



Case Report

# Management of spinal dural arteriovenous fistula in a child with myelopathy

Jeremy Rajadurai<sup>1</sup>, Saeed Kohan<sup>1</sup>, Jason Wenderoth<sup>2</sup>

Departments of <sup>1</sup>Neurosurgery and <sup>2</sup>Interventional Neuroradiology, Sydney Childrens Hospital, Randwick, New South Wales, Australia.

E-mail: \*Jeremy Rajadurai - jeremiahraj@hotmail.com; Saeed Kohan - saeedkohan@yahoo.com; Jason Wenderoth - jdwenderoth@gmail.com



**\*Corresponding author:**

Jeremy Rajadurai,  
Department of Neurosurgery,  
Sydney Childrens Hospital,  
High Street, Randwick - 2031,  
New South Wales, Australia.  
jeremiahraj@hotmail.com

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## ABSTRACT

**Background:** Spinal dural arteriovenous fistulas (DAVF) are rare intradural spinal lesions. Patients with DAVF are typically in the 40's or 50's, and classically present with acute neurological deterioration. Notably, these lesions are exceedingly rare in the pediatric age group.

**Case Description:** A 2-year-old child presented with the sudden onset of lethargy, and 4/5 weakness of the left lower extremity with accompanying ataxia. The cervicothoracic MR scan revealed central cord edema from C5 to T4. A DAVF was diagnosed based on the multiple dilated intradural perimedullary veins. Following endovascular treatment, the child markedly recovered and remained stable 2 years later.

**Conclusion:** DAVFs are treatable lesions that rarely occur in the pediatric age group. They are associated with serious neurological morbidity. When suspected, they should be immediately diagnosed with magnetic resonance imaging/magnetic resonance angiography, and formal angiography. Prompt neurosurgical and neuroradiological/endovascular opinions and intervention should be sought to provide and the best treatment strategy.

**Keywords:** Dural arteriovenous fistula pediatric, Dural arteriovenous malformation paediatric, Spinal myelopathy

## INTRODUCTION

Spinal vascular malformations comprise 3–4% of all intradural lesions.<sup>[1,10]</sup> Pediatric dural arteriovenous fistulas (DAVF) are a rare subset of these entities, and occur when a single or multiple feeding arteries from a radiculomedullary artery (or its dural branch) enters and arterializes an intervertebral or radiculomedullary vein. Presenting symptoms may include acute, subacute, or chronic spinal cord dysfunction.<sup>[2]</sup> Here, we diagnosed a 2 years old with a spinal DAVF who required endovascular coiling.

## CASE REPORT

### Clinical presentation with cervical and thoracic MR/magnetic resonance angiography

A 2-year-old female presented with the sudden onset of lethargy, gait ataxia, and paresis in the left leg. On examination, she exhibited focal 4/5 motor weakness in the left lower extremity. The cervical and thoracic MR studies revealed a vascular malformation with venous congestion and central cord edema extending from C5 to T4. There were multiple accompanying dilated intradural

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perimedullary veins [Figure 1]. A large intercostal branch of the aorta supplied the region with the largest mass effect.

### Digital subtraction angiography (DSA) and coiling/embolization of DAVF

DSA showed a large feeding vessel on the left at the T6 level; this was supplied by a single “hole fistula” extending through the left T3/4 foramen and was accompanied by a venous “side” aneurysm [Figure 2]. Endovascularly, the fistula and venous aneurysm were closed with two coils and a small amount of embolic agent – precipitative hydrophobic injectable liquid-25 [Figure 2]. Following a period of inpatient rehabilitation, the child’s mobility and weakness improved, and she was discharged home.

### 2-year follow-up

Two years following embolization, the child was mobilizing independently and had a normal neurological examination. The subsequent DSA 12 months later showed no residual DAVF. Further, the magnetic resonance imaging (MRI) showed no recurrence, and the resolution of cord edema [Figure 3].

