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The Adult with Coarctation of the Aorta

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Abstract

The manuscript will discuss the epidemiology and etiology of the adult with coarctation of the aorta (CoA) as well as describe the embryology, anatomy, pathophysiology, and clinical presentation in order to recognize and appropriately diagnose an adult patient with CoA. This chapter will also review diagnostic testing, management, therapeutic interventions including percutaneous and surgical procedures, and long-term complications that can arise in an adult with repaired CoA. It contains images with examples from echocardiography, cardiac computed tomography (CT), magnetic resonance imaging (MRI), and angiograms as part of the description.

Keywords: congenital heart disease, coarctation of the aorta, percutaneous intervention, balloon dilatation, stenting, surgery, management, complications

1. Introduction

Coarctation of the aorta is a congenital cardiac defect. It usually manifests as a discrete constriction of the aortic isthmus. However, it is more likely to represent a spectrum of aortic narrowing from this discrete entity to tubular hypoplasia, with many variations seen in between these two extremes. Morphologists argue that tubular hypoplasia, although it may coexist with discrete coarctation, should be considered as a separate entity [1]. On rare occasions there can be a gap between the ascending and descending thoracic aorta, known as an interrupted aortic arch. Interventions can be required as an infant however procedures may be needed later in life for native coarctation or patients with recurrent coarctation. The presence of associated arch hypoplasia is relevant to longer term risk for the development of hypertension so in addition to re-coarctation, these patients are at increased risk for developing other comorbidities and should have lifelong follow up care.

2. Epidemiology

Coarctation of the aorta (CoA) is the fifth most common congenital heart defect, accounting for 6–8% of live births with congenital heart disease, with an estimated incidence of 1 in 2500 births [2–5]. It affects more male babies than female, with a reported ratio in males of between 1.27:1 and 1.74:1 [6, 7]. Patients with CoA can have other defects like atrial septal defect (ASD), ventricular septal defect (VSD), atrioventricular canal defect (AVCD), bicuspid aortic valve (BAV), transposition of great arteries (TGA), patent ductus arteriosus (PDA), hypoplastic left heart syndrome. CoA often coexists with other left heart obstructive lesions like mitral stenosis, subaortic stenosis and aortic stenosis. About 50–60% patients with coarctation of the aorta or interrupted aortic arch have a BAV [8]. Compared to right-sided lesions, left-sided cardiac obstructions are more frequently seen in males than female [2]. One genetic condition noted to be associated with an increased risk of having coarctation of the aorta (12–35%) is Turner syndrome [9]. Lastly, the etiology of CoA is not well understood and thought to be affected by various factors including a genetic component, environmental factors, and arteriopathy.

3. Etiology and cardiac associations

The etiology of the discrete isthmus constriction of the aorta seen in patients with CoA remains controversial and is thought to be multifactorial. Although the precise pathogenesis is unknown, the two theories for the development of congenital coarctation of the aorta have been postulated: reduced antegrade intrauterine blood flow causing underdevelopment of the fetal aortic arch [10] and migration or extension of ductal tissue into the wall of the fetal thoracic aorta [11]. Histologic examination of localized aortic coarctation lesions has demonstrated the presence of a tissue ridge extending from the posterior aortic wall and protruding into aortic lumen. This ridge consists of ductal tissue with in-folding of the aortic media [12]. Prenatal environmental exposures have been associated with CoA and other left-sided lesions. However, there is a growing body of literature that suggests a genetic basis for development of these lesions [13]. There has been evidence of genetic contribution to CoA [14, 15]. Vascular endothelial growth factor (VEGF) plays a vital role in aortic development, acting as a chemo-attractant, stimulating angioblast migration toward the midline before formation of the aorta. Indeed, targeted disruption of VEGF in mice leads to significant disruption of the developing aorta [16]. Whether an initial mutation leads to secondary effects on VEGF or on other signaling systems involved in recruiting mural cells in fetuses, leading to CoA, is unknown. An increase in collagen and decrease in smooth muscle content of the pre-coarctation aorta in humans has been demonstrated in comparison to post-coarctation aorta or to proximal aorta of young transplant donors [17]. Recently, mutations in the NOTCH1 gene have been identified in individuals with left ventricular outflow tract malformation, including coarctation [18]. Up to 18–30% of patients with Turner syndrome have coarctation [19]. Genetic testing for Turner syndrome (i.e., karyotype analysis) should therefore be performed in female patients diagnosed with coarctation of the aorta [20, 21]. Mechanical models have suggested that abnormalities of blood flow, defective endothelial cell migration, and excessive deposition

of aortic duct tissue at the aortic isthmus can result in coarctation [22]. Epidemiological studies have found that for left ventricular outflow tract lesions, there is a higher chance of concordant diagnosis in multiple family members [23]. The co-existence of CoA with other left heart obstructive pathologies like aortic stenosis and hypoplastic left heart syndrome suggests that there could be a common pathogenic mechanism at a molecular level [24, 25]. Williams syndrome, a congenital and multisystem genetic disorder, has been associated with supra-ventricular aortic stenosis. Aortic arch abnormalities, including coarctation, are present in 10% of patients with Williams syndrome [26]. Coarctation can also be present in congenital cardiovascular anomalies involving multiple left-sided lesions, including Shone syndrome and hypoplastic left heart syndrome [22]. Environmental factors could also play a role in the incidence of CoA since there is increase CoA rate along the US-Mexico border [27]. Seasonal variations have been reported in the incidence of CoA we well [28].

4. Noncardiac associations

The link between intracranial aneurysms and CoA was described well before the surgical era, accounting for 5% deaths in patients with aortic coarctation on autopsy review [29]. Most of the aneurysms described are small, and therefore have a low risk of spontaneous rupture. Currently the benefits of routine screening for intracranial aneurysms in coarctation remain unclear [22].

