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CLINICAL PRACTICE GUIDELINES

The Society of Thoracic Surgeons Clinical Practice Guidelines on the Management of Neonates and Infants with Coarctation

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ABSTRACT

BACKGROUND Although coarctation of the aorta without concomitant intracardiac pathology is relatively common, there is lack of guidance regarding aspects of its management in neonates and infants.

METHODS A panel of experienced congenital cardiac surgeons, cardiologists, and intensivists was created, and key questions related to the management of isolated coarctation in neonates and infants were formed using the PICO (Patients/Population, Intervention, Comparison/Control, Outcome) Framework. A literature search was then performed for each question. Practice guidelines were developed with classification of recommendation and level of evidence using a modified Delphi method.

RESULTS For neonates and infants with isolated coarctation, surgery is indicated in the absence of obvious surgical contraindications. For patients with risk factors for surgery, medical management before intervention is reasonable. For those stable off prostaglandin E_1, the threshold for intervention remains unclear. Thoracotomy is indicated when arch hypoplasia is not present. Sternotomy is preferable when arch hypoplasia is present that cannot be adequately addressed through a thoracotomy. Sternotomy may also be considered in the presence of a bovine aortic arch. Antegrade cerebral perfusion may be reasonable when the repair is performed through a sternotomy. Extended end-to-end, arch advancement, and patch augmentation are all reasonable techniques.

CONCLUSIONS Surgery remains the standard of care for the management of isolated coarctation in neonates and infants. Depending on degree and location, arch hypoplasia may require a sternotomy approach as opposed to a thoracotomy approach. Significant opportunities remain to better delineate management in these patients.

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Coarctation of the aorta is commonly encountered, comprising 4% to 5% of congenital heart disease[1] and the second most common congenital heart disease requiring

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Abbreviations and Acronyms

CHSD = Congenital Heart Surgery Database
COR = classification of recommendation
DHCA = deep hypothermia circulatory arrest
LCCA = left common carotid artery
LOE = level of evidence
MRI = magnetic resonance imaging
PICO = Patients/Population, Intervention, Comparison/Control, Outcome
PTFE = polytetrafluoroethylene
SACP = selective antegrade cerebral perfusion
SCP = selective cerebral perfusion
STS = The Society of Thoracic Surgeons
VSD = ventricular septal defect

neonatal intervention.[2] Management of coarctation has changed considerably since the first operations in the 1940s,[3,4] with prostaglandins, introduced in the 1970s, allowing neonates with critical coarctation to be stabilized before surgical repair.[5] Surgical techniques have also evolved to emphasize autogenous tissue-to-tissue anastomoses that could also be extended to address coexisting transverse aortic arch hypoplasia. The surgical approach varies between thoracotomy and sternotomy, usually driven by coexistent transverse arch hypoplasia and/or concomitant intracardiac lesions.

Over the years, mortality has decreased. The Society of Thoracic Surgeons (STS) Congenital Heart Surgery Database (CHSD) reported an operative mortality of 0.98% for off-bypass repair of coarctation from July 1, 2016, to June 30, 2020.[6] Nevertheless, important short-term and long-term morbidities are still prevalent. Recoarctation, especially for neonatal repair, is a vexing issue, and long-term systemic hypertension is another concern. Ideally, surgical correction for coarctation would include a single operation with minimal perioperative complications that results in no residual long-term obstruction, no need for reintervention, and no need for antihypertensive medications.

Although coarctation is commonly encountered, there is no consensus regarding the optimum management of isolated coarctation in neonates and infants. The heterogeneity in phenotype has been increasingly recognized, including variable severity of associated arch hypoplasia, arch anomalies, such as bovine arch, as well as concomitant lesions such as ventricular septal defect (VSD). Such heterogeneity makes consensus in management challenging. Management strategies are also frequently impacted by institutional and surgeon-specific expertise and algorithms.

Given the prevalence of coarctation and the lack of consensus about its management, the STS Congenital Heart Surgery Task Force on Evidence Based Surgery saw isolated coarctation as an entity ripe with opportunity to examine the available literature and provide curated information and guidance regarding management. For the purposes of this practice guideline, isolated coarctation refers to coarctation of the aorta that may have associated arch hypoplasia, but without other congenital heart defects, such as VSDs or other major anomalies.

We recognize the limitations regarding available data, including the lack of randomized trials, the frequent implicit selection bias within single-institutional case series for techniques (based on the previously mentioned provider preferences and heterogeneous definitions), and inconsistent follow-up. Recommendations should be taken in the context of the individual patient as well as the expertise of a given surgeon and institution. Guidance regarding management will and should evolve with time as more data are reported.

PATIENTS AND METHODS

This project was performed under the auspices of the STS Workforce on Evidence Based Surgery and its Task Force on Congenital Heart Surgery. The multidisciplinary Task Force identified key questions related to the care of these patients using the PICO (Patients/Population, Intervention, Comparison/Control, Outcome) Framework. A literature search of PubMed was performed using key words related to each PICO question. The search was limited to studies published in the English language, focusing on papers published in 2000 onwards. Studies including patients with concomitant VSD or other lesions, as well as studies related to arch augmentation in other clinical settings, were reviewed for applicability. Studies including nonprimary data, such as meta-analyses, were not used for forming recommendations, but were used within the development of the document, with meta-analyses used in part for determining level of evidence (LOE).

Clinical practice guidelines with classification of recommendation (COR) and LOE were developed using a modified Delphi method. To consider the statement having reached consensus, 80% of the members must have voted, with 75% of the members who voted to agree.[7] COR and LOE were determined based on guidance provided by the American College of Cardiology and the American Heart Association (Figure 1).[8]

