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Research Article

Validity of Jones criteria for diagnosis of rheumatic fever and rheumatic heart disease

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ABSTRACT

Background: Acute rheumatic fever (ARF) remains one of the major causes of cardiovascular disease burden throughout the world particularly among the young. Availability of echocardiography / Doppler adds a different angle to the diagnosis of ARF and RHD in India.
Methods: The present one year cross-sectional study was conducted on patients of rheumatic fever (RF) and rheumatic heart disease (RHD) of age group of 5-15 years from in and around Kanpur, India to test the validity of Jones criteria.
Results: Out of 950 patients, majority had arthralgia (85%) and fever (82%). Nearly 23% had arthritis, 67% patients had an increased PR interval in ECG and increased ASO titre was seen in 93% of patients. Only1% was having subcutaneous nodules, 2% had Erythema marginatum and no patient had chorea while 24 % had a positive throat swab culture.
Conclusions: Jones criteria can be made more simple and easy to apply. The two diagnostic features of RF and RHD were arthritis/arthralgia and carditis. The other major as well as minor criteria had no importance for diagnosis of new cases.

Keywords: ARF, RHD

INTRODUCTION

Acute rheumatic fever (ARF) remains one of the major causes of cardiovascular disease burden throughout the world particularly among the young.[1] With respect to India, there have been reports that the incidence and prevalence studies have reported a decline in incidence of ARF and a decrease in prevalence of rheumatic heart disease (RHD). The introduction of echocardiography/Doppler and its easy availability in many urban and many rural settings adds a different angle to the diagnosis of ARF and RHD in India. [2]

For checking the validity of Jones criteria to diagnose rheumatic fever and rheumatic heart disease, a large number of cases of rheumatic fever and rheumatic heart disease have to be studied. This was only possible by covering the population in and around Kanpur, INDIA especially the villages of Kanpur district by telemedicine. By use of telemedicine, nearly all of the features of Jones criteria and new criteria could be evaluated. The present study was conducted on patients seeking health advice from in and around Kanpur to test the validity of Jones criteria for diagnosis of rheumatic fever and rheumatic heart disease.

METHODS

The present one year cross-sectional study was conducted on patients of rheumatic fever (RF) and rheumatic heart disease (RHD) of age group of 5-15 years from in and around Kanpur after taking permission from the institutional ethical committee. Informed consent was taken from all the study subjects.

Patients with any complaint related to Jones criteria for diagnosis of rheumatic fever and rheumatic heart disease (whether present or past history) were included in the study.

A structured performa was used to collect data regarding the socio-demographic variables, chief complaints, history of present and past illness, and relevant family history. General and systemic Examination was done. Investigations like Hb%, TLC, DLC, CBP, ESR, CRP, chest X-ray, throat Swab culture, echo Doppler and ECG were done. All cases were tested against the Jones criteria in order to assess its reliability as a diagnostic tool.

The following Jones criteria were used

Major criteria: Carditis**,** arthritis**,** chorea, sub- cutaneous nodules and erythema marginatum

Minor criteria: Fever, arthralgia, prolonged P-R interval and elevated acute phase reactants

Essential criteria: Supporting evidence of a recent group a streptococcal infection (e.g. positive throat culture or raped antigen detection test, and/or increasing streptococcal antibody titre).

Evidence of previous group: A streptococcal infection was considered positive if history revealed an episode of upper airways streptococcal infection within 5 weeks preceding the onset of symptoms or if ASO levels were greater than 320 IU/mL. SPSS was used for data analysis. Relevant tests were applied.

RESULTS

Table 1: Gender wise distribution of patients according to the criteria used for the diagnosis of RF and RHD.

Criteria	No. of Females	No. of males	Total
Arthritis	138	85	223
Arthralgia	503	308	811
Erythema margintaum	11	7	18
Subcutaneous nodules	4	7	11
Chorea	0	0	0
Fever	480	303	783
↑ RD interval	393	248	641
↑ ASO titre	538	341	879
Culture Positive	150	81	231

This study was done on 950 patients of rheumatic fever (RF) and rheumatic heart disease (RHD).

In our study there were 811 patients (85%) had arthralgia, 223 patients (23%) had arthritis, 783 patients (82%) had fever. Patients with increased PR interval in ECG were 641 (67%) and increased ASO titre was seen in 879 patients (93%). Only 11 patients (1%) were having subcutaneous nodules, 18 patients (2%) had erythema marginatum and no patient had chorea, throat swab culture was positive in 231 patients (24%). After clinical and laboratory evaluation for erythema marginatum, it was found that rashes were due to urticaria in 11 patients, 5 patients had drug eruption and in 2 patients were due to viral infections. Thus causes of subcutaneous nodules and erythema marginatum were non-rheumatic in our study, as in 5 patients due to parasitic infections, 3 patients due to lipoma and 3 patients were due to fibroma arthralgia was more common among the females (62.02%) as compared to males (37.98%) (Table 1).

Arthralgia was the most common symptom encountered, 88% in the age group of 15 years and 87% in the age group of 11-13 years (Table 2).

