

Clinical profile of children with rheumatic heart disease in Indonesia

*I Ketut Alit Utamayasa^{1,2}, Ajeng Indriastari¹, Taufiq Hidayat¹, Rendi Aji Prihaningtyas¹, Mahrus Rahman¹, Teddy Ontoseno¹

Sri Lanka Journal of Child Health, 2021; **50**(2): 200-202

Abstract

Introduction: In developing countries, rheumatic heart disease (RHD) affects 19 per 100.000 children. There are few systematically collected data on children with RHD in developing countries.

Objective: To describe the clinical profile of children with RHD in Indonesia.

Method: A cross-sectional study was conducted on children diagnosed with RHD who came to the paediatric cardiology outpatient clinic of Dr. Soetomo General Hospital from June 2014 to June 2015. Demographic data and clinical characteristics of RHD were collected.

Results: Fifty children were diagnosed with RHD during the study period. The median age was 10.9 years. Female to male ratio was 1.2:1. At the time of initial diagnosis, 28% children had acute rheumatic fever (ARF) and 72% had RHD. Major clinical manifestations were carditis (86%), polyarthritis migrans (10%) and chorea (2%). Minor clinical manifestations were fever (36%), arthralgia (8%), positive C-reactive protein (60%), elevated erythrocyte sedimentation rate (68%) and prolonged PR interval on electrocardiogram (2%). Raised anti-streptolysin O titre was present in 44%. The common valve lesions were MR (86%), tricuspid regurgitation (42%) and aortic regurgitation (38%).

Conclusions: Carditis was the commonest major clinical manifestation in past ARF and MR the commonest valve lesion in children with RHD.

¹Child Health Department of Faculty of Medicine Universitas Airlangga, Indonesia and Dr. Soetomo General Hospital Surabaya, Indonesia, ²Medical Doctoral Programme Student at Faculty of Medicine Universitas Airlangga, Indonesia and Dr. Soetomo General Hospital Surabaya, Indonesia
*Correspondence: alit_tusari@yahoo.com

 <https://orcid.org/0000-0002-2510-8689>

(Received on 03 April 2020; Accepted after revision on 22 May 2020)

The authors declare that there are no conflicts of interest

Personal funding was used for the project.

Open Access Article published under the Creative Commons Attribution CC-BY  License

Conclusions: Carditis was the commonest major clinical manifestation in past ARF and MR the commonest valve lesion in children with RHD.

DOI: <http://dx.doi.org/10.4038/sljch.v50i2.9554>

(Keywords: acute rheumatic fever, rheumatic heart disease)

Introduction

Rheumatic heart disease (RHD) is an important sequela of acute rheumatic fever (ARF)¹. Around 40% of children with ARF can develop pancarditis². In developing countries RHD affects 19 per 100.000 children^{1,3,4}. Jones criteria for diagnosis of ARF include 5 major and 4 minor criteria with an absolute need for evidence (microbiologic or serologic) of recent group A streptococcal infection^{1,5}. All children with a past history of acute RF or murmurs suggestive of valve disease require echocardiography. Serial echocardiography is crucial in diagnosis and follow-up of RHD and in timing of any required surgery⁶. Although secondary prophylaxis is important to reduce the morbidity and mortality associated with the disease, ensuring adequate compliance is a challenging task². There are few systematically collected data on children with RHD in developing countries.

Objective

To describe the clinical profile of children with RHD in Indonesia.

Method

A cross-sectional study was conducted in a tertiary government medical centre. Subjects were children diagnosed with RHD who visited the paediatric cardiology outpatient clinic of Dr. Soetomo General Hospital, Indonesia from June 2014 to June 2015. Demographic data and clinical characteristics of RHD were collected. No sample size was calculated as all children registered during the study period were eligible for inclusion. The first working diagnosis at the first visit to the paediatric cardiology outpatient clinic was also determined.

Ethical issues: Approval was obtained from the Institutional Ethics Committee of the Faculty of Medicine, University of Indonesia to collect data and review the records of included subjects.

