A 29 years old male patient presented with proptosis, ecchymoses of the left eye with both subjective and objective bruit in the left eye and left ear, and tinnitus in the left ear. There was no history of trauma, venous thrombosis, or surgical operations. The condition is dated since birth.

Figure 1. Postcontrast CT scan showing a left temporal dilated venous channel with some wall calcifications and moderate mass effect. The densely enhanced venous channels are mixed with parenchymal hypodensities. The dilated, densely enhanced venous channel reflects venous hypertension which commonly results in retrograde venous reflux, congestive encephalopathy and white matter changes (petechial hemorrhages, white matter edema, reactive astrogliosis). Dilated venous pouches with all calcification are evident in (A).
Figure 2. Postcontrast CT scan showing a left hemispherical markedly dilated, densely enhanced venous channels (venous ectasia). The disease is involving both then superficial (Cortical) and deep venous systems. Both systems are interconnected by transhemispherical dilated collaterals. The dilatation predominately affects the deep venous system. The white matter changes most probably reflects retrograde venous reflux and congestive encephalopathy. The dilated transhemispherical venous channels and congestive encephalopathy most probably denotes the existence of cortical venous reflux. Notice the marked dilations of the the vein of Galen.

Figure 3. Precontrast MRI T1 images showing markedly dilated signal void tortuous venous channels predominately involving the deep venous systems of the left hemisphere, with some dilated cortical veins, in particular the vein of Galen is markedly dilated. Notice the significant white matter changes in the form of a mixture of T1 hypointensities, hyperintensities and positive mass effect. The precontrast T1 hyperintensities most probably represents blood products secondary to venous congestion.
Figure 4. MRI T2 images showing markedly dilated signal void tortuous venous channels predominately involving the deep venous systems of the left hemisphere, with some dilated cortical veins, in particular the vein of Galen is markedly dilated. Notice the significant white matter changes in the form of a mixture of T2 hypointensities, hyperintensities and positive mass effect. The term venous congestive encephalopathy (VCE) was introduced in 1994. On CT, the venous congestion may be evident as an area of edema and mass effect. In patients with cortical venous reflux, MR imaging often shows prominent flow voids on the surface of the brain. Hydrocephalus may be secondary to the venous hypertension in the superior sagittal sinus. On MR imaging, T2 hyperintensity deep in the brain parenchyma may be evident secondary to the venous hypertension and passive congestion of the brain. The cerebellum, cerebrum, and deep gray nuclei or brainstem may be affected. In chronic cases, the proton density or T2-weighted images may show a central hypointensity that may be related to hemosiderin deposition from chronic venous congestion. In the cerebral hemispheres, the deep white matter is the most vulnerable to the venous congestion. The T2 hyperintensity may be reversible after treatment. The differential diagnosis of the T2 hyperintensity would include a superior sagittal sinus thrombosis with a venous infarction or venous congestion, demyelination, or a dysmyelination and neoplasm. But the combination of a surplus of pial vessels, T2 hyperintensity deep within the brain, and peripheral enhancement is highly suggestive of a dural arteriovenous fistula and mandates prompt angiography.

Figure 5. MRI T2 images showing markedly dilated signal void tortuous venous channels predominately involving the deep venous systems of the left hemisphere, with some dilated cortical veins, in particular the vein of Galen is markedly dilated. Notice the significant white matter changes in the form of a mixture of T2 hypointensities,
hyperintensities and positive mass effect. The term venous congestive encephalopathy (VCE) was introduced in 1994. On CT, the venous congestion may be evident as an area of edema and mass effect. In patients with cortical venous reflux, MR imaging often shows prominent flow voids on the surface of the brain. Hydrocephalus may be secondary to the venous hypertension in the superior sagittal sinus. On MR imaging, T2 hyperintensity deep in the brain parenchyma may be evident secondary to the venous hypertension and passive congestion of the brain. The cerebellum, cerebrum, and deep gray nuclei or brainstem may be affected. In chronic cases, the proton density or T2-weighted images may show a central hypointensity that may be related to hemosiderin deposition from chronic venous congestion. In the cerebral hemispheres, the deep white matter is the most vulnerable to the venous congestion. The T2 hyperintensity may be reversible after treatment. The differential diagnosis of the T2 hyperintensity would include a superior sagittal sinus thrombosis with a venous infarction or venous congestion, demyelination, or a dysmyelination and neoplasm. But the combination of a surplus of pial vessels, T2 hyperintensity deep within the brain, and peripheral enhancement is highly suggestive of a dural arteriovenous fistula and mandates prompt angiography.

Figure 6. MRI T2 images showing markedly dilated signal void tortuous venous channels predominately involving the deep venous systems of the left hemisphere, with some dilated cortical veins, in particular the vein of Galen is markedly dilated. Notice the significant white matter changes in the form of a mixture of T2 hypointensities, hyperintensities and positive mass effect. The term venous congestive encephalopathy (VCE) was introduced in 1994. On CT, the venous congestion may be evident as an area of edema and mass effect. In patients with cortical venous reflux, MR imaging often shows prominent flow voids on the surface of the brain. Hydrocephalus may be secondary to the venous hypertension in the superior sagittal sinus. On MR imaging, T2 hyperintensity deep in the brain parenchyma may be evident secondary to the venous hypertension and passive congestion of the brain. The cerebellum, cerebrum, and deep gray nuclei or brainstem may be affected. In chronic cases, the proton density or T2-weighted images may show a central hypointensity that may be related to hemosiderin deposition from chronic venous congestion. In the cerebral hemispheres, the deep white matter is the most vulnerable to the venous congestion. The T2 hyperintensity may be reversible after treatment. The differential diagnosis of the T2 hyperintensity would include a superior sagittal sinus thrombosis with a venous infarction or venous congestion, demyelination, or a dysmyelination and neoplasm. But the combination of a surplus of pial vessels, T2 hyperintensity deep within the brain, and peripheral enhancement is highly suggestive of a dural arteriovenous fistula and mandates prompt angiography.
The primary pathology in the above reported case is two dural arteriovenous fistulas at the cavernous sinus and the
Abnormal communications between the arterial and venous systems may be congenital or acquired. Most extracranial arteriovenous malformations (AVMs) are apparent clinically, although imaging studies are helpful in delineating the extent of the abnormality. Brain parenchymal arteriovenous malformations may be more obscure clinically, but are usually quite readily identified on contrast-enhanced CT and MR studies. Conventional angiography identifies arterial supply and venous drainage. CT angiography may also prove useful. Time-of-flight MR angiography complements spin-echo sequences.

