Management and clinical outcome of posterior fossa arteriovenous malformations: report on a single-centre 15-year experience

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ABSTRACT

Objectives: Posterior fossa brain arteriovenous malformations (PFbAVMs) are rare lesions. Management is complicated by eloquence of adjacent neurological structures, multimodality treatment is often necessary, and obliteration is not always possible. We describe a 15-year experience in the management of posterior fossa brain AVMs with a focus on clinical outcome.

Methods: From 1989 to 2004, prospectively collected information on 108 patients with diagnosis of a PFbAVMs was obtained. Clinical and angiographic characteristics, management options and complications are described and reviewed to evaluate their impact on final outcome as measured by the Modified Rankin Score (mRS).

Results: Ninety-eight patients were followed for an average of 3.3 years (1–14.6). The male-to-female ratio was 1:1. Ninety-five out of 98 patients (98.9%) were symptomatic at presentation, with 61 (62.2%) intracranial haemorrhages. Sixty-two patients were treated (46 cerebellar, 16 brainstem). Ten haemorrhages occurred in follow-up (4.1%/year). The mRS was obtained in 62 patients and was classified as low (good, mRS $\leq$ 2) or high (poor, mRS $\geq$3). Haemorrhage was the only predictor of poor mRS at presentation (p = 0.0229). A poor clinical outcome was correlated with the presence of AA (p = 0.0276), a poor initial mRS (p < 0.0001) and the number of treatments needed (p = 0.0434). Patients were significantly more likely to improve than to deteriorate over time (p = 0.0201).

Conclusion: The final clinical outcome in PFbAVMs relates directly with the presence of associated aneurysms, number of treatments needed to obliterate the AVM and mRS at presentation. Despite the fact that patients tend to improve after brain AVM haemorrhage, the relationship of MRS at presentation and final outcome suggests that an expedited, more definitive treatment is probably a better choice, especially in patients with good grades after the initial bleeding.

Posterior fossa brain arteriovenous malformations (PFbAVMs) are considered rare lesions, comprising about 5% to 7% of all intracranial arteriovenous malformations (AVMs) in clinical series. Often diagnosed after haemorrhage, management of these lesions is complicated by the concentration of eloquent neurological structures within the small contents of the posterior fossa. The putative factors that may contribute to an increased risk of haemorrhage have been addressed in many publications, sometimes with different results, and the impact of brain AVM haemorrhage in neurological function has also been re-evaluated. Series reporting on results of treatment usually deal with single modality and/or focus mostly on radiological AVM obliteration. However, the major goal of the treatment of brain AVMs is preservation of neurological function. Therefore, probably as important as identifying predictors of haemorrhage or radiological obliteration of the AVM is the identification of factors affecting final clinical outcome. This may be especially true regarding posterior fossa AVMs (PFbAVMs), a subgroup where complete occlusion is not always possible without significant risks. We describe our experience in the management of posterior fossa brain AVMs in one of the largest prospectively collected posterior fossa AVM series. In a subset of patients where mRS information was prospectively available, we attempted to identify factors related to a worse clinical outcome.

METHODS

The Toronto Brain Vascular Malformation Study Group has maintained since 1989 an ongoing prospective database where demographic, angiographic and clinical information regarding patients with intracranial and intraspinal vascular disease is collected. From 1989 to 2004, 678 brain AVM patients with complete and prospectively collected information were included. One hundred and six patients with diagnosis of a posterior fossa AVM confirmed with MRI or angiography were identified. Eight patients were lost for follow-up, and 98 were included in this analysis. Cavernous malformations, dural arteriovenous fistulas and vein of Galen malformation were excluded. Follow-up started at the time of presentation/diagnosis and ended with last visit, AVM occlusion or death.

