Pediatr Cardiol 22:44–52, 2001
DOI: 10.1007/s002460010151

Pediatric Cardiology

Valve Repair in Children with Congenital Mitral Lesions: Late Clinical Results

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Abstract. Mitral valve repair may be performed without ring support with advantages related to results and complications. The objective of this study was to analyze the long-term clinical results following surgical repair and reconstruction without the use of rings in cases of congenital mitral lesions in children less than 12 years of age. Twenty-one patients who had undergone surgery during the period from 1975 to 1998 were evaluated. The mean age was 4.6 ± 3.4 years. Females represented 47.6% of the total. Mitral regurgitation was present in 57.1% (12 patients), stenosis in 28.6% (6 patients), and the mixed lesion group represented 14.3% (3 patients). Perfusion time was 43.1 ± 9.5 minutes and ischemic time 29.4 ± 10.5 minutes. Follow-up time was 41.5 ± 53.6 months for the regurgitation group, 46.3 ± 32.0 months for the stenosis group, and 39.41 ± 37.51 months for the mixed lesion group. Echocardiographical follow-up time was 37.17 ± 39.51 months for the regurgitation group, 42.61 ± 30.59 months for the stenosis group, and 39.41 ± 37.51 months for the mixed lesion group. Operative mortality was 9.5% (two cases). There were no late deaths. In the regurgitation group, 10 patients (83.3%) were asymptomatic ($p = 0.004$). In the echocardiographical follow-up, most of the patients had minimal regurgitation. In the clinical follow-up of the stenosis group all patients were in functional class I (NYHA). The mean transvalvular gradient measured by echocardiography was from 8 to 12 mmHg with a mean gradient of 10.7 mmHg. In the mixed lesion group there was one reoperation at postoperative month 43. There were no cases of endocarditis or thromboembolism. Mitral valve repair in congenital lesions is associated with good late results. The majority of cases in the regurgitation group remain asymptomatic and do not require reoperation. Rings or annular support are not necessary in such cases. Satisfactory repair is more difficult to achieve in cases of mitral stenosis due to valvular abnormalities and the seriousness of the associated lesions.

Key words: Congenital heart disease — Mitral valve lesion — Valve repair

Congenital mitral valve malformations are complex lesions that involve diverse morphological abnormalities which generally affect more than one valvular component [4, 8, 9, 31] and because they occur in a population group with a high prevalence of associated cardiac anomalies [1, 30].

Isolated congenital lesions are rare [4, 6, 32]. Stenosis or mitral regurgitation affects 1% of the population of patients with congenital heart disease [18]. Isolated mitral congenital regurgitation is extremely uncommon [6]. In infants it is usually found in association with other cardiac defects, in connective tissue disorders, and in acquired inflammatory conditions such as myocarditis, endocarditis, rheumatic fever, Kawasaki's disease, and other collagenoses involving vascular damage [6]. Flow obstruction in congenital mitral stenosis results from different morphological anomalies [4, 18, 31].

The aim of this study is to present an analysis of late clinical results following surgical treatment of congenital mitral anomalies in cases with or without associated malformations (with the exception of total atrioventricular septal defects) in children up to 12 years of age, who were treated using repair and reconstruction techniques without annular support, we also provide a review of the related literature.

Patients and Methods

From 1975 to 1998 at the Instituto de Cardiologia do Rio Grande do Sul, 21 patients with mitral valve congenital lesions underwent operations. The mean age was 4.67 ± 3.44 years, and females represented

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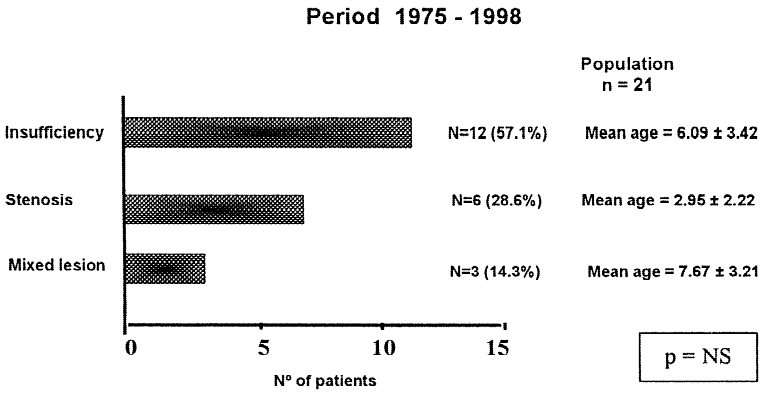

Fig. 1. Patients classification by type of lesion (atrioventricular septal defects were not included).

