VASCULAR DISORDERS

VASCULAR MALFORMATIONS AND INTRACTABLE EPILEPSY

A retrospective study of 20 consecutive patients with cerebral vascular malformations who were treated surgically for medically refractory partial epilepsy is reported from the Departments of Neurology and Neurologic Surgery, Mayo Clinic, Rochester, MN. MRI was more sensitive than CT and showed 36 vascular malformations (32 cavernous and 4 A-V malformations). Previous hemorrhage was verified in 18 patients. After complete resection of the lesion, 15 patients were free of seizures and 3 had a 90% control. Age of onset and duration of seizures did not affect outcome. A focal corticectomy in addition to lesionectomy was necessary in 11 patients with MRI-disclosed ipsilateral medial temporal lobe atrophy. (Dodick DW, Cascino GD, Meyer FB. Vascular malformations and intractable epilepsy: Outcome after surgical treatment. Mayo Clin Proc Aug 1994;69:741-745). (Reprints: Dr GD Cascino, Department of Neurology, Mayo Clinic, 200 First Street SW, Rochester, MN 55905).

COMMENT. The authors advocate early surgical intervention and excision of the lesion in patients with refractory partial seizures associated with vascular malformations. Corticectomy may be necessary in patients with dual temporal lobe pathologies.

SURGERY OF MOYAMOYA DISEASE

The manifestations of moyamoya disease in children and adults and the results of various surgical procedures are reviewed from the Department of Neurological Surgery and Section of Child and Adolescent Neurology, Mayo Clinic, Rochester, MN. Onset is in the first or the fourth decades of life. Of 518 patients registered in Japan, 155 were children <15 years, and 234 were adolescents or adults. In children, recurrent ischemic attacks (in 39%), strokes (in 39%), and seizures (14%) were the most common features, whereas in adults, hemorrhage (65%), and strokes (18%) were most frequent. Mortality was 7.5% for the total series; 10% for adults and 4.3% for children. Intracranial bleeding was the cause of death in 5 of 9 children (55%) and in 19 of 30 adults (63%). Of 27 children with only TIAs who were untreated, the TIAs gradually resolved but more than one-half showed cognitive impairment at 5 to 10 year follow-up. Ischemic symptoms diminished in 10 Mayo patients treated surgically at 13.5 years (mean age), using STA-MCA anastomosis (5), EDAS (2), and EDAS/EMS (3). Most published reports find surgical intervention of benefit in children but not in adults. (Ueki K, Meyer FB, Mellinger JF. Moyamoya disease: The disorder and surgical treatment. Mayo Clin Proc Aug 1994;69:749-757). (Reprints: Dr FB Meyer, Department of Neurologic Surgery, Mayo Clinic, 200 First Street SW, Rochester, MN 55905).

COMMENT. The natural history of moyamoya disease is characterized by neurologic deterioration, strokes and hemorrhage, seizures, and mental deterioration. Current evidence favors surgery to revascularize ischemic brain tissue by collateral pathways, especially in children with ischemic symptoms.

An 11 year-old girl with acute unilateral chorea as the presenting manifestation of moyamoya disease is reported from Hospital Plaza de Cruces, Vizcaya, Bilbao, Spain (Garaizar C, Prats JM et al. Acta

Neuropediatr June 1994;1:59-64). Chorea resolved after 15 days haloperidol therapy, and no further ischemic episodes had occurred in 16 months during treatment with nicardipine. The authors cite two previous reports of moyamoya and chorea.

MYASTHENIA GRAVIS

JUVENILE MYASTHENIA AND PUBERTY

The influence of race, sex, and puberty on incidence, severity, and outcome of juvenile myasthenia gravis beginning before age 20 years was evaluated in 115 patients seen at the University of Virginia, Duke University, and University of North Carolina at Chapel Hill. White patients with prepubertal disease onset had an equal sex ratio, and female predominance increased during and after puberty. Males had less severe disease than females. Black patients showed a constant F:M ratio of 2:1 in all pubertal-onset groups. Spontaneous remissions only occurred in white patients with prepubertal onset; and persistent symptoms for more than 10 years were least frequent in this group. Early thymectomy in white patients was followed by more remissions and milder symptoms than late thymectomy. Black patients had infrequent remissions, and similar disease severity after early or late thymectomy. (Andrews PI et al. Race, sex, and puberty influence onset, severity, and outcome in juvenile myasthenia gravis. Neurology July 1994;44:1208-1214). (Reprints: Dr P Ian Andrews, Division of Pediatric Neurology, Box 3533, Duke University Medical Center, Durham, NC 27710).

COMMENT. This study documents the importance of race, sex, and puberty on the incidence, severity, response to thymectomy, and outcome in juvenile myasthenia gravis. Thymectomy was most effective in white patients when performed within 1 year of peripubertal disease onset. See Progress in Pediatric Neurology II, Chicago, PNB Publ, August 1994, for further reports of juvenile myasthenia gravis from the University of Iowa, a multicenter study in Italy, and from the Mass General Hospital, Boston.

TOXIC DISORDERS

LEAD EXPOSURE: INAPPROPRIATE SCREENING PRACTICES?

Physician screening practices at a hospital-based, university-affiliated pediatric primary care center serving an urban high-risk population in Rochester, NY were evaluated to determine the feasibility of the 1991 Centers for Disease Control guidelines. Among 632 children aged 9 to 25 months who attended the center between 1989 and 1991, screening was deficient in 55%, 34%, and 29% at ages 9-13 months, 14-19 months, and 20-25 months, respectively. Many high-risk children living in houses built before 1950, including those making well-child visits, were not appropriately screened for lead toxic effects, and opportunities for testing were frequently missed. (Campbell JR, McConnochie KM, Weitzman M. Lead screening among high-risk urban children. Are the 1991 Centers for Disease Control and Prevention Guidelines feasible? Arch Pediatr Adolesc Med July 1994;148:688-693). (Reprints: Dr Campbell, Department of Pediatrics, Rochester General Hospital, 1425 Portland Ave, Rochester, NY 14621).