

SHORT COMMUNICATIONS

Traditional Jones Criteria: Limitation in the Diagnosis of Rheumatic Fever in Patients with Mitral Valve Repair

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ABSTRACT

The present study aims to determine the limitations of traditional Jones criteria during the first episode of acute rheumatic fever (ARF) at the initial referral hospital, in a cohort of patients below 18 years old who had undergone mitral valve repair in National Heart Institute (IJN) from 2011 to 2016. Carditis followed by fever and joint involvement were the most frequent manifestations at first diagnosis. Of the 50 patients, only seven (14%) fulfilled the traditional Jones criteria for the diagnosis of the first episode of ARF. When compulsory evidence of a previous group A Beta hemolytic streptococcus (GABHS) was disregarded, this figure rose to 54%. Therefore, strict adherence to Jones criteria with absolute documentation of GABHS will lead to underdiagnoses of ARF. The application of echocardiographic diagnostic criteria of rheumatic heart disease (RHD) needs to be emphasized to allow early diagnosis and administration of secondary prophylaxis to prevent progression to severe valvular disease.

Keywords: Jones criteria, Rheumatic fever, Rheumatic heart disease, Mitral valve repair

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INTRODUCTION

Rheumatic fever (RF) and rheumatic heart disease (RHD) are the major causes of cardiac morbidity and mortality in the developing world, although a declining trend is seen in some of these countries. Likewise in Malaysia, despite improved medical care and standard of living, RF remains a serious healthcare concern (1). RHD is the only chronic sequela to acute rheumatic fever (ARF), with life-long consequences. Jones criteria was established to aid the diagnosis of ARF in the mid 40's, at the height of its outbreak in the United States. The traditional Jones criteria were based on 5 major and 4 minor criteria with an absolute requirement for evidence (microbiologic and serologic) of recent Group A Beta haemolytic Streptococcus (GABHS) infection. The criteria have been revised, modified and updated since then.

Previous studies have looked at the reliability of Jones criteria in children who were followed up for ARF during the first episode of their disease. (2,3). However, ours is slightly different as we attempt to apply the criteria at their initial diagnosis of ARF at the referral hospital, on a

cohort of patients who had progressed into severe RHD requiring mitral valve repair. This study highlights the limitations of strict application of Jones criteria and the need to resort to other means of diagnosing RF especially in tandem with appropriate echocardiographic findings.

MATERIALS AND METHODS

This is a cross-sectional retrospective study of children below 18-years -old who underwent mitral valve repair at the National Heart Institute from 2011 to 2016. All except two patients were initially followed-up and subsequently referred from public hospitals in Malaysia. The study was approved by the Institutional Ethics Committee. A total of 50 children was identified. Data regarding the demographics, clinical presentation and laboratory findings for the initial presentation of ARF at the referral hospital were recorded in standardized proforma. Data were analyzed with the SPSS 22.0 Windows package.

RESULTS AND DISCUSSION

A large proportion of the patients in the study population was Malays (39/50; 78%). The remaining comprised of 8 indigenous patients from East Malaysia, 1 Indian and 2 foreigners. The mean age (IQR) at first diagnosis was 11.7 years (4.8-17 years). The mean duration interval from the time of the first diagnosis to the operation was

1.3 years (range 0.1 to 10.7 years). There were slightly more females (28/50, 56%) as compared to males (22/50, 44%).

Table I shows the frequency of clinical features based on Jones' criteria during their first episode of ARF at the referral hospital. Of the 50 patients who eventually required the mitral valve surgery, a majority (43/50, 86%) presented with carditis at the point of initial diagnosis; 14 of whom presented as the sole manifestation. This was followed by fever and articular involvement. Polyarthritis was less manifested as compared to arthralgia and none of the patients had aseptic monoarthritis. Sydenham chorea, subcutaneous nodules, and erythema marginatum were rarely seen at the first presentation. Table II shows the characteristic of valvular pathology prior to surgical intervention. One patient had combined incompetence and stenosis of both mitral and aortic valves. Two other patients had mitral stenosis with incompetence of both mitral and aortic valves.

Table I: Clinical Parameters according to Jones Criteria on First Diagnosis of Acute Rheumatoid Fever

Clinical Characteristics of Jones Criteria	Numbers (n)	Percentage (%)
Major		
Polyarthritis	9	18
Carditis	43	86
Sydenham chorea	2	4
Subcutaneous nodule	1	2
Erythema marginatum	1	2
Minor		
Fever	25	50
Arthralgia	12	24
ESR	14	28
Prolonged PR interval	1	2
Absolute requirement		
ASOT	10	20
Throat culture	0	0

Table II: The frequency of valvular abnormalities

Valvular abnormalities	Frequency
Mitral regurgitation	50 (100%)
Aortic incompetence	27 (54%)
Mitral stenosis	3 (6%)
Aortic stenosis	1 (2%)

In terms of evidence of GABHS pharyngitis, 10 patients were found to have a significant increase in ASO titer and none was found to have a previous history of sore throat nor a positive throat culture for GABHS. Only seven (14%) of the total 50 patients fulfilled the traditional Jones' criteria for diagnosis (with the inclusion of the absolute requirement of a GABHS infection) during the first presentation at the referral hospital. When

the compulsory evidence of a previous GABHS was disregarded, this figure rose to 54%.

