Clinical presentation and prognostic factors of spinal dural arteriovenous fistulas: an overview

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Spinal dural arteriovenous fistulas (AVFs), the most common type of spinal cord vascular malformation, can be a challenge to diagnose and treat promptly. The disorder is rare, and the presenting clinical symptoms and signs are nonspecific and insidious at onset. Spinal dural AVFs preferentially affect middle-aged men, and patients most commonly present with gait abnormality or lower-extremity weakness and sensory disturbances. Symptoms gradually progress or decline in a stepwise manner and are commonly associated with pain and sphincter disturbances. Surgical or endovascular disconnection of the fistula has a high success rate with a low rate of morbidity. Motor symptoms are most likely to improve after treatment, followed by sensory disturbances, and lastly sphincter disturbances. Patients with severe neurological deficits at presentation tend to have worse posttreatment functional outcomes than those with mild or moderate pretreatment disability. However, improvement or stabilization of symptoms is seen in the vast majority of treated patients, and thus treatment is justified even in patients with substantial neurological deficits. The extent of intramedullary spinal cord T2 signal abnormality does not correlate with outcomes and should not be used as a prognostic factor.

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KEY WORDS • dural arteriovenous fistula central nervous system vascular malformation

- spinal cord disease
- spinal cord ischemia

ROGRESSIVE neurological damage due to a vascular disorder of the spine has been recognized for more than 40 years. The advent of selective spinal catheter angiography has allowed the identification of the most common type of spinal cord vascular malformation, the spinal dural arteriovenous fistula (SDAVF) or Type I spinal vascular malformation. These lesions are defined by abnormal connections between a radicular feeding artery and the coronal venous plexus of the spine without an intervening capillary bed. The actual fistula site is at the dural sleeve of the nerve root, and the pathophysiology underlying the neurological symptoms and signs is well understood. Arterial blood flow is shunted directly into the venous plexus, under arterial pressure. The venous plexus subsequently becomes "arterialized," and obstruction of venous outflow leads to venous congestion, venous hypertension, and progressive ascending myelopathy (Figs. 1 and 2).² If untreated, this can lead to venous infarction and irreversible neurological deficits. In this review, we provide an update on this disorder with special emphasis on more recent information about clinical signs and symptoms and prognostic factors after successful treatment.

Epidemiology

Spinal dural AVFs constitute approximately 70% of spinal arteriovenous malformations and affect about 5–10 cases per million people annually.¹⁷ These lesions are thought to be acquired conditions and affect predominantly middle-aged men; at least 80% of patients in most series are male.^{15,18,20,22,26,30} Aside from speculation about genetic influence, the reason for such a strong male predominance is unknown. The average age at diagnosis is 58–63 years, and more than two-thirds of patients are in the 6th or 7th decade of life at the time of diagnosis.¹⁷ It is very uncommon for an SDAVF to be diagnosed in a patient younger than 30 years.¹⁵

Abbreviations used in this paper: AVF = arteriovenous fistula; PC-FIESTA = phase-cycled fast imaging employing steady-state acquisition; SDAVF = spinal dural AVF.

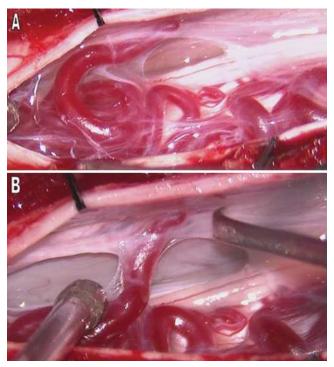


Fig. 1. A: Intraoperative photograph of an SDAVF showing the arterialized, dilated, and tortuous veins of the coronal plexus running on the dorsal aspect of the spinal cord. B: Intraoperative photograph showing isolation of the proximal portion of the draining vein emerging from the dura. Typically the draining vein emerges from the dura of the foramen underneath the corresponding pedicle.

