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Adult congenital heart disease: experience with the surgical approach

Cardiopatia congênita no adulto: experiência com a abordagem cirúrgica

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Abstract

Objective: **To report the institution experience with the surgical treatment of adults with congenital heart disease due to the increasing number of these patients and the need for a better discussion of the subject.**

Methods: **Retrospective analysis describing demographic data, risk factors and results.**

Results: **191 patients between 16 and 74 years old were operated on. Primary correction was done in 171 cases, 93 (55%) for atrial septal defect repair. Among 20 (12%) reoperations, pulmonary valve replacement was done in six cases. The mean intensive care and hospital stay were 2.7 and 8.5 days respectively, significantly greater for the reoperated cases (*P*=0.001). The mean bypass and clamping times were 68.6 and 44.7 minutes respectively, greater for the reoperated cases (*P*<0.0001 and *P*=0.0003 respectively). Hospital mortality was 4.2% and male sex, functional class III-IV and older age at operation were predictive risk factors. Significant complications were more frequent in the reoperated cases (*P*<0.003), mainly atrial flutter and fibrillation. Among 183 patients discharged, 149 (82%) are being followed and atrial flutter and fibrillation are common. The mean functional class value improved significantly after operation (1.66 to 1.11; *P*<0.0001). The estimated survival was 96.2% in six years.**

Conclusion: **Heart surgery in adults with congenital heart disease can be accomplished with low mortality and functional class improvement. Immediate and late complications are frequent. Multicenter studies are important to better characterize this patient population in the country.**

Descriptors: **Heart defects, congenital. Adult. Cardiac surgical procedures.**

Resumo

Objetivo: **Relatar a experiência da instituição com o tratamento cirúrgico de adultos com cardiopatia congênita devido ao crescente aumento no número desses pacientes e consequentes necessidades de maior discussão do tema.**

Métodos: **Análise retrospectiva dos pacientes operados, com análise de dados demográficos, fatores de risco e resultados.**

Resultados: **Cento e noventa e um pacientes, com idade entre 16 e 74 anos, foram operados. Cirurgia primária foi realizada em 171 pacientes, 93 (55%) com comunicação interatrial. Dentre 20 (12%) reoperações, substituição de valva pulmonar ocorreu em seis casos. Os tempos médios de unidade de terapia intensiva e hospitalar foram 2,7 e 8,5 dias, respectivamente, maiores nas reoperações (*P*=0,001). Os tempos médios de cir-**

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Abbreviations, acronyms & symbols	
ASD	Atrial septal defect
CLAMP	Clamping
CPB	Cardiopulmonary bypass
ECHO	Echocardiogram
NYHA	New York Heart Association

culação extracorpórea e pinçamento aórtico foram 68,6 e 44,7 minutos, respectivamente, maiores nas reoperações (*P*<0,0001 e *P*=0,0003, respectivamente). Mortalidade hospitalar foi 4,2%, sem relação com reoperação. Sexo masculino, classe funcional III-IV e idade avançada foram fatores preditivos de risco. Complicações importantes foram mais frequentes nas reoperações (*P*<0,003), principalmente fibrilação e flutter atrial. Entre 183 pacientes com alta hospitalar, 149 (82%) foram seguidos e a fibrilação e flutter atrial foram bastante prevalentes. O valor médio da classe funcional mudou significativamente após a cirurgia (1,66 para 1,11; *P*<0,0001). A estimativa de sobrevida geral foi de 96,2% em seis anos.

Conclusão: **Cirurgia em adultos com cardiopatia congênita pode ser realizada com baixa mortalidade e melhora funcional na maioria dos pacientes. Complicações imediatas e tardias são frequentes. Estudos multicêntricos são importantes para melhor caracterização dessa população de indivíduos no país.**

Descritores: **Cardiopatias congênitas. Adulto. Procedimentos cirúrgicos cardíacos.**

INTRODUCTION

The progressive improvement in the available therapeutic options for treating congenital heart diseases has allowed the great majority of these patients to be treated during pediatric age. The success of these interventions, most of them palliative, and the routine follow up assessment have been responsible for a new population of adolescents and adults [1] with residual cardiovascular lesions [2]. This group of patients, added to those with late diagnosed congenital heart disease or with late symptoms presentation might need an invasive treatment in a period when age related problems also require attention.

