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Original Research Article

Pattern of critical congenital heart disease and their outcome in Northern India: a medical school based study

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ABSTRACT

Background: Critical congenital heart disease (CCHD) comprise a group of morphologically heterogeneous disorders which have one thing in common that early surgical or catheter interventional therapy is mandatory to achieve survival. Our aim in this study was to look for pattern of Critical congenital heart disease in a tertiary centre in Northern India.
Methods: Our study was a hospital based prospective study conducted on all patients with suspected congenital heart disease who were brought to neonatology section of a tertiary care hospital. The methods employed for case detection used were clinical examination, pulse oximetry, chest radiograph, electrocardiogram and echocardiography.
Results: Forty two neonates with suspected congenital heart disease were admitted in our NICU (Neonatal intensive care unit) and nine cases of CCHD were diagnosed. Twenty seven cases were having Non Critical cardiac lesions whereas six cases had respiratory illness. Out of nine cases of CCHD seven patients died while two are alive.
Conclusions: With the advent of more screening tools the prevalence of CCHD has increased and the mortality associated with this condition is also very high. It is thus imperative to go for early surgical correction to achieve survival.

Keywords: CCHD, Echocardiography, Neonates

INTRODUCTION

Congenital heart diseases are relatively common with a prevalence of 5-10 in every 1000 live births.[1] With improvements in diagnosis and treatment, the outlook for newborns with congenital heart diseases has changed considerably. But these malformations still contribute to significant morbidity and mortality in this age group. Children with congenital heart diseases are at approximately two times higher risk of mortality in the first year of life.[2]

Newborns with Critical congenital heart disease (CCHD), a group of morphologically heterogeneous disorders, have one thing in common that they need early surgical or catheter intervention for their survival. In the current era, surgeries are done for repair or palliation for nearly all types of congenital heart malformations.[3] Intervention needs to be done in first weeks of life so as to optimize hemodynamics and to prevent end-organ injury associated with delayed diagnosis. A timely recognition of CCHD could improve outcome, so it is important to identify and evaluate strategies to enhance early

detection. A study in California reported 2 deaths per 100000 live births related to delayed diagnosis of CCHD.[4]

Among sequelae of CCHD is neonatal hemodynamic compromise, the most important long term effect relate to the consequences of brain injury from ischemia and reperfusion, because the brain has the highest oxygen requirement than any other organ.

Children with CCHD are reported to experience more frequent impairments in motor function, speech and language, visual motor perceptual function and executive function, as well as increased use of special services.[5-13] Neonates with CCHD may be diagnosed on the basis of physical examination findings, such as heart murmurs, tachypnea or overt cyanosis. Apart from physical examination, pulse oximetry, chest radiography, electrocardiogram and echocardiography are useful tools in identifying many cases of serious congenital heart disease postnatally.

METHODS

Our study was hospital based prospective study over a period of three years from 1st March 2016 to 28th February 2019. The study was conducted in the department of Pediatrics and neonatology SKIMS Medical College Srinagar, Jammu and Kashmir, India which is a tertiary care hospital in Northern India.

The hospital has the catchment area of both rural and urban population. All neonates who were brought to the OPD/Nursery and NICU were screened for congenital heart disease.

Inclusion criteria

All neonates who presented with any of symptom plus Murmer

- Overt cyanosis,
- Tachypnea (respiratory rate >60/min),
- Grunting, flaring,
- Retractions,
- Murmur,
- Active precordium or
- Diminished pulses were taken as patients with suspected congenital heart disease and were enrolled in the study.

Exclusion criteria

All neonates who presented with,

- Normal APGAR score at birth
- Normal general and systemic examination
- No or minimal cyanosis
- Neonates with probable sepsis

Statistical analysis

The analytic tools that were used in the study for the presentation and interpretation of results were mostly those of time tested tabular analysis and percentages, besides certain specific statistical indices and tests. Data was compiled and all the analysis was done through SPSS-10 (Statistical Package for Social Science) software package.

Approach

All patients with suspected congenital heart disease were screened for congenital heart disease by following tools;

- Pulse oximetry,
- Chest radiograph,
- Electrocardiogram
- Echocardiography

The cardiac lesions were divided into two broad groups- Non Critical Cardiac lesions which does not pose any immediate threat to life and Critical Cardiac lesions which were incompatible to life if not corrected by surgical or catheter intervention.

RESULTS

Our study was a hospital based prospective study conducted on all patients with suspected congenital heart diseases who were brought to neonatology section OPD/Nursery/NICU of our tertiary care hospital over a period of three years. A total of 1800 patients were brought to hospital and 42 neonates with suspected congenital heart disease were screened for congenital heart disease, 9 cases of CCHD were diagnosed giving prevalence of CCHD as 9 in 1800 (0.5 %). Twenty seven (27) cases were having Non Critical cardiac lesions while whereas 6 cases had Respiratory illness. Out of 9 cases of CCHD seven patients died while two are alive.

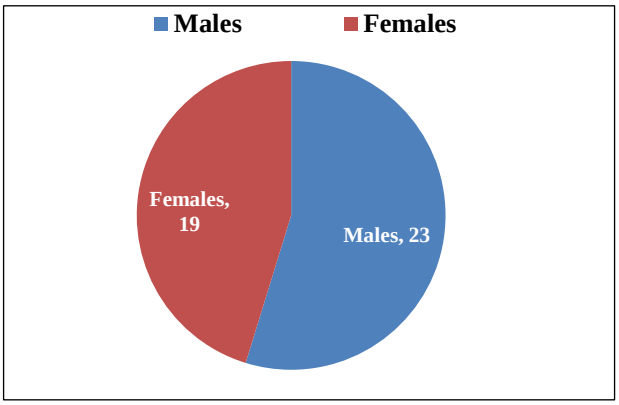

Figure 1: Case distribution as per sex.

