Evaluation of the American Heart Association 2015 revised Jones criteria versus existing guidelines

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ABSTRACT
Objectives To compare the diagnostic yield of acute rheumatic fever (ARF) by the American Heart Association/ American College of Cardiology (AHA/ACC) 2015 revised Jones criteria with the WHO 2004 and Australian guidelines 2012.

Methods Retrospective observational study in 93 cases of suspected ARF admitted to the Division of Paediatric Cardiology between January 2012 and December 2014. WHO 2004, Australian guidelines and AHA/ACC 2015 Jones criteria were applied to assess definite and probable ARF.

Results Of the 93 cases, 50 were diagnosed as the first episode of ARF and 43 as a recurrence of the condition. Subclinical carditis was a predominant presentation (38%) in the first episode group (p<0.01) whereas in the recurrence group carditis (88%) was the main presentation (p<0.01). Among the joint manifestations, the majority of patients in both the first episode group and the recurrence group presented with arthritis. Of all the patients with suspected ARF (50), 34% of cases did not fulfill the standard Jones criteria 2004; however, 86% qualified as having ARF on applying the Australian and AHA/ACC 2015 criteria. Surprisingly in the recurrence group only 67% of the patients fulfilled AHA/ACC 2015 despite the modifications incorporated beyond WHO 2004; however, all the patients fulfilled the Australian guidelines either as definite (88.4%) or probable (11.6%). Inclusion of subclinical carditis, polyarthralgia and monoarthritis as major criteria influenced the diagnosis to definite ARF in 20%, 10% and 4% of patients, respectively.

Conclusions The clinical manifestations of ARF, comprising subclinical carditis and arthralgia, are possibly milder in the Indian population; hence, inclusion of subclinical carditis, polyarthralgia and monoarthritis as major criteria in the newer guidelines has improved the diagnostic yield of ARF. In the absence of a gold standard for the diagnosis of ARF, it is not possible to comment on sensitivity and specificity.

INTRODUCTION
The global distribution of acute rheumatic fever (ARF)/rheumatic heart disease (RHD) is essentially disproportionate. The incidence of ARF is high in low-income countries, in certain geographic regions, and among certain socioeconomic groups, whereas in other regions the disease has become rare. Also, the disease has become less severe, and certain manifestations that were common in the past (severe carditis and subcutaneous nodules) are now rare.1

The Jones criteria represent the clinical standard to establish the diagnosis and have undergone several revisions over the years due to the declining incidence of ARF in the West. These revisions increased the specificity but at the cost of sensitivity, thereby undermining the diagnostic abilities of physicians in low-income nations where the disease is endemic. Thus, a single set of diagnostic criteria may no longer be appropriate for all population groups which may result in overdiagnosis in low incidence populations and underdiagnosis in high-risk populations. Recurrences can be prevented by secondary penicillin prophylaxis thereby underlining the need to establish a correct diagnosis.

The idea of re-evaluating the diagnostic criteria, particularly in high-prevalence areas, gained momentum with the introduction of separate diagnostic guidelines by Australia and New Zealand.2,3 These changes were prompted by new findings of the disease following the application of echocardiographic techniques such as quantitative Doppler and colour flow mapping, underscoring the need to diagnose carditis even in the absence of overt clinical findings (‘subclinical carditis’).4,6 Also, publications from selective high-risk populations have indicted monoarticular arthritis and polyarthralgia as the major manifestation of arthritis.7

Recently, the American Heart Association/ American College of Cardiology (AHA/ACC) (2015) has come up with a revision of the Jones criteria which has incorporated major modifications for high-prevalence areas to improve the diagnostic yield.1 ARF and RHD occur at high rates in the Indian subcontinent, predominantly affect children, adolescents and young adults and are important causes of premature mortality. Until recently we have followed the Jones criteria (WHO 2004)8–9 and have possibly missed a large number of patients with ARF. Hence, it was planned to study the clinical characteristics of patients with first episode of ARF and recurrence visiting our tertiary care centre and to evaluate the appropriateness and strength of the new variable clinical manifestations of ARF included in AHA/ACC 2015 guidelines towards modifying the classification of suspected cases. We compared the diagnostic yield of ARF by the AHA/ACC 2015 guidelines over WHO 2004 and also with the Australian guidelines 2012.