47.6% and males 52.4%. Mitral valve regurgitation was present in 57.1% (12 cases), stenosis in 28.6% (6 cases), and mixed lesion in 14.3% (3 cases) (Fig. 1). The most frequent valvular morphological lesions found in the regurgitation group were annular dilatation in 75%, anterior leaflet cleft in 33.3%, and posterior leaflet cleft in 16.6%. In the stenosis group parachute valve was found in 50% of the cases and commissural fusion in 33.3%. In the mixed lesion group leaflet mobility decrease was found in 100% of cases (Table 1). Patients with total atrioventricular septal defects were excluded from the study. Associated intracardiac malformations were found in 61.9% of patients: in 83.3% of the stenosis group, 41.6% of the regurgitation group, and all patients in the mixed lesion group. More severe malformations were found in 28.6% of the patients, all having valvular stenosis; parachute valve was found in 3 cases, all having stenosis, 2 of which had associated Shone's syndrome. Isolated annular dilatation without other malformations was found in 2 cases.

All patients were operated by means of median sternotomy with conventional extracorporeal circulation using a bubble or membrane disposable oxygenator and moderate hypothermia at 28°C to 30°C. For myocardial protection crystalloid hyperkalemic cardioplegic solution and pericardial cavity irrigation with cold saline at 4°C were employed. None of the patients had prior surgery for mitral correction. Mitral valve approach was done through left longitudinal atriotomy. In two (33.3%) patients isolated mitral commisurotomy was performed. In the regurgitation group, annuloplasty was carried out in nine patients (75%) using Wooler's technique [42]. Associated procedures in 75% of cases were chord shortening in four, anterior leaflet cleft suture in four, and posterior leaflet cleft suture in two (Table 1). The associated cardiac lesions were corrected after mitral valvoplasty (Table 1). The extracorporeal circulation mean time was 43.1 ± 9.5 minutes and that of aortic cross clamping was 29.4 ± 10.5 minutes.

Postoperative control took the form of periodical clinical, radiological, and/or ecchocardiographic evaluation. Follow-up time was up to 15.7 years (mean 3.8 ± 4.1 years).

Results

The operative mortality rate was 8.9% (two deaths) in cases of mitral stenosis. One 12-day-old patient had mitral valve hypoplasia associated with left ventricular firbroelastosis, and a 7-month-old patient had parachute mitral valve (Table 1).

Overall morbidity incidence was 28.5% of the total patient population. The highest incidence occurred in the regurgitation group (two patients): one case of atelectasia after drain removal and one of pleural hemorrhage. In the stenosis group, one patient had repetitive respiratory infection; in the mixed lesion group, one patient had operative wound infection.

No episodes of endocarditis or thromboembolism were reported during the observation period (Table 1). The incidence of reoperation was 13% (three cases), all by failure repair.

In the regurgitation group the mean follow-up time was 41.5 ± 53.6 months. At the last evaluation, 10 patients (83%) were asymptomatic and off medication. There was 1 patient in NYHA functional class I and another in functional class II, both on medication (Table 1). One patient was reoperated at postoperative month 48, when a new valvuloplasty was performed. Echocardiographic evaluation was made on average 37.17 ± 39.51 months, and most of the patients were found to have a light reflux ($p = 0.002$; Table 1);

In the stenosis group (six cases), there were two deaths of the four surviving patients (66%), two are in functional class I (and use medication) and two are in class II. Echocardiographic mean follow-up time on these patients was 42.6 ± 30.5 months and showed a mean transvalvular gradient between 8 and 12 mmHg.

