The first clinical description of a spinal vascular malformation was in 1890 by Berenbruch, who did not recognize the lesion as a vascular entity at surgery but only later at autopsy. Two decades later, Krause recognized a spinal vascular malformation at surgery. Credit for first recognizing that spinal vascular malformations can cause SAH must be given to Heboldt in 1885 and Gaupp in 1888, who referred to these lesions as “hemorrhoids of the pia mater.” Since then, multiple eponyms and classifications have been used for the various types of vascular malformations of the spine and spinal cord. Although reflecting the continuous evolution in our understanding of the subject, the many classifications and variable nomenclature attest to our difficulty in understanding and comparing them. In 1943, Wyburn-Mason recognized 2 main types of spinal vascular lesions: the venous type, consisting of an abnormal mass of sinuous, turgid, blue, pial veins, usually located posteriorly and below the midthoracic region of the cord; and the arteriovenous type, consisting of a fistulous capillary bed, preferentially located in the cervical enlargement anteriorly or at the lumbar enlargement, indicating for the first time that true medullary AVMs are different from what we know today as spinal dural fistulas.

The introduction of selective spinal angiography in the late 1960s and early 1970s and the studies of large series of patients by Houdart, Di Chiuro, Baker, Djindjian, and their colleagues further improved our anatomical and hemodynamic understanding of those rare lesions. Aminoff and colleagues later extensively reviewed the pathophysiology, clinical presentation, and prognosis of vascular malformations of the spinal cord. Confusion was propagated by attempts to classify spinal AVMs based on morphological aspects. From an anatomical point of view, we prefer to distinguish paraspinous, epidural locations from the AVM or AVF, realizing that the neurological symptoms will nearly always be related to the venous drainage pattern of the lesion no matter where the nidus is situated.

Vascular malformations of the spine and spinal cord are considered uncommon lesions, representing 3–16% of spinal cord vascular malformations.
of all spinal space-occupying lesions. Dural AVFs are more common than spinal cord AVMs, the latter of which have an incidence of ~10% that of brain AVMs.

In this article, we aim to demonstrate the way spinal AVMs, specifically spinal cord AVMs and spinal DAVFs, are assessed, classified, and managed at our institution, highlighting some aspects of classification and management that may be different from the views of others.

**Spinal Cord AVMs**

This group of arteriovenous shunts includes intradural lesions supplied by branches of the radicular arteries that follow the vertebral (radiculomedullary or anterior spinal arteries) or the dorsal nerve roots (radiculo-pial or posterior spinal arteries). This includes AVMs located along the intradural portion of the spinal nerves and the terminal filum. The lesions may be located along the surface of the cord (perimedullary), may be intramedullary, or both. They can be further classified into direct AVFs (macro- or microfistulas), representing ~20% of the cases, invariably located in the surface of the cord, and true nidal types of AVMs (80% of spinal cord AVMs) with an (at least partially) intramedullary nidus (Fig. 1). Multiplicity of shunts, with obvious therapeutic implications, is found in up to 20%. Syndromes associated with spinal vascular malformations are also included in this group, such as the spinal arteriovenous metameric, Klippel-Trenaunay, or Parkes-Weber syndromes.

Several classifications have been proposed, dividing the AVMs into subgroups based on morphological data. Based on angiographic interpretations that can be subjective and the relationship of the shunt with anatomical compartments of the vertebral column and spinal cord, these classifications lump together dural fistulas and true spinal cord AVMs, and sometimes even spinal tumors. The classifications assume that the lesions are frozen static in time and are not dynamic entities capable of change and subjected to genetic, hemodynamic, and biological influences. The permanent connection of a lesion to a treatment option solely on its placement in a given category is probably unwise, considering the continuous evolution of our understanding of vascular anatomy of the spinal cord, spinal cord arteriovenous shunts, and improvements in treatment tools and materials. Rather, we adopt a classification that considers the supposed origin of each shunt, its angioarchitecture, and acknowledges the concept of "host" and the relationship between the malformation and the spinal cord itself, with the knowledge that changes in this equilibrium may lead to changes in the clinical picture and prognosis.