PATIENTS AND METHODS
A retrospective observational study was conducted. The hospital records for all children with suspected first episode of ARF and recurrence in the paediatric age group (5–18 years) admitted between January 2012 and December 2014 to the Division
of Paediatric Cardiology, PGIMER, and associated Dr Ram Manohar Lohia Hospital New Delhi were reviewed. Those fulfilling either Jones criteria WHO 2004 or Australian guidelines 2012 were identified and included in the study. Files were reviewed meticulously for demographic data, clinical findings and laboratory reports on admission and during the hospital stay and those with incomplete information were removed. Children with congenital heart disease and other acquired heart diseases like cardiomypathies, Kawasaki disease, juvenile rheumatoid arthritis and other connective tissue disorders were excluded.

Carditis was diagnosed on account of any of the following: a precordial murmur, pericardial rub or effusion, cardiac failure and radiographic cardiomegaly. In people with known RHD, carditis consisted of a new, worsened, or changed murmur or cardiac failure in the presence of other evidence of active rheumatic inflammation.

Arthritis was defined as inflammation (swollen, red and warm) and restriction of movement of joints; monoarthritis was defined as involvement of a single joint, whereas in the presentation and the male:female ratio between the two groups.

Involvement of the joint without swelling. Arthralgia was defined as involvement of the joint without swelling. Echocardiograms were reviewed for valvular lesions particularly to determine whether pathological regurgitation (minor degrees of mitral or aortic valve regurgitation with colour Doppler manifestations) was present as described by the Australian guidelines 2012 and ACC/AHA 2015. Subclinical carditis was considered in those patients with no audible murmur but echocardiographic evidence of valvular regurgitation.

The patients were analysed with respect to first episode of ARF and recurrence according to the Jones criteria WHO 2004, Australian guidelines 2012 and ACC/AHA 2015 (table 1).

Statistical analysis was conducted using SPSS V.17.0 (Chicago, Illinois, USA). Results are expressed as mean±SD, numbers and percentages. Categorical variables were analysed using either the χ² or Fisher’s exact test. For all statistical tests, a value of p<0.05 was considered significant. Clearance was obtained from institutional ethical board.

RESULTS

Of the 93 cases of suspected ARF and recurrence, 50 were diagnosed as first episode of ARF and 43 as recurrence. The overall male:female ratio was 1.6:1 with a mean age of 11.3 years (range: 6–18). There was no difference between the mean age at presentation and the male:female ratio between the two groups. The clinical profile of each group is given in table 2.

Clinical carditis was the hallmark lesion (88%) in the recurrence group of patients, whereas in the first episode patients subclinical carditis was a significant presenting finding (38%, n=19, table 2). Six (6.4%) patients had no evidence of cardiac involvement even on echocardiography; four of them were from the first episode of ARF group.

Joint symptoms were present in the majority of patients (74%) with first episode of ARF while it was less common (37%) in patients with recurrence. Arthralgia (81%, n=13) was the characteristic form of joint involvement while arthritis (19%, n=3) occurred in a minority of patients with recurrence unlike in the cases with first episode of ARF where arthralgia (40%, n=20) and arthritis (34%, n=17) were almost equal in their distribution.

Chorea was present in 16% (n=8) of the cases and all of them had subclinical carditis except one. Joint involvement was uncommon (arthralgia n=2, arthritis n=1). Two patients (4.7%) presented with recurrence of chorea. They had subclinical carditis and no joint symptoms.

None of the patients had subcutaneous nodules or erythema marginatum. The median value of ESR and CRP, ASO and anti-DNase B was similar in both groups (table 3). In the first episode of ARF, ASO was positive in 60% of patients, anti-DNase B in 80% while ASO and anti-DNase B combined were positive in 92% of cases. Similar values were found in the recurrence cases.

