Intracranial Dural Arteriovenous Fistulae
Clinical Presentation and Management Strategies

Timothy R. Miller, MD; Dheeraj Gandhi, MD

Dural arteriovenous fistulae (DAVF) are a rare type of acquired intracranial vascular malformation consisting of a pathological shunt located within the dura matter of the brain. In contradistinction to brain arteriovenous malformations, DAVFs do not harbor a focal nidus. These lesions may arise anywhere along the dura, but most commonly are found in the region of the transverse, sigmoid, and cavernous sinuses. DAVF are typically supplied by meningeal arteries and exhibit venous drainage either directly into the dural venous sinuses or via cortical and meningeal veins. Larger, more complex lesions may recruit pial arterial supply. Although a single lesion is present in a majority of patients, multiple shunts can occur in ≤8%.7

DAVF can present with a myriad of clinical signs and symptoms. In general, it is the location and most importantly the venous drainage pattern of DAVF that determines their clinical presentation and potential for serious sequelae. Management strategies for DAVF are consequently guided by these features and include conservative management, as well as endovascular and surgical treatments. This article will begin by reviewing the epidemiology, natural history, and classification of these lesions. Subsequently, clinical presentations, imaging characteristics, and treatment of DAVF will be discussed.

Epidemiology and Pathophysiology

DAVF account for only 10% to 15% of intracranial vascular malformations but are slightly over-represented in the posterior fossa (35% of such lesions). DAVF are typically encountered in middle-aged adults with a median age of onset in the sixth decade. Although there is a female predominance, DAVF in men are more likely to display aggressive neurological symptoms and present with hemorrhage. Rarely, DAVF may be encountered in the pediatric population, where these lesions tend to be more extensive.

The cause of DAVFs remains uncertain. However, these lesions have been associated with dural sinus or venous thrombosis, the causal role of venous thrombosis is supported by the association of DAVF with hypercoagulable states, such as hyperhomocysteinemia, factor V Leiden, and antithrombin, protein C and S deficiencies. However, it is important to note that dural sinus thrombosis or occlusion can also be a sequelae of DAVF, likely resulting from associated venous hypertension. Finally, although many cases of DAVF are sporadic with no identifiable cause, some patients will report a history of intracranial surgery, infection, radiation exposure, pregnancy, or trauma.

Dural sinus or venous thrombosis may promote the formation of DAVF by 2 pathophysiologic mechanisms. First, occlusion of a dural sinus or cerebral vein may cause alterations in local hemodynamics, allowing for the opening of already present, but dormant, small arteriovenous shunts within the dura. Alternatively, venous thrombosis may result in abnormalities in local vascular growth factors, which subsequently incite the development of the fistula by neoangiogenesis. These potential mechanisms are not mutually exclusive, and it is possible that both contribute to the development of DAVF.

Natural History and Classification

The natural history of DAVFs can broadly be characterized by benign or aggressive behavior based on their venous drainage. Fortunately, most DAVF without aggressive angiographic features will follow a benign clinical course with a low risk of serious sequelae. However, DAVF demonstrate that venous drainage directly into leptomeningeal veins, or retrogradely to cortical veins from an involved dural sinus, is associated with intracranial hemorrhage and nonhemorrhagic neurological deficits (NHND). Malik et al were one of the first groups to identify this cortical venous drainage (CVD) as the main risk factor for serious sequelae from DAVF, which has been confirmed by subsequent reports. DAVF with direct or exclusive leptomeningeal drainage may be particularly prone to an aggressive presentation.

Reported rates of hemorrhage, NHND, and mortality associated with DAVF exhibiting CVD have varied, possibly because of the small number of patients available for such studies. Davies et al noted annual rates of hemorrhage, NHND, and mortality of 19.2%, 10.9%, and 19.3%, whereas Shin et al found that these figures to be 4.5%, 7.2%, and 11.6%. Finally, van Dijk et al reinforced the poor long-term natural history of these lesions by reporting rates of intracranial hemorrhage, NHND, and mortality of 35%, 30%, and 45% in nontreated or partially treated DAVF, exhibiting CVD (n=20) over a mean follow-up of 4.3 years.
CVD, in turn, has been associated with stenosis or occlusion of adjacent dural sinuses, high-flow shunts, and lesion location. Location likely influences the development of CVD because of the lack of adjacent dural sinuses to provide venous drainage in certain sites, such as the anterior cranial fossae and tentorium. DA VF at these locations are, therefore, more prone to aggressive presentations. For example, Malik et al noted a hemorrhagic presentation in 51% (20 of 39) of DA VF located remote from a dural sinus, whereas Lawton et al found that this figure to be 55% (17 of 31) for tentorial DA VF. However, no location in the intracranial compartment is completely immune to aggressive behavior. Furthermore, DA VF are dynamic lesions, and benign fistulae may occasionally evolve into aggressive lesions with CVD, often secondary to venous stenosis or thrombosis, or occasionally increased arterial supply. Often, this transformation is heralded by a change in patient symptomatology. Satomi et al identified 117 consecutive patients with benign DA VF that were either conservatively followed (n=73) or treated palliatively (n=44). Of 112 patients with available follow-up, 110 (98.2%) experienced a benign disease course, whereas 2 developed CVD because of progressive venous outlet obstruction. In a smaller, retrospective study, Shah et al calculated an annual rate of conversion to a higher grade DA VF of 1%

