Dural Arteriovenous Shunts
A New Classification of Craniospinal Epidural Venous Anatomical Bases and Clinical Correlations

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Background and Purpose—The craniospinal epidural spaces can be categorized into 3 different compartments related to their specific drainage role of the bone and central nervous system, the ventral epidural, dorsal epidural, and lateral epidural groups. We propose this new classification system for dural arteriovenous shunts and compare demographic, angiographic, and clinical characteristics of dural arteriovenous shunts that develop in these 3 different locations.

Methods—Three hundred consecutive cases (159 females, 141 males; mean age: 47 years; range, 0 to 87 years) were reviewed for patient demographics, clinical presentation, multiplicity, presence of cortical and spinal venous reflux, and outflow restrictions and classified into the 3 mentioned groups.

Results—The ventral epidural group (n=150) showed a female predominance, more benign clinical presentations, lower rate of cortical and spinal venous reflux, and no cortical and spinal venous reflux without restriction of the venous outflow. The dorsal epidural group (n=67) had a lower mean age and a higher rate of multiplicity. The lateral epidural group (n=63) presented later in life with a male predominance, more aggressive clinical presentations, and cortical and spinal venous reflux without evidence of venous outflow restriction. All differences were statistically significant (P<0.001).

Conclusion—Dural arteriovenous shunts predictably drain either in pial veins or craniofugally depending on the compartment involved by the dural arteriovenous shunt. Associated conditions (outflow restrictions, high-flow shunts) may change that draining pattern. The significant differences between the groups of the new classification support the hypothesis of biological and/or developmental differences in each epidural region and suggest that dural arteriovenous shunts are a heterogeneous group of diseases. (Stroke. 2008;39:2783-2794.)

Key Words: classification ▪ cranial dural arteriovenous fistula ▪ epidemiology ▪ epidural veins, clinical aspects ▪ spinal dural arteriovenous fistula

Dural arteriovenous shunts or fistulas (DAVSs or DAVFs) are a group of diseases that share involvement of the epidural space and adjacent dura mater and bony structures.

Different classifications have been proposed for these lesions,1-3 but the most widely used are those that focus on the presence or absence of cortical venous reflux; it is generally accepted that this angiographic finding constitutes the major clinical consideration regarding the natural history and, therefore, the therapeutic strategies for these lesions.4

Until now, the classification systems, being based on the various topographies, arterial feeders, and the drainage pattern, have been mainly descriptive. The different role played by each epidural area involved, as revealed during development and embryology of the venous system, were not considered. Issues concerning spinal versus cranial homologies or classic sex dominance in some localizations were not specifically addressed.

The basis for the new classification of DAVSs proposed in this study relies on the characteristics of venous afferent patterns of the various epidural spaces according to the evolution and embryological development of the venous drainage of the central nervous system and surrounding structures (vertebra, skull base, calvarium) by which 3 different epidural spaces can be differentiated as outlined subsequently.

Developmental Considerations of the Venous System of the Brain, Spine, and Adjacent Bony Structures

The notochord forms within the mesoderm in the third week and is considered a “patterning” embryonic structure of the
neural tube that regulates the development of the surrounding structures (such as nerves, blood vessels, and somites). Among other roles, it also controls the arterial versus venous identification of the major axial blood vessels and specifies a variety of cell types in forming primitive segments. It extends from the level of the sacrum to the basiphenoid and adjacent portion of the sphenoid wings; it will contribute to the future development of the vertebral bodies and bony structures within these regions.6

During the third week, angioblasts initially form small cell clusters (blood islands) within the embryonic and extraembryonic mesoderm. Recent studies have shown that arteries and veins are in part genetically determined by their endothelial types before the blood starts to flow inside the lumen.6

The neural tube starts to develop into the ventricular spaces at the fourth week.7 In the same period (corresponding to Paget’s 5-mm stage embryo), the blood traversing the capillary network at the surface of the brain is drained on either side by 3 “meningeal” (leptomeningeal) venousplexuses arranged craniocaudally (anterior, middle, and posterior); cranially, these open into a corresponding “primary head sinus” (epidural) and which will further develop by coalescence into “primitive sinuses.”7,8

The development of the choroid plexus starts at Paget’s 8- to 11-mm stage embryo10 with venous drainage into the median prosencephalic vein, which functions as the venous drainage route for the early immature brain and choroidplexuses. Secondarily, it may collect the deep cerebral venous afferents and basal vein, thereafter becoming the great cerebral vein or vein of Galen.

