REVIEW ARTICLE

Acute rheumatic fever diagnosis and management: Review of the global implications of the new revised diagnostic criteria with a focus on Saudi Arabia

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Rheumatic fever (RF) is a common cause of acquired heart disease in children worldwide. It is a delayed, nonsuppurative, autoimmune phenomenon following pharyngitis, impetigo, or scarlet fever caused by group A β-hemolytic streptococcal (GAS) infection. RF diagnosis is clinical and based on revised Jones criteria. The first version of the criteria was developed by T. Duckett Jones in 1944, then subsequently revised by the American Heart Association (AHA) in 1992 and 2015. However, RF remains a diagnostic challenge for clinicians because of the lack of specific clinical or laboratory findings. As a result, it has been difficult for some time to maintain a balance between over- and underdiagnosis of RF cases. The Jones criteria were revised in 2015 by the AHA, and the main modifications were as follows: the population was subdivided into moderate- to high-risk and low risk; the concept of subclinical carditis was introduced; and monoarthritis was included as a feature of musculoskeletal inflammation in the moderate- to high-risk population. This review will highlight the major changes in the AHA 2015 revised Jones criteria for pediatricians and general practitioners.

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1. Introduction

Rheumatic fever (RF) is a common autoimmune disorder, particularly in developing countries. It is caused by group A β-hemolytic streptococcal (GAS) infection in genetically susceptible individuals [1]. The five cardinal manifestations of RF outlined by Dr. Jones and published in 1944 were carditis, arthritis, chorea, erythema marginatum, and subcutaneous nodules [2]. These features have been memorized by health professionals for several decades with little amendment over time. In Saudi Arabia, a few studies from different regions have demonstrated the mild nature of acute rheumatic fever (ARF) attacks, with frequent carditis ranging from 58% to 65% of patients with ARF [3–06]. Incidence of RF has declined remarkably in developed countries, but it has not yet been eliminated. In developing countries, it remains a major health challenge and results in lifelong, devastating sequelae.

In view of the high prevalence of the disease, especially in developing countries, it is important to emphasize the revision of the Jones criteria for pediatricians and general practitioners to improve diagnostic sensitivity and achieve earlier disease detection. This will consequently lead to better clinical outcomes for the disease.

Abbreviations

RF	Rheumatic fever
RHD	Rheumatic heart disease
GAS	Group A β-hemolytic streptococcal infection
AHA	American heart association
ASO	anti-streptolysin O titer
ESR	Erythrocyte Sedimentation Rate
CRP	C-reactive protein
NSAIDs	non-steroidal anti-inflammatory drugs
Penicillin V	Phenoxymethylpenicillin
Penicillin G	Benzylpenicillin

2. Methods and discussion

Peer-reviewed articles written in English and indexed in PubMed and EMBASE were screened by four investigators to review ARF diagnosis and management. The following keywords were used: acute rheumatic fever, rheumatic heart disease, Jones criteria, acute rheumatic fever management guidelines, and Saudi Arabia. Article selection was not restricted by year of publication.

3. Epidemiology

RF affects school-age children in the 5- to 14-years age range [7,8]. Although it occurs all over the world, its epidemiology largely varies. Nowadays, the annual incidence ranges from <0.5/100,000 in developed countries to >100/100,000 in developing countries [9]. The mean incidence rate of RF in the first attack is from five to 51 per 100,000 population [10]. The annual incidence rate is lowest in America and Western European countries (<10/100,000), and there is a relatively higher incidence rate in Eastern Europe, Asia, Australasia, and the Middle East (>10/100,000) [10]. Globally, it has been estimated that approximately 500,000 new RF cases occur annually and that about 230,000 people die each year from the disease [11]. The long-term sequela of acute RF is rheumatic heart disease (RHD), which is the most common cause of heart failure in poor populations [9].

