

Incidence and Clinical Characteristics of Congenital Heart Disease among Neonates in Neonatal Intensive Care Unit

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Abstract: Background: Congenital heart diseases affect a large number of newborns and account for a high proportion of infant mortality worldwide. There are regional differences in the prevalence and distribution pattern of congenital heart diseases. We aimed to evaluate the relative frequencies, risk factors and diagnostic clues of congenital heart disease in newborns at our neonatal intensive care unit. Materials and methods: Among 326 cases admitted to the Abant Izzet Baysal University Neonatal Intensive Care Unit between February 2013 - September 2014, thirty-five newborns with congenital heart disease were evaluated, retrospectively. Data was collected on a predesigned proforma containing information regarding gender, gestational age and weight at birth, family history, and associated malformations. Results: The prevalence of congenital heart disease was 10.7%. The most common cause for cardiology consultation was cardiac murmur (85.5%). The relative percentage of acyanotic and cyanotic heart defects were 91.6% and 8.4%, respectively. The most frequent acyanotic defect was ventricular septal defect with a prevalence of 22.8%. Maternal diabetes mellitus and Down syndrome were observed in 17.1% and 2.8% of cases, respectively. Conclusion: The prevalence of congenital heart disease in newborns at the neonatal intensive care unit was higher than that in all live births and most common defect was muscular VSD. The most frequent diagnostic clue was cardiac murmur.

Keywords: Congenital Heart Diseases, Echocardiography, Neonatal Intensive Care Unit, Newborn

1. Introduction

Congenital heart diseases (CHDs) are the most common congenital fetal malformations and are responsible for a high rate of child mortality and morbidity. While prenatal incidence of CHD ranges from 2.4 to 52% (1-6), postnatal incidence is around 0.3 to 1.2% of live births (7, 8). CHD is more frequent in neonates in the neonatal intensive care unit than in live born neonates in population. Shima Y et al suggested that the occurrence rate of CHD was approximately 4.5% of neonates admitted to the neonatal intensive care unit (9). In current study we found the prevalence of CHD was 10.7% among neonates in neonatal intensive care unit that was higher according to literature. Higher frequency of emergence of CHD is an expected result, because of being sick babies, inspection of every day and

more frequent evaluation of echocardiography in this group. In a study that performed on 3334 neonates in neonatal intensive care unit, the frequency was found to be 4%. Heart murmur, central cyanosis and major congenital abnormalities were found statistically higher in the neonates with cardiac disorder (10). Archer et al. (11) reported that CHD is probably more frequent in very low birth infants treated in neonatal intensive care units than in the general live-born population. According to Aydogdu et al (12) and Shima et al (9) the most common cyanotic congenital heart disease was tetralogy of Fallot and the most common acyanotic heart disease was VSD in the neonatal intensive care unit. In our study results were same with those published reports.

Neonatal period because of the hemodynamic differences,

there are difficulties in establishing the diagnosis of CHD. On the other hand, late diagnosis can be fatal in some cases (13). CHD may be associated with chromosomal abnormalities that are related to familial transition or part of syndromes (14). The clinical finding of CHD varies according to anatomic abnormalities in neonatal period. Besides severe symptoms such as respiratory distress, cyanosis and shock, murmur may be a single symptom.

In this study, we aimed to investigate the distribution, prevalence and pattern of various CHD in neonates followed in the neonatal intensive care unit at Abant İzzet Baysal University.

2. Materials and Methods

Among 326 cases admitted to the Abant İzzet Baysal University Neonatal Intensive Care Unit between February 2013 - September 2014, thirty-five newborns with CHDs were evaluated, retrospectively. The initial examination and evaluation of newborns were performed by newborns experts and patients were evaluated by a specialist pediatric cardiology if necessary. Echocardiography was performed to patients with abnormal heart murmur, abnormal electrocardiography findings, respiratory distress or cyanosis with unexplained pulmonary disease, Down's syndrome, dysmorphic features and diabetic mothers. The patients included in the study that was evaluated by echocardiography among sick babies in neonatal intensive care unit.

Interatrial septum opening less than 3 mm and the minimal patent ductus arteriosus (PDA) cases determined in the first three days (the cases of constant pressure difference between ductus arteriosus and pulmonary artery in doppler USG; and continuous minimal flow from left to right and no enlargement in the left ventricular and left atrial in colored doppler USG) were excluded in the study. Sociodemographic characteristics, perinatal characteristics, maternal history, physical examination, echocardiography results were recorded. Maternal age, miscarriage and stillbirth, brother history of CHD, diabetes, systemic lupus erythematosus (SLE), exposure to radiation, TORCH infection in pregnancy, smoking and alcohol use, intermarriage, CHD in the family were questioned.

