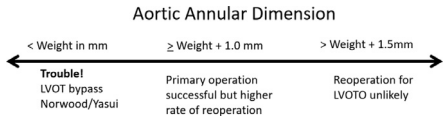
How Small Is Too Small? Decision-Making and Management of the Small Aortic Root in the Setting of Interrupted Aortic Arch

Kyle W. Riggs, MD, and James S. Tweddell, MD

Interrupted aortic arch is commonly associated with a posterior malalignment ventricular septal defect (VSD) and left ventricular outflow tract (LVOT) hypoplasia. Standard repair is carried out in the neonatal period and includes re-establishing arc continuity and VSD closure. Reintervention on the LVOT for obstruction is a common and an ongoing source of morbidity and mortality. A variety of preoperative echocardiographic measurements have been identified to identify patients at risk for developing LVOT obstruction but an aortic valve annulus dimension (mm) < patient's weight (kg) + 1 mm is a reasonable threshold to identify a patient at increased risk for future LVOT reintervention. Prophylactic direct approaches to prevent future LVOT obstruction include myectomy/myotomy and left-sided placement of the VSD patch but do not reliably prevent late LVOT obstruction. Patients amendable to a biventricular repair but with important LVOT hypoplasia are probably best served with a Yasui operation, either as a primary operation or staged with a Norwood procedure. In the case of complex redo operations, a Ross-Konno provides another valuable option for a durable repair. Though smaller preoperative LVOT structures predict the need for reoperation, careful preoperative planning may minimize the need for LVOT reintervention and improve long-term survival.

Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 22:21–26 © 2019 Published by Elsevier Inc.

Keywords: Interrupted aortic arch, Aortic root, Aortic valve, Yasui operation



When to intervene on the LVOT in IAA.

Central Message

For small aortic annuli in IAA, a Yasui operation reduces late LVOT obstruction, but a Ross-Konno is best for complex redo operations.

THE ROAD TO DISCOVERING "HOW SMALL IS TOO SMALL?"

Interrupted aortic arch (IAA) is a congenital heart lesion in which the aortic arch is discontinuous and flow to the descending thoracic aorta is dependent on a patent arterial duct. The interruption may be distal to the left subclavian artery (type A), between the left common carotid and the left subclavian artery (type B) or between the innominate artery and the left common carotid artery (type C). Type B interruptions account for about two-thirds of cases, type A occurs in about one-third of cases, and type C is extremely rare, present in less than 1% of cases. The right subclavian artery arises anomalously in 50% of type B interruptions either distal to the left subclavian artery (aberrant right subclavian artery) or from the arterial duct (isolated right subclavian artery). A ventricular septal defect (VSD) is nearly always present for types B and C interruption and when a VSD is present there is commonly posterior malalignment of the outflow septum and the left ventricular outflow tract (LVOT) is smaller than normal. The degree of LVOT hypoplasia is greater in the presence of an anomalous origin of the right subclavian artery. Standard repair of IAA with VSD involves re-establishing arch continuity and VSD closure. As many as 40% of survivors will require reintervention on the LVOT tract by 15 years of follow-up [1,2]. The presence of the small aortic root complicates the initial operation and results in increased long-term morbidity especially the potential for multiple reoperations directed at the LVOT (Fig. 1) [2]. Identifying preoperative anatomic factors that would predict the development of LVOT obstruction (O)

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https://doi.org/10.1053/j.pcsu.2019.02.004

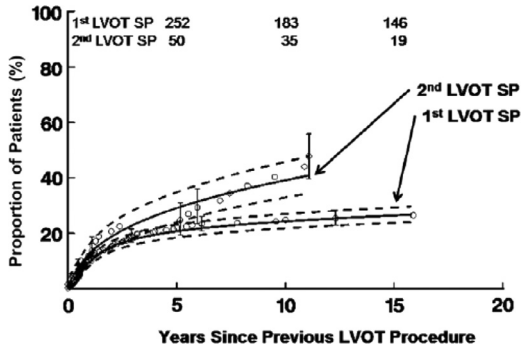

Figure 1 The risk of subsequent surgical procedures for left ventricular outflow tract obstruction increases after the first reoperation. (Reproduced with permission from Jegatheeswaran A et al [2].)

and would allow for alternative surgical strategies would benefit this patient group.