Over the past few years, the global burden of RHD has dramatically reduced in developed countries [12]. However, RHD remains a significant issue in many developing countries, with approximately 1% of all school-age children showing signs of the disease [12]. Arab Gulf countries, Asia, Africa, the Pacific, and the indigenous populations of Australia and New Zealand are most commonly affected by RHD [12,13]. In Saudi Arabia, published data about the prevalence of RHD are limited. However, it is known that the percentage of children with RHD in Saudi Arabia is still higher than the global rate [14]. In Saudi Arabia, a study was conducted in patients with RF between 1994 and 2003 [15]. The study found that over a 10-year period, 96 children (mean age, 9 years) were diagnosed with RF. The annual incidence was 17 cases in 2 years (1994 and 1995), but only two in 2003, signifying a dramatic decrease in incidence in Saudi Arabia. The average incidence of RF in Saudi Arabia is eight cases annually [15]. Another study conducted on all regions in Saudi Arabia showed a high RF prevalence of 0.3 in 1000 and chronic RHD prevalence of 2.8 in 1000, with an overall rate of 3.1 in 1000 school-age children [4]. Other published studies on different regions in Saudi Arabia reported children older than 5 years as having a high prevalence rate [5,6]. RF could be the first attack or a relapse. For example, in the study of Abbag et al [6], it was found that 34 out of 40 (85%) patients had initial attacks and 12 (30%) were relapse cases, whereas Al-Eissa et al's [5] study reported 51 initial attacks out of 67 (76%) children and 22 (32%) relapse cases.

4. Rheumatic fever diagnostic criteria

The diagnosis of RF is based on Dr. Jones' criteria, which were recently revised (2015). The criteria include major and minor manifestations, and risk stratification has recently been applied to populations, dividing them into low risk and moderate- to high-risk [9]. The major diagnostic criteria are carditis, arthritis, chorea, erythema marginatum, and subcutaneous nodules, whereas the minor criteria are arthralgia, hyperpyrexia, high erythrocyte sedimentation rate (ESR), and/or high C-reactive protein (CRP), and prolonged PR interval [9]. To diagnose a patient with RF as a first episode of the disease, a confirmation of two major criteria or one major and two minor criteria is required, along with evidence of antecedent GAS infection. The diagnosis of subsequent episodes of RF requires either two major criteria, one major and two minor criteria, or three minor criteria [9]. The evidence of GAS infection is confirmed by one of the following: a positive throat culture for GAS; increasing trend anti-streptolysin O titer (ASO) readings rather than a single titer result; or a positive rapid group A streptococcal carbohydrate antigen test in a child who clinically suggests a high pretest probability of streptococcal pharyngitis [9].

4.1. Difference between low-risk and moderate- to high-risk populations

The differences between low-risk and moderate- to high-risk populations in the major criteria are as follows. Arthritis must be polyarthritis in the low risk population, whereas in the moderate- to high-risk population it can be polyarthritis, polyarthralgia, and/or monoarthritis. Meanwhile, the differences in the minor criteria are the following. In arthralgia, the number of affected joints is important in risk stratification. Polyarthralgia is considered a minor criterion in the low-risk population, whereas monoarthralgia is a minor criterion in the moderate- to high-risk population. Moreover, a $\geq$30 mm/h ESR is considered a minor criterion in the moderate- to high-risk population, but in the low-risk population it must be $\geq$60 mm/h. Regarding fever, 38.5 °C is considered febrile in the low-risk population, whereas in the moderate- to high-risk population, 38.0 °C is considered as fever [9]. The revised criteria are summarized in Table 1.

Universally, the most common presentations of RF major criteria are carditis (50–70%) and arthritis (35–66%) [12,16–20]. These are followed by chorea (10–30%), which predominates in females, and then subcutaneous nodules (<10%) and erythema marginatum (<6%), which are rare but highly specific manifestations of RF [9,18–20].

In Saudi Arabia, a study published in 2009 reported that the most common presentation of the RF major criteria is arthritis, which was present in 73%, followed by carditis in 17%, and chorea in 10% of cases [15]. None of the patients included in the study presented with erythema marginatum or subcutaneous nodules. For the minor criteria, an elevated ESR was most commonly seen in 94% of patients, followed by high grade fever in 83%, prolonged PR interval in 23%, and arthralgia in the absence of arthritis in 11% [15].

Table 1. Summary of major and minor criteria of RF in low-risk and moderate- to high-risk population.

