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TO STUDY THE CLINICAL PRESENTATION, ASSOCIATED EXTRA-CARDIAC ANOMOLIES AND OUTCOME OF PATIENTS OF NEONATAL CARDIAC DISORDERS

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ABSTRACT

Background: Congenital heart disease (CHD) is the commonest of all congenital lesions and is the most common type of heart disease among children.[1]

Congenital heart diseases are not fixed anatomic defects that appear at birth, but are instead a dynamic group of anomalies that originate in fetal life and change considerably during the postnatal development.[2] The incidence of moderate-to-severe structural CHD in live born infant is 6–8 per 1000 live births.[3] About 2–3 per 1000 newborns will be symptomatic with heart disease in the 1st year of life. CHD is considered as one of the leading causes of neonatal mortality.[4]

Aim: To study the clinical presentation, associated extra-cardiac anomolies and outcome of patients of Neonatal Cardiac Disorders.

Material and Methods: It was an observational prospective study carried out in the Neonatology Section, Department of Pediatrics, GB Panth Childrens hospital Srinagar from November 2017 to August 2019.

Results: During the two year study, 471 neonates were diagnosed with congenital heart disease in which 335 (71.1%) were acyanotic and 136 (28.9%) were cyanotic. Males and females comprised of 253 and 218 respectively with a male to female ratio of 1.2:1.

Nearly half of the cases presented in the 4th week 217 (46.1%) followed by those in 3rd week 121 (25.7%), 2nd week 79 (16.8), and 1st week 54 (11.5%). the neonates presented with various symptoms of respiratory distress, feeding difficulty, and combination of symptoms; however, the most common presenting cardinal complaint was fast breathing in 201 (42.7%), followed by difficult feeding 98 (20.8%), cyanosis 83 (17.6%), shock manifested as decreased urinary output in 40 (8.5%), fast breathing and cyanosis were present in 21 cases (4.5%). Thirty-eight cases (5.9%) were asymptomatic and clinically only murmur was present. Associated (extra-cardiac) anomalies were seen in 87 (18.5%) in our study in which Down's syndrome was the commonest association with congenital heart disease. The immediate outcome of the neonates showing 57 (41.9%) of cyanotic CHD died within one month whereas only 35 (10.4%) acyanotic CHD expiring at one month.

Conclusion: congenital heart diseases in neonates have a varied presentation.high index of suspicion and early timely treatment should be done.

KEYWORDS: congental heart diseases, cyanosis, acyanotic heart diseases. To study the clinical presentation, associated extra-cardiac anomolies and outcome of patients of Neonatal Cardiac Disorders

INTRODUCTION

Congenital heart disease (CHD) is the commonest of all congenital lesions and is the most common type of heart disease among children.[1]

Congenital heart diseases are not fixed anatomic defects that appear at birth, but are instead a dynamic group of anomalies that originate in fetal life and change considerably during the postnatal development.[2] The incidence of moderate-to-severe structural CHD in live born infant is 6–8 per 1000 live births[3]. About 2–3 per 1000 newborns will be symptomatic with heart disease in the 1st year of life. CHD is considered as one of the leading causes of neonatal mortality.[4]

Asian race is found to be more affected than non-Asian race due to high rate of consanguineous marriages.[5] The worldwide prevalence of CHD is estimated to be 8-10 in every 1000 live births but the prevalence greatly varies between regions. Nearly 1/3rd of the congenital heart diseases (CHD) are critical requiring interventions in the first year of life.[6] CHDs contribute to infant mortality significantly as 7% of the neonatal deaths are due to congenital malformations, 25% of which are cardiovascular.[6]

In India, 10% of the present infant mortality may be accounted for by Congenital Heart Disease as reported by Saxena et al.[7] The incidence of severe CHD requiring expert cardiologic care is around 2.5-3/1000 live births.[8]

The neonates with CHD may present with, feeding difficulty, fast breathing, cyanosis, cardiovascular collapse, and congestive heart failure or combination of these presentations. Pure versions of specific defects may present in some patients, but many neonates have various combinations of defects.[9]

The initial evaluation of any newborn suspected of having critical CHD includes a meticulous physical examination, four extremity blood pressures, preductal and postductal saturations, hyperoxia test, ECG and chest radiograph. Echocardiography, with Doppler and color Doppler, has become the primary diagnostic tool for CHD. In addition, it reduces the requirement for invasive studies such as cardiac catheterization.[10]

MATERIAL AND METHODS

The study was conducted in the Postgraduate Department of Pediatrics, GB Pant Children's Hospital an associated hospital of Government Medical College, Srinagar. This study was in continuation of our previous study, To study the spectrum of neonatal congenital cardiac disorders.The hospital has a catchment area of both rural and urban populations and is the referral tertiary care hospital of valley.

