Research Article

Interventional Cardiology

Clinical-epidemiological profile of congenital and acquired heart diseases admitted to a private hospital

Abstract

Objective: To characterize the clinical-epidemiological profile of children with heart diseases (congenital, acquired and arrhythmias), conducted by the pediatric cardio group of a private general hospital, from January 2016 to March 2020.

Method: Retrospective longitudinal observational interference study, with quantitative design, carried out in a private hospital. A total of 502 patients were selected according to the following inclusion criteria: patients who had congenital heart disease, acquired heart disease and arrhythmia, all managed by the pediatric cardiomyopathy group. Descriptive analysis of absolute and relative frequencies were represented through tables and graphs.

Results: Of the 502 patients included in the study, 86.5% had congenital heart disease, 7.7% acquired heart disease and 5.8% arrhythmias. 50.2% were female, 49.6% male and 0.2% were not defined so far. Prematurity occurred in 19.3% of cases, presence of syndromes in 31.2 % and associated comorbidities in 34.7%. In total, 530 procedures were performed, of which 77% were surgeries, 18.5% hemodynamic procedures and 4.5% hybrid procedures. A risk score called RACHS-1 (adjusted risk for surgery and congenital heart disease) was used to categorize the surgeries. There was a predominance of category 3 (38.4%), followed by category 2 (28.6%), category 1 (18.2%), category 4 (9.6%) and category 6 (5.2%).

Conclusion: The complexity of the profile of the found heart diseases reiterates that measures are needed for an integrated approach of different specialties and professionals for the treatment of these patients.

Keywords: Congenital heart diseases ; Epidemiology ; Incidence ; Prevalence

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Introduction

Congenital Heart Diseases (CHDs) are defined as abnormalities in the cardio circulatory structure or function that are present since birth and can have a significant functional impact on the life of the individual [1]. They represent the majority of all malformations in live-born babies and are the most common cause (30% to 40%) of mortality among birth defects [2].

The prevalence of CHDs in the world population is uncertain due to the lack of records in many countries, but it ranged from 5 to 8 per 1,000 live births before the introduction of Doppler echocardiography and 8 to 12 after the widespread use of this diagnostic method [3].

In Brazil, the presentation of CHDs is similar to those observed in other nations, generating an alert that the health system should be prepared to diagnose and treat these patients as early as possible to reduce costs, prevent possible sequelae, and mitigate the emotional distress of the patients and their families [4]. In addition, due to substantial advances in the management of CHD, including improvements in

surgical techniques, intensive support, pharmacotherapy, and early detection, treatment and results have improved drastically in recent decades [5]. Thus, many of these new-borns will survive and thus increase the population of adults with CHD, which reinforces the importance of understanding the epidemiology of these diseases in order to support valuable changes in health policies and clinical practices [6].

Recognizing the profile of this population is essential because CHDs are among the main causes of neonatal morbidity and mortality, and their prevalence in the population is growing; however, cases are underreported and few studies on them have been published in Brazil, which reinforces the importance of producing epidemiological studies. The compilation of relevant information on the characteristics of children with CHD can reveal indicators related to the prevalence of cases, good care practices, and specific outcomes, contributing to the improvement of health services.

In this context, the objective of this study was to understand the case series of congenital heart disease patients monitored in a private hospital of São Paulo and to report the clinical profile of these patients. These records will serve as a basis for future studies and will allow for national and international collaborations.

Materials and Methods

This was a retrospective, longitudinal, observational interference study with a quantitative design that was conducted at private hospital and philanthropic in the city of São Paulo that has neonatal intensive care unit, paediatric intensive care unit, and paediatric ward beds. It has a monthly average of 4.6 admissions/month for treatment related to paediatric heart diseases (congenital, acquired, and arrhythmias). The patients were selected through the electronic medical records database, the TASY System, through the analysis of medical records of hospitalizations from January 2016 to March 2020.

The cardiac paediatric group of this private hospital selected 502 patients according to the following inclusion criteria: patients with congenital heart disease, acquired heart disease, or arrhythmias. Data were collected through the use of an instrument that covered clinical, sociodemographic, preoperative, and postoperative variables. It was formulated in RED CAP software, a clinical research management platform that is designed to create databases and that meets all regulatory requirements related to information security.