Major criteria	
Low-risk population	Moderate- to high-risk population
- Carditis (clinical or subclinical)	- Carditis (clinical or subclinical)
- Arthritis (polyarthritis only)	- Arthritis (polyarthritis, polyarthralgia, and/or monoarthritis)
- Chorea	- Chorea
- Erythema marginatum	- Erythema marginatum
- Subcutaneous nodule	- Subcutaneous nodule
Minor criteria	
Low-risk population	Moderate- to high-risk population
- Polyarthralgia	- Polyarthralgia
- Fever (≥38.5 °C)	- Fever (≥38.0 °C)
- Elevation of ESR (≥60 mm in the 1st hour) and/or CRP ≥3 mg/dL	- Elevation of ESR (≥30 mm in the 1st hour) and/or CRP ≥3 mg/dL
- Prolonged PR interval, corrected for age (only when there is no carditis)	- Prolonged PR interval, corrected for age (only when there is no carditis)

CRP = C-reactive protein; ESR = erythrocyte sedimentation rate; RF = rheumatic fever.

5. Differences between the 1992 Jones criteria and American heart Association criteria 2015

There have been two substantial changes in the recently published 2015 American Heart Association (AHA) criteria compared to the 1992 Jones criteria [21]. One is that susceptible children are divided into two groups on the grounds of epidemiological variation regarding the risk of developing the disease. The reason for this change is that ARF incidence varies significantly from one country to another. Dividing the population into low and moderate- to high-risk would help prevent overdiagnosis in low-risk populations and prevent underdiagnosis in moderate- to high-risk populations [9]. The risk stratification depends on the incidence of RF in the area. Low-risk populations are shown in orange and moderate- to high-risk populations in purple on the world map shown in Fig. 1. Further details of each country are listed in Table 2. For example, children aged 5–14 years living in a community with an incidence of RF of <2/100,000/year, or children of any age where the prevalence of chronic rheumatic carditis is one or more/1000 per year are considered low risk (Class IIa, Level of Evidence C). Meanwhile, children living in areas with an incidence of two or more/100,000/year in children aged 5–14 years or a prevalence of chronic rheumatic carditis more than one/1000/year at any age are considered at a moderate to high risk of developing the disease (Class IIa, Level of Evidence C) [21].

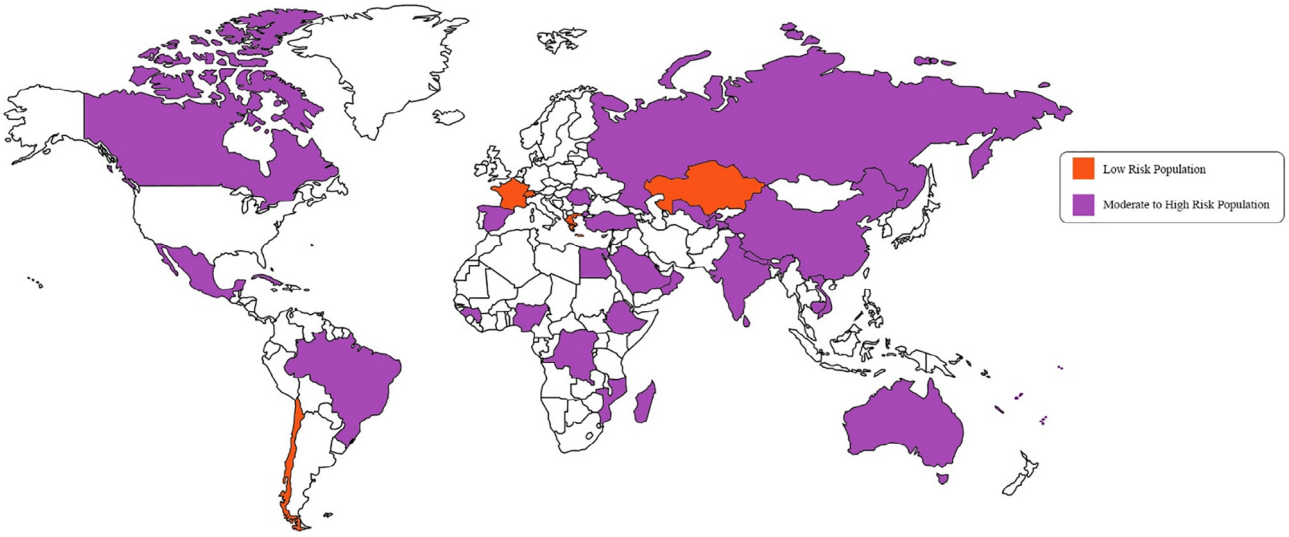

Figure 1. World map showing low- and moderate- to high-risk populations depending on the reported literature and the new American Heart Association criteria.

