Received:
07 May 2017

Revised:
01 December 2017

Accepted:
22 January 2018

<https://doi.org/10.1259/bjr.20170337>

Cite this article as:

Wendl CM, Aguilar Pérez M, Felber S, Stroszczyński C, Bätzner H, Henkes H. Paraspinal arteriovenous fistula: Stuttgart classification based on experience and a review of the literature. *Br J Radiol* 2018; **91**: 20170337.

REVIEW ARTICLE

Paraspinal arteriovenous fistula: Stuttgart classification based on experience and a review of the literature

^{1,2}CM WENDL, ¹M AGUILAR PÉREZ, ³S FELBER, ²C STROSZCZYŃSKI, ⁴H BÄZNER and ^{1,5}H HENKES, MD

¹Neuroradiologische Klinik, Neurozentrum, Klinikum Stuttgart, Stuttgart, Germany

²Institut für Röntgendiagnostik, Universitätsklinikum Regensburg, Regensburg, Germany

³Institut für Diagnostische und Interventionelle Radiologie und Neuroradiologie, Gemeinschaftsklinikum Mittelrhein, Koblenz, Germany

⁴Neurologische Klinik, Neurozentrum, Klinikum Stuttgart, Stuttgart, Germany

⁵Medizinische Fakultät der Universität Duisburg-Essen, Essen, Germany

Address correspondence to: Dr H Henkes

E-mail: hhenkes@aol.com

ABSTRACT

The term “paraspinal arteriovenous shunts” (PAVSs) summarizes an inhomogeneous variety of rare vascular disorders. PAVSs have been observed as congenital or acquired lesions. The clinical course of PAVSs may be asymptomatic or present with life-threatening symptoms. Based on a collection of individual cases from three institutions and a literature evaluation, we propose the following classification: PAVSs that are part of a genetic syndrome are separated from “isolated” PAVSs. Isolated PAVSs are subdivided into “acquired”, “traumatic” and “congenital” without an identifiable genetic hereditary disorder. The subgroups are differentiated by the route of venous drainage, being exclusively extraspinal or involving intraspinal veins. PAVSs associated to a genetic syndrome may either have a metamer link or occur together with a systemic genetic disorder. Again extra- vs intraspinal venous drainage is differentiated. The indication for treatment is based on individual circumstances (e.g. myelon compression, vascular bruit, high volume output cardiac failure). Most PAVSs can be treated by endovascular means using detachable coils, liquid embolic agents or stents and derivatives.

INTRODUCTION

Spinal vascular malformations can in general be classified into arteriovenous malformations (AVM) and the more common arteriovenous shunts (AVSs). AVSs account for 60–80% of all spinal vascular disorders.¹ In 1926, Foix and Alajouanine described the clinical features of what was later called “spinal dural arteriovenous fistula”.² Another 60 years later, Kendall and Logue understood the underlying vascular pathology, pointing out that the majority of “intradural spinal AVMs” were actually the venous drainage of dural AVSs located on a nerve root sleeve.¹ Over the last 20 years, different efforts have been made to subdivide AVSs with respect to their location, their venous drainage pattern, and their etiology to gain better understanding of their pathophysiology.^{3–5} In 2003, Rodesch and Lasjaunias proposed a classification scheme for spinal cord AVSs (SCAVSs), separating them into four different groups according to their localization and relationship to the dura: paraspinal, epidural, dural and intradural.⁴ Furthermore, SCAVSs, in their opinion, can be described according to the type of shunt involved (direct fistula vs nidus) and their potential relationship with genetics, vascular biological features and angiogenesis.

Dural AVSs have been the subject of many investigations, as they are the most common type of spinal AVSs.⁶ Paraspinal AVSs (PAVSs), also termed paravertebral shunts, on the contrary, are rare lesions, presenting with a female predominance. Their frequency is below 5% of the incidence of spinal dural arteriovenous fistulas. Rodesch and Lasjaunias described paraspinal (or parachordal) AVSs as malformations arising along the embryonic notochord.⁴ In contrast to epidural and intradural shunts, the AVS of PAVSs is located outside the spinal canal. Nevertheless, PAVSs can cause neurological symptoms due to cord compression by ectatic draining veins, venous congestion or by arterial steal due to reflux in the epidural or perimedullary venous system.

The literature consists of singular case reports and only a few case series. Understanding the angioarchitecture of different subtypes of PAVSs is important for estimating the course of the disorder and planning of the treatment. Based on a review of the literature and our experiences, we propose a simple classification exclusively for paraspinal shunts, illustrating it by representative cases. To our knowledge, no classification dealing exclusively with PAVSs exists in the literature.

Table 1. Classification scheme of paraspinal arteriovenous shunts, mainly divided into their origins (isolated or associated with a metameric link or genetic disorder) and their drainage pathways

<i>Stuttgart lassification of PAVSs</i>	
Isolated	
Acquired	
Extraspinal venous drainage	
Intraspinal venous drainage	
Traumatic	
Extraspinal venous drainage	
Intraspinal venous drainage	
Congenital without identifiable genetic hereditary disorder	
Extraspinal venous drainage	
Intraspinal venous drainage	
Associated	
Metameric link	
Extraspinal venous drainage	
Intraspinal venous drainage	
Systemic genetic dysplasia	
Extraspinal venous drainage	
Intraspinal venous drainage	

PAVS, paraspinal arteriovenous shunt.