## DISCUSSION

### Historical and clinical presentation of pediatric spinal DAVF

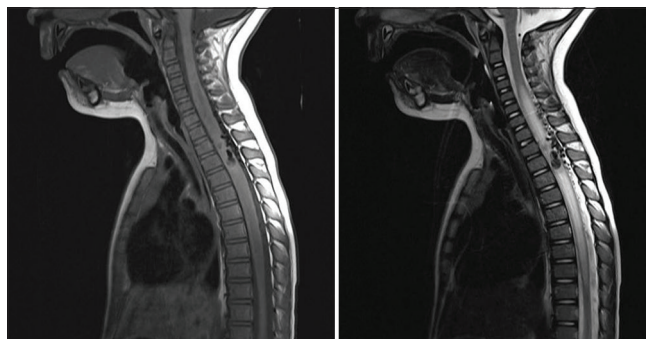
Spinal DAVFs differ in their anatomical location, size, and etiology. Intradural DAVFs usually affect the spinal cord, nerve roots or filum terminale, and typically link dural branches of radicular arteries with radiculomedullary veins.<sup>[3,7]</sup>

### History of DAVF’s

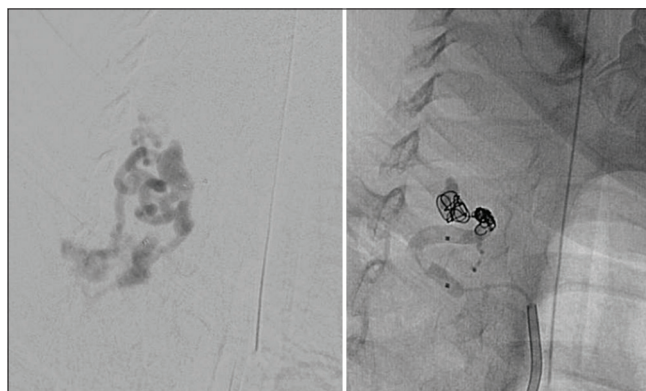
There is currently limited literature regarding the natural history of spinal DAVFs in children.<sup>[5]</sup> Rodesch *et al.* (1999) investigated the difference in natural history between pediatric and adult populations with DAVF; of 155 patients analyzed over a period of 18 years, 10 were pediatric cases versus 20 occurring in adults.<sup>[9]</sup> Compared with adults, bleeds were more frequent in children, and they exhibited increased rates of hematomyelia. Nonhemorrhagic deficits could be seen in about 22% of children versus 7% of adults usually attributed to thrombosis of portions of these DAVF.

### MR identification of spinal DAVF

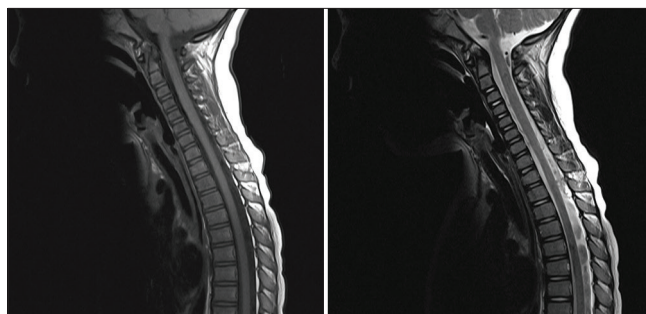
Pediatric spinal DAVFs are easily identifiable on MRI scans. They typically show resultant cord edema intrinsic cord signal changes over multiple levels<sup>[4]</sup> that typically enhance with gadolinium. T2-weighted MRI imaging can also confirm many dilated serpiginous vessels and flow voids.<sup>[8]</sup> Cord atrophy may also be present in the late stages.



**Figure 1:** T1 and T2 axial MRI images showing dural arteriovenous fistulas.



**Figure 2:** Digital subtraction angiography showing dural arteriovenous fistulas pretreatment (left) and following coil-assisted embolization (right).



