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Research Article

The Effect of Multidisciplinary Approach: A Retrospective Analysis of Congenital Heart Defects from a Tertiary Referral Centre In Bucharest-Romania

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Abstract

Background: Congenital heart defects (CHD) are a common type of internal birth defect affecting newborns globally, with a prevalence ranging from 0.8% to 1.2% of all live births. CHD are a significant cause of morbidity and mortality in newborns and children worldwide. They can range in severity from simple defects to complex defects that require multiple surgeries and ongoing medical management. The mortality rate for CHD has decreased over the last decades due to improved surgical techniques and medical management. However, they still represent an important public health problem. Prenatal detection and confirmation of the diagnosis after birth, or screening of newborns suspected of having a heart anomaly, allows for early and accurate diagnosis for timely optimal care. **Material and methods:** We conducted a retrospective cross-sectional study, over five years (2016-2020), at Clinical Hospital of Obstetrics and Gynecology "Prof. Dr. P. Sârbu" in Bucharest, Romania, a level III maternity hospital. Patients were selected from the hospital database. **Results:** We included 375 patients, identified through prenatal or neonatal screening, that had clinically diagnosed, echocardiographic confirmed, and computerized angiography-confirmed congenital heart malformations. In our study, the incidence of congenital heart malformations was 2%. Among the identified cases, 41 had complex CHD, and 13 newborns had various genetic syndromes associated, with Down syndrome being the most common. Out of the total patients, 27 were sent to cardiovascular surgery clinics during the neonatal period. Eight patients, especially preterm infants, died due to the complicated nature of the heart malformation and other conditions. **Conclusions:** In CHD evaluation, there should be a multidisciplinary team involved. In our clinic, the number of newborns with CHD has increased due to effective collaboration between obstetricians, neonatologists, and pediatric cardiologists. This facilitated intrauterine transfer at any gestational age to a higher-level maternity hospital, intending to diagnose, monitor, and treat heart anomalies. Our results demonstrate the effectiveness of a multidisciplinary team and support the inclusion of a pediatric population surveillance program in the national public health strategy.

Keywords: Congenital Hearts Defects; Neonatology; Neonatal Intensive Care Unit

Introduction

Cardiovascular compromise with or without Congenital Heart Defects (CHD) is a common finding in many premature infants and severely ill term newborns. It is associated with an increased incidence of significant short- and long-term morbidity and mortality [1]. Often the cause of CHD remains unknown. Pregnancy-related medical history is useful in diagnosing certain congenital heart diseases risk factors, such as biological teratogenic factors (rubella virus, cytomegalovirus, herpes viruses, and Coxsackie type B), physical, chemical, drug-related risk factor (antidepressants, amphetamines, anticonvulsants, certain antibiotics, lithium, retinoic acid) or traumatic factors [2-4]. CHDs can be associated with known genetic syndromes: Trisomy 13, 18, 21, Noonan syndrome, Turner syndrome, DiGeorge syndrome [2,5,6]. Signs and symptoms suggestive of CHD depend on the type of malformation, with some newborns being asymptomatic, while others present with nonspecific manifestations. All newborns with suspected CHD should be investigated during the neonatal period [2,3,5]. Echocardiography is the most important diagnostic modality for CHD and is readily available to the clinician. There are also other diagnostic options such as Computed Tomography (CT), Magnetic Resonance Imaging (MRI) and CT angiography, which help establish an accurate diagnosis in cases of complex CHD [2,3,6,7].

