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Diagnosis and management of dural carotid–cavernous sinus fistulas

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✓A carotid–cavernous sinus fistula (CCF) is an abnormal communication between the cavernous sinus and the carotid arterial system. Some fistulas are characterized by a direct connection between the cavernous segment of the internal carotid artery (ICA) and the cavernous sinus, whereas others consist of a communication between the cavernous sinus and one or more meningeal branches of the ICA, external carotid artery, or both. These dural fistulas usually have low rates of arterial blood flow and until recently were difficult to diagnose and treat. In this paper, the author discusses the anatomy, pathogenesis, clinical manifestations, diagnosis, treatment, and prognosis of dural CCFs.
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KEY WORDS • carotid–cavernous sinus fistula • internal carotid artery • external carotid artery • superior ophthalmic vein

THE blood supply to the region of the cavernous sinus is provided by interconnecting branches of the ICA and ECA, and it is from these vessels that dural CCFs—often called dural arteriovenous fistulas—arise. Such fistulas usually are separated anatomically into three types: 1) shunts between meningeal branches of the ICA and the cavernous sinus; 2) shunts between meningeal branches of the ECA and the cavernous sinus; and 3) shunts between meningeal branches of both the ICA and ECA and the cavernous sinus (Figs. 1–3).[7,20] Of these three types, the third type is by far the most common.

Pathogenesis

Dural CCFs usually become symptomatic spontaneously. The pathogenesis of these fistulas is controversial.[21] It was once speculated that spontaneous dural CCFs form after rupture of one or more of the thin-walled dural arteries that normally traverse the cavernous sinus.[79] Extensive, preformed, dural arterial anastomoses not directly involved in the fistula may dilate and contribute collateral blood supply after rupture, resulting in an angiographic appearance indistinguishable from that of a congenital vascular malformation. Sequential arteriography demonstrates that the feeder vessels of dural CCFs change with time as the vessels spontaneously open and close.[100] Although this theory is favored by some investigators,[7] it fails to explain why spontaneous dural CCFs are more common in elderly women than in men. A second theory for the origin of dural CCFs is that most of these lesions develop in response to spontaneous venous thrombosis in the cavernous sinus and represent an attempt to provide a pathway for collateral venous outflow.[40] This theory is favored by most investigators because it also explains the pathogenesis of arteriovenous fistulas that develop in the sigmoid and other dural sinuses.[21]

Certain factors may predispose patients to the development of symptomatic dural CCFs. These factors include pregnancy, systemic hypertension, atherosclerotic vascular disease, connective tissue disease (such as Ehlers–Danlos syndrome), and minor trauma (Fig. 4).[75,91,102] In addition, iatrogenic dural CCFs occasionally occur.[14]

Clinical Manifestations

Dural CCFs usually occur in middle-aged or elderly women, but they may produce symptoms in either sex at

Abbreviations used in this paper: CCF = carotid–cavernous fistula; CN = cranial nerve; CT = computed tomography; ECA = external carotid artery; ICA = internal carotid artery; MR = magnetic resonance; SOV = superior ophthalmic vein.

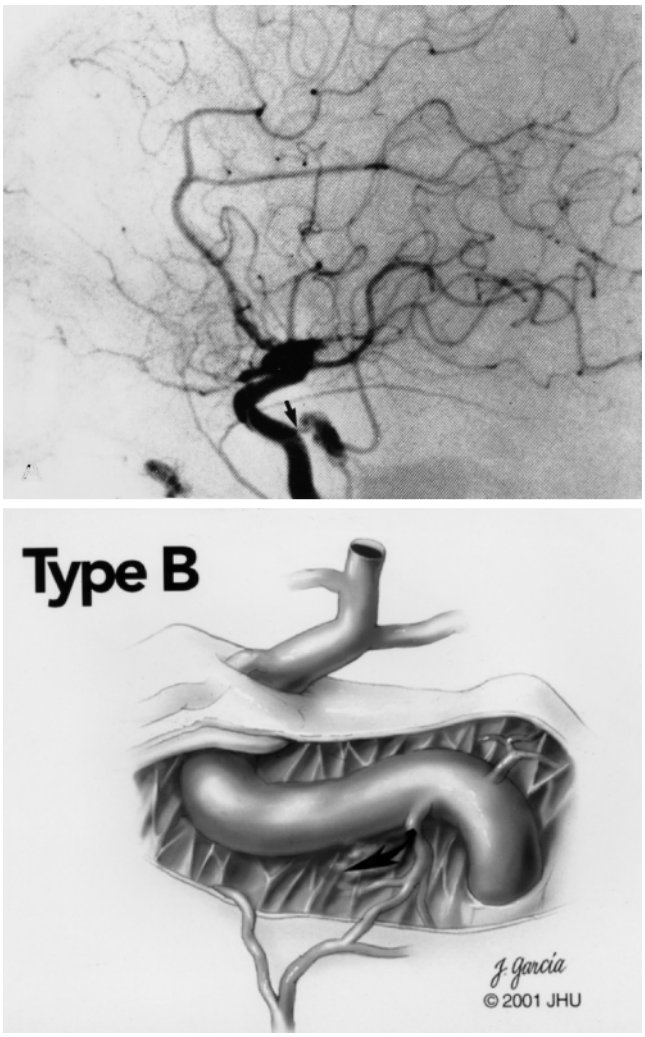

FIG. 1. Images showing the appearance of a dural CCF in which the only contribution is from extradural branches of the ICA. *Upper:* A selective left ICA arteriogram shows a fistula at the posterior portion of the cavernous carotid artery *(arrow)*. The left ECA arteriogram was normal. *Lower:* Artist's drawing shows that this type of fistula is fed only by extradural branches of the ICA with no contribution from the extradural branches of the ipsilateral ECA (Type B according to Barrow and colleagues, 1985).

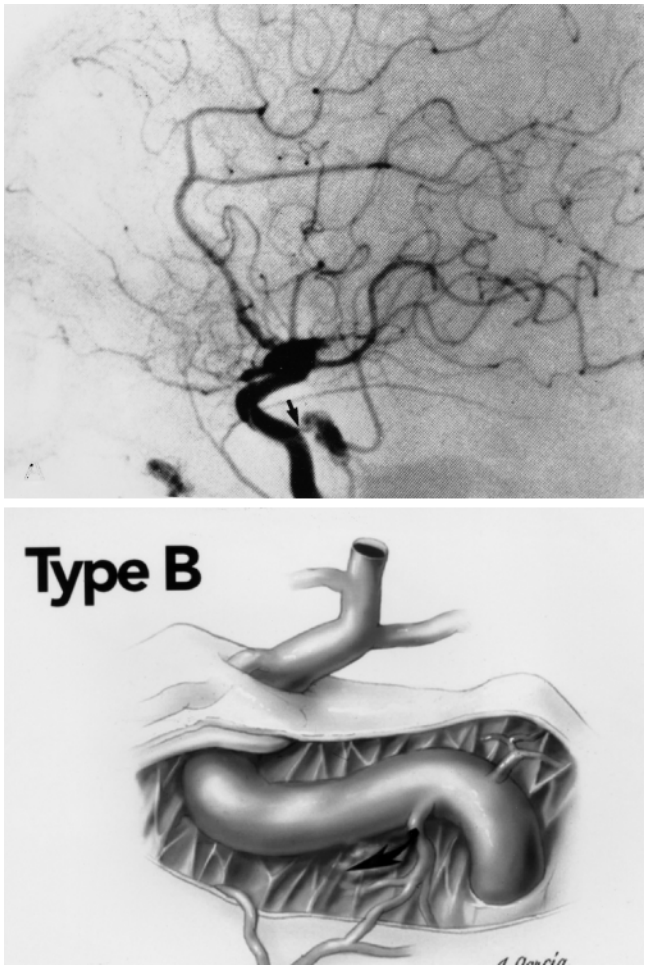

FIG. 2. Images showing the appearance of a dural CCF in which the only contribution is from extradural branches of the ECA. *Upper:* A selective left external carotid arteriogram shows a high-flow fistula fed by extradural branches of the left ECA, particularly the internal maxillary artery. The fistula drains anteriorly into the ipsilateral SOV *(arrows)*. There was no contribution from the ipsilateral ICA or from the contralateral ICA or ECA. The patient was a 65-year-old woman with gradual onset of redness and swelling of the left eye. *Lower:* Artist's drawing of the appearance of this type of fistula. Note that the only contribution is from the extradural branches of the ECA (Type C according to Barrow and colleagues, 1985).

any age, even in childhood or infancy.[95] The symptoms and signs produced by these lesions are influenced by a number of factors, including the size of the fistula, the location within the cavernous sinus, the rate of blood flow, and drainage route, especially if the drainage route is posterior, anterior, or both.[98,99] The drainage route of the fistula is probably related to its basic anatomical configuration, although Grove[30] postulated that many, if not all, fistulas initially drain posteriorly into the inferior petrosal sinus, basilar venous plexus, or both. Grove believed that when this normal pathway for drainage becomes thrombosed, the fistula begins to drain anteriorly, producing visual symptoms and signs. I and others have examined patients whose clinical course suggests that this theory is correct.[38] Such patients initially may experience an acute, isolated, ocular motor nerve paresis, the evaluation of which reveals a posteriorly draining fistula (Fig. 5, *upper*). Shortly thereafter, these patients develop typical signs of an anteriorly draining fistula (Fig. 5, *lower*).

