

# Echocardiographic Stigmata in the First Attack of Acute Rheumatic Fever as a Major Criterion for the Diagnosis of Rheumatic Carditis in Misan, Iraq

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## Abstract

**Background:** Doppler Echocardiography facility is usually available in the most areas of the world and its routine use in the initial diagnosis of rheumatic heart disease may promote early detection, much earlier than clinical examination does.

**Objectives:** To evaluate the utility of Doppler Echocardiography for the diagnosis of carditis in patients whom clinical examination did not indicate any evidence of carditis.

To describe the spectrum of cardiac abnormalities in patients with primary episode acute rheumatic fever.

**Patients and Method:** A cross sectional and prospective study conducted during a period of five years in Misan province (South East Iraq). Referred patients for an echocardiographic evaluation in Al-Sader Teaching Hospital with diagnosis of acute rheumatic fever as first attack were selected. All patients were examined clinically then subjected to Doppler Echocardiographic evaluation in the first 48-72 hours of acute phase and 2 weeks later.

**Results:** A total of 36 patients with median age of 13±4.69 years and male predominance were presented with diagnosis of primary attack of acute rheumatic fever. The majority were presented with migratory polyarthritides (80.6%) and clinical carditis (44.4%). Clinical carditis was found in 16/36 patients while the remaining (20 patients) had no clinical carditis. On the other hand, Doppler Echocardiography confirmed the diagnosis of carditis in additional 10 cases without clinical carditis.

About almost all patients with acute rheumatic carditis (25/26) had mitral valve /aortic valve thickness >4mm. Valvular regurgitations were constituted the major manifestation; mitral regurgitation was found in the majority (21/26 patients) followed by aortic and tricuspid regurgitation.

**Conclusion:** Doppler Echocardiography is more sensitive than a clinical assessment in the detection of carditis in patients with an initial attack of acute rheumatic fever. It is a useful method in identifying the subclinical mitral and aortic valvular disease when carditis cannot be detected clinically. Therefore, it is reasonable and valuable to support the addition of echocardiographic stigmata of rheumatic carditis to the current diagnostic criteria as an isolated major criterion by itself for the validation of rheumatic carditis with subclinical carditis.

**Keywords:** Rheumatic fever, carditis, echocardiography, Misan, Iraq.

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## Introduction

Acute rheumatic fever (ARF) is a common serious public health problem in the developing countries<sup>(1)</sup>. Despite the decline in its incidence in Europe and

North America with the tremendous progress made in cardiology, ARF is still forming a major healthcare concern for the vast majority of the populations in the world including children, adolescent, and young adults (1, 2, 3, 4).

Rheumatic heart disease (RHD), the sequel of ARF, is a very common cause of cardiovascular morbidity and mortality, as well as, the predominant indication for cardiac surgery in the developing countries (1, 5).

At 2001, World Health Organization (WHO) reported that a clinical examination is still the basis of the diagnosis of ARF and carditis, and the role of an echocardiographic study is considered supportive. However, an Echocardiography/Doppler examination should be used if the facilities are available (5).

In addition, the WHO reported that an involvement of echocardiography to detect valvular regurgitation and confirm the diagnosis of subclinical rheumatic carditis is still controversial (5).

Moreover, different studies showed that the inclusion of imaging study of echocardiography into Jones criteria cannot be justified unless long term results of prospective studies are available to emphasize the importance of detecting subclinical rheumatic carditis in treatment and prognosis (6, 7).

Nowadays, Doppler Echocardiography facility is usually available in the most areas of the world and its routine use in the initial diagnosis of RHD may promote early detection, much earlier than clinical examination does (8). Echocardiography has been proven to be more specific and sensitive than auscultation. It can detect early evidence of valvular involvement, can confirm the presence of valvular regurgitation, and can exclude the non-rheumatic etiologies of valvular involvement (9, 10).

Consequently, in 2012, the World Heart Federation echocardiographic criteria for RHD have been developed. Three groups are identified on the basis of assessment by 2-Dimensional, continuous-wave, and color-Doppler echocardiography: definite RHD, borderline RHD, and normal. Additionally, even the morphological features of RHD, as well as, the criteria for pathological aortic and mitral regurgitations are also defined (11).

Lately, in 2015, the American Heart Association released recommendation for using echocardiography with Doppler in ARF; one of these recommendations is

to assess for the presence of carditis in the absence of auscultatory findings, especially in moderate to high risk groups and whenever RHD is a possible diagnosis (12).