In a subset of patients where mRS was available (62 patients), we evaluated clinical and angiographic characteristics and management options to identify which factors could influence outcome. Poor outcome was defined as a Modified Rankin Score (mRS) equal to or higher than 3. The following factors were analysed: clinical presentation, initial mRS, eloquence, presence of deep venous drainage (DVS), associated aneurysms (AA), venous stenosis or ectasia, age and treatment. Clinical presentation was divided into haemorrhagic and non-haemorrhagic. AVMs located in the deep cerebellar nuclei, cerebellar peduncles or brainstem were considered eloquent, as opposed to AVMs located in the cerebellar cortex. Venous drainage was classified as deep if
major veins drained towards the deep venous system. Associated aneurysms (AA) were defined as any arterial dilatation in an intracranial vessel measuring at least twice the parent vessel diameter. Further subclassification in intranidal, prenidal and remote was used according to Redekop et al. Size was classified using the Spetzler–Martin\(^6\) classification, and the AVMs with a diameter above 3 cm were grouped together as large, due to the small number of large AVMs. The influence of treatment was analysed considering a dichotomous model (treatment yes/no) and the effects of multimodality management. Univariate analysis and multiple logistic regression were used to evaluate the impact of each factor in the final clinical outcome.

RESULTS
From the 106 patients, eight were lost for follow-up, leaving 98 patients (92.4%) with complete follow-up information. The follow-up time varied from 1 to 14.6 years (average 3.3 years). Patient demographics and AVM characteristics are shown in table 1.

At initial assessment, 95 out of 98 (96.9%) patients were symptomatic, with 61 (62.3%) intracranial haemorrhages. Sixty-two patients (63.3%) were treated (97 procedures), 46 with cerebellar and 16 with brainstem AVMs. In this group, 60 (96.7%) were symptomatic, with 46 (74.1%) haemorrhages. Third-six patients were managed conservatively. From this group, 34 (94.4%) were symptomatic. Treatment information is shown in table 2.

Appropriate prospective mRS classification was available in 62 patients, once it was not used in earlier phases of data collection (fig 1). The following results apply to these patients only. Haemorrhage was strongly correlated with a poorer mRS (p = 0.0229). No significant difference was identified between the treated and untreated patients. There was a trend towards treating more often AVMs with AA (p = 0.0837).

In univariate analysis, the presence of AA (p = 0.0249), a poor initial mRS (p<0.0001) and treatment—single (p = 0.0045) or multimodality (p = 0.0175)—were associated with poor final mRS. Males were less likely than females to have a poor mRS at final follow-up (p = 0.0414). Interestingly, presentation with haemorrhage, although associated with a poor initial mRS, did not correlate directly with a poor final mRS (p = 0.4075). In multiple logistic regression analysis, the institution of treatment was the only factor associated with a poor clinical outcome. Subjects receiving a single treatment are on average 5.79 (95% CI 1.09 to 30.83) times more likely to have a final MRS classification of 3 to 6 than those who did not receive treatment. Subjects receiving multiple treatments are 8.19 (95% CI 1.41 to 47.67) times more likely to have a final MRS classification of 3 to 6 than those who did not receive treatment, after controlling for the other terms in the model. AVMs located in eloquent regions received treatment more often (p = 0.0047) and were also more likely to require multimodality treatment (p = 0.0168).

Thirty-one patients (48%) had a good mRS on initial and final visit, 18 (28%) were classified as poor on both occasions, 12 (19%) started out as poor but improved at the final visit, and only three subjects (5%) started out as good grades and had a poor mRS at final visit. The McNemar test shows that among subjects whose mRS classification changed over the course of the study, improving was significantly more likely than worsening (p = 0.0201). Treatment-related complications occurred in 12 out of 97 procedures (12.4%).

Radiological follow-up was complete in 52 patients (83.9%) at the time of data analysis. Ten patients were still waiting for final imaging after radio-surgery. Thirty (48.9%) AVMs were completely obliterated, seven had a smaller but still patent nidus, and five were unchanged. Ten haemorrhages occurred in follow-up, generating a yearly risk of haemorrhage of 4.1% (excluding haemorrhage at presentation). There was no difference between the yearly risk of haemorrhage in partially treated and untreated AVMs.

