



Clinical characteristics of pediatric patients with first-attack acute rheumatic fever following the updated guideline

Güncellenen kılavuz sonrasında ilk atak akut romatizmal ateş tanısı alan çocuk hastaların klinik özellikleri

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Cite this article as: Güler M, Laloğlu F, Olgun H, Ceviz N. Clinical characteristics of pediatric patients with first-attack acute rheumatic fever following the updated guideline. *Turk Pediatri Ars* 2019; 54(4): 220–4.

Abstract

Aim: To evaluate the clinical features of children diagnosed as having acute rheumatic fever between June 2015 and November 2018, and the changes observed in patient groups in comparison with data obtained in previous years. The diagnosis of acute rheumatic fever was made using the updated Jones criteria.

Material and Methods: The medical records of pediatric patients who were diagnosed as having acute rheumatic fever between June 2015 and November 2018 using the updated criteria, were examined retrospectively. The data of a previous study that used the old criteria were reorganized and the two groups were compared.

Results: A total of consecutive 50 patients [22 males (44%)] who presented in the study period and were diagnosed as having first-attack acute rheumatic fever, were included in our study. Carditis was found in 42 (84%) patients. Manifest carditis was found in 24 patients and silent carditis was found in 18 patients. Joint involvement was present in 34 (68%) patients. Accompanying carditis was present in all 14 patients (28%) who were found to have chorea. Erythema marginatum and subcutaneous nodules were not found in our patients. When evaluated in terms of the updated criteria, a diagnosis of rheumatic fever was made with silent carditis+polyarthralgia in two patients, with silent carditis+monoarthritis in two patients, with polyarthralgia in four patients, and with monoarthritis in one patient in our study. A diagnosis could be made by means of the updated criteria in a total of 9 (18%) patients. When compared with the previous study, an increase in the rate of silent carditis (from 21.8% to 36%) and a reduction in the rate of total carditis (from 92% to 84%) were found.

Conclusion: Our results show that the updated Jones criteria prevent under diagnosis of acute rheumatic fever in an important number of patients.

Keywords: Acute rheumatic fever, children, updated Jones criteria

Öz

Amaç: Bu çalışmada kliniğimizde Haziran 2015–Kasım 2018 tarihleri arasında akut romatizmal ateş tanısı almış çocuk hastaların klinik bulgularının değerlendirilmesi ve geçmiş yıllardaki verilerle karşılaştırılarak hasta gruplarında gözlenen değişimleri değerlendirmektir. Hastalarımıza güncellenen Jones ölçütleri ile tanı konuldu.

Gereç ve Yöntemler: Haziran 2015–Kasım 2018 tarihleri arasında yeni ölçütler ile akut romatizmal ateş tanısı almış çocuk hastaların tıbbi kayıtları geriye dönük olarak incelendi. Eski tanı ölçütleri kullanılarak daha önce yapılmış bir çalışmanın verileri tekrar düzenlendi ve iki grup karşılaştırıldı.

Bulgular: Çalışmamıza belirtilen tarihler arasında başvuran ve ilk atak akut romatizmal ateş tanısı alan 22'si (%44) erkek ardışık toplam 50 hasta alındı. Kırk iki (%84) hastada kardit saptandı. Belirgin kardit 24 ve sessiz kardit 18 hastada saptandı. Eklem tutulumu 34 hastada (%68) vardı. Kore saptanan 14 (%28) hastanın tümünde eşlik eden kardit vardı. Hastalarımızda eritema marginatum ve deri altı nodülü saptanmadı. Çalışmamızda yenilenen ölçütler yönünden değerlendirildiğinde iki hastaya sessiz kardit+poliartralji, iki hastaya sessiz kardit+monoartrit, dört hastaya poliartralji, bir hastaya monoartrit ile tanı konuldu. Toplam 9 (%18) olguya yenilenen ölçütler sayesinde tanı konulabildi. Daha önce yapılan çalışma ile karşılaştırıldığında sessiz kardit oranında artma (%21,8'den %36'ya), toplam kardit oranında azalma (%92'den %84'e) saptandı.