Posteriorly Draining Fistulas

When dural CCFs drain posteriorly into the superior and inferior petrosal sinuses, they are usually asymptomatic. In some cases, however, such fistulas produce a cranial neuropathy, such as a trigeminal neuropathy,[87] facial nerve paresis,[77] or an ocular motor nerve paresis.[24] In most of these cases, there is no evidence of orbital congestion.

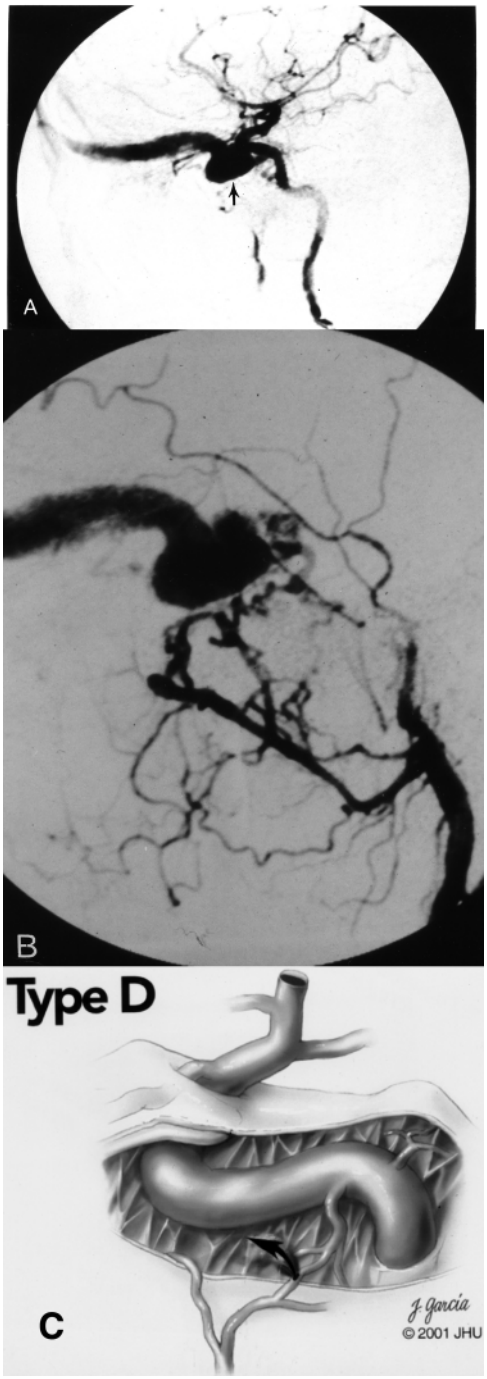

FIG. 3. Images showing the appearance of a dural CCF that is fed by extradural branches from both the ICA and ECA. A: Lateral view of a selective left internal carotid arteriogram shows a large collection of contrast material in the cavernous sinus *(arrow).* The fistula drains anteriorly into the left SOV, which is markedly enlarged. B: Lateral view of a selective left external carotid arteriogram shows multiple contributions from extradural branches of the left ECA. C: Artist's drawing of this type of fistula, which is fed by extradural branches of both the ICA and ECA (Type D according to Barrow and colleagues, 1985).

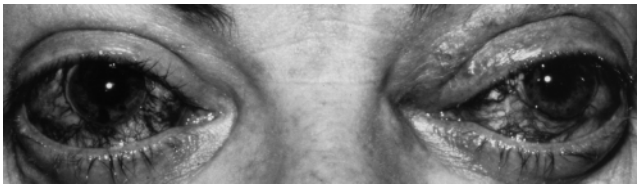

FIG. 4. Photograph of a 39-year-old woman with Ehlers–Danlos syndrome who developed spontaneous bilateral dural CCFs. The CCFs were successfully closed using an endovascular approach, but the patient died several months later from unrelated vascular complications of the underlying disease.

In most cases of ocular motor nerve paresis caused by a posteriorly draining dural CCF, the onset of the paresis is sudden, and only one of the ocular motor nerves is affected. The oculomotor nerve (CN III) is most often affected, and the resulting paresis may be complete with involvement of the pupil, incomplete with pupil involvement, or incomplete with pupil sparing. I have never seen a patient with a complete, pupil-sparing oculomotor nerve paresis in this setting. In almost all cases, the paresis is associated with ipsilateral orbital or ocular pain, a presentation that initially suggests an intracranial aneurysm (Fig. 6).[38,65] The correct diagnosis in such cases is not evident until cerebral angiography is performed. In other cases, the posteriorly draining fistula produces an abducent (CN VI) or trochlear (CN IV) nerve paresis, again usually associated with ocular or orbital pain (Fig. 7).[24,52,79,92]

The cranial neuropathies that are caused by a posteriorly draining dural CCF are usually the initial sign of the fistula. In many of these cases, failure to diagnose and treat the fistula leads eventually to a change in the direction of the flow of blood in the fistula. The flow becomes anterior, and the patient develops evidence of orbital congestion. In other cases, the blood flow in the fistula initially is anterior, producing orbital manifestations. With time, however, the anterior drainage ceases, and posterior flow is associated with the development of the cranial neuropathy.

Dural fistulas that drain posteriorly sometimes cause brainstem congestion that may be associated with neurological deficits.[101] In addition, such fistulas rarely may produce intracranial hemorrhage.[35]

Anteriorly Draining Fistulas

Similarly to their direct counterparts, dural CCFs usually produce visual symptoms and signs when they drain anteriorly into the superior and inferior ophthalmic veins. The clinical manifestations of patients with dural CCFs that drain anteriorly are therefore similar to, but usually much less severe than, those of patients with direct fistulas, because most dural fistulas contain blood flowing at a low rate. Dural fistulas usually produce an important and rather characteristic syndrome that, nevertheless, often is misdiagnosed.[12,30,47,48,60,79,84,98,99] Unlike direct fistulas, there often is no objective or subjective bruit with dural fistulas; even when a subjective bruit is present, the patient may not mention it, either because it is mild or because the patient does not associate the sound with his or her ocular symptoms and signs. In the mildest cases, there is redness of one, or rarely, both eyes, caused by dilation and arterialization of

