Surgical aortic valvuloplasty in children and adolescents with aortic regurgitation: Acute and intermediate effects on aortic valve function and left ventricular dimensions

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Objective: Surgical aortic valvuloplasty is increasingly employed in the management of children and adolescents with aortic regurgitation, but the durability of this approach and factors associated with outcome are not well defined.

Methods: From 1989 to 2005, a total of 81 patients younger than 19 years with moderate or severe aortic regurgitation underwent surgical aortic valvuloplasty. Aortic regurgitation was congenital in 20 cases, after treatment of aortic stenosis in 30, from other injuries to the aortic valve in 12, and from other causes in 19. Eighteen patients had moderate or severe aortic stenosis. Preoperative left ventricular end-diastolic dimension z score was 4.9 ± 2.7 .

Results: After surgical aortic valvuloplasty with various techniques, including pericardial leaflet augmentation in 80%, aortic regurgitation was improved in 77 patients and was mild or less in 68. Ten of 18 patients with moderate or severe aortic stenosis before repair had a decrease to mild, whereas 2 had progression from mild to moderate. Left ventricular end-diastolic dimension z score decreased by 2.9 ± 2.1 ($P < .001$). During follow-up (median 4.7 years), 33 patients underwent aortic valve reinterventions, including aortic valve replacement in 25. Estimated freedoms from aortic valve replacement were $72\% \pm 6\%$ at 5 years and $54\% \pm 9\%$ at 7.5 years and were shorter in patients with moderate or severe aortic stenosis before surgical aortic valvuloplasty. Among surviving patients who did not undergo aortic valve replacement, aortic regurgitation at follow-up was moderate in 21 and trivial or mild in 34; left ventricular and aortic root dimensions were preserved.

Conclusion: Surgical aortic valvuloplasty is a valid option with good intermediate results for children and adolescents with aortic regurgitation from a variety of causes, particularly for patients with less than moderate aortic stenosis.

Several surgical options are available for the treatment of aortic regurgitation (AR) in young patients, including various techniques of aortic valve repair, or surgical aortic valvuloplasty (SAVP), and aortic valve replacement (AVR) with mechanical,^{1,2} bioprosthetic,^{3,4} or autologous pulmonary^{5,6} valves. SAVP may have particularly important advantages in children relative to the alternatives, including freedom from the risks of a small mechanical prosthesis, avoidance of anticoagulation, and avoidance of the drawbacks of an autograft AVR, such as aortic root dilation and need for a right ventricle–pulmonary artery conduit. The apparent advantages of SAVP in young patients depend, however, on low morbidity and durable aortic valve function after repair. To date, few reports of SAVP in children, most with small numbers of patients and limited follow-up, have been published.⁷⁻¹⁴ The purpose of this study was to analyze results of SAVP in a large number of children and young patients with AR and to explore factors associated with improved outcome.

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Abbreviations and Acronyms

AR	= aortic regurgitation
AS	= aortic stenosis
AVR	= aortic valve replacement
LV	= left ventricular
LVEDD	= left ventricular end-diastolic dimension
LVFS	= fractional shortening
SAVP	= surgical aortic valvuloplasty

Materials and Methods

This protocol was approved by the Children's Hospital Committee on Clinical Investigation.

Patients

The Cardiovascular Program database at Children's Hospital (Boston, Mass) was queried for children and adolescents (<19 years) who underwent SAVP for AR with or without associated valvular aortic stenosis (AS) from 1989 to 2005. Only patients who underwent repair of the aortic valve were included. Patients who had only a root reduction or annular plication without alteration of the valve leaflets were excluded. Other exclusion criteria included the following: a univentricular circulation, a morphologically right ventricle functioning as the systemic ventricle, and other associated causes of left ventricular (LV) volume overload, such as a ventricular septal defect or significant mitral regurgitation. Patients with an anatomic pulmonary or truncal valve in the aortic position were included if they did not meet other exclusion criteria. The decision to undertake surgery was based on the judgment of the treating physicians and not necessarily on strict anatomic or functional criteria. Cross-sectional follow-up was obtained by April 2007.

