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Vertebral-Venous fistulas: Single center experience and practical treatment approach

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Abstract

Background: Vertebral-venous fistulas (VVFs) are rare. Scarce literature exists to guide our understanding and management. We report our experience and propose a classification based on flow, feeder number, and involvement of accessible veins. Additionally, we include a practical treatment approach.

Methods: Retrospective chart and imaging review of cerebrovascular arteriovenous fistulas treated in our center between July 2013 and April 2022. We reviewed patient demographics, presentation, imaging, treatment strategies, and outcomes.

Results: Nine patients with VVFs were identified, six were females. Ages ranged between 38–83 years. There were six high-flow and three low-flow. Most VVFs originated at the level of V3. Additional feeders from the internal carotid artery, external carotid artery, and/or subclavian artery were present in four cases (two were high-flow). Four cases had multiple arterial feeders. All cases were symptomatic. Origin was spontaneous in eight and iatrogenic in one case. Most common presenting symptoms were pain (7) and pulsatile tinnitus (4). Neurological deficits were present in two cases (1 high- and 1 low-flow). Four cases were treated with vertebral artery segmental sacrifice alone, three required multiple transarterial embolizations with or without VA sacrifice, one case had single transvenous approach, and one was treated with single targeted transarterial embolization. One patient had a minor transient neurological complication. No treatment-related mortality was seen.

Conclusion: Treatment of high-flow and symptomatic low-flow VVFs is feasible and safe. Our classification and treatment approach might help guide patient selection and choice of endovascular approach. However, our approach warrants further validation with a larger number of patients.

Keywords

Arteriovenous fistula, vertebral artery, cerebrovascular, neurosurgery

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Abbreviations: CTA, computed tomography angiography; ICA, internal carotid artery; IJV, internal jugular vein; MRI, magnetic resonance imaging; mRS, modified Rankin Scale; PICA, posterior inferior cerebellar artery; VA, vertebral artery; VVF, vertebral-venous fistula; VVP, vertebral venous plexus

Background

Vertebral-venous fistulas (VVFs) are rare. There is no consensus in the literature about the definition and nomenclature. Vertebral-vertebral fistulas and vertebral arteriovenous fistulas are other reported names used interchangeably with VVF.[1–6] They are defined by the presence of an abnormal connection (without intertwining capillaries) between the vertebral artery (VA) or one of its branches and an adjacent vein.[2,3,5–7] The most extensive systematic review on the subject defines them as a fistulous connection between the VA (or one of its branches) and the vertebral venous plexus (VVP).[1] However, despite the narrow definition that the authors used, they, in effect, included all reported VVFs that had drainage to surrounding veins, including the VVP, internal jugular vein (IJV), and vertebral veins.[1,6,8–10] Additionally, arterial contribution through the internal carotid artery (ICA), branches of the external carotid artery, and cervical arteries (deep or ascending) have also been observed.[3,6,11]

VVFs can arise spontaneously or be a product of iatrogenic injury or trauma.[1,7,8,12] Spontaneous VVFs are thought to be caused by microtrauma secondary to activities

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such as sneezing or sudden neck movement. Moreover, they can be associated with conditions such as neurofibromatosis or fibromuscular dysplasia.[1,11,13–18] The clinical presentation of VVFs is variable, ranging from asymptomatic to mildly symptomatic, and in the worst cases, with significant neurological impairment caused by venous congestion, compression, and/or ischemic events.[1,3,19]

We aimed to evaluate the clinical course, radiological findings, endovascular management, and treatment outcomes of patients with VVFs treated at our center. In addition, we developed an anatomical classification system and a practical treatment approach.

Methods

Patient selection and data collection

After approval by our Institutional Review Board, we retrospectively reviewed the charts of patients diagnosed with a VVF at our center between July 2013 and April 2022. Patients were identified by entering the keywords "arteriovenous, fistula, craniocervical, vertebral-venous, dural, and vertebral-vertebral. Consent to participate was waived due to the retrospective nature of this study. Data collected included baseline demographics, medical history, clinical presentation, imaging results, operative reports, and clinical outcomes. Statistical analysis was based on tabulated results with descriptive statistics. The study is reported following the STROBE guidelines.

Angiographic characteristics and classification of the VVFs

We suggest a classification based on the blood flow through the fistulous connection and the number of arterial feeders. The blood flow pattern was divided into high or low. A "Type A" VVF has a high blood flow and is defined by a direct shunt between the main trunk of the VA and the surrounding veins. On the other hand, a "Type B" shunt has a low flow, and it is indirectly fed by dural or muscular branches of the VA. Finally, each category can be subdivided according to the number of arterial feeders (single or multiple). Details of the classification are presented in Table 1.

Table 1. Proposed classification of vertebral-venous fistulas.

	High-flow (indirect)		Low-flow (direct)
Type A	High-flow VVF. Direct VA feeder	Type B	Low-flow VVF. VA branch feeder.
Type A+	A plus other feeders from ICA, ECA, and/or subclavian artery.	Type B+	B plus other feeders from ICA, ECA, and/or subclavian artery.

Abbreviations: ECA, external carotid artery; ICA, internal carotid artery; VA, vertebral artery; VVF, vertebral-venous fistula.

Results

Patient characteristics and clinical presentation

Seventy-two patients were initially screened. Among these, only nine presented a true VVF. Ages ranged between 38 and 83 years. Six patients were females. All patients were treated through an endovascular approach. Patient characteristics are summarized in Table 2.

