Curr Pediatr Rep (2017) 5:64–72
DOI 10.1007/s40124-017-0128-5

CARDIOLOGY (W LAI AND W ZUCKERMAN, SECTION EDITORS)

Aortic Valve Surgery in the Pediatric Population

Elizabeth H. Stephens[1] · **Paul Chai**[1]

Published online: 10 May 2017

Abstract
Purpose of Review In this paper, current approaches to pediatric aortic valve surgery are discussed.
Recent Findings While many pediatric aortic valve operations are palliative until an adult-sized prosthetic can fit, some recent advances in adult aortic valve repair techniques can be applied to the pediatric population yielding more definitive repair. Systematic analysis of the valve pathology is necessary to decide on the appropriate operation and refer patients to surgeons with the appropriate expertise.
Summary The Ross operation remains an ideal operation for pediatric patients with significant aortic valve disease and a competent pulmonic valve, given that it allows the autograft to grow with the child. Depending on aortic valve pathology, those with regurgitation secondary to aneurysmal dilation of the aorta may be candidates for various valve-sparing techniques. Critical congenital aortic stenosis may be managed via balloon valvuloplasty or surgical valvotomy depending on the specific scenario. Valve replacement is not ideal in the pediatric population, particularly if the patient is not full-grown, but can be necessary.

Keywords Aortic valve surgery · Ross operation · Bicuspid aortic valve · Aortic stenosis · Balloon valvuloplasty · Aortic valve repair

This article is part of the Topical Collection on *Cardiology*

✉ Elizabeth H. Stephens
Ehs4947@gmail.com

[1] Cardiothoracic Surgery, Columbia University, New York, USA

Introduction

Aortic valve surgery in children is distinct from that of adults on multiple levels. In pediatrics, there is a wide range of etiologies underlying valve dysfunction in a given patient who often has undergone previous interventions. In many cases, their management is a stepping stone intended to sustain the patient until they are large enough for a more definitive solution of valve replacement. Aortic valve surgery in adults, and specifically valve repair, has undergone substantial advances in the last decade. Despite the aforementioned differences between pediatric and adult aortic valve disease, these advances in adults have informed pediatric valve repair. In this review, we will start by discussing the Ross operation, which has long been the standard operative technique for children with valve pathology who are too small for valve replacement. Advances in adult aortic valve repair surgery will then be reviewed, with focus on application to pediatrics, followed by discussion of other operative techniques and the management of specific congenital heart diseases with aortic valve involvement. Lastly, options for aortic valve replacement as well as long-term post-repair management considerations will be discussed.

Etiologies of Valve Disease in Children

Aortic valve dysfunction in children can vary tremendously in terms of underlying etiology, associated cardiac lesions, degree of dysfunction, and timing of presentation. Critical aortic stenosis presents in the neonatal period and is secondary to malformed aortic cusps (often unicuspid or bicuspid). Intervention is required in the neonatal period and debate continues as to whether balloon valvuloplasty or surgical valvotomy is superior (see discussion below). Critical aortic

stenosis can be associated with other cardiac lesions such as aortic coarctation and underdeveloped left-sided structures (i.e., a spectrum of hypoplastic left heart syndrome). Other etiologies causing aortic valve and/or left ventricular outflow tract dysfunction in the pediatric population are supravalvular lesions, such as in Shone's disease (congenital mitral stenosis and aortic stenosis) or Williams syndrome; subvalvular lesions, such as a sub-aortic membrane; and aortic regurgitation associated with aortic aneurysmal disease, such as Marfan syndrome or bicuspid aortic valve disease. Echocardiography is critical to differentiate the level(s) of obstruction (i.e., subvalvular, valvular, supravalvular) as their surgical management differs. As discussed below, echocardiography is also critical to further differentiate valvular etiologies. Aortic valve regurgitation in aneurysmal disease, such as associated with Marfan syndrome, more commonly presents later in the teenage years or young adulthood. Less common etiologies include familial hypercholesteremia that can cause an equivalent of senile, calcific aortic stenosis with accelerated atherosclerosis of the aortic valve. Patients with bicuspid valves or previous cardiac surgery are also at increased risk for endocarditis that can affect the aortic valve.

Some patients who have previously undergone cardiac surgery for a wide range of etiologies later develop issues involving the aortic valve and/or root requiring surgical management. At particular risk are patients with conotruncal anomalies status post-repair, such as truncus arteriosus or tetralogy of Fallot. These patients are at risk long-term for aortic dilatation, which may have associated aortic valve regurgitation. Similarly, as discussed in the Ross operation section below, patients who have previously undergone a Ross operation are at risk for progressive dilation of the pulmonary autograft, especially if an inclusion technique or annular stabilization was not used. If the cusps of the pulmonary autograft are of adequate quality, a valve-sparing aortic root replacement can be applied with excellent results in expert hands [1–4].

