Rheumatic heart disease (RHD) remains one of the largest preventable burdens of disease in the world. It is perceived as a disease of childhood, acquired by streptococcal throat infection of the tonsillo-pharynx, leading to an inflammatory reaction that involves many organs, including the heart. However, cases in children of 5 to 14 years of age are likely to represent only 15% to 20% of all cases within all age groups of vulnerable populations.1

As a disease of poverty and low socioeconomic circumstances, RHD is prevalent in the developing world. This association with the poorest and weakest of society represents a double tragedy: whereas the lack of access to education, infrastructure, and medical facilities turns a preventable and surgically treatable condition into a major cause of suffering, incapacity, and death, global awareness remains depressingly low.

Although RHD has received increasing attention in the past few years, leading to declarations,2 definition of areas of research,3 and new diagnostic criteria on echocardiography, as well as suggestions for prevention programs, insight into incidences, disease progression, and mortality remains largely speculative because of a lack of data.

With the adoption of echocardiography to project-funded screening programs, it has emerged that previous estimates may have distinctly underestimated the prevalence of the disease.4 Because screening programs generally focus on school children, they concentrate on the most accessible and reliable cohort for the assessment of disease incidence. Yet, echo-based incidences of RHD in school children are age-dependent,5 because a 71% increase in children younger than 9 years of age, to those older than 12 years of age,6 was shown. Although the 10× higher sensitivity of echo screening over clinical assessment4 detects in 90% of cases clinically silent RHD, almost half of these valve lesions are already functionally moderate to severe.6 Therefore, together with the effectiveness of secondary penicillin prophylaxis, the case for echo screening programs is strong.

However, as important as school-screening programs are for secondary prophylaxis, they address only the tip of the iceberg, in view of the disease progression in the undiagnosed post–school-age population groups. In contrast to the assessment of early disease incidence in school children, data on disease progression rely on the screening of adult population cohorts, years after manifestation of the disease. Historical knowledge largely comes from the developed world, before disease eradication, when the natural history of RHD was already mitigated by high socioeconomic standards. In developing countries, pregnancy screenings provide some insight6 but require a more advanced level of socioeconomic development of a society than the screening of school children. One of the few follow-up studies to date reports a 16% progression rate from acute rheumatic fever to severe, clinically relevant chronic valve lesions within less than 15 years in Brazil.7 Given an estimated 60% progression rate from rheumatic fever to RHD,8 the actual progression rate of echocardiographically diagnosed cases would therefore be closer to 25%. The proportion of these patients who eventually die remains the biggest unknown in the natural history of RHD. Sporadic reports from countries like Ethiopia suggest that 70% of patients die before they are 26 years of age.9 Previous global estimates of 500 000 deaths per year8 have been recently increased to 1.4 million per year.10 Apart from prophylactic prevention, the majority of these deaths might be preventable through surgical valve repairs or replacements.

Thus, as the political will for funding and implementation of both prophylaxis programs and the rollout of cardiac surgery will depend on better insight into the impact of RHD on population health, productivity, and premature mortality, the next big challenge in the combat of RHD will be better insight into the natural history of the disease.11 Yet, early detection by using the newest screening guidelines (2006 National Institutes of Health/World Health Organization) does represent a crucial first step.

The study by Beaton et al,12 performed in Kampala, Uganda and published in this issue of Circulation, addresses the limited data from Sub-Saharan Africa on echocardiography in children. Auscultation and portable echocardiography was used to screen 4869 randomly selected schoolchildren, 5 to 16 years of age. The strengths of the study are the large sample size, the carefully planned prospective design, and the systematic application of criteria for diagnosis. The study design is interesting and novel in that it provided simple prescreening and, if any abnormality was detected, very detailed assessment by a cardiologist at Mulago Hospital, Kampala. Through this novel approach the researchers were
able to screen 200 to 250 children with 1 sonographer, in 1 day, while 2 other staff members served as organizers and data tabulators. Each school-based screening took approximately 2 minutes to complete. Echocardiography detected 3 times as many cases of RHD than auscultation. Lower socioeconomic groups had significantly more RHD, with also more advanced disease presentation. The burden of disease in that cohort was 15 cases per 1000. The authors interestingly expanded their calculation to the Ugandan population of an estimated 18 million children in the age group 5 to 16 years. By this calculation, 266 400 cases would be detected, whereas only 88 200 would be detected by clinical examination. Their data suggest that screening efforts targeting 10-year-old children in lower socioeconomic cohorts may maximize subclinical detection.

