Cranial dural arteriovenous fistulas: modification of angiographic classification scales based on new natural history data

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This article presents a modification to the existing classification scales of intracranial dural arteriovenous fistulas based on newly published research regarding the relationship of clinical symptoms and outcome. The 2 commonly used scales, the Borden-Shucart and Cognard scales, rely entirely on angiographic features for categorization. The most critical anatomical feature is the identification of cortical venous drainage (CVD; Borden-Shucart Types II and III and Cognard Types IIb, IIa + b, III, IV, and V), as this feature identifies lesions at high risk for future hemorrhage or ischemic neurological injury. Yet recent data has emerged indicating that within these high-risk groups, most of the risk for future injury is in the subgroup presenting with intracerebral hemorrhage or nonhemorrhagic neurological deficits. The authors have defined this subgroup as symptomatic CVD. Patients who present incidentally or with symptoms of pulsatile tinnitus or ophthalmological phenomena have a less aggressive clinical course. The authors have defined this subgroup as asymptomatic CVD. Based on recent data the annual rate of intracerebral hemorrhage is 7.4–7.6% for patients with symptomatic CVD compared with 1.4–1.5% for those with asymptomatic CVD. The addition of asymptomatic CVD or symptomatic CVD as modifiers to the Borden-Shucart and Cognard systems improves their accuracy for risk stratification of patients with high-grade dural arteriovenous fistulas. (DOI: 10.3171/2009.2.FOCUS0928)

KEY WORDS • intracranial dural arteriovenous fistula • classification •
natural history • cerebral hemorrhage • neurological deficits

CRANIAL DAVFs are direct shunts between dural arteries and a dural venous sinus or cortical vein. These DAVFs represent 6% of supratentorial and 35% of infratentorial vascular malformations.⁴⁰ Since their original angiographic description by Sachs in 1931,⁴³ numerous studies have been published describing DAVF angiographic features,^{3,10,15,43} mode of presentation,^{1,3,10,13,15,19,28,36,43} and subsequent clinical course.^{5,13,16,45,46,48} From these studies, several important clinical characteristics have been discerned. First, most DAVFs are idiopathic, while a minority are associated with antecedent events such as craniotomy, trauma, or dural sinus thrombosis.^{2,8} Second, DAVFs may present incidentally or with symptoms related to the location and pattern of venous drainage.²⁷ For example, lesions with increased dural sinus drainage often present with symp-

toms related to the involved sinus, such as pulsatile tinnitus for lesions of the transverse or sigmoid sinus, and ophthalmoplegia, proptosis, chemosis, retroorbital pain, or decreased visual acuity for lesions involving the cavernous sinus.^{30,48,49} Those patients with CVD—an angiographic finding in which the DAVF shares venous drainage with the normal cortical circulation—often present with ICH or NHND, including cognitive dysfunction, seizures, or focal neurological deficits.^{22,23,27,32,34,35,39,46,48,49} Third, the natural history of DAVFs depends greatly on the presence or absence of CVD. Although the association between CVD and presentation with symptoms of ICH or NHND has long been appreciated,^{1,3,6,11,13,15,16,25,36,48} only in the past decade has it become clear that CVD also increases the risk of subsequent neurological events.^{16,48} Appropriately, the importance of CVD has been reflected in the angiographic classification systems of Borden and Shucart³ and Cognard et al.¹⁰ (Table 1).

Recently, 2 independent reports have provided new information on the natural history of DAVFs with CVD.^{45,46} Both studies demonstrate a strong relationship between the mode of presentation and natural history.

Abbreviations used in this paper: AVF = arteriovenous fistula; CBF = cerebral blood flow; CVD = cortical venous drainage; DAVF = dural AVF; ICH = intracranial hemorrhage; NHND = nonhemorrhagic neurological deficits.

TABLE 1: Angiographic classification systems for DAVFs

Borden-Shucart Type	Cognard Type	Site of Venous Drainage	Flow Characteristics*
CVD absent			
I	I	dural sinus	antegrade sinus flow
I	Ila	dural sinus	retrograde sinus flow
CVD present			
II	IIb	dural sinus	antegrade sinus flow, CVD
II	Ila + b	dural sinus	retrograde sinus flow, CVD
III	III	CVD	nonectatic cortical vein
III	IV	CVD	ectatic cortical vein
III	V	CVD	spinal perimedullary vein

* Applicable only to the Cognard scale.