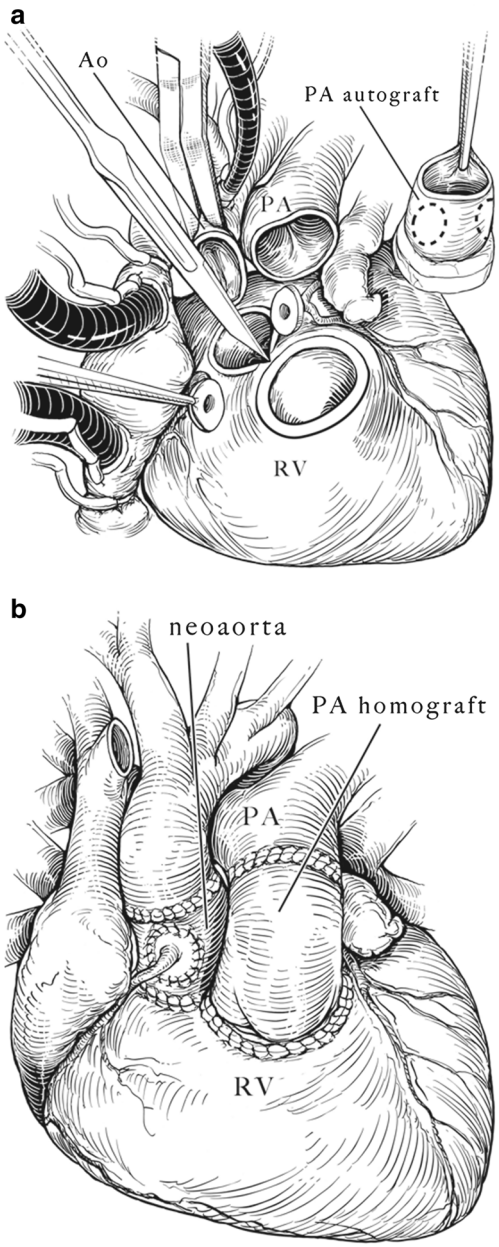

Fig. 1 **a** Operative technique for Ross showing resection of the pulmonary root to be used as an autograft in the aortic position. The diseased aortic valve and root has been resected and removed leaving the coronary buttons. *PA* pulmonary artery, *RV* right ventricle, *Ao* aorta [from Backer/Mavroudis Atlas of Ped Cardiac Surgery]. **b** Completed Ross operation with the pulmonary autograft sitting in the aortic position and coronary buttons reimplanted. A homograft has been used to reconstruct the right ventricular outflow tract. *PA* pulmonary artery, *RV* right ventricle, *Ao* aorta [from Backer/Mavroudis Atlas of Ped Cardiac Surgery]

Ross Operation

The Ross operation, developed by Donald Ross (Fig. 1) [5], has long been the standard technique for children with aortic valve pathology in whom a simple valve repair is inadequate and who are not large enough for a valve replacement [6]. In this procedure, the patient's pulmonary valve and root is resected and used as an autograft to replace the diseased aortic valve and root, while a homograft is used to reconstruct the right ventricular outflow tract. The key advantages of this operation are the ability of the autograft to grow with the child, freedom from anti-coagulation, durability, and excellent hemodynamics [7]. However, this is a technically difficult operation and leaves the patient with two replaced valves [7]. While long-term results after the Ross procedure have been excellent [7], there still exists the risk of autograft dilation and the pulmonary homograft will inevitably need replacement [7]. If the pulmonary valve is dysfunctional, this technique is not an option. A bicuspid pulmonic valve is considered a relative contraindication to the procedure, even if functioning well. It also should not be used with patients with connective

tissue disease or auto-immune diseases such as ankylosing spondylitis and rheumatoid arthritis as these patients are at risk for early failure [7]. In many cases, the pulmonary autograft annulus may be substantially larger than the native aortic annulus; if >2 mm larger, some surgeons plicate between the trigones to correctly size the autograft [7].

Long-term results of the Ross have been excellent in the hands of experts. Yacoub et al. found that in their patients less than 19 years old who underwent a Ross, long-term survival was equivalent to the age-matched general population [7]. David et al. also reported in a cohort of 212 patients that long-term survival at 20 years was similar to the general population [8••], and Sievers et al. reported a remarkable 92% freedom from autograft and homograft replacement at 10 years in 501 patients undergoing a subcoronary Ross procedure [9]. Similarly, Elkins et al. reported on a series of 150 patients with a median age of 12 at the time of surgery and found a survival of 97% at 8 years, freedom from reoperation on the autograft of 90%, and pulmonary homograft 89% [10]. Other series have shown similar results with an 83–90% freedom from reoperation after 5–7 years of follow-up [11, 12–14]. Not unsurprisingly, a trial randomizing patients to either Ross operation or aortic homograft confirmed a survival advantage for Ross patients at 10 years [15] and a study of young adults found patients undergoing a Ross procedure demonstrated a survival advantage compared to patients undergoing a mechanical aortic valve replacement [16].

Generally, the incidence of pulmonary autograft dilation is approximately 20% long-term [7, 17], although reports have been as high as 55% at 7 years [18]. Risk factors for dilation include preoperative annular dilation [15, 17], younger age [17], male sex [15, 17], and preoperative regurgitation [15, 19]. Various modifications to the Ross operation have been designed to stabilize the annulus and prevent dilation. Modifications have included inclusion of the autograft within a Dacron tube [20, 21] or Valsalva graft [22, 23], with some surgeons also including a reduction annuloplasty [24]. With these modifications, groups have reported superior results [21, 24, 25]. However, David et al. found no differences between three commonly used techniques (the root replacement, subcoronary or root inclusion technique) after 20 years [15]. According to Yacoub, the most important factor for preventing dilation is ensuring the autograft fits well within the native aortic annulus; measures such as using a Dacron graft to encompass the autograft in their eyes reduce full mobility of the aortic root and of course would inhibit growth [7]. Similarly, David emphasizes the importance of securing the autograft within the aortic annulus and does not support annular stabilization techniques [15]. Therefore, the Ross operation requires significant experience with the technique, and no particular modification of the original operation has been shown to be uniformly superior.