Classification of AR

The causes of AR were classified as follows: (1) congenital, (2) resulting from transcatheter or surgical treatment of congenital valvular AS, (3) caused by trauma during other cardiac interventions, (4) caused by endocarditis or rheumatic heart disease, (5) related to abnormal truncal valve, and (6) resulting from dilation or distortion of the neo-aortic root or an abnormal neo-aortic (native pulmonary) valve after arterial switch operation.

Echocardiographic Measurements

Aortic valve function and dimensions. AR was graded echocardiographically according to a composite assessment scale as none or trivial, mild (no LV dilation, no retrograde flow in the descending aorta, AR vena contracta width <4 mm), moderate (LV end-diastolic volume or LV end-diastolic dimension [LVEDD] z score >2 and <4, retrograde flow in the descending aorta, vena contracta 4–6 mm), or severe (LV end-diastolic volume or LVEDD z score >4, retrograde flow in the descending aorta, vena contracta >6 mm).

Valvular AS was estimated by Doppler evaluation from multiple views, and the highest maximum instantaneous gradient was recorded. AS was categorized as moderate or greater (gradient >50 mm Hg) or less than moderate ($\leq$ 50 mm Hg). Aortic valve annulus and root diameters were measured from parasternal long-axis images, with the root diameter taken as the maximum dimension at

the level of the sinuses. These measurements were indexed to body surface area and reported as z scores.

Ventricular dimensions and function. LV dimensions, volumes, mass, and fractional shortening (LVFS) were measured and calculated from apical four-chamber and cross-sectional echocardiographic images,¹⁵ indexed to body surface area, and reported as z scores. Short-axis LVEDD and LV end-systolic dimension were measured. In some studies, LV end-diastolic volume and LV end-systolic volume were also calculated. Because LV volumes were assessed adequately at the desired time points for fewer patients than were LV linear dimensions, however, only LVEDD and LV end-systolic dimension z scores were used for analysis.

Aortic Valvuloplasty

A variety of aortic valvuloplasty techniques were used, with multiple techniques often used in a single patient. Most of these, or variations thereof, have been described previously.^{7–13} These were categorized as pericardial augmentation or extension of leaflets, creation of a leaflet, repair of a torn or perforated leaflet, reattachment or resuspension of a dehiscence or prolapsing leaflet, and commissuroplasty. Pericardial augmentation or extension consisted of a variety of techniques, including augmentation of prolapsing or deficient leaflets, use of a pericardial patch to close a rudimentary commissure or combine a rudimentary leaflet with a larger leaflet, and extension of leaflets, usually after excision of thickened or malformed edges. Autologous pericardium was fixed in 0.6% glutaraldehyde for 5 to 10 minutes. Creation of a leaflet was used to replace absent or severely deficient leaflet tissue that was resected; it involved fashioning a patch of autologous pericardium into a leaflet, which was then sutured to the aortic root. Closure of perforated or torn leaflets involved either direct suture closure or closure with a pericardial patch. Reattachment or resuspension of leaflets involved reattachment of dehiscence leaflets, resuspension of prolapsing leaflets, or reattachment of leaflets deliberately separated from the aortic wall to resect a rudimentary leaflet and plicate a dilated aortic root (mostly in patients with truncus arteriosus). Commissuroplasty involved repair or tightening of commissures with mattress sutures placed from outside the aorta to inside the lumen. Patients with coexisting AS often underwent commissurotomy along with the listed valvuloplasty techniques. Thinning of leaflets was frequently performed in conjunction with these techniques and was not specified separately.

Intraoperative transesophageal echocardiography was typically used, particularly during the latter part of our experience, to assess the repair and guide revision if necessary. In our more recent experience, preoperative and intraoperative 3-dimensional echocardiography was frequently used to define the anatomy of the regurgitant valve and to assess the repair (Figure 1).

