Pathomechanisms of Symptomatic Developmental Venous Anomalies

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**Background and Purpose**—Although it is generally accepted that developmental venous anomalies (DVAs) are benign vascular malformations, over the past years, we have seen patients with symptomatic DVAs. Therefore, we performed a retrospective study and a literature study to review how, when, and why DVAs can become clinically significant.

**Methods**—Charts and angiographic films of 17 patients with DVAs whose 18 vascular symptoms could be attributed to a DVA were selected from a neurovascular databank of our hospital. MRI had to be available to rule out any other associated disease. In the literature, 51 cases of well-documented symptomatic DVAs were found. Pathomechanisms were divided into mechanical and flow-related causes.

**Results**—Mechanical (obstructive or compressive) pathomechanisms accounted for 14 of 69 symptomatic patients resulting in hydrocephalus or nerve compression syndromes. Flow-related pathomechanisms (49 of 69 patients) could be subdivided into complications resulting from an increase of flow into the DVA (owing to an arteriovenous shunt using the DVA as the drainage route; n=19) or a decrease of outflow (n=26) or a remote shunt with increased venous pressure (n=4) leading to symptoms of venous congestion. In 6 cases, no specific pathomechanisms were detected.

**Conclusions**—Although DVAs should be considered benign, under rare circumstances, they can be symptomatic. DVAs, as extreme variations of normal venous drainage, may represent a more fragile venous drainage system that can be more easily affected by in- and outflow alterations. The integrity of the DVA needs to be preserved irrespective of the treatment that should be tailored to the specific pathomechanism. *(Stroke. 2008;39:3201-3215.)*

**Key Words:** arteriovenous shunting ■ compression ■ developmental venous anomaly ■ DVA ■ flow imbalance ■ hemorrhage ■ thrombosis ■ venous congestion

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Developmental venous anomalies (DVAs), that have been previously called venous angiomas, are extreme variations of normal transmedullary veins that are necessary for the drainage of white and gray matter. They consist of converging dilated medullary veins that drain centripetally and radially into a transcerebral collector that opens either into the superficial subcortical or deep pial veins. The DVAs have no proliferative potential, no direct arteriovenous shunts, and normal brain parenchyma between the dilated veins. DVAs serve as normal drainage routes of the brain and radially into a transcerebral collector that opens either into the superficial subcortical or deep pial veins. The integrity of the DVA needs to be preserved irrespective of the treatment that should be tailored to the specific pathomechanism.

DVAs are benign anatomic variations and are, therefore, usually incidentally discovered. Although in the past, different clinical symptoms were attributed to be caused by DVAs, MRI has changed the understanding of DVAs’ natural history and associated clinical symptoms; most hemorrhages are related to associated cavernomas rather than to the DVA, epilepsies are due to associated cortical dysplasias, and pseudotumoral effects can be secondary to associated lymphatic malformations.

Although it is thus generally accepted that DVAs are only rarely symptomatic, their exact clinical significance still remains controversial. Most series described the epidemiology, distribution, radiological characteristics, and associated conditions of DVAs. However, these studies did not differentiate whether symptoms arose from the DVA itself or rather from pathologies associated with the DVA (ie, cavernomas). The aim of this article is to describe, by the aid of a
retrospective series of cases and a review of the literature (after MRI has been introduced), how and when DVAs can become clinically significant. In all our patients and the cases from the literature, we systematically looked for the cause of the complication of the DVA. More specifically, we studied the relation of the DVA to neighboring structures and we analyzed the balance of the in- and outflow of the DVA. Therefore, we aimed at reviewing all possible pathomechanisms and describe potential therapeutic options.

**Table 1. Own Series of 17 Patients With 18 Episodes of Vascular Complications Attributable to a DVA**

<table>
<thead>
<tr>
<th>Patient Number</th>
<th>Age</th>
<th>Sex</th>
<th>Mechanism of Complication</th>
<th>Morphological Presentation</th>
<th>Clinical Symptoms</th>
<th>Topography</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1 year</td>
<td>F</td>
<td>Mechanical</td>
<td>Right proptosis</td>
<td>Proptosis and eye pain</td>
<td>Left temporal</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>2002—flow misbalance with outflow obstruction</td>
<td>2002—hemorrhage</td>
<td>2002—vertigo, ataxia</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>1 month</td>
<td>M</td>
<td>Flow misbalance with outflow obstruction</td>
<td>Hemorrhagic venous infarction</td>
<td>Seizures at birth</td>
<td>Left temporal</td>
</tr>
<tr>
<td>4</td>
<td>32 years</td>
<td>F</td>
<td>Flow misbalance with outflow obstruction</td>
<td>Venous congestion</td>
<td>Headache, Parinaud syndrome</td>
<td>Mesencephalon/cerebellum</td>
</tr>
<tr>
<td>5</td>
<td>8 months</td>
<td>F</td>
<td>Flow misbalance with outflow obstruction</td>
<td>Hemorrhagic venous infarction and SAH</td>
<td>Headache, seizures</td>
<td>Left temporal</td>
</tr>
<tr>
<td>6</td>
<td>11 months</td>
<td>F</td>
<td>Flow misbalance with outflow obstruction</td>
<td>SAH</td>
<td>Syncope and seizures</td>
<td>Right deep nuclei</td>
</tr>
<tr>
<td>7</td>
<td>5 years</td>
<td>F</td>
<td>Flow misbalance with outflow obstruction</td>
<td>Venous infarction</td>
<td>Seizures and left hemiparesis</td>
<td>Right temporal</td>
</tr>
<tr>
<td>8</td>
<td>29 years</td>
<td>M</td>
<td>Flow misbalance with outflow obstruction</td>
<td>Venous congestion</td>
<td>Headache, somnolence, aphasia</td>
<td>Left frontal</td>
</tr>
<tr>
<td>9</td>
<td>58 years</td>
<td>F</td>
<td>Flow misbalance with outflow obstruction</td>
<td>Venous congestion</td>
<td>Headache, right hemiparesis</td>
<td>Left deep ganglia</td>
</tr>
<tr>
<td>10</td>
<td>41 years</td>
<td>M</td>
<td>Flow misbalance with outflow restriction due to remote shunt</td>
<td>Hemorrhage</td>
<td>Headache, vomiting</td>
<td>Right temporal</td>
</tr>
<tr>
<td>11</td>
<td>9 years</td>
<td>M</td>
<td>Flow misbalance with increased inflow due to microshunt</td>
<td>Hemorrhage</td>
<td>Headache and seizures</td>
<td>Left temporal</td>
</tr>
<tr>
<td>13</td>
<td>24 years</td>
<td>M</td>
<td>Flow misbalance with increased inflow due to microshunt</td>
<td>Hemorrhage</td>
<td>Headache, left hemiparesis</td>
<td>Right frontal</td>
</tr>
<tr>
<td>14</td>
<td>8 years</td>
<td>M</td>
<td>Flow misbalance with increased inflow due to microshunt</td>
<td>Hemorrhage</td>
<td>Ataxia and somnolence</td>
<td>Right cerebellar hemisphere</td>
</tr>
<tr>
<td>15</td>
<td>2 days</td>
<td>F</td>
<td>Spontaneous/idiopathic</td>
<td>Hemorrhage</td>
<td>Seizures</td>
<td>Left frontal</td>
</tr>
<tr>
<td>16</td>
<td>32 years</td>
<td>F</td>
<td>Spontaneous/idiopathic</td>
<td>Hemorrhage</td>
<td>Headache</td>
<td>Left basal ganglia</td>
</tr>
<tr>
<td>17</td>
<td>42 years</td>
<td>F</td>
<td>Spontaneous/idiopathic</td>
<td>Venous infarction</td>
<td>Right hemiparesis</td>
<td>Left deep ganglia</td>
</tr>
</tbody>
</table>

