# Perimedullary arteriovenous fistula classification

- Arteriovenous Malformations of the Central Nervous System
- Transient fourth nerve palsy preceding cervical myelopathy due to carotid-cavernous fistula
- Angioarchitecture Classification and Treatment Modalities of Craniocervical Junction Arteriovenous Fistulas: A Cohort Study of 155 Patients
- Vascular Malformations of the Spinal Cord in Children
- Intradural spinal cord arteriovenous shunts in a personal series of 210 patients: novel classification with emphasis on anatomical disposition and angioarchitectonic distribution, related to spinal cord histogenetic units
- Perimedullary arteriovenous fistulas of the craniovertebral junction: A systematic review
- Surgical Management for Craniocervical Junction Arteriovenous Fistula Targeting the Intradural Feeder
- Cervical spinal arteriovenous fistula with ventral perimedullary venous drainage

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Subtype I: single arterial supply (ASA), single small fistula, slow ascending perimedullary venous drainage

subtype II: multiple arterial supply (ASA and PSA), multiple medium fistulae, slow ascending perimedullary venous drainage

subtype III: multiple arterial supply (ASA and PSA), single giant fistula, large ectatic venous drainage

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## Type IV perimedullary arteriovenous fistula

According to the generally accepted Anson and Spetzler classification, three subtypes are identified according to shunt flow and degree of vascular enlargement.

In type IVa PMAVF, there is a slow-flow single shunt between a non-dilated anterior or posterolateral spinal artery and a spinal vein.

Type IVb lesions show greater flow than in type IVa and an ampullary dilatation of the venous side of the shunt. Increased shunt flow causes dilated tortuous intradural veins. PMAVFs generally have more than one feeder, usually a dilated anterior spinal artery and one or two posterior spinal arteries.

Type IVc PMAVFs, called giant perimedullary AVFs, have multiple high-flow dilated feeding arteries and gross dilatation of draining veins; varices or true venous aneurysms are encountered either near the shunt or more distally [1)].

Sometimes there is difficulty in accurately labeling type IVa and IVb lesions. A revised classification by Rodesch et al. simplifies the task and separates PMAVFs into macro-AVF and micro-AVF. Macro-AVFs are high-flow direct shunts fed by one or more spinal cord arteries ending in a venous ectasia with

secondary perimedullary venous drainage. Micro-AVFs are small lesions fed by one or more slightly enlarged arteries draining into veins that are not ectatic [2].

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Mizutani et al. proposed another vision of ID-SAVSs allowing a reappraised classification based on analysis of the anatomical disposition, angioarchitecture, and histogenetic location of these vascular malformations.

Methods: The radiological and clinical records of 210 patients with ID-SAVSs were retrospectively reviewed, considering their localization, vascular architectonics, and correlation with the 5 histogenetic units of the spinal cord. Among these, 183 files with complete data allowed precise analysis of the ID-SAVSs.

Results: Among these 183 files (162 and 21 cases with single and multiple lesions, respectively), different entities were identified: 13 pial macro arteriovenous fistulas (MAVFs), 92 pial micro arteriovenous fistulas (mAVFs), 33 superficial pial niduses, and 69 intramedullary niduses. Thirteen sulcal shunts (either fistulas or niduses) were considered subtypes of pial lesions. Among the 21 multiple cases, 11 were monomyelomeric while 10 were multimyelomeric. Pial lesions, either fistulas or niduses, were dominantly vascularized by pial arteries (anterior or posterior depending on the localization of the shunt) and occasionally (except for MAVFs) by transmedullary arteries. Pial niduses occasionally extended into the funiculus by recruiting intrinsic veins or by extension of the nidus itself inside the white matter. Intramedullary niduses were always vascularized by both centrifugal and centripetal feeders, respectively, from sulcal arteries (SAs) and pial arteries. Sulcal lesions are pial lesions located within the ventral median sulcus and vascularized by SAs and veins. Single or multiple ID-SAVSs can be part of various syndromes such as hereditary hemorrhagic telangiectasia, Parkes-Weber, RASA1, CLOVES, and spinal arteriovenous metameric syndromes. Histogenetic analyses revealed a specific distribution of each ID-SAVS in the 5 histogenetic units of the spinal cord: intramedullary niduses were found almost equally from cervical to thoracic units, while MAVFs and mAVFs were mostly found from thoracic to postcrural ones. Pial niduses showed intermediate features between intramedullary and fistulous lesions and were mostly distributed from brachial to crural segments.