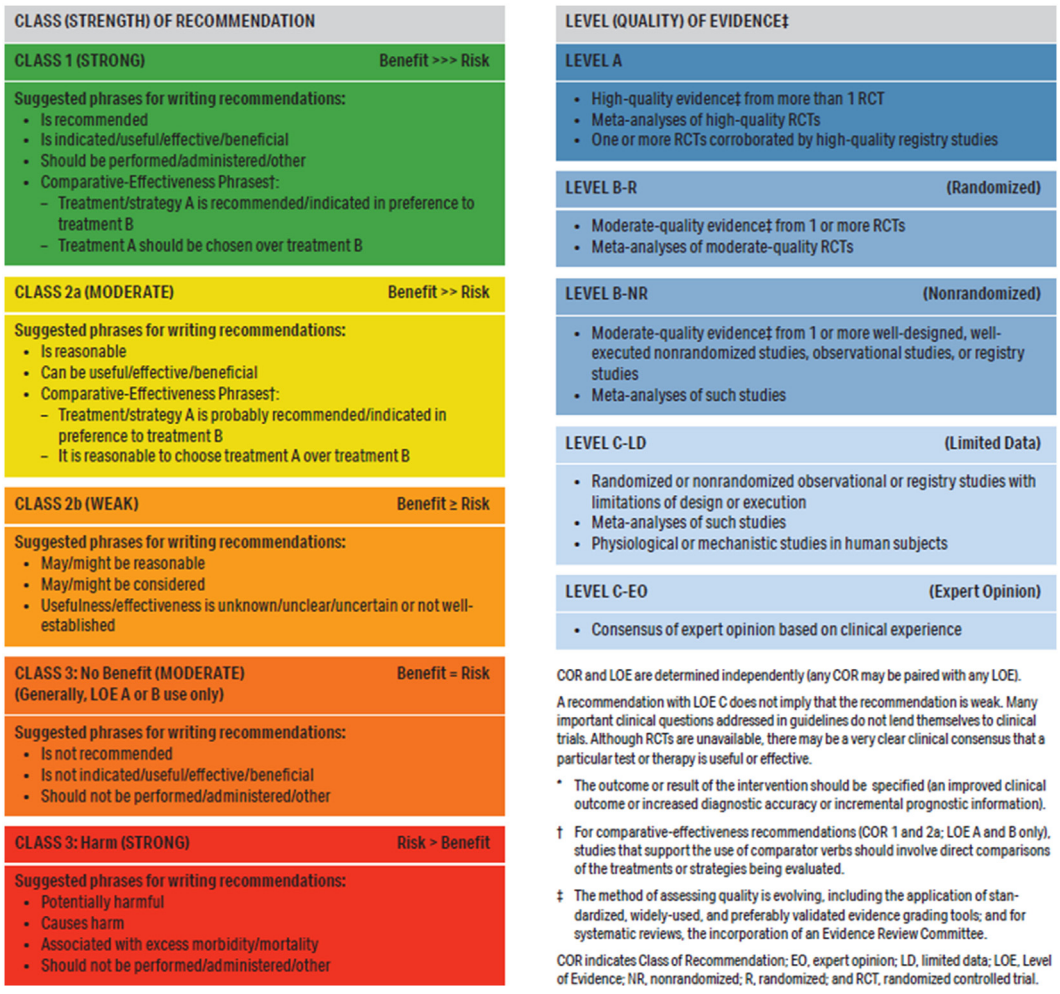

FIGURE 1 Classification of recommendation and level of evidence. Reproduced from Isselbacher and colleagues[8] with permission from the American College of Cardiology Foundation.

LIMITATIONS AND BIAS. The data on management of isolated coarctation are limited on multiple levels. The vast majority of studies were performed at single institutions, with inconsistent definitions of diagnoses, procedures, and outcomes. Most studies are generally biased toward particular techniques. Centers and surgeons who routinely publish on such techniques have become "experts" in that technique, limiting knowledge about real-world outcomes and learning curves. Thresholds to use different techniques—or to intervene at all—differ by provider, complicating comparisons across studies.

Although we can learn about outcomes for different techniques by comparing large single-institutional case series, such comparisons have significant limitations and unclear applicability to a surgeon or center that might or might not have experience in a particular technique. Furthermore, long-term outcomes, including hypertension, health-related quality of life, and neurodevelopment, should eclipse the current focus on short-term outcomes and perioperative complications.

TERMINOLOGY, DEFINITIONS, AND IMAGING. Please see the Supplemental Material for details on terminology and definitions, including that of coarctation and arch hypoplasia, as well as imaging. Briefly, for the purposes of this practice guideline, a hypoplastic arch is defined as a z-score of the distal transverse arch diameter of < -2.0, where the distal transverse arch is the portion of the aorta between the left common carotid artery

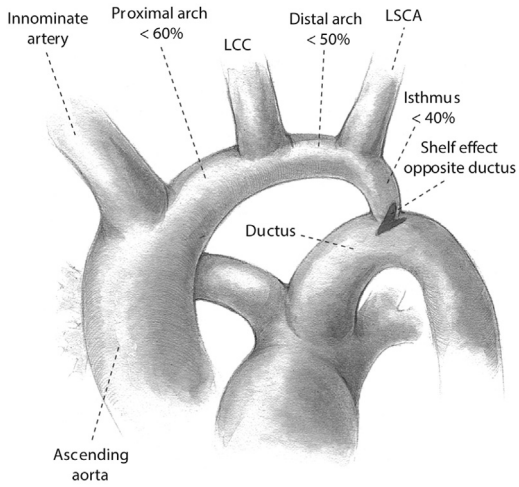

FIGURE 2 Segments of the aorta. (LCC, left common carotid; LSCA, left subclavian artery.) © 2014 From Comprehensive Surgical Management of Congenital Heart Disease (2nd ed.) by Richard A. Jonas.[9] Reproduced by permission of Taylor and Francis Group, LLC, a division of Informa plc.

(LCCA) and left subclavian artery (Figure 2).[9] Whenever possible and when available, discussions of data related to arch hypoplasia include details regarding how arch hypoplasia was defined within that study. The consensus statements use the term *arch hypoplasia* broadly, with specific discussion of evidence regarding degree of arch hypoplasia, and portion of the arch, contained within the text discussion following the statements.

RECOMMENDATIONS

1. **For neonates and infants with isolated coarctation, surgical intervention is recommended in the absence of obvious contraindications.**
   - **COR: I**
   - **LOE: B-NR**

Surgical repair is the treatment of choice for neonates and infants with isolated, primary (unrepaired; ie, native) coarctation. Balloon angioplasty or stent placement is reserved for the rare circumstances in which the patient is not considered a surgical candidate and medical management until surgery is recommended. Limited data are available regarding the results of percutaneous interventions for primary (native) neonatal and infant coarctation, in part because operative repair is considered the standard of care.

However, Sen and colleagues[10] recently analyzed 75 neonates and infants with native coarctation, of whom 28 underwent balloon angioplasty and the remaining 47 underwent surgical repair. They found that 29% in the balloon angioplasty group had significant coarctation on predischarge echocardiography. Further, among neonates, 64% who underwent balloon angioplasty required reintervention, and 17% who underwent surgical repair required reintervention, with a 6-month median follow-up. For patients >30 days, the reintervention rate was 29% in the balloon group and 42% in the surgical group. It is notable that the rate of reintervention for the surgical group in this study is higher than most modern published case series, including those including patients with arch hypoplasia,[11-15] with the meta-analysis by Dias and colleagues reporting a recoarctation rate of 14% in studies from 1989 to 2019.[16]

A study by Fiore and colleagues[17] from 2005 analyzed patients aged <40 days old who underwent balloon angioplasty (n = 23) or surgical repair (n = 34). With a mean follow-up of 3 years, 57% of the balloon angioplasty group required operative repair, 8 of 23 required a second balloon, and 3 of those had an aortic aneurysm. In the surgical arm, 18% had recoarctation. The surgical arm also demonstrated improved arch growth compared with the balloon group.

Similarly, the 2013 study by Chiu and colleagues[18] examined 88 patients with coarctation aged <3 months old, with or without VSD, of whom 17 underwent balloon angioplasty and the remainder underwent surgical repair. The reintervention rate was 65% for the balloon group and 42% in the surgical group, and 29% of the balloon group required repeat reinterventions.

In summary, although there are rare clinical scenarios in which catheter-based intervention can be beneficial and potentially even lifesaving, those scenarios are generally limited to cases in which operative repair is considered too high risk and medical management also is not possible.

2. **For neonates and infants with isolated coarctation and prematurity, low weight, or other risk factors for surgical intervention, medical optimization before intervention or primary operation is reasonable.**
   - **COR: IIa**
   - **LOE: B-NR**

Select studies have demonstrated certain factors, such as weight and age, are risk factors for

adverse events with surgical repair of coarctation. However, these risk factors have not been shown consistently across studies, and no studies have demonstrated that among ductal-dependent newborns, prostaglandin infusion awaiting somatic growth and chronological age before surgical repair mitigates these risk factors specifically in patients with coarctation. Therefore, each surgeon and institution should consider these factors within the context of the specific clinical scenario, their experience, and results.

