



Classifications of Cranial and Spinal Dural Arteriovenous Fistulas and Their Endovascular Embolization

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Abstract

Dural arteriovenous fistula (DAVF) represents approximately 10–15% of all intracranial vascular malformations. DAVFs can be benign lesions, but the angioarchitecture of retrograde venous drainage or cortical venous reflux makes these lesions an aggressive high risk of neurological deficits and mortality caused by rupture or venous congestion. Endovascular embolization is the first line treatment for DAVF. Both transarterial and transvenous approaches have expanded the complex DAVFs that we can treat. The selection of endovascular approach depends on the angioarchitecture of the DAVF and experiences of neurosurgeons, the feeding arteries, lesion location, venous outflow direction. Surgery and stereotactic radiosurgery are used in few cases that endovascular approaches are impossible or unsuccessful.

Keywords

Cranial · Spinal · Dural arteriovenous fistula
Pathology · Classification · Therapy

1.1 Introduction

Dural arteriovenous fistulas (DAVFs) are the most common type of spinal vascular malformations (AVMs), but account for 10–15% of all intracranial arteriovenous malformations [1]. These abnormal connections between dural arteries and dural/pial veins can develop at everywhere of the dural mater. The most common places are transverse-sigmoid and cavernous sinuses, following tentorium, sagittal sinus, torcula, anterior cranial fossa, clivus, occipital foramen or dural sleeve of spinal nerve root [1, 2]. DAVFs cause a set of clinical signs and symptoms arising from venous congestion and pial venous rupture [1, 3, 4]. Patients with unruptured DAVFs may develop pulsatile tinnitus, bruit, headaches, visual changes, alterations in mental status, seizure, myelopathy, cranial nerve palsies and motor or sensory deficits depending on the location of the fistula [5–11]. The importance of venous drainage is well known when they distinguished between drainage directly into the dural sinuses and the pial veins in the clinical presentation of DAVFs (Table 1.1) [1, 3, 4]. The DAVF symptoms are produced by disruption of normal venous drainage causing changes in flow dynamics. Approximately 20–33% of DAVFs present with intracranial hemorrhage [6, 12].

CT scan is often useful for determining hemorrhage or oedema in DAVF diagnosis (Fig. 1.1). MRI can show parenchymal edema and flow void

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Table 1.1 Previous classifications of DAVFs and Zipfel classification

Djindjian-Merland classification (1977) [8]	Borden classification (1995) [3]	Cognard classification (1995) [5]	Zipfel classification (2009) [28]	Symptoms
I, Drainage into a sinus	I, Venous drainage directly into dural venous sinus or meningeal vein	I, Venous drainage into dural venous sinus with antegrade flow	I, DAVFs drain directly into dural venous sinuses; spinal extra-dural AVM without perimedullary vein reflux	No symptom or tinnitus
		IIa, Venous drainage into dural venous sinus with retrograde flow	II, DAVFs drain into dural sinuses but also have retrograde drainage into ophthalmic and bridging veins; spinal extra-dural AVM with perimedullary vein reflux	Proptosis, chemosis, headaches, epilepsy, cognitive dysfunction, dementia, intracranial hemorrhage, myelopathy
II, Sinus drainage with reflux into cerebral veins	II, With cortical vein reflux	IIb, Venous drainage into dural venous sinus with cortical vein reflux		
III, Drainage solely into cortical veins	III, Cortical vein drainage	III, Venous drainage directly into cortical veins	III, DAVFs drain into pial veins and do not have dural sinus drainage; spinal DAVF drained with perimedullary vein only	Intracranial hemorrhage, myelopathy
IV, With supra or infratentorial venous lake		IV, Type III with venous ectasias of the draining subarachnoid veins		
–	–	V, Spinal perimedullary vein drainage	II or III, Cranial or spinal DAVF	Intracranial hemorrhage, myelopathy

Cavernous fistula-modified Borden type I (petrosal sinus drainage) and II (ophthalmic vein drainage or cortical reflux) Spinal DAVF-modified Borden type II (extradural dural fistula) and III (intradural dural fistula)

signals of venous hypertension, such as pial vein engorgement, dilated venous pouch or abnormal vascular enhancement [1] (Fig. 1.2). Susceptibility-weighted imaging can clarify arteriovenous shunting of DAVF by demonstrating hyperintense venous signal due to rapid wash-in of oxygenated blood [1] (Fig. 1.3). Cranial CT angiography (CTA) or MR angiography (MRA) can demonstrate engorged arterial or venous vessels, enhanced transosseous vessels or asymmetric sinuses.

Negative cranial CTA or MRA can not completely exclude the diagnosis of DAVF. Cerebral digital subtraction angiography (DSA) is the gold

standard imaging to characterise the DAVF [1]. A full DSA, including bilateral internal carotid arteries (ICA), external carotid arteries (ECA) and vertebral arteries, is usually required to depict the DAVF. Superselective evaluation of smaller arteries is also helpful to clarify particular arterial feeders that can be chosen for embolization approach.

The Borden and Cognard classifications are the most well-known schemes to predict aggressiveness of intracranial DAVFs [3, 4]. The Borden classification system has unified spinal and intracranial DAVFs based on surgical practices [3]. The Borden classification

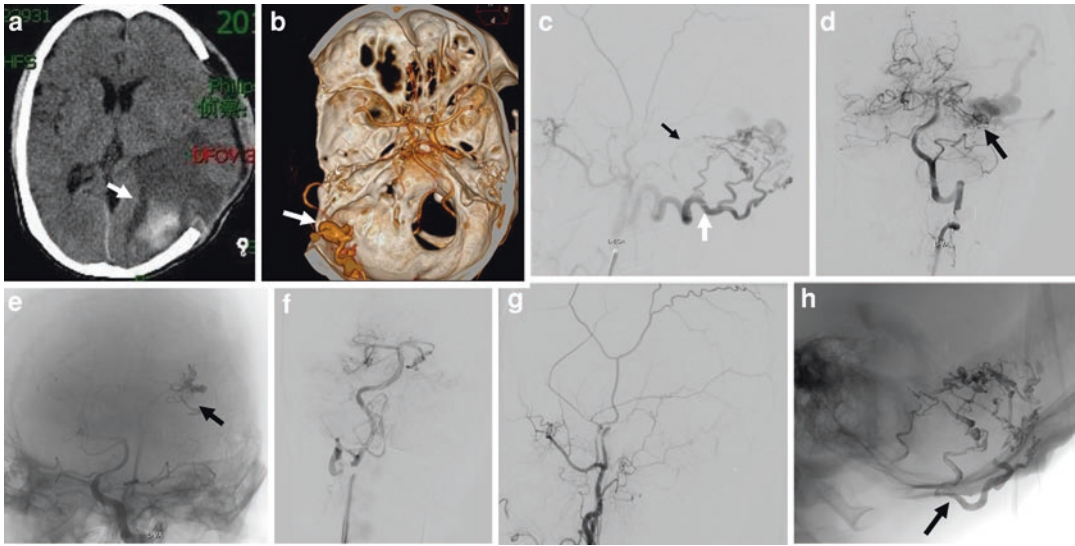


Fig. 1.1 A 63-year-old male patient presented with intracerebral hematoma. (a) CT scanning showing hematoma in left occipital lobe treated by surgery and decompression treatment (arrow). (b) CT angiography showing a giant venous ectasia of the left occipital lobe (arrow). (c) The left external carotid artery angiogram (lateral view) showing a Zipfel type 3 dural fistula supplied by the dural branches of the ascending pharyngeal artery (black arrow) and the occipital artery (white arrow). (d) The left vertebral artery angiogram (frontal view) showing the dural fistula supplied by the temporal branches of the left posterior cerebral artery (arrow). The dural fistula was drained

directly by cortical veins with a giant venous pouch. (e) Unsubtracted image (frontal projection) showed the posterior artery branches was embolized with Onyx firstly (arrow). (f) Control angiogram of the left vertebral artery (frontal view) showed the dural fistula was completely occluded. (g) Control angiogram of the left external carotid artery (lateral view) showed the dural fistula was completely occluded after left occipital artery embolization. (h) Unsubtracted image (lateral projection) showed the Onyx was injected to occlude the fistula via the occipital artery (arrow)

designates types I–III lesions as those with dural venous drainage without cortical venous reflux, dural venous drainage with cortical venous reflux, and cortical venous drainage without dural venous drainage, respectively. The Cognard classification designates types I, IIa, IIb, IIa + b, III, IV and V lesions as those antegrade dural venous drainage without cortical venous drainage (type I), retrograde dural venous drainage without cortical venous reflux (type IIa), antegrade dural venous drainage with cortical venous reflux (type IIb), retro-

grade dural venous drainage with cortical venous reflux (type IIa + b), cortical venous reflux without dural venous drainage (type III), cortical venous reflux with venous ectasias (type IV), and cervical perimedullary venous drainage (type V). Borden types II and III and Cognard types IIa, IIb, IIa + b, III, IV, and V DAVFs constitute aggressive lesions which must be treated [4].

