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Research Article

THE SPECTRUM OF CYANOTIC HEART DEFECTS IN A TERTIARY CARE HOSPITAL IN NORTH INDIA

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A R T I C L E I N F O

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A B S T R A C T

Background: There is a lack of data about the present spectrum of congenital cyanotic heart disease [CCHD] in the pediatric age group. The present study was undertaken to determine the spectrum of patients with CCHD in the paediatric age group in tertiary paediatric cardiac care centre.
Methods: The study was carried out in the Department of Paediatrics Government Medical College Srinagar. All patients referred with complaints or clinical examination suggestive of CHD were further evaluated .On basis of clinical examination, echocardiography findings patients having congenital heart defects were included as cases which were further divided into cyanotic and acyanotic heart defects. The profile and mode of presentation of various cyanotic CHDs were further described in detail.
Results: There were 543 subjects with CCHD with a male to female ratio of 1.6:1. The children were aged between 2 days to 14 years. The most common type of CCHD (both isolated and multiple CCHD) was tetralogy of Fallot (TOF) followed by transposition of the great arteries (TGA) and double outlet right ventricle (DORV). The most common mode of presentation was cyanosis.
Conclusion: This study have yielded important data on epidemiology and incidence of CCHDs. This has raised the important need to prioritise the foetal and early neonatal diagnosis of cyanotic CHD through various modalities like foetal echocardiography and neonatal echocardiography screening as the majority of the neonates with congenital cyanotic heart disease showed survival with appropriate management.

INTRODUCTION

The incidence and prevalence of cyanotic congenital heart diseases (CCHD) vary worldwide. It accounts for a third of the congenital heart disease (1). CCHD can be classified into (2):

i. CCHD due to right-to-left shunt with decreased pulmonary flow: [Ventricular Septal Defects with Pulmonary stenosis physiology (VSD+PS physiology), Tetralogy of Fallot (TOF), DORV(double outlet left ventricle) VSD+PS, Congenitally corrected Transposition CCTGA+VSD+PS , Complete atrioventricular septal defect (CAVSD +PS), Pulmonary atresia with VSD and Pulmonary atresia with intact septum]
ii. CCHD due to right-to-left shunt with the decreased aortic flow: [Left-sided hypoplastic heart, interrupted arch, severe coarctation]
iii. CCHD disease due to admixture lesions: [TGA(transposition of great arteries), Tricuspid atresia with VSD (TA +VSD), Truncus arteriosus, etc. (2)

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Few prospective studies have demonstrated the different spectrum of cyanotic CHD in neonatal age; however, no previous study has tried to find out the relative spectrum of congenital cyanotic heart disease in overall children of age 0-15 years.[3] This prospective observational study was undertaken to describe the frequency of various forms of congenital cyanotic heart disease diagnosed by echocardiography in children. This study has demonstrated the relative proportion of cases with congenital cyanotic heart disease evaluated in a tertiary cardiac care centre. It is important to mention that this part of India is unique concerning its demographic and geographical location, as different Himalayan mountain ranges surround the valley. Furthermore, the rate of consangious marriage in our part of the country is also high, hence more predisposition to genetic disorders.

MATERIALS AND METHODS

This was a prospective observational study conducted in a tertiary cardiac care centre in North India over a period of 24 months (Jan 2016 to Jan 2018). All patients under 15 years of age who visited the paediatric cardiac clinic within this study period of our hospital with clinically suspected congenital

cyanotic heart disease were provisionally included in this study. History and physical examinations were obtained as necessary. Plain chest radiograph, electrocardiography and other ancillary investigations were done as required. They underwent thorough echocardiographic evaluation (2D echocardiography and Doppler echocardiography). Cardiac CT angiography was done wherever necessary. After echocardiographic and Cardiac CT evaluation, those who had a definite diagnosis of CCHD were finally included in this study. We had also included those patients who had palliative shunt operation or corrective surgery for CCHD and came for follow-up during our study period. Those patients who were not clinically cyanosed, although echocardiography revealed cardiac anomaly suggestive of CCHD with increased pulmonary blood flow or admixture lesion, were included in our study.