(Continued)

**Methods**

Patients were selected after a retrospective search through the databank of our hospital into which, since 1989, patients were prospectively entered. To date (May 2007), there is a total of 4217 patients of which 80 patients were found whose principal diagnosis was “DVA.” Within those, 17 patients presenting with 18 direct vascular complication or a neurological symptom related directly to the region of the brain that is drained by the DVAs and its anatomic structure, diagnosed by angio-CT, MRI, and confirmed by digital angiography, were included in this series. We analyzed epidemio-
logical variables like age, gender, associated risk factors as well as clinical presentation, radiological data, treatments options, and follow-up. Considering the angioarchitecture of the DVA, we examined size, topography, venous drainage, morphology of medullary veins and venous collector, presence or absence of capillary ectasia, and medullary blush or associated pathology. Increased vascular transit time through the DVA, associated cerebral arteriovenous malformations (AVMs) or dural arteriovenous shunts remote or close to the DVA.

We restricted this review to complications considered to be directly related to DVAs to recognize under which conditions they could become symptomatic. The following exclusion criteria were therefore chosen: (1) unspecific symptoms like headaches or longstanding symptoms such as epilepsy were not considered if there was no MR evidence of a causative link to the DVA (such as congestive edema in the immediate vicinity);7,9–14 (2) MRI abnormalities without symptoms (T2* hypointensities, T2/flair hyperintensity), although related to DVAs, were not included;5,15,16 (3) patients with DVAs and symptomatic cavernomas because of their established natural history (hemorrhage, epilepsy, mass effect);5,14,17–25 and (4) patients and series presenting incomplete radiological or clinical data necessary to

Table 1. Continued

<table>
<thead>
<tr>
<th>Scan/MRI</th>
<th>Angioarchitecture/Venous Drainage</th>
<th>Risk Factors/Associated Conditions</th>
<th>Treatment/Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Focal atrophy</td>
<td>Bihemispheric and complex → frontal v. → ophthalmic v.</td>
<td>Stenosis of venous collector/blue rubber bleb nevus syndrome</td>
<td>Conservative</td>
</tr>
<tr>
<td>1998—hydrocephalus</td>
<td>Large and complex</td>
<td>Stenosis of venous collector, venous ectasia</td>
<td>Conservative/venous ectasia</td>
</tr>
<tr>
<td>2002—venous congestion</td>
<td>Bilateral cerebellum → left precentral cerebellar v. → VG</td>
<td>Stenosis of venous collector, venous ectasia</td>
<td>Conservative/normal development</td>
</tr>
<tr>
<td>Congestive edema surrounding the venous collector</td>
<td>Large and complex thalamostriate v. → ICV → subependymal collector → VG</td>
<td>Stenosis of venous collector, venous ectasia</td>
<td>Conservative/normal development</td>
</tr>
<tr>
<td>Venous congestion, edema in the DVA territory</td>
<td>Complex and large bilateral cerebellum → precentral cerebellar v. → VG</td>
<td>Thrombosis of venous collector (precentral cerebellar v.)</td>
<td>Conservative/spontaneous recovery</td>
</tr>
<tr>
<td>Venous infarction</td>
<td>Complex left temporal → atrial v. → basal v. → VG</td>
<td>Thrombosis of venous collector</td>
<td>Conservative/good recovery with normal development</td>
</tr>
<tr>
<td>Partial thrombosis of DVA collector</td>
<td>Right deep nuclei → ICV → internal parietal v. → SSS</td>
<td>Stenosis of venous collector</td>
<td>Conservative/no follow-up</td>
</tr>
<tr>
<td>Venous infarction</td>
<td>Right temporal → subtemporal v. → epidural sinus → transverse sinus</td>
<td>Thrombosis of venous collector</td>
<td>Conservative/good recovery</td>
</tr>
<tr>
<td>Cytotoxic and vasogenic edema</td>
<td>Frontopolar v. → SSS</td>
<td>Stenosis and thrombosis of venous collector</td>
<td>Anticoagulation/good recovery</td>
</tr>
<tr>
<td>Vasogenic and cytotoxic edema</td>
<td>Left basal ganglia → inferior striate v. → basal v.</td>
<td>Stenosis venous collector</td>
<td>Anticoagulation/good recovery</td>
</tr>
<tr>
<td>Right temporal intraparenchymal hematoma</td>
<td>Right temporal → subependymal v. → inferior temporal v. → …</td>
<td>Left frontal AVM</td>
<td>AVM embolization/good recovery</td>
</tr>
<tr>
<td>Ventricular hemorrhage/large medullary zone</td>
<td>Complex with capillary ectasia and tortuous and dilated medullary veins close to fistula/left temporal → transsural collector → Labbé v. and atrial v. → lateral atrial v. → basal v.</td>
<td>Pseudoaneurysm—medullary microshunts</td>
<td>Arterial embolization/good recovery</td>
</tr>
<tr>
<td>Left cerebellum hematoma</td>
<td>Large with capillary ectasia and tortuous and dilated medullary veins close to fistula/bilateral cerebellum → basal v. → VG</td>
<td>Microshunts</td>
<td>Arterial embolization/good recovery</td>
</tr>
<tr>
<td>Frontal intraparenchymal hematoma</td>
<td>Right paracentral lobule → venous collector → SSS</td>
<td>Microshunts</td>
<td>Arterial embolization/good recovery</td>
</tr>
<tr>
<td>Intraparenchymal cerebellar hematoma</td>
<td>Right cerebellar hemisphere → Lateromesencephalic v. → precentral cerebellar v. → VG</td>
<td>Microshunts</td>
<td>Arterial embolization/good recovery</td>
</tr>
<tr>
<td>Left frontal hematoma</td>
<td>Left frontal → pericallosal v. → frontal v. → SSS</td>
<td>Normal</td>
<td>Conservative/good recovery, normal development</td>
</tr>
<tr>
<td>Left basal hematoma</td>
<td>Large with capillary ectasia</td>
<td>Normal</td>
<td>Conservative/good recovery</td>
</tr>
<tr>
<td>Vasogenic edema</td>
<td>Left basal ganglia → inferior striate v. → basal v.</td>
<td>Normal</td>
<td>Conservative/no follow-up</td>
</tr>
</tbody>
</table>

F indicates female; M, male; SAH, subarachnoid hemorrhage; v., vein; VG, vein of Galen; ICV, internal cerebral vein; SSS, superior sagittal sinus.  

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exclude completely an associated disease (eg, patients investigated or manuscripts before the MRI era).