METHODS AND MATERIALS

A search for articles on Pubmed (National Library of Medicine) was conducted to find studies dealing with paravertebral shunts and their diagnosis, classification and treatment from December 1980 to March 2016. The following keywords were used: “spinal arteriovenous fistula/shunt”, “paraspinal arteriovenous fistula/shunt”, “paravertebral arteriovenous fistula/shunt” or “vertebro-vertebral fistula/shunt”. Additional articles were found by searching the reference list of relevant articles. PAVSs were defined as AVSs located paravertebral with extraspinal, epidural or perimedullary venous drainage.

In addition, the records of the three participating hospitals were reviewed and patients with AV shunts outside but adjacent to the spinal canal were identified. All available examinations were reviewed by senior neuroradiologists and representative cases were chosen for illustrating the developed classification for PAVS.

Case based classification of paraspinal AV shunts

Isolated paraspinal AV shunts

Most of the paraspinal arteriovenous shunts can be categorized as isolated shunts without associated genetic, hereditary disorders. We subdivided this group according to their etiology in acquired, traumatic and congenital PAVSs, the later without identifiable genetic, hereditary disorder according to the proposed classification (Table 1). Table 2 summarizes the cases 1 to 5 to illustrate our classification scheme.

Acquired paraspinal AV shunts

Acquired PAVSs occur spontaneously in late adolescence or adulthood. According to the literature, 44% of the PAVSs cannot be linked with a history of trauma.⁷ Most acquired PAVSs show an epidural or paraspinal venous drainage.^{8–12} Vertebrojugular or vertebrovertebral AVSs are a subtype of PAVSs, predominantly draining into paraspinal venous plexuses or epidural veins.

Depending on the location of the PAVSs, neurological symptoms can vary from minor disorders as audible bruit to serious symptoms as radiculopathy or paraparesis (Figure 1). A gradual onset of symptoms is typical for this subgroup of PAVSs, although it should also be considered that a so-called “acquired” PAVSs could be hardly differentiated from a formerly asymptomatic congenital PAVS. Dilated epidural veins and feeding arteries cannot only cause myelon compression but also bony erosions. Schmidt et al reported two patients with osseous erosions of the adjacent vertebrae.¹⁰

Subarachnoid hemorrhage is a rare complication of PAVSs, probably due to sudden occlusion of perimedullary draining veins or rupture of arterialized veins.¹³ It can be accompanied by severe neurological symptoms such as paraparesis, bladder and sphincter dysfunction. Our literature search revealed only four patients, who suffered subarachnoid hemorrhage from a spontaneous PAVS.^{13–15}

Traumatic paraspinal AV shunts

Traumatic PAVSs are caused or associated with a regional trauma of different severity.^{15–18} Most often, traumatic PAVSs are located at the cervical level and are shunts between the vertebral artery and the paraspinal venous plexus or the jugular veins. Symptoms vary depending on the location and volume of the shunt and comprise an audible bruit, motor weakness, sensory disturbance, radiculopathy, pulsatile tinnitus or headache (Figure 2).^{15–17,19}

Besides vertebrovertebral or vertebrojugular PAVSs, only a few traumatic paraspinal shunts are reported in the literature. Cognard et al published the case of a 17-year-old girl, presenting with a history of progressive lower back pain, which begun 6 months after she had fallen from a horse. Spinal angiography revealed a PAVS, supplied by an iliolumbar artery and a branch of the medial sacral artery. The shunt showed venous drainage via the ascending lumbar vein to the epidural and perimedullary veins, causing a backward shift of the conus medullaris due to the space-occupying effect.²⁰

Congenital paraspinal AV shunts without identifiable genetic hereditary disorder

Paraspinal AV shunts also occur in children.^{13,21} Congenital PAVSs with and without a detectable genetic disorder can be distinguished. If diagnosed shortly after birth without history of trauma, a congenital origin of the paraspinal shunt can be assumed. Most of the reported children had first been submitted, because of a continuous heart murmur over the paraspinal or parasternal area (Figure 3). Neurological symptoms due to spinal venous congestion or compression of the spinal cord are extremely rare.²² Congestive heart failure secondary to a high shunt volume has been reported in only one patient.²³

Table 2. Overview of the example cases illustrating the Stuttgart classification for PAVSs compared to the classification according to Iizuka et al³

Case number	Age	Clinical presentation	Location of shunt	Assumed etiology of the PAVS	Venous drainage	Therapy	Clinical outcome	Stuttgart classification	Classification according to Iizuka et al ³
1	62 years	Pulsatile tinnitus, dizziness	Cervical	Unknown	Paraspinal venous plexuses	None	Persisting clinical symptoms	Isolated acquired PAVS, extraspinal venous drainage	Isolated extradural arteriovenous shunt
2	78 years	Progressive paraparesis	Lumbar	Trauma	Paraspinal and epidural venous plexuses	Transarterial Onyx embolization	Clinical improvement	Isolated traumatic PAVS, intraspinal venous drainage	Isolated extradural arteriovenous shunt
3	7 years	Audible bruit on the back	Thoracic	Congenital	Paraspinal and epidural venous plexuses	Coiling	Complete occlusion after 4 years, symptoms completely resolved	Isolated congenital PAVS without genetic hereditary disorder, intraspinal venous drainage	Isolated extradural arteriovenous shunt
4	44 years	Progressive paraparesis (due to medullary AVM), no symptoms associated with the PAVS	Sacral	Klippel-Trénaunay syndrome	Paraspinal venous plexuses	None	Paraparesis improved after endovascular treatment of the medullary AVM, PAVS asymptomatic	Associated PAVS with a metameric link, extraspinal venous drainage	Associated PAVS with metameric link
5	50 years	Clumsiness of the right hand	Cervical	Neurofibromatosis	Paraspinal and epidural venous plexuses	Coiling	Complete occlusion after 6 months, symptoms completely resolved	Associated PAVS with systemic genetic disorder, intraspinal venous drainage	Associated PAVS with systemic dysplasia

AVM, arteriovenous malformation; PAVS, paraspinal arteriovenous shunt.

