Hypoplasia of the descending thoracic and abdominal aorta: A report of two cases and review of the literature

Thomas T. Terramani, MD, Ali Salim, MD, Douglas B. Hood, MD, Vincent L. Rowe, MD, and Fred A. Weaver, MD, Los Angeles, Calif

Hypoplasia of the thoracic and abdominal aorta, referred to as atypical, elongated, or diffuse coarctation, is an exceedingly rare cardiovascular anomaly. Congenital, acquired, inflammatory, and infectious etiologies have been described. Symptoms typically occur within the first three decades of life and include hypertension, lower extremity claudication, and mesenteric ischemia. The condition is considered a life-threatening emergency as a result of the complications associated with severe hypertension. Diagnosis is best made with angiography. Surgical bypass grafting is the optimal method of treatment and must be tailored depending on the distribution of disease. We report two cases of diffuse hypoplasia involving the thoracic and abdominal aorta treated with thoracic aorta to abdominal aorta bypass. (J Vasc Surg 2002;36:844-8.)

Coarctation of the aorta is defined as significant luminal narrowing of the aorta that produces hemodynamically significant obstruction to the flow of blood. This process may affect a short isolated segment of the aorta or a longer and more diffuse segment. The most common location for segmental coarctation is at the attachment of the ductus arteriosus to the thoracic aorta, which accounts for 98% of focal lesions. Coarctation in the abdominal aorta is exceedingly rare, accounting for less than 2% of all cases.

When longer segments of the aorta are narrowed, the term hypoplasia is often used. Much less common than segmental coarctation, aortic hypoplasia has been described in all portions of the thoracic and abdominal aorta. The etiology of this condition is poorly understood, and both congenital and acquired causes have been proposed.

The most common clinical manifestation of aortic hypoplasia is severe uncontrolled hypertension in adolescents and young adults. This article describes our experience in the treatment of two patients with diffuse hypoplasia of the descending thoracic and suprarenal abdominal aorta.

**CASE REPORT**

**Case 1.** A 26-year-old Central American man was seen at the hospital with right-sided weakness that began suddenly. The medical history was significant for untreated hypertension that was first noted during childhood and was associated with frequent episodes of epistaxis. Physical examination on admission revealed a normally developed man with right-sided hemiparesis who needed a cane for ambulation. The patient had normal upper extremity pulses, and the pulses in the lower extremities were diminished. A grade 2/6 systolic murmur was heard over the left sternal border. No cervical or abdominal bruits were noted. The patient was found to be profoundly hypertensive in both arms, with a brachial blood pressure of 240/145 mm Hg and with thigh blood pressures of 122/66 mm Hg. A 12-lead electrocardiogram revealed normal sinus rhythm with left ventricular hypertrophy, and a chest roentgenogram revealed no rib notching or cardiomegaly. A computed tomographic (CT) scan of the brain showed acute hemorrhage in the left basal ganglia. Renal function was found to be normal, with a blood urea nitrogen level of 10 mg/dL and a creatinine level of 0.8 mg/dL. Renal artery duplex scanning showed normal velocities in the renal arteries bilaterally with normal kidney sizes. Of note was an area of probable stenosis in the supraceliac aorta, at which level the peak systolic velocity was 668 cm/s, with a velocity ratio of 5.1. Contrast arteriography obtained via a femoral artery access showed diffuse narrowing of the descending thoracic aorta distal to the subclavian artery and extending to the level of the diaphragm, with a focal area of severe stenosis just above the celiac trunk (Fig 1). The systolic pressure gradient across the area of focal stenosis measured 100 mm Hg. The abdominal aortogram was normal except for bilateral duplicated renal arteries. Magnetic resonance angiography confirmed the previous findings (Fig 2). No periarteric inflammatory changes were noted on cross-sectional views.

This patient ultimately needed three antihypertensive agents for adequate blood pressure control. He was discharged from the hospital and, after completing rehabilitation therapy for the stroke, was readmitted 6 weeks later for operative repair of the aortic hypoplasia. The operation was performed with the patient in a modified right lateral decubitus position. A left posterolateral thoracotomy through the fourth intercostal space was made for access to the descending thoracic aorta. The infrarenal abdominal aorta was accessed from a retroperitoneal approach through a separate oblique incision beginning at the tip of the 11th rib. The descending thoracic and proximal abdominal aorta was found to have a small diameter of approximately 1 cm. No periarteric inflam-
Inflammatory changes were noted. A 20 mm–diameter, prosthetic tube graft was used to perform an aortoaortic bypass, sutured end-to-side to the proximal descending thoracic aorta above and to the infrarenal abdominal aorta below and passing through a small incision made in the posterior lateral aspect of the diaphragm. The patient’s postoperative course was uneventful, with normalization of the lower extremity pulses. He was discharged on two antihypertensive medications. At 1-year follow-up, the patient no longer needed any antihypertensive therapy and had no further episodes of epistaxis.

