Natural History of Dural Arteriovenous Shunts

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Background and Purpose—Dural arteriovenous shunts with cortical venous reflux or drainage may cause neurological symptoms and death with or without intracranial hemorrhage. Present knowledge about the natural history of these lesions is limited, however. We investigated the incidences of intracranial hemorrhage, progressive dementia syndrome, and death in patients diagnosed in our neurovascular center.

Methods—We evaluated the records of 85 patients with dural arteriovenous shunts with cortical venous drainage or reflux hospitalized in our institution from 1978 to 2007. The annual incidences of intracranial hemorrhage, progressive dementia syndrome, and death were calculated.

Results—Fifty-three patients did not have an intracranial hemorrhage as the presenting event. One of these patients bled after diagnosis. Thirty-two patients had an intracranial hemorrhage as the presenting event. Three patients bled after diagnosis. One of these patients died. Apart from deficits caused by hemorrhage, no patient reported adverse neurological symptoms. In patients presenting with an intracranial hemorrhage the annual risk for hemorrhage is approximately 7.4% and in those not presenting with a hemorrhage it is approximately 1.5%.

Conclusion—The risk of intracranial hemorrhage from a dural arteriovenous shunt with cortical venous drainage is most likely smaller than previously proposed. Presentation with hemorrhage is a risk factor for hemorrhage. The risks of developing neurological symptoms not related to hemorrhage are also less than previously reported. (Stroke. 2008;39: 1735-1739.)

Key Words: dural arteriovenous fistula □ dural arteriovenous shunts □ hemorrhage □ natural course □ natural history

Dural arteriovenous shunts (DAVSs), also called dural arteriovenous fistulae or malformations, are acquired lesions in the dura mater. In the adult population, intracranial DAVSs are approximately 10 times less frequent than brain arteriovenous malformations. Common revealing events are pulse-synchronous bruit or neurological symptoms secondary to intracranial hemorrhage. The venous drainage pattern of the shunt is crucial in the risk assessment of these lesions. DAVSs with direct or indirect cortical venous drainage (CVD) are considered to carry a high risk for hemorrhage that mandates prompt diagnosis and treatment. However, data on the natural history are limited, the most complete reference on the subject being based on a long-term follow-up of 14 untreated and 6 partially treated patients. The aim of this study was to assess the natural course of untreated DAVSs with cortical venous drainage, in particular the risk for intracranial hemorrhage.

Materials and Methods

Hospital neurovascular and radiosurgical databases were searched for patients with DAVS diagnosed during the era of CT scanning in our hospital. From 1993 onward, data have been collected prospectively in a dedicated neurovascular database. Data about patients admitted for the first time before 1993 were collected from the database of the gamma knife unit and from the databases of individual vascular neurosurgeons.

We found and evaluated the radiological and clinical records of 163 consecutive patients with DAVSs hospitalized in our institution from January 1978 to the end of June 2007. Hospital files were scrutinized for the date and nature of the presenting event, for prior and subsequent hemorrhagic episodes, and for evidence of progressive neurological deficit. All intracranial hemorrhages had been confirmed with CT or MRI.

When data were missing or ambiguous, we contacted the patients or his or her relatives directly for additional information. All pertinent neuroradiological examinations were meticulously reexamined. Thus, each angiographic workup was reviewed by 2 of the authors (M.S. and L.P.) with special emphasis on presence of CVD.

For each patient, the day of the first therapeutic attempt was the study end point. Patients who did not undergo treatment were followed until death or until the end of June 2007. Causes of death were investigated for every patient who died before the end of the study.

Venous Drainage Patterns

We analyzed the venous drainage patterns of the DAVSs in the 163 patients with DAVS with regard to the presence or absence of cortical venous drainage or reflux. Ninety-nine patients had 100
DAVSs with direct or indirect reflux to the cortical venous system. The DAVS classification systems of Borden and Cognard take the venous drainage pattern into consideration.5,7 All 100 DAVSs with CVD corresponded to Borden types II and III, which is equivalent to Cognard types IIb, IIa+b, III, and IV. That is, they drained either directly to a cortical vein or first to a dural sinus and secondarily to a cortical vein. The patient with 2 DAVSs was in the present study considered as a single patient. No patient fulfilling the inclusion criteria was lost to follow up.