In patients with a diminutive aortic annulus, the Ross-Konno operation is frequently used (Fig. 2). The Konno portion of the operation is an aortic annular enlargement technique. An incision is made through the right coronary sinus into the anterior interventricular septum and a patch is placed increasing the aortic annular dimension [7]. Once this portion of the operation is performed, the pulmonary autograft can be placed over the left ventricular outflow tract and the homograft placed in the right ventricular outflow tract position as in the original Ross operation. In fact, the Ross-Konno was the most commonly used technique in neonates and infants undergoing aortic valve surgery according to the 2012 STS Congenital Heart Surgery Database (81% of neonates and 32% of infants compared to 32 and 12% for Ross operations and 12 and 2% for homografts, respectively) [6]. However, in some expert's minds, this technique should not be used in children, but rather other techniques such as septal myectomy, mobilization of the fibrous trigones, or posterior root enlargement with a Manouguian or Nicks procedure (described below) are more appropriate [7]. This added complexity to the procedure does add a finite additional risk, including relatively infrequent distortion of the mitral valve causing regurgitation.

Patients who have undergone a Ross procedure are at risk for homograft dysfunction, particularly in younger children, as well as autograft dilation and progressive regurgitation [26]. Most commonly, the homograft in the right ventricular

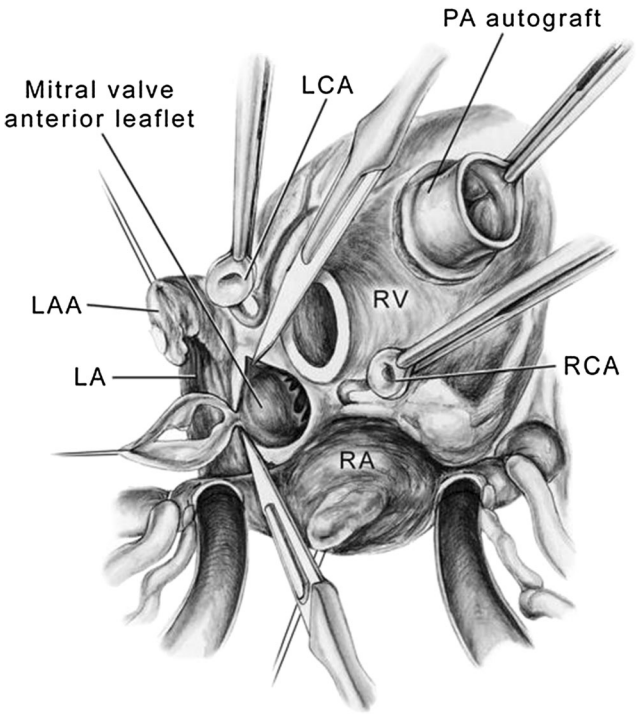

Fig. 2 Incision for expanding the aortic annulus during a Ross-Konno operation for patients with a small aortic annulus. *LA* left atrium, *LAA* left atrial appendage, *RCA* right coronary artery, *LCA* left coronary artery, *RA* right atrium, *PA* pulmonary artery [from Backer/Mavroudis Atlas of Ped Cardiac Surgery]

outflow tract develops obstruction [10], which often can be managed in the cath lab with balloon dilation or placement of a Melody valve (Medtronic Inc., Minneapolis, MN) within the homograft. Melody valve implantation may cause coronary compression and therefore must be assessed prior to placement [27].

Application of Advances in Adult Aortic Valve Repair

In recent years, advances in the understanding of aortic valve disease have led to the development of a number of valve repair techniques largely applied in the adult population. These techniques have allowed valve repair for patients who previously may have required valve replacement. The repair-oriented classification of del Kerchove et al. [28] identifies the underlying cause of aortic regurgitation, allowing specific techniques to be applied to address the underlying pathophysiology. Type I lesions have normal cusp motion with either functional aortic annular dilation (types Ia–Ic) or cusp perforation (type Id), while type II lesions are due to cusp prolapse, and type III lesions are due to restricted cusp(s) [28]. Utilizing echocardiography to identify the type of valve dysfunction aids the surgeon in the most appropriate repair for a given patient. Based on the type of dysfunction, various types of repairs are more likely to be performed, some requiring significant experience in complex techniques. In type Ia lesions where regurgitation stems from underlying dilation at the sinotubular junction (STJ), STJ remodeling with an ascending aortic graft can be used without having to address the root (which would be a higher risk, more complex operation) [29]. In type Ib lesions, where dilation of the STJ and annulus has led to regurgitation, a valve-sparing procedure can be applied if the cusps are of adequate quality (Fig. 3). However, this is a more complex operation and patients should be referred to surgeons with significant experience with valve-sparing procedures. In type Ic lesions, in which dilation of the ventriculoaortic junction has led to regurgitation, a subcommissural annuloplasty can correct the regurgitation, which is a less complex operation. Type Id lesions are due to cusp perforation that can be salvaged with a relatively simple patch repair. Type II lesions are due to cusp prolapse that can be corrected with a number of strategies depending on the specifics of the lesion including free margin shortening, plication, resuspension of commissures, and triangular resection. In type III lesions, the regurgitation is secondary to restricted cusp motion. This can be corrected by resection of raphe if appropriate, shaving, decalcification, and/or patch repair [29]. Type II and III lesions require more complex repair techniques; patients with these pathologies should be referred to surgeons expert in these techniques.

