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Original Article

# Etiological Profile of Congestive Cardiac Failure in Children in a Tertiary Care Center in Tamil Nadu

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## Abstract

**Background:** The availability of adequate, accessible health-care delivery systems, modern diagnostic, and therapeutic modalities has paved the way for prompt diagnosis of congenital heart defects and the institution of appropriate intervention before complications set in, thus reducing the high levels of morbidity and mortality due to these defects in developed countries. In developing countries where there is a scarcity of similarly advanced health-care delivery set up, that goal has not been attained as in case of developed countries. Hence, a regular update in the knowledge of the prevalence and causes of heart failure in children will remain a necessity for their appropriate treatment.

**Objectives:** The objectives of the study are to analyze the incidence of cardiac failure among children and its etiological profile in a tertiary care center in Tamil Nadu, India.

**Materials and Methods:** Children between 1 month and 12 years of age admitted with a clinical diagnosis of cardiac failure over a period of 1 year.

**Results:** Among 7095 cases admitted during the study period, 2.09% (*n* = 148) of children had cardiac failure. The majority of the cases were males constituting 57.4% (*n* = 85) of cardiac failure cases. Among the cardiac etiologies for heart failure, congenital heart diseases (CHDs) tops the list (59.85%). The most common CHD causing heart failure is ventricular septal defect (*n* = 29). Anemia is the most common noncardiac cause of heart failure (*n* = 8).

**Key words:** Anemia, Cardiac failure, Children, Congenital heart diseases, Tertiary care center, Ventricular septal defect

## INTRODUCTION

Why is heart failure in children important? If we just consider the number of individuals affected, adult heart failure is clearly a more compelling public health problem. However, the relatively small numbers belie the overall economic and social impact of pediatric heart failure. When a child is admitted to the hospital for heart failure; the costs are considerably higher for children than adults because of the frequent need for surgical or catheter-based intervention. The demands of medical care can fray the family structure and adversely affect parental economic productivity. When a child dies of heart failure, the economic impact is magnified enormously because of the number of potentially productive years lost per death. For these and other reasons, heart failure in children is a serious public health concern. Cardiac failure is the inability of the heart to deliver the adequate cardiac output to meet the metabolic demands of the body. It is a common problem encountered in emergency pediatric practice. The majority of the children with congenital heart defects in the developing countries have been noted to present in advanced cardiac failure with the result of increased morbidity and mortality. It is also known that failure to carefully seek for the evidence of cardiac failure in children admitted with anemia, bronchopneumonia and other illnesses known to predispose to heart failure, has led to failure to recognize, diagnose and promptly treat heart failure in such children. The result of this is increased mortality from many of these treatable conditions.

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### Definition[1]

Congestive heart failure (CHF) is a clinical syndrome in which the heart is unable to pump enough blood to the body to meet its needs, to dispose of systemic or pulmonary venous return adequately, or a combination of the two.

Andrews *et al.* report the incidence of heart failure assessed at first presentation to hospital to be around 0.87 per 100,000.[2] Kay *et al.* report that the incidence of heart failure as a result of congenital defects is between 1 and 2 per 1000 live births.[3]

The heart may fail if it is confronted with:

1. An excessive preload - Left to right shunts, mitral regurgitation (MR) and complete heart block
2. High afterload - Hypertension, aortic stenosis
3. Impaired myocardial contractility - Myopathy, myocarditis
4. Inadequate diastolic filling – Constrictive pericarditis and tachyarrhythmias.

Under these conditions, initially, various compensatory mechanisms come into play which has got salutary effects but the same compensatory mechanisms if pressed into play indefinitely cause nonsalutary effects and potentiates heart failure.

The well-established NYHA Heart Failure Classification[4] is not applicable to most of the pediatric population.

The Ross Heart Failure Classification[5] was developed to provide a global assessment of heart failure severity in infants and has subsequently been modified to apply to all pediatric ages. The Modified Ross Classification incorporates feeding difficulties, growth problems and symptoms of exercise intolerance into a numeric scale.

