# Aortic Coarctation/Arch Hypoplasia Repair: How Small Is Too Small

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Aortic coarctation/arch hypoplasia is a relatively common congenital heart disease that leads to severe cardiovascular complications if left untreated. During the modern era, the mortality of the primary surgical repair is very low but the long-term issues, such as recurrent coarctation/arch reobstruction and hypertension, are still significant challenges. The former is related to the surgical repair performed particularly in the management of the smallish distal aortic arch, and for the latter, despite the "successful" repair of the aortic coarctation, the intrinsic vascular anomaly remains a significant long-term morbidity.

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**Central Message**

Respect the natural profile of the aortic arch at surgery for coarctation of the aorta and hypoplastic arch for the long-term benefits.

Aortic coarctation (CoA) accounts for 5–8% of all congenital heart diseases with an incidence of 4/10,000 live births [1,2]. If left untreated, the condition will eventually lead to death caused by systemic hypertension, congestive heart failure, cerebral hemorrhage, infective endocarditis, or aortic dissection [3]. The first successful repair of aortic coarctation was performed in 1944 [4] when Crafoord and Nylin operated on 2 patients using an interposition graft technique in Sweden. Ever since and three-fourths of a century later, many surgical techniques have been introduced to repair the coarctation, but there are still short- and long-term issues related to the operation. Notably, recoarctation and late hypertension have been known to be the major challenge.

## ADDRESSING THE PATHOPHYSIOLOGICAL CHANGES

The coarctation causes perfusion defects after the ductus arteriosus closes. If CoA is not severe, the collateral flow will gradually compensate perfusion deficiency, and the clinical manifestations are mitigated. These patients typically develop problems later in adult life, such as persistent hypertension, intracranial hemorrhage, and heart failure due to left ventricular hypertrophy. The clinical picture is more prominent in neonates with inadequate collateral circulation after the ductal closure: the infant is suffering from the severe hypoperfusion of the lower body. By treating CoA surgically or percutaneously (if indicated), the complications as mentioned above can be avoided. The clinical criteria for CoA have been developed (Table 1).

Coarctation of the aorta is defined as a medial wall thickening of the aortic lumen, which leads to a localized narrowing. As the CoA is composed of abnormal histology of the arterial wall, it is considered as an arteriopathy because it is in association with the long-term cardiovascular pathologies. The narrowing is usually located juxtaductally, that is, opposite to the insertion of the patent ductus arteriosus [5]. The underlying pathophysiology is not clearly understood, but several theories have been presented: abnormal fetal blood flow, excessive distribution of duct-like tissue around the aortic isthmus, or unusual cellular migration patterns during the arch development [6,7]. These theories have been supported in clinical

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**Table 1** The Definition of the Aortic Coarctation

| Group | Characteristics |
|---|---|
| Infants | The arch/isthmus morphology and ductal flow |
| Children | Arm-to-leg blood pressure gradient >20 mm Hg |
| | $V_{max}$ > 2.5 m/s in echocardiography |
| | Weak femoral pulses |
| | Left ventricular hypertrophy |
| | Impaired LV function with 2D narrowing |
| | CoA to descending aortic diameter <0.7 |

CoA, coarctation of the aorta; LV, left ventricle; $V_{max}$, maximum velocity.

studies as it seems that the arterial reactivity is severely impaired also in normotensive young adults after CoA repair in childhood [8]. This may indicate that the structural and functional changes relevant to the pathology may persist, despite the corrective repair of CoA. In addition, the aortic arch shape seems to play a role in the development of postoperative hypertension [9]. The angulated gothic-shaped arch especially appears to be related to the postoperative problems. Moreover, a recent study showed that the specific high and narrowed aortic arch shape features might compromise the left ventricular function in the long term [10]. Not surprisingly, the preservation of the favorable shape in the CoA/arch repair is desirable, but unfortunately the means to address the nonfavorable shape after the initial repair are limited. As we know well, the primary operation often determines the destiny.

The current strategies for CoA repair include surgical repair, intravascular stent placement, and balloon angioplasty. Restenosis occurs after both endovascular and surgical repair [11,12], but the impact of specific surgical technique which is the focus of this paper on the recoarctation rate remains controversial. Some studies have suggested that the subclavian flap aortoplasty is superior concerning recoarctation rate [13,14], but some suggest extended end-to-end anastomosis [15,16].

## WHAT TO DO WITH HYPOPLASTIC DISTAL AORTIC ARCH?

There is still controversy regarding the definition of hypoplastic aortic arch [17]. Physiologically, a 50% reduction of the luminal dimension would have important flow dynamic effect. Other measures include the transverse arch dimension less than the left carotid artery, or the presence of a common brachiocephalic trunk, the distal arch is less than half the diameter of descending aorta at diaphragm, or the *z* scores. The last measure needs to be treated with care; a small change in actual dimension can cause a big change in the *z* scores especially at the extreme end of the Bell curve. However, we use the practical definition by Karl et al [18] that the dimension (mm) of transverse arch should be the baby weight +1, for example, in a 3 kg baby, the transverse arch is acceptable at 4 mm.

