Prevention of Rheumatic Heart Disease in Saudi Arabia: Fact or Fiction?

Y. A. Al-Eissa


Thirty-four children, 6–14 years of age, with recurrent acute rheumatic fever or already established rheumatic heart disease were studied to highlight the reasons and consequences of rheumatic fever recurrences. The majority of cases belonged to low and middle-class large families with ready access to medical care. Although 62% of the patients had a history of pharyngitis prior to the onset of rheumatic fever and the majority of them sought medical attention, they received inadequate treatment. All patients developed rheumatic recurrences because they were not on prophylaxis at all or they took their medications irregularly. The main reasons for non-compliance were lack of proper counselling, children’s resentment of injections, or inaccessibility to penicillin injections. Cardiac involvement was detected in 32 patients of whom 56% had isolated mitral regurgitation, 6% had isolated aortic regurgitation, 34% had multivalvular regurgitation, 3% had mitral regurgitation and stenosis, and 28% had congestive heart failure. Current strategies for primary and secondary prevention must be intensified if rheumatic fever and rheumatic heart disease are to become diseases of the past.

Acute rheumatic fever (ARF) is an inflammatory sequela which occurs in 0.3–3% of children afflicted with streptococcal pharyngitis.1 One of the striking features of ARF is its tendency to recur; a subsequent untreated streptococcal pharyngitis is associated with a 25–75% risk of a recurrence.2 Although an initial attack of ARF can heal without damaging the heart, the risk of chronic rheumatic heart disease (RHD) is higher with recurrences.3 Acute rheumatic fever occurs predominantly in developing countries where it remains the leading cause of acquired heart disease with its high rate of morbidity and mortality.3 This is partly attributable to low socio-economic conditions, overcrowding, and poor access to medical care. This paper intends to highlight the reasons and consequences of rheumatic recurrences in a group of
34 children, and to suggest strategies for more effective control of ARF and RHD in Saudi Arabia.

Patients and Methods
From January 1985 to December 1989, 79 consecutive children aged 14 years and younger with ARF or RHD were seen at KKUH; 34 of these who presented with recurrences of ARF or already established RHD form the body of subjects of this study. The remaining 45 patients with an initial attack of ARF were excluded. All patients came directly to the paediatric primary-care clinics which are operated on a walk-in basis. Patients were carefully assessed at presentation to confirm a present or past diagnosis of ARF and to differentiate initial attacks from recurrences and established RHD. The diagnosis of ARF was based on the revised Jones criteria. Rheumatic recurrences were defined as the reappearance, after an interval of at least 2 months of freedom from rheumatic activity following the cessation of suppressive therapy, of manifestations that fulfilled the revised Jones criteria. Rheumatic heart disease was defined as inactive valvular heart disease of presumed rheumatic origin (i.e. history of previous attack of ARF) in the absence of clinical and laboratory signs of active inflammation, and where congenital causes or cardiomyopathies were excluded.

For each patient the following information was sought: age at diagnosis of initial attack; subsequent recurrences and their timing; sex; residence; family size and housing condition; parents' education and occupation; previous history of pharyngitis and possible antibiotic treatment; the cardinal presenting features at initial diagnosis and subsequent recurrences; parents' knowledge of the disease; adherence to the prophylactic regimen; and family history of ARF. Pertinent laboratory investigations and a two-dimensional echocardiography with Doppler examination were carried out for each case to verify the diagnosis of ARF and valvular lesions respectively.

All patients were conventionally managed and intensive counselling was made to ensure continuous secondary prophylaxis and follow-up at specific intervals.

Results
Twenty-two children presented with recurrent ARF, while 12 had a definite previous history of ARF and signs of established RHD. The demographic features of all 34 children are summarized in Table 1. Of the 34 patients, 85% shared a bedroom with one or more family members. All the patients, except one young child, were attending school during their initial attack of ARF. Seven patients (21%) had an immediate family member with a history of ARF.

Twenty-one (62%) of the 34 patients recalled symptoms suggestive of pharyngitis within 1 month before the onset of their initial or recent recurrent attack of ARF. Three (14%) of these reportedly received a full course of antibiotics, but 14 (67%) either received no antibiotics, or failed to complete a full 10-day course of antibiotics. In the remaining four patients, the parents considered the symptoms to be too mild to bring their children for medical care.

