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*Editorial Comment*

# Echocardiography and subclinical carditis: guidelines that increase sensitivity for acute rheumatic fever

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ECHOCARDIOGRAPHY HAS BEEN THE PIVOTAL INVESTIgation used by cardiologists assessing cardiac valvar disease for the last three decades. It is diagnostic and often prognostic whatever the aetiology of the cardiac valvar disease. Echocardiography is more accurate than clinical evaluation by experienced cardiologists.[1] In rheumatic cardiac disease echocardiography is the key investigation for serial assessment of valvar and ventricular function.

The entity of subclinical valvar regurgitation, defined as pathological mitral or aortic regurgitation inaudible to skilled auscultation, is not a clinical concern in most cardiac disease as these lesions are not of haemodynamic importance. However in acute rheumatic fever, subclinical carditis is of considerable diagnostic importance, as it may be the only manifestation of carditis in acute rheumatic fever.

There is convincing evidence that subclinical or silent echocardiographic carditis is part of the spectrum of rheumatic carditis and should not be ignored. It has been found by all investigators in regions of the world with endemic rheumatic fever, including Europe[2,3] the Middle East,[4] Asia,[5–7] Oceania,[8–12] North America,[13–15] and South America.[16–20] Three of the above publications specifically address follow-up of subclinical carditis,[17,19,20] confirming that it persists in similar proportions to that of mild clinical carditis.[10,14,21] Not surprisingly many of these authors advocate that subclinical carditis is incorporated into the Jones criteria as evidence of carditis. This is relevant globally as 80% of the world's children still live in regions with high prevalence of rheumatic fever and rheumatic cardiac disease.[22]

Duckett Jones proposed the guidelines for acute rheumatic fever in 1944,[23] at which time echocardiography did not exist. Successive revisions and updates of the Jones criteria by the American Heart Association have occurred, each change aiming to increase the specificity of the criteria but decrease sensitivity. Before the last major review of the guidelines in 1992,[24] there were few reports of subclinical carditis but there were many by 2002,[25] when once again subclinical carditis was not endorsed as a major criterion for acute rheumatic fever. The American Heart Association committee may not have appreciated that it is not just the lower end of mild valvar regurgitation that is subclinical, but in the acute phase even moderate mitral or aortic regurgitation may be inaudible with a stethoscope. The committee was also comprised mainly of non-cardiologists.

Today echocardiography is used to its fullest extent by cardiologists to assess not only subclinical carditis but clinical carditis by defining myocarditis, pericarditis and the severity of valvulitis. Echocardiography is used to visualize the anatomy of the mitral and aortic valves, defining rheumatic structural changes as well as the severity of valvar regurgitation, especially helpful in the presence of mixed valvar disease including tricuspid valvulitis. Echocardiography will confirm the presence of a pericardial effusion in suspected pericarditis, and may reveal valvar regurgitation that is inaudible due to the pericardial fiction rub. It defines ventricular function as a measure of myocardial involvement in carditis. Portable echocardiograms and mobile echocardiographers should enable use of echocardiography to be introduced into more regions of the world, even in countries with very low income.

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Table 1. Echocardiographic criterions for subclinical carditis.

| Minimal echocardiographic criterions to allow a diagnosis of pathological valvar regurgitation* |
|---|
| *Aortic regurgitation* |
| Colour: |
| Substantial **colour jet seen in 2 planes extending greater than or equal to 1 cm beyond the valve leaflets. |
| Continuous wave or pulsed Doppler: |
| Holodiastolic with well defined high velocity spectral envelope. |
| *Mitral regurgitation* |
| Colour: |
| Substantial colour jet seen in 2 planes extending greater than or equal to 2 cm beyond the valve leaflets. |
| Continuous wave or pulsed Doppler: |
| Holosystolic with well defined high velocity spectral envelope. |

*From the NZ RF guidelines Table 6[28] Reproduced with permission. **The WHO suggested a minimal jet length of 1 cm from the mitral valve leaflets[32] but experience shows that many central 1 cm colour jets will not have a well defined high velocity spectral envelope. Most 2 cm long colour jets usually do meet these Doppler requirements and this jet length is more likely to avoid over diagnosis. However, posterior jets of less than 2 cm hitting the back wall of the left atrium may then dissipate out of the echocardiographic plane yet clearly be "substantial" and pathological, hence the importance of assessment in more than one echocardiographic plane.

