



# Echocardiography in Acute Rheumatic Fever

V Jacob Jose\*, Paul George\*\*

\*Professor, Head of the Department, Department of Cardiology; \*\*Associate Professor, Department of Cardiology, Christian Medical College, Vellore 632 004, Tamil Nadu.

21

## ABSTRACT

Echocardiography is an important diagnostic tool for the diagnosis of valvular lesions. Now it is recognized that echo can be used also in the diagnosis of acute rheumatic fever. In this way, patients with no auscultatory evidence of valvular regurgitations can also be identified and this will help in the early and correct diagnosis of acute rheumatic fever. In acute rheumatic fever small nodules have been demonstrated in the acute phase. In addition, there can be prolapse of the leaflet and annular dilatation. The same changes have been observed in aortic valve as well. Colour Doppler can confirm the valvular regurgitation. Patients with acute rheumatic fever who do not have cardiac involvement by echo are known to have a benign prognosis. In this way, echo plays a very important role not only in the diagnosis, but also in the prognosis of acute rheumatic fever.

## INTRODUCTION

The first attack of acute rheumatic fever is diagnosed based on Jones criteria. Though echocardiography is not included in the diagnostic criteria of Jones, there are many experts in the field who advocate the need to include Echo into the existing criteria. An international expert panel and The American Heart Association Committee on Rheumatic Fever, Endocarditis and Kawasaki Disease met in the year 2000, to review the guidelines for the diagnosis of rheumatic fever especially in the utility of newer investigations like Doppler and echocardiography in diagnosis.<sup>1</sup> Though acknowledging the differences in prevalence and the time to presentation of rheumatic fever between developing and developed countries, the committee decided not to alter the current status of using Jones criteria, and so did not include echocardiography as part of diagnostic criteria for acute rheumatic fever.

## ROLE OF ECHO IN ACUTE RHEUMATIC FEVER

1. Affirms the dominant role of valvular pathology rather than myocardial disease in the clinical manifestations of rheumatic heart disease.
2. Confirms the clinical findings and allows assessment of (a) severity of valve involvement, (b) chamber size and ventricular function and (c) presence and size of pericardial effusion.
3. Helps in serial assessment of disease progression.
4. May help in exclusion of valve involvement in suspected cases of rheumatic fever.

5. Earlier detection of carditis in cases of acute rheumatic fever.
6. In some cases help in detecting silent subclinical carditis.

## Echocardiographic features of rheumatic carditis

Rheumatic carditis is a pan-carditis involving all three layers of heart. Pericarditis and myocardial involvement in the form of congestive cardiac failure is rare in the absence of significant valvular involvement. The finding of the valvular involvement is critical to the diagnosis of rheumatic carditis and this can easily be done with the use of echocardiogram. Most cases the valvular involvement may not be severe enough to be symptomatic and in many patients the regurgitation lesions may not be clinically detected by auscultation. However, echocardiogram can pick up regurgitation lesions very easily. Thus the role of echocardiogram in acute rheumatic carditis is to identify valvular involvement in the form of regurgitations.

### *Subclinical carditis*

The presence of valvular regurgitation detected by Echo Doppler only without clinical evidence of carditis is termed "silent carditis" (subclinical carditis). This is especially relevant, as up to 50% of chronic rheumatic heart disease patients do not give history of rheumatic fever. In the Utah outbreak, 19% had Doppler evidence of cardiac involvement even though clinically considered to have isolated arthritis or pure chorea.<sup>2</sup> A number of authors have reported echocardiographically detected carditis in cases of acute rheumatic fever.<sup>3-6</sup> A trial from New Zealand showed that 'echocardiography alone' detected regurgitant murmurs became audible within two weeks of diagnosis.<sup>7</sup> A larger trial from India

showed that all cases of regurgitant lesions in cases of rheumatic detected by echocardiography were also diagnosed by careful auscultation.<sup>8</sup>

### **Echo findings of Mitral Valve**

The mitral valve is the commonest valve involved in acute rheumatic fever with carditis in 94% and also in rheumatic fever recurrences by 84%.

Diffuse thickening is universal in recurrences but only in 40% of cases in the first attack.

1. Focal nodular thickening of the body and tips of the leaflet is another important feature that can be picked up by echocardiogram. These nodules are 3–5mm in size and have different echogenicity from the valve structures and do not have the characteristic mobility of endocarditic vegetations with which it has to differentiate (Fig. 1). These are considered rheumatic verrucae equivalents and are seen commonly in the mitral valve; these are seen on tricuspid also and rarely on the aortic valve.<sup>8</sup>
2. Prolapse of the leaflet though less common is also responsible for the regurgitation in some cases (8.6% in the first attack and 13% in the second attack.) The regurgitant jet is directed posteriorly in 70% of cases. It can also be central, anteriorly directed, or multidirectional (16%, 12%, 10% respectively). In rheumatic carditis, only the coapting portion of the anterior mitral leaflet prolapses and there is no billowing of the medial portion of this leaflet. This particular feature permits us to readily differentiate from the mitral valve prolapse due to myxomatous degeneration of the valve.
3. Annular dilatation is also common with acute rheumatic fever recurrence. In a study by Marcus et al<sup>9</sup> in 73 patients with acute rheumatic carditis and mitral regurgitation who subsequently went for surgery they found that 96% had annular dilatation.
4. Elongation of the chordae to the anterior leaflet is also common. This makes the leaflet not to appose at the edges resulting in anterior leaflet prolapse. It is important to point out here that it is the chordae to the anterior mitral valve leaflet that is elongated in acute rheumatic fever.
5. Rupture of the chordae tendinae can also be responsible for mitral regurgitation, though it is rare.

