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# Bypass for the Treatment of Vasculitis Affecting the Central Nervous System

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## 1. Introduction

Vasculitis affecting the central nervous system (CNS) comprises a large group of diseases characterized by inflammation of arteries that narrows the arterial lumen, compromises cerebral blood flow, and causes a variety of ischemic symptoms. These diseases share similar pathology, but differ in their ages of onset and involved arteries (Begelman & Olin, 2000; Sheikhzadeh et al., 2002; Weyand & Goronzy, 2003). Large-vessel vasculitides are characterized by inflammation of the aorta and the carotid arteries. Giant-cell arteritis, for example, affects patients older than 50 years, while Takayasu's arteritis affects women in their second and third decades of life. With giant-cell arteritis, incidences range from 15 to 25 cases per 100,000 with a predilection for people of northern European descent; Takayasu's arteritis is most commonly seen in Japan, Southeast Asia, India, and Mexico and has an annual incidence in North America of 2.6 per million people. Large-vessel vasculitides in the pediatric population are rare (Gardner-Medwin et al., 2002). Nevertheless, most childhood vasculitides, such as Henoch-Schonlein purpura, Kawasaki disease, polyarteritis nodosa, Wegener granulomatosis, and Behcet disease, are medium- and small-vessel syndromes that often present pathognomonically and rarely, if ever, lead to aortitis or large-vessel obliterative arteriopathy.

Management of vasculitis is almost always medical and individualized to the presenting symptoms, involving a combination of corticosteroids, cytotoxic immunosuppressive agents (intended to decrease inflammation and prevent arterial occlusion) and antibiotics in infectious causes. In the later stages of these diseases, cerebral ischemia may result from chronic scarring or occlusion of arteries rather than acute inflammation. Therefore, revascularization is sometimes necessary to prevent cerebral ischemia and infarction, and bypass surgery has an important role in the treatment of selected cases of vasculitis outside of the acute inflammatory phase.

The aforementioned variable pattern of involvement of different vasculitides can affect conventional donor and recipient sites for common extracranial-intracranial bypasses. Consequently, bypasses for vasculitis often require unusual and complex reconstructions that are not part of the standard bypass repertoire. In this chapter, we review the special techniques and nuances of bypass for vasculitis patients and we demonstrate our experience with case examples.

## 2. Ilustrative cases

**Case 1**

A previously healthy 10-month old boy presented with sudden right-sided hemiparesis. Magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) demonstrated acute ischemia in the left middle cerebral artery territory. Cerebral angiography demonstrated severe obliterative arteriopathy of the great vessels with complete occlusion of all vessels originating from the aortic arch and reconstitution of the cerebral perfusion through unique collateralization patterns. The cerebral vasculature appeared to be structurally unaffected by the underlying vascular pathology.
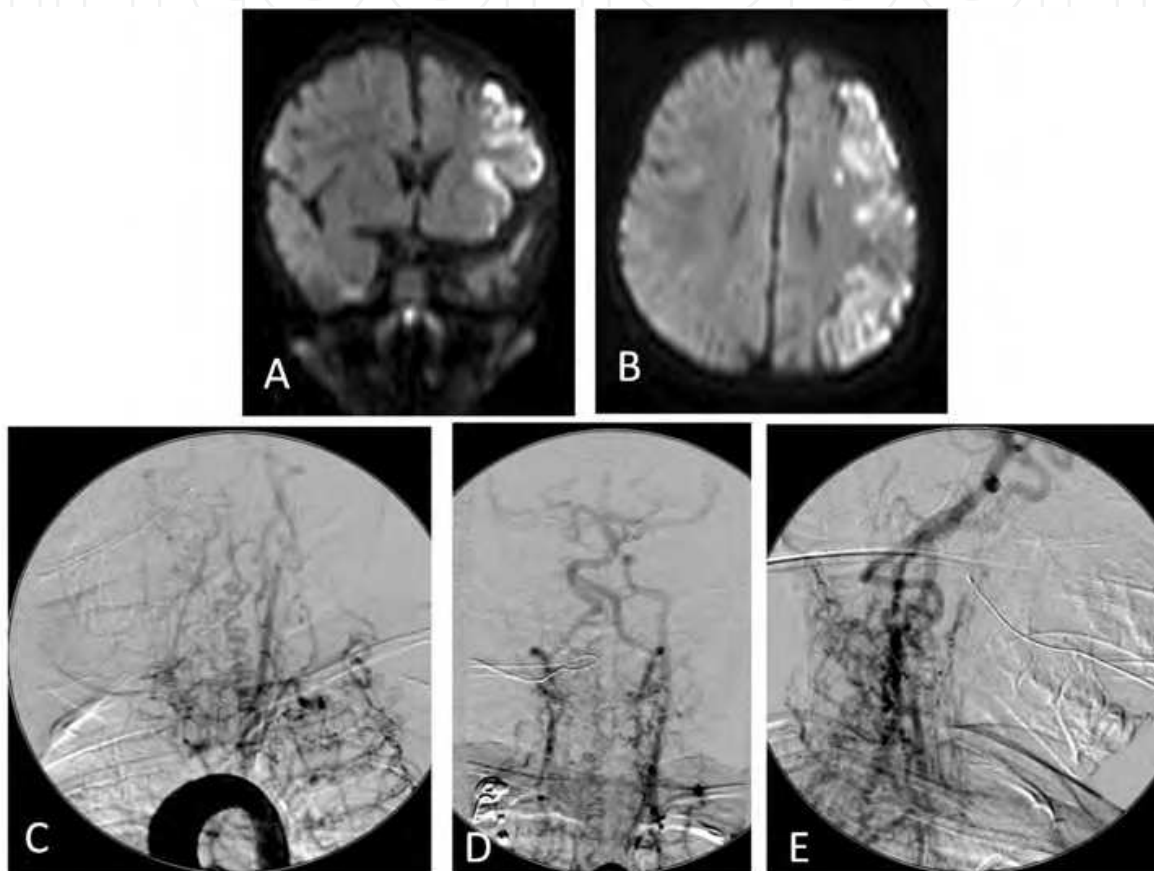


