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Review Article

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Management of Coarctation of The Aorta in Adult Patients: State of The Art

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ABSTRACT

Coarctation of the aorta (CoA) is a common form of congenital heart disease. Adult patients with CoA may be asymptomatic or may present with hypertension. Over the last few years, endovascular management of adult patients with CoA emerged as the preferred strategy. Stent implantation, though technically challenging, offers the best and most lasting therapy. In this paper, we will review technical considerations and outcome of patients undergoing stent implantation for CoA.

Keywords: Stents; Aortic Coarctation; Balloon angioplasty; Cardiac Catheterization

INTRODUCTION

Coarctation of the aorta (CoA) is the sixth most common congenital heart disease (CHD) accounting for 4–8% of all CHD and occurs in 4 out of 1,000 live births with a male predominance. CoA can occur as an isolated lesion, but is often associated with other cardiovascular lesions, such as bicuspid aortic valve (BAV) in 50–75% of the cases, aortic arch hypoplasia, subaortic stenosis, mitral valve abnormalities, ventricular and atrial septal defects and patent ductus arteriosus (PDA).[1)2)] The most important non-cardiac associated lesion is cerebral aneurysm present in up to 10% of patients, which is approximately 5 times higher than that in the general population.[3)4)]

CoA is defined as a localised narrowing of the aortic lumen by a ridge, composed of medial wall thickening and infolding of aortic wall tissue. The narrowing is commonly located opposite to the insertion of the PDA (juxtaductal); however, it may also be located proximal (preductal) or distal (postductal) to the PDA. Infrequently, it can also exist in the transverse aortic arch and abdominal aorta, or be a part of a long segment arch hypoplasia.

PATHOGENESIS

The underlying pathogenesis of CoA is not fully understood. However, there are three theories that may shed some light on this[5)6)]:

1. Abnormal genetic mutation.
2. Distal aortic arch underdevelopment due to reduced anterograde intrauterine blood flow leading to underdevelopment of the fetal aortic arch.
3. Aberrant PDA tissue extrusion into the wall of the fetal thoracic aorta.

Most cases of CoA are due to sporadic mutations. Several genes have been implicated in CoA etiology, including the *NOTCH1* gene, which plays an important role in cardiovascular development. A higher incidence of CoA is seen in children with Turner's, Noonan's, Williams-Beuren syndromes, PHACE, DiGeorge, and velocardiofacial syndromes. Children with Turner's in particular are at higher risk of aortic dissection than the general population and should be followed closely.[7] An association has been described with sympathomimetic medication use by the mother in the Baltimore Washington Infant Study.[8]

CoA is also considered a general arteriopathy, given the often-abnormal histology of the arterial wall adjacent to and distant to the site of CoA. In particular medial wall degeneration has been observed in pre- and post-stenotic specimens and this may lead to increased incidence of aortic aneurysm and dissection. However, some argue that these changes occur as results of increased blood pressure and the hemodynamic disturbance due to the narrowing.[9]

NATURAL HISTORY

Without treatment, the outcome for patients with CoA is poor. Historical data on the natural history of patients with CoA who survived beyond infancy showed a mean age of death of 34 years and 75% mortality by age 43 years. Death was from congestive heart failure, aortic dissection or rupture, endocarditis, and intracranial bleeding.[10]

PATHOPHYSIOLOGY

The presence of narrowing increases the left ventricular (LV) pressure afterload, exposes the upper body to hypertension, causes flow disturbance in the thoracic aorta, and decreases perfusion to the lower body. As a compensatory mechanism, the LV hypertrophies and collateral vessels will develop to overcome the obstruction. In addition, also pre and post stenotic dilatation develops.[11] Depending on the balance between the degree of narrowing and the compensatory mechanisms available to overcome it, the clinical presentation may vary from the critically ill neonate with heart failure to the asymptomatic child or adult. The possible presence of intrinsic aortopathy and hypertension will increase the risk of aortic aneurysm/dissection and cerebral berry aneurysm formation.[12] As result of flow disturbance and/or associated CHD especially BAV, the risk of endocarditis will increase. Furthermore, re-CoA and future aneurysm formation can occur following successful surgical and endovascular repair which mandates life-long surveillance.

CLINICAL PRESENTATION

Two populations of adults with CoA exist: those, who underwent previous CoA repair in childhood and those with native (un-corrected CoA). It is uncommon to be diagnosed incidentally for the first time during routine medical check-up. The usual presentation in

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adults is hypertension. Liberthson et al.[13] reported that 10.3% of patients presented with CoA after the age of 40 years. Occasionally, patients present with leg fatigue and claudication and abdominal angina. Other typical symptoms in adults with untreated CoA may include headache, tinnitus, epistaxis, dizziness, cold feet and in advance cases of angina and shortness of breath. On clinical examination, the femoral pulses are often weak, delayed or absent. A continuous systolic-diastolic murmur between the scapulae is typical for significant CoA with blood flow through collateral vessels. Auscultation may also reveal a suprasternal thrill and a systolic ejection murmur in aortic area in cases with concomitant aortic valve disease. Complications of CoA may include premature coronary artery disease, heart failure, arrhythmias, aortic aneurysm, aortic rupture/dissection, infective endocarditis, and intracranial aneurysm/haemorrhage.

Interestingly, a recent analysis of 80 adolescents who had had CoA repair in childhood did not find any intracranial aneurysms on screening MRA,[3] raising the possibility that the aneurysms develop with age and preferentially in the presence of hypertension.

EVALUATION

The electrocardiography (ECG) may show signs of LV hypertrophy and dilatation, and also it may show ischemic changes (**Figure 1**).

IMAGING

The primary goals of imaging modalities are to establish the diagnosis, preoperative or procedural guidance, or post repair surveillance.