The surgical treatment of congenital heart disease in the adult patient has peculiar characteristics. The great variety in diagnosis as well as the complications and residual lesions frequently found makes this therapeutic approach particularly challenging requiring adequate human and structural facilities in order that good results are achieved.

The aim of this study was to review the experience with the surgical treatment offered to adults with congenital heart disease in a tertiary center situated in the São Paulo state where the pediatric invasive treatment for this type of disease has mainly developed during the last twenty years. Reports of global experiences with this type of patients in the country were not available until recently when a relevant experience was published [3]. However, results with specific groups of patients may be found [4-7].

METHODS

A retrospective analysis of all patients 16 years and older submitted to a surgical procedure for treatment of a congenital heart defect at the Hospital das Clínicas da Faculdade de Medicina de Ribeirão Preto da Universidade de São Paulo (HCFMRP/USP) was performed. The cases available for the study underwent surgery between 1st January 1970 to 1st April 2010. The great majority of the patients lived in the city area (n=40, 20%) or in the region (n=133, 70%) while 17 (10%) were referred from other states. An Excel data base was created containing general information of the patients, possible preoperative risk factors such as diabetes, cyanosis, smoking, systemic hypertension, obesity and chronic lung disease, detailed information regarding the surgical procedure, hospital complications and follow up information.

The preoperative data collected, using the information available in the patients notes were: diagnosis [echocardiogram (ECHO) and/or catheter], clinical function according to the New York Heart Association (NYHA) and left ventricular function assessed by ECHO or angiography and classified based on the ejection fraction value as good (>50%), reasonable (between 50% and 30%) and bad (<30%). Pulmonary hypertension was defined when the systolic pulmonary artery pressure was greater than 60 mmHg during ECHO or cardiac catheterization. The surgical informations noted were the type of procedure, the cardiopulmonary bypass (CPB) and clamping (CLAMP) time and also the intensive care and hospital length of stay. The hospital complications were classified as major (high risk) or minor (low risk). After hospital discharge, the informations noted were the length of follow up, functional class at the last outpatient visit and also the complications and comorbidities detected. The comorbidities diagnosed during the long term follow up were classified as major or minor according to the degree of clinical impairment at the assessment. The functional class was numerically transformed from I to IV to 1 to 4 [8].

Statistical analysis was performed using the GraphPad Instat (GraphPad Software, Inc) software. The non-parametric Mann Whitney test was used for comparison of the mean values. The Fisher exact test was used for univariate analysis when the following parameters were assessed: gender, age at operation, functional class, left ventricular function, pulmonary hypertension, arrhythmias, bypass and clamping time, systemic hypertension, cyanosis, plasmatic creatinin and smoking habit. Significant differences were considered when the *P* value was <0.05. A tendency to statistical significance was considered

when the *P* value was situated between 0.05 and 0.10. The survival probability was assessed using the Kaplan-Meier method. Individual informed consent was not requested and the study was approved by the Ethical Committee of the institution.

RESULTS

One hundred and ninety one patients underwent surgery and the annual distribution of the procedures can be seen in Figure 1. One hundred and seventy-two (90%) patients were treated in the last eleven years. The mean age at operation was 34±14 years (16 to 74 years). One hundred and seventeen (61%) were female and seventy-four (39%) were male. The preoperative information can be seen in Table 1. In one hundred and seventy one (89%) patients a primary correction was done while twenty (11%) were re-operated. The diagnoses and number of procedures can be seen in Figures 2 and 3.

The mean intensive care stay was 2.76±3.3 days (range 1 to 37 days). CPB was used in 171 (89%) patients and the mean time duration was 68.65±46 minutes (range 18 to 220 minutes). Procedures without CPB (11%) included relief of aortic coarctation (n=14), ductus ligation (n=5) and pulmonary valvotomy (n=1). Aortic clamping was employed in 166 patients and the mean time duration was 44.71±34 minutes (range 7 to 167 minutes).