In a 2-center study, Geva et al looked for preoperative anatomic risk factors that predicted postoperative development of left ventricular outflow tract obstruction (LVOTO). They used a stringent definition of LVOTO, a 20 mm Hg peak gradient by echo, and found that among a variety of areas measured, a LVOT (subaortic) cross-sectional area ≤0.7 cm^2/m^2 was a sensitive predictor of postoperative LVOTO after repair of IAA [1]. A study by Apfel et al of 26 patients, using a slightly more generous echo gradient of >40 mm Hg as a definition of recurrent LVOTO found that a preoperative LVOT cross-sectional area greater than 1.6 cm^2/m^2 predicted freedom from LVOTO [3]. This study was also noteworthy in that it demonstrated that an early gradient of >25 mm Hg predicted the need for reintervention (Fig. 2). In a study of 16 patients, Salem et al found that postoperative LVOTO (defined as an echo gradient of >2.5 m/s) correlated with an aortic valve annulus diameter <4.5 mm or a *z*-score <−5 (Fig. 3) [4]. Chen et al, in an analysis of 70 patients, identified an aortic root size of <6.5 mm as the threshold predicting future reintervention [5]. Hirata et al looked at 38 patients and again using reintervention as the definition of

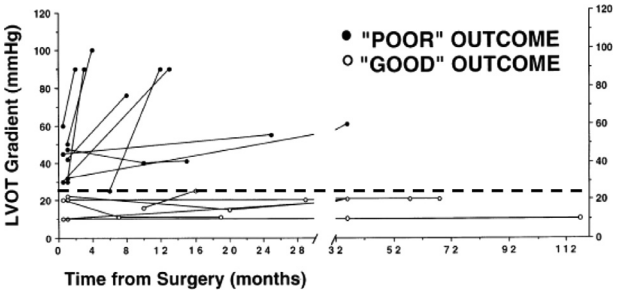

Figure 2 The risk of developing important left ventricular outflow tract (LVOT) obstruction was highly correlated with the early postoperative echo gradient. Those patients with a good outcome (open circles) had an LVOT gradient by echo of less than 25 mm Hg. (Reproduced with permission from Apfel HD et al [3].)

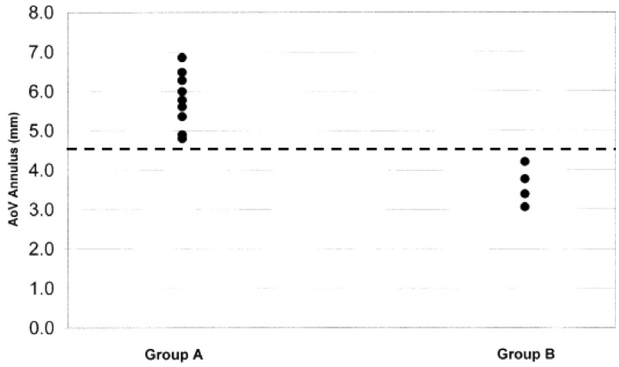

Figure 3 The risk of developing left ventricular outflow tract obstruction (LVOTO) defined as an echo gradient of >2.5 m/s was correlated with aortic annulus size. Development of LVOTO was not seen in patients with an aortic annular dimension >4.5 mm. (Reproduced with permission from Salem MM et al [4].)

LVOTO identified a preoperative aortic annulus dimension in millimeters that was less than the patient's weight in kilograms plus 1.5 [aortic annulus (mm) < patient's weight (kg) + 1.5] as predicting the need reoperation (Fig. 4) [6]. The measurement of the annular dimension and application of this algorithm are straightforward to apply in the operating room. In summary, preoperative identification of hypoplasia of the LVOT or evidence of a residual gradient after repair predicts recurrent LVOTO (Table 1).