Development of the intrinsic venous drainage of the spinal cord starts after closure of the neural tube. Two longitudinal collector systems form in the subarachnoid space at the dorsal and ventral surface of the cord, later joining the epidural space laterally through numerous emissary-bridging veins. Contrary to arteries, these veins are not embryologically (metamerically) linked with the spinal nerves and thus do not follow a nerve root nor do they exit the subarachnoid space with them. Up to 40% of them exit through a separate dural foramen between the spinal nerves; thus, they should not be confused with the minute radicular veins draining the roots. They may instead be named emissary-bridging veins (transdural) to differentiate them from the so-called emissary veins (transosseous), which drain the intracranial venous sinuses outside of the skull.

The spinal venous system, being the basic one, will exhibit homologous features at its cranial counterpart for the 3 cephalic vesicles. However, the volume of developing brain will progressively alter the simple pattern of the spinal and spinal cord venous drainage. The emissary-bridging vein of the rhombencephalon is the medullary vein opening laterally into the condyloid vein at the level of the foramen magnum. For the mesencephalon, it is the petrous vein that drains the middle and upper brainstem. Longitudinal pial veins anastomoses will allow significant additional cranial contributions from the basal vein system. At the prosencephalic level, the base of the brain venous circle (interbasal venous anastomotic circle)11 and its outlets will represent the most cranial equivalent of the basic spinal and hindbrain pial venous system. During development, the earliest venous outlet of the anastomotic circle corresponds to the lateromesencephalic vein opening into the petrous vein and in the superior petrosal sinus as illustrated in the epsilon-shaped drainage in aneuysmal malformations of the vein of Galen.12 Posteriorly, the great cerebral vein (Galen vein) and anteriorly the deep sylvian vein openings correspond to phylogenetically more recent pathways as, respectively, shown by their late or postnatal development in humans.

During development, the draining veins of the archicerebellum (flocculonodular and lateral recess veins) and archicerebrum (anterior cerebral, uncate, and olfactory veins) will open into these older (previously existing) venous systems (basal vein, lateromesencephalic vein, pontine vein, and medullary vein) and thereafter through the petrosal vein or medullary emissary-bridging vein to the superior petrosal sinus or condyloid vein.

As the intrinsic venous system of the cortex further develops with its increase in cerebral territory (volume), it tends to be directed laterally to drain into the primitive tentorial sinus.9 The embryonic “tentorial sinus” at that point is likely to represent the most cranial portion of the lateral epidural venous system of the spine. Subsequently, the Breschet sinus, superior petrosal sinus, and straight sinuses are also included in that group. In short, the basal vein anastomotic circle; its afferent veins; and its mesencephalic, cavernous, and galenic (telencephalic and diencephalic) outlets are the cranial homologs of the emissary-bridging veins of the hindbrain and spinal cord.13

Considering the volume increase of the cerebrum and cerebellum (paleo- and neopallium) observed throughout phylogeny, the venous drainage will finally require additional outlets, which will be illustrated by the appearance of new dorsal pathways, like the superior sagittal (for the brain) and transverse sinuses (for the cerebellum) that are not present at the spinal level.

The development of the cranial vault sinuses into the epidural space results from the coalescence of the leptomeningeal vein contribution from the central nervous system with the osseous venous drainage. The secondary modeling into the adult-type sinuses is largely dependent on the growth of the skull.8,9 At the spine and skull, 2 types of bone can be distinguished, the endochondral type (cartilaginous bone–vertebral bodies, basioccipital, basisphenoid, and the petromastoid bone) and the intramembranous type (membranous bone–spinosus process, laminae, and cranial vault).13–15 The creation of the dorsal midline-located sinuses and confluences is, nevertheless, linked to the falces (cerebri and cerebelli) and the tentorium cerebelli16; both are associated with an epidural confluence and coalescence of the venous vascular spaces at their bony attachment later to become the superior sagittal sinus, medial occipital sinus, and transverse sinuses.17 Their role in the venous drainage of the paleo- and neocerebrum and paleo- and neocerebellum is a recent phylogenetic acquisition; they will further evolve postnatally after regression of the medial occipital sinus and maturation of the jugular bulbs.

