

Intracranial Dural Arteriovenous Fistulas: The Sinus and Non-Sinus Concept

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Introduction

Dural arteriovenous fistulas (dAVFs) represent 15% of all intracranial arteriovenous shunts. In particular, 7% of supratentorial and 35% of infratentorial shunts are dAVFs [1]. Dural arteriovenous fistulas are a group of acquired pathological vascular malformations, defined by an abnormal communication within the dural leaflets, between arteries and dural venous sinuses and/or subarachnoid veins. The feeding arteries are commonly branches of the external carotid artery, tentorial branches of the internal carotid artery, meningeal branches of the vertebral artery or, rarely, pial branches of the cerebral arteries [2]. Dural arteriovenous fistulas are more commonly supratentorial than infratentorial in location. The transverse-sigmoid sinus junction is the most common location for dAVFs, with a slight left-sided predominance. Through the years many classification systems have been proposed for intracranial dAVFs. These systems are based on the lesion's venous drainage patterns, as this factor dictates the behavior of the lesion itself. Djindjian and Merland first classified dAVFs according to their venous angioarchitecture in 1977 [3]. In 1995, Cognard further classified both cranial and spinal arteriovenous fistulas according to their venous outflow with prognostic and treatment implications [4]. Borden simplified the Cognard classification, emphasizing that the major factor in predicting an aggressive clinical course is the presence of cortical venous drainage. Unlike venous sinuses, cortical veins are not protected by the dura and cannot withstand arterial pressures. Therefore, dAVFs with cortical venous drainage (Borden types II and III) have a higher risk of rupture and hemorrhage [5, 6].

The Vascular Malformations Study Group of Toronto identified in 1997 the same rate of bleeding for dAVF and cerebral AVMs. Intracranial hemorrhage and neurologic deficit is likely in 2% of the Borden classification type I, 39% of type II, and 79% of type III dAVFs [7]. However, there is a 45% mortality rate over 4 years among patients with cortical venous drainage, a 19.2% intracranial hemorrhage rate per year, and 10.9% new neurologic deficit rate per year. For patients who present with hemorrhage, the rate of repeated hemorrhage is 35% within the first 2 weeks of the initial ictus [1, 6, 8-10]. According to our experience [11-13], we suggest a simplified classification of dAVFs: sinus and nonsinus. This classification guides the choice of surgical treatment. As previously observed by other authors, posterior fossa localization of the fistulas is not a real adjunctive riskfactor [14].

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Our Experience

Sinus Fistula: We define as sinus dAVF all the fistulas having the dural arteries shunting directly into dural sinus. This direct communication between the artery and the sinus can subsequently recruit cerebral veins. The classification for this type of fistula, despite venous recruitment, does not change and still represents a sinus fistula (Fig. 1a).

Non-Sinus Fistula: In non-sinus fistula the sinus maintains functional for the venous discharge of the brain. The pathological shunt is embedded into the dural leaflet without communication with the sinus. Drainage of the fistula depends entirely upon cerebral veins (Fig. 1b).

SINUS FISTULA

NON SINUS FISTULA

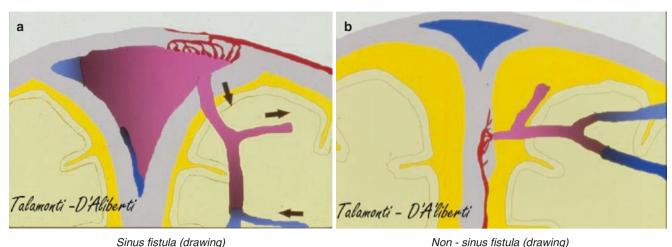


Fig. 1 (a) Sinus fistula (drawing). (b) Non-sinus fistula (drawing)

From 2006 to 2016, at our Neurosurgical Department and our Interventional Neuroradiology at Niguarda Hospital, Milan, Italy, 477 intracranial dural arteriovenous fistulas have been treated: 376 underwent endovascular treatment and 101 underwent surgical treatment. Cavernous sinus dAVFs and Galen ampulla malformations have been excluded from this series, as they represent a different pathology per se. Of the 376 dAVFs treated by the endovascular approach, 180 were sinus and 179 were non-sinus. Of the 101 dAVFs treated with the surgical approach: 15 were sinus and 86 were non-sinus.