Table 2: Gender wise distribution of patients according to the criteria used for the diagnosis of RF and RHD.

Criteria	Age group (in years)				Total
	5-7	8-10	11-13	≤15	
Arthritis	27	52	92	186	223
Arthralgia	116	193	316	186	811
Erythema marginatum	5	3	5	5	18
Subcutaneous nodules	2	4	3	2	11
Chorea	0	0	0	0	0
Fever	128	193	294	148	783
↑ PR interval	99	155	261	126	641
↑ ASO titre	130	215	344	190	879
Culture Positive	37	62	81	51	231

DISCUSSION

The overall prevalence estimated to be about 1.5-2/1000 in all age groups, in India (total population about 1.3 billion) suggests that there are about 2.0 to 2.5 million patients of RHD in the country.

T. Duckett Jones proposed the Jones criteria; it was modified and later revised by the American Heart Association (AHA). This was reconfirmed in and AHA sponsored Work shop in 2002. No modification or change was suggested after the introduction of Doppler Echocardiography (DE) for the diagnosis of clinical and "subclinical carditis". When a clinical diagnosis by the clinician fails to show evidence of carditis, DE may pick up cases of "subclinical carditis". Moreover, previous recommendations did not include the Classification of

Recommendations (Class I, II a, II b, and III) and Levels of Evidence (Levels A, B and C). Recently, however, a scientific statement from the American Heart Association (AHA) has published an update to the Jones criteria including the use of DE in the diagnosis of ARF.[3]

This study found that the major criterion Erythema marginatum to be very rare as it was present only in 2% patients and the diagnosis of Erythema marginatum itself became a retrospective diagnosis after having examined and confirmed the cases to be rheumatic. It is difficult to recognize Erythema marginatum in our country due to darker complexion of the individuals. Another major criterion proposed by Jones is subcutaneous modules, which was again very rare in this study (1%). After clinical and laboratory evaluation, the causes of subcutaneous nodules were found to be non-rheumatic. Chorea was not present in any patient in this study, leaving only arthritis and carditis as major criteria. Arthralgia was found to be the commonest presentation (87%) in this study, what has been defined several years ago in modified Jones criteria as a minor criterion. It should be included in the major criteria.

Similar findings were reported by Lalchandani et al who analysed 200 patients of acute rheumatic carditis with regard to Jones criteria.[4] No patient had even one of the 3 major criteria such as Chorea, erythema maginaum, subcutaneous nodules and arthritis was present in 20% of patients.

Our findings are also comparable to the study by Ralph et al; where out of 35 patients with suspected Acute Rheumatic Fever, 49% had a diagnosis of definite Acute Rheumatic Fever and 20% had a non- rheumatic fever diagnosis.[5] The remaining 31% posed diagnostic difficulties because of mild symptoms that failed to fulfil Jones criteria (having unexplained arthralgia) or atypical features such as older age. Two patients whose illness initially failed to fulfil the Jones criteria, who were neither diagnosed with ARF nor commenced on secondary benzathine penicillin prophylaxis, were found on follow up to have definite and probable ARF, respectively. At least 29% of patients without prior recognized ARF/RHD had echocardiographic evidence of established RHD, indicating that previous episodes were missed.

The introduction of echocardiography/Doppler and its easy availability in many urban and mny rural settings adds a different angle to the diagnosis of ARF and RHD in India. In another study by Misra et al, 61 school going children with RHD were identified by ECHO evaluation giving a prevalence of 0.5/1000 children.[6]

Evengelisto et al also found the patients having subcutaneous nodules to remain a diagnostic challenge.[7] They reviewed the causes of nodules with emphasis on those associated with rheumatic disease and provided guidelines for nodules evaluation better characterizing the associated disease and diagnostic assessment.

It is difficult to diagnose ARF when carditis is the only manifestation of the disease particularly in a recurrence. When patient has sub clinical carditis the clinicians fail to detect clinically. Clinically apparent carditis is present but supportive minor criteria are not fulfilled. When previous cardiac status is unknown it is not possible to know in a new case whether the findings are due to acute carditis or it is recrudescence or it is established old case of RHD. In cases of polyarthralgia, which is a minor criterion, if the patient is neglected and not evaluated for ARF, they would go undiagnosed, and could end up with RHD, allegedly without any past history of ARF, as the patient and the parents would have long forgotten the joint pain.[8]

Therefore diagnosis of past RF is only possible when a patient gives a history of arthritis, arthralgia, chorea or has an established RHD. Hence, retrospective diagnosis or identification of past RF is often missed.

RF and RHD are so widely prevalent in India that a very long rope should be given as far as diagnostic criteria are concerned. Considering the higher incidence amongst Indians we cannot apply very stringent rules to our patients. Therefore, we strongly believe the diagnosis of RF requires a major modification in criteria so that a wide spectrum and a larger number of cases of RF may be included and timely treatment started.

CONCLUSION

Jones criteria can be made more simple and easy to apply. The two diagnostic features of RF and RHD were arthritis/arthralgia and carditis. The other major as well as minor criteria had no importance for diagnosis of new cases.

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