All children were screened through proper history and examination followed by ancillary tests like ABG, SpO2, ECG and Chest X-ray

Study Design: It was an observational prospective study carried out in the Neonatology Section, Department of Pediatrics, GB Panth Childrens hospital Srinagar from November 2017 to August 2019.

Inclusion Criteria

All neonates suspected of congenital heart disease presenting to Pediatric OPD/IPD/Nursery, on the basis of history and clinical examination were included and diagnosis was confirmed by echocardiography.

Details of all neonates having CHD diagnosed by echocardiography were noted in the preformed proforma and their incidence, clinical profile, associated extra-cardiac anomalies and immediate outcome was studied.

Echocardiography was done by a single pediatric cardiologist on SIEMENS ACUSON SC2000 using M-mode, two-dimensional color Doppler cardiac imaging.

STATISTICAL METHODS

The recorded data was compiled and entered in a spreadsheet (Microsoft Excel) and then exported to data editor of SPSS Version 20.0 (SPSS Inc., Chicago, Illinois, USA). The data was summarized as frequencies and percentages. Graphically the data was presented by bar and pie diagrams. Chi-square test was employed to compare various parameters between cyanotic and acyanotic heart disease. A P-value of 0.05 was considered statistically significant.

RESULTS AND OBSERVATION

During the two year study, 471 neonates were diagnosed with congenital heart disease in which 335 (71.1%) were acyanotic and 136 (28.9%) were cyanotic. Males and females comprised of 253 and 218 respectively with a male to female ratio of 1.2:1.

During first week 54 (11.5%) neonates presented to hospital, 79 (16.8%) presented in the 2nd week, 121 (25.7%) presented in 3rd week while as majority i.e. 217 (46.1%) neonates presented in the 4th week.

Among cyanotic and acyanotic CHDs 44 (32.4%) and 10 (3%) respectively presented to our hospital in the first week followed by 34 (25%) cyanotic and 45 (13.5%) acyanotic CHDs presented in the 2nd week, 31 (22.8%) cyanotic and 90 (26.9%) acyanotic CHDs presented in the 3rd week while 27 (19.9%) cyanotic and 190 (56.7%) acyanotic CHDs presented in the 4th week.

Table 1: Distribution of study neonates as per type of CHD.

Type of CHD	Frequency	Percentage
Cyanotic	136	28.9
Acyanotic	335	71.1
Total	471	100

In our study, majority of neonates had acyanotic CHD 335 (71.1%) while cyanotic CHD comprised of 136 (28.9%).

Table 2: Age of presentation in study neonates.

Age (Weeks)	Frequency	Percentage
1st Week	54	11.5
2nd Week	79	16.8
3rd Week	121	25.7
4th Week	217	46.1
Total	471	100

Nearly half of the cases presented in the 4th week 217 (46.1%) followed by those in 3rd week 121 (25.7%), 2nd week 79 (16.8), and 1st week 54 (11.5%).

In our study males were more than females with 253 (53.7%) and 218 (46.3%) with a male to female ratio of 1.2:1.

Table 3: Time of presentation according to type of CHD in study neonates.

Age (Weeks)	Cyanotic		Acyanotic	
	No.	%age	No.	%age
1st Week	44	32.4	10	3.0
2nd Week	34	25.0	45	13.4
3rd Week	31	22.8	90	26.9
4th Week	27	19.9	190	56.7
Total	136	100	335	100
Chi-square=109.64; P-value<0.001 (Statistically Significant)				

Above table shows the time of presentation of newborns with cyanotic CHD versus acyanotic CHD. Majority of neonates with acyanotic CHD 190 (56.7%) presented in the 4th week, while majority of cyanotic and critical CHD 44 (32.4%) presented in the 1st week.

Table 4: Chief complaints at presentation in study neonates.

Complaint	Frequency	Percentage
Fast breathing	201	42.7
Difficult feeding	98	20.8
Cyanosis	83	17.6
Shock	40	8.5
Fast breathing and cyanosis	21	4.5
Murmur	28	5.9
Total	471	100

The neonates presented with various symptoms of respiratory distress, feeding difficulty, and combination of symptoms; however, the most common presenting cardinal complaint was fast breathing in 201 (42.7%), followed by difficult feeding 98 (20.8%), cyanosis 83 (17.6%), shock manifested as decreased urinary output in 40 (8.5%), fast breathing and cyanosis were present in 21 cases (4.5%). Thirty-eight cases (5.9%) were asymptomatic and clinically only murmur was present.

Table 5: Types of acyanotic heart disease.

