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ORIGINAL ARTICLE

Clinical, epidemiological and therapeutic characterization of children with congenital heart disease

Caracterización clínica, epidemiológica y terapéutica de niños con cardiopatía congénita

Yoandro Rosabal-García[1]✉, Lorchen Torres-Quiñones[2], Elisa Juy-Aguirre[3], Pablo Antonio Hernández-Dinza [4], Magyoris Malo de Molina-Sariol[5]

[1]University of Medical Sciences of Santiago de Cuba. Saturnino Lora Provincial Hospital. Santiago de Cuba, Cuba.
[2]University of Medical Sciences of Santiago de Cuba. Santiago de Cuba, Cuba.
[3]University of Medical Sciences of Santiago de Cuba. Juan de la Cruz Martínez Maceira Pediatric Teaching Hospital. Santiago de Cuba, Cuba.
4University of Medical Sciences. Antonio María Béguez César South Pediatric Teaching Hospital. Intensive Care Service. Santiago de Cuba.
[5]University of Medical Sciences of Santiago de Cuba. Carlos Juan Finlay Polyclinic. Santiago de Cuba. Cuba.

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ABSTRACT

Introduction: pediatric cardiology has evolved considerably in recent years on the basis of new embryological, pathological and physiological knowledge. This progress has not been unrelated to the development of new technologies. It is currently estimated that between 650,000 and 1,300,000 adults have cardiac malformations.
Objective: to characterize clinically, epidemiologically and therapeutically the patients with congenital heart disease admitted to the Hospital Infantil Norte from January 2017 to December 2019. Clinical, epidemiological and therapeutic variables.
Methods: a descriptive and cross-sectional study was carried out with the aim of characterizing patients admitted to the Hospital Infantil Norte Docente "Juan de la Cruz Martínez Maceira" with the diagnosis of Congenital Heart Disease.

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Results: it was evidenced that 71 % corresponded to the male sex, in patients from one year to five years old were the ones with the highest number (58 %) in this group the male sex predominated with 39 %, the symptom that had the highest presence was the cardiac murmur with 53 %, this in turn in the male sex with 20 %. Regarding the diagnosis, 37% of the patients with atrial septal defect were male, 20 % of whom were male.
Conclusions: patients with congenital heart disease constitute a group with specificities such as age, time of diagnosis and therapy used, which places them as an entity of relatively frequent observation in the pediatric population.

Keywords: Heart Defects, Congenital; Complications.

RESUMEN

Introducción: la cardiología pediátrica ha evolucionado considerablemente en los últimos años sobre la base de nuevos conocimientos embriológicos, patológicos y fisiológicos. Este progreso no ha sido ajeno al desarrollo de nuevas tecnologías. Estimándose actualmente entre 650,000 y 1,300 000 adultos con malformaciones cardiacas.
Objetivo: caracterizar de forma clínica, epidemiológica y terapéutica de los pacientes con cardiopatías congénitas ingresados en el Hospital Infantil Norte en el período comprendido de enero 2017 a diciembre del año 2019. Variables clínicas, epidemiológicas y terapéuticas.
Métodos: se realizó un estudio descriptivo y transversal con el objetivo de caracterizar los pacientes ingresados en el Hospital Infantil Norte Docente "Juan de la Cruz Martínez Maceira" con el diagnóstico de Cardiopatías Congénitas.
Resultados: se evidenció que el 71 % correspondió al sexo masculino, en pacientes de un año a cinco años fueron los de mayor número con (58 %) en este grupo predomino el sexo masculino con 39 %, el síntoma que tuvo mayor presencia fue el soplo cardiaco con 53 %, este a su vez en el sexo masculino con un 20 %. En lo referente al diagnóstico se observó dominio de los pacientes con comunicación interauricular, con un 37 % de estos un 20 % fueron del sexo masculino.
Conclusiones: los pacientes con cardiopatías congénitas constituyen un grupo con especificidades tales como; la edad, el momento del diagnóstico y la terapéutica empleada, la cual los ubica como una entidad de observación relativamente frecuente en la población pediátrica.

Palabras claves: Cardiopatía Congénita; Complicaciones.