Patients who present with symptoms of cortical venous hypertension have a substantially higher risk of new neurological events compared with those who present incidentally or with symptoms of increased dural sinus drainage.^{45,46} The best hypothesis to explain this observation is that the angiographic demonstration of CVD does not necessarily indicate cortical venous hypertension.

The purpose of this article is to provide a systematic review of the clinical and radiological features of DAVFs including patient characteristics, angiographic features, manner of presentation, and neurological risk if not treated. Common therapeutic approaches for DAVFs are also reviewed with an emphasis on appropriate patient selection based on recent natural history data. Finally, the authors propose a modification to the existing classification systems based on the aforementioned insights on neurological risk in patients with or without presenting symptoms of ICH or NHND.

Classification Systems

There are 2 widely used angiographic classification systems for DAVFs: the Borden-Shucart³ and Cognard¹⁰ classifications (Table 1). In the Borden-Shucart system, classifications are based on the site of venous drainage (dural sinus and/or cortical vein) and the presence or absence of CVD. Type I lesions drain directly into a dural sinus without CVD, Type II lesions drain into a dural sinus and have CVD, and Type III lesions drain directly into a cortical vein and therefore have CVD. Lesions are subclassified as having a single fistula (subtype a) or multiple fistulas (subtype b).³

In the Cognard system, an adaptation of the Djindjian system,¹⁵ classifications are based on the direction of dural sinus drainage (antegrade or retrograde), the presence or absence of CVD, and venous outflow architecture (nonectatic cortical vein, ectatic cortical vein, or spinal perimedullary vein).^{10,15} Type I lesions drain antegrade into a dural sinus without CVD, Type IIa lesions drain retrograde into a dural sinus without CVD, Type IIb lesions drain antegrade into a dural sinus with

CVD, and Type IIa + b lesions drain retrograde into a dural sinus with CVD. Type III, IV, and V lesions all have CVD, absent dural venous drainage, and varying cortical venous outflow architecture. Both the Borden-Shucart and Cognard systems highlight the importance of CVD. Because many researchers have documented the association of CVD with clinical symptoms,^{3,10,12,14} the higher the type in either classification system the more likely the DAVF is to be symptomatic as a result of inferred venous congestion.^{3,10}

Imaging

Catheter angiography remains the most accurate method for the detection and classification of a DAVF (Figs. 1–3).^{4,26} Recent advances in CT angiography and MR angiography have allowed improved lesion detection with these modalities.³⁷ These tools, particularly CT angiography, may aid in surgical planning by locating draining veins relative to brain structures. On MR imaging, flow voids from draining veins and T2 hyperintensity from prior ischemia caused by venous hypertension are often identified.²

Presentation

Dural AVFs present at a mean patient age of 50–60 years, yet a wide age range at the time of diagnosis is well documented.^{6,10,13,16,50} No sex preference exists overall, although multiple patient series suggest that a hemorrhagic presentation is more common in males than in females.^{10,16,49} No linkages to family history or genetics have been identified. Antecedent symptoms such as headaches, orbital bruits, pulsatile tinnitus, and ophthalmological phenomena have been noted, but the time course between these symptoms and aggressive presentation with ICH or NHND is poorly defined.^{6,10}

Although uncommon in the past, an increasing number of DAVFs are being diagnosed incidentally, most likely due to the wide availability and frequent use of high-resolution MR imaging. Many other DAVFs, however, are diagnosed after the development of symptoms directly related to the DAVF. These symptoms are highly dependent on the characteristics and location of venous outflow,⁴ and result from either increased dural sinus drainage or the development of cortical venous hypertension. Regarding dural sinus drainage, pulsatile tinnitus is the most frequent complaint. Although this symptom may occur in the context of any DAVF, it is particularly common in lesions with venous outflow into the sigmoid sinus, most likely due to the close proximity of this sinus to the auditory apparatus.⁶ Increased dural sinus drainage may also cause ophthalmoplegia, proptosis, chemosis, retroorbital pain, or decreased visual acuity, all of which result from drainage into the cavernous sinus.^{30,48} On the other hand, symptoms due to cortical venous hypertension are generally more severe and include ICH and NHND. Intracranial hemorrhage is believed to result from the rupture of fragile parenchymal veins subjected to increased pressure due to arterialized CVD, whereas NHND stems from the downstream consequences of arterialized CVD including venous congestion and ultimately cerebral ischemia. Two PET studies of cerebral hemodynamics in patients with

