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Cerebral aneurysms in childhood and adolescence

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✓ In this study, 24 aneurysms occurring in 23 patients under the age of 18 years (mean 12 years) are analyzed. The male:female ratio was 2.8:1, and the youngest patient was 3 months old. Mycotic lesions and those associated with other vascular malformations were excluded. Forty-two percent of the aneurysms were located in the posterior circulation, and 54% were giant aneurysms. Presenting symptoms included subarachnoid hemorrhage in 13 and mass effect in 11. Several of these aneurysms were documented to rapidly increase in size over a 3-month to 2-year period of observation. All aneurysms were surgically treated: direct clipping was performed in 14; trapping with bypass in four; trapping alone in four; and direct excision with end-to-end anastomosis in two. The postoperative results were excellent in 21 aneurysms (87%), good in two (8%), and poor in one. The pathogenesis of cerebral aneurysms is reviewed.

KEY WORDS • **cerebral aneurysm** • **congenital anomaly** • **traumatic aneurysm** • **children**

INTRACRANIAL aneurysms in the pediatric age group are rare neurosurgical lesions, occurring at a frequency of approximately 0.5% to 4.6% in large aneurysm series.[15,19–21,28] Analysis of previous reports indicates several distinct characteristics of this entity.[2,4,11,15,16,18,19,21,25,27,29] First, there is a predominant male:female ratio approaching 2:1 to 3:1. Second, a disproportionately high number of these aneurysms (40% to 45%) arise in the posterior circulation. Third, a high percentage of the aneurysms that develop in the anterior circle of Willis are located at the carotid bifurcation. Fourth, approximately 30% to 45% are giant aneurysms.

These identifiable characteristics suggest that aneurysms in the younger age group may be a distinct pathophysiological entity from aneurysms in the adult population. The goal of this report is to review our surgical experience with 24 aneurysms which occurred in 23 patients under the age of 18 years.

Summary of Cases

Clinical Material

During the period from January, 1967, through September, 1987, 1387 aneurysms were operated on at the Mayo Clinic. Of these, 24 aneurysms occurred in 23 patients under the age of 18 years. These were evaluated and surgically treated. Excluded from this group are patients who suffered from mycotic lesions, associated vascular anomalies including arteriovenous malformations, and vein of Galen aneurysms. The average age was 12 years (range 3 months to 18 years), and 14 patients were under this age. There were 17 males and six females (ratio 2.8:1). Presenting signs and symptoms included subarachnoid hemorrhage (SAH) in 13 aneurysms and mass effect in 11.

The location and size of these aneurysms are listed in Table 1. Aneurysms were defined as being saccular (0 to 10 mm in size), globular (11 to 25 mm in size), and giant (> 25 mm in size) on the basis of angiographic and intraoperative measurements. Eleven (46%) were located in the posterior circulation. Of the 13 (54%) giant aneurysms, eight were in children under the age of 12 years, and eight occurred in the posterior circulation. All four aneurysms of the posterior cerebral artery (PCA) were giant in size.

There were no instances of associated systemic diseases like hypertension, coarctation of the aorta, polycystic kidney disease, or connective tissue disorders. However, it is recognized that these latter two conditions may not be manifest before adulthood. One patient did have diffuse dilatation of the entire vertebrobasilar system suggestive of an intrinsic vascular disorder. This patient (Case 3) was initially treated for

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TABLE 1

Location and size of 24 aneurysms in 23 patients

Arterial Site of Aneurysm	Saccular (0–10 mm)	Globular (11–25 mm)	Giant (> 25 mm)
anterior circulation			
cavernous internal carotid	0	0	3
internal carotid bifurcation	0	1	1
internal carotid-ophthalmic	1	1	0
posterior communicating	3	0	0
middle cerebral	0	1	1
anterior cerebral	1	0	0
posterior circulation			
basilar	0	1	3
posterior cerebral	0	0	4
superior cerebellar	0	1	0
posterior inferior cerebellar	0	1	0
vertebral	0	0	1
total	5	6	13

a giant internal carotid artery (ICA) aneurysm and subsequently underwent a second operation 2 years later for a giant basilar artery aneurysm. This case is counted as two surgical procedures in the computation of results.

