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Pattern and Diagnosis of Congenital Heart Disease in Children Admitted to Al-Sadaqa Teaching Hospital, Aden, January 2017-December 2019

Mariam T. Bin Yehya, Adnan A. Hadi, Iman A. Ba-Saddik

Abstract

Introduction: Congenital heart disease (CHD) is a lifelong problem of the heart's structure and function present at birth. The objective of this study is to determine the pattern of congenital heart diseases (CHD) among children of both sexes at Al-Sadaqa Teaching Hospital in Aden city.

Methods: This is a retrospective, hospital-based study from January 2017 to December 2019. Data were collected from the medical records at the Statistical Department of Al-Sadaqa Teaching Hospital. Children with CHD were identified and diagnosed, via history taking, clinical examination, electrocardiography and echocardiography.

Results: The total number of patients who were holding the diagnosis of heart diseases were 148, out of whom 10 children were excluded because of having acquired heart diseases. The total number of patients who were diagnosed with CHD comprised 138 with females predominating males (58% vs. 42% respectively). Their mean age was 18.8±3.0 months and the median age was 8 months. Eighty-seven percent of patients were diagnosed with acyanotic CHD. Ventricular septal defect (VSD) constituted 35.5%, atrial septal defect (ASD) 14.5%, patent ductus arteriosus (PDA) 12.3% and atriovenous canal 5.8% of the children with CHD. Tetralogy of Fallot (TOF) was the commonest cyanotic CHD diagnosed (7.2%). Children with isolated CHD constituted 82.6% while 17.4% had multiple lesions. Male preponderance was documented in VSD and PDA while females predominated in ASD. A total of 66.7% children were diagnosed with CHD at one year of age.

Conclusion: The commonest acyanotic CHD were VSD, followed by ASD. Isolated congenital heart defects constituted 82.6%. A total 66.7% CHD children were diagnosed during infancy. VSD and PDA encountered more among males while ASD predominated among females.

Keywords: Congenital Heart Defects (CHD), Acyanotic Heart Defects, Ventricular Septal Defect, Atrial Septal Defect, Tetralogy of Fallot.

Pediatrics Department, Faculty of Medicine and Health Sciences, University of Aden, Republic of Yemen.
Corresponding Author: Mariam T. Bin Yahia **Email:** mariambinyahia@yahoo.com

نمط وتشخيص أمراض القلب الخلقية لدى المرضى المترددين على مستشفى الصداقة التعليمي، مدينة عدن، اليمن، يناير 2017 - ديسمبر 2019

مريم طاهر بن يحيى، عدنان أأمين هادي، ايمان علي باصديق

ملخص الدراسة

المقدمة: أمراض القلب الخلقية هي مشكلة في بنية القلب ووظيفته منذ الولادة تستمر مدى الحياة. هدفت الدراسة تحديد نمط أمراض القلب الخلقية بين أطفال اليمن من الجنسين في مدينة عدن.

المنهجية: هذه دراسة استرجاعية أجريت في مستشفى الصداقه من يناير 2017 إلى ديسمبر 2019. تم جمع البيانات من السجلات الطبية في وحدة القلب بمستشفى الصداقة التعليمي وقد تم تشخيص الأطفال المصابين بأمراض القلب التاجية من خلال أخذ التاريخ والفحص السريري وايكو للقلب.

النتائج: مائة وأربعة وثمانون مريضا في وحدة القلب تم تشخيصهم بأمراض القلب الخلقية. تم استبعاد 10 أطفال بسبب الإصابة بأمراض القلب المكتسبة. إجمالي المرضى 138 طفلاً، شكلت الإناث 58% مقابل 42% للذكور. كان متوسط العمر18.8± 3.0 شهرًا ووسيط العمر 8 أشهر. سبعة وثمانون بالمائة مصابون بأمراض الشرايين التاجية غير المسببة للإزرقاق. كان عيب الحاجز بين البطينين VSD هو الأكثر انتشارًا 35.5%، يليه عيب الحاجز بين الأذينين (ASD) 14.5%، القناة الشريانية السالكة (PDA) 12.3% والقناة الأذينية - البطينية 5.8%. رباعية فالو (TOF) كانت النسبة الأكبر من أمراض الشرايين التاجية المزرقة 7.2%. أمراض القلب التاجية المنفردة شكلت 82.6% بينما 17.4% لديهم أكثر من عيب خلقي في القلب. الذكور أكثر اصابه بـ VSD وPDA بينما سادت الإناث في ASD. تم تشخيص 66.7% بأمراض الشرايين التاجية في عمر سنة.