STRATEGIES TO PREVENT THE DEVELOPMENT OF LVOTO

If we can reliably identify patients at increased risk of recurrent LVOTO, are there strategies we could apply at the time of repair to prevent the development of obstruction? There are 2 direct approaches to the hypoplastic LVOT that have been

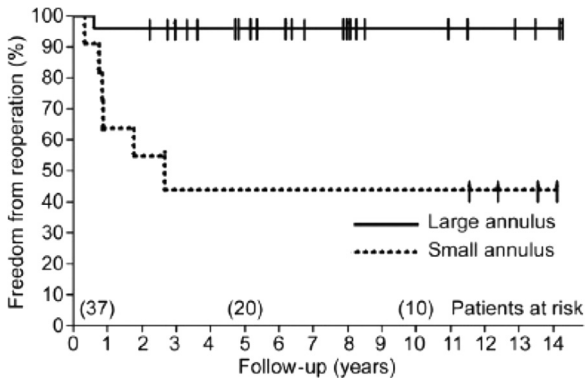

Figure 4 The risk of developing left ventricular outflow tract obstruction (LVOTO) defined as reintervention was dependent on the aortic annular dimension at the time of repair. If the patient's aortic annulus size was greater than patient's weight in kilograms + 1.5 (large annulus), there was a low likelihood of reintervention. (Reproduced with permission from Hirata Y et al [6].)

Table 1 A Compilation of Previously Proposed Definitions and Predictors of LVOTO Put Forth Since 1993

Author	Year	*n*	Definition of LVOTO	Predictor of LVOTO
Geva et al [1]	1993	37	Echo gradient >20 mm Hg	LVOT area <0.7 cm^2/m^2
Apfel et al [3]	1998	26	Echo gradient >40 mm Hg	LVOT area <1.6 cm^2/m^2
Salem et al [4]	2000	16	Echo velocity >2.5 m/s	Aortic valve annulus <4.5 mm
Hirata et al [6]	2010	38	Reintervention	Aortic annular dimension in millimeters < patient's weight in kilograms plus 1.5
Chen et al [5]	2013	70	Reintervention	Aortic root size <6.5 mm

LVOT, left ventricular outflow tract; LVOTO, left ventricular outflow tract obstruction.

used. The first is the myectomy/myotomy (Fig. 5). In a series of 27 patients undergoing repair of IAA, 15 patients were identified as at risk for LVOTO. The criteria identifying those thought to be at higher risk for reintervention were not precisely defined and there was overlap of the aortic valve dimension between the myotomy/myectomy group and those undergoing standard repair. Of the 15 patients subjected to myotomy/myectomy, 6 (40%) underwent LVOT reintervention, suggesting that myotomy/myectomy was not completely successful [7,8]. Luciani et al in a small series of 9 patients described an approach in which at the time of repair the cephalad end of the VSD patch is secured to left ventricular side of the subaortic aorta and the patch is secured such that traction is applied in order to pull the conal septum anteriorly (Fig. 6).

