Pattern of Congenital Heart Disease in Newborn at a Tertiary Care Hospital

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Abstract

Background: Congenital heart disease (CHD) has been defined as a gross structural abnormality of the heart or intrathoracic great vessels that are actually or potentially of functional significance. CHD accounts for about 10% of newborn deaths and nearly half of all deaths due to congenital malformations in developed countries. The vast majority of newborns escape early intervention. The most important role of a pediatrician today is to ensure that, as far as possible, serious CHD should not be missed, especially in the neonatal period and infancy where maximum attrition for CHD occurs.

Objective: The objective of the study was to identify the pattern of CHD using echocardiography in newborns born in a tertiary care hospital in Tamil Nadu.

Methodology: A cross-sectional observational study conducted for 1 year. One hundred and fifty newborns diagnosed to have CHD after they underwent routine clinical examination and pulse oximetry, followed by echocardiography, were included in this study. Study design: This is a cross-sectional study. Place of study: Government Theni Medical College. Study period: 1 year. Sample size: 150.

Inclusion Criteria: Newborns diagnosed to have CHD confirmed with echocardiography were included in this study.

Exclusion Criteria: Newborns whose parents refused to provide consent were excluded from the study.

Results: This study group includes 56% of girls and 44% of boys. It shows that 83% are term babies and the remaining 17% from preterm groups. Among these, 146 babies had acyanotic heart disease as 97% and rest four newborns as 3% had cyanotic disease. Among 146 acyanotic heart disease, 76 newborns (52%) had atrial septal defect (ASD), followed by patent ductus arteriosus (PDA) in 59 newborns (40%), ventricular septal defect (VSD) in 15 newborns (10%), pulmonary hypertension in eight newborns (5%), and magnetic resonance in only one newborn (0.6%). Among four newborns presented with cyanotic heart disease, two newborns presented with total anomalous pulmonary venous connection (50%), followed by transposition of great arteries (25%) and tetralogy of Fallot TOF (25%).

Conclusion: In our study, the pattern of CHD is ASD, followed by PDA and VSD.

Keywords: Congenital heart disease, Echocardiography, Newborn, Pulse oximetry

INTRODUCTION

Heart diseases constitute an important group of pediatric illness and major causes of childhood morbidity and mortality. They may be symptomatic or asymptomatic. Late diagnosis of heart disease in children carries a high risk of mortality and morbidity. To avoid this, mortality and morbidity early diagnosis is important. Heart diseases in children may be congenital or acquired. Congenital heart disease (CHD) is the one present since birth. Some cases of CHDs are asymptomatic and are diagnosed on routine health visits. It is important to mind that children with CHDs are at increased risk of poor growth. The factors which play a role in poor growth may be feeding difficulties, excessive caloric requirement, and the cardiac lesions on growth and development.

Classification of CHDs in Children

Congenital lesions are divided into two major categories: Acyanotic and cyanotic. Acyanotic lesions further divided
into three main categories.\[^5\]

1. **Left to right shunt**
2. **Acyanotic obstructive lesions**
3. **Acyanotic regurgitant lesions**

Cyanotic lesions are further divided into two major categories.

1. **Lesions with decreased pulmonary blood flow.**
2. **Lesions with increased pulmonary.**

**Definition of CHDs:**

1. **Normal:** No echocardiographic abnormality or any of the following:
   - Patent ductus arteriosus (PDA) < 2 mm in size without volume overload of the left ventricle,
   - Patent foramen ovale or atrial septal defect (ASD) < 5 mm without volume overload of the right ventricle,
   - Mild turbulence at branch pulmonary arteries.
2. **Insignificant CHDs:**
   - Very small muscular ventricular septal defect (VSD), as these is likely to close spontaneously.
3. **Significant CHDs:**
   - These are divided into minor and major CHDs:
     a. **Minor CHD:**
        - ASD > 5 mm, PDA > 2 mm with left ventricle volume overload, restrictive VSD, and valvular aortic/pulmonary stenosis with gradients < 25 mmHg.
     b. **Major CHD:**
        - CHD was likely to require intervention within the 1st year, including newborns with critical CHD that require intervention within the first 4 weeks of life.

