Endovascular management of fusiform aneurysms in the posterior circulation: the era of flow diversion

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Fusiform aneurysms are uncommon compared with their saccular counterparts, yet they remain very challenging to treat and are associated with high rates of rebleeding and morbidity. Lack of a true aneurysm neck renders simple clip reconstruction or coil embolization usually impossible, and more advanced techniques are required, including bypass, stent-assisted coiling, and, more recently, flow diversion. In this article, the authors review posterior circulation fusiform aneurysms, including pathogenesis, natural history, and endovascular treatment, including the role of flow diversion. In addition, the authors propose an algorithm for treatment based on their practice.

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FUSIFORM aneurysms are defined as circumferential dilations of an intracranial artery without an ostium or neck.¹² They are commonly located in the posterior circulation, especially the vertebral artery (VA), basilar artery (BA), and posterior cerebral artery (PCA).¹¹ Fusiform aneurysms are uncommon compared with their saccular counterparts, yet they remain very challenging to treat. The first case of a vertebral fusiform aneurysm was described by Wells in 1922,³⁵ and since then several terms have also been used, including dolichoectatic aneurysm, transitional aneurysm, and giant serpentine aneurysm.

Posterior circulation fusiform aneurysms (PCFAs) have a significant male predominance (approximately 70%) and most commonly present as posterior circulation ischemic stroke.^{12,31} In addition, they may cause cranial nerve palsies, brainstem compression, and subarachnoid hemorrhage (SAH). Contrary to the more common saccular aneurysms, fusiform aneurysms are associated with high rates of rebleeding and morbidity.

In this article, we review PCFAs, including pathogenesis, natural history, cerebrovascular surgical treatment, and endovascular treatment, including the role of flow diversion. In addition, we propose an algorithm for treatment based on our practice.

Pathogenesis and Natural History

Fusiform aneurysms may occur due to a variety of underlying pathologies affecting the wall of the blood vessel. The most common proposed causes are dissection and atherosclerosis.^{6,7,30} Our understanding of the natural history of PCFAs is very limited and largely depends on the presenting signs and symptoms. Symptomatic patients have a poor natural history if they do not undergo treatment, especially if they present with brainstem ischemia or compression. In patients with ruptured aneurysms, the rebleeding rate is high and ranges between 30% and 85%.^{1,23} The mortality rate is also high for untreated ruptured aneurysms. In a study that evaluated conservative management in ruptured PCFAs, the mortality rate was 38% after a mean follow-up period of 18 months.¹⁰

In a prospective study of vertebrobasilar aneurysms over a 12-year period at the Mayo Clinic, the annual rupture rate of fusiform aneurysms was 2.3%.^{13,21} The initial diameter of an aneurysm is a significant predictor of lesion rupture. The authors also found that an initial diameter larger than 10 mm in fusiform aneurysms was a significant risk factor for aneurysm enlargement and future rupture. The mortality rate was approximately 6 times higher in

ABBREVIATIONS AICA = anterior inferior cerebellar artery; BA = basilar artery; EC-IC = extracranial-intracranial; IEL = internal elastic lamina; MRA = MR angiography; PCA = posterior cerebral artery; PCFA = posterior circulation fusiform aneurysm; PED = Pipeline embolization device; PICA = posterior inferior cerebellar artery; SAC = stent-assisted coiling; SAH = subarachnoid hemorrhage; SCA = superior cerebellar artery; STA = superficial temporal artery; VA = vertebral artery.

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patients with aneurysm growth than in those with no enlargement.²¹

Therefore, based on observations of the natural history, the vast majority of ruptured PCFAs should be treated. Additionally, unruptured PCFAs larger than 10 mm also likely warrant treatment.

Classification

There are 2 widely accepted classification systems for nonsaccular aneurysms, including the fusiform type, that stratify patients into risk groups. Flemming's classification for nonsaccular vertebrobasilar circulation aneurysms is based on radiographic appearance.¹² Lesions are defined as having an arterial dilation greater than 1.5 times the normal diameter without any neck (Huber's definition), and the types are as follows: A) fusiform (14%), aneurysmal dilation of the vessel without an identifiable neck involving a portion of arterial segment; B) dolichoectasia (45%), uniform dilation involving the entire artery with any degree of tortuosity; C) transitional (19%), uniform aneurysmal dilation of the artery with superimposed dilation of a portion of the involved arterial segment; and the indeterminate type (20%). Fusiform and transitional types are most likely to be symptomatic, while the dolichoectatic type has a more benign nature. Acute dissecting aneurysms were excluded because of the known distinctive behavior.