In the mixed lesion group (three cases) there was one patient in which echocardiography indicated light regurgitation at the time of discharge from the hospital and for whom there was no further follow-up; one patient in whom the valve was replaced by a bioprothesis at 43 months following the first operation; and one patient who, at 75 months following surgery, was in functional class II and echocardiography showed mild stenosis and regurgitation.

The echocardiographic evaluation of the whole sample population with mean follow-up time of 39.8 months showed most of the patients had mild lesions ($p = 0.002$) (Table 1 and Fig. 2). In 83.3% of the patients in the regurgitation group, reflux was either absent or light (Fig. 3).

Table 1. Morphology, surgical techniques, and late clinical results

Patient No./age	Morphology	Associated malformations	Surgical techniques	Surgical morbidity		Surgical mortality		Reoperations	Functional class		Eco	
				Hospital	Late	Hospital	Late		Preop	Postop[a]	Preop	Postop[b]
Mitral incompetence												
1/6 years	ALC + ACDP	ACDP	Anterior cleft suture	No	No	No	No	No	I	ASS	R severe	Moderate regurgitation
2/4 years	AD	TI	Wooler + Devega	Hem. Pl. Efusion	No	No	No	No	I	ASS	R severe	Light regurgitation
3/7 years	AD + PLC + ACDP	ACDP	Wooler + posterior cleft suture	No	No	No	No	No	III	ASS	R severe	Light regurgitation in the discharge
4/2 years	AD	LPH	Wooler	Atelectasis	No	No	No	No	III	ASS	R severe	Absent in the discharge
5/9 years	PLC + ACDP	ACDP	Posterior cleft suture	No	No	No	No	No	II	II	R severe	Light regurgitation
6/6 years	AD + ALP	No	Wooler + Chordal shortening	No	No	No	No	4 years PO (repair)	IV	ASS	R severe	Light regurgitation
7/6 years	ALC	No	Anterior cleft suture	No	No	No	No	No	I	ASS	R severe	Light regurgitation
8/5 years	AD + ALC + ACDP	ACDP	Wooler + anterior cleft suture	No	No	No	No	No	I	ASS	R severe	Light/moderate regurgitation
9/1 years	AD + ALP + AIPM	No	Wooler + chordal shortening	No	No	No	No	No	IV	I	R severe	Moderate regurgitation
10/13 years	AD	No	Wooler + chordal shortening	Atelectasis	No	No	No	No	II	ASS	R severe	Light regurgitation
11/8 years	AD + ALP	No	Wooler + chordal shortening	No	No	No	No	No	I	ASS	R severe	Light regurgitation
12/2 years	AD + ALC	No	Wooler + anterior cleft suture	No	No	No	No	No	I	ASS	R severe	No eco control
Mitral stenosis												
1/3 years	P	PDA + VSD + SS	Pappilotomy	No	No	No	No	No	III	I	R severe	Light mixed lesion
2/16 years	CF	No	Comissurotomy + pappilotomy	No	No	No	No	8 years PO (repair)	I	I	R severe	Light stenosis
3/12 days	H	AS + FE	Comissurotomy	No	No	IPO	No	No	III	No	R severe	—
4/18 months	CF	AS + AC	Comissurotomy	No	No	No	No	No	III	I	R severe	Light mixed lesion
5/3 years	P	AS + SS	Pappilotomy	RRI	No	No	No	No	II	I	R severe	Light mixed lesion
6/7 months	P	FE	Comissurotomy	No	No	IPO	No	9 days PO	IV	No	R severe	No

Continued next page

Table 1. *Continued*

Patient No./age	Morphology	Associated malformations	Surgical techniques	Surgical morbidity		Surgical mortality		Reoperations	Functional class		Eco	
				Hospital	Late	Hospital	Late		Preop	Postop[a]	Preop	Postop[b]
Mixed lesions												
1/6 years	DM + CF	No	Comissurotomy	No	No	No	No	No	II	I	R severe	Moderate mixed lesion
2/10 years	DM + ALC + FTPM	SS, S. Noonan	Cleft closure + pappilotomy	No	No	No	No	No	III	I	R severe	Light regurgitation (discharge)
3/4 years	DM + FTPM	TI + SFS	Pappilotomy	OWI	No	No	No	4 years PO (prosthesis)	III	II	R severe	—

