AHA Scientific Statement

Proceedings of the Jones Criteria Workshop

Patricia Ferrieri, MD, for the Jones Criteria Working Group*

On April 8 and 9, 2000, the members of the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the American Heart Association met with a group of international experts on rheumatic fever, rheumatic heart disease, and streptococcal infections to review guidelines for the diagnosis of acute rheumatic fever using the Jones criteria, including the 1992 statement on the "Jones Criteria Updated."[1] Formal presentations during the meeting included an historical review of the Jones criteria, beginning with the original publication in 1944[2] through several revisions of the guidelines, including the latest statement in 1992.[1] A review of the worldwide impact of rheumatic fever and rheumatic heart disease was presented, as well as an overview of the recent resurgence of rheumatic fever in certain regions of the United States. Extensive discussion focused on the diagnostic potential of Doppler echocardiography and its possible role in the diagnosis of acute rheumatic carditis. In addition, data on "silent" rheumatic valvular disease were presented, as well as clinical dilemmas in the diagnosis of rheumatic fever, with special reference to epidemiological challenges in both developing and developed countries.

The purpose of the workshop was to review the adequacy of the existing guidelines[1] for the diagnosis of first attacks of rheumatic fever and to assess whether contemporary diagnostic techniques, specifically two-dimensional and Doppler echocardiography, could be added to the criteria for the diagnosis of rheumatic carditis/valvulitis. Other issues that were addressed included the diagnosis of recurrences of rheumatic fever, the adequacy of one set of guidelines (the current Jones criteria) for the diagnosis of rheumatic fever both in developed countries and in developing countries that have a much higher prevalence of rheumatic fever, and monoarticular arthritis as a possible major criterion for the diagnosis of rheumatic fever.

The working group reaffirmed the validity of the major and minor Jones criteria and that these criteria should continue to be considered the accepted standard for diagnosis of initial attacks of acute rheumatic fever. The consensus opinion was that no new version of the Jones criteria was justified at this time. The 1992 revision of the Jones criteria was not intended to apply to recurrent attacks of acute rheumatic fever. However, the working group recommended that healthcare workers maintain a high index of suspicion of recurrent rheumatic fever in patients with a history of a previous attack who present with symptoms or signs consistent with acute rheumatic fever. Workshop members acknowledged that it might be particularly difficult to establish evidence of acute carditis in patients with preexisting rheumatic heart disease. Caution was suggested in using an isolated clinical finding (eg, monoarthritis, fever, arthralgia, elevation of acute-phase reactants) in this patient population as a sole criterion for diagnosis of acute rheumatic fever.[3]

The working group recognized that the prevalence of rheumatic fever varies widely throughout the world.[4] Ideally, a presumptive diagnosis of recurrent rheumatic fever may be made when a single major or several minor manifestations are present in a patient with a history of rheumatic fever or rheumatic heart disease, provided there is supporting evidence of a recent group A streptococcal infection. Strict adherence to the Jones criteria in areas of high prevalence may result in underdiagnosis and lack of treatment of patients with recurrent episodes of rheumatic fever. Accordingly, in areas where microbiological and immunologic tests may not be available, the workshop panel acknowledged the importance of clinical findings for the presumptive diagnosis of recurrent rheumatic fever. Thus, the epidemiological setting in which the diagnosis of rheumatic fever and its recurrence are considered is particularly important. For example, acute monoarticular arthritis among the indigenous people of Australia or the Maoris of New Zealand may be much more likely to represent rheumatic fever than monoarticular arthritis among a cohort population in the United States.[3]

Echocardiography has enhanced our understanding of the pathophysiology of acute[5–8] and chronic rheumatic heart disease[9,10] and is commonly used in the evaluation of patients with acute rheumatic fever. It has affirmed the dominant role of valvular pathology, rather than myocardial disease, in the clinical manifestations of rheumatic heart disease. Echocardiography is useful for confirming clinical findings and allows assessment of (1) the severity of valvular stenosis and

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*Members of the Jones Criteria Working Group are listed in the Appendix.

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regurgitation, (2) chamber size and ventricular function, and (3) the presence and size of pericardial effusions. Serial evaluations may help guide the management of patients with evolving rheumatic heart disease (ie, mitral stenosis and/or regurgitation with pulmonary hypertension, severe valvular regurgitation with marked ventricular dilatation, or ventricular dysfunction). In addition, echocardiography may exclude mitral and/or aortic regurgitation (and, therefore, rheumatic valvulitis) as the cause of a cardiac murmur in some patients with suspected acute rheumatic fever.[11]

Although echocardiography is of established value in the evaluation and management of acute and chronic rheumatic heart disease,[12–17] the use of Doppler findings for the diagnosis of acute rheumatic fever in patients who lack cardiac findings on physical examination remains controversial.[1,7] Clearly, Doppler is more sensitive than auscultation for detecting valvular regurgitation,[18] but physiological valvular regurgitation can be detected by Doppler in normal individuals.[19–22] Because pathological and physiological regurgitation are not distinct entities but are part of a continuum, the challenge is to distinguish physiological from pathological valvular regurgitation. Doppler criteria to distinguish physiological from pathological mitral regurgitation have been proposed by investigators who care for large numbers of patients with acute rheumatic fever.[23–26] At present, however, these criteria have not been universally accepted.

