

## Clinical Profile, Pattern of Presentation and Short Term Outcome of Cyanotic Congenital Heart Disease in Neonates

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| Received: 04.10.2020 | Accepted: 15.10.2020 | Published: 17.10.2020

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## Abstract

## Original Research Article

**Background:** Cyanotic heart disease refers to a group of many different heart defects that are present at birth (congenital) that results in a low blood oxygen level, many of which are life threatening in neonatal period necessitating early diagnosis and prompt treatment. Thus, by early intervention for critical cyanotic congenital heart diseases in neonatal period or early infancy, children can survive beyond the pediatric age. **Aim of the study:** To determine the clinical profile and pattern of presentation and short term outcome of cyanotic congenital heart disease in neonates. **Methods and Materials:** An observational type of cross-sectional study was conducted in the Department of Pediatric Cardiology of Dhaka Shishu hospital between the study period of July to December 2019 where total 74 newborn with cyanosis were recruited from outpatient and inpatient departments. Cyanotic congenital heart disease was diagnosed with the help of clinical examination and investigations finding and associated risk factor and treatment and short term outcome was documented. **Results:** Among the 74 neonates 58 (78%) were male and 16(22%) were female and male : female ratio was 3.6 :1. Age of presentation of majority of patient (n=36) was 16-28 days whereas only 3 patient presented in between 0-2 days. The neonates mainly had Transposition of Great Arteries (TGA) (n=22), Tetralogy of Fallot (TOF) (n=12), Tricuspid Atresia(n=7), DORV(n=7), Total Anomalous Pulmonary Venous Return(n=5), Hypoplastic Left Heart Syndrome(n=3). The median age of presentation of common cyanotic congenital heart defects like TGA, TOF, Tricuspid Atresia, DORV are 12-15 days. Median age of presentation for TAPVC was highest that is 23 days and Ebstein anomaly was lowest that is 8 days. In this study all of the study neonates (n=74) had cyanosis. Out of 74 patient 69(93%) had tacyhpnea, 61(82%) had tachycardia, 38(51%) had murmur, 26(35%) had hepatomegaly. Crepitation, rhonchi and oedema were found respectively 34(45%), 24(32%) and 19(25%) patients. 2 patient (2.7%) presented with shock. In this study it has been seen that majority of the patient that is 54 (72%) were term neonate. Family history of CHD were found in 9 (12%) of patient and consanguinity was found in 31 (41%) of patient. Other risk factors found were maternal diabetes mellitus in 19 (26%) patients, maternal hypertension in 28 (37%) patients, infection during pregnancy in 15 (20%) patient and parental smoking in 39 (53%) patients In this study 9 (12%) patient received prostaglandin, 21 (28%) required use of Iontrops, antifailure drugs were used in 36 (48%) patients and emergency procedures were performed in 13 (17%) patients. Regarding outcome, 11 (14%) patient required ventilatory support, 19 (25%) had an ICU stay and death occurred in 6 (8%) patients. **Conclusion:** Cyanotic congenital heart disease in neonates is often undiagnosed which is main the cause of high case fatality. Development of neonatal screening program for early detection of such cases and prompt measure can surely save life of many newborns. Therefore gathering knowledge about the clinical profile and pattern of presentation of cyanotic congenital heartdisease in neonates in our country's perspective will definitely help to avoid complication, reduce mortality and improve quality of life of the newborns.

