Time to diagnosis, clinical features of patients admitted with congenital heart diseases in a teaching hospital

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Abstract
Objectives: The present study was conducted to evaluate age of diagnosis, clinical profile and echocardiographic findings of congenital heart disease in children of age-group 0-12 years admitted in tertiary care hospital

Material and Method: It was a prospective study conducted in the department of Pediatrics of Government Medical College & Hospital over a period of one year from January 2017 to January 2018. 207 children from birth to 12 years of age admitted in the hospital who had congenital heart disease confirmed by echocardiography were included. All patients were treated conservatively and observed for immediate outcome during the hospital stay.

Results: Out of 207 cases, 151 cases were diagnosed Acyanotic congenital heart disease [ACHD] and 56 were Cyanotic Congenital Heart Disease [CCHD]. Male (59.9%) outnumbered Female. Total of 143 children [69.08%] were below one year of age at the time of diagnosis. Common clinical features in ACHD were breathlessness (60%), Recurrent chest infections [53%], Failure to thrive [39.2%]. In CCHD blue discoloration was commonest feature [94.6%], breathlessness [58.8%], failure to thrive [25.6%]. Increased respiratory rate was most common physical sign [58.9%] in ACHD and cyanosis [100%] in CCHD. Frequently observed complications were heart failure and growth failure.

Keywords: Congenital Heart Disease; Cyanotic Congenital heart disease; Acyanotic congenital heart disease.

Introduction
Congenital heart disease (CHD) is defined as abnormality in ‘cardiocirculatory’ structure or function that is present since birth, even though it may be discovered later. CHDs remains the leading cause of death in children with malformation. Incidence of CHDs being 8 per 1000 live births and is the most common severe congenital abnormality[11]. With the currently available treatment modalities over 75% of infants born with critical heart disease can survive beyond the first year of life and many can lead a near normal life thereafter.[2]
The clinical presentation of congenital heart disease varies according to the type and severity of the defect.\[^3\]\ In neonatal period the presenting feature of congenital heart disease are cyanosis (with or without respiratory distress), heart failure (with or without cyanosis), collapse, an abnormal clinical sign detected on routine examination (e.g. absent femoral pulse or a heart murmur).\[^3\]\ In infancy and childhood, the usual presenting features are cyanosis, digital clubbing, murmur, syncope, squatting, heart failure, arrhythmia, failure to thrive.\[^4\]\ The adolescent and adults present with heart failure, murmur, arrhythmia, cyanosis, hypertension, late consequences of previous cardiac surgery (e.g. arrhythmia, heart failure).\[^4\]\ This study was undertaken to find out the age of diagnosis, clinical profile and immediate outcome of congenital heart disease among the admitted children in our institution.

Patients and Methods

This prospective study was carried out over a period of one year among the admitted children age ranging from newborn to 12 years. The cases were included in the study when the diagnosis of CHD was established by echocardiography. After enrolment, the detailed history of the studied patients was taken to know their clinical presentation. Moreover, a thorough clinical examination was done to evaluate specific heart lesion. Apart from echocardiography other investigations like chest x-ray, electrocardiography and other relevant investigations were also done.

Results

In this study total of 207 children with CHD between age of newborn to 12 years were admitted during study period. Table 1 depicts the baseline characteristic of children with CHD who were hospitalized during the study period. 151 children had Acyanotic CHD [72.9%], whereas cyanotic CHD was present in 56 [27%]. 124 children were male while as 83 were females with M: F ratio 1.5:1.

Of total children 15.9% presented in the neonatal period, 52.3% in infancy, 25.6% from 1 to 6-year. Least number of children were more than 6 years at diagnosis [5.8%].

| Table 1: Baseline characteristic of children with CHD |
|-----------------------------------------------|
| **Baseline characteristic** | **N [percentage]** |
| **Type of CHD** | |
| Acyanotic | 151[72.9] |
| Cyanotic | 56[27] |
| **Gender** | |
| Male | 124[59.9] |
| Female | 83[40] |
| **Age** | |
| 0-28 days | 33[15.9] |
| 1 month-1 year | 110[52.3] |
| 1-6 year | 52[25.6] |
| 6-14 years | 1[5.8] |

| Table 2: Age of presentation of acyanotic congenital heart diseases |
|---------------------------------------------------------------|
| **Diagnosis** | **0-28 days** | **1 month-1 year** | **1-6 years** | **6-14 years** |
|----------------|----------------|-----------------|-------------|----------------|
| VSD | 4[6] | 46[69.6] | 13[19.6] | 3[4.5] | 66 |
| ASD | 1[4] | 8[32] | 12[48] | 4[16] | 25 |
| PDA | 4[22.2] | 10[55.5] | 3[16.6] | 1[5.5] | 18 |
| AVSD | 2[18.18] | 7[63] | 2[18.18] | | 11 |
| COA | 3[23] | 8[61] | 1[7.6] | 1[7.6] | 13 |
| IAA | 1[50] | 1[50] | | | |
| ALCAPA | 2[50] | 2[50] | | | 4 |
| CCTGA/VSD | 2[25] | 4[50] | 1[12] | 1[12] | 8 |
| AP WINDOW | 1[25] | 2[50] | 1[25] | | 4 |