Material and Methods

We conducted a longitudinal retrospective observational study that aimed to evaluate the prevalence of CHD in a tertiary referral center in Bucharest, Romania, investigating infants born with complex cardiovascular malformations admitted to our clinic over a period of 5 years (January 2016- December 2020). Objectives included describing the characteristic of newborn babies suspected and diagnosed with CHD. The inclusion criteria were antenatal or postnatal suspicion of congenital heart disease based on clinical or paraclinical data. Exclusion criteria were incomplete data, aneurysm of the systemic arteries, ductus arteriosus or patent foramen ovale without hemodynamic involvement. Patients with late-onset symptoms such as feeding difficulties and fatigue during meals, and growth failure were not included in this study. Data was collected from the hospital's electronic register using simple random sampling to reduce bias. Data collection was collected using Microsoft Excel software. IBM SPSS version 26 software was used for data analysis and graphical representation. The distribution was tested using the Shapiro-Wilk test. Prematurity refers to a gestational age below 37 weeks, with subcategories of late preterm (32-37 weeks of gestation), very preterm (28-32 weeks), and extremely preterm (<28 weeks) [8,9,10].

Results

After neonatal screening in the maternity ward the study cohort included 375 patients with suspected congenital heart disease. Out of them only 104 (27,73%) were confirmed with Congenital Heart Disease (CHD) following a cardiological consultation and echocardiography.

A quarter of the newborns included in the study (25.2%) were diagnosed antenatally. They were reevaluated postnatal by a cardiology specialist in order to confirm their diagnosis. Thirty cases (29.1%) had signs and symptoms on clinical examination suggestive of heart malformations such as cyanosis, SpO2<95%, respiratory distress, systolic murmur, heartbeat to the right of the sternum, and single umbilical artery. Other reasons for performing an echocardiography were multiple malformation syndromes, chromosomal anomalies, and newborns from diabetic mothers. Due to immaturity and fetal distress, premature babies, and those with Intrauterine Growth Restriction (IUGR) were referred for cardiological consultation. Table 1 contains the reasons the attending physicians had for requesting a cardiology consult and echocardiography.

Reason for referral to cardiology specialist		**No of neonates (n=375)**
Suspected CHD (antenataly diagnosed)		26 (6,9%)
Finding on clinical examination	Systolic murmur	231 (61,6%)
	Cyanosis, SpO2<95% or major clinical signs of cardiorespiratory distress	30 (8%)
	Heartbeat to the right of the sternum	2 (0,5%)
Congenital anomalies	Extracardiac anomaly (multiple malformations, Prune Belly syndrome, Smith-Lemli-Opitz, Lobstein malady)	7 (1,86%)
	Chromosomal anomaly (Down syndrome)	6 (1,6%)
	Two-vessel ombilical cord	4 (1,06%)
Prematurity		38 (10,1%)
Late preterm		26 (6,93%)
VLBW		8 (2,13%)
ELBW		4 (1,06%)
IUGR		20 (5,3%)
Maternal diabetes mellitus		11 (2,93%)

Table 1: Reason for referral of newborn babies from our medical center for cardiology consult and echocardiography in the timeframe 2016-2020.

CHD, congenital heart disease, IUGR intrauterine growth restriction, VLBW very low birth weight, ELBW extremely low birth weight.

This study includes 104 newborns diagnosed with cardiovascular complex malformations. More than half (59,2%) were male. Table 2 presents the characteristics of the study group. Most of them (95,1%) were born in our maternity ward. Only 4,9% were transferred from another hospital. Most of the patients (92,3%) were conceived naturally. Eight patients (7,7%) came from pregnancies obtained through IVF (in vitro fertilization). 19.2% of the patients of the infants diagnosed with CHD included in this study were diagnosed with intrauterine growth restriction. 20 patients (19,2%) had IUGR (intrauterine growth restriction). A considerable number of patients (36,5%) were preterm babies. Out of them, 26 newborn babies (68,4%) were late preterm, followed by very preterm, 8 (21,1%), and extremely preterm, 4 (10,5%).

