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Dural arteriovenous fistulas without cortical venous drainage: presentation, treatment, and outcomes

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OBJECTIVE Current evidence suggests that intracranial dural arteriovenous fistulas (dAVFs) without cortical venous drainage (CVD) have a benign clinical course. However, no large study has evaluated the safety and efficacy of current treatments and their impact over the natural history of dAVFs without CVD.

METHODS The authors conducted an analysis of the retrospectively collected multicenter Consortium for Dural Arteriovenous Fistula Outcomes Research (CONDOR) database. Patient demographics and presenting symptoms, angiographic features of the dAVFs, and treatment outcomes of patients with Borden type I dAVFs were reviewed. Clinical and radiological follow-up information was assessed to determine rates of new intracranial hemorrhage (ICH) or nonhemorrhagic neurological deficit (NHND), worsening of venous hyperdynamic symptoms (VHSs), angiographic recurrence, and progression or spontaneous regression of dAVFs over time.

RESULTS A total of 342 patients/Borden type I dAVFs were identified. The mean patient age was 58.1 ± 15.6 years, and 62% were women. The mean follow-up time was 37.7 ± 54.3 months. Of 230 (67.3%) treated dAVFs, 178 (77%) underwent mainly endovascular embolization, 11 (4.7%) radiosurgery alone, and 4 (1.7%) open surgery as the primary modality. After the first embolization, most dAVFs (47.2%) achieved only partial reduction in early venous filling. Multiple complementary interventions increased complete obliteration rates from 37.9% after first embolization to 46.7% after two or more embolizations, and 55.2% after combined radiosurgery and open surgery. Immediate postprocedural complications occurred in 35 dAVFs (15.2%) and 6 (2.6%) with permanent sequelae. Of 127 completely obliterated dAVFs by any therapeutic modality, 2 (1.6%) showed angiographic recurrence/recanalization at a mean of 34.2 months after treatment.

ABBREVIATIONS CONDOR = Consortium for Dural Arteriovenous Fistula Outcomes Research; CVD = cortical venous drainage; dAVF = dural arteriovenous fistula; ICH = intracranial hemorrhage; NHND = nonhemorrhagic neurological deficit; VHS = venous-hyperdynamic symptoms.

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Progression to Borden-Shucart type II or III was documented in 2.2% of patients and subsequent development of a new dAVF in 1.6%. Partial spontaneous regression was found in 22 (21.4%) of 103 nontreated dAVFs. Multivariate Cox regression analysis demonstrated that older age, NHND, or severe venous-hyperdynamic symptoms at presentation and infratentorial location were associated with worse prognosis. Kaplan-Meier curves showed no significant difference for stable/improved symptoms survival probability in treated versus nontreated dAVFs. However, estimated survival times showed better trends for treated dAVFs compared with nontreated dAVFs (288.1 months vs 151.1 months, log-rank $p = 0.28$). This difference was statistically significant for treated dAVFs with 100% occlusion (394 months, log-rank $p < 0.001$).

CONCLUSIONS Current therapeutic modalities for management of dAVFs without CVD may provide better symptom control when complete angiographic occlusion is achieved.

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KEYWORDS dural arteriovenous fistula; low-grade; cerebral venous drainage; venous hypertension; intracranial hemorrhage; nonhemorrhagic neurological deficit; natural history; treatment; endovascular embolization; radiosurgery; microsurgery; prognosis; vascular disorders

Dural arteriovenous fistulas (dAVFs) are abnormal arteriovenous connections between an arterial feeder and a dural venous sinus or leptomeningeal vein, with the nidus located within the dural leaflets.[1] Dural AVFs may present with hemorrhage, nonhemorrhagic neurological deficits (NHNDs), and venous-hyperdynamic symptoms (VHS) such as headache, tinnitus, ophthalmoplegia, proptosis, and chemosis.[2]

Intracranial hemorrhage (ICH) is associated with significant neurological morbidity and mortality.[3] The best predictor of symptoms and risk of ICH is the venous drainage pattern: dAVFs with cortical venous drainage (CVD) have higher incidence of ICH, venous infarction, and venous hypertension.[4,5] Dural AVFs without CVD are classified as Borden-Shucart type I or Cognard type I or IIa.[6,7] The risk of ICH in these lesions is low (1.5%), and approximately 2% may develop CVD over time.[8,9] Given the "benign" natural history of dAVFs without CVD, routine treatment has been discouraged. However, the generally considered benign symptoms often prompt treatment due to patient discomfort and decreased quality of life.[10]

To date, no studies have assessed the safety or efficacy of current treatments in patients with dAVFs without CVD. We describe the natural history, treatment, and outcomes of dAVFs without CVD in the largest multicenter database of dAVFs: the Consortium for Dural Arteriovenous Fistula Outcomes Research (CONDOR).