None of the 22 children with recurrent ARF and eight of the 12 patients with RHD were on antistreptococcal chemoprophylaxis at the time of presentation. No prophylactic medications had been prescribed for ten (29%) of the 34 children, and failure to comply with antistreptococcal chemoprophylaxis was noted in the remainder. Among the 24 children who had penicillin prophylaxis prescribed, 13 (54%) dropped completely the continuous use of medication within 1 year after the attack of ARF and three (13%) within 1–5 years. Six (25%) patients were taking their medication irregularly. The prophylactic regimen was terminated by a physician in two patients 5 years after the onset of ARF, but both patients had recurrences within 6 months of the cessation of prophylaxis. An attempt was made to determine precisely the main reasons for failure of the 24 patients to comply with adequate prophylaxis. The families of eight (33%) patients were not properly instructed regarding the importance and/or duration of chemoprophylaxis for prevention of the disease and its grave sequelae; prophylaxis was discontinued in these patients on the basis of apparent recovery. Six (25%) children refused to take their monthly benzathine penicillin injections because of pain, and four of them were switched to oral penicillin which was irregularly taken. The parents of ten (42%) children prematurely terminated prophylaxis because of inaccessibility to medications and because of the long waiting time for injections at out-patient clinics.

Recurrent rheumatic attacks developed once in each of 19 patients (56%), twice in nine patients (26%), and three

### Table 1
Demographic data on 34 children with rheumatic recurrences

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>6–14</th>
<th>10.5 (2.3)</th>
</tr>
</thead>
<tbody>
<tr>
<td>range</td>
<td></td>
<td></td>
</tr>
<tr>
<td>mean (SD)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Males (%)</td>
<td>56</td>
<td></td>
</tr>
<tr>
<td>Residence (%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>urban</td>
<td>62</td>
<td></td>
</tr>
<tr>
<td>non-urban</td>
<td>38</td>
<td></td>
</tr>
<tr>
<td>Socio-economic stratum (%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>low</td>
<td>47</td>
<td></td>
</tr>
<tr>
<td>middle</td>
<td>38</td>
<td></td>
</tr>
<tr>
<td>high</td>
<td>15</td>
<td></td>
</tr>
<tr>
<td>Average family size (SD)</td>
<td>8.9</td>
<td>(2.7)</td>
</tr>
</tbody>
</table>

### Table 2
Cardiac effects of rheumatic recurrences in 32 patients

<table>
<thead>
<tr>
<th>Cardiac effect*</th>
<th>Total (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>(n = 32)</td>
<td></td>
</tr>
<tr>
<td>Isolated MI</td>
<td>18 (56)</td>
</tr>
<tr>
<td>Isolated AI</td>
<td>2 (6)</td>
</tr>
<tr>
<td>Combined MI and AI</td>
<td>9 (28)</td>
</tr>
<tr>
<td>Combined MI, AI, and TI</td>
<td>2 (6)</td>
</tr>
<tr>
<td>Combined MI and MS</td>
<td>1 (3)</td>
</tr>
<tr>
<td>Cardiomegaly</td>
<td>8 (25)</td>
</tr>
<tr>
<td>Pericarditis</td>
<td>1 (3)</td>
</tr>
<tr>
<td>Congestive heart failure</td>
<td>9 (28)</td>
</tr>
</tbody>
</table>

*MI: mitral insufficiency; AI: aortic insufficiency; TI: tricuspid insufficiency; MS: mitral stenosis.
Prevention of Rheumatic Heart Disease

patients could have benefited from primary prevention, but majority of them received inadequate treatment as a result of poor compliance. Bergman and Werner reported that 56% of patients treated in an emergency room had stopped taking oral penicillin by the 3rd day, 71% by the 6th day, and 82% by the 9th day after the diagnosis of streptococcal pharyngitis.16

It is well known that the prevalence and severity of RHD increases with the number of recurrences,1,6,17 and this is due mainly to the increased tendency to recurrences in patients with RHD rather than to the de novo appearance of cardiac involvement in patients initially free of it.18 Exceptions, however, are frequent, and continuous prophylactic antibiotics should be prescribed for all patients with a diagnosis of ARF irrespective of whether or not carditis is present during the initial attack.1,6,19

The discrepancy between a high prevalence and greater severity of cardiac involvement in this series and a low incidence and mild nature of carditis in the initial attack of ARF in Saudi Arabia20 most likely reflects the end result of multiple recurrences due to poor compliance with antistreptococcal chemoprophylaxis.21 This lack of faithful adherence to long-term chemoprophylaxis was partly due to inadequate professional counselling and interest. The education of practitioners is, therefore, vital for rectifying this apparent problem. Recent studies have found a recurrence rate of 2% or less in patients who were consistently maintained on regular antistreptococcal regimens.22 Moreover, if recurrences of ARF were prevented, the heart murmur of mitral regurgitation would disappear in 70% of patients and that of aortic regurgitation in 27% of patients within 10 years of first being noted, and no patient would develop valvular stenosis.21