INTRODUCTION

Congenital heart defects (CHDs) vary widely among epidemiological reports of disease. They are the most common congenital malformation, with an incidence of 2,5 to 3 per 1,000 births, are the most common congenital defects, and account for nearly one-third of all major congenital anomalies.[1]

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Congenital heart disease (CHD) is currently a major health problem, some of which do not severely impact the patient's daily life, while others are more serious, limiting functional capacity and compromising quality of life, and therefore require a significant reduction in the risk of congenital heart disease. It has been reported that 30 % of cases of critical congenital heart disease are diagnosed within three days of birth in the United States.[2]

Its worldwide prevalence is estimated to range from 4 to 9 per 1 000 births, depending on the diagnostic capacity and acuity and reporting process of each registry, as well as genetic and environmental factors in each region.[3]

In 2018, Argentina presented an infant mortality of 8,9 per 1000 births and congenital anomalies accounted for 28 % of these deaths.[4] Mexico has a little more than two million births per year, we estimate around 20,000 children with congenital heart disease, which makes these pathologies a major health problem.[5] Congenital heart disease is the second leading cause of mortality in children under one year of age and, in the period from one to 14 years, it rose from the third to the second leading cause, displacing childhood cancer.[6]

In Cuba, about 1000 children are born each year with congenital heart disease (6,5 per thousand live births), between 20 and 40 % are considered critical or very severe heart disease. In 2018, 25 343 patients were attended at the "William Soler" Cardiocenter, of whom 3918 needed hospitalization with the diagnosis of congenital heart disease, of them 1993 adults and 1925 children, 264 under one year of age (52 newborns).[7] According to Valentín,[8] congenital cardiovascular anomalies are the second cause of mortality in patients under 1 year of age, surpassed only by infections and perinatal conditions.

From a global perspective, the fight against mortality in childhood has been remarkably effective in recent decades, but a more detailed analysis shows that the neonatal mortality rate has stagnated in the same period, without following a similar rate of decrease and, therefore, its weight is increasing among deaths in the first years of life. At the "William Soler" Cardiocenter, the results of the joint work are documented with the decrease in the number of infant mortality due to heart disease in Cuba, of more than 80%, from a rate of more than five per thousand live births at the beginning of the 80's, it was reduced to 0,4 per thousand live births in 2018.[9]

In view of the morbidity due to these causes, their impact on health and society, survival and quality of life of children, this situation motivated the research with the aim of characterizing the clinical, epidemiological and therapeutic characterization of patients with congenital heart disease admitted to the Hospital Infantil Norte.

METHODS

A descriptive and cross-sectional study was carried out with the objective of characterizing the patients admitted to the Hospital Infantil Norte Docente "Juan de la Cruz Martínez Maceira" with the diagnosis of congenital heart disease with the objective of clinically, epidemiologically and therapeutically characterizing the patients with congenital heart disease admitted to the Hospital Infantil Norte Docente "Juan de la Cruz Martínez Maceira", the following variables were studied, collected in a data collection form.

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Demographic variables
Age: age groups as follows:
- < 1 year
- 1-5 years
- 5 years

Sex: it was divided according to its two biological aspects into:
- Male
  Female

Race: it was divided according to skin color
- White
- Mestizo
- Black

Clinical variables

Timing of diagnosis:
- prenatal (diagnosis before birth)
- postnatal (diagnosis after birth)

Cardinal or presenting symptoms

- heart murmur
- dyspnea
- palpitations
- cyanosis
- inadequate weight gain
- marked physical exhaustion

Type of heart disease according to severity classification and prognosis: It was divided into:

a). Critical: complex cyanotic cardiopathies that endanger the patient's life and that require eminent interventional or surgical procedures.
b). Potentially critical: simple cardiopathies (cyanotic and/or acianotic) with hemodynamic repercussion that could have an interventional or surgical management in the medium term.
c). Non-critical: Asymptomatic patient, without hemodynamic repercussions, usually with medical treatment and clinical follow-up.

