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Recommendations for the Management of Intracranial Arteriovenous Malformations : A Statement for Healthcare Professionals From a Special Writing Group of the Stroke Council, American Stroke Association

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AHA Scientific Statement

Recommendations for the Management of Intracranial Arteriovenous Malformations

A Statement for Healthcare Professionals From a Special Writing Group of the Stroke Council, American Stroke Association

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I. Introduction

Intracranial arteriovenous malformations (AVMs) are relatively uncommon but increasingly recognized lesions that can cause serious neurological symptoms or death. Although AVMs can present with hemorrhage or seizure, since the advent of contemporary brain imaging techniques, an increasing number are detected before rupture. Over the last decade, there have been significant developments in the management of intracranial AVMs. There has been an evolution of microsurgical as well as endovascular and radiosurgical techniques to treat these lesions. As the management options have evolved, individual and combined modality treatment protocols have been developed in different institutions for the management of AVMs.

A writing group was formed by the Stroke Council of the American Stroke Association to review published data for intracranial AVMs to develop practice recommendations regarding epidemiology, natural history, potential treatment strategies, and outcomes. The reports reviewed for this synthesis were selected on the basis of study design, sample size, and relevance to a particular topic. Each report was graded according to previously defined criteria.[1,2] After review of the available literature, recommendations for current practice standards have been made according to 3 separate grades (Table 1).

By the design of this type of review, the recommendations in this report represent an overview of existing treatment protocols that may vary considerably. These guidelines were developed to serve as a basis for the development of treatment strategies for AVMs, which overall represent a fairly heterogeneous group of cerebrovascular lesions and which may demonstrate different natural histories. In addition, for brain AVMs, no level I or II data are available in the literature. Because of the heterogeneity of these lesions and their relatively infrequent occurrence, strictly defined subcategories for comparison of the efficacy of various treatment modalities is difficult. Therefore, the recommendations presented here are potentially open to a wide interpretation.

II. Epidemiology

The incidence and prevalence of intracranial vascular malformations are not known with certainty, although there are data available from autopsy series and limited population-based studies. Autopsy data suggest that there is an overall frequency of detection of AVMs in ≈4.3% of the population.[3,4] In another autopsy series, 46 AVMs were noted among 3200 brain tumor cases, for a frequency of detection of 1.4%; 12.2% of the cases were symptomatic.[5,6] Autopsy data are affected by the aggressiveness with which pathologists search for the lesions, the age and cause of death of the patient, and the presence of neurological symptoms.

Population-based data are limited regarding intracranial vascular malformations. In the Netherlands between 1980 and 1990, the annual incidence of symptomatic AVMs was 1.1 per 100 000 population.[7] In a population-based study in Olmsted County, Minnesota,[8] the detection rate was 1.1 per 100 000 for AVMs when autopsy cases were excluded and 2.1 per 100 000 for all cases. The detection rate for symptomatic cases was 1.2 per 100 000 person-years.[8] The most common type of vascular malformation detected was AVM, followed by venous malformation and cavernous malformation.

III. Diagnosis and Clinical Manifestations: Natural History of AVMs

Intracranial AVMs may be diagnosed with a variety of diagnostic imaging studies. Computed tomography (CT) without contrast has a low sensitivity, but calcification and hypointensity may be noted; enhancement is seen after contrast administration.[9] Magnetic resonance imaging (MRI) is very sensitive, showing an inhomogeneous signal void on T1- and T2-weighted sequences, commonly with hemosiderin suggesting prior hem-

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TABLE 1. Levels of Evidence in Grading of Recommendations for Treatment of Patients With Subarachnoid Hemorrhage

Levels of evidence
Level I: Data from randomized trials with low false-positive (alpha) and low false-negative (beta) errors
Level II: Data from randomized trials with high false-positive (alpha) and high false-negative (beta) errors
Level III: Data from nonrandomized concurrent cohort studies
Level IV: Data from nonrandomized cohort studies using historical controls
Level V: Data from anecdotal case series
Strength of cumulative data
Grade A: Supported by level I evidence
Grade B: Supported by level II evidence
Grade C: Supported by level III, IV, or V evidence

Data are from References 1 and 2.

orrhage.[10,11] MRI can also provide critical information detailing the localization and topography of an AVM as intervention is being considered. Magnetic resonance angiography can provide some data noninvasively, without detailing factors such as presence of intranidal or feeding artery aneurysms, comprehensive data on venous drainage patterns, or subtle AVM nidus characterization. Arteriography is the "gold standard" for defining the arterial and venous anatomy. In addition, superselective angiography can provide functional and physiological data important to clinical decision analysis. On the basis of available information, it is strongly recommended that an MRI study and a 4-vessel angiogram be obtained to delineate the anatomy of an AVM.

Intracranial AVMs are occasionally seen in the elderly but are typically diagnosed before the patient has reached the age of 40 years. More than 50% of AVMs present with intracranial hemorrhage.[12] Intracerebral hemorrhage occurs more commonly, although subarachnoid hemorrhage and intraventricular hemorrhage can occur. Severe vasospasm from AVM-related hemorrhage is distinctly uncommon, although it is occasionally noted.[13]

The next most common presentation is seizure, which occurs in ≈20% to 25% of cases.[14,15] Seizures can be either focal or generalized and may be an indicator of the location of the lesion. Other presentations include headaches in 15% of patients, focal neurological deficit in fewer than 5% of cases, and pulsatile tinnitus. In children younger than 2 years of age, presentation can include congestive heart failure, large head due to hydrocephalus, and seizures. Vascular malformation–related steal phenomena that cause focal neurological deficit by altering perfusion in the tissue in the region of the AVM are distinctly uncommon.[14,16]

The overall frequency of hemorrhage caused by vascular malformations in stroke registries indicates an ≈1% occurrence of AVM-related hemorrhage among all strokes.[17] The long-term risk of hemorrhage among people with AVMs and the outcome from this hemorrhage are controversial. There are a number of potential biases that can affect natural history studies, including selection bias, treatment-intervention bias, inconsistent follow-up, and lack of arteriography for all cases. The available natural history studies (Table 2) indicate an overall risk of initial hemorrhage of ≈2% to 3% per year.[14–16,18–22] Mortality from the first hemorrhage is between 10% and 30%, although some data suggest that the mortality rate may be lower,[23] and 10% to 20% of survivors have long-term disability.[14–16,18–24] All available natural history data are level V data.

