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Original Article

Study of Clinical Profile and Immediate Outcome of Congenital Heart Disease in Neonates at a Tertiary Care Hospital in India: A Single Center experience

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ABSTRACT

Background: Data on congenital heart diseases in neonates, especially critical congenital heart disease, varies worldwide. Developing countries have higher mortality than developed countries. To study the clinical profile of congenital heart diseases in neonates.

Methods: Prospective observational study. Neonates admitted to a tertiary care hospital with congenital heart disease. Clinical details, investigations, and management of neonates admitted to a tertiary care hospital were studied prospectively.

Results: Among the study neonates, 60% had acyanotic heart disease, the most common being ASD, while 40% had cyanotic heart disease, the most common being TGA—outborn cases comprised 60% of the total neonates. The most common presenting features were fast breathing and cyanosis; the most common clinical sign was murmur(59%). Among the study neonates, 15 cases had low calcium levels. Definitive surgery was done in 21 neonates, of which 7 succumbed. The staged repair was done in 9 neonates, of which 2 succumbed. Of 12 TGA cases,10 underwent definitive repair, and 7 did well at follow-up. Among the 10 TOF babies, 6 underwent staged repair. Out of the 7 cases of CoA, 6 underwent definitive surgery, and 1 expired. Among Critical CHD, mortality was 23.3%.

Conclusion: The most common cyanotic heart disease was ASD, and the most common cyanotic heart disease was TGA. Surgery was done in 30 cases, out of which 9 succumbed, and mortality among surgical cases was 30%. At follow up TGA cases with definitive surgeries were doing well. Mortality was lower compared to other studies in India. Hypocalcemia was observed in 16.6% of the CHD cases, which needs to be studied in the future.

Keywords: Cardiac murmur, Cardiac surgical procedures, Congenital, Cyanosis, Heart defects, Neonates

Introduction

Congenital heart diseases (CHD) refer to structural heart diseases at birth. These are primarily seen in neonates, infants, or children (1). CHD occurs in 0.8% of live births. Congenital cardiac defects have a broad spectrum of severity in infants: approximately 2-3 in 1000 newborn infants will be symptomatic with heart disease in the 1st year of life (2). The diagnosis is established by 1 week of age in 40-50%patients with congenital heart disease and by 1 month of age in 50-60% (2). A study by Hoffman on the incidence of congenital heart disease showed the earlier

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incidence of 4 to 5/1,000 live births, (3) but the incidence has increased up to 12 to 14/1,000 live births.[3] The low incidence was due to the non-availability of good echocardiography, and the diagnosis was not made if it was not evident clinically and cardiac catheterization was needed. Prenatal diagnosis leads to abortion, leading to reports of low incidence of CHDs (4). However, a recent study by Bhat et al (5) showed a prevalence between 8.5 and 13.6/1000 live births. In another recent study from China, the incidence of CHD is mentioned as 8/1000 live births globally (6). Ventricular septal defect (VSD) is the most common CHD, followed by Patent ductus arteriosus(PDA). Other CHDs include atrial septal defect (ASD), pulmonary stenosis (PS), and Coarctation of aorta(COA). It is observed that maximum CHDs were detected in the first year of life concerning later years of life. A prevalence of 5.3 per 1000 population was found in a study by Naik et al in Kashmir. VSD was the most common lesion (30.1%), followed by PDA in 21.6 % and ASD in 20.2%. Among the Cyanotic CHD, Tetralogy of Fallot(TOF) was the most common CHD (8.0%) (7). Friedberg et al. observed that the two most influencing factors in prenatal diagnosis of CHD depend on the expertise and experience of the medical practitioners doing the screening ultrasound and the type of heart disease (8). Infants with critical CHD present immediately after birth, often with life-threatening features requiring immediate intervention (9). However, some cases with CHD may appear normal on routine examination, and features of critical CHD are not evident until after discharge. Depending upon the presence of Patent ductus arteriosus, the timing of diagnosis of CHDs differs. Closure of the PDA within the initial days can present with life-threatening complications such as metabolic acidosis, seizures, shock, and end-organ damage. Neonates with critical CHD undiagnosed postnatally, the risk of mortality is increased to 30 percent (10). Before pulse oximetry was used routinely for screening, 30 % of infants born with critical CHD were undiagnosed at discharge (11). There are two reasons for separate guidelines for treatment in our country. First, the result of surgery is different from the Western world due to higher infections, underweight, anemia, and several co-morbidities in our children. Secondly, most often, late presentation occurs; this calls for modification in treatment guidelines to optimize the outcome.