Chest radiography

Chest X-ray in adults may show a normal cardiac contour. Double contouring of the descending aorta known as the "3 sign" beneath the aortic notch is characteristic and represents narrowing of the aorta at the level of CoA and dilatation of the aorta pre and post

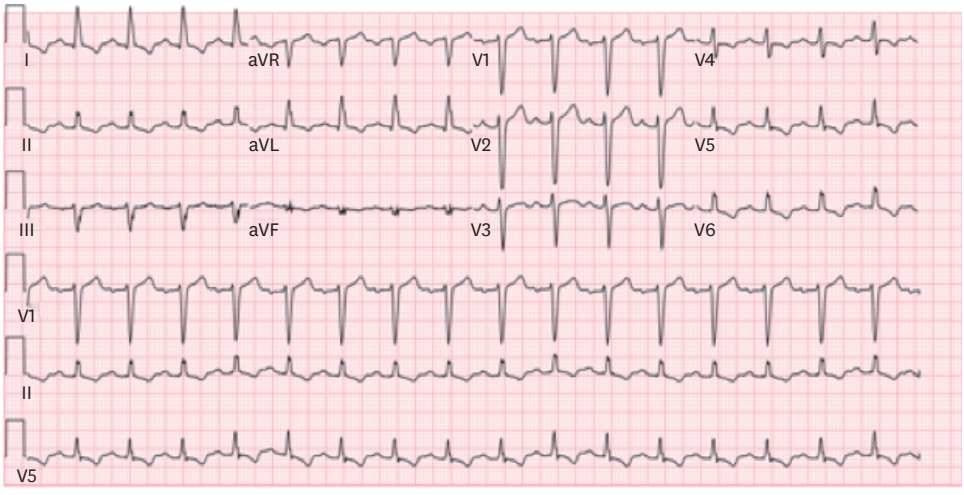

Figure 1. An electrocardiogram of a 45-year old male with coarctation of the aorta. There is evidence of ischemic changes.

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CoA. Rib notching of the posterior fourth to eight ribs caused by the intercostal collateral arteries can be visible in patients with longstanding severe CoA (**Figure 2**).

Echocardiography

Transthoracic echocardiography (TTE) is the primary imaging modality for suspected CoA, given its ready availability and safety. It has the capacity to provide us with diagnosis and to assess severity. Continuous wave Doppler can reveal the peak pressure gradient (PPG) across the CoA and continuation of the flow during diastole (**Figure 3**). Appropriate visualisation of the CoA-site can, however, be difficult due to a poor acoustic window and operator dependence. In addition, when the aorta leaves the suprasternal echocardiographic plane, it normally has a tapering appearance, thereby confounding the assessment of severity. TTE can assess cardiac function and associated cardiac and valvular abnormalities. TTE has limited value in the evaluation of extracardiac structures and collateral circulation. Although transesophageal echocardiography can provide accurate imaging of the aorta, it is seldom used due to its invasive nature and limited additional value.

Cardiac magnetic resonance imaging

Cardiac magnetic resonance (CMR) imaging is the preferred advanced imaging modality for non-invasive diagnosis and follow-up of CoA. A major advantage of CMR is the lack of ionising radiation and contrast exposure, making it ideally suitable for repeated imaging. CMR can accurately identify the location and hemodynamic (phase contrast flow analysis) significance of the CoA, determine arch sidedness, branching patterns, and the length of segment involved (**Figure 4**). CMR also enable selection of ideal stent size and length. Assessment of the cardiac function and the presence of other CHDs can be decided accurately. In addition, it is very helpful for surveillance after stent implantation and surgery for complication.

Computed tomography

Although CMR is a core imaging modality in patients with CoA, cardiac computed tomography (CT) (computed tomography angiography; CTA) has also become a very important tool in the diagnosis and management of such patients. It offers several advantages

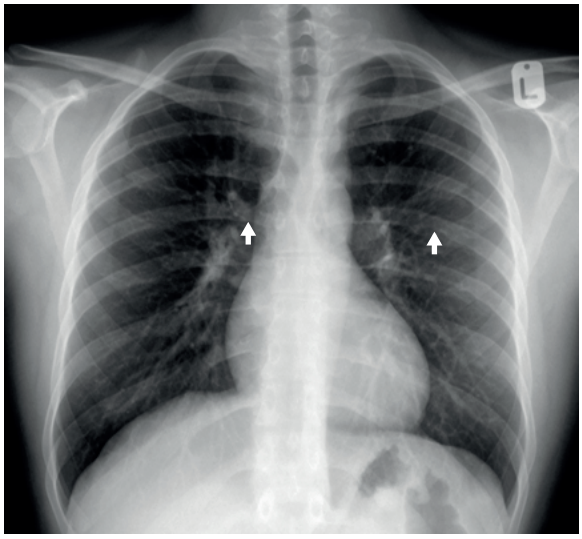

Figure 2. Chest radiograph of a patient with long-standing coarctation of the aorta demonstrating typical findings of coarctation: rib notching (arrows) and '3 sign' beneath the aortic notch.