Of the 13 aneurysms presenting with SAH, 10 were in patients classified preoperatively in Botterell Grade 1, two were in Grade 2 patients, and one was in a Grade 3 patient. Outcome was defined as: excellent, with no neurological deficit and normal development; good, with minimal residual deficit but normal development; and poor, with significant neurological deficit and impairment of intellectual or physical development.

Operative Results

The surgical procedures utilized to treat these aneurysms included direct clipping in 14, trapping in four, trapping with bypass in four, and resection with end-to-end anastomosis in two. The surgical results were excellent in 21 cases (87%), good in two cases (8%), and poor in one case. The poor result occurred in the patient who was preoperatively in Grade 3 whose deficit did not significantly improve after surgery. There was one late death from SAH which occurred 5 months after surgical repair of a giant fusiform aneurysm of the basilar artery.

There was no significant correlation between surgical results and the patient's age, size of aneurysm, or clinical presentation. The mean overall follow-up period was 3.5 years (1 to 7 years).

Illustrative Cases

Case 1

This 13-year-old left-handed boy had the gradual onset of a left occipital headache which persisted for 2 days and was associated with a stiff neck. Seven days after the ictus he was noted to have a normal examination by a neurologist. Despite a diagnosis of migraine headache, a contrast-enhanced computerized tomography (CT) scan was performed which demonstrated a 1-cm lesion adjacent to the left ambient cistern in the mesial temporal lobe (Fig. 1 *left*). Although this appearance was typical of aneurysms in this location, conservative observation was recommended. Six weeks later, a second CT scan showed that the lesion had increased threefold in size as compared to the original CT scan. At this point, an angiogram demonstrated a 3-cm aneurysm of the P_2 segment of the left PCA (Fig. 1 *right*).

The patient was transferred to our institution and underwent a left subtemporal craniotomy with spinal drainage. The aneurysm arose without an obvious neck from the P_2 segment of the PCA just distal to the medial posterior choroidal artery. The aneurysm was initially ligated with a 7-0 Prolene suture as it was thought that a clip would occlude the PCA. The aneurysm was then opened, thrombus was removed, and a neck was fashioned from the sac to facilitate placement of a clip. The patient made an excellent recovery.

Case 2

This 3-month-old baby boy was born at full term without intrauterine complications. His growth curve and head circumference were at the 50% level, and he was achieving the predicted milestones. He had the acute onset of obtundation, and on examination was drowsy and febrile with meningismus but without a focal neurological deficit. A lumbar puncture was suggestive of SAH. A CT scan with and without contrast enhancement confirmed the presence of widespread subarachnoid blood in the basal cisterns and identified an area of enhancement in the region of the left circle of Willis. On Day 6 following the SAH, he underwent retrograde angiography of the left ICA under general anesthesia through a catheter placed into the left external carotid artery since cannulation of his femoral artery proved not technically feasible. This intraoperative angiogram demonstrated an aneurysm of the distal left ICA.

A left pterional craniotomy was performed and a giant, partially thrombosed aneurysm was identified arising from the anterior wall of the ICA between the posterior communicating artery and the bifurcation. Clip placement was attempted but proved inadequate as the clip compromised the lumen of the ICA. The ICA, middle cerebral artery (MCA), and anterior cerebral artery were temporarily occluded for 20 minutes to facilitate resection of the aneurysm with primary closure of the ICA by means of a 9-0 Prolene suture. Inspection after removal of the clips demonstrated good flow through all vessels without lumen compromise. The child made an excellent recovery. Pathological examination demonstrated that this was a true congenital aneurysm in that there was fragmentation of both the internal elastic membrane and muscularis layer on the parent vessel adjacent to the neck. There was absence of both layers in the wall of the aneurysm (Fig. 2).