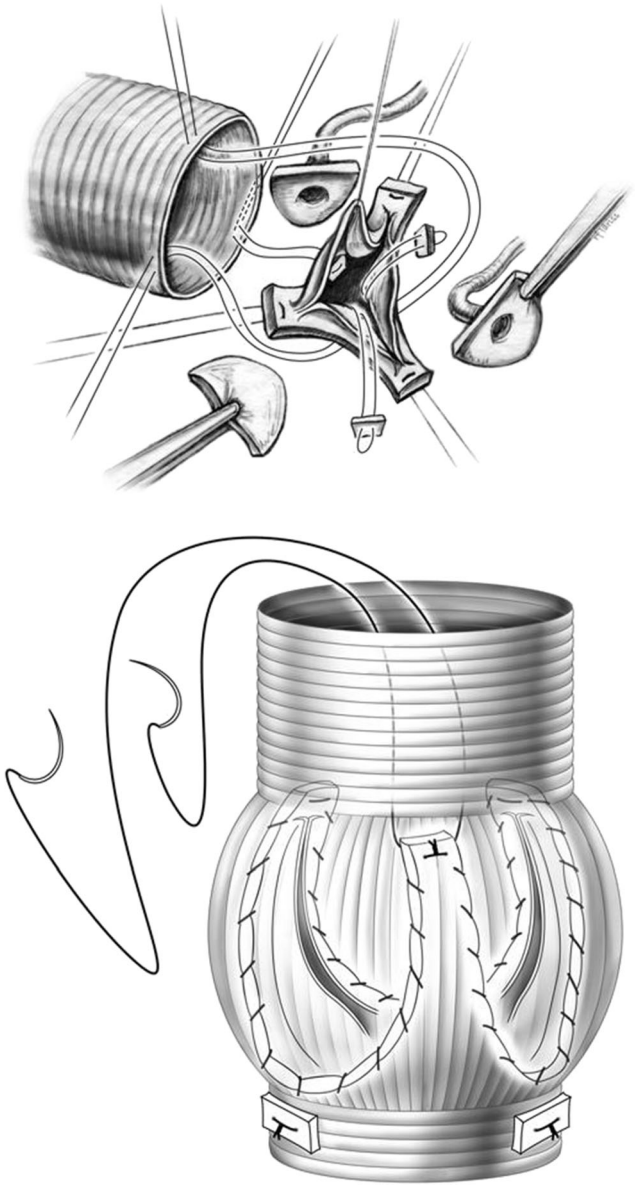

Fig. 3 Tirone David valve-sparing aortic root replacement. The native aortic valve is sutured as shown inside a graft with the coronary buttons reimplanted [from Backer/Mavroudis Atlas of Ped Cardiac Surgery]

This systematic, repair-oriented approach has applications for select pediatric patients. For instance, patients having undergone aortic balloon valvuloplasty for congenital aortic stenosis often have cusp tears that can either be repaired with direct suturing or patch material [7]. Often children presenting with aortic valvular disease have dysplastic, thickened cusps limiting their mobility. In these cases, thinning of the cusps by shaving them and dividing fused commissures may help [7]. In bicuspid patients with prolapse, free margin shortening or even triangular resection of the nodule of Arantius can be performed [7]. Some connective tissue disorder patients present as children with dilation of the root and/or ascending aorta. As discussed

below, in these patients, a valve-sparing aortic root replacement may be the optimal strategy.

Studies in adult aortic valve repair have identified factors predictive of a durable repair including cusp coaptation length (>9 mm in adults), height of coaptation relative to the aortic annulus in the case of valve-sparing procedures, and quality of aortic cusps [30, 31]. Assessment of these preoperative and postoperative characteristics can predict successful durable repair in children [7]. The cusps should be carefully assessed on echocardiography and intraoperatively for thickening, nodules or calcification, mobility, excursion, geometry, tears, raphe, and relative lengths of coaptation (2 mm recommended) [7].

Various materials have been utilized in valve repair, including untreated autologous pericardium, glutaraldehyde-treated pericardium, and various commercially available materials such as CorMatrix (CorMatrix Cardiovascular, Roswell, GA) made from small intestinal submucosa extracellular matrix. Some experts believe that untreated autologous pericardium allows it to adapt better to its environment, may be more resistant to infection, and appears equally durable to treated pericardium [7], while others advocate treatment in 0.5–0.7% glutaraldehyde for 8 min citing prevention of pericardial fibroblast activation and subsequent thickening/calcification [32]. However, others have shown that repairs performed without patch material are less likely to need reintervention [33].

