

### Intracranial Dural Arteriovenous Fistulae

Intracranial dural arteriovenous fistulae (DAVF) are uncommon lesions. Their true incidence is unknown, although selected series suggest that they occur only one in ten times as frequently as intraparenchymal arteriovenous malformations (AVM). Since many may remain clinically silent or involve spontaneously the incidence may be an underestimate. DAVF tend to present later in life than AVMs, lending support to the theory that these are acquired lesions, although presentation can be at any age. A presentation with aggressive neurological symptoms is more common in males.

The fistula represents an abnormal connection between dural arteries or pachymeningeal branches of cerebral arteries and dural veins. Occasionally, as a fistula grows or becomes more diffuse, pial recruitment from parenchymal veins may occur, predisposing the patient to intracranial hypertension, which acts as the initiating factor opening up microscopic vascular connections within the dura. Maturation of these channels secondary to progressive venous stenosis or occlusion results in the development of direct shunts between the arteries and dural veins. In addition, a secondary complementary mechanism of DAVF evolution may occur with the release of angiogenic growth factors such as vascular endothelial growth factor (VEGF) and basic fibroblast growth factor (bFGF) promoting neovascularisation and development of a DAVF.

### Presentation and natural history

A wide spectrum of symptoms exists, ranging from the benign to the more aggressive. Individual lesions may regress spontaneously or follow a benign course over years. Drainage of a petrous region DAVF to the transverse or sigmoid sinus commonly produces pulsatile tinnitus, sometimes in association with an audible bruit. Depending upon the pattern of venous drainage such patients may be managed conservatively. Cavernous sinus DAVFs may develop orbital signs such as congestion, chemosis and ophthalmoplegia. Treatment is usually undertaken to protect against ocular and visual complications.

More aggressive behaviour may manifest as focal neurological deficits, a dementia-type of syndrome or cerebrovascular events. Treatment is usually undertaken to protect against ocular and visual complications. The two commonly used classification systems are shown in Tables 1 and 2.

### Classification

Various classification methods have been adopted that attempt to explain the significance of the angiographic anatomy; namely, the pattern of venous drainage and the clinical presentation and outcome. The two commonly used classification systems are shown in Tables 1 and 2. However, the Cognard system is more detailed and elaborates on the direction of flow, whether normal (anterograde) or retrograde and the presence or absence of cortical venous reflux. Such definition enables more accurate comparison of clinical and radiological parameters. In addition, spinal perimedullary venous drainage is specifically recognised.

In a retrospective review of 102 DAVFs in 98 patients Davies et al. reported a significant correlation between

---

**Table 1: The Borden Classification system.**

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>DAVF drainage into a dural venous sinus or meningeal vein with normal anterograde flow. Usually benign clinical behaviour.</td>
</tr>
<tr>
<td>II</td>
<td>Anterograde drainage into dural venous sinus and onwards but retrograde flow occurs into cortical veins. May present with haemorrhage.</td>
</tr>
<tr>
<td>III</td>
<td>Direct retrograde flow of blood from the fistula into cortical veins causing venous hypertension with a risk of haemorrhage.</td>
</tr>
</tbody>
</table>

**Table 2: The Cognard classification system.**

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I (Figure 1)</td>
<td>Normal anterograde flow into a dural venous sinus.</td>
</tr>
<tr>
<td>IIA</td>
<td>Drainage into a sinus with retrograde flow within the sinus</td>
</tr>
<tr>
<td>IIB (Figure 2)</td>
<td>Drainage into a sinus with retrograde flow into cortical vein(s)</td>
</tr>
<tr>
<td>IIA + b</td>
<td>Drainage into a sinus with retrograde flow within the sinus and cortical vein(s)</td>
</tr>
<tr>
<td>III (Figure 3)</td>
<td>Direct drainage into a cortical vein without venous ectasia</td>
</tr>
<tr>
<td>IV (Figure 4)</td>
<td>Direct drainage into a cortical vein with ectasia &gt;5mm and 3x larger than the diameter of the draining vein</td>
</tr>
<tr>
<td>V (Figure 5)</td>
<td>Direct drainage into spinal perimedullary veins</td>
</tr>
</tbody>
</table>
progressive tinnitus and ataxia a few weeks after an ear infection. She had sustained a minor head injury 2 years previously. Figure 2a top: Type IIb DA VF - A lateral projection left common carotid artery injection. This 68-year-old lady presented with obliteration of the DA VF on 12 month follow-up angiography.

Figure 1: Type I DA VF - Left common carotid artery DSA lateral projection in a 47-year-old lady with disabling tinnitus in the left ear. This shows a Type 1 DAVF filling via transmastoid perforators of the occipital artery and draining to the sigmoid sinus. Despite some improvement of symptoms with particulate injection she requested further treatment. Injection of a 25/75 mixture of NBCA and Lipiodol into the occipital artery, with further particulate injection of the middle meningeal artery, led to a resolution of symptoms and sustained improvement of symptoms with particulate injection she requested into the occipital artery and draining to the sigmoid sinus. Despite some
delay in the introduction of symptoms attributable to mass effect from the ectatic vein. The presence of direct cortical venous drainage was therefore a strong predictor of haemorrhage. Of those 12 patients with spinal perimedullary venous drainage, 6 presented with myelopathy.

