
**VASCULAR DISORDERS**

**IRON DEFICIENCY AND STROKE**

A series of six iron deficient children, 6 to 18 months of age, presenting with an ischemic stroke or venous thrombosis after a viral syndrome, are reported from the University of Saskatchewan, and the Children's Hospital of Eastern Ontario, Canada. Other known etiologies for stroke were excluded. Iron deficiency, in association with a nonspecific viral illness, was a contributing factor and not a coincidental finding. (Hartfield DS, Lowry NJ, Keene DL, Yager JY. Iron deficiency: a cause of stroke in infants and children. Pediatr Neurol Jan 1997;16:50-53). (Respond: Dr Yager, Room 3717, F Wing, Department of Pediatrics, Royal University Hospital, 103 Hospital Drive, Saskatoon, Saskatchewan, Canada S7N 0W8).

**COMMENT.** Iron deficiency anemia in infancy and early childhood can be a contributing factor in the etiology of stroke. Iron deficiency has also been related to other neurological illness, including breath holding spells, headache, pseudotumor, diplopia, and cranial nerve palsies. (see Progress in Pediatric Neurology I. PNBPubl, Chicago, 1991;pp397-398).

**INFECTION DISORDERS**

**TREATMENT OF RAMSAY HUNT SYNDROME**

The effect of acyclovir-prednisone treatment in 80 patients with Ramsay Hunt syndrome was analyzed retrospectively at the Department of Otolaryngology, Ehime University School of Medicine, Ehime, Japan. Ages ranged from 15 to 75 years. All presented with facial paralysis and herpetic eruption on the pinna or oral mucosa, and 22 had associated hearing loss. Treatment with acyclovir, 250 mg tid by intravenous drip or 800 mg 5x daily orally, and prednisone, 1 mg/kg day iv or orally, was started 1 to 10 days after onset of facial paralysis. Recovery from paralysis was complete in 21 (75%) of 28 patients treated within 3 days of onset, and in only 7 (30%) of 23 whose treatment was delayed more than 7 days. Nerve excitability testing showed that nerve degeneration was reduced by early administration of acyclovir-prednisone therapy. Recovery of hearing was better in patients treated early. In facial nerve recovery and outcome, intravenous administration of acyclovir was not superior to oral treatment. (Murakami S, Hato N, Horiuchi J et al. Treatment of Ramsay Hunt syndrome with acyclovir-prednisone: significance of early diagnosis and treatment. Ann Neurol March 1997;41:353-357). (Respond: Dr Murakami, Department of Otolaryngology, Ehime University School of Medicine, Shigenobu-cho, Onsen-gun, Ehime 791-02, Japan).

**COMMENT.** Ramsay Hunt syndrome manifested by facial pain and nerve paralysis, herpetic eruption on the pinna, and frequent vestibulocochlear involvement, is caused by varicella-zoster virus infection. Early administration of acyclovir and prednisone, within the first 3 days of onset, results in 75% rate of facial nerve recovery and less likelihood of residual nerve deafness. Absence of herpetic eruption in 8 to 25% of cases leads to misdiagnosis as Bell's palsy and poorer prognosis because acyclovir treatment
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