Although there have been many classification schemes suggested for DA VF, the 2 proposed by Borden and Cognard are the most widely used. The Borden classification system (Table 1) categorizes DA VF based on the site of venous drainage (dural sinus versus cortical vein) and the absence or the presence of CVD. The Cognard system (Table 2) not only notes these features but also takes into account the direction of flow in an involved dural sinus (antegrade versus retrograde), as well as the absence or presence of venous ectasia(s) in recruited cortical veins.

Clinical Presentations
Clinical manifestations of DA VF vary depending on their location, arterial supply, degree of arteriovenous shunting, and most importantly, their venous drainage pattern. DA VF lacking CVD may be asymptomatic, or present with symptoms related to increased dural sinus blood flow, such as pulsatile tinnitus, the latter particularly common for transverse and sigmoid sinuses lesions. Generalized central nervous system symptoms that may be related to venous hypertension or cerebrospinal fluid malabsorption, while resulting cranial nerve palsies, are often because of an arterial steal phenomenon or occasionally mass effect from an enlarged arterial feeder. In addition, DA VF involving the cavernous sinus may present with orbital symptoms, including chemosis, ptosis, ophthalmoplegia, and decreased visual acuity.

DAVF with CVD typically have more aggressive clinical presentations, including the sudden onset of severe headache, seizures, NHND, and intracranial hemorrhage, including intraparenchymal, subarachnoid, and subdural hemorrhage. In a meta-analysis, Lasjaunias et al reviewed 195 cases of DA VF and found that focal neurological deficits were related to the presence of associated CVD and venous congestion in the affected vascular territory. Less common aggressive presentations include brain stem or cerebellar dysfunction secondary to venous congestion, parkinsonism-like symptoms, and focal neurological deficits.

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<thead>
<tr>
<th>Type</th>
<th>Venous Drainage</th>
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<tr>
<td>1</td>
<td>Dural Sinus</td>
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<tr>
<td>2</td>
<td>Dural Sinus with associated CVD</td>
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<tr>
<td>3</td>
<td>Cortical Veins (CVD)</td>
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CVD indicates cortical venous drainage.
extra-axial hemorrhage in the cervical spine, as well as cervical and upper thoracic myelopathy.3,38,40–42 DAVF with extensive arteriovenous shunting, particularly in the setting of dural sinus thrombosis, can result in impaired venous drainage from the brain and the global venous hypertension.3,44 This can lead to cerebral edema, encephalopathy, and cognitive decline.3,44,47–49

### Diagnosis and Imaging

The diverse range of clinical presentations of DA VF can make their diagnosis a challenge. Furthermore, common symptoms of DAVF, such as headache and tinnitus, are nonspecific and most often because of another cause. However, in patients presenting with intracranial hemorrhage or unexplained neurological deficits, the clinician must have a high level of suspicion for DAVF. This is also true for patients with objective pulsatile tinnitus (bruit auscultated on examination), which most often has a vascular cause because of increased or turbulent dural sinus flow.

