



Case Report

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Acute Rheumatic Fever and Rheumatic Carditis in Izmir



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Introduction

Acute rheumatic fever is a non-suppurative complication of *Streptococcus pyogenes* which only licks the joints, however bites the heart valves, particularly the mitral, aortic or both, rarely the tricuspid valve, resulting in valvular endothelial ulceration, collagen degeneration, neovascularization, interstitial calcification and fibrosis associated with lymphocytic infiltration [1]. Currently in Turkey, it is believed that there are tens of thousands of heart valve diseased adult patients half of whom are class III or higher according to New York Heart Association (NYHA) classification [2]. It is stated that the other half is in mild or moderate heart failure condition, ie. Class I or II, possibly due to previous interventions [3]. Currently, the incidence of rheumatic fever (RF) and rheumatic heart disease (RHD) is really low at certain parts of the globe compared to the incidence in Fiji, South Africa, New Zealand etc., and Turkey, with a high incidence following right after these countries [3]. Many countries have recognized the problem and tried to manage the disease so the diagnosis could be made earlier, using hand held echocardiography, as a screening test [4]. Some others have integrated using computer assisted auscultation in addition to two dimensional (2D) echocardiography, in an effort to improve the diagnostic methods. The reason behind such efforts has been to initiate secondary prophylaxis without delay, hoping to minimize the upcoming valvular damage. Once the valve is damaged, valvular mechanics of the beating heart damages the valve even further. Hence, the valve will require a suitable type of intervention, such as repair, balloon valvuloplasty and/or prosthetic valve replacement. In Turkey, the most frequent etiology of heart valve disease (HVD) is still of rheumatic origin [2]. The question is, is this disease preventable? Many publications support the fact that RF and RHD are preventable. For many years, the prevalence of the disease has been shown to decline dramatically within decades in certain populations at a certain place when certain precautions are carried out. Recently, the decline in RF was reported to be in association with the changes in M protein of Group A Streptococci, which is one of the main

portions of the microorganism responsible for the mechanism of antigenic mimicry within the host [5]. Certain strategies should be proposed in order to increase the possibility of earlier diagnosis and to keep track of the patient's compliance to secondary prophylaxis, as well as to infective endocarditis prophylaxis. Due to variations of the disease incidence between regions, even within the same country, it is our belief that many doctors had to make their clinical judgements as a personal initiative. One of the main reasons for this has been the inadequacy of the criteria used within the last few decades [6]. However, the disease is not only a problem of how healthcare is provided to the patient; but also mainly related to low economic income, crowded living conditions, low socioeconomic status overall. The aim is to discuss the most current recommendations in the diagnosis of definite and/or probable RF with special emphasis on subclinical carditis and preventive measures.

What is Novel Regarding Jones Criteria

Antecedent group a beta streptococcal infection

The Jones criteria were introduced in the diagnosis of RF more than half a century ago. Meanwhile, Jones criteria have been revised several times. One of the mainstays that has never changed until recently is the proof of *Streptococcus pyogenes* infection prior to the major and minor clinical manifestations. An antecedent Group A Beta Streptococcal pharyngitis is a must. Recently, in New Zealand, pyoderma with the same microorganism has been proposed to cause RF. This is a novel finding [7]. Secondly, a latent period has to evolve before the initiation of the manifestations. The evidence of Streptococcal infection are as in the following: recent history of scarlet fever, or rising titers of anti-streptolysin O (ASO) to a level of ≥ 276 IU, which begins rising at 2-3 weeks and starts declining by 6-8 weeks. The sensitivity and specificity of the laboratory data in the diagnosis of an antecedent streptococcal infection is increased to 92% when anti-deoxyribonuclease B (anti-DNAase B) titer elevation is determined besides raising levels of ASO. Anti-DNAase B titers are elevated usually around 3

months of the infection [6]. Currently, in our daily practice, serial evaluation of rising ASO titers are the optimal tool for the evidence of antecedent streptococcal throat infection.

Major and minor Jones criteria in the moderate to high risk versus low risk populations

The most recent major Jones criteria have been revised according to the risk of the population; ie. high and low risk populations. Low risk has been defined as RF incidence of <2/100,000 school age children; or, all age rheumatic heart disease prevalence of <1/1000 population per year. For the high risk populations, monoarthritis, instead of polyarthritis, and polyarthralgia (in the absence of arthritis) are now considered as major criteria [6]. On the other hand, polyarthritis only is considered as a major criterion in the low risk populations. We have been considering monoarthritis as a major criteria in our country [8]. For the high risk populations, polyarthralgia, low grade fever of 38 or more, sedimentation rate of 30 mm/hour or more, CRP: 3.0 mg/dl or more, PR prolongation in the absence of carditis have been considered as minor criteria. On the other hand, for the low risk populations, fever of 38.5 or more, sedimentation rate of 60 mm/hour or more, CRP>3.0 mg/dl or more, polyarthralgia (in the absence of arthritis), prolonged PR on ECG (in the absence of carditis) are considered as minor criteria [6]. Chorea, erythema marginatum and subcutaneous nodules are the other major criteria in both risk groups. In the presence of chorea, elevation of sedimentation rate and CRP should not be expected. Chorea by itself is diagnostic for RF and may be associated with carditis up to half of the patients with chorea.