Fig. 1. Magnetic resonance imaging demonstrated acute ischemia in the left MCA territory (diffusion-weighted images, (A) coronal and (B) axial views). (C) Catheter angiography (aortic injection, anteroposterior view) revealed severe obliterative arteriopathy of the great vessels originating from the aortic arch. Cerebral blood flow was reconstituted by collaterals from the anterior spinal artery to the vertebral arteries, and from intercostal and cervical arteries to the left CCA. Delayed phase of the angiogram showed late filling of the left ICA and vertobasilar circulation (aortic injection, (D) anteroposterior and (E) lateral views)

The patient's hemiparesis improved with hypervolemic therapy and pressor agents, but his deficits returned when these pressor agents were weaned. Despite reconstitution of blood flow in his left carotid system by his native cervical collateral vessels, his left hemisphere remained the symptomatic side. After several unsuccessful attempts to wean him from hypertensive therapy, a revascularization procedure was planned to address his hemispheric hypoperfusion and inadequate collateral blood flow.

### Microsurgical Technique

An aortocarotid bypass from the aortic arch to the common carotid artery was determined to be the shortest and best method of augmenting blood flow to the ischemic left hemisphere. An oblique incision along the medial border of the left sternomastoid muscle was made just above the clavicle and the proximal left common carotid artery was exposed, revealing an indurated and completely obstructed artery. The aortic arch was then exposed through a median sternotomy. The aortic arch appeared normal, but the common carotid artery was also indurated and completely obstructed. The origin of the common carotid artery was occluded with a Weck clip and transected from the arch. There was no retrograde flow from the CCA, and a longitudinal incision was made along the carotid until a lumen with back bleeding was encountered. The lumen was probed with a 4-mm dilator and back-bleeding became more brisk. The end of the CCA was cleaned and prepared for the distal anastomosis.
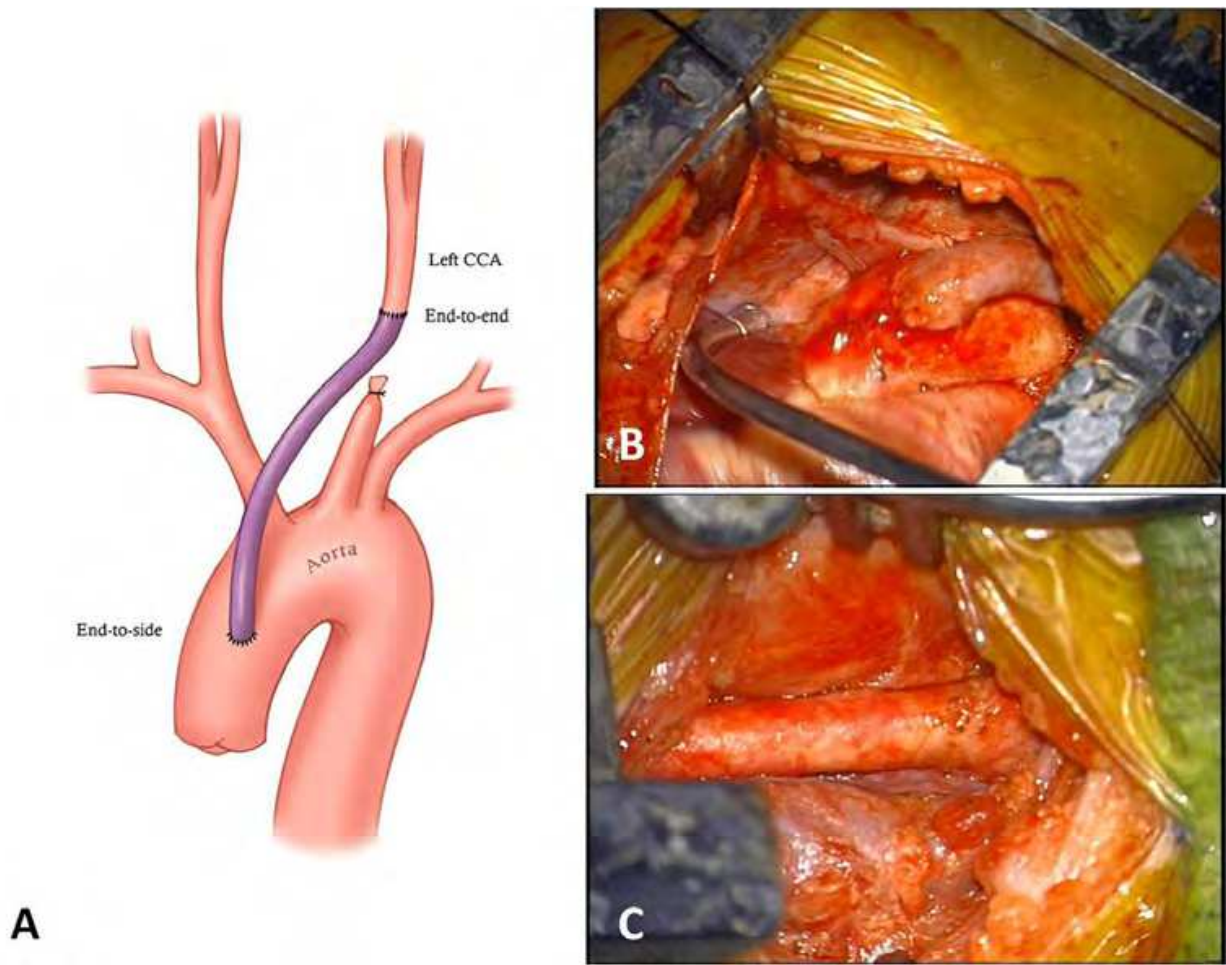