Type of CHD	Frequency	Percentage
ASD	98	20.8
VSD	153	32.5
PDA	47	10.0
AVSD	15	3.2
BAV (severe AS)	2	0.4
Valvular PS (mild/moderate)	3	0.64
Peripheral PS	11	2.3
COA	2	0.4
ALCAPA	1	0.2
CCTGA/VSD	2	0.4
AP window	1	0.2
Total	335	71.1

CHD, congenital heart disease; ASD, Atrial septal defect; VSD, Ventricular septal defect; PDA, Patent ductus arteriosus; AVSD, Atrioventricular septal defect; BAV, Bicuspid aortic valve; AS, Aortic stenosis; PS, pulmonary stenosis; CAO, Coarctation of aorta; ALCAPA, Anomalous left coronary artery from pulmonary artery; CCTGA = Congenitally corrected transposition of great arteries; AP = Aortapulmonary window

In our study the most common CHD was ventricular septal defect (VSD) 103 (32.5%) followed by atrial septal defect (ASD) 98 (20.8%) and PDA 47 (10.0%).

Table 6: Types of cyanotic heart disease and their age of presentation.

Type of CHD	1st Week	2nd Week	3rd Week	4th Week	Total
d-TGA	23	3	5	8	39
TOF	2	5	8	8	23
TAPVC	3	4	2	2	11
HLH with interrupted aortic arch	5	3	1	0	9
VSD, pulmonary atresia	2	3	3	2	10
DILV with PAH/PS	3	7	5	2	17
Truncus arteriosus	0	2	1	0	3
Critical PS with PFO/ASD R-to-L	1	2	1	0	4
Tricuspid atresia, VSD	1	1	2	1	5
Ebstein anomaly	1	2	0	0	3
CCTGA/VSD/pulmonary atresia	0	0	1	1	2
Isomerism with complex CHD	2	1	1	3	7
Taussig-Bing anomaly	1	1	1	0	3
Total	44	34	31	27	136

CHD, congenital heart disease; d-TGA, D-transposition of great arteries; TOF, Tetralogy of fallot; TAPVC, Total anomalous pulmonary venous connection; HLH, Hypoplastic left heart; VSD, Ventricular septal defect; DILV with PAH/PS, Double inlet left ventricle with pulmonary arterial hypertension/ pulmonary stenosis; PFO, Patent foramen ovale; ASD, Atrial septal defect; CCTGA = Congenitally corrected transposition of great arteries.

The above table depicts the age of presentation of various cyanotic CHD with d-transposition of great arteries (d-TGA) as the most common. Age of presentation was first week in majority of cases except tetralogy of fallot.

Table 7: Types of acyanotic heart disease and their age of presentation.

Type of CHD	1st Week	2nd Week	3rd Week	4th Week	Total
ASD	4	17	25	52	98
VSD	2	3	34	114	153
PDA	3	17	19	8	47
AVSD	0	3	5	7	15
BAV (severe AS)	1	1	0	0	2
Valvular PS (mild/moderate)	0	0	1	2	3
Peripheral PS	0	2	3	6	11
COA	0	1	1	0	2
ALCAPA	0	1	0	0	1
CCTGA/VSD	0	0	1	1	2
AP window	0	0	1	0	1
Total	10	45	90	190	335

CHD, congenital heart disease; ASD, Atrial septal defect; VSD, Ventricular septal defect; PDA, Patent ductus arteriosus; AVSD, Atrioventricular septal defect; BAV, Bicuspid aortic valve; AS, Aortic stenosis; PS, pulmonary stenosis; CAO, Coarctation of aorta; ALCAPA, Anomalous left coronary artery from pulmonary artery; CCTGA = Congenitally corrected transposition of great arteries; AP = Aortapulmonary window

The above table depicts the age of presentation of various acyanotic CHD with ventricular septal defect (VSD) as the most common.

Age of presentation was fourth week in majority of cases.

Table 8: Associated extra-cardiac anomalies in study neonates.

Associated extra-cardiac anomalies	Frequency	Percentage
Downs syndrome	32	6.8
Skeletal deformity	20	4.2
Anorectal malformations	14	3.0
TEF	10	2.1
Renal anomaly	7	1.5
Cleft lip with palate	4	0.8
Total	87	18.5

TEF, Tracheo-esophageal fistula

Associated (extra-cardiac) anomalies were seen in 87 (18.5%) in our study in which Down's syndrome was the commonest association with congenital heart disease.

Table 9: Immediate Outcome.

Expired	No. of Patients	Percentage
Cyanotic	57	41.9%
Acyanotic	35	10.4%
Total	92	19.5
Chi-square=60.93; P-value<0.001 (Statistically Significant)		

Above table shows the immediate outcome of the neonates showing 57 (41.9%) of cyanotic CHD died within one month whereas only 35 (10.4%) acyanotic CHD expiring at one month.

