Journal of Health and Medical Sciences

Almawazini, Abdulmajid Mustafa, Alzahrani, Abdulrahman Matar Abdullah, Saadi, Zaher Hassan, Almawazini, Hazem Abdulmajid, Almawazini, Mohammad Abdulmajid, Zahrani, Saeed Daifalla, Alsaadi, Saud Hussien, Eidoh, Mohammed Abdulla, Alghamdi, Abdulrahman Mohammed, and Antar, Ahmed Mohammed Al. (2019), The Prevalence and the Common Types of Cyanotic Congenital Heart Diseases in Albaha, Saudi Arabia. In: *Journal of Health and Medical Sciences*, Vol.2, No.3, 332-336.

ISSN 2622-7258

DOI: 10.31014/aior.1994.02.03.53

The online version of this article can be found at:
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The Asian Institute of Research

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The Asian Institute of Research
Journal of Health and Medical Sciences
Vol.2, No.3, 2019: 332-336
ISSN 2622-7258
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DOI: 10.31014/aior.1994.02.03.53

The Prevalence and the Common Types of Cyanotic Congenital Heart Diseases in Albaha, Saudi Arabia

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Abstract
Background: Cyanotic congenital heart disease accounts about one-third of all congenital heart disease, and the prevalence of it vary from center to another due to related factors. Objectives: The study aimed to provide the prevalence and the most common types and distribution of CCHD in children less than 14 years of age in Albaha area Saudi Arabia. Methods: Hospital-based retrospective observational cross-sectional study involved all cases of Cyanotic congenital heart disease (CCHD) diagnosed at the King Fahad Hospital Albaha, Saudi Arabia, between January 2006 and January 2019. All patients were screened by echocardiography to confirm the diagnosis. Results: Total of 314 patients were diagnosed as cyanotic congenital heart disease (CCHD) in our center. There were 158 males (50.32%) and 156 females (49.68%). The age of patients was from 1 day to 14 years with a mean of 35.42±4.1 months. The neonatal period was the modal age at diagnosis. Approximately 80% of the children had been diagnosed at the age of < 1 yr and 20% of cases diagnosed after that. Tetralogy of Fallot (TOF) was the most common CCHD; it was diagnosed in 125 pts 39.81%. Double Outlet Right Ventricle (DORV) diagnosed in 48 pts 15.29%. Transposition of the Great Arteries (TGA) diagnosed in 34 pts 10.82. Pulmonary atresia 20 pts, 6.37%., Truncus arteriosus 20 pts, 6.37%., and Hypoplastic Left Ventricle Syndrome 20 pts, 6.37%. The other cyanotic congenital heart disease (CCHD) was 13.16%. Cyanosis was the common clinical presentation in all patients. Conclusions: The prevalence of cyanotic congenital heart disease in Albaha area was 14.62% of all congenital cardiac abnormalities, and no significant difference between male and female patients was seen. The most common types were TOF. Cyanosis was the most common clinical presentation. The outcome can be improved by early diagnosis. CCHD screening program associated with physical examination can help in early diagnosis.