The suspected congenital heart disease cases comprised of 23 males and 19 females (Figure 1). Neonates with first birth order were 17 cases, second, third and fourth or

greater birth order was seen in 11 cases, 6 cases and 8 cases respectively (Figure 2).

In this study 25 (59.5%) cases were from rural areas and 17 (40.5%) cases belong to urban area. Consanguinity was seen in 6 cases and family history of CHD was seen in only one neonate. Echocardiography was taken as gold standard for case detection, which detected congenital heart disease in 36 cases and ruled out in 6 cases. Among 36 cases, 9 had Critical congenital heart disease (CCHD) while the rest had Non Critical Cardiac lesions (Table 1 and 2).

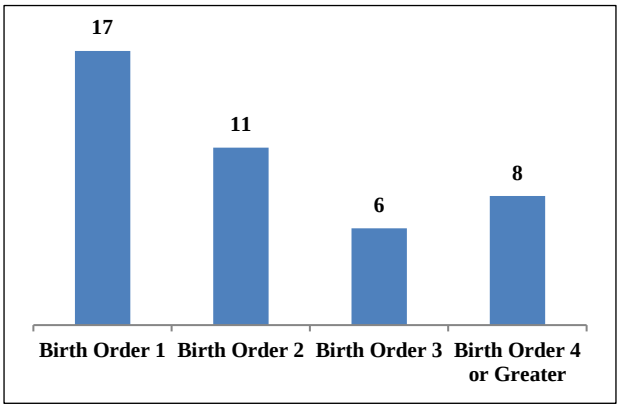

Figure 2: Case distribution as per Birth Order

Table 1: Patients with Critical Congenital Heart Disease (CCHD).

Echocardiography findings	Number of cases	Percentage
TGA/VSD	2	22.2
Truncus Arteriosus	1	11.1
Tricuspid Atresia/VSD	1	11.1
TOF	2	22.2
HLHS	1	11.1
TAPVD	2	22.2
Total	9	

TGA=Transposition of great arteries, TOF=Tetralogy of Fallot, HLHS=Hypoplastic left heart syndrome, TAPVD=Total Anomalous Pulmonary Venous Drainage

Table 2: Patients with non-critical cardiac lesions.

Echocardiography findings	Number of cases	Percentage
ASD	12	44.4
ASD/PDA	4	14.8
VSD	5	18.5
VSD/PDA	2	7.41
PDA	4	14.8
Total	27	

[ASD=Atrial septal defect, VSD=Ventricular septal defect, PDA=Patent ductus arteriosus]

DISCUSSION

Critical congenital heart disease (CCHD) is a term that refers to a group of serious heart defects that are present from birth. These abnormalities result from problems with the formation of one or more parts of the heart during the early stages of embryonic development. CCHD is often clinically unapparent until the fetal circulatory pattern transitions to the postnatal pattern, which includes closure of the ductus arteriosus during the days following birth. Because prenatal ultrasound examinations and newborn physical examination fail to identify a significant number of CCHD cases, late diagnosis may occur. Delayed diagnosis can result in significant morbidity, permanent injury of vital organs including the brain, and in some cases, death. Well-designed screening of newborns improves detection of CCHD before it becomes clinically apparent.

Early detection of major congenital heart defects (i.e. those leading to death or requiring invasive intervention before 1 year of age), might improve the outcome of newborn babies[14]. Improvement with early detection is critical for duct-dependent lesions in which closure of the ductus arteriosus can result in acute cardiovascular collapse, acidosis and death.[15-17]

Male predominance found in our study has been documented in previous studies also.[18] This may highlight a possible change in genetic substrate in South Asian population resulting in higher incidence in males rather than equal gender distribution in Western population. Another factor is cultural wherein a male gets more attention and is more likely to be brought to attention earlier than a female child.

Almost 59.5% children belonged to rural areas, a figure consistent with the general population, figures of rural population of Pakistan (62%) as well as South Asia (65%).[19] This finding reiterates the fact that CHD is equally distributed in urban and rural populations and highlights the importance of provision of screening facilities in rural areas as well as early detection.

In the current era, surgeries are done for repair or palliation for nearly all types of congenital heart malformations[3]. Intervention needs to do in first weeks of life so as to optimize hemodynamics and prevent end-organ injury associated with delayed diagnosis. A timely recognition of CCHD could improve outcome, so it is important to identify and evaluate strategies to enhance early detection.

Screening for congenital heart defects mainly relies on mid trimester ultrasound scan in which the fetal heart is imaged and postnatal physical examination that includes assessment of pulses and heart sounds and inspection for cyanosis, suspected cases are then evaluated with pulse oximetry, chest radiography, electrocardiogram and echocardiography.

Out of 1800 neonates, only 9 cases of CCHD were detected. Thus, prevalence of CCHD was found to be 1 in 200 (0.5%). Our study found results similar to Tautz et al, (0.1 %) and by Arlettaz et al, (0.33%).[20,21] Twenty seven (27) cases were having Non Critical Cardiac lesion where as six (6) cases had respiratory cause for their illness. Out of nine (9) cases of CCHD four cases were operated but only two achieved survival, which included one TGA and other TOF patient. All other patients of CCHD who were managed conservatively died in their first month of life.

Data was collected from a single tertiary care hospital setting and that was the study limitation. It only included patients who did reach a tertiary care facility and could not account for critical conditions and could not make it to such facility.

CONCLUSION

With the advent of more screening tools the prevalence of CCHD has increased and the mortality associated with this condition is also very high. It is thus imperative to go for early surgical correction to achieve survival.

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