Introduction

Despite the development of different modalities of treatment and the great advances in the field of neuroimaging, the management of AVMs remains one of the most difficult problems in neurosurgery.

Arteriovenous malformations are generally handled by microsurgical resection, interventional neuroradiology, radiosurgery, or by a combination of these treatments. Treatment should only be indicated after factors such as occupation, age, clinical condition of the patient, and the personal experience and results of the specialists involved with the treatment have been carefully considered and confronted with the natural history of AVMs. In cases in which the risks of a given treatment surpass those of the natural history the best option is to leave the lesion untreated.

The ultimate therapy for most cases of AVMs is still microsurgical resection. The early poor surgical results have led to a more conservative attitude towards AVMs. The challenge of AVM surgery became such that even at this time only a few neurosurgeons are adequately prepared to deal with this pathology. With the development of microsurgical techniques, advances in neuroimaging, neuroanesthesia, intra and post-operative monitoring, the surgical treatment of these lesions became possible.

Endovascular embolization plays an important role as an adjuvant therapy to the microsurgical resection of large lesions. Even in the best hands the rate of cure of AVMs by endovascular techniques alone is still very low.

Radiosurgery is the latest modality of treatment for AVMs. One of the disadvantages of this technique is the time required for the obliteration of the AVMs (around 2 years).

During this period, the patient is unprotected from hemorrhagic events. Also, the results of treatment of large lesions are dismal.

Despite the various treatment options, there has been great debate regarding the best therapeutic approach to AVMs.

It is known that many neurosurgical centers still do not have the facilities for endovascular or radiosurgery techniques or lack trained professionals in those areas. Unfortunately it is also known that even in those places where all treatment options are available many neurologists and neurosurgeons refer their patients to either modality of treatment without considering them as part of a whole therapeutic plan. The different types of treatment for AVMs have advantages and disadvantages. Thus, the best therapy should be discussed by a team of specialists that led by the neurosurgeon would decide the best strategy for each patient.

Natural History and the Role of the Neurosurgeon

The surgeon should confront all the benefits and limitations of the available treatment with the natural history of the lesion. This is particularly true when one indicates surgery as the best therapy.

The risk of bleeding for an AVM is 3-4% per year. The mortality of these cases is 1-1.5%. In other words, in a ten-year period, roughly 30-40% of the patients with untreated AVMs will bleed and
10–15% of them will die.

The surgeon's results must achieve better figures than those of the natural history of the disease. This is particularly important if we consider that most of the surgical disasters in the treatment of AVMs are due to inadequate planning and inexperience of the surgeon.

The surgeon interested in dealing with AVMs must be specially prepared. A detailed knowledge of the anatomy of the normal brain structures is a must. The surgeon must be able to identify the anatomical structures from different angles and approaches, in a three-dimensional way. Although descriptive anatomy is extensively described in the medical literature the only possible way to learn microsurgical anatomy in a three-dimensional fashion is in the microsurgical laboratory. Also the fine and delicate microsurgical technique necessary to deal with the fragile vessels of an AVM can only be adequately obtained with a diligent work with laboratory animals.

Although most surgeons find the training in the laboratory boring or unnecessary the time spent in the lab is the most important step in the long journey of the training program for a surgeon interested in dealing with the treatment of AVMs.

The availability of a good microscope, microsurgical instruments and a well equipped intensive care unit cannot be overemphasized.

It is also necessary to have a team of neuroanesthesists, intensivists, neurologists and nurses specially trained to deal with this pathology. It is also desirable to work with neuropsychologists to better evaluate the cognitive function of the patient before and after the surgery.

**General Standards**

It may seem that AVMs are lesions in which the vasculature is in complete disarray. However, the vessels involved in the arterial supply and venous drainage of such lesions usually follows the patterns of the normal vasculature.

As an example, a temporal AVM around the sylvian fissure (Fig. 1) will be supplied by branches of the middle cerebral artery. The venous drainage will be done through the superficial sylvian vein and/or through the complex of Labbé and Trollard. If this AVM extends to temporal horn of the lateral ventricle there may be branches of the anterior choroidal artery going to the lesion.

