On-line Table 1: Presentation and fistula architecture stratified by fistula progression[a]

	All Subjects (*n* = 34)	Progressive (*n* = 19)	Nonprogressive (*n* = 15)	*P* Value
Baseline clinical characteristics				
Mean age at dx ± SD	45 ± 23 y	36 ± 25 y	57 ± 13 y	.0059[b]
Median age (range)	52.5 y (0.2–77 y)	38 y (0.2–71 y)	59 y (19–77 y)	
Male	35% (12/34)	42% (8/19)	27% (4/15)	.48[c]
Trauma	21% (7/34)	21% (4/19)	20% (3/15)	1.0[c]
Hemorrhage	26% (9/34)	37% (7/19)	13% (2/15)	.24[c]
Seizures	18% (6/34)	21% (4/19)	13% (2/15)	.67[c]
Dizziness	21% (7/34)	11% (2/19)	33% (5/15)	.20[c]
Syncope	6% (2/34)	11% (2/19)	0% (0/15)	.49[c]
Tinnitus	38% (13/34)	42% (8/19)	33% (5/15)	.73[c]
Bruit	41% (14/34)	37% (7/19)	47% (7/15)	.73[c]
Thrill	6% (2/34)	5% (1/19)	7% (1/15)	1.0[c]
Headache	47% (16/34)	42% (8/19)	53% (8/15)	.73[c]
CHF	0% (0/34)	0% (0/19)	0% (0/15)	NA
Cranial neuropathy	47% (16/34)	47% (9/19)	47% (7/15)	1.0[c]
Focal neurologic deficit	32% (11/34)	37% (7/19)	27% (4/15)	.72[c]
Hearing loss	9% (3/34)	11% (2/19)	7% (1/15)	1.0[c]
Developmental delay	6% (2/34)	11% (2/19)	0% (0/15)	.49[c]
Altered mental status	15% (5/34)	11% (2/19)	20% (3/15)	.63[c]
Cognitive dysfunction	9% (3/34)	5% (1/19)	13% (2/15)	.57[c]
Chemosis or proptosis	29% (10/34)	32% (6/19)	27% (4/15)	1.0[c]
Papilledema	9% (3/34)	11% (2/19)	7% (1/15)	1.0[c]
Visual changes	35% (12/34)	32% (6/19)	40% (6/15)	.72[c]
Retinal hemorrhage	6% (2/34)	0% (0/18)	13% (2/15)	.19[c]
Median baseline mRS (25%, 75%)	1 (1, 3)	1.5 (1, 3)	1 (1, 3)	.27[d]
Range baseline mRS	1–5	1–5	1–5	
Baseline angioarchitecture				
Ectasia of draining vein		58% (11/19)	27% (4/15)	.09[c]
Retrograde venous drainage (0, 1, 2)		2 (0, 2)	1 (0, 2)	.36[d]
Periventricular drainage		21% (4/19)	13% (2/15)	.67[c]
Venous stenosis (1–5)		3 (1, 5)	3 (1, 4)	.64[d]
Venous varix		47% (9/19)	13% (2/15)	.064[c]
Sinus stenosis or occlusion (1–5)		5 (1, 5)	1 (1, 5)	.091[d]
Persistent fetal venous structures		26% (5/19)	7% (1/15)	.20[c]
Median number of AV connections (25%, 75%)		4 (2, 4)	3 (3, 4)	.72[d]
Range of AV connections		1–7	1–5	
Pseudophlebitic cortical venous pattern (0, 1, 2)		1 (0, 2)	0 (0, 1)	.048[d]
Venous sinus dilation		41% (8/19)	7% (1/15)	.045[c]
Borden-Shucart (1–3)		2 (2, 2)	2 (1, 3)	.77[d]
Cognard (1–5)		4 (3, 4)	4 (1, 5)	.57[d]
Fistula location				
Transverse sigmoid sinus		53%	73%	.30[c]
Superior sagittal sinus		26%	20%	.71[c]
Torcula		5%	33%	.07[c]
Cavernous sinus		26%	20%	.71[c]
Marginal		16%	13%	1.00[c]
Petrosal		5%	20%	.30[c]
Occipital		11%	0%	.49[c]
Other intracranial location		26%	27%	1.00[c]
Baseline imaging				
Prior hemorrhage		29% (5/17)	7% (1/14)	.20[c]
Parenchymal edema		27% (4/15)	10% (1/10)	.61[c]
Current hemorrhage		24% (4/17)	0% (0/14)	.11[c]
WM calcifications		14% (2/14)	11% (1/9)	1.0[c]
Ischemia		7% (1/15)	11% (1/9)	1.0[c]
Encephalomalacia		13% (2/15)	11% (1/9)	1.0[c]
Hydrocephalus		6% (1/16)	0% (0/10)	1.0[c]
Mass effect		29% (5/17)	0% (0/12)	.059[c]