Methods

Population

The CONDOR database repository compiles data from 12 different institutions in the United States, the United Kingdom, the Netherlands, and Japan. IRB approval was obtained at each institution. Patients with intracranial dAVFs who presented to any of the participating institutions were identified and retrospectively reviewed. Only patients with radiologically confirmed Borden type I or Cognard grade I/IIa dAVFs diagnosed between 1990 and 2017 were included in this study. The collected data were de-identified, pooled, and transmitted to our institution for analysis. Verification and attestation of data accuracy were performed by each contributing institution.

Data Collection and Definitions

NHND was defined as focal or global neurological deficit due to venous hypertension, including dementia, aphasia/dysarthria, ataxia, sensory and motor deficits, cranial nerve palsies (except for cranial nerves III, IV, and VI), hydrocephalus, seizures, and psychiatric symptoms. VHS were defined as symptoms of increased venous drainage such as isolated headache not due to ICH, tinnitus, bruit, ophthalmoplegia/diplopia, and chemosis/proptosis.

Demographic information, presence of dAVF-related ICH, NHND, and VHS were collected and analyzed. Severity of VHS was considered mild/moderate if nondisabling or severe if it limited the patient's ability to perform activities of daily living.

General angiographic features such as location, size, arterial feeders, sinus/venous drainage, and sinus drainage obstruction were evaluated. Obliteration success and complications were assessed after each therapeutic intervention. Radiological follow-up information was reviewed to determine rates of recurrence or recanalization, progression to CVD, development of new dAVFs, or spontaneous regression. A subgroup analysis of patients treated with liquid embolics (Onyx, Medtronic; *N*-butyl cyanoacrylate, Johnson & Johnson; and PHIL, MicroVention) versus any other endovascular modality was performed.

Statistical Analysis

A Cox proportional hazards regression model was used to calculate hazard ratios. Evaluated censoring events included death due to dAVF and development or worsening of NHND/VHS over time. "Initial time" was the date of initial clinical presentation (for symptomatic cases) or initial imaging diagnosis (for incidental cases). Kaplan-Meier curves for stable/improved symptoms survival probability were plotted and compared between treated and nontreated dAVFs. We excluded from the survival analysis carotid-cavernous fistulas because of their overall good prognosis and ease of endovascular sinus embolization, and type I dAVFs with ICH at presentation (extremely rare). For those rapidly evolving dAVFs that developed new ICH, NHND, or VHS soon after diagnosis and before intervention, initial time was censored to treatment date. A subgroup survival analysis adjusted for degree of obliteration after treatment was also performed.

