

is withheld. A test for early recognition of varicella-zoster virus infection could improve prognosis.

HOPKINS SYNDROME WITH MYCOPLASMA INFECTION

A 3-year-old boy with acute flaccid paralysis of the right lower limb developing one week after treatment and recovery from an acute asthma attack is reported from the Department of Child Health, Milton Keynes General Hospital, UK. Sensation was normal, plantar responses were flexor, deep tendon reflexes were absent in the involved limb, and meningeal signs were negative. MRI of the spine was normal. EMG was consistent with anterior horn cell damage and persisting amyotrophy. Mycoplasma complement fixation test titer and agglutination titer were significantly increased. Erythromycin treatment was without benefit, and paralysis persisted at 12 month follow up. (Acharya AB, Lakhani PK. Hopkins syndrome associated with Mycoplasma infection. *Pediatr Neurol* Jan 1997;16:54-55). (Response: Dr Lakhani, Department of Child Health, Milton Keynes General Hospital, Eaglestone, Milton Keynes MK6 5LD, UK).

COMMENT. Since the first description of 10 cases of a poliomyelitis-like illness associated with acute asthma in childhood (Hopkins IJ, 1974), 18 additional reports of Hopkins syndrome have been published. Non-polio enteroviruses, varicella and herpes virus type 1 have been implicated in some cases. Mycoplasma is known to exacerbate asthma and has been linked to various neurological disorders, including Guillain-Barre syndrome and transverse myelitis. Amyotrophy and Hopkins syndrome are additional complications of Mycoplasma infection.

CONGENITAL DEVELOPMENTAL DISORDERS

CONGENITAL FIBROSIS OF EXTRAOCULAR MUSCLES

The intracranial and orbital pathology and muscle pathology of chromosome 12-linked congenital fibrosis of the extraocular muscles are described in 3 affected members of a family in a report from Children's Hospital, Harvard Medical School, Boston, and other centers. Postmortem examination of 1 and muscle biopsies of 2 patients showed abnormalities in the brain stem, cranial nerves, and extraocular muscles (EOMs): absence of motor neurons of the oculomotor nucleus, loss of axons in III cranial nerve, absence of the superior division of III CN, and atrophic superior rectus and levator palpebrae muscles, showing only a clump of myofibers, connective tissue, and fat. Increased numbers of internal nuclei and central mitochondrial clumping found in other extraocular muscles pointed to an extension of the process beyond the superior division of III CN, and an abnormal development of the EOM lower motor neuron system. (Engle EC, Goumnerov BC, McKeown CA et al. Oculomotor nerve and muscle abnormalities in congenital fibrosis of the extraocular muscles. *Ann Neurol* March 1997;41:314-325). (Respond: Dr Engle, Division of Genetics, Enders 512, Children's Hospital, 300 Longwood Avenue, Boston, MA 02115).

COMMENT. Congenital fibrosis of extraocular muscles, an autosomal dominant inherited disorder, is characterized by bilateral ptosis and fixation of eyes in a downward and strabismic position. It resembles Brown's syndrome (vertical retraction), and Duane's syndrome (horizontal retraction), associated with fibrosis and aberrant innervation of the rectus lateralis. The pathology

of these congenital EOM fibrosis syndromes is more diffuse than previously recognized and extends beyond the extraocular muscles and involves also the cranial nerves and brain stem.

CSF CIRCULATION AND CONGENITAL HYDROCEPHALUS

A new model of the cerebrospinal fluid (CSF) circulation, involving brain capillaries as the main site of CSF absorption, is proposed from the Karolinska MR Research Center and Department of Neuroradiology, Karolinska Hospital, Stockholm, Sweden. In communicating hydrocephalus, a decrease of CSF flow through the foramen magnum demonstrated by MRI is explained by restricted expansion of the intracranial arteries. In the new classification, communicating hydrocephalus is called "restricted arterial pulsation (RAP) hydrocephalus," a primary hemodynamic disturbance, caused by any process that decreases compliance of arteries, eg ectasia, arteritis, spasm. In obstructive hydrocephalus, ventricular dilatation causes compression of cortical veins, a secondary hemodynamic disturbance called "venous congestion (VC) hydrocephalus." Pharmacological treatment for hydrocephalus should consist of selective venous constrictor drugs such as dihydroergotamine. (Greitz D, Greitz T, Hindmarsh T. A new view on the CSF-circulation with the potential for pharmacological treatment of childhood hydrocephalus. *Acta Paediatr* Feb 1997;86:125-132). (Respond: Dr D Greitz, Karolinska MR Research Center, Karolinska Hospital, S-171 76, Stockholm, Sweden).

COMMENT. The authors caution that treatment of hydrocephalus with dihydroergotamine should not be employed in children until more studies are completed in adults. The new hemodynamic mechanism for hydrocephalus is more in keeping with studies involving flow sensitive MR imaging and radionuclide cisternography. As noted by Whitelaw A, in an editorial commentary, one of the strongest arguments against the conventional model of CSF circulation and absorption is the absence of Pacchionian granulations in children less than 12 months of age. However, the new hemodynamic concept is only a hypothesis which needs to be proven by further studies.

MOVEMENT DISORDERS

DYSTONIA AND WRITER'S CRAMP TRIGGERED BY EXERCISE

An 11-year-old girl with a gait disturbance and foot dystonia, especially after exertion at the end of the day, and handwriting difficulty with cramps after a short school exercise, was evaluated at the Neuropaediatric Unit, CHUV, Lausanne, Switzerland. Her gait was better in the morning and after rest, but when lying down, her feet were inverted and plantar flexed in a dystonic posture. Treatment with L-dopa, 100 mg, and benzerazide 25 mg, 2 to 3 times a day, relieved all symptoms and signs, but dystonia returned when L-dopa was discontinued. The mother had parkinsonism relieved by L-dopa. (Deonna T, Roulet E, Ghika J, Zesiger P. Dopa-responsive childhood dystonia: a forme fruste with writer's cramp, triggered by exercise. *Dev Med Child Neurol* Jan 1997;39:49-53). (Respond: Dr Thierry-W Deonna, Neuropaediatric Unit, CHUV, 1011 Lausanne, Switzerland).

COMMENT. Children presenting with dysgraphia, fatigue, and cramps should be examined for possible dopa responsive dystonia, especially if the symptoms are associated with a gait disturbance at the end of the school day.