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Background

AVF are characterized by abnormal shunting of blood between the arterial system and the venous system, without the presence of a normal intervening capillary bed. Because the capillary bed represents the source of resistance to blood flow in the circulatory system, AVF are low resistance, high flow lesions. They represent a group of disorders that are characterized by abnormal shunting from the ECA, ICA, vertebrobasilar system, or spinal arteries into normal or abnormal venous channels. The primary treatment is often endovascular because of the feasibility of catheterizing the lesion directly and placing embolic material directly at the site of the abnormal arteriovenous connection. For purposes of this practice guideline, AVMs will be considered as a separate entity and are discussed in a separate guideline. Additionally, spinal lesions and lesions contained entirely in the extracranial space will be separately addressed, although many principles of the treatment of fistulae with cerebral components also apply to the treatment of all fistulae.

Carotid Cavernous Fistulae

Fistulae occurring between major cerebral arteries and/or their branches, which drain directly or indirectly into the cavernous sinus (CS), are collectively called carotid cavernous fistulae (CCF). CCF are the most frequent and among the most complex examples of AVF of the CNS. This discussion of their treatment therefore also serves as a model for the treatment of other fistulae (1).

CCF arising from a defect in the cavernous segment of the ICA are called direct fistulae because the drainage into the CS arises directly via the ICA. CCF resulting from AVF supplied by ECA and ICA dural branches are traditionally called *indirect*, or *dural*, fistulae (dural AVF of the cavernous sinus).

Under normal circumstances, the CS contributes to cerebral venous drainage by receiving blood flow from the sphenoparietal sinus and from the orbit by way of the ophthalmic veins. Thus, venous drainage from the globe is directed toward the CS by way of the ophthalmic veins. Maintenance of normal intraocular pressure is a function of maintenance of normal venous pressure in the orbit, among other factors. In the presence of an arteriovenous shunt in the CS, flow becomes reversed in the orbital veins and is then directed in a sinofugal fashion toward communicating channels, including anteriorly toward the superior ophthalmic vein. Venous congestion occurs in the territory drained by the ophthalmic veins. This leads to many of the classic symptoms of CCF (2, 3).

The diagnosis of CCF can usually be made clinically. In the absence of a history of trauma, patients usually present because they have orbital complaints related to increased venous pressure. Imaging studies with either CT or MR imaging may confirm the diagnosis by the presence of distended ophthalmic veins (6). The lack of dilation of the superior ophthalmic vein or other imaging features does not exclude the diagnosis and should not preclude angiography. The diagnosis is confirmed by angiography, which should be performed with consideration of the options for the ultimate treatment of the disease. Diagnostic arteriography is best performed by those who are charged with the ultimate treatment of the disease.

Indications

Patients with CCF present with a combination of symptoms and signs of venous hypertension and congestion, including proptosis, extraocular muscle enlargement and edema in the orbital fat, chemosis, scleral injection, and conjunctival hemorrhages. Diminished visual acuity can result from ocular ischemia due to reduced perfusion pressure, papilledema, retinal detachment, or the production of glaucoma due to decreased drainage of vitreous into the pressurized venous system. Visual decline can be fulminant, necessitating emergent therapy. Thus, close ophthalmologic monitoring is an important component in management. Pharmacologic management of glaucoma requires close monitoring of intraocular pressure by the ophthalmologic team members.

Headaches may be related to stretching of dural structures in the CS, venous thrombosis, venous hemorrhage, or pathologic involvement of the Vth cranial nerve, either by way of direct pressure in the CS or pulsatile pressure from abnormal arterial flow into the CS. Another common complaint, diplopia, may be secondary either to direct venous compression or pulsatile trauma on the IIIrd, IVth, or VIth cranial nerves in the CS or may be due to ischemic injury from decreased venous drainage of the nerves from the elevated venous pressure (4).