Clinical diagnosis

- Ventricular septal defect (VSD)
- Aortic Stenosis
- Atrial Septal Defect (ASD)
- Dilated Cardiomyopathy
- Persistent Ductus Arteriosus (PCA)
- Tricuspid Atresia
- Pulmonary Atresia
- Aortic Coarctation
- Double Emergency Right Ventricle
- Atrioventricular Septal Defect Atrioventricular Septal Defect (AVSD)

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- Ebstein's anomaly
- Tetralogy of Fallot
- Transposition of great vessels

Anatomical Stratification: Congenital Heart Disease
- Simple
- Complex

Therapeutic Approach
- Surgical
- Medical
- Interventional
- Surgical-Interventional (Combined)

Complications presented

- Cardiovascular (heart failure, acute cardiac arrhythmia, cardiac arrest, pericardial effusion, infective endocarditis, pulmonary thromboembolism, dysfunction of fistulas and/or intracardiac devices).
- Neurological (acute ischemic neurological damage, seizures, transient motor deficits).
- Respiratory (healthcare-associated pneumonia, pleural effusion, pneumothorax, Respiratory Distress Syndrome (RDS))
- Surgical (surgical wound infection, mediastinitis, presence of surgical residual lesions)
- Other complications (hematologic, acid-base balance disorders, hydroelectrolytic).
- No complications (there were no pathologies that aggravated the patient's prognosis and evolutionary status).

Information Processing

Once the information was obtained, a database was created using the SPSS system version 22.0, with which summary measures such as number and percentage were made.

RESULTS

When analyzing the patients admitted for congenital heart disease according to demographic variables and time of diagnosis (Table 1), it was found that the predominant age group was between one and five years with 101 patients, 57 % of whom 39 % were male, and the majority race was mestizo with 59 %, 16 % of whom were female. Prenatal diagnosis was performed in 58 % of the patients.

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Table 1. List of patients according to demographic variables (age, race and sex) admitted to the Hospital Infantil Norte Docente "Juan de la Cruz Martínez Maceira".

Demographic Variables	Nº	%*
Age		
< 1 year	10	6
1-5 years	101	57
> de 5 years	66	37
Sex		
Male	125	71
Female	52	29
Race		
White	52	29
Mestizo	105	59
Black	20	11
Time of diagnosis		
Prenatal	102	58

Source: Medical records. * % of total columns

When evaluating the patients with congenital heart disease according to main clinical variables and sex (Table 2), the symptom with the greatest presence was the presence of cardiac murmur with 53 %, this in turn in the male sex with 20 %, in turn dyspnea, inadequate weight gain, physical exhaustion had similar percentages in both sexes. As for the severity of the cases, the female sex predominated the non-critical with 46 %, in the male sex they had similar percentages among the critical, moderately critical and non-critical with 36 %, 34 %, 30 % respectively.

Table 2. Patients with congenital heart disease according to clinical variables and sex.

CLINICAL VARIABLES		Sex			
		Male		Female	
		Nº	%*	Nº	%*
Symptoms	heart murmur	25	20	17	33
	Dyspnea	16	13	8	15
	Palpitations	11	9	6	12
	Cyanosis	40	32	9	17
	inadequate gain	14	11	5	10
	Exhaustion	19	15	7	13
Severity	Critical	45	36	11	21
	Non-critical	37	30	24	46
	Moderately Critical	43	34	17	33

Source: Medical records. % of total columns

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Regarding clinical diagnoses (Table 3). Of the patients with a diagnosis of Atrial Septal Defect (ASD), 37 % were male (20 %), patients with ventricular septal defect were predominantly female (23 %), although pathologies such as dilated cardiomyopathy, persistent ductus arteriosus (PCA), atrioventricular septal defects (AVSD) were observed (2 3%, 21 %, 18 % respectively), the latter mostly in the female sex.

Table 3. Patients with congenital heart disease according to clinical diagnosis and sex.

