PEER REVIEW, Vol. 6, Nº 1, 2024
DOI: 10.53660/PRW-1731-3330
ISSN: 1541-1389

Congenital Heart Disease: Clinical Characterization in Newborns from a Referral Hospital, Manaus, Amazonas, Brazil

Cardiopatias Congênitas: Caracterização Clínica em Recém-Nascidos de um Hospital de Referência, Manaus, Amazonas, Brasil

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Recebido: 10/12/2023 | Aceito: 12/01/2024 | Publicado: 18/01/2024

ABSTRACT

In Brazil, the impact of congenital heart disease (CHD) remains obscured by limited data, especially in the northern region. For this reason, our study seeks to uncover the prevalence and clinical variations of CHD in Manaus, Amazonas. This study followed a cross-sectional model performed in 110 newborns with CHD diagnosed between March 2022 to April 2023. Complex CHD was present in about 80% of the newborns, which increased the risk of requiring multiple surgical interventions. We found and characterized 19 CHD phenotypes, with a higher prevalence of complex CHD cases. We documented 12 fatalities, mostly attributed to septic and cardiogenic shock related to complex CHD with high levels of urea, creatinine, AST, ALT, CRP, GGT, NT-proBNP and acidosis ($p<0.001$). Our findings suggest a close relationship between CHD and a systemic impact with chronic/acute kidney failure and hepatopathies. Inadequate data from the information system on live births (SINASC) results in underreporting and increased CHD mortality rates, emphasizing the urgency for new strategies to enhance case identification and improve early diagnosis of critical CHD with important systemic repercussions.

Keywords: Heart disease; Congenital disease; Cyanosis; Newborn; Manaus.

RESUMO

No Brasil, o impacto das doenças cardíacas congênitas (DCC) permanece ofuscada por dados limitados na literatura, especialmente na região Norte do Brasil. Um estudo transversal foi realizado em 110 recém-nascidos diagnosticados com DCC entre março/2022 a abril/2022. A maioria dos casos de DCC (80%) foram do tipo complexa, ocasionando a necessidade de múltiplas intervenções cirúrgicas. Encontramos e caracterizamos 19 fenótipos de DCC, com maior prevalência de casos de DCC complexos. Dureante o período de estudo, foram documentadas 12 mortes, a maioria atribuídas a choque séptico e cardiogênico relacionado à DCC complexas com níveis elevados de uréia, creatinina, AST, ALT, PCR, GGT, NT-proBNP e acidose ($p<0,001$). Nossos achados sugerem uma estreita relação entre doença coronariana e impacto sistêmico com insuficiência renal crônica/aguda e hepatopatias. Dados inadequados do SINASC resultam em subnotificação e aumento das taxas de mortalidade por doença coronariana, enfatizando a urgência de novas estratégias para melhorar a identificação de casos e melhorar o diagnóstico precoce de DCC com importante repercussão sistêmica.

Palavras-chave: Cardiopatia; Doença congênita; Cianose; Recém-nascido; Manaus.

INTRODUCTION

Characterized as alterations that impact the heart and circulatory system, congenital heart disease (CHD) varies in its complexity and physiological repercussions, and makes up about one-third of all congenital anomalies, affecting around 1% of all live births (KHATAMI et al., 2018). In Brazil, CHD is the second leading cause of death in the first year of life, following infectious diseases, and the primary cause of deaths in children up to the age of five (SALIM et al., 2020; SOARES, 2020). It is estimated that up to 80% of individuals with CHD require surgical intervention, half of them, within the first year of life (SOARES, 2020).

