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Epidemiology and Evaluation of Congenital Cyanotic Heart Disease in Children- A Review

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Review Article

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ABSTRACT

Structure abnormalities of the heart or intrathoracic great vessels that arise during fetal development are known as congenital heart disease (CHD). It is the most frequent type of birth defect. Despite great success in surgical and medicinal care of CHD. many operations are palliative rather than curative. and some survivors still have significant residual hemodynamic and electrical conduction abnormalities. as well as long-term cardiovascular problems. The field of congenital heart defect treatment has come a long way in its infancy. With heart problems that were an

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automatic death sentence 60 years ago, to contemporary surgical survival rates of more than 96 percent for all defects evaluated together. No other branch of research or medicine has done so much in such a short time. In this paper, we overview epidemiology, evaluation and management of congenital cyanotic heart diseases in children.

Keywords: Heart disease; congenital heart disease; cardiovascular problems.

1. INTRODUCTION

Structure abnormalities of the heart or intrathoracic great vessels that arise during fetal development are known as congenital heart disease (CHD). Which is the most frequent type of birth defect. Non-cyanotic CHD and cyanotic CHD. often known as serious congenital heart disease (CCHD), are two types of CHD [1]. The cause of coronary heart disease (CHD) is still largely unknown. About 15% to 20% of newborns with CCHD have chromosomal abnormalities that have been identified. cardiovascular defects are believed to be responsible for 35% of infant mortality due to congenital malformations. and Children with congenital cardiac abnormalities has considerably lower weight and age [2].

Manv coronary heart diseases occur multifactorial, meaning they are caused by a combination of genetic susceptibility and environmental risk factors. but can occasionally be related to genetic diseases. Maternal including illnesses. diabetes and phenylketonuria. maternal exposure to chemicals or drugs, and viral infections during pregnancy are potential environmental risk factors [3,4].

Despite great success in surgical and medicinal care of CHD. many operations are palliative rather than curative. and some survivors still have significant residual hemodynamic and electrical conduction abnormalities. as well as long-term cardiovascular problems [5-9]. studies have shown that CVD risk factors such as hypertension and obesity are more common in patients with CHD than in people without CHD [10-15].

Since 1990. mortality has decreased by 60%. however even after surgical repair of the anatomical abnormality. people with congenital heart disease might have cardiac problems such arrhythmias. heart failure. and valve insufficiency [16].

In children with cyanotic congenital heart disease, mental-motor retardation is common and they are still observed many years after successful surgery and potentially manifest themselves in learning or behavioral difficulties. Patient-associated factors such as gender. lower birth weight. existence of genetic/phenotypic abnormalities. mother's educational position. and ethnicity. in addition to medical features linked to the heart problem. explained up to 30% of the variance in the child's development [17].

After heart surgery. infarction. ischemic strokes. and cerebral hemorrhage can occur. Children with cyanotic heart defects. such as TGA or TOF. have been reported to have poorer developmental outcomes than children with acyanotic heart malformations [14].

2. EPIDEMIOLOGY

Congenital heart disease (CHD) is the most common congenital abnormality detected in newborns. with a birth prevalence of 10% of live births recorded globally. 8.9% in china. Cyanotic congenital heart disease (CHD) accounts for up to 25% - 27.4% of all cases of CHD. The most common CCHD is Tetralogy of Fallot (fah-LO) . and TGA. premature babies have increased risk of having CHD. males also have more risk and that risk is increased if the child father has one of the risks factors such as age. poor obstetric history. prenatal febrile disease. Between 1970 and 2017. the global prevalence of coronary heart disease grew by 10% per five years. with over 90% of this rise likely owing to greater identification of milder lesions (VSD. ASD and PDA). The higher frequency of mild lesions is likely due to increased global usage of echocardiography and better echocardiographic methods. Among the six worldwide areas. Africa had the lowest CHD prevalence. which was reported as less than a 25% of some of the other regions. and that's probably due to the lack of diagnosing techniques such ECG and thus the number of cases is significantly lower than the other regions [18-24].

In 2000, 1 million adults in the United States were living with congenital heart disease. An estimated half of them had relatively simple residual disease, one quarter had moderately complex residual disease, and one quarter had severe residual disease. However, functional disability is not limited to those with "severe" disease, as even an individual with "simple" disease may be disabled due to a complication [25].

Every year. around 6500 children in Germany are born with a congenital heart defect. Eight to nine out of every 1.000 live births have congenital heart disease (CHD), of which CCHD accounts for about 25%. and they require surgical catheter-based immediate or intervention. The most common CCHD is Tetralogy of Fallot (fah-LO) (which accounts for 5% of all CCHDs). and TGA which accounts for 2% of all CCHDs. Patent ductus arteriosus is the most common noncyanotic lesion. followed by ventricular septal defect and atrial septal defect [18].

