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Original Research Article

Spectrum of congenital heart disease in a tertiary care centre of Northern India

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ABSTRACT

Background: Congenital heart disease (CHD) is very common disease and it is the major cause of childhood mortality and morbidity. Not much of Indian data are available particularly from the northern part of the country. It is important to mention that this part of India is unique with respect to its demographic and geographical location. There are needs to further explicate the spectrum and epidemiology of the CHD in this part of the country.
Methods: A prospective hospital-based study carried out over a period of 24 months (January 2016 to January 2018) where all suspected children (<14 years) of CHD were subjected to echocardiographic study. The age, sex, clinical presentation and echo findings were well documented.
Results: Out of total 3210 cases CHD was diagnosed in 2072 cases (64.54%). Most CHDs were diagnosed between 1 month and 6 years of age in both cyanotic and a cyanotic disease group. Incidence of a cyanotic CHD was n=1529 (47.6%) and cyanotic CHD was n=543 (17%) with the ratio of acyanotic to cyanotic 2.8:1. Ventricular septal defect was commonest CHD (35.85%) among acyanotic CHD and Tetralogy of Fallot was the commonest (12.2%) among cyanotic CHD.
Conclusions: Burden of CHD is highly underestimated and unrecognised, especially in this part of the country. This study can provide observed data that can help in policy making in the health sector. The contrasting result with respect to complex CHD in present study indicates need for good and effective antenatal cardiac screening in high risk mothers.

Keywords: Acyanotic, Congenital heart disease, Cyanotic, Echocardiography

INTRODUCTION

Congenital heart disease (CHD) (con, together; genitus, born) is a group of gross structural abnormalities that are present at birth. Congenital malformations of the heart and circulation are not fixed anatomic defects that appear at birth but instead are anomalies in flux that originate in the early embryo, evolve during gestation, survive the dramatic circulatory alterations at birth, and change considerably during extrauterine life.[1]

Congenital heart defects are the most common type of congenital disability, which constitutes an important group of pediatric illness and major cause of childhood mortality and morbidity.[2] Different genetic, environmental, and medical factors affect the CHDs worldwide. CHDs are primarily seen in neonates, infants, and children even though it is also seen in adults with undiagnosed and uncorrected CHD.

The prevalence of CHD is not uniform across the country and varies from 0.8 to 5.2/1000 patients in community-based studies while the prevalence ranges between 3.9 and 26.4/1000 live births in hospital-based studies in India, which is not uniform across the country.[3-8] Ten percent of the present infant death may be accounted for by CHD.[5]

Table 1: Classification of congenital heart diseases.

Cyanotic congenital heart diseases	Acyanotic congenital heart diseases
TOF	ASD
DORV with VSD and PS	VSD
DORV with transposed great vessels	PDA bicuspid aortic valve
d-TGA	AVSD
TA	Valvular pulmonary stenosis
TAPVC	CCTGA
SVP	Supravalvular pulmonary stenosis
Truncus arteriosus	Coarctation of aorta
PA	PAPVC
Ebstein anamoly	APW
AVSD with pulmonary stenosis	Infundibular pulmonary stenosis
CCTGA with VSD and PS	Subaortic membrane
Mitral atresia	ALCAPA
Pulmonary AV fistula	Congenital mitral stenosis
Common atrium	Interrupted aortic arch
	Dysplastic aortic valve
	Double-aortic arch
	Cortriatriatum

TOF: Tetralogy of fallot, ASD: Atrial septal defect, DORV: Double-outlet right ventricle, VSD: Ventricular septal defect, Pulmonary stenosis, PDA: Patent ductus arteriosus, TA: Tricuspid atresia, D-TGA: D-transposition of great arteries, AVSD: Atrio-ventricular septal defect, TAPVC: Total anomalous pulmonary venous connection, SVP: Single ventricular physiology, CCTGA: Congenitally corrected transposition of the great arteries, PA: Pulmonary atresia, APW: Aorto pulmonary window, PAPVC: Partial anomalous pulmonary venous connection, PS: Pulmonary stenosis, ALCAPA: Anomalous left coronary artery from pulmonary artery, AV: Arteriovenous.

They affect eight out of every 1000 newborns.[9] It is very difficult to calculate the prevalence of CHD due to home deliveries and unavailability of routine neonatal screening. Among CHD, two/third are acyanotic CHDs and the rest are cyanotic CHDs. With the improvement of pediatric cardiac care, their survival to adulthood has increased. Majority of acyanotic CHD are potentially correctable due to the evolution in percutaneous interventional therapies without exposing the patients to an open surgical procedure in early life. Only few studies from India have addressed the overall childhood spectrum (0-14 years) of CHD.[10,11] There are few studies on pattern of CHD from northern part of India. Similarly, very few studies from Kashmir valley has addressed the issue. Authors aimed to study the burden and pattern of CHD in children under 14 years of age in this northern part of the country. It is important to mention that this part of India is unique with respect to its demographic and geographical location, as the valley is surrounded by different Himalayan mountain ranges.