Figure 1. Time-of-flight MRA of a 62-year-old patient presenting with a pulsatile tinnitus of the left ear, dizziness and no history of trauma, showing arterialization of the left transverse sinus (a). Digital subtraction angiography confirms two different arteriovenous shunts, an arteriovenous shunt of the skull base [(b), injection of the ascending pharyngeal artery, black arrow] and a vertebrovertebral shunt (white arrow) originating from the right vertebral artery, the latter draining via paraspinal venous plexuses in the ipsilateral internal jugular vein (c, d). Both shunts were not treated yet, because the patient refused any kind of therapy. MRA, MR angiography.

Paraspinal AV shunts associated with genetic disorders

Only a small number of case reports describe PAVSs related to an underlying genetic disease. Depending on the stage in cellular evolution when the vascular cells are affected, two different groups of PAVSs associated with a genetic disorder can be distinguished: non-hereditary lesions that share a developmental metamer link and hereditary lesions caused by a mutation in the vascular germinal cells (Table 1).²⁴

Paraspinal AV shunts with metamer link

Spinal arteriovenous metamer syndrome (SAMS) is a very rare disease including spinal vascular malformations of non-hereditary genetic metamer origin, which affect the spinal cord as well as other tissues originating from the same metamere. The cause of the

development of multiple malformations in parts or all tissues of the same metamere lies in a disturbed mesodermal cell migration during embryogenesis due to the mutation of mother cells. SAMS can involve the spinal cord, adjacent bone, subcutaneous tissues, and tissues of the epidural or paraspinal space and the skin.²⁵ According to the largest series of cases published by Niimi et al in 2012, the fistulous variant of SAMS is even rarer than the nidus form (86%). Among 28 cases presented, 13 showed a paraspinal involvement, although it remains unclear whether there were fistulous malformations in the paraspinal region.²⁶

The most important representative of this group of diseases is the Cobb syndrome, an association of a skin lesion with a spinal vascular malformation at the level corresponding to the respective dermatome.²⁷ Moreover, overgrowth syndromes such as the

Figure 2. A 78-year-old female patient suffering from progressive paraparesis. The sagittal T_2 weighted images (a, b) show intramedullary oedema and serpiginous leptomeningeal veins indicating a spinal AVS. The contrast-enhanced MR angiography (c) reveals a primary shunt into the epidural and paraspinal venous plexuses. DSA of the third lumbar radicular artery on the left confirms a paraspinal AVS that drains primarily into the segmental epidural spinal venous plexus (d) and into a leptomeningeal vein of the longitudinal venous drainage of the spinal cord (e). AVS, arteriovenous shunt; DSA, digital subtraction angiography.

Figure 3. Large calibre arteriovenous shunt from the descending aorta to the vena cava inferior in a 7-year-old boy with an audible bruit on his back. Venous congestion of the para- and intraspinal veins (a, b). The arteriovenous connection was occluded with coils in 2012 (c). A follow-up DSA, 4 years later confirmed the persistent occlusion of the paraspinal arteriovenous shunt (d). DSA, digital subtraction angiography.

Klippel–Trénaunay syndrome and CLOVES syndrome, which also seem to have a metameric link, can be accompanied by vascular malformations (Figure 4).²⁸ Alomari et al reported six cases of CLOVES syndrome with complex spinal–paraspinal high flow lesions.²⁹

Paraspinal AV shunts associated with genetic dysplasia

Several genetically inherited conditions are known to be associated with vascular anomalies, most notably neurofibromatosis (Figure 5) and hereditary hemorrhagic telangiectasia (Rendu–Osler–Weber disease).^{4,11,30} The latter is characterized by an early affection of the vascular cells in the germinal stage.⁴ Besides, a few case reports concerning PAVSs in patients with fibromuscular dysplasia exist in the literature.^{28,30}

DISCUSSION

Numerous classifications for spinal AVSs have been published, since the first AVSs around the spinal cord were described in the late 19th century by Hebold and Gaupp.^{5,31–40} These classifications divide spinal AVSs into subgroups on the basis of morphological criterion which, according to Rodesch and Lasjaunias, create unfortunate confusion due to subjective interpretation of angiographic data or imprecise description. Thus, in 2003 Rodesch and Lasjaunias proposed a classification scheme for spinal AVSs, categorizing them according to (1) their precise localization, (2) the type of shunt involved (fistula or nidus) and (3) their potential association with genetic alterations, vascular biological features, and angiogenesis. Thereby, they took an additional step separating extradural shunts in extradural/epidural and PAVSs.⁴

In our opinion, a classification system for PAVSs is worthwhile, as they represent an independent entity of AV shunts, which can be distinguished from extradural or epidural AV shunts by differences in their arterial supply and their venous drainage pattern. Subdivision could help to draw information about

natural course, treatment options and outcomes together and, thereby could help to determine therapy strategies in the future.