Variables	Patients (n=104)
Gender of newborn babies male female	61 (59,2%) 43 (41,3%)
Pregnancy natural conception in vitro fertilization	96 (92,3%) 8 (7,7%)
The birth our maternity other maternity	99 (95,1%) 5 (4,9%)
Apgar score 8-10 6-7 <5	73 (70,1%) 19 (18,4%) 12 (11,6%)
Complications mild-moderate pulmonary hypertension severe pulmonary hypertension Other	22 (21,3%) 5 (4,8%) 3 (2,9%)
Orotracheal intubation	31(30,1%)
Underwent surgery Our country San Donato Lyon	27(25,9%)
Deaths	8 (7,8%)

Table 2: Characteristics of the study group.

The most common congenital heart defect (CHD) diagnosed was ventricular septal defect (VSD) (31,7%), followed by pulmonary artery stenosis (19,2%) and patent ductus arteriosus (11,5%). A high percentage of patients in the study experienced complications. Most of them had pulmonary hypertension and 31 (30.1%) required orotracheal intubation and mechanical ventilation.

Of the 104 patients diagnosed with complex CHD, 25.9% were managed in neonatal intensive care before being transfered to surgical correction centers (in Bucharest, Italy, Germany, and France). Despite the complexity of CHD, the percentage of preoperative surviving children was 92.2%, with a low percentage of deaths, which were also accompanied by comorbidities (multiple malformation syndrome, genetic syndrome, extreme prematurity, and others). Eleven angioCT scans were performed in five years (Table 3) (Figure 1-4).

Congenital heart disease diagnosed	Patients (n = 104)
Ventricular septal defect	33 (31,7%)
Pulmonary stenosis	20 (19,2%)
Patent ductus arteriosus	12 (11,5%)
Transposition of the great vessels	5 (4,8%)
Tetralogy of Fallot	5 (4,8%)
Coarctation of the Aorta	5 (4,8%)
Atrio-ventricular canal, pulmonary valve atresia	3(2,9%)
Aortic arch hypoplasia	3(2,9%)
Pulmonary valve atresia, wide subaortic VSD	3(2,9%)
Hypoplastic left heart	2 (1,9%)
Severe valvular pulmonary stenosis	2(1,9%)
Severe aortic valve stenosis	1(0,96%)
Dextrocardia + congenital corrected TGV	1(0,96%)
RV hypoplasia, Pulmonary valve atresia, tricuspid valve atresia	1(0,96%)
RV hypoplasia, complete atrio-ventricular canal	1(0,96%)
Tricuspid valve dysplasia	1(0,96%)
Common arterial trunk + VSD	1(0,96%)
Totally aberrant pulmonary venous return in the coronar sinus	1(0,96%)
Clamp the isthmic aorta	1(0,96%)
Aortic bicuspid	1(0,96%)
Total abnormal venous return, IVC interrupt	1(0,96%)
Aortic arch interrupt, dextrocardia	1(0,96%)

Table 3: Congenital heart diseases diagnosed in the study group.

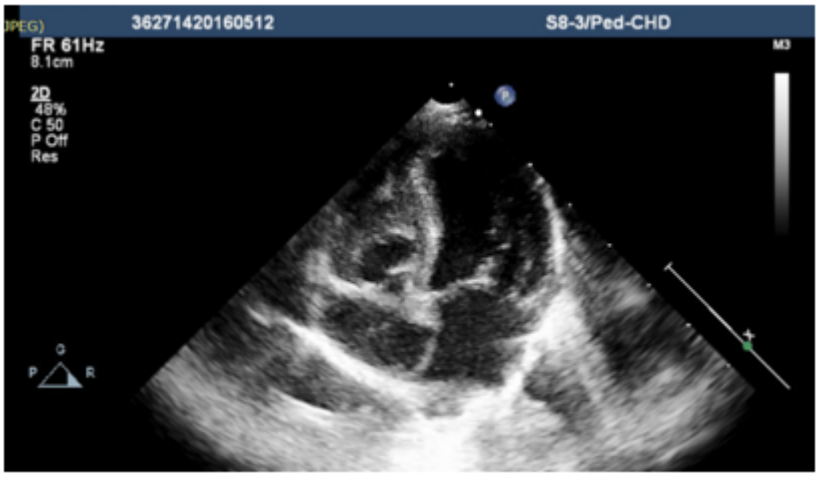

Figure 1: Echocardiography performed for one of the patients included in the study. Apical 4-chamber view: right ventricle hypoplasia with intact interventricular septum.

