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Received, April 16, 2008.
Accepted, August 26, 2008.

CRANIAL DURAL ARTERIOVENOUS FISTULAE: ASYMPTOMATIC CORTICAL VENOUS DRAINAGE PORTENDS LESS AGGRESSIVE CLINICAL COURSE

OBJECTIVE: Cranial dural arteriovenous fistulae (dAVF) with cortical venous drainage (CVD) (Borden Types 2 and 3) are reported to carry a 15% annual risk of intracranial hemorrhage (ICH) or nonhemorrhagic neurological deficit (NHND). The purpose of this study was to compare the clinical course of Type 2 and 3 dAVFs that present with ICH or NHND with those that do not.

METHODS: Twenty-eight patients with Type 2 or 3 dAVFs were retrospectively evaluated. CVD was classified as asymptomatic (aCVD) if patients presented incidentally or with pulsatile tinnitus or orbital phenomena. CVD was classified as symptomatic (sCVD) if patients presented with ICH or NHND. Occurrence of new ICH or new or worsening NHND between diagnosis and disconnection of CVD or last follow-up (if not disconnected) was noted. Overall frequency of events was compared using Fisher's exact test. Cumulative, event-free survival was compared using Kaplan-Meier analysis with log-rank testing.

RESULTS: Of 17 patients with aCVD, 1 (5.9%) developed ICH and none experienced NHND or death during the median 31.4-month follow-up period. Of 11 patients with sCVD, 2 (18.2%) developed ICH and 3 (27.3%) experienced new or worsened NHND over the median 9.7-month follow-up period. One of these patients subsequently died. Overall frequency of ICH or NHND was significantly lower in patients with aCVD versus sCVD ($P = 0.022$). Respective annual event rates were 1.4 versus 19.0%. aCVD patients had significantly higher cumulative event-free survival ($P = 0.0016$).

CONCLUSION: Cranial dAVFs with aCVD may have a less aggressive clinical course than those with sCVD.

KEY WORDS: Central nervous system vascular malformation, Clinical course, Cortical venous drainage, Cranial dural arteriovenous fistula, Outcome

Neurosurgery 64:241–248, 2009 DOI: 10.1227/01.NEU.0000338066.30665.B2 www.neurosurgery-online.com

Cranial dural arteriovenous fistulae (dAVF) are direct shunts between dural arteries and a cortical vein or dural sinus (37). Most dAVFs are idiopathic. Others are associated with antecedent craniotomy, trauma, or dural sinus thrombosis and may result from aberrant dural angiogenesis (2, 6). Clinical presentation depends primarily on the location and pattern of venous drainage (25). dAVFs may be found incidentally or may present with symptoms of increased dural sinus drainage such as pulsatile tinnitus or orbital phenomena. The latter are related to elevated cavernous sinus flow and include ophthalmoplegia, proptosis, chemosis, retro-orbital pain, and decreased visual acuity (27, 41). More severe presentations include intracranial hemorrhage (ICH) and nonhemorrhagic neurological deficits (NHND) such as progressive dementia, seizures, parkinsonism, cerebellar symptoms, and other focal deficits. ICH and NHND are attributable to cortical venous hypertension (21, 22, 25, 28, 30, 31, 35).

ABBREVIATIONS: aCVD, asymptomatic cortical venous drainage, **BI**, Barthel Index; **CVD**, cortical venous drainage; **dAVF**, dural arteriovenous fistulae; **ICH**, intracranial hemorrhage; **mRS**, modified Rankin Scale; **NHND**, nonhemorrhagic neurological deficit; **sCVD**, symptomatic cortical venous drainage

TABLE 1. Classification of cranial dural arteriovenous fistulae[a]

Classification	Venous drainage site	Flow characteristics
Borden et al. (3)		
1	Dural venous sinus	No CVD
2	Dural venous sinus	Reflux into cortical veins (CVD)
3	Cortical vein	CVD
Cognard et al. (7)		
I	Dural venous sinus	Antegrade flow within sinus, no CVD
IIa	Dural venous sinus	Retrograde flow within sinus, no CVD
IIb	Dural venous sinus	Antegrade flow within sinus, reflux into cortical veins (CVD)
IIa+b	Dural venous sinus	Retrograde flow within sinus, reflux into cortical veins (CVD)
III	Nonectatic cortical vein	CVD
IV	Ectatic cortical vein	CVD
V	Spinal perimedullary vein	CVD

[a] CVD, cortical venous drainage.

