Carotid-cavernous fistulas are acquired abnormal communications between the cavernous sinus and the carotid artery or its branches. Carotid-cavernous fistulas are divided into 4 categories according to the Barrow classification. Type A CCF is a direct shunt between the cavernous sinus and the ICA, whereas Types B, C, and D are indirect dural fistulas with arterial feeders from the ICA, ECA, or both, respectively. Endovascular therapy has become the mainstay of treatment for CCFs. Given the direct nature of the shunt in Type A CCFs, transarterial embolization of the cavernous sinus through the fistulous site is the recommended treatment. For indirect fistulas, however, the transvenous approach is the preferred modality given the size and number of involved arterial feeders, as well as the difficulties and potential hazards associated with transarterial embolization. In a transvenous approach, the inferior petrosal sinus is the easiest and shortest path to the cavernous sinus. Access can also be obtained through the facial vein-SOV route. When all transarterial and transvenous options have been exhausted, direct operative cannulation of the SOV provides a direct, reasonable, and safe route to the cavernous sinus. Liquid embolic agents—mainly cyanoacrylate—served only as an adjunct to coils to achieve better obliteration of the fistula.

Object. Endovascular therapy is the primary treatment option for carotid-cavernous fistulas (CCFs). Operative cannulation of the superior ophthalmic vein (SOV) provides a reasonable alternative route to the cavernous sinus when all transvenous and transarterial approaches have been unsuccessful. The role of the liquid embolic agent Onyx in the management of CCFs has not been well documented, especially when using an SOV approach. The purpose of this study is to assess the safety and efficacy of Onyx embolization of CCFs through a surgical cannulation of the SOV.

Methods. The authors retrospectively reviewed all patients with CCFs who were treated with Onyx through an SOV approach between April 2009 and April 2011. Traditional endovascular approaches had failed in all patients.

Results. A total of 10 patients were identified, 1 with a Type A CCF, 5 with a Type B CCF, and 4 with a Type D CCF. All fistulas were embolized in 1 session. Onyx was the sole embolic agent used in 7 cases and was combined with coils in 3 other cases. Complete obliteration was achieved in 8 patients and a significant reduction in fistulous flow was achieved in 2 patients, which later progressed to near-complete occlusion on angiographic follow-up. All patients experienced a complete clinical recovery with excellent cosmetic results and were free from recurrence at their latest clinical follow-up evaluations.

Conclusions. Onyx embolization is an excellent therapy for CCFs in general, and through an SOV approach in particular. Direct operative cannulation of the SOV followed by Onyx embolization may be the best treatment option in patients with CCFs when all other endovascular approaches have been exhausted.

Key Words • carotid-cavernous fistula • embolization • Onyx • superior ophthalmic vein

Abbreviations used in this paper: CCF = carotid-cavernous fistula; ECA = external carotid artery; ICA = internal carotid artery; IOP = intraocular pressure; SOV = superior ophthalmic vein.
Recently, the introduction of Onyx (ev3) has added an important element to the endovascular armamentarium and revolutionized the treatment of intracranial fistulas. Onyx offers several advantages over N-butyl cyanoacrylate or coil embolization that allow safer and better treatment of intracranial arteriovenous malformations and fistulas.\(^1\),\(^4\),\(^26\) However, the role of Onyx in the endovascular management of CCFs has yet to be defined, as studies have been limited to case reports and small patient series.\(^9\),\(^27\) Furthermore, the effectiveness of Onyx embolization of CCFs through a transorbital approach has not been investigated. In the present paper, we assess the safety and efficacy of Onyx embolization of CCFs through the SOV approach in a series of 10 patients treated at our institution.

**Methods**

**Study Sample**

Between April 2009 and April 2011, 10 patients were treated at the Jefferson Hospital for Neuroscience for CCFs through a direct SOV approach using Onyx. In all patients, treatment of their CCFs had failed using traditional transvenous and transarterial access. Medical charts and angiograms were retrospectively reviewed to determine patient characteristics, type of fistula (based on the Barrow classification\(^2\)), type of embolic material used, degree of fistula occlusion after embolization, procedural complications, clinical outcome, and angiographic follow-up.