Except for one iatrogenic case, the etiology of the fistula was spontaneous in all of the remaining patients. One case (patient 5) was associated with neurofibromatosis type 1, and another (patient 7) had fibromuscular dysplasia. The most common presenting symptoms were pulsatile tinnitus/bruit, headaches, and neck pain. Other manifestations included nausea, vomiting, radicular pain, motor deficits, postural imbalance or poor coordination, and sensory deficits. In two cases (patients 4 and 5), magnetic resonance imaging (MRI) studies showed signs of spinal cord hypertension or ischemia. In addition, patient 4 presented with a spinal cord compression caused by the engorged venous structures. Baseline mRS was 3 in 6 patients and 2 in 3 patients.

Angiographic characteristics

Most VVF was distributed between vertebrae levels C1 and C2, being the V3 segment of the VA the most common feeder. Six cases were located on the right side and three on the left. Venous drainage involved the vertebral venous plexus and several neighboring veins, such as the vertebral vein and the IJV. In addition, the bulbar and perimedullary spinal veins were involved in one case each (patients 6 and 8, respectively). Six patients had a high-flow pattern, while three had a low-flow. Ipsilateral or bilateral VA anomalies, including dysplasia, marked dilatation, or tortuosity, were observed in most cases.

Endovascular treatment

All patients in our series were treated endovascularly. In all cases, a codominant VA was observed, which allowed us to perform VA sacrifice in six cases. Treatment with only VA sacrifice was performed in four cases, all of which were high-flow VVF (patients 3, 4, 5, and 7).

Three cases (patients 2, 6, and 9) with multiple feeders underwent staged (in multiple sessions) transarterial targeted embolization of the different feeders, and patients 2 and 9 also underwent VA sacrifice as part of that strategy.

In patient 9, the approach had to be converted to transvenous due to the inability to treat the fistula through the transarterial route. Therefore, the fistula was treated purely through the veins. Patient 1 presented multiple feeders and drainage to the IJV and was treated with a single transvenous embolization (IJV obliteration). Single-targeted transarterial embolization (constructive) was sufficient in patient 8.

Table 2. Summary of vertebral-venous fistula cases.

N	Age / Sex	Clinical presentation	Baseline mRS	Side, Classification, Flow	Arterial feeders	Direct venous drainage	Endovascular treatment	Complications	Clinical Outcome	Follow-up mRS	Radiological Outcome
1	66, M	Bruit, pulsatile tinnitus, HA, neck pain, dizziness, vomiting	3	L, B+, Low	Dural branches of VA, APhA, OA	IJV	Trans-venous IJV sacrifice	None	Resolved symptoms	1	Complete obliteration
2	61, F	Bruit, pulsatile tinnitus, HA, dizziness	2	R, A+, High	V3, deep cervical artery	IJV	1st–TA V3 sacrifice 2nd–TA Deep Cervical Artery embolization	1st–XII palsy/ radicular pain 2nd–Deep cervical artery rupture/ neck hematoma	Resolved symptoms after the second treatment	1	Recurrence after 1st treatment, Complete obliteration after the second treatment
3	38, F	Bruit/pulsatile tinnitus, neck pain, radicular pain	3	R, A, High	V3	VVP	TA V3 sacrifice	None	Resolved symptoms	1	Complete obliteration
4	44, M	Neck pain, UL weakness, four limb numbness	3	L, A, High	V3	VVP	TA V3 sacrifice	None	Persistence of clinical manifestations	1	Complete obliteration Improved cord edema
5	40, F	Imbalance, poor coordination [a]	3	R, A, High	V2	VV, VVP	TA V2 sacrifice	None	Resolved symptoms	1	Complete obliteration
6	65, F	HA, dizziness, neck pain, radicular pain, LUL radicular numbness, and weakness	3	L, B+, Low	V3, ECA, cervical ICA, R OA	Bulbar veins to SPV and SPS	1st–TA ICA targeted onyx 2nd–TA VA and Rt OA targeted onyx	None	Resolved symptoms	2	Complete obliteration
7	63, F	Bruit/pulsatile tinnitus, HA, dizziness [b]	2	R, A, High	V3	DCV, ACV to IJV	TA V3 sacrifice	None	Resolved symptoms	1	Complete obliteration
8	83, F	Myelopathy, vertigo	3	R, B, Low	V3	Peri-medullary vein, SPV	TA targeted onyx embolization	Onyx migration to PICA origin (retrieved by aspiration)	Improved symptoms	1	Complete obliteration
9	64, M	C8 radiculopathy [c]	2	R, A+, High	V1, thyrocervical artery, costo-cervical artery	Subclavian vein	1st–TA VA sacrifice 2nd–TA thyrocervical coil, TV coil	None	Improved symptoms	0	Complete obliteration

[a]Associated to neurofibromatosis type 1; [b]Associated to fibromuscular dysplasia; [c]Traumatic case (post thyroid surgery).

Abbreviations: ACV, anterior condylar vein; APhA, ascending pharyngeal artery; DCV, deep cervical vein; ECA, external carotid artery; F, female; HA, headaches; ICA, internal carotid artery; IJV internal jugular vein; IMAX, internal maxillary artery; L, left; LUL, left upper limb; M, male; mRS, modified Rankin Scale; OA, occipital artery; PICA, posterior inferior cerebellar artery; R, right; SPV, superior petrosal vein; TA, transarterial; TV, transvenous; UL, upper limbs; VA, vertebral artery; V2, second segment of VA; V3, third segment of VA; VV, vertebral vein; VVP, vertebral venous plexus.

