Abstract

Background: Congenital heart disease (CHD) refers to various abnormalities with the structure and functions due to anomalous development of the heart before birth. Usually classified into two types: acyanotic and cyanotic, CHD is the most common type of all birth defects and is responsible for more deaths in the first year of life than any other birth defects.

Objectives: To determine the prevalence and clinical profile in a tertiary medical centre.

Materials and Methods: The study was conducted in the department of Pediatrics, RIMS, Imphal, during a period of two years from August 2011 to July 2013 in which all admitted cases of CHD (0-12 years), were evaluated with detailed history, clinical examinations and routine investigations including chest X-ray (CXR), electrocardiogram (ECG) and confirmed by echocardiography.

Results: 83 children (1.76%) were diagnosed with CHD during the study period of 2 years. Majority of cases (51.8%) were observed in the 1-12 months age group followed by 26.5% in 0-1 month, 18.1% in 1-5 years and 3.6% in 5-12 years age group respectively. Acyanotic congenital heart disease (ACHD) was the commonest type accounting for 86.8% of all CHDs out of ventricular septal defect (VSD) constituted 38.6% of the total CHDs. Among the cyanotic congenital heart disease (CCHD), Tetralogy of Fallot (TOF) was the commonest type constituting 10.8% of all CHDs. The most predominant symptom was breathing difficulty (85.5%) followed by fever (62.7%), cough (60.2%), recurrent chest infection (57.8%), excessive cry (56.6%), reduced feeding (48.2%), increased sweating during feeds (44.6%), failure to thrive (26.5%) and blue lips (13.2%). There were 3(3.6%) mortalities in infants with complex CHDs.

Conclusion: Early diagnosis of CHD will significantly contribute in reducing morbidity and mortality by prioritizing appropriate management.

Keywords: Congenital heart disease, clinical profile, ventricular septal defect, breathlessness.

Introduction

Congenital heart disease (CHD) is the most common type of birth defects and also the leading cause of death in children with malformations. These defects may be diagnosed during fetal life or soon after birth requiring urgent intervention, while some may be totally asymptomatic. Other children who have not been diagnosed early may
present late with shortness of breath, difficulty in feeding, tachycardia, cyanotic episodes (esp. during feeding), murmurs on routine screening, heart failure, failure to thrive, sudden collapse etc.²

The reported incidence of CHD is 8/1000 live births.³ Data available for prevalence of CHD in India varies from as low as 0.8 to 5.2/1000 live births.⁴,⁵ Generally divided into two types - acyanotic and cyanotic, the commonest CHD remains ventricular septal defect, closely followed by atrial septal defect and patent ductus arteriosus. Tetralogy of Fallot, transposition of great arteries is seen in one fifth of neonates having CHD.⁶ Severity with cardiac malformations vary. Many patients have little clinical dysfunction while others require surgery, and for a few the malformation may be incompatible with life. Even though cardiac malformations occur in few live births, the number of years of potential life lost because of cardiac malformations is great because most deaths occur in infancy and early childhood. Rapid advances have taken place in the diagnosis and treatment of CHD over the last six decades. Presently, accurate diagnosis of CHD can be made antenatally and with current available treatment modalities, over 75% of infants born with critical heart disease can survive beyond the first year of life and many can lead near normal lives thereafter. However, the privilege of early diagnosis and appropriate management is restricted to children in developed countries only. Unfortunately, majority of affected children born in developing countries lack access to necessary care leading to high morbidity and mortality.⁵

Timely accurate diagnosis and reliable information regarding CHD will not only help in defining the problem but will also help in prioritizing efforts in their management. Very few studies and surveys have been conducted in the north-eastern states of India, including Manipur, on CHD. This study is undertaken to describe the prevalence and clinical profile of CHD in a tertiary centre in Imphal, Manipur.

Aims and Objectives
To determine the prevalence and clinical profile of CHD in Regional Institute of Medical Sciences, Imphal, Manipur.