Clinically, CHD can manifest as either an isolated malformation (simple) or in multiple forms (complex), involving two or more concurrent malformations in the patient. In the latter situation, surgical correction is challenging and often requires sequential surgeries (DOLGNER et al., 2020). Syndromic patients, such as those with DiGeorge syndrome, Noonan syndrome, Down syndrome, and others, carry an elevated risk for the occurrence of complex CHD (PIERPONT et al., 2018; SHABANA et al., 2020). CHD may also be classified into two distinct groups: cyanotic and acyanotic (HOFFMAN & KAPLAN, 2002). The most frequent acyanotic malformations are ventricular septal defect (VSD), atrial septal defect (ASD), atrioventricular septal defect (AVSD), patent ductus arteriosus (PDA), and aortic coarctation (AoC). Cyanotic conditions are more severe due to reduced arterial hemoglobin, and commonly include tetralogy of Fallot (TOF) and transposition of the great arteries (TGA) (VAN DER LINDE et al., 2011; LIU et al., 2019).

Over the past few decades, progress in diagnostic tools and surgical procedures has led to a reduction in mortality rates associated with CHD, enabling up to 85% of those with CHD to reach adulthood (MARINO et al., 2012; KRISHNA & KUMAR, 2020). Despite advances in early detection methods, CHD in Brazil is a significant public health concern and it is estimated that up to 50% of newborns may remain undiagnosed (BRASIL, 2017). Early diagnosis is essential to prevent shock, acidosis, cardiac arrest or neurological complications prior to treatment (MARINO et al., 2012; DESAI et al., 2019). Additionally, underserved remote areas often struggle to provide proper treatment due to a lack of qualified professionals and limited access to healthcare (MATTOS et al., 2015).

The importance of early diagnosis of CHD is underscored by its social impacts, effects on quality of life, and the substantial associated long-term healthcare costs (BERTOTLETTI et al., 2014; KRISHNA & KUMAR, 2020).

The true extent of CHD among the Brazilian population, particularly in the northern region, remains insufficiently understood due to incomplete and non-comprehensive data in the the information system on live births (Sistema de Informações Sobre Nascidos Vivos - SINASC). With this in mind, our objective was to outline and delineate the occurrence and diverse clinical manifestations of CHD cases diagnosed at Fundação Hospital do Coração Francisca Mendes (FHCFM).

MATERIALS AND METHODS

This study followed a cross-sectional design, in partnership to Fundação Hospital do Coração Francisca Mendes (FHCFM) from March 2022 to April 2023. The FHCFM is a cardiology referral center located in Manaus, Amazonas, which provides comprehensive services to the state capital Manaus, as well as to remote areas and other states. The study covered all patients under one year of age who were seen for emergency room consultations, hospitalizations and cardiac surgery at FHCFM.

Participation in the study required the legal guardian's signature on the informed consent form. Clinical and demographic information was collected through interviews and medical records. This data was then organized into individual patient profiles, which were compiled, and structured on an Excel spreadsheet. Subsequently, after being categorized according to variable type, the data was subjected to statistical analysis using SPSS (version 23) software. Descriptive statistics were applied to describe the main clinical and demographic data. Laboratory data were analyzed and those with p-values <0.05 were considered statistically significant.

RESULTS

The study sample comprised 110 pediatric patients under 1 year of age, who had been referred to FHCFM for CHD treatment. Of the total, 57 (51.8%) were male and 53 (48.2%) were female, with 8 (16%) and 7 (15.9%) being diagnosed with Down syndrome,

respectively. Nine (8.2%) mothers reported the existence of heart disease in the family, eight (7.2%) had diabetes and eight (7.2%) had high blood pressure. Twelve (10.9%) children died during the study, with no deaths associated with mothers with diabetes and/or high blood pressure.

The Francisca Mendes Hospital is the largest referral hospital for heart disease in the state of Amazonas and is a referral center not only for Manaus, but also for the other cities in the state of Amazonas and the northern region of Brazil. The majority of newborns resided in Manaus (70,1%), with 23 (20.9%) coming from other cities in the interior of the state of Amazonas. The full list of cities and the corresponding number of patients referred to FHCFM for CHD treatment can be found in **Table 1.**

Table 1. Patients treated at Fundação Hospital do Coração Francisca Mendes according to their city of residence in the state of Amazonas (2021-2022).