Data Analysis

The primary outcome was freedom from AVR after SAVP. Secondary outcomes included acute and chronic changes in AR and left heart dimensions and function, severity of AS, freedom from any aortic valve reintervention, and survival. Independent variables analyzed for association with outcomes included age; etiology, severity, and time course of AR; associated anomalies and interventions; preoperative AS; preoperative left heart dimensions and function; type of SAVP; and early postoperative AR. Time-dependent outcomes were assessed with Kaplan–Meier analysis and multivariable Cox regression analysis. For comparison of continuous or

Figure 1. Matrix array 3-dimensional echocardiographic images of aortic valve in patient with severe congenital aortic regurgitation before (*Pre*) and after (*Post*) surgical aortic valvuloplasty. Image before surgical aortic valvuloplasty (*top*) demonstrates partial left-right leaflet fusion, large central deficiency in fused left-right leaflet, and thickening of all leaflets. Image after surgical aortic valvuloplasty (*bottom*) demonstrates three well-formed leaflets that coapt nicely. There was no aortic regurgitation after surgical aortic valvuloplasty. *R*, Right leaflet; *L*, left leaflet; *N*, noncoronary leaflet.

categoric variables between groups, independent samples *t* test and χ^2 analyses, respectively, were used. For comparison of preoperative and postoperative data within patients, paired *t* test analysis was used. Multivariable analysis of discrete outcomes was performed with multiple logistic regression. Correlation between continuous variables was assessed with linear regression. Data are presented as mean $\pm$ SD or median with range.

Results

Patients

From 1989 to 2005, a total of 81 patients met the inclusion criteria for this study and underwent SAVP at a median age of 8.6 years (1 month–18.4 years). Twenty-one of these patients were reported on previously.⁷ Twenty-three patients (28%) were infants and children younger than 5 years, 29 (36%) were children between 5 and 11 years old, and 29 (36%) were adolescents (>11 and <19 years). Sixty-two patients (77%) had associated cardiovascular anomalies, and 57 (70%) had undergone at least one previous intervention (Table E1).

Severity and Causes of AR

Preoperative AR was severe in 52 patients (64%) and moderate in 29 (36%). The distribution of different etiologies of AR is summarized in Table 1. Among patients with congenital

AR, 1 had a quadrileaflet aortic valve with partial deficiency of multiple leaflets, 3 had a completely absent right coronary leaflet, 5 had a tricommissural valve with deficiency of 1 or more leaflets (usually the right or noncoronary leaflets), 1 had a unicommissural valve and central deficiency, and the remaining patients had a bicommissural valve with central deficiency. In all but 1 of these patients, the AR emanated from the area of the right or noncoronary cusp.

Among patients with only moderate AR, indications for surgery included concomitant moderate or severe AS in 11, additional surgical procedures in 8, young age in 4, endocarditis in 2, traumatic AR with difficulty weaning from ventilator support in the early postoperative period in 1, and other in 3.

Among patients with AR caused by intervention for congenital AS, the most recent AS procedure was a median of 3.6 years (10 days–14 years) before SAVP. Among patients with AR resulting from other types of valve injury, the procedure causing valve damage was a median of 3.5 years (1 day–9 years) before SAVP.

Preoperative AS

Moderate or greater AS was present in 18 patients (22%); 15 with AR after previous treatment of AS ($P < .001$ vs other

TABLE 1. Types and causes of aortic regurgitation

Type or cause	No.	%
Congenital	20	25
After previous treatment of aortic stenosis*	30	37
Balloon aortic valvuloplasty	28	35
Surgical aortic valvotomy	4	5
Other traumatic or iatrogenic cause	12	15
Leaflet perforation during subaortic stenosis resection	3	4
Leaflet perforation during modified Konno procedure	2	2
Other leaflet perforation (VSD closure, Rastelli operation, or atrioventricular canal defect repair)	3	4
Leaflet tethering from VSD closure during ASO	2	2
Leaflet or commissure tethering from repair of aortopulmonary window with tetralogy of Fallot	1	1
Leaflet partially avulsed during transcatheter closure of multiple muscular VSDs	1	1
Truncus arteriosus, abnormal truncal valve	9	11
Transposition of the great arteries with root distortion or abnormal neo-aortic valve after ASO	5	6
Rheumatic fever or endocarditis	5	6

