Aortic Valve Interventions in Pediatric Patients

Ismail Bouhout, MD, MSc,[*,‡] *Papa Salmane Ba, MD,*[*,‡] *Ismail El-Hamamsy, MD, PhD,*[†,‡] *and Nancy Poirier, MD*[*,†,‡]

Aortic valve (AV) disease in pediatric patients requires a complex decision process that has an impact on decades of life. The aim of this review is to summarize the current evidence surrounding AV interventions in this patient population. In neonates with critical aortic stenosis, the relative merit of surgical vs balloon valvuloplasty is debated and practices vary depending on centers' experience with little comparative literature. In children and adolescents, AV repair has regained interest in the last decades with encouraging early and mid-term results. The Ross procedure represents the best AV replacement option as it offers growth potential, excellent hemodynamics, low rates of endocarditis, and thromboembolism without the risks of anticoagulation. Based on contemporary literature, we propose a management algorithm for children AV disease.

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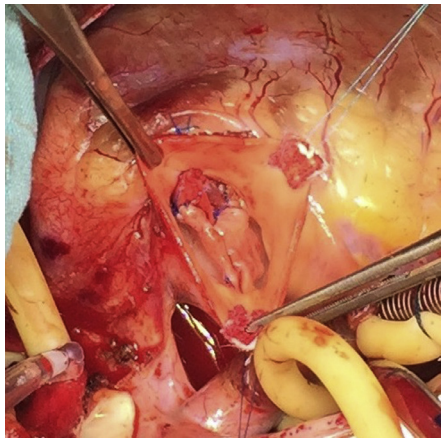
Keywords: Aortic valve, Repair, Replacement, Ross procedure, Congenital



Repair of a unicuspid aortic valve with a patch extension.

Central Message

The treatment of aortic valve disease in children often requires more than one intervention over the course of their lifetime.

Perspective Statement

Surgical aortic valve repair offers a precise and tailored solution and should be considered over balloon valvuloplasty in an effort to preserve the valve and ultimately postpone as much as possible an aortic valve replacement. The Ross operation represents the ideal replacement option with excellent durability, growth, and low valve-related complications.

INTRODUCTION

Treating aortic valve disease in pediatric patients entails choosing not one but a series of interventions that have a lifelong impact. The ideal management strategy must offer durable solutions for a growing child with optimal hemodynamic performance and low rates of valve-related complications. This has long been a subject of debate, and is all the more so with developments in transcatheter approaches. While balloon valvuloplasty (BVP) more widely used at different ages to address severe aortic stenosis, there is a regained popularity AV repair using principles used till recently in adults. Furthermore, recent long-term data on the Ross procedure and prosthetic replacement options in this population provide further insight on best practices. The aim of this review is to summarize the current evidence on aortic valve interventions in pediatric patients and propose a contemporary management algorithm based on published data from the last decade (Fig. 1).

Abbreviations: AR, aortic regurgitation; AS, aortic stenosis; AV, aortic valve; AVR, aortic valve replacement; BVP, balloon valvuloplasty; LVOTO, left ventricle outflow tract obstruction; RVOT, right ventricle outflow tract; SV, semilunar valve; VV, venous valve

*Cardiovacsular Surgery, CHU Sainte Justine, Montreal, Québec, Canada
†Cardiac Surgery, Montreal Heart Institute, Montreal, Québec, Canada
‡Division of Cardiac Surgery, University of Montreal, Montreal, Québec, Canada

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Address reprint requests to Nancy Poirier, MD, Division of Cardiac Surgery, University of Montreal, CHU-ME Ste-Justine, 3175 Côte Ste-Catherine, Montréal, Québec H3T1C5, Canada. E-mail: ncpoirier@videotron.ca

LITERATURE SEARCH

We performed a search on PubMed and EMBASE with the following keywords: "aortic valve" and ([pediatric] or [congenital] or [neonate] or [infant] or [children] or [adolescent]) from 2007 to 2017. A total of 3349 abstracts were screened by 2 independent reviewers (IB and PSB) using a systematic review software (www.convidence.com). In case of duplicate papers with accumulating numbers of patients or increased follow-ups, the most informative studies were included. Papers not written in English, case reports, editorials, expert opinions, and animal studies were excluded. Discrepancies among the