Outcomes

Treatment-related neurological morbidity was noted in only one patient (patient 2), who developed hypoglossal nerve palsy 10 days after coil embolization of the VA (proximal and distal to the fistulous point). In this case, the MRI and cerebral angiogram showed a markedly engorged hypoglossal venous plexus which probably caused hypoglossal nerve compression. Additionally, this case had a residual fistula at follow-up which was caused by a feeder from the ipsilateral deep cervical artery that was overlooked in the first procedure. During retreatment, direct endovascular access through that vessel resulted in vessel rupture, which was then successfully managed with coil sacrifice of that vessel. A minor neck hematoma was evident in the immediate postoperative period, which resolved spontaneously. The 3-month follow-up showed mild improvement in the hypoglossal nerve neuropathy. The initial presenting symptoms resolved entirely.

Another case (patient 8) suffered from an intraoperative migration of embolization material (Onyx) with subocclusion at the origin of the posterior inferior cerebellar artery (PICA). Fortunately, Onyx was successfully retrieved with mechanical suction without any permanent neurological sequela.

No treatment-related mortality was seen in our series. Follow-ups ranged from 1 to 8 months (mean 3.6 months) and included catheter-based diagnostic angiography. In eight (89%) patients, clinical improvement (modified Rankin scale [mRS] of ≤ 1) was evident at the last follow-up. Complete fistula obliteration after treatment was confirmed with catheter-based diagnostic angiography. In one case (patient 4), the clinical symptoms persisted even though the fistula resolved on imaging. Please refer to Figures 1–4 for some illustrative cases.

Discussion

Spontaneous VVFs are a very rare entity, with only 136 cases found in a recent literature review.[1] In this publication, the authors reported eight of them with a male/female ratio of 1:2. About one-third of the cases present with a concomitant disorder, most commonly neurofibromatosis type 1.[1] In our series, one case was associated with FMD and another one with NF1. In general, our results converge with the findings reported in the previous literature. Only one case in our series was iatrogenic in origin.

The location of the fistula can vary according to the etiology. Most spontaneous VVFs involve the V3 segment of the VA, while traumatic and iatrogenic VVFs can be found in variable locations between the intracranial compartment and the T1 vertebral level.[1–3] In our series, all spontaneous VVFs were located at the C1 to C2 levels, which corresponds to the V3 segment of the VA. However, the only iatrogenic case of our series had the fistula located at the level of the V1 segment. On the other hand, venous drainage was observed most frequently to be the vertebral venous plexus, with other venous structures less commonly involved. Significant engorgement of the veins inside the spinal canal can be seen occasionally, which can potentially cause spinal cord compression or venous hypertension and lead to ischemic cord damage.[1,2,19]

The clinical evolution can vary according to several factors, including shunt flow rate, lesion chronicity, and venous drainage configuration.[20] Patients can be asymptomatic in up to 15% of cases, with some presenting only with an audible bruit.[1] Symptomatic cases commonly present with pulsatile tinnitus, headaches, and neck pain. However, severe clinical manifestations exist, with neurological deficits due to vertebrobasilar insufficiency, arterial stealing phenomena, venous hypertension (with congestive myelopathy or radiculopathy), and venous congestion (with direct spinal cord compression).[2,3,19–23]

In our series, five patients (55.5%) followed a benign clinical course dominated by non-life-threatening complaints. In three patients (33.3%), cervical radicular pain was evident, most probably due to radicular compression by venous engorgement into the cervical foramina. Only one patient (11.1%) presented radicular motor deficits, consisting of upper motor neuron weakness. The same patient showed imaging changes (MRI fluid-attenuated inversion recovery/T2 sequence) in the C3 to C6 spinal cord levels. We hypothesize this was secondary to ischemia from venous hypertension. Even though radiological improvement was achieved after treatment, it was not followed by clinical improvement.

Initial VVF diagnosis is usually performed with Doppler ultrasonography and computed tomography angiography (CTA). We consider six-vessel digital subtraction angiography essential to obtain a detailed anatomical understanding of arterial feeders, venous outflow, and possible leptomeningeal venous drainage. Although VVFs are most commonly fed directly from the VAs or their muscular, radicular or dural branches, contributions through ICAs, branches of the external carotid artery like the occipital artery, and cervical arteries (deep or ascending) have also been observed, reinforcing the need of a precise anatomical characterization.[3,6,11] Finally, brain and spinal cord MRI might play an important role in the differentiation of VVFs from other lesions in the craniocervical region.[13,21] Venous cord compression or ischemic changes might indicate the need for a more expeditious treatment before permanent neurological deterioration occurs.

Currently, no consensus exists on VVF management.[4,6] The goal of treatment is the obliteration of the fistulous point to prevent recurrence or recruitment of additional feeders, and options include open surgical and endovascular surgical strategies.[1,3] Several factors must be considered when approaching the management of VVFs. First, the configuration and contribution of each VA to the posterior circulation and possible retrograde filling of the fistula from the contralateral VA need to be studied. This can guide us around the option of a