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FIG. 1. Borden-Shucart Type I DAVF in a 64-year-old woman presenting with a 3-month history of pulsatile tinnitus. Angiography demonstrated a Borden-Shucart Type I/Cognard Type IIa dural fistula involving the left transverse sigmoid sinus. Arterial supply was from the left middle meningeal, occipital, ascending pharyngeal, and dural branches of the meningohypophyseal trunk. There was no CVD. *Upper:* A digital subtraction (DS) angiogram obtained in the lateral projection after microcatheter injection of the left occipital artery (OA) demonstrates branching of the OA (arrow) into innumerable transosseous arteries that then fill the left transverse and sigmoid sinuses. A focal stenosis at the junction of the sigmoid sinus with the jugular bulb (arrowhead) is present. Some retrograde flow into the superior petrosal sinus is also evident (asterisk). *Lower:* An anteroposterior projection from the same injection shows filling of the transverse and sigmoid sinuses, as well as retrograde filling of the transverse sinus (asterisk) to the torcula (arrowhead) and the retrograde superior petrosal sinus as well. The arrow indicates the OA. Due to the intolerable pulsatile tinnitus, the patient opted to have her DAVF treated. She underwent transarterial embolization using *N*-butyl cyanoacrylate followed by stereotactic radiation for the residual fistula. Her symptoms resolved.

CVD support this hypothesis. Both studies reported reductions in cerebral blood flow and increased O_2 extraction in brain regions with CVD, which improved after treatment in most patients.^{29,31} Nonhemorrhagic neurological

deficits reported in the context of DAVFs include progressive dementia, seizures, Parkinsonism, cerebellar symptoms, and other focal neurological deficits.^{22,23,27,32,34,35,39} In severe cases, clinical signs or symptoms of increased intracranial pressure may be present.⁹

The mode of presentation is also intrinsically linked to DAVF location.^{1,6,10,34} Anterior fossa lesions are fed by ethmoidal arteries and frequently drain into the cavernous sinus. Therefore, these lesions often present with proptosis, chemosis, or other orbital phenomena due to increased cavernous sinus drainage.^{28,30,49} They also have a high propensity for hemorrhagic presentation.³⁴ A similarly high risk for ICH at presentation has been noted for tentorial DAVFs.³⁴ Middle fossa lesions, on the other hand, more commonly present with pulsatile tinnitus due to frequent drainage into the transverse or sigmoid sinus.⁶ Brainstem DAVFs often present with symptoms such as quadriparesis or lower cranial nerve palsies that result from venous congestion and ischemia within the brainstem.^{3,30} Superior sagittal sinus DAVFs frequently produce global venous congestion that may manifest as hydrocephalus, papilledema, seizures, or dementia.^{23,27,30} A case report documenting alexia and amnesia in a dominant hemisphere, temporal lobe DAVF underscores the importance of functional anatomical relationships in the venous congestive pattern.^{22,34}

In addition, numerous investigators have examined which angiographic features of DAVFs are associated with aggressive presentations, including ICH and NHND. Of the most common features examined, CVD has been the strongest and most consistently identified risk factor for aggressive presentation.^{1,3,6,10,13,15,16,25,36,48} Other angiographic characteristics also potentially linked to clinical presentation include venous varix^{1,6} and galenic drainage.¹

Clinical Course

Dural AVFs Without CVD

Dural AVFs without CVD have a benign natural history, and overwhelmingly present incidentally or with symptoms of increased dural sinus drainage rather than symptoms of cortical venous hypertension.¹³ In a study of 112 patients with Borden-Shucart Type I DAVFs in which 68 patients were conservatively treated and available for clinical follow-up, Satomi and colleagues⁴⁴ found that 1 patient suffered an ICH and none developed NHND over a mean follow-up of 27.9 months. Type I lesions can develop CVD over time and become a higher classification through venous stenosis, venous thrombosis, increased arterial flow, or de novo fistula formation;^{11,44} however, the risk of this conversion appears relatively low. In the aforementioned study by Satomi et al.,⁴⁴ only 2 of 50 patients with follow-up catheter angiography developed CVD.