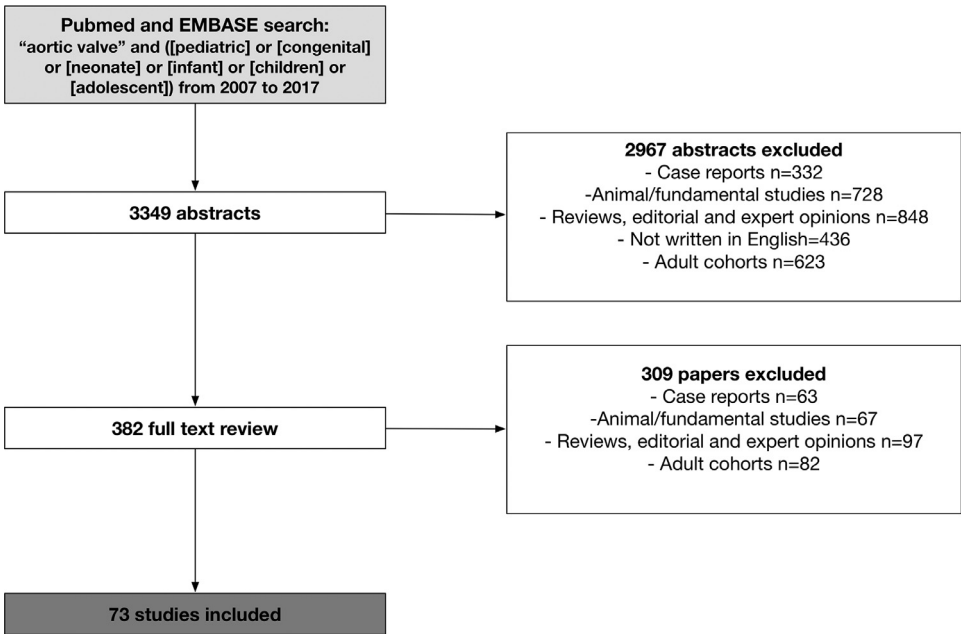


Figure 1. Study flow chart.

reviewers were resolved by consensus. A total of 73 studies were included after full text review. The study flow chart is presented in Figure 1.

INAUGURAL INTERVENTION FOR CRITICAL AORTIC STENOSIS: SURGERY VS BALLOON VALVULOPLASTY

In neonates and infants, critical aortic valve stenosis can be relieved by either percutaneous BVP or surgical intervention. The best treatment strategy remains controversial, and practices vary between centers depending on local expertise and not necessarily reflected in the results published in contemporary literature as summarized in Table 1.

Though BVP has been associated with low early mortality and efficacy in relieving aortic stenosis (AS),[1–9] it carries a high mid- and long-term risk of reintervention: 50–80% of neonates and infants undergoing BVP require surgical intervention at 10 years due to significant aortic regurgitation (AR).[1–9] Furthermore, BVP appears to jeopardize the potential for subsequent AV repair.[2,4–8,10] Alternatively, open surgical techniques that include extensive resection of dysplastic tissue associated with cusps and commissural reconstruction allow a more tailored and lesion-specific approach.[10] There are few studies comparing BVP vs surgical valvulotomy with early and long-term outcomes. Siddiqui et al[10] reported long-term outcomes in 123 neonates and infants undergoing aortic intervention for critical AS. Surgical valvotomy was associated with a significantly higher freedom from aortic reintervention when compared to BVP (27% vs 65% at 5 years).[10] Similarly, Benson et al[11] reported a lower freedom from aortic valve reintervention in the long term after BVP when compared to SVP (25% vs 50% at 10 years) in 84 neonates. However, this result did not reach statistical significance ($P = 0.60$). The Ross procedure in neonates is mostly reserved for unrepairable AV disease and remains a high-risk operation, with early mortality of up to 40%, including high-volume centers (Table 2).[12–20] Interestingly, neonates who survive to hospital discharge have subsequent long-term survival comparable to older children. In addition, a low hazard of reintervention on the pulmonary autograft (<5% at 10 years) is observed, probably due to better adaptation of the autograft to systemic conditions at this early stage in life.

Therefore, given the high-operative risks associated with neonatal Ross procedures, BVP or surgical repair should be the favored approach in neonates and infants, with the aim of postponing aortic valve replacement (AVR) while optimizing AV function.[26] The current shift to percutaneous treatments is not supported by the available evidence as BVP is associated with a lower freedom from AR and may affect subsequent aortic valve repair.[6,8–13,15] Nevertheless, the choice of approach should depend on surgical expertise, availability of interventional facilities, patient conditions, left ventricle function, and overall center outcomes.