Materials and Methods
This was a hospital based cross sectional prospective study carried out in the Department of Pediatrics, Regional Institute of Medical Sciences, Imphal, Manipur from 1st August 2011 to 31st July 2013. Admitted children, aged 0-12 years clinically diagnosed with CHD were included in the study. Relevant details such as age, sex, religion, socioeconomic status, pertinent maternal history, presenting complaints (feeding difficulties, fast breathing, easy fatigability, excessive cry/sweating, respiratory difficulties, cyanotic spells etc.), duration of illness at presentation, nutritional status were obtained and recorded in a predefined proforma along with detailed physical examination findings. Each enrolled patient was subjected to routine investigations including roentgenogram and electrocardiogram. Echocardiograms were performed for definite diagnosis and findings analyzed. A written consent from the parents or legal guardians was taken for each enrolled case. Infants or children whose parents/legal guardians refused consent and those clinically suspected to have acquired heart diseases were excluded from the study.

Results and Observations
A total of 4714 children (aged 0-12 years) were admitted during the study period, out of which 83 children were diagnosed with CHD constituting 1.76% of the total pediatric admissions in the hospital. Maximum number of cases - 43(51.8%) were observed in the 1-12 months age group. 22(26.5%) cases were seen in the 0-1 month age group, followed by 1-5 years with 15(18.1%) and 3(3.6%) in 5-12 years age group (Table – 1). The youngest patient was a 2 days old neonate having VSD with patent ductus arteriosus (PDA) and double outlet right ventricle (DORV). CHD was
found to be distributed equally among males – 42(50.6%) and females – 41(49.4%) respectively (M : F = 1:1).

Breathing difficulty (85.5%) was the most common presenting symptom at admission followed by fever (62.7%), cough (60.2%) and recurrent chest infection (57.8%) respectively. Other symptoms were excessive cry (56.6%), reduced feeding (48.2%), increased sweating during feeds (44.6%), failure to thrive (26.5%) and blue lips (13.2%) [Table – 2]. Among acyanotic CHDs breathing difficulty, fever and cough were more common while blue lips, breathing difficulty, excessive cry and chest retractions were predominant in cyanotic CHDs. One case of tetralogy of Fallot reported with loss of consciousness and cyanosis following bouts of loose stools, 

Acyanotic CHD (ACHD) was the commonest type accounting for 72 (86.8 % of all CHDs. Cyanotic CHD (CCHD) constituted the remaining 11(13.2%). Of all the ACHDs, ventricular septal defect (VSD) was the most common, being found in 32(38.6%) of the total CHDs & 44.4% of the total ACHDs(Table - 3). Tetralogy of Fallot (TOF) was the major CCHD type being observed in 9 cases (81.8% of all CCHDs and 10.8% of all CHDs(Table - 4). 

The number of preterm in the study group were 12(14.5%) and PDA was found in 8(66.7%) of them. The most common non-cardiac congenital anomaly observed in association with CHD was Down syndrome -4(4.8%). The other associated non-cardiac anomalies were congenital talipes equinus varus (CTEV) – 2(2.4%), situs inversus – 2(2.4%), polydactylly -1(1.2%) and Fanconi’s anemia -1(1.2%). 

Oral Ibuprofen treatment resulted in closure of patent ductus arteriosus in 8(66.7%) of the preterm neonates.

A marginally higher number of cases (44;53%) came from rural areas as compared to those from urban regions (39;47%) and the cases were mainly concentrated between upper lower class IV and lower middle class III comprising 42.2% and 34.9% respectively (Table – 5).

3(3.6%) infants with complex CHD anomalies expired during hospitalization. 

**Table 1:** Prevalence of congenital heart disease in different age groups

| Age group | Number of cases | Percentage (%) |
|-----------|-----------------|----------------|
| 0 – 1 month | 22              | 26.5           |
| 1 – 12 months | 43              | 51.8           |
| 1 – 5 years     | 15              | 18.1           |
| >5 – 12 years   | 3               | 3.6            |

**Table 2:** Clinical manifestations of congenital heart disease at admission

| Clinical manifestations | Number of cases | Percentage (%) |
|-------------------------|-----------------|----------------|
| Breathing difficulty    | 71              | 85.5           |
| Fever                   | 52              | 62.7           |
| Cough                   | 50              | 60.2           |
| Recurrent chest infection | 48              | 57.8           |
| Excessive cry           | 47              | 56.6           |
| Reduced feeding         | 40              | 48.2           |
| Increased sweating      | 37              | 44.6           |
| Failure to thrive       | 22              | 26.5           |
| Blue lips               | 11              | 13.2           |
| Loose stools             | 2               | 2.4            |
| Loss of consciousness   | 1               | 1.2            |