**Keywords:** Cyanotic congenital heart disease, neonates, clinical profile, outcome.

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## INTRODUCTION

Congenital heart disease (CHD) is the most common congenital problem in children. In order to avoid complications, reduce mortality and for proper

management early detection of congenital heart disease is of utmost importance [1]. Congenital heart diseases are mainly grouped as Acyanotic and Cyanotic. Acyanotic heart disease includes: Ventricular septal

defect, Atrial septal defect (ASD), Patent ductus arteriosus (PDA), Coarctation of aorta (COA) and Aortic stenosis (AS). Cyanotic heart disease includes Tetralogy of fallot (TOF), Transposition of the great vessels (TGA), Total Anomalies pulmonary versus return (TAPVR), Pulmonary atresia, Hypoplastic left heart syndrome etc [2]. Cyanotic congenital heart diseases are critical congenital heart disease, that cause serious and often life threatening symptoms and accounts for more deaths than any other type of congenital malformation [3, 4]. Nearly 4,800 babies born each year in the United States have CCHD, and nearly 280 infants with unrecognized CCHD are discharged annually [4]. CCHDs often go undetected because some babies will appear healthy at first and are discharged from the hospital before any defect is detected. However, due to the severity of these conditions, immediate follow up to determine the best method of treatment or intervention is necessary. CCHD is associated with hypoxemia (insufficient levels of oxygen in the blood or tissues) and among infants during the newborn period, hypoxemia represents 17-31 percent of all CHDs [5, 6]. The primary targets of CCHD screening include: Hypoplastic left heart syndrome, Pulmonary atresia (with intact ventricular septum), Tetralogy of Fallot, Total anomalous pulmonary venous return, Transposition of the great arteries, Tricuspid atresia, and Truncus arteriosus. Infants with these conditions require surgical or catheter intervention within the first few days or first year of life. Without early diagnosis or treatment, infants with CCHDs are at significant risk of morbidity or mortality due to physiological changes of the heart (e.g, closing of the Ductus arteriosus) [8]. In the United States an estimated 4,800 babies born each year have CCHD, and nearly 280 of those infants may be missed by routine newborn cardiac screening [9]. Prenatal ultrasound has been used for many years in advanced countries for screening of congenital abnormalities and plays an important role in identifying CCHD but this scenario is quite different in a developing country like Bangladesh [10]. This is especially true for critical congenital heart disease, a group of morphologically heterogeneous disorders which have in common that early surgical or catheter interventional therapy is mandatory to achieve survival. Clinical examination is unable to detect all forms of CHD [11]. Heart murmurs, one of the hallmarks of non-critical heart disease typically diagnosed later in the life may be absent or misleading in CCHD because of the underlying anatomy, prolonged decline of pulmonary vascular resistance or reduced ventricular function. Despite of the increasing number of prenatal and postnatal diagnostic facility, a significant proportion of the affected newborns are still not diagnosed as the stay at home after home delivery in Bangladesh or not detected before discharge after birth in case of health facility delivery [12]. The first manifestation of CCHD may be acute due circulatory collapse and need cardio-pulmonary resuscitation or death occurs if delay in diagnosis. So this condition is associated with

significant morbidity and mortality [13, 14]. The current incidence of this severe physiologic compromise resulting from previously unrecognized CCHD has been estimated to be 1 per 15,000 to 1 per 26,000 live birth. Hence, there is broad consensus that screening for CCHD is warranted [15]. Cyanotic heart disease refers to a group of many different heart defects that are present at birth (congenital) that results in a low blood oxygen level. There are many physical defects in the heart that can cause cyanotic congenital heart disease. Many cases of CHD attend Pediatric OPD as well as in patient department of Dhaka Shishu Hospital but data regarding relative frequency, pattern, age and sex distribution of CHD are very scanty from this region. Although cyanotic congenital heart disease accounts for less than 25% of cardiac defects, many of these are life threatening in neonatal period, thereby necessitating early diagnosis and prompt treatment. Availability of diagnostic tools like echocardiography, fetal scanning, advancement in surgical techniques and postoperative care can radically improve the life expectancy of affected children. Thus, by early intervention for critical cyanotic congenital heart diseases in neonatal period or early infancy, children are now surviving beyond the pediatric age. This study will help us to determine the relative frequency, predisposing risk factors and gravity of critical cyanotic cardiac lesions presenting in neonatal period. The findings will hopefully have implications in development of cardiac service in other tertiary care centers.