The minimal criteria for a diagnosis of abnormal mitral or aortic valvar regurgitation are summarised in Table 1. To be classified as pathological, both the colour and Doppler signals must be holodiastolic for aortic regurgitation, or holosystolic for mitral regurgitation. The Doppler signal must be a high velocity signal with a well-defined spectral envelope, using pulsed or continuous wave. Those experienced in echocardiography in areas with endemic rheumatic fever agree that these features can usually readily separate the normal child with a small colour jet of physiological regurgitation from the child with pathological regurgitation.[2–4,8,9,14–20]

There are now guidelines incorporating subclinical carditis as a major manifestation for acute rheumatic fever in New Zealand[26] and Australia.[27] The full guidelines can be found on the world wide web through these countries' national Heart Foundations.[28,29]

The guidelines are evidence-based, and were developed by consensus expert opinion with wide endorsement by Australian and New Zealand health organisations. As there is no laboratory test for the diagnosis of acute rheumatic fever, it is not possible to prove scientifically that a new set of clinical criterions are more accurate than a previous set. The current article by Vijayalakshmi and colleagues[30] has confirmed that the current Jones criterions lack sensitivity for the presence of carditis in acute rheumatic fever. Fifty-two of 333 (16%) more patients were diagnosed as having definite acute rheumatic fever than would have been using the American Heart Association Jones criterions. This is similar to a recent experience in New Zealand, where subclinical carditis influenced the diagnosis to definite acute rheumatic fever in 11% more cases than using the American Heart Association criterions even though it was present in 31% of cases.[31] Vijayalakshmi and colleagues[30] also add a requirement that valvar morphological features should also be present to diagnose subclinical carditis. This will help increase specificity and will help avoid over-diagnosis. Morphologic changes, however, are not always present early in acute rheumatic fever, so most authors have not recommended them as a requirement for the diagnosis of subclinical carditis.

Consider where a clinician makes the diagnosis of acute rheumatic fever based on the auscultatory findings of a systolic murmur misjudged to be mitral regurgitation, but subsequently found to have no mitral regurgitation by echocardiography. Clinicians have had no hesitation in correcting their false positive diagnosis of carditis. It would be unethical to subject a child to years of prophylaxis with penicillin based on incorrect auscultatory findings. Yet this situation has undoubtedly occurred since the inception of the Jones criterions[32] and still occurs where echocardiography is not available. Clinicians are not as accurate as echocardiography.[1,33] The likelihood of false positive misclassification is even higher now if echocardiography is not utilized, as young physicians are less proficient at auscultation than in earlier years.[34] Cardiologists have frequently observed that flow murmurs are mistakenly diagnosed as mitral regurgitation by hospital clinicians assessing children considered to have acute rheumatic fever.

Consider the converse, that of false negative diagnosis. When a child has subclinical carditis its use will avoid under-diagnosis of acute rheumatic fever if the child has no other major criterion of acute rheumatic fever. Experienced clinicians in countries with endemic rheumatic fever are not radically changing the Jones criterions, rather, they are applying a more sensitive and more accurate tool for the diagnosis of carditis. As noted above, using subclinical carditis as a major Jones manifestation influences the diagnosis of definite acute rheumatic fever in an extra 11–16% only. In the majority of patients the major criterion of acute rheumatic fever is either polyarthritis, clinically overt carditis or chorea. Subclinical carditis does play a role of helping to reinforce to families the diagnosis of acute rheumatic fever and need for secondary prophylaxis with penicillin.

## Atypical arthritis

Successive versions of the American Heart Association Jones criteria have emphasized that arthritis is a migratory polyarthritis that is extremely painful in its classical presentation. However, many arthritic symptoms of acute rheumatic fever are atypical, and monoarthritis is frequently observed.[35–37] All too often individuals who did not fulfil the Jones criterions of migratory polyarthritis, and who were not started on prophylaxis with penicillin, have returned with a recurrence of acute rheumatic fever and significant cardiac disease. Atypical presentations of arthritis are not accepted as major manifestations of acute rheumatic fever by the American Heart Association,[25] but the 1992 update[24] did describe that where migratory polyarthritis may have been "aborted by premature administration" of anti-inflammatory medication this be regarded as a major criterion. The Australian and New Zealand guidelines have thus also widened the spectrum of arthritis as a major criterion for the diagnosis of acute rheumatic fever.[28,29]

Recognising the migratory and evanescent nature of arthritis, the guidelines accept a history of arthritis rather than just documentation by a clinician to satisfy the criterion of arthritis. Moreover, in high risk populations in Australia it has been documented that aseptic mono-arthritis or polyarthralgia are common manifestations of acute rheumatic fever.[36] They are accepted as a major criterion for acute rheumatic fever, with the proviso that alternative causes are sought.[29] In New Zealand, mono-arthritis is accepted as a major criterion only when anti-inflammatory medication has been given early in the illness.[28,38] It is worth remembering that Duckett Jones proposed that polyarthralgia be a major manifestation when acute rheumatic fever was endemic in the USA in 1944.[23] Some clinicians have the adage that a child who presents with arthritis and a fever have acute rheumatic fever until proved otherwise. Septic arthritis must of course be excluded urgently in those with a mono-arthritis who are toxic with a high fever.