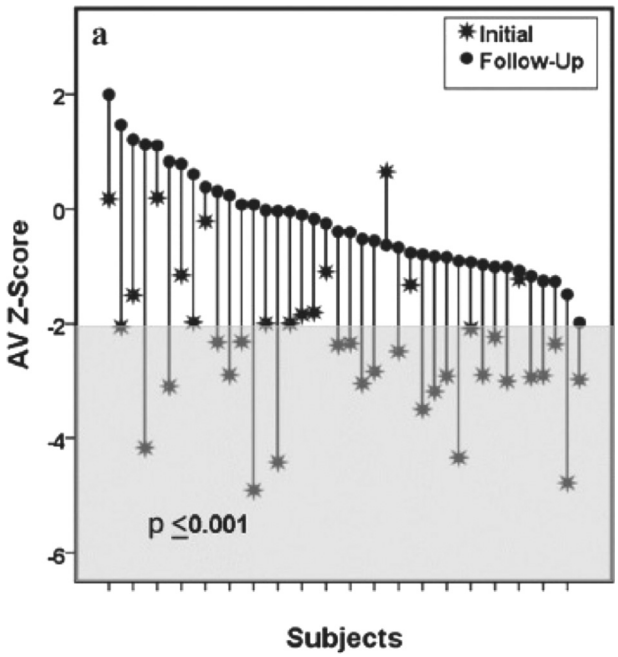

Figure 5 In contrast to the patient with critical aortic stenosis, the aortic valve of the patient with interrupted aortic arch with a ventricular septal defect, while typically bicuspid, is thin and pliable. The annular dimension can increase after repair in response to increased flow. In this review of 73 subjects with arch abnormalities undergoing biventricular repair, the initial median aortic valve *z*-score was −2.34, with normalization to a median *z*-score of −0.4 at follow-up ($P < 0.001$). (Reproduced with permission from Plymale JM et al [23].)

This approach resulted in no subaortic significant obstruction at a median follow-up of 12 months but many of the patients had aortic annular dimensions that would have predicted a low likelihood of reintervention [9]. In addition, equivalent repositioning of the outflow septum that is attributed to VSD patch traction might be accomplished as a consequence of standard repair as repositioning of the outflow septum away from the subaortic area and toward the right ventricle would be anticipated due to the pressure differential between the left and right ventricles after VSD closure. Finally, it is unclear how crowding the subaortic space with the addition patch material would be beneficial. The potential benefits of pre-emptive use of myotomy/myectomy and left-sided VSD patch placement are derived from retrospective studies without uniform indications for prophylactic intervention, furthermore there are no control groups and it is unclear if they are effective in preventing postrepair LVOTO compared with a standard repair.

Another strategy is to bypass the hypoplastic LVOT using the pulmonary outflow tract. Yasui et al described an approach that combines arch repair with a Damus-Kaye-Stansel, patch baffle of the VSD to the combined pulmonary and aortic outflows and placement of a right ventricle to pulmonary artery conduit (Fig. 7) [10,11]. This can be performed as a primary repair or can be staged, using the Norwood operation as the initial procedure. Single-center reports describe 50–80% 5-year survival with frequent reintervention; however, the reinterventions are primarily for conduit replacement, while reintervention for LVOTO is relatively rare at 6% (4 of 70) in late

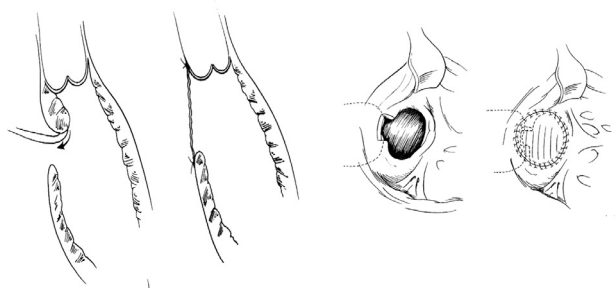

Figure 6 The technique of myotomy/myectomy to prevent reoperation on the left ventricular outflow tract following repair of interrupted aortic arch. The outlet septum below the aortic valve is resected. (Reproduced with permission from Bove EL et al [7].)

