

Evaluation of Surgical Treatment of Congenital Heart Disease in Patients Aged Above 16 Years

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Abstract

Background: The increasing number of children with evolving congenital heart diseases demands greater preparation of professionals and institutions that handle them.

Objective: To describe the profile of patients aged over 16 years with congenital heart disease, who have undergone surgery, and analyze the risk factors that predict hospital mortality.

Methods: One thousand five hundred twenty patients (mean age 27 ± 13 years) were operated between January 1986 and December 2010. We performed a descriptive analysis of the epidemiological profile of the study population and analyzed risk factors for hospital mortality, considering the complexity score, the year in which surgery was performed, the procedure performed or not performed by the pediatric surgeon and reoperation.

Results: There was a significant increase in the number of cases from the year 2000. The average complexity score was 5.4 and the septal defects represented 45% of cases. Overall mortality was 7.7% and most procedures (973 or 61.9%) with greater complexity were performed by pediatric surgeons. Complexity (OR 1.5), reoperation (OR 2.17) and pediatric surgeon (OR 0.28) were independent risk factors influencing mortality. Multivariate analysis showed that the year in which the surgery was performed (OR 1.03), the complexity (OR 1.44) and the pediatric surgeon (OR 0.28) influenced the result.

Conclusion: There is an increasing number of patients aged 16 years which, despite the large number of simple cases, the most complex ones were referred to pediatric surgeons, who had lower mortality, especially in recent years. (Arq Bras Cardiol 2012;98(5):390-397)

Keywords: Heart defects / surgery; adolescent; hospital mortality.

Introduction

Surgical treatment of congenital heart disease has changed the natural history of the disease. Prior to this specialty, only those patients with diseases of lesser complexity managed to survive and reach maturity¹. More complex diseases are rarely seen in adult patients. The dramatic success of pediatric heart surgery and, more recently, advances in interventional cardiology applied to this population have reversed this situation. Diseases with poor prognosis began to evolve into adulthood more often^{2,3}. The knowledge required to take care of these patients is more complex and must include training in internal medicine, general cardiology, pediatric cardiology and their subspecialties. The significant increase in this population, the lack of professionals with expertise in this area and Medical Centers prepared to provide proper health care is a worrying challenge to the existing health system, especially for tertiary healthcare centers^{1,3}.

There is much discussion on the surgical treatment of adult patients with congenital heart disease. What is the best scenario to get the best results? Should these patients be operated by pediatric surgeons in general cardiac surgery services or by general heart surgeons in pediatric hospitals? Should they be operated in pediatric hospitals or general hospitals?²⁺⁷ In Brazil, this question becomes a little more difficult to be answered, because there are few pediatric cardiovascular surgery services located in pediatric hospitals, and there are few specialist pediatric surgeons or others prepared for such cases. Most services are located in general hospitals and some are specialized in cardiology, such as the Heart Institutes, where pediatric cardiac surgery coexists with general cardiovascular surgery.

Being acquainted with our reality, the profile of patients and the results, and planning an appropriate program for taking care of this population has become an obligation that the Brazilian tertiary institutions are able to fulfill.

The purpose of this study was to describe the profile of adult patients with congenital heart disease operated at a large Health Care Service in the last 24 years. In addition, hospital survival was analyzed and correlated with the complexity of the cases, the type of surgeon involved in the procedure, the occurrence of reoperation, group of diagnosis and procedures as well as the year in which these were carried out.

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Manuscript received September 08, 2011, revised manuscript September 08, 2011; accepted January, 03, 2012.

Methods

We studied patients aged over 16 years, operated by the Surgical Division of Instituto do Coração do Hospital das Clínicas of Universidade de São Paulo from January 1986 to December 2010. The epidemiological profile of patients, the procedures performed, the distribution of according to diagnoses (grouped according to the international nomenclature modified for institutional use⁸ with the surgeon who performed the procedure (pediatric and non-pediatric surgeon) and the results obtained make up the profile of this analysis.

