

Interventional Therapy for Coarctation of the Aorta in Adults

What we have learned over the last 2 decades.

BY DAISUKE KOBAYASHI, MD, AND THOMAS J. FORBES, MD

Coarctation of the aorta (CoA) is one of the more common congenital heart lesions and occurs in 5% to 8% of all congenital heart defects. Optimal treatment of adults presenting with native CoA remains controversial. Surgical repair is the traditional treatment of native CoA. During the past 2 decades, balloon angioplasty (BA) and stent treatment (ST) have gained wider acceptance in the treatment of native CoA in older children and adults. In this article, we present current data regarding surgery, BA, and ST in the treatment of native CoA and will not discuss treatment of recurrent CoA.

SURGERY

The preferred surgical approach to CoA is end-to-end anastomosis with CoA resection. However, in adults, due to increased difficulties in mobilizing the aorta, other less optimal approaches are frequently required. In the Congenital Cardiovascular Interventional Study Consortium (CCISC) experience of 99 patients undergoing surgical repair of their CoA, none of the patients older than 16 years underwent end-to-end repair of their CoA.¹

In the CCISC CoA registry, 111 patients were enrolled in the surgical group as of July 2015. The primary surgical technique was end-to-end anastomosis in young children (10 years or younger). Tube graft and patch augmentation were used more frequently in adolescents and adults. No adult patients underwent end-to-end anastomosis repair (Figure 1). The CCISC results regarding the type of surgical repair in adults are similar to those of the Mayo clinic experience, as noted in Figure 2.²

Short-term follow-up by integrated imaging showed surgery had a high incidence of aortic complications (23%), including moderate to severe reobstruction and aortic aneurysm formation.¹ In the current CCISC registry, 19.2% of 99 patients undergoing surgery experienced acute com-

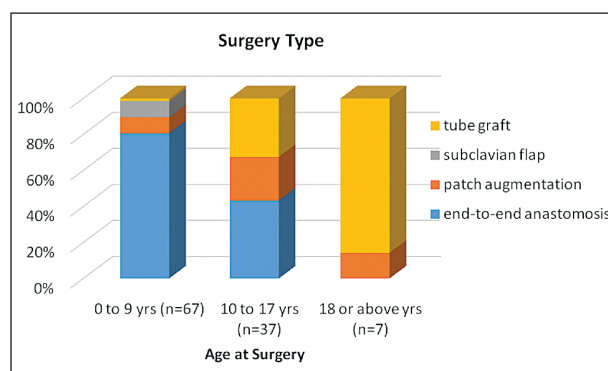


Figure 1. Type of surgical repair for CoA in three age groups.

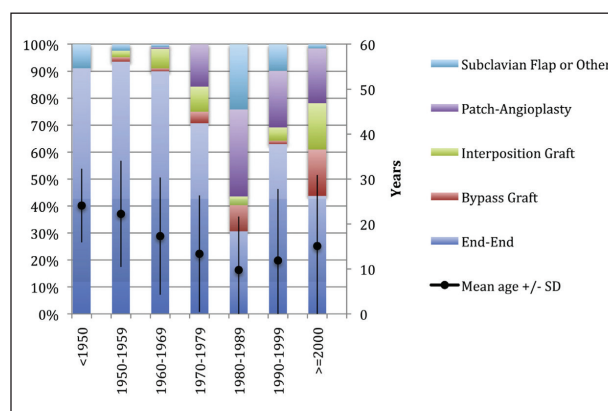


Figure 2. Type of repair by decade. The type of CoA repair was stratified by decade of repair. Mean $\pm$ SD age, by decade, at time of operation is also included. Reprinted with permission J Am Coll Cardiol, 62, Brown ML, Burkhardt HM, Connolly HM, et al, Coarctation of the aorta lifelong surveillance is mandatory following surgical repair, 1020-1025, 2013, with permission from Elsevier.²

