

## Prognosis of the Newborns with Congenital Heart Diseases

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### Abstract

**Background:** Congenital heart disease (CHD) is defined as problems affecting the heart of the fetus. According to previous studies, the incidence rate varies from 4-12 in 1000 live births. This study aimed to evaluate the demographic characteristics, clinical presentations, and findings that have impact on the prognosis of newborns hospitalized in Tabriz children Hospital, northwest of Iran. **Methods:** This cross-sectional study was conducted on 82 neonates with CHD admitted in Tabriz children Hospital, northwest of Iran from December 2011 to December 2012. Demographic characteristics, main complaints, symptoms, clinical presentations, associated anomalies, diagnoses, and also the correlation between the treatment process and mortality were evaluated. All data were analyzed using SPSS™ version 16 statistical software. **Results:** A total of 82 neonates with CHD were enrolled. The mean age and birth weight were 8.54 days and 2828.17 grams, respectively. The most common chief complaint was respiratory distress (58 cases, 70.73%). The most common diagnosis was d-transposition of great arteries and the most frequent therapeutic procedure was medical therapy in combination with mechanical ventilation (31 cases, 37.8%). 10 neonates (12.1%) had complications while seizure was the most common complication of the study (4 case, 4.87%). Of 82 newborns in this study, 30 patients (36.6%) died of which 13 cases (43.33%) had DTGA; the most common therapeutic procedure was mechanical ventilation plus medical therapy which was performed in 22 patients (73.33%). In our study, no significant correlation could be observed between age, sex or weight of neonates and final outcome. **Conclusion:** Mortality of neonates with critical CHD is high. Also, the neonates treated with more invasive methods have higher mortality rates. It is obvious that both early detection and timely management affect ultimate prognosis of these patients. Hence, prenatal (fetal echocardiography) and postnatal (pulse oximetry and echocardiography) diagnostic evaluations and instituting appropriate referral system are crucial. Furthermore, establishment of neonatal and pediatric intensive care and pediatric cardiac, cardiac surgery and catheterization laboratory units in the same tertiary center is suggested.

**Keywords:** Prognosis; Congenital heart disease; Newborn

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## Introduction

Congenital heart disease (CHD) refers to structural and functional problems of the heart which occur due to abnormal heart development. Being associated with substantial noninfectious perinatal morbidity and mortality, CHD is one of the most common congenital malformations among newborns. With an estimated incidence of 4-12/1000 (0.4-1.2%) of live births, CHD accounts for 4% of all neonatal deaths; 50% of all deaths in patients with CHD occur during the first year of life.<sup>1-10</sup>

Despite remarkable progresses in medical sciences, the major cause of CHD is not well-known yet. However, previous studies demonstrated that both genetic and environmental factors have significant impacts on CHD pathogenesis.<sup>8-11</sup> CHD could lead to a wide range of defects.<sup>12</sup> Administration of some medicines such as anticonvulsants, lithium or progesterone during pregnancy may be blamed for its occurrence. Ventricular Septal Defect (VSD) is the most common congenital heart anomaly accounting for approximately 30-40% of CHDs.<sup>13</sup> Substantial advances have been made in diagnosis (even before birth), treatment (medical, transcatheter, and surgical), and care methods among these patients in developed countries. These factors have led to significantly improved longevity, lifestyle and quality of life among these patients.<sup>14</sup>

Although the prevention of CHD is beyond the capability of current medical sciences (except for the ability of controlling the effective factors such as consanguinity, drugs consumption and other teratogenic supplements during the pregnancy), progress in treatment and care methods has been associated with hopeful results.

