

Coarctation of Aorta-Management Options and Decision Making

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Research Article

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Abstract

Coarctation of the Aorta (CoA) is a congenital cardiac anomaly consisting of a constricted aortic segment with a prevalence of 5 to 8% of all Congenital Heart Defects (CHD). The classic CoA is located in the thoracic aorta distal to the origin of the left subclavian artery, at about the level of the ductal structure. Significant hypertension and/or congestive heart failure are indications of intervention. If hypertension (rather that heart failure) is the clinical problem, it is better to relieve the aortic obstruction promptly rather that attempting to "treat" hypertension with antihypertensive medications. Surgical relief of the aortic obstruction and catheter interventional techniques (balloon angioplasty and stents) are available alternatives. Since the introduction of surgical correction by Crafoord and Nylin and Gross and Hufnagel in early 1940s, surgical therapy has been the treatment of choice for aortic coarctation. Gruntzig's technique of balloon angioplasty was adopted by Singer and Sperling and their associates to enlarge coarcted aortic segments in post-surgical recoarctation and native coarctation, respectively in early 1980s. This is followed by the application of the technique by other cardiologists to treat native CoA. The procedure consists of inserting a balloon angioplasty catheter across the site of coarctation and inflating the balloon with diluted contrast material. Both immediate and followup results are reasonably good. Residual and recurrent obstructions following surgery and prior balloon angioplasty are also amenable for balloon angioplasty. Despite reasonably good short-term and long-term results of balloon angioplasty, some problems remain and include restenosis, probability of aortic rupture, formation of aneurysms and inability to effectively treat long-segment tubular narrowing. Because of these and other reasons, endovascular stenting of aortic coarctation has gained acceptance over the last decade. The balloon catheter, with the stent mounted on it, is advanced over a stiff guide wire and positioned across the coarctation segment and the balloon inflated, thus implanting the stent. Most cardiologists use stents in adolescents and adults and restrict their use in younger children because of issues related to growth. Stent therapy appears to be an attractive method for treatment of native or recurrent coarctation, aneurysm formation following prior surgical or balloon intervention and for long segment hypoplasia. Role of covered stents to manage aortic coarctation is limited and are used when the assessed risk for development of aneurysm or dissection is high. A comparison of all available treatment modalities was made and while it is difficult to make a definitive statement, the overall data seem to indicate that transcatheter methods may be better than surgery. Selection of the method of therapy is largely based on the age at presentation and anatomy of the coarcted segment and surrounding structures; surgery for infants and long segment coarctations, balloon angioplasty for discrete native and post-surgical coarctations in children and stents for long segment coarctations as well as any type of coarctations in adolescents and adults seem to be the current trend.

Keywords: Coarctation of the aorta; Surgery; Balloon angioplasty; Stents; Covered stents; Treatment options

Introduction

Coarctation of Aorta (CoA) is a congenital abnormality of the heart producing obstruction to blood flow through the aorta; it consists of a constricted aortic segment comprising localized medial thickening with some infolding of the media and superimposed neointimal tissue. It may be a shelf-like structure or a membranous curtain-like structure with an eccentric or a central opening. Most commonly it is located at the junction of the ductus arteriosus with the aortic arch, just distal to the left subclavian artery. Rarely, the coarcted segment is present in the lower thoracic or abdominal aorta, long and fusiform with irregular lumen and may be considered as a variant of Takayasu arteritis. Coarctation can occur in isolation, in association with bicuspid aortic valve or with major cardiac malformations [1]. CoA accounts for 5-8% of children born with congenital heart disease [2,3]. The majority of coarctations are newly diagnosed in childhood; less than 25% are recognized beyond 10 years of age [4]. Despite of our extensive experience and knowledge regarding CoA, there are controversies when it comes to its management in children. Surgical correction was first performed in mid 1940s [5,6] and since then it has become standard method of therapy for CoA. Transcatheter balloon angioplasty of CoA began in late 1970s but became more popular in late 1980s thru' early 1990s [7]. Over the past decade we have started using intravascular stents for management of coarctation, mainly in adolescents and adults [8]. In this paper we will review the indications for intervention, available treatment options, selection of method of intervention along with results and compare various treatment modalities.