The sigmoid sinuses and jugular bulbs are dependent on the growth of the petrous bone and skull base as they collect
petromastoid veins and do not receive any direct afferent from the leptomeningeal space. Thus, they are rather behaving like a conduit to bring the transverse and medial occipital sinus blood to the jugular vein.

The cavernous plexus at birth and the basilar venous plexus do not drain any cerebral vein either. After birth, the so-called cavernous capture may bring the remnant of the tentorial sinus or the deep sylvian vein to open into the cavernous plexus. The sigmoid sinus and cavernous and basilar plexuses therefore correspond to the ventrally located venous plexus of the spine. They share large midline communications as well as their unique role in draining the spongiosus parts of the related bones. Because the cranial sutures serve as intramembranous growth sites, the evolution of the dorsal dural sinuses closely follows development of the adjacent sutures.

In summary, 3 main observations of the venous development of the brain and spinal cord during embryology therefore contribute to the generation of the new classification:

1. The venous system of the notochord and corresponding sclerotome extends from the basisphenoid (cavernous plexus) to the sacrum and gives rise to the ventral epidural drainage group. It collects the blood from spongiosus bony structures and has no primary role in the drainage of the central nervous system.

2. The dorsal epidural venous space is normally poorly developed at the spinal level. Presence of dorsally located dural sinuses intracranially is therefore the major difference between the venous systems of the brain and spine. Their formation is linked to the appearance, during evolution, of the dural falces and tentorium and associated with the development of the paleo- and neopallial structures. These sinuses result from the confluence in the epidural space of 2 different venous systems: the osseous system draining the cranial vault and the leptomeningeal system draining the brain.

3. The veins draining the central nervous system (both spinal and cranial) are not related to the peripheral nerves, as are the arteries; these “emissary-bridging veins” join the lateral epidural venous spaces as connecting drainage system. This leptomeningeal venous drainage has no direct confluent communication with the ventral and dorsal epidural venous plexuses, which drain the skull and spine.

Although the epidural space changes its configuration, from the spinal to the intracranial regions at the level of magnum foramen, it keeps the same analogous characteris-
tics. All 3 epidural venous spaces remain compartmented and converge secondarily to join the jugular, azygos, lumbar, or sacral, paraspinous venous collectors. We suggest that DAVSs in these 3 different areas will drain according to the specific role of the venous system of each region, unless there are associated conditions such as constraint placed on the venous outlet or high-flow arteriovenous shunts. These venous outlet restrictions (ie, occlusion or stenosis) can be either adjacent to the shunting area, most commonly from thrombosis rather than endothelial proliferation, and/or remote from the shunt such as seen in jugular bulb dysmaturation in pediatric patients.

New Classification

As pointed out previously, there are 3 venous subgroups according to the relationship of the epidural venous spaces with the afferent veins from the bone and central nervous system (Figure 1): the ventral epidural (VE) or osteo(cartilaginous)-epidural group, the dorsal epidural (DE) or osteo(membranous)-epidural group, and the lateral epidural (LE) or leptomeningeal–epidural group. Based on these spaces, DAVSs are therefore classified into the following 3 groups.

Ventral Epidural Shunts
(osteocartilaginous]-epidural)

Dural arteriovenous shunts in these regions involve mainly the epidural space and are in direct contact with the adjacent osseous structures that they may invade or recruit the blood supply from (Figures 2 and 3). The venous afferents of these regions are closely related to the bony structures and they drain outside the bony limits, thus resulting in no subarachnoid (spinal or cortical) venous reflux (CVR). However, some factors may precipitate development of CVR in these

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**Figure 2.** Spinal VE DAVSs (A). Lateral sacral artery angiogram. Anteroposterior view reveals spinal VE DAVSs at the sacrum (B) formerly known as “epidural arteriovenous fistula.” Venous drainage is related to the bone and therefore typically drains through the epidural plexus of the vertebral body without perimedullary reflux.
regions such as thrombosis within the epidural space surrounding, remote, or distal to the shunt. This will produce reflux in the lateral epidural space as seen in rare cases of ventral epidural spinal shunts with perimedullary venous reflux.19,20 CVR can also be encountered in high-flow shunts forcing the emissary-bridging vein opening after reflux in the lateral epidural space.21 In this group, the following locations are encountered: “vertebral body,” basioccipital, sigmoid sinus, petrous pyramid, basisphenoid and adjacent sphenoid wings, and related dural structures.