dAVFs sharing venous drainage with the normal cortical circulation have been associated not only with ICH and NHND at presentation (1, 4, 14, 32), but also with poor subsequent clinical course if not treated (14, 41). The grading systems of Borden et al. (3) and Cognard et al. (7) differentiate dAVFs on the basis of their venous drainage (Table 1). Borden Type 1 dAVFs are without cortical venous drainage (CVD). They are generally benign and treated only when associated with intolerable tinnitus or orbital phenomena (12, 16). Borden Type 2 and 3 dAVFs have CVD. Urgent treatment has been recommended for all Type 2 and 3 dAVFs, given the reported high rate of future ICH or NHND in partially treated and untreated lesions (ranging from 15% per year to 35% within 20 days of presentation) (14, 41). However, the number of patients with Type 2 and 3 dAVFs in whom the clinical course after initial presentation was specifically examined is very small (i.e., 40 patients from 2 published studies), and nearly all of these patients had presented with ICH or NHND (14, 41). Yet, many Type 2 and 3 dAVFs are discovered incidentally or present with isolated symptoms of increased dural sinus drainage. The clinical course and appropriate management of these lesions remain unclear. The purpose of our study was to further validate the reported high risk associated with Type 2 and 3 dAVFs presenting with symptoms of cortical venous hypertension (referred to as symptomatic CVD [sCVD]) and to compare this risk with the risk of those presenting incidentally or with isolated symptoms of increased dural sinus drainage (referred to as asymptomatic CVD [aCVD]).

PATIENTS AND METHODS

After institutional review board approval, neurosurgery and interventional neuroradiology records were searched for all patients with dAVFs diagnosed, evaluated, or treated at our institution between 1996 and 2007. We identified 108 consecutive patients with dAVFs. Their angiographic reports were reviewed for documentation of the presence or absence of CVD. Thirty-four patients were reported to have CVD, and the remainder were excluded from the study. Catheter angiograms obtained at the time of presentation were reviewed to determine dAVF location and confirm CVD in these 34 patients. The lesions were classified using the Borden et al. (3) and Cognard et al. (7) scales.

Patients were divided into 2 groups on the basis of a review of medical records: those presenting with aCVD (incidental finding, pulsatile tinnitus, or orbital phenomena) and those presenting with sCVD (ICH or NHND, defined as neurological symptoms other than pulsatile tinnitus and orbital phenomena without evidence of hemorrhage). Medical records were reviewed for endovascular, surgical, or radiosurgical treatments and associated complications. Follow-up angiograms were reviewed for persistence or elimination of CVD. For each patient, a follow-up period was defined a priori as the interval between angiographic diagnosis and either elimination of CVD (for dAVFs with CVD eliminated) or last follow-up (for dAVFs with persistent CVD). The clinical course during this period was then assessed using a review of medical records and a telephone survey. Occurrence of new ICH, new or worsening NHND, or death was specifically noted. Levels of disability and independence at the time of the last follow-up evaluation were quantified using the modified Rankin Scale (mRS) and Barthel Index (BI), respectively. mRS and BI outcomes were classified as favorable (mRS score, $\leq$1; BI score, $\geq$95) or unfavorable (mRS score, $\geq$2; BI score, $\leq$90).

Age at the time of presentation was compared using 2-tailed Student's *t* test. Sex distribution and frequencies of ICH or NHND (excluding events at presentation), favorable mRS, and favorable BI were compared using Fisher's exact test. Yearly event rates were calculated, and cumulative event-free survival was compared using Kaplan-Meier analysis with log-rank testing.

RESULTS

Of the 34 patients with Type 2 or 3 dAVFs, 5 patients (all with sCVD) underwent surgical or endovascular disconnection of CVD within 1 week of diagnosis. One additional patient did not present for follow-up after the incidental finding of a dAVF. These 6 patients were excluded from the study, leaving a total of 28 patients followed with persistent CVD. There was no significant difference in age between patients with aCVD versus those with sCVD (mean age, 55 versus 63 years; $P = 0.096$). Patients with aCVD were more frequently female (11 [65%] of 17 patients versus 3 [27%] of 11 patients), although the difference was not significant ($P = 0.060$).

Of the 17 patients with aCVD (Table 2), all were offered treatment. This advice was followed by 13 patients and declined by 4 patients. Of those accepting treatment, 10 underwent endovascular embolization alone, 2 underwent endovascular embolization followed by stereotactic radiosurgery, and 1 underwent endovascular embolization followed by surgery. Complete elimination of CVD was achieved in 8 patients, whereas persistent CVD was noted in

TABLE 2. Clinical and imaging features, treatment, and outcome of dural arteriovenous fistulae with asymptomatic cortical venous drainage[a]