Fig. 2. (A) Illustration showing the aortocarotid bypass from the aortic arch to the proximal left CCA, using a cryopreserved saphenous vein allograft. (B) The aortic arch was exposed through a median sternotomy and a proximal end-to-side anastomosis was performed using a side-biting clamp on the aorta. (C) The graft was tunneled to a cervical incision and an end-to-end anastomosis to the CCA completed the bypass, with strong pulsations in the carotid bifurcation.

The boy's saphenous vein was considered for the bypass, but the caliber was too small and instead, a 5-mm diameter, cryopreserved, allograft saphenous vein was selected (CryoLife, Kennesaw, GA). The ends of the vein graft and the common carotid artery were beveled and anastomosed end-to-end. The vein graft was then tunneled to the mediastinum and trimmed to reach the aorta. A side-biting clamp was placed on the aortic arch, obviating the need for cardiopulmonary bypass. A small incision was made in the aorta and the proximal end of vein graft was sewn to the aorta in an end-to-side fashion. After completing the anastomosis, the side-clamp was removed and flow was established in the bypass with good pulsation in the vein graft and in the downstream carotid artery. Heparin (1mg/kg) was used during the procedure and was not reversed.

**Clinical Course**

Postoperatively, the patient continued to improve and his hemiparesis resolved completely. His pressor agents were weaned and he remained neurologically intact, independent of his blood pressure. An angiogram demonstrated the widely-patent interpositional vein graft between the aortic arch and left common carotid artery. Furthermore, the arterial vessels of the cerebral vasculature appeared well-perfused via the left internal carotid artery.

At last clinical follow-up 12 months after presentation, the bypass remained patent based on carotid ultrasound examination, and the patient demonstrated no new neurological symptoms. He was advancing steadily with his developmental milestones and his language function was normal.
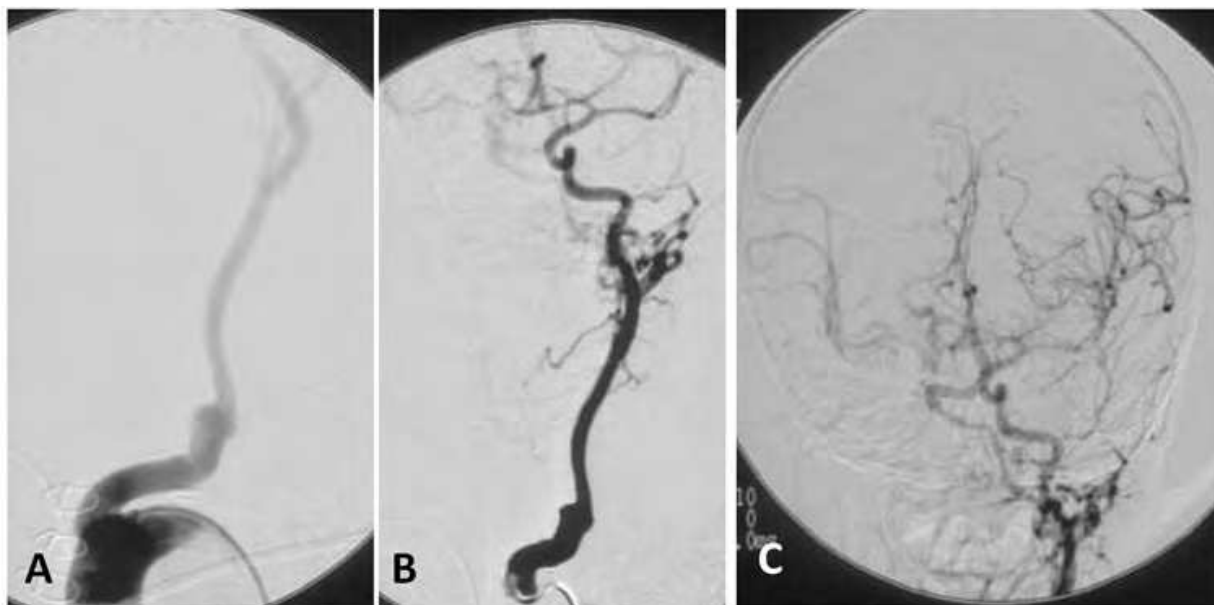


Fig. 3. Postoperative catheter angiography demonstrated a patent aortocarotid bypass ((A) aortic injection, anteroposterior view) and improved cerebral perfusion through the left ICA ((B) graft injection, anteroposterior view). (C) Delayed phase of the angiogram showed filling of the right carotid circulation via anterior communicating artery and the basilar circulation via the posterior communicating artery.