The other classification system is that of Mizutani et al. and consists of 4 types based on histopathology.²⁴ Type I, classic dissecting aneurysm characterized by widespread disruption of the internal elastic lamina (IEL) without intimal thickening. This type typically presents with SAH and high rates of rebleeding. Type II, segmental ectasia, with a more benign clinical course than Type I. This type is characterized by extended and/or fragmented IEL with intimal thickening. In addition, the luminal surface is smooth without thrombus formation. Type III, dolichoectatic dissecting aneurysm. This type is distinguished pathologically from Type II by dissections in the thickened intima and organized luminal thrombus. Most Type III aneurysms are symptomatic, grow over time, and are frequently associated with hemorrhage and a mortality rate of 50%. Lastly, Type IV is saccular aneurysm characterized by minimally disrupted IEL without intimal thickening and is associated with a high risk of rupture.

Treatment

Choice of Treatment

All fusiform aneurysms have been historically treated with different open surgical treatment modalities, including Hunterian ligation, trapping, surgical bypass, and clip reconstruction techniques.³¹ However, endovascular therapy has emerged as the primary treatment modality for PCFAs over the past decade. Recently, endovascular treatments have been successfully used in treating PCFAs with good outcomes. In fact, microsurgical treatment is generally reserved for cases that cannot be treated with endovascular therapy. The endovascular options include parent vessel coil occlusion, stenting alone, stent-assisted coiling (SAC), and flow-diverting stents (Fig. 1).

Microsurgical Management

Open surgical treatment of PCFAs is becoming a less popular option given the recent advancements in endovascular therapy. Microsurgical treatment modalities often involve flow reduction or bypass/trapping in cases of poor collateral supply, flow reversal in cases of adequate collateral supply, or trapping with aneurysm decompression for lesions with mass effect. Since fusiform aneurysms do not have a true neck, they are usually not amenable to clip reconstruction techniques. Additionally, it is not uncommon for PCFAs to be partially calcified and/or thrombosed, further complicating the open surgical approach. Hence, trapping with or without bypass is considered the main microvascular modality.

Drake and colleagues published extensively on their operative experience with fusiform aneurysms in the posterior circulation.^{8,9,33} The authors used different modalities based on patient presentation, clinical status, and collateral supply. Outcomes were almost comparable between the different modalities, with approximately 70% of treated patients achieving good to excellent outcomes.

Kalani et al.¹⁶ reported the most recent experience at the Barrow Neurological Institute with giant aneurysms in the posterior circulation. The 12-aneurysm cohort included 8 fusiform aneurysms. The primary treatment modality was extracranial-intracranial (EC-IC) bypass. Superficial temporal artery–superior cerebellar artery (STA-SCA) bypasses were performed in 7 cases and STA-PCA was performed in 1 case. Flow was reserved or reduced by complete ($n = 6$) or partial ($n = 1$) occlusion of the BA, or by occlusion of the VA distal to the posterior inferior cerebellar artery (PICA) ($n = 1$). Recurrence and complications were high and the mortality rate among fusiform aneurysms was approximately 40%. The authors did admit that despite their aggressive surgical approach, the long-term outcome was poor for most patients.

More recently, Lawton et al. published an evolved technique of surgical bypass for treating fusiform aneurysms in the basilar trunk.¹⁹ The study included 37 patients, and the bypass evolved in 3 distinct phases, each with different hemodynamic alternations. Surgical bypasses consisted of EC-IC (STA-SCA and STA-PCA) bypasses in Phase 1 for flow reversal, IC-IC (VA-SCA) bypasses in Phase 2 for flow reduction, and Phase 3 (middle cerebral artery–PCA) for distal occlusion. Phase 1 led to extensive flow reduction that prompted BA thrombosis and was associated with 100% mortality. On the other hand, Phase 2 was safer (67% mortality rate) but did not prevent aneurysm growth or progression of symptoms. As a result, the authors revised their technique to distal occlusion, achieving an improved surgical outcome and aneurysm stabilization with a better mortality rate (62%). However, this technique reduced the flow to brainstem perforators causing ischemic damage, despite treatment with antiplatelet agents.

Endovascular Management

The lack of a true aneurysm neck usually makes simple coil embolization impossible and more advanced techniques are required, including SAC and, more recently, flow diversion (Figs. 2 and 3). Parent vessel occlusion is a

FIG. 1. Suggested treatment algorithm for fusiform aneurysms in the posterior circulation based on presentation. FD = flow diverter; PVO = parent vessel occlusion.

reasonable option for nondominant VA aneurysms as well as distal aneurysms of the nondominant PICA, anterior inferior cerebellar artery (AICA), or SCA.

