

## The Brain Aneurysm Institute

Multidisciplinary Care of Patients with Hemorrhagic and Ischemic Stroke

SPRING 2022

# Neurovascular News



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## Idiopathic Intracranial Hypertension: Vascular Origins and Defining a Modern Management Protocol at the BIDMC Brain Aneurysm Institute

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Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri or benign intracranial hypertension, is a syndrome characterized with elevated intracranial pressure. It is often associated with obesity, elevated intra-abdominal pressure, decreased conductance of CSF outflow, certain drugs (Vitamin A, tetracycline) and systematic diseases (such as lupus erythematosus, uremia, hypothyroidism). The overall incidence of IIH is approximately 1.2/100 000, and predominantly seen in young women at childbearing age. In addition to headaches, increased ocular pressure can occur with associated papilledema and visual loss.

The initial management of IIH is largely through medical interventions. However, there is no consensus on the optimal management of IIH in refractory cases. There are several systematic reviews on the surgical treatment of IIH, and different groups have adopted various aspects of the treatment options based on the available local expertise. Venous sinus stenosis has been defined as a cause of IHH in some patients and there is one meta-analysis that performed

a stratified, rigorous analysis on the effectiveness of venous sinus stenting (VSS) in the management of IIH.<sup>1</sup> A similar analysis is lacking for CSF shunting procedures and herein, we performed a systematic review and meta-analysis on CSF shunting in refractory IIH. We used this analysis in conjunction with VSS data to develop a modern protocol for the management of IIH that would be applicable at most centers.

### Systematic review and meta-analysis on cerebrospinal fluid shunting

PubMed and Embase were systematically searched for studies describing CSF shunting for medically refractory IIH. Relevant information including study characteristics, patient demographics, clinical outcomes, peri-procedural complications, and long-term outcomes were subjected to meta-analysis.

Fifteen studies published between 1988 and 2019 met our inclusion and exclusion criteria, providing 372 patients for analysis, 311 (83.6%) of which were female. The mean age of patients was 31.2 years (range 0.5-71). Of the 372 patients included,

**Table 1:** Study of characteristics and patient demographics

Author and year	number of patients	Female (%)	Age (range or SD)	CSF opening pressure	Shunt type	Follow up months (range or SD)
Johnston et al 1988	36	72.2%	24.7 (0.5-71)	NR	LPS (34) and CAS (2)	42.9
Eggenberger et al 1996	27	88.9%	30 (8-52)	NR	LPS	77 (21-278)
Burgett et al 1997	30	93.3%	32.9 (11-68)	NR	LPS	35 (0-143)
Maher et al 2001	13	76.9%	31.5 (6-54)	NR	VPS	15.2 (1-38)
Bynke et al 2004	17	70.6%	34 (13-63)	NR	VPS	78 (21.6-153.6)
Woodworth et al 2005	21	81.0%	42±17	NR	VPS	20±17
Abu-Serieh et al 2007	9	55.6%	26.4 (4-63)	NR	VPS	44.3 (6-110)
Kandasamy et al 2011	17	29.4%	23.9 (3-52)	NR	VPS	21 (9-49)
Sinclair et al 2011	53	94.3%	30.3±8.5	39.5±8.2 cm H2O	LPS (49) and VPS (4)	24 (1-84)
Tarnaris et al 2011	34	94.1%	35±7.9	39.4±10.3 mmHg	LPS (25) and VPS (9)	28.9±31.8
El-Saadany et al 2012	22	81.8%	28.5 (20-38)	NR	LPS	12 (NR)
Yadav et al 2012	24	91.7%	39 (17-58)	NR	LPS	51 (18-137)
Niotakis et al 2013	7	71.4%	8.7 (6-14)	41.35±5.42cmH2O	LPS	26 (8-60)
Karsy et al 2016	34	91.2%	34.4 (18-50)	39.6±9 cmH2O	VPS	24.8 (1.3-71.9)
Bjornson et al 2019	28	92.9%	29.3 (6-48)	NR	VPS	17 (4-96)
Totals	372	83.6%	31.2			33.9

