Curable Cause of Paraplegia
Spinal Dural Arteriovenous Fistulae

Nozar Aghakhani, MD; Fabrice Parker, MD, PhD; Philippe David, MD; Pierre Lasjaunias, MD, PhD; Marc Tadie, MD, PhD

Background and Purpose—The rarity of spinal dural arteriovenous fistulae makes physicians often overlook this potential diagnosis in patients with progressive gait disturbance and paraparesis. Consequently, patients with spinal dural arteriovenous fistulae can gradually become completely paraplegic if the final diagnosis is delayed considerably. The objective of the current study is to demonstrate that, particularly in patients with paraplegia, surgical treatment of fistula is necessary and often has a favorable outcome.

Methods—Of 42 patients with spinal dural arteriovenous fistulae treated in our institution (surgery or endovascular treatment), 6 were paraplegic preoperatively (Grade IV on the McCormick scale and Grade V on the Aminoff scale, Grade 5 of modified Rankin Scale with motor ASIA between 0 and 10 for both lower limbs). Their clinical history revealed that paraplegia appeared progressively within a period of <3 months. All patients were clinically evaluated at 6 weeks, 6 months, and then annually during an average follow-up of 3 years. Patients received at least one spinal angiography and MRI test during the follow-up period.

Results—Total exclusion of the fistula was performed surgically in all cases and was confirmed by spinal angiography. No surgical complications were recorded. All patients improved postoperatively. Three patients showed almost normal walking (Grade I on the McCormick scale, I on the Aminoff scale, Grade 1 of modified Rankin Scale) and 3 were able to walk with a cane (Grade II on McCormick, Grade III on Aminoff scale, Grade 2 of modified Rankin Scale). MRI tests were normal in all patients.

Conclusions—Our results indicate that treatment of fistula is a necessary intervention, even in patients with complete paraplegia. (Stroke. 2008;39:000-000.)

Key Words: paraplegia ■ spinal dural arteriovenous fistulae ■ surgery ■ treatment

Spinal dural arteriovenous fistulae (SDAVF) are the most common type of spinal vascular malformations.1 Although considered a well-known pathology in the neurological and neurosurgical milieu, SDAVF is frequently ignored and misdiagnosed by other physicians. Early diagnosis is difficult due to the rarity of SDAVF and the nonspecific symptoms (progressive myelopathy). In a large series reported by Jellema et al.2 the median delay of diagnosis was 15 months by which time 62 of 80 patients had severe weakness of the legs and 15 had become wheelchair-bound.

There are several reports on the clinical presentation, treatment, and outcome of patients with SDAVF in the literature. Steinmetz1 analyzed data collected from 20 papers and 532 patients. The typical clinical presentation of SDAVF is progressive myelopathy with neurological deterioration such as paraparesis, sensory disturbance, and sphincter dysfunction. If the SDAVF diagnosis is missed on initial evaluation, the neurological deterioration can worsen over time. As reported by Aminoff et al1 within 3 years after the onset of symptoms, over 90% of patients are unable to walk independently.

The appropriate treatment option in these deteriorated patients is a matter of controversy. To contribute to this debate, we analyzed data from 6 patients with paraplegia with SDAVF treated in our institution. Clinical and radiological data, patient outcomes, and our pathophysiological hypothesis are presented and discussed.

Patients and Methods
We reviewed all patients treated for SDAVF in our institution (through endovascular or surgical intervention) between December 1992 and December 2004. During this period, 42 patients were treated (22 by endovascular procedure and 20 by surgery). We analyzed 6 patients who were already paraplegic at the time of the treatment. Clinical symptoms were evaluated with regard to motor deficit (using the Aminoff scale,3 McCormick scale,4 modified...
Results

Table 3 shows the clinical presentation, the localization of the fistulae, and follow-up results in our patients.

The mean duration of symptoms before surgery was 39 months (range, 11 to 144 months). The mean duration of paraplegia before surgery was 1.6 months (range, 24 hours to 3 months). At the time of treatment, all patients had paraplegia (Grade IV on the McCormick scale and Grade V on the Aminoff scale, Grade 5 on modified Rankin Scale) with a mean duration of symptoms before surgery of 39 months (range, 24 hours to 144 months). The mean duration of symptoms before surgery was 39 months (range, 11 to 144 months). The mean duration of symptoms before surgery was 39 months (range, 11 to 144 months).

Of 4 patients experiencing pain, 3 patients (75%) reported that their pain was less severe after surgery and the fourth reported it was unchanged. Only 2 of 5 patients (40%) experiencing splinter disturbance before surgery reported an improvement.