The Borden and Cognard classifications have both focused chiefly on venous drainage direction of DAVFs excluding direct carotid cavernous

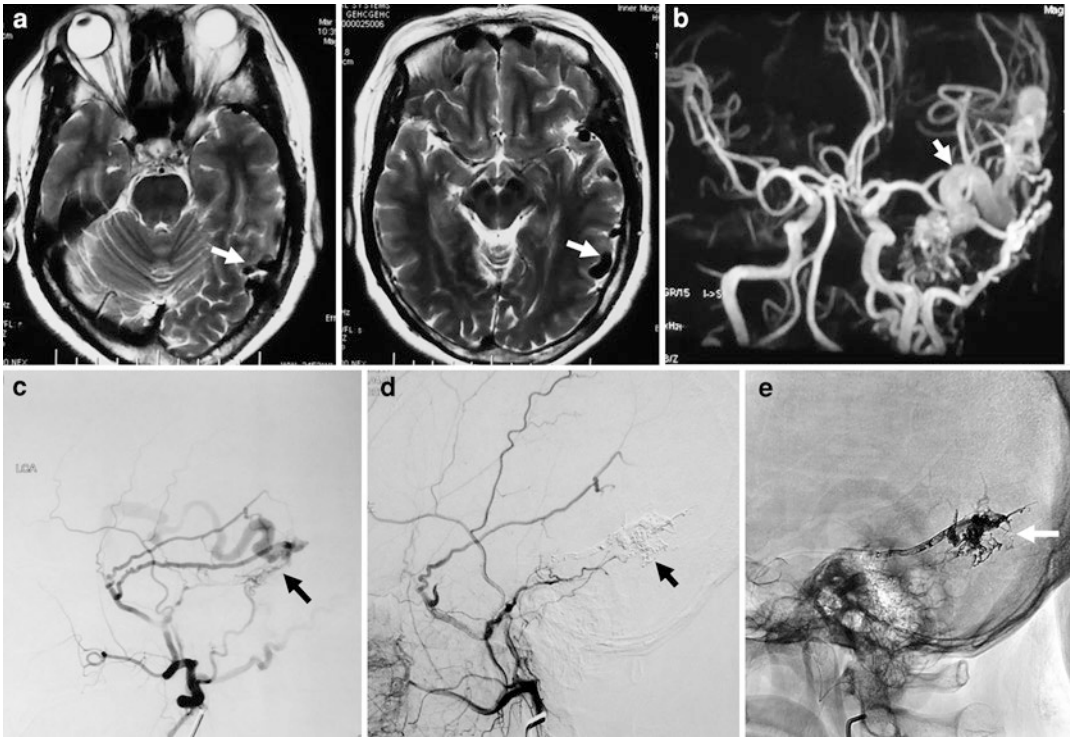


Fig. 1.2 A 50-year-old female patient presented with headaches. **(a)** Axial MR imaging, T2-weighted, showing flow void signals of cortical veins engorgement in the occipital lobe (arrow). **(b)** MR angiography (MRA) demonstrate engorgement of the left external carotid arteries and cortical veins of the left occipital lobe. **(c)** The left external carotid artery angiogram (later view) showing a Zipfel type 3 transverse sinus dural fistula (arrow) supplied by

the middle meningeal artery branches and cortical vein drainage. **(d)** The left external carotid artery angiogram (lateral view) showed the dural fistula was completely occluded after Onyx embolization. **(e)** Unsubtracted image (lateral view) showing the Onyx cast after successful infusion through the middle meningeal artery (arrow)

fistulas [3, 4]. Though adequately predicting extent of venous flow and properly categorizing the lesions angioarchitecturally, these grades lack of correlation with patients' presentation, natural history, and hemorrhagic risk. While the Cognard

system is more complicated to use, it can be changed. Therefore, Zipfel et al. proposed a classification of DAVFs based on natural history data, which is a modifier of the Borden grading system (Table 1.1) [13].

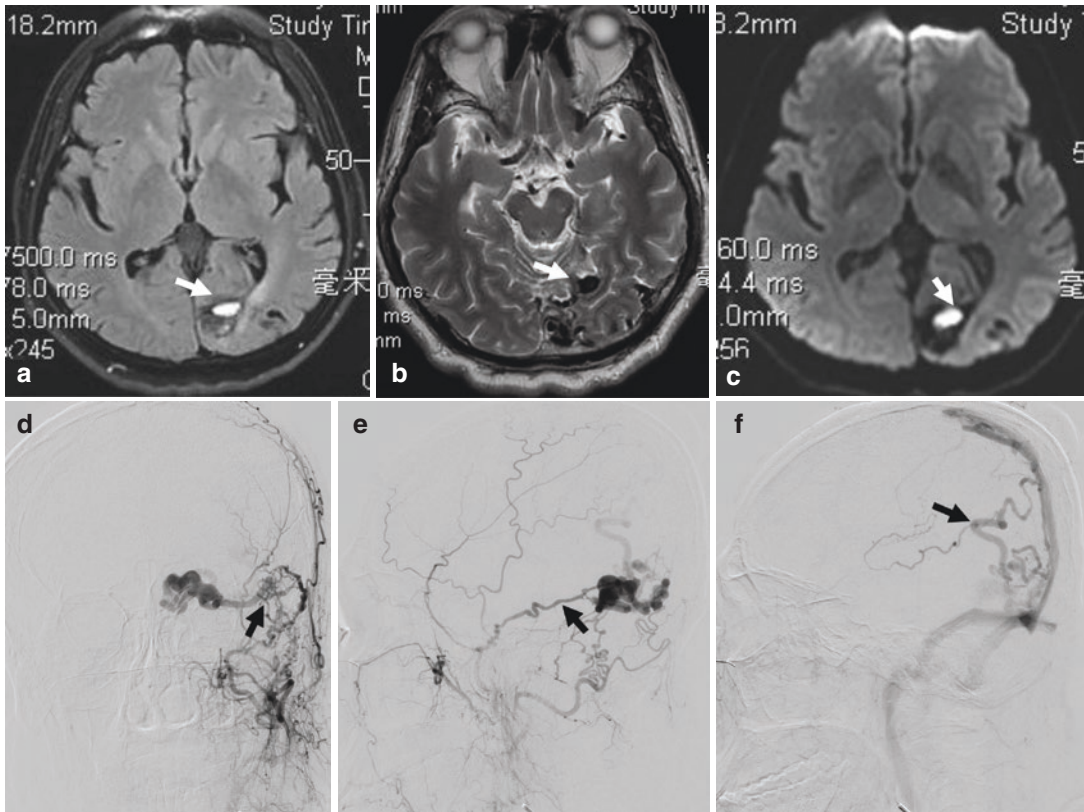


Fig. 1.3 A 59-year-old male patient suffered a second intracranial bleeding after 15 years of bleeding. (a) MR imaging, T1-weighted. (b) MR imaging, T2-weighted. (c) DWI MR imaging. Showing a subacute hemorrhage of the left occipital lobe and flow void signals of the engorged cortical veins. (d) the left external carotid artery angio-

gram (frontal view). (e) The left external carotid artery angiogram (later view). (f) The late phase of the left external carotid artery angiogram (later view). Showing a Zipfel type 3 dural fistula (arrow) supplied by the middle meningeal artery branches and cortical veins drainage

1.2 Zipfel Classification of DAVFs [13]

Type I DAVFs are those that drain into the dural sinus with antegrade venous flow, e.g. the flow of the veins draining from the parenchyma or spinal cord into the dural sinuses or epidural veins is antegrade, a carotid cavernous fistula (CCF) without cortical or superior ophthalmic vein (SOV) reflux.

Type II DAVFs drain into dural sinus with retrograde venous flow, a CCF with SOV reflux without cortical reflux. Intracranial type II DAVFs can drain into spinal perimedullary veins (Fig. 1.4).

Type III DAVFs are those that drain into the pial veins and the spinal coronal or perimedullary veins, a CCF with cortical drainage (Fig. 1.5). Type III spinal DAVFs can drain intracranially.

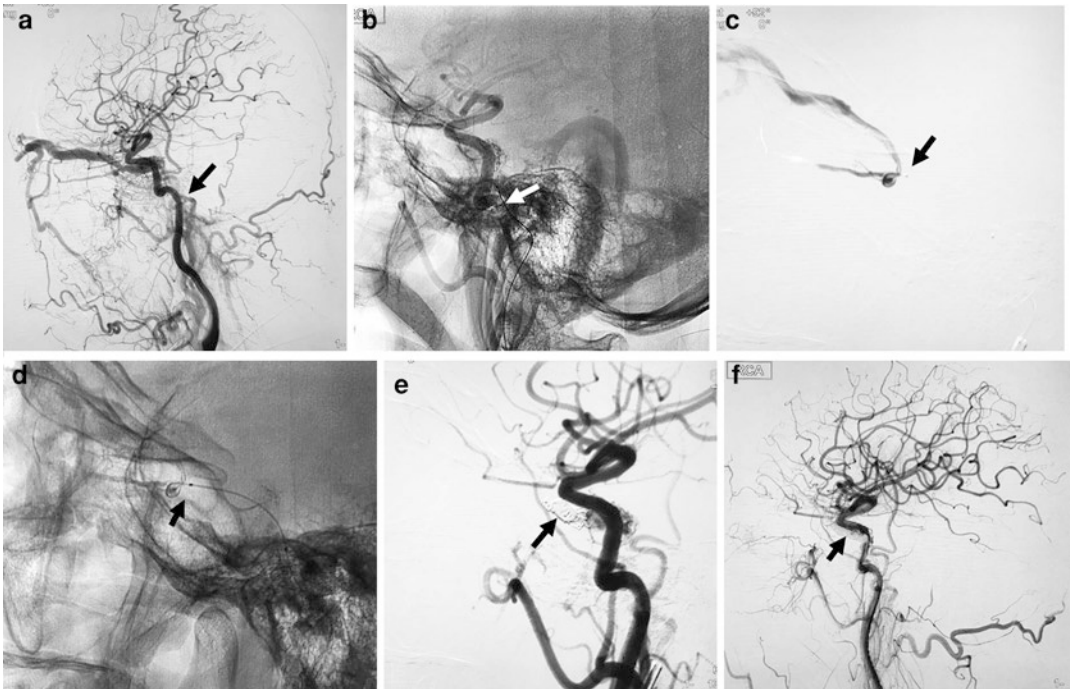


Fig. 1.4 A 46-year-old male patient presented with 6 months of right-sided proptosis. Transvenous embolisation of a right Zipfel type II cavernous sinus dAVF with Onyx and microcoils. **(a)** Lateral projection pre-embolisation angiogram, right carotid artery injection, showing multiple dural feeders arising from the external carotid artery and the internal carotid artery draining into superior ophthalmic vein and inferior petrosal sinus (arrow). **(b)** Unsubtracted lateral view showing successful catheterization of the inferior petrosal sinus (arrow). **(c)** Selective

injection showing the Echelon-10 microcatheter positioned within the origin of the superior ophthalmic vein (arrow). **(d)** Unsubtracted lateral view showing successful deployment of microcoils within the origin of the superior ophthalmic vein (arrow). **(e)** Lateral view of the carotid artery injection showing successful deployment of microcoils within the origin of the superior ophthalmic vein (arrow). **(f)** Lateral view of the carotid artery injection showing successful occlusion of the dural fistula after Onyx injection (arrow)