Statistical analysis: Data were encoded using SPSS. Descriptive statistics were used for all variables. For nominal data, frequencies and percentages were used. For numerical data, mean ± SD was used. Appropriate graphs and tables were used to summarize data.

Results

During the study period, 50 children were diagnosed to have RHD. The characteristics of these children are shown in Table 1.

Table 1
Characteristics of study population (n=50)

Variable	n (%)
<i>Sex</i>	
Female	23 (46)
Male	27 (54)
<i>Age group in years</i>	
6-11	26 (52)
12-18	24 (48)
<i>Nutritional status</i>	
Normal	27 (54)
Underweight	23 (46)
<i>Working diagnosis</i>	
Acute rheumatic fever	14 (28)
Rheumatic heart disease	36 (72)

The median age of the study population was 10.9 years (range 6–15 years). Female to male ratio was 1.2:1. At the time of first diagnosis, 28% had acute rheumatic fever (ARF) and 72% had RHD (Table 1).

The clinical and laboratory findings are shown in Table 2.

Table 2
Clinical and laboratory findings (n=50)

Variable	n (%)
<i>Major manifestations</i>	
Carditis	43 (86)
Polyarthritits migrans	05 (10)
Chorea	01 (02)
Erythema marginatum	0 (0)
Subcutaneous nodules	0 (0)
<i>Minor manifestations</i>	
Fever	18 (36)
Arthralgia	04 (08)
Elevated acute phase reactants:	
• ESR	34 (68)
• C-reactive protein	30 (60)
Prolonged PR Interval	01 (02)
<i>Evidence of preceding streptococcal infection</i> ASOT (>333 units)	22 (44)

ASOT: Antistreptolysin O titre,
ESR: Erythrocyte sedimentation rate

The abnormalities on electrocardiography were prolonged PR interval in one child and axis deviation in 2 children.

Echocardiographic findings are shown in Table 3. Common valve lesions were mitral regurgitation (MR) in 43 (86%) children, aortic regurgitation (AR) in 19 (38%) children and tricuspid regurgitation (TR) in 22 (44%) children (Table 3).

Table 3: Echocardiography findings (n=50)

Variable	n (%)
<i>Mitral regurgitation</i>	
Mild	04 (08)
Moderate	17 (34)
Severe	20 (40)
Prolapse (Anterior mitral leaflet)	02 (04)
<i>Aortic regurgitation</i>	
Mild	10 (20)
Moderate	04 (08)
Severe	05 (10)
<i>Tricuspid regurgitation</i>	
Mild	17 (34)
Moderate	01 (02)
Severe	04 (08)

Discussion

ARF characteristically affects the cardiac valves, but can affect other cardiac structures¹. Prevalence of RHD is very low in developed countries being <0.5 per 1000³. In our study of 50 children diagnosed with RHD the median age was 10.9 years and the female to male ratio was 1.2 :1. On initial diagnosis 14 (28%) had ARF and 36 (72%) had RHD. Echocardiographic anomalies suggestive of carditis have been found in children with normal clinical cardiac examination⁷. Carditis as an initial sign might be mild or even remain unrecognized⁸.

Rheumatic carditis affects the endocardium, myocardium and pericardium in varying degrees². Valvulitis is a special sign of rheumatic carditis, so that if there is myocarditis or pericarditis without valvulitis, it may not be caused by ARF². If there is functional disturbance in the pericardium, myocardium and endocardium, it could cause haemodynamic alteration and cause cardiomegaly². For diagnosis of ARF, using Jones criteria as a guideline, there should be 2 major manifestations or 1 major manifestation and 2 minor manifestation along with evidence (microbiologic or serologic) of recent group A streptococcal infection^{1,3,5}.

