Rheumatic Heart Disease: How Big is the Problem?

A Ibrahim, MRCP* A R A Rahman, PhD** * Department of Medicine ** Department of Pharmacology and Medicine School of Medical Sciences, Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan

Malaysia is progressing very rapidly in all aspects of life. It is fast developing and very soon it will be a fully developed country. Many dangerous diseases, especially those of infectious aetiology have been eradicated and controlled. However, there are other diseases which are quite prevalent and result in high morbidity and mortality. Cardiovascular diseases are good examples of these, and they are escalating in their prevalence. They are the leading cause of death in many countries of the world and in Malaysia they rank as the number one cause of death since the beginning of the last decade¹. Among the cardiovascular diseases, coronary artery disease is the top killer. Systemic arterial hypertension comes next and valvular heart disease is the third killer of which rheumatic fever is the main cause. Of all new patients referred to the adult cardiac clinic in Hospital Universiti Sains Malaysia between 1991-1992, the commonest diagnosis made was rheumatic valvular heart disease².

Rheumatic fever and rheumatic heart disease continue to be major public health problems, especially in the developing countries of the world³. Although the incidence of acute rheumatic fever and its sequelae, chronic rheumatic heart disease, has declined in Western Europe and North America, the disease remains common in Eastern Europe and is increasing in Asia. Worldwide, it remains the single most important cause of valvular heart disease. In developed countries a steep decline has been noted in recent years⁴. Incidence of rheumatic fever in Denmark decreased from 200 cases per 100,000 between 1862 to 1888 to fewer than 5 cases per 100,000 in 1962⁵. By contrast, in developing countries like Asia, South America and Africa, rheumatic fever and rheumatic heart disease are still prevalent^{6,7}. In Brazil, Chile, Uruguay and Venezuela, rheumatic fever constituted 6-7% of hospital admissions up to 12 years of age^{8,9}. In Thailand, the reported prevalence rate of rheumatic fever and rheumatic heart disease among school children was 1.2-2.1 per 1000¹⁰. With the backing of the World Health Organization and the Thai government, a national control program for rheumatic fever and rheumatic heart disease was established in 1986¹¹.

Although there was a decline in the incidence of acute rheumatic fever in many developed European countries and in the United States of America, there have been reports lately of resurgence of this disease¹². In Norway, acute rheumatic fever has been a curiosity during the last 30 years and only five cases were reported to the Norway's National Notification System For Infectious Diseases in the 1990s. However, during the period 1990 to 1992, 99 patients were discharged from Norwegian hospitals with the diagnosis of acute rheumatic fever¹³.

In some developing countries, not only is the incidence of acute rheumatic fever not decreasing, it is if anything increasing. In Turkey, the 1990s has seen an increase in the frequency of the disease, compared with 1980s. Between 1990 to 1992 alone, there were 228 patients with acute rheumatic fever being admitted to a paediatric hospital in Ankara¹⁴. In Malaysia, rheumatic fever and its sequelae are still common but their exact incidences are not known as these diseases are not reportable, and no study had so far been

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carried out to assess them. However, a prevalence study of rheumatic heart disease in primary school children in Kelantan revealed a prevalence rate of 0.11 per thousand¹⁵. Nearly forty-seven thousand children (46,986) from 144 primary schools in three districts of Kelantan were screened between August 1988 to December 1990. Five children (0.2% of those detected to have a heart murmur) had echocardiographic evidence of rheumatic valvular heart disease. This prevalence rate is low, compared to other prevalence studies in Asia. The number of school children screened in other studies ranged from 6,004 in the Indian study¹⁶ to 67,125 in the Thai study¹⁰. Our study subjects were younger, and this may have inadvertently missed out the age group in which other studies showed high prevalence of rheumatic heart disease^{10,16,17}. In the University Hospital Kuala Lumpur, as reported in this issue of the Journal, 134 children with acute rheumatic fever were admitted between January 1981 to December 1990. This gave a hospital incidence of 21.2/100,000 paediatric admission per year. The reported incidence of first attack was 15.8/ 100,000 per year and recurrent attack of 5.38/100,000 per year¹⁸. In Kelantan, a total of 158 hospital cases of acute rheumatic fever were documented between January 1987 to June 1994. The age ranged from 1 to 35 years, the commonest affected group being from 6 to 15 years old, and a male to female ratio of $0.9:1^{19}$.