We classify spinal cord arteriovenous shunts into 3 broad groups: those that are part of genetic hereditary disorders, those that are genetic but a nonhereditary disorder, and those with isolated focal lesions. The first group (genetic hereditary lesions) is represented by hereditary hemorrhage telangiectasia (Rendu-Osler-Weber syndrome) in which single shunt fistulas are present due to the compromise of vascular cells at the germinal stage. The second group (genetic nonhereditary lesions) involves multiple shunts that, although they are not related to a hereditary disorder, may share metameric links. In this group, cells are affected very early in their biological and/or embryonic life, and all organs to which these cells migrate in the future may harbor the malformations. A clear metameric disposition can be demonstrated (Cobb syndrome) or strongly suspected. The third group (focal lesions) includes single lesions, either AVMs or microfistulas, in which a genetic component cannot be shown.

In our experience with children, the phenotype of a spinal cord AVM is nearly always an AVF; the nidus type of AVMs is more prevalent during the 2nd and 3rd decades of life. As one can already suspect, management may be completely different for each group (an AVF in a patient with a systemic vascular compromise will be very different from an isolated focal AVF and requires a different decision-making process in management). We believe that this classification places the spinal cord AVM into a broader context, taking into consideration new physiological and genetic data, treating these lesions as expressions of more complex disease processes that may not be completely evident on imaging alone and may not simply be a morphological target. Management is then chosen based on the characteristics of each lesion and not simply by its placement in a given category.

Spinal cord AVMs usually present in the 3rd decade of life, but they can be diagnosed in children <16 years old in 20% of the cases. When the pediatric population is included, a male predominance is shown, but no sex predominance is found in the adult population. Hemorrhage is the most common presentation, occurring in 50% of all patients with spinal cord AVMs, and it is often associated with sudden onset of new neurological deficits or with worsening of preexisting deficits. The typical syndrome of spinal hemorrhage is characterized by acute severe back pain spreading along the spinal axis and legs. Motor and sensory symptoms and bladder and bowel dysfunction can occur. In a small percentage of patients (25%), preexisting spinal cord or nerve root symptoms is present. Intracranial SAH can occur with se-
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Fig. 2. Images obtained in a young boy presenting with multiple episodes of SAH. Multiple cerebral angiograms revealed no findings. A: A contrast-enhanced CT scan showing SAH and intraventricular blood. B: Sagittal T1-weighted MR image obtained after sudden onset of back pain, demonstrating thoracic intradural flow voids (arrow). C–F: Spinal angiograms demonstrating a dorsal perimedullary spinal cord AVF (small black arrow) supplied by right (C) and left (D) lumbar radiculo-pial (posterior spinal) arteries as well as the left thoracic radiculo-pial artery (E) and without direct supply from radiculomedullary (anterior spinal) arterial system (F), draining into an enlarged dorsal perimedullary venous system (open arrow in D and E).

vere headaches and disturbance of consciousness, and an intracranial origin can be wrongly suspected (Fig. 2). The natural history is based on small series that may include different lesions, but these are thought to be severe lesions with a rebleeding rate of nearly 10% in the 1st month and 40% in 1 year.3–5 The hemorrhage rate seems to be different in the pediatric population in which hemorrhage and hematomyelia are more frequent.44,49 Nonhemorrhagic symptoms include root or back pain, weakness, sensory changes, sexual, bowel, and bladder dysfunction, and rarely a bruit.

Computed tomography scanning and myelography no longer have a significant role in the initial screening of suspected spinal cord AVMs. Magnetic resonance imaging is the initial imaging modality of choice when a vascular pathological entity of the spinal cord is suspected. The T2-weighted images are useful for small lesions with slow flow. Axial T1-weighted imaging slices show areas of low signal intensity in the center of the cord, with high signal on T2-weighted images. Magnetic resonance imaging studies may be needed to accurately localize the lesion in relation to the cord tissue and the meningeal spaces, and MR imaging may also be useful in identifying spinal cord hemorrhage, thrombosis, intramedullary cavities, or atrophy, demonstrating extraspinal extension of the spinal arteriovenous metameric syndromes and distant signal changes that may explain otherwise confusing neurological symptoms.