5. Anatomy

Although most patients have a discrete narrowing of the descending aorta at the insertion of the ductus arteriosus, there is a spectrum of aortic narrowing that encompasses the usual discrete thoracic lesions, long-segmental defects, tubular hypoplasia, and, rarely, coarctation located in the abdominal aorta. In simple terms, coarctation is characterized by discrete narrowing of the thoracic aorta adjacent to the ligamentum arteriosum. Importantly, discrete coarctation is an aortopathy that lies within a spectrum of arch abnormalities ranging from discrete narrowing to a long segment of arch hypoplasia. Morphologically it appears as a localized shelf in the posterolateral aortic wall. There are some anatomic variations of coarctation. It maybe appear as (a) discrete narrowing (b) tubular hypoplasia of any part of the arch or (c) aortic arch interruption [30]. CoA has also been described as a diffuse arteriopathy with abnormalities in the elastic properties of the aorta. Increase in collagen and decreases in smooth muscle component of the pre-coarctation aorta have been reported [31].

Although CoA can be an isolated CHD, it is also commonly found in other congenital syndromes and cardiovascular anomalies. Thus, deliberate investigation for the presence of coarctation should be made in these patients. The most common cardiovascular malformation associated with CoA is BAV. Prior autopsy examination showed 46% of patients with CoA have congenital BAV [32]. The relative frequency of associated cardiac lesions in patients with CoA differs somewhat based upon the age of the population studies. Adult patients with CoA

evaluated with magnetic resonance imaging, 17% had no additional cardiovascular anomalies however, in this cohort, BAV, arch hypoplasia, VSD, and PDA were detected in 60, 14, 13, and 7% of patients, respectively [33]. The coincidence of BAV and CoA is difficult to determine, because BAV is very common and not everyone is screened for the presence of coarctation.

6. Pathophysiology and presentation

The clinical presentation of coarctation differs significantly in pediatric patients in comparison with adults. Although infants with severe coarctation may present with signs and symptoms of heart failure and cardiogenic shock as the ductus closes, most adults with unrepaired coarctation are generally asymptomatic. A common presentation of coarctation is systemic arterial hypertension. The causes of hypertension in this cohort of patients are not fully understood, but malfunction in a number of individual systems have been implicated, including imbalance within the autonomic nervous system [34], impaired vascular function [35, 36] and hyperactivation of the rennin–angiotensin system [37, 38]. It is likely that more than one of these systems is involved. In young adults presenting with severe upper extremity hypertension, coarctation should be excluded. Patients presenting with severe hypertension may experience symptoms including angina, headache, epistaxis, and heart failure [22]. Coarctation causes upper extremity hypertension, which leads to systemic hypertension and left ventricular hypertrophy. There have been several mechanisms proposed for hypertension in patients with coarctation, which include reduced arterial compliance, blunted baroreceptor sensitivity and endothelial dysfunction [39]. Age at repair is an important determinant of developing late hypertension. Patients who get the repair in infancy have less than 5% chance of developing hypertension by early adulthood, whereas those operated on after the age of one have a 25–33% chance of developing hypertension [40–42]. Late hypertension is associated with residual or recurrent obstruction. Despite the variability in blood pressure in the upper and lower extremities, regional blood flow is generally maintained within normal limits by autoregulatory vasoconstriction in the hypertensive areas and by vasodilation in the hypotensive areas [43]. Nonetheless some patients with satisfactory repair can still develop late hypertension due to vascular dysfunction [44]. In a systemic review of literature, the median prevalence of late hypertension after satisfactory repair was reported to be 32% with a range of 25–68% [45]. Ambulatory blood pressure monitoring can help in early diagnosis of late hypertension [46]. Patients with coarctation remain at a high risk of developing complications like premature coronary atherosclerosis, cerebrovascular events, left ventricular systolic dysfunction and endocarditis. With early repair, timely recognition of late hypertension and treatment of risk factors, the overall survival has improved. However, the life expectancy of these individuals is not as normal as the unaffected peers [47].

In full term newborns one of the important causes of congestive heart failure is aortic coarctation. Beyond the neonatal period most patients are asymptomatic and present with difficult to control hypertension in later years. In previously undiagnosed adults, the classic presenting sign is hypertension. Older patients might complain of headaches, leg fatigue with exercise and cold extremities. As mentioned previously, coarctation can be a part of syndromes like

Turner syndrome, Williams Syndrome or Shone's complex. Almost 50% of the cases are associated with BAV. Other associated abnormalities include intracranial aneurysms (most commonly of the circle of Willis) in 2–10% case and acquired intercostal artery aneurysms [30]. Data on the natural history of coarctation of the aorta are largely derived from hospital post-mortem records and from case series prior to the availability of operative repair that was first done in 1945 [48]. The average survival age of individuals with unoperated coarctation was approximately 35 years of age, with 75% mortality by 46 years of age [49]. Common complications in unoperated patients or in those operated on during later childhood or adulthood were systemic hypertension, accelerated coronary artery disease, stroke, aortic dissection, and heart failure. Causes of death include heart failure, aortic rupture, aortic dissection, endocarditis, endarteritis, intra-cerebral hemorrhage, and myocardial infarction [48, 50]. Patients with an associated BAV may also develop significant aortic stenosis, aortic regurgitation, and dilated ascending aorta from myxomatous degeneration of the medial wall of the aorta.

6.1. Pregnancy and coarctation

Coarctation of the aorta and associated lesions, particularly BAV, aortic stenosis, and ascending aorta dilation should be evaluated before pregnancy for appropriate counseling and advice. Rarely the first manifestation is during pregnancy. In the absence of hemodynamically significant stenotic lesion, pregnancy is well tolerated in patients with repaired aortic coarctation. However there is a greater propensity of developing hypertension during pregnancy [51]. Outcome of pregnancy in patients after repair of aortic coarctation have been reported over the last decade [52]. During pregnancy and delivery, there were no serious cardiovascular complications. Hypertension alone was reported in 21 pregnancies in 14 women, and preeclampsia in 5 pregnancies in 4 women. In another study, serious complications were uncommon in women with a hemodynamically significant gradient (≥ 20 mmHg) after repair [53]. These women were more likely to have systemic hypertension related to the increased coarctation gradient. However, there are case reports of aortic rupture or dissection that occur with pregnancy after coarctation repair due to the hemodynamic and aortic medial changes of pregnancy, which remain rare.

7. Physical exam

On physical examination, femoral arterial pulses are diminished and usually delayed. Rarely, claudication may be reported because of lower extremity ischemia. Auscultation of the left sternal border may demonstrate a harsh systolic murmur with radiation to the back. An associated thrill may be palpable in the suprasternal notch. If left ventricular pressure or volume overload have developed, a left ventricular lift can be present. The finding of a continuous murmur may suggest the presence of arterial collaterals in those with long-standing unrepaired significant coarctation [22].