Keywords: Cyanotic Congenital Heart Disease, Prevalence, and Presentation

Introduction

The cyanotic congenital heart diseases (CCHD) include many types of structures abnormalities with impaired cardiac function. It can be a result of decreased pulmonary flow as in Tetralogy of Fallot (TOF), pulmonary atresia, and right side hypoplastic heart, or with decreased aortic flow as in left-sided hypoplastic heart, interrupted arch, and severe coarctation, or due to blood mixing inside or outside the heart as in TGA, DORV, and TA, etc. **(**Allen HD, Driscoll DJ, Shaddy RE, et al., 2013. Jeffrey R. Boris, Marie J. Béland Lisa J. Bergensen, et al. 2017**).** CCHD accounts about one-third of the congenital heart disease**,** and the prevalence of it varies worldwide (Hoffman JI, Kaplan S 2002**,** Kennedy N, Miller P 2013**)**. The common CCHD include tetralogy of fallot (TOF), double outlet right ventricle (DORV), transposition of the great arteries (TGA), total anomalous pulmonary venous return (TAPVR), truncus arteriosus and tricuspid atresia (TA), **(**Van der Linde D, Konings EE, Slager MA, et al 2011**).** Ebstein's anomaly, Hypoplastic left heart syndrome (HAHS), pulmonary atresia (PA), tricuspid atresia (TA), pulmonary atresia (PA), and single ventricle, are the less types of CCHD, (Van der Linde D, Konings EE, Slager MA, et al. 2011). The most common type is TOF **(**O'Brien P, Marshall AC, et al. 2014**).** The presentation depends on the type of abnormalities, the severity of obstruction, and the cardiac output. Some CCHD may present early like TGA, HLHS**,** PA, and TA, while the signs may be presented late in the 1st year in the patients with TOF **(**Animasahun BA, Madise-Wobo AD, Gbelee HO, et al. 2017; Okoromah CA et al. 2011**).** The cyanosis, feeding difficulties, failure to thrive, and respiratory problems are the common clinical findings in patients have CCHD**.** Some chemical risk factors, genetic disorders, consanguinity, intrauterine infections like rubella, medication used in pregnancy, and chromosomal abnormalities as Down, Noonan, and Turners syndromes, can be considered as risk factors of CCHD, but the exact etiology is unknown **(**Gorini F, Chiappa E, Gargani L, et al 2014; Campbell M et al 1961**;** Shawky RM, Elsayed SM, Zaki ME, et al 2013**).** The management of children with CCHD is multidisciplinary**.** Surgical correction is the main stone of the management combined with medical treatment, is important to give the proper care for the patients **(**Radu S, Floria M, Baroi GL, et al. 2016**).** Limited reports were available about the cyanotic congenital heart disease in Saudi Arabia, and the data about CCHD is included in reports about all congenital heart diseases **(**Alabdulgader AA et al. 2006**).** Critical congenital heart disease screening (CCHD) program has a good rule in early detection of cyanotic cardiac defects **(***Abdulmajid M. Almawazini et al. 2017;* Movahedian AH, Mosayebi Z, Sagheb S, et al. 2016**).** Antenatal diagnosis can be done in utero by fetal echocardiography (Chew C, Halliday JL, Riley MM, et al. 2007; Hoffman JIe et al. 2013).

Objectives

The study aimed to provide the prevalence and the most common types, clinical presentation, and distribution of CCHD in children less than 14 years of age in Albaha area Saudi Arabia.

Methods

Hospital-based retrospective observational cross-sectional study included the cases of CCHD. The study was conducted in the Pediatric and Neonatology Department, King Fahad Hospital, Albaha, Saudi Arabia from January 2006 to January 2019. The study was approved by the Scientific Research and Ethical Committee in the hospital. Statistical Package for Social Sciences (SPSS) was used to analyze the data and the outcome. Statistical significance were $P<0.05$, 95%CI, OR, and RR. All the patients were aged 0–14 years. Mean age ± standard deviation was considered. The inclusion criteria were all patients diagnosed to have cyanotic congenital heart. Patients had acyanotic heart disease, neonates with patent ductus arteriosus (PDA) or patent foramen ovale (PFO), and other types of heart diseases were excluded. Echocardiography was performed using Philips IE33 Ultrasound (Philips, Bothel, WA, USA). 2-D echocardiography, colored Doppler, and M-mode were used for the diagnosis of structures abnormality and the evaluation of cardiac function and hemodynamic status for all patients, (Alabdulgader AA et al. 2006). A detailed history and physical examinations were performed for all. Cyanotic congenital heart disease screening program was applied in our center in the last 4 years, and was done for all neonates (*Abdulmajid M. Almawazini et al. 2017*). Chest radiograph, electrocardiography, and laboratory investigations were done as required. For surgical intervention, patients were referred for higher cardiac centers in Saudi Arabia. The outcomes were prevalence of CCHD, most common types, and the clinical presentation. The files of the patients were reviewed retrospectively and findings recorded. Level of significance was P value

< 0.05. Odd ratio (OR), Relative Risk (RR), and 95% confidence interval were considered. The protocol used in this study was convinced with AHA/ACC circulation for the diagnosis, classification, evaluation of heart structures and function, and guidelines for the care of children with congenital heart disease, **(**Jeffrey R. Boris, Marie J. Béland Lisa J. Bergensen, et al. 2017**).**

Results

Table 1: Distribution of all children screened during the time of the study.

Patients		Number	Notes
Total children		8035	Screened
Normal		1640	Excluded
PFO closed in 1st year		2557	Excluded
PDA closed in 1st month		1623	Excluded.
Heart Disease (no CHD)		67	Excluded.
CCHD 2148 Pts	Acyanotic CHD	1834 (85.38%)	Excluded
	Cyanotic CHD	314 (14.62%)	Included
CCHD: cyanotic congenital heart disease; CHD: congenital heart disease; PFO: patent foramen ovale; PDA: patent ductus arteriosus; Pts: patients.			