Management at presentation

Early presentation during neonatal period or infancy: In neonates with critical coarctation, Prostaglandin E1 is infused intravenously to open the ductus arteriosus [9] and to maintain the systemic circulation (Figure 1). Assisted ventilation and inotropic drug (dopamine, dobutamine, epinephrine) infusion should be used, as indicated. In neonates umbilical artery line is very useful to assess the response to prostaglandins with direct post ductal blood pressure measurement. Most patients should undergo surgical or catheter intervention to relieve the coarctation after initial stabilization. In non-critical patients presenting with congestive heart failure symptoms, diuretics and inotropic drugs are used for initial management.

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Late presentation: Older infants and children may present because of hypertension or a cardiac murmur. In children presenting because of hypertension, treatment of severe hypertension is important. Mild hypertension does not require treatment. Beta- blockers are the first choice of drugs with secondary hypertension with coarctation. Therapy should be guided to achieve near normal upper extremity blood pressure while taking care not to compromise systemic perfusion. Beta-blocker therapy has been shown to decrease the severity of postoperative hypertension but most patients do require brief period of therapy with short acting drugs such as nitroprusside or short acting intravenous beta-blocker, esmolol. For long term postoperative therapy beta-blockers are first choice with addition of angiotensin converting enzyme inhibitors and angiotensin II antagonists as adjuncts [10]. One should remember that relieving the aortic obstruction promptly is better than attempting to achieve normal blood pressures with antihypertensive medications [10,11].

Indications for surgical or transcatheter intervention

Significant hypertension and congestive heart failure are indications for intervention. A peak-to-peak systolic pressure gradient more than 20 mmHg across aortic coarctation is generally required for intervention. Surgery and catheter interventional techniques (balloon angioplasty and stents) are available alternatives for management CoA. Neonates and infants who are symptomatic should have urgent intervention as soon as the infant is stabilized. Asymptomatic infants, children, adolescents, and adults should undergo the procedure electively. If neither hypertension nor heart failure is present, elective surgical or balloon therapy in children between the ages of one and five years is recommend. Waiting beyond that age is not advisable because of the evidence of residual hypertension if intervention is performed after 5 years of age [12]. More recent studies [13] support the idea that intervention prior to first birth day is likely to avoid hypertension during follow-up.

Surgical therapy

Surgical correction for CoA was first introduced by Crafoord and Nylin [5] and Gross and Hufnagel [6]. A variety of techniques have been used in repairing aortic coarctation and these include resection and end-to-end anastomosis [5,6] subclavian flap angioplasty[14], prosthetic patch aortoplasty [15] and tubular bypass grafts [16,17]. Several modifications of the initially described techniques [18-20] have been introduced over the time. Age of the patient, aortic arch anatomy, associated defects and preference of the surgeon influence the type of surgical procedure used. Surgical correction has had a major favorable impact on the prognosis of the patients with CoA. However, the surgical procedures are also associated with significant risks and complications. Operative mortality in neonates and infants is high (4 to 50%) [21,22] while that for older children is low (0 to 5%) [21,22]. Overall prognosis after surgery is largely dependent upon the associated cardiac anomalies. Other known complications associated with surgical repair are significant recoarctation (6 to 33% in infants, 0 to 18% in older children) [21,22] formation of aneurysms in all types of coarctation repair [23], particularly well-documented following prosthetic patch angioplasty [23], development of paraplegia [24,25], paradoxical hypertension [26,27] and vascular complications[28-32] related to subclavian flap repair continue to be problems.

A recent multicenter observational study by Forbes et al. [33] presented short and intermediate term outcomes in 72 patients with body weight \geq 10 kg who had surgery between 2002 and 2009. At the time of discharge after surgery, upper to lower extremity gradient was 7.7 \pm 18.2 mmHg. 73% patients had reduction in upper and lower extremity gradient to \leq 15 mmHg. Average length of stay was 6.4 days. Over a 5-year follow up period, 25% of patients had complications; 12.5% patients were reported to have aneurysm and 18.8% patients had recoarctation (mild, moderate and severe - 6.3% each). Mean coarctation to descending aorta ratio was 0.98. Four of 72 patients required unplanned reintervention. These results are not significantly different than those observed in previous reviews [21,22].

Balloon angioplasty

Gruntzig's technique of balloon angioplasty [34] was adopted by Sos [35], Singer [36] and Sperling [37] and their associates to enlarge coarcted aortic segments in a postmortem specimen, post-surgical recoarctation and native coarctation respectively. This is followed by a report of successful use of this technique in a sizeable population of patients with native aortic coarctation by Lababidi [38]. Subsequently, the procedure was adopted by other groups of workers [39-41] including our own group [21,22,42-44].

