Coarctation of the Aorta

8

Rachel D. Torok, Michael J. Campbell, Gregory A. Fleming, and Kevin D. Hill

Introduction

Coarctation of the aorta was first described by Morgagni in 1760, and in its simplest form refers to isolated and discrete stenosis of the proximal thoracic aorta. However, coarctation may also be associated with a longer segment of narrowing, with hypoplasia of the transverse aortic arch, or with stenosis of the lower thoracic or abdominal aorta [1–3]. While more severe cases typically present in the neonatal period, aortic coarctation may be diagnosed at any age, either in isolation or in association with other cardiac defects. Crafoord was the first to perform a successful surgical repair of aortic coarctation in 1944 [4]. Since then, various surgical and transcatheter approaches have been developed, which have enabled significantly improved outcomes. In this chapter, we will focus our attention on the etiology, evaluation, and management of coarctation of the thoracic aorta and then discuss the less common presentations of abdominal aortic coarctation and pseudocoarctation.

Prevalence and Etiology

Coarctation of the aorta accounts for 5–7% of all congenital heart disease [5], with an incidence of approximately 3 cases per 10,000 births [6]. Males are more commonly affected than females [5]. Coarctation may be seen in isolation or with additional cardiac lesions, including left ventricular outflow tract lesions, such as bicuspid aortic valve, aortic valve stenosis, and hypoplastic left heart syndrome [7, 8], as well as ventricular septal defect, patent ductus arteriosus, transposition of the great arteries, and atrioventricular canal defects [9–13]. Genetic syndromes including Turner syndrome (45XO), Down syndrome (Trisomy 21), and Jacobsen syndrome (11q terminal deletion) are associated with coarctation of the aorta [14]. In neonates undergoing coarctation repair, Turner syndrome is the most common genetic syndrome (4%), followed by Down syndrome (2.1%) [15, 16]. The exact embryologic development of coarctation of the aorta is unclear, but two main hypotheses exist. One theory is that in utero, ductal tissue abnormally migrates into the aortic isthmus. With ductal constriction after birth, there is also abnormal constriction of the aortic isthmus, leading to coarctation [1]. An alternative hypothesis proposes that decreased blood flow through the ductus arteriosus leads to abnormal growth of the aortic isthmus, which acts as a vulnerable "watershed" region. This abnormal flow could be caused by proximal obstruction to flow in the left ventricular outflow tract or an abnormal angle of entry of the ductus arteriosus at the aortic isthmus [17]. This theory is supported by fetal echocardiography data, which shows a prevalence of transverse aortic arch hypoplasia in fetuses who eventually go on to have coarctation [18, 19].

Diagnosis

Clinical Presentation

Coarctation can present at any age. In the United States approximately 1 in 4 neonates requiring surgical intervention for coarctation is diagnosed prenatally [20, 21]. Neonates with "critical" or ductal dependent aortic coarctation that are not diagnosed prenatally often present with heart failure, acidosis, and shock following closure of the ductus arteriosus. Without prompt medical resuscitation and surgical intervention, death occurs rapidly in these patients [22, 23].

Patients with less severe coarctation may not be diagnosed until later in life and can present with a murmur or

Electronic Supplementary Material The online version of this chapter (https://doi.org/10.1007/978-3-030-11322-3_8) contains supplementary material, which is available to authorized users.

R. D. Torok
Pediatric Cardiology, Pediatrics, Duke University Medical Center, Durham, NC, USA

M. J. Campbell · G. A. Fleming · K. D. Hill (✉)
Pediatrics, Duke University Medical Center, Durham, NC, USA

R. S. Dieter et al. (eds.), *Diseases of the Aorta*, https://doi.org/10.1007/978-3-030-11322-3_8

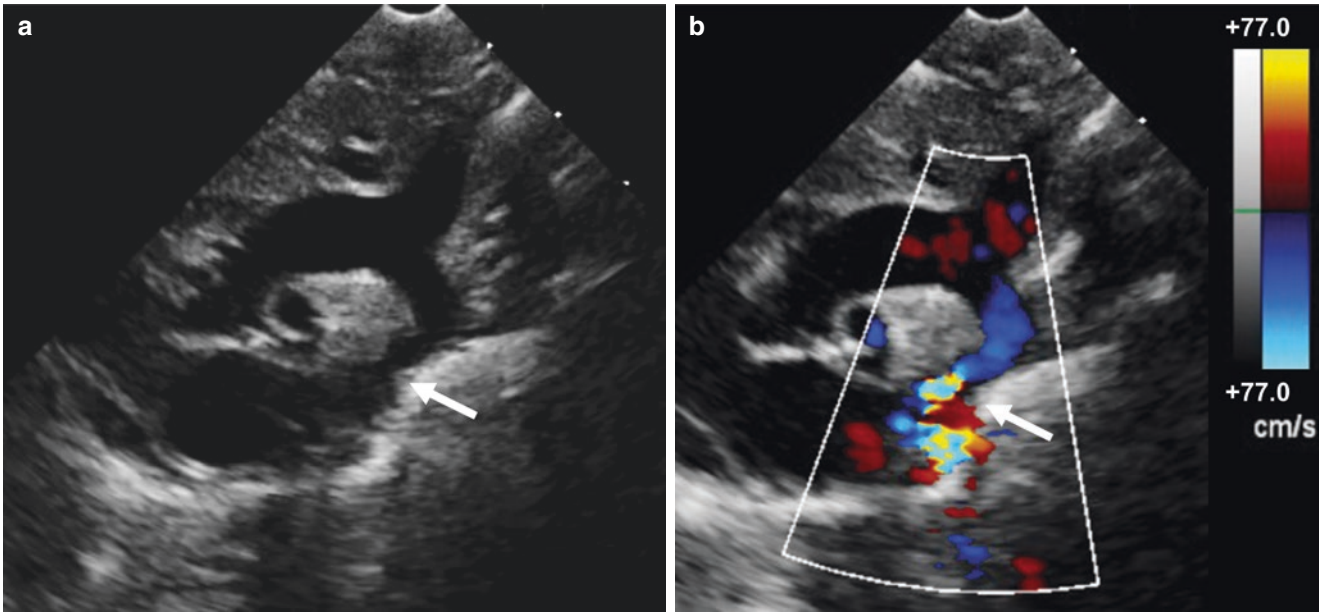

Fig. 8.1 Echocardiogram of coarctation. (**a**) Two-dimensional transthoracic echocardiogram in an 11-day-old infant with discrete coarctation (arrow). (**b**) Color Doppler of the same image demonstrating aliasing of flow at the coarctation site (arrow)

hypertension. Murmurs can be a manifestation of associated congenital heart defects (e.g. ventricular septal defects or aortic stenosis) or may be due to flow through collateral vessels that develop from the internal thoracic and subclavian arteries, thyrocervical trunks, and vertebral and anterior spinal arteries [24, 25]. In adults with previously undiagnosed coarctation, hypertension is the most common presenting symptom [26]. Frequent headaches or symptoms of claudication of the lower extremities with exertion may also be reported. In these patients, decreased lower extremity pulses or a significant systolic blood pressure gradient between the upper and lower extremities is highly suggestive of aortic coarctation [22, 26]. However, the absence of these physical exam findings does not exclude a diagnosis of coarctation, as exam findings may be diminished or even absent in the setting of significant collateral blood flow, which often develops in patients diagnosed later in life [27].

Imaging Studies

Chest x-ray may be nonspecific, especially in young patients, but in adult patients, indentation of the aorta at the site of coarctation creates a classic "3 sign," and notching of the posterior fourth to eighth ribs due to dilated intercostal arteries may also be seen [27, 28]. Electrocardiogram is often normal in infants, but in older children and adults, ventricular pressure overload typically leads to evidence of left ventricular hypertrophy [27]. Transthoracic echocardiography is the diagnostic test of choice in neonates and young children with concern for coarctation, and it enables detection of the presence and severity of aortic coarctation and any associated cardiac defects (Fig. 8.1). In adult sized patients, transthoracic echocardiography remains the initial test of choice for evaluation of coarctation [27], but echocardiographic windows may be suboptimal in this population. Computed tomography (CT) scan or magnetic resonance imaging (MRI) can provide excellent anatomic detail and are commonly used to create three-dimensional images for interventional planning (Fig. 8.2, Video 8.1). MRI has the additional benefit of defining and quantifying collateral vessel flow. Cardiac catheterization is also a valuable diagnostic tool for the diagnosis of coarctation. Although cardiac catheterization is now used less frequently as a primary diagnostic modality due to advances in other imaging modalities, such as echocardiography and MRI, catheterization remains the gold standard for quantification of pressure gradients across the region of coarctation. The addition of rotational angiography to fluoroscopic equipment will allow improved imaging of coarctation of the aorta in the catheterization lab. Moreover, transcatheter approaches are increasingly used for therapeutic intervention, particularly in older children, adolescents, and adults [27, 28].

Treatment

Fortunately, surgical and transcatheter interventions are now available for coarctation of the aorta, and the outcomes are very good. Treatment guidelines exist for both children and adults with coarctation, with intervention warranted in patients with a peak-to-peak gradient ≥20 mmHg across