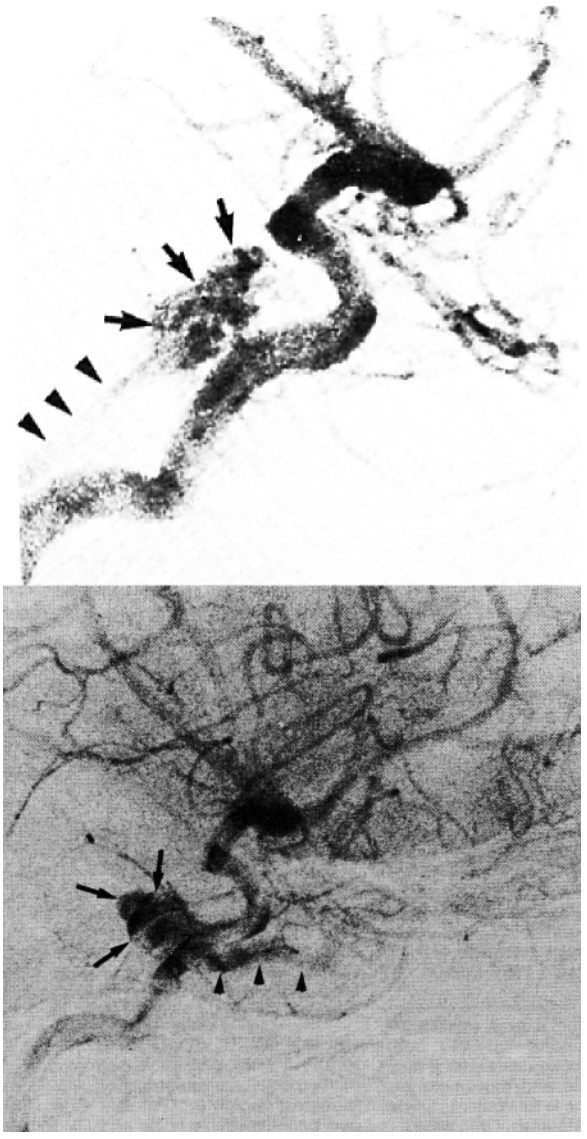

FIG. 5. Lateral views of a selective right ICA arteriogram in a patient with a dural CCF and an acute, painful CN III. *Upper:* Initial view shows a posteriorly draining dural CCF (shunt). The patient was a 58-year-old woman who developed an acute right-sided frontoorbital headache. Four weeks later she developed diplopia, and 7 days afterward developed right ptosis and a dilated right pupil. She was believed to have an intracranial aneurysm, and an arteriogram was obtained. The arteriogram shows a dural arteriovenous fistula of the cavernous sinus *(arrows)* that drains posteriorly into the inferior petrosal sinus *(arrowheads)*. *Lower:* Repeated cerebral angiography was performed 5 weeks later, after the patient developed redness, conjunctival chemosis, and proptosis of the right eye. Arteriogram shows that the dural CCF *(arrows)* now drains anteriorly into the SOV *(arrowheads)* rather than posteriorly. From Hawke SH, Mullie MA, Hoyt WF, et al: Painful oculomoter nerve palsy due to dural-cavernous sinus shunt. **Arch Neurol 46:**1252–1255, 1989. Reproduced with permission of the *Journal of the American Medical Association.*

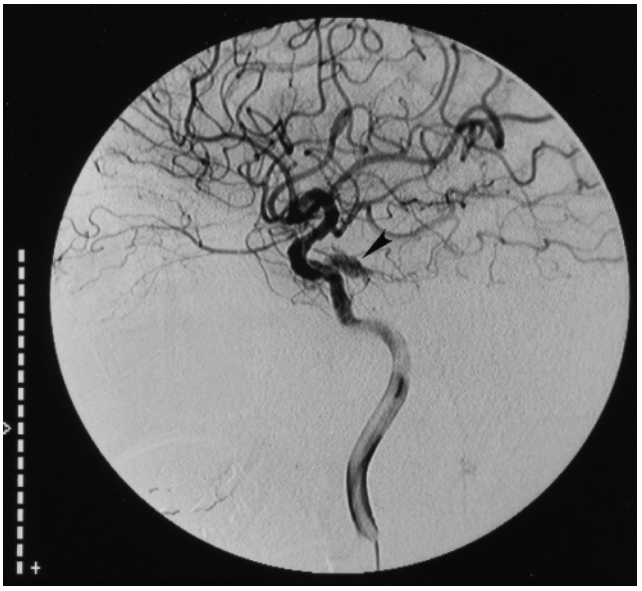

FIG. 6. Lateral view of a selective left internal carotid arteriogram showing a large dural CCF *(arrowhead)* that drains posteriorly into the inferior petrosal sinus rather than anteriorly into the orbit. The patient was an 86-year-old woman with facial pain and acute binocular diplopia. The examination revealed a partial left CN III palsy with involvement of the pupil. The patient had no signs or symptoms of orbital disease. From Lee AG: Third nerve palsy due to a carotid cavernous fistula without external eye signs. **Neuro-Ophthalmology 16:**183–187, 1996. Reproduced with permission of the publisher.

both conjunctival and episcleral veins (Fig. 8). The appearance of the eye in these cases may suggest conjunctivitis, episcleritis, or thyroid eye disease; however, a careful examination of the dilated vessels usually demonstrates a typical tortuous corkscrew appearance that is virtually pathognomonic of a dural CCF (Fig. 9). There also may be minimal eyelid swelling, conjunctival chemosis, proptosis, or a combination of these symptoms. Diplopia from abducent nerve paresis may be present (Fig. 10). Ophthalmoscopy results may be normal, or there may be mild dilation of retinal veins.

In more advanced dural CCFs, particularly those with a high flow rate, the symptoms and signs are identical to those in patients with direct CCFs.[41,71,98,99] In these cases, proptosis, chemosis, and dilation of conjunctival vessels are obvious (Fig. 11). Diplopia may result from ophthalmoparesis caused by ocular motor nerve pareses, orbital congestion, or both mechanisms, and there may be significant periorbital or retroocular discomfort or pain, initially suggesting an inflammatory process or even the Tolosa–Hunt syndrome.[15,85] Some patients develop facial pain, facial weakness, or both.[43] Increased episcleral venous pressure may produce increased intraocular pressure that occasionally is quite high.[41,47,84,108] Angle–closure glaucoma may develop from elevated orbital venous pressure, congestion of the iris and choroid, and forward displacement of the iris–lens diaphragm.[103] In other cases, chronic ischemia produces neovascular glaucoma. Ophthalmoscopic abnormalities include venous stasis retinopathy with intraretinal hemorrhages, central retinal vein occlusion, proliferative retinopathy, retinal detachment, vitreous hemorrhage, choroidal folds, choroidal effusion, choroidal detachment, or optic disc swelling (Fig. 12).[25,29,55,61,76,90]

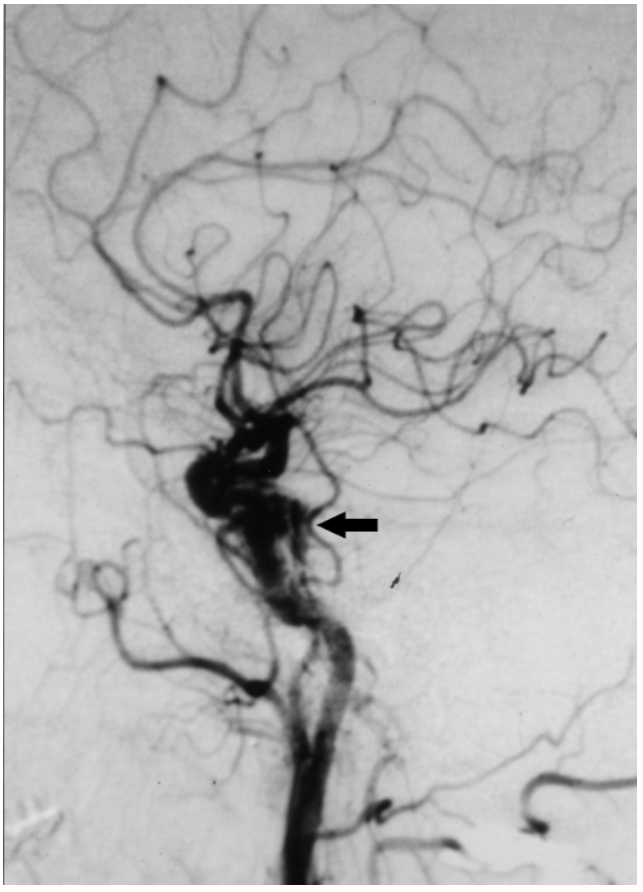

FIG. 7. Lateral view of a left common carotid arteriogram showing a dural CCF *(arrow)* draining posteriorly into the inferior petrosal sinus. The patient was a 44-year-old woman with a severe headache and horizontal diplopia. The examination revealed a left CN VI palsy. Courtesy of Eric Eggenberger, D.O.

Visual loss, although less frequent than in patients with direct CCFs, occurs in 20–30% of patients with dural CCFs.[30,79] This visual loss may be caused by ischemic optic neuropathy, chorioretinal dysfunction, or uncontrolled glaucoma.[61,84]

The ocular manifestations of unilateral dural CCFs almost always are ipsilateral to the fistula, but they may be solely contralateral or bilateral (Fig. 13).[98,99] When unilateral fistulas cause bilateral manifestations, there is a high probability that the fistula is draining into cortical veins (Fig. 14).[98]

Although most dural fistulas are unilateral, bilateral spontaneous dural fistulas have been described.[37] Patients with bilateral dural CCFs often have severe systemic hypertension, atherosclerosis, or some type of systemic connective tissue disease such as Ehlers–Danlos syndrome Type IV. Most patients with bilateral dural CCFs have bilateral manifestations; however, I once examined a patient with bilateral dural fistulas who demonstrated only left-sided ocular manifestations: the left-sided fistula drained anteriorly into the left orbit via the left SOV, and the right-sided fistula drained across the intercavernous sinus and then anteriorly into the left orbit.

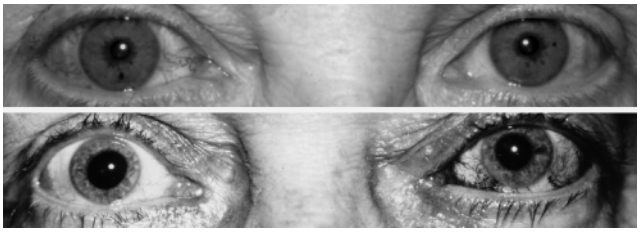

FIG. 8. Photographs of two different patients with spontaneous dural CCFs. *Upper:* A 58-year-old woman with mild monocular redness of the right eye with minimal fullness of the eyelids. This appearance is often mistaken for conjunctivitis. *Lower:* A 61-year-old man with moderate redness of the left eye associated with mild proptosis. This appearance is often mistaken for episcleritis or dysthyroid orbitopathy.