**Case 2**

A 48 year-old female with a history of Takayasu's arteritis presented to the ER with a sudden, severe headache. CT scan demonstrated subarachnoid hemorrhage. Cerebral

angiography revealed a fusiform aneurysm at the basilar apex. Both common carotid arteries and her right vertebral artery were occluded. Her entire cerebral circulation was supplied by the left vertebral artery. Since microsurgical clipping of the aneurysm would likely require some temporary occlusion of the basilar artery, a revascularization procedure was planned.
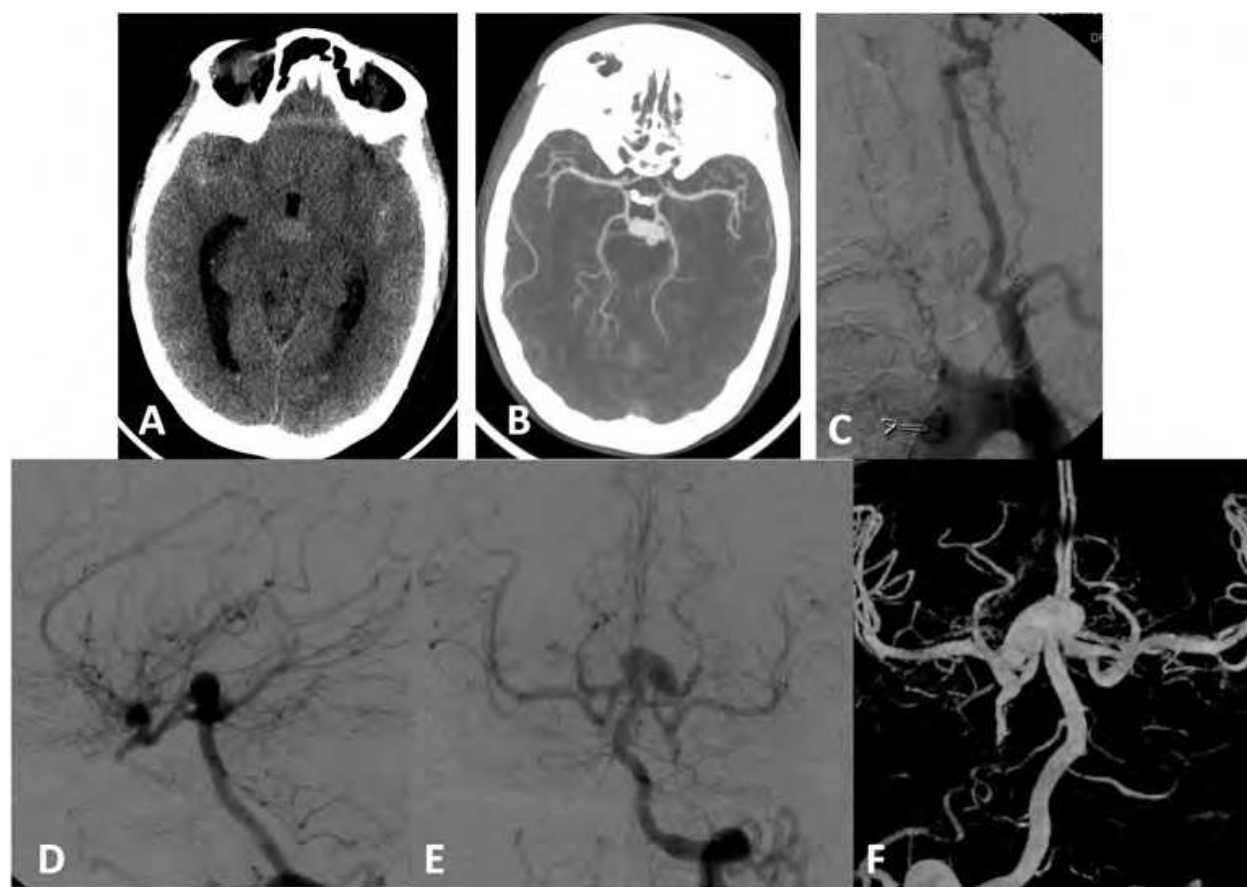


Fig. 4. (A) A brain CT scan revealed subarachnoid hemorrhage and (B) CT angiography revealed a fusiform basilar tip aneurysm. (C) Angiography showed that her innominate artery and left common carotid artery were occluded from her prior disease and surgery (aortic arch injection, anteroposterior view), and only her right vertebral artery was supplying her cerebral circulation. Two large posterior communicating arteries were collateralizing the anterior circulation (D, vertebral artery injection, lateral and (E) anteroposterior views). (F) Angiography with 3D reconstruction depicted the fusiform aneurysm at the basilar apex.

**Microsurgical technique**

A subclavian-middle cerebral artery bypass was thought to be the most suitable revascularization option. A saphenous vein graft was harvested from the thigh and down into the midcalf using endoscopic technique. The subclavian donor site was prepared by reopening a prior surgical incision from previous aorto-subclavian bypass that had occluded. After trimmed appropriately, the vein graft was first anastomosed to the recipient M2 branch. It was tunneled underneath the inferior edge of the craniotomy incision down to the previously prepared subclavian site, cut to the appropriate size, and sewn in and end-to-side fashion to the subclavian artery.

