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ORIGINAL ARTICLE

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

Ashraf Abou-Taleb *, **Marwa A. Abdelhamid, Mohamed A.M. Bahkeet**

Pediatric Department, Sohag University, Sohag, Egypt

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KEYWORDS

Cyanotic congenital heart disease;
Echocardiography;
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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.

* Corresponding author at: Pediatric Department, Faculty of Medicine, Sohag University, Sohag, Egypt. Mobile: +20 1004063306.
E-mail address: ashmaabu@yahoo.com (A. Abou-Taleb).

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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 (SpO_2) < 85%], was detected in 96% of cases. However by ABG analysis, all cases were hypoxemic [partial pressure of arterial oxygen (PaO_2) < 85 mmHg]. P value between pulse oximetry and ABG was 0.137.

Table 1 The possible risk factors of CCHD.

Risk factors	No.	(%)
Consanguinity	17	34
Family history	6	12
Maternal infection	8	16
Maternal drugs	10	20

CCHD, cyanotic congenital heart disease.