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# Benign cranial dural arteriovenous fistulas: outcome of conservative management based on the natural history of the lesion

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*Object.* Cranial dural arteriovenous fistulas (DAVFs) can be classified into benign or aggressive, based on their patterns of venous drainage. A benign condition requires the absence of cortical venous drainage (CVD). The clinical and angiographic features of a consecutive single-center group of 117 patients harboring benign cranial DAVFs were evaluated over time to validate the behavior and appropriate management of these lesions.

*Methods.* At the initial assessment four patients were asymptomatic. Two infants presented with congestive heart failure. All other patients presented with other benign symptoms: chronic headache, bruit, or orbital phenomena. Observational management was instituted in 73 patients (62%). Intolerable bruit or ophthalmological sequelae were deemed indications for palliative embolization in 43 patients and surgical treatment in one patient. A median follow-up period of 27.9 months (range 1 month–17.5 years) was available in 112 patients (95.7%), among whom repeated angiography was performed in 50. Overall, observational and palliative management resulted in a benign and tolerable level of disease in 110 (98.2%) of 112 cases. In two cases managed conservatively CVD developed. In both of these cases the conversion from benign to aggressive DAVF was associated with spontaneous progressive thrombosis of venous outlets.

*Conclusions.* The disease course of a cranial DAVF without CVD is indeed benign, obviating the need for a cure of these lesions. Symptoms are well tolerated with either observation or palliative treatment. After a long-term follow-up review of 68 patients, this conservative management resulted in a benign and tolerable level of disease in 98.5% of cases. It is noteworthy, however, that a benign DAVF carries a 2% risk of developing CVD, mandating close clinical follow-up review in such cases and renewed radiological evaluation in response to any deterioration in the patient's condition.

**KEY WORDS • dural arteriovenous fistula • cortical venous reflux • natural history**

It is generally recognized that the severity of clinical signs and symptoms of cranial DAVFs correlates with their patterns of venous drainage.[1,3,13,18] The absence of CVD can be considered a potential predictor of a benign clinical course.[9,10] Davies, et al.,[8] have reported that the majority of cranial DAVFs that are not accompanied by CVD behave in a benign fashion and that the focus of therapeutic efforts, if necessary at all, should be directed toward palliation rather than toward an angiographically verifiable cure. Cognard and associates,[7] however, have reported that seven patients, six of whom were partially treated, experienced an alteration in venous drainage during the follow-up period. In five cases CVD developed, indicating that a complete curative treatment for lesions that are not associated with CVD is warranted. Unfortunately, the total number of patients in that study is unknown.

*Abbreviations used in this paper:* CVD = cortical venous drainage; DAVF = dural arteriovenous fistula; NHND = nonhemorrhagic neurological deficit.

In the present study, we reviewed both the clinical and angiographic data in 117 patients harboring cranial DAVFs without CVD to determine the behavior of these lesions and to assess their risks of aggressive conversion during a period of conservative management.

## Clinical Material and Methods

Members of the University of Toronto Brain Vascular Malformation Study Group consecutively assessed 285 patients with cranial and spinal DAVFs who presented at Toronto Western Hospital between June 1984 and May 2001. Since 1989, data from these patients have been collected prospectively. One hundred seventeen patients with cranial DAVFs were assigned to a subgroup characterized by benign lesions based on the absence of CVD (Borden Type 1[2]/ Cognard Type I or IIa[6]).

The following clinical data were collected at the initial presentation: patient age and sex, presenting signs and symptoms, and location and previous treatment of the

TABLE 1

*Intracranial locations of the DAVFs in 117 patients*

| Location | No. of Patients (%) |
|---|---|
| cavernous sinus | 50 (42.7) |
| transverse sinus | 44 (37.6) |
| jugular & sigmoid sinuses | 18 (15.4) |
| foramen magnum | 2 (1.7) |
| temporal fossa | 3 (2.6) |

DAVF. All patients underwent a full neurological examination. The diagnosis of a DAVF was made based on findings of a digital subtraction, six-vessel cerebral angiographic study, which was interpreted by a neuroradiologist, with special attention directed to the venous phase. Hemorrhage, NHND, and death were considered aggressive phenomena. Chronic headache, congestive heart failure, bruit, and orbital symptoms were considered benign.