This classification was found to be very useful in our daily practice. In a sinus fistula there is a direct communication between the arterious dural branch and one dural sinus, and cerebral veins can subsequently be recruited. These recruited veins, called by some authors "red veins" due to the inverted internal blood flow, are not functioning veins. Red veins must be excluded from the circulation together with the excision of the portion of sinus interested by the fistula. In non-sinus fistula, the shunt is embedded into the dural leaflet and it doesn't communicate with any sinus, whereas it drains itself into a cortical vein. Therefore, the sinus is functional for the brain. This draining vein—a red vein—has to be occluded proximally ("piè" of the vein) through a surgical or endovascular transvenous approach.

Correct individuation of the type of fistula, sinus or not sinus, is important in order to plan a proper surgical treatment, regardless of the presence of venous varices in the malformation. Those varices, in fact, do not represent an anatomopathological variant and do not require a different surgical approach [13].

The draining vein in a non-sinus fistula, with one or more associated varices on its course, so-called Type IV of some classification systems [4], does not configure a different type

of fistula in terms of another anatomopathological entity, so it is not an adjunctive surgical risk factor. Therefore, surgical treatment of both infratentorial and supratentorial fistulas is the same: clipping or occlusion of the vein at its emergence (See "Illustrative Cases": Fig. 2, Cases 1 and 2)

Illustrative Cases

Case 1 A 65-year-old man come to observation for ictal onset of headache and coma. At admittance in ER, he presented with GCS 3 and anisocoria (> right), onset <1 h. A CT scan showed a left temporal ICH and a CT-Angio with 3D reconstruction showed a non-sinus dural fistula of pterional dura (a-c). The patient underwent an urgent surgery, with carotid exposure at neck. A wide craniotomy was performed, the hemorrhage was drained, and non-sinus fistula was excluded through multiple clipping of drainage vein, after temporary (7 min) occlusion of external carotid artery (d). The postoperative stay was good, a postoperative DSA showed the complete exclusion of dAVF, and a postoperative CT scan showed the results of previous hemorrhage (e-f). At 2 months follow-up the patient was able to walk alone and he had just a slight expressive dysphasia.

Case 2 A 45-year-old female came to our ED for progressive onset of nucal headache and instability. A first CT scan showed a right, small intracerebellar hemorrhage (g). Six hours after admittance, the patient complained of a new headache attack and the onset of a coma. A new CT scan showed a new hemorrhage and a DSA showed a falco-tentorial nonsinus fistula with associated varix on the drainage vein (h, i, l). The patient underwent urgent surgery with drainage of