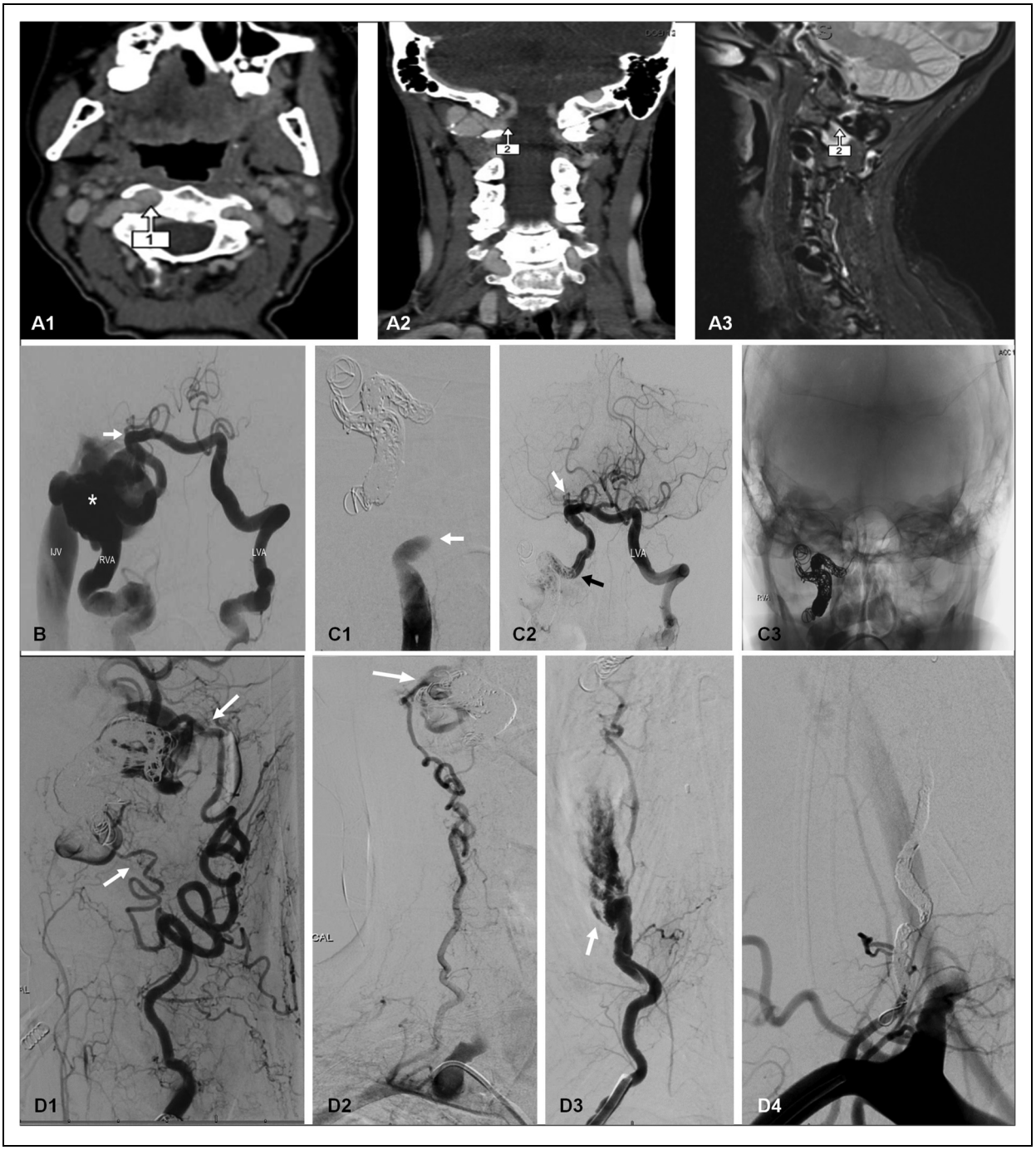

Figure 1. Type A + right VVF, case number 2 in table 2. Neck computed tomography angiography (CTA) and magnetic resonance imaging (MRI). Axial cut (A1) depicting an evident enlargement of the right vertebral foramina at the level of the V2 segment of the vertebral artery (VA) with a big venous pouch [arrow # 1]. Coronal CT (A2) and sagittal (A3) cut MRI T2 STIR sequence depicting the fistulous connection between the V3 segment of the VA and venous structures in the vicinity of the right jugular fossa [arrow # 2]. Anterior-posterior (AP) digital subtraction angiography (DSA) of the right VA [RVA]. (B) A left VA [LVA] injection showing that fistula [asterisk] is fed retrogradely in addition to the anterograde RVA filling. The high-flow extracranial fistula is shunting finally into the internal jugular vein [IJV]. When planning a deconstructive strategy, the origin of the posterior inferior cerebellar artery (PICA) informs the safety and guides the extent of parent vessel sacrifice. In this case, the PICA is distal to the fistulous connection [white arrow]. Post-embolization DSA. Right VA, AP view (C1) depicting vessel sacrifice with coils and microvascular plug [white oval mark]. Flow stagnation creates a stump [white arrow]. Left VA, AP view (C2) depicts no fistula filling. The embolization extends beyond the fistulous point [black arrow] to avoid retrograde filling of the fistula from the left VA [LVA]. Flow to the PICA is conserved [white arrow]. AP fluoroscopy image (C3) shows the coil mass. Fistula recurrence at 1-month DSA follow-up. AP (D1) and lateral (D2) views of the right subclavian artery. Filling of the fistula through branches of the deep cervical artery [white arrows] is seen. AP view (D3) of the deep cervical artery [DCA]. Rupture of the vessel after a manual run is evident with contrast extravasation [white arrow]. AP view (D4) of the right subclavian artery showing vessel sacrifice with coils. The fistula has no filling. A small asymptomatic neck hematoma developed, which resolved spontaneously within a few days.