TABLE 1. AMERICAN HEART ASSOCIATION RECOMMENDATIONS FOR STENT PLACEMENT IN NATIVE AND RECURRENT CoA

Class	Indications
Class I	1. Recurrent CoA with gradient > 20 mm Hg, in which the stent can be expanded to an adult size
Class IIa	1. Native or recurrent CoA, with (a) Gradient > 20 mm Hg (b) Gradient < 20 mm Hg but with systemic hypertension (c) A long-segment CoA with gradient > 20 mm Hg 2. Native or recurrent CoA with failed balloon angioplasty
Class IIb	1. Complex aortic arch obstruction that persists despite surgical or catheter-based therapy, when further surgery is considered as high risk, in neonates and infants 2. Native or recurrent CoA, with (a) Gradient < 20 mm Hg but with elevated left ventricular end-diastolic pressure (b) Gradient < 20 mm Hg with significant aortic collaterals, resulting in an underestimation of CoA

Data summarized from Feltes TF, Bacha E, Beekman RH 3rd, et al. Indications for cardiac catheterization and intervention in pediatric cardiac disease: a scientific statement from the American Heart Association. Circulation. 2011;123:2607–2652.

plications, including severe prolonged hypertension, atrial fibrillation, pleural effusion, neurologic/spinal cord injury, chylothorax, and vocal cord paralysis. Aneurysm formation was seen at 12.3% at short term and 10.5% at intermediate follow-up.

BALLOON ANGIOPLASTY

BA is effective for the treatment of native CoA, especially in older children and adults.³ However, an important associated complication observed with BA is aneurysm formation. CCISC recently reported the outcome of BA in 130 patients with native (n = 76) and recurrent (n = 54) CoA.⁴ BA was found to be effective in the treatment of CoA acutely and at intermediate follow-up. At short-term follow-up (mean, 12.6 months), the rate of aortic wall complications (dissection, aneurysm formation, or intimal tear) was high, at 48% in native CoA (n = 45) and 21% in recurrent CoA (n = 30). The aortic wall complication rate increased at intermediate-term follow-up (mean, 32 months; native 57% and recurrent 33%). In the CCISC CoA registry, BA clearly is not considered the treatment of choice in adults with native CoA. Of 144 patients undergoing BA for treatment of CoA, only eight adults (6%) underwent BA of their native CoA. One study sticks out as being a strong advocate for performing BA in adults with native CoA. Hassan et al reported on 49 patients at a mean age of 24 years (range, 14–59 years) who underwent BA of their isolated native CoA. An immediate decrease in systolic gradient from 66 (± 23) mm Hg to 10.8 (± 7) mm Hg was observed, which persisted at 1-year follow-up catheterization at which the systolic gradient was 6.2 (± 6) mm Hg. At a mean follow-up of 11.5 years, aneurysm formation was observed in 7.5% patients with no reobstruction noted. No reinterventions were required in this group.³

STENT PLACEMENT

ST is considered the preferred approach at many centers. Indications for stent placement in native and recurrent CoA are outlined in Table 1. In the CCISC experience, ST and surgical treatment of CoA was superior to BA both acutely and at short-term follow-up, although there were no differences between the three groups at intermediate follow-up. Importantly, technical complications of CoA stenting have significantly decreased during the last 2 decades,^{1,5} mainly due to improvements in balloon catheter and stent technology. In our early experience, the CCISC analyzed the procedural results and acute complications of 565 CoA stent procedures; 87% of patients were older adolescents and adults.⁵ Although ST effectively treated CoA in the majority (97.7%), the rate of acute complication (14.3%) was not negligible.

Adults With CoA Stenting

From the CCISC study, we have learned important findings in treating adults with native CoA through ST.^{1,5} Patients aged 40 years or older had the highest complication rates (31%), primarily aortic wall complications. We suspect that aortic wall compliance is decreased in this group—it is likely that the aorta may be less resistant to expansion in adults. In this high-risk adult group, we recommend aiming for hemodynamic success rather than angiographic resolution of the CoA. The prophylactic use of covered stents for CoA would be a safer alternative in older patients. Currently, the COAST II trial is assessing the safety and efficacy of covered Cheatham-Platinum stents (NuMed, Inc.) for prevention or treatment of aortic wall injury associated with CoA. Covered stents are important to have in your cath lab for bailout procedures when aortic wall injury occurs in higher-risk, older patients. The risk of