DISCUSSION
Despite significant advances, management of brain AVMs is still challenging. Posterior fossa malformations constitute a subset of AVMs where management is further complicated by the large concentration of highly eloquent neurological structures in a confined space, where a neurological deficit either from AVM haemorrhage or treatment complications is more likely and severe.\(^{16–17}\) Series reporting on those lesions are likely to have significant bias, since brainstem AVMs are more likely to be submitted to radio-surgery or not treated, and cerebellar hemisphere AVMs may be amenable to surgical resection and/or endovascular obliteration.\(^{10–14\text{, }18–21}\) Data from Khaw et al\(^2\) and from our own institution\(^2\) showed the association of PFbAVMs with haemorrhagic presentation. This strong association was again confirmed in this subset of patients. This, however, did not translate into a higher risk of haemorrhage in follow-up (4.1%/year in our series). The high rate of haemorrhagic presentation is more likely related to the fact that due to their location, these AVMs are unlikely to manifest by symptoms such as headache or seizures,\(^4\) leaving unruptured, asymptomatic AVMs undiagnosed.

The association between brain AVMs and intracranial aneurysms has been well described, but the analysis of its impact on the risk of haemorrhage resulted in conflicting

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**Table 1 Patient demographics and arteriovenous malformation characteristics**

<table>
<thead>
<tr>
<th>Total no of patients</th>
<th>98</th>
<th>100%</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Demographics</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male:female</td>
<td>53:45</td>
<td>54.6%:46%</td>
</tr>
<tr>
<td>Mean age (range)</td>
<td>38.8 (1 to 79)</td>
<td></td>
</tr>
<tr>
<td><strong>Location</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cerebellar</td>
<td>71</td>
<td>72.4%</td>
</tr>
<tr>
<td>Brainstem</td>
<td>21</td>
<td>21.5%</td>
</tr>
<tr>
<td>Both</td>
<td>6</td>
<td>6.12%</td>
</tr>
<tr>
<td><strong>Presentation</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Haemorrhage</td>
<td>61</td>
<td>62.2%</td>
</tr>
<tr>
<td>Seizure</td>
<td>7</td>
<td>7.14%</td>
</tr>
<tr>
<td>Bruit</td>
<td>2</td>
<td>2.04%</td>
</tr>
<tr>
<td>Headache</td>
<td>55</td>
<td>56.12%*</td>
</tr>
<tr>
<td>Asymptomatic</td>
<td>3</td>
<td>3.06%</td>
</tr>
<tr>
<td><strong>Neurological deficits at presentation</strong></td>
<td>47</td>
<td>47.9%</td>
</tr>
<tr>
<td><strong>Angioarchitecture</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Deep drainage</td>
<td>68</td>
<td>69.4%</td>
</tr>
<tr>
<td>&lt;3 cm</td>
<td>78</td>
<td>79.6%</td>
</tr>
<tr>
<td>3–6 cm</td>
<td>17</td>
<td>17.4%</td>
</tr>
<tr>
<td>&gt;6 cm</td>
<td>3</td>
<td>3.0%</td>
</tr>
<tr>
<td>Associated aneurysms</td>
<td>21</td>
<td>21.4%</td>
</tr>
<tr>
<td>Prenidal</td>
<td>15</td>
<td>15.3%</td>
</tr>
<tr>
<td>Intranidal</td>
<td>4</td>
<td>4.1%</td>
</tr>
<tr>
<td>Remote</td>
<td>2</td>
<td>2.1%</td>
</tr>
</tbody>
</table>

*Includes patients presenting with haemorrhage.
In our series, the presence of an associated aneurysm was an independent predictor of poor mRS at final follow-up. Other series have shown an increased risk of haemorrhage when comparing those AVMs with bAVMs without AA, which may reflect a poorer outcome. Target treatment of these aneurysms, even if the AVM cannot be cured, is at least intuitively attractive, in the hope that this would decrease the risk of future haemorrhage. In our series, however, partial treatment (including a small number of target embolisations of AA) did not translate in a lower risk of haemorrhage. This could perhaps be explained by the large heterogeneity of bAVMs, representing a continuum of lesions with a different prognosis, depending on lesion angioarchitecture and location. There was a trend to treat AVMs with AA more often, aiming at either total obliteration, when possible, or elimination of the aneurysms, perceived as a "weak point" in the AVM. We were not able to access the impact of this specific type of target treatment in the rate of haemorrhage due to the small numbers.

The impact of haemorrhage from brain AVMs has been recently reviewed. Although studies from Columbia suggest a lower morbidity and mortality than previously reported, those series include a small percentage of AVMs located in the posterior fossa. A large proportion of our patients presented with neurological deficits (47 out of 98) and in the subgroup with prospective mRS, 18 (28%) were classified as high mRS (>3), with 12 (66.6%) improving to a good mRS at final visit. Despite a significant relationship between presentation and final mRS score (p < 0.0001), only three subjects (5%) deteriorated over the course of the study. Patients whose mRS classifications changed were significantly more likely to improve than to deteriorate (p = 0.0201).

