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CLINICAL PROFILE OF CYANOTIC CONGENITAL HEART DISEASE IN NEONATAL INTENSIVE CARE UNIT AT TERTIARY CARE HOSPITAL IN NORTH INDIA

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ABSTRACT

OBJECTIVE: To determine the clinical profile and assess the outcome of all neonates diagnosed with cyanotic congenital heart disease.

METHODS: The study was conducted prospectively in NICU, G.B Pant Children Hospital, Department of Paediatrics and Neonatalogy, GMC Srinagar from January 2016 to January 2018. All included patients were subjected to thorough clinical history, full clinical examination, initial and frequent measurement of oxygen saturation by pulse oximetry, blood gas analysis, and echocardiography

RESULTS: Total 529 cases of CHD diagnosed by Echocardiography in neonatal period were studied, out of which 280 were males and 259 were females (male:female =1.5:1). 147 (27.7%) had cyanotic CHD[CCHD], The most common type of CCHD was d-transposition of great arteries (D-TGA) (27.8%) followed by TOF(17%) and complex CHD[12.2%], All cases presented with central cyanosis and needed medical treatment whereas balloon atrial septostomy was performed in 6% of cases. 87 neonates [59.1%] improved and referred to higher centers while 40.08% expired during hospital stay.Intractable heart failure was commonest cause of death[13.6%].All neonates with HLHS ,truncus arteriosus and pulmonary atresia died

CONCLUSION: CCHD is a leading cause of neonatal morbidity and mortality. CCHD frequency was significant (27.7%) in our study population with D-TGA being the commonest type. Majority of neonates with CCHD showed survival with suitable management. Early diagnosis and referral to pediatric cardiac center for proper management will improve the outcome. Neonatologists and pediatric cardiologists should be familiar with diagnosis and management of CCHD

KEYWORDS :

INTRODUCTION

The congenital heart disease are not fixed anatomic defects that appear at birth, but are instead a dynamic group of anomalies that originates in fetal life and changes considerable during the postnatal development.[1] The incidence of moderate to severe structural congenital heart disease in live born infant is 6 to 8 per 1000 live births.[2] About 2-3 per 1000 newborn will be symptomatic with heart disease in 1st year of life. The diagnosis is established by1 week of age in 40-50% of patients. CHD is considered one of the leading causes of neonatal mortality [3]. According to a status report on CHD in India, 10% of the present infant mortality may be accounted to CHD. [4] Many cases are asymptomatic and discovered incidentally during routine health check-up. [5]

The neonates with CCHD may present with cyanosis, cardiovascular collapse, and congestive heart failure or combinations of these presentations. Pure versions of specific defects may present in some patients, but many neonates have various combinations of defects [6] Echocardiography, with Doppler and color Doppler has become the primary diagnostic tool for CCHD. In addition, it reduces the requirement for invasive studies such as cardiac catheterization [7]

MATERIALS AND METHODS

The study was conducted prospectively in NICU, G.B Pant Children Hospital, Department of Pediatrics, GMC Srinagar from January 2016 to January 2018. Echocardiography screening of all neonates suspected of having CHD was done. Admitted neonates diagnosed with cyanotic congenital heart disease (CCHD) were included in the study.

Exclusion criteria: Neonates diagnosed as persistent pulmonary hypertension of neonates (PPHN), and acyanotic CHD were excluded from the study.

METHODS

All neonates included in this work were subjected to the following: thorough clinical history, full clinical examination, initial and frequent measurement of oxygen saturation by pulse oximetry and arterial blood gas (ABG) analysis. Echocardiography has been performed by a single pediatric cardiologist at NICU on SEIMENS ACCUSON S 2000 using M-mode, Two-dimensional and color doppler cardiac imaging.

RESULT

During the 2-year study period, total admissions in our NICU with CHD were 529. Out of them 147 neonates were diagnosed with cyanotic congenital heart disease (CCHD) and included in the study. CCHD frequency was found to be 27.7% (147/529) in our NICU population. Out of CCHD cases, 90 (61.2%) were males with male to female ratio1.5:1. Their age of presentation ranged from 2 to 28 days .The most common presenting symptoms were central cyanosis (100%), followed by respiratory distress (70%), then feeding difficulties (53%).followed by shock 8.1%

Hypoxemia by pulse oximetry, [peripheral capillary oxygen saturation (SpO_2) < 85%], was detected in 90% of case, However by ABG analysis, all cases were hypoxemic [partial pressure of arterial oxygen (PaO_2) < 85 mmHg].

