

## Surgical Mitral Valve Repair in Children with Rheumatic Fever

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### Summary

**Background:** Mitral repair is well accepted in children with rheumatic fever.

**Objective:** To analyze the outcomes of surgical mitral repair in children with rheumatic lesions after four years of follow-up.

**Methods:** Retrospective study of 40 patients younger than 18 years, who underwent surgery in the National Institute of Cardiology (Rio de Janeiro) between January 1998 and January 2003. The echocardiographic degree of mitral regurgitation; surgical technique used; pre and postoperative functional class; patient outcome; need for valve replacement; and deaths were analyzed.

**Results:** Twenty one patients (52.5%) were females. Severe mitral regurgitation was observed in 32 patients (80%) and moderate in eight (20%). Three immediate deaths occurred (7.5%). Three months after surgery, echocardiography showed no valve regurgitation or mild regurgitation in 35 of 37 cases (94.6%) patients, and severe regurgitation in two (5.2%). Thirty three cases (82.5%) were in functional class III or IV in the preoperative period, and three months after surgery all the 37 cases (100%) were in functional class I or II. The differences between the degree of mitral regurgitation and functional class in pre and postoperative periods were statistically significant ( $p < 0.01$ ). Seven (19%) patients underwent heart valve replacement before four years of follow-up.

**Conclusion:** Mitral valve repair showed favorable results in most of the cases as regards the degree of mitral regurgitation and the pre and postoperative functional class. Only 19% of the patients required surgical valve replacement before four years of follow-up. (Arq Bras Cardiol 2009; 92(6) : 400-404)

**Key words:** Mitral valve/surgery; mitral valve insufficiency; rheumatic fever.

### Introduction

The incidence of rheumatic fever among children, adolescents and young adults is of 0.3% to 3% of the individuals<sup>1</sup>. There are few Brazilian studies available on the prevalence of this disease. Among children from public schools of the city of Belo Horizonte this rate was 3.6/1,000<sup>2</sup>. In Brazil, rheumatic fever accounts for 30% of the cardiac surgeries in general, especially for mitral valve regurgitation<sup>3</sup>.

Mitral regurgitation results from annulus dilatation and changes in the papillary muscles, chordae tendineae and leaflets<sup>4-7</sup>. The available prostheses lead to complications which are difficult to be controlled, especially calcification and rupture in bioprostheses, thromboembolism in mechanical prostheses, and the need for continuous use of anticoagulation and its monitoring with periodic laboratory tests<sup>8</sup>. In lower age ranges, prosthesis replacement is necessary because of height and weight growth. Based on these facts, surgeons have used reconstructive techniques in the treatment of mitral valve regurgitation with increasing frequency<sup>9,10</sup>.

The objectives of the present study were to analyze the results of mitral valve repair in patients with rheumatic fever, to evaluate the patients' outcome, and to determine the factors that could influence the need for valve replacement before four years of follow-up.

### Methods

This is a retrospective study including 40 patients younger than 18 years of age, who underwent surgical reconstruction of the mitral valve (mitral valve repair). They were followed in the Department of Child and Adolescent Cardiology and the surgeries were performed from January 1988 to January 2003 at the Department of Pediatric Cardiovascular Surgery, both from the National Institute of Cardiology (NIC), a Ministry of Health agency, situated in the city of Rio de Janeiro (RJ). The variables studied were: gender, patient age at onset of disease and at surgery, medications used, type of valve lesion, surgical technique, pre and postoperative New York Heart Association (NYHA) functional class, need for vasoactive amines or blood products, duration of extracorporeal circulation and of anoxia, outcome of the cases and need for heart valve replacement. Surgeries were performed by the same surgical team in all patients, using the median sternotomy approach. After heparinization with 4ml/kg of weight, the ascending

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Manuscript received July 29, 2008; revised manuscript received October 16, 2008; accepted October 24, 2008

aorta and venae cavae were cannulated, and extracorporeal circulation (ECC) was established. All patients underwent moderate hypothermia (28°C), continuous total aortic clamp, and myocardial protection with blood cardioplegia at 4°C. The mitral valve was approached via the left atriotomy, and both mitral valve cusps were preserved in all patients.

