Clinical profile of cyanotic congenital heart disease in neonatal intensive care unit at Sohag University Hospital, Upper Egypt

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Abstract  Background: Cyanotic congenital heart disease (CCHD) accounts for 25% of congenital heart defects and has a high morbidity and mortality in neonates.

Objective: The aim of this work was to study clinical profile and available therapeutic modalities used in the management of CCHD.

Subjects and methods: This was a prospective study carried out for a period of one year, starting from January 2011, and included 50 neonates admitted to neonatal intensive care unit (NICU) of Sohag University Hospital, Sohag, Upper Egypt. All included patients were subjected to thorough clinical history, full clinical examination, initial and frequent measurement of oxygen saturation by pulse oximetry, blood gas analysis, and echocardiography. Also, therapeutic modalities used in management of CCHD were noted.

Results: 50 neonates were diagnosed as having CCHD, out of them 39 (78%) were males with male to female ratio 3.55:1. The mean age of presentation was 11.78 ± 9.4 days. CCHD frequency was found to be 9.5% (50/524) in our NICU population. The most common type of CCHD was d-transposition of great arteries (D-TGA) (66%) followed by complex CCHD (12%) and hypoplastic left heart syndrome (HLHS) (12%), whereas the less common type was hypoplastic right ventricle (2%). All cases presented with central cyanosis and needed medical treatment whereas balloon atrial septostomy was performed in 26% of cases. Seventy-four percent of cases were improved and referred to higher centers while 26% were expired during hospital stay.

Conclusion: CCHD is a leading cause of neonatal morbidity and mortality. CCHD frequency was significant (9.5%) in our study population with D-TGA being the commonest type. Majority of neonates with CCHD showed survival with suitable management. Early diagnosis and referral to pediatric cardiac center for proper management will improve the outcome. Neonatologists and pediatric cardiologists should be familiar with diagnosis and management of CCHD.

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1. Introduction

Congenital heart disease (CHD) can be defined as an anatomic malformation of the heart or large vessels which occurs during intrauterine development, irrespective of the age at presentation [1]. It is the most common congenital problem in children representing nearly 25% of all congenital malformations [2]. The incidence of CHD in different studies varies from about 4/1000 to 50/1000 live birth [3]. Congenital cyanotic heart disease (CCHD) accounts for 25% of all cases of CHD [4].

Etiology of most cases of CHD is thought to be multifactorial and they result from a combination of genetic predisposition and environmental factors [5]. The majority of genetic causes of CHD are sporadic genetic changes or large chromosomal abnormalities [6]. Environmental factors for the development of CHD include maternal disease and drug exposure [7]. Positive family history is considered one of the most common risk factors for CHD [8].

The neonates with CCHD may present with cyanosis, cardiovascular collapse, and congestive heart failure or combinations of these presentations. Pure versions of specific defects may present in some patients, but many neonates have various combinations of defects [9]. Echocardiography, with Doppler and color Doppler has become the primary diagnostic tool for CCHD. In addition, it reduces the requirement for invasive studies such as cardiac catheterization [10].

There is lack of data about spectrum of CCHD in neonates in our locality. The aim of the study was to describe clinical profile and available therapeutic modalities used in the management of CCHD in neonatal intensive care unit (NICU) at Sohag University hospital, Upper Egypt.

2. Subjects and methods

2.1. Study design

This was a prospective, cross sectional, descriptive study carried out for a period of one year, starting from January 2011, at neonatal intensive care unit (NICU) in Sohag University Hospital, Sohag, Upper Egypt. Sohag University Hospital is a tertiary care referral hospital for patients from hospitals in Sohag and Qena Governorates. Admitted neonates diagnosed with cyanotic congenital heart disease (CCHD) were included in the study.

2.2. Exclusion criteria

Neonates diagnosed as acyanotic congenital heart disease, other causes of central cyanosis and persistent pulmonary hypertension of neonates (PPHN) were excluded from the study.

2.3. Ethical consideration

The protocol of the study was approved by Research Ethics Committee at Sohag Faculty of Medicine. Informed consent was obtained from parents of all enrolled subjects. The work has been carried out in accordance with The Code of Ethics of The World Medical Association (Declaration of Helsinki) for experiments on humans.

2.4. Methods

All neonates included in this work were subjected to the following: thorough clinical history, full clinical examination, initial and frequent measurement of oxygen saturation by pulse oximetry and arterial blood gas (ABG) analysis. Echocardiography has been performed by a single pediatric cardiologist at NICU using M-mode, Two-dimensional Color Doppler cardiac imaging, 7 MHz, 5 MHz-transducers frequency by the mean of Two-dimensional ECHO (Vivid 3). All therapeutic modalities used in management of CCHD were documented.

2.5. Statistical analysis

The data were subjected to statistical analysis and tabulation using SPSS version 18. P value is considered significant if less than 0.05 then the results were presented to fulfill the objectives of the study.