In some instances, dural CCFs drain both anteriorly and posteriorly. In most of these cases, the only manifestations are those related to the anterior drainage; however, some patients develop manifestations from the posterior drainage, such as facial nerve paresis or acute hemiparesis associated with neuroimaging evidence of brainstem congestion.[77,94]

Differential Diagnosis

Because the symptoms and signs of a dural CCF often are mild, usually developing spontaneously and rather slowly, this lesion is often misdiagnosed initially. When the patient simply has a red eye, perhaps with minimal eyelid swelling, it may be believed that he or she has a chronic conjunctivitis or blepharoconjunctivitis that is refractory to topical therapy (Fig. 15). In patients who develop diplopia from abducent nerve paresis, the significance of a slightly red eye may be missed (Fig. 16).

In patients with evidence of orbital congestion, red eye, conjunctival chemosis, and other symptoms, diagnoses other than a spontaneous dural CCF, such as dysthyroid orbitopathy, orbital pseudotumor, orbital cellulitis, episcleritis, sphenoorbital meningioma, or Tolosa–Hunt syndrome, may be considered.[15,30,79,81,84,85] The correct diagnosis in such cases may not be able to be made until symptoms and signs worsen, new symptoms and signs develop, or appropriate diagnostic studies are performed. In addition, trauma to the posterior orbit in the region of the superior orbital fissure may produce such manifestations,[70] and I and others have examined patients with congenital or acquired

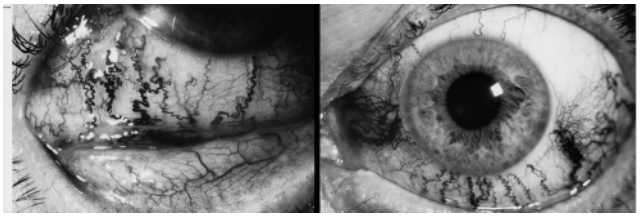

FIG. 9. Photographs of conjunctival and episcleral vessels in two patients with spontaneous dural CCFs. Note dilation, tortuosity, and corkscrew appearance of the veins in each eye.

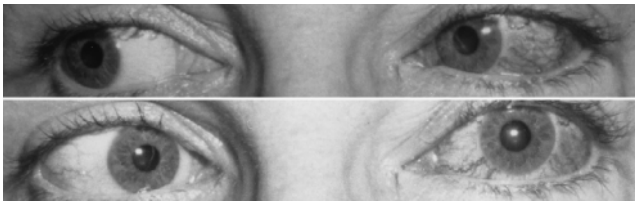

FIG. 10. Photographs of left CN VI palsy in a 34-year-old woman with a left-sided dural CCF. *Upper:* The left eye adducts fully during an attempted right lateral gaze. *Lower:* When the patient attempts to look to the left, the left eye abducts only to just beyond the midline. Note the mild left proptosis and the dilated conjunctival veins of the left eye.

anomalous intracranial venous drainage who developed clinical manifestations suggesting a dural CCF.[105]

Diagnosis

It should be clear from the previous discussion that the diagnosis of a dural CCF should be considered in any patient who spontaneously develops a red eye, chemosis of the conjunctiva, abducent nerve paresis, or mild orbital congestion with proptosis. Auscultation of the orbit may disclose a bruit, but this is relatively uncommon. Tonometry, however, usually shows asymmetry of the ocular pulse with greater pulse amplitude on the side of the lesion. The asymmetry in the amplitude of the ocular pulse can be discovered using any tonometer, although I prefer to use a pneumotonometer that provides both a direct measurement and an objective record of the ocular pulse amplitude (Fig. 17).[28]

When a dural CCF is suspected, CT scanning, CT angiography, MR imaging, MR angiography, orbital ultrasonography, transorbital and transcranial color Doppler imaging, or a combination of these tests may be beneficial in confirming the diagnosis (Figs. 18–21).[5,17,46,50,88] The gold standard diagnostic test, however, as in the case of the direct CCF, is a catheter angiogram (Figs. 1–3).[19,21] Because many dural CCFs are fed either by meningeal branches of the ECA or by meningeal branches of both the ICA and ECA and others are fed by arteries from both sides or are fed by unilateral arteries but produce bilateral symptoms and signs, selective angiography of both the ICA and ECA on both sides should always be performed.[19] When performed by an experienced neuroradiologist, catheter angiography has a morbidity rate of less than 1% and virtually no mortality rate, except in patients with connective tissue disorders such as Ehlers–Danlos syndrome, in whom the risks are much greater (because of excessive fragility of the extracranial and intracranial vessels).[91]

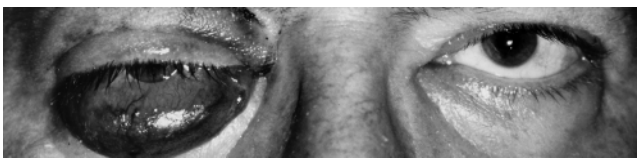

FIG. 11. Photograph of a right-sided, high-flow, dural CCF in a 54-year-old man. The right eye is moderately proptotic, and there is significant chemosis of the conjunctiva. The appearance of this patient is indistinguishable from that of a patient with a high-flow, direct CCF.

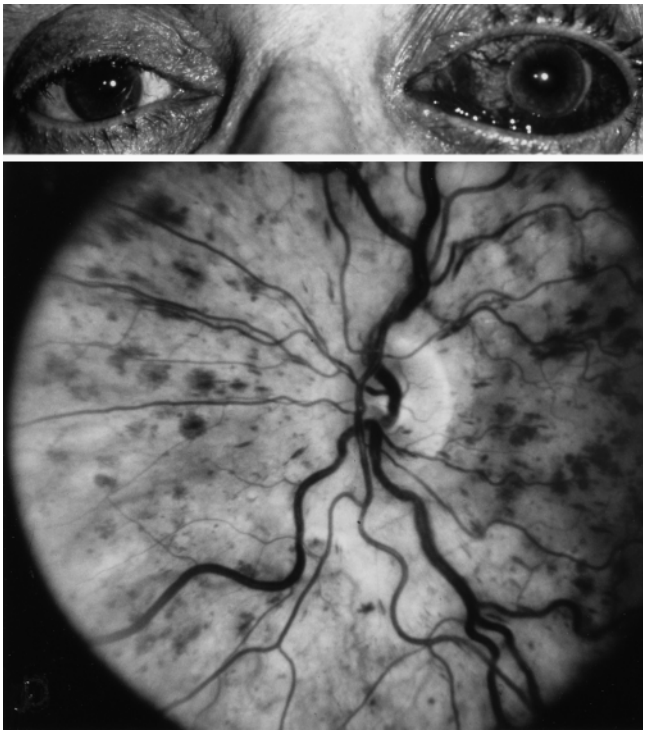

FIG. 12. Images of central retinal vein occlusion in a 62-year-old woman with a spontaneous dural CCF. *Upper:* Photograph shows moderate proptosis of the left eye, associated with conjunctival chemosis and arterialization of conjunctival and episcleral vessels. The patient noted progressive visual loss in the left eye over several days. *Lower:* Ophthalmoscopic photograph of left ocular fundus showing changes consistent with a mild central retinal vein occlusion. The left optic disc is normal, but the retinal veins are dilated, and there are numerous intraretinal "dot and blot" hemorrhages. The visual acuity in this eye was 20/80, but improved to 20/20 as the hemorrhages cleared spontaneously.

Natural History of Dural CCFs

The majority of patients with a dural CCF have no difference in mortality rates from those of the normal population because the lesion usually affects only the eyes. Spontaneous intracranial hemorrhage is exceptionally rare.[35] Thus, when one considers the natural history of a dural CCF, one is usually concerned with ocular morbidity.

Regardless of whether they drain anteriorly or posteriorly, 20–50% of dural CCFs close spontaneously, after angiography, or after air flight travel (Figs. 10 and 22).[69] In

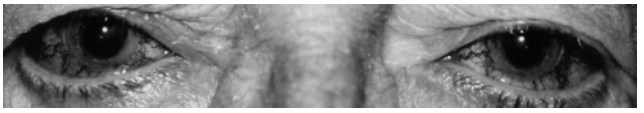

FIG. 13. Photograph showing bilateral ocular manifestations in a patient with a right-sided spontaneous dural CCF. The patient has bilateral red eyes, with dilated, tortuous conjunctival and episcleral veins.