TREATMENT OPTIONS FOR CHILDREN AND ADOLESCENTS

Aortic Valve Repair

Aortic valve repair is an attractive option in pediatric patients as it avoids the long-term risks of anticoagulation, allows growth, and postpones AVR. In the last decade, there has been a 3-fold increase in AV repairs[21–23] partly because of favorable results published in adults with low reintervention rates and long-term valve-related complications.[24] A systematic approach to AV repair in young adults with recent refinements has led to better reproducibility and improved durability of the

Table 1. Outcomes of Balloon Aortic Valvuloplasty and Surgical Aortic Valvulotomy in Neonates and Infant Published Between 2007 and 2017

Study	Year	Age	n	FU	Early Mortality	Survival	Freedom From Reintervention	Freedom From AVR
				Balloon valvuloplasty				
Han[1]	2007	3.5 d (0.5–30 d)	53	3.2 y	13%	86% at 10 y	33% at 10 y	59% at 10 y
Fratz[2]	2008	8.1 d (0–28 d)	68	17.5 y	10%	71% at 10 y	47% at 10 y	NR
Crespo[3]	2009	<1 y	66	5.5 y	9.1%	>75% in infant and 55% in neonates at 10 y	50% at 10 y	NR
Brown[4]	2010	<1 y	232	9.3 y	NR	NR	NR	80% at 10 y
Ewert[5]	2011	0.3 y	1004	2.7 y	3%	NR	50% at 10 y	NR
Rossi[6]	2011	22 d (0–92 d)	30	6.8 y	3%	NR	50% at 6.7 y	NR
Petit[7]	2012	13 d	48	4.8 y	2%	51% at 10 y	51% at 10 y	34% at 10 y
Patel[8]	2015	4 d (1–54 d)	30	9 y		82% at 10 y	55% at 10 y	NR
Sullivan[9]	2016	34 d (5–243 d)	154	6.1 y	NR	85% in neonates and 94% in infants at 10 y	32% in neonates and 44% in infants at 10 y	45% in neonates and 62% in infants at 10 y
				Aortic valvulotomy				
Hraska[90]	2012	15 d (1–28 d)	34	11 y	9%	57% at 20 y	NR	68% at 10 y
Galoin-Bertail[91]	2016	17 d (0–111 d)	83	4.2 y	6%	87% at 5 y	58% at 5 y	67% at 5 y
				Comparative studies				
Siddiqui[10]	2013	27 d	BVP 37 SVP 86	10 y	BVP 3% SVP 3%	88% at 10 y	BVP 27% at 10 y SVP 65% at 10 y $P = 0.004$	55% at 20 y
Benson[11]	2015	7.9 d	BVP 57 SVP 27	12 y	NR	BVP 91% at 10 y SVP 82% at 10 y $P = 0.15$	BVP 25% at 10 y SVP 50% at 10 y $P = 0.60$	BVP 70% at 10 y SVP 90% at 10 y $P = 0.32$

AVR, aortic valve replacement, BVP: balloon valvuloplasty, d, days; m, months; SVP, surgical valvuloplasty; y, years.

repair.[25] This same approach is now being proposed for young children, tailored to their aortic valve anatomy and size. The link between the dynamic anatomy of the aortic root (including AV cusps, annulus, and sinuses of Valsalva) and the pathophysiology of AV dysfunction can be objectively assessed and addressed using a variety of techniques (Table 3).[26] Nevertheless, AV repair in congenital patients is a more complex procedure, especially in patients with previous BVP and varying degrees of cusp retraction and thickening. In addition, the prevalence of unicuspid aortic valves is higher in children than in middle-aged adults.[27] These patients often present with significant degrees of AI and aortic annulus dilatation. Repair of unicuspid AVs using a bicuspidization technique provides excellent immediate results.[28,29] The frequent need for autologous or bovine pericardial patch extensions to repair AV cusps in pediatric patients appears to be associated with limited long-term durability of the repair possibly due to retraction and calcification of these materials.[21,30–32] Ongoing investigations into novel processing techniques of pericardial patches and research into alternative materials may lead to improvements in long-term outcomes.