Dorsal Epidural Shunts (osteo[membranous]-epidural)
At the spinal level, they are exceptional and clinically present with epidural hematoma (Figures 4 and 5). The venous pressure within the dural sinuses is usually low, thus antegrade, craniofugal flow of the shunts is normally observed. CVR may occur after associated venous outflow restriction, or with high-flow shunts; in case of variations of the venous opening into this system such as presence of an accessory epidural sinus, distal restriction of the sinus opening will rapidly produce CVR. In this group, the following DAVS locations are encountered: “dorsal spinal epidural DAVS,” marginal sinus (dorsal portion), medial occipital sinus, torcular, transverse and accessory epidural sinuses, and superior sagittal sinus.

Lateral Epidural Shunts
At the spinal level, these shunts correspond to the typical spinal DAVFs and are located lateral where the pial emissary bridging vein pierces the dura (Figures 6 and 7). Intracranial locations include DAVS draining into emissary-bridging veins of the brainstem and their homologs draining deep cerebral structures. The drainage is always directed to the cortical or perimedullary veins; therefore, the DAVS developing there are always considered to be aggressive.22–24 In this group, the following locations are encountered: spinal dural arteriovenous shunts, marginal sinus (lateral portion-foramen magnum) with the emissary-bridging vein to the condyloid vein, falcotentorial (vein of Galen), petrosal and

Figure 3. A, Internal carotid artery and (B) external carotid artery angiograms in lateral views reveal cranial VE DAVSs at the cavernous sinus in 2 different patients. Venous drainage is related to the bone at the skull base and therefore no CVR occurs as seen in the first patient (A) unless there is associated outflow restriction (B), ie, thrombosis of the jugular bulb in the second patient (*) causing severe CVR through the superficial middle cerebral vein.
basientorial, Breschet sinus, paracavernous region (embryonic tentorial sinus remnants), intraorbital shunts, and lamina cribrosa.

The goal of our study was to compare in 300 consecutive patients the demographic, angiographic, and clinical aspects of DAVSs that develop in these 3 different epidural spaces and to assess whether they truly present according to the predicted role of each epidural space in the venous drainage of the central nervous system and adjacent bony structures. Although the latter would confirm the validity of the classification system, the former may demonstrate that the 3 different DAVSs also constitute separate clinical entities with possible different age and sex distributions and pathomechanisms.

Materials and Methods
This is a retrospective review of 300 consecutive patients, 150 patients each from 2 different institutions (Ramathibodi Hospital, Bangkok, Thailand, during July 1998 to December 2006 and Bicêtre Hospital, Paris, France, during September 1992 to December 2006) for patient demographics, clinical symptoms, shunt localization, multiplicity, presence of CVR, and outflow modifications (stenosis/occlusion). The study was approved by the local ethics committees of both hospitals.

The clinical symptoms were separated into 2 groups, benign and aggressive.25 The benign group consists of incidental diagnosis, nonspecific headaches, cranial nerve deficits, chemosis/proptosis, midline bruit, mass lesions, and cardiac insufficiency. Aggressive symptoms included seizures, intracranial hemorrhage, motor or sensory deficits, visual field defects, aphasia, global neurological deficits (dementia, delayed psychomotor development, macrocrania), and other nonhemorrhagic neurological deficits such as incontinence.

On angiographic terms, benign and aggressive lesions were defined by absence or presence of CVR, respectively,26 and were grouped also using the classification systems of Borden27 and Cognard.3 Borden 1 (sinus drainage only), Cognard I (antegrade sinus drainage) and Iia (antegrade and retrograde sinus drainage)
were considered as “benign” DAVSs, whereas all the higher grades (which have cortical and spinal drainage with or without sinus drainage) were grouped as “aggressive” DAVSs.27

Venous outflow restrictions (stenosis/occlusion) were defined as adjacent to the shunt, remote from it, or both (mixed). In addition, we evaluated whether the restriction occurred proximal or distal to the shunt or both. The cavernous sinus lesions were excluded from this item, because nonvisualization of the outflow pathways could mean either thrombosis or compartmentalization of the cavernous sinus. However, if there was stenosis or occlusion of the venous system elsewhere, they were added and classified as a remote type. The spinal lesions were also excluded because nonvisualization of the draining spinal cord veins is part of the diagnosis of this location and reflects the spinal cord venous congestion leading to neurological symptoms.28 In addition, thrombosis or fibrosis of pial emissary-bridging veins of the cord occur with normal aging.8

The DAVSs were classified into the 3 groups of the new classification. Crosstabulations with the sex, age groups, clinical symptoms, presence of CVR, and types of venous outflow modifications were performed. Statistical significance was calculated for each group using $\chi^2$ and one-way analysis of variance tests.