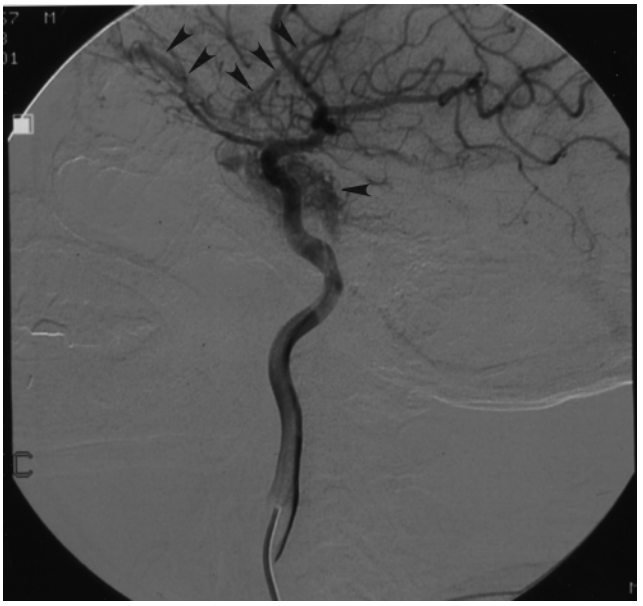

FIG. 14. Selective left ICA arteriogram shows a dural CCF *(single arrowhead)* with drainage into the SOV *(double arrowheads)* and also into several cortical veins *(triple arrowheads)*.

some cases, the symptoms and signs begin to resolve within days to weeks after angiography. In others, they do not resolve until months to years after the fistula has become symptomatic.

I believe it is appropriate to follow up clinically patients who have mild ocular manifestations to determine if the fistula will close spontaneously. During the waiting period, patients do not need to alter their lifestyle. They should, however, be examined at regular intervals so that their visual function, intraocular pressure, and ophthalmoscopic appearance can be monitored.[89] During this time, exposure keratopathy caused by proptosis can be treated using ocular lubrication, and persistent bothersome diplopia can be treated using prism therapy or occlusion of one eye. Increased intraocular pressure rarely is so severe that it requires treatment.[41] If intraocular pressure is substantially elevated, one can attempt to lower it with one of the many topical agents that reduce the production of aqueous humor. Because in most cases the cause of the elevated intraocular pressure is raised episcleral venous pressure, such agents may not be helpful, however, and even agents such as latanoprost, a prostaglandin receptor agonist that increases uveoscleral outflow of aqueous humor, may not be effective in patients with a dural CCF because of the significant backup of arterial blood in the orbital veins. Nevertheless,

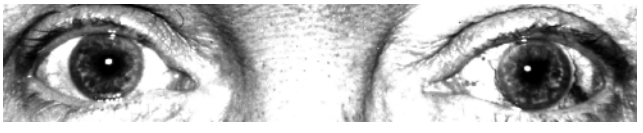

FIG. 15. Photograph of a 54-year-old woman with a spontaneous dural CCF, which was mistaken for chronic conjunctivitis. Note redness and minimal swelling of the left eye without proptosis or chemosis.

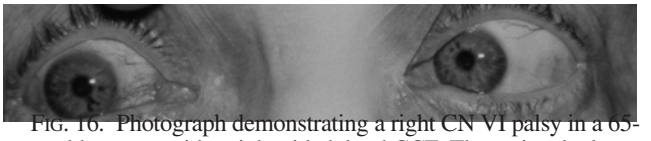

FIG. 16. Photograph demonstrating a right CN VI palsy in a 65-year-old woman with a right-sided dural CCF. The patient had systemic hypertension and presented with acute horizontal diplopia. Initially, she was believed to have experienced a vasculopathic CN VI palsy; however, on a follow-up examination 6 weeks later, it was noted that the conjunctival and episcleral veins of the right eye were dilated and tortuous compared with those of the left eye.

it is still worthwhile to administer these drugs for a few weeks because even a small reduction in intraocular pressure may protect the patient's vision. In the final analysis, however, the best treatment for severely increased intraocular pressure is closure of the fistula.

Patients with a dural CCF may experience acute worsening of ocular manifestations. This clinical deterioration results from an increase in blood flow through the fistula in some cases, but in others, it is caused by spontaneous thrombosis of the SOV.[28,93] Patients in whom clinical worsening is caused by spontaneous progressive thrombosis of the SOV usually begin to improve within several weeks, and most eventually experience complete resolution of symptoms and signs (Fig. 23). Systemic corticosteroids administered when deterioration occurs may lessen the severity of symptoms and signs and perhaps reduce the length of time until recovery occurs.[93]

Patients in whom a dural CCF persists or in whom such a fistula is not recognized may experience major hemorrhagic and other complications when they undergo intraocular or orbital surgery performed for other reasons, such as for cataracts or strabismus.[84] Others may undergo uncom-

FIG. 17. Image of the Pascal dynamic contour tonometer. This pneumotonometer may be used to diagnose dural CCFs. Photograph courtesy of Ziemer Ophthalmic Systems AG.

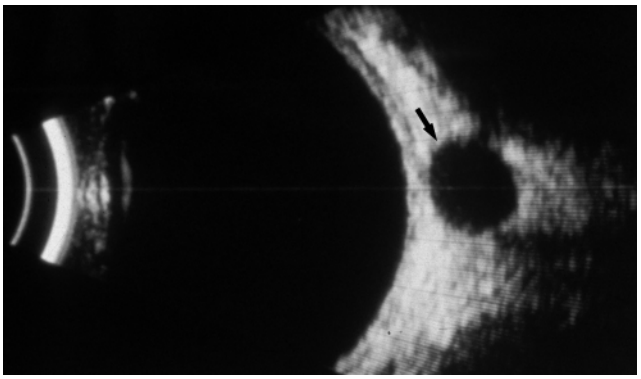

FIG. 18. Image showing ultrasonography of the orbit in a patient with an ipsilateral dural CCF. Note the large round void *(arrow)* representing a cross-section of an enlarged SOV.

plicated surgery, only to lose vision subsequently from ischemic complications of the fistula.[96]

Treatment

The visual manifestations of a dural CCF usually do not require local treatment. Occasionally, increased intraocular pressure requires treatment with topical or oral pressure-lowering agents. Although pressure-lowering ocular surgery has been advocated for patients in whom medical therapy does not reduce the intraocular pressure to an acceptable level,[48,84] I believe that if intraocular pressure remains unacceptably elevated despite maximum medical therapy, definitive treatment of the fistula should be performed instead of ocular surgery. Only if treatment of the fistula cannot be performed or is unsuccessful, or if the intraocular pressure remains elevated despite closure of the fistula, should ocular surgery be considered.[25] Similarly, although the proliferative retinopathy that may occasionally accompany a severe, high-flow dural CCF can be treated

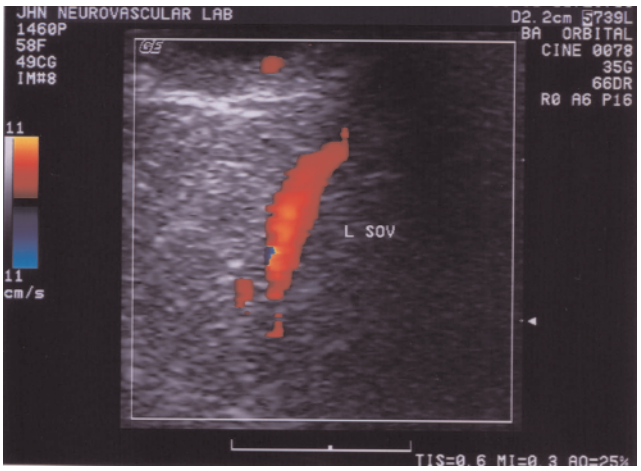

FIG. 19. Color Doppler flow image of the SOV in a patient with an ipsilateral dural CCF. The red color of the blood in the SOV indicates that it is flowing toward the eye rather than away from it, indicating that it is arterial rather than venous in origin. If the blood were flowing away from the eye, it would appear blue. Image courtesy of Dr. Peter J. Savino.

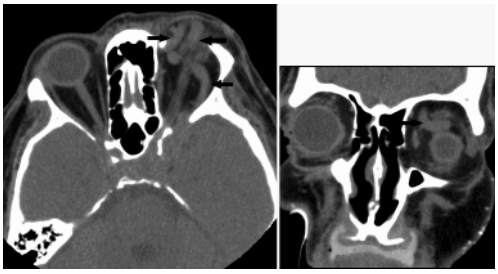

FIG. 20. Axial *(left)* and coronal *(right)* CT scans in a patient with a right-sided high-flow dural CCF. Note the enlarged SOV *(arrow)*.

successfully with photocoagulation,[25,36] it is best to treat the fistula producing the retinopathy whenever possible. Again, if the fistula cannot be treated or treatment is unsuccessful, photocoagulation may be needed to preserve vision.