Analysis of 4.538 cases conducted for all children who were admitted to a pediatric hospital for the first time from Jan 1995 to December 1997. The diagnosis of all patients with CHD is at least confirmed by ECG. There were 2.017 (44.4%) children with congenital heart disease. 201 (4.4%) children with acquired heart disease. 52 (1.2%) children with arrhythmias. and 2.268 (50%) children who were healthy. The most common heart defect is a ventricular septal defect [19].

Regarding Congenital Heart Disease in Jordanian Infants. a total of 1.028 newborns were evaluated in the cardiology service. 865 people had abnormal echo findings. CC was found to be 25 per 1.000 live births. and premature babies accounted for 51% of the total. Cyanotic CHD accounts for 6% of all CHD cases [20]. Another study from India that indicated the epidemiology and risk factors reported that; (27.50%) Cyanotic CHD and (72.50%) noncyanotic. males accounted for 261 (65.25%). It was found that father's age. poor obstetric history. prenatal febrile disease and advanced age increased the risk of coronary heart disease [21].

In Nigeria; out of 352 people with CCHD were studied. male to female ratio of 1.34:1. Tetralogy of Fallot (TOF) was the most common kind of CCHD. followed by double outlet right ventricle (DORV) and transposition of the great arteries (TGA) [22].

In Hunan Province. China. CHDs were discovered in 6289 newborns out of 673.060.

The total frequency was 93.44 per 10.000 Pls. the risks of CHD were higher in urban newborns than in rural infants. The Predominant subtypes of CHDs; VSD. TOF. and ASD were the most prevalent forms among early-gestation fetuses. whereas ASD. PDA. and VSD were the most common types among Pls [1].

3. EVALUATION OF CHD

There's no particular symptoms for CHD. but Shortness of breath and limited capacity to exercise are common signs of congenital heart disease. as are exhaustion and an irregular sound in the heart called a heart murmur. which is confirmed by a physician while listening to the heart beats [26].

Physical examination. echocardiography. magnetic resonance imaging. cardiac computed tomography. cardiac catheterization. Continuous ambulatory blood pressure monitoring. MRI. PET CT. and other invasive radioisotope modalities. cardiac catheterization with angiocardiography and open-heart surgery are used to diagnose congenital heart disease [27,28].

Two-dimensional and/or Doppler echocardiography read by an investigator with experience in echocardiography in early newborns is the noninvasive reference standard for detecting structural congenital heart disease in newborn infants. Lactate. troponin. creatine kinase (CK). and creatine kinase myocardial band (CK-MB) are examples of cardiac indicators which can help in diagnosing [29,30].

The ICD-9 diagnosis codes for congenital cardiac disease are mostly in the range of 745.0 to 747.9 [31]. For TOF. A normal-sized heart silhouette with an upturned apex and a concave major pulmonary artery section. also described as "boot-shaped." can be seen on chest radiographs [32].

All children with HF should have their blood glucose and serum electrolytes. such as calcium and phosphorus. checked since imbalances can induce reversible ventricular dysfunction. In newborns with HF. screening for hypoxia and sepsis is recommended. In many researches about CHD the heart failure was the main underlying diagnosis. when the ductus arteriosus closes in people with duct dependent systemic circulation. it causes acute cyanosis or cardiogenic shock. other cardiac Patients with other various cardiac abnormalities will present with cardiac failure and/or respiratory distress. However, the majority of diagnostic criteria for HF are clinical. and numerous standardized diagnostic classification systems have been developed. Because of the wide range of ages at which HF manifests. pediatric HF encompasses a greater spectrum of concomitant symptoms than adult HF [32].

Cardiomegaly is indicated by a cardiothoracic ratio of >60% in newborns and >55% in older children on pediatric CXR. Cardiomegaly has a high specificity and negative predictive value for ventricular dilatation on echocardiography. but a poor sensitivity and positive predictive value [32].

Sinus tachycardia. LV hypertrophy. ST-T alterations. myocardial infarction patterns. and conduction blocks are the most prevalent ECG abnormalities in children with HF. In all cases of pediatric HF, a transthoracic echocardiogram is recommended to rule out any structural issues [33].

4. EVALUATION OF THE SITUATION

It's difficult for countries with high fertility rates and those of which that have more than 8 children per one family to afford the medical support and treatment for CHD. and unfortunately that makes family with more than one child with CHD in such countries unable to afford right medical support which increase the burden on the health care system overall that's why Improving local health services and infectious managing illnesses (diarrhoea. rheumatic fever. measles. and rotoviral infection) are vital. but they are only band-aids when compared to bettering education. empowering women. and lowering birth rates [34].

In TOF the most predictive risk factors with the age are QRS duration. right ventricular hypertrophy. and left ventricular dysfunction. Also, systemic ventricular dysfunction. according to current studies. becomes a substantial risk factor after the age of 20. However, Children have a better overall prognosis with HF than adults because HF in children is more often caused by structural heart disease and reversible diseases that may be treated. following the development of early surgical treatments [35]. The result of HF due to CHD has altered drastically. In the "early surgical period." the incidence of symptomatic HF has also decreased. Only 10% of their patients in a

tertiary care pediatric cardiology environment had symptomatic HF, according to Massin et al. [36].