If indicated, CoA with hypoplastic distal aortic arch can be dealt with concomitantly using a resection of the CoA and an extended end-to-end anastomosis augmenting the distal arch.

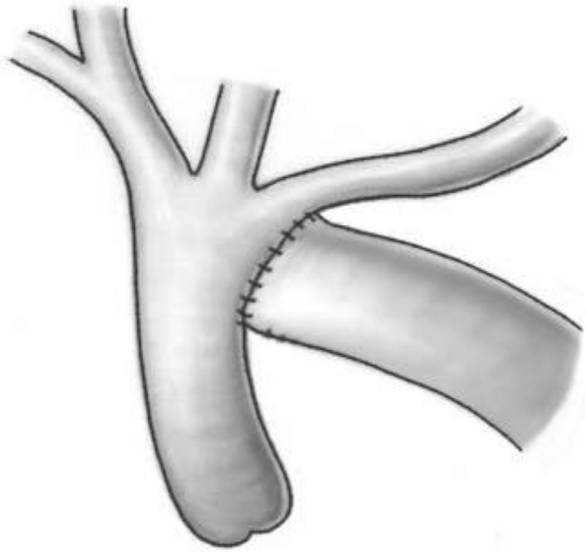

**Figure 1** A schematic of an end-to-side repair of coarctation/hypoplastic arch from sternotomy.

A curved vascular clamp can be applied proximally on the aortic arch (beware of innominate artery) with an incision on the under surface of the arch and toward distal ascending aorta, this would potentially create a tension-free anastomosis with a diameter as large as both the adjacent and distal aorta [19]. However, a recent article by Rakhra et al [20] suggested that patients undergoing an end-to-side repair from sternotomy had less arch reobstruction than those undergoing an extended end-to-end repair by thoracotomy (92% vs 61% freedom from reobstruction at 10 years, $P < 0.001$). The end-to-side approach from sternotomy or aortic arch advancement [21] is a popular approach to deal with the smallish aortic arch in the presence of CoA (Fig. 1). One technical aspect of the operation requires special attention. The arteriotomy on the aortic arch must not be performed too anterior, otherwise the end result will be a twisting effect which can be very significant if the ascending aorta is smallish resulting in a supravalvar stenosis. If the connection is too low, there is a possibility of compression of the left airway and pulmonary artery and vein. If done correctly, the advantages of this approach include a native tissue-to-tissue reconstruction with the potential for growth.

## THE GREAT ORMOND STREET HOSPITAL APPROACH

When it comes to the preferred strategy at Great Ormond Street Hospital (GOSH), in 2000−2006 end-to-end anastomosis was the most common repair. Until 2016, a rise of the extended end-to-end repair, patch aortoplasty repair, and arch augmentation repair was seen. Currently, we tend to follow the "Norwood type" of aortic arch repair for CoA with a hypoplastic arch. In this repair, the coarctation and ductal tissue are excised, creating a wide posterior tissue-to-tissue connection. A tailored pulmonary homograft patch with a curve is used to augment the hypoplastic arch/CoA with preservation of the

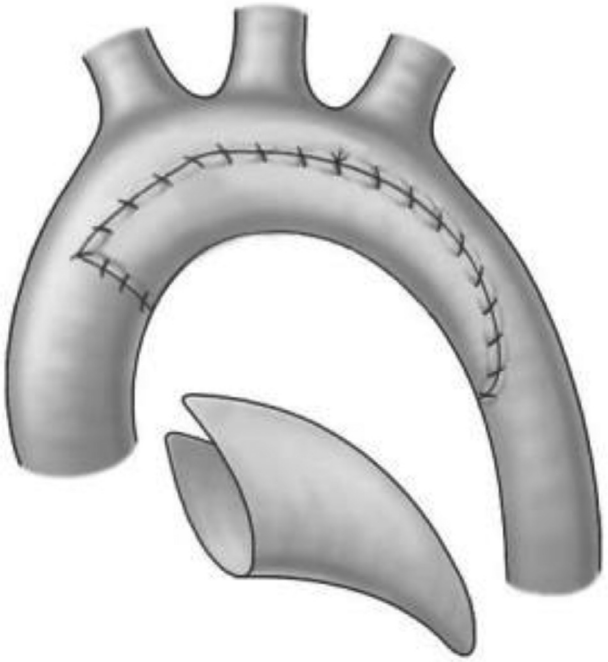

**Figure 2** A schematic of coarctation/hypoplastic arch repair using a tailored pulmonary homograft patch in the inner curve of the arch.

shape of the arch (Fig. 2). To put this into perspective, the change of the most common strategies is accompanied by an increased preference for sternotomy. From 2000 until 2016, 380 patients underwent primary surgical repair for CoA at GOSH. Of these patients, in the beginning only 7.8% underwent sternotomy for CoA repair. Over the years, this number has risen to 49.7%. The incidence of recoarctation and reintervention across time is being evaluated.