**Operative Technique**

All procedures were performed in the angiography suite under general anesthesia and continuous electrophysiologic monitoring. After femoral access is obtained and a catheter placed in the carotid artery, the orbital cutdown to expose the SOV is performed by a neuroophthalmologist and a neurosurgeon. The orbicularis oculi muscle and the orbital septum are opened following a small upper eyelid crease incision. With meticulous dissection into the superomedial orbit, the arterialized SOV is identified and subsequently catheterized with an 18-gauge angiocatheter connected to a rotatory hemostatic valve. Under roadmap guidance, an Echelon microcatheter (ev3) is advanced into the cavernous sinus. Under full heparinization with 100 units/kg, the cavernous sinus is embolized using Onyx 18 or Onyx 34, depending on flow intensity across the fistula. The forward penetration of the material is closely monitored to detect and prevent any migration into an inappropriate area, in which case the injection is momentarily interrupted to allow polymerization of the Onyx cast. Extreme care is taken to maintain the patency of at least 1 of the ophthalmic veins (the superior or the inferior ophthalmic vein), because embolization of both veins leads to a transient dramatic increase in the IOP. Coils are used only for high-flow fistulas to provide a framework for Onyx, thus minimizing the risk of distal embolic material migration.

Once embolization is complete, cerebral angiograms from the arterial supply are performed to confirm obliteration of the fistula. At that time, attention is redirected to the orbit. After removal of the access catheter, the SOV is ligated distal to the puncture site. With bipolar cautery, a meticulous hemostasis is achieved, and the orbital incision is subsequently closed. Follow-up angiography was typically obtained in patients who showed any clinical suspicion of recurrence of the fistula. Clinical follow-up evaluations were scheduled at 3 and 6 months after surgery for all patients.

**Results**

The mean patient age in the series was 65 years, with a range of 36–78 years. Eight patients were women and 2 were men. The mean follow-up duration was 5.5 months (range 3–12 months). All patients had intractable orbital symptoms (Table 1). One patient had a Type A CCF, 5 had a Type B, and 4 had a Type D fistula. All patients showed some degree of dilation of the SOV, and 4 patients had CCFs that displayed bilateral arterial supply. In all these patients, multiple attempts to catheterize the cavernous sinus through a transarterial or a transvenous approach had failed.

All CCFs were obliterated in 1 session. Onyx (18 or 34) was the sole embolic agent used in 7 cases (70%). Coils were deployed in addition to Onyx in 3 patients with high-flow fistulas to achieve better obliteration and decrease the risk of distal Onyx migration (Table 2). Complete obliteration of the CCF was achieved in 8 (80%) of 10 patients. Complete occlusion of the fistula was achieved in all 7 cases in which Onyx was used as the sole embolic agent. In 2 patients, high-flow CCFs were incompletely occluded after embolization but showed significant flow reduction (80%) through the fistula. Follow-up angiography in these 2 patients demonstrated further thrombosis and near-complete obliteration of the fistula. In 9 (90%) of 10 patients, no complications were noted. One patient developed high IOPs in his left eye postoperatively and was noted to have an afferent pupillary defect. This defect was attributed to a probable treatment-induced thrombosis of the superior and inferior ophthalmic veins. Because the patient continued to experience high uncontrolled IOP despite maximum medical therapy, a cantholysis had to be performed to relieve orbital pressure. He subsequently recovered well and was discharged on postoperative Day 3. All 10 patients in the series had a rapid clinical recovery and demonstrated complete reversal of their symptoms. No signs of recurrence were noted at the latest clinical follow-up evaluation in all 10 patients. Excellent cosmetic results were achieved as well.