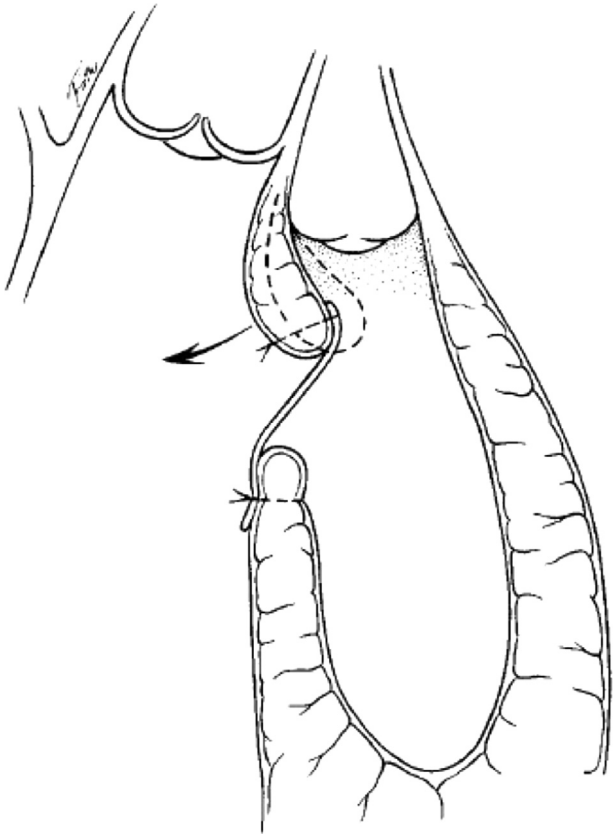

Figure 7 A technique to prevent reoperation on the left ventricular outflow tract (LVOT) following repair of interrupted aortic arch. The cephalad end of the patch is secured to the left side of the outlet septum below the aortic valve. The patch is intended to pull the outlet septum out of the LVOT. (Reproduced with permission from Luciani GB et al [9].)

survivors with a median follow-up of 5.9 years [12–16]. Abarbanell et al reported on 77 patients operated on from 2003 to 2017 and found that for neonates with an aortic root *z*-score of <−2.5, the probability of reintervention was significantly higher in the primary repair group compared to the Yasui group. Therefore, an aortic annular *z*-score of <2.5 may be a reasonable cut-off to identify those patients best managed with a Yasui procedure rather than a standard repair [17].

The Ross-Konno procedure can be used to reconstruct the LVOT with a small aortic root. This can be performed as part of the primary repair for IAA or as an operation for postrepair LVOTO (Fig. 8) [18]. Single-center results show excellent 5-year survival between 77% and 95% in neonates and young infants [19–21]. Despite these reports, an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database found high operative mortality for the Ross-Konno operation at 29% for neonates and 14% for infants [22].

HOW SHOULD WE APPROACH THE PATIENT WITH IAA AND HYPOPLASIA OF THE LVOT?

Comprehensive preoperative evaluation is critical to determining the optimal operative strategy. In contrast to isolated aortic stenosis with an intact ventricular septum, the aortic valve of patients with IAA with a VSD, while commonly bicuspid is thin and pliable and there is potential for acute enlargement and long-term freedom from stenosis (Fig. 8) [23]. There is little data to indicate that pre-emptive procedures, myotomy/myectomy, or left-sided VSD patch are likely to change the risk of developing LVOTO. The choice is therefore between standard repair and a Yasui or Ross-Konno. A reasonable and easy-to-apply rule is to base the decision on the aortic valve