## CURRENT CHALLENGES IN COARCTATION REPAIR DURING THE NEONATAL PERIOD

The evolution of pediatric cardiac surgery is moving toward early physiological repair if possible, even in the smaller infants. However, the weight <2.5 kg at the time of heart surgery remains a significant risk factor for early mortality [22]. Accordingly, the prematurity (<35 gestational weeks) also has an almost 4-fold risk of infant mortality after congenital heart surgery [23]. Similarly, the CoA repair of very low-weight neonates appears to be related to the higher mortality and recoarctation rate [24]. Although the mortality of low birth weight children after cardiac surgery is still considerable, the outcome has improved over the decades [25]. The technical advancements and improvement in surgical techniques have contributed favorably to the peri- and postoperative care. During the current era, the preoperative management has developed remarkably, and the controversies remain whether it is beneficial or not to delay the definitive treatment to allow the low-weight neonate to grow [26].

**Table 2** Surgical Strategies for CoA/Arch Repair

| Type | Strategy |
|---|---|
| Simple CoA | Repair via left thoracotomy |
| Simple CoA with large VSD | Arch repair via median sternotomy, with closure of VSD<br>OR repair of CoA via left thoracotomy and PA banding |
| Arch hypoplasia with CoA | Repair via median sternotomy |
| Complex CoA and arch hypoplasia with TGA | Arterial switch and arch repair via median sternotomy |

CoA, coarctation of the aorta; PA, pulmonary artery; TGA, transposition of the great arteries; VSD, ventricular septal defect.

Already mentioned earlier, the trend is toward even smaller infant surgery. Therefore, we need to enhance the safety and durability of neonatal/infant coarctation surgery as much as possible. Our strategy to the neonatal CoA operation is determined by the anatomy of the distal aortic arch which probably eventually defines the long-term outcome. For isolated juxtaductal CoA, it is reasonable to perform a resection of CoA and an end-to-end anastomosis via a left thoracotomy. For a neonate or small infant with CoA and a hypoplastic distal arch, we undertake a sternotomy and a short period of deep hypothermic circulatory arrest for the surgical repair described earlier. Similarly, a one-stage repair via a sternotomy is chosen if the child has a CoA associated with an intracardiac defect, for example, a large ventricular septal defect. Our practical strategy is briefly demonstrated in Table 2.

## RECOARCTATION

The early operative risk of primary repair is very low nowadays. The main challenge of CoA surgery is the avoidance of recoarctation. This may contribute to the late postoperative hypertension, and the rate of recurrent CoA varied from 5% to 24% of CoA repaired patients [11,27–29]. In particular, the neonates (<30 days of age) and infants (<1 year of age) with hypoplastic arch segment are at the higher risk for recurrent CoA. The surgical strategy of dealing with recoarctation is outside the remit of this article, and understandably the residual arch anomaly determines the surgical and/or endovascular approach (Table 3).

## LATE SYSTEMIC HYPERTENSION

Relatively little long-term data surfaced until Clarkson et al reported rather worrying results in 1983 [30]. With a follow-up

**Table 3** Recoarctation Strategies

| Type | Strategy |
|---|---|
| >6 wk after surgery | Balloon dilatation |
| If resistant/insufficient to balloon dilatation | Surgical approach |
| >30 kg* | Stent angioplasty with covered stent |

*Both recoarctation and native coarctation.

of 10−28 years, the recoarctation rate was 6% and the likelihood of being alive without complication and with a normal blood pressure was 69% at 10 years, but only 20% at 25 years after the operation. In 1994, Gardiner et al [8], using a method of reactive hyperemia and an ultrasonic measurement of right brachial arteries, demonstrated that the arterial reactivity was significantly impaired in normotensive young adults 15 years after a successful repair of CoA. Some of these patients had the CoA repair in infancy/early childhood. The assumption that early surgery prevents vascular abnormalities with consequent cardiovascular complications may not necessarily apply in many cases. The persistence of structural and functional vascular changes relevant to the original pathology warrants long-term follow-up.

## CONCLUSION

With the passage of time, CoA has been well recognized as a "simple" surgical defect to repair but a complex disease to follow. The primary operation often determines the destiny, and repair of the CoA/hypoplastic distal arch during the neonatal period requires the understanding of the limitations included in the operation. Most importantly, the profile of the aortic arch surgery needs to be improved in the current era for the long-term benefits.

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