Echocardiographic studies for the analysis of mitral regurgitation were performed in a SONUS instrument (Pharmaceutical-Abbott Laboratories, USA), and regurgitation was rated as mild, moderate or severe, according to the characteristics of the regurgitant jet. The main surgical techniques used for valve repair were: mitral ring placement, quadrangular resection of the posterior leaflet, chordal shortening, chordal transfer, and commissurotomy. The patients were followed-up for at least four years postoperatively.

Data were described as relative frequencies (percentage), mean, median, minimum and maximum values, and the groups were compared using the Chi square test. Fisher's correction was used when indicated. The possible risk factors for heart valve replacement before four years of clinical follow-up were analyzed using simple logistic regression. Statistical significance was set at 95%.

The cases studied are filed in the NIC database for the long-term follow-up and further description of the outcomes.

This study was approved by the Research Ethics Committee of NIC under number 0181/26.11.07.

## Results

Twenty one patients (52.5%) were females. The age at bout of rheumatic fever and at surgery ranged from three to 14 years (median of eight years), and from four to 17 years (median of 12 years), respectively. The difference between the age at the first bout and at surgery ranged from zero to 10 years (median of three years) (Table 1).

In the first visit prior to surgery, 12 patients (30%) were not taking any medication, 16 (40%) were on four or more medications, and the remaining twelve cases were on continuous use of one to three medications. The drugs more frequently used were: furosemide, spironolactone, captopril, hydralazine, and penicillin G benzathine.

The degree of mitral regurgitation was analyzed by echocardiography in the preoperative period and was classified as mild, moderate or severe, according to the regurgitant jet. Mitral regurgitation was severe in 32 patients (80.0%) and moderate in eight (20.0%). Immediate echocardiographic analysis was performed in all patients, except for two, due to death. The analyses showed that 20 of the 38 patients

(52.6%) had mild or no valve regurgitation, and 18 (47.4%) had moderate regurgitation. Another death occurred three days after surgery.

The echocardiographic studies performed three months after surgery in the 37 patients that survived showed that 19 (51.3%) had mild or no valve regurgitation, 16 (43.3%) had moderate regurgitation, and two (5.4%) had severe regurgitation (the latter presented a new bout of rheumatic fever). Therefore, 35 patients (94.6%) presented mild or moderate mitral regurgitation in the postoperative period. Statistically significant difference was observed ( $p < 0.001$ ) (Table 2).

The length of hospital stay ranged from four to 103 days (median of 12). ECC time ranged from 50 to 220 minutes, with mean of  $120 \pm 36$  minutes, and time of anoxia from 35 to 170 minutes, with mean of  $93 \pm 32$  minutes. The mean length of stay in the children's postoperative care unit ranged from one to 60 days (median of four) (Table 3).

The main surgical techniques used were: mitral ring placement ( $n = 28$ ), quadrangular resection of the posterior leaflet ( $n = 17$ ), chordal shortening ( $n = 23$ ), chordal transfer ( $n = 5$ ), and commissurotomy ( $n = 11$ ). Eleven patients (27.5%) required only one surgical technique, whereas 29 patients (72.5%) required two or more techniques (Table 4).

The preoperative NYHA functional class ranged from II to IV: FC II ( $n = 7$ ) 17.5%, FC III ( $n = 17$ ) 42.5%, and FC IV ( $n = 16$ ) 40.0%, that is, 33 patients (80.5%) were in functional class III and IV in the preoperative period. In the outpatient follow-up three months after surgery, all the 37 patients (100%) became FC I ( $n = 28$ ) and II ( $n = 29$ ). Statistically significant difference was observed ( $p < 0.001$ ) (Table 5; Figure 1)

**Table 2 – Preoperative and late postoperative severity of mitral regurgitation as assessed by echocardiography in the patients**

Severity of mitral regurgitation	Preoperative period	Late postoperative period (3 months)	$\chi^2$ *	p value
	n= 40 (100%)	n=37 (100%)		
Mild to Moderate	8 (20)	35 (94.6%)	46.005	$p < 0.001$
Severe	32 (80)	2 (5.4)		

\* Cochran's method and Fisher's correction.