AD, annulus dilatation; ALC, anterior leaflet cleft; PLC, posterior leaflet cleft; ALP, anterior leaflet prolapse; AIPM, anomalous implantation of the papillary muscle; P, parachute valve; CF, comissure fusion; HV, hipoplastic valve; DM, decrease motion; FTPM, fibroelastic tissue in papillary muscle; ACDP, atrioventricular canal defect partial; ASD, atrial septal defect; VSD, ventricular septal defect, Hem. Pl. efusion, Hemorrhagic pleural efusion; Ss, subaortic stenosis; TI, Tricuspid incompetence; FE, fibroelastosis; LPH, left pulmonary hipoplasia; PS, pulmonary stenosis; AS, aortic stenosis; RRI, repetitive respiratory infection; OWI, operative wound infection; IPO, immediate postoperative; ASS, asintomatic. No endocarditis tromboembolism episodes were reported. None of the patients reported previous surgery before mitral valve correction.

[a] Mitral regurgitation group $p = 0.004$.

[b] Mitral regurgitation group $p = 0.002$.

Fig. 2. Apical four-chamber view shows mild postoperative mitral regurgitation. *LA,* left artery; *LV,* left ventricle; *RA,* right artery; *RV,* right ventricle.

Actuarial probability of survival in the regurgitation group was 90% at 5, 10, and 15 years (Fig. 4). Actuarial probability of freedom from reoperation was 72% at 5 years and 46% at 10 and 15 years (Fig. 5) for the whole patient population ($N = 21$). In the regurgitation group, the actuarial probability of survival free from reoperation was 86% at 5, 10, and 15 years (Fig. 6).

Discussion

Clinical presentation and surgical indication depend on the severity of the mitral lesion and associated intracardiac defects [2]. In our series, the indication for surgery was intractable cardiac failure or pulmonary hypertension or both. If possible, surgery should be avoided before 6 months of age. In infants younger than 3 months, the collagen is not adequately mature and the tissue is particularly friable, making manipulation difficult [9]. In our series the mean age at surgery for regurgitation was 6.1 ± 3.4 years and for stenosis it was 4.0 ± 5.9 years, with the difference being statistically nonsignificant (Fig. 1). In Carpentier's [9] series, it was 6.1 ± 3.2 years for regurgitation and 5.1 ± 3.2 years for stenosis. In Uva et al.'s [41] it was 7.4 ± 2.7 months for regurgitation and 5.8 ± 3.9 months for stenosis. The following are factors that may explain the lower age for surgical intervention for stenosis: Stenotic lesions are less well tolerated than regurgitation, and this group is associated with a higher incidence and more severe intracardiac abnormalities than the regurgitation group.

Congenital mitral stenosis occurs in 0.6% of autopsies and in 0.2% to 0.42% of clinical series [4, 33]. In Ruckman and Van Praagh's [38] series from 49 autop-

