Original Research Article

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Pattern of congenital heart diseases among children attending a pediatric cardiology OPD of a tertiary level hospital in West Bengal, India: a retrospective study

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ABSTRACT

Background: Congenital heart disease (CHD) is one of the most common congenital defects worldwide. The prevalence of CHD is low in India mostly due to under detection. The knowledge regarding socio-demographic factors, other congenital anomalies/syndromes and social impacts associated with CHDs in India is grossly lacking. **Methods:** This was a retrospective study. Details of all diagnosed pediatric (age 0-14 years) cases of CHD were recorded by a single cardiologist from the year 2013 to 2020. Duplicate entries were removed and CHDs were classified according to Q20-Q28 of tenth revision of international classification of diseases (ICD). For description purposes, the CHDs were divided into 10 sub-groups.

Results: Total 9247 cases of CHDs were recorded during the study period. Ventricular septal defect was the most commonly identified CHD (32.1%). Most cases were acyanotic (69.8%) and presented first time at age of more than two years. Almost 1.5% cases were associated with other non-cardiac congenital defect or syndrome. In 1.0% of cases either mother or the child faced social or family problem due to the disease.

Conclusions: CHD is an emerging public health problem. It is associated with other congenital anomalies/syndrome. Mandatory screening for birth defects needs to be initiated at all birth facilities. At birth counseling of the parents need to be implemented due to the associated social issues.

Keywords: Congenital heart diseases, Congenital anomalies, Social impact

INTRODUCTION

CHDs are a rapidly emerging global health problem.¹ CHDs are defined as structural abnormalities of the heart and/or great vessels that are present at birth.² These are one of the most frequently diagnosed congenital disorders with a global prevalence of approximately 0.8-1.2%.^{3,4} In India, the reported prevalence of CHDs has been found to be ranged between 0.04-0.9% in various studies.^{5,6} Although the reported prevalence of CHDs in our country seems to have increased dramatically in recent years, the prevalence is assumed to be a gross underestimation of the actual burden as many of the cases remain undiagnosed at birth and thereafter for a long duration.⁷ Nonetheless, with 25 million births every year in India, the absolute number of children born every year with CHDs is large.

CHDs are associated with high morbidity and mortality. In the latest report, it has been found that in 2017 CHDs were associated with almost 0.2 million deaths among infants globally.⁸ The deaths associated with CHD are relatively higher in low and middle-income countries (LMIC) than the developed countries.⁹ It has also been found that many times either the patient or the mother faces multiple social problems.¹⁰ However, very few studies have been done so far in the eastern region of India to evaluate various aspects of CHDs and factors or social issues associated with them.

To fill these gaps, the objectives of our study were to assess the pattern of CHDs attending pediatric cardiology OPD of a tertiary care hospital in West Bengal, India and to study the selected socio-demographic factors, other congenital malformations/syndromes, selected complications and social impacts associated with the CHDs.

METHODS

This was a retrospective study. We undertook a retrospective study of all cases of structural CHDs aged between 0-14 years and attended pediatric cardiology clinics organized by Healthworld hospitals, City Centre,

Durgapur in West Bengal, India. The patients attending the clinic were from West Bengal, Jharkhand and Bihar states of India. Patients from neighboring countries like Nepal and Bhutan also attended these clinics. One single cardiologist entered all the data in the prescribed format which is the routine procedure of the hospital. All new patients were provided with a registration number. The OPD cards of repeat patients were stamped only with the date of the repeat visit.

The data used for analysis in this study was restricted to new patients only who attended this clinic from January 2013 to December 2020. The treating doctor maintained a register where the diagnosis along with registration number, age and sex of the patient were recorded. In case of multiple morbidities, the primary diagnosis was recorded first. Only the primary diagnoses were included for analysis.

Data abstraction from the diagnosis register was carried out by the clinician himself. A preliminary check for duplication was performed using a combination of names and the patient's registration number and thus, the duplicate entries were identified and removed.

The diagnoses were confirmed by echocardiography and classified according to Q20-Q28 of the tenth revision of the ICD.¹¹ For description purposes, the CHDs were divided into 10 sub-groups, group 1-4 were cyanotic heart diseases whereas group 5-10 were acyanotic heart diseases (Table 1).

Table 1: Operational subgroups of CHDs.