Technique: Cardiac catheterization and selective cineangiography are performed percutaneously to confirm the clinical diagnosis, to evaluate associated cardiac defects and to assess suitability for balloon angioplasty. Once balloon angioplasty is decided upon, a # 4– to 5– French pigtail or multipurpose catheter is introduced into the femoral arterial sheath and is positioned across the coarctation segment. A 0.018 to 0.035 inch J-tipped guide wire is passed through the catheter into the ascending aorta and the tip of the wire positioned either in





the ascending aorta or in the right subclavian artery. The selected balloon angioplasty catheter is positioned across the aortic coarctation and the balloon is inflated (Figure 2) with diluted contrast material to approximately three to five atmospheres of pressure or higher, depending upon the manufacture's recommendations. The balloon is inflated for duration of about 5 seconds. A total of two to four balloon inflations are performed 5 minutes apart. Then repeat aortography and measurement of pressure gradients across the CoA are performed. Recording of heart rate, systemic pressure, and cardiac index prior to and fifteen minutes after balloon dilatation are made to assure that change in pressure gradient is not related to changes in patient status, but is, indeed related to balloon dilation [22,45].

We generally perform this procedure under general anesthesia. Most cardiologists use percutaneous femoral artery approach with ultrasound guidance, as necessary. Occasionally, femoral artery cutdown [40,46] or left axillary artery cut-down [40] may have to be used if percutaneous approach is unsuccessful. Transumbilical approach [47,48] is also used in neonates in an attempt to avoid potential injury to the femoral arteries. Transvenous approach has been used in patients with associated transposition or double outlet right ventricle or large ventricular septal defect [49,50].

The size of the balloon chosen for angioplasty is two or more times the size of the coarcted segment, but no larger than the size of the descending aorta at the level of the diaphragm, as measured from a frozen frame of cineangiogram. Usually a balloon that is midway between the size of the aortic isthmus (or transverse aortic arch) and the size of the descending aorta at the level of diaphragm is chosen. If there is not an adequate relief or obstruction (pressure gradient reduction to <20 mmHg and angiographic improvement), a balloon as large as the diameter of the descending aortic at the level of diaphragm is chosen for additional dilatation [22,45,51]. Heparinization of the patient with 100 units/kg is undertaken; heparin is usually given prior to introducing the left heart catheter. Activated clotting times should be measured every 30 minutes and maintained between 200 and 250 sec [52,53]. It is highly important that tips of a catheter or a guide wire are not manipulated over the site of freshly dilated coarctation in order to prevent inadvertent aortic perforation [46]. A guide wire should always be left in place across the coarctation segment, and all angiographic and balloon-dilatation catheters should be exchanged over the guide wire.

Immediate results: There have been multiple studies involving small number of patients looking at outcomes of balloon angioplasty of coarctation. Forbes et al. [33] showed that post-intervention discharge upper and lower extremity gradient was 10.3 \pm 12.9 mmHg. Mean percent increase in coarctation segment measurement was 125%. Approximately 10% patients had aortic wall injury (dissection/ intimal tear) and no aneurysms were reported in short term follow up. Average length of stay for patients was 3.6 days [33]. These results are comparable to previous reports by Rao et al.[11,21,44]. Reduction of pressure gradient (Figures 3 and 4) across the coarctation and increase in the size of the coarctation segment (Figures 5-7) were observed in all age groups. They also reported diminution of collateral vessels (Figure 8). The femoral pulses, which had been either absent or markedly reduced and delayed (when compared to brachial pulses), became palpable with increased pulse volume after balloon angioplasty. The infants who were in heart failure improved as did their hypertension. The infants who were ventilator dependent could be weaned off of the ventilator support and were extubated. Most infants (beyond neonate



Figure 3: Bar graph of peak systolic pressure gradients (mmHg) across the coarctation of the aorta prior to (Pre) and immediately following (Post) balloon angioplasty in neonates (<30 days), infants (1 to 12 mo) and children (1 to 15 years). Note significant (P < 0.001) decrease in pressure gradient in each subgroup. Mean + standard error of mean (SEM) are shown. N indicated the number of patients in each subgroup.



period) and children are discharged from the hospital within 24 hours after balloon angioplasty. None of their patients required immediate surgical intervention [11,21,44].

