A proposed grading system for arteriovenous malformations

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An important factor in making a recommendation for treatment of a patient with arteriovenous malformation (AVM) is to estimate the risk of surgery for that patient. A simple, broadly applicable grading system that is designed to predict the risk of morbidity and mortality attending the operative treatment of specific AVM's is proposed. The lesion is graded on the basis of size, pattern of venous drainage, and neurological eloquence of adjacent brain. All AVM's fall into one of six grades. Grade I malformations are small, superficial, and located in non-eloquent cortex; Grade V lesions are large, deep, and situated in neurologically critical areas; and Grade VI lesions are essentially inoperable AVM's. Retrospective application of this grading scheme to a series of surgically excised AVM's has demonstrated its correlation with the incidence of postoperative neurological complications. The application of a standardized grading scheme will enable a comparison of results between various clinical series and between different treatment techniques, and will assist in the process of management decision-making.

KEY WORDS • arteriovenous malformation • grading system • surgical resection • prognosis

The surgical treatment of arteriovenous malformations (AVM's) of the brain is primarily intended to eliminate the continued risk of disastrous intracranial hemorrhage. The decision to recommend surgery rests on an objective comparison of the long-term risks presented by an untreated AVM, with the more immediate risks of operative treatment. Clearly, the individual patient with an AVM is benefited only if his operation is accomplished without mortality or disabling morbidity. As there is now information on the long-term risks or natural history of untreated AVM's, educated surgical decision-making requires an objective method — a grading system — for predicting the risks of operation in individual cases of AVM's.

An ideal grading system would define, for each specific AVM, the degree of difficulty involved in safely removing the malformation. Such a grading system should provide a reasonably accurate estimation of operative morbidity and mortality, and be simple yet comprehensive enough to be readily applied to all cerebral AVM's. Previously proposed grading schemes, which have been based only on AVM size or on the number and distribution of feeding arteries, are simple enough to be easily applicable. These fail, however, to consider such important variables as anatomic location, degree of vascular steal, eloquence of adjacent brain, and pattern of venous drainage. Other systems are so complicated that they are difficult to be applied easily and rapidly. In order to correct these shortcomings a simple system that weighs the important features of an individual AVM, and thus grades the lesion according to its degree of surgical difficulty, is proposed.

Description of Grading System

Graded Variables

The major factors important in determining the difficulty of resecting an AVM include: size, number of feeding arteries, amount of flow through the lesion, degree of steal from surrounding normal brain, location, surgical accessibility, eloquence of adjacent brain, and pattern of venous drainage. A grading system based on all of the above variables would be too cumbersome for practical use. However, experience with the management of AVM's has established that many of these factors are interrelated, allowing us to simplify the grading process. By reducing the graded variables to three generalized features of AVM's, the scheme can be simplified without ignoring critical factors. The three variables considered are: 1) size of the AVM; 2) pattern of venous drainage; and 3) neurological eloquence of the brain regions adjacent to the AVM.
Grading system for arteriovenous malformations

Size of the AVM. The size of the AVM is determined by measuring on angiograms the largest diameter of the nidus of the malformation. When magnified angiographic views are considered, a correction for the magnification factor is required. The size of the AVM is determined to be small (< 3 cm), medium (3 to 6 cm), or large (> 6 cm), and the AVM is scored appropriately.

The size of the malformation is responsible for much of the technical difficulty in removing AVM’s. The larger an AVM, the larger the amount of normal adjacent neural tissue that is exposed to injury during microsurgical resection of the nidus. Large AVM’s mandate longer operating time, thereby increasing the risk of anesthesia-related complications. Furthermore, the criterion of size encompasses several of the other important factors that determine the degree of surgical difficulty. In general the size of an AVM determines, or is closely related to, the number of feeding arteries, the amount of flow, and the degree of steal.

Pattern of Venous Drainage. The course of the draining veins is determined from the angiogram. The venous pattern is considered superficial if all the drainage from the AVM is through the cortical venous system. The venous pattern is considered deep if any or all of the drainage is through deep veins (such as internal cerebral veins, basal veins, or precentral cerebellar vein). In the posterior fossa, only cerebellar hemispheric veins that drain directly into the straight sinus or transverse sinus are considered to be superficial veins.

Clearly, the pattern of venous drainage is closely related to the surgical accessibility of an AVM. Deep venous drainage, no matter how small, further complicates AVM excision. Often the vast majority of an AVM will have been separated from the surrounding brain when the small arterialized subependymal veins of the deep component are encountered. These veins are friable, resist bipolar coagulation, and have the dangerous propensity to retract and bleed into the parenchyma or ventricle when disrupted.