In addition, we reviewed all manuscripts from 1980 to 2007 in the Medline, Embase, and Scopus databases using the following search terms: “venous angioma,” “developmental venous anomaly,” “venous malformation,” “medullary malformation,” and “medullary venous malformation.” The selection and exclusion criteria described previously were the same for the series of patients selected from our databank as for the literature review.

Results
In our databank, we found 80 cases with the principal diagnosis of “DVA.” Of those, there were 17 cases with 18 vascular complications directly linked to the DVA and that could not be associated with other pathologies (Table 1). One patient (Case 2) had 2 separate complications from his DVA. Within the literature, 51 cases were found that fulfilled our criteria of well-documented truly symptomatic DVAs and in whom MRI was available to rule out any other associated disease. We restrict our review of the literature and the case series to these 68 patients with 69 distinct clinical presentations owing to vascular complications of the DVA. Values of incidence and prevalence of symptomatic DVAs could not be given, because data from our center are likely to be biased by referral. Based on the imaging features and clinical symptoms, 2 major groups of presumed pathophysiological mechanisms could be identified: mechanical and flow-related. Patients in whom complications were present and in whom no pathomechanism could be identified were grouped separately (idiopathic pathomechanism; Figure 1).

Mechanical
Mechanical complications were considered when some component of the DVA (typically the draining collector vein) compressed an intracranial structure (parenchyma, cranial nerves, ventricles, or bone), thereby producing compressive symptoms that could be documented by imaging. We found 2 patients from our series and 12 additional cases from the literature (Table 2).

The mean age of patients in this group was 30 years with a range from 1 to 62 years; there was no gender predominance (male:female = 7:7). Most cases were related to the collecting vein of a posterior fossa DVA (n = 9 [64.3%]); in 42.8% of cases, the venous collector of the DVA was dilated. There was no relation between compressive symptoms and the size of the drained medullary zone. A detailed description of the clinical symptoms can be seen in Tables 1 and 2. Obstructive hydrocephalus (n = 7 [50%]) and neurovascular nerve compression syndromes (n = 6 [42.8%]), being trigeminal neuralgia, facial hemispasm, or tinnitus, were the most common findings. The structure most typically compressed was the mesencephalic aqueduct (n = 6 [42.8%]) followed by the trigeminal nerve (n = 3 [21.4%]) and the acousticofacial complex (n = 3 [21.4%]). The orbital contents (Case 1) and the interventricular foramen were compressed in one patient each.

The patients presenting with hydrocephalus had the occlusion at the level of the aqueduct (n = 6) or, in a single case, at the level of the interventricular foramen producing unilateral ventricular dilatation. Shunting surgery was performed in 3 patients, endoscopic third ventriculostomy in 2 (Case 2), whereas 3 patients (28%) were kept under close clinical observation without published surgical treatment. Three patients with nerve compression underwent decompressive treatment with excellent results (Figure 2). For the remaining patients, the treatment was conservative.

Flow-Related
Flow-related complications were characterized as a misbalance of the in- and outflow of blood in the DVA system raising the pressure in the DVA either due to an increased inflow into the DVA or to an obstruction of the outflow.
<table>
<thead>
<tr>
<th>Reference</th>
<th>Morphology</th>
<th>Age, Years</th>
<th>Sex</th>
<th>Clinical Presentation</th>
<th>Localization</th>
<th>Drainage of Venous Collector</th>
<th>Treatment/ Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Truwit, 1992</td>
<td>Unilateral ventricular dilatation</td>
<td>37</td>
<td>F</td>
<td>Headaches</td>
<td>Left basal ganglia and thalamus</td>
<td>Dilated venous collector with compression of interventricular foramen</td>
<td>Left striae v. → ICV</td>
</tr>
<tr>
<td>Oka, 1993</td>
<td>Hydrocephalus</td>
<td>43</td>
<td>F</td>
<td>Headaches and seizures</td>
<td>Tectum</td>
<td>Dilated transmesencephalic venous collector with aqueductal obstruction</td>
<td>Precentral cerebellar v. → VG</td>
</tr>
<tr>
<td>Nagata, 1995</td>
<td>Vessel–nerve contact</td>
<td>35</td>
<td>M</td>
<td>Left trigeminal neuralgia</td>
<td>Left cerebellar hemisphere</td>
<td>Large and complex, dilated venous collector with compression of CN V</td>
<td>Left petrosal v.</td>
</tr>
<tr>
<td>Blackmore, 1996</td>
<td>Hydrocephalus</td>
<td>16</td>
<td>F</td>
<td>Intermittent throbbing occipital headache, associated with photophobia and motion sickness</td>
<td>Left thalamus</td>
<td>Dilated subependymal venous collector with aqueductal compression</td>
<td>VG (direct)</td>
</tr>
<tr>
<td>Chen, 1996</td>
<td>Vessel–nerve contact</td>
<td>53</td>
<td>F</td>
<td>Left facial hemispasm</td>
<td>Left cerebellar hemisphere</td>
<td>Large dilated venous collector with compression of CN VII</td>
<td>Precentral cerebellar v. → petrosal v.</td>
</tr>
<tr>
<td>Kuker, 1997</td>
<td>Vessel–nerve contact</td>
<td>62</td>
<td>M</td>
<td>Left trigeminal neuralgia and slight dysesthesia V2 and V3</td>
<td>Lobus semilunaris superior and inferior of the left cerebellar hemisphere</td>
<td>Transpontine v. without venous ectasia, compression of CN V</td>
<td>v. of the lateral recess of the fourth ventricle → subependymal v. → VG</td>
</tr>
<tr>
<td>Korinth, 2002</td>
<td>Vessel–nerve contact</td>
<td>37</td>
<td>F</td>
<td>Trigeminal neuralgia</td>
<td>Left cerebellar hemisphere</td>
<td>Large DVA compressing CN V, no venous ectasia</td>
<td>Lateral mesencephalic v.</td>
</tr>
<tr>
<td>Bannur, 2002</td>
<td>Hydrocephalus</td>
<td>11</td>
<td>M</td>
<td>Persistent headache + acute ataxia, vomiting, vertigo, papilledema</td>
<td>Midbrain close to aqueduct</td>
<td>Aqueductal stenosis by dilated venous collector</td>
<td>subependymal v. → VG</td>
</tr>
<tr>
<td>Yagmurlu, 2002</td>
<td>Hydrocephalus</td>
<td>7</td>
<td>F</td>
<td>Severe, progressive headaches</td>
<td>Multiple DVAs (thalamic, bilateral cerebellar)</td>
<td>Signs of compression of the aqueduct by dilated venous collectors</td>
<td>v. of the lateral recess of the fourth ventricle → subependymal v. → VG</td>
</tr>
<tr>
<td>Malinvaud, 2006</td>
<td>Vessel–nerve contact</td>
<td>55</td>
<td>M</td>
<td>Permanent, nonpulsatile tinnitus in right ear</td>
<td>Right cerebellar hemisphere</td>
<td>Dilated venous collector, compressing CN VIII</td>
<td>Precentral cerebellar v. → petrosal v.</td>
</tr>
<tr>
<td>Shim, 2007</td>
<td>Vessel–nerve contact</td>
<td>5</td>
<td>M</td>
<td>Progressive hearing loss</td>
<td>Right cerebellar hemisphere</td>
<td>Dilated venous collector, compressing CN VIII; associated scalp hemangiomas</td>
<td>Petrosal v. (direct)</td>
</tr>
</tbody>
</table>