**Table 3:** Relative frequency of different sub-types of acyanotic congenital heart disease (ACHD)

| Type of ACHD | No. of cases | % of ACHD | % of total CHD |
|--------------|--------------|-----------|----------------|
| VSD          | 32           | 44.4      | 38.6           |
| ASD          | 16           | 22.2      | 19.3           |
| PDA          | 13           | 18.1      | 15.7           |
| AVSD         | 5            | 6.9       | 6.0            |
| RVOTO        | 4            | 5.6       | 4.8            |
| LVOTO        | 2            | 2.8       | 2.4            |
| Total        | 72           | 100       | 86.8           |

CHD – congenital heart disease; ACHD- acyanotic congenital heart disease; VSD-ventricular septal defect; ASD-atrial septal defect; PDA – patent ductus arteriosus; AVSD – atrioventricular septal defect; RVOTO-right ventricular outflow tract obstruction; LVOTO-left ventricular outflow tract obstruction

**Table 4:** Relative frequency of sub-types of cyanotic congenital heart disease (CCHD)

| Type of CCHD | No. of cases | % of CCHD | % of total CHD |
|--------------|--------------|-----------|----------------|
| TOF          | 9            | 81.8      | 10.8           |
| TGA          | 1            | 9.1       | 1.2            |
| EA           | 1            | 9.1       | 1.2            |
| Total        | 11           | 100       | 13.2           |

CCHD-cyanotic congenital heart disease; TOF-tetralogy of Fallot; TGA-transposition of great arteries; EA- Ebstein’s anomaly
Discussion

In the present study, clinically diagnosed 83 children with CHD were enrolled out of a total of 4714 pediatric admissions (0-12 yrs age) giving a prevalence of 1.76%. This may not be representative of the true prevalence as the study is of limited two years duration. However in India the prevalence ranged between 0.8 to 5.2/1000 live births.4,5 Maximum number of cases of CHD was seen in the age group 1-12 months (51.8%) followed by 0-1 month group (26.5%). Kula S et al7 reported 39.3% of CHDs during the first year of life. Ibadin MO et al8 also had similar findings of 46.9% in infants less than one year, 4.1% in neonatal period and the rest in 1 year and above.

The male to female ratio was almost 1:1 which was comparable to the findings of other workers.9,10 Mughal AR et al11 however reported higher preponderance of males (59.7%) over females (40.3%) with a male:female ratio of 1.5:1.

In our study, the common presenting symptoms were breathing difficulty (85.5%), fever (62.7%), cough (60.2%), recurrent chest infection (57.8%), excessive cry (56.6%), reduced feeding (48.2%), increased sweating during feeds (44.6%), failure to thrive (FTT) and blue lips (13.3%). This finding is similar to the one reported by Ibadin MO et al8 with breathing difficulty as the commonest presenting symptom (75.5%) followed by cough (57.1%), fever (38.8%), blue lips (12.2%). Sharmin LS et al12 too reported breathlessness as the major presenting symptom (60%), followed by fatigue (54.8%), cough (43.5%) and blue lips (20%). Tank et al13 also found breathlessness to be present in 74.83% of all CHD’s followed by symptoms of lower respiratory tract infection (LRTI - 44.89%), cyanosis (37.4%), FTT (38.77%), congestive cardiac failure (CCF - 30.6%), refusal of feeds (30.6%), cyanotic spell in 12.92%. Suraj Gupte et al14 in a study at Jammu and Kashmir in 60 subjects reported presenting complaints of CHD to be 45.8% (breathlessness), 36.66% (failure to thrive), 35.77% (recurrent respiratory tract infection) and 21.66% (cyanosis).