Prevention of intracranial haemorrhage is the main goal of treatment for bAVMs, and evidence suggests that only complete AVM obliteration is protective. In the posterior fossa, this is sometimes not possible. Despite the recent questioning of the role of treatment for unruptured bAVMs, there is no controversy regarding the appropriateness of treating a ruptured lesion. In our series, the vast majority of the AVMs presented with haemorrhage. However, treatment was instituted in only 60%. In approximately 40% of the cases, the risks of treatment were perceived to be too high, and a conservative management was adopted. Even with this careful selection of patients for treatment, the modified Rankin Scores at final visit were worse in the group of AVMs submitted to treatment, especially multiple treatments. These results, however, should be viewed with care. They reflect the experience of a tertiary care centre with a significant referral bias, and multimodality treatment is more likely to be employed when dealing with more complex AVMs, with the outcomes maybe reflecting a more aggressive disease process, more prone to haemorrhage. Further identification of bAVM subgroups according to risk of haemorrhage and neurological deficit is clearly needed. AVMs located in more eloquent regions of the posterior fossa were more likely to require multimodality treatment (p = 0.0168), also reflecting a higher difficulty level of management. This may also have affected our results, once deficits in more eloquent regions are less likely to improve completely.

Our study has limitations. Despite the fact that the data were collected prospectively, it was retrospectively reviewed with the inherent limitations. Bias in the selection of patients for treatment and therapeutic modalities certainly influenced our results. Series on the results of P/FbAVMs treatment had been published, including results from each of the three modalities of treatment available (radio-surgery, surgery and embolisation) and their combinations. Those series, however, are focused mostly on treated patients, and the outcomes are mainly related to AVM obliteration. No comparison is made with untreated patients at the same institution, and the number of patients seen but not treated is unknown. Our cohort includes conservatively managed patients, adding treatment as a variable and focusing on the final clinical outcome rather than AVM obliteration as a measure of treatment success. A piece of useful information that is missing is how lesions were selected for treatment or not. The reasons why an AVM patient would not be offered treatment were mainly age, associated comorbidities or when the treatment was perceived to carry a very high risk. Unfortunately, there were no clear, objective criteria to define a "high risk" treatment. However, no major difference between the groups (treated and untreated AVMs).

![Figure 1](https://example.com/figure1.png)

**Figure 1** Initial and final modified Rankin Score (mRS) for the whole cohort.
was present, except for the fact that lesions with AA tended to be treated more often.

The single most important measure of success in the management of any disease is clinical outcome. The significant relationship between a poor clinical grade at presentation and a poor outcome demonstrates clearly the importance of preventing neurological damage, especially in such eloquent region. The relationship of initial haemorrhage and poor mRS at presentation is clear, and the latter correlates strongly with a poor final mRS. The increasing availability of high-quality MRI is likely to increase the number of asymptomatic posterior fossa AVMs diagnosed, raising once more the issue of “prophylactic” brain AVM treatment. On the other hand, it is clear that the number of treatments needed to obliterate a lesion has a direct impact on the outcome, probably reflecting the unforgiving nature of complications in highly eloquent brain regions. When possible, expeditious and definitive single modality treatment is probably the best choice of management, especially for patients in good clinical grade.

CONCLUSION

Posterior fossa bAVMs are complex lesions and probably represent a continuum of intracranial vascular disease. Lesions requiring multiple treatments and/or with associated aneurysms are more likely to have a poor final clinical outcome. Overall, patients are likely to improve after brain AVM haemorrhage, but a poor initial modified Rankin Score correlates strongly with poor outcome after haemorrhage from posterior fossa brain AVMs. This association should be taken into account when management decisions are made, and expedited, effective and, if possible, single modality treatment aimed at complete AVM obliteration should be implemented in order to prevent further haemorrhage and deterioration. If obliteration is not expected, partial treatment should probably be avoided, except maybe in very specific cases such as intranidal aneurysms.

Competing interests: None.

REFERENCES

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