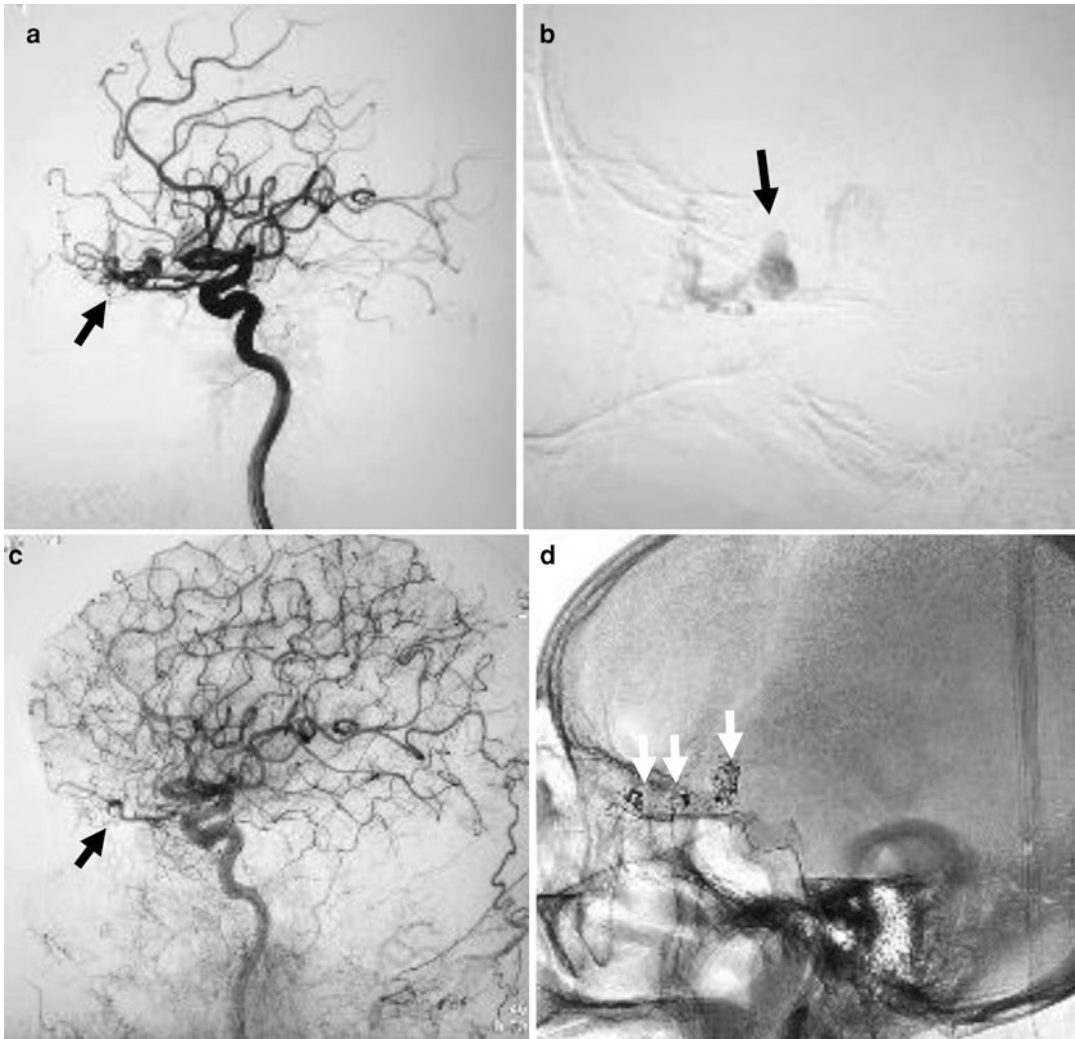


Fig. 1.5 A 55-year-old male patient presented with headaches. **(a)** The left internal carotid artery angiogram (lateral view) showing a Zipfel type 3 dural fistula arising from the anterior cranial fossa (arrow) supplied by the ethmoidal arteries arising from the ophthalmic artery and cortical vein ectasia. **(b)** Superselective angiography showing the microcatheter positioned through the left

ophthalmic artery and the large venous pouch (arrow). **(c)** The left internal carotid artery angiogram (lateral view) showed the dural fistula was completely occluded after Onyx embolization. **(d)** Unsubtracted image (lateral view) showing the Onyx cast after successful infusion the venous pouch (arrows)

1.3 Endovascular Approaches

Endovascular approach is the first-line treatment for most DAVFs. The mainstay for endovascular treatment involves occlusion of the arteriovenous shunts and its initial venous components while preventing adverse effects [14]. Inappropriate embolisation could cause sudden changes of the

flow dynamics in its venous drainage and potentially worsen the patient outcome (Fig. 1.6). Therefore, it is imperative to have comprehensive understanding of the arterial and venous components prior to initiating treatment [15].

The fistulous connection can be approached by either transarterial or transvenous approaches. Detachable balloons, polyvinyl alcohol, silk

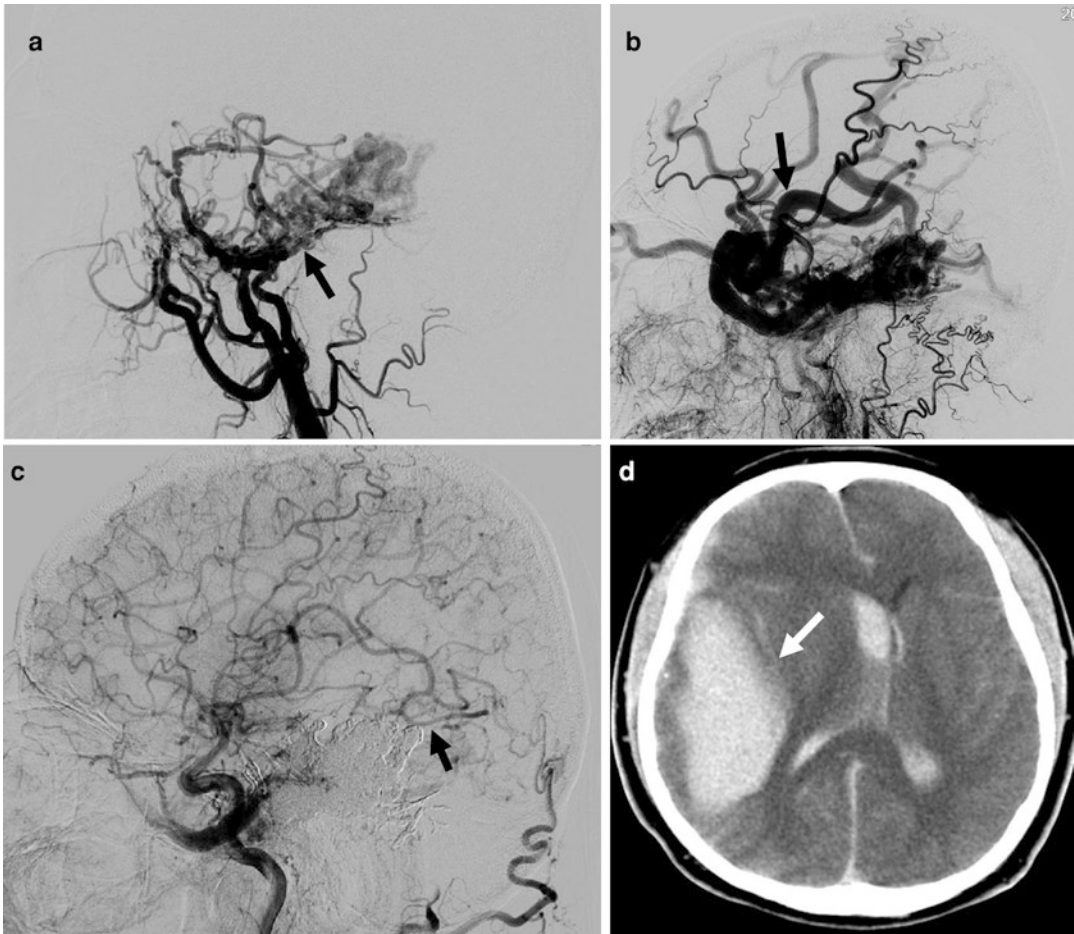


Fig. 1.6 A 47-year-old male patient presented with headaches. **(a)** The right external carotid artery angiogram (later view) showing a Zipfel type 3 dural fistula arising from the middle cranial fossa (arrow) supplied by the dural branches arising from the middle meningeal artery. **(b)** The late phase of the right external carotid artery angiogram (later view) showing the dural fistula drained

by multiple dilated cortical veins. **(c)** The right carotid artery angiogram (lateral view) showed the residual fistula supplied by the middle cerebral artery after Onyx embolization (arrow). **(d)** The patient was in coma half an hour after embolization. CT scanning showed a large intracerebral hematoma (arrow)

sutures, microspheres and detachable microcoils were used for treatment of cerebral AVM and DAVFs in the past decades. However, they have been widely replaced by current embolic agents, including *n*-butyl-2-cyanoacrylate (nBCA, glue, Trufill, DePuy Synthes, Raynham, MA) and Onyx (Medtronic, Irvine, CA) [15]. Recent advancements have introduced newer embolic agents, such as Squid (Emboflu, Switzerland), precipitating hydrophobic injectable liquid (PHIL; MicroVention, Aliso Viejo, California)

[15–17]. Flow diverters, such as the pipeline embolisation device (Medtronic Neurovascular, Irvine, California), have been reported to treat direct or indirect CCF, in which arterial feeder were ICA or its dural branch, a type of DAVF [18]. Endothelialisation of the flow-diverting stent allowed for occlusion of the CCF.

