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1. Introduction

1.1. Classification systems and prognosis of DAVM

DAVM are typically classified by location of the involved sinus or shunt as well as by the pattern of venous drainage. The pattern of venous drainage is a key factor predicting the natural history of these lesions and provides the foundation for the widely adopted Borden and Cognard classification systems. Borden type I DAVM are mostly benign lesions that exhibit normal antegrade flow into a dural sinus. Type II lesions exhibit a degree of AV shunting exceeding the capacity of antegrade outflow from the involved sinus resulting in retrograde venous drainage into cortical veins. Type III lesions drain exclusively into cortical veins or a trapped sinus segment (sinus thrombosis at both ends from high-flow venous congestion). The Cognard classification is a modified version of the Djindjian classification and identifies five types of DAVM based on the pattern of venous outflow. Briefly, type I lesions exhibit normal antegrade flow into a dural sinus. Type II DAVM show retrograde venous drainage into the adjacent sinus segments (type IIa), cortical veins (type IIb) or both (type IIa +b). Types III and IV DAVM drain directly into cortical veins, either with (type IV) or without (type III) venous ectasia. Type V DAVM are typically localized to the tentorium or dural coverings of the posterior fossa and are further characterized by drainage inferiorly into spinal perimedullary veins.

Both classification systems have been validated and appear to correlate well with the risk of intracranial hemorrhage or non-hemorrhagic neurologic deficit (NHND). [9] Borden types II and III DAVM are associated with a more aggressive natural history and when technically
feasible are generally treated because of their increased risk of symptomatic presentation. In a study that included 20 patients with Borden type II and III lesions, Duffau et al [10] found that rebleeding within 2 weeks after the first hemorrhage occurred in as many as 35% of patients and noted that rehemorrhage carried a worse prognosis than the initial hemorrhage. The authors recommended complete and early treatment of ruptured DAVM with cortical venous drainage (CVD). On the other hand, Borden type I DAVM generally have a benign natural history. In a study where 68 patients with Borden Type I DAVM were conservatively treated and followed for a mean of 27.9 months, Satomi and colleagues [11] found that only 1 patient suffered an intracranial hemorrhage and noted a benign and tolerable level of disease in 98.5% of cases. However, according to a recent report, the risk of conversion of a Type I lesion to a more aggressive lesion with CVD appears to be higher than previously reported (annual rate of 1%). [12] Any change in patient symptoms should therefore prompt repeat angiographic imaging to rule out the development of alternative drainage routes.

The annual risk of hemorrhage of DAVM varies between 1.8% and 15%. [13-14] The first hemorrhagic episode of a DAVM is associated with >30% mortality or serious disability.

The annual mortality rate for DAVM with CVD may be as high as 10.4% with a combined annual risk of intracranial hemorrhage and NHND of 15%. [15] Recent data suggest that the natural history of DAVM dependents not only on the pattern of venous drainage, but also on the mode of presentation. As such, Soderman et al [16] found that the annual hemorrhage risk of Borden Type II or III DAVM was 7.4% for patients presenting with intracranial hemorrhage compared with only 1.5% for those with non-hemorrhagic presentation. Likewise, in a study by Strom et al [17] that included 28 patients with Borden Type II or III DAVM, the risks of hemorrhage and NHND were 7.6% and 11.4% respectively for lesions with symptomatic CVD versus only 1.4% and 0% for those with asymptomatic CVD. Based upon these data, Zipfel et al [18] proposed the inclusion of the mode of presentation (symptomatic or asymptomatic CVD) into the Borden and Cognard classification systems to allow for more accurate risk stratification in patients with high-grade DAVM.

<table>
<thead>
<tr>
<th>Type</th>
<th>Borden</th>
<th>Cognard</th>
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<tr>
<td>I</td>
<td>Drains directly into venous sinus or meningeal vein</td>
<td>Normal antegrade flow into dural sinus</td>
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| II   | Drains into dural sinus or meningeal veins with retrograde drainage into cortical veins | a) Retrograde flow into sinus(es)  
     |       | b) Retrograde filling of cortical vein(s)  
     |       | a+b) Retrograde drainage into sinus(es) and cortical vein(s) |
| III  | Drains into cortical veins without dural sinus or meningeal involvement | Direct drainage into cortical veins without venous ectasia |
| IV   | Direct drainage into cortical veins with venous ectasia  
     | >=5 mm and 3x larger than diameter of draining vein. |
| V    | Drainage to spinal perimedullary veins |

Table 1. The Borden and Cognard classification systems of DAVM
More recently, Geibprasert et al [19] have proposed an ambitious classification, conceptually unifying pathophysiologic consequences of cranial and spinal DAVM on an embryologic basis. The scheme is based on the venous afferent patterns of three epidural spaces: 1) the ventral drainage group derived from the notochord and corresponding sclerotome extending from the base of the sphenoid to the sacrum, 2) the dorsal epidural space derived primarily from the dorsally located intracranial dural sinuses as this space is not well-developed in the spine, and 3) the lateral epidural shunts located where lateral pial emissary bridging veins pierce the dura. By examining 300 patients with DAVM and categorizing their lesions by their respective afferent venous patterns, Geibprasert et al were able to establish some clinical generalizations about each group. Ventral epidural shunts demonstrated a 2.3:1 female predominance and were less likely associated with cortical venous reflux unless there was extensive thrombosis of the epidural drainage or an especially high-flow shunt. Similarly, dorsal epidural shunts were less likely to reflux into the cortical veins unless thrombosis was present. These lesions did not demonstrate sex predominance but did tend to occur in a lower age group (pediatric) and more frequently occur as multiple lesions. The lateral epidural shunts tended to present in older patients and were more common in men. These lateral lesions were always clinically aggressive and demonstrated significant cortical venous reflux.