Weight has been shown to be associated with adverse events in patients undergoing coarctation repair in select studies. In a study by Costopoulos and colleagues[14] of 66 infants undergoing coarctation repair by sternotomy (62%) or thoracotomy (38%), the reintervention rate was 8% for those <2.5 kg and 5% for those >2.5 kg. Those <2.5 kg also had longer length of stay, more complications, and a greater incidence of hypertension. However, this study did not use multivariable analysis and was limited by type II error. A study by Gorbatykh and colleagues[19] of 114 patients found weight <3 kg was a risk factor for recoarctation, with an odds ratio of 2. A study by Truong and colleagues[20] of 87 infants with coarctation repaired by thoracotomy found lower birth weight was associated with recoarctation on univariable but not multivariable analysis, and similarly, a study by Soynov and colleagues[21] of 54 infants repaired through a thoracotomy found lower weight was associated with recoarctation.

In terms of age, a study by Lehnert and colleagues[22] of 353 patients with coarctation, with or without VSD, found age <15 days and prostaglandin infusion were the only independent risk factors for reintervention on multivariable analysis, but not weight <2.5 kg. Similarly, McElhinney and colleagues[23] reported on 103 infants undergoing coarctation repair by thoracotomy and found younger age was independently associated with reintervention on multivariable analysis. A multi-institutional study by Quaegebeur and colleagues[24] of 322 patients from 1994 found age at repair was independently associated with mortality on multivariable analysis.

Weight and prematurity have been repeatedly shown to be risk factors for adverse events in congenital heart surgery overall. In the 2008 analysis of STS data by Curzon and colleagues,[25] patients aged <90 days undergoing surgical repair who weighed <2.5 kg had significantly higher mortality, including patients with coarctation repair (7.1% vs 2.7%, $P < .01$). Costello and colleagues[26] examined the STS CHSD and found that early gestational age, even 37 to 38 weeks, was a risk factor for in-hospital mortality (odds ratio, 1.34) for those undergoing various congenital heart operations, with more complications and longer length of stay. Other studies have demonstrated similar findings. For example, Kalfa and associates[27] analyzed 766 undergoing cardiac surgery and found weight <2.5 kg yielded worse outcomes (operative mortality 11% vs 5%, $P = .007$), including for coarctation repair performed through a sternotomy (25% vs 0%, $P = .02$). Despite these results, Hickey and colleagues[28] demonstrated medical management and delaying surgery in patients with low weight (often also with prematurity) did not improve survival. Within this study, there were 11 patients of the 149 infants with isolated coarctation, but the study did not allow for analysis of individual congenital heart disease entities.

In the setting of contraindications for surgical coarctation repair, such as intraventricular hemorrhage in a patient requiring sternotomy, medical management allowing recovery before operative repair is recommended. In such scenarios, the decision-making is multidisciplinary in nature considering the risks of bypass and heparinization in light of recent neurologic injury. Other instances in which medical optimization before repair may be indicated include decreased left ventricular function, where prostaglandin can be used to restore ductal patency and allow the left heart function to recover before repair.

3. **For neonates and infants with isolated coarctation without associated arch hypoplasia, repair through a thoracotomy is indicated.**

- **COR: I**
- **LOE: C-EO**

Repair through a thoracotomy is standard of care for coarctation without associated arch hypoplasia and in the absence of intracardiac pathology. This repair technique presents certain key advantages compared with the sternotomy approach, including avoiding the potential adverse effects of bypass and heparinization on a neonatal and infant brain. In the current era, this approach has been shown to have low operative mortality and relatively low incidence of complications such as recurrent laryngeal nerve injury and chylothorax.[11,15,20,22,29] Recurrent or residual coarctation requiring reintervention varies,

depending on the study and the threshold for criteria, but would be expected to be low. For other complications, paraplegia is devastating and is thankfully rare in neonates and infants. When it occurs, it mainly is associated with inadequate collaterals in children and also with an aberrant subclavian. Monitoring of distal perfusion in children is recommended, and partial bypass should be used if distal perfusion is inadequate.[30]

4. **For neonates and infants with isolated coarctation and associated arch hypoplasia that cannot be adequately addressed through a thoracotomy, repair through a sternotomy is preferable.**

- **COR: IIa**
- **LOE: B-NR**

Arch hypoplasia is heterogeneous, varying in location(s) and degree; how this is imaged, quantified, and defined, has varied across studies and institutions. Data related to arch hypoplasia are discussed in more detail subsequently. Nevertheless, ideally, the operation for coarctation of the aorta with a hypoplastic aortic arch should result in normal caliber of the ascending aorta and all aortic arch segments, with no gradient between the proximal ascending aorta and descending aorta.

In coarctation repair through a thoracotomy, the vascular clamp is applied between the innominate artery and LCCA, allowing the aortotomy to extend on to the undersurface of the arch to opposite the origin of the LCCA. The clamp can be positioned abutting the innominate artery and angled, allowing some access to the aorta between the innominate artery and LCCA, but the thoracotomy approach does not allow more proximal augmentation without compromising brain blood flow. Application of the clamp between the innominate artery and LCCA can be challenging in patients with a bovine arch, where the clamp could compromise cerebral blood flow. The most common operation performed through a thoracotomy in the current era is the extended end-to-end repair. Although the extended end-to-end repair addresses distal arch hypoplasia, it might not restore the diameter of the proximal arch to normal.

In patients with long-segment distal arch hypoplasia, several approaches have been described to allow augmentation through a thoracotomy. The reverse subclavian flap allows augmentation of the arch segment distal to the LCCA while maintaining ductal patency, followed by coarctation resection with end-to-end repair.[31] This, however, requires ligation of the left subclavian artery, which is usually well-tolerated, but incurs some long-term risks, including decreased left upper arm length,[32] decreased length and circumference of the left forearm,[33] and decreased left arm muscle mass and grip strength compared with the right arm.[34] Additionally, isolated incidences of left arm ischemia[35] or dizziness due to subclavian steal syndrome,[36,37] both requiring surgery in adulthood, have been reported. Patch augmentation[38] and native tissue approximation[39] have also been described to address long-segment hypoplasia through a thoracotomy, but are similarly unable to address proximal arch hypoplasia between the innominate artery and LCCA.

Repair of coarctation and a hypoplastic arch through a sternotomy addresses arch hypoplasia at all segments, including hypoplasia proximal to the LCCA and those with arch branch anomalies. Studies comparing outcomes between thoracotomy and sternotomy approaches have often been limited by selection bias, with those undergoing sternotomy more likely to have higher incidence of arch hypoplasia, more proximal arch hypoplasia, and a higher incidence of associated intracardiac defects.[5,11,40-45]

Select studies of repair through a thoracotomy have demonstrated that a hypoplastic arch, variably defined within each study, is a risk factor for recoarctation. For example, a study by Hager and colleagues[46] of 191 infants with isolated coarctation undergoing repair almost exclusively through a thoracotomy (all except 3 patients) found a hypoplastic arch, as described by the surgeon but not with specification about location or extent, was associated with death or recoarctation with an odds ratio of 2.9. A study by McElhinney and colleagues[23] of 103 patients undergoing coarctation repair through a thoracotomy found smaller transverse arch size, particularly when indexed, was associated with recoarctation. It should be noted that recoarctation can occur at the site of the anastomosis or at the site of an unoperated on or operated on arch that failed to grow to normal size. This distinction is not usually specified in studies.

