BRIEF REPORT

PATTERN OF HEART DISEASES AMONG PATIENTS REFERRED TO A PAEDIATRIC CARDIOLOGY CLINIC IN SRI LANKA, THE SITUATION FROM 2010 TO 2012

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Abstract

Background:

Epidemiology of cardiac diseases among paediatric population remain largely undocumented in Sri Lanka. The objective of this study was to identify the pattern of heart diseases among patients referred to the Paediatric cardiology clinic at Teaching Hospital Kandy and Sirimavo Bandaranaike Specialized Childrens’ Hospital (SBSCH) Peradeniya.

Methods: Demographic, clinical details and echocardiography details of patients referred for cardiac assessment between 1st 2010 to 30th November, 2012 were descriptively analysed.

Results: Ten thousand, six hundred and seventy-eight (10678) newly referred patients were evaluated over the study period. Of the study cohort 6582 patients had some form of heart disease. Among them, 5363 (50.2%) had congenital heart diseases (CHDs). The most frequent acyanotic heart disease was Atrial Septal Defect (ASD-3024 patients). Ventricular Septal Defect (VSD) ranked second in frequency (697 patients). The commonest cyanotic CHD was Tetralogy of Fallot (79 patients). There were 1200 patients with acquired heart diseases. The most frequent preventable acquired heart disease was Rheumatic valvular heart disease (80 patients).

Conclusions: The commonest CHD among the study cohort was ASD while Tetralogy of Fallot was the commonest cyanotic heart disease. Early identification is important to ensure appropriate interventions and prevent complications.

Key words: Congenital Heart Disease; CHD

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Introduction

Congenital Heart Disease (CHD) is the most common congenital abnormality in newborns\(^1\). It accounts for 25\% of all congenital anomalies\(^2\). The reported prevalence of CHDs varies between 3 to 12 per 1000 live births in Western countries\(^3\)-\(^6\). According to World Health Organization reports, among all cardiovascular diseases, the incidence of congenital heart disease is 10\% in Sri Lanka, 6\% in Bangladesh, 15\% in India and 6\% in Burma\(^7\). But these are not based on data sources from individual countries but were based on US, UK, Canadian or Australian prevalence or incidence statistics, which were, extrapolated with statistical modelling\(^8\).

Heart diseases seen in children are mainly congenital and some like Rheumatic carditis are acquired. Etiology of CHD is multifactorial and a large collection of environmental and genetic causes have a role in their pathogenesis\(^9\). CHDs contribute appreciably to childhood mortality and morbidity worldwide but more in developing countries where facilities for their modern management are often lacking\(^10\). They could be life threatening in early childhood\(^11\)-\(^13\). They not only contribute to significant mortality and morbidity but also causes a tremendous psychological stress and economic burden to the whole family and to the society. Early recognition and appropriate interventions may be life saving in some patients while in others; they could mitigate the morbidity burden\(^14\). Previous studies on the CHDs have shown that the most frequent lesions are VSD, ASD, and PDA\(^11\)-\(^12\).

Malformations of the cardiovascular system are also associated with significant medical morbidity, which requires use of costly medical facilities\(^15\). Thus determining the prevalence and pattern of CHD is necessary to recommend changes in health policies\(^16\). Over the last two to three decades, major advances have been made in the diagnosis and management of CHD\(^17\). Paediatric cardiology services in Sri Lanka are still in its infancy. Over the last decade, there has been a rapid development of services like advanced echocardiography, cardiac catheterization and availability of sufficiently trained paediatric cardiologists and cardiothoracic surgeons in the country. This means that children now have access to early diagnosis and definitive treatment for heart diseases. As a result, more children with CHD are surviving into adolescence and adulthood.

There are no data on prevalence and pattern of CHDs or mortality due to them in Sri Lanka. Thus, there is a need for identification of pattern of heart diseases in children to recommend changes in health policies. This study was done with the objective of identifying the pattern of heart diseases among patients referred to paediatric cardiology clinics at Teaching Hospital Kandy and Sirimavo Bandaranaike Specialised Children’s Hospital (SBSCH), Peradeniya. We also wanted to identify the socio-demographic characteristics of patients with CHDs and age at presentation. Furthermore, we also wanted to study the geographical distribution of patients with rheumatic heart diseases.

Material and Method

This descriptive cross sectional study was conducted at Teaching Hospital Kandy and SBSCH Peradeniya in Sri Lanka. The study period of 2 years extended from 1\(^{st}\) December 2010 to 30\(^{th}\) November 2012. During the study period this was the only dedicated paediatric cardiology clinic which served the central Sri Lanka and its surroundings.
All children who attended the paediatric cardiology clinic for the first time were included and patients attending for reviews were excluded.

All the patients referred with suspected cardiac problems were evaluated by a Specialist Paediatric Cardiologist and had echocardiograms done on them. Ten thousand six hundred seventy eight (10678) patients were studied. Age ranged from 1 day to 64 years, including adults with congenital heart diseases who attended the clinic.