Groups	Subgroups	Types
	1	TOF, TOF-PA, TETCANAL
Cronotia	2	Complex CHD, univentricular anatomy, tricuspid anomaly
Cyanotic	3	TAPVC
	4	TGA, ebstein, hemitruncus, truncus
	5	Pre tricuspid anomalies-ASD, ASD-P
	6	Post tricuspid anomalies-VSD, PDA, APW
A	7	Left obstructive-AS COA
Acyanotic	8	Right obstructive-PS
	9	Combined-AVSD, VSD-DCRV, VSD-SAM
	10	Others like CHB, MVP

The final data set, before its use for analysis was delinked from the name and registration number to anonymize the information of any personal identifier. Collection of information from all the patients visiting the pediatric cardiology clinic is being done routinely. Its purpose is to improve healthcare delivery in the future. Routinely collected service data were utilized for this study. The study adhered to the principles laid out as per the declaration of Helsinki. The data were entered in Microsoft excel, scrutinized for duplicates and cleaned. The final analysis was done in STATA version 12 (StataCorp College Station, Texas, USA). Categorical data were presented as percentages (%). Normally, distributed data were presented as means and standard deviation. Bi-variate analysis (Chi squared test or Fisher Exact test) was done for studying the association between selected socio-demographic and other variables. The p value <0.05 was considered statistically significant.

RESULTS

Type of CHDs

A total of 9247 children aged 0-14 years with CHDs attended the clinic from January 2013 to December 2020. The maximum number of the cases came with the presenting diagnosis of VSD (32.1%), followed by TOF (19.5%) and ASD (18.3%) (Table 2). Almost 2/3rd of the children (69.8%) had acyanotic CHDs (Table 3).

Socio-demographic factors and CHDs

The mean (SD) age of the children was 5.1 (4.6) years at the time of presentation at our clinic. The majority of the children were aged more than two years and only 16.3% of children presented within one month of their birth. The presenting mean age for cyanotic group was 5.4 years (SD=4.7) and for acyanotic group it was 5.2 years (SD=4.5).

The presenting age of the two group was not significantly different (p=0.099). The proportions of children with CHDs were almost equal in both the sexes; 50.9% were males and 49.2% were females. The proportion of males was higher among cyanotic CHDs, whereas, in acyanotic CHDs, the proportion of males and females were similar. This difference was statistically significant (p<0.001). In the majority of the cases, the maternal ages were between 20 to 30 years. There was no statistically significant difference when compared between cyanotic and acyanotic group in terms of maternal age (p=0.076). Out of the total children, 4/5th (79.2%) of them came from rural areas. The rural-urban difference in terms of cyanotic and acyanotic CHDs was found to be statistically significant (p=0.002). We also found that the proportion of cyanotic heart diseases was significantly higher in consanguineous marriages than in acyanotic heart diseases (p=0.004). However, the absolute number of CHDs associated with consanguineous marriage was less (Table 3).

Birth history and CHDs

Overall, 4.9% of the children were born preterm. We also found a statistically significant difference between cyanotic and acyanotic groups in terms of birth maturity (p=0.046). Acyanotic heart diseases were more common in children who were born preterm than those who were born at term. The majority was born in hospital settings (96.7%) and by normal vaginal delivery (57.5%). Only 0.5% of children have conceived via IVF induced pregnancy. Out of all the children, 11.3% cases had a maternal history of at least one miscarriage in previous pregnancies. There was no statistically significant difference between cyanotic and acyanotic CHDs in terms of place of birth, types of delivery, mode of pregnancy and history of miscarriage (Table 3).

Co-existing birth defects/syndromes

Non-cardiac birth deformities were found in 87 (0.9%) of the CHDs. Bone deformity (34.5%), cleft lip/palate (16.1%), malformation of gut (9.2%) and pre-auricular sinus (8.0%) were the major deformities (N=87). Various congenital syndromes were also found to coexist with 0.6% of CHDs. Down's syndrome was found to coexist in 75.4% of the CHDs. Congenital rubella syndrome, Turner syndrome, William syndrome were among the others.

Social issues and CHDs

A total of 103 (1.1%) cases of CHDs or their mother faced some sort of social issues due to the disease. Physical abuse and mental harassment of a mother by the spouse or other family members or dissolution of marriage were associated in 91 (1.0%) of the cases. The child was neglected in 12 of the cases.

