The challenge of screening for asymptomatic rheumatic heart disease in South Africa

Liesl Zühlke* and Bongani M. Mayosi*

*Western Cape Paediatric Cardiac Services, Red Cross War Memorial Children’s Hospital and Tygerberg Hospital, Universities of Cape Town and Stellenbosch, Cape Town

*Department of Medicine, Groote Schuur Hospital and University of Cape, Cape Town

Address for correspondence:
Professor B.M. Mayosi
Department of Medicine
J Floor, Old Main Building
Groote Schuur Hospital
Observatory
7925
South Africa

Email: bongani.mayosi@uct.ac.za

Acute rheumatic fever and rheumatic heart disease remain common in the population of South Africa. A recent screening study of asymptomatic schoolchildren in Mozambique and Cambodia makes a compelling case for a shift in the approach to screening for rheumatic heart disease from auscultation to portable echocardiography. Rheumatic heart disease meets all the epidemiological criteria for screening in the South African population. The incorporation of echocardiographic screening programmes into the school health system and in antenatal clinics for the pre-symptomatic diagnosis of rheumatic heart disease could result in the reduction of morbidity and mortality through the early and wide application of secondary antibiotic prophylaxis. SAHeart 2009; 6:100-103
A series of echocardiographic screening studies confirm that rheumatic fever and rheumatic heart disease are of sufficient importance to warrant the urgent attention of the international public health and research communities. These echocardiographic screening studies have major implications for clinical and public health practice in South Africa. These data raise questions on whether screening for asymptomatic rheumatic heart disease is required, how it should be conducted, what diagnostic criteria should be used, and the cost-effectiveness of screening for asymptomatic rheumatic heart disease.

RATIONALÉ FOR SCREENING ASYMPTOMATIC PEOPLE FOR RHEUMATIC HEART DISEASE

For a disease to be suitable for screening as a tool for prevention, it must satisfy the following conditions: (1) there must be evidence of a significant burden of disease, (2) the condition must have an initial latent stage, (3) which can be detected by simple, accessible, sensitive and specific tests, and (4) there must be evidence that early intervention improves prognosis. It can be argued that rheumatic heart disease meets all the criteria for screening in many developing countries such as South Africa. There is evidence that the condition may affect up to 3% of school children; that large numbers of pre-symptomatic cases exist, that portable echocardiography is a sensitive tool for screening; and that the early introduction of penicillin prophylaxis prevents recurrent attacks of acute rheumatic fever.

A 2002 report from a paediatric cardiology workshop highlights the belief among clinicians that South Africa continues to experience a high burden of cases of symptomatic rheumatic heart disease. Recent reports show that rheumatic heart disease is present in 1% of pregnant patients presenting to tertiary centres and is associated with high maternal morbidity and mortality and poor foetal outcomes.

Clinical and echocardiographic screening studies indicate that the overwhelming majority of cases (over 85%) with definite rheumatic heart disease are asymptomatic. These observations indicate that the disease has a pre-symptomatic stage that can be detected by portable echocardiography and penicillin prophylaxis is available to prevent recurrent attacks of rheumatic fever and potentially ameliorate the development of chronic rheumatic heart disease. There is therefore a strong case to be made for the screening of the general population for rheumatic heart disease in South Africa and other countries where the disease remains endemic.

WHAT IS THE APPROPRIATE METHOD FOR SCREENING ASYMPTOMATIC PEOPLE FOR RHEUMATIC HEART DISEASE?

Several studies present a compelling argument for the use of portable echocardiography as the screening test of choice for asymptomatic individuals who are at high risk of rheumatic heart disease, as opposed to screening by cardiac auscultation. If clinical diagnosis had been relied upon in the Mozambique study, approximately 90% of echocardiographically detected cases would have been missed. In the Mozambican study, clinical examination underdiagnosed rheumatic heart disease more than 10 fold, whereas in the Tongan study, the opposite held in that junior auscultators overestimated rheumatic heart disease. This variability in the performance of cardiac auscultation in screening probably reflects differences in the clinical skill and experience of the clinicians, a factor that is not likely to improve in an era of over-reliance on technology and tests in clinical medicine.

The time has come to replace the stethoscope with portable cardiac ultrasound in screening for rheumatic heart disease. The cost of portable ultrasound equipment is falling and the technical capability of these devices is improving. There is therefore a need to increase the availability and use of portable echocardiography in endemic regions of the world. In addition the feasibility of training a large number of local healthcare workers in basic echocardiography to screen for rheumatic heart disease should be investigated.