Dural AVFs With CVD

Dural AVFs with CVD, which commonly present with ICH or NHND,^{1,3,6,10,13,15,16,25,36,48} carry significant risk of future neurological events. Duffau and colleagues¹⁶ were the first to document this poor natural history of dural AVFs with CVD. These investigators studied 20 pa-

TABLE 2: Proposed modification to angiographic classification systems for DAVFs*

Modified Type	Borden-Shucart Type	Cognard Type	Venous Drainage	CVD	Annual Risk (%)		Treatment Recommendation
					ICH	Death	
1	I	I, IIa	dural sinus	no	<1†	0‡	elective treatment for intractable symptoms
2 w/ aCVD	II	IIb, IIa + b	dural sinus	yes	1.4–1.5‡§	0§	elective treatment to prevent ICH/NHND
2 w/ sCVD	II	IIb, IIa + b	dural sinus	yes	7.4–7.6‡§	3.8§	immediate treatment to prevent ICH/NHND
3 w/ aCVD	III	III, IV, V	CVD	yes	1.4–1.5‡§	0§	elective treatment to prevent ICH/NHND
3 w/ sCVD	III	III, IV, V	CVD	yes	7.4–7.6‡§	3.8§	immediate treatment to prevent ICH/NHND

* aCVD = asymptomatic CVD; sCVD = symptomatic CVD.

† Satomi et al., 2002.

‡ Soderman et al., 2008.

§ Strom et al., 2009.

tients with angiographically proven Borden-Shucart Type II or III DAVFs who presented with ICH, and noted a 35% rate of rebleeding over the 20-day mean interval between diagnosis and treatment. The authors strongly advocated “complete and early treatment” of DAVFs with CVD.¹⁶ In a more recent study, expanding on a series published by Davies et al.,¹³ van Dijk and colleagues⁴⁸ studied 20 partially treated or untreated Borden-Shucart Type II or III DAVFs over a mean period of 4.3 years. The majority of these lesions presented with symptoms of cortical venous hypertension (5 patients with ICH, 11 patients with NHND). Annual risks of ICH and NHND were found to be 8.1 and 6.9%, respectively. These authors also concluded that immediate treatment is necessary for all DAVFs with CVD.⁴⁸

Mode of Presentation Affects Natural History of DAVFs With CVD

Two recent studies provide new insights into the natural history of DAVFs.^{45,46} Soderman and colleagues⁴⁵ performed a retrospective analysis of 85 patients with Borden-Shucart Type II or III DAVFs. Thirty-two patients presented with ICH, and 53 patients did not. One of the patients in the latter group presented with progressive dementia. The clinical course between angiographic diagnosis and either the 1st day of therapeutic attempt or the last clinical follow-up (if untreated) was recorded. An annual hemorrhage risk of 7.4% was noted for patients who presented with ICH, as compared with 1.5% for those who did not. This difference in risk was statistically significant. Annual mortality rates were not reported.

Strom and colleagues⁴⁶ retrospectively analyzed 28 patients with partially treated or untreated Borden-Shucart Type II or III DAVFs. Eleven patients presented with symptoms of cortical venous hypertension (ICH or NHND) and were categorized as having symptomatic CVD. Seventeen patients presented incidentally or with symptoms of increased dural sinus drainage (tinnitus or orbital phenomena) and were categorized as having asymptomatic CVD. The clinical course between angiographic diagnosis and either CVD obliteration or last clinical follow-up (if untreated or partially treated) was recorded. An annual hemorrhage risk of 7.6% and an an-

nual risk of NHND of 11.4% was noted for patients presenting with symptomatic CVD, as compared with 1.4% and 0% (respectively) for those presenting with asymptomatic CVD. An annual mortality rate of 3.8% for those with symptomatic CVD and 0% for those with asymptomatic CVD was noted. These differences in neurological risk and death were statistically significant. Results from this study and those of Soderman et al.⁴⁵ clearly suggest that the natural history of patients with DAVFs is dependent not only on the presence or absence of CVD, but also on the mode of presentation, because patients presenting with ICH or NHND (with symptomatic CVD) carry greater risk than those who do not (with asymptomatic CVD).