Exclusion Criteria
Fourteen patients had their first angiogram in an institution outside of the primary recruitment area of our center and were sent to our institution only at a later stage for treatment. These patients were excluded from the study. The remaining 85 patients comprise the material in this study.

Total Time at Risk
The total time at risk, ie, the time period when the patient had a DAVS with CVD present, is comprised of 3 different periods. The first period, before diagnosis, began with the debut of symptoms or with no nonangiographic neuroradiological examination indicating the presence of a DAVS and ended with the conventional angiography that verified the diagnosis.

The second period, the radiological follow-up period, began with the diagnostic conventional angiography and lasted until the last conventional angiography was performed with the DAVS still present or until the time of treatment. In almost all cases, the last angiography before treatment was done immediately before endovascular or gamma knife therapy. The third period, the clinical follow-up period, began with the last angiography showing the DAVS and concluded with the death of the patient, angiographic proof of spontaneous DAVS closure, or the end of the study period, ie, June 2007.

Statistical Methods
The event rates for intracranial hemorrhage, progressive dementia syndrome, and death were calculated per patient year and shown as percentages. The $\chi^2$ test ($df=1$) was used to compare categorical data. All probability values lower than 0.01 were counted for as $<0.01$.

Results

Demographics
Forty-nine of the 85 patients (58%) were male. The mean age at diagnosis was 58 years. Seventeen of the patients were diagnosed during the period 1978 to 1992 and the remaining 68 patients were diagnosed with a DAVS during 1993 to 2007.

No patient had any history of intracranial hemorrhage or DAVS before the study period. No patient had been subjected to any previous neuroradiological examinations (ie, angiography) proving the absence or presence of a DAVS. Follow-up data were available for all patients.

Locations and Classification
The DAVSs were found in various locations with preponderance for the sigmoid–transverse complex and parietal–occipital dura mater. Thirty-two of the lesions were classified as Borden II and 53 as Borden III. For details, see the Table.

Presentation
Thirty-two patients presented with intracranial hemorrhages. Thirteen presented with a subarachnoid hemorrhage, 17 with cerebral or cerebellar hemorrhages, one with a combined subarachnoid and intracerebral bleed, and one with an intraventricular bleed. Eight of the patients who presented with a hemorrhage had a Borden type II DAVS, whereas 24 patients had a DAVS classified as Borden type III.

Of the 53 patient not presenting with an intracranial hemorrhage, 22 presented with an objective pulse-synchronous bruit. In 12 patients, the DAVS was an incidental finding at a neuroradiological workup for unrelated reasons. The remaining 19 patients presented with various symptoms, one of them revealing a progressive dementia syndrome.

In this cohort, 24 patients had a Borden type II DAVS and 29 patients had a Borden type III DAVS. Mean ages were similar between patients with and without hemorrhage as the presenting event.

Incidence of Hemorrhage Before Diagnosis
Two of the 32 patients presenting with hemorrhage bled a second time before diagnosis, one a few days and the second a few weeks after the first hemorrhage. Time at risk for these patients, from the first bleed to diagnosis, was 6.3 patient-years; thus, the annual incidence of repeated hemorrhage before diagnosis was 3.3%.

None of the 53 patients without intracranial hemorrhage as the presenting event bled before angiographic diagnosis. Time at risk was 38 patient-years and the annual incidence of intracranial hemorrhage was consequently 0.2%. The difference between the 2 groups was significant at the $P<0.01$ level.