This study aims to study the clinical profile and immediate outcome of congenital heart diseases in neonates at a tertiary care hospital in India.

Methods

It is a prospective observational study from April 2020 to August 2021 over 17 months. The Study population included all neonates admitted to NICU(inborn and outborn and Postnatal wards) at a tertiary care hospital with congenital heart disease from April 2020 to May 2021; the cases were followed up for 3 months till Aug 2021. They were studied for clinical profile and their immediate outcome. The protocol was submitted to the institution's Ethics Committee, and IEC clearance was sought. IEC Number: IEC 897-2019, CTRI Reg: CTRI/2020/04/024525. Informed consent was obtained from parents after they explained the study's purpose to them. Neonatologists diagnosed the cases and referred them to pediatric cardiologists and cardiothoracic surgeons. Data collected in the form of history as well as clinical examination and investigations (Chest x-ray, ECHO done in suprasternal, subcostal, apical, and long axis parasternal view, and CT angio if done) was recorded according to the format of the proforma designed for this study. Echo and chest X-ray was done in all cases, and CT angio was done in 17 cases. Treatment given was noted. Details of surgery, if done, with a postoperative period were recorded. Length of hospital stay and condition at discharge/mortality were recorded. The study population was assessed throughout the study period to infer the outcome of Congenital heart disease. The outcomes were evaluated as follows: 1) Neonates who received medical treatment and their outcome. 2) Neonates who underwent surgical intervention and their follow-up. 3) Outcome at the end of the 3-month follow-up period.4) Neonates who expired during the course of study.

Descriptive Statistics was used in this study as frequencies and percentages because it is a descriptive study.

Ethical Approval

The protocol was submitted to the institution's Ethics Committee, and IEC clearance was sought. IEC Number: IEC 897-2019. CTRI Reg: CTRI/2020/04/024525.

Results

The total number of Neonates admitted In NICU and Postnatal wards during the study period was 3315 (Inborn and Outborn -3315 from April 2020-May2021). Neonates were followed up for 3 months till Aug 2021. Among these, the total

Table1. Types of congenital heart disease among the study population

Acyanotic chd	N= 54 (60%)
Asd	22 (40%)
Vsd	11(20%)
Pda	10(18%)
Coa	7(13%)
Asd+vsd	2(3.7%)
Asd+ pda	2(3.7%)
Cyanotic chd	N=36(40%)
Tga	12(33%)
Tof	10(27%)
Truncus arteriosus	2(5.5%)
Tapvc	2(5.5%)
Tricuspid atresia	2(5.5%)
Hlhs	2(5.5%)
Dorv+ps+complete av septal defect	1(2.7%)
Ebsteins anomaly	1(2.7%)
Peripheral ps	1(2.7%)
L tga+asd+vsd+ps	1(2.7%)
Dysplastic pulmonary valve +ps	1(2.7%)
Cchd + ps+vsd+asd + dextrocardia	1(2.7%)

number of babies diagnosed with congenital heart disease was 90(2.7%).

