Retrospective epidemiology of acute rheumatic fever: a 10-year review in the Waikato District Health Board area of New Zealand

Victoria Pennock, Anita Bell, Te Aro Moxon, Peter Reed, Fraser Maxwell, Diana Lennon

Abstract

Background Acute rheumatic fever (ARF) is a preventable disease which remains a prominent burden of health in New Zealand, with an annual incidence comparable to that of developing countries.

Aim The aim of this study was to describe the epidemiology of ARF and recurrent ARF cases in the Waikato District Health Board (DHB) area of New Zealand from 1 January 2002 to 31 December 2011.

Methods A total of 106 cases of ARF and four cases of recurrent ARF were identified through the Public Health Database – EpiSurv and the Hospital coding system, ICD-10.

Results The overall Waikato DHB annual incidence of ARF was 3.1 per 100,000 population with Māori children aged 5–14 years experiencing higher rates of 46.1 per 100,000 population. Eighty-five percent of the cases were of Māori ethnicity, and 10% Pacific. Almost three-quarters of all cases lived in areas of the three most deprived deciles as described by the New Zealand Deprivation Index 2006.

Discussion The rates of ARF seen in the Waikato DHB are comparable to that seen previously locally and nationally. High risk groups have been identified as children aged 5–14 years, Māori and Pacific ethnicity, and those living in lower socioeconomic areas which could be targeted by the Rheumatic Fever Prevention Programme (RFPP) with the intention to reduce the incidence of ARF nationally to 0.4 cases per 100,000 population by 2017.

Acute rheumatic fever (ARF) is an autoimmune response to a sore throat caused by the bacteria, Group A Streptococcus (GAS). This can subsequently lead to chronic damage to the heart – rheumatic heart disease (RHD).1

ARF rates have reduced worldwide, with the highest rates experienced in developing countries within the school-aged population.2,3

New Zealand has yet to see the removal of ARF as a public health burden, and consistently experiences higher rates in Māori and Pacific children compared to European.4–6 The disproportionally high rates of AFR in Māori and Pacific children have been attributed to poor socioeconomic conditions, in particular overcrowding and inadequate access to health care.5–7

ARF and RHD remain a significant cause of morbidity, mortality and cost to patients, families and the New Zealand Health Service.8,9 A 2012 study showed that the
average annual diagnosis related group based cost of hospital admissions for ARF and RHD in New Zealand was $12.0 million, yet a ten day course of oral penicillin can treat the preceding streptococcal throat infection that leads to ARF.

To prevent a recurrence of ARF, secondary prophylaxis is instigated. In New Zealand this involves an intramuscular injection of antibiotic every 28 days, for a minimum of 10 years, depending on the extent of carditis.

The Waikato District Health Board area (DHB) has a population of just under 340,000; approximately 8% of the population of New Zealand. Locally, over 30 years ago, the Hamilton Health District established a register for ARF and RHD with community nurses delivering penicillin, but it was not until 1986 that ARF became a notifiable disease.

In the early 1990s the Hamilton register was discontinued as a result of health systems reorganisation. Subsequently, administration of prophylaxis became disorganised as patients moved location or missed injections.

A Waikato DHB study through 1998–2004 found an annual ARF incidence of 12.9 per 100,000 in the 5–14 year age group, with over 80% of cases of Māori ethnicity. In view of this, in 2008 the Waikato DHB Rheumatic Fever Prevention and Management group was established to reduce the incidence and burden of the disease. This included community and health professional awareness raising, in particular regarding sore throats, and an audit of cases to improve their management both in the community and in hospital. A register was set up within Population Health for the Waikato DHB, however a national register has never been implemented.

In other countries such as the