Table 2:	Top ten	presenting	diagnosis	of the	CHDs.
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Sr. No.	Diamagic	Male	Female	Total	
	Diagnosis	Number (%)	Number (%)	Number (%)	
1	VSD	1,597 (53.8)	1,371 (46.2)	2968 (32.1)	
2	TOF	986 (54.7)	818 (45.3)	1804 (19.5)	
3	ASD	764 (45.1)	930 (54.9)	1694 (18.3)	
4	PDA	407 (40.1)	610 (59.9)	1017 (11.0)	
5	SV anatomy	219 (53.5)	190 (46.5)	409 (4.4)	
6	PS	75 (43.4)	98 (56.6)	173 (1.9)	
7	AVSD	86 (51.5)	81 (48.5)	167 (1.8)	
8	TAPVC	69 (57.1)	52 (42.9)	121 (1.3)	
9	AS	61 (68.5)	28 (31.5)	89 (0.9)	
10	TGA	49 (64.5)	27 (35.5)	76 (0.8)	

Table 3: Description of the different variables across different diagnosis subgroup categories.													
Diagnosis	Cyanotic hea	rt diseases (N	(=2794)			Acvanotic heat	rt diseases (N=6	(453)					Total
category	Group 1 (N=1937)	Group 2 (N=559)	Group 3 (N=121)	Group 4 (N=177)	Total (A)	Group 5 (N=1695)	Group 6 (N=3941)	Group 7 (N=187)	Group 8 (N=188)	Group 9 (N=301)	Group 10 (N=141)	Total (B)	(N=9247)
Age catego	ory of child (0: <	(1month;1: 1-	12 months; 2:	12 months to	5 years; 3: 5 ye	ears to 11 years;	4: > 11 years) (p value for cor	nparison betw	een cyanotic a	nd acyanotic g	group =0.099	
0	202 (10.4)	109 (19.5)	44 (36.5)	47 (26.5)	402 (14.4)	173 (10.2)	794 (20.2)	40 (21.4)	38 (20.2)	27 (8.9)	30 (21.3)	1102 (17.1)	1,504 (16.3)
1	290 (14.9)	63 (11.3)	16 (13.2)	18 (10.2)	387 (13.9)	112 (6.6)	572 (14.5)	16 (8.6)	24 (12.8)	37 (12.3)	13 (9.3)	774 (11.9)	1,161 (12.6)
2	617 (31.8)	159 (28.4)	23 (19.1)	49 (27.7)	848 (30.4)	488 (28.8)	1150 (29.2)	65 (34.7)	44 (23.4)	98 (32.6)	34 (24.1)	1879 (29.1)	2,727 (29.5)
3	520 (26.8)	142 (25.4)	25 (20.7)	46 (25.9)	733(26.2)	598 (35.3)	1,009 (25.6)	39 (20.8)	50 (26.6)	85 (28.2)	31 (21.9)	1812 (28.2)	2,545 (27.5)
4	308 (15.9)	86 (15.4)	13 (10.7)	17 (9.6)	424 (15.2)	324 (19.1)	416 (10.6)	27 (14.4)	32 (17.0)	54 (17.9)	33 (23.4)	886 (13.7)	1,310 (14.2)
Sex (0=female; 1=male) (p value for comparison between cyanotic and acyanotic group <0.001)													
0	881 (45.5)	256 (45.8)	52(42.9)	82 (46.3)	1271 (45.5)	930 (54.8)	1,961 (49.7)	69 (36.9)	103 (54.8)	140 (46.5)	68 (48.2)	3271 (50.7)	4,542 (49.1)
1	1,056 (54.5)	303 (54.2)	69 (57.1)	95 (53.7)	1523 (54.5)	765 (45.1)	1,980 (50.2)	118 (63.1)	85 (45.2)	161 (53.5)	73 (51.8)	3182 (49.3)	4,705 (50.9)
Maternal age category (0: <20 years; 1: 20-30 years; 2: >30 years) (p value for comparison between cyanotic and acyanotic group=0.076)													
0	597 (30.8)	195 (34.9)	46 (38.0)	62 (35.0)	900 (32.2)	561 (33.1)	1,352 (34.3)	69 (36.9)	75 (39.9)	94 (31.2)	46 (32.6)	2197 (34.1)	3,097 (33.5)
1	1,175 (60.7)	314 (56.2)	63 (52.1)	97 (54.8)	1649 (59.0)	1,000 (59.0)	2,301 (58.4)	102 (54.5)	97 (51.6)	178 (59.1)	84 (59.6)	3762 (58.3)	5,411 (58.5)