If aortic coarctation is suspected blood pressure should be measured in both arms and legs in supine position. Normally BP in the lower extremities is 10–20% higher than the upper

extremities due to wave amplification. If BP in the leg is lower than the arm BP by 10 mmHg or more then coarctation should be suspected. A pressure gradient of 35 mmHg or greater is considered highly specific for coarctation [54]. The presence of collateral vessels may diminish the pressure gradient. Arterial pulsations from collaterals to the intercostal and interscapular arteries can also be palpated. In patients with suspected coarctation, it is important to assess for systolic blood pressure discrepancy between upper and lower extremities. The upper extremity systolic blood pressure is usually 20 mmHg higher than the lower extremities in patients with significant coarctation. In rare instances of coarctation patients with concomitant anomalous subclavian artery origin distal to the coarctation, systolic blood pressure differences may not be detected between ipsilateral arm and legs. On auscultation a continuous murmur of aorto-aortic collateral arteries would be audible in the interscapular space. Simultaneous palpitation of radial and femoral artery might reveal a delay or absence of the femoral pulse.

8. Diagnostic evaluation

8.1. Initial workup

The electrocardiogram of a patient with coarctation may be normal or demonstrate evidence of left ventricular hypertrophy from chronic left ventricular pressure overload. On chest radiograph, a “figure of three” sign formed by the aortic knob, the stenotic segment, and the dilated post stenotic segment of the aorta suggests CoA. The heart border can be normal or mildly enlarged. Inferior rib notching can also be seen in the third to eighth ribs bilaterally caused by the presence of dilated intercostal collateral arteries [22, 30].

8.2. Echocardiography

Transthoracic echocardiography is the most accessible and the main stay for the practicing physician. A comprehensive echocardiogram is recommended in the initial evaluation of a patient with repaired or suspected CoA. In addition to characterization of the coarctation itself, it is important to evaluate for evidence of left ventricular pressure or volume overload, left ventricular hypertrophy, size, and left ventricular systolic and diastolic dysfunction. Particular attention should be placed in identifying associated cardiac defects especially left-sided lesions. The morphology of the aortic valve, and evidence of subvalvular, valvular, and supra-valvular aortic stenosis should be interrogated. The dimensions of the aortic root and ascending aorta can be followed serially to assess for associated aortopathy. Suprasternal windows are important to view the aortic arch from the long-axis view, in two-dimensional imaging and by color flow Doppler. Visualization of the aortic arch in the long axis may demonstrate a focal area of narrowing of the thoracic aorta distal to the takeoff of the left subclavian artery with associated flow turbulence on color flow Doppler.

Coarctation is imaged from the high left parasternal views with lateral angulation of the probe toward the left shoulder. The suprasternal notch view is used for obtaining Doppler gradient (**Figures 1 and 2**). Subcostal imaging is used to evaluate the distal thoracic and upper

abdominal aorta. The stenotic segment may be discrete, segmental or long therefore the entire aortic arch should be imaged particularly the origin of the left subclavian artery as in transverse arch hypoplasia the distance between the origin of the left common carotid artery and left subclavian artery may be increased. Low frequency imaging and harmonic imaging can improve the image quality.

On doppler imaging color flow aliasing would be seen at and beyond the narrow segment. Systolic velocity in the descending aorta is increased. If transverse arch hypoplasia is present the proximal velocity increases as well therefore the systolic pressure gradient should be calculated with the expanded Bernoulli equation $4(V_2^2 - V_1^2)$ [55, 56]. In severe cases there is a gradient during both systole and diastole across the stenosis, which results in the classic saw tooth pattern. The presence of collateral arteries can cause doppler to underestimate the severity of obstruction [57]. Some of the other factors, which can affect the Doppler gradient, include severe obstruction, long tortuous vessels or eccentric gradient. Yet with long-standing coarctation, significant collaterals may have developed thereby reducing the peak systolic gradient across the site of stenosis. A saw-tooth pattern seen on continuous-wave Doppler reflects the persistent forward flow in diastole because of diastolic run-off. Higher gradient across the coarctation and longer duration of diastolic forward flow in the thoracic aorta suggest more significant coarctation [22].

In the absence of proximal obstruction when the pulse wave doppler is placed in the abdominal aorta, the wave form shows a rapid systolic upstroke, short deceleration time, followed by a brief early diastolic flow reversal and little antegrade flow throughout diastole. In the presence of coarctation there is loss of early diastolic flow reversal, which is highly sensitive

