Rheumatic Fever in the Kimberley Region of Western Australia

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Summary

The objective of this study was to assess the incidence and clinical presentation of acute rheumatic fever in children and young adults in the north west of Western Australia—a region known as the Kimberley. In this five year retrospective study, residents aged less than 30 years, suspected of having had acute rheumatic fever from 1988 to 1992, were identified and their hospital and clinic records were reviewed. There were 96 cases of acute rheumatic fever that met the revised Jones criteria during the five year period. There were 59 initial attacks and 37 recurrences that occurred in 81 people, of whom 80 were of Aboriginal descent. The overall incidence of acute rheumatic fever in the Kimberley region for those aged under 30 years is 136 cases per 100,000 per year and in the Aboriginal population is 241 per 100,000 per year. The group most at risk are Aboriginal school children aged 5-14 years who accounted for 50 cases and have an annual incidence of 375 cases per 100,000 per year. The clinical presenting features were carditis in 50 per cent, polyarthritis in 59 per cent, chorea in 22 per cent, and subcutaneous nodules in 1 per cent. The hospitalization rate was 80 per cent for new cases and 73 per cent for recurrences with average stays of 14.6 and 1.5 days respectively. Rheumatic heart disease (RMD) developed in 31 of the cases; two required surgery and there was one death attributable to RHD in this group. Rheumatic fever continues to be a significant health problem in the Aboriginal population in this region and the recurrence rate, despite secondary prophylaxis, is also unacceptably high.

Introduction

The incidences of rheumatic fever and rheumatic heart disease have fallen dramatically in industrialized nations in the past 50 years, but they continue to be a major health problem in the developing countries where there has been little change in incidence. Certain populations are at high risk in developed countries, such as the Maoris in New Zealand and Polynesians in Hawaii. A high incidence of hospitalized cases of rheumatic fever in Aboriginal children in Derby was documented by Patten for the period 1975 to 1979 and similar rates have been reported in Aboriginal communities in Queensland and the Northern Territory, although population sizes have been small.

Methods

The Kimberley region is in the far north of Western Australia and covers an area of 421,450 km². It has a total population of 23,375 people and 48 per cent are of Aboriginal descent. There are six towns in the region and a large number of remote Aboriginal communities and outstations. There are six hospitals and health care is provided by Health Department, Community Health, and Kimberley Aboriginal Medical Service staff.

Potential cases were identified retrospectively from each of the hospital’s discharge diagnoses coded for rheumatic fever, rheumatic heart disease, and potentially related articular and valvular disorders for the period January 1988 to December 1992. Rheumatic fever registers and Rural Paediatric Service records provided additional cases. Their hospital and clinic notes were then examined for all patients, between September 1993 and June 1994. Demographic, medical, and laboratory data were recorded.

The diagnosis of rheumatic fever was made if there was documentation fulfilling the revised Jones criteria.
TABLE I
Cases of acute rheumatic fever in the Kimberley, 1988–1992

<table>
<thead>
<tr>
<th>Year</th>
<th>New cases</th>
<th>Recurrences</th>
<th>Total cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>1988</td>
<td>13</td>
<td>9</td>
<td>22</td>
</tr>
<tr>
<td>1989</td>
<td>15</td>
<td>3</td>
<td>18</td>
</tr>
<tr>
<td>1990</td>
<td>9</td>
<td>8</td>
<td>17</td>
</tr>
<tr>
<td>1991</td>
<td>13</td>
<td>7</td>
<td>20</td>
</tr>
<tr>
<td>1992</td>
<td>9</td>
<td>10</td>
<td>19</td>
</tr>
<tr>
<td>1988–1992</td>
<td>59</td>
<td>37</td>
<td>96</td>
</tr>
</tbody>
</table>

and supporting evidence of an antecedent streptococcal infection. The exceptions to these criteria were cases of chorea and indolent carditis where other causes had been excluded. Rheumatic carditis was diagnosed on auscultatory findings with supportive electrocardiograph (ECG) and chest X-ray findings. Echocardiography was generally not available for acute episodes. A recurrent attack was defined as a new episode of rheumatic fever in a patient with an adequately documented initial episode. Those suspected cases of recurrence with only minor criteria were not included. In the case of chorea it may be difficult to differentiate low grade residual chorea with relapse from a recurrence. For this study, relapse was defined as symptoms appearing at least 12 months after resolution of previous chorea.

The Health Services Statistics and Epidemiology branch of the Health Department of Western Australia provided estimates for the resident population under 30 years old in the Kimberley region for 1988 to 1992 which were broken down by age and Aboriginal and non-Aboriginal populations. Incidence rates were calculated only for those who were permanent residents of the region.

Results
There were 96 cases of rheumatic fever identified during the five year period, which occurred in 81 patients. There were 59 new cases and 37 recurrences. The number of cases was similar in each year and is shown in Table 1. The age at diagnosis for new cases (Fig. 1) was 4 to 29 years (mean 13.3) and for recurrences it was 11 to 29 years (mean 17.6). There were 42 female and 39 male patients. They were all Aboriginal except one 20 year old Caucasian stockman. Two cases occurred in visitors to the region. Eighty per cent (47/59) of new cases and 73 per cent (27/37) of recurrences required hospitalization with an average stay of 14.6 and 11.5 days respectively.