N	**Cities**	**n**	**%**
1	Manaus	87	79.1
2	Tefé	3	2.7
3	Coari	2	1.8
4	Maués	2	1.8
5	Parintins	2	1.8
6	Tabatinga	2	1.8
7	Alvarães	1	0.9
8	Autazes	1	0.9
9	Carauari	1	0.9
10	Humaita	1	0.9
11	Iranduba	1	0.9
12	Labrea	1	0.9
13	Jutaí	1	0.9
14	Manacapuru	1	0.9
15	Manicoré	1	0.9
16	Rio Preto da Eva	1	0.9
17	São Gabriel da Cachoeira	1	0.9
18	São Paulo de Olivença	1	0.9

	Total	110	100

In this study, 99 (90.0%) patients were confirmed as having CHD, resulting in a total of 80 (81.6%) hospitalizations due to the need for surgical procedures. Additionally, 11 (10.0%) patients were referred to FHCFM with cardiovascular issues and were still under investigation of the diagnosis at the end of our study period. It was found that 9 (8.2%) cases involved premature birth and had a low birth weight (1.8±0.53 kg), with no significant difference between genders. The mean duration of the hospitalization of the patients was 28.29 ± 26.6 days. Some cases required multiple and sequential surgeries that exceeded 90 days of hospitalization, with 80% of these cases needing more 30 days of follow-up. The remaining analyzed variables are described according to gender in **Table 2**.

Table 2. Demographic and clinical data of pediatric patients with congenital heart disease and under 1 year of age and treated at Fundação Hospital do Coração Francisca Mendes between 2021 and 2022.

Demographic and clinical data	**Male (%)**	**Female %)**	**Total (%)**
	57 (51.8)	53 (48.2)	110
Age (months)	4.35±3.61	5.18±3.31	4.75±3.48
Weight (kg)	2.7±0.69	2.8±0.74	2.7±0.71
Low weight at birth (<2,500g)	4 (7.0)	5 (9.4)	9 (8.2)
Multiple heart surgeries	47 (82.5)	44 (83.0)	91 (82.7)
Complex cardiac disease	42 (78.7)	38 (86.0)	80 (72.7)
Received blood transfusions	50 (87.7)	44 (83.0)	94 (85.5)
Red Blood Cells (Concentrate)	25 (43.9)	23 (43.4)	48 (43.6)
Platelet (Concentrate)	19 (33.3)	22 (41.5)	41 (37.3)
Fresh Frozen Plasma	16 (28.1)	21 (39.6)	37 (36.6)
Hospitalization (days)	27±21.27	29±31.7	28.29±26.6
Blood group			
A +/-	18 (31.6)	16 (30.2)	34 (30.9)
B+/-	2 (3.5)	4 (7.5)	6 (5.5)

AB+/-	1 (1.8)	2 (3.8)	3 (2.7)
O+/-	36 (63.1)	31 (58.5)	67 (60.9)

The patients' laboratory hematological and biochemical data are shown in **Table 3**, with no significant differences when compared between genders.

Table 3. Laboratory data (according to gender) of pediatric patients with congenital heart disease under 1 year of age and treated at Fundação Hospital do Coração Francisca Mendes between 2021 and 2022.

Laboratory data	Male Mean ± SD	Female Mean ± SD	p-value
Hemolysis			
RBC, x 10^6/mm^3	4.53±0.93	4.35±0.81	.348
Hemoglobin, g/dL	12.63±2.71	12.16±2.23	.392
Hematocrit, %	38.17±7.96	36.81±6.57	.410
Mean cell volume, fL	84.58±9.82	85.61±9.16	.643
Mean cell hemoglobin, pg	28.11±.47	28.12±3.17	.994
RDW, %	17.11±3.31	16.73±2.43	.666
Leukocytes			
WBC, x 10^6/L	12.19±6.84	10.54±3.64	.181
Neutrophils x 10^6/L	6.52±3.97	5.13±2.16	.055
Lymphocytes x 10^6/L	4.47±2.19	4.42±1+99	.916
Monocytes x 10^6/L	0.74±0.62	0.57±0.32	.155
Eosinophils x 10^6/L	0.41±0.38	0.39±0.31	.863
Basophils x 10^6/L	0.06±0.04	0.03±0.02	.281
Platelets			
Platelet count, x 10^9/L	313.18±128.52	312.34±98.78	.975
Hemolysis plus Hepatic			
Aspartate aminotransferase, U/L	57.34±45.38	71.42±54.68	.411
Alanine aminotransferase, U/L	40.13±35.63	59.81±43.41	
Gamma glutamyl transferase, U/L	118.57±108.68	91.73±85.21	
Lactate dehydrogenase, U/L	19.77±13.51	19.65±13.71	.980
Kidney			
Blood urea nitrogen, mg/dL	32.65±21.70	32.69±27.87	.996
Creatinine, mg/dL	0.54±0.22	0.62±0.53	.354
Glucose			

