# Intracranial Arteriovenous Malformations Associated with Aneurysms

Hunt Batjer, M.D., Richard A. Suss, M.D., and Duke Samson, M.D.

Division of Neurosurgery, Department of Surgery (HB, DS), and Division of Neuroradiology (RAS), University of Texas Health Science Center, Dallas, Texas

The presence of intracranial aneurysm in association with arteriovenous malformation has been well documented. Aneurysms have been described in typical proximal sites along the feeding system to the arteriovenous malformation, in abnormal distal locations along feeding vessels, and in sites remote and apparently hemodynamically unrelated to the arteriovenous malformation. Little attention has been focused on the most appropriate medical and surgical care of patients harboring these lesions. Since 1977, 22 patients with this combination of lesions have been evaluated at our institution. Nine patients (41%) presented after intracranial hemorrhage. The remaining 13 patients were investigated because of seizures in 5 patients (23%), headaches in 4 patients (18%), and progressive ischemia in 4 patients (18%). Among the patients suffering intracranial hemorrhage, 78% had bled from an aneurysm, with 22% having hemorrhaged from their arteriovenous malformation. All 7 of the patients who suffered aneurysmal hemorrhage bled from atypical distal aneurysms on major feeding vessels. Our experience and that of others has led us to believe that the safest approach to patients with this combination of lesions is to treat the aneurysm before microsurgical resection of the associated arteriovenous malformation. Hemodynamic changes associated with the abrupt elimination of an arteriovenous malformation may place associated aneurysms at immediate risk. (Neurosurgery 18:29–35, 1986)

Key words: Aneurysm, Arteriovenous malformation, Subarachnoid hemorrhage

Walsh and King in 1942 reported the first example of a patient harboring an aneurysm and an arteriovenous malformation (AVM) (19). After their publication, numerous case reports and small series began to appear in the literature (2, 3, 5, 17). That this combination of lesions was particularly hazardous both with and without surgical therapy was emphasized by Perret and Nishioka in 1966 in the Cooperative Aneurysm Study (13). In their report, 9 of 18 conservatively managed patients and 6 of 16 surgically managed patients died. Gamache et al. in 1981 reviewed 42 medically managed patients from the literature. They found that 38% died, 7% had significant neurological difficulties, and only 14% were doing well (6).

Several published series have suggested that between 2.7% (12) and 9.3% (9) of patients with AVMs have associated aneurysms. Most recent series using current diagnostic measures imply that the incidence of this combination is probably higher than the expected natural occurrence of aneurysms in the general population. These reports have described aneurysms in typical proximal sites along the feeding system to the AVM, in abnormal distal locations along feeding vessels, and in sites remote and apparently hemodynamically unrelated to the AVM. The apparent clustering of aneurysms along feeding vessels has provoked considerable discussion about the mechanism of formation of aneurysms in general. Most publications have dealt with the descriptive aspects of this patient population; significantly less attention has been paid to the most appropriate medical and surgical management of these patients.

Since 1977, 22 patients with this combination of intracranial vascular lesions have been evaluated and treated at The University of Texas Health Science Center at Dallas. This group comprises 14% of our total operative AVM series during this time. The present retrospective review was undertaken to examine this population carefully in terms of symptoms, the anatomical relationship of aneurysms to AVMs, and therapeutic morbidity and mortality.

## CLINICAL MATERIAL

Twenty-two patients with an intracranial AVM and at least one arterial aneurysm have been evaluated and treated by us since 1977. Most of these patients were referred to our institution by neurologists and neurosurgeons for the consideration of surgical therapy. The remaining patients presented to Parkland Memorial Hospital from the Dallas area after the development of neurological symptoms. The referral nature of our patient population may in part explain the heavy predominance of aneurysms in our AVM patients.

Fifty-nine per cent of our patients were male, and 41% were female. The patients ranged in age from 11 to 55 years. Nine patients (41%) presented after intracranial hemorrhage (Table 3). There were 7 cases of subarachnoid hemorrhage (SAH), 1 case of intraparenchymal hemorrhage, and 1 case of isolated intraventricular bleeding. The remaining 13 patients were investigated because of seizures in 5 patients (23%), headaches in 4 patients (18%), and progressive ischemia in 4 patients (18%). Nineteen of the 22 patients underwent surgical procedures; operation was directed toward the aneurysm alone in 8 cases, toward the AVM in 3 cases, and at both lesions in 8 patients.