Incidence of Hemorrhage or Dementia During the ‘Radiological Follow-Up Period’
In 4 patients, diagnostic angiography was performed after which the patients were only followed clinically. Consequently, it was possible to define a “radiological follow-up period” beginning and ending with a conventional angiography in 81 patients comprising a total of 49.6 patient-years (mean, 0.60 years per patient). During this time, 3 patients had an intracranial hemorrhage, and 2 of these had also presented with a hemorrhage. One patient bled 3 weeks and the second one had his hemorrhage 2 years 8 months after the initial intracranial bleed. The third patient had dysgraphia and excessive fatigue as presenting events and had his first

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### Table. All 85 DAVSs According to Location and Drainage

<table>
<thead>
<tr>
<th>Localization</th>
<th>Borden II</th>
<th>Borden III</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sigmoid–transverse sinus</td>
<td>22</td>
<td>3</td>
</tr>
<tr>
<td>Petrosal sinus</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Midline sinus</td>
<td>2</td>
<td>4</td>
</tr>
<tr>
<td>Cavernous sinus</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>Temporal</td>
<td>0</td>
<td>7</td>
</tr>
<tr>
<td>Tentorial</td>
<td>1</td>
<td>16</td>
</tr>
<tr>
<td>Parietal–occipital</td>
<td>1</td>
<td>6</td>
</tr>
<tr>
<td>Anterior fossa</td>
<td>0</td>
<td>5</td>
</tr>
<tr>
<td>Other</td>
<td>0</td>
<td>8</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>32</strong></td>
<td><strong>53</strong></td>
</tr>
</tbody>
</table>
hemorrhage 3 years 4 months after diagnosis by cerebral angiography.

In 3 patients, the DAVS had occluded spontaneously before a second angiography. One of those patients harbored 2 separate DAVSs, both of which had occluded at the time of the second angiography 9 years 8 months after the initial diagnostic investigation. In another patient, the DAVS spontaneously occluded during the 7 months between his 2 last angiographies performed. Lastly, in a third patient, the DAVS occluded during the 3 months in between 2 angiographies. The timespans until the last angiographies were considered radiological follow-up periods and were used as such in the risk calculations. There were no deaths or additional neurological deficits in the radiological follow-up period besides those that were caused by the intracranial hemorrhages nor were there any new presentations of progressive dementia syndrome.

In this cohort that represented the radiological follow-up period, the annual incidence of hemorrhage was 12% for the 30 patients who presented with an intracranial hemorrhage (time at risk=16.3 patient-years), and it was 3.0% for the 51 patients with a nonhemorrhagic presentation (time at risk=33.3 patient-years). The annual incidence of an intracranial hemorrhage in the complete cohort of 81 patients was 6.1% (time at risk=49.6 patient-years). The difference between the 2 groups was significant at the P<0.05 level.

Incidence of Hemorrhage or Dementia During the ‘Clinical Follow-Up Period’

Five patients with Borden type III DAVSs were followed clinically until death. Four of those patients refused any treatment or radiological follow-up after the diagnostic angiography. One patient had one follow-up angiogram performed that showed the DAVS to be still patent, but after that refused treatment and further angiographic controls.

The first of these patients who refused treatment presented with a temporal lobe hemorrhage and was subsequently diagnosed with a DAVS in 1979. She died of a deep frontal–parietal hemorrhage in 1991. At that time, the patient was on long-term anticoagulation with warfarin because of problems with lower limb venous thrombosis. The second patient died because of a ruptured aortic aneurysm, the third from severe alcohol abuse, the fourth from a traumatic event, and the fifth patient died of congestive heart failure. There were no indications of an intracranial hemorrhage in these last 4 patients during the clinical follow-up period. None of these 5 patients developed progressive neurological symptoms.

The follow-up was 58.5 patient-years (mean=11.7 patient-years per patient), during which time one patient had a bleed and died. Thus, the annual incidence of intracranial hemorrhage as well as the mortality was 1.7% in this cohort. The limited number of patients did not permit a statistical test with reasonable power.