Results

Of the 300 patients included in this article, the general patient data are summarized in Table 1. There were 150 patients (50%) who had lesions classified to the VE group, 67 patients (22.3%) in the DE group, and 63 patients (21%) in the LE group. There were 20 (6.7%) patients with multiplicity of lesions: 16 had both a VE and a distant DE DAVF, one had VE + LE types, and 3 patients had DE + LE types. When calculating the statistics to evaluate the differences for each of the 3 groups (VE, DE, LE), the patients having multiplicity of lesions were excluded. Refer to supplemental Table I for details of the Borden and Cognard classification and presence of CVR.

Male versus Female

There was a female predominance in patients having VE lesions (117 of 167 [70%]; $P<0.001$) and a strong male predominance in patients with LE-type lesions (54 of 67 [81%]; $P<0.001$). There was no definite sex predominance (female: male = 41:46; $P=0.225$) for the DE lesions.

Age

The mean consultation ages of patients with only VE, DE, and LE lesions were 51, 28, and 56 years, respectively. The mean age in the DE group was significantly lower than the
VE and LE lesions \((P<0.001)\) with 31 of 67 (46.3\%) being in the pediatric population. Patients with LE-type lesions also presented later in life with no patients presenting before the age of 30.

**Benign Versus Aggressive Clinical Symptoms**

In 150 patients of the VE group, 139 (92\%) had benign clinical symptoms, whereas in the LE group, 54 of 63 (86\%) patients had aggressive clinical symptoms \((P<0.001)\). There was a slight trend to more aggressive symptoms in the DE group (42 of 67 [63\%]) without statistical significance at 99\% CI \((P=0.038)\). Concerning the patients with multiplicity of DAVS, whenever a LE lesion was present in these patients, the symptoms were aggressive, demonstrating that these lesions are dominant in terms of risks for the patient, whereas in only 7 of 16 patients who had both VE and DE shunts, aggressive clinical symptoms were present.

**Multiplicity**

The DE shunts had a higher rate of multiple lesions, 31 of 86 (36\%), whereas only 20 of 167 (12\%) shunts of the VE type and 6 of 67 (9\%) shunts of the LE type had multiple lesions \((P<0.001)\).

**Comparison With Cognard/Borden**

Because the LE lesions are located within the laterally located epidural space, which drains primarily into the cortical or spinal veins, all of them were classified as aggressive and,
subsequently, had classifications of Types III, IV, and V according to Cognard and Type 3 according to Borden. If multiplicity of shunts was present, the lateral epidural location always determined the aggressiveness and could therefore be determined as the primary risk factor. Drainage pattern of the cranial VE and DE type of DAVSs was primarily through the sinuses, resulting in Types I, IIa, IIb, and IIab according to the Cognard classification and Type 1 and Type 2 according to the Borden classification depending on outflow restrictions. Refer to supplemental Tables II and III, available online at http://stroke.ahajournals.org, for a detailed comparison of the Cognard and Borden classifications with the new classification scheme.

Cortical/Spinal Venous Reflux
Cortical/spinal venous reflux was less common in patients with only VE shunts, being present in only 45 of 150 (30%) of the patients, all of the noncavernous shunts having stenosis/thrombosis of the venous outflow. The percentage of CVR increased in patients with DE and LE shunts, being 43 of 67 (64%) of the only DE shunts and all (100%) of the LE shunts ($P<0.001$). In the multiple type of VE+DE lesions, CVR was present in 13 of 16 (81%) cases.

Venous Outflow Restriction
In the noncavernous VE lesions, 39 of 52 cases (75%) had evidence of venous outflow restriction, which occurred surrounding the shunt in 30 of 39 cases (77%; $P<0.001$). Sixty of 67 cases (90%) with only DE lesions had stenosis/occlusion of the venous outflow and the majority (35 of 60 [58%]) were of the multiple type that had thrombosis surrounding and remote to the shunt location ($P<0.001$). In the 35 patients with nonspinal LE type of DAVSs, only 6 patients (17%) had restrictions of venous outflow, being remote from the shunt location in 4 of those ($P<0.001$).