Angiography of the spine remains the gold standard in the diagnosis and treatment planning of vascular lesions of the spine and spinal cord. Different institutions use different angiography protocols, but we favor selective angiography with a territorial approach, performed after induction of general anesthesia with controlled respiration. The angiographic investigation must outline the normal vasculature around the disease, ensuring that the full extent of the lesion is visualized and its effect on the spinal cord is understood to allow for proper treatment planning. Differentiation of the primary malformation from acquired features reflecting the arterial or venous response to the AVM over time is of utmost importance. Aspects to be observed in superselective angiography are arterial features such as peri- and intramedullary anastomosis, direct and indirect AVM supply (which may repre-
sent the “sump” effect and collateral recruitment), associated aneurysms and pseudoaneurysms, venous drainage, and most important, spinal cord supply. The nidus arrangement is difficult to appreciate at the cord level due to the complex axial and longitudinal anastomosis and the presence of sulcal perforating vessels. The situation of the venous drainage should be evaluated given that venous hypertension or thrombosis can be associated with progressive neurological deterioration, mimicking spinal dural DAVFs, or hemorrhage. Venous thrombosis responsible for clinical symptoms may be suspected on angiography but is best demonstrated on MR imaging.

The goal in the management of spinal cord AVMs is to preserve neurological function and not to obtain a perfect radiological picture. Cure of these lesions is seldom obtained without morbidity. Partial targeted treatment to obliterate weak portions like arterial or nidal aneurysms, size reduction, decreased flow, and decongestion of the venous drainage may improve a clinical situation or modify the natural history, and in our experience may represent a better choice than aggressive treatment aimed at total obliteration of the AVM. The treatment plan should consider the patient’s age, the clinical presentation (single or multiple hemorrhages), the morphology (associated aneurysms or venous varix), and flow (impaired venous drainage or stagnation) of the lesion. Ideally of course all symptomatic lesions should be obliterated. However, it is of utmost importance to identify the cases in which total anatomical cure is not possible without worsening the neurological status of the patient. Under such circumstances, an anatomical goal or objective should be defined before starting the treatment. Obliteration of a high-risk portion of the lesion, such as an associated aneurysm, the occlusion of a direct AVF within the nidus, or of a portion draining into a territory with outflow restriction, can result in a favorable long-term outcome.

In our center, treatment options for spinal cord AVMs include excision, endovascular obliteration or partial targeted therapy, or conservative management. If the lesion is deemed suitable for both endovascular and surgical treatment, embolization is first attempted (Fig. 3), often in a staged fashion. Despite the risk of rebleeding, we tend not to treat the lesion during the acute period to avoid interfering with the natural history, given that most patients show some improvement over a couple of months. An exception will be made to exclude a pseudoaneurysm (Fig. 4) or other potentially risky angioarchitecture feature, especially if > 1 hemorrhage has occurred, but early aggressive intervention is rarely needed.

Endovascular options have evolved significantly since the first report in 1971 by Doppman et al.\textsuperscript{21} of occlusion of spinal cord AVMs. From proximal arterial ligations it
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Evolved to superselective injections of permanent embolic material in specific points of the AVM, being able to preserve important blood supply to the cord. In our combined experience of 3 major centers for cerebrovascular disease, outcome measurements after embolization were graded as excellent (normal neurological status or AVM obliteration), good (better or stable neurological status, with ≥ 90% obliteration), fair (not better or in those with mild worsening), and poor (clearly worse or permanent complication from the procedure). Of 47 treated spinal cord AVMs, outcome was excellent in 49% of patients and good in 28%; 77% of the cases had a favorable result. None of the patients with a previous fixed deficit returned to their normal condition. Permanent worsening directly related to the procedure occurred in 11% of the cases, and transient complications occurred in another 11%. One patient died of ascending myelopathy 15 months after treatment. Immediate complete obliteration of the malformation was achieved in almost half of the patients by embolization alone. In the remaining patients, partial targeted embolization or combined approaches associating embolization and surgery were used. Recurrent hemorrhage occurred in 4% patients, all with cervical spinal cord AVMs incompletely treated at the time of rebleeding, while they were waiting for complementary embolization.

 Yaşargil and colleagues\textsuperscript{58} reported good results in 41 patients with spinal cord AVMs who underwent surgical treatment, although in 73% of patients no angiographic follow-up was available, despite intraoperative report of complete removal. Clinical improvement occurred in 48% of the cases and clinical worsening in 19.5%; there was 1 death directly related to surgery. Spetzler et al.\textsuperscript{51} reported on their surgical experience that 68% of the patients (27 spinal cord AVMs) improved and 29% remained the same, with 92% of complete resection. Considering the significant complexity of some approaches to these lesions, which may include anterior and anterolateral spinal approaches and postoperative stabilization, we believe it is justified to consider endovascular treatment as a first choice.