3. Results

During the 1-year study period, total admissions in our NICU were 524. Out of them 50 neonates were diagnosed with cyanotic congenital heart disease (CCHD) and included in the study. CCHD frequency was found to be 9.5% (50/524) in our NICU population. Out of CCHD cases, 39 (78%) were males with male to female ratio 3.55:1. Their age of presentation ranged from 2 to 28 days with mean age of presentation 11.78 ± 9.4 days. The most common presenting symptoms were central cyanosis (100%), followed by respiratory distress (82%), then feeding difficulties (62%). The less common symptoms were generalized convulsions (12%) and bleeding tendency (4%).

The commonest risk factors encountered among the studied neonates were positive parental consanguinity (34%), followed by maternal drug intake in 1st trimester of pregnancy (20%). These drugs included antibiotics, analgesics, antipyretics or folic acid. Maternal infection during 1st trimester and positive family history of CCHD, were found in 16% and 12%, respectively (Table 1).

Arterial blood gas (ABG) analysis at time of admission revealed metabolic acidosis in 56% of cases and normal PH in 44% of cases.

Hypoxemia by pulse oximetry, [peripheral capillary oxygen saturation (SpO2) < 85%], was detected in 96% of cases. However by ABG analysis, all cases were hypoxemic [partial pressure of arterial oxygen (PaO2) < 85 mmHg]. P value between pulse oximetry and ABG was 0.137.

<table>
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<th>Table 1 The possible risk factors of CCHD.</th>
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<td>CCHD, cyanotic congenital heart disease.</td>
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According to echocardiography findings of our studied cases, the most frequent type of CCHD was d-transposition of great arteries (D-TGA) (33/50,66%), followed by complex CCHD (6/50,12%) and hypoplastic left heart syndrome (HLHS) (6/50,12%) then single ventricle (2/50,4%) and pulmonary atresia (2/50,4%), whereas the less common type was hypoplastic right ventricle (1/50,2%). Complex CCHD included 3 cases of D-TGA with pulmonary atresia and 3 cases of D-TGA with tricuspid atresia (Table 2).

All cases needed medical treatment in the form of one or more of the following [Prostaglandin E1 (PGE1) infusion, antifailure, inotropes, antibiotics, vitamin K or sodium bicarbonate]. Balloon atrial septostomy was performed in (13/50,26%) of cases. Thirteen neonates (26%) required mechanical ventilation while oxygen therapy was used in (26/50,52%) of cases (Table 3).

In our study, heart failure was the most common complication (50%), followed by severe chest infection (42%) and signs of septicemia (42%) while the less common complications were brain insult (12%) and polycythemia (8%) (Table 4).

Regarding the outcome of studied cases, 37 (74%) cases were improved and referred to a higher center as Abou-Elreesh Children Hospital, Cairo University to complete their treatment. Thirteen (26%) babies expired during hospital stay. There was no mortality among the cases with D-TGA, hypoplastic right ventricle and single ventricle, while mortality rate was 100% in cases of pulmonary atresia and hypoplastic left heart syndrome (HLHS), and (5/6,83.3%) in cases with complex CCHD (Table 5).

As regards the possible causes of death, the most common causes were irreversible shock (16%), followed by delayed referral to a higher center (8%) and the less common cause was central apnea (2%).

4. Discussion

Congenital heart disease (CHD) has already been known as an important cause of significant morbidity and mortality in neonatal period. Neonatal unit is the best place for screening and diagnosis of CHD [11]. The objective of the present study was to describe the clinical profile and available therapeutic modalities used in the management of CCHD in NICU at Sohat University hospital. During the one year study period 50 neonates were diagnosed with cyanotic congenital heart disease. Cyanotic congenital heart disease frequency was found to be 9.5% (50/524) in our NICU population. Shima et al. [12] reported that the occurrence rate of CHD is nearly 4.5% of admitted neonates in neonatal intensive care unit. In another study, it was found to be 4% of NICU population [13]. Ekici et al. [14], in a prospective study for one year, found that the incidence of CHD in all admitted neonates to NICU to be 3.49%. Our frequency rate is higher than other studies; this may be due to the reason that our NICU is the only tertiary care unit and a referral unit for critically sick neonates from 2 Governorates (Sohag and Quena).

In the present study of CCHD, there was male predominance with male to female ratio 3.55:1. This is consistent with a recent study in Pakistan (50 males/37 females) [15]. Also, Zahid et al. [16], in Peshawar, Pakistan, stated a male to female ratio of 1.3:1. Shah et al., in Nepal, found that male to female ratio was 1.5:1 [17]. However, our finding is not similar to that reported in Saudi Arabia [18] and Iceland [19], where the frequency was the same for males and females while in a study in Nigeria, there was female predominance in CHD [20]. This can be explained by differences in race and ethnic groups.

In our study the most common presenting symptoms were central cyanosis (100%), followed by respiratory distress (82%), and feeding difficulties (62%) in the form of poor sucking, poor feeding and interrupted feeding whereas the less...
common presentations were generalized convulsions (12%), and bleeding tendency (4%). This was in agreement with other studies done by Humayun and Atiq [21] and Dorfman et al. [22]. However in a study by Masood et al. [23] more than two-thirds of cases presented with respiratory distress while cyanosis was present in 40% of cases. This difference may be due to difference in age of studied cases.

