

# Aortic Coarctation and Interrupted Aortic Arch

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Coarctation of the aorta (CoA), aortic arch hypoplasia, and interrupted aortic arch (IAA) are congenital conditions where a portion of the aorta (not including the aortic valve) measure small for BSA or a segment of the arch is atretic or missing. These lesions can present in the newborn period, childhood, and more rarely, in the adult period. Timing of presentation is dependent on the severity of the lesion (degree and extent of arch narrowing) as well as the presence of a PDA and aortopulmonary collaterals. Arch lesions may also be accompanied by other left-sided obstructive lesions including an abnormal aortic valve, mitral valve, or LV. For multilevel left-heart hypoplasia, see Chapter 26.

The aortic arch is usually divided into different segments (Figure 25-1):

- **Proximal arch:** between the innominate artery and left carotid artery
- **Distal arch:** between the left carotid artery and the left subclavian artery
- **Isthmus:** between the left subclavian artery and the ductus arteriosus or ligamentum arteriosum

The dimensions of each of the aortic arch segments are important to define the clinical presentation and management of patients with aortic arch hypoplasia. Focal CoA classically presents with significant narrowing at the level of the isthmus.

IAA is classified into 3 types depending on the aortic arch segment involved with the interruption:

- **Type A:** the interruption is distal to the left subclavian artery
- **Type B:** the interruption is between the left common carotid artery and the left subclavian artery
- **Type C:** the interruption is between the innominate artery and the left common carotid artery

## Pathophysiology and Clinical Presentation

### Fetal

Mild aortic arch lesions are often difficult to detect in utero, and may only be detected postnatally, after the PDA closes. On the contrary, if there is significant ascending, transverse, or isthmus arch narrowing, there will be retrograde flow in the aortic arch, which can be detected on fetal echocardiogram.

IAA can also be detected in utero, when a fetal echocardiogram reveals a lack of communication between parts of the transverse and descending aorta. However, the type of interruption may be difficult to determine depending on the size, position, and gestational age of the fetus.

Regardless of severity, there are typically minimal clinical fetal manifestations of arch obstruction in utero due to the patency of the PDA. However, it is critical to determine which arch lesions will be severe or critical (such as the case in IAA), as it will help

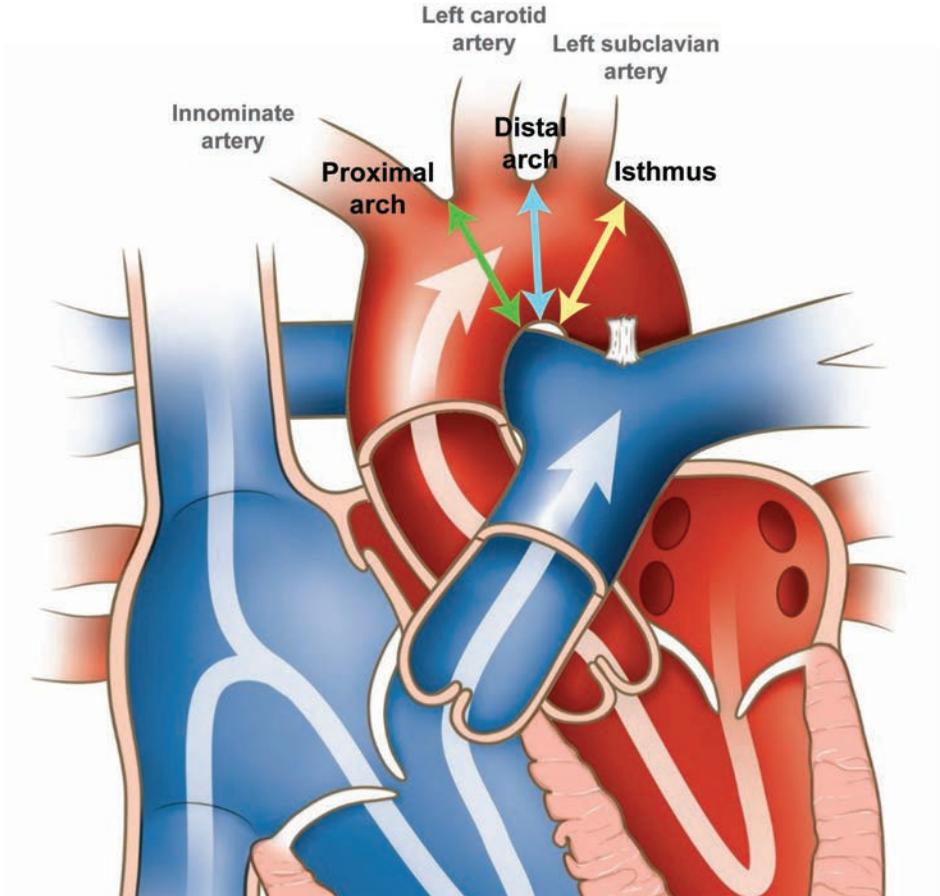


Figure 25-1. Aortic arch segments.