Endovascular approaches for DAVF embolization include transarterial, transvenous or a combination of both techniques. Endovascular DAVF embolization is often done under general

anesthesia to decrease patient motion. Heparinised saline flushes are routinely used to prevent catheter-associated thrombosis and embolic events. Various imaging techniques, including superselective angiography through microcatheter, three-dimension angiography, roadmapping and negative roadmapping, are often used to obtain clear images throughout the procedure [19].

1.4 Transarterial Embolization

Transarterial approach is the preferred treatment for DAVFs, in which transvenous approach is limited. In transarterial embolization, microcatheters are tracked over microwires to distal locations in feeding arteries, with the goal of getting the microcatheter as close to the fistula point as possible. Different liquid embolic agents can be used: nBCA, Onyx, Squid or PHIL. Injection of an embolic agent through a compatible microcatheter. Advantages of transarterial embolisation include decreased complications specific to commonly used transvenous approaches (e.g., venous perforation from catheterisation and bleeding from premature venous occlusion), ability to save functional venous system, decreased chance of flow redirection into an alternate venous pathway and avoidance of post-treatment de novo DAVF formation from venous hypertension [20].

1.4.1 nBCA

Before the appearance of Onyx, nBCA was widely used as “glue” for the embolisation of DAVFs [20, 21]. NBCA can quickly solidify when it comes in contact with blood. Ethiodol (ethiodised oil) must be added to nBCA for radiopaque and identifiable on fluoroscopy [22]. The concentration of nBCA is an important consideration as it changes the extent of its penetration before polymerisation. A mixture with high nBCA-to-Ethiodol ratio (high concentration of glue) polymerises more rapidly to embolize more proximal targets. A mixture with low nBCA-to-

Ethiodol ratio can achieve more distal penetration. NBCA of 25–33% concentrations are commonly used.

Prior to nBCA injection, the microcatheter must be advanced to the fistula point and helps successfully deliver the glue to the fistulous collateral networks [20]. Microcatheter angiography is obtained to help estimate the optimal glue concentration according to the flow velocity. Prior to injecting the nBCA and Ethiodol mixture, the microcatheter should be flushed with non-ionic solution of 5% dextrose, to prevent glue polymerisation within the microcatheter. The glue is then injected under negative road map as either a continuous column or a bolus [23]. Depending on the position of the microcatheter and the distance to the target vessel, 5% dextrose can be simultaneously injected through the guide or distal intracranial support catheter to promote more distal penetration of the glue [21]. After nBCA embolisation, it is important to rapidly remove the microcatheter to prevent the catheter from being entrapped to the vessel.

1.4.2 Onyx

Onyx is the preferred option for DAVF embolization. It is a liquid mixture of ethylene–vinyl alcohol copolymer (EVOH) suspended in dimethyl sulfoxide (DMSO). Catheters and syringes involved in the procedure must be DMSO compatible. Tantalum powder is added for radio-opacity. Before use, the Onyx mixture must be shaken for 20 min to evenly distribute the tantalum and obtain uniform radio-opacity [24]. When onyx is in contact with blood, it will precipitate into a non-adhesive substance with a characteristic “lava” flow pattern, resulting in blood vessel occlusion. After an in-depth understanding of the anatomy and hemodynamics of the fistula, the microcatheter must be flushed with DMSO before the injection of Onyx. The negative road-map imaging is used to inject Onyx under direct visualization to identify any premature leaks. The injection speed can be customized to optimize vessel penetration. Once Onyx reflux is observed, the injection should be stopped for about 30–90 s

to allow Onyx to solidify before next injection. When the Onyx reflux forms an ideal plug around the tip of the microcatheter, it can push Onyx into the fistula. Multiple control angiograms are usually necessary during the entire operation to carefully monitor the progress of the embolization. Since the injection of Onyx needs to be very precise, it is not uncommon for treatment with a single pedicle to last more than an hour or to divide the embolization into multiple stages. After embolization, the microcatheter can be removed from the Onyx cast by continuous gentle traction. The ability of prolonged Onyx injections allows curative embolization of DAVF with multiple feeders that form a complex vascular network with a single injection, especially when Onyx reaches the venous side [14, 25–27] (Fig. 1.7). Therefore, endovascular Onyx embolization has shown a higher DAVF cure rate than nBCA [28].

1.4.3 PHIL and Squid

PHIL is an iodinated copolymer dissolved in DMSO and will precipitate to form a non-adhesive substance when it contacts with blood. The agent can be used immediately (without shaking) because the radiopaque iodine is covalently bonded to the compound. Since it does not contain tantalum, very few artifacts can be seen in the postoperative follow-up CT scan. The agent can penetrate into blood vessels less than 10 μm . PHIL can be used in three concentrations, the lowest concentration is most commonly used for DAVF embolization. PHIL is currently not available for commercial use in China, but an active trial is underway to evaluate its effectiveness in the treatment of AVM. Leon et al. describes the use of PHIL in eight cases using Apollo (Medtronic) or Marathon (Medtronic) microcatheter to treat five skulls and three spinal

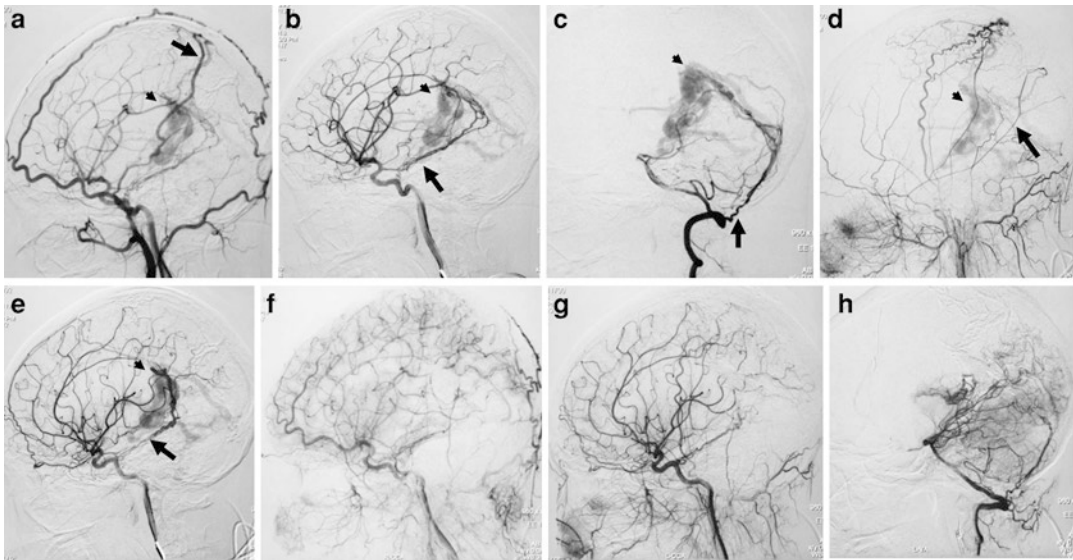


Fig. 1.7 A 29-year-old female patient presented with headaches. (a) The right external carotid artery angiogram (lateral view). (b) The right internal carotid artery angiogram (lateral view). (c) The right vertebral artery angiogram (lateral view). (d) The left external carotid artery angiogram (lateral view). (e) The left internal carotid artery angiogram (lateral view). Showing a Zipfel type 3 tentorial dural fistula multiple arterial feeders arising from bilateral middle meningeal arteries (arrow), bilateral

meningohypophyseal arteries (arrow), and bilateral posterior meningeal arteries (arrow), draining into dilated pial veins (arrowhead). Post-embolization angiograms. (f) The right carotid artery angiogram (lateral view). (g) The left carotid artery angiogram (lateral view). (h) The left vertebral artery (lateral view). Showing the dural fistula was completely occluded after transarterial Onyx embolization

cords DAVF [29]. Laming et al. reported that Apollo, Marathon and Headway Duo (MicroVenton, Tustin, California, USA) microcatheters are used in 30 PHIL embolization procedures [30].

Squid is another liquid embolic agent used to treat DAVF, but it is not currently on the market in China. Squid is an ethylene vinyl alcohol copolymer, available in two versions: Squid 12 and Squid 18 (Emboflu, Switzerland). Squid has 30% less tantalum content than Onyx and has micronized tantalum, which may help to better observe the structure behind the embolic material and provide a more uniform solution than Onyx [30, 31]. Akmanjit et al. reported a case series of nine cases of DAVF treated with Squid and Sonic detachable microcatheter [31]. Much like Onyx, the Squid is injected in a “plug and push” manner. In their case series, they used a higher density Squid 18 for initial plug formation and a lower viscosity Squid 12 for distal penetration. Gioppo et al. reported the use of Squid with the Headway Duo microcatheter (MicroVenton, Tustin, California, USA) to treat a complex case of DAVF [17].