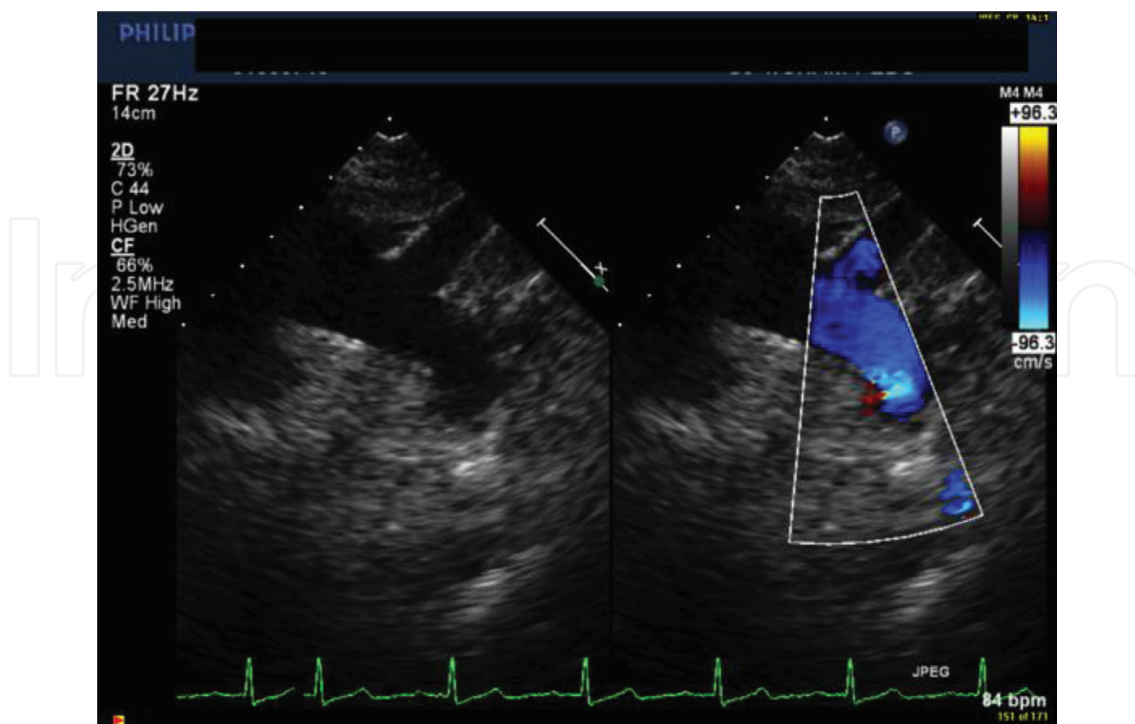


Figure 1. 2D transthoracic echo imaging showing coarctation of the aorta distal to the left subclavian artery.

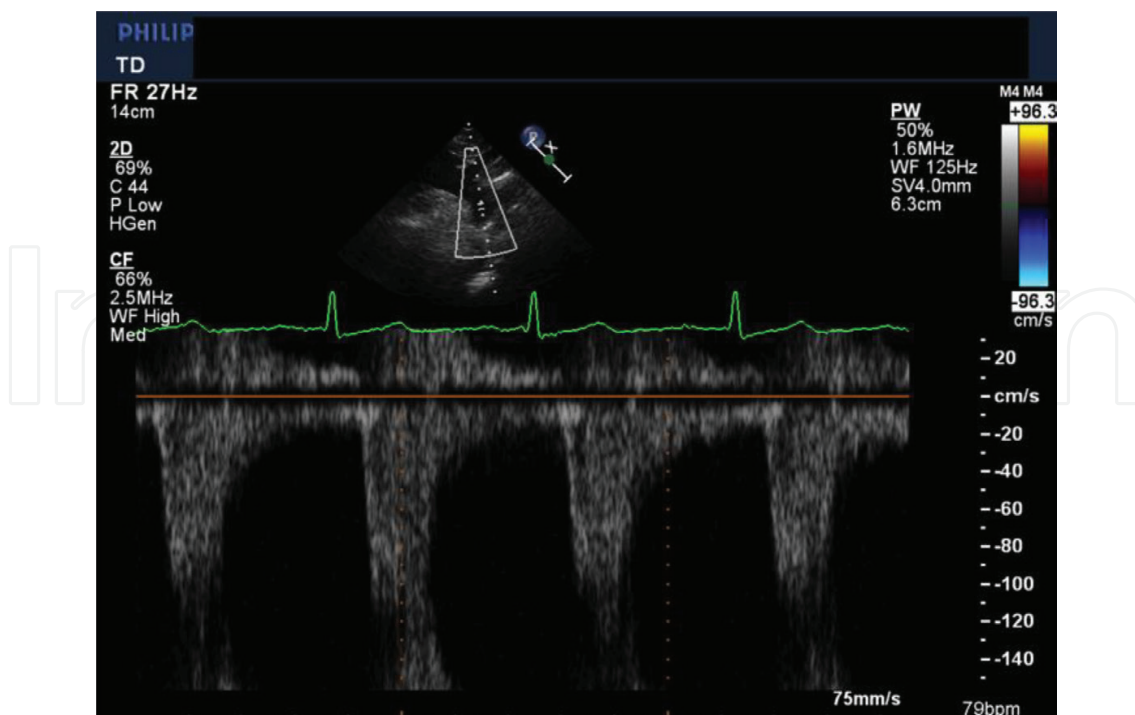


Figure 2. Pulse wave doppler profile through the coarctation segment demonstrating a pressure gradient.

for detection of upstream obstruction. The systolic velocity is blunted, there is continuous antegrade flow and increased diastolic flow velocity. If the delay between R wave on ECG and peak velocity of the abdominal aorta is >50 ms it is associated with coarctation [58].

8.3. Magnetic resonance imaging

Magnetic resonance imaging (MRI) is the most comprehensive method of evaluating coarctation of the aorta. MRI does not expose the patients to ionizing radiations, which is an important consideration for young patients who would have to undergo serial imaging. Cardiac MRI (cMRI) has become a valuable noninvasive modality to assess patients with unrepaired and repaired coarctation. In adults with suboptimal echocardiographic imaging window, cMRI can be used to characterize the aortic valve, aortic root, left ventricular size, and function. cMRI, along with gadolinium-enhanced magnetic resonance angiography, provides excellent resolution of cardiac anatomy and vascular structures. Compared with echocardiography, cMRI demonstrates superior visualization of the aortic arch with precise characterization of the location and extent of coarctation, and assessment of the presence and extent of collateral vessels. In the unrepaired patient, the measured minimum aortic cross-sectional area and heart rate–corrected deceleration time in the descending aorta can be used to predict a significant gradient by cardiac catheterization [59] and future need for interventions. cMRI provides exceptional visualization of the aortic arch and detection of post repair complications including arch “kinking” and pseudoaneurysms. Thoracic aortic magnetic resonance angiography also provides assessment of post stenotic dilation or aneurysmal formation at the site of a previous repair. Importantly, the lack of ionizing radiation provides an advantage of cMRI over CT, in the serial evaluation of late complications after repair [22, 59].

A stack of half-Fourier acquisition single shot turbo spin-echo (HASTE) images of the mediastinum are acquired in transverse, coronal and oblique sagittal plane parallel to the plane of the aortic arch. These provide dark blood images, which give anatomical overview of the coarctation. Black blood images are less susceptible to artifact from metallic objects. For evaluation of left ventricular function and mass a stack of steady state free precession (SSFP) cine images is acquired in the left ventricular short axis plane. Long axis cine images are acquired in the four, two and three chamber planes. SSFP cine images are then performed through the aortic root in the plane of the aortic valve, the aortic arch and the region of the aortic isthmus. Phase contrast flow imaging is performed to quantify the flow volumes and velocity [60].