VSD, Ventricular septal defect; ASO, arterial switch operation. *A total of 40 previous balloon dilation or surgical aortic valvotomy procedures were performed in 30 patients, with multiple procedures in 9 patients (2 procedures in 8 and 3 procedures in 1).

causes), 2 with congenital AR, and 1 with truncus arteriosus. Patients with moderate or greater pre-SAVP AS were more likely to have moderate AR than severe AR (11 of 29 with moderate AR vs 7 of 52 with severe AR, $P = .01$) and had lower z scores for both aortic annulus diameter (1.1 ± 1.6 vs 3.9 ± 3.2 , $P = .001$) and LVEDD (2.9 ± 2.1 vs 5.5 ± 2.5 , $P < .001$) than patients without significant AS.

LV Dimensions and Function

LV dimensions and function at baseline are summarized in Table E2. Pre-SAVP LV dilation was less severe in younger (<5 years) patients than in older patients (LVEDD z score 3.7 ± 1.2 vs 5.3 ± 2.9 , $P = .02$), in patients with moderate AR than in those with severe AR (LVEDD z score 5.8 ± 2.4 vs 2.9 ± 2.1 , $P < .001$), in and patients with at least moderate AS than in those with mild or no AS (LVEDD z score 2.9 ± 2.1 vs 5.5 ± 2.5 , $P < .001$). Seven patients, all with moderate AR, had a normal pre-SAVP LVEDD z score (<2); 5 had moderate or greater AS and 2 had AR caused by aortic valve injury during surgery for other anomalies.

Aortic Valvuloplasty

Surgical procedures on the aortic valve are summarized in Table 2. In 80% of patients, including 28 of 30 with AR after treatment of AS, leaflets were augmented or extended with pericardium, often in conjunction with other techniques. Additional procedures were performed in 25 patients (Table 3).

Outcomes

Survival. Cross-sectional follow-up was obtained for all but 1 patient (3%) a median of 4.5 years (1 month–16 years) after SAVP. There were 3 deaths. Estimated survivals according to Kaplan–Meier analysis were $97\% \pm 2\%$ at 1 year and $96\% \pm 2\%$ at 5 years. The first death was that of

a 14-year-old with congenital AR and LV dysfunction who had a cardiac arrest as a result of an aortic dissection 2 days after SAVP. This patient died despite AVR. The second patient died during AVR, 2 years after SAVP, as the result of a platelet transfusion reaction. The third death was that of an infant with an atrioventricular canal defect, multiple muscular ventricular septal defects, and a prosthetic mitral valve who underwent SAVP for traumatic AR and died suddenly at home 1 year after SAVP of unknown causes.

Postoperative AR. Early after SAVP (1–26 days), AR was improved in 77 patients (95%): by 1 grade in 19 patients, by 2 grades in 46, and by 3 or 4 grades in 12. AR was trivial or none in 22 patients (27%), mild in 46 (57%), moderate in 13 (16%), and severe in none. None of the variables analyzed were associated with worse postoperative AR.

At most recent follow-up of the 78 surviving patients (before AVR if applicable), the degree of AR was severe in 11 patients, moderate in 32, and trivial or mild in 35. Among the 55 surviving patients that did not undergo AVR, a median of 4.2 years after repair (1 month–12 years), the degree of AR was moderate in 21 and trivial or mild in 34.

Follow-up aortic root diameter z scores, including measurements before surgery for patients who underwent AVR, did not differ from pre-SAVP z scores ($P = .45$). One patient who underwent AVR had endocarditis diagnosed 4.4 years after SAVP.