Long-term results of AV repair in pediatric patients are summarized in Table 4.[21,23,30,31,33–38] The early mortality is reported between 0% and 3%, and long-term survival exceeds 95% at 10 years. However, freedom from aortic valve reintervention ranges between 50% and 80% at 10 years, with a median time to reoperation between 4 and 6 years.[21,23,30,31,33,34,37] In contrast, most studies report freedom from AVR >70% at 10 years. Various factors are predictive of reintervention including preoperative AS,[33] unicuspid AVs,[36] lack of coaptation symmetry,[39] and the use of patch material for cusp extension.[21,36] In addition, moderate AS following AV repair is reported in up to 10% of pediatric patients,[21,32] especially in those with cusp extensions. In contrast, in patients with rheumatic valve disease, cusp extension techniques are associated with a freedom for valve reintervention >80% at 10 years.[30,32] Predictors of reintervention in this patient population are older age at intervention and recurrence of rheumatic carditis.

Balloon Valvuloplasty

The debate on BVP vs surgical repair as an initial procedure also extends to older children. Contemporary studies reported high rates of reintervention following BVP varying from 50% to 70% at 10 years.[4,40–45] Recent studies showed a lower median time to reoperation, a higher rate of reintervention, and a lower gradient reduction with BVP when compared to SVP.[42,43,45] In addition, recent published cohorts suggest that a previous BVP may reduce the durability of a subsequent aortic valve repair.[37,38] Nonetheless, the relative merit of the BVP as a first intervention in older children have to be better defined in larger comparative studies.

Table 2. Outcomes of the Ross Procedure in Pediatric Cohorts Published Between 2007 and 2017

Studies	Year	Technique	n	Age	FU	Konno	Early Mortality	Survival	Overall Freedom From Reintervention	Freedom From Autograft Reintervention	Freedom From RVOT Reintervention
					Neonates and infant cohorts						
Shinkawa[12]	2010	Full root	31	18 d (1–349 d)	6 y	80 %	16%	76.7% at 10 y	50.6% at 10 y	95.2% at 10 y	71.5% at 10 y
Maeda[13]	2012	Full root	24	28 d (1–236 d)	6.8 y	100%	4%	95% at 10 y	36.9% at 5 y	NR	43.8% at 5 y
Elder[14]	2013	Full root	34	6 m (4 d–18 m)	10.6 y	76%	12%	88% at 10 y	NR	96% at 10 y	64% at 10 y
Mookhoek[15]	2015	Full root	76	85 d (6 d–347 d)	3.6 y	68 %	17%	75% at 10 y	NR	98% at 10 y	51% at 10 y
					Mixed age cohorts						
Pasquali[55]	2007	Full root	121	8.2 y (4 d–34 y)	6.5	20%	2.5%	NR	70% at 8 y	81% at 8 y	83% at 8 y
Kadner[56]	2008	Full root	52	5 y (4 d–15 y)	3.6 y	4%	10% N&I (33%) C&A (0%)	82% at 10 y	57% at 10 y	NR	60% at 10 y
Kirkpatrick[57]	2008	Full root	60	11.5 y (9 d–17 y)	5.6 y	38%	3.3% N&I (40%) C&A (0%)	96.7% at 12 y	66.2% at 10 y	81.2% at 10 y	76% at 10 y
Horer[58]	2009	Full root (96%) Subcoronary (4%)	152	10.1 y (54 d–15 y)	6.1 y	5.9%	2.6%	90.4% at 10 y	77.5% at 10 y	95.5% at 10 y	79.6% at 10 y
Alsoufi[47]	2010	Full root	227	12.1 y (1 w–18 y)	7.8 y	14 %	1.3%	95% at 10 y	NR	84% at 10 y	81% at 10 y
Frigiola[59]	2010	Full root (83%)	95	12 y (8 m–17 y)	7 y	8.4%	NR	96% at 10 y	NR	80% at 10 y	NR
Clark[60]	2011	Full root (80%) Root inclusion (17%) Dacron inclusion (3%)	54	13.5 y (0.5–35 y)	6.4 y	4%	0%	100% at 10 y	NR	100% at 10 y	71% at 10 y
Tan Tanny[61]	2013	Full root (91%) Root inclusion (6%) Subcoronary (3%)	100	8.6 y (3 d–18 y)	7 y	29%	6% N&I (32%) C&A (0%)	90.8% at 10 y	65.8% at 10 y	86% at 10 y	75.5% at 10 y
Lo Rito[19]	2014	Full root	140	9.6 y (6 d–35 y)	10.8 y	14%	2.8% N&I	N&I: 81% at 10 y C&A: 98% at 10 y	NR	N&I: 100% at 10 y	N&I: 64.6% at 10 y

(continued on next page)

Table 2. (*continued*)