2	165 (8.5)	50 (8.9)	12 (9.9)	18 (10.2)	245 (8.8)	134 (7.9)	288 (7.3)	16 (8.6)	16 (8.5)	29 (9.6)	11 (7.8)	494 (7.7)	739 (7.9)
Area of rea	sidence (0=rural	l; 1=urban) (j	o value for con	nparison betw	een cyanotic a	nd acyanotic gro	oup=0.002)						
0	1,576 (81.4)	457 (81.7)	99 (81.8)	136 (76.8)	2268 (81.2)	1,292 (76.2)	3,132 (79.5)	140 (74.9)	134 (71.3)	247 (82.1)	108 (76.6)	5053 (78.3)	7,321 (79.2)
1	361 (18.6)	102 (18.3)	22 (18.2)	41 (23.2)	526 (18.8)	403 (23.8)	809 (20.5)	47 (25.2)	54 (28.7)	54 (17.9)	33 (23.4)	1400 (21.7)	1,926 (20.8)
Consangui			or comparison			otic group=0.004							
0	1,934 (99.8)	556 (99.5)	121 (100.0)	177 (100.0)		1,695 (100.0)	3,940 (99.9)	187 (100.0)	188 (100.0)	301 (100.0)	141 (100.0)	6452 (99.9)	9,240 (99.9)
1	3 (0.2)	3 (0.5)	0	0	6 (0.2)	0	1 (0.03)	0	0	0	0	1 (0.02)	7 (0.1)
Birth matu	urity of child (0=	· ·	term) (p value	for compariso	n between cya	notic and acyan	otic group=0.00	4)					
0	76 (3.9)	27 (4.8)	3 (2.5)	13 (7.34	119 (4.3)	98 (5.8)	208 (5.3)	5 (2.7)	9 (4.8)	12 (3.9)	6 (4.3)	338 (5.2)	457 (4.9)
1	1,861 (96.1)	532 (95.2)	118 (97.5)	164 (92.7)	2675 (95.7)	1,597 (94.2)	3,733 (94.7)	182 (97.3)	179 (95.2)	289 (96.0)	135 (95.74)	6115 (94.8)	8,790 (95.1)
Mode of d	elivery (0=norm	al delivery; 1	=caesarean se	ction) (p value	for compariso	on between cyan	otic and acyano	tic group=0.67	8)	_	_	_	-
0	1,095 (56.5)	323 (57.8)	71 (58.7)	126 (71.2)	1615 (57.8)	1,016 (59.9)	2,209 (56.1)	108 (57.7)	104 (55.3)	171 (56.8)	92 (65.3)	3700 (57.3)	5,315 (57.5)
1	842 (43.5)	236 (42.2)	50 (41.3)	51 (28.8)	1179 (42.2)	679 (40.1)	1,732 (43.9)	79 (42.3)	84 (44.7)	130 (43.2)	49 (34.7)	2753 (42.7)	3,932 (42.5)
Place of de						otic and acyanot	ic group=0.678)						
0	57 (2.9)	14 (2.5)	2 (1.6)	12 6.78	85 (3.1)	49 (2.9)	138 (3.5)	3 (1.6)	3 (1.6)	13 (4.3)	10 (7.1)	216 (3.4)	301 (3.3)
1	1,880 (97.1)	545 (97.5)	119 (98.4)	165 (93.2)	2709 (96.9)	1,646 (97.1)	3,803 (96.5)	184 (98.4)	185 (98.4)	288 (95.7)	131 (92.9)	6237 (96.6)	8,946 (96.7)
Mode of pregnancy (0=natural; 1=IVF) (p value for comparison between cyanotic and acyanotic group=0.448)										-			
0	1,924 (99.3)	556 (99.5)	121 (100.0)	176 (99.4)	2777 (99.4)	1,689 (99.6)	3,924 (99.6)	185 (98.9)	188 (100.0)	301 (100.0)	140 (99.3)	6427 (99.6)	9,204 (99.5)
1	13 (0.7)	3 (0.5)	0	1 (0.6)	17 (0.6)	6 (0.4)	17 (0.4)	2 (1.1)	0	0	1 (0.7)	26 (0.4)	43 (0.5)
Miscarriag	ge (0=no; 1=yes)	(p value for	comparison be	etween cyanoti	c and acyanoti	ic group=0.756)							
0	1,738 (89.7)	486 (86.9)	99 (81.8)	159 (89.8)	2482 (88.8)	1,495 (88.2)	3,500 (88.8)	164 (87.7)	166 (88.3)	267 (88.7)	126 (89.4)	5718 (88.6)	8,200 (88.7)
1	199 (10.3)	73 (13.1)	22 (18.2)	18 (10.2)	312 (11.2)	200 (11.8)	441 (11.2)	23 (12.3)	22 (11.7)	34 (11.3)	15 (10.6)	735 (11.4)	1,047(11.3)