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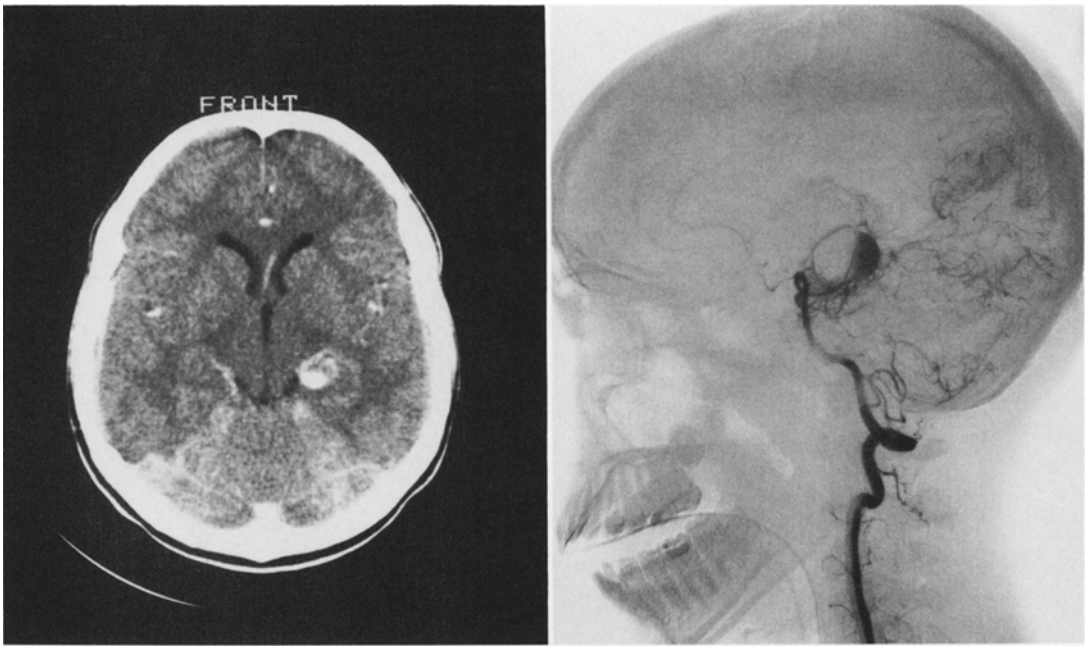

FIG. 1. Case 1. *Left:* Contrast-enhanced computerized tomography scan demonstrating a 1-cm lesion in the mesial left temporal lobe. This has a typical appearance of aneurysms in this location with a contrast layer within a lumen surrounded by a peripheral rim without significant mass effect. In spite of this, conservative treatment was recommended. *Right:* Angiogram demonstrating a 3-cm giant aneurysm of the P_2 segment of the posterior cerebral artery. This was treated by direct clipping of aneurysm neck.

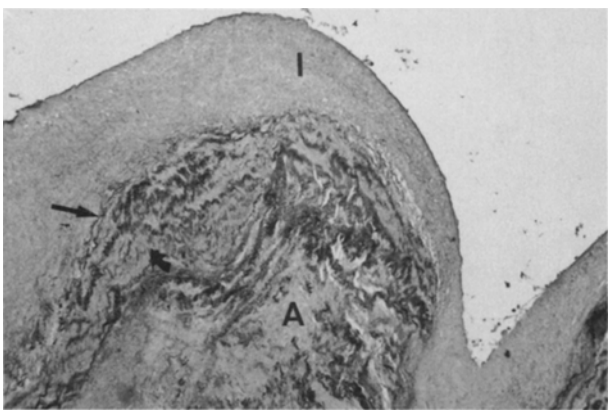

FIG. 2. Case 2. Photomicrograph of the aneurysm neck showing fragmentation and nearly total absence of both the internal elastic membrane *(straight arrow)* and muscularis layer *(curved arrow)*, thickened intima (I), and adventitia with reactive fibrosis (A). EIVG, × 70.

