



## Research

### Disease Burden of Rheumatic Heart Disease Among Children Attending the Paediatric Cardiology Unit Teaching Hospital Jaffna.

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#### Abstract

**Background:** Rheumatic heart disease is the most common cause of acquired heart disease in children worldwide. It causes significant morbidity and mortality in developing countries.

**Objective:** To identify the disease burden of rheumatic heart disease and identify the spectrum of disease with its complications.

**Method:** It was a retrospective, descriptive study carried out at the Paediatric cardiology unit Teaching Hospital Jaffna. The echocardiographic data was collected over a period of three and a half years starting from September 2013 till March 2017. Children with echocardiographic diagnosis of Rheumatic heart disease were selected as study population.

**Results:** One hundred and twenty three children were diagnosed with Rheumatic heart disease during the study period. The male to female ratio was 1.4:1. Majority (96%) of the study population were within the age group of 5-15 years. One hundred and nineteen (96.7%) children had mitral regurgitation, and sixty seven children (54.4%) had aortic regurgitation. Both valves were affected in 63 children (51.2%). Complications were noted only in 7 Children (5.7%). Complications included heart failure and recurrence of rheumatic carditis. Six children needed surgical repair of mitral and/or aortic valves. Mitral valve repair was the commonest (3.3%).

#### Introduction

Rheumatic heart disease is an immune mediated disease caused by group A Streptococcus. It is the most common cause of acquired heart disease in children worldwide. According to the World health Organization (WHO) rheumatic fever or rheumatic heart disease (RHD) affects about 15.6 million people worldwide with 282,000 new cases and 233,000 deaths each year<sup>(1)</sup> The majority of these cases (79% of the rheumatic heart disease) came from less developed countries<sup>(1)</sup>. There are 2.4 million affected children between five and fourteen years of age in developing countries<sup>(2)</sup>. The peak incidence of RHD is noted in children between 5-15 years. This indicates the disease burden of the illness worldwide.

#### Objectives

The objection and the study were to identify the disease burden of rheumatic heart disease & evaluate the age and sex distribution, geographical areas where the disease is more prevalent and identify the spectrum of disease with its complications and assess the percentage of children needing surgical intervention.

#### Methods:

In this retrospective descriptive study, the echocardiographic data was collected over a period of three and a half years starting from September 2013 till March 2017.

The study was carried out at the Paediatric cardiology unit, Teaching Hospital Jaffna. Children with echocardiographic diagnosis of rheumatic heart disease were selected as the study population

The study instrument was a questionnaire. It had details of personal demographic profile, type of cardiac valve involved, the severity of cardiac involvement and the complications of rheumatic heart disease. The study instrument was pretested before commencing the study.

Out of electronic pooled data in the Paediatric cardiology unit, the children with echocardiographic diagnosis of rheumatic heart diseases were selected and their details were retrieved from the database.

Ethical clearance was obtained from Ethical Review committee, Faculty of Medicine, University of Jaffna and approval from Director Teaching Hospital was obtained before commencing the study.



**Results**

A total of 18949 echocardiographic examinations were done at the paediatric cardiology unit, Teaching Hospital, Jaffna during the study period of three and a half years. 63% (11956) were new referrals. 38% (4449) of children who were newly referred had abnormal echocardiographic findings. Out of those children with abnormal echocardiographic findings 89 children were diagnosed with rheumatic heart disease. Thirty four (34) children were already on follow up with rheumatic heart disease. A total of one hundred and twenty three children were diagnosed with rheumatic heart disease. This indicates that rheumatic heart disease contributes to 2% of disease burden on newly diagnosed heart disease in the paediatric cardiology unit at Teaching hospital Jaffna. Out of 123, 72 were male and 51 were females with the ratio of 1.4:1

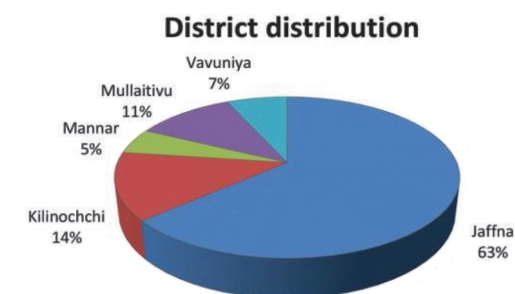


Figure 1 Geographic distribution of children with Rheumatic heart disease

Even though the above figure indicates the prevalence is more in Jaffna district, when we analyze the disease burden with regards to population density the prevalence is more in the Kilinochchi and Mullaitivu district. Table 2 clearly indicates the disease burden of rheumatic heart disease with regards to population density in all districts.