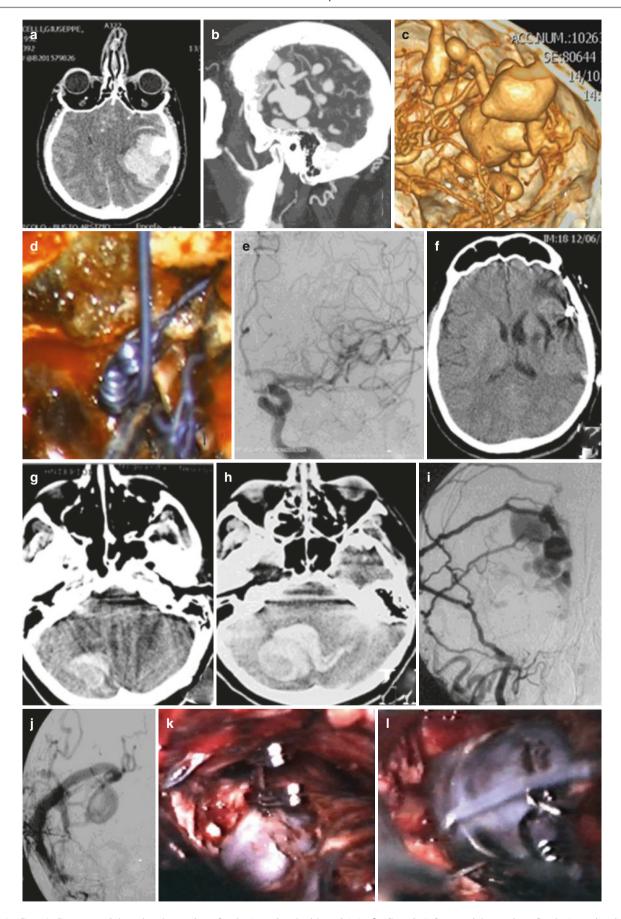


Fig. 2 Case 1. Supratentorial pterional non-sinus fistula (associated with varix) (\mathbf{a} - \mathbf{f}). Case 2. Infratentorial non-sinus fistula (associated with varix) (\mathbf{g} - \mathbf{n})

hemorrhage and clipping of "piè" of the vein. Varix was excised with the hemorrhage (m, n). Postoperative stay was good with a prompt recovering of consciousness. The patient was discharged after 2 weeks with a slight instability.

The different surgical approach in FAVD next to a venous sinus depends on the type of the shunt and it doesn't depend on the localization but on type of drainage and on the recruited cerebral veins. In the case of a sinus fistula, therefore, it is possible the resection of the fistulous sinus tract and the occlusion of the recruited nonfunctional veins and the absolute preservation of the veins still functional to the brain (See "Illustrative Cases": Fig. 3, Cases 3 and 4).

Case 3 A 48-year-old male came to our observation for an intracranial hypertension syndrome with slow onset (some months before), with headache and progressive cognitive impairment. Some days before the patient complained of a severe impairment of awareness and vomiting with associated visual impairment. A DSA showed a sinus fistula of the torcular with engorgement of straight sinus and severe recruitment of many cerebral veins, which seemed to persist long because of engorgement (a–c). The patient underwent a surgical resection of torcular with occlusion of recruited veins (red veins), both not functional for the brain. Postoperative DSE showed exclusion of the FAVD and the non-visualization of straight sinus in arterial phase, for the restoring of a normal situation (d, e).

Case 4 A 17-year-old male with a slow onset symptomatology (6 months) with headache and visual blurring. A DSA showed a sinus FAVD of the middle third of SSS with recruitment of cerebral veins (f-h). The anterior one is not functional; the posterior one, instead, remains normally visible in venous phases of angiography when dAVF is not visible. The patient underwent surgical resection of fistulous sinus tract with occlusion of anterior recruited vein (i, l, m) and avoidance of the posterior one, functional for the brain. The postoperative DSA shows a normal venous angiogram (n).

Different types of dAVFs (sinus or non-sinus) in posterior cranial fossa may localize in the same place, but they require different surgical treatment, in particular those located at the petrous bone apex (See "Illustrative Cases": Fig. 4, Cases 5 and 6). Finally, non-sinus fistulas with associated varix and perimedullary drainage, so-called Type V of Cognard-Gobin Classification [4], are not a different type of fistula and they do not add an adjunctive surgical risk factor [15] (See "Illustrative Cases": Fig. 5, Cases 7 and 8).

Case 5 A 44-year-old female fashion designer come to our observation for a progressive brainstem symptomatology. An MRI showed a right ischemic lesion in the pons. Three years before, she was treated in another institution with embolization of a sinus transverse-sigmoid dAVF with an initial recruitment of the superior petrous sinus (a–c). A new DSA showed sinus recanalization associated with an evident recruitment of the superior petrous sinus and reflux in engorged infratentorial veins (d, e). The postoperative angiography showed a disappearance of sinus fistula after sinus removal and clipping of main recruited vein (f). After 6 months of swallowing rehabilitation the patient was able to go back to her job.