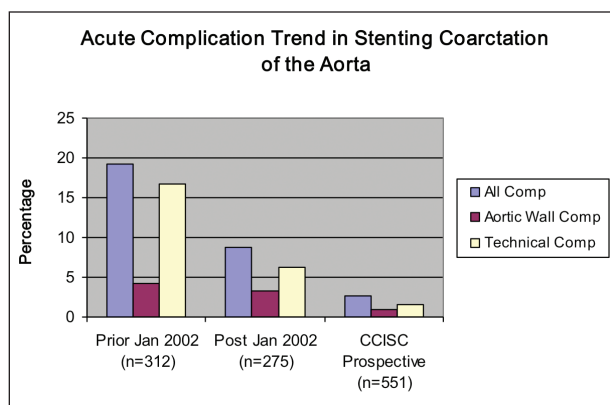


Figure 3. Current trends in encountering acute complications for stent treatment of CoA.

cerebrovascular accident is higher during stent deployment in older patients. Extra caution should be taken to manipulate a wire or catheter in a brachiocephalic vessel, and heparinization should be maintained at an activated clotting > 250 seconds to provide adequate anticoagulation during procedure.

TECHNIQUE

In the CCISC experience, ST for CoA is performed under general anesthesia in 95% of patients. The most frequently used balloon is the BIB (Balloon-in-Balloon, NuMed, Inc.), followed by Z-Med (B. Braun Interventional Systems Inc.) and PowerFlex (Cordis Corporation). Palmaz 10 series and Genesis XD stents (Cordis Corporation) are the most widely used stents. The IntraStent LD stent (Medtronic) is an excellent alternative due to its open cell design when there is a possibility the stent will cover the origin of a brachiocephalic vessel.

COMPLICATIONS

Over the past 2 decades, we have observed a significant decrease in the complication rates associated with stenting CoA. Acute complications are broken down into aortic wall complications (dissection, intimal tear, aneurysm formation) and technical complications (vascular injury, balloon rupture, stent embolization). Figure 3 shows a twofold decrease in aortic wall complications and nearly an eightfold decrease in technical complications. The reasons for this appear to be multifactorial.

The main decrease in aortic wall complications appears to be related to a decrease in short-term aneurysm formation. Early in our experience, aggressive dilation of a tight coarctation segment, sometimes up to eight to 10 times the size of the original diameter, was performed. This was at a time when covered stents were not (and are still not) widely available in the United States. The aggressive dilation of the tight coarctation segment was believed to have played a



Figure 4. Stent placement in a native coarctation segment that measured 2.4 mm in diameter and was dilated up to 16 mm. Eight months later, a large aneurysm was noted to have developed at the coarctation site. The aneurysm was successfully treated with a covered stent.

role in the development of large aneurysms at short-term follow-up (Figure 4).⁵

Over the past 18 years, a staged approach to stenting CoA has been used in which the narrowest segment is not dilated greater than four times its original size in the initial setting. Since staging has been used, the CCISC has not observed large aneurysm formation at either the short or intermediate term for the past 15 years. Aneurysms are still observed in approximately 3% of the patients, although they are the more typical bubble aneurysms that are observed at intermediate follow-up (Figure 5).

In the CCISC experience, the majority of these smaller aneurysms have not increased in size, with 8% requiring covered stent treatment due to progression in size. Thus far, all of these aneurysms are located on the greater curvature of the aorta.

Another aortic wall issue, especially noted in adults, relates to the formation of acute dissection or frank aortic wall rupture. Fortunately, this is an exceedingly rare entity but can have devastating consequences for the patient. Some investigators have advocated balloon compliance testing of the coarctation segment. A balloon catheter is inflated up to 4 atm, and if an indentation still persists in the balloon, that patient is believed to have a noncompliant aorta and would not be a candidate for either aggressive BA or stent placement. We take a different approach. The stent is loaded on the balloon catheter and inflated up to 3 atm across the coarctation site. If a significant waist is observed in the stent, no further intervention is done in that setting. If, at 6-month follow-up, there is a significant upper to lower extremity systolic gradient (> 20 mm Hg), or if the patient remains hypertensive, more aggressive dilation is performed. Covered stents, either handmade using Gore-Tex or the Cheatham-Platinum stent under the COAST II trial research protocol, should be available for emergency use.

