

# Prevalence and Pattern of Congenital Heart Disease in Pediatric Population—A Study from Central India

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Abstract **Introduction** Prevalence of congenital heart disease (CHD) is variable depending on the region and setting of study ranging from 0.8 to 6/1,000. Age of presentation depends on types of CHD and its severity. Burden of CHD is not known in this tribal belt of Central India. Main objective of this study was to find out prevalence of CHD in children. We have also studied the pattern and age of presentation of various types of CHDs. Methods This retrospective observational study was conducted at tertiary care teaching institute in tribal belt of Central India. Children aged 0 to 15 years reported to pediatric outpatient department (OPD) were included. CHD was confirmed by echocardiography in suspected cases. Prevalence rate was calculated as number of CHDs per 1,000 OPD patients. Pattern of CHD was categorized as per standard guidelines and age-wise presentations of various types of CHD were studied. **Results** The prevalence rate of CHD in our study population was 27.7/1,000, which is **Keywords** high when compared with most of the other hospital-based studies. Most of the ► congenital heart patients, 60.36 and 83.26% were detected before the age of 1 year and 5 years, disease respectively. All critical CHD cases were detected in early infancy. India Conclusion There is a high burden of CHD seen in this study. Possible cause of this prevalence may be lack of specialized facility in this reason and study period included first echocardiography and second wave of COVID. Further, large sample size studies and/or nationwide registry/database are needed to know the exact burden of CHD. pattern

# Introduction

Congenital heart disease (CHD) is any gross structural abnormality of the heart that is present since birth.<sup>1</sup> The

article published online July 14, 2022 DOI https://doi.org/ 10.1055/s-0042-1751085. ISSN 2455-7420. reported prevalence of CHD in India is 8 to 10 out of every 1,000 live births.<sup>2</sup> CHD accounts for approximately one-fourth of all congenital malformations and is a cause for 10% of the present infant mortality.<sup>3</sup> Prevalence of CHD is not

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uniform in different regions and settings of world. In community-based studies from India, the prevalence of CHD ranges from 0.8 to 5.2 out of every 1,000 patients<sup>4,5</sup> while hospital-based studies had reported a wide variation in prevalence of CHD ranging from 2.25 to 26 out of every 1,000.<sup>6,7</sup> CHDs are grossly classified into acyanotic, cyanotic, and obstructive heart diseases, former being more common. Ventricular septal defect (VSD) (30-35%) and tetralogy of Fallot (TOF) (5–7%) are most common among acyanotic and cyanotic CHDs, respectively.<sup>8</sup> Age of presentation and clinical findings vary as per types and severity of CHD. Despite various studies conducted in India, burden and pattern of CHD in this tribal belt of Central India are lacking. So we planned this study to assess prevalence, types, and age-specific distribution of CHD among children in this region.

#### **Materials and Methods**

This retrospective observational study was conducted at tertiary care center in tribal belt of Central India from February 2020 to September 2021. Aim of study was to detect the burden of CHD in children of this region. Patients aged 0 to 15 years of age, who reported to pediatric and pediatric cardiology outpatient department (OPD) were included. Patients having signs and symptoms related to CHD were evaluated and subjected for chest X-ray and electrocardiogram. Transthoracic echocardiogram (TTE) was performed to confirm the diagnosis of CHD. TTE was performed by pediatric cardiologist using GE Vivid E95 echo Machine (Wipro GE Healthcare Pvt Ltd, India) with pediatric and neonatal probe using two dimensional, color doppler, Mmode, pulse and continuous wave echocardiogram.<sup>9</sup> We excluded all patent foramen ovale (PFO) and patent ductus arteriosus (PDA) of <2 mm in less than 1 month of age as spontaneous anatomical closure of majority of insignificant defects like PFO and majority of PDAs in preterm and term infants occurs by 44-weeks postmenstrual age or 2 to 3 weeks of life.<sup>10-12</sup> Bicuspid aortic valve (BAV) is considered most common type of CHD with an incidence of 1.3% of total population.<sup>13</sup> To avoid overestimation of CHD prevalence, we have also excluded uncomplicated/isolated BAV.

Primary objective was to find the prevalence of CHD per 1,000 OPD cases of similar age group and secondary objective was to see the age-wise distribution of various types of CHD. The CHDs were grossly classified into acyanotic, cyanotic, and obstructive lesions. These defects were further categorized according to the International Pediatric and Congenital Cardiac Code (IPCCC) and the 11th iteration of the International Classification of Diseases (ICD-11).<sup>14,15</sup> Ethical approval was obtained from the Institutional Ethics Committee. Informed consents were obtained from the participant's guardian prior to study enrollment.