Patient no.	Age (y)/sex	Presentation	Location	Classification		Treatment	CVD eliminated?	F/u (mo)	New events	mRS	BI
				Borden et al. (3)	Cognard et al. (7)						
1	50/F	Incidental	R tentorium	2	IIb	$Emb_{TA, TV}$	No	166.8	SAH	1	100
2	41/F	Incidental	L occipital area	3	III	Emb_{TA}	Yes	5.3	None	0	100
3	72/F	L pulsatile tinnitus	L transverse sinus	2	IIb	Emb_{TV}	Yes	1.7	None	0	100
4	57/F	B pulsatile tinnitus	L transverse sinus	2	IIb	$Emb_{TA, TV}$	Yes	9.7	None	0	100
5	71/M	Incidental	Torcular	3	III	Emb_{TA}, Surg	Yes	24.8	None	0	100
6	33/F	L pulsatile tinnitus	L transverse sinus	2	IIb	$Emb_{TA, TV}$	Yes	31.4	None	0	100
7	73/F	Incidental	Clivus	3	III	None	No	38.6	None	1	100
8	33/F	Incidental	R petrous region	3	III	None	No	53.6	None	1	100
9	73/F	L pulsatile tinnitus	L sigmoid sinus	2	IIa+b	$Emb_{TA, TV}$	No	105.0	None	0	100
10	52/M	L pulsatile tinnitus	L transverse sinus	2	IIa+b	$Emb_{TA, TV}$	Yes	4.7	None	1	100
11	68/M	Incidental	R transverse sinus	3	III	None	No	132.1	None	0	100
12	46/F	L pulsatile tinnitus	L cranial base	3	III	Emb_{TA}	No	67.1	None	0	100
13	64/F	R pulsatile tinnitus	R sigmoid sinus	2	IIa+b	$Emb_{TA, TV}$	Yes	12.6	None	0	100
14	37/M	R pulsatile tinnitus	R cavernous sinus	3	IV	Emb_{TA}, SRS	No	22.3	None	0	100
15	47/F	L pulsatile tinnitus	L sigmoid sinus	3	III	$Emb_{TA, TV}$	Yes	15.7	None	1	100
16	55/M	Incidental	L frontal region	3	III	None	No	97.2	None	0	100
17	69/M	↓ acuity L eye	L petrous apex	2	IIa+b	Emb_{TA}, SRS	No	51.2	None	1	95

[a]CVD, cortical venous drainage; F/u, follow-up time; mRS, modified Rankin Scale score at last follow-up; BI, Barthel Index score at last follow-up; R, right; $Emb_{TA, TV}$, transarterial and transvenous embolization; SAH, subarachnoid hemorrhage; L, left; Emb_{TA}, transarterial embolization; Emb_{TV}, transvenous embolization; B, bilateral; Surg, surgery; SRS, stereotactic radiosurgery; ↓, decreased.

5 patients. Among those who underwent treatment, the median time between angiographic diagnosis and the first therapeutic procedure was 9.4 months, with the most common reasons for delayed therapy being delayed referral from an outside hospital and patient preference. For the entire cohort, the median follow-up time was 31.4 years. One (5.9%) of the 17 patients experienced ICH 13 years after refusing treatment. The dAVF was then embolized with minimal residual CVD, and the patient was independent and without disability at the 11-month follow-up examination. There were no cases of NHND or death during the observation period.

Of the 11 patients with sCVD (Table 3), all were offered treatment. This advice was followed by 10 patients and declined by 1 patient. Of those accepting treatment, 5 underwent endovascular embolization alone, 1 underwent surgery alone, and 4 underwent endovascular embolization followed by surgery. Complete elimination of CVD was achieved in 7 patients, whereas persistent CVD was noted in 4 patients. Among those who underwent treatment, the median time between angiographic diagnosis and the first therapeutic procedure was 8.4 months, with the most common reasons for delayed therapy again being delayed referral from an outside hospital and patient preference. For the entire cohort, the median follow-up time was 9.7 months. During this period, ICH occurred in 2 patients (18.2%) and new or worsened NHND occurred in 3 patients (27.3%). Of the latter 3 patients, 1 experienced progressive dementia associated with an increased magnetic resonance imaging T2 signal throughout the bilateral hemispheres, consistent with venous hypertensive encephalopathy; 1 patient experienced progressive dementia, dysarthria, and ataxia without follow-up imaging; and the third developed cortical vein thrombosis leading to tonicoclonic seizures, which were complicated by aspiration pneumonia, and the patient died. Three of the recorded events occurred before treatment, and 2 occurred after partial treatment with persistent CVD.

Excluding events at presentation, the overall frequency of ICH or NHND was significantly lower in the aCVD group than the sCVD group (1 [5.9%] of 17 patients versus 5 [45.5%] of 11 patients; $P = 0.022$). The respective annual event rates were 1.4 versus 7.6% for ICH, 0 versus 11.4% for NHND, and 1.4 versus 19.0% for either event. Annual mortality rates were 0 versus 3.8%. Cumulative event-free survival (Fig. 1) was significantly higher in the aCVD group ($P = 0.0016$, log-rank test). Patients with aCVD more commonly had a favorable mRS score (17 [100%] of 17 patients versus 4 [36%] of 11 patients; $P = 0.00028$) and BI (17 [100%] of 17 patients versus 6 [55%] of 11 patients; $P = 0.0047$) at the time of the last follow-up evaluation.