Having placed the bypass, a temporary clip on the basilar trunk softened the aneurysm which allowed complete dissection of the neck and its definitive clipping occluding the rupture site. A fusiform segment of the basilar artery apex was also reinforced with cotton wrapping.
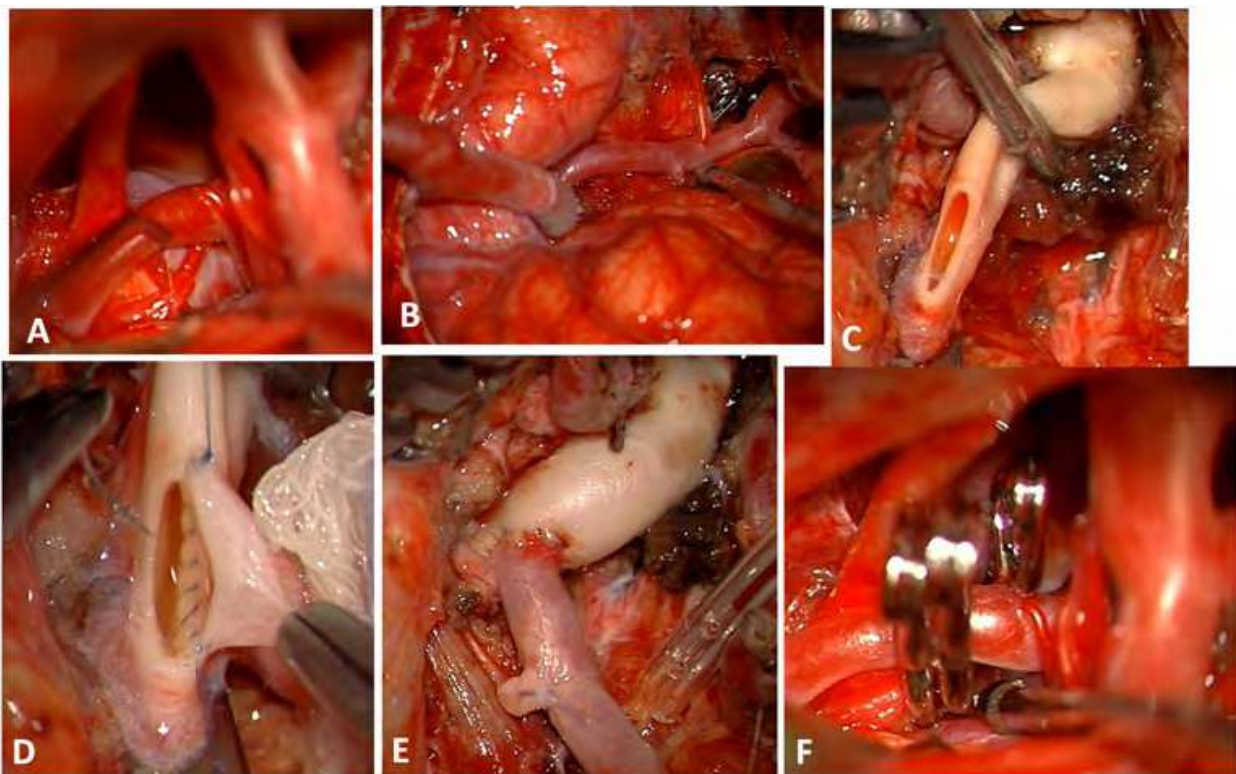


Fig. 5. (A) A left orbitozygomatic approach exposed the basilar apex aneurysm and the need for temporary occlusion was confirmed. (B). A shapenous vein graft was harvested and anastomosed first to the recipient M2 branch. (C) The subclavian artery was exposed through a supraclavicular incision and an arteriotomy was made with an aortic punch. (D,E) The tunneled graft was anastomosed with running 9-0 sutures end-to-side. (F) With the bypass in place, a temporary clip on the basilar trunk softened the aneurysm and allowed complete dissection of the neck. The aneurysm was clipped with fenestrated clips transmitting the PCA and its perforators.

**Clinical course**

Postoperatively, the patient developed a pulmonary embolism which required the placement of a vena cava filter. However, she recovered well and was discharged home after a 2 week hospitalization without any neurological symptom. At last follow-up 4 years after the procedure, the patient remained neurologically asymptomatic and an angiogram demonstrated the patent vein graft between the left subclavian and left middle cerebral artery.

**Case 3**

A 43-year-old woman presented with right sided neglect and expressive aphasia due to left parietal stroke. An angiogram showed an occluded left internal carotid artery with compromised blood flow to the left hemisphere. The right internal carotid artery had a high-

grade stenosis in the proximal half of the cervical segment, but restored caliber in the distal cervical segment and normal blood flow to the right hemisphere. These findings suggested large vessel vasculitis and a revascularization procedure was planned with a STA-MCA bypass.
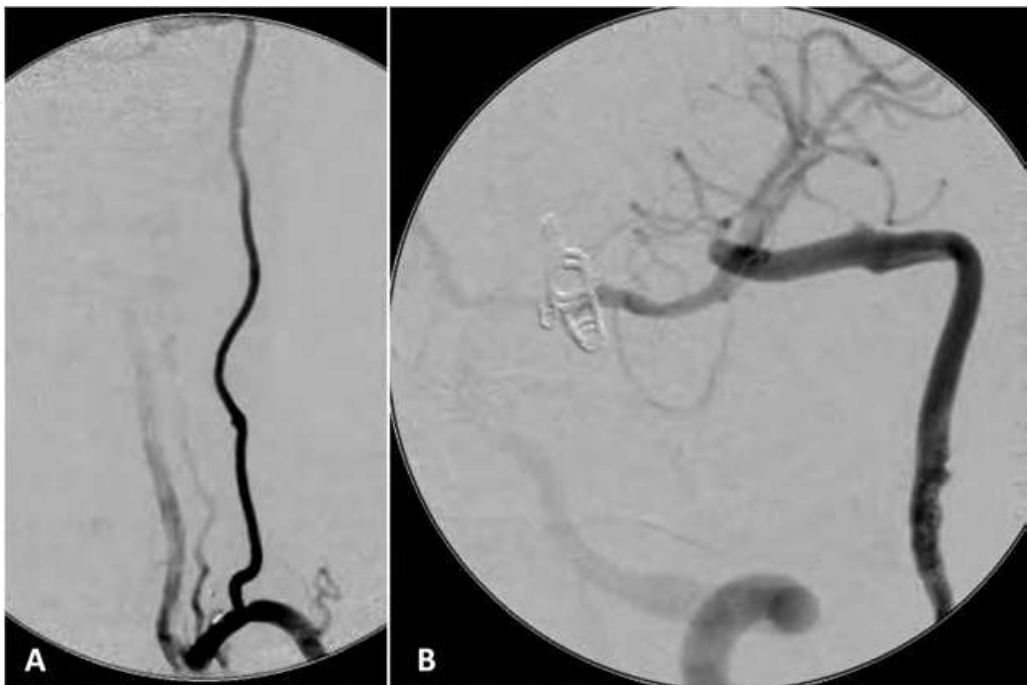


