Early and Mid-Term Outcome of Pediatric Congenital Mitral Valve Surgery

Ramin Baghaei [1]; Avisa Tabib [2,*]; Farshad Jalili [1]; Ziae Totonchi [1]; Mohammad Mahdavi [1]; Behshid Ghadrdoost [1]

[1]Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, IR Iran
[2]Heart Valve Disease Research Center, Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, IR Iran

Corresponding author: Avisa Tabib, Heart Valve Disease Research Center, Rajaie Cardiovascular Medical and Research Center, Vali-Asr St., Niayesh Blvd, Tehran, IR Iran. Tel: +98-2123922199, Fax: +98-2122663213, E-mail: avtabib@yahoo.com

Background: Congenital lesions of the mitral valve are relatively rare and are associated with a wide spectrum of cardiac malformations. The surgical management of congenital mitral valve malformations has been a great challenge.

Objectives: The aim of this study was to evaluate the early and intermediate-term outcome of congenital mitral valve (MV) surgery in children and to identify the predictors for poor postoperative outcomes and death.

Patients and Methods: In this retrospective study, 100 consecutive patients with congenital MV disease undergoing mitral valve surgery were reviewed in 60-month follow-up (mean, 42.4 ± 16.4 months) during 2008 - 2013. Twenty-six patients (26%) were under one-year old. The mean age and weight of the patients were 41.63 ± 38.18 months and 11.92 ± 6.12 kg, respectively. The predominant lesion of the mitral valve was MV stenosis (MS group) seen in 21% and MR (MR group) seen in 79% of the patients. All patients underwent preoperative two-dimensional echocardiography and then every six months after surgery

Results: Significant improvement in degree of MR was noted in all patients with MR during postoperative and follow-up period in both patients with or without atrioventricular septal defect (AVSD) (P = 0.045 in patients with AVSD and P = 0.008 in patients without AVSD). Decreasing trend of mean gradient (MG) in MS group was statistically significant (P = 0.005). In patients with MR, the mean pulmonary artery pressure (PAP) had improved postoperatively (P < 0.001). Although PAP in patients with MV stenosis was reduced, this reduction was not statistically significant (P = 0.17). In-hospital mortality was 7%. Multivariate analysis demonstrated that age (P < 0.001), weight (P < 0.001), and pulmonary stenosis (P = 0.03) are strong predictors for mortality. Based on the echocardiography report at the day of discharge from hospital, surgical results were optimal (up to moderate degree for MR group and up to mild degree for MS group) in 85.7% of patients with MS and in 76.6% of patients with MR. Age (P = 0.002) and weight (P = 0.003) of patients are strong predictors for surgical success in multivariate analysis.

Conclusions: Surgical repair of the congenital MV disease yields acceptable early and intermediate-term satisfactory valve function and good survival at intermediate-term follow-up. Strong predictors for poor surgical outcome and death were age smaller than 1 year, weight smaller or equal than 6 kg, and associated cardiac anomalies such as pulmonary stenosis.

Keywords: Congenital Heart Defect; Mitral Regurgitation; Mitral Stenosis; Pediatrics

1. Background

Congenital lesions of the mitral valve (MV) are relatively rare and are associated with a wide spectrum of cardiac malformations (1). Isolated congenital MV disease is uncommon, occurring in approximately four of 1000 children with congenital heart disease (2), but malformation of the MV often coexist with other congenital heart lesions, especially those of the left heart chambers. The anomaly of the valve may involve any component of the MV apparatus that result in stenosis, regurgitation, or mixed hemodynamic disturbances. Congenital MV stenosis (MS) may result from supravalvular ring, annular and leaflet hypoplasia, or subvalvular obstruction due to anomalies of the papillary muscle or chordae (3). Congenital MR (MR) may result from annular dilatation, leaflet prolapse, chordal shortening, or clefts (3).

For many years, the surgical management of congenital MV malformations has been a great challenge. Severe symptoms and signs of pulmonary hypertension is the definitive indication for surgery. Operation is indicated when pulmonary hypertension is severe, even in the absence of symptoms. Another indication of surgery is when moderate or severe MS or MR coexists with other important congenital cardiac lesions (3). Based on the difficulties of the valve replacement in children, MV repair remains the most acceptable option. There are several reports that demonstrate excellent results of surgical repair in complex congenital MV lesions (4-7). Recent studies demonstrated improved result and long-term freedom from reoperation among pediatric patients undergoing MV surgery (1, 8, 9). We have studied our five-year experience with congenital MV surgery.

2. Objectives

The aim of our study was to determine the early and intermediate-term outcome of patients undergoing MV surgeries as well as the risk factors of poor surgical outcome.

