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Research Article

Congenital Heart Disease: Spectrum and Distribution at a Tertiary Health Care Centre in Western India

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Abstract

Background: Changing pattern and incidence of congenital heart diseases (CHD) have been observed in various geographical locations.

Objectives: To study the frequency, age-wise distribution, and spectrum of congenital heart diseases (CHD) at a tertiary health care centre in Ajmer, Rajasthan.

Methods: A retrospective analysis of case records of patients in the age group of 0 - 18 years referred for 2D echocardiography from January 2008 to July 2015 was done to ascertain the spectrum and distribution of CHDs. Clinical examination, electrocardiography, chest X ray and transthoracic echocardiography (TTE) were used as diagnostic tools.

Results: Out of 8,641 patients, 2052 (23.75%) were found to have CHD. Male preponderance was observed (male to female ratio = 1.43). Study group comprised of 12.62% neonates, 39.38% infants and 47.81% of more than 1 year age. 1742 (84.89%) were acyanotics, and 310 (15.11%) suffered from cyanotic heart disease. Among the acyanotic heart diseases ventricular septal defect (VSD) was the most frequent lesion seen in 700 (40.18%), followed by atrial septal defect (ASD) in 370 (21.24%) children**.** Tetralogy of Fallot (TOF) was the most frequent cyanotic heart disease seen in 196 (63.23%) patients.

Conclusion: The frequency of CHD at a tertiary care centre in western India was 23.75 percent. VSD and ASD were the most common acyanotic while TOF was the commonest cyanotic congenital heart defect observed. TTE plays an important role in the diagnosis of CHD. When clinical evidences lead to suspicion of congenital heart defect is suspected, an echocardiography should be performed.

Keywords: *Congenital Heart Disease; Spectrum; Transthoracic Echocardiography*

Key Message

This study aimed to fill in the caveat in epidemiology of congenital heart disease by studying the spectrum and distribution of CHD in western India. TTE plays major role in diagnosis of CHD. The spectrum and distribution of CHD in western India was found to be similar to other areas.

Introduction

Congenital heart disease (CHD), was defined by Mitchell., *et al.* as "a gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance" [1]. 10% of the present infant mortality in India may be accounted for by CHD [2]. Studies have reported that CHD spectra differ according to geographical location [3]. The reported prevalence of CHD in India ranges from 0.8 - 5.2/1000 patients [4,5]. Prevalence of CHDs is reported higher in India than other parts of the world, being 5% in Chan-

digarh, 3% in Punjab and 2.5% in South India. Prevalence studies of CHD are required to establish baseline rates, and geographical trends that may help to raise the awareness of early medical and surgical intervention. It is critical in understanding the social and economic burdens placed on the families, demands placed on the health care system and health planning. 2-dimensional echocardiography with colour Doppler has revolutionized the diagnosis and management of cardiac malformations. We conducted this study to assess the prevalence of CHD among patients attending a tertiary care centre in western India.

Materials and Methods

A retrospective analysis of case records of patients in the age group of 0-18 years from January 2008 to July 2015 referred for 2D echocardiography was done to ascertain the spectrum and distribution of CHDs.

Inclusion criteria

Any patient having the signs and symptoms like shortness of breath, difficulty in feeding, excessive sweating, bluish discoloration of lips and tongue, failure to thrive, clubbing, palpitation, feeling of impending doom, fainting, light headedness, rapid breathing, discrepancy in pulse, cyanosis, heart murmur, abnormal chest X-ray, or strong family history, recurrent chest infections, high blood pressure, swelling of abdomen and feet, chest and abdomen pain, and arrhythmias and loss of consciousness, etc. were evaluated further. Those suspected of cardiac disease were subjected to chest X-ray, electrocardiogram (ECG), followed by echocardiography.

Exclusion criteria

Neonates less than 2 weeks of age with diagnosis of patent ductus arteriosus (PDA), known CHD patients presenting on follow up visits were excluded.