SD= Standard deviation; NR= Not reported; LPS= Lumboperitoneal shunting; VPS= Ventriculoperitoneal shunting; CAS= Cisternoatrial

218 (58.6%), 152 (40.9%) and 2 (0.5%) had lumboperitoneal shunting (LPS), ventriculoperitoneal shunting (VPS) and cisternoatrial shunting (CAS), respectively. Average follow up duration was 33.9 months (range 0-278 months). (Table 1)

Overall, the rate of improvement in headache was 91% (95% CI 84-97%). The improvement rate for papilledema was 96% (95% CI 85-100%). Outcomes for visual impairment were recorded in 14 studies. Most studies reported visual impairment as an independent symptom while a few studies separated it into decreased visual field and visual acuity. The overall improvement rate for visual impairment after shunting was 85% (95% CI 72-95%).

All of the studies recorded periprocedural outcomes or complications. However, the definition of complication was not standardized. In terms of over-drainage of CSF through shunting, 74 patients out of 372 experienced periprocedural low-pressure headache reported in 12 studies. The overall rate for low pressure headache was 20% (95% CI 11-32%). There was significant heterogeneity between studies (I<sup>2</sup>=81.07%).

In terms of durability of shunting, all the studies recorded the number of patients who had revision and total number of revisions. Out of 372 patients 155 had 436 revisions in total during 33.9 months of follow up. The overall revision rate was 42% (95% CI 26-59%). There was no significant correlation between average follow up duration and revision rates in studies (P=0.627). As shown in the table, the most common reason for revision was obstruction with recurrence of symptoms and signs of increased intracranial pressure. Other common reasons included low pressure headache from over-drainage, catheter migration and infection. The reasons for shunt obstruction were only described for 28 cases out of the 107 reported obstructions. All of these cases received VPS. Distal catheter obstruction was the reason for obstruction in 23 cases (82.1%), proximal obstruction in 3 cases (10.7%), and valve obstruction/malfunction in 2 cases (7.1%).

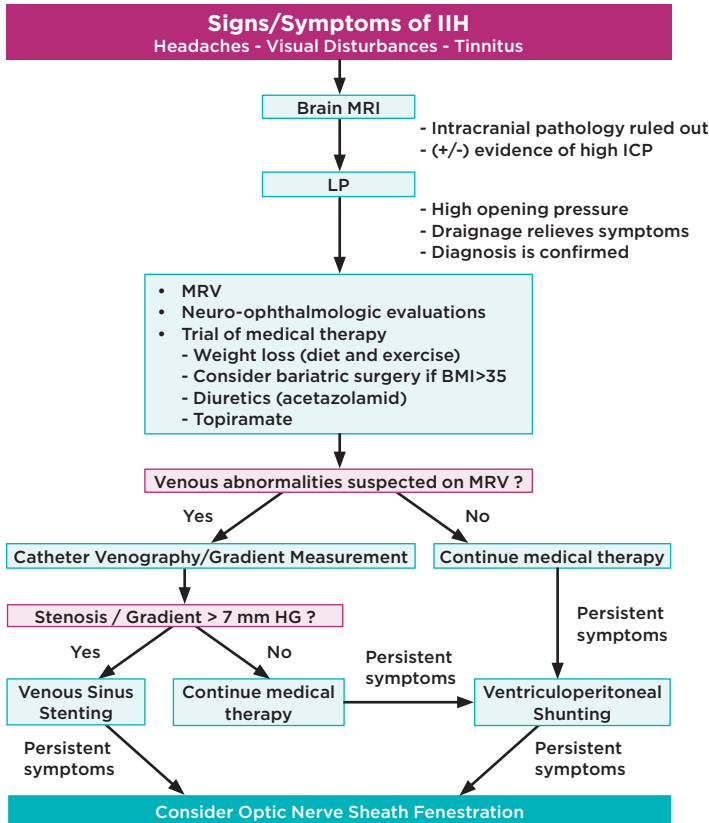
**Current management protocol of IIH** Based on available literature, our current meta-analysis, and the evolution in treatment options for patients with IIH, a reasonable algorithm for the management of

these patients is presented in Figure 1. Early establishment of the diagnosis is critical; this is based on the most common presenting symptoms of headache, visual loss or visual changes, and tinnitus, followed by objective evaluations that consist of neuroimaging studies, a formal ophthalmology evaluation, and a lumbar puncture performed under carefully controlled circumstances. The ophthalmology evaluation includes assessment of visual acuity, visual fields, papilledema grading, color vision testing, and optical coherence tomography (OCT) testing.