A detailed anatomical knowledge of the brain structures gives the surgeon the opportunity to understand the anatomy and the radiological exams in a three-dimensional picture. With this in mind, the surgeon is able to develop a "fourth dimension" that is the ability to understand all the distortions of the normal anatomical structures in the presence of an AVM (or any other pathology) and to choose the most adequate route to approach the lesion.

**TREATMENT STRATEGY**

**Radiological examination**

CT-scans, MRI and a digitally subtracted angiography is essential to diagnose, localize and decide the better treatment. According to the radiological exams the AVMs will be classified and the surgical strategy will be planned. Consequently, the radiological exams must be of a very good quality.

Among the complementary studies angiography plays the most important role. The knowledge of the normal vasculature as seen in the angiogram is essential for the surgeon to evaluate the arterial supply and venous drainage of an AVM. The neurosurgeon should study carefully the radiological exams before surgery, in order to plan the best surgical approach and anticipate all the possible complications and difficulties of that specific case. Unfortunately, younger neurosurgeons are not prepared to interpret an angiography in detail, as needed to approach AVMs. This skill must be developed.
Approaching the Arteriovenous Malformation

The craniotomy must be large enough to expose all the AVM as well some extension of normal brain tissue around its borders. Small craniotomies play no role in the surgical treatment of AVMs. Large craniotomies will expose normal cisterns and fissures through which the surgeon can be shown the best way to the lesions.

Following the patterns of the normal anatomy of the cisterns, fissures, vessels, sulci and gyri, the surgeon can access the lesion with less difficulty and perform its surgical resection in a more rational way.

It is essential to follow the principles of good microsurgery: a clean operative field with minimal or no bleeding, opening of the cisterns and fissures, minimal brain retraction, no damage to the normal vessels or to the brain tissue.

It is important to find and preserve the arachnoid plane. If the surgeon loses the arachnoid plane there is a greater risk to enter the AVM’s nidus before an adequate exposure of the lesion is achieved. Serious bleeding or damage to the normal brain may result.

The surgeon must avoid bleeding during the surgical procedure. Bleeding is the worse enemy of a good microsurgical technique. The blood fills the subarachnoid and subpial spaces, overshadowing important reference points for the surgeon.

The myth that surgery of AVM is a bloody one is wrong. With tenacity and diligent work bleeding can be controlled with bipolar cauterization, and clean operative fields can be the rule rather than the exception.

In our series, blood transfusion during surgery is rare.

With the normal anatomical pattern in mind, the surgeon must achieve proximal control of the vessels going to the AVM. Ideally, the surgeon must dissect all the vessels around the AVM and only obliterate them very close to the lesion. This will avoid the obliteration of the so-called vessels “en passage” which although passing through the AVM, are in reality going to normal areas of the brain.

Bipolar cauterization should be done only when the surgeon is sure that a specific vessel belongs to the AVM. The use of temporary clips may be useful, during this process.

The greatest difficulty lies on the coagulation of the deep feeders. These vessels are usually small, with a high flow and fragile walls that are difficult to obliterate.

After coagulation of most afferents, the arterio-venous shunt is interrupted and the AVM “dies”.

The last step is the resection of the lesion and cauterization of the draining veins. We use bipolar cauterization to obliterate the vessels of an AVM. These vessels are very fragile, as compared to normal vessels. Therefore, their cauterization is much more difficult. The vessels may rupture during coagulation, requiring patience from the surgeon to deal with the resulting bleeding.

Although the perfect bipolar unit is yet to be developed, it is the most important surgical tool during the surgery of AVMs. Laser coagulation is not superior to any good bipolar available, and does not play an important role in the surgery of AVMs.

Temporary mini-clips applied in small vessels prior to coagulation may be helpful. Very rarely, we use aneurysm clips or any other metallic clip to obliterate the vessels.

CLASSIFICATION

In order to decide whether to operate and the necessity of any adjunctive therapy we use the grading system proposed by Spetzler and Martin 1) (Table 1).

This classification takes into consideration the size, localization and venous drainage of an AVM and gives an approximate idea of the risks and difficulties involved with the surgical resection of each lesion. Accordingly these lesions can be graded from I to V. However the classification can be
misleading in some cases. As an example a large frontal AVM with a superficial venous drainage in a non eloquent area of the brain is a grade III according to the Spetzler-Martin classification. A small thalamic AVM, near the foramen of Monro draining into the internal cerebral veins is also classified as grade III in the Spetzler-Martin classification system.

