Rheumatic Heart Disease in India: ‘Buried alive’

Rheumatic fever (RF) and rheumatic heart disease (RHD) are still widely prevalent in most parts of India. RHD may have declined in India from the pandemic levels seen earlier but it remains a major public health concern. Unfortunately, despite its continuing presence, our attention on RHD has declined. There is no national or state programme to control it and there is little if any research to understand the pathogenesis, the efficacy of primary prophylaxis or the role and efficacy of secondary prevention in high-risk groups. Increasing privatization of healthcare has further marginalized patients with RHD as they are usually poor and unable to afford healthcare at corporate hospitals. Thus, a large number of practising physicians, cardiologists and healthcare administrators may never have encountered a patient with RHD and hence believe that the disease is no longer a public health problem in India. The study by Kumar et al., published in this issue of the Journal, is important as it establishes that RF/RHD have not declined even in Punjab, one of the most economically developed states of India. They found a prevalence of 1/1000 (95% CI 0.8–1.3) in schoolchildren in the age group of 5–14 years. Based on the school survey, the prevalence of RF/RHD in the general population was estimated to be 143/100 000. More importantly, episodes of RF were still seen and the annual incidence of acute RF was estimated to be at least 8.7/100 000 children in the age group of 5–14 years.

The majority of studies on prevalence of RHD in India are school-based surveys with only a handful of studies focusing on the entire population (Table I). The drawback of these studies is that they estimate a point-prevalence with no follow-up. The present study is prospective and well-designed; it included active surveillance data from health centres/practitioners and did two sample surveys in schools to ascertain the epidemiology of RHD in a defined population. Even though a serious attempt was made to include the absentees, around 2% of children could not be included. Despite its prospective nature and careful design, it may still not replace a population-based survey, but such surveys are difficult to organize and conduct. However, the study has important advantages over simple school surveys. The overall estimates for population prevalence are likely to be more accurate and the burden among the non-school-going age group could be estimated. Unlike school surveys, the study was able to estimate the mortality and morbidity due to chronic RHD using active surveillance and verbal autopsy methods.

School surveys on RHD have gone through three major phases—auscultation, auscultation followed by echocardiography, and more recently echocardiographic screening in all. The reported prevalence of RHD varied from 1.8 to 11/1000 schoolchildren (average 6/1000) during the 1970s and 1980s, and 1–3.9/1000 during the 1990s. However, these studies used clinical auscultation to diagnose RHD. Studies using echocardiographic validation of clinical diagnoses show a much lower prevalence of RHD. The prevalence of RHD estimated by the present school surveys is comparable to the older studies using the method of auscultation followed by echocardiographic confirmation. The prevalence estimates range from 0.5/1000 in Gorakhpur (2007) to 0.67/1000 in Vellore (2003) and Bikaner (2006). Paradoxically, the reported prevalence of RHD increases dramatically if echocardiographic screening is done for all schoolchildren. The estimated prevalence of echocardiographically detected RHD in India is as high as 20.5/1000 (Ballabgarh, 2011). This is contrary to what was happening earlier—clinical evaluation including auscultation was done and all those with heart murmurs or clinical suspicion of a heart disease had an echocardiogram. Echocardiography decreased the true prevalence as it showed that a lot of the murmurs were benign or due to congenital heart disease. When using echocardiography alone, the numbers increased as a new disease
profile was found, that of clinically silent, echocardiography-identified RHD. The presence of mitral stenosis or elevated anti-streptococcal antibodies could help in confirming that these are indeed RHD. However, in the absence of the above, no other investigation can prove or disprove the valve being ‘rheumatic’. The natural history of people whose valves get labelled as ‘rheumatic’ is not known and there is no consensus about whether they should receive secondary penicillin prophylaxis.

The estimated prevalence of RF/RHD is substantially higher than what was reported by a population-based study from Punjab in 1993 (0.09% or 90/100 000). This is despite the fact that the surveillance methods used in the present study are likely to under-report prevalence. The annual incidence of acute RF is also substantially higher than the estimates of RF in various countries. The prevalence did not differ between the two school surveys but that may not mean much as these were only 3 years apart.

