Efficacy of Echocardiographic Criteria for Diagnosis of Carditis in Acute Rheumatic Fever

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Abstract

Background: There is a great need for ECHO criteria for accurate diagnosis of carditis in acute rheumatic fever.

Aim: To propose and test the efficacy of ECHO criteria for accurate diagnosis of carditis

Material and Methods: The 333 cases underwent detailed clinical examination, laboratory tests and meticulous Echocardiographic study. Vijay's ECHO criteria for the diagnosis of carditis / subclinical valvulitis was used. 220 (66.06%) cases were both Jones' positive and ECHO positive [True +ve], 52 cases (15.61%), probably had subclinical carditis as murmur was not heard (Jones'-ve) but ECHO was positive [False - ve]. Four cases, clinically diagnosed as carditis were Jones'+ve ,but ECHO showed congenital heart disease [False +ve]. 57 cases (17.11%) were clinically , echocardiographically and Jones' negative were taken as control (True –ve). Sensitivity is 81% and specificity is 93%.

Conclusions: Precise diagnosis of both carditis /subclinical valvulitis is possible with Vijay's ECHO criteria. ECHO should be included as a major criterion in Jones' criteria.

Key words: Rheumatic carditis, subclinical valvulitis, ECHO criteria

Introduction

Acute Rheumatic Fever (ARF) and its long-term sequel is still a major burning problem in children, adolescents and young adults, in a developing country like India.[1] Among the various manifestations of ARF, carditis is the only one that can cause death during the acute stage of the disease or lead to permanent damage with long-term morbidity and mortality due to RHD and congestive heart failure (CHF). Despite the modification of Jones' criteria[2] and its revision four times, [3,4], carditis in ARF is either under diagnosed, leading to nearly 50% of established RHD not receiving prophylaxis or over diagnosed, depending on traditional characteristic auscultatory findings for diagnosis of carditis.[5,6]. Precise and early diagnosis of carditis in acute rheumatic fever is eluding the clinicians. Hence, accurate diagnosis is very essential, as timely management can make the heart normal in 35 - 40% of cases, prevent recrudescence of rheumatic activity and further damage to the valves. Despite the fact that ECHO can help to diagnose carditis more accurately than traditional auscultatory findings[7] and can prevent both over diagnosis and under diagnosis[8], the question is being asked. "Could echocardiography have been of much help?" Albert et al say probably not, and Stollerman agree[9,10]. Unfortunately committee for Jones' criteria is skeptical about including ECHO as a major criteria for the fear of over diagnosis of carditis[11]. Therefore there is a great need for appropriate ECHO criteria for the precise and early diagnosis of both clinical and subclinical valvulitis.

Background

As a pilot study[6]we performed echocardiography in 452 consecutive patients with

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acute rheumatic fever, clinically diagnosed by the strict Jones's criterions, using the patients as part of a multi-centric and double blinded prospective study. The youngest was aged 1 year 11 months, while the oldest was a 51-year-old female. Out of the 452 cases of ARF, diagnosed by Jones's criteria though 164 cases were clinically diagnosed as carditis, only 141 cases had echocardiographic evidence of carditis (85.97%). The remaining 23 patients (14%) had functional murmurs, tachycardia, or anaemia and 2 patients also had congenitally malformed hearts. Out of 40 patients with rheumatic chorea, 28 (70%) had echocardiographic evidence of carditis or valvulitis. Polyarthralgia was seen in 213 cases (47.12%), from which only 38 patients (17.8%) had carditis clinically, albeit that 88patients (41.3%) showed echocardiographic evidence of subclinical carditis or valvulitis. Echocardio-graphy, when carried out in patients with acute rheumatic fever diagnosed strictly according to the Jones criterion, can avoid both overdiagnosis and underdiagnosis of carditis. A high incidence of carditis, or subclinical carditis, is detected by echocardiography when performed in patients with rheumatic chorea or arthralgia.

Aim

Our study is to propose Vijay's echocardiographic criteria and test its efficacy to make precise and early diagnosis of carditis and/or subclinical valvulitis.

