CASE REPORTS

From the Midwestern Vascular Surgical Society

Long-segment thoracoabdominal aortic occlusions in childhood

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Developmental coarctation, hypoplasia, and occlusion of the abdominal aorta is a rare disease encompassing many differing etiologies and diverse methods of treatment. Long-segment thoracoabdominal aortic occlusion, an extreme manifestation of this disorder, has not previously been reported in children. Two pediatric patients with this entity, a 5- and 13-year-old with uncontrolled hypertension, underwent extensive arterial reconstructions for this entity and provided the impetus for this report. An ascending thoracic aorta to infrarenal aortic expanded polytetrafluoroethylene bypass was undertaken in the younger child. A distal thoracic aorto-bi-iliac artery expanded polytetrafluoroethylene bypass, with implantation of the left renal artery to one graft limb and a right renal artery bypass originating from the other limb, was performed in the older child. There were no major perioperative complications. Both patients were discharged with easily controlled blood pressures. They have remained normotensive at 13 and 14 months follow-up. (J Vasc Surg 2012;56:482-5.)

Developmental abdominal aorta narrowing is a rare disease that is infrequently associated with an occlusion. Anecdotal case reports in the literature have described this disease in adults, but lengthy occlusions of the distal thoracic and abdominal aorta in children have not been reported. Two pediatric patients who recently underwent extensive aortic reconstructions for this disease are reported. This review has been approved by the University of Michigan Institutional Review Board (HUM 00006223).

CASE REPORTS

Case 1. A 5-year-old girl was diagnosed with hypertension during a routine physical examination. She was of normal stature, being 115 cm tall (>90th percentile) and weighing 26 kg (>95th percentile). She was the product of an uncomplicated, full-term pregnancy; there was no preceding systemic or febrile illness suggestive of vasculitis. Her mother reported that the child experienced dyspnea with heavy exertion.

An electrocardiogram at the referring children's hospital revealed left ventricular hypertrophy, and an echocardiogram revealed aortic narrowing distal to the juxtaductal region. A magnetic resonance angiogram (MRA) revealed occlusion of a long segment of the thoracic and upper abdominal aorta with preserved flow to the distal aorta and its branches through extensive collaterals (Fig 1, *A*). There were no renal or splanchnic arterial stenoses.

The child was subsequently admitted to the University of Michigan Mott Children's Hospital. At that time, she was hypertensive with a pressure of 191/104 mm Hg despite being on amlodipine 4 mg BID, salbutamol 2 mg BID, and doxazosin 3.5 mg QD. Her serum creatinine was 0.6 mg/dL.

It was elected to proceed with an aortic reconstruction. The ascending aorta and abdominal aorta were exposed through a median sternotomy extended inferiorly onto the abdominal wall. A diminutive infrarenal aorta was noted without obvious pulsatile flow. There was no palpable aortic tissue at the diaphragmatic hiatus. Partial aortic occlusion of the left anterior ascending aorta was obtained with a side-biting vascular clamp, and an aortotomy was fashioned to which a 16-mm expanded polytetrafluoroethylene (ePTFE) graft was anastomosed. The graft was placed through the posterior pericardium and positioned anterior to the main pulmonary artery along the left lateral aspect of the heart in such a way to avoid compression of the pulmonary veins. Modest graft redundancy was allowed for anticipated later growth. The graft was then advanced further through the left diaphragmatic crus, into the retroperitoneum behind the left kidney, to the infrarenal aorta. The distal anastomosis of the graft to the aorta at this level was constructed proximal to the inferior mesenteric artery in an end-to-side fashion. Her postoperative course was unremarkable other than for prolonged ventilator support. She was discharged 17 days later, being normotensive on two antihypertensive agents. A postoperative computed tomography angiogram obtained just prior to discharge