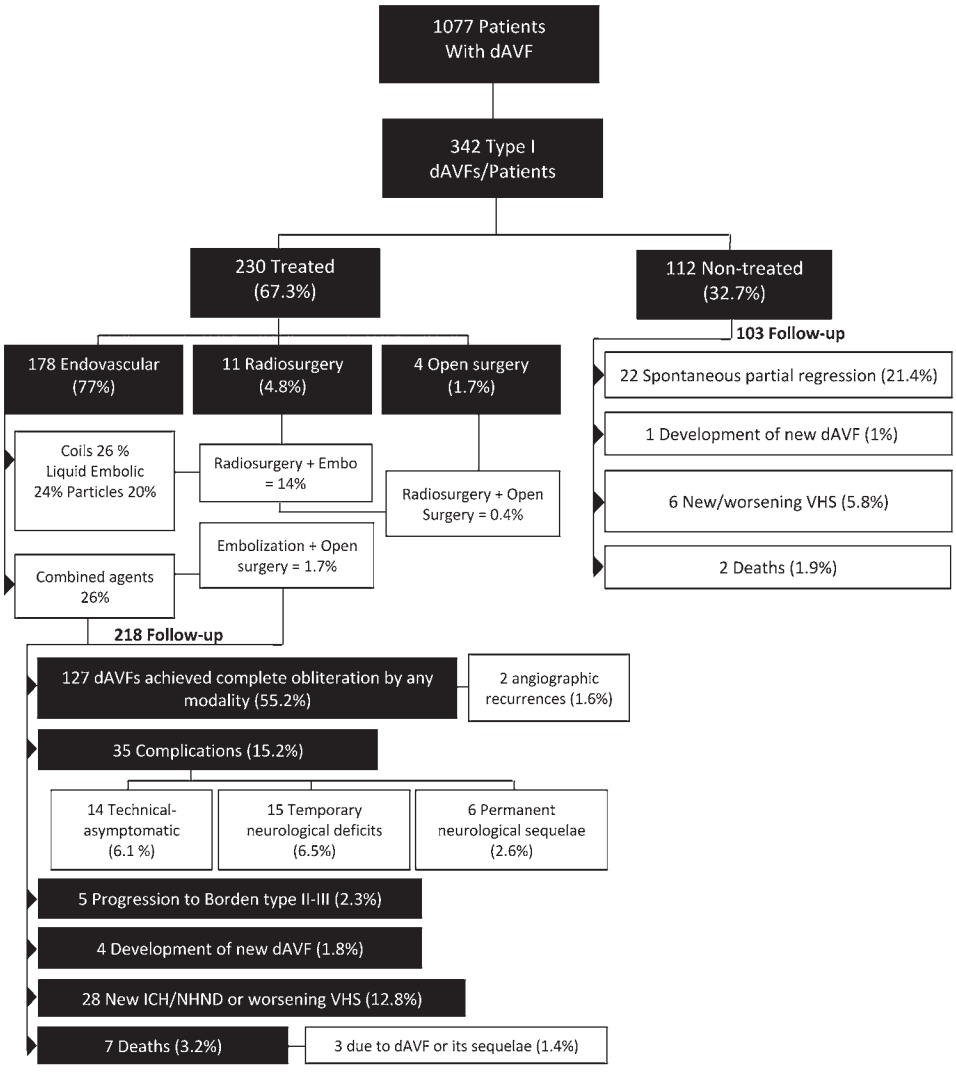

FIG. 1. Flowchart of patient selection and follow-up outcomes.

All analyses were performed using the IBM SPSS (version 24, IBM Corp.).

Results

A total of 342 patients/type I dAVFs without CVD were identified in the CONDOR database (Fig. 1 and Table 1). The mean age was 58.1 ± 15.6 years, and 212 patients (62%) were women. At presentation, 277 patients (81.2%) demonstrated dAVF-related VHS, and 31 patients (9.1%) had severe symptoms: 19 with pulsatile tinnitus or bruit, 11 with chemosis or proptosis, 11 with ophthalmoplegia or diplopia, and 9 with severe headache. Sixteen patients (4.7%) presented with NHND (14 focal neurological deficits and 2 global neurological deficit/seizures). Sixty-three dAVFs (18.4%) were diagnosed incidentally. Most dAVFs were located in the transverse/sigmoid sinus (51.8%), cavernous sinus (21.9%), tentorial/petrosal sinus (6.7%), superior sagittal sinus/convexity (4.7%), foramen magnum (5.8%), torcular (1.8%), ethmoidal or anterior fossa (1.8%), sylvian or middle fossa (0.9%), and other/straight sinus (4.1%).

Two hundred thirty dAVFs (67.3%) were treated (Table 2). The mean time from diagnosis to treatment was 5.9 months. In this time frame, 15 patients developed new VHS, 2 developed new NHND, and 1 patient had a new dAVF-related ICH. Among treatment modalities, 178 dAVFs (77%) underwent mainly endovascular embolization, 11 (4.8%) radiosurgery, and 4 (1.7%) open surgery. Several dAVFs were treated with a combined approach: 32 (14%) with radiosurgery and embolization, 4 (1.7%) with embolization and surgery, and 1 (0.4%) with radiosurgery with open surgery. The most common approach for endovascular intervention was transarterial (130/214, 60.7%); coils were used for embolization in 26.2% of patients, liquid embolics in 24.3%, and particles in 20%. Combined embolic materials were used in 27% (57/214) of patients with endovascular embolization. The most common com-

TABLE 1. Characteristics of 342 patients with dAVFs without CVD in CONDOR

Variable	Value
Demographics	
Mean age, yrs	58.1 ± 15.6
Women	212 (62)
White	235 (68.7)
Past medical history	
Past/current smoker*	71 (20.8)
Hypertension*	127 (37.1)
Diabetes mellitus*	26 (7.6)
History of cancer*	47 (13.7)
Hypercoagulability*	24 (7.0)
Previous ischemic stroke/TIA*	17 (5.0)
Pregnant/<6 wks postpartum*	11 (3.2)
Cranial surgery in last 6 mos/severe head trauma*	54 (15.8)
Symptoms at presentation	
Incidental	63 (18.4)
Symptomatic	279 (81.6)
mRS score at presentation*	
Mean	0.82 ± 0.93
0–2	321 (93.9)
3–5	16 (4.7)
NHND at presentation	16 (4.7)
Global neurological deficit†	2 (0.6)
Focal neurological deficit‡	14 (4.1)
VHS at presentation§	277 (81.2)
Mild/moderate VHS	246 (72)
Severe VHS	31 (9.1)
Etiology*	
Spontaneous/idiopathic	265 (77.5)
Traumatic	33 (9.6)
Iatrogenic/previous intracranial surgery	6 (1.8)
Infectious/thrombophilia	12 (3.5)
Pregnancy	3 (0.9)
Congenital/other	8 (2.3)