A dural AVM or arteriovenous fistula (AVF) is a well-known cause of headache and hemorrhagic infarction. However, dural AVM or arteriovenous fistula is also the most frequent cause of objective pulsatile tinnitus in the patient with a normal otoscopic examination. The transverse, sigmoid, and cavernous sinuses are the most frequent locations of dural arteriovenous malformations and arteriovenous fistulas; transverse and sigmoid sinus involvement causes pulsatile tinnitus. Branches of the external carotid artery supply these dural arteriovenous malformations; venous drainage may be extracranial, intracranial, or both.

CT or MR studies may demonstrate a dilated dural venous sinus, unusually large or numerous cortical veins, or abnormal vessels in the soft tissues beneath the skull base. However, dural arteriovenous malformations or arteriovenous fistulas are often invisible on CT and MR studies. A normal contrast-enhanced CT or MR study therefore does not exclude a dural AVM or AVF. Conventional angiography may be the only modality that shows the abnormality. When a patient has convincing history and physical findings and normal cross-sectional imaging studies, conventional angiography is an important diagnostic option.

Brain arteriovenous shunts (AVSS) develop from a primary defect or malformation of the neurovascular system. There are two broad categories of Brain arteriovenous shunts: arteriovenous malformations (AVMS) and arteriovenous fistulas (AVFs). Brain arteriovenous shunts are characterized by the direct connection of one or more arteries to one or more draining veins, without intervening capillary beds. The shunting of arterial blood into the low-resistance venous network produces a high flow that typifies these lesions and results in distention, tortuosity, and reactive changes in the affected arteries and veins.

The venous compartment of brain arteriovenous shunts is relatively difficult to evaluate, even on angiography, compared with the arterial or nidal compartments, because of superimposed venous outlets and poor opacification of the draining veins. Venous stenosis, venous dilatation, deep venous drainage, and venous reflux have been reported to be potential risk factors for hemorrhagic events. Analysis of the venous compartment is crucial for the evaluation and management of patients with the brain arteriovenous shunts.

The radiologic findings of the venous compartment of brain arteriovenous shunts include developmental variations, suggesting a congenital origin of the disease, and secondary responses to the brain arteriovenous shunts, such as collaterals. These structural changes can be regarded as the anatomic adaptations to high-flow shunts. Thus, the radiologic findings are extremely variable according to the hemodynamics and are modified by the development of venous stenosis or ectasia.

The venous ectasia of brain arteriovenous shunts is a progressive response of the venous wall to the high-flow situation. Thus, this anatomic change is regarded as a failure to find an efficient exit to relieve the increased pressure. The venous ectasia can be defined as "any change in venous caliber in the venous runoff or drainage from the brain arteriovenous shunts, with a >2-fold caliber change in any draining venous channel". The venous stenosis is defined as a narrowing of any draining venous outflow pathway in two angiographic views. Focal venous ectasia (venous pouch) can resemble a cystic intracranial mass lesion. Because there is little evidence that most brain arteriovenous shunts are present at birth, cystic intracranial lesions during the intrauterine or neonatal period seen by ultrasonography can easily be misinterpreted as arachnoid cysts or cystic brain neoplasms. Further investigations, such as with Doppler ultrasonography or MRI, might be considered to differentiate the previously described diagnostic possibilities. Venous pouches are one of characteristic findings in patients with hereditary hemorrhagic telangiectasia (HHT) who have brain arteriovenous fistulas. A large venous pouch on MRI and multiple brain arteriovenous shunts should raise the suspicion of HHT. In the author experience, multiple brain arteriovenous malformations in the same patient occurred in 50% of the patients with HHT.

Venous ectasias can become so large that they cause local or generalized mass effect and symptoms of increased intracranial pressure. Large venous pouches are more frequently seen in children with brain arteriovenous shunts compared with adults. In adults, large venous ectasias may reflect the size of the malformation and the insufficient
Anatomic venous obstacles, such as the tentorial ridge, deep sylvian fissure, or Monro foramen, may compress the draining vein directly. This results in a proximal stenosis with distal ectasia of the draining vein. When the compression is caused by bridging arteries, however, the venous stenosis may not be associated with an upstream ectasia. In addition, converging venous systems (deep venous systems, posterior portion of the basal vein, and deep Sylvian vein) have a higher chance of developing venous ectasias than do diverging venous systems, such as cortical veins.

Thrombosis of the venous drainage pathway also represents a venous flow disturbance. The draining vein may thrombose partially or completely. The sequential CT and MRI examinations may show progressive clot formation within the venous compartment of an brain arteriovenous shunts. With partial or complete thrombosis of a common venous channel for both the surrounding brain parenchyma and brain arteriovenous shunts, brain edema may be demonstrated.