At least two other factors complicate the use of Doppler echocardiography for identifying pathological valvular regurgitation. First, the degree of valvular regurgitation considered "normal" increases with age.[22] Second, the frequency and degree with which Doppler-detected valvular regurgitation occurs in children with febrile illnesses unrelated to acute rheumatic fever, such as viral myocarditis, endocarditis, and systemic lupus erythematosus, are unknown. Finally, even if one could reliably distinguish pathological from physiological mitral regurgitation, the long-term prognostic significance of such a finding remains unknown.[27] It was the opinion of the working group that Doppler echocardiographic findings alone should not be classified as either a major or minor Jones criterion in the guidelines for the diagnosis of acute rheumatic fever at this time. There has been a subsequent expression of opposing opinions, which highlights the controversy about the role of echocardiography in the diagnosis of acute rheumatic fever,[18,27,28] including its important role in the "unlabeling" of individuals in whom echocardiography documents the absence of mitral regurgitation.[11,29]

Another area of discussion was the entity of poststreptococcal reactive arthritis (PSRA). The panelists reaffirmed the conclusion of the 1992 statement that the potential relationship of PSRA to acute rheumatic fever remain unresolved.[1] Features of PSRA include the development of arthritis 3 to 14 days after streptococcal pharyngitis; fever and a scarlatiniform rash are often present during the acute phase of pharyngitis but are absent by the time arthritis appears.[30] Typically, these patients have prolonged symptoms and protracted arthritis that fails to respond promptly to salicylate therapy, in contrast to the vast majority of patients with polyarthritis of acute rheumatic fever. Some of these patients fulfill the Jones criteria and should be diagnosed as having acute rheumatic fever. For those who do not fulfill the Jones criteria, the diagnosis of PSRA should be made only after other rheumatologic diagnoses (eg, Lymearthritis, rheumatoid arthritis) have been carefully excluded. The workshop did not address the potential need for antibiotic prophylaxis in these patients.

In summary, the workshop participants agreed that there are insufficient data to support a revision of the Jones criteria and reaffirmed the guidelines iterated in the 1992 statement.[1] In the absence of a "gold standard" for the diagnosis of rheumatic fever, no single specific laboratory test exists that is pathognomic of acute rheumatic fever or its recurrences. At present, Doppler echocardiography should be used as an adjunctive technique to confirm clinical findings and to evaluate chamber sizes, ventricular function, degree of valvular regurgitation, and morphological features of the valves. It should not be used as a major or minor criterion for establishing the diagnosis of carditis associated with acute rheumatic fever in the absence of clinical findings. It is hoped that future refinements of Doppler echocardiography and prospective studies of its predictive value will prompt reassessment of its role in the diagnosis of acute rheumatic fever. Data are not sufficiently compelling to designate monoarthritis as a criterion for diagnosis of acute rheumatic fever in the absence of other Jones criteria. However, it is acknowledged that this finding must be interpreted within the clinical and epidemiological setting of rheumatic fever prevalence in various populations.

Clinical research in several areas is needed, including epidemiological studies and determination of the prognostic implications of subclinical valvular regurgitation. In addition, research on basic pathogenetic mechanisms that result in rheumatic fever in "at-risk" individuals should continue. Future revisions of the Jones criteria statement will depend on data generated from these areas of research.

It is acknowledged that this is a consensus document from the participants of the workshop, but differences of opinion were expressed on some issues.

Appendix

The Jones Criteria Working Group included the following individuals.

Members of the American Heart Association Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease

Patricia Ferrieri, MD (Chair); Larry Baddour, MD; Ann Bolger, MD; Adnan Dajani, MD; Thomas Pallasch, DDS, MS; Lloyd Tani, MD; Walter Wilson, MD; Michael Gerber, MD; Matthew Levison, MD; Jed Jacobson, DDS, MS, MPH; Stanford T. Shulman, MD; Kathryn Taubert, PhD; Michael Gewitz, MD; and Jane Newburger, MD, MPH.

Invited Experts

Alan Bisno, MD; Edward L. Kaplan, MD; Milton Markowitz, MD; Jagat Narula, MD, PhD; and L. George Veasy, MD.

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