Incidence
The overall incidence (Table 2) in the general population under 30 years of age for all attacks was 136 per 100 000 per year and 241 per 100 000 per year in the Aboriginal population. The group at highest risk are Aboriginal school children aged 5–14 years who accounted for 50 cases (37 new and 13 recurrences) and have an incidence of 375 per 100 000 per year. In this age group, 37 cases required hospitalization with an incidence of 278 hospitalized children per 100 000 per year.

Clinical data
See Table 3. Polyarthritis was present in 58 (60 per cent) cases and was the sole major criterion in 32 (33 per cent), most often affecting the knees, ankles, hips, and wrists. There were 20 cases of chorea (21 per cent) in 17 patients (13 female and 4 male) with two patients having prolonged courses with multiple relapses over several years. Subcutaneous nodules were seen in one patient with polyarthritis, and erythema marginatum was not documented.

Carditis was a presenting feature in 48 (50 per cent) of the cases (43 patients) with a similar rate for both first and recurrent attacks. There was associated polyarthritis in 25 (26 per cent) patients and chorea in five (5 per cent) others. Mitral regurgitation (MR) was present in 34 cases, aortic regurgitation (AR) in two, and both AR and
Table 2
Annual incidence of rheumatic fever in Kimberley and Aboriginal population by age category

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>Estimated population</th>
<th>Cases of rheumatic fever</th>
<th>Annual incidence per 100000 population (95% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total population</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>0–4</td>
<td>2788</td>
<td>1</td>
<td>7.2 (0.2–40)</td>
</tr>
<tr>
<td>5–14</td>
<td>4327</td>
<td>50</td>
<td>231 (172–305)</td>
</tr>
<tr>
<td>15–29</td>
<td>6671</td>
<td>43</td>
<td>129 (93–173)</td>
</tr>
<tr>
<td>Total 0–29</td>
<td>13786</td>
<td>94</td>
<td>136 (110–167)</td>
</tr>
<tr>
<td>Aboriginal population</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>0–4</td>
<td>1804</td>
<td>1</td>
<td>11.1 (0.3–62)</td>
</tr>
<tr>
<td>5–14</td>
<td>2664</td>
<td>50</td>
<td>375 (285–495)</td>
</tr>
<tr>
<td>15–29</td>
<td>3258</td>
<td>42</td>
<td>258 (186–348)</td>
</tr>
<tr>
<td>Total 0–29</td>
<td>7726</td>
<td>93</td>
<td>241 (194–295)</td>
</tr>
</tbody>
</table>

Table 3
Major manifestations of acute rheumatic fever

<table>
<thead>
<tr>
<th>Manifestation</th>
<th>New cases</th>
<th>Recurrences</th>
<th>Total cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Carditis (isolated)</td>
<td>13</td>
<td>5</td>
<td>18</td>
</tr>
<tr>
<td>Carditis and polyarthritis</td>
<td>12</td>
<td>13</td>
<td>25</td>
</tr>
<tr>
<td>Carditis and chorea</td>
<td>4</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>Chorea (isolated)</td>
<td>9</td>
<td>6</td>
<td>15</td>
</tr>
<tr>
<td>Polyarthritis (isolated)</td>
<td>20</td>
<td>12</td>
<td>32</td>
</tr>
<tr>
<td>Polyarthritis and subcutaneous nodules</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
</tbody>
</table>

MR in 12 cases. Pericarditis was seen in four patients. Severe carditis with cardiac failure occurred in six patients, none of whom had associated arthritis or chorea. One patient with persistent cardiac failure required mitral valve plication. These patients required prolonged hospitalization with an average stay of 50 days. There were three patients who presented with indolent carditis.

Rheumatic heart disease (RHD) was diagnosed clinically in 31 patients (confirmed in 23 with echocardiography). This occurred in 18 of 29 new cases of carditis and 13 of 14 patients with recurrences of carditis. All patients with severe carditis developed RHD. Isolated mitral regurgitation occurred in 14 patients, mitral stenosis (MS) and MR in three, MR and AR in eight, isolated aortic regurgitation in two and MR, MS, and AR in five patients. Two patients have required valve replacements: one patient declined surgery and one patient was not fit for surgery due to poor left ventricular function and died of cardiac failure aged 18 years.

Recurrences
There was a total of 37 recurrent episodes documented in 28 patients of whom six had had their first episode during the study period. The length of time from previous attack ranged from four months to 17 years with a median interval of four years (see Table 4). Penicillin prophylaxis prior to recurrence was not being used in 10 cases, oral penicillin V (phenoxymethylpenicillin) in eight cases, and intramuscular procaine penicillin LA Bicillin in the other 19 cases although poor compliance was noted in 17. The major criteria differed in seven of the recurrences from their first attack, including four with new carditis. The recurrence rate for those with first attacks during this period was 4.8 per 100 patient years (95 per cent confidence interval 1.8–10).