Dural CCFs may be treated using direct surgery,[18,21,39,60,80] conventional radiation therapy,[107] stereotactic radiosurgery,[32] intermittent manual self-compression of the affected ICA with the contralateral hand,[44] or even occlusion of the ipsilateral ICA.[60] In general, however, endovascular embolization is the optimum treatment for those lesions that produce progressive or unacceptable symptoms and signs including visual loss, diplopia, an intolerable bruit, severe proptosis, and, most importantly, cortical venous drainage.[4,16,20,21,39,53,60,71,102] A number of synthetic and natural materials can be used for embolization, including absorbable gelatin (Gelfoam); Silastic; platinum coils; low-viscosity silicone rubber; autogenous clot, muscle, or dura; tetradecyl sulfate (a sclerosing agent); polyvinyl alcohol particles (Ivalon); ethanol; oxidized cellulose (Oxycel); and isobutyl-2-cyanoacrylate glue (Bucrylate).[21,31,49,51,53,54,60,106]

In patients with a fistula fed only by meningeal branches of the ECA or by meningeal branches from both the ECA and ICA, the embolization material is introduced via a microcatheter placed in the ECA and passed into the spe-

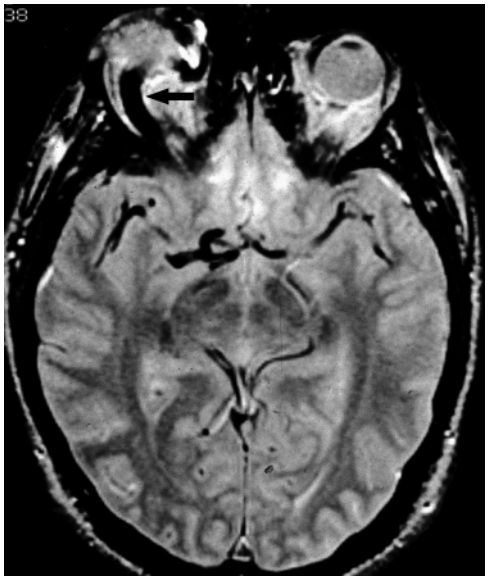

FIG. 21. A T2-weighted MR image in a patient shows a right-sided dural CCF. Note the enlarged SOV *(arrow)*.

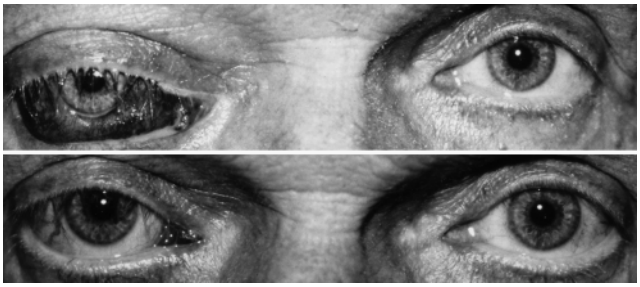

FIG. 22. Photographs showing a patient's appearance before *(upper)* and after *(lower)* spontaneous closure of a dural CCF. *Upper:* The patient was a 59-year-old man who developed progressive redness and swelling of the right eye. The patient's right eye is swollen and red, and significant conjunctival chemosis is apparent. Ocular pulse amplitudes were asymmetric, with the higher pulse on the right side. Cerebral angiography confirmed a right-sided dural CCF fed by branches of the right ICA and ECA. The treating physician chose to follow up the patient without intervention. Within 1 week after angiography, the patient began to experience a reduction in swelling and redness of the right eye. *Lower:* One month after angiography, the patient shows minimal swelling and redness of the right eye. Intraocular pressure and ocular pulse amplitudes are normal and symmetrical.

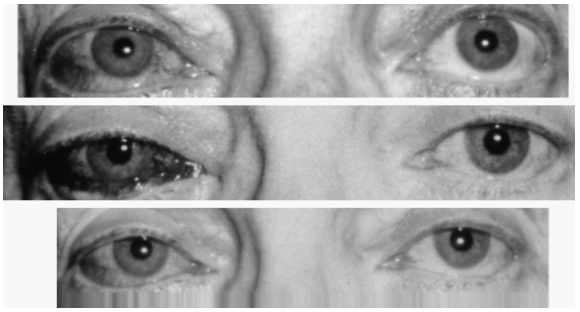

FIG. 23. Photographs showing a progressive worsening of a patient's ocular appearance in the course of spontaneous closure of a dural CCF. *Upper:* The appearance of the patient when first examined. The patient was a 73-year-old woman who developed a red right eye. Note the mild redness of the right eye, associated with swelling of the right eyelid and mild proptosis. An evaluation revealed a right-sided dural CCF fed by branches of the right ICA and ECA. The treating physician elected to follow up the patient without intervention. *Center:* Two months later, the patient experienced an abrupt increase in the swelling and redness of the right eye. The right eye now shows increased redness, swelling, conjunctival chemosis, and proptosis. Color Doppler imaging showed that the right SOV was occluded, and cerebral arteriography confirmed that the fistula was in the process of spontaneously occluding. *Lower:* One month after the onset of worsening and 3 months after the onset of visual manifestations, the right eye shows only minimal redness and swelling. The patient had a normal appearance 3 months later.

cific branch or branches that feed the fistula.[21,60,68,106] The ICA usually is not embolized unless the interventionalist can successfully catheterize the meningohypophyseal trunk or other meningeal feeders from the artery.[14] When the fistula is fed only by branches from the ECA, successful closure of these branches is often possible and associated with rapid resolution of all ocular symptoms and signs.[7,60]

When a dural fistula is fed by branches from both the ECA and ICA, embolization of the feeders from the ECA using various agents such as polyvinyl alcohol or glue may reduce the blood flow in the fistula sufficiently such that nonembolized feeders from the ICA will thrombose spontaneously.[54,69] Embolization of feeders from the ICA is almost never appropriate because of the significant potential neurological morbidity from distal embolization.[60] If thrombosis does not occur with this technique, the fistula can be treated by placement of detachable platinum coils or detachable balloons within the cavernous sinus using a transvenous route. The favored approach usually is via the femoral or internal jugular vein into the inferior or superior petrosal sinus, and from there into the cavernous sinus, but if this approach fails, a variety of other approaches may be used, most of which involve cannulation of the superior or inferior ophthalmic vein.[1,5,7–11,13,16,20–22,26,27,34,42,49,51,53,56,57,59,63,67,73,78,83,97,102] In some cases, more than one session and more than one approach are needed.[11,57]

At my institution, we prefer the direct SOV approach performed in most cases by surgical exposure of the vessel (Fig. 24 and accompanying video). All procedures are performed in a neurosurgical operating room under fluoroscopic guidance. With the patient in a state of general anesthesia, a sheath is placed in a common femoral artery to permit intraoperative angiography. Following appropriate preparation and draping of the affected eye and orbital regions, a curvilinear skin incision is made at the level of the superior lid crease or the superior sulcus of the upper eyelid nasally, using magnification provided either by an operating microscope or magnifying loupes (Figs. 24A and 24B). The incision is continued down through the orbicularis oculi muscle, with careful attention to hemostasis. The orbital septum is identified and opened with sharp spring-action scissors, exposing the retroseptal orbital fat. The SOV is identified using blunt dissection. The vein appears as a reddish-blue vessel that varies in size from 3 to 8 mm in diameter (Fig. 24C). The vein is carefully cleaned from its attachments to surrounding orbital fat until a segment measuring 10–20 mm long is exposed (Fig. 24D). Two ligatures, consisting of 2–0 black silk sutures for small veins and silicone vascular loops 1 mm in diameter for large veins, are passed underneath the vessel using a Kelly or right-angled clamp (Fig. 24E), and the two ends of each ligature are then passed through a piece of tubing that varies in size from a pediatric feeding tube to a French catheter, depending on the diameter of the ligatures (Fig. 24F). The two ligatures are then placed as far apart as possible to isolate a segment of the vein.