Case 6 A 48-year-old male journalist come to our observation for a progressive tetraparesis with sphincter disorders. An MRI showed a severe cervical spine myelopathy, and because of the presence of a disk herniation at C4–C5, initial diagnosis was spondylotic cervical myelopathy. Because of the presence, in MRI, of vascular structures in cervical subarachnoid space (g), a second opinion suggested a complete angiographic study of the brain and medulla. The latter showed a non-sinus fistula of the apex of the right petrous bone, with feeder from the Bernasconi-Cassinari artery and drainage in a perimesencephalic vein with associated engorgement of perimedullary veins (h, i, 1). The patient underwent surgical intervention through a suboccipital craniotomy and clipping of drainage vein at the apex of the petrous bone (m, n). Postoperative DSA showed the complete exclusion of the dAVF (o). After 6 months of rehabilitation, the patient went back to work autonomous life and with slight sphincter with impairment.

Case 7 A 52-year-old male presented with a progressive myelopathy, and MRI showed a cervical spine myelopathy without a cervical spondylosis and presence of vascular structures in posterior subarachnoid space (a). The patient underwent complete DSA of brain and medulla, which showed a non-sinus fistula on the dura close to the entrance into intracranial space of the left vertebral artery and engorgement of perimedullary veins of the cervical tract (b). Through a far lateral approach, the "piè" of the vein was exposed in the anterior part of the intracranial entry point of vertebral artery (c, d). The malformation was excluded through clipping of the drainage vein at its origin. Postoperative DSA showed the complete exclusion of the dAVF (e). At 5 months follow-up, the patient walked with aid and still had sphincter disorders.

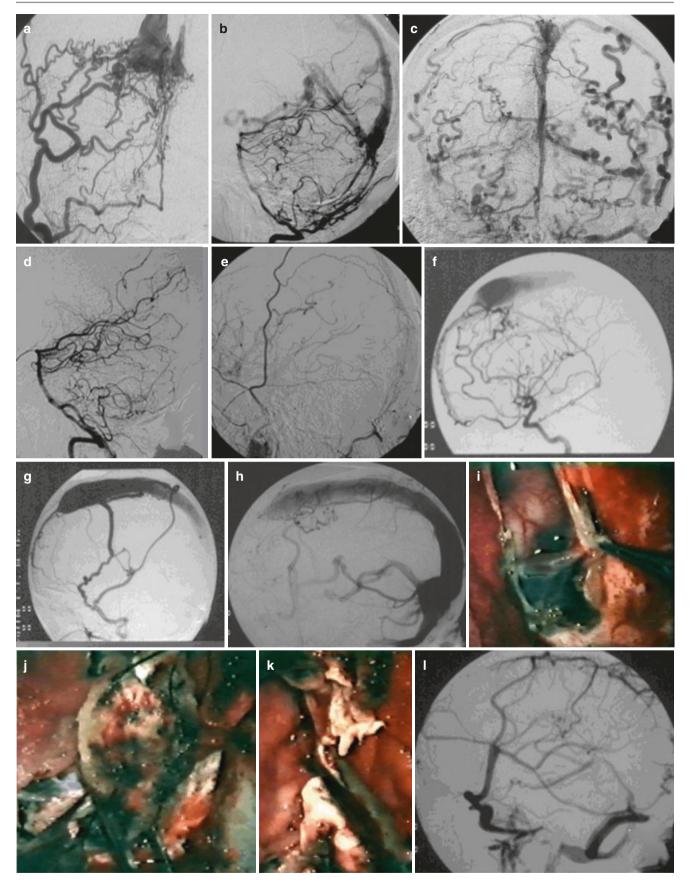


Fig. 3 Case 3. Sinus Fistula of Torcular (a–e). Case 4. Non sinus fistula SSS (f–n)