### Modified Ross Heart Failure Classification for Children

- Class I – Asymptomatic
- Class II:
  - Mild techypnea (or) diaphoresis with feeding in infants
  - Dyspnea on exertion in older children
- Class III – Marked dyspnea on exertion prolonged feeding times and growth failure
- Class IV – Symptoms such as tachypnea, retraction, grunting, or diaphoresis at rest.

The heart failure syndrome may arise from diverse causes. The causes of heart failure vary with age. Common causes of CHF are volume or pressure overload, or both, caused by congenital or acquired heart disease and myocardial diseases. Tachyarrhythmia and heart block can also cause heart failure at any age. By far the most common causes of CHF in infancy are congenital heart diseases (CHDs). Beyond infancy, myocardial dysfunctions of various etiologies are important causes of CHF. Among the rare causes of CHF are metabolic and endocrine disorders, anemia, pulmonary diseases, collagen vascular diseases, systemic or pulmonary hypertension, neuromuscular disorders, and drugs.

Symptoms of cardiac failure are poor weight gain, difficulty in feeding, fast breathing, persistent cough and wheezing, irritability, restlessness, and pedal edema.

### Aim of the Study

The objective of this study is to find the incidence of congestive cardiac failure (CCF) among children admitted in a tertiary care center and analyzing the etiological profile of cardiac failure.

## MATERIALS AND METHODS

### Study Center

The study was conducted in the pediatric ward of a tertiary care center in Tamil Nadu.

### Sampling

This is a prospective observational study conducted over a period of 1 year. In children between 1 month and 12 years of age admitted with a clinical diagnosis of CHF, detailed history, general, and systemic examination were done. X-ray chest, electrocardiogram (ECG), and echocardiogram (ECHO) were taken. And investigation to ascertain the other etiologies such as complete blood count, peripheral smear, hemoglobin (Hb) electrophoresis, bone marrow aspiration, and renal function tests were done. Enzyme assays were performed in suspected metabolic causes causing cardiac failure. High-resolution computed tomography (CT) was done for cases of bronchiectasis and 64 slice angiography was done for a case of pulmonary hypertension.

Data collected were recorded in a master chart. Data analysis was performed with the help of computer using SPSS software. Data were analyzed using simple descriptive statistics.

## RESULTS

A total number of children admitted in the ward during our study period were 7095. Among them, 148 were admitted with cardiac failure. It constituted 2.09% of the total admissions (Table 1 and Figure 1).

Males admitted with CCF were 85 and females 63 making 57.4% and 42.6%, respectively. The male to female ratio is 1.3:1 (Table 2 and Figure 2).

In our study, the majority of the children admitted with cardiac failure were below 1 year of age (52.70%, $n = 78$). 13.51% ($n = 20$) were between 1 and 3 years of age, 10.14% ($n = 15$) between 4 and 6 years of age, 10.14% ($n = 15$) between 7 and 9 years of age, 13.51% ($n = 20$) were 10 to 12 years old (Table 3 and Figure 3).

Around 92.57% ($n = 137$) of the cardiac failure cases were due to cardiac etiologies whereas non cardiac causes constituted the remaining 7.43% ($n = 11$) (Table 4 and Figure 4).

CHDs contributed 59.85% ($n = 82$) of the heart failure cases due to cardiac etiology. The next common cardiac causes were rheumatic heart diseases and dilated cardiomyopathy making 15.33% ($n = 21$) and 7.30% ($n = 10$), respectively. The proportion of cases due to myocarditis, supraventricular tachycardia, infective endocarditis, pulmonary hypertension, scorpion sting, Pompe's disease, mucopolysaccharidosis (MPS), and undiagnosed causes were 5.11%, 1.46%, 2.19%, 2.19%, 2.92%, 0.73%, 1.46%, and 1.46%, respectively (Table 5 and Figure 5).