In the VE group, no cortical or spinal reflux was present when there was no modification of the venous outflow pattern (13 of 13 [100%]). In the remaining patients in the VE group, the venous outflow restriction usually occurred surrounding the shunt and most of the patients with this type did not have

![Figure 7. Internal carotid artery angiogram (A) and occipital artery angiogram (B) reveal cranial LE DAVSs in 2 patients at the anterior cranial fossa (A) and tentorium (B), respectively. In these locations, CVR is always present. Branches of the ophthalmic artery (A) supply the ethmoidal LE DAVSs with drainage into ectatic venous pouches of the frontal cortical vein before entering the superior sagittal sinus (arrows). Stylomastoid branches of the occipital artery (B) supply the tentorial LE DAVSs and drain into the basal vein group, which joins the vein of Galen (arrowheads).](http://stroke.ahajournals.org/content/full/1/6/625/F7.large.jpg)
CVR (27 of 30 [90%]; \( P = 0.001 \)). There was also a higher chance of CVR when the restriction occurred surrounding the shunting zone (5 of 8 [63%]) or in mixed type patterns (31 of 35 [89%]) in this group than when the restriction occurred remote to the shunt (6 of 17 [35%]; \( P = 0.001 \)). All of the LE lesions drained directly into the cerebral or spinal cord veins with CVR and in the majority (29 of 35 [83%]) of the nonspinal patients, there was no venous outflow restriction. Refer to supplemental Table IV for tabulation of the data concerning the type of thrombosis in relation to CVR in each group.

In the DE shunts, the venous stenosis was usually distal to the shunt or of the mixed type with the rate of CVR being higher in the mixed type group (20 of 35 [57%] versus 22 of 25 [88%]; \( P = 0.001 \)). No statistical significance was found in the other groups. Refer to supplemental Table V for tabulation of the data concerning the location of thrombosis in relation to CVR in each group.

**Discussion**

**Ventral Epidural Shunts**

There was an approximately 2.3:1 female predominance in this group, which is also seen in other lesions involving the same regions such as cavernous aneurysms.\(^{29}\) The characteristics are summarized in Table 2. Cranial lesions usually present with benign clinical symptoms attributed to the low rate of CVR. In the spine, these shunts are usually asymptomatic; there have been only few case reports describing associated perimedullary reflux causing congestive myelopathy and stimulated considerations about a possible valve-like mechanisms normally impeding retrograde flow from the epidural plexus to perimedullary veins; however, in our opinion, it is more likely an extensive thrombosis of the normal epidural outlets that leads to retrograde drainage to perimedullary veins as a seldom variant of spinal VE arteriovenous shunts.

**Dorsal Epidural Shunts**

There was no significant sex predominance. The characteristics are summarized in Table 3. The infantile type and dural sinus malformations of the pediatric DAVSs are within this group,\(^{30,31}\) therefore significantly reducing the average age of patients compared with the other groups in which children are poorly represented. CVR is more frequently observed in patients who have venous outflow restriction; the more extensive the thrombosis or venous occlusion, the higher the percentage of CVR.

Multiplicity of the shunts was more commonly found in the DE group (36% increasing up to 46% with the pediatric group excluded), frequently within adjacent regions of the same epidural type. Multifocality of dural arteriovenous lesions in children has already been reported previously.\(^{32}\) Although the etiology of these shunts is not known, there seems to be clinical and radiological evidence that they are evolutionary lesions and it has been suggested that they may have been induced by venous sump from the sinus draining the DAVSs.\(^{32}\) In the adult population, multifocality may be due to venous hypertension, typically from sinus thrombosis,\(^{33}\) although the exact pathomechanism is as yet unclear. In the literature, it has been reported that the subgroup with multiple DAVFs had a 3 times higher rate of hemorrhagic presentation compared with a single DAVFs due to a higher rate of CVR.\(^{34}\)

**Lateral Epidural Shunts**

There was a strong male predominance, approximately 4:1, within this group. The characteristics are summarized in