M indicates male; F, female; v., vein; CN, cranial nerve; ICV, internal cerebral vein; VG, vein of Galen.
**Increase of Developmental Venous Anomalies’ Inflow**

An augmentation of inflow into the DVA was either due to microshunts into the DVA or AVMs that used the DVA as the drainage route. We found 4 cases in our databank and 15 cases in the literature with a mean age of 28.5 years ranging from 1 to 62 years and a discrete male predominance (male:female = 12:7; Table 3). The initial clinical presentations included headaches (n=11 [61%]), neurological deficits (n=7 [38%]), seizures (n=4 [22.2%]), and coma (n=4 [22.2%]). The morphological presentation was mainly hemorrhages in 12 cases (66.6%), including intraparenchymal (n=8 [66%]), intraventricular (n=2 [17%]), and both (n=2 [17%]). The remaining 6 cases (33.3%) had venous infarction in the drainage territory of the DVA, presumably due to venous congestion after arterialization. Thirteen lesions (72%) were located supratentorially and 6 lesions (28%) infratentorially. The angioarchitectural aspects were microshunts into capillary veins at the medullary zone of the DVA (n=11 [55%]; Figures 3 and 4) and typical nidal-type AVMs draining through the venous collector (n=8 [45%]). Among them, only 3 cases were larger than 5 cm and had complex venous drainage with no relationship with the clinical manifestation. Asymmetrical dilatation of the capillary veins in the medullary zone of the DVA of the patients with microshunts was observed in 76% (8 of 11). This finding helped to support the diagnosis in some cases, which was subsequently confirmed by superselective injections.

Treatment strategies were extremely variable according to architecture, morphological presentation, and the treating center. Radiosurgery was the most frequent option (n=7 [38.8%]), even for hemorrhagic or ischemic presentations, and was focused on the AVM and DVA (70%) or on the AVM alone (30%). Five patients (including all 4 cases from our series) were treated with endovascular embolization of the lesion (microshunt or AVM) with careful preservation of the patency of the DVA using transarterial glue (n-butyl cyanoacrylate, Histoacryl; B. Braun, Melsungen, Germany) injections. All patients recovered from their bleeding without new neurological deficits.

Among the 13 patients who presented with hemorrhage, 3 patients had their hematoma surgically drained preserving the DVA itself. Two other patients with an associated AVM were operated with the goal of AVM resection. In one of them, the DVA was occluded unintentionally and the patient had severe venous ischemia resulting in new and permanent neurological deficits.

**Developmental Venous Anomaly Outflow Restriction**

An imbalance of blood flow can also occur if the venous outlets of the DVA are restricted while the inflow is normal. This category can be further subdivided into anatomic and functional causes, the latter being due to a remote arterial overload to the venous system due to a distant shunt/AVM, whereas the former can be secondary to thrombosis of the DVA channels, stenosis or occlusion of the venous collector, or the distal draining sinus.

**Anatomic Obstacle**

Concerning mechanical obstruction of DVA outflow, we report 8 cases from our databank and 18 previously published cases (Table 4). The mean age was 32.1 years with no gender predominance (male:female = 14:12). There were 7 patients (27.6%) who presented with hemorrhage (either intraparenchymal or subarachnoid), whereas the major presentation was venous congestion with edema (Figure 5). Clinical symptoms consisted of neurological deficits (n=20 [68.9%]), headaches (n=17 [58.6%]), seizures (n=12 [41.3%]), and alteration of consciousness or altered mental status (n=6 [20.7%]). Twenty-three (79.3%) were located supratentorially. No difference in size (only 55% were larger than 3 cm) nor venous drainage (55% draining to the deep venous system and 45% to the superficial veins) was found within this group of patients. Fifteen (51.7%) had thrombosis on the venous collector, 24.1% (n=7) had stenosis at some point of the DVA itself. Two other patients with an associated AVM were operated with the goal of AVM resection. In one of them, the DVA was occluded unintentionally and the patient had severe venous ischemia resulting in new and permanent neurological deficits.