الاستنتاج: أكثر أنواع العيب الخلقي انتشارًا هو VSD، يليه ASD. تشكل عيوب القلب الأحادية 82.6%. تم تشخيص 66.7%خلال السنة الأولى من العمر. أظهرت الدراسة أن VSD وPDAأكثر انتشارًا بين الذكور بينما ASD بين الإناث.

الكلمات المفتاحية: عيوب القلب الخلقية، عيوب القلب المسببة للإزرقاق، ثقب بين البطينين، ثقب بين الاذينين ورباعية فالو.

قسم طب الأطفال، كلية الطب والعلوم الصحية، جامعة عدن، الجمهورية اليمنية.

Introduction

Congenital heart disease (CHD) is a lifelong problem of the heart's structure and function present at birth, affecting the heart or adjacent great blood vessels and detected either at the time of birth or detected later in life[1,2]. These defects generally result from the aberrant development of a normal structure in fetus or failure of progress beyond the early stage of embryonic or early fetal development [3]. The defects can be broadly classified into cyanotic and acyanotic depending on their physiology [4,5]. Cardiac mal-development early in the embryo leads to significant morbidity and mortality [3]. Furthermore, delayed presentation of congenital heart defects and appearance of some complications significantly increases the mortality and morbidity in pediatric age group [6,7].

CHD is the most common congenital problem in children accounting for nearly 25% of all congenital malformations [8]. It affects 0.8 to 1.2% of live births globally until year 1990. In 2013, the global prevalence of CHD at birth was approximated to be around 1.8 cases per 1000 live births, a 4.2% (95% UI 2.8–5.6) increase since 1990. This high increment is due to the fact that the birth prevalence is likely to be higher and because of the inclusion of data for low- and middle-income countries [9]. It was indicated that nine-tenths of the world's children born with congenital heart disease live in locations with little to no care where mortality remains high. Several reports have documented in the past two decades' better survival rates for neonates with CHD in high-income countries; while those with severe forms of CHD in low and middle-income countries who have no available access to surgical treatment are more likely to die before their fifth birthday [3,9].

CHD may present at different ages from birth to adolescence [10]. Many cases are asymptomatic and discovered incidentally during routine health check-up [11]. The clinical manifestations of congenital heart disease depend on the type, severity of the defect, age and stage of developmental of the child. In neonatal period the presenting feature of CHD is cyanosis, respiratory distress, heart failure, failure to thrive and shock. In infancy and childhood, the usual presenting features are signs of cyanosis, digital clubbing, murmur, syncope, respiratory distress, squatting episodes, heart failure, arrhythmia, failure to thrive and recurrent lower respiratory tract infections [3]. The diagnosis is based on a careful history taking and thorough physical examination that may that will require the urgent guide for further evaluation, referral, and collaboration with acute care and cardiac specialists to manage this complex chronic illness [9-12].

The etiology of CHD is multifactorial with important contributions from both environmental and genetic factors [12]. Both genetic and environmental factors have been associated to play a role in the pathogenesis of the diseases. Extensive studies on human and animal experiments have been conducted on cardiac transcription factors, cardiac-specific genes, and signaling pathways, which are responsible for early cardiac morphogenesis. Moreover, novel

genetic methods were discovered as next generation sequencing and chromosomal microarrays that have led to further study the genes, non-coding RNAs and subtle chromosomal changes, clarifying their effects to the etiology of CHD. Literature reports have also implicated non-hereditary risk factors such as congenital rubella infection, teratogens, maternal age, diabetes mellitus, drug exposure, smoking during pregnancy, maternal obesity, and abnormal hemodynamics in causing CHDs. These etiological factors raise questions on multifactorial etiology of CHDs. There is a strong correlation between drug exposure, viral infections as congenital rubella infection, smoking during pregnancy, maternal obesity, and maternal hyperglycemia are also among the important non-genetic risk factors implicated to CHDs [3]. The most common malformation is ventricular septal defect (VSD) which accounts for approximately 25% of CHD. Majority of these structural abnormalities occurs as isolated cardiac defects. Twenty per cent of patients with CHD, however, have more than one cardiac defect, for example a VSD with a patent ductus arteriosus (PDA). In addition, approximately 25 per cent of patients with CHD also have non-cardiac malformation [13].