On echocardiography, mitral regurgitation was the most common lesion followed by aortic regurgitation in both the groups. Isolated mitral regurgitation was the predominant single valvular lesion while mitral and aortic regurgitation together constituted the most common combination lesion. Combination valvular lesions were more common in the recurrence group than in the first episode ARF group. Mitral regurgitation was usually severe in the recurrence group while it was mild in the first episode of ARF group (table 4).

In the first episode of ARF group using WHO 2004 and AHA/ACC 2015 criteria, 66% and 86% of cases respectively, qualified as ARF. Using Australian guidelines, definite ARF was identified in 86% of the patients while seven (14%) patients were labelled as probable ARF (table 5).

In the recurrence group, 67% of cases fulfilled WHO 2004 but there was no improvement in the yield with the new AHA/ACC 2015 criterion. All patients fulfilled the Australian guidelines either as definite 88.4% or probable 11.6% (5).

Impact of subclinical carditis on the diagnosis of rheumatic fever

In the suspected first episode of ARF group, the incidence of subclinical carditis was 38% (n=19). However, in nine of these patients other major (chorea 7, polyarthritis 2) and/or minor criteria envisaged in WHO 2004 were present, completing the diagnosis of ARF, and the presence of subclinical carditis only reinforced the diagnosis.

However, by inclusion of subclinical carditis as a major criteria it influenced the diagnosis in 10 (20%) suspected cases who had either only one single major (polyarthralgia (five cases), polyarthritis (two cases) and monoarthritis (one case)) or insufficient minor criteria to be labelled as ARF (table 6).

In the recurrence group subclinical carditis (n=3) did not increase the number of cases with recurrence activity as these patients already met the WHO 2004 criteria for diagnosing ARF.

Impact of polyarthralgia and monoarthritis on the diagnosis of rheumatic fever

In the first episode of ARF, inclusion of monoarthritis as a major criteria enhanced the diagnosis of ARF in two (4%) cases
Second, arthralgia was the dominant joint manifestation in our patients (overall 35%, first episode 40% and recurrence 30%) in comparison to arthritis which was present in a smaller proportion of cases, markedly so in the recurrence group (overall 21%, first episode 34% and recurrence 19%). This is in contrast to the studies Carapetis and Currie from Australia and Wilson et al from New Zealand where arthritis was the predominant manifestation than arthralgia (arthralgia 78%/28% and 62%/27%, respectively). However, Carapetis and Currie also observed that arthralgia was more common in recurrence (46%) than first episode (28%).

Third, in the current study arthritis and polyarthritis were diagnosed more commonly in first episodes (34% and 24%, respectively) than recurrences (6.9% and 4%, respectively), but the frequency of these manifestations were quite lower than that reported by Carapetis and Currie (first episode: arthritis: 34% and polyarthritis 27%; recurrence: 6% and 3%, respectively).

The presentation and spectrum of disease had subtle variation between the first episode and recurrence of ARF. Subclinical carditis was a predominant presentation in the first episode

only as the remaining three cases already fulfilled WHO 2004 guidelines on account of the presence of criteria as envisaged for diagnosing ARF (table 6).

In our study, 32% (n=16) of patients presented with polyarthralgia as a minor manifestation with first episode of ARF and 18% (n=9) of patients could make it to the final diagnosis of ARF using WHO 2004 criteria. However, with the ACC/AHA 2015 and Australian guidelines 28% (n=14) of cases could qualify as ARF on account of the upgrading of polyarthralgia to a major criteria thus yielding 10% more cases.

However, this intervention did not increase the tally of patients being diagnosed with ‘activity’ in the recurrence group.