Their data are different from another study using echocardiography-based screening in sub-Saharan Africa. The study performed by Marijon et al² in 2170 otherwise healthy school children in Mozambique, between 2001 to 2002, found a substantially higher burden of disease (30.4 cases per 1000), corresponding with 33/1000 in Tonga,¹³ 24/1000 in New Zealand’s Maoris,¹¹ and 51/1000 in India.¹² Apart from including children as young as 5 years of age and, as such, encountering an expectably lower incidence, the difference might also be explained by difference in region, socioeconomic circumstances, or echocardiographic criteria used.

The authors critically discuss various levels of screening by echocardiography via careful clinical assessment and comment on the use of various guidelines, including the new criteria from the World Heart Federation, published February 2012.¹⁴ The study supports the inclusion of portable echocardiography in screening protocols, even in resource- and medical staff–constrained settings.

A recent study from the Heart of Soweto cohort, reporting on the incidence and clinical characteristics of newly diagnosed RHD in adulthood from an urban African community, found an estimated incidence of new cases of RHD for those >14 years of age to be in the region of 23.5 cases/100 000 per annum.⁶ Many of those patients presented late, with an echocardiographic ejection fraction of <45% in 17% of the cases and an elevated right-sided pressure in 18% of the cases. Therefore 22% of this cohort of 344 cases had valve replacement or repair within 1 year, with a further 26% being admitted for initial diagnosis of suspected bacterial endocarditis within 30 months.

A further burden of late diagnosis of RHD is the fact that women often only present with symptomatic RHD when pregnant. A 4-year audit of cardiac disease in pregnancy in a South African hospital found an etiology of 63.5% of RHD and 20.1% of prosthetic VHD, probably of RHD origin, which contributes to the unacceptably high maternal mortality in South Africa.¹⁵

Although a predominant lesion in RHD patients presenting in pregnancy is mitral stenosis, which is amenable to balloon- or surgically-closed mitral valvotomy, 60% to 84% of cases presenting outside of pregnancy involve mitral regurgitation and 15% to 40% aortic regurgitation, eventually making open heart surgery inevitable.

Even extrapolating the authors’ relatively low incidence of 15/1000 to an African population of 1 billion would result in 15 million RHD patients with almost 400 000 new cases per year, of which 100 000 per year may need heart valve surgery at some stage in their lives. Taking the higher incidences reported from Mozambique, this number would be as high as 200 000 per year.⁴ At present, only South Africa, Egypt, Sudan, Kenya, Ghana, and to a limited extent, Algeria, and,
most recently, Namibia have established independent cardiac surgical programs that do not rely on sporadic fly-in missions. Yet, even there, a majority of hospitals cater for the few private patients, whereas the largely indigent RHD patients rely on a small number of public hospitals. Therefore, the already dismal access to 18 open heart surgeries/million population, as opposed to 1222/million in the United States, is in reality distinctly lower. In South Africa—which boasts two thirds of all hospitals that provide cardiac surgery on the African continent—only 15% of these cater to the indigent patients with RHD. Thus, >50 million patients compete for limited access to 7 public centers in the one African country that has the best established cardiac surgical facilities. The situation is even more dramatic in countries like Uganda, where the incidence highlighted by the authors contrasts with the almost complete inability of routine referral to cardiac surgery.

This study, as with all studies, also has weaknesses. Only left-sided valves were examined for features of RHD. This is by far the most common area for RHD, but a number of cases involving the right side may have been missed. Spending only approximately 2 minutes per child and doing screening on more than 200 cases a day, by one sonographer, might have led to missing a number of cases. The number of views recorded for secondary evaluation must have been limited by this approach.

The authors did not include a cost calculation. This would have been of particular interest, as this study was carried out in a developing world set-up with limited funding. In the cost calculation the salaries of the three staff members, as well as the equipment needed for cardiac ultrasound, would need to be included.

Furthermore, the use of the much cheaper hand-held portable echocardiography to detect RHD in children and adults with symptoms should be explored, as the image quality might be sufficient for screening purposes (Figure). Interestingly, the World Health Organization supported the development of a simple, affordable, solar-powered blood-pressure device. Support for other cheaper devices, as for cardiac ultrasound, would be laudable.

In conclusion, the authors have provided a significant contribution to the knowledge of childhood incidence of RHD in a hitherto unreported part of Sub-Saharan Africa. Well-conducted studies like this will continue to be important for creating awareness of the fact that we only begin to see the tip of an iceberg, which developed countries have long erased from their radar screen. Without deeper understanding of the disease progression and the actual impact of RHD on the respective countries, however, the necessary political pressure for the rollout of secondary prophylaxis and surgical therapy will be insufficient.

Disclosures

None.

References


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