Introduction

Spinal arteriovenous (AV) shunts are abnormal connections between arteries and veins of the spine with the passage of blood from arteries to veins without going through the capillary network. The precise diagnosis of spinal AV shunts is more difficult than that of cerebral arteriovenous malformations (AVM): the sizes of the lesions are very small; spinal angiography is technically challenging; and their angioarchitecture differs according to their site, type, and spinal level. Many classification systems have been reported and have changed over time in the literature. The purposes of this study were 1) to review historical changes in the classification of spinal AV shunts after the introduction of spinal angiography, 2) to present representative neuroradiological imaging of spinal AV shunts, and 3) to propose a practical and easy-to-follow classification system.

Materials and Methods

The PubMed database was searched for English literature regarding the classification of spinal AV shunts published after the introduction of spinal angiography (1967–2015). The classification systems of spinal AV shunts were tabulated in systematic order. The proposed classification system was described and representative angiographic and magnetic resonance images of spinal AV shunts were presented.

Results

The first description of a classification (1971)

In 1967, Di Chiro G (a neuroradiologist at the National Institute of Health, USA) and colleagues reported selective angiography of spinal AV shunts for the first time. In 1971, Di Chiro G described the first classification of spinal AV shunts based on angiographic findings: single coiled vessel type I, glomus type II, and juvenile type III (Table 1). The majority of single...
Historical Changes in the Classification of Spinal AV Shunts

In 1971, Dichiro G (a radiologist at the National Institute of Neurological Diseases, New York) reported a new classification of spinal AV shunts.

Table 1 Classification of spinal AV shunts reported by Dichiro (1971)

<table>
<thead>
<tr>
<th>Type</th>
<th>Academic name</th>
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</thead>
<tbody>
<tr>
<td>I</td>
<td>Single coiled vessel type</td>
</tr>
<tr>
<td>II</td>
<td>Glomus type</td>
</tr>
<tr>
<td>III</td>
<td>Juvenile type</td>
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</table>

In 1975, Yasargil MG (a neurosurgeon at the University of Zürich, Switzerland) and colleagues reported a surgical case series of spinal AV shunts. This report included 11 cervical lesions. All patients were diagnosed using angiography of the vertebral artery; however, the spatial resolution of the images was very low. These lesions presumably corresponded to intradural glomus type, juvenile type, or perimedullary AVF. In Yasargil's report, AV shunts were described as "extramedullary and intramedullary vascular malformations".

In 1977, Kendall BE (a neuroradiologist at the National Hospital for Nervous Diseases, England) and Logue V (a neurosurgeon at the same hospital) reported a surgical case series of spinal AV shunts. This report included eight thoracolumbar lesions and one sacral lesion. All patients were diagnosed using selective angiography of spinal segmental arteries. The spatial resolution of the images was higher in Kendall's report than in those by Dichiro G and Yasargil. The angiographic findings of eight thoracolumbar lesions corresponded to single coiled vessel type (dural AVFs), while the remaining sacral lesion corresponded to an intradural perimedullary AVF at the filum terminale. In Kendall's report, the AV shunt was described as "a spinal epidural angiomatous malformation draining into intrathecal veins".

In 1983, Cogen P (a neurosurgeon at the Columbia-Presbyterian Medical Center, USA) reported a surgical case series of spinal AV shunts. This report included one cervical and five thoracolumbar lesions. The angiographic and surgical findings of these lesions corresponded to glomus AVM.

In 1983, Oldfield EH (a neurosurgeon at the National Institute of Health, USA), Dichiro G, and colleagues reported a surgical case series of spinal AV shunts. This report included six thoracolumbar lesions. The angiographic findings of this series corresponded to single coiled vessel type (dural AVFs). Occlusion of the proximal intradural arterialized vein was performed on five patients, whereas stripping of the intradural vein over multiple spinal segments was performed on the remaining patient. They concluded that occlusion of the proximal vein was sufficient, with more extensive removal of the vein being unnecessary.

In 1984, Symon L (a neurosurgeon at the National Hospital for Nervous Diseases, England), Kendall BE, and colleagues reported a large case series of 55 patients with dural AVF. Occlusion of the proximal intradural arterialized vein was performed on 50 patients, whereas decompressive laminectomy only was performed on the other five cases. Occlusion of the vein led to neurological improvements in 85% of 50 patients.