Angiography seldom reveals a partially thrombosed vein but may show a relatively delayed visualization of the venous compartment of the brain arteriovenous shunts. If the brain arteriovenous shunts has more than one draining vein and some of them are thrombosed, the shunted blood flow may be rerouted to a nonthrombosed draining vein or veins or recruit adjacent veins that have not been used by the brain arteriovenous shunts. Because the drainage of brain arteriovenous malformations is usually predictable from the location of the nidus, unusual venous drainage patterns for the nidus location may suggest partial or complete thrombosis of the venous compartment of the brain AVM.

The venous reflux into a sinus or a deep vein has been reported to be a risk factor for hemorrhagic events. Venous reflux into the cortical veins can also be demonstrated on angiography. The venous reflux should be differentiated from venous collaterals. The venous reflux is the reversal of flow in any venous outflow pathway in a direction other than the normal pathway, but venous collaterals maintain their normal drainage pathway.

**DIAGNOSIS:**

**MULTIPLE DURAL ARTERIOVENOUS FISTULAS IN THE TRANSVERSE SEGMOIDAL SINUS AND CAVERNOUS SINUS.**

**DISCUSSION:**

Cranial dural arteriovenous fistulas (DAVFs) are a unique neurovascular entity, representing 10-15% of all intracranial arteriovenous lesions [1]. In the literature many dural arteriovenous fistulas have been referred to as "malformations" in adults, however, we prefer the term "fistulas". dural arteriovenous fistulas consist of one or more direct arteriovenous connections within the dura mater. This anatomic location clearly discerns dural arteriovenous fistulas from the pial arteriovenous malformations (AVMs). It is generally accepted that dural arteriovenous fistulas are acquired [2-5] as opposed to the pial arteriovenous malformations that are thought to be congenital [6,7]. The rare exception is in the pediatric age group where congenital dural malformations are associated with high-flow single or multifocal fistula.

DAVFs can be found anywhere along the dura mater, both cranial and spinal. Spinal dural arteriovenous fistulas present with a chronic progressive myelopathy caused by venous hypertension in the perimedullary venous plexus. There are only a few reports of hemorrhage related to a spinal dural arteriovenous fistula, and these rare lesions are in the craniocervical location [8]. By contrast, cranial dural arteriovenous fistulas present with a diverse spectrum of clinical signs and symptoms including hemorrhage. The clinical findings in intracranial dural arteriovenous fistulas may be related to the fistula itself (ie, bruit) or the venous hypertension in the involved venous territory. The venous hypertension can involve the orbit or the brain depending on the venous drainage. If the brain is involved, both hemorrhagic and nonhemorrhagic neurologic deficit can occur.

- **History**

Dural arteriovenous fistulas were rarely identified before 1960. In the seventies, dural arteriovenous fistulas were recognized as a distinct entity caused by the advances in angiography that included magnification, subtraction techniques, and selective arterial catheterization [9]. At first, these dural lesions were regarded as a benign disease in
In 1984, Malik et al. [16] published their review of 223 cases, emphasizing that restriction of venous outflow was a key factor in the clinical expression of the disease. They did not emphasize, however, the importance of the cortical venous reflux. Lasjaunias et al. [17] published a meta-analysis of 191 cases in addition to their four illustrative cases, concluding that focal neurologic symptoms were dependent on the territory of the draining veins, and that cortical venous reflux carries a high risk of intradural bleeding. Awad et al. [18] reviewed 377 cases, mostly from the literature, and introduced the term "aggressive" for lesions presenting with a hemorrhage or a focal neurologic deficit. Awad et al. also emphasized the importance of cortical venous reflux and included venous ectasias and galenic drainage as risk factors for an aggressive course.

**Pathogenesis**

The etiology of cranial dural arteriovenous fistulas is unknown. Dural arteriovenous fistulas have been described after surgery, head trauma, and in relation to sinus thrombosis. Two hypotheses of the pathogenesis have been proposed. The first hypothesis claims that cranial dural arteriovenous fistulas arise from already existing "dormant" channels between the external carotid circulation and the venous pathways within the dura mater. Histopathologic and radioanatomic studies have shown that these communications are normally present in the dura [19]. These channels open because of the venous hypertension associated with sinus thrombosis or sinus outflow obstruction [4,12,15,18,20]. A variation to this theme is the reported existence of thin-walled venous pouches within the dura, close to small arteries. Rupture of these fragile pouches easily induces arteriovenous communications within the dura [21,22]. The second hypothesis claims that dural arteriovenous fistulas are the result of new vascular channels that are stimulated by angiogenetic factors. These factors, such as VEGF and bFGF, originate either directly from the organization of a sinus thrombosis or indirectly because of tissue hypoxia related to an increased intraluminal venous pressure [3,5,23-25].

Histopathologically, the true arteriovenous fistula has no intervening capillary bed and consists of small venules with a diameter of approximately 30μ. These vessels have been called "crack-like vessels," because they look like cracks in the dural sinus wall after histologic staining [26]. Furthermore, intimal thickening of both dural arteries and dural veins has been observed [27]. Although the fistula initially has been described within a thrombosed sinus, it is generally accepted that the fistula is located within the wall of the sinus. This explains the existence of dural arteriovenous fistulas that drain directly into the pial venous network [25,28].

**Classification**

The terms "benign" and "aggressive" can be applied to the symptomatology and natural history of cranial dural arteriovenous fistulas [18,29-32]. Nonhemorrhagic neurologic deficits (NHND), hemorrhage, and death are considered aggressive. Chronic headache, pulsatile bruit, and orbital symptoms including cranial nerve deficit are considered benign.