Although these two AVMs are grade III, the difficulty and risks involved with the surgical resection and the prognosis of both lesions are completely different. Likewise, a grade III AVM localized in the uncus is less difficult to approach than a similar one localized in the walls of the ventricles.

Since AVMs of the same grade offer different difficulties for the surgeon according to their localization and size many subgroups could be created.

With this in mind and in order to better understand and treat grade III lesions we have modified the classification of Spetzler-Martin to include two subgroups: grade IIIA and grade IIIB. grade IIIA AVMs are considered grade III due to its large size. Grade IIIB AVMs are classified as grade III due to the venous drainage and/or eloquence. The treatment and surgical results in the two subgroups are different.

### Surgical Results

In a ten-year period we have operated on 344 AVMs (Table 2).

Based on the modified Spetzler-Martin classification, our surgical results allowed us to adopt our own criteria for indication of the modality of treatment.

**Garde I and II**

All grade I and II lesions should be operated, regardless of their clinical presentation. In our series, total surgical resection was achieved in 100% of the cases with no mortality and minimal morbidity (Table 4).

For elderly patients with limited life expectancy or in poor clinical condition surgery is not indicated. For those patients that refuse surgical treatment, radiosurgery or endovascular embolization can be an alternative.

For all the other grades, the choice of the best treatment must be individualized.

**Grade IIIA**

AVMs classified in this subgroup are lesions that are considered Grade III due to its large size. For this subgroup we also indicate surgical resection. However, since the surgery of a large AVM is often difficult, prior embolization usually helps the surgical resection of these lesions. Therefore, unless the clinical conditions prove to be a limiting factor we manage grade IIIA patients with preoperative endovascular embolization followed by microsurgical resection of the AVM.

**Grade IIIB**

AVMs classified in this subgroup are usually small lesions located in areas where surgical resection is either too difficult or prohibitive.

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**Table 1** AVM Classification (Spetzler-Martin)

<table>
<thead>
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<tr>
<td>eloquent area</td>
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<tr>
<td></td>
<td>0</td>
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<td></td>
<td>yes</td>
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**Table 2**

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<td>grade IIIA</td>
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<tr>
<td>grade IIIB</td>
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<tr>
<td>grade IV</td>
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<td>13%</td>
</tr>
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<td>17</td>
<td>5%</td>
</tr>
<tr>
<td>total</td>
<td>344</td>
<td>100%</td>
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Management of AVMs
For grade IIIB AVMs the treatment of choice is radiosurgery. However, this group of lesions is not uniform and there are many exceptions to this rule: with refined microsurgical techniques, grade IIIB lesions localized in the mesial aspect of the temporal lobe, for example, can be surgically excised (Fig. 2). Also, deep seated lesions can be reached and resected without brain damage. For deep seated lesions the use of a stereotactic guide is very useful.

The results of the treatment of patients with AVMs grade IIIA and B are shown in Table 5. We consider bad result those patients that became hemiplegic or aphasic. Fair outcome refers to patients with neurological deficits but that have a Karnofsky scale of 70 or more. Good results refer to patients without neurological deficits.

The morbidity of subgroup IIIB is significantly higher than in subgroup IIIA. Also, in group IIIB we had 2 deaths (2.1%). It is worth noting that 90% of the patients in group IIIB that had morbidity or mortality had pre-operative neurological deficits. In our series, grade IIIB AVMs bleed more often, and the patients have a poor neurological status as compared with the patients of the former groups.

Grade IV

Grade IV lesions are preferably treated conservatively. Even if the AVM has bled, the risks of causing a permanent deficit with the operative treatment should be carefully weighed. This is also true for grade IV lesions presenting with repeated episodes of bleeding or with progressive neurological deficits.

However, in those patients with fixed neurological deficits caused by the lesion or for those AVMs located in non eloquent areas, surgery with prior embolization should be considered. When surgery is indicated, prior endovascular embolization, in our experience, is mandatory (Fig. 3).

We have operated on 45 patients with grade IV AVMs. In 34 patients we have achieved good results (75%). We had 9 patients with fair outcome (20.6%) and 2 deaths (4.4%).

Grade V

As the excision of grade V malformation is often associated with a significant risk of postoperative deficits, we prefer to follow the patient without surgical treatment.