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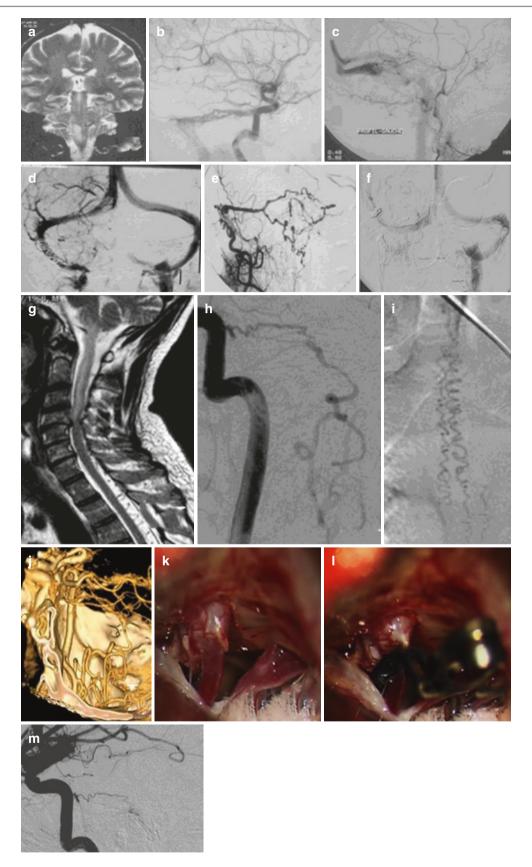


Fig. 4 Case 5. Sinus Fistula of Petrous bone apex (a–f). Case 6. Non-sinus fistula of Petrous bone apex (g–o)

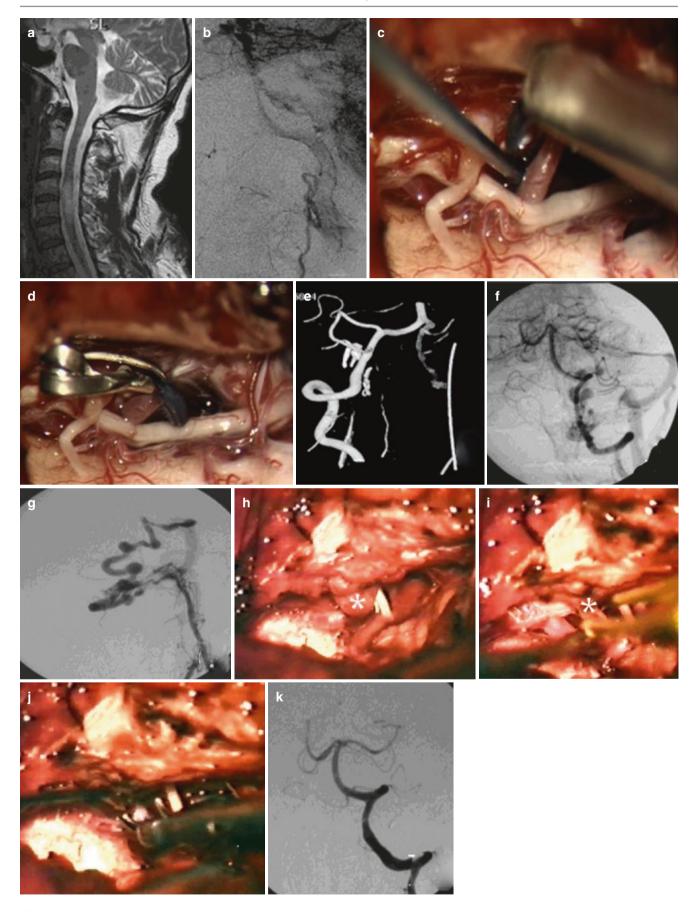


Fig. 5 Case 7 Non-sinus Fistula of Foramen Magnum (a–e). Case 8 Non-sinus fistula of Jugular Foramen (f–m)

Table 1 Surgical results

Mortality	2/101 (2%)
Severe morbidity	1/101 (1%)
Minor morbidity	4/101(4%)
Complete obliteration	101/101 (100%)
F.U 5 years: recurrence	1/101 (1%)

Case 8 A 58-year-old Italian politician come to our observation for a left tinnitus with onset 1 year previous. Some weeks before, the patient experienced onset of progressive motor impairment in walking with sensitive deficit. Angiography showed a non-sinus fistula close to the jugular foramen (f, g). The fistula was excluded through a surgical intervention with clipping of "piè" of the vein (h, i, l). Postoperative DSA showed the complete disappearance of the malformation (m).

