General Principles of AVM Management

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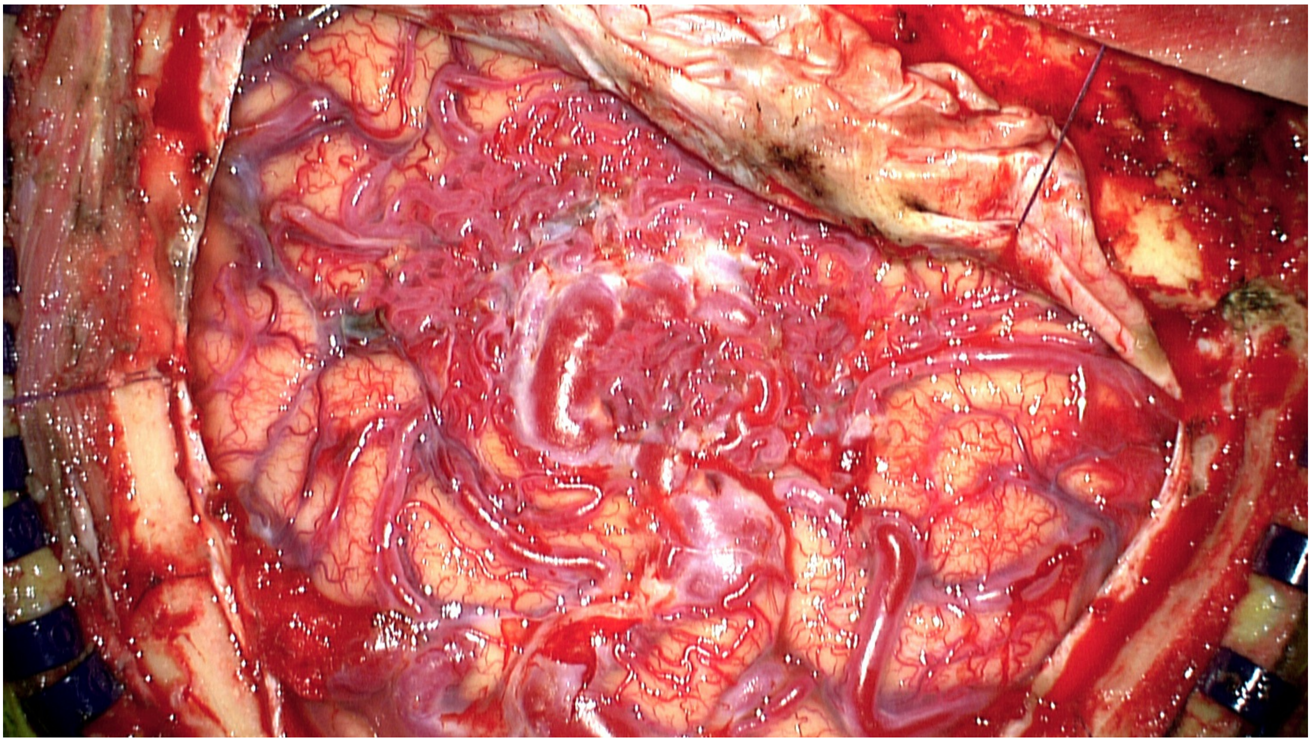

Figure 1: A giant (>6cm) right temporoparietal AVM is exposed.

The appropriate treatment modality for arteriovenous malformations (AVMs), like any other pathology, should offer results that are better than the natural course of the disease. This is especially true for unruptured AVMs, which have been encountered more frequently in recent years because of the high frequency of magnetic resonance imaging (MRI) studies performed for a variety of other symptoms.

Brain arteriovenous malformations present a unique technical challenge in their diagnosis, evaluation and management. Despite advances in endovascular, radiosurgical, and microsurgical treatment modalities, these vascular lesions are a heterogeneous pathological entity whose behavior is often difficult to predict without intervention. The goal of AVM treatment is to alleviate the risk of future hemorrhage without incurring treatment-related morbidity. Microsurgical resection remains the treatment of choice for carefully selected AVMs because it immediately and definitively excludes the lesion.

In cases where microsurgical resection is accompanied by a high risk of morbidity, radiosurgery is a desirable option; however, radiosurgical cure takes years during which a risk of rupture persists. Endovascular treatment is available in many centers, but cure is difficult to achieve through this modality alone, so it is most useful as a microsurgical or radiosurgical adjunct.

In this chapter, I first briefly review the natural course of AVMs and their surgical outcome. I will then discuss classifications of AVMs and the decision-making process for rendering suitable treatment. The results of recent trials have added controversy to this process.