3. Patients and Methods

3.1. Patient Population

From May 2008 to December 2013, 100 patients were operated for congenital MV disease in our department by the same surgical team. The local ethical committee approved this retrospective study.

There were 47 males (47%). The mean age and weight of the patients were 41.63 ± 38.18 months (range, 1 - 156 months) and 11.92 ± 6.12 kg (range, 3 - 30 kg), respectively. Twenty-six patients (26%) were under one year old. There were important associated cardiac lesions in 66% of cases (Figure 1) and 24 patients had overt symptoms related to their diseases.

All patients underwent two-dimensional echocardiography with color flow Doppler study preoperatively. Cardiac catheterization and angiography was performed in 70% of patients. Although some patients had mixed MV lesions, they were divided into two groups based on predominant lesion: MS in 21% and MR in 79% of the patients. The MR group consisted of patients with atrioventricular septal defect (AVSD), who required reparative procedures other than cleft closure, and patients without AVSD. Table 1 demonstrates preoperative demographic and clinical characteristics of patients.

3.2. Surgical Technique

Aortobicaval cannulation with mild systemic hypothermia (32°C) was used in all cases. The method of myocardial protection was antegrade blood cardioplegia. The exposure of the MV was through either a left atriotomy parallel to the interatrial groove, or interatrial septum, depending on the associated procedures. The leaflets and subvalvular apparatus were carefully examined. Cold saline was injected to the LV cavity to assess the leaflet motion and coaptation. Based on the mechanism of the disease and anatomical considerations, one or more of the repair techniques were used. The efficacy of corrective procedure was assessed by pressure monitoring of the left atrium as well as transesophageal echocardiography (TEE) whenever feasible. Weaning of cardiopulmonary bypass (CPB) was usually achieved by combined inotropes (adrenaline, 0.05 - 0.1 µg/kg/min; and milrinone, 0.25 - 0.5 µg/kg/min).

3.3. Echocardiographic Evaluation

Our method for measuring the echocardiographic parameters was based on the following criteria:

The MR severity (10) was graded as follows in the Table 2.

3.3.1. Mitral Stenosis

We used mean gradient (MG) of MV for grading of MS. It is an invaluable measurement in pediatric patients. The MS severity was graded as follows (11):

Mild MS, MG of < 5 mmHg; moderate MS, MG of 5 to 10 mmHg; and severe MS, MG of > 10 mmHg. The TR severity (12) was graded as follows in the Table 3.

According to the guideline, severity of MR and tricuspid regurgitation (TR) were categorized as mild, moderate, and severe, but in cases that the patient had all criteria of a certain grade in addition to one criteria of next grade, severity of regurgitation was defined as mild to moderate or moderate to severe.

3.3.2. Mean Pulmonary Arterial Pressure

The mean pulmonary artery pressure (PAP) was estimated by pulmonary regurgitate jet, as highest protodiastolic gradient, plus estimated right atrium pressure (13).

Table 1. Preoperative Demographic and Clinical Characteristics [a]

Variables	Values
Age	
Mean age	41.6 ± 38.1
Range, mo	1 - 156
Patients ≤ 12 months old	26 (26)
Male/female	47 : 53
Weight	
Mean weight, kg	11.9 ± 6.1
Range, kg	3 - 24
Mean body surface area, m²	0.55 ± 0.21
Mitral stenosis (MS group)	21
Mitral regurgitation (MR group)	79
AVSD	41 (51.9)
Non-AVSD	38 (48.1)
Symptoms (CHF)	
Asymptomatic	76
Mild	11
Moderate to severe	13
Co-existing cardiac lesions	66
Moderate to severe regurgitation	70 (70)
Moderate to severe stenosis	42 (42)
Mean preoperative ejection fraction	60.92 ± 8.11

[a] Data are presented as Men ± SD or No (%) or %.

Table 2. Severity of Mitral Regurgitation [a]

	Severity Mitral Regurgitation		
	Mild	**Moderate**	**Severe**
LA size	Normal	Normal or dilated	Usually dilated
LV size	Normal	Normal or dilated	Usually dilated
Color flow jet area	Small central jet < 20% of LA area	Sign of MR > mild present but no criteria for severe MR	Large jet > 40% LA area or variable size wall impinging jet swirling in LA
Jet density-CW	Incomplete or faint	Dense	Dense
Pulmonary vein flow	Systolic dominance	Systolic blunting	Systolic flow reversal

[a] Abbreviations: LA, left atrium; LV, left ventricle; MR mitral regurgitation; CW, continuous wave Doppler.