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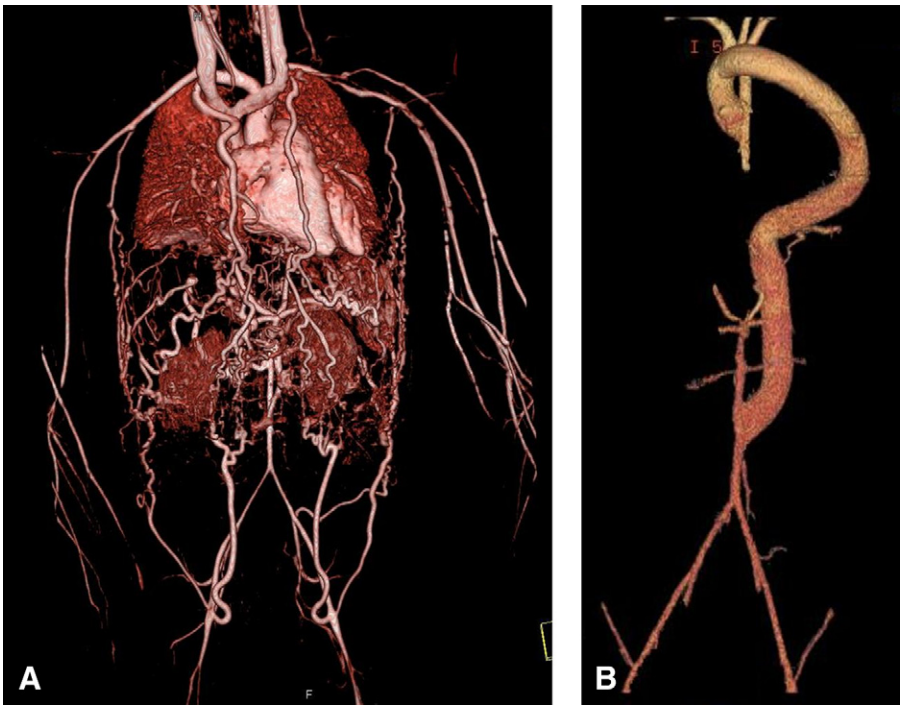

Fig 1. A, Reformatted preoperative magnetic resonance angiogram (MRA) in a 5-year-old, documenting occlusion of the thoracic aorta and upper abdominal aorta. Extensive collateral circulation has restored blood flow to the midabdominal aorta just above the renal arteries, as well as the pelvic and lower extremity circulation. **B,** Postoperative computed tomography angiogram documenting patency of a 16-mm expanded polytetrafluoroethylene (ePTFE) graft originating from the ascending thoracic aorta, with infrarenal abdominal aortic insertion.

revealed the thoracoabdominal graft extending from distal ascending aorta to infrarenal abdominal aorta (Fig 1, *B*).

The 6-month follow-up MRA confirmed patency of the thoracoabdominal graft and well-opacified visceral and renal arteries. At the 13-month follow-up, her blood pressure was normal on amlodipine 2.5 mg BID alone. Her weight had increased to 33 kg and her left ventricular hypertrophy had resolved. Her exercise capacity was normal.

Case 2. A 13-year-old girl underwent evaluation at another hospital for uncontrolled hypertension while on enalapril 10/5 mg-am/pm, atenolol 50/37.5 mg-am/pm, and minoxidil 5/2 mg-am/pm. Her preoperative blood pressures averaged 140 to 150/80 mm Hg. She was 168 cm tall (95th percentile) and weighed 46 kg (50th percentile). There was no history of lower extremity fatigue or claudication. A known abdominal aortic coarctation had been diagnosed at 3 months of age by MRA. There was no history suggesting a vasculitis manifest by a systemic or febrile illness. She was born following a full-term pregnancy complicated by premature labor and polyhydramnios.

The referring physicians documented a 30- to 40-mm Hg gradient between the upper and lower extremity arterial pressures. An electrocardiogram raised suspicion for left ventricular hypertrophy. An MRA revealed occlusion of the descending thoracic aorta just above the diaphragmatic hiatus with reconstitution of both distal common iliac arteries, and no evidence of any intervening aorta (Fig 2, *A*). Bilateral renal arteries as well as the distal superior mesenteric artery were reconstituted through multiple collaterals. The kidneys were normal in size. Her serum creatinine was 0.6 mg/dL.

