Mitral valve reconstruction in a pediatric population: Late clinical results and predictors of long-term outcome

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Objective: Pediatric mitral valve anomalies present complex management challenges to the surgeon, who may have to choose between valve replacement or repair. We review our 18 years of experience to establish the long-term outcomes of pediatric mitral repair.

Methods: Forty-five children (22 boys) with mitral valve anomalies were studied. Mitral reconstruction was performed in all cases at the first instance. The median age at operation was 2.16 years with 18 (40%) younger than 1 year. Patients were divided into two groups: group 1, isolated (mitral anomaly with or without atrial septal defect or patent ductus arteriosus), contained 30 patients (66.6%), and group 2, complex (mitral anomaly with concurrent intracardiac disease), contained 15 patients (33.3%).

Results: In-hospital (30-day) mortality in group 1 was 3.3% (1/30); overall in-hospital mortality was 11.1%. Group 2 had a significantly higher in-hospital death rate of 26.6% (4/15; $P < .05$). There was 1 late death, that of a child who required reoperation. The median follow-up was 5.08 years (range 1-211 months). The 15-year survival in group 1 was 93%, versus 73% in group 2. Seven patients required 9 revision surgical procedures. Two mitral valve replacements were required at reoperation. The 15-year freedom from reoperation was 81.7%. There were no thromboembolic events. The event-free rate at 15 years was 73.5%.

Conclusion: This series compares favorably with others, with 74% to 85% survival and 66% to 85.7% freedom from reoperation reported with valve replacement. Patients with significant associated congenital cardiac abnormalities are at a higher risk of early death after mitral reconstructive surgery.

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Since the early 1980s, we have adopted an aggressive approach to repairing mitral valve anomalies in children.¹ These anomalies present complex management challenges for the surgeon. Not only must the mitral disease be addressed, there are frequently associated congenital abnormalities also requiring reconstruction.² Mitral valve disease is most commonly seen in the setting of a partial or complete atrioventricular septal defect (AVSD), multiple left-sided obstructive lesions, or isolated mitral valve dysplasia. In managing the mitral pathology, the surgeon may have an opportunity to choose between mitral valve replacement and repair. Reconstruction and retention of the native mitral valve and subvalvular apparatus offers distinct advantages. In comparison with valve replacement, mitral reconstruction conserves the subvalvular apparatus and ventricular geometry, preserving left ventricular function and resulting in long-term survival benefits.³ Mechanical valve use in this age group is significantly challenged by patient-prosthesis size mismatch, which is further complicated as the child grows. Mitral repair also reduces the risk of thromboembolism and avoids the need for anticoagulation, which is particularly difficult to manage in the pediatric population.

Several groups have reported excellent results with mitral valve repair.^{1,4-11} Mitral valve abnormalities in the pediatric population are rare, and so there is comparatively small and limited experience with mitral valve repair at each institution. We reviewed a single-operator (A.W.) experience to evaluate whether an aggressive advocacy of repair is justified by calculating survival and freedom from reoperation and identifying the predictors of adverse outcomes in this group of patients.

Patients and Methods

Patients

A retrospective review was performed of all children considered for mitral valve surgery from January 1, 1986, to December 31, 2003. All procedures were performed by a single operator at this institution, which is the sole provider of national pediatric cardiothoracic services. Mitral reconstruction was performed in all cases in the first instance. Records of 45 children (22 boys and 23 girls) with mitral valve anomalies were identified. There was 1 case of acquired mitral incompetence after cardiac catheterization. The other patients had congenital mitral valve anomalies and concordant atrioventricular and ventricular arterial connection. Cases of primary correction of total or partial atrioventricular canal defect and univentricular hearts were excluded. Data were obtained from medical records and, where these were inadequate regarding follow-up, by contact with the patient's general practitioner or family. The median age at operation was 2.16 years (range 2 months to 14.17 years), with 18 (40%) younger than 1 year. The median weight at operation was 11.5 kg (range 1.5-55.2 kg). Pulmonary hypertension was moderate (mean 35-45 mm Hg) in 7 cases (15.5%) and severe (mean >45 mm Hg) in 10 cases (22.2%). Mitral valve reconstruction was possible in all cases reviewed. Complete follow-up was obtained in all but 1 case, that of a female patient emigrated with her family after 9 years of follow-up. She had been well to that point. Of note, 1 male patient committed suicide 13 years after surgery. He had been well before that and is not included as a cardiac-related late death. The median follow-up was 5.08 years (range 1 month to 17.6 years).

