Update on diagnosis of acute rheumatic fever: 2015 Jones criteria

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Abstract

In the final Jones criteria, different diagnostic criteria were established for the diagnosis of acute rheumatic fever for low risk and moderate-high risk populations. Turkey was found to be compatible with moderate-high risk populations as a result of regional screenings performed in terms of acute rheumatic fever and rheumatic heart disease. The changes in the diagnostic criteria for low-risk populations include subclinical carditis found on echocardiogram as a major criterion in addition to carditis found clinically and a body temperature of 38.5°C and above as a minor criterion. In moderate-high risk populations including Turkey, subclinical carditis found on echocardiogram in addition to clinical carditis is used as a major criterion as a new amendment. In addition, aseptic monoarthritis and polyarthralgia are used as major criteria in addition to migratory arthritis and monoarthritis is used as a minor criterion among joint findings. However, differentiation of subclinical carditis from physiological valve regurgitation found in healthy individuals and exclusion of other diseases involving joints when aseptic monoarthritis and polyarthralgia are used as major criteria are very important. In addition, a body temperature of 38°C and above and an erythrocyte sedimentation rate of 30 mm/h and above have been accepted as minor criteria. The diagnostic criteria for the first attack have not been changed; three minor findings have been accepted in presence of previous sterptococcal infection in addition to the old cirteria for recurrent attacks. In the final Jones criteria, it has been recommended that patients who do not fully meet the diagnostic criteria of acute rheumatic fever should be treated as acute rheumatic fever if another diagnosis is not considered and should be followed up with benzathine penicilin prophylaxis for 12 months. It has been decided that these patients be evaluated 12 months later and a decision for continuation or discontinuation of prophylaxis should be made. In countries where the disease is prevalent, it is very important for physicians to make an accurate diagnosis of acute rheumatic fever with their own logic and assessment in addition to the criteria proposed. (Turk Pediatri Ars 2016; 51: 1-7)

Keywords: Acute rheumatic fever, echocardiography, Jones criteria, rheumatic heart disease, subclinical carditis

Introduction

The clinical picture of acute rheumatic fever (ARF) has been known since 1500s. In 1800s, the relation between ARF and the heart was described as “from tonsillitis to carditis”. This condition was described as “ARF licks the joints and bites the heart” by Laseque in 1884. As in other conditions which are difficult to diagnose, Jones criteria were established for the diagnosis of ARF in 1944 and these criteria were updated in 1965, 1984 and 1992 (1). While the incidence of ARF was decreased with the measures taken in Europe and North America, it has continued to be a serious public health problem in developing countries (2, 3). Acute rheumatic fever is the most common cause of acquired heart disease in children and young adults in many areas of the world and especially in developing countries. At least 15 600 000 patients with rheumatic heart disease live in the world (4). Each year 500 000 new ARF cases are seen. About 280 000 of these acquire rheumatic heart disease and 233 000 individuals are lost due to ARF or rheumatic heart disease yearly.

Updates performed in time after the first description of Jones criteria increased the specificity of the diagnostic criteria, but decreased the sensitivity. In areas where ARF is endemic or epidemic, the diagnostic criteria are not sensitive enough. Therefore, the number of patients with rheumatic heart disease increase, because the diagnosis is missed and secondary prophylactic treatment is not initiated. The World Health Organization published a guideline for the diagnosis of ARF in 2002, recommended the Jones criteria established in 1992 for the diagnosis in the first attack and attenuated the conditions for the diagnosis of recurrent attacks (5). By the time, subclinical (silent) carditis which is detected by echocardiography in absence of clinical findings came to the fore with the availability and widespread use of echocardiography in the world in the last 20 years (6-8). In the 1992 Jones criteria and 2002 World Health...
Organization recommendations, subclinical carditis which is detected by echocardiography is not included as a diagnostic criterion. At the present time, access to healthcare facilities has become easy for patients. Patients with arthritis present to healthcare facilities at the time of the first joint involvement. In addition, use of non-steroid antinflammatory agents including ibuprofen as analgesic and antipyretic changes the clinical course of arthritis and makes the diagnosis difficult. Considering all these factors, amendments have been made to increase the sensitivity of the diagnostic criteria in countries where the incidence of ARF is high.