Immediate results

Eight (4.2%) patients died during the 30 days after the procedure, all of them during the same admission. Mortality occurred in three patients with an atrial septal defect (ASD), two of them older than 60 years and one submitted to a simultaneous coronary bypass graft; in two patients submitted to a correction of an atrioventricular septal defect, one of them a complete form in a 63-year-old patient; one case of Fallot's tetralogy repair; one patient submitted to a mechanical aortic valve replacement plus coronary artery bypass graft and one

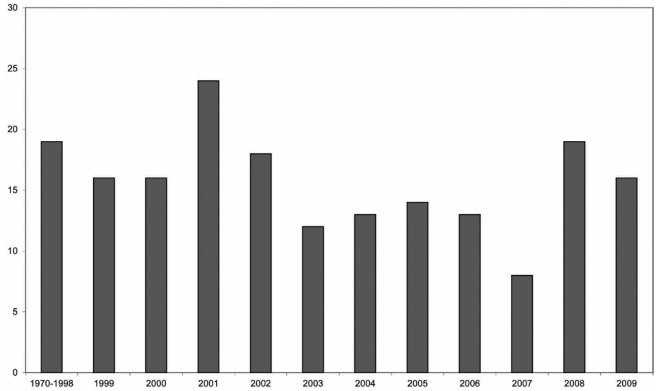

Fig. 1 - Anual number of operations in 191 adult patients operated on for congenital heart disease

Table 1. Preoperative data in 191 patients operated on for treatment of congenital heart disease

	number (%)
Female sex	117 (61%)
Sinus rhythm	166 (97%) n=170
Left ventricular function	
Good	184 (96%)
Reasonable	7(4%)
Bad	0
Pulmonary hypertension	12 (6%)
Cyanosis	9 (5%)
Systemic hypertension	63 (33%)
Smoking	44 (25%) n=175
Diabetes	6 (3%)
Obesity	16 (8%)
CLD	6 (3%)
NYHA	
I	80 (41%)
II	91 (48%)
III	19 (10.5%)
IV	1 (0.5%)

CLD: chronic lung disease; NYHA: functional class according to the New York Heart Association

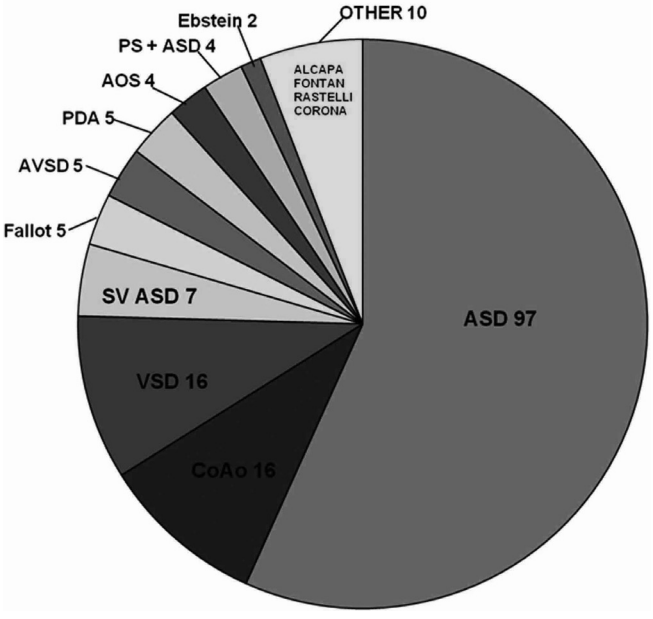

Fig. 2 - Diagnosis and number of patients who underwent a primary correction
ASD: atrial septal defect; CoAo: coarctation of the aorta; VSD: ventricular septal defect; SV ASD: sinus venosus atrial septal defect; Fallot: Fallot's tetralogy; AVSD: atrioventricular septal defect; PDA: persistent ductus arteriosus; AOS: aortic stenosis; PS: pulmonary stenosis; Ebstein: Ebstein's anomaly of the tricuspid valve; ALCAPA: anomalous origin of the left coronary artery from the pulmonary artery; Fontan: Fontan's operation; Rastelli: Rastelli's operation; CORONA: anomalous origin of the coronary artery from the wrong sinus of Valsalva

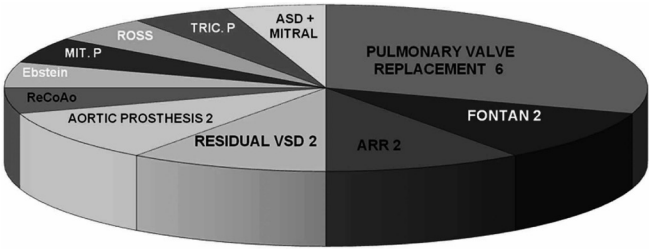

Fig. 3 - Diagnosis and number of patients who underwent a reoperation
ARR: aortic root replacement; VSD: ventricular septal defect; ReCoAo: recoarctation of the aorta; MIT. P: mitral prosthesis; ROSS: Ross operation; TRIC. P: tricuspid prosthesis; ASD + MITRAL: closure of an atrial septal defect plus mitral prosthesis

case of Fontan operation. The causes of death were sepsis due to respiratory infection (n=3), low cardiac output (n=2), cardiac tamponade, sudden death and pulmonary edema, one case each.