**DISCUSSION**

Clinical manifestations of rheumatic fever in the Indian subcontinent may be different from the West and other high-prevalence areas of the world. First, the joint manifestations were markedly less common (overall 59%, first episode ARF 72% and recurrence 37%) in our patients as compared with studies from aboriginal population of Australia and New Zealand (first episode ARF: 91% and 89%, respectively).7 11

| Table 1 Criteria used for defining ARF |
|----------------------------------------|
| **Criteria** | **WHO 2004** | **Australian guidelines** | **AHA/ACC 2015** |
| **Major manifestations** | ▶ Carditis | ▶ Carditis (including subclinical carditis) | ▶ Carditis (clinical and/or subclinical) |
| | ▶ Polyarthritis | ▶ Polyarthritis | ▶ Polyarthritis |
| | ▶ Monoarthritis | ▶ Monoarthritis | ▶ Polyarthritis |
| | ▶ Aseptic monoarthritis | ▶ Aseptic monoarthritis | ▶ Monoarthritis |
| | ▶ Polyarthritis | ▶ Polyarthritis | ▶ Polyarthritis |
| | ▶ Erythema marginatum | ▶ Erythema marginatum | ▶ Erythema marginatum |
| | ▶ Subcutaneous nodules | ▶ Subcutaneous nodules | ▶ Subcutaneous nodules |
| | ▶ Fever | ▶ Fever | ▶ Fever |
| | ▶ ESR ≥30 mm/h or CRP ≥30 mg/L | ▶ ESR ≥30 mm/h or CRP ≥30 mg/L | ▶ ESR ≥30 mm/h or CRP ≥30 mg/L |
| **Minor manifestations** | ▶ Prolonged PR interval | ▶ Prolonged PR interval | ▶ Prolonged PR interval |
| | ▶ Monoarthritis | ▶ Monoarthritis | ▶ Monoarthritis |
| | ▶ Erythema marginatum | ▶ Erythema marginatum | ▶ Erythema marginatum |
| | ▶ Chorea | ▶ Chorea | ▶ Chorea |
| | ▶ Arthralgia | ▶ Arthralgia | ▶ Arthralgia |
| | ▶ Febrile | ▶ Febrile | ▶ Febrile |
| | ▶ ESR or raised leucocyte count | ▶ ESR or raised leucocyte count | ▶ ESR or raised leucocyte count |
| | ▶ Polyarthralgia | ▶ Polyarthralgia | ▶ Polyarthralgia |
| | ▶ Elevated acute phase reactants | ▶ Elevated acute phase reactants | ▶ Elevated acute phase reactants |
| | ▶ Carditis (clinical and/or subclinical) | ▶ Carditis (clinical and/or subclinical) | ▶ Carditis (clinical and/or subclinical) |
| | ▶ Polyarthritis | ▶ Polyarthritis | ▶ Polyarthritis |
| | ▶ Monoarthritis | ▶ Monoarthritis | ▶ Monoarthritis |
| | ▶ Aseptic monoarthritis | ▶ Aseptic monoarthritis | ▶ Aseptic monoarthritis |
| | ▶ Polyarthritis | ▶ Polyarthritis | ▶ Polyarthritis |
| | ▶ Erythema marginatum | ▶ Erythema marginatum | ▶ Erythema marginatum |
| | ▶ Subcutaneous nodules | ▶ Subcutaneous nodules | ▶ Subcutaneous nodules |
| | ▶ Fever | ▶ Fever | ▶ Fever |

**ARF, acute rheumatic fever; ASO, anti-streptolysine O; CRP, C reactive protein; ESR, erythrocyte sedimentation rate; GAS, group A streptococci; PR, pulmonary regurgitation; RHD, rheumatic heart disease.**
Inclusion of subclinical carditis as a major criteria qualified 20% more cases as definite ARF which is slightly more than that in the studies by Wilson et al\textsuperscript{8,13} (8%) and Cann et al\textsuperscript{19} (9%) from Australia and New Zealand, respectively. This is the first study from India to show the influence of subclinical carditis as a major criterion for the diagnosis of ARF.

In the current AHA/ACC guidelines the ambit of arthritis has been increased by including polyarthralgia and monoarthritis as major criteria and monoarthralgia as a minor criterion for high-prevalence areas.

Several studies from Northern India reported much higher percentages of arthritis (52%–87%)\textsuperscript{10,11,15} than in our study. Indian literature is replete with arthralgia as a predominant manifestation of rheumatic fever (33%–90%)\textsuperscript{15-17} Roy\textsuperscript{16} even suggested in 1960 that arthralgia be included as a major manifestation in the Jones criteria. Until 1956, it was considered to be a major criterion for the diagnosis of ARF. We also found that upgrading of polyarthralgia to a major criterion influenced the diagnosis in 10% of cases.