Once the ligatures are in place around the vein and are tightened down to prevent bleeding (Fig. 24G), a small incision is made in the wall of the portion of the vein between the ligatures using sharp spring-action iris scissors (Fig. 24H). Brisk arterial bleeding indicates that a full-thickness opening has been achieved. The ligatures are then tightened using the feeding tubes or catheters. A microcatheter, the size of which is determined by the diameter of the vein, is placed into the vein opening, using a jewelers' forceps to steady and direct it (Fig. 24I). The placement is facilitated using a two-person technique, with one person threading the catheter and the other manipulating the ligatures to allow passage of the catheter while limiting bleeding. The catheter is threaded posteriorly under fluoroscopic control

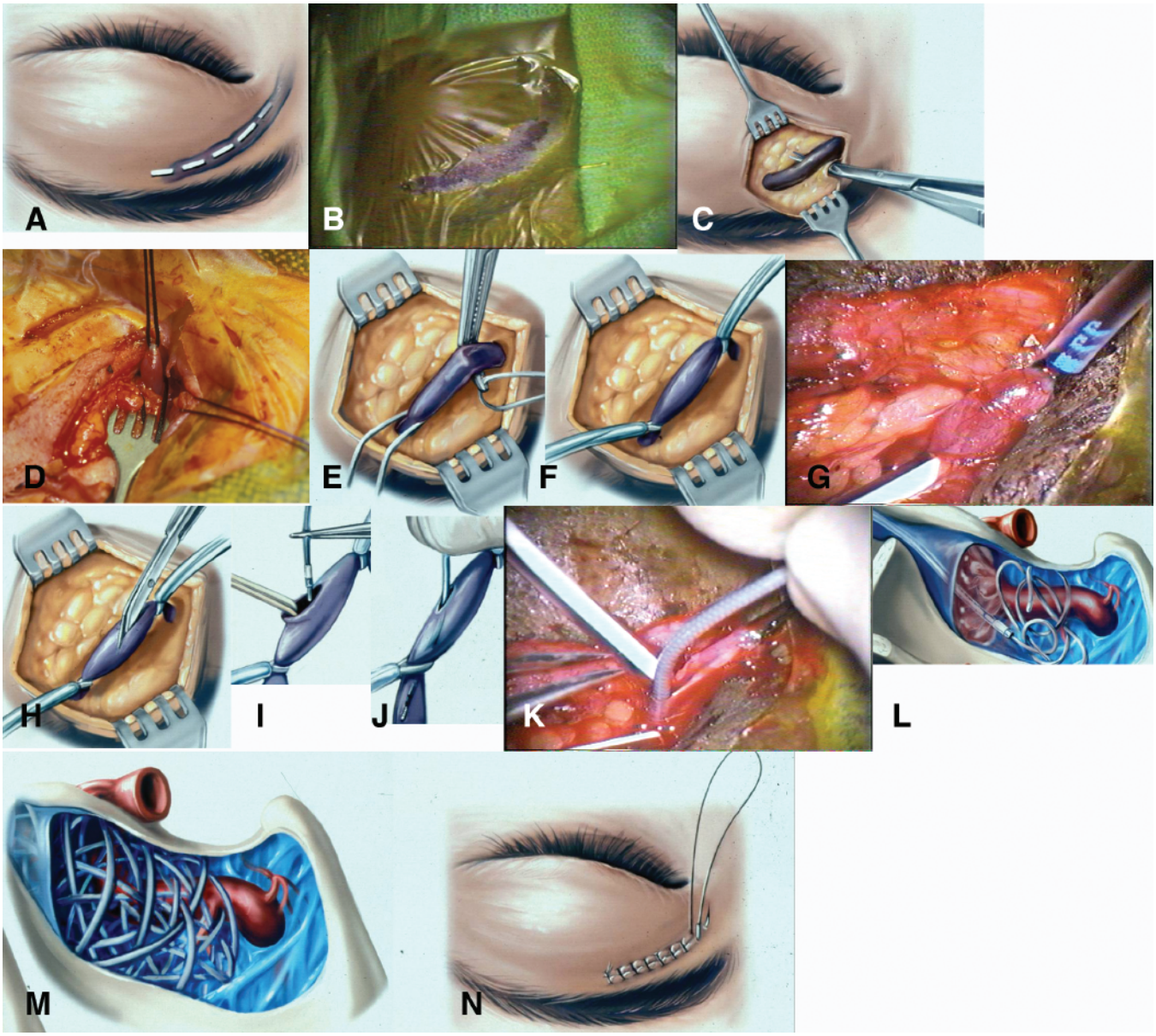

FIG. 24. Intraoperative photographs (B, D, G, K) and artist's illustrations (A, C, E, F, H–J, L–N) demonstrating the direct SOV approach for closure of a dural CCF. A: The location of the skin incision (dashed line) for a left SOV approach. B: The location of the incision site. C: The exposure of the SOV after opening of the orbital septum. D: Another case of a left CCF in which the left SOV is isolated by 4-0 black silk suture. E: Isolation of a segment of the SOV between two silicone vascular loops. F: Both ends of each of the loops have been passed through small pediatric feeding tubes, and the tubes have been secured down against the vein to control bleeding. G: Isolation of a portion of the SOV between two pediatric feeding tubes that have been secured down against the vein, thus permitting control of blood flow during the procedure. H: Opening of the SOV using sharp spring-action iris scissors. I: Placement of a microcatheter into the SOV. Note the use of a vein guide to elevate the wall of the vessel to allow easier access to the catheter. Also note that the catheter is placed in a medial-to-lateral direction. J: Threading of the catheter through the SOV. The proximal feeding tube controlling blood flow in the SOV has been loosened slightly to allow passage of the catheter through the vein. K: Threading of the microcatheter through the SOV. L: A microcatheter has been placed in the cavernous sinus and a thrombogenic coil is being introduced into the sinus. M: The appearance of coils in the cavernous sinus. N: Closure of the skin incision using a running 8-0 black silk suture.

until the tip is observed to be within the cavernous sinus (Fig. 24J and K), at which time multiple platinum coils are detached in the cavernous sinus (Fig. 24L and M) until the fistula is closed as determined by intraoperative angiography (Fig. 25). Once it is clear that the fistula is closed, the catheter is withdrawn, and the SOV is permanently occluded using bipolar cautery and ligatures. When the vein is quite large and its wall is thick, the incision can be closed

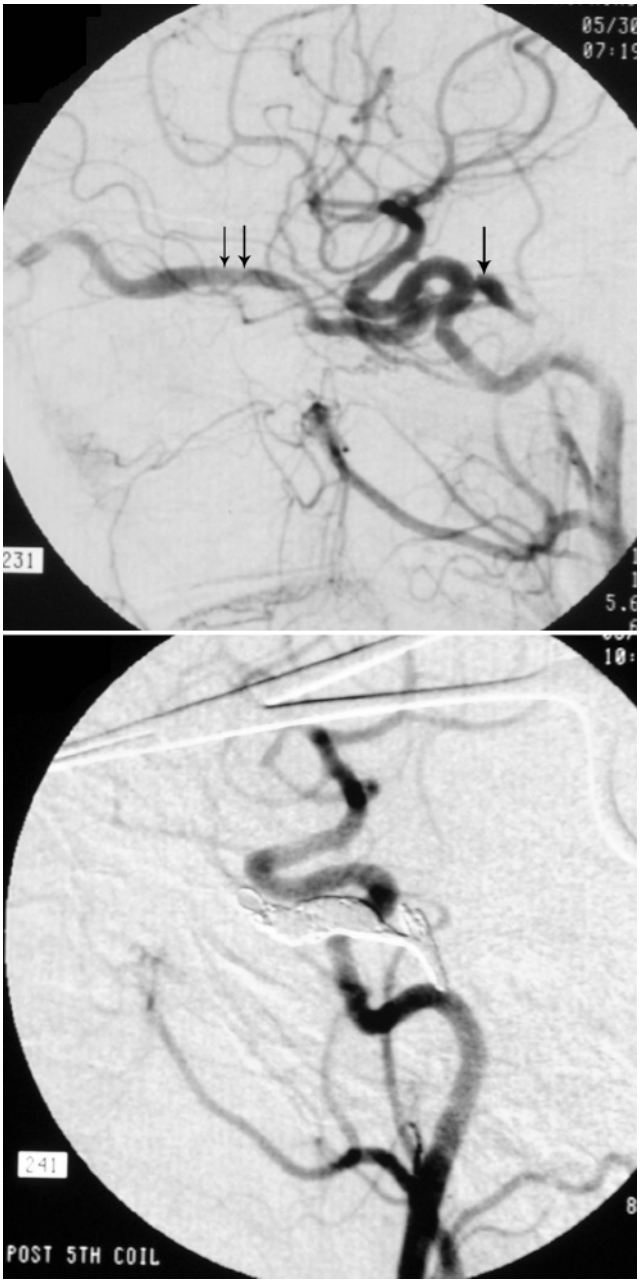

FIG. 25. Preoperative *(upper)* and postoperative *(lower)* angiograms in a patient who has undergone transvenous closure of a left-sided dural CCF using the direct SOV approach. Closure was achieved by placing five platinum coils in the left cavernous sinus. The left ICA has been preserved. *Double arrows* indicate the enlarged left SOV, and the *single arrow* indicates the fistula.