A wide range of reported mortality rates could be found in the literature. Although there are few studies on the management of CHD in

children with low birth weight, there is not any report on the incidence of CHD in children with low birth weight; however, the incidence of these diseases are much higher in preterm infants. Thanks to more appropriate obstetric care and more available prenatal diagnosis, prognosis of preterm infants has been improved compared to last decades. Thus, it is likely that these factors may affect the incidence of mortality in children with CHD.<sup>15</sup>

Prematurity and birth weight of less than 2500 gr are the risk factors for increasing hospital mortality. Optimal timing and type of intervention are controversial in premature infants with complex congenital heart disease. The mortality rate is almost 30% in premature children with very low birth weight.<sup>16</sup> On the other hand, evaluating factors with the ability of affecting the preoperative status and side effects of surgery would be useful in improving the long-term prognosis. Considering the large number of hospitalized patients in pediatric hospitals and the significant morbidity and mortality in neonates with CHD, we decided to investigate causes and risk factors for mortality in these neonates and evaluate the efficacy of the therapeutic procedures in the neonatal intensive care unit (NICU) on prognosis of newborns with CHD.

## Material & methods

In this cross-sectional retrospective study, lasting for 12 month, medical records of 82 newborns with CHD admitted in NICU of Tabriz children hospital, northwest of Iran were reviewed. All required information was obtained from hospital records and all patients' information remained confidential. This study was approved by local medical ethical committee of Tabriz University of medical sciences.

Medical records of patients were reviewed and all data related to prenatal and postnatal



findings, also any complications having required further intervention during the treatment courses and those associated with the outcome and prognosis of patients were collected using the designed checklists. Also, clinical symptoms, diagnosis, complications and patients' health status at discharge from the hospital were evaluated. Studied variables were: age, sex, weight, chief complaint, associated symptoms, associated disease, complications, final diagnosis, treatment methods and final status of the patients. Measurements were expressed as mean  $\pm$  standard deviation (SD), frequency and percentage. Chi square test and Fishers' exact test were used to compare the study variables. The obtained data were analyzed using SPSS™ version 16 statistical software (SPSS Inc., Chicago, IL, USA). In this study,  $P < 0.05$  was considered statistically significant.

## Results

Of 82 cases, 51 patients (62%) were male. The mean age and weight of the patients were 8.54 days (range=1-30 days) and 2828.17 gram (range=1350.00-4500.00 gr), respectively.

In this study, the mean age at diagnosis in hospitalized patients was 8.54 days and the mean birth weight of patients was 2828.17 gr. Respiratory distress was the most common complaint in patients (58 patients, 70.7%) and cyanosis was the second most common complaint (20 patients, 24.4%). Mechanical ventilation combined by medical therapy was the most common treatment method used (31 patients, 37.8%). Also, 2 patients (2.43%) were given interventional treatment, 29 cases (35.4%) medical therapy only and 6 patients (7.3%) were treated surgically. 13 patients (15.8%) had coexisting diseases and 69 cases (84.1%) had no coexisting diseases. In addition, multiple congenital anomalies were the most common associated anomaly in (3 patients, 3.7%). Down syndrome was observed in 2 patients (2.4%). 12.1% of the patients (10

cases) experienced complications; seizure was observed in 4 patients (4.87%) as the most common complication. 2 patients (2.4%) experienced pneumothorax and the other 2 (2.4%) cases experienced Pneumonia. Moreover, 1 patient (1.2%) had gastrointestinal bleeding and 1 patient (1.2%) had pleural effusion. Of 82 newborns in this study, 30 patients (36.6%) died of which 13 cases (43.33%) had DTGA.

Respiratory distress was the most common complaint among the patients (Table-1). Medical therapy and mechanical ventilation plus medical therapy were the most commonly administered therapeutic methods (Table-1).

There was no coexisting disease in 69 (84.1%) of patients; esophageal atresia (EA) was the most common associated disease in (8 patients, 9.75%) (Table-1). Regarding the associated complications with CHD, our study revealed that 72 (87.8%) patients had no associated complications; meanwhile, seizure was the most common associated complication in 4 (4.87%) patients (Table-1).

Among the treatment methods, the only significant association was observed between the medical therapy method and the final outcome of the discharged patients ( $P < 0.001$ ,  $R = 0.317$ ). Also, the combination of medical therapy and mechanical ventilation was of significant correlation with the mortality ( $P < 0.001$ ,  $R = 0.442$ , Table 3).