1.5 Transvenous Approach

In the transvenous method, the catheters are retrogradely inserted into the affected sinus or the affected pial vein to occlude them using a microcoils, liquid embolic agent, or a combination of them (Fig. 1.4). For the transvenous approach, appropriate patient selection is essential to achieve complete occlusion and avoid complications. The selected sinus or pial vein should fully participate in fistula drainage, not in normal cerebral venous drainage, and the venous access should be completely occluded for proper treatment [32]. When the DAVF is supplied by many small tortuous artery, the transvenous approach is preferred because of the lack of transarterial access. When the DAVF is only supplied by the branch directly from the ICA or the vertebral artery, when the DAVF is supplied by the dangerous extracranial to intracranial anastomosed artery, or when the DAVF is supplied by

the nutrient arteries of the cranial nerve, the transvenous embolization should be considered [33, 34]. The transvenous route is the first-line treatment for indirect CCF, in which dangerous anastomosis, arteries supplying cranial nerves, and very small supply arteries supplying fistulas are very likely to be involved [34, 35].

Various approaches are used to achieve transvenous access. A transvenous access can be established through the femoral vein, the internal jugular vein, or directly puncture the draining venous pouch under the guidance of drilling, craniotomy, or ultrasound [36, 37]. In the case of a thrombotic sinus, the transvenous approach can be achieved by recanalization of the closed venous segment, such as traversing the ipsilateral occluded inferior petrosal sinus to access the trapped cavernous sinus or traversing the occluded sigmoid sinus to access an isolated transverse sinus [38, 39]. In this method, neuro-interventionists need to carefully manipulate the catheter and guide wire to avoid the risk of thrombosis sinus or vein perforation [39]. Sometimes the contralateral approach can also be used, such as the contralateral jugular vein can be used to pass through torcula to the sigmoid sinus [40]. The trapped sinus can also be directly punctured by drilling or craniotomy [36]. The direct access to the trapped cavernous sinus in the indirect CCF can be achieved by a transforaminal approach via foramen ovale or transorbital puncture [41, 42]. The hybrid angio-operating room is the ideal choice for this type of combined endovascular surgical method.

Coil embolization is very effective for filling and occluding affected sinuses or venous pouch, especially an isolated transverse sinuses [24, 33]. Sometimes it is necessary to combine Onyx with coils to help seal the associated sinus or venous pouch [33, 43, 44]. Some authors prefer the “dual catheter technique,” one is a proximal microcatheter used to deploy the coils, and the other is a distal microcatheter used to inject Onyx after coiling [33].

For ethmoidal or anterior cranial fossa DAVF, the blood supply arteries are usually small and very tortuous, making it difficult or impossible to perform a safe superselective transarterial catheter

terization [45]. In these cases, transvenous coil or Onyx embolization may be safe when there is an uncurved draining vein that allows the catheter to enter the venous pouch [45, 46]. Albuquerque and colleagues describe transvenous Onyx embolization for the treatment of high-risk transverse sigmoid sinus DAVF, in which a microcatheter is passed through the sinus into the venous pouch and positioned to the arterial ostium [45]. In these cases, Onyx penetrates into multiple arterial suppliers, and a small amount of reflux enters the venous pouch [45]. According to this report, this technique is safe when there is a venous pouch or an isolated sinus.

When a DAVF flows directly into the transverse sigmoid sinus and there is no venous pouch, the possibility of Onyx reflux to the normal sinus is high. A large amount of reflux to the normal sinuses can cause pulmonary embolism or inadvertent sinus thrombosis. In these cases, reconstructive transvenous balloon-assisted embolization was introduced as an option. For this technique, a microcatheter and a DMSO-compliant balloon are simultaneously guided to the affected transverse sigmoid sinus [47]. After the balloon is inflated, Onyx 18 is injected around the balloon and slowly penetrates into the blood suppliers resulting in complete occlusion of DAVFs [47]. In this technique, the preservation of normal cortical veins, such as Labbé vein and normal bridging vein, is essential to prevent complications such as venous infarction and cerebral or cerebellar hemorrhage [48]. It is also possible to temporarily block the main feeding artery during the Onyx embolization of the fistula venous pouch, especially in treatment of direct CCF [49]. When the venous drainage of the brain depends

on the transverse sinus of the fistula, the use of a stent to reconstruct the transverse sinus, with or without transarterial Onyx embolization may be a good choice [50].

1.6 Stereotactic Radiosurgery

Stereotactic radiosurgery (SRS) is usually reserved as the last resort option for the treatment of DAVF. Endothelial cell damage and thrombosis are considered to be the main mechanism of DAVF occlusion caused by radiation [51]. Like SRS for the treatment of brain AVM, the occlusion of DAVF may take 1–3 years, and there is still a risk of bleeding during this incubation period [52–54]. When endovascular treatment and surgery fail, it can also be used as a supplementary treatment [52] (Fig. 1.8). When intravascular or surgical methods are too dangerous or fail, SRS can be used for high-risk DAVF [51]. According to reports, 50–93% of DAVFs are completely occluded and treated with SRS [52, 53, 55]. After SRS treatment, the complete occlusion rate and symptom improvement rate of indirect CCF are higher than them of the DAVF of the transverse and sigmoid sinuses [53]. According to reports, the average incubation period of DAVF closure after SRS is 23 months [54]. After SRS of DAVF, the annual rebleeding rate is reported to be as high as 2.6% [55], but this will depend on the initial fistula grade. For follow-up, MRI is recommended once a year, and angiography must be performed to accurately confirm complete DAVF occlusion [52, 53]. Complications include cranial nerve palsy, cerebral edema, latent hemorrhage and radiation effects [52, 53, 55].

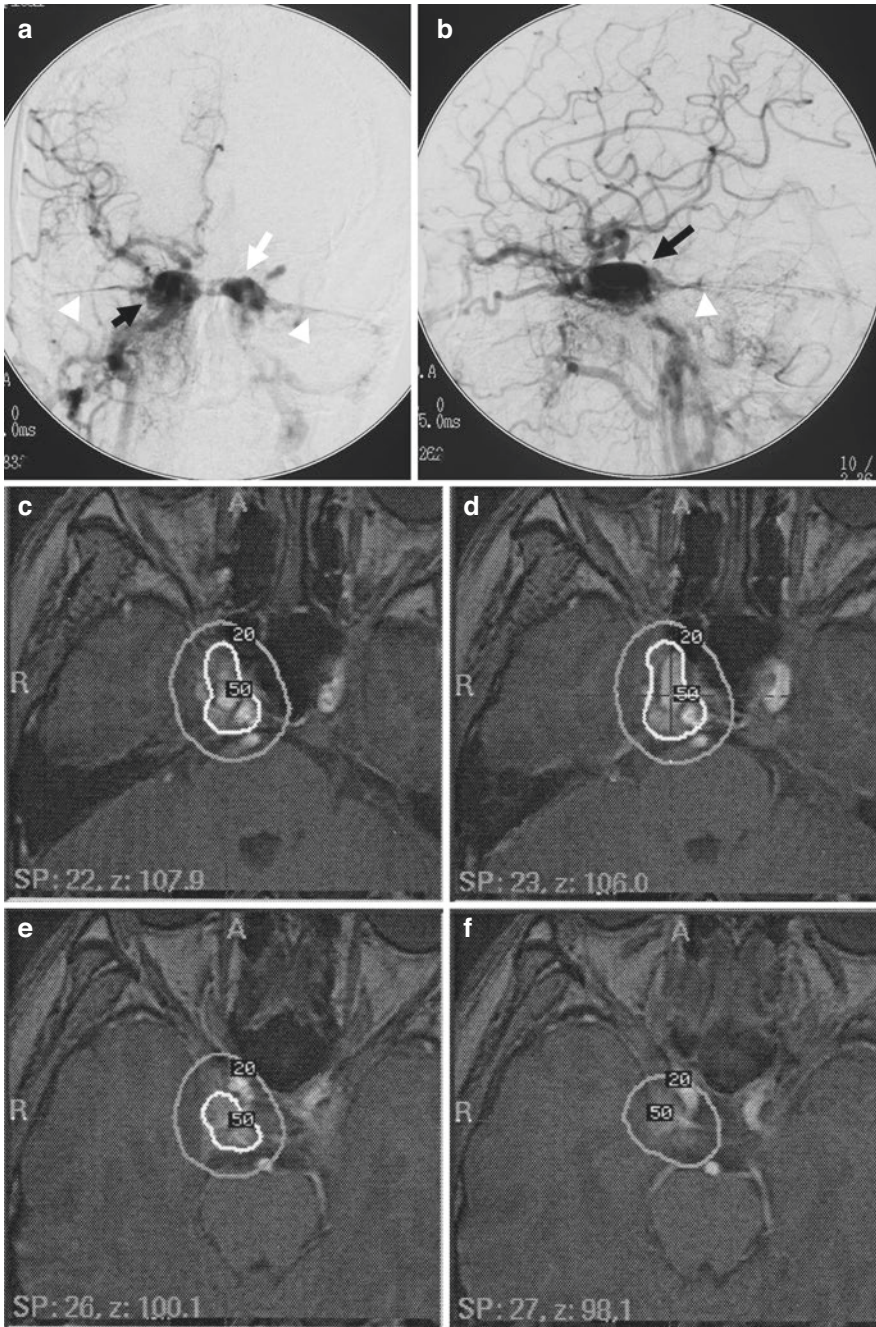


Fig. 1.8 A 53-year-old male patient presented with 2 months of bilateral proptosis. (a) Frontal projection angiogram, right carotid artery injection. (b) Lateral projection angiogram, right carotid artery injection. Showing multiple dural feeders arising from the external carotid artery and the internal carotid artery draining into superior oph-

thalmic vein (black arrow), contralateral cavernous sinus (white arrow) and superior petrosal sinus (white arrowheads). The right Zipfel type II cavernous sinus dAVF was treated with radiosurgery employing a dose of 19 Gy to the 50% isodose line, as seen on axial views of planning MRI T1-weighted sequence with contrast (c-f)

1.7 Surgery

Although endovascular techniques are generally considered the first-line treatment for DAVFs, surgery is still an effective and safe alternative [56]. For a DAVF in the transverse sigmoid sinus, surgery involves extensive exposure of the involved sinus by separating and coagulating the dural arteries and arterialized cortical veins [56]. If the affected sinus is not functional and is not involved in normal brain venous drainage, it can be completely removed. Care must be taken to avoid massive blood loss during surgery, especially in complex fistulas with highly developed and hypertrophic arterial networks. In surgical treatment of non-sinus DAVF, the cortical draining vein is disconnected from the fistula point by clip or coagulation [56, 57]. Frameless stereotactic navigation helps to locate the fistula connection during craniotomy [58]. Occlusion of the DAVF during surgery is usually confirmed by indocyanine green angiography with or without intraoperative DSA [59].