Acyanotic CHD (ACHD) accounted for 86.8% of all CHD’s which was similar to the findings of other workers.15,16,17 VSD contributed 38.6% to the total cases of CHD which was followed by atrial septal defect (ASD-19.3%). VSD as a percentage of total CHD in this study was found to be almost similar as compared to those observed by Alabdulgader AAA9 of 39.5% and Smitha R et al18 of 40.47%. The relatively high prevalence of PDA may be due to the fact that preterm babies were included in the study which contributed to 14.5% of the total CHD’s and routine echocardiogram screening of newborn including preterm infants with persistent respiratory distress. Among the cyanotic CHDs, tetralogy of Fallot was the commonest lesion being observed in 9(10.8%) of the cases followed by 1(1.2%) each of TGA and EA, which was also reported by others.15,16,17

In two of the 4 patients with Down syndrome, two had VSD followed by tetralogy of Fallot and atrioventricular septal defect (1 each). Tandon et al19 had reported atrioventricular septal defect as the major lesions patients with Down syndrome, as the commonest lesion.

Oral Ibuprofen resulted in closure of PDA in 75% of the preterm neonates as it had been shown that oral Ibuprofen is effective as Indomethacin in closing a PDA.20

Number of CHD cases from rural areas (53%) in our study were slightly higher as compared to those from urban regions (47%). This contrasts with the study of Kumari NR et al21 who reported higher concentration in the urban population (75%). Miranda JJ et al22 also found higher

Table 5: Distribution of socioeconomic status with congenital heart disease

| Socioeconomic status | No. of cases | Percentage (%) |
|----------------------|-------------|----------------|
| Upper (I)            | 0           | 0              |
| Upper middle (II)    | 7           | 8.4            |
| Lower middle (III)   | 29          | 34.9           |
| Upper lower (IV)     | 35          | 42.2           |
| Lower (V)            | 12          | 14.5           |
| Total                | 83          | 100.0          |
prevalence of CHD among urban (33%) and rural-to-urban migrants (20%) than the rural population (3). The higher percentage of rural population in our study may be due to preference of urbanites in seeking treatment in private health facilities due to their better economic status.

The socio-economic status of the families of the affected patients offers an overview of their ability in the management of the cases. Employing Kuppuswamy’s scale\(^\text{23}\) as an index of socioeconomic status of the enrolled patients’ families, cases were seen most commonly in the upper lower class IV (42.2%) followed by lower middle III (34.9%). 14.5% cases were seen in lower class V while 8.1% were observed in upper middle II class. Ibadin MO et al\(^8\) too reported similar finding of higher prevalence in middle and lower socioeconomic class (85%) as compared to high class (15%). Mughal AR et al\(^11\) also found 93.4% in middle and lower class and observed that the adverse effect on families after definitive therapy of the patient (surgery or angiographic intervention) ranged from leave without pay to losing jobs or business (46%), and selling their assets (11.3%). It also affected the schooling and health of siblings (22.7% and 26.1% respectively). Although medical treatment of the patients in our study entailed some degree of financial expenditure, the impact of socioeconomic status on treatment could not be assessed as no definitive surgical treatment facility is available in our hospital.

In this study, 3(3.6%) infants with complex CHDs expired during hospitalization. One case was diagnosed with PDA and severe pulmonary arterial hypertension; second case with ASD with double outlet right ventricle and the last infant died secondary to complex CHD with atrioventricular septal defect with ostium-primum ASD with mild right ventricular outflow tract obstruction.

Conclusion
Congenital heart disease in infants could be benign or very significant and a high level of suspicion along with meticulous evaluation will immensely contribute in establishing an accurate diagnosis. In the developing countries, the burden of heart disease constitutes a big public health problem and as facilities for treatment are expensive or not available, many children die before getting optimal treatment. Early recognition of CHD will help to treat the child and if the lesions are amenable to corrective surgery, some of these identified children may have a better future.

Contributors
Dr NG conceptualized the study, collected the data and wrote the manuscript. Dr. CS, Dr. HBB and Dr MM did the analysis and reviewed the manuscript. Dr. SD and CM approved the final manuscript. Dr NG and Dr CS will be the guarantors of the manuscript.

