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# Endovascular management of pediatric cerebral arteriovenous malformations (AVMs)

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**Abstract**

Cerebral AVMs are congenital vascular abnormalities composed of multiple fistulous connections between arteries & veins without the presence of a normal intervening capillary bed. AVMs in the pediatric population represent 3% of all AVMs. They are usually presented by spontaneous intracerebral haemorrhage followed by recurrent attacks of seizures, intractable headaches, and focal neurological deficits. Several imaging techniques are used for obtaining diagnosis including brain CT scan, CT angiography, MRI, MRA, and DSA. Regarding the management process, several modalities are currently used including conservative management, endovascular therapy (EVT), microsurgery, and stereotactic radiosurgery (SRS). EVT has been proven to be an effective and safe modality for treating cerebral AVMs in the pediatric population showing a high cure rate either as a single treatment modality or as a pre-procedural technique. Despite good post-embolization angiographic and clinical outcomes during the early follow-up period, still longer angiographic and clinical follow-ups are needed for all cases.

**Keywords:** Pediatric cerebral arteriovenous malformations, endovascular embolization, microsurgery, stereotactic radiosurgery

## Introduction

Cerebral arteriovenous malformations (AVMs) are congenital vascular abnormalities composed of multiple fistulous connections between arteries and veins without the presence of a normal intervening capillary bed. This direct Arterio-venous (AV) connection with a lack of capillaries in between leads to hypertrophy in both AV components of the AVM [1]. AVMs can be present at any age with a mean age at diagnosis of 31.5 years. AVMs in the pediatric population represent about 3% of all AVMs and are usually presented with spontaneous intracerebral hemorrhage (ICH) followed by recurrent attacks of seizures, intractable headaches, and focal neurological deficits [2, 3]. Several imaging modalities are used for diagnostic evaluation including CT brain, CT angiography (CTA), MRI, MRA, and DSA which is considered the golden standard method of diagnosis [4]. AVMs are classified through the Spetzler-Martin grading system which is mainly based on the size of the nidus, eloquence of the adjacent brain, and pattern of the venous drainage of the AVM [5]. Complete obliteration is commonly achieved by multimodal therapy that includes microsurgery, endovascular therapy (EVT), and stereotactic radiosurgery (SRS) or a combination of two or more of those techniques [6, 7, 8]. Endovascular embolization showed recently a rapid technical evolution in catheter designs and embolization materials e.g. Onyx and a variety of mixes of NBCA [9, 10]. This continuous progression in the endovascular technique has led to an increase in the use of this procedure as a single modality or as a part of a multimodal process [11, 12].

## Angiographic Anatomy of Cerebral AVMs

### Geometric classification of Feeding Arteries [13-17]

AVM Feeders can be classified anatomically, hemodynamically, and geometrically. Anatomically, feeders may have:

- **Pial supply:** (Cortical or medullary branches).
- **Meningeal supply:** (Direct or through pial anastomoses).
- **Choroidal supply:** (Extra-ventricular or the intra-ventricular portions of choroidal arteries).

Hemodynamically, feeders are classified into dominant and supplementary feeders. Geometrically, feeders can be classified 3 main types including terminal, indirect, and pseudo-terminal feeders.

**1. AVM Nidus and its angioarchitecture [18, 19]**

The nidus is composed of single (mono-compartmental AVMs) or multiple (multi-compartmental AVMs) vascular compartments supplied by a feeding artery and drained into a single draining vein. The nidus may be a pure plexiform nidus consisting of A-V shunting across a plexiform network of vascular channels or a pure fistula (arteriovenous fistula) or both; which is then called a mixed nidus.

**2. Venous Angio-architecture of cerebral AVMs [20]**

Veins draining an AVM can be classified into 3 main types: pure cortical AVMs (sulcal & gyral type)→ drain into the adjacent dural venous sinus through cortical veins, cortical AVMs with ventricular or subcortical extensions→ drain into both superficial cortical and deep veins and deep AVMs→ drain into subependymal deep veins.

### Classification of Cerebral AVMs [21, 22]

**Table 1:** Classification of cerebral AVMs according to location

| Superficial (Cortical) AVMs | Deep AVMs |
|---|---|
| Sulcal type. | Sub-arachnoid type. |
| Gyral type. | Deep parenchymal type. |
| Mixed type (sulco-gyral) | Plexal type. |
| | Mixed type. |

### Grading systems for Cerebral AVMs

### Spetzler-Martin (SM) grading system

SM grading system is the most practical, widely used grading system for cerebral AVMs [1]. According to the system, AVM grades I, II, or III have low treatment-associated morbidity (low-grade AVMs). On the other hand, grades IV and V lesions were found to have a much higher morbidity rate up to 31% and 50% in G IV & V respectively (high-grade lesions) [22, 23, 24].