Surgery is usually considered when the endovascular method fails to completely cure the disease, such as such as DAVF in the anterior fossa [60, 61] (Fig. 1.9). According to reports, the long-term morbidity and mortality rate of DAVF surgery is as high as 13% [58]. The main complications of surgery include infection, hydrocephalus, cerebrospinal fluid leakage, stroke, cranial nerve palsy, and severe blood loss [58].

Preoperative embolization before surgery can help reduce blood loss during surgery.

1.8 Endovascular Management Based on Zipfel Classification of DAVFs [13]

Cranial and spinal DAVFs are classified into three types based on Zipfel et al.'s classification scales [13]. Zipfel et al. proposed a modified Borden classification based on natural history data correlating symptoms with outcome in 2009 [13]. The Zipfel classification designates types I, II, and III lesions as those with dural venous drainage without venous reflux, dural venous drainage with venous reflux, and cortical venous drainage, respectively. Zipfel type I lesions exhibit a benign clinical course. Patients harboring type II lesions usually present intracranial hypertension. Hemorrhagic ruptured occurred in more than 30% of patients harboring type III DAVFs. Type II and III spinal DAVFs exhibiting drainage into spinal perimedullary veins might present progressive myelopathy in all of cases. Some spinal DAVF patients characterized by spasticity of muscular hypertonia caused by the upper motor neuron syndrome.

Zipfel type I DAVFs can be treated by TAE and major sinus drainage needs to be preserved (Fig. 1.10). Because of the benign nature, DAVFs without venous reflux (type I) are often managed

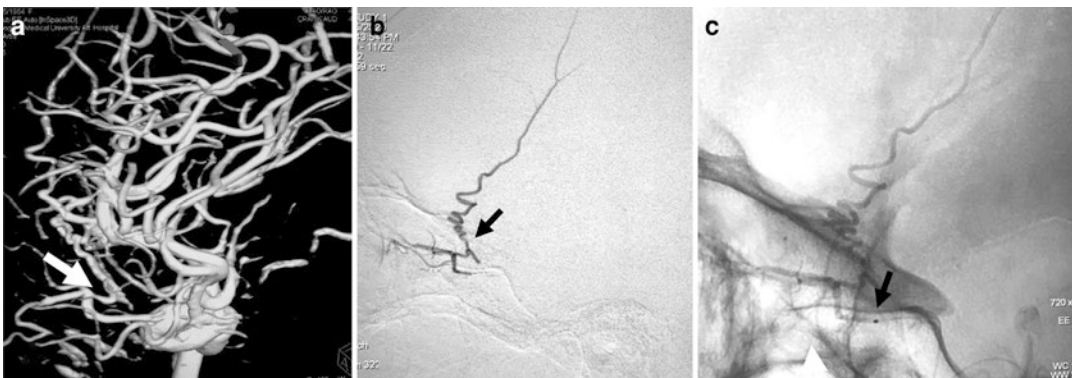


Fig. 1.9 A 54-year-old female patient presented with subarachnoid hemorrhage caused by a Zipfel type III dAVF supplied by the right ophthalmic artery. (a) 3-D reconstruction of the right internal carotid artery injection demonstrated a modified Borden type III dAVF supplied

by the ophthalmic artery (arrow). (b) Lateral super selective injection of the right ophthalmic artery showing the arteriovenous shunt (arrow). (c) Unsubtracted image demonstrated the microcatheter tip (arrow) and the modified Borden type III dAVF in the right anterior cranial fossa



Fig. 1.10 A 38-year-old male patient presented with 3 years of pulsatile tinnitus. Transarterial Onyx embolisation of a left sphenoid parietal sinus dAVF. **(a)** Frontal projection pre-embolisation angiogram, selective the left external carotid artery injection. **(b)** Lateral projection pre-embolisation angiogram, selective the left external carotid artery injection. Showing arterial feeder arising from the middle meningeal artery (arrow), draining into

sphenoid parietal sinus. **(c)** Superselective angiogram showing the microcatheter within the middle meningeal artery (arrow). **(d)** Unsubtracted lateral view showing Onyx cast after single infusion through the middle meningeal artery. **(e)** Final digital subtraction angiography of the left external carotid injection, frontal view **(e)** and lateral view **(f)**, showing no residual dAVF

conservatively. Compression therapy is sometimes used for these DAVFs. Compression of ipsilateral carotid artery or occipital artery is performed by contralateral hand for more than 20 min, three times a day. Contrast to type I DAVFs, which are usually benign, types II and III DAVFs presented a high proportion of worse clinical status before treatment. Their pathophysiology was consistent as venous congestion and hypertension. Type II and III DAVFs can be successfully treated with endovascular treatment. Since being associated with moderate risk for type II DAVFs, endovascular techniques should be used in selective patients (Fig. 1.11). When the venous drainage could be sacrificed safely, the DAVF can be cured by the venous obliteration. Type III DAVFs are usually cured by TAE embolization of the pial draining vein.

Classifications developed by Borden et al. and Cognard et al. have both inadequately distinguish between anterior and posterior drainage in the cavernous sinus region, which is the most common location [62–66]. Cavernous sinus DAVFs drained only inferior petrosal sinus and spinal extradural DAVFs only involving epidural venous plexus is defined as Zipfel type I DAVFs. Type I DAVFs are often asymptomatic or bruits. Cavernous sinus DAVFs that drain retrogradely into the intraorbital/ophthalmic veins with or without reflux to the Sylvian veins are classified as type II DAVFs because the intraorbital veins corresponds to dural sinuses [64] (Fig. 1.12). Spinal DAVFs that drain into the epidural venous plexus with reflux to the perimedullary veins are classified as type II DAVFs because the epidural plexus corresponds to dural sinuses and the intradural perimedullary veins are pial veins. Intra-orbital arteriovenous fistula exhibit an exceedingly low incidence and prevalence, presenting chiefly with proptosis and chemosis and also is classified as Zipfel type II. There are two specific locations, such as the orbit and the spine. The intra-orbital veins are located between the orbital periosteum and ocular nerves sheath dura and the spinal epidural veins are located in the spinal canal between the spinal periosteum and meningeal dura [3]. Thus, the intraorbital venous plexus and spinal epidural veins correspond to the dural sinuses.

Zipfel type III DAVFs are drained directly into pial veins with or without giant venous pouch (Cognard type III and IV and Borden type III) because their natural history and treatment are the same as for other type III DAVFs. In type III patients, the venous pouch can cause subarachnoid hemorrhage, supratentorial ventricular dilatation and trigeminal neuralgia and facial nerve spasm. Cognard type V DAVFs can be Zipfel type II or III as those lesions drained into spinal perimedullary veins indirectly or directly [7, 9] (Fig. 1.13). The most common group of type III DAVFs are tentorial [12, 67] (Fig. 1.14). The most common spinal DAVFs are type III DAVFs supplied by dural branches of a radicular artery and drained into the perimedullary veins [68]. Spinal DAVFs drained into intradural spaces causing myelopathy.

When a DAVF drains into a major dural sinus that cannot be safely occluded, it is often difficult to cure the DAVF, especially if it has multiple fistulas [69]. The hemorrhagic complication risk of a major dural sinus sacrifice combined with the benign natural history of type I or II DAVFs makes this procedure an unwarranted option. When the sinus must be preserved and the arterial supply cannot be completely eliminated, our treatment goal should aim to restore the hemodynamic of brain or spine circulation, e.g. the arteriovenous shunts can be significantly reduced by endovascular embolization.