**Figure 2.** DVA causing mechanical compression. The patient had severe trigeminal pain in the distribution of left V2 and V3. A (contrast-enhanced T1-weighted axial section), the enlarged venous collector of a transspontine DVA encroaches on the trigeminal nerve at its exiting zone from the brain stem (arrow). B–C, Vertebral artery angiograms in anteroposterior and lateral views in the venous phase demonstrate a classical umbrella-shaped pattern of the DVA with the medullary vein draining into an enlarged collector that further drained into the superior petrosal vein. D–E, Surgical view with compression of the left trigeminal nerve (arrow in E). A Teflon patch to separate the nerve from the vein was placed with excellent results and complete recovery from the trigeminal pain immediately after surgery.
<table>
<thead>
<tr>
<th>Reference</th>
<th>Mechanism</th>
<th>Morphology</th>
<th>Age, Years</th>
<th>Sex</th>
<th>Clinical Presentation</th>
<th>Localization</th>
<th>Angiography</th>
<th>Venous Drainage</th>
<th>Risk Factor/Associated Conditions</th>
<th>Treatment/Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bergui, 1997</td>
<td>Flow misbalance: increased inflow (microshunt)</td>
<td>Parenchymal hemorrhage</td>
<td>35</td>
<td>F</td>
<td>Diplopia, hemiparesis</td>
<td>Brainstem</td>
<td>Capillary ectasia; tortuous and dilated medullary veins close to shunt</td>
<td>v. of the lateral recess of the fourth ventricle → precentral cerebellar v. → VG</td>
<td>Medullary microshunts</td>
<td>Conservative/good recovery</td>
</tr>
<tr>
<td>Bergui, 1997</td>
<td>Flow misbalance: increased inflow (microshunt)</td>
<td>Parenchymal hemorrhage</td>
<td>39</td>
<td>M</td>
<td>Headache, aphasia, right hemiparesis</td>
<td>Left temporal</td>
<td>Classical aspect without capillary ectasia</td>
<td>Superior striate v. → basal v.</td>
<td>Medullary microshunts</td>
<td>Radiosurgery/partial recovery</td>
</tr>
<tr>
<td>Komiyama, 1999</td>
<td>Flow misbalance: increased inflow (microshunt)</td>
<td>Intraventricular hemorrhage</td>
<td>22</td>
<td>M</td>
<td>Headache, dizziness</td>
<td>Left parietal</td>
<td>Venous ectasia; asymmetric opacification, tortuous and dilated medullary veins</td>
<td>SSS (direct)</td>
<td>Medullary microshunts</td>
<td>Conservative/good recovery</td>
</tr>
<tr>
<td>Trueit, 1982</td>
<td>Flow misbalance: increased inflow (AVM)</td>
<td>Intraparenchymal hemorrhage</td>
<td>1</td>
<td>M</td>
<td>Seizures</td>
<td>Left frontal</td>
<td>Classical pattern without capillary ectasia</td>
<td>SSS (direct)</td>
<td>AVM draining directly into DVA</td>
<td>Surgery with resection of the AVM/good recovery</td>
</tr>
<tr>
<td>Awad, 1993</td>
<td>Flow misbalance: increased inflow (microshunt)</td>
<td>Intraparenchymal hemorrhage</td>
<td>54</td>
<td>M</td>
<td>Seizures, headache</td>
<td>Right temporal</td>
<td>Not reported</td>
<td>Not reported</td>
<td>Medullary microshunts</td>
<td>Surgery with drainage of hematoma/good recovery</td>
</tr>
<tr>
<td>Awad, 1993</td>
<td>Flow misbalance: increased inflow (microshunt)</td>
<td>Intraparenchymal hemorrhage</td>
<td>39</td>
<td>F</td>
<td>Seizures</td>
<td>Right frontal</td>
<td>Not reported</td>
<td>Not reported</td>
<td>Medullary microshunts</td>
<td>Surgery (drainage of hematoma)/good recovery</td>
</tr>
<tr>
<td>Awad, 1993</td>
<td>Flow misbalance: increased inflow (microshunt)</td>
<td>Venous congestion</td>
<td>36</td>
<td>F</td>
<td>Coma</td>
<td>Left parietal</td>
<td>Not reported</td>
<td>Not reported</td>
<td>Medullary microshunts</td>
<td>Radiosurgery/good recovery</td>
</tr>
<tr>
<td>Lindquist, 1993</td>
<td>Flow misbalance: increased inflow (AVM)</td>
<td>Intraparenchymal hemorrhage</td>
<td>60</td>
<td>F</td>
<td>Not reported</td>
<td>Right temporal</td>
<td>Not reported</td>
<td>Not reported</td>
<td>AVM draining directly into DVA</td>
<td>Radiosurgery (DVA/AVM)</td>
</tr>
<tr>
<td>Lindquist, 1993</td>
<td>Flow misbalance: increased inflow (AVM)</td>
<td>Intraparenchymal hemorrhage</td>
<td>20</td>
<td>M</td>
<td>Not reported</td>
<td>Bilateral cerebellum</td>
<td>Not reported</td>
<td>Not reported</td>
<td>AVM draining directly into DVA</td>
<td>Radiosurgery (DVA/AVM)</td>
</tr>
<tr>
<td>Meyer, 1995</td>
<td>Flow misbalance: increased inflow (AVM)</td>
<td>Intraparenchymal hemorrhage</td>
<td>32</td>
<td>F</td>
<td>Headache, aphasia, right hemiparesis and progressive to coma</td>
<td>Right parietal</td>
<td>Not reported</td>
<td>Not reported</td>
<td>SSS (direct)</td>
<td>AVM draining directly into DVA</td>
</tr>
<tr>
<td>Nussbaum, 1996</td>
<td>Flow misbalance: increased inflow (microshunt)</td>
<td>Venous congestion</td>
<td>24</td>
<td>M</td>
<td>2 episodes of acute headache, vertigo, longstanding blurred vision and nystagmus</td>
<td>Right cerebellum</td>
<td>Large multicycle microshunts; asymmetric medullary veins, tortuous and dilated close to microshunts</td>
<td>v. of the lateral recess of the fourth ventricle → precentral cerebellar v. → VG</td>
<td>Medullary microshunts</td>
<td>Conservative/good recovery</td>
</tr>
<tr>
<td>Kurita, 1999</td>
<td>Flow misbalance: increased inflow (AVM)</td>
<td>Venous infarction</td>
<td>39</td>
<td>M</td>
<td>Transient dysarthria</td>
<td>Right temporal</td>
<td>Not reported</td>
<td>Superficial sylvian v. → sphenopalatine sinus</td>
<td>AVM draining directly into DVA</td>
<td>Radiosurgery/good recovery</td>
</tr>
<tr>
<td>Yanaka, 2001</td>
<td>Flow misbalance: increased inflow (AVM)</td>
<td>Venous congestion</td>
<td>37</td>
<td>F</td>
<td>Loss of consciousness</td>
<td>Right basal ganglia and lateral wall of right ventricle</td>
<td>Significant dilatation of medullary veins</td>
<td>Superior striate v. → longitudinal caudate v. → lateral subependymal v. → VG</td>
<td>AVM draining directly into DVA</td>
<td>Radiosurgery/good recovery</td>
</tr>
<tr>
<td>Aksoy, 2001</td>
<td>Flow misbalance: increased inflow (AVM)</td>
<td>Venous congestion</td>
<td>11</td>
<td>M</td>
<td>Temporal lobe seizures</td>
<td>Right temporal lobe</td>
<td>Capillary ectasia</td>
<td>Anterior temporal v. → basal v.</td>
<td>AVM draining into 2 DVAs</td>
<td>Radiosurgery for AVM/good recovery</td>
</tr>
<tr>
<td>Kuc, 2007</td>
<td>Flow misbalance: increased inflow (microshunt)</td>
<td>Intraparenchymal hemorrhage</td>
<td>56</td>
<td>M</td>
<td>Aphasia, hemiparesis</td>
<td>Left temporal</td>
<td>Large; asymmetric medullary veins, tortuous and dilated close to the hematoma</td>
<td>Not reported</td>
<td>Medullary microshunts</td>
<td>Drainage of hematoma</td>
</tr>
</tbody>
</table>

F indicates female; M, male; v., vein; VG, vein of Galen; SSS, superior sagittal sinus.
corresponded to the DVA drainage territory in 21 cases (72.4%). The management was variable due to delay of the diagnosis in most of the patients. There were 16 patients (55.2%) who received conservative treatment without anticoagulation and antiaggregation. Systemic heparinization was administered in 9 patients (31%), similar to the treatment in cortical venous thrombosis. In 3 cases, decompressive craniectomy for refractory and malignant regional edema was deemed necessary and in one patient, a ventricular shunt for hydrocephalus treatment due to posterior fossa hypertension after cerebellar infarction was placed. The overall outcome was good in 24 patients (82.8%).

Functional Outflow Restriction:
Functional impairment of the venous drainage of the DVA was suspected in patients with a remote arteriovenous shunt not draining directly into the DVA but competing and hindering the normal DVA drainage due to venous hypertension (Figure 6). Although 3 such cases are present in the literature (2 dural arteriovenous shunts and one pial AVM), we report one additional patient with a pial AVM distant to the DVA but likely to produce venous hypertension. The mean age was 37 years old and all 4 patients were male. All patients became symptomatic due to venous congestion of the area drained by the DVA either with hemorrhagic venous infarction or congestive edema. Management included treatment of the primary shunt to decrease the venous hypertension in 2 cases, whereas in one patient, conservative management with anticoagulation therapy was performed. The follow-up was uneventful and the patients had a good recovery.