Note:—NA indicates not applicable; AV, arteriovenous; CHF, congestive heart failure; dx, diagnosis.

[a] Progressive group includes patients with enlarging fistulas or de novo fistula formation with time. Nonprogressive group includes patients with multiple synchronous fistulas or nonenlarging fistulas that recurred after treatment. Note that for some variables, complete patient data were either not acquired (eg, a particular angioarchitectural feature could not be assessed on available angiograms; no cross-sectional imaging study was performed) or results were not obtainable; in these cases, the denominator has been provided. Altered mental status includes acute confusion, cognitive impairment (including by self-report), and/or altered level of consciousness.

[b] Two-tailed *t* test.

[c] Two-sided Fisher exact test.

[d] Wilcoxon rank sum (Mann-Whitney) test.

On-line Table 2: Detailed initial clinical presentation and outcomes with modified Rankin Scale scores

Patient No.	DAVF Characteristics	Malignant or "Runaway" Course[a]	Clinical Presentation	mRS Before	Clinical Outcome	Last mRS
1	P	1	Normal neonatal period; multifocal seizures at 68 days, CT head with R frontal and L occipital SAH/SDH; MRA/MRV complex vascular malformation	4	Code status DNI/DNR; died with home hospice; last f/u: poor neuro exam, no interaction with environment; 3–7 seizures per day, on phenobarbital; 3–4 episodes of autonomic storming per day	6
2	P	1	Focal and generalized seizures at 3 mo; R occipital DAVF with pial component; global developmental delay, nystagmus	4	No residual DAVF; mild gait ataxia; myopia/cortical visual impairment; speech therapy, OT/PT, making global progress; falls from time to time but no significant injuries; interacts appropriately	3
3	P	1	R frontotemporal HA, R peripheral CN VII palsy for 4 mo; R eye proptosis/ex-opthalmos progressing to R ophthalmoplegia; CT showed R retro-orbital mass	1	Also developed right orbital mixed venous lymphatic malformation, s/p orbitotomy; last f/u at 13 y: no HA or eye pain, no tinnitus; no neuro symptoms, interactive, doing well in school	1
4	P	1	L forehead angioma at 9 mo; minor trauma to L eye at 18 mo; led to progressive proptosis, diplopia; at 2 y, diagnosed with multiple DAVFs; multiple residual DAVF after tx at OSH; proptotic L eye, chemosis, discoloration; doing well on modified school curriculum; acute-onset severe HA, n/v, dizziness	2	Last angio: extensive, multiple DAVFs of intracranial sinuses and cortical veins; L eye proptosis due to left orbital apex and cavernous sinus fistula, not amenable to embo; at time of d/c: no new neuro symptoms; unchanged L eye proptosis, L orbital/cranial bruit	1
5	P	1	At 9 y, macrocephaly, skin lesions, cranial bruit, prominent neck pulses, cardiac murmur, RVH; overall function: ADD with "above average IQ" but c/o decline in cognitive function	2	Grand mal seizure after final embo, had not taken AED for several days; was at baseline mental status at death; died 6 mo later	6
6	P	1	Fell from tree at 10 y; chronic HA at 12 y; found to have diffuse dural vein thrombosis; anticoagulated; new subtle papilledema, L ear bruit, then c/o tinnitus, worsening HA; at 13 y, admitted for confusion, aphasia, R weakness	4	Last f/u: no neurologic problems noted	0
8	P	1	HA at 16 y for 6 y; presented to OSH at 21 y with progressive RUE numbness; pulsatile tinnitus when bending over; exam: palpable thrill in R post auricular and occipital regions, pulsatile cranial bruit, mild RUE dysmetria	1	R hearing loss; enrolled in junior college	1
9	P	1	Progressive facial asymmetry, L ear tinnitus when post partum at 18 y; embo twice at 19 y and 22 y in Philippines; developed proptosis, worsening tinnitus; exam: skull base bruit, blurred vision, facial weakness, tongue weakness, hearing deficit	2	Last admit for L thalamic intracerebral hemorrhage related to DAVF; multiple fistulas not amenable to embo or surgery; started minocycline in hopes of reducing fistula growth; last exam: mild general limb weakness; slow, wide-based gait with short shuffling steps, ambulating independently; failed swallow evaluations, on tube feeing; patient died 1 mo later on comfort care	6