Conclusions: Precise analysis of the anatomical disposition of ID-SAVSs in relation to functional histogenetic units allows a better understanding of these lesions and improved therapeutic management [3].

# Perimedullary arteriovenous fistulas of the craniovertebral junction

A systematic literature search was conducted using MEDLINE, Scopus, and Google Scholar databases, searching for the following combined MeSH terms: (perimedullary arteriovenous fistula OR dural arteriovenous shunt) AND (craniocervical junction OR craniovertebral junction). We also present an emblematic case of PMAVF at the level of the craniovertebral junction associated to a venous pseudoaneurysm. A total of 31 published studies were identified; 10 were rejected from our review because they did not match our inclusion criteria. Our case was not included in the systematic review. We selected 21 studies for this systematic review with a total of 58 patients, including 20 females (34.5%) and 38 males (65.5%), with a female/male ratio of 1:1.9. Thirty-nine out of 58 patients

underwent surgical treatment (67.2%), 15 out of 58 patients were treated with endovascular approach (25.8%), 3 out of 58 patients underwent combined treatment (5.2%), and only 1 patient was managed conservatively (1.7%). An improved outcome was reported in 94.8% of cases (55 out of 58 patients), whereas 3 out of 58 patients (5.2%) were moderately disabled after surgery and endovascular treatment. In literature, hemorrhagic presentation is reported as the most common onset (subarachnoid hemorrhage in 63% and intramedullary hemorrhage in 10%), frequently caused either by venous dilation, due to an ascending drainage pathway into an intracranial vein, or by the higher venous flow rates that can be associated with intracranial drainage. Hiramatsu and Sato stated that arterial feeders from the anterior spinal artery (ASA) and aneurysmal dilations are associated with hemorrhagic presentation. In agreement with the classification by Hiramatsu, we defined the PMAVF of the CVJ as a vascular lesion fed by the radiculomeningeal arteries from the vertebral artery and the spinal pial arteries from the ASA and/or lateral spinal artery. Considering the anatomical characteristics, we referred to our patient as affected by PMAVF, even if it was difficult to precisely localize the arteriovenous shunts because of the complex angioarchitecture of the fine feeding arteries and draining veins, but we presumed that the shunt was located in the point of major difference in vessel size between the feeding arteries and draining veins. PMAVFs of CVJ are rare pathologies of challenging management. The best diagnostic workup and treatment are still controversial: more studies are needed to compare different therapeutic strategies concerning both long-term occlusion rates and outcomes [4].

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Cho KT, Lee DY, Chung CK, Han MH, Kim HJ. Treatment of spinal cord perimedullary arteriovenous fistula: embolization versus surgery. Neurosurgery. 2005 Feb;56(2):232-41; discussion 232-41. Review. PubMed PMID: 15670371.
2)
Rodesch G, Hurth M, Alvarez H, Tadie M, Lasjaunias P. Spinal cord intradural arteriovenous fistulae: anatomic, clinical, and therapeutic considerations in a series of 32 consecutive patients seen between 1981 and 2000 with emphasis on endovascular therapy. Neurosurgery. 2005 Nov;57(5):973-83; discussion 973-83. PubMed PMID: 16284566.
3)
Mizutani K, Consoli A, Maria FD, Condette Auliac S, Boulin A, Coskun O, Gratieux J, Rodesch G. Intradural spinal cord arteriovenous shunts in a personal series of 210 patients: novel classification with emphasis on anatomical disposition and angioarchitectonic distribution, related to spinal cord histogenetic units. J Neurosurg Spine. 2021 Apr 2;34(6):920-930. doi: 10.3171/2020.9.SPINE201258. PMID: 33799293.
4)
Umana GE, Scalia G, Chaurasia B, Fricia M, Passanisi M, Graziano F, Nicoletti GF, Cicero S. Perimedullary arteriovenous fistulas of the craniovertebral junction: A systematic review. J Craniovertebr Junction Spine. 2020 Jul-Sep;11(3):157-162. doi: 10.4103/jcvjs.JCVJS_106_20. Epub 2020 Aug 14. PMID: 33100763; PMCID: PMC7546045.