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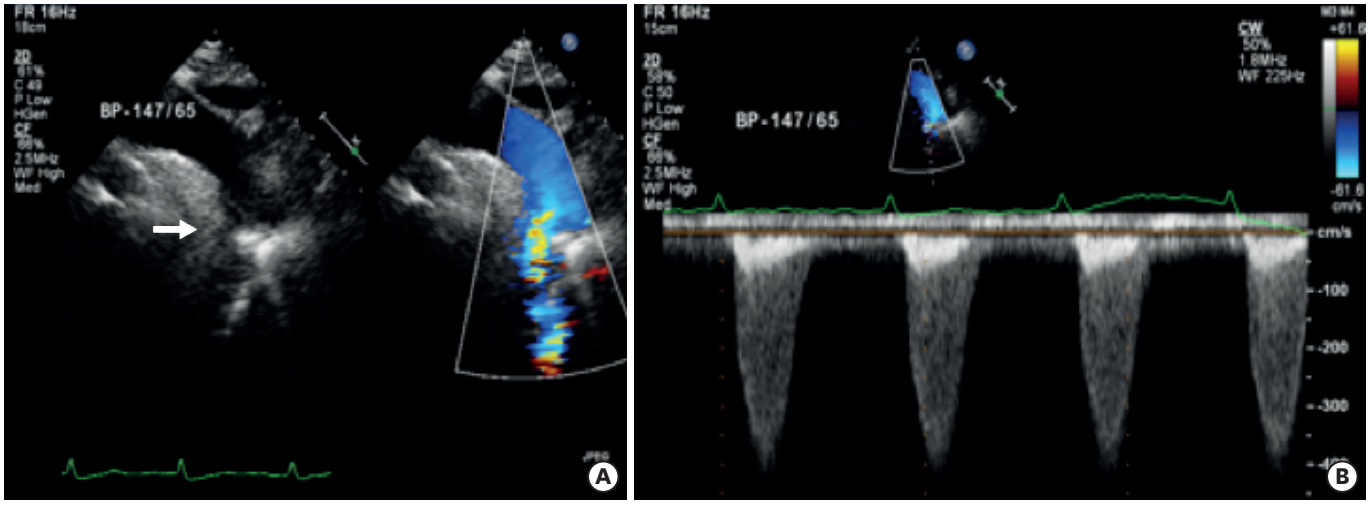

Figure 3. (A) A two-dimensional TTE from the suprasternal view demonstrating a discrete narrowing just distal to the take-off of the left subclavian artery (arrow) with continuous high velocity color Doppler signal across the coarctation. (B) Continuous wave Doppler TTE showing continuation of anterograde flow during diastole and peak pressure gradient of 60 mmHg across the coarctation.
TTE = transthoracic echocardiography.

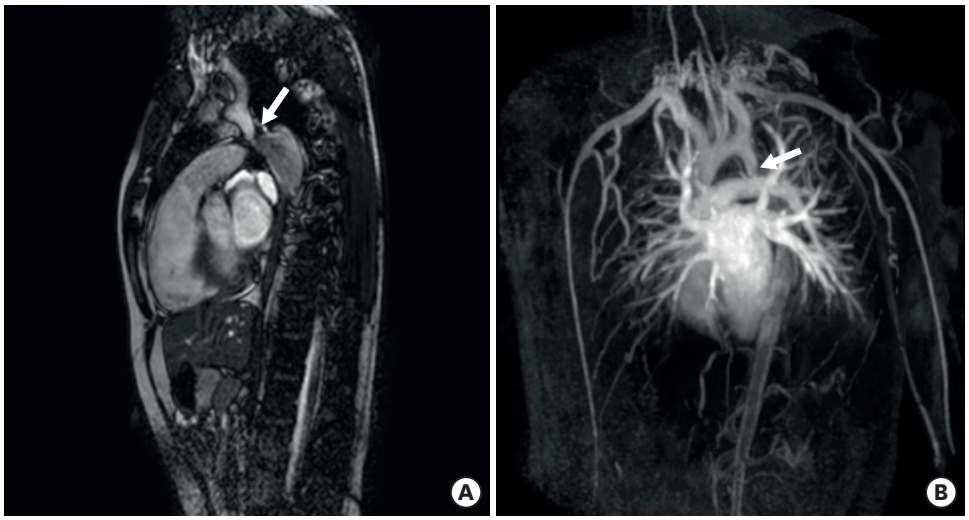

Figure 4. (A) Cardiac magnetic resonance image in sagittal view revealing discrete coarctation of the aorta (arrow). (B) Different view is cardiac magnetic resonance image revealing extensive collaterals.

over CMR: high spatial resolution, shorter acquisition times, presence of metallic implants is not a contraindication, and presence of previous stent implantation does not lead to a signal noise artefact. It is less claustrophobic compared to CMR. The radiation and contrast are a concern in this modality but the new low dose protocols of radiation and contrast can give the same excellent image (**Figure 5**). However, CT cannot provide haemodynamic information such as the PPG across the CoA-site and the degree of collateral circulation.

Catheter angiography

Catheter angiography (CA) is the gold standard for the assessment of the PPG across the CoA and provides high-resolution images of the aorta. With the emergence of CMR and CTA, CA is currently limited for coronary angiography before intervention/surgery and when catheter-based intervention is considered.

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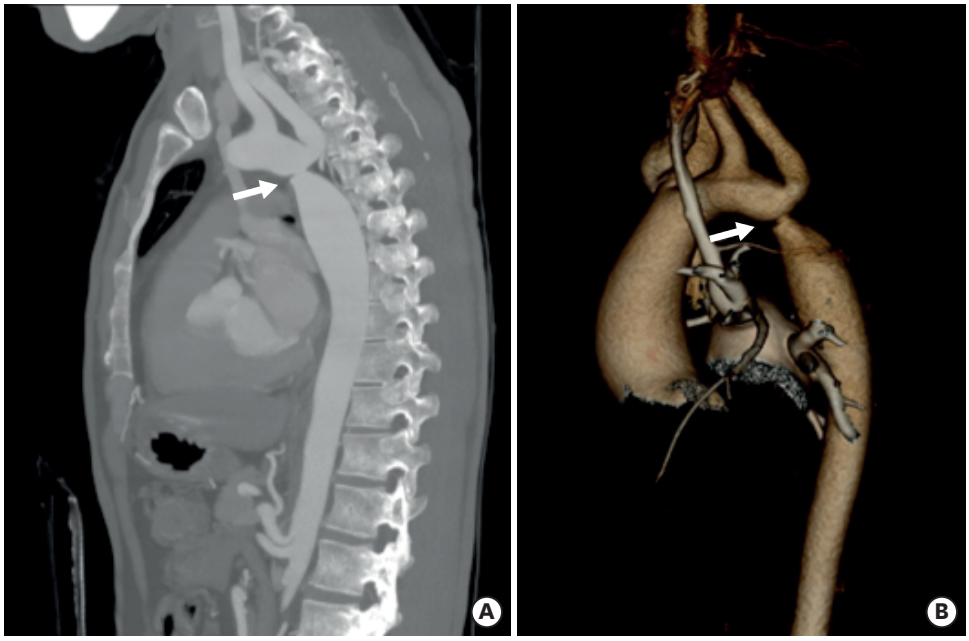