Idiopathic
Idiopathic complications were attributed to symptomatic cases with no obvious vascular modification attributable to the DVA,
Table 4. Findings in All Patients Reported in the Literature With Decreased Outflow From the DVA

<table>
<thead>
<tr>
<th>Reference</th>
<th>Mechanism</th>
<th>Morphology</th>
<th>Age, Years</th>
<th>Sex</th>
<th>Clinical Presentation</th>
<th>Localization</th>
<th>Angiography/Imaging</th>
<th>Drainage of the Venous Collector</th>
<th>Risk Factors/Associated Conditions</th>
<th>Treatment/Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bouchacourt, 1986&lt;sup&gt;15&lt;/sup&gt;</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Venous infarction</td>
<td>37</td>
<td>F</td>
<td>Headache, seizures, right hemiparesis</td>
<td>Left frontal</td>
<td>Thrombosis of venous collector of DVA</td>
<td>Thalamostriate v. → ICV</td>
<td>...</td>
<td>Heparin/good recovery</td>
</tr>
<tr>
<td>Yamamoto, 1989&lt;sup&gt;16&lt;/sup&gt;</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Venous infarction</td>
<td>26</td>
<td>F</td>
<td>Headache, left hemiparesis</td>
<td>Right parietal</td>
<td>Large DVA, thrombosis of superior sagittal sinus (SSS)</td>
<td>SSS (direct)</td>
<td>Postpartum</td>
<td>Decompressive craniotomy/recovery with mild residual symptoms</td>
</tr>
<tr>
<td>Truwit, 1992&lt;sup&gt;17&lt;/sup&gt;</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Venous infarction</td>
<td>12</td>
<td>M</td>
<td>Seizures</td>
<td>Left frontal</td>
<td>Large, complex DVA with stenosis of venous collector</td>
<td>SSS (direct)</td>
<td>...</td>
<td>Conservative/no follow-up</td>
</tr>
<tr>
<td>Field and Russell, 1995&lt;sup&gt;18&lt;/sup&gt;</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Congestive hemorrhage</td>
<td>34</td>
<td>F</td>
<td>Persistent headache, left hemianopia</td>
<td>Right parieto-temporal</td>
<td>Thrombosis of venous collector of DVA</td>
<td>Tentorial venous plexus → sup. petrosal sinus</td>
<td>...</td>
<td>Conservative/good recovery</td>
</tr>
<tr>
<td>Kim, 1996&lt;sup&gt;19&lt;/sup&gt;</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Venous infarction</td>
<td>13</td>
<td>M</td>
<td>Ataxic gait, emesis, left hemiparesis, right mydriasis</td>
<td>Right temporo-parietal</td>
<td>Large, complex DVA with thrombosis of venous collector</td>
<td>Labbé → transverse sinus</td>
<td>...</td>
<td>Decompressive craniotomy/death</td>
</tr>
<tr>
<td>Guerrero, 1996&lt;sup&gt;20&lt;/sup&gt;</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Venous infarction</td>
<td>62</td>
<td>M</td>
<td>Vertigo, diplopia, ataxia and occipital headache and hydrocephalus</td>
<td>Mesencephalon + right cerebellar hemisphere</td>
<td>Large DVA with thrombosis of venous collector</td>
<td>Precentral cerebellar v. → VG</td>
<td>...</td>
<td>Conservative/slow recovery</td>
</tr>
<tr>
<td>Merten et al, 1996&lt;sup&gt;21&lt;/sup&gt;</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Congestive hemorrhage</td>
<td>50</td>
<td>F</td>
<td>Headache, aphasis, right hemiparesis</td>
<td>Left basal ganglia</td>
<td>Large collector significant dilatation of medullary veins; thrombosis of venous collector</td>
<td>Subependymal v. → Sylvian v.</td>
<td>...</td>
<td>Heparin followed by warfarin/good recovery</td>
</tr>
<tr>
<td>Konan, 1999&lt;sup&gt;22&lt;/sup&gt;</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Venous infarction</td>
<td>31</td>
<td>M</td>
<td>Severe headache, vomiting, ataxia, and right-sided facial paresis and coma</td>
<td>Bilateral cerebellar</td>
<td>Large DVA, presence of a clot inside the DVA collector</td>
<td>Precentral cerebellar v. → VG</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>Herbreteau, 1999&lt;sup&gt;23&lt;/sup&gt;</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Venous infarction</td>
<td>45</td>
<td>M</td>
<td>Seizures</td>
<td>Left parietal</td>
<td>Venous collector stenosis</td>
<td>Thalamostriate v. → ICV</td>
<td>...</td>
<td>Conservative/good recovery</td>
</tr>
<tr>
<td>Lai et al, 1999&lt;sup&gt;24&lt;/sup&gt;</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Venous infarction</td>
<td>56</td>
<td>F</td>
<td>Seizures, left hemiparesis</td>
<td>Right parietal</td>
<td>Thrombosis of venous collector</td>
<td>SSS (direct)</td>
<td>...</td>
<td>Conservative/good recovery</td>
</tr>
<tr>
<td>Thobois et al, 1999&lt;sup&gt;25&lt;/sup&gt;</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Venous infarction</td>
<td>25</td>
<td>F</td>
<td>Headache, seizures, and left hemianopia homonym</td>
<td>Right parieto-occipital</td>
<td>Thrombosis of venous collector</td>
<td>SSS (direct)</td>
<td>Contraceptives</td>
<td>Anticoagulation/good recovery</td>
</tr>
<tr>
<td>Masson, 2000&lt;sup&gt;26&lt;/sup&gt;</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Venous infarction</td>
<td>68</td>
<td>M</td>
<td>Headache, seizures, right hemiplegia, and aphasis</td>
<td>Left frontoparietal</td>
<td>Thrombosis of SSS</td>
<td>SSS (direct)</td>
<td>...</td>
<td>Heparin/mild motor sequelae</td>
</tr>
<tr>
<td>Hammoud, 2002&lt;sup&gt;27&lt;/sup&gt;</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Venous infarction</td>
<td>26</td>
<td>F</td>
<td>Right-sided acute numbness and weakness</td>
<td>Left frontoparietal</td>
<td>Thrombosis of venous collector</td>
<td>...</td>
<td>Postpartum/family history of thrombosis/smoke/contraceptives</td>
<td>Conservative/good recovery</td>
</tr>
<tr>
<td>Lovrencic-Huzjan, 2004&lt;sup&gt;28&lt;/sup&gt;</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Subarachnoid hemorrhage</td>
<td>39</td>
<td>M</td>
<td>Ocipital headache, nausea and vomiting</td>
<td>Cerebellum/SAT</td>
<td>Thrombosis of venous collector</td>
<td>Precentral cerebellar v. → VG</td>
<td>...</td>
<td>Conservative/good recovery</td>
</tr>
<tr>
<td>Peltier, 2004&lt;sup&gt;29&lt;/sup&gt;</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Venous infarction, subsequent obstructive hydrocephalus</td>
<td>32</td>
<td>M</td>
<td>Headache, vomiting, and coma</td>
<td>Pons/left cerebellar hemisphere</td>
<td>Complex, large DVA, thrombosis of venous collector</td>
<td>v. of the lateral recess of the fourth ventricle → petrosal v.</td>
<td>...</td>
<td>External drainage/recovery with mild deficits</td>
</tr>
</tbody>
</table>

(Continued)
no associated vascular condition nor systemic factor. We report 3 cases of our series and 3 previously reported cases in this category presenting with hemorrhage in 4 and venous infarction in 2. The global mean age was 33.5 years old (range, 0 to 56 years); all cases had neurological deficits. Three patients had unusually large and complex DVAs with deep venous drainage in 2 patients. Because no risk factor could be found, there was no treatment considered in these cases (Table 5).