Data was collected on a predesigned proforma containing information regarding gender, gestational age and weight at birth, family history, and associated malformations. SPSS was used for statistical analysis.

3. Results

The prevalence of CHD among infants admitted to our unit was 10.7%. Non-cyanotic CHD was observed in 32 patients and cyanotic CHD was in 3 patients. The most frequently isolated non-cyanotic heart diseases were, ventricular septal defect (VSD) atrial septal defect (ASD) and patent ductus arteriosus (PDA) respectively, while cyanotic CHDs as, transposition of the great arteries coarctation of the aorta and hypoplastic left heart syndrome (Table 1). One case was

Down syndrome. Maternal diabetes frequency was 17.1%.

The most common cause for cardiology consultation was murmur (85.5%). Cyanosis, respiratory distress, dysmorphic features and maternal diabetes were the other main reasons for the consultation. 62% of cases were born in our hospital. 40% of cases were female and 60% of were male. 51% of cases were premature (28-37 weeks), 49% of were mature (38-41 weeks). The birth weight was ranged between 1035-4290gr. The average maternal age was 28±5 year. Three parents (8.5%) had consanguineous marriages and four parents (11.4%) had miscarriage history. Two patients (5.7%) had family history of CHD.

There was no history of SLE, alcohol or tobacco use during pregnancy, exposure to radiation in any mothers. One patient (% 2.8) was diagnosed as Down syndrome and atrioventricular septal defect was detected in echocardiographic examination. Six patients with a diagnosis of CHD (17.1%) had diabetes in their mother. Heart diseases in infants of diabetic mothers are presented in Table 2 and the most common finding was muscular VSD.

Table 1. The frequency of cyanotic and noncyanotic heart diseases.

Cardiac disease	isolated	with other cardiac diseases	total
	n (%)	n (%)	n(%)
VSD	8(22.8)	12(34.2)	17(57)
ASD	7 (20)	11(31.4)	18(51.4)
PDA	6(17.1)	3 (8.5)	9(25.6)
Transposition of the great arteries	1 (2.8)	0	1(2.8)
Hypoplastic left heart syndrome	1 (2.8)	0	1(2.8)
Coarctation of the aorta	1 (2.8)	0	1(2.8)

VSD: Ventricular septal defect, ASD: Atrial septal defect, PDA: Patent ductus arteriosus

Table 2. The frequency of congenital heart disease in infants of diabetic mothers.

Cardiac disease	n
Muscular VSD	3
Secundum ASD	2
ASD+VSD	1
Total	6

VSD: Ventricular septal defect, ASD: Atrial septal defect

4. Discussion

CHDs are important major anomalies observed more common in neonatal period and may require immediate treatment. In current study we found that the frequency of CHD in hospitalize newborns was 10.7%. Aydogdu et al was found 6.6% (12) and Guven et al 4.9% (15). The prevalence of CHD was higher in this study than other reports; this may be due to high prematurity rate as 51%. Aydogdu et al reported the prematurity rate as 18% (12).

The incidence of CHDs is summarized in table 3.

Table 3. The incidence of congenital heart disease (16).

Lesion type	%
VSD	25-30
ASD (secundum)	6-8
PDA	6-8
Coarctation of the aorta	5-7
Tetralogy of Fallot	5-7
Pulmonary valve stenosis	5-7
Aorta valve stenosis	4-7
Transposition of the great arteries	3-5
Hypoplastic left heart syndrome	1-3
Truncus arteriosus	1-2
Tricuspid atresia	1-2
Single ventricle	1-2
Double outlet right ventricle	1-2
Others	5-10

VSD: Ventricular septal defect, ASD: Atrial septal defect, PDA: Patent ductus arteriosus

This retrospective study showed that the most common reason for cardiology consultation was heart murmur. In the study of Aydogdu et al suggested that the most common reason for cardiology consultation was again murmur; alone or in combination with other signs with a ratio of 87.5% (12). In addition to pathological murmurs, the innocent systolic murmurs can be heard more than 50% of healthy and term infants in the first week (17).