Patients with heart diseases due to congenital or acquired causes with any structural or functional derangement were included in the analysis. All the other patients referred for cardiac evaluation i.e. due to syncopal attacks, chest pain, suspected endocarditis, Kawasaki disease who had structurally and functionally normal hearts were considered as normal.

Results

During the study period of 2 years, total of 10678 patients were evaluated. There were 5994 males and 4684 females, and male: female ratio was 1.2: 1. According to ethnicity, 8679(81.2%) of them were Sinhalese, 1102 (10.3%) Muslims and 897 (8.4%) Tamils.

Of the patients considered for the study, 6582 (61.6%) patients were diagnosed to have some form of heart disease. Among them 5363 had congenital heart disease, 1200 had an acquired heart disease and 17 had arrhythmia. Pattern of heart diseases are shown in Table 01. Among CHDs, 5102 (95.1%) were acyanotic and 261(4.9%) had cyanotic CHDs.

The commonest acyanotic heart disease was Atrial Septal Defect (ASD). The frequencies of specific forms of CHD are shown in the table 02.

Atrial Septal defect was the most frequent form of CHD which was found in 3024 patients. Out of the ASDs 2974 (98.3%) patients had ostium secundum Atrial sepal defect, 29 (0.95%) had Sinus Venous ASD and 21 (0.69%) had Ostium Primum ASDs. Atrial Septal Defects were commonly associated with Patent ductus arteriosus (352), ventricular Septal Defects (281) or pulmonary stenosis (212). Patent Foramen Ovale was also found in 983 patients (18.32%) which usually does not need any intervention.

Occurrence of Atrial septal defects were followed by ventricular septal defects (697), Patent ductus arteriosus (589) and pulmonary stenosis (304).

Among the cyanotic congenital heart diseases, Tetralogy of Fallot 79 (1.4 %) was the most common and it was followed by Complete Atrio ventricular Septal defects 37(0.6%) and d-transposition of great arteries 33(0.6%) as shown in the table 03.

| Pattern of heart diseases   | Number (%) |
|-----------------------------|------------|
| Acyanotic CHD               | 5102 (47.8%) |
| Cyanotic CHD                | 261 (2.4%)  |
| Acquired heart disease      | 1200 (11.2%) |
| Arrhythmia                  | 17 (0.3%)   |
| Normal heart                | 4098 (38.3%) |
| **Total**                   | **10678 (100%)** |
Table 02: Specific form of common CHDs

| Specific form of common CHDs | Frequency (% of any heart diseases - 6580) |
|-----------------------------|------------------------------------------|
| Acyanotic CHDs              |                                          |
| Atrial Septal Defect (ASD)  | 3024 (46)                                |
| Ventricular Septal Defect (VSD) | 697 (11)                             |
| Patent Ductus Arteriosus (PDA) | 589 (9)                                 |
| Valvular Pulmonary Stenosis (PS) | 304 (4.6)                             |
| Branch Pulmonary Stenosis   | 61 (1)                                   |
| Coarctation of Aorta COA    | 23 (0.35)                                |
| Other lesions (e.g.- ALCAPA, AP window etc.) | 410 (6.2) |

The total is different as the common disorders were considered ignoring the combined lesions.

Table 03: Specific form of common CHDs in detail

| Congenital heart disease              | Frequency (Percentage %) |
|---------------------------------------|--------------------------|
| Acyanotic CHDs                        |                          |
| Atrial Septal Defect (ASD) only       | 2130 (39.72)             |
| Ventricular Septal Defect (VSD) only  | 350 (6.52)               |
| Patent Ductus Arteriosus (PDA) only   | 158 (2.94)               |
| Valvular Pulmonary Stenosis (PS) only | 56 (1.04)                |
| Branch Pulmonary Stenosis             | 61 (1.13)                |
| ASD+VSD                               | 281 (5.23)               |
| ASD+PDA                               | 352 (6.59)               |
| VSD+PDA                               | 12 (0.22)                |
| ASD+VSD+PDA                           | 38 (0.70)                |
| ASD+PS                                | 212 (3.95)               |
| PS+VSD                                | 07 (0.13)                |
| PS+PDA                                | 08 (0.14)                |
| PS+ASD+PDA                            | 12 (0.22)                |
| PS+VSD+PDA                            | 07 (0.13)                |
| PS+ASD+VSD+PDA                        | 02 (0.03)                |
| Coarctation of Aorta COA              | 23 (0.42)                |
| Other acyanotic CHDs                  | 410 (7.64)               |
| Patent Foramen Ovale (PFO) only       | 983 (18.32)              |
| Total acyanotic CHDs                  | 5102                     |

Cyanotic CHDs

| Cyanotic CHDs                        | Frequency (Percentage %) |
|--------------------------------------|--------------------------|
| Tetralogy of Fallot (TOF)            | 79 (1.47)                |
| Complete AV canal defect             | 37 (0.68)                |
| d-Transposition of great arteries    | 33 (0.61)                |
| Total anomalous pulmonary venous drainage | 27 (0.5)               |
| Single ventricle                     | 22 (0.41)                |
| Double outlet right ventricle        | 20 (0.4)                 |
| Hypoplastic Left Heart Syndrome      | 10 (0.2)                 |
| Ebstein Anomaly                      | 09 (0.2)                 |
| Other cyanotic CHDs                  | 24 (0.45)                |
| Total Cyanotic CHDs                  | 261                      |