## ILLUSTRATIVE CASE REPORTS

Case 6

This 47-year-old, right-handed man developed double vision insidiously over several weeks, followed by a SAH. He was admitted to our institution in Hunt-Hess Grade I status. His initial computed tomographic (CT) scan revealed a small amount of localized blood in his left ambient cistern. Angiography demonstrated a distal superior cerebellar aneurysm located on a major arterial feeder to a left cerebellar hemisphere AVM (Fig. 1). At operation on the following day, his superior cerebellar aneurysm was encased in fresh clot. This

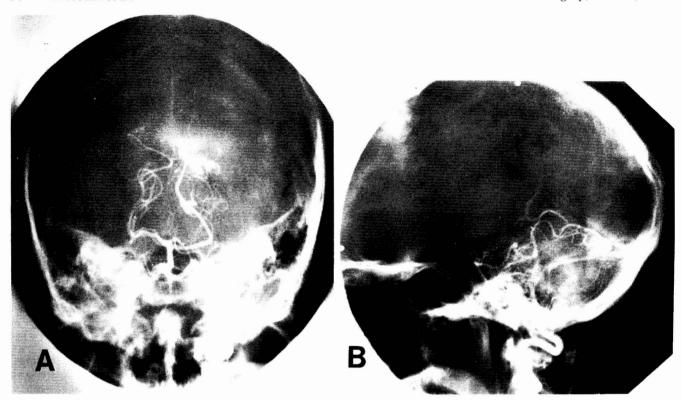


FIG. 1. Case 6. A 47-year-old man with a new left 4th nerve palsy and SAH. Anteroposterior (A) and lateral (B) vertebral arteriograms demonstrate a ruptured distal left superior cerebellar artery aneurysm and a left cerebellar hemisphere AVM fed by the left superior cerebellar artery.

aneurysm was clipped, sparing the 4th nerve, which was stretched over its fundus. One month later, the patient was readmitted for the clipping of an incidental right middle cerebral artery aneurysm and the subsequent removal of his small AVM. He remains neurologically intact.

# Case 3

This 45-year-old, right-handed man was transferred to our hospital 2.5 weeks after a SAH from which he had completely recovered. A CT scan showed relatively diffuse subarachnoid blood, perhaps most apparent in the interhemispheric fissure. Angiography revealed a large AVM involving the left mesial occipital lobe, the left pulvinar and splenium, which were fed by the left anterior cerebral artery, and the left posterior cerebral artery (Fig. 2). Also present were a large, basilar P-1 aneurysm on the right, a left P-1-P-2 junction aneurysm, a small intracavernous carotid aneurysm, an anterior communicating artery aneurysm, and a distal left pericallosal artery aneurysm. At operation, the large basilar aneurysm and then the smaller P-1-P-2 junction aneurysm were clipped. Next, the small and unruptured anterior communicating artery aneurysm was clipped. As the source of hemorrhage had not yet been found, the dissection through the gyrus rectus was continued following the left A-2 branch. Fresh hemorrhage was noted in the subarachnoid space and in the brain parenchyma immediately surrounding the patient's pericallosal aneurysm. This aneurysm was clipped and evacuated. The patient made a slow but steady recovery from his craniotomy and is now back to his premorbid state of health. We have elected to defer resection of his large and asymptomatic AVM.

## Case 15

This 17-year-old, right-handed boy sustained a sudden ictus and was admitted to the hospital in a bilaterally decerebrate condition with a dilated and fixed left pupil. A CT scan revealed a large parietal hemorrhage with significant subarachnoid blood (Fig. 3). Immediate arteriography revealed a small parietal AVM fed predominantly by the contralateral anterior cerebral artery. There was an irregular fusiform aneurysm on the anterior cerebral artery in immediate proximity to the malformation and hematoma. Emergency operation for evacuation of the hematoma and clipping of the ruptured aneurysm was performed, but the patient's neurological deficit did not improve. He ultimately died a neurological death 2 weeks postoperatively.

#### Case 8

This 22-year-old, right-handed woman presented with SAH during pregnancy. Arteriography demonstrated a right frontal AVM with feeding from the right pericallosal artery (Fig. 4). Two proximal carotid wall aneurysms were noted. It was presumed, probably incorrectly, that the patient had bled from her AVM, and she was managed as a high risk pregnancy. After the completion of her pregnancy, she underwent the resection of her AVM and the clipping of her two proximal carotid wall aneurysms. Unfortunately, 2 weeks postoperatively she sustained a second SAH. She subsequently underwent the clipping of her previously unrecognized distal anterior cerebral artery aneurysms and ultimately made a full neurological recovery.