Table 3 summarizes the available literature for PAVSs, classifying them according to the proposed classification for PAVSs. In the literature, the isolated PAVSs are much more common than the associated ones. Only a few case reports and small case series with coherence of hereditary genetic or metameric origin and paraspinal shunts exist.^{28,43–45}

Iizuka et al reported a case of high-flow paraspinal osseous epidural AVF, presenting a classification for epidural/paraspinal AVMs/AVFs at the same time. In this classification, they further subcategorized the isolated epidural/paraspinal AVMs/AVFs in (a) paraspinal AVFs along the segmental nerve route, (b) vertebral bone AVMs, (c) slow flow epidural AVFs and (d) other extradural AVFs/AVMs. In contrast, our classification scheme is based on the supposed origin of the PAVS and its potential relationship to an underlying disease with respect to the venous drainage pattern. This way of classifying PAVSs is more consistent and, thereby superior to the one proposed by Iizuka et al whose classification is a mixture of morphological aspects, etiology and drainage characteristics.³

Anatomy and pathophysiology

The vascular anatomy of spine and myelon is relevant for the understanding of the pathophysiology of PAVS and the potential complications of their treatment.

PAVSs arise from segmental arteries and are located either at the main trunk or its branches.⁴ Every segmental artery supplies all the tissues on one side of a given metamere with exception of the spinal cord. The segmental arteries are connected via a system of extradural longitudinal and transversal anastomoses. In the cervical region, the vertebral arteries together with the deep cervical and ascending cervical arteries form the effective chain of longitudinal anastomoses.^{39,46} Therefore, vertebrovertebral

Figure 4. A 44-year-old male patient with Klippel-Trénaunay syndrome. He presented with progressive paraparesis due to a medullary arteriovenous malformation, supplied by the anterior spinal artery, which carries a fusiform aneurysm (a). The supply of the AV-malformation and the aneurysm were occluded with coils in order to eliminate the associated bleeding risk (b), achieving a significant reduction of the arteriovenous shunt. While the clinical symptoms were caused by the spinal AVM, injection of the right common iliac artery shows an asymptomatic paraspinal arteriovenous shunt with paraspinal venous drainage (c–e). This AV-shunt remains asymptomatic and, therefore untreated. The condition of the patient stabilized, he walks with a cane. AVM, arteriovenous malformation.

fistulas should also be rated among the group of PAVS.⁴ The venous drainage of the spinal cord parenchyma is effected through intrinsic veins of the parenchyma in the anterior and posterior median spinal veins. These longitudinal collectors drain into the epidural venous plexus through radicular veins. The epidural venous plexus is connected with the paraspinal venous system, namely the intercostal or segmental veins, the ascending lumbar vein, and the vertebral and deep cervical veins.⁴⁶ Normally, the paravertebral veins drain into the azygos and hemiazygos venous system but can in presence of a PAVS also lead to venous congestion in the epidural venous plexus and even the perimedullary veins.

The reason of retrograde venous flow in the spinal venous system was widely discussed in literature.^{47–49} In anatomic studies the existence of venous valves was already excluded for the spinal venous system.⁵⁰ Quite the contrary, an antireflux mechanism was discovered, consisting of narrowing and zigzagging of the

Figure 5. A 50-year-old female patient with neurofibromatosis. Clumsiness of her right hand and a suspected intraspinal neurofibroma (a) led to a surgical exploration of her cervical spine. During this operation, no neurofibroma but intraspinal arterialized veins were found. The injection of the right vertebral artery showed several fusiform dilatations of this artery and an arteriovenous shunt to the para- and intraspinal veins (b, c). Both, the aneurysms and the arteriovenous shunts were occluded with detachable coils (d). Follow-up MRI, 6 months later confirmed the complete disappearance of the intraspinal veins (e).

radicular veins while crossing the dura.⁴⁶ Gilian hypothesized that a failure in the antireflux mechanism of the epidural space as a result of chronic venous hypertension could be a possible explanation for the reflux into the epidural and perimedullary venous system.⁵⁰

Clinical presentation

Symptomatic PAVSs most often present with reflux into the epidural venous system with venous hypertension in the epidural veins or even venous congestion. The latter could be a result of venous stenosis, pouches or thrombosis due to longstanding high-flow shunts.^{14,26} Compressive myelopathy, isolated nerve root compression or infrequently steal of blood flow with high-output cardiac failure are possible clinical symptoms, however, there is no classical presentation.^{3,10,14,26,27,51,52}

Congenital PAVSs often cause a murmur as the only symptom, especially before the age of 10.⁵³ Therefore, the need for treatment in this subgroup of PAVSs is discussed controversial, especially because the natural course of this rare entity is unknown and epidural venous engorgement occurs infrequently.²¹ Nevertheless, there seems to be a tendency towards prophylactic treatment in asymptomatic children to avoid the possible development of