Table 3. Severity of Tricuspid Regurgitation [a]

	Severity of Tricuspid Regurgitation		
	Mild	**Moderate**	**Severe**
Color flow jet	Small central	Intermediate	Very large or eccentric wall impinging jet
CW signal of tricuspid regurgitation jet	Faint	Dense	Dense
Hepatic vein flow	Systolic dominance	Systolic blunting	Systolic Flow reversal

[a] Abbreviation: CW, continuous wave Doppler.

3.4. Follow up

All patients were assessed by transthoracic echocardiography (TTE) immediately on arrival to the intensive care unit (ICU) and before discharge from hospital. All surviving patients were examined periodically in the clinic and underwent TTE (by the same physician) every six months after surgery.

3.5. Statistical Analysis

Statistical analysis was performed with SPSS 15 for Windows (SPSS Inc, Chicago, Illinois, the United States). Data were expressed as mean ± SD for interval and frequency (percentage) for categorical variables. All variables were tested for normal distribution with Kolmogorov-Smirnov test. Categorical values were compared by Chi square test or Fisher's exact test. Independent-samples t test or Mann-Whitney U test were used to compare the mean variables between two groups. Continuous variables were compared using the two-tailed Wilcoxon test. Repeated measures ANOVA followed by Bonferroni post hoc test was used to assess parametric distributions. For nonparametric distributions, Friedman test was applied. P values < 0.05 were considered statistically significant.

4. Results

Patient demographics and baseline data are summarized in Table 3. Fourteen patients (14%) weighted less than 6 kg and 26 children (26%) aged less than 1 year. Overall, 21% of patients had MS and 79% had MR.

The key point of MV repair is the mechanism of the stenosis or regurgitation of the valve. The characteristics of the lesions responsible for hemodynamic dysfunction of MV are shown in Table 4. There were 102 lesions in 79 patients in MR group. The most common responsible lesions were anterior leaflet cleft (44.1%) and annular dilatation (42.1%). The high percentage of leaflet cleft may reflect the significant number of AVSD patients among the MR group. There were 33 lesions in 21 patients in MS group. In this group, the most common lesion was complete or incomplete supramitral fibrous ring (45.5%). In fact, multiple lesions can be responsible for stenosis or regurgitation of the valve in a single patient. The frequency of this figure was 39.2% in MR group and 38% in MS group.

Table 4. Classification of Responsible Lesions for Valvular Hemodynamic Dysfunction

Mitral Valve Malformation	Number of lesion
Mitral regurgitation group	102
Anterior leaflet cleft	45
Annular dilatation	43
Leaflet prolapse	11
Retracted leaflet	2
Leaflet perforation	1
Mitral stenosis group	33
Complete/incomplete supramitral ring	15
Commissural fusion	9
Leaflet and subvalvular fibrosis	7
Parachute mitral valve	2

There were 156 associated cardiac lesions in 66 patients (Figure 1): 19 patients (90.5%) in MS group and 61 (77.2%) in MR group. There was no statistically significant association between associated cardiac lesions and MS or MR (P = 0.17). Ventricular septal defect (VSD) (62%) and atrial septal defect (ASD) (50%) were the most commonly associated lesions. Most patients (76%) were asymptomatic preoperatively.

The mean CPB time and cross-clamp time were 95 and 74 minutes, respectively. Different techniques were used in these patients (Table 5). The most commonly used technique were cleft closure and suture annuloplasty/commissuroplasty for MR group and commissurotomy ± subvalvular resection for MS group. All patients were assessed by TTE immediately on arrival to the ICU. The mean ICU stay time was 3.97 ± 2.98 days in MR group and 3.90 ± 2.36 days in MS group. There was no significant differences between two groups in length of ICU stay (P = 0.75). The mean follow-up period was 42.5 ± 16.4 months (range, 2 - 60 months). We could achieve complete follow-up in 89% of surviving patients. The freedom from reoperation was 100%. The freedom from recurrence was 83% in MS group and 75% in MR group.

Changes in the echocardiography parameters, from the preoperative period to the end of follow-up, are shown in Table 6. There were significant differences between preoperative and postoperative ejection fraction (EF) at ICU in MR group (P < 0.001), but there were no significant differences between preoperative and postoperative EF at ICU in MS group (P = 0.54). Trend of changes in EF during follow-up was not significant in any of the groups (P = 0.71 in MS group vs. P = 0.53 in MR group).