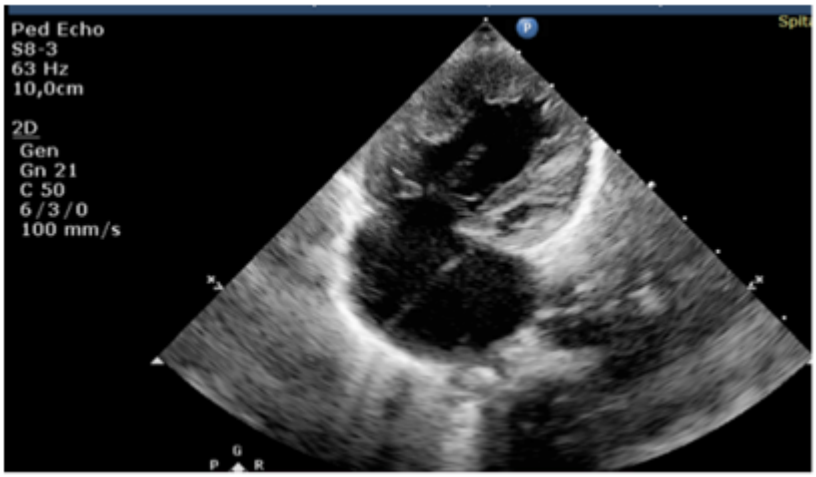

Figure 2: Echocardiography performed for one of the patients included in the study. Modified Apical 4 chamber view: hypoplastic left ventricle hypoplasia with dilated right cavities.

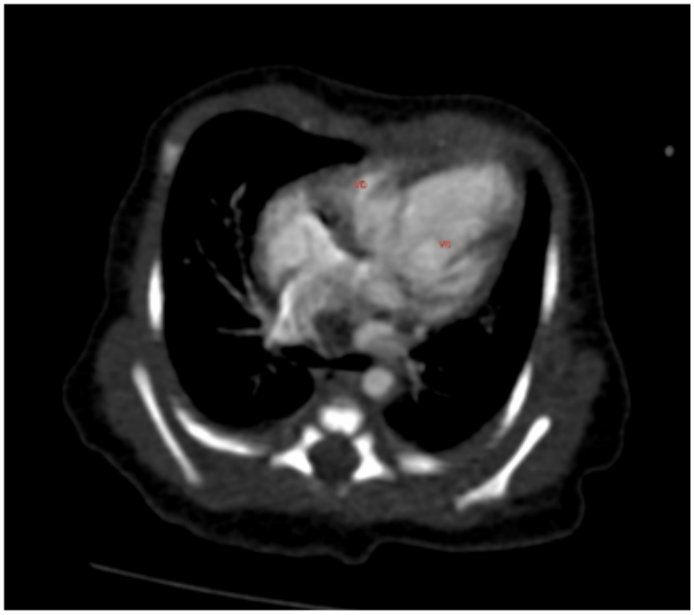

Figure 3: Computer tomography performed for one of the patients included in the study. Four Chamber plan showing severe hypoplastic right ventricle.

