Highlights in congenital cardiothoracic surgery: 2020–2021

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Advances in congenital cardiothoracic surgery continue at a rapid pace. This year, for the first time, the Editorial Board of the *Journal* provides focused summaries of the key articles published in the American Association for Thoracic Surgery (AATS) journals during the year preceding the annual AATS meeting. These series of focused summaries will be published at the time of the AATS meetings. This particular compendium features articles on 3 topics: aortic valve surgery, the arterial switch operation (ASO), and atrioventricular septal defect (AVSD) repair.

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AORTIC VALVE SURGERY

It is becoming increasingly clear that surgical aortic valve repair achieves better outcomes than balloon aortic dilatation in children, particularly when repair can be performed without patch material.^{1,2} If the valve is of sufficiently good quality to avoid the use of patches, the long-term results are better. A recent report on aortic valve repair without the use of patches in 102 children between 1980 and 2016 found that freedom from aortic valve replacement at 15 years was 50% in neonates and 60% in older children.³ Thus, when aortic valve could be repaired without patches, replacement could be delayed until much later in childhood, when a stabilized Ross procedure might be performed.⁴

An interesting and somewhat controversial technique for aortic valve repair using a ring annuloplasty for a regurgitant bicuspid aortic valve has been described in a child.⁵ Although good initial results have been reported,⁶ the role of this annuloplasty technique is yet to be defined.

Another novelty in aortic valve surgery in children has been the application of complete or partial aortic valve replacement using the Ozaki technique. A recent report of 58 children who underwent the Ozaki technique for aortic valve surgery between 2015 and 2019 demonstrated stable gradients at a median follow-up of 14 months and freedom

CENTRAL MESSAGE

This compendium of recent articles on aortic valve surgery, the arterial switch operation, and atrioventricular septal defect repair highlights current trends in the surgical management of these patients.

from moderate or greater aortic regurgitation of 80% at 3 years.⁷ Similarly, another recent study reported the outcomes of 57 children who underwent the Ozaki procedure between 2015 and 2019 and demonstrated stable gradients and degree of aortic valve regurgitation at a median follow-up of 17 months.⁸ Although the Ozaki procedure could be useful in selected patients, its role in children will be determined by the long-term outcomes.⁹

The Ross procedure remains an important technique in children, with the current focus on improving autograft longevity.^{10–12} An attempt has been made to determine molecular causes for autograft failure in a proteomic study comparing dilated pulmonary autografts following a Ross procedure, with age- and sex-matched controls.¹³ The exact molecular mechanisms and causation are difficult to establish, however.¹⁴ Yet, it is becoming increasingly clear that autograft stabilization is crucial to prevent autograft failure and achieve long-term durability.^{10,15}

ARTERIAL SWITCH OPERATION

The ASO is performed worldwide with excellent results^{16,17}; however, certain subgroups of patients present

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additional surgical challenges. A recent study described the impact of coronary anatomy on outcomes of the ASO in 1033 children who had surgery between 1983 and 2013.¹⁸ Early mortality was 3% in patients with usual coronary arrangement, compared with 4% in the overall group with an anomalous coronary arrangement and 6% in those with a single coronary artery. None of these differences was statistically significant, however. Reintervention on the coronaries was uncommon, occurring in <1% of patients. Patients at risk for these rare coronary events potentially could be identified by computed tomography scanning of the coronary arteries.¹⁹

Another higher-risk group comprises patients with aortic arch obstruction undergoing ASO. A recent study of 83 patients who underwent surgery between 1983 and 2015 demonstrated an early mortality of 6%, no late deaths, and freedom from reintervention of only 68% at 20 years.²⁰ Reintervention was most commonly required for right-sided obstruction; reintervention on the aortic arch was uncommon. These patients may require intraoperative resection of the right ventricular outflow tract obstruction, and thus detailed 3-dimensional (3D) imaging and preoperative simulation may be helpful.