mRS = modified Rankin Scale; TIA = transient ischemic attack.
Values represent the number of patients (%) or mean ± SD.
* Data were not available for all patients. Missing values: past/current smoker = 67, hypertension = 5, diabetes mellitus = 5, history of cancer = 5, hypercoagulability = 8, previous ischemic stroke/TIA = 4, pregnant/<6 weeks postpartum = 4, cranial surgery in last 6 months/severe head trauma = 5, mRS at presentation = 5, etiology = 15.
† Includes seizures, dementia, psychiatric symptoms, altered consciousness, hydrocephalus, and symptoms of increased intracranial pressure.
‡ Includes cranial nerve palsies (except ophthalmoplegia), weakness, numbness, aphasia, cerebellar signs (tremor, dysmetria, ataxia), and parkinsonism.
§ Includes symptoms of increased venous drainage (e.g., isolated headache not due to hemorrhage, tinnitus or bruit, orbital phenomena: ophthalmoplegia, decreased vision, nausea/vomiting, chemosis, proptosis, diplopia).

bined endovascular treatment modality was liquid embolics + coils in 49% (28/57).

Partial reduction in early venous filling was achieved in 101 (47.2%) of 214 endovascular cases after the first treatment session. Fifty-three (24.8%) dAVFs underwent more than one embolization, increasing the complete obliteration rate from 37.9% to 46.7% by embolization only. Complete dAVF obliteration was achieved in 21 (47.7%) of 44 patients after radiosurgery and 5 (55.6%) of 9 patients undergoing open surgery. One hundred twenty-seven of 230 treated dAVFs were successfully obliterated by any modality (55.2%). A subgroup analysis of endovascular cases treated with liquid embolics (92 patients, 42%) versus treatment by any other endovascular modality (132 patients, 58.9%) demonstrated similar angiographic obliteration rates: 43.4% vs 56.6% ($p = 0.75$).

Treatment-related complications were reported in 15.2% (35/230) of patients: 14 were technical/asymptomatic complications, 15 experienced temporary neurological sequelae after the intervention, and only 6 (2.6%) had permanent deficits (present at last follow-up).

Follow-up was available for 321 patients (93.9%) during a mean time of 37.7 months (Table 2). The most common follow-up imaging modality was conventional angiography (61.7%). Among 218 treated patients with follow-up, 45.4% (99/218) experienced complete improvement/resolution of symptoms, 18.3% (40/218) had some improvement (not to baseline), 23.4% (51/218) remained symptomatically stable, and 12.8% (28/218) experienced new/worsening NHND/VHS. Two (1.6%) of 127 successfully obliterated dAVFs showed recurrence during follow-up angiography in a mean time of 34.2 months after treatment; progression to Borden type II or III was documented in 2.2% of dAVFs and subsequent development of new dAVF in 1.8% of treated fistulas (1.6% of total). Spontaneous partial regression was documented in 22 (21.4%) of 103 nontreated dAVFs with follow-up available. Nine patients died in this cohort; however, only 3 deaths (0.9%) were related to the presence of a dAVF. The dAVF of one patient who experienced bleeding and died was located in the tumoral bed of a frontal oligodendroglioma. The other two patients who died had dAVFs located in the posterior fossa/cerebellar regions, with arterial feeders that were not successfully embolized after multiple attempts.

Results from the univariate and multivariate Cox proportional hazards regression models are presented in Table 3. Older age, presence of NHND/severe VHS at diagnosis, and infratentorial location were significantly associated with dAVF-related death and worsening of symptoms over time in multivariate analysis. Treatment showed a nonsignificant protective effect in both uni- and multivariate models.

Kaplan-Meier curves demonstrated a nonsignificant trend favoring stable/improved symptoms in treated compared with nontreated type I dAVFs (Fig. 2A). Estimated survival times were higher for treated dAVFs (288.1 months, 95% CI 125.5–450.6) than nontreated dAVFs (151.1 months, 95% CI 120.2–181.9) (log-rank $p = 0.28$). This difference turned statistically significant for treated dAVFs with complete angiographic occlusion (394 months, 95% CI 34.7–753.2; log-rank $p < 0.001$) (Fig. 2B). On the other hand, partially treated lesions demonstrated a trend toward worse symptomatic outcome compared with nontreated type I dAVFs.