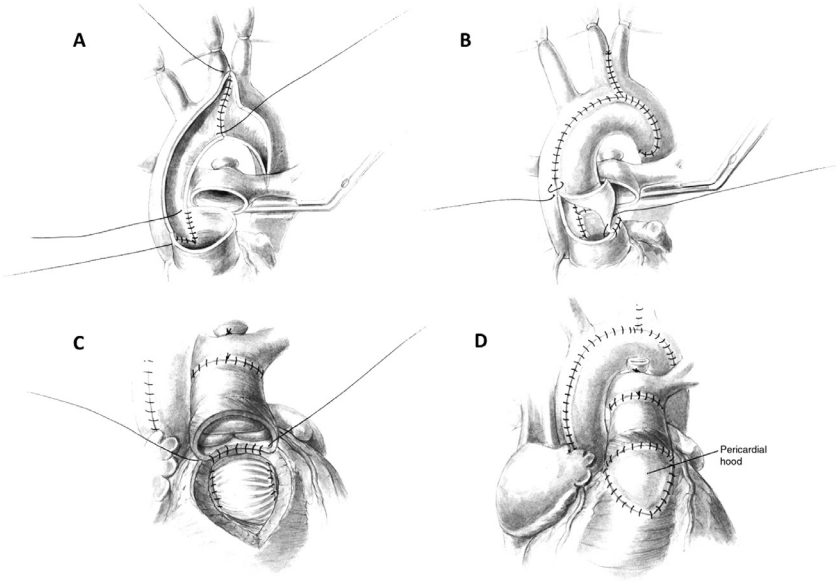

Figure 8 The Yasui procedure severe left ventricular outflow tract obstruction, with an interrupted aortic arch and a ventricular septal defect (VSD). (A) Repair of interrupted aortic arch combined with a Damus-Kaye-Stansel (DKS). (B) Completion of the aortic arch repair and DKS with a patch. (C) The VSD patch baffle is shown connecting the left ventricle to the DKS root. A pulmonary homograft is being used to establish right ventricle to pulmonary artery continuity. (D) The completed reconstruction. (Reproduced with permission from Kanter K [11].)

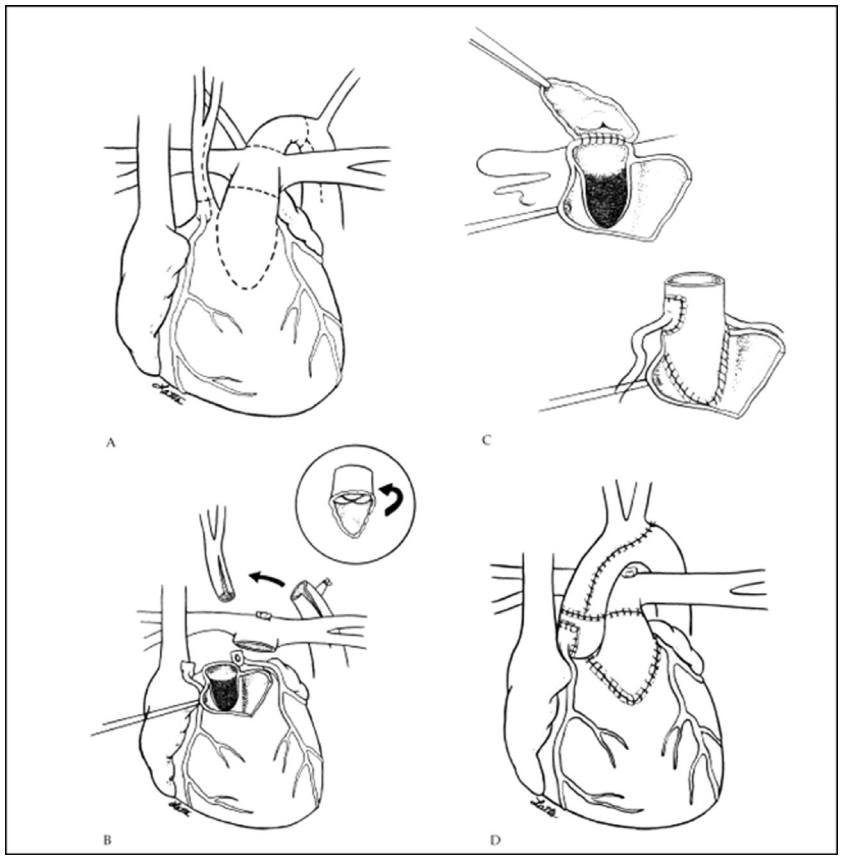

Figure 9 The Ross-Konno procedure as performed for interrupted aortic arch. (A) The anatomy of interrupted aortic type B with the planned incision indicated dashed lines. (B) The pulmonary autograft is harvested with a skirt of right ventricular muscle. Coronary buttons are excised and the proximal coronary arteries are mobilized. The aortic root is incised through the annulus across the outflow septum into the ventricular septal defect. The ascending and the descending aorta are incised vertically to be anastomosed in side-to-side fashion. (C) The pulmonary autograft is implanted and the ventricular septal defect is closed with the muscle flap. Coronary buttons are implanted in appropriate positions. (D) A valved conduit used for reconstruction of the right ventricular outflow tract. The neoaorta and the autograft are anastomosed in end-to-end fashion behind the pulmonary artery. (Reproduced with permission from Hirooka K and Fraser CD [18].)