Among the study population, 60% of the neonates presented with Acyanotic CHDs; ASD was seen in 22 cases, followed by VSD in 11 cases, PDA in 10 cases, and Coarctation of Aorta in 7 cases. 36 neonates presented with Cyanotic CHDs. Transposition of Great arteries (TGA) was the most common, followed by Tetralogy of Fallot(TOF) in this study. (Table 1). All Cyanotic heart disease cases were referred cases postnatally except for 3 cases diagnosed antenatally and referred to our hospital for delivery (1 case of double outlet right ventricle (DORV)+ pulmonary stenosis(PS),1 case of Tricuspid Atresia (TA), 1 case of Total anomalous pulmonary venous connection(TAPVC) and PS).

In this study,60% were outborn cases since ours is a referral center. Demographic characteristics of the study population are given in Table 2. All critical congenital heart diseases except one case of coarctation of aorta and one TGA were diagnosed in 1st week of life. 87% of the babies were born to mothers aged 25-35. Anomaly scan was done for all 90 cases. Among the 90 anomaly scans, cardiac anomalies were detected in 6 babies. Among 6 abnormal anomaly scans, 5 were done outside and one at our hospital(case of Ventricular foci and ASD). Fetal ECHO was done for 38 cases, of which 25 were reported as normal. Among the 90 babies, abnormal foetal ECHO was 13. Among the foetal ECHO reported as normal (25cases),7 babies had TGA,3 cases had VSD, 2 cases had TOF,1 case had Ebstein anomaly,6 cases had ASD,5 cases had PDA, and 1 case had dextrocardia in postnatal ECHO. Sensitivity for fetal echocardiographic screening ranged from

Table 2. Demographic characteristics of the study population

Chracteristics	N=90	Percentage
Gender	Male-50	55
	Female-40	44
Place of Delivery	Outborn- 54	60
	Inborn- 36	40
Period of Gestation	>37 weeks- 57	63
	>34 to 37 weeks- 12	13
	32 to 34 weeks- 8	9
	28 to 32 weeks- 7	7.7
	<28 weeks - 6	6.6
Birth weight	2.5 to 4kgs- 55	61.1
	1.5 to 2.5kgs- 21	23.3
	1 to 1.5kgs- 5	5.5
	<1kg- 9	10

30% to 80%. Among the abnormal fetal ECHO reports(n=13),6 had similar post-natal ECHO reports, and a difference was seen in 7 cases. Consanguinity history in the parents of neonates with CHD in our study was 3.3%. In our research, Gestational Diabetes mellitus(GDM) was the most common risk factor in the antenatal period. Among the mothers, GDM was seen in 14 women. In one case of Ebstein anomaly, the mother was obese and had GDM and hypothyroidism.

In the present study, the most common presenting complaint was hurried respiration and cyanosis. In some cases, an overlap of symptoms was seen; among 12 cases of TGA,3 presented with cyanosis and low saturation, cyanosis and respiratory distress in 2 cases of TOF, and cyanosis and seizure in one case of TGA. The majority of acyanotic CHD cases were asymptomatic and were detected by incidental murmurs. Among the 90 neonates, 35 (39%) were asymptomatic at presentation. Murmur was the most common clinical finding in the study. Among the neonates with CHD,59% of neonates presented with murmur. Clinical features at presentation among the studied neonates is described in table 3. Most were systolic murmurs,

Table 3. Clinical features at presentation

Symptoms	N=90(%)
Fast Breathing	23(25%)
Cyanosis	23(25%)
Feeding Problem	6(6.6%)
Seizure	2(2.2%)
Shock	1(1.1%)
Signs On Examination	N=90(%)
Murmur	53(59%)
Low Saturation	15(16%)
Dysmorphic Features	11(12%)
Cardiomegaly	7(7.7%)
Hepatomegaly	5(5.5%)
Feeble Femoral Pulses	3(3.3%)
Hypotonia	2(2.2%)