The second classification (1987)

In 1986, Hero RC (a neurosurgeon at Massachusetts General Hospital, USA) and colleagues reported a new type of spinal AV shunt that did not fit any type. They described a patient with a direct AV connection between the anterior spinal artery and vein that was located ventral to the spinal cord. Since this patient initially underwent spinal angiography in Belgium and was transferred to...
USA, Lasjaunias P (a French neuroradiologist) was listed as a co-author. Angiography revealed that the anterior spinal artery from the vertebral artery descended caudally and connected directly to the anterior spinal vein at the T3 level. The vein ascended cephalad and flowed into the vein in the posterior cranial fossa. This patient underwent direct surgery via anterior half corpectomy of the thoracic spine. The AVF was obliterated by dividing the two arterial feeders, the ascending and descending branches of the anterior spinal artery, both of which entered the vein through the fistula. On the 4th postoperative day, paraparesis deteriorated and spinal angiography revealed no filling of the ascending branch of the anterior spinal artery. In this case report, the AV shunt was described as a direct spinal AVF, Type IV.

The report by Hero was not the first case report of a direct spinal AVF. In 1977, Djindjian R (a neuroradiologist at the Lariboisière Hospital, France) and colleagues reported a series of intradural AV shunts fed by the anterior spinal artery. This report included six lumbosacral lesions. The angiographic findings of this case series did not fit any type of Di chiro G’s first generation classification, but corresponded to direct spinal AVF reported by Hero RC.

In 1987, Rosenblum B, Oldfield EH (neurosurgeons at the National Institute of Health, USA), Doppman JL, and Di chiro G (neuroradiologists at the same institution) reported a large case series of 81 patients with spinal AV shunts. In this report, spinal AV shunts were classified into four types: type I as dural AVF; type II as intradural intramedullary glomus AVM; type III as intradural intramedullary juvenile AVM; and type IV as direct AVF (Table 2). Medical illustrations were used to give a representation of these types. They compared dural AVFs and intradural AVMs in terms of clinical epidemiology, radiological imaging, and treatment outcomes.

The third classification (1993)

In 1993, Mourier KL and Merland JJ (neuroradiologists at the Lariboisière Hospital, France), and colleagues reported a series of intradural direct AVFs. In this report, AV shunts were described as perimedullary AVFs; thus, intradural direct AVFs were referred to as perimedullary AVFs thereafter in the literature. This study included 35 patients, 25 of whom underwent endovascular embolization. Perimedullary AVFs were classified according to the number of feeding arteries and size of AVF into the following three subtypes: Type I with a single
Table 2 Classification of spinal AV shunts reported by Rosenblum (1987)

<table>
<thead>
<tr>
<th>Type</th>
<th>Academic name</th>
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<tbody>
<tr>
<td>I</td>
<td>Dural AVF</td>
</tr>
<tr>
<td>II</td>
<td>Intramedullary glomus AVM</td>
</tr>
<tr>
<td>III</td>
<td>Intramedullary juvenile AVM</td>
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<tr>
<td>IV</td>
<td>Intradural direct AVF</td>
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Table 3 Classification of spinal AV shunts reported by Mourier (1993)

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<tr>
<th>Academic name</th>
<th>Subtype</th>
<th>Feeder, AVF, and venous drainage</th>
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<tbody>
<tr>
<td>Perimedullary AVF</td>
<td>I</td>
<td>A single feeder and small AVF</td>
</tr>
<tr>
<td></td>
<td>II</td>
<td>Multiple feeders and medium AVFs</td>
</tr>
<tr>
<td></td>
<td>III</td>
<td>Multiple feeders and a giant AVF</td>
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</table>

was performed. These five lesions corresponded to perimedullary AVF subtype I.

The Fourth and fifth classifications (2002)

In 2002, Spetzler RF (a neurosurgeon at the Barrow Neurological Institute, USA) reported a modified classification of spinal vascular malformations that changed the conventional concept of lesions (Table 4).<ref> He classified spinal vascular malformations including cavernous malformations and spinal AV shunts on the basis of his surgical experience of more than 130 lesions (89 spinal AV shunts). From the standpoint of a neurosurgeon, he classified spinal AV shunts based on anatomical locations between the lesion and spinal cord. Since spinal dural AVFs commonly develop at the dorsolateral portion of the dura mater, dural AVFs were defined as dorsal AVFs. In contrast, because perimedullary AVFs commonly develop at the ventral portion of the spinal cord, perimedullary AVFs were defined...
as ventral AVFs. Intramedullary glomus AVMs were defined as compact AVMs, and intramedullary juvenile AVMs as extradural-intradural AVMs. In addition, three new types were described: extradural AVFs, diffuse type AVMs, and conus AVMs. Extradural AVFs in the Spetzler classification were cervical lesions that were fed by the extradural branch of the vertebral artery and drained into the extradural venous plexus. In this lesion, although the spinal cord was severely compressed by the enlarged venous plexus, there was no venous congestion because there was no intradural venous drainage. Diffuse AVMs were intramedullary AVMs with subpial