Although hospital-based studies do not give a true picture of the occurrence of the disease, the data in this series raise an urgent need for a comprehensive preventive campaign directed at both primary and secondary prophylaxis of ARF. Primary prophylaxis entails early detection and adequate treatment of streptococcal pharyngitis.10,15,23 The drug of choice is still oral penicillin or, in the case of penicillin allergy, oral erythromycin, and a full 10 days of therapy is mandatory.17,23,24 Recent studies have confirmed that a treatment regimen for 7 days is significantly less effective than that for 10 days.24 If compliance is in doubt, then long-acting intramuscular benzathine penicillin is preferable.15,23,24 Primary prophylaxis should be focused on public education about the need for prompt and adequate treatment of sore throats.1,22 Primary prophylaxis is the ideal form of prevention, but it can be difficult to implement in many developing countries.1,22 However, Costa Rica has recently been referred to as an example of a country in

times or more in five patients (15%). One patient had apparent, continuous rheumatic activity over a period of months. The mean interval between the initial attack of ARF and first recurrence was 15.3 months with a range of 5 months to 5.5 years; 62% of the patients had their first recurrence within the first year, 24% in the second year, 9% in the third year, and 6% after the first 5 years.

Among the 22 patients with recurrent ARF, two had no cardiac involvement. The cardiac effects of rheumatic recurrences in 32 patients are shown in Table 2. Congestive heart failure was massive and intractable in two patients. Both patients had multivalvular involvement and underwent prosthetic mitral-valve replacement because of massive mitral regurgitation.

Discussion

It is generally believed that RHD is widely prevalent in Saudi Arabia and a strikingly high proportion of paediatric rheumatic patients is undergoing open heart surgery.5,6 Although ARF is a notifiable disease in this country,7 reliable statistics are lacking. However, a prevalence of RHD of 2.4 per 1000 schoolchildren has recently been reported in the western district.8

The incidence of ARF and prevalence of RHD have long been considered to be closely related to environmental factors and in particular to socioeconomic conditions.9 In the present study, these school-age children came predominantly from low and middle-class and relatively large families living under crowded conditions, but all had ready access to free medical care. Crowding, especially within the home, bedroom and school, has been found to facilitate the rapid person-to-person spread of group A streptococci and may increase not only the number of infections but also their virulence and rheumatogenicity.10 The positive family history of ARF in 21% of patients in the present series is certainly high when compared with that of a previous Western study.11 This finding may be attributed to large family size and to similarity of environmental factors within each rheumatic family, but the role of a genetic susceptibility to ARF cannot be ruled out as a contributory factor.12,13 However, improved housing and living conditions, good nutrition, and education are important factors in the control of streptococcal infections and their sequelae.

Earlier studies have shown that 90% of initial attacks of ARF can be prevented by adequate treatment of the preceding streptococcal pharyngitis.14 However, the observation that at least 30–50% of patients with ARF may have no history of preceding throat infection1,15 emphasizes the great difficulty of primary prevention of the disease. In the present series, 38% of the patients had no known history of an apparent antecedent respiratory infection, and, therefore, had no opportunity for primary prevention of ARF. The remaining
which primary prevention has achieved more than 90% decline in ARF over a 10-year period.26

Secondary prophylaxis involves prevention of rheumatic recurrences, by continuous antimicrobial administration (preferably intramuscular benzathine penicillin, 3-weekly or monthly), to individuals who have experienced a prior rheumatic attack.9,22,23 The optimum duration of chemoprophylaxis is a subject of dispute, but in developing countries lifetime continuous prophylaxis has been recommended.1,16,17,19 There is an obvious need for this country to establish a highly organized national rheumatic fever control programme to identify rheumatic individuals, to enrol them into prophylactic programmes, and to assure consistency of prophylaxis. Such a programme can be easily integrated into the currently existing network of primary-care health centres and school health services. Early case-finding gives the greatest return if it is carried out in primary and secondary schools by public health nurses. All detected cases are then referred to the nearby primary-care health centre, given a rheumatic fever identification card, and entered in a register which serves as a tool for follow-up.

This scheme has been tried successfully and discussed in detail elsewhere.1,3,27 It is more cost-effective to institute and maintain an effective rheumatic fever control programme than to manage patients with RHD medically or surgically. In addition, there is a need for carefully designed epidemiological studies to evaluate the complex geographical, host, and microbial factors that influence the distribution of ARF and RHD in this country.

In conclusion, RHD remains a significant public health challenge in Saudi Arabia and the persistence of a largely preventable disease should bring it automatically to the focus of medical attention.

References