Eloquence of Adjacent Brain. Eloquent brain regions are defined as those that speak to readily identifiable neurological function and, if injured, result in a disabling neurological deficit. For the purpose of this grading scheme, the following are considered eloquent areas (Fig. 1): the sensorimotor, language, and visual cortex; the hypothalamus and thalamus; the internal capsule; the brain stem; the cerebellar peduncles; and the deep cerebellar nuclei. Areas with much more subtle neurological function, or areas in which injury does not cause permanent disabling deficit (such as the anterior portion of the frontal or temporal lobes, or the cerebellar cortex) are considered non-eloquent. To preserve simplicity, it is assumed that eloquent cortical regions occupy their normal anatomic location. For the purpose of grading, Wada testing or electrophysiological mapping techniques designed to precisely localize eloquent cortex are not required. Data from these techniques, if available, can easily be used for grading brain eloquence in this scheme. The removal of AVM’s adjacent to eloquent areas carries a much greater risk of disabling neurological morbidity than does excision of these lesions from less critical regions. The brain that is adjacent to the AVM is liable to experience damage during surgery from AVM dissection and retraction, and after surgery from postoperative hemorrhage or edema.

Determination of Grade

In order to assign an AVM grade, the size, the venous drainage, and the eloquence of the adjacent brain are determined from angiography, computerized tomography (CT), and/or magnetic resonance imaging (MRI). Particularly with deep supratentorial or posterior fossa malformations, CT and MRI are helpful in delineating the topographical relationship of the AVM to eloquent regions. A numerical value is assigned for each of the
Historical article

TABLE 1

<table>
<thead>
<tr>
<th>Graded Feature</th>
<th>Points Assigned</th>
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<tbody>
<tr>
<td>size of AVM</td>
<td></td>
</tr>
<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 only</td>
<td>0</td>
</tr>
<tr>
<td>deep</td>
<td>1</td>
</tr>
</tbody>
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* Grade = [size] + [eloquence] + [venous drainage]; that is (1, 2, or 3) + (0 or 1) + (0 or 1).

categories (Table 1). The grade of the lesion is derived by summing the points assigned for each category. The lowest grade possible is Grade I; such a lesion would be small (1 point), located in a non-eloquent region such as the anterior frontal lobe (0 points), and have exclusively superficial drainage (0 points) (Figs. 2 and 3). Complete surgical excision of such an AVM would present relatively minor technical difficulties and would entail very little risk of resultant morbidity or mortality. The highest grade within this scheme is Grade V; an AVM of this type would be larger than 6 cm (3 points), located within or immediately adjacent to eloquent brain (1 point), and a portion of the drainage would empty into the deep venous system (1 point) (Figs. 7

Fig. 2. Carotid angiograms, lateral view (left) and anteroposterior view (right), showing a Grade I arteriovenous malformation (AVM). This AVM is less than 3 cm in diameter (small: 1 point), located in the anterior frontal lobe (non-eloquent: 0 points), and drains through cortical veins (arrows) (superficial drainage: 0 points).

Fig. 3. Vertebral angiograms, lateral view (left) and anteroposterior view (right), showing a Grade I arteriovenous malformation (AVM). This cerebellar AVM is less than 3 cm in diameter (small: 1 point), located superficially on the cerebellar cortex (non-eloquent: 0 points), and drains only through superficial cerebellar hemispheric veins (arrows) (superficial drainage: 0 points).

R. F. Spetzler and N. A. Martin

Fig. 4. Carotid angiograms, lateral view (left) and anteroposterior view (right), showing a Grade II arteriovenous malformation (AVM). The AVM is less than 3 cm (small: 1 point), located in the dominant hemisphere adjacent to the receptive language area (Wernicke's area) (eloquent: 1 point), and has exclusively superficial venous drainage (arrow) (superficial drainage: 0 points).

Fig. 5. Carotid angiograms, lateral view (left) and anteroposterior view (right), showing a Grade III arteriovenous malformation (AVM). The AVM is less than 3 cm (small: 1 point), located adjacent to the thalamus (eloquent: 1 point), and drains into the galenic venous system (arrow) (deep drainage: 1 point).
Grading system for arteriovenous malformations

Fig. 6. Carotid angiograms, lateral view (left) and anteroposterior views, arterial phase (center) and venous phase (right), showing a Grade IV arteriovenous malformation (AVM). This AVM is slightly less than 6 cm in diameter (medium: 2 points), located in the dominant parietal lobe adjacent to Wernicke's area (eloquent: 1 point), and drains in part into the galecic system (arrow) (deep drainage: 1 point).