Postoperative AS. Among 18 patients with at least moderate AS before SAVP, 10 (56%) had a decrease to mild or less in the early post-SAVP period and 8 continued to have moderate AS. There was a trend toward lower aortic annulus z scores among the 8 patients whose AS did not improve (median z score 0.8 vs 2.2, $P = .11$). Two patients with less than moderate AS before SAVP had an increase to moderate AS in the early post-SAVP period. According to

TABLE 2. Techniques of aortic valve repair and additional surgical procedures

Procedures	No.	%
Aortic valve repair procedures*		
Pericardial augmentation of leaflet(s)	65	80
Commissuroplasty	30	37
Repair of perforated or torn leaflet(s)	18	22
Resuspension or reattachment of dehiscenced, avulsed, or prolapsing leaflet	12	15
Creation of a leaflet	6	7
Additional surgical procedures		
Patients	25	31
Procedures		
Aortic root reduction or aneurysmorrhaphy	5	6
Subaortic stenosis resection or modified Konno procedure	6	7
Replacement of a pulmonary outflow conduit	6	7
Augmentation of ascending aorta for supravalvular aortic stenosis	5	6
Mitral valvuloplasty for mitral stenosis or mild mitral regurgitation	3	3
Reoperative mitral valve replacement	2	2
Other	2	2

*Multiple aortic valve repair procedures were performed in 47 patients (57%), so the number of procedures listed does not total 81.

multivariable logistic regression, the only factor associated with moderate or greater postoperative AS was moderate or greater preoperative AS ($P < .001$).

Eight patients with less than moderate AS both before SAVP and early after SAVP had a diagnosis of moderate or greater AS a median of 3 years (0.5–6 years) after SAVP. Two of these patients had peak Doppler gradients early after SAVP of 45 mm Hg, just below the threshold for moderate AS. In all 8 cases, the SAVP included pericardial augmentation or extension of aortic valve leaflets; in 6, SAVP was originally performed for AR resulting from treatment of congenital AS.

Postoperative LV dimensions and function. On early postoperative echocardiography ($n = 66$), the LVEDD z score was lower than the preoperative z score in all but 4 patients ($P < .001$). The average postoperative LVEDD z score of 2.1 ± 2.6 was 2.9 ± 2.0 lower than the pre-SAVP z score ($P < .001$; Figure 2). Among patients with a pre-SAVP LVEDD z score greater than 4 ($n = 45$), the LVEDD z score decreased in all but 1; overall, the decrease in LVEDD z score in these patients was greater than in those with less severe dilation (decrease by 3.8 ± 1.8 vs 1.5 ± 1.7 , $P < .001$).

At most recent follow-up of patients who did not undergo AVR, the post-SAVP decrease in LVEDD was preserved (z score 1.7 ± 2.4), whereas among those who underwent AVR, the pre-AVR LVEDD z score was 4.2 ± 2.7 . LVFS z score at most recent follow-up among patients who did not undergo AVR was 1.7 ± 2.2 ; among patients who did undergo AVR, the pre-AVR LVFS z score was -0.2 ± 2.6 . In only 2 patients was the LVFS z score below the normal range (< -2); both underwent AVR.

Aortic valve reintervention. During a median follow-up of 4.7 years (1 month–17 years), 33 patients underwent 1

or more reinterventions on the aortic valve: repeated SAVP in 3 cases, balloon aortic valvuloplasty in 8, and AVR in 25. This includes 3 patients who underwent early postoperative reintervention—for revision of the valvuloplasty in 2 cases and AVR in 1—because of dehiscence of a created or augmented leaflet. Freedoms from aortic valve reintervention were $91\% \pm 3\%$ at 1 year, $63\% \pm 6\%$ at 5 years, and $41\% \pm 8\%$ at 7.5 years. Factors associated with shorter freedom from aortic valve reintervention according to multivariable Cox regression were the presence of moderate or greater AS at the time of SAVP (5-year freedom from reintervention $33\% \pm 13\%$ vs $70\% \pm 7\%$, $P < .001$; Figure 3) and larger pre-SAVP LVEDD z score ($P = .05$). None of the other factors analyzed were associated with freedom from AVR. Notably, freedom from reintervention did not differ according to age at SAVP, type or cause of AR, technique of SAVP, or moderate AR early after SAVP.