The Australian Heart Association established their own guideline in 2006 and the New Zeland Heart Association established their own guideline in 2008 and these guidelines were updated in 2012 (9, 10). Since ARF is observed commonly in the natives living in Australia and New Zealand and rarely in the others, the recommendations were established for different groups as moderate-high-risk and low-risk populations. In 2015, the Jones criteria were also updated and different diagnostic criteria were established for low-risk and moderate- and high-risk populations similar to the guidelines of the Australian and New Zealand Heart Associations (11).

The populations with an ARF incidence of ≤2/100 000 in school age children and a rheumatic heart disease prevalence of ≤1/1 000 at all ages were defined as low-risk populations and the others were defined as high-risk populations. In addition, it was recommended that the diagnostic criteria used in moderate and high risk populations be applied in populations for which reliable epidemiological studies are lacking.

No study investigating the incidence of ARF throughout Turkey has been conducted and the studies performed reflect local data. In a study conducted in primary school children around Ankara, the annual incidence of ARF was found to be 56.6/100 000 between 1970 and 1973, while it was found to be 36.7/100 000 15 years later (12). The incidence of ARF increased from the mid 1980s in Turkey as throughout the world. In a study in which ARF patients were evaluated in a period of 30 years between 1980 and 2000 in Ankara, the incidence was calculated to be 37/100 000 between 1980 and 1989, 60/100 000 between 1990 and 1999 and 21/100 000 between 2000 and 2009 (13).

The prevalence of rheumatic heart disease is 5.7/1 000 in Africa in the South Sahara Desert, 3.5/1 000 in the natives in Australia, New Zeland and Pacific, 2.2/1 000 in Middle-South Asia and below 0.5/1 000 in developed countries (14). There is no new study conducted to determine the prevalence of rheumatic heart disease in Turkey. In a regional study conducted in a low socio-economical group in 1988, the prevalence was found to be 5.6/1 000 (15). According to the final Jones criteria, Turkey is compatible with moderate and high risk populations as a result of regional screenings performed in terms of ARF and rheumatic heart disease.

According to the final Jones criteria, subclinical carditis found on echocardiogram in addition to clinical carditis is used as a major criterion and a body temperature of 38°C and above is used as a minor criterion in low-risk populations (Table 1) (11). In moderate- and high-risk populations including Turkey, subclinical carditis found on echocardiogram in addition to clinical carditis is used as a major criterion as a new amendment. Additionally, aseptic monoarthritis and polyarthritis are used as major criteria in addition to migratory arthritis among joint findings and monoarthralgia is used as a minor criterion. However, differentiation of subclinical carditis from physiological valve failures found in healthy individuals and exclusion of other conditions involving joints when aseptic monoarthritis and polyarthralgia are used as major criteria are very important for an accurate diagnosis. In addition, a body temperature of 38°C and above and an erythrocyte sedimentation rate (ESR) of 30 mm/h and above are accepted to be minor criteria in moderate and high risk populations including Turkey. As in the previous Jones criteria, joint findings are not used as minor criteria when they are used as major criteria in the new Jones criteria. In individuals who have had carditis, PR prolongation on electrocardiogram (ECG) is not accepted as a minor criterion. In the final Jones criteria, it has been recommended that patients who do not fully meet the diagnostic criteria of ARF should be treated as ARF if another diagnosis is not considered and should be followed up with benzathine penicilin prophylaxis for 12 months. It has been decided that these patients be evaluated 12 months later and a decision for continuation or discontinuation of prophylaxis should be made.

The diagnostic criteria of ARF are valid for the acute period of the disease and patients who are compatible with these criteria are diagnosed with ARF with a high probability. These criteria are not used for the diagnosis of rheumatic heart disease and for evaluation of the prognosis.

The recurrence risk is higher in patients who have had ARF compared to the normal population. In addition, the risk of heart failure and mortality is higher in recurrent attacks compared to the first attack. Considering these conditions, the criteria with a high sensitivity rather than a high specificity should be used in the di-
agnosis of recurrent attacks. In the final Jones criteria, it was reported that recurrent attacks can be diagnosed with two major criteria or one major criterion and two minor or three minor criteria in presence of evidence of preceding streptococcal infection. However, it should be kept in mind that upper respiratory tract infection endemies related with group A streptoccoci may be present and the antistreptolysine O (ASO) level may be high in the population when making a diagnosis of recurrent attack in areas where the incidence of ARA is high. In countries where the disease is prevalent, physicians have a great role in terms of an accurate diagnosis. It is very important that physicians make an accurate diagnosis in ARF patients with their own logic and assessments in addition to using the diagnostic criteria proposed for ARF.