Figure 5. (A) Computed tomographic angiogram is revealing coarctation of the aorta just at the origin of the left subclavian artery (arrow). (B) 3D volume rendered reconstruction of the same patient revealing the coarctation (arrow), note the incidental finding of persistent left superior vena cava to left atrium.

INDICATIONS FOR INTERVENTION

According to the most recent recommendations for the management of CoA as issued by the European Society of Cardiology and the American heart association/American College of Cardiology,[14][15] interventional therapy is indicated in all patients with a non-invasive blood pressure difference >20 mmHg between upper and lower limbs, regardless of symptoms but with upper limb hypertension (>140/90 mmHg in adults), pathological blood pressure response during exercise, or significant LV hypertrophy (class I, level C). Independent of the PPG, hypertensive patients with ≥50% aortic narrowing relative to the aortic diameter at the level of the diaphragm should also be considered for treatment (class IIa, level C).

Milder obstructions may also benefit from intervention by decreasing LV diastolic pressure and preserving LV function in the long term, especially in the presence of hypertension at rest, abnormal blood pressure response during exercise, progressive LV hypertrophy, elevated LV end diastolic pressure and in cases of complex heart disease particularly those post Fontan operation.[16]

TREATMENT OPTIONS

The treatment of CoA could be either surgical or catheter based.

Surgical management

In order to evaluate individual treatment options in adulthood one needs to have a detailed understanding of the possible operations performed in infancy or childhood. Several surgical

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techniques have been described.[17] The surgical approach of choice is based on the age and aortic arch anatomy. The types of surgical repair include:

1. Resection with end-to-end anastomosis. The first surgical therapy documented for CoA was performed through a lateral thoracotomy and involved resection of the coarcted segment followed by a direct suture anastomosis of the transected ends. High re-CoA rates complicated this approach due to the circumferential suture lines particularly when performed in neonates.
2. Patch aortoplasty. To minimize the concerns of re-CoA, patch augmentation, initially with Dacron, was used where the aorta was incised longitudinally through the coarcted segment on the lateral wall of the aorta and a prosthetic patch is sutured across the incision which enlarges the vessel diameter. This technique initially resulted in less frequent re-CoA compared to the end-to-end repair but it fell out of avour when aneurysms formed in 20–40% of cases on the opposite wall of the patch augmentation. Changing the patch material to polytetrafluoroethylene (PTFE) reduced the aneurysm formation to about 7% but still demonstrated a 25% risk of re-CoA. Patch aortoplasty is still being used but only in the context of complex arch reconstruction.[18]
3. Subclavian flap aortoplasty. In this technique the subclavian artery (SCA) is ligated close to the origin of the left vertebral artery. The flap is generated by an incision of the SCA that is extended down onto the aortic isthmus and across the coarcted segment. The re-CoA rate of this technique seems to be relatively low when performed in older children up to 3%. However, when applied to neonates, re-CoA may occur in up to 23%.[19] Although sacrificing the SCA does not result in left arm ischemia, it may cause claudication of the affected arm in the long term.
4. Extended end-to-end anastomosis. In contrast to a direct end-to-end anastomosis, the proximal clamp is placed across the aortic arch including the SCA or even the left carotid artery including the aortic arch. Distally, the aorta is clamped below the coarcted segment. After ligation and division of the ductus arteriosus, the coarcted segment is resected and the aortic arch is opened on its inferior aspect, followed by end-to-end anastomosis of the opened arch and the descending aorta. The procedure can be performed with low peri-operative mortality and reports show relatively low re-CoA rates of 4% to 13%.[20]
5. Interposition graft. This technique has been reserved for patients in whom outgrowth of the graft is not a concern, or in patients with long-segment CoA. After the aorta is cross-clamped and the obstructive tissue resected, a tube graft of either aortic homograft or Dacron is sewn into the aorta, creating an unobstructed path for blood. The main disadvantage of this technique is that it requires a longer cross-clamp time for two surgical anastomoses to be sewn, and the tube graft will not grow with the patient. Yet, for adult-sized patients presenting with long-segment CoA, this technique may be preferable at many centers.

Overall, surgery is performed as early as possible balancing the prognostic benefit of early surgery versus the age-related risks for procedural complications. Surgical mortality in CoA-intervention is rare (<1%), with an overall survival rate of 98% at a median follow-up of 4.8 years of age for infants and 76% one year after initial repair for preterm infants.[21][22] Early postoperative morbidity includes paradoxical hypertension, injury to the recurrent laryngeal nerve (or other adjacent nerves) and subclavian steal syndrome (with subclavian patch angioplasty). Although very rare, spinal cord ischaemia with paraplegia (due to prolonged

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clamping of the aorta) has been reported.[23] Surgical repair in adulthood is characterised by an increased mortality risk due to degenerative changes in aortic wall, coronary artery disease and end-organ damage due to long-standing hypertension.

Catheter based repairs

Transcatheter procedures have a role in native CoA as well in the treatment of re-CoA or aneurysm formation after initial repair.

