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## **Abdominal Aortic Coarctation and Hypoplasia**

Developmental narrowing of the distal thoracic and midabdominal aorta usually becomes evident during the first or second decade of life. There is no gender predilection. Multiple renal arteries to one or both kidneys occur in more than 70% of these children. Stenoses of these latter arteries causing renovascular hypertension and narrowings of the celiac and superior mesenteric artery are common.

The classic clinical presentation includes severe hypertension, diminished or absent femoral pulses, and an abdominal bruit, with 25% experiencing lower extremity claudication or exercise-related fatigue. Untreated patients do poorly, with death from cardiac failure or stroke often occurring before age 40 years.

Thoracoabdominal bypasses employing ePTFE or knitted Dacron, in conjunction with renal or splanchnic revascularization, were the most frequent procedures undertaken in the past. Contemporary treatment favors a primary patch aortoplasty with ePTFE and, if needed, direct implantation of the renal or splanchnic arteries into the native aorta. The timing of aortoplasty must take into consideration age-related size limits. An older child is likely to receive long-term benefits from a single operation. Infants less than 2 years of age may require reoperation when older to correct subsequent narrowings because of the small patch or bypass used initially. Percutaneous angioplasty with or without stenting has not been advocated by experienced interventionists because of the truly hypoplastic nature of these vessels. In a collective review, performance of 42 thoracoabdominal bypasses, 13 aortoplasties, and 18 miscellaneous aortic reconstructive procedures, accompanied by concomitant renal artery reconstructive procedures or primary nephrectomy in nearly a third of patients, the reported operative mortality was 8% and excellent or good results occurred in 89% of surviving patients. At the University of Michigan, thoracoabdominal bypass or patch aortoplasty in 24 children, combined with splanchnic and renal arterial reconstructions, yielded salutary results in 91% with no operative mortality.

## **Abdominal Aortic Aneurysms**

Abdominal aortic aneurysms in children are often associated with congenital cardiovascular malformations, most notably bicuspid aortic valves and aortic coarctation; connective tissue disorders including Marfan syndrome, Ehlers-Danlos syndrome, and tuberous sclerosis; infection, most notably following umbilical artery catheterizations; and inflammatory processes accompanying an aortoarteritis.

Clinical manifestations range from asymptomatic lesions recognized during imaging for other illnesses to exsanguinating hemorrhage following rupture. Few childhood aortic aneurysms are thought to be benign. Operative intervention should be considered soon after the diagnosis is established. Rupture may necessitate ligation of the aorta with reliance on collateral circulation to the pelvis and lower extremities, but such is not a logical therapeutic option. Closed or open aneurysmorrhaphy may be appropriate in very young infants when no form of arterial reconstruction may otherwise be possible. Interposition grafts of either Teflon or knitted Dacron have been successful, and in some instances a patch aortoplasty has sufficed. Reoperation may be required as the child grows and the blood flow through the aortic reconstruction becomes inadequate. Fewer than 50 preadolescent children have been reported to have undergone operation for aortic aneurysms.

## **References**

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