Case 2. An 18-year-old South American man was seen at an outside clinic with sudden onset of severe headaches. In addition, the patient had vague postprandial abdominal pain and one block claudication affecting both legs. His condition was found to be hypertensive, with a brachial blood pressure of 180/110 mm Hg, and the patient was started on two antihypertensive medications. He then was referred for vascular consultation. Physical examination revealed an 80-kg, well-built man. Pulses were normal in the upper extremities and diminished in the lower extremities. No abdominal bruit was noted. Brachial blood pressures were elevated (175/105 mm Hg on the right and 180/100 mm Hg on the left), and the thigh blood pressures were 125/70 mm Hg. Serum blood urea nitrogen and creatinine levels were normal (16 mg/dL and 0.9 mg/dL, respectively). Renal artery duplex scan revealed normal renal artery velocities and kidney sizes. Chest roentgenogram and electrocardiogram were both unremarkable. CT scanning showed marked narrowing of the descending thoracic aorta distal to the left subclavian artery with no surrounding inflammatory changes on cross-sectional views (Fig 3). Arteriography revealed severe diffuse narrowing of the aorta from the mid-descending thoracic (Fig 4) to the suprarenal aorta associated with an area of severe stenosis in the celiac trunk and an occluded superior mesenteric artery. The inferior mesenteric artery was patent with good collaterals to the superior mesenteric artery.

The operative course was similar to that of the previous patient, with an end-to-side aortoaortic bypass from the proximal descending thoracic aorta to the infrarenal aorta with a 22 mm–diameter prosthetic tube graft. The postoperative course was uneventful. The patient had normalization of the lower extremity pulses, and he was discharged on one antihypertensive agent. At 1-year follow-up, he no longer needed antihypertensive therapy. The headaches had resolved, and the patient no longer had claudication or postprandial abdominal pain.

DISCUSSION

Hypoplasia has been described in all portions of the thoracic and abdominal aorta. Hypoplasia that involves both the thoracic and abdominal portions is extremely rare and has been previously described in case reports totaling only 16 patients. Of these 16 patients, 13 were female (81%), the ages ranged from 3 to 49 years, seven had associated renal artery stenosis (44%), and nine had aortoaortic bypass reconstruction (56%). The etiology of aortic hypoplasia is poorly understood. Possible mechanisms that have been proposed include developmental defects, a response to infection, and inflammation.
Most authors favor a developmental defect from the frequent association of hypoplasia with other anomalies and the absence of any obvious inflammatory or atherosclerotic changes. Duplicated renal arteries are seen in more than 70% of patients with abdominal aortic coarctations. One proposed developmental mechanism for the occurrence of duplicated renal arteries is the reduction of infrarenal aortic blood flow caused by the coarctation. With normal aortic blood flow conditions, only the principle renal artery would have a hemodynamic advantage and the other renal arteries would undergo apoptosis or involution.

During development, the abdominal aorta is formed at approximately the 25th day by fusion of the paired embryonic dorsal aortae. Incomplete fusion may lead to kinking of the aorta, resulting in a permanent constriction. Conversely, overfusion of the primitive dorsal aortae may occur, including the origins of the posterior aortic branches. That the latter mechanism may contribute to the development of aortic hypoplasia is supported by studies that have shown decreased aortic diameters in patients with single origins for the lowest pair of lumbar arteries. Growth of a developmentally normal aorta may be disturbed by an acquired insult, either in utero or during early life. Certain viruses, notably rubella, may interfere with cell growth and may have direct cytopathic effects on developing tissue. Support for this mechanism in the development of aortic hypoplasia is the observed association between hypoplasia and other manifestations of the rubella syndrome. It is thought that viral inhibition of smooth muscle cell organization and proliferation may prevent normal aortic growth. Finally, an inflammatory process has been suggested because there may be an association with arteritis, such as Takayasu’s disease. However, little evidence exists of an inflammatory process in most cases. Neurofibromatosis, fibromuscular dysplasia, syphilis, tuberculosis, and radiation therapy at an early age have all been associated with hypoplasia of the abdominal aorta, but no causal relations have been identified.