2. Clinical features of DAVM and anatomic considerations for embolization

The clinical features associated with DAVM generally depend on the location of the lesion, the extent of the AV shunting, and associated abnormalities of venous drainage. [20-21] Symptoms may be indistinguishable from those associated with pial brain arteriovenous malformations and may include headache, diplopia, blurred vision, or neurologic dysfunction. Focal neurologic deficits and seizures may develop in relation to disturbances in regional cortical venous drainage resulting from the redirection of venous flow from the shunt into pial veins, potentially congesting venous territory remote from the site of the dural fistula. In patients with severe compromise of the deep venous drainage of the brain or with diffuse intracranial hypertension resulting from the obstruction of both sigmoid sinuses, the clinical presentation may include dementia. [5] Dementia may also develop in patients with superior sagittal sinus DAVM due to venous congestion of bilateral frontal lobes. Closure of the arteriovenous shunts may successfully reverse this state only when there are adequate residual venous channels available for the normal venous drainage of the brain. Rarely, cranial neuropathy or unilateral visual phenomena may arise secondary to arterial steal without evidence of associated venous hypertension. [22] Focal symptomatology may worsen or change as a result of the redirection of venous outflow from a DAVM. For example, progressive thrombosis and occlusion of the inferior and superior petrosal sinuses may be associated with worsening of signs in a patient with a cavernous sinus DAVM draining anteriorly through the ipsilateral ophthalmic veins. If contralateral drainage is available, the venous sinus hypertension may be transmitted to the contralateral cavernous sinus, leading to development of bilateral orbital symptoms.
The signs and symptoms of increased intracranial pressure occasionally complicate cases of DAVM. In certain cases this can be attributed to diminished CSF absorption through the arachnoid villi resulting from the transmission of increased venous pressure throughout the superior sagittal sinus. Patients may present with typical symptoms of normal pressure hydrocephalus, such as progressive dementia, gait disturbance, and urinary incontinence. [23] Alternatively, obstruction of the cerebral aqueduct secondary to compression of the mesencephalon by an ectatic draining vein may occur, leading to obstructive hydrocephalus. [24] Moreover, aneurysmal venous ectasia may unusually cause symptomatic mechanical compression of adjacent neurologic structures, most commonly in DAVM draining into pial veins of the posterior fossa. This is particularly true for type IV DAVM, which not infrequently present with clinical symptoms related to mass effect caused by pronounced venous ectasia. [5]

Approximately 20 to 33 percent of patients with symptomatic DAVM present with an intracranial hemorrhage. This most frequently is encountered in lesions involving the floor of the anterior cranial fossa or the tentorium cerebelli; however, it may occur in any case associated with cortical venous drainage, particularly in the presence of significant cerebral venous ectasia.² In a recent large study by Bulters et al, [25] DAVM associated with venous ectasia had a 7-fold increase in the incidence of hemorrhage (3.5% no ectasia vs 27% with ectasia). Therefore, patients with venous ectasia may represent a high-risk group that requires rapid intervention.

For those DAVM that present in ways other than hemorrhage, the clinical presentation depends entirely on the grade, location and venous afferent pattern of the fistula. This allows DAVM to be categorized clinicoanatomically into those involving the cavernous sinus, transverse and sigmoid sinuses, superior sagittal sinus, petrosal sinus, torcular, tentorial incisura, and anterior cranial base.