Clinical Symptoms and Signs

Because the disorder is rare, and initial symptoms can be nonspecific, SDAVFs are challenging to diagnose early, and misdiagnoses are frequent. The most common disorders that are incorrectly initially considered in patients who ultimately are found to have an SDAVF include spinal stenosis, demyelinating disease, and spinal cord tumors. More rarely, conditions such as Guillain-Barre syndrome, amyotrophic lateral sclerosis, or peripheral vascular disease are incorrectly considered.²² Diagnosis is delayed by an average of 11–18 months after symptom onset, and it is not unusual for patients to undergo invasive surgical procedures other than interruption of the fistula for symptoms that in retrospect are attributable to the SDAVF.^{15,20,30}

Although there are characteristic clinical presentations of SDAVF, the symptoms are not specific. The majority of patients develop myelopathic symptoms that start insidiously and progress gradually over time, or occur in a stepwise fashion. In about one-quarter of patients, superimposed episodes of acute neurological worsening punctuate this decline.¹⁵ Much more rarely, in approximately 5% of cases, the presenting symptom is acute in nature. It is well known that activities that increase venous pressure (for example, exercise, the Valsalva maneuver) may worsen symptoms, while resting often improves them.

Foix-Alajouanine syndrome is a classic but frequently misunderstood syndrome associated with spinal cord vascular malformations.¹³ Traditionally this was concep-

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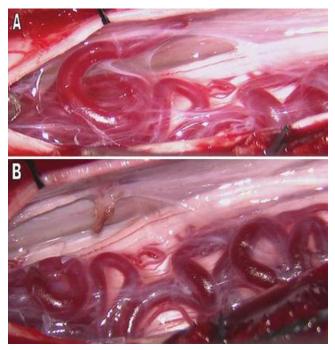


Fig. 2. Intraoperative photographs obtained before (A) and after (B) coagulation and division of the draining vein. In panel B, after surgical interruption of the fistula, the dilated arterialized veins are collapsed and have assumed a dark blue coloration typical of venous blood.

tualized as an acute or subacute neurological deterioration attributed to spinal cord venous thrombosis related to an arteriovenous malformation, resulting in venous infarction and necrotic myelopathy.¹⁰ Spinal dural AVFs had not yet been described at the time of the original report in 1926 that described 2 young men with progressive myelopathy. In retrospect, it has been speculated that the patients in the original report by Foix and Alajounaine had SDAVFs.¹² Pathological analysis of these initial cases did not show evidence of thrombosis, and symptoms may have been attributable to venous hypertension.

The most common early symptoms of SDAVF include gait disturbances and sensory changes.^{15,26} Lowerextremity weakness is also a frequent initial symptom, usually affecting approximately half of patients, ranging from 32% to 81%.^{3,8,9,15,22,24,26,30} By the time of diagnosis or surgery, symptoms have typically progressed, and almost all patients will have lower-extremity weakness.^{15,18,20,22,30} Lower-extremity weakness usually involves both legs, but is asymmetrical about half of cases,⁶ and even unilateral in up to 16%.²⁰ Symptoms infrequently involve the upper extremities, although tetraparesis has been reported with SDAVFs located in the cervical spine.^{6,28}

Sensory loss or paresthesias has been reported in 50% of cases at symptom onset and more than 90% by the time of diagnosis.^{26,30} Sensory levels to pinprick are described in 18%–37% of patients.^{15,20} In 1 series of 153 cases, the most common sensory level was at the first lumbar segment, but this did not correlate well with fistula level.²⁰ Other types of sensory disturbances at presentation include impaired vibration sense in the legs (49%), a stocking-like pattern of sensory loss (16%), and perineal numbness (13%).¹⁵

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Sphincter disturbances, including urinary or fecal incontinence or retention, are characteristic as the disorder progresses. Although a minority (4%–12%) of patients have sphincter disturbances as an initial symptom, the vast majority are affected by the time of diagnosis.^{9,18,22,26,30} About one-third of men with SDAVF have erectile dysfunction.¹⁵

Pain afflicts one-quarter¹⁵ to more than half of patients with SDAVF and can be a disabling symptom.^{9,30} Pain is commonly dull in quality, centered in the lower back, and may radiate into a lower extremity, which sometimes is misinterpreted as being due to degenerative vertebral disease. Burning pain involving the lower extremities and perineum also may occur.