Mitral Pathology

Mitral pathology was classified according to the Carpentier pathophysiologic classification (Table 1).¹² Mitral stenosis was the dominant lesion in 15 children, and incompetence was dominant in 30. The most common type of mitral valve malformations resulting in incompetence were anterior leaflet cleft (type I) in 13 patients (43.3%) and annular dilatation (type I) in 11 patients (36.6%). Multiple left heart obstructions (Shone anomaly) involving the mitral valve were found in 1 patient. Seven patients (15.5%) required repair after AVSD correction. All in this group had undergone closure of the cleft at primary AVSD repair, which had been performed within the study era. Patients were divided into two groups: group 1, isolated (mitral anomaly with or without atrial septal defect or patent ductus arteriosus), consisted of 30 patients (66.6%), and group 2, complex (mitral anomaly with intracardiac disease or left ventricular outflow tract obstruction [LVOTO]), consisted of 15 patients (33.3%). In the complex group, 8 patients underwent simultaneous repair of ventricular

TABLE 1. Distribution of anatomic abnormalities in cases of mitral stenosis and incompetence, as classified by Carpentier and colleagues¹²

Mitral valve abnormality	No.
Mitral valve stenosis	15
Type A (normal papillary muscle)	6
Supravalvular ring	4
Leaflet fusion	2
Type B (Abnormal papillary muscle)	9
Parachute	6
Papillary muscle abnormality	3
Mitral valve incompetence	30
Type I (normal leaflet motion)	27
Cleft anterior leaflet	13
Annular dilatation	11
Leaflet defect	3
Type II (leaflet prolapse)	1
Elongated chordae	1
Type III (restricted leaflet motion)	2
Type A (normal papillary muscle)	0
Type B (abnormal papillary muscle)	2
Papillary muscle hypoplasia	2

TABLE 2. Repair strategies used in the reconstruction of mitral valve anomalies at first operative intervention in this series

Repair strategy	No.	%
Stenosis		
Papillary muscle split	9	60
Resection of supravalvular ring	7	46.6
Commissurotomy	5	33.3
Repair of chordae	2	13.3
Wooler-Kay annuloplasty	1	6.6
Annuloplasty ring	1	6.6
Incompetence		
Cleft suture	14	46.6
Posterior annuloplasty	5	16.6
Wooler-Kay annuloplasty	6	20
De Vega anuloplasty	5	16.6
Papillary muscle split	4	13.3
Annuloplasty ring	3	10
Leaflet replacement with autologous pericardium	1	3.3
Leaflet resection	1	3.3
Paneth annuloplasty	1	3.3

septal defect (VSD), and LVOTO was simultaneously addressed in 5, with a subaortic membrane resection in 2 and aortic commissurotomy in 3. One of the 7 patients with previous AVSD repairs had a subaortic stenosis and was placed in group 2. There were 2 patients with mitral incompetence and aberrant coronary arteries requiring reimplantation. In these cases, the degree of mitral incompetence was severe, and on assessment it was considered that mitral function would not improve sufficiently with coronary correction alone.