**Table 3 – Length of hospital stay and operative time**

Maximum time	Mean $\pm$ SD	Median	Minimum value	Maximum value
Days of hospital stay	20 $\pm$ 20 dias	12	4	103
Time of ECC *	120 $\pm$ 36 min	120	50	220
Time of anoxia *	93 $\pm$ 32 min	97	35	170
Days of IPO	6 $\pm$ 9 dias	4	1	60

\* ECC – extracorporeal circulation; IPOI – infant postoperative period.

**Table 1 – Demographic characteristics of the patients undergoing mitral valve repair (n=40)**

Variables	Minimum value	Maximum value	Median
Age at bout	3	14	8
Age at surgery	4	17	14
Time between bout and surgery	0	10	3

**Table 4 – Most frequently used surgical techniques (n=40)**

Technique	n	%
Mitral ring placement (Carpentier's)	28	70
Chordal shortening	23	57.5
Quadrangular resection	17	42.5
Commissurotomy	11	27.5
Chordal transfer	5	12.5

More than one technique was used in several cases.

Transesophageal echocardiography was performed in 28 patients (70.0%) during surgery. Blood products were used in 21 cases (52.5%). Vasoactive amines were necessary in 26 patients (65.0%) in the postoperative period. Three immediate deaths occurred (7.5%): one in the operation room, one 24 hours after surgery, and one three days after surgery (two patients of cardiogenic shock and one of coagulation disorders).

Heart valve replacement was required in eight patients (20.0%), seven of whom with less than four years of mitral repair (Table 6).

Risk factors that could affect the outcome of patients in relation to the mitral valve replacement before four years, and whose analysis is shown in Table 6, were not statistically significant and included functional class and degree of pre and postoperative mitral regurgitation; number of medications used by the patients at the moment of surgery; time of perfusion and anoxia; number of surgical techniques used; and need for vasoactive amines in the immediate postoperative period.

## Discussion

Mitral valve repair is a technique universally accepted as superior to valve replacement, especially in children, in whom further replacements may be necessary as they grow<sup>6,7,11</sup>. The

**Table 5 – Pre (n=40) and postoperative (n=37) variation of NYHA functional class**

Assessment of the Functional Class	Preoperative	Postoperative	X <sup>2</sup>	p value
	n = 40	n = 37		
Functional class I and II	7 (17.5%)	37 (100%)		
Functional class III and IV	33 (80.5%)	0	54.340	p<0.001

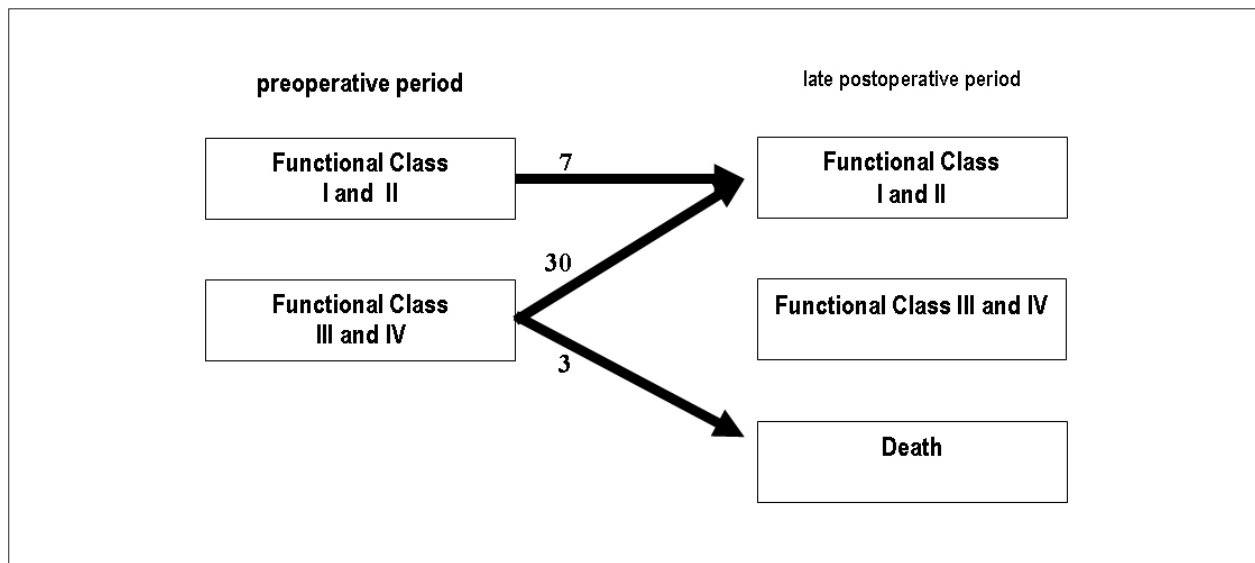
\* Cochran's method and Fisher's correction.