The greater overall proportion of females as compared to males is a universal finding in most RHD populations; females occur 1.5 to 2 times more as compared to males (4). It is also interesting that the indigenous communities from East Malaysian (16%) make up quite a number of patients in this study population in contrast to the minority 0.5% proportion the community represents in the Malaysian population. This finding concurs with other countries in Australia and New Zealand, where the major burden of the disease is found in this selected underprivileged communities (5).

Articular and cardiac manifestations have been described as the most frequent features of RF (4,6). Our study showed that carditis was a more common feature with twice in frequency to articular manifestation. This could be explained by the nature of the study as it was confined to only RHD patients with valvular lesions for mitral repair. We could not ascertain the nature of the arthralgia, whether it was poly or monoarthralgia as this was a retrospective study. None of our patients presented with aseptic monoarthritis. In the revised Jones criteria (2015) for moderate- and high-risk population, monoarthritis and polyarthralgia are now included as major criteria in addition to polyarthritis (7). On the other hand, Sydenham's chorea, erythema marginatum, and subcutaneous nodules were rarely seen at the presentation of the first diagnosis, as concurred in several previous studies (4-6).

Our finding that of 43 patients with carditis, 14 (32%) were found to have carditis as the sole manifestation in RF, is also reported in previous studies (3,6). This further reinforced that these patients were detected symptomatic late in the course of the disease after several episodes of silent or undetected attacks of ARF and at a time when the titer level was low. In the revised American guidelines for the diagnosis of RF, echocardiography constitutes as part of the diagnostic criteria for confirmation of carditis in patients with suspected ARF, even if documented carditis is not present on diagnosis, particularly in high-risk populations.⁷ This subclinical carditis is considered as a major criterion of the revised Jones criteria for the diagnosis of ARF in the current Doppler echocardiography era.

RHD should be rigorously sought out, and echocardiographic screening for early detection of RHD is recommended especially in endemic areas as early secondary prophylaxis administration prevents progression to severe valvular disease hence reduce the morbidity and mortality related to complications of RHD. In 2015 a guideline on echocardiographic diagnostic criteria for RHD was developed to allow early identification of individuals with RHD without a clear history of ARF and hence allow enrollment into

secondary prophylaxis programs (7).

Our study further highlights that application of stringent traditional Jones criteria would lead to underdiagnosis of ARF at first presentation especially if the absolute requirement for microbiologic or serologic evidence of recent GABHS must be fulfilled. This finding is not surprising as many of these patients could not recall of a recent episode of a sore throat. Furthermore, their antibody levels have invariably normalized when treatment is sought especially six months after the infection. A common but major problem is that of obtaining serum for convalescent titers in children and thus lacking serial streptococcal titers to improve diagnosis (8).

Moreover, the microbiologic evidence on throat swab culture is also usually negative with most affected individual as the onset of ARF is 1 to 5 weeks after the infection. In addition, the gold standard of a positive immunologic response is not seen in 20% of these patients following the infection (9). It is recommended that serology for anti-DNase B antibody in addition to ASOT be performed in these patients in providing further evidence of a preceding streptococcal infection. Pyoderma has been implicated in a few cases of RF (10,11). Kaplan et. al has shown that response to anti-DNase B response was strong after an infection in either the skin or throat whereas poor for ASOT following a skin infection (10). Furthermore, the upper level of both serological markers for streptococcal infection remains a challenging issue in the diagnosis of ARF. Globally, the level varies as it is age-specific and dependent on endemicity of the regions. An inappropriately high upper limits of normal may have limited the detection of preceding streptococcal infection and hence potentially undercounts definite and probable ARF diagnoses (8). The historic Jones criteria were established in the early days for guidance in the diagnosis of ARF. Today the application of the traditional Jones criteria may not be appropriate to the population where ARF remains endemic, including Malaysia. It needs to be further emphasized that physicians be aware of the changing in variability of clinical presentation of RF in high-risk population. Subclinical carditis, polyarthralgia and monoarthritis are currently the clinical features incorporated to the revised diagnosis of Jones criteria. Suspected patients should be subjected for echocardiography for early detection of RF. A misdiagnosis of the disease would lead to a potentially devastating outcome in terms of a more severe valvular damage following repeated attacks of ARF.

CONCLUSION

Strict adherence to Jones criteria with absolute documentation of GABHS will lead to underdiagnoses of ARF. Therefore, the application of echocardiographic diagnostic criteria of RHD is important to allow early

diagnosis of cases where the traditional Jones criteria is not fulfilled or in cases without a clear history of ARF to warrant appropriate management to prevent morbidity and mortality related to complication of RHD.

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