There were 2 complications of the 38 endovascular treatments. In 1 patient, diffuse migration of liquid embolization material into the left hemispheric venous system led to acute

TABLE 3. Clinical and imaging features, treatment, and outcome of dural arteriovenous fistulae with symptomatic cortical venous drainage[a]

Patient no.	Age (y)/sex	Presentation	Location	Classification		Treatment	CVD eliminated?	F/u (mo)	New/worsened events	mRS	BI
				Borden et al. (3)	Cognard et al. (7)						
18	74/F	L temporal IPH	L transverse sinus	3	III	$Emb_{TA,}$ Surg	Yes	0.5	IPH	2	100
19	49/M	R basal ganglia IPH	R petrous region	3	III	None	No	83.5	None	4	45
20	65/F	Dementia, ataxia, R HH	L transverse sinus	2	IIb	$Emb_{TA,}$ Surg	Yes	6.8	None	1	100
21	63/M	L parietal IPH	L tentorium	3	III	Emb_{TA}	Yes	9.7	None	0	100
22	67/M	Dementia, dysarthria, ataxia	L sigmoid sinus	2	IIa+b	Emb_{TA}	Yes	11.9	Dementia	4	75
23	73/F	Dementia, ↓ L hearing	L transverse sinus	2	IIa+b	$Emb_{TA, TV}$	Yes	61.9	None	1	100
24	46/M	R cerebellar hemorrhage	R petrous apex	3	III	Emb_{TA}	No	60.1	None	2	95
25	68/M	Dysarthria, ataxia	B transverse sinuses	2	IIa+b	$Emb_{TA, TV,}$ Surg	No	65.9	Dementia, dysarthria, ataxia	3	80
26	59/M	Seizure, R HH	L transverse sinus	3	IV	$Emb_{TA,}$ Surg	Yes	5.5	None	0	100
27	60/M	L< R quadriparesis	R tentorium	3	V	Surg	Yes	0.6	IPH	5	5
28	69/M	R HH	Sup sag sinus	2	IIb	$Emb_{TA, TV}$	No	4.2	CVT, death	6	0

[a] CVD, cortical venous drainage; F/u, follow-up time; mRS, modified Rankin Scale score at last follow-up; BI, Barthel Index score at last follow-up; L, left; IPH, intraparenchymal hemorrhage; $Emb_{TA,}$ transarterial embolization; Surg, surgery; R, right; HH, homonymous hemianopsia; ↓, decreased; $Emb_{TA, TV,}$ transarterial and transvenous embolization; B, bilateral; Sup sag, superior sagittal; CVT, cortical vein thrombosis.

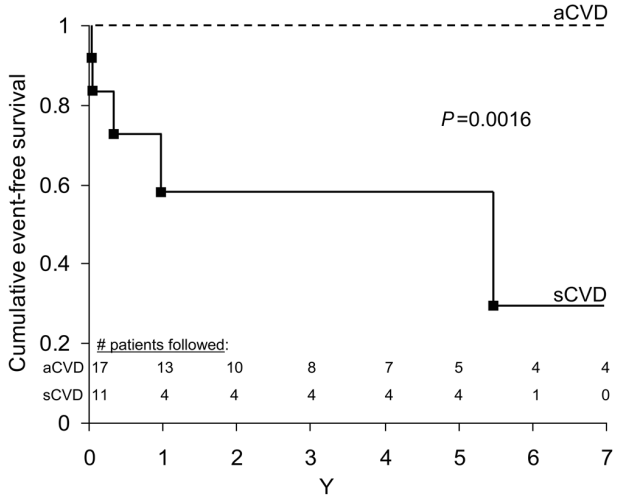

FIGURE 1. *Graph of Kaplan-Meier analysis demonstrating significantly higher cumulative event-free survival in patients with asymptomatic cortical venous drainage (aCVD) versus symptomatic cortical venous drainage (sCVD) (P = 0.0016, log-rank test). One patient with aCVD experienced a nonfatal intracranial hemorrhage 13 years after diagnosis (not shown).*

global aphasia, which spontaneously resolved by the time of discharge. In another patient, there was nontarget embolization of 2 pushable coils into pulmonary arteries; these were retrieved without clinical significance. A cerebrospinal fluid leak complicated 1 of the 6 surgical treatments. There were no complications of the 2 radiosurgical treatments.

Figure 2 presents the angiograms of a representative patient in the symptomatic group (Patient 21), and Figure 3 presents the angiograms of a patient in the asymptomatic group (Patient 2).