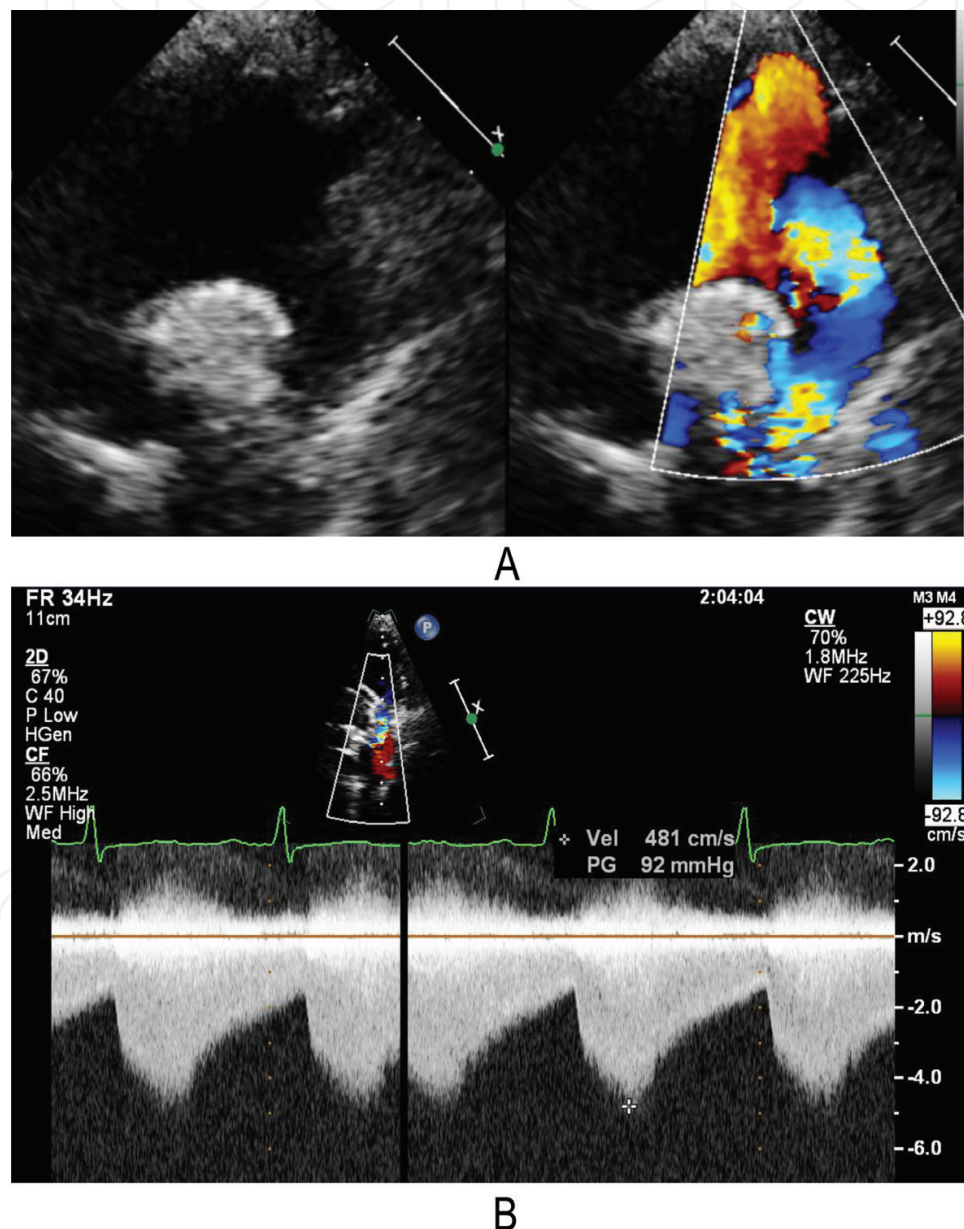


Figure 3. (A) 2D echo with color flow doppler showing severe narrowing of the proximal descending aorta with significant turbulence and a peak velocity of 4.8 m/s consistent with severe aortic coarctation. (B) Doppler tracing shows delay in return to baseline in diastole (diastolic drag) and blunting of the abdominal aortic doppler pattern consistent with significant aortic coarctation.

With phase contrast imaging the degree of collateral flow can be determined. The flow volume is assessed in the aorta just proximal to the stenosis and then at the level of the diaphragm. Usually there is a 7% decrease in total flow from proximal to distal aorta, if there is increase in flow by 5% or more, it is highly indicative of collateral flow joining the descending aorta [61]. Four-dimensional flow MR imaging is an emerging tool to evaluate hemodynamic significance of collateral blood flow (**Figure 3**) [62].

8.4. Computed tomographic angiography (CTA)

Although cMRI is the preferred mode of serial follow-up for patients after coarctation repair, the use of cardiovascular CT may be considered in selected patients. In particular, cMRI in patients with transcatheter stents may have susceptibility artifact precluding accurate assessment of late complications associated with these interventions. With cMRI, metallic artifact can lead to difficulty in the assessment of vessel lumen patency, identifying restenosis, aneurysm, or stent fracture [22]. Use of CT obviates concerns about metallic artifact impairing accurate assessment of stented segments of the aorta. Other advantages of cardiac CT over cMRI include improved image resolution, shorter scan time, and greater availability [22]. CTA is also used to assess concomitant coronary anomalies that may not be well visualized with cMRI. Patients with pacemakers or implantable cardioverter defibrillators that are not cMRI compatible may benefit from surveillance with cardiovascular CTA. Similar to cMRI, cardiovascular CT can be performed to image the coarctation segment, any aneurysmal dilation distal to the coarctation segment, recoarctation post repair, (**Figures 4–7**), hypoplasia of the



Figure 4. 3D reconstruction (CT angiogram) showing discrete segment of CoA and mild dilatation of the descending thoracic aorta distal to coarctation segment.



Figure 5. CT angiogram sagittal view of discrete coarctation segment distal to the left subclavian artery.