Table 3. Review of the literature

Author	Year	No. of cases	Assumed etiology of the PAVS	Clinical presentation	Location of shunt	Venous drainage	Therapy	Stuttgart classification	Classification according to Iizuka et al ³
Alomari et al ²⁹	2011	6	Syndromal (CLOVES syndrome)	Weakness of upper extremity, back pain, urinary incontinence, leg weakness	Thoracic, lumbar	Paravertebral, epidural	Surgery, embolization (material unknown)	Associated PAVS with metamerik link, extra- and intraspinal drainage	Associated PAVS with metamerik link
Ashour et al ⁴¹	2015	2	Congenital	Heart murmur	Cervical	Epidural	Coils + occlusion device	Isolated congenital PAVS, intraspinal drainage	Isolated extradural arteriovenous shunt
Briganti et al ³⁰	2014	29 (review)	Spontaneous (4), syndromal (NF; 10), trauma/iatrogen (15)	Inferior limb weakness, pulsatile bruit, cervical pain, tinnitus	Cervical	Paravertebral	Balloon, coils, transarterial glue, surgery	Isolated acquired (4)/traumatic (15) PAVS, extraspinal drainage; associated (10) PAVS with systemic genetic dysplasia, extraspinal drainage	Isolated PAVSs along the segmental nerve, type a (vertebrovertebral shunts)
Chen et al ¹¹	1996	1	Spontaneous	Leg weakness	Thoracic	Epidural	Transarterial glue	Isolated acquired PAVS, intraspinal drainage	Isolated extradural arteriovenous shunt
Cognard et al ²⁰	1995	1	Trauma	Pain, radicular hypoesthesia	Sacral	Ascending lumbar vein	Balloon occlusion	Isolated traumatic PAVS, extra- and intraspinal drainage	Isolated extradural arteriovenous shunt
Farhat et al ³¹	2015	2	Congenital	Heart murmur	Cervicothoracic (1), thoracic (1)	Epidural	None	Isolated congenital PAVS, intraspinal drainage	Isolated extradural arteriovenous shunts
Fotso et al ⁴²	2006	1	Congenital	Heart murmur	Thoracic	Epidural	Coils	Isolated congenital PAVS, intraspinal drainage	Isolated extradural arteriovenous shunt
Hui et al ²³	1993	2	Congenital	Leg spasm, underweight, heart murmur	Cervical (1), Thoracic (1)	Paravertebral, epidural, perimedullary	Transarterial glue (2), coils (1)	Isolated congenital PAVS, extra- and intraspinal drainage	Isolated extradural arteriovenous shunts
Kähärä et al ⁸	2003	1	Spontaneous	Pain, bladder dysfunction	Sacral	Epidural	Transarterial glue	Isolated acquired PAVS, intraspinal drainage	Isolated extradural arteriovenous shunt
Niimi et al ²⁶	2005	5	Congenital	Heart murmur	Thoracic (4), Lumbar (1)	Paravertebral, epidural	Transarterial glue (4), coils (1)	Isolated congenital PAVS, extra- or intraspinal drainage	Isolated extradural arteriovenous shunts
Schmidt et al ¹⁰	2008	3	Spontaneous	Pain, no neurological symptoms	Thoracic (1), Lumbar (1), Sacral (1)	Epidural	Transarterial onyx	Isolated acquired PAVS, intraspinal drainage	Isolated extradural arteriovenous shunt

(Continued)

Table 3. (Continued)

Author	Year	No. of cases	Assumed etiology of the PAVS	Clinical presentation	Location of shunt	Venous drainage	Therapy	Stuttgart classification	Classification according to Iizuka <i>et al</i> ³
Toi <i>et al</i> ¹⁴	2011	1	Trauma	Pain, paraplegia, bladder and sphincter dysfunction	Sacral	Perimedullary	Transarterial glue + surgery	Isolated traumatic PAVS, intraspinal drainage	Isolated extradural arteriovenous shunt
Yamaguchi <i>et al</i> ⁹	2008	1	Spontaneous	Tetraparesis, bowel dysfunction	Cervical	Epidural	Transarterial glue	Isolated acquired PAVS, intraspinal drainage	Isolated extradural arteriovenous shunt
Yeh <i>et al</i> ¹⁵	2014	5	Trauma (3), Spontaneous (2)	Headache, pulsatile tinnitus, neck pain, subarachnoidal hemorrhage (2)	Cervical (vertebro-vertebral PAVSs)	Paravertebral	Covered stent (3), coils (3)	Isolated acquired (2) or traumatic (3) PAVS, extraspinal drainage	Isolated extradural arteriovenous shunts

NF, neur of fibromatosis; PAVS, paravertebral arteriovenous shunt. Cases were classified according to the Stuttgart classification and the classification according to Iizuka *et al*³.

congestive heart failure, which in fact is an infrequent if not inexistent phenomenon.⁵⁴

In contrast, traumatic and acquired PAVSs may present with local pain, but also severe symptoms for example paraparesis is possible and therefore, immediate treatment should be favored.^{11,53}

The natural course of the different subtypes of PAVSs is still.

Treatment

The indication for treatment of PAVSs is based on clinical signs and symptoms. Incidentally detected PAVSs with exclusive extraspinal drainage often remain untreated. Reasons for treatment include intraspinal drainage with congestion or compression of the myelon, intraspinal hemorrhage, vascular bruit, local pain or a high-output cardiac failure.

Endovascular as well as surgical treatment aims at a complete interruption of the pathological arteriovenous connection.

Before the development of endovascular methods, surgery of PAVSs was the only option. Due to unsatisfactory results, neurosurgical therapy is now only indicated in the case of compressive extradural hematoma.^{8,12} Endovascular embolization became, meanwhile, the accepted first line treatment method for PAVSs.^{23,26,55,56}

Given the frequently present anastomoses and direct arterial supply to the myelon in the surrounding territory of PAVSs, particle embolization should be avoided. No matter if polyvinyl alcohol or one of the more technical embolization particles [e.g. Embosphere Microspheres, Boston Scientific (Marlborough, MA)] is used, the control of the distribution of these particles can be difficult and any inadvertent occlusion of the pial supply of the myelon may result in a major neurological deficit. Vessel occlusion after particle embolization is frequently not permanent, especially with polyvinyl alcohol, and recurrences can be more difficult to treat than the original lesion.