These data strongly suggest that angiographic evidence of CVD may not be associated with clinically significant cortical venous hypertension in all patients. There is some data from PET studies of hemodynamics to support this hypothesis. Iwama et al.²⁹ studied 10 patients with DAVFs using PET. Five of these patients had angiographic evidence of CVD. Evidence of hemodynamic impairment was identified in the regions with CVD in 4 of these 5 patients with CVD (reduced CBF in 3 patients and reduced CBF with increased O₂ extraction fraction in 1 patient). Interestingly, the 4 patients with hemodynamic impairment all presented with NHND, whereas the 1 patient with CVD and normal hemodynamics presented with tinnitus. None of the 5 patients with Borden-Shucart Type I fistulas demonstrated hemodynamic abnormalities. Similar data were reported by Kuroda and colleagues³¹ in a PET study of 8 patients with DAVFs and CVD. Positron emission tomography measurements obtained before and after definitive treatment demonstrated significant improvements in CBF and O₂ extraction fraction. Improvement in cerebral blood volume was also reported, but the validity of this measurement as a reflection of autoregulatory vasodilation is questionable given the very large quantity of blood in pial veins because of the CVD.

Proposed Modification to the Angiographic Classification Systems

Based on the new natural history data of Soderman

FIG. 2. Borden-Shucart Type II DAVF with asymptomatic CVD in a 52-year-old man with a 15-year history of pulsatile tinnitus. The enlarged vein of Labbe (A, *arrowheads*) had been documented using MR imaging and MR angiography 10 years prior to evaluation at our institution. The left vertebral artery injection in the lateral projection (A) demonstrates a Borden-Shucart Type II/Cognard Type IIb DAVF with arterial supply from muscular branches of the vertebral artery (*arrows*). Other feeding arteries included the posterior meningeal and middle meningeal arteries and the OAs. The drainage occurs via 2 separate channels in the sigmoid sinus (*asterisks*) and an enlarged vein of Labbe (*arrowheads*). Not well-demonstrated on these images is a severe stenosis at the junction of the sigmoid sinus and the jugular bulb, as well as occlusion of the left transverse sinus. The lateral (B) and anteroposterior (C) projections are later images obtained after the same injection demonstrating extensive retrograde CVD (*arrowheads*). The patient underwent transvenous and transarterial embolization with successful obliteration of the fistula.

et al.⁴⁵ and Strom et al.,⁴⁶ we propose a modification to the existing angiographic classification systems (Table 2). The new proposed classification scales are as follows: 1) Type 1 DAVFs drain solely into a dural sinus and have no CVD; 2) Type 2 DAVFs drain into a dural sinus and have either asymptomatic or symptomatic CVD; and 3) Type 3 DAVFs drain directly into cortical veins and have either asymptomatic or symptomatic CVD. A stepwise increase in neurological risk is noted between Type 1 lesions, Type 2 or 3 lesions with asymptomatic CVD, and Type 2 or 3 lesions with symptomatic CVD (Table 2). We believe such differences should be considered when developing a treatment strategy for patients harboring DAVFs (see *Treatment*).

Treatment

Treatment options include transarterial or transvenous endovascular delivery of embolic agents for DAVF obliteration or selective CVD disconnection, microsurgical intervention for DAVF obliteration or selective CVD disconnection, stereotactic radiosurgery for DAVF obliteration, or multimodality therapy. Selective CVD disconnection via surgical or endovascular means has gained popularity in recent years due to documentation of a comparable efficacy to DAVF obliteration with significantly lower periprocedural risks.^{17,18,24,38,42,47,49} At many institutions, the first therapeutic attempt for most DAVFs is via an endovascular approach, with microsurgery reserved for patients with persistent CVD following maximal endovascular therapy. In select DAVFs in which endovascular therapy carries considerable procedural risk (such as anterior fossa lesions with ethmoidal feeders originating from the ophthalmic artery), surgical intervention is the treatment of choice. Stereotactic radiosurgery is also a proven efficacious therapy for some DAVFs,^{7,20,41} although

this treatment approach includes the caveat that obliteration occurs over time with the risk of new neurological events existing in the interim.