Table 2: The Types and Prevalence of CCHD diagnosed in this study.

Type CCHD	Pts number	% CHD	% CHD	Male	Female	P value
All Pts	314	100%	15.13	158 (50.32%)	156 (49.68%)	0.68
TOF	125	(39.81)	5.82	65 (52%)	60 (48%)	0.65
DORV	48	(15.29)	2.23	20 (41.67%)	28 (58.33%)	0.54
TGA	34	(10.82)	1.58	21 (61.76%)	13 (38.24%)	0.48
PA	20	(6.37)	0.93	9 (45%)	11 (55%)	0.64
TRUNCUS	20	(6.37)	0.93	6 (30%)	14 (70%)	0.08
HLHS	20	(6.37)	0.93	15 (75%)	5 (25%)	0.06
CCCHD	10	(3.18)	0.47	6 (60%)	4 (40%)	0.63
TAPVR	8	(2.48)	0.37	2 (25%)	6 (75%)	0.06
Ebstein's anomaly	6	(1.9)	0.28	3 (50%)	3 (50%)	0.72
Interrupted Arch	5	(1.6)	0.23	3 (60%)	2 (40%)	0.59
Single ventricle	5	(1.6)	0.23	3 (60%)	2 (40%)	0.59
Inoperable CCHD	5	(1.6)	0.3	3 (60%)	2 (40%)	0.59
DILV	3	(0.9)	0.14	2 (66.67%)	1 (33.33%)	0.08
TA	3	(0.9)	0.14	1 (33.33%)	2 (66.67%)	0.08
HRHS	2	(0.6)	0.1	1 (50%)	1 (50%)	0.71

CCHD: cyanotic congenital heart disease; CHD: congenital heart disease; DORV: Double outlet right ventricle; DILV: Double inlet left ventricle; HLHS; Hypoplastic left ventricle syndrome; HRVS: Hypoplastic right ventricle syndrome; PA: pulmonary atresia; AVSD: atrio-ventricular septal defect; TOF: Tetralogy of Fallot; TGA: transposition of great arteries.

Between January 2006 and January 2019, a total of 2148 patients diagnosed as congenital heart disease. Cyanotic congenital disease (CCHD) was diagnosed in 314 patients (14.62%) in our center **(table 1).** There were 158 males (50.32%) and 156 females (49.68%) with no significant difference, (OR 1.1. P= 0.68). The age of patients was from 1 day to 14 years with a mean of 35.42±4.1 month, and the neonatal period was the modal age at diagnosis. Approximately 80% of the children had been diagnosed at age of less than 1 year and 20% of cases diagnosed after that. As mentioned in (**table 2)**, our results revealed that the most common type of CCHD was Tetralogy of Fallot (TOF), which was diagnosed in 125 pts 39.81%; there were 65 males 52% and 60 females 48% with a male to female ratio of 1.1:1. Double Outlet Right Ventricle (DORV) diagnosed in 48 pts 15.29%, with 20 males 41.67% and 28 females 58.33%, a female to male ratio of 1.4:1. Transposition of the Great Arteries (TGA) diagnosed in 34 pts 10.82%, with 21 males 61.76% and 13 females 38.24% with a male to female ratio of 1.6:1. Pulmonary atresia diagnosed in 20 pts, 6.37%, there were 9 males 45% and 11 females

55% with a female to male ratio of 1.2:1. Truncus arteriosus diagnosed in 20 pts, 6.37%, there were 6 males 30% and 14 females 70% with a female to male ratio of 2.3:1. Hypoplastic Left Ventricle Syndrome diagnosed in 20 pts, 6.37%, there were 15 males 75% and 5 females 25% with a female to male ratio of 3:1. Total Anomalous pulmonary veins connection (TAPVR) diagnosed in 8 patients 2.48%, and there were 2 males 25% and 6 females 75% with a female to male ratio of 3:1. Ebstein anomaly diagnosed in 6 patients 1.9%, there were 3 males 50% and 3 females 50% with no difference between male and female. The other types of cyanotic congenital heart disease were diagnosed in 33 pts 10.51%. Inoperable complex cyanotic congenital heart disease was diagnosed in 5 pts (1.6%), two patients had Dextrocardia with pulmonary atresia and large ventricular septal defect (VSD) and atrial septal defect (ASD), two pts had truncus arteriosus with severe hypoplasia in the all branches of pulmonary arteries, and one patient has complex CCHD. Cyanosis was the common presentation, documented in 300 cases 95.54% of all patients, cardiac murmur in 275 pts 78.57%. Features of Down syndrome presented in 25 pts 7.97% and the majority of them diagnosed to have TOF. Signs of heart failure were confirmed in 20 pts 6.37%. Feeding difficulties and weight affected seen in 10 pts 3.18%, and associated congenital anomalies in 5 pts 1.6%.

