SPECIAL ARTICLE

Diagnosis of Reumatic Fever: Which Modification?

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ABSTRACT We evaluated the implementation of diagnostic criteria on 547 ambulatory patients with rheumatic fever (RF) and rheumatic heart disease (RHD) between January 1, 1983 and December 31, 1992. The diagnosis of RF and reactive RHD was established by eiher revised Jones criteria, modified Jones criteria, or clinical judgment. The patient's age ranged from 4-18 years, 255 (46.6%) of them were boys. The clinical manifestations found were fever (58.1%), arthritis (41.9%), chorea (8.6%), subcutaneous nodule (1.3%), erythema marginatum (1.1%), holosystolic murmur (56.9%), mid-diastolic murmur (25.4%), and early diastolic murmur (29.5%). The laboratory changes were hemoglobin <10 g/dl (16.5%), BSR > 20 mm/h (56.3%), ASTO >200 U (29.6%), PR interval > 0.16 sec (13.3%), and CTR > 0.55 (27.6%). Revised Jones criteria were met in 162 cases (29%), modified criteria in 474 cases (86,6%), and clinical judgment in 521 cases (95.2%). **[Paediatr Indones 1994; 34:141-148]**

Introduction

In developed countries, the incidence of rheumatic fever (RF) and the prevalence of rheumatic heart disease (RHD) have decreased significantly in the last 3 decades in accord with the improvement in health, nutrition, and environment.[1,2] A very different situation is encountered in most developing countries including Indonesia, where RF and RHD are still a public health problem that need serious management.[3-5] The high incidence of RF as well as high prevalence of RHD in Indonesia need accurate clinical and laboratory assessments to avoid either under-diagnosis or over-diagnosis commonly encountered .[6,7]

The aim of this study was to evaluate the implementation of diagnostic criteria

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for RF and RHD besides analyzing and elaborating the clinical and laboratory findings of patients with RF and RHD at the Outpatient Clinic of the Division of Cardiology, Department of Child Health, University of Indonesia-Cipto Mangunkusumo Hospital, Jakarta, during the last 10 years.

Methods

This retrospective study was conducted in patients with RF and RHD at the Outpatient Clinic of the Cardiology Division, Department of Child Health, University of Indonesia Cipto Mangunkusumo Hospital during the period of January 1, 1983 until December 31, 1992. The diagnosis of RF and RHD was based on the following:[2,8,9] (a) revised Jones criteria, i. e., the presence of 2 major criteria or 1 major criterion and 2 minor criteria supported by evidence of beta-hemolytic group A streptococcal infection (Table 1); (b) modified Jones criteria, i.e., the presence of 2 major criteria or 1 major with 2 minor criteria without evidence of recent streptococcal infection; or (c) clinical judgment, i.e., the presence of 1 major criteria only, especialy carditis or chorea with or without one additional minor criterion.

Patients were divided into two groups according to their date of diagnosis. The first consisted of patients treated at the OPD during the first 5-year period (from January 1, 1981 to December 31, 1987) while the second group consisted of patients treated during the second 5-year period (from January 1, 1988 to December 31, 1992).

Table 1. Revised Jones criteria[2]

Major manifestation	Minor manifestation
Carditis	Clinical
Polyarthritis	Fever
Chorea	Arthralgia
Erythema marginatum	History of previous RF/RHD
Subcutaneous nodule	Laboratory
	ESR /, leukocytosis, CRP +
	Prolonged PR interval

plus

Evidence of previous streptococcal infection such as increased of ASTO test or others antibodies again streptococcus, finding of beta-hemolytic Streptococcus group A in throat swab, or evidence of recent scarlet fever. The presence of two major criteria or of one major and two minor criteria, indicates a high probability of acute rheumatic fever, if supported by evidence of streptococcal infection except in the presence of chorea or carditis with has occurred for a long time.

The variables assessed were age, sex, clinical presentations (i.e., fever, arthritis, chorea, subcutaneous nodule, erythema marginatum, heart murmur), laboratory findings (i.e., hemoglobin content, blood sedimentation rate, ASTO), PR interval on electrocardiography (ECG), and cardiothoracic ratio (CTR) on standard A-P photo. The clinical and laboratory data were available in the medical record of the Division of Cardiology, Department of Child Health. The medical record had been prepared for the purpose of cardiac investigations.