Glucose, mg/dL	95.59±19.63	110.62±38.74	.126

WBC: White blood cell count RBC: Red blood cell count

RDW: Red blood cell distribution width

Over the course of the study, a total of nineteen distinct types of CHD were identified. Among these, patent ductus arteriosus (PDA), ventricular septal defect (VSD) and atrial septal defect (ASD) represented over 60% of the cases, whether occurring in isolation or in combination with other malformations. Of all the diagnosed CHDs, the majority (73.6%) was of the acyanotic type, with tetralogy of Fallot (TOF) being most frequent cyanotic type, accounting for 14.5% of cases. Isolated cases (<1%) of rare and severe conditions such as Anomalous left coronary artery from the pulmonary artery; aortic coarctation; interrupted aortic arch; systemic to pulmonary anastomosis and truncus arteriosus **(Table 4)**.

Table 4. Prevalence of acyanotic and cyanotic heart disease (according to gender) of pediatric patients with congenital heart disease under 1 year of age and treated at Fundação Hospital do Coração Francisca Mendes between 2021 and 2022.

Heart Disease	Type	Male n=57 (%)	Female n=53 (%)	Total n=110 (%)
Anomalous Left coronary artery from the pulmonary artery	Cyanotic	--	1 (1.9)	1 (0.9)
Aortic coarctation	Acyanotic	1 (1.8)	--	1 (0.9)
Aortic stenosis	Acyanotic	1 (1.8)	2 (3.8)	3 (2.7)
Interrupted aortic arch	Cyanotic	1 (1.8)	--	1 (0.9)
Atrial septal defect	Acyanotic	21 (36.8)	25 (47.2)	46 (41.2)
Atrioventricular septal defect	Acyanotic	6 (10.5)	5 (9.4)	11 (10.0)
Double outlet right ventricle	Cyanotic	2 (3.5)	--	2 (1.8)
Congenital mitral stenosis	Cyanotic	1 (1.8)	1 (1.9)	2 (1.8)
Patent ductus arteriosus	Acyanotic	32 (56.1)	26 (49.1)	58 (52.7)
Pulmonary atresia	Cyanotic	4 (7.0)	1 (1.9)	5 (4.5)
Pulmonary stenosis	Acyanotic	8 (14.0)	6 (11.3)	14 (12.7)
Pulmonary valve atresia	Cyanotic	2 (3.5)	2 (3.8)	4 (3.6)

Right aortic arch anomalies	Cyanotic	--	2 (3.8)	2 (1.8)
Tetralogy of Fallot	Cyanotic	8 (14.3.0)	8 (15.1)	16 (14.5)
Systemic to pulmonary anastomosis	Cyanotic	1 (1.8)	--	1 (0.9)
Transposition of the great arteries	Cyanotic	4 (7.0)	1 (1.9)	5 (4.5)
Tricuspid atresia	Cyanotic	1 (1.8)	3 (5.7)	4 (3.6)
Truncus arteriosus	Cyanotic	--	1 (1.9)	1 (0.9)
Ventricular septal defect	Acyanotic	23 (40.4)	25 (47.2)	48 (43.6)

During this study, we reported 12 cardiac-related deaths involving five males and seven females, with age in weeks of a minimum of 4 and a maximum of 44. Among these, half were attributed to septic shock and cardiogenic shock. It is noteworthy that, barring three patients, all the others presented complex congenital heart conditions (**Table 5**).