The above data along with the study published in this issue of the Journal suggest that the prevalence of RHD has not declined, even in one of the more economically advantaged areas of India. This statement, however, needs to be ‘tempered’ as India is a large country. A prospective two-point survey, 15 years apart in schoolchildren aged 5–15 years living in the hills of Shimla, did show a 3-fold decline in the prevalence of RF/RHD in both rural and urban areas. More data are required and quickly from different parts of India as the disease remains common in the poorer and more populous states such as Bihar, Odisha, Chhattisgarh, Uttar Pradesh, etc. while cardiologists in southern India contend that RHD is uncommon. Most importantly, while the point-prevalence of RF/RHD may have decreased, the total burden in India may be same or more as the total population at risk has increased.

The risk factors for RHD are consistent across recent studies. The prevalence of RHD generally peaks in 10–14 years and is more common in females. It is not known whether the female preponderance of RHD is due to increased incidence of streptococcal sore throat, increased susceptibility for subsequent RHD or due to neglect of healthcare in Indian girls compared to boys.

The prevalence in government schools and private schools was 1.2 and 0.3/1000, respectively and such a 4–5-fold difference has been reported by other large school surveys in India. Most children belonging to the lower socioeconomic class study in government schools that are poorly maintained and overcrowded. The middle class and

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ICMR Indian Council of Medical Research
the affluent have a much lower incidence of RF in the same cities wherein people in slums have a much higher number. Given the large difference in the prevalence of RHD, control programmes in India must focus on government schools.

One major drawback of the study was that the echocardiographic interpretation was not based on standard echocardiographic criteria. Echocardiography is useful to diagnose severity of valve lesions in RHD and to rule out other causes of murmurs. The routine use of echocardiography to diagnose RF is still not recommended and is not included in the Jones’ criteria. The use of echocardiography to confirm clinically suspected RHD/RF is desirable, but milder forms of valve involvement, especially mitral regurgitation (MR) alone, pose a major challenge in interpretation even to experts in RHD. The WHO has proposed criteria to differentiate physiological and pathological valvular regurgitations. Recently, the World Heart Federation (WHF) has proposed criteria to define rheumatic involvement of a valve using echocardiography and Doppler. Given the lack of a gold standard and the variable disease progression in different populations, validation of these criteria may take several years.

The present study also took throat swabs to assess the presence of group A streptococcal (GAS) infection/carriers among these children. The prevalence of GAS was 2% (13/656) in those with a sore throat and 0.5% (14/2920) among those who did not have a sore throat. Only 27 GAS typing was done that revealed 16 emm types. The prevalence of GAS among children with sore throat is highly variable with high estimates in urban slums and rural areas of India. The investigators calculated that about 1000 episodes of GAS pharyngitis would lead to one episode of acute RF in the study community. These estimates are slightly lower than those of the classical studies of 1960s from the West which estimated that 100 episodes of GAS throat infections led to 0.3 to 3 cases of RF in endemic and epidemic situations, respectively. Such a low rate of conversion into RF following streptococcal pharyngitis will raise serious questions on the efficacy of a primary prophylaxis programme for the control of RHD. In developed countries, rates of streptococcal sore throats have not decreased, yet RF and RHD have disappeared. It is believed that widespread use of antibiotics for sore throat is one of the reasons for the decline of RF/RHD in southern India and in Ladakh where it was rampant till a few years ago. A reduction in episodes of acute RF following a streptococcal sore throat may be an indicator of the epidemiological decline of RHD.

The present study reiterates that implementation of prevention strategies for primary and secondary prophylaxis within the existing primary healthcare setting may be cost-effective to control RHD in developing countries. Most (95%) of the RF/RHD cases received secondary prophylaxis regularly as against 50%–60% reported earlier. Despite good adherence to penicillin, the consequences of RHD remain serious. The study reports 42 deaths among 813 registered (5.2%) and the estimated annual death rate due to RF/RHD was 3/100,000 population. More importantly, young people with RHD die; the median age at death was 35 years.

The defined population for the study is representative of an average Indian district, with a majority in rural areas and a large proportion of the population in the young age group, though the average per capita income may be higher than the national average. The prevalence of RHD could be higher in states with poorer socioeconomic indicators. The study also shows that economic development alone may not translate into adequate and desirable improvements in parameters of healthcare. As suggested earlier, human development indices may be better correlates of the prevalence of RHD in the community than economic indices. The burden of RHD is unlikely to decrease as the population at risk of RHD is steadily increasing. The burden of RF/RHD could be similar to that of cancer in India, yet there is little focus on RHD. Therapy of valve involvement requires surgery, which is scantily available for the needy population in India. Valve replacement by itself is not a solution as secondary prophylaxis has to continue and anticoagulation has to be strictly adhered to. Regular monitoring of prothrombin time/international normalized ratio (INR) is expensive and needs high-quality laboratories, something not possible in rural India even today. Thus, making these patients believe that valve replacement is going to cure them is a half-truth at best. It seems the only hope for this population is early identification and diligent secondary prophylaxis with penicillin before valve damage becomes severe. Severe regurgitation may not decrease in some cases as MR begets MR even if there is no further organic insult.

