TO STUDY THE SPECTRUM OF NEONATAL CONGENITAL CARDIAC DISORDERS

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Background: Congenital heart disease (CHD) is the commonest of all congenital lesions and is the most common type of heart disease among children. Congenital heart disease, in a definition proposed by Mitchell et al is “a gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance.” It is the most common cause of major congenital anomalies, representing a major global health problem. Twenty-eight percent of all major congenital anomalies consist of cardiac anomalies. In India, 10% of the present infant mortality may be accounted for by Congenital Heart Disease as reported by Saxena et al.

Aim: To study the Spectrum of Neonatal Congenital Cardiac Disorders

Material and methods: It was an observational prospective study carried out in the Neonatology Section, Department of Pediatrics, GB Pant 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. The most common cyanotic CHD was d-transposition of great arteries (d-TGA) 39 (8.3%), followed by tetralogy of Fallot (TOF) 23 (4.9%) and single ventricle (double inlet left ventricle [DILV] with pulmonary arterial hypertension/pulmonary stenosis [PAH/PS]) 17 (3.6%). In our study the most common acyanotic CHD was ventricular septal defect (VSD) 103 (32.5%) followed by atrial septal defect (ASD) 98 (20.8%) and PDA 47 (10.0%).

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.

Introduction:

Congenital heart disease (CHD) is the commonest of all congenital lesions and is the most common type of heart disease among children1. Congenital heart disease, in a definition proposed by Mitchell et al is “a gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance.” It is the most common cause of major congenital anomalies, representing a major global health problem. Twenty-eight percent of all major congenital anomalies consist of cardiac anomalies2.
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. The incidence of moderate-to-severe structural CHD in live born infant is 6–8 per 1000 live births.

The diagnosis is established by 1 week of age in 40%–50% of patients. CHD is considered as one of the leading causes of neonatal mortality.

The prevalence of congenital heart diseases in neonates has been studied thoroughly and is usually reported to be 5-8/1000 live births.

Asian race is found to be more affected than non-Asian race due to high rate of consanguineous marriages. Nearly 1/3rd of the congenital heart diseases (CHD) are critical requiring interventions in the first year of life. CHDs contribute to infant mortality significantly as 7% of the neonatal deaths are due to congenital malformations, 25% of which are cardiovascular.

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

Methods And Study Design:
The study was conducted in the Postgraduate Department of Pediatrics, GB Pant Children’s Hospital an associated hospital of Government Medical College, Srinagar. 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 and the diagnosis was confirmed by Echocardiography (2D Echocardiography). On this basis the profile of various congenital heart defects were studied.

Study Design:
It was an observational prospective study carried out in the Neonatology Section, Department of Pediatrics, GB Pantn Children 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. A suspected case was defined as:
1. Any child with SpO2 <93% at room air/or visible cyanosis
2. Congestive heart failure (CHF)
3. Murmur
4. Abnormal ECG
5. Abnormal heart sounds

Abnormal Blood Pressure
1. Differential Peripheral pulses
2. Abnormal chest X-Ray

Exclusion Criteria
1. All preterm newborns with PDA and PFO.
2. Children more than one month of age.

Details of all neonates having CHD diagnosed by echocardiography were noted in the preformed proforma. 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. 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%).

Table 3: Gender distribution of study neonates.

| Gender  | Frequency | Percentage |
|---------|-----------|------------|
| Male    | 253       | 53.7       |
| Female  | 218       | 46.3       |
| Total   | 471       | 100        |

Male:Female=1.2:1

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 4: 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       |
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 5: Types of cyanotic heart disease.

| Type of CHD                                      | Frequency | Percentage |
|-------------------------------------------------|-----------|------------|
| d-TGA                                           | 39        | 8.3        |
| TOF                                             | 23        | 4.9        |
| TAPVC                                           | 11        | 2.3        |
| HLH with interrupted aortic arch                | 9         | 1.9        |
| VSD, pulmonary atresia                         | 10        | 2.1        |
| DILV with PAH/PS                                | 17        | 3.6        |
| Truncus arteriosus                              | 3         | 0.64       |
| Critical PS with PFO/ASD R-to-L                 | 4         | 0.85       |
| Tricuspid atresia, VSD                         | 5         | 1.1        |
| Ebstein anomaly                                 | 3         | 0.64       |
| CCTGA/VSD/pulmonary atresia                    | 2         | 0.42       |
| Isomerism with complex CHD                      | 7         | 1.5        |
| Taussig-Bing anomaly                            | 3         | 0.64       |
| Total                                           | 136       | 28.9       |

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 most common cyanotic CHD was d-transposition of great arteries (d-TGA) 39 (8.3%), followed by tetralogy of Fallot (TOF) 23 (4.9%) and single ventricle (double inlet left ventricle [DILV] with pulmonary arterial hypertension/pulmonary stenosis [PAH/PS]) 17 (3.6%).

Table 6: 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%).
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. 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. The neonatal unit is the best place for screening and diagnosis of CHD.

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). wherein the cyanotic CHD constituted 31% and acyanotic 69%. Similarly, in a study by Deo B et al. (2015) 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) 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) in Nepal wherein the male-to-female ratio was 1.5:1. Humayun KN et al (2008) 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.

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) and Farooqui et al (2010). However, in studies done by Patra S et al. (2015) and Hussain S et al. (2013) 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), Khalil A et al (1994) 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.

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.

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