Intermediate-term follow-up: A 5-year follow up of 25 patients by Forbes et al. [33] showed that upper and lower extremity gradient was 5.5 ± 14.3 mmHg. About 82% of patients had upper and lower extremity gradient ≤ 15 mmHg; 72% of patients had normal blood pressure during follow-up visits. Only 16 % of patients were on antihypertensive therapy at the time of follow up. There was 19.2 % incidence of aneurysms. Total of 18.8% patients had evidence of reobstruction; all of them were mild. Mean coarctation/descending aorta ratio was found to be 0.79. Six of 61 patients required reintervention of which 2 were planned procedures. Rao et al. [11,21,44] reported intermediate (and long-term) follow up of sixty patients (58 catheterization, 2 clinical). The residual gradients 14 months following angioplasty remained low at 16+15 mmHg (Figures 4 & 9). These gradients continue to be lower (p<0.001) than those prior to angioplasty (46+17 mmHg) and are slightly higher (p<0.05) than the gradients (11+9 mmHg) immediately following angioplasty (Figure 9). Angiographically measured coarctation segment remained

COARCTATION OF THE AORTA BALLOON ANGIOPLASTY – 3-MONTH OLD INFANT



Figure 5: Selected cineangiographic frames from aortic arch cineangiograms in 20°left anterior oblique view in a three-month-old infant demonstrating aortic coarctation (arrow) prior to (A) and immediately after balloon angioplasty (B). Note improvement in coarctation segment size. C, catheter in the superior vena cava; CC, common carotid artery; DAo, Descending aorta; Inn, innominate artery; LSA, left subclavian artery.

COARCTATION OF THE AORTA BALLOON ANGIOPLASTY – 9-MONTH OLD INFANT



Figure 6: Left ventricular (LV) cineangiogram in a postero-anterior view (A) showing discrete aortic coarctation (white arrow - Coarct) in a 9-month-old. Following balloon angioplasty (B) the size of coarcted segment improved markedly (arrow). C, catheter.

wide (Figures 10 and 11) in most patients. Aneurysms developed in 3 (5%) of 58 who underwent follow-up catheterization and angioplasty; one of these patients required surgical excision of the aneurysm and the other two were small and are followed clinically. Recoarcation, defined as peak to peak gradient in excess of 20 mmHg developed in 15 (25%) of 60 patients; younger the baby, the greater is the probability for recoarctation. Repeat balloon dilatation has been found to be useful [44,54] in relieving the recurrent or residual obstruction (Figure 12).

Long-term follow-up: There is scanty data on long-term followup after balloon angioplasty of native Coarctation [44,55,56]. Despite the problems of recoarctation and aneurysms, some requiring repeat intervention at intermediate-term follow-up, the long-term follow-up results (5 to 9 years) appear encouraging in that there was minimal incidence of late recoarctation and no late aneurysm formation. In the majority of children, the arm blood pressure remained normal and blood pressure-determined gradient between arms and legs remained low (Figure 9).

Stent therapy

O'Laughlin et al. [57] were the first to report the use of stents for

the treatment of aortic coarctation, although the result in a 12 year old child was marginal. The first series of stents in 10 patients was reported by Suarez de Lezo et al. [58] followed by others [59-62].

Indications for stents: The indication for employing stents are: a) long segment coarctation, b) associated hypoplasia of the isthmus or aortic arch, c) tortuous coarctation with mal-alignment of proximal with distal aortic segment and d) recurrent aortic coarctation or an aneurysm following prior surgical or balloon therapy. Most interventionists use stents in adolescents and adults while some cardiologists may opt for their use in younger children.

Technique: The diagnostic component is similar to that described in the balloon angioplasty section. Following angiography (left anterior oblique and lateral projections), measurements are made of the diameter of the coarctation segment, diameter of the aorta proximal and distal to the obstruction and "stentable" length of the vessel; all measurement are made in two views and averaged. There are various descriptions of stent delivery in practice with individual modifications. At our institution, we use balloon in balloon (BIB) catheter to implant the stent in adolescents and adults. Balloon-mounted stents may be



Figure 7: Selected cine frame from a postero-anterior view of left ventricular (LV) cineangiogram (A) showing discrete aortic coarctation (white arrow) in a 10-year-old child. Following balloon angioplasty (B), aortogram revealed no evidence for significant residual coarctation (black arrow). Ao, aorta; DAo, descending aorta.



Figure 8: A selected cineangiographic frame from a postero-anterior view of an aortogram prior to balloon angioplasty, showing aortic coarctation (arrow) and a large number of collateral vessels (a). Immediately following balloon angioplasty (b), the aortogram shows marked decreased in collateral vessels. In b, the site of dilated coarctation segment (arrows) is wide open. Also, note a greater opacification of the descending aorta (DAo) in b than in a. Ao, aorta.