Case 3

This 12-year-old boy had a 3-month history of right retro-orbital headaches and more recent complaints of loss of vision in the right eye. Examination was remarkable for optic atrophy in the right eye and a left superior quadrantanopsia. A contrast-enhanced CT scan demonstrated a large lesion in the right parasellar region with extension into the sphenoid sinus. On angiography there was a giant fusiform aneurysm of the right ICA extending from the sphenoid sinus to the proximal MCA (Fig. 3 *left*). The giant ICA aneurysm was treated by right cervical ICA ligation with maintenance of hemispheric blood flow by means of a superficial temporal artery-MCA bypass. Intraoperative xenon blood flow measurements indicated that the bypass had a flow of approximately 40 ml/100 gm/min immediately after the anastomosis (Fig. 3 *right*). The patient made an excellent recovery from this surgery. It is pertinent to the outcome of this patient that posterior circulation angiography performed at this time demonstrated mild dilatation of the left vertebral and basilar arteries (Fig. 4 *upper left*).

Two years later, the child had the onset of renewed headaches associated with emesis. He also exhibited diplopia on right lateral gaze. His examination was remarkable for a complete right sixth nerve palsy and partial third nerve paresis. Cerebral angiography demonstrated a bilobed giant fusiform aneurysm of the basilar artery which had markedly increased in size when compared to his earlier angiogram (Fig. 4 *upper right* and *lower left*). The patient tolerated a trial balloon-occlusion of the left vertebral artery for 10 minutes without demonstrable neurological deficit. Angiography performed during the occlusion demonstrated a patent left posterior communicating artery along with collateral muscular branches to the vertebral artery. With intraoperative monitoring, the base of the aneurysm was clipped with preservation of the left anterior inferior cerebellar artery which arose just proximal to the neck. The patient awoke with a mild hemiparesis

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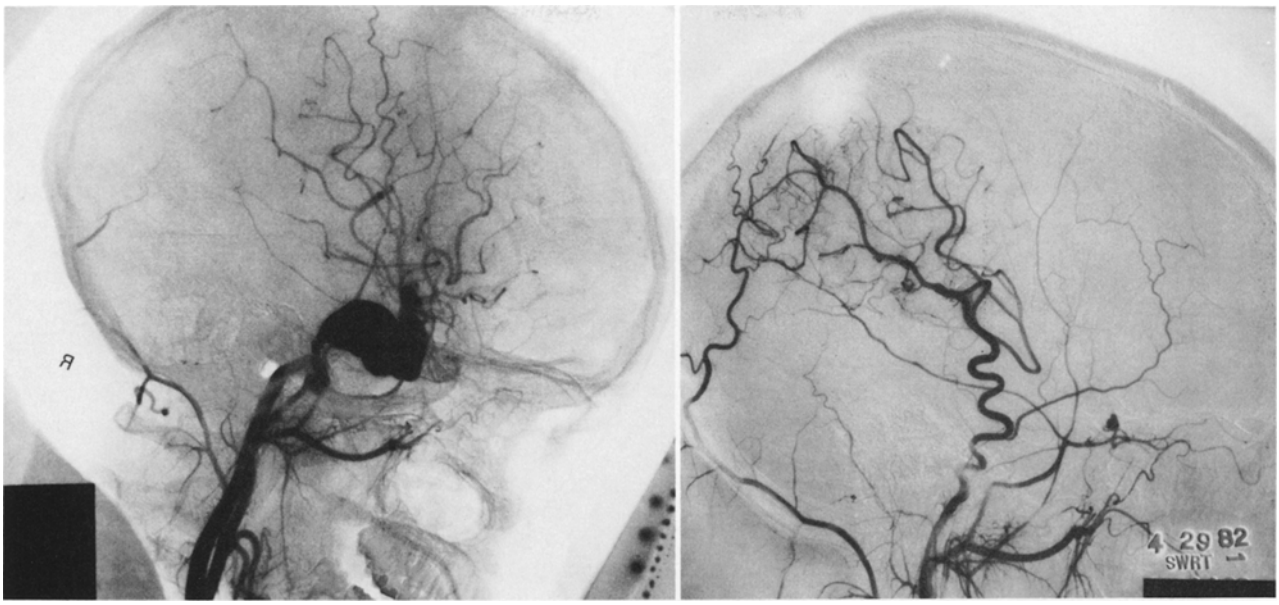

FIG. 3. Case 3. *Left:* Angiogram demonstrating a giant aneurysm of the internal carotid artery extending from the sphenoid sinus to the bifurcation. *Right:* Angiogram obtained after the carotid aneurysm was treated by cervical internal carotid artery ligation. Protection of hemispheric blood flow was insured through a superficial temporal-middle cerebral artery bypass. Intraoperative cerebral blood flow measured 40 ml/100 gm/min through the graft.

which resolved to a significant degree over the ensuing 10 days. Prior to discharge, he had only a slight decrease in manual dexterity and postoperative angiography demonstrated successful clipping of the aneurysm (Fig. 4 *lower right*).