Critical Aortic Stenosis

The management of congenital critical aortic stenosis continues to be debated. Balloon valvuloplasty is performed by interventional cardiologists via arterial catheter access. In this technique, a balloon is inflated that tears the valve allowing more cusp mobility but at the risk of causing significant regurgitation. The particular portion of the valve that tears is dependent on the material properties of the valve and cannot be targeted. In surgical valvotomy, the aorta is opened and the valve is inspected. Depending on the findings, incisions are made to create commissures allowing more cusp mobility. Thickened cusps and/or raphe may be thinned to also improve mobility. If a patient has an associated cardiac lesion, those lesions can also be corrected during this operation. Both balloon valvuloplasty and surgical valvotomy have distinct advantages and disadvantages, and certain techniques appear more appropriate for an individual patient. Randomized studies have not been performed directly comparing the two techniques, but multiple cohort studies have examined the techniques, including their perioperative and long-term outcomes. McCrindle et al. found a greater decrease in systolic gradient in the balloon valvuloplasty group with no difference in survival at 5 years; however, the balloon valvuloplasty group was more likely to have higher grades of regurgitation afterwards [34]. Freedom from reintervention was 48% at 5 years and also was not statistically significantly different between groups [34]. However, another study found that surgical valvotomy demonstrated better gradient reduction, less regurgitation, and less need for reintervention compared to balloon valvuloplasty [35]. Both surgical and balloon valvuloplasty can leave the patients with significant residual valvular lesions, either stenosis or regurgitation. Significant regurgitation is more common in the balloon valvuloplasty group [34], and patients with residual regurgitation after either surgical or balloon valvuloplasty are more likely to need aortic valve replacement sooner [36]. Studies also indicate that regurgitation after balloon valvuloplasty tends to be progressive [37] while the mild stenosis commonly present after surgical valvotomy is not progressive and is well-tolerated by patients [38].

A number of studies have examined the long-term results of balloon valvuloplasty documenting the frequent need for reinterventions. In the study by Maskatia et al. [39], aortic valve replacement was required in 15% at a median of 3.5 years after balloon valvuloplasty and was associated with a gradient after balloon valvuloplasty of at least 25 mmHg and the presence of regurgitation [39]. Death or transplant was necessary in 9% and was associated with decreased left ventricular function [39]. Repeat balloon valvuloplasty was required in 15% at a median of 0.51 years and was associated with neonatal initial intervention, gradient after balloon valvuloplasty of at least 25 mmHg, and depressed left ventricular function [39]. At 15 years, freedom from repeat valvuloplasty was 65%, from aortic valve replacement was 61%, and death or transplant was 87% [39]. Brown et al. [40] similarly found frequent need for reinterventions. After a median follow-up of 9.3 years, 44% underwent reintervention including repeat balloon dilation in 23%, aortic valve repair in 13%, and aortic valve replacement in 23% [40]. Lower gradient post-dilation and lower grade of regurgitation were associated with lower risk of later aortic valve replacement, but age and pre-dilation severity of stenosis was not [40]. In the short-term, balloon valvuloplasty is successful: Torres et al. [41•] found a 71% acute procedural success (defined as peak gradient ≤35 mmHg and no more than mild regurgitation in the critical aortic stenosis group and peak gradient ≤35 mmHg and no increase in regurgitation in the patients with mixed stenosis and regurgitation). Predictors of a successful result included first-time intervention, lack of prostaglandin dependence, and isolated critical stenosis [41•]. Predictors of an unsatisfactory result in neonates included smaller annular size and critical stenosis, while left ventricular function was not predictive [41•]. In those greater than 1 month of age, older age, presence of ≥1+ regurgitation, and history of previous balloon valvuloplasty were associated with an unsatisfactory result [41•].

Neonates who undergo surgical valvotomy also are at risk for requiring reintervention. In a study of 34 neonates followed for a mean of 11 years, there were three early deaths and

freedom from aortic valve replacement at 15 years was 68% [42]. Tricuspid valve morphology gave the best outcome with event-free survival of 90% at 20 years and 100% freedom from aortic valve replacement [42]. Alexiou et al. reported on a smaller group of neonates (18) undergoing open valvotomy and found that 6 required reoperation, 3 of them requiring aortic valve replacement at 9–11 years old [43]. Generally, risk of mortality for neonatal aortic valve replacement is very high, reported as 28% in the recent analysis of STS data [6].

Taking all these data together, balloon aortic valvuloplasty is best employed for neonates in shock and with poor left ventricular function. Surgical intervention is best utilized in patients with a small annulus, aortic regurgitation, ductal-dependent circulations, or those undergoing interventions on the arch or other cardiac lesions. Those with unicuspid, bicuspid, or unbalanced valves (i.e., "functional" bicuspid aortic valves [44]) are also at high risk for procedural failure with balloon valvuloplasty and surgical intervention should be considered [45]. Both groups of patients will need reintervention(s) in the future and need to be carefully followed.

Bicuspid/Unicuspid Aortic Valve and Aortopathy

As in adults, patients with bicuspid aortic valves frequently present with bicuspid aortopathy. While previously the cut-off for intervention on aortopathy in bicuspid patients was akin to those with connective tissue disease [46] [47], more recent guidelines support a more conservative approach [48, 49], with increasing recognition that bicuspid aortic valve patients with regurgitation appear to have a more malignant phenotype [50, 51]. However, specific cut-offs have not been given for bicuspid aortopathy in the pediatric population.

Various surgeons have described techniques for reconstructing the bicuspid aortic valve into a competent tricuspid valve, including commissurotomy and resuspension techniques [52]. Some surgeons prefer performing commissurotomy first for bicuspid valve patients and if an adequate orifice area is not created then proceeding to a commisuroplasty as described by Tolan [53], with reconstruction of the commissural attachments using pericardium [54]. However, some surgeons do not recommend incision of the raphe due to the feeling that residual regurgitation is inevitable [7].

