Arteriovenous malformations in the brain are arterial-to-venous shunts formed by abnormal, often tortuous blood vessels. These tangles of blood vessels occur in 0.5% of the general population. There is a higher incidence of arteriovenous malformations in patients who have angioplastic syndromes, including Ostler-Weber-Rondu syndrome, hereditary hemorrhagic telangiectasia, and Wyburn-Mason syndrome, and in first-degree relatives whereby there is a familial occurrence. The incidence of multiplicity in familial arteriovenous malformations is also higher: 5.6% versus 1.9% in nonfamilial arteriovenous malformations. Imaging of asymptomatic family members may detect previously undiagnosed arteriovenous malformations.

CO-EXISTENCE OF SACCULAR ANEURYSMS

The frequency of the co-existence of saccular aneurysms ranges from 3.7% to 8.7%. That the aneurysms form on arteriovenous malformations feeding vessels speaks for a causal role for the increased blood flow through the arteriovenous malformation. Other pathogenetic factors include enlargement of the feeding arteries, higher flow velocities, increased turbulence and hydrostatic pressure in the feeding arteries, and decreased resistance in these arteries. Hemodynamic abnormalities in the arteries leading to an arteriovenous malformation may predispose...
the vessels to aneurysm formation in the arteriovenous malformation feeding systems.

Actuarial analysis has demonstrated the risk for intracranial hemorrhage in patients who have a co-existing aneurysm and arteriovenous malformation to be 7% per year at 5 years after diagnosis, as compared with 1.7% for patients who only have an arteriovenous malformation. There is a significant difference between the groups in terms of survival without intracranial hemorrhage. The higher risk for hemorrhage when an arteriovenous malformation and an aneurysm co-exist, as compared with only an arteriovenous malformation, is probably secondary to rupture from the co-existing aneurysm.

GENETICS OF INTRACRANIAL ANEURYSMS

Intracranial aneurysms are associated with certain disease states. Most notable and accounting for 5% of cases are the heritable connective tissue disorders: Ehlers-Danlos syndrome type IV, Marfan syndrome, neurofibromatosis type I, and autosomal dominant polycystic kidney disease. Familial intracranial aneurysms represent 7% to 20% of patients who have aneurysmal subarachnoid hemorrhage without associated heritable connective tissue disorders. First-degree relatives of patients who have aneurysmal subarachnoid hemorrhage are at a fourfold increased risk for having a ruptured intracranial aneurysm, compared with the general population.

Families that have two or more affected members were advised to consider screening for asymptomatic intracranial aneurysms with MR angiography followed by digital subtraction angiography if the MR angiogram was positive. A recent study, however, indicates that screening is not an effective way of reducing morbidity and mortality from ruptured intracranial aneurysms in people who have two or more first-degree relatives with ruptured intracranial aneurysms, unless the suspected incidence of asymptomatic aneurysms is considerably greater than 10%. Individuals considering screening for familial aneurysm who have a history of no more than two first-degree relatives with ruptured intracranial aneurysms are reassured that the small chance of bleeding from an aneurysm and the risk from treatment do not warrant screening.

OCCURRENCE OF ARTERIOVENOUS MALFORMATIONS

Supratentorial arteriovenous malformations are more common than cerebellar and brain stem arteriovenous malformations in all age groups, accounting for 94% of arteriovenous malformations in adults. Infratentorial arteriovenous malformations constitute 7% of cases unselected for age. Pial arteriovenous malformations are the most common symptomatic vascular abnormality of the posterior fossa, comprising
70% of a series of all age groups. Although supratentorial arteriovenous malformations predominate in children, infratentorial arteriovenous malformations occur more commonly than in adults and have a high mortality rate (35%), especially the cerebellar arteriovenous malformations.

**RECURRENT OF ARTERIOVENOUS MALFORMATIONS**

Complete surgical excision of arteriovenous malformations, as documented by postoperative angiography, is believed to eliminate the risk for subsequent rebleeding. There have been reports, however, of pediatric patients and an adult whose cerebral arteriovenous malformations recurred in or adjacent to the site of the original lesion after negative postoperative angiograms. Actual regrowth of the arteriovenous malformation may occur in children because of their immature cerebral vasculature, and may involve angiogenesis mediated by humoral factors, such as vascular endothelial growth factor.

Although the risk for recurrence after a negative postoperative angiogram is low, follow-up imaging studies should be considered in children at least 1 year after their initial negative postoperative angiogram to exclude a recurrent arteriovenous malformation—especially if they are planning to undergo a diagnostic or surgical procedure, because the morbidity and mortality from recurrent hemorrhage are high. Routine delayed angiography in an adult after initial negative postoperative angiogram is not recommended. These patients are followed clinically; CT scanning or MR imaging and angiography are performed if symptoms and signs of regrowth of the arteriovenous malformation appear.

**THE SPETZLER-MARTIN SYSTEM**

The Spetzler-Martin System for grading cerebral arteriovenous malformations is a scale that correlates well with the morbidity from the therapeutic modalities for arteriovenous malformations: microsurgery, embolization, and radiosurgery. There is also good interobserver correlation. The system includes the following criteria:

- Maximum diameter of the arteriovenous malformation (1–3 points)
- Presence of deep venous drainage (0 or 1)
- The proximity of the arteriovenous malformation to eloquent areas of the brain (0 or 1)

**PRESENTATION OF ARTERIOVENOUS MALFORMATIONS**

The most common presentation of patients who have intracranial arteriovenous malformations is intracranial hemorrhage, which occurs...
in 20% to 50% of cases. The severity of clinical presentation is related directly to the size of the acute hematoma, which, itself, is a reasonable predictor of outcome. The mortality rate for patients who suffer an initial hemorrhage is 10% to 29%.