Type III DAVFs can be fed by vessels that also supply important structures [70]. DAVFs of the tentorium are fed by meningohypophyseal trunk branches of the ICA. DAVFs at the anterior cranial fossa are fed by the ethmoidal branches of the ophthalmic artery. Spinal DAVFs are fed by dural branches of the radicular arteries adjacent to the radiculomedullary arteries. When these arteries are embolized, there are potential risks of neurological deficits. Inadvertent embolization of the cavernous and petrosal branches of the middle meningeal artery, which triumphantly emerges into the middle cranial fossa through foramen spinosum, may precipitate infarctions of the trigeminal and facial nerves [71–73]. As to the cavernous sinus DAVFs, the cause of hemorrhagic or venous infarction complications are

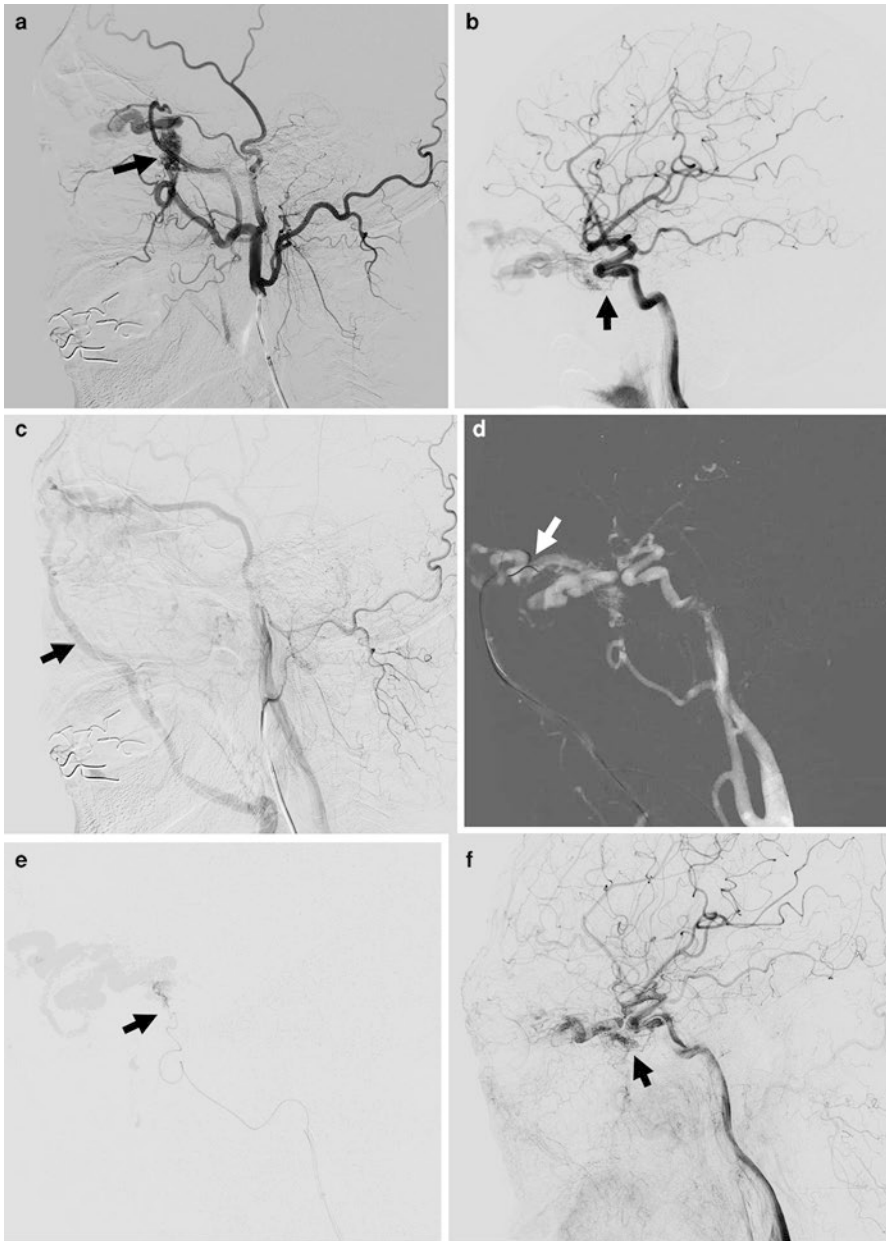


Fig. 1.11 A 45-year-old male patient presented with congestion of the right bulbar conjunctiva, exophthalmos, and right palpebral edema. **(a)** Arterial phase of a lateral view on selective right external carotid arteriography before treatment. Sphenopalatine and infraorbital arteries branched from the maxillary artery and the middle meningeal artery (arrow), exhibiting inflow to the shunt site via aggregated blood vessels. The entire shunt was visualized from the artery of the superior orbital fissure. Outflow to the ophthalmic veins was observed. **(b)** Arterial phase of a frontal view on right internal carotid arteriography before treatment. A portion of the shunt was visualized from small dural branches (arrow) from the cavernous segment of the internal carotid artery. **(c)** Venous phase of a lateral view on right

carotid arteriography before treatment. The main drainers outflowed to the front are superficial temporal vein and facial vein (arrow) mediated by the angular vein. **(d)** Under roadmap, the transvenous treatment through the facial vein to the superior ophthalmic vein was attempted, but failed to access the fistula site because of the tortuous angular vein (arrow). **(e)** Selective angiography through a Marathon microcatheter guided to the area adjacent to the shunt at the periphery of the artery of the superior orbital fissure (arrow). The shunt and drainer were visualized. **(f)** Late phase of a lateral view on the right carotid arteriography after surgery showing a small residual arteriovenous shunt from the small dural branches (arrow) from the cavernous segment of the internal carotid artery

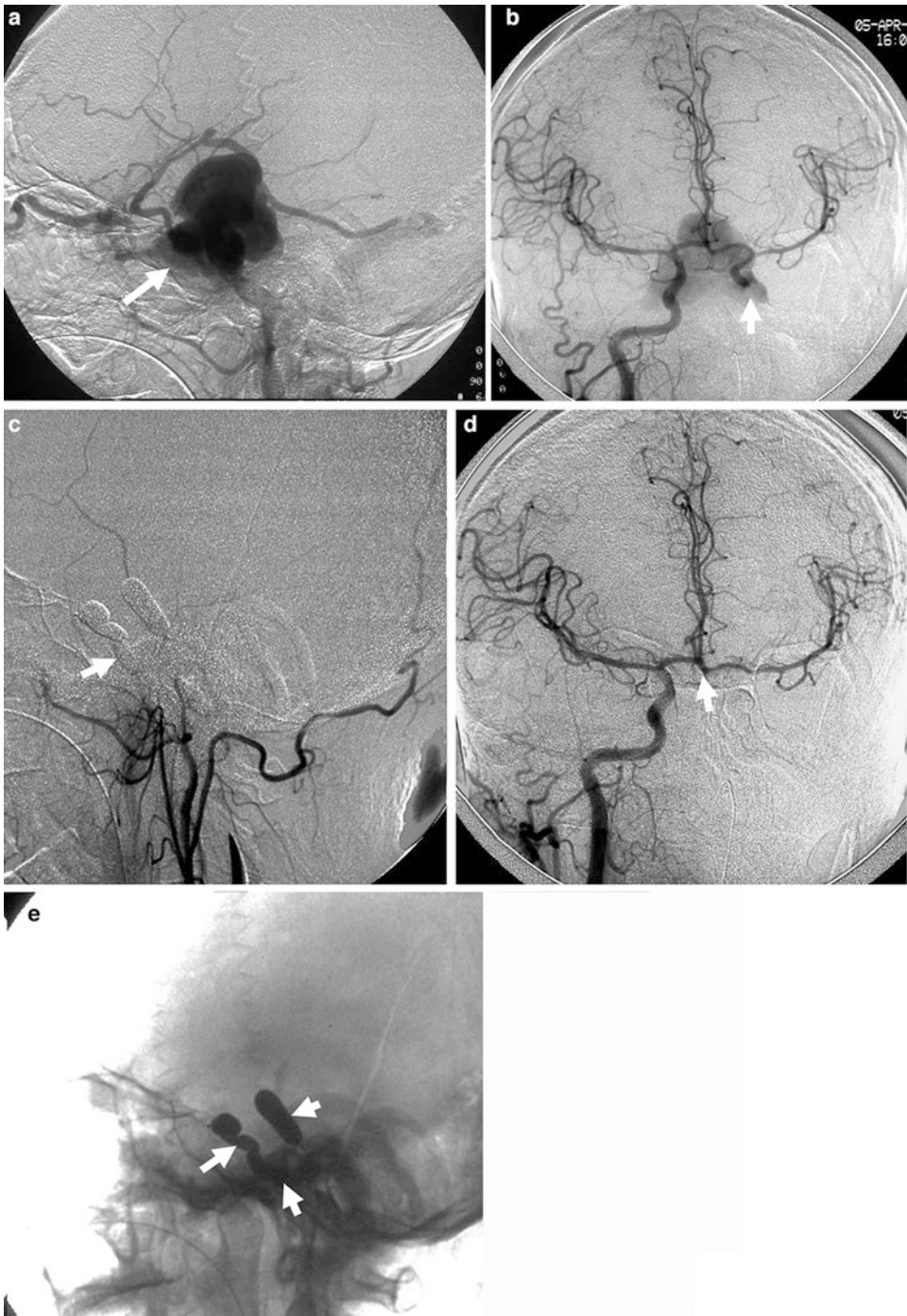


Fig. 1.12 A large carotid-cavernous fistula was treated with detachable balloons. (a) Left carotid artery angiogram (lateral view) showing a Zipfel type III carotid-cavernous fistula supplied by the internal carotid artery (arrow). (b) Right carotid artery angiogram (frontal view) showing the blood flow reflux from the contralateral

carotid artery circulation (arrow). (c) The carotid artery angiogram (lateral view) showing the internal carotid artery and fistula was complete occluded (arrow). (d) Control angiogram (frontal view) showing there was no blood flow to the dural fistula. (e) Unsubtracted image (lateral view) showing the balloons