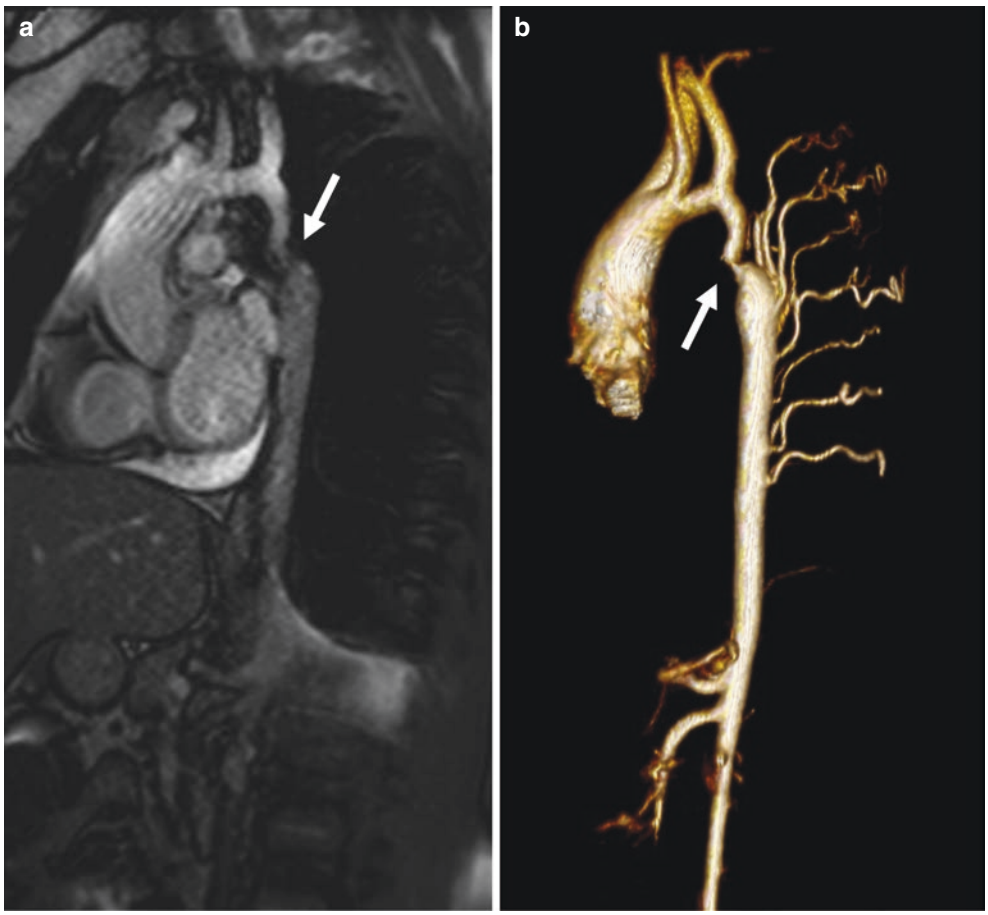

Fig. 8.2 Magnetic resonance imaging of coarctation. (**a**) Sagittal magnetic resonance image (steady-state free precession) in a 12-year-old boy showing transverse arch hypoplasia and long segment coarctation distal to the left subclavian artery (arrow). (**b**) Three-dimensional reconstruction of a gated contrasted angiogram of the same patient, again highlighting hypoplasia of the transverse arch, coarctation at the distal transverse aortic arch and isthmus (arrow), and dilated intercostal arteries functioning as collaterals

the site of coarctation. Intervention is also warranted with lesser gradients in the presence of significant anatomic evidence of stenosis and extensive collateral flow [27, 29]. Factors such as systemic hypertension, additional cardiac defects, left ventricular hypertrophy, or elevated left ventricular end diastolic pressure must also be considered when determining possible intervention [27, 29–31].

Surgical Repair

In 1944, Crafoord described resection with end-to-end anastomosis as the first surgical repair for coarctation [32] (Fig. 8.3a). Subsequent studies showed recoarctation in over half of the patients repaired with this technique, which was largely attributed to the use of a circumferential suture line [33, 34]. Gross reported using an interposition graft after resection of the segment of coarctation in 1951 [35]. While less appealing in pediatric patients due to somatic growth, this approach can be appropriate in adult patients with aneurysm, long segment coarctation, or recoarctation after primary repair [36].

As an alternative approach, Vosschulte described prosthetic patch aortoplasty for coarctation repair in 1961. In this technique, the ductal tissue is excised, a longitudinal incision across the coarctation is made, and a prosthetic patch is used to enlarge the area of stenosis (Fig. 8.3b). This technique avoids a circumferential suture line, can address longer segments of coarctation, and minimizes mobilization of the aorta and ligation of intercostal arteries [37]. Recoarctation rates of 5–12% [38] were lower compared to the resection and end-to-end anastomosis technique, but aortic aneurysm was a long-term problem with this technique, occurring in 18–51% of patients [39–42].

Subclavian flap aortoplasty was introduced in 1966 as a surgical treatment option for coarctation by Waldhausen and Nahrwold. In this approach, the left subclavian artery is ligated and divided, and the proximal left subclavian stump is folded down and used to enlarge the area of coarctation (Fig. 8.3c). This technique allows for improved growth by avoiding the use of a circumferential suture line and prosthetic material and can be used in long segment coarctation [43, 44]. However, the need to sacrifice the left subclavian artery has been a major concern with this technique, which has been associated with decreased length and muscle bulk of the left upper extremity as well as claudication with exercise [45, 46].

In 1977 Amato described a modification to Crafoord's resection and end-to-end anastomosis, called the *extended*

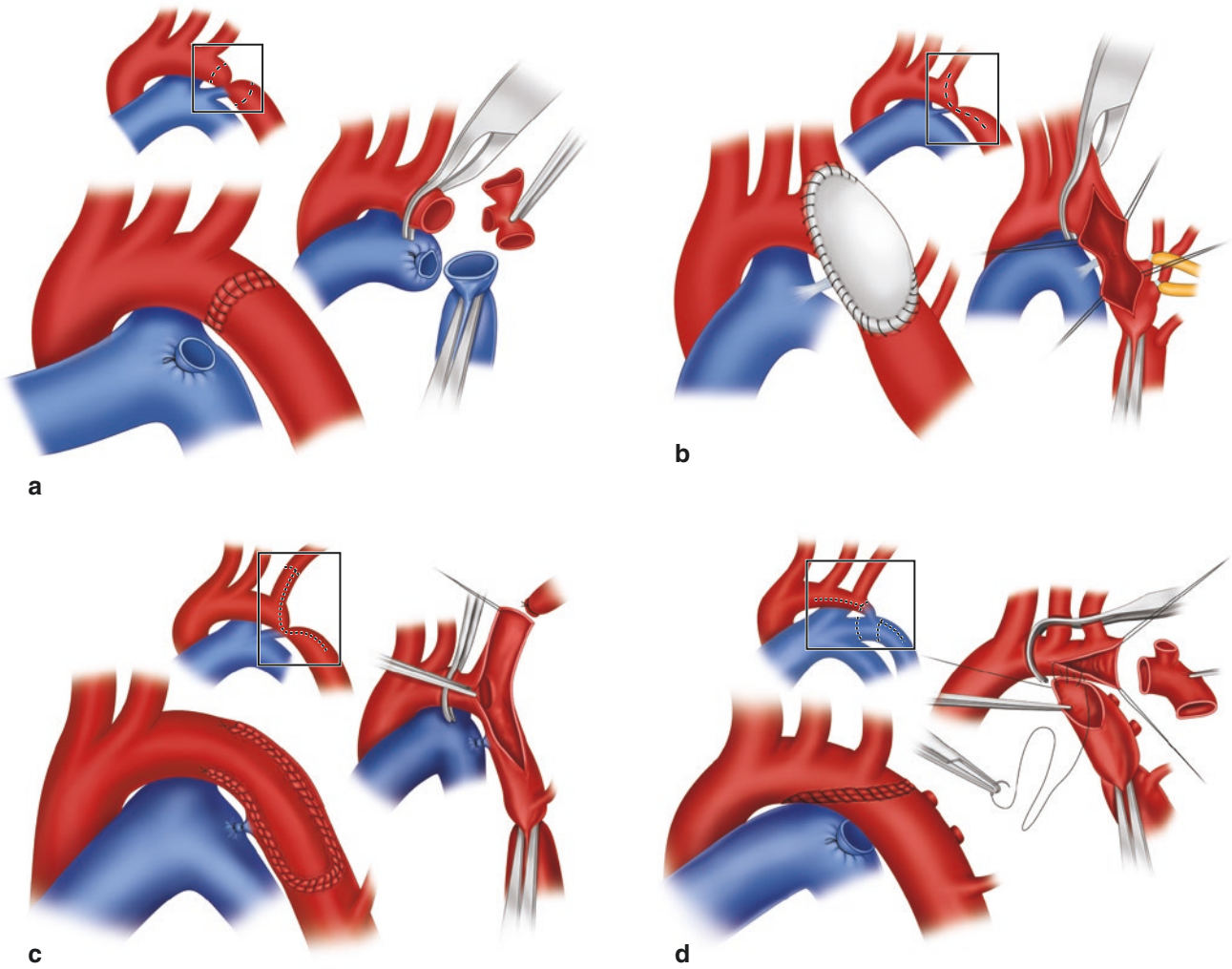

Fig. 8.3 Surgical techniques in coarctation repair. (**a**) Crafoord's original resection with end-to-end anastomosis. The coarctation is resected, and an end-to-end, circumferential anastomosis is created. (**b**) Patch aortoplasty. An incision is extended across the coarctation, followed by patch augmentation of the stenotic region. (**c**) Subclavian flap aortoplasty. The left subclavian artery is ligated and divided, and a longitudinal incision is extended from the proximal left subclavian artery beyond the area of coarctation. The proximal left subclavian flap is then folded down to enlarge the area of coarctation. (**d**) Resection with *extended* end-to-end anastomosis. The coarctation is resected using a broad, longitudinal incision, and an oblique anastomosis is used to join the undersurface of the transverse arch and descending thoracic aorta. (Adapted and reprinted from Dodge Khatami A et al. [32], with permission from John Wiley and Sons)

end-to-end technique. In the extended end-to-end approach, a broader, longitudinal incision and anastomosis across the proximal aorta is performed (Fig. 8.3d). While still avoiding prosthetic material and enabling ductal tissue resection, the wider incision is less prone to restenosis and enables enlargement of the transverse aorta [47, 48]. Currently, extended end-to-end anastomosis is one of the preferred techniques for surgical repair, due to low restenosis rates (4–11%) and low mortality rates [47, 49–51].

Finally, in neonates considerable debate has revolved around the merits of performing coarctation repair through a left thoracotomy versus a median sternotomy. While most surgeons would advocate for a left thoracotomy approach in all patients with discrete coarctation, for neonates with a hypoplastic transverse aortic arch, a slight variation of the extended end-to-end anastomosis has been called aortic arch advancement (AAA). In this procedure, a median sternotomy is performed, the infant is placed on cardiopulmonary bypass, all ductal tissue is removed, and a longitudinal incision is made on the convex aspect of the distal ascending aorta/proximal transverse arch. The descending aorta is mobilized and anastomosed in an end-to-side approach to the proximal aortic arch [52]. While one must accept the increased risks associated with cardiopulmonary bypass, compared to a left thoracotomy, proponents of the AAA approach cite overall low morbidity and mortality rates, greater exposure of the aortic arch, and lower reintervention rates [49, 52, 53].