Approximately one-third to one-half of symptomatic intracranial DAVM involve the transverse and sigmoid sinuses. [26] Nearly half present with a subjective bruit as the first clinical manifestation due to proximity of the draining sinus to the middle ear. The tinnitus is synchronized to arterial pulsations and results from increased blood flow into the sigmoid or transverse sinuses. Auscultation over the retroauricular area usually reveals the pulsatile bruit. As with the other DAVM, additional neurologic symptoms and findings generally depend on the pattern of venous drainage encountered in the individual patient. Symptoms may include chronic signs of increased intracranial pressure potentially leading to papilledema and optic atrophy in addition to disturbances related to balance and hearing. Aggressive neurologic symptoms may occur in up to 27% of patients with transverse and sigmoid DAVM. In progressive cases, associated with obstruction of the ipsilateral jugular outflow redirected venous drainage into pial veins of the posterior fossa may result in brain stem or cerebellar dysfunction as well as posterior fossa hemorrhage. Rerouting of drainage into the supratentorial cortical venous compartment may be associated with the development of focal neurologic deficit or seizures as well as increased risk of intracranial hemorrhage. Spontaneous occlusion of transverse and sigmoid DAVM is rare (5%) and generally occurs after hemorrhagic events.
DAVM involving the superior sagittal sinus, tentorial incisura, petrosal sinuses, and anterior cranial base occur less frequently than DAVM involving the transverse, sigmoid, or cavernous sinuses. [5, 27] In these lesions, symptoms typically depend on the route of abnormal venous drainage and associated pattern of venous hypertension, and may include dysphasia, hemiparesis, hemisensory deficits, and abnormal visual phenomena. Several specific features deserve particular attention. (1) Dural fistulas involving the floor of the anterior cranial fossa are usually associated with drainage into ectatic parasagittal cortical veins and often present with intracranial hemorrhage. [27] Moreover, these patients may exhibit unilateral visual loss secondary to arterial steal from the ophthalmic circulation into ethmoidal and recurrent meningeal supplies to the shunt. Although a majority of these lesions are treated with open surgery, embolization through the ophthalmic artery can be undertaken with a reasonably high success rate and low complication risks. [28] (2) DAVM of the petrosal sinuses or tentorial incisura may occasionally drain inferiorly into perimedullary veins of the spinal cord (type V), resulting in progressive myelopathy similar to that encountered in spinal dural AVMs. [26] Assuming the venous sinus drainage of the brain is otherwise unimpaired, these symptoms usually respond well to endovascular or surgical closure of the shunt. Tentorial DAVM drain only via the leptomeningeal-cortical venous system. Consequently, they behave aggressively with severe hemorrhagic and nonhemorrhagic symptoms occurring in 19% and 10% of cases per year. Also, it is not uncommon for such lesions to cause fatal bleeding in the posterior fossa. Therefore, they should be treated aggressively by endovascular and/or surgical means to disconnect the venous drainage system and minimize the risk of hemorrhage and NHND. Superior sagittal sinus DAVM are frequently associated with restrictive change of the superior sagittal sinus and retrograde CVD. Thus, aggressive neurologic symptoms are common and occur in nearly one-half of cases. [29]

DAVM most frequently involve the transverse sinuses. Arterial supply to fistulae of this region predictably derive from identifiable supratentorial and infratentorial sources. The supratentorial group is usually organized around 1) contributors to the basal tentorial arcade, typically including the petrosal and petrosquamosal divisions of the MMA and the lateral division of the meningohypophyseal trunk off the ICA, and, occasionally 2) transosseous branches of the posterior auricular artery. The infratentorial group commonly involves the jugular division of the ascending pharyngeal artery, transmastoid and more distal transosseous branches of the occipital artery, and the posterior meningeal arteries and artery of the falx cerebelli, either of which can variably arise from the occipital, vertebral or ascending pharyngeal arteries as well as rarely directly from the PICA. With higher flow lesions indirect contribution from contralateral sources may be seen but this usually involves anastomosis with one of the above-mentioned conduits as a final common pathway.

In terms of embolization hazards, the petrosal branch of MMA notably gives rise to a branch which anastomoses with the stylomastoid branch of the occipital or posterior auricular arteries forming an arterial arcade within the facial canal which if aggressively embolized (inadvertently) may result in damage to the facial nerve. In that the petrosal branch of the MMA usually participates in the supply of transverse sinus DAVM through the basal tentorial arcade, its contribution to the lesion commonly can be indirectly devascularized by accessing the basal
tentorial arcade posterolaterally through the petrosquamosal division of the MMA avoiding the need for direct catheterization and embolization of the petrosal branch altogether. The basal tentorial arcade is an arterial network extending along the insertion of the tentorium into the petrous ridge from the petroclinoid ligament laterally to the transverse sinus. The jugular division of the APA enters the cranial vault via the jugular foramen supplying CN 9,10,11 before dividing into medial and lateral divisions. The medial division courses along the inferior petrosal sinus where it supplies CN 6 and anastomoses with the medial division of the lateral clival branch of MHT. The lateral division runs superiorly along the sigmoid sinus and vascularizes the dura along the transverse sigmoidal confluence. In very high flow fistulae of the distal transverse sinus or lesions of the sigmoid sinus and foramen magnum, recruitment of supply through the hypoglossal division of the ascending pharyngeal artery may be seen (particularly where this artery gives rise to the ipsilateral posterior meningeal artery. Transarterial embolizations through this division of the APA (particularly with liquid embolic agents) may result in injury to CN 12 leading to ipsilateral paresis of the tongue.

DAVM of the cavernous sinus (CSDAVM) are most commonly seen in female patients and generally associated with orbital signs and symptoms that fluctuate depending on alterations in orbital venous outflow, which develop secondary to thrombosis and changes in head position. Patients typically present with the gradual onset of focal or diffuse chronic eye redness distinguishable from uveitis. Close inspection reveals dilated tortuous conjunctival and epibulbar vessels that exhibit an acute angulation near the ocular limbus. [30-31] These lesions are often associated with an elevation of episcleral venous pressure leading to a persistent rise in intraocular pressure in the affected eye, potentially resulting in the development of glaucoma. If both cavernous sinuses become involved in the venous drainage secondary to a change in the ipsilateral venous outflow of the affected cavernous sinus, the ocular findings may become bilateral. The patient may complain of pulsatile tinnitus, and in 25 percent of cases a bruit can be auscultated over the orbit. [5] Cranial neuropathies, most commonly involving the sixth nerve, frequently lead to ocular motor dysfunction, which also may be exacerbated by orbital venous congestion and proptosis. More important to the planning of embolization are the hypoxic ischemic retinal changes that develop in approximately 15 percent of patients. [21] Rarely, if thrombosis in the cavernous sinus is extensive, abnormal drainage into cerebral veins may occur, increasing the likelihood of an intracranial hemorrhage or venous infarction. [5] Unfortunately, frequently cited classification schemes of intracranial DAVM are deficient in their handling of CSDAVM due to the lack of explicit consideration given to ophthalmic venous drainage and the clinical consequences of orbital venous congestion. Despite the lack of a coherent classification scheme for CSDAVM, the implications of venous outflow from these lesions are similar to DAVM at other locations and the analysis of venous drainage is important in understanding the pathophysiology of the disease at this site. An excellent study of the clinical manifestations in 85 patients with CSDAVM relative to their angiographic characteristics was reported by Stiebel-Kalish et al. [32-33] In this study, the clinical symptoms found in patients with CSDAVM were related to the abnormal venous drainage and could be predicted by analysis of the aberrant venous drainage patterns. Interestingly, central nervous system symptoms or dysfunction, were found in 7 (8%) of these patients, attesting to the potential danger of cortical venous drainage even
among patients with CSDAVM. Spontaneous regression of CSDAVM is a well-described phenomenon that is observed in 10%–50% of cases. [29]