Neuroimaging evaluation includes a high-quality MRI; signs suggestive of IIH include flattening of the posterior scleral margins, widening of the peri-optic cerebrospinal fluid space, tortuosity of the optic nerves, “empty-sella”, cerebellar tonsillar descent and very small or slit-like ventricles. This is complemented by an MRV study to assess the status of the intracranial venous sinus pathways and look for any focal stenosis, atresia, or other venous abnormalities that may predispose an individual to developing IIH.

If no contraindications exist, this is followed by a lumbar puncture (LP)

**Figure 1:**



to assess the opening CSF pressure and also allow removal of CSF from the lumbar subarachnoid cistern to ascertain any therapeutic benefit from CSF drainage. Routine CSF studies are also performed, and the patient is followed in clinic in 1-2 weeks to assess the response to CSF drainage. The diagnosis of IIH is confirmed by this collation of clinical symptomatology, neurological and ophthalmological evaluations, LP, and neuroimaging studies. Diagnostic criteria such as Modified Dandy's criteria and Friedman's criteria can be helpful in establishing the diagnosis of IIH.<sup>2,3</sup>

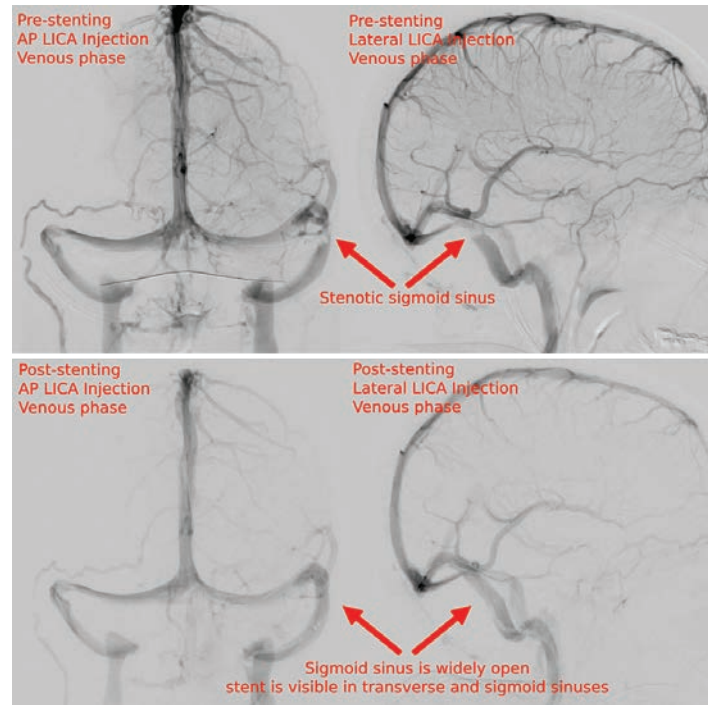
The initial step in all patients is optimization of medical therapy. Randomized controlled trials have substantiated the role for the carbonic anhydrase inhibitor, acetazolamide (Diamox) as being effective in alleviating the symptoms of IIH and decreasing papilledema and improving visual acuity and visual constrictions. Topiramate (Topamax) is also a carbonic anhydrase inhibitor

that has been used for IIH. In addition to optimal medical therapy, referral to a weight loss specialist for diet and activity modification and consideration for bariatric surgery for patients with a BMI>35 is initiated.