Clinical follow-up studies were conducted in a multidisciplinary clinic, which is attended by both neurosurgeons and interventional neuroradiologists. The patients were advised to undergo repeated angiography after treatment or following worsening of symptoms to exclude the development of CVD.

## Results

Among the 117 patients harboring a benign cranial DAVF, there were 44 male and 73 female patients (51.5 ± 19.6 years of age, mean ± standard deviation). The different locations of the lesions are outlined in Table 1. In the majority of cases (95.7%) the DAVFs were related to a major sinus.

The patients' initial presenting symptoms are summarized in Table 2. One hundred fifteen patients were asymptomatic or presented with chronic headache, bruit, or orbital phenomena. Two infants presented with congestive heart failure due to the high flow through the fistula. No patient was found to have a hemorrhage or an NHND. Follow-up data were available in 112 patients (95.7%) and covered 348.1 patient-years (median 27.9 months, range 1 month–17.5 years). Observational management was chosen for 73 (62.4%) of the 117 patients. Five patients were lost to follow-up review and were therefore excluded from this review. Observation resulted in tolerable, stable disease in 67 (98.5%) of the remaining 68 patients. One patient (1.5%) experienced a seizure during the follow-up period as a result of an intracerebral hematoma after development of CVD.

TABLE 2

*Presenting symptoms in 117 patients*

| Presenting Symptom | No. of Patients (%) |
|---|---|
| asymptomatic | 3 (2.6) |
| hemorrhage | 0 (0.0) |
| NHND | 0 (0.0) |
| chronic headache | 38 (32.5) |
| bruit | 76 (65.0) |
| orbital phenomena | 46 (39.3) |
| congestive heart failure | 2 (1.7) |

TABLE 3

*Complications of diagnostic angiography and palliative treatment*

| Procedure & Complication | Duration | Location of DAVF |
|---|---|---|
| angiography | | |
| partial abducent palsy | transient | cavernous sinus |
| balance difficulties | transient | transverse sinus |
| embolization | | |
| pulmonary edema | transient | transverse sinus |
| raised intraocular pressure | transient | cavernous sinus |
| facial pain | transient | transverse sinus |
| embolus, pericallosal artery | permanent (asymptomatic) | transverse sinus |

Endovascular embolization was performed in 43 (36.8%) of the 117 patients, aimed at palliation of unbearable symptoms or pressing ophthalmological phenomena. In 15 patients, multiple (up to four) treatment sessions were required to obtain a satisfactory result. Surgical therapy was performed in one patient (0.9%). Treatment resulted in a tolerable level of disease in all but one patient. That patient died a few years after undergoing palliative embolizations, most likely as a result of venous congestion.

Long-term angiographic follow-up review was performed in 50 patients because of a sudden or unexpected change in symptoms. A changed venous drainage pattern was revealed in five cases that had been managed conservatively; in two of these cases angiography demonstrated conversion of the benign DAVF into a lesion with CVD. Both cases were treated by disconnection of the CVD.

Complications occurred after two diagnostic angiographic sessions and after four palliative embolizations (Table 3). There was one asymptomatic arterial embolus; the five other complications were transient.

## Discussion

It is generally accepted that the presentation of a DAVF is dictated by its venous drainage pattern. The correlation between the presence of CVD and grave neurological events or death is well known. Venous ectasias of refluxing pial veins, venoocclusive disease in an involved dural sinus, and galenic venous drainage are associated with an aggressive presentation.[1,13,14,18,20] In itself, the anatomical location of a DAVF has no direct correlation with aggressive events, although this was suggested in the past. It seems more appropriate to state that, in some locations, due to the local venous anatomy DAVFs have a higher likelihood of developing CVD with all its consequences.[1,3,18] Based on the venous features of DAVFs, several classifications have been proposed,[2,6,17] each of which is useful in estimating the severity of the disease and in determining an indication for treatment.[9] A factor mostly ignored by these scales is the presence of venous congestion of the brain. This concept was suggested by Gelwan, et al.,[11] in 1988 in their explanation of two cases with papilledema, and was further elaborated by Cognard and associates[5] and by Hurst and colleagues.[15] Willinsky, et al.,[26] highlighted the presence of tortuous, engorged veins on cerebral angiograms in a case of venous congestion and labeled this the pseudophlebitic pattern.