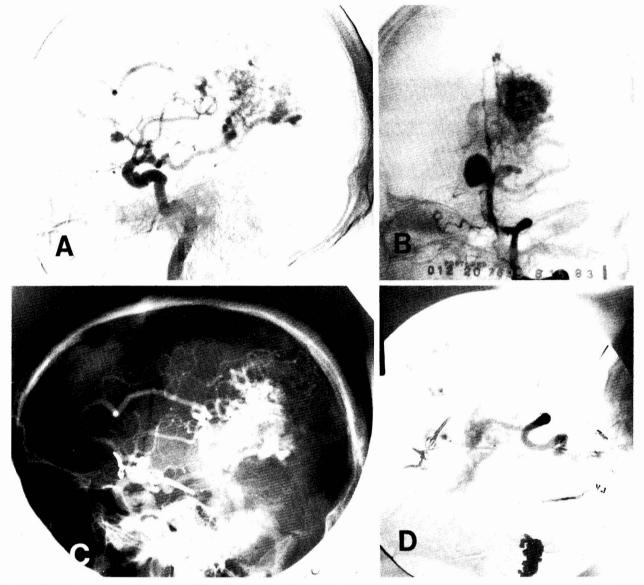


FIG. 2. Case 3. A 45-year-old man with diffuse SAH. A and B, preoperative arteriograms demonstrate pericallosal, carotid wall, basilar, and P-1-P-2 junction aneurysms involved with the feeding system to a deep thalamic mesial occipital AVM. C and D, postoperative arteriograms show clipping of the ruptured anterior cerebral aneurysm and other asymptomatic intradural aneurysms.

## **RESULTS**

Five of the 19 operative patients have died, for an operative mortality rate of 26%. One of these patients (Case 10) underwent the clipping of his aneurysms and multiple feeders to his AVM as the initial stage of a planned two-stage resection. He tolerated the procedure well, but died suddenly 1 week later of pulmonary embolism.

A second patient (Case 15) was left comatose after a devastating subarachnoid and intraparenchymal hemorrhage. His operation was directed at the removal of his life-threatening clot and the clipping of his aneurysm. He unfortunately did not improve after operation and ultimately died of uncontrolled intracranial hypertension.

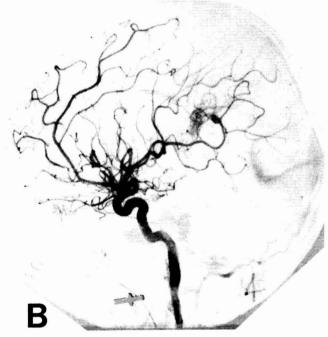
Two patients (Cases 1 and 11) died as a result of massive intraoperative hemorrhage. Patient 1 harbored a large (5-cm), right, prerolandic malformation that had been treated by

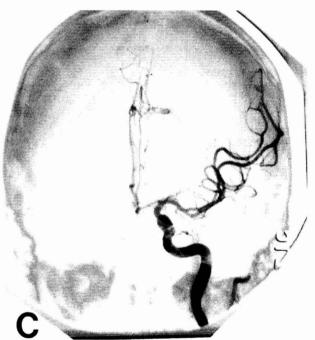
staged ligation of middle cerebral and anterior cerebral feeders and by clipping of her incidental distal middle cerebral aneurysm. One week later, she returned to the operating room for definitive resection. Late in the dissection, after her lateral ventricle had been entered and the major thalamostriate draining vein had been dissected but not sacrificed, the brain became tense and numerous previously coagulated vessels in the gyral bank surrounding her malformation suddenly began to bleed. Her AVM was quickly removed without abatement of the hemorrhage. Her carotid artery was ultimately sacrificed intracranially after hypotension failed to improve the situation. Carotid ligation allowed control of this catastrophic bleeding, but the patient developed severe neurological deficit postoperatively and died 6 days later. We suspect that the resection of this large, high flow malformation may have resulted in normal perfusion pressure breakthrough.