Clinical diagnosis	Sex					
	Male		Female		Total	
	Nº	%	Nº	%	Nº	%
CIV	15	12	12	23	27	35
Clinical diagnosis	7	6	2	4	9	9
CIA	25	20	9	17	34	37
Dilated Cardiomyopathy	12	10	7	13	19	23
PCA	9	7	7	13	16	21
Tricuspid atresia	4	3	2	4	6	7
Pulmonary atresia	2	2			2	2
Aortic Coarctation	10	8	3	6	13	14
Double RV Emergency	4	3			4	3
DSAVC	13	10	4	8	17	18
Ebstein's anomaly	3	2	1	2	4	4
Tetralogy of Fallot	6	5			6	5
Transposition of great vessels	7	6	2	4	9	9

Source: Medical records. % of total patients

Table 4 shows that 50 patients underwent surgical therapy, 70 % of which were in heart diseases with complex anatomical stratification. 100 % of the interventional procedures were in heart diseases with simple anatomical stratification, while medical therapy was the most used in 97 patients, mostly in patients with simple anatomical stratification (98 %).

Table 4. Patients with congenital heart disease according to therapy used and anatomic stratification.

Therapeutics used	Anatomical Stratification			
	Complex		Simple	
	Nº	%*	Nº	%*
Surgical	35	70	15	30
Medical	2	2,06	95	97,9
Interventional	0	0	17	100
Surgical-interventional	10	76,92	3	23,08
Total	47	26,55	130	76,45

Sources: medical records; * % to row total

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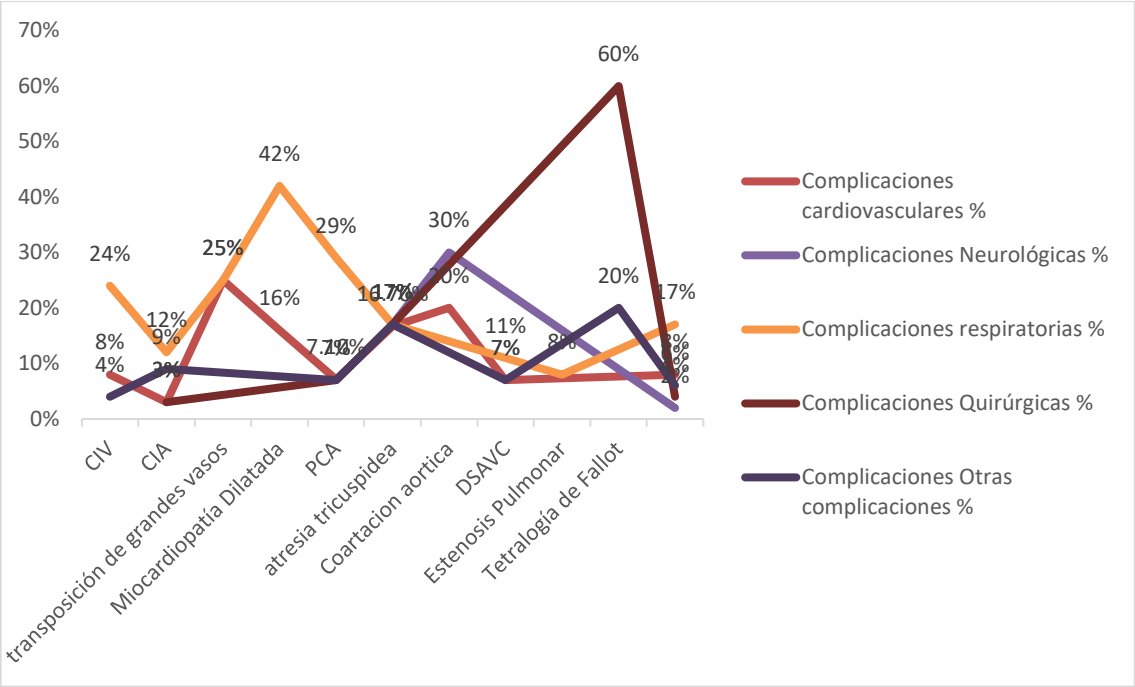
Graph 1 shows the predominance of respiratory complications with 30 patients representing a total of 17% and cardiovascular complications with 14 patients for 8 %, in turn dilated cardiomyopathy had higher percentage representation in both groups, other entities such as VSD and PCA had percentage figures of 29 %, 24 % respectively. In patients with tetralogy of Fallot, there were three patients with surgical complications, representing 60 % of the total patients with this entity. It should be added that 63 % of the patients did not present complications.



Sources: Medical records.