Statistical analysis: Data were collected and compiled into MS Excel spread sheets. Continuous variables were summarized as mean and standard deviation and categorical variables were summarized as proportion (%).

# **Observations and Results**

A total of 17,921 patients aged 0 to 15 years attended our pediatric and pediatric cardiology department during study period. A total of 843 patients were detected to have CHD. We have excluded 346 patients (PDA-106, PFO-183, and BAV-57) as per the exclusion criteria and 497 patients were included. Male to female ratio was 1.26:1 (277:220). Median age of children with CHD was 9 months and most of the patients, i.e., 300 (60.4%) out of 497 were less than 12 months of age, while the number of cases in age group 6 to 10 years and 11 to 15 years were almost same. Prevalence of CHD in children came out to be 27.7 per 1,000 study population. Major classification and types of CHD in present study are summarized in **-Table 1**. Acyanotic CHDs (n = 316) were about twothirds and cyanotic about one-fourth of all CHD. Together atrial septal defects (ASDs) and VSDs constituted about half of all CHDs. TOF was the most common (43%) and total anomalous pulmonary venous return was the second most common type of cyanotic CHD. Patients with isolated aortic stenosis and isolated pulmonary stenosis were almost equal in number and constituted more than 60% of total obstructive lesions. Various subtypes of acyanotic, cyanotic, and obstructive CHD are shown in diagrammatic presentation in **► Fig. 1**.

Age-wise distribution of various CHDs is depicted in **- Table 2**. In comparison to acyanotic and cyanotic CHDs the proportion of patients with obstructive CHD was less in infants. In all the groups of CHDs there was a gradual decline in number of cases with increasing age.

## Discussion

The prevalence of CHD varies in different region of the world and in different settings. As per study from China, prevalence of CHD was 9.3 out of every 1,000 live births<sup>16</sup> and Fatema et al<sup>17</sup> got a prevalence rate of 25 per 1,000 live birth in Bangladesh. Dixit et al<sup>2</sup> in their study had summarized the overall prevalence estimate of CHD from various other studies. They found prevalence ranging from 0.77 to 5.2 per 1,000 in school-based studies and a range of 3.9 to 26.4 per 1,000 in hospital-based studies. In their own study, Dixit et al<sup>2</sup> have found a prevalence of 19.4 out of every 1,000 but this might have underestimated as they had included adults in their study, as prevalence of CHD in adult is low compared with children. We found a prevalence of 27.7 out of every 1,000 study population. Most of the other studies reporting prevalence of CHD have included either newborns or school children. Studies including newborn overestimate the prevalence rate by including small ASD or PDA while school-based studies exclude a large number of children of pre-school age group. Hoffman et al<sup>18</sup> in his study stratified the major form of CHD into trivial, moderate, and severe categories and found a variation in incidence ranging from 6 to 75 out of every 1,000 based on inclusion of insignificant CHD (small VSD, PDA, and PFO) and how early the diagnosis was made. We have followed strict exclusion criteria by excluding small PDA in early age, isolated BAV, and PFO to

Table 1 Major classification of CHD and types of CHD

Major classification	Types of CHD	Subtypes	
Acyanotic (Left to right shunt) lesions $(n = 316; 63.6\%)$	VSD (n = 138; 27.7%)	Membranous	95 (68.8%)
		Muscular	26 (18.8%)
		Inlet	9 (6.5%)
		Outlet	8 (5.8%)
	ASD (n = 98; 19.7%)	OS ASD	85 (86.7%)
		SV ASD	8 (8.16%)
		OP ASD	5 (5.1%)
	PDA		67 (13.48%)
	AVSD		10 (2%)
	AP window		3 (0.6%)
Cyanotic lesions (n = 127; 25.5%)	TOF (n = 55; 11.06%)	PS	33 (60%)
		PA	17 (30.9%)
		Absent PV syndrome	5 (9.1%)
	TAPVC ( <i>n</i> = 19; 3.82%)	Supracardiac	8 (42.1%)
		Mixed	5 (26.3%)
		Cardiac	4 (21%)
		Infracardiac	2 (10.5%)
	Single ventricle physiology $(n = 12, 2.4\%)$	Increased pulmonary blood flow	7 (58.3%)
		Decreased pulmonary blood flow	5 (41.7%)
	Tricuspid atresia $(n = 10, 2\%)$	ΙA	2 (20%)
		1B	5 (50%)
		1C	3 (30%)
	TGA		12 (2.4%)
	DORV + PS		6 (1.2%)
	HLHS		5 (1%)
	Truncus arteriosus		4 (0.8%)
	PA + Intact IVS		3 (0.6%)
	CCTGA + VSD + PS		1 (0.2%)
Left or right side obstructive lesions (n = 48; 9.6%)	AS(BAV)		16 (3.2%)
	PS		15 (3%)
	Coarctation of aorta		7 (1.4%)
	Shone's disease		4 (0.8%)
	Peripheral PS		4 (0.8%)
	Subaortic membrane		2 (0.4%)
Others ( <i>n</i> = 6; 1.2%)	Ebstein		4 (0.8%)
	Situs inversus totalis		2 (0.4%)