For small calibre single hole or plexiform arteriovenous connections, we prefer nBCA/Lipiodol (e.g. Glubran2, GEM) rather than ethylene-vinyl alcohol copolymer (Onyx; Medtronic). During the glue injection, arterial reflux should be avoided but a well-controlled and limited venous passage with occlusion of the draining vein adjacent to the shunt is the aim.

In a large calibre shunt, the propagation of any liquid embolic agent can be difficult to control. For the time being, coil occlusion is the preferred technique, if needed in combination with liquid embolic agents. Both selective obliteration of the shunt as well as parent artery occlusion can be part of the treatment strategy.

In selected PAVSs, a reconstruction of the artery from which the supply of the PAVSs arises can be achieved using telescoping porous stents, stent grafts or flow diverters. The vertebral artery is the preferred vessel for this treatment strategy.

CONCLUSION

PAVSs occur isolated or associated with genetic disorders. Isolated PAVSs are either acquired, due to trauma or congenital.

PAVSs associated with a genetic disorder have a metamerick link or a systemic genetic background. The route of venous drainage, mainly responsible for the symptoms, is either extraspinal or intraspinal.

The presented classification system can intuitively be used and the subclassification of this complex and rare type of AVS may

lead to better understanding of the clinical course and, therefore, can help to reliably determine treatment strategies in the future. Currently, the indication for and technique of endovascular treatment is based on individual clinical and anatomical criteria. With this classification, a standardized nomenclature is proposed, which could also help to create a comprehensive database in the future.

REFERENCES

- Kendall BE, Logue V. Spinal epidural angiomatous malformations draining into intrathecal veins. *Neuroradiology* 1977; **13**: 181–9. doi: <https://doi.org/10.1007/BF00344211>
- Foix CH, Alajouanine T. La myélite nécrotique subaigue. *Rev Neurol* 1926; **2**: 1–42.
- Iizuka Y, Suzuki M, Suzuki K, Shimoji K, Komura S. High-flow paraspinal osseous epidural arteriovenous fistula. A case report. *Neuroradiol J* 2008; **21**: 433–9. doi: <https://doi.org/10.1177/197140090802100322>
- Rodesch G, Lasjaunias P. Spinal cord arteriovenous shunts: from imaging to management. *Eur J Radiol* 2003; **46**: 221–32. doi: [https://doi.org/10.1016/S0720-048X\(03\)00093-7](https://doi.org/10.1016/S0720-048X(03)00093-7)
- Spetzler RF, Detwiler PW, Riina HA, Porter RW. Modified classification of spinal cord vascular lesions. *J Neurosurg* 2002; **96**(2 Suppl): 145–56. doi: <https://doi.org/10.3171/spi.2002.96.2.0145>
- Krings T, Mull M, Gilsbach JM, Thron A. Spinal vascular malformations. *Eur Radiol* 2005; **15**: 267–78. doi: <https://doi.org/10.1007/s00330-004-2510-2>
- Vinchon M, Laurian C, George B, D'arrigo G, Reizine D, Aymard A, et al. Vertebral arteriovenous fistulas: a study of 49 cases and review of the literature. *Cardiovasc Surg* 1994; **2**: 359–69.
- Kähärä V, Lehto U, Sajanti J. Presacral arteriovenous fistula: case report. *Neurosurgery* 2003; **53**: 774–7. doi: <https://doi.org/10.1227/01.NEU.0000080066.56242.C4>
- Yamaguchi S, Hida K, Asano T, Yano S, Kashiwazaki D, Iwasaki Y. Case of cervical paraspinal arteriovenous fistulae with a huge intracanalicular varix successfully treated with transvenous embolization. *No Shinkei Geka* 2008; **36**: 547–53.
- Schmidt C, Lonjon J, Costalat V, Menjot De Champfleury N, Seris C, Brunel H, et al. Paraspinal arteriovenous malformations with perimedullary venous drainage. *J Neuroradiol* 2008; **35**: 165–72. doi: <https://doi.org/10.1016/j.neurad.2008.01.081>