The vascular supply to the dura of the cavernous sinus is complex because of extensive regional anastomoses between dural branches of the internal carotid and branches of the internal maxillary artery (middle meningeal, and accessory meningeal arteries, and the artery of the foramen rotundum). Moreover, the ophthalmic artery may participate indirectly via a tentorial branch of the recurrent meningeal artery. From the perspective of angiographic workup and embolization, these lesions may be divided conceptually into two groups: (1) an anterolateral group, arising from the orbital apex and lateral cavernous sinus, and (2) a posterior group, including the posterior cavernous sinus, petroclinoid ligament, and dorsum sella.

The meningeal supply to anterior division lesions may be considered to reflect the hemodynamic balance between branches arising from the horizontal segment of the cavernous internal carotid artery, most notably the inferolateral trunk (ILT) and meningeal branches of the internal maxillary artery. This latter group includes cavernous and recurrent meningeal branches of the MMA, cavernous meningeal branches of the accessory meningeal artery, and the artery of the foramen rotundum. As expected, embolization of these meningeal arteries should be preceded by superselective angiographic analysis to prevent inadvertent embolization into the internal carotid artery or possible damage to the orbit or regional cranial nerves.

The supply to posterior division lesions is derived primarily from medial and lateral clival (meningo-hypophyseal) branches of the internal carotid artery and their potential anastomotic connections with branches of the ascending pharyngeal and middle meningeal arteries. These most notably include the ascending clival and inferior petrosal arcades, derived from the hypoglossal and jugular divisions of the ascending pharyngeal artery, respectively; the posterior cavernous branches of the MMA; and the basal tentorial arcade supplied by the petrosal and the petrosquamosal branches of the MMA.

Three critical points should be considered before embolization of fistulae involving this territory. (1) The vascular supply to the intrapetrous facial nerve should be determined. This may arise primarily from the petrous branch of the MMA. For this reason, petroclinoid lesions supplied by the basal tentorial arcade should be embozilated preferentially from the petrosquamosal branch of the MMA, thereby avoiding the proximal petrosal artery. (2) Potential contributions from the contralateral internal carotid and ascending pharyngeal arteries via transclival anastomoses should be evaluated, particularly in lesions involving the dorsum sellae. (3) Because embolization of upper clival and petroclinoid lesions may involve the hypoglossal or jugular division of the ascending pharyngeal artery, attention must be directed to the possibility of iatrogenic lower cranial neuropathy when using NBCA, Onyx, or ethanol. Midline lesions requiring aggressive embolization of pedicles from both ascending pharyngeal arteries should be performed as a staged procedure on different days, specifically to avoid development of bilateral hypoglossal nerve deficits.

The simplest and most commonly utilized route to access the cavernous sinus is through the inferior petrosal sinus. Guidewire or microcatheter navigation through the sinus may be complicated by vessel rupture. Alternatively, access to the cavernous sinus may be obtained
through the facial vein or the superficial temporal vein. Direct operative cannulation of the superior ophthalmic vein is also an acceptable route to the cavernous sinus when other approaches have been exhausted. [34]

3. Imaging of DAVM

Recent advances in both computed tomography (CT) and magnetic resonance imaging (MRI) have significantly contributed to the initial diagnostic evaluation of patients with suspected DAVM. Because CT and MRI findings are nonspecific, however, the diagnosis can be delayed or missed. Routine conventional head CT is the first-line investigation of patients presenting with tinnitus, headache or other neurological symptoms. Its value is limited to identifying intracranial hemorrhage and edema due to venous congestion (area of low density). Focal or generalized atrophy of the brain, possibly accompanied by hydrocephalus, are nonspecific secondary findings that may be appreciated. Although not infrequently diagnostically equivocal, MRI is more helpful than CT because it can reveal dilated vessels, thrombosed venous structures, and prominent vascular enhancement particularly in patients with DAVM associated with CVD. The combination of prominent flow voids on the cortical surface and high-intensity lesions in the deep white matter on T2-weighted images secondary to venous hypertension/congestion is highly suggestive of a DAVM. Despite the presence of these secondary signs that suggest the presence of a DAVM, conventional MRI alone is generally unsuccessful in defining the exact site of fistulization. Any suspicious findings on CT/MRI should prompt catheter angiographic evaluation.