AVR with a mechanical valve prosthesis ($n = 20$), pulmonary autograft ($n = 4$), or aortic allograft ($n = 1$) was performed in 25 patients a median of 3.7 years (2 days–9 years) after SAVP, at a median age of 13.5 years (10 months–26 years). At the time of AVR, the degree of AR was severe in 11 patients, moderate in 10, and mild in 4, and 11 had at least moderate AS. One of the patients who underwent pulmonary autograft AVR had endocarditis. In patients whose SAVP included pericardial augmentation or recreation of leaflets, the pericardium was often found at the time of AVR to be highly calcified and immobile, which presumably contributed to the ultimate failure of the repair. Among the 4 patients with mild AR, 2 had moderate or severe AS and LV dysfunction, 1 underwent early post-SAVP AVR after an aortic dissection and cardiac arrest, and 1 underwent AVR at the time of reoperation for the primary indication of

Figure 2. Line graph demonstrating change in left ventricular (LV) short-axis end-diastolic dimension (EDD) z score from before surgical aortic valvuloplasty (Pre-SAVP) to early (<1 month) after surgical aortic valvuloplasty (Post-SAVP) for each patient.

a progressive aortic root aneurysm. Freedoms from AVR after SAVP were $95\% \pm 3\%$ at 1 year, $72\% \pm 6\%$ at 5 years, and $54\% \pm 9\%$ at 7.5 years. The only factor associated with shorter freedom from AVR was moderate or greater AS at the time of SAVP ($74\% \pm 7\%$ vs $60\% \pm 14\%$ at 5 years, $P = .05$; Figure E1). None of the other factors analyzed were associated with time to AVR, including age; among children 11 years old or younger at the time of SAVP, freedoms from AVR were $77\% \pm 7\%$ at 5 years and $59\% \pm 10\%$ at 7.5 years ($P = .3$ vs patients >11 years old).

A second SAVP for recurrent AR was performed in 3 patients: in the early postoperative period in 2 patients and 3.4

Figure 3. Kaplan-Meier graph demonstrating freedoms from any aortic valve reintervention and from aortic valve replacement (AVR) after surgical aortic valvuloplasty in entire cohort.

years after SAVP in a patient with truncus arteriosus and a dysplastic aortic valve, whose AR decreased from moderate to mild after repeat SAVP and was unchanged 4.8 years later.

Eight patients (6 with congenital AS and 2 with congenital AR and AS) underwent balloon aortic valve dilatation for AS a median of 5.4 years (0.4–8.3 years) after SAVP, which had included pericardial aortic valve augmentation in 7 patients. Angiographically, the augmented leaflets appeared calcified and immobile in these 7 patients. Balloon dilatation was successful in all cases, reducing the peak AS gradient from a median of 80 mm Hg (55–110 mm Hg) to 40 mm Hg (20–60 mm Hg) without increasing AR to more than mild. An additional balloon dilatation procedure was performed in 3 patients (9–30 months later), with similarly successful results. Three patients (none of whom had a repeated balloon dilatation) subsequently underwent AVR, all within 11 months of balloon dilatation, either for recurrent severe AS ($n = 1$) or for residual or recurrent moderate AS with LV dysfunction ($n = 2$). The 5 patients who did not undergo AVR were followed up for 3.9 to 10 years after SAVP and 0.1 to 3.6 years after the most recent balloon valvuloplasty procedure.

Discussion

Acute Benefits of SAVP in Children and Adolescents with AR

In our experience, SAVP was effective at acutely reducing AR and improving LV dilation in children and adolescents with AR from a variety of causes. Other investigators have reported similarly promising early outcomes after SAVP in children and adolescents with congenital heart disease, although most series are relatively small, include patients with primarily AS, or include simple as well as complex repairs.⁷⁻¹⁴ Nevertheless, the growing body of literature on this topic suggests that SAVP is effective at decreasing AR in the short term and, as we have shown in this study, allows rapid reverse LV remodeling even in patients with severe LV dilation.