In one study,[24] a cohort of 281 consecutive, prospectively enrolled patients was investigated to evaluate the risk for hemorrhage. Among those patients who presented with symptoms other than hemorrhage, the annual risk of hemorrhage was 2.2% (3.3% per year for men and 1.3% per year for women). The annual risk of intracranial hemorrhage among people with AVMs who present with symptoms other than hemorrhage is ≈2% to 3% per year. If one assumes an annual hemorrhage risk among people with previously unruptured AVMs of ≈2% to 4% per year, the lifetime risk of intracranial hemorrhage in a person with an AVM is approximated by the following formula[25,26]:

$$\text{Lifetime risk (\%)} = 105 - \text{the patient's age in years}$$

The risk of recurrent intracranial hemorrhage is slightly elevated for a short period of time after the first hemorrhage. In 2 studies,[18,21] the risk during the first year after initial hemorrhage was 6% and then dropped to the baseline rate, whereas in another study,[19] risk of recurrence during the first year was 17.9%. The risk of recurrent hemorrhage may be even higher in the first year after the second hemorrhage and has been reported to be 25% during that year.[21]

In a prospective study,[24] during a short mean follow-up of 8.5 months, the risk of recurrent hemorrhage was 17.8% per year after presentation with hemorrhage. In that study, only 20 patients were still being followed up who were untreated at 1 year after hemorrhage; the risk of recurrent hemorrhage was 32.9% in the first year after hemorrhage and decreased to 11.3% in subsequent years.[24] The increased rate in the first year after initial hemorrhage has not been noted consistently, however.[22]

TABLE 2. Risk of Hemorrhage in Intracranial AVMs Among Cases Presenting Without any Prior History of Intracranial Hemorrhage

Series Author	Description	Number of Cases	Follow-Up, y	Hemorrhages, n	Annual Initial Hemorrhage Rate, Crude	Annual Hemorrhage Rate, Life-Table Analysis
Brown et al[14]	Retrospective, referral series	168	8.2	31	2.3 (at 15 y)	2.2
Graf et al[18]	Retrospective, referral series	71	4.8	14	4.1	...
Crawford et al[20]	Retrospective, referral series	217	10.4	77	3.4	3
Ondra et al[22]	Retrospective, referral, population-based	160	23.7	64	1.7	...
Mast et al[24]	Prospective, consecutive, referral series	139	1.0	3	2.2	2.2

Comprehensive evaluation of a patient with an AVM includes a detailed clinical examination and radiological clarification of the anatomy with MRI scanning and arteriography. After the comprehensive evaluation has been performed, decisions can be made regarding the best management approach by comparing the natural history of the lesion with the intervention-related morbidity and mortality.

There is evidence suggesting that radiological parameters may be predictive of hemorrhage risk. A complex combination of variables may predict the risk for hemorrhage from an AVM. Some studies have noted that patients with seizures may be at slightly higher risk for hemorrhage, but this has not been noted consistently.[14,18] There are also data that suggest that prior hemorrhage is a strong predictor of hemorrhage.[23] Small AVM size in terms of maximal diameter[18,27] or volume[28] may also be a predictor for higher risk of hemorrhage; however, these are level IV data and have not been noted consistently.[14,23] Feeding artery pressures may also be related to bleeding risk.[29] AVMs in a periventricular or intraventricular location may also be at increased risk,[30,31] although this has not been found consistently,[32] and location was not found to be a risk factor in another large series.[14]

Characteristics of the venous drainage system, including presence of deep venous drainage, have been reported to be a predictor of presentation with hemorrhage[33–35] or occurrence of hemorrhage during follow-up in cases initially presenting with or without hemorrhage.[23] The angiographic characteristics of an AVM are complex. There are likely both arterial and venous factors that are predictive of an increased risk of hemorrhage, although studies are not definite. In one retrospective study (level V data), independent predictors of presentation with hemorrhage included central venous drainage, intranidal aneurysm, and periventricular or intraventricular location.[30]

In another study,[33] univariate analysis predictors of presentation with hemorrhage included deep venous drainage, arterial supply via perforators, intranidal aneurysms, multiple aneurysms, vertebrobasilar supply, and basal ganglia location. Single draining vein, impaired venous drainage, and deep venous drainage alone were factors in another study.[34] Both of the latter studies examined features retrospectively associated with hemorrhage rather than risk factors of future hemorrhage, and these studies lacked multivariate analyses. Impaired venous drainage was not an important factor in 2 other studies,[30,33] nor was a single draining vein.[33] Presence of a venous varix was also not predictive of hemorrhage.[14,33,34]

The nature of the arterial system may also be important; detection of intranidal or saccular aneurysms appears to be an important finding.[21,30,36] When selected clinical factors are combined, a profile for risk of hemorrhage may be developed. One such approach used history of prior hemorrhage, angiographic presence of a single draining vein, and diffuse AVM morphology. The lowest-risk group (risk of 1.0% per year) had no history of prior hemorrhage and >1 draining vein in a compact nidus, whereas the highest-risk group (8.9% per year) comprised those who had a prior hemorrhage, a single draining vein, and/or a diffuse nidus.[37]

Treatment Risks Versus Benefits

We have outlined the natural history of AVMs previously. A crucial question is how the natural history for a patient of a given age with a specific AVM compares with the risk of treatment. The answer to this comparison typically dictates the final recommendation of whether to treat an AVM and, if so, how to treat it (lowest risk/highest efficacy technique). In the next sections, we will discuss the various treatment modalities and recommendations for usage.

IV. Direct Surgical Treatment

Timing of Surgery

The recommendation for surgery for AVMs should generally be elective. Occasionally, one must operate emergently to remove a large, life-threatening hematoma. Under these conditions, only superficial AVMs that are readily controllable are removed with the hematoma. When the hematoma is caused by a complicated AVM, the blood clot can be removed and the patient allowed to recover until further details are known regarding the exact angiographic AVM architecture. In a nonemergent situation, the lesion is approached as are other elective intracranial operations.

Lesions are typically excised by standard microsurgical techniques with the operating microscope. The arterial feeders are generally attacked first, followed by excision of the nidus of the lesion and finally resection of the draining vein.[38,39] In general, the veins are preserved until the very end of the operation. When a brain AVM is resected, the goal should be complete obliteration. To this end, intraoperative or postoperative angiography is usually recommended. If there is residual lesion, immediate resection should be considered to avoid subsequent hemorrhage from the remaining vessels. Another treatment consideration for the residual lesion may include stereotactic radiosurgery, although there remains a risk of hemorrhage during the intervening period until lesion obliteration (see below).

