Current management of coarctation of the aorta

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ABSTRACT
Coarctation of the aorta (C) is the sixth most common lesion in congenital heart disease and represents a spectrum of aortic narrowing that varies from a discrete entity to tubular hypoplasia. This condition was once thought to be a relatively simple lesion that would be “cured” upon repair of the narrowing, however, despite relief of the anatomical obstruction the subsequent risk of early morbidity and death persists. This review outlines the optimal management strategy of this disease from neonatal to adult life and provides insights to approach this straightforward but challenging condition.
INTRODUCTION
Coarctation of the aorta (C) is the sixth most common congenital lesion accounting for 4–6 per cent of live births with congenital heart disease. Although most patients have a discrete narrowing of the thoracic aorta at the insertion of the ductus arteriosus, the anatomical spectrum may vary from this discrete entity to tubular hypoplasia, with many variations in between these extremes. Despite these anatomical variations, the effect of the narrowing has the commonly shared features of increased afterload on the left ventricle, exposure of the upper body to hypertension, flow disturbance in the thoracic aorta, and decreased perfusion to the lower body. Depending on the balance between the degree of flow disturbance and the compensatory mechanisms available to overcome it, the clinical presentation may vary from the critically ill neonate in heart failure to the asymptomatic child or adult with hypertension. Untreated coarctation carries a poor prognosis with average survival age of 35 years of age; with 75% mortality by 46 years of age (Figure 1). Long term complications are the consequence of long-term hypertension including premature coronary artery disease, stroke, endocarditis, aortic dissection and heart failure. Furthermore, recurrent coarctation and future aneurysm formation can occur following successful surgical and endovascular repair which mandates long-term close surveillance.

This review focuses on the current management of C from neonatal to adult life and provides insights to approach the straightforward but challenging complications of this disease.

CLASSIFICATION
Native coarctation of the aorta
The classic native C describes the discrete narrowing of the descending aorta resulting from ridge-like thickening of the media of the aortic wall that protrudes into the lumen opposite the insertion of the ductus arteriosus (Figure 2). The origin of the subclavian artery can occasionally be involved with post-stenotic dilatation of the aorta commonly encountered. Tubular hypoplasia is a less common form of native C which involves the isthmus and part of the transverse arch.

Recurrent coarctation of the aorta
Recoarctation of the aorta refers to restenosis after an initially successful surgical or catheter-based repair and is thought to be secondary to either a residual obstruction or development of restenosis. The incidence of recoarctation after surgery is about 10% and occurs independently of the type of
surgical repair used⁵. It is also encountered, to a lesser extent, following balloon angioplasty⁶. It is seen primarily in children due to inadequate aortic wall growth at the site of repair as surgery was performed before the aorta has reached adult size.

INDICATIONS AND TIMING FOR INTERVENTION

The most widely accepted indication for intervention in children and adults is the presence of systemic arterial hypertension, with an upper and lower extremity systolic blood pressure difference $\geq 20$ mg. Milder obstructions may also benefit from intervention by decreasing left ventricular diastolic pressure and preserving left ventricular function in the long term⁷, especially in the presence of hypertension at rest, abnormal blood pressure response during exercise, progressive left ventricular hypertrophy, and in cases of complex heart disease particularly Fontan patients.

In 2008, The American College of Cardiology and American Heart Association (ACC/AHA) guidelines for adults with congenital heart disease recommended intervention for coarctation in the following settings⁵:

- Peak-to-peak coarctation gradient $\geq 20$ mg; which is the difference in peak pressure proximal and beyond the narrowed segment.
- Peak-to-peak coarctation gradient $< 20$ mg with imaging evidence of significant coarctation and radiologic evidence of significant collateral flow. The resting gradient alone may be an unreliable indicator of severity when there is significant collateral circulation.

Infants with “critical” coarctation are at risk for developing heart failure and death when the ductus arteriosus closes. Identification of these patients is essential in order to maintain patency of the ductus prior to surgical repair by continuous intravenous infusion of prostaglandin E1.