Discussion
In the Kimberley region, the incidence of acute rheumatic fever in Aboriginal school children and young adults is extremely high, confirming the susceptibility in this population reported elsewhere in Australia. Despite considerable efforts to improve access to health care, living conditions, and health education in this population, there is no evidence of a downward trend during this period and the incidence of
hospitalized cases is similar to that in the late 1970s in
Derby reported by Patten (227–353 per 100 000 children
per year). The incidence is higher than that reported in
developing countries and other high risk populations
such as New Zealand Maoris and Samoan children
in Hawaii. This current incidence is similar to that in
Western Europe at the turn of the century.

The limitations of this retrospective study include
inaccuracies of available data, availability of specialist
opinions, and appropriate investigations in remote areas.
Trendy to overdiagnosis was minimized by adherence
to the revised Jones criteria. Case underestimation may
have resulted from lack of data, particularly streptococ-
cal serology, premature administration of salicylates in
monoarthritis, and patients in remote areas not present-
ing to medical services.

The accuracy of population estimates of Aboriginal
people is affected by their mobility and problems in
collecting data; however, the Australian Bureau of
Statistics now uses special procedures to improve accuracy
of census figures and minimize underestimation.

This study has shown that rheumatic fever remains a
major health problem in the Aboriginal population in the
Kimberley. The first attack usually occurs in school age
children with a slight preponderance in females,
reflecting their increased susceptibility to chorea. The
sex difference is less than that noted by Patten. New
cases in adults may represent recurrences where the
initial episode is missed. The absence in Caucasian
children is seen despite contact in schools with a high
risk population.

There are a number of reasons for the high rates seen
in the Aboriginal population. They have poor living
conditions with overcrowding and reduced access to
health care which are associated with increased inci-
dence of RF. The high prevalence of streptococcal
infections in Aboriginal children is well known but there
is little information regarding pharyngitis and asympto-
matic carriage. The importance of adequate treatment of
pharyngitis in this high risk population is not always
appreciated by health professionals, particularly new
staff. There is also a reluctance to seek treatment in this
age group and compliance with oral medication is often
poor. The role of virulent serotypes of streptococci has
assumed more importance with outbreaks of RF in the
USA but their role here is not known. There may be a
genetic predisposition in Aboriginals to rheumatic fever
but this has not been studied.

The clinical presentation in this series is similar to that
generally reported, with arthritis being the most common
manifestation. Carditis, occurring in half the cases, is
similar to that seen in other series and was associated
with the highest morbidity. The presence of associated
arthritis or chorea appears to be protective against severe
carditis. The significance of recurrent carditis progres-
sing to rheumatic heart disease and new carditis
occurring in a recurrence after a long interval illustrate
the importance of maintaining good compliance with
prophylaxis in all patients.

The cost to the community of rheumatic fever is
significant, with immediate costs for hospitalizations and
for subsequent rheumatic heart disease, as documented
in New Zealand, and underlines the need for more
effective primary and secondary prevention. Primary
prevention requires the identification and adequate
treatment of streptococcal upper respiratory tract
infections. This requires education of the at-risk
population to encourage early presentation, better
compliance with treatment, and awareness in health
professionals of the problem. In Costa Rica, there was
a marked decline in the incidence of RF following a
policy of treating all cases of tonsillitis with benzathine
penicillin, although this policy may make people more
reluctant to seek medical assistance. In a Queensland
Aboriginal community, regular surveillance in school
children with throat swabs and appropriate treatment of
positive children and family contacts resulted in the
virtual disappearance of rheumatic fever during the
study. However, with decreased community interest and
reduced compliance, the quick return to previous rates
highlighted their susceptibility. This illustrates the
problems of maintaining health programmes in Abori-
ginal communities and this would be more difficult over
a whole region where it is not perceived as a high
priority.

An effective secondary prevention programme is
essential with such a high incidence of rheumatic
fever. The recurrence rate in this region remains
unacceptably high. The maintenance of regularly
updated registries is required, but although this does
occur in the Kimberley Aboriginal Medical Service,
other registries were found to have a significant dropout
rate. In addition, the highly mobile nature of the
population causes problems in ensuring regular prophy-
laxis where registries are not all linked. The notification
of all cases of rheumatic fever in the region to a
centralized source with standardized data may facilitate
this process as well as enable prospective monitoring of
incidence and morbidity.

Conclusions

Rheumatic fever remains a significant health problem in
the Aboriginal population in the Kimberley. This
regional survey has documented an incident rate that is
one of the highest reported in the literature and is
not decreasing. The recurrence rate, despite secondary
prophylaxis, is also unacceptably high. This has important
implications for health professionals and planning in the
region. Community awareness of the problem needs to be
raised.

References

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2. Neutze J. Rheumatic fever and rheumatic heart disease in