Major criteria
Arthritis: Arthritis is the most common sign observed in ARF and the least specific one. It is observed in older patients. In 1992 Jones criteria, migratory polyarthritis was used as a major criterion (1). The disease frequently involves large joints unilaterally. It frequently involves the knee, elbow, wrist and ankle. Although the hip and shoulder joints are large joints, they are involved rarely. Arthritis is migratory and passes from one joint to another. Arthritis in each joint lasts shorter than two weeks. Migratory arthritis observed in ARF generally resolves spontaneously in a few days or weeks (in 4 weeks at the latest) even if it is not treated. Arthritis of ARF responds very well to treatment with salicylates. In case of arthritis lasting longer than 48 hours under salicylate treatment at an appropriate dose, another diagnosis should be considered.

In the final Jones criteria, only migratory polyarthritis was considered a major criterion again in low-risk populations, whereas migratory polyarthritis, aseptic monoarthritis or polyarthralgia were considered major criteria in moderate- and high-risk populations including our country (11). In studies conducted in countries where the disease occurs with a high frequency, aseptic monoarthritis has been reported up to a rate of 17% in patients with ARF (16). Polyarthralgia is a nonspecific finding which is observed in many rheumatoid diseases. Polyarthralgia was accepted to be a major criterion according to the Jones criteria until 1956. However, it was excluded from the major criteria with updates of the Jones criteria, because it caused to misdiagnosis in low-risk populations and was accepted to be

### Table 1. Diagnosis of acute rheumatic fever, modified Jones criteria, 2015*

<table>
<thead>
<tr>
<th>A. For all patient populations with evidence of preceding group A streptococcal infection (other than chorea)</th>
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<tr>
<td>Diagnosis: initial ARF</td>
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<td>Diagnosis: recurrent ARF</td>
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<th>B. Major criteria</th>
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<td>Low-risk populations</td>
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<td>Carditis (Clinical and/or subclinical)</td>
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<td>Arthritis (Polyarthritis only)</td>
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<td>Chorea</td>
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<td>Subcutaneous nodules</td>
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<th>C. Minor criteria</th>
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<tr>
<td>Low-risk populations</td>
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<tr>
<td>Polyarthralgia</td>
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<tr>
<td>Fever (≥38.5°C)</td>
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<td>ESR ≥60 mm/h and/or CRP ≥3 mg/dL</td>
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<td>Prolonged PR on ECG (for age)</td>
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<td>(unless carditis is a major criterion)</td>
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ARF: acute rheumatic fever; CRP: C-reactive protein; ESR: Erythrocyte sedimentation rate

*Prepared by utilizing the reference number 10.

1. Low-risk populations are those with ARF incidence ≤2/ per 100 000 school-aged children or all-age rheumatic heart disease prevalence of ≤1/ per 1000 population per year.

2. Subclinical carditis is pathological echocardiographic valvulitis.

3. Polyarthralgia should only be considered as a major manifestation in moderate- to high-risk populations after exclusion of other causes. As in past versions of the criteria erythema marginatum and subcutaneous nodules are ‘stand-alone’ major criteria. Additionally, joint manifestations can only be considered in either the major or minor categories but not both in the same patient.

4. CRP value must be greater than upper limit of normal for the laboratory. Also because ESR may evolve during the course of ARF, peak ESR values should be used.
a minor criterion. In the final Jones criteria, it remained as a minor criterion in low-risk populations and was accepted to be a major criterion in moderate- and high-risk populations in order to increase the sensitivity of the criteria. Excluding other diseases with differential diagnosis is very important especially in patients in whom aseptic monoarthritis or polyarthralgia is used as a major criterion. Erythrocyte sedimentation rate is increased and fever may accompany in most of the arthritis cases related with conditions other than ARF. Therefore, Jones diagnostic criteria (1 major and 2 minor) are easily met and a misdiagnosis may be made in arthritis cases related with other conditions. Evidence of preceding group A streptococcal infection should be definitely searched in these patients. However, it should be kept in mind that ARF does not develop in all patients who have had group A streptococcal infection, positive growth in throat culture may be a carrier state and the antistreptolysin O (ASO) level makes a peak in 2-4 weeks following streptococcal infection may remain high for 3-6 months and even for one year. The chance of detection of a high titer of ASO in the community is high, because group A streptococcal infection endemia occurs frequently in countries where ARF is prevalent. An increased ASO level alone is not significant in patients who do not meet the diagnostic criteria. The contribution of ASO titer to the diagnosis is only important in terms of exclusion of the disease when low levels are found. If the ASO titer is low in a patient in whom ARF is considered (excluding chorea), it should be repeated 1-2 weeks later. If it is high or increasing, this finding supports the diagnosis.