As for group IV, there are some exceptions: patients with progressive neurological deficit, repeated hemorrhagic events or with raised intracranial pressure syndrome should be considered for
endovascular embolization plus surgical excision.

Embolization plus surgery may also be considered for those patients with fixed neurological deficits or for AVMs located in areas where the surgical excision will result in minimal neurological deficits.

We have operated on 17 patients harboring a grade V AVM. Twelve of them had a good outcome (70%), while 4 had fair results (23.5%). There was no death in this group, but 1 (0.8%) patient had a poor outcome.

**Integrated Approach**

The best treatment of AVMs should be decided by a team in which the neurosurgeon, the neuroradiologist, and the radiosurgeon work together.

We find the modified Spetzler-Martin classification a useful guide for the best treatment, even though still many are the exceptions to the classification.

The goal of AVM surgery is the total resection of the lesion. The success of total removal is related to the grade of the AVM, as shown in Table 7.

Total surgical resection was successful in all patients with grade I, II, and IIIA AVMS. In 4 patients (3%) with grade IIIB AVM, the resection was incomplete. In our series incomplete obliteration of the AVM was more common in Grades IV and V.

**AVMs and Aneurysms**

Sacular aneurysms associated with AVMs are not uncommon. As a rule, they should be treated first.

Aneurysms arising in the main feeders of the AVM can be treated followed by excision of the AVM in the same procedure. In such cases, preoperative embolization should not be performed. After embolization, the pressure inside the vessel feeding the AVM grows and the risk of aneurysm bleeding is higher.

**Posterior Fossa AVMs**

It is our impression that posterior fossa AVMs that are not located in the brainstem are lesions for which surgical resection is significantly less difficult than those located in the supratentorial compartment.

These AVMs usually receive supply from branches of either the superior cerebellar artery, the PICA and/or the AICA, which can frequently be easily accessed during surgery.

All cortical and subcortical lesions related to the cerebral hemispheres and to the vermis and located posteriorly to the flocculus usually do not involve the brainstem, and therefore can be resected with very low morbidity.

**AVMs of the Basal Ganglia Region**

Arteriovenous malformations arising at the region of the basal ganglia still carry a considerable risk of morbidity and mortality with any given treatment modality. Surgical indications are questionable and the majority of the cases are usually considered inoperable. Surgical intervention for those
AVMs located in the caudate, globus pallidus, and internal capsule usually results in major postoperative neurological deficits. Consequently, interventional neuroradiology and radiosurgery are considered the treatments of choice for most cases.

However, the precise localization of such lesions through neuroimage studies allied with a thorough knowledge of the anatomy of the region, allows some selected small AVMs located in the vicinity of the basal ganglia to receive definitive surgical treatment with minor postoperative morbidity.

**Intrinsic Brainstem AVMs**

We see no indications for any of the currently available types of treatment for those purely intrinsic brainstem AVMs. We never perform surgery for these lesions, except for small extensions of the AVM to the pial surface of the brainstem.

**Breakthrough**

There are many publications dealing with the breakthrough phenomenon, explained as the lack of autoregulation of the normal vessels around a large AVM after its excision. Brain swelling follows with important neurological manifestation.

However, many cases of breakthrough are, in reality, problems with the surgical technique employed, particularly regarding incomplete resection of the AVM.

In our series, post-operative changes suggestive of breakthrough phenomenon were found in very few cases.

Post-operative hemorrhagic complications were all due to residual AVMs.

**Embolization**

Endovascular embolization requires a well-trained specialist working cooperatively with the neurosurgeon. Endovascular embolization is not a simple procedure and should not be considered as a “conservative” approach.

In our service, the patient is prepared for embolization with the same pre-operative routine used for a surgical patient. This is particularly true when one considers that a few patients may have complications related to the procedure and may require urgent surgical treatment.

The pre-operative embolization makes the surgical excision of an AVM less difficult. Nevertheless the embolization of the afferents only, does not help the surgeon. It is very important to obliterate the AVMs nidus.

