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

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Introduction

Virchow in 1863 described many varieties of angiomatous malformations and cited a number of isolated cases described in the first half of the 19th century. In 1928, Cushing and Bailey as well as Dandy '28 reported on small series of arteriovenous “aneurysms” of the brain and described their gross appearance and the signs and symptoms produced by them. Dandy’s illustrative drawings are classics. Since the advent of angiography, many large series of arteriovenous “aneurysms” of the brain have been published: Olivecrona and Riives in 1948 reported 60 cases; Mackenzie '53, 50 cases; Tolus and Lange-Cosack '53, 72 cases; Paterson and McKissock '56, 110 cases; Olivecrona and Ladenheim '57, 125 cases; Olivecrona and Ladenheim '57, 125 cases; and McRae '65, 95 cases. Most of the authors follow a classification derived from Bergstrand '36: (1) angioma cavernosum, and (2) angioma racemoseum, which also includes telangiectasia, Sturge-Weber disease, angioma racemoseum arte-
rioie, angioma racemoseum venosum, and aneurysma arterio venosum. The pathology and a comprehensive classification of vascular “arteriovenous” malformations was published recently by McCormick '66. The arteriovenous “aneurysms” or malformations are by far the most common and are clinically the most important. They comprise approximately 1.5 to 4 per cent of verified intracranial tumors. According to most authors, they are about twice as common in males as in females.

Kaplan et al. '61 believed that arteriovenous malformations originate as a congenital maldevelopment of the blood vessels in early embryonic existence. They believed that the underlying lesions represented a perpetuation of the primitive arteriovenous communications, a shunt which normally would be replaced by an intervening capillary network. The component structures of the vascular deformity other than the deeply imbedded sinusoids are individually normal and each responds to the altered local hemodynamic incident to the shunt. Arteries open directly into large sinusoids or alter their state and become veins without an intervening capillary system. There are progressive dilatation, phlebsclerosis, and occasional rupture of the veins. The sinusoids lie within or below the cortex. The malformation has a wedge-shaped configuration with the base at the cortex and the apex at the edge of the lateral ventricle. They follow, thus, a pattern of the transcerebral veins. The blood flow to and from the malformation is through enlarged, normally placed channels. These cerebral malformations are supplied by branches of the anterior, middle, or posterior cerebral arteries alone or in combination. Many of the supratentorial

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malformations receive blood supply from both carotid arteries and occasionally also from the vertebral-basilar system.

These malformations have been described as equally common in the right and left cerebral hemispheres and are most frequent in the parietal region. They may involve the corpus callosum, the structures of the posterior fossa, the ventricles, the dura, the tentorium (Laine et al. '63), the cranium, and the scalp alone or in combination.

The most common symptoms described are convulsions, hemorrhage, headaches, progressive neurologic deficit, and mental deterioration. A bruit or systolic murmur was reported in the majority of cases when branches of the external carotid artery contributed to the blood supply of the malformation. The first symptoms usually occurred during the second or third decade of life; when hemorrhage occurred, it was more frequently between the ages of 10 and 40.

In the 125 cases of Olivercrona and Ladenheim '57, intracranial hemorrhage occurred in 48 (38 per cent). Hemorrhage was the initial symptom in 42 per cent of the 110 cases reported by Paterson and McKissock '56. Sixty-four (64) per cent of their patients had large “angiomas” while the malformation was small in 36 per cent. The small ones bled as frequently as the large ones. Krayenbühl and Siebenmann '65, analyzing 24 cases of spontaneous intracerebral hemorrhages resulting from small vascular malformations, reported that half of their patients bled during the first three decades of life. In the report of 134 cases by Tönnis et al. '58, subarachnoid hemorrhage occurred in 33 per cent, 17 per cent had seizures and hemorrhage. Recurrent hemorrhage occurred in six per cent of patients, most of them after intervals of several years. The longest interval between two hemorrhagic episodes was 28 years.

Studies of the cerebral blood circulation in the presence of an arteriovenous malformation (Tönnis and Lange-Cosauck '53), showed that the circulating cerebral blood volume was reduced approximately 50 per cent after extirpation of the angioma. The circulation was also diminished immediately after an intracerebral hemorrhage but not changed after ligation of the external or common carotid artery.

Treatment of arteriovenous malformations consisted of roentgen therapy, ligation of the carotid artery with or without roentgen therapy, ligation of the local arteries feeding the malformation, excision of the malformation, and artificial embolization of the malformation (Luessenhop et al. '65).

Among the 16 cases reported by Cushing and Bailey '28 and the eight cases of Dandy '28, no successful radical removal of the lesion was reported. Pilcher '46 believed that radical extirpation of the malformation should be undertaken if the lesion is located in a suitable area and is the cause of severe and progressive symptoms. The majority of patients who were in poor condition, usually resulting from hemorrhage, were rarely treated surgically. In the series reported by Pool '62, 24 patients were treated medically: one died and two had poor results. There were also 24 patients treated surgically, either by excision of the malformation, by coagulation or clipping of the surface vessels, or by carotid ligation; four died and nine had poor results. In the majority of patients, most authors agree that carotid occlusion in the neck or occlusion of the feeding vessels intracranially and x-ray therapy have not proved effective. Svien and McRae '65 suggested that conservative management was appropriate for 80 to 85 per cent of patients with arteriovenous malformations.

Of the 68 conservatively managed cases reported by Svien and McRae '65, five died of hemorrhage and six of other related causes. Sixty-eight (68) per cent led normal lives, twenty-two (22) per cent were self-supporting despite their symptoms, and ten (10) per cent were invalids. They estimated that hemorrhage carried a six per cent mortality and that the risk of dying from a subsequent hemorrhage after recovery from initial hemorrhage was also six per cent.

Aneurysmal dilatation of the great vein of Galen associated with other midline cerebral arteriovenous anomalies was described by Dandy '28, Litvak et al. '60, and others. These occur often in infants but have also been seen at all ages.

The association of cerebral arteriovenous malformations with intracranial aneurysms has been reported by Walsh and King '42, Arieti and Gray '44, Aring '45, Boyd-Wilson '59, and others. Most of these patients had