Material and Methods

The 333 consecutive, suspected cases of ARF, 165 males and 168 females (M:F Ratio 1:1), from January 2005 to June 2006, underwent a detailed clinical examination; laboratory tests and meticulous echocardiographic and Doppler study with Philips Sonos 5500 machine. The data was entered in specially designed database. The thickness of anterior mitral leaflet (AML), posterior mitral leaflet (PML), tricuspid valve (TV), aortic valve (AV) were measured in mm in parasternal long axis and apical four and five chamber views. Similarly measurements were taken by tissue harmonics. Also note was made whether the thickness of the valve was at the tip/base or through out the valve. Increased echogenicity of submitral structures, present or not, was noted. By M-Mode the excursion of AML and PML was recorded to know the mobility, whether normal/reduced or increased. Mitral valve prolapse (MVP) was looked for clinically and by ECHO. Whether the valve in MVP is thin, redundant, myxomatous with large excursion or thick valve with reduced mobility, were noted. Whether there is prolapse of AML / PML / both leaflets was noted. The chordal tear with rolled up chordae and flail valve was recorded. Similarly tricuspid and aortic valve were looked for TVP / AVP. The grade of mitral regurgitation (MR) clinically and by echocardiography was noted. Whether MR jet was central / eccentric and jet velocity was also noted. Chordal tear of AML / PML were looked for. Carefully auscultation was done for early diastolic murmur of aortic regurgitation (AR). ECHO Grade of AR was recorded. Similarly tricuspid regurgitation (TR) murmur was looked for clinically and ECHO Grade and Jet velocity were noted. Beaded appearance of mitral and tricuspid valves was carefully looked for. All the patients were carefully auscultated for pericardial rub and looked for mild / moderate/ severe pericardial effusion on ECHO. Chamber dimensions and Ejection Fraction (EF%) were noted in all. Vijay's ECHO criteria for the diagnosis of carditis / subclinical valvulitis evolved, depending on the various ECHO features observed from our previous study, as shown in Table I. Finally, total ECHO Score was calculated by giving 2 points for each of the eight ECHO features of carditis. The cases with an ECHO score > 6 out of 16 were taken as ECHO positive, so as to avoid over diagnosis of valvulitis. Mitral regurgitation, was taken as pathological only when colour mosaic jet persisted throughout systole and the jet was identified in at least two planes. The length of the colour jet was not necessarily > 1 cm in all cases, and were graded further as trivial, grade I, grade II and grade III. Aortic regurgitation and tricuspid regurgitation were also graded as trivial, grade I,

grade II and grade III. Mitral valve thickness was measured for both anterior mitral leaflet and posterior mitral leaflet in parasternal long axis view, with and without tissue harmonics, to find out the difference. Clinical echocardiographic, and laboratory data on the evolution of each episode were collected in specially designed computerized database and was analyzed systematically. Out of 333 suspected cases of ARF, 272 cases, that fulfilled the ECHO criteria for carditis / subclinical valvulitis, formed the material for this study. The 4 cases had various congenital heart disease and the remaining 57 cases were taken as control group.

Results

Out of 333 ECHO positive and Jones' positive were 220 (66.06%) [True +ve], 52 cases (15.61%), where clinically murmur was not heard but ECHO was positive. Though they were Jones' negative, were taken as probably, subclinical carditis presenting late, as biochemical parameters were negative [False - ve]. 4 cases though, clinically were diagnosed as carditis and were +ve by Jones' criteria , actually ECHO showed, one case each of Atrial Septal Defect, myxomatous MVP with mitral regurgitation, aorto arteritis with mitral regurgitation, and subaortic membrane [False positive]. Remaining 57 cases (17.1%) who, were Jones'-ve and clinically as well as echocardiographically negative were taken as control group (True –ve). Sensitivity of Vijay's ECHO criteria is 81% and specificity is 93%.