Table 1: Types of cyanotic congenital heart disease and their age of presentation

Type of CHD	1st week	2nd week	3rd week	4th week	Total (n)	(%)
d-TGA	26	5	3	7	41	27.8
TOF	2	6	8	9	25	17
TAPVC	3	4	3	2	12	8.16
HLH with Interrupted Aortic arch	6	3	1		10	6.8
VSD, Pulmonary atresia	2	4	3	3	12	8.16
DILV with PAH/PS	3	7	6	2	18	12.2
Truncus arteriosus		2	1		3	2
Critical PS with PFO/ASD R-to-L	2	3	-		5	3.4
Tricuspid atresia, VSD	1	1	3	1	6	4.0
Ebstein anomaly	2	1			3	2.0
CCTGA/VSD/Pulmonary atresia			1	1	2	1.36
Isomerism with complex CHD	2	2	1	2	7	4.7
Taussing bing anomaly	1	1	1		3	2.0
TOTAL	50	39	31	27	147	

d-TGA: D-transposition of great vessel, TAPVC: total anamolous pulmonary venous connection, 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.

Table 1 depicts various cyanotic CHD and age of presentation. The commonest cyanotic CHD is d-TGA 27.8%, followed by TOF (17%)

and single ventricle [DILV with PAH/PS] (12.2%). Majority of cyanotic CHD presented in first week of life n=50 [34.01%]

Table 2 depicts various therapeutic modalities ,used in neonates. All cases needed medical treatment in the form of one or more of the following [Prostaglandin E1 (PGE1) infusion, antifailure, inotropes, antibiotics, vitamin K or sodium bicarbonate].

Therapeutic modality	Number	percentage
Medical therapy [PGE1]	147	100%
Balloon septostomy	10	6%
Oxygen therapy	123	83%
Mechanical ventilation	43	29%

Table 3 depicts the immediate outcome of neonates, showing 40.8% of cyanotic CHD died within one month

Table 3

Expired	No of cases (n)	Percentage
Cyanotic	60	40.8%

In our study 20 neonates [13.5%] died due to heart failure ,followed by intractable shock (8.8%) severe hypoxemia due to closure of ductus arteriosus 6.1% ,sepsis in 6.1% .In nine neonates death was secondary to CNS,renal causes etc.[table 4]

Table 4

Complication	Number	Percentage
Heart failure	20	13.6%
Closure of ductus arteriosus	9	6.1%
Intractable shock	13	8.8%
Sepsis	9	6.1%
Others	9	6.16%

There was 36.5% mortality among the cases with D-TGA.100% mortality in neonates with HLH and Truncus arteriosus .75% mortality in neonates with pulmonary atresia [Table 5].

DISCUSSION

Congenital heart disease (CHD) has already been known as an important cause of significant morbidity and mortality in neonatal period. Neonatal unit is the best place for screening and diagnosis of CHD [8]. The objective of the present study was to describe the clinical profile and available therapeutic modalities used in the management of CCHD in NICU at tertiary care hospital.

Table 5: Mortality in various CHD's

Type of CHD	Total (n)	Expired	Percentage
d-TGA	41	15	36.5%
TOF	25	5	20%
TAPVC	12	3	24%
HLH with Interrupted Aortic arch	10	10	100%
VSD, Pulmonary atresia	12	9	75%
DILV with PAH/PS	18	2	11.1%
Truncus arteriosus	3	3	100%
Critical PS with PFO/ASD R-to-L	5	1	20%
Tricuspid atresia, VSD	6	4	66.6%
Ebstein anomaly	3	2	66.6%
CCTGA/VSD/Pulmonary atresia	2	1	50%
Isomerism with complex CHD	7	4	57.1%
Tausing bing anamoly	3	1	33.3%
	147	60	

During the two year study period 529 neonates were diagnosed with congenital heart disease of which 147 (27.7%) were cyanotic CHD, which was comparable to a study by Shah GS, et al where-in the cyanotic congenital heart disease constituted 31%[9] Similarly in a study by Bhushan Deo et al, 32.5% belonged to cyanotic group[10] Most of the cyanotic variety (34.01%) presented in the first week of life, which is comparable to study conducted by Humayun et al [11] in which the mean age of presentation of neonates with congenital heart disease was 5 days and all had cyanotic type of congenital heart disease. Hence, most of the critical and cyanotic CHD present in first week of life indicating that early detection of these neonates is critical for their survival.