Figure 1. Associated Cardiac Lesions in Patients With Mitral Valve Lesions
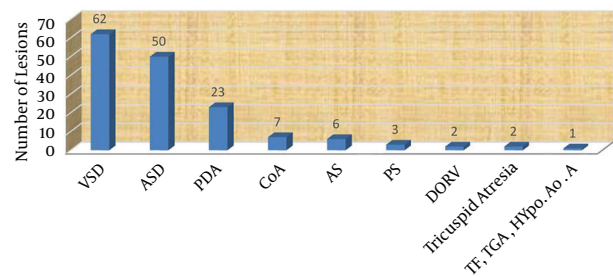
Abbreviations: AS, aortic stenosis; ASD, atrial septal defect; CoA, coarctation of aorta; DORV, double outlet right ventricle; Hypo. Ao. A, hypoplastic aortic arch; PDA, patent ductus arteriosus; PS, pulmonary stenosis; TF, tetralogy of Fallout; TGA, Transposition of the great arteries; VSD, ventricular septal defect.

Table 5. Different Techniques of Surgical Repair of Mitral Valve Lesions [a]

Repair Technique	Frequency
Cleft closure	41
Suture annuloplasty/ commissuroplasty	40
Posterior pericardial band annuloplasty	27
Commissurotomy ± Subvalvular resection	16
Neochordal insertion	12
Supramitral ring resection	11
Papillary muscle splitting	5
Quadrangular resection	4
Leaflet extension with pericardial patch	2
Prosthetic ring annuloplasty	2

[a] Data are presented as %.

Table 6. Echocardiographic Variables Changes During Sixty Month Follow-up [a,b]

	Pre-op	ICU	6 m post op	12 m post op	24 m post op	36 m post op	48 m post op	60 m post op	P Value
EF									
MS	63.52 ± 9.0	62.19 ± 7.0	65.53 ± 6.1	64.36 ± 6.4	64.29 ± 4.3	64 ± 5.6	64.38 ± 5.6	63.75 ± 7.5	0.71
MR	60.21 ± 7.7 [c]	55.77 ± 10.5 [c]	59.98 ± 7.1	60.81 ± 5.3	61.33 ± 5.6	64.80 ± 6.0	66.67 ± 7.5	64.17 ± 5.8	0.53
MG, mmHg									
MS	15.05 ± 8.0 [c]	5.12 ± 1.8 [c]	5.15 ± 1.4 [c]	4.08 ± 2.1	4.03 ± 2.2	3.47 ± 1.2	4.50 ± 1.5	5.00 ± 2.4	0.005
MR	6.93 ± 12.6 [c]	3.54 ± 2.9 [c]	3.64 ± 2.8 [c]	3.66 ± 2.7	3.78 ± 2.7	3.34 ± 3.8	4.79 ± 6.7	5.50 ± 7.2	0.553
TR									
MS	2 (3 - 4)	1.5 (1 - 2)	1 (1 - 2)	1 (1 - 2)	2 (1 - 2)	1.5 (1 - 2)	1.5 (1 - 2)	2 (1 - 2)	0.945
MR	5 (5 - 6)	2 (2 - 3)	1.5 (1 - 2)	3.5 (1 - 6)	2 (1 - 3)	3 (0 - 6)	1.5 (1 - 2)	1.5 (1 - 2)	< 0.001
TR									
AvCanal+	4 (4 - 5)	2 (2 - 3)	2 (2 - 3)	2 (2 - 3)	2 (2 - 3)	2 (2 - 3)	2 (2 - 3)	2 (2 - 3)	< 0.001
AvCanal-	3 (2 - 3)	2 (2 - 3)	1 (1 - 2)	2 (1 - 2)	1 (1 - 2)	1 (1 - 2)	1 (1 - 2)	2 (1 - 2)	0.073
MR									
AvCanal+	6 (5 - 6)	2 (2 - 3)	2 (2 - 3)	2 (2 - 3)	2 (2 - 3)	3 (2 - 3)	2 (2 - 3)	1 (1 - 2)	0.045
AvCanal-	6 (5 - 6)	2 (2 - 3)	2 (2 - 3)	2 (2 - 3)	2 (2 - 3)	2 (2 - 3)	2 (2 - 3)	1 (1 - 2)	0.008

[a] Abbreviations: EF, ejection fraction; MG, mean gradient; MR, mitral regurgitation; Pre-op, preoperation; post-op, postoperation; TR, tricuspid regurgitation.
[b] Degree of MR or TR severity: 1, trivial; 2, mild; 3, mild to moderate; 4, moderate; 5, moderate to severe; 6, severe.
[c] Statistically significant in paired-wise comparison (P < 0.05).

Decreasing trend of MG in MS group was statistically significant (P = 0.005). In patients with MR, the mean PAP was improved to 28.40 ± 16.61 mmHg compared with 37.63 ± 17.48 mmHg at baseline (P = < 0.001), but in patients with MS, the mean PAP was not changed significantly (46.76 ± 18.17 mmHg before operation vs. 29.25 ± 21.04 mmHg after operation; P = 0.17). Although reduction of PAP in patients with MS was clinically important, this reduction was not statistically significant. It seems that this result is due to the low number of patients with MS.