Regarding the final status of the patients, 13 (15.85%) cases were clinically followed up, 39 (47.56%) cases were medically followed up and 30 (36.58%) cases died.

## Discussion

Over the past 10-15 years, the medical world has been witnessing dramatic developments in diagnosis and treatment (or mitigation) of CHD in newborns. Advanced diagnostic methods such as echocardiography have played a major



Parameter	Patients (n=82)
Main complaints:	
Respiratory distress	58 (70.7%)
Cyanosis	20 (24.4%)
poor feeding	1 (1.2%)
oliguria	1 (1.2%)
palpitation	2 (2.4%)
Treatment method:	
Medical therapy	29 (35.4%)
Transcatheter Intervention	2 (2.4%)
surgery	6 (7.3%)
clinical follow up	6 (7.3%)
mechanical ventilation + Medical therapy	31 (37.8%)
mechanical ventilation + surgery	8 (9.8%)
Associated diseases with CHD:	
Without any diseases	69 (84.1%)
Down syndrome:D.S	2 (2.4%)
Cleft palate:C.P	1 (1.2%)
Bile duct atresia:B.D.A	1 (1.2%)
Multiorgan anomaly: MOA	3 (3.7%)
Choanal atresia:C.A	1 (1.2%)
Esophageal atresia:EA	8 (9.75%)
Intestinal atresia: IA	2 (2.4%)
IA+EA	1 (1.2%)
Associated complications with CHD:	
Without any complication	72 (87.8%)
Seizures	4 (4.87%)
gastrointestinal(GI)Bleeding	1 (1.2%)
Pneumothorax	2 (2.4%)
Pneumonia	2 (2.4%)
Pleural effusion	1 (1.2%)

**Table 1:** frequency of main complaints, treatment methods and associated diseases and complications with CHD

role in this regard. In addition, sophisticated surgical techniques have made it possible to complete treatment or mitigation in infants with CHD weighing even less than 1.5-2Kg.<sup>17</sup>

In a study by Henry et al. on 180 patients, the authors reported that 37 cases (21%) died at the hospital. Mechanical ventilation plus medical therapy were reported as one of the associated risk factors for mortality.<sup>15</sup> In the current study, of 82 patients 36.6% (30 cases) died and of 31 patients treated by mechanical ventilation plus medical therapy, 22 patients

(70.96%) died. Our results about the mortality and the requirement for mechanical ventilation are in line with the results of previous studies.

Kecskes et al. studied 47 infants with congenital heart disease; VSD and coarctation of aorta were the most common diagnoses in their study. Death due to congenital heart disease was reported in 32% of the cases with coarctation of aorta having the highest mortality rate (62%).<sup>14</sup>

Similarly, in our study the mortality rate was 36.6% (30 cases); while, DTGA was the most

Final diagnosis	Patients (n=82)
d-transposition of the great arteries:DTGA	20 (24.4%)
coarctation of the aorta: COA	2 (2.4%)
ADS+PDA	11 (13.4%)
patent ductus arteriosus: PDA	3 (3.7%)
atrial septal defect:ASD	3 (3.7%)
ventricular septal defect:VSD	1 (1.2%)
pulmonary hypertention(PH)+ Mitral Supravalvular ring	1 (1.2%)
hypoplastic left heart syndrome:HLHS	1 (1.2%)
COA+VSD	2 (2.4%)
Pulmonary Atresia +Tricuspid Atresia +PDA	1 (1.2%)
COA+PDA+ Sever aortic valve stenosis(AS)	1 (1.2%)
VSD+PDA	5(6.09%)
Sever AS	1 (1.2%)
COA+ASD	1 (1.2%)
DTGA+ single ventricle	1 (1.2%)
PDA+ Pulmonary Atresia	4 (4.9%)
congenital complete heart block (CCHB)+ Dilated Cardiomyopathy	1 (1.2%)
Primary pulmonary hypertention(PPHN)	1 (1.2%)
Cardiac Tumor	1 (1.2%)
double outlet right ventricle(DORV) + VSD+PH	1 (1.2%)
Single Atrium +Single Ventricle +PH	1 (1.2%)
pulmonary stenosis(PS)+dextrocardy	1 (1.2%)
Pulmonary Atresia+ Intact ventricular septum	1 (1.2%)
VSD+ASD	3 (3.7%)
COA+ Complete atrio-ventricular Canal defect(CAVCD)	1 (1.2%)
Complete AV Canal defect	1 (1.2%)
COA+PDA	4 (4.9%)
PS+ Single ventricle	1 (1.2%)
Pulmonary Atresia +VSD	2 (2.4%)
COA+VSD+ASD	1 (1.2%)
COA+PDA+VSD	1 (1.2%)
total anomalous pulmonary connection( TAPVC)	1 (1.2%)
Idiopathic Dilated Cardiomyopathy	2 (2.4%)