Disability from symptoms related to SDAVF is commonly classified in a modified disability scale (known as the Aminoff-Logue disability scale).⁴ Mobility is scored from Grade 1 (activity not restricted despite symptoms) through Grade 5 (unable to stand; confined to wheelchair or bed) (Table 1). The median score at the time of diagnosis is 3 (require a cane for walking),^{20,30} with one-half to nearly two-thirds of patients depending on a walking aid or unable to ambulate.^{68,9}

Diagnosis

The clinical presentation of a middle-aged man with gradual-onset, progressive pain, weakness or sensory changes in the legs, and micturition disturbance should thus raise suspicion for SDAVF, a diagnosis that can be established with neuroimaging. Magnetic resonance imaging is essential, and it should encompass the entire spine, as localizing physical examination findings (such as sensory level) do not correlate well with fistula level.²⁰ Venous hypertension results in spinal cord ischemia or edema, which manifests as homogenous, longitudinally extensive T2 signal abnormality within the central spinal cord. This signal abnormality characteristically involves the conus medullaris in as many as 95% of patients.20 It should be noted that while MRI is invaluable for the diagnosis of SDAVF, T2 signal abnormality on MR images does not reliably predict postoperative outcomes, and should thus not be used for prognostic purposes.^{16,20} In addition to abnormal cord signal, the MR images typically show cord enlargement, and dilation of perimedullary veins may be seen, particularly on T2-weighted sagittal sequences. Following MRI, selective spinal angiography provides definitive diagnosis and allows localization of the fistula by identification of the feeding artery; it also allows evaluation of the venous drainage. Inability to detect the fistula on catheter angiography is not uncommon especially in older patients due to vessel tortuosity and in patients with more advanced disease because of the relative venous hypertension that may limit the degree of shunting. In the presence of typical clinical and MRI findings, PC-FIESTA MRI can be helpful in those cases in which catheter angiography fails to visualize the lesion.19

Treatment

Two effective treatment options—surgical disconnection and endovascular occlusion—are available for SDAVFs.⁵ Surgical treatment of the fistula includes a tar-

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TABLE 1: Aminoff-Logue disability scales for gait and micturition

Description	Score
gait	
leg weakness, abnormal stance or gait w/o restriction of local motor activity	1
restricted exercise tolerance	2
need for a cane or some support for walking	3
need for 2 canes or crutches for walking	4
unable to stand, confined to bed or wheelchair	5
micturition	
normal	0
urinary hesitancy, urgency, increased frequency, or altered sensation	1
occasional urinary incontinence or retention	2
total urinary incontinence or persistent retention	3

geted laminectomy, opening of the dura, and disconnection of the draining vein.¹ Surgery is durable and safe, resulting in long-term shunt occlusion in 98% of cases, with a morbidity rate of less than 2%.²⁷ In recent years, endovascular treatment has emerged as a tenable alternative for treatment of SDAVF. Complete obliteration of the proximal portion of the draining vein is key to prevent recurrences. Although endovascular procedures have achieved favorable outcomes with low morbidity rates,^{5,26} not all fistulas are amenable to endovascular therapy (which may be precluded by arterial feeders too small to catheterize or a common origin of the artery of Adamkiewicz, or not uncommonly a small posterior spinal artery, from the same segmental artery as the feeder), and long-term shunt occlusion rates may not be as high as those obtained with surgery.²⁷

Prognosis

There are no prospective studies of the natural history of untreated SDAVFs. Such studies would be difficult to justify considering that available treatments are simple, safe, and effective. Attempts have been made to deduce the natural history through retrospective analysis of patients with spinal vascular malformations who were studied before the introduction of selective spinal arteriography (and thus before a diagnosis or treatment of SDAVF). In 1974 Aminoff and Logue reported on a series of 60 cases involving men with a clinical diagnosis of spinal vascular malformations; one-fifth of the patients were nonambulatory or ambulatory with crutches after 6 months, half were severely disabled within 3 years of onset of gait impairment, and 91% were restricted in their activities within 3 years of symptom onset.⁴ In retrospect, the characteristics of the vast majority of these cases are consistent with SDAVF, and at present there is general agreement that myelopathy due to SDAVF is progressive without fistula treatment.