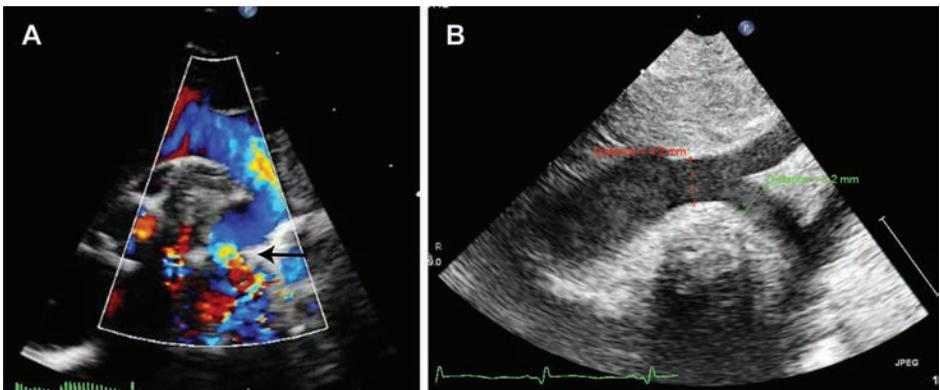
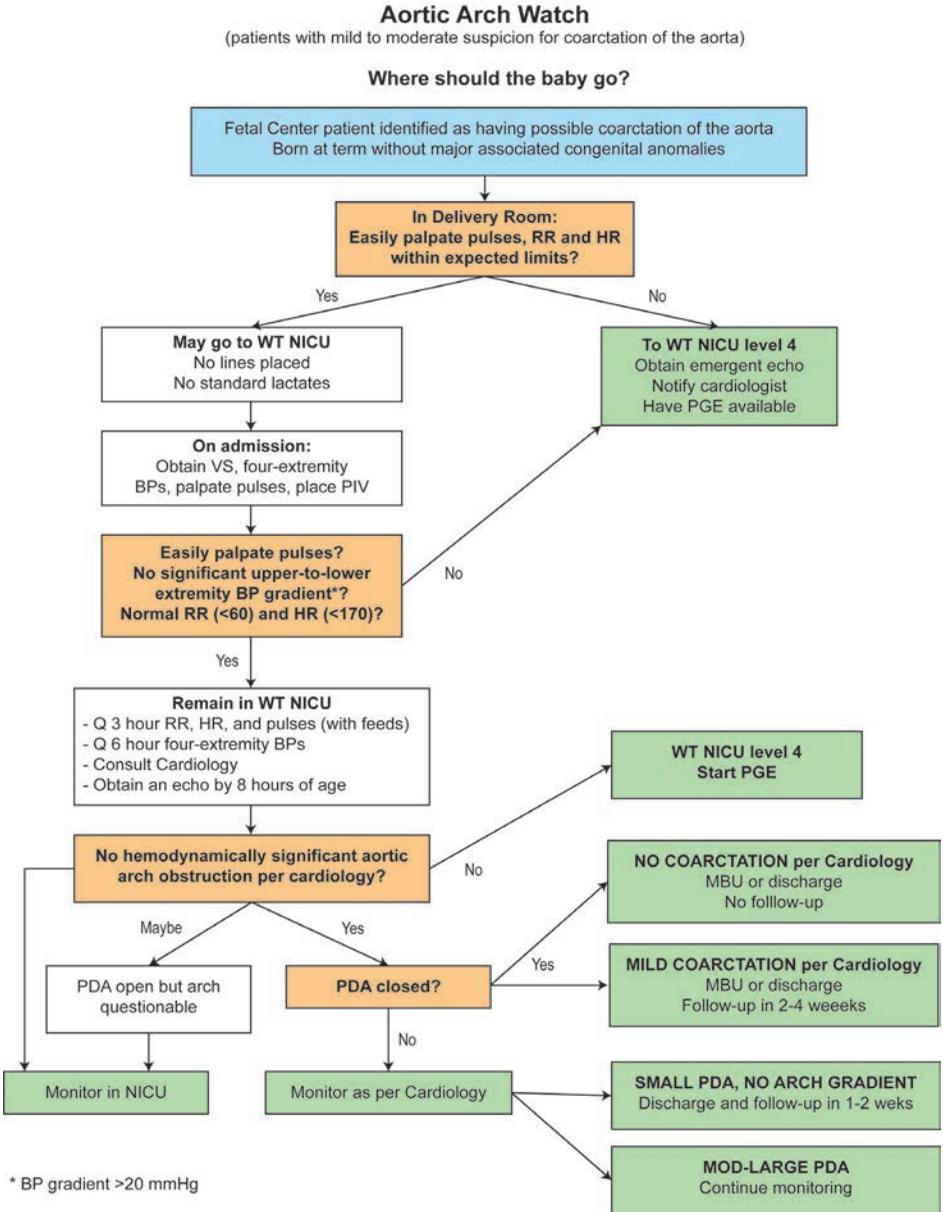
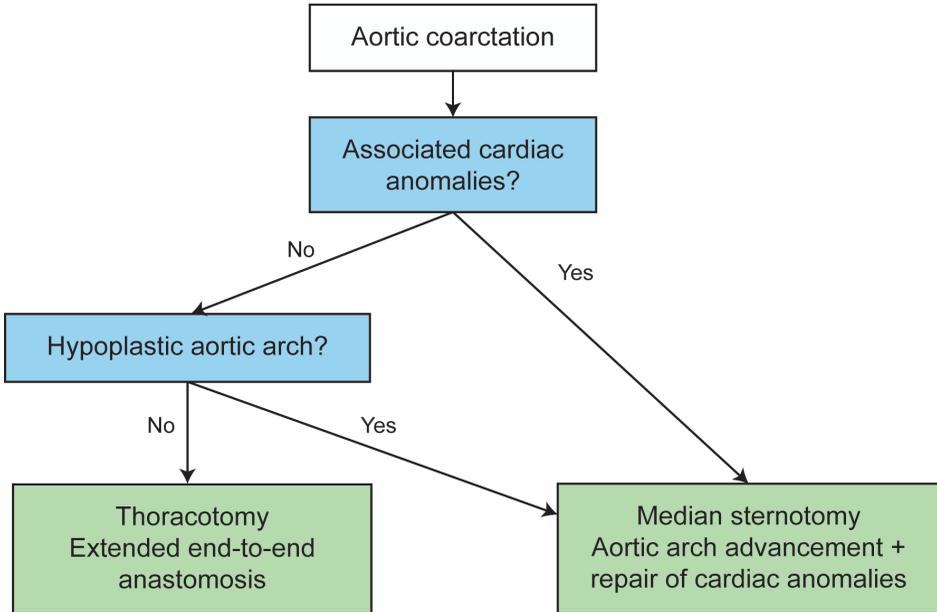


Figure 25-2. Echocardiographic images of a patient with an isolated aortic coarctation (A, arrow) and a patient with a hypoplastic aortic arch (B).