Data are presented descriptively in proportions or mean values with a confidence interval of 95% (CI 95%). In comparing the mortality in major groups of procedures performed by pediatric surgeons and non-pediatric surgeons, the Fisher exact test was used. Influence of variables: the year in which the operation was performed, patient age, reoperation, complexity of the procedure and the surgeon responsible for the surgery versus mortality was assessed by the logistic regression model. The variables were studied in isolation and those that had a significance level below 0.1 were subjected to multivariate analysis. Statistical analysis was performed using the software SPSS for Windows version 17.0.

Results

During the period of study, 1,571 procedures were performed in 1,520 patients aged over 16 years (mean age 27 ± 13 years). The oldest patient was 78 years old, diagnosed with atrial septal defect.

The growth trend in the number of procedures performed at the institution may be observed in three different periods: phase I, from 1986 to 1989, phase II, from 1990 to 1999, and phase III, from 2000 to 2010. The trend of steady growth in the total number of patients operated for congenital heart disease regardless of age contrasts with that observed in the subgroup of patients operated for congenital heart disease, older than 16 years. The average annual number of all procedures in these phases was: 403 in phase I; 454 in phase II; and 685 in phase III. The annual averages of procedures for the subgroup older than 16 years were: 52.3 in Phase I; 51.3 in phase II; and 84.3 in phase III. That is, there was a steady increase in the number of procedures from 12.7% between phase I and II, and 50.8% between phase II and III, considering the group as a whole. However, in the subgroup of patients aged over 16 years, there was virtually no change between phases I and II, but there was a growth of 62% between phases II and III (Figure 1).

In this subgroup of patients aged over 16 years, the global average of scores for the scale of complexity Aristotle Basic Score (ABC)⁹ was 5.4, and the overall mortality rate was 7.68%. Most procedures consisted of surgeries performed in patients without any previous surgery, and reoperations were only 180 (11.45%). As for the distribution of the leading groups of diagnosis, it is observed that the group of septal defects corresponded to the highest number of cases ($n = 698$ or 45%), followed by the group of right heart anomalies ($n = 278$ or 18%) and anomalous pulmonary venous return ($n = 241$ or 16%) (Figure 2).