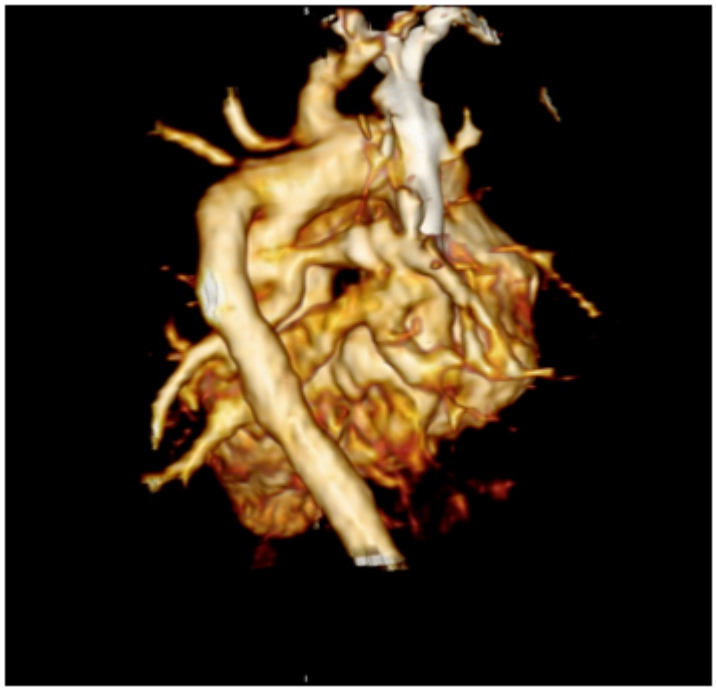

Figure 4: 3D reconstruction performed for one of the patients included in this study. The pulmonary veins form a common collector draining in the right atrium. there is also an aorta-pulmonary duct-like collateral.

Discussion

During the five-year study period (2016-2020), 18,519 newborns were admitted to our clinic. Out of them 375 were suspected to have Congenital Heart Defects (CHD), some based on fetal morphologies and others after neonatal screening performed in the maternity ward. The resulting prevalence for suspected CHD was 2.02%. Only 0.56% were confirmed with CHD, corresponding to the data reported by other studies [3,11–13].

The family medical history, especially the maternal one, including pathological finds during pregnancy such as raising suspicion of congenital heart disease on fetal echocardiography, inadequate fetal development, and/or problems at birth or postnatal, represent additional risk factors for congenital heart anomalies. These newborns require postnatal evaluation [5].

In 2020, 10 newborns were prenatally diagnosed with CHD, accounting for 40% of all prenatal diagnoses, indicating excellent collaboration between the pediatric cardiologist and the team of doctors in our NICU-equipped maternity unit, despite the start of the Covid-19 pandemic. This collaboration led to a reduction in neonatal morbidity and mortality. When the attending physician of the pregnant mother suspects a fetal CHD, they sometimes ask for second opinion. The next step would be requesting a pediatric cardiology consult during which the diagnosis is reevaluated treatment options are discusses. Birth in a 3rd level unit is recommended. The neonatology team is informed about the future birth of a child with CHD in the clinic in order to prepare for the management of the infant. The neonatology team continues communication with the pediatric cardiology specialist after the birth of the child. In our country the parents of babies with such conditions often experience multifaceted stress due to a lack of information, emotional impact, or financial strain. They must be informed and referred to specialized units equipped with specific equipment to complete their pregnancy.

During the last two years of the study, there were no newborns with CHD transferred to our clinic from other hospitals, and antenatal addressability has increased considerably. Thanks to good collaboration between pediatric cardiologists, obstetricians, and neonatologists, some fetuses were diagnosed prenatally and transferred intrauterine to our level 3 maternity hospital, where the pregnancy was completed, and the newborns received superior management.

Echocardiography is a non-invasive investigation used to diagnose Congenital Heart Disease (CHD) at any age, including newborns, with results significantly correlated with those obtained by invasive investigations [2,14]. It can contribute to establishing therapeutic indications, including interventional cardiology or cardiovascular surgery [15,16]. This fact was also reflected in

our study. Echocardiography helped finding optimal management for each case among the 104 newborns diagnosed with various degrees of CHD.

Often, newborns with cardiac anomalies are asymptomatic at birth and in the first few hours postnatal, with cardiac distress symptoms occurring later [5]. Current guidelines recommend initial screening for congenital cardiovascular diseases in the neonatal period (at 24 hours and 36 hours post-birth) , followed by a re-examination at 6-8 weeks of age [5,17].