## OBJECTIVES

### General Objective

- To determine the clinical profile and pattern of presentation of cyanotic congenital heart disease in neonates.

### Specific Objectives

- To determine the clinical profile of cyanotic congenital heart disease in neonates.
- To determine the pattern of cyanotic congenital heart disease in neonates.
- To determine the short term outcome of cyanotic congenital heart disease in neonates.

## METHODOLOGY AND MATERIALS

This study was designed as a cross sectional study which was carried out from 1<sup>st</sup> July 2019 to 31<sup>st</sup> December 2019 in Dhaka Shishu (Children) hospital. Dhaka Shishu (Children) Hospital (Total bed 650) has both paediatric medicine and surgery unit. A large number of patients is admitted over there each day due to various complaints and modern laboratory facilities also available for their proper diagnosis and treatment by skilled doctors and other health professional. Patients were seen in outpatient department or admitted in inpatient department of Dhaka Shishu Hospital from July to December 2019 where neonates presenting with various systemic complaints or significant cyanosis.

**Inclusion Criteria**

- Children admitted between age 0-28 days with cyanotic congenital heart disease in the outdoor & indoor patients department of Pediatric and Cardiology, DSH.
- Both sexes

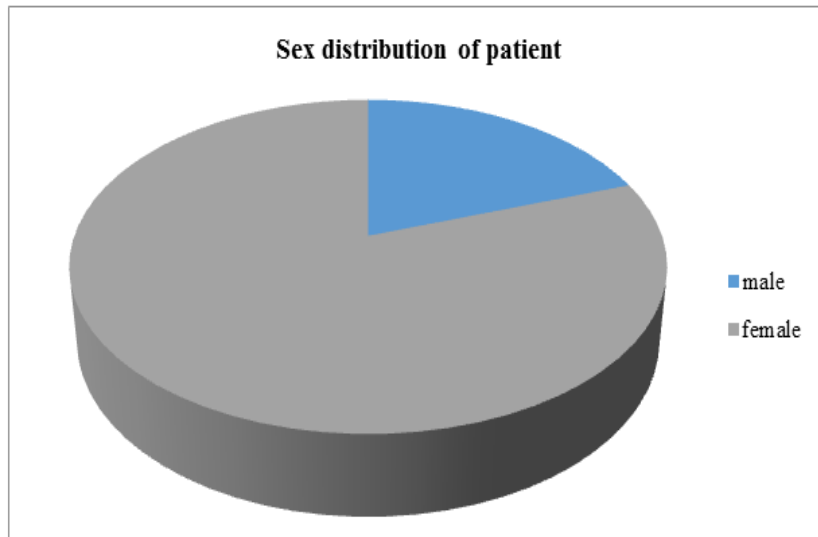
**Exclusion Criteria**

- Patients/legal guardians who would not give consent
- Old/cases already evaluated by echocardiography
- Age >28 days
- Unstable patients who died before the confirmation of diagnosis.