Age at diagnosis	Frequency
<5 years old	3.3% (4)
5-10	56.9% (70)
11-15	39.8% (49)

Table 1: Age at diagnosis of Rheumatic heart disease

Table 1 demonstrates the age distribution of the study population at the time of diagnosis. Mean age at diagnosis of rheumatic heart disease was 9.9 years with a standard deviation of 2.49. The youngest age of diagnosis was two years. Two children were diagnosed at the age of two years and one child was diagnosed at the age of three.

Figure 1 demonstrates the geographic distribution of the children with rheumatic heart disease. Majority (63%) of the patients were from Jaffna district and it was followed by Kilinochchi (13%).

District	Frequency	Per 100,000	Population (According to last census)
Jaffna	75	12.9	583071
Kilinochchi	16	14.2	112872
Mannar	6	6.1	99063
Mullaitivu	13	14.1	92228
Vavuniya	9	7.3	172789

Table 2: Disease burden of rheumatic heart disease with regards to population density

All the children were regularly followed up in the paediatric cardiology unit with regular echocardiograms except 13 % (n=17) who defaulted their follow up after the initial visit.

Mitral regurgitation is the commonest echocardiographic finding in our study population. 96% (n=119) had mitral regurgitation and 54% (n=67) had aortic regurgitation. Both valves were involved in 51% (n=53). Out of 119 children who had mitral valve involvement 15% (n=18) had severe mitral regurgitation and 40% (n=48) had moderate mitral regurgitation.

During the follow up imaging valvular lesions had completely disappeared in 1.6% (n=2) of the study population.



The severity of lesions improved in most of the children. The percentage of children who had severe mitral regurgitation has come down from 15% to 4.2% even though only 3% underwent surgical repair of the lesion.

Two children had congenital heart disease in addition to the rheumatic heart disease. One had a patent foramen ovale and the other one had a ventricular septal defect with subaortic membrane and ruptured sinus of Valsalva.

Complications were noted only in 5% (n=7) of children. Table 3 demonstrates type and frequency of complications. 94% (n=116) of children did not have any complication. Among those who had recurrence of rheumatic heart disease one child defaulted follow up for three months and didn't receive benzathine penicillin prophylaxis. Other one was followed up in other hospitals and no previous records were available.

Complications	Frequency
Heart failure	2.4% (3)
Pericardial effusion	0.8% (1)
Mitral stenosis and Pulmonary hypertension	0.8% (1)
Recurrence of rheumatic heart disease	1.6% (2)

Table 3 Complications of rheumatic heart disease

All the children with rheumatic heart disease were treated with high dose of aspirin in the initial period and 2.4% (n=3) needed oral steroids at the acute stage. The indications to start steroids in these children were heart failure (one child) and progression of valve lesion in spite of high dose of aspirin. With steroid treatment two had clear improvement in echocardiographic findings and the third one had defaulted the follow up and could not be assessed for the outcome.

All three children with heart failure received anti failure treatment which was needed nearly 6-9 months. Regular intramuscular benzathine penicillin prophylaxis was recommended and given for all the children with rheumatic heart disease three weekly.

Six children needed surgical intervention for the complications of rheumatic heart disease. Four (3.3%) underwent surgery and one (0.8%) was waiting for surgery at the time of last visit. Table 6 shows the indications for surgery.

Four children (3.3%) needed surgery involving the mitral valve and one (0.81%) involving the aortic valve.

Three patients with mitral valve disease developed mitral stenosis with the time and needed repair.