Higashida et al.¹⁵ reported the first case of SAC in PCFAs. They used an intravascular stent in conjunction with Guglielmi detachable coils in a ruptured fusiform aneurysm in the BA. Since then, several case series have been published regarding the use of SAC in the treatment of PCFAs.^{6,29,34} Interestingly, it appears that certain locations of fusiform aneurysms in the PCA have distinct entities with different outcomes. In the 14-year Stanford experience of treating PCFAs,⁶ Steinberg et al.³³ reported that PCA aneurysms had the best outcome (90%), followed by VA and PICA aneurysms (60%); aneurysms located in the BA and vertebrobasilar junction had the worst outcome (39%).

Initial experience with flow diversion in PCFAs was mixed with poor outcomes.³² However, when used in carefully selected patients, PCFAs have been recently reported to have a good to excellent outcome in the majority of those patients. Table 1 summarizes large series (> 5 cases) on the use of flow diverters for PCFAs.

Byrne and colleagues published their initial experience

with Silk (Balt Extrusion) flow diverters in the treatment of intracranial aneurysms in a multicenter prospective study.⁴ Of 70 patients, there were 11 cases of PCFAs. Of these patients, 2 patients died, and there were 2 device-related complications. There were no available data on postoperative angiographic obliteration.

In 2014, both Thomas Jefferson University and Rush University groups published their experience with the Pipeline embolization device (PED; Medtronic) for fusiform aneurysms in the posterior circulation.^{25,26} These reports included 7 and 12 cases of PCFAs, respectively. The device-related complication rate was approximately 25%, and 70%–90% of patients had good to excellent neurological outcome. During angiographic follow-up, complete occlusion varied significantly between the 2 studies, ranging between 30%²⁵ and 75%.²⁶

Contrary to their initial experience with poor outcomes,³² a team from the University at Buffalo reported excellent results in their latest experience with PED for PCFAs.²⁷ Eleven (90%) of 12 patients recovered to a modified Rankin Scale score of 0 or 1 after a clinical follow-up duration of 22 months, with only 1 patient experiencing a perforator territory infarction with poor clinical outcome

FIG. 2. Angiograms obtained in a 46-year-old woman with a good-grade SAH. **A and B:** Initial digital subtraction (DS) angiograms (right VA injection [A] and left VA injection [B]) demonstrating bilateral fusiform VA aneurysms, of which the right-sided aneurysm appeared irregular and the likely source of SAH. **C and D:** The patient was treated acutely with coil occlusion of the right VA (right vertebral injection, unsubtracted [C] and subtracted [D] views). Given that the right VA aneurysm was the likely source of SAH, the patient was acutely allowed to recover from her SAH. **E–G:** The fusiform left VA aneurysm (left VA DS angiogram lateral view [E]) was treated with flow diversion a few weeks later (left VA injection, unsubtracted [F] and subtracted [G] views). The patient will undergo delayed angiography to evaluate for aneurysm occlusion and vessel remodeling.

(modified Rankin Scale Score 4). At last follow-up, the complete occlusion rate was 100%, and the PEDs were patent. The authors attributed the dramatic improvement in outcomes to careful patient selection. All patients presented early, and none had evidence of stroke on MRI before treatment. The second factor was the strict dual antiplatelet regimen with confirmation of the therapeutic effect of antiplatelet therapy by using response testing before flow diversion. Technically, the authors used fewer but longer (35 mm) PEDs compared with more and shorter (20 mm) devices in their initial report. In addition, the new experience included adjunctive coiling, which might reduce stent prolapse by acting as a scaffold.

More recently, Bhogal and his colleagues from Germany published the largest series of flow diversion in PCFAs.² Of the 56 patients with nonsaccular aneurysms, there were 24 fusiform aneurysms. The study used 2 types of flow-diverter devices: PED and p64 flow modulation device (Phenox). The mortality rate was low, with only 1 death (4%). The complete aneurysm occlusion rate was 75% with minor residual filling seen in 12.5% of cases and an unchanged appearance in 1 patient (4%). In the 4 patients without angiographic occlusion, the aneurysm de-

creased in maximum diameter, with increased intraaneurysmal thrombus in 3 cases (75%) on MRI follow-up.