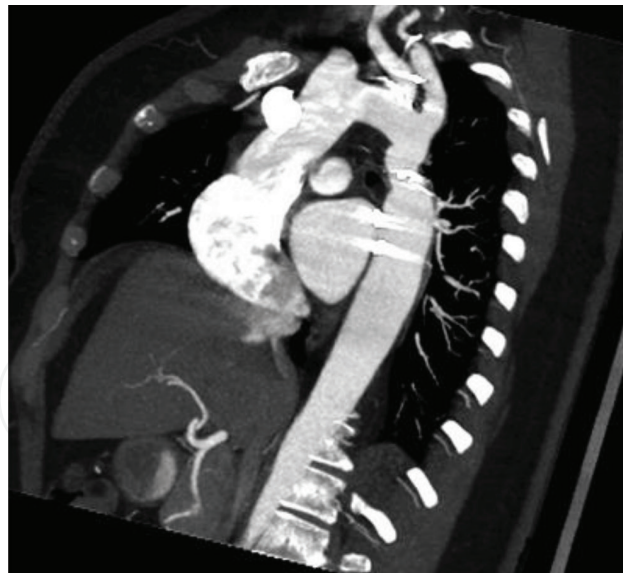


Figure 6. CT angiogram showing recurrent CoA.

aortic arch, follow serial aortic dimensions and can also show associated vascular anomalies such as double superior vena cava or aberrant great vessels. Collateral vessel formation can also be visualized with CTA. The main disadvantage of CTA is radiation exposure, therefore dose-saving algorithms are very important in reducing radiation exposure for patients (Figures 8–11).



Figure 7. CT 3D reconstruction of the aorta postsurgical repair of CoA.

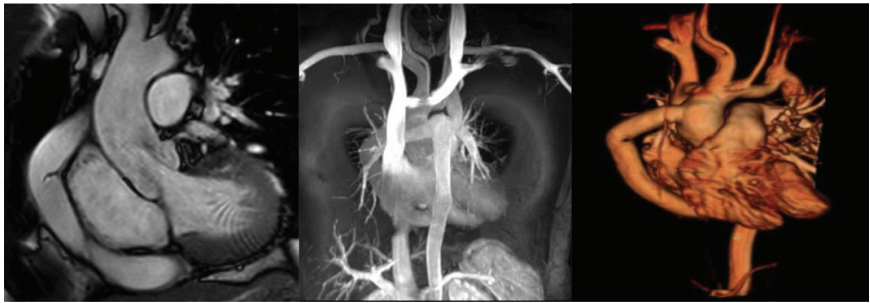


Figure 8. Cardiac MRI of interrupted aortic arch Type A status post a vascular jump graft resulting in a C-shaped appearance of the distal arch and multiple areas of stenosis now with a 20 mm extra-anatomic bypass graft from the mid ascending aorta to the distal descending aorta at the level of the diaphragm. The last picture in this figure shows a three dimensional (3D) reconstructed image of the graft.

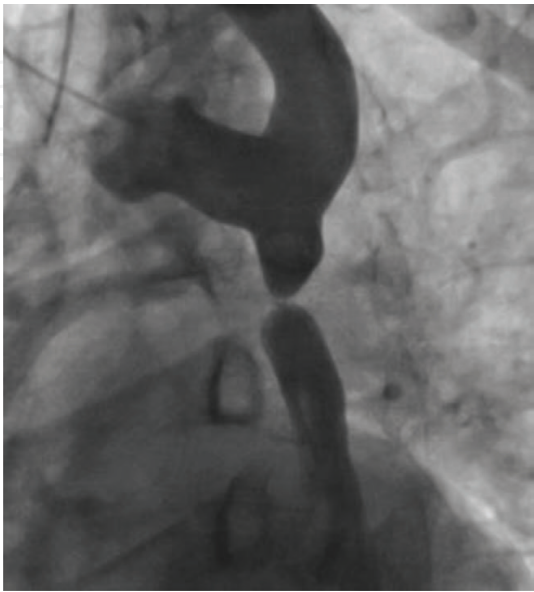


Figure 9. Invasive angiogram showing Type A interruption of the aorta.

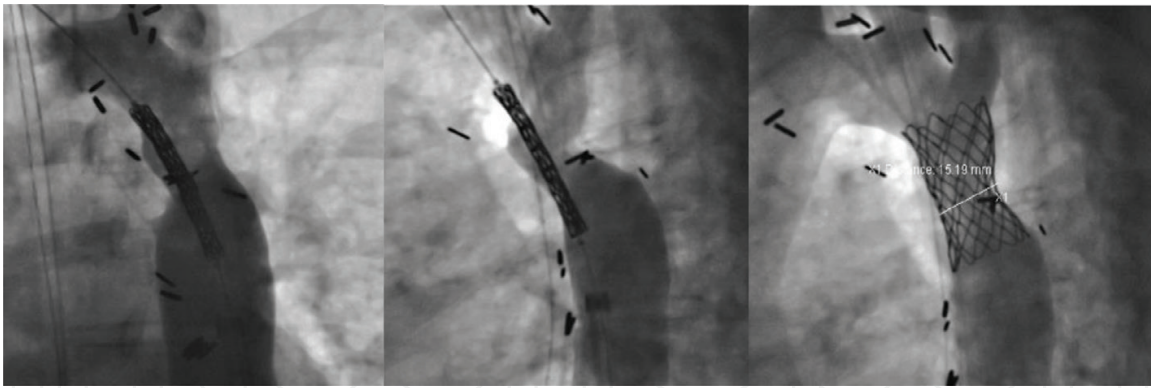


Figure 10. Angiography demonstrating stent placement in a patient with coarctation of the aorta.