If venous sinus stenosis or abnormalities are identified on MRV or other non-invasive imaging, catheter venography is performed, and the pressure gradient is measured across the area of abnormality. A gradient greater than 7 mm Hg is considered indication for venous sinus stenting. Figure 2 shows such a patient with sinus stenosis before and after stenting. If there is no intracranial venous sinus stenosis, or the venous gradient is less than 7 mm Hg, then consideration is given to CSF diversionary procedures.<sup>4</sup>

The two main options for CSF diversionary procedures are the VP and LP shunt. The VP shunt is preferable when possible, considering numerous previous studies indicating

**Figure 2: (A)** Cerebral angiogram demonstrating venous sinus stenosis of the right transverse sinus in a 58-year old patient with atretic left transverse sinus. She had intractable headaches and papilledema with visual loss. The measured pressure gradient across the stenosis was 9 mm Hg. **(B)** Angiogram after venous sinus stenting demonstrating a widely patent transverse sinus with resultant abolished pressure gradient in the sinus. The patient had improvement of her headaches and improvement of her visual acuity and resolution of her papilledema.



that LPS is associated with an increased risk of revision by 2- to 2.5-fold compared to VPS.<sup>5,6</sup> Moreover, VPS does not harbor the risk of secondary Chiari 1 malformation and radiculopathy, although it adds the rare risk of brain parenchymal damage. Overdrainage is also a more prominent concern in LPS as the hydrostatic pressure effect of gravity on the cerebrospinal fluid column in upright position can aberrantly increase the drainage rate. If the ventricle cannot be cannulated, then an LP shunt is considered. However, with advent of frameless image-guided stereotactic shunt placement techniques, accurate VPS placement is now feasible in the majority of cases even with compressed ventricles. Presence of cerebellar tonsillar descent on neuroimaging studies is a contraindication for an LP shunt. All patients are evaluated with a CT head and shunt series to confirm optimal catheter placement. In patients

with a BMI>30 and with significant obesity, laparoscopic insertion of the peritoneal catheter by the general surgery team is performed, and CT of the abdomen obtained ahead. The use of a programmable valve with the LP shunt is also an option.

Re-evaluation and regular neurological and ophthalmological follow up is essential to ascertain the response to medical therapy, venous sinus stenting, or CSF shunting. Patients who have persistent visual loss that is refractory to these interventions are considered for optic nerve sheath fenestration procedures. In recent years, the option of endoscopic medial optic nerve decompression via a transnasal, transsphenoidal route has been

discussed but the long-term results are forthcoming.

The algorithm presented here is in line with recent systematic reviews of treatments of IIH, our understanding of the pathophysiology of the condition, and based on existing medical and surgical therapeutics. It allows the optimal utilization of resources to care for a complex group of patients; the treatments discussed above are complementary and will likely provide the best outcomes for patients with IIH.

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## Endovascular Treatment of Chronic Subdural Hematomas

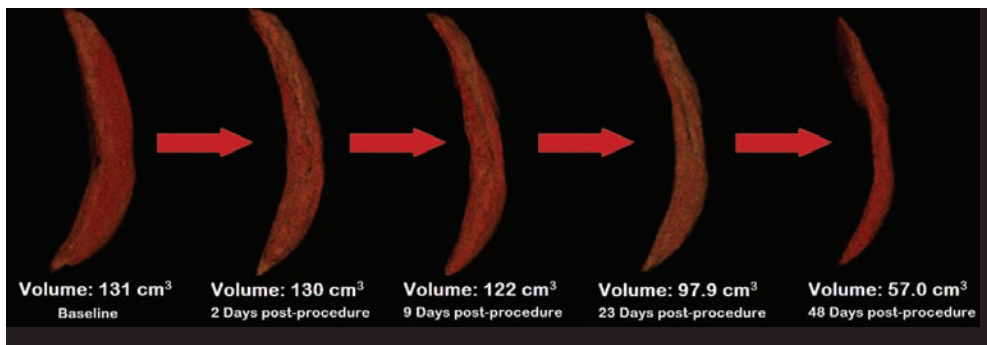
MirHojjat Khorasanizadeh, MD and Christopher Ogilvy, MD