Positive parental consanguinity was encountered in 34% of our cases. This in line with the study done by Settin et al. [24] in Mansoura Locality, Egypt, where positive parental consanguinity was found in 18.8% of CHD cases. Also, Bassili et al. [25] reported high rate of positive parental consanguinity as a risk factor for CHD. In addition, in a study done in Iran, parental consanguinity was found in 39.6% of cases with CHD [26].

Other risk factors for the development of CCHD in our study were maternal drug intake (20%), maternal infection during 1st trimester of pregnancy (16%) and positive family history of CCHD (12%). This came in agreement with other studies [27,28]. However positive family history was found only in 2.2% of neonates with CHD in a study done by Hussain et al. [15].

In our study 96% of cases were hypoxemic (SpO2 < 85%) by pulse oximetry. Meanwhile all cases were hypoxemic (PaO2 < 85 mmHg) by ABG analysis. P value between pulse oximetry and ABG was P = 0.137. So pulse oximetry is a good, easy, non invasive, and cheap method for early diagnosis and detection of degree of hypoxemia but it is less accurate and should be confirmed by ABG analysis. This was in line with a study done by Riede et al. [29] as they found that pulse oximetry was a good screening tool for detection of duct-dependent pulmonary circulation, as it has detected 100% of cases.

Transposition of great arteries (D-TGA) was the most frequent type of CCHD in our patients with a frequency of 66%, followed by HLHS (12%) and complex CCHD (12%), meanwhile the less common types were single ventricle (4%) and pulmonary atresia (4%) followed by hypoplastic right ventricle (2%). Our work was in agreement with studies done by Islam et al. [28] and Farooqui et al. [30]. However in studies done by Patra et al. [4], Hussain et al. [15], Humayun and Atiq [21] and Zahid et al. [16], the most common types of CCHD were Tetralogy of Fallot (TOF) followed by D-TGA. This difference can be because of inclusion of only neonates in our study while other studies included older children and usually Tetralogy of Fallot presents after few months of life.

All cases needed medical treatment in the form of one or more of the following (Prostaglandin E1 “PGE1”, antifailure, inotropes antibiotics, vitamin K, sodium bicarbonate). Balloon atrial septostomy was performed in 26% of cases. Thirteen neonates (26%) required mechanical ventilation while oxygen therapy was used in 52% of cases. The majority of our cases (74%) were improved by using these modalities. Neutze et al. [31] stated that PGE1 infusions have been used to increase ducatal patency in 11 cases with CCHD, all of them showed a satisfactory increase in oxygen saturation attributed to dilatation of the ductus. In a study done by Kovacekova et al. [32], continuous infusion of PGE1 was administered in 84.6%, dopamine in 30%, and Na bicarbonate was used in 34.6% of the cases. Meanwhile 30.8% of cases were mechanically ventilated and 34% were referred to tertiary hospital.

In our work the most common complications were heart failure (50%), followed by severe chest infection (42%), signs of septicemia (42%), while the less common complications were brain insult (12%) and polycythemia (8%). This was in accordance with other studies [17,21].

Regarding the outcome of studied cases, 74% of cases were improved and then referred to a higher center as Abo-Elreesh Children Hospital, Cairo University to complete their treatment. Thirteen patients (26%) expired during hospital stay. The mortality rate of 26% is more than that reported in other studies where Shah et al. [17], in Nepal, reported 20% mortality rate among patients with CHD and also, Jacobs et al. [33], in a study from Hong Kong reported 20% mortality in cyanotic heart disease patients. However, the mortality rate is higher in the study by Humayun and Atiq [21], where mortality rate was 36.4%. The difference may be due difference in the study population and the availability of cardiac facilities.

The most common causes of death were irreversible shock (16%) mostly carcinogenic shock and septic shock, followed by delayed referral to a higher center (8%) and the less common cause was central apnea (2%) due to extensive cerebral infarction and severe ICH. In the study done by Humayun and Atiq [21], the causes of death were surgical complications in 2 (4.5%) cases and medical problems in 14 (31.8%) babies such as sepsis and pneumonia and associated extra-cardiac malformations.

5. Conclusion

Congenital heart disease (CHD) is the most common congenital malformation presenting in the neonatal period with cyanotic defects representing 25% of all cases of CHD. Cyanotic congenital heart disease (CCHD) is a leading cause of neonatal morbidity and mortality. CCHD frequency was found to be 9.5% in our study population. Transposition of great arteries was the commonest CCHD diagnosed in our patients. Majority of neonates with CCHD showed survival with suitable management. Early diagnosis and referral to pediatric cardiac center for proper management will improve the outcome. Significant proportion of studied cases had complex cyanotic heart defects, so neonatologists and pediatric cardiologists should be familiar with diagnosis and management of these defects.

6. Limitations of the study

Our study had some limitations as the study included only a single center. Also the study was for only one year duration, this was not an enough period for catching all the cases with CCHD and we do not know the outcome of neonates referred to the cardiac centers for surgery. It is a hospital based study and may not reflect the true incidence or prevalence of CCHD in our community as the study included only neonates admitted in NICU at Sohag University Hospital.

Conflict of interest

We have no conflict of interest to declare.

References