Correction of coarctation should be performed in infancy or early childhood to prevent the development of chronic systemic hypertension as delayed repair after early childhood does not prevent persistence or late recurrence of systemic hypertension⁸. If coarctation escapes early detection, repair should be performed at the time of subsequent diagnosis.
TREATMENT OPTIONS

Surgical repair

Surgical repair of coarctation was first described in 1944 and since then many modifications have been developed depending on the anatomy. The types of surgical repair include:

- Resection with end-to-end anastomosis (Preferred repair in patients older than 1 year) (Figure 3A)
- Patch aortoplasty, which is avoided whenever possible because of the frequent occurrence of aortic aneurysm or rupture9 (Figure 3B)
- Bypass graft insertion across the area of coarctation when the distance to be bridged is too long for an end-to-end repair (Figure 3C)
- Subclavian flap aortoplasty in infants with long segment coarctation (Preferred in patients younger than 1 year) (Figure 3D)

Surgery is the preferred treatment of infants with coarctation with an overall survival rate of 98 percent at a median follow-up of 4.8 years of age10. Surgical repair is also possible in preterm infants with birth weights less than 2.5 kg, with an overall survival rate of 76 percent one year after initial repair11. However, in older patients, complications are more common and can be detrimental as surgical repair is associated with extended recovery time, potential phrenic nerve and recurrent laryngeal nerve injury, and the serious, although uncommon, lower body paralysis secondary to ischemic spinal cord injury.

Early postoperative morbidity includes paradoxical hypertension, left recurrent laryngeal nerve paralysis, phrenic nerve injury and subclavian steal3. Paraplegia due to spinal cord ischemia and mesenteric arteritis with bowel infarction are rare complications. Recoarctation is an important long-term complication resulting due to inadequate aortic wall growth at the site of repair when surgery is performed before the aorta has reached adult size. The rate of recoarctation appears to be similar with the different surgical techniques12.

Endovascular repair

Balloon angioplasty

Balloon angioplasty has been an acceptable technique for three decades for the relief of coarctation13. It produces controlled tear of the intima and part of the media which results in an improvement of the vessel diameter. Balloon angioplasty is particularly safe and successful in infants between one and six
months of age with discrete narrowing and no evidence of arch hypoplasia\textsuperscript{14}. It is also considered in critically ill patients regardless of age who have heart failure due to severe ventricular dysfunction, mitral regurgitation or low cardiac output. Nevertheless, extending the application of this technique to all age groups fell out of favor due to the high incidence of future aneurysm formation (up to 9\%)\textsuperscript{15,16} and the availability of stents in most centers.

Recoarctation is a common complication following balloon angioplasty and tends to occur in most neonates after angioplasty requiring repeat balloon angioplasty within 5 to 12 weeks following the initial procedure. As a result, balloon angioplasty is not recommended for infants less than four months of age especially if the lesion is accompanied by arch hypoplasia. Compared to surgery, balloon angioplasty is equally effective in reducing pressure gradient early after intervention; however, the risks of recoarctation and aneurysm formation are greater\textsuperscript{17,18}.

**Stenting**

While balloon angioplasty is effective in relieving vascular obstructions, it has some limitations in that elastic recoil of vessel wall and intimal dissection may not result in effective relief of the blood vessel narrowing. Stent placement after balloon angioplasty or surgery reduces the complications, improves luminal diameter, results in minimal residual gradient, and sustains hemodynamic benefit\textsuperscript{19 – 21}. Stent placement is not generally recommended in patients less than 25 kg due to the small aortic size and the potential injury to the femoral artery from the large sheath required for stent delivery. Children with aortic stent placement are more likely to require a planned reintervention as the stent often needs to be dilated as the child grows. In our practice, we consider stent placement only in patients who are large enough to receive a stent that can be expanded to an adult size.