Outcome of Direct Surgery

Outcome reports regarding the results of surgical excision of brain AVMs are level V data. The majority of this information is gathered in a retrospective fashion.[40,41] However, with the Spetzler-Martin grading system,[40] it is possible to estimate risks of surgery for AVM patients. For grade I patients, published reports include a high probability (92% to 100%) of favorable outcome.[40,41] For grade II levels, a 95% chance of excellent or good outcome has been reported.[40] In grade III lesions, the rate of excellent or good outcome has been reported as 68.2% in the short term and 88.6% in longer follow-up.[41] For grade IV lesions, the rate of excellent outcome drops to 73% of patients.[40] In grade V patients, the reported good/excellent rate is 57.1%, with a 14.3% rate of poor outcome and a 4.8% mortality rate in longer-term follow-up.[41] Although these results have a heavy selection bias, they also provide a framework within which to consider risks of treatment in individual patients (see below).

V. Grading Systems and Risk of Therapy

Grading schemes were initially developed as a means to predict surgical risk during obliteration. The criteria used for selecting a scheme included applicability to all AVM types and ease of use. The important variables include size, number of feeding arteries, velocity of flow through the lesion, degree of steal from surrounding brain, location (including surgical

TABLE 3. Spetzler-Martin AVM Grading Scale

Size	
0–3 cm	1
3.1–6.0 cm	2
>6 cm	3
Location	
Noneloquent	0
Eloquent	1
Deep venous drainage	
Not present	0
Present	1

accessibility), eloquence of adjacent brain, presence of associated aneurysms, and finally, the pattern of venous drainage. Grading scales have been applied both prospectively and retrospectively, and the results support their use.

The method proposed by Malik et al[42] for preoperatively grading AVMs was an anatomically based system. The authors focused on arterial supply and the number of arteries feeding supratentorial malformations. AVMs with grades I through IV were derived by use of this system, with special categories for vascular supply from lenticulostriate vessels, vessels from the choroid plexus, and the region of the corpus callosum. These authors used 2 additional factors, including a clinical grading scale and anatomic location. The system of Malik et al proved to be too complex for general use, but it did confirm that increasing grade was associated with greater surgical morbidity.

Nearly a decade later, 2 grading scales were published simultaneously.[40,43] The anatomically based system proposed by Shi and Chen[43] focused on size, location, depth, complexity of feeding arteries, and complexity of draining veins. Although the system did appear to predict surgical morbidity, it proved to be too complex for bedside use. Currently, the most commonly used grading scale is the system described by Spetzler and Martin.[40] Experience suggested that many important factors were interrelated. The authors provided a simplified scheme based on size, location, and venous drainage (Table 3). The score ranged between 1 and 5, with 1 point given for a lesion <3 cm, 2 points for a lesion from 3 to 6 cm, and 3 points for a lesion >6 cm. Location within eloquent cortex provided an additional point, as did deep venous drainage. The score was calculated by summing the points for each category. When this system was retrospectively applied by the authors, grade I and II lesions had very low morbidity and higher-grade lesions were associated with gradually increasing morbidity; however, no deaths were reported. This system was also applied by other surgeons.[41,44] Again, lower-grade lesions were associated with minimal surgical morbidity; however, grade V lesions were found to convey up to 33% permanent and serious morbidity.

The Spetzler-Martin grading scale has also been applied prospectively.[45] Lesions graded I, II, or III were found to have low treatment-associated morbidity. However, grade IV lesions conferred 31.2% treatment-associated morbidity, and grade V lesions had 50% new treatment-associated morbidity. In addition, the rate of permanent deficit was 29.9% for grade IV lesions and 16.7% for grade V lesions. This led the authors to recommend surgery for all grade I and II lesions. Grade III lesions should be treated on a case-by-case basis; however, in general, the authors recommend surgery for both symptomatic and asymptomatic patients. Grade IV and V lesions require a multidisciplinary approach with individual analysis. Many grading scales have been proposed,[26,42,43,46–53] all of which focus on anatomic, hemodynamic, and physiological properties associated with AVMs. The Spetzler-Martin grading system has become the scale most often used by treating physicians to perform a relative risk analysis for selecting the appropriate therapy for a specific AVM.

Although the Spetzler-Martin grading scale was designed to predict surgical outcome, it has also been evaluated in the combined management of AVMs, including resection, surgery plus embolization, embolization alone, or radiosurgery, with various combinations.[54] Deterioration due to treatment was seen in 19% of grade I and II patients, 35% of patients with grade III lesions, and 42% of patients with grade IV and V lesions. The scale does not include characteristics such as associated aneurysms, venous stasis, or venous aneurysms that have been associated with hemorrhagic risk. There are no reliable data, in fact, correlating such features with treatment risk. In the future, this grading scale will need to be refined, integrating concepts of eloquence in relation to functional imaging and the potential impact of neurological deficit on the patient's quality of life.

VI. Treatment Options

At present, there are 4 major treatment options available for patients with an AVM of the brain. The lesion can be monitored expectantly with the understanding that the patient would have some risk of hemorrhage or other neurological symptoms such as seizures or focal deficit. Alternatively, intervention can be undertaken with the goal of complete AVM obliteration, because subtotal therapy does not confer protection from hemorrhage. Management strategies include single or combined therapy applying microsurgery, endovascular techniques, or radiosurgery (focused radiation). Each treatment option has associated risks and benefits that will be outlined in the subsections below.

Outcomes of treatment in subsequent sections generally include associated mortality and morbidity, although these are not reported consistently. Selection, assessment, and reporting biases often prevent all but gross comparisons among various series. Glasgow Outcome Scale or other broad disability outcome scales (eg, excellent, good, fair, poor, or death) are frequently used in larger series, but the definitions of categories are inconsistent, and the timing of assessment is rarely standardized. The current literature rarely includes patient-generated functional outcome assessment (quality of life) for various management modalities or third-party adjudication of outcomes. Rates of major and minor treatment-related neurological morbidity are often useful in comparing various therapeutic approaches, but these should be considered in light of such paucity of control. Treatment efficacy is a critical outcome parameter (total/permanent angiographic obliteration of the lesion), as is delay in or failure of lesion obliteration.

Anesthetic and Perioperative Considerations for Microsurgical Resection

Recommendations for anesthetic management are based primarily on level V evidence. In general, conduct of anesthesia

for AVM resection follows the same recommendations for neuroanesthetic management for any intracranial lesion[55] regarding choice of monitoring, vascular access, anesthetic agents, vasoactive drugs, and muscle relaxants.