Chronic subdural hematoma (cSDH) is among the most frequently encountered neurosurgical pathologies, the incidence of which is rising in concert with the aging population.<sup>1</sup> Surgical evacuation has long been the mainstay of treatment, but is associated with certain limitations, including high risk of recurrence. The rate of recurrence in surgically-treated cSDHs is often reported to exceed 20%.<sup>2</sup> Recently, middle meningeal artery embolization (MMAE) has been investigated as a therapeutic option for cSDH patients. Numerous studies have reported safety and efficacy of MMAE in treatment of cSDH.<sup>3-6</sup> Contrary to earlier assumptions, it is now established that cSDH is not purely due to the rupture of cortical bridging veins in the subdural space, as a cSDH is maintained by the leaky neovascularity formed in hematoma's membrane which are fed by the middle meningeal artery. MMAE studies have demonstrated how this procedure can reduce the risk of recurrence through devascularizing these immature leaky capillary networks.<sup>7</sup>

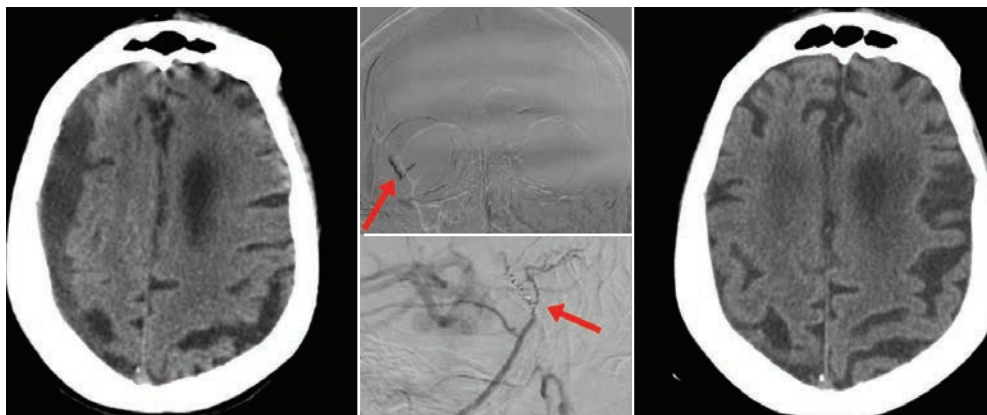
A typical MMAE procedure involves injection of polyvinyl alcohol particles (100-300 microns in diameter) or liquid embolic agents into the MMA, sometimes followed by coiling of the MMA branches by helical detachable coils. The procedure is generally safe and well tolerated. The most dreaded complications associated with MMAE are visual complications due to penetration of the embolic material from the main MMA trunk or early anterior division through the small meningolacrimal or sphenoidal branches into potential anastomoses with the ophthalmic artery.<sup>8,9</sup> If anastomoses between the MMA main trunk and the ophthalmic artery are identified, isolated use of coils or injection of the particles selectively into one or both distal branches should be attempted, so as to avoid the risk of inadvertent embolization of the central retinal artery resulting in vision loss. Anatomic features of the MMA including accessibility, tortuosity and diameter of the branches are other factors that are considered when planning the location for coiling

and particle injection.

The division of endovascular and operative cerebrovascular surgery at the Beth Israel Deaconess Medical Center has been one of the pioneers in adopting and optimizing this novel minimally invasive therapeutic approach. BIDMC has been one of the top 3 institutes in the United States in terms of the number of MMAE procedures performed. According to the latest data available, MMAE has enabled effective and safe treatment of cSDH in a large group of BIDMC patients. In our center, out of the 94 treated hematomas, 72 (76.6%) received upfront MMAE as treatment, 14 (14.9%) hematomas received burr-hole surgery in adjunction to MMAE, and 8 (8.5%) hematomas underwent MMAE following a prior failed burr-hole drainage of the cSDH. After a mean radiologic follow-up duration of 113.9 days, mean cSDH axial thickness and volume on the latest follow-up imaging were 5.7 mm and 29.6 ml respectively. Mean reduction in volume and axial thickness of the



**Figure 1.** Changes in volume of a chronic subdural hematoma following treatment with upfront middle meningeal artery embolization.