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<th>Table 1. General Data of the 300 Patients</th>
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<tr>
<td>Demographics/Characteristics</td>
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<td>Sex</td>
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<td>Female</td>
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<td>Male</td>
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<td>Age, mean years (range)</td>
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<td>Clinical symptoms</td>
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<td>Benign</td>
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<td>Aggressive</td>
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<td>History of procoagulatory states, sinus thrombosis</td>
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<td>Multiple shunts</td>
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<td>Bilateral cavernous lesions</td>
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<td>CVR</td>
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<td>Venous outflow restriction (VOR)</td>
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<td>(excluded 93 cavernous and 28 spinal lesions)</td>
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<td>Type of VOR</td>
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<td>Adjacent (surrounding) to the shunt</td>
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<td>Remote to the shunt</td>
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<td>Mixed (both)</td>
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<td>Location of VOR</td>
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<td>Upstream to the shunt</td>
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<td>Distal to the shunt</td>
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<td>Both</td>
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<tr>
<th>Table 2. Ventral Epidural Shunts (osteo/cartilaginous)-epidural</th>
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<tr>
<td>Female predominance</td>
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<tr>
<td>No reflux unless</td>
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<td>Extensive thrombosis of epidural drainage</td>
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<td>High-flow shunts</td>
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<th>Table 3. Dorsal Epidural Shunts (osteo(membranous)-epidural)</th>
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<tr>
<td>No sex predominance</td>
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<tr>
<td>Lower mean age (pediatric group)</td>
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<td>Multiplicity</td>
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<td>Usually no CVR unless</td>
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<tr>
<td>Extensive thrombosis of the mixed type, both proximal and distal</td>
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<td>High-flow shunts</td>
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<td>Special anatomic variations; compartmentalization of the sinusal space if full coalescence of the venous lumen does not occur</td>
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<th>Table 4. Lateral Epidural Shunts</th>
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<td>Male predominance</td>
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<td>Present at later age</td>
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<td>Always aggressive lesion; CVR always present</td>
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Table 4. The patients also present at a later age compared with the other groups. As previously described in patients with spinal DAVFs, these shunts most likely manifest themselves after spinal cord draining veins have been obliterated by fibrosis from aging.35,36 The drainage into the cortical or perimedullary spinal cord veins results in increased pial venous pressure with a high rate of neurological symptoms ranging from venous infarctions, global neurological deficits to hemorrhage in the brain or congestive venous myelopathy in the cord.37,38 Due to the role played by veins in cranial locations, the shunts will involve the subarachnoid portion of the draining cortical vein causing venous ectasias that may be associated with hemorrhage from rupture of the venous pouches.22–24

Treatment Considerations

The decision to treat or not to treat still depends on the presence of cortical or spinal venous reflux regardless of the symptoms present.26 As multiple other previous reports have already emphasized, the presence of cerebral or spinal venous reflux should prompt immediate treatment, because their natural history is always aggressive with a high rate of morbidity and mortality.4,27,39

Conclusions

The roles of venous afferents of the 3 epidural spaces established during embryological development of the central nervous system and craniospinal structures are different. This reference to embryology allows analyzing both cranial and spinal homologs and, therefore, is the basis for a classification system that can be applied to all types of lesions developing in these spaces.

This new classification has the ability to predict the drainage of shunts located in different locations based on the craniospinal venous anatomy and the presence of associated venous outflow restriction (stenosis/occlusion), usually thrombosis, which could be either an etiology or comorbidity of the disease. Because venous thrombosis is uncommon in the general population, it may reflect an underlying biological disorder in these patients with possible different geographic (population) impact.

The significant difference of the patient characteristics (sex and age) between the groups of the new classification suggests biological/hormonal differences within the epidural venous system in each region and may point toward a different etiology of the 3 different groups of arteriovenous shunts. Grouping DAVFs into a single entity may hide the fundamental differences existing in each location. We presume that the new classification proposed in this study will contribute to a better understanding of these lesions and to their respective disease mechanisms.

Source of Funding

SG received a Research Grant of the World Federation of Interventional Neuroradiology (WFITN).

Disclosures

None.

References


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Stroke. 2008;39:2783-2794; originally published online July 17, 2008; doi: 10.1161/STROKEAHA.108.516757

Stroke is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0039-2499. Online ISSN: 1524-4628

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://stroke.ahajournals.org/content/39/10/2783

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