Patient 11 had suffered SAH from a distal right superior



Fig. 3. Case 15. A 17-year-old boy who presented bilaterally decerebrate after a sudden ictus. *A*, the CT scan reveals a large parietal hemorrhage with intraventricular and subarachnoid blood. *B* and *C*, preoperative angiograms demonstrate the ruptured anterior cerebral aneurysm with its associated AVM.





cerebellar artery aneurysm on a feeding vessel to a 3-cm cerebellar hemispheric AVM. After recovery from her SAH and the subtemporal clipping of her aneurysm, she underwent an elective suboccipital craniectomy. Because of the broad representation of her malformation on the superior surface of the cerebellar hemisphere, we planned to operate with her in the sitting position. During the initial stages of the dissection, uncorrectable major air embolism was encountered. We quickly rolled her into the prone position and redraped.

During this episode, she never became hypotensive, but her pulmonary pressures were altered significantly. Although the cerebellum was slightly more full than it had been while she was in the sitting position, we unfortunately decided to proceed with the removal of her malformation. Relatively early in the resection of her malformation, a small tuft of vessels was amputated from the vascular mass. The resultant bleeding very shortly became uncontrollable. Despite attempts to extirpate her lesion quickly, massive hemorrhage from within

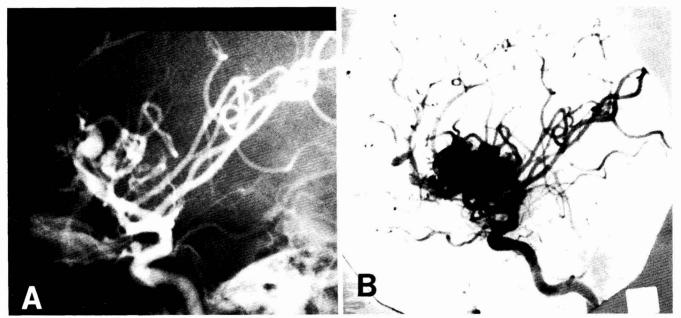


FIG. 4. Case 8. A 22-year-old woman with SAH. A, the preoperative early phase arteriogram shows proximal carotid and distal anterior cerebral aneurysms. B, later phase arteriogram reveals a mesial frontal AVM.

the 4th ventricle developed, and she quickly became hypotensive and suffered cardiac arrest. We suspect that the hemodynamic alterations produced by her air emboli contributed to the escalation of a technical error that should have been relatively easy to correct. In retrospect, the procedure should have been terminated at the onset of serious air embolism.

The final patient (Case 5) never awakened after the resection of a large vermian AVM. Postoperative CT scans revealed that he had sustained brain stem hemorrhage during his operative procedure.

One patient (Case 20) developed a new expressive aphasia after his AVM resection; fortunately, the aphasia is improving. The remaining 13 patients in the operative series (69%) have had good outcomes, with a return to their premorbid occupations.

A striking 50% of our 22 patients harbored multiple aneurysms. A total of 44 aneurysms were detected, with 82% of the aneurysms being located in hemodynamic association with the malformation (see Tables 1 and 2). Fifty per cent of these aneurysms were located proximally along the feeding system of the AVM. If one considers the pericallosal origin a distal site for aneurysmal development, 32% of all aneurysms were located in atypical distal points along major feeding vessels. The remaining 18% of the aneurysms were found in sites remote from the patients' AVMs.

All nine patients who suffered intracranial hemorrhage underwent surgical procedures (Table 3). Two patients in that group died (22%), one of whom was comatose at the time of operation. At present, six of these nine patients (67%) have returned to their previous occupations. The source of hemorrhage in the nine patients who had bled was determined by surgical findings correlated with CT scan and angiographic evidence. Seven of the nine patients (78%) had bled from an aneurysm, with the remaining two patients (22%) having bled from their AVM. All seven patients who had suffered aneurysmal SAH had bled from aneurysms located in atypical distal sites on major feeding vessels. None of the aneurysms at sites remote from the AVM ever bled, to the best of our

TABLE 1
Summary of Clinical Data

	No.
Patients	22
Aneurysms	44
Patients with single aneurysms	11
Patients with multiple aneurysms	11

TABLE 2
Location of Aneurysms

	No. Aneurysms		
	Anterior circulation	Posterior circulation	Total
Proximal on the feeding vessel	17	5	22 (50%)
Distal on the feeding vessel	10	4	14 (32%)
Remote from the AVM	5	3	8 (18%)

Table 3

Outcome in Patients with Hemorrhage

Case	Location of Hemorrhage	Result <sup>a</sup>
1	SAH	Died
3	SAH	Good
6	SAH	Good
7	SAH	Good
8	SAH	Good
15	SAH, intraparenchymal	Died
16	Intraventricular	Good
20	Intraparenchymal	New aphasia (resolving)
22	SAH	Good

<sup>&</sup>lt;sup>a</sup> Good, return to previous occupation.

knowledge. Looked at another way, 68% of our patients presented with symptoms referrable to their AVMs and 32% presented with symptoms from associated aneurysms.