Among the various congenital cardiac diseases causing cardiac failure, ventricular septal defect (VSD) is the most

**Table 1: Prevalence of cardiac failure among admissions**

| Gender | Total number of admissions during study period | Number of cases admitted with cardiac failure (%) |
|---|---|---|
| Male | 4275 | 85 (1.20) |
| Female | 2820 | 63 (0.89) |
| Total | 7095 | 148 (2.09) |

**Table 2: Gender distribution of the disease**

| Gender | Frequency (%) |
|---|---|
| Male | 85 (57.4) |
| Female | 63 (42.6) |
| Total | 148 (100) |

**Table 3: Age distribution of cardiac failure cases**

| Age | Frequency (%) |
|---|---|
| 1 month – 1 year | 78 (52.70) |
| 1-3 years | 20 (13.51) |
| 4-6 years | 15 (10.14) |
| 7-9 years | 15 (10.14) |
| 10-12 years | 20 (13.51) |
| Total | 148 (100) |

**Table 4: Etiology of cardiac failure cases**

| ??? | Frequency (%) |
|---|---|
| Cardiac | 137 (92.57) |
| Non cardiac | 11 (7.43) |
| Total | 148 (100) |

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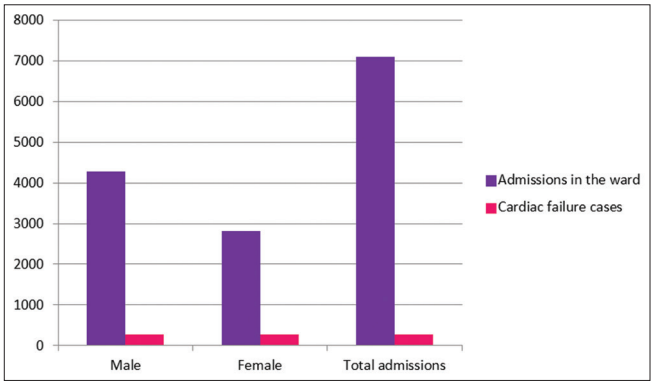


**Figure 1: Prevalence of cardiac failure among admissions**

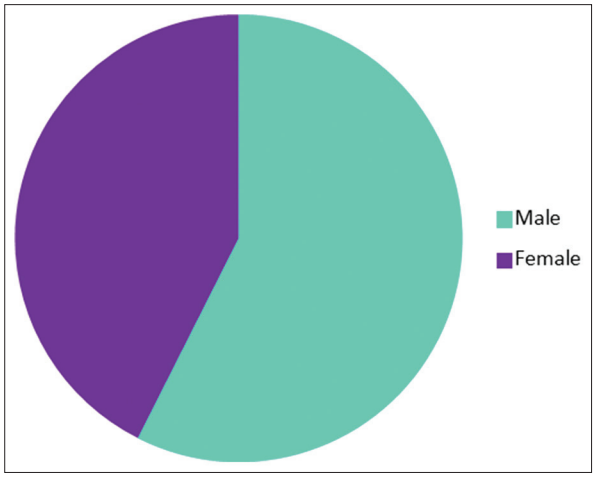


**Figure 2: Gender distribution of the disease**

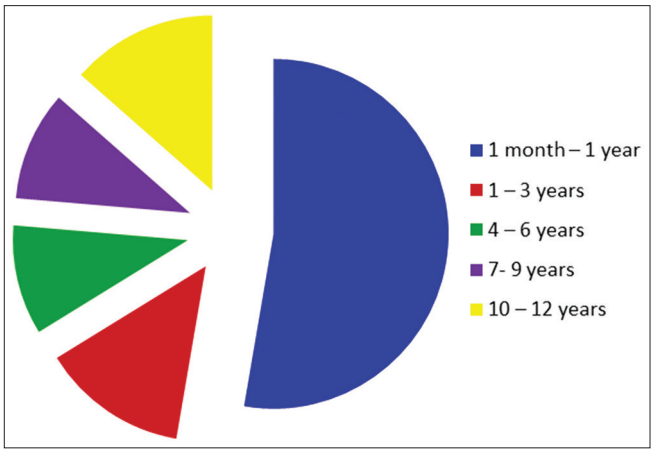


**Figure 3: Age distribution of cardiac failure cases**

common making 35.37% ($n$ = 29) followed by combined left to right shunt lesions in various combinations and atrioventricular septal defects. The common cyanotic CHD causing failure in our study is transposition of great arteries (4.88%, $n$ = 4) (Table 6 and Figure 6).