using a 10-0 nylon suture. The orbit is then irrigated with an antibiotic solution, and the skin incision is closed using a running 8-0 black nylon suture (Fig. 24N). No attempt is made to close the orbital septum in most cases. A Xeroform gauze pad is placed over the incision site, and a light eye patch is placed over the pad for 24 hours. The incision site is then treated with a topical antibiotic ointment, such as erythromycin or bacitracin, and the skin suture is removed in 5–7 days. When performed by an experienced team, this approach is successful in the majority of cases and, in fact, may be the best initial treatment for all dural CCFs.[27,60,73,74,86] It is important to be aware, however, that should an attempt at closing a dural CCF transvenously via the SOV be unsuccessful and the SOV sacrificed or ligated during the procedure, there is a significant risk of increased venous pressure in the orbit with subsequent neovascular glaucoma and severe visual loss.[33] For this reason, physicians attempting this form of treatment should always have an alternative available, such as a transvenous approach through the inferior petrosal sinus, endovascular occlusion of the artery or arteries supplying the fistula, or direct surgery on the fistula.[66,72]

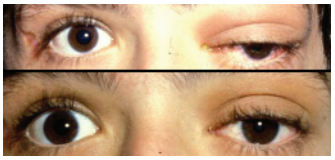

FIG. 26. Appearance of patient with a left-sided dural CCF before *(upper)* and immediately after *(lower)* transvenous occlusion of the fistula using an SOV approach. The patient's orbital and lid swelling as well as ocular injection improved immediately after occlusion.

Successful closure of dural CCFs by standard particulate or glue embolization is possible in 70–95% of all cases.[3,39,60,62,69] When transvenous coil or balloon occlusion of the fistula is used, the rate of successful closure is 90–100%.[71,73]

Complications from endovascular treatment of dural CCFs are rare except in patients with connective tissue disorders such as Ehlers–Danlos syndrome.[45,91] Nevertheless, significant complications may occur, including hemorrhage at the catheter site or in the orbit, local infection, sepsis, ophthalmic artery occlusion,[104] and both transient and permanent neurologic deficits, particularly ocular motor nerve pareses.[2,60,82] Devoto and associates[23] reported the development of increasing proptosis, chemosis, and markedly elevated intraocular pressure associated with a mid-dilated and poorly reactive pupil during attempted transvenous closure of a dural CCF. The patient was treated with intravenous mannitol and acetazolamide, as well as topical timolol maleate and apraclonidine. At the same time, the interventionalist introduced larger coils into the anterior portion of the cavernous sinus. Within several minutes, the condition had resolved, and the patient had a successful result with vision of 20/20 the next day.

Prognosis After Treatment

It is not unusual for dural CCFs to recanalize or form new abnormal vessels after transarterial embolization with particles or other material.[64] Recurrence of ocular symptoms and signs indicate the recurrence of the fistula, and patients in whom manifestations recur require repeat angiography and consideration of further treatment. I am less concerned about incomplete closure or recurrence if the fistula has been closed transvenously using detachable coils or balloons. Symptoms and signs usually begin to improve within hours to days after successful closure of a dural CCF (Fig. 26).[27,60,73] Any preexisting bruit immediate-

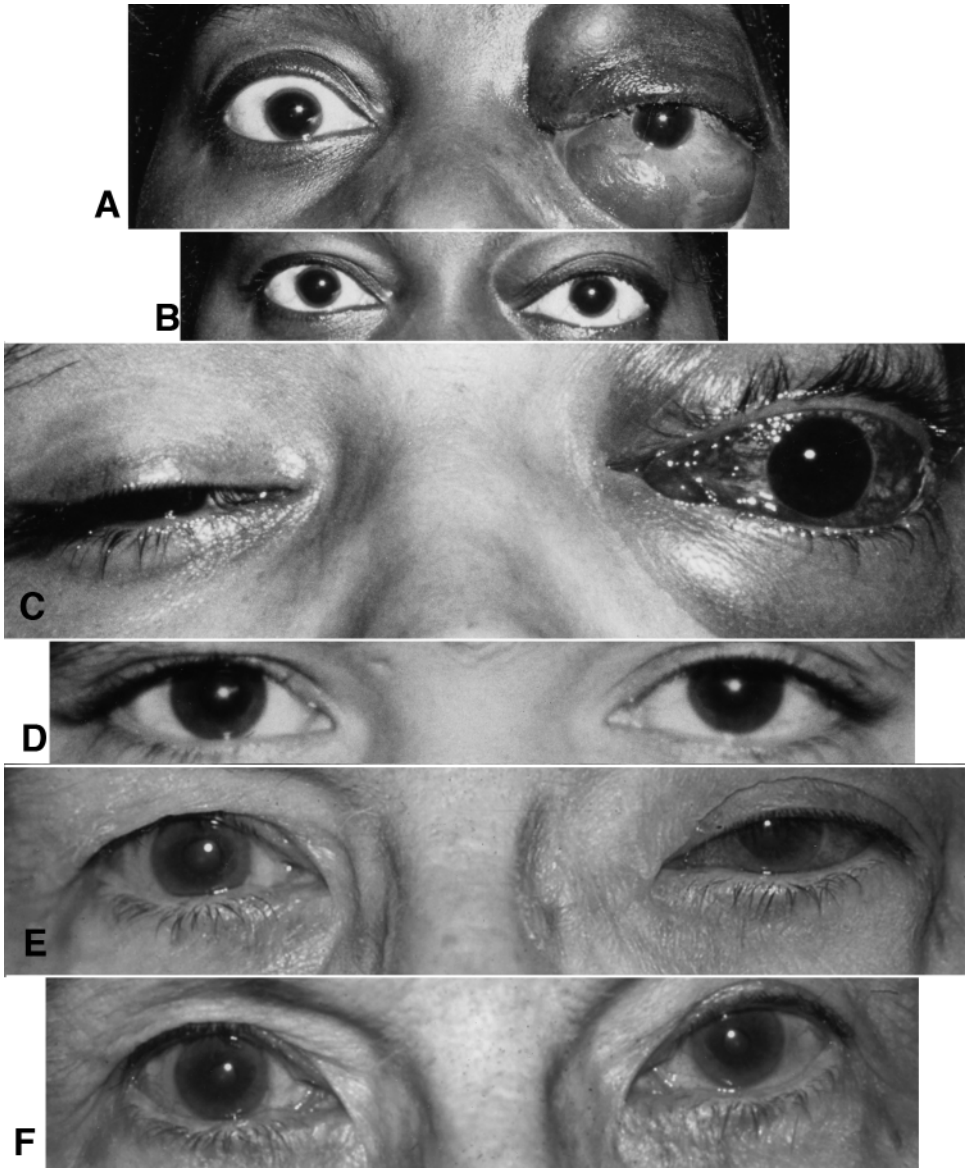

FIG. 27. Photographs of patients before and after transvenous occlusion of dural CCFs. A: Preoperative appearance of a 35-year-old woman with a long-standing, left-sided CCF. B: Postoperative appearance of the patient 4 months after occlusion of the fistula through the left SOV using a detachable balloon. C: Preoperative appearance of a 34-year-old man with a left-sided CCF. D: Postoperative appearance of the patient 6 months after balloon occlusion via the left SOV. E: Preoperative appearance of a 65-year-old woman with a left-sided CCF. F: Postoperative appearance of the patient 2 months after occlusion of the fistula through the left SOV using detachable platinum coils.

ly disappears, and intraocular pressure immediately returns to normal. Proptosis, conjunctival chemosis, redness of the eye, and ophthalmoparesis (whether caused by orbital congestion or an ocular motor nerve paresis) usually resolve completely within weeks to months, and most patients have a normal or near-normal external appearance within 6 months (Fig. 27). At the same time, patients with visual loss caused by choroidal effusion or detachment usually experience substantial if not complete recovery of visual function.[64] Unfortunately, patients with visual loss caused by retinal damage (for example, central retinal vein occlusion) usually have persistently poor visual function.[61]

Patients whose dural CCFs are treated using techniques other than endovascular closure, such as stereotactic radiosurgery, often take longer to improve than patients whose fistulas are closed using endovascular techniques.[6,58] Nevertheless, these techniques may provide excellent results over time.

Conclusions

The diagnosis and management of dural CCFs have dramatically improved in recent years. The widespread availability of noninvasive imaging techniques, combined with improvements in catheter angiography, permit rapid and accurate diagnosis in most cases, and new endovascular

therapeutic techniques allow most patients with these lesions to be treated successfully with little or no morbidity or death and with resolution of most, if not all, clinical manifestations.

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