Outcomes

Following successful surgical or endovascular treatment of SDAVFs, almost 90% of patients experience stabilization of or improvement in their symptoms.²⁷ In a meta-analysis of 9 studies, 55% of patients had symptom improvement after treatment, 34% had no change, and 11% had neurological deterioration. When improvement occurs, patients gain 1–2 points on average on the Aminoff-Logue disability scale for ambulation^{6,9,26,27} This is largely accounted for by an improvement in motor symptoms, the symptoms that tend to respond best to treatment, which occurs in approximately two-thirds of treated patients (R. Muralidharan, unpublished data).⁹

Sensory symptoms such as numbness, dysesthesia, or burning pain tend to improve less than motor symptoms, but improvement has been reported in 12%–43% of patients (R. Muralidharan, unpublished data).^{6,9} While most patients at least have stabilization of sensory symptoms, these worsen in 14%–22% of patients, and can be a cause of significant pain.^{6,9} Sphincter disturbances tend to recover less well, remaining impaired in up to 73% of those affected and improving in only 15%.⁹

Predictors of Outcome

Physicians face a challenge when trying to predict whose symptoms will improve and whose will stabilize or worsen on an individual basis. Establishing accurate predictors has been limited by the small size of most published series and inconsistent results among studies. Clinical characteristics, rather than radiological findings, seem to have the most predictive value.

One consistent finding in the literature is that patients with severe preoperative neurological deficits have worse functional outcomes when compared with those with mild or moderate preoperative impairment (R. Muralidharan, unpublished data).^{7,8,21} Patients with preoperative gait disability scores of 2-3 improve more than those in other grade categories.^{6,31} Only 11% of patients who have severe preoperative disability (defined by a total score of 6-8 on the combined Aminoff-Logue disability scale for gait and micturition) have substantial improvement postoperatively, while 78% of those with mild disability (score 0-3) improve, and 29% of those with moderate disability (score of 4–5) improve.⁸ Nevertheless, clinical recovery is possible even for patients with severe deficits, including paraplegia. Treatments should not be withheld from patients who are severely affected, because they still may benefit from surgery (R. Muralidharan, unpublished data).^{22,29}

One large retrospective study of 153 patients with SDAVF treated surgically at a single institution found that the presence of preoperative claudication without a pinprick level was associated with greater chances of independent ambulation after surgery (R. Muralidharan, unpublished data). The presence of a sensory level to pinprick stimulation on preoperative examination was associated with a lower likelihood of postoperative improvement, suggesting that this could be a marker of irreversible spinal cord ischemia (R. Muralidharan, unpublished data).

Other clinical findings are not as clearly associated with outcomes. The time elapsed from symptom onset to treatment is not predictive of outcome in most studies (R. Muralidharan, unpublished data).^{8,9,21,26} This may in part be explained by the variable progression of irreversible neurological injury, which occurs acutely in a minority of patients. Although results of 1 study showed that older

age may be associated with worse functional outcomes,²¹ this finding has not been replicated in other studies, and there is general agreement that patients should not be excluded from treatment solely based on advanced age (R. Muralidharan, unpublished data).^{8,25,27}

Radiological findings on MRI have been studied as possible predictors of outcome, but clinicians and patients should not necessarily be discouraged by a radiological finding of a longitudinally extensive myelopathy, as neither the extent of preoperative nor the change in postoperative T2 signal abnormality correlates with postoperative clinical disability (R. Muralidharan, unpublished data).¹⁶ Single studies have found other imaging associations. For example, one found that patients with fistulas located in the lower thoracic region had an increased chance of improvement compared with those with fistulas located in the upper thoracic to midthoracic segments. Only 66% of patients with a fistula above T-9 showed symptomatic improvement, whereas improvement was seen in more than 90% of patients with a fistula located between the T-9 and T-12 levels.⁹ Another study found that spinal cord atrophy may be a predictor of unfavorable outcomes.²⁵ These associations were found in small single-center studies and still need to be replicated in other cohorts.