Data are presented in the form of text, tables, or pictures, elaborated manually or by using a computer program (EPI INFO version 5).

Results

There were 547 patients with RF or RHD who visited the Pediatric Cardiology Outpatient Clinic during in the last 10 years; it means that approximately 50 new cases of RF and RHD were treated each year. Their age ranged between 4 and 18 years with the mean of 10 years, most of them (61.4%) were between 8-12 years old. The age distributions of the patients are shown in Figs. 1, 2, and 3.

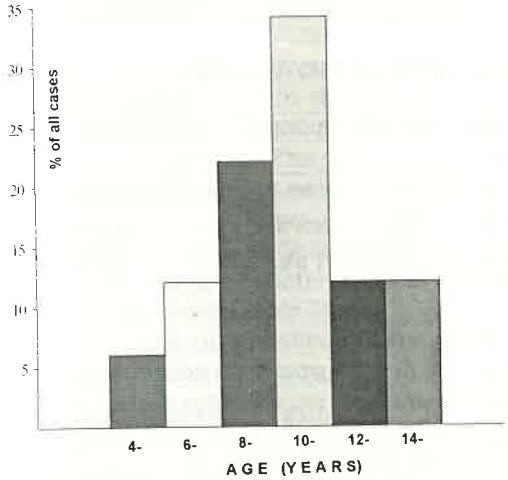

Figure 1. Age distribution of 547 RF and RHD cases (1983-1992)

Out of the total patients, there were 255 boys (46.6%) and 292 girls (53.4%). The clinical manifestations of all patients were described in Table 2, consisted of fever (85.1%), arthritis (41.9%), chorea (8.6%) subcutaneous nodule (1.3%), erythema margination (1.1%), pansystolic murmur (56.9%), middiastolic murmur (25.4%), and early diastolic murmur in 29.5%. This table shows that the cli-

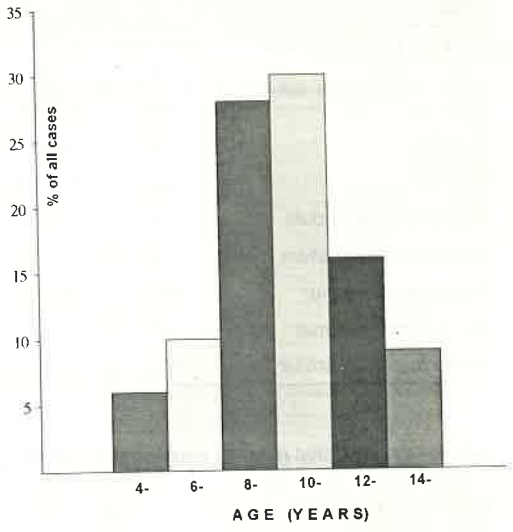

Figure 2. Age distribution of 290 RF and RHD cases (1983-1987)

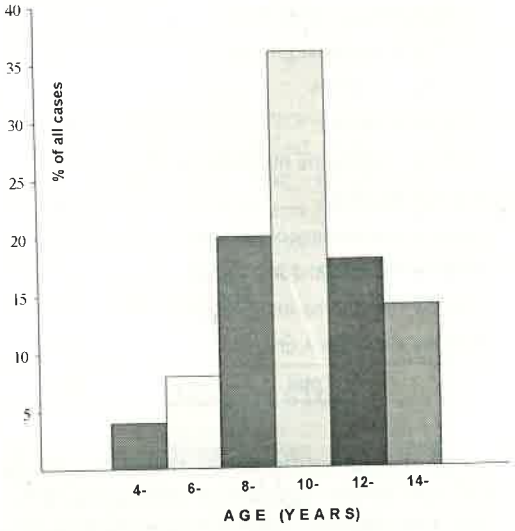

Figure 3. Age distribution of 257 RF and RHD cases (1988-1992)

