Chapter

60 Coarctation of the Aorta

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A 16-day-old female presents for a sedated echocardiogram. She was at the pediatrician for a well-child visit when the physician noted weaker femoral pulses compared with her radial pulses. Vital signs are: BPs: RUE: 98/60 LUE: 62/31 RLE: 66/32 LLE: 66/30; HR 126/min; RR 25/min; SpO_2 100% on room air. Weight 4.2 kg. Physical examination was notable for excessive tissue over the posterior neck. Chest X-ray reveals prominent pulmonary vascular markings.

What Is Coarctation of the Aorta?

Coarctation is simply a narrowing of the aorta. Most commonly, the narrowing is present at the site of the ductus arteriosus attachment (or former attachment; Figure 60.1). Less commonly, the coarctation appears in the ascending aorta and is also known as a preductal coarctation. Coarctation may present in the neonatal period (first 30 days of life) or at any time following. Sometimes it is undiagnosed until later in life. Coarctation may be associated with other congenital heart defects, such as hypoplastic left ventricle (LV), ventricular septal defect (VSD), or atrial septal defect (ASD). Up to 30% of children with Turner syndrome have coarctation.

What Are the Clinical Manifestations of Turner Syndrome?

Turner syndrome is a genetic mosaicism in which part or all of an X chromosome is lost. These patients can be monosomy X or contain a chromosomal mosaicism (e.g., 45,X/46,XX). Common clinical signs are:

- Wide-spaced nipples
- Short stature
- Congenital lymphedema of hands and feet
- Webbed neck
- Low hairline
- Hearing loss
- Ovarian insufficiency
- Hypertension
- Prolonged QT interval
- Renal anomalies
- Cardiovascular anomalies
- Metabolic syndrome

What Are the Common Cardiac Issues Affecting Patients with Turner Syndrome?

Cardiovascular anomalies are exceedingly common in patients with Turner syndrome. In addition to coarctation, aortic valve anomalies, especially bicuspid aortic valves, are present in as many as 30%. These patients are also at a higher risk for progressive aortic dilation and aortic dissection.

What Are the Major Hemodynamic Consequences of a Preductal Coarctation?

Coarctation of the aorta essentially functions as an LV outflow tract obstruction. Severe coarctation may result in LV failure and poor distal perfusion.

Critical coarctation in infancy generally requires early intervention. The patient is stabilized with prostaglandin to promote ductal patency which allows for perfusion to the lower extremities. Inotropic agents are administered when needed to improve myocardial contractility, especially in the setting of heart failure.

What Is Differential Cyanosis?

In the absence of an intracardiac mixing lesion (e.g., VSD), the upper body is oxygenating via the LV and ascending aorta while the lower body is perfused with deoxygenated blood from the RV via the PDA. This

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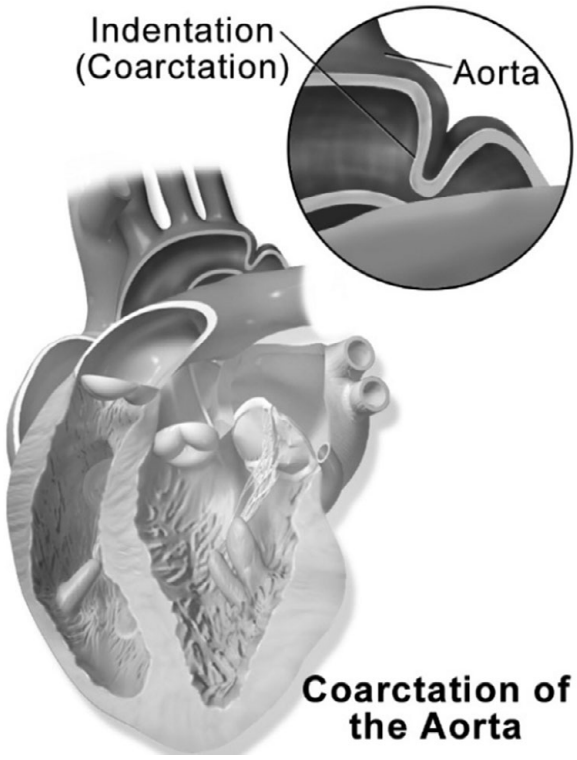

Figure 60.1 Pictorial illustration of aortic coarctation which can occur as illustrated or as a circumferential narrowing of the aortic lumen. Image reproduce from Blausen.com staff (2014). "Medical gallery of Blausen Medical 2014". *WikiJournal of Medicine* 1 (2) under CC BY 3.0 license https://creativecommons.org/licenses/by/3.0/

results in a differential cyanosis between the lower and upper extremities.

How Is the Diagnosis of Coarctation Made?

Coarctation is diagnosed prenatally by ultrasound or suspected postnatally based on physical examination findings such as radial-femoral pulse delay or discrepant blood pressures particularly between the right upper extremity and lower extremities. Transthoracic echocardiography can often identify the region of coarctation, especially in the neonate as well as resultant LV hypertrophy. Gradients across the narrowing are estimated by Doppler to assess the severity of the narrowing. In the setting of LV failure, the estimated gradient may be artificially low. In larger children, CT angiography or cardiac MRI may be done to assess the location and extent of the coarctation and resultant collateral vessels. Cardiac catheterization allows for direct measurement of pressures pre- and post-narrowing and calculation of the true gradient.