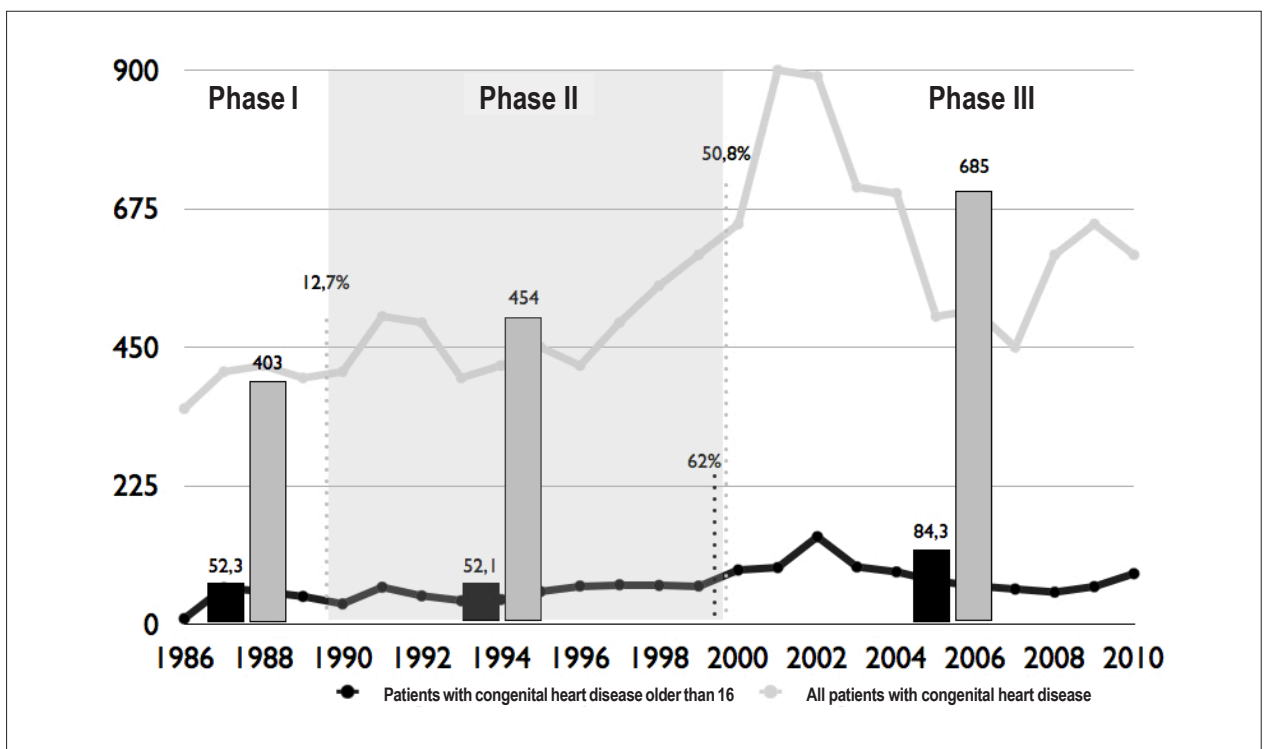


Figure 1 - Number of procedures performed annually (lines), comparing the averages (bars) by phases (I: 1986-89, II: 1990-99 and III: 2000-10). There is a steady growth in the group of patients operated with congenital heart disease (phase II: 12.7% and phase III: 50.8%) and a growth of 62% in the year 2000 (phase III) in the subgroup of patients aged over 16 years. Considering phase III alone, we observe that the number of cases in patients aged over 16 has grown more.