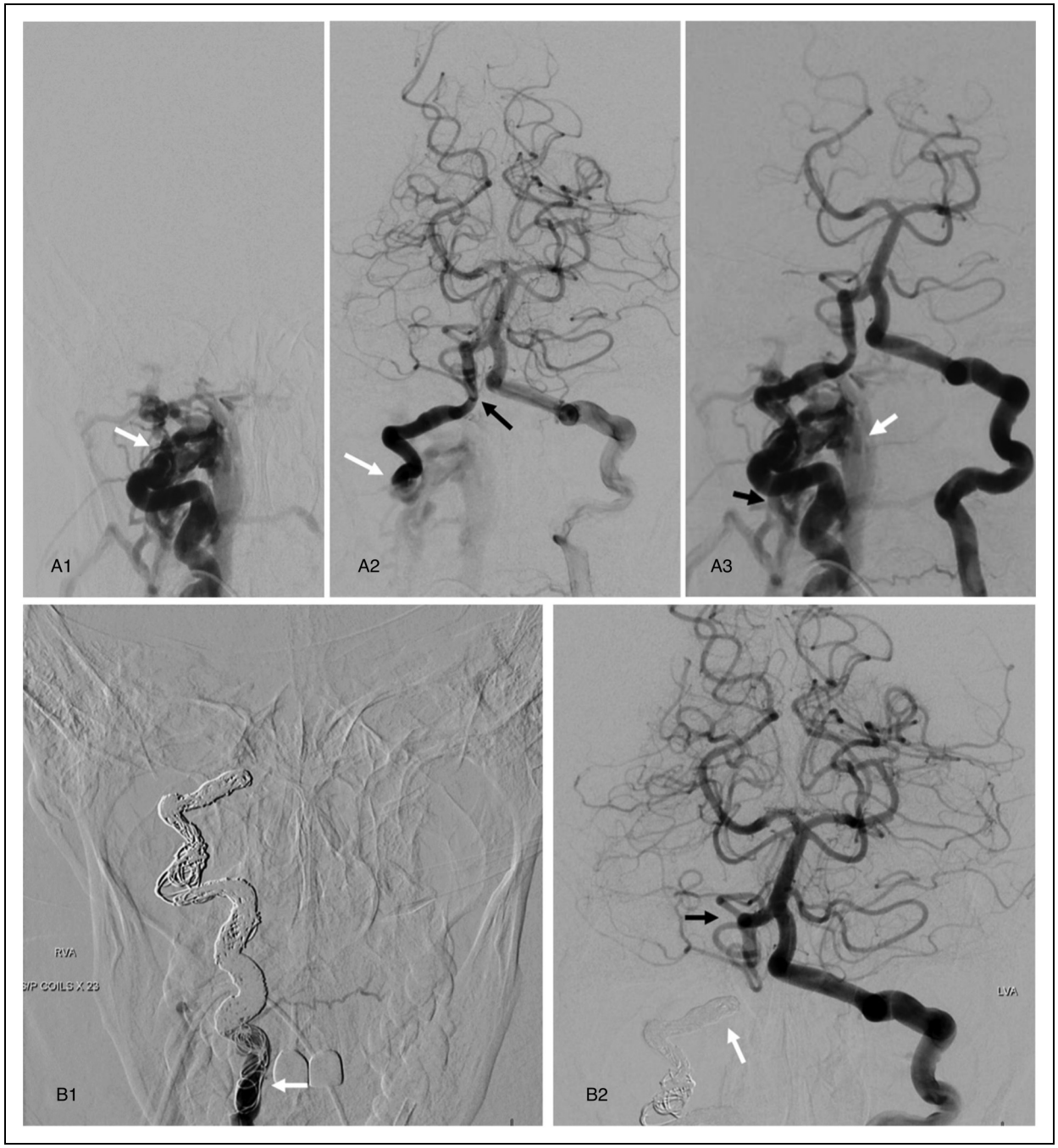

Figure 2. Type A VVF, case number 3 in Table 2. Digital subtraction angiography (DSA) of the right vertebral artery (VA) Anterior-posterior (AP) (A1) view depicting the fistulous pouch [white arrow] located at the end of a blind VA. The fistula has a high flow. The AP (A2) view shows that the left VA also feeds the fistula in a retrograde fashion [white arrow]. The origin of the right posterior inferior cerebellar artery (PICA) is distal to the fistulous point [black arrow]. AP view (A3) of a DSA run with a power injector depicts the hypertrophied and tortuous VAs. The fistula drains into the internal vertebral venous plexus [white arrow] and external vertebral venous plexus [black arrow]. Post-embolization DSA. AP view (B1) of the right VA showing vessel sacrifice with coils and a microvascular plug system. Note the flow stagnation in the VA and the stump without anterograde flow [white arrow]. AP view (B2) of a left VA shows no fistula filling from this side. The embolization extended beyond the fistulous point [white arrow] to avoid retrograde filling of the fistula by the left VA. The origin of the right PICA is preserved [black arrow].

deconstructive treatment, which can only be performed with the backup of a contralateral VA. The origin of the PICA is another relevant consideration. A PICA origin that is proximal to the fistulous connection and does not present a safe gap with the fistula will probably contraindicate VA sacrifice on that side. Additionally, patients with associated conditions such as neurofibromatosis type 1, Ehlers-Danlos syndrome, fibromuscular dysplasia, and Marfan syndrome can present with underlying vasculopathy, which can potentially increase the risk of vessel dissection or rupture when endovascular treatment is performed.[24] Finally, surgical treatment with either