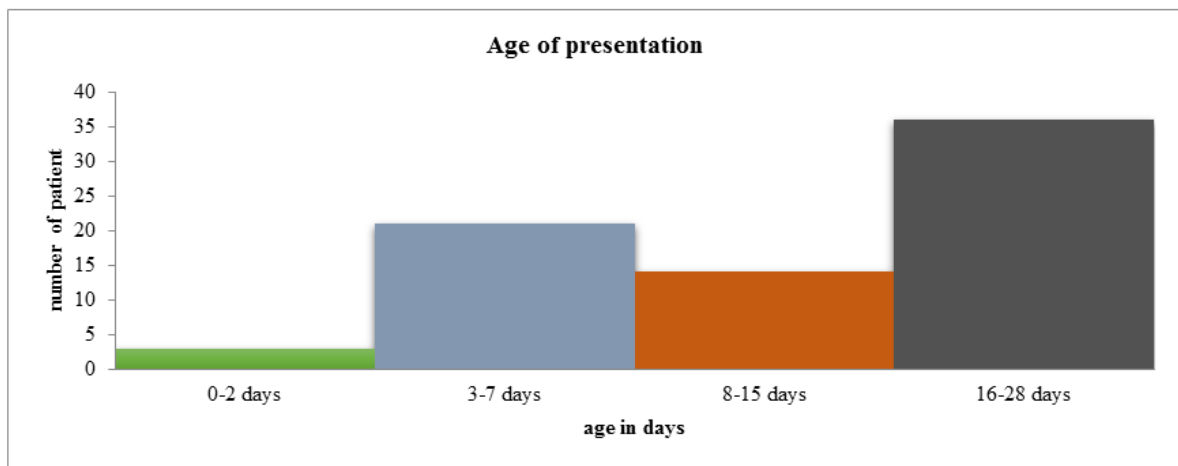
Among the 0-28 days' neonates with clinical features of chest infection, respiratory distress, cyanosis or cyanotic spells, poor feeding or difficulty in feeding & other relevant history and clinical findings of critical cyanotic congenital heart disease in Dhaka shishu (children) hospital. The study included neonates of both male and female. For each baby detailed history of age, sex, parental age, family history, onset of cyanosis and preceding/existing symptoms were recorded. A thorough general examination and other systemic examinations were done. A chest X ray, ECG and echocardiography were routinely done in all patients and if needed other investigation were performed depending upon the clinical presentation. Chest X-ray was done by professional Radiologists and echocardiography by Pediatric Cardiologists of the study places. Clinical profile and interpretations of other investigation findings were then correlated with echo cardio graphic findings. Prior to the commencement of this study ethical permission was taken from ethical review committee of Bangladesh Institute of Child Health, Dhaka Shishu (Children) Hospital. The aim and objectives of the study along with its procedure was explained to the child's parents in easily understandable local language and informed consent was taken from each guardian. It was assured that all the information and records would be kept confidential and the procedure would be helpful for both patients and physicians in making rational approach of the case management. All the data were collected and recorded systematically in a questionnaire and were analyzed manually.

**RESULTS**

In this study out of 74 neonates with cyanotic congenital heart disease 78% (n=58) were male and 22% (n=16) were female. Male: Female ratio was 3.6:1 (Figure-1). In this study majority of patient that is 36 neonates presented in between age of 16-28 days of age (48%). 21 patient presented at age range of 3-7 days (28%), 14 patient presented in between 8-15 days of age (18%) and only 3 neonates presented at 0-2 days (4%) (Figure-2). This table shows out of 74 neonates 22 (29%) were diagnosed of having Transposition of Great Arteries (TGA). Tetralogy of Fallot (TOF) were found in 12 neonates (16%), Pulmonary Atresia, DORV, Tricuspid Atresia were found in 7 patients (9.4%) respectively. Other cyanotic heart diseases observed were Total Anomalous Pulmonary Venous Return, Truncus Arteriosus, Hypoplastic Left Heart Syndrome etc (Table-1). In this study all of the study neonates (n=74) had cyanosis. Out of 74 patients 69(93%) had tachypnea, 61(82%) had tachycardia, 38(51%) had murmur, 26(35%) had hepatomegaly. Crepitation, rhonchi and oedema were found respectively 34(45%), 24(32%) and 19(25%) patients. 2 patients (2.7%) presented with shock (Figure-3). This study reveals median age of presentation for Transposition of great arteries, Tricuspid atresia and Double outlet right ventricle were 15 days. Median age of presentation for TAPVC was highest that is 23 days and Ebstein anomaly was lowest that is 8 days (Table-2). Chest X-ray findings were oligoemic lung in 39%, plethoric lung in 44%, cardiomegaly in 46%, egg on side shaped heart in 14% patients and boot shaped heart was found in 5% patients (Table-3). In this study it has been seen that majority of the patient that is 54 (72%) were term neonate. Family history of CHD were found in 9 (12%) of patient and consanguinity was found in 31(41%) of patient. Other risk factors found were maternal diabetes mellitus in 19 (26%) patients, maternal hypertension in 28 (37%) patients, infection during pregnancy in 15 (20%) patient and parental smoking in 39 (53%) patients (Table-4). In this study 9 (12%) patient received prostaglandin, 21 (28%) required use of Inotropes, antifailure drugs were used in 36 (48%) patients and emergency procedures were performed in 13 (17%) patients. Regarding outcome, 11 (14%) patient required ventilatory support, 19 (25%) had an ICU stay and death occurred in 6 (8%) patients (Table-5).