- Chen CJ, Huang CC, Hsu YY, Hsu WC. Small isolated paraspinal arteriovenous fistula. *AJNR Am J Neuroradiol* 1997; **18**: 359–61.
- Honarmand AR, Ansari SA, Alden TD, Soltanolkotabi M, Schoeneman SE, Hurley MC, et al. Endovascular management of pediatric high-flow vertebro-vertebral fistula with reversed basilar artery flow. A case report and review of the literature. *Interv Neuroradiol* 2013; **19**: 215–21. doi: <https://doi.org/10.1177/159101991301900211>
- Hiu T, Yoshioka T, Kitagawa N, Izumo T, Okunaga T, Suyama K, et al. Vertebral arteriovenous fistula presenting with subarachnoid hemorrhage: a case report. *No Shinkei Geka* 2009; **37**: 667–71.
- Toi H, Matsubara S, Watanabe S, Yamashita T, Uno M. Paraspinal arteriovenous fistula presenting with subarachnoid hemorrhage and acute progressive myelopathy--case report. *Neurol Med Chir* 2011; **51**: 846–9. doi: <https://doi.org/10.2176/nmc.51.846>
- Yeh CH, Chen YL, Wu YM, Huang YC, Wong HF. Anatomically based approach for endovascular treatment of vertebro-vertebral arteriovenous fistula. *Interv Neuroradiol* 2014; **20**: 766–73. doi: <https://doi.org/10.15274/INR-2014-10072>
- Vergouwen MD, Majoie CB, van Rooij WJ, Poll-The BT. A vertebro-vertebral fistula as a complication of a jugular line. *J Pediatr* 2006; **149**: 576.e1–576.e2. doi: <https://doi.org/10.1016/j.jpeds.2006.06.049>
- Mortimer A, Stubbs E, Cookson D, Dawson R, Fleet M. Delayed presentation of a vertebral arterio-venous fistula secondary to penetrating cervical trauma: endovascular management using coil embolisation - a case report. *J Radiol Case Rep* 2009; **3**: 9–15. doi: <https://doi.org/10.3941/jrcr.v3i6.81>
- Kypson AP, Wentzensen N, Georgiade GS, Vaslef SN. Traumatic vertebrojugular arteriovenous fistula: case report. *J Trauma* 2000; **49**: 1141–3. doi: <https://doi.org/10.1097/00005373-200012000-00031>
- Shirakawa M, Nishioka T, Yamashita K, Maeda Y, Arita N. Traumatic vertebro-vertebral arteriovenous fistula manifesting as radiculopathy. Case report. *Neurol Med Chir* 2008; **48**: 167–70. doi: <https://doi.org/10.2176/nmc.48.167>
- Cognard C, Semaan H, Bakchine S, Miaux Y, Thibault S, Sola Martinez MT, et al. Paraspinal arteriovenous fistula with perimedullary venous drainage. *AJNR Am J Neuroradiol* 1995; **16**: 2044–8.
- Farhat N, Desprechins B, Otto B, Ramaekers V, Seghaye MC. Paraspinal arterio-venous fistula in children: two more cases of an exceptional malformation. *Clin Pract* 2015; **5**: 707. doi: <https://doi.org/10.4081/cp.2015.707>
- Meisel HJ, Lasjaunias P, Brock M. Modern management of spinal and spinal cord vascular lesions. *Minim Invasive Neurosurg* 1995; **38**: 138–45. doi: <https://doi.org/10.1055/s-2008-1053474>
- Hui F, Trossello MP, Meisel HJ, Alvarez H, Sequeira E, Lasjaunias P. Paraspinal arteriovenous shunts in children. *Neuroradiology* 1994; **36**: 69–73. doi: <https://doi.org/10.1007/BF00599202>
- Naidich TP, Castillo M, Cha S, Raybaud C, Smirniotopoulos JG, Kollias S. *Imaging of the spine: expert radiology series*. 1st ed. Elsevier Health Sciences; 2010.
- Choi IS. Spinal arteriovenous metamerick syndrome: angioarchitecture and their prognosis. *AJNR Am J Neuroradiol* 2013; **34**: 464–5. doi: <https://doi.org/10.3174/ajnr.A3318>
- Niimi Y, Berenstein A, Fernandez PM, Brismar JL, Song JK. Pediatric nonvertebral paraspinal arteriovenous fistulas along the segmental nerve: clinical, imaging, and therapeutic considerations. *J Neurosurg* 2005; **103**(2 Suppl): 156–62. doi: <https://doi.org/10.3171/ped.2005.103.2.0156>
- Cobb S. Haemangioma of the spinal cord: associated with skin naevi of the same metamere. *Ann Surg* 1915; **62**: 641–9.