We propose an action plan for the control of RHD (Table II). The efforts of primary and secondary prevention of RHD should be integrated with school health services. The school health syllabus should include better knowledge of RHD and other common
diseases. RHD does not get a mention in the list of heart diseases and even well-educated children and adults are not aware of the disease. Linking sore throat with joint pains and RF should be included in school curriculum. Newspapers and the electronic media should be used to propagate that a simple sore throat could end up with a serious heart valve disease. It is also important to stress on the need for regular parenteral benzathine penicillin prophylaxis and remove the fear of anaphylaxis from the minds of caregivers as its incidence in children receiving penicillin is <1/100 000. Patients, caretakers and caregivers need to understand that valve replacement is not a cure for this disease.

Since non-communicable diseases (NCDs) are overtaking communicable ones, even in India, there has been much awareness about heart diseases. Unfortunately, the focus is on coronary artery disease and its complications; till recently, RHD did not even find a mention in the list of NCDs.

We believe most of India continues to have a high burden of RF and RHD. The disease generally affects children and young adults who are in the prime of their life and leads to a high morbidity and mortality. There is an urgent need to tackle the disease at the national level and its prevention should be incorporated in the national agenda. Concerted action is needed to eradicate this potentially preventable disease.

**Table II. Suggested policy initiatives to control rheumatic heart disease (RHD)**

**Broad initiatives**

A. Integrating early detection, primary and secondary prevention of RHD in school health services

1. All school children should have an annual health check-up wherein cardiac auscultation should be mandatory. Explore the possibility of using electronic stethoscopes with recording facilities to get offline expert opinion.

2. School teachers, school medical and paramedical staff should be educated about the importance of joint pains and sore throat so that they are able to detect children early in the course of the disease.

3. Medical, paramedical staff and social workers should be trained to detect streptococcal sore throats in children using scoring systems.

4. Oral antibiotics such as ampicillin, amoxycillin and cephalosporins should be made available free of cost to treat children in government schools.

5. Injectable benzathine penicillin should be made available free at all primary health centres, district hospitals and private nursing homes/hospitals. Resuscitation equipment and drugs for treating anaphylaxis should be kept at all such centres and basic life support (BLS) training should be mandatory for all health workers.

B. Maintenance of nationwide RF/RHD registers to improve secondary prophylaxis and also monitor data

C. Increase awareness of the disease

1. School textbooks do not mention RF/RHD and hence there is little awareness of this disease. Incorporate information on heart in school textbooks. Linking sore throat with RF should be included in school curriculum.

2. The print and electronic media should be used to increase awareness that an untreated sore throat could end up as a serious heart valve disease.

**Targeted actions**

1. Declare RF/RHD as a national health programme, make it reportable and maintain a comprehensive database of patients suffering from the disease. Make injectable benzathine penicillin available in primary health centres, district hospitals, health-related non-governmental organizations and the private sector free of cost.

2. Ensure that injectable penicillin is given in a room well-equipped with resuscitation facilities and in the presence of a doctor or nurse capable of managing anaphylaxis. Public awareness should be increased about the safety of penicillin and the low incidence of anaphylaxis in children. Doctors refusing to administer penicillin need to be warned by the state medical council and action taken on repeated defaulters.

3. Maintenance of nationwide RF/RHD registers/digital data to improve secondary prophylaxis by sending reminders, etc. Even the poorest of people in the remotest corners of India have access to mobile phones which should be used for communication about compliance, etc.

4. Give RHD its importance in the list of NCDs.

5. Make heart valve replacements and mitral valve balloon dilatation procedures free to all poor patients in tertiary care centres.

6. Anticoagulation monitoring for patients of RHD with atrial fibrillation and those who have had mechanical prosthetic valves implanted should be standardized, funded and made available free for poor patients.

7. Dedicated centres of excellence for RF/RHD that focus on all aspects of RF/RHD—from basic research to the most sophisticated training in 3D imaging and imparting cardiothoracic surgeons’ skills to repair mitral valves and do a maze surgery for atrial fibrillation. These centres should be headed by cardiologists who are willing to dedicate themselves to this field.
REFERENCES


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