The patients were coded according to the International Classification of Diseases (ICD-10) and the International Pediatric and Congenital Cardiac Code (IPCCC). The main diagnosis was defined as the most complex congenital cardiac injury.

After collection, we organized the data in Excel spreadsheets, in which we ran descriptive analyses of the absolute and relative frequencies, represented by tables and graphs. This study was approved by the ethics committee, which waived the need for informed consent.

Results

In total, 502 patients were treated by the cardiac paediatric group from January 2016 to March 2020. Of these, 86.5% had congenital heart defects, 7.7% had acquired heart defects, and 5.8% had arrhythmias. A total of 50.2% were female, 49.6% were male, and 0.2% had an unrecorded sex.

Most of the patients admitted to hospital were admitted for elective surgery, followed by transfer from an external hospital and being born in our obstetric center.

Approximately 34.7% had related comorbidities. Seizures, protein-calorie malnutrition, and encephalopathies were the main symptoms found. Syndromes were diagnosed in 126 patients, while 31 were under diagnostic investigation. Down syndrome was the most common, representing 58% of the syndromes.

Regarding prematurity, 97 patients were born at less than 37 weeks, representing 19.3% of the study patients. The table shows the distribution between extremely preterm, very preterm, moderate preterm, and late preterm infants [7].

The distribution of the types of congenital heart disease, acquired heart disease, and arrhythmias are described. The most frequent cyanogenic heart diseases found were tetralogy of Fallot (10.56%), transposition of the great arteries (6.38), hypoplasia syndrome of the left heart (5.98%), truncus arteriosus (2.59%), right ventricular outflow tract double (2.99%), and pulmonary atresia with interventricular communication (4.18%). The most frequent acyanogenic heart diseases were total atrioventricular septal defect (10.56%), interventricular communication (9.96%), aortic coarctation (9.56%), interatrial communication (8.36%), and persistence of the ductus arteriosus (6.57%).

The distribution of the type of cardiac physiology showed that 81.1% of patients had biventricular physiology and 18.9% had univentricular physiology. Among the patients treated by the cardiac paediatric team, 445 (88.65%) underwent some type of invasive intervention, such as surgery, haemodynamic procedure, or hybrid procedure. The need for combined interventions was observed in several cases. In total, 530 procedures were performed, of which 77% were surgeries, 18.5% were haemodynamic procedures, and 4.5% were hybrid procedures. Categorizing the surgical procedures by the adjusted Risk for Surgery in Congenital Heart Disease (RACHS) score, we found a predominance of category 3 (38.4%), followed by category 2 (28.6%), category 1 (18.2%), category 4 (9.6%), and category 6 (5.2%). Most of the patients subjected to invasive procedures were neonates (31%) or infants (45.8%). The tables shows the age distribution at the intervention date (Table 1-4).

Table 1: Presence of syndromes		
Syndromes	**N**	**%**
Down syndrome	91	58
Genetic syndromes to be clarified (dysmorphisms and malformations)	31	19.7
Goldenhar syndrome	3	1.9
Scimitar syndrome	1	0.6
Pentalogy of Cantrell	2	1.3
Edwards syndrome	6	3.8
Dandy–Walker syndrome	3	1.9
VACTERL syndrome	2	1.3
Cornelia Lange syndrome	1	0.6
Treacher–Collins syndrome	1	0.6
Chromosomal alterations	3	1.9
Bland–White–Garland syndrome	1	0.64
Branchio-Oto-renal syndrome	1	0.64
Aarskog syndrome	1	0.64
George syndrome	7	4.46
Turner syndrome	1	0.64
Disabsorptive syndrome	1	0.64
Kartagener syndrome	1	0.64

Table 2: Prematurity in weeks		
Classification	**N**	**%**
Extreme preterm (<28 weeks)	13	13.4
Very preterm (28 to <32 weeks)	13	13.4
Moderate preterm (32 to <33 weeks)	13	13.4
Late preterm (34 to <37 weeks)	54	55.6
Prematurity without known weeks	4	4.2

