Echocardiographic screening for rheumatic heart disease

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For a disease to be suitable for screening as a tool of preventive medicine, it must satisfy the following criteria: (1) the evidence of an obvious burden of the disease, (2) an initial latent stage of the condition, (3) which can be detected by appropriate tests, (4) and treated by adequate therapy, and finally (5) to prove that intervention at an early stage can improve prognosis.1

Rheumatic fever is a delayed sequel to throat infection by a group A streptococcus. More than one third of affected children develop carditis, followed many years later – especially in the case of recurrent rheumatic infections – by progressive and permanent valvular lesions, known as rheumatic heart disease.2 Rheumatic heart disease is now largely restricted to developing countries and most of the reduction in its incidence in wealthy countries is attributable to better living conditions with consequent reductions in the transmission of group A streptococci. Nevertheless, with the decline in rheumatic fever in industrialized countries and the emphasis on diseases such as malaria and HIV in developing countries, there has been a parallel reduction in research into this still prevalent and important condition.3 Rheumatic heart disease remains an obvious public health burden across the developing world causing more than 200 000 deaths a year.4

An exaggerated immune response to specific bacterial epitopes in a susceptible host is thought to be the basis of the disease.5 Rheumatic heart disease usually results from the cumulative damage from recurrent episodes of acute rheumatic fever. It has been reported that after a first episode of carditis, cardiac auscultation becomes unremarkable in one-third of children but even these children may progress to significant rheumatic valve disease in later life, as confirmed by echocardiography.6

Thus early detection of “subclinical” rheumatic valve disease is vital, as it presents an opportunity for case detection at a time when prophylactic penicillin – to prevent recurrent episodes – can prevent progression to important valve disease in young adult life. Early detection of rheumatic heart disease in school children is traditionally done by listening for murmurs by stethoscope, followed by echocardiographic confirmation only in suspected cases. Recently, we did systematic echocardiographic screening in schools in Cambodia and Mozambique, to assess whether early case detection could be improved by using portable ultrasound.7 We achieved a case detection rate by echocardiography approximately 10-fold that achieved by clinical examination only. Such screening thus makes it possible to identify children at risk of developing severe rheumatic valve disease for whom secondary prevention with penicillin prophylaxis may be effective.

Acute rheumatic fever episodes can be prevented by antibiotic treatment of group A streptococcal throat infections, a strategy known as primary prophylaxis. Nevertheless, highly sensitive and specific clinical diagnostic algorithms for group A streptococcus pharyngitis are not available, microbiological diagnosis is expensive, and at least two-thirds of the patients with acute rheumatic fever do not get a sore throat. The key to primary prevention is reducing exposure to group A streptococci, which requires dramatic improvements in housing, hygiene infrastructure and access to health care for inhabitants of the developing world.8 Even with comprehensive programmes for primary prophylaxis of rheumatic fever, not all acute episodes can be prevented. Moreover, systematic screening and treatment of sore throats – for example, in school-based programmes – has not been proven to be cost-effective.9

Unfortunately, at the present time there are no other proven strategies to prevent initial episodes of acute rheumatic fever.9,10 Secondary prophylaxis consists of monthly penicillin injections in children after a first episode of acute rheumatic fever and follow-up until the third decade of life. This strategy has been shown to be inexpensive and efficient, and is best delivered as part of a register-based control programme. The World Health Organization and the World Heart Federation have recommended this approach since the 1980s.

It is not yet proven that penicillin prophylaxis for all children with subclinical rheumatic valve lesions is the best option in clinical practice. Antibiotic prophylaxis is currently recommended in endemic areas, only in those children with “significant subclinical rheumatic heart disease”.10 Therefore, we cannot currently recommend that all children with echocardiographically detected subclinical rheumatic heart disease lesions start prophylaxis, but only those with “significant” lesions (defined as “very mild regurgitant jet, more than 1.0 cm, localized immediately above or below the valve, throughout systole at the mitral valve or diastole at the aortic valve”).10 Research into the risks and benefits of including all children with subclinical rheumatic heart lesions in follow-up prevention programmes will be essential to assessing the best strategy for rheumatic heart disease prevention.

While echocardiography has been shown to be a useful tool in detecting children with subclinical rheumatic heart disease, studies are needed to examine the cost-effectiveness of such screening. The cost of a basic portable echocardiographic device is approximately US$ 15 000, and educating assistants to use echocardiography in diagnosing clinical and subclinical rheumatic heart disease should be feasible. We hope that our data stimulate a re-evaluation of the feasibility, efficacy and cost-effectiveness of ultrasound-based screening for subclinical rheumatic valve involvement in high-prevalence settings.

References

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