Although significant progress has been achieved in the field of genetics in terms of diagnosis. which then influences prognosis and genetic counselling. the great majority of the etiology of congenital cardiac abnormalities is still unknown or incomplete [37].

Before the age of 2.5. all of the children had their lesion surgically corrected. Those who were at a higher risk of having difficulty in school due to known consequences of their heart illness or for reasons not directly related to the heart disease were eliminated. Children with cyanotic illness performed significantly worse in all academic subjects [38].

Many children are unable to feed regularly prior to heart surgery. necessitating frequent extended or enteral feedings. Children who lack typical muscle strength may experience developmental delays. Because of their weakness or intolerance to respiratory infections. they may be unable to attend day care [39].

Children may experience varying degrees of disability following cardiac surgery. At school age. 11 to 17% of children with congenital heart disease who have had an operation have severe impairments in adaptive behavior. sociability. communication skills. and daily living abilities [40].

Patients who have only received palliative surgery often have substantial functional restrictions for the rest of their lives. Individuals born with single ventricles. for example. have just 50 to 60% of their usual activity capacity for their age [41].

So even after receiving CHD treatment in many cases children left to suffer lifetime consequences or at least they still have drawbacks and cannot do some of the same tasks as their corresponding healthy mates do [42].

5. MANAGEMENT AND PROGNOSIS

Early detection and treatment of congenital heart disease (CHD) are critical for a positive result. Although HF caused by CHD is one of the main causes of death among children with congenital



Fig. 1. Low oxygen Levels in the blood cause the lips, fingers and toes to be cyanotic (blue)

diseases [43]. Treatment and management of HF in children in general has better overall prognosis than the adult and that because it's often caused by structural diseases and the reversible diseases can be treated. Treatment of the cause. correction of any triggering event. and treatment of systemic or pulmonary congestion are among the management concepts [44].

In many cases with CHD surgery is the main treatment and it can be divided into three categories:

- Curative: After surgical treatment of these disorders in childhood. patients with these conditions rarely experience long-term consequences. Patent ductus arteriosus. secundum atrial defect. and uncomplicated ventricular septal defect are the three conditions [45].
- Reparative: Patients with these defects improve following remedial surgery, but the effects last a lifetime, and some will experience considerable late impairment. Aortic stenosis, atrioventricular canal, coarctation of the aorta, partial anomalous pulmonary venous return. pulmonary tetralogy of Fallot, stenosis total anomalous pulmonary venous return, dtransposition of the great arteries, and Itransposition of the great arteries are all examples of these defects (also called congenital corrected transposition of the great arteries) [46].
- Palliative: Because surgery does not fully address the underlying abnormality in

these people (even if it is performed). they are likely to have considerable functional disability for the rest of their lives. Eisenmenger syndrome. hypoplastic left heart syndrome. malaligned atrioventricular canal with single ventricle repair. single ventricle. Tricuspid atresia. and untreated cyanotic heart disease are all examples of these disorders [47].

If not detected early, numerous causes of HF in newborns can manifest with acute circulatory collapse or proceed to shock. Many of these diseases necessitate the use of prostaglandin infusions or emergency operations like ductal stenting and balloon atrial septostomy to keep the ducts open [48].

In severe cases of endocarditis. a catheter technique and surgery are required to replace heart valves or undergo heart transplantation. however. persistent abnormalities exist in a considerable percentage of patients. some of which necessitate repeat catheter or surgical intervention. necessitating continuing follow-up after surgery [49].

Diuretics are used to relieve pulmonary or systemic congestion. inotropes are used to increase contractility. and vasodilators and other treatments are used to reduce the disproportionately increased afterload. Diuretics, digoxin, angiotensin-converting enzyme inhibitors (ACEIs), spironolactone, beta-blockers, and inotropes are all commonly used medicines in the treatment of heart failure in children [50].

Survival in individuals with congenital heart disease who reach adulthood is reduced. Death is commonly due to heart failure or sudden death, and risk is increased by arrhythmia, endocarditis, myocardial infarction, and pulmonary hypertension [51].

6. CONCLUSION

CHD is one of the most serious congenital diseases that may result in secondary HF which can lead to the death of the newborn. and even if he didn't die patients are left to face moderate to severe symptoms that in many cases non-curative.

The field of congenital heart defect treatment has come a long way in its infancy. With heart problems that were an automatic death sentence 60 years ago, to contemporary surgical survival rates of more than 96 percent for all defects evaluated together. No other branch of research or medicine has done so much in such a short time.

When we talk about Congenital heart disease, we mostly refer to a genetic drawback for the newborn. such a newborn was being put-aside by the natural laws of natural selection. today the situation has changed. and with the major breakthroughs that have been made in the medical field that has managed to give large number of newborn new chances to live. but even that is not enough because large number of surgeries that being made are not curative and the patient still have to face in many cases lifetime drawback. so, we still have long way to go. Improving the life of the current CHD patients and finding new curative methods. and also, early detection and if possible, prevention can sometimes provide an early solution.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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