**Table 2:** Final diagnosis of CHD

common diagnosis (20 cases, 24.4%) and had the highest mortality rate (43.33%).

In the studies by Brown et al. and Anagnostou et al., the mortality rates were 12% (of 286 patients) and 10% (of 105 patients), respectively which were lower than the mortality rate in our study.16, 18 Moreover, in

a study by Sawanta and colleagues, VSD (42.86%) and ASD (25.71%) were the most common CHDs, respectively. Also, VSD and TOF were more common in male patients and ASD was more common among female patients. Associated diseases were observed in 22.86% of patients with Down syndrome being the most common disease in this regard.19 In the

present study, DTGA and ASD+PDA were the most common diseases in both groups and multiple congenital anomalies (3 Patients,

3.7%) was the most common associated anomaly.

Parameter	Patients (n=82)	Dead (n=30=36.58%)
Medical therapy	29 (35.4%)	5 (17.24%)
Transcatheter Interventional therapy	2 (2.4%)	0 (0%)
Surgery	6 (7.3%)	1 (16.6%)
Mechanical ventilation + medical therapy	31 (37.8%)	22 (70.96%)
Mechanical ventilation + surgery	8 (9.8%)	2 (25%)
Clinical follow up	6(7.3%)	0(0%)

**Table 3:** Mortality rate occurred by the used therapeutic methods

Padley and colleagues studied the final outcome of infants with congenital heart disease undergoing palliative or curative surgery in Australia during 5 years. The total mortality rate at hospital discharge and after 30 days of surgery was 1.9% and 1.3%, respectively.<sup>20</sup> In our study, of 82 patients, 6 cases underwent surgery, 1 patient (16.6%) died which is higher than previous studies. Alfieris et al. studied the final status of patients who underwent the surgery with DTGA diagnosis; in their study, of 96 patients who underwent surgery, the mortality rate of 0% was reported.<sup>21</sup> Nevertheless, in our study of 20 patients with DTGA diagnosis, 6 cases were operated of which 3 cases (50%) died, indicating the higher mortality rate.

In our study, no significant correlation between age, sex or weight of neonates and final outcome was observed.

### Conclusion

Mortality rate of newborns with critical congenital heart disease is high, especially in those requiring invasive treatments such as mechanical ventilation or surgery. Mortality

rate can be higher when the management is established with delay due to either delayed referral or diagnosis. Furthermore, separation of neonatal intensive care units from cardiac wards and cardiac surgery centers has significant effect on final outcome of these newborns. Hence, the following recommendations are suggested to improve the outcome of these patients:

- 1- Establishment of prenatal diagnostic procedures such as fetal echocardiography in high risk pregnancies
- 2- Routine upper and lower pulse oximetry in addition to precise physical examination before home discharge of neonates
- 3- Establishment of appropriate referral system for timely referral of these neonates to pediatric cardiologists
- 4- Establishment of neonatal intensive care units, pediatric cardiology catheterization laboratory, and pediatric cardiac surgery with trained and expert staff for timely and appropriate management of these patients in the same centers.

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