Other studies comparing sternotomy and thoracotomy approaches have found decreased risk of recoarctation with sternotomy. Rakhra and colleagues[44] reviewed 305 patients who

underwent coarctation and hypoplastic arch repair through a sternotomy (n = 74) or thoracotomy (n = 231) and found those with a sternotomy approach had less recoarctation (92% vs 61% freedom from recoarctation at 10 years, $P < .001$), along with a lower incidence of hypertension (14% vs 20%, $P = .4$).[44] In this study, arch hypoplasia was defined in the oldest era by reports of a "small" arch, in the intermediate era as arch diameter less than weight in kilograms +1, and most recently using transverse arch *z*-scores. Similarly, Sakurai and colleagues[45] reviewed 288 consecutive patients who underwent coarctation repair through a sternotomy (n = 51) or thoracotomy (n = 237) and found in patients with hypoplastic arch (defined as arch diameter less than the innominate artery diameter or arch diameter less than weight in kilograms +1), sternotomy had a lower reintervention rate (4% vs 13%, $P < .001$). These studies suffer from significant selection bias, but in a manner such that sternotomy patients would be at higher risk of recoarctation.

Studies have attempted to determine the ideal threshold for transverse arch hypoplasia at which repair should be performed through a sternotomy as opposed to a thoracotomy. Gropler and colleagues[11] studied 251 patients, with 62% having transverse arch hypoplasia defined as *z*-score of < -2. The repair in 91% was through a thoracotomy and on analysis, the cut point predicting repair through a sternotomy was a proximal transverse arch *z*-score of < -4.1 or distal transverse arch *z*-score of < -2.8. This cut point, however, only defines the threshold by which the surgeons felt a sternotomy would be better, and excellent results were obtained with this threshold. The data do not provide information about whether a transverse *z*-score of < -2.8 can be approached through a thoracotomy with good outcomes. Tulzer and colleagues[12] performed a similar analysis of 183 patients, all of whom had arch hypoplasia, defined as a proximal transverse arch *z*-score of < -2.0. Thoracotomy was a risk factor for reintervention with a cut-off *z*-score of −4.6. Kotani and colleagues,[15] based on their analysis of 140 patients, recommended thoracotomy if the proximal transverse arch *z*-score was > -6.0.

However, others performing similar analyses have not been able to find an association with various indicators of arch hypoplasia and recoarctation. Truong and colleagues,[20] in examining 84 patients with isolated coarctation repaired through a thoracotomy, analyzed *z*-scores of various portions of the arch and did not find differences in the *z*-scores between those who developed recoarctation and those who did not.

Similarly, Ramachandran and colleagues[29] analyzed 102 patients repaired with various techniques through a thoracotomy and compared those with "good outcomes" (no hypertension, no antihypertensive medications, blood pressure differential <15 mm Hg, echocardiographic gradient <20 mm Hg, and no reintervention) vs those who did not meet those criteria and found similar median proximal arch *z*-scores between the 2 groups. No significant predictor of "good outcomes" could be found, although a larger transverse arch dimension tended to be associated with success ($P = .06$).

Callahan and colleagues[47] similarly analyzed 153 patients who underwent repair through a thoracotomy and used a variety of different criteria for categorizing proximal arch hypoplasia, but none were associated with reintervention. A recent study by Minotti and colleagues[48] analyzing 218 neonates with coarctation, 39% with complex congenital heart disease and 47% with hypoplastic proximal and/or distal arch (defined as a *z*-score of < -3.0) also did not find a cut point associated with reintervention. Other studies that have analyzed risk factors for reintervention have also not found arch hypoplasia (defined differently depending on the study) to be associated with recoarctation.[19,21,22,49-52]

Apart from the few aforementioned studies, the incidence of late hypertension is not well reported. Similar to the association between hypertension and recoarctation, there is also a correlation between residual arch hypoplasia and hypertension in those with inadequate catch-up growth after thoracotomy. Recent advancements in 3-dimensional imaging and flow dynamics studies have suggested some association between the shape of the reconstructed arch and flow patterns and the subsequent impact on late hypertension, left ventricular systolic and diastolic function, and exercise performance.[53-55] These changes were independent of the presence of recoarctation. More research is needed to correlate the type of operation with the eventual shape of the arch, to assess the effect of patch material and other modifiable surgical factors on distensibility of the arch and flow patterns, and to associate these factors with late morbidity.

COMPLICATIONS RELATED TO OPERATIVE APPROACH. Operative approach is also informed by whether sternotomy is associated with more complications or longer recovery. On the basis of the limited available evidence, neither approach has demonstrated clear superiority with regard to mortality, global neurologic outcomes, incidence of recurrent nerve injury, or incidence of chylothorax, as will be discussed in detail. The current literature consists of mainly single-institutional studies and systematic reviews with varying outcomes of interest analyzed and no head-to-head comparisons of postoperative morbidity between the 2 surgical techniques. The current literature is further hindered by small sample sizes and heterogeneous definitions. As a result, limited inferences can be made.

Operative mortality is largely comparable between patients who undergo a sternotomy or thoracotomy approach.[5,11,40-45]

The incidence of a neurologic catastrophe appears to be very small, and overall, neurologic outcomes seem to be similar. Studies have used varying definitions of what constituted a seizure and/or a stroke, and it cannot be clearly extrapolated whether these patients had additional corroborative investigations (ie, electroencephalogram and/or magnetic resonance imaging [MRI]).

Backer and colleagues[56] in 1998 reported on 55 patients who underwent resection with end-to-end anastomosis, including 34 through a thoracotomy and 20 through a sternotomy with circulatory arrest. Seizures occurred in 3 patients in the sternotomy group, whereas no seizures occurred in the thoracotomy group. Kaushal and colleagues[57] updated the Chicago experience, looking at an additional 156 patients (201 in total) who underwent resection and end-to-end anastomosis, of which 44 had sternotomy with circulatory arrest. Three sternotomy and no thoracotomy patients had seizures, though again these results did not reach statistical significance. A study by Gray and colleagues[58] analyzed 62 patients with coarctation and proximal arch hypoplasia who were repaired through a sternotomy with circulatory arrest in all except 2 patients. One patient had a seizure, and none had other neurologic complications.

Data are also inconsistent with regard to the prevalence of injury to the recurrent laryngeal or the phrenic nerve. Practices vary in the threshold to check for these injuries, as well as how the diagnosis is defined and confirmed. Frequently, when a study reports an injury, there is paucity of information on how this was diagnosed, the threshold for testing, and importantly, whether this injury resolved with time or required further intervention. Studies have shown that vocal cord dysfunction, in particular, is underdiagnosed without routine surveillance.[13]

Recurrent laryngeal nerve injury appears to be quite low in the thoracotomy approach, whereas it may be higher in the sternotomy approach, particularly when surveillance testing is used. Wright and colleagues[59] reviewed 83 patients with coarctation, 72 of whom were repaired through a thoracotomy, and found 1 patient with vocal cord dysfunction. Similarly, Kaushal and colleagues[57] analyzed 201 patients, 78% repaired through a thoracotomy, and reported a 3% incidence of recurrent laryngeal nerve paresis.