Balloon angioplasty

Balloon angioplasty (BA) for CoA was first reported in 1982,[24] with use having become widespread over the past two decades. The mechanism of BA involves controlled tearing of the intimal and medial walls of the aorta and there is concern that this damage to the aortic wall predisposes to later aneurysm formation especially when treating native CoA with this technique. A relatively high incidence of re-stenosis is also reported for BA alone.[25] In order to overcome these concerns, stenting of CoA was introduced in 1991.[26]

Stenting

Stenting is considered advantageous when compared to BA alone; it improves luminal diameter, results in minimal residual PPG, and sustained hemodynamic benefit. It also can tack intimal flaps to the aortic wall potentially allowing healing to occur this way reducing the risk of dissection and aneurysm formation. Furthermore, stent implantation does prevent vascular recoil resulting in re-CoA.[27] Nowadays, BA with simultaneous stenting is considered to be the preferred treatment option for adolescent and adult patients with native or re-CoA. Stent implantation in young children remains controversial due to the need for frequent re-dilation to accommodate the growing aorta. In our practice, we consider stent placement only in patients who are large enough (usually over 15 kg in weight) to receive a stent that can be expanded to an adult size. However very few stents are expandable to the average diameter of a large adult aorta (21.1+3.2 mm for women, 26.1+4.3 mm for men).

Stent technology has evolved in recent years and there is now a wide variety of stents available to treat CoA which all vary in the degree of flexibility, profile, foreshortening on expansion, maximal expansion diameter, and radial strength. The choice of stent depends on the CoA anatomy, size of the patient, the preference of the operator and availability.

1) Stent types

The most commonly implanted type of stent, and the only one with Food and Drug Administration and CE mark approval for this use, is the Cheatham Platinum (CP) stent (NuMED Inc., Hopkinton, NY, USA). Other stent types are used off-label. They include Palmaz series (Johnson & Johnson Interventional Systems Co., Warren, NJ, USA), Genesis XD (Cordis Corp., Miami, FL, USA), Atrium Advanta V12 (Maquet, Rastatt, Germany), IntraStent (Medtronic, Minneapolis, MN, USA), Formula (Cook Medical, Bloomington, IN, USA), and AndraStent (Andramed GmbH, Reutlingen, Germany). In small children and in bail-out situations, coronary stents have been used as well, because of the small diameter of the aorta. Stents vary both in material and architecture. There are 3 commonly used designs, namely closed-cell, open-cell, and a hybrid design. In closed-cell design all internal inflection points of the structural components are connected (fixed points), thereby creating small closed cells. Therefore, it is less flexible and conformable compared with stents with an open-cell design. In the open-cell designs some of the internal inflection points of the structural components are not connected. The absence of connections allows more

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longitudinal flexibility of the cells preferred for treatment of transverse aortic arch narrowing, because they adapt better to the arch shape. Furthermore, separate BA through the cell into the aortic branches can be performed when using an open-cell design. Stents with hybrid design, closed-cell and open-cell designs are combined. Another architectural difference is the use of single wires welded together (for instance in the case of the CP stent) versus a single tube that is slotted without junctions between components. Welds represent weak points in the structure of the stent; thus, in the newer CP stents, gold soldering was added to reinforce the welds. One of the other important differences between the stents is the metal made of, namely, stainless steel, platinum-iridium alloy, and chromium-cobalt alloy. The chromium-cobalt alloy is stronger than stainless steel. Stents can be bare or covered. The most commonly used material for the cover is PTFE, which makes the stent impermeable. Conventionally, covered stents were solely used to treat aortic wall complications. However, currently, covered stents are implanted as the primary treatment of CoA in patients at risk of complications. Risk factors for lesions to the aortic wall include narrow CoA, tortuous aorta, genetic aortic pathologies, and aneurysms/pseudoaneurysms derived from previous interventions. The characteristics of each type of stent are summarized in **Table 1**.

2) Procedural steps

At our institution, we elected to perform all procedures that involve BA or stent implantation for CoA under general anesthesia. The procedure incites pain at the time of dilatation and this may result in patient movement which may compromise the success of the procedure. Trans-femoral artery (FA) approach is the standard access, if crossing of the CoA segment is impossible from the descending aorta, a secondary arterial access via the radial or brachial artery allows for an antegrade catheterization of the CoA. Subsequently, the wire has to be exteriorized through the FA sheath. Some operators may prefer the second access from a transseptal puncture. We also obtain venous access to perform full hemodynamic study and for the possibility of rapid right ventricular pacing during stent deployment. Intravenous heparin is administered achieving an activated clotting time (ACT) greater than 200 seconds. A straight-tip catheter (multipurpose) or a right judkins catheter is then advanced into the descending aorta followed by advancing a soft-tipped wire (e.g., Glide wire) through the CoA site. Once the catheter is advanced into the ascending aorta, it is replaced over the wire by a pigtail catheter or multitrack catheter followed by careful pullback to obtain PPG across the CoA. Biplane angiography is then performed using a calibrated pigtail catheter positioned just proximal to the CoA. Good angiographic visualization of the aortic arch in a lateral and shallow left or right anterior oblique projection view is essential. The angiogram needs to show the entire transverse aortic arch, including brachiocephalic vessel location, isthmus, CoA segment, and the aorta down to the level of the diaphragm. Following angiography, measurements are made at different points of the aorta in

Table 1. Characteristics of the different types of stents

Stent types	Manufacture	Metal	Cell types	Cover	Advantages	Disadvantages
CP	NuMED Inc.	Platinum-Iridium	Closed cell	Outer cover PTFE	Good radial strength, rounded edges, good radio-opacity	Large delivery sheath for covered stents
Atrium Advanta V12	Maquet	Stainless steel	Open cell	Outer cover PTFE		
AndraStent	Andramed GmbH	Cobalt chromium	Hybrid (open and closed)		Minimal foreshortening, good flexibility	
IncraStent	EV3 Inc.	Stainless steel	Open cell			
Formula	Cook Medical	Stainless steel	Open cell			
Palmaz 8	Cordis Corp.	Stainless steel	Closed cell		Good radial strength	Sharp edges stiff
Palmaz XL	Cordis Corp.	Stainless steel	Closed cell		Good radial strength	Sharp edges stiff
Genesis XD	Cordis Corp.	Stainless steel	Closed cell		Broader expansion range, good radial strength, good conformability	

CP = Cheatham Platinum; PTFE = polytetrafluoroethylene.