Significant improvement in the degree of MR was noted in all patients with MR during postoperative and follow-up period. Postoperative echocardiography showed that the prevalence of severe MR decreased to 17% while before surgery 58% of patients had severe MR. There were mild or moderate residual MR in 61 patients (62.3%) in immediate echocardiographic assess at ICU; however, there was no progressive MR or evidence of excessive MV annular dilatation during the follow-up period in these patients. Indeed, 28% of the patients maintained the same MR grade during the follow-up period while the severity decreased in 60% of patients with MR. In this group, grade of regurgitating decreased significantly in both patients with AVSD (P = 0.045) or without AVSD (P = 0.008). Patients with MR and AVSD showed significant reduction in the TR severity (P < 0.001). Severity of TR in these patients decreased in follow-up echocardiography compared with immediate postoperative measurements. The TR severity further decreased in the patients who did not have AVSD; however, this change was not statistically significant.

Based on the report of echocardiography at the day of hospital discharge, surgical results were optimal (up to moderate degree for MR group and up to mild degree for MS group) in 85.7% of patients with MS and in 76.6% of patients with MR.

Univariate analysis of the preoperative variables demonstrated that age < 1 year old (P = 0.05) and severity of symptoms (P = 0.008) had statistically significant association with optimal surgical results (Table 7). Age (β, 2.45; 95% CI, 2.51 - 54.3; P = 0.02) and weight (β, -0.16; 95% CI, 0.76 - 0.94; P = 0.03) were strong predictors for surgical success in multivariate analysis.

Table 7. Comparison Between Failed and Successful Surgical Results [a,b]

Variables	Successful (n = 79)	Failed (n = 21)	P Value
Age, y			0.05
> 1	60 (77.9)	12 (57.1)	
≤ 1	17 (22.1)	9 (42.9)	
Sex			0.34
Male	35 (45.5)	12 (57.1)	
Female	42 (54.5)	9 (42.9)	
Weight	11.46 ± 4.95	13.81 ± 9.34	0.07
BSA	0.54 ± 0.18	0.59 ± 0.29	0.50
Diagnosis			0.36
MS	18 (23.4)	3 (14.3)	
MR	59 (76.6)	18 (85.7)	
MR			0.20
AVSD+	26 (44.1)	11 (61.1)	
AVSD-	33 (55.9)	7 (38.9)	
Kind of op			
Mitral cleft closure	38 (49.4)	9 (42.9)	0.59
Annuloplasty/pericardial Band	18 (23.4)	7 (33.3)	0.35
Repair commissuroplasty	31 (40.3)	9 (42.9)	0.83
Neochorda	11 (14.3)	1 (4.8)	0.23
Supramitral ring	14 (18.2)	2 (9.5)	0.34
MV commissurotomy	8 (10.4)	2 (9.5)	0.90
Annuloplasty ring	1 (1.3)	1 (4.8)	0.32
Patch Plasty	1 (1.3)	1 (4.8)	0.32
Symptoms (CHF)			0.008
Asymptomatic	63 (81.8)	12 (57.1)	
Mild	9 (11.7)	2 (9.5)	
Moderate to severe	5 (6.5)	7 (33.3)	
Co-existence anomaly			
VSD	51 (66.2)	10 (47.6)	0.11
ASD	40 (51.9)	7 (33.3)	0.13
COA/VSD	5 (6.5)	2 (9.5)	0.63
PDA	17 (22.1)	6 (28.6)	0.53
LSVC	3 (3.9)	2 (9.5)	0.29
PA banding	10 (13)	1 (4.8)	0.29
Severe AS	4 (5.2)	1 (4.8)	0.93

[a] Data are presented as mean ± SD or No. (%).
[b] Abbreviations: AS, aortic stenosis; ASD, atrial septal defect; BSA, body surface area; CoA, coarctation of Aorta; LSVC, left superior vena cava; PA banding, pulmonary artery banding; PDA, patent ductus arteriosus; VSD, ventricular septal defect.

All complication rates were 21%, almost one-third of which were severe enough to need specific treatment. Complications included gastrointestinal (10 patients), respiratory (9 patients), cardiovascular (7 patients), infectious (5 patients), neurologic (1 patient), and hematologic (1 patient) problems.