In 171 patients who had undergone a primary correction, 33 (19%) had 30 major and 12 minor complications. Among the 30 major complications, the more frequent were the arrhythmias (n=14), mainly atrial flutter or fibrillation (n=9). Among these nine patients with atrial arrhythmias, six of them had been submitted to a secundum ASD closure (Table 2).

In 20 reoperated patients, eight (40%) had 10 major and 3 minor complications. Among the 10 major complications, the most frequent was the acute renal failure (n=3) (Table 2).

The major and minor complications were more frequent in the reoperated patients (n=8, 40%) than in the patients submitted to a primary repair (n=33, 19%) with a tendency for statistical significance (P<0.08).

The major complications were more frequent in the reoperated cases (n=10, 50%) than in the patients submitted to a primary repair (n=30, 18%) (P<0.003).

There was no mortality among the patients submitted to a re-operation albeit the duration of the CPB (P<0.0001) and CLAMP (P=0.0003) were significant longer and having a longer hospital length of staying (P=0.001) than the patients submitted to a primary repair.

Regression results analysis for 30 day-mortality disclosed that male gender (P=0.006), older age at operation (P=0.004) and preoperative functional class III-IV (P=0.004) were considered risk factors for surgical mortality.

Table 2. Immediate postoperative complications in 191 adult patients operated on for treatment of congenital heart disease

PRIMARY CORRECTION (n = 171)		REOPERATIONS (n = 20)	
MAJOR		MAJOR	
Arrhythmias	14	Acute renal failure	3
Atrial flutter	5	Sternotomy review	1
Atrial fibrilation	4	Paraplegia	1
Complete AV block	2	Pleural efusion	1
Junctional rhythm	2	Pulmonary emboli	1
Ventricular taquicardia	1	Stroke	1
Reoperation for bleeding	3	Respiratory insuficiency	1
Pleural efusion	3	Complete AV block	1
Pericardium efusion	3		
Pneumotorax	2		
Hemotorax	1		
Chock	1		
Stroke	1		
Respiratory insuficiency	1		
Arterial right leg thrombosis	1		
MINOR		MINOR	
Systemic hypertension crisis	3	Infection	2
Subcutaneous enphysema	1	Diabetes	1
Recurrent laryngeal lesion	1		
Broncoaspiration	1		
Pneumonia	1		
Hipoglicemia	1		
Pericarditis	1		
Liver disease	1		
Subglotic stenosis	1		
Malnutrition	1		

AV: atrioventricular

Late results

Among the 183 patients discharged from hospital, two died suddenly (one with an ASD and the other a Rastelli operation in a case of congenitally corrected transposition) and another of non cardiac cause. Information regarding late follow up were available in 149 (82%) patients who had a mean follow up period of 5.4±6.1 years (range 4.4 to 6.4 years). One hundred and eleven (74%) of these patients were seen in the last three years.

In 129 patients who underwent a primary correction, 18 (14%) had 11 major and 10 minor complications/morbidities detected during late follow up. Among the 11 major complications/morbidities, the arrhythmias (n=7) were the most frequent, and among them, atrial flutter or fibrillation (n=6). Among the six cases with atrial flutter or fibrillation, five had been submitted to a secundum ASD closure (Table 3).

In the 20 patients who underwent a reoperation, 4 (20%) had two major and two minor complications/morbidities detected during late follow up (Table 3).

There was no significant difference regarding the occurrence of complications between the patients submitted to a primary correction and those who were re-operated (P=0.515).

After operation, a significant number of patients changed their functional class from II to I (41% to 90%) (P<0.0001). At the last outpatient visit, 134 (90%) were in NYHA class I, while 13 (9%) were in class II and 2 (1%) in class III (Figure 4). The numeric transformation of the functional class disclosed a significant difference (P<0.0001) between the mean values before (1.66) and after (1.11) the operation. The actuarial survival curve showed that 96.2% of the patients were alive within six years of follow up (Figure 5).