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Figure 9: Bar graph of immediate and follow-up results after balloon angioplasty of aortic coarctation. Peak-to peak systolic pressure gradients across the coarctation in mmHg (mean + SEM) are shown. Note significant (p < 0.001) drop in the gradient following balloon angioplasty (Pre, prior to vs. Post, immediately following). The gradient increases (p < 0.05) slightly at a mean follow-up to 14 mo (range, 4 to 56 mo). However, these values are lower (p < 0001) than prior to angioplasty. At late follow-up (LFU) (median 5 years) following balloon angioplasty, blood pressure-measured arm-leg peak pressure difference is lower than catheterization measured peak gradients prior to (p < 0.01) balloon angioplasty and those obtained at intermediate-term follow-up (p < 0.01). Mean + standard error of mean (SEM) are shown. N indicated the number of patients undergoing balloon angioplasty.

 COARCTATION OF THE AORTA

 BALLOON ANGIOPLASTY - FOLLOW-UP RESULTS

 PRE
 POST

 FOLLOW-UP



Figure 10: Selected cine frames from angiograms of a nine-month-old infant prior to (A), immediately following (B), and 1 year after (C) balloon angioplasty. Note that the coarcted aortic segment (arrow) improved markedly after angioplasty (B) which remains wide open at follow-up (C). Ao, aorta; C, catheter; DAo, descending aorta; LV, left ventricle.

used in infants and young children which we do not advocate [63,64]. Once the percutaneous access of the femoral artery is established, heparin is administered intravenously to keep activated clotting time between 200 and 250 seconds. A multipurpose catheter is advanced over the wire to navigate through the coarctation. After traversing the coarctation segment, the catheter is positioned in the right subclavian artery, and an extra- stiff, J-tipped, Amplatzer guide wire is passed and it tip placed deeply in the right subclavian artery and the multipurpose catheter removed. Next a long blue Cook or a similar sheath with a multipurpose curve is inserted over the guide wire and positioned across the stenotic lesion. Once the tip of the sheath is positioned proximal to intended site, the dilator is removed. The guide wire is left in place and the sheath flushed. The selection of the sheath size is dependent on the sheath size necessary for stent deployment. When using BIB catheters, a sheath size 1-French larger than that required by the BIB catheter is used.

The selected stent is hand-crimped onto the balloon and we use

an umbilical tape to finish crimping on to the balloon. Angiographic roadmap is used for positioning the stent. The balloon should preferably be slightly longer than the stent to help with even deployment of the stent. Use of a BIB catheter will generally prevent complications such as balloon rupture and stent migration. The balloon/stent assembly is introduced thru' the valve of the delivery sheath either with the help of a tubular tool that is supplied with some of the stents or slowly ease the stent across the valve of the sheath, to prevent inadvertent dislodgement of the stent from the balloon. The stent is positioned across the coarctation site and its position verified by test angiogram (Figure 13A) via the side arm of the implantation sheath. The inner balloon of the BIB catheter is then inflated (Figure 13B), and angiogram are performed through the sheath to confirm position of the stent (Figure 13C). With the stent in the desired position, the outer balloon is inflated to implant the stent within the lesion (Figure 13D). Once the stent is expanded, both the outer and inner balloons are deflated and removed (Figure 13E). During the removal of the balloon catheter, the tip of the sheath is advanced over the deflated balloon (into stent) and the balloon catheter is slowly withdrawn into the sheath to prevent inadvertent displacement of the stent. A multi



Figure 11: Selected cine frames from angiograms of a three-month-old infant prior to (a), immediately following (b), and 1 year after (c) balloon angioplasty. Note that the coarcted aortic segment (arrow) improved markedly after angioplasty (b) which remains improved at follow-up (c). Ao, aorta; DAo, descending aorta; LV, left ventricle.



Figure 12: Bar graph of immediate and follow-up results of patients who had recoarctation following balloon angioplasty of aortic coarctation. Note significant (p < 0.001) drop in the gradient following balloon angioplasty (Pre, prior to vs. Post, immediately following). However, the gradient increases (p < 0.001) slightly at a mean follow-up to 14 mo (FU). After repeat balloon angioplasty (2^{nd} BA), the gradient decreased remarkably (P < 0.001); this residual gradient did not increase (P > 0.1) at further follow-up (2^{nd} FU). The gradients at second follow-up are significantly lower (p < 0.001) compared to those prior to initial and repeat balloon angioplasty. Mean + standard error of mean (SEM) are shown. N indicated the number of patients undergoing repeat balloon angioplasty.



