

## Original Research Article

# Gamut of congenital heart diseases in a tertiary center in South India: an ode to echocardiography

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### ABSTRACT

**Background:** Echocardiography has supplanted clinical acumen in diagnosis of congenital heart diseases (CHDs). Prevalence rates of CHDs across various regions of the world are subject to change over the course of time, with increasing use of this diagnostic modality. Objective: To assess the prevalence and types of CHDs.

**Methods:** The study was conducted at a tertiary care center in South Karnataka, India. Transthoracic echocardiographic records of all patients suspected to have congenital heart disease, over a period of 60 months were analyzed. Categorization of data into acyanotic and cyanotic congenital heart disease, and further, into different types was done after an exhaustive search. Specific variables such as age, frequency and gender distribution of all kinds of CHDs were computed.

**Results:** Of a total 112,372 pediatric patients who attended our center, 1451 reports of subjects suspected to have CHDs were analyzed. The prevalence was found to be 6.22 per 1000 subjects. Of the 700 subjects (48.24%) with CHD, 664 (94.85%) were diagnosed to have Acyanotic Congenital Heart Disease and 36 (5.14%) were diagnosed to have Cyanotic Congenital Heart Disease. Among the Acyanotic CHD, Atrial Septal Defect (ASD) was found to be the most common (40.21%) seconded by Ventricular Septal Defect (VSD) (21.53%). Among the 36 subjects diagnosed to have Cyanotic CHD, it was found that Tetralogy of Fallot (TOF) was the most commonest lesion (61.11%).

**Conclusions:** Increased utilization of Echocardiography as a diagnostic modality significantly helps to better appreciate ever varying prevalence rates and types of CHDs in different parts of India. Frequent longitudinal studies in this regard help in enhanced allocation of available resources and updating of available databases.

**Keywords:** Acyanotic, Congenital heart disease, Cyanotic, Echocardiography

### INTRODUCTION

Congenital heart disease (CHD) is defined as any structural defect of the heart or the intrathoracic great vessels, that may be responsible for an immediate, or gradual functional consequence.<sup>1</sup> CHDs are one of the notable causes of infant mortality. In approximately 90% of the CHD cases, there may be no identifiable cause. More commonly, asymptomatic subjects may be

identified during routine examinations.<sup>2</sup> The overall incidence and prevalence of CHD has been found to vary worldwide, and these differences may be attributable to diversity in genetics, environmental and cultural factors. In Asia, the estimated prevalence of congenital heart diseases has been noted to be 9.3/1000 live births with relatively higher prevalence observed among subjects with pulmonary outflow obstructions, as compared with those with left ventricular outflow tract obstructions. This

has been found to be higher in comparison to prevalence rates noted across both Europe and North America (8.2/1000 live births and 6.9/100 live births respectively).<sup>3</sup> Congenital heart disease has been classified as per Merck Manual of diagnosis broadly into Acyanotic and Cyanotic CHD. Atrial Septal Defect (ASD), Ventricular Septal Defect (VSD), Tetralogy of Fallot (TOF), Patent Ductus Arteriosus (PDA), Pulmonary Stenosis (PS), Aortic Stenosis (AS), Coarctation of Aorta (COA) and Atrioventricular Septal Defect (AVSD) totally constitute 85% of all CHDs.<sup>4</sup>

The remainder of 15% of CHDs is accounted for by rare and complex defects such as Truncus Arteriosus (TA), Tricuspid Atresia (TA), Total Anomalous Pulmonary Venous Connection (TAPVC), Hypoplastic Left Heart Syndrome (HLHS), Double Outlet Right Ventricle (DORV), Single Ventricle (SV), Ebstein Anomaly (EA) and Dextrocardia. Over the last two decades, echocardiography has proved to be the most important diagnostic modality in identification of CHDs, sometimes even supplanting clinical acumen. It stands to reason then, that with the increasing utilization of echocardiography, our understanding and knowledge of prevalence rates and types of CHDs are subject to change, more so in the developing world, empowering the need for further studies in this regard.

