Management of arteriovenous malformations: A surgical perspective

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The management strategies for arteriovenous malformations (AVMs) continue to evolve, spurred by advancing technology and improved understanding of the natural history of these lesions. In general, intervention is reserved for Spetzler-Martin Grade I-III lesions or for those with certain high-risk features. Grade IV-V AVMs, in contrast, are usually managed conservatively. Although multimodality therapy incorporating endovascular and/or radiosurgical techniques is increasingly common, microsurgical removal remains the definitive form of treatment.

Key Words: Arteriovenous malformation, endovascular, radiosurgery, Spetzler-Martin Grade

The risk of a chosen management strategy must include the sum of the risk of all the interventions. Arteriovenous malformations (AVMs) represent some of the most complex lesions that neurosurgeons confront. The management principles outlined below have evolved from more than three decades of experience on the part of the senior author. We use a multidisciplinary approach for both the evaluation and treatment of these lesions, as any given lesion may need to be treated with some combination of surgery, embolization, and/or stereotactic irradiation. The risk of a chosen management strategy must include the sum of the risk all the interventions in the treatment plan, including diagnostic angiography, embolization, radiosurgery, and/or microsurgery.

Grading of arteriovenous malformations

The Spetzler-Martin grading system is a simple yet comprehensive scheme that allows evaluation of the salient anatomic characteristics of AVMs with regard to natural history and the morbidity and mortality rates associated with treatment. AVMs are defined by three variables: size, neuroanatomical eloquence of surrounding brain tissue, and the pattern of venous drainage. A simple summation of assigned point values yields the grade of the lesion (Table 1).

Size determines much of the surgical difficulty that a given AVM represents. Lesions are graded as small (0 to 3 cm – 1 point), medium (3 to 6 cm – 2 points), or large (greater than 6 cm – 3 points). AVM location is divided into eloquent (1 point) and non-eloquent (0 points) regions. Eloquent brain includes sensorimotor, language, and visual cortex; the hypothalamus; thalamus; internal capsule; brainstem; deep cerebellar nuclei; and cerebellar peduncles. The pattern of venous drainage affects surgical accessibility and is divided into superficial (0 points) and deep (1 point). Only cortical venous drainage and cerebellar venous drainage into the straight or transverse sinuses are considered to be superficial.

Table 1: Spetzler-Martin grading system*

<table>
<thead>
<tr>
<th>Feature</th>
<th>Points</th>
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<tbody>
<tr>
<td>Size</td>
<td></td>
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<tr>
<td>Small (&lt;3 cm)</td>
<td>1</td>
</tr>
<tr>
<td>Medium (3-6 cm)</td>
<td>2</td>
</tr>
<tr>
<td>Large (&gt;6 cm)</td>
<td>3</td>
</tr>
<tr>
<td>Eloquence of Adjacent Brain</td>
<td></td>
</tr>
<tr>
<td>Non-eloquent</td>
<td>0</td>
</tr>
<tr>
<td>Eloquent</td>
<td>1</td>
</tr>
<tr>
<td>Pattern of Venous Drainage</td>
<td></td>
</tr>
<tr>
<td>Superficial</td>
<td>0</td>
</tr>
<tr>
<td>Deep</td>
<td>1</td>
</tr>
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* Grade VI denotes a truly unresectable untreatable AVM, (e.g., holohemispheric or involving a substantial amount of brainstem)
Endovascular and radiotherapy

Endovascular therapy and radiosurgery have made welcome adjuncts to the therapeutic arsenal available for treating this disease. These technologies continue to improve steadily. Preoperative endovascular embolization of feeding arteries has rendered many previously difficult AVMs much easier to remove surgically, decreasing operating time and blood loss. In general, endovascular embolization is most useful for Grade III AVMs. If endovascular access to the feeding vessels can be achieved without placing normal vessels at substantial risk, we often use it for lower-grade AVMs as well. As a stand-alone therapy, endovascular embolization is not definitive treatment. The rate of complete occlusion associated with endovascular therapy alone is estimated at 10%,[10] and the durability of occlusion in this setting is unknown.

In contrast, stereotactic radiosurgery can produce an angiographic and clinical cure for many lesions. Optimally, the lesion should be small (<10 cc total volume). The efficacy of radiosurgical obliteration of lesions less than 2.5 cm in diameter ranges from 74 to 80% to about 50% for lesions between 2.5 and 3 cm in diameter.[16] The full effect of radiosurgery does not become apparent for at least 2 years. Many centers are now aggressively recommending radiosurgery for the treatment of AVMs. We recommend surgery for most accessible lesions and reserve radiosurgery for lesions in locations where the morbidity associated with the surgical approach is high, such as in the basal ganglia or thalamus. As part of multimodality treatment, radiosurgery can be an important adjunct to surgery. After endovascular embolization, surgery, or both have reduced the size of the nidus of a difficult AVM as much as possible, radiotherapy can be used to treat the remnant definitively. As of yet, the benefit of this strategy has not been substantiated. For large lesions (>3 cm in diameter), surgery typically offers the most realistic hope of cure.