Valve Reconstruction

An innovative technique developed by Shigeyuki Ozaki for use in pediatric and adult patients is reconstruction of the aortic valve using autologous pericardium [55]. With this technique, a template is used to create neo-cusps out of autologous glutaraldehyde pericardium [55]. In the most recent report of his series of 416 patients (including children and young adults), freedom from reoperation was 97% at 73 months of follow-up [55]. The mean aortic valve gradient was 15 mmHg 5 years after surgery with a mean aortic regurgitation grade of less than one [55].

Options for Aortic Valve Replacement

Unfortunately, there are no good options for aortic valve replacement in small children. The 19 Regent mechanical valve (St. Jude Medical, St. Paul, MN) is the best option for small patients; if the patient's annulus will not allow that valve, one should consider a Ross operation or an annular enlargement technique. Homografts can be used if necessary but quickly calcify in children and do not grow like autografts. Advantages of homografts include the excellent hemodynamics, resistance to infection, and lack of need for anti-coagulation [7]; however, as shown in the STS database, homografts had the highest operative mortality (40%) [6]. The four principal annular enlargement techniques utilized include the Manougian, the Nicks, the Nunez, and the Konno. In the Manougian, an incision is made between the left and non-coronary aortic valve cusps and continued down onto the anterior mitral valve leaflet. In the Nunez, the same incision is made but stops at the level of the annulus. In the Nicks annular enlargement, an incision is made in the middle of the non-coronary cusp that terminates at the level of the annulus. In each of these annular enlargements, the incision is closed using a patch. In the Konno, anterior aortic root enlargement is performed with an incision thru the right coronary sinus with a separate incision in the right ventricular outflow tract.

When an aortic valve replacement is performed in children, most commonly a mechanical valve is utilized. As has been well-documented, structural valve degeneration is age-dependent, occurring much more rapidly with younger age [48]. The hemodynamics of a mechanical valve is much better than bioprosthetic valves, especially at smaller annular sizes. However, in high school and college athletes or young women anticipating pregnancy in which anti-coagulation is not an option, a bioprosthetic valve is utilized. The ability for patients to undergo transcatheter valve replacement (TAVR) in the future after bioprosthetic valve replacement may be a good option for some patients; however, there is only limited annular real estate in which a TAVR valve can be placed. As with adults, the risk of thromboembolic events and bleeding is significant in children with mechanical valves on anticoagulation; however, based on limited studies, it appears those risks may be less than reported in adults at approximately 0.3%/patient-year [56–59].

Management of Patients After Aortic Valve Surgery

Patients who have undergone aortic valve repair remain at risk for recurrent regurgitation or stenosis and require lifelong monitoring. The frequency of cardiology clinic visits and echocardiography is determined by the cardiologist and depends on the lesion, type of repair, and the patient's clinical status. Medical management depends on any residual lesions (stenosis or regurgitation), overall cardiac function, and symptoms. For the general pediatrician, particular attention should be paid to murmurs and symptoms and continued communication with the patient's cardiologist and surgeon is paramount.

Future Directions in Pediatric Aortic Valve Surgery

Options remain limited in particular for small, young patients with significant aortic valve disease not amenable to repair. As discussed above, there are no good valve replacement options for such patients; even for patients who are large enough to undergo valve replacement, these valves have limited durability. Ongoing work to develop a tissue-engineered valve that would grow with the child remains relatively far from clinical implementation but would revolutionize the paradigm of valve surgery in the pediatric population [60].

Compliance with Ethical Standards

Conflict of Interest Elizabeth H. Stephens and Paul Chai each declare no potential conflicts of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

References

Recently published papers of particular interest have been highlighted as:
- • Of importance
- •• Of major importance