## METHODS

The study was conducted at K. S. Hegde Hospital, a tertiary care center in Mangalore, Karnataka. In this retrospective analysis, transthoracic echocardiographic

records of all patients suspected to have congenital heart disease, over a period of 60 months (January 2011-January 2016) were analyzed. All in-patient and out-patient data of subjects suspected to have CHD were extensively reviewed. This also included records of live births in the center. The diagnosis of CHD was done as defined by Mitchell et al that is, any gross structural aberration of the heart or intrathoracic great vessels that is actually or potentially of functional consequence, but discounting the systemic great arteries and veins.<sup>1</sup> Records of subjects who presented with signs and symptoms such as shortness of breath, feeding difficulties and excessive forehead sweats, bluish discoloration of lips and tongue, recurrent respiratory tract infections, failure to thrive and syncopal episodes were studied.

Furthermore, the records of subjects who were found to present with clubbing, cyanosis, pedal edema and abdominal distension, effortless tachypnea, abnormal blood pressure for age, heart murmur and who were subject to chest X-rays, electrocardiogram (ECG) and echocardiography were reviewed. CHDs associated with syndromes and acquired heart disease constituted the exclusion criteria. Echocardiography was performed by a cardiologist and as per standards laid down by the American Society of Echocardiography, using M-mode, two-dimensional and color Doppler, pulse and continuous wave echocardiogram.<sup>5</sup> Categorization of data into acyanotic and cyanotic congenital heart disease, and furthermore, into different types was done after an exhaustive search. Specific variables such as age, frequency and gender distribution of all kinds of CHDs were computed.

**Table 1: Spectrum of acyanotic congenital heart disease.**

CHD Type	≤ 1 month	1 month-1 year	1-5 years	5-12 years	> 12years	Total
ASD	103	62	43	48	11	267 (40.21%)
VSD	10	30	50	38	15	143 (21.53%)
PDA	27	27	22	15	05	96 (14.45%)
ECD	06	01	03	01	01	12 (1.81%)
ASD+VSD	08	08	04	03	-	23 (3.46%)
ASD+PDA	08	02	02	01	-	13 (1.96%)
ASD+VSD+PDA	03	02	01	-	-	06 (0.9%)
An. IAS	01	04	-	01	01	07 (1.05%)
Bicuspid AV	-	-	02	04	02	08 (1.20%)
PAPVC	-	02	-	03	-	05 (0.75%)
COA	-	-	-	02	-	02 (0.30%)
Cardiomyopathy	-	02	01	01	-	04 (0.60%)
VSD+PS	-	01	-	01	01	03 (0.45%)
VSD+AR	-	-	-	-	01	01 (0.15%)
Dextrocardia	-	02	-	04	02	08 (1.20%)
AS	-	-	01	04	01	06 (0.9%)
PS	-	03	06	05	01	15 (2.26%)
MR	-	03	-	-	-	03 (0.45%)
MVP	02	01	-	20	19	42 (6.33%)

ASD-Atrial Septal Defect, VSD-Ventricular Septal Defect, PDA-Patent Ductus Arteriosus, ECD-Endocardial cushion defect, An. IAS-Aneurysmal interatrial septum, AV-atrioventricular valve, PAPVC-Partial anomalous pulmonary venous return, COA-Coarctation of aorta, PS-Pulmonary stenosis, AR-Aortic regurgitation, AS- Aortic stenosis, MR-Mitral regurgitation, MVP-Mitral valve prolapse

**Table 2: Spectrum of cyanotic congenital heart diseases.**

CHD Type	≤ 1 month	1 month-1 year	1-5 years	5-12 years	>12 years	Total
TOF	03	09	06	03	01	22 (61.11%)
TGA	-	-	-	-	01	01 (2.78%)
TAPVC	-	01	-	01	01	03 (8.34%)
TA	-	-	-	01	-	01 (2.78%)
SV	01	-	-	-	-	01 (2.78%)
HLHS	01	-	-	-	-	01 (2.78%)
EA	01	-	01	-	01	03 (8.34%)
DORV	-	01	-	01	-	02 (5.56%)
Dextrocardia with TOF	01	-	-	-	-	01 (2.78%)
Dextrocardia with DORV	-	-	-	01	-	01 (2.78%)

TOF-Tetralogy of Fallot, TGA-Transposition of Great arteries, TAPVC-Total anomalous pulmonary venous return,TA-Tricuspid atresia, SV-Single ventricle, HLHS- Hypoplastic left heart syndrome, EA-Ebstein anomaly, DORV-Double outlet right ventricle.