Surgical Results

Surgical series results are described in Table 1. Mortality and severe disability was 3% and morbidity less than 4%. All patients underwent a postoperative DSA with nearly 100% of complete occlusion of the fistula. At a mean follow up of 5 years in one case there was a non-sinus fistula recurrence, due to the presence of a partial clipping of "the shunting foot." Our surgical experience underlines the fact that posterior fossa localization, per se, does not represent and adjunctive risk factor for treatment. Moreover, based on shunt location, dAVFs can be classified in the same way as meningiomas: falcotentorial, straight sinus-vein of Galen-torcular, transverse and jugular sinus, CPA and foramen magnum (before or at the entrance of the vertebral artery).

Discussion

Dural arteriovenous fistulas (dAVFs) account for 10–15% of all intracranial arteriovenous lesions. Symptoms and prognosis are highly variable. Some dural AVFs produce neurological symptoms depending on the locations and the involved structures; others are associated with intracranial hemorrhage [1, 7, 16]. It is very important to have a good understanding of the natural history of dural AVFs when it comes to decision-making about treatment (See Fig. 6). This challenging and interesting topic has been reviewed since 1984, when Malik et al. [17] studied 223 previously reported cases and concluded that lesions related to large dural sinuses are less likely to bleed than lesions with restricted dural outflow. In this first review there were no angiographic features, in particular, the pattern of venous drainage taken into consideration. In 1986, Lasjaunias et al. [18] presented a meta-

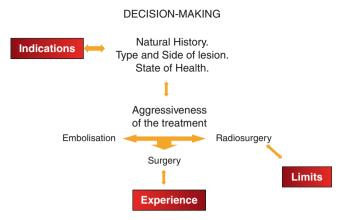


Fig. 6 Decision-making workflow

analysis of 191 cases. They analyzed the mechanism of neurologic manifestations and concluded that apart from the peripheral cranial nerve palsy due to arterial steal phenomena, central nervous system symptoms seem to be related to passive venous hypertension. In 1990, Awad et al. [19] reviewed 360 cases reported in the literature and 17 of their own cases to compare the angiographic features of 100 aggressive cases and 277 benign cases. They concluded that leptomeningeal venous drainage, variceal or aneurysmal venous dilatations, and galenic drainage were indicative of possible aggressive neurologic signs. In 1995 Cognard et al. [4] reviewed a series of 205 consecutive patients with dural AVFs over 18 years. The purpose was to complete and validate the classification of dural AVFs proposed in 1978 by Djindjian et al. [3]. In the same year Borden proposed his classification system [5], which stratifies lesions on the basis of the site of venous drainage and the presence or absence of cortical venous drainage. Borden type I lesions have the direct communication of meningeal arteries with a meningeal vein or dural venous sinus and exhibit normal antegrade flow. Type II lesions have shunts between the meningeal arteries and dural sinus, with retrograde flow into the subarachnoid veins, causing venous hypertension. Type III lesions have direct drainage of meningeal arteries into subarachnoid veins or an "isolated" sinus segment. The latter phenomenon is often the result of thrombosis on either side of the arterialized sinus segment, which directs retrograde flow into the subarachnoid venous system. The Borden classification scheme further subclassifies lesions as single-hole (a) or multiple-hole (b) fistulas. In our experience, the most useful classification for a merely surgical point of view is to distinguish intracranial dAVFs as sinus or non-sinus, because the simple understanding of this point leads to the correct treatment. On the basis of venous drainage, Borden [5] classified intracranial dAVFs into three main types: sinusal, sinusal with recruitment of one or more cerebral veins, and fistulas draining into cortical cerebral veins.