In contrast, studies of patients repaired through a sternotomy report an incidence of 4% to 60%. Costopoulos and colleagues[14] reviewed coarctation repair in 66 patients, including 51 through a median sternotomy, and found 3 patients had vocal cord injury, all in the sternotomy group (8% of that group). Pham and colleagues,[60] analyzed 155 patients undergoing Norwood and arch reconstruction (n = 55) and found 58% had abnormal vocal cord motion that was not different between the Norwood and arch repair groups. Pourmoghadam and colleagues[61] studied 101 neonates undergoing Norwood and arch repair (n = 26) and found 61% of arch reconstruction patients had vocal cord dysfunction, which was higher than Norwood patients (41%). At the last follow-up, mean of 11 months, vocal cord function had recovered in 86% of the arch repair patients. Mery and colleagues[13] investigated 275 infants undergoing arch advancement. Before 2007, routine laryngoscopy was not performed, and vocal cord dysfunction was diagnosed in 4%. However, after routine laryngoscopy surveillance was instituted, 38% of the patients had vocal cord dysfunction (20 had paresis and 16 paralysis). Of these, only 1 patient had clinical evidence of persistent vocal cord dysfunction at the last follow-up (median of 6 years).

A study by Ungerleider and colleagues[5] using STS CHSD data to examine coarctation repair found the overall incidence of recurrent laryngeal nerve injury was 3.5% in those with and without additional repairs and 1.6% in those with isolated coarctation repair. The isolated coarctation group was repaired at a median age of 32 days, with a variety of techniques: 56% extended end-to-end, 33% end-to-end, 4% patch

aortoplasty, 3% interposition graft, and 3% subclavian flap, and 86% were performed without bypass.[5] However, as discussed previously, recurrent laryngeal nerve injury is thought to be underdiagnosed in the absence of surveillance. The incidence of phrenic nerve injury was 0.9% across all groups and was 0.4% for the isolated coarctation group.[5]

Dysphagia and the need for a gastrostomy tube may also be higher in sternotomy than in thoracotomy. Costopoulos and colleagues[14] analyzed 66 patients, including 51 repaired through a median sternotomy, and found 5 patients needed gastrostomy tube placement, 1 (7%) in the thoracotomy group and 4 (11%) in the sternotomy group, although the difference did not meet statistical significance. This may relate to the concern for vagal nerve injury during arch dissection from the front, similar to the Norwood procedure. In the study by Pham and colleagues[60] of 155 patients, including 55 undergoing arch reconstruction, vocal cord paralysis occurred in 69% of arch patients, and 16% of those patients required gastrostomy tube placement.

Chylothorax is relatively uncommon and not different between techniques. A study by Backer and colleagues[56] compared thoracotomy and sternotomy patients after resection and end-to-end anastomosis. Chylothorax occurred in 2 of the 20 patients (10%) in the sternotomy group and in none of the 35 patients in the thoracotomy group. Costopoulous and colleagues[14] studied 66 patients, and chylothorax developed in 2 patients (13%) in the thoracotomy group and in 3 patients (8%) in the sternotomy group. In a study of 83 patients by Wright and colleagues,[59] of whom 72 patients underwent thoracotomy, chylothorax occurred in 2 patients (3%) in the thoracotomy group and in none in the sternotomy group, although the sternotomy group was admittedly small. Other studies have found the incidence of chylothorax is relatively low and not notably different between approach.[29,39,43,57] The study by Ungerleider and colleagues[5] using STS data to examine patients undergoing coarctation discussed previously, found the overall incidence of chylothorax was 3.3%, with 2.1% in the isolated coarctation group. These data did not provide information on the incidence relative to surgical approach.

The length of recovery is rarely described in existing studies; sternotomy may be associated with longer recovery. Although this might be explained by the effect of cardiopulmonary bypass and incidence of delayed sternal closure, the comparison is similarly contaminated by the fact that the infants who underwent sternotomy vs thoracotomy were often not comparable.[5]

RESIDUAL ARCH HYPOPLASIA. A remaining question in the setting of coarctation and hypoplastic arch is what is the fate of residual arch hypoplasia for patients in whom a thoracotomy approach is taken and whether the thoracotomy approach exposes these patients to higher need for early or late reintervention, persistent gradient, and subsequent hypertension or ventricular dysfunction, or diminished late survival. Studies analyzing the fate of the transverse arch in such patients demonstrate conflicting data regarding the reliability of arch growth after isolated coarctation repair.

Some theorize the residual hypoplastic arch will be stimulated to grow in response to increased blood flow after relief of distal obstruction, as described in the "hemodynamic molding theory."[62] Several clinical studies corroborate this. In a review of 17 patients with a "small arch" who underwent coarctation repair through a subclavian flap angioplasty, Myers and colleagues[62] observed growth of the transverse arch and found that arch growth was more pronounced in younger patients (aged <1 month) compared with older children (1 month-1 year). Siewers and colleagues[49] analyzed 33 patients with transverse arch hypoplasia who underwent subclavian flap angioplasty or classic coarctation resection with end-to-end anastomosis. They observed significant transverse arch growth in all patients without any attempt to enlarge the transverse arch. Brouwer and colleagues[63] analyzed the evolution of the aortic arch *z*-score on echocardiograms in 15 infants undergoing resection and end-to-end anastomosis. Eight of these infants had a hypoplastic arch (mean *z*-score, −7.14), and at the 6-month follow-up, the arch *z*-score had increased to −1.08, despite no attempt at arch enlargement.

More recent studies by Karamlou and colleagues[50] (2009) and Kotani and colleagues[15] (2014) further support this notion. Karamlou and colleagues[50] analyzed the echoes of 36 neonates who underwent coarctation repair using a variety of repair techniques over a 9-month follow-up, and the transverse arch *z*-score increased in all patients. Neonates with the smallest transverse arch *z*-scores had accelerated arch and isthmic growth trajectories. Importantly, extended end-to-end anastomosis, compared

with other surgical techniques, was associated with increased transverse arch growth. This study did not specifically look at *proximal* arch growth, so some of the observed transverse arch growth potentially was related to surgical augmentation of the arch in certain patients (ie, extended end-to-end anastomosis).

Kotani and colleagues[15] assessed the proximal aortic arch after extended end-to-end repair in 140 patients. Follow-up echocardiography in those with arch hypoplasia (n = 80) showed catch-up growth of the proximal arch, with the *z*-score improving from −2.58 at discharge to −1.17 at a median follow-up of 18 months ($P = .002$). Notably, multivariable analysis demonstrated a lower proximal arch *z*-score at discharge was a predictor of poor proximal arch growth. So, although arch growth was observed, there are likely limits to how hypoplastic an arch can be and still demonstrate reliable growth.

Although these studies suggest that arch growth may occur if the hypoplastic arch is not addressed at the time of repair, other data challenge the reliability of this phenomenon. Liu and colleagues[64] studied proximal vs distal arch growth in 20 neonates who underwent coarctation repair through a left thoracotomy. They did observe sustained growth of the distal transverse arch, evidenced by normal *z*-scores of the distal arch, but minimal proximal arch growth, with 35% (7 of 20) having a proximal transverse arch *z*-score of < -2 (mean follow-up, 9.5 years).