Finally, a subgroup survival analysis demonstrated that

TABLE 2. Treatment and follow-up of dAVFs without CVD

Variable	Value
Treatment	230/342 (67.3)
Mean time from diagnosis to treatment, mos	5.9 ± 20.1
Symptoms from diagnosis to treatment*	
New symptoms	18/230 (7.8)
New dAVF-related hemorrhage	1/230 (0.4)
New dAVF-related NHND	2/230 (0.9)
New dAVF-related VHS	15/230 (6.5)
Stable symptoms	173/230 (75.2)
Improving symptoms	7/230 (3.0)
Never had symptoms (n = 6)/symptoms not dAVF-related (n = 21)	27/230 (11.7)
Endovascular embolization	178/230 (77)
Radiosurgery	11/230 (4.8)
Open surgery	4/230 (1.7)
Embolization + radiosurgery	32/230 (14)
Embolization + surgery	4/230 (1.7)
Radiosurgery + surgery	1/230 (0.4)
Complete dAVF obliteration by any modality*	127/230 (55.2)
Procedural complications*	35/230 (15.2)
Technical (asymptomatic, no neurological deficit)	14/230 (6.1)
Vessel dissection/perforation	5/230 (2.2)
Ischemic stroke/venous infarct	5/230 (2.2)
Hemorrhagic stroke	3/230 (1.3)
Microcatheter retention/entrapment	1/230 (0.4)
Temporary neurological deficit	15/230 (6.5)
CN neuropathy	5/230 (2.2)
Motor/sensory deficit	10/230 (4.3)
Permanent neurological sequelae	6/230 (2.6)
CN neuropathy	3/230 (1.3)
Motor/sensory deficit	3/230 (1.3)
Follow-up	321/342 (93.9)
Mean time from diagnosis to last follow-up, mos	37.7 ± 54.4
Clinical assessment	
Symptoms from diagnosis to last clinical follow-up	
New/worsening NHND/VHS	34/321 (10.6)
Treated	28/218 (12.8)
Not treated	6/103 (5.8)
Stable symptoms	96/321 (29.9)
Treated	51/218 (23.4)
Not treated	45/103 (43.7)
Improvement but not to baseline	52/321 (16.2)
Treated	40/218 (18.3)
Not treated	12/103 (11.7)
Complete improvement/resolution of symptoms	139/321 (43.3)
Treated	99/218 (45.4)
Not treated	40/103 (38.8)

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TABLE 2. Treatment and follow-up of dAVFs without CVD

Variable	Value
Follow-up (*continued*)	321/342 (93.9)
Clinical assessment (*continued*)	
mRS score at last follow-up*	
Mean	0.73 ± 0.99
Treated	0.76 ± 1.04
Not treated	0.68 ± 0.89
0–2	300/321 (93.5)
Treated	204/218 (93.6)
Not treated	96/103 (93.2)
3–6	17/321 (5.3)
Treated	11/218 (5)
Not treated	6/103 (5.8)
Radiological assessment	
Angiographic recurrence (recanalization of previously cured dAVF)*	2/127 (1.6)
Mean time from dAVF cure to recurrence, mos	34.2 ± 41.2
Progression to Borden type II or III	5/321 (1.6)
Treated	5/218 (2.3)
Not treated	0/103 (0)
New dAVF	5/321 (16)
Treated	4/218 (1.8)
Not treated	1/103 (1)
Spontaneous partial regression (in nontreated patients w/ follow-up)†	22/103 (21.4)
Death*	9/321 (2.8)
Treated	7/218
Not treated	2/103
Due to dAVF or its sequelae	3/321 (0.9)
Treated	3/218 (1.4)
Not treated	0/103 (0)
Due to another cause	5/321 (1.6)
Treated	3/218 (1.4)
Not treated	2/103 (2)
Unknown cause	1/321 (0.3)
Treated	1/218 (0.5)
Not treated	0/103 (0)

CN = cranial nerve.
Values represent the number of patients (%) or mean ± SD.
* Data not available for all subjects. Missing values: symptoms from diagnosis to treatment = 5, complete dAVF obliteration by any modality = 5, procedural complications = 3, mRS at last follow-up = 4, imaging modality during last follow-up = 4, angiographic recurrence = 23, death = 2.
† Refers to any angiographic hemodynamic improvement (not resolution) of the dAVF.

rapidly evolving dAVFs developed new ICH, NHND, or VHS soon after diagnosis and before intervention. The overall survival probability of this group was similar to the group with nontreated dAVFs (estimated survival time = 317.5 months, 95% CI 58.9–576.2; log-rank p = 0.55) (Fig. 2C).

TABLE 3. Evaluation of risk factors for dAVF-related death and new or worsening NHND/VHS

Factor	Univariate Analysis		Multivariate Analysis	
	HR (95% CI)	p Value	HR (95% CI)	p Value
Older age	1.02 (0.99–1.05)	0.11	1.05 (1.003–1.09)	**0.03**
Sex (female)	1.35 (0.66–2.8)	0.41	1.47 (0.49–4.40)	0.48
NHND/severe VHS at diagnosis	4.38 (2.15–8.94)	**<0.001**	14.2 (3.78–53.26)	**<0.001**
Location (infratentorial)	1.02 (0.47–2.19)	0.96	7.2 (1.53–33.78)	**0.01**
Arterial feeders (≥2)	1.53 (0.65–3.58)	0.33	2.76 (0.82–9.26)	0.10
Venous ectasia	1.63 (0.62–4.36)	0.32	2.61 (0.53–12.79)	0.24
Occlusion/stenosis of sinus drainage	1.25 (0.46–3.40)	0.66	1.32 (0.41–4.19)	0.64
Flow direction (retrograde)	1.11 (0.49–2.56)	0.79	1.46 (0.46–4.61)	0.52
Treatment	0.61 (0.25–1.49)	0.28	0.69 (0.19–2.58)	0.59

Boldface type indicates statistical significance.