Table 2. Reported data on low and moderate- to high-risk areas.

Low-risk population	Moderate-to high-risk population		
Chile [22]	Australia [23,24]	India [25]	Samoa [26]
France [27]	Brazil [28]	Madagascar [29]	Saudi Arabia [30]
Greece [31]	Cambodia [12]	Mexico [32]	Spain [33]
Kazakhstan [12]	Canada [34]	Mozambique [12]	Sri Lanka [35]
Switzerland [36]	China [37]	Nepal [38]	Sub-Saharan Africa [39]
	Cuba [40]	New Caledonia [41]	Tajikistan [12]
	Democratic Republic of Congo [12]	Nigeria [42]	Tonga [43]
	Egypt [44]	Oman [45]	Turkey [46]
	Ethiopia [47]	Qatar [13]	Uzbekistan [12]
	Fiji [48]	Romania [12]	Vietnam [49]
	Guinea [12]	Russia [50]	

The diagnostic criteria for an initial RF episode in low-risk patients have not changed from the Jones criteria published in 1992. These state that the patient should have two major manifestations or one major plus two minor manifestations [9]. However, in the update published in 2015, polyarthralgia and monoarthritis are considered as major criteria in patients belonging to the moderate- to high-risk group. Moreover, in this risk group, monoarthralgia is considered a minor criterion [9]. The minor criteria stipulate an ESR $\geq$60 mm in the 1st hour in low-risk individuals but an ESR $\geq$30 mm/h in moderate- to high-risk patients. There are no other changes in the minor criteria, regardless of the risk stratification group [9].

Most importantly, regardless of the risk stratification, the latest update recommends using echocardiography with Doppler to diagnose carditis and subclinical carditis in all patients [9]. Echocardiography has a high sensitivity and is more reliable in diagnosing valvular involvement in ARF [51]. In addition, it is recommended to repeat echocardiography in case of uncertainty [9]. Carditis has been accepted globally as a major criterion [9]. However, the concept of subclinical carditis, which is defined as positive findings of mitral or aortic valvitis on an echocardiogram without heart murmurs or other clinical signs, has emerged as a major criterion [9]. The echocardiographic features of rheumatic carditis are focused on the aortic and mitral valves. The American College of Cardiology has described, in brief, mitral valve regurgitation detected in two or more views, jet length $\geq$2 cm, peak velocity >3 m/s, and pansystolic. It also described aortic valve regurgitation if detected in two or more views, jet length $\geq$1 cm, peak velocity >3 m/s, and pandiastolic. All four above-mentioned criteria must be met to diagnose mitral valve regurgitation and aortic valve regurgitation, respectively [9]. The detailed echocardiographic features of rheumatic carditis are shown in Table 3 [52].

In all patients diagnosed with rheumatic carditis, the mitral valve is usually involved, and the most common finding in color flow imaging is mitral regurgitation [53]. Mitral regurgitation in rheumatic carditis is associated with restriction of leaflet mobility and/or ventricular dilatation [53]. Rheumatic carditis does not result in congestive heart failure without hemodynamically significant valve lesions [53]. Moreover, it is observed in patients with rheumatic carditis that valve nodules could show echocardiographic equivalents of rheumatic verrucae [53]. Echocardiography is widely available worldwide, and numerous studies have reported echocardiography/Doppler evidence of mitral or aortic valve regurgitation in patients with ARF despite the absence of classic auscultatory findings. However, expert cardiac sonographers are not widely available, particularly in developing countries.

Arthritis in the 1992 Jones criteria is described as migratory polyarthritis in the larger joints, mainly the knees, ankles, wrists, and elbows, which tends to improve significantly with salicylates or nonsteroidal anti-inflammatory drugs (NSAIDs) [54]. However, in the 2015 update, the consideration

Table 3. Echocardiographic features of rheumatic carditis.