Conservative treatment includes management of the various complications of persistent neurological deficit; physical therapy, pain control management, nursing, and

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Fig. 4. \textit{Upper}: Images obtained in a 20-year-old man who presented with 2 episodes of a sudden neurological deficit 3 months apart with incomplete recovery. A vertebral angiogram (early arterial phase) (A1) demonstrating anterior arterial supply of a spinal cord AVM associated with intranidal aneurysms (arrows) likely to be the cause of repeated hemorrhagic episodes. Angiograms prior to (A2) and after (A3) superselective catheterization and embolization with glue (NBCA) of the anterior spinal arterial sulcal-commissural branches containing the aneurysms, demonstrating that aneurysms are no longer being filled as well as reduced flow and size of the residual nidal. No complications occurred, and no further hemorrhage was documented on >15-year follow-up. \textit{Lower}: Images obtained in a 40-year-old woman who presented with repeated episodes of SAH. A spinal angiogram (B1) demonstrating a spinal cord AVM at the level of the conus supplied mostly from anterior spinal artery (black arrows) and possible associated intranidal aneurysm (white arrow). Superselective catheterization of the ASA and selective injection (B2) of the sulcal-commissural branch showing the nidal aneurysm (arrow). Postembolization angiogram (B3) with glue (NBCA) into the aneurysm demonstrating elimination of the aneurysm (arrow). No complications occurred, and no further hemorrhages have been documented at the 6-month follow-up.
psychotherapy should be included. After hemorrhage has been excluded, anticoagulation with heparin may be indicated in situations in which thrombosis and/or slow flow in the AVM results in worsening venous hypertension of the cord (Fig. 5).

Spinal DAVFs

As opposed to the spinal cord AVMs, spinal DAVFs are acquired shunts located within or adjacent to the dura along the spinal canal. Anatomically, a spinal DAVF is an abnormal arteriovenous shunt in the dura, which can be located anywhere along the dura but is most commonly situated near the nerve root exit.\textsuperscript{31,37,48,52} The arterial supply usually arises from a dural branch of the dorsospinal artery, draining into a usually single, very dilated, tortuous vein that can pierce the dura quite far from a nerve root, eventually reaching the perimedullary venous system\textsuperscript{9} and producing venous hypertension that leads to a slowly progressive mixed motor and sensory myelopathy that can progress to irreversible myelopathy.\textsuperscript{3,4,12,37,52} The chronic venous hypertension reduces the arteriovenous pressure gradient and decreases tissue perfusion, resulting in progressive hypoxia to the spinal cord, producing intramedullary vasodilation, further decreasing blood flow, with possible exhaustion of autoregulation in the affected areas. This will decrease tissue perfusion and generate edema and progressive loss function.

Spinal DAVFs are the most frequent arteriovenous shunts in older adults, usually presenting after the 4th or 5th decade of life with a heavy male predominance (5:1). The most frequent location is in the midthoracic spine. A cervical localization is rare, but one should be aware that dural shunts located at the level of the foramen magnum or above can present with spinal cord myelopathy due to spinal cord venous drainage.\textsuperscript{13,34,55} The clinical presentation is characterized by progressive motor and sensory symptoms, with spasticity, paresthesias, pain, bladder, and bowel disturbances, and sexual dysfunction.\textsuperscript{37,46,48,53} Hemorrhage is extremely rare.\textsuperscript{30} A lag between initial symptoms and diagnosis is common, on average 10.5 months in our series.\textsuperscript{53}

In a recent paper, Geibprasert et al.\textsuperscript{24} made an important contribution to our understanding of these fistulas. These authors were able to connect cranial and spinal DAVFs based on embryological and developmental aspects of the venous drainage of the central nervous system, classifying them according to their venous drainage into ventral epidural, dorsal epidural, and lateral epidural,
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generating 3 groups with very different demographics, presentation, and outcomes. As they pointed out in their paper, the decision to treat is still based on the presence of venous reflux into cortical or perimedullary veins, but as with spinal cord AVMs, grouping these lesions based solely on anatomical and angiographic characteristics ignores very important differences and hinders the advancement of our understanding.