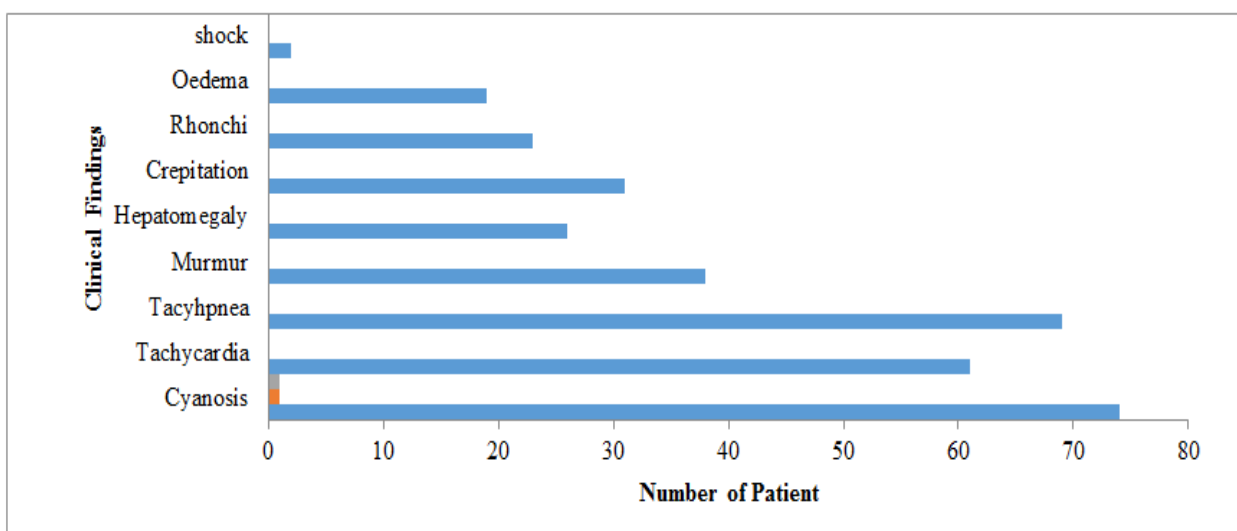
**Fig-1: Sex distribution of neonates with cyanotic heart disease**



**Fig-2: Frequency distribution of patient according to age of presentation**

**Table-1: Types of cyanotic congenital heart disease among study neonates (N=74)**

Type of cyanotic heart disease	Number of total patient	Percentage	Male	Female
Transposition of great arteries (TGA)	22	29%	20	2
Tetralogy of fallot (TOF)	12	16%	6	6
Tricuspid atresia	7	9.4%	5	2
Double outlet right ventricle (DORV)	7	9.4%	4	3
DORV with Pulmonary atresia	2	2.7%	2	0
Pulmonary atresia	7	9.4%	5	2
Total anomalous pulmonary venus return (TAPVC)	5	6.7%	3	5
Hypoplastic left heart syndrome (HLHS)	3	4%	2	1
Truncua arteriosus	2	2.7%	2	0
Single ventricle	2	2.7%	2	0
Ebstein anomaly	2	2.7%	0	2
Mitral atresia with double inlet left ventricle	2	2.7%	2	0
Critical pulmonary stenosis	1	1.3%	0	1