Table 3: Prematurity in weeks	
Aorta	**N**
Coarctation of the aorta	48
Coarctation of the aorta - heterotaxis	1
Isthmic hypoplasia	2
Interruption of the aortic arch	9
Double aortic arch	1
Aorta tortuosa	1
Hypoplasia of the aortic arch	2
shone syndrome	1
Kommerell diverticulum	1
Atrium and Large veins	**N**
Partial anomalous drainage of pulmonary veins	1
Total anomalous pulmonary vein drainage	4
Cor triatriatum	1
Atrioventricular Connection	**N**
Transposition of the great arteries	32
Corrected transposition of the great arteries	1

Dysplasia of the tricuspid valve	1
Atresia of the tricuspid valve	10
Mitral valve stenosis	2
Dysplasia of the mitral valve	1
Ebstein's disease	2
Ventricular artery connection	**N**
Truncus arteriosus	13
Double outflow tract of the right ventricle	**N**
Double outflow tract of the right ventricle	15
Double outflow tract of the right ventricle - heterotaxis	4
Left heart hypoplasia syndrome	**N**
Left heart hypoplasia syndrome	32
Tetralogy of Fallot and variants	**N**
Pulmonary atresia with interventricular communication	21
Tetralogy of Fallot	53
Atrioventricular septum	**N**
Total atrioventricular septal defect	53
Total atrioventricular septal defect - heterotaxis	4
Partial atrioventricular septal defect	7
Single ventricle	**N**
Single ventricle	9
single ventricle - heterotaxis	2
Left ventricular outflow tract	**N**
Subvalvular aortic stenosis	2
Aortic stenosis	3
Right ventricular outflow tract	**N**
Pulmonary stenosis	10
Pulmonary valve stenosis - heterotaxis	1
Pulmonary valve stenosis	1
Pulmonary atresia + intact interventricular septum	17
Septal defect	**N**
Interventricular communication	50
Interventricular communication - heterotaxis	1
Interatrial communication	40
Persistence of the ductus arteriosus	**N**
Persistence of the ductus arteriosus	33
Coronary arteries	**N**
Anomalous origin of the left coronary artery	3
Arrhythmias	**N**
Total atrioventricular block	3
Acquired	**N**
Hypertrophic cardiomyopathy	1
Pericardial effusion	1
Pericarditis	1
Intracardiac thrombus	1

Table 4: Age of the patient at the invasive intervention

Age group	N	%
Neonate (0 to 28 days)	138	31
Infant (29 days to 12 months)	204	45.8
Preschool (1 to 6 years)	77	17.3
Elementary school (7 years to 10 years)	11	2.5
Preadolescent (10 to 13 years)	4	0.9
Adolescent (13 to 18 years)	3	0.7
Adult	8	1.8

Discussion

The global prevalence of CHDs shows an increase of 10% every 5 years, which is due to the expansion of the use of echocardiography and the improvement of technologies, according to a meta-analysis on the epidemiological situation of congenital heart diseases [8,9].

In the study hospital, 502 patients were referred by the cardiopaediatrics group, most often coming to us for elective surgery, but many also were transferred from other hospitals or were born in the obstetric centre of our hospital. In Brazil, a study on the local epidemiological situation estimated 25,757 new cases of CHD/year, without considering the large amount of underreporting, serving as a warning about the growing number of new cases and the need to direct investments to the care of this local population [10].

We saw no predominance of one sex over another, in line with several other studies; however, there are studies that report significant sex-related differences in specific heart diseases [11-13].

Some 19.3% of our sample was born prematurely. This high number is in line with the finding that 16% of children born with cardiovascular malformations are premature, according to a large English study [14]. In addition, an important issue in this population is the high mortality rate [14]. Premature infants who have CHD have a worse performance than those born at term because they are more vulnerable, and this condition remains a clinical and surgical challenge [15].

The most frequent CHDs found in the present study were acyanogenic, which is in agreement with the epidemiological data from national publications [16]. In international data, there is also a predominance of acyanogenic heart diseases [17]. The prevalence of mild injuries is due to the greater use of echocardiography worldwide, which has increased the diagnosis rate. In addition, abortion in complex heart diseases is an option in many countries, which also influences the reduction of the incidence of these specific CHDs .