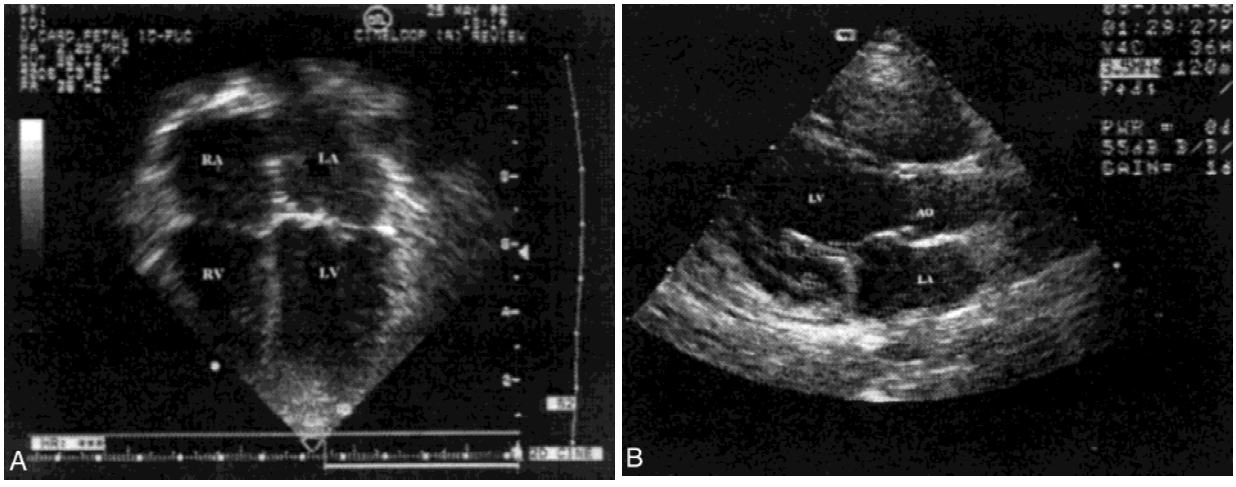

Fig. 3. (A) Apical four-chamber view in patient No. 10 (see Table 1) showing the absence of regurgitation and adequate mitral valve function. **(B)** Parasternal long-axial view (systole) from patient No. 10 (see Table 1) showing nearly normal mitral apparatus. *LA,* left artery; *LV,* left ventricle; *RA,* right artery; *RV,* right ventricle.

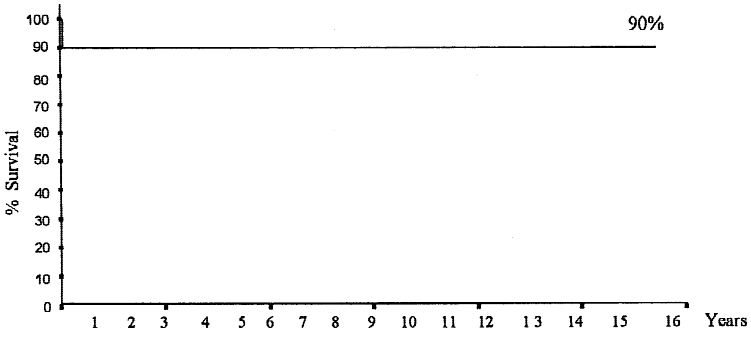

Fig. 4. Actuarial probability of survival in the regurgitation group.

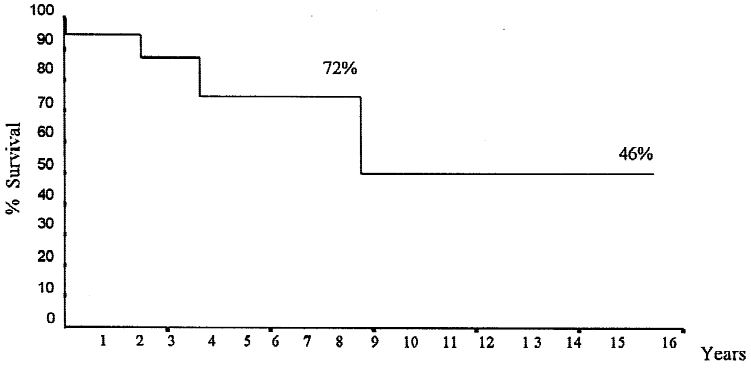

Fig. 5. Actuarial probability of survival free from reoperation for all patients.

sies with stenosis, typical stenosis was found in 49%, with the most common associated lesion being aortic coartation. Left ventricle (LV) size was normal in 96% of these patients. Congenital mitral hypoplasia was the second most common cause of stenosis (41%) and hypoplasia of the LV was associated with each case. Supravalvular ring was found in 12% of cases and parachute valve was found in 8%.