Finally, the success of the treatment procedure undoubtedly influences outcomes. Complete and permanent fistula obliteration provides the best chance for symptomatic improvement and a favorable outcome. In the past, endovascular treatment has been criticized because of low initial success rates and high fistula recurrence rates (up to 83% with the use of polyvinyl alcohol).¹¹ More recent data indicate a higher success rate and lower recanalization rates with the use of liquid adhesive embolization. The initial success rate of embolization was 69%²² in 1 recent study and has ranged from 30% to 90% depending on whether penetration of the proximal vein is required.^{23,26,31} Recanalization—a complication more often seen in endovascular therapy compared with microsurgical clipping-is another consideration. In 1 recent study of 26 patients treated endovascularly, 19% had recanalization and all had accompanying worsening neurological symptoms.14

The majority of patients in both surgical and endovascular series improve after treatment, and postoperative imaging to confirm resolution of the fistula is not always performed. In addition, there are insufficient data detailing long-term outcomes for patients treated with endovascular therapy, and thus it is difficult to make direct comparisons of clinical outcomes between these patients and those treated with microsurgery. Nevertheless, once the fistula is obliterated, clinical evolution after the procedure is expected to be similar in those treated by endovascular means and those treated surgically.¹¹

Summary

The most common type of spinal cord vascular malformation, SDAVF, is still overall a rare disorder. This acquired lesion affects mostly middle-aged men, who typically present with insidious symptoms of myelopathy and experience progressive or stepwise decline. Motor weakness is among the most common symptoms at pre-

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sentation and it is the symptom that responds best to treatment. After surgical or endovascular disconnection of the fistula, the prognosis is reasonably good, particularly if disability is not already severe at the time of treatment. Thus, early recognition, diagnosis, and treatment are important for patients to have the best possible chances of favorable recovery.

Disclosure

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References

- Afshar JK, Doppman JL, Oldfield EH: Surgical interruption of intradural draining vein as curative treatment of spinal dural arteriovenous fistulas. J Neurosurg 82:196–200, 1995
- ral arteriovenous fistulas. J Neurosurg 82:196–200, 1995
 Aminoff MJ, Barnard RO, Logue V: The pathophysiology of spinal vascular malformations. J Neurol Sci 23:255–263, 1974
- Aminoff MJ, Logue V: Clinical features of spinal vascular malformations. Brain 97:197–210, 1974
- 4. Aminoff MJ, Logue V: The prognosis of patients with spinal vascular malformations. **Brain 97:**211–218, 1974
- Andres RH, Barth A, Guzman R, Remonda L, El-Koussy M, Seiler RW, et al: Endovascular and surgical treatment of spinal dural arteriovenous fistulas. Neuroradiology 50:869– 876, 2008
- Atkinson JL, Miller GM, Krauss WE, Marsh WR, Piepgras DG, Atkinson PP, et al: Clinical and radiographic features of dural arteriovenous fistula, a treatable cause of myelopathy. Mayo Clin Proc 76:1120–1130, 2001
- Behrens S, Thron A: Long-term follow-up and outcome in patients treated for spinal dural arteriovenous fistula. J Neurol 246:181–185, 1999
- Cecchi PC, Musumeci A, Faccioli F, Bricolo A: Surgical treatment of spinal dural arterio-venous fistulae: long-term results and analysis of prognostic factors. Acta Neurochir (Wien) 150:563–570, 2008
- Cenzato M, Versari P, Righi C, Simionato F, Casali C, Giovanelli M: Spinal dural arteriovenous fistulae: analysis of outcome in relation to pretreatment indicators. Neurosurgery 55:815–823, 2004
- Criscuolo GR, Oldfield EH, Doppman JL: Reversible acute and subacute myelopathy in patients with dural arteriovenous fistulas. Foix-Alajouanine syndrome reconsidered. J Neurosurg 70:354–359, 1989
- Dehdashti AR, Da Costa LB, terBrugge KG, Willinsky RA, Tymianski M, Wallace MC: Overview of the current role of endovascular and surgical treatment in spinal dural arteriovenous fistulas. Neurosurg Focus 26(1):E8, 2009
- Ferrell AS, Tubbs RS, Acakpo-Satchivi L, Deveikis JP, Harrigan MR: Legacy and current understanding of the often-misunderstood Foix-Alajouanine syndrome. Historical vignette. J Neurosurg 111:902–906, 2009
- Foix C, Alajouanine T: [Subacute necrotic myelitis, slowly progressive central myelitis with vascular hyperplasia, and slowly ascending, increasingly flaccid amyotrophic paraplegia accompanied by albuminocytologic dissociation.] **Rev Neurol (Paris) 33:**1–42, 1926 (Fr)
- Guillevin R, Vallee JN, Cormier E, Lo D, Dormont D, Chiras J: N-butyl 2-cyanoacrylate embolization of spinal dural arteriovenous fistulae: CT evaluation, technical features, and outcome prognosis in 26 cases. AJNR Am J Neuroradiol 26:929–935, 2005
- 15. Jellema K, Canta LR, Tijssen CC, van Rooij WJ, Koudstaal