Five months after the second surgery, the patient complained of dizziness, headache, and diplopia. Two days later he became acutely obtunded with decerebrate posturing. A CT scan with and without contrast demonstrated subarachnoid blood in the basal cisterns and a 25-mm enhancing lesion compatible with a recurrent basilar aneurysm. The child died 10 hours later, and an autopsy was declined.

Discussion

From a clinical perspective it is important to emphasize that, although rare, aneurysms in the pediatric population do occur. A recurrent theme in the history of those patients who presented initially with SAH was the general failure of the attending physician to consider an aneurysm in the initial differential diagnosis. With current microsurgical and neuroanesthetic techniques, the general failure of the attending physician to consider an aneurysm in the initial differential diagnosis. With current microsurgical and neuroanesthetic techniques, an excellent result can be obtained in the large majority of these patients.

The patient profile in this surgical group composed largely of individuals referred from other institutions reconfirms the observations previously noted. First, the male:female ratio shows a male predominance in pediatric aneurysms in series reported by Matson[16] (12:1), Locksley[15] (3:1), Thompson, *et al.*[27] (2.5:1), and Amacher and Drake[2] (2:1). Although several reviews found a ratio approaching 1:1, this incidence is still contrary to the definitive female predominance found in adult aneurysm series.

Second, the disproportionately large number of posterior circulation aneurysms in our series (46%) has been found in series by Amacher and Drake[2] (59%) and Storrs, *et al.*[25] (35%). This may reflect the referral nature of these neurosurgical practices, however, because large series analyzing SAH in children have failed to substantiate this. The high frequency of carotid bifurcation aneurysms recorded by Sedzimir and Robinson[21] (36%), Pasqualin, *et al.*[18] (37%), Gerosa, *et al.*[11] (32%), Patel and Richardson[19] (34%), Storrs, *et al.*[25] (31%), and Almeida, *et al.*[1] (54%), was not reconfirmed in our series.

Third, the high number of giant aneurysms in our group of patients (54%) is similar to results obtained by Storrs, *et al.*[25] (31%), Amacher and Drake[2] (45%), and Gerosa, *et al.*[11] (20%). This may again reflect the referral nature of our practice, as an equal number of reports do not verify this fact.

Despite these apparent discrepancies, it appears reasonable to conclude that pediatric aneurysms are distinct from their counterparts in adults on the basis of sex predominance, location, and size. Perhaps this information can facilitate an understanding concerning the genesis of cerebral aneurysms.[4] Discussion on the evolution of cerebral aneurysms has always focused on the question of the congenital versus the acquired nature of these lesions. In aneurysms in the adult popu-

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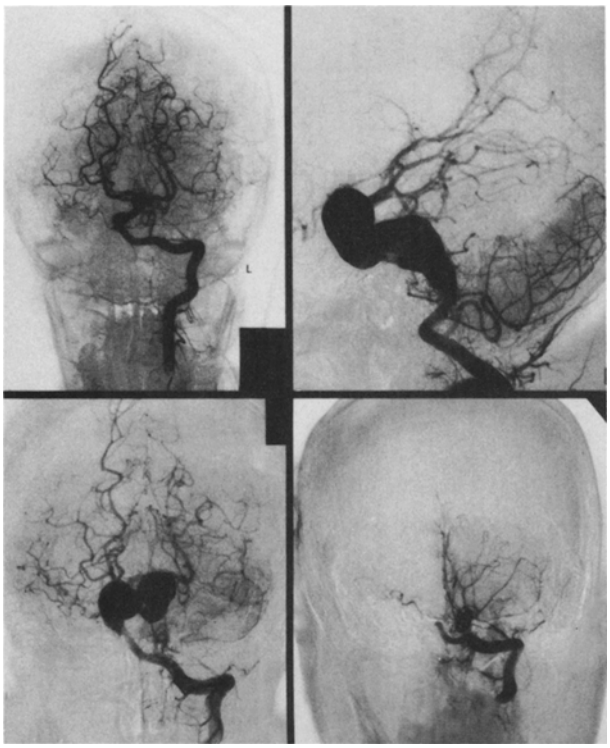