Carditis

Pancarditis is the most important factor for morbidity and mortality in RF. The valvular endothelial surface is effected in up to 70-80% of the patients involving one or more cardiac valves. Myocardial and pericardial involvement may be seen. Myocarditis in RF is not associated with necrosis [1].

Troponin levels are not elevated in the pediatric RF patients [9]. Pericardial involvement is extremely rare in RF, unlike other rheumatic diseases, eg. systemic lupus erythematosus in which pericarditis may be the first manifestation of carditis.

Clinical scenario of rheumatic carditis is the presence of a systolic or diastolic murmur. Rapid heart rate at rest, fatigability and chest pain may accompany. Low grade fever, monoarthritis or polyarthralgia, elevated both sedimentation rate and CRP is present typically. Sometimes, 1st degree atrio-ventricular (AV) block or rarely higher degrees of AV block may be present on ECG. Recently, the presence of 2 major or 1 major and 2 minor or 3 minor criteria is considered diagnostic for recurrent disease.

Silent Carditis and 2d Echocardiography

As per se, monoarthritis was extrapolated from the major criteria not to overdiagnose RF once, Doppler echocardiographic

criteria set by World Health Organization (WHO) are considered too strict, or even may be considered inadequate [8]. Recently many countries have developed their own echocardiographic criteria in order not to underdiagnose the entity of silent carditis [7-10]. In my experience, in children more than 10 years or older, the observation of mitral valve prolapse especially at the tip of the valve and/or anterior valve thickening, whether associated with regurgitation or not, should be further questioned for family history. Recently, the presence of E148Q mutation on the second exon of MEFV gene was found to be associated with carditis in the Turkish population. In case of lack of family history, a personal history for epistaxis, frequent upper respiratory infections and a latent period or an event of self resolving discomfort or pain while moving medium to large sized joints should be obtained. History should be followed by a complete physical examination and supportive laboratory data. Echocardiography is the most important diagnostic tool for detection and prevention of sequela in any patient suspected of RF. Every child with acute rheumatic fever must undergo echocardiography at initial presentation followed by another at the end of the second week. Morphological changes such as mitral annular dilation, chordal elongation, anterior mitral leaflet prolapse just at the tip of the valve, nodularity of the leaflet tips may not be evident on the first echocardiographic examination. Therefore a second exam preferably at the end of the second week of the disease is recommended. Serial echocardiographic follow up is mandatory in the presence of valvular regurgitation and presence of any of the other changes. Among the echocardiographic and clinical parameters, predictors for the development of chronic RHD were initial big left ventricular end diastolic dimension and an audible murmur at the first presentation [11].

Adult cardiology uses the following echocardiographic criteria in the diagnosis and management of chronic valvulitis: right atrial and coronary sinus volumes, mitral valve area, ejection fraction, vena contracta, effective orifice area, etc. However in the pediatric age group, the presence of such criteria would be extraordinarily rare. Many of the pediatric patients fall into the category of, at risk of developing mitral regurgitation. Before progressive mitral regurgitation develops, it is important to define which patients are to be administered secondary prophylaxis. Regarding this, the most recent guidelines from AHA/ACC have introduced the concept of definite RF/probable RF in relevance to the administration of secondary prophylaxis. In case of probable RF, secondary prophylaxis is recommended for at least one year [6]. Such an approach is satisfactory, however, may be inadequate in some cases where the expert has to put his/her initiative regarding whether to continue or discontinue the prophylaxis. The weighted pooled prevalence of silent carditis has been published as 17%. However, without appropriate management, the disease is known to progress over time [6]. If compliance to secondary prophylaxis is established 80% of mild mitral regurgitation is known to diminish within 5 years [11]. Within 3 years of follow-up, mitral

regurgitation diminished in patients with good compliance to secondary prophylaxis [11]. Although secondary prophylaxis with Penicillin G is inadequate during the third week, a 3 weekly regimen is satisfactory [12]. We recommend a 2 weekly Penicillin G prophylaxis during the 1st 6 months period following an initial acute attack and to be continued life long every 3 weeks.

More on Prevention

In the etiopathogenesis, viral infections have never been proven, however Coxsackie B, enterovirus, Hepatitis B particles were found to a greater extent in the myocardium of the patients with rheumatic carditis compared to normal population. The Australian RF Group recommends influenza vaccine for patients on ASA [8]. In our practice, influenza vaccine and infective endocarditis prophylaxis is recommended to all rheumatic carditis patients with valvar insufficiency in the long term follow-up.

In conclusion, rheumatic carditis continues to be a health issue particularly in crowded, low income parts of the World. The prognostic importance of early diagnosis and treatment of silent carditis using echocardiography and the need for initiation of a policy to increase the awareness to the disease to minimize the extent of the disease worldwide is emphasized.

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