Association Between Hemorrhage and Prior Hemorrhage

Four patients had an intracranial hemorrhage after the diagnostic angiography but before treatment, DAVS obliteration, or patient death. The total follow-up time for the 85 patients was 108 patient-years. In the cohort of 32 patients who had an intracranial hemorrhage as the presenting event, 3 patients had a second hemorrhage. Time at risk was 40.4 patient-years, and the annual incidence of intracranial hemorrhage was 7.4%.

In the cohort of 53 patients who did not have an intracranial hemorrhage as the presenting event, one patient had an intracranial hemorrhage during this period. Time at risk was 67.1 patient-years and the annual incidence for intracranial hemorrhage was 1.5%. The difference between the 2 groups was significant at the P<0.05 level.

Comparison to Previous Studies

The calculated incidence in our material for an intracranial hemorrhage, progressive neurological deficit, and death after presentation of the DAVS was compared with that of van Dijk et al. The incidences were significantly lower in our material (P<0.01).

The calculated incidence in our material for a second bleeding during the first 2 weeks after presentation with hemorrhage was compared with that of Duffau et al. Also in this comparison, the incidence was significantly lower in our material (P<0.01).

Discussion

Previous Studies

Intracranial DAVSs draining into the cortical or deep venous system of the brain may cause intracranial hemorrhage, sometimes with severe consequences. Nonhemorrhagic neurological symptoms, including pulse-synchronous bruit, are also frequent. There are several published studies concerning DAVSs with a significant number of patients, although they focus mainly on the presenting event. Apart from case reports, we know of 4 published studies regarding the prognosis for patients harboring a DAVS, in particular the risk of intracranial hemorrhage.

Brown and colleagues followed 52 patients with DAVSs over a mean period of 6.6 years. They reported an annual risk of hemorrhage of 1.8% but did not discriminate between lesions with CVD and without. Among their patients, 14 had “lesions draining into leptomeningeal veins” and in that group, 3 had an intracranial hemorrhage. However, the presence of leptomeningeal drainage was not significantly associated with the occurrence of such bleeds.

Davies et al followed 14 nontreated patients with Borden type II and III DAVSs over a total period of 21 patient-years. They reported an annual hemorrhage rate of 19% per lesion year. Van Dijk and colleagues studied the same patient cohort as Davies et al over a substantially longer period of time and added 6 patients with CVD that persisted despite treatment. The total follow-up time in this study was 87 patient-years with an annual hemorrhage rate of 8.1%, a progressive dementia syndrome rate of 6.9%, and accordingly an annual event rate of 15%. The annual mortality rate was 10%. Duffau et al investigated 20 patients with an intracranial hemorrhage from a DAVS and followed the patients over a mean time of 20 days after the presenting hemorrhage, reporting a 35% early rebleeding rate.
Knowledge about the natural course of the disease is essential for correct management of these patients. However, as discussed previously, current understanding of the natural history is based mainly on the findings in the 2 small single-center series by Duffau et al and van Dijk et al.\textsuperscript{3,4}

**Presentation**

In our material, there was a relative overrepresentation of Borden type III DAVSs among those who presented with intracranial hemorrhage. However, because these lesions usually do not create a bruit, they will quite often go undetected until they cause a hemorrhage. For these patients, time at risk before the initial bleed is therefore often unknown. Thus, this overrepresentation can be interpreted in several ways, one being an increased risk of hemorrhage and another being the lack of other means of disclosure for the DAVS. The hemorrhage incidence of 1.7% in the "clinical follow-up group," in which all 5 patients had a Borden type III DAVS, does not support the hypothesis of a high risk of hemorrhage from these lesions.

**Time at Risk**

The time at risk comprised 3 different periods. The first period was from the first indication of a DAVS until diagnosis by angiography. Any number of patients may have dropped out before diagnosis and this data has limited value.

The period from diagnosis until censoring was split into the radiological follow-up period and the clinical follow-up period. The rationale is that DAVS may close spontaneously.\textsuperscript{3,4,13,15} Therefore, the time at risk is certain only if there is a neuroradiological examination at the end of the period proving the DAVS is still present. In the patients who died from other causes or refused further radiological follow-up, we do not know for certain whether the DAVS was present until censoring. Therefore, the 2 periods have different weight as evidence.

