Aortocarotid bypass for hemispheric hypoperfusion in a child

Technical note

NADER SANAI, M.D.,1 HEATHER FULLERTON, M.D.,2 TOM R. KARL, M.D.,3 AND MICHAEL T. LAWTON, M.D.1

Departments of 1Neurological Surgery and 2Pediatric Neurology; and 3Pediatric Heart Center, University of California at San Francisco, California

Large-vessel vasculitis syndromes in the pediatric population are rare and highly morbid. The authors here report on the microsurgical revascularization of a unique case of presumed vasculitis with aortitis and severe obliterative arteriopathy in a 10-month-old child with symptomatic hemispheric hypoperfusion. Using a cryopreserved saphenous vein, this unilateral aortocarotid bypass restored normal intracranial perfusion bilaterally and led to a resolution of the patient’s ischemic symptoms. The aortocarotid bypass is clinically effective and technically feasible in young children when a saphenous vein allograft is used. The bypass graft is amenable to angioplasty with or without stenting if delayed stenosis becomes an issue later in life. (DOI: 10.3171/PED/2008/1/4/343)

KEY WORDS • aortocarotid bypass • cerebral ischemia • hypoperfusion • obliterative arteriopathy • pediatric stroke • revascularization • vasculitis

LARGE-VESEL vasculitides, such as giant-cell or temporal arteritis and Takayasu arteritis, are characterized by inflammation of the aorta and its major branches. When the carotid arteries are involved, cerebral ischemia is clinically manifested as a stroke, postural dizziness, seizures, and amaurosis. These conditions typically involve similar histological abnormalities, but distinct ages of onset and targeted vascular structures. Giant-cell arteritis, for example, almost exclusively affects a population older than 50 years of age, whereas Takayasu arteritis typically afflicts adolescent girls and women in the 2nd and 3rd decades of life. In the former, incidence rates reach 15–25 cases per 100,000 persons with a predilection for those of northern European descent; the latter syndrome is most commonly seen in Japan, Southeast Asia, India, and Mexico and has an annual incidence in North America of 2.6 cases per 1 million people.

In addition to cerebrovascular consequences, the systemic vascular inflammation associated with most known large-vessel vasculitides can lead to severe hypertension from renal artery stenosis and congestive cardiac failure due to hypertension, aortic regurgitation, and dilated cardiomyopathy. In patients with giant-cell or Takayasu arteritis, obliterative vascular lesions are also accompanied by fever, malaise, arthralgias, myalgias, weight loss, and anemia. Management paradigms are almost always medical and tailored to the patient’s symptomatic profile, involving a combination of corticosteroids, aspirin, and immunosuppressive agents intended to limit luminal occlusion.

Interestingly, comparable large-vessel vasculitis syndromes in the pediatric population are rare. Pediatric Takayasu arteritis is particularly rare and usually manifests systemic, nonneurological findings such as hypertension, cardiomegaly, and fever as well as angiographic abnormalities such as abdominal aortic stenosis and aneurysms, both intra- and extracranial. Nevertheless, most childhood vasculitides—Henoch–Schönlein purpura, Kawasaki disease, polyarteritis nodosa, Wegener granulomatosis, and Behçet disease—are medium- and small-vessel syndromes that often manifest pathognomonically and rarely, if ever, lead to aortitis or large-vessel obliterative arteriopathy. Treatment of these vasculitides, as in adults, is nonsurgical and relies on corticosteroid therapy to suppress immune-mediated responses.

Here, we describe microsurgical revascularization of a
unique case of presumed vasculitis with aortitis and severe obliterative arteriopathy in a symptomatic 10-month-old child with hemispheric hypoperfusion.

Materials and Methods

Patient Presentation and Examination

This previously healthy 10-month-old boy presented with an inability to move his right side following several hours of agitation and a series of complex partial seizures. He had no known history of viral illness and he was afebrile and normotensive. Neurological examination confirmed a right-sided paresis with 2/5 strength in his arm and leg.

Magnetic resonance imaging and MR angiography (Fig. 1) demonstrated acute ischemia in the left middle cerebral artery territory. Although a prothrombotic state was suspected, a laboratory workup was nondiagnostic. Multisys-

Fig. 1. Preoperative coronal (A and B) and axial (C and D) diffusion-weighted MR images demonstrating left hemispheric regions of ischemia. Axial T2-weighted MR images (E and F) revealing diffuse T2 prolongation in the affected areas.

Fig. 2. Preoperative catheter angiography demonstrating severe obliterative arteriopathy of all vessels arising from the aortic arch (A and B) with no direct connection of the arch vessels to distal cervical vasculature extending into the brain (C and D). Extensive collateralization of the great cervical vessels from intercostals and the anterior spinal artery can also be seen, predominantly affecting the left carotid and vertebral arteries (E and F).

tem imaging suggested that the distal aorta as well as the abdominal and renal circulation were normal. Additionally, serologic tests for Takayasu arteritis were negative.

Cerebral angiography (Fig. 2) demonstrated severe obliterative arteriopathy of the great vessels with complete occlusion of all vessels originating from the aortic arch and reconstitution of the cerebral vasculature through unique collateralization patterns. The anterior spinal artery was markedly dilated, supplied by branches from the thoracic aorta, and contributed flow to the vertebrobasilar circulation. The cervical vertebral arteries were collateralized by an extensive network of small intercostal arteries. These intercostal arteries also collateralized the left CCA, with antegrade filling of the left ICA seen in delayed phases of the angiogram. He had large posterior communicating arteries bilaterally that contributed blood supply to the anterior circulation. No reconstitution of blood flow was seen in the right ICA. The cerebral vasculature appeared to be struc-
turally unaffected by the underlying vascular pathophysiology.