annulus diameter. If the aortic valve diameter in millimeters is greater than the patient's weight in kilograms + 1 than a standard repair (repair of the IAA and patch closure of the VSD) is typically feasible and important LVOTO is not likely to be present in the early postoperative period (Fig. 9). If the aortic annulus in millimeters is less than the patient's weight in kilograms, the patient is at high risk of recurrent obstruction with a standard repair and a LVOT bypass (Norwood/Yasui operation) should be pursued. Patients between these 2 points are in a gray zone and require case-by-case evaluation (Fig. 10). In those rare cases where the pulmonary valve is not suitable as the systemic valve, a standard repair may be considered with the expectation of reoperation on the LVOT. In those patients with a well-functioning pulmonary valve, an LVOT bypass procedure should be considered. Due to the high mortality associated with the Ross-Konno, it is generally reserved for patients with recurrent complex LVOT obstruction or a challenging VSD to pulmonary artery pathway, which is unlikely to allow unobstructed blood flow.

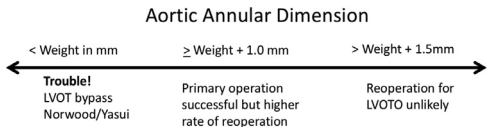

Figure 10 A simplified decision-making algorithm based on aortic valve diameter for management of the interrupted aortic arch with left ventricular outflow tract hypoplasia.

SUMMARY AND CONCLUSION

Recurrent LVOTO is common after repair of IAA with a small aortic root and is an important source of ongoing morbidity and mortality. Preoperative imaging demonstrating smaller LVOT structures is predictive of reoperation. Prophylactic efforts do not appear to reliably protect against the development of LVOTO and need for reoperation. Annular dimensions [annulus (mm) + 1 < weight (kg)] should prompt pre-emptive primary or staged Yasui repair. Lastly, the Ross-

Konno operation seems to be best suited for management of late/recurrent LVOT obstruction as opposed to being used as the initial strategy in infants.

REFERENCES

[1] Geva T, Hornberger LK, Sanders SP, Jonas RA, Ott DA, Colan SD: Echocardiographic predictors of left ventricular outflow tract obstruction after repair of interrupted aortic arch. J Am Coll Cardiol 1993;22:1953–1960

[2] Jegatheeswaran A, McCrindle BW, Blackstone EH, et al: Persistent risk of subsequent procedures and mortality in patients after interrupted aortic arch repair: a Congenital Heart Surgeons' Society study. J Thorac Cardiovasc Surg 2010;140:1059–1075.e1052

[3] Apfel HD, Levenbraun J, Quaegebeur JM, Allan LD: Usefulness of preoperative echocardiography in predicting left ventricular outflow obstruction after primary repair of interrupted aortic arch with ventricular septal defect. Am J Cardiol 1998;82:470–473

[4] Salem MM, Starnes VA, Wells WJ, et al: Predictors of left ventricular outflow obstruction following single-stage repair of interrupted aortic arch and ventricular septal defect. Am J Cardiol 2000;86:1044–1047.A1011

[5] Chen PC, Cubberley AT, Reyes K, et al: Predictors of reintervention after repair of interrupted aortic arch with ventricular septal defect. Ann Thorac Surg 2013;96:621–628

[6] Hirata Y, Quaegebeur JM, Mosca RS, Takayama H, Chen JM: Impact of aortic annular size on rate of reoperation for left ventricular outflow tract obstruction after repair of interrupted aortic arch and ventricular septal defect. Ann Thorac Surg 2010;90:588–592