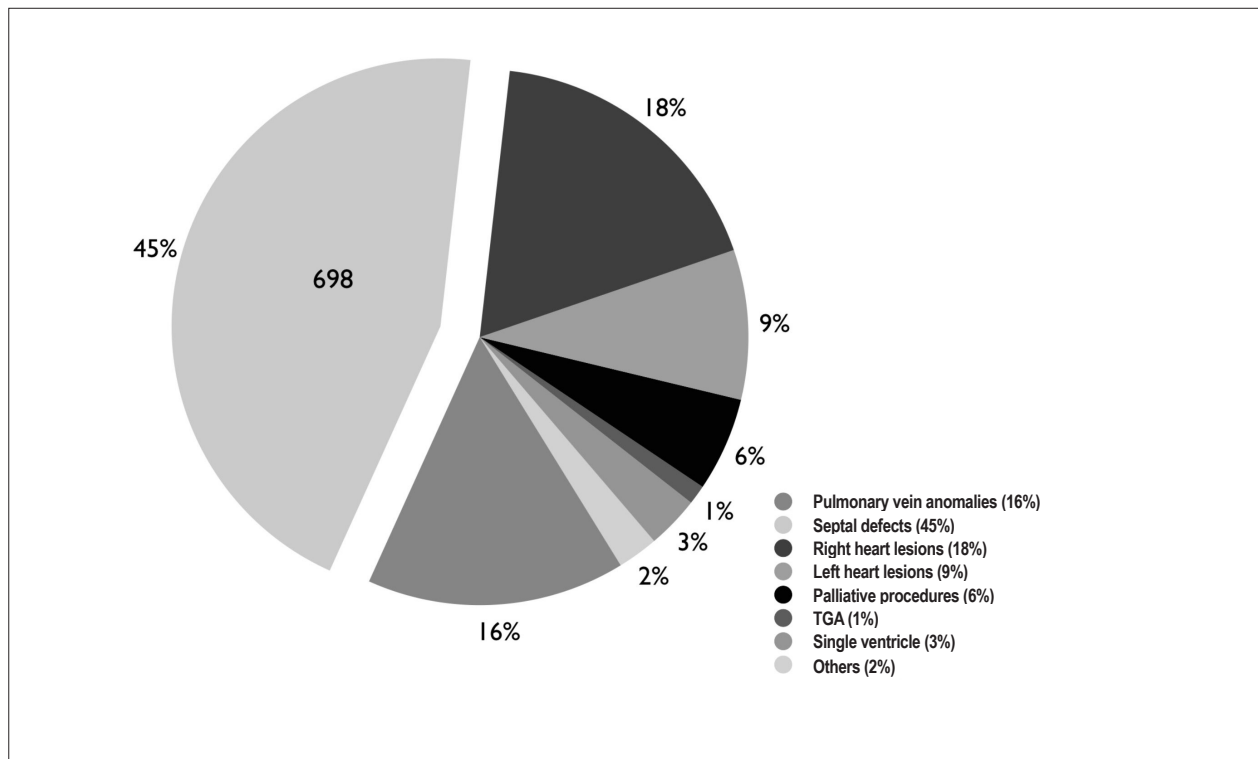


Figure 2 - 1,571 procedures performed in patients aged over 16, according to anatomical classification.

The closure of isolated atrial septal defects (n = 530) and atrial septal defects associated with partial anomalous connection of pulmonary veins (n = 54) accounted for more than one third (37%) of all procedures performed.

In 180 patients who had previously undergone surgery (reoperation), either or not performed in our department, diagnoses of right heart anomalies (35% of cases), left heart anomalies (16.7%) and patients with atrioventricular univentricular connection (single ventricle) (15.5%) were more common. The most common procedure among these cases, 23.4%, was reoperations for reconstruction of the right ventricular outflow tract (RV-PT exchange in 22 patients and prosthetic valve implantation in pulmonary position in 21). In the group of left heart anomalies, changes of valves in mitral and aortic positions accounted for 60% of procedures performed in this group. In those diagnosed with single ventricle, 14 patients completed total cavopulmonary operation over the age of 16 years, 9 underwent bidirectional Glenn surgery and shunt was performed in 3 patients.

The procedures performed by pediatric surgeons comprise the largest volume (973/1520 = 61.9%) and the highest proportion of more complex cases (average complexity of 5.99 - Aristotle Basic Score - compared to the average complexity of 4.39 observed in patients operated by general cardiac surgeons).

Figure 3 shows the relative distribution of the main groups of procedures performed by pediatric surgeons and non-pediatric surgeons.

The absolute distribution of the type of surgeon according to the main groups of diagnosis and observed mortality is summarized in Table 1.

Although the more complex cases were operated mainly by the group of pediatric surgeons, there was no statistically significant difference in mortality rate among such cases compared with those of smaller complexity operated by general surgeons.

The historical series presented in Figure 4 shows the significant improvement in the mortality rate in the last 10 years. In the early years of this experience, the year of the surgery proved to be an independent risk factor associated (OR = 1.03 - 95% CI: 1.001-1.059) to the complexity and type of surgeon who operated the patient.

In the multivariate analysis, the type of surgeon, the complexity of the procedure and the presence of reoperation were independent risk factors for mortality (Table 2). The analysis of these risk factors combined, complexity, type of surgeon and year in which the operation was carried out shows that the more complex cases operated by pediatric surgeons showed better results than when operated by non-pediatric surgeons (Table 3).

Discussion

About 5 to 8 cases per 1,000 live births present some type of congenital heart disease. It is considered the first non-infectious cause of death in newborns¹⁰. Of these children, 30% require surgical treatment and/or intervention