For these reasons, this study had arisen to evaluate the utility of Doppler Echocardiography for the diagnosis of carditis in patients whom clinical examination did not indicate any evidence of carditis and to describe the spectrum of cardiac abnormalities in patients with primary episode ARF.

## Patients and Method

A cross sectional and prospective study conducted during a period of five years from September 2010 to September 2015 in Misan province (South East Iraq).

Referred patients for an echocardiographic evaluation in Al-Sader Teaching Hospital with diagnosis of ARF as initial attack were selected. Diagnosis of ARF in these cases was according to the revised Jones criteria (12, 13).

Total sample size was 40 patients with first episode of acute rheumatic fever, but only 36 patients came for follow-up evaluations. So the final sample size was 36 cases; male were 22 cases and female were 14 cases. Age was ranging from 6-22 years.

Initially, all patients were examined clinically at the referral time. So patients were classified into 2 groups:

16 cases with clinical carditis.

20 cases without clinical carditis.

Then all patients were subjected to Two-Dimensional Echocardiography and Doppler Color Flow (2D Echo Doppler) evaluation in the first 48-72 hours of acute phase and 2 weeks later. All patients were examined by the same specialist doctor to exclude personal variation.

Echocardiographic imaging was performed with GE machine, model vivid, version E9 following the recommendation and guidelines of using echocardiography with Doppler in ARF (11, 12).

**Exclusion criteria:** Previous history of ARF or recurrent attacks of ARF, known case of chronic RHD, and echocardiographic stigmata of chronic valvular disease (such as restricted excursion or doming of valve leaflets or significant mitral or aortic stenosis). In addition, any case rejected to be involved in this study

or did not complete the follow-up evaluations was excluded.

The study protocol was reviewed; ethical approval and official permission were obtained from Ministry of Higher Education, College of Medicine in Misan, Misan directorate of health and Al-Sader Teaching Hospital to carry out this study. An informed written consent was obtained from each patient or from their parents.

The analysis of data was carried out using Microsoft Excel and was presented in form of tables of numbers and percentages.

### Results

A total of 36 patients with median age of 13±4.69 years and male predominance were presented with diagnosis of initial attack of ARF according to revised Jones criteria. The majority were presented with migratory polyarthritits (80.6%) and clinical carditis (44.4%) as shown in **table 1**.

Clinical carditis was found in 16 patients with a first attack of ARF while the remaining (20 patients) had no clinical carditis. On the other hand, 2D Echo Doppler confirmed the diagnosis of carditis in additional 10 cases without clinical carditis as shown in **table 2**.

About almost all patients with acute rheumatic carditis (25/26) had mitral valve /aortic valve thickness >4mm. Valvular regurgitations were constituted the major manifestation; mitral regurgitation was found in the majority (21/26 patients) followed by aortic and tricuspid regurgitation. In addition, acute heart failure was present in 2/26 patients and the same number was seen with pericardial effusion as shown in **table 3**.

**Table1. Base-line characteristics and frequency of major Jones criteria of patients with initial attack of acute rheumatic fever at the time of clinical evaluation.**

Parameter	Number (%)
<b>Gender:</b>	
Male	22 (61.1%)
Female	14 (38.9%)
<b>Age (year):</b>	
Age range	6-22
Age mean ±SD	13±4.69

**Cont... Table1.**

<b>Major Jones criteria:</b>	
Migratory polyarthritits	29 (80.6%)
Clinical carditis	16 (44.4%)
Subcutaneous nodules	2 (5.6%)
Erythema marginatum	1(2.8%)
Chorea	0 (0%)

**Table 2. The role of Two-Dimensional Echocardiography and Doppler Color Flow in the diagnosis of carditis in patients with initial attack of acute rheumatic fever.**

Acute rheumatic fever	Carditis confirmed by Two-Dimensional Echocardiography and Doppler Color Flow
With clinical carditis	16/16
Without clinical carditis	10/20
Total	26/36

**Table3. Features of acute rheumatic carditis (first attack) detected by Two-Dimensional Echocardiography and Doppler Color Flow in 26 cases.**

Features detected by Two-Dimensional Echocardiography and Doppler Color Flow	Number (%)
Mitral valve /Aortic valve thickness >4mm	25 (96.2%)
Mitral regurgitation grade I-II	21 (80.8%)
Mitral valve prolapse	13(50.0%)
Rheumatic nodule (beaded appearance )	7 (26.9%)
Aortic regurgitation	5(19.2%)
Tricuspid regurgitation	4(15.4%)
Pancarditis	2 (7.7%)
Combined mitral and aortic regurgitation	2 (7.7%)
Pericardial effusion	2 (7.7%)
Signs of cardiac failure	2 (7.7%)

### Discussion

The recent guidelines of Australian <sup>(14)</sup> and New Zealand <sup>(13)</sup> on ARF have been accepted an echocardiographic subclinical carditis as a major criterion. So, all patients with suspected or definite diagnosis of ARF should undergo an echocardiography to detect the evidence of carditis.