Noncontrast computed tomography (CT) and conventional magnetic resonance (MR) imaging often seem unremarkable with benign DA VF.50 However, these studies may demonstrate the sequelae of aggressive lesions with CVD, including hemorrhage, venous congestion with edema, venous aneurysms, tortuous cortical veins in a pseudophlebitic pattern, and parenchymal or leptomeningeal enhancement (Figure 2).50–52 With larger fistulae, CT or MR can demonstrate prominent vessels or flow voids associated with a dural sinus.1,50–52 Cross-sectional imaging may also reveal engorged orbital veins and proptosis with cavernous sinus DAVF.51 Finally, color-coded duplex sonography has also been successfully used to evaluate DA VF and their response to treatment.53,54

CT and MR angiography (CTA and MRA, respectively) may be used to screen patients suspected of harboring DA VF, grade or classify these lesions when detected, as well as evaluate for response to treatment (Figure 3).51,55–58 Both CTA and MRA may visualize the fistula itself as prominent vessels associated with the meninges or dural sinus wall, as well as detect enlarged feeding arteries, early dural sinus opacification, and prominent draining veins.51,55–58 Narvid et al58 evaluated CTA imaging of a small group of patients with known DA VF presenting with pulsatile tinnitus and found that the technique was both sensitive (86%) and specific (100%) for the detection of associated arterial feeders. Kwon et al51 similarly evaluated the sensitivity of time-of-flight MRA for the detection of DA VF, and found that the technique had a sensitivity of 91% (n=11) for the detection of abnormal venous flow-related enhancement.

More recent imaging advances have allowed for the development of time-resolved CTA and MRA, which tracks the passage of a contrast bolus through the cerebrovasculature.57,59–61

### Cognard Classification of Dural Arteriovenous Fistulae

<table>
<thead>
<tr>
<th>Type</th>
<th>Venous Drainage</th>
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<tbody>
<tr>
<td>I</td>
<td>Venous drainage into dural sinus with antegrade flow</td>
</tr>
<tr>
<td>II a</td>
<td>Venous drainage into dural sinus with retrograde flow</td>
</tr>
<tr>
<td>II b</td>
<td>Venous drainage into dural sinus with antegrade flow and CVD</td>
</tr>
<tr>
<td>II a+b</td>
<td>Venous drainage into dural sinus with retrograde flow &amp; CVD</td>
</tr>
<tr>
<td>III</td>
<td>Venous drainage into cortical veins (CVD)</td>
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<tr>
<td>IV</td>
<td>CVD with associated venous ectasia(s)</td>
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<tr>
<td>V</td>
<td>Venous drainage into spinal perimedullary veins</td>
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CVD indicates cortical venous drainage.
These techniques allow for differentiation of arterial and venous phases similar to catheter angiography and may permit better visualization of early arteriovenous shunting into a dural sinus. Willems et al used time-resolved CT angiography to successfully detect and classify 90% of a small group of DAVF (n=11) compared with catheter angiography. Similarly, Meckel et al found that time-resolved dynamic MRA correctly identified the presence and lateralization of all DAVF in a small group of patients with no false-positives. Pekkola et al compared another dynamic, time-resolved MRA technique for the detection of arteriovenous shunting in a small group of posterior fossa DAVF patients (n=19) and reported 94.4% sensitivity and 83.3% specificity compared with 64.7% sensitivity and 80% specificity for time-of-flight MRA.

Despite these advances in MR and CT imaging, catheter angiography remains the definitive imaging study for evaluation of DAVF because of its superior spatial and temporal resolution. Catheter angiography can delineate both the arterial supply and the venous drainage of the fistula, as well as identify high-risk features including CVD, venous outflow obstruction, and arterial pedicle or venous aneurysms. Catheter angiography is also excellent for evaluation of any associated dural venous sinus thrombosis or occlusion. Finally, the information provided by catheter angiography is essential for planning endovascular or surgical treatment.

Management Strategies

General Considerations and Conservative Management

Management of DAVF should be based on patient characteristics, symptom severity, and risk of serious sequelae, the latter being primary determined by the presence or absence of cortical venous reflux. DAVF without high-risk features may be managed conservatively with an acceptably low rate of serious complications. In these instances, treatment should be tailored to palliation of intolerable symptoms. Complete fistula obliteration may not be necessary although any residual lesion could potentially grow and recruit new arterial supply. Spontaneous thrombosis of DAVF may occasionally occur, most often involving slow flow cavernous sinus lesions. Patient carotid self-compression may help promote fistula closure in a minority of such cases. However, patients with benign DAVF electing conservative management should undergo clinical and imaging follow-up given the small risk of conversion to an aggressive lesion.

Alternatively, DAVF with severe clinical presentations or high-risk angioarchitecture features should be promptly treated. Here, the goal is complete fistula closure although elimination of aggressive features such as CVD or venous ectasias may be sufficient to prevent significant morbidity and mortality. Fortunately, DAVF are often completely curable by endovascular, and less commonly by surgical methods, with consequent reversal of symptoms and subsequent risk of hemorrhage. However, fistula treatment may be easier and pose lower risk when the lesion is lower grade.