Graph 1 Patients with congenital heart disease according to complications and diagnosis.

DISCUSSION

Sometimes the finding of the malformation can occur late, because the symptoms do not appear from birth and the cardiovascular physical examination is normal from the first weeks, and even in later months; In a study by Gonzalez et al,[10] they evaluated patients according to age and sex. Regarding age it was observed that most of the patients were betweenone and nine years old, boys represented higher numbers than girls. At that point Machado et al 11; refer that 58 % of patients with congenital heart disease belonged to the male sex, and an age range of 23,8 months, according to Méndez et al ,[12] the predominant gender was female (53,5 %) in their study, with a prevalence of one to 11 months of life (40,6 %).

Inés Reyes-Roig and cols,[13] in a study on congenital heart disease and genetic syndrome refer to the types of heart disease related to sex, where it is seen that heart disease with short circuit predominated with 33 patients and, of these, interatrial and interventricular communications with 11 patients each, for 24 %, and among them two correspond (30,5 %) to the female sex. In a Colombian study it was found that the female sex predominated over the male population.[14]

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In a publication presented by Aguilera Sánchez,[15] a female/male ratio of 1,17 % prevailed. It also showed the time of postnatal diagnosis with 60,78 %. In this study, the most frequent congenital heart diseases were atrial septal defects, followed by ventricular septal defects.

The clinical findings appear later, depending on the severity of the defect. Studies on congenital heart disease showed that the predominant clinical presentation was the murmur present in a high number of patients.[10]

Machado et al,[11] referring to the presence of complex congenital cardiopathies showed in their work that 61,9 % of children were carriers of complex cardiopathies, in eight of them the diagnosis was prenatal. Six of the 10 children with other malformations or chromosomopathies had complex congenital heart disease.

Alonso et al,[16] reflects in a study the most frequent congenital cardiopathies were the acyanotic ones, which agrees with other works such as the one presented by Groisman et al,[17] Referring to a study carried out in Argentina on congenital cardiopathies, a total of 1814 (71,84 %) isolated or simple cases were determined, 519 (20,55 %) with multiple or complex anomalies. Tetralogy of Fallot was the most frequent specific criticism. Double emergence of the right ventricle was the most frequent associated with syndromes or multiple anomalies.

ravieso et al, [18] carried out a study on genetic characterization in the province of Pinar del Río, the most frequent defects were septal anomalies, represented mainly by ventricular septal defect and complete atrioventricular canal; however, ventricular septal defect is not in the list of prioritized international surveillance defects, since it is not included among the main defects to be monitored. It is reasonable, considering that small isolated VSD usually show a very favorable postnatal evolution without the need for surgical treatment. Even in larger defects that do not close spontaneously, very high survival rates with good quality of life are reported despite surgical treatment.[19]

Gonzalez et al,[10] evidenced that surgery occupied the first place in the order of applied therapy, followed in order of frequency by spontaneous resolution and resolution by catheterization. Another smaller percentage of patients continue with the cardiac defect. In another point of analysis, Mariño et al,[20] report that percutaneous closure of simple heart defects such as ASD and PCA closure is the most frequent interventional procedure performed. This coincides with the results of the present investigation. This places the results of interventional procedures in a superior role in the management of congenital heart disease at present.

CONCLUSIONS

Patients with congenital heart disease constitute a group with specificities such as age, time of diagnosis and therapy used, which makes them a relatively frequent entity to be observed in the pediatric population.

Conflict of interest
The authors declare that they have no conflicts of interest.

Authors' contributions
YRG: Conceptualization; formal analysis; research; methodology; project management; resources; software; supervision; validation; visualization; writing the original draft, (reviewing and editing).
EJA: Conceptualization; data curation; formal analysis; research.
PAHD: Formal analysis; research; methodology; statistical analysis; critical review.

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MS MM: Literature search; tabulation; statistical analysis; writing; participated in collective discussion of the final version.
LTQ: Search for bibliographies; tabulation; participated in the collective discussion of the final version.

Additional material
Additional material to this article can be consulted in its electronic version available at: www.revcmpinar.sld.cu/index.php/publicaciones/rt/suppFiles/5721

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