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On-line Table 2: Continued

Patient No.	DAVF Characteristics	Malignant or "Runaway" Course[a]	Clinical Presentation	mRS Before	Clinical Outcome	Last mRS
10	P	1	Pulsatile tinnitus in R ear for 4 months when pregnant; miscarried; pulsatile tinnitus became progressively louder and more bothersome	1	No residual DAVF, no symptoms	0
11	P	1	Rushing noises in ears, vertical diplopia, dizziness, LUE/LLE shaking and weakness at 36 y; papilledema; LP with elevated pressures; symptoms progressed at 38 y with pulsatile tinnitus, episodes of confusion/disorientation, possible LOC; MRI/MRV showed thrombosis of anterior half of SSS, prominent collateral venous structures in parasagittal, supraorbital, infraorbital regions; tx: warfarin, high-volume LP; no improvement	3	No new DAVF; syncope multiple times per day; found to have ICP spikes due to venous sinus thrombosis causing intermittent translocation of cerebellar tonsils; VP shunt improved symptoms; last f/u: reports occasional syncopal episodes or confusion; unilateral weakness resolved	2
12	P	1	Severe whiplash injury and neck injury due to MVA at 39 y; developed progressive pulsatile tinnitus at 48 y; pulsatile tinnitus, odd taste in mouth, bilateral face and neck numbness, HA; 1 episode of "near-syncope"; MRI showed AVM/AVF	1	C/o headache, constant tinnitus waxing/waning in severity for several years but MRI and angio negative for fistula; normal neuro exam findings; low-grade symptoms attributed to stress and fatigue	1
13	P	1	Horse accident with LOC at 33 y; mild migraine HA, severe R occipital HA, R pulsatile tinnitus at 51 y	1	Last progress note: normal neuro exam findings, feeling well, headaches overnight relieved by oral opiates, dorsal R foot pain without deficits	1
15	P	1	Presented at 52 y with L frontal ICH; work-up in Korea revealed multiple DAVFs, partially embolized	2	New low-risk DAVF not embolized; last progress note: patient doing well	0
19	P	1	R tinnitus since 48 y; fell off exercise equipment with severe bump to head at 56 y; slight bruit over occiput at 57 y; acute onset HA 1 y later; CT/MRI at OSH revealed small ICH and DAVF; embo, repeat angio showed residual DAVF	1	Stable low-risk left DAVF and stable right carotid ophthalmic aneurysm; R arm and leg numbness without motor weakness; no significant symptoms	1
25	P	1	Diplopia, intermittent nausea for 1–2 mo; OSH exam: R chemosis, proptosis, ophthalmoparesis, decreased visual acuity, R carotid pulse synchronous bruit; outside CT confirmed CCF	1	Last f/u: R CN VI and minimal R eye proptosis, unchanged bruit; no new plans for repeat imaging	1
29	P	1	HA since youth; at 62 y, HA worsened, now anterior aspect of head; at 63 y, diplopia, L eye pain; exam: L chemosis, proptosis, visual loss, partial CN VI palsy; CT confirmed indirect left cavernous DAVF	1	Normal neuro exam findings; no symptoms reported after final embo	0
32	P	1	Diplopia with mild R chemosis, bruit; OSH DSA showed DAVF	1	No symptoms noted	0