Methods

Patients with suspected CHD were further evaluated with12- lead ECG, chest X ray and diagnosis was confirmed by transthoracic 2D echocardiography (TTE).

Echocardiography was performed by senior cardiologists as per standards laid down by the American Society of Echocardiography, using the M-mode, two-dimensional and color Doppler, pulse and continuous wave echocardiogram [9]. TTE was done in subcostal, apical four chamber, apical two chamber, apical long axis, parasternal long axis, parasternal short axis (at various level of left ventricle like: basal, mid cavity or at level of papillary level and apical part), parasternal high short axis (at aortic valve, pulmonary valve level) and suprasternal view. The following age groups were considered: Neonates (1 - 30 days), infants (1 - 12 months), children and adolescents (> 1 - 18 years). Informed written consent was obtained from parents and/or attendants from all enrolled patients. The data was analyzed in Microsoft office excel spread sheet.

Results

During the study period of 6.5 years, TTE was performed on 8,641 patients. 3942 patients were found to have CHD. 214 neonates with PDA aged less than 2 weeks and were excluded. 1676 patients were excluded as they came on follow up visits after initial diagnosis of CHD. 2052 (23.75%) patients with CHD fulfilled the study criteria and were included in the study (Figure 1). Study group comprised of 12.62% neonates (n = 259), 39.38% infants (n = 808) and 47.81% of more than 1year age (n = 985).

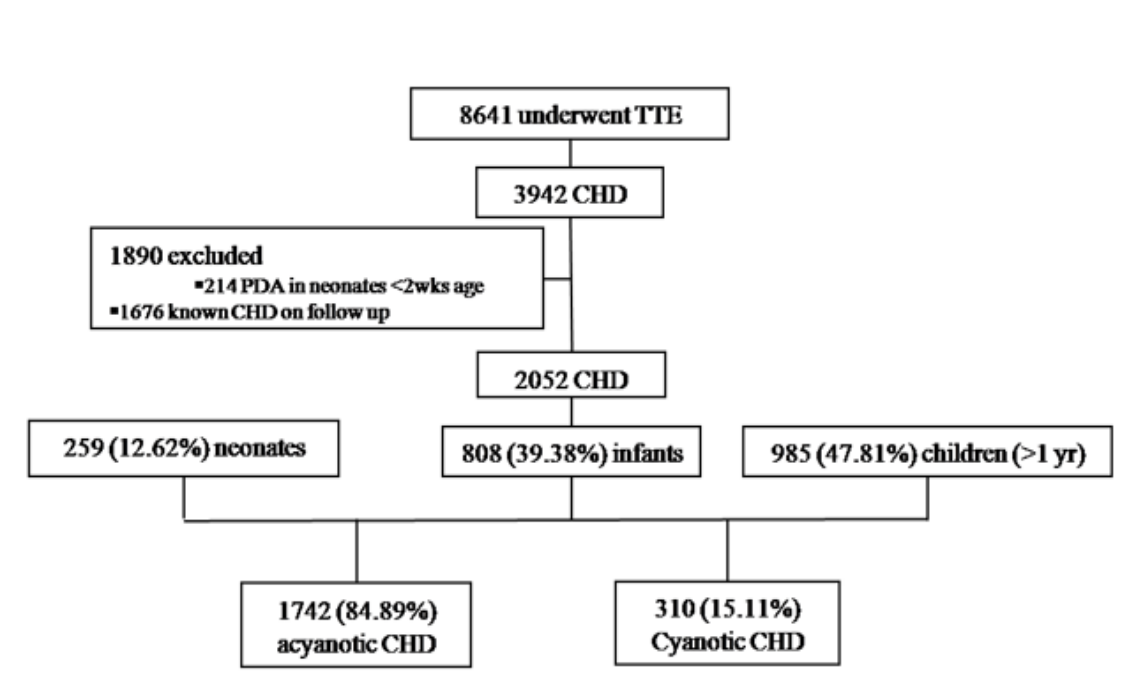

Figure 1: *Flow of patients in the study.*
CHD: Congenital Heart Disease; TTE: Transthoracic Echocardiography; PDA: Patent Ductus Arteriosus

Most common symptom was dyspnea (55.56%) followed by recurrent lower respiratory tract infections (42.54%) and failure to thrive (27.39%). 230 (11.21%) patients were asymptomatic and were evaluated on ground of cardiac murmurs (Table 1).