Discussion

Data were reviewed for all cases of CCHD over 13 years (2006-2019). It was documented the distribution, prevalence, and clinical presentation, of cyanotic congenital heart disease in King Fahad Hospital Albaha, Saudi Arabia. There are previous reports from Albaha Area about congenital heart disease, but no previous reports about CCHD. The critical cyanotic congenital heart disease screening program (CCHD) was applied in our hospital, and other centers in Albaha area for all newborns at the age of 24 hours for early diagnosis of CCHD, **(***Abdulmajid M. Almawazini et al. 2017;* Movahedian AH, Mosayebi Z, Sagheb S, et al. 2016**).** About 80% of the patients were diagnosed in early infancy while only 20% of cases diagnosed in older ages (P= 0.036), same as mentioned in international reports **(**Hoffman JI, Kaplan S 2002**,** Kennedy N, Miller P 2013**,** Van der Linde D, Konings EE, Slager MA, et al. 2011**).** Some cases diagnosed in utero by fetal echocardiography **(**Chew C, Halliday JL, Riley MM, et al. 2007**;** Hoffman JIe et al. 2013**).** Less than 1% of the cases discharged before screening and less than 1% missed and did not diagnose till the age of 2 years and more when patient had clear central cyanosis **(**Shawky RM, Elsayed SM, Zaki ME, et al. 2013; Radu S, Floria M, Baroi GL, et al. 2016**).** There was no significant male to female ratio difference in total CCHD, but we found male more affected in TOF, TGA, HLHS, while female more affected in TAPVR, Truncus arteriosus, and DORV, and no significant difference in residual types of CCHD, as documented in international studies **(**Kennedy N, Miller P 2013**,** Van der Linde D, Konings EE, Slager MA, et al 2011**;** Sadoh WE, Uzodimma CC, Daniels Q et al 2013**).** Tetralogy of Fallot (TOF) was the most common, it was diagnosed in 125 patients (39.81), and associated in 25 patients by down syndrome features, same as mentioned in other published studies **(**O'Brien P, Marshall AC et al 2014**;** Alabdulgader AA et al 2006; Sadoh WE, Uzodimma CC, Daniels Q et al 2013**).** DORV and TGA were established as the other most common CCHD (Hoffman JI, Kaplan S 2002**,** Kennedy N, Miller P 2013**;** Okoromah CA et al. 2011**)**.

Truncus Arteriosus, TA 3 pts, and TAPVR 8 pts were diagnosed in our study, maybe of the long period covered by this study, as reported in national and international reports **(**Hoffman JI, Kaplan S 2002**,).** The less common defects were Single Ventricle, and Ebstein's anomaly, which is similar to what was published in some previous national and international studies. The cyanosis and cardiac murmur were the most common clinical presentation and the reasons to transfer the patients for cardiac evaluation. Easy fatigability, tachypnea, tachycardia, feeding difficulties, failure to thrive, and signs of heart failure were also seen and common signs in the patients with cyanotic congenital heart disease (Allen HD, Driscoll DJ, Shaddy RE, et al 2013**;** Van der Linde D, Konings EE, Slager MA, et al 2011**;** Okoromah CA et al 2011**)**. The patients managed as multidisciplinary team **(**Jeffrey R. Boris, Marie J. Béland Lisa J. Bergensen, et al. 2017**;** Radu S, Floria M, Baroi GL, et al. 2016**),** medical treatment started, and all patients were transferred to a higher cardiac center for surgical correction. Complete correction is done for the majority of the patients while univentricular repair done for 10 patients. Five patients had complex inoperable CCHD were kept on medical treatment because no surgical intervention could be done for them.

Conclusions

Cyanotic congenital heart disease is common in Albaha area, and males to female ratio vary in different types of CCHD. Echocardiography was the safe and easy investigation can confirm the diagnosis. Cyanosis and cardiac murmur were the common presentation in the majority of the cases. Early diagnosis can improve the outcome. Multidisciplinary team is an ideal for management of patients with cyanotic congenital heart diseases.

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