DISCUSSION

In managing patients with cranial dAVFs, risks of treatment must be weighed against the lesion's natural history (9, 33). Unfortunately, dAVFs may have a highly variable course, ranging from spontaneous obliteration to an apoplectic event (14, 41, 43). ICH may result from increased pressure on delicate cortical veins (9, 35). NHNDs such as progressive dementia (25), seizures (41), venous infarction (36), and reversible focal deficits (22, 28, 30, 31) are also likely the result of cortical venous hypertension (25, 30). An ideal treatment paradigm would involve differential management of dAVFs on the basis of known risk factors for ICH or NHND.

Numerous studies have documented CVD in patients with ICH or NHND (1, 3, 4, 8, 11, 13, 14, 24, 32, 41). It is important to note that nearly all of these studies reported a strong association between CVD and presenting symptoms, but not future events. In 1972, Houser et al. (24) observed CVD in 4 patients presenting with ICH. Djindjian et al. (13) later graded dAVFs according to flow characteristics, including CVD. Malik et al. (32) combined 10 personal cases with 203 dAVFs from the literature and noted CVD in all 33 lesions presenting with ICH. The authors also observed that hemorrhagic lesions were frequently located outside the transverse sigmoid and cavernous sinuses, but dAVFs at these locations were later shown to simply have a greater propensity for CVD (10, 18, 26). In a meta-analysis of 377 dAVFs, Awad et al. (1) found a significant association between ICH or NHND

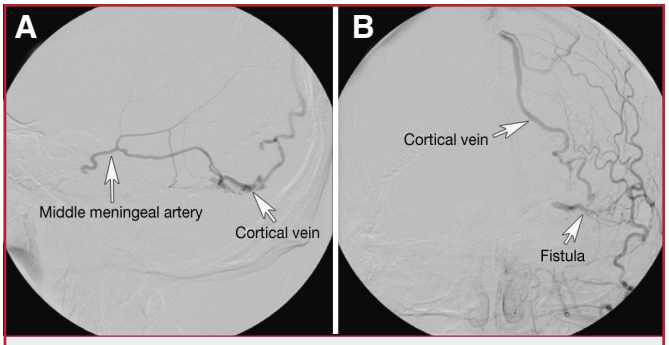

FIGURE 2. *Angiograms of a symptomatic 63-year-old man (Patient 21) who presented with a left parietal hemorrhage. Angiography demonstrated a left parietal dural arteriovenous fistula fed primarily by the left middle meningeal artery.* **A**, *microcatheter injection, lateral projection, in the middle meningeal artery demonstrating a direct dural-to-pial fistula.* **B**, *anteroposterior projection after injection of the left external carotid artery. The cortical venous drainage was eliminated 9.7 months after angiographic diagnosis using transarterial injection of ethylene vinyl copolymer (Onyx; Micro Therapeutics, Inc., Irvine, CA).*

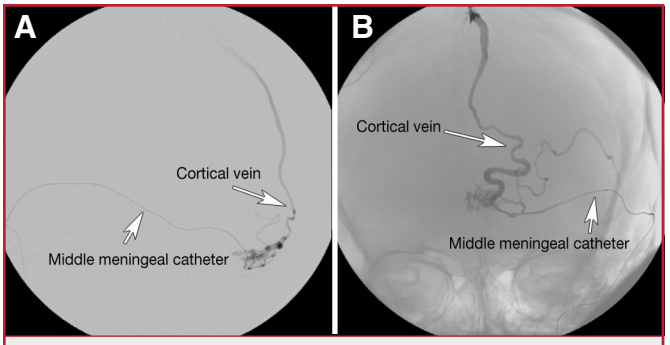

FIGURE 3. *Angiograms of an asymptomatic 41-year-old woman (Patient 2) with an incidental direct dural-to-pial arteriovenous fistula.* **A,** *lateral projection after injection of a microcatheter in the distal middle meningeal artery, demonstrating immediate flow into a cortical vein.* **B**, *unsubtracted anteroposterior projection of the same injection. This was successfully treated with ethylene vinyl copolymer (Onyx) injection 5.3 months after diagnosis.*

at presentation and the presence of CVD, galenic drainage, or venous aneurysms. In 1995, Cognard et al. (7) elaborated on the Djindjian et al. classification scheme, associating 5 angiographic grades with increasing frequency of neurological events. The same year, Borden et al. (3) devised a simplified grading system that was later validated to predict ICH or NHND at presentation (10). These studies in the aggregate clearly established an association between CVD and an aggressive presentation but did not address the consequences of persistent CVD in a known dAVF. Stated differently, the clinical course of Borden Type 2 and 3 dAVFs, once identified, remained unknown.