Symptoms	Number (%)
Dyspnea	1140 (55.56)
LRTI	873 (42.54)
FTT	562 (27.39)
Cyanosis	310 (15.11)
CCF	256 (12.48)
Feeding difficulty	232 (11.31)
Cyanotic spells	123 (5.99)
Asymptomatic	230 (11.21)

Table 1: *Symptomatology of CHDs.*
Data are presented as n (%). LRTI: Lower Respiratory Tract Infection;
FTT: Failure to Thrive; CCF: Congestive Cardiac Failure.

CHD was more common in males in our study (male to female ratio = 1.43). PDA, mitral valve prolapse (MVP), bicuspid aortic valve (BAV) and pulmonary atresia (PA) were more common in females. Rupture of sinus of valsalva (RSOV), double outlet right ventricle (DORV) and tricuspid atresia (TA) had no gender preponderance. Rest of the CHDs were more common in males (Table 2).

Lesion	Total	Male	Female
VSD	700 (34.48)	411 (58.71)	289 (41.29)
ASD	370 (18.23)	207 (55.95)	163 (44.05)
PDA	163 (8.03)	78 (47.85)	85 (52.15)
PS	134 (6.60)	87 (64.93)	47 (35.07)
TOF	196 (9.66)	118 (60.20)	78 (39.80)
PFO	120 (5.91)	75 (62.5)	45 (37.5)
MVP	102 (5.02)	39 (38.24)	63 (61.76)
AS	33 (1.63)	26 (78.79)	7 (21.21)
Malposition	32 (1.58)	23 (71.88)	9 (28.12)
TGA	32 (1.58)	22 (68.75)	10 (31.25)
BAV	28 (1.38)	6 (21.43)	22 (78.57)
LVNC	14 (0.69)	10 (71.43)	4 (28.57)
AVSD	20 (0.99)	14(70.00)	6 (30.00)
DORV	18 (0.89)	9 (50.00)	9 (50.00)
TA	14 (0.69)	7 (50.00)	7 (50.00)
SV	10 (0.49)	6 (60.00)	4 (40.00)
PA	8 (0.39)	2 (25.00)	6 (75.00)
HCM	10 (0.49)	10 (100.00)	0 (0.00)
COA	6 (0.30)	4 (66.67)	2 (33.33)
EBSTEIN	6 (0.30)	4 (66.67)	2 (33.33)
SA	6 (0.30)	2 (33.33)	4 (66.67)
RSOV	4 (0.20)	2 (50.00)	2 (50.00)
IAA	2 (0.10)	2 (100.00)	0 (0.00)
COR T	2 (0.10)	2 (100.00)	0 (0.00)

Table 2: *Overall distribution of congenital heart diseases including multiple congenital cardiac defects. Data are presented as n (%). VSD: Ventricular Septal Defect; ASD: Atrial Septal Defect; PDA: Patent Ductus Arteriosus; AS: Aortic Stenosis; PS: Pulmonary Stenosis; TOF: Tetralogy of Fallot; TGA: Transposition of Great Arteries; DORV: Double Outlet Right Ventricle; AVSD: Atrioventricular Septal Defect; PFO: Patent Foramen Ovale; MVP: Mitral Valve Prolapsed; BAV: Bicuspid Aortic Valve; LVNC: Left Ventricle Non Compaction; HCM: Hypertrophic Cardiomyopathy; COA: Coarctation of Aorta; RSOV: Rupture Sinus of Valsalva; IAA: Interrupted Aortic Arch; TA: Tricuspid Atresia; SV: Single Ventricle; PA: Pulmonary Atresia; SA: Single Atrium; Cor T: Cortriatriatum.*