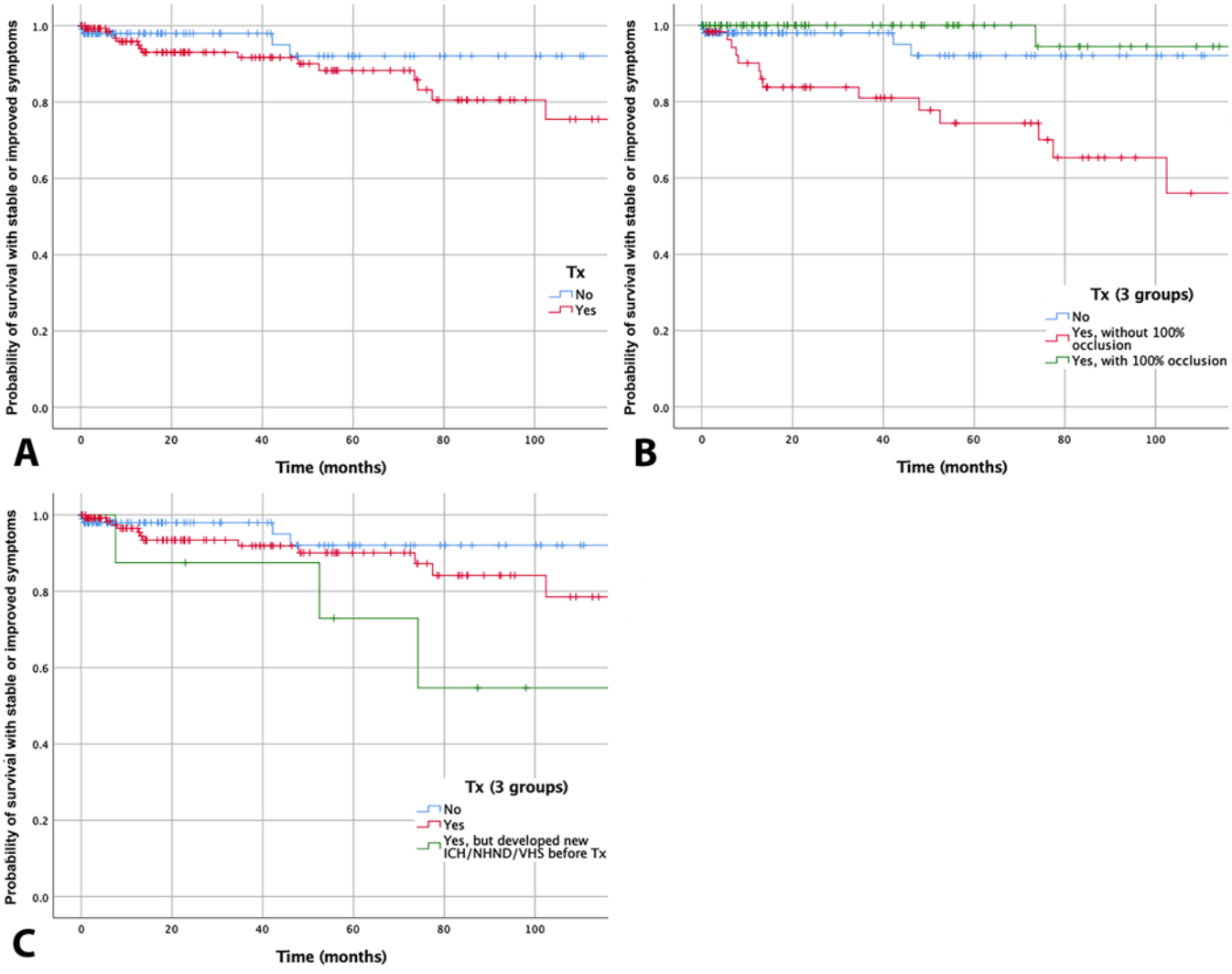

FIG. 2. Kaplan-Meier curves. **A:** Stable/improved symptoms survival probability in treated versus nontreated type I dAVFs. **B:** Subgroup survival analysis for treated dAVFs that achieved complete angiographic obliteration. **C:** Subgroup survival analysis for "rapidly evolving" treated dAVFs that developed new ICH, NHND, or VHS soon after diagnosis. Tx = treatment. Figure is available in color online only.