Total atrioventricular septal defect (10.56%), interventricular communication (9.96%), and aortic coarctation (9.56%) are the most prevalent acyanogenic CHDs. Of the congenital cyanogenic heart diseases, the most commonly found were tetralogy of Fallot (10.56%), transposition of the great arteries (6.38), and left heart hypoplasia syndrome (5.98%). Other studies have found that tetralogy of Fallot was the most frequent cyanogenic CHD. Total atrioventricular septal defect was the most common acyanogenic CHD, but it is not the most prevalent in the world literature, which can be explained by the significant number of patients with Down syndrome (n=91) in our study population.

In CHDs, the anatomy determines the cardiac physiology, and patients with univentricular physiology have a more complex clinical management and undergo numerous invasive interventions throughout life [18]. In this study, the classification of patients according to physiology showed a prevalence of 81.1% with biventricular physiology, whereas univentricular patients represented 18.9% of the sample. This significant proportion presents a challenge for the entire multidisciplinary intensive care team. The physiology of a single ventricle may result from a series of anatomical lesions that are associated with a variety of physiological manifestations, requiring great expertise from the professionals involved in conducting treatment and surgical correction [18].

Syndromes were diagnosed in 31.2% of our sample, which is in agreement with the range found in the literature of 25 to 30% [19,20]. Chromosomal diseases stood out, especially Down syndrome, representing 58% of the syndromes found. The high prevalence of chromosomal diseases corroborates the recommendation of the need to perform chromosomal studies in new-borns with multiple anomalies.

A total of 34.7% of our sample had associated comorbidities. The abnormal circulatory physiology in children with CHD influences the development and functioning of other systems, with a significant impact on the clinical management and outcome of cases [21]. Seizures, protein-calorie malnutrition, and encephalopathies were the main comorbidities found in this study.

Syndromes, extracardiac anomalies, prematurity, low birth weight, and other associated comorbidities aggravate the challenges

associated with the treatment of CHD and are significant risk factors that affect the survival of these patients [22]. The vast majority of patients included in the study (88.65%) underwent invasive intervention. In total, 530 procedures were performed, of which 77% were surgeries, 18.5% were haemodynamic procedures, and 4.5% were hybrid procedures. A risk score called RACHS-1 was used to categorize the surgeries. Despite some shortcomings, such as the low individual predictive power and the inability to classify all cardiac procedures, RACHS-1 is a well-used and widespread tool that categorizes CHD according to the expected mortality in six categories. Each of them is used as a predictor of postoperative mortality. In the present study, there was a predominance of category 3 (38.4%), followed by category 2 (28.6%), category 1 (18.2%), category 4 (9.6%), and category 6 (5, 2%). According to a Brazilian study of mortality in CHD using RACHS-1 in a single centre, the mortality rate of category 3, the main one in this study, was 8.5%, though it ranged from 10.4% to 60% in international reports [23]. On top of the high risk scores for the surgeries themselves, surgical complexity and treatment outcome are also strongly influenced by comorbidities and associated syndromes, both present in one-third of our sample [24]. This information reflects the need for highly specialized care, presenting a challenge for clinical and surgical teams.

This study's main limitation was its retrospective nature. Because of this and the changes in the hospital evolution system, the data were limited.

Conclusion

This study comprehensively illustrates the epidemiological characteristics of a private hospital that also does philanthropic care. The complexity of the profile of the CHDs revealed that the integrated approach calling on professionals of various specialties for the treatment of these patients is essential for a successful outcome. Efforts must be made so that the team responsible for the care of complex CHD patients is highly specialized, as they have numerous associated comorbidities and syndromes and need specific, high-level care. Organizing an intensive care unit focussed on paediatric cardiac care in hospitals that have a demand for it is a strategy to achieve high specialization of care and continuous improvements focussed on this population. The issues raised are important because they serve as the basis of national comparative studies, which are scarce in the literature, and because they explain the clinical profile of CHD treated in the private sector, which is erroneously thought to absorb less complex cases. Thus, the importance of each service knowing the particularities of the CHD population served is of paramount importance to perform investment management for quality care.

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Availability of Data and Materials

All original (deidentified) data and materials are available upon request from the corresponding author.

Ethics Approval and Consent to Participate

This study was carried out in accordance with the principles of the Declaration of Helsinki. Approval was granted by the Ethics Committee of the Municipal Secretary of Health of São Paulo. Ethics approval number: 25442019.3.0000.0086.

Competing Interests

The authors declare that they have no competing interests.

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