Traditionally, the lesions in which it is most difficult to ensure an effective and lasting repair are those that exhibit alterations to the subvalvular apparatus with abnormal papillary muscles, such as parachute valve, hammock valve, and papillary muscle agenesia. These lesions often determine the stenosis and are associated with a high frequency of complex malformations [2]. Barbero-Marcial et al. [5] reported good short- and long-

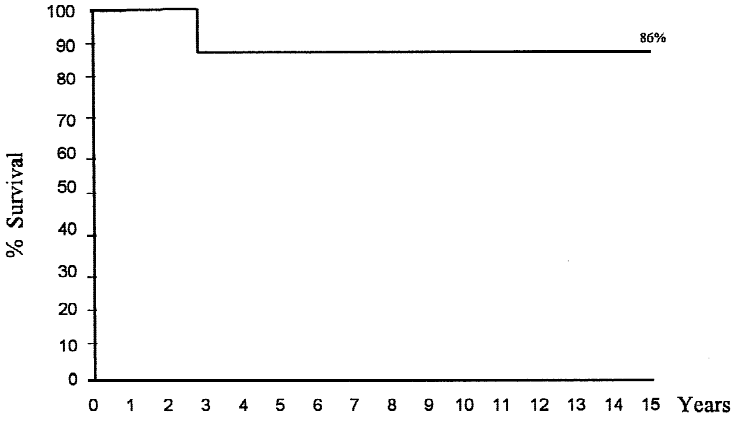

Fig. 6. Actuarial probability of survival free from reoperation for mitral regurgitation.

Table 2. Results of valve repair in anomalous pappilary muscle

	Hammock valve		Parachute	
	Repair	Replace	Repair	Replace
Uva et al. [41]	1	1	2	2
Barbero-Marcial et al. [5]			9	
McCarthy et al. [30]			5	
Stellin et al. [39]			4	1
Fuzellier et al. [19]	11	1	4	1
Kalil (in this series)			3	

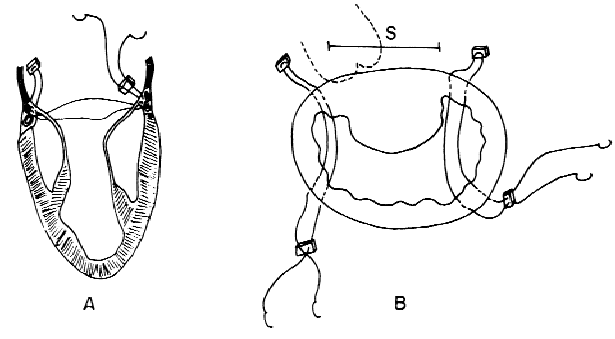

Fig. 7. Wooler annuloplasty. **(A)** Polyester suture through insertion annulus of the mitral leaflet. **(B)** The aim of the suture is to reduce the mural leaflet without compromising the septal leaflet width. The aortic leaflet projection is represented by the dashed line.

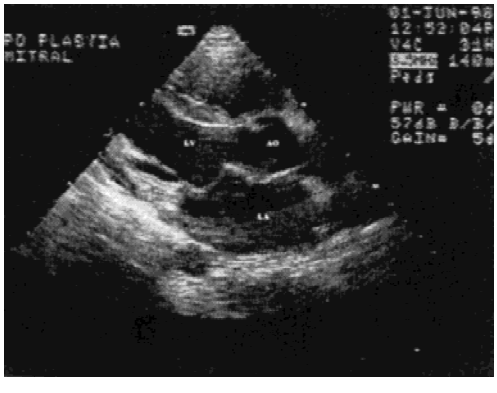

Fig. 8. Parasternal long-axial view of the left ventricle (*LV*) (sistole frame) depicting a prolapse of leaflets still present (patient No. 11; see Table 1). *AO,* aorta; *LA,* left artery.

term results in seven patients with parachute mitral valve. According to the review presented in Table 2, studies published in the past decade have registered important advances in terms of the results in which it was possible to carry out repairs in 91.4% of the cases of mitral stenosis with parachute valve and 50% in cases with hammock valve.

Moore et al. [33] states that the typical mitral hypoplasia with symmetrical papillary muscles was the first cause of stenosis (52% of cases), followed by supravalvular ring (20%). However, Embrey and Behrendt [18] affirm that the ring is rarely so small as to result in stenosis unless LV hypoplasia is present. According to Embrey and Behrendt, chordal malformation is the most common cause of stenosis.