Table 2. Clinical manifestations of 547 patients with RF and RHD

Clinical manifest ations	1983 - 1987 n=290		1988 - 1992 n=257		Total n=547	
	n	%	*n*	%	*n*	%
Fever	178	61.4%	140	54.5%	318	58.1%
Arthritis	127	43.8%	102	39.7%	229	41.9%
Chorea	33	11.4%	14	5.4%	47	8.6%
Subcutaneous nodule	2	0.7%	6	2.3%	8	1.4%
Erythema marginatum	2	0.7%	4	1.6%	6	1.1%
Pansystolic murmur	165	56.9%	146	56.8%	311	56.9%
Mid-diastolic murmur	67	23.1%	72	28.0%	139	25.4%
Early diastolic murmur	72	24.2%	89	34.6%	161	29.5%

Table 3. Combination of major manifestations of 547 patients with RF and RHD (1983-1992)

Major manifestation	1983 - 1987	(n=290)	1988-1992	(n=257)	Total	(n=547)
Arthritis only	44	15.2%	68	26.5%	112	20.5%
Carditis only	133	45.9%	144	44.4%	247	45.1%
Chorea only	13	4.5%	5	1.9%	18	3.3%
Subcutaneous nodule only	0	0	0			
Erythema marginatum only	0	0	0			
Arthritis + carditis	73	25.2%	51	19.9%	124	22.7%
Arthritis + chorea	8	2.8%	3	1.2%	11	2.0%
Arthritis + erythema marginatum	2	0.7%	4	1.6%	6	1.1%
Carditis + chorea	9	3.1%	3	1.2%	12	2.2%
Carditis + subcutaneous nodule	1	0.3%	6	2.3%	7	1.3%
Chorea + subcutaneous nodule	1	0.3%	1	0.44%	2	0.4%
Chorea + erythema marginatum	1	0.3%	0	1	1	0.2%
Arthritis + carditis + chorea	5	1.7%	2	0.8%	7	1.3%
Total	290		257		547	

nical manifestations in these 2 groups did not show significant difference. Chorea was slightly more frequently found in the first 5-year period (11.4% vs 5.4%), while subcutaneous nodule was found more frequently in the second 5-year period (0.7% vs 2.3%).

Table 3 depicts the combination of major symptoms in 547 patients with RF and RHD. Most of the RF and RHD cases showed signs of carditis (92.6%). Patients with carditis as the only symptom were found in 45.1%, arthritis only in 20.5%, and chorea only in 3.3% of the

Table 4. Abnormalities in laboratory findings of 547 patients with RF and RHD (1983-1992)

Laboratory	1983-1987 (n=290)		1988-1992 (n=257)		Total (n=547)	
Hemoglobin < 10 g/dl	55	(19.0%)	35	(13.6%)	90	(16.5%)
ESR > 20 mm/hr	175	(60.3%)	133	(51.8%)	308	(56.3%)
ASTO > 200 U	88	(30.3%)	74	(28.8%)	162	(29.6%)
PR interval > 0,16 sec	40	(13.8%)	33	(12.8%)	73	(13.3%)
CTR > 0,55	83	(28.6%)	68	(26.5%)	151	(27.6%)

cases. Neither subcutaneous nodule nor erythema marginatum was found alone, but was always accompanied by other major symptom(s). The most frequently found combinations were arthritis and carditis (22.7%), chorea and other major symptom (4.8%), while a combination of the three manifestations was only found in 1.3% of cases (Table 3).

There was no difference of the clinical manifestations between the 2 groups, except that arthritis as a single manifestation was more frequently found in the last 5-year period (26.5%) compared with patients in previous 5-year period (15,2%). It seems that RF with cardiac involvement occurred more frequently in the first 5 year period (80.3% vs 71.6%).

Laboratory findings in all patients are described in Table 4. Hb level of < 10 g/dl was found in 16.5% of cases, BSR > 20 mm/h in 56.3%, ASTO > 200 U in 29.6%, prolonged PR interval (> 0,16) in 13.3%, CTR > 0.55 in 27.6%. No significant difference was noted between the laboratory findings in the 2 groups.

Table 5 revealed that the diagnosis of RF and reactivation of RHD could be established by revised Jones criteria in 162 cases (29%). If modified Jone's criteria were used then the diagnosis was established in 474 cases (86.6%) while if it were based merely on clinical manifestations the diagnosis was established in 521 cases (95.2%).