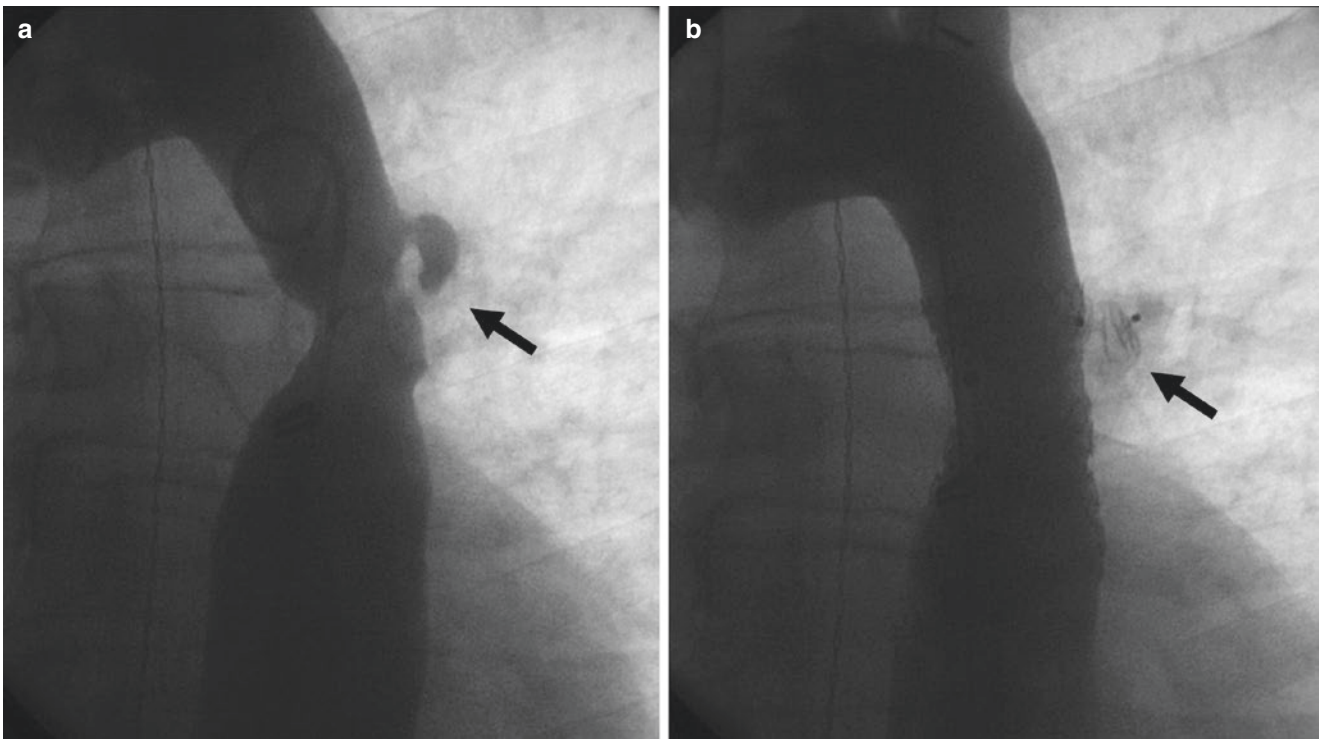

Fig. 8.4 Endovascular stent placement for coarctation. (**a**) Angiogram (LAO 30°, caudal 30°) in a 45-year-old man with a discrete coarctation and intercostal aneurysm (arrow). (**b**) Angiogram in the same projection after endovascular bare metal stent placement with no significant residual stenosis. An Amplatzer Vascular Plug II was used to successfully occlude the intercostal aneurysm (arrow)

Balloon Angioplasty: Native Coarctation

An alternative to surgical intervention for coarctation of the aorta emerged in 1982, when the use of balloon angioplasty was described by Lock [54]. Several studies have shown balloon angioplasty to be a relatively effective acute intervention for native coarctation, however recoarctation and aneurysm formation have limited the widespread adoption of balloon angioplasty in native coarctation [55–57]. In a prospective, multicenter study performed by the Congenital Cardiovascular Interventional Study Consortium (CCISC), the rate of recoarctation, defined as a systolic upper to lower extremity blood pressure gradient >15 mm Hg at intermediate term follow-up (1.5–5 years after balloon angioplasty), was 27% for native coarctation in patients greater than or equal to 10 kg. In the same study, aneurysms were observed in 8 of 21 (38%) patients with available imaging data from CT, MRI, or repeat angiography [58]. This risk of aneurysm formation was similar to that seen in a small, single-center, randomized trial comparing balloon angioplasty versus surgical repair of coarctation in 36 children (20 balloon angioplasty versus 16 surgical) ages 3–10 years of age. In this study 35% of the balloon angioplasty patients developed aneurysm, compared to none in the surgical cohort [59]. With balloon angioplasty, aneurysm formation is thought to be provoked by tearing of the intima and media during the procedure with subsequent disruption of vascular integrity [60–63].

Balloon Angioplasty: Recurrent Coarctation

In contrast to native coarctation, balloon angioplasty is typically the intervention of choice for recoarctation, especially in patients who are too small for stent placement [29]. Short-term success rates range from 80% to 93% [64], and the incidence of aortic wall injury is low at 1–2%. Fibrosis at the site of recoarctation is thought to be protective against aneurysm formation in these patients. However recoarctation remains a significant concern, with a broad incidence reported of 6–53% [65, 66].

Bare Metal Endovascular Stent Placement

Endovascular stents were introduced as a treatment option for coarctation in 1991 [67], which broadened the utility of transcatheter treatment for coarctation. Stents offer decreased rates of aortic wall injury compared to balloon angioplasty alone because stents do not require overdilation of the vessel and provide structural support to the vessel wall [5] (Fig. 8.4).

The Coarctation of the Aorta Stent Trial (COAST) has provided definitive prospective data on the safety and efficacy of stent placement for aortic coarctation. COAST began in 2007 as a multicenter, single-arm clinical trial to evaluate the safety and efficacy of the Cheatham Platinum (CP, NuMED, Hopkinton, NY) stent in children and adults with

coarctation. Because of the controversy in using endovascular stents in small children due to large sheath size and the need to accommodate for somatic growth [68, 69], patients less than 8 years of age and under 35 kg were excluded. In COAST, CP stent implantation was attempted in 105 patients from 8 to 52 years of age, with no significant acute adverse events and only one failure of stent implantation. In the catheterization lab, no patient had a significant gradient across the CP stent, and at 1 month follow-up, 99% of patients had a gradient <20 mmHg. Two-year follow-up data was available for 86% of patients, with 90% demonstrating a blood pressure gradient <20 mmHg between the upper and lower extremities. Stent fracture was relatively common, reported in 23 patients, though none of these cases had decreased stent integrity, stent migration, aortic wall injury, or hemodynamic obstruction. Aortic aneurysm was found in 6 patients, one of which spontaneously resolved. To date, there have been no reported surgical reinterventions for any COAST trial patient, but the need for stent dilation or development of aortic wall injury has prompted 19 patients to undergo repeat transcatheter intervention [70]. The COAST trial is planned to include 5 years of follow-up from stent placement, which will enable further understanding regarding the safety and efficacy of the CP stent for aortic coarctation [6].

Covered Endovascular Stents

The use of covered endovascular stents is one of the most recent transcatheter innovations for the treatment of patients with coarctation, first described in 1999. The material within the stent provides additional structural support and creates a protective barrier against shear stress and presumably subsequent aortic wall injury. When aortic aneurysm or stent fracture occur with bare metal stent placement, covered stents can serve as a rescue therapy. Covered stents can also be used as a primary treatment option for coarctation and are especially helpful in the setting of complex coarctation anatomy or when friable and calcified aortic wall tissue exists in older patients. Some limitations of covered stent placement exist, and due to the need for large sheath sizes and to accommodate for somatic growth, covered stent placement is often precluded in small children. Furthermore, caution must be taken to avoid stent occlusion of significant aortic branches, including paraspinal branches off of the descending aorta, which are often challenging to identify [71].

In 2010, the COAST II trial was developed to investigate the safety and efficacy of the covered CP stent in treating or preventing aortic wall injury in patients with coarctation. Short-term outcomes at 1-month follow-up for the COAST II trial were released in 2016. A total of 158 patients with either a history of coarctation with aortic wall injury or an increased risk of aortic wall injury underwent placement of a covered CP stent. At 1 month of follow-up, the average gradient across the aortic arch had declined from 27 ± 20 mmHg to 4 ± 6 mmHg. Complete coverage of pre-existing areas of aortic wall injury was achieved in 92% of patients, and there were no cases of acute aortic wall injury, repeat interventions, or death. This led to FDA pre-market approval of the covered CP stent in April 2016 for preventing aortic wall injury in high-risk patients with coarctation and for the treatment of existing aortic wall injury related to complications from previous interventions for coarctation. Follow-up to 24 months after covered stent placement is planned for the COAST II trial [72].

A recent randomized clinical trial was performed comparing bare CP stent placement to covered CP stent placement in 120 patients. No procedural complications occurred in either group, and at a mean follow-up of 31.1 months, the bare CP stent group had an increase in the rate of recoarctation (6.7% versus 0%) and decrease in the rate of pseudoaneurysm (0% versus 3.3%) compared to the covered CP stent group, though neither comparison reached statistical significance. In both cases of pseudoaneurysm in the covered stent group, the aneurysm developed at the proximal end of the stent and was able to be treated by placing a second covered stent. Neither case developed any further complications [73].

Management Algorithm

In the setting of various treatment options, determination of the optimal treatment strategy for coarctation of the aorta can be complicated, and there is no comprehensive evidence-based standard of care or algorithm. Guidelines from the American College of Cardiology and the American Heart Association provide some insight, but the level of evidence supporting these recommendations is suboptimal (Level B or C for all recommendations) [27, 29]. Treatment decisions must be made after careful consideration of the age at presentation, complexity of the coarctation, and whether the coarctation is native or recurrent. In general, surgical repair is preferred for infants and young children with native coarctation due to the risk of recurrent coarctation and aortic wall injury with angioplasty, the need for large sheath sizes, and challenges in accounting for somatic growth with stent placement [59]. Surgical repair may also be more appropriate at any age when repair of associated cardiac defects is indicated or in patients with complex coarctation anatomy, including those with transverse arch hypoplasia, tortuous segments of recoarctation, and distorted arterial branch anatomy [27]. For uncomplicated native coarctation in the older child or adult, stent placement with either a bare metal or covered stent can offer a less invasive approach than surgical repair with good long-term outcomes [27, 29, 74]. For recurrent coarctation, balloon angioplasty is typically per-

formed. If the anatomy is favorable, stent placement should also be considered when the chosen stent can be dilated to near adult size [29, 58].

Treatment Approach for Coarctation of the Aorta in Children and Adults

Indications for Treatment [27, 29]:
Intervention is indicated with a peak-to-peak systolic pressure gradient ≥20 mmHg across the site of coarctation upon initial presentation or in the setting of recurrent coarctation.
Intervention is also warranted with lesser gradients in the presence of significant anatomic evidence of stenosis and extensive collateral flow.
Intervention may be considered with a peak-to-peak systolic pressure gradient <20 mmHg but with systemic hypertension associated with anatomic narrowing that explains the hypertension.
Intervention may be considered with a peak-to-peak systolic pressure gradient <20 mmHg but with an elevated left ventricular end-diastolic pressure and an anatomic narrowing.

