroanatomical abnormalities in adolescents with attention-deficit/hyperactivity disorder. *J Am Acad Child*