Intermediate Outcomes After SAVP in Children and Adolescents with AR

In this series of children and adolescents with moderate or severe AR who underwent SAVP, freedoms from any aortic valve reintervention at 5 and 7.5 years were $63\% \pm 6\%$ and $41\% \pm 8\%$, respectively, and freedoms from AVR at 5 and 7.5 years were $72\% \pm 6\%$ and $54\% \pm 9\%$. Our experience includes many young children, and the durability of repair was just as good in these patients as in older children and adolescents. With regard to intermediate-term aortic valve function, the literature is mixed. Other published studies have demonstrated progressively decreasing freedom from aortic valve reintervention during the several years after SAVP, although the frequency and time course of reintervention vary.^{9,11,12,14} This series was intended to focus on complex repairs in patients with significant AR and did not

include patients with isolated AS, less than moderate AR, or ventricular septal defect–associated AR, for whom the usual techniques of repair have proved durable.^{12,16}

Despite the limited follow-up data on SAVP in children with congenital heart disease and complex AR, more abundant literature concerning the use of pericardial leaflet extension in both children and adults with rheumatic AR and in adults with AR and a structurally normal aortic valve demonstrates that repairs incorporating pericardial augmentation of leaflets can function well for many years.¹⁷⁻²⁰ The optimal approaches to certain technical elements of pericardial leaflet augmentation are not well defined, particularly the duration of pericardial fixation. In patients undergoing AVR after SAVP in our series, the pericardial patches were often heavily calcified and contracted, presumably contributing to progressive post-SAVP aortic valve dysfunction. Our current practice is to fix the pericardium in glutaraldehyde for 5 to 10 minutes, shorter than in our early experience, which leaves the pericardium more pliable; however, the effect of this modification has yet to be determined.

In patients without significant AR after SAVP, LV dimensions and function improved after SAVP and remained stable at follow-up, regardless of pre-SAVP LV size. This is in contrast to the findings of Tafreshi and colleagues,²² who observed that a preoperative LVEDD *z* score greater than 4 was associated with worse functional outcome and persistent LV dilation after AVR in children and adolescents with AR.²¹

Postoperative AS After SAVP in Children and Adolescents with AR

One of the potential adverse outcomes of SAVP for AR in patients with congenitally abnormal valves is postoperative AS. This is a particular concern in patients who have AR develop after balloon dilatation for AS. In our experience, 22% of patients had moderate or greater AS in addition to AR before SAVP, and more than half of these had a reduction in AS to mild or less after SAVP, probably in part as a result of reductions in LV volume load and forward stroke volume. Patients with moderate or greater AS had lower aortic annulus *z* scores than did those with mild or less AS, and there was a trend toward lower aortic annulus *z* scores among patients whose AS did not improve.

New or increased AS after SAVP was uncommon. Even when significant AS was not present early after SAVP, however, it sometimes developed with time. The cause of this progressive AS was not clear, but all affected patients underwent SAVP with pericardium to repair the aortic valve, and pericardial calcification or contraction may have been a contributing factor, as discussed previously.

A unique feature of this series was the subset of 8 patients who underwent balloon aortic valvuloplasty for AS after SAVP. Most of these patients had AR after previous balloon valvuloplasty for congenital AS, and all had significant AS at

the time of SAVP. In all cases, balloon dilatation decreased AS in the acute term. Although 3 of the 8 patients ultimately underwent AVR, the other 5 were alive without AVR as long as 10 years after SAVP. There was no significant increase in AR after balloon dilatation in any of these patients. Accordingly, balloon aortic valvuloplasty may be an effective and safe first-line treatment for patients with AS but minimal AR after SAVP.

Management of AR in Children and Adolescents

Although published reports present guidelines for management of AR in young patients,²² the optimal timing and method of intervention for AR in children remain challenging, and there are limited data to help guide the choice between SAVP and AVR in this patient population. Because serious adverse outcomes were uncommon, our data do not allow inferences that might provide firmer guidance regarding timing and method of intervention in children and adolescents with AR. Of note, age at the time of SAVP was associated neither with worse short- or long-term outcome nor with the duration of freedom from reintervention. Our results do support several important conclusions: there are few short- to intermediate-term drawbacks of SAVP, LV dimensions typically normalize or improve substantially after SAVP even when severe dilation is present, and SAVP allows substantial delay of AVR for most patients, which may facilitate eventual AVR with a larger prosthesis or bioprosthesis. With improved patient selection and surgical techniques, the durability of SAVP should continue to improve.