Use of nonsteroid antiinflammatory drugs including ibuprofen as antipyretic and analgesic disrupts the course of arthritis, changes the clinical picture and makes the diagnosis difficult. Use of these drugs should be interrogated and the history and findings should be evaluated under the light of this information in patients in whom ARF is suspected. Use of nonsteroid antiinflammatory drugs as analgesic until the duration of arthritis may affect both clinical and laboratory findings. Findings of arthritis will resolve spontaneously in four weeks even if no treatment is administered in ARF, whereas use of nonsteroid antiinflammatory drugs with an insufficient dose and duration may cause the severity of arthritis to show a fluctuating course and prolong the duration of arthritis. In addition, it may also cause the laboratory findings to change. It is very important to use drugs like paracetamol instead of nonsteroid antiinflammatory drugs as antipyretic and analgesic until the diagnosis is made in patients in whom ARF is suspected.

In patients in whom joint findings are used as major criteria, arthralgia is not used as a minor criterion.

Poststreptococcal reactive arthritis is an arthritis which develops after streptococcal infection with a short latent period (one week-10 days) involving small joints with a long duration (approximately two months, may extend up to eight months) and which does not respond to salicylate treatment. In some patients, the clinical picture transforms to ARF. In 2012, the American Heart Association recommended that patients who are considered to have poststreptococcal reactive arthritis should be monitored carefully in terms of carditis. Secondary prophylaxis should be administered against streptococci for one year in patients who do not have cardiac involvement, prophylaxis should be discontinued at the end of one year if cardiac involvement is not present and prophylaxis should be continued in patients who have cardiac involvement (17, 18). The Australian Heart Association recommended secondary prophylaxis for one year in patients in whom poststreptococcal reactive arthritis is considered in populations with a low ARF risk and for five years in populations with a moderate and high ARF risk. They proposed that prophylaxis may be discontinued at the end of this period if there is no cardiac involvement and prophylaxis should be continued in patients with cardiac involvement (9). Although studies in this area are continuing, it seems to be appropriate to administer prophylaxis for five years in patients who are thought to have poststreptococcal reactive arthritis, to discontinue prophylaxis at the end of this period in patients who do not have cardiac involvement and to continue prophylaxis in patients with cardiac involvement, because Turkey carries a moderate and high risk in terms of ARF.

The differential diagnosis of joint findings related with ARF includes septic arthritis, juvenile idiopathic arthritis, familial Mediterranean fever, serum sickness, systemic lupus erythematosus, dermatomyositis, Henoch-Schönlein purpura, viral arthropathy, reactive arthropathy, poststreptococcal reactive arthritis, Lyme disease, infective endocarditis, gout, sickle cell anemia, leukemias and lymphomas.

Carditis: Carditis is observed with the second highest frequency after arthritis and is more common at younger ages in contrast to arthritis. Since carditis may lead to acute heart failure and chronic valve disease, it is the most severe major finding in ARF. Carditis most commonly involves the endocardium and leads to valvulitis by involving valvular endocardium. The myocardium and pericardium are affected in approximately 10% of
the patients. Carditis generally does not lead to complaint. It is detected during examination performed because of arthritis or chorea. Heart failure is rare. Mitral insufficiency is found most commonly in patients with carditis. The second most common valvular disorder is aortic insufficiency. Aortic insufficiency is rarely alone. It is frequently associated with mitral insufficiency. Involvement of the tricuspid and pulmonary valves occurs rarely. Mitral stenosis and aortic stenosis may emerge years after the acute attack (generally at adult ages).