Studies	Year	Technique	n	Age	FU	Konno	Early Mortality	Survival	Overall Freedom From Reintervention	Freedom From Autograft Reintervention	Freedom From RVOT Reintervention
							(13.6%) C&A (0.8%)			C&A: 84.4% at 10 y	C&A: 84.4% at 10 y
Andreas[62]	2014	Full root	70	10 y	10 y	6%	1.6%	90% at 10 y	NR	NR	NR
Brancaccio[18]	2014	Full root	55	5.9 y (2 d−17 y)	5.5 y	48 %	13% N&I (46%) C&A (3%)	85% at 10 y	48% at 10 y	74% at 10 y	56% at 10 y
Luciani[20]	2014	Full root (66%)	305	9.4 y (2 d−18 y)		24%	3.3% N&I (22%) C (0.7%)	93% at 10 y		86% at 10 y	90% at 10 y
Bansal[17]	2015	Full root	305	13.1 y (0.01−70.3 y)	8.2 y	27%	N&I (17.1%) C (1.2%) A (1.2%)	N&I: 71% at 8 y C: 98% at 8 y A: 97% at 8 y	N&I: 58% at 8 y C: 69% at 8 y A: 66% at 8 y	N&I: 100% at 8 y C: 78% at 8 y A: 68% at 8 y	N&I: 58% at 8 y C: 74% at 8 y A: 90% at 8 y
Nelson[16]	2015	Full root	240	NR	10.7 y	32.5%	4.2% N&I (18%) C&A (1%)	89% at 10 y N&I: 72% at 10 y C&A: 92% at 10 y	NR	75% at 10 y N&I: 95% at 10 y C&A: 70% at 10 y	77% at 10 y N&I: 47% at 10 y C&A: 84% at 10 y
Brown[54]	2016	Full root	115	11 y (1 m−18 y)	7.8 y	12%	0.8%	94% at 10 y	NR	NR	NR
Tran[63]	2017	Full root	75	10.2 (5.3 m−18 y)	5.2	0%	0%	100% at 10 y	92% at 10 y	NR	65% at 10 y
Schneider[64]	2017	Full root (96%) Root inclusion (2%) Subcoronary (2%)	154	12 (19 d−48 y)	10 y	32%	5%	87% at 10 y	81% at 10 y	93% at 10 y	89% at 10 y

A, adolescent; C, children; d, days; I, infants; m, months; N, Neonates; NR, not reported; RVOT, right ventricle outflow tract; y, years.

Table 3. Techniques Used for Aortic Valve Repair in Pediatric Patients

Techniques	Lesions
Triangular resection of cusp-free edge	Cusp prolapse
Cusp plication	
Commissural resuspension	
Cusp shaving	Cusp thickening
Cusp repair using interrupted suture or patch	Torn or perforated cusp
Patch extension	Cusp retraction
Circumferential annuloplasty	Annular dilatation
Subcommissural plication	

The Ross Procedure

The Ross procedure is the preferred aortic valve substitute in children and adolescents as it offers growth potential, excellent hemodynamics and quality of life, and avoids the risk of anticoagulation and thromboembolic events.[46] It is the procedure of choice when the aortic valve is not amenable to repair in the absence of connective tissue disorders, active rheumatic disease, or autoimmune pathologies.[35,47]

The initial subcoronary technique described by Donald Ross in 1967[48] has been modified over the years in order to decrease the risk of reoperation due to autograft dilatation. Most centers now use a full-root replacement technique in pediatric patients because it is more reproducible and better preserves pulmonary root geometry.[20,49] Recently, there has been growing enthusiasm for adding a synthetic reinforcement graft to the autograft in adults.[50,51] The main concern with this approach is constraining the dynamic physiology of the living pulmonary root in the aortic position, which may impact long-term outcomes. In addition, in young children, this could blunt growth of the autograft root in parallel with somatic growth of the patients. Nevertheless, early experience suggests a reduced incidence of long-term reintervention in adolescents when using this approach.[17,52] Additionally, concomitant aortic ring annuloplasty to adjust autograft-to-aortic annular geometry, as well as stabilization of the sinotubular junction with sutures or synthetic bands have also demonstrated a reduction in the rate of reintervention in young children.[53,54] Furthermore, strict blood pressure control during the 6 months following the Ross procedure[26] is thought to be critical in avoiding early autograft dilatation, while allowing it to adapt to systemic hemodynamics.[12,14] The effectiveness of these adjunct techniques remains, however, to be better ascertained in larger cohort studies with longer follow-up.