Diagnosis of CHD depends on the symptoms and signs of the child and the underlying provisional diagnosis who may require one or more of the following investigations of blood tests, chest radiography, electrocardiogram echocardiography, computed tomography, cardiac magnetic resonance imaging cardiac catheterization and cardiac angiography [12,13].

Despite advanced diagnostic facilities and improved cardiovascular medicine and surgery, it has enabled neonates, infants and children to reach to adulthood. Nevertheless, CHD is still considered one of the leading causes of neonatal and infant mortality with a heavy burden of disease within the pediatric population [10,12]. Limited data are available regarding pattern of CHD in this area. The objective of this study was to describe the pattern of CHD admitted in the pediatric cardiac ward at Al-Sadaqa teaching hospital in Aden city.

Methods

Study design

This is a retrospective hospital-based study conducted over a three years' period. The primary data source was the medical records of patients which were extracted from the Statistics Department in the hospital.

Study setting

The place of study was the cardiac unit of the general ward at Al-Sadaqa Teaching Hospital which is the main pediatric and maternity referral hospital in Aden, South Yemen. It is a public health facility serving people from Aden and adjacent governorates. The cardiac unit receives all patients suspected to have cardiac diseases.

Participants

All patients who were holding the final diagnosis of heart diseases admitted to the cardiac unit in the general ward of the hospital, during the study period 1st January 2017 to 31st December 2019 were included in this study who fulfilled the following inclusion requirements:

- **Inclusion criteria**
  All patients of both sexes with Yemeni nationality from 1month to 14 years of age holding the diagnosis of CHD.

- **Exclusion criteria**
  All patients who were diagnosed with acquired heart diseases.

Variables

Variables studied included age of patient, gender, clinical physical examination of patients, such as cyanosis, heart examination, electrocardiography and results of Echocardiography. Age of the patients, classified as 1month to one year, more than one year to 6 years and more than > 6 years to 14 years. The CHD were classified as cyanotic and non-cyanotic types.

Statistical analysis

The data collected were analyzed using SPSS for Windows version 20. Descriptive statistics were presented as frequency and percentage of various types of heart defects. Quantitative variables were presented as mean and SD or median and range as appropriate.

Ethical clearance

The permission for undertaking this study was obtained from the administrative office in the referred hospital. Informed consent was not required since the study was retrospective and performed by chart review without any potential risk. Ethical approval was obtained from the Ethical Committee at the Faculty of Medicine and Health Sciences, University of Aden, Yemen.

Results

A total of 148 patients of both sexes suspected to have heart problems were seen in the cardiac unit of the General Ward of Al-Sadaqa Teaching Hospital, Aden City, Yemen. Children who were diagnosed with acquired heart disease accounted for 10 who were consequently excluded from this study (n=10). The echocardiography proven patients with CHD accounted for a total of 138 children who were enrolled in this study (58% females and 42% males), with a female to male ratio of 1.4:1.0. The percentage of congenital heart diseases in the current study was 13.7% of the total admitted children. The ages of patient ranged from one month to 14 years with a mean age of 18.7±3.0 months and median age of 8 months at time of admission.

One hundred and twenty children (87%) out of the total 138 patients were diagnosed clinically, by electrocardiography and echocardiography as acyanotic heart defects and only 18 patients (13%) of them comprised cyanotic heart defects.