TABLE 3. Demographic, operative, and outcome data from patients in group 1 (isolated mitral disease with or without atrial septal defect or patent ductus arteriosus) and group 2 (mitral anomaly with intracardiac disease or LVOTO)

	Group 1	Group 2	P value
Total (No.)	30	15	—
Male (No.)	13	9	.292
Age (mo, mean $\pm$ SD)	62.3 $\pm$ 52.9	16.1 $\pm$ 14.1	.002
Mean preoperative pulmonary pressure (mm Hg, mean $\pm$ SD)	35 $\pm$ 16.4	39 $\pm$ 16.6	.526
Mitral stenosis (No.)	9	6	.502
In-hospital death (No.)	1	4	.019
Late death (No.)	1	0	.475
Reoperation (No.)	4	3	.561
Cardiopulmonary bypass (min, mean $\pm$ SD)	54.8 $\pm$ 24.6	80.1 $\pm$ 46.1	.02
Crossclamp (min, mean $\pm$ SD)	26 $\pm$ 14.1	30 $\pm$ 13	.405
Weight (kg, mean $\pm$ SD)	18.2 $\pm$ 12.7	8.4 $\pm$ 3.8	.01
Intensive care unit stay (d, mean $\pm$ SD)	4.4 $\pm$ 4.2	8.5 $\pm$ 7.63	.06
In-patient stay (d, mean $\pm$ SD)	12.15 $\pm$ 7.33	14.4 $\pm$ 9.3	.423

Group 2 had a significantly higher hospital mortality and required surgery at a significantly younger age.

Surgical Indications and Procedure

Children with increasing symptoms (lethargy, fatigue, noted diminution in exercise tolerance), evidence of rising pulmonary pressures (half systemic or greater), or increasing left ventricular chamber size on surveillance echocardiography were considered for surgical intervention. Where possible, associated congenital cardiac defects such as patent ductus arteriosus or aortic coarctation were addressed first, and mitral surgery was delayed. The surgical aim was to restore a widely patent and competent pathway between the inflow and outflow tracts of the left ventricle. More recent surgical management included evaluation of the mitral valve anomaly with intraoperative transesophageal echocardiography, although this constituted only 3 cases of the series, none with reoperation. Surgery was performed with aortobicaaval cannulation and cardiopulmonary bypass, with most group 1 patients (23/30) kept at moderate systemic hypothermia (28°C). Profound hypothermia (18°C) was more frequently used in more complex cases (11/15). Circulatory arrest was not used. The mitral valve was approached through a left atriotomy posterior to the interatrial groove. Initially, antegrade cold crystalloid cardioplegia was used, but the practice evolved to antegrade cold potassium-enriched blood cardioplegia after 1993. The annulus, leaflets, chordae tendineae, and papillary muscles were carefully inspected to determine the precise anatomy of the lesion and to plan the procedure. The mitral orifice size was measured with a Hagar dilator and compared with the expected mean normal valve diameter for the mitral valve relative to body surface area (BSA).¹³ Annular dilatation producing central valve incompetence was treated by means of reduction annuloplasty techniques. The posterior leaflet was advanced forward to coapt with the anterior leaflet, according to the principles of Carpentier.⁶ The annular orifice was downsized to the size predicted by BSA. The choice of annuloplasty performed was determined by the BSA of the child. A Wooler-Kay annuloplasty was performed on the very young with BSA less than 0.5m². The Wooler-Kay annuloplasty consists of single mattress sutures reinforced with pledgets at the commissural sites.¹⁴ Other suture plication techniques were used between 0.5 and 0.9m². In a Paneth annuloplasty, a double-armed, pledgeted 2-0 polypropylene suture was placed at the margin of the central fibrous body, sutured circumferentially around the annulus in 2- to 3-mm steps to

the posterior portion of the annulus, and tied over a second pledget. A second suture was placed on the opposite side of the annulus.¹⁵ The De Vega annuloplasty was performed in two sections with a continuous suture that involved the entire ventricular free ventricular wall, with pledgets placed at both commissures.¹⁶ Complete annuloplasty rings were not used. A posterior annuloplasty ring was used in children with a mean BSA greater than 0.9m².¹³ An incomplete annuloplasty ring was used in 4 cases. In 1 case, a Puig-Massana ring was used, whereas the Mitral Repair System (MRS; Kohler Chemie, Alsbach-Hahnlein, Germany) ring was used in the other 3. Anticoagulation was not used in any patient after mitral reconstruction. Splitting and fenestration of interchordal spaces were performed in cases with single papillary muscle. Chordal transfer techniques or artificial replacement were not used. The techniques used in the repair of mitral disease varied according to morphology (Table 2). Inhaled nitric oxide has been routinely available in our unit since 1992. The 2 patients with anomalous left coronary artery underwent direct aortic reimplantation of the artery and a modified De Vega mitral annuloplasty. When valve replacement is required, it has been our current practice to use a St Jude Medical device (St Jude Medical, Inc, Minneapolis, Minn). To date, the youngest patient at implantation has been a 2 year old and a total of three devices have been placed in our unit during this period. Extracorporeal membrane oxygenation was not used to salvage any patient in this series.