Significantly elevated levels of renal and hepatic serum markers among pediatric patients who died and those with surgical success are shown in **Figure 1**, highlighting the average GGT values which were 8 times higher in those who died (1,543±259.1 vs 190.1±15.58). Other average values were also significantly high, such as N-terminal prohormone of brain natriuretic peptide - NT-proBNP (24,875.99±1,429,60 vs 5,546.51±3,338.25) and pCO_2 (47.04±3.46 vs 38.89 ± 0.76). However, blood pH values were significantly lower in those who died (7.29±0.06 vs 7.41±0.01).

Table 5. Description of the cause of death of pediatric patients under 1 year of age with congenital heart disease in this study (2021-2022).

Patient	**Age in weeks**	**Days hospitalized**	**Medical reason for surgery**	**Cause of death**
1 / F	38	12	ASD; VSD, CMS, AoC	Congenital mitral insufficiency/aortic hypoplasia
2 / M	4	8	ASD, PDA	Not specified
3 / F	20	8	ASD, VSD	Septic shock; left ventricular insufficiency
4 / F	12	17	ALCAPA	Not specified
5 / F	8	2	CVSD, TA	Not specified

6 / M	8	28	ASD, PDA, SPA	Cardiogenic shock
7 / M	4	21	ASD, VSD, PDA, TA	Septic shock; acute renal failure
8 / F	24	10	TOF	Septic shock
9 / F	8	42	ASD, PDA	Septic shock; acute renal failure
10 / F	44	3	TOF	Cardiogenic shock
11 / M	4	4	PDA, CMS	Respiratory failure; septic shock; acute renal failure
12 / M	8	20	ASD, PDA, TGA	Cardiogenic shock; left ventricular failure

M: Male F: Female

ALCAPA: Anomalous left coronary artery from the pulmonary artery

AoC: Aortic coarctation ASD: Atrial septal defect

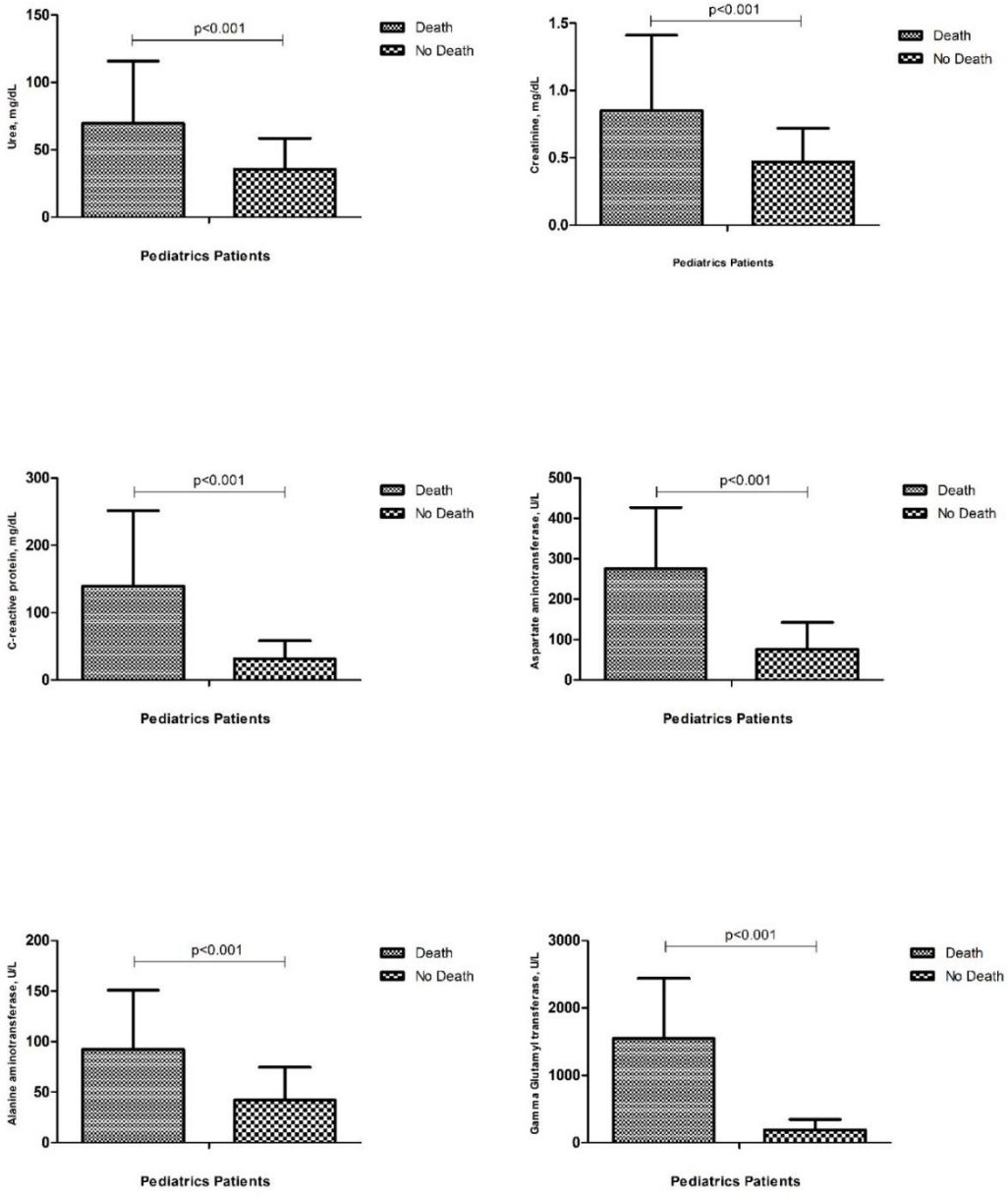
AVSD: Atrioventricular septal defect CMS: Congenital mitral stenosis