A common alternative to SAVP in children and adolescents with aortic valve disease is the Ross procedure.^{4-6,23} In contrast to SAVP, the Ross procedure involves replacement of the entire aortic root, with reimplantation of the coronary arteries, as well as placement of a valved conduit between the right ventricle and pulmonary arteries. Apart from an important incidence of need for conduit replacement, there is increasing evidence of significant progressive neo-aortic root dilation and autograft failure.^{5,23}

Limitations

This study is limited by its retrospective nature, by the heterogeneous patient population, and by the fact that referral for surgery was subjective in most cases. Furthermore, this experience encompasses a period when the techniques of SAVP were under development and thus may include a learning curve. Regular serial measurements of post-SAVP LV geometry and function were not available, which limits our insight into the evolution of changes in aortic valve function and LV size and function with time. Although the relative benefits and drawbacks of SAVP relative to therapeutic alternatives, such as mechanical or pulmonary autograft AVR, would be useful, this study does not allow direct comparison between SAVP and AVR.

Conclusions

This study demonstrates that SAVP is a valid option with good intermediate results for children and adolescents with AR from a variety of causes, particularly those with less than moderate AS. We now consider primary SAVP for essentially every patient with AR, aside from those with associated greater than moderate AS or a small aortic annulus, for whom a Ross or Ross-Konno procedure may be preferred.

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Figure E1. Kaplan–Meier graphs demonstrating freedom from aortic valve replacement (AVR) among patients with moderate or greater aortic stenosis (AS) before surgical aortic valvuloplasty and those with less than moderate aortic stenosis. Numbers of patients at risk at 0, 1, 3, 5, 7, 9, and 11 years are listed.

TABLE E1. Associated cardiovascular anomalies and previous interventions

Anomalies and interventions	No.	%
Associated anomalies	62	77
Valvular AS	35	43
Subvalvular AS	11	14
Supravalvular AS	4	5
Coarctation of the aorta	5	6
Mitral stenosis	2	2
Transposition of the great arteries or double-outlet right ventricle	10	12
Truncus arteriosus	9	11
Ventricular septal defect or atrioventricular canal defect	5	6
Tetralogy of Fallot with aortopulmonary window	1	1
Previous interventions	57	70
Previous surgical interventions	35	43
Truncus arteriosus repair	9	11
Arterial switch operation	9	11
After previous pulmonary artery band	4	5
Subaortic stenosis resection or modified Konno procedure	5	6
Surgical aortic valvotomy for AS	4	5
Repair of atrioventricular canal defect	3	3
Coarctation repair	3	3
Mitral valve replacement	2	2
Other	3	3
Previous catheter interventions	32	40
Balloon aortic valve dilatation	28	35
Coarctation dilatation or stenting	3	3
Ventricular septal defect closure	2	2
Pulmonary outflow conduit dilatation	1	1

AS, Aortic stenosis.

TABLE E2. Baseline left ventricular and aortic dimensions and functional parameters

Parameter	Mean $\pm$ SD	Range
LV fractional shortening (%)	37% $\pm$ 7%	25%–55%
LV fractional shortening z score	0.5 $\pm$ 2.1	–3.7–5.2
LV end-diastolic diameter z score	4.9 $\pm$ 2.7	–1.5–12.7
LV end-systolic diameter z score	3.0 $\pm$ 2.6	–3.1–8.5
LV mass z score (n = 67)	3.9 $\pm$ 1.9	–0.4–8.6
Aortic valve annulus diameter z score	3.2 $\pm$ 3.2	–2.9–13.2
Aortic root diameter z score	3.3 $\pm$ 3.2	–0.6–15.7

Adequate data were available for 75 patients unless otherwise specified. *LV*, left ventricular.