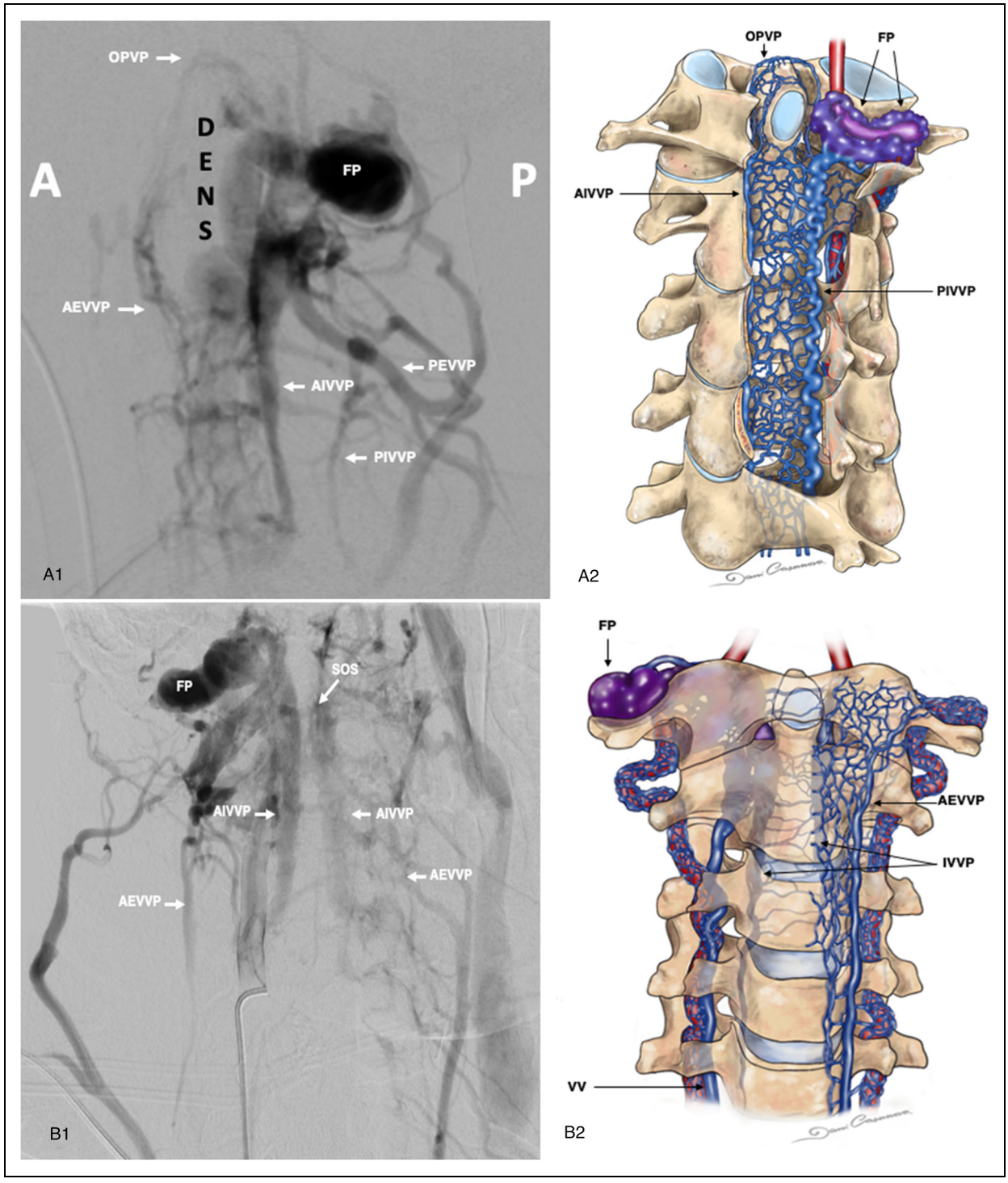

Figure 3. Type A VVF, case number 3 in Table 2. DSA of the right VA, lateral view (A1). The odontoid process [DENS] is highlighted for orientation. Anatomy is very clear due to the high pressure transferred through the fistula into the venous system. Relevant structures include the odontoid process venous plexus [OPVP], fistulous pouch [fp], anterior external vertebral venous plexus [AEVVP], anterior internal vertebral venous plexus [AIVVP], posterior internal vertebral venous plexus [PIVVP], and posterior external vertebral venous plexus [PEVVP]. Schematic illustration (A2) highlighting the following anatomical landmarks: OPVP, FP, AIVVP. The AP view (B1) shows the FP, suboccipital sinus [SOS], AIVVP, and AEVVP. Schematic illustration (B2) highlighting the following anatomical landmarks: FP, AEVVP, IVVP, and vertebral vein [VV].

ligation of the fistulous connection or surgical sacrifice of the pathologic segment of the VA might be reserved for cases with complex fistulas or as an alternative after the failure of an endovascular approach.[1,25]

Endovascular treatment for VVFs has gained popularity recently and can be classified as constructive (obliteration of the fistulous connection) and deconstructive (sacrifice of the parent VA). Constructive interventions allow for the