**Figure 25-3.** “Arch-watch” protocol at TCH. BP: blood pressure, HR: heart rate, MBU: Mother and Baby Unit, NICU: Neonatal Intensive Care Unit, PDA: patent ductus arteriosus, PGE: prostaglandin E, PIV: peripheral intravenous line, RR: respiratory rate, VS: vital signs, WT: West Tower.



**Figure 25-4.** Algorithm for management of CoA and hypoplastic aortic arch in newborns and infants.

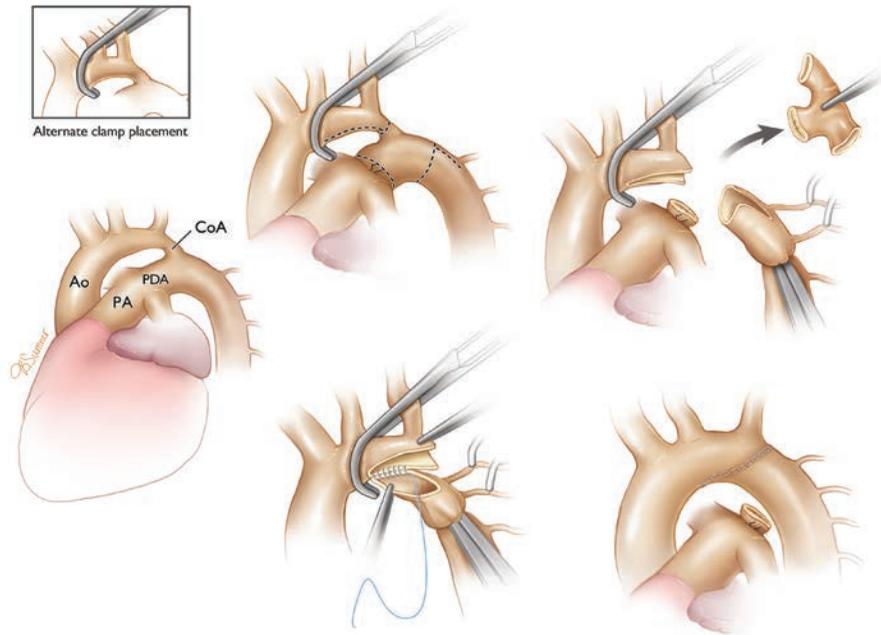
dictate immediate postnatal care including location of delivery and initiation of PGE to maintain patency of the ductus.

### Neonatal

The clinical presentation of aortic arch lesions after birth will depend on the degree of obstruction. Mild CoA may not present in the neonatal period, as it typically allows adequate blood flow to the body, making it clinically less obvious. Often, these milder lesions are discovered and/or become clinically apparent during childhood (see Childhood/Adolescence below).

On the contrary, most patients with severe CoA and IAA tend to present in the first 24-72 hours after birth as the PDA closes, unless PGE is started immediately after birth due to prenatal diagnosis. Closure of the PDA can result in significant narrowing of either one or multiple parts of the aortic arch in addition to impeding flow through the PDA into the distal body. Furthermore, the neonatal myocardium is uniquely sensitive to increased afterload and any significant narrowing of the arch can cause a severe strain on the LV, potentially leading to LV dysfunction.

Early in the course of PDA closure (often first 12-24 hours), it may be noted that femoral and pedal pulses are diminished, or that patients have a BP gradient difference between upper and lower extremities. Compromise to blood flow distal to the PDA may not be very clinically obvious at this point. If detected later in the neonatal course (after 24-72 hours), patients may have severe compromise of blood flow distal to the