Total 5363
Out of all the referred patients, 1351 (12.65%) were neonates. Among them 1226 (90.7%) had acyanotic congenital heart disease, 97 (7%) had cyanotic heart disease and only 28 (2.07%) had a normal heart. Age distribution at the time of presentation to the Paediatric cardiology clinic (which was started in 2010) is shown in table 4. Nearly thirty seven percent of the cyanotic heart diseases presented during neonatal period while most of the acyanotic heart disease patients were also less than one year old.

|                | Neonate | 1-12months | 1-5yrs | 6-12yrs | >12yrs |
|----------------|---------|------------|--------|---------|--------|
| Acyanotic      | 1226 (24.3%) | 1676 (32.8%) | 1317 (25.8%) | 710 (13.9%) | 173 (3.4%) |
| Cyanotic       | 97 (37.2%) | 67 (25.7%) | 37 (14.2%) | 32 (12.3%) | 28 (10.7%) |

Table 4: Age of diagnosis of CHDs

Among the acquired heart diseases, 80 patients were found to have rheumatic valvular heart disease which was the commonest preventable acquired heart disease. Rheumatic fever (with or without cardiac involvement) was diagnosed using Modified Jones Criteria. Geographical distribution of rheumatic fever is described in table 5.

Table 5: Geographical Distribution of Rheumatic Fever

| District       | N (%)  |
|----------------|--------|
| Nuwara Eliya   | 36 (44.9%) |
| Kandy          | 28 (35.9%) |
| Matale         | 6 (7.7%)  |
| Kegalle        | 4 (3.8%)  |
| Badulla        | 3 (3.8%)  |
| Ampara         | 3 (3.8%)  |
| **Total**      | **80** |
international studies. It could be due to diagnosis of cyanotic CHDs in other cardiac centers before establishing this new clinic out of Colombo or transferring them to other centers with better facilities as only a clinic is available in our unit or death of patients before accessing available medical facilities. It could also be due to inclusion of minor cardiac defects like Patent Foramen Ovale and mild branch pulmonary artery stenosis as acyanotic CHDs which cannot be considered as normal too.

The most frequent type of CHD was found to be ASD which in accordance with two studies in Iran \(^1\)\(^,\)\(^1\)\(^8\), and one study in Bangladesh \(^1\)\(^9\) while in other studies the most frequent type of CHD was VSD. VSD ranked the second in frequency accounting for 697 patients. Patent ductus arteriosus was seen in 589. These differences could be due to detection of severe form of congenital heart diseases at other cardiac clinics before establishing this new unit, spontaneous or surgical closure of VSD or death before accessing the medical facilities. This might also be due to racial and genetic factors in different populations too.

Out of all the referred patients, 1351 (12.65%) were neonates. Among them 90.96% had acyanotic congenital heart disease, 6.95% had cyanotic heart disease and only 2.07% had normal heart. This shows that after establishing the new unit, there were more positive patients among neonates with higher percentage of babies with cyanotic CHDs.

The multifactorial etiology of CHD involves the chromosomal abnormalities, maternal diabetes, maternal infections during early pregnancy, smoking - and exposure to teratogenic drugs. These can lead to differences in incidence in different countries.

As there is multifactorial inheritance, genetic counseling is important to patients with family history of CHDs. Fetal echocardiography is also helpful to identify and treat the CHDs early which is available in the clinic.

In our study, the most frequent preventable acquired heart disease was rheumatic valvular heart disease accounting for 1.2% of patients. Majority of them (80%) were from Nuwara Eliya and Kandy districts. There were no similar studies to compare this result to identify the trend.

As this is a preventable heart disease, preventive measures can be implemented at local and/ or national level to reduce or prevent it. More attention should be given to the most prevalent areas and further studies are necessary to identify the reasons for that. We recommend further statistical studies in all hospitals in Sri Lanka.

Echocardiography is noninvasive and considered the gold standard for diagnosis of heart diseases. Early referral of children with suspected heart diseases to the nearest paediatric cardiac center for early treatment is essential to improve the outcome.

A limitation of this study is that the data was obtained from the service records which were not specifically designed for this research. As this was a newly established unit, some patients were referred for the review. Therefore, age of presentation to our clinic is not the age of diagnosis in some patients.
Conclusions

The study shows the pattern of the CHDs presenting to a newly established Paediatric Cardiology Clinic in Central Sri Lanka from 2010 to 2012. Majority of patients with congenital heart diseases detected to have acyanotic CHDs. ASD was the commonest acyanotic lesion while TOF was the commonest cyanotic lesion. Rhematic fever was the most frequent preventable acquired heart disease. Early detection of heart diseases is essential to manage them effectively to reduce morbidity and mortality. These finding will help to establish health policies for the improvement of diagnostic, therapeutic and preventive facilities.

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