Sometimes an undiagnosed CHM can be a significant cause for the delay in growth of the infant's stature and weight [2,5]. During their hospital stay, all newborns with Congenital Heart Disease (CHD) received a diagnosis via a clinical examination at birth and daily assessments during the early neonatal phase. Infants who showed signs of late-onset CHD symptoms during the neonatal period, such as feeding difficulties, fatigue during meals, or growth failure, were included in this study. Children with such symptoms may escape early diagnosis due to the lack of prenatal diagnosis and birth in lower-level (I and II) maternity units or the lack of screening [18].

The distribution of premature infants who required referral to a cardiologist was relatively uniform during the five-year study period. One of them, a Very Low Birth Weight (VLBW) infant, required surgical ligation of PDA in our maternity unit, avoiding the stress of transfer, by an Italian-Romanian cardiovascular surgery team. Initially, the neonatologist tried to close the arterial duct medically, but without a favorable response. The VLBW newborn was born in 2017 at 25 weeks of gestation, with a birth weight of 750g. He requiring long-term orotracheal intubation.

More than 70% of the 104 newborns with CHD had an Apgar score>8, and only 11.6% had moderate-severe perinatal hypoxia, an Apgar score <5. A transient systolic murmur of tricuspid regurgitation is an innocent murmur that disappears after 1-2 days but may persist if there is a much-increased pulmonary vascular resistance, disappearing when the resistance decreases. Fetal distress and severe perinatal hypoxemia, which maintain high levels of pulmonary vascular resistance, lead to a longer persistence of this murmur [5,19].

Murmurs detected in newborns can be innocent, functional, or pathological, with innocent murmurs being ten times more common than pathological ones [5,19]. Murmurs are most often detected during a routine clinical examination of a newborn and represent an essential element in guiding towards a congenital heart anomaly in 80% of cases [5]. More than half of newborns included in this study (61.6%) had a systolic murmur on clinical examination in the early neonatal period (0-7 days), requiring referral to a cardiologist. But only 42.91% of them had pathological murmurs, being diagnosed with CHD. The rest of newborns with heart murmurs were excluded from the study since it was due to secundum atrial septal defect, SIA aneurysm, or PDA in the process of closure. Good collaboration with pediatric cardiologists and a thorough clinical examination in the maternity unit resulted in a reduction of morbidity and mortality due to CHD in our maternity unit. The data aligns with previous information from literature indicating that most murmurs identified within the initial few days of life (up to 54%) are indicative of pathological conditions [5].

In our study 8% of the newborns referred for cardiology consult, and echocardiography had cyanosis, SpO2<95% and/or respiratory distress. They all underwent pre- and postductal SpO2 monitoring, hyperoxia test, ABG, cardio-pulmonary X-ray, measurement of blood pressure in all four limbs, and echocardiography. Screening for CHD should include prenatal ultrasound, physical examination and pulse oximetry screening [17]. According to the American Heart Association and the American Academy of Pediatrics, the sensitivity of pulse oximetry for critical congenital heart disease is 70% [5,20]. We have to keep in mind that pulse oximetry is not able to detect acidosis, hypercapnia and hypoxemia [21]. Analysis Of Blood Gases (ABG) is indicated for all newborns with respiratory distress, even if pulse oximetry is normal [5,19,20,21–24].

In this study, measuring blood pressure in all limbs, helped diagnosing 5 (4.8%) newborns with aortic coarctation. According to literature, the incidence of aortic coarctation represents 5-8-10% of CHD with a male predominance (2/1). In these patients arterial blood gases (ABG) are normal. The diagnosis is confirmed by 2D echocardiography and color Doppler [5,23–27].