A recent study²¹ using a 3D printed model for training surgeons to perform the ASO demonstrated that repeated practice increased technical performance scores and decreased the time needed to perform the mock procedure, suggesting that 3D training models should be included in congenital cardiac surgery training curricula.

As early mortality has become very low following the ASO, the focus has shifted to the long-term outcomes of these patients as they reach adulthood. A recent study described the long-term outcomes of 844 patients from a single center who underwent ASO between 1983 and 2015.¹⁷ The study demonstrated that although the long-term survival was 95% at 25 years, with the vast majority of patients having normal left ventricular function, 15% of patients had moderate neo-aortic regurgitation or required neo-aortic valve replacement at 25 years. Moreover, the functional health status of 201 adult survivors of surgery for transposition of the great arteries has been reported recently.²² Overall, patients had equal or better scores for health status compared with age-matched normal controls. Patients who had undergone the ASO had higher scores compared with those who received Mustard and Senning palliations, whereas patients who required Rastelli procedure had the lowest scores. This important finding shows that patients undergoing ASO have excellent quality of life into their 30s. Thus, it has now become apparent that despite a low rate of coronary events and excellent functional outcomes, there is an emerging subgroup of patients in their 30s who may need neo-aortic root surgery.²³

ATRIOVENTRICULAR SEPTAL DEFECT SURGERY

The 2 dominant techniques for repair of complete atrioventricular septal defect (AVSD), double patch and modified single patch, both achieve excellent early results; however, whether there is a difference in long-term risk of left atrioventricular valve (LAVV) reoperation or left ventricular outflow tract obstruction (LVOTO) remains unclear.²⁴ A recent multicenter study of 819 children who underwent repair of complete AVSD between 1990 and 2015 demonstrated no difference in survival or freedom from LAVV reoperation or LVOTO in a propensity-score matched analysis of 223 pairs.²⁵

The optimal approach to children with AVSD who develop early heart failure has remained unclear, with some groups preferring an initial pulmonary artery band (PAB) and others proceeding to primary complete repair. A recent study compared outcomes of patients with AVSD who underwent PAB with those who underwent primary complete repair in the Congenital Heart Surgeons' Society Database between 2012 and 2018.²⁶ The study reviewed the outcomes of 50 children who underwent initial PAB and those of 333 children who underwent initial complete repair. The median duration of follow-up was 2.1 years. The use of PAB did not affect the degree of LAVV regurgitation. Of 25 patients with balanced ventricles, 23 progressed to a biventricular repair. They demonstrated that survival and freedom from LAVV reoperation was comparable in those who underwent primary repair and those who underwent complete repair after PAB; however, the analysis began at the time of complete repair in both groups.

The issue of PAB in children with balanced AVSD has been further explored in 2 long-term follow-up studies. One study of 194 patients who underwent AVSD repair at age <3 months between 1990 and 2019 demonstrated improved survival after AVSD repair as a primary procedure.²⁷ In a propensity-matched cohort, the primary complete repair group had significantly better long-term survival compared with the initial PAB group. Furthermore, the subgroup of neonates who underwent complete repair had no mortality and similar freedom from reoperation compared with age 1 to 3 months. The second study described the outcomes of 304 children who underwent repair of AVSD between 1993 and 2018,²⁸ comparing the outcomes in those who underwent repair at age <3 months and those who did so at age >3 months. The study demonstrated no significant difference in the risk of operative mortality, long-term survival, or LAVV reoperation between the 2 groups.

Thus, it appears that primary complete AVSD repair should be preferred when surgery is required at age <3 months and is safe even in neonates. The PAB is certainly an important strategy for patients with unbalanced

AVSD. In those progressing to staged univentricular repair, restoration of the fibrous skeleton of the heart appears to be crucial to achieve the long-term competency of the leaking common atrioventricular valve.²⁹

CONCLUSIONS

The past year has seen important advances in our understanding of the management of the aforementioned congenital anomalies that will result in the improved outcomes.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

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