The Fate of Covered Branch Vessels With Flow Diverters

The location of aneurysms in the distal VA (V_4) and vertebrobasilar junction in relation to the branch vessels, especially PICA and AICA, often requires covering the arterial ostium, theoretically increasing the risk of branch vessel occlusion and infarction. A meta-analysis published in 2013 showed that the rate of perforator infarction is 3% with significantly higher odds in posterior circulation aneurysms.³

Initial experiences with flow diverters have shown mixed results regarding the fate of covered branch vessels, ranging between complete patency on all follow-up studies to immediate occlusion after flow-diverter deployment or shortly after.^{5,17,18,28,36} However, recent experiences reported a 0% rate of branch occlusion in the posterior circulation on immediate or follow-up angiography.^{14,20,22} Mazur and colleagues' series specifically reported the patency of PICA and aneurysm occlusion on angiography.²² This series of 11 aneurysms located predominantly in the

FIG. 3. Angiograms obtained in a 57-year-old man who presented with poor-grade SAH. **A:** Initial DS angiogram (left VA injection) demonstrated a single fusiform aneurysm involving the left VA and encompassing the origin of the PICA. **B:** Given the nature of the aneurysm, flow diversion was thought to be the only treatment option that could exclude the aneurysm. Therefore, the patient received dual antiplatelet therapy (aspirin and Plavix), and a flow diverter was placed in the left VA (left VA injection, unsubtracted view). The patient required ventriculoperitoneal shunting, for which Plavix was discontinued. **C:** Delayed DS angiogram demonstrated complete aneurysm occlusion and arterial remodeling (left VA injection).

VA included fusiform aneurysms (80%). The flow diverter spanned the PICA ostium in all cases, with 1 patient experiencing an occluded PICA and in-stent stenosis on immediate angiography. The in-stent stenosis was resolved after abciximab administration, and the covered PICA was noted to have recanalized on follow-up imaging 6 months later. Follow-up angiography was reported in 8 patients (the remaining 3 cases are awaiting follow-up) and demonstrated thrombosis of the aneurysm with patency of the PICA in all of them.

Clinical and Radiographic Follow-Up

Our practice protocol involves a clinical follow-up at 1 month, 3–6 months, and 12–18 months. We find that, in general, these time periods end up synching well with the stages of a patient's recovery and clinical progress. Additionally, they coincide with our imaging follow-up. Depending on the case and symptoms, later follow-up can be scheduled at 1- to 3-year intervals. Our imaging follow-up protocol consists of immediate postoperative control conventional angiography, then at 3–6 months and another session at 12–18 months. We also recommend MR angiography (MRA) at 12–18 months after treatment, and every 1–3

years subsequently, depending on the degree of aneurysm obliteration. We have settled on this follow-up paradigm based on both of the most common practices reported in current literature and from discussion with colleagues around the globe. We feel that, in a stable aneurysm, MRA is an adequate surrogate for conventional angiography, hence our switching to MRA after the 12- to 18-month follow-up angiography. However, the limitations of resolution of MRA make us feel that gold-standard angiography with maximal detail and resolution is still worthwhile for most patients during the first 12–18 months, although this is certainly debatable and, in high-risk patients, we switch to MRA follow-up sooner. Patients undergoing SAC and flow-diverter placement are kept on a strict regimen of pre- and postoperative antiplatelet therapy, and dual therapy is maintained at least until the 3- to 6-month angiogram, after which aspirin is continued for life.

Conclusions

Given the evolving endovascular technologies over the last 2 decades in addition to high rates of complications and mortality associated with open surgery, endovascular therapy should be considered as the primary treatment

TABLE 1. Summary of large series (> 5 patients) on the use of flow diverters for PCFAs

Authors & Year	No. of PCFAs	Flow Diverter Device	No. of Device-Related Complications	No. of Deaths	No. w/ Complete Obliteration on Angiography
Byrne et al., 2010	11	Silk	2	2	NA
Siddiqui et al., 2012	7	PED (6), Silk (1)	6	4	2
Monteith et al., 2014	7	PED	2	1	2
Munich et al., 2014	12	PED	3	1	9 (75%)*
Natarajan et al., 2016	12	PED	1	0	12 (100%)
Bhogal et al., 2017	24	PED, p64	NA	1	18 (75%)

NA = not available.

* Data were not available for 2 patients. Of the 10 patients who underwent angiography, 9 patients (90%) had complete occlusion.

modality for PCFAs. For aneurysms that are not treatable by endovascular methods, microsurgical treatment should be considered. Flow diversion is a new endovascular method and can achieve excellent outcomes in carefully selected patients with PCFAs.

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Disclosures

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Author Contributions

Conception and design: Mocco, Awad, Mascitelli. Acquisition of data: Awad. Drafting the article: Awad, Mascitelli, Haroun. Critically revising the article: all authors.

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