DISCUSSION

Mainly in view of the surgical techniques improvement during the last 60 years [9], particularly verified in the neonatal treatment, a growing number of patients with congenital heart disease are reaching adulthood [10]. The great majority of these patients, however, can not be considered cured. Sequela and residual lesions are frequent [2] demanding special attention during a period of life when problems related to the individual activities and to the natural aging process might interfere with the lesion physiopathology [11,12]. An interesting and still not well discussed subject, especially in Brazil, is the surgical approach in adults with congenital heart disease, frequently necessary in those patients with late diagnosis and those in need of a reoperation. The data here presented are concerned

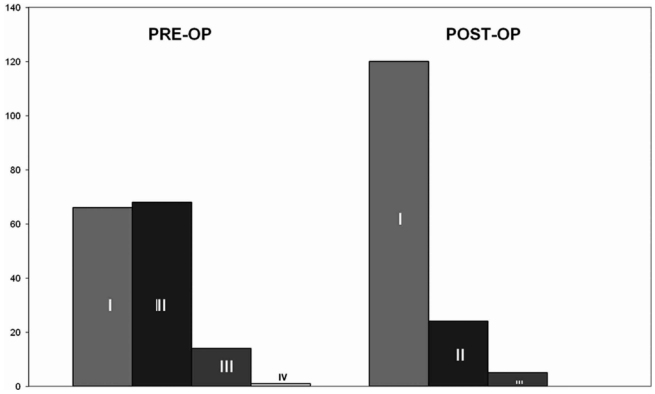

Fig. 4 - Functional class according to the NYHA in 149 patients followed in the long term

Table 3. Late postoperative complications and comorbidities diagnosed in 149 adult patients operated on for treatment of congenital heart disease and followed in the long term

PRIMARY CORRECTION (n = 129)		REOPERATIONS (n = 20)	
MAJOR		MAJOR	
Arrhythmias	7	Arrhythmias	1
Atrial flutter	4	Atrial flutter	1
Atrial fibrilation	2	Aortic regurgitation	1
Supraventricular taquicardia	1		
Pericardial efusion	1		
Aortic recoarctation	1		
Cardiac failure	1		
Right ventricular disfunction	1		
MINOR		MINOR	
Systemic hypertension	8	Systemic hypertension	2
Chronic lung disease	1		
Keloide	1		

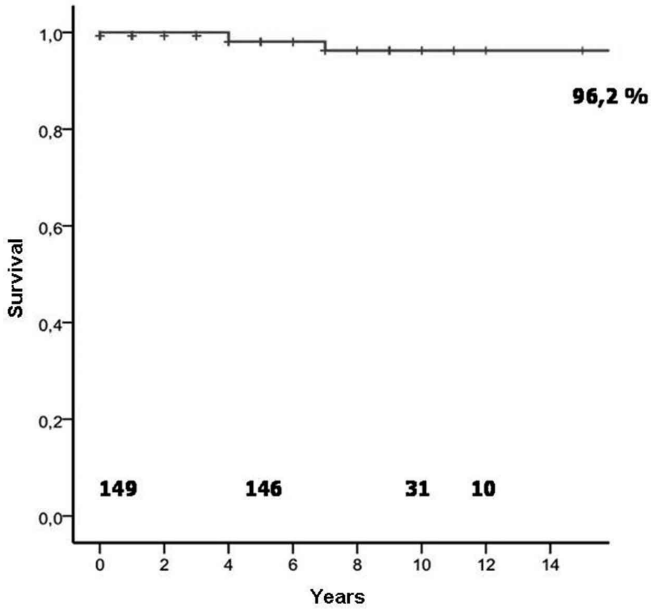

Fig. 5 - Actuarial survival curve in 149 patients followed in the long term

to a tertiary institution dedicated to congenital heart disease treatment for the last 40 years. However, it should be noted that the complex forms of defects started being treated in the last 20 years, which gives a peculiar characteristic to our experience, with an important prevalence of mild and moderately complex lesions differing, in some respect, of the experience of pioneer institutions in the country [3].