Çıkarımlar: Sonuçlarımız güncellenen Jones ölçütlerinin önemli sayıda hastada akut romatizmal ateş tanısını koymadaki yetersizliğini önlendiğini göstermektedir.

Anahtar sözcükler: Akut romatizmal ateş, çocuk, yeni tanı ölçütleri

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Received /Geliş Tarihi: 13.01.2019 **Accepted /Kabul Tarihi:** 04.04.2019

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DOI: 10.14744/TurkPediatriArs.2019.69376

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Introduction

Acute rheumatic fever (ARF) is the most important cause of acquired heart disease in children. The diagnosis is made using the modified Jones criteria. The update made in 2015 introduced important modifications in the diagnostic criteria. In this study, we aimed to evaluate the clinical features of children diagnosed as having acute rheumatic fever between June 2015 and November 2018, and the changes observed in patient groups in comparison with data obtained in previous years.

Material and Methods

This study was conducted in Atatürk University, Faculty of Medicine, Division of Pediatric Cardiology between June 2015 and November 2018. The medical records of children who were diagnosed as having ARF between the study dates (group A), were examined retrospectively. The patients' clinical data and laboratory results were recorded.

Since June 2015, the diagnosis of ARF has been made in our clinic using the criteria recommended for populations in which the disease occurs with a moderate-high prevalence (1).

The data of patients who were diagnosed as having ARF between June 2005 and July 2008 in our clinic and whose records were collected because of a thesis study, were rearranged (group B) and compared with the data of the new study (2). In that study, the diagnosis was made using the old diagnostic criteria (3).

Ethics committee approval was obtained from Atatürk University, Faculty of Medicine, Clinical Researches Ethics Committee for the present study (Meeting No. 07, Decision No. 10). The study was conducted in accordance with the Declaration of Helsinki. No financial support was received for the study. Informed consent was not obtained from the patients because the study was a retrospective study.

Results

A total of 50 consecutive patients [22 males (44%)] who presented during the study period and were diagnosed as having first-attack acute rheumatic fever, were included in our study. The female/male ratio was 1.27:1. The mean age was 13.5±3.3 years and the mean body weight was 40±14.2 kg.

The frequencies of major findings in both groups are shown in Table 1.

In group A, carditis was found in a total of 42 (84%) patients. Other accompanying major findings were present in 40 (80%) of these patients, whereas carditis was the single major finding in two patients (5%). Manifest mitral insufficiency and silent aortic insufficiency were present in both of these patients who had carditis alone. Forty patients had mild carditis and two patients had moderate carditis. When we compared the groups in terms of the frequency of carditis, we found that the rate of manifest carditis was 48% and the rate of silent carditis was 36% in group A. In group B, the rate of manifest carditis was found as 70% and the rate of silent carditis was 21.8%.

Joint involvement was present in a total of 34 patients (68%), carditis accompanied in 26 of these patients (76.6%), and 11 (32.3%) of these patients had silent carditis. Twenty patients (58%) had polyarthritis, nine (26.4%) had polyarthralgia, and five (16%) had monoarthritis. Joint findings were the only major finding in nine patients. In the patients who had polyarthritis and polyarthralgia, the joints involved included the ankles, knees, wrists, and hips, and these patients had migratory joint involvement. Among the patients who had monoarthritis, the right ankle was involved in three patients, the left ankle was involved in one patient, and the left knee was involved in one patient. The frequency of joint involvement was found as 68% and 65.9% in group A and group B, respectively.

Accompanying carditis (manifest=7, silent=7) was present in all patients who were found to have chorea. The frequency of chorea was found as 28% and 17% in group A and group B, respectively. Erythema marginatum and subcutaneous nodules were not found in our patients in either period.

When group A and group B were compared in terms of minor criteria (Table 2), it was observed that the most common minor finding was increased acute-phase reactants in both groups. However, a reduction occurred in the frequency of fever and arthralgia, and an increase occurred in the frequency of prolonged PR over time.

A diagnosis of ARF was made with two major findings in 40 patients, with one major+two minor findings in seven patients, with one major+three minor findings in two patients, and with one major+one minor finding in one patient.