**Table 6 – Possible risk factors for surgical valve replacement before four years of mitral valve repair (n=7)**

Variable analyzed	OR*	IC 95%	p
Functional class**	2.61	0.25 – 24.38	0.397
Medications***	1.08	0.25 – 4.63	0.914
Perfusion <sup>∞</sup>	1.16	0.20 – 6.80	0.864
Surgery <sup>•</sup>	2.44	0.56 – 10.55	0.231
Anoxia <sup>∞</sup>	2.18	0.47 – 10.05	0.314
Amines <sup>◊</sup>	1.47	0.32 – 6.83	0.620

OR - odds-ratio; 95% CI, 95% confidence interval; \* Odds-ratio - adjusted for gender, age, functional class, medications, time of perfusion, operative time, time of anoxia, use of vasoactive amines; \*\* - New York Heart Association functional class heart failure; \*\*\* - Number of medications used per patient at the moment of surgery; <sup>∞</sup> - Time in minutes; <sup>•</sup> - Number of surgical techniques used by the surgeon; <sup>◊</sup> - Number of patients using vasoactive amines.

age of the patients ranged from four to 17 years at the moment of the surgical mitral repair. Due to the long-term complications related to anticoagulation, thromboembolism, bleeding, rapid prosthetic degeneration in the young population, the higher



**Figure 1 - Progression of NYHA functional class in patients undergoing mitral repair**

risk of endocarditis, and the poorer preservation of the ventricular function, mitral valve repair is superior to mitral valve replacement<sup>12</sup>. An important advantage of the surgical reconstruction technique is the low thrombogenicity of the repaired valve, which eliminates the need for anticoagulation therapy. Absence of thromboembolism after mitral repair is reported in 87% to 99% of the cases after five years. Most of the embolic manifestations are cerebrovascular events<sup>10-12</sup>.

The following valve repair techniques were used: mitral ring placement, quadrangular resection, chordal shortening, commissurotomy and chordal transfer, with the aim of preserving both the mitral apparatus and the ventricular function. In rheumatic fever, valve reconstruction is technically more difficult and the mid and long-term outcomes may be influenced by new bouts of the disease<sup>6,9</sup>. Initial studies presented techniques which addressed only the valve annulus treatment<sup>13</sup>. One of the largest personal experiences in conservative procedures in the mitral valve is that of Carpentier's<sup>14</sup>, with 1,421 patients with mitral regurgitation. The techniques used in his series were partial resections, chordal shortening or transfer, reinsertion of the papillary muscles, and annuloplasty with rigid ring.

In the present study, 39% of the patients underwent surgery with significant impairment of the whole valvular structure and myocardial dysfunction. These situations, which are not uncommon in our midst (surgical treatment performed when the patient already presents ventricular dysfunction and/or significant clinical symptoms), mainly result from social factors and difficulties found in our health system. However, mitral repair should be indicated early, on the basis of the severity of regurgitation and on the degree of valve impairment; thus, favorable results can be achieved with the conservative treatment, even when symptoms are scarce or absent<sup>15</sup>. For valve reconstruction, perfect knowledge of the anatomy and multiplicity of techniques available is required of the surgical team. Additionally, excellent visualization of the valve is important, especially of the subvalvular plane, to enable good assessment of the chordae tendineae and papillary muscles. If necessary, these structures are shortened, in an attempt to achieve good cusp coaptation. In the Department of Pediatric Cardiovascular Surgery of NIC, surgeons perform papillary muscle shortening by means of a cross-sectional wedge resection because they believe this technique permits more homogeneous shortening of the chordae tendineae. In addition to this technique, they also use mitral ring placement, quadrangular resection, commissurotomy, and chordal transfer.