<table>
<thead>
<tr>
<th>Types of AV shunts</th>
<th>Academic name</th>
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<tbody>
<tr>
<td>AVF</td>
<td>Extradural AVF</td>
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<tr>
<td></td>
<td>Intradural ventral AVF</td>
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<tr>
<td></td>
<td>Intradural dorsal AVF</td>
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<tr>
<td>AVM</td>
<td>Extradural-intradural AVM</td>
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<tr>
<td></td>
<td>Intradural compact AVM</td>
</tr>
<tr>
<td></td>
<td>Intradural diffuse AVM</td>
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<tr>
<td></td>
<td>Intradural conus AVM</td>
</tr>
</tbody>
</table>

Fig. 4 Intradural perimedullary AVF subtype II (multiple feeders, multiple medium AVFs). (A) A sagittal thoracic image using T2-weighted MR imaging showing congestion of the spinal cord (asterisk). (B) An angiogram of the thoracic intercostal artery showing intradural perimedullary AVF subtype II (arrowhead) fed by pial branches of the anterior and posterior spinal arteries (arrows). (C) An angiogram of the lumbar artery showing the same intradural perimedullary AVF (arrowhead) fed by pial branches of the posterior spinal arteries (arrow).

Fig. 5 Intradural perimedullary AVF subtype III (multiple feeders, single giant AVF). (A) A sagittal thoracic image using T2-weighted MR imaging showing compression of the spinal cord by an enlarged venous varix. (B) An angiogram of the thoracic intercostal artery showing intradural perimedullary AVF subtype III (arrowhead) fed by pial branches of the anterior and posterior spinal artery (arrows). (C) An angiogram of the thoracic intercostal artery showing the same intradural perimedullary AVF (arrowhead) fed by pial branches of the posterior spinal artery (arrow).

Table 4 Classification of spinal AV shunts reported by Spetzler (2002)
Historical Changes in the Classification of Spinal AV Shunts

Historical Changes in the Classification of Spinal AV Shunts

Extensions over multiple spinal segments. Conus AVMs were complex intramedullary AVMs that developed at the conus medullaris with multiple feeders and AV shunts. Surgical procedures for these lesions differed.

This report resulted in controversy. Barrow DL (a neurosurgeon at Emory University, USA) criticized the modified classification via a long letter to the Editor (1500 words); he anticipated the likelihood of interobserver variations among intramedullary AVMs between the compact and diffuse types. He also pointed out that complex AVMs are not always located at the conus medullaris; they may develop at different spinal levels. Lasjaunias P (a neuroradiologist at the Lariboisière Hospital, France) criticized the modified classification via a longer letter to the Editor (2000 words); his criticism was too acerbic, but worth reading.

In 2002, the same year in which Spetzler reported the modified classification, Rodesh G and Lasjaunias P (neuroradiologists at the Lariboisière Hospital, France) reported a new classification system from the standpoint of an endovascular interventionist. According to their type and size, spinal AV shunts were divided into AVMs and AVFs, with the latter being subdivided into macro and micro AVFs (Table 5). All spinal AV shunts corresponded to three categories: genetic hereditary lesions, genetic non-hereditary lesions, and single lesions.

Neither Spetzler’s nor Rodesh’s classification has been universally accepted because both classifications are biased toward either microsurgery or endovascular treatment.

The sixth and seventh classifications (2011)

In 2008, Geibprasert S, Lasjaunias P (a neuroradiologist at the Lariboisière Hospital, France), and colleagues reported a new classification system for cranio-spinal AV shunts in which AV shunts were divided into ventral, lateral, and dorsal epidural groups (Table 6). In this classification, which included intracranial and spinal AVFs, extradural AVFs corresponded to the ventral epidural group. In this report, the ventral epidural group was not associated with intradural venous drainage. These angiographic characteristics do not fit subsequent reports, as described below.

In 2009, Clarke MJ (a neurosurgeon at the Mayo Clinic, USA) reported a surgical case series of extradural AVFs. This report included six lumbosacral lesions that presented with venous congestion of the spinal cord because of the presence of intradural venous drainage. These six AVFs differed from Spetzler’s extradural AVFs, which did not have intradural venous drainage.