1742 (84.89%) patients were acyanotic while 310 (15.11%) patients suffered from cyanotic heart disease (Figure 1). Among the acyanotic heart diseases isolated ventricular septal defect (VSD) was the most frequent lesion seen in 700 (40.18%), followed by isolated atrial septal defect (ASD) in 370 (21.24%) children (Figure 2). Tetralogy of Fallot (TOF) was the most frequent cyanotic heart disease seen in 196 (63.23%) patients followed by transposition of great arteries in 32 (10.32%) patients (Figure 3). Age-wise distribution of CHD showed that VSD was the most common acyanotic CHD irrespective of the age, but among cyanotic CHD neonates most commonly had transposition of great arteries (TGA) while TOF was commonest in rest (Table 3).

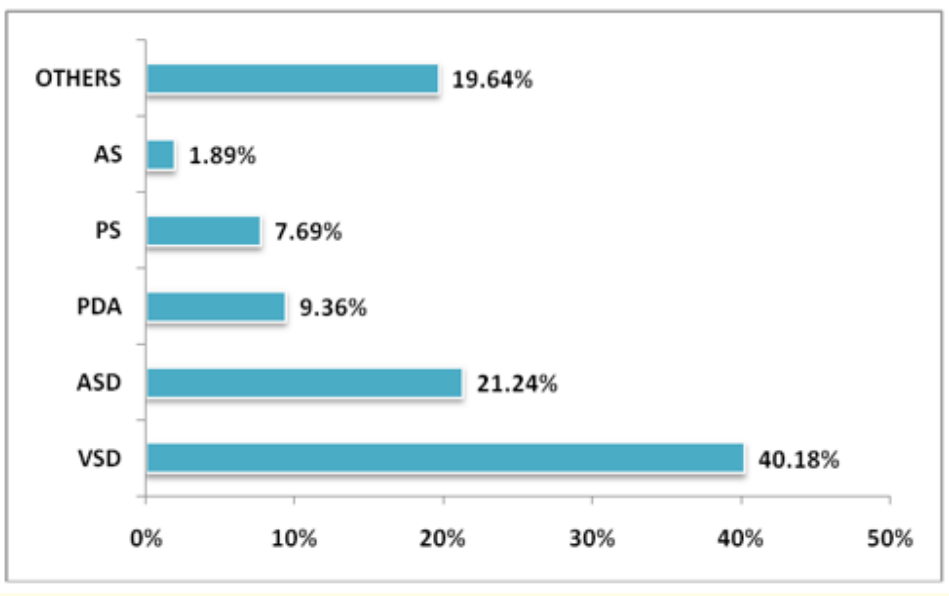

Figure 2: *Bar diagram showing distribution of acyanotic congenital heart diseases. Data are presented as %.VSD: Ventricular Septal Defect; ASD: Atrial Septal Defect; PDA: Patent Ductus Arteriosus; AS: Aortic Stenosis; PS: Pulmonary Stenosis; Others (PFO: Patent Foramen Ovale; MVP: Mitral Valve Prolapsed; BAV: Bicuspid Aortic Valve; LVNC; Left Ventricle Non Compaction; HCM: Hypertrophic Cardiomyopathy; COA; Coarctation of Aorta; RSOV: Rupture Sinus of Valsalva; IAA: Interrupted Aortic Arch).*