Because AVM resection is usually not emergent, preexisting medical conditions should be optimized, and neurological dysfunction, either as a result of presenting hemorrhage, presumed effect of the AVM, or preoperative embolization (infarction or edema), should be factored into the intraoperative and postoperative management plan. An important consideration throughout the operative period is the potential for massive, rapid, and persistent blood loss. Choice of intraoperative monitoring is tempered by this eventuality, and adequate amounts of blood, along with access for its administration, must be readily available.

The risk of AVM rupture during induction is probably low based on inferential evidence.[56,57] Nevertheless, blood pressure control that approximates the patient's normal range is sound anesthetic practice in the absence of mitigating circumstances. However, it should be borne in mind that ≈10% of AVM patients harbor intracranial aneurysms[1,4] that may increase the risk of rupture during increases in arterial blood pressure.

Although intracranial pressure control is rarely a problem with the AVM patient who presents for elective resection, intracranial compliance may be abnormal. Therefore, the usual caveats about avoidance of anesthetics and vasoactive agents that cause cerebral vasodilation seem prudent, ie, high inspired concentration of volatile anesthetics and high doses of vasodilators that directly relax vascular smooth muscle.

There is no anesthetic regimen that has been rigorously shown to confer "cerebral protection" in neurosurgical patients. The choice of anesthetic agent must be consistent with safe conduct of intracranial surgery, including brain relaxation, excellent blood pressure control, and rapid emergence. Euvolemia, normotension, isotonicity, normoglycemia, and mild hypocapnia are recommended.[58,59] Profound hypocapnia is not recommended unless indicated for control of brain swelling or surgical exposure.[59]

An ongoing randomized, controlled study (Intraoperative Hypothermia in Aneurysm Surgery Trial 2 [IHAST2]) is evaluating the use of mild induced hypothermia (33°F) for cerebral protection during craniotomy for aneurysm clipping.[60] If successfully completed, this study will provide the first opportunity to gain level I evidence of intraoperative cerebral protection. The induction of general anesthesia results in an obligatory core temperature decrease as peripheral vasodilation redistributes heat to the periphery. The current recommendation is to maintain normothermia or accept the mild decrease in body temperature that results from general anesthesia and not aggressively rewarm patients until timing for emergence is planned. This recommendation is based only on level V data.

Induced hypotension is frequently useful during AVM resection, especially in large AVMs that have a deep arterial supply. Bleeding from these small, deep feeding vessels may be difficult to control, and decreasing arterial pressure facilitates surgical hemostasis. The subject of induced hypotension is discussed extensively in the anesthesiology literature.[55] There is no compelling evidence to use one particular agent. Choice of agent must be placed in the context of the clinical situation (eg, avoidance of β-adrenergic blockers with bronchospastic airway disease or use of nitroglycerin with coronary artery disease) and the experience of the practitioner.

The intraoperative appearance of diffuse bleeding from the operative site or brain swelling and the postoperative occurrence of hemorrhage or swelling have been attributed to normal perfusion pressure breakthrough (NPPB) or "hyperemic" complications.[61] There is no universally accepted definition of what constitutes a hyperemic state, and it should be a diagnosis of exclusion after all other correctable causes for malignant brain swelling or bleeding have been considered. α-Adrenergic blockade may be of use in preventing and treating this syndrome, based on anecdotal information and suggestive observations.[62] Emergence hypertension is frequently encountered after AVM resection. Data suggest that elevated plasma renin and norepinephrine levels are associated with this phenomenon.[62]

The upper and lower limits of blood pressure control have potential opposing effects. Ischemic deficits due to intraoperative sacrifice of an en passage feeding vessel (a vessel feeding an AVM and also sending distal branches to normal brain), for example, may result in a deficit ascribed to brain retraction or to the resection itself. Marginally perfused areas may be critically dependent on collateral perfusion pressure. Maintenance of low or even normal blood pressure may be inadequate and may result in infarction if hypoperfusion is unrecognized. Verification of potential borderline perfusion states may require imaging modalities such as intraoperative or immediate postoperative angiography.

Postoperative hyperthermia may be detrimental[63] and may even be exacerbated by mild, intraoperative-induced hypothermia.[64] Therefore, careful attention should be paid to control of patient temperature in the intensive care unit.

Associated Aneurysms

Intracranial aneurysms are found in ≈7% to 17% of patients.[14,65,66] Intracranial aneurysms can occur on the feeding artery to the AVM. These may involute after resection or obliteration of the brain AVM. Alternatively, patients may also harbor more saccular intracranial aneurysms at typical locations in the circle of Willis. It is recommended that these be approached during the same surgery if the operative field is adequate or that they be treated separately with endovascular or open surgical obliteration. There are no natural history data regarding this point in the literature, and therefore the rationale for treatment of aneurysms that are not associated with AVMs is used.[2]

Brain Edema/Hemorrhage

Two hypotheses for the cause of brain edema and hemorrhage during or after surgery have been proposed: NPPB or occlusive hyperemia. The NPPB theory suggests that postoperative hemorrhage and edema are caused by a failure in autoregulation in the ischemic brain around the AVM. Chronic hypoperfusion in brain surrounding an AVM may cause maximal chronic vasodilation, which results in an inability of these vessels to vasoconstrict in response to the resumption of normal perfusion pressure after the AVM has been resected. According to this theory, the key to prevention of malignant postoperative hemorrhage and edema is staged reduction of

blood supply to the malformation. This can be accomplished by staged surgical ligation of the feeders[67–72] or by endovascular embolization. With the technological advance of interarterial embolization, this is the current recommended route, although admittedly this recommendation is based on apparent safety without statistical documentation in the literature. Surgical resection of the AVM should occur shortly (ie, several days) after the final feeding artery embolization to prevent development of new collateral flow.

A number of observations suggest that the details of this theory are not applicable to most cases of malignant postoperative hemorrhage and edema. Intraoperative studies[73–80] demonstrate maintained autoregulation in the region surrounding an AVM both before and immediately after its resection, even in cases subsequently complicated by edema and hemorrhage. This observation argues against the value of staged operation or embolization in the resection of AVMs.[81] It has also led to the proposal of an alternative hypothesis regarding the cause of malignant postoperative edema and hemorrhage termed "occlusive hyperemia."