RESULTS

Demographics of the study participant

Between January 2016 and January 2018 a total of 543 children had CCHD. There were 335 males and 208 females with a male to female ratio of 1.6:1. The children were age zero days to fourteen years .136 cases [25%] patients were less than one month old. Majority of patients were aged between 1 month to 1 year, n=225[41.4%], of which males were 135 and females were 90. 151[27.8%] patients were between 1 to 6year while 31 patients with CCHD were more than 6 years. By the age of 5 years, 512 [94%] of the children had been diagnosed. [Table 1]

Table 1 Baseline presentation of the population (n=543)

		Male	female
Total	**N=543**	**335[61.62%]**	**208[38.3%]**
Age			
0-28 days	136[25%]	80	56
1 month – 1 year	225[41.4%]	135	90
1-6 years	151[27.8%]	90	61
6-14 years	31[5.7%]	21	10

Age-wise distribution of congenital heart disease

Within the study period, 543 were diagnosed with having cyanotic CHD through clinical, echocardiographic and CT angiographic evaluation. The various diagnoses of cyanotic CHD in each age group are presented in Table 2 and 3.

Table 2 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
TGA/VSD	40	17	9	4
TAPVC	12	10	2	2
Tricuspid atresia/VSD	5	13	9	4
AVSD PS	1	3	3	
VSD Pulmonary atresia	8	4	1	
Pulmonary atresia with intact IVS	4	4	1	
Ebstein anomaly	1	9	10	1
Truncus arteriosus	3			
HLHS	9	3		
Isomerism with single ventricle	10	8		
CCTGA /VSD/PS	1	2	2	
Common atrium	1	2	2	2
Total	136	225	151	31

TOF Tetralogy of Fallot ;d-TGA: D-transposition of great vessel,VSD ventricular septal defect; TAPVC: total anomalous pulmonary venous connection,AVSD atrioventricular septal defect HLH:Hypoplastic left heart, DILV with PAH/PS: double inlet left ventricle with pulmonary arterial hypertension, PS: pulmonary stenosis, CCTGA; congenitally corrected transposition of great vessel, TOF: tetralogy of Fallot,IVS Interventricular septum

Table 3 Distribution of cyanotic congenital heart diseases

Diagnosis	n (%)
TOF	254 (46.77)
d-TGA/VSD	70 (12.8)
DORV/VSD	67 (12.3)
Tricuspid atresia/VSD	31 (5.7)
TAPVC	26 (4.7)
Ebstein anomaly	21(3.8)
Isomerism with single ventricle	18 (3.3)
VSD Pulmonary atresia	13(2.3)
Pulmonary atresia Intact IVS	9[1.6]
HLHS	12 (2.2)
AVSD with PS	7[1.2]
Common atrium	7(1.2)
CCTGA /VSD/PS	5[0.9]
Truncus arteriosus	3 (0.5)

Types of CCHD

Of the 543 patients of CCHD. These were 254 patients of TOF, 67 of double outlet right ventricle (DORV),70 had TGA, 31 Tricuspid Atresia with VSD,26 children with Total anomalous pulmonary venous connections (TAPVC),21 with Ebstein anomaly,18 with single ventricle PS/PAH physiology , 22 had pulmonary atresia of which 13 had VSD and 9 had Intact IVS.12 patients with hypoplastic left ventricle (HLHS), 7 each of common atrium and AVSD /PS . 5 had CCTGA/VSD/PS. Only 3 had Truncus arteriosus. The demographics of the different subtypes of CCHD are highlighted in Table 2.

TOF (Tetralogy of Fallot)

TOF was the most common CCHD, documented in 46.7% of the children with CCHD. There were a total of 254 cases of TOF. Most of the patients, (47.1%) were between 1month to 1 year. Only 25 presented in neonatal age [9.8%]. These neonates had severe pulmonary stenosis with little antegrade flow. Three quarter, 75% of the patients were cyanosed at presentation. 5 had no visible cyanosis [Pink TOF].4 children presented with polycythemia and vertigo. Two patients had brain abscess. Majority of children had classical TOF, 5 had TOF with absent pulmonary valve. 5 patients had DiGeorge syndrome while trisomy 21 was present in 2 patients.

TGA : There were 70 cases of TGA. 40 cases [57 %] presented in neonatal age. Most of them had Intact Interventricular septum. 13 children were more than 1 year at diagnosis. Cyanosis was less conspicuous in them and they usually presented with Congestive cardiac failure. All of them had Ventricular septal defect.

DORV VSD: A total of 67 cases of DORV were recorded. Of the 67 cases, 45 DORV occurred without other cyanotic heart lesions. TGA was the most common cyanotic congenital heart lesion that occurred with the DORV [Taussing Bing Anomaly]. All the patients with DORV had co-existing intracardiac lesions. The most common was pulmonary stenosis. Majority of children were between 1 month to 1 year.

TAPVC: This lesion was seen in 26 patients.Out of the 26 cases, 15 had Supracardaic TAPVC, 4 cases of Infracardiac TAPVC and 4 cases were of Cardiac TAPVC, however, 3 had mixed TAPVC. Most of the cases presented in neonatal age.