Imaging
CT, MRI and angiography all have roles to play in the investigation of patients with a possible DAVF. Because the clinical and imaging features can be non-specific, the diagnosis of a DAVF is often delayed or missed. Occasionally plain films can demonstrate grooving within the skull vault due to chronic compression from enlarged middle meningeal vessels. If haemorrhage is suspected, non-enhanced CT is a pre-requisite. Venous congestion may appear as an area of low density on CT. In most institutions CT is more readily available and cheaper than MRI and so becomes the first-line investigation of patients presenting with tinnitus, headache or other vague neurological symptoms. Multi-detector CT angiography (MDCTA) can now provide high resolution detail of vascular anatomy. In the investigation of tinnitus it has the additional advantage that it can detect inner and middle ear abnormalities such as aberrant vascular anatomy or glomus tumours. Linear bony defects formed by enlarged emissary veins, similar to the grooving abnormality seen on plain film, can indicate the presence of a fistula. MDCTA, because of its rapid acquisition, has a temporal advantage over static CT. This is important because of the likelihood of altered flow dynamics within a fistula. Subtle changes in contrast intensity of the cerebral vessels may be evident.

Arterialised venous blood within the veins draining a DAVF has increased density when compared to non-arterialised blood. Careful scrutiny of the source images with narrow window settings are required to make this distinction.

T2 weighted MRI is more sensitive to the white matter changes of venous congestion or infarction when compared to CT. It has the drawback of being less sensitive to the changes of acute haemorrhage. If dilated cortical veins are present they may be seen on conventional spin echo sequences and visualised using MR angiographic techniques such as phase contrast venography or contrast enhanced MR angiography. Benign disease, without cortical venous reflux can be missed using both CT and MRI. Conventional catheter angiography therefore remains the investigation of choice if there is a strong clinical suspicion of a fistula.

Treatment
Treatment is dependent on the clinical picture and the grade of fistula. A multidisciplinary approach involving a neurosurgeon and neuroradiologist is required. A DAVF without angiographic evidence of retrograde sinus or cortical venous drainage and presenting with a well-tolerated or non-disabling tinnitus can be managed conservatively. Techniques such as occipital artery or carotid manual compression have been reported to occasionally lead to obliteration of the DAVF although this may correspond to the natural history of the disease. If possible, patients should avoid anti-platelet agents which might prevent thrombosis. A change in symptoms warrants repeat assessment with formal angiography.

Both surgical and endovascular techniques have proven efficacy for more troublesome DAVFs. At the benign end of the spectrum, particulate trans-arterial embolisation may afford palliation (Figure 1). This can be regarded as fairly low-risk but the risks should not exceed the natural history of the disease. In addition, further recruitment of fistulous channels may cause re-emergence of symptoms at a later date. For more severe disease and particularly for those patients presenting with intracranial haemorrhage or progressive neurological symptoms, treatment is indicated (Figures 2, 3 and 4). If the DAVF drains directly to cortical veins without involvement of the sinus, surgical disconnection of the arteriised draining vein(s) can be employed. This minimises the risk of future intracranial haemorrhage and is facilitated by the use of neuronavigation to localise the cranialotomy. Surgery can be combined with pre-operative particle embolisation one to two days before surgery to reduce the risk of intra-operative bleeding and in some cases permit complete resection of the involved dura.
Cortical veins. Figure 4: Type IV DA VF - A 74-year-old man presented with a coma positioned at the level of the fistula completely obliterated the DA VF. Injection into the left external carotid artery shows several small radicles in cortical branches feeding into the transverse sinus. The veins sacrifice of a vessel. When occluding the cortical sinus it is important to ensure that any cortical venous reflux is abolished in order to minimise the risk of intracranial haemorrhage. This technique is commonly employed to treat fistulas of the cavernous sinus via a petrosal sinus. Occasionally, a sinus may no longer communicate with an internal jugular vein because of thrombosis. In these cases a direct percutaneous approach can be successful via a burr hole (Figure 2b) or orbital cut down procedure in the case of a carotico-cavernous fistula.

Radiosurgery has been used in the treatment of DAVFs. Söderman et al. treated 53 patients over a 25 year period with gamma knife radiosurgery. 36 patients had aggressive shunts exhibiting cortical venous drainage. 19 of these presented with haemorrhage. 41 patients were followed up by formal angiography and 28 DAVFs were obliterated. The risk of haemorrhage exists however until complete obliteration has occurred. Radiosurgical treatment should therefore be considered if occlusion by surgical or endovascular means is not possible or carries unacceptable risks.

Summary

DAVs can present in a variety of ways and their diagnosis can be missed on conventional cross-sectional imaging. Conventional catheter angiography remains the investigation of choice if the diagnosis is clinically suspected. A spectrum of pathology exists ranging from the benign to life-threatening. Treatment is indicated in more aggressive disease. This is characterised by cortical venous reflux on angiographic investigations. A multi-disciplinary approach is required before considering treatment, which can be surgical, endovascular or occasionally radiosurgical.

References