## Conclusions

Regions with high prevalence of rheumatic fever should broaden the definition of carditis by using accurate echocardiography to its fullest extent, and accept subclinical carditis as a major manifestation of acute rheumatic fever. Similarly, mono-arthritis and polyarthralgia may be manifestations of acute rheumatic fever. Guidelines that increase the sensitivity for acute rheumatic fever will help avoid under-diagnosis of acute rheumatic fever compared to the 1992 Jones criterions. This is an issue for about 80% of the world's population. The benefits of secondary prophylaxis with penicillin to prevent recurrences of acute rheumatic fever and worsening carditis have been known for decades.[22] The ultimate aim is to reduce the burden of significant rheumatic cardiac disease for both the individual and the population.

## References


1. Jaffe WM, Roche AH, Coverdale HA, McAlister HF, Ormiston JA, Greene ER. Clinical evaluation versus Doppler echocardiography in the quantitative assessment of valvar heart disease. Circulation 1988; 78: 267–275.
2. Karaaslan S, Demiroren S, Oran B, Baysal T, Baspinar O, Ucar C. Criteria for judging the improvement in subclinical rheumatic valvitis. Cardiol Young 2003; 13: 500–505.
3. Cotrim C, Macedo AJ, Duarte J, Lima M. The echocardiogram in the first attack of rheumatic fever in childhood. Rev Port Cardiol 1994; 13: 581–586.
4. Folger GM Jr, Hajar R, Robida A, Hajar HA. Occurrence of valvar heart disease in acute rheumatic fever without evident carditis: colour-flow Doppler identification. Br Heart J 1992; 67: 434–438.
5. Agarwal PK, Misra M, Sarkari NB, Gupta AK, Agarwal P. Usefulness of echocardiography in detection of subclinical carditis in acute rheumatic polyarthritis and rheumatic chorea. J Assoc Physicians India 1998; 46: 937–938.
6. Vijaykumar M, Narula J, Reddy KS, Kaplan EL. Incidence of rheumatic fever and prevalence of rheumatic heart disease in India. Int J Cardiol 1994; 43: 221–228.
7. Regmi PR, Pandey MR. Prevalence of rheumatic fever and rheumatic heart disease in school children of Kathmandu city. Indian Heart J 1997; 49: 518–520.
8. Abernethy M, Bass N, Sharpe N, et al. Doppler echocardiography and the early diagnosis of carditis in acute rheumatic fever. Aust N Z J Med 1994; 24: 530–535.
9. Wilson NJ, Neutze JM. Echocardiographic diagnosis of subclinical carditis in acute rheumatic fever. Int J Cardiol 1995; 50: 1–6.
10. Voss LM, Wilson NJ, Neutze JM, et al. Intravenous immunoglobulin in acute rheumatic fever: a randomized controlled trial. Circulation 2001; 103: 401–406.
11. Ralph A, Jacups S, McGough K, McDonald M, Currie BJ. The challenge of acute rheumatic fever diagnosis in a high-incidence population: a prospective study and proposed guidelines for diagnosis in Australia's Northern Territory. Heart Lung Circ 2006; 15: 113–118.
12. Maheu B, Costes P, Lionet P, et al. Contribution of Doppler echocardiography to the diagnosis of the first attack of acute rheumatic fever. Arch Mal Coeur Vaiss 1995; 88: 1833–1839.
13. 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.
14. Veasy LG, Tani LY, Hill HR. Persistence of acute rheumatic fever in the intermountain area of the United States [see comment]. J Pediatr 1994; 124: 9–16.
15. Minich LL, Tani LY, Pagotto LT, Shaddy RE, Veasy LG. Doppler echocardiography distinguishes between physiologic and pathologic "silent" mitral regurgitation in patients with rheumatic fever. Clin Cardiol 1997; 20: 924–926.
16. Mota CC. Doppler echocardiographic assessment of subclinical valvitis in the diagnosis of acute rheumatic fever. Cardiol Young 2001; 11: 251–254.
17. Lanna CCD, Tonelli E, Barros MVL, Goulart EMA, Mota CCC. Subclinical rheumatic valvitis: a long-term follow-up. Cardiol Young 2003; 13: 431–438.