FIG. 4. Case 3. *Upper Left:* Left vertebral angiogram obtained during evaluation of the giant carotid aneurysm demonstrating mild irregularity and dilatation of both the vertebral and basilar arteries. *Upper Right* and *Lower Left:* Angiograms obtained 2 years later demonstrating progression of the basilar dilatation into a giant bilobed aneurysm. The patient tolerated a 10-minute trial balloon-occlusion of the vertebral artery due to collateral flow from a patent left posterior communicating artery. The base of the aneurysm was clipped from a suboccipital approach with preservation of the left anterior inferior cerebellar artery. *Lower Right:* Postoperative angiogram demonstrating obliteration of the aneurysm. The patient awoke with a mild hemiparesis which improved over the next 10 days. Upon discharge, the patient's only deficit was a mild decrease in manual dexterity.

lation, light microscopy routinely demonstrates fragmentation and absence of both the internal elastic membrane and muscularis layer of the media. Forbus[10] noted that there were often developmental defects in the muscularis layer of the media in nonpathological cerebral artery bifurcations. Presumably, medial wall defects could facilitate distention of a thinning internal elastic membrane. However, the frequent occurrence of medial wall defects is significantly greater than the incidence of aneurysms.[26] Although it was originally thought that the internal elastic membrane herniated through medial wall defects,[13,28] Glynn[12] demonstrated that the internal elastic membrane alone could withstand distending pressures of 600 mm Hg. In his analysis of 149 autopsy cases, Crompton[7] noted medial wall defects in cerebral artery bifurcations which appeared to increase with age. He observed that degeneration first appeared in the intimal pads proximal to bifurcations which then tended to extend to intima underlying the medial wall defects. Therefore, it is now generally accepted that injury to the internal elastic lamina by hemodynamic forces is the initial pathophysiological alteration.[8] This usually occurs at arterial bifurcations because this is the site of greatest shearing forces against the arterial wall. In addition, pathological analyses of arterial bifurcations have shown that fenestrations of the internal elastic membrane are greatest at the apex.[6] Based on these facts, the development of cerebral aneurysm is considered to be an acquired degenerative phenomenon perhaps associated with congenital defects of the media.[24]

A congenital basis for aneurysms has been promoted by some authors, who have cited the rare existence of saccular aneurysms in children as an argument against a degenerative process. One early theory proposed that the aneurysms arose from remnants of small vascular trunks stemming from arterial bifurcations.[5,14] Unfortunately, pathological studies of pediatric aneurysms are limited. Most reported cases indicate that the histology of these aneurysms resembles that of adult saccular aneurysms, showing absence of both the internal elastic membrane and muscularis layer of the media. Our Case 2 confirms this finding in a 3-month-old baby. There are, however, a few reports of pediatric aneurysms with absent media but intact elastic membrane.[4,9,17] Stehbens[22–24] has critically reviewed published histological reports of pediatric aneurysms and has concluded that most studies contain insufficient evidence to support a congenital mechanism. He also appropriately noted that the existence of a few true congenital saccular aneurysms does not refute a degenerative mechanism for the vast majority of these lesions.

Nonetheless, it is difficult to explain the unique features of pediatric aneurysms when compared to those in adults: male predominance, a high percentage of giant aneurysms, and an unusual location.[3] One possible explanation for at least some of these aneurysms is a traumatic etiology. In four patients, there were aneurysms of the PCA at the P_2 segment as it traverses the tentorium. Possibly, during times of increased intracranial pressure (for example, during delivery), there was herniation of the PCA over the tentorium causing vessel injury. Since the hemodynamic stress would be minimal as compared to a bifurcation, it might be anticipated that this type of aneurysm would be prone to grow slowly into a giant lesion prior to rupture.

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