

*From the Guest Editor*

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## Rheumatic fever in the 21st century

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In this issue of the Journal, we publish three important papers concerned with rheumatic fever. So as to put these contributions in their world-wide perspective, I asked Cleonice Mota, a member of our Editorial Board, if she would prepare the “From the Editor” section for this issue. It is our pleasure to publish her thoughtful introduction, and we thank her for her continuing efforts on our behalf.

*Robert H. Anderson  
Editor-in-Chief*

**T**HE PROBLEMS RELATED TO RHEUMATIC FEVER remain an old challenge, but as we enter the 21st century, they demand a new approach, taking into particular consideration the difficulties in the diagnosis and treatment. The profile of this disease, involving multiple systems of organs, depends on the severity, association, and predominance of the manifestations, thus resulting in a wide range of presentations. Sadly, there are neither pathognomonic clinical features, nor specific laboratory tests. From a historical perspective, since the characterization of “the acute rheumatism” in the 17th century as being different from other rheumatisms, several authors independently described the clinical manifestations of rheumatic fever. Despite these efforts, it took two centuries for Cheadle<sup>1</sup> to identify them as part of the same disease. No modifications have subsequently been incorporated into this clinical profile. And, although the same author assigned the determination of a genetic pattern of susceptibility to the disease, the pathogenesis has still not been completely elucidated. At the beginning of a new millennium, therefore, rheumatic fever

remains worldwide as an unsolved problem of public health.

Considering the high incidence, severity, and the necessity of unifying the diagnosis, Jones, in 1944, assembled the clinical and laboratory manifestations to create a guideline for diagnosis. Later, these Jones criteria were modified, aiming at minimizing overdiagnosis. The three subsequent revisions have added more detailed information by introducing changes based on clinical observation, and have enhanced the specificity of the diagnosis by requiring supporting evidence for the antecedent streptococcal infection. As regards the incorporation of new technology to improve the accuracy of the diagnosis, however, few contributions have been registered. After a long period since the original criteria were proposed, the updated revision, published in 1992, highlighted the exceptions to the criteria, recognising that the risks of underdiagnosis would be higher facing the requirement of a strict adherence to the criteria in three conditions: indolent carditis, chorea as the only manifestation of the disease, and recurrences. The adequacy of the previous statements has also been recently assessed, and the conclusion was reached that, based on the available data, there was insufficient evidence to support a revision of the Jones criteria for first episodes, especially regarding the introduction of new clinical criteria and diagnostic techniques.<sup>2,3</sup>

Despite all the investment, the disease continues to challenge all those involved with its diagnosis and

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repercussions. Overdiagnosis can still present problems by means of the undesirable stigma, and exposure of patients to the rigors of an unnecessary prophylaxis. On the other hand, the difficulties in detecting any mild and unusual presentation could contribute to underdiagnosis, with all the undesirable implications in the prognosis.

Although it is a fact that, based on the results of applied and basic research, knowledge has been gathered, and considerable progress has been made, rheumatic fever is still the major cause of acquired disease in developing countries for those aged from 5 to 24 years. It is also one of the most frequent diseases requiring cardiac surgery in adults, thus having a great social and economic impact. In contrast, the disease has become rare in the developed areas. This decline has been attributed to the improvements in the standards of living and the environment, mainly related to a decrease in overcrowding and the access to medical care, including antibiotic therapy. Even in areas where rheumatic fever is still prevalent, there has been a decrease in its frequency and seriousness, resulting in a less characteristic presentation of its clinical manifestations. Based on the examination of the possible causes for the trends of modifications of streptococcal diseases, the changing pattern has been attributed to the presentation of decreasing rheumatogenic potential of group A streptococcal strains, the changing susceptibility of the human host, the widespread use of antimicrobial therapy, and improvement of the diagnostic abilities resulting in early and correct treatment. On the other hand, rheumatic fever has resurged in areas where it had almost disappeared, which has rekindled the interest in the disease. These outbreaks remain to be clearly explained, and should be viewed as a warning. Rheumatic fever has yet to be totally overpowered, and we should remain aware of the potential risks for its resurgence. As has been pointed out, it is clear that the control of factors related as the possible causes of the decline has not been enough to control the disease.<sup>4</sup>

It is a fact that there is not an isolated factor responsible for the epidemiological changes in either the disappearance or the reappearance of this intriguing disease. In this context, a set of questions emerges, to which current knowledge is insufficient to offer the complete answers and means to solve the diagnostic doubts. Fifty years after their introduction, the Jones criteria maintain their great usefulness and validity for the diagnosis in the classical forms of the disease. But there are still controversies about the role of echocardiography, and difficulties remain in the diagnosis and management of the mild forms, including recently introduced conditions such as subclinical valvitis, subclinical chronic valvar disease, and poststreptococcal reactive arthritis. Are their sequels

less severe or not? Do these valvar lesions have the same potential for evolution when compared with valvar lesions with auscultatory findings? If the available information is insufficient for the cardiac involvement diagnosed exclusively by cross-sectional and Doppler echocardiography to be considered as carditis, how should the patients be managed? Accuracy in the diagnosis is very important for proper prevention, and pre-existing valvar lesions can be made worse by recurrences. Thereby, should or should not the patients with isolated arthritis or pure chorea, and diagnosis of subclinical valvitis based on the current echocardiographic criteria, receive the same therapy in the acute phase, and the same scheme for prophylaxis, as those with clinical findings of cardiac involvement? Could the difficulties in identifying the acute phase of patients with rheumatic chronic cardiac disease also be attributed to a subclinical course of carditis? Should the potential risk of developing significant valvar lesions, and the necessity for surgery, be reduced by the identification of a subclinical valvar lesion in those patients? From these uncertainties, other questions emerge, such as the impact of actions on the different realities around the world regarding the economic aspects and epidemiological data. The answers to these questions will only be found through prospective investigations, the results of which will determine the most convenient approach. As emphasized by Kaplan<sup>5</sup> "clinicians and scientists there have both the opportunity and the obligation to further the understanding of this enigmatic disease, and then to implement these advances into practical techniques for improving the cardiovascular health of a significant proportion of children in the world today". Fortunately, a more intensive movement on this direction has now been registered, proved by the publication of three investigations in this issue of the *Cardiology in the Young*.<sup>6-8</sup> The recent data have shown a larger number of patients with subclinical valvitis, enhancing the quality of information. Additionally, the first publications regarding a longer follow-up seem to show a similar pattern of evolution found among patients with clinical manifestations of the cardiac involvement during the acute phase of rheumatic fever.<sup>6,7,9,10</sup> On account of the available information, although not recognized as complete, we should be cautious, and offer the same therapeutic approach for both groups of patients, considering the risks and implications on prognosis, mainly in those areas retaining a higher prevalence of rheumatic fever.

Faced with these dilemmas it is, therefore, essential that a renewed interest must be addressed to research in both the developed and developing world. Only in this way we will increase our understanding of this important disease, giving the answers for these new questions, as well as perhaps for the old ones that remain unanswered.

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