**Table 2:** Spetzler-Martin system grading system for an AVM [23]

| Characteristic of lesion | Points |
|---|---|
| **Nidal size** | |
| 1. Small (diameter < 3 cm). | 1 |
| 2. Medium (diameter 3-6 cm). | 2 |
| 3. Large (diameter > 6 cm). | 3 |
| **Eloquence of adjacent brain (location)** | |
| 1. Non-eloquent site. | 0 |
| 2. Eloquent site. | 1 |
| **Pattern of venous drainage** | |
| 1. Superficial drainage only. | 0 |
| 2. Any deep drainage. | 1 |

### Clinical Presentations of Cerebral AVMs [25-28]

1. **Intracerebral haemorrhage (ICH):** The most common clinical presentation of cerebral AVMs involving more than 70% of cases. Haemorrhages may be parenchymatous (41%), subarachnoid (24%), intra-ventricular (12%) or combination of any of the previous types (23%).
2. **Seizures:** The 2nd most common presentation of cerebral AVMs. Pathophysiology of seizures can be explained by several hypothesises including vascular steal, cortical irritation, venous congestion, neuronal loss & glial proliferation.
3. Headache.
4. Focal neurological deficits.
5. Asymptomatic lesions.

### Diagnosis of Cerebral AVMs

1. **Brain CT scan:** An initial screening tool which can show a ICHs, parenchymal edema, mass effect, ischemic changes, and the presence of hydrocephalus [2].
2. **CT Angiography of the brain (CTA):** CTA is considered to be a non-invasive, safe, and rapid diagnostic imaging tool for cerebral AVMs. CTA also provides 3D reconstruction images to help understand the vascular anatomy of the lesion and its components [29].
3. **Brain MRI:** Provides more specific data about the angio-architectural features of an AVM including its morphology (sponge-like structure, formed of tightly packed blood vessels and patchy signal voids), the vascular anatomic relations of the nidus, mass effect and ischemic changes [18].
4. **Brain (MRA) [30]:** A non-invasive imaging tool used to evaluate intracranial vascular lesions with a technique based upon the flow relationships more than the anatomical images of the cerebral blood vessels. Several MRA cross-sectional imaging techniques like three-dimensional time of flight MRA (3D TOF-MRA), 3D contrast-enhanced MRA (CE-MRA) & time-resolved MRA (TR-MRA) have been applied recently in clinical use.
5. **Digital subtraction angiography (DSA):** The golden standard diagnostic tool for cerebral AVMs providing precise data about the lesion's characteristics because it provides the highest temporospatial resolution of all previously mentioned diagnostic tools [20].

### Management of Cerebral AVMs

**1. Endovascular Embolization**

**The role of endovascular therapy (EVT) in the management of cerebral AVMs can be summarized in five main scenarios [9]:**

- **Curative embolization:** Cerebral AVMs can be cured using EVT alone. The published embolization cure rates range from 9.7% to 25% [11]. The chance of cure rate is inversely proportional to the AVM size and the number of its feeders [31].
- **Pre-operative embolization:** It is frequently performed before microsurgical resection aiming at nidal filling with occlusion of the arterial feeders lying deep within the brain parenchyma, thus minimizing the risk of surgery (with a mean nidal occlusion efficacy of approximately 72%) [32].
- **Pre-radiosurgery embolization:** Performed to reach certain goals including; decrease the nidal size into a single smaller target, obliteration of large A-V fistulae

which are more refractory to the effects of radiosurgery and elimination of high-risk features of the AVM such as nidal aneurysms [33].

- **Targeted embolization:** Performed to occlude high-risk lesions such as intranidal or flow-related aneurysms [34].
- **Palliative embolization:** Can be done in selected circumstances to relieve symptoms caused by arterial steal phenomenon or venous hypertension thus improving the perfusion pressure in the surrounding brain parenchyma [35].

**2. Microsurgery**

Microsurgery has shown the highest rates of complete nidal obliteration/ removal plus hematoma evacuation in cases of acute ruptured cases [36] with an overall obliteration rate ranging from 65 to 100%, and with complication rates ranging from 5 to 33% [15]. Including haemorrhage, brain tissue damage, vasospasm, seizures, vascular thrombosis, and stroke. Significant evidence of recurrence (5.5-14.4%) has been reported with paediatric AVMs, even in cases with complete microsurgical resection [37].

**3. Stereotactic Radiosurgery (SRS)**

SRS major platforms are Gamma knife, modified LINACs and particle beam units. The current and most widely used technique for AVM radiosurgery is Gamma knife radiosurgery. The radiation implied on the lesion causes changes in cellular components of the blood vessels resulting in progressive obliteration of their lumens [33]. The choice of this modality for the paediatric population was restricted for fear of the drawbacks of radiation therapy over growing brain tissues in children [8]. Studies showed that the overall rate of obliteration following SRS range from 72% to 85% with certain complications reported in 10% of cases including cyst formation, memory deficits, cognitive deficits, encapsulated hematoma, and radiation-induced tumour [38, 39].

**4. Multimodality treatment**

Multi-modality treatment is usually considered for cases with large and complex cerebral AVMs. The 1st and most common combination applied includes EVT followed by microsurgical resection. The 2nd combination applied includes EVT followed by SRS providing an obliteration rate of approximately 40–83%. The 3rd combination includes SRS followed by micro-surgery or multiple sessions of EVT [40, 41].

## Conclusion

The optimal management for pediatric cerebral AVMs remains controversial. EVT has been proven to be an effective and safe modality for treating pediatric cerebral AVMs as it showed a high cure rate either as a single treatment modality or as a pre-procedural technique facilitating other treatment modalities. Despite good post-embolization angiographic & clinical outcomes during the early follow-up period, still longer angiographic and clinical follow-ups are needed for all cases especially those presented with intracranial hemorrhage because initial complete obliteration is not a guarantee for permanent follow-up.

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