PDA: Patent ductus arteriosus SPA: Systemic-Pulmonary anastomosis

TA: Tricuspid atresia TGA: Transposition of the great arteries

TOF: Tetralogy of Fallot VSD: Ventricular septal defect

Figure 1. Description of the diferent biochemical levels of pediatric patients under 1 year of age with congenital heart disease that died vs those for whom cardiac surgery was successful.



DISCUSSION

This study tracked 110 patients and identified nineteen distinct phenotypes of CHD, including some rare and severe conditions (<1%). Patients received their diagnoses at an average age of 4.75±3.48 months, with 80 (72.7%) undergoing surgical procedures. Notably, 79.1% of the participants were from Manaus. This outcome was anticipated due to challenges in accessing diagnosis and arranging transportation for patients from rural and remote areas to receive appropriate treatment. These challenges highlighting the need for proactive efforts in these regions (MATTOS et al., 2015).

The current epidemiological indicators are not sufficiently accurate to establish the precise incidence of CHD, a concern in all of Latin America (SOLA et al., 2020). At present, the primary methods considered as the gold standard for diagnosing CHD are imaging examinations, which are still a challenge in terms of accessibility and availability for the entire population. Even though pulse oximetry has emerged as a common diagnostic screening strategy, providing universal access continues to be an obstacle (LEITE et al., 2010; SUN & LIU et al., 2015; MA & HUANG, 2018),

In the state of Pará, a neighboring state with a size comparable to that of Amazonas, a different reality is observed. There still a substantial waiting period for surgery, averaging 23 ± 18.3 months. A significant portion (63.4%) of patients come from rural areas within the state (JESUS et al., 2018). Even as of 2022, more than half still await treatment, facing considerable challenges in accessing both diagnosis and treatment, particularly in municipalities distant from the state capital Belém (ALVES et al., 2022).

Despite a higher incidence of acyanotic CHD (such as PDA, VSD, and ASD), close to 80% of the patients presented those with multiple concurrent CHD conditions. Our findings are in line with existing literature, demonstrating a comparable distribution of conditions ASD, VSD, and PDA in conjunction with other malformations, with TOF as the most prevalent among cyanotic CHD cases (VAN DER LINDE et al., 2011; LIU et al., 2019).