Almost all rheumatic heart disease cases in our study had involvement of mitral valves. The most common rheumatic valvular lesion is isolated MR. The second most common valve involved was aortic valve. One child had isolated mitral stenosis (MS) and two children had MS with associated MR (Table 7 and Figure 7).

**Table 5: Cardiac etiologies of heart failure**

| Cardiac etiologies | Frequency (%) |
|---|---|
| CHDs | 82 (59.85) |
| Rheumatic heart diseases | 21 (15.33) |
| Dilated cardiomyopathy | 10 (7.30) |
| Myocarditis | 7 (5.11) |
| Supraventricular tachycardia | 2 (1.46) |
| Infective endocarditis | 3 (2.19) |
| Pulmonary hypertension | 3 (2.19) |
| Scorpion sting | 4 (2.92) |
| Pompe's disease | 1 (0.73) |
| MPS | 2 (1.46) |
| Undiagnosed | 2 (1.46) |
| Total | 137 (100) |

MPS: Mucopolysaccharidosis, CHDs: Congenital heart diseases

**Table 6: Breakup of CHDs causing heart failure**

| ??? | Frequency (%) |
|---|---|
| VSD | 29 (35.37) |
| ASD | 3 (3.66) |
| Patent ductus arteriosus | 4 (4.88) |
| Aortopulmonary window | 1 (1.22) |
| Combined shunt lesions | 15 (18.29) |
| Atrioventricular septal defect | 12 (14.63) |
| Aortic stenosis | 1 (1.22) |
| Congenital MR | 1 (1.22) |
| Transposition of great arteries | 7 (8.54) |
| Total anomalous pulmonary venous return | 4 (4.88) |
| Tetrology of fallot | 1 (1.22) |
| Tricuspid atresia | 1 (1.22) |
| Ebstein anomaly | 2 (2.44) |
| Double outlet right ventricle | 1 (1.22) |
| Total | 82 (100) |

MR: Mitral regurgitation, VSD: Ventricular septal defect, ASD: Atrial septal defect, CHDs: Congenital heart diseases

**Table 7: Breakup of rheumatic heart diseases causing heart failure**

| ??? | Frequency (%) |
|---|---|
| MR | 13 (61.90) |
| MR and AR | 5 (23.81) |
| MS with MR and AR | 1 (4.76) |
| MS with regurgitation | 1 (4.76) |
| MS | 1 (4.76) |
| Total | 21 (100) |

AR: Aortic regurgitation

Among the 148 cases of cardiac failure, 11 had noncardiac etiologies. Anemia was the commonest noncardiac cause of cardiac failure in our study (72.72%, $n$ = 8). Two