• Valvular regurgitation	• Leaflet	• Annular dilatation
– A regurgitant jet >1 cm in length	– Prolapse	• Chordal elongation/rupture
– A regurgitant jet in at least two planes	– Coaptation failure	• Increased echogenicity of subvalvular apparatus
– A mosaic color jet with a peak velocity >2.5 m/s	– Thickening (>4 mm)	• Pericardial effusion
– Jet persists throughout systole (mitral valve) and diastole (aortic valve)	– Reduced mobility	• Ventricular dilatation and dysfunction (almost always with significant regurgitation)
	– Nodules	

of monoarthritis and monoarthralgia as major and minor criteria, respectively, make a migratory description impractical. The second change in the 2015 Jones criteria is in the criteria used to diagnose RF relapse. For individuals who have previously had RF, the number of criteria required for diagnosis has been modified. In addition to the typical two major criteria or one major and two minor criteria for diagnosing RF, RF relapse can be diagnosed when three minor criteria are present in the patient [9].

5.1. Impact of AHA 2015 on clinical practice in Saudi Arabia

We believe the study conducted by Kumar et al [55] has provided good insight into how the AHA 2015 update on Jones criteria might impact clinical practice, especially in developing countries. The authors compared the updated AHA criteria to the World Health Organization (WHO) 2004 and Australian guidelines 2012 [52,56] and found that newer criteria that incorporated subclinical carditis and monoarthritis as major criteria led to a modest increase in the diagnosis of ARF cases. Until local data are available for Saudi Arabia, we think a similar situation is applicable to our population.

6. Treatment of ARF

There are two main goals in treating ARF; the first is to eliminate GAS infection via anti-streptococcal treatment and the second is to treat the clinical manifestations such as arthritis, carditis, and chorea [1,57].

6.1. Antimicrobial options for eradicating group A β-hemolytic streptococcus infection

(1) Phenoxymethylpenicillin (penicillin V) oral is the anti-streptococcal management of choice for streptococcal pharyngitis. The correct dose of phenoxymethylpenicillin (penicillin V) for patients weighing >27 kg is 500 mg two or three times a day for 10 days. Children weighing ≤27 kg should be given 250 mg two or three times a day for 10 days [1,57].
(2) Another choice in anti-streptococcal treatment is benzylpenicillin (penicillin G), which is used only in hospital facilities because it is given intramuscularly. It is given as an individual dose of 1.2 MIU for patients with a body weight >27 kg or age >6 years, and 600,000 IU for pediatric patients with a body weight <27 kg or age <6 years [1,57].
(3) Also, amoxicillin can be given orally for 10 days at a dose of 50 mg per kg with a maximum dose of 1 g every 8 hours. As per AHA guidelines, it is essential to start penicillin treatment because it could prevent initial episodes of RF up to the 9th day of disease onset [57].

6.2. Cases in which the patient is allergic to penicillin

In cases of known allergy to penicillin, a narrow-spectrum oral cephalosporin (cefadroxil or cefalexin) should be administered for 10 days. However, in immediate type hypersensitivity, macrolides such as clindamycin 20 mg/kg/d divided into three times daily (max 1.8 g/d) or clarithromycin 15 mg/kg/d divided into twice daily (max 250 mg, BID) should be administered orally for 10 days, except for azithromycin, which is given for 5 days at a dose of 12 mg/kg once daily (max 500 mg) [57].

One of the causative agents of ARF is GAS infection. A study was conducted by Bhardwaj et al. [58] in 2018 on 296 patients who had GAS infection and who showed antimicrobial resistance. Seventy-nine percent of these patients were resistant to tetracycline, 46% to erythromycin, and 9.5% to ciprofloxacin. Meanwhile, 30.6% showed resistance to both erythromycin and tetracycline [52]. In Saudi Arabia, in the most recent study conducted among 13,750 patients, 7.1% had GAS infection and 1% of them showed antimicrobial resistance to cancomycin [59].