She was subsequently admitted to the University of Michigan Mott Children's Hospital for reconstruction of her aorta and renal arteries. Exposure of the thoracic and abdominal aorta was facilitated by a thoracoabdominal incision through the ninth intercostal space that extended onto the abdomen. Exploration revealed a normal-sized distal thoracic aorta and reconstitution of the distal common iliac arteries. The distal right and left renal arteries appeared normal. Both were exposed following a medial reflection of the overlying viscera. A retroperitoneal tunnel through which the graft was to be placed was created from the left thorax, through the left leaf of diaphragm, behind the left kidney, to the iliac vessels.

A distal thoracic aorta-to-bilateral common iliac arterial bypass was performed using a 16- × 8-mm bifurcated ePTFE graft. The left renal artery was directly reimplanted onto the graft's left limb, and the right renal artery bypassed with a 7-mm ePTFE conduit originating from the graft's right limb.

Her postoperative course was uncomplicated, and she was discharged on the 11th postoperative day. A computed tomography arteriogram confirmed the patency of the aortobiiliac graft and the reconstructed left and right renal arteries (Fig 2, *A* and *B*). She was on two antihypertensive agents and her creatinine remained stable, 0.4 mg/dL, at the time of discharge.

She has subsequently returned to normal activities, including competitive swimming. At 14-months follow-up, she was normotensive while on amlodipine 5 mg BID and atenolol 25

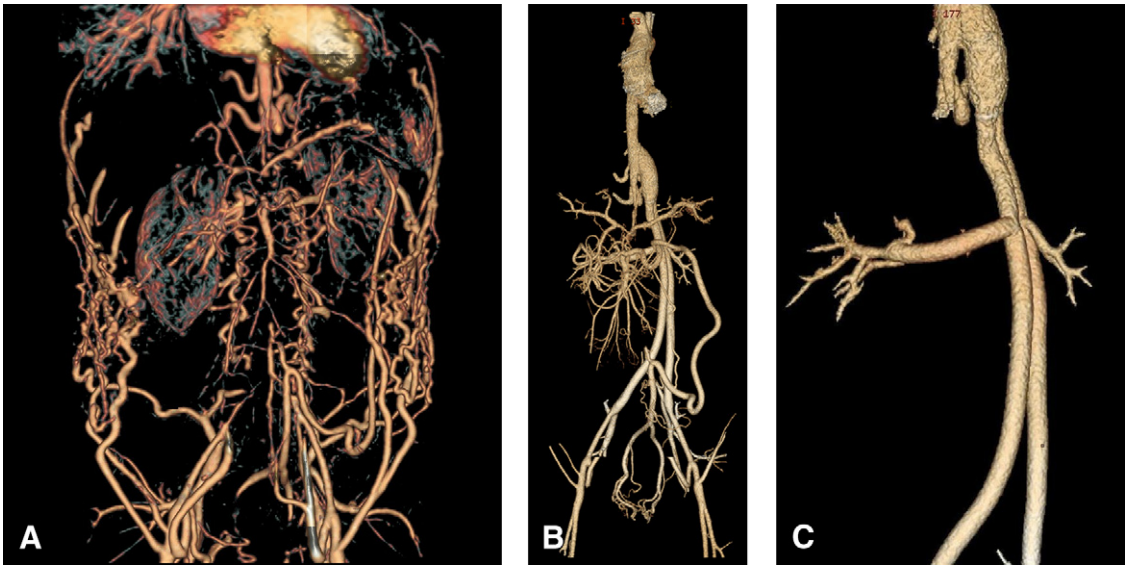

Fig 2. A, Reformatted preoperative magnetic resonance angiogram (MRA) in a 13-year-old, documenting absence of the distal thoracic aorta and entire abdominal aorta and extensive collateral reconstitution of the renal and superior mesenteric arteries, as well as of the distal iliac arteries. **B,** Postoperative computed tomography angiogram documenting the 16- × 8-mm expanded polytetrafluoroethylene (ePTFE) bifurcated graft originating from the midthoracic aorta with its bilateral limbs terminating at the origin of the external iliac arteries. **C,** Reformatted postoperative computed tomography angiogram of the bifurcated ePTFE graft with a 7-mm ePTFE conduit reconstruction of the right renal artery arising from the right graft limb, and reimplantation of the left renal artery onto the left graft limb.

mg BID. Her postoperative resting ankle-brachial indices were normal.