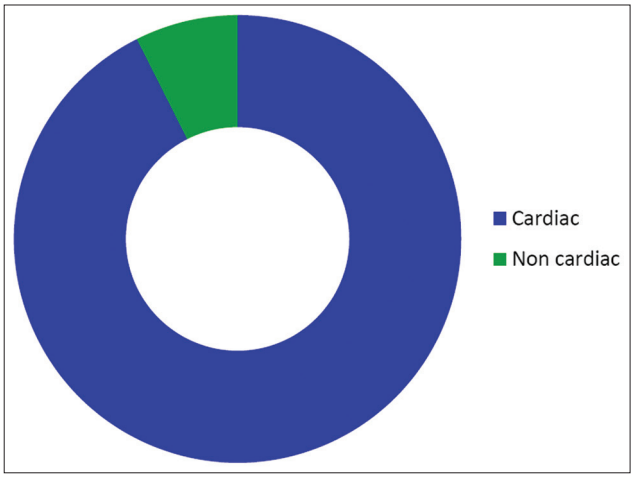


**Figure 4: Etiology of cardiac failure cases**

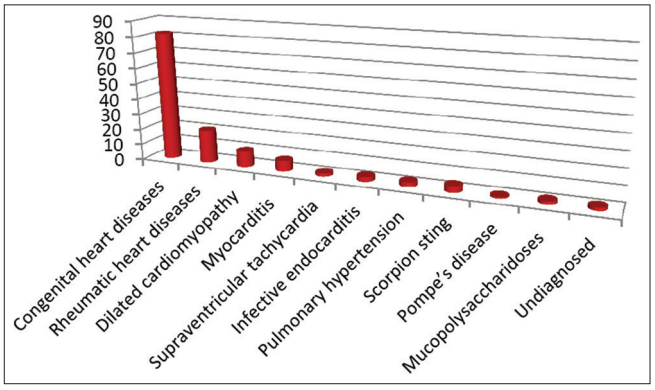


**Figure 5: Cardiac etiologies of heart failure**

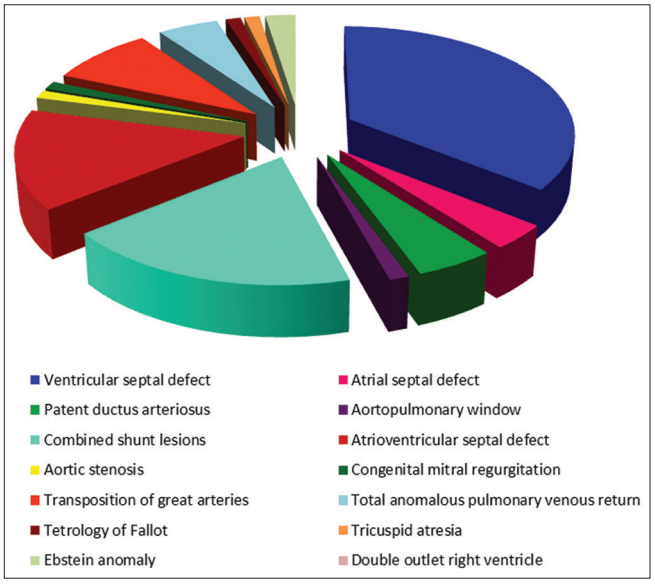


**Figure 6: Breakup of congenital heart diseases causing heart failure**

children developed cor pulmonale due to bronchiectasis and 1 had uremia as a cause of cardiac failure. (Table 8 and Figure 8).

## DISCUSSION

In our study, the proportion of children admitted with cardiac failure among the total admissions during 1 year period in the pediatric ward in a tertiary care center is 2.09%.

Massin *et al.*[6] in a prospective study of 1196 children over 10 years, reported heart failure in 124 children (10.4%) with congenital and acquired heart diseases.[6] 5.8% of total pediatric admissions in Lagunju and Omokhodion's study of Ibadan study[7] is due to CHF. The prevalence rate of CCF in Adekanmbi *et al.*'s[8] study is 7.02%.

In our study, among the total children admitted with cardiac failure the male to female ratio is 1.3:1. The prevalence was higher among males than females in Rashid *et al.*'s[9] study. In the study by Lagunju and Omokhodion,[7] the male to female ratio is 1.2:1. Amoah and Kallen,[10] in their study, reported the incidence of CCF among male to female was 1.2:1.

**Table 8: Noncardiac etiologies of heart failure**

| Non cardiac causes | Frequency (%) |
|---|---|
| Anemia | 8 (72.73) |
| Bronchiectasis | 2 (18.18) |
| Renal failure | 1 (9.09) |
| Total | 11 (100) |