28. Jacob AG, Driscoll DJ, Shaughnessy WJ, Stanson AW, Clay RP, Gloviczki P. Klippel-trénaunay syndrome: spectrum and management. *Mayo Clin Proc* 1998; **73**: 28–36. doi: [https://doi.org/10.1016/S0025-6196\(11\)63615-X](https://doi.org/10.1016/S0025-6196(11)63615-X)
29. Alomari AI, Chaudry G, Rodesch G, Burrows PE, Mulliken JB, Smith ER, et al. Complex spinal-paraspinal fast-flow lesions in CLOVES syndrome: analysis of clinical and imaging findings in 6 patients. *AJNR Am J Neuroradiol* 2011; **32**: 1812–7. doi: <https://doi.org/10.3174/ajnr.A2349>
30. Briganti F, Tedeschi E, Leone G, Marseglia M, Cicala D, Giamundo M, et al. Endovascular treatment of vertebro-vertebral arteriovenous fistula. A report of three cases and literature review. *Neuroradiol J* 2013; **26**: 339–46.
31. Farhat N, Desprechins B, Otto B, Ramaekers V, Seghaye MC. Paraspinal arterio-venous fistula in children: two more cases of an exceptional malformation. *Clin Pract* 2015; **5**: 707. doi: <https://doi.org/10.4081/cp.2015.707>
32. Grote EH, Voigt K. Clinical syndromes, natural history, and pathophysiology of vascular lesions of the spinal cord. *Neurosurg Clin N Am* 1999; **10**: 17–45.
33. Bao YH, Ling F. Classification and therapeutic modalities of spinal vascular malformations in 80 patients. *Neurosurgery* 1997; **40**: 75–81.
34. Merland JJ, Reizine D, Laurent A, Khayata M, Casasco A, Aymard A. Embolization of spinal vascular lesions. In: Vinuela F, Halbach VV, Dion J, eds. *Interventional neuroradiology: endovascular therapy of the central nervous system*. New York: Raven Press; 1992. pp. 153–65.
35. Anson JA, Spetzler RF. Classification of spinal cord arteriovenous malformations and implications for treatment. *BNI Quarterly* 1992; **8**: 2.
36. Heros RC, Debrun GM, Ojemann RG, Lasjaunias PL, Naessens PJ. Direct spinal arteriovenous fistula: a new type of spinal AVM. Case report. *J Neurosurg* 1986; **64**: 134–9. doi: <https://doi.org/10.3171/jns.1986.64.1.0134>
37. Malis LI. Arteriovenous malformations of the spinal cord. In: Youmans JR, ed. *Neurological surgery: A comprehensive reference guide to the diagnosis and management of neurosurgical problems*. vol. 3. Philadelphia: WB Saunders; 1982. pp. 1850–74.
38. Marsh WR. Vascular lesions of the spinal cord: history and classification. *Neurosurg Clin N Am* 1999; **10**: 1–8.
39. Hebold O. Aneurysmen der kleinsten rückenmarksgefäße. *Arch Psychiatr Nervenkr* 1885; **16**: 813–23. doi: <https://doi.org/10.1007/BF02057574>
40. Gaupp J. Casuistische Beiträge zur pathologischen Anatomie des Rückenmarks und seiner Häute. *Beitr Pathol Anat* 1888; **2**: 510–24.
41. Ashour R, Orbach DB. Lower vertebral-epidural spinal arteriovenous fistulas: a unique subtype of vertebrovertebral arteriovenous fistula, treatable with coil and Penumbra Occlusion Device embolization. *J Neurointerv Surg* 2016; **8**: 643–7. doi: <https://doi.org/10.1136/neurintsurg-2015-011677>
42. Fotso A, Aubert D, Saltoun K, Galli G, Bonneville JF, Bracard S. Congenital paravertebral arteriovenous fistula: a case report. *J Pediatr Surg* 2006; **41**: e21–e23. doi: <https://doi.org/10.1016/j.jpedsurg.2005.12.031>
43. Siddhartha W, Chavhan GB, Shrivastava M, Limaye US. Endovascular treatment for bilateral vertebral arteriovenous fistulas in neurofibromatosis 1. *Australas Radiol* 2003; **47**: 457–61. doi: <https://doi.org/10.1046/j.1440-1673.2003.01221.x>
44. Paolini S, Colonnese C, Galasso V, Morace R, Tola S, Esposito V, et al. Extradural arteriovenous fistulas involving the vertebral artery in neurofibromatosis Type 1. *J Neurosurg Spine* 2008; **8**: 181–5. doi: <https://doi.org/10.3171/SPI/2008/8/2/181>
45. Hughes DG, Alleyne CH. Rare giant traumatic cervical arteriovenous fistula in neurofibromatosis type 1 patient. *BMJ Case Rep* 2012; **2012**: bcr1220115354. doi: <https://doi.org/10.1136/bcr.12.2011.5354>
46. Krings T. Vascular malformations of the spine and spinal cord*: anatomy, classification, treatment. *Clin Neuroradiol* 2010; **20**: 5–24. doi: <https://doi.org/10.1007/s00062-010-9036-6>
47. Manelfe C, Lazorthes G, Roulleau J. Arteries de la dure-mère rachidienne chez l'homme. *Acta Radiol* 1972; **13**: 829–41.
48. Merland JJ, Riche MC, Chiras J. Les fistules arterioveineuses intracanales extra-medullaires a drainage veineux medullaire. *J Neuroradiol* 1980; **7**: 221–30.
49. Tadie M, Hernet J, Aaron C. Le dispositif protecteur anti-reflux des veins de la moelle. *Neurochirurgie* 1979; **25**: 28–30.
50. Gillilan LA. Veins of the spinal cord. Anatomic details; suggested clinical applications. *Neurology* 1970; **20**: 860–8. doi: <https://doi.org/10.1212/WNL.20.9.860>
51. Goyal M, Willinsky R, Montanera W, terBrugge K. Paravertebral arteriovenous malformations with epidural drainage: clinical spectrum, imaging features, and results of treatment. *AJNR Am J Neuroradiol* 1999; **20**: 749–55.
52. Heier LA, Lee BC. A dural spinal arteriovenous malformation with epidural venous drainage: a case report. *AJNR Am J Neuroradiol* 1987; **8**: 561–3.
53. Krings T, Geibprasert S, Luo CB, Bhattacharya JJ, Alvarez H, Lasjaunias P. Segmental neurovascular syndromes in children. *Neuroimaging Clin N Am* 2007; **17**: 245–58. doi: <https://doi.org/10.1016/j.nic.2007.02.006>
54. Kitagawa RS, Mawad ME, Whitehead WE, Curry DJ, Luersen TG, Jea A. Paraspinal arteriovenous malformations in children. *J Neurosurg Pediatr* 2009; **3**: 425–8. doi: <https://doi.org/10.3171/2009.2.PEDS08427>
55. Djindjian R. Clinical symptomatology and natural history of arteriovenous malformations of the spinal cord—a study of the clinical aspects and prognosis, based on 150 cases. In: Pia HW, Djindjian R, eds. *Spinal angiomas: advances in diagnosis and therapy*. New York: Springer; 1978. pp. 48–83.
56. Berenstein A, Lasjaunias P. Spinal cord arteriovenous malformations. In: Berenstein A, Lasjaunias P, eds. *Surgical neuroangiography: endovascular treatment of spine and spinal cord lesions*. vol. 5. New York: Springer; 1992. pp. 24–76.