Treatment Approach:

Surgical Repair

Surgical repair is typically preferred over transcatheter approaches in infants and young children with native coarctation, all patients requiring repair of associated cardiac defects, or in the setting of complex coarctation anatomy.

Extended end-to-end anastomosis is typically the preferred surgical technique, which avoids prosthetic material, includes resection of the coarctation, and involves a wider incision that is less prone to restenosis.

In neonates with a hypoplastic transverse aortic arch, a variation of the extended end-to-end anastomosis technique, called aortic arch advancement, may be preferred.

Balloon Angioplasty

Typically balloon angioplasty is the preferred intervention for recurrent coarctation in children and adults.

Balloon angioplasty is not often used in native coarctation due to concern for recoarctation and aneurysm formation.

Endovascular Stent

For uncomplicated coarctation in the older child or adult, either a bare metal or covered endovascular stent can offer a less invasive approach than surgical repair with good long-term outcomes. If the anatomy is favorable, stent placement should be considered when the chosen stent can be dilated to near adult size.

Endovascular stents provide structural support and decreased rates of aortic wall injury and aneurysm compared to balloon angioplasty, but care must be taken to avoid overlying vital branch vessels.

Covered stents may be considered as an alternative to bare metal stents, particularly in patients felt to be at increased risk for aortic wall injury.

The use of stents in small children remains controversial due to the need for large sheath sizes and limitations in accommodating for somatic growth.

Patient Follow-Up

Without intervention, the outcome for patients with coarctation of the aorta is overwhelmingly poor. In his classic 1970 natural history study, Campbell examined autopsy and clinical records of 465 patients with coarctation who survived beyond 1 year of age, and the mean age of death was 34 years, with 75% mortality by 43 years of age. Causes of death included congestive heart failure (26%), aortic rupture (21%), bacterial endocarditis (18%), and intracranial hemorrhage (12%) [75]. Fortunately, surgical and transcatheter techniques have evolved, and outcomes for patients with repaired coarctation are now overall quite good. After coarctation repair, patients still must be followed at least annually by a cardiologist to assess for long-term issues, such as hypertension and associated left ventricular hypertrophy and dysfunction, exercise intolerance, intracranial aneurysms, and recoarctation [27].

Hypertension

Hypertension is endemic in patients with repaired aortic coarctation and represents the most common long-term morbidity [76, 77]. In a contemporary analysis of the Swedish National Registry on Congenital Heart Disease (SWEDCON), hypertension was present in 344/653 (52.7%) adults (mean age 36.9 ± 14.4 years) with a prior history of coarctation repair (mean age at repair 9.5 ± 11 years) [78]. Risk factors for hypertension in multivariable analysis included male sex (OR = 3.35 [95% CI: 1.98–5.68]), age (OR = 1.07 per year [95% CI: 1.05–1.10]), increased body mass index (OR = 1.09 per unit increase [95% CI: 1.03–1.06]), and a residual right upper to lower extremity systolic blood pressure gradient of 10–19 mm Hg (OR = 3.58 [95% CI: 1.70–7.55]) or >20 mm Hg (OR = 11.38 [95% CI: 4.03–32.11]) [78]. The etiology of such high rates of baseline and exercise-induced hypertension remain unclear but may be due to any combination of underlying arteriopathy, decreased aortic wall compliance, abnormal streaming of blood flow, or renal abnormalities [76]. In addition to traditional hypertension risk factors such as age and body mass index, the substantially increased risk of hypertension in patients with a residual blood pressure gradient serves to highlight the importance of evaluating for recoarctation as a cause of hypertension.

Intracranial Aneurysm

Adults with a history of coarctation have a five-fold increased risk of developing an intracranial aneurysm [79]. Recognizing this risk, the current American College of Cardiology and American Heart Association adult congenital guidelines recommend screening for intracranial vasculature abnormalities in patients with coarctation by CT or MRI, but the exact timing and frequency of follow-up imaging is not defined [27]. A prospective trial using CT angiography for screening of intracranial aneurysm in patients with coarctation found increased age to be the sole risk factor for

the development of intracranial aneurysm in patients with coarctation, with the fourth and fifth decade of life being the most common age at presentation [79]. While further studies are needed, this data suggests screening for intracranial aneurysm should be performed in patients with coarctation at least by the fourth decade of life.

Impact of Arch Type

Even when no evidence of recoarctation exists, patients with a history of coarctation are at risk for hypertension, vascular remodeling, and decreased left ventricular function. Some evidence suggests that the morphology of the aortic arch after coarctation repair can impact outcomes in patients who have undergone coarctation repair. Three types of aortic arch morphology have been described: (1) Romanesque with a normal, rounded aortic arch, (2) Crenel with a rectangular shaped aortic arch and normal horizontal aortic width, and (3) Gothic with an acutely angled, triangular aortic arch and an exaggerated height-to-width ratio (Fig. 8.5) [80]. Interestingly, in repaired coarctation patients, Gothic aortic arch morphology has been associated with increased prevalence of hypertension both at baseline [80] and with exercise [81]. Ou et al. demonstrated that the Gothic aortic arch morphology is also associated with other maladaptive aortic features including increased carotid artery intima-media thickness, higher aortic stiffness index, and impaired vasoreactivity proximal to the site of coarctation repair. Bruse et al. described an association between Gothic arch morphology after coarctation repair and impaired ventricular performance including lower left ventricular ejection fraction, larger indexed left ventricular end-diastolic volume, and elevated indexed left ventricular mass [82]. However, the relationship between arch morphology and long-term outcome has not been completely consistent, and other reports have shown no association between arch type and exercise induced hypertension [83, 84]. In fact, one study proposed that hypoplasia of the transverse arch and isthmus, not arch curvature, were the major factors associated with exercise induced hypertension [84]. Indeed, patients with repaired coarctation with normal or Romanesque aortic arch morphology also have significantly higher carotid artery intima-media thickness, aortic stiffness, and impaired vasoreactivity suggesting that arch morphology is not the sole determinant of abnormal arch physiology [85]. Overall, further study is needed regarding the utility of assessing arch morphology for long-term risk prediction in patients with repaired coarctation.

Consensus Follow-up Recommendations

According to guidelines from the American Heart Association and American College of Cardiology for management of adults with congenital heart disease, patients with repaired coarctation should be followed at least annually by a cardiologist, or sooner if concerns arise. In adults, it is recommended that there be consultation with a specialist in adult congenital heart disease.

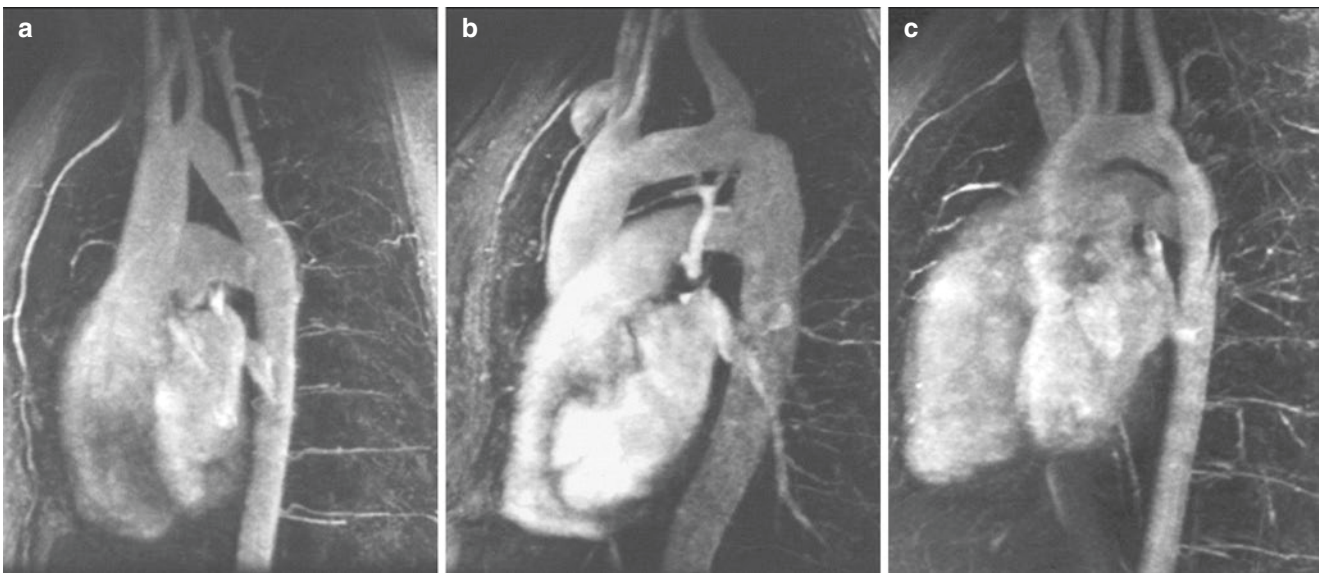

Fig. 8.5 Three described categories of arch morphology. (**a**) Gothic arch with acute angulation of the aortic arch, creating an exaggerated height-to-width ratio. (**b**) Crenel arch with rectangular shaped aortic arch and normal width. (**c**) Romanesque arch with a normal, rounded shape. (Adapted and reprinted from Ou P, et al. [85], with permission from Elsevier)

Screening for baseline and exercise induced hypertension, including evaluation for the presence of a right upper to lower extremity blood pressure gradient is recommended. Cardiac evaluation in patients with hypertension should include evaluation for associated left ventricular hypertrophy and dysfunction. Re-imaging of the repaired coarctation is recommended at least every 5 years, or sooner based on the original anatomy and symptoms, to assess for complications such as aortic aneurysm or recurrent stenosis [27]. At least by the fourth decade of life, CT or MRI angiography of the brain should be performed to assess for the presence of intracranial aneurysms. Exercise should be encouraged in patients with no significant upper to lower extremity blood pressure gradient, no evidence of aneurysm or associated heart defects, and normotension at rest and with exercise. Patients should only be restricted from activities with a large static component [86]. Although not addressed in the American Heart Association and American College of Cardiology guidelines, the European Society of Cardiology recommends consideration of reintervention regardless of symptoms for all patients with repaired coarctation and a noninvasive systolic blood pressure gradient >20 mmHg between upper and lower limbs with upper limb hypertension (>140/90 mmHg in adults), pathologic blood pressure response during exercise, or significant left ventricular hypertrophy [87]. Finally, according to the most recent American Heart Association guidelines, endocarditis prophylaxis is not routinely recommended after the first 6 months following surgical or transcatheter intervention, unless a previous history of infectious endocarditis exists [88].

