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

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.1,2 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.3,4 The DVAs have no proliferative potential, no direct arteriovenous shunts, and normal brain parenchyma between the dilated veins.5 DVAs serve as normal drainage routes of the brain tissue because the habitual venous drainage of their territory is absent. Their etiology and mechanism of development are unknown, but it is currently accepted that they act like a compensatory system of cerebral parenchyma venous drainage due to early failure, abnormal development, or an intrauterine occlusion of normal capillaries or small transcerebral veins.2

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,6 epilepsies are due to associated cortical dysplasias,7 and pseudotumoral effects can be secondary to associated lymphatic malformations.8

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
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>Seizures at birth</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, hemisensomnolence, 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>

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;15,16 (3) patients with DVAs and symptomatic cavernomas because of their established natural history (hemorrhage, epilepsy, mass effect);13,14,17–24 and (4) patients and series25,26 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></td>
<td>Conservative</td>
</tr>
<tr>
<td>2002—venous congestion</td>
<td>Bilateral cerebellum → left precentral cerebellar v. → VG</td>
<td></td>
<td>2002—Conservative</td>
</tr>
<tr>
<td>Congestive edema surrounding the venous collector</td>
<td>Large and complex</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</td>
<td>Thrombosis of venous collector (precentral cerebellar v.)</td>
<td>Conservative/spontaneous recovery</td>
</tr>
<tr>
<td>Venous infarction</td>
<td></td>
<td></td>
<td></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 → transinsular 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/no follow-up</td>
</tr>
<tr>
<td>Left basal hematoma</td>
<td>Left basal ganglia → inferior striate v. → basal v.</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/good recovery</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.
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. In brief, 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, 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 or a restricted outflow and were present in 49 patients.
<table>
<thead>
<tr>
<th>Reference</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 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 striate v. → ICV</td>
<td>Not reported</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>
<td>Endoscopic III ventriculostomy</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>
<td>Decompressive surgery</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>
<td>Conservative clinical follow-up unchanged</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>
<td>Decompressive surgery/good recovery</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>
<td>Conservative</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>
<td>Decompressive surgery/good recovery</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>
<td>Shunt/good recovery</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>
<td>Conservative</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>
<td>Conservative; clinically unchanged</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>
<td>Conservative</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 angiographic 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. Brain, Melsungen, Germany) injections. All patients recovered from their bleeding without new 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.

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 system, 24.1% (n = 7) had stenosis at some point of the DVAs drainage (Figure 5), 13.8% (n = 4) had complete thrombosis of the DVA with a systemic procoagulating factor or state (ie, puerperium), and 10.4% (n = 3) had complete thrombosis without an identifiable cause. This mechanical obstruction lead to venous congestion resulting in hemorrhagic or venous infarction in all symptomatic patients, which
### Table 3. Findings in All Patients Reported in the Literature With Increased Inflow Into the DVA

<table>
<thead>
<tr>
<th>Reference</th>
<th>Mechanism</th>
<th>Morphology</th>
<th>Age, 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><strong>Bergui, 1997</strong></td>
<td>Flow misbalance:</td>
<td>Parenchymal hemorrhage</td>
<td>35 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><strong>Bergui, 1997</strong></td>
<td>Increased inflow</td>
<td>Parenchymal hemorrhage</td>
<td>39 M</td>
<td>Headache, aphasis, right hemiparesis</td>
<td>Left temporomotor</td>
<td>Classical aspect without capillary ectasia</td>
<td>Inferior striate v. → basal v.</td>
<td>Medullary microshunts</td>
<td>Radiosurgery/partial recovery</td>
</tr>
<tr>
<td><strong>Kociyama, 1999</strong></td>
<td>Flow misbalance:</td>
<td>Intraventricular hemorrhage</td>
<td>22 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><strong>Koc, 2007</strong></td>
<td>Flow misbalance:</td>
<td>Intraventricular hemorrhage</td>
<td>56 M</td>
<td>Aphasia, right hemiparesis</td>
<td>Left temporal lobe</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, 19865</td>
<td>Flow misbalance:</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.</td>
<td>ICV</td>
<td>...</td>
</tr>
<tr>
<td>Yamamoto, 198970</td>
<td>Flow misbalance:</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, 19924</td>
<td>Flow misbalance:</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, 19957</td>
<td>Flow misbalance:</td>
<td>Congestive hemorrhage</td>
<td>34</td>
<td>F</td>
<td>Persistent headache, left hemianopsia</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, 199656</td>
<td>Flow misbalance:</td>
<td>Venous infarction</td>
<td>13</td>
<td>M</td>
<td>Ataxic gait, emesis, left hemiparesis, right mydriasis</td>
<td>Right temporoparietal</td>
<td>Large, complex DVA with thrombosis of venous collector</td>
<td>Labbé → transverse sinus</td>
<td>Paucity of alternate superficial venous drainage pathways</td>
<td>Decompressive craniotomy/death</td>
</tr>
<tr>
<td>Guerrero, 198660</td>
<td>Flow misbalance:</td>
<td>Venous infarction</td>
<td>62</td>
<td>M</td>
<td>Vertigo, diplopia, ataxia and occipital headache</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, 199654</td>
<td>Flow misbalance:</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, 199670</td>
<td>Flow misbalance:</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>Right-sided lower midbrain cavernous angioma</td>
<td>Conservative/mild residual cerebellar symptoms</td>
</tr>
<tr>
<td>Herbreteau, 199971</td>
<td>Flow misbalance:</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.</td>
<td>ICV</td>
<td>...</td>
</tr>
<tr>
<td>Lai et al, 199970</td>
<td>Flow misbalance:</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, 199972</td>
<td>Flow misbalance:</td>
<td>Venous infarction</td>
<td>25</td>
<td>F</td>
<td>Headache, seizures, and left hemianopsia 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, 20007</td>
<td>Flow misbalance:</td>
<td>Venous infarction</td>
<td>68</td>
<td>M</td>
<td>Headache, seizures, right hemiplegia, and aphasia</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>Hammadou, 20027</td>
<td>Flow misbalance:</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>SSS (direct)</td>
<td>Postpartum/ family history of thrombosis/ smoke/ contraceptives</td>
<td>Conservative/good recovery</td>
</tr>
<tr>
<td>Lovrencic-Huzjan, 200427</td>
<td>Flow misbalance:</td>
<td>Subarachnoid hemorrhage</td>
<td>39</td>
<td>M</td>
<td>Occipital headache, nausea and vomiting.</td>
<td>Cerebellum/SAS</td>
<td>Thrombosis of venous collector</td>
<td>Precentral cerebellar v. → VG</td>
<td>...</td>
<td>Conservative/good recovery</td>
</tr>
<tr>
<td>Peltier, 20048</td>
<td>Flow misbalance:</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