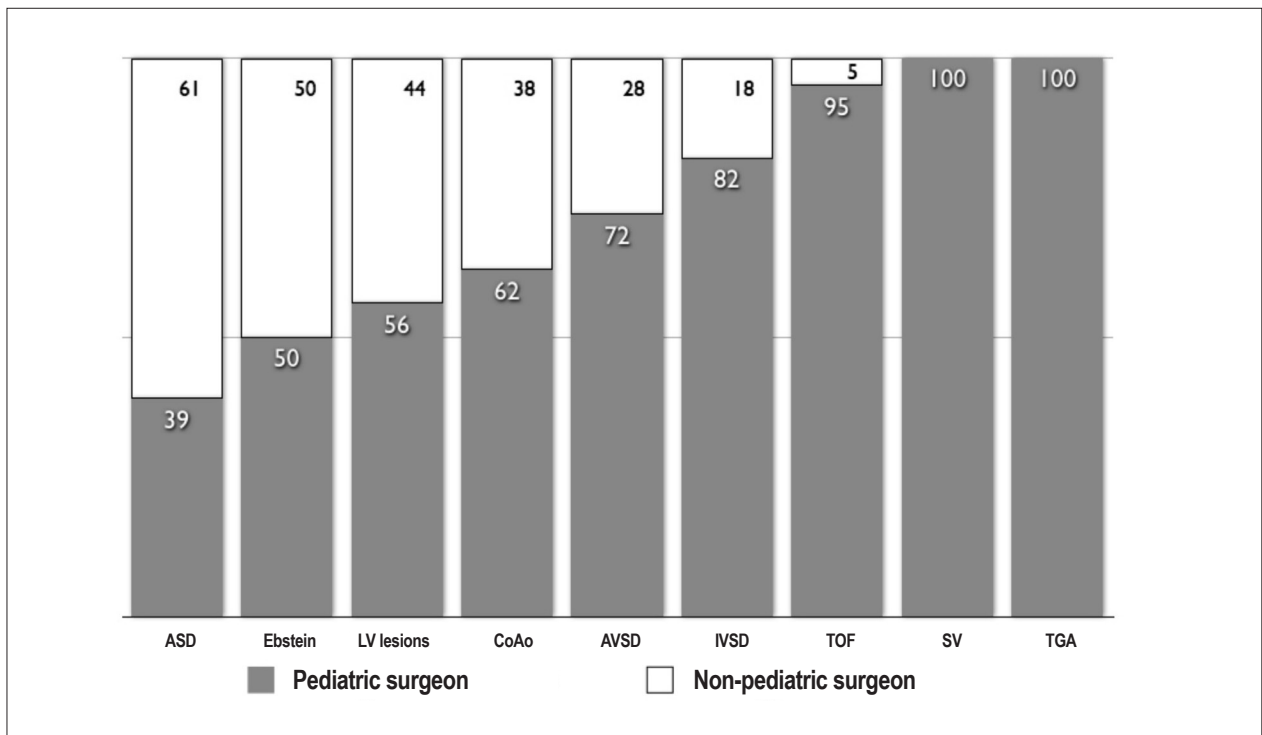


Figure 3 - Percentage distribution of procedures performed by type of surgeon (either pediatric or non-pediatric), main diagnoses, from lower to higher complexity

Table 1 - Main procedures performed according to the type of surgeon, distributed through different groups according to the primary diagnosis

	Pediatric surgeon (mortality %)	General heart surgeon (mortality %)	p value (Fisher's exact test)
Septal defects	328 (5)	355 (4)	0,7442 (ns)
ISD	212	318	
CIV	83	19	
AVSD	33	18	
LV anomalies	55 (11)	45 (4)	0,1624 (ns)
RV anomalies	102 (3)	28 (3)	1,00 (ns)
TOF	78	3	
Ebstein	24	25	
Single ventricle	60 (10)	0	
Thoracic arteries and veins	129 (2)	58 (3)	0,1739 (ns)
CoAo	72	38	
PDA	57	20	
TGA	21 (3)	0	

ASD - Atrial septal defect; VSD - Ventricular septal defect; AVSD - Atrioventricular septal defect; LV - left ventricle; RV - Right ventricle, TOF - Tetralogy of Fallot; CoAo - Coarctation of the aorta; PDA - Patent ductus arteriosus; TGA - Transposition of the great arteries.