This theory postulates that malignant postoperative hemorrhage and edema are caused by either arterial stagnation and obstruction or venous outflow obstruction, which are in turn direct results of resection of the AVM.[76,82,83] Evidence for the role of outlet obstruction in spontaneous hemorrhage presented above tends to support this hypothesis, as does the observation that long feeding arteries correlate with a greater risk of postoperative deterioration than do short vessels of similar diameter and flow.[84] Moreover, given this theory, indications for staged resection would be limited to those cases necessitated by technical factors,[82] and hypotensive therapy in the management of postoperative edema may prove more deleterious than beneficial. All of the data presented regarding these theories are level V, and therefore, their impact on AVM management is only moderate.

Postoperative Care

The recommendations for postoperative care include neurological intensive care monitoring for at least 24 hours. Blood pressure is monitored with an arterial catheter and urine output with an indwelling catheter. Typically, normotensive and euvolemic conditions are maintained; however, tight blood pressure control with agents that do not act in the central nervous system may be appropriate for selected individuals. Perioperative antibiotics, steroids, and seizure medication are used variably. After being monitored in the intensive care unit, the patient is transferred to a standard surgical floor, where mobilization occurs. An angiogram is also performed to confirm complete resection of the AVM during the immediate postoperative period. A new neurological deficit after surgery is usually investigated with a CT scan to rule out a hemorrhage or hydrocephalus. MRI scanning with diffusion-weighted imaging may be appropriate if an infarction is entertained.

In summary, AVM surgery is usually elective and frequently preceded by preoperative embolization. The surgical approach allows complete resection of the nidus, resecting the feeding vessels and subsequently the draining veins. Management of associated aneurysms is determined on an individual basis.

Recommendations

In general, surgical extirpation should be strongly considered as the primary mode of therapy for Spetzler-Martin grade I and II lesions. For patients with small lesions, where surgery offers some increased risk based on location or feeding vessel anatomy, radiosurgery should be strongly considered. For grade III lesions, a combined modality approach with embolization followed by surgery is often feasible (see below). Surgical treatment only is often not recommended for grade IV and V lesions because it confers a high risk.

VII. Endovascular Treatment

Technical advances in interventional neuroradiology/endovascular neurosurgery have afforded new alternatives in the treatment of cerebral AVMs. Flow-directed and flow-assisted microcatheters have made navigation of intracranial vessels safer and have allowed more accurate delivery of embolic materials. Current embolic materials are divided into solid or liquid agents. Solid agents consist of polyvinyl alcohol particles, fibers, microcoils, and microballoons.[84–89] Liquid agents consist of cyanoacrylate monomers such as IBCA (*I*-butyl cyanoacrylate) and NBCA (*N*-butyl cyanoacrylate), as well as polymer solutions such as ethylene vinyl alcohol (EVAL copolymer).[90–94] Other liquid agents include absolute ethanol, with and without the use of contrast agents for visualization under digital subtraction fluoroscopy.[95–97] NBCA has recently been officially approved by the Food and Drug Administration for use in brain AVMs.

Embolization of cerebral AVMs is only one aspect of a multimodality approach to these lesions. Current indications for embolization can be divided into presurgical embolization in large or giant cortical AVMs and embolization before radiosurgical intervention to reduce nidus size. In addition, palliative embolization may be used in large nonsurgical or nonradiosurgical AVMs in patients presenting with progressive neurological deficit secondary to high flow or venous hypertension. In this group of patients, the goal is flow reduction in an attempt to minimize or halt symptom progression. Finally, embolization of a pseudoaneurysm that seems to be related to a hemorrhage is also possible.[98]

Anesthetic and Perioperative Considerations for Endovascular Therapy

Although many of the risks and responses are for the most part conceptually the same, there are also many important differences in the working environment.[99–101] There are generally two schools of thought on how to manage the patient undergoing AVM embolization. One is to rely on knowledge of neuroanatomy and vascular architecture to ascertain the likelihood of neurological damage after embolization. The "anatomy school," therefore, will prefer to embolize under general anesthesia. Arguments for this approach include improved visualization of structures with the absence of patient movement, especially with temporary apnea or when the ventilator is correlated with digital subtraction angiography contrast injection. Furthermore, it can be argued that if the embolic material is placed intranidally, then by definition, no normal brain is threatened.

The "physiological school" trades off the potential for patient movement against the increased knowledge of the true functional anatomy of a given patient, given the wide variability described in these patients.[102,103] At the present time, the physiological approach demands deep intravenous sedation to render the patient comfortable during catheter placement and yet keep the patient appropriately responsive for selective neurological testing.

There is no evidence that either general endotracheal anesthesia or intravenous sedation is associated with a lower rate of complications (level IV evidence).[100] Recommendations for premedication with corticosteroids, anticonvulsants, aspirin, calcium channel blockers, and antibiotics have been made, but none have rigorous support for their use.

Direct transduction of arterial pressure is indicated for intracranial embolization procedures, especially with manipulation of systemic pressure with vasoactive agents. The femoral artery introducer sheath is easily used to monitor arterial pressure. Intravascular pressures may also be monitored from the coaxial (guiding) catheter, as well as via the superselective catheter.

In addition to the recommended American Society of Anesthesiology monitors, additional considerations include placement of an additional pulse oximeter on the foot of the leg that will receive the femoral introducer catheter as an early warning of femoral artery obstruction or distal thromboembolism and overly vigorous compression for postprocedure hemostasis. Bladder catheters assist in fluid management as well as patient comfort. Supplemental oxygen should be given to all patients who have received sedative-hypnotic agents.

General endotracheal anesthesia considerations are conceptually similar to those for open craniotomy. Primary goals of anesthetic choice for intravenous sedation include alleviation of pain or discomfort, anxiety, and patient immobility, but at the same time, the anesthetic must allow for a rapid decrease in the level of sedation when neurological testing is required. There is no evidence one regimen is superior to any other; propofol and midazolam have been directly compared and found to be similarly effective (level II evidence).[104] Choice should be based on the experience of the practitioner and the aforementioned goals of anesthetic management. Common to all intravenous sedation techniques is the potential for upper airway obstruction. Placement of nasopharyngeal airways may cause troublesome bleeding; it may be prudent to place them before anticoagulation. Careful management of coagulation is required to prevent thromboembolic complications during and after the procedures, although algorithms for anticoagulation remain controversial.[105–107]

Profound deliberate systemic hypotension may be induced while the interventionist prepares the glue for injection. Hypotension slows the flow through the fistula and provides for a more controlled deposition of embolic material, particularly the glues. Blood pressure reduction can be achieved with vasoactive agents, general anesthetics, or even by brief, adenosine-induced cardiac pause.[108]

Complications during endovascular navigation of the cerebral vasculature can be rapid and dramatic and require interdisciplinary collaboration. The primary responsibility of the anesthesia team is to preserve cardiovascular function and gas exchange and, if indicated, secure the airway. If emergent endotracheal intubation is necessary, a thiopental and relaxant induction should not be avoided because of the possibility of a transient decrease in perfusion pressure.