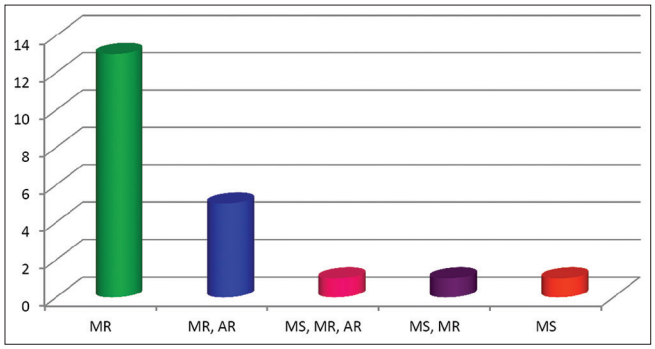


**Figure 7: Breakup of rheumatic heart diseases causing heart failure**

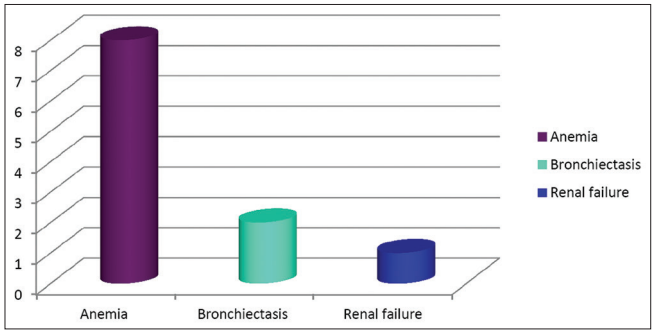


**Figure 8: Noncardiac etiologies of heart failure**

The prevalence of CHF is higher in children under 1 year of age in our study. 52.70% of the total admissions due to cardiac failure are between 1 month and 1 year of age. Keith[11] reported that 90% of failure due to CHD occurred in 1st year of life. Keith in 1956 pointed out that if a patient of CHD does not develop failure within the 1st year of life, he is not likely to develop so in the next 10 years, unless complicated by anemia, infection or infective endocarditis.[12] In Rashid *et al.*'s study,[9] only 36% of the children with CCF fall under 1 year of age and the common age group with CCF is between 1 and 5 years of age. In Lagunja *et al.*'s study,[7] 54% of admissions due to CHF is between 1 month and 1 year of age.

In this study, the overall common cause of CCF in children is CHDs. (55.4%). In Adenkambi *et al.*'s[8] study, anemia forms the common cause (46%). In Lagunju and Omokhodion's study,[7] the common cause of CHF was found to be acute lower respiratory tract infections (LRTIs) (36%).

Cardiac causes constitute 92.57% of all admissions due to cardiac failure and noncardiac causes forms 7.43%. Cardiac causes are more common than noncardiac causes. In Rashid *et al.*'s[9] study, cardiac causes are 84% and noncardiac causes are 16% of cases of CCF. Luganju *et al.*[7] report 31% cardiac causes and 69% of noncardiac causes as the etiology of CHF. In Adenkambi *et al.*'s study,[8] cardiac causes from 12.5% and noncardiac causes from 87.5%.

In this study, among the cardiac causes, CHDs from the common cause of CCF admissions under 1 year of age and also between the age of 1 and 3 years. In more than 3 years to <6 years age group, myocardial dysfunction due to scorpion sting forms the common etiology. Rheumatic heart disease is the most common cause in more than 6 years of age. Rashid *et al.*[9] reported that CHDs are the common cause of CHF in <1 year age group. Myocarditis constitutes the common cause between 1 and 5 years age group. Rheumatic carditis is the common cause in children more than 5 years of age.

CHDs from 59.85% of the cardiac etiology of CCF in this study. Among the CHDs, left to right shunt lesions constitute the major cause for CCF in <3 years of age. VSD is the most common among the CHDs causing CCF in this study forming 35.37% of the heart failure cases due to CHD. Rashid *et al.*[9] reported that 44% of the CHD cases causing CCF in their study were due to VSD.

In Luganju *et al.*'s study,[7] CHDs constituted 25% of total cases due to CCF and VSD constituted 44% of cases of CCF due to CHDs.

Combined shunt lesions constitute 18.29% of admissions due to CHD causing CCF in our study. Among them, 9 were due to VSD and atrial septal defect (ASD), 1 VSD and patent ductus arteriosus (PDA), 1 VSD, ASD and PDA, 3 VSD and pulmonic stenosis (PS) and 1 ASD, mitral valve prolapse and MR. Lagunju and Omokhodion[7] reported 5 cases of combined shunt lesions all of them being VSD and ASD which constituted 20% of the CHD causing CCF.