The radiologist must try to initiate the nidus embolization through the small vessels that feed the AVM, and not through the larger ones. If a large vessel going to the AVM is obliterated the flow is

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**Fig. 1** (A) Surgical view of a right-sided sylvian arteriovenous malformation. Compare figure 1A with the anatomical specimen of figure (B) and note that the AVM follows the pattern of the normal anatomy.
Fig. 2A: Artistic representation of the approaches to anterior mesial temporal lobe AVMs (1. Trans-sylvian; and 2. Pretemporal approaches)

B: Right side digital carotid angiogram showing an arteriovenous malformation located in the anterior portion of the mesial temporal lobe region.

C: Post-operative right side digital carotid angiogram showing complete resection of the lesion.

D: Intra-operative view of the same case. Through a pretemporal approach the temporal lobe has been elevated posteriorly after wide opening of the sylvian fissure and the AVM can be seen on the anterior superior surface of the uncus. (1) Frontal lobe; (2) Optic nerve; (3) Internal carotid artery; (4) Bridging vein; (5) Middle fossa; (6) Oculomotor nerve; (7) AVM; (8) Retractor over the temporal lobe.

E: Intra-operative view after resection of the AVM. (1) Right optic nerve; (2) Internal carotid artery; (3) Anterior cerebral artery; (4) Middle cerebral artery; (5) Oculomotor nerve; (6) Middle fossa; (7) Pons.
Fig. 3 Parieto-occipital AVM.
A: Right side digital carotid angiogram showing the middle cerebral artery supply to the arteriovenous malformation.
B: Right side digital vertebral angiogram showing the posterior cerebral artery supply to the arteriovenous malformation.
C: Intraoperative view of the arteriovenous malformation.
D: Intraoperative view of the arteriovenous malformation after its resection.

Averted to the smaller vessels, making the control of bleeding during surgery much more difficult.

The pressure of the vessels close to the AVM is low, due to the steal from the arteriovenous shunt. After the obliteration of the AVM, the pressure inside these vessels rises abruptly. Due to the lack of autoregulation vasoplegcy with brain swelling may occur resulting in the breakthrough phenomenon.
Management of AVMs

With prior embolization, the flow to the AVM becomes lower and so the vessel pressure close to the lesion grows. The surgical resection of the AVM in such cases is less risky because it does not cause abrupt alterations in the perfusion of normal areas of the brain.

The material we use for is bucrilate, tantalum powder and lipiodol, which mixes into a soft substance. The surgical manipulation of the AVM embolized with this material is less difficult.

Embolization triggers an inflammatory response that may manifest with headaches, fever, meningismus, deficits or seizures. Patients receive anticonvulsants and corticosteroids routinely. As this inflammatory response may cause adherence of the AVM to the surrounding brain tissues or to the dura-mater, surgery should ideally be performed 4 to 5 days after embolization.

Barbiturate coma and or hypotension is indicated only in those cases where the venous drainage was obliterated and the risk of bleeding is higher. Hypertension is avoided after the procedure.

Radiosurgery

This elegant modality of treatment for AVMs has an important role in small lesions (less than 3 cm), particularly those deep seated or in eloquent areas of the brain (grade III B). It is also an alternative when surgery of larger AVMs can not be performed. However, as said before, for larger lesions the results of radiosurgery are very dismal. The drawback of this treatment is that it requires a variable number of months to work, leaving the patient untreated during this period. Also, the total obliteration rate of this modality is not superior to that achieved with surgery.

Although not invasive, radiosurgery also has its complications. Focal or generalized seizures may follow the treatment, within the first 24 hr. Long term morbidity is directly related to the dose used for treatment. Vasculopathy and/or radiation necrosis caused by the treatment may result in neurological deficits.

Conclusion

The surgical management of arteriovenous malformations demands the integrated approach of a specially trained neurosurgeon, of an interventional neuroradiologist and a radiosurgeon. The interventional radiologist and radiosurgeon must work as team led by the neurosurgeon. Each AVM is studied by the three specialists together and the best modality of treatment is decided for each case.

The idea of a team approach should also involve neuroanesthesists, intensivists, neurologists, neuropsychologists, nurses and a dedicated contingent of professionals adequately prepared for the management of these lesions.

For the neurosurgeon, a detailed anatomical knowledge of the brain structures and a refined microsurgical techniques is mandatory. Training in a microsurgical laboratory is essential.

Finally, the surgical treatment must offer the patient better chances than the natural history of the AVMs.

References