The implications of residual arch hypoplasia may be significant. Jahangiri and colleagues[65] found persistent arch hypoplasia after subclavian flap angioplasty was a risk factor for mortality and recoarctation, even though coarctation recurrence was noted to be more distally, in the region of ductal tissue. Weber and colleagues[66] studied 28 patients who had undergone a "good" coarctation repair (defined by normal blood pressure and absence of blood pressure gradient at rest after isolated coarctation repair without arch augmentation). With exercise or provocative testing, systolic hypertension and a significant arch gradient developed in 8 patients (29%). Aortography demonstrated transverse arch hypoplasia in these 8 patients compared with controls.

Residual gradient has been associated with recoarctation. Truong and colleagues,[20] analyzing 87 infants with coarctation repaired through a thoracotomy, found postoperative peak Doppler velocity was associated with recoarctation on multivariable analysis, with a hazard ratio of 1.13. In a study by Sen and colleagues[10] of 75 infants with native coarctation, of which 28 underwent balloon angioplasty and the remaining 47 underwent surgical repair, the only factor found on univariable analysis to be associated with recoarctation was the gradient on the predischarge echocardiogram. A study by Adamson and colleagues[52] of 74 patients with biventricular circulation and coarctation who underwent repair using a variety of techniques similarly found the systolic gradient on discharge was associated with recoarctation on univariate analysis. These studies did not differentiate whether the gradient was due to residual arch hypoplasia proximal to the repair or inadequate relief of obstruction at the repair site.

In summary, when a coarctation repair is performed through a left thoracotomy, every reasonable effort should be made to address the hypoplastic arch. If significant hypoplasia might be left behind, "catch-up growth" is not guaranteed, and consideration should be given to arch reconstruction through a sternotomy.

5. **For neonates and infants with isolated coarctation and bovine arch anatomy, repair through a sternotomy may be reasonable given the potential increased risk of recoarctation with bovine arch anatomy repaired through a thoracotomy.**
   - **COR: IIb**
   - **LOE: C-LD**

On the basis of the limited available data, neonates with isolated coarctation and bovine arch anatomy having coarctation repair performed through a thoracotomy can be reasonably expected to have higher rates of recoarctation/reintervention compared with those with isolated coarctation and typical arch anatomy repaired by thoracotomy. The current literature consists largely of single-institutional, retrospective studies. The one retrospective study demonstrating the influence of arch anatomy on reintervention comes from a small study (49 patients), yet represents the most rigorous assessment of bovine arch prevalence.[67] This limits the inferences that can be made regarding the relationship between bovine arch anatomy and reintervention rates.

Few investigations have assessed bovine arch anatomy as a variable that may impact the risk of

recoarctation. In the rare study that did assess arch anatomy, all but one[67] showed no significant impact on recoarctation. Yet, each of these negative studies only demonstrated a 3% to 5% prevalence of bovine arch anatomy in their cohorts.[23,51,68] These low rates of bovine arch prevalence are suspected to be secondary to retrospective review of medical records with inadequate attention to arch anatomy, as imaging and autopsy analysis specifically directed at ascertaining arch anatomy reveal bovine arch frequencies of 15% to 37%.[69-71]

A review of echocardiography reports in a study by Turek and colleagues[67] only identified 6.1% of patients as having bovine arch anatomy, whereas reads of the actual echocardiographic images of the same patients specifically looking at arch anatomy yielded a 28.6% prevalence. Meanwhile, in this same cohort, 28.6% of the patients identified by echocardiographic review to have a bovine arch had recurrent arch obstruction, as defined by a gradient of >20 mm Hg, compared with only 5.7% recoarctation for patients with a normal arch anatomy. All repairs were done through a thoracotomy with extended end-to-end anastomoses. This small study also shows through angiographic imaging measurements that the available length of aorta for clamping to facilitate repair is diminished in patients with bovine arches. Analysis of infant aortic arches as assessed by computed tomography further lends credence to the suggestion that bovine arches are shorter and could limit clamping during coarctation repair.[72]

Given the data related to the bovine arch having an apparent increased propensity for recoarctation, consideration can be made for repair through a sternotomy, although there are no data comparing the approaches in this subgroup.

Other limited data exist related to an aberrant subclavian and its impact on recurrence. In a study by Kaushal and colleagues[57] of 201 patients undergoing coarctation repair, an aberrant subclavian was a risk factor for recurrence on univariate analysis, and actually was the only risk factor for recoarctation identified in that study.

Another important aspect related to bovine arch anatomy is clamp placement. In this anatomy, there is potential to significantly impair cerebral blood flow with clamp placement, and this can make repair more challenging and impact the extent to which the arch can be addressed.

6. **For neonates and infants with isolated coarctation undergoing repair through a sternotomy, antegrade cerebral perfusion or limited-duration deep hypothermic circulatory arrest may be reasonable.**
   - **COR: IIb**
   - **LOE: B-R**

In neonates undergoing repair of coarctation, regional perfusion techniques, including selective antegrade cerebral perfusion (SACP/SCP) or dual aortic cannulation, might be considered given the existing literature leans toward equivocal or occasionally better neurologic outcomes when those techniques are used instead of deep hypothermic circulatory arrest (DHCA), as detailed subsequently. This recommendation is based on 4 studies: 1 randomized clinical trial,[73] 2 retrospective observational studies,[74,75] and 1 systematic review.[76] The recommendation should be taken in the context of the limitations of the studies it is based on, the sample sizes, the lack of mid- to long-term follow-up data related to neurologic outcomes, and variability in perfusion techniques. Further, inclusion in most of these studies was not restricted to isolated coarctation repair.

Kulyabin and colleagues[73] in 2020 reported the results of a prospective randomized trial of infants (most were neonates) who underwent surgical coarctation repair with arch hypoplasia. There were 45 patients randomized to DHCA, SCP, or dual aortic cannulation groups (15 per group). They found that only DHCA (with a mean duration of 23 minutes) had a high risk of neurologic injury compared with the other perfusion methods. Neurologic injury was defined as any new brain lesion after surgery that was visible on MRI. In patients who underwent DHCA, the risk of new MRI findings was the highest (80% vs 33% in both of the other groups, $P = .019$), with white matter injury being the most common type of lesion observed. Thus, continuous visceral perfusion strategies under mild and moderate hypothermia were associated with a reduced risk of new brain MRI findings.

In a retrospective review from 2015 by Kornilov and colleagues,[74] 62 patients, not strictly neonates, underwent coarctation repair. Patients requiring concomitant procedures were included, but hypoplastic left heart syndrome patients were excluded. Of these, 27 patients underwent DHCA and 35 underwent SACP. The odds ratio for neurologic complications, defined as any "new temporary or permanent focal or global neurological dysfunction after surgery,"

was significantly lower in the SACP group (6% of patients undergoing SACP had neurologic complications vs 31% of patients undergoing DHCA; odds ratio, 0.14 for SACP). A combination of computed tomographic imaging and clinical assessment was used to define neurologic complications. However, this study also showed a higher incidence of renal dysfunction in the SACP group (61% vs 19%; odds ratio, 6.49).

In another retrospective observational analysis by Uemura and colleagues,[75] 56 infants underwent reconstruction of interrupted aortic arch or coarctation of the aorta. These patients underwent total circulatory arrest (n = 23), perfusion of the carotid arteries while the descending aorta was cross-clamped (n = 21), or dual aortic cannulation (n = 12). There were no differences among the 3 groups in cerebral bleeding or seizures.