The various ECHO features observed and their incidence are given below in table II. Out of 239 cases of mitral regurgitation detected by ECHO, only 144 cases had systolic murmur on auscultation. Aortic regurgitation was detected in 60 patients on ECHO, whereas in only 11 cases the early diastolic murmur was heard clinically. In 60 cases, tricuspid regurgitation was detected by ECHO, but systolic murmur of tricuspid regurgitation was heard only in 9 cases.

Increased AML thickness is the commonest feature found in 279 cases (83.8%). The valve thickness (Figure-1) is more often detected (310 cases-93%), if tissue harmonics is used. The PML thickness was found in 188 cases (56.5%), whereas tissue harmonics increased the yield to 250 cases (75%).Though the AML is thickened more often, the reduced mobility is seen more in PML (121 cases-36%). Reduced mobility of AML is seen less frequently (55 cases-16.5%). Rheumatic nodules or beaded appearance (Figure-2 A,B,C), was seen in 159 cases (47.7%). MVP with thickened valve was seen in 202 patients (83.69%), while TVP was seen in only 42 (12.6%). Out of 5 cases of torn chordae (01.5%), 3 cases had severe mitral regurgitation due to flail valve (0.9%). Though pericardial rub was not heard clinically, pericardial effusion was found in 12 cases (3.6%).

Discussion

More than 50% of RHD detected in surveys and health check up camps are unaware of the disease! More than 70% do not receive secondary prophylaxis (WHO Bulletin 1981) [6]because carditis especially subclinical carditis or valvulitis is mostly missed by the best of the clinicians. Detection of active rheumatic carditis is of great prognostic and therapeutic importance and is currently based on the Jones' criteria[7]. Kaplan EL in the editorial says -Rheumatic fever and rheumatic heart disease continue to be a problem for medical and public health communities- the fact that penicillin has failed to eradicate this disease process is irrefutable proof of the need for more laboratory, epidemiological and clinical research[12]. In fact, it is the lack of penicillin prophylaxis rather than the failure of penicillin . If this large chunk of patients with subclinical carditis could be detected and put on penicillin prophylaxis, one can prevent recrudescence of rheumatic activity and further damage to the valves. Our study clearly shows that a simple non-invasive, reproducible ECHO can fill this grey zone effectively.

Hence, it is essential to realize that precise and early diagnosis of carditis in ARF is very pertinent. The diagnosis of carditis in ARF, traditionally depends on characteristic auscultatory findings like (i) detection of new murmur not present earlier (ii) cardiomegaly (iii) congestive cardiac failure (CCF) (iv) pericardial rub / effusion. This could be unreliable, because if the patient has

come for the first time, how to make out it is a new murmur and it did not exist earlier? Cardiomegaly could be due to any other cause like congenital heart disease, aortoarteritis or dilated cardiomyopathy etc. CCF could be due to any other cause, not necessarily ARF and pericardial rub / effusion could be due to tubercular pericarditis, especially in our country. Hence, clinically there is no way that one could be sure of rheumatic carditis, especially in an era when cardiac auscultation has been taught less extensively and is used with less confidence by young clinicians! In our own experience, a 10 year old boy diagnosed to have ARF with carditis was put on steroids by pediatrician, developed tubercular meningitis due to flaring of primary complex. On ECHO he was found to have a congenital subaortic membrane! Rheumatic carditis is almost always associated with valvulitis. The diagnosis of carditis therefore remains a problem and the solution is obviously not more laboratory tests or the formulation of yet another set of clinical criteria. Because valvulitis constitutes the sine qua non of rheumatic carditis, echocardio-graphic documentation of valvular and subvalvar changes should, theoretically, be of significant help. Often asked question is 'Does Echocardiography perform better than clinical examination in the detection of carditis?' The answer is 'Yes'. Our study showed that mitral regurgitation was clinically detected in only 144 cases, where as ECHO picked it up in 239 patients. This means that 95 cases would not have received penicillin prophylaxis but for ECHO. Similarly early diastolic murmur of aortic regurgitation was detected clinically only in 11 cases, but ECHO picked up in 60 cases and so also the systolic murmur of tricuspid regurgitation was clinically detected in only 9 cases, but ECHO picked it up in 60 patients. It is not that we are over diagnosing the physiological regurgitation. Because according to Vijay's ECHO criteria, unless we have three or more parameters like thickened valve / beaded appearance / reduced mobility / increased echogenicity of sub mitral structures etc we do not take regurgitation as rheumatic. For example one patient with grade II mitral regurgitation was clinically diagnosed as carditis, did not fulfill the criteria as other features were not there. That case turned out to be aortoarteritis. Similarly, another clinically diagnosed case of carditis with MVP and grade II mitral regurgitation had myxomatous redundant valve with grade III MVP, the ECHO score was 4 (Figure - III). Yet another case though clinically diagnosed as carditis with MR had atrial septal defect with MVP, there was mitral regurgitation by ECHO. The ECHO not only helps to make precise diagnosis but also helps in management strategy when severe MR is due to chordal tear (Figure- IV).