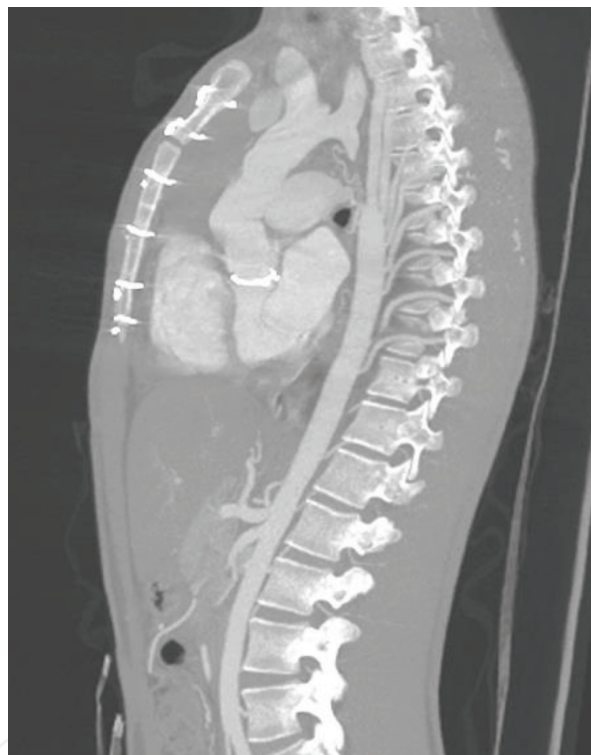


Figure 11. CT angiogram showing Type A interruption of the aortic arch.

9. Management

In 2008, the American College of Cardiology and American Heart Association (ACC/AHA) guidelines for adults with congenital heart disease recommended intervention for coarctation for the following indications:

- a. Peak to peak coarctation gradient ≥ 20 mmHg. The peak to peak gradient is a measurement derived from catheterization data in which the peak pressure beyond the coarctation is subtracted from the peak pressure proximal to the coarctation.
- b. Peak to peak coarctation gradient < 20 mmHg with anatomic imaging evidence of significant coarctation and radiologic evidence of significant collateral flow [49, 50].

Systemic hypertension, accelerated coronary heart disease, stroke, aortic dissection, and heart failure are common complications in adults who have not undergone correction for their coarctation or were operated on later in life [49]. Coarctation repair after early childhood does not prevent persistence or late recurrence of systemic hypertension. As a result, correction of coarctation should be performed in infancy or early childhood to prevent the development of chronic systemic hypertension [42]. If coarctation escapes early detection, repair should be performed at the time of subsequent diagnosis if clinically indicated. Management with antihypertensive medications is important to prevent long-term complications [2]. According to guidelines, the first line medications in the treatment of hypertension in patients with CoA are angiotensin converting enzyme (ACE) inhibitors, angiotensin-receptor blockers (ARB), and beta blockers (BB) [50]. Hypertension can be treated with medical management, but coarctation or recoarctation of the aorta need to be repaired either percutaneously or surgically [47, 63]. Choosing one intervention over another depends on the individual patient and should be done in collaboration with an interdisciplinary team including an adult congenital heart disease (ACHD) cardiologist, interventionalist and surgeon with training in ACHD. For example, patients with a long segment of coarctation of the aorta, complex arch anatomy, or with interruption of the aorta are more likely to need open-heart surgery as opposed to a transcatheter intervention [64, 65].

9.1. Percutaneous intervention

In the mid-1900s, repair of coarctation of the aorta was entirely surgical. Balloon angioplasty is a percutaneous alternative to surgical repair for older infants and young children (greater than 4 months) with native discrete coarctation. It remains the preferred intervention for all patients with isolated recoarctation regardless of age [49, 66]. However, stent placement has replaced balloon angioplasty as the procedure of choice in older children and adults with native coarctation [66]. Currently, balloon dilatation and stenting remain the transcatheter interventions that can be used for the treatment of CoA [63, 67]. Although balloon angioplasty was the treatment of choice for discrete native coarctation in adults in the past, most centers currently perform stent implantation for older children and adults with native discrete or long-segment coarctation. Through-out the years, continuous advancements in technology and catheter-based techniques have made a variety of percutaneous intervention possibilities available. Improvements in the field have allowed interventions to evolve from balloon angioplasty to endovascular stents to covered stents. The patients who underwent balloon angioplasty were noted to develop residual or recurrent stenosis, aneurysms and dissections or femoral artery complications including occlusion [68]. Stenting, on the other hand, have been shown to be superior to balloon dilatation in relieving the aortic coarctation, with less recurrent narrowing of the aorta, as well as having a smaller amount of complications. Studies have shown that balloon angioplasty and surgical correction are equally effective in reducing the peak systolic pressure gradient early after intervention [69]. The development of covered stents has helped decrease the number of problems associated with injury to the aortic wall and have allowed providers to avoid surgical interventions for aneurysms [2]. Overall, the repair of complex coarctation of the aorta with stents has been shown to be safe with improvements in outcomes [70]. Bare metal stents may be sufficient in many, if not most, patients that undergo stent placement and that further research is needed to determine if there is a subset

of patients who truly benefit from the implantation of a covered versus bare stent. Follow-up data will also be important to see if there is a long-term benefit regarding maintaining normal blood pressure using covered stents. Stenting may be less successful in patients with suboptimal anatomy with vessel tortuosity and transverse arch hypoplasia [71]. For these patients, the decision to perform stent placement versus surgical correction must be made on a case-by-case decision by the clinical team.

9.2. Surgical

Resection and direct end-to-end anastomosis or subclavian flap arterioplasty are the most commonly used techniques for the treatment of CoA in the infantile period because anatomic conditions are more favorable. Subclavian flap arterioplasty and patch graft aortoplasty have been developed as an alternative to resection and direct end-to-end anastomosis in which more than one-half of patients experience late-onset re-coarctation problems [72]. However, CoA in adolescents and adults is often complicated with the occurrence of associated comorbidities like aortic aneurysms, dissections, aortic valve disease, and other cardiovascular diseases. Studies actually show that having a BAV is a risk factor for mortality [73]. This is most likely because BAV have been associated with aortic insufficiency and stenosis in addition to dilatation and dissection of the aorta resulting in a potential need for open heart surgery. In a retrospective study of patients with CoA undergoing surgical interventions, aortic aneurysm or dissection and disease of the aortic valve were the most common comorbidities. Within this cohort, 38% had a BAV [72]. Patients with CoA who have left ventricular dysfunction and a brachial-ankle gradient of 20 mmHg or greater have also shown to be at risk for significant cardiovascular events [47]. Earlier, CoA was evaluated as the localization anomaly of the aorta; however, it is currently considered as part of a broad-spectrum pathology. The main goal of surgical treatment in CoA is the removal of stenosis. The surgical technique is selected according to the length of the coarcted segment, localization with the ductus, status of the collateral circulation in the distal aorta, and atherosclerotic alterations in the aortic wall [72]. The resection and graft interposition were first described by Gross in 1951 [74, 75]. This technique is not suitable for pediatric patients, because it restricts the development of the aorta. However, bypass grafting is an appropriate technique particularly for patients with aneurysms, long-segment coarctation or post-recovery aneurysms, and adult patients with diffuse collateral circulation and coarctations. Therefore, artificial bypass grafting was preferred in these patients to prevent complications (i.e., spinal cord complications, bleeding, and aneurysm development) during and after surgery [76]. Prosthetic patch aortoplasty is avoided whenever possible because of the frequent occurrence of aortic aneurysm or rupture [77]. When surgical repair of the coarctation is done at a later age, the possibility of these cardiovascular comorbidities should be kept in mind. Some of the other risks in surgery to consider are related to spinal ischemic injuries and intraoperative bleeding from extensive amounts of collaterals [2]. In general, repair of the CoA surgically has been shown to have a low mortality rate. However, as these patients continue to follow up with their cardiologists, re-coarctation is often seen in the long-term. Other than additional percutaneous procedures, these patients sometimes need to be evaluated for further surgical interventions [78]. Currently, there are no clinical trials showing a direct comparison between transcatheter approaches are superior to surgical interventions or vice versa. More research is needed in this area to compare the different approaches [64].