Initially, attention should be drawn to the fact that 90% of the patients were treated during the last 11 years. Another interesting aspect is the high proportion of female patients (61%), certainly related to the great number of ASD, a common feature reported in other experiences [13-15]. It should also be emphasized that only 40 (20%) patients lived in the city area (population 600.000 inhabitants). This information is important when the number of patients in continuing follow up in a specific institution is discussed. Extrapolating recently published data [16], the estimated number of adults with congenital heart disease in the Ribeirão Preto city is 1920 patients. In our recently published outpatient data [17] only 23% of 413 patients lived in the city, showing that the great majority of them were not followed in a specialized unity, a finding already reported [18].

Immediate results

The 30-day mortality was 4.2% and numerically comparable with recent publications available where this number varies from 1.5% to 6.3% [13-15,19]. However, a detailed analysis of these experiences shows that comparison with our data is not adequate due to their high number of reoperations [13] and also of very complex cases [19] based on the NYHA functional class. Albeit our patients with III and IV functional class corresponded to only 11% before operation, two of our early deaths occurred in patients with not complex forms of congenital heart disease, ASD and partial atrioventricular septal defect. Despite the relative benignity of the intervention in these types of defects, the possibility of complications has already been reported [20]. Another fact which certainly influences results is the number of operated patients. Our experience is relatively small when compared with other publications where the numbers varied from 438 to 2012 [3,13-15,19].

In the univariate analysis we found that male gender, older age at operation (47±17 years) and functional class III-IV were considered risk factors for death. Albeit the significant statistically difference found, we believe this information should be assessed with caution in view of the small number of deaths. Particularly regarding the age at operation, we found it difficult to establish a number above what the surgical risk should be considered high. Also, known risk factors as pulmonary hypertension [13] and increased CPB time [21,22] had only a tendency for statistical significance in our data.

Postoperative complications occur frequently and can be multiple [13-15]. The prevalence in our material of 21% patients with major complications is similar to the experience reported by Vida et al. [14] where this number was 22%. However, it differs considerably from the results of an Italian multicentre study: 29% of 856 patients who had undergone surgery [15] and two factors could possibly explain this difference. Unlike this last investigation, we did not consider as complication the prolonged ventilatory time in the intensive care unit. Also, in a multicentre study, it is possible that the increased number of complications is contaminated by events originated from groups with small experience which participated in the study.

It is interesting to note that the occurrence of complications was greater in the patients who underwent a reoperation (40%) than in those submitted to a primary correction (20%). However, the statistical analysis showed that this difference had only a tendency to significance ($P<0.08$). When we assess only the major complications, it is clear that they tend to occur more frequently in the reoperated patients ($P<0.003$). It should be remembered that the number of reoperated patients was small (n=20) with no early mortality despite the fact that they have required a prolonged time of CPB, CLAMP and intensive care stay. It is well known the fact that reoperations are more demanding and have a significant morbidity, albeit low mortality can be accomplished when the procedure is done in centers dedicated to adult congenital heart disease treatment [22].

As already reported [13-15], the arrhythmias are the most frequent complications and, among them, atrial flutter or fibrillation, which also occurred in our patients and were

commonly associated to a secundum ASD closure. This type of complication deserves special attention, mainly due to the fact that it is the most common cause of hospital admission in adults with congenital heart disease [23].

Late results

Among 183 patients discharged, three of them died during follow up, one of a non cardiac cause and two probably due to a cardiovascular complication since the nature of the death was sudden. This low late mortality rate makes a risk factor analysis impossible. However, it is well known that arrhythmias, cyanosis, smoking and depressed left ventricular function are considered risk factors for late death in this group of patients [13].

The analysis of the late results, usually hard to be accomplished in our country, was possible in 149 (82%) patients and, despite better than the index reported by a multicenter European study (68%) [14], it is inferior to those registered in a multicenter Italian study (87%) [15] and in a Dutch study (89%) [13].

The mean follow up period of our patients was 5.2 ± 6.1 years (median: four years, CI 95%: 4.41 – 6.38 years) which characterize our follow up information as a medium term one. During the last three years 111 (74%) of these patients have routinely been seen at the outpatient clinic. This 26% of patients lost to follow up is worrisome, and, as already emphasized [14] a policy of active search of patients should be undertaken by any tertiary unit in order that the intervention results are known.

Despite the incidence of complications and comorbities found during late follow up were more prevalent in the reoperated patients (20%) than in those submitted to a primary repair (14%) the difference had no statistical significance. It should be noted the important occurrence of arrhythmias in these patients (8/22) with significant prevalence of atrial flutter or fibrillation (six cases), frequently associated to a secundum ASD closure.