**Incidence of Hemorrhage and Progressive Dementia After Diagnosis**

In the present study, the incidence of a second hemorrhage after diagnosis was 7.4% for patients with a bleeding as the presenting event. For the cohort of patients that did not present with a hemorrhage, the incidence was 1.5%. In the entire material, there was only one death possibly related to the DAVS and no cases of progressive dementia syndrome.

Two patients who presented with an intracranial hemorrhage bled a second time before diagnosis. There were no bleedings before diagnosis in the cohort not presenting with hemorrhage. However, these data are not fully reliable, because any number of patients could have had a second hemorrhage and died before diagnosis. Despite these objections, these findings indicate that the risk of hemorrhage may be higher in patients who have experienced a previous hemorrhage.

Van Dijk et al had an annual hemorrhage rate of 8.1% in their material. There was a significant ($P<$0.05) difference between their results and the ones from the present study regarding the risk for a hemorrhage from a DAVS with CVD. Furthermore, only 25% of their 20 patients had presented with an intracranial hemorrhage, which makes the disparity between the studies even more remarkable.\textsuperscript{4} Our data also do not concur with the 35% early rebleeding rate reported by Duffau et al.\textsuperscript{3} The difference was significant at the $P<$0.01 level.

The reasons why the results are different in the present study are not clear. Several explanations are possible such as demographic differences, inclusion and publication bias. However, both the number of patients included and the calculated patient-years at risk are higher in our study, which can be argued to be in favor of a greater reliability.

Our study highlights the fact that in DAVS with CVD, much like the situation for brain arteriovenous malformations, hemorrhage is a risk factor for hemorrhage.\textsuperscript{14} In our material, the annual incidence of hemorrhage was approximately 7.4% for patients presenting with an intracranial hemorrhage. In those not presenting with a hemorrhage, it was approximately 1.5%.

This finding is not really surprising, because the lesions are similar in that they drain into the intracranial venous system. Both shunts can therefore be the cause of an increased venous load that may secondarily create weak spots prone to rupture. To summarize, the natural course of DAVSs with CVD is most probably more benign than previously proposed, in particular for patients with a presenting event other than an intracranial hemorrhage.

**Management**

Previous publications have proposed a dismal prognosis in DAVSs with CVD.\textsuperscript{3,4,11,13} This has had impact on management recommendations.\textsuperscript{15,16} Many patients are probably treated rather rapidly after diagnosis, often by transarterial or transvenous embolization supplemented by surgery and, more rarely, radiosurgery.\textsuperscript{15–18} All treatments obviously involve a risk for complications, a risk that has to be balanced against the natural course.\textsuperscript{2,17,19–22}

Because the natural course probably is less perilous than previously assumed, DAVS management algorithms may have to be revised toward a less aggressive approach. This is in particular the case for patients with lesions that have never bled and where the treatment alternatives that can be offered are considered to entail a substantial risk for complications.

**Limitations to the Study**

This is a single-center study over a timespan of approximately 25 years. Criteria for inclusion into the databases (or personal patient series) on which the study relies have obviously changed over time. Some patients who were managed in the beginning of the study period have for a number of reasons almost certainly never been entered into the databases. This may be the reason for apparent increase in detection rate of patients with DAVS. Three patients experienced spontaneous closure of the DAVSs proven at angiography. It is unclear exactly when the obliteration happened. The same uncertainty applies for the patients who died without any follow-up angiography, including the patient who died from a cerebral hemorrhage. Accordingly, the time at risk may be slightly shorter than calculated and the risk of hemorrhage therefore correspondingly higher.
Conclusion
The risk of intracranial hemorrhage from a dural arteriovenous shunt with cortical venous drainage is most likely smaller than previously proposed. Presentation with hemorrhage is a risk factor for hemorrhage. The risks of developing neurological symptoms not related to hemorrhage are also less than previously reported.

Disclosures
None.

References
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