The advent of CT angiography (CTA) and magnetic resonance angiography (MRA) has provided more power to the noninvasive screening of patients with suspected DAVM. In addition to providing anatomic details, these modalities may be coupled with perfusion studies to evaluate the effect of a DAVM on regional blood flow.

CTA aids in the accurate diagnosis and characterization of DAVM by localizing the fistula and demonstrating the pattern of venous drainage and supplying arteries. Overlapping bone structures may make it difficult to demonstrate the detailed vascular pattern of DAVM especially for smaller lesions. The sensitivity of CTA for diagnosis of DAVM is reportedly lower than the sensitivity of MRA (15.4% versus 50%). [35] Lee et al [36] recently introduced a CTA algorithm for bone removal (hybrid CTA) that eliminates bone structures while preserving enhancing transosseous vascular structures. They found that the technique provides valuable information for treatment planning and carries a sensitivity of 93% and a specificity of 98%. In addition, recent studies have shown that 4D CTA with high spatial and temporal resolution are suitable for the diagnosis, classification, treatment planning, and follow-up imaging of DAVM. [37-38]

MRA may be performed using a three-dimensional time-of-flight (3D TOF) technique or MR digital subtraction angiography (MR DSA). [39-42] The presence of multiple high-intensity curvilinear or nodular structures adjacent to a sinus, in conjunction with high-intensity foci within the sinus is considered suspicious for a DAVM; however, the technique still suffers
from a high false positive rate, with as many as 14% of otherwise healthy patients incorrectly identified as possibly harboring a DAVM by 3D TOF MRA. Although the current spatial resolution of MR DSA is less than 3D TOF MRA, the benefit of MR DSA would be related to the temporal resolution of the technique and the ability to depict flow within cortical veins, particularly important in those patients with retrograde flow from a DAVM.

Despite the advances in both CTA and MRA, conventional digital subtraction angiography remains paramount in the diagnosis and pretreatment evaluation of intracranial DAVM. The angiographic evaluation usually includes selective studies of the internal and external carotid arteries bilaterally as well as of both vertebral arteries when evaluating lesions of the posterior fossa or tentorium. The pretherapeutic examination must be tailored to the clinically suspected location of the fistula and must disclose the entire arterial supply, as well as any anastomoses between the supplying vessels and arterial distributions to the orbit, brain, or cranial nerves. This usually requires superselective arterial catheterization and angiography before the use of embolic materials. The venous anatomy must be studied with respect to the pattern of drainage from the fistula, and the adequacy of normal venous drainage of the brain must be assessed.

4. Therapeutic approaches to DAVM

Because many DAVM regress spontaneously or remain asymptomatic throughout the patient’s life, it is crucial to weigh the risks of treatment against the natural history of these lesions. Management should be tailored to the type of lesion (location, classification, and angiographic features) and individual patient history (age, clinical presentation, and comorbidities) and may include relief of symptoms or complete occlusion of the DAVM. Although spontaneous resolution of clinical signs related to DAVM is not uncommon, most symptomatic lesions require some form of treatment. Treatment options include observation, carotid-jugular compression, transarterial embolization, transvenous embolization, open surgery and stereotactic radiosurgery. In the majority of patients, a multimodality approach with a combination of treatment offers the best chance for success.

Patients with Type I DAVM are at low risk of hemorrhage and should be managed conservatively unless they have disabling clinical symptoms like tinnitus or develop new neurological deficits or CVD at follow-up. Expectant follow-up of asymptomatic lesions should include serial MRI to detect changes in the DAVM anatomy. Angiographic follow-up should also be considered every few years especially for DAVM of the anterior cranial fossa or the tentorial incisura, which commonly develop CVD. Patients with symptomatic type II or III DAVM should be treated aggressively to minimize the risk of hemorrhage and NHND. The management of asymptomatic type II and III lesions should take into consideration the patient’s age, treatment decision and risk of future hemorrhage. Intervention is often favored over observation because of the long-term risk to the patient and the dismal natural history of an intracranial hemorrhage.
Carotid-Jugular Compression. Patients with Borden type I transverse or sigmoid sinus DAVM or with fistulas of the cavernous sinus and otherwise normal ophthalmologic examinations may be treated conservatively with ipsilateral carotid or occipital artery compression. Intermittent manual compression of the carotid artery may be effective in eliminating DAVM involving the ipsilateral cavernous sinus in patients with mild findings and no evidence of carotid vascular disease or other contraindications to carotid compression. The ipsilateral carotid artery is compressed, using the contralateral hand, for approximately 5 minutes every waking hour for 1 to 3 days. If this is tolerated, the compression time is increased to 10 to 15 minutes of compression per waking hour. The compression, if properly performed, produces concomitant partial obstruction of the ipsilateral carotid artery and jugular vein. This results in the transient reduction of arteriovenous shunting by decreasing arterial inflow while simultaneously increasing the outlet venous pressure, thereby promoting spontaneous thrombosis within the nidus. Nearly 30% of cavernous sinus and 25% of transverse/sigmoid sinus DAVM will thrombose with compression therapy. Compression therapy is usually not recommended in patients with CVD.