Albeit the criteria for functional class classification recommended by the New York Heart Association is widely applied, recent studies have demonstrated that this method underestimates the real individual capacity [24]. However, it is generally used as a parameter of cardiovascular function and being particularly useful when a group of patients is assessed before and after an intervention. In the data here presented an analysis of the total number of patients was done aiming to have an idea of the general degree of improvement after operation. Obviously a detailed analysis by groups of patients is desirable, however difficult in our cases due to the small proportion of patients in some groups. Taking into account this information, a significant migration from class II before operation to class I after the procedure (41% to 90%) was noted, which is comparable to the experiences of Putman et al. [13] (80%), Vida et al. [14] (90%) and Padalino et al. [15] (79%). The numeric transformation of the functional class has not been frequently used [8]. However, we believe it can be useful when a group of patients is assessed, since it reflects the occasional difficulty in classifying a specific patient in one or another functional class. The mean value difference was significant, which also allowed us to conclude that our patients were not so physiologically impaired. We believe this parameter should be continually evaluated in this group of patients, however, recent investigations have suggested that a more objective assessment be used for cardiovascular function analysis, like the ergoespirometry. The results of this test may disclose patients under higher cardiovascular risk for complications and death, indicating which of them should be more carefully followed up [25].

The survival probability according to the Kaplan-Meier method was 96.2% at six years, very similar to the numbers of Padalino et al. [15] (96% at five years) and Vida et al. [14] (97% at five years). Due to the great diagnostic variability usually found in groups of adult patients with congenital heart disease, an actuarial analysis based on groups of defects is difficult in our cases, which would be feasible with more patients.

Despite the parameters above discussed have been widely employed for late results evaluation of patients followed in the long term, we share the opinion that the individual quality of life is an important aspect which should be considered in the outpatient assessment because it can give a more complete idea of the individual status [12,26].

Based on these pieces of information, we believe that the surgical treatment of adults with congenital heart disease can be accomplished with low mortality, depending on the severity of the disease. The immediate complications are frequent, particularly the arrhythmias. The long term follow up of these patients is crucial in order to know the results of the procedures. During this period, the residual lesions are frequent, specially the arrhythmias. Also, the occurrence of other morbidities is important and should be adequately addressed. Proper advice to the patient at discharge is mandatory, also to the physician in charge of the follow up [27].

Due to the excellent improvement in the invasive treatment of children with congenital heart disease a growing number of patients with complex lesions will reach adulthood. Consequently, the profile of these adult patients will change with more reoperations and percutaneous treatment being needed [28]. Considering the special characteristics of the material here presented, it should not be considered as the usual spectrum of patients to be found in centers with large experience in congenital heart disease treatment. Due to this regional characteristic, certainly present in other centers in the country, to the great diagnostic variability usually found and to the different surgical strategies employed in the treatment of these patients, we believe that multicenter studies are of extreme importance. This model of investigation [29], of crescent interest in the international literature will allow us

to know the profile of our patients, to verify the results of the procedures adopted and to discuss the efficacy of different therapies. Adults with congenital heart disease constitute a very special group of individuals, many of them survivors of several operations. The tendency is that the number of these patients will increase dramatically [30] and the institutions dedicated to this type of assistance should be ready to work efficiently. Due to the absolute absence of reports regarding this subject among us, comparison of our results becomes difficult.

Despite the comparison of our data with those disclosed by a recent report [3] coming from a pioneer center in the country is possible in some aspects, they both differ fundamentally in terms of patient population, institutional characteristics, detailment of postoperative complications and information regarding follow-up. These differences, as we can see it, add to the informations we consider important regarding the management to these patients.

Study limitations

The data here presented reveals the institution experience. The number of old patients with lost informations is probably very small and we believe it would hardly change the results. The great diagnostic diversity makes it difficult to assess the results by groups of patients. Some important parameters related to the immediate results like the intensive care length of mechanical ventilation were not studied but we believe that the information available reflect the usual outcome of these patients. The long term results analysis was improved through an active search of patients lost to follow up and should be considered when the results with other studies are compared.

Authors' roles & responsibilities	
FTVA	Research design, medical records review, data analysis and writing
AJR	Satistical analysis and final writing
PHM	Medical records review and writing
AS	Research design and final writing
MFK	Medical records review and data organization
CC	Data organization, medical records review
RNS	Medical records review and data analysis
WVAV	Research design, data analysis and final writing

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