[7] Bove EL, Minich LL, Pridjian AK, et al: The management of severe subaortic stenosis, ventricular septal defect, and aortic arch obstruction in the neonate. J Thorac Cardiovasc Surg 1993;105:289–295, discussion 295–286

[8] Suzuki T, Ohye RG, Devaney EJ, et al: Selective management of the left ventricular outflow tract for repair of interrupted aortic arch with ventricular septal defect: management of left ventricular outflow tract obstruction. J Thorac Cardiovasc Surg 2006;131:779–784

[9] Luciani GB, Ackerman RJ, Chang AC, Wells WJ, Starnes VA: One-stage repair of interrupted aortic arch, ventricular septal defect, and subaortic obstruction in the neonate: a novel approach. J Thorac Cardiovasc Surg 1996;111:348–358

[10] Yasui H, Kado H, Nakano E, et al: Primary repair of interrupted aortic arch and severe aortic stenosis in neonates. J Thorac Cardiovasc Surg 1987;93:539–545

[11] Kanter K: The Yasui procedure. Oper Tech Thorac Cardiovasc Surg 2010;15:206–222

[12] Gruber PJ, Fuller S, Cleaver KM, et al: Early results of single-stage biventricular repair of severe aortic hypoplasia or atresia with ventricular septal defect and normal left ventricle. J Thorac Cardiovasc Surg 2006;132:260–263

[13] Nathan M, Rimmer D, del Nido PJ, et al: Aortic atresia or severe left ventricular outflow tract obstruction with ventricular septal defect: results of primary biventricular repair in neonates. Ann Thorac Surg 2006;82:2227–2232

[14] Moorthy PS, McGuirk SP, Jones TJ, Brawn WJ, Barron DJ: Damus-Rastelli procedure for biventricular repair of aortic atresia and hypoplasia. Ann Thorac Surg 2007;84:142–146

[15] Hoashi T, Bove EL, Devaney EJ, Hirsch JC, Ohye RG: Intermediate-term clinical outcomes of primary biventricular repair for left ventricular outflow tract obstruction and ventricular septal defect. J Thorac Cardiovasc Surg 2011;141:200–206

[16] Kanter KR, Kirshbom PM, Kogon BE: Biventricular repair with the Yasui operation (Norwood/Rastelli) for systemic outflow tract obstruction with two adequate ventricles. Ann Thorac Surg 2012;93:1999–2005, discussion 2005–1996

[17] Abarbanell G, Border WL, Schlosser B, Morrow G, Kelleman M, Sachdeva R: Preoperative echocardiographic measures in interrupted aortic arch: which ones best predict surgical approach and outcome? Congenit Heart Dis 2018;13:476–482

[18] Hirooka K, Fraser Jr CD: Ross-Konno procedure with interrupted aortic arch repair in a premature neonate. Ann Thorac Surg 1997;64:249–251

[19] Alsoufi B, Al-Halees Z, Manlhiot C, et al: Intermediate results following complex biventricular repair of left ventricular outflow tract obstruction in neonates and infants. Eur J Cardiothorac Surg 2010;38:431–438

[20] Shinkawa T, Bove EL, Hirsch JC, Devaney EJ, Ohye RG: Intermediate-term results of the Ross procedure in neonates and infants. Ann Thorac Surg 2010;89:1827–1832, discussion 1832

[21] Maeda K, Rizal RE, Lavrsen M, et al: Midterm results of the modified Ross/Konno procedure in neonates and infants. Ann Thorac Surg 2012;94:156–162, discussion 162–163

[22] Woods RK, Pasquali SK, Jacobs ML, et al: Aortic valve replacement in neonates and infants: an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. J Thorac Cardiovasc Surg 2012;144:1084–1089

[23] Plymale JM, Frommelt PC, Nugent M, Simpson P, Tweddell JS, Shillingford AJ: The Infant with aortic arch hypoplasia and small left heart structures: echocardiographic indices of mitral and aortic hypoplasia predicting successful biventricular repair. Pediatr Cardiol 2017;38:1296–1304