Discussion

This is the largest reported cohort of patients with dAVFs without CVD. This study confirms the benign natural history of these lesions when only the risk of ICH is accounted for. However, almost 12.6% of dAVFs without CVD presented with NHND or severe VHS. Despite multiple interventions, complete angiographic cure was achieved in only 55.2% of patients. This group of patients exhibited better estimated survival time than untreated or partially treated patients.

Natural History

Data regarding the natural history of fistulas without CVD is scarce. Gross et al. reported no ICH or NHND in 126 type I dAVFs;[9] no patient developed ICH or NHND over a total of 177 lesion-years (mean 1.4 lesion-years). Satomi et al. described the second largest cohort of dAVFs without CVD.[8] No ICH or NHND at presentation was reported among 117 fistulas. One symptomatic ICH and no NHND occurred over a mean follow-up of 2.3 years; the calculated annual neurological event rate was 0.6%. However, 36.8% of patients underwent treatment because of unbearable symptoms as described by the authors.

Defining these lesions as benign solely based in the incidence of ICH may underrepresent the disease process. NHND and VHS could be disabling and prompt treatment. Most prospective survival studies have censored their analyses only for development of new ICH or NHND, based on the concept that other VHS (e.g., tinnitus or ophthalmological symptoms) have a less aggressive clinical course.[5] In the CONDOR database, approximately 12.6% of patients with type I dAVFs presented with NHND or severe VHS. Hence, we used death and new or worsening NHND or VHS in our survival analysis as censoring events over time. CCFs were excluded from the outcomes analysis since these fistulas behave differently from classic dAVFs without CVD.

Treatment Outcomes

Previous studies have suggested that treatment of dAVFs without CVD is only aimed at symptom palliation without the need to achieve complete cure.[10–12] Our results suggest that complete angiographic obliteration is necessary to achieve clinical improvement and stop symptom progression. Despite the low complete angiographic obliteration rate achieved in this cohort (55.2%), our survival analysis demonstrated a trend for better symptom-free survival in treated dAVFs compared with nontreated dAVFs. In fact, this difference was statistically significant when complete angiographic obliteration was achieved. Partially treated lesions demonstrated a trend toward worse symptomatic outcome compared with nontreated type I dAVFs. This suggests that partial treatment confers no benefit in the natural history of these fistulas. The CONDOR database includes a broad array of treatment modalities, as it spans from 1990 to 2017. With the advent of newer liquid embolics and embolization devices, endovascular treatment may improve success rates and symptom resolution.[13,14]

Davies et al. described "palliative transarterial embolization" in 22 patients with type I dAVFs, with a 9% complication rate.[15] The indications for intervention were disabling bruit and headache and progressive ocular symptoms. Satomi et al. performed palliative embolization in 43 (36.8%) of 117 patients with type I dAVFs because of unbearable symptoms or pressing ophthalmological symptoms.[8] A 9.3% complication rate was reported. Shah et al. reported a 7.6% complication rate in treating 13 (56%) of 23 type I dAVFs.[10] The main indications for treatment were intolerable tinnitus or ophthalmological symptoms. Complete obliteration of the fistula was achieved in only 4 (30%) of 13 cases. Treatment of these complex and heterogeneous lesions may be challenging. Success and complications depend on the treatment modality and the operator's experience.[16] Our survival analysis censored by symptom improvement suggests that palliative treatment does not provide better outcomes. Treatment should be aimed at achieving complete angiographic closure of the fistula. In the CONDOR database, 99 (45%) of 218 treated patients achieved complete improvement and/or resolution of symptoms versus 40/103 (38%) of untreated patients. The rate of permanent treatment complications was 2.6%.

Evolution

Approximately 21.4% of nontreated dAVFs in our cohort showed partial spontaneous regression on angiographic follow-up. Gross et al. reported that 3% of dAVFs thrombosed spontaneously on follow-up.[9] Previous studies have suggested that the slow flow of some of these low-grade dAVFs may trigger spontaneous thrombosis.[17] A small subset of these lesions, albeit extremely rarely, can develop CVD and subsequent venous stenosis, thrombosis, increased arterial flow, and/or de novo fistula formation.[18] Shah et al. estimated a 1% annual risk of conversion of type I dAVF to high-grade dAVFs.[10] Satomi et al. reported a 1.7% risk of development of CVD in type I dAVFs.[8] The conversion from benign to aggressive dAVF was associated with spontaneous progressive thrombosis of venous outlets. In our cohort, 2.2% of fistulas developed CVD and 1.6% developed a new dAVF. Additionally, we found a 2.3% recurrence rate in patients with reported complete occlusion after treatment. The mechanisms behind dAVF recanalization are complex, as they may reappear after recruitment of a new arterial feeder due to incomplete occlusion of the venous pouch.[19] This heterogeneous evolution warrants close follow-up of these patients. In this study, the time from type I dAVF cure to recurrence was 34 months.