The anatomic location of a dural arteriovenous fistula initially was felt to be important in its association with the venous drainage pattern and symptomatology [33]. Cavernous sinus and transverse sinus lesions often have sinosal drainage only, whereas anterior cranial fossa and tentorial lesions almost all have cortical venous reflux. Recently, it has become evident that any location can develop cortical venous reflux, including the cavernous sinus and transverse sinus lesions. Aminoff grouped dural arteriovenous fistulas by location, separating them into an anteroinferior group and a posteroinferior group [12]. Later, authors pointed out a relationship between the location of the dural arteriovenous fistula and the behavior of the dural arteriovenous fistula [18,34]. Malik et al. hypothesized that dural arteriovenous fistulas in specific locations have a higher likelihood of developing cortical venous reflux [16]. Malik et al. related cortical venous reflux to the presence or absence of a dural sinus at the site of the fistula. Although no location is immune from aggressive behavior [18], certain locations are more prone to have cortical venous reflux.

Several classification schemes for cranial dural arteriovenous fistulas have been introduced [15,35-37]. Classification schemes must be used with caution because dural arteriovenous fistulas are a dynamic process and their angioarchitecture can be altered by venous thrombosis. The classifications of Borden et al. and Cognard et al. are the most widely used (Table 1). Although the three-step classification of Borden has the advantage of being relatively
simple to apply, the Cognard classification (a revision of the Djindjian classification scheme) incorporates the additional influence of retrograde flow in the sinus and spinal venous drainage. Retrograde flow can prohibit the cortical veins from draining into the involved sinus because of venous hypertension in the sinus leading to venous congestion of the brain without cortical venous reflux. Both classifications have been validated by Davies et al [33]. Borden I, Cognard I, and Cognard IIa lesions are considered benign, whereas all other Borden and Cognard grades should be considered aggressive.

In the pediatric age group, Lasjaunias has recognized three types of dural arteriovenous fistulas [6]. These include the dural sinus malformations with arteriovenous shunting, infantile dural arteriovenous shunts, and the adult-type dural arteriovenous fistulas. The dural sinus malformations and high-flow infantile arteriovenous shunts in the pediatric age group are beyond the scope of this article.

- **Symptomatology**

The majority of the fistulas present with the so-called "benign" symptoms, either a pulsatile tinnitus or orbital congestion (Table 2). The patients with pulsatile tinnitus usually have an objective bruit (Fig. 9). Other than pulsatile tinnitus, the symptoms relate to the venous drainage of the fistula and are commonly remote from the fistula itself. Fistulas can present at any age, although patients are often over 50 years of age. Adult-type dural arteriovenous fistulas in childhood are rare. Patients presenting with orbital congestion frequently have cavernous sinus dural arteriovenous fistulas (Fig. 10). The orbital signs include chemosis, conjunctival injection, and proptosis. The orbital symptoms may be progressive and to the patient may not be considered "benign." Patients may develop an ophthalmoplegia related to a cranial nerve dysfunction or extraocular muscle swelling. Orbital congestion may result in raised intraocular pressure and lead to loss of visual acuity.

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**Table 1. Classification of cranial dural arteriovenous fistulas**

<table>
<thead>
<tr>
<th>Borden classification</th>
<th>Cognard classification</th>
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<tbody>
<tr>
<td></td>
<td>I Venous drainage into dural venous sinus with antegrade flow</td>
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<tr>
<td></td>
<td>IIa Venous drainage into dural venous sinus with retrograde flow</td>
</tr>
<tr>
<td></td>
<td>IIb Venous drainage into dural venous sinus with antegrade flow and cortical venous reflux</td>
</tr>
<tr>
<td></td>
<td>IIa+b Venous drainage into dural venous sinus with retrograde flow and cortical venous reflux</td>
</tr>
<tr>
<td></td>
<td>III Venous drainage directly into subarachnoid veins (cortical venous reflux only)</td>
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<tr>
<td></td>
<td>IV Type III with venous ectasias of the draining subarachnoid veins</td>
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*Abbreviation: CVR; cortical venous reflux.*

**Table 2. Clinical findings in cranial dural arteriovenous fistula**

<table>
<thead>
<tr>
<th>Common signs and symptoms</th>
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</thead>
<tbody>
<tr>
<td>Pulsatile tinnitus</td>
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<tr>
<td>Objective bruit</td>
</tr>
<tr>
<td>Proptosis, conjunctival injection, chemosis</td>
</tr>
<tr>
<td>Ophthalmoplegia</td>
</tr>
<tr>
<td>Visual loss</td>
</tr>
<tr>
<td>Glaucoma</td>
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<tr>
<td>Transient ischemic attacks</td>
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<td>Aphasia</td>
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<tr>
<td>Motor weakness</td>
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Figure 9. "Benign" type dural arteriovenous fistula in a 40-year-old male patient with pulsatile tinnitus. (A) Lateral left ascending pharyngeal artery (straight arrow) and (B) left occipital artery (straight arrow) angiograms show a network of arterial branches participating in a shunt into the jugular bulb (curved arrow) (A) and sigmoid sinus (curved arrow) (B).

Figure 10. Anterior cavernous sinus dural arteriovenous fistula. Transvenous endovascular treatment in 36-year-old female patient with visual loss. (A) Coronal T1-weighted and (B) T2-weighted MR image show an enlarged superior ophthalmic vein (arrow) (A) and a dilated inferior lateral trunk artery (ILT) (arrow) (B). (C) Lateral internal carotid
artery angiogram shows a high-flow shunt into the anterior cavernous sinus (open arrow) and drainage exclusively into the orbit. (D) Lateral radiograph shows a microcatheter (small arrows) coursing through the transverse facial vein and superior ophthalmic vein. Coils (solid arrow) have been deposited into the cavernous sinus at the fistula site. (E) Postembolization lateral internal carotid angiogram shows that the fistula is closed."Aggressive" symptoms include neurologic dysfunction related to intracranial hemorrhage or venous congestion. The hemorrhage can be subarachnoid, subdural, or intracranial. The neurologic dysfunction can include aphasia, weakness, transient ischemic attacks, seizures, and dementia. Papilledema and communicating hydrocephalus can be presenting findings. Heart failure and craniomegaly are only seen in the dural sinus malformations and infantile dural arteriovenous fistulas of childhood.