Table 5 Classification of spinal AV shunts reported by Rodesh (2002)

<table>
<thead>
<tr>
<th>Types of AV shunts</th>
<th>Academic name</th>
</tr>
</thead>
<tbody>
<tr>
<td>AVM</td>
<td></td>
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<tr>
<td>AVF</td>
<td>Micro AVF</td>
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<td></td>
<td>Macro AVF</td>
</tr>
<tr>
<td>Genetic classification</td>
<td>Genetic hereditary lesions</td>
</tr>
<tr>
<td></td>
<td>Genetic non-hereditary lesions</td>
</tr>
<tr>
<td></td>
<td>Single lesions</td>
</tr>
</tbody>
</table>

Table 6 Classification of cranio-spinal dural AV shunts reported by Geibprasert (2008)

<table>
<thead>
<tr>
<th>Types of AV shunts</th>
<th>Academic name</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventral epidural group</td>
<td>Ventral epidural shunts</td>
</tr>
<tr>
<td>Dorsal epidural group</td>
<td>Dorsal epidural shunts</td>
</tr>
<tr>
<td>Lateral epidural group</td>
<td>Lateral epidural shunts</td>
</tr>
</tbody>
</table>

Table 7 Classification of spinal extradural AVFs reported by Rangel-Castilla (2011)

<table>
<thead>
<tr>
<th>Types of AV shunts</th>
<th>Subtype</th>
<th>Venous drainage, neurological deficit</th>
</tr>
</thead>
<tbody>
<tr>
<td>Extraludal AVF</td>
<td>A</td>
<td>w intradural venous drainage</td>
</tr>
<tr>
<td></td>
<td>B1</td>
<td>w/o intradural venous drainage, w neurological deficits</td>
</tr>
<tr>
<td></td>
<td>B2</td>
<td>w/o intradural venous drainage, w/o neurological deficits</td>
</tr>
</tbody>
</table>

In 2011, Rangel-Castilla L (a neurosurgeon at the Methodist Hospital, USA), Diaz OM (a neuroradiologist at the same hospital), and colleagues reported an endovascular case series of extradural AVFs. This report included 8 lesions in 7 patients: 1 cervical, 1 thoracic, and 6 lumbar lesions. In this report, four lesions were associated with congestive myelopathy, three were compressive radiculopathy, and the remaining 1 was asymptomatic. Extradural AVFs were subdivided into three types: types A, B1, and B2 with or without intradural venous drainage and with or without neurological deficits (Table 7).

In 2012, the author conducted a systematic review on extradural AVFs in order to clarify the clinical and neuroradiological characteristics of these lesions. Forty-five cases were found in the PubMed database between 1990 and 2011. Type A spinal extradural AVFs were diagnosed in patients in their 6th decade.

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of life and exhibited diffuse high signal intensity in
the spinal cord on T2-weighted magnetic resonance
(MR) images with no mass effect. They also commonly
occurred in the thoracolumbar and lumbar regions.
In contrast, type B lesions exhibited a severe mass
effect due to an enlarged extradural venous plexus,
and commonly occurred in the cervical and upper
thoracic regions.

Representative cases of extradural AVFs, subtypes
A and B, are shown in Fig. 6.

In 2013, Niizuma K, Endo T (a neurosurgeon at
Tohoku University, Japan), and colleagues reported
a surgical case series of 5 type A extradural AVFs.\(^{23}\)
In the same year, Kiyosue H (a neuroradiologist at
Oita University, Japan) reported an endovascular
case series of 5 type A extradural AVFs referred to
as ventral epidural AVFs.\(^{24}\)

Discussion

Historical changes in the classification of spinal AV
shunts in the angiography era were reviewed herein.
There were seven major classification systems based
on historical reports (Tables 1–7): the first description
of spinal AV shunts diagnosed and classified using
spinal angiography in 1971; the second classification
system based on a case report of intradural direct
perimedullary AVFs treated by microsurgery in
1987; the third classification system based on a case
series of intradural perimedullary AVFs treated by
endovascular interventions in 1993; the fourth and
fifth classification systems based on a case series
of spinal AVFs and AVMs treated by microsurgery
or endovascular interventions in 2002; the sixth
classification based on a case series of cranio-
spinal dural AV shunts in 2009; and the seventh
classification based on a case series of extradural
AVFs treated by microsurgery and endovascular
interventions in 2011. By learning the historical
background, we may obtain a clearer understanding
of the complex and confusing classification system
of spinal AV shunts.