In 2012, Hirsch and colleagues[76] reported a systematic review on neuroprotective strategies for infants undergoing congenital cardiac surgery that assessed DHCA, regional cerebral perfusion, and low-flow cardiopulmonary bypass. The studies included were from 1990 to 2010, and all congenital cardiac operations were considered, with the exception of cardiac transplantation. They assessed perfusion strategy in 44 manuscripts: 2 had Class IIB LOE B, 6 had Class III (no benefit) LOE B, and 1 had Class III LOE C. In this analysis, they found none was superior to the others in neurologic outcomes.

Additional studies also compared different perfusion methods; however, outcomes were reported for the whole cohort of patients and not for coarctation repair only. For example, in a randomized trial by Algra and colleagues,[77] no difference was found in new cerebral lesions assessed by MRI between DHCA and SACP perfusion methods in neonates undergoing arch reconstruction/repair with associated congenital cardiac defects. Although the most commonly observed neurologic lesion was white matter injury, central infarctions were seen exclusively after SACP.

Theoretically, neurologic outcomes may be improved with a reduced duration of DHCA from shorter operative times or adjunctive perfusion techniques such as SACP. A retrospective observational study by Algra and colleagues[78] on neonates undergoing aortic arch repair with associated congenital cardiac defects found that a longer duration of DHCA was associated with longer intensive care unit length of stay, longer duration of mechanical ventilation, and decreased kidney function.

It is also important to note that some studies of SACP did not provide adequate perfusion, especially to the lower body.[79] As noted above, significant renal dysfunction has been noted with SACP as well as intraoperative lactate correlating with rectal temperature,[74] suggesting that the remainder of the body may not be adequately protected at the higher temperatures frequently used in SACP; DHCA provides additional protection in that regard. Although it seems logical that providing cerebral blood flow, in particular, should yield improved neurologic outcomes, conclusive data are lacking to date. Brain and body protection, however, solely relying on hypothermia in DHCA is time limited.

A recent study by Starr and colleagues[80] examined neuroprotection practices within the STS database for neonates undergoing cardiac surgery. Among those, 18% underwent arch repair, 24% Norwood/stage 1 procedure, and 23% arterial switch. The use of DHCA across procedures decreased during the study period from 2010 to 2019 from approximately 40% to 36%, and the mean nadir temperature increased from 23.9 to 25.6 °C. On analysis, after risk adjustment, temperature was not independently associated with adverse neurologic outcomes across procedures, but factors independently associated with major neurologic risk included younger age, Norwood/stage 1, longer bypass time, longer duration of DHCA, younger gestational age, noncardiac anomaly, and chromosomal anomaly. Specifically, among the aortic arch cohort, a composite neurologic event (seizure, stroke, persistent neurological deficit) occurred in 3.2%, and temperature was not associated with this outcome. Prematurity, chromosomal abnormality, cardiopulmonary bypass time, and lower annual hospital volume all were independently associated with neurologic outcome. Specific perfusion strategies were not analyzed in this study.

7. **For neonates and infants with isolated coarctation undergoing repair through a sternotomy, extended end-to-end, arch advancement (end-to-side reconstruction with ligation of isthmus), and patch augmentation are all reasonable techniques.**
   - **COR: IIa**
   - **LOE: B-NR**

Neonates with coarctation presenting for surgical repair through a sternotomy can reasonably undergo reconstruction with a variety of techniques with satisfactory results, including relatively low rates of reintervention and perioperative complications. Data largely consist of single-institutional case series with a bias toward one technique, with other techniques reserved for specific situations leading to dissimilar cohorts within the study. Data directly comparing techniques are very limited, particularly in similar populations. Although outcomes can be examined across techniques and institutions, limited conclusions can be drawn, and applicability to surgeons and institutions without experience with particular techniques is unknown. Therefore, the choice between direct anastomosis, such as extended end-to-end or arch advancement/end-to-side, and patch material will largely be governed by surgeon and institutional expertise, as well as by anatomy. Independent of specific technique, key objectives of the reconstruction include complete resolution of narrowing and minimizing potential complications such as recurrent laryngeal nerve injury and left main bronchus compression.

The major techniques used do have theoretical risks and benefits relative to one another. With an extended end-to-end technique, there is more extensive dissection compared with other techniques, risking recurrent laryngeal nerve injury and the potential for more tension on the repair, which is thought to relate in some cases to recoarctation. Additionally, significant proximal arch hypoplasia may be challenging to address with this technique. The technique has the advantage, however, of avoiding prosthetic material. The end-to-side or arch advancement technique also involves significant dissection and additionally puts the left main stem bronchus at risk for compression, although in experienced centers this complication is relatively low. This technique also has the advantage of avoiding prosthetic material. Patch augmentation requires less dissection risking the recurrent laryngeal nerve and does not have the same risks of tension on the anastomosis or left mainstem bronchus compression; however, there has been a wide variation of reported performance of materials, with some demonstrating distinctly worse performance than other materials.

With respect to extended end-to-end repair, studies show excellent results. Gropler and colleagues[11] reported on 251 patients with a median age at repair of 15 days and 9% performed through a sternotomy. Excellent results were obtained, with 0% mortality, a 2% rate of reintervention (1 of 2 catheter ballooning), and 18% incidence of hypertension, with a median follow-up of 5.4 years. On analysis, a proximal arch z-score of < -4.1 predicted sternotomy. Extended end-to-end repair has replaced end-to-end repair given the better outcomes with extended end-to-end repair.

However, not all studies have shown such good results with extended end-to-end anastomosis. For example, in a study by Tulzer and colleagues,[12] 183 neonates and infants with coarctation/hypoplastic arch, with or without VSD, with a median age of 15 days underwent repair through a thoracotomy in 111 and sternotomy in 72. Among the sternotomy patients, 41 had repair with extended end-to-end anastomosis and 30 end-to-side. They found a reintervention rate of 8%, and 10 of the 11 had undergone extended end-to-end through a thoracotomy. Similarly, a study by Wright and colleagues[59] of 83 infants with a median age of 21 days, with 11 undergoing repairs through a sternotomy, found a recoarctation rate of 8%.

The end-to-side technique, also called arch advancement, has had excellent results in experienced hands. In a study by Mery and colleagues,[13] 275 patients underwent arch advancement for a variety of conditions, including isolated coarctation (n = 29). The mortality was 3% (none in isolated coarctation group) and the incidence of left main-stem bronchus compression was 0.7%; however, recurrent laryngeal nerve injury was noted in 38%, with the vast majority recovering function. The median follow-up was 6 years, and the reintervention rate was 3%, occurring at a median of 5 months. Lee and colleagues[81] also showed good results with this technique. In their study of 170 patients undergoing end-to-side repair for interrupted arch (n = 95) or coarctation (n = 75) with accompanying VSD in 143 patients (and 93% undergoing single-stage repair), there were 4 early arch reinterventions, including 2 for bronchial compression. Bronchial compressing occurred in 8 total patients, including 3 noted after discharge. After a mean of 10 years, the reintervention rate was 22 patients (15%), including 15 (8%) arch reoperations. Of note, 14% had recurrent laryngeal nerve palsy. However, not all studies have shown such good results. In a study by Li and colleagues[82] discussed in more detail subsequently, the recoarctation rate was 51%, and the end-to-side

technique was a risk factor for recoarctation relative to patch angioplasty.