Fig. 6. Postoperative angiogram showed a patent subclavian-MCA bypass ((A) left subclavian injection, anteroposterior cervical view) with good filling of the left MCA territory distally ((B) left subclavian injection, anteroposterior cranial view).
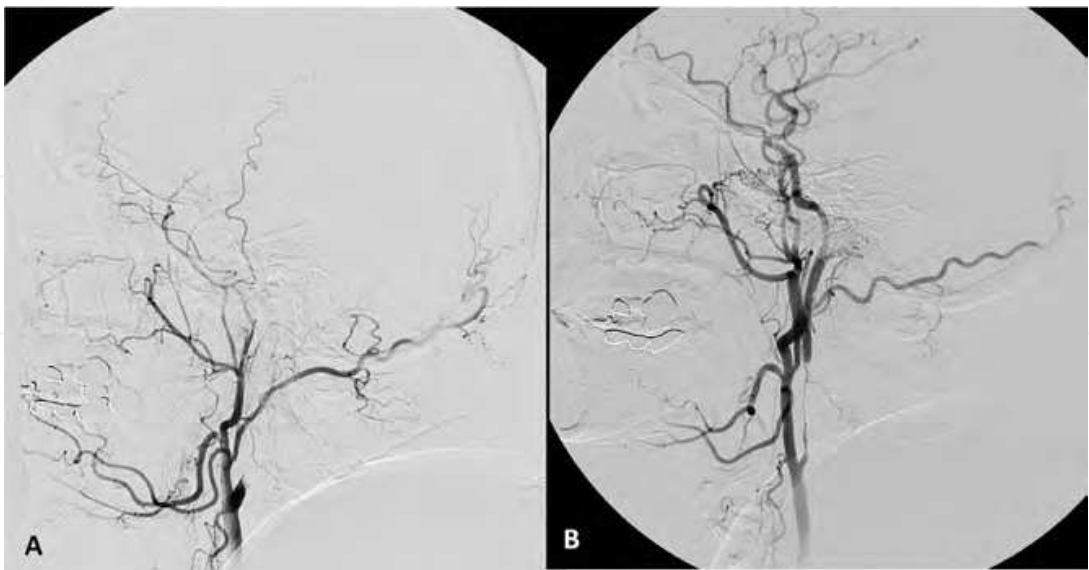


Fig. 7. (A) Angiography showed a completely occluded left internal carotid artery (left common carotid artery injection, anteroposterior view) and (B) a high grade stenosis in the cervical segment of the right internal carotid artery (right common carotid artery injection, anteroposterior view.

**Microsurgical technique**

The left temporal artery was harvested from the zygoma all the way up to the superior temporal line. A small fronto-temporal craniotomy was then performed and the possible recipient arteries on the cortical surface were exposed. As previously noticed on the angiogram and most likely related to the patient vasculitis, all the cortical vessels had a smaller caliber than usually observed. An adequate recipient branch was dissected and the temporal artery was swung to it in and end-to- side fashion. An ICG videoangiography confirmed the patency of this technically demanding anastomosis.

**Clinical course**

Postoperatively, a control angiogram demonstrated patency of the left STA-MCA bypass. The patient´s vasculitis was diagnosed as Takayasu´s arteritis. She was discharged home with no neurological deficits.
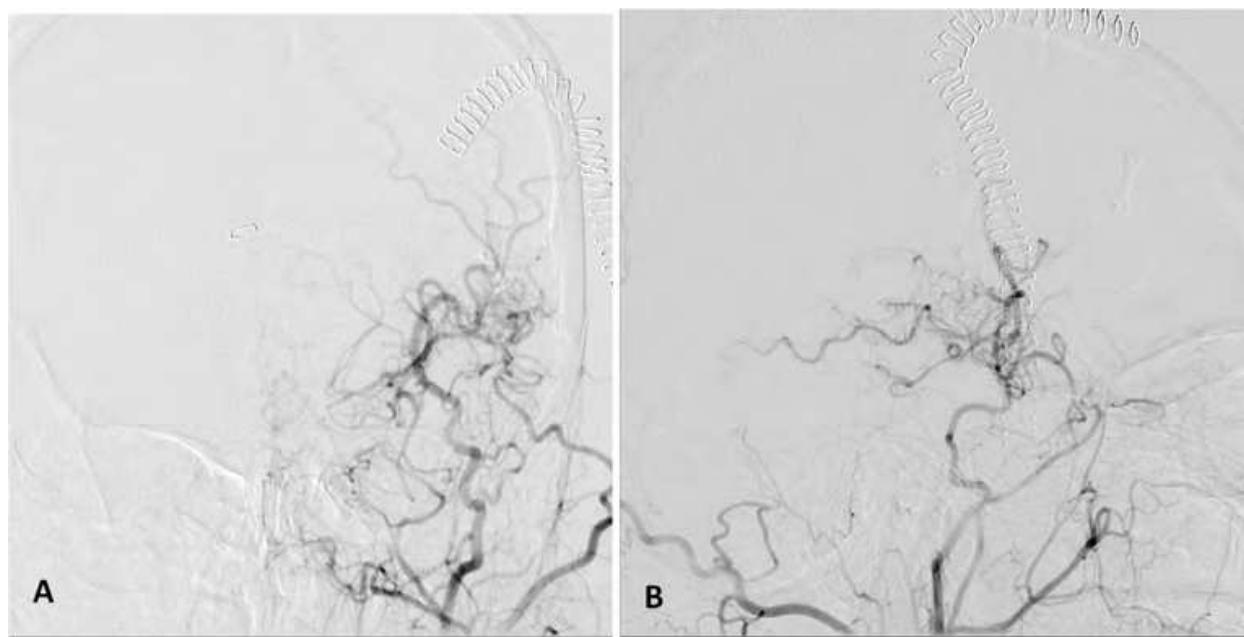