The commonest acyanotic CHD were ventricular septal defect (VSD) (35.5%), followed in order of frequency by atrial septal defect (ASD) (14.5%), patent ductus arteriosus (PDA) (12.3%) and pulmonary valve stenosis (2.9%). Cyanotic heart disease was less frequent with Tetralogy of Fallot (TOF) constituting 7.2% followed by transposition of the great arteries (TGA) in 3.6% of patients as shown in Table 1

Table 1: Frequency of Acyanotic and Cyanotic CHD (n=138)

	Congenital Heart Disease	No.	%
Isolated CHD	Ventricular septal defect (VSD)	49	35.5
	Atrial septal defect (ASD)	20	14.5
	Patent ductus arteriosus (PDA)	17	12.3
	Tetralogy of Fallot (TOF)	10	7.2
	AV Canal	8	5.8
	Pulmonary Stenosis (PS)	4	2.9
	Mitral regurgitation (MR)	3	2.2
	Others	3	2.2
Multiple CHD	VSD + ASD	7	5.1
	VSD + PDA	7	5.1
	TOGA + ASD	3	2.2
	TOGA + VSD	2	1.4
	VSD + Pulmonary hypertension	2	1.4
	Others	3	2.2

Table 2 shows a higher frequency of acyanotic CHD in females in comparision with males for VSD (32.5% vs. 39.7%), ASD (17.5% vs.10.3%); TOF (7.5% vs. 6.9) and multiple defect VSD with ASD (6.2% vs. 3.4%) and VSD with PDA (7.5% vs.1.7). On the other hand, more males had PDA compared to females (15.5% and 10% respectively).

The age of presentation ranged from infancy to 14 years whereby the 138 patients of CHD were divided into different age groups to identify the age group at which initial diagnosis of CHD was identified. Two-thirds (92 or 66.7%) was found among those younger than one year of age, whereas 37 (26.8%) found in the age group 1-6 years and 9 (6.5%) in those aged more than 6 years as illustrated in Table 3.

Table 2: CHD by Sex (n=138)

CHD	Males (n=58)		Females (n=80)		Total (n=138)
	No.	%	No.	%	
VSD	23	39.7	26	32.5	49
ASD	6	10.3	14	17.5	20
PDA	9	15.5	8	10	17
TOF	4	6.9	6	7.5	10
AV Canal	4	6.9	4	5	8
VSD + ASD	2	3.4	5	6.2	7
VSD + PDA	1	1.7	6	7.5	7
PS	2	3.4	2	2.5	4
MR	3	5.2	0	0	3
TGA + ASD	0	0	3	3.8	3
TGA + VSD	1	1.7	1	1.3	2
VSD + PH	2	3.4	0	0.0	2
Others	1	1.7	5	6.2	6

Table 3: CHD by the Age Group (n=138)

Type of CHD	Age group by year						Total	
	1 month-1year		>1year -6 year		>6year-14year			
	No.	%	No.	%	No.	%	No.	%
VSD	37	75.5	10	20.4	2	4.1	49	35.5
ASD	14	70	4	20	2	10	20	14.5
PDA	12	70.6	5	29.4	0		17	12.3
TOF	3	30	5	50	2	20	10	7.2
AV Canal	8	100	0	0.0	0	0.0	8	5.8
VSD + ASD	3	42.9	4	57.1	0	0.0	7	5.1
VSD + PDA	5	71.4	2	28.6	0	0.0	7	5.1
Pulmonary stenosis (PS)	3	75	1	25	0	0.0	4	2.9
Mitral regurgitation	2	66.7	0	0.0	1	33.3	3	2.2
TGA + ASD	1	33.3	1	33.3	1	33.3	3	2.2
TGA + VSD	0	0.0	2	100	0	0.0	2	1.4
VSD + PH	0	0.0	2	100	0	0.0	2	1.4
Others	4	66.6	1	16.7	1	16.7	6	4.3
Total	92	66.7	37	26.8	9	6.5	138	100.0

Discussion

CHD is the most common congenital anomaly among childhood with a relatively higher mortality rate more than other birth defects during the first year of life. The importance of CHD as a cause of childhood mortality has shown a significant global decrease in the frequency of deaths from the years 1990 to 2017, regardless of sex, age and socio-demographic index. In addition, the mortality rates did not show a homogenous pattern globally due to the variations in socio-economic status and the available medical and surgical care facilities in the different countries [14].

Furthermore, failure to thrive, recurrent infections, and undernutrition adversely affects the long-term health of these children. Limitations in daily activities also have a significant impact on social development of these children. The economic and social impacts on families are also significant in the absence of state-sponsored social support [15]. In the industrialized countries, noteworthy improvements are seen in terms of the expectancy and quality of life of children with CHD. Nevertheless, it is important to acknowledge that the expense and complexity of treating CHD is much greater extent than for many other childhood illnesses [14] .