ASD: Atrial septal defect, VSD: Ventricular septal defect, PDA: Patent ductus arteriosus, AVSD: Atrioventricular septal defect, CCTGA: Congenitally corrected transposition of the great arteries, IAA: Interrupted aortic arch, ALCAPA: Anomalous left coronary artery from the pulmonary artery, AP: aortopulmonary window
Table 2 depicts the age of presentation of various Acyanotic CHD. VSD was commonest Acyanotic CHD. The majority [69.6%] of children with VSD were diagnosed in infancy. Most children with PDA and AVSD were also diagnosed in infancy. Age of presentation ASD was later with the majority presenting more than 1 year of age. More complex Acyanotic CHD like COA, IAA, ALCAPA, AP window presented in neonatal and early infancy.

Table 3: Age of presentation of cyanotic congenital heart diseases

| Diagnosis                  | 0-28 days | 1 month-1 year | 1-6 years | 6-12 years |
|----------------------------|-----------|----------------|-----------|------------|
| TOF                        | 1[4]      | 12[48]         | 10[40]    | 2[8]       |
| DORV/VSD                   | 2[33.3]   | 2[33.3]        | 2[33.3]   | 6          |
| dTGA/VSD                   | 5[71]     | 2[28]          |           | 7          |
| TAPVC                      | 1[50]     | 1[50]          |           | 2          |
| Tricuspid atresia/VSD      | 1[50]     | 1[50]          |           | 2          |
| DILV with PAH/PVS          | 2[50]     | 2[50]          |           | 4          |
| VSD Pulmonary atresia      | 1[33.3]   | 1[33.3]        | 1[33.3]   | 3          |
| Ebstein anomaly            | 1[25]     | 3[75]          |           | 4          |
| Truncus arteriosus         | 1[100]    |                |           | 1          |
| HLHS                       | 2[100]    |                |           | 2          |

Table 3 depicts the age of presentation of various cyanotic CHD. TOF was commonest cyanotic CHD. 12 patients presented in infancy [48%]. TGA presented in neonatal age [71%]. Complex cyanotic CHD DILV, Truncus arteriosus, HLHS mostly presented in early neonatal age.

Table 3a: Symptomatology

| ACYANOTIC CHD | NUMBER OF CASES | PERCENTAGE |
|---------------|-----------------|------------|
| Breathlessness| 91              | 60.2%      |
| Recurrent chest infections | 80 | 53% |
| Failure to thrive | 59 | 39.2% |
| Cough | 54 | 36% |
| Feeding difficulty | 51 | 33.1% |
| Fever | 37 | 24.5% |
| Asymptomatic | 13 | 8.6% |

Table 3b: Important physical findings in CHDs

| ACYANOTIC CHD | N=151 | ACYANOTIC CHD | N=56 |
|---------------|-------|---------------|------|
| Cyanosis      | 56    | 100%          |      |
| Increased respiratory rate | 12 | 23.5% |
| Clubbing      | 24    | 44.6%         |      |
| Clinical signs | Polycythemia | 11 | 19.6% |
| Tachycardia   | 5     | 9.8%          |      |
| Pallor        | 2     | 3.7%          |      |

Table 4: Complication of CHDs

| Complication | Number | Percentage |
|--------------|--------|------------|
| Failure to thrive | 130 | 62.8% |
| Congestive cardiac failure | 111 | 53.6% |
| Recurrent chest infections | 66 | 31.8% |
| Irreversible pulmonary hypertension | 29 | 14% |
| Hypoxic seizures | 13 | 6.2% |
| Brain abscess | 2 | 0.9% |

Table 4 depicts the various complication among children admitted with various CHD. In the present study failure to thrive was commonest found in 62.8% cases followed by congestive cardiac failure occurring in 53.6%, recurrent chest infections occurred in 31.8%. Irreversible pulmonary hypertension occurred in 29 cases [14%].
Discussion

The present study was undertaken to know the age of diagnosis of congenital heart diseases admitted in the Department of Paediatrics Government medical college and hospital, over a period of 1 year. 151 children had Acyanotic CHD [72.9%], whereas cyanotic CHD was present 56 [27%] with M: F ratio 1.5:1. This is comparable to many studies by Shah GS, et al in Nepal where in the male to female ratio was 1.5:1[5]. Similarly in a study conducted by Humayun et al[6] in Pakistan, male to female ratio was 1.7:1. Male preponderance in congenital heart disease was seen in majority of the studies conducted worldwide. This male dominance pattern could be due to Indian social and cultural factors. Neglect, differential treatment, or poor access to health-care facilities is putting girls at disadvantages. Moreover, this could be the reason for the less female child seeking health-care facilities.