Fig. 8. Postoperative angiogram showed a patent left STA-MCA bypass with good filling of the left MCA territory distally (A) Left common carotid artery injection, anteroposterior view. (B) Left common carotid artery injection, lateral view.

## 3. Discussion

### 3.1 Indications for bypass

The treatment of vasculitis in the acute phases begins with medical therapy with corticosteroids and/or cytotoxic immunosuppressants, the specific medication depending on the type of vasculitis. The indications for brain revascularization in vasculitis are symptomatic cerebral ischemia despite medical therapy, caused by significant reduction in blood flow that is not compensated by collateral circulation.

Catheter angiography helps defining a patient's underlying vasculopathy, but also his or her compensatory collateral circulation. An intact circle of Willis can compensate for a carotid occlusion with cross-filling from the contralateral ICA through the anterior communicating

artery (ACoA) and from the vertebrobasilar circulation through the posterior communicating artery (PCoA). Retrograde flow in the ophthalmic artery can provide collateral circulation from the external carotid artery (ECA). Leptomeningeal collaterals can be recruited over the cerebral convexity connecting PCA and MCA territories as well as ACA and MCA territories. Other less common sources of collateral circulation include: anterior, middle, and posterior meningeal arteries in the dura mater connecting to cortical arteries; ACA-PCA connections through a "limbic loop;" extracranial connections between ECA or vertebral artery branches and the distal ICA; and anterior spinal artery collateralizing the vertebrobasilar circulation. Well developed collateral circulation can compensate for severe occlusive vasculitis, whereas the absence of collateral circulation may indicate surgical bypass (Hofmeijer et al, 2002; Yamauchi et al, 2004).

In addition to the diagnosis of an underlying vasculopathy and the evaluation of the collateral circulation, confirming the presence of ischemic tissue that is at risk for stroke, but still salvageable, is another important criterion for bypass surgery. A large evolving or completed stroke on CT scans or MR imaging contraindicates bypass surgery because this brain tissue is not salvageable and revascularization could precipitate hemorrhagic conversion of an acute infarction. Bypass candidates have small or no strokes and significant areas of ischemia that can be rescued by revascularization. The ability to identify mismatches between blood supply and brain demand helps facilitate patient selection and increases the likelihood of a benefit to those who undergo bypass surgery. Perfusion CT (PCT) imaging is a simple, quick, and accessible technique that provides physiological, qualitative information about those mismatches called the ischemic penumbra (Hemphill et al., 2005; Kamath et al, 2008). PCT imaging uses a bolus injection of iodinated contrast and spiral CT imaging during the passage of the contrast bolus through the brain. Perfusion maps show parameters including: time to peak (TTP, the time between the first arrival of contrast intracranially and its peak concentration), mean transit time (MTT, the average time for blood to travel through a volume of brain), cerebral blood flow (CBF), and cerebral blood volume (CBV). In acute stroke, ischemic penumbra has increased TTP and MTT, relatively normal CBV due to vasodilation and recruitment of collateral flow, and decreased CBF. In contrast, infarcted brain has increased TTP and MTT, but CBV and CBF are both decreased. The identification of ischemic but salvageable penumbra encourages stroke intervention. Measuring salvageable tissue via PET is a more quantitative method that is more difficult to obtain due to limited availability.

Our current algorithm for surgical selection identifies patients with symptomatic lesions refractory to medical management, no significant infarction in the involved vascular territories, and poor collateral circulation on angiography. PCT imaging then identifies hemodynamic insufficiency as increased MTT, typically with near normal CBV and decreased CBF.

## 3.2 Surgical technique

Selecting the appropriate donor and recipient vessels can be challenging due to the distribution of disease, the lack of normal anatomy, and the lack in some cases of an autologous graft that is available or unaffected by the disease. For example, traditional donor arteries like the STA may be involved with the pathology and therefore, alternative bypasses are needed.

### STA-MCA bypass

STA-MCA bypass is by far the most common bypass for any stroke patient, whatever the etiology. The patient is positioned supine and with the head turned 90 degrees to align the lateral convexity parallel to the floor. The course of the STA is mapped with a Doppler flow probe and the posterior limb is usually selected. The anterior limb is used only when the posterior limb is too small. The microscope is used to dissect the STA from zygoma to the superior temporal line. The artery is left in continuity and vasodilated with papaverine extraluminally. Temporalis muscle is divided under the STA and mobilized to each side. Alternatively, temporalis can be flapped anteriorly to expose directly over or in the Sylvian fissure for a larger recipient artery. A small craniotomy is made by working under the STA and staying below the superior temporal line. Dura is opened in flap that preserves middle meningeal arteries if they have developed collateral circulation.