- Location

The location of dural arteriovenous fistula is defined by the site of the arteriovenous shunting. The venous drainage of the fistula is influenced by the degree of venous obstruction of the involved sinus. Direct fistulas into cortical veins result from complete obstruction of the adjacent sinus. Table 3 outlines the common locations of dural arteriovenous fistulas and their venous drainage. The venous drainage is variable and may evolve with time. Change in the venous drainage is caused by venous thrombosis or may be the consequence of a hemorrhage.

<table>
<thead>
<tr>
<th>Location</th>
<th>Venous drainage</th>
</tr>
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<tbody>
<tr>
<td>Anterior cranial fossa</td>
<td>Frontal and olfactory veins</td>
</tr>
<tr>
<td>Anterior cavernous sinus</td>
<td>Ophthalmic and deep sylvian veins</td>
</tr>
<tr>
<td>Posterior cavernous sinus</td>
<td>Superior and inferior petrosal sinuses; ophthalmic, deep sylvian, uncal and anterior pontomesencephalic veins</td>
</tr>
<tr>
<td>Sphenoid wing</td>
<td>Deep sylvian veins</td>
</tr>
<tr>
<td>Transverse sinus</td>
<td>Sigmoid sinus; temporal and cerebellar veins</td>
</tr>
<tr>
<td>Torcular</td>
<td>Transverse sinus; medial occipital and infratemporal veins</td>
</tr>
<tr>
<td>Tentorial</td>
<td>Basal vein of Rosenthal, lateral mesencephalic and tentorial veins</td>
</tr>
<tr>
<td>SSS</td>
<td>SSS; cortical veins</td>
</tr>
<tr>
<td>Inferior straight sinus</td>
<td>Vermian veins</td>
</tr>
<tr>
<td>Foramen magnum</td>
<td>Marginal sinus; lateral pontomesencephalic and spinal veins</td>
</tr>
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Abbreviation: SSS; superior sagittal sinus.

**TYPES OF CRANIAL DURAL ARTERIOVENOUS FISTULAS**

- Benign fistulas

Awareness of the natural history of a given disease is essential for patient management. The results of any active treatment must be compared with the outcome of the natural history of that disease. The recognition of the so-called benign fistulas is helpful in planning a management strategy; the evolution of the disease must be carefully monitored, however. A change in symptomatology may be related to either reduced flow through the malformation or rerouting of the venous drainage. A fistula that originally had anterograde sinosal drainage could convert to a fistula with retrograde sinosal drainage or develop cortical venous reflux.

In the absence of cortical venous reflux, cranial dural arteriovenous fistulas have a benign presentation and, in most cases, an uneventful clinical course [33,38]. There are only three publications focused on the natural history and angiographic follow-up of dural arteriovenous fistulas without cortical venous reflux. Davies et al [29] reported their experience with a cohort of 54 cases without cortical venous reflux over a mean follow-up period of 33 months. Only one of the cases (2%) died after palliative endovascular treatment; there was, however, no evidence of angiographic conversion into a lesion with cortical venous reflux. This unusual course resulted from the development of venous hypertension caused by functional obstruction of the superior sagittal sinus. In conclusion, Davies et al reported that
the majority of cranial dural arteriovenous fistulas without cortical venous reflux behave in a benign fashion and that the focus of therapeutic efforts, if necessary, should be directed toward palliation rather than toward angiographic cure. Over 80% of the patients not treated had clinical improvement during the mean follow-up of over 2 years.

Cognard et al [39] reported seven patients that initially had a dural arteriovenous fistula without cortical venous reflux and later converted to a dural arteriovenous fistula with cortical venous reflux. The rate of conversion of dural arteriovenous fistulas without cortical venous reflux to those that develop cortical venous reflux cannot be ascertained as the total number of patients was not included in the report. Five patients were embolized with particles, one patient had proximal ligation of the occipital and middle meningeal artery, and one patient had conservative management. All of them showed a worsening in the venous drainage pattern during a follow-up between 1 month and 20 years (mean, 7 years). Two cases, both embolized, demonstrated a change from antegrade to retrograde flow into the draining sinus, and five cases developed cortical venous reflux. In all cases, the change in venous pattern was accompanied by a worsening of the clinical symptoms.

In a more recent study by Satomi et al [31], the chronologic change in clinical symptoms and angiographic features of 117 patients harboring cranial dural arteriovenous fistula without cortical venous reflux were evaluated. None of the cases presented with either intracranial hemorrhage or Nonhemorrhagic neurologic deficits, and the majority of these lesions were managed conservatively. Palliative treatment, never aimed at cure, was performed if the patient had intolerable symptoms or if there was a persistent high intraocular pressure or decreasing visual acuity. Sixty-three percent of the patients had no invasive treatment. Using this conservative management, 98% of the patients had a well-tolerated clinical course. In Satomi et al's series, five cases showed a change in their venous drainage pattern associated with progressive thrombosis of the venous outlets during the follow-up period. Three patients with cavernous sinus dural arteriovenous fistulas (conservative management in 1 case, palliative embolization in 2 cases) had an alteration in sinus drainage direction from antegrade to retrograde, after which the lesions eventually resolved spontaneously. The two other cases showed angiographic conversion into a lesion with cortical venous reflux, with symptom resolution in one case and symptom aggravation in the other. This demonstrates that dural arteriovenous fistulas without cortical venous reflux have the potential to develop cortical venous reflux even without treatment. All five cases with angiographic conversion were associated with progressive steno-occlusive change in the affected sinus without arterial flow increase or de novo arteriovenous shunts. Stenosis or thrombosis of the secondarily occluded venous outlets was not present on the initial angiogram in these cases.