Of total children 15.9% presented in the neonatal period. 52.3% in infancy, 25.6% from 1 to 6-year, least number of children were more than 6 years at diagnosis 5.8%. VSD was commonest ACHD and the majority of children were diagnosed between 1 month to 1 year. Complex ACHD like COA, IAA, was diagnosed less than 1 year. TOF was commonest cyanotic CHD and most common age of diagnosis was between 1 month to 1 year. Total of 143 children [69.08%] was below one year of age at the time of diagnosis. The time of diagnosis of CHD in our study was earlier as compared to studies in developing countries[7]. Early age of diagnosis has been reported in many studies Turkey[8], Kenya[9] and an earlier report from Nigeria by Ibadin[10]. The earlier age of diagnosis in our study can be explained as our hospital is the main paediatric referral hospital in Kashmir and we have dedicated paediatric cardiology facilities in our hospital. The earlier age of diagnosis has prognostic significance A higher mean age may translate to a significant number of a patient not having optimal surgical intervention[11].

The modes of presentation of ACHD seen in this study includes breathlessness [60.2%], frequent respiratory tract infections [53%], failure to thrive [39%], cough [36%] feeding difficulty [33.1%] and fever [24.5%]. 8.6% were asymptomatic and were incidentally diagnosed. In CCHD, blue discoloration as noticed by parents [94.6%] breathlessness [58%], failure to thrive [25.4%]. Cyanotic spell occurred in 10.7% and hypoxic seizures in 3.9%. 8.9% were asymptomatic cases were parents missed blue discoloration. Padedum et al[12] in his study identified the most common presentation were chest retractions (57.4%) followed by cough (53.7%), breathlessness (35.1%), failure to thrive (25.9%), feeding difficulty (14.8%). In CCHD, it was observed that cyanosis was the most common presenting complaint in 100% cases followed by feeding difficulty (80%), breathlessness (60%), failure to thrive (60%), chest retractions (40%) and cyanotic spell in 20% cases. Sandeep et al[13] in his study conducted at a tertiary care hospital observed the commonest symptom in CHD as breathlessness (78%) followed by LRTI (60%), FTT (40%), cyanosis (20%) and fever (24%). Similar results were also observed by Dipendra et al[14] were, breathlessness was the most common presenting symptom reported in 69.2% followed by fatigue (62.6%), fever (59.3%), cough (54.9%), failure to thrive (42.8%), recurrent LRTI (35.1%), CCF (27.4%), cyanosis (26.3%), refusal of feed (17.5%), cyanosis (9.8%) and clubbing (15.3%). Shamima Sharmin et al[15] observed similar results. Recurrent LRTI was the most common in ACHD Most common physical finding in ACCHD was increased respiratory rate [58.9%], tachycardia [43.1%], chest indrawing 33.3% and tender hepatomegaly 23.1%. Pallor increased JVP and chest signs occurred in 19.8%, 11.9% and 15.3% respectively. In CCHD cyanosis was present in 100%, clubbing in 44.5%, increased respiratory rate 23.5%; clinical signs of polycythemia like congested eyes were present in 19.6%. Tachycardia in 9.8%; pallor was least commonly
found only in 3.7%. Physical findings in our study are similar to those found by\textsuperscript{[15]}. In the complication of CHD, the commonest was Failure to thrive occurring in 62.8%. The congestive cardiac failure occurred in 53.6% followed by recurrent chest infections in 31.8%. Irreversible pulmonary hypertension was present in 14%. Hypoxic seizures secondary to cyanotic spell occurred in 6.2%. Brain abscess occurred in two patients. Study by Rao et al\textsuperscript{[16]} reported growth failure as commonest complication of CHD. Study by Jain et al\textsuperscript{[17]} found CCF as commonest complication of CHD.

**Conclusion**

Breathlessness, chest retractions, FTT, feeding difficulty, cyanosis were the common clinical presentations in congenital heart disease. CHD should be suspected in all cases of recurrent chest infections and failure to thrive. A high index of suspicion, a detailed history, physical examination, chest x-ray, electrocardiogram along with Echocardiography helps us to diagnose most of the congenital heart disease. With limited resources, clinical acumen forms the backbone for diagnosis for CHD. Early detection and intervention reduces the morbidity and mortality of CHDs.

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