Abbreviations: AP window, aortopulmonary window; AS, aortic stenosis; ASD, atrial septal defect; AVSD, atrioventricular septal defect; BAV, bicuspid aortic valve; CCTGA, congenitally corrected TGA; DORV, double outlet right ventricle; HLHS, hypoplastic left heart syndrome; IVS, interventricular septum; OP, ostium primum; OS, ostium secundum; PA, pulmonary atresia; PDA, patent ductus arteriosus; PS, pulmonary stenosis; PV, pulmonary valve; SV, sinus venosus; TAPVC, total anomalous pulmonary venous return; TGA, transposition of great arteries; TOF, tetralogy of Fallot; VSD, ventricular septal defect.

Note: Shone's disease: Left heart obstruction at multiple places.

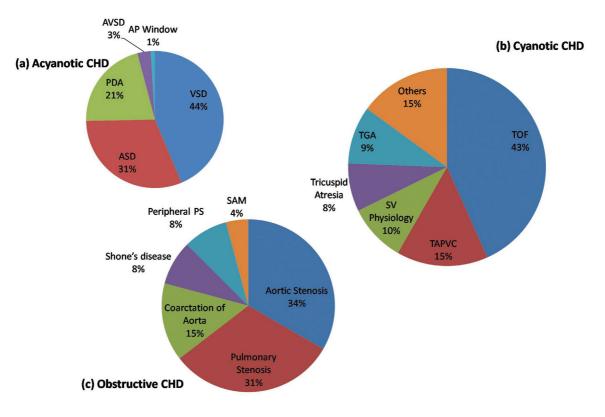


Fig. 1 Diagrammatic presentation of various subtypes of (a) acyanotic CHD, (b) cyanotic CHD, and (c) obstructive CHD. CHD, congenital heart disease.

avoid overestimation of CHD prevalence. Despite following strict exclusion criteria, we have observed highest prevalence (27.7/1,000) when compared with similar types of study. There were two major COVID outbreaks during our study period. This might have led to only reporting of patients with significant symptoms/morbidity and thus overestimating the prevalence rate. Another cause of high burden of CHD in this region might be poor availability of specialized facility, so majority of cases were reported at our center.

In this study, 276 (55.5%) children were male with male to female ratio of 1.24. Qazi and Saqib<sup>19</sup> in their study found 60.3% male patients with CHD while Wanni et al<sup>20</sup> and Chatterjee et al<sup>21</sup> in 52.4 and 51.9% cases, respectively. In our study we had categorized the CHD in acyanotic, cyanotic, and obstructive lesions. Most of the previous studies have included obstructive lesions into acyanotic CHD and reported acyanotic CHD ranging from 66.7 to 88.7%.<sup>7,22,23</sup> More than half of the patients (63.6%) in our study had acyanotic CHD, followed by cyanotic (25.5%) and obstructive (9.6%) lesions. The pattern of classification of CHD and prevalence rate in our study is seconded by the study of Dixit et al<sup>2</sup> and Reller et al.<sup>15</sup> Although Dixit et al had included adult also in their study but the prevalence of acyanotic, cyanotic, and obstructive lesions was same in both pediatric and adult age.<sup>2</sup>

Most common acyanotic CHD in our study was VSD (27.7%). Similarly, other reports like Meshram and Gajimwar (20.7%), Kapoor and Gupta et al (21.3%), and Wanni et al

(31.9%) reported VSD as the most common CHD.<sup>7,22,23</sup> Second most common CHD in our study was ASD (19.7%) similar to study by Meshram and Gajimwar.<sup>23</sup> But Naik et al<sup>20</sup> and Wanni et al<sup>22</sup> reported PDA as the second most common CHD followed by ASD. PDA was the third most common CHD in our study. We found atrioventricular septal defect in 2% patients similar to Naik et al<sup>22</sup> and Meshram et al<sup>23</sup>. As aortopulmonary (AP) window is a very rare CHD, none of the other similar studies reported prevalence rate of AP window.