### Table 2: Clinical characteristics of patients with first episode ARF and recurrence

|                      | First episode ARF | Recurrence | p Value |
|----------------------|-------------------|------------|---------|
| **Age**              | 10.90±2.32        | 11.79±2.47 |         |
| **Sex**              |                   |            |         |
| Female               | 16 (32%)          | 19 (44.2%) | 0.22    |
| Male                 | 34 (68%)          | 24 (55.8%) |         |
| **Religion**         |                   |            |         |
| Hindu                | 41 (82%)          | 38 (88.4%) | 0.39    |
| Muslim               | 9 (18%)           | 5 (11.6%)  |         |
| **Clinical carditis**|                   |            |         |
| Subclinical          | 19 (38%)          | 3 (7%)     | <0.01   |
| Clinical             | 27 (54%)          | 38 (88%)   | <0.01   |
| **Chorea**           | 8 (16%)           | 2 (4.7%)   | 0.07    |
| **Arthritis**        |                   |            |         |
| Monoarthritis        | 17 (34%)          | 3 (6.9%)   | 0.01    |
| Polyaarthritis       | 5 (29%)           | 1 (34%)    | 0.14    |
| **Polyarthritis**    | 12 (70%)          | 2 (66%)    | <0.01   |
| **Subcutaneous nodule** | 0          | 0           |         |
| **Erythema marginatum** | 0            | 0           |         |
| **Fever**            | 34 (68%)          | 28 (65%)   | 0.76    |
| **Arthralgia**       | 20 (40%)          | 13 (30.2%) | 0.32    |
| Mono arthralgia      | 4 (20%)           | 3 (23%)    | 0.852   |
| Polyaarthesis        | 16 (80%)          | 10 (77%)   | 0.35    |
| **High ESR**         | 34 (68%)          | 33 (76.7%) | 0.35    |
| **High CRP**         | 16 (32%)          | 24 (55.8%) | 0.02    |
| **ASO titre positive** | 30 (60%)      | 21 (48.8%) | 0.28    |
| Anti-DNase B positive | 40 (80%)       | 35 (81.4%) | 0.86    |

**ARF**, acute rheumatic fever; **ASO**, anti streptolysine O; **CRP**, C reactive protein; **ESR**, erythrocyte sedimentation rate.

### Table 3: Mean (IQR) of laboratory parameters

|                      | Mean (IQR) |
|----------------------|------------|
| ESR                  | 44.4 (20–57.25) |
| CRP                  | 13.50 (9.55–32.50) |
| ASO                  | 418.50 (189.75–611.75) |
| Anti-DNase B         | 667 (389.25–1050) |

**ASO**, anti streptolysine O; **CRP**, C reactive protein; **ESR**, erythrocyte sedimentation rate.

### Table 4: Valvular involvement in patients with first episode ARF and recurrence

| Valvular involvement | First episode ARF | Recurrence | p Value |
|----------------------|-------------------|------------|---------|
| MR                   |                   |            |         |
| Mild                 | 20                | 4          | <0.001  |
| Moderate             | 9                 | 7          | 0.83    |
| Severe               | 16                | 26         | 0.005   |
| AR                   |                   |            |         |
| Mild                 | 13                | 14         | 0.48    |
| Moderate             | 7                 | 4          | 0.48    |
| Severe               | 5                 | 6          | 0.56    |
| TR                   |                   |            |         |
| Mild                 | 8                 | 14         | 0.06    |
| Moderate             | 1                 | 6          | 0.03    |
| Severe               | 3                 | 4          | 0.55    |
| PR                   |                   |            |         |
| Mild                 | 2                 | 5          |         |
| Moderate             | 0                 | 3          | 0.16    |
| Severe               | 0                 | 0          | 0.09    |
| MS                   |                   |            |         |
| Mild                 | 0                 | 2          | 0.24    |
| Moderate             | 0                 | 2          | 0.24    |
| Severe               | 0                 | 8          | 0.001   |