**Illustrative Case**

This 60-year-old woman (Case 1) presented to our institution with a 3-week history of worsening left-sided chemosis, proptosis, and diplopia. On examination, she was found to have a sixth cranial nerve palsy with an elevated IOP (28 mm Hg) in the left eye. A digital subtraction angiogram revealed a left-sided Type B CCF supplied by meningeal branches of the left and right ICA with retrograde venous flow in the SOV and cortical veins (Fig. 1). Inferior
petrosal sinuses were noted to be occluded bilaterally, and multiple attempts to catheterize the fistula through the facial venous system were unsuccessful. The decision was made to perform a surgical cutdown to expose the SOV. The vein was isolated and subsequently catheterized with an 18-gauge angiocatheter (Fig. 2). An Echelon-10 microcatheter was navigated into the cavernous sinus, and the fistula was subsequently embolized with 1.6 ml of Onyx 18 (Fig. 3). Control angiography of the left and right ICAs confirmed complete obliteration of the fistula with no evidence of distal embolic material migration (Fig. 4A and B). The patient’s postoperative course was uneventful and by the time of discharge, her IOPs had decreased to 17 mm Hg. She displayed complete reversal of her symptoms at the 3- and 6-month clinical assessments and maintained complete occlusion of her fistula at the 3-month follow-up angiography (Fig. 4C and D).

Discussion

Carotid-cavernous fistulas usually present with ocular symptoms such as chemosis, proptosis, ocular bruist, loss of visual acuity, and diplopia. Because 20%–50% of CCFs close spontaneously, treatment is indicated only in the presence of visual loss, elevated IOP despite optimal medical therapy, refractory ocular symptoms, and cortical venous drainage (given the potential risk of intracranial bleeding). Access to the cavernous sinus can be obtained through the ipsilateral and contralateral inferior petrosal sinus. Alternatively, the cavernous sinus can be approached retrograde via the facial vein, to the angular vein and to the SOV. This approach is rarely successful, however, due to the tortuosity of the facial venous system. When traditional transarterial and transvenous endovascular approaches have failed, surgical exposure of the SOV is an acceptable alternative. In the presence of a neuroophthalmologist and an experienced neuroendovascular team, this approach is almost always successful in embolizing CCFs. Some investigators have even described the direct SOV approach as the best initial treatment for all CCFs. Potential complications of this approach include infection, damage to orbital structures, orbital hematoma, and neovascular glaucoma (if the SOV is ligated) with subsequent loss of vision. Thus far, the majority of reported cases of CCFs that were treated through an SOV approach were embolized using coils or detachable balloons. Quiñones et al. rewrote

<table>
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<tr>
<th>Case No.</th>
<th>Type of Fistula</th>
<th>Embolic Material</th>
<th>Initial Angiographic Occlusion</th>
<th>Complications</th>
<th>Follow-Up Angiographic Occlusion</th>
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<tr>
<td>1</td>
<td>B</td>
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<td>complete</td>
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<tr>
<td>2</td>
<td>D</td>
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<td>not performed</td>
</tr>
<tr>
<td>4</td>
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<td>intractable high IOP</td>
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</tr>
<tr>
<td>6</td>
<td>A</td>
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<td>none</td>
<td>not performed</td>
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<tr>
<td>7</td>
<td>D</td>
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<tr>
<td>8</td>
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<td>9</td>
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<td>Onyx 18</td>
<td>complete</td>
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<td>D</td>
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<td>partial (80%)</td>
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<td>near-complete</td>
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* CN = cranial nerve; DVA = decreased visual acuity.
ported complete angiographic obliteration with coil embolization in 12 of 13 patients with CCFs, with complications developing in 2 patients. Derang et al.\(^7\) successfully treated 21 patients with balloons and coils through direct operative cannulation of the SOV and noted new cranial nerve deficits in 2 cases.

We used Onyx through am SOV approach for the treatment of 10 patients with various types of CCFs. Onyx was the sole embolic agent used in 7 cases and was combined with coils in 3 other cases. We were able to achieve adequate obliteration of fistulas and reversal of symptoms in all patients with a very low rate of complications. Interestingly, all 7 cases that were embolized solely with Onyx were completely obliterated at the end of the procedure. It appears that Onyx is an excellent agent for embolizing CCFs even when used as a monotherapy (without coils). Onyx offers the advantage of filling the interstices of the cavernous sinus along with small delicate fistulous channels that would otherwise be left patent with coil embolization. Due to its lava-like flow pattern and its nonadhesive nature, Onyx allows longer, slower, and more controlled injections with better penetration of the fistula. Onyx embolization through the SOV might well be the best available option in patients failing more traditional endovascular approaches. Impressive cosmetic results were obtained as well, given that a small incision was made within the natural crease of the upper eyelid.