Figure 13: Selected cine-angiographic/radiographic frames demonstrate stent deployment in aortic coarctation using a balloon-in-balloon catheter showing the position of an un-inflated stent (A), following inflation of the inner balloon (IB) (B), test angiogram after inflation of inner balloon (C), after outer balloon (OB) inflation (D) and after balloon deflation and withdrawal (E). White arrow in E demonstrates the expanded stent (St). The guide wire (GW) is positioned in the right subclavian artery in an attempt to keep the stent straight. Radio-opaque tip of the sheath (Sh) is seen. DAo, descending aorta.



Figure 14: Selected cine frames from an aortic arch (AA) cineangiogram in lateral view demonstrating severe coarctation (White arrow) with isthmic hypoplasia (A) in an adolescent which improved (B) LSA, left subclavian artery of stent (St).

track catheter [65] is positioned over the guide wire to the area just proximal to the stent and pressure pullback performed to document any residual gradient post stent followed by an angiogram (Figure 14). Additional stent dilatation with an appropriately sized low-pressure balloon is performed by some workers [66]; we try to avoid this step by initially selecting the outer balloon diameter of the BIB catheter to be equal to the intended diameter of the stent. Cefazolin (25 mg/kg) is given in the catheterization laboratory and repeated two more times every 6-8 hours. Aspirin is started at doses of 5-10 mg/kg/day for the next 6 weeks.

Immediate results: In the first series reported, Suarez de Lezo et al. [58] showed that peak systolic pressure gradient decreased from 43 ± 12 to 2 ± 3 mmHg and the ratio of isthmus/descending aortic diameter improved from 0.65 \pm 0.14 to 1 ± 0.08 following stent procedures. Similar results have been found in subsequent studies involving series with a larger number of patients. Increase in the diameter of the coarcted segment (Figure 14) and reduction in peak systolic pressure gradient across the coarctation were observed after stent placement. Stents are effective in post-surgical and post-balloon angioplasty recoarctation as

well as native coarctation. Improvement of the diameter of hypoplastic isthmus or transverse arch and exclusion of aneurysms has also been shown. In a more recent study, Forbes et al. [33] presented data of 217 pediatric patients who had stent placement for CoA. Immediately after the stent implantation, upper and lower extremity pressure gradient decreased to 4.9 ± 13.2 mmHg. At the time of discharge 81% of the patients had gradient ≤ 15 mm Hg. The complication rate was as low at 2.3% with 1.4% related to stent migration. Average length of stay in the hospital after the procedure was 2.4 days.

Intermediate follow up: At 5 year follow up of 77 patients, Forbes et al. [33] showed that upper and lower extremity gradient was 1.9 ± 13.7 mmHg. About 85% of patients had upper and lower extremity gradient ≤ 15 mmHg. 82% of patients had normal blood pressures and 31% of patients were on antihypertensive therapy at the time of follow up. There was 12.5% incidence of complications with aneurysm comprising 5.4%. 14.3% patients had evidence of reobstruction with 12.5% being mild, 1.8% moderate and no severe recoarctation. Mean coarctation/descending aorta ratio was found to be 0.8. Fortyfour out of 217 patients required re-intervention with 35 being planned procedures.

Covered stents



Figure 15: Bar graph depicts immediate results of balloon angioplasty and surgery for aortic coarctation; note significant (p<0.005) reduction in peak-to-peak gradient across the coarctation both after surgery and balloon angioplasty. This gradient reduction between the groups is not significant (p=0.28). Mean + standard error of mean (SEM) are shown. PRE, before; POST, after intervention.



Figure 16: Bar graph illustrating residual gradients at follow-up after initial surgery (SURG) and balloon angioplasty (BALL); these are similar (p>0.1). In both groups, repeat balloon angioplasty resulted in significant (p<0.001) reduction in coarctation gradients. Mean + standard error of mean (SEM) are shown. FU, follow-up; PRE, before; POST, after intervention.



Figure 17: Bar graph portraying morbidity and complication rates following surgery (SURG) and balloon angioplasty (BALL). Note that the duration of hospitalization (p<0.05), mechanical ventilation (p<0.01) and complication rate (p<0.05) are lower in the balloon group than surgical group. Mean + standard error of mean (SEM) are shown.