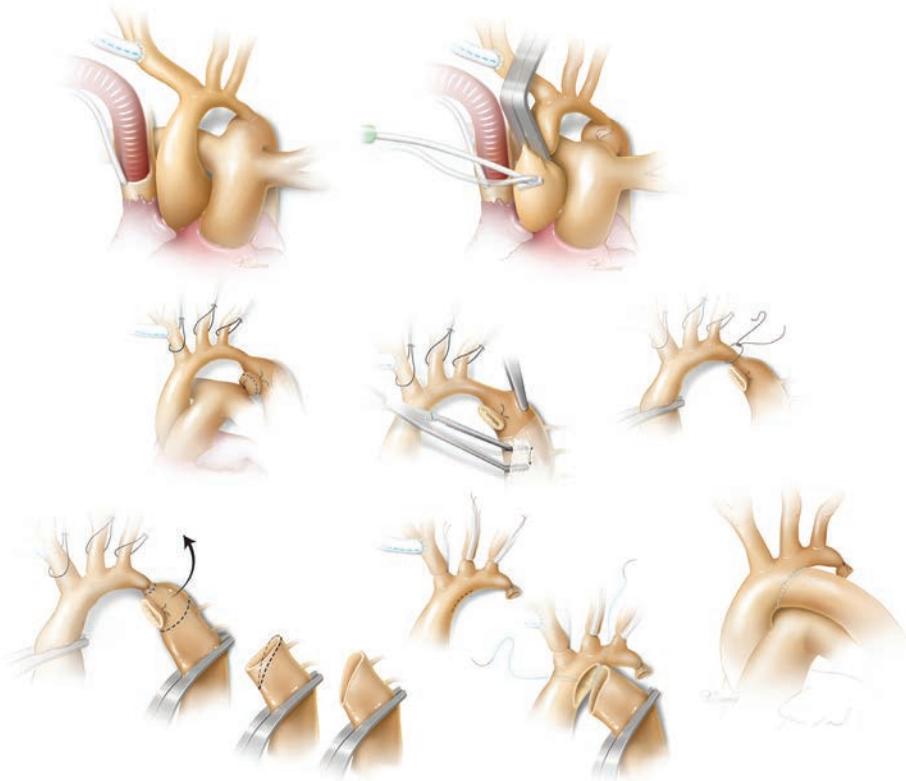
**Figure 25-5.** Coarctectomy and extended end-to-end anastomosis repair of CoA. The aortic arch, brachiocephalic vessels, descending aorta, and ductus arteriosus / ligamentum arteriosum are fully mobilized. The intercostal vessels / collaterals are controlled with fine tourniquets. A Castaneda clamp is used to clamp the left subclavian artery and the aortic arch. An angled or straight clamp is placed on the descending aorta. The ductus arteriosus is divided and the coarctation segment with all ductal tissue is removed. The proximal incision is extended into the undersurface of the aortic arch. The distal incision is extended posteriorly. A large anastomosis is created between the descending aorta and the aortic arch with polypropylene suture (7-0 for newborns). The clamps are released.

area of narrowing or PDA, and they may present with poor feeding, tachypnea, lethargy, irritability, and cardiogenic shock with acidosis and multisystem organ failure within the first week of life.

### Childhood / Adolescence

Aortic arch obstructive lesions that present in childhood and adolescence are typically less severe and there is time for collateral flow to develop to deliver blood to the lower body despite obstruction in the aortic arch. These lesions can also develop as a result of aortic injury due to trauma or inflammatory processes.

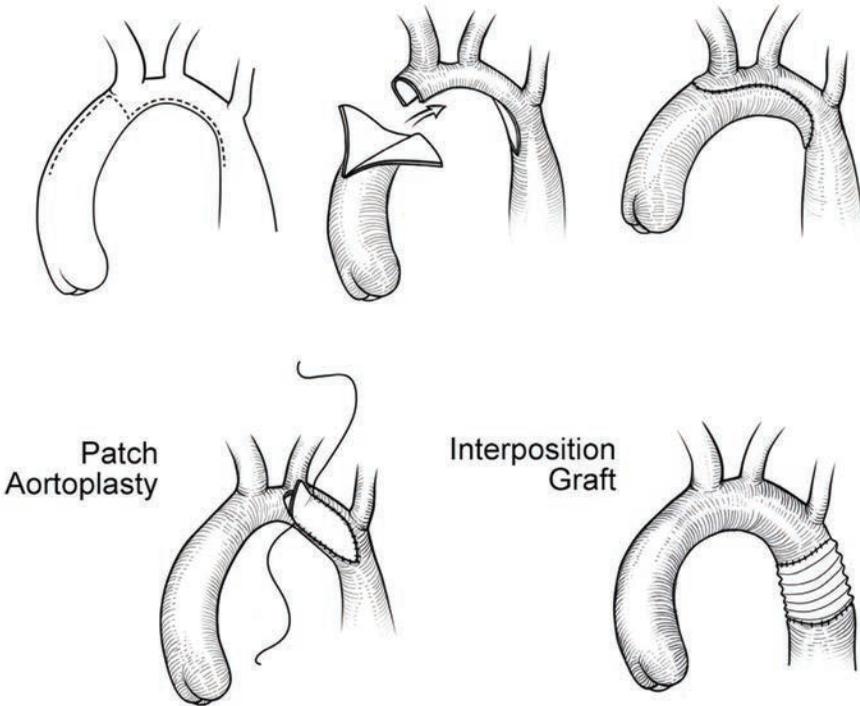
Otherwise asymptomatic patients may present with a murmur or during evaluation for hypertension. Patients may also present with frequent headaches and stomach aches that are worse with activity. On examination, they will have decreased or delayed



**Figure 25-6.** Aortic arch advancement for repair of aortic arch hypoplasia in newborns and infants. CPB is instituted through a graft on the innominate artery and bicaval cannulation. The aortic arch, brachiocephalic vessels, and the descending aorta are widely mobilized as distal as possible to allow adequate mobility. After division of the ductus arteriosus / ligamentum arteriosum, the isthmus is divided and sutured closed. All ductal tissue is removed and the descending aorta is spatulated. An incision is made on the proximal aortic arch and an end-to-side anastomosis is performed between the descending aorta and the arch. It is important to properly align the anastomosis to avoid torsion of the ascending aorta with resultant obstruction.

pulses, though the amount of decrease or delay depends on the amount of collateral vessel flow that has developed over the duration of the obstruction. These patients will also have a significant systolic BP gradient ( $>20$  mmHg) on 4-extremity BP. A systolic murmur that persists into diastole may be heard in the left paravertebral area and represents flow across the area of obstruction. Continuous murmurs may also be heard due to flow through large collateral vessels. Presentation at this age is typically not associated with poor cardiac function or cardiovascular collapse.