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order to select the size of balloon or stent to be used. The decision on the maximal diameter of the balloon on which the stent is mounted is based on the diameter of the transverse or distal arch and isthmus with the diameter not exceeding the size of aorta at the level of the diaphragm. It is important to note that post-stenotic dilatation is common and this should not be used as reference for balloon size selection. A soft tipped wire is initially used to cross the CoA. Thereafter, it is exchanged for a stiffer wire (usually an Amplatz super or extra stiff exchange guidewire) which is secured in the ascending aorta or the right SCA. A long Mullins sheath (at least 75-cm) (Cook Medical) is advanced from the FA site and placed across the CoA segment for deployment of the stent. The stent is hand-crimped on the balloon catheter. We use an umbilical tape to finish crimping on to the balloon if available. A guidewire is inserted into the distal end of the balloon catheter while crimping the stent onto the balloon in order to avoid compromising the lumen of the catheter. Variety of balloons such as the BIB and Z-Med (NuMEM Corp., Hopkinton, NY, USA), and Cristal (Balt, Montmorency, France) balloons are used. The BIB balloon catheter is the most commonly used balloon in our practice. This balloon has an inner balloon and a 1-cm longer outer balloon which is double the diameter of the inner balloon. They are available in outer balloon diameters of 12 to 30 mm, requiring sheaths from 8 to 16 Fr. Because the inner balloon is usually shorter than the stent, the balloon expands the stent uniformly without flaring the ends of the stent, thus decreasing the risk of balloon perforation or wall injury. The expanded inner balloon provides a better anchoring mechanism, which results in more precise control during inflation of the outer balloon. As such it provides controlled stent expansion and the ability to adjust stent position following inner balloon expansion. The single large diameter balloon catheters can be used, but as these balloons first expand at the ends, they carry the risk of wall injury by the edges of the stent especially in small anatomy. The already mounted bare and mounted covered CP Stents are available. The stent is pre-mounted on a BIB balloon catheter. This already pre-mounted system will save time and reduces the risk of dislodgement. It is ready to be introduced through the delivery sheath over the stiff wire. Recently introduced is the NuDEL™ (NuMED Inc.) which is all-in-one stent delivery system designed for the efficient and effective treatment of CoA. It includes the covered CP stent, mounted on a BIB balloon catheter, which is then covered by a sheath (stent, balloon, sheath: all-in-one system). Pre-loaded system saves time and allows for quick actions in emergency situations.

The stent/balloon assembly is advanced through the Mullins sheath over the stiff wire and the stent is positioned across the CoA site before the sheath is withdrawn gently to uncover the stent/balloon completely. Prior to full uncovering, repeated injections can be made from the other access line or via the side-arm of the delivery sheath. Once position of the stent is confirmed to be at the right location, expansion of the stent follows. If the coarcted segment is very tight, no need for right ventricular pacing. However, if the CoA is mild or the segment is in the transverse arch, we recommend rapid RV pacing to ensure no stent movement. We usually like to see mean aortic pressure about 50 mmHg during pacing while expanding the stent. Pacing is turned off after the outer balloon is fully deflated. The various steps of CoA stenting are demonstrated in **Figure 6**.

We advise against pre-stent BA unless the CoA is very tight to allow the delivery sheath passage as this approach is associated with more complications than primary direct stenting.[28] If a BIB balloon catheter is used, the inner balloon is inflated first and the stent repositioned, if needed, before inflation of the outer diameter. Both balloons are then deflated and the balloon catheter is removed over the wire while keeping the sheath in position. Following stent deployment, another PPG across the CoA is documented with