Further divided into two categories; acyanotic and cyanotic CHDs;

i. **Acyanotic CHD:** Nonrestrictive VSD, valvular aortic/pulmonary stenosis with gradients > 25 mmHg, and coarctation of aorta (nonduct dependent).

ii. **Cyanotic CHD:** Hypoplastic left heart syndrome, transposition complexes, aortic arch interruption, univentricular heart, tetralogy of Fallot (TOF), and TOF-such as conditions associated with pulmonary stenosis or atresia, total anomalous pulmonary venous connection (TAPVC), persistent truncus arteriosus, and Ebstein's anomaly.

4. **Other cardiac findings on echocardiography**
   - Persistent pulmonary hypertension (PHT) of newborn, cardiac tumor, situs inversus dextrocardia, and arrhythmias (complete heart block, and supraventricular tachycardia).\[^9\]

**Clinical Evaluation of the Child with Heart Diseases**

The clinical history, physical examination, chest radiography, pulse oximetry, and echocardiogram are the keystones in the diagnosis of CHDs in newborns.

Symptoms of CHDs are variable, sometimes asymptomatic, and subtle and may manifest anytime from fetal period to adulthood. Usually, critical CHDs present early in life. Some may go undiagnosed and identified in later life incidentally because of asymptomatic murmur.

Some cases of CHDs are asymptomatic and presents with an only heart murmur.\[^6\] The main symptoms of CHDs are mostly non-specific such as poor feeding, failure to thrive or poor weight gain, and poor exercise tolerance. Developmental delay may be a presenting symptom. Easy fatigability and diaphoresis may be the other presenting symptoms. Easy fatigability presents as difficulty in feeding in newborns and in infants.

CHD affects neurodevelopment across the lifespan. In infants, the developmental delay can occur, ranging from mild hypotonia to persistent delay affecting many aspects of development including language, social skills, and feeding. The spectrum of neurodevelopmental impairment is wide. Some children have minimal or no impairment, whereas others are severely affected. In general, children with milder forms of diseases such as ASD and VSD have fewer neurodevelopmental sequelae than those with complex lesions such as single ventricle or hypoplastic heart syndromes. However, medical, environmental, and genetic factors all play a role.\[^8\]

All newborns recruited into the study were screened using transthoracic echocardiography after the initial evaluation but within 48 h of life.

**Some Common CHDs**

**ASD**

Fossa ovalis ASD: Located in the central portion of atrial septum, in the position of foramen ovale. This type is amenable to closure by cardiac catheterization. Overall, the most common type (Ostium secundum type).

Sinus venosus ASD: At the junction of Superior vena cava and right atrium most commonly (Superior vena caval type).

Ostium primum type: Due to failure to over seal the septum primum most commonly seen in Down's syndrome.

Coronary sinus ASD: Defect in the roof of the coronary sinus.

**Treatment:** Surgical closure is better done before school entry to avoid late complications. Small defects < 8 mm can be observed. Fossa ovalis with good margins may be closed percutaneously in the catheterization lab. Other defects need surgical closure.\[^9\]
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VSD
Most common type of CHD is VSD (15-20%) of which Perimembranous VSD is most common type. Other types are inlet, outlet, and trabecular. Spontaneous closure is possible in small muscular types, about 30%.

Treatment: Medical treatment for congestive cardiac failure (CCF), RRTI, IEC, and anemia if occurs.

Surgical closure indicated in larger defects, evidence of ventricular volume overload, progressive aortic valve diseases, infundibular defects, chamber enlargement, and pulmonary arterial pressure more than 50% of systemic pressure.[10]

PDA
It is the persistence of normal fetal channel connection between aorta and pulmonary artery. Functional closure usually occurs between 12 and 24 h. Prematurity is associated with delayed closure of PDA.

Treatment of PDA: Indomethacin and ibuprofen can be tried in preterm infants in the neonatal period. Paracetamol therapy also has promising results and an alternative for PDA closure when indomethacin is contraindicated.[11]

After 10 days of post-natal age, the ductus rarely responds to medical therapy. Such patients need non-surgical closure such as coil closure and with occlusive devices.