### Sliding Arch Aortoplasty

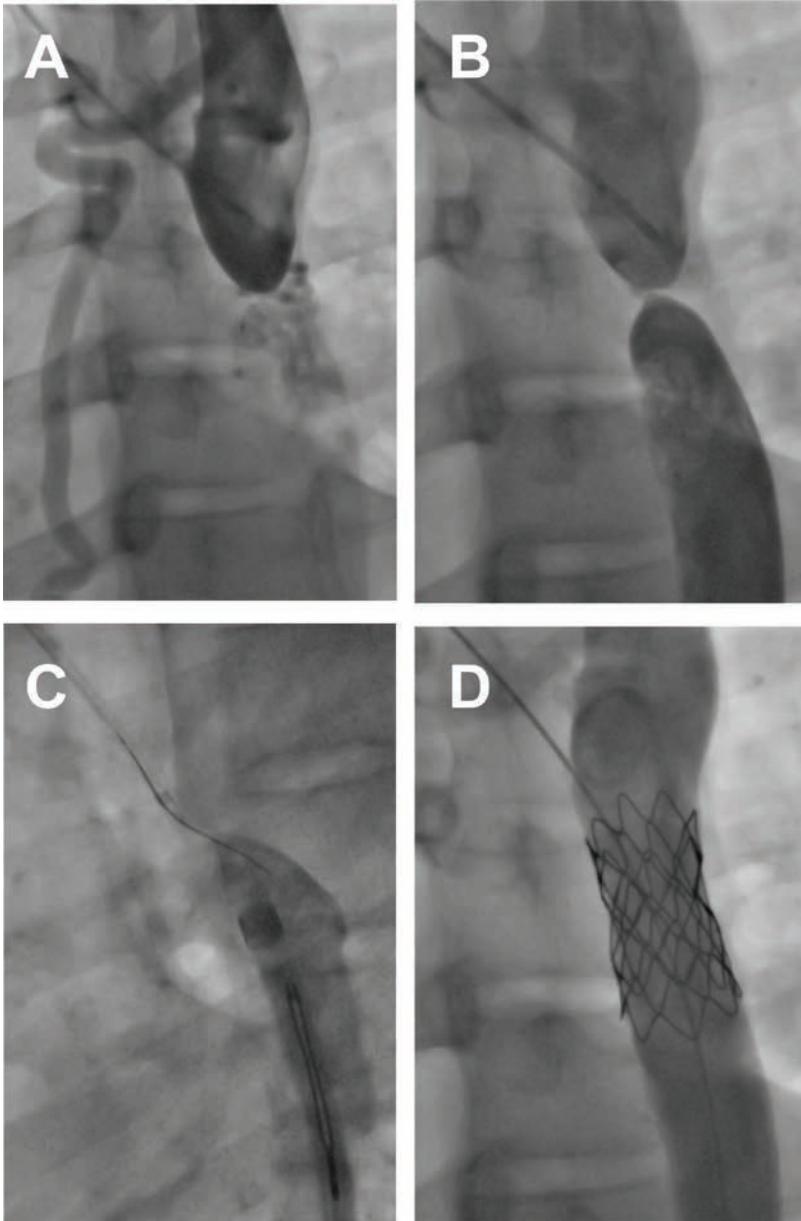


**Figure 25-7.** Alternative surgical approaches for aortic arch reconstruction. The sliding arch aortoplasty is an all-autologous repair in which a tongue of ascending aorta is used to enlarge the aortic arch. If all-autologous repairs are not possible, a patch aortoplasty or an interposition graft placement may be used.

### Diagnosis

- **Fetal echocardiogram.**

- *Mild CoA.* There can be subtle signs on fetal echocardiogram that suggest a possible CoA (often mild) ex utero, including persistent color flow in the area of the mild arch narrowing, and borderline or mildly small measurements of the aortic arch for gestational age by 2D. Presence or severity of a potentially mild CoA in these cases cannot be fully realized until postnatal assessment, as the PDA closes and BP gradients and femoral/pedal pulses can be assessed.
- *Severe CoA, moderate-to-severe tubular hypoplasia, and IAA.* These lesions can be detected in utero by fetal echocardiogram. In the case of severe CoA and moderate-to-severe tubular arch hypoplasia, arch 2D measurements are typically small for gestational age and retrograde flow is noted in the narrowed aortic arch by color and pulse-wave Doppler. Arch sidedness is able to be determined



**Figure 25-8.** Angiograms of a patient with near atresia of the aorta at the level of CoA. A) Note extensive arterial collateral vessels typical in severe aortic obstruction. B) Simultaneous injections are performed above and below the obstruction demonstrating the severity of the CoA. C) A wire is passed through the narrowed segment, to be snared from below. D) Angiography after placement of a covered stent demonstrating normal caliber of the treated area.

in most cases. In the case of an IAA, the ascending aorta typically takes a very linear course superiorly and does not connect with the descending aorta, which arises from a large PDA. It can be very challenging to determine arch sidedness prenatally in a patient with IAA.