Regarding the course of the disease after presentation of DAVFs without CVD, only two publications have been published that contain clinical and angiographic follow-up data. Davies and coworkers[8] have reported their experience with a cohort of 54 patients who harbored DAVFs without CVD over a mean follow-up period of 33 months. One (2%) of these patients died after receiving palliative endovascular treatment; however, in that case there was no angiographic evidence of conversion into a lesion with CVD. This unusual course of a predicted benign disease was attributed to venous hypertension caused by functional obstruction of the superior sagittal sinus. Cognard and associates[7] reported on their experience with seven patients who initially presented with a DAVF without CVD, but these authors did not mention the number of patients in the entire group. Five patients underwent embolization with particles, one patient proximal ligation of the occipital and middle meningeal artery, and one patient conservative management. A worsening in the venous drainage pattern was observed in all these patients during a follow-up period ranging from 1 month to 20 years (mean 7 years). In two patients who underwent embolization there was a change from antegrade to retrograde flow into the draining sinus and in five cases CVD developed. In all cases, the change in venous pattern was accompanied by a worsening of clinical symptoms. Unfortunately, without knowing the total number of patients in this series, it is impossible to calculate the frequency of progression that is associated with more severe venous drainage.

In the present study, cranial DAVFs that appeared on the initial angiogram without any sign of CVD were categorized as benign. Chronological changes in clinical symptoms and angiographic features in 117 patients harboring a benign cranial DAVF were evaluated. None of the patients presented with either intracranial hemorrhage or NHND and, therefore, the preferred management of these lesions was conservative. Palliative treatment, the goal of which was never a cure, was performed if the patient experienced intolerable symptoms or if there were pressing ophthalmological indications. Using this conservative management, 98% of patients achieved a tolerable level of disease.

Venous thrombosis is likely to be a key factor in the existence and development of cranial DAVFs.[4,13,21,22] Along with additional venous hypertension, venous thrombosis has been shown to contribute to the genesis of de novo cranial DAVFs in experimental studies.[12,24] Histologically, thrombosis was demonstrated in all surgical specimens of involved dural sinuses (nine cases).[25] Regarding the pathophysiological basis of spontaneous resolution of cranial DAVFs, again progressive thrombosis and possible intracranial hemorrhage have been mentioned as causative.[16,19] Among others, Piton, et al.,[23] suggested the theory of staged progression in the disease course of cranial DAVFs. In this concept, venoocclusive changes (thrombosis) in a dural sinus following trauma or surgery promote the growth of the dural arteries and generate the DAVF, followed by progressive restriction in venous drainage and spontaneous closure.

In five cases in our series, angiography demonstrated changes in the venous drainage patterns due to progressive thrombosis of venous outlets. In three patients harboring cavernous sinus DAVFs (one treated with observational management and two with palliative embolization) alterations in the direction of sinus drainage from antegrade to retrograde were identified; these alterations subsequently resolved spontaneously. In two other conservatively managed cases (one patient with a transverse sinus lesion and one with a cavernous sinus lesion), angiography revealed conversion into a lesion with CVD; this conversion was associated with symptom resolution in the first case and symptom aggravation in the latter. This illustrates that benign DAVFs have a 2% potential for angiographically verified conversion, even without treatment. All five cases in which there was angiographically verified conversion were associated with progressive thrombosis in the affected sinus and not with an increase in arterial flow or with de novo arteriovenous shunts, as described by Cognard and associates.[7] Stenosis or thrombosis of the venous outlets, a factor reported to forecast later worsening,[7] was not present on the initial angiogram in the converted cases. Based on this series, however, it is not possible to calculate the exact rate of angiographic conversion, because less than half of the patients underwent repeated angiography. Nevertheless, the majority (98%) of the benign cranial DAVFs followed their predicted benign clinical course.

## Conclusions

The disease course of a cranial DAVF without CVD is indeed benign, obviating the need for a cure of these lesions. Symptoms are well tolerated when either observation or palliative treatment is applied. After long-term follow up, conservative management resulted in a benign and tolerable level of disease in 98% of cases. Nevertheless, a benign DAVF carries a 2% risk for the development of CVD, mandating close clinical follow-up review of such cases and renewed radiological evaluation if there is any worsening of symptoms.

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