In conclusion, the disease course of a cranial dural arteriovenous fistula without cortical venous reflux is benign in most cases, obviating the need for a cure of these lesions. Symptoms are well tolerated with either observation or palliative treatment. A dural arteriovenous fistula of the benign type has a 2-3% potential of developing cortical venous reflux, however, mandating close clinical follow-up and renewed radiologic evaluation with any deterioration or change in symptoms.

- **Aggressive fistulas**

Cranial dural arteriovenous fistulas with cortical venous reflux are considered aggressive lesions. They often present with intracranial hemorrhage or Nonhemorrhagic neurologic deficits [4,14,16-18]. The disease course after their aggressive presentation is less well understood, however. The only studies predominantly focused on events after presentation were performed by Duffau et al [40], Brown et al [34], and Davies et al [30]. Duffau et al looked at their short time frame between the diagnosis and treatment, which lasted a mean of only 20 days. Brown et al followed patients for mean 6.6 years but did not specifically select for cortical venous reflux. In 1997 Davies et al [30] calculated an annual mortality of 19.2%, with a 19.2% annual rate of hemorrhage and a 10.9% annual rate of Nonhemorrhagic neurologic deficits during the disease course of dural arteriovenous fistulas with persistent cortical venous reflux. A recalibration of Davies et al's series was performed in 2002 by Van Dijk et al [32], based on a larger population and four times the follow-up time. This yielded an annual mortality rate of 10.4%. In addition, disregarding aggressive events at presentation, the annual risk of intracranial hemorrhage or Nonhemorrhagic neurologic deficits was 8.1% and 6.9%, respectively, adding up to a 15.0% annual adverse event rate. These numbers mandate a prompt, accurate diagnosis and treatment of these aggressive lesions.

**NEUROIMAGING OF CRANIAL DURAL ARTERIOVENOUS FISTULAS**

- **Dural arteriovenous fistulas without cortical venous reflux**

MR imaging of the brain parenchyma in patients with dural arteriovenous fistulas without cortical venous reflux is typically normal. The involved dural sinuses may be irregular, stenotic, or septated on MR imaging. MR angiography may confirm the occlusive changes within the involved sinus. Hydrocephalus may be present in any dural
arteriovenous fistula that causes venous hypertension in the superior sagittal sinus. The venous hypertension may result from retrograde flow in the superior sagittal sinus. Cortical venous reflux may not be present. The venous hypertension interferes with the cerebrospinal fluid absorption by the pacchionian granulations.

- **Dural arteriovenous fistulas with cortical venous reflux**

MR imaging is often positive in dural arteriovenous fistulas with cortical venous reflux. In Willinsky et al's review, 10 of the 13 patients (77%) with dural arteriovenous fistulas and cortical venous reflux had dilated pial vessels. Two patients had hydrocephalus. Diffuse white matter edema, in the cerebellar or cerebral hemispheres, was present on MR imaging in 4 patients and correlated with neurologic deficits. In two of these four patients gadolinium enhancement surrounding the area of T2 hyperintensity was seen in the periphery of the involved hemisphere.

![Image of cerebellar venous congestion secondary to a straight sinus dural arteriovenous fistula with cortical venous reflux.](figure11)

**Figure 11.** Cerebellar venous congestion secondary to a straight sinus dural arteriovenous fistula with cortical venous reflux. (A) T2-weighted MR image shows central hyperintensity (open arrow) in the cerebellar hemisphere with peripheral hypointensity and prominent flow voids (closed arrows). (B) T1-weighted gadolinium-enhanced MR image shows peripheral enhancement adjacent to the T2 hyperintense lesion. (C) Early and (D) late phases of the selective angiogram of a dural branch (small arrows) of the right vertebral artery show a fistula in the wall of the straight sinus. The drainage is retrograde into the inferior vermian vein (large arrows) that then reflexes into the cerebellar hemisphere veins (D).
Figure 12. Post-treatment resolution of venous congestion. (A) FLAIR MR image shows central hyperintensity (arrow) in the cerebellum. (B) Gadolinium-enhanced MR image shows peripheral enhancement (arrow) surrounding the hyperintense region seen on the FLAIR. (C) Lateral right vertebral angiogram shows a dural arteriovenous fistula (open arrow) fed by a dural branch (arrowhead) of the vertebral artery and draining into the inferior vermian vein (curved arrow). (D) Late phase of the AP right vertebral angiogram shows a PPP in the right cerebellar hemisphere related to venous congestion. (E) Posttreatment (embolization and surgery) FLAIR MR image shows resolution of the venous congestion.