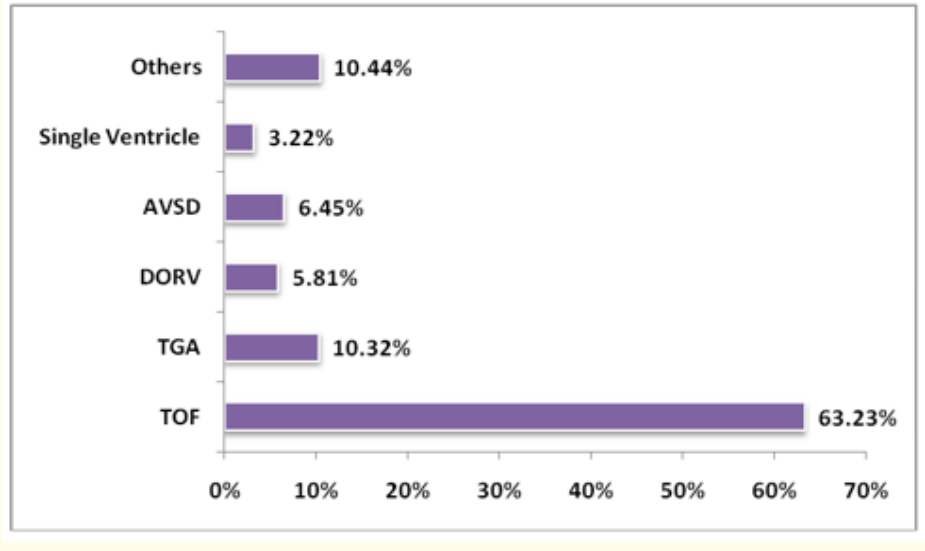

Figure 3: *Bar diagram showing distribution of acyanotic CHD. Data are presented as %. TOF- tetralogy of fallot; TGA: Transposition of Great Arteries; DORV: Double Outlet Right Ventricle; AVSD: Atrioventricular Septal Defect; Others (TA: Tricuspid Atresia; SV: Single Ventricle; PA: Pulmonary Atresia; SA: Single Atrium; Cor T: cortriatriatum).*

Children (> 1 year)			
Acyanotic CHD	**Total number (%)**	**Cyanotic CHD**	**Total number (%)**
VSD	306 (37.27)	TOF	120 (75.00)
ASD	160 (19.49)	DORV	12 (7.5)
PDA	92 (11.21)	TGA	8 (5.0)
MVP	86 (10.48)	SV	6 (3.75)
PS	48 (5.85)	EBSTEIN	4 (2.5)
AS	33 (4.62)	AVSD	4 (2.5)
BAV	26 (3.17)	PA	4 (2.5)
Malposition	16 (1.95)	TA	2 (1.25)
LVNC	10 (1.22)	COR T	2 (1.25)
Infants (>1 Month- 1 Year)			
Acyanotic	**Total number (%)**	**Cyanotic**	**Total number (%)**
VSD	318 (46.49)	TOF	72 (58.06)
ASD	146 (21.35)	AVSD	16 (12.90)
PDA	38 (5.56)	TGA	12 (9.68)
PFO	32 (4.68)	TA	10 (8.06)
PS	18 (2.63)	DORV	4 (3.23)
Malposition	10 (1.46)	PA	4 (3.23)
Neonates (Upto 1 month)			
Acyanotic	**Total number (%)**	**Cyanotic**	**Total number (%)**
VSD	318 (46.49)	TOF	72 (58.06)
ASD	146 (21.35)	AVSD	16 (12.90)
PDA	38 (5.56)	TGA	12 (9.68)
PFO	32 (4.68)	TA	10 (8.06)
PS	18 (2.63)	DORV	4 (3.23)
Malposition	10 (1.46)	PA	4 (3.23)

Table 3: *Age-wise distribution of congenital heart diseases*

Data are presented as n (%). VSD: Ventricular Septal Defect; ASD: Atrial Septal Defect; PDA: Patent Ductus Arteriosus; AS: Aortic Stenosis; PS: Pulmonary Stenosis; TOF: Tetralogy of Fallot; TGA: Transposition of Great Arteries; DORV: Double Outlet Right Ventricle; AVSD: Atrioventricular Septal Defect; PFO: Patent Foramen Ovale; MVP: Mitral Valve Prolapsed; BAV: Bicuspid Aortic Valve; LVNC: Left Ventricle Non Compaction; TA: Tricuspid Atresia; SV: Single Ventricle; PA: Pulmonary Atresia; SA: Single Atrium; Cor T: Cortriatriatum.