Acyanotic Obstructive Lesions
Aortic stenosis
The most common cause is the bicuspid aortic valve. Severe forms of stenosis present early in the newborn. Less severe forms present later in life. Supra valvular aortic stenosis is seen in association with Williams’s syndrome.

Treatment: Pressure gradient <50 needs regular follow-up.
1. Balloon valvuloplasty (Isolated stenosis without aortic regurgitation)[12] indicated in pressure gradient more than 50 with symptoms or ST-segment and T-wave changes in the electrocardiogram. Pressure gradient more than 75 without any symptoms.
2. Aortic valve surgery or valve replacement is indicated in failed valvuloplasty and cases associated with aortic regurgitation. Aortic valve replacement with a mechanical prosthesis or Ross surgery with pulmonary autograft is the surgical procedure.[13]

Coarctation of aorta
COA manifestation in CHD is 6-8%. Usually manifests as a discrete constriction of the aortic isthmus. Presence of aortic arch hypoplasia is relevant in developing hypertension.[14]

Clinical presentation depends on the presence of other lesions such as VSD and PDA. Severe disease presents as collapse or shock-like state in newborn after ductal closure. The infant may present with CCF. The hallmark of physical finding is discrepancy between upper limb and lower limb pulses and blood pressure.

Treatment: Treatment of CCF with inotropes and diuretics, followed by surgical repair in newborn. Percutaneous Balloon Angioplasty and stenting can be done as a bridging procedure in children with CCF also.[15]

Resection and end to end anastomosis and subclavian flap repair are the most common surgical approaches. Aneurysm and rupture are the most common complication, and this needs reintervention.[16-18]

Pulmonary stenosis
Valvular form is the common type. Other types are subvalvular and supra valvular. In the common form, the valve is thickened with fused or absent commissures. The pulmonary valve is dysplastic in Noonan syndrome. Supra valvular pulmonary stenosis often refers to the narrowing of the pulmonary artery branch. Most patients are asymptomatic and well developed. Critical pulmonary stenosis presents with cyanosis.

Treatment: Mild stenosis (Pressure gradient <50) needs yearly follow-up. Moderate (pressure gradient 50–79) and severe stenosis (pressure gradient more than 80) need balloon pulmonary valvotomy. Balloon pulmonary valvotomy has excellent short-term and long-term outcome.[19] Surgical valvotomy is indicated in patients with dysplastic valves and severe stenosis, which failed to respond to balloon valvotomy.

Common Cyanotic CHDs
Tetralogy of Fallot’s
The four components of TOF are VSD, aortic override of VSD, right ventricular outflow tract obstruction, and right ventricular hypertrophy. Other associated anomalies are valvular pulmonary artery stenosis, right-sided aortic arch, and ASD. About 5% of cases have an anomalous origin of the left anterior descending artery from the right coronary artery. Clinical severity of TOF varies with the degree of pulmonary stenosis. With mild obstruction, systolic murmur is the only presenting symptom known as pink tetralogy. With severe obstruction, the patient presents with cyanosis.[20]

Treatment: Surgical correction accomplished by patch closure of VSD and right ventricular muscle resection with or without pulmonary valvotomy. Cases with anomalous left coronary artery may need temporary BLALOCK
TAUSSING shunt followed by more complicated intracardiac repair and takedown of the shunt.

**Transposition of great arteries (TGA)**
Aorta and pulmonary artery arising from wrong ventricles leading to deoxygenated blood in systemic circulation and oxygenated blood back to lungs. VSD is the common associated anomaly in 25% cases. Newborn with intact VSD presents with severe cyanosis within 24 h of life. Reverse differential cyanosis is the hallmark of TGA with intact septum. Other presentations in TGV are a failure to thrive and CCF.