in the first year of life^{11,12}. With the huge advance in pediatric cardiac surgery and improvement of results, it is estimated that today there are more than 1 million survivors with congenital heart disease in the United States¹³. The

exact size of the population of patients with congenital heart disease in adulthood is estimated between 80% and 85% of patients born with some type of congenital heart disease¹⁴. Although the age limit to be considered an adult

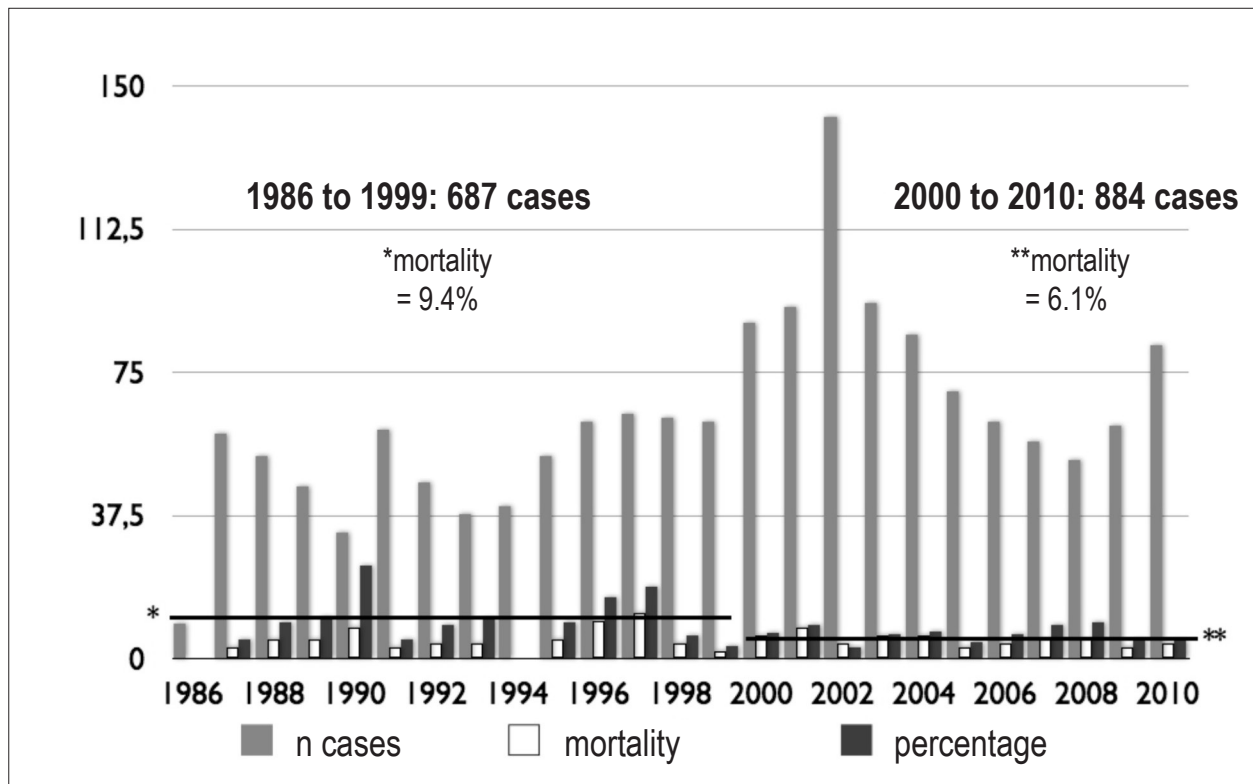


Figure 4 - Mortality in two different periods of the 1,571 procedures with an overall mortality of 7.6% divided into 2 periods: 1986 to 1999 (687 cases) and 2000 to 2010 (884 cases). There was an improvement in the last period, although such improvement was not observed in the year of operation in isolation from the beginning to the end of this experience.