Endovascular Therapy

Endovascular embolization has become the primary treatment of DAVF. The goal of endovascular therapy is obliteration of the site of fistulization between feeding arteries and draining veins. In contradistinction to cerebral arteriovenous malformations, this can safely be accomplished by occlusion of the fistula’s draining vein(s), which often results in complete

Figure 3. Elderly men with acute change in mental status.

A. Axial noncontrast computed tomographic (CT) imaging demonstrates subarachnoid hemorrhage (white arrows) centered along left frontal and temporal lobe sulci.

B. Axial delayed phase CT angiography imaging demonstrates multiple prominent enlarged cortical and meningeal veins (white arrows) along the left cerebral hemisphere.

C. Reconstructed 3-dimensional image from delayed phase CT angiography better demonstrates the enlarged meningeal veins with a venous ectasia also noted (white arrow).

D. Lateral view from a right vertebral artery angiogram demonstrates innumerable transosseous muscular and meningeal feeders (white arrows) arising form the distal right vertebral artery supplying a posterior fossa Borden type 3/Cognard type II a+b DAVF. There is retrograde opacification of the superior sagittal, transverse, and sigmoid sinuses (black arrows), as well as the deep venous system (black asterisk).
closure of the lesion. However, partial embolization, either by proximal arterial occlusion or incomplete venous drainage closure, may temporarily alleviate symptoms, but it is unlikely to result in a long-term cure. In addition, partial occlusion could negatively alter the venous drainage pattern, potentially inducing CVD. Complete lesion closure may sometimes not be possible in a single treatment session when the fistula is large, and staged treatment is sometimes required.

Endovascular embolization of DAVF can be accomplished using particulates, microcoils, or liquid embolic agents. Access to the lesion may be transarterial, transvenous, or both. In addition, some authors have treated transverse and sigmoid sinus DAVF by angioplasty and placement of an endovascular stent into the involved sinus (most often when the latter has become stenotic or occluded). This technique may treat DAVF by restoring normal venous flow, as well as by closing arteriovenous shunts in the dural sinus wall. Although endovascular embolization has generally been shown to be safe and effective, potential complications include nontarget embolization resulting in stroke or blindness, vessel injury, and intracranial hemorrhage. Because DAVF often involve branches of the external carotid artery, it is imperative that treating physicians remain cognizant of potential dangerous anastomoses between the external and the internal carotid artery systems, as well as the normal external carotid artery supply to cranial nerves.

Transvenous Embolization

Transvenous embolization is performed by retrograde catheterization of the involved draining vein or dural sinus with a microcatheter, which is then subsequently closed by placement of multiple detachable microcoils or liquid embolic agent (Figure 4). Transvenous embolization is ideal when the involved draining veins or dural sinus do not contribute to normal cerebral venous outflow. Under these circumstances, the diseased venous drainage may be euthanized with little risk of resulting venous infarct. Transvenous embolization is also helpful to treat extensive DAVF that are supplied by multiple arterial pedicles, which may make a transarterial approach challenging. Rates of complete fistula obliteration by this technique are high, with reported ranges of 71% to 87%. Specific risks include venous infarction and dural sinus perforation.

DAVF involving the cavernous sinus are particularly amenable to transvenous embolization, with high rates of successful lesion closure. Here, the cavernous sinus may be approached via the inferior petrosal sinus, facial and superior ophthalmic veins, or contralateral cavernous sinus. Rarely, direct percutaneous puncture of the cavernous sinus or an orbital vein may be used if other venous access routes are unavailable. The technique is well tolerated although cavernous sinus embolization may result in cranial neuropathies or worsening of the venous drainage pattern when fistula closure is incomplete.

Transarterial Embolization

Transarterial embolization of DAVF is both safe and effective and is currently the preferred endovascular approach to such lesions. Furthermore, in cases where draining veins are stenotic or tortuous, or when there is common venous drainage with normal brain parenchyma, transarterial access may be the only safe route to the fistula. Transarterial treatment...
was first attempted using particulate embolization, which involves navigation of a microcatheter into a feeding artery followed by injection of a mixture of particulates and contrast into the lesion. However, this technique has now fallen out of favor because closure of treated vessels is often temporary, with reconstitution of the fistula expected in weeks to a few months.\(^{1,66}\) There is also the theoretical risk of systemic venous emboli.