The term venous congestive encephalopathy (VCE) was introduced in 1994 to describe those patients who present with cranial neurologic deficits caused by venous hypertension [41]. This entity is analogous to the venous congestive myelopathy of the spinal cord in the presence of a spinal dural arteriovenous fistula [42]. On CT, the venous congestion may be evident as an area of edema and mass effect. In patients with cortical venous reflux, MR imaging often shows prominent flow voids on the surface of the brain. Hydrocephalus may be secondary to the venous hypertension in the superior sagittal sinus. On MR imaging, T2 hyperintensity deep in the brain parenchyma may be evident secondary to the venous hypertension and passive congestion of the brain. The cerebellum, cerebrum, and deep gray nuclei or brainstem may be affected. In chronic cases, the proton density or T2-weighted images may show a central hypointensity that may be related to hemosiderin deposition from chronic venous congestion. In the cerebral hemispheres, the deep white matter is the most vulnerable to the venous congestion [41]. The T2 hyperintensity may be reversible after treatment. The differential diagnosis of the T2 hyperintensity would include a superior sagittal sinus thrombosis with a venous infarction or venous congestion, demyelination, or a dysmyelination and neoplasm [43]. But the combination of a surplus of pial vessels, T2 hyperintensity deep within the brain, and peripheral enhancement is highly suggestive of a dural arteriovenous fistula and mandates prompt angiography.
Figure 13. CT/MR image correlation of venous congestion. (A) Enhanced CT shows central edema in the left temporal lobe with tortuous, prominent vessels deep in the sulci (arrows). (B) Proton density weighted MR image shows central hypointensity (curved arrow) in the left temporal lobe and dilated, tortuous vessels in the sulci (small arrows). (C, D) Early and late phases of a lateral left occipital artery angiogram shows shunting into the transverse sinus and cortical venous reflux (closed arrows). Note that the transverse sinus is occluded both proximally and distally (open arrows). The late phase (D) shows the extent of the venous reflux, both supratentorial and infratentorial.
Figure 14. Chronic venous congestion in the temporal/occipital region. (A) Sagittal T1-weighted MR image shows linear hyperintensity (arrow) in the occipital cortex with a central hypointensity. The T1 hyperintensity relates to infarction from the chronic venous congestion. (B) T2-weighted MR image shows central hyperintensity (curved arrow) in the temporal/occipital region and tortuous, dilated vessels (closed arrows) on the surface of the brain. (C) Gadolinium-enhanced MR image better delineates the abnormal vessels (arrows). Note the burr hole (curved arrow) from the previous biopsy. (D) Lateral distal external carotid artery angiogram shows a shunt into the transverse sinus (open arrow) which is occluded distally. Note the cortical venous reflux (closed arrows).

At angiography, a delayed circulation time is compatible with venous congestive encephalopathy [44]. This is analogous to the delayed circulation time seen in the venous congestive myelopathy related to a spinal dural arteriovenous fistula [45]. Angiography outlines the arterial feeders to the dural arteriovenous fistula, as well as the venous drainage of the fistula. Often, venous sinus occlusions are evident and contribute to the venous hypertension and reflux into the cortical or cerebellar veins. Careful scrutiny is needed in the detection of cortical venous reflux. Global nonselective angiography should be avoided as subtle cortical venous reflux will be missed. Selective, magnification, subtraction angiography is essential to visualize the cortical venous reflux. A complete assessment of the cranial circulation is important as 7-8% of patients have multiple dural arteriovenous fistulas [46,47].
Figure 15. Hydrocephalus and the PPP. (A) T2-weighted MR image shows dilated lateral ventricles and prominent flow voids deep in the sulci (arrows). (B, C) Early and late phases of a left occipital artery angiogram shows shunting into the distal transverse sinus with retrograde flow into an anomalous dural sinus in the parietal region (open arrow) (B). (C) In the late phase, cortical venous reflux is evident (closed arrows). (D) Late phase of the right internal carotid artery angiogram shows a PPP in the cerebral hemisphere indicative of venous congestion. (From Willinsky R, Goyal M, TerBrugge K, et al. Tortuous, engorged pial veins in intracranial dural arteriovenous fistulas: correlations with presentation, location, and MR findings in 122 patients.

The exact anatomic position of the cortical venous reflux must be clearly defined to allow appropriate treatment planning. Angiography is critical for the detection of the venous reflux and the assessment of the venous drainage of the brain. Focal areas of delayed venous drainage in the brain correspond to the site of cortical venous reflux. Often tortuous, dilated collateral veins develop in the region of the cortical venous reflux. This is in response to the venous hypertension secondary to the cortical venous reflux. Willinsky et al [44] described this finding of tortuous, dilated, engorged veins and referred to it as a pseudophlebitic pattern (PPP). PPP is associated with venous rerouting into dilated transosseous venous channels or retrograde flow into orbital veins. PPP is as a sign of venous congestion of the brain and may correlate with an aggressive natural history. In their report, no location of a cranial dural arteriovenous fistula was immune to PPP. Although the presence of PPP is typically concurrent with the existence of cortical venous reflux, sporadic cases of PPP without cortical venous reflux were found and may be a sign that correlates with a more aggressive natural history.
Figure 16. Venous congestion of the brainstem in a foramen magnum dural arteriovenous fistula with cortical venous reflux. (A) T2-weighted MR image shows a central hyperintensity in the medulla and a few prominent vessels on the surface of the brainstem. (B) AP right vertebral artery angiogram shows a shunt at the foramen magnum (open arrow) with drainage into the anterior spinal vein (closed arrow). (C) Lateral selective angiogram of a dural branch from the right vertebral artery shows the drainage into the spinal vein (single arrow) and the posterior fossa veins (double arrow). (D) Post-treatment (embolization and surgery) T2-weighted MR image shows resolution of the congestion.

MANAGEMENT OF CRANIAL DURAL ARTERIOVENOUS FISTULAS

Treatment of dural arteriovenous fistulas is done to improve on the natural history of the disorder. Therefore, observation should be considered as a completely valid treatment option in patients with dural arteriovenous fistulas without cortical venous reflux who are tolerating their symptoms. No treatment is especially important in the elderly patient with a well-tolerated slow-flow fistula of the cavernous sinus without cortical venous reflux. Patients with cranial dural arteriovenous fistulas without cortical venous reflux have a 98% chance of having a benign course with no curative treatment, indicating that observation with timely imaging re-evaluation is the best available treatment [31]. The imaging used in follow-up should include MR with MR angiography. MR angiography with gadolinium may be more accurate in the assessment of cortical venous reflux and therefore should be helpful for the routine follow-up of dural arteriovenous fistulas that are being managed conservatively. A 3-year-follow-up angiogram is advised in patients with stable clinical signs and symptoms. If there is any change in the clinical status, either worsening or improvement in symptoms, repeat angiography is needed to look for the development of cortical venous reflux or progressive venous thrombosis and retrograde flow in the dural sinuses.