Later series followed patients after their diagnosis to determine this clinical course. Brown et al. (4) calculated a 1.8% annual risk of ICH in 52 conservatively treated patients having dAVFs that presented predominantly with symptoms such as pulsatile tinnitus, headache, or orbital phenomena. Because only 13 patients in this series had documented CVD and because the clinical course of these patients was not separately reported, the hemorrhagic and neurological risk after initial diagnosis for Type 2 and 3 dAVFs could not be determined. Duffau et al. (14) followed 20 patients with angiographically proven Type 2 and 3 dAVFs who presented with ICH and observed a 35% rate of rebleeding over the 20-day mean interval between diagnosis and treatment. This report provided strong evidence that these lesions carry a high subsequent risk, at least for those presenting with cerebral hemorrhage. Later, expanding on a cohort published by Davies et al. (12), van Dijk et al. (41) followed 20 partially treated or untreated dAVFs over a mean period of 4.3 years. Most of these patients presented with symptoms referable to cerebral venous hypertension including ICH (5 patients) and NHNDs (11 patients). Excluding events at presentation, the annual risks of ICH and NHNDs were 8.1 and 6.9%, respectively. Given their results, Duffau et al. (14) and van Dijk et al. (41) concluded that immediate treatment is necessary for all Type 2 and 3 dAVFs. It should be emphasized, however, that nearly all patients in these 2 series presented with ICH or NHND (100% in the series of Duffau et al. [14] and 80% in the series of van Dijk et al. [41]). Yet, many Type 2 and 3 lesions, including 17 of the 28 dAVFs in our series, are discovered incidentally or present with symptoms of increased dural sinus drainage such as pulsatile tinnitus or orbital phenomena. Given the relative paucity of data regarding the risk associated with Type 2 and 3 dAVFs as a whole and the lack of data regarding the risk associated with Type 2 and 3 lesions that are discovered incidentally or after symptoms of increased dural sinus drainage, we sought to separately examine and then compare the clinical course of Type 2 and 3 dAVFs with sCVD or aCVD.

Excluding events at presentation, patients with sCVD in our series had an overall frequency of ICH or NHND of 45.5% and an annual risk of ICH or NHND of 19.0%. These results strongly confirm the very poor clinical course documented by Duffau et al. (14) (whose cohort included 100% sCVD patients as categorized by our schema) and van Dijk et al. (41) (whose cohort included 80% sCVD patients as categorized by our schema). In fact, the 19.0% annual risk in our sCVD patients closely matched the 15.0% annual risk reported by van Dijk et al. (41). This very poor clinical course for sCVD patients is in contrast with that for aCVD patients in our study. We documented an overall frequency of ICH or NHND of 5.9% and an annual risk of ICH or NHND of 1.4% for aCVD patients. This marked difference in risk between groups was strongly statistically significant.

These data raise interesting questions regarding the variable physiological effects of angiographically identified CVD associated with dAVFs. The less aggressive clinical course of dAVFs with aCVD suggests that CVD may not be a reliable indicator of cortical venous hypertension. Although they share venous drainage with the cortical circulation, lesions with aCVD may lack or have less severe cortical venous hypertension. This may be attributable to less flow through the fistula, more efficient collateral pathways to uninvolved dural sinuses, or other factors at a microvascular level. Direct measurements of pressure in the

draining pedicles of a dAVF obtained at the time of surgical or endovascular treatment may provide more information about possible physiological differences between patients with symptomatic and asymptomatic CVD. Physiological imaging tools may also have a role, but the CVD would likely cause significant errors in the measurements of mean transit time and blood volume.

Our study has several limitations. First, our series was a retrospective analysis and, therefore, subject to the inherent biases of this study design. Second, many patients in our series underwent partial treatment, which may have influenced their clinical course. However, there is precedence for inclusion of partially treated patients in studies examining the clinical course of dAVFs: 6 of 20 patients from the van Dijk et al. (41) series had undergone incomplete treatment and were included in their annual risk calculations. Moreover, the rates of partial treatment in our aCVD group (29%), our sCVD group (27%), and the van Dijk et al. series (30%) were all similar, making a confounding effect on comparisons between these groups unlikely. Finally, and most importantly, when we redefined clinical follow-up for our partially treated patients as the interval between angiographic diagnosis and first treatment (rather than last clinical follow-up), the aCVD group continued to have significantly higher cumulative event-free survival compared with the sCVD group ($P = 0.0019$, log-rank test). A third limitation of our study is that 5 of the 6 excluded patients had sCVD, making the study prone to selection bias. However, it is unlikely that their inclusion (if left untreated) would have reduced the observed differences between groups. In fact, inclusion of these patients (based on the documented high risk of Type 2 and 3 dAVFs in studies that exclusively [14] or predominantly [41] examined patients having sCVD by our categorization schema) would have, if anything, widened the disparity between groups. Fourth, although all patients were offered treatment, it is possible, owing to the uncontrolled nature of this study, that other unrecognized factors could have affected treatment decisions in some way. Fifth, 1 patient with aCVD was lost to follow-up; however, even if this patient experienced an ICH or NHND related to the fistula, this would not have significantly changed the results of the study or our conclusions. Last, our series was relatively small, and the length of follow-up was short for many patients, making our conclusions regarding risk of new neurological events limited to the first few years after presentation. Despite this, we observed a significant difference in outcome between patients with aCVD and those with sCVD.