We hypothesize that the lower frequency of simple CHD presentation is linked to challenges in diagnosing asymptomatic presentations, which leads to underreporting (BAUMGARTNER et al., 2010; SPICER et al., 2014; MAHMOUD et al., 2019). Remote regions, distant from major urban centers, face obstacles in providing appropriate treatment, access to healthcare, and often have a shortage of qualified professionals (MATTOS et al., 2015; LOPES et al., 2018). These factors contribute to a mistaken perception of a low incidence of CHD, which results in undiagnosed cases in adults and more severe manifestations (BRASIL, 2017; SOARES, 2020).

Fortunately, a minority of patients exhibited low birth weight and prematurity. Age, weight, prematurity and the type of cardiac condition serve as risk indicators that influence mortality rates in congenital heart surgery (ARAGÃO et al., 2013). Furthermore, our study aligns with a positive correlation between the number of malformations and the duration of hospitalization, with instances of patients staying in hospital for up to four months (BELO et al., 2016).

In Campo Largo, Paraná, Brazil, a high frequency of VSD and PDA was identified (BELO et al., 2016). Similarly, in the state of Pará, frequent CHD cases included VSD (29%), PDA (18.4%), ASD (11%), and TOF (8.6%) (JESUS et al., 2018; LOPES et al., 2018). Despite our findings indicating a comparable distribution of PDA, VSD, ASD, and TOF, there is a trend towards an increase in the incidence of congenital malformations due to improved detection and diagnostic methods (LIU et al., 2019).

We believe that the varying prevalences of CHD phenotypes might be attributed to limitations in detection strategies for these malformations, accounting for regional characteristics, or even endemic variations in each area. This emphasizes the need for further studies focused on region-specific characterization of populations with CHD. Unfortunately, the SINASC data lacks sufficient information, possibly due to underreported frequencies. This deficiency results in an elevated mortality rates of CHD in neonatal population, stemming from the absesnce of strategies for ealy diagnosis and treatment (BRASIL, 2017; SALIM et al., 2020; SOARES, 2020).

Unfortunately, 12 fatalities were documented, half of which were attributed to septic shock and cardiogenic shock, and most related to complex CHD cases; consecutive surgical procedures and corrections naturally pose significant challenges and an increased risk of mortality (DOLGNER et al., 2020).

Systemic inflammation has been documented in pre-operative cyanotic CHD patients (MCPHILLIPS et al., 2019). In our study, a noteworthy proportion of our patients with cyanotic CHD who died exhibited a higher prevalence of septic shock as opposed to cardiogenic shock. This finding contrasts with the study by Chan et al. (2020), which reported a higher incidence of cardiogenic shock, particularly in relation to cyanotic episodes. Additionally, several risk factors that may contribute to the increased incidence of sepsis include prematurity, low birth weight, and hospitalization in the neonatal intensive care unit, all of which are commonly necessities observed in CHD patientes (FEIL et al., 2018).

The quantification of NT-proBNP levels in cardiogenic shock patients reveals a direct correlation with the severity of their heart disease – higher NT-proBNP levels are indicative of increased disease severity (BRISSAUD et al., 2016). In the pediatric population, NT-proBNP concentrations are notably elevated in individuals with complex congenital heart defects when compared to those with simple cardiac anomalies such as ASD, VSD, or PDA (FERNANDES et al., 2016). Our study's observations of increased NT-proBNP, AST, ALT, Creatinine, and GGT levels align with the literature, which not only highlights these substantial alterations in cases of CHD but also underscores their intricate relationship with hepatopathy (KEHL et al., 2021).

Conversely, another study has shown that elevated NT-proBNP levels are associated with adverse events in CHD patients with pulmonary arterial hypertension, though no such association was found with urea or ALT/AST levels (DENG et al., 2019). Furthermore, right/left heart dysfunction and the necessity for re-operation or re-intervention have been identified as significant risk factors for the development of hepatopathy. These findings emphasize the imperative need for further investigations aimed at reducing hepatic complications and enhancing the prognosis of complex congenital heart disease patients (KEHL et al., 2021; XIE et al., 2021).