**AR**, aortic regurgitation; **AS**, aortic stenosis; **MR**, mitral regurgitation; **MS**, mitral stenosis; **TR**, pulmonary regurgitation; **TR**, tricuspid regurgitation.
We found monoarthritis in 10% (n = 5) of the patients with suspected first episode of ARF which is similar to studies from India (4%–13%) \(^{10, 11}\) and several high-risk indigenous populations (16%–18%).\(^2\) Generally, monoarthritis without non-steroidal anti-inflammatory drug (NSAID) use has rarely been observed; in Australian guidelines it is now accepted as a major criterion. In India, over the counter NSAIDs are freely available, and is a cause of concern for both underdiagnosis as well as complicating the diagnosis of ARF. Monoarthritis as a major criterion influenced the diagnosis of definite ARF in a small number of our patients (4%) similar to that reported by Wilson et al.\(^{13}\) (8%) and Cann et al.\(^{12}\) (12%) thereby justifying its inclusion as a major criterion.

Anti-DNase B was positive in 80% of cases in both the groups whereas ASO was positive in only 60% and 48.8% of cases of first episode and recurrence, respectively. The immune response to Group A streptococci skin infection in the form of antistreptolysin O is weak compared with throat infection, whereas the anti-DNase B response after infection at either site is strong.\(^{18}\) It raises questions that possibly the throat infection complicating the diagnosis of ARF. Monoarthritis as a major criterion influenced the diagnosis of definite ARF in the high-prevalence areas.

The Australian guidelines (ARF (definite)): 86%). Cann et al\(^{11}\) found 20% more cases being diagnosed as definite ARF on modifying the 1992 AHA/ACC Jones criteria (including subclinical carditis and monoarthritides) as opposed to 16% by Wilson et al.\(^{13}\) on usage of New Zealand guidelines when compared with the American Heart Association 1992 Jones update criteria. Thus, the newer guidelines have also resulted in increased yield of ARF, justifying the inclusion of these clinical features as major and minor manifestations for high-prevalence areas.

The sensitivity and specificity of any set of new criteria for ARF cannot be determined as there is no laboratory or objective test for ARF.

The Australian and AHA/ACC 2015 guidelines have the same set of major and minor manifestations in their Jones criteria for diagnosing recurrence activity but the standard defining norms for labelling recurrences are slightly different.

The definition logistics for recurrence activity in the AHA/ACC 2015 and Australian guidelines are similar with respect to the need for two major criteria or three minor criteria towards the diagnosis. However, the differential requirement of one major and one minor criterion in Australian guidelines against one major and two minor criteria in AHA/ACC 2015 substantially increased the overall diagnostic yield with the Australian guidelines.

### Table 5 Comparative statement of ARF diagnosis (first episode and recurrence) WHO 2004 against newer guidelines

| Variables                  | WHO 2004 | Australian guidelines | AHA/ACC 2015 |
|----------------------------|----------|-----------------------|--------------|
| Total suspected cases (n=93) |          | % (n)                 | % (n)        |
| First episode ARF (50)     | 66 (33)  | Definite: 86 (43)     | 86 (43)      |
| Recurrence (43)            | 67 (29)  | Definite: 88.4 (38)   | 67 (29)      |

**ARF, acute rheumatic fever.**

### Table 6 Comparison of Australian and AHA 2015 guidelines with WHO 2004 guidelines: major criteria for ARF

| Variables                  | Australian and AHA 2015 guidelines | WHO 2004 |
|----------------------------|------------------------------------|----------|
| Total joint symptoms       | 33 (66)                            | 28 (56)  |
| Monoarthritis              | 5 (8)*                             | 0 by definition |
| Polyarthralgia             | 16 (32)†                           | 16 (32) minor criteria |
| Polyarthritis              | 12 (24)                            | 12 (24)  |
| Total carditis             | 46 (92)                            | 27 (54)  |
| Clinical carditis          | 27 (54)                            | 27 (54)  |
| Subclinical carditis       | 19 (38)†                           | 0 by definition |

*Influence on diagnosis to change to definite ARF in 2 (4%).
†Influence on diagnosis to change to definite ARF in 5 (10%).