Data Analysis

Statistical analysis was performed with SPSS version 12.0 software (SPSS Inc, Chicago, Ill). Actuarial analysis of events was made with the Kaplan-Meier method (Figure 1). Comparisons between groups with nominal data were made with a χ^2 test, and those of parametric data with a Student *t* test.

Results

Mortality

The in-hospital (30-day) mortality in group 1 was 3.3% (1/30). Group 2 had a significantly higher in-hospital mortality, with 4 deaths (26.6%, $\chi^2_1 = 5.5125$, *P* = .019). The

death in group 1 was that of a 9-month-old boy with mitral regurgitation, a single papillary muscle, and pulmonary hypertension. He died of a tachyarrhythmia (junctional ectopic tachycardia) on postoperative day 1. The 4 deaths in group 2 were those of 2 boys and 2 girls: a 20-month-old boy with a preoperative mean pulmonary pressure of 53 mm Hg who had a postoperative hypertensive crisis, a 41-month-old girl with complex LVOTO and mitral incompetence who underwent a Paneth type repair and aortic valvotomy and died when brought back to theatre the next day for further management of residual LVOTO, a 2-month-old boy with anomalous left coronary artery and ischemic mitral incompetence who underwent reimplantation of the left coronary artery and a De Vega repair of the incompetent valve and died as a result of low output syndrome on day 5 while being weaned from ventilation, and a 7-month-old girl who underwent a commissurotomy and papillary muscle split for a parachute mitral valve and also VSD closure and died as a result of a tachyarrhythmia (junctional ectopic tachycardia) at postoperative 2 days. In-hospital mortality was not significantly associated with type of mitral pathology, preoperative pulmonary pressures, extracorporeal circulation duration, or type of cardioplegia used. Although there was a significant difference in age and weight between groups 1 and 2 (Table 3), analysis of age ($P = .132$) and weight ($P = .097$) separately from anatomic factors discerned no significant relationship between age and risk of 30-day mortality, although 3 of the in-hospital deaths were among the 18 children younger than 1 year at the time of surgery (16.6%). There were no early deaths (30 days to 1 year). The only late death (>1 year) was that of a female patient 13 months after initial mitral repair who required reoperation for a second reconstruction and died of a tachyarrhythmia associated with a postoperative low output syndrome. This patient had mitral stenosis as the dominant pathology. In group 1, the 15-year survival was 93%, versus 73% for group 2. The overall population survival at 15 years was 86.5%.

Morbidity

Nonfatal complications included the case of a patient who was brought back to the operating room for management of postoperative bleeding. One patient required a permanent pacemaker for complete heart block, and another had a chylothorax develop. Seven patients (4 with mitral incompetence, 3 with stenosis) required 9 revision surgical procedures. The first repair was performed within 30 days of the primary procedure in 3 cases. These three cases predated the availability of intraoperative transesophageal echocardiography in our unit. In 2 cases the dominant mitral pathology was incompetence, and in 1, stenosis. In the mitral stenosis case and one of the incompetence cases, there was significant postoperative mitral regurgitation requiring rein-