Two classification systems have been developed to describe the variations of the hypoplastic segment with respect to the renal arteries. Ben-Shoshan, Rossi, and Korns proposed an anatomic classification system on the basis of two major coarctations groups: hypoplastic and segmental. Each major group was subdivided on the basis of the location of the affected aorta in relation to the renal arteries: suprarenal, interrenal, and infrarenal. Hallett et al developed a classification system on the basis of the relationship of the coarctation to the renal arteries (suprarenal versus infrarenal coarctation) and whether renal artery stenosis was present or absent. These classifications systems help define the extent of disease and the operative plan. Hypoplasia of the abdominal aorta is slightly more common among females, who account for roughly 60% of cases. Most patients with aortic hypoplasia are seen in the second or third decade of life.

Hypoplasia of the suprarenal or perirenal aorta most commonly is seen with hypertension and its sequelae, including fatigue, headaches, and epistaxis. The hypertension is most likely the result of global renal hypoperfusion and activation of the renin-angiotensin system.
tension is typically severe and, if left untreated, can result in life-threatening complications, such as stroke and heart or renal failure. Bjork and Intonti26 found that the average age of death was 30 years in their series of patients with aortic hypoplasia and hypertension who were managed medically. Patients with hypoplasia limited to the infrarenal aorta typically are seen with lower extremity claudication. Hypoplasia of the infrarenal segment may be associated with the early development of atherosclerosis in the aortic wall.7,19 Although up to 22% of patients with hypoplasia of the abdominal aorta have occlusive disease of the celiac and superior mesenteric arteries, mesenteric ischemia is uncommon, presumably because of collaterals from an intact inferior mesenteric artery. Physical examination of patients with abdominal aortic hypoplasia usually reveals normal upper extremity pulses, diminished pulses in the lower extremities, and occasionally an abdominal bruit. Blood pressure in the upper extremities is usually significantly greater than in the lower extremities.

Any young patient with hypertension should be evaluated for aortic coarctation or hypoplasia, which may be suspected on the basis of the history and physical examination. Historically, aortography has been the diagnostic test of choice.11,18,22 Aortography not only shows the location and extent of the aortic disease but also may reveal associated abnormalities of the visceral and renal vessels. Direct measurement of pressure gradients can be performed to assess the hemodynamic significance of stenotic lesions.22 Newer generation CT scanning and magnetic resonance imaging/angiography have been used with increasing frequency to delineate vascular anatomy. The cross-sectional images that these methods can provide may give additional information regarding the presence of any associated inflammatory changes that angiography cannot evaluate. With true hypoplasia, no inflammatory changes can be shown. Laboratory studies to assess for the presence of arteritis are nonspecific, and the presence of aortic wall thickening on CT scan imaging is a better predictor of an inflammatory process.

The prognosis of aortic hypoplasia is generally poor in untreated cases, depending on the location with respect to the renal arteries, the functional significance of involved arterial branches (ie, renal and visceral vessels), and the severity and complications of the associated hypertension.11 The treatment of choice is surgical revascularization. The operation must be individualized on the basis of the length of the hypoplastic segment and associated renal or visceral artery involvement. Options include resection of the involved segment and end-to-end anastomosis or placement of an interposition graft, which is generally more appropriate for segmental coarctation rather than hypoplasia. For patients with longer segments of hypoplasia, bypass grafting around the narrowed segment is more appropriate. End-to-side proximal and distal anastomoses allow preservation of important side branches that may originate from the narrowed segment, such as the intercostal arteries. The bypass graft should originate from uninvolved aorta proximally, usually the descending thoracic aorta. Origination from the ascending aorta has also been described.27 The distal anastomosis can generally be made to the infrarenal aorta. In patients whose coarctation does not involve the perirenal aorta, anastomosis to the infrarenal aorta improves renal artery blood flow via retrograde aortic flow.28 In patients with coarctation involving the visceral or renal arteries, bypass to these vessels may also be necessary. The use of moderate total body hypothermia as an adjunct to bypass grafting has been described to minimize spinal complications in patients without collateral circulation.9 Also, bypass of the aorta with an extramammary axillofemoral bypass graft has been described with a good surgical outcome.9 Balloon dilatation has been used for segmental coarctation of the thoracic aorta.29,30 However, in patients with longer segments of hypoplasia, this method is not likely to achieve a favorable outcome.

CONCLUSION

Hypoplasia of the aorta refers to nonatherosclerotic and noninflammatory narrowing of the aorta and is a rare vascular lesion that typically presents between the first and third decades of life. It tends to be slightly more common among females, and the usual presenting symptoms are hypertension, claudication, and visceral vessel ischemia. Surgical bypass graft is the optimal management strategy for this condition to relieve the systemic hypertension and to restore circulation to the lower extremities, renal arteries, and visceral organs. A review of the literature found a limited number of case reports (16 patients) on combined hypoplasia of the thoracic and abdominal aorta.2,11,16-17 We report our experience of two such patients.

REFERENCES


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