As shown in Table 8, the author proposes a practical
and easy-to-follow classification system based on
the historical background. Spinal AV shunts were
classified into five types: type I as dural AVF; type II

![Fig. 6 Extradural AVF. (A) A sagittal image using T2-weighted MR imaging showing congestion of the spinal cord (asterisk). (B) An antero-posterior angiogram of the lumbar artery showing the feeding pial arteries (white arrow), fistula (white arrowhead), venous pouch (black arrow), and draining vein (black arrowhead). (C) A sagittal image using T2-weighted MR imaging showing compression of the spinal cord by the extradural venous plexus (asterisk). (D) A lateral angiogram of the ascending cervical artery showing the feeding branch of the ascending cervical artery (white arrow) and fistula (white arrowhead).](image-url)
Historical Changes in the Classification of Spinal AV Shunts

Table 8 Proposed classification of spinal AV shunts by the author (2016)

<table>
<thead>
<tr>
<th>Type</th>
<th>Academic name</th>
<th>Subtype</th>
<th>Feeder, AVF, and venous drainage</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Dural AVF</td>
<td></td>
<td></td>
</tr>
<tr>
<td>II</td>
<td>Intramedullary glomus AVM</td>
<td></td>
<td></td>
</tr>
<tr>
<td>III</td>
<td>Intramedullary juvenile AVM</td>
<td></td>
<td></td>
</tr>
<tr>
<td>IV</td>
<td>Perimedullary AVF</td>
<td>IVa</td>
<td>A single feeder and small AVF</td>
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<td></td>
<td></td>
<td>IVb</td>
<td>Multiple feeders and medium AVFs</td>
</tr>
<tr>
<td></td>
<td></td>
<td>IVc</td>
<td>Multiple feeders and a giant AVF</td>
</tr>
<tr>
<td>V</td>
<td>Extradural AVF</td>
<td>Va</td>
<td>w intradural venous drainage</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Vb</td>
<td>w/o intradural venous drainage</td>
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</table>

as intradural intramedullary glomus AVM; type III as intradural intramedullary juvenile AVM; type IV as perimedullary AVF; and type V as extradural AVF. Perimedullary AVFs were subdivided into Subtypes IVa, IVb, and IVc and extradural AVFs were subdivided into Subtypes Va and Vb (Fig. 7).

The main advantage of the proposed classification system is that it is based on the classic second classification system with the type numbers i–iV, which have been the most widely used in the literature (Table 2). Extradural AVFs were added as a new type, which resulted in a type i–V classification system. Since the type numbers in the classification are consistent with those reported previously, treatment results may be easily compared among historical and current reports. In this classification system, perimedullary AVFs and extradural AVFs are subdivided into subtypes according to the feeding artery, the sizes of AV shunts, and drainage system. The advantage of this subclassification is that treatment approaches may be indicated based on the subtypes. Type IVa perimedullary AVFs are more amenable to microsurgery, and type IVc perimedullary AVFs to endovascular embolization. Type Va extradural AVFs may be treated by microsurgery and endovascular embolization, whereas type Vb extradural AVFs are more amenable to endovascular embolization. The proposed classification system is suitable for planning treatment strategies using microsurgical and/or endovascular interventions because it is based on the microvascular anatomy and hemodynamics of AV shunts.

However, this classification system also has disadvantages. We need to consider that spinal AV shunts differ not only according to the site and type of the AV shunt, but also to the spinal level at which the AV shunt develops. For example, the angioarchitecture of dural AVFs in the thoracic region differs from that in conus/lumbar lesions.25) The proposed classification system does not include anatomical differences at each spinal level. Furthermore, there may be another type of spinal AV shunt that does

Fig. 7 Illustration showing the different types of spinal arteriovenous shunts (Type I–V). Arrows indicates the arteriovenous connection(s).

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not fit any type of this classification. For example, the angioarchitecture of spinal AV shunts in the cervical region is complex. Further studies on the complex angioarchitecture of this rare disease are needed in more cases by analyzing neuroradiological imaging with high spatial and temporal resolution as well as surgical findings.

**Conclusion**

There were seven major classification systems based on historical reports, based on which the author proposed the current classification system according to the sites (dural, intradural, and extradural) and types (AVF and AVM) of AV shunts. By learning the historical background, we may obtain a clearer understanding of the complex and confusing classification system of spinal AV shunts.

**Conflicts of Interest Disclosure**

The author declares that there are no conflicts of interest.

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