Fuzellier et al. [19], in their series of 50 patients, showed that the most common cause of stenosis with normal papillary muscles is commisural fusion (17 patients). In abnormal papillary muscles the most common cause was Hammock mitral valve (11 patients). In our series, 50% of stenosis cases had some abnormal papillary muscle condition, the most common being parachute mitral valve in 3 cases, 2 of which were associated with Shone's syndrome which allows for the possibility of recuperation (Table 1). Typical mitral stenosis represented 38% of cases and mitral valve hypoplasia 12.5% (Table 1). Left ventricle hypoplasia was present in 50% of the stenosis cases, with 50% of these cases having associated LV fibroelastosis.

According to McGiffin [31] congenital mitral regurgitation occurs less frequently than congenital mitral ste-

Table 3. Reported results for mitral insufficiency

Reference/year	Mean age (years)	*n*	Hospital mortality (%)	Late mortality (%)	Surviving functional (%)	Free from reoperation (%)	Kind of annuloplasty
Okita et al. [36]/1988	5.5	66	1.5	6.0	93.1 (7 years)	89 (10 years)	Kay–Reed
Kirklin and Barret-Boyes [27]/1993	—	18	18	—	—	—	Reed
Bordignon et al. [7]/1996[a]	12.4	13	7.7	0	89 (5 years)	91 (5 years)	Wooler
Uva et al. [41]/1991	1	10	0	0	100 (7 years)	61.2 (7 years)	Wooler
Carpentier [9]/1994	6.1	105	5	4	62 (15 years)	62 (15 years)	Ring
This series	6.09	12	0	0	100 (15 years)	86 (15 years)	Wooler

[a] Includes rheumatic mitral regurgitation.

Table 4. Reported results for mitral stenosis

Reference/year	Mean age (years)	*n*	Hospital mortality (%)	Late mortality (%)	Surviving functional (%)	Free from reoperation (%)
Kirklin and Barret-Boys [28]/1993	4.5	19	21	—	—	—
Uva et al. [41]/1991	0.5	10	0	10	94.1 (7 years)	54.8 (7 years)
Carpentier [9]/1994	5.1	50	26	3	47 (10 years)	47 (10 years)
Geha et al. [20]/1997	4.5	58	25.8	5	67 (15 years)	—
Barbero-Marcial et al. [5]/1991	1.6	9	0	11	89	100

nosis. In our series this was not confirmed; there were 12 cases of regurgitation and 6 of stenosis. The most frequent cause of regurgitation was annular dilatation [3, 31], as confirmed in our series. In the regurgitation group, 75% of patients had this malformation, and in 25% of patients it was the only malformation.

Surgical Treatment

Mitral valve congenital or acquired lesions in children may be surgically treated using mechanical or biological prostheses or valvuloplasty [20, 30]. Valve replacement is accompanied by high mortality, problems with anticoagulation, and the difficulty of annular growth which makes reoperation unavoidable [11, 20, 21, 29–31, 37, 40].

The use of rigid or flexible rings as a prerequisite for efficacious and durable annular repair [8] is currently being questioned for adults [10, 12, 15, 39] and their use will likely diminish.

In children and teenagers the use of the prosthetic rings should be avoided because they inhibit normal annular growth [18], represent a risk factor for cardiac cavity distortion, and contribute to LV outlet obstruction [11, 34]. Given the knowledge that the aortic mitral annular segment has the capacity to contract and relax during the cardiac cycle on LV outlet [10, 12, 34] and is the only annular portion which does not dilate, it is possible to conclude that no prosthetic structure should exist at that level.

Long-term stability of mitral valve repair within the remodeling annuloplasty concept does not require the use of a rigid or flexible ring in order to decrease the annular anteroposterior angle. Since 1975 we have maintained that stability in annuloplasty without support, as with Wooler's technique [42] (Fig. 7), is provided by anchorage to the right and left fibrous trigones, maintaining the normal variable anatomical relationship with aortic valve and LV outlet, enabling the annular segment of mitroaortic continuation to contract and relax during the cardiac cycle [34] (Fig. 8).

When using Wooler's technique, it is essential for long-term stability that the sutures go through the fibrous trigones, and that their relationship with the anterior cusp be considered [23]. This technique showed late results comparable with more complex support techniques [24–26] but with a lower incidence of technical failure. Reported failure of repair due to technical problems with prosthetic rings on adults varies: Aharon et al. [2], 2.9%; Cosgrove [13], 3.2%; Deloche et al. [16], 4.3%; Cosgrove [12], 3.3%.