Table 2 - Independent of predictors of hospital mortality

Variable	Odds Ratio	p	Confidence Interval 95%
Reoperation	2.17	0.001	1.355 to 3.477
Complexity (Aristotle Basic Score)	1.51	<0.0001	1.366 to 1.672
Performed by a pediatric surgeon	0.28	<0.0001	0.170 to 0.475
Year in which the procedure was performed	0.99	0.074	0.976 to 1.004
Patient's age	125 ± 14	1.02	0.997 to 1.055

Table 3 - Multivariate analysis, Cox proportional regression

Variable	Odds Ratio	p	Confidence Interval 95%
Year in which the procedure was performed	2.17	0.103	1.001 to 1.059
Complexity (Aristotle Basic Score)	1.51	1.446	1.300 to 1.607
Performed by a pediatric surgeon	0.28	0.48	0.280 to 0.823

is 18 years, as defined by the World Health Organization (WHO), in our country it is a bit confusing in clinical practice. According to the Ministry of Health, the services of high cardiovascular complexity can only serve patients

aged 12 or more, requiring a special accreditation for the service to be considered a pediatric cardiovascular service (Ordinance 210, SAS-MS, 2004). The exclusive pediatric cardiovascular service, in turn, will be authorized to serve

patients in the neonatal period up to 18 years of age. Our hospital specializes in cardiovascular diseases and, although it is not a pediatric hospital, it has a group of professionals dedicated to medical and surgical treatment of children with heart disease. Although patients with congenital heart disease are referred to the pediatric ward, some patients older than 16 years are admitted in other places, especially patients with valvular disorders or atrial septal defect. Thus, we have arbitrarily included all patients aged 16 years or older in this study, which appears to be more representative of the Brazilian clinical practice in the care of patients considered "adults" in our country.

Considering the current U.S. population, we observe an annual increase of 2,500 adults with congenital heart disease^{2,15}. In Brazil, the database of our Public Healthcare System (DATASUS) shows that 70% of patients diagnosed with congenital heart disease survive to the age of 18. Some interesting studies have attempted to scale the growth prospect of a clinic for congenital heart disease in adults, indicating that for every 100,000 live births, 200 cases will reach adulthood each year (18). This represents an annual increase of about 5,800 cases of adults with congenital heart disease requiring medical follow-up at the outpatient care in our country. Patients operated may account for about 53% to 55% of the volume of follow-ups as shown by all published national experiences^{16,17}.

Considering the referential data mentioned above, after 18 years of operation, a healthcare service with surgical volume averaging 500 cases per year should reoperate 110 to 120 patients more, considering only their patients being followed up. That would mean that a cardiovascular surgery service like the InCor-HCFMUSP, in the past seven years, in order to assist their own surgical patients, should have been performing a linearly higher number of congenital heart disease procedures of at least 100 more cases per year. In the period studied, taking the average of operated congenital heart diseases, this was not observed. Considering the number of adult patients operated, although we have observed an increase in the number of cases since 2000, the annual average was only 78 cases per year in the last seven years.

The population of adults with congenital heart disease includes those previously operated in childhood, as well as those who had their disease diagnosed in adulthood. This is due to the difficulty faced by these patients in accessing specialized tertiary centers or due to the diagnosis not established by the doctor who followed them up in the primary healthcare service. The difficult access to specialized centers can be explained by the insufficient number of centers or by their geographical distribution. Imprecise diagnosis and lack of proper behavior can be explained by inadequate training.

Among the different types of procedures required for these patients, we could group them as follows¹⁸: 1) primary corrections of untreated congenital defects (congenital heart diseases not operated before age 18); 2) surgical treatment of residual lesions in patients previously operated; 3) surgical treatment of new diseases associated with congenital heart diseases; 4) surgical treatment of valve injuries operated or

not; and 5) treatment of acquired diseases in patients with congenital heart disease operated or not.