Table 4. Management of chd in the study population

Management	ASD N=22	VSD N=11	TGA N=12	TOF N=10	CoA N=7	TAPVC N=2	Truncus Arteriosus N=2	Hlhs N=2	Pda N=10	Tricuspid Atresia N=2
Medical	0	6	0	0	0	0	0	0	9	0
Stagedrepair	0	0	1	6	0	0	1	0	0	1
Definitive surgery	0	1	10	0	6	2	1	0	1	0
Conservative	22	1	0	3	0	0	0	0	0	1
Expired			3	1	1	1	1	0	2	0
Dama	0	0	1	0	1	0	0	2	0	0

ejection systolic murmur in 9, pan systolic murmur in 14, and continuous murmur in 10 cases. Renal anomalies had the most common association with CHD in the present study, followed by Tracheo esophageal fistula(TEF) and Gastrointestinal anomalies. Among the 12 renal anomalies,7 were detected antenatally (2 in anomaly scan,4 in 7th month and 1 at 5th month). Among the study population, 4 babies were suspected of having Digeorge syndrome I/v/o hypocalcemia, CHD, and dysmorphic features; among the 4 cases, 2 of them opted to do genetic testing at a later date; in 1 case, genetic testing was reported as normal(VSD, hypocalcemia, dysmorphic features). Trisomy 21(2 cases) and Trisomy 13(1 case) were diagnosed by karyotyping. Among the 90 cases,15 cases were found to have hypocalcemia. All cases were term AGA babies except for 2 babies (Late preterm and early preterm). Three babies had seizures (Calcium levels were 6.1mg/dl, 6.6mg/dl, 7.5mg/dl). Among 15 cases with low calcium levels ,(5-6mg/dl were 2 ,with 6-7mg/dl were 3 And 7-7.5mg/dl were 10). Low calcium and CHD should be studied in the future.

CT Angio was done in 17 cases. They include 6 cases of TOF,1 baby with VSD,2 babies with truncus arteriosus,4 babies with coarctation of Aorta, 2cases with TAPVC and one Heterotaxy syndrome with CCHD and one Dysplastic Pulmonary Valve.

Definitive surgery was done in 21 neonates, out of which 7 succumbed. The staged repair was done for 9 cases, and 2 succumbed. All the 22 ASD cases were managed conservatively. Out of 12 TGA cases, 10 underwent definitive surgery, 3 expired, 1 case had staged repair, and 1 case went discharged against medical advice (DAMA). Among the 10 TOF babies, 6 underwent staged repair, and 3 cases were managed conservatively, and 1 case expired. Out of the 7 cases of CoA, 6 underwent definitive surgery, 1 case expired, and 1 case went DAMA. Babies discharged against medical advice were 5 in number[2 cases of Hypoplastic left heart syndrome(HLHS), one case of TGA with TEF and 1 case of COA and 1 case of Heterotaxy syndrome with complex congenital heart disease(CCHD)]. Management of CHD among the study population is described in table 4.

Postoperative complications: Out of the total surgical cases(30),right upper lobe collapse was seen in 6(20%) cases, sepsis was seen in 6(20%) cases, wound dehiscence in 3(10%), seizures in 3(10%), ventricular bigeminy in 1(3.3%) cases and hypertension in 1(3.3%) case.

In the present study mortality rate(total) was 10%. Among Critical CHD mortality was (23.3%).

The mean NICU stay for a surgical case was 27 days.

Follow up

Total cases followed up were 69. Out of the total 90 cases, Expired and DAMA cases were 9 and 5 respectively and 7 cases were lost for follow up. Among 2 cases with Tricuspid Atresia who underwent repair(BD Glenn), attained age appropriate developmental milestones and adequate weight gain. Among 12 cases of TGA, 10 cases under went definitive repair and 7 cases were doing well at follow up. Among 2 cases of TAPVC-1 baby expired due to surgical complication, the other child had adequate weight gain. Among 2 cases of Truncus Arteriosus-1 baby succumbed due to surgical complication and one baby had poor weight gain at follow up. Among 2 cases of ASD+PDA,both attained spontaneous closure. Among 2 cases of ASD+VSD- both were on follow up. Neonate with Ebstein anomaly had adequate weight gain .spo2 at follow up was 86%. Congenital dysplastic PV with Severe PS case had Inadequate weight gain at follow up. Atrioventricular septal defect(AV) septal defect+ Double outlet right ventricle (DORV)+ Pulmonary stenosis(PS)case was followed up and inadequate weight gain was seen.