In the previous Jones criteria and in the World Health Organization’s recommendations, carditis which was a major criterion was based on clinical findings and echocardiography was recommended as an assistive method for confirmation of valvular lesions and pericardial effusion (1, 5). It was proposed that echocardiography should be performed in all patients in whom ARF was considered and it was helpful in confirming the diagnosis and diverging from false positive diagnoses. However, evidence which supported inclusion of echocardiography findings in the diagnostic criteria has been obtained in the last 20 years (6-8). Pathological valve involvement found by echocardiogram without clinical signs of carditis is called subclinical carditis or silent carditis. In a meta-analysis in which studies related with subclinical carditis were examined, it was reported that subclinical carditis was found in 0-53% of the patients with ARF (mean rate 16.8%) (8). In the ARF diagnostic criteria of the Australian and New Zealand Heart Association and in the final Jones criteria updated in 2015, subclinical carditis was accepted to be a major criterion in both low-risk and moderate- and high risk populations (9-11). Evaluation by echocardiogram in terms of heart involvement is very important to detect subclinical carditis both at the time of disease onset and during the later follow-up period even if it is not found at the beginning in patients in whom ARF is suspected.

Many studies have been conducted to differentiate mild valvular insufficiency in subclinical carditis from physiological valve insufficiency in healthy individuals and criteria have been established (6-8, 19). However, it has been proposed that the guideline published by the World Heart Association in 2012 and used to detect valve involvement related with rheumatic heart disease should be adapted in order to differentiate mild valvular insufficiency in subclinical carditis from physiological valve insufficiency in healthy individuals in the final Jones criteria (11, 19). Nevertheless, findings detected in chronic valvular problems may not be found in acute valvular involvement. In addition, it is clear that the criteria in which the degree of valvular insufficiency on colored Doppler is used are not appropriate for patients who are aged younger and who have a low body weight, considering that ARF may be observed at the age of 5 years and rarely at younger ages. In addition, valvular insufficiency flow rate is closely related with blood pressure. Physiological insufficiency may be observed in the three valves except for the aortic valve in healthy children. Insufficiency found in the aortic valve in the childhood is related with congenital or acquired valvular problems. Here, the problem is to differentiate mitral insufficiency related with ARF from physiological mitral insufficiency. New criteria which would be established according to age and/or body weight are needed to differentiate mild mitral insufficiency related with subclinical valvulitis from physiological mitral insufficiency in healthy children.

Chorea: It is observed more rarely compared to polyarthritis and carditis. No amendment related with chorea was made in the final Jones criteria (11). Chorea is more common in girls and in the adolescence. Its latent period is long (1-6 months, mean period four months). Since chorea is frequently a late finding of ARF, major findings, minor findings and findings of preceding streptococcal infection may not be observed when chorea is found. Unintentional, involuntary, rapid and bouncing movements, involuntary contraction in the tongue, muscle weakness, disruption in the fine motor movements (disruption in handwriting, inability to button up etc.) and emotional disorders (hyperactivity, crying and laughing attacks etc.) occur. These findings disappear in sleep and increase during voluntary movements. They are generally bilateral and sometimes unilateral (hemichorea). Handwriting is specially affected. It is a self-limited disease. It lasts for a mean period of three months and may continue up to 17 months. The movement of chorea should be differentiated from tics, athetosis, conversion reactions, hyperkinesia and drug reactions. Subclinical carditis which is found by echocardiography and not found clinically is found in many patients with chorea (6-8). Therefore, echocardiographic examination should be performed to investigate subclinical carditis when chorea is suspected.
Erythema marginatum: This finding is observed rarely. No amendment related with erythema marginatum was made in the final Jones criteria (11). Erythema marginatum is in the form of map-like, painless and unitchy erythema which becomes pale from the center with irregular borders generally observed on the trunk and the internal side of the arms and legs and not observed on the face. Erythema marginatum is migratory like arthritis. It is aggravated when the environmental temperature increases and may disappear in cold environment. It spontaneously resolves after a few days. Although it is a major criterion of ARF, it is not diagnostic alone without the presence of another major criterion, because it may also be found in other conditions including acute glomerulonephritis and drug-induced rash.

Subcutaneous nodules: Subcutaneous nodules are observed rarely. No amendment related with subcutaneous nodule was made in the final Jones criteria. Subcutaneous nodules are painless nodules which are not attached to the surrounding with a size of 0.5-2 cm observed on the extensor surfaces of joints, scapula or mastoid. They disappear in a few days-weeks. Although they are a major criterion of ARF, they are not diagnostic alone without the presence of another major criterion, because they may also be found in other conditions including juvenile idiopathic arthritis and systemic lupus erythematosus.