Modern treatment options for dAVFs include the following: 1) transarterial or transvenous endovascular delivery of embolic agents (typically, liquid acrylic *n*-butyl 2-cyanoacrylate or ethylene vinyl copolymer [Onyx; Micro Therapeutics, Inc., Irvine, CA]) for selective CVD disconnection or dAVF obliteration; 2) microsurgery for selective CVD disconnection or dAVF obliteration; and 3) stereotactic radiosurgery for dAVF obliteration. The strategy of choice for many centers has become selective CVD disconnection because this approach has proven as effective as complete dAVF obliteration, but with significantly lower periprocedural risk (15, 17, 23, 34, 39, 40, 42). Often, this is first attempted via an endovascular approach, with surgical intervention being reserved for patients with persistent CVD that is not amenable to further endovascular therapy. However, certain dAVFs (e.g., anterior fossa lesions having ethmoidal feeders originating from the ophthalmic artery) are best managed with immediate surgery because of the relatively high risks associated with endovascular treatment of these specific lesions. Finally, in a minority of patients having significant medical comorbidities or dAVFs that are poorly amenable to endovascular and surgical therapy, stereotactic radiosurgery is a viable alternative with a proven record of efficacy for some dAVFs (5, 19, 38).

It is the risks of these therapeutic modalities that must be compared with the expected clinical course of a given dAVF. For those lesions with sCVD, the very high risk of new ICH or NHND (19.0% per year in our series) (14, 41) will almost always mandate early and definitive intervention with endovascular or surgical therapy. The therapeutic delay of stereotactic radiosurgery is unacceptable for these lesions unless surgical or endovascular therapy is contraindicated for other reasons (5, 19, 38). For those lesions with aCVD, the apparent less aggressive clinical course (1.4% annual event rate in our series) must be carefully weighed against the risks of therapy. For most aCVD patients, endovascular or surgical intervention will remain the mainstay of dAVF management. However, for some aCVD patients, particularly those who are elderly or medically infirm or who have complicated lesions expected to carry high risk of endovascular and surgical complications, the delayed therapeutic effect of stereotactic radiosurgery may be acceptable. Even conservative management may be reasonable for a minority of patients. Follow-up for these individuals might involve serial angiograms to evaluate for changes associated with aggressive behavior, such as the development of new fistula sites, increased arterial flow, venous engorgement, venous aneurysms, or dural sinus stenosis (7, 20, 29, 44).

CONCLUSION

In this series of 28 patients with dAVFs and persistent CVD, the risk of future ICH or NHND was significantly lower among patients presenting without ICH or NHND (aCVD) compared with patients presenting with these symptoms (sCVD). The respective annual event rates were 1.4% for patients with aCVD versus 19.0% for patients with sCVD. Although immediate treatment is mandated for Type 2 or 3 dAVFs causing ICH or NHND, a more elective approach can be considered for patients presenting without ICH or NHND.

Disclosure

This study was financially supported with the following funds granted to Gregory J. Zipfel, M.D.: unrestricted educational grants from Synthes CMF (West Chester, PA) and Boston Scientific (Natick, MA), and a Barnes-Jewish Hospital Foundation (St. Louis, MO) grant. The remaining authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

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COMMENTS

Strom et al. retrospectively reviewed the charts of patients with dural arteriovenous fistulae (dAVF), specifically those with cortical venous drainage (CVD). They found 34 patients; 6 of these patients were eliminated because of treatment within 1 week (5 patients) or no follow-up (1 patient). Of the 28 total remaining patients, 17 presented with asymptomatic CVD. The mean time between angiographic diagnosis and first therapeutic procedure was 1.7 years, and the mean follow-up time was

4.1 years. Thirteen of the 17 patients accepted treatment (in 8 of the 13 patients, CVD was eliminated, and in 5 of the 13 patients, CVD persisted after treatment). One (5.9%) of the 17 patients experienced an intracranial hemorrhage, which occurred 13 years after the patient declined treatment. Eleven patients had symptomatic CVD; 10 of these 11 patients underwent treatment (mean time between diagnosis and first therapeutic procedure, 0.7 year); in 7 of the 10 patients, the CVD was eliminated with treatment, and in 3 of the 10 patients, CVD persisted after treatment. The mean follow-up time was 2.4 years, during which intracranial hemorrhage occurred in 2 patients (18.2%), and new or worsened nonhemorrhagic neurological deficit occurred in 3 patients (27.3%). The authors conclude that their data do not support the recommendation that all Type 2 and 3 dAVFs require urgent treatment.