Pseudocoarctation

Pseudocoarctation is a rare anomaly that refers to kinking or buckling of the aorta at the isthmus without significant obstruction to flow or development of collateral circulation (Fig. 8.6). This condition is thought to arise embryologically from abnormal compression of the third through seventh dorsal aortic segments, leading to a superiorly displaced distal aortic arch and redundancy and kinking of the aorta at the ligamentum arteriosum [89, 90]. Patients with pseudocoarctation are typically asymptomatic but may present with hypertension [91]. On routine chest x-ray pseudocoarctation may resemble a mediastinal mass due to the superior displacement of the distal transverse aortic arch. Patients with pseudocoarctation may also appear to have true coarctation on chest x-ray due to an indentation at the isthmus where the aorta is kinked, giving a classic "3 sign." In these challenging scenarios, a high degree of suspicion for pseudocoarctation must exist, prompting further evaluation [89, 92].

In isolation, pseudocoarctation is typically felt to be a benign condition due to the lack of actual obstruction to aortic blood flow. While pseudocoarctation often exists in isolation, it is important to consider the association of additional congenital heart defects, especially a bicuspid or stenotic aortic valve, which warrant further evaluation and potential intervention [93]. Furthermore, patients with pseudocoarctation are at risk for aortic aneurysm and dissection distal to the kinked segment, which is thought to be related to abnormal, turbulent blood flow beyond the area of pseudocoarctation [94]. Therefore, patients with suspected pseudocoarctation should first undergo transthoracic echocardiography to rule out associated congenital heart defects. The area of aortic kinking may be difficult to visualize by echocardiography, and CT angiography or MRI should then be performed to rule out true coarctation as well as the development of aortic aneurysm or dissection distal to the kinked segment [90, 95]. If less invasive imaging techniques remain inconclusive, cardiac catheterization should be performed, which remains the gold standard to determine the anatomy and pressure gradient across the aorta. Surgical intervention for pseudocoarctation is typically reserved for patients with significant symptoms or when concern for aortic aneurysm and/or dissection exist [90].

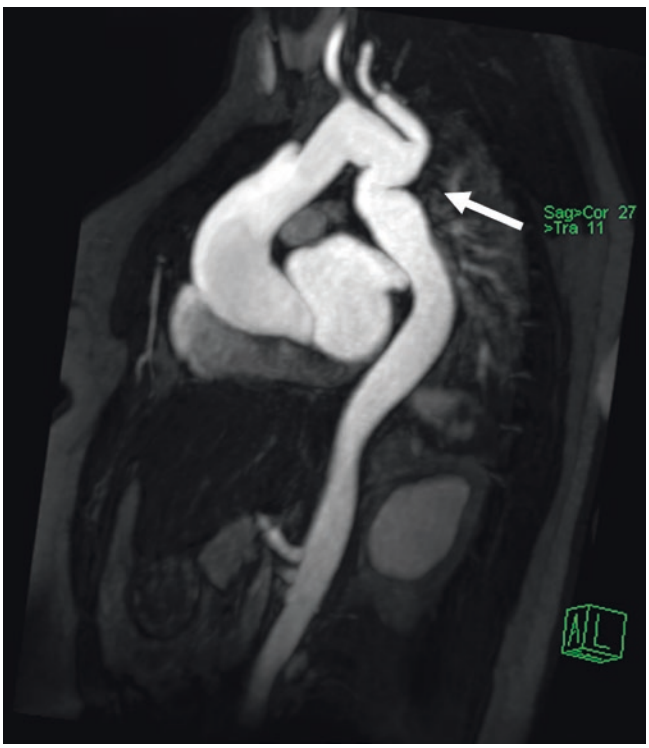

Fig. 8.6 Magnetic resonance image of pseudocoarctation. Sagittal magnetic resonance image angiogram of the thoracic aorta in a 39-year-old man with a history of pseudocoarctation demonstrating significant tortuosity of the proximal descending aorta (arrow)

Abdominal Coarctation

Abdominal coarctation, also known as Middle aortic syndrome (MAS), occurs when there is narrowing of the distal thoracic and/or the abdominal aorta (Fig. 8.7). It is a rare disease found in children and young adults, making up only 0.5–2% of all cases of aortic coarctation [96]. While uncommon, this is an important etiology for hypertension in children and young adults. A recent systematic review of MAS showed that most cases are idiopathic (64%), 15% are associated with genetic disease such as neurofibromatosis type I, Alagille syndrome, and William syndrome, and an additional 17% of cases are caused by inflammatory diseases such as Takayasu arteritis or intrauterine infection [97]. The exact embryologic development of MAS remains unclear, but one theory is that it results from abnormal fusion of the two dorsal aortas in fetal life. An inflammatory response with resultant fibrosis in the setting of an intrauterine infection prenatally or postnatal vasculitic diseases may also explain some cases of MAS [98].

Clinical Presentation

Patients with MAS most commonly present with refractory hypertension, and the severity depends on the location and degree of vessel stenosis. An abdominal bruit may be heard, and patients may have absent femoral pulses and symptoms of claudication. While stenosis of visceral vessels is quite common in MAS, reports of intestinal angina and weight loss occur rarely, though renal dysfunction occurs more commonly [97, 99].

Evaluation

As with classic coarctation, patients with MAS may demonstrate evidence of left ventricular hypertrophy on an electrocardiogram. A transthoracic echocardiogram should be obtained to assess the anatomy of the thoracic and abdominal aorta and to screen for associated intracardiac defects and end-organ damage from hypertension [98]. If an abdominal bruit exists on exam, a dedicated abdominal or renal ultrasound may also be indicated but can be of limited quality in adult patients due to technical challenges. Typically CT angiography or MRI is then utilized to better define the exact areas of the aorta that are affected, the presence of collaterals, and to define any extra-aortic vessel involvement [100, 101]. Cerebrovascular disease occurs in as many as 45% of patients with MAS and should be evaluated on CT angiography or MRI of the brain as well [102]. The abdominal aorta is the site of narrowing in 97% of cases, with only 3% of MAS affecting the distal thoracic aorta. In a large systematic review of MAS in adults by Rumman et al., 57% of the 630 reviewed cases defined the site of abdominal coarctation, and the most common site of coarctation was the suprarenal aorta

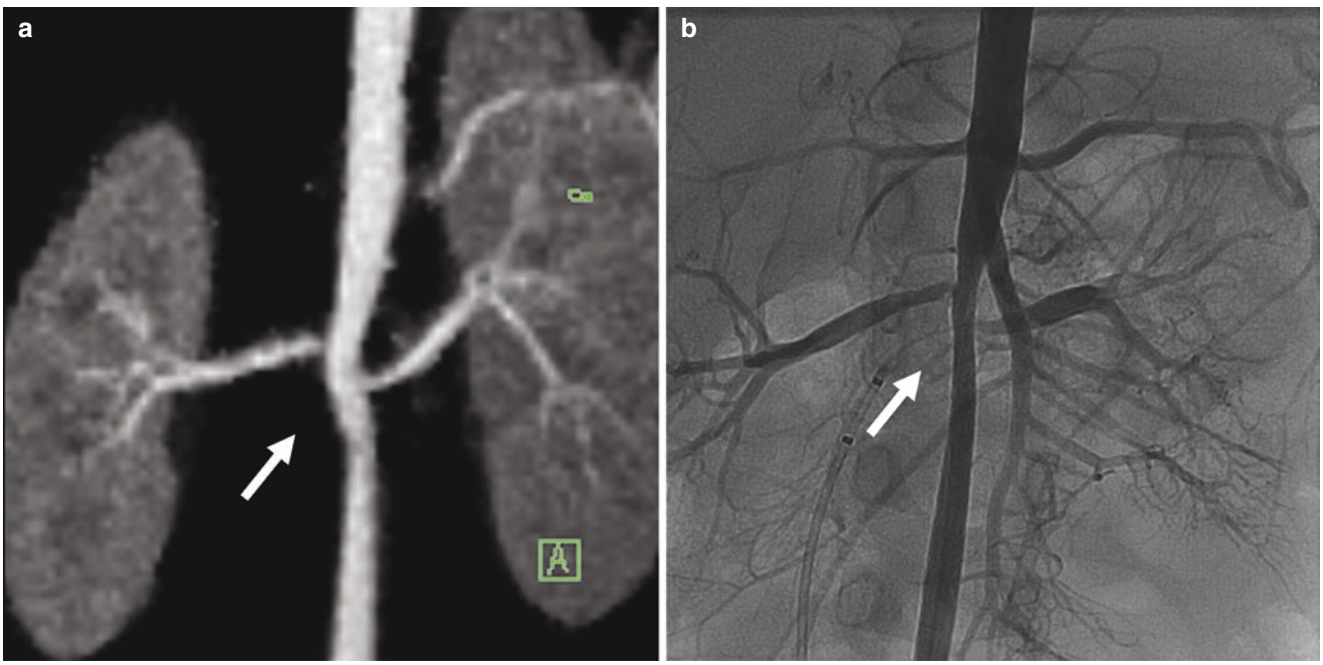

Fig. 8.7 Abdominal coarctation. (**a**) Maximum intensity projection (MIP) image of an abdominal magnetic resonance contrast angiogram in a 3-year-old girl with refractory systemic hypertension showing abdominal coarctation and bilateral renal artery stenosis (arrow). (**b**) Anterior–posterior projection of an angiogram of the descending aorta in a 6-year-old girl showing abdominal coarctation and renal artery stenosis (arrow)

(29%), followed by stenosis from the suprarenal to infrarenal aorta (12%), and the infrarenal aorta (8%). Visceral branch vessels are affected in about 70% of cases of MAS, with renal artery stenosis being the most common (66%), followed by stenosis of the superior mesenteric artery (30%) and celiac trunk (22%). Interestingly the inferior mesenteric artery is typically not affected [97].

Management

With severe hypertension being the most common symptom, antihypertensive agents are the first line treatment option in MAS. Unfortunately, there is a high rate of refractory hypertension, and in their series of 36 patients with MAS, Tummolo et al. report only an 8% success rate of medical management with antihypertensives [102]. Failure to achieve blood pressure control or evidence of end-organ damage are often cited as reasons to pursue endovascular or surgical interventions, but specific guidelines for intervention do not exist. The region and length of stenosis need to accommodate for somatic growth in young patients, extra-aortic vessel involvement, and degree of symptoms must be considered on an individual basis [97]. The use of a stent in percutaneous transluminal angioplasty (PTA) can be quite effective in relieving stenosis, but care must be taken to avoid occlusion of important visceral arteries [103]. In their systematic review of 630 adult cases of MAS, Rumman et al. reported that 28% of patients underwent PTA with or without stenting. Complications were reported in 13% of patients, mortality in 2.3%, and technical failure or need for reintervention was described in 28% of cases [97]. In their report of outcomes in 36 patients with MAS, Tummolo et al. report that 36% of patients underwent PTA, with 46% requiring repeat PTA. Because of failure to adequately control blood pressure after PTA, 53% of these patients went on to require surgical intervention [102].