Any death that occurs during 30 days of hospitalization was defined as hospital mortality. Seven deaths occurred among patients (two in MS group and five in MR group). All patients except one (a 10-month-old boy with pure severe MS) had major associated cardiac defects. Four patients were < 6 months old, two were between six and 12 months old, and just one patient was > 12 months old. Regarding the immediate surgical result and based on echocardiography,

the result was suboptimal in just one case, a five-month old girl with multilevel left heart obstruction (Shoen's Complex). The cause of death was low cardiac output in three, multiorgan failure/sepsis in two, and pulmonary hypertension crisis in two patients. The mean of interval from surgery to death was six days. There was no late mortality. Several variables presumed to be the risk factor for survival were analyzed (Table 8). Among variables, age < 1 year (P = 0.004) and weight ≤ 6 kg (P = 0.003) had a significant association with mortality. Multivariate analysis showed that age (β, -0.16; 95% CI, 0.7 - 0.98; P = 0.03), weight (β, -0.65; 95% CI, 0.32 - 0.91: P = 0.02), and pulmonary stenosis (β, 4.48; 95% CI, 1.43 - 54; P = 0.03), as an important associated cardiac anomaly, were strong predictors for mortality.

Table 8. Association Between Some Variables and Survival After Mitral Valve Surgery [a,b]

Variables	Dead (n = 7)	Alive (n = 93)	P Value
Age, mo	12.14 ± 9.19	43.87 ± 38.63	0.003
Weight, kg	6.76 ± 2.31	12.31 ± 6.76	0.003
Diagnosis			0.65
MS	1 (14.3)	20 (21.5)	
MR	6 (85.7)	73 (78.5)	
MR			0.34
AVSD+	2 (33.3)	39 (53.4)	
AVSD-	4 (66.7)	34 (46.6)	
Operation Method			
Mitral cleft closure	3 (42.9)	45 (48.4)	0.77
Annuloplasty Pericardial Band	4 (57.1)	23 (24.7)	0.06
Repair commissuroplasty	1 (14.3)	39 (41.9)	0.15
Neochorda	0 (0)	12 (12.9)	0.31
Supramitral ring	0 (0)	16 (17.2)	0.23
MV commissurotomy	2 (28.6)	9 (9.7)	0.12
Annuloplasty ring	0 (0)	2 (2.2)	0.69
Patch Plasty	0 (0)	2 (2.2)	0.69
Symptoms of CHF			0.75
Asymptomatic	5 (71.4)	71 (76.3)	
Mild	1 (14.3)	10 (10.8)	
Moderate to severe	1 (14.3)	12 (12.9)	
Coexistence anomaly			
VSD	5 (71.4)	2 (28.6)	0.59
ASD	3 (42.9)	4 (57.1)	0.82
COA/VSD	1 (14.3)	6 (85.7)	0.43
PDA	3 (42.9)	4 (57.1)	0.19
LSVC	1 (14.3)	6 (85.7)	0.33
Severe PS	1 (14.3)	6 (85.7)	0.07
PFO	1 (14.3)	6 (85.7)	0.07
Length of ICU stay	3.29 ± 3.72	4.01 ± 2.79	0.07

[a] Abbreviations: ASD, atrial septal defect; AVSD, atrioventricular septal defect; CHF, congestive heart failure; COA, coarctation of aorta; ICU, intensive care unit; LSVC, left superior vena cava; MR, mitral regurgitation; MS, mitral stenosis; PFO, patent foramen ovale; PS, pulmonary stenosis; VSD, ventricular septal defect
[b] Data are presented as mean ± SD or No. (%).

5. Discussion

In this study, we described our five-year experience with congenital pediatric MV surgeries. There is quite a controversy in the surgery literature regarding the outcome of surgery in children, but the trend in the recent years has been very hopeful (14, 15).

The approach to the patient with congenital MV lesion consists of thorough preoperative evaluation. This evaluation should focus not only on anatomical diagnosis, but also on the mechanism of stenosis or regurgitation. Echocardiography has a determinative role in the diagnosis as well as planning for surgery. In other words, we could not achieve an acceptable surgical result and good postoperative outcome without a thorough preoperative understanding of the valve lesion (1).

One of the critical aspects of cardiac operations is the myocardial protection. We found no deterioration of myocardial systolic function as indicated by EF. The significant decrease of EF in MR group, based on immediate postoperative echocardiography, does not indicate deterioration of myocardial function. We know that in the presence of MR, EF is falsely high and its postoperative decrease is normally expectable. In all survived patients, EF remained unchanged throughout the follow-up period. The preservation of myocardial function in both MR and MS groups may reflect the optimal cardiac protection techniques.

The optimal outcome for MV repair is restoration of valvular function with minimal residual stenosis and regurgitation. In our study, up to moderate degree of residual regurgitation for MR group and up to mild degree of residual stenosis for MS group were defined optimal surgical outcomes. Improvement was noted in the severity of both regurgitation and stenosis during the 60 months after surgery.