10. Complications and long-term follow up

All patients with coarctation (repaired or not) should be monitored with lifelong congenital cardiology follow-up and imaging because long-term survival is reduced compared with normative populations and there is potential need for reintervention [79, 80]. The European Society of Cardiology and the American Heart Association recommends continuous life-long follow up of patients with coarctation of the aorta even though they have been repaired [50]. As mentioned previously, even though patients with coarctation are repaired, they are at risk for re-coarctation later in life as well as develop other comorbidities such as hypertension and coronary artery disease. The unoperated mean survival rate of adults with coarctation of the aorta is 35 years of age, with a mortality rate of 75% by 46 years of age [49]. In general, the patients with CoA who are repaired at a later age are more likely to remain hypertensive. This is because in addition to the narrowing of the aorta, they can also develop arterial stiffness and vascular abnormalities besides from alternations in their renin-aldosterone angiotensin system [81]. Investigators have also postulated that the mechanical stress associated with increased pressure load may initiate rapid gene expression for collagen production, leading to re-enforcement and reorganization of the vessel musculo-elastic fascicle, and thereby reducing the degree of pressure-induced aortic dilatation. However, a clear disadvantage of this is that the resultant stiffer vessel will lead to augmented central aortic systolic pressure and systolic hypertension, which is the major cause of longer term morbidity and mortality in these patients, even despite early repair [82].

Other than being hypertensive at rest, it is also common for these patients to be hypertensive with exercise. In a prospective study of 74 patients with coarctation, the systolic blood pressure at peak exercise was an indicator for long term hypertension [83]. Exercise stress testing is useful to assess the patients' hypertensive response, evaluate their need for future interventions and determine prognosis in the long run [84]. Other studies have shown a link between exercise-induced hypertension and left ventricular hypertrophy (LVH) in patients with CoA. LVH has been shown to be associated with a higher incidence of adverse events [85]. Overall, more research is needed in this area to determine the risks and benefits of exercise in patients with CoA and whether there is a need for exercise restrictions. Patients with CoA are also at risk for developing intracranial aneurysms (ICA). With five times the risk of developing ICA, guidelines recommend advanced imaging such as CT or MRI to assess the intracranial vessels. Studies show that screening these patients is reasonable, especially as they get older, since age is one of the main risk factors in the prevalence of ICA [86]. Hypertension will put these patients with aneurysms at risk for cerebrovascular accidents (CVA) and intracranial hemorrhage. In one of the studies comparing patients with congenital heart disease with and without CoA, the patients with CoA (especially adults, men, and the patients without a VSD) have a higher risk of developing hypertension, therefore increasing their risk for CVA [87].

Endocarditis prophylaxis is not required for patients with uncomplicated native coarctation or 6 months after successful repair of native or re-coarctation. Antibiotic prophylaxis is

indicated in patients with a past history of endocarditis, in those whose repair involved insertion of a conduit, or for 6 months after intervention if prosthetic material or stent was used. The 2015 scientific statement of the AHA/ACC provides competitive athletic participation guidelines for patients with congenital heart disease (CHD), including coarctation [88]. As with any other guidelines, recommendations need to be tailored to the patient and a comprehensive evaluation by an experienced clinician is required. Before a decision is made regarding sports participation, a detailed evaluation should be conducted, which should include a physical examination, electrocardiography (ECG), chest radiograph, exercise testing, and cardiac/aortic imaging (with transthoracic echocardiogram, MRI, and/or computed tomography angiography [CTA]) when appropriate. The time interval for repeating this extensive testing is unclear and should be individualized to the specific patient.

It is known that morbidity and mortality are higher in patients with CoA given their risk of complications. These patients can have aortic aneurysms, chronic hypertension, re-coarctation, and the potential need for additional transcatheter and surgical interventions. However, for patients with coarctation of the aorta that survive into adulthood, studies have shown that their overall long-term survival rate is high. These patients should be followed up in a center specialized in adult congenital heart disease, where these morbidities are recognized and close observation is provided to prevent devastating complications.

11. Conclusion

Patients with CoA who have undergone repair require lifelong surveillance. Because this type of CHD is associated with many long-term complications, collaborative management by cardiologists with expertise in adult CHD is recommended. As patients with CHD are now surviving into adulthood, with 5–8% of these patients having coarctation of the aorta, it is important to understand the anatomy, pathophysiology, and management of these patients. Although echocardiography is a fundamental tool in the assessment of patients after coarctation repair, advanced imaging is often necessary for comprehensive evaluation. cMRI is the preferred imaging modality for repaired and unrepaired CoA. Alternatively, cardiovascular CT is best suited to evaluate patients with endovascular stents or those with contraindications to cMRI. It is not uncommon for this cohort to develop complications or require additional percutaneous or surgical interventions during their lifetime. This chapter emphasizes the importance of long-term follow up care, especially in a center specializing in the care of patients with congenital heart disease.

Conflict of interest

There is no conflict of interest.

Author details

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