**Fig-3: Clinical findings of the neonates with cyanotic congenital heart disease**

**Table-2: Median age of presentation of cyanotic congenital heart diseases**

Types of cyanotic heart disease	Median age of presentation
Transposition of great arteries (TGA)	15 days
Tetralogy of fallot (TOF)	14 days
Tricuspid atresia	15 days
Double outlet right ventricle (DORV)	15 days
DORV with Pulmonary atresia	18 days
Pulmonary atresia	19 days
Total anomalous pulmonary venus return (TAPVC)	23 days
Hypoplastic left heart syndrome (HLHS)	10 days
Trunca arteriosus	19 days
Single ventricle	21 days
Ebstein anomaly	8 days

**Table-3: Chest X-ray findings among the study patients (n=74)**

X-ray finding	Number of patient	Percentage
Oligaemic lung	29	39%
Plethoric lung	33	44%
Cardiomegaly	34	46%
Egg on side appearance of heart	11	14%
Boot shaped heart	4	5%
Figure of 8 shaped heart	1	1.3%

**Table-4: Baseline demographic characteristics of study patient (n=74 )**

Variable	Number (%)
Gestational age: Term	54 (72%)
Preterm	20 (27%)
Family history of CHD	9 (12%)
Consanguinity	31(41%)
Maternal diabetes mellitus	19 (26%)
Maternal hypertension	28 (37%)
Infection during pregnancy	15 (20%)
Exposer to teratogenic drugs	2 (2.7%)
Parental smoking	39 (53%)

**Table-5: Treatment required and outcome of study patient**

Variable	Number (%)
Received prostaglandin	9 (12%)
Required use of Inotropes	21 (28%)
Use of anti-failure drugs (frucemide, digoxin)	36 (48%)
Ventilatory support	11 (14%)
Emergency procedures	13 (17%)
Required ICU stay	19 (25%)
Death	6 (8%)

## DISCUSSION

This study was done to see the clinical profile, pattern and frequency of presentation of cyanotic congenital heart disease in neonates. Total 74 neonates were diagnosed as cases having cyanotic congenital heart disease during the study period of July to December 2019 on the basis of inclusion criteria. In this study out of 74 neonates with cyanotic congenital heart disease 78% (n=58) were male and 22% (n=16) were female. Male: Female ratio was 3.6: 1. In a study by Mahapatra A *et al.*, where among 231 patients with CHD, CHD were more common among males (54.5%) with the male to female ratio is 1.2:1 [33]. In this study majority of patient that is 36 neonates presented in between age of 16-28 days of age (48%). 21 patient presented at age range of 3-7 days (28%), 14 patient presented in between 8-15 days of age (18%) and only 3 neonates presented at 0-2 days (4%). Out of 74 neonates 22 (29%) were diagnosed of having Transposition of Great Arteries (TGA). Tetralogy of Fallot (TOF) were found in 12 neonates (16%), Pulmonary Atresia, DORV, Tricuspid Atresia were found in 7 patients (9.4%) respectively. Other cyanotic heart diseases observed were Total Anomalous Pulmonary Venous Return, Truncus Arteriosus, Hypoplastic Left Heart Syndrome etc. A study conducted by Tsao P-C et al. where pulse oxymetric screening could detect 5 cyanotic congenital defects in neonates and among them 2 cases were TGA, 1 of HLHS, 1 case of Ebstein anomaly and 1 case of DORV with single ventricle [34]. In another study by Nadia M *et al.*, out of 150 study patient cyanotic lesions tetralogy of Fallot (TOF) was the commonest lesion found in 11.2% of the patients, this number was higher as compared to international data [35, 36]. However recent study of Agha Khan University, reported an even higher percentage of 25% [37]. In this study, the second common cyanotic lesion was TGA seen in 5.2%, this is in accordance with study at Atlanta<sup>38</sup> but contrast with study of Taiwan in which TGA was twice common than TOF.<sup>35</sup> Dextrocardia was seen in 1.5% (2 cases) presented in neonatal period [39]. This study reveals median age of presentation for Transposition of great arteries, Tricuspid atresia and Double outlet right ventricle were 15 days. Median age of presentation for TAPVC was highest that is 23 days and Ebstein anomaly was lowest that is 8 days. A study by Colaco S *et al.*, where among the 71 newborn with critical congenital heart disease, median age at diagnosis was