tervention. In this incompetence case, further repair was successful; in the mitral stenosis case, however, valve replacement was required. The third patient was brought back on day 1, not to address mitral pathology but rather to further address LVOTO (subaortic stenosis), which had become more significant in the presence of a competent mitral valve. Other required reoperations were performed within 1 year in 1 case and between 1 and 4 years in the remaining 3 cases. All 9 procedures were performed within 4 years of the primary mitral repair. There was no significant difference between groups 1 and 2 in reoperation rate (Table 3). Patient age at operation, type of cardioplegia, and dominant type of mitral pathology were not associated with failure of repair. Two mitral valve replacements were required at reoperation, 1 in a patient with stenosis and 1 in a patient with incompetence (25-mm CarboMedics valve; and 19-mm St Jude Medical valve). In group 1, the 10-year freedom from reoperation was 82.9% versus 72.7% for group 2. The overall freedom from reoperation at 15 years was 81.7%. There were no detected incidences of thromboembolism, anticoagulation-related hemorrhage, endocarditis, or sudden death. The event-free rate at 15 years was 73.5%.

Previous AVSD Repair

Separate analysis of the group undergoing mitral repair after a previous AVSD reconstruction was performed. This group consisted of 7 patients with a mean age of 62 months. There were three boys. There were no deaths and only 1 reoperation. One patient was in group 2 with subaortic stenosis. The patient with reoperation underwent two further procedures and ultimately a mitral valve replacement. The 10-year survival therefore was 100%, with a freedom from reoperation of 85.7%. Among the children without previous AVSD surgery, there were 38 patients with a mean age of 44.7 months (19 boys). This latter group had a 10-year survival of 83.9% and a freedom from reoperation of 81.2%.

Discussion

The results of mitral reconstruction in children with acquired mitral incompetence are excellent, with a reported 10-year survival of 90%.¹⁷ However, congenital mitral anomalies represent a more complex population with a high prevalence of associated cardiac anomalies. During the 18-year experience from a single operator presented here, mitral valve reconstruction techniques learned from adult practice were successfully applied to the pediatric setting. Mitral valve repair was performed in all cases at the first surgical intervention, yielding survival and freedom from reoperation comparable with those of other published series (Table 4).

Mitral valve repair has many advantages relative to replacement. Prosthetic replacement-associated problems

TABLE 4. Demographic, operative, and outcome data from patients grouped by presence or absence of significant associated intracardiac disease at the time of mitral valve surgery

Reference	Period (y)	Age <1 y	Mean age (y)	Median age (y)	Valve repair	Valve replacement
Current series	18	40%		2.16	45	0
McCarthy et al, ¹ 1996	10	43.4%	3.2		23	0
Serraf et al, ² 2000	18	44%		1.5	72	0
Prifti et al, ⁴ 2002	10	27%	5.2		94	0
Stellin et al, ⁵ 1988	14	16.6%		6	26	4
Chauvaud, et al, ⁶ 1998	25	—*	5.7		138	7
Zias et al, ⁷ 1998	7	—*	6.0		26	0
Aharon et al, ⁸ 1994	11	24% < 2 y	4.9		79	0
Stellin et al, ⁹ 2000	12	11.7%	5.9		34	0
Uva et al, ¹⁰ 1995	13	100%	0.55		19	1
Yoshimura et al, ¹¹ 1999	20	—*	3.6		36	20
Caldarone et al, ¹⁹ 2001	17	38.8%		1.42	0	139
Eble et al, ²² 2003	18	20.7%		5.0	0	53
Kadoba et al, ²³ 1990	14	100%	0.58		0	25
Lorier et al, ²⁸ 2001	23	—*	4.6		21	0
Moran et al, ²⁹ 2000	11	—*		2.8	37	9
Okita et al, ³⁰ 1988	17	12% < 2 y	5.5		66	0
Erez et al, ³¹ 2003	20	—*	8.1		0	90

Patients with associated intracardiac disease (group 2) were significantly younger and smaller and had higher early mortality and longer intensive care stay. *E*, Endocarditis; *VT*, valve thrombosis; *SCD*, sudden cardiac death; *TE*, thromboembolic event; *CVA*, cerebrovascular accident. *Data not available.