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On-line Table 2: Continued

Patient No.	DAVF Characteristics	Malignant or "Runaway" Course[a]	Clinical Presentation	mRS Before	Clinical Outcome	Last mRS
34	P	1	At 77 y, admitted to OSH for R hemiplegia, aphasia, LOC, generalized seizures; found to have L frontal ICH; status epilepticus 6 mo later; MRA: TS DAVF	5	Altered mental status and neurologic deficits; complex partial seizures and CSF leak after craniotomy; underwent leak repair but CT scan 12 hours postoperation showed large R holohemispheric acute-on-chronic subdural hematoma with midline shift; patient transitioned to comfort care and died secondary to respiratory failure	6
30	P	1	Seizures at 17 y, well-controlled on AED; at 42 y, acute-onset HA, n/v, L hemianesthesia, L hemiparesis; OSH CT/MRI: R parietal IPH	3	No symptoms, normal neuro exam findings at last follow-up	0
7	NP	0	Severe HA at 16 y; found to have cerebellar bleed; 2-mo DSA negative; 4 mo later, new HA, dizziness, diplopia; OSH imaging: extensive SSS thrombosis; exam: bilateral papilledema and CN VI palsy	1	Last angio: no residual DAVF, dural venous sinuses patent; on life-long anticoagulation; migraine HA resolves with topiramate; last progress note: no neuro deficits, no papilledema, normal visual fields	0
27	NP	0	MVA with whiplash injury at ~47 y; severe R temporal HA, retro-orbital pain; exam: retinal hemorrhage; treated for pseudotumor cerebri then developed bruit in ear; OSH angio: DAVF	1	Normal neuro exam findings	0
16	NP	0	Left posterior auricular bruit for 4 mo; normal MRI findings at OSH	1	Last angio: residual L sigmoid sinus DAVF, interval decrease in AV shunting; asymptomatic	0
17	NP	0	MVA at 34 y; L occipital thrill and bruit at 48 y; MRI: L dural sinus fistula, not treated; symptoms improved then worsened at 52 y with new HA	1	Tinnitus resolved after final embo; no symptoms; barely audible bruit on exam	0
21	NP	0	57 y, L HA, bruit in L ear; tx with prednisone at OSH, resolved; recurrence 3 mo later in R ear (bruit, tinnitus); R diplopia/blurred vision 2 months later; proptosis, chemosis; OSH DSA: bilateral CC fistulas, AVM; at 59 y, minimal L temporal bruit	1	One low-risk DAVF remaining; Last f/u: headache, possible bruit over R transverse sinus	1
22	NP	0	58 y "spacey" feeling, n/v, HA, imbalance, difficulty walking, increased fatigue, cognitive dysfunction, including short-term memory loss, episodes of confusion; MRI/MRV at OSH: R TS occlusion; tx: warfarin, rehab; some improvement	3	Last angio normal findings; neuro exam: impaired coordination and gait difficulty; nausea for 8 mo; had LP to rule out NPH	3
23	NP	0	Seizures since 20 y; at 59 y AMS, intermittent L face and arm twitching at OSH; recurrent partial status epilepticus/focal seizures with secondary generalization; exam: drowsy, R peripheral facial droop	4	No overt seizures since last embo, though patient has periods of continuous, very-low-amplitude shaking of the left hand, not disabling per patient; moderate weakness in left arm and leg; walking with a walker	3