A total of 10 patients (20%) had a single major finding in group A. Presence of two additional minor findings found in these patients is shown in Table 3.

Table 1. Major criteria found at the time of diagnosis

Finding	Group A				Group B			
	n	%	Manifest carditis	Silent carditis	n	%	Manifest carditis	Silent carditis
Carditis+polyarthritits	17	34	10	7	26	40.8	26	None
Carditis+polyarthralgia	5	10	3	2	–	–	–	–
Carditis+monoarthritits	4	8	2	2	–	–	–	–
Carditis+chorea	14	28	7	7	5	7.8	5	None
Carditis+polyarthritits+chorea	None				2	3.1	2	None
Carditis	2	4	2	None	12 ^a	18.7	12	None
Polyarthritits	3	6	None		13	20.3	None	9
Polyarthralgia	4	8	None		–	–	–	–
Monoarthritits	1	2	None		–	–	–	–
Chorea	None				6	9.3	None	5
Total	50	100	24 (48%)	18 (36%)	64	100	45 (70%)	14 (21.8%)

^aFour of these patients had monoarthritits, but it was not considered major criterion

Table 2. Minor criteria found at the time of diagnosis^a

	Group A		Group B	
	n	%	n	%
Increased ERS+CRP	41	82	55	85.9
Fever	16	32	53	82.8
Prolonged PR	20	40	14	21.8
Arthralgia	1	2	11	17.2

^aAll these criteria were not used in the diagnosis because of accompanying major findings. ESR: Erythrocyte sedimentation rate; CRP: C reactive protein

Discussion

Acute rheumatic fever is one of the nonsuppurative complications of pharyngitis caused by group A beta-hemolytic streptococci. It constitutes 50% of all cardiovascular diseases and hospitalizations due to heart disease in many developing countries. There are at least 15–16 million people with rheumatic heart disease worldwide. Currently, 233 000 people die of ARF or rheumatic valve disease yearly (4).

There is still no clinical finding or laboratory test that is diagnostic by itself. The Jones criteria were established in 1944 for the diagnosis, and updated in 1965, 1984, 1992, and finally in 2015 (1, 3).

In the last update, populations were classified as low and moderate-high risk according to prevalences of ARF, and the criteria were rearranged according to these classifications (1). In Turkey, the data related to ARF prevalence are based on local studies. In a study conducted in 2012 that

Table 3. Minor criteria used in the diagnosis in ten patients who had a single major finding (Group A)

	n	%
Increased ERS/CRP ^a	1	10
Increased ERS/CRP+fever	5	50
Increased ERS/CRP+prolonged PR	2	20
Increased ERS/CRP+fever+prolonged PR	1	10
Increased ERS/CRP+monoarthralgia	1	10

^aA diagnosis of ARF was made because manifest mitral insufficiency and silent aortic insufficiency found in this patient, which could not be explained by other valve pathologies. ESR: Erythrocyte sedimentation rate; CRP: C reactive protein

included 30 years of data, the prevalence of ARF in Ankara was reported as 37/100 000 for 1980–1989, 60/100 000 for 1990–1999, and 21/100 000 for 2000–2009 (5). The Turkish Pediatric Cardiology and Heart Surgery Association estimated the prevalence of ARF in Turkey as 9/100 000 (unpublished data, 2017) in children aged between 5 and 15 years in a prestudy conducted throughout the country. Thus, it is understood that Turkey is in the moderate-high risk population group.

There are few studies presenting the clinical features of patients diagnosed newly in the light of the updated criteria, and we are not aware of the presence of studies conducted in this area (6).

An important modification introduced by the final update was the inclusion of silent carditis, polyarthralgia, and monoarthritits in the major criteria. Modifications for limiting values were also recommended in minor criteria (1).