**Figure 2.** Complete resolution of a chronic subdural hematoma following middle meningeal artery embolization by isolated use of coils.

hematomas on the last follow-up were 71.4% and 59.2% respectively. Volume of the hematoma decreased by at least 50% in 79.8% of cases. Complete resolution (undetectable hematoma) was achieved in 42.6%. The overall rescue surgery rate was 8.5% (8 cases). Figure 1 demonstrates temporal changes in a cSDH following MMAE treatment in one of the cases treated at our center.

Our center has also been productive in MMAE research, aiming to improve this novel technique and provide evidence-based answers to the gaps in knowledge. In a recent study<sup>10</sup>, morphological features of MMAE-treated cSDHs, including internal architecture and density at the time of treatment were described, and nature, sequence, and timing of changes on follow-up, as well as prognostic significance of these features were explored. The findings provided valuable information regarding natural history of cSDHs and the expected

course of changes during follow-up of upfront MMAE, which can enhance prognostication and patient selection for this recently emerging therapeutic approach. In another study, we demonstrated how isolated use of coils for endovascular treatment of cSDHs can be as effective as particle embolization (Figure 2). This method eliminates the risks of cranial nerve and visual complications associated with MMAE, and potentially reduces the odds of procedural abortion due to presence of dangerous anastomoses, which would drastically reduce this procedure's costs and technical complexity. And finally, our groups also found that distal or proximal or combined proximal and distal MMA embolization do not result in significantly different rates of cSDH resolution and need for rescue surgery, and embolizing more than one branch does not translate into enhanced MMAE therapeutic efficacy. On this basis, a general decision-making algorithm for MMAE technical

planning was suggested, to reduce technical complexity of the procedure and maintain optimal safety and efficacy.

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# Cranial Dural Arteriovenous Fistula in a Young Adult

Max Shutran, MD and Carlos Baccin, MD

Cranial dural arteriovenous fistula (dAVF) is a type of arteriovenous shunt distinct from a brain arteriovenous malformation (AVM). The location of the arteriovenous shunt in a dAVF is within the leaves of the dura mater, often but not always in the wall of a venous sinus, while the shunt in a brain AVM is located within brain parenchyma. Dural AVFs are acquired lesions. Although the pathogenesis is not fully understood, many dAVFs are felt to be a consequence of dural venous sinus thrombosis. They have been associated with hypercoagulable states, including malignancy. Common sites for dAVFs include the junction of the transverse and sigmoid sinuses, the tentorium, the ethmoidal dura, and the superior sagittal sinus. Dural AVFs of the cavernous sinus are also common but are more frequently referred to as indirect carotid-cavernous (CC) fistulas.

The clinical presentation of dAVFs depends on the location of the fistula and its venous drainage pattern. Pulsatile tinnitus is one common presentation, especially for dAVFs located at the transverse sigmoid sinus junction. In such cases, a bruit can often be heard by auscultating over the mastoid with a stethoscope. Dural AVFs of the cavernous sinus often present with orbital symptoms including chemosis (redness of the eye), proptosis, increased intraocular pressure, eye pain, vision loss, and extraocular motion restriction (ophthalmoparesis). Dural AVFs with cortical venous drainage can present with neurological dysfunction due to venous hypertension, including cognitive decline, focal neurological deficit, seizures, and hemorrhage.

The natural history of a dAVF depends on the venous drainage pattern. The two commonly used

grading schemes for dAVFs, the Borden and Cognard classifications, both reflect this<sup>[1][2]</sup>. Lesions with venous drainage into the dural venous sinuses only (Borden grade 1) have a low risk of hemorrhage and in some cases observation may be appropriate. Dural AVFs with drainage into venous sinuses and cortical veins (Borden grade 2) and especially those with exclusively cortical venous drainage (Borden grade 3) are at risk of causing intracranial hemorrhage and treatment is indicated to prevent this. More recent analysis has shown that among patients with cortical venous drainage, patients presenting with neurological symptoms are at the highest risk of future neurological injury compared to those without<sup>[3]</sup>.