In the setting of inadvertent vascular occlusion, a method to increase distal perfusion is blood pressure augmentation with or without direct thrombolysis. The systemic blood pressure may be increased to drive adequate flow via collaterals to the area of ischemia as a temporizing measure.[101] Given the best available evidence, deliberate hypertension in the face of symptomatic cerebral ischemia from vascular occlusion during AVM embolization should not be avoided because of fear of rupturing the malformation.[39] If the problem is hemorrhagic, immediate reversal of heparin is indicated. Protamine is given as rapidly as possible to reverse heparin without undue regard for systemic blood pressure.[101]

Presurgical Embolization

Preoperative embolization of AVMs has become part of the treatment for many AVMs, especially larger lesions.[109–113] Studies comparing surgery with and without embolization do not exist in a prospectively controlled fashion (level I or II study) because the introduction of this technique was immediately believed to be advantageous, and subsequent randomization was deemed inappropriate. Advantages include diminished blood loss and shorter surgical times, the applicability of strategically targeted embolization, and the ability to occlude vessels deemed difficult to control by the operating surgeon, as well as the theoretical benefits of staging flow reduction in the nidus.[114]

The goals of presurgical embolization are to decrease the nidus size of the AVM and to attempt to occlude deep, surgically inaccessible or deep arterial feeding vessels such as the anterior/posterior perforating vessels, choroidal vessels, or posterior cerebral vessels to facilitate surgical excision. Other goals of presurgical embolization are to occlude intranidal aneurysms and high-flow fistulas to presumably promote progressive thrombosis of the nidus of the AVM. Proximal occlusion of arterial feeding vessels and failure to occlude the AVM nidus with embolic material may have a deleterious effect on surgery because of the inevitable development of cortical transmedullary and transdural collaterals.[115,116]

The results and efficacy of intravascular embolization have been presented as level V data. Vinuela et al[113] in their series of 405 patients were able to totally cure the lesion in 9.9% of cases. This was primarily in small and medium AVMs with fewer than 4 pedicles. Hemorrhagic complication rates associated with embolization in more recent series range from 2% to 4.7%. The source of hemorrhagic complications may be arterial perforation, intranidal aneurysm rupture, or untoward venous occlusion. Mortality rates during embolization have been reported to be 1.08% or less, and neurological morbidity rates of 2% to 5% have been reported with the use of superselective Amytal testing and new-generation microcatheters.[112,113,117–119] Numerous studies describe the beneficial effect of presurgical embolization in reducing operative time and blood loss, as well as converting high Spetzler-Martin grade lesions to lower-grade lesions, with a concurrent reduction in morbidity and mortality (level V and level III evidence, respectively).[112,114,117,120] No prospective randomized trials have been performed to verify this observation.

Preradiosurgical Embolization

Endovascular therapy has 3 potential goals when used before radiosurgical intervention for AVMs[121–123]: (1) to decrease target size to <3 cm in diameter, because smaller volumes have a higher cure rate with less morbidity; (2) to eradicate angiographic predictors of hemorrhage, such as intranidal aneurysms or venous aneurysms; and (3) to attempt to reduce symptoms related to venous hypertension. No ideal embolic material has been identified for preradiosurgical use.[123–125] Several reports have documented delayed recanalization of AVMs after angiographic obliteration with polyvinyl alcohol embolization. Recanalization in 16% of patients embolized with particulate agents and treated with radiosurgery has also been reported.[126]

Most centers recommend the use of more permanent agents, such as polymers of cyanoacrylate. However, numerous studies indicate that the use of such agents may also result in a recanalization rate of 14%. This may be dependent on the concentration of acrylic deposited within the nidus.[123–125] There is no evidence that flow reduction alone without reduction of the AVM volume provides any benefit before radiosurgery, and in fact, it may make it more difficult to provide a conformal dose plan at the time of radiosurgical planning (level III evidence).[126]

Palliative Embolization

Palliative embolization may be recommended for patients who have large, inoperable cortical and subcortical AVMs and in patients presenting with seizures resistant to medical management or with progressive neurological deficit thought to be secondary to venous hypertension and/or arterial steal.[127–129] Partial embolization may be successful in reversing these signs and symptoms; however, it is usually only temporary, because collaterals develop rapidly, reducing the effectiveness of such therapy (level V data). Palliative embolization should be used as part of a strategy aimed at staged AVM obliteration, to treat a specific AVM-associated feature (eg, associated aneurysm), or to reverse a specific symptom. There is no evidence that partial AVM embolization alters long-term hemorrhagic risk, and as such, it is not recommended as a broad treatment strategy for AVMs.

Intravascular embolization of AVMs as a sole therapeutic modality is usually only achieved in small lesions fed by no more than 4 arterial pedicles.[113] In many series, permanent occlusion of brain AVMs by embolization was achieved in 10% to 30% of cases.[113,117,127] Current evidence is incomplete and mandates long-term follow-up even when the lesion is embolized with agents such as liquid acrylics and other copolymers, because recanalization can occur.[124,125,130]

Recommendations

Recommendations for endovascular management of AVMs can be divided into presurgical, preradiosurgical, or palliative management for focal neurological symptoms or uncontrolled seizures. The decision to perform embolization of an AVM should take into consideration Spetzler-Martin grade as well as the combined surgical and endovascular risk for a particular patient. The risks of embolization must be weighed against other risks in terms of combined morbidity and mortality for surgery and/or radiosurgery. Currently, all data available are either level III or IV, because no prospective randomized trials exist concerning embolization therapy.

In general, Spetzler-Martin grade II or III lesions may be embolized before surgery or radiosurgery. Grade IV or V lesions should not be embolized unless this is to be done in conjunction with other treatment modalities (surgery or radiosurgery) for the goal of complete care. The only exception to this may be in a patient with a grade IV or V lesion with venous outflow obstruction, in whom embolization is used to reduce arterial inflow to control edema, or in a patient with true "steal," in whom embolization is used to relieve the amount of shunt through the AVM.