Embolization. The development of improved superselective angiographic catheter systems and embolic agents has increased the role of interventional neuroradiology in the management of these lesions, both primarily and preoperatively. Two strategies, transvenous and transarterial have been employed and their appropriate selection depends on the location and complexity of the DAVM, its vascular features and the potential complications inherent to each technique. Treatment aims to completely occlude the arteriovenous fistula. If this is not an option, selective disconnection of the CVD is an acceptable option that is equally effective in reducing the risk of hemorrhage and NHND associated with DAVM. Transvenous embolization with metallic coils or detachable balloons has been advocated primarily for the treatment of DAVM involving the transverse, sigmoid or cavernous sinuses. The technique involves a transfemoral or intraoperative approach to the affected venous sinus following which coils, balloons or liquid embolic agents are deposited adjacent to the shunt. Several features are critical in appropriate patient selection for this method of treatment. (1) The segment of sinus to be occluded must be in proximity to the fistula and receive its entire venous drainage. (2) The sinus to be occluded should not be essential to the normal venous drainage of the brain. The cerebral venous drainage must be thoroughly evaluated before embolization to determine the alternate pathways for cerebral venous outflow and avoid potential venous infarction or hemorrhage. (3) The target sinus must be completely occluded throughout the involved segment to avoid diversion of fistulous flow into confluent cerebral veins and worsening of CVD. Such redirection of a high flow shunt into previously uninvolved low capacitance venous channels may precipitate an acute venous infarct or hemorrhage.

Levrier et al developed a novel way to treat DAVM that would preserve the venous sinus. In ten patients including fistulas grade I-IV both with and without sinus stenosis, the researchers used a transvenous approach to angioplasty the involved sinus and then placed stents with high radial force to bridge the ostia of cortical veins draining into the sinus. Their follow-up at 7 months by conventional angiography revealed that four patients had complete DAVM occlusion and four had significantly reduced flow through the fistula. Two subjects
refused repeat angiography. At two years, CTA confirmed stent patency in eight out of nine patients imaged. The safety and long-term efficacy of this technique, however, require further investigation.

Transvenous embolization is particularly useful for DAVM with multiple arterial feeders. Typically, involved arteries are small and torturous arterial feeders, which renders selective catheterization extremely challenging or hazardous. Ease of access to the fistulous site and the ability to obliterate the fistula in a single session are important advantages of this approach. Transvenous embolization is associated with a low complication rate and high rates of cure and complete occlusion of the fistula. [47-48] Transvenous embolization, however, is less suited for DAVM involving the superior sagittal sinus. It can also be associated with severe complications, including vessel rupture, sinus venous thrombosis, venous infarction, hemorrhage, and neurological deficits related to disruption of venous drainage. [49] Hemorrhage may be related to vessel injury or to the sacrifice of a dural sinus draining normal brain parenchyma. Additionally, transvenous embolization is rarely associated with the development of de novo DAVM at secondary intracranial sites following occlusion of the primary lesion. While the etiology of these secondary de novo fistulas is unclear, they may arise from angiogenesis induced by venous hypertension secondary to the occlusion of the major dural sinuses targeted by transvenous embolization.

Under some circumstances, transarterial embolization with liquid embolic agents offers advantages over a transvenous approach. Not infrequently, transvenous access to the DAVM is limited by venous sinus occlusion or high-grade stenoses preventing transvenous catheterization. Likewise, high-grade lesions emptying directly into remote small cerebral veins may be inaccessible to uncomplicated venous catheterization. As such, tentorial incisura and anterior cranial fossa DAVM, which frequently behave aggressively, may not be accessed transvenously. Transarterial delivery of a liquid embolic agent capable of permeating the vascular apparatus of the shunt provides the means for discrete definitive occlusion of the fistula site and reduces the likelihood for diversion of shunt flow into more dangerous alternate venous pathways while enabling closure of the fistula without necessarily sacrificing an entire venous conduit that may be critical to the drainage of normal brain parenchyma. Conversely, incomplete occlusion of the fistula by transarterial embolization is usually complicated by recruitment of new collateral vessels that are smaller, more tortuous and less amenable to embolization. Complex fistulas may require a multistaged approach combining transarterial and transvenous techniques to eliminate CVD and occlude the fistula.

Transarterial embolization may be effective in palliating disabling symptoms through occlusion of arterial feeders even without angiographic cure of the DAVM. Transarterial embolization also plays an important role in decreasing flow through DAVM before surgical intervention, transvenous obliteration, and radiosurgery. [50-51]

The transarterial approach requires selective catheterization of individual feeding vessels followed by superselective angiography to evaluate the vascular supply to the fistula, particularly with respect to potential anastomoses with the orbit or cerebral vasculature. It is important to understand that such anastomoses may not be demonstrable on the initial
angiograms but may become manifest as progressive embolization produces alterations in flow within the target vascular territory.

Guidewire-directed microcatheters are typically employed in the catheterization of meningeal branches supplying such lesions. The embolic agents commonly used in transarterial embolization of DAVM are liquid cyanoacrylate (NBCA), Onyx, polyvinyl alcohol foam (PVA), or ethanol. Ideally, liquid embolic agents, delivered close to the shunt under wedged-microcatheter induced flow arrest, present the best opportunity for embolotherapeutic cure of the lesion as it enables permeation of the collateral complex supplying the fistula and its immediate venous receptacle thus permanently occluding the shunt. Such a degree of permeation is not possible using particulate agents that characteristically lodge within arterioles of the peri-fistula microcollateral network at a point proximal to the shunt. If not fully permeated, these microcollateral networks will then evolve and reestablish flow through the shunt complex.