In children, seizures are the second most frequently presenting symptom, occurring in 15% to 20% of cases. Other significant presentations include headache, progressive neurologic deficit, pulsatile tinnitus, and unrelated complaints leading to a fortuitous diagnosis. The risk for hemorrhage among those who present with conditions other than an initial hemorrhage is 2.2% per year.

TREATMENT MODALITIES

The modalities available for treatment of cerebral arteriovenous malformations include microsurgery with or without endoscopic assistance; embolization; and stereotactic radiosurgery with heavy particles (e.g., protons, helium ions), cobalt (γ knife), or x-radiation (linear accelerator), alone or in combination. Although operative extirpation is desirable, surgery may not be possible for arteriovenous malformations in inaccessible or eloquent areas of the brain or for patients in poor medical condition. Similarly, although radiosurgery is effective for obliterating small arteriovenous malformations, the technique is limited by a decrease in obliteration and an increase in radiation-induced complications for larger arteriovenous malformations and the chance of hemorrhage during the latent period before complete obliteration has occurred.

Embolization has been used to reduce the size of the arteriovenous malformations before microsurgery or radiosurgery. The residual nidus then can be irradiated with results almost comparable with radiosurgery alone for an arteriovenous malformation of the same size. Because conservatively treated arteriovenous malformations, especially the deep-seated ones, are more likely to bleed—with an increase in patient mortality, the multimodal approach has improved clinical outcome by facilitating the successful treatment of difficult cases.

ORDER OF TREATMENT

Aneurysms may rupture before, during, and immediately after treatment of an arteriovenous malformation, complicating the management of patients with arteriovenous malformations and associated intracranial aneurysms. To reduce the risk for intracranial hemorrhage, more likely from the aneurysm than from the arteriovenous malformation, the aneurysm is treated by surgical or endovascular means before definitive therapy for the arteriovenous malformation is undertaken. Close attention to intraoperative blood pressure control and fluid management during surgery to secure the aneurysm is essential to avoid hemorrhage from the arteriovenous malformation.
INTERVENTION

Microsurgery is recommended for smaller, surgically accessible arteriovenous malformations. This is superior to radiosurgery because of the reduced risk for hemorrhage, as may occur during the latent period after radiosurgery, with reduced mortality and permanent neurologic morbidity. Surgery for arteriovenous malformations is delayed until patients have recovered from the effects of the initial intracranial hemorrhage. If the patient has bled from an associated aneurysm, however, early surgery is recommended to secure the aneurysm, with appropriate attention to prevention of hemorrhage from the arteriovenous malformation. Immediate operation is also necessary if the hematoma from the arteriovenous malformation is causing clinical deterioration because of increased intracranial pressure.

ANESTHETIC MANAGEMENT

Whether therapeutic intervention is immediate or delayed, all patients require definitive diagnosis by CT scan, MR imaging, and cerebral angiography after initial evaluation of their condition. Older children who are alert and oriented may be able to cooperate, but younger, disoriented, and comatose children require general anesthesia for airway protection and optimization of the neurodiagnostic studies.

The induction and maintenance of anesthesia in children who have arteriovenous malformations with or without associated aneurysms are predicated on avoidance of further intracerebral hemorrhage and improvement of intracranial compliance, especially because the location and size of the hematoma are not known precisely before the imaging studies are performed. It must be assumed that the patients have full stomachs.

Intravenous access is secured before the induction of anesthesia. Preoxygenation is followed by the intravenous sequence of pentothal, 4 mg/kg, or propofol, 2 mg/kg (i.e., if blood pressure permits), rocuronium, 2 mg/kg, and fentanyl, 2 to 3 μg/kg. The dose of sedative-hypnotic and narcotic is reduced if the patient is hypotensive. With the advent of short-acting, nondepolarizing muscle relaxants, the airway may be secured expeditiously without the use of succinylcholine, which may elevate intracranial pressure.

Anesthesia is maintained with an intravenous infusion of propofol and minimal inhalational anesthesia. Ventilation is controlled to achieve moderate hypocapnia. Spontaneous ventilation with the possibility of hypercarbia may be detrimental. Even when the patient seems neurologically intact on initial evaluation, the degree of intracranial elastance is unknown before the brain is visualized. A small increase in intracranial volume from any increase in cerebral blood flow may cause herniation.

The CT and angiography suites can accommodate all anesthesia monitoring, delivery, and ventilation equipment. The MR suite requires
nonferromagnetic equipment. Anesthesia machines and monitoring equipment are available that are compatible with the MR scanners. In the absence of MR-compatible machines, induction and maintenance of anesthesia are totally intravenous.

**SUMMARY**

Intracranial arteriovenous malformations can occur singly, multiply, and in conjunction with aneurysms and denovo, famially, or in conjunction with connective-tissue disorders. Intracranial hemorrhage is the most common presentation, occurring in 20% to 50% of cases. In children, seizures are the second most common presentation occurring in 15% to 20% of cases. The modalities available treatment of arteriovenous malformations are microsurgery, embolization, and stereotactic radiosurgery with heavy particles, α knife, or linear accelerator. Induction, maintenance, and emergence from anesthesia are designed to prevent rupture of arteriovenous malformation and aneurysm and to improve intracranial compliance in the presence of an intracranial hematoma, during both diagnostic (CT, MR scanning) and therapeutic procedures.

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