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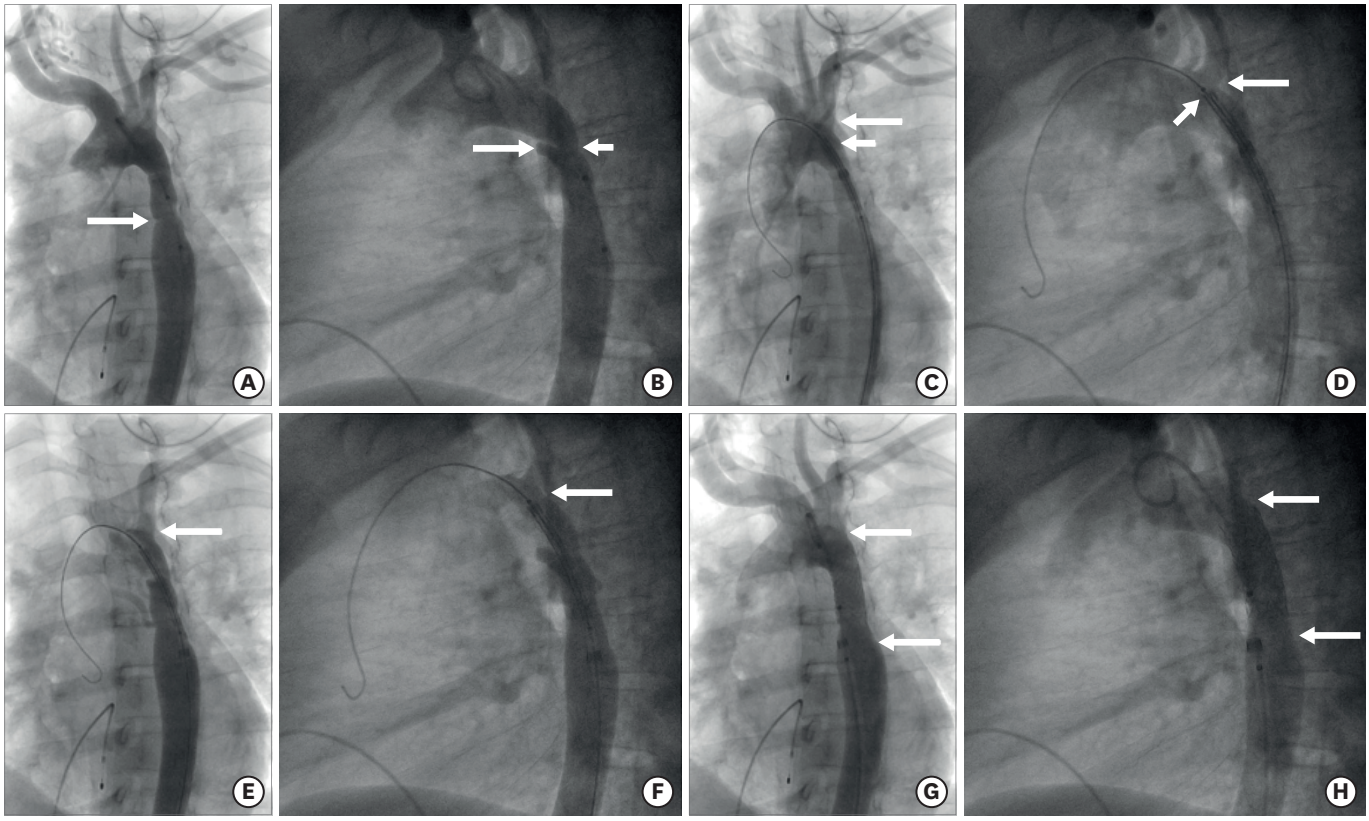

Figure 6. Angiogram in the transverse arch in left anterior oblique 30° (A) and straight lateral 90° (B) in a 28 years female patient with severe coarctation of the aorta, with a gradient of 30 mmHg. (A) The coarctation ridge (arrow). (B) Ductal diverticulum (long arrow) and the coarctation juxta ductal (shorter arrow). Note, there is a pacing catheter in the right ventricle. (C, D) Angiogram via side arm of delivery sheath (covered stent [Bentley 14 mm×39 mm] is uncovered half way). (C) Origin of the left subclavian artery (long arrow) and proximal tip of the stent (short arrow). (E, F) Angiogram via side arm of sheath after uncovering the stent completely. Note, origin of the left subclavian artery (arrow). (G, H) Angiogram via pigtail catheter in transverse arch after complete stent deployment showing good position (arrows) and remodelling of the area of coarctation. Residual gradient was 0 mmHg.

pullback or simultaneous pressure measurement using the catheter above the CoA and the sheath positioned below the CoA. A final angiogram is performed to assess the result and to look for any complication (aortic wall injury). Stent implantation is considered successful if a PPG <10 mmHg and improvement in vessel calibre >80% of the normal adjacent aortic arch is achieved.[29] Post-dilatation of the stent with a larger balloon can be performed if CoA relief or stent apposition to the aortic wall is deemed suboptimal. However, we recommend against full expansion of the stent at the site of CoA. In adults with tight CoA, we are satisfied with a diameter of 12–14 mm at the site. Above and below can be gently expanded making sure not to expand the narrow area to that diameter. Some operators, specially using covered stents they try to achieve full expansion to the diameter of isthmus. At the end of the procedure we remove the sheath manually (ACT <160 seconds). A perclose proglide suture mediated closure system (Abbot Vascular, Abbott Park, IL, USA) has been used to close the access site. However, it needs to be inserted in the common FA prior to the introduction of the large sheath and the diameter of the vessel to be at least 5–6 mm.

Procedure related complications are not common, and these may include: vascular access related complications and aortic wall injury. Aortic dissection and aortic rupture may occur in 0.9% and 0.4%, respectively. Stent migration/embolization may occur in 2.4% of the cases. Occlusion of aortic side branches and stroke may occur, and death is uncommon (0.4%).[30]

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Patients are usually discharged home next day on aspirin 81 mg daily and are instructed to avoid contact sports for 6 months. A chest radiograph, ECG, TTE are obtained prior to discharge.

Outcome

Success

Since the first cases of CoA stenting were reported in the 1990s, the encouraging outcome of this procedure has been described in considerable numbers of individual series reporting both short-term and intermediate-term results.[31-34] In 2007, a large multi-center retrospective series was reported by the Congenital Cardiovascular Interventional Study Consortium (CCISC) on 565 procedures of stent implantation performed in 555 patients with CoA between 1989 and 2005.[28] The success rate of reducing PPG to less than 20 mmHg was 98%. There was a mean±standard deviation (SD) reduction of the PPG from 31.6±16 to 2.7±4.2 mmHg and an increase in diameter from 7.4±3.0 to 14.3±3.2 mm. These results showed high degree of success rates which were similar to those reported in other smaller series.[31-34] These series and the CCISC in a follow-up study reported that the relief of the PPG persisted in the majority of patients at medium-term follow-up.[31] Recently, Hartman et al.[30] reviewed reports published between 1990 and 2014 after stenting CoA involving 1,612 patients with some reports included in this review had long follow-up reaching up to 45 months.[30] The CoA diameter increased from 6.43±0.86 mm to 15.1±0.6 mm, PPG deceased from 38.58±3.66 mmHg to 3.93±1.3 mmHg, and systolic blood pressure from 153±9.3 mmHg to 132±10.8 mmHg. In another recent meta-analysis by Yang et al.[35] for the reports published between 1991 and 2015 including 561 CoA patients after stenting. The procedures success rate was 98%, the CoA diameter increased from 5.1± 5.4 mm to 14.4±7.7 mm, PPG deceased from 47.4±13.4 mmHg to 3.6±3.6 mmHg (all CoA diameter and PPG values are presented as mean±SD.).