Discussion

Congenital heart disease is one of the major diseases in the paediatric age group occurring in 8 per 1000 live births [10]. Along with neural tube defects, CHD accounts for two-thirds of all major birth defects, being an important cause of morbidity and mortality in infancy [11]. We carried out this study as there are very few Indian studies stating the epidemiology of CHD in our country. Khail., *et al.* studied 10964 live births and reported the incidence of CHD to be 3.9 per 1000 live births in India [11]. In this study, CHD was found in 23.75% cases. This is not a real picture of the prevalence rate of CHD in community as it is a hospital-based study and included only those patients who were born in our hospital, and referred from other hospitals.

Clinically dyspnea was the commonest presenting symptom followed by respiratory tract infections and failure to thrive. The same picture is reported in studies all over the world [11-13].

In accordance with previous studies, our study also showed a male preponderance [8,14,15]. Some CHDs have sex predilection. TGA and left sided obstructive lesions occur more commonly in boys, whereas VSD, PDA, ASD and pulmonary stenosis (PS) are more common in girls. However, in our study only PDA, pulmonary atresia (PA), MVP and bicuspid aortic valve were more common in females. TGA, left sided obstructive lesions, TOF, ASD, VSD, PS were all common in males.

Acyanotic CHD formed the major bulk (84.89%) in our study which is in congruence with other Indian and western studies [3,16-18]. VSD was the commonest congenital heart defect similar to other studies [3,19-21]. ASD was the second most common CHD, occurring in 21.4% which is higher than the frequency of 6 - 8% reported in western countries but correlated well with the frequency of Indian studies (10% - 23%) [22]. TOF was the most common cyanotic CHD correlating well with other studies [23-25].

Maximum number of cases of CHD was of the age group 0 - 1 year (52.0%) including neonates and infants which is in accordance with other studies from rest of India and rest of the world [3,26-28]. The frequency of the complex and rare types of CHDs was less when compared to the western data but similar to other Indian studies [29,30]. This could be either due to racial and genetic factors or due to death of the babies in utero and in early neonatal period from severe CHD before accessing medical facilities. Various CHD escape diagnosis like neonates born at home, who die without medical attention and those individuals who are asymptomatic with mild to moderate degree of CHD. This might have led to falsely low prevalence of CHD in our study.

The magnitude of CHD burden is like tip of iceberg being considerable but largely unrecognized and underestimated. However, recent advances in the treatment of CHDs from developed countries and their encouraging results should prompt clinicians to take up the challenge of managing these complex problems. Given a decline in contribution of infectious and nutritional diseases to infant mortality, CHDs are likely to be important contributor in future. Hence, exact prevalence of CHD is to be determined, so that appropriate changes in health policies can be recommended [31].

Study Limitations

This is a single centre study. Patients referred to the cardiology department for 2D echocardiography were only evaluated. So, true prevalence of CHD cannot be ascertained with this study. A larger population-based study is required.

Conclusion

The frequency of CHD at a tertiary care centre in western India was 23.75 percent. VSD and ASD were the most common acyanotic congenital heart defect while TOF was the commonest cyanotic congenital heart defect observed. TTE plays an important role in the diagnosis of CHD. Early diagnosis and treatment of CHD is the best approach to minimize the morbidity and mortality. Hence, when CHD is suspected, an echocardiography should be performed.

Conflict of Interest

No conflict of interest.

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