About 40% of these cases are considered of low complexity and can be easily treated in most centers of cardiology and cardiovascular surgery¹⁹. This sample corroborates these data, where it was observed that 45% of cases are within the group of septal defects, and correction of atrial septal defect is among the leading procedures in the whole sample. In turn, 20% to 25% of these cases are considered complex, rare and require extremely specialized and multidisciplinary care¹⁹, requiring huge efforts from the center responsible for the care they need. These cases require clinical cardiologists specializing in adult congenital heart disease, intensive care experts, electrophysiologists, imaging specialists (MRI, computed tomography), interventional cardiology, onsite pregnancy risk service, active transplant program with family support, trained nurses, cardiovascular rehabilitation services and psychology, among others²⁰. The more complex cases are usually directed to the pediatric surgeon, as seen in this experiment, and these surgeons often required, in addition to knowledge in congenital heart disease, preparation of all cardiovascular surgical subspecialties.

In our environment, access to specialized centers is often hampered by geographical distances, the concentration of these centers in more developed areas and the small number of centers available. This implies that less symptomatic patients seek medical help late in the natural evolution of the disease and that could explain the large number of patients operated in this service for the first time in adulthood, over 16 years. Those previously operated and under follow-up return to the procedures necessary in evolution, such as tetralogy of Fallot, in patients with univentricular physiology under staged surgical treatment and congenital valvular lesions previously treated, as shown in the results of this analysis.

From the surgical point of view, patients with congenital heart disease in adults present a large number of associations of various cardiac and non-cardiac diseases, requiring the surgeon's knowledge not only of congenital heart disease, but also of how to handle valvular operations, diseases of the aorta, arrhythmia and other subspecialties¹. The possibility of applying all this knowledge in an integrated manner can perhaps provide a better outcome in this group of patients. Although the result may be more successful when the patient is treated by a pediatric surgeon as shown in the literature³⁻⁷, the real impact of a pediatric surgeon specializing in adults in these results had not been analyzed systematically. In this study, the fact that the patient of higher complexity has been treated by a pediatric surgeon had an impact on the result observed when we analyzed mortality. These data, however, do not allow a deeper analysis of the results regarding complications and the surgical effectiveness²¹. In this analysis, we used the score of complexity described for congenital heart diseases in general, the Aristotle Basic Score (ABC). The ABC seems to have a suboptimal predictive power in adults²². However, as one does not have any specific score validated for this group of patients, the ABC is a possible way to stratify the complexity of patients operated.

Among the current structural challenges, we can cite the difficulty in identifying the best center to perform the service, which can be a pediatric hospital, a hospital for adults or a center specializing in cardiology and cardiovascular surgery⁵⁻⁷. Providing outpatient prenatal care for women pregnant with fetuses with cardiac disease and care in the postpartum period, as well as proper infrastructure and personnel specializing in non-cardiac fields like nephrology, hepatology, pulmonology, hematology, among others, is the great challenge to serve this population.

Providing full care of patients with congenital heart disease, either operated or not, requiring surgical treatment as a result of progression of the disease operated or diagnosed in adulthood, becomes a challenge for the future. Organizing a working group specializing in adult congenital heart disease, in line with the group of pediatric cardiology and other specialties within cardiology is mandatory to ensure best results and most appropriate care for these patients. Moreover, it is important to support the specific needs of this group of patients (such as arrhythmia, for example), establishing protocols for care and follow-up.

Although this study is a retrospective analysis of medical records of patients treated at the institution, the data supports the findings in the literature. Hence, there is a need for surgeons specializing in the treatment of this important group of adult patients with congenital heart disease, as well as centers dedicated and modified to treat them in a specialized and integrated manner, from the perspective of their needs and particularities.

We have observed in this study an increasing number of patients aged over 16 years in the last 10 years. Although most of these procedures are of low complexity, the more complex ones were referred to pediatric surgeons. Separately, the complexity of reoperation and whether it was performed by a pediatric surgeon or a non-pediatric surgeon have influenced mortality.

The results found emphasize that the most complex cases of congenital heart disease should be referred to the pediatric surgeons and a learning curve is expected in the management of these patients, since we observed a lower mortality in recent years.

Acknowledgement

Our thanks to Prof. Dr. Rodolfo A. Neirotti for his detailed review of the content and helpful comments.

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 study is not associated with any post-graduation program.

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