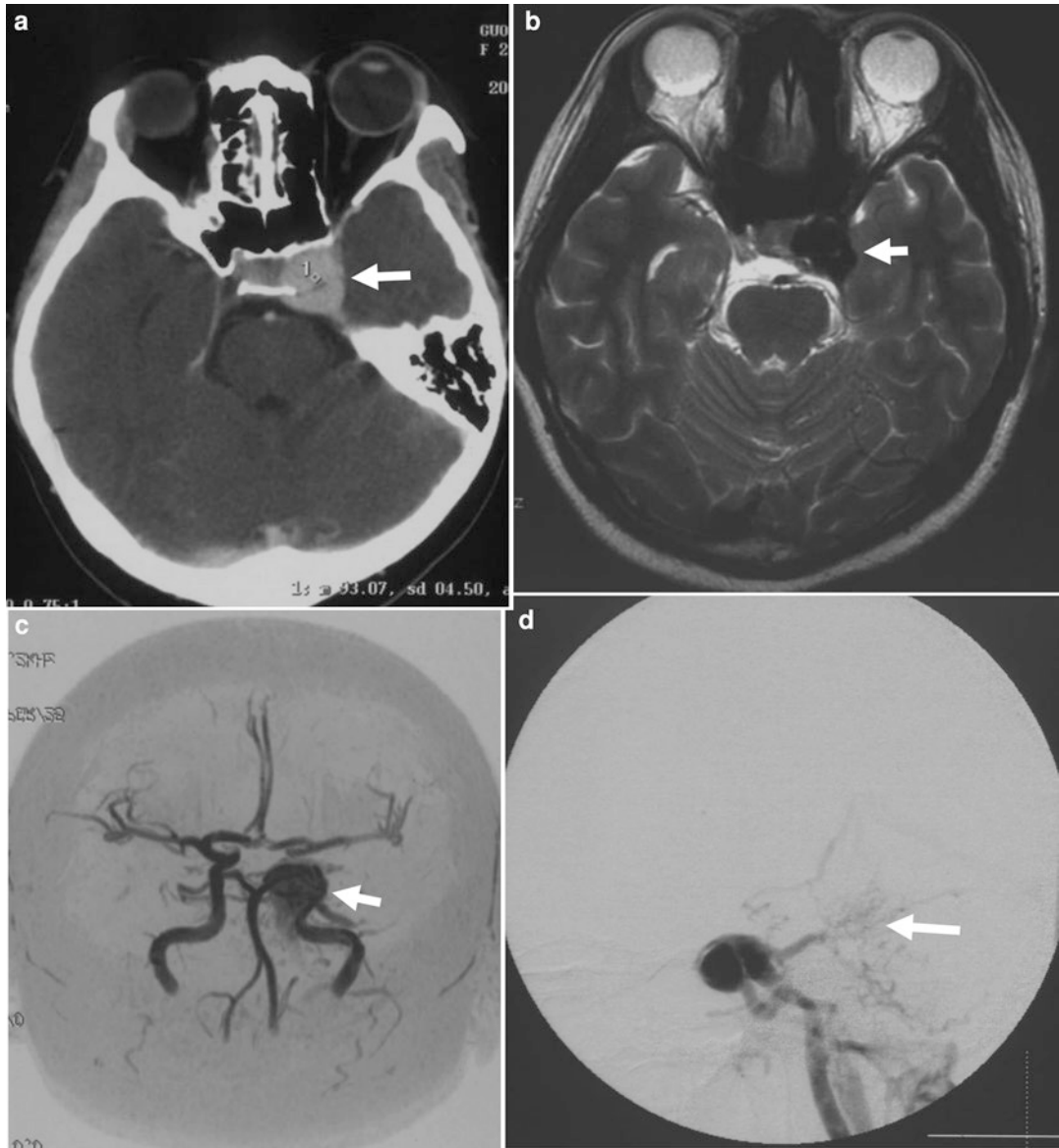


Fig. 1.13 A 24-year-old female patient presented with the left oculomotor nerve palsy and intracranial bruits. **(a)** Cranial CT scanning showing a hyperdensity lesion at the left parasellar location (arrow). **(b)** MR imaging showing a flow void signal at the left parasellar location (arrow).

(c) MR angiography demonstrating engorged left cavernous sinus. **(d)** Left carotid artery angiogram (lateral view) showing a Zipfel type III carotid-cavernous fistula supplied by the internal carotid artery drained to brain stem and cerebellar veins

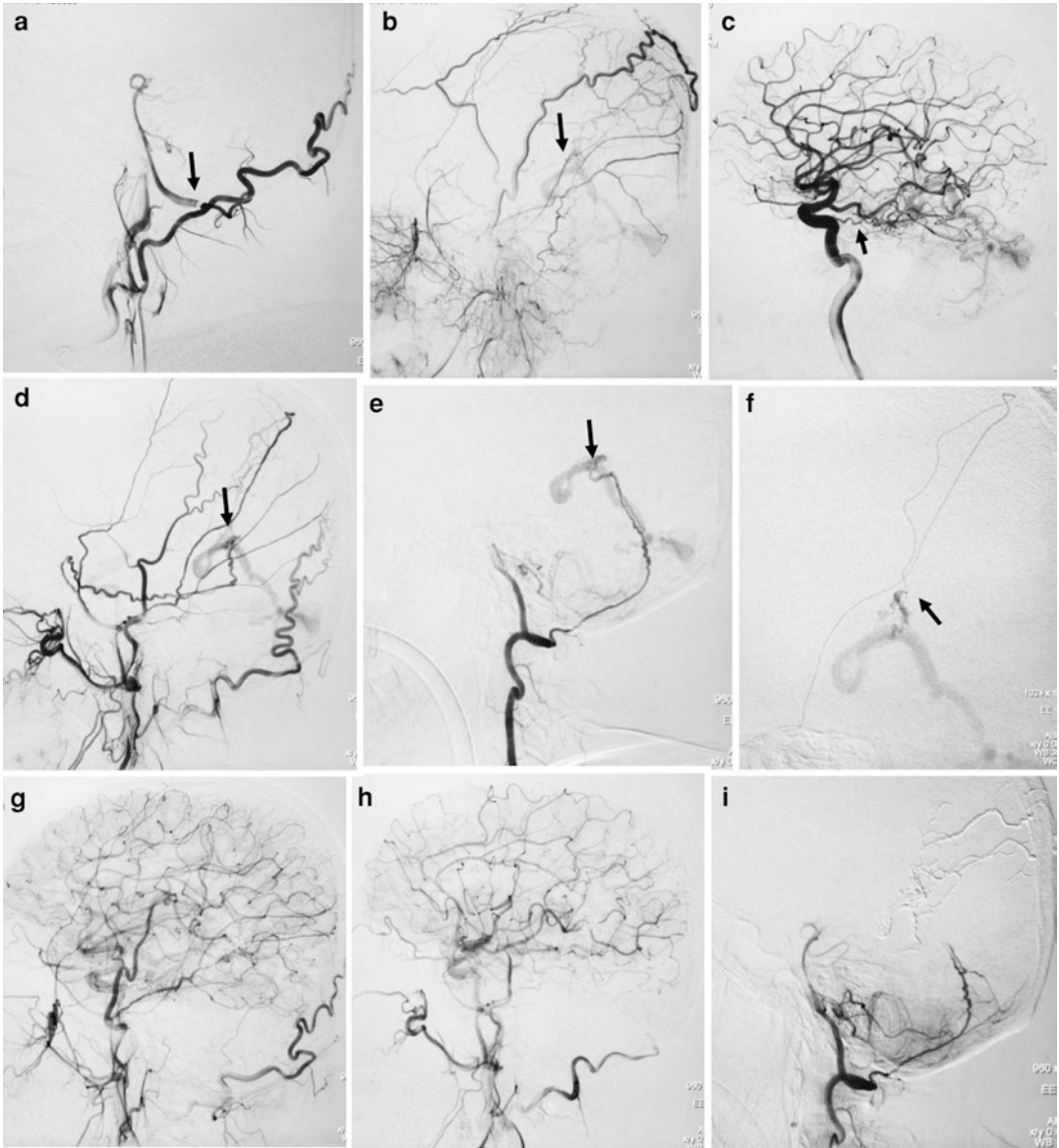


Fig. 1.14 A 53-year-old male patient presented with dizziness. (a) Lateral projection pre-embolisation angiogram, selective right external carotid artery injection, showing the occipital artery anastomosed to the vertebral artery with the transosseous vessel (arrow). (b) Late phase of the lateral projection pre-embolisation angiogram, selective right external carotid artery injection. (c) The lateral projection pre-embolisation angiogram, selective right internal carotid artery injection. (d) The lateral projection pre-embolisation angiogram, selective left external carotid artery injection. (e) The lateral projection

pre-embolisation angiogram, selective left vertebral artery injection. Showing multiple arterial feeders arising from the bilateral middle meningeal artery (arrow), right meningohypophyseal artery (arrow) and left posterior meningeal artery (arrow), draining into a common channel (arrow). (f) Selective injection showing the Marathon microcatheter positioned within the arteriovenous shunt (arrow). Postembolisation lateral angiograms, (g) right carotid artery injection, (h) left carotid artery injection and (i) left vertebral artery injection showing no residual dAVF

attributed to the venous perforations or hemodynamic changes in direction of venous drainage [65, 74, 75]. Among patients harboring pial artery supply, giant venous pouch and high volume Onyx injection in one session constituted the chief causes of hemorrhagic complications [76]. When collateral arteriovenous shunts exist between dural and pial arterial suppliers, the injection force can cause Onyx reflux across these collateral vessels into the brain, which requires special attention during DAVF embolization.

1.9 Conclusions

Endovascular treatment is usually the first-line treatment of DAVF. Before intervention, it is necessary to have a complete understanding of the vascular structure of the fistula. This includes identifying blood supply arteries, fistula connection points, venous drainage pathways, and venous flow direction. According to the location and anatomical structure of the fistula, the intravascular approach is carried out from the artery, vein or combined access. Surgery and SRS remain as alternative treatment options, especially when endovascular methods are unsuccessful or considered dangerous. Follow-up angiography is needed to confirm the long-term occlusion of DAVF and the durability of treatment. The Borden and Cognard classifications do not explicitly cover cavernous DAVF, which is a very common type of DAVFs. In this study, based on the understanding of vascular structure and natural history of DAVFs, the Zipfel classification was proposed to guide the proper evaluation of DAVF and the treatment of these lesions.

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