Table 5. Diagnostic criteria of 547 patients with RF and RHD

Diagnostic criteria	n	(%)
Revised Jones criteria	162	(29,6%)
Modified Jones criteria	474	(86,6%)
Clinical judgement	521	(95,2%)
Inactive RHD	26	(4,8%)

Discussion

Our data indicated that the number of patients with RF and RHD visiting Pediatric Cardiology Outpatient Clinic, Department of Child Health, Medical School University of Indonesia, has been steady. There have been on the average 50 new

cases each year. It should be noted that the development of other hospitals in the city which are also caring for patients with RF and RHD does not reduce the number of patients visiting our hospital. This strongly suggests that RF and RHD cases in Jakarta do not show the tendency to decline.

This study showed that there was no change in the age distribution of patients during the last 10 years; most of patients in this series were between 8-12 years of age. Similarly, age and sex distribution in this series did not vary much from the previous reports from other developing countries which have high incidence of RF and high prevalence of RHD such as Iran, Bolivia, Sudan, and Zambia.[3,10]

How is the situation in other area in Indonesia? It is reasonable to assume that some big cities in Indonesia with similar environtment to Jakarta, such Surabaya and Semarang, face the same problems. This is one of the reasons why a periodic collaborative study involving all university hospitals in Indonesia is necessary.[11,12]

In general, there was no difference in the clinical manifestations and laboratory findings between the 2 groups, except that arthritis as a single manifestation was more frequently found in the last 5-year period. Although it seems that RF with cardiac involvement did occur a slightly more frequently in the first 5-year period, it can be stated that there was no significant change in the clinical manifestation of RF and RHD disease in Jakarta during the last 10 years. The major clinical manifestations in these patients were also similar to those in other developing countries,[3,11] namely most of the patients showed carditis, less than 50% with polyarthritis and only a small portion of them showed chorea, and very few presented with erythema marginatum or subcutaneous nodule. Erythema marginatum or subcutaneous nodule has never existed as a single manifestation; they always accompanied other major manifestations. It means that no single patient was diagnosed by the presence of erythema marginatum or subcuatenous nodule. Therefore, it is questionable whether including erythema marginatum or subcutaneous nodule as major signs is still relevant.

In developing countries active carditis is frequently found without history of RF and other clinical or laboratory manifestations.[3,11,12] Therefore many cases are not diagnosed as RF or reactive RHD, especially if we used revised Jone's criteria which need evidence of streptococcal infection. As seen in this study only one fourth of the cases (29.6%) were diagnosed as RF and reactive RHD based on revised Jones criteria, while the others (57,2%) were based on 2 major criteria or 1 major criterion plus 2 minor criteria without supporting evidence of previous streptococcal infection. Futhermore in 8,6% of cases, diagnosis of RF and reactive RHD could only be established based on clinical judgment, namely the presence of carditis or specific chorea.

The role of color Doppler echocardiography, which is routinely used at our Pediatric Department is quite important in detecting active carditis.[13] With periodic echocardiographic evaluations started as early as possible, valve leakage which can't be detected clinically, may be seen clearly, and any changes can be detected early so that appropriate management could be given to the patient without delay.[14-16] If echocardiographic and Doopler examinations are done routinely in every case with rheumatic fever and rheumatic heart disease, in general this will certainly provide a better outlook to the patients.

In a small number of patients who distinctly suffered from inactive RHD and did not meet the forementioned criteria, the management was just secondary prophylactic measures with the use of three to four weekly administration of intramuscular benzatine penicillin. Since the cardiac lesion was considered to be inactive, no antiinflammatory drug was needed. Such kind of treatment has also been reported by other researchers.[17]

In conclusion, although revised Jones criteria are still used as a 'gold standard' in the diagnosis of acute rheumatic fever in our Department, some modifications are also made. In many cases definite criteria of modifications are appropriate; however, in some patients no rigid criteria can be used, so that clinical judgement, which is mainly depends on the physician's experience, must be used. Care should be exercised, to avoid over-diagnosis, since it will give a definite consequence, i. e., unnecessary long-term treatment to the patient which will give further burden to the family.

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