#### DISCUSSION

There has been considerable variability in the literature regarding the exact location of aneurysms associated with AVMs. Cronqvist and Troupp reported a series in which 69% of the aneurysms were located on the main arterial feeders of the malformations (5). Suzuki and Onuma noted a 44% incidence of aneurysms located on major feeders (18). In the Cooperative Study report by Perret and Nishioka, only 37% of the aneurysms were on major feeders, with 42% of the aneurysms in sites remote from the malformation (13).

Several theories have been advanced to explain the apparent association between aneurysms and AVMs. Paterson and McKissock thought that the increased blood flow through the arterial feeders of the AVM contributed to the development of aneurysms (12). Anderson and Blackwood wrote that both lesions were simply part of a constellation of multiple vascular anomalies in the same patient (1). Boyd-Wilson thought that the association of these lesions was probably coincidental (4).

There is considerable circumstantial evidence to support the theory of Paterson and McKissock. The hemodynamic effect of an AVM is reflected primarily in decreased intravascular resistance in its feeding vessels (15) and the attendant marked increase in arterial flow (4). Somach and Shenkin reviewed six patients who had undergone carotid artery ligation for the treatment of intracranial aneurysm (16). Repeat arteriography 3 to 10 years postoperatively revealed that two of these patients had developed aneurysms on the contralateral internal carotid artery. The authors concluded that increased flow in a cerebral artery predisposes to the development of an aneurysm. Conversely, Hayashi et al. (8), Shenkin et al. (14), and Koulouris and Rizzoli (11) all described significant decreases in the size of previously documented aneurysms on feeding vessels to AVMs after surgical obliteration of the AVM. Whether the arteriographic changes are due to intraluminal thrombosis or to intimal thickening is unclear, as is the long term fate of these aneurysms that have apparently diminished in size. Nevertheless, these important cases do lend credence to the relationship between increased flow and the development or growth of intracranial aneurysms. As a result of these latter studies, some authors have concluded that this phenomenon offers a possible alternative to the radical surgical treatment of both intracranial lesions in that excision of the AVM in some cases may result in the eventual obliteration of the associated aneurysm.

Our personal philosophy regarding the surgical approach to these patients is colored by our experience (Case 8) and that of Drake (personal communication, May 1984) and Kassell (10), each of whom has described individual cases of the operative resection of an AVM followed during the immediate postoperative period by disastrous SAH from an aneurysm on a feeding vessel. In spite of the few reported instances of this complication, such occurrences suggest that the sudden increase in vascular resistance associated with the abrupt resection of an AVM puts aneurysms in its feeding system at immediate risk of distension and possibly subsequent rupture. This risk is present even if the aneurysm has not previously bled.

A similar but perhaps less persuasive argument can be made regarding the risk for aneurysm rupture remote from the AVM. After elimination of the sump effect with resection of the AVM, the vessels on which these aneurysms are located may be subjected to increased flow, with a resultant heightened risk of future aneurysmal rupture.

These concerns have led us to modify our surgical approach to patients with combined lesions. In almost all patients presenting with SAH from whatever source, seizures, or ischemic neurological deficits, the intracranial aneurysm is treated first. Data from the Cooperative Study suggest that rebleeding from AVMs is not common during the first several posthemorrhage weeks (13). In fact, Graf et al. found that the risk of rebleeding is only 6% over the 1st year (7). Exceptions to this general rule are cases of large intraparenchymal clots from AVMs or patients with unrelated remote aneurysms. Obliterating the aneurysms and then microsurgically resecting the AVM is the safest approach to symptomatic lesions of this type. In our series, aneurysms on the feeding vessels to AVMs bled commonly, and aneurysms remote from the AVMs did not bleed. This experience causes us to wonder whether the natural history of aneurysms on feeding vessels is more malignant than that of uncomplicated unruptured aneurysms. For this reason, we most frequently recommend elective obliteration of aneurysms associated with AVMs in asymptomatic patients even if subsequent resection of the malformation is not contemplated.