Also, the particles degrade within days to weeks, resulting in high recurrence rates of the fistula and possibly in extensive shunting into leptomeningeal veins. Transarterial embolization with PVA is therefore used to relieve symptoms or in combination with other procedures such as radiation, surgery, or transvenous embolization. As with PVA, embolization with coils alone does not provide complete obliteration of DAVM. [52]

Nevertheless, PVA may find use in several situations. First, the initial use of PVA in embolizing the less favorable arterial supplies to a multi-pedicle fistula, may facilitate more complete subsequent embolization of the shunt with liquid embolic agents through a safer conduit. The embolization of competing inflows to the shunt with PVA allows the undiluted permeation of the fistula by the liquid embolic agent without fragmentation of the glue column. PVA may also be useful in reducing flow through low-velocity shunts, thereby facilitating thrombosis in these DAVM. This can be particularly applicable in managing low flow CSDAVM, and may be combined with manual compression in treating lesions also supplied by cavernous segment dural branches of the ipsilateral ICA.

In certain situations, partial embolization of dural fistulas may be performed in an attempt to alleviate disabling symptoms. For example, partial embolization of a cavernous sinus DAVM can reduce intraocular pressure in a patient suffering acute deterioration of visual acuity. Aggressive treatment in such cases may not be needed unless symptoms are particularly disabling or the DAVM is associated with CVD. Partial embolization may also be advocated in patients presenting with new-onset dementia or in those patients with severe tinnitus. Lastly, PVA and liquid embolic agents are used in the preoperative devascularization of dural fistulas prior to surgical excision. In this situation, particulate embolic agents, because of their low morbidity, are generally preferred and should be applied 1 to 2 days before surgery.

NBCA has been widely utilized for transarterial embolization of DAVM with fairly good results. Guedin et al [53] treated 43 patients with Borden Type II or III DAVM using NBCA and reported complete obliteration of the fistula in 34 patients (79%) and occlusion of CVD in all remaining cases. There was no treatment-related mortality or permanent morbidity in the series. Interestingly, they reported post-embolization secondary thrombosis in 5 patients in whom residual flow was noted on the immediate post-treatment angiograms. In a recent large study by Kim et al [54] that included 121 DAVM treated with transarterial glue embolization, immediate cure was achieved in 14.0% of lesions, and progressive complete thrombosis of the residual shunt at follow-
up in 15.7% of lesions. Surgical CVD disconnection or transvenous coil embolization was necessary for clinically important residual shunts in as many as 45.2% of all cases. Procedural complications were seen in 7.8% of patients in the series.

However, use of NBCA has some disadvantages. It is an adhesive agent that undergoes rapid polymerization at contact with blood, which may increase the risk of microcatheter retention or avulsion of the feeding artery upon removal of the microcatheter. The injection must be performed quickly and continuously, which may diminish the precision of injection and result in suboptimal penetration into the fistulous site. Use of a wedged microcatheter technique with low-concentration glue may maximize glue penetration into the venous drainage route (Figure 1).

![Figure 1](http://dx.doi.org/10.5772/55219

Recently, the introduction of Onyx has added an important element to the endovascular armamentarium and improved the endovascular treatment of DAVM. Onyx is comprised of
ethylene vinyl alcohol copolymer dissolved in DMSO (dimethyl sulfoxide), and suspended micronized tantalum powder to provide contrast for visualization under fluoroscopy. Onyx offers several advantages over NBCA, which allow for safer and more efficient treatment of DAVM. Due to its lava-like flow pattern and its nonadhesive nature, Onyx facilitates longer, slower, and more controlled injections with better penetration of the fistula. It also allows embolization of a substantial portion of the lesion from a single pedicle injection because the agent can efficiently penetrate the depths of the fistulous connection and then flow into adjacent arterial feeders, thereby obviating the need for multiple embolizations. The interventionalist can even discontinue Onyx injection for angiographic assessment of the embolization and evaluation of collateral and en passage feeders that may become evident during the course of embolization. Additionally, Onyx is less adherent to the microcatheter than NBCA with possibly a lower risk of catheter retention and arterial rupture. The middle meningeal artery provides an excellent route for Onyx injection with particularly high curative rates according to several reports. [55-57] The middle meningeal artery is easy to catheterize and its branches are anchored to the dura and calvarium, which facilitates removal of the microcatheter and minimizes the risk of arterial avulsion.

Onyx has some disadvantages compared to NBCA, namely an increase in fluoroscopy time, procedure time, and procedure cost. Cranial nerve injury and DMSO-induced angioxicity are additional disadvantages of Onyx. There is also a risk of distal embolization of the embolic material into the venous system and the pulmonary circulation.