	Indications for surgery
Patient 1	Moderate mitral stenosis
Patient 2	Severe MR, Mild MS, Surgery was declined as high risk for surgery due to poor LV function(He also had VSD, AR, SAM,RVOT obstruction)
Patient 3	Severe MR
Patient 4	MS, mild to moderate pulmonary hypertension
Patient 5	Severe MR
Patient 6	Severe MR, LA/LV dilated- acute on chronic MR

Table 7 Indications for surgery in children with Rheumatic heart disease

## Discussion

Rheumatic heart disease remains the commonest acquired heart disease in children in developing countries although its incidence had declined in developed countries over the past few decades. Not only the increased incidence but also the late presentation which lead to the severe valvular involvement contribute to the significant burden on paediatric cardiology and paediatric cardiac surgery units in the developing countries.

The sex predominance of rheumatic heart disease varies in different parts of the world. There was female predominance in Nepal and Nigeria with the rate of 64%, 52% respectively<sup>(3,4)</sup>. Male predominance was seen in Lebanon (58%)<sup>(5)</sup>. It is found in a study done in Sri Lanka that initially girls had been predominantly affected whereas now RF and RHD appear to be commoner in boys<sup>(6)</sup>. Our study shows male predominance (59%) which is similar to the previous study done at LRH (57%)<sup>(6)</sup>.

Rheumatic heart disease is common in the age group of 5-15 years. The mean age of diagnosis varies slightly between studies. When compared to the Nepal study where the mean age of diagnosis was 11.9 years and youngest age of diagnosis was 5.9 years<sup>(3)</sup> our study shows a younger population with rheumatic heart disease. Our mean age of diagnosis was 9.9 years with the youngest age of diagnosis was two years. This may indicate that younger populations are exposed to more group A streptococcal disease, in our context.

Mitral regurgitation is the commonest echocardiographic finding in our study population which is similar with other studies<sup>(3-6)</sup>.



But the percentage of children with both valve involvement is more in our study population (51%) when compared to other studies where it is between 10-30% only<sup>(3-6)</sup>. Severity of mitral regurgitation is also high in our study population. Out of those who had mitral regurgitation moderate to severe mitral regurgitation was seen in 65% in our study when compared to the Lebanon study where it was only 21%<sup>(5)</sup>. Both of this may be due to the late presentation and later diagnosis in our study. Late diagnosis and late referral may reflect the poor primary care system at the moment in Sri Lanka

Complications were noted less frequently in our study population. This may be due to the short period of follow up of our study population. Forty four percent of children developed heart failure and 11% developed pericardial effusion in the Lebanon study,<sup>(5)</sup> whereas only 2.4% developed heart failure and 0.8% developed pericardial effusion in our study. In an Indian study recurrence of rheumatic fever occurred in 22 children out of 85 children with acute rheumatic fever, in which study 10 children had rheumatic heart disease in the initial period and recurrence were noted<sup>(7)</sup>. In our study only 1.6% had recurrence. Both in our study and Indian study the recurrence has been observed when they missed the penicillin prophylaxis. This indicates the importance of good compliance of secondary prophylaxis to prevent the recurrence of illness.

Steroids were given for three children with rheumatic heart disease during the acute phase of illness with rapidly progressing valve regurgitation on echocardiogram in spite of high dose aspirin. Even though it is not indicated in the literature to use steroids for progressive valve damage without heart failure, we have used steroids in three children with good evidence of improvement in the valve function. This needs further studies with larger sample size to prove the benefits of steroids in limiting the valve damage.

Rheumatic heart disease was the second common cause for open heart surgery in children at Nepal. Out of 103 children with rheumatic heart disease 40 children under went surgical correction in Nepal study<sup>(3)</sup> and 31% of the study population underwent surgical correction in Lebanon study<sup>(5)</sup>. In our study even though 4.4% needed surgical intervention only 3.3% underwent surgical correction and 1.2% are waiting for surgical intervention.

The less number of children needing surgical correction may be due to the short duration of follow up on our study population.

But our study shows that those needing surgical correction also had to wait for a longer period due to non-availability of the surgical correction in time.

## Conclusion

Our study highlights that

- Rheumatic heart disease in children still contributes to a significant burden both on cardiology and cardiac surgery in our country.
- Recurrence of rheumatic heart disease is seen when there is poor compliance of penicillin prophylaxis.
- Steroids play a role in the improvement of valvular lesions when there is progression of valve damage in spite high dose of aspirin. This needs further studies with larger study group.

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