Long term complication

Recently, the results of the Coarctation of the Aorta Stent Trial (COAST) trial, a study that aimed to assess the safety and efficacy of CP stents in children and adults with native or re-CoA were published.[36] During the two-year follow-up no deaths, serious adverse events or surgical interventions were reported. All patients experienced satisfactory post procedural results with low rates of complications including aneurysms (5.7%) and stent fracture (11% at 2 year) without loss of stent integrity, stent embolization, aortic wall injury, or re-obstruction. Re-interventions (8.7%) that occurred during the two-year follow-up period were for stent re-dilatation in order to account for the patients' somatic growth and address of aneurysms. Of note, there was evidence of fracture progression and additional reinterventions occurred after 2 years. In the review by Hartman et al.[30] the rate of aneurysms was 1.5%, stent fracture 1.6%, and reintervention 11%. Relatively similar findings were obtained by Yang et al.[35] in his meta-analysis.

Covered stent versus bare stent

It has been proposed that the use of covered stents reduces the risk of aneurysms, dissection, and rupture. The recently published, COAST II trial which is a multi-center, single arm trial using the covered CP stent for the treatment and/or prevention of acute wall injury (AWI) in native or re-CoA associated with one or more of the following: acute or chronic aortic wall injury, nearly atretic descending aorta to 3 mm or less in diameter, genetic syndromes associated with aortic wall weakening (e.g., Marfan syndrome, Turner syndrome, familial BAV with ascending aortic aneurysm), or advanced age (60 years or older). A total of 158 patients were involved. The PPG decreased from 27 mmHg to 4 mmHg. The overall success rate was 92%. There was no AWI, repeat intervention or death.[37] However, in a randomized trial of 120 patients with severe native CoA, there was no difference in the rate of re-CoA

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and pseudoaneurysm formation after 31 months of follow-up between patients who underwent implantation using a bare metal stent and those with a covered stent and this result was similar to what previously has been published in case reports and small series.[38] Nevertheless, covered stents offer the advantage of excluding any stretch-induced wall trauma from the endoluminal aspect of the aorta, particularly in the catastrophic event of aortic rupture as revealed by COAST II trial.

Surgery versus transcatheter intervention

Forbes et al.[39] published the results of a multi-center observational study comparing the safety and efficacy of surgical repair (72 patients), stent (217 patients), and BA (67 patients) as treatment of native CoA acutely and at follow-up. All 3 treatment modalities showed significant improvement in systolic blood pressure and PPG. The rate of acute complications was lower after stent implantation compared to BA or surgery. However, planned re-intervention was more likely in the group of stented patients. Stent implantation and surgery achieved superior hemodynamic results than BA. This finding was relatively consistent with the previously reported results by Carr et al.[40]

Follow-up

Despite excellent long-term survival after CoA repair, patients have a reduced life expectancy with increased risk of morbidity. Long-term complications occur despite adequate and timely repair, warranting life-long and careful follow-up. In our practice, we schedule the clinical visits at 3, 6, and 12 months for the first year, then once every year. A blood pressure evaluation (four limbs), ECG, CXR, and TTE are obtained at each visit. Due to the possibility of late complication such as aneurysm formation and re-CoA, follow-up CTA/CMR is mandatory in all patients at approximately 6 to 12 months after the procedure, then we repeat at intervals of 5 years or less, depending on the specific anatomic findings before and after repair and at any time before pregnancy.

Hypertension

Chronic hypertension remains present in 35–68% of the patients with CoA, even in the presence of an anatomically satisfactory repair.[41-43] Furthermore, exercise-induced hypertension occurs in over one-third of the normotensive patients.[44] Prevalence of systemic hypertension is significantly lower in patients treated in the neonatal period or infancy and in patients who underwent CoA resection with end-to-end anastomosis.[42] The exact pathophysiology of late-onset hypertension after CoA-repair remains to be elucidated. Reduced aortic compliance and an abnormal baroreceptor function may explain this.[41] Hypertension is also associated with an abnormal geometry of the aortic arch.[42] Mild residual morphological obstruction and a disturbed renin–angiotensin–aldosterone system appear to play no major role in late-onset hypertension.[45] Post-treatment hypertension is a risk factor for premature death and requires aggressive treatment. Follow-up should include ambulatory 24 hours blood pressure measurement and exercise testing, as exercise-induced hypertension can predict future systemic hypertension.[41]

Exercise/sports

Patients without residual obstruction who are normotensive at rest and with exercise can usually lead normally active lives without restriction, except for extensive static sports at a competition level. Patients with arterial hypertension, residual obstruction, or other complications should avoid heavy isometric exercises, in proportion to the severity of their problem.[14][15]

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Pregnancy

After successful treatment of CoA, many women tolerate pregnancy without major problems. In particular, women with unrepaired CoA, but also those after repair with arterial hypertension, residual CoA, or aortic aneurysms have an increased risk of aortic rupture and rupture of a cerebral aneurysm during pregnancy and delivery. An excess of miscarriages and hypertensive disorders is reported.[14)15)]

CONCLUSION

Despite adequate treatment, patients with CoA have a reduced life expectancy and increased risk of cardiovascular complications later in life, related to hypertension, LV dysfunction and hypertrophy, restenosis, aneurysm formation and cardiovascular and cerebrovascular diseases. Timely intervention is crucial, as early treatment may prevent cardiovascular complications. Re-CoA should be proactively addressed. Cardiovascular complications may occur decades after initial treatment, warranting lifelong follow-up. Blood pressure control should be performed yearly, and regular evaluation of the heart and aorta with TTE and/or CMR are recommended.

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