Neurosurg Focus / Volume 32 / May 2012

PJ, van Gijn J: Spinal dural arteriovenous fistulas: clinical features in 80 patients. J Neurol Neurosurg Psychiatry 74: 1438–1440, 2003

- Kaufmann TJ, Morris JM, Saladino A, Mandrekar JN, Lanzino G: Magnetic resonance imaging findings in treated spinal dural arteriovenous fistulas: lack of correlation with clinical outcomes. Clinical article. J Neurosurg Spine 14:548–554, 2011
- Koch C: Spinal dural arteriovenous fistula. Curr Opin Neurol 19:69–75, 2006
- Logue V: Angiomas of the spinal cord: review of the pathogenesis, clinical features, and results of surgery. J Neurol Neurosurg Psychiatry 42:1–11, 1979
- Morris JM, Kaufmann TJ, Campeau NG, Cloft HJ, Lanzino G: Volumetric myelographic magnetic resonance imaging to localize difficult-to-find spinal dural arteriovenous fistulas. Report of 3 cases. J Neurosurg Spine 14:398–404, 2011
- Muralidharan R, Saladino A, Lanzino G, Atkinson JL, Rabinstein AA: The clinical and radiological presentation of spinal dural arteriovenous fistula. Spine (Phila Pa 1976) 36:E1641– E1647, 2011
- Nagata S, Morioka T, Natori Y, Matsukado K, Sasaki T, Yamada T: Factors that affect the surgical outcomes of spinal dural arteriovenous fistulas. Surg Neurol 65:563–568, 2006
- 22. Narvid J, Hetts SW, Larsen D, Neuhaus J, Singh TP, McSwain H, et al: Spinal dural arteriovenous fistulae: clinical features and long-term results. **Neurosurgery 62:**159–167, 2008
- Niimi Y, Berenstein A, Setton A, Neophytides A: Embolization of spinal dural arteriovenous fistulae: results and followup. Neurosurgery 40:675–683, 1997
- 24. Saladino A, Atkinson JL, Rabinstein AA, Piepgras DG, Marsh WR, Krauss WE, et al: Surgical treatment of spinal dural arteriovenous fistulae: a consecutive series of 154 patients. Neurosurgery 67:1350–1358, 2010
- Shinoyama M, Endo T, Takahash T, Shimizu H, Takahashi A, Suzuki M, et al: Long-term outcome of cervical and thoracolumbar dural arteriovenous fistulas with emphasis on sensory disturbance and neuropathic pain. World Neurosurg 73: 401–408, 2010
- Song JK, Vinuela F, Gobin YP, Duckwiler GR, Murayama Y, Kureshi I, et al: Surgical and endovascular treatment of spinal dural arteriovenous fistulas: long-term disability assessment and prognostic factors. J Neurosurg 94 (2 Suppl):199–204, 2001
- Steinmetz MP, Chow MM, Krishnaney AA, Andrews-Hinders D, Benzel EC, Masaryk TJ, et al: Outcome after the treatment of spinal dural arteriovenous fistulae: a contemporary singleinstitution series and meta-analysis. Neurosurgery 55:77–88, 2004
- Symon L, Kuyama H, Kendall B: Dural arteriovenous malformations of the spine. Clinical features and surgical results in 55 cases. J Neurosurg 60:238–247, 1984
- Tacconi L, Lopez Izquierdo BC, Symon L: Outcome and prognostic factors in the surgical treatment of spinal dural arteriovenous fistulas. A long-term study. Br J Neurosurg 11: 298–305, 1997
- Van Dijk JM, TerBrugge KG, Willinsky RA, Farb RI, Wallace MC: Multidisciplinary management of spinal dural arteriovenous fistulas: clinical presentation and long-term follow-up in 49 patients. Stroke 33:1578–1583, 2002
- Westphal M, Koch C: Management of spinal dural arteriovenous fistulae using an interdisciplinary neuroradiological/ neurosurgical approach: experience with 47 cases. Neurosurgery 45:451–458, 1999

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