Among 16 congenital cyanotic heart diseases that presented with heart failure, transposition of great vessels (TGV) – 7 cases – 8.54% of the CHDs causing CCF. Three cases of TGV had associated ostium secundum ASD, 1 with VSD, 1 with PDA, 1 with AVSD and the remaining 1 associated with single ventricle. In Laganju *et al.*'s study, among the 25 CHDs as the cause of CCF, there were 4 transient global amnesia (TGA) cases forming 16%.

According to Rao,[13] Group I TGA with intact ventricular septum the infant usually become symptomatic within the 1st week of life. Group II TGA with VSD presents with symptoms of CCF between 4 and 8 weeks of life. Moreover, Group III TGA with VSD and PS, the presentation is variable depending on the severity of PS.

In this study, the second common cardiac cause being rheumatic heart disease with failure – 21 cases, constitutes 14.19% of the admissions due to cardiac failure and 15.33% CCF due to cardiac cause. Acute rheumatic carditis was noticed in 11 cases and pulmonary hypertension in 7 cases. Rupture chordae was recognized in 3 of 21 cases. In Adekanmbi *et al.*'s study,[8] 1% of CCF cases are due to rheumatic heart disease constituting 54% of the cases. In Laganja *et al.*'s study,[7] rheumatic heart disease is responsible for 6% of CCF cases and the most common valvular lesion in affected patients was MR and it was seen in all. MS was associated in 3 cases.

About 10 out of 137 admissions due to cardiac causes of CCF in this study were due to dilated cardiomyopathy, forming 7.30%. One case had associated left ventricular (LV) clot, one case was associated with proximal renal tubular acidosis and rickets. Absent right depressor angularis muscle was noticed in one child. Moreover, one case presented with left hemiparesis. Lipshultz *et al.* report the incidence of cardiomyopathy in pediatric patients as 1.13 per 100,000 in the United States.[14] Nugent *et al.* report that in pediatric patients the incidence of cardiomyopathy in Australia is 1.24 per 100,000.[15] Primary cardiomyopathies are the principal cause of heart failure signs and symptoms in children with a structurally normal heart. In Jeffrey *et al.*'s study, the annual incidence of dilated cardiomyopathy in children <18 years was 0.57 cases per 100,000 per year. Incidence was higher in boys than in girls (0.66 in boys and 0.47 in girls per 100,000). Among them, 66% of the cases are idiopathic. Among the known causes, 46% are due to myocarditis and 26% are due to neuromuscular disorders. 1 and 5 years rates of death or transplantation were 31% and 46%, respectively.[16]

Seven cases of myocarditis, 3 male and 4 female got admitted with CCF in this study. It forms 5.11% of the cardiac causes of CCF in this study. Among them, pneumonia was present in four cases. Two cases had diarrhea. Myocardial dysfunction due to scorpion sting was noticed in four cases. Myocarditis constituted 1% of CCF cases in the study by Adekanmbi *et al.*[8] Acute viral myocarditis formed the overall common etiology of CCF in Rashid *et al.*[9] constituting 53% of cases between 1 and 5 years age group.

Das *et al.* in a study of 32 children admitted with scorpion envenomation reported that 16 (50%) children developed myocarditis and also reported that ECG is a sensitive indicator of myocarditis.[17]

A 2-month-old child presented with cardiac failure in this study, his ECG showing high voltage QRS complexes, cardiomegaly in chest X-ray, ECHO showing concentric LV hypertrophy with global LV dyskinesia, mild LV dysfunction and Grade II MR. The level of acid maltase in his serum was 24 n mol/h/mg (normal level >60 n mol/h/mg), and a diagnosis of Pompe's disease was made. It constitutes 0.73% of cardiac causes of CCF.