Conflict of Interest: None
Source of Funding: None

References
1. Bernstein D. Congenital heart disease. Kleigman RM, Bonita, Staton, Geme JS, Schor N, Behrman RE editors. Nelson Textbook of Pediatrics, 19th Edition, International: Elsevier, 2011, 1549.
2. Moller JH, Taubert KA, Allen HD. Cardiovascular health and disease in children: Current status. A special writing group from task force on children and youth, American Heart Association. Circulation 1994;89(2):923-30.
3. Frommelt MA, Frommelt PC. Advances in echocardiographic diagnostic modalities for the pediatrician. Pediatric Clinics of North America, Philadelphia: Saunders 1999; 46(2):427-39.
4. Gupta I, Gupta ML, Parihar A, Gupta CD. Epidemiology of rheumatic and congenital heart disease in school children. J Indian Medical Assoc 1992; 90: 57-59.
5. Vashishtha VM, Kalra A, Kalra K, Jain VK. Prevalence of congenital heart disease in school children. Indian Pediatr 1993;30:1337-1340.
6. Saxena A. Congenital heart disease in India: A status report. Indian J Pediatr 2005;72(7):595-8.
7. Kula S, Cevik A, Olgunturk FR, Tunaoglu FS, Oguz AD, Ilhan MN. Distribution of congenital heart disease in Turkey. Turk J Med Sci 2011;41(5):889-93.
8. Ibadin MO, Sadoh WE, Osarogiagbon W. Congenital heart diseases at the university of teaching hospital. Niger J Pediatr 2005;32(2):29-32.
9. Alabdulgader AAA. Congenital heart disease in 740 subjects: epidemiological aspects. Annals of Tropical Paediatrics: International Child Health 2001;21(2):111-8.
10. Marelli AJ, Mackie AS, Ionescu-Ittu R, rahme E, Pilote L. Congenital heart disease in the general population. Changing prevalence and age distribution. Circulation 2007;115:163-72.
11. Mughal AR, Sadiq M, Hyder SN, Qureshi AU, Shah SSA, Khan MA et al. Socioeconomic status and impact of treatment on families of children with congenital heart disease. J Coll Physicians Surg Pak 2011;21(7):398-402.
12. Sharmin LS, Haque MA, Bari MA, Ali MA. Pattern and clinical profile of congenital heart disease in a teaching hospital. J Teach Assoc 2008;21(2):58-62.
13. Tank S, Malik S, Joshi S. Epidemiology of CHD among hospitalized patients. Bombay Heart Journal. 2004; 46(2):144-150.
14. Gupte S, Saini G. Congenital heart disease: Clinico echocardiographic profile in children. Asian Journal of Pediatric Practice. 2004; 8(2):30-34.
15. Jain KK, Sagar A, Beri S. Heart disease in children. Indian J Pediatr 1971;38:441-48.
16. Kasturi L, Kulkarni AV, Amin A, Mahashankar VA. Congenital heart disease: clinical spectrum. Indian Pediatr 1999;36:953.
17. Shah GS, Singh MK, Pandey TR, Kalakheti BK, Bhandari GP. Incidence of congenital heart disease in tertiary care hospital. Kathmandu Univ Med J. 2008 Jan-Mar;6(1):33-6.
18. Smitha R, Karat SC, Narayanappa D, Krishnamurthy B, Prasanth SN, Ramchandra NB. Prevalence of congenital heart disease in Mysore. Indian J Hum Genet 2006;12(1):11-16.
19. Tandon R, Edwards JE. Cardiac Malformations Associated with Down's Syndrome Circulation, Volume XLVII, June 1973; 1349-55.
20. Ohlsson A, Walia R, Shah SS. Ibuprofen for the treatment of patent ductus arteriosus in preterm and/or low birth weight infants. Cochrane Database Syst Revs. 2010;4:CD003481.
21. Kumari NR, Raju IB, Patnaik AN, Barik R, Singh A, Pushpanjali A, et al. J Cardiovasc Dis Res 2013;4(1):11-4.
22. Miranda JJ, Gilman RH, Smeeth L. Differences in cardiovascular risk factors in rural, urban and rural-to-urban migrants in Peru. Heart 2011;97(10):787-96.
23. Kumar N,Sekhar C, kumar P, Kundu AS. Kuppuswamy's socioeconomic status scale-updating for 2007. Indian j Pedatr 2007;74:131-2.