REVISTA BRASILEIRA DE REUMATOLOGIA

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Review article

Rheumatic fever: update on the Jones criteria according to the American Heart Association review – 2015

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ARTICLE INFO

Article history:
Received 5 June 2016
Accepted 20 December 2016
Available online 10 April 2017

Keywords:
Childhood
Carditis
Rheumatic fever
Jones criteria
Classification

ABSTRACT

Rheumatic fever is still currently a prevalent disease, especially in developing countries. Triggered by a Group A β-hemolytic *Streptococcus* infection, the disease may affect genetically predisposed patients. Rheumatic carditis is the most important of its clinical manifestations, which can generate incapacitating sequelae of great impact for the individual and for society. Currently, its diagnosis is made based on the Jones criteria, established in 1992 by the American Heart Association. In 2015, the AHA carried out a significant review of these criteria, with new diagnostic parameters and recommendations. In the present study, the authors perform a critical analysis of this new review, emphasizing the most relevant points for clinical practice.

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Febre reumática: atualização dos critérios de Jones à luz da revisão da *American Heart Association* – 2015

Palavras-chave:
Infância
Cardite
Febre reumática, Critérios de Jones
Classificação

RESUMO

A febre reumática ainda é uma doença prevalente nos tempos atuais, sobretudo nos países em desenvolvimento. Deflagrada por uma infecção pelo Streptococcus β-hemolítico do grupo A, pode afetar pacientes geneticamente predispostos. A cardite reumática é a mais importante das manifestações clínicas, podendo gerar sequelas incapacitantes e de grande impacto para o indivíduo e para a sociedade. Atualmente, seu diagnóstico é feito baseado nos Critérios de Jones, estabelecidos em 1992 pela American Heart Association (AHA). Em 2015, a AHA procedeu uma significativa revisão destes critérios, com novos parâmetros e recomendações diagnósticas. No presente estudo, os autores realizam uma análise crítica desta nova revisão, enfatizando os pontos de maior relevância para a prática clínica.

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http://dx.doi.org/10.1016/j.rbre.2017.03.001

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Introduction

Rheumatic fever (RF) is an inflammatory, systemic disease, triggered by the group A β-hemolytic *Streptococcus* infectious agent, which occurs in genetically predisposed individuals. The most relevant clinical manifestation of the disease consists of heart disorders and is characterized, in most part, by valvulitis, especially in the mitral and aortic valves, which can be chronic and cause disabling sequelae.

Note: the levels of evidence indicated throughout the text are those endorsed by the Guidelines of the Federal Medical Council (CFM) with the Brazilian Medical Association (AMB) and are listed in Table 1.

Diagnosis

Currently, the diagnosis of rheumatic fever is still based on a set of criteria, i.e., the Jones criteria, which have been reviewed at irregular intervals by the American medical associations – currently, by the American Heart Association (AHA). According to the latest review, published in 2015,[1] 2 major changes have been made in relation to the criteria established in 1992.[2]

The first one consisted in the stratification of susceptible individuals into 2 groups, based on epidemiological considerations regarding the risk to acquire the disease. A low-risk group is one in which the incidence of RF is less than 2/100,000 schoolchildren (aged 5–14 years) per year or that has a prevalence of chronic rheumatic carditis in any age group lower than or equal to 1/1000 per year. Children from communities that exhibit levels above these would have moderate-to-high risk for acquiring the disease.

The second important change was to include the possibility of using the Jones criteria to diagnose rheumatic fever relapses (until then its purpose was only to diagnose early episodes of the disease).

- At the diagnosis of initial RF outbreaks:

For low-risk individuals, the interpretation of the diagnostic criteria remains the same as for the 1992 review.[2] In these cases, an initial outbreak (1st episode) of rheumatic fever will be highly likely when, in the presence of evidence of a previous infection by group A β-hemolytic *Streptococcus*, 2 major criteria are met, or 1 major and 2 minor.

Moreover, regardless of the risk classification, the Echocardiography with Doppler findings suggestive of carditis – even if they are not accompanied by heart murmur or other clinical signs ("subclinical carditis"), are considered enough to contemplate 1 major sign of the criteria.

Concerning joint involvement, for individuals with moderate to high risk, polyarthralgia and monoarthritis (and not only polyarthritis, as in the past) also started to be considered as major signs of the criteria. Additionally, monoarthralgia was also considered as a minor sign for this risk group.

In both risk groups, there were no changes in the other minor signs of the criteria. Isolated Chorea, with an undefined etiology, remains sufficient for the diagnosis to be attained, even in the absence of other manifestations[1] (D).

- At the diagnosis of disease relapse (recurrent RF):

For patients that have already had the initial RF outbreak, the criteria remain the same as those listed above for low-risk and moderate-to-high risk populations – what changes is the minimum number of criteria to be met. For these individuals, in addition to meeting 2 major criteria or 1 major and 2 minor ones (as in the initial outbreak), one can also consider the possibility of meeting three minor criteria as a disease recurrence diagnosis, regardless of the risk group to which the patient belongs (Table 2).

There are some characteristics that are specific to the clinical manifestations of rheumatic fever, which, when identified, increase the positive predictive value of that finding. Although it cannot be said that there is a typical clinical picture of rheumatic fever, the most common forms of involvement are:

- Arthritis – Large joints such as knees, elbows, wrists, and ankles are the most affected ones. The pattern of involvement is migratory and can be fully resolved, most often leaving no sequelae. The response to nonsteroidal inflammatory drugs is excellent, with symptom remission within 48–72 h[1,3] (D)[4] (B).
- Carditis – the affected leaflet is the endocardium in more than 90% of the cases, which is expressed as mitral regurgitation, manifesting as an apical systolic murmur. In approximately 50% of the cases, it may be accompanied by basal diastolic murmur, due to aortic regurgitation. The concomitance of mitral and aortic regurgitation in a previously healthy patient is highly suggestive of rheumatic fever. Occasionally, myocarditis and pericarditis may be present. In the absence of valvulitis, these manifestations are rare in rheumatic fever[1] (D)[4] (B)[5] (D)[6] (B).
- Chorea – disordered, involuntary, abrupt movements of skeletal striated muscle groups. Complaints include stumbling during ambulation, slurred speech, dropping or throwing objects such as dishes, cups, notebooks, and bad calligraphy. It affects more females than males, in the adolescent age group. There is considerable emotional lability, easily alternating between crying and laughter. The differential diagnosis with systemic lupus erythematosus is necessary, especially in cases of difficult therapeutic control[1,3] (D).

Table 1 – Level of Scientific Evidence by Type of Study – "Oxford Centre for Evidence-Based Medicine" – last updated in May 2001.[19]

Level of recommendation	Strength of scientific evidence
A	Experimental or observational studies of better consistency
B	Experimental or observational studies of lesser consistency
C	Case reports, uncontrolled studies
D	Opinion without critical evaluation, based on consensuses, physiological studies or animal models

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