1. Sundt TM, Moon MR, Xu H. Reoperation for dilatation of the pulmonary autograft after the Ross procedure. J Thorac Cardiovasc Surg. 2001;122:1249–52.
2. Leyh RG, Kofidis T, Fischer S, Kallenbach K, Harringer W, Haverich A. Aortic root reimplantation for successful repair of an insufficient pulmonary autograft valve after the Ross procedure. J Thorac Cardiovasc Surg. 2002;124:1048–9.
3. Masetti P, Davila-Roman VA, Kouchoukos NT. Valve-sparing procedure for dilatation of the autologous pulmonary artery and ascending aorta after the Ross operation. Ann Thorac Surg. 2003;76:915–6.
4. El-Essawi A, Sanger J, Ulrich S, Kutschka I, Harringer W. Aortic valve-sparing operation for autograft failure after the Ross procedure. J Thorac Cardiovasc Surg. 2005;130:1215–6.
5. Ross DN. Replacement of aortic and mitral valves with a pulmonary autograft. Lancet. 1967;2:956–8.
6. Woods RK, 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;144:1084–9.
7. El-Hamamsy I, Yacoub MH. Repair, replacement, Ross: how I approach the older child with mixed aortic stenosis/aortic insufficiency. Seminars in thoracic and cardiovascular surgery: Pediatric cardiac surgery annual. 2009:133–8.
8. •• David TE, David C, Woo A, Manlhiot C. The Ross procedure: outcomes at 20 years. J Thorac Cardiovasc Surg. 2014;147:85–93. **Excellent summary of Ross operation in an expert's hands.**
9. Sievers HH, et al. Fourteen years' experience with 501 subcoronary Ross procedures: surgical details and results. J Thorac Cardiovasc Surg. 2010;140:816–22. 822 e1-5
10. Elkins RC, Knott-Craig CJ, Ward KE, Lane MM. The Ross operation in children: 10-year experience. Ann Thorac Surg. 1998;65:496–502.
11. Lupinetti FM, Duncan BW, Lewin M, Dyamenahalli U, Rosenthal GL. Comparison of autograft and allograft aortic valve replacement in children. J Thorac Cardiovasc Surg. 2003;126:240–6.
12. Laudito A, et al. The Ross procedure in children and young adults: a word of caution. J Thorac Cardiovasc Surg. 2001;122:147–53.
13. Turrentine MW, Ruzmetov M, Vijay P, Bills RG, Brown JW. Biological versus mechanical aortic valve replacement in children. Ann Thorac Surg. 2001;71:S356–60.
14. Hraska V, et al. Ross and Ross-Konno procedure in children and adolescents: mid-term results. Eur J Cardiothorac Surg. 2004;25:742–7.
15. El-Hamamsy I, et al. Long-term outcomes after autograft versus homograft aortic root replacement in adults with aortic valve disease: a randomised controlled trial. Lancet. 2010;376:524–31.
16. Andreas M, et al. The Ross procedure offers excellent survival compared with mechanical aortic valve replacement in a real-world setting. Eur J Cardiothorac Surg. 2014;46:409–13. discussion 413-4
17. Brown JW, Ruzmetov M, Rodefeld MD, Mahomed Y, Turrentine MW. Incidence of and risk factors for pulmonary autograft dilation after Ross aortic valve replacement. Ann Thorac Surg. 2007;83:1781–7. discussion 1787-9
18. Simon-Kupilik N, et al. Dilatation of the autograft root after the Ross operation. Eur J Cardiothorac Surg. 2002;21:470–3.
19. Sievers HH, et al. Major adverse cardiac and cerebrovascular events after the Ross procedure: a report from the German-Dutch Ross Registry. Circulation. 2010;122:S216–23.
20. Slater M, Shen I, Welke K, Komanapalli C and Ungerleider R. Modification to the Ross procedure to prevent autograft dilatation. Seminars in thoracic and cardiovascular surgery: Pediatric cardiac surgery annual. 2005;181–4.
21. Ungerleider RM, Ootaki Y, Shen I, Welke KF. Modified Ross procedure to prevent autograft dilatation. Ann Thorac Surg. 2010;90:1035–7. discussion 1037
22. Juthier F, et al. Modified Ross operation with reinforcement of the pulmonary autograft: six-year results. J Thorac Cardiovasc Surg. 2010;139:1420–3.
23. Gebauer R, Cerny S. A modification of the Ross procedure to prevent pulmonary autograft dilatation. Eur J Cardiothorac Surg. 2009;36:195–7.
24. Brown JW, Ruzmetov M, Shahriari AP, Rodefeld MD, Mahomed Y, Turrentine MW. Modification of the Ross aortic valve