Cotrim [11], conclude that early ECHO is very important in all children suspected to have ARF, especially, because mitral regurgitation can be demonstrated by colour flow mapping in absence of cardiac murmur (Figure V). Neutze et al[12], feel pulse and colour Doppler ECHO provide a method to detect minor degree of pathological regurgitation without characteristic clinical signs. Doppler echocardiography is more sensitive than clinical assessment in the detection of carditis in acute rheumatic fever, and can contribute to earlier diagnosis [16]. In fact addition of ECHO features in the criteria for diagnosis, not only prevents over diagnosis of carditis by clinicians but also helps in detecting the subclinical cases of carditis / valvulitis that would otherwise go undetected and would not receive secondary prophylaxis, due to absence of Jones' criteria. As in our study, 108 cases had subclinical carditis detected on ECHO by the ECHO criteria out of which 56 were Jones's +ve and 52 cases were Jones's negative. Probably these large groups of patients are the ones who present later as RHD, without the past history of ARF and secondary prophylaxis. The cost of ECHO at the beginning of the disease and the cost of penicillin prophylaxis is negligible, when compared to the human suffering and cost of management of RHD with or without surgery.

Vijay's ECHO criteria plays an important role in early, precise diagnosis of carditis / subclinical valvulitis .The sensitivity of this criteria is 81% and specificity is 93%. Therefore, apart

from valvular regurgitations, other features of rheumatic carditis in ECHO and colour Doppler findings should be accepted as a major criterion for the diagnosis of rheumatic fever. Therefore, there is no doubt that if echocardiography is used as a primary diagnostic modality and is included in Jones' criteria it will change the epidemiological face of ARF and RHD completely. However, long-term follow-up is necessary to determine the outcome for young children with subclinical echocardiographic evidence of valvular disease[17].

Conclusions

Early and precise diagnosis of carditis in ARF, though difficult is very important to prevent the serious consequences, morbidity and mortality in young. Vijay's ECHO criteria plays an important role in precise diagnosis of carditis / subclinical valvulitis. These subclinical changes detected only by ECHO, can persist and probably belong to a large group of patients who present later as RHD, without the past history of ARF and prophylaxis. Therefore ECHO should be included as a major criterion in Jones' criteria

Table-I Vijay's Echo Criteria

Sl.No	Echo feature	Score
1	MV and AV thickness ≥ 4mm	2
2	Increased Echogenicity of Submitral structures	2
3	Rheumatic nodules (beaded appearance)	2
4	Mitral valve prolapse (MVP)/AVP/TVP	2
5	MV regurgitation and / AVR/TVR	2
6	Reduced mobility of valves	2
7	Chordal tear	2
8	Pericardial Effusion	2
	Total Score	**16**