In MR group, the severity of MR was significantly reduced not only in the postoperative period but also during the follow-up. Postoperative echocardiography showed that the rate of severe MR decreased significantly. There was no progressive or excessive MR during the follow-up period in most of patients with mild or moderate residual MR. In other words, the severity of MR improved in two-thirds of the patients and was not deteriorated during the follow-up period. Honjo et al. (16) showed that 90% of the patients who had undergoing MV repair during childhood, remained in the same MR grade during the follow-up period. By indicating the MV annulus grew along the normal growth curve after MV repair, they concluded that despite the presence of residual MR in some cases, their approach provided an almost appropriate growth pattern of the MV annulus regardless of the degree of residual MR.

The success rate of corrective surgery for MS is usually indicated by transvalvular MG. The MG improved significantly in patients who originally presented with MR (P = 0.005). The decreased MG was found by serial echocardiographic studies during the follow-up period. On the other hand, we did not find any significant change in MG across MV in MR group. This may reflect that corrective surgery in MR group could make acceptable competence of the valve without producing stenosis.

The reported mortality in our study was comparable with other same publications (14, 17, 18). In contrast with other studies, mortality rate was less in the MS group than in MR group. In MR group, patients who died had more severe MR. Studies demonstrated that postoperative degree of MR was a strong predictor of reoperation or death; this means that Grade III residual regurgitation on the contrary of the Grade I residual regurgitation was a strong risk factor for reoperation or death (1, 19, 20). The etiology of MR is an important factor for poor surgical outcome. Kalfa (21) showed that left ventricular outflow tract obstruction (LVOTO)-related etiology of MR was a significant risk factor for recurrent MR, MV reoperation, and MV replacement because the MR in this pathologic condition is only the marker of a global disease of the left heart.

Based on the multivariate analysis, age of < 1 year old was defined as the risk factor for death (P = 0.004). Many studies identified age < 1 year as a strong predictor for postoperative mortality. The associated heart defects also increased the rate of mortality due to significantly longer cardiopulmonary and aortic cross clamping times, complex used procedures, and complex anatomy (1, 21, 22).

Wood et al. emphasized on associated congenital anomalies as predictors of poor outcome and in-hospital mortality. The reported in-hospital death rate in their study was higher than that in other reports; therefore, they believed it was related to the high proportion of cases with associated intracardiac anomalies and a large number of patients younger than one year old with stenosis, which are factors that have been associated with poorer outcome in other studies (7).

In lee et al. (17) study on long-term results after MV repair in children no early death in was reported, but they had three mortalities in long-term; hence, they reported survival rate in their 15-year study to be up to 97%. Residual MR after first surgery was the most important risk factor for mortality.

In our study, we could show that surgical repair of the congenital MV disease yields acceptable early and intermediate-term valve function and good survival at intermediate-term follow-up. Strong predictors for poor surgical outcome and death were age of less than one year, weight ≤ 6 kg, and associated cardiac anomalies such as pulmonary stenosis.

5.1. Limitations

This study has several limitations. First, it was a retrospective study and all limitations of this kind of studies might interfere with the results. Second, both hemody-

namic lesion categories (MS and MR) were included and evaluated simultaneously. On the other hand, patients with AVSD who needed further reparative surgery were also included.

Acknowledgements

Authors' Contributions

Ramin Baghaei: conducting and management of the project; Avisa Tabib: revising the manuscript and final approval the article; Farshad Jalili: collecting the data and review of literature; Behshid Ghadrdoost: data analysis and preparing the manuscript; Mohammad Mahdavi and Zia Toutounchi: collecting the data.