The results of recently published Ross cohorts are summarized in Table 2,[16–20,47,54–64] showing an early morality rate of 0–3% in children and adolescents. Long-term survival ranges from 82% to 100% at 10 years and was higher in older patient cohorts. Freedom from reintervention on the autograft varies from 74% to 100% at 10 years. In a recent meta-analysis, Takkenberg et al[46] reported a pooled annualized risk of autograft reintervention of 1.38% patient-years. Patients with isolated AR and those with rheumatic valve disease were at a higher risk of autograft dilatation and reintervention following the Ross procedure.[16,47,65] In addition, freedom from reintervention on the right ventricular outflow tract (RVOT) was reported between 50% and 81% at 10 years. The risk of RVOT reoperation is higher in younger patients and is inversely related to the conduit size implanted.[13,15,65] Furthermore, pulmonary homografts perform better than aortic homografts and heterografts, and should be favored to reconstruct the RVOT.[15,18,60,61,65] Interestingly, the use of transcatheter techniques for RVOT failure is increasingly used as first-line therapy, which effectively delays or obviates the need for surgical reintervention.[14,60]

THE LAST OPTION—PROSTHETIC VALVE REPLACEMENT

Mechanical Aortic Valve Replacement

Mechanical AVR has been reserved for children with aortic valve disease that is not amenable to a valve-sparing or a Ross procedure. It is more reproducible than techniques previously described and can be performed with low early mortality in young children[66–69] but has significant long-term morbidity. However, the need for lifelong anticoagulation is associated with a risk of major bleeding or thromboembolic events of 1% per patient-year in pediatric cohorts.[35,54,67–72] In addition, concerns over compliance with medication and avoidance of contact sports in adolescents pose an inherent risk and can translate into reduced quality of life in this young patient population.[73] Although mechanical prostheses have been considered durable, there is a non-negligible risk of reintervention in children reported between 5% and 20% at 10 years,[66,69,71,72] mainly due to patient-prosthesis mismatch, endocarditis and nonstructural valve degeneration. Finally, mechanical AVR and anticoagulation in female patients is associated with a higher risk of maternal and fetal adverse events during pregnancy.[74,75]

The long-term survival in pediatric cohorts after mechanical AVR was excellent, with >90% 10-year survival in most recently published cohorts.[35,66–68,71] However, Alsoufi et al recently performed a propensity score analysis comparing pediatric patients undergoing the Ross procedure vs mechanical AVR.[69] The latter was associated with a 2-fold higher risk of early and late mortality. Younger age and smaller prostheses (18 mm) were identified as risk factors for death during follow-up. This mandates performing root enlargement procedures in children to accommodate larger prostheses in order to decrease the risk of patient-prosthesis mismatch. Nevertheless, contemporary studies showed that about one-third of these patients have a size 19 mm or lower prosthesis implanted in the aortic position.[67–69]

Nonliving Biological Substitute

Few reports have been published on the utilization of bioprostheses and aortic homografts in children. Their use in pediatric patients is limited due to a high risk of early valve

Table 4. Outcomes of Aortic Valve Repair in Pediatric Cohorts Published Between 2007 and 2017

Study	Year	Previous BVP	AV Pathology	Patch Extensions	n	Age	FU	Early Mortality	Survival	Freedom From Reintervention	Freedom From AVR
Hawkins[31]	2007	20%	AR 100%	15%	54	8.5 y (2 d–18 y)	6.3 y	2%	98% at 10 y	58% at 10 y	67% at 10 y
Bacha[33]	2008	30%	AR 78 % Mixed 22%	80%	81	8.6 y (1 mo–18.4 y)	4.5 y	NR	96% at 5 y	41.5% at 7.5 y	54% at 7.5 y
Myers[30]	2010	0%	Rheumatic AR	100%	78	12 y	9.6 y	1.2%	NR	80.7% at 10 y	
Polimenakos[34]	2010	14%	NR	100%	142	9.3 y	14.4 y	0%	100% at 10 y	81% at 10 y	72% at 10 y
d'Udekem[21]	2013	12%	AS 54% AR 39 % Mixed 7%	36%	142	9 y (0–20 y)	3.4 y	2%	95% at 10 y	80% at 7 y	81% at 7 y
Khan[35]	2013	8%	AR 47% AS 18% Mixed 32%	NR	97	2.6 y (1–18 y)	3.7 y	2%	70% freedom from death or reintervention at 10 y		NR
Kari[36]	2015	30%	AS	10%	30	8.2 y (2 d–28 y)	3.2 y	3.3%	98% at 8 y	NR	88% at 5 y
Poncelet[23]	2016	20%	AR 55% AS 19% Mixed 26%	15%	66	8 y (4–13.5 y)	4.2 y	1.5%	95% at 10 y	72% at 10 y	81% at 10 y
Wilder[37]	2016	NR	AS 43%	73%	56	10.8 y		0%	NR	64% at 6 y	NR
Vergnat[38]	2017	45%	AR 33% AS 64% Mixed 4%	11%	193	9.2 y (0.1–28.7 y)	5.1 y	0.5%	97% at 10 y	57% at 7 y	68% at 7 y
Poncelet[23]	2017	20%	AR 20% AS 26% Mixed 54%	15%	66	8.8 y	4.2 y	1.5%	95% at 10 y	72% at 10 y	81% at 10 y