There are very little data regarding the use of Onyx to treat CCFs. Elhammady et al.\(^9\) treated 12 patients using Onyx, 8 via a transvenous route and 4 via a transarterial route. The authors were able to obliterate all CCFs and reported 3 new cranial nerve palsies following the procedure. Lv et al.\(^18\) reported complete angiographic obliteration in 3 of 5 patients with CCFs following Onyx embolization, with cranial nerve palsies developing in 2 cases. In their series, Zaidat et al.\(^32\) used Onyx in combination with coils or stents in 5 cases and achieved complete occlusion in all 5 cases. Given that traditional endovascular approaches were used in these studies, no direct comparison can be made with the findings of the present series. Nevertheless, the results of these studies are consistent with our findings and underline the high success rate of Onyx in the treatment of CCFs, regardless of approach. However, we could not confirm that Onyx embolization is associated with a substantial risk of new cranial nerve palsies as no such complications were noted in the present series. Investigators have postulated that Onyx induces thrombosis and swelling in the cavernous sinus, which could lead to compression of surrounding nerves.\(^9\) This type of complication has been previously described with coils that can exert mass effect in the cavernous sinus, leading to cranial nerve paralysis.\(^7,21,22,24\)

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**Fig. 1.** Case 1. Frontal (left) and lateral (right) views of digital subtraction angiography of the left ICA demonstrating a left-sided indirect CCF (Barrow Type B).

**Fig. 2.** Case 1. Intraoperative picture of the transorbital approach, showing the angiographic setup with an 18-gauge angiocatheter connected to a rotatory hemostatic valve.

**Fig. 3.** Case 1. Fluoroscopic image of the left cavernous sinus demonstrating the Onyx cast.
Some authors have described treatment of CCFs through transorbital punctures of the SOV or the cavernous sinus. In a recent article, Dashti et al. successfully treated 2 patients with indirect CCFs through a direct transorbital puncture of the superior and inferior ophthalmic veins. The authors concluded that their technique represents a reasonable alternative to cutdown procedures in most cases. We believe that a direct puncture of the SOV is an unnecessarily hazardous procedure that carries a high risk of damage to the cranial nerves, ICA, orbit structures, and trochlea. Furthermore, given the arterialized nature of the SOV, a direct puncture of the vessel could lead to a disastrous retroorbital hematoma with the risk of vision loss. While experience with transorbital punctures has been limited to case reports (and 1 series of 8 patients), surgical exposure of the SOV is an established procedure that is straightforward, safe, cosmetically appealing, and almost always reliable.

Conclusions

In this paper we present the results of the first case series that assesses the safety and efficacy of Onyx embolization of CCFs through a surgical cutdown. We were able to achieve excellent clinical and angiographic outcomes with a low rate of complications for various types of CCFs. Onyx can also be used efficiently as a monotherapy to embolize CCFs. When attempts to catheterize the cavernous sinus through transvenous and transarterial approaches have failed, surgical exposure of the SOV followed by Onyx embolization (in combination with coils when necessary) is safe and effective and should be considered the preferred treatment modality.

Disclosure

Dr. Tjoumakaris serves as a consultant to Stryker Neurovascular. Author contributions to the study and manuscript preparation

Fig. 4. Case 1. Digital subtraction angiograms of the left ICA. Frontal (A) and lateral (B) views show complete obliteration of the fistula following embolization with Onyx 18. Frontal (C) and lateral (D) projections at the 3-month follow-up confirm complete occlusion of the fistula.

Fig. 5. Artist’s illustrations of the procedure. Left: The incision in the upper eyelid crease is shown, as well as the exposure of the dilated SOV in the superomedial quadrant of the orbit. Right: Illustration showing the relatively straight course of the SOV (arrow) draining into the cavernous sinus. The microcatheter is introduced through the SOV (A) and positioned in the cavernous sinus. Onyx is slowly injected into the cavernous sinus (B) to achieve obliteration of the fistula.
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