Funding/Support

References

1. Prifti E, Vanini V, Bonacchi M, Frati G, Bernabei M, Giunti G, et al. Repair of congenital malformations of the mitral valve: early and midterm results. *Ann Thorac Surg.* 2002;**73**(2):614–21.
2. Allen HD, Driscoll DJ. In: *Moss and Adams. Heart Disease in infants, children and adolescents.* 7 editor. Philadelphia: LWW; 2008.
3. Kouchoukos N, Blackstone E. In: *Kirklin/Barrat-boyes Cardiac Surgery.* 4 editor. England: Churchill Livingstone; 2013.
4. Serraf A, Zoghbi J, Belli E, Lacour-Gayet F, Aznag H, Houyel L, et al. Congenital mitral stenosis with or without associated defects: An evolving surgical strategy. *Circulation.* 2000;**102**(19 Suppl 3):III166–71.
5. Moran AM, Daebritz S, Keane JF, Mayer JE. Surgical management of mitral regurgitation after repair of endocardial cushion defects: early and midterm results. *Circulation.* 2000;**102**(19 Suppl 3):III160–5.
6. Uva MS, Galletti L, Gayet FL, Piot D, Serraf A, Bruniaux J, et al. Surgery for congenital mitral valve disease in the first year of life. *J Thorac Cardiovasc Surg.* 1995;**109**(1):164–76.
7. Wood AE, Healy DG, Nolke L, Duff D, Oslizlok P, Walsh K. Mitral valve reconstruction in a pediatric population: late clinical results and predictors of long-term outcome. *J Thorac Cardiovasc Surg.* 2005;**130**(1):66–73.
8. Kadoba K, Jonas RA, Mayer JE, Castaneda AR. Mitral valve replacement in the first year of life. *J Thorac Cardiovasc Surg.* 1990;**100**(5):762–8.
9. Chauvaud S, Perier P, Touati G, Relland J, Kara SM, Benomar M, et al. Long-term results of valve repair in children with acquired mitral valve incompetence. *Circulation.* 1986;**74**(3 Pt 2):I104–9.
10. Baumgartner H, Hung J, Bermejo J, Chambers JB, Evangelista A, Griffin BP, et al. Echocardiographic assessment of valve stenosis: EAE/ASE recommendations for clinical practice. *J Am Soc Echocardiogr.* 2009;**22**(1):1–23.
11. Rudski LG, Lai WW, Afilalo J, Hua L, Handschumacher MD, Chandrasekaran K, et al. Guidelines for the echocardiographic assessment of the right heart in adults: a report from the American Society of Echocardiography endorsed by the European Association of Echocardiography, a registered branch of the European Society of Cardiology, and the Canadian Society of Echocardiography. *J Am Soc Echocardiogr.* 2010;**23**(7):685–713.
12. Lancellotti P, Moura L, Pierard LA, Agricola E, Popescu BA, Tribouilloy C, et al. European Association of Echocardiography recommendations for the assessment of valvular regurgitation. Part 2: mitral and tricuspid regurgitation (native valve disease). *Eur J Echocardiogr.* 2010;**11**(4):307–32.
13. Zoghbi W. Recommendations for evaluation of the severity of native valvular regurgitation with two-dimensional and doppler echocardiography. *J Am Soc Echocardiogr.* 2003;**16**(7):777–802.
14. Stellin G. Repair of congenital mitral valve dysplasia in infants and children: is it always possible? *Eur J Cardio Thorac Surg.* 2000;**18**(1):74–82.
15. Yoshimura N, Yamaguchi M, Oshima Y, Oka S, Ootaki Y, Murakami H, et al. Surgery for mitral valve disease in the pediatric age group. *J Thorac Cardiovasc Surg.* 1999;**118**(1):99–106.
16. Honjo O, Ishino K, Kawada M, Akagi T, Sano S. Midterm outcome of mitral valve repair for congenital mitral regurgitation in infants and children. *Interact Cardiovasc Thorac Surg.* 2006;**5**(5):589–93.
17. Lee C, Lee CH, Kwak JG, Park CS, Kim SJ, Song JY, et al. Long-term results after mitral valve repair in children. *Eur J Cardiothorac Surg.* 2010;**37**(2):267–72.
18. Hoashi T, Bove EL, Devaney EJ, Hirsch JC, Ohye RG. Mitral valve repair for congenital mitral valve stenosis in the pediatric population. *Ann Thorac Surg.* 2010;**90**(1):36–41.
19. Alghamdi AA, Yanagawa B, Singh SK, Horton A, Al-Radi OO, Caldarone CA. Balancing stenosis and regurgitation during mitral valve surgery in pediatric patients. *Ann Thorac Surg.* 2011;**92**(2):680–4.
20. Caldarone CA, Raghuveer G, Hills CB, Atkins DL, Burns TL, Behrendt DM, et al. Long-term survival after mitral valve replacement in children aged <5 years: a multi-institutional study. *Circulation.* 2001;**104**(12 Suppl 1):I143–7.
21. Kalfa D, Vergnat M, Ly M, Stos B, Lambert V, Baruteau A, et al. A standardized repair-oriented strategy for mitral insufficiency in infants and children: midterm functional outcomes and predictors of adverse events. *J Thorac Cardiovasc Surg.* 2014;**148**(4):1459–66.
22. Oppido G, Davies B, McMullan DM, Cochrane AD, Cheung MM, d'Udekem Y, et al. Surgical treatment of congenital mitral valve disease: midterm results of a repair-oriented policy. *J Thorac Cardiovasc Surg.* 2008;**135**(6):1313–20.