# Management of Arteriovenous Malformations Associated with Developmental Venous Anomalies: A Literature Review and Report of 2 Cases

*Michael Zhang[1], Ian D. Connolly[1], Mario K. Teo[1,2], George Yang[3], Robert Dodd[1], Michael Marks[1], Mario Zuccarello[3], Gary K. Steinberg[1]*

■ BACKGROUND: **Classification of cerebrovascular malformations has revealed intermediary lesions that warrant further review owing to their unusual presentation and management. We present 2 cases of arteriovenous malformation (AVM) associated with a developmental venous anomaly (DVA), and discuss the efficacy of previously published management strategies.**

■ METHODS: **Two cases of AVMs associated with DVA were identified, and a literature search for published cases between 1980 and 2016 was conducted. Patient demographic data and clinical features were documented.**

■ RESULTS: **In case 1, a 29-year-old female presenting with parenchymal hemorrhage and left homonymous hemianopia was found to have a right parieto-occipital AVM fed from the anterior cerebral, middle cerebral, and posterior cerebral arteries, with major venous drainage to the superior sagittal sinus. In case 2, imaging in a 34-year-old female evaluated for night tremors and incontinence revealed a left parietal AVM with venous drainage to the superior sagittal sinus. Including our 2 cases, 22 cases of coexisting AVMs and DVAs have been reported in the literature. At presentation, 68% had radiographic evidence of hemorrhage. Stereotactic radiosurgery was performed in 7 cases, embolization in 6 cases, surgical resection in 4 cases, and multimodal therapy in 5 cases. Radiography at follow-up demonstrated successful AVM obliteration in 67% of cases (12 of 18).**

■ CONCLUSIONS: **Patients with coexisting AVMs and DVAs tend to have a hemorrhagic presentation. Contrary to traditional AVM management, in these cases it is important to preserve the draining vein via the DVA to ensure a safe, sustained circulatory outflow of the associated brain parenchyma while achieving safe AVM obliteration.**

## INTRODUCTION

The commonly recognized subgroups of cerebrovascular malformations include arteriovenous malformation (AVM), capillary telangiectasia, cavernous malformation, and developmental venous anomaly (DVA).[1] The estimated incidence of AVM in population-based studies is generally approximately 1 in 100,000.[2] AVM occurs when a collection of arteries and veins forms without an intervening capillary bed. This results in a high-pressure shunting of blood, thought to be responsible for the symptomatic presentation of hemorrhage at an annual rate of 2%–3%.[2-5]

In comparison, a DVA usually appears as a collection of radiating veins converging on a large and centrally located draining vein. This is classically described as having a fan-shaped "caput medusae" appearance, although this is not always seen.[6] Histologically, the DVA consists of a collection of thickened and hyalinized veins with little smooth muscle and elastic tissue.[1] DVA is the most common cerebrovascular abnormality, with an overall incidence of 2%–4%,[7-11] much more common than AVMs. However, DVAs, usually discovered incidentally, rarely if ever bleed, and the

**Key words**
- Arteriovenous malformation
- Coexisting malformation
- CyberKnife radiosurgery
- Developmental venous anomaly
- Glue embolization
- Multimodal treatment
- Stereotactic radiosurgery

**Abbreviations and Acronyms**
**ACA**: Anterior cerebral artery
**AVM**: Arteriovenous malformation
**DVA**: Developmental venous anomaly
**MCA**: Middle cerebral artery
**MRI**: Magnetic resonance imaging
**PCA**: Posterior cerebral artery

*From the [1]Department of Neurosurgery and Stanford Stroke Center, Stanford University Medical Center, Stanford, California, USA; [2]Department of Neurosurgery, Bristol Institute of Clinical Neuroscience, North Bristol University Hospital, Bristol, UK; and [3]Department of Neurosurgery, Mayfield Brain & Spine, Cincinnati, Ohio, USA*

*To whom correspondence should be addressed: Gary K. Steinberg, M.D., Ph.D. [E-mail: gsteinberg@stanford.edu]*

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reported annual bleeding rate of 0.2%–0.3% is likely due to bleeding from coexisting cavernous malformations.[12,13]

Despite their contrasting properties, the convergence of AVM and DVA presents a tenuous hemodynamic balance that warrants neurosurgical intervention. Presumably, when the DVA's low resistance drainage system receives a high-pressure inflow, the bleeding risk exceeds that associated with either entity alone. Perhaps the additional strain on the hemodynamic balance from increased inflow leads to the commonly reported symptoms of coexisting AVMs and DVAs, including headache, seizures, and neurologic deficits.[14]

Given the ongoing need to understand the clinical manifestations and management of these colocalizing cerebrovascular malformations, we present 2 cases of coexisting AVM and DVA from our institutions. In 1984, Huang et al.[15] reported the first case of a coexisting AVM and DVA. To aid in the treatment of future cases, we also review the decision making and outcomes reported in the literature for this rare transitional malformation.