Several investigators have reported remarkably high cure rates with this embolic agent, with a high proportion of treatments completed in a single session. [49] Cognard et al. enrolled 30 patients in a prospective trial: ten were graded type II, eight type III and twelve type IV fistulas. [55] They reported complete anatomic cure in 24/30 patients with only two complications, including a temporary cranial nerve palsy and post-procedure hemorrhage secondary to venous outlet thrombosis. Lv et al. report their experience with 40 patients suffering from DAVM. [27, 58] They report a complete occlusion rate of 25/40 or 62.3%. Nine patients suffered complications including reflexive bradyarrhythmia in 3 patients, hemifacial hypoesthesia in 3, hemifacial palsy in 2, posterior infarction in 2, jaw pain in 1, hallucinations in 1, Onyx migration in 1 and retention of a microcatheter tip in 1. Abud et al. [59] treated 44 DAVM with Onyx and achieved occlusion of the shunt in all but 9 patients, 5 of whom were successfully treated by complimentary transvenous embolization with coils and Onyx. In as many as 81% of cases, a cure was obtained in a single session. Six complications were observed including 4 cranial nerve injuries and 2 cases of venous thrombosis post-embolization. In a series of 29 DAVM treated with Onyx embolization, mostly through a transarterial approach, Stiefel et al. [56] achieved an angiographic cure in 72% of all lesions, with complications occurring in 9.7% of cases and leading into permanent morbidity in only 2.4%. We have recently reviewed our experience in 39 patients with DAVM treated between 2001 and 2009 at Jefferson Hospital for Neuroscience. We found no major procedure-related complications in the series and achieved an obliteration rate of 75% with elimination of CVD in up to 85% of patients with Onyx embolization (Figure 2-3).
A few investigators have used Onyx in a transvenous approach to carotid-cavernous fistulas. After an unsuccessful embolization of a C-C fistula using detachable coils and liquid adhesion agents, Arat et al successfully completed the embolization by injecting Onyx into the cavernous sinus forming a cast of the structure. [60-61] Similarly, He et al report their experience in 6 patients using a combination of detachable coils and Onyx via a transvenous approach. [62] Four of the six cases were completely embolized in one attempt, whereas the other two required staged procedures. In these latter two cases the patient suffered minor transient cranial nerve palsies. Suzuki et al report equally good results in three patients with spontaneous C-C fistulas. [63] In all these studies patients experienced rapid relief of their neuro-ophthalmologic symptoms. El Hammady et al [64] treated 12 patients with C-C fistulas using Onyx, 8 through a transvenous route and 4 through a transarterial route. All lesions in their series were obliterated in a single session with resolution of presenting symptoms in 100% of patients by 2 months. Cranial neuropathies, however, were noted in 3 patients likely from post-embolization cavernous sinus thrombosis and swelling or from cranial nerve ischemia/infarction from deep penetration of Onyx. We have recently reported on Onyx embolization of C-C fistulas through a surgical cannulation of the superior ophthalmic vein in a series of 10 patients. [34] We achieved complete obliteration of the fistula in 8 patients and a significant reduction in fistulous flow in 2 patients, with no procedural complications.
5. The role of surgery

With the advent of Onyx, most lesions can now be successfully managed with endovascular therapy. However, surgical treatment of DAVM may still be necessary when endovascular options have failed. Surgery consists of disconnection of draining veins, disconnection of arterial feeders, resection/packing of the dural sinus, and/or direct puncture and embolization of large varices or meningeal arteries. Hoh et al [65] described a technique whereby the draining vein is clipped close to the fistula with extensive dural coagulation.

DAVM of the anterior cranial fossa and the superior sagittal sinus are more suitable for surgical treatment than other types of DAVM. Transarterial embolization plays an important role in decreasing flow through the DAVM prior to surgical intervention and facilitates operative
exposure of the involved segment of the dural sinuses, thus affecting the quality and completeness of surgical excision.

6. The role of stereotactic radiosurgery

Stereotactic radiosurgery is an acceptable option for DAVM not amenable to surgical or endovascular therapies. It is best suited for benign lesions without CVD (type I) and for low-flow cavernous DAVM (which typically do not have CVD). Radiosurgery induces thrombogenic obliteration of DAVM with a latency period of up to two years. This treatment modality is therefore not suitable for DAVM with CVD because such lesions have a malignant natural history and require rapid and definitive treatment via surgical or endovascular means. A recent review of 14 studies found that stereotactic radiosurgery with or without adjunctive embolization results in DAVM obliteration in 71% of cases and post-treatment hemorrhage in 1.6% of cases (4.8% of lesions with CVD). [66] Despite these promising results, experience with radiosurgery in DAVM treatment remains limited and the efficacy of the technique requires more investigation in large prospective studies. Meanwhile, stereotactic radiosurgery should be reserved for lesions that are not amenable to surgical or endovascular interventions.

7. Conclusion

Recent advances in endovascular therapies and studies of the anatomical and functional properties of DAVM led to a rapid evolution in their diagnosis and management. Onyx embolization through transarterial or transvenous approaches has emerged as safe and highly efficient treatment for even the most complex lesions. However, the decision of which approach and embolic agent to use for treatment of a DAVM must be tailored to each individual case, recognizing that the most effective approach for permanent DAVM treatment, particularly in high-flow shunts, may require a combination of approaches and embolic agents. Treatment of DAVM should be entrusted to a multidisciplinary team with ample expertise in the management of these often challenging lesions.

Author details

Nohra Chalouhi, Pascal Jabbour, Aaron S. Dumont, L. Fernando Gonzalez, Robert Rosenwasser and Stavropoula Tjoumakaris*

*Address all correspondence to: stavropoula.tjoumakaris@jefferson.edu

Department of Neurosurgery, Thomas Jefferson University and Jefferson Hospital for Neuroscience, Philadelphia, Pennsylvania, USA
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