Treatment: Arterial switch operation is corrective surgery. In addition, the coronary artery must be moved to the new aortic root. Most successful when performed age <2 weeks. If the diagnosis is made late, then atrial switch operation (Mustard and Senning) may be performed. Rastelli operation is the procedure of choice when TGA is complicated with pulmonary artery stenosis or left ventricular outflow tract obstruction and a VSD.[21,22]

**Total anomalous pulmonary venous return**
All the pulmonary veins drain to systemic veins or the right atrium instead of draining to the left atrium. The obstructive type of TAPVR presents within few hours of life with cyanosis and respiratory distress, and the cyanosis never responds to any non-surgical intervention. The unobstructed type presents in the neonatal period or infantile period with CCF, RRTI, and FTT.

Treatment: Reanastomosis of pulmonary venous confluence to the posterior wall of the left atrium is choice.

**Aim of the Study**
The aim of the study was to identify the pattern of CHD using echocardiography in newborns born in a tertiary care hospital in Tamil Nadu.

**MATERIALS AND METHODS**

It is a cross-sectional observational study conducted over a period of 1 year at Government Theni Medical College, Tamil Nadu, we screened newborns for CHDs using echocardiography. Nearly 7000 deliveries occur in this hospital every year, but babies after confirmed to have CHD with echocardiography were recruited into the study. The ethical committee approval was obtained.

**Informed Consent**
An informed consent sheet with details of the study protocol was given to one of the parents and approval sought in writing before recruiting the newborn in the study. None of the parents refused to provide consent.

**RESULTS**

After an initial evaluation, 150 newborns with significant heart disease are recruited in the study. Among these, 84 (56% of total sample size) are girls and 66 (44% of total sample size) are boys [Figure 1]; 124 term and 26 preterm babies [Figure 2]. The pattern of CHD in our study is illustrated in the bar diagram below. Among these, 146 newborns had acyanotic heart disease (97%) and the rest four newborns (3%) had cyanotic heart disease. Among 146 acyanotic heart disease, 76 newborns (52%) had ASD followed by PDA in 59 newborns (40%), VSD in 15 newborns (10%), PHT in eight newborns (5%), and magnetic resonance in only one newborn (0.6%). Among four newborns with cyanotic heart disease, two newborns presented with TAPVC (50%), followed by TGA (25%) and TOF (25%) [Figure 3].

**DISCUSSION**

In our study, girl babies outnumbered boy babies with a ratio of 1.27:1. We found that CHD was more common in female births, which was very similar to the study conducted in Nigeria[23] by Antia et al. which showed a female preponderance. However, our finding is not similar to that reported by Nikyar et al. from Gorgan[24] where there is a male preponderance and found out the ratio of male:female is 1.35. Alabdulgader et al. from Saudi Arabia[25] and Stephensen et al. from Iceland[26] reported that the frequency was the same for males and females.

In this study of pattern of CHD in our hospital, term child (83%) is affected more with CHD than preterms (17%). In
The most frequent type of CHD that we found in our study is to be ASD, which is in accordance with a recent study done in Saudi by Majeed-Saidan et al.\cite{28} Furthermore, another study in Iran\cite{29} cited the most common CHD in the newborn as ASD, while in other studies, the most frequent type of CHD was VSD.\cite{30-34} In our study, the pattern of CHD is ASD, followed by PDA and VSD. Majeed-Saidan et al.\cite{28} reported the pattern as ASD, followed by VSD. The study by Nikyar et al. in Gorgan, Northern Iran, reported the pattern as ASD followed by VSD and PDA.

**CONCLUSION**

CHD is one of the leading causes of morbidity and mortality in growing children in developing countries. Most of the babies born with CHD are expected to have a normal life when diagnosed and treated as early as possible. However, in developing countries, high incidence of preterm delivery and birth asphyxia CHD remains neglected and often overlooked.

The lack of awareness, nonavailability of trained staffs, and absence of screening programs for CHD in peripheral level leads to delayed diagnosis and poor outcome of CHD. By various training programs, recruiting skilled staff and availability of instruments at the peripheral level, we can ensure early diagnosis and treatment of CHD, to prevent a serious risk of avoidable morbidity, mortality, and handicaps.

This study shows that the most common acyanotic CHD is ASD, and the most common cyanotic CHD is TAPVC and TOF. Our study has certain limitation since it was done at the periphery hospital level.

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