Surgical options for patients with MAS include thoracoabdominal bypass grafts, patch aortoplasty, interposition aortoaortic grafts, and renal autotransplantation. In the systematic review by Rumman et al., 55% of 630 patients underwent surgical treatment for MAS, with 12% of these cases following failed endovascular intervention. Of these surgical cases, 42% were done by aortoaortic bypass, 23% involved reconstruction patch graft, and renal autotransplantation was performed in 11% of cases. While most patients tolerated surgery well, a complicated postoperative course was reported in 9% of cases, technical failure in 8%, and surgical mortality occurred in 2.9% of cases. Interestingly, cases involving arteritis were the highest risk [97]. Tummolo et al. reported 47% of their patients proceeding to surgery, with 41% of these cases following failed endovascular intervention. At mean follow-up of 5.6–7.2 years (patients who underwent surgery only versus surgery after failed PTA, respectively), 25% of patients no longer required antihypertensives, 58% required antihypertensive therapy with improved BP control, 14% of patients continued to have refractory hypertension, and 3% were reported as a technical failure. In their series of 53 patients who underwent surgical treatment for MAS, Stanley et al. reported resolved hypertension in 53% and improved hypertension in 34% of patients. There was no improvement in blood pressure in 7% of patients, who underwent repeat surgical intervention [99].

Outcome

Left untreated, MAS leads to a shortened life expectancy, typically in the fourth decade of life [103]. Residual hypertension is the most common long-term problem in MAS, and, even after endovascular or surgical intervention, hypertension is reported in over one-third of patients [97]. Restenosis, especially in-stent stenosis, and outgrowth of a previously placed stent are typical reasons for surgical reintervention after PTA [103]. In surgical patients, reintervention due to somatic growth relative to an aortic bypass graft of patch aortoplasty is not unusual, with one surgical cohort describing a reintervention rate of 6% for these reasons [99]. Exact guidelines regarding follow-up in MAS patients after intervention do not exist. However, it would seem reasonable to extrapolate recommendations for typical aortic coarctation by suggesting at least annual evaluation by a cardiologist with screening for hypertension, exercise intolerance, left ventricular hypertrophy and ventricular dysfunction. Regarding follow-up imaging, one proposed regimen is to perform at least yearly surveillance with CT angiography or MRI, and once several scans are documented to be stable, spacing imaging intervals to every 2–3 years [98].

Conclusion

Coarctation of the aorta is a very heterogeneous disease that can present at any age, sometimes requiring a high index of suspicion to make the appropriate diagnosis. Fortunately in the past 70 years, a great deal of progress has been made in the ability to both diagnose and treat aortic coarctation. Advances in echocardiography, CT, and MRI have aided the diagnosis, treatment planning, and follow-up in these patients. Modifications of various surgical techniques have led to low mortality and morbidity rates, even in the smallest patients. Development of transcatheter balloon angioplasty and subsequently endovascular stent place-

Table 8.1 Important studies and guideline statements in the treatment and outcome of coarctation in adults and children

Author	*N*	Follow-up	Outcome
Cowley et al. (2005) [59]	36	Mean 14 years	Randomized trial comparing BA and surgery for native coarctation in children. Aortic aneurysm developed in 35% of BA patients and none of the surgical patients
Carr (2006) [104]	846	Mean 36 months for catheter-based group and 7.8 years for surgical group	Meta-analysis comparing catheter versus surgical intervention for adults with coarctation. Higher risk of restenosis and need for reintervention found in catheter-based group
Forbes et al. (2007) [105]	578	Median 12 months	Retrospective multicenter analysis at intermediate follow-up after stent placement for coarctation. Exceeding a balloon/coarctation ratio of 3.5 and pre-stent BA increased risk of aortic wall injury
Warnes et al. (2008) [27]	–	–	ACC/AHA guidelines for management of coarctation in adults
Baumgartner et al. (2010) [87]	–	–	ESC guidelines for management of coarctation in adults
Holzer et al. (2010) [106]	302	3–60 months	Prospective analysis of acute, intermediate, and long-term follow-up after stent placement for coarctation using CCISC registry. At long-term follow-up, recoarctation in 20% of patients, 4% required unplanned reintervention, and 1% had aortic wall injury
Feltes et al. (2011) [29]	–	–	AHA guidelines for transcatheter intervention in children with coarctation.
Forbes et al. (2011) [107]	350	Mean 1.7 years	Multicenter observational study comparing surgery, BA, and stent placement for native coarctation in children using CCISC registry. Significantly lower acute complication rates in stent group but higher planned reintervention rates. Hemodynamic and arch imaging outcomes superior in stent and surgical patients compared to BA group.
Harris et al. (2014) [58]	130	3–60 months	Prospective, multicenter analysis of short and intermediate outcomes for BA in native and recurrent coarctation in children. Trend toward increased acute aortic wall injury and restenosis in native coarctation patients.
Sohrabi et al. (2014) [73]	120	Mean 31.1 months	Randomized clinical trial comparing covered and bare CP stents for native coarctation in adolescents and adults. Trend of increased rates of restenosis and lower rates of pseudoaneurysm in bare stent group.
Meadows et al. (2015) [70]	105	2 years	Prospective, multicenter, single-arm study assessing safety and efficacy of CP stent in children and adults with coarctation. Two-year follow-up of 86% showed 23 fractured stents with no significant clinical effects, 6 aortic aneurysms, 19 repeat catheter interventions, and no surgical interventions
Rumman et al. (2015) [97]	630	Median 4 years	Systematic review examining the features of MAS in children. There is a high prevalence of stenosis of the visceral arteries, with renal artery stenosis being most common (70% of cases). Most cases of MAS are idiopathic, but disease severity is worse in the setting of genetic or inflammatory etiologies.
Rinnström et al. (2016) [78]	653	Mean 27.4 years	Analysis of the SWEDCON registry demonstrated hypertension in 52.7% of patients with repaired coarctation. Associated risk factors for hypertension in these patients were increasing age, male sex, elevated body mass index, and a residual right upper to lower extremity systolic blood pressure gradient.

BA balloon angioplasty, *ACC* American College of Cardiology, *AHA* American Heart Association, *ESC* European Society of Cardiology, *CCISC* Congenital Cardiovascular Interventional Study Consortium, *CP* Cheatham Platinum, *MAS* middle aortic syndrome, *SWEDCON* Swedish National Registry on Congenital Heart Disease

ment have expanded treatment options and allowed less invasive approaches for some patients. However, even after seemingly uncomplicated repairs, patients with coarctation of the aorta are still at risk for long-term health issues, most notably hypertension, exercise intolerance, and left ventricular hypertrophy and dysfunction (Table 8.1). Ongoing efforts to understand and potentially mitigate these long-term problems are underway.

Conflict-of-interest Statement Fleming GA is the site principal investigator for the Covered Cheatham Platinum Stents for the Prevention or Treatment of Aortic Wall Injury Associated with Coarctation of the Aorta (COAST II) trial at Duke University Medical Center. There are no other conflicts of interest to disclose.

References

1. Ho SY, Anderson RH. Coarctation, tubular hypoplasia, and the ductus arteriosus. Histological study of 35 specimens. Br Heart J. 1979;41(3):268–74.
2. Price TP, Whisenhunt AK, Policha A, Ayad MT, Gardiner GA Jr, Abai B, et al. Middle aortic coarctation. Ann Vasc Surg. 2014;28(5):1314 e15–21.
3. Mullen MJ. Coarctation of the aorta in adults: do we need surgeons? Heart. 2003;89(1):3–5.
4. Kvitting JP, Olin CL. Clarence Crafoord: a giant in cardiothoracic surgery, the first to repair aortic coarctation. Ann Thorac Surg. 2009;87(1):342–6.
5. Padua LM, Garcia LC, Rubira CJ, de Oliveira Carvalho PE. Stent placement versus surgery for coarctation of the thoracic aorta. Cochrane Database Syst Rev. 2012;5:CD008204.