Jacob *et al.* reported a 5 months old child with typical features of Pompe's disease in the form of hypotonicity, hyporeflexia, X-ray evidence of cardiomegaly, ECG evidence of short PR interval, ECHO evidence of LV hypertrophy, and histopathological evidence of increased glycogen accumulation in skeletal muscle biopsy. Death occurred 2 months after symptom onset. There is a history of similar illness in previous two siblings present.[18]

About 1.46% of the cardiac causes of CCF in this study are due to valvular lesions due to mucopolysaccharidoses in this observational study. One 12-year-old child had increased urinary glycosaminoglycan concentration (357.22 mg GAGs/g of creatinine against the normal value of 10.77-77.5) and undetectable levels of arylsulfatase B activity and grouped as Type VI MPS, Maroteaux-Lamy syndrome. Her ECHO revealed MS, dilation of the left atrium and right ventricular hypertrophy.

Wippermann *et al.* in a study of 84 children with MPS reported 64.3% of MR and 40.5% of aortic regurgitation (AR). The frequency of aortic and/or MR was 75% in all patients, 89% in MPS I, 94% in MPS II, 66% in MPS III, 33% in MPS IV, and 100% in MPS VI. Combined MR and AR was present in 29%.[19]

Nearly 7.43% of CCF admissions were due to noncardiac causes. Adekanmbi *et al.* reported 87.5% of CCF cases were due to noncardiac causes. Noncardiac causes constituted 16% of etiology of CCF in Rashid *et al.*'s study. In Lagunju and Omokhodion's study, 69% of CCF cases were of noncardiac etiology.

Among 11 admissions due to noncardiac causes, 8 cases were due to anemia (72.73%), 2 cases were due to bronchiectasis (18,18%), and one case due to chronic renal failure (9.09%).

All 8 anemia cases had Hb <6 g and all had cardiomegaly. Thalassemia – 1, hypochromic microcytic anemia – 1, dimorphic anemia – 2, pure red cell aplasia – 1, osteopetrosis – 2, and undiagnosed – 1. Anemia constitutes 5.41% of total admissions due to CCF in our study. In Adekanmbi *et al.*'s study,[8] anemia is the most common cause of CCF, anemia alone constituting 46% and anemia and LRTIs constituting 11.5%. In their study, malaria was the most common cause of anemia. Lagunju and Omokhodion[7] reports 28% of CCF is due to anemia in their study, malaria being the common cause constituting 16% of anemia cases followed by sickle cell anemia forming 5%, septicemia 5%, prematurity 1%, and acute lymphoblatic leukemia 1%.

Khan *et al.* in a study of 212 patients with beta Thalassemia observed clinical CHF in 33 (15.6%) patients with the age range between 8 and 21 years.[20]

Two female children, 7 years and 11 years old admitted with bronchiectasis and CHF in this study. Both had CT evidence of bilateral bronchiectasis and dilated right atrium and ventricle with pulmonary hypertension in ECHO. LRTIs constitute 1.35% of the total CCF admissions in this study. Lower respiratory infections are the most common cause of CCF in Lagunju and Omokhodion's[7] study constituting 36%. In Adekanmbi *et al.*'s[8] study, isolated LRTI contributed 29% of CCF cases and 11.5% of CCF cases are due to LRTI and anemia.

One male child of 12 years, who is a known case of posterior urethral valve with chronic renal failure presented with CCF in our study constituting 0.68% of total admissions due to CCF. CRF is the cause of 1% and 3% of CCF cases in Adenkambi *et al.*[8] and Lagunju and Omokhodion,[7] respectively. Liang *et al.* presented a 1.5-year-old girl developed CCF 9 months after she presented with hypertension. The hypertension was caused by renal artery stenosis.[21] Parekh *et al.* cardiac complications are the major cause of death in 25% of children with advanced chronic renal failure.[22]

## CONCLUSIONS

In infants and children, early clinical and ECHO is needed to identify the structural heart lesions. Regular follow-up by applying Modified Ross Classification of Heart Failure to assess the progress of diseases will help in early recognition of failure and early referral for surgery. Early surgical intervention is needed for cases amenable to surgery to prevent morbidity and mortality. Precipitating causes like anemia, infections are to be identified and treated early. Valvoli Thittam which was pioneered by Government of Tamil Nadu 20 years back, empowering the school teachers to recognize sore throat and to treat with oral penicillin for 10 days needs to be reintroduced.

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