6.3. Managing clinical manifestations of ARF

Arthritis:

- NSAIDs such as acetyl-salicylic acid, ibuprofen (30–40 mg/kg/d), and ketoprofen (1.5 mg/kg/d) are used in cases of arthritis with or without mild carditis [21]. Acetyl-salicylic acid (aspirin) is used for 2–3 weeks at a dose of 100 mg/kg/d, and once the symptoms completely improve, aspirin should be tapered down to 60–70 mg/kg/d. In case of aspirin sensitivity, naproxen at a dose of 10–20 mg/kg/d divided into twice daily can be used instead [60].

Carditis:

- In case of mild cardiac involvement, treatment is aspirin in a similar dose to arthritis mentioned above [60].
- In case of moderate to severe carditis, oral prednisone (2 mg/kg/d, max. 80 mg/d) is used and should be tapered down over 2–4 weeks by decreasing the dose to 2.5–5 mg every 3 days. Aspirin at a dose of 50–75 mg/kg/d should be administered concomitantly and should continue for 12 weeks [60].

Chorea:

- In rare cases in which mild chorea has manifested, a sedative such as diazepam or phenobarbitone is administered. If the patient does not improve, haloperidol (0.25–0.5 mg/kg/d) or valproic acid (15 mg/kg/d) should be administered for 12 weeks or 2–4 weeks after clinical improvement [60].

Table 4. Duration of prophylaxis based on the extent of cardiac involvement.

Category of patient based on cardiac involvement	Duration of prophylaxis
In children with no cardiac involvement	Prophylaxis should continue for 5 yr subsequent to the recent episode or until the age of 21 yr, whichever is longer
In children with preceding carditis and mild residual mitral regurgitation or valve lesion which resolved completely	Prophylaxis should continue for 10 yr subsequent to the recent episode or until the age of 21 yr, whichever is longer
In children with preceding carditis with moderate to severe valve damage	Prophylaxis should continue for 10 yr subsequent to the recent episode or until the age of 40 yr, whichever is longer
In children with relapses or high risk of infection	Prophylaxis must continue forever
In children with valve replacement	Prophylaxis must continue forever

Conspicuously, extrapyramidal syndrome could occur in high doses of haloperidol, and valproic acid should be given as a substitute [21].

6.4. How to prevent recurrent attacks of ARF

Relapsing episodes of RF could result in RHD or exacerbate the current cardiac condition. The best way to prevent severe RHD is by preventing relapsing episodes of GAS pharyngitis (secondary prevention). Moreover, a patient with a history of RF episodes will be at higher risk of another episode of RF. Benzathine penicillin G is administered intramuscularly as a longstanding prophylaxis every 28 days in this case. (In high-risk patients, it should be administered every 3 weeks.) In children weighing >27 kg, 1.2 million U is administered; however, in children weighing ≤27 kg, 600,000 U is administered. Penicillin V potassium 250 mg orally every 12 hours can be given as an alternative. The duration of the prophylaxis mainly depends on the extent of cardiac involvement; details are summarized in Table 4 [57,61].

6.5. Challenges in diagnosis and management of ARF in Saudi Arabia

A previous study conducted by Al Qurashi [15] showed that GAS was isolated in only 11% of Saudi children compared to 20–25% in the general literature [62,63], and this has been attributed to previous antibiotic use. Prevalence of brucellosis-related arthritis and post-streptococcal reactive arthritis make diagnosis of ARF-isolated arthritis very difficult. Al Qurashi [15] also described a high recurrence rate of ARF because of poor compliance with prophylaxis.

7. Conclusion

ARF is still prevalent in developing countries. The updated 2015 AHA criteria provide clinicians with more insight to classify the population into two major groups based on the risk stratification: low risk and moderate to high risk. The concept of subclinical carditis is becoming a widely accepted major criterion in all patients, regardless of their risk group. Primary prevention with eradication of GAS infection remains the most important step in the management of acute RF. However, the high index of suspicion helps in early detection and avoiding the devastating sequela of RF. Extrapolating data from developing countries with a similar ARF risk to Saudi Arabia and the application of newer criteria incorporating subclinical carditis and monoarthritis as major criteria might lead to an increase in the diagnosis of ARF cases. However, this is an area that should invite the interest of local researchers.

Conflicts of interest

The authors declare that they have no conflict of interests.

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