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On-line Table 2: Continued

Patient No.	DAVF Characteristics	Malignant or "Runaway" Course[a]	Clinical Presentation	mRS Before	Clinical Outcome	Last mRS
26	NP	0	Migraine HA, fell off barn onto head as child; at 51 y, L pulsatile tinnitus; papilledema noted at 56 y; LP: elevated opening pressure; had a grand mal seizure, poor balance, HA, d/w pseudotumor cerebri; various medication for 18 mo, symptoms resolved; at 61 y, simple partial motor seizure of LUE/LLE with postictal weakness lasting many months; MRI: vascular malformation, no tx; stroke 3 mo later, several simple partial motor seizures; tinnitus resolved spontaneously; DSA at OSH: DAVF, partially embolized (aborted due to hemorrhage or seizure)	3	No DAVF on last angio; residual mild L hemineglect, impaired attention, short-term memory loss, and mild weakness of L body	3
18	NP	0	P/w progressive ecchymosis, proptosis, ophthalmoplegia, bruit for unknown duration	1	Patient c/o heaviness over R eye	1
20	NP	0	R pulsatile tinnitus of unknown duration; OSH work-up: DAVF	1	Last f/u: normal neurologic exam findings and function; no symptoms	0
24	NP	0	MVA at 61 y, admitted to OSH GCS 3; head CT: R frontal and sphenoid fx, R frontal contusion, L temporal contusion, facial fx, global injury and IPH; transferred for treatment of CCF after ~3 mo; chemosis, proptosis, no extraocular motions, and no light perception in his left eye; L pupil fully dilated; bruit in the left side of his head; dysmetria; altered mental status, amnesia	5	Last neuro exam before discharge: no light perception OS, no change to baseline vision OD; no symptoms	1
28	NP	0	64-y persistent dizziness, HA, intermittent diplopia, visual changes for 1 week, followed by severe eye pain; dx: CC fistula; tx: Inderal (propranolol), steroid taper; symptoms improved, then recurred at 1 mo; "thrills in both ears," tinnitus, pins and needles feeling in head/scalp/face, nausea, tingling in L foot/shin; thick serous discharge in both eyes, mild chemosis of L eye, bruit along R posterior mastoid process	1	No residual DAVF; no symptoms	0
31	NP	0	Clicking sound in head for unknown duration, pulsatile tinnitus, dizziness, nausea; bruit; occasional minor tic since childhood	1	No residual DAVF; patient reports mild numbness over left ear after craniotomy	1
33	NP	0	History of transient global amnesia; MRI: fusiform aneurysm; c/o, dizziness, occasional vertigo, sinus HA, L sharp frontal HA; reports twice of having a sudden vision change, visualizing something, and having a different vision suddenly come to mind	1	No residual DAVF; no symptoms	0

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On-line Table 2: Continued

Patient No.	DAVF Characteristics	Malignant or "Runaway" Course[a]	Clinical Presentation	mRS Before	Clinical Outcome	Last mRS
14	NP	1	Dizziness if turning head to L for "decades"; significant R hearing loss for "many years," also some L hearing loss; R facial palsy in LMN distribution at 71 y; horizontal diplopia of R eye attributed to previous embolizations	1	Seizure 481 days after last embo; negative head CT findings; impaired balance, multiple falls, cognitive difficulties, impaired speech production, change in writing; floaters in R > L eye; bilateral hearing loss unchanged; neuro exam: truncal ataxia, transient fleeing horizontal diplopia to R gaze, deficits in fine motor and coordination of RUE and RLE	3

Note:—P indicates progressive; NP, nonprogressive; R, right; L, left; SDH, subdural hematoma; DNI/DNR, Do Not Intubate, Do Not Resuscitate; HA, headache; OSH, outside hospital; RVH, right ventricular hypertrophy; ADD, attention-deficit disorder; IQ, intelligence quotient; OT/PT, occupational therapy and physical therapy; f/u, follow-up; AED, antiepileptic drug; RUE, right upper extremity; LUE/LLE, left upper extremity/left lower extremity; LP, lumbar puncture; LOC, loss of consciousness; ICP, intracranial pressure; VP, ventriculoperitoneal; c/o, complained of; CCF, carotid cavernous fistula; CN, cranial nerve; ICH, intracerebral hemorrhage; Ts, transverse sigmoid; AED, antiepileptic drug; IPH, intraparenchymal hemorrhage; SSS, superior sagittal sinus; MVA, motor vehicle collision: CC, carotid cavernous; AMS, altered mental status; NPH, normal pressure hydrocephalus; GCS, Glasgow Coma Scale; OS, left eye; OD, right eye; neuro, neurological; embo, embolization; s/p, status post; tx, treatment; angio, angiography; TS, transverse sigmoid; n/v, nausea vomiting; d/w, diagnosed with; d/c, discharged; P/w, presented with; fx, fracture; LMN, lower motor neuron; rehab, rehabilitation; dx, diagnosis; AV, arteriovenous.

[a] 0 = no, 1 = yes.