Diagnosis and Evaluation

Most patients present for neurosurgical evaluation after an acute hemorrhage or after an AVM was found on imaging during the work-up of other complaints such as seizures, focal neurologic deficit, or headaches. Increasingly, AVMs are found incidentally on imaging obtained for other unrelated reasons.

Once an AVM is identified, all patients receive a thorough preoperative evaluation including history and neurologic exam. Additional imaging is obtained as needed so each patient has a CT, an MRI, and a catheter angiogram. The most important imaging modality for thorough evaluation of an AVM is a catheter angiogram, which contains a wealth of information about the anatomy and hemodynamics of a given lesion.

Natural Course of AVMs

Risk of Bleeding

In general, the annual risk of hemorrhage from an AVM is approximately 2-4%. However, this rate is not applicable to all AVMs because some AVM’s harbor anatomic and flow-related features that prone them to more likelihood of hemorrhage.

Two main factors increase the likelihood of AVM bleeding: 1) recent history of hemorrhage, and 2) presence of an intranidal aneurysm(s) or venous stenosis/stasis. The annual bleeding risk is 7% during the first 2 years after the initial hemorrhage and this rate then declines steadily to reach 3% in 5 years and afterwards.

There are other potential risk factors that increase the chance of bleeding from an AVM, namely single deep venous drainage, deep location, and the patient’s age.

The long-term risk of bleeding may change over time due to dynamic alterations in the angioarchitecture of the AVM. Therefore, neither of the above factors may reliably predict the long-term outcome.

It goes without saying that the longer the patient’s life expectancy, the higher the chance of AVM bleeding. Therefore, the physician has more reason to propose treatment to younger patients with minimal comorbidity and a longer life expectancy.

Assuming an annual hemorrhage rate of 2% to 4% and an average life expectancy of 70 years, the cumulative risk for AVM rupture can be calculated by the following formula: 105 minus the patient's age in years.

Pregnancy is most likely not associated with an increased the risk of AVMs’ hemorrhagic transformation. The method of delivery should be primarily based on obstetric principles. Additionally, size and the risk of hemorrhage have not been conclusively correlated. Whether smaller AVMs lead to an increased risk of rupture is controversial.

There are certain limitations regarding the available natural history studies. First, most studied AVMs were identified because they were symptomatic (about half of them were due to hemorrhage and one-quarter due to seizures). Therefore, the applicability of these studies to the increasing number of truly incidental AVMs discovered via the increased use of neuroimaging is limited and their results may overestimate the risk of hemorrhage for truly asymptomatic lesions. Second, patients who appear to be at higher risk because of the presence of risk factors (e.g., associated aneurysms) would be disproportionately treated compared with patients who have less threatening lesions; this fact would, conversely, underestimate the risk of observation.

Risks of Morbidity and Mortality

The initial hemorrhage has a 15% chance of causing death, whereas subsequent hemorrhages lead to major disability or death in about 40% to 50% of patients. This results in an almost 1% annual rate of mortality for all patients.

Although major morbidities are usually due to a hemorrhagic event, there is a 1.5% annual incidence of functional decline because of seizure activity or progressive neurologic deficits caused by venous hypertension or arterial hypoperfusion (“steal” phenomenon) in the neighboring cerebral tissues.

Treatment Considerations

Location (vicinity of indispensable brain,) size, location and pattern of arterial supply, presence of sizable deep white matter perforators, location and pattern of venous drainage, evidence of outflow obstruction, associated nidal aneurysms, evidence of “steal” phenomenon, AVM configuration (compact versus diffuse) are all factors that must be considered in determining the feasibility of surgery.

For proper patient selection, the risks of microsurgical resection or radiosurgery alone should not outweigh the risks involved in the AVMs’ natural history.

Three major criteria are used with Spetzler-Martin classification, namely size of the AVM, the presence of deep venous drainage, and the eloquence of the AVM’s location. These are all considered predictors of surgical outcome. Other important risk factors include the presence of robust lenticulostriate feeders and deep meningeal supply, as well as the diffuseness of the nidus.

Table 1: Spetzler-Martin Arteriovenous Malformation

Grading System

AVM grading scale predictive of operative outcome (Grades 1-6)

	0	1	2	3
Size of Nidus	-	Small (<3cm)	Medium (3-6cm)	Large (>6cm)
Eloquence of Adjacent Brain	Non-eloquent	Eloquent	-	-
	• Eloquent = sensorimotor, language, visual cortex, hypothalamus, thalamus, brain stem, cerebellar nuclei, or adjacent regions • Non-eloquent = frontal lobe, temporal lobe, cerebellar hemispheres			
Venous Drainage	Superficial only	Deep	-	-
Summation of scores (range 1-5) = Grade (falls into 1 of 6 grades) Note: Grade 6 = inoperable				

These are all angiographically-based criteria that emphasize the importance of careful scrutiny of preoperative imaging, including angiography for operative planning. However, some of these criteria may not be as effective as we once thought. For example, the presence of deep venous drainage does not increase the risk of resection, and in fact it may ease the technical challenges of preserving the draining vein during the early stages of the operation and AVM disconnection.