Because of the high rate of fistula recurrence using particulates, transarterial embolization of DAVF is now typically performed with liquid embolic agents, such as n-butyl cyanoacrylate (n-BCA) or ethylene vinyl alcohol copolymer (Onyx). Liquid embolic treatment can result in permanent fistula closure if the agent penetrates the lesion, crossing from artery to proximal draining vein. To achieve this, it is essential to wedge the microcatheter as distally as possible in the feeding artery, otherwise, proximal arterial occlusion with eventual lesion reconstitution is likely.\(^{74}\) In cases of suboptimal catheter position, fistula penetration may be achieved by creation of an Onyx plug around the microcatheter tip, or distal delivery of NBCA by the sandwich and D5 push techniques. Specific risks of liquid embolic embolization include microcatheter entrapment, retrograde penetration of noncatheterized arterial pedicles, and excessive venous penetration, which may result in worsening of venous hypertension or venous infarction.\(^{1}\)

With high flow lesions, the latter may be avoided by the adjuvant placement of coils before liquid embolic injection to slow the rate of arteriovenous shunting.\(^{1}\)

Outcomes of liquid embolic treatment of DAVF have been favorable.\(^{8,23,74–79}\) Guedin et al\(^{79}\) reported successful fistula closure of 83% (35 of 42) of DAVF with CVD primarily treated by n-BCA embolization, whereas the remainder had suppression of CVD. There was no permanent morbidity or mortality. Wakhloo et al\(^{85}\) reported complete occlusion of 14 complex indirect carotid cavernous fistula treated primarily with n-BCA, again with no permanent morbidity or mortality. Finally, De Keukeleire et al\(^{80}\) noted an 85.7% immediate occlusion rate of DAVF treated transarterially with Onyx, whereas Hu et al\(^{85}\) reported a 79% (50 of 63 DAVF) rate of angiographic cure after transarterial Onyx embolization, with only 1 case of permanent morbidity secondary to cranial nerve palsies.

**Surgical Approaches**

Microsurgical repair is a safe and effective treatment method for DAVF, either alone or in combination with endovascular embolization.\(^{23,80–82}\) Surgical treatment methods for DAVF include surgical excision of involved meningeal arteries and veins, packing of the diseased dural sinus, as well as skeletonization of the involved dural sinus with disconnection of draining leptomeningeal veins.\(^{10,32,81}\) A surgical approach can be ideal for anterior cranial fossa DAVF that are supplied by meningeal branches arising from the ophthalmic artery, which typically are not favorable for endovascular treatment. Risks of surgical repair of DAVF include blood loss, intracranial hemorrhage, arterial infarct, venous infarct, and cerebrospinal fluid leakage.\(^{83,84}\) Given recent advances in endovascular treatment methods of DAVF, surgical repair is often reserved for lesions that fail, or are not amenable to, endovascular therapy.

**Stereotactic Radiosurgery**

Stereotactic radiosurgery, or gamma knife surgery (GKS), has been successfully used to treat DAVF, either alone or in combination with other treatment methods, such as endovascular embolization.\(^{85–89}\) A recent meta-analysis by Chen et al\(^{88}\) including 19 studies and 729 patients who underwent GKS for DAVF demonstrated complete obliteration of 73% for fistulae involving the cavernous sinus, compared with 58% for lesions located at other sites. A higher obliteration rate was also noted for DAVF without CVD (75% versus 56%). Patients who experience incomplete fistula closure after GKS will often still report palliation of symptoms, such as pulsatile tinnitus.\(^{85–89}\) Reported complication rates after GKS of DAVF have been low or even zero in some series, with a few isolated reports of induction of CVD after treatment.\(^{85–89}\) However, the major drawback of GKS is the delay in fistula closure when this treatment modality is used alone. Consequently, GKS by itself may be inappropriate for DAVF that demonstrate aggressive behavior or high-risk angioarchitecture features.

**Conclusions**

DAVF are a rare type of acquired intracranial vascular malformation whose natural history is primarily determined by the route of venous drainage. Benign fistulae may be conservatively managed if associated symptoms are tolerable, whereas aggressive lesions with CVD carry a significant risk of neurological deterioration and should be promptly treated. Although no treatment may be indicated in benign fistulae, close follow-up is warranted as they may progress. Endovascular embolization is the primarily treatment modality for DAVF although microsurgical repair and stereotactic radiation therapy may also be efficacious in selected cases.

**Disclosures**

None.

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**KEY WORDS:** aneurysm ■ cerebellar diseases ■ magnetic resonance imaging ■ meninges ■ tomography
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