At the BIDMC Brain Aneurysm Institute, we recently encountered and treated a patient with a Borden grade 3 dAVF that illustrates the above concepts. A 34-year-old man with a history of alcohol-induced liver cirrhosis presented to an urgent care clinic with several weeks of worsening headaches and was noted to have a dilated right pupil. A head CT and CT angiogram were obtained, which showed dilated vascular structures over the right hemisphere without evidence of acute hemorrhage. On neurological exam, he had slow speech and a dilated and sluggishly reactive right pupil, but no other focal neurological signs. We performed a diagnostic cerebral angiogram which confirmed a dural AVF with extensive cortical venous drainage. The fistula was located in the dura of the right middle cranial fossa, adjacent to the cavernous sinus but without any venous drainage into the cavernous sinus. Instead, the sole venous drainage was into Sylvian veins, causing hypertension of the cortical venous system. Arterial supply was seen from the ophthalmic

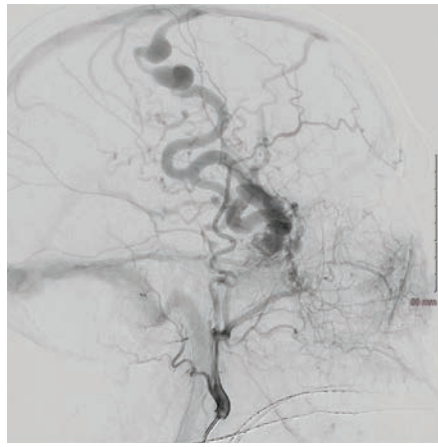
artery, middle meningeal artery, and distal internal maxillary artery. It is worthwhile to re-emphasize that the venous drainage, rather than the arterial supply, dictates the risk and symptomatology of a dAVF, and that any arteries supplying the adjacent dura mater may supply the fistula.

Because the fistula was symptomatic and carried a risk of hemorrhage, we felt that urgent treatment was indicated. Our first attempt was to inject Onyx (a non-adhesive, lava-like embolic agent which solidifies after injection) via the middle meningeal artery, but we were unable to achieve adequate 'penetration' of the fistula (i.e. occlusion of the vein) by this route. Two days later, we injected Onyx through the distal internal maxillary artery and were successfully able to fill the fistula and proximal draining vein with embolic material, curing the fistula. The patient's headaches improved post-procedure and he felt more alert.

The treatment for dAVF is to occlude the arteriovenous shunt on the venous side. This may be done with trans-arterial or trans-venous endovascular approaches, or with surgery. Radiosurgery has also been used as an effective treatment option, although this comes with a one to two year lag time before regression of the fistula can be expected. As mentioned, some fistulas may be also be safely observed. At the BIDMC Brain Aneurysm Institute, all modalities of therapy are considered and available for use. The chosen treatment option depends on the specific anatomy of the fistula. In the case above, there was no readily available trans-venous access to the fistula, so trans-arterial embolization was our first choice. Most dAVFs of the cavernous sinus are treated by trans-venous embolization, as the



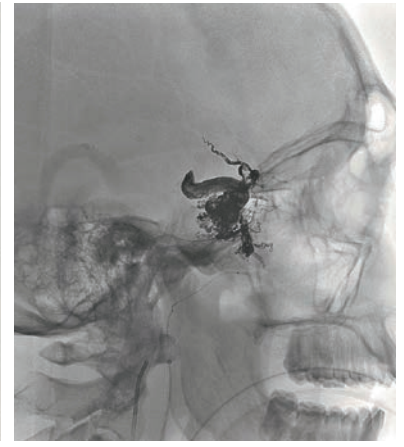
**Figure 1:** CT angiogram of the head showing dilated veins in the right Sylvian fissure.



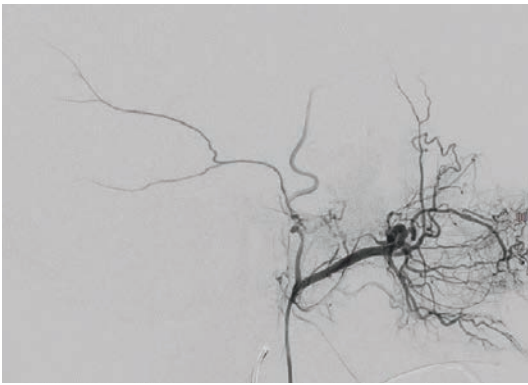
**Figure 2:** Lateral angiogram of the right external carotid artery showing an arterIALIZED and dilated cortical draining vein.