In our study ratio of male-female ratio was1.5:1 which is similar to many studies by Shah GS, et al in Nepal where in the male to female ratio was 1.5:1 (9). Similarly in a study conducted by Humayun et al (11) in Pakistan, male to female ratio was 1.7:1.

In our study the most common presenting symptoms were central cyanosis (100%), followed by respiratory distress (70%), and feeding difficulties (53%) in the form of poor suckling, poor feeding and interrupted feeding common presentations whereas shock was present in 8%. This was in agreement with other studies done by Humayun and Atiq [11]and Dorfman et al [12]

In our study 96% of cases were hypoxemic (SpO_2 < 90%) by pulse oximetry. Meanwhile all cases were hypoxemic (PaO_2 < 85 mmHg) by ABG analysis So pulse oximetry is a good, easy, non invasive, and cheap method for early diagnosis and detection of degree of hypoxemia but it is less accurate and should be confirmed by ABG analysis. This was in line with a study done by Riede et al [13] as they found that pulse oximetry was a good screening tool for detection of duct-dependent pulmonary circulation, as it has detected 100% of cases.

Transposition of great arteries (d-TGA) was the most frequent type of Cyanotic CHD in our patients with a frequency of 27.8%, followed by TOF (17%) and DILV with PAH/PS (12%). Our work was in agreement with studies done by Islam et al.[14] and Farooqui et al.[15]. However in studies done by Patra et al.[16], Hussain et al[.17] the most common types of Cyanotic CHD were Tetralogy of Fallot (TOF) followed by D-TGA. This difference can be because of inclusion of only neonates in our study while other studies included older children and usually Tetralogy of Fallot presents after few months of life. Higher incidence of complex CHD in our study can be due to high rate of consangious 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.

All cases needed medical treatment in the form of one or more of the following (Prostaglandin E1 "PGE1", antifailure, inotropes antibiotics, vitamin K, sodium bicarbonate). oxygen therapy was used in 123 [82%] of cases. 43 neonates (29%) required mechanical ventilation. Balloon atrial septostomy was performed in 6% of cases. 87 neonates of our cases (59.1%) were improved by using these modalities. Neutze et al.[18]stated that PGE1 infusions have been used to increase ductal patency in 11 cases with CCHD, all of them showed a satisfactory increase in oxygen saturation attributed to dilatation of the ductus.

In our study 40.8% of neonates with cyanotic CHD died within one month. This mortality rate is comparable to study by Humayun et al 36.4% [11], 50% by Ravilala et al[19]This mortality rate is higher than in other studies by Shah et al which showed mortality rate of 20% (9). The difference may be due the difference in the study population and the limited availability of cardiac facilities.

The most common causes of death were intractable heart failure, followed by profound shock [8.8%].severe hypoxemia due to duct closure . In the study done by Humayun and Atiq 11, the causes of death were surgical complications in 2 (4.5%) cases and medical problems in 14 (31.8%) babies such as sepsis and pneumonia and associated extra-cardiac malformations.

Mortality rate was high in critical CHD,100% in HLH,truncus arteriosus and 75 % in pulmonary atresia.High mortality ratein critical CHD has been highlighted in many studies like Selma Alves et al[20].In this study the lethality attributed to critical congenital heart diseases was 64.7%, with proportional mortality of 12.0%.

CONCLUSION

Congenital heart disease (CHD) is the most common congenital malformation presenting in the neonatal period Cyanotic congenital heart disease (CCHD) is a leading cause of neonatal morbidity and mortality with 60% of neonates expiring in neonatal period suggesting dire need of good paediatric cardiac centre

Limitations of the study: Our study had some limitations as the study included only a single center. The incidence of CHD could not be calculated as we don't have data regarding total live birth. It is a hospital based study and will not reflect the true incidence or prevalence of CHD in our community.

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