The hospital prevalence of CHD in the current study was 13.7/1000 of total admissions that does not fall in agreement with what was reported from Pakistan (4 per 1000) [16]. Kiran and Kapoor from India reported prevalence rates of 1.9 per 1000 and 25 per 1000 among hospitalized children respectively [17,18].

This study does not give true prevalence of CHD in general among the children of the Yemeni population since it was conducted in the cardiac unit of Al-Sadaqa teaching hospital at Aden city. It is generally accepted that the improvement of diagnosis,

attention or awareness among the general pediatricians and early referral to the pediatric cardiologists has resulted in an increase of prevalence of real reported children with CHD.

Eighty seven percent of cases were acyanotic CHD and the remaining were cyanotic, this is well correlated with international studies [19,20].

The current study showed that females have higher frequency of CHD compared to males that does not coincide with the results of Alqurashi *et al* [21], nor in agreement with a study done in Northern Iran [12]. In this study, the most common CHD was the acyanotic type of VSD that was diagnosed in 35.5% which is similar to findings from India [18], Saudi Arabia [21], and Iran [22] but does not coincide with a study done by Kiran *et a l* [17] who found ASD as the most common form of CHD. The other forms of acyantoic heart defects in this study showed ASD comprising 14.5% and PDA (12.3%) which is similar to a study done by Alabdulgader in Saudi Arabia [23]. The most common acyanotic congenital heart defects constituted VSD, ASD and PDA among children in this study. Among the cyanotic lesions, TOF was the most common (7.2%) followed by AV Canal and Transposition of the great arteries, (5.8%, 2.9% respectively) which was comparable with others studies elsewhere [18,22,24,25] .

Kamal *et al* found no association between sex and types of CHD. In the present study, males showed higher frequencies of VSD, PDA, and females had higher frequencies of ASD [25].

Similarly, the age at which the initial diagnosis of CHD showed some differences in the literature. In this study, a higher proportion of children 66.7% were diagnosed in infancy, and 26.8% in 1-6 years. These results did not show any difference from the studies done in a tertiary care cardiac hospital of Karachi and Akash Hospital in Banglore [17] and Pediatric Cardiac Centre in Birj [19] and, Iran [22]. Thus, early screening and diagnosis of CHD is important for better prognosis and early intervention of medical and surgical treatment.

Such variation in the frequencies of defects and the age of diagnosis, in this study and other literature reports may be explained by the fact that this study was conducted in a tertiary care pediatric hospital within a limited period of time. This hospital receives some referrals from the private hospitals and polyclinics and other hospitals from neighboring governorates but other children diagnosed with CHD may also be admitted, diagnosed and receive their required treatment in other private hospitals in Aden city.

The etiology of CHD is still heterogenous with lack of substantial knowledge on the specific underlying cause for each anomaly worldwide. This in turn leads to problems in the prevention and screening programs of congenital heart disease in children. It is noteworthy to mention that the burden of congenital heart disease falls most heavily on low income countries and in those with the highest fertility rates [26].

Diagnostic and treatment capabilities for congenital heart disease have dramatically improved over the past

80 years. CHD is a worldwide, large, rapidly evolving problem in the field of infant and child health. Although there are large innovative global advances in cardiac medicine and surgery, reports have not yet documented to reveal significant alterations in the prevalence of CHD. Therefore, interventions and resources must be used to improve survival and quality of life [9]. The findings in this study highlights some of the results regarding inequities in the demographic characteristics of congenital heart disease and can serve as a starting point for policy changes to improve screening, treatment, and data collection.

Conclusion

The frequency of CHD reported in this study is consistent with other studies worldwide with the commonest acyanotic CHD were VSD, ASD and PDA while the commonest cyanotic CHD was TOF. Patients with isolated CHD were of very higher frequency that those with multiple lesions. Males showed a higher predominance in VSD and PDA while females in ASD. Echocardiography found to be important for confirmation of CHD in this study. Early referral of suspected children with CHD is mandatory for better medical and surgical management in under-developed countries like Yemen where parents of patients have shortage of money and inadequate resources for abroad treatment. These current alarming problems will lead to a helpful and partial supportive solution in the near future based on the speedy establishment of cardiac centers at different regions of the country.

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