- **Pulse oximetry.** Unlike other cyanotic congenital heart lesions that present in the neonatal period, pulse oximetry screening cannot accurately detect CoA (whether mild or severe). However, in the setting of IAA, pulse oximetry screening can detect differential cyanosis (higher  $\text{SaO}_2$  levels in the arms than in the legs).
- **CXR.** In the neonatal period, CXR is not very helpful for diagnosis of a CoA, although it may help with determination of arch sidedness. In the childhood/adolescent period, where a CoA may have been present for some time and aortopulmonary collaterals exist, CXR may demonstrate proximal rib notching along the sternal border. Additionally, with later presentation, one may see the “3” sign where there is indentation in one portion of the aorta where the localized narrowing exists.
- **ECG.** In the childhood/adolescent period there may be LV hypertrophy.
- **Echocardiogram (Figure 25-2).** Mainstay of diagnosis. Evaluation should include arch sidedness, arch branching, and measurements of the ascending aorta, proximal and distal transverse aortic arch, and aortic isthmus. A complete evaluation of the aortic arch must be performed. The primary goal is to define the site and extent of obstruction with additional focus on evaluation of function, myocardial changes, and other coexistent lesions (e.g., bicuspid aortic valve, small left-heart structures, VSD). Multiple views must be obtained to provide adequate definition. Retrograde flow may be noted in the narrowed aortic arch by color and pulse-wave Doppler if there is a PDA or if there are collaterals supplying flow in retrograde fashion to a portion of the transverse aortic arch. Color-Doppler echocardiography can show increased turbulence at the site of narrowing and spectral Doppler can show increased velocity of flow through the area. The abdominal aorta should be evaluated to determine if there is flow continuation throughout the cardiac cycle. Reduced or delayed systolic amplitude with flow that continues into diastole would be concerning for aortic arch obstruction. Additional images should be obtained to evaluate cardiac function and provide function estimates. One can also ascertain injury to the myocardium by looking for echobrightness suggestive of endocardial fibroelastosis. In older patients, the arch may not be well seen and alternative imaging modalities may be necessary.
- **CTA/MRI.** May be useful in cases where the degree of arch narrowing is unclear, arch sidedness cannot be determined, or arch anatomy cannot be fully delineated by echocardiogram (as is often in the case in late presenting CoA with several aortopulmonary collaterals). In patients with IAA and unclear anatomy on echocardiogram, CTA/MRI may be helpful in identifying the type of interruption, distance between the proximal and distal segments, and other defects that may interfere with surgical planning (including anomalous subclavian arteries and arch sidedness).
- **Genetics.** Arch anomalies may be associated with genetic abnormalities, such as DiGeorge syndrome. For details on genetic testing, see Chapter 50.

### Indications/Timing of Intervention

In some situations, it is unclear whether a newborn will develop a significant CoA upon closure of the PDA. In those cases, the patient may be placed on an “arch watch”. The purpose of this “arch watch” is to closely monitor hemodynamic and anatomic parameters as the PDA is allowed to close off PGE. If upon PDA closure, the patient develops a significant CoA, PGE is started and the patient is referred for surgical intervention. The “arch watch” protocol at TCH is shown in Figure 25-3.

The presence of a severe CoA and/or aortic arch hypoplasia that is dependent on PGE to keep the PDA open is an indication for intervention. Similarly, the diagnosis of IAA warrants intervention in the newborn period.

Patients that present in cardiogenic shock a few days or weeks after birth due to closure of the PDA are started on PGE in an attempt to reopen the PDA and/or relax the ductal tissue of the isthmus and relieve the obstruction. If PGE is successful in reestablishing blood flow to the lower body, the patient is medically stabilized for a few days to allow end-organ function recovery prior to surgical intervention. If administration of PGE is unsuccessful in reestablishing adequate blood flow, the patient is taken emergently to the OR for intervention.

Patients that present beyond the newborn period with milder forms of CoA still benefit from intervention in order to decrease the risks associated with long-standing hypertension. However, intervention (cath or surgical) can be performed in a more elective fashion.

### Anesthetic Considerations

The anesthetic management of patients with COA is straightforward, particularly in neonates. It is customary to have the ability to monitor BP both proximal and distal to the CoA, either with a right-radial arterial line and a BP cuff on the lower extremity or a right-radial arterial line with an umbilical or femoral arterial line. Be cognizant of the arch anatomy and the possibility of an aberrant right subclavian origin (which will interfere with assessment of proximal aortic pressures). Discuss the surgical approach and the need for invasive arterial pressure distal to the CoA with the surgeon: in very small infants or infants with Down syndrome, the risk may outweigh the benefit.

For CoA repair via left thoracotomy, patients can be allowed to cool during the initial dissection, with core temperatures reaching the 34-35 °C range. Small infants may cool passively in the room, while a larger patient might need more active cooling measures. Likewise, as patients become larger, specialized airway techniques can be used as tolerated to isolate the ipsilateral lung. There is not generally an indication for TEE in CoA repair via thoracotomy.

### Surgical Repair

The ideal type of surgical repair depends on the anatomy of each particular patient (Figure 25-4). In general, an all-autologous repair is favored, avoiding the use of patch or graft material. If the proximal aortic arch is adequate in size, the procedure is performed through a left thoracotomy. On the contrary, if the proximal arch is small and there

**CoA repair via left thoracotomy (1995-2013)** (Mery et al. 2015)

- Median number of procedures done per year: 18 (0-26)
- Median age at surgery: 2 months
- Median hospital length of stay: 7 days (2-194 days)
- Perioperative mortality: 1%
- Reintervention rate (median follow-up 6 years): 4%

**Aortic arch advancement (1995-2012)** (Mery et al. 2014)

- Median number of procedures per year: 14 (0-30)
- Median age at surgery: 14 days
- Vocal cord dysfunction on routine laryngoscopy: 38%
- Clinical vocal cord residual dysfunction at last follow-up: <1%
- Median hospital length of stay: 16 days (5-235 days)
- Perioperative mortality: 1%
- Reintervention rate (median follow-up 6 years): 3%

is concern that addressing the arch through a left thoracotomy would leave a more proximal gradient, or if there are concomitant intracardiac lesions to be addressed, the procedure is performed through a median sternotomy using CPB. In general, for neonates and infants, the proximal arch size in centimeters should be equal or greater than the weight in kilograms of the patient plus 1. For example, a 3.5-kg newborn is expected to

have a proximal arch that measures at least 4.5 cm.