In recurrent carditis, the mechanism of mitral regurgitation includes restriction of leaflet mobility, ventricular dilatation, annular dilatation and leaflet prolapse.

### **Echo findings of aortic valve**

Although rheumatic carditis predominantly involves the mitral valve, the aortic valve may be involved as well. The aortic valve involvement was detected in 39% of patients with the acute rheumatic carditis in one study.<sup>10</sup> In 17% of these patients, only echo was showing evidence of aortic regurgitation. Since aortic regurgitation is not commonly seen physiologically in children, all aortic regurgitation detected by echocardiogram in children suspected to have rheumatic carditis should be considered as evidence of an abnormality.

The mechanism for aortic regurgitation can be as follows:

1. Leaflet thickening with deficient coaptation
2. Leaflet prolapse
3. Annular dilatation
4. Aortic leaflet retraction is seen in recurrent attacks

### **Tricuspid valve**

Isolated tricuspid valve and pulmonary valve involvement is almost unknown in rheumatic carditis and a diagnosis of carditis based on isolated tricuspid or pulmonary valve involvement is never done.

### **Left ventricular function and myocardial involvement:**

Left ventricular dilatation is more common with recurrent carditis and when present with the first attack it is usually associated with moderate to severe regurgitation lesions. Uncommon with the first attack, recurrent carditis will produce annular dilatation. Fractional shortening is not altered even with congestive heart failure in the initial attack in up to 79% of cases with carditis and any decrease in fractional shortening was associated with significant valvular regurgitation and congestive heart failure.<sup>8</sup>

### **Pericarditis**

Pericarditis is unusual without evidence of significant valvular involvement. Pericardial effusion can be easily detected by echocardiography (being the gold standard). Massive effusions and pericardial tamponade have been reported in acute rheumatic fever with carditis.

## **ECHO AND PROGNOSIS**

It has been suggested that Echo detected carditis is not so benign. Folger et al<sup>11</sup> found that four of the six patients with Doppler valve involvement continued to have it 18 to 36 months later.

Vason et al<sup>12</sup> in another study showed that if Echo did not show carditis, then these patients did not develop regurgitation on follow-up.

Thus, Echo is useful not only in identifying the involvement of the heart in acute rheumatic fever; also a negative study will ensure a benign prognosis.

## **CONCLUSION**

Although echocardiogram has not been included as a diagnostic test for the diagnosis of acute rheumatic fever, Echo provides an accurate assessment of the presence and severity of valvulitis. Echo can identify valvulitis even in patients without auscultatory evidence of valvular lesions. With cardiac auscultation taking a back seat after the advent of echocardiogram, it becomes more important for us to rely on Echo to identify valvular lesions in patients with acute rheumatic fever.

## **REFERENCES**

1. Ferrieri P and for the Jones Criteria Working Group. Proceedings of the Jones criteria workshop. *Circulation* 2002;106:2521-2523.

2. Veasy LG, Wiedmeier SE, Orsmond GS, et al. Resurgence of acute rheumatic fever in the intermountain area of the United States. *N Engl J Med* 1987;316:421-427.
3. Folger GM, Hajar R, Robida A, Hajjar HS. Occurrence of valvar heart disease in acute rheumatic fever without evident carditis: colour-flow Doppler identification. *Br Heart J* 1992;67:434-439.
4. Figueroa FE, Fernandez MS, Valdes P, Wilson C, Lanas F, Carrion F, Berrios X, Valdes F. Prospective comparison of clinical and echocardiographic diagnosis of rheumatic carditis: long term follow up of patients with subclinical disease. *Heart* 2001;85:369-70.
5. Ozkutlu S, Ayabakan C, Saraclar M. Can subclinical valvulitis detected by echocardiography be accepted as evidence of carditis in the diagnosis of acute rheumatic fever? *Cardiol Young* 2001;11:255-60.
6. Hilario MO, Andrade JL, Gasparian AB, Carvalho AC, Andrade CT, Len CA. The value of echocardiography in the diagnosis and follow up of rheumatic carditis in children and adolescents: a 2 year prospective study. *J Rheumatol* 2000 4:1082-6.
7. Abernathy M, Bass N, Sharpe N. Doppler echocardiography and the early diagnosis of carditis in acute rheumatic fever. *Aust NZ J Med* 1994;24:530-35.
8. Vasan R S, Shrivastava S, Vijaikumar M, et al. Echocardiographic evaluation of patients with acute rheumatic fever and rheumatic carditis. *Circulation* 1996;94:73-82.
9. Marcus RH, Sareli P, Pocock W A, et al. Functional Anatomy of severe mitral regurgitation in active rheumatic carditis. *Am J Cardiol* 1989;63:577-84.
10. Veasy LG, Tani LY, Hill HR. Persistence of acute rheumatic fever in the intermountain area of the United States. *J Pediatrics* 1994;124:9-16.
11. Folger GM, Hajar R, Robida A, Hajjar HS. Occurrence of valvar heart disease in acute rheumatic fever without evident carditis: colour-flow Doppler identification. *Br Heart J* 1992;67:434-439.
12. Vasan R, Shrivastava S, Vijaya Kumar K, Narang R, Lister BC, Narula J. Echocardiographic evaluation of patients with acute rheumatic fever and rheumatic carditis. *Circulation* 1996;94:73-82.