Patch augmentation has been used with a wide variety of materials, including Dacron (DuPont), polytetrafluoroethylene (PTFE), varieties of autologous and xenograft pericardium, autologous vascular tissues, and homograft. In some studies, patch augmentation was used in a limited subset of the study population with the surgeon's discretion, and often when other techniques were not favorable. Such studies limit meaningful comparison of outcomes given the patch cohort was inherently a more challenging substrate.

Nevertheless, some differences have been noted between different materials. Dacron, in particular, has been noted to have poor outcomes due to aneurysm formation and risk of rupture,[83,84] and although not commonly used in neonates, should nonetheless be avoided. A study by Cramer and colleagues[84] of 63 patients with a median age of 63 months, including some neonates, who underwent patch augmentation for primary repair of coarctation (92%) or recoarctation, found a 47% incidence of aneurysm formation, and 7 of those 29 died suddenly of rupture.

Limited data exist regarding the performance of CardioCel (tissue-engineered bovine pericardium, Ademedus Regen Pty Ltd), with 1 study directly comparing CardioCel and homograft for various types of arch reconstruction in neonates and infants.[85] Among 41 patients, including 7 isolated arch repairs, there was a 70% incidence of recoarctation requiring reintervention within the first year for CardioCel vs 23% in the homograft group.

PTFE is uncommonly used in neonatal primary coarctation repair, and evidence suggests that the rate of recurrence is high (50%) when used in patients aged <1 year,[86] but lower for patients >1 year. A study by Walhout and colleagues[87] of 262 children, 42% with isolated coarctation, and 158 patients undergoing repair through a sternotomy, compared PTFE (48%; mean age, 1 year) and resection and end-to-end (52%; mean age, 1.8 years). Recoarctation occurred in 21% with a median 12-year follow-up, which was not significantly different between the groups.

Limited data also exist for autologous pericardium. A study by Onalan and colleagues[88] examined 60 patients with a median age of 20 days undergoing extended patch aortoplasty with various types of pericardium for coarctation/arch hypoplasia in a diverse population of congenital heart disease, including 3 patients with isolated coarctation/arch hypoplasia. Mean follow-up was 22 months, and they found a 22% recoarctation rate, with no difference between glutaraldehyde-treated pericardium (n = 21), bovine pericardium (n = 19), and porcine pericardium (n = 20). A study by Roussin and colleagues[89] examined materials used for arch reconstruction in 51 neonates and young infants with interrupted arch (n = 28) or coarctation with VSD (n = 23), with pericardium as a patch material, but pericardium patients were grouped with the homograft patients for analysis. The pericardium/homograft group had a 28% recurrence rate with median follow-up of 29 months.

Other studies using pulmonary homograft material demonstrate better results, however, suggesting that its use is reasonable in experienced hands. In a study by Tchervenkov and colleagues[90] of 40 patients undergoing repair of coarctation along with other procedures using homograft augmentation in 36 and extended end-to-end repair in 4 patients, they found no recoarctation at a mean of 36 months of follow-up among those without left-sided disease. Similarly, in a study by Sakurai and colleagues[45] of 237 patients undergoing coarctation repair with a variety of techniques, those who had patch repair (n = 22, usually pulmonary homograft, rarely bovine pericardium) demonstrated no recoarctation or aneurysm formation, whereas there was an 11% reintervention rate overall with a follow-up of 7 years. A study by Gray and colleagues[58] of 62 patients with a median age of 10 days undergoing repair of isolated coarctation by homograft aortoplasty (92%) found a reintervention rate of 10% with a median 41-month follow-up.

However, other centers have not shown such promising results. Whiteside and colleagues[91] reported on 101 patients with a median age of 8 days who underwent Norwood-type arch reconstruction using a homograft of SynerGraft (CryoLife) in the setting of single-ventricle and biventricular physiology. They found a recurrence rate of 18% in the biventricular group, with mean time to recurrence of 0.5 years.

Small case series have been published using autologous vascular materials, such as autologous pulmonary artery. In the previously mentioned study by Roussin and colleagues,[89] pulmonary autograft showed the best results with no recoarctation. Similarly, Lee and colleagues[92] reported on 33 patients with a median age of 17 days undergoing arch repair for coarctation and arch hypoplasia (n = 31) or interrupted arch (n =

2) with a variety of autologous vascular patches and showed no recoarctation at a median follow-up of 25 months. In a study by Li and colleagues[82] of 121 infants undergoing arch reconstruction using end-to-side (n = 37), patch with autologous pericardium (n = 53), bovine pericardium (n = 20), or autologous pulmonary artery (n = 11), with a mean follow-up of 1042 days, the recoarctation rate was 51% in the end-to-side technique group and 30% in the patch group, with the lowest recoarctation rate in the autologous pulmonary artery group at 9%. On multivariable analysis, technique approached significance ($P = .097$), with autologous pericardium best and end-to-side technique worst.

Other studies in which patch augmentation was not the primary repair strategy have found patch augmentation was a risk factor for recurrence. For example, a study by Adamson and colleagues[52] of 74 patients with biventricular circulation and coarctation who underwent repair using a variety of techniques, with 25 patients having patch and 31% performed through a sternotomy, found patch was a risk factor for recurrence on multivariable analysis. The details of the surgical repair strategies, how each repair strategy was determined, and the patch material(s) used were not included.

A study by Sen and colleagues[10] of 47 infants and neonates who underwent repair of isolated coarctation at a median age of 1.2 months, all repaired through a sternotomy, showed a recoarctation rate of 43% with pericardial patch (n = 23) and 19% with resection and end-to-end anastomosis (n = 21), but there were no further details on decision-making for the surgical strategy, which also included 2 subclavian flap aortoplasties and 1 interposition graft. On univariate analysis, only the gradient on the predischarge echocardiogram was associated with recurrence.

A study by Rakhra and colleagues[44] that examined 305 patients with arch hypoplasia, with a median age of 11 days, undergoing repair through a sternotomy in 24%, found that those repaired with patch material had a 60% incidence of recoarctation; however, that technique was reserved for complex cases, and details regarding patch material were not included.

Although there are limitations as noted regarding the data on patch augmentation, it appears that pulmonary homograft or autologous material are likely better choices for patch material than CardioCel, Dacron, and PTFE in neonatal primary coarctation repair.

Patients undergoing coarctation repair are also at risk for aneurysm formation in the long-term. The overall risk varies by surgical repair, at 50% when Dacron patch material is used, 17% subclavian flap, 6% interposition graft, and 3% end-to-end.[83] The average time to aneurysm formation is 12 years, with false aneurysms known to develop at suture sites and true aneurysms opposite patch material.[83]

INTERPRETATION AND APPLICATION OF STATEMENTS. Although these statements provide guidance considering the available data, these are not intended to be prescriptive, and practitioners should apply these based on their experience, as well as within the clinical setting in which they work. Surgeons and institutions, for example, with extensive experience in performing repair through a sternotomy with a certain technique and have demonstrated excellent results will have a lower threshold to use this approach as opposed to thoracotomy in a patient with a hypoplastic arch that is "borderline" compared with a surgeon for whom this is not the case. Furthermore, we anticipate that such recommendations will continue to change as more data are acquired.

CONCLUSIONS. Surgery remains the standard of care for the management of isolated coarctation in neonates and infants. Depending on degree and location, arch hypoplasia may require a sternotomy approach as opposed to a thoracotomy approach. Significant opportunities remain to better delineate management in these patients, including better elucidation of variables favoring the sternotomy approach.

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