Describe the Setting in Which Administration of Prostaglandin E1 (PGE1) Is Indicated in Children with a Coarctation

In cases of preductal coarctation (often with a co-existing hypoplastic aortic arch) identified at birth, flow to the descending aorta may be dependent on blood flow through a PDA. If the PDA closes in the first days of life, blood flow past the coarctation may be restricted and the child may suffer a shock-like condition of poor tissue perfusion. Administration of PGE1 as a continuous infusion will maintain patency of the PDA and enable distal tissue perfusion until the coarctation can be surgically dilated.

Initiation of PGE1 causes central apnea in 15–20% of neonates and thus requires close monitoring. Boluses of PGE1 may result in hypotension or seizure activity. Due to the long half-life of prostaglandin, the infusion is often stopped before transport to the operating room for definitive treatment to avoid accidental bolus dosing.

Continued dilatation of the PDA will also allow for decompression of the pulmonary circulation and RV. When the ductus remains patent, a pressure gradient between the upper and lower extremities may exist (Figure 60.2).

What Are the Surgical Approaches to Repair an Aortic Coarctation?

The main surgical approach is an end-to-end anastomosis. This approach allows the surgeon to avoid graft material (which does not grow with the child) and it avoids division of the subclavian artery (an older technique). The end-to-end anastomosis technique allows for the resection of any remaining ductal tissue that may serve to be a lead point for a future restenosis. The disadvantage of the end-to-end anastomosis is a 10–15% rate of restenosis. Balloon dilation can be used to dilate areas of restenosis, however there is an increased risk of subsequent aortic aneurysmal formation.

The other method of repair is placement of an interposition graft. This technique utilizes a graft that closes the gap after the narrowed section of the aorta

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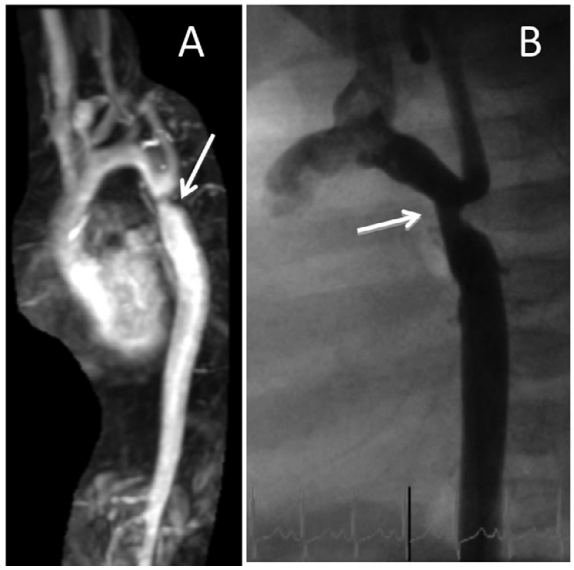

Figure 60.2 (A) Computed tomographic angiography demonstrating a distinct area of coarctation in the "juxta-ductal" region of the aorta distal to the left subclavian artery (arrow). (B) Cardiac catheterization with area of narrowing in the proximal segment of the descending aorta (arrow)

is resected. This may be necessary in older children in whom a larger segment of the aorta is narrowed, resulting in an area too long for end-to-end anastomosis without tension on the sutures. This technique is generally reserved for patients large enough to get a full size (diameter) aortic tube graft.

The main surgical approach is via a left chest thoracotomy. One-lung ventilation is generally unnecessary, as the surgeon will usually retract the left lung for visibility. Alternatively, the approach may be via sternotomy and utilization of cardiopulmonary bypass (CPB). This may be required due to the location of the coarctation or the need for work on the aortic arch or other cardiac defects.

What Are the Major Anesthetic Considerations for Coarctation Procedures?

With the proximity to the great vessels and potential for brisk bleeding, cross-matched blood should be at the bedside, with a warmer immediately available for transfusion. Arterial access, while beneficial for monitoring and sampling, is not necessarily required for this procedure and is largely institution dependent. Ideally, a monitoring device should be applied to each extremity in the form of a NIBP cuff or pulse oximeter to assess for residual gradients postoperatively. Heparin is generally given during cross-clamping with the dose being institution dependent. A cooling blanket can be employed to avoid hyperthermia during aortic cross-clamp.

Older patients develop collaterals to bypass the area of coarctation. These collaterals increase spinal cord and distal perfusion during aortic cross-clamping and reduce the incidence of hemodynamic changes generally associated with cross-clamping. However, large collaterals can pose an increased bleeding risk during thoracotomy.

Postoperative pain after thoracotomy may be severe. PCA should be used for older developmentally appropriate patients. Some centers prefer regional anesthesia in the form of paravertebral single-shot injection or placement of a catheter.

What Are the Most Common Side Effects or Complications Associated with Coarctation Repair?

- The most catastrophic complication of coarctation repair is paraplegia resulting from ischemia of the distal aorta and spinal cord blood supply. The risk is greater in the absence of collaterals and with prolonged cord clamp time (>20–30 minutes);
- Hyperthermia during aortic cross-clamp is associated with poor neurologic outcome. Postoperative hypertension may occur and is treated with nicardipine or nitroprusside (institution dependent);
- Postoperative abdominal pain from mesenteric reperfusion;
- Esophageal damage or accidental ligation;
- Injury or ligation of the recurrent laryngeal nerve;
- Accidental ligation of other vessels;
- Chylothorax from damage to lymphatic networks crossing the pleura of peri-aortic regions;
- Residual aortic coarctation (residual gradient of > 20 mmHg).

What Is Re-coarctation?

The incidence of recurrent coarctation is approximately 5–20%. Intervention is often reserved for re-coarctation

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with residual gradients of >20 mmHg by cardiac catheterization or 35 mmHg by echocardiography. Initial intervention for re-coarctation is generally via balloon dilatation.

Suggested Reading

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