18. Caldas AM, Terreri MT, Moises VA, Silva CM, Carvalho AC, Hilario MO. The case for utilizing more strict quantitative Doppler echocardiographic criterions for diagnosis of subclinical rheumatic carditis. Cardiol Young 2007; 17: 42–47.
19. Figueroa FE, Fernandez MS, Valdes P, et al. Prospective comparison of clinical and echocardiographic diagnosis of rheumatic carditis: long term follow up of patients with subclinical disease [see comment]. Heart 2001; 85: 407–410.
20. Hilario MO, Andrade JL, Gasparian AB, Carvalho AC, Andrade CT, Len CA. The value of echocardiography in the diagnosis and followup of rheumatic carditis in children and adolescents: a 2 year prospective study. J Rheumatol 2000; 27: 1082–1086.
21. Wilson NJ, Voss LM, Neutze JM, Ameratunga RV, Lennon DR. The natural history of acute rheumatic fever to one year in the echocardiographic era. Proceedings of the 2nd World Congress of Paediatric Cardiology and Cardiac Surgery. Futura Publishing Co, New York, 1997, pp 971–972.
22. Carapetis JR. Rheumatic heart disease in developing countries. N Engl J Med 2007; 357: 439–441.
23. Jones TD. Diagnosis of rheuamatic fever. Jama 1944; 126: 481–484.
24. Guidelines for the diagnosis of rheumatic fever. Jones Criteria, 1992 update. Special Writing Group of the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the Council on Cardiovascular Disease in the Young of the American Heart Association. Jama 1992; 268: 2069–2073.
25. Ferrieri P. Proceedings of the Jones Criteria workshop. Circulation 2002; 106: 2521–2523.
26. Atatoa-Carr P, Lennon D, Wilson N. Rheumatic fever diagnosis, management, and secondary prevention: a New Zealand guideline. N Z Med J 2008; 121: 59–69.
27. Carapetis JR, Brown A, Wilson NJ, Edwards KN. An Australian guideline for rheumatic fever and rheumatic heart disease: an abridged outline. Med J Aust 2007; 186: 581–586.
28. New Zealand Guidelines for Rheumatic fever 1. Diagnosis, Management and Secondary prevention. Heart Foundation of New Zealand, Cardiac Society of Australia and New Zealand. 2006 [cited; Available from: http://www.nhf.org.nz].
29. National Heart Foundation of Australia (RF/RHD guideline development working group) and the Cardiac Society of Australia and New Zealand. Diagnosis and management of acute rheumatic fever and rheumatic heart disease in Australia – an evidence based review. 2006 [cited; Available from: http://www.heartfoundation.com.au].
30. Vijayalakshmi IB, Vishnuprabhu RO, Chitra N, Rajasri R, Anuradha TV. The efficacy of echocardiographic criterions for the diagnosis of carditis in acute rheumatic fever. Cardiol Young 2008; 18: in press.
31. Wilson NJ, Morreau J, Voss L, Stewart J, Lennon D. The influence of subclinical carditis on the diagnosis of acute rheumatic fever. Heart Lung Circ 2005; 14 (Suppl 1): S1117.
32. Rheumatic fever and rheumatic heart disease. World Health Organization technical report series 2004: 1–122.
33. Wilson NJ, Neutze JM. Echocardiographic diagnosis of mitral insufficiency [comment]. J Pediatr 1994; 125: 673–674.
34. Mahnke CB, Nowalk A, Hofkosh D, Zuberbuhler JR, Law YM. Comparison of two educational interventions on pediatric resident auscultation skills. Pediatrics 2004; 113: 1331–1335.
35. Harlan GA TL, Byington CL. Rheumatic fever presenting as monoarticular arthritis. Pediatr Infect Dis J 2006; 25: 743–746.
36. Carapetis JR, Currie BJ. Rheumatic fever in a high incidence population: the importance of monoarthritis and low grade fever. Arch Dis Child 2001; 85: 223–227.
37. Pileggi GC, Ferriani VP. Atypical arthritis in children with rheumatic fever. Jornal de Pediatria 2000; 76: 49–54.
38. Wilson E, Wilson N, Voss L, Morreau J, Lennon D. Monoarthritis in rheumatic fever? [comment]. Pediatr Infect Dis J 2007; 26: 369–370.