DISCUSSION

Among all coarctations affecting the aorta, <2% involve its abdominal segment.[1-6] While the cause of these aortic narrowings often remains ill-defined, the pathogenesis is thought most often to relate to events occurring around day 25 of fetal development when the two embryonic dorsal aortas fuse and lose their intervening wall to form a single vessel. Both overfusion and failure to fuse with subsequent obliteration of one vessel could predictably result in aortic narrowing.[3]

Viral-mediated injury of mesodermal tissues during embryonic development and certain genetically determined events associated with neurofibromatosis, Alagille syndrome, and Williams' syndrome have been implicated in these cases.[5] Panaortitis (active or chronic) with adventitial or periadventitial fibrosis and associated inflammatory cell infiltrates is another cause of certain of these coarctations. Both patients in the present report were believed to have developmental aortic disease.

Abdominal aortic narrowings are often associated with coexisting splanchnic and renal occlusive disease, with 87% having concomitant renal artery stenosis and 62% exhibiting splanchnic occlusive disease.[5] Most patients with abdominal aortic coarctation present with uncontrolled hypertension due to an activated renin-angiotensin system. Blood pressure elevations in this setting are typically refractory to simple medical-drug control. Untreated patients risk progressive left ventricular hypertrophy, congestive heart failure, flash pulmonary edema, and stroke.

Symptomatic intestinal ischemia due to splanchnic arterial disease is very uncommon, affecting only 6% of our previously published cases of abdominal aortic coarctations.[5] As such, the celiac and superior mesenteric arteries are mandated to be reconstructed only in clinically symptomatic cases. Lower extremity ischemia is similarly uncommon.

Aortic reconstructions in growing children present a unique problem. In constructing a thoracoabdominal aortic bypass in these cases, the graft diameter is selected so as to be as big as possible, short of being so large that excessive luminal thrombus might accumulate.[5] Oversizing grafts, compared with the initial aortic diameter, anticipates growth. In the ideal circumstance, a graft should be chosen that will not represent an energy-consuming narrowing as the patient grows into maturity. We have found that this translates into the use of 8- to 12-mm grafts in young children, 12- to 16-mm grafts in older children and early adolescents, and 14- to 20-mm grafts in late adolescents and adults. At times a smaller graft must be used for a bypass, with an understanding that a replacement will be required at a later time.

Graft length is a nonissue in older children. Axial growth from the diaphragm to the pelvis is minimal after reaching the age of 9 or 10 years. This is not necessarily the case within the thorax. It has been our practice to leave as

much graft redundancy as feasible in the thoracic cavity without risking graft kinking. The optimal amount of thoracic graft redundancy is unknown, but a reasonable strategy mimics the placement of pediatric pacer wires in which the bulk of redundancy resides in the thoracic cavity and not the abdominal cavity.

Furthermore, a certain amount of native vessel growth occurs through conformational changes, similar to that observed following an extracardiac Fontan modification in which an ePTFE conduit is fashioned between the inferior vena cava and the distal right pulmonary artery, or in Blalock-Taussig shunts where an ePTFE conduit is placed between the innominate artery and the pulmonary artery. These former grafts often are placed at birth and never require revision secondary to inherent vessel accommodation.

Certain young children whose initial aortic reconstruction cannot be safely deferred to an older age will outgrow their original graft and require reoperation. In our earlier published experience with abdominal aortic coarctations in 47 children, 6% required reoperation as a result of outgrowing the adequacy of their bypass graft of patch aortoplasty.[5] Clearly, regular clinical follow-up is integral to the care of these individuals. Yearly graft surveillance with MRA is recommended, with the time interval extended to every 2 to 3 years in the setting of several consecutive stable scans.

To our best knowledge, the present report is the first description of long-segment thoracoabdominal aortic occlusion in pediatric-aged patients. The dramatic anatomic presentation of a complete obstruction of such a long aortic segment stands in distinct contrast to the disease's near-silent clinical manifestation. Surgical reconstruction of the occluded aorta in these patients requires careful planning and a modicum of technical expertise. Extensive aortic reconstructions, including appropriate treatment of branch lesions, represent the optimal treatment of long-segment thoracoabdominal aortic occlusions in young patients.

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