

# Clinical profile of children with rheumatic heart disease in Indonesia

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## Clinical profile of children with rheumatic heart disease in Indonesia

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### 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%), polyarthritides 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.

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

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 (Keywords: acute rheumatic fever, rheumatic heart disease)

### Introduction

Rheumatic heart disease (RHD) is an important sequela of acute rheumatic fever (ARF)<sup>1</sup>. Around 40% of children with ARF can develop pancarditis<sup>2</sup>. In developing countries RHD affects 19 per 100.000 children<sup>1,3,4</sup>. 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<sup>1,5</sup>. 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 surgery<sup>6</sup>. Although secondary prophylaxis is important to reduce the morbidity and mortality associated with the disease, ensuring adequate compliance is a challenging task<sup>2</sup>. 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) |
| Polyarthritis migrans                                | 05 (10) |
| Chorea   | 01 (02) |
| Erythema marginatum                                  | 0 (0)   |
| Subcutaneous nodules                                 | 0 (0)   |
| <i>Minor manifestations</i>                          |         |
| Fever  | 18 (36) |
| Arthralgia   | 04 (08) |
| <i>Elevated acute phase reactants:</i>               |         |
| • 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 structures<sup>1</sup>. Prevalence of RHD is very low in developed countries being <0.5 per 1000<sup>3</sup>. 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 examination<sup>7</sup>. Carditis as an initial sign might be mild or even remain unrecognized<sup>8</sup>.

Rheumatic carditis affects the endocardium, myocardium and pericardium in varying degrees<sup>2</sup>. Valvulitis is a special sign of rheumatic carditis, so that if there is myocarditis or pericarditis without valvulitis, it may not be caused by ARF<sup>2</sup>. If there is functional disturbance in the pericardium, myocardium and endocardium, it could cause haemodynamic alteration and cause cardiomegaly<sup>2</sup>. 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<sup>1,3,5</sup>.

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<sup>9,10</sup>. World Heart Federation

guidelines for echocardiographic diagnosis of RHD in children without a history of ARF aims to differentiate mild RHD from normal findings<sup>11</sup>. 2D Doppler echocardiography is more sensitive than auscultation of the chest for detection of heart anomalies<sup>7</sup>. However, the degree to which it aids specific diagnosis of subclinical rheumatic carditis remains controversial and a subject of debate<sup>7</sup>. 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 involved<sup>8</sup>. Chronic MR is the commonest type of RHD in children and young adults, whereas mitral stenosis is commoner in the fourth to sixth decade<sup>3</sup>.

### Conclusions

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

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