

PEDIATRIC NEUROLOGY BRIEFS

A MONTHLY JOURNAL REVIEW

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Vol. 20, No. 3

March 2006

VASCULAR DISORDERS

PRESENTATION AND OUTCOME OF CRANIAL ANEURYSMS

The presentation, location, types, and long-term post-surgical outcome of 43 intracranial aneurysms in 32 patients (14 male, 18 female), aged 2 months to 18 years (mean 11.7 years), seen between 1977 and 2003, were reviewed at the Department of Neurological Surgery, University of California at San Francisco. Outcomes were assessed during office examinations and telephone interviews. Of 9 patients (28%) presenting with neurological symptoms and signs caused by mass effect of giant aneurysms, 4 had a cavernous sinus syndrome. Seven patients (22%) presented with rupture of the aneurysm and subarachnoid hemorrhage (SAH). Six (19%) were diagnosed during evaluation of chronic headaches not caused by SAH, and another 6 (19%) in the work-up for tuberous sclerosis, optic atrophy, cranial neuropathy, coarctation of the aorta, facial angioma, and hypothalamic pilocytic astrocytoma. Overall, 9 patients (28%) had medical comorbidities (eg. polycystic kidney disease, Henoch-Schonlein purpura, protein C deficiency, dwarfism), and 6 (19%) had a history of head trauma. Of 8 patients with more than one aneurysm, 4 developed after treatment of another aneurysm.

Most aneurysms were located in the anterior circulation (72%), including the middle cerebral (MCA) and cavernous internal carotid (ICA) arteries. The posterior circulation was involved in 28%, the vertebrobasilar junction (VBJ) and basilar artery (BA) being the most common sites. Seventeen (40%) were giant aneurysms, nine (21%) were large, 22 (51%) fusiform or dolichoectatic, and the remainder saccular. Treatment was by microsurgery in 13 patients (clipping, bypass/trapping etc) and endovascular in 16 (balloon occlusion, coil etc). Comparison of the two methods of treatment showed that complete obliteration of the aneurysm was obtained in 94% of the microsurgical and 82% of the endovascular group. None of the microsurgical group recurred, whereas 3 (14%) of the endovascular group recurred (mean follow-up 5.7 years). De novo aneurysms formed in 1 (8%) following

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microsurgery and in 3 (19%) with endovascular surgery. Treatment-related neurological morbidity occurred in 2 (7%) patients, 1 in each group, and no patient died as a result of surgery. (Sanai N, Quinones-Hinojosa A, Gupta NM, et al. Pediatric intracranial aneurysms: durability of treatment following microsurgical and endovascular management. **J Neurosurg (2 Suppl Pediatrics)** Feb 2006;104:82-89). (Reprints: Michael T Lawton MD, Department of Neurological Surgery, University of California, 505 Parnassus Avenue, M-780C, San Francisco, CA 94143).

COMMENT. Diagnostic risk factors for intracranial aneurysm in children include symptoms and signs of an acute mass lesion, subarachnoid hemorrhage, chronic headaches, a history of head trauma, and certain medical comorbidities, including polycystic kidneys, aortic coarctation, and bleeding disorders. The aneurysms in children are usually large and located anteriorly at the internal carotid bifurcation or posteriorly; in adults they are small, saccular and located on the circle of Willis. Treatment of pediatric aneurysms by microsurgery may be more effective than balloon occlusion, and the long-term benefits more durable.

Heros RC, in an editorial, notes the differences in the current and previous reviews of pediatric intracranial aneurysms compared to adults (**J Neurosurg (2 Suppl Pediatrics)** 2006;104:77-88). Treatment is more successful in children, especially with collaboration of pediatric neurosurgeon and neurovascular surgeon, a practice favored by Dr Sanai and colleagues.

NEONATAL PORENCEPHALY, ADULT STROKE, AND COLLAGEN IV A1 MUTATION

The relation of leukoencephalopathy, lacunar infarcts, micro- and macro-bleeds to a defect in collagen IV A1 gene was examined in a family with the mutation and autosomal dominant porencephaly followed at VU University Medical Center, Amsterdam, the Netherlands. Porencephaly was diagnosed in the mother at age 24 years, while her 2 children had symptoms and signs of porencephaly in infancy. The mother developed recurrent strokes beginning at age 42 years. All 3 patients had leukoencephalopathy. MRI showed lacunar infarcts, and micro- and macro-bleeds. Electron microscopy of skin vessels showed interruptions and thickening of the basement membrane. A heterogeneous mutation in the collagen IV A1 gene, a component of the vascular basement membrane, was found in all 3 patients, but was absent in the father and in 192 matched Dutch controls. This mutation is a risk factor for a microangiopathy, resulting in perinatal hemorrhage and porencephaly, leukoencephalopathy, and later onset ischemic and hemorrhagic strokes. (van der Knaap MS, Smit LME, Barkhof F, et al. Neonatal porencephaly and adult stroke related to mutations in collagen IV A1. **Ann Neurol** March 2006;59:504-511). (Respond: Dr van der Knaap, Department of Child Neurology, VU University Medical Center, PO Box 7057, 1007 MB Amsterdam, the Netherlands).

COMMENT. Porencephaly, a term first coined by Heschl in 1859 to designate a congenital defect extending from the surface of the brain to the lateral ventricle, was later defined as any cavity within the brain tissue, with or without extension to the ventricle or subarachnoid space, of prenatal or postnatal origin (Norman RM. In Greenfield's