METHODS

This was a prospective study conducted in Postgraduate Department of Pediatrics, GMC Srinagar. The cases included all patients attending the out-patient or inpatient section of our department as well as the neonatal and Pediatric intensive care units within the age range of 0-14 years over a period of 24 months (January 2016 to January 2018).

Inclusion criteria

All cases suspected of having a CHD on clinical examination were included in the study.

Patients from neonatal intensive care unit were subjected to echocardiography due to the presence of murmur, appearance of cyanosis and tachypnoea. The usual presentation of patients from infancy was failure to thrive, breathlessness, cyanosis, presence of murmur and arrhythmias. The presence or absence of CHD and its character was confirmed by echocardiography. The echocardiography was done by a single paediatric cardiologist. The data of all patients regarding age of presentation, gender, signs and symptoms, clinical features and echo findings were documented.

RESULTS

Table 2 shows the baseline presentation of population. A total of 3210 new cases of suspected CHD attended tertiary cardiac care institute between January 2016 and Jan 2018. The total number of CHDs diagnosed were 2072 (64.54%).

Table 2: Baseline presentation of population (n=2072).

Variables		N (%)
Type of CHD	Cyanotic	543 (26.2)
	Acyanotic	1529 (73.8)
Gender	Male	1250 (60.32)
	Female	822 (39.68)
Age	0-28 days	338 (16.3)
	1 month to 1 year	1096 (53.3)
	1-6 years	517 (24.9)
	6-14 years	121 (5.8)

Around 25 congenital heart anomalies were diagnosed in our population. CHDs were more common among the

male 1250 (60.32%), with a male-to-female ratio of 1:0.66. In present study, most CHDs (n= 1434, 69.2%) were diagnosed between age of 1month to 6 years of age. Incidence of cyanotic CHD was n=543 (26.2%) and acyanotic CHDs was n=1529(73.8%), with acyanotic to cyanotic ratio of 2.8:1 (Table 2).

Age-wise distribution of congenital heart disease

Tables 3 and 4 show the pattern of CHDs identified at different age group.

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

Diagnosis	0-28 days	1month-1 year	1-6 years	6-14 years
VSD	51	502	170	20
ASD	54	143	115	34
PDA	39	150	52	14
AVSD	18	33	6	1
BAV	2	6	10	15
Valvular PS	9	18	10	3
SVPS	9	8	5	1
COA	3	7	1	1
IAA	4	-	-	-
ALCAPA	1	2	-	-
CCTGA/VSD	2	4	1	1
AP WINDOW	1	2	1	-
Total	193	875	371	90

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

Diagnosis	0-28 days	1month-1 year	1-6 years	6-12 years
TOF	25	120	93	16
DORV/VSD	16	30	19	2
dTGA/VSD	40	17	9	4
TAPVC	12	7	2	2
Tricuspid atresia/VSD	5	13	9	4
DILV with PAH/PS	9	4	3	-
VSD pulmonary atresia	12	4	1	-
Ebstein anomaly	1	9	10	1
Truncus arteriosus	3	-	-	-
HLHH	8	-	-	-
Isomerism with single ventricle	10	8	-	-
DOMV/MV abnormality	-	3	-	-
Common atrium	4	7	-	2
Total	145	221	146	31

In age-wise distribution, authors found that most diseases were diagnosed between 1 month and 6 years of age in both cyanotic and acyanotic disease groups.

The most common cyanotic and acyanotic CHDs were also found between the age of 1 month and 6 years. In cyanotic disease group, totally 254 cases of tetralogy of Fallot were diagnosed, out of which most cases 213 (83.8%) were diagnosed between 1 month and 6 years. Similarly, in acyanotic disease group, maximum cases of atrial septal defect, ventricular septal defect, and patent ductus arteriosus were diagnosed between ages of 1 month and 6 years.

Cyanotic congenital heart diseases

The distribution of different cyanotic congenital heart diseases is shown in (Table 5). Out of different types of simple cyanotic CHDs, tetralogy of Fallot (n = 254, 12.2%) was most prevalent. Other cyanotic CHDs found in present study were d-transposition of great arteries (3.3%), double-outlet right ventricle (3.2%), tricuspid atresia (1.4%), total anomalous pulmonary venous connection (1.1%), Ebstein anomaly (1.01%) in the descending order of their prevalence in present study.

Table 5: Distribution of cyanotic congenital heart diseases.