We found elevated renal serum markers (urea and creatinine) among pediatric patients who died when compared to those for whom surgery was successful. The intricate relationship between the heart and kidney is necessary for the maintenance of cardiovascular homeostasis, and disruptions in hemodynamics within one organ can exert substantial effects on the hemodynamics of the other (TRIPOSKIADIS et al., 2022).

The physiological changes occurring in the kidneys of cyanotic CHD patients are multifaceted and have not yet been fully elucidated, though we know that they involve multiple causal pathways such as chronic hypoxia, intraglomerular hemodynamic alterations, and cardiac surgery (EL SAYEGH et al., 2022). We posit that the heightened biomarker levels are correlated with the severity of CHD, potentially culminating in acute kidney failure. Some studies suggest the potential for kidney injury following congenital heart surgery in the pediatric population, although this topic remains a subject of ongoing debate and investigation in the current literature (KHUONG et al., 2021; VAN DEN EYNDE et al., 2022). However, it is well-documented that CHD patients exhibit a heightened incidence of chronic kidney disease. Early detection and the timely implementation of corrective heart surgery for CHD may offer notable benefits for kidney function (FANG et al., 2021).

The severe acidosis observed in deceased individuals (pH: 7.29±0.06) may be attributed to the gravity of cyanotic CHD. Metabolic acidosis in CHD is typically contingent on systemic circulation and often associated with factors like obstructed ducts or pulmonary hyperperfusion (KHALIL et al., 2019). Another study reported presence of acidosis in 83% and 68%, respectively, in patients that presented cardiogenic and septic shock (CHAN et al., 2020). Acidosis is considered to be one of the most crucial predictors of adverse events, and a delayed diagnosis of CHD with concurrent acidosis also correlates with hypoxia, an increased occurrence of post-operative pulmonary hypertensive crises and respiratory failure with cyanotic episodes (DESAI et al., 2019).

This situation underscores the need for new strategies to enhance case detection, improve treatment and increase healthcare vigilance. It is crucial to invest more in assistive technology, expand referral unit capacity, decentralize care, boost specialized workforce numbers, improve detection methods and diagnostic speed and offer comprehensive professional training. Additionally, establishing new referral centers in both urban and remote areas is essential due to existing limitations in the public healthcare system’s ability to adequately support the population.

This study faced limitations due to the limited number of available medical records and the absence of information in the majority of the records that were evaluated. Many rare congenital malformations have such a low frequency of occurrence that no other comparative studies were found.

CONCLUSION

A total of 19 phenotypes of CHD were identified and characterized, with complex CHD showing a higher frequency. Our findings demonstrate a comparable distribution of the conditions ASD, VSD, and PDA in conjunction with other malformations, with TOF as the most prevalent among cyanotic CHD cases. We documented 12 fatalities, mostly attributed to septic and cardiogenic shock related to complex CHD, which presented significant differences ($p<.001$) with higher levels of urea, creatinine, AST, ALT, CRP, GGT, NT-proBNP and acidosis when compared to those with who had had successful cardiac surgery and a good prognosis. Our findings suggest and reinforce the literature findings that report a close relationship between CHD and a systemic impact, including a chronic inflammation profile, chronic/acute kidney failure and hepatopathies. There is a lack of literature data regarding the actual situation of the northern region of Brazil, the state of Amazonas, and the city of Manaus concerning CHD. Studies that report epidemiological data and contribute to the development of clinical practices are of paramount importance. These efforts can enhance healthcare delivery and help comprehend the true extent of the issue.

ACKNOWLEDGMENT

This work was funded by Fundação de Amparo à Pesquisa do Estado do Amazonas (FAPEAM) (POSGRAD Program [#002/2023]) and Coordenação de Aperfeiçoamento de Pessoal de Nível Superior (CAPES) (PDPG-CONSOLIDACAO-3-4 Program - #88887.707248/2022-00).

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