The most frequently diagnosed CHD in our study was ventricular septal defect (31,7%). The rate was higher than the one reported in other studies [28–30]. Symptoms of patients with Ventricular Septal Defect (VSD) depends on the presence of hemodynamically significant shunt, being directly related to the size of the defect. Therefore, many patients can be asymptomatic, and some ventricular septal defects close with time. There prevalence of VSD within population varies between studies depending on mode of diagnosis and age of patients [31]. It tends to be higher in those where echocardiography was used [32,33] as was in our study.

One of the most frequent complications of CHD in our study was pulmonary hypertension (26,1%). Persistent Pulmonary Hypertension Of The Newborn (PPHN) reflects a disruption in the transition from normal fetal circulation to neonatal circulation. The disease is characterized by persistent high Pulmonary Vascular Resistance (PVR), as opposed to the normal decrease observed at birth [22]. Delayed diagnosis/adequate therapy in some cases of complex CHD with significant left-right shunts favors the

early onset of irreversible Pulmonary Vascular Disease (PVD), even at a young age, reducing the chances of subsequent surgical intervention [5,22]. In our Neonatal Intensive Care Unit (NICU), which is 3rd a level unit, the management of these patients was prompt, including endotracheal intubation, mechanical ventilation, and nitric oxide administration.

Recently, the number of newborns with genetic and multiple malformations syndromes has decreased due to better access to family doctors, obstetricians, geneticists, pediatric cardiologists (fetal echocardiography), maternal-fetal medicine (morphology TI and II). Pediatric cardiology has developed a lot in our country in recent years [2].

This study has certain limitations. It was a single-center study that serve as an argument for requiring better care for newborn babies with congenital heart disease in our country. It offers only a glimpse of the national situation. In addition, due to the lack of a national registry for patients with CHD we were not able to include those infants who showed late signs of CHD, not being diagnosed during their stay in the maternity ward.

Conclusions

Screening for detection of Congenital Heart Defects (CHD) is crucial in all newborns in the maternity ward using clinical evaluation. If any suspicion of CHD exists, the clinical assessment must be accompanied by paraclinical investigations. Delaying early diagnosis during the neonatal period can lead to an increased in morbidity and mortality associated with this pathology. Therefore, it is essential to implement CHD screening protocols in all maternity hospitals, regardless of their grade.

To prevent the onset of CHD in fetuses, it is necessary to have a multidisciplinary team that can recognize all risk factors and ensures early diagnosis in the maternity ward, followed by monitoring and treatment of congenital heart defects. Neonatologists are responsible for identifying, coordinate care and medically manage newborns with these pathologies. Therefore, it is essential for all doctors, especially neonatologists, to stay up to date with the latest research, guidelines, and techniques related to the diagnosis, treatment, and management of CHD. By improving medical knowledge and promoting collaboration among healthcare professionals, we can ensure that newborns with CHD receive the best possible personalized care (based on their specific needs and medical condition) and achieve the best possible outcomes.

Although Pediatric Cardiology became a separate specialty in our country in 2017, there is a shortage of centers and specialists in the field of pediatric cardiac interventional surgery. As a result, many cases must be transferred directly from the maternity ward to other countries.

Based on this study, recommendations for the development of a surveillance program for pediatric population with CHD were formulated. The first step would be establishing a national registry of children with CHD that includes demographic, clinical, and outcome data. The registry can be useful to monitor disease trends, evaluate treatment outcomes, and identify areas where further research is needed. Implementing a system for screening newborns for CHD using non-invasive methods such as pulse oximetry would be the next step. Early detection can lead to better outcomes and reduce the risk of complications. There is a need to develop guidelines for the management of children with CHD that are based on the latest scientific evidence and best practices. These guidelines should be regularly updated to reflect new research findings and evolving medical practices. Last but not least, increasing funding for research on CHD in children is needed to improve our understanding of the disease and develop new treatment strategies. Finally, quality control measures should be implemented, and regular evaluation of the surveillance program should be conducted to assess its effectiveness and identify areas for improvement. We hope that our recommendations can serve as a roadmap for policymakers, healthcare professionals, and researchers working to address this critical public health issue.

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