When treating a patient with a DAVF, the risks of intervention must be carefully weighed against the lesion's expected clinical course based on available natural history studies. Traditionally, given the high reported risk of DAVFs with CVD as studied by Duffau et al.¹⁶ and van Dijk et al.,⁴⁸ urgent treatment has been strongly advocated. However, recent results from Soderman and colleagues⁴⁵ and Strom and associates⁴⁶ indicate that further refinement to the treatment algorithm for high-grade DAVFs is warranted. For patients with symptomatic CVD, the well-documented high risk of subsequent ICH or NHND (as high as a 19% risk per year⁴⁶) mandates immediate endovascular or surgical intervention in the vast majority of cases. The therapeutic delay of stereotactic radiosurgery would be unacceptable for all but the most medically frail patients or those with complex lesions that carry very high procedural risks. For patients with asymptomatic CVD, the newly documented lower risk of subsequent ICH or NHND (1.4–1.5% risk per year^{45,46}) indicates that a more judicious approach toward therapeutic intervention is warranted. In most cases, endovascular or surgical intervention is still indicated, although the timing of therapy may be more elective in nature. In others, particularly in patients who are elderly, medically frail, or harbor complex DAVFs, stereotactic radiosurgery may be a reasonable alternative. In a minority of patients with asymptomatic CVD, even conservative treatment may be reasonable. Close clinical monitoring for signs or symptoms of aggressive behavior would be necessary for these patients. Imaging follow-up could include serial angiograms to identify changes associated with aggressive behavior, such as the development of new

FIG. 3. Borden-Shucart Type II DAVF with symptomatic CVD in a 50-year-old woman presenting with headache with a history of previous left internal carotid artery aneurysm clip placement 12 years prior. A head CT (A) demonstrated right-sided subarachnoid hemorrhage (arrow). Catheter angiography found no evidence of new or recurrent aneurysm. A Borden-Shucart Type II/Cognard Type IIa + IIb DAVF of the right transverse sinus was found. Arterial supply was from the right middle meningeal, occipital, and posterior meningeal arteries. The right sigmoid sinus was occluded and drainage was retrograde out the right transverse sinus to the torcula, as well as through extensive retrograde CVD. The right external carotid DS angiography injection, lateral projection (B), shows several branches of the middle meningeal (asterisks) draining into a varix of the transverse sinus (arrow). A guide catheter is also present in the left jugular bulb with a microcatheter placed in the torcula in anticipation of transvenous embolization. Extensive retrograde CVD is observed (arrowheads) in the occipital region with retrograde filling of a large vein of Labbe more anteriorly. The anteroposterior projection from the same injection (C) shows the venous drainage in the transverse sinus (arrow) and cortical veins (arrowheads) to better advantage. This lesion was treated acutely with transvenous coil embolization and transarterial *N*-butyl cyanoacrylate injection. The fistula was eliminated.

fistula sites, increased arterial flow, venous engorgement, venous aneurysms, or dural sinus stenosis.^{11,21,33,50} In addition, MR imaging studies could be used to monitor for the development of signal changes on T2 or FLAIR that might indicate subclinical ischemic changes secondary to venous hypertension. Perfusion imaging using CT or MR could also be considered given the wide availability of these tools.

Conclusions

Cortical venous drainage and aggressive presentation with ICH or NHND are strong predictors of poor natural history for DAVFs. Inclusion of both factors in a classification system allows for more accurate risk stratification when counseling patients. To this end, the following modification to existing angiographic classification scales was proposed: DAVFs with CVD would be subdivided into those with asymptomatic CVD (presenting incidentally or with symptoms of increased dural sinus drainage) or those with symptomatic CVD (presenting with ICH or NHND). When applied to the Borden-Shucart scale, Type 1 lesions have no CVD and carry a low risk for ICH (< 1% risk per year⁴⁴), Type 2 and 3 DAVFs with asymptomatic CVD carry intermediate risk for ICH (1.4–1.5% risk per year^{45,46}), and Type 2 or 3 DAVFs with symptomatic CVD carry a high risk for ICH (7.4–7.6% risk per year^{45,46}). Regarding treatment guidelines for patients with symptomatic CVD, the high risk of subsequent neurological events mandates immediate treatment via endovascular or surgical means for the vast majority of patients. For patients with asymptomatic CVD, on the other hand, the interme-

diate risk of subsequent neurological events suggests that endovascular or surgical therapy is typically required but that this may be performed in a more elective fashion. Moreover, the delayed therapeutic effect of stereotactic radiosurgery may be acceptable in select patients with asymptomatic CVD.

Disclosure

Gregory J. Zipfel, M.D., receives financial support from a Synthes CMF unrestricted educational grant, a Boston Scientific unrestricted educational grant, and a Barnes-Jewish Hospital Foundation grant.

Acknowledgment

The authors gratefully acknowledge the editorial support provided by Abbie Bradley, B.S.N., M.S.W., in the preparation of this manuscript.

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Manuscript submitted January 15, 2009.

Accepted February 11, 2009.

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