DIAGNOSTIC CRITERIA FOR RHEUMATIC HEART DISEASE IN ASYMPTOMATIC INDIVIDUALS

Until now, the diagnosis of rheumatic heart disease in asymptomatic individuals has been based on the detection of pathological murmur in combination with typical echocardiographic morphological changes and functional abnormalities (e.g., Doppler regurgitation or stenosis) of the heart valves. The patients with no history of acute rheumatic fever nor a clinically audible murmur but who have typical structural and functional abnormalities of rheumatic heart disease on echocardiography are regarded as probable rheumatic heart disease. Individuals with isolated Doppler regurgitation on echocardiography with no history of rheumatic fever, no audible murmur, and no structural valve abnormality are regarded as possible rheumatic heart
disease. The natural history of possible and probable rheumatic heart disease (i.e., subclinical rheumatic heart disease) is not known.\(^{(17)}\) In addition, it is not known whether penicillin prophylaxis is indicated in subjects with subclinical rheumatic heart disease.\(^{(19)}\) The World Health Organisation, however, recommends antibiotic prophylaxis for those with “significant” subclinical rheumatic mitral regurgitation, who are defined on the basis of the following echocardiographic criteria: (1) the presence of a colour jet of more than 1 centimetre in length, (2) that is evident in at least two imaging planes, (3) with the mosaic jet having a peak velocity of greater than 2.5 metres per second, and that the Doppler signal is holosystolic.\(^{(19,20)}\) The WHO criteria differentiates physiological from pathological regurgitation, but these criteria do not cover many of the morphological changes seen in chronic rheumatic heart disease. Investigators in the A.S.A.P. programme propose to use an amalgamation of the WHO criteria with the findings of Viyashlaskmi and others to detect definite, probable and possible disease on screening echocardiograms.\(^{(20)}\) This approach allows the classification of cases into definite, probable and possible rheumatic heart disease using criteria that consider all possible functional and structural abnormalities that are associated with rheumatic fever and rheumatic heart disease (Tables 1 and 2).

**COST-EFFECTIVENESS OF SCREENING FOR ASYMPOTOMATIC RHEUMATIC HEART DISEASE**

There is good evidence that primary prevention (i.e., treatment of suspected streptococcal pharyngitis with penicillin) and secondary prophylaxis (i.e., regular intramuscular penicillin injections) for rheumatic fever are cost-effective interventions for the control of rheumatic heart disease.\(^{(13,21)}\) Secondary prophylaxis is best delivered as part of a register-based control programme, providing education and enabling better clinical follow-up.\(^{(22)}\) It is not known, however, whether the use of portable echocardiography to screen asymptomatic people in rheumatic heart disease-endemic regions is cost-effective.\(^{(23)}\) The major costs are related to staff, equipment, and other health service costs of community-based screening. One way of reducing the costs related to screening may be to integrate rheumatic heart disease screening with the existing school health system and antenatal care services for school children and pregnant women, the two high-risk groups who may be targeted initially.

**TABLE 1: Proposed echocardiographic diagnostic criteria for rheumatic heart disease used in screening studies of asymptomatic participants without a history of rheumatic fever**

<table>
<thead>
<tr>
<th><strong>Definite RHD</strong></th>
<th><strong>Probable RHD</strong></th>
<th><strong>Possible RHD</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Significant mitral stenosis (mean gradient: &gt;4 mm Hg)</td>
<td>Significant structural and/or functional changes involving both mitral and aortic valves, i.e., multiple valve disease</td>
<td>Isolated structural OR functional changes involving either mitral or aortic valve</td>
</tr>
</tbody>
</table>

**TABLE 2: Definitions of Structural and Functional changes in rheumatic heart disease**

<table>
<thead>
<tr>
<th><strong>Significant structural changes:</strong></th>
<th><strong>Significant functional changes:</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Thickness of mitral and aortic leaflets greater than 4 mm</td>
<td>Significant mitral regurgitation: defined as a mitral regurgitant jet at least 1 cm from the coaptation point of the valve leaflets, seen in two planes, high velocity (mosaic pattern) and persisting throughout systole. Additional changes that may be present include multiple regurgitant jets and/or a posterolaterally-directed jet</td>
</tr>
<tr>
<td>Increased echogenicity of submitral structures</td>
<td>Significant aortic regurgitation: defined as an aortic regurgitant jet at least 1 cm from the coaptation point of the valve leaflets, of high velocity (mosaic pattern) and seen in two planes</td>
</tr>
<tr>
<td>Rheumatic nodules giving a beaded appearance</td>
<td>Elbow or dog leg deformity of the anterior mitral valve leaflet</td>
</tr>
<tr>
<td>Prolapse of mitral, aortic or tricuspid valves</td>
<td>Fixed or markedly restricted motion of the posterior mitral leaflet</td>
</tr>
<tr>
<td>Reduced mobility of leaflets</td>
<td>Chordal tears</td>
</tr>
</tbody>
</table>

**CONCLUSION**

It is clear from the new evidence on the superior performance and high yield of echocardiographic screening of rheumatic heart disease in high-risk communities that we cannot continue with “business as usual” in the management of rheumatic heart disease in South Africa.\(^{(24)}\) First, healthcare practitioners in South Africa need to be reminded that rheumatic fever is a notifiable condition and be encouraged to develop registry-based follow-up clinics for their patients with rheumatic heart disease.
health care workers to perform a focused echocardiogram with possibility of using technologists, nurses or even local community to clinical cardiology centres for reporting by trained staff. The auscultatory and echocardiographic findings of screening can be transmitted to clinical cardiology centres for reporting by trained staff. The possibility of using technologists, nurses or even local community health care workers to perform a focused echocardiogram with specialised staff reviewing these echocardiograms should be explored. With the shortages in trained staff in all strata of healthcare, we need to develop innovative and creative strategies for the implementation of effective screening programmes in resource-poor settings. The A.S.A.P. programme provides an evidence-based framework for building a national programme of prevention of rheumatic heart disease in South Africa and other African countries. Finally, prospective studies are required to evaluate the natural history and management of subclinical rheumatic heart disease, and to explore cost-effective strategies for the screening of rheumatic heart disease in the general population of South Africa.

REFERENCES