The extension of the intramedullary T2-weighted hyperintensity on pretreatment MRI and its evolution on posttreatment MRI was not correlated with the outcome.

Considering all 36 other patients, long-term follow-up showed that 78% of patients with pretreatment gait disturbance improved, 10% worsened, and 10% remained stable. No patient becomes paraplegic after treatment.

Discussion

Although several studies have analyzed SDAVFs, none have focused particularly on patients with paraplegia. Steinmetz, Cenzato, and Mourier reported on patients with Grades IV and V on the Aminoff scale. Among patients confined to bed, Mourier et al reported that 60% were able to walk with crutches, but only one could return to work. Among the 18 patients reported by Steinmetz, 4 were Grade V on the Aminoff scale. After treatment, one patient reached Grade 0, another reached Grade II, and 2 reached Grade III on the Aminoff scale. In these 2 series, no information concerning the duration of the paraplegia was available. Jellema et al reported in 2004 a series of 44 treated patients and mentioned 13 patients who were wheelchair-bound at the time of...
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system includes patients who are unable to stand and are
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whether these patients are completely paraplegic. Atkinson
et al12 published a series of 94 dural arteriovenous fistulas
(spinal and cranial fistulas) and proposed a modified
Aminoff-Logue scale dividing the Grade V into 2 sub-
groups: Grade 5T (patients with ability to aid in transfer
with some movement of legs but confined to bed or
wheelchair and unable to stand) and Grade 5P (those who
are paraplegic with no ability to aid in transfer nor
functional lower extremity movement and who are also
confined to bed or a wheelchair). They reported 7 patients
with paraplegia (Grade 5P patients of their modified
Aminoff-Logue scale). One patient was lost for long-term
follow-up, 3 remained paraplegic (Grade 5T), and 2 could
ambulate with aids (Grade IV). In these cases, the duration
of paraplegia was not precise.

We used the ASIA score to precisely quantify the motor
deficit. All of our patients had a motor ASIA score of <10.
They had no voluntary movement of legs and the duration
of their paraplegia before treatment is clearly noted.
Paraplegia in our patients aged from 24 hours to 3 months,
and we admit that recovery is more hazardous in long-
standing paraplegia.

SDAVFs are an abnormal communication between a ra-
dicular artery and a vein within the dura matter near a nerve
root. This direct communication provokes venous hyperpres-
sure at the level of the valveless spinal cord veins. Using
intraoperative pressure recording, Hassler et al13,14 clearly
demonstrated an increase in intrinsic venous pressure (the
venous pressure in dural arteriovenous fistula was approxi-
ately 70% of the systemic arterial pressure) and the fact that
this venous pressure increases concomitantly with blood
pressure. This leads to a reduction in the intramedullary arte-
riovenous pressure gradient and thus in blood stagnation at
the spinal cord level. A decrease in tissue perfusion and
progressive hypoxia of neural tissue may follow. This
provokes histological changes, including a thickening of
intramedullary and pial vessel walls with hyalinization and
often with thrombus within the vessels. There can also be
neuronal depopulation, gliosis, and lipid-laden macro-
phages. Necrosis of the white or gray matter may follow.15
All these modifications explain the progression of the
neurological deficit. Given the gradual and progressive
installation of these phenomena, it is possible that treat-
ment of the venous hyperpressure could arrest the progres-
sion of symptoms. If there are no definitive histological
lesions, it is possible that all signs of disease would decline
once the fistula is treated.

The value of MRI changes as a predicting factor of
outcome remains controversial. Willinsky et al16 consid-
ered that there was some correlation between MRI and
eclinical outcome but at the same time they pointed out that
MRI evaluation could not distinguish those who had
improved from those who stabilized. However, the major-
ity of authors7,10,12 admit that no correlation is found
between clinical outcome and pre- or posttreatment MRI
changes. There is especially no correlation between the
extent of abnormal T2-weighted hyperintensity on pre-
treatment MRI and the clinical outcome.10 Our results
support this feature.

**Conclusion**

Our results demonstrate that endovascular or surgical
treatment is beneficial for completely paraplegic SDAVF
patients. All of our patients were able to walk indepen-
dently within 6 months after surgery. Thus, in light of
these encouraging results, we believe that paraplegia due
to SDAVF is typically a reversible symptom, and therefore
treatment should be performed even in patients with
paraplegia.

**Disclosures**

None.
Table 3. Continued

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