10 days and median age of presentation was 13.5 days [40]. In this study all of the study neonates (n=74) had cyanosis. Out of 74 patients 69(93%) had tachypnea, 61(82%) had tachycardia, 38(51%) had murmur, 26(35%) had hepatomegaly. Crepitation, rhonchi and oedema were found respectively 34(45%), 24(32%) and 19(25%) patients. 2 patients (2.7%) presented with shock. Mahapatra A *et al.*, showed out of 231 of his study patients 196 had murmurs, 96 had tachycardia, 84 had tachypnea, 28 had clubbing, 21 had oedema, 41 had enlarged liver [33]. Chest X-ray findings were oligoemic lung in 39%, plethoric lung in 44%, cardiomegaly in 46%, egg on side shaped heart in 14% patients and boot shaped heart was found in 5% patients. In this study it has been seen that majority of the patient that is 54 (72%) were term neonate. Family history of CHD were found in 9 (12%) of patient and consanguinity was found in 31(41%) of patient. Other risk factors found were maternal diabetes mellitus in 19 (26%) patients, maternal hypertension in 28 (37%) patients, infection during pregnancy in 15 (20%) patient and parental smoking in 39 (53%) patients. Vaidyanathan B *et al.*, demonstrated in his study where among 5487 newborns 4950 were term, 107 patient had consanguinity, 542 neonates mother had diabetes, maternal hypertension was present in 571 patient and maternal infection during pregnancy was found in 264 patients [41]. In this study 9 (12%) patient received prostaglandin, 21 (28%) required use of Inotropes, antifailure drugs were used in 36 (48%) patients and emergency procedures were performed in 13 (17%) patients. Regarding outcome, 11 (14%) patient required ventilatory support, 19 (25%) had an ICU stay and death occurred in 6 (8%) patients. A study conducted by Colaco S *et al.*, where comparison of neonates with cyanotic HD diagnosed antenatally and postnatally was done. Among the postnatally group out of 71 patient 19 were transferred on prostin, 4 patient required inotropes, 9 patient had convulsion and 4 patient died [40].

## LIMITATIONS OF THE STUDY

A limitation of this study is the relatively small sample size and study period was short. For evaluation of consequence of cyanotic congenital heart disease in neonates all facilities were not available in the place of study. Echocardiography was expensive and many poor patients couldn't afford to do that. Lowering the cost of investigation would be beneficial for the patients.

## CONCLUSION AND RECOMMENDATIONS

Cyanotic congenital heart disease in neonates is often undiagnosed which is main the cause of high case fatality. Development of neonatal screening program for early detection of such cases and prompt measure can surely save life of many newborns. Therefore, gathering knowledge about the clinical profile and pattern of presentation of cyanotic congenital heart disease in neonates in our country's perspective will definitely help to avoid complication, reduce mortality and improve quality of life of the newborns. Pediatric echocardiography as diagnostic tool should be made more widely available especially in tertiary institutions to enable early diagnosis and, screening for possible cardiac defects during pregnancy. There is an urgent need for the government to establish pediatric cardiac surgical centers with specialized medical cardiology, intensive care, imaging and interventions. All newborn babies should be examined thoroughly for any evidence of CHD by pediatrician before hospital discharge and on follow up visits in the early neonatal period. There is a need to intensify efforts to educate general practitioner and other health professionals who come across children with heart disease, as they are the ones who can screen, diagnose and refer these babies to cardiac facility and the earlier the diagnosis, the easier it would be to do timely management in appropriate manner.

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