Finally, we identified a subgroup of patients with type I dAVFs who developed new ICH, NHND, or VHS soon after diagnosis. In these patients, treatment does not seem to significantly improve prognosis compared with nontreatment. Further characterization of the angioarchitecture and clinical characteristics of these lesions would facilitate prompt recognition of these "fast decliners" in order to offer appropriate treatment. Our preliminary data suggest that older age, NHND or severe VHS at presentation, and infratentorial location are predictors of worse outcome.

Limitations

The CONDOR database is an international collaboration to gather data of these uncommon neurovascular lesions; therefore, management, treatment decision, and sur-

gical protocols are not homogeneous. However, this is a reflection of real-life scenarios and endovascular practices around the world. The selection of endovascular technique and embolic materials and the wide time frame during which patients were recruited generate potential interinstitutional and technical disparities that affect outcomes. Additionally, current data collected in the common consortium database did not allow us to adjudicate the etiology of angiographic venous ectasia in this cohort (10.5%). However, this could be attributed to 1) complete occlusion of the sinus drainage (9.4%); or 2) occlusion of a common arterial collector, which is sometimes confounded with a cortical vein.[20] Also, since symptomatic dAVFs undergo treatment more often than incidental dAVFs, there is an intrinsic severity imbalance between treated and nontreated cases. Inadvertently, this could exert a dual effect over our survival analysis. First, it could increase the odds of reporting apparent clinical improvement in the treated arm of our study, thus increasing survival probability. On the other hand, some treated dAVFs could have a considerably poorer pretreatment clinical status compared with nontreated dAVFs, making them more likely to develop events sooner in time and decrease their survival probability. To overcome these biases, we redefined "initial time" as time of treatment (instead of time of presentation/diagnosis) for patients in the treated arm who developed new ICH, NHND, and VHS soon after diagnosis.

Conclusions

Complete angiographic occlusion of dAVFs without CVD provides longer symptom-free survival when compared to partially treated or untreated fistulas. Although these lesions are relatively benign, a smaller group of patients may require prompt therapeutic intervention to prevent significant neurological deterioration over time. This group of patients is older, has NHND or severe VHS at presentation, and an infratentorial dAVF location.

Appendix

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Disclosures

Dr. Samaniego: consultant for MicroVention/Medtronic. Dr. Levitt: ownership in Synchron, Cerebrotech, and Proprio. Dr. Kansagra: consultant for MicroVention and Penumbra; and support of non–study-related clinical or research effort from MicroVention and Medtronic. Dr. Lanzino: consultant for Superior Medical Editing and Nested Knowledge. Dr. Polifka: consultant for DePuy Synthes. Dr. Gross: consultant for Medtronic and MicroVention. Dr. Alaraj: consultant for Cerenovus and Siemens. Dr. Derdeyn: ownership in Pulse Therapeutics; consultant for Penumbra, Rapid Medical, and NoNo; and clinical or research support for this study from Siemens Healthineers.

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Supplemental Information

Companion Papers

Zipfel GJ: Introduction. The Consortium for Dural Arteriovenous Fistula Outcomes Research (CONDOR). DOI: 10.3171/2021.1.JNS2174.

Cockcroft KM: Editorial. The challenges of managing "benign" disease. DOI: 10.3171/2020.10.JNS203420.

Guniganti R, Giordan E, Chen CJ, Abecassis IJ, Levitt MR, Durnford A, et al: Consortium for Dural Arteriovenous Fistula Outcomes Research (CONDOR): rationale, design, and initial characterization of patient cohort. DOI: 10.3171/2021.1.JNS202790.

Chen CJ, Buell TJ, Ding D, Guniganti R, Kansagra AP, Lanzino G, et al: Intervention for unruptured high-grade intracranial dural arteriovenous fistulas: a multicenter study. DOI: 10.3171/2021.1.JNS202799.

Abecassis IJ, Meyer RM, Levitt MR, Sheehan JP, Chen CJ, Gross BA, et al: Assessing the rate, natural history, and treatment trends of intracranial aneurysms in patients with intracranial dural arteriovenous fistulas: a Consortium for Dural Arteriovenous Fistula Outcomes Research (CONDOR) investigation. DOI: 10.3171/2021.1.JNS202861.

Abecassis IJ, Meyer RM, Levitt MR, Sheehan JP, Chen CJ, Gross BA, et al: Recurrence after cure in cranial dural arteriovenous fistulas: a collaborative effort by the Consortium for Dural Arteriovenous Fistula Outcomes Research (CONDOR). DOI: 10.3171/2021.1.JNS202033.

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