The electrocardiogram (ECG) is not very helpful in the diagnosis of ARF. In our study prolonged PR interval was found only in one patient. Whilst echocardiography is highly sensitive for detecting valve abnormalities, questions have been raised regarding its specificity^{9,10}. World Heart Federation

guidelines for echocardiographic diagnosis of RHD in children without a history of ARF aims to differentiate mild RHD from normal findings¹¹. 2D Doppler echocardiography is more sensitive than auscultation of the chest for detection of heart anomalies⁷. However, the degree to which it aids specific diagnosis of subclinical rheumatic carditis remains controversial and a subject of debate⁷. In our study, echocardiography revealed that in children the common valve lesions were MR (86%) AR (38%) and TR (44%). In the literature, RHD is typically characterized by leaflet inflammation of the mitral and/or aortic valves, pulmonary and tricuspid valves being more uncommonly involved⁸. Chronic MR is the commonest type of RHD in children and young adults, whereas mitral stenosis is commoner in the fourth to sixth decade³.

Conclusions

Carditis was the commonest major clinical manifestation in past ARF and MR the commonest valve lesion in children with RHD.

Acknowledgements

Authors thank Director and staff of Dr. Soetomo General Hospital, Surabaya for permitting study.

References

1. Gerber AM. Rheumatic fever. In: Behrman RE, Kliegman RM, Jenson HB, editors. *Nelson Textbook of Pediatrics* 19th ed. Philadelphia: Saunders, 2011: 920-8.
2. WHO global programme for the prevention of RF/RHD. Report of a consultation to review progress and develop future activities. Geneva, World Health Organisation, 2000. (WHO document WHO/CVD/00.1).
3. Tani LY. Rheumatic fever and rheumatic heart disease. In: Allen HD, Driscoll DJ, Shaddy RE, et al., editors. *Moss and Adam's Heart Disease in Infants, Children, and Adolescent: Including the Fetus and Young Adults*. 7th ed. Philadelphia: Lippincott Williams & Wilkins, 2008:1257-80.
4. Working Group on Paediatric Acute Rheumatic Fever, Cardiology Chapter of Indian Academy of Paediatrics, Saxena A, et al. Consensus guidelines on pediatric acute rheumatic fever and rheumatic heart disease. *Indian Pediatrics* 2008; **45**(7):565-73.
5. Park MK. Acute rheumatic fever. In: Park MK, ed. *Pediatric Cardiology for Practitioners*. 5th ed. Philadelphia: Elsevier; 2008:381-8. <https://doi.org/10.1016/B978-0-32304636-7.50029-0> PMID:PMC3523718
6. Wilson NJ, Neutze JM. Echocardiographic diagnosis of subclinical carditis in acute rheumatic fever. *International Journal of Cardiology* 1995; **50**:1-6. [https://doi.org/10.1016/01675273\(95\)02325-Q](https://doi.org/10.1016/01675273(95)02325-Q)
7. Caldas AM, Terreri MT, Moises VA, Silva CM, Len CA, Carvalho AC, et al. What is the true frequency of carditis in acute rheumatic fever? A prospective clinical and Doppler blind study of 56 children with up to 60 months of follow-up evaluation. *Pediatric Cardiology* 2008; **29**(6):1048-53. <https://doi.org/10.1007/s00246-008-9242-z> PMID:18825449
8. De Rosa G, Pardeo M, Stabile A, Rigante D. Rheumatic heart disease in children: from clinical assessment to therapeutical management. *European Review for Medical and Pharmacological Sciences* 2006; **10**(3):107-10.
9. Roberts K, Maguire G, Brown A, Atkinson D, Reményi B, Wheaton G, et al. Echocardiographic screening for rheumatic heart disease in high and low risk Australian children. *Circulation* 2014; **129**:1953-61. <https://doi.org/10.1161/CIRCULATIONAHA.113.003495> PMID:24622384
10. Roberts K, Colquhoun S, Steer A, Reményi B, Carapetis J. Screening for rheumatic heart disease: current approaches and controversies. *Nature Reviews Cardiology* 2013; **10**:49-58. <https://doi.org/10.1038/nrcardio.2012.157> PMID:23149830
11. Reményi B, Wilson N, Steer A, Ferreira B, Kado J, Kumar K, et al. World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease—an evidence-based guideline. *Nature Reviews Cardiology* 2012; **9**:297-309. <https://doi.org/10.1038/nrcardio.2012.7> PMID: 22371105 PMID: PMC5523449