Halbach et al (5) reported pulse synchronous bruit in approximately 50% of patients with dural fistulae involving the CS. The bruit can often be heard on examination with a stethoscope, either over the involved eye or elsewhere over the cranium. Observation of a bruit is an important component in management, because such surveillance represents a noninvasive means for repeated as-

sessment of closure or recurrence of a fistula during treatment. Its presence or absence should be recorded at the time of the initial examination.

Although cerebral symptoms are unusual in cases of CCF, they are a particular risk in association with high flow fistulae. They may occur secondary to increased pressure in cerebral cortical veins, resulting in cerebral edema or intracerebral hemorrhage. They may also be secondary to cerebral ischemia related to arterial steal into the fistula, resulting in deprivation of flow to the brain with reduced cerebral perfusion. Subdural or subarachnoid hemorrhage may also occur when the arterialized blood drains pathologically into dural or cortical veins.

Efficacy

Treatment of CCF is directed toward relief of symptoms. In the case of most direct CCF, this also involves elimination of the fistula. In the case of indirect CCF, although elimination of the fistula is the goal, cure is sometimes difficult to achieve. In those cases, symptomatic relief can sometimes be achieved by elimination of the venous outflow to the orbit. Intermittent carotid compression may be an appropriate first treatment maneuver for many patients.

A secondary goal in each case is preservation of the carotid artery flow to the brain. For some patients, this is a necessity insofar as carotid sacrifice would not be tolerated. In cases of direct CCF, treatment is therefore directed to the venous side of the fistula. Historically, the most successful and well-established treatment modality involves the placement of a detachable balloon across the CCF, using flow direction from the arterial side. The balloon is then inflated such that it is wedged in the CS against the fistula. The balloon thereby creates a tamponade of the CCF, eliminating flow and permitting healing across the orifice of the fistula. In particularly large CCF or in fistulae in which the CS is distended, multiple balloons are sometimes needed. The goal is to position the balloons such that the last one placed is wedged against the arterial rent, causing cessation of flow across it. However, even in the case of direct CCF, it is not always possible to get a balloon to cross the arterial defect to the venous side.

It is possible to cure 90% of direct CCF using intra-arterial therapy and detachable balloons (7, 8). However, if one is restricted to the use of intra-arterial therapy and detachable balloons, carotid preservation is not possible in all situations. Use of a transvenous approach (9) or a surgical approach to the CS (10) increases the rate of cure of CCF without decreasing the rate of carotid preservation. However, detachable balloons can present substantial technical difficulties when these approaches are used. Direct CCF can also be treated with coils, by both transarterial and transvenous approaches. Balloon-assisted techniques for coil placement may be required.

When carotid sacrifice is contemplated or anticipated as a possible complication of a procedure, it is often useful to assess the patient's potential for neurologic tolerance of that maneuver. Balloon occlusion testing is complicated by the hemodynamics of the fistula, which can produce cerebral steal via retrograde flow into the fistula when only proximal occlusion is achieved. Temporary occlusion testing is sometimes performed using a nondetachable balloon placed at or distal to the fistula. Although this may increase flow into the venous side of the fistula by temporarily cutting off distal carotid outflow, it should not affect the cerebral hemodynamics. If necessary, a second balloon can also be placed proximally, temporarily trapping the fistula while neurologic testing is performed.

Indirect CCF can be treated from an arterial, venous, or surgical approach. The surgical approach is again reserved for cases in which the arterial and venous approaches either are unavailable or fail. Achievement of cure is often much more difficult with indirect than with direct CCF. Transarterial embolization can sometimes produce improvement in symptoms. However, as with other AVF, partial embolization often provides only temporary relief. With indirect CCF, it is rarely possible to cross an arterial fistula to the venous side from an arterial approach.

In cases of indirect CCF, the easiest approach for cure is by way of the inferior petrosal sinus to the CS. Arterial in-flow to the fistula is disconnected from the superior ophthalmic vein by packing the CS between the fistula and the superior ophthalmic vein. If the ipsilateral inferior petrosal sinus is occluded, it is sometimes possible to access the CS via the contralateral inferior petrosal sinus and basilar plexus or the circular sinus, through the facial, angular, and superior ophthalmic veins, or through the pterygoid plexus.