Several different endovascular stents are commercially available, however very few 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). The choice of stent depends on the coarctation anatomy, size of the patient, the preference of the operator and availability. Balloon-expandable bare metal stents are the most commonly used and are made from stainless steel (Palmaz Genesis, Johnson and Johnson; Mega LD and Maxi LD series, ev3), platinum-iridium alloy (Cheatham-Platinum stent, NED), or chromium-cobalt alloy (Andrtent XL and XXL, Andramed). The chromium-cobalt alloy is stronger than stainless steel and, therefore, thinner struts allow for a lower crimped profile without compromising radial strength. Closed cell stents are strong and rigid (Palmaz Genesis) and markedly foreshorten whereas open cell stents (Mega LD and Maxi LD), although weaker, foreshorten less, conform to the anatomy, and allow access to side branches. A hybrid open-closed cell stent (Andrtent) combines the advantages of these designs. Balloon-expandable expanded polytetrafluoroethylene (TFE)-covered stents are available in a closed cell design (CP stent) and open cell design (Advanta V12 LD, Atrium Medical). It has been proposed that the use of covered stents reduces the risk of aneurysms. However, in a randomized trial of 120 patients with severe native coarctation, there was no difference in the rate of recoarctation 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\textsuperscript{22}. 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.

Data from a multicenter case series of over 500 patients, treated from 1989 to 2005, who were over the age of four years, demonstrated the efficacy and relative safety of stent placement for both native and recurrent coarctation\textsuperscript{21,23}. The success rate of stenting was 98\% in these cases, defined as reduction in the gradient to $< 20$ mg or a ratio of post-stent coarctation to descending aorta of $> 0.8$. Acute complications occurred in 14 percent of patients including two procedure-related deaths.

**Procedural steps**

At our institution, all procedures that involve balloon angioplasty or stent implantation for C are performed under general anesthesia because pain at the dilatation site and patient movement may compromise the success of the procedure. After obtaining arterial access, intravenous heparin is administered achieving an activated clotting time (ACT) goal greater than 200 seconds. A straight-tip catheter (e.g. multipurpose, right judkins) is then advanced into the descending aorta followed by advancing a soft-tipped wire (e.g. Glide wire) through the coarctation site. Once the catheter is advanced into the ascending aorta, it is replaced over the wire by a pigtail catheter followed by careful pullback to obtain pressures across the entire aortic arch and descending aorta. Biplane angiography
is then performed using a calibrated pigtail positioned just proximal to the lesion. The initial angiogram is usually obtained in the left anterior oblique and straight lateral projections (Figure 4A, 4B). The recent introduction of 3D rotational angiography provides an additional tool to understand complex coarctation anatomies. Following angiography, measurements are made at different points of the aorta in order to select the size of balloon or stent to be used. The size of the balloon selected should be no more than 1 to 2 mm larger in diameter than the smallest normal aortic diameter proximal to the coarctation, in cases of recurrent coarctation, the size of the balloon should not be larger than the size of the 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. For balloon angioplasty, we favor using low-profile/low pressure balloons (e.g. Tyshak II balloon (NED Inc., Hopkinton, NY) that permit their passage through smaller sheaths. The balloon is inflated until the wasting of the balloon is abolished. In our experience, high-pressure balloons are rarely needed and low-pressure balloons are effective in treating coarctations.

As stated above, stents have the advantage of exerting radial force that prevents vessel wall elastic recoil and by helping compress dissection flaps against vessel wall, therefore, providing effective relief of obstruction. The choice of stent depends on the coarctation anatomy, size of the patient, the preference of the operator and availability. It is important to note that all stents suffer progressive shortening with deployment and this should be considered when choosing the stent’s length (Palmaz XL stent shortens the most). The selected stent is hand-crimped onto the balloon and an umbilical tape is used to finish crimping on to the balloon. 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. Previously, only single large diameter balloon catheters were available for stent implantation. As these balloons first expand at the ends, they carry the risk of wall injury by the edges of the stent. The subsequent introduction of the balloon-in-balloon (B) (NED, Hopkinton, NY) catheters offered several advantages for stent implantation. These catheters have 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 sizes of 8 to 24 mm, whereas the shaft of the catheter is either 8 or 9 French. Compared to single-balloon catheters, B balloon offers several advantages. 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 a long sheath is required for the delivery of the balloon catheter via the femoral artery. We prefer using the Mullins sheath (Cook, Bloomington, IN), which has a radiopaque tip that can be visualized under fluoroscopy and a side arm that can be used for hand injection of contrast to aid in stent implantation. The delivery sheath selected should be one to two French larger than the indicated for the balloon catheter alone. Once the stent and balloon catheter are ready, an end-hole catheter is maneuvered retrograde across the coarctation followed by exchanging the catheter with a long extra or super stiff wire (e.g. Amplatz extra or super stiff wire). We prefer parking the wire in the ascending aorta as compared to the right or left subclavian arteries. The regular sheath is then exchanged for the selected long delivery sheath and advanced retrograde until the tip of the sheath is slightly beyond the lesion. Once the stent is positioned across the coarctation segment, it is uncovered by gently
withdrawing the sheath while holding the stent in place. The stent position is then verified by test angiogram performed by injecting contrast via the side arm of the sheath. Some operators prefer to place a catheter via either a transeptal puncture to the area above the coarctation or simply via placing a 5 Fr sheath in the left brachial artery. To ensure precise placement during balloon inflation, rapid right ventricular pacing is performed to decrease stroke volume in order to prevent the stent from moving. If the coarctation is very tight, there is no need for rapid right ventricular pacing. Furthermore, we advise against pre stent balloon angioplasty as this approach is associated with more complications than primary direct stenting. If a B balloon 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 catheter is removed over the wire. The distal end of the stent may occasionally lack complete wall apposition, however, we recommend against flaring the stent end as it offers no hemodynamic benefit and may increase the risk of aortic wall injury. At the end, hemodynamic assessment is repeated with pullback or simultaneous pressure measurement using the sheath side-arm to document any residual gradient across the stent (Figure 4A–C).