Key messages

**What is already known about this subject?**

The Jones criteria are the standard for diagnosing acute rheumatic fever. It has undergone many revisions to improve its specificity based on the declining prevalence of the disease in the West. However, rheumatic fever is endemic in India, South East Asia and Africa. Hence, a single diagnostic criterion cannot be used worldwide. Keeping this in view the AHA/ACC has revised the Jones criteria in 2015 instituting modifications for the high-prevalence areas.

**What does this study add?**

The presentation of acute rheumatic fever (ARF) is different in our country from the West and other high-prevalence areas. Subclinical carditis and arthralgia were the predominant manifestations in our cohort. Applying the WHO 2004 guidelines led to underdiagnosis of disease in a significant proportion of cases. Inclusion of subclinical carditis, polyarthralgia and monoarthritis led to a greater detection rate of ARF and recurrence with the AHA/ACC 2015 and Australian guidelines. This is the first study to validate the use of the AHA/ACC guidelines 2015 in a high-prevalence area like India.

**How might this impact on clinical practice?**

Rheumatic heart disease is a cause of significant morbidity and health expense in our country. Recurrences which lead to further cardiac damage can be prevented by secondary benzathine prophylaxis. Hence, it is very important to prevent misdiagnosis and underdiagnosis. The AHA/ACC 1992 WHO 2004 update, which is more appropriate for Western countries, leads to underdiagnosis of cases in our country. We found that newer guidelines which have incorporated subclinical carditis and broader definitions of arthritis as major criteria lead to a modest increase in cases of rheumatic fever. Nevertheless, the findings have implications for the management of definite cases of acute rheumatic fever in India who receive benzathine penicillin G secondary penicillin prophylaxis and for the probable or possible cases who were until now being denied prophylaxis in the absence of clear-cut guidelines.
guidelines which is very well illustrated in our study. Of the 43 patients with suspected recurrence, only 67% fulfilled WHO 2004 and AHA/ACC 2015 while 88.4% patients had definite recurrence of rheumatic fever by Australian guidelines (p value 0.01, 95% CI 0.03 to 0.38).

A substantial number of patients in our study could not be classified as having first episode of ARF (14%, n=7) or recurrence (11.6%, n=5) despite being highly suspected (in the absence of any other plausible diagnosis) to be, due to the absence of either essential criteria or one major or minor criteria. These patients have been addressed by the Australian guidelines as probable rheumatic fever and advised secondary penicillin prophylaxis. ACC/AHA 2015 has also created a niche ‘possible rheumatic fever’, and advised penicillin prophylaxis for those patients not fulfilling the Jones criteria on account of non-availability of laboratory tests for acute phase reactants or for confirmation of recent streptococcal infection, poor documentation of clinical features or unreliable history.1

Although the ACC/AHA 2015 revision has made the Jones criteria more sensitive for the Indian subcontinent, the diagnostic dilemma in some of our patients with first episode ARF and recurrence remains to be addressed. Australian guidelines have a better case definition rate vis-a-vis WHO 2004 both for first episode ARF and recurrence and also it makes a caveat for inclusion of probable case definitions which is very much needed in the Indian context. However, we could be overdiagnosing some of these cases but in a high-prevalence country like India underdiagnosis is better than underdiagnosis. By subscribing to the still recent WHO 2004 criteria, 32% of our suspected cases of ARF and 33% of suspected recurrence cases would be refused treatment and prophylaxis, contributing significantly to the increase in the load of rheumatic valvular heart disease. However, the current revision of the Jones criteria respects the fact that there has to be a separate subset of recommendations to diagnose ARF in high-prevalence areas thereby addressing the issue of underdiagnosis and continued morbidity and mortality from chronic valvular heart disease.

Answers to the validity of the recently introduced guidelines can only be found through extensive studies on the high number of patients with rheumatic fever, which are hard to design in the developed countries due to the lower incidence of the disease. Experience and data from studies involving large series from low-income/middle-income countries shall provide answers in due course. The limitations of our study were the retrospective observational design and small sample size.

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