Experience in the use of covered stents to manage aortic coarctation is limited [67-74]. Different types of stents to treat aortic coarctation have been used and include Jostent grafts (Jomed International, Helsingborg, Sweden), C-P stents (NuMed Inc, Hopkinton, NY), AneuRx (Medtronic, Watford, UK), Advanta V12 (Atrium Medical, Hudson, NH) and others. Although these stents are available outside USA, none are, as of this date approved for clinical use by FDA. However, customizing and off-label use of available endoluminal grafts are feasible, when necessary [75]. General indications for covered stents are similar to those used for balloon angioplasty and standard stent. Specific indications for use of covered stents include postangioplasty aneurysm, tortuous aortic arch and isthmus, associated patent ductus arteriosus, prior surgical conduit, Takayasu arteritis and extremely narrow (subatretic) coarcted segment. When the assessed risk for development of aneurysm or dissection is high, a covered stent should be used. The results of the limited use of covered stents appear to be good [67-77]. Some of the stents can be expanded to only an 18-mm diameter. In addition, the stent shortens when expanded to larger diameters. Use of covered stents has another disadvantage in that the vessels that arise from the aorta are blocked. Aortic rupture remains an important, though an infrequent complication following primary stenting for aortic coarctation. Covered stents have been used to reduce this risk. However, aortic rupture has been reported even with covered stents [76]. Renarrowing of covered stents has been reported and data in limited number of patients suggest Covered Cheatham-Platinum stents can be re-expanded [77]. Based on the currently available data, the covered stents may be useful in highly selected patients with aortic coarctation.

Comparison of different treatment modalities

There are scanty data directly comparing surgical intervention with balloon angioplasty procedure. In 1993, Shaddy et al. [78] presented a prospective randomized study involving 36 patients, aged 3-10 years. The children were assigned to either balloon angioplasty (20 patients) or surgery (16 patients) and showed similar immediate pressure gradient relief in both groups. The complications of aneurysm formation and restenosis were higher in the balloon angioplasty group compared to surgical counterpart but the risks of neurologic complications were higher in the surgical group. They concluded that balloon angioplasty of coarctation of the aorta may provide an effective initial alternative to surgery in children and suggested that further long term studies following angioplasty.

In 1994, Rao et al. [79] compared efficacy and safety of balloon angioplasty with surgical correction in infants<3 months of age. They presented 29 infants undergoing intervention for aortic coarctation from 1982 to 1992. Fourteen infants underwent surgical repair while 15 had balloon angioplasty. The data [79] indicated that the degree of relief of aortic obstruction both immediate (Figure 15) and follow-up (Figure 16) and the frequency and effectiveness of reintervention (Figure 16) were similar in both groups. However, the morbidity and complications were lower with balloon than with surgical therapy (Figure 17). They suggested that the balloon angioplasty may be an acceptable alternative to surgery in the treatment of symptomatic aortic coarctation in infants<3 months of age [79].

Pooled data from senior author's study group and nine published reports of balloon angioplasty and surgical series derived from 11 reports were compared [22]. The balloon group is made up of 72 patients who had balloon angioplasty between 1982 and 1991. The surgical group is comprised of 607 patients operated on between 1979 and 1990. The prevalence of associated cardiac defects in the balloon group (43 of 61 [70%]) was similar (p> 0.1) to that in the surgical group (360 of 516 [70%]), assuring the comparability of the groups. The initial mortality rates were similar (p>0.1) as were the recoarctation rates (p>0.05). The late mortality was higher (p<0.01) in the surgical group. A similar comparison between surgical and balloon groups [80] suggested better outcome following surgery, but this study was flawed as stated elsewhere [81] since all balloon angioplasty papers were not included and the time periods during which intervention was performed was not similar.

The same group of children reviewed above [78] was followed for a mean of 10 to 11 years and results were compared between balloon angioplasty and surgery for native aortic Coarctation [82]; similar resting blood pressures, residual gradients across the coarctation, exercise performance, aortic arch anatomy by MRI angiography and re-intervention rates were found. However, a higher incidence of aneurysms and greater arm to leg blood pressure difference during exercise was found in the balloon than in the surgery group. The authors [82] concluded that surgery is preferable to balloon angioplasty in the treatment of CoA in children. While randomization is a virtue of this study, the study only involves 36 children with only 21 (58%) returning for reevaluation. In addition, other studies (in which larger numbers of balloon angioplasty procedures were evaluated at longterm follow-up) revealed 5% aneurysm formation. And, aneurysm formation was detected in surgical patients [23] as well. Consequently we recommended [11] multi-institutional randomized studies with a larger number of subjects, to resolve the issues brought out by this paper.