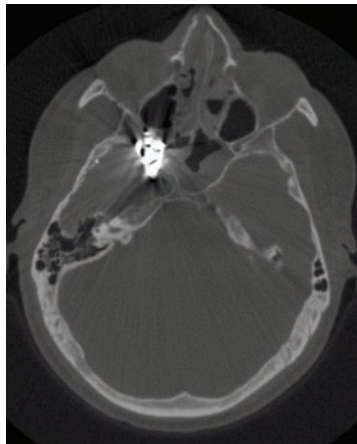
**Figure 3:** A microcatheter is positioned in the distal right internal maxillary artery, preparing to inject Onyx.



**Figure 4:** Onyx casting the proximal aspect of the draining vein and multiple small feeding arteries.



**Figure 5:** Right external carotid artery angiogram showing complete occlusion of the fistula.



**Figure 6:** Post-procedure CT scan showing the location of the embolic material.

efficacy is higher and risk of cranial neuropathy lower than with trans-arterial embolization. Ethmoidal dAVFs are often treated surgically, as the endovascular approaches to these lesions carry higher risk. Treatment strategies for dAVF can become quite complicated and can involve staged approaches or combined approaches, such as trans-arterial plus trans-venous embolization. In some cases, open surgery is combined with endovascular surgery to achieve access to vascular compartments that would be inaccessible through pure endovascular approaches<sup>[4]</sup>. When planning a treatment strategy, it is critical to understand the anatomy of the arteriovenous shunt and its relationship to normal structures to be able to design an effective approach with minimal risk.

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## NEWS

**The BIDMC Brain Aneurysm Institute is proud to announce the appointment of Phil Taussky, MD, FACS to the team.**



He served the Department of Neurosurgery at the University of Utah as the Section Chief of Neurovascular Surgery and Chief Value Officer. He earned his MD degree at the University of Basel. He completed a skull base/cerebrovascular fellowship at the University of Utah and an endovascular

fellowship at the Mayo Clinic, focusing on minimally invasive techniques to treat stroke, aneurysms, AVMs, and other vascular diseases. Because of his dual training, he has a unique perspective, offering his patients both microsurgical and minimally invasive endovascular treatment for vascular disease. He is currently involved in multiple trials further expanding the use of minimal invasive endovascular devices to treat intracranial aneurysms and stroke. Dr. Taussky has published over 120 papers and book chapters and has lectured both nationally and internationally. He has proctored physicians from all over the world in complex flow diversion cases. He provides consultation and management for patients with AVMs, aneurysms, cavernous angiomas, meningiomas, Moya-Moya, stroke, carotid disease, and other cerebral problems.

## SAVE THE DATES

**29<sup>th</sup> Annual Ischemic and Hemorrhagic Update:  
Current Practices and Future Directions  
MONDAY, MAY 9, 2022**

VIRTUAL: <https://cmeregistration.hms.harvard.edu/ischemic2022>

COURSE DIRECTORS: Christopher S. Ogilvy, MD,  
Sandeep Kumar, MD, and Magdy Selim, MD, PhD

This is a unique course focused on recent advances in the field of neurovascular disease including up-to-date theories of carotid disease, cerebral hemorrhage, and brain aneurysms. Topics covered will include assessment, management, and specific issues of carotid disease, cerebral hemorrhage, and brain aneurysms. (7.75 AMA PRA Category 1 Credits™)

For information about registration, contact Deidre Buckley, NP, at [dabuckle@bidmc.harvard.edu](mailto:dabuckle@bidmc.harvard.edu).

**21<sup>st</sup> Annual Arterial Challenge: 5K Run/2.5K Walk  
SUNDAY, APRIL 24, 2022**

9:30 am – 12:30 pm

Castle Island, 2010 William J Day Boulevard, Boston, MA

For more information, contact Brain Aneurysm Foundation, at [office@bafound.org](mailto:office@bafound.org) or call 888-272-4602.