The limits to the study are outlined in the Discussion and include its retrospective nature; in addition, many patients underwent partial treatment during the follow-up period (5 of the 6 excluded patients had symptomatic CVD, and 1 patient was lost to follow-up); and the study had a limited follow-up time period with a small number of patients. Because of these limitations, it is impossible to draw a conclusion that patients with dAVF and CVD should not have timely treatment.

From a common sense standpoint, the only difference between an asymptomatic CVD patient and a symptomatic CVD patient is that the latter just had an event. The data presented in this article, the above-noted caveats notwithstanding, merit further clinical investigation. On the basis of this report, it would seem ethically sound to follow asymptomatic patients, ideally in a randomized fashion with minimal exclusions, to confirm or refute these findings.

Daniel Surdell
Christopher Eddleman
H. Hunt Batjer
Chicago, Illinois

Cranial dAVFs with CVD have long been known to have a worrisome course. Certainly, in the past, symptomatic lesions were most likely to have been treated aggressively, but this study certainly indicates (although the numbers are small) that symptomatic CVD has roughly 18 to 19 times the incidence of hemorrhage per annum as compared with the asymptomatic presentation.

Be that as it may, even at an annual event rate of 1.4%, the findings indicate that these lesions should be treated aggressively and effectively to remove the source of the CVD, even if the dAVF is not entirely cured. This information is useful in understanding the disease process and planning timely intervention.

Robert H. Rosenwasser
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Strom et al. present an interesting analysis of the risk of neurological complications, including hemorrhage, in a retrospectively identified cohort of patients with asymptomatic versus symptomatic dAVFs with CVD. There was a significantly greater risk of bleeding and serious complications in symptomatic as compared to asymptomatic cases during follow-up or between diagnosis and treatment.

The study is retrospective and uncontrolled. All patients were offered treatment, and only 4 asymptomatic cases and 1 symptomatic case were, in fact, not treated. So this hardly qualifies as a "natural history" study. The untreated cases were too few to allow a meaningful comparison of factors and risks associated with the symptomatic state. The reasons for selecting nontreatment were uncontrollable and, therefore, could have introduced unknown biases toward treating more dangerous symptomatic and asymptomatic lesions. Hence, we would be careful about false generalization and advising patients with CVD against treatment, unless more robust follow-up data become available on cases who have received a recommendation against treatment. There may be a subgroup of patients with dural fistulae with CVD who remain asymptomatic, but this subgroup is small compared with the group having lesions without CVD, and there is no guarantee that such patients would remain asymptomatic, until we better understand the common features which portend a benign course.

Let us remember the foremost fact that each symptomatic case with CVD was, in fact, asymptomatic until the first symptom or bleed, which is often catastrophic. So symptomatic state alone is not a guarantee of benign clinical course, but a Darwinian selection of cases that have not yet developed serious sequelae.

Issam A. Awad
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This is a thoughtful attempt to stratify the risk of subsequent neurological events for patients diagnosed with cranial dAVFs with CVD. The rarity of dAVFs and factors that affect selection criteria for treatment make understanding of the natural history of untreated patients difficult. In this series, the authors noted a more benign clinical course for patients with either pulsatile tinnitus or orbital symptoms, as compared with patients with dAVFs who presented with bleeding or other nonhemorrhagic neurological symptoms. Also, despite a multimodality treatment approach using endovascular techniques, surgical disconnection, and radiosurgery, only 15 (65%) of 23 patients had complete elimination of their dAVF. Although the information presented should not affect the management of dAVF patients who are considered at high risk in the present analysis, it does prompt discussion about the need for aggressive treatment for all dAVF patients with CVD. I hope that continued advances in endovascular techniques will provide higher cure rates in the future.

Bruce E. Pollock
Rochester, Minnesota

In this well-written article, Strom et al. elucidate an important subgroup of dAVF patients that have a better prognosis than what has previously been suggested. And, vice versa, there are patients with previous hemorrhages or nonhemorrhagic neurological deficits that may need more aggressive treatment from the start. As there still are complications related to treatment, it is important to separate these groups of patients. In the group with a better prognosis, we have more time to carefully weigh the optimal means and timetable of treatment to prevent inadvertent sequelae. With noninvasive and continually developing magnetic resonance angiographic techniques, we may, in the future, better identify patients with CVD without the necessity of invasive and risky digital subtraction angiography.

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