Magnetic resonance imaging is the initial investigative modality of choice, usually showing signal changes within the cord, no mass effect, and nonspecific slight enhancement with Gd, suggesting a nonneoplastic or nondemyelinating cause of the symptoms.25,35 The identification of abnormal perimedullary flow voids points to a vascular abnormality. These findings on MR imaging are nonspecific, and spinal angiography is necessary to confirm the diagnosis and establish the exact location of the nidus and the point at which the vein pierces the dura reaching the intradural space. Improvements in MR imaging technique now allow the establishment of an area of interest and help to focus spinal angiography at a specific segmental level (Fig. 6). During angiography, it is important to assess the blood supply to the spinal cord, particularly the origin of the ASA. The origin of the ASA can be located at the same level as the radicular supply to the dural fistula, a contraindication for endovascular treatment (Fig. 7).2 Bimetameric supply (a location of the shunt in the dura between 2 adjacent nerve roots resulting in dual arterial supply) can occur in a percentage of cases, with important implications for treatment and postembolization angiographic control.

Considering that endovascular and surgical treatments have low risk and good results as well as the known natural history of a progressive neurological deficit if untreated, we favor treatment for virtually all spinal DA VFs, with the goal of occluding the draining vein as it exits the dura, disconnecting the fistula from the spinal cord venous system. An attempt at endovascular treatment is always performed as a first treatment choice, usually at the time of diagnostic angiography, done after induction of general anesthesia. A permanent, low-viscosity liquid adhesive agent, NBCA, is used when attempting to reach the nidus and the proximal vein. Particulate agents, such as polyvinyl alcohol or Gelfoam are contraindicated because of very high recanalization and recurrence rates.27,38 The fistula is considered treated only if NBCA reaches and occludes the proximal portion of the draining vein. Our initial success rate with endovascular occlusion of spinal DA VFs is 25%, with no evidence of recanalization.53 Complications are rare and are usually associated with the failure to recognize a common origin for the spinal DAVF supply and the anterior or posterior spinal arterial systems or inadvertent deposition of the embolic material beyond the nidal-venous junction, reaching the perimedullary venous system.

If endovascular therapy is unsuccessful, surgery is indicated. We usually perform the surgical disconnection during the same hospital stay. Although in the past excision of the dura at the site of fistula was proposed,38,52 we, as most groups, treat spinal DA VFs by simple venous disconnection.1,40,53 Outcomes are usually good, with the majority of patients showing clinical improvement or at least stabilization of symptoms after interruption of the
abnormal venous drainage. The best results are seen in patients treated soon after onset of symptoms. Motor and deep sensory symptoms are more likely to improve than superficial sensation and sphincter disturbances.\textsuperscript{3,36,50,53} Patients submitted to surgical disconnection should have a postoperative angiogram confirming the interruption of the intradural venous drainage before leaving the hospital. If there is no clinical improvement or recurrent symptoms occur, additional MR imaging and angiography is advised. It is important to remember that signal changes within the cord can persist for a while even in patients with clinical improvement.\textsuperscript{56} Therefore, absence of improvement on MR imaging does not necessarily reflect residual or recurrent spinal DAVF.

Although multiplicity of spinal dural arteriovenous shunts is extremely rare as is the metachronous occurrence of such lesions,\textsuperscript{3} any development of new or worsening neurological deficit should warrant additional investigation with MR imaging to exclude such diagnosis or to exclude venous thrombosis.

Conclusions

Spinal arteriovenous shunt is a broad term that encompasses many entities, from true arteriovenous shunts located inside the spinal cord to epidural shunts with venous drainage directed toward the spinal canal. Even within similar locations, important differentiation should be made. Spinal cord AVMs in a patient with a systemic disease such as hereditary hemorrhage telangiectasia are clearly different from sporadic spinal cord AVMs. Management should be based on the idea that preservation of function is the goal, not complete radiological cure, and understanding the basic aspects of the disease will help to differentiate between patients in whom treatment is advised and patients in whom only more harm can be done. We favor classifications that allow us to consider all the aspects involved in each lesion instead of grouping lesions into artificial categories, which, as demonstrated recently by Geibprasert et al.,\textsuperscript{24} can impair our ability to recognize differences that can be crucial for proper management.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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