The age of presentation of CHD depends on the hemodynamic effect of the heart defect. More than half (300/497, 60.4%) of all the CHD patients in our study had presented in first year of life. Our finding is consistent with the study of Meshram and Gajimwar<sup>23</sup> (56.28%) while Mahapatra et al<sup>24</sup> (40.25%) and Jatav et al<sup>25</sup> (37.06%) have reported lesser number of cases in this age group. About 25% (114/497) were diagnosed in 1 to 5 years of age. Large VSD, PDA, AP window and other most of the cyanotic CHD cases become symptomatic in early infancy. This explains high number of these cases in early infancy. ASD usually presents later in life but in our study ASD cases were also detected in infancy. We could not found any reason for this. All critical duct-dependent lesions like hypoplastic left heart syndrome and pulmonary atresia with intact interventricular septum and most of the cyanotic lesions except TOF were detected before 1 year of age. Surgical correction of CHD in these patients is needed in early life. Unlike shunt lesions the age of presentation of obstructive lesions is usually late. We had found significant proportion (45.8%) of obstructive lesions in

Major classification	Type of CHD	0–1 Y	1–5 Y	6–10 Y	11–15 Y	Total
Acyanotic CHD	VSD	84	39	9	6	138
	ASD	58	24	8	8	98
	PDA	43	15	6	3	67
	AVSD	7	2	0	1	10
	AP window	2	0	1	0	3
	Total	194 (61.4%	80 (25.3%)	24 (7.6%)	18 (5.7%)	316
Cyanotic CHD	TOF	27	15	5	8	55
	TAPVC	12	5	2	0	19
	Single ventricle	8	1	1	2	12
	Tricuspid atresia	8	2	0	0	10
	TGA	8	2	1	1	12
	DORV + PS	4	1	1	0	6
	HLHS	5	0	0	0	5
	Truncus arteriosus	4	0	0	0	4
	PA +Intact IVS	3	0	0	0	3
	CCTGA + VSD + PS	0	0	0	1	1
	Total	79 (62.2%)	26 (20.5%)	10 (7.9%)	12 (9.4%)	127
Obstructive CHD	AS	8	3	3	2	16
	PS	5	1	4	5	15
	Coarctation	3	2	1	1	7
	Shone's disease	3	1	0	0	4
	Peripheral PS	3	0	1	0	4
	Subaortic membrane	0	1	0	1	2
	Total	22 (45.8%)	8 (16.6%)	9 (18.7%)	9 (18.7%)	48
Others	Ebstein	3	0	0	1	4
	Situs inversus totalis	2	0	0	0	2
	Total	5 (83.3%)	0	0	1 (16.6%)	6
	Grand total	300(60.4%)	114 (22.9%)	43 (8.6%)	40 (8.0%)	497

Table 2 Age-wise distribution of various types of CHD

Abbreviations: AP window, aortopulmonary window; AS, aortic stenosis; ASD, atrial septal defect; AVSD, atrioventricular septal defect; BAV, bicuspid aortic valve; CCTGA, congenitally corrected TGA; DORV, double outlet right ventricle; HLHS, hypoplastic left heart syndrome; PA, pulmonary atresia; PDA, patent ductus arteriosus; PS, pulmonary stenosis; TAPVC, total anomalous pulmonary venous return; TGA, transposition of great arteries; TOF, tetralogy of Fallot; VSD, ventricular septal defect.

patients less than 1 year of age. This can be explained by incidental detection of these CHDs by routine clinical examination even in the absence of symptoms. Almost half of the obstructive CHD patients were under 5 years of age. This is consistent with the study reported by Dixit et al<sup>2</sup> and Jatav et al,<sup>25</sup> who found 50% of obstructive CHDs in patients less than 5 years of age.

None of the previous studies reporting the prevalence of CHD in India had subclassified it further. In our study, we have further subclassified the types of CHD. Isolated VSD was seen in 82% while in other 18% cases VSD was associated with ASD and/or PDA.

Among cyanotic CHD, TOF (11%) was the most common CHD. Various studies have reported the prevalence of TOF ranging from 5.5 to 21%.<sup>20–23</sup> Transposition of great arteries (TGAs) was the second most common cyanotic CHD in reports by Naik et al<sup>22</sup> and Meshram et al.<sup>23</sup> In our study TPAVC was the second common cyanotic CHD followed by TGA and single ventricle physiology.

### Conclusion

We have observed high burden of CHD in this region compared with other hospital-based studies. VSD was the most common acyanotic CHD while TOF was the commonest cyanotic CHD. More than half of the patients had presented in their first year of life. Study was conducted in small sample size and study period included the COVID outbreak, these are the limitations of our study. Further studies including large sample size in non-COVID era are needed to estimate the true prevalence of CHD in our region. Nationwide registry/database will help to know the exact burden of CHD in different regions of India.

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Conflict of Interest None declared.

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