Commonly, type I fistulas have a benign behavior and are conservatively treated. If a disabling bruit is present, a palliative transarterial embolization can be indicated, while surgery is chosen in very selected cases.

However, Type II and III dAVFs usually show an aggressive behavior including hemorrhage and progressive neurological deficits. Davies [1, 7] report an intracranial hemorrhage rate of 19.2% lesion/year and non-hemorrhagic deficit of 10.7% lesion/year. The complete obliteration of Type II or Type III fistula by embolization can be obtained in less than 50% of cases via the transarterial route and in more than 80% of cases when the venous route is used.

Urtasun, Roy, Link and Lucas [20–23] reported rates of 72% complete obliteration and 28% partial obliteration in patients treated by radiosurgery at 1 and 3 years of follow-up. On the basis of shunt location, posterior cranial fossa dAVFs can be classified in the same way as meningiomas: falcotentorial, straight sinus/vein of Galen, torcular, transverse sinus, jugular sinus, CPA, and foramen magnum dAVFs (Fig. 7). From a practical surgical point of view we

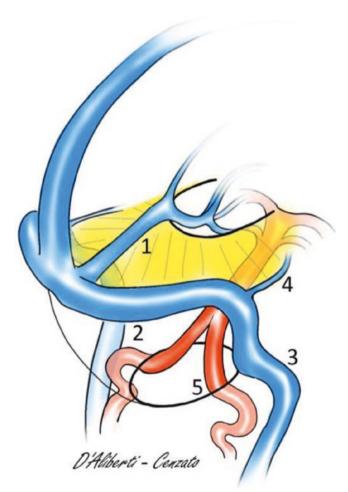


Fig. 7 Localization of posterior cranial fossa dAVFs (See Table 2 for description)

suggest to simply classify dAVFs as "sinusal" or "non sinusal fistula." In the former type there is a direct communication between the dural shunt and one sinus, and cortical veins can sometimes be recruited; in the latter type the shunt is embedded into the dura and the drainage always involves a cerebral vein. As for all diseases, the decision-making process must take into account surgeon experience, aggressiveness of the lesion, and limits of the treatment. Three different approaches can be indicated in dAVFs: surgical, endovascular (i.e., transarterial/transvenous embolization), and radiosurgical with Gamma Knife.

Surgical treatment is different in sinus and non-sinus fistulas and comprise involved sinus resection and clipping or endovascular occlusion of recruited cerebral veins. Recruited veins, the so-called "red veins," are not functional and show an inverted internal blood flow. Distal to the involved sinus there could be functional blue veins that must be spared. On the other hand, treatment of non-sinusal fistula consists in clipping or endovascular occlusion of the draining red vein which, as mentioned before, is a non-functional vein.

Conclusions

We want to underline the role of venous drainage in the management of dAVFs. Prioritization of treatment options is crucial to achieving a good result. As a consequence, identifying sinus vs non-sinus fistulas is crucial from a surgical point of view. Sinus fistulas require sinus excision and clipping and subsequent occlusion of recruited veins. Nonsinus fistulas require, on the other hand, clipping or endovascular venous embolization of the drainage vein. A posterior cranial fossa location does not represent an adjunctive risk factor in the treatment of dural fistulas. Type IV and V in Cognard and Gobin's classification are not to be considered as independent entities: varices and perimedullary drainage can be seen in both sinus and non-sinus fistulas.

Table 2 Localization of posterior cranial fossa meningiomas and dAVFS

Localization of posterior cranial	Localization of posterior cranial
fossa meningioma	fossa non sinus-sinus DAVFs
(Castellano-Yasargil-AlMefty)	(D'Aliberti-Cenzato)
Tentorial-Falcotentorial	1. Straight sinus, Vein of Galen and Torcular
Petrous-Petroclival	2. Trasverse Sinus
Jugular Foramen	3. Jugular Foramen
CPA	4. Petrous bone apex
Foramen Magnum	5. Foramen Magnum (Before or at the entrance of VA)

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