6. Ringel RE, Gauvreau K, Moses H, Jenkins KJ. Coarctation of the Aorta Stent Trial (COAST): study design and rationale. Am Heart J. 2012;164(1):7–13.
7. McBride KL, Zender GA, Fitzgerald-Butt SM, Koehler D, Menesses-Diaz A, Fernbach S, et al. Linkage analysis of left ventricular outflow tract malformations (aortic valve stenosis, coarctation of the aorta, and hypoplastic left heart syndrome). Eur J Hum Genet. 2009;17(6):811–9.
8. Loffredo CA, Chokkalingam A, Sill AM, Boughman JA, Clark EB, Scheel J, et al. Prevalence of congenital cardiovascular malformations among relatives of infants with hypoplastic left heart, coarctation of the aorta, and d-transposition of the great arteries. Am J Med Genet A. 2004;124A(3):225–30.
9. Anderson RH, Lenox CC, Zuberbuhler JR. Morphology of ventricular septal defect associated with coarctation of aorta. Br Heart J. 1983;50(2):176–81.
10. Shinebourne EA, Tam AS, Elseed AM, Paneth M, Lennox SC, Cleland WP. Coarctation of the aorta in infancy and childhood. Br Heart J. 1976;38(4):375–80.
11. Shone JD, Sellers RD, Anderson RC, Adams P Jr, Lillehei CW, Edwards JE. The developmental complex of "parachute mitral valve," supravalvular ring of left atrium, subaortic stenosis, and coarctation of aorta. Am J Cardiol. 1963;11:714–25.
12. Warnes CA. Bicuspid aortic valve and coarctation: two villains part of a diffuse problem. Heart. 2003;89(9):965–6.
13. Becker AE, Becker MJ, Edwards JE. Anomalies associated with coarctation of aorta: particular reference to infancy. Circulation. 1970;41(6):1067–75.
14. Ware SM, Jefferies JL. New genetic insights into congenital heart disease. J Clin Exp Cardiol. 2012:S8.
15. Eckhauser A, South ST, Meyers L, Bleyl SB, Botto LD. Turner Syndrome in Girls Presenting with Coarctation of the Aorta. J Pediatr. 2015;167(5):1062–6.
16. Patel A, Costello JM, Backer CL, Pasquali SK, Hill KD, Wallace AS, et al. Prevalence of noncardiac and genetic abnormalities in neonates undergoing cardiac operations: analysis of the society of thoracic surgeons congenital heart surgery database. Ann Thorac Surg. 2016;102(5):1607–14.
17. Rudolph AM, Heymann MA, Spitznas U. Hemodynamic considerations in the development of narrowing of the aorta. Am J Cardiol. 1972;30(5):514–25.
18. Allan LD, Chita SK, Anderson RH, Fagg N, Crawford DC, Tynan MJ. Coarctation of the aorta in prenatal life: an echocardiographic, anatomical, and functional study. Br Heart J. 1988;59(3):356–60.
19. Head CE, Jowett VC, Sharland GK, Simpson JM. Timing of presentation and postnatal outcome of infants suspected of having coarctation of the aorta during fetal life. Heart. 2005;91(8):1070–4.
20. Gomez-Montes E, Herraiz I, Mendoza A, Escribano D, Galindo A. Prediction of coarctation of the aorta in the second half of pregnancy. Ultrasound Obstet Gynecol. 2013;41(3):298–305.
21. Quartermain MD, Pasquali SK Hill KD, Goldberg DJ, Huhta JC, Jacobs JP, Jacobs ML, Kim S, Ungerleider RM. Variation in prenatal diagnosis of congenital heart disease in infants. Pediatrics. 2015 (In Press).
22. Rosenthal E. Coarctation of the aorta from fetus to adult: curable condition or life long disease process? Heart. 2005;91(11):1495–502.
23. Ward KE, Pryor RW, Matson JR, Razook JD, Thompson WM, Elkins RC. Delayed detection of coarctation in infancy: implications for timing of newborn follow-up. Pediatrics. 1990;86(6):972–6.
24. Steffens JC, Bourne MW, Sakuma H, O'Sullivan M, Higgins CB. Quantification of collateral blood flow in coarctation of the aorta by velocity encoded cine magnetic resonance imaging. Circulation. 1994;90(2):937–43.
25. Leschka S, Alkadhi H, Wildermuth S. Images in cardiology. Collateral circulation in aortic coarctation shown by 64 channel multislice computed tomography angiography. Heart. 2005;91(11):1422.
26. Strafford MA, Griffiths SP, Gersony WM. Coarctation of the aorta: a study in delayed detection. Pediatrics. 1982;69(2):159–63.
27. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, et al. ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing committee to develop guidelines on the management of adults with congenital heart disease). Circulation. 2008;118(23):e714–833.
28. Karaosmanoglu AD, Khawaja RD, Onur MR, Kalra MK. CT and MRI of aortic coarctation: pre- and postsurgical findings. AJR Am J Roentgenol. 2015;204(3):W224–33.
29. Feltes TF, Bacha E, Beekman RH 3rd, Cheatham JP, Feinstein JA, Gomes AS, et al. Indications for cardiac catheterization and intervention in pediatric cardiac disease: a scientific statement from the American Heart Association. Circulation. 2011;123(22):2607–52.
30. Rao PS. Coarctation of the aorta. Curr Cardiol Rep. 2005;7(6):425–34.
31. Vergales JE, Gangemi JJ, Rhueban KS, Lim DS. Coarctation of the aorta - the current state of surgical and transcatheter therapies. Curr Cardiol Rev. 2013;9(3):211–9.
32. Dodge-Khatami A, Backer CL, Mavroudis C. Risk factors for recoarctation and results of reoperation: a 40-year review. J Card Surg. 2000;15(6):369–77.
33. Kappetein AP, Zwinderman AH, Bogers AJ, Rohmer J, Huysmans HA. More than thirty-five years of coarctation repair. An unexpected high relapse rate. J Thorac Cardiovasc Surg. 1994;107(1):87–95.
34. Williams WG, Shindo G, Trusler GA, Dische MR, Olley PM. Results of repair of coarctation of the aorta during infancy. J Thorac Cardiovasc Surg. 1980;79(4):603–8.
35. Gross RE. Treatment of certain aortic coarctations by homologous grafts; a report of nineteen cases. Ann Surg. 1951;134(4):753–68.
36. Charlton-Ouw KM, Codreanu ME, Leake SS, Sandhu HK, Calderon D, Azizzadeh A, et al. Open repair of adult aortic coarctation mostly by a resection and graft replacement technique. J Vasc Surg. 2015;61(1):66–72.
37. Vossschulte K. Surgical correction of coarctation of the aorta by an “isthmusplastic” operation. Thorax. 1961;16:338–45.
38. Backer CL, Paape K, Zales VR, Weigel TJ, Mavroudis C. Coarctation of the aorta. Repair with polytetrafluoroethylene patch aortoplasty. Circulation. 1995;92(9 Suppl):II132–6.
39. Rheuban KS, Gutgesell HP, Carpenter MA, Jedeikin R, Damman JF, Kron IL, et al. Aortic aneurysm after patch angioplasty for aortic isthmic coarctation in childhood. Am J Cardiol. 1986;58(1):178–80.
40. Clarkson PM, Brandt PW, Barratt-Boyes BG, Rutherford JD, Kerr AR, Neutze JM. Prosthetic repair of coarctation of the aorta with particular reference to Dacron onlay patch grafts and late aneurysm formation. Am J Cardiol. 1985;56(4):342–6.
41. Ala-Kulju K, Heikkinen L. Aneurysms after patch graft aortoplasty for coarctation of the aorta: long-term results of surgical management. Ann Thorac Surg. 1989;47(6):853–6.
42. Parks WJ, Ngo TD, Plauth WH Jr, Bank ER, Sheppard SK, Pettigrew RI, et al. Incidence of aneurysm formation after Dacron patch aortoplasty repair for coarctation of the aorta: long-term results and assessment utilizing magnetic resonance angiography with three-dimensional surface rendering. J Am Coll Cardiol. 1995;26(1):266–71.
43. Waldhausen JA, Nahrwold DL. Repair of coarctation of the aorta with a subclavian flap. J Thorac Cardiovasc Surg. 1966;51(4):532–3.

44. Pierce WS, Waldhausen JA, Berman W Jr, Whitman V. Late results of the subclavian flap procedure in infants with coarctation of the thoracic aorta. Circulation. 1978;58(3 Pt 2):I78–82.
45. Pandey R, Jackson M, Ajab S, Gladman G, Pozzi M. Subclavian flap repair: review of 399 patients at median follow-up of fourteen years. Ann Thorac Surg. 2006;81(4):1420–8.
46. van Son JA, van Asten WN, van Lier HJ, Daniels O, Vincent JG, Skotnicki SH, et al. Detrimental sequelae on the hemodynamics of the upper left limb after subclavian flap angioplasty in infancy. Circulation. 1990;81(3):996–1004.
47. Kaushal S, Backer CL, Patel JN, Patel SK, Walker BL, Weigel TJ, et al. Coarctation of the aorta: midterm outcomes of resection with extended end-to-end anastomosis. Ann Thorac Surg. 2009;88(6):1932–8.
48. Amato JJ, Rheinlander HF, Cleveland RJ. A method of enlarging the distal transverse arch in infants with hypoplasia and coarctation of the aorta. Ann Thorac Surg. 1977;23(3):261–3.
49. Thomson JD, Mulpur A, Guerrero R, Nagy Z, Gibbs JL, Watterson KG. Outcome after extended arch repair for aortic coarctation. Heart. 2006;92(1):90–4.
50. Burch PT, Cowley CG, Holubkov R, Null D, Lambert LM, Kouretas PC, et al. Coarctation repair in neonates and young infants: is small size or low weight still a risk factor? J Thorac Cardiovasc Surg. 2009;138(3):547–52.
51. Wright GE, Nowak CA, Goldberg CS, Ohye RG, Bove EL, Rocchini AP. Extended resection and end-to-end anastomosis for aortic coarctation in infants: results of a tailored surgical approach. Ann Thorac Surg. 2005;80(4):1453–9.
52. Mery CM, Guzman-Pruneda FA, Carberry KE, Watrin CH, McChesney GR, Chan JG, et al. Aortic arch advancement for aortic coarctation and hypoplastic aortic arch in neonates and infants. Ann Thorac Surg. 2014;98(2):625–33. discussion 33.
53. Sakurai T, Stickley J, Stumper O, Khan N, Jones TJ, Barron DJ, et al. Repair of isolated aortic coarctation over two decades: impact of surgical approach and associated arch hypoplasia. Interact Cardiovasc Thorac Surg. 2012;15(5):865–70.
54. Lock JE, Bass JL, Amplatz K, Fuhrman BP, Castaneda-Zuniga W. Balloon dilation angioplasty of aortic coarctations in infants and children. Circulation. 1983;68(1):109–16.
55. Tynan M, Finley JP, Fontes V, Hess J, Kan J. Balloon angioplasty for the treatment of native coarctation: results of Valvuloplasty and Angioplasty of Congenital Anomalies Registry. Am J Cardiol. 1990;65(11):790–2.
56. Mendelsohn AM, Lloyd TR, Crowley DC, Sandhu SK, Kocis KC, Beekman RH 3rd. Late follow-up of balloon angioplasty in children with a native coarctation of the aorta. Am J Cardiol. 1994;74(7):696–700.
57. Fawzy ME, Fathala A, Osman A, Badr A, Mostafa MA, Mohamed G, et al. Twenty-two years of follow-up results of balloon angioplasty for discreet native coarctation of the aorta in adolescents and adults. Am Heart J. 2008;156(5):910–7.
58. Harris KC, Du W, Cowley CG, Forbes TJ, Kim DW. Congenital Cardiac Intervention Study C. A prospective observational multicenter study of balloon angioplasty for the treatment of native and recurrent coarctation of the aorta. Catheter Cardiovasc Interv. 2014;83(7):1116–23.
59. Cowley CG, Orsmond GS, Feola P, McQuillan L, Shaddy RE. Long-term, randomized comparison of balloon angioplasty and surgery for native coarctation of the aorta in childhood. Circulation. 2005;111(25):3453–6.
60. Ho SY, Somerville J, Yip WC, Anderson RH. Transluminal balloon dilation of resected coarcted segments of thoracic aorta: histological study and clinical implications. Int J Cardiol. 1988;19(1):99–105.
61. Ino T, Kishiro M, Okubo M, Akimoto K, Nishimoto K, Yabuta K, et al. Dilatation mechanism of balloon angioplasty in children: assessment by angiography and intravascular ultrasound. Cardiovasc Intervent Radiol. 1998;21(2):102–8.
62. Lock JE, Castaneda-Zuniga WR, Bass JL, Foker JE, Amplatz K, Anderson RW. Balloon dilatation of excised aortic coarctations. Radiology. 1982;143(3):689–91.
63. Sohn S, Rothman A, Shiota T, Luk G, Tong A, Swensson RE, et al. Acute and follow-up intravascular ultrasound findings after balloon dilation of coarctation of the aorta. Circulation. 1994;90(1):340–7.
64. Saxena A. Recurrent coarctation: interventional techniques and results. World J Pediatr Congenit Heart Surg. 2015;6(2):257–65.
65. Yetman AT, Nykanen D, McCrindle BW, Sunnegardh J, Adatia I, Freedom RM, et al. Balloon angioplasty of recurrent coarctation: a 12-year review. J Am Coll Cardiol. 1997;30(3):811–6.
66. Reich O, Tax P, Bartakova H, Tomek V, Gilik J, Lisy J, et al. Long-term (up to 20 years) results of percutaneous balloon angioplasty of recurrent aortic coarctation without use of stents. Eur Heart J. 2008;29(16):2042–8.
67. O'Laughlin MP, Perry SB, Lock JE, Mullins CE. Use of endovascular stents in congenital heart disease. Circulation. 1991;83(6):1923–39.
68. Bentham J, Shettihalli N, Orchard E, Westaby S, Wilson N. Endovascular stent placement is an acceptable alternative to reoperation in selected infants with residual or recurrent aortic arch obstruction. Catheter Cardiovasc Interv. 2010;76(6):852–9.
69. Mohan UR, Danon S, Levi D, Connolly D, Moore JW. Stent implantation for coarctation of the aorta in children <30 kg. JACC Cardiovasc Interv. 2009;2(9):877–83.
70. Meadows J, Minahan M, McElhinney DB, McEnaney K, Ringel R. Investigators* C. intermediate outcomes in the prospective, multicenter Coarctation of the Aorta Stent Trial (COAST). Circulation. 2015;131(19):1656–64.
71. Tzifa A, Ewert P, Brzezinska-Rajszys G, Peters B, Zubrzycka M, Rosenthal E, et al. Covered Cheatham-platinum stents for aortic coarctation: early and intermediate-term results. J Am Coll Cardiol. 2006;47(7):1457–63.
72. Taggart NW, Minahan M, Cabalka AK, Cetta F, Usmani K, Ringel RE, et al. Immediate outcomes of covered stent placement for treatment or prevention of aortic wall injury associated with Coarctation of the Aorta (COAST II). JACC Cardiovasc Interv. 2016;9(5):484–93.
73. Sohrabi B, Jamshidi P, Yaghoubi A, Habibzadeh A, Hashemi-Aghdam Y, Moin A, et al. Comparison between covered and bare Cheatham-Platinum stents for endovascular treatment of patients with native post-ductal aortic coarctation: immediate and intermediate-term results. JACC Cardiovasc Interv. 2014;7(4):416–23.
74. Kische S, D'Ancona G, Stoeckicht Y, Ortak J, Elsasser A, Ince H. Percutaneous treatment of adult isthmic aortic coarctation: acute and long-term clinical and imaging outcome with a self-expandable uncovered nitinol stent. Circ Cardiovasc Interv. 2015;8(1).
75. Campbell M. Natural history of coarctation of the aorta. Br Heart J. 1970;32(5):633–40.
76. Morgan GJ, Lee KJ, Chaturvedi R, Bradley TJ, Mertens L, Benson L. Systemic blood pressure after stent management for arch coarctation implications for clinical care. JACC Cardiovasc Interv. 2013;6(2):192–201.
77. Hager A, Kanz S, Kaemmerer H, Schreiber C, Hess J. Coarctation Long-term Assessment (COALA): significance of arterial hypertension in a cohort of 404 patients up to 27 years after surgical repair of isolated coarctation of the aorta, even in the absence