MANAGEMENT APPROACH

Societal guidelines recommend correction of coarctation as early as possible, optimally early in childhood, to reduce the long-term morbidity and improve survival. The choice of intervention should be determined by a multidisciplinary team experienced in treating patients with congenital heart disease and is dependent on the underlying morphology, age of the patient, and the presence or absence of other cardiac lesions.

Neonates and young infants

In neonates with critical coarctation, an infusion with prostaglandin E1 (alprostadil) to maintain patency of the ductus arteriosus should be administered until corrective surgery can be performed. For patients with heart failure, general supportive care and inotropic agents should be administered. Palliative balloon angioplasty may be considered to stabilize the critically ill patient. Surgical correction is recommended once patient is stable, as it is associated with lower risk for reintervention compared with balloon angioplasty.

Older infants and young children

For younger pediatric patients, surgical correction has been the primary treatment of native coarctation at most centers, however, there has been an increased use of balloon angioplasty in this age group. The decision between these two modalities is determined by the multidisciplinary team, expertise of the center, and the underlying morphology of the coarctation. Our approach is consistent with the 2011 AHA pediatric guidelines. In infants and children between four months and five years of age (weight below 25 kg), we prefer balloon angioplasty if the lesion is discrete and there is no evidence of arch hypoplasia. In the presence of complex coarctation anatomy, the decision to use balloon angioplasty versus surgical repair is made on a case by case basis.

Older children and adults

For larger patients (weight > 25 kg), stenting has become the preferred approach in many centers. In the presence of complex coarctation anatomy like vessel tortuosity and arch hypoplasia, the decision to perform stent placement versus surgical repair is made on a case by case basis.

LONG-TERM CARDIOVASCULAR COMPLICATIONS

Long term problems may occur after all forms of treatment. The most important complications are arterial hypertension, recoarctation and aneurysms of the ascending aorta or at the site of intervention. Further sequelae may develop due to coronary artery disease, bicuspid aortic valve, mitral valve anomalies, infective endocarditis or cerebral aneurysms.

Recoarctation

Restenosis at the site of coarctation is an important cause of morbidity after an initially successful repair. The reported prevalence of recoarctation ranges between 3% and 41% in a survey of 11 major studies. Recoarctation may occur with all surgical techniques (5 – 14%); no single technique appears...
to be superior to the others. This occurs due to inadequate aortic wall growth at the site of repair when surgery is performed before the aorta has reached adult size. Recoarctation can also occur following balloon dilatation. Higher rates of recoarctation tend to occur in smaller patient size, younger age at repair, and in the presence of associated transverse arch hypoplasia. Indications for re-intervention are similar to those for native coarctation (see above). Transcatheter intervention with stenting is our preferred treatment of choice as the mortality for surgical reoperation is higher than for primary repair (1 to 3% versus <1%) and can be as high as 5–10% if there are significant comorbidities or left ventricular dysfunction. Nevertheless, surgical repair is indicated for recoarctation when confounding features are present such as long recoarctation segment, hypoplastic arch, or aortic aneurysm or pseudoaneurysm.