Although acute rheumatic fever at present may not be as common in Malaysia as compared to other developing countries, there are still many patients who are suffering from its sequelae, namely mitral valve disease, predominantly mitral stenosis. These people have increased mortality and morbidity especially those with severe valvular defects complicated by cardiac failure and atrial fibrillation. Infective endocarditis is a real problem if it occurs in these patients. Thrombotic stroke is a major tragedy and is fairly common in patients with rheumatic mitral valve disease complicated by atrial fibrillation. In Malaysia, there are also many patients on the waiting list for mitral valve surgery or percutaneous transvenous mitral commisurotomy for rheumatic mitral valve disease.

In the meantime, what can we do to reduce the incidence of this disease? Eradicating group A beta

haemolytic streptococcus pharyngitis would be ideal, but it would not be easy, unless a vaccine can be developed. Further research is needed until a thorough knowledge of the various clinical manifestations of group A streptococcus infections is available in order to apply the preventive and therapeutic measures in a rational manner. This is more pressing, in view of the changing epidemiology of the prevalent group A streptococcus strains. An increase in the M - protein serotypes associated with higher virulence is being recognised in Europe and The United States²⁰. Fortunately, group A streptococcus remained susceptible to penicillin. The next step then would be to make penicillin easily available to children who are suspected to have streptococcal pharyngitis and treat them adequately. Indeed evidence from Nepal showed that the high rates of acute rheumatic fever and rheumatic heart disease may relate not to increased prevalence of streptococcal infection, but to inadequate antibiotic therapy (proper dosage and duration) of streptococcal pharyngitis²¹. Evidence from the Philippines even went to the extent of suggesting that by giving penicillin to school children with pharyngitis (prior to confirmation of its aetiology), the attack rate of rheumatic fever can be reduced by ten fold²².

Secondary prophylaxis is another area where we can all improve on. It is important to make penicillin easily available to this group of patients. The Philippines experience showed that compliance to penicillin decreased as the drug became less accessible (when supply of drug ran out) and this in turn can cause an increase in the prevalence rate²². Dosage and duration of penicillin therapy is another issue we need to reappraise. The classical practise of giving intramuscular benzathine penicillin 1.2 mega unit every four weeks has come under close scrutiny²³. Recent evidence showed that this may not be adequate in rheumatic fever prophylaxis. Injections every three weeks²⁴ or even fortnightly²⁵ is more appropriate in controlling rheumatic fever recurrences. Discontinuation of rheumatic fever prophylaxis is another area where there is yet no consensus. Recent prospective evidence from Chile suggested that prophylaxis can be stopped relatively safely in patients who a) have reached their early twenties b) have had their most recent attack more than five years ago c) did not have carditis with their previous attack(s)

and d) are free of rheumatic disease²⁶. Caution must however be exercised in discontinuing prophylaxis and decisions made must be individualised, after assessing the risks and benefits involved²⁷. Finally patients with established cardiac complications must be regularly followed up and decisions on therapeutic or surgical intervention made at the most appropriate juncture. This requires cooperation and proper understanding of prognosis by patients and relatives and proper counselling on the doctors' part.

Rheumatic heart disease remains the most common acquired heart disease worldwide. In tropical countries, it is the commonest form of heart disease with high mortality, prevalence, incidence and disability rates⁶. Control of the disease to a very low prevalence and even to the stage of eradication is not impossible. The World Health Organization (WHO) and the International Society and Federation of Cardiology (ISFC) have jointly embarked on a global program for the prevention and control of rheumatic fever and rheumatic heart disease¹⁶. Although Malaysia is not directly involved in this program (Indonesia and Vietnam are), it is our hope that we can tap local expertise towards a more concerted effort in controlling this disease to the minimum, if not complete eradication. This will require collaboration and cooperation between the different specialities; microbiologists, epidemiologists, public health educators, clinicians and not forgetting the public. This will hopefully lead in future to the decline if not demise of an important public health hazard.

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