The definition of silent carditis was made for the first time when carditis was shown clinically in 10 and echocardiographically in nine of 20 patients who had rheumatic chorea (7). In the last 20 years, subclinical (silent) carditis found by echocardiography in the absence of clinical findings, has frequently come to the forefront with widespread use of echocardiography throughout the world; a metaanalysis revealed that silent carditis was found with a rate of 0–53% in patients with ARA (mean rate: 16.8%) (8). However, it was not considered a major finding until 2015. The data obtained in our study indicated that there was an increase in the frequency of silent carditis and a reduction in the frequency of total carditis in our region between 2015 and 2018 compared with the previous period. This can be interpreted such that the disease severity gradually decreased. On the other hand, this finding may also be related to the fact that the patients with ‘manifest’ carditis were being followed up by new pediatric cardiology centers that started to operate in our region. In addition, the referral of patients with polyarthralgia and monoarthritis to our outpatient clinic with a prediagnosis of ARF might also have increased the frequency of silent carditis.

In group B patients, the only major joint finding was polyarthritis (40/40). In group A patients, the frequency of polyarthritis was found as 58.8% (20/34), and the major joint finding in 14 patients was monoarthritis. Monoarthritis was present in 10% of the patients in group A. Although monoarthritis was not considered a major finding in the period when group B patients were being followed up, monoarthritis accompanied in 6.25% of these patients. In a study conducted in 2001, it was reported that monoarthritis was observed with a rate reaching up to 17% in patients with ARF in countries where the disease occurred with a high prevalence (9).

In the last update, no modification related to chorea was made. The increase in the frequency of chorea may be related to the fact that a pediatric neurologist was not available in either pediatric cardiology clinic found in the vicinity, and these patients were referred to our hospital.

We think that acceptance of polyarthralgia as a major criterion caused a reduction in the frequency of arthralgia as a minor finding (Table 2).

When evaluated in terms of updated criteria, a diagnosis of rheumatic fever was made with silent carditis+polyarthralgia in two patients, with silent carditis+monoarthritis in two patients, with polyarthralgia in four patients, and with monoarthritis in one patient in our study. The diagnosis could be made by means of the

updated criteria in a total of nine (18%) patients. In a similar study conducted recently in Italy, this rate was reported as 20.7% (6). This finding shows that the last update will decrease the difficulty in making the diagnosis for populations in which ARF is observed with a high frequency.

The limited number of subjects and short follow-up time were the basic limitations of our study. There may be patients whose diagnoses were missed because all patients with joint symptoms who presented to the general pediatrics outpatient clinic were not included in our study.

Our results show that the updated Jones criteria prevented under diagnosis of ARF in an important number of patients in our region.

Ethics Committee Approval: Atatürk University, Faculty of Medicine, Clinical Researches Ethics Committee, Date: 11/29/2018, Meeting number: 7, Decision number: 10.

Informed Consent: As it was a retrospective study, consent was not obtained from the patients.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - N.C., H.O.; Design - N.C., M.G.; Supervision - N.C., H.O.; Data Collection and/or Processing - M.G., F.L.; Analysis and/or Interpretation - N.C., M.G.; Literature Review - M.G., F.L.; Writing - M.G., N.C.; Critical Review - Z.K.

Conflict of Interest: The authors have no conflicts of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

Etik Kurul Onayı: Atatürk Üniversitesi Tıp Fakültesi Klinik Araştırmalar Etik Kurulu, 29.11.2018 tarihli, Toplantı no; 7 ve karar no; 10 onayı.

Hasta Onamı: Geriye dönük bir çalışma olduğu için hastalardan onam alınmamıştır.

Hakem Değerlendirmesi: Dış bağımsız.

Yazar Katkıları: Fikir - N.C., H.O.; Tasarım - N.C., M.G.; Denetleme - N.C., H.O.; Veri Toplanması ve/veya İşlemesi M.G., F.L.; Analiz ve/veya Yorum - N.C., M.G.; Literatür Taraması - M.G., F.L.; Yazıyı Yazan - M.G., N.C.; Eleştirel İnceleme - H.O., F.L.

Çıkar Çatışması: Yazarlar çıkar çatışması bildirmemişlerdir.

Mali Destek: Yazarlar bu çalışma için finansal destek almadıklarını beyan etmişlerdir.

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