VIII. Radiosurgery

Stereotactic radiosurgery has become an important treatment technique for the management of cerebral AVMs. The purpose of radiosurgery is to irradiate the blood vessels of the AVM to cause progressive luminal obliteration and thereby prevent hemorrhage. Involution of the irradiated mass is the final stage of the healing response, as well as the final stage of inflammation.[131] At that time, the AVM vessels are occluded and AVM volume is reduced. With focused radiation, the dose of radiation to brain tissue surrounding the AVM can be minimized.

Indications for AVM Radiosurgery

A large number of studies (level V evidence) indicate that radiosurgery provides satisfactory results for AVM cure with few complications. Radiosurgery is most appropriate for patients with small AVMs, especially when such AVMs are located in eloquent brain locations. Lesions most effectively treated with radiosurgery have volumes <10 cm^3 or maximum diameter <3 cm.[132–134] Candidates for treatment are selected on the basis of AVM volume and location, patient age, and relative risk analysis compared with surgical and endovascular therapies as predicted by the Spetzler-Martin grading scale.

Clinical Experience

The goal of radiosurgery is to obliterate the AVM, prevent rehemorrhage, improve seizure control, and relieve headaches.[132,133,135] The results from patient series have been published (1971 through the present), and radiosurgery has been found to be a safe and effective treatment for specific AVMs based on numerous level V studies.[132,134,136–139] One historical control study by Pollock et al[140] contained level IV data. Radiosurgery leads to complete AVM obliteration (elimination of the hemorrhage risk) in ≈80% of patients within 2 to 3 years, a result that is stratified by AVM size. Smaller AVMs (<10 cm^3) respond better because more radiation can be delivered safely.[141] Angiography is still the standard to confirm complete obliteration.

Postradiosurgery Effects

Immediate postradiosurgery complications are rare. The potential morbidity of radiosurgery is delayed and corresponds with the time course for AVM obliteration, as well as for the inflammatory-mediated effects discussed above.[132,142–144] Symptomatic imaging changes are found in 10% of treated

patients. These changes resolve in half the patients within 3 years of onset. Permanent changes as a result of radiation necrosis occur in 2% of patients. Thus, there is a 5% to 7% risk of treatment-related complications with radiosurgery. In addition, symptomatic patients are exposed to a 3% to 4% risk per year of hemorrhage during the time to obliteration. Therefore, over a 3-year period, the patient has a 14% to 19% risk of complication or hemorrhage in addition to possible incomplete obliteration.

Data regarding protection from rehemorrhage during the 2- to 3-year interval after treatment with radiosurgery are inconclusive. Although Karlsson et al[145] reported protection from rehemorrhage in the interval before complete obliteration, other series[146,147] have not identified such a benefit. In our experience, the hemorrhage rate after radiosurgery remains the same as the hemorrhage rate before radiosurgery until the AVM obliterates. However, there has not been an observed hemorrhage after complete obliteration.

Recommendations

Radiosurgery can be considered in lesions thought to be at high risk from a surgical or endovascular standpoint. The overall efficacy of radiosurgery is higher for small lesions, and therefore, this modality may be considered as a primary treatment for smaller as opposed to larger lesions. However, size is not the only factor in the final determination of treatment. The exact location, patient age, symptoms, and angiographic anatomy must be considered in this decision process. For small, surgically accessible lesions (Spetzler-Martin grade I or II), surgery has fewer risks than radiosurgery. Radiosurgery may be considered in larger lesions (Spetzler-Martin grade II through V) only if the overall goal is complete obliteration of the lesion. Partial treatment of a larger lesion with radiosurgery or embolization subjects the patient to the risks of the procedure without eliminating the risk of hemorrhage.

IX. Multimodality Treatment of AVM

AVMs are often treated by more than one treatment modality. This occurs in one of two fashions. It is done as either a planned maneuver, typically with embolization followed by surgical resection or radiosurgery, or as an unplanned maneuver where one treatment modality fails and a second treatment modality is necessary to obliterate the AVM. This can occur in situations such as residual AVM after subtotal surgical resection or resection of an AVM after incomplete radiosurgical treatment.

There are only level V studies in the literature regarding this form of therapy. In these studies, both strategies of planned and unplanned combined modality therapy are reported. Initial descriptions of combined treatment modality included endovascular and surgical resection. Although results have been reported in series of fairly large numbers of patients,[113,148] patients are still evaluated on a case-by-case basis. Therefore, there are no specific recommendations that can be made as to which patients benefit from multimodality therapy. This level of recommendation is in keeping with the great variability of AVMs in terms of their angioarchitecture, as well as the risk of specific treatment given the factors outlined above.

Recommendations

Multimodality therapy should be performed only if it is part of a total treatment plan to eradicate an AVM. The goals of the different modalities should be clear at the outset. Because of the variability of resources available in any one area of the country or world, some patients are offered partial treatment with a single technique. Such treatment is unjustified. Although it is difficult to make generalizations about specific uses of multimodality treatment, such treatment does appear to play a helpful role in larger lesions (Spetzler-Martin grade III or V) for which complete obliteration is the goal. The hope is that with combined techniques, the overall risk of therapy will be reduced, although this is yet to be proven statistically.

X. Specific Considerations

Pregnant Patients

The data regarding AVM hemorrhage risk during pregnancy are inconclusive. Some studies suggest that the risk of hemorrhage during pregnancy is similar to that at other times.[149–151] In a study of 451 patients who had 540 pregnancies, the incidence of hemorrhage was found to be 3.5% during the 52 weeks after the patient's last menstrual period. In patients with a history of hemorrhage before pregnancy, the incidence of hemorrhage was 5.8% during the year after the last menstrual period, but the number of patients with hemorrhage was small, which made the data nondefinitive. Neither of these rates was significantly different from similar nonpregnant populations.[152]

The rebleeding rate during the same pregnancy for patients who present with hemorrhage during pregnancy may be higher than the early rebleeding rate in nonpregnant patients. Among 27 women with hemorrhage during pregnancy who did not have immediate resection of the AVM, there were 7 recurrences of hemorrhage before or immediately after delivery.[149,151,153] This rebleeding rate of 26% (with a 95% confidence interval of 9% to 49%) is well over the 6% expected in the first year after a hemorrhage in nonpregnant patients. Although these studies represent level V evidence, there is a suggestion that some pregnant patients who present with hemorrhage may benefit from early definitive therapy.