AR, aortic regurgitation; AS, aortic stenosis; AVR, aortic valve replacement; BVP, balloon valvuloplasty; d, days; m, months; NR, not reported; y, years.

degeneration that is inversely correlated with age.[35,76–78] More than 50% of patients will require a reintervention at 10 years.[77,78] These substitutes may be considered in patients nonamenable to a valve-sparing or Ross procedure when anticoagulation should be avoided.[77]

DISCUSSION

In the present review, the recent literature is outlined in the proposed management algorithm (Fig. 2). While the controversy between surgical or percutaneous management of the aortic valve in pediatric patients is still ongoing, few studies have directly compared SVP and BVP. Nonetheless, the choice of the best treatment should be tailored depending on patients' conditions (namely the LV function and the feasibility of biventricular repairs) and the availability of surgical or interventional cardiology expertise. Mirroring the recent adult experiences with the transcatheter valve therapies, a pediatric heart team approach should be privileged to achieve the best long-term aortic valve durability without compromising a potential surgical repair.

The aortic valve repair is an interesting alternative as it postpones the need of AVR to an older age where a more definite aortic valve substitute could be implanted. These repair techniques warrant a systematic approach to the aortic valve and root and therefore should be offered in centers with surgical expertise in order to achieve good long-term results. However, aortic valve repair in pediatric patients is complicated by the frequent lack of valve tissue that precludes durable repair. The Ozaki technique consists on replacing all 3 aortic valve cusps with glutaraldehyde-treated autologous pericardium and is an attractive option for children to address this issue.[79, 80] Indeed, a complete reconstruction of aortic leaflets with fixed pericardium has been reported in small pediatric case series with an acceptable immediate result.[81,82] Though this technique has been sufficient for young children to buy time, the mid-term durability remains limited.[81] In addition, the presence of a nongrowing tissue inside a growing vessel could lead eventually to aortic valve regurgitation. Interestingly, Hammer et al[83] have compared semilunar valves (SV) to venous valves (VV) in response to vessel growth in a simulation model. On this simulation model, VVs better maintain a competent closure over a wide range of vessels sizes. Indeed, the leaflet-free edge is only slightly longer than twice the radius of the valve root in diastole in the SV, whereas the leaflet-free edge is more than 4 times the vessel radius in diastole in the VV design. This allows the leaflet to span the distance to the valve center at greater vessel sizes. Furthermore, VV are bileaflet valve with 2 interleaflet commissures approximately 180° apart. As a consequence, the midpoint of a leaflet-free edge moves axially upward along the valve center in response to vessel growth, preserving valve coaptation. However, other factors such as the degree of stretch of the material used[84] and the difference into pressures between veins and aortic roots[85] should be taken into account before extrapolating these results into clinical practice. Nevertheless, further biomechanical studies are needed to develop aortic valve repair techniques that take into account the growing potential of the aortic root.

Some specific situation could be encountered when treating pediatric patients with aortic valve disease. Insufficient quadricuspid truncal valves could be repaired using various techniques namely subcomissural annuloplasty, bicuspidization, or tricuspidization depending on the cusp symmetry and regurgitation mechanism.[86,87] However, the durability of these repair techniques remains poor in the long term.[31,35,87] While aortic regurgitation after the switch operation is uncommon,[88] repair of these neoaortic valves is associated with a higher risk of long-term reintervention.[31] In these specific patient populations, the risk of reintervention given the repair complexity should be weighed against the morbidity associated with a prosthetic AVR.