Fig. 7. Carotid angiograms, arterial phase (left) and venous phase (right), showing a Grade V arteriovenous malformation (AVM). This AVM is larger than 6 cm in its greatest diameter (large: 3 points), located in the corpus callosum with a deep thalamic component (eloquent: 1 point), and drains predominantly into a hugely dilated internal cerebral vein (arrow) (deep drainage: 1 point).

and 8). A Grade V lesion is associated with significant risk of surgical morbidity and mortality. These large, critically located malformations require extensive dissection in close proximity to important brain regions; their removal may be complicated by difficulties with controlling fragile deep veins. Obliteration of the large AVM shunt presents surrounding normal vessels with a sudden increase in perfusion that may result in vasogenic edema or even hemorrhage (a phenomenon that has been termed "normal perfusion pressure breakthrough"). Various combinations of lesion size, location, and venous drainage pattern result in the intermediate grades of AVM (Figs. 4 to 6). A schematic representation of all 12 possible combinations of the graded criteria is found in Fig. 10.

There are certain lesions that should not currently be considered for surgery. Within this group are extremely large diffuse AVM's that are dispersed through critical neurologically eloquent areas, or malformations with a diffuse nidus that encompasses critical structures such as the hypothalamus or brain stem (Fig. 9). As surgical resection of such lesions would almost unavoidably be associated with a totally disabling deficit or death, these AVM's fall into a separate category that can be termed "Grade VI" or, more simply, "inoperable."

Fig. 8. Vertebral angiograms, anteroposterior view (left) and lateral view, venous phase (right), showing a Grade V arteriovenous malformation (AVM). This posterior fossa AVM is larger than 6 cm (large: 3 points), located in the cerebellar hemisphere with extension into the inferior cerebellar peduncle and region of the deep cerebellar nuclei (eloquent: 1 point), and drains primarily into the galecic system (deep drainage: 1 point).
Application of the Grading Scheme

In order to test the predictive value of this system, 100 consecutive AVM's were completely resected by one of us (R.S.) were retroactively analyzed. The lesions were graded on the basis of their radiographic studies, and the complications of surgery for the malformations in each grade were tabulated and presented in numerical and graphic form (Table 2 and Fig. 11). The complications were broken down into the categories of minor, major, and mortality. Temporary neurological deficits lasting less than 3 days are not included. There was no mortality. The specifics of morbidity are listed in Table 3. The results indicate that the correlation between AVM grade and the incidence of neurological complications is good. Grade I and II lesions were resected with very low incidence of surgically induced neurological deficit, while surgery for Grade IV and V AVM's was accompanied by a significant number of neurological complications.

In order to test the reliability and consistency of the grading system, 25 angiograms of patients with AVM's were graded independently by one of the authors and by two other neurosurgeons. There was complete agreement among all of the observers in 23 of the 25 cases, and in 24 of 25 cases between one of the authors and a neurosurgeon. In two cases the grades varied by one; in each instance the presence or absence of eloquence was in question. In one case an AVM was situated in the anterior portion of the dominant temporal lobe, and in the other the AVM was situated in the periventricular area of the left frontal horn. Even in retrospect the judgment is difficult. Thus, there is excellent correlation between independent observers in grading AVM's based on this system.

The need for all three variables becomes quickly apparent when the operative difficulty is compared between an AVM of 2 cm located in the frontal pole and a similarly sized AVM located in the thalamus. By just using size, both AVM's would be considered as Grade I; however, by including both eloquence and deep venous drainage the AVM's are, respectively, in Grades I and III, assessing correctly the expected morbidity and mortality in the surgical management of each lesion.

Discussion

Simply put, surgical removal of an AVM is indicated if the risk of operation is less than the risk determined by the natural history of the AVM. As more data have accumulated, it has become apparent that the long-term prognosis for patients with AVM's is grim. The ongoing risk of intracranial hemorrhage from an AVM is 2% to 3% per year, and the risk for recurrent hemorrhage is higher than this, at least temporarily, after an AVM has bled.\textsuperscript{4,5,6} The risk of death associated with initial AVM rupture is approximately 10%, and the mortality rate increases with each subsequent hemorrhage.\textsuperscript{5,6,12} The incidence of neurological deficit is approximately 50% for each episode of hemorrhage.\textsuperscript{14} In

\begin{table}
\centering
\begin{tabular}{|c|c|c|c|c|c|c|}
\hline
\textbf{Grade} & \textbf{No. of Cases} & \textbf{No Deficit} & \textbf{Minor Deficit} & \textbf{Major Deficit} & \textbf{Death} \\
\hline
\textbf{I} & 23 & 100 & 0 & 0 & 0 & 0 \\
\textbf{II} & 21 & 95 & 1 & 7 & 0 & 0 \\
\textbf{III} & 25 & 84 & 3 & 12 & 4 & 0 \\
\textbf{IV} & 15 & 73 & 3 & 20 & 1 & 7 & 0 \\
\textbf{V} & 16 & 69 & 3 & 19 & 2 & 12 & 0 \\
\hline
total & 100 & 86 & 10 & 4 & 4 & 0 \\
\hline
\end{tabular}
\caption{Correlation of AVM grade with surgical results*}
\end{table}