#### **ACKNOWLEDGMENTS**

The authors extend special thanks to the physicians who referred these difficult patients to us and for the secretarial support of Ms. Jolene Jackson and Mrs. Vicki Rankin.

Received for publication, March 12, 1985; accepted, August 24, 1985.

Reprint requests: Hunt Batjer, M.D., Department of Surgery, Division of Neurosurgery, The University of Texas Health Science Center at Dallas, 5323 Harry Hines Boulevard, Dallas, Texas 75235.

#### **REFERENCES**

- Anderson RM, Blackwood W: The association of arteriovenous angioma and saccular aneurysm of the arteries of the brain. J Pathol Bacteriol 77:101-110, 1959.
- Arieti S, Gray EW: Progressive multiform angiosis: Association of a cerebral angioma, aneurysms and other vascular changes in the brain. Arch Neurol Psychiatry 51:182–189, 1944.
- Aring CD: Neurological clinical pathological conference of the Cincinnati General Hospital. Dis Nerv Syst 6:119–124, 1945.
- Boyd-Wilson JS: The association of cerebral angiomas with intracranial aneurysms. J Neurol Neurosurg Psychiatry 22:218– 316, 1966.
- Cronqvist S, Troupp H: Intracranial arteriovenous malformation and arterial aneurysm in the same patient. Acta Neurol Scand 42:307–316, 1966.
- Gamache FW, Drake CG, Peerless SJ, Girvin JP: Intracranial arteriovenous malformations associated with intracranial aneurysm: A review. Presented at the 50th Annual Meeting of the American Association of Neurological Surgeons, Boston, Massachusetts, April 8, 1981.
- Graf CJ, Perret GE, Torner JC: Bleeding from cerebral arteriovenous malformations as part of their natural history. J Neurosurg 58:331–337, 1983.
- 8. Hayashi S, Arimoto T, Itakura T, Fujii T, Nishiguchi T, Komai N: The association of intracranial aneurysms and arteriovenous malformation of the brain: Case report. J Neurosurg 55:971–975, 1981.
- Higashi K, Hatano M, Yamashita T, Inoue S, Matsumura T: Coexistence of posterior inferior cerebellar artery aneurysm and arteriovenous malformation fed by the same artery. Surg Neurol 12:405–408, 1979.
- 10. Kassell NF: Comment on Koulouris S, Rizzoli HV: Coexisting

- intracranial aneurysm and arteriovenous malformation: Case report. Neurosurgery 8:222, 1981.
- Koulouris S, Rizzoli HV: Coexisting intracranial aneurysm and arteriovenous malformation: Case report. Neurosurgery 8:219– 222, 1981.
- Paterson JH, McKissock WA: A clinical survey of intracranial angiomas with special reference to their mode of progression and surgical treatment: A report of 110 cases. Brain 79:233–266, 1956.
- Perret G, Nishioka H: Report on the Cooperative Study of Intracranial Aneurysms and Subarachnoid Hemorrhage: Section VI. Arteriovenous malformations: An analysis of 545 cases of cranio-cerebral arteriovenous malformations and fistulae reported to the Cooperative Study. J Neurosurg 25:467–490, 1966.
- 14. Shenkin HA, Jenkins F, Kim K: Arteriovenous anomaly of the

- brain associated with cerebral aneurysm: Case report. J Neurosurg 34:225–228, 1971.
- Shenkin HA, Spitz EB, Grant FC, Kety SS: Physiologic studies of arteriovenous anomalies of the brain. J Neurosurg 5:165–172, 1948
- Somach FM, Shenkin HA: Angiographic end-results of carotid ligation in the treatment of carotid aneurysm. J Neurosurg 24:966–974, 1966.
- 17. Sugar O: Pathological anatomy and angiography of intracranial vascular anomalies. J Neurosurg 8:3–22, 1951.
- Suzuki J, Onuma T: Intracranial aneurysms associated with arteriovenous malformations. J Neurosurg 50:742–746, 1979.
- Walsh FB, King AB: Ocular signs of intracranial saccular aneurysms: Experimental work on collateral circulation through the ophthalmic artery. Arch Ophthalmol 27(New Series):1–33, 1942.