Management of high-grade AVMs

Although some neurosurgeons still advocate an aggressive approach,[7,8] a trend toward conservative management of high-grade AVMs (Spetzler-Martin Grades IV and V) has been increasing. Substantial risk is associated with the microsurgical removal of these lesions. Although the possibility of achieving a morbidity-free surgical cure is attractive, we believe a critical analysis of the risk-benefit ratio for a large number of patients does not support the routine treatment of Grade IV and V lesions.[11] The natural history of these AVMs predicts a better outcome than that which can be achieved surgically. Incomplete treatment of an AVM appears to increase rather than decrease the risk of hemorrhage.[11] The only indication for partial treatment of a high-grade AVM is to ameliorate the symptoms of vascular steal in a patient with a lesion that turns to 3% per year thereafter. Grade IV and V AVMs appear to have a lower rate of hemorrhage, which is estimated at 1.5% per year.[1] The predicted lifetime risk of hemorrhage is equal to 1 – (1 - annual hemorrhage risk)n, where n = the remaining years of life expected. This risk is weighed against the risk of surgery as predicted by the Spetzler-Martin Grading scale. Clearly, the younger a patient is and the lower the surgical risk, the more likely the patient is to benefit from treatment.

Rationale for specific therapy

The indications for surgical treatment of AVMs of the brain continue to evolve. Given the relatively high rate of hemorrhage associated with AVMs without treatment and the prospect of cure with treatment, their obliteration is usually desirable. Microsurgical removal remains the mainstay of definitive treatment. Nonetheless, given the advances in endovascular therapy and radiosurgery, all options must be considered when a therapeutic plan is formulated. Endovascular therapy rarely achieves a cure alone, but it is a valuable adjunct. In practice, two or more modalities are often used to treat any given AVM.

A number of factors are considered in the decision to treat an AVM. The most important elements are the patient’s symptoms, age, and medical condition; the size of the AVM; its location; the type of venous drainage; the history of the AVM; and the natural history of AVMs in general. In children and younger patients, we are aggressive about surgical removal in an effort to avoid irradiating these patients. Complete resection also eliminates the risks of rebleeding and vascular steal in symptomatic patients. In some cases, it can also improve headaches and seizures.

A higher risk of bleeding is associated with small AVMs; impaired venous drainage; and the presence of feeding artery, nidal, or venous aneurysms.[14] Accordingly, we favor treating most small lesions because they are both easier to remove and more likely to bleed than larger AVMs. AVMs located in eloquent brain and associated with deep venous drainage are technically more difficult to remove without complications, relatively favoring their non-surgical management. Many large lesions located in non-eloquent cortex are still reasonable candidates for surgery.

A critical analysis of outcomes has shown that no treatment is the optimal course for many patients with high-grade Spetzler-Martin AVMs.[1] Although these lesions per se represent a moderate risk based on their natural history, the risk of their treatment is still higher. In recent years, the practice at our institution has been to defer intervention on these lesions unless they have high-risk features as discussed above. If a given AVM has already hemorrhaged and caused a fixed deficit or if recurrent hemorrhages, steal, or venous hypertension is producing a stepwise decline, we favor its surgical removal.

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poses an unacceptably high risk for total microsurgical resection.

Although we have attempted to distill the principles upon which our general treatment philosophy of high-grade lesions is based, each patient must be evaluated individually. Associated aneurysms, progressive neurological deficit related to vascular steal, and repeated hemorrhages are all relative indications for surgery, even in patients with a high-grade AVM. In equivocal cases, the patient’s preference ultimately may determine the decision to pursue or forgo excision.

**Conclusion**

Surgical treatment of AVMs is favored in patients in a good medical condition with a good life expectancy who harbor small to medium-sized AVMs in anatomically accessible locations within the brain. Additional indications include vascular steal or flow-related symptoms, AVMs with a high risk of hemorrhage (such as those associated with aneurysms or venous outflow obstruction), and AVMs that have failed complete treatment with other modalities. Any post-treatment residual AVM should be considered to have at least the same risk of bleeding as a native lesion and should be treated. Both radiosurgery and endovascular embolization have important roles in the management of AVMs. The former is a viable alternative to surgical resection when small lesions are either deep or in eloquent areas. The latter is a useful adjunct in facilitating microsurgical or radiosurgical therapy. Expectant management should be pursued in the case of Spetzler-Martín Grade IV or V lesions unless high-risk features such as intranidal aneurysms or progressive clinical deterioration manifest.

**References**


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