Minor criteria

Fever: Currently, fever is observed less frequently. While a body temperature of 39 °C and above was used as a minor criterion in the previous Jones criteria, a body temperature of 38.5 °C and above in low-risk populations and a body temperature of 38 °C and above in moderate- and high-risk populations were accepted as minor criteria in the final Jones criteria. A body temperature of 38 °C and above should be used as a minor criterion in our country. Fever which is found in the beginning of the disease decreases in a few days.

Arthralgia: According to the final Jones criteria, polyarthralgia in low-risk populations and monoarthralgia in moderate- and high-risk populations including our country are used as minor criteria. However, exclusion of the other causes by differential diagnosis is very important in terms of making an accurate diagnosis when joint findings are used for the diagnosis. When joint findings are used as major criteria, they can not be used as minor criteria.

PR interval: PR prolongation on ECG is observed frequently in patients with ARF. In the final Jones criteria, a PR interval longer for age was proposed as a minor criterion, but a PR interval longer for age and heart rate should be used as a minor criterion, because accompanying fever and myocarditis may lead to tachycardia in these patients. PR prolongation is not a specific finding for the disease. Prolongation in PR interval may last for a long time after the acute period in a small number of patients. PR prolongation can not be used as a minor criterion in patients who have had carditis.

Laboratory findings indicating acute infection: Erythrocyte sedimentation rate and C-reactive protein (CRP) are used. An ESR above 60 mm/h in low-risk populations and an erythrocyte sedimentation rate above 30 mm/h in moderate- and high-risk populations is used as a minor criterion.

Both in low-risk and moderate- and high-risk populations, a CRP value above 3 mg/L or above the upper normal limit of the laboratory is used as a minor criterion.

Findings supporting preceding group A streptococcal infection

Antibody tests, throat culture and rapid streptococcal antigen tests are used. The most commonly used antibody tests include ASO and antideoxyribonuclease B. High or increasing values are significant for the diagnosis. If the ASO level is low in the beginning, it is repeated 1-2 weeks later. Antistreptolysine O ASO starts to increase one week after throat infection related with streptococci, reaches a peak after 3-5 weeks and may remain high for 6 months-1 year. A high ASO titer indicates only preceding streptococcal infection and should not be considered to be in favour of ARF if clinical findings of ARF are not present.

In conclusion, different diagnostic criteria were established for moderate- and high-risk populations in terms of ARF in the 2015 Jones criteria (11). Turkey was found to be compatible with moderate- and high risk populations as a result of regional screenings performed in terms of ARF and rheumatic heart disease. The amendments in low-risk populations are as follows: subclinical carditis found by echocardiography in addition to carditis detected clinically is used as a major criterion and a body temperature of 38.5 °C and above is used as a minor criterion. In moderate- and high-risk populations including Turkey, the following amendments have been made: subclinical carditis found by echocardiography in addition to carditis detected clinically is used as a major criterion and aseptic monoarthritis and polyarthritis besides migratory ar-
thritis among joint findings are used as major criteria and monoarthralgia is used as a minor criterion. The diagnostic criteria for the first attack have not been changed and three minor criteria were accepted in presence of preceding streptococcal infection in addition to the old diagnostic criteria for recurrent attack. In addition, the final Jones criteria recommended to treat patients who do not meet the diagnostic criteria of ARF completely as possible ARF if another diagnosis is not considered, to follow up these patients for 12 months with benzathine penicillin prophylaxis and to decide continuation or discontinuation of prophylaxis after 12 months by reassessment. The diagnostic criteria of ARF are valid for the acute phase of the disease and patients who meet these criteria are diagnosed with ARF with a high probability. Physicians have a great role in terms of accurate diagnosis in countries where the disease is prevalent. It is very important that physicians make an accurate diagnosis in ARF patients with their own logic and assessments in addition to using the diagnostic criteria proposed for ARF.

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**References**

3. Tibazarwa KB, Volmink JA, Mayosi BM. Incidence of acute rheumatic fever in the world: a systematic review of population-based studies. Heart 2008; 94: 1534-40. [CrossRef]