In the current study; the frequency of primary episode ARF occurrence during 5 years was 36 (plus 4 cases were excluded) which was much less than the

frequency of Turkey study in which 118 cases were diagnosed during the period of 3 years only<sup>(2)</sup>. This frequency of occurrence would not reflect the exact rate in Misan because some patients may be already referred to other centers.

Additionally, the present study showed that migratory polyarthritis (80.6%) was the commonest manifestation of the first attack ARF while a clinical carditis (44.4%) was the second presenting features. These rates were higher than Turkey<sup>(2)</sup>.

A clinical carditis was diagnosed in less than half of cases (16/36 cases), which was slightly less than Caldas et al study who reported about 50% of cases had a clinical carditis<sup>(15)</sup>. This may be related to a personal variation in the clinical skills of auscultation.

On the other hand, this study revealed that rheumatic carditis was more frequently diagnosed by 2D Echo Doppler than clinical examination, especially in sub-clinical carditis. These results were consistent with different studies<sup>(16, 17)</sup>. A study by Marijon et al in Cambodia and Mozambique revealed that echocardiography can detect up to ten times the number of carditis in ARF in comparison to a clinical examination alone<sup>(18)</sup>.

These findings suggest that carditis was missed by a clinical examination. Also, it was reported that the clinical skill of auscultation by the training physicians may be not the optimum one, especially, in areas where ARF is declining<sup>(19)</sup>.

In addition, the precise diagnosis of both clinical and subclinical carditis by using 2D Echo Doppler has been documented and it is more accurate than clinical auscultation, and if integrated in the Jones criteria, it can prevent both over- and under-diagnosis of valvular pathology in ARF patients<sup>(20)</sup>.

Diffuse thickening > 4 mm and valvulitis of the mitral valve leaflets was a universal feature in acute rheumatic carditis (25/26 cases). Therefore, any carditis without valvulitis or valvular thickening is unlikely to be rheumatic in origin<sup>(21)</sup>.

In rheumatic carditis cases; there was 2/26 cases of acute heart failure, both of them were associated with valvular regurgitation. Obviously, echocardiography is more sensitive for diagnoses of valvular regurgitation with increasing rate in detecting mitral, aortic, and

tricuspid regurgitation.

In spite of all these findings, till now, there is a fear from increase the chance of over diagnosing carditis in large proportion of ARF patients<sup>(5)</sup>. The over interpretation of valvular regurgitation (physiological or trivial) may lead to misdiagnosis of an iatrogenic valvular disease<sup>(22)</sup>. This was attributed to a higher sensitivity of 2D Echo Doppler in detecting the abnormal blood flow and valvular regurgitations<sup>(9, 10)</sup>. On the other hand, the use of 2D Echo Doppler can decrease the over diagnosis of the functional murmur as a valvular heart disease<sup>(9, 10)</sup>.

So, an accurate interpretation, as well as, the technical expertise with 2D Echo Doppler with following the World Heart Federation echocardiographic criteria are essential to reach a correct diagnosis and to avoid the over diagnosis in RHD patients<sup>(6, 11)</sup>.

The current study clearly revealed a significant incremental diagnostic utility of 2D Echo Doppler in the first attack of ARF especially in subclinical carditis and can eliminate the over diagnosis of rheumatic carditis depending solely on cardiac auscultatory findings, hence, better diagnosis, treatment, and prophylaxis<sup>(23, 24)</sup>.

## Conclusion

The imaging study of Two-Dimensional Echocardiography and Doppler Color Flow is more sensitive than a clinical assessment in the detection of carditis in patients with an initial attack of acute rheumatic fever. It is a useful method in identifying the subclinical mitral and aortic valvular disease when carditis cannot be detected clinically. Therefore, it is reasonable and valuable to support the addition of echocardiographic stigmata of rheumatic carditis to the current diagnostic criteria as an isolated major criterion by itself for the validation of rheumatic carditis with subclinical carditis.

**Disclosure statement:** The authors report no conflict of interest and this research is not funded.

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