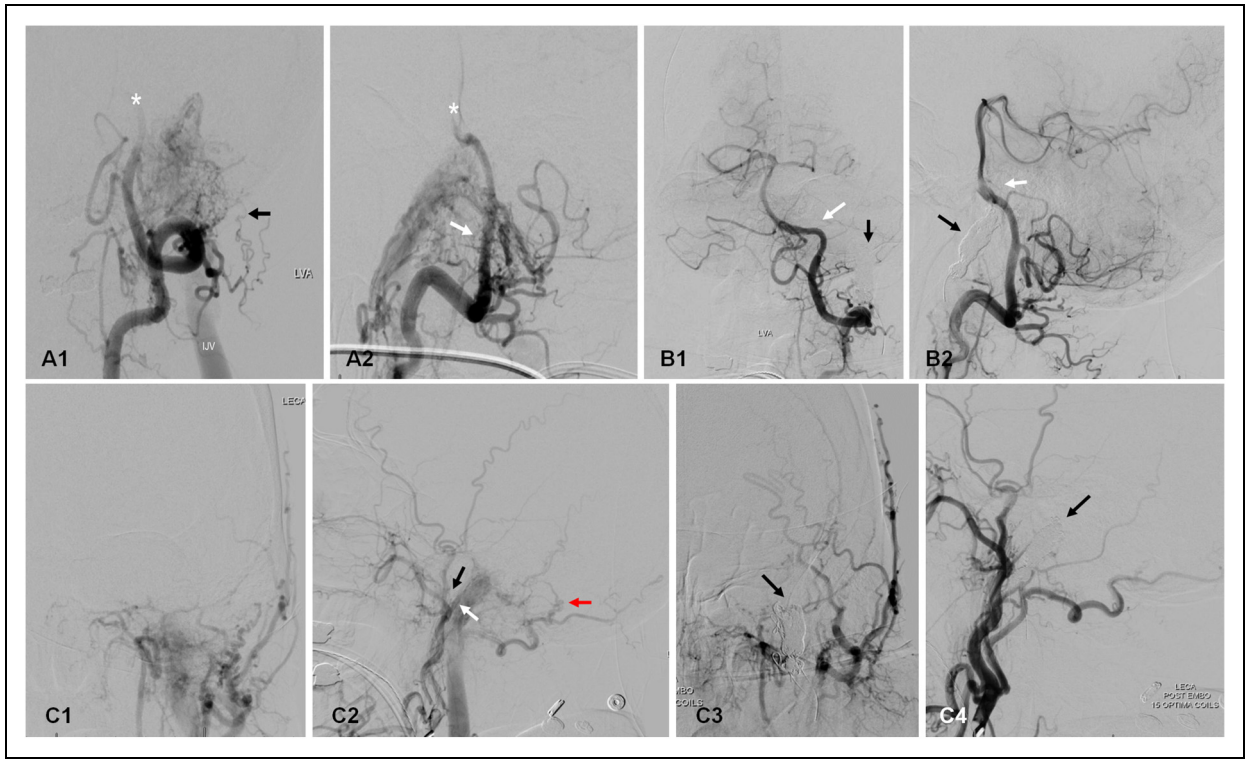

Figure 4. Type B + left VVF, case number 1 in Table 2. Digital subtraction angiography (DSA) of the left vertebral artery (VA). Anteroposterior (AP) (A1) and lateral (A2) views depicting multiple fistulous connections to the left internal jugular vein [IJV] from multiple muscular and dural branches of the left VA [black arrows]. Due to flow steal through the fistula, a poor anterograde flow through the VA is apparent [asterisk]. After transvenous embolization of the IJV. DSA of the left VA. AP (B1) and lateral (B2) views. There is no more filling of the fistula, and anterograde flow through the left VA is improved [white arrows]. The embolic coiling material is visible [black arrows]. DSA of the left external carotid artery. AP (C1) and lateral (C2) views before the transvenous embolization show additional feeders from the occipital artery [red arrow] and ascending pharyngeal artery [white arrow]. Additional AP (C3) and lateral (C4) views of the left ECA DSA after the transvenous embolization of the IJV. No more fistula filling is evident, and embolic coiling material is visible [black arrows].

preservation of the parent artery, targeting specifically the fistulous point or fistulous pouch. However, the isolation of the fistulous point can be challenging. On the other hand, the deconstructive approach aims to sacrifice the VA (proximal and distal to the fistula). Of note, in the setting of fistula recurrence, access after a deconstructive approach can be challenging since the access route to the VA can be blocked by the previous treatment.[1]

Several embolic materials have been employed, being coils the most commonly used. Additionally, they can be combined with liquid embolic materials if needed.[1,3,4,23] Detachable balloons were popular mainly in earlier reports [3,5,12,23,26,27] but have largely been abandoned as an option. The use of covered stents has also been described. However, stent migration and fracture/bending, as well as endoleak, are potential problems which can lead to fistula recurrence and ischemic events. Furthermore, a higher risk of complications has been observed when the stent is placed in the V3 segment of the VA (probably due to the high mobility of this segment at the craniocervical junction) or in VAs with significant tortuosity.[6,11] Finally, the employment of liquid embolic materials has also been described.[20] They have the disadvantage of possible migration and embolization to eloquent vascular territories, especially due to the high flow present in VVFs and the dangerous anastomoses around the area.

Treatment-related complications range between 0% and 12.5% in previously reported large case series (patient cohorts ranged from 8 to 46 patients).[3,5,12,23] In an extensive systematic review, the overall permanent neurological morbidity reported was 3.3%.[1] Minor complications include neck pain and headaches. Major complications manifest as ischemic and/or hemorrhagic events which can be caused by disruption of the normal venous outflow or ischemic arterial insults. Notably, reperfusion injury or normal perfusion pressure breakthrough rarely happens after embolizing high-flow VVFs.[1,28] The available literature has reported a 1.5% treatment-related mortality,[1] with no cases found in our series.