Hospital mortality was related to NYHA functional class IV. In our series it represented 100% mortality (in

Kirklin and Barret-Boyes' [27, 28] series, patients with stenosis in functional class IV had 50% mortality); both patients had endocardiac fibroelastosis. A 12-day-old patient had left ventricular hypoplasia and a 7-month-old patient had Shone's syndrome. Recent research by Ni et al. [35] suggests that viral infection as the etiology of endocardiac fibroelastosis supporting the hypothesis that this disease is an aftereffect of viral miocarditis, in particular, due to the mumps virus.

Mitral repair was carried out by means of longitudinal atriotomy of the right wall of the left atrium. No other approach was used [5, 14, 17, 22, 36].

Comparative results of studies on regurgitation repair shown in Table 3 verify the long-term good results of annuloplasty without annular support in this population. However, the population of patients with stenosis exhibits a large variety of mitral valvular malformations, an increased association of cardiac defects, and a variability of age that make it difficult to generalize conclusions (Table 4).

In conclusion, mitral valvuloplasty without support in congenital lesions, either isolated or in association with other cardiac malformations, shows good long-term results. Most patients are asymptomatic and free of reoperation. Congenital mitral regurgitation is associated with better results than those obtained by mitral stenosis. Repair failure is associated with the complexity of malformations.

For congenital mitral regurgitation, mitral valvuloplasty without support using Wooler's technique proved to have a low surgical risk and good long-term results, suggesting that prosthetic rings are unnecessary in this group of patients.

Acknowledgments. We thank all the staff in the research department of the IC/FUC, who are always there to help, and my (G.L.) dear wife for the unconditional support she offered. I dedicate this research paper to my wife and my children.

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DOI: 10.1007/s002460010152

Around PediHeart: Hyperlipidemia

For many of us, the treatment of children with hyperlipidemia has become an increasingly important part of our pediatric cardiology practice. Hyperlipidemia has been firmly established as a risk factor for coronary artery disease (CAD) in older adults and more recently in young adults [3]. Evidence has been published linking hyperlipidemia and early atherosclerotic changes in childhood [1]. Finally, and most recently, the development of 3-hydroxy-3-methylglutaryl coenzyme A (HMG CoA) reductase inhibitors or "statins" have made it possible to both treat hyperlipidemia and prevent myocardial infarction as demonstrated in men without previously documented CAD [2]. In light of all this, one of our group presented brothers, aged 9 and 12 years, with similar fasting lipid profiles; total cholesterol 300 mg/dl, LDL cholesterol 240 mg/dl, HDL cholesterol 60 mg/dl, and normal triglycerides. They were athletic, non-obese and had not responded to fat restriction (i.e., the American Heart Association "Step-Two" Diet). The family history was strongly positive for both hyperlipidemia and premature CAD. He wanted advice on treatment options.

Many who wrote back thought that statin therapy would be an appropriate next step. They believed that diet and exercise alone was often not enough in patients with a familial hyperlipidemia and that statins were safe, effective, and better tolerated than bile acid sequestrants or niacin. Others who supported the use of statins in children at the same time pointed out that there was no, and never will be, direct evidence that initiating statin therapy in children prevents CAD. Further, no guidelines exist on when to start statin therapy and what the long-term side-effects may be. Finally, they pointed out the enormous cost of a lifetime of these medicines.

Those opposed to treatment at this time agreed with the above concerns but also felt that statins, in blocking the synthesis of cholesterol, may interfere with neuronal maturation. Ancedotal evidence was offered promoting vigorous daily exercise for the reduction of elevated cholesterol without resorting to medications.

There are no easy answers here. This is largely because we will never have direct evidence as a guide. All one can do is extrapolate from the adult data and circumstantial pediatric data. The most recent policy statement from the American Academy of Pediatrics "Cholesterol in Childhood" (RE9805) published in 1998 provides the scientific justification and recommended strategies for the screening and treatment of hyperlipidemia in children.

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