**Figure 3:** T1 and T2 images 24 months post digital subtraction angiography embolization of lesion.

### Use of DSA (angiography) for diagnosis/documentation of DAVF

DSA remains the gold standard for DAVF imaging. Classically, intradural DAVFs show dilated perimedullary or intramedullary vasculature with deformation of the cord. Angiography also facilitates the reconstruction of these lesions for treatment planning, along with the identification of critical vessels such as the artery of Adamkiewicz.<sup>[11]</sup>

## Endovascular versus surgical treatment of DAVF

DAVFs can be treated endovascularly or surgically. Endovascular treatment may involve embolization with coils in conjunction with utilization of liquid embolic agents. Where endovascular treatment has failed, surgery may be warranted to close the shunt (e.g., by clipping, coagulating, and securing the exit intradural vein).<sup>[12,13]</sup> Further, surgery may be required when the feeding vessel arises from the same pedicle as an artery supplying the cord (e.g., artery of Adamkiewicz), and/or where embolization may cause ischemia.<sup>[6]</sup>

## Genetic predisposition

As spinal DAVFs can be associated with inherited and/or genetic disorders, referral to geneticists should be discussed with the child's parents or caregivers.<sup>[2,14]</sup>

## CONCLUSION

Spinal DAVFs are rare and are often difficult to diagnose in the pediatric population. Notably, DAVFs should be considered when children exhibit acute neurological deterioration. MRI and DSA should be performed with careful attention given to discerning whether these lesions can be embolized/coiled versus surgically managed.

## Statement of ethics

The research was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. The parent of the subject presented has given their written consent to publish this case (including images).

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

## REFERENCES

1. Berenstein A. Spinal Cord Arteriovenous Malformations. New York: Springer Verlag; 1992.
2. Du J, Ling F, Chen M, Zhang H. Clinical characteristic of spinal vascular malformation in pediatric patients. *Childs Nerv Syst* 2009;25:473-8.
3. Flores BC, Klinger DR, White JA, Batjer HH. Spinal vascular malformations: Treatment strategies and outcome. *Neurosurg Rev* 2017;40:15-28.
4. Hurst RW, Kenyon LC, Lavi E, Raps EC, Marcotte P. Spinal dural arteriovenous fistula: The pathology of venous hypertensive myelopathy. *Neurology* 1995;45:1309-13.
5. Hurth MH, Djindjina R, Rey A, Djindjian M. Arteriovenous malformations of the spinal cord. Clinical, anatomical and therapeutic considerations: A series of 150 cases. *Progr Neurol Surg* 1978;9:238-66.
6. Jeng Y, Chen DY, Hsu HL, Huang YL, Chen CJ, Tseng YC. Spinal dural arteriovenous fistula: Imaging features and its mimics. *Korean J Radiol* 2015;16:1119-31.
7. Miyasaka K, Asano T, Ushikoshi S, Hida K, Koyanagi I. Vascular anatomy of the spinal cord and classification of spinal arteriovenous malformations. *Interv Neuroradiol* 2000;6 Suppl 1:195-8.
8. Morris JM. Imaging of dural arteriovenous fistula. *Radiol Clin North Am* 2012;50:823-39.
9. Niimi Y, Berenstein A, Setton A, Neophytides A. Embolization of spinal dural arteriovenous fistulae: Results and follow-up. *Neurosurgery* 1997;40:675-82.
10. Rangel-Castilla L, Russin JJ, Zaidi HA, Martinez-Del-Campo E, Park MS, Albuquerque FC, *et al.* Contemporary management of spinal AVFs and AVMs: Lessons learned from 110 cases. *Neurosurg Focus* 2014;37:E14.
11. Rodesch GH, Alvarez H, Ducot B, Tadie M, Lasjaunias P. Angio-architecture of spinal cord arteriovenous shunts at presentation. Clinical correlations in adults and children. *Acta Neurochirur* 2004;146:217-27.
12. Rodesch G, Lasjaunias P. Spinal cord arteriovenous shunts: From imaging to management. *Eur J Radiol* 2003;46:221-32.
13. 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* 2002;33:1578-83.
14. Willinsky R, Lasjaunias P, Terbrugge K, Hurth M. Angiography in the investigation of spinal dural arteriovenous fistula. A protocol with application of the venous phase. *Neuroradiology* 1990;32:114-6.

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