Suggested classification and practical approach for the management of VVFs

A previous classification of VVFs separated them into Type I, in which the fistula arises only from the parent VA; Type II, in which the fistula arises from the parent and contralateral VA; and Type III, in which the fistula comes from both VAs and has additional arterial steal

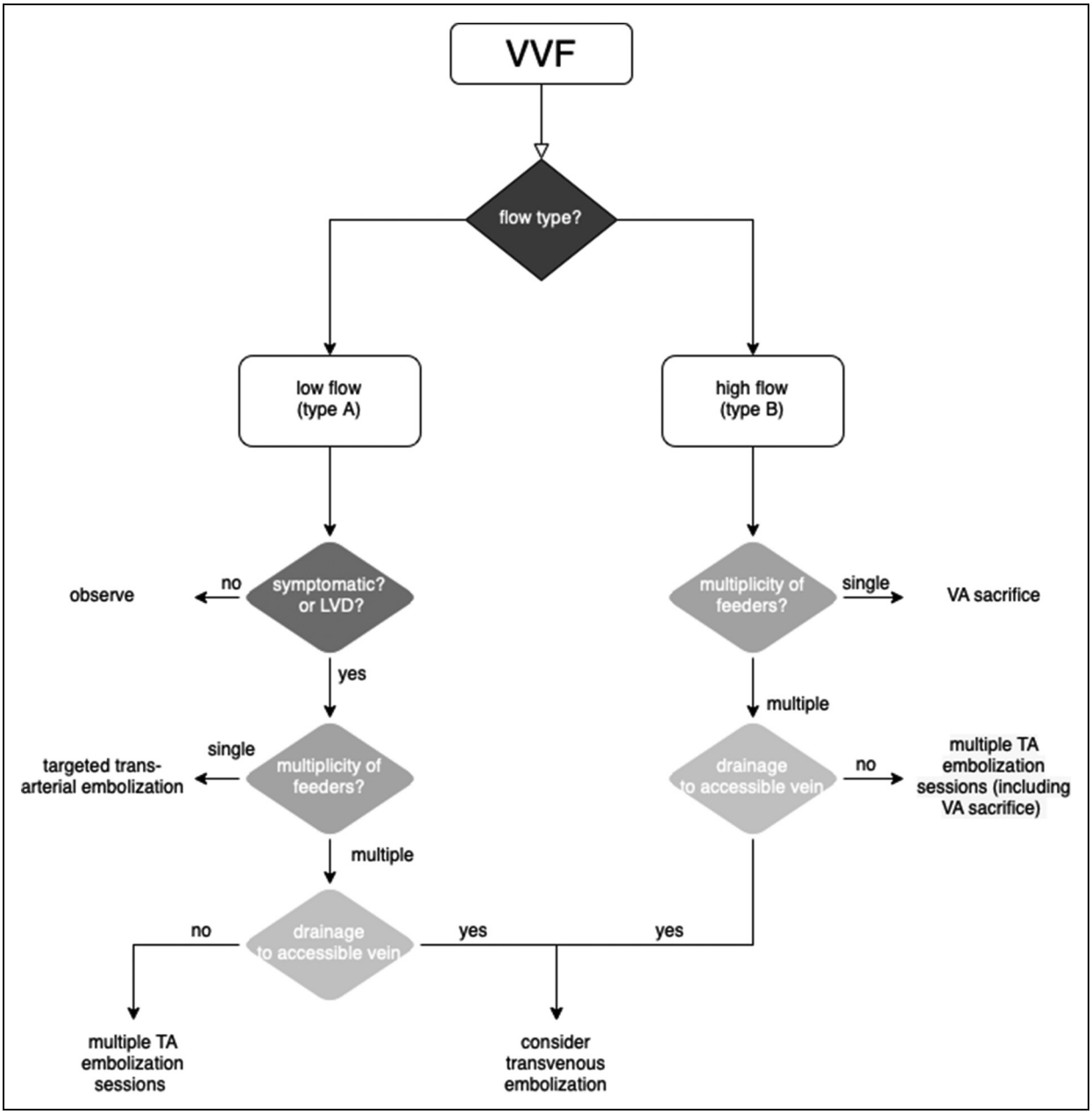

Figure 5. Algorithm of treatment for VVFs. Abbreviations: CVD, cortical venous drainage; TA, transarterial; VA, vertebral artery; VVF, vertebral-venous fistula.

from the ICA via the basilar artery.[29] We consider this classification useful for the general understanding of the pathology but limited due to the absence of relevant considerations that facilitate a treatment strategy. Our suggested treatment approach considers flow type, number of feeders, and presence of drainage into an accessible major neck vein (e.g., IJV or subclavian vein). A descriptive algorithm is presented in Figure 5.

Generally speaking, high-flow VVFs (Type A) behave more aggressively and present progressive neurological deficits more often, which warrants a timely intervention. Conversely, low-flow VVFs (Type B) are usually asymptomatic, and treatment is indicated only if they become symptomatic. Type B VVFs, on the other hand, can be quiescent for long periods before symptoms appear and may be underdiagnosed. We suggest that this might explain the low number of low-flow VVFs in our series. We consider that only high-flow lesions fed by the VA should be managed with endovascular VA sacrifice. Alternatively, low-flow single-feeder VVFs can be treated with a single targeted embolization when feasible, surgical ligation of that feeder if accessible or VA sacrifice if none of them is possible. Regardless of the flow status, the presence of multiple feeders usually dictates the need for multiple transarterial embolizations or a single transvenous approach if the VVF drains into an endovascularly accessible vein.

Limitations

A major limitation of our study is the small sample size in our series of this rare pathology. Furthermore, the retrospective nature of this study carries its inherent limitations. It may be argued that a classification system cannot be built based on only a few cases. However, we intended to create a framework on which a treatment strategy can be planned. Additionally, our classification intends to be the building block in which future modifications can be implemented according to future evidence and experience.

Conclusion

VVFs are an uncommon type of extracranial arteriovenous fistula. They can present with a direct connection (high-flow) between the VA and the surrounding veins or an indirect (low-flow) connection between branches of the VA and the surrounding veins. Our classification and management algorithm considers flow type, number of feeders, and the presence of drainage into an accessible major vein. We consider that our classification might help to organize the thought process for patient selection and treatment approach for this rare pathology. However, larger studies are needed to validate our approach and classification.

Declaration of conflicting interests

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