**Discussion**

The presumed origin of DVAs is considered to be venous thrombosis during Padget’s fourth to seventh stage that

Table 4. Continued

<table>
<thead>
<tr>
<th>Reference</th>
<th>Mechanism</th>
<th>Morphology</th>
<th>Age, Years</th>
<th>Sex</th>
<th>Clinical Presentation</th>
<th>Localization</th>
<th>Angiography/Imaging</th>
<th>Drainage of the Venous Collector</th>
<th>Risk Factors/Associated Conditions</th>
<th>Treatment/Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Flacke, 200677</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Ischemic—venous congestion</td>
<td>49</td>
<td>M</td>
<td>Seizures</td>
<td>Left frontal</td>
<td>Thrombosis of venous collector</td>
<td>SSS (direct)</td>
<td>Elevated factor VIII activity of 227%</td>
<td>Anticoagulation/good recovery</td>
</tr>
<tr>
<td>Vieira Santos, 200655</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Venous infarction</td>
<td>9</td>
<td>F</td>
<td>Right hemiparesis</td>
<td>Left parietal</td>
<td>Thrombosis of venous collector</td>
<td>SSS (direct)</td>
<td>Homozygous for 4G allele of PAI-1</td>
<td>Heparin/slow recovery</td>
</tr>
<tr>
<td>Seki, 200778</td>
<td>Flow misbalance: outflow obstruction</td>
<td>Congestive hemmorhage</td>
<td>33</td>
<td>M</td>
<td>Headache, right hemiparesis, and coma</td>
<td>Left temporoparietal</td>
<td>Ectasia and thrombosis of venous collector</td>
<td>VG (direct) → straight sinus</td>
<td>Straight sinus stenosis</td>
<td>Cranietomy, hematoma drainage/recovery with mild deficits</td>
</tr>
<tr>
<td>Kuncz, 200157</td>
<td>Flow misbalance: outflow restriction due to remote shunt</td>
<td>Venous infarction</td>
<td>31</td>
<td>M</td>
<td>Seizures and right hemiparesis</td>
<td>Left frontal</td>
<td>Concurrence of venous drainage with DAVFs and slow flow in DVA</td>
<td>SSS (direct)</td>
<td>Distant bilateral DAVFs from ethmoidal arteries, draining into SSS</td>
<td>Surgery for DAVFs/left frontal atrophy and cognitive deficits</td>
</tr>
<tr>
<td>Agazzi, 200153</td>
<td>Flow misbalance: outflow restriction due to remote shunt</td>
<td>Venous infarction</td>
<td>39</td>
<td>M</td>
<td>Seizures, mild left hemiparesis, homonymous left hemianopia</td>
<td>(1) Right basal ganglia; (2) right temporal</td>
<td>Two DVAs, one of which had microhunts and early drainage; tortuous and dilated medullary veins close to fistula</td>
<td>(1) Subependymal v. → tentorial sinus; (2) inferior temporal v. → Labbé</td>
<td>Restricted venous drainage due to anatomic variations</td>
<td>Aspirin/good recovery</td>
</tr>
<tr>
<td>Dudeck, 200479</td>
<td>Flow misbalance: outflow restriction due to remote shunt</td>
<td>Venous congestion</td>
<td>16</td>
<td>M</td>
<td>Pulsatile tinnitus 14 months after posterior fossa surgery for brain stem cavernoma resection</td>
<td>Right cerebellar hemisphere</td>
<td>Complex DVA, concurrence of venous DVA drainage with DAVF and slow flow in DVA</td>
<td>v. of the lateral recess of the fourth ventricle → precentral cerebellar v. → VG</td>
<td>Right posterior fossa DAVF (Cognard type II/a + b), fed by ipsilateral cerebellar arteries</td>
<td>Conservative/stable</td>
</tr>
</tbody>
</table>

F indicates female; M, male; SAH, subarachnoid hemorrhage; DAVF, dural arteriovenous fistula; v., vein; ICV, internal cerebral vein; VG, vein of Galen.

Figure 5. Flow-related complication due to an anatomic outflow restriction: This 58-year-old woman had an acute onset of right hemiparesis and headaches. A (axial nonenhanced coronal CT), a hypodensity of the left basal ganglia region that was not related to a typical vascular territory. B–C, CT angiography in axial (B) and coronal views (C) demonstrate a DVA with dilated medullary veins draining into the deep venous system. D–E, the 3-dimensional reconstruction of the venous phase of a left internal carotid artery injection demonstrates a basal ganglia DVA draining into the basal vein of Rosenthal. There is a stenosis of the venous collector (arrow in D) and a dilatation of the vein proximal to the stenosis (small arrows in E). Presumably, the stenosis had led to a decreased outflow of the DVA and venous congestive edema in the medullary zone normally drained by the DVA.
Table 5. Findings in All Patients Reported in the Literature With Idiopathic Vascular Complications and DVAs

<table>
<thead>
<tr>
<th>Reference</th>
<th>Mechanism</th>
<th>Morphology</th>
<th>Age, Years</th>
<th>Sex</th>
<th>Clinical Presentation</th>
<th>Localization</th>
<th>Angiography</th>
<th>Venous Drainage</th>
<th>Risk Factors/Associated Conditions</th>
<th>Treatment/Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Uchino, 1996</td>
<td>Idiopathic</td>
<td>Hemorrhage</td>
<td>49</td>
<td>M</td>
<td>Aphasia</td>
<td>Left frontoparietal</td>
<td>Large typical pattern</td>
<td>SSS (direct)</td>
<td>None</td>
<td>Conservative/good recovery</td>
</tr>
<tr>
<td>Masson, 2000</td>
<td>Idiopathic</td>
<td>Venous infarction</td>
<td>43</td>
<td>M</td>
<td>Seizures and right hemiplegia</td>
<td>Left parietal</td>
<td>Classical pattern</td>
<td>SSS (direct)</td>
<td>None</td>
<td>Heparin/good recovery</td>
</tr>
</tbody>
</table>

M indicates male; SSS, superior sagittal sinus.
according to their presumed pathomechanism and were able to
divide them into 2 subsets: those caused by mechanical
compression of intracranial structures (being due to an
atypical location of the DVA) and those caused by a misbal-
ance of either the in- and outflow in the DVA (therefore being
related to their relative inflexibility of changes in the venous
equilibrium). More than 92.7% of truly symptomatic DVAs
harbored either one of these mechanisms. However, an
important caveat to keep in mind is that it cannot be
completely ruled out that the concurrence of a DVA with
venous thrombosis within its collecting vein could be a mere
coincidence because both entities represent frequent
diseases/variants.