![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.](image-url)
subsequently leads to variations of the normal venous drainage. These variations are either constituted by dilated medullary veins, that converge into a large transcerebral collector, or by persistence of intrinsic venous anastomoses with an absence of normal draining veins in the region. In comparison to normal veins, histologically, DVAs are characterized by a composition of thin-walled vessels spread in normal neural parenchyma draining into a large caliber vein with a thicker wall without a smooth muscle layer nor elastic lamina.

On contrast-enhanced CT, the venous collector of the DVA is readily detectable as a linear or curvilinear focus of enhancement, typically coursing from the deep white matter to a cortical vein or a deep vein or to a dural sinus. On MRIs, DVAs typically have a transhemispheric flow void on both T1- and T2-weighted images. After the administration of gadolinium, because of the slow flow, significant enhancement of the caput medusae of the medullary veins and venous collector are observed. Digital subtraction angiography is rarely necessary to diagnose a DVA. Classical angiographic appearance is that of a caput medusae appearance of transmedullary veins visualized during the early to middle venous phase, draining into a large venous collector, which can extend either to the superficial or deep venous system depending on the type of the DVA.

DVAs are classified as deep (ie, draining into deep subependymal veins and the galenic system) or superficial (ie, draining into cortical veins). In 70%, the latter pattern is present, deep drainage is found in 20%, whereas a combination of deep and superficial drainage occurs in 10%. Apart from the classical appearance, complex DVAs can have multiple collectors, drain a large area, and can be associated with both deep and superficial drainage. In general, DVAs occur most often in the frontal lobe (36% to 56%) followed by the parietal (12% to 24%), occipital (4%), and the temporal lobes (2% to 19%); the cerebellum (14% to 29%); the basal ganglia (6%); the thalamus, the ventricles (11%); and the brainstem (less than 5%).

Radiological and autopsy series demonstrated that DVAs occur in 2.5% to 3% of the population and they are the most common vascular “malformation” of the central nervous system constituting approximately 60% of all vascular malformations, whereas capillary telangiectasias represent 20%, cavernomas 10%, AVMs 9%, and dural arteriovenous shunts 1% in larger autopsy series. They have been seen or diagnosed in patients presenting with symptoms such as seizures, vertigo, syncope, tinnitus, and headaches; however, because these symptoms are among the most often to lead to MRI investigation, a direct relationship could not be established and it is currently accepted that they represent an incidental finding in the vast majority of cases.

DVAs are considered as stable and benign conditions; however, as an anatomic variation, they occur in atypical locations and they may be less flexible to changes of the intracranial venous equilibrium. We classified both our symptomatic DVAs and those reported in the literature

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>Uchino, 1996</td>
<td>Idiopathic</td>
<td>Venous infarction</td>
<td>53</td>
<td>M</td>
<td>Seizures</td>
<td>Right frontal</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/variations.

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 obstruc-
tion 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 transparentchymal 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-
recanalization of the venous collector will presumably prevent this complication.\textsuperscript{55} 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.\textsuperscript{53,56}

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.\textsuperscript{51,53} Dural arteriovenous shunts invariably increase the pressure in the dural venous sinuses. This causes mld 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.\textsuperscript{57} 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.\textsuperscript{58} 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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**Disclosures**

None.

**References**


Pathomechanisms of Symptomatic Developmental Venous Anomalies
Vitor M. Pereira, Sasikhan Geibprasert, Timo Krings, Thaweesak Aurboonyawat, Augustin Ozanne, Frederique Toulgoat, Sirintara Pongpech and Pierre L. Lasjaunias

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