However, the diffuseness of the AVM and predominance of deep white matter feeders, especially lenticulostriate arteries, are most likely to be as or more important than the size of the AVM.

Decision making Process for Treatment

There are three main treatment options available for managing newly diagnosed AVMs. These options include, microsurgery (with and without embolization,) radiosurgery (with or without embolization) or observation.

Embolization

Embolization can rarely cure small AVMs with a single pedicle and should not be used indiscriminately as the hemorrhagic risk of overembolization without reaching complete cure can be significant. Embolization is used if microsurgical resection is contemplated in cases where some of the feeders may not be readily available to devascularization during the early parts of the surgery.

Palliative embolization is indicated for the treatment of high-grade lesions deserving of no other treatment because of the attended risks. The progressive symptoms that could be related to steal phenomenon (intractable headaches, seizures and fluctuating neurologic deficits) may be tapered via selective pedicle obliteration. The hemorrhagic risk of the AVM is not altered and inadvertent partial nidal occlusion may in fact increase the risk of rupture through hemodynamic alteration.

Coil embolization of deeding or pedicle aneurysms is indicated, especially for unresectable high-grade AVMs. These aneurysms are often situated in remote and inaccessible locations complicated by presence of the AVM and its draining veins. Resection of the AVM alone will lead to involution of the aneurysm and its potential disappearance. The presence of subarachnoid hemorrhage in the absence of intracerebral clot should increase the suspicion of aneurysmal subarachnoid hemorrhage.

Radiosurgery

Radiosurgery is used for surgical high-risk/inaccessible small to medium size AVMs in the brainstem, basal ganglia and thalamus. It can also be used as an adjunct to “downgrade” an AVM so that the lesion is more amenable to resection. I have occasionally used radiosurgery to treat the high-risk portions of the AVM (deep white matter feeders in close proximity of deep structures) in preparation for complete removal of the malformation in a delayed fashion.

Hypofractionated and volume-staged radiosurgery are generally reserved for AVMs>3cm or volumes >10cc. The rates of obliteration are substantially lower in the larger AVMs. Embolization followed by radiosurgery is associated with a risk of radiosurgical failure with the large AVM size as the confounding factor for such a failure. The latency period of radiosurgery allows recanalization of the partially embolized AVM. Therefore, embolization followed by radiosurgery is not an effective option for AVM cure.

Suggested Classifications

Various classifications of AVMs with their corresponding prognostic values have been proposed, but so far, no classification by itself accurately predicts the risk of microsurgical resection.

Spetzler-Martin grading, mentioned above, is most commonly used in practice. In 2011, this grading system was simplified into three grading classes with different risk-prediction values and proposed treatment options.

Table 2: Modified Spetzler-Martin Grading System

Spetzler-Martin Class	Spetzler-Martin Grade	Management
A	I & II	Surgical resection
B	III	Multimodality treatment
C	IV & V	Conservative

Microsurgery is the suitable treatment modality for Class A lesions. Conservative approach is recommended for AVMs categorized as Class C unless the patient experiences progressive neurologic deficit or repeated hemorrhagic attacks.

Management of Class B AVMs, which truly demand a good classification system, is said to be individualized based on the following factors: size of the lesion, its eloquent location, history of hemorrhage, presence of intranidal or feeding aneurysms, and the diffuseness of the nidus.

A multimodality treatment paradigm, such as microsurgery, radiosurgery, embolization, or a combined approach, is probably most appropriate to tackle Class B AVMs, whereas conservative management is a viable option for a subset of surgically high-risk patients.

Lawton introduced a supplementary classification scheme in 2010 that assigned scores from 1 to 5 based on patient's age, ruptured presentation, and diffuseness of the nidus (Table 2). He proved this classification to be a more reliable predictor of outcome than the conventional Spetzler-Martin grading system in cases when the two classifications contradict each other.

On the other hand, the scores obtained from both classifications may be added to make a new score ranging from 2 to 10; in this system, patients with a total score of less than 6 are considered to have an acceptable surgical risk, and patients with a total score of 4 or less harbor minimal risk.

Table 3: Supplementary Classification by Lawton and Young

Variable	Definition	Points
Patient age (years)	<20	1
	20-40	2
	>40	3
Unruptured presentation	No	0
	Yes	1
Diffuse	No	0
	Yes	1

Management of AVMs after the ARUBA Study

A Randomized Trial of Unruptured Brain Arteriovenous Malformations (ARUBA) study was prematurely completed recently and showed no advantage for intervention (radiosurgery, embolization, or surgery) versus medical treatment for unruptured AVMs.