## RESULTS

A total of 112,372 pediatric patients attended our center during the study period. Among these,1451 reports of subjects suspected to have CHDs were analyzed. Of these, 673 were purported to be normal, whereas 78 reports were reported as Patent Foramen Ovale, and hence considered insignificant.

Of the remaining 700 subjects (48.24%), 664 (94.85%) were diagnosed to have Acyanotic Congenital Heart Disease and 36 (5.14%) were diagnosed to have Cyanotic Congenital Heart Disease. Of the 664 Acyanotic CHDs, 332 (50%) were male subjects and 332 (50%) were female subjects, reflecting an equal gender predilection. Of the 36 Cyanotic CHDs, 21 (58.33%) were male and 15 (41.67%) were female. Among the 664 subjects diagnosed to have Acyanotic CHD, Atrial Septal Defect (ASD) was found to be the most common (n=267, 40.21%) seconded by Ventricular Septal Defect (VSD) (n=143, 21.53%) and followed closely by Patent Ductus Arteriosus (PDA) (n=96, 14.45%). It was also observed that 8 subjects were diagnosed to have Dextrocardia, of which 3 were found to have an associated VSD and 3 had a concomitant situs inversus.

The various Acyanotic CHDs observed and their age distribution is as depicted in Table 1. Among the 36 subjects diagnosed to have Cyanotic CHD, it was found that Tetralogy of Fallot (TOF) represented the commonest lesion (n=22, 61.11%), followed by equal distribution of Ebstein anomaly (EA) and Total Anomalous Pulmonary Venous Connection (TAPVC) (n=3, 8.34%). An interesting observation was the occurrence of dextrocardia with 2 different cyanotic lesions, namely Double Outlet Right Ventricle (DORV) and TOF. The various Cyanotic CHDs observed and their age distribution is as depicted in Table 2. The gender-wise distribution of acyanotic and cyanotic CHDs are as depicted in Table 3 and Table 4 respectively.

**Table 3: Gender-wise distribution of acyanotic congenital heart diseases.**

Congenital heart disease	Male	Female
ASD	115 (43.07%)	152 (56.93%)
VSD	95 (66.43%)	48 (33.57%)
PDA	41 (42.7%)	55 (57.3%)
ECD	05 (41.67%)	07 (58.33%)
ASD+VSD	12 (52.17%)	11 (47.83%)
ASD+PDA	05 (38.47%)	08 (61.53%)
ASD+VSD+PDA	04 (66.67%)	02 (33.33%)
An. IAS	03 (42.86%)	04 (57.14%)
Bicuspid AV	04 (50%)	04 (50%)
PAPVC	05 (100%)	-
COA	02 (100%)	-
Cardiomyopathy	01 (25%)	03 (75%)
VSD+PS	01 (33.33%)	02 (66.67%)
VSD+AR	01 (100%)	-
Dextrocardia	05 (62.5%)	03 (37.5%)
AS	04 (66.67%)	02 (33.33%)
PS	09 (60%)	06 (40%)
MR	-	03 (100%)
MVP	20 (47.61%)	22 (52.39%)

**Table 4: Gender-wise distribution of cyanotic congenital heart diseases.**

CHD	Male	Female
TOF	13 (59.09%)	09 (40.91%)
TGA	-	01 (100%)
TAPVC	02 (66.67%)	01 (33.33%)
TA	01 (100%)	-
SV	01 (100%)	-
HLHS	-	01 (100%)
EA	01 (33.33%)	02 (66.67%)
DORV	02 (100%)	-
Dextrocardia with TOF	-	01 (100%)
Dextrocardia with DORV	01 (100%)	-

## DISCUSSION

Congenital Heart Diseases account for significant morbidity and mortality. Diagnostic modalities such as Echocardiography have provided increased insight into the ever-growing prevalence of CHDs, especially in the developing world. The prevalence rates noted across the western world currently, both in Europe and North America are 8.2/1000 live births and 6.9/100 live births respectively.<sup>3</sup>

This differs from earlier reports describing prevalence as being 5.51 in Europe and 6.6 in North America, highlighting the obvious importance of echocardiography in improving surveillance.<sup>6,7</sup> The different prevalence rates in various geographical zones are as shown in Table 5.<sup>8-14</sup>