***Tricuspid Atresia With* VSD:** This was present in 31 cases. Those presenting in neonatal age had severe pulmonary stenosis. Infants usually presented with CCF. 3 Patients had undergone palliative surgery. (Bidirectional Glenn in two and Fontan completion in one)

Ebstein Anomaly: 21 patients presented with Ebstein anomaly. Majority of children were more than 1year old. 8 patients presented with CCF, 2 with arrthymia. 5 patients were diagnosed while being evaluated for cardiomegaly.

***Single Ventricle With* PS/PAH**: Out of the 543 cases of CHD, 18 cases were of single ventricle. Most of the cases of single ventricle had presentation since birth, cyanosis as most common presentation.10 patients presented with cyanosis only while 8 were having cyanosis along with features of increased pulmonary blood flow.

HLHS: It was present in 12 cases. Majority presented in neonatal age with shock and cyanosis. Three cases had large PDA and presented beyond the neonatal period.

***Pulmonary Atresia*:** 22 patients had pulmonary atresia of which 13 had VSD and 9 had intact ventricular septum. Majority 13 presented in the early neonatal period with cyanosis. Four children had cyanosis along with heart failure due to large major aortopulmonary collateral.

CCTGA VSD PS, AVSD PS; each present in 7 children.3 children with AVSD PS had Trisomy 21 .2 children with CCTGA VSD PS had situs inversus with dextrocardia

Common Atrium: was present in 7 children. The most common presentation was sutble cyanosis with poor weight gain.

Truncus Arteriosus: Only 3 neonates had Truncus arteriosus of which one had DiGeorge syndrome.

DISCUSSION

The present study has documented a recent report on the clinical profile of children with cyanotic congenital heart disease in a tertiary hospital in a developing country. Most of the studies had shown male preponderance in having CHDs. Similarly, our study also found M: F ratio 1.60 similar to the studies of Hussain *et al* [4] and Kumar *et al* [5] which were 2.08:1 and 1.78:1 respectively. In contrast, Khalid *et al* [6] didn't find much of gender disparity. Gender discrepancy is quite striking, which might be due to genetic reasons or because of early reporting of male progeny by the parents.

The age at diagnosis of CCHD depends on the type of heart lesion and other associated factors such as severity of the lesion, other co-morbidity and where the children reside (western countries or under-developed regions of the world). It was observed that the children with TGA, TAPVC, TA, Truncus Arteriosus and DORV were diagnosed earlier than the patients with TOF The finding in this regard shows that most diagnoses are made in the first 12 months of life and by their 5th birthday most children with CCHD had been diagnosed [7,8]. In our study 41.4% of children were diagnosed in infancy and 94% of total CCHD were diagnosed before 6 years. Early diagnosis can be due to the earlier presentation of some critical CCHD and due to the increasing availability of facilities and expertise in the diagnosis of heart disease. Least number of patients were diagnosed more than 6 years of age. Possible reasons for the late presentation and diagnosis includes difficulty in assessing specialized care, poverty and poor health seeking behaviour [9].

Concerning the distribution of the different types of cyanotic heart lesions, TOF was the most common defect keeping in view with the fact that TOF is commonest CCHD [10]. The second and third most common CCHD were TGA and DORV as found in many studies [10]. The frequency of complex CHD (3.3%) in our study is comparable to the incidence of complex CHD in the west [11], but more when compared to data available from India [12,13]. This increased evidence of complex CHD can be due to the high rate of consangious marriage in this part of India. Besides, being the main referral unit for sick neonates, most the 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 a very small percentage of pregnant ladies going for fetal echocardiography.

The number of cases with critical CCHD like HLHS, pulmonary atresia intact ventricular septum were higher in our study than other reported studies[14] as we have obstetrics and neonatal department in our hospital, congenital cyanotic heart disease, which occurred right after birth and which needed to be addressed immediately.

CONCLUSION

Our centre is the main paediatric tertiary care centre in our area. It also has large catchment area and the population of varied ethnicity are admitted here. We are used to getting good numbers of congenital cyanotic heart disease to assess the proportion of different congenital cyanotic heart disease in the community. We need to prioritise the foetal and early neonatal diagnosis of cyanotic CHD through foetal echocardiography, neonatal oxygen saturation testing, neonatal echocardiography, and colour Doppler screening as the majority of the neonates with congenital cyanotic heart disease showed survival with approprivate management[15].

Burden of CCHD 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 have yielded important data on epidemiology and incidence of CCHDs. 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 our study indicates need for good and effective antenatal cardiac screening in high risk mothers.

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