Score >6 is Diagnostic of Rheumatic Carditis

Table-II Showing type o cardiac involvement by ECHO

Sl. No.	Type of Involvement	No. of Cases	%
1a.	Mitral valve thickness ≥ 4 mm		
	anterior mitral leaflet (AML)	279	83.8%
	posterior mitral leaflet (PML)	188	56.5%
1b.	Mitral valve thickness by tissue harmonics		
	Anterior mitral leaflet (AML)	310	93.0%
	Posterior mitral leaflet (PML)	250	75.0%
2.	Increased echogenicity of submitral structures	230	75.0%
3.	Reduced mobility		
	Anterior mitral leaflet (AML)	55	16.5%
	Posterior mitral leaflet (PML)	121	36.3%
4.	Mitral valve prolapse (MVP)	202	83.69%
	Flail valve	3	00.9%
	TVP	42	12.6%
5.	Rheumatic nodules (Beaded appearance)	159	47.7%
6a.	Mitral regurgitation		
	Grade trivial	72	21.6%
	Grade I	48	17.6%
	Grade II	56	16.8%
	Grade III	63	18.9%
6b.	Aortic regurgitation (AR)		
	Grade trivial	11	03.3%
	Grade I	23	06.9%
	Grade II	13	03.9%
	Grade III	13	03.9%
6c.	Tricuspid regurgitation (TR)		
	Grade trivial	18	05.4%
	Grade I	31	09.3%
	Grade II	11	03.3%
7.	Pericardial effusion (PE)	12	03.6%
8	Chordal tear	5	01.5%

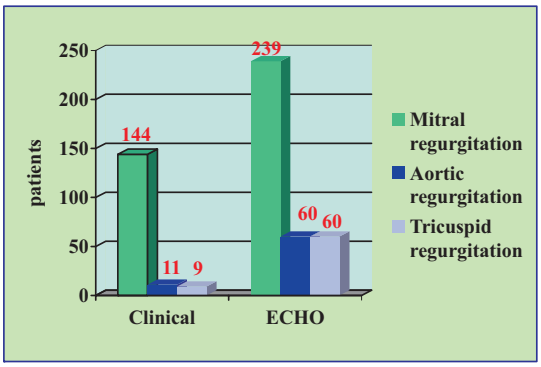

Table III – Clinical and ECHO correlation in mitral, aortic and tricuspid regurgitation

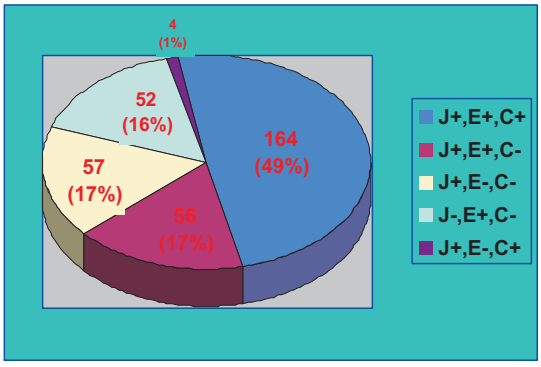

Table IV J= Jones criteria, E = Echo criteria, C= Clinical criteria

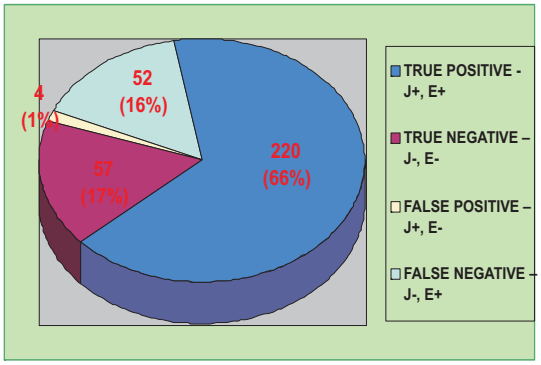

Table V

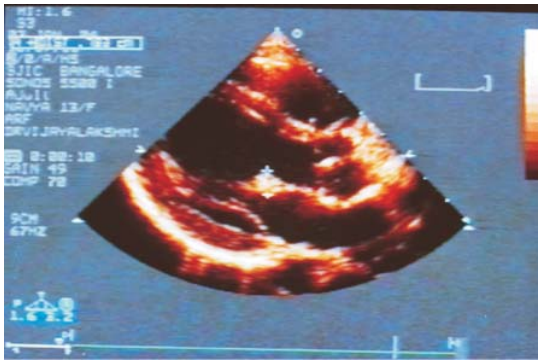

Figure I- ECHO in PLAX view shows thickened AML (6mm)