Other routes of transvenous access in cases of CCF might include percutaneous catheterization of the angular vein on the scalp or retrograde catheterization of the facial vein, gaining access to the superior orbital vein and then the CS. Direct placement of a cannula in the orbital vein may be performed under surgical exposure (11, 12). More than one attempt or method of approach might be required to achieve success.

Most embolizations of CCF can be performed with the patient awake but sedated. This allows better monitoring of the patient's vision or neurologic condition. In some cases, such as acute occlusion of high flow fistulae in which normal perfusion pressure breakthrough may be of concern, performance of the procedure with the patient awake may be specifically necessitated for monitoring purposes. When prolonged procedures are expected or in cases in which patient cooperation is questionable because of age, mental status, or anxiety, general anesthesia may assist considerably in the successful and efficient performance of a procedure. Efficacy thresholds are outlined in the accompanying table.

Efficacy Indicator	Threshold (%)
Indirect CCF	
Complete or significant relief (may require multiple stages)	90
Direct CCF	
Complete or significant relief	90
CCF occlusion with or without parent vessel occlusion	90

Safety

During treatment, complications may be encountered involving either the eye or the cerebral circulation. As mentioned earlier, use of particulate or liquid embolic material can cause acute venous outflow obstruction, resulting in worsening of the elevation of intraocular pressure.

Neurologic decline during balloon embolization of a high flow fistula has been reported, with relief afforded with partial balloon deflation (13). This was attributed to normal perfusion pressure breakthrough after reestablishment of intracranial arterial pressurization when the fistula was occluded. This can be successfully treated with graded occlusion.

As is the case with any transarterial procedure, embolic complications that can result in stroke are possible. Routine use of heparin during these procedures limits the incidence of that complication. The use of smaller introducer catheters may also limit the incidence of intimal injury from the introducer catheter.

Special mention should be made of the hazards inherent in the treatment of patients with Ehlers-Danlos syndrome. The arteries are extremely fragile, arterial dissection and rupture may occur, and venous packing may be difficult to achieve. Safety thresholds are outlined in the accompanying table.

Safety Indicator	Threshold (%)
Indirect CCF	
Death	0
Major complication (permanent deficit)	1
Minor complication (permanent deficit)	2
Transient (> 24 hr) deficit	5
Direct CCF	
Death	1
Major complication (permanent deficit)	3
Minor complication (permanent deficit)	5
Transient deficit	5

Applications of These Principles to AVF Occurring Elsewhere

The clinical constellation of signs and symptoms that occurs with other AVF depends on the neuroanatomic structures involved, but most commonly relate to sequelae of venous hypertension also. Some common problems associated with AVF of the CNS include cerebral hemorrhage (either from associated aneurysms or from venous rupture related to venous hypertension), stroke (either from embolic phenomena or from cerebral steal by the fistula), transient ischemic symptoms related to cerebral steal, progressive deficit from increasing mass effect from the lesion or adjacent cerebral edema, and progressive deficit from cerebral steal. High flow fistulae can also be associated with increased intracranial pressure, especially when associated with venous outflow restriction. More global symptoms, such as headache, memory loss, and decreasing level of consciousness, can also be seen and may be a harbinger of either progression of the primary disease or of the development of complications. However, the endovascular physician must be sufficiently familiar with the clinical neurologic examination and with the broad spectrum of clinical signs and symptoms associated with these lesions, so that he is able to integrate the findings with the therapeutic plan. This is necessary for follow-up purposes as well.

Quality Improvement

All fistula embolizations should be monitored within the overall quality improvement program of the facility. Incidence of complications and unexpected admissions should be recorded and periodically reviewed for the opportunity to improve care. These data should be collected in a manner that complies with statutory and regulatory peer review procedures to protect the confidentiality of the peer review data.

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