\begin{table}
\centering
\begin{tabular}{|l|}
\hline
\textbf{Minor Deficit} \\
\hline
1. very mild increase in brain-stem deficit \\
2. temporary increase in visual field deficit \\
3. temporary increase in aphasia and weakness \\
4. mild increase in aphasia (resolved 90%) only detectable with rapid speech \\
5. temporary mild increased weakness \\
6. increase in trigeminal nerve deficit (anticipated) \\
7. mild temporary increase in hemiparesis \\
8. mild residual ataxia \\
9. temporary mild dysphasia \\
\hline
\textbf{Major Deficit} \\
\hline
1. hemiparesis \\
2. increase in aphasia \\
3. homonymous hemianopsia (anticipated) \\
4. severe neurological deficit (presumed normal perfusion pressure breakthrough) with major aphasia and hemiparesis \\
\hline
\end{tabular}
\caption{Details of instances of deficit}
\end{table}

\textsuperscript{*} See also Fig. 11. AVM = arteriovenous malformation.
Grading system for arteriovenous malformations

Addition to the complications of intracranial hemorrhage, patients with AVM’s face the lesser risks of developing flow-related symptoms, such as ischemic deficit due to steal.

An estimation of the risk that confronts an individual patient with an AVM requires specific consideration of certain characteristics of both the AVM itself and the patient harboring the AVM. For instance, untreated AVM’s located in the posterior fossa appear to have a particularly poor prognosis, and small AVM’s seem to
have a greater propensity to bleed spontaneously than do large malformations. Children face a greater danger than do adults in that they have before them the period of life of highest risk for AVM rupture (ages 15 to 40 years). Patient-specific characteristics such as these must be considered in the formulation of prognosis for each case. Recently published reviews provide a more detailed analysis of the clinical features that influence the behavior of individual AVMs and of the natural history of these lesions as a group.

There is less available information that can be applied to a determination of the risks that attend surgical removal of an individual AVM. In the past, this estimation has been dependent on the knowledge and personal experience of the surgeon involved in the case, and has often been imprecise and nonsystematic. It has been difficult to apply the surgical results as reported in the literature because the AVMs in these reports have been incompletely characterized.

As discussed above, categorization of AVM's according to size or number of feeding arteries only is insufficient. For these reasons this grading system was constructed. It is simple, easy to learn, and easy to apply. No relatively simple grading system will accurately classify all the variations of such complex vascular lesions. For instance, relatively small AVM's may have more or less direct arteriovenous connections resulting in very high flows — a feature that considerably complicates their surgical treatment. This system may assign an inappropriately low grade to a lesion, since size rather than flow is scored. The location and organization of eloquent areas, particularly those adjacent to an AVM, may vary from the norm and thus be another potential source of error. This may result in under- or overestimation of the risk of causing significant neurolological deficit in a specific case. In spite of occasional inappropriateness of the assigned grade, we believe that the simplicity of the system is an advantage. In order to take into consideration every possible anatomic and physiological permutation of AVM's, the scheme would have to be so complex as to be impractical.

This grading system has demonstrated a relationship to the technical difficulty of removing individual AVM's. Virtually all the Grade I and II AVM's were removed without much difficulty in single-staged procedures. The Grade IV and V lesions, however, often required pre- and intraoperative embolization and multistaged surgical resection.

This grading system is designed to be predictive of the results of complete surgical resection of AVM's. This remains the "gold standard" for the treatment of AVM's, as it is the only method that virtually eliminates the risk of future hemorrhage. Long-term follow-up review of patients whose AVM's were completely excised has confirmed that they are almost fully protected from rebleeding. Incomplete surgical removal and feeding artery ligation do not prevent later bleeding. Incomplete obliteration of the AVM by embolization also fails to provide protection from subsequent hemorrhage. Even in cases that show complete angiographic obliteration after embolization, there is no assurance that hemorrhage cannot occur in the future. In such thoroughly embolized cases, the nidus of the AVM nevertheless remains in the brain and the possibility for delayed recanalization of portions of the malformation through collateral channels exists.

Stereotactically directed radiation therapy is another technique available for the treatment of AVM's. Our grading system may be applied to lesions treated either by radiation therapy or by embolization for the purpose of comparing the results of these techniques with those of surgical excision. It is, however, quite possible that this system will not accurately predict the results of embolization or radiation therapy, as the complications of these types of treatment differ from those of surgery.

A prospective application of this grading scale in AVM cases is currently underway. If this scale in its simplicity proves to predict operative morbidity in other series, it will provide a useful tool for decision-making in the management of patients with AVM's. As a further benefit, it will facilitate comparison of AVM treatment results between series or between techniques.

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Grading system for arteriovenous malformations


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