The study ignited many critics who believed the trial was riddled with methodological errors and that the follow-up period was too brief to allow valid conclusions. Because of its shortcomings and limitations, this trial's results should be interpreted with caution and most likely should not significantly affect our treatment decisions for unruptured AVMs.

According to the generally accepted consensus pertaining to this topic, I recommend treatment for all small and moderate size AVMs that have bled. I also recommend treatment of symptomatic AVMs to prevent progression of seizures and neurologic deficits. Unruptured asymptomatic small and moderate size AVMs in non-eloquent territories should also undergo resection because the lifetime risk of hemorrhage and development of focal deficits and seizures, especially in younger patients, is significant compared with the risk of microsurgical excision.

Large AVMs in the proximity of eloquent locations require an individualized treatment approach based on the diffuseness and the precise proximity of the AVM to the functional cortices and white matter tracts. Unfortunately, there will likely never be a reliable grading system to handle these challenging and controversial lesions. The experience of the AVM surgeon plays an important role under these circumstances. Radiosurgery followed by microsurgery has been proposed as a potential alternative treatment strategy for surgically high-risk AVMs.

The efficacy of endovascular embolization as the sole modality for AVM obliteration is questionable, and long-term follow-up information for this practice is not available at this time.

I do not routinely use embolization in preparation for microsurgical resection. I believe nonselective pedicle embolization leads to expansion of white matter feeders. These feeders add significant technical complexity to the procedure. Instead, I request selective embolization of surgically inaccessible feeding arteries in expectation of microsurgical excision of the nidus.

Table 4: Management Options and Factors Favoring Each Treatment Approach

	Microsurgery with and without adjunctive embolization	Radiosurgery	Observation
Patient age	Younger	Older	Older
Presentation	Previous hemorrhage neurologic deficit seizure	Asymptomatic	Asymptomatic
Angiography architecture	Associated aneurysm	Compact nidus	
Size or Spetzler-Martin grade	Small or low grade more favorable	Small	High grade
Location	Noneloquent	Eloquent	Eloquent

Operative Considerations

Instrumentation

The operation is performed with the surgeon in the sitting position using an armrest. Please refer to the chapter on Surgeon's Philosophy and Operating Position for further details. The surgeon's comfort prevents fatigue and provides smooth microsurgical dissection maneuvers during the critical portions of the case. These critical steps often occur later in the case when fatigue can be an important factor.

The initial dissection may be performed with loupes to prevent working into a deep hole, but as the dissection proceeds deeper, the microscope is used with the mouthpiece to minimize redundant and nondeliberate movements such as using the microscope handles to refocus the microscope. The mouthpiece is critical for microsurgical efficiency. Monitors showing the oculars' view of the surgical field should be available to the other members of the operating room staff to allow them to follow the flow of the procedure and inform them regarding the dissecting instruments needed for the next step.

Irrigating bipolar forceps of various lengths and tips are the most important tool for AVM dissection. The irrigation and coagulation levels of this device must be appropriately balanced to prevent accumulation of char on the tips. The tips of these forceps may be kept in cold saline solution to minimize charring at their tips during their use.

Next, a variety of dissecting instruments are needed. The width of the tips of dissectors should be matched to the level of microdissection. The angled dissectors provide more flexibility in working around the corners as along as all the dissection is performed under direct visualization. Blunt and blind dissection is the cause of significant morbidity because of perforator injuries and unintended parenchymal transgressions. Small aneurysm clips may be used to control bleeding from white matter feeders that are not amenable to bipolar coagulation.

Anesthesia Considerations

The administration of general anesthesia and intraoperative monitoring of physiologic parameters during AVM surgery must be a team effort with open communication among the members of the surgical team.

Standard noninvasive monitoring is used along with direct arterial pressure monitoring to ensure that the patient’s blood pressure is aggressively treated and maintained within designated parameters. Central venous access is used for larger lesions to ensure rapid restoration of fluid and blood volume if necessary.

Hemodynamic stability, cerebral perfusion, and management of intracranial pressure are important for maximizing patient outcomes. Mild intraoperative hypotension can be used to decrease blood loss and facilitate surgical resection; however, this maneuver should be used with caution as profound hypotension can cause ischemia in parenchyma already subject to hypoperfusion due to arteriovenous shunting.

Emergence from the anesthesia can often be associated with coughing and bucking, causing periodic dramatic increases in blood and intracranial pressures, leading to intracerebral rebleeding within friable territories. Therefore, following difficult operations associated with high blood loss, large AVMs, or any other concern for postoperative bleeding, the patient remains intubated and sedated overnight to avoid these potential complications.

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