Hemorrhage during delivery has been a major concern of obstetricians and patients; however, the available data would suggest that in most cases, vaginal delivery does not carry a higher risk for hemorrhage than delivery by cesarean section.[152] There are no data available to address whether cesarean section helps to reduce the already low incidence of AVM-associated complications during delivery, although there is evidence that increased venous pressure during a Valsalva maneuver is not directly transmitted to the draining veins.[56]

Recommendations

If a woman anticipates pregnancy and has a known AVM, treatment should be considered before the pregnancy. If the lesion is discovered during pregnancy, a decision should be made regarding the treatment risks versus the risk of hemorrhage during the remainder of the pregnancy if the lesion is left untreated. This also must include the potential risk to the fetus during intervention, whether it be by embolotherapy, surgical extirpation, or radiation and the associated diagnostic

tests. In most cases, such risk-benefit analysis will not support elective treatment of AVMs during pregnancy.

Pediatric Lesions

Pediatric patients make up ≈12% to 18% of surgical AVM series from experienced centers.[154–157] AVMs account for 30% to 50% of hemorrhagic strokes in children,[156,158,159] and pediatric patients are more likely to present with hemorrhage than adults, with some series reporting an 80% to 85% hemorrhage rate as their initial presentation.[154,160] Because of the large degree of arteriovenous shunting relative to cardiac output, neonates and infants can present in cardiac failure from arteriovenous shunting.[161–166] The remaining pediatric AVM patients present with seizures, headache, neurological deficit, or incidentally.

The long potential life span of a pediatric patient with an AVM leads to a high lifetime risk of hemorrhage.[24,25] Hemorrhagic events from an AVM in children have also been associated with a 25% mortality rate.[167] This high risk of hemorrhage from a pediatric AVM would tend to warrant treatment whenever possible; however, 10% to 42% of children with AVMs have been managed without treatment, depending on referral pattern and bias of the institution.[156,160,168,169] Moreover, pediatric AVMs are more commonly found in eloquent locations such as the basal ganglia and thalamus.[160,170,171]

Pediatric AVMs have been treated with surgical excision,[154–157,160,167,169,170,172–176] endovascular embolization,[168,177] radiosurgery,[137,178–182] and multimodality management.[171,183] The efficacy of treatment reported in these series, however, constitutes level V evidence. Most of the large series of pediatric AVMs, regardless of treatment modality, have been associated with higher rates of morbidity and mortality than adult series, except for a few that have reported favorable results.[170,171] The largest surgical series comes from Humphreys et al,[160] who reported a series of 160 pediatric AVMs in which the morbidity and mortality rates were 18% and 11%, respectively. The largest endovascular series is a series by Lasjaunias et al[168] of 179 pediatric AVMs, in which the morbidity and mortality rates were 28% and 16%, respectively. The largest radiosurgical series is a series by Levy et al[180] of 40 pediatric AVMs, in which they reported a 30% rate of permanent neurological deficits.

Several authors have reported AVM recurrence in pediatric patients after total surgical resection, with postoperative angiogram confirming complete obliteration.[157,160,171,184] These authors have had no cases of recurrent AVMs in their adult AVM series, thus suggesting a pathophysiological difference between AVMs that occur in children and those that occur in adults. This could be a consequence of the relatively immature cerebral vasculature in children. It also suggests that AVMs may not strictly be congenital lesions. One study has suggested that certain pediatric AVMs may express higher astrocytic vascular endothelial growth factor than adult AVMs, which may in part explain their ability to recur.[184] The rare recurrence of pediatric AVMs after complete surgical excision may be an indication for postoperative radiographic follow-up in these patients; however, there have been no prospective studies conducted to support this.

Recommendations

The younger the patient, the more conclusively treatment is warranted. More aggressive treatment strategies can be justified in dealing with pediatric patients, whereas only low-risk strategies should be offered to elderly patients.

Management of Complications

Hydrocephalus

Hydrocephalus may occur as a result of intraventricular hemorrhage secondary to an AVM. When this occurs soon after hemorrhage, urgent insertion of ventricular drainage catheters may be necessary. These catheters can also be used to monitor intracranial pressure in patients in the intensive care unit setting. As the ventricular blood is cleared, patients may have chronic hydrocephalus and thus may warrant ventriculoperitoneal shunting. This decision should be made on an individual basis, based on the size of the ventricles and the cerebrospinal fluid pressure. In rare instances, hydrocephalus can result from compression of the aqueduct of Sylvius by large draining veins of AVMs.

Seizures

Obliteration of AVMs may reduce the incidence of seizures. After surgery, one report of level V evidence documented no statistical difference in seizures between surgically treated and medically treated groups of patients with AVMs.[185] Several other reports, however, documented the efficacy of surgical resection of AVMs in decreasing the seizure rate. In one report in 27 patients undergoing surgical resection of an AVM and an epileptogenic center, seizure control was believed to be excellent in 21 of the 27 patients and poor in 1 patient.[186] In a larger series of 200 patients with AVMs, 163 had experienced no seizures preoperatively. Of this group, 8 patients (6%) had new-onset seizures. Of the 102 surviving patients who had presented with seizures, 85 (83%) were seizure free over a 2-year minimum follow-up. Of these patients, 48% no longer received anticonvulsant therapy. Although 17% suffered intermittent seizures, 13 of these patients reported improved control compared with before surgery.[187]

Other surgical series are equally promising and suggest seizure control may correlate with age at seizure onset, duration of seizures, and location of lesion and cortical excision.[188] Similar results have been reported in the radiosurgical literature, with seizure control noted after radiosurgery for AVMs in 55% to 70% of patients with AVM obliteration.[189] Although all of the reports available are level V data, it can generally be expected that surgical or radiosurgical obliteration of an AVM will reduce seizure activity. No studies exist from which recommendations can be made in terms of duration or type of anticonvulsant prophylaxis after treatment.

XI. Other Vascular Malformations

This document specifically addresses intracranial parenchymal or pial AVMs and does not cover recommendations for angiographically occult AVMs, cavernous malformations, dural AVMs or fistulae (including vein of Galen AVM), or spinal AVMs. These other lesions reflect unique considerations of epidemiology, diagnostic evaluation, natural history, risk-benefit analysis, and therapeutic strategies. Other special considerations in rare familial AVMs and those associated with hereditary hemorrhagic telangiectasia (Osler-Weber-

Rendu disease), including vascular malformations affecting multiple organ systems, are also beyond the scope of this report.

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KEY WORDS: AHA Scientific Statements ■ arteriovenous malformations ■ stroke ■ cerebrovascular disease ■ hemorrhage