replacement to prevent late autograft dilatation. Eur J Cardiothorac Surg. 2010;37:1002–7.
25. Al Rashidi F, Bhat M, Hoglund P, Meurling C, Roijer A, Koul B. The modified Ross operation using a Dacron prosthetic vascular jacket does prevent pulmonary autograft dilatation at 4.5-year follow-up. Eur J Cardiothorac Surg. 2010;37:928–33.
26. David TE, Woo A, Armstrong S, Maganti M. When is the Ross operation a good option to treat aortic valve disease? J Thorac Cardiovasc Surg. 2010;139:68–75.
27. Gillespie MJ, et al. Transcatheter pulmonary valve replacement for right ventricular outflow tract conduit dysfunction after the Ross procedure. Ann Thorac Surg. 2015;100:996–1003.
28. El Khoury G, et al. Functional classification of aortic root/valve abnormalities and their correlation with etiologies and surgical procedures. Curr Opin Cardiol. 2005;20:115–21.
29. Boodhwani M, et al. Repair-oriented classification of aortic insufficiency: impact on surgical techniques and clinical outcomes. J Thorac Cardiovasc Surg. 2009;137:286–94.
30. Kunihara T, et al. Preoperative aortic root geometry and postoperative cusp configuration primarily determine long-term outcome after valve-preserving aortic root repair. J Thorac Cardiovasc Surg. 2012;143:1389–95.
31. Pethig K, Milz A, Hagl C, Harringer W, Haverich A. Aortic valve reimplantation in ascending aortic aneurysm: risk factors for early valve failure. Ann Thorac Surg. 2002;73:29–33.
32. Grinda JM, et al. Aortic cusp extension valvuloplasty for rheumatic aortic valve disease: midterm results. Ann Thorac Surg. 2002;74:438–43.
33. Al Halees Z, Al Shahid M, Al Sanei A, Sallehuddin A, Duran C. Up to 16 years follow-up of aortic valve reconstruction with pericardium: a stentless readily available cheap valve? Eur J Cardiothorac Surg. 2005;28:200–5. discussion 205
34. McCrindle BW, et al. Are outcomes of surgical versus transcatheter balloon valvotomy equivalent in neonatal critical aortic stenosis? Circulation. 2001;104:I152–8.
35. Brown JW, Rodefeld MD, Ruzmetov M, Eltayeb O, Yurdakok O, Turrentine MW. Surgical valvuloplasty versus balloon aortic dilation for congenital aortic stenosis: are evidence-based outcomes relevant? Ann Thorac Surg. 2012;94:146–53. discussion 153-5
36. Karamlou T, et al. The influence of valve physiology on outcome following aortic valvotomy for congenital bicuspid valve in children: 30-year results from a single institution. Eur J Cardiothorac Surg. 2005;27:81–5.
37. Balmer C, Beghetti M, Fasnacht M, Friedli B, Arbenz U. Balloon aortic valvoplasty in paediatric patients: progressive aortic regurgitation is common. Heart. 2004;90:77–81.
38. Keane JF, et al. Second natural history study of congenital heart defects. Results of treatment of patients with aortic valvar stenosis. Circulation. 1993;87:I16–27.
39. Maskatia SA, et al. Twenty-five year experience with balloon aortic valvuloplasty for congenital aortic stenosis. Am J Cardiol. 2011;108:1024–8.
40. Brown DW, Dipilato AE, Chong EC, Lock JE, McElhinney DB. Aortic valve reinterventions after balloon aortic valvuloplasty for congenital aortic stenosis intermediate and late follow-up. J Am Coll Cardiol. 2010;56:1740–9.
41.• Torres A, et al. Balloon valvuloplasty for congenital aortic stenosis: multi-center safety and efficacy outcome assessment. Catheter Cardiovasc Interv. 2015;86:808–20. **Excellent study of results of balloon valvuloplasty for congenital aortic stenosis**
42. Hraska V, et al. The long-term outcome of open valvotomy for critical aortic stenosis in neonates. Ann Thorac Surg. 2012;94:1519–26.
43. Alexiou C, et al. Open commissurotomy for critical isolated aortic stenosis in neonates. Ann Thorac Surg. 2001;71:489–93.
44. Reich O, et al. Long term results of percutaneous balloon valvoplasty of congenital aortic stenosis: independent predictors of outcome. Heart. 2004;90:70–6.
45. Thomson JD. Management of valvar aortic stenosis in children. Heart. 2004;90:5–6.
46. American College of Cardiology/American Heart Association Task Force on Practice G, et al. ACC/AHA 2006 guidelines for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing committee to revise the 1998 Guidelines for the Management of Patients With Valvular Heart Disease): developed in collaboration with the Society of Cardiovascular Anesthesiologists: endorsed by the Society for Cardiovascular Angiography and Interventions and the Society of Thoracic Surgeons. Circulation. 2006;114:e84–231.
47. Hiratzka LF, et al. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM guidelines for the diagnosis and management of patients with thoracic aortic disease: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, American Association for Thoracic Surgery, American College of Radiology, American Stroke Association, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society of Interventional Radiology, Society of Thoracic Surgeons, and Society for Vascular Medicine. Circulation. 2010;121:e266–369.
48. Nishimura RA, et al. 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Thorac Cardiovasc Surg. 2014;148:e1–e132.
49. Erbel R, et al. 2014 ESC guidelines on the diagnosis and treatment of aortic diseases: document covering acute and chronic aortic diseases of the thoracic and abdominal aorta of the adult. The Task Force for the Diagnosis and Treatment of Aortic Diseases of the European Society of Cardiology (ESC). Eur Heart J. 2014;35:2873–926.
50. Girdauskas E, Disha K, Raisin HH, Secknus MA, Borger MA, Kuntze T. Risk of late aortic events after an isolated aortic valve replacement for bicuspid aortic valve stenosis with concomitant ascending aortic dilation. Eur J Cardiothorac Surg. 2012;42:832–7. discussion 837-8
51. Girdauskas E, Disha K, Secknus M, Borger M, Kuntze T. Increased risk of late aortic events after isolated aortic valve replacement in patients with bicuspid aortic valve insufficiency versus stenosis. J Cardiovasc Surg. 2013;54:653–9.
52. Kadri MA, Hovaguimian H, Starr A. Commissurotomy and bileaflet pericardial augmentation-resuspension for bicuspid aortic valve stenosis. Ann Thorac Surg. 1997;63:548–50.
53. Tolan MJ, Daubeney PE, Slavik Z, Keeton BR, Salmon AP, Monro JL. Aortic valve repair of congenital stenosis with bovine pericardium. Ann Thorac Surg. 1997;63:465–9.
54. Tweddell JS, et al. Aortic valve repair. Seminars in thoracic and cardiovascular surgery: Pediatric cardiac surgery annual. 2005:112–21.
55. Ozaki S, et al. Aortic valve reconstruction using autologous pericardium for aortic stenosis. Circ J. 2015;79:1504–10.
56. Lubiszewska B, et al. Mechanical valve replacement in congenital heart disease in children. J Heart Valve Dis. 1999;8:74–9.

57. Alexiou C, McDonald A, Langley SM, Dalrymple-Hay MJ, Haw MP, Monro JL. Aortic valve replacement in children: are mechanical prostheses a good option? Eur J Cardiothorac Surg. 2000;17:125–33.
58. Champsaur G, et al. Mechanical valve in aortic position is a valid option in children and adolescents. Eur J Cardiothorac Surg. 1997;11:117–22.
59. Shanmugam G, MacArthur K, Pollock J. Mechanical aortic valve replacement: long-term outcomes in children. J Heart Valve Dis. 2005;14:166–71.
60. Fioretta ES, Dijkman PE, Emmert MY, Hoerstrup SP. The future of heart valve replacement: recent developments and translational challenges for heart valve tissue engineering. J Tissue Eng Regen Med. 2016;