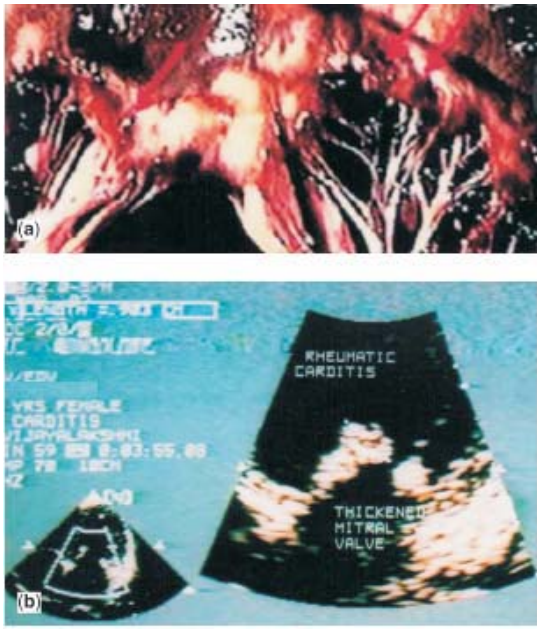

Figure II Mitral valvar verrucous nodules as seen (a) at autopsy, and com-pared (b) to an echocardiogram showing nodules on the mitral leaflets

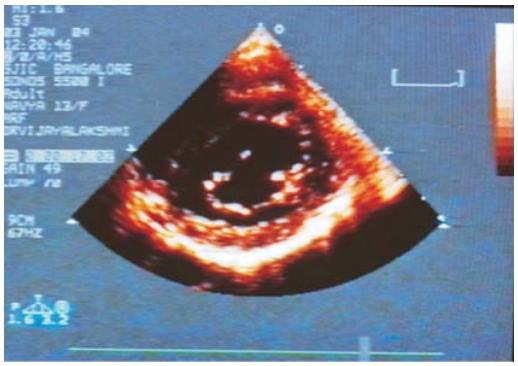

Figure II C -Short axis view shows beaded appearance of mitral valve

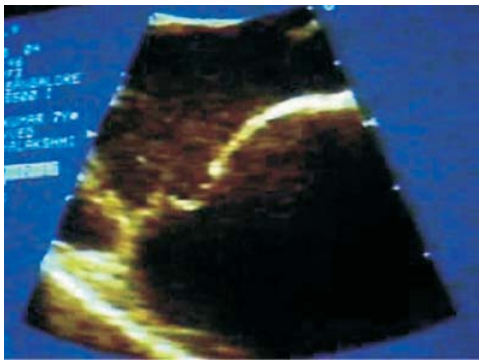

Figure III- 12 years old boy with Mitral regurgitation. ECHO-Grade III MVP with Myxomatous redundant valve with MR. The ECHO Score - 4 (MVP-2, MR-2)

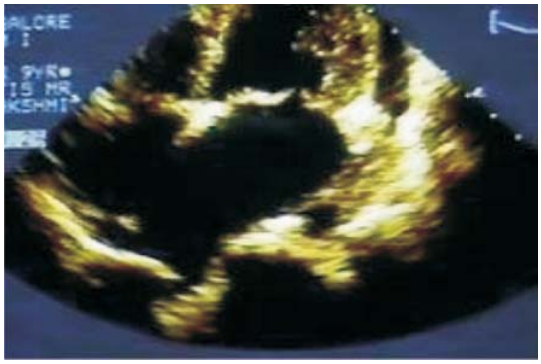

Figure IV- 10 years old girl with Mitral regurgitation. Apical four chamber view shows MVP, PE, MR with chordal tear ECHO Score-10 (MVP-2,MR-2, MV thick-2, PE-2 Chordal tear-2).

Figure V - Pancarditis with mitral regurgitation.

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