Table 3: Description of the different variables across different diagnosis subgroup categories.

DISCUSSION

Our study presented a descriptive picture of the newly diagnosed CHDs among children who attended pediatric cardiology OPD of a tertiary care hospital in West Bengal, India. In our study, the most commonly identified congenital heart disease was VSD, followed by TOF and ASD. The finding was consistent with the available literature from India. A hospital-based crosssectional study by Saxena et al reported VSD as the most common CHD.¹² Another study done by Sawant et al in a tertiary care hospital also reported VSD as the most common CHD.¹³ VSD, ASD and TOF have also been identified as the most commonly occurring CHDs in a study done in China.¹⁴ In our study, we found that majority of CHDs were acyanotic (70%). This finding is also consistent with other studies done in India and outside.15,16 In both cyanotic and acyanotic CHDs, the post tricuspid anomalies were most prevalent. VSD, PDA, APW were the most common anomalies in acyanotic group, whereas the TOF, TOF-PA, TETCANAL group of anomalies were the most common cyanotic anomalies. Meshram et al reported similar findings.17

Most of the cases presented in the cardiology OPD for the first time at the age above two years. Although adults presenting with congenital hearts defects are not uncommon in India, such late presentation may pose clinical and ethical challenges in the management decision by the clinician.^{18,19} The high number of home deliveries, lack of awareness and delay in diagnosis are major contributing factors for such delayed presentation.⁵ In this study, female had more number of acyanotic heart disease where male had more number of cyanotic heart diseases and the difference was statistically significant. This finding was similar to the study by Naik et al where sex difference was present but it was not significant, whereas, another study found a significant difference.^{20,21} Data from European countries showed that gender differences existed in congenital heart diseases. The same study reported that male had higher mortality and females had higher functional limitations when the data were adjusted for age and type of defects.²² In the Indian context, this gender differentiation has greater importance as on many occasions girl child gets unequal importance in terms of health-seeking behaviour. In our study, we found that majority of the cases came from rural background. This increases the generalizability of the study findings. This also warrants that doctors working in the primary and secondary healthcare levels need to be trained for early identification and quick referral of the cases to specialized facilities. The proportion of caesarian delivery was relatively higher in our study. The finding was consistent with the evidence given by a systematic review by Prefumo et al where proportion of casesarian delivery was found near the proportion of normal vaginal delivery in case of babies having CHDs.²³ The proportion of CHD cases with a birth history of preterm deliveries was less than most of the available literature.²⁴ This finding may be due to the smaller sample size in most of the earlier studies. However, the finding was similar to a prevalence study conducted in India.²⁵ Regarding the proportion of mothers having a history of miscarriage in a previous pregnancy, it was similar to a national level finding by Maharana where the prevalence of miscarriage was 10% among mothers of the CHD cases.²⁶

We found that almost 1.5% of the CHD cases were associated with some other non-cardiac congenital defect or syndrome. A study by Stoll et al reported that 26.3% of the CHDs were associated with some other non-cardiac anomalies.²⁷ This study by Stoll et al was done in Europe. The robust screening mechanism at birth might be the explanation of such discordance with our study findings. This also warrants the implementation of a proper screening mechanism to identify birth defects in hospital settings in India. To our best effort, we could not find any study describing non-cardiac congenital defects/syndrome associated with CHDs in the Indian setting.

Congenital birth defects are associated with multiple socio-economic issues like an economic burden, anxiety of parents and fear of having similar problems in other offspring.²⁸ In our study, we found that the mother and children faced physical abuse, mental abuse, dissolution of marriage and neglect in around 1% of the cases. This being a record based study, we could not evaluate other socio-economic consequences associated with CHDs. However, we recommend establishing setups for counselling of parents regarding all birth defects including CHDs at every delivery point.

As early identification and management of congenital heart diseases have been included under rashtriya bal swasthya karyakram (RBSK), our study finding is a value to add to the existing knowledge on this issue.²⁹ However, mandatory screening of all babies at birth needs to be strengthened further along with sensitization of all the doctors especially those who are coming in contact with newborns at very early days of life. Adequate allocation of resources, training and sensitization of other health care professionals are also necessary.

The large sample size in our study itself provides strength to the study. The diagnosis was made by a trained pediatric cardiologist. Moreover, the data was collected by the cardiologist from the register and clinical notes.

Our study had few limitations. Since this was a hospitalbased study so prevalence couldn't be calculated. Moreover this was a record based study which prevented from studying associated other factors.

CONCLUSION

In this study, we have described the pattern of CHDs attending in a pediatric cardiology OPD of a tertiary care

hospital of West Bengal India. VSD was the most common congenital heart defects followed by TOF and ASD. Similarly, acyanotic heart diseases were most common and most of the cases presented for the first time after 2 years. Almost 1.5% of the CHD cases were associated with some other non-cardiac congenital defect or syndrome. In 1% of the cases, either mother or the child faced some sort of social or family problem because of the disease. We recommend considering the issue as an emerging public health problem and necessary actions are taken at the policy level.

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