Discussion

In the present study male to female ratio was 1.2:1.In the study conducted by Ravilala et al M:F ratio was 1.1:1 (1). Many studies done showed

male preponderance corresponding to the present study (11, 12). In this study, 82% of the cases were diagnosed in 1st week of life. In the study by Ravilala et al among 60 cases, half of the cases presented in the first week (45%) followed by those in fourth week (40%), third week (8%), second week (7%) in decreasing order (1). In the study by Ravilala et al, Out of 60 cases studied, 24 cases were born to consanguineously married couples (1). In our study, consanguinity was seen in 3.3% of cases. A study done by Lindsey E et al. suggests that women with GDM may have a similar risk for the occurrence of fetal CHD as women with pre-existing Type 1 diabetes (13). In a study from Iran, maternal obesity, history of abortions, and consanguineous marriages were found to be significant risk factors for CHD (14). In another study, there was a high correlation between diabetic mothers and CHD (15). Ravilala et al. studied 60 newborns; the commonest presenting complaint was hurried respiration (68), followed by a feeding problem, cyanosis. Breathlessness was the most common clinical symptom identified by a study by Mesharam et al (16). In the study from China, the most common symptoms were fast breathing and cyanosis in ACHD and CCHD, respectively. In our study, the most common presenting features were hurried breathing and cyanosis, similar to the study from China. Among the neonates with CHD 72%presented with murmur in the study by Ravilala et al.[1] In our study, 59% of the neonates presented with murmur. In the study from China by Song L et al most common finding was murmur((90.2%). In the best centres mortality for arterial switch operation is now 1%, (17) considering ours as a developing country, mortality of arterial switch operation was 30%. In a series from Toronto, 4% of neonates presenting with simple TGA died before surgery (18). The overall mortality rate among Indian studies like the study by Ravilala et al. was 50%, (1) Humayun and Atiq was 36.4% (19), and another study by Miret al (20) showed 40.8% mortality. In our study, mortality among the total 90 cases was 10%, and among critical congenital heart disease was 23.3%. CHD mortality has a greater variability worldwide. Low industrialized or developing countries have higher mortality rates than developed countries. In a study done in Malaysia, the overall mortality rate was 34.8% among CCHD, with a median age at death being 2.7 months, 17% of patients died before surgery, 9.8% died within 30 days of cardiac surgery, and 11.5% died after 30 days of surgery. No patient with HLHS in this study was offered surgical treatment (21). In our study also, 2 cases of HLHS went DAMA in v/o poor prognosis following surgery. In a study done in New Zealand between 2006 and 2010, mortality rates were 18% in critical CHD versus 8% in noncritical CHD; mortality among early diagnosis was 12%, whereas among those with late diagnosis was 27% (22). In this study, they stated that antenatal detection reduced mortality among CCHD. In our study, out of the total 90 cases, fetal echo were done in 38 cases, and 13 were abnormal(34.2%). In a study done in Norway, mortality among severe CHD was 10%, of whom 58% died before surgery (23).

Limitations of the study: We followed up the cases for 3 months; long term follow up is necessary to see the long-term outcome of these cases.

Conclusion

Most common acyanotic heart disease was ASD and most common cyanotic heart disease was TGA. Surgery was done in 30 cases, out of which 9 succumbed, and mortality among surgical cases was 30%. At follow up TGA cases with definitive surgeries were doing well. Mortality was lower compared to other studies in India. Hypocalcemia was observed in 16.6% of the CHD cases, which needs to be studied in the future.

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None.

Conflicts of interest

There is no conflict of interest of the authors and / or family members regarding this study.

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