of restenosis and prosthetic material. J Thorac Cardiovasc Surg. 2007;134(3):738–45.
78. Rinnstrom D, Dellborg M, Thilen U, Sorensson P, Nielsen NE, Christersson C, et al. Hypertension in adults with repaired coarctation of the aorta. Am Heart J. 2016;181:10–5.
79. Cook SC, Hickey J, Maul TM, Zumberge N, Krieger EV, Valente AM, et al. Assessment of the cerebral circulation in adults with coarctation of the aorta. Congenit Heart Dis. 2013;8(4):289–95.
80. Ou P, Bonnet D, Auriacombe L, Pedroni E, Balleux F, Sidi D, et al. Late systemic hypertension and aortic arch geometry after successful repair of coarctation of the aorta. Eur Heart J. 2004;25(20):1853–9.
81. Ou P, Mousseaux E, Celermajer DS, Pedroni E, Vouhe P, Sidi D, et al. Aortic arch shape deformation after coarctation surgery: effect on blood pressure response. J Thorac Cardiovasc Surg. 2006;132(5):1105–11.
82. Bruse JL, Khushnood A, McLeod K, Biglino G, Sermesant M, Pennec X, et al. How successful is successful? Aortic arch shape after successful aortic coarctation repair correlates with left ventricular function. J Thorac Cardiovasc Surg. 2016.
83. De Caro E, Trocchio G, Smeraldi A, Calevo MG, Pongiglione G. Aortic arch geometry and exercise-induced hypertension in aortic coarctation. Am J Cardiol. 2007;99(9):1284–7.
84. Ntsinjana HN, Biglino G, Capelli C, Tann O, Giardini A, Derrick G, et al. Aortic arch shape is not associated with hypertensive response to exercise in patients with repaired congenital heart diseases. J Cardiovasc Magn Reson. 2013;15:101.
85. Ou P, Celermajer DS, Mousseaux E, Giron A, Aggoun Y, Szezepanski I, et al. Vascular remodeling after "successful" repair of coarctation: impact of aortic arch geometry. J Am Coll Cardiol. 2007;49(8):883–90.
86. Graham TP Jr, Driscoll DJ, Gersony WM, Newburger JW, Rocchini A, Towbin JA. Task Force 2: congenital heart disease. J Am Coll Cardiol. 2005;45(8):1326–33.
87. Baumgartner H, Bonhoeffer P, De Groot NM, de Haan F, Deanfield JE, Galie N, et al. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J. 2010;31(23):2915–57.
88. Wilson W, Taubert KA, Gewitz M, Lockhart PB, Baddour LM, Levison M, et al. Prevention of infective endocarditis: guidelines from the American Heart Association: a guideline from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young, and the Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and the Quality of Care and Outcomes Research Interdisciplinary Working Group. Circulation. 2007;116(15):1736–54.
89. Nasser WK, Helmen C. Kinking of the aortic arch (pseudocoarctation). Clinical, radiographic, hemodynamic, and angiographic findings in eight cases. Ann Intern Med. 1966;64(5):971–8.
90. Singh S, Hakim FA, Sharma A, Roy RR, Panse PM, Chandrasekaran K, et al. Hypoplasia, pseudocoarctation and coarctation of the aorta - a systematic review. Heart Lung Circ. 2015;24(2):110–8.
91. Munjal AK, Rose WS, Williams G. Magnetic resonance imaging of pseudocoarctation of the aorta: a case report. J Thorac Imaging. 1994;9(2):88–91.
92. Cheng TO. Pseudocoarctation of the aorta. An important consideration in the differential diagnosis of superior mediastinal mass. Am J Med. 1970;49(4):551–5.
93. Steinberg I, Engle MA, Holswade GR, Hagstrom JW. Pseudocoarctation of the aorta associated with congenital heart disease: report of ten cases. Am J Roentgenol Radium Therapy, Nucl Med. 1969;106(1):1–20.
94. Amico A, Michaud JL, Baron O, Al Habash O, Despins P, Gaudin R, et al. Pseudocoarctation as cause of a refractory hypertension. J Cardiovasc Med (Hagerstown). 2007;8(4):289–90.
95. Atalay MK, Kochilas LK. Familial pseudocoarctation of the aorta. Pediatr Cardiol. 2011;32(5):692–5.
96. Connolly JE, Wilson SE, Lawrence PL, Fujitani RM. Middle aortic syndrome: distal thoracic and abdominal coarctation, a disorder with multiple etiologies. J Am Coll Surg. 2002;194(6):774–81.
97. Rumman RK, Nickel C, Matsuda-Abedini M, Lorenzo AJ, Langlois V, Radhakrishnan S, et al. Disease Beyond the Arch: A Systematic Review of Middle Aortic Syndrome in Childhood. Am J Hypertens. 2015;28(7):833–46.
98. Coleman DM, Eliason JL, Ohye RG, Stanley JC. Long-segment thoracoabdominal aortic occlusions in childhood. J Vasc Surg. 2012;56(2):482–5.
99. Stanley JC, Criado E, Eliason JL, Upchurch GR Jr, Berguer R, Rectenwald JE. Abdominal aortic coarctation: surgical treatment of 53 patients with a thoracoabdominal bypass, patch aortoplasty, or interposition aortoaortic graft. J Vasc Surg. 2008;48(5):1073–82.
100. Moresco KP, Shapiro RS. Abdominal aortic coarctation: CT, MRI, and angiographic correlation. Comput Med Imaging Graph. 1995;19(5):427–30.
101. Orellana-Barrios MA, Rosales A, Karkoutly A, Meyerrose GE. Middle aortic syndrome: imaging and endovascular treatment. Tex Heart Inst J. 2015;42(6):588–9.
102. Tummolo A, Marks SD, Stadermann M, Roebuck DJ, McLaren CA, Hamilton G, et al. Mid-aortic syndrome: long-term outcome of 36 children. Pediatr Nephrol. 2009;24(11):2225–32.
103. Day E, Stojanovic J, Karunanthy N, Qureshi S, Reidy J, Sinha MD. Middle aortic syndrome--an 8-year story of pills, pretty balloons and struts. Pediatr Nephrol. 2015;30(8):1361–5.
104. Carr JA. The results of catheter-based therapy compared with surgical repair of adult aortic coarctation. J Am Coll Cardiol. 2006;47(6):1101–7.
105. Forbes TJ, Garekar S, Amin Z, Zahn EM, Nykanen D, Moore P, et al. Procedural results and acute complications in stenting native and recurrent coarctation of the aorta in patients over 4 years of age: a multi-institutional study. Catheter Cardiovasc Interv. 2007;70(2):276–85.
106. Holzer R, Qureshi S, Ghasemi A, Vincent J, Sievert H, Gruenstein D, et al. Stenting of aortic coarctation: acute, intermediate, and long-term results of a prospective multi-institutional registry--Congenital Cardiovascular Interventional Study Consortium (CCISC). Catheter Cardiovasc Interv. 2010;76(4):553–63.
107. Forbes TJ, Kim DW, Du W, Turner DR, Holzer R, Amin Z, et al. Comparison of surgical, stent, and balloon angioplasty treatment of native coarctation of the aorta: an observational study by the CCISC (Congenital Cardiovascular Interventional Study Consortium). J Am Coll Cardiol. 2011;58(25):2664–74.