Mechanical-Related Symptomatology
The venous collector of a DVA can compress intracranial
structures, especially if dilated or ectatic and in close prox-
imity to vulnerable structures. The neurological symptoms
were caused by mechanical compression in 32.7% of all cases
of symptomatic DVAs with hydrocephalus, tinnitus, brain-
stem deficits, facial hemispasm, and trigeminal neuralgia
being the most common presenting symptoms. Obstruction of
the ventricles has not only been described for DVAs, but also for
dilated drainage veins of an AVM. Potential sites for obstruction are at the level of the
interventricular foramen (here related to dilated thalamo-
striate veins) or at the aqueduct (due to a dilated vein of
Galen or transparenchymal venous collectors). These cases
should be differentiated from hydrocephalus secondary to
a hydrovenous imbalance that can be seen in young
patients with high-flow fistulae and vein of Galen
malformations.

In hydrocephalus related to the venous collector of a DVA,
that cannot be managed conservatively, the management
should be exclusively the treatment of the hydrocephalus
either using a shunt or a ventriculocisternostomy. Neurovas-
cular compression syndromes on the other hand can be
successfully treated by microvascular decompression. For
other intracranial compression syndromes related to a DVA,
management should be conservative with preservation of the
integrity and patency of the venous collector to avoid venous
ischemic complications.

Flow-Related Symptomatology
In DVAs, as extreme variations of normal venous drainage, a
single collector drains an abnormally large parenchymal
territory. This can lead to a more fragile venous outflow
system because the single venous collector can be overloaded
accounting for the dilated medullary veins. In the group of
“flow-related” complications are those DVAs subsumed in
which this fragile equilibrium of in- and outflow is disturbed
and which thereby become symptomatic.

Increase of the Inflow
Considering the increased inflow into a DVA, we found 18
cases of AVMs draining directly through a DVA. In
comparison to AVMs draining through regular veins, these
patients presented with a high rate of parenchymal hem-
orrhage. In our cases, and in those documented by angiog-
raphy in the literature, the morphology of the medullary
veins draining into the DVA were characteristically dilated
and ectatic. This chronic increased pressure within the
DVA may change its natural history by increasing the risk
of venous rupture because of an already fragile venous
outlet.

Associations of 2 or more different cerebral vascular
malformations are not uncommon, the most well known
being the association of cavernomas and DVAs. Although
the latter are most likely due to a common pathomecha-
nism, the exceedingly rare combination of a DVA with an
AVM is presumably purely by coincidence. There have
been reports describing a hybrid malformation consisting
of an AVM and a DVA as a rare subset of mixed
cerebrovascular malformations. In certain large and com-
plex DVAs, a slightly early venous filling can be present,
which has led to the description of so-called “mixed
vascular malformations” with what has been described as
“microshunts.” In our experience, an increased medul-
lar blush is not related to a true shunt, but rather
demonstrates a rapid transit time because of enlarged
medullary veins and we have found no symptomatic cases
in our databank nor in the literature. Therefore, it is our
opinion that the association of a true AVM with a DVA
exists as a distinct and rare entity that is associated with a
higher risk of hemorrhage and complications. Following
this line of thought, there is in our practice a place for
preventive treatment in an asymptomatic patient with a
shunt draining through a DVA.

The management of these lesions is aimed at treatment of
the AVM with surgery, radiosurgery, or embolization
with preservation of the patency of the DVA because it has
been described that the proper treatment of the AVM
decreases the risk of complication of the DVA.

Restriction of Outflow
Restriction of the venous drainage from a DVA can occur by
2 pathomechanisms: by an anatomic obstacle to the normal
drainage (secondary to stenosis or thrombosis of the DVA
or its drainage vein) or by a “functional” obstacle that can
be caused by an increase in the venous pressure secondary
to a distant arteriovenous shunt (dural arteriovenous shunt
or AVM).

The restriction of outflow can produce a variety of
morphological and clinical presentations ranging from
venous congestive edema to hemorrhage similar to sinus
cortical venous thrombosis. Clinical symptomatology
is therefore highly variable and dependent on the cause,
localization, extension, and time of development of the
venous occlusion. Signs of increased intracranial pressure
may be present; neurological deficits and seizures can occur
in the group of patients in whom focal congestive and
hemorrhagic lesions occur. A congestive (ie, vasogenic)
edema with a breakdown of the blood–brain barrier is a
potential presentation that can proceed to hemorrhagic or
true ischemic transformation, the latter being most likely
due to a critical diminution in cerebral blood flow with
subsequent cytotoxic edema. Early
recanalization of the venous collector will presumably prevent this complication. Consequently, anticoagulation was suggested as the first-line treatment in symptomatic DVAs even in the presence of hemorrhage, similar to the treatment of sinus or venous thrombosis.

Functional obstruction of the venous drainage is present in associated venous hypertension after an arteriovenous shunt. The association between a symptomatic DVA and a dural arteriovenous shunt has been reported previously. Dural arteriovenous shunts invariably increase the pressure in the dural venous sinuses. This causes mild to severe disturbances in the draining functions of other veins. A more fragile venous system with decreased flexibility that may be present in DVAs will be more prone to becoming symptomatic leading to venous congestive edema or ischemia. As already described, the aim should be the treatment of the arteriovenous shunt with preservation of the DVA.

**Idiopathic Symptomatology**

Given the previously mentioned considerations, the pathomechanisms of 92.7% of all symptomatic DVAs could be explained, however, in a fraction of the cases described; no clear pathomechanism could be identified. They presented mainly with intraparenchymal hemorrhage. Whether this was due to an unrecognized small cavernoma or a resolved thrombosis or truly due to a rupture of the DVA can therefore not be decided. It is of interest, however, that in most symptomatic idiopathic cases, large and complex DVAs were present.

**Evaluation of Symptomatic Cases**

In cases of hemorrhage related to DVAs, cavernomas are the most often encountered etiology. However, especially in large and complex DVAs, other mechanisms have to be kept in mind. We have identified the following characteristics for symptomatic DVAs: large and complex DVAs with changes on MRI suggesting venous congestion, acute or subacute ischemic changes, asymmetrical appearance of the medullary zones, and association with true arteriovenous malformation. MRI is the diagnostic modality of choice to diagnose DVAs, their potential complications, and associated pathologies. Although not routinely performed in this series, diffusion and perfusion sequences will be helpful for detecting venous congestion. In our opinion, thrombotic complications of DVAs require the same treatment and laboratory workup as cortical venous or sinus thrombosis, i.e., anticoagulation treatment with investigation of procoagulating factors or prothrombotic conditions. Although MRI is sufficient for routine evaluation, we think that angiography can add to the understanding of the hemodynamics of the DVA, potential ruptured points, venous stenosis, and other associated pathologies such as dural arteriovenous shunts or AVMs. A superselective injection may be required if a conventional angiogram is not capable to define the diagnosis in suspicious cases (repetitive hemorrhages with focal hematoma, venous asymmetry).

**Conclusion**

The true incidence of vascular complications related to a DVA is unknown. DVAs should still be considered to be benign lesions, although in exceedingly rare cases, they can be symptomatic according to the aforementioned conditions. The pathomechanism should be identified for proper management. The integrity of the DVA needs to be preserved irrespective of the treatment of choice.

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None.

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