### Coarctation Repair Via Left Thoracotomy

The procedure is performed using a serratus-sparing left posterolateral thoracotomy through the third or fourth intercostal space. The descending aorta, aortic arch (up to the innominate artery), and ductus arteriosus/ligamentum arteriosum are completely dissected with care not to injure the vagus and recurrent laryngeal nerves (RLN) (the RLN wraps around the ductus arteriosus/ligamentum arteriosum) or the intercostal vessels and collaterals that may be quite prominent in this disease. 100 Units/kg of heparin are administered 2-3 minutes prior to clamping.

For newborns and young children, the coarctectomy and extended end-to-end anastomosis (Figure 25-5) is the surgical technique of choice for repair of CoA. In older children and adults, the decreased elasticity of tissues will preclude using this technique. In those cases, other techniques such as simple end-to-end anastomosis, patch aortoplasty, or placement of an interposition graft may be needed.

### Aortic Arch Reconstruction Via Median Sternotomy

In cases of aortic arch hypoplasia or intracardiac anomalies, the procedures are performed through a median sternotomy under CPB. These procedures are usually performed using antegrade cerebral perfusion through a Gore-Tex® graft (3 or 3.5 mm for newborns) that is sutured to the innominate artery after administration of 100 Units/kg of heparin. Bicaval cannulation is commonly used. After institution of CPB, the patient is cooled down to 18 °C and the aortic arch, ductus arteriosus/ligamentum arteriosum, and descending aorta are completely dissected. Once the goal temperature is reached, an aortic cross-clamp is placed and antegrade cardioplegia administered. The RA is opened and the left heart is vented through the atrial septum.

The procedure of choice for newborns and infants is the aortic arch advancement (Figure 25-6) which entails an end-to-side anastomosis between the descending aorta

and the proximal aortic arch. This technique is not used in older patients due to the lack of mobility of the tissues. In those patients, alternative techniques include the sliding arch aortoplasty, patch aortoplasty, or placement of an interposition graft (Figure 25-7).

### **IAA**

In addition to arch interruption, patients with IAA usually have a posterior malalignment VSD and a hypoplastic aortic annulus. However, in the majority of patients, the aortic valve annulus is sufficient to support the systemic circulation (~5 mm in a 3 kg newborn). The surgical repair is performed in the newborn period and entails an aortic arch advancement, resection of the posteriorly malaligned ventricular septum causing subaortic obstruction, and transatrial VSD repair with an autologous pericardial patch. The distance between the ascending aorta and the descending aorta after ductal tissue resection in these patients can be quite significant. As such, in order to reduce tension, it may be useful in some patients to perform a posterior direct anastomosis between the ascending and descending aortas and place an anterior patch of glutaraldehyde-treated pericardium or homograft.

### **Catheter-Based Intervention**

Catheter-based techniques serve an important role in the treatment of children of all ages with aortic obstruction (Figure 25-8). In general, intervention is indicated when the pressure gradient is >20 mmHg. It is also indicated in patients with gradient <20 mmHg in the setting of significant collaterals, univentricular heart, or ventricular dysfunction.

### **Balloon Angioplasty**

Balloon angioplasty involves dilation of the CoA, and is effective in providing relief of obstruction. However, in the native CoA, recurrence of obstruction can occur, especially in younger patients (<6 months), and reintervention can be required. In postoperative recurrent CoA (i.e., after repair for CoA, IAA, or hypoplastic left heart syndrome with a Norwood procedure), relief of obstruction is more durable than in native CoA, and balloon angioplasty is the therapy of choice. Balloon angioplasty is also helpful in providing early relief of aortic obstruction for young infants who have other associated congenital heart lesions, depressed ventricular function, severe MR, or low cardiac output and who will benefit from delay of surgery.

### **Balloon-Expandable Bare-Metal Stents**

Balloon-expandable bare-metal stents provide for safe, more effective, and more durable relief of aortic obstruction than standard balloon angioplasty. They are employed when standard balloon angioplasty does not achieve a durable result, as may happen in cases of intraprocedural vessel recoil or rapid recurrence after standard balloon angioplasty. Smaller pediatric patients who receive stents will undergo reintervention for dilation of the stent as they grow. All efforts are made to adhere to the tenet of implanting stents that are capable of being expanded to adult size.

Stenting is frequently avoided in infants and small children, in whom small vessel size may preclude easily implanting stents with potential to reach adult size. Exceptions to this can occur when individual clinical factors render implantation of smaller stents

necessary (e.g., surgery undesirable in a small patient with recurrent CoA, intimal flap raised during angioplasty). In such cases, these smaller stents may later be overdilated to the point of intentional fracture to keep up with somatic growth. The stented area is then restented with a stent that has adult-size potential.

For lesions near the head-and-neck vessels, open-cell stents can be used. Open-cell stents that are by necessity implanted near or across the orifice of a branch vessel allow for selective dilation of the cell adjacent to the vessel orifice, such that flow through the orifice remains completely unobstructed.

### Covered Stents

Covered stents are composed of a metal stent covered with fabric or graft material such as polytetrafluoroethylene (PTFE). Covered stents are used more frequently in patients that are at higher risk for aortic wall injury, including older pediatric and adolescent/adult patients with severe CoA/near-interruption of the aorta. They may also be used in patients who have already developed evidence of aortic wall injury (e.g., pseudoaneurysm). The covering serves to seal off the affected or potentially injured area. An additional benefit of the covering is the prevention of neointimal ingrowth.

### Postprocedural Management

Approach to postoperative care depends on the procedure performed. In general, appropriate support of end-organ perfusion, control of hypertension, and early diagnosis of any postoperative complications are the most important goals.