MCA vessels exiting the Sylvian fissure are inspected under the microscope and a recipient artery is selected with suitable size and accessibility. The STA is transected, stripped of its adventitia, cut at a 60 degree angle, and spatulated with an axial incision in the arterial wall. After the patient is placed into propofol-induced EEG burst suppression, the recipient artery is trapped between temporary clips, arteriotomized, and flushed with heparinized saline. Two 10-0 sutures are placed on either end of the arteriotomy to approximate the STA and MCA. Running continuous sutures are placed from one end of the arteriotomy to the other, then tightened and tied. The opposite wall is sutured similarly. The temporary clips are removed and hemostasis is achieved with fibrillar Nu-Knit packing. Indocyanine green videoangiography confirms the patency of the bypass.

The closure must avoid compromising flow in the bypass. The dural closure should not constrict the STA is it passes through. A small passageway for the STA is rongeured from the bone flap. Temporalis muscle is closed loosely around the graft, and galea sutures and skin staples are placed carefully to avoid pinching the graft.

### High-flow bypasses

Vasculitis patients have variable and multifocal disease. Traditional donor arteries like the STA may be involved with the pathology and, therefore, alternative bypasses are needed. Bypasses with interposition grafts like saphenous vein or radial artery have higher flow than traditional EC-IC bypasses.

The cervical carotid artery is an accessible donor artery, with anastomotic sites on the ICA, ECA, and common carotid artery (CCA). The ICA is used when a patent stump is available and collateral circulation from the ECA must be preserved. ECA is used when the cerebral circulation is critically dependent on ICA flow and temporary ICA or CCA occlusion during the anastomosis would be poorly tolerated. CCA is used when the carotid bifurcation is high-riding and exposure is compromised by the mandible (Auguste et al., 2001; Sanai et al., 2008). High-flow bypasses connect the ICA, ECA, or CCA with the MCA, typically along the M2 segment distal to the lenticulostriate arteries originating from the M1 segment.

A similar bypass can be constructed for high-flow bypasses to the posterior circulation. The PCA is preferred over SCA because its caliber matches that of the bypass graft. A shorter bypass alternative is the MCA-PCA bypass, which uses the MCA as the donor site rather than the cervical carotid artery. This bypass is entirely intracranial (IC-IC), spares the patient

a cervical incision, and requires only a short radial artery graft. Shorter grafts generally have higher long-term patency rates than longer grafts to the neck.

Vasculitis can also compromise patency of cervical carotid arteries. In these rare cases, a more proximal donor artery is needed (**see case 1**). The subclavian artery can be accessed through a supraclavicular incision and serve as a proximal donor. The distance to the recipient site is much longer than with other high-flow bypasses involving the cervical carotid artery, and therefore saphenous vein interposition grafts are typically required. The subclavian artery-to-MCA bypass can be used in these vasculitis patients and those with previous surgical carotid sacrifice for aneurysm, tumor, or other intervention.

## 4. Conclusion

Bypass surgery has an infrequent, but important role in the treatment of symptomatic vasculitis refractory to medical therapy causing cerebral ischemia. Vasculitis patients with symptomatic cerebral ischemia caused by significant reduction in blood flow that is not compensated by collateral circulation should be considered for revascularization procedures. However, bypass techniques can be especially challenging in these heterogeneous diseases and neurosurgeons should be aware of the technical difficulties associated with these procedures.

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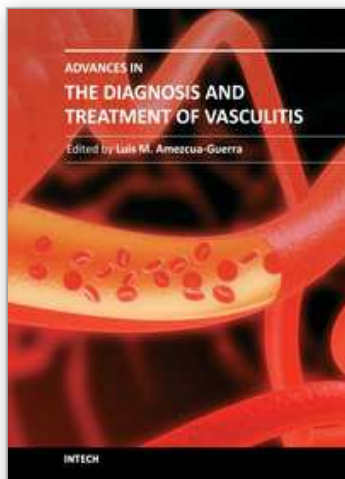
**Advances in the Diagnosis and Treatment of Vasculitis**
Edited by Dr. Luis M Amezcua-Guerra

ISBN 978-953-307-786-4
Hard cover, 366 pages
**Publisher** InTech
**Published online** 09, November, 2011
**Published in print edition** November, 2011

This book represents the culmination of the efforts of a group of outstanding experts in vasculitis from all over the world, who have endeavored to draw themselves into this volume by keeping both the text and the accompanying figures and tables lucid and memorable. The book provides practical information about the screening approach to vasculitis by laboratory analysis, histopathology and advanced image techniques, current standard treatment along with new and more specific interventions including biologic agents, reparative surgery and experimental therapies, as well as miscellaneous issues such as the extra temporal manifestations of "temporal arteritis" or the diffuse alveolar hemorrhage syndrome. The editor and each of the authors invite you to share this journey by one of the most exciting fields of the medicine, the world of Vasculitis.

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Ana Rodrĩguez-Hernández, S. Andrew Josephson and Michael T. Lawton (2011). Bypass for the Treatment of Vasculitis Affecting the Central Nervous System, Advances in the Diagnosis and Treatment of Vasculitis, Dr. Luis M Amezcua-Guerra (Ed.), ISBN: 978-953-307-786-4, InTech, Available from: http://www.intechopen.com/books/advances-in-the-diagnosis-and-treatment-of-vasculitis/bypass-for-the-treatment-of-vasculitis-affecting-the-central-nervous-system

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