with hemolysis, perivalvular leak, valve thrombosis, thromboembolism, and endocarditis are avoided or minimized. Repair conserves the chordae and subvalvular apparatus, preserving normal left ventricular geometry and function.¹⁸ Conservation of the normal geometry preserves left ventricular function and leads to survival benefits in the long term.³ Valve replacement is difficult in infants younger than 1 year, as was the case in 40% of our series population. This is because of size mismatch between the prosthesis and annulus, which interferes with cardiac function, distorting the geometry of the cavity and frequently obstructing the left ventricular outflow tract.¹⁹ This may be minimized with supra-annular placement of the prosthesis, but the problem remains that these very young children will quickly outgrow the prosthetic valve, requiring reoperation, and the long-term survivorship has yet to be confirmed. Annular dilation was the major factor in mitral incompetence in 36.6% of cases in this series. In these cases, repairs with rigid annuloplasty rings were avoided, particularly in infants younger than 1 year. This was done to avoid interference with valve annulus growth and the complications of endocarditis and thromboembolism. In most cases a commissural plication annuloplasty technique to correct annular dilatation was used. However, in 4 cases annuloplasty rings were required. Annuloplasty rings are associated with interference in the normal cardiac wall motion and function, but incomplete

rather than complete rings were used to minimize any restriction of movement of the anterior mitral leaflet and trigone area.²⁰ All 4 patients survived, and none have yet required revision surgery. The management of anticoagulation in a pediatric population is challenging for child, parent, and physician. An incidence of serious hemorrhage of 9% and a 12% incidence of thromboembolic events were reported in a review of children anticoagulated with warfarin after cardiac valve replacement.²¹ This is avoided with mitral repair, which obviates the need for anticoagulation.

In considering alternatives to repair, data regarding mortality, freedom from reoperation, and event-free survival are essential (Table 4). Most series report significant mortality from mitral valve replacement, with death or transplant rates of 36% when mechanical valves were used exclusively²² and 47% to 56% from populations of mixed bioprosthesis and mechanical valves^{23,24} where the 5-year survival was reported as 43%.²³ In contrast, one Japanese center has reported no in-hospital mortality for mechanical valve replacement in children and a 10-year survival of 90.3%.¹¹ However, the group's freedom from reoperation at 10 years was similar to other studies at 66% to 67.3%.^{11,22} The 15-year freedom from reoperation rate in this series of mitral valve reconstructions, 81.7%, compares favorably with the rates after valve replacement. Although biologic valve replacements have been used, porcine mitral biopros-

In-hospital death	Late death	Reoperation	Survival	Freedom from reoperation	Adverse events
5	1	7	86.5% at 15 y	81.7% at 15 y	0
3	2	3	77.4% at 7 y	84.1% at 7 y	0
9	8	24	69.6% at 15 y	70.8% at 15 y	2 E, 2 VT
8	9	6	89.2% at 5 y	76.3% at 5 y	0
5	1	3	—*	—*	1 SCD
7	10	21	Repair 88% at 10 y, replacement 51% at 10 y	Repair 67.7% at 15 y, replacement —*	2 TE, 1 CVA, 1 E, 1 SCD
1	0	4	—*	—*	0
3	3	10	82% at 5 y	83% at 8 y	1 TE
0	1	3	96.8% at 12 y	85.9% at 12 y	0
0	1	11	94% at 7 y	58% at 7 y	2 VT
2	2	6	Repair 87.2% at 10 y, replacement 90.3% at 10 y	Repair 72.7% at 10 y, replacement 67.3% at 10 y	3 VT, 1 valve dysfunction
25	4	32	74% at 10 y	81% at 5 y	8 E, 4 VT, 3 CVA
6	8	18	—*	66% at 10 y	6 VT, 1 CVA
9	5	9	43% at 5 y	45% at 3 y	0
2	0	4	—*	46 % at 10 y	0
1	4	*	Repair 92.6% at 10 y, replacement 85.7% at 10 y	Repair 78.5% at 9 y, replacement 85.7% at 9 y	1 E
1	4	*	88.4% at 17 y	86% at 17 y	0
17	4	12	70.5% at 10 y	—*	6 TE, 9 bleeding events

theses in the pediatric population are associated with accelerated tissue calcification and failure.²⁵ This poorer performance with biologic valves is also consistent with reports of high early mortality and 5-year failure from adult use of mitral homograft valves.²⁶ Gulbins and colleagues,²⁷ again in an adult population, report that 3 of 8 complete homografts required replacement within 3 years of surgery. Among the published articles reviewed, only Chauvaud and associates⁶ commented on event-free survival, reporting a 69% event-free rate at 10 years.