Patients following catheter interventions are admitted to CICU for observation overnight focusing primarily on management of hypertension and puncture site complications. Compression safeguards remain in place and must be passively deflated every 2 hours and removed to ensure appropriate extremity perfusion. The extremity used for arterial access must be frequently assessed for signs of bleeding and poor perfusion. In cases of compromised extremity perfusion, anticoagulation therapy with heparin or enoxaparin should be immediately initiated. If poorly responsive to intervention, Doppler study of the vessels should be completed to document the location and degree of obstruction. Any compromise in perfusion leading to loss of pulses or change in color and appearance must be treated as an emergency. Vascular surgery should be involved immediately. Other rare but reported complications related to catheterization include pneumothorax and retroperitoneal bleeding.

Surgical repair through thoracotomy involves an incision through several muscular layers as well as physical compression of the lung to achieve visualization. Postoperative management is focused around close neurologic monitoring, tight BP control, symptoms of lung contusion, bleeding, and management of pain.

Postoperative hypertension warrants aggressive treatment to avoid anastomotic bleeding and is usually multifactorial. The early catecholamine storm-related hypertension is treated with agents such as nitroprusside sodium or nicardipine. In older children, esmolol drip or intermittent labetalol should be considered. Additional therapy with long-term agents such as enalapril or captopril may be initiated on postoperative

days 1-2 to address the elevated renin levels reported in this patient population. Renal perfusion and function should be satisfactory before initiating these agents.

Pain is also a significant and common contributor to postoperative hypertension, particularly in patients undergoing a left thoracotomy. We recommend the use of nonsedating agents such as IV acetaminophen, and patient-controlled analgesia. Other agents to consider include dexmedetomidine drip during the first postoperative night and ketorolac for older children (>1 year) provided the platelet count and renal function are satisfactory.

The management of patients that undergo aortic arch repair through a median sternotomy is similar to other patients undergoing CPB. Older children are commonly extubated in the OR unless other surgical repairs accompany the repair. Newborns and infants usually return from the OR intubated. Once hemodynamic stability is observed within the first 6-12 hours, the patients are progressed to extubation. Attention should focus around symptoms of upper-airway obstruction as manipulation of RLN may lead to temporary paresis or injury. RLN injury commonly presents as stridor or even significant upper-airway obstruction; evaluation is warranted by ENT.

## Complications

### Surgical Repair

The main complications after surgical repair of CoA and hypoplasia of the aortic arch include:

- **Injury to the RLN.** If routine laryngoscopy is performed, approximately 25-30% of patients undergoing repair through a median sternotomy will have left vocal cord paresis due to stretching of the nerve during dissection (Dewan et al. 2012). The vast majority of these patients will recover vocal cord function within a few weeks. However, newborns with RLN injury may exhibit swallowing difficulties and may need thickened formula or nasoenteral feeds until recovery.
- **Chylothorax.** The disruption of lymphatic channels in the thorax, in particular during repairs using a left thoracotomy, may lead to a chylothorax (see Chapter 77).
- **Postcoarctectomy syndrome.** Thought to be a reperfusion reaction of the organs below the prior CoA and is commonly accompanied by abdominal tenderness, feeding intolerance, and leukocytosis at 2-3 days following repair. Appropriate BP control helps prevent these symptoms in the majority of patients.
- **Left mainstem bronchus compression.** Aortic arch reconstruction reduces the window between the ascending and descending aorta, potentially causing compression of the left mainstem bronchus. However, it is very rare for bronchial compression to be significant (<1%).

### Catheter-Based Treatment

Aortic wall injury can occur in catheter-based interventions. This may take the form of early or late pseudoaneurysm formation or dissection; both can be treated with transcatheter techniques. Stent embolization may occur intraprocedurally or in the immediate postcatheterization period. Stent fracture can occur late and results in recurrence of obstruction; this is treated with restenting.

Patients undergoing balloon angioplasty will by definition sustain some degree of intimal tear; such therapeutic tears can sometimes be visualized on angiography. These tears are intrinsic to the treatment strategy and as such, are not considered a complication, but rather as evidence of efficacy.

### Long-Term Follow-Up

Repair of aortic arch obstruction is not without long-term risk. Lifelong follow-up with clinical exam and echocardiographic imaging is needed to continue to monitor and treat issues as they arise. Potential long-term complications include:

- **Hypertension.** The long-term development of hypertension or exercise-induced hypertension in patients after aortic arch repair is high, likely related to an abnormal wall structure causing increased stiffness and poor compliance. The patients more likely to develop postintervention hypertension are those with preintervention hypertension, although most patients will develop hypertension whether or not they had preexisting hypertension. A normal BP in the physician's office does not rule out hypertension as a significant portion of those with normal BP in the clinic will have elevated BP on ambulatory or exercise evaluation. Exercise-induced hypertension is a predictor for developing hypertension in the future. Many who develop hypertension will be resistant to treatment and require multiple medications to control or improve their BP.
- **Recurrent aortic arch obstruction.** Recurrent arch obstruction requires lifelong monitoring. Echocardiogram, MRI, and CTA may be used to evaluate for recurrent obstruction and help define need for reintervention. The incidence of recurrent arch obstruction at TCH after repair through a left thoracotomy or a median sternotomy is approximately 3-4% at a median follow-up of 6 years.
- **Aortic arch aneurysm/dissection.** Aortic arch aneurysms may develop mainly in patients that have undergone arch repairs using patch aortoplasty or subclavian flaps.
- **Premature coronary artery disease.** Patients with aortic arch intervention are at higher risk of developing premature coronary artery disease. Abnormal flow dynamics in muscularized arteries can lead to a high atherosclerotic potential. Additionally, due to abnormal flow dynamics in these patients, there is an increase in LV pressure that can result in LV hypertrophy. Continued evaluation of cardiac hypertrophy and function is important.

### Suggested Readings

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## PART II. DISEASES

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