Endovascular Treatment of Cerebral Dural and Pial Arteriovenous Fistulas

Pascal Jabbour, MD*, Stavropoula Tjoumakaris, MD,
Nohra Chalouhi, MD, Ciro Randazzo, MD,
Luis Fernando Gonzalez, MD, Aaron Dumont, MD,
Robert Rosenwasser, MD

KEYWORDS

• Dural • Pial • Fistula • Endovascular • Arteriovenous • Embolization

KEY POINTS

- Dural arteriovenous fistulas (DAVFs) are arteriovenous shunts from a dural arterial supply to a dural venous channel, typically supplied by pachymeningeal arteries and located near a major venous sinus.
- DVAFs with retrograde venous drainage can result in hemorrhage or cause decreased regional cerebral blood flow in cortical regions involved.
- DAVFs can be treated with surgery, endovascular embolization (transarterial or transvenous approach), and radiosurgery.
- Elimination of the retrograde cortical venous drainage is the goal of any type of treatment for DAVFs.
- Pial Arteriovenous fistulas (PAVFs) are direct fistulas from an intracranial arterial feeder into a single venous channel and typically have high risk of intracranial hemorrhage and death if left untreated.
- PAVFs can be treated with surgery or endovascular embolization. Radiosurgery is not used because of the latent effect and difficulties in targeting the fistula.

DURAL ARTERIOVENOUS FISTULAS

Dural arteriovenous fistulas (DAVFs) are arteriovenous shunts from a dural arterial supply to a dural venous channel, typically supplied by pachymeningeal arteries and located near a major venous sinus.[1] The etiology of these lesions is not fully understood; some are congenital, and others are acquired. DAVFs in the pediatric population are associated with structural venous abnormalities,[2] but most DAVFs are thought to be acquired.[3,4] The development of venous obstruction and hypertension with aberrant angiogenesis[5] can contribute to the pathogenesis of these lesions. This altered angiogenesis occurs within the dura following an inciting event such as trauma, surgery, chronic infection, or sinus thrombosis. As microshunts proliferate in association with venous hypertension, these mature into clinically significant DAVF. The degree of progression or involution determines the significance of the abnormality. This can then result in hemorrhage or other focal manifestations including hemodynamic insufficiency. DAVFs can also cause decreased regional cerebral blood flow in cortical regions where there is retrograde venous drainage.

Department of Neurosurgery, Jefferson Hospital for Neuroscience, Thomas Jefferson University Hospital, 909 Walnut Street, Philadelphia, PA 19107, USA
* Corresponding author. 909 Walnut Street, 2nd Floor, Philadelphia, PA 19107.
E-mail address: Pascal.jabbour@jefferson.edu

Neuroimag Clin N Am 23 (2013) 625–636
http://dx.doi.org/10.1016/j.nic.2013.03.010
1052-5149/13/$ – see front matter

At the same time, some cases of DAVF have no clear inciting event or are at a site that is clearly distinct from the presumed inciting event. It is thought that the development of a DAVF in these settings requires a common unclear mechanism as well as a possible anatomic or genetic predisposition.[6,7]

DAVFs have been reported in all age groups, but mainly in the fifth and sixth decades of life.[8–10] The estimated incidence of DAVFs is 0.17 cases per 100,000 population, and they are one-fifth as common as arteriovenous malformations (AVMs).[11,12] They represent 10% to 15% of all intracranial vascular malformations,[11–17] with a higher incidence in women. A female-to-male ratio of 2:1 exists in certain anatomic sites such as the cavernous and transverse–sigmoid sinuses.[4,8,9,18–20] DAVFs are usually solitary, although in 5% of cases, multiple lesions have been described.[12,21,22] The goal of this article is to describe the natural history, clinical presentation, and treatment of dural and pial fistulas with emphasis on the endovascular treatment.

Natural History

An established DAVF may follow 1 of several unpredictable natural courses. Some lesions remain asymptomatic or maintain stable clinical symptomatology and angiographic features over many years. Others undergo spontaneous regression, involution, and resolution with stabilization or improvement of neurologic symptoms.[23–28] Features that may predispose to such spontaneous involution are not known. DAVFs in the region of the cavernous sinus are particularly prone to this phenomenon, with as many as 40% of reported cases having undergone spontaneous involution.

In contrast, some DAVFs may demonstrate an increase in size from either arterial or venous enlargement[1,11,29] or even de novo development of a DAVF.[30] Pachymeningeal arterial feeders may be progressively recruited causing enlargement of the nidus due to unknown mechanisms. This results in hypertrophy of dural arteries and the reappearance of involuted embryonic arteries that may not normally be visible in the adult dura mater. In some DAVFs there is also progression on the venous side. Progressive arterialization of the pathologic dural leaflets results in hypertension in adjacent leptomeningeal venous channels, leading to retrograde leptomeningeal venous drainage. Under arterialized pressures these channels may become tortuous and become varices or aneurysms. The catastrophic consequence that ensues is a cerebral hemorrhage from retrograde cortical venous drainage (CVD).

Clinical Presentation and Assessment

Clinical manifestations of DAVFs are highly variable and are related primarily to the location of the fistula as well as retrograde CVD. These range from minor symptoms to intracranial hemorrhage. The vast majority of symptoms can be attributed to the anatomy of the DAVF.

Patients' symptoms may be sudden or slowly progressive. The degree and type are determined by venous topography, venous flow pattern, and the capacity of surrounding compensatory venous drainage. The most serious neurologic sequelae from DAVFs are associated with retrograde CVD,[11] leading to a propensity to rupture. Focal neurologic deficits likely result from venous hypertension and intracranial hemorrhage from rupture of arterialized leptomeningeal veins.

There are a wide variety of nonhemorrhagic symptoms with which DAVF can present.[31,32] Relatively benign symptoms such as pain, tinnitus, or bruit are related to arteriovenous shunting and flow within the DAVF. Pulsatile tinnitus or other auditory symptoms may occur with or without pain. These symptoms are likely related to high flow through dural vascular channels at the base of the skull. Other more painful complaints may be related to orbital congestion, stretching of dural leaflets by engorged vascular channels, or to direct compression of the trigeminal nerve by arterialized venous structures near the petrous apex.

Various neuro-ophthalmologic manifestations of DAVFs include visual and gaze abnormalities caused by venous hypertension. Orbital or ocular venous hypertension with resulting orbital crowding, venous stasis retinopathy, and glaucoma can also be seen.

Other intracranial DAVFs may present with symptoms of increased intracranial pressure (ICP) or a poorly defined headache.[33,34] While the headaches are nonspecific, there does appear to be an association with the dysplastic changes in meningeal vessels that are often present in DAVFs. There are also a wide spectrum of neurologic symptoms including seizure, hearing loss, cranial nerve palsy, papilledema, vision changes, and motor/sensory deficits that can be seen with intracranial DAVF.

DAVFs may also result in altered cerebrospinal (CSF) flow.[35] Dilated venous structures may act as mass lesions, obstructing the CSF circulation and causing hydrocephalus. In other cases, dural venous hypertension may result in decreased absorption of CSF with secondary intracranial hypertension and papilledema. This latter complication appears to be more common in high-flow lesions draining into large dural venous sinuses in the setting of concomitant sinus outflow obstruction.