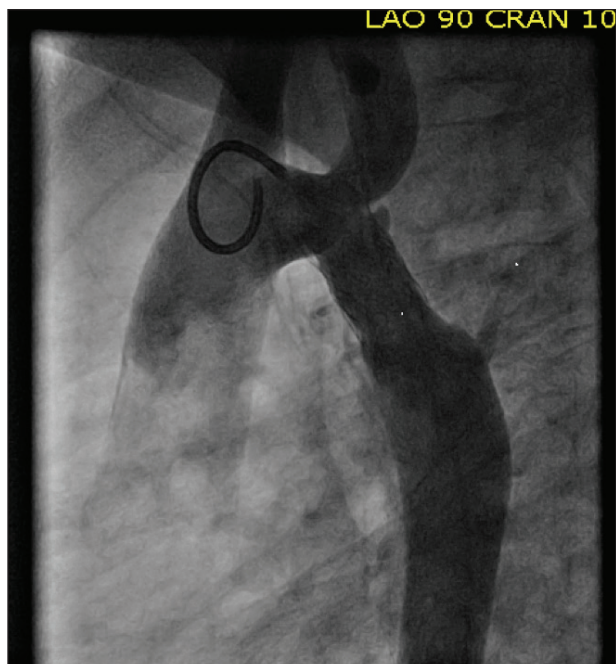


Figure 5. Small “bubble” aneurysm noted on the greater curvature of the aorta, proximal to the coarctation site, 2 years after stent placement for severe native CoA.

The most dramatic decrease in acute complications has been seen in the technical aspects of the procedure. Improvements in balloon technology have significantly decreased the amount of balloon ruptures observed with initial stent deployment and have aided interventionists in achieving adequate stent placement across the coarctation site. In fact, stent malposition has decreased by nearly 80% from the 1990s to 2012 in the CCISC registry. Although there has been no single technique that has been proven to decrease the likelihood of stent malposition, two techniques have been more frequently used to improve the likelihood of achieving proper stent deployment across the coarctation site.

The first technique is rapid right ventricular pacing during stent deployment. In the CCISC experience, nearly 15% of operators use rapid right ventricular pacing in the majority of their cases. They are more likely to use pacing for the treatment of transverse aortic arch obstruction and less likely to use it for discrete isolated, severe coarctation located in the distal isthmus.

The second technique that has been used is distal balloon inflation. All balloon catheters inflate proximal to distal. There is the possibility of the stent migrating distally off the balloon catheter during initial deployment, although it is more likely that the balloon catheter, inflating proximal to distal, milks itself (along with the stent) distal to the coarctation lesion, thereby embolizing the fully expanded stent into

the distal aorta. This latter situation was the main technical complication encountered, observed in 6% of all coarctation stent cases in our initial multicenter report. To minimize this risk, the sheath is used to constrain the proximal balloon from expanding, with the distal balloon and stent fully deployed in a controlled setting, locking it into place before the sheath is fully pulled off the balloon catheter and the proximal stent is fully deployed.

In the same light, the CCISC has seen a dramatic decrease in vascular arterial complications (42%) over the past 8 years. We believe this is primarily related to extra attention given to properly accessing the femoral artery and the use of closure devices at the completion of the procedure.

Follow-up integrated imaging is essential for any patient undergoing BA, surgical repair, or ST of their native CoA. In the CCISC study regarding ST of CoA, long-term procedural success was 77%.⁶ In the ST group, the rate of aortic wall complications and recurrent obstruction was 2% and 20% at follow-up, respectively.⁶ Repeat intervention was required in 12% of cases. However, this rate decreased to only 4% when planned/staged repeat interventions were excluded. At intermediate follow-up, 25% of patients continued to have hypertension and one-third were taking antihypertensive medication, which is similar to their surgical and BA counterparts. Regardless of what treatment is chosen to treat adult CoA, lifelong blood pressure management and occasional integrated imaging is warranted.^{1,2} ■

Daisuke Kobayashi, MD, is with the Division of Pediatric Cardiology, Children's Hospital of Michigan, Carman and Ann Adams Department of Pediatrics, Wayne State University School of Medicine in Detroit, Michigan. He has stated that he has no financial interests related to this article.

Thomas J. Forbes, MD, is with the Division of Pediatric Cardiology, Children's Hospital of Michigan, Carman and Ann Adams Department of Pediatrics, Wayne State University School of Medicine in Detroit, Michigan. He has stated that he has no financial interests related to this article. Dr. Forbes may be reached at (313)745-5481; tforbes@dmc.org.

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