Diagnosis	N (%)
TOF	254 (12.2)
d-TGA/VSD	70 (3.3)
DORV/VSD	67 (3.2)
Triscuspid atresia/VSD	31 (1.4)
TAPVC	23 (1.1)
Ebstein anamoly	21(1.01)
Isomerism with single ventricle	18 (0.87)
VSD Pul atresia	17 (0.8)
DILV with PAH/PS	16 (0.7)
Common atrium	13 (0.6)
HLHH	8 (0.38)
Truncus arteriosus	3 (0.14)
DOMV/MV abnormality	3 (0.14)

Acyanotic congenital heart diseases

The distribution of different acyanotic congenital heart diseases is presented in (Table 6). Ventricular septal defect (35.85%) was the most common acyanotic CHD found in present study. In the acyanotic CHD group, after ventricular septal defect, atrial septal defect 346 (16.7%) and patent ductus arteriosus 255 (12.3%) were the commonly occurring CHDs. Other acyanotic CHDs were atrioventricular septal defect (2.7%), valvular pulmonary stenosis (1.9%), bicuspid aortic valve (1.5%), supravalvular pulmonary stenosis (1.1%), coarctation of aorta (0.5%), congenitally corrected transposition of the great arteries (0.38%), aortopulmonary window (0.12%)

in the descending order of them of prevalence in present study.

Table 6: Distribution of acyanotic congenital heart diseases.

Diagnosis	N (%)
VSD	743 (35.85)
ASD	346 (16.7)
PDA	255 (12.3)
AVSD	58 (2.7)
BAV	33 (1.5)
Valvular PS	40 (1.9)
SVPS	23 (1.1)
COA	12 (0.5)
CCTGA/VSD	8 (0.38)
IAA	4 (0.19)
AP window	4 (0.19)
ALCAPA	3 (0.14)

DISCUSSION

There are only few Indian studies which show the pattern and prevalence of CHD. This is the one of main referral pediatric hospital in Kashmir which caters large population of varied ethnicity. Most of CHDs were diagnosed between 1 month and 6 years (69.2%), which is comparable to other studies.[12] The highest number of cases was seen between 1 month and 6 years of age which could be because of a large number of referrals from peripheral health center and increasing use of echocardiography. In present study, CHDs were more common among the male 1250 (60.32%), with a male-to-female ratio of 1.52:1. Present study shows that the male preponderance which is similar to other studies showed male-to-female ratio of 1.78:1 and 2.08:1, respectively.[13-15] This male dominance pattern could be due to Indian social and cultural factors. Neglect, differential treatment, or poor access to health-care facilities is putting girls at disadvantages. Moreover, this could be the reason for less female child seeking the health-care facilities.

In the present study, incidence of cyanotic CHD was 543 (26.2%) and acyanotic CHDs was 1529 (73.8%)

Ventricular septal defect (38.85%) was the most common CHD found in present study. In acyanotic CHD, after ventricular septal defect, atrial septal defect 350 16.89%) and patent ductus arteriosus 259 (12.5%) were the commonly occurring CHDs. Present study results are in line with the study done by Bhat et al, who stated that ventricular septal defect was most common in 30.4% patients, followed by atrial septal defect in 17.63% and patent ductus arteriosus in 9.62%.[10] The higher incidence of VSD can be due to inclusion of newborns in present study. This incidence of VSD actually overestimates the haemodynamically significant VSDs. The lower incidence of ASD can be due to reason that ASD yields soft murmur and relatively asymptomatic course in early infancy leading to low diagnosis rate. Hence, the incidence of atrial septal defect in childhood actually underestimates the true incidence. The low incidence rate of PDA can be attributed to exclusion of hemodynamically insignificant PDA in neonatal age. The lower incidence rate of AVSD is attributed that AVSD is usually associated with triosomy 21 and higher attrition rate of children with triosomy may lead to lower incidence of AVSD.

In present study the most common cyanotic CHD was tetralogy of Fallot (n = 250, 12.05%). Tetralogy of Fallot according to natural history usually presents late and has favorable natural history, which can be the reason that it is the most common cyanotic CHD encountered in present study has found in many other studies.[16] However this incidence of TOF is low as compared to many studies like Abqari et al.[17] This low incidence can be explained as authors have included only classical TOF and the other variants of VSD PS physiology are grouped separately.

The frequency of complex CHD (2.2%) children with univentricular heart is comparable to the incidence of complex CHD in west, but more when compared to data available from India.[18-20] This increased evidence of complex CHD can be due to high rate of consangious marriage in this part of India. Besides, being the main referral unit for sick neonates, most of neonates with complex CHD are diagnosed at our centre. Furthermore, the antenatal diagnosis of congenital heart defects is still in infancy in this region, with very small percentage of pregnant ladies going for fetal echocardiography.

CONCLUSION

Burden of CHD is highly underestimated and unrecognised, especially in this part of the country. This study included large number of patients over period of 2 years from main paediatric centre of the Kashmir valley. Hence, this study has yielded important data on epidemiology and incidence of CHDs. This study can provide observed data that can help in policy making in the health sector. This study will also provide awareness in people with family history of CHD especially where rate of consangious marriages are high. The contrasting result with respect to complex CHD in present study indicates need for good and effective antenatal cardiac screening in high risk mothers.

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