**Table 5: Prevalence rates of CHDs in different geographical zones.**

Country	Year	Frequency
UK	1981	5.51
USA	1990	6.6
Hong-Kong	1991	6.35
Norway	1994	10.6
Pakistan	1997	4
Saudi Arabia	2001	10.7
South Africa	1979	3.9
Spain	2005	8.96
Iran	2007	8.6

**Table 6: Prevalence of CHDs in various regions across India.**

Regions (study population)	Period	Prevalence rates (per 1000)
Punjab	1962	31
South India	1968	25.6
J and K	1979	9.7
Shimla	1995	2.25
New Delhi	2001	4.2
J and K	2014	1.12
Mysore	2006	6.6-13.06
Present study	2016	6.22

The average prevalence of CHDs in various regions of India has been purported to be 14.7 per thousand live births.<sup>15</sup> The relative prevalence in specific zones and trends over the last five decades is as depicted in Table 6.<sup>3,16-21</sup>

It is worthwhile noting that attempts have been made to estimate the prevalence of CHDs in various population groups over the past decades, which has been supported with echocardiographic evidence. Our study encompassed all children up to the age of 18 years, which included live born subjects and those referred from outlying centers.

Our hospital based study delivered a prevalence rate of 6.22 per 1000, which is comparable to prevalence rates noted elsewhere, although it must be reiterated that only one study, by Khalil et al identifies the prevalence among live births to be 3.9 per thousand.<sup>21,22</sup>

**Table 7: Profile of CHD in India.**

Author	Year	Profile of selected CHD in India (expressed as percentage)								
		ASD	VSD	PDA	ECD	PS	TOF	TGA	TA	COA
Shrestha et al	1980	23	30	11	-	-	04	-	-	-
Kinare et al	1981		04	09	-	-	12	12	-	08
Vashishtha et al	1993	11	41	04	-	-	14	-	-	-
Thakur et al	1995	38	32	-	-	-	-	-	-	-
Gowda et al	2006	11.4	26.3	10.9	-	-	6.4	0.4	-	-
Kapoor	2008	18.9	21.3	14.6	-	03	4.6	1.1	-	-
Misra et al	2009	6.7	28.3	8.7	3.5	9.5	6.8	3.4	1.2	4.2
Haleja	2014	26	58	10	2	2	52	18	04	-
Jatav et al	2014	18.1	28.4	10.3	-	6.8	6.0	4.3	-	1.7
Wanni et al	2014	22.9	31.4	23.9	4.1	7.3	48	27	02	0.1
Present study	2016	40.2	21.5	14.4	1.8	2.2	61.1	2.7	2.7	0.3

The present study observed that among acyanotic CHDs, ASD (40.21%) was the commonest lesion, followed by VSD (21.53%) and PDA (14.45%). This finding did not corroborate with any of the previous literature cited (Table 7), all of which favored greater involvement of

VSD in their studies, except for Thakur et al who observed greater prevalence of ASD.<sup>3,15,19,21,23-28</sup>

Also of relevance is the observation of interesting findings such as PAPVC (0.75%) and aneurysmal IAS that has not been noted previously, even among recent

cited literature such as those of Gowda et al and Wannan et al.<sup>3,21</sup> Among the cyanotic lesions, the most common lesion was TOF (61.11%). It was also observed that frequency of ASD was much higher compared to previous studies, whereas the frequencies of VSD and PDA favored comparably with the same. The prevalence of TOF in particular, was manifold in comparison with other reports and may be attributable to the fact that our hospital is a reputed tertiary referral center. The profile of CHDs in various Indian literatures is as depicted in Table 7.

It is worthwhile noting that 31.78% of acyanotic CHDs and 30.56% of cyanotic CHDs were diagnosed beyond the age of 5 years, which still raises questions with regard to accessibility of health services such as echocardiography in our populace. Previously, sharing of infrastructure with adult cardiology units and utilization of their resources where feasible has been mooted, as in a country like India, pediatric cardiology units remain a luxury. Optimal utilization of said infrastructure is also inclusive of creation and updating of databases catering to congenital heart diseases.

With advent of newer technology, understanding of various CHDs as well as their prevalence is subject to change. This study stresses on the requirement of longitudinal surveillance at frequent intervals, with focus on identification of prevalence of various CHDs in our population, which would definitely go a long way in optimal resource allocation and utilization, in a country where such resources are at a premium.

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