DISCUSSION

According to Mitchell et al's definition, congenital heart disease is a gross structural malformation of the heart or great intrathoracic vessels with a real or potential functional importance.[1] Therefore this definition excludes anomalies such as bicuspid aortic valve without valve dysfunction, mitral valve prolapse, persistent left superior vena cava, anomalous origin of the left subclavian artery, mild valve regurgitation, and functional alterations without a structural component. This definition was adopted in this study.

Congenital heart disease (CHD) has already been known as an important cause of significant morbidity and mortality in the neonatal period.

During the 2-year study, 471 neonates were diagnosed with CHD of which 335 (71.1%) were acyanotic CHD and 136 (28.9%) were cyanotic CHD, which was comparable to a study by **Shah GS et al (2008).**[18] wherein the cyanotic CHD constituted 31% and acyanotic 69%. Similarly, in a study by **Deo B et al. (2015)**[19] 32.5% belonged to cyanotic group and 67.5% belonged to acyanotic group.

Most of the cyanotic variety 44 (32.4%) presented in the 1st week of life, while acyanotic 190 (56.7%) lesions presented in the 4th week of life, which is comparable to a study conducted by **Humayun KN et al (2008)**[13] in which the mean age of presentation of neonates with CHD was 5 days and all had cyanotic type of CHD. Hence, most of the critical and cyanotic CHD present in the first week of life indicating that early detection of these neonates is critical for their survival.

In our study, the ratio of male-to-female was 1.2:1. This is comparable to many studies viz. **Shah GS *et al* (2008)**[18] in Nepal wherein the male-to-female ratio was 1.5:1. **Humayun KN et al (2008)**[13] in Pakistan wherein male-to-female ratio was 1.7:1. The male preponderance in CHD was seen in majority of the studies conducted worldwide.

Majority of neonates presented with breathing difficulty 201 (42.7%), feeding difficulty 98 (20.8%), and cyanosis 83 (17.6%). A study by **Molaei A et al (2015)**[20] in Iran, observed that respiratory distress (70%) was the most common complaint and cyanosis (24.4%) was the second-most common complaint.

In our patients d-TGA was the most frequent type of cyanotic CHD with a frequency of 39 (8.3%), followed by TOF in 23 (4.9%) and DILV with PAH/PS in 17 (3.6%). Our work was in agreement with studies done by **Islam MN et al. (2013)**[15] and **Farooqui et al (2010)**[14] However, in studies done by **Patra S et al. (2015)**[21] and **Hussain S et al. (2013)**[16] the most common types of cyanotic CHD were TOF followed by d-TGA. This difference can be because of the inclusion of only neonates in our study, while other studies included older children and usually TOF presents after a few months of life. Higher incidence of complex CHD in our study can be due to high rate of consanguineous marriage in this part of India. Besides, it also reveals low rate of antenatal

diagnosis of complex CHD. There is less awareness about fetal echocardiography.

Among acyanotic CHD, the most common CHD is VSD in 153 (32.5%), followed by ASD in 98 (20.8%) and PDA in 47 (10%). Our results are comparable to study by **Hussain S et al (2013),[16] Khalil A et al (1994)[12]** noted VSD and PDA were the most common lesions found in 34.8% and 18.6%, respectively. The lower rates of PDA in our study are due to inclusion of only hemodynamically significant PDA in our study with left atrium:aorta ratio of 1.2:1.

It is well known that extra-cardiac anomalies are associated with congenital heart disease. Associated non-cardiac malformations noted in identifiable syndromes may be seen in as many as 25% of patients with congenital heart disease.

Associated (extra-cardiac) anomalies in our study were seen in 87 (18.5%) in which Down's syndrome was the commonest association with congenital heart disease. In a study conducted by **Tank S et al (2004),[22]** 10% of cases of congenital heart disease had syndromes and other associated somatic anomalies among which Down's syndrome was the commonest. Similarly **Khalil et al (1994)[12]** noted an incidence of 17.9% of somatic anomalies in patients with congenital heart disease with Down's syndrome as the commonest one. In another study conducted by **Kasturi L et al (1999),[23]** 20% of cases with congenital heart disease had extra cardiac anomalies.

In our study, 57 (40.8%) of neonates with cyanotic CHD died within 1 month. This mortality rate is comparable to the study by **Humayun KN et al (2008)[13]** 36.4%, 50% by **Ravilala VK et al (2018).[17]** This mortality rate is higher than in other studies by Shah *et al*. which showed mortality rate of 20%. The difference may be due to the difference in the study population and the limited availability of cardiac facilities.

CONCLUSION

Congenital heart disease (CHD) is a common congenital disorder of the neonatal population. Early diagnosis and timely management are key factors for optimal outcome of this problem. It can be easily diagnosed by detailed and careful clinical examination by trained medical personnel, pulse oxymetry and echocardiography. Exercising high index of suspicion during the neonatal examination can significantly change the outcome of CHD. Neonates presenting with multiple anomalies should be screened for any underlying structural heart disease.

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