In 2003, Hernández-González et al. [83] compared the results of balloon angioplasty with surgical aortic resection in a randomized, multicenter study and found similar effectiveness in reducing coarctation pressure gradient and blood pressures with both treatment modalities. However, higher re-coarctation rates and persistence of arterial hypertension at follow-up were found in the balloon than in surgical group, although complications associated with surgery were more serious than with balloon therapy. The authors concluded that overall differences are not statistically significant.

In 2004, Walhout et al. [84] performed retrospective review and

comparison of surgical repair with balloon angioplasty for native coarctation in 46 patients and found similar immediate success with respect to decrease in pressure gradients and recoarctation rates and freedom from reintervention at follow-up. No aneurysms were seen in either group. They conclude that both surgical repair and balloon angioplasty for native coarctation result in low reintervention rates.

Recently Forbes et al. [33] presented comparison between surgical, balloon and stent therapy for children weighing ≥ 10 kg with up to 60 months of follow up. They noted that there was no significant difference in terms of re-coarctation in any of the 3 groups. The incidence of aneurysm was highest with balloon therapy and least with stent therapy. Stent group was noted to have highest number of follow up planned interventions. They showed that stent therapy has better outcomes compared to surgery and balloon therapy groups when used in appropriate sized patient [33].

Complications such as paraplegia [24,25] and paradoxical hypertension [26,27,85] are seen with significant frequency following surgical repair while such complications are either rare, or if present, very mild and inconsequential following balloon angioplasty. Aneurysms following balloon angioplasty [86,87], through are of concern and need longer follow up, such aneurysms are also seen with surgery [23]. Femoral artery occlusion rate following balloon angioplasty may be higher than that seen with surgical therapy [44]. However, vascular complications can occur in the left upper limbs following coarctation repair with subclavian flap aortoplasty and these complications include gangrene [22,29], reduction in the length and muscle mass of upper arm and forearm [30-32] and abnormal Doppler blood flow velocities in brachial arteries [32] suggesting potential for symptoms of ischemia.

Based on the above and other reviews [7,11,21,22,43,88-90], it appears that effectiveness of balloon angioplasty is comparable to surgery, mortality rates are similar (and are probably related to the associated cardiac defects, not related to type of intervention performed) and morbidity and complication rates are lower with balloon than with surgical therapy and it may be suggested that balloon angioplasty is an effective alternative to surgery for relief of aortic coarctation. In older children, adolescents and adults, stent therapy has shown better outcomes and complication profile compared to other treatment options.

Therapy selection

Based on an extensive review of the literature and personal experience of the senior author for a period in excess of 25 years, some generalizations with regard to management of aortic coarctation may be made:

a. Children older than 6 months with discrete native coarctation are candidates for balloon angioplasty. There is a reasonable consensus on this issue among most cardiologists. Long-segment coarctations or those associated with significant isthmic hypoplasia are good candidates for stent placement, especially in older children, adolescents and adults.

b. Recurrent coarctation following previous balloon angioplasty may be treated with repeat balloon angioplasty [54]; others prefer surgery [91]. If the recoarcted segment is long, surgical treatment in young children and stents in older children is appropriate.

c. Treatment of neonatal and infant (< six months) coarctation

is perhaps the most controversial issue. Many centers prefer surgical intervention whereas a few centers opt for balloon angioplasty. We believe that balloon angioplasty in neonates and young infants has been very useful in critically ill babies, particularly in those in whom avoidance of anesthesia or aortic cross-clamping required for surgery is beneficial in the overall management. Such special circumstances include infants with shock-like syndrome [48], severe myocardial dysfunction and hypertensive cardiomyopathy [92], prior spontaneous cerebral hemorrhage [88], severely compromised ventricular function [90] and biliary atresia awaiting liver transplantation [88]. In summary, balloon angioplasty is useful in the management of extremely ill neonates and infants with severe Coarctation [7,48,88,90,92,93]; in this subset of patients and has a significant advantage over surgical approach.

d. Although we have not discussed the management of postsurgical recoarctations [94] in this review, there is general agreement that balloon angioplasty is the treatment of choice for treatment of these aortic recoarctations

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