VSD is most common defect among CHD with 25% percent and seen 2.5/1000 in live births. In our study, the most frequent CHD was VSD and among these patients the most common seen VSD type was small-medium with muscular VSD. VSD combined with other cardiac abnormalities were detected in 34.2% of infants. In the study of Aydogdu et al single VSD was found as the most frequent CHD (19.6%) (12). Hussain et al found that the frequency of CHD among 5800 neonates in a neonatal unit of a tertiary care hospital was % 1.5 with male preponderance and VSD was found to be most common defect (%31.3), (18). VSD is usually asymptomatic and often closes spontaneously (19). The reported rates of spontaneous closure vary between 50% and 75% in small defects (20-22). Therefore, the prevalence of VSD should be higher in neonates. Some studies done by performing echocardiography on newborns with no murmurs, have found huge numbers of tiny muscular VSD with the incidence of 2-5% (23, 24). Roguin et al (23) reported that the prevalence of muscular VSD in neonates of 53.2/1,000 live births and patients were asymptomatic, and 88.9% had defects that closed spontaneously within 1 to 10 months.

ASD represents 8-9% of CHD with the prevalence of 0.5/1000 in live births. Many infants have a patent foramen ovale with a tiny left-to-right shunt, and inclusion of these will inflate the incidence of ASD. ASD is usually asymptomatic therefore murmurs are often soft; these defects frequently do not lead to early diagnosis or referral. Because diagnosed in adulthood, the incidence in childhood usually

underestimates the true incidence of the lesion (25, 26). We found the incidence of ASD among neonates 51.4%. 20% of cases were isolated and 31.4% was combined with other cardiac defects.

PDA is another common defect which has an increased incidence among preterm infants with the prevalence of 10% among CHD. In term infant, the ductus arteriosus is almost always closed by four to seven days after birth. In current study 25.6% of neonates had PDA which was the third common defect that observed in neonatal intensive care unit. The reason for the high incidence may be due to high rate of prematurity in this study.

Atrioventricular septal defects (AVSDs) (endocardial cushion defects) have an incidence that varies with the age of the involved mothers. It's incidence is 0.20-0.25/1000 of neonates in live births and creates 4-5% of CHD. Trisomy 21 (Down syndrome) is much more common in mothers more than 34 years old, and AVSDs are much more frequent in those with trisomy 21 than with normal chromosomes. Thus, the incidence of AVSDs increases in older mothers children. It is known that, 40% of children who was diagnosed as Down syndrome have CHD and most common defect is endocardial cushion defect (27). Reinhold et al (28) found that the genetic syndrome prevalence was 5.6% among 814 patients with CHD and they reported the frequency of Down's syndrome was 1.4%. In our study, this ratio was found to be 2.8%. Aydogdu et al found the prevalence of Down syndrome as 3.6% (12).

Coarctation of the aorta is an abnormality with the combination of upper body hypertension and weak or absent femoral pulses. But, studies in children's hospitals have shown that the diagnosis is often missed by the referring pediatric cardiologist (29). We found 2.8% of newborns with coarctation of the aorta among hospitalized neonates in the neonatal intensive care unit.

Transposition of the great arteries (TGA) is the most common form of CHD, presenting within the first week of life. The aorta and the pulmonary artery take their origins from the right ventricle and left ventricle respectively. A dual parallel circulation exists where fully saturated blood from the pulmonary veins is returned to the pulmonary circulation, and desaturated blood from the systemic veins is returned to the systemic arterial circulation. This results life-threatening hypoxia in the newborn period. TGA affects 5% of infants with CHD with 90% of mortality within the first year of life (30). We observed 2.8% TGA among hospitalized neonates.

Hypoplastic left heart syndrome is a rare congenital heart defect in which the left side of the heart is underdeveloped. It was previously regarded as fatal but surgical management of hypoplastic left heart syndrome has changed the prognosis (31). The incidence of this syndrome is 1-4% among CHD. We found 2.8% of our infants were with this syndrome.

In our study, 17.1% of infants with CHD had diabetic mothers and most common defect was muscular VSD among this infants. In general, asymmetric septal hypertrophy, VSD, ASD, pulmonary stenosis are common abnormality in infants of diabetic mothers (32).

In conclusion, the incidence of CHD in newborns at the neonatal intensive care unit was higher than that in all live births. Its incidence varies from centre to centre due to different factors like nature of the sample, method of detection and early examination by a neonatologist / pediatrician. In this study a higher incidence is reported because it was carried out in a tertiary care unit and prematurity prevalence was higher. The most frequent diagnostic clue was cardiac murmur and the most frequently observed risk factors were maternal diabetes mellitus and genetic syndrome.

Study Limitations

We have carried out a small regional study; there is a need to establish a database for CHDs among sick babies in neonatal intensive care unit. Therefore further studies with the large number of neonates are recommended.

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