Few reports on congenital mitral valve anomaly repair have identified predictors of poor outcome. This may reflect the small numbers generally found in such series. The major factor associated with increased in-hospital mortality in this series was the presence of associated significant intracardiac disease. This group was also found to be significantly younger and hence lighter at the time of operation, but analysis of age or weight separately from the anatomic division between groups 1 and 2 showed no significant relationship between age or weight and outcomes. The cardiopulmonary bypass time was longer in the groups with complex anatomic pathology, which could be explained by the time required to address the associated pathology. However, crossclamp time was not significantly longer. The longer cardiopulmonary bypass time is more likely to reflect the practice of using profound hypothermia in patients with

more complex problems. Other groups have also observed poorer survival in cases with associated cardiac anomalies.^{2,4} The associated intracardiac anomalies identified in this series as high risk consisted of concurrent repair of VSD, LVOTO, and anomalous coronary artery. The increased risk may be due to more complex anatomy, LVOTO, VSD, and the presence of impaired ventricular function, which struggles postoperatively with corrected circulatory flow. In considering whether any associated intracardiac disease should be addressed before mitral valve repair, the combined mortality of two separate procedures would have to be superior, and Serraf and coworkers² reported a better outcome when all anomalies were repaired in a single stage rather than in a two-stage fashion. In studies of mitral valve replacement, the outcome of patients with significant associated cardiac disease has not been analyzed separately, so there are no comparative data enabling a true analysis of whether this group would have superior outcomes with valve replacement at first attempt. This would seem unlikely, however, because the placement of a mechanical valve is unlikely to offer benefits relative to repair in a situation of poor left ventricular function or to reduce the operative times significantly. Other reported predictors of poor outcome include age younger than 1 year, which was not statistically significant in this study, hammock mitral valve, and cardiomegaly.^{2,4} No significant fac-

Figure 1. Kaplan-Meier curve shows survivals and event-free rates for 18-year period. Cumulative reoperation rates (numbers in parentheses) are also shown.

tors in this study were found for failure of repair and reoperation, although mitral stenosis is reported to have a higher reoperation requirement.^{4,9}

The in-hospital death rate reported in this study is higher than in some recent reports (Table 4). We believe this is related to the high proportion of cases with associated significant intracardiac disease (33.3%), which we have identified as a mortality risk factor. In addition, there was a large proportion of patients younger than 1 year (40%) and with stenosis (33.3%) which, although not statistically significant factors in this study, are factors that have been associated with poorer outcome in other studies.⁴ This series covers a longer period than many contemporary reports, and differences in mortality compared with shorter recent studies may reflect improvements in preoperative optimization, intraoperative cardiac protection, cardiopulmonary bypass management, and postoperative intensive care management, especially in relation to the pharmacologic management of pulmonary hypertension.

Conclusion

This review covers a significant follow-up period, and only a few published reports cover similar periods.^{6,11,28} Higher mortality with repair was observed when associated with significant intracardiac disease. Whether this group would benefit from mitral valve replacement at the first attempt has yet to be determined. Every effort should be made to preserve the natural valve, even when very dysplastic, by reconstructive techniques. Our experience with mitral valve repair rather than replacement as the first surgical intervention has proved it to be a reliable tool and satisfactory alternative to replacement of the valve, with no other events other than reoperation. Later valve replacement is still possible, but it is at least postponed, preferably until adulthood,

when the problems of mechanical valve size and growth are less significant and anticoagulation can be managed independently.

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