The Ross procedure offers many advantages in pediatric patients. However, it is a more complex procedure that requires an intervention in 2 valves. In addition, it is associated with a significant risk of reintervention which is higher the younger the patient is. As demonstrated in the adult population, results following the Ross procedure are related to the center surgical volume.[89] Therefore, the Ross procedure should be performed in expert centers and should be postponed as much as possible. On the other hand, the management of patient with hypoplastic annulus or multilevel left

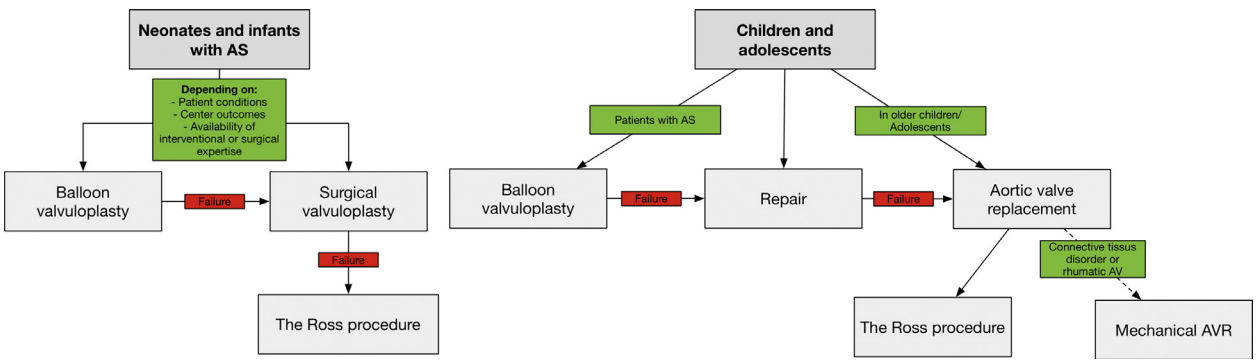


Figure 2. Proposed management algorithm for pediatric patients with aortic valve disease. Dotted line: area of significant controversy. AS, aortic stenosis; AV, aortic valve; AVR, aortic valve replacement.

ventricular outflow obstruction (LVOTO) continues to pose significant clinical challenge. While serial BVP or univentricular palliation is suboptimal, the Ross-Konno procedure represents a more efficient treatment option.[13] As highlighted in this review, the Ross-Konno operation is associated with a high early mortality in neonates and infants. However, this higher operative mortality is probably more related to the patients' substratum than the procedure itself. Therefore, in selected patients with complex LVOTO, the Ross-Konno procedure could represent an interesting alternative.

Limitation

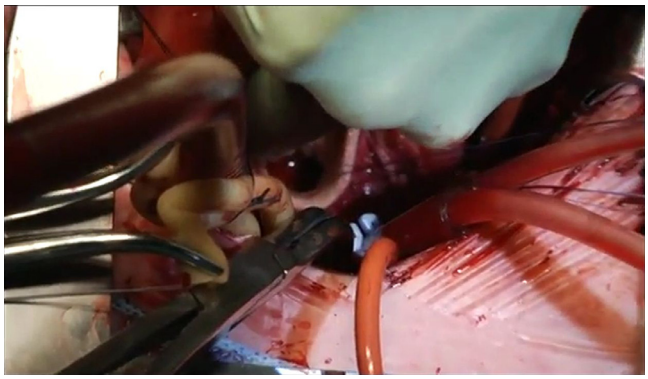
The main limitation of the present review is the lack of robust evidence. Indeed, most of the available literature includes small observational series from expert centers. Therefore, it is difficult to make any strong recommendations. However, this extensive review of the current literature stresses the need for a large multicenter database with a prospective follow-up. In addition, the few number of comparative studies make any meta-analysis of the current data difficult.

CONCLUSION

The treatment of aortic valve disease in children often requires more than one intervention over the course of their lifetime. The choice of these interventions aims to optimize life expectancy and quality of life, while limiting the risk of valve-related complications. Surgical AV repair offers a precise and tailored solution that should be considered over BVP in an effort to preserve the valve and ultimately postpone as much as possible an AVR. The Ross operation represents the ideal replacement option with excellent durability, growth, and low valve-related complications. Although prosthetic AVR is not ideal in the pediatric population, it is an alternative in selected patients that are not Ross candidates.

SUPPLEMENTARY MATERIAL

The following is the supplementary data to this article:



Video 1. Quadricuspid truncal valve repair using a bicuspidization technique.

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