## MATERIALS AND METHODS

We identified cases of coexisting AVM and DVA from a locally held institutional database at Stanford University. In addition, M.Z. and I.D.C. conducted a literature search for cases published between 1980 and 2016 using the PubMed database to identify case reports of this unique entity. The following terms were used: arteriovenous malformation and developmental venous anomaly, AVM and DVA, venous angioma and AVM, venous malformation and AVM, and associated DVA. We also reviewed the references of these journal articles for additional reports. M.Z., M.K.T., and I.D.C. reviewed and extracted data from the studies. Clarification of information from the literature was sought from senior authors when appropriate.

Patient demographic data and clinical features were examined, including clinical and radiographic presentation and outcomes, feeding artery distribution, AVM location, draining vein termination site, and treatment modality. Univariate analyses were performed with Stata statistical software (StataCorp, College Station, Texas, USA), using the appropriate tests. A $P$ value $< 0.05$ was considered statistically significant.

## RESULTS

### Case 1

A 29-year-old female who had sustained an intracerebral hemorrhage presented with an episode of right-sided headache and partial left homonymous hemianopsia. Computed tomography scan and magnetic resonance imaging (MRI) of the brain demonstrated rupture of the right parieto-occipital AVM component. Angiography demonstrated a 3.5-cm Spetzler–Martin grade IV mixed AVM fed from the right anterior cerebral artery (ACA), middle cerebral artery (MCA), and posterior cerebral artery (PCA), with primary venous drainage into a high-flow DVA. Functional MRI demonstrated the AVM did not involve the calcarine or motor cortices, and magnetic resonance tractography confirmed that the AVM nidus was located adjacent to but not intimately involving the corticospinal and geniculocalcarine tracts (Figure 1).

Despite the high-grade lesion, in view of the patient's history of rupture, the decision was made to proceed with staged multimodal treatment. The first stage involved embolization of the 2 pericallosal artery pedicles, resulting in angiographic occlusion of one-third of the AVM nidus. After the embolization, the decision was made to perform radiosurgery instead of microsurgery owing to the size and eloquence of the AVM. A second angiogram performed 2 months later demonstrated interval (asymptomatic) occlusion of the right ACA supply to the AVM. The patient was treated with single-fraction frameless stereotactic radiosurgery at 10 months after presentation and tolerated the procedure without difficulty. At last follow-up (18 months since presentation), she reported decreased frequency and severity of headaches with improvement in visual symptoms. MRI and magnetic resonance angiography also showed reduction in the AVM nidus with preservation of the DVA.

### Case 2

A 34-year-old right-handed female initially presented to an outside hospital with hemisensory changes and weakness. Computed tomography scan and MRI of the brain revealed a left parietal occipital AVM. Embolization was attempted, but was determined to be unsafe. The patient was seen at Stanford Health Care 2 months later, where her husband reported jerking movements of her right arm during sleep and nocturnal incontinence, which was concerning for seizures. Angiogram showed an extremely diffuse high-flow, Spetzler–Martin grade III AVM approximately 4 cm in diameter associated with a large DVA. It was supplied by multiple enlarged branches from the left MCA and PCA, with terminal drainage into the superior sagittal sinus (Figure 2).

Embolization was attempted once again, but was not completed because of the diffuse vasculature. The AVM component was eventually treated via partial resection of several of the AVM feeders with adjuvant CyberKnife radiosurgery, taking care to leave the DVA component undisturbed. After surgery, the patient's hemisensory changes, weakness, and seizures resolved. At a 14-month follow-up, she was neurologically normal and had remained seizure-free.

### Previous Reports

Including our 2 cases presented here, 22 cases of coexisting AVM and DVA have been described in the literature.[6,15-26] Clinical and demographic data for all reported cases are summarized in Table 1. The mean patient age was 30 years, and 12 of the 22 patients (55%) were male. The mean duration of follow-up was 30 months (range, 4–161 months). Fifteen patients (68%) presented with hemorrhage, and in all patients in whom the site of bleeding was identified radiographically, the bleeding was localized to the AVM component. There was no significant difference in patient age by sex or event of hemorrhage. Eight of 12 patients (66.6%) with superficial venous drainage and 6 of 8 patients (75%) with deep venous drainage had a hemorrhagic presentation ($P = 0.54$, Fisher's exact test). Therefore, the venous drainage pattern and hemorrhagic risk are comparable.

Among the 17 patients with complete documentation of clinical presentation, 11 (65%) experienced new-onset weakness. Loss of consciousness (13.6%) and seizures (10%) were reported less frequently. Radiographically, the site of AVM was distributed in the parietal, frontal, cerebellar, and temporal territories in 32%, 32%, 23%, and 18%, respectively. Meanwhile, feeding arteries