

# PEDIATRIC NEUROLOGY BRIEFS

A MONTHLY JOURNAL REVIEW SERVICE

J. GORDON MILLICHAP, M.D., F.R.C.P., EDITOR

---

Vol 1, No 2

July 1987

---

## CEREBROVASCULAR DISEASE

### SURGICAL TREATMENT OF CHILDHOOD MOYAMOYA DISEASE

Neurosurgeons at the Hospital for Sick Children and Western Hospitals, Toronto, Canada, review their experiences during a 6 month to 15 year follow up of 25 children with moyamoya disease 15 of whom underwent revascularization procedures. Five patients had superficial temporal artery-middle cerebral artery (STA-MCA) bypass procedures and 13 underwent encephalo-duro-arterio-synangiosis (EDAS), a procedure expected to induce collateral branches to sprout from the STA.

Of the untreated cases, 75% developed permanent neurological sequelae, including seizures, hemiparesis and mental retardation, and 2 died. Among the 13 treated cases, 10 were neurologically intact and the remainder were improved. The STA-MCA was the procedure of choice in older children whereas EDAS, including opening of the arachnoid, a simpler technique, gave good results and was preferred in infants and young children. The authors suggest that all pediatric patients be offered the benefit of early surgical treatment before permanent neurological deficits have developed. (Olds MV, Griebel RW, Hoffman HJ et al. J Neurosurg 1987; 66:675-680).

**COMMENT:** Moyamoya, a Japanese term meaning hazy or misty like a "puff of smoke", describes the angiographic appearance of the net-like collateral circulation that develops at the base of the brain as a result of stenosis of the distal internal carotid artery and its branches. Moyamoya disease has been classified in two forms: (1) a "primary" idiopathic nonprogressive form with alternating hemiplegia, predominantly in girls of Japanese origin, often familial and representing a hereditary malformation of the elastic lamina of the cerebral vasculature, and (2) an "acquired" usually progressive form in both children and adults with varying underlying diseases. The diseases associated with moyamoya have included neurofibromatosis, basal meningitis, hypertension, atherosclerosis, myopathy, sickle-cell anemia, Fanconi's anemia, type 1 glycogenosis, congenital heart disease, Down's syndrome, and conditions requiring radiation therapy. The postradiation pathogenesis was reported by Drs. Rajakulasingam, Cerullo, and Raimondi at Northwestern UMS (Child's Brain 1979;5:467),

---

**PEDIATRIC NEUROLOGY BRIEFS** c 1987 covers selected articles from the world literature and is published monthly. Subscription requests (£15 UK or \$25 US) may be sent to: **PNB 1987/88 - J. Gordon Millichap MD FRCP - Editor**, P.O. Box 1605, London W8 7JD, UK, or P.O. Box 11391, Chicago, Ill 60611, USA.

The Editor is Professor of Neurology and Pediatrics at Northwestern University Medical School, Chicago, USA. He is presently Visiting Professor at the Hospital for Sick Children, Great Ormond Street, London WC1, England.

one case occurred in the present series, and a patient of mine, a girl aged 16, developed the syndrome following irradiation of the neck for Hodgkin's disease.

The present authors point out that the early concepts of moyamoya disease as a relatively benign disorder in Japanese girls have been modified in recent reports. The sexes were equally affected and the majority of patients were Caucasian in their series. The clinical course depends on the rapidity and extent of vascular occlusion and the ability to develop a collateral circulation. The progressive nature and serious sequelae of cases left untreated are stressed.

#### ARTERIOVENOUS MALFORMATIONS OF THE BRAIN

Twenty-three children with A-V malformations have been treated by neurosurgeons at the Children's Hospital of Eastern Ontario, Ottawa, Ontario. Fourteen were boys and 9 girls. The average age at presentation was 10 years. The majority (83%) presented with spontaneous hemorrhage and only one (4.3%) with seizures. Angiography was performed in 21 patients. The AVM could not be demonstrated in 5 (24%) who had an occult or cryptic AVM. Contrast-enhanced CT also failed to show abnormal vessels in 3 of the 5 occult AVMs.

Of 18 survivors, 15 were normal and 3 slightly disabled (2 with epilepsy). Aggressive surgical intervention resulted in improved survival and low morbidity. Overall mortality in the group was 22% while complete surgical excision carried a 7% mortality. The authors conclude that a spontaneous cerebral hemorrhage in a child is probably due to a vascular malformation, even when angiography and enhanced CT are negative. CT contrast enhancement is not a reliable indicator of occult AVM and direct surgery is needed for diagnostic confirmation and prevention of further hemorrhages. Also, children presenting with symptoms other than hemorrhage (e.g. seizures), should undergo surgery to prevent bleeding, providing the lesion is accessible with low risk to healthy brain. (Ventureyra ECG, Herder S. Child's Nerv Syst 1987;3:12-18).

COMMENT: AVMs that bleed usually require neurosurgical management, and total excision seems the treatment preferred when feasible. Stereotaxic radiosurgery and the proton beam are available in some centers for cases not amenable to excision. The treatment of the AVM presenting with seizures or recurrent headaches but without spontaneous hemorrhage is often a neurologist's responsibility. When should he involve his neurosurgical colleague? In the past, neurologists have sometimes chosen conservative management. The present study argues in favor of surgery for all AVMs in children diagnosed radiologically, given accessibility and low risk to surrounding brain tissue. Even the neonate with the AVM that involves the vein of Galen and presents with congestive heart failure has a better chance of survival with surgery. (see HOFFMAN et al J Neurosurg 1982;57:316).

#### CNS NEOPLASMS

##### PINEAL REGION TUMOURS OF CHILDHOOD

At the University Hospital, Hamburg, from 1980-85, 17 children had pineal region tumours among 102 children with CNS tumours. The authors conclude: (1) that the incidence in Germany is higher than assumed and at least equal to that in Japan, (2) radiation without