Intermittent manual carotid compression by the patient has been used by Halbach et al [48] to treat dural arteriovenous fistulas. The patient is taught to compress the carotid artery with his contralateral hand and stop compressing if any weakness develops. They report a cure rate of 30% with this technique after 4-6 weeks. This treatment has been the subject of debate because the 30% cure rate in the short term may reflect the natural history of the disease.

Endovascular embolization has been demonstrated to be a valid treatment of cranial dural arteriovenous fistulas with cortical venous reflux [13,49-53]. The same has been demonstrated for surgical therapy [54-58]. It is still a debate whether total resection or obliteration of the fistula is necessary, or if simple disconnection of the refluxing cortical veins from the fistula will yield the same result [59]. There are a limited number of reports on the radiosurgical treatment of cranial dural arteriovenous fistulas [60-63]. In patients with cortical venous reflux, the delayed efficacy of radiosurgery is not acceptable as the adverse event rate in these patients is 15% per year. Because most cranial dural arteriovenous fistulas without cortical venous reflux do not need treatment, the risk of radiosurgery in these patients may not be acceptable, especially in view of the limited data available on the efficacy of this treatment.

Cranial dural arteriovenous fistulas with cortical venous reflux require a multidisciplinary approach to treatment. Endovascular treatment is often the primary treatment modality. The primary goal of treatment, either endovascular or surgery, is to eliminate the cortical venous reflux. Complete closure of the fistula is the ideal treatment but not critical, as eliminating the cortical venous reflux should only be effective in eradicating the risk of intracranial hemorrhagic or nonhemorrhagic deficits. Endovascular treatment often begins with arterial embolization. The ideal goal is to occlude the fistula itself which involves reaching the venous side of the fistula. Liquid adhesive agents are the best agent to permanently occlude a fistula. Particle embolization through the feeding pedicles is satisfactory as a palliative treatment in dural arteriovenous fistulas without cortical venous reflux or as an adjunct to the definitive treatment on the venous side, either endovascular or surgical. The endovascular treatment on the venous side is done
retrograde through the veins with deposition of coils into the venous compartment at the fistula site. Transvenous packing of the involved dural sinus is commonly done for transverse sinus and cavernous sinus dural arteriovenous fistulas with cortical venous reflux. Sacrifice of an involved transverse sinus can only be considered after a thorough understanding of the venous drainage of the brain. In many of these patients, the brain has developed alternative pathways for its venous drainage because of the high pressure in the involved venous sinus and/or pre-existing venous occlusive disease.

Endovascular treatment is indicated for cavernous dural arteriovenous fistulas without cortical venous reflux when the intraocular pressures cannot be controlled medically. If left untreated, these patients will develop permanent visual loss. The transvenous femoral approach to the cavernous sinus can be done through the petrosal sinus or, infrequently, through facial veins. After transvenous coil deposition into the cavernous sinus, there may be worsening of the venous hypertension in the orbit caused by venous thrombosis within the orbit. This may require oral steroids and anticoagulation in order to spare the vision.

Patients with cavernous dural arteriovenous fistulas that are being followed conservatively need careful monitoring by a neuro-ophthalmologist. Spontaneous venous thrombosis in the orbit may develop in these patients. This results in worsening of the eye signs and symptoms and raised intraocular pressure. Initially, the raised intraocular pressure may respond well to topical medical treatment. Patients may require steroids and systemic anticoagulation to save their vision. The neuro-ophthalmologist can supervise the medical therapy and is the key person to decide if surgical treatment of the hypertensive glaucoma is needed. Failure of medical therapy is an indication for prompt endovascular treatment.
Surgical treatment may involve one of two strategies. The first strategy is surgical disconnection of the cortical venous reflux without treating the actual fistula. The second strategy is skeletonization of the involved dural sinus with interruption of the dural arterial supply and preservation of the cortical veins. Both types of surgical treatment benefit from preoperative embolization of the arterial feeders. The goal of disconnection alone is to eliminate the future risk of hemorrhagic or nonhemorrhagic deficits. Disconnection alone will convert a Borden 2 dural arteriovenous fistula to a Borden 1 dural arteriovenous fistula and thereby favorably alter the natural history. Disconnection alone is often a less formidable task compared with skeletonization of the involved sinus. Disconnection alone is only feasible when the cortical venous reflux is anatomically limited to a limited area. Skeletonization of the dural sinus requires a large operation as the entire dural sinus must be isolated from its dural arterial supply.

SUMMARY

Cranial dural arteriovenous fistulas present with a wide spectrum of clinical findings from pulsatile tinnitus alone to intracranial hemorrhage and Nonhemorrhagic neurologic deficits. The neurologic sequelae are a consequence of venous hypertension and venous congestion. Dural arteriovenous fistulas with cortical venous reflux can present with or develop a venous congestive encephalopathy that can be recognized on MR imaging as a diffuse T2 hyperintensity in the deep white matter of the cerebral or cerebellar hemispheres. The T2 hyperintensity has a characteristic peripheral enhancement. The telltale sign on MR imaging is the plethora of prominent pial vessels on the surface of the brain that are the engorged cortical veins participating in the cortical venous reflux. Selective angiography is critical for the accurate assessment of the cortical venous reflux. Dural arteriovenous fistulas with cortical venous reflux require prompt treatment, either endovascular alone or a combination of endovascular treatment and surgery.

Addendum

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