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

**Microsurgical Technique**

An aortocarotid bypass from the aortic arch to the CCA was determined to be the shortest and best method for augmenting blood flow to the ischemic left hemisphere. An oblique incision along the anterior border of the left sternomastoid muscle was made just above the clavicle, and the proximal left CCA 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 CCA was also indurated and completely obstructed. The origin of the CCA 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 artery 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.

The boy’s saphenous vein was considered for the bypass, but the caliber was too small and a 5-mm-diameter, cryopreserved saphenous vein allograft was selected instead (CryoLife; Fig. 3). The ends of the vein graft and the CCA were beveled and anastomosed end-to-end. The vein graft was then tunneled into anatomical position to the mediastinum and trimmed to reach the aorta. A side-biting clamp was placed on the aortic arch, obviating cardiopulmonary bypass. A small incision was made in the aorta, and the proximal end of the 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 (1 mg/kg) was used during the procedure and was not reversed.

**Results**

Postoperatively, the patient’s condition continued to improve and the hemiparesis resolved completely. The pressor agents were weaned and he remained neurologically intact, independent of his blood pressure. An angiogram (Fig. 4) demonstrated the widely patent interpositional vein graft between the aortic arch and left CCA. Furthermore, the arterial vessels of the cerebral vasculature appeared well perfused via the left ICA.

Anatomical and immunohistochemical analysis of the diseased proximal carotid artery were notable for severe intimal proliferation and abundant reticulin fibers. The tissue weakly expressed CD45, indicating a modest lymphocytic infiltration. Additional pathological studies of the spe-

**Fig. 3.** Intraoperative photographs and drawing showing the 5-mm-diameter, cryopreserved saphenous vein allograft used for the bypass.  
A: Arrow indicates the proximal anastomosis to the aortic arch, as viewed through the sternotomy exposure.  
B: The CCA and its bifurcation are seen through the cervical incision after blood flow has been augmented via the bypass.  
C: Note the locations of the bypass anastomoses relative to the local vascular anatomy.
cimen were nonspecific, however, and an exact diagnosis was not assigned, although all known childhood vasculitides were ruled out.

At the last clinical follow-up 12 months after the initial presentation, the bypass remained patent according to carotid artery ultrasonography examination, and the patient demonstrated no new neurological symptoms. He was advancing steadily with his developmental milestones, and his language function was normal. There were no new episodes of seizure or stroke.

Discussion

We described childhood vasculitis associated with a severe, large-vessel oblitative arteriopathy that was amenable to an aorto-carotid bypass using a cadaveric vein graft. Although large-vessel vasculitides have been treated with direct bypass in adults, the present case is the first report of such a bypass in a young child.

Although the patient’s disease was not defined, its sudden onset, multifocality, and severity are suggestive of an acquired large-vessel vasculitis. Interestingly, extensive laboratory testing for a hypercoagulable state, viral illness, or immune-related disorders were all nondiagnostic. Furthermore, tissue analysis from the grossly abnormal proximal CCA was also nonspecific, although suggestive of an inflammatory reaction that precipitated complete obliteration of the proximal vessel lumen.

Whereas previously reported adult aortocarotid bypasses often have involved the use of a harvested saphenous vein graft, the young age of our patient precluded the use of such a small-caliber vessel. We considered using a polytetrafluoroethylene graft, but we were concerned about the necessity for long-term patency in this young child and the relatively short follow-up in published experiences with these synthetic grafts (usually < 50 months). Therefore, we elected to use an cadaveric adult saphenous vein graft with a reliability that has been established with peripheral vascular reconstructive surgeries. Recent studies in adolescents undergoing lower-extremity revascularization procedures with saphenous vein grafts have demonstrated a > 90% patency rate, but these studies have, at most, 11 years of clinical follow-up. Similarly, in moyamoya disease in pediatric patients undergoing direct bypass procedures, an even greater long-term patency rate has been observed, although the reported clinical follow-up was often shorter. Data in the brief clinical follow-up in the present case do not suggest any significant autoimmune or inflammatory reaction to the graft. Given that we have performed cerebral revascularization with such a graft in the youngest patient to date, we will follow his course with great interest. Unlike synthetic grafts, saphenous vein allografts can be treated with angioplasty and stenting if stenotic changes occur. This capacity of the saphenous vein allograft was another important reason for its selection in the present case.

Cerebral hyperperfusion and hemorrhage are recognized complications of aorto-carotid bypass surgery and other cranio-cervical revascularization procedures. However, these complications can be avoided with strict postprocedural blood pressure control and minimal or no anticoagulation. Despite the middle cerebral artery territory ischemia evident on preoperative MR imaging in this patient, there were no sequelae attributable to increasing his cerebral perfusion.

Conclusions

This unusual vasculitic condition affecting this 10-month-old boy was amenable to a direct aortocarotid bypass. This simple bypass effectively compensated for the sudden hemispheric hypoperfusion and restored a normal neurological state. The long-term efficacy and durability of this bypass strategy will be interesting to follow.

Disclaimer

The authors are not involved in any financial relationships with the companies that make the products related to this study.
References


Address correspondence to: Michael T. Lawton, M.D., Department of Neurological Surgery, University of California at San Francisco, 505 Parnassus Avenue, M779, Box 0112, San Francisco, California 94143-0112. email: lawtonm@neurosurg.ucsf.edu.