Annuloplasty was the single procedure used in only eight of the patients studied (20%), thus demonstrating that in most of the cases (80%) the association with more than one technique was necessary, and in many occasions with modifications of the initial planning, a situation that has also been described by other authors<sup>6,10,16</sup>. Mitral annulus dilatation is present in almost all patients with mitral regurgitation. Thus, ring implantation offers more stability in valve reconstruction, preventing further dilatations of the posterior cusp as suggested by Carpentier<sup>14</sup>, according to whom remodeling the valve with repair is always important for the long-term outcomes. Deloche et al demonstrated excellent durability of mitral repair

using Carpentier's technique, and only 6.2% of their series required further mitral valve replacement<sup>9</sup>. In our study, 19% underwent early mitral valve replacement.

Mitral regurgitation may be secondary to multiple lesions in the commissures, in the leaflets, and in the subvalvular apparatus. As previously mentioned, mitral annulus dilatation is observed in all patients. This is due to the displacement of its posterior portion, since the anterior portion is limited by the right and left fibrous trigones of heart. According to the literature, repair is considered a good option, because it permits the conservation of the native valve cusps and of the subvalvular apparatus, thus preserving the left ventricular geometry and ventricular function<sup>10</sup>.

In-hospital mortality in the present study was 7.5%. The three patients who died were in functional class IV, and two of them were in the acute phase of the disease. This mortality rate may be considered high if compared with those of most of the authors, which range from 0 to 4.8%<sup>17-20</sup>. Carpentier reported his 10-year experience with repair, with an in-hospital mortality of 4.2% in adults and children<sup>14</sup>. Surgical mortality is higher in the acute phase of rheumatic fever when operation is indicated in patients with severe valve dysfunction and ineffective clinical treatment. Even in this group, repair may provide better results than valve replacement, as it preserves the whole subvalvular apparatus, which results in better postoperative ventricular function<sup>12,19</sup>.

In our case series, reoperation with indication of valve replacement before four years was necessary in seven (19%) of the 37 cases, a very high rate in comparison with that of other authors. Pomerantzeff et al<sup>21</sup> reported reoperation-free survival in 70% after 17 years<sup>21</sup>. Patients with rheumatic valve disease have had a high incidence of reoperation, as demonstrated by Deloche et al<sup>9</sup>. Reoperation rates range from 8% to 10%<sup>7,18</sup>. Additionally, the outcomes also depend on the status of the valve at the moment of reconstruction, that is, the more severely it is damaged, the worse the outcome<sup>16</sup>. Gillinov et al reported reoperation-free survival in 93% after 10 years<sup>22</sup>; the reason for the need of a new surgery was new bouts of rheumatic fever in most of the patients. In Machado et al<sup>18</sup> case series, 72.6% of the cases were free from reoperation after 188 months. The major causes of failure in rheumatic mitral repair are wrong indications, use of inadequate techniques, and progression of the rheumatic valve disease<sup>7,16</sup>.

Transesophageal echocardiography has been established as the most adequate method for the assessment of immediate outcomes, guiding the need for valve replacement in the cases of failure in mitral repair or significant residual regurgitation<sup>10</sup>. Murad et al<sup>6</sup> reported the use of transesophageal echocardiography in only 4% of their surgeries. In the present study, transesophageal echocardiography was used in most of the patients (68.3%) during the surgical procedure. Also, we observed improved regurgitation in 94.6%, a result similar to that of other Brazilian studies<sup>16,18</sup>.

In conclusion, there was a significant improvement of mitral regurgitation as well as of the functional class, as

verified in the assessment at the third postoperative month, however with a high need for reoperation before four years of follow-up.

#### Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

#### Sources of Funding

There were no external funding sources for this study.

#### Study Association

This article is part of the thesis of master submitted by Andréa Rocha e Silva, from Universidade Federal Fluminense and Instituto de Cardiologia de Laranjeiras.

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