**Aortic aneurysm**

Aneurysms of the ascending aorta or in the region of the aortic isthmus are the most dangerous complications because they carry the risk of life-threatening rupture. Bicuspid aortic valve, aortic wall changes, and systemic hypertension may together be responsible for aneurysmal formation of the ascending aorta.

Aneurysm formation occurs at and around the coarctation site due to an inherent aortic wall medial abnormality (Figure 5A), which is characterized by fragmentation of elastic fibers, an increase in ground substance, and a reduction in the number of smooth muscle cells. Practically all surgical techniques carry the risk of postoperative aneurysms at the site of prior repair. The occurrence depends on the age at the time of surgery and the surgical technique employed. Dacron patch aortoplasty has the highest reported frequency of postoperative aneurysm formation. On the other hand, the lowest incidence is reported after end-to-end anastomosis or after extra-anatomic tube grafts. Aneurysms may also occur following balloon angioplasty of either native or recurrent coarctation (4% to 12%). Because the incidence of aneurysm formation after surgery or balloon angioplasty appears to increase with longer follow-up periods, all patients need careful periodic surveillance. Generally, aortic aneurysms are treated surgically. Alternatively, endovascular stent grafts have been used to repair aortic aneurysms successfully without major complications (Figure 5B).

**Systemic hypertension**

Hypertension is among the factors that contribute to premature death from coronary and cerebrovascular disease following repair of coarctation of the aorta. Although the blood pressure typically falls after successful repair, persistent or recurrent hypertension and disproportionate systolic hypertension with exercise are observed. Hypertension is more common in patients whose repair was performed after 20 years of age compared with those who were corrected in early childhood. The factors responsible for the persistent risk of hypertension are not well understood, but probably related to the decreased compliance of the arterial wall. As noted in the 2008 ACC/AHA adult congenital heart disease guidelines, hypertension should be controlled by beta blockers, angiotensin converting enzyme (ACE) inhibitors, or angiotensin receptor blockers.
LONG-TERM SURVEILLANCE

All coarctation patients, whether repaired or not, should be monitored with life-long congenital cardiology follow-up because of the potential long-term complications. Per 2008 ACC/AHA guidelines, monitoring should include the following:\(^3\):

- Close observation for the presence of systemic hypertension
- Imaging of the coarctation repair site by MRI or CT performed at intervals of 5 years or less depending on the specific anatomic findings before and after repair. In infants and younger children, echocardiography may suffice due to better acoustic windows.
- Periodic echocardiography to assess bicuspid aortic valve function, ascending aorta, and ventricular function.
- Cranial imaging with either MRI or CT should be performed in all patients with aortic coarctation to evaluate for intracranial (berry) aneurysms (Figure 6). There is 10% prevalence of such aneurysms in patients with coarctation compared to 3% in the general population.
- The role of routine exercise testing for follow-up in adults is not well established; however it should be performed in all patients who wish to participate in competitive sports.
- Endocarditis prophylaxis is only indicated in patients with a past history of endocarditis, in those whose repair involved insertion of a conduit, or for six months after intervention if prosthetic material or stent was used.

![Figure 6. Angiogram of a young patient with coarctation and cerebral aneurysm (arrow).](image)

CONCLUSIONS

C is a lifelong disease that affects the entire pre-coarctation arterial tree. Close follow-up and aggressive management of complications is crucial. Despite the advances in the recognition and treatment of patients with coarctation, future endeavors should be directed towards improving the procedural aspects and long-term morbidity and survival of this condition. One of the promising recent advances in stent technology is the development of biodegradable stents which may offer a solution for endovascular therapy for neonatal and infant coartations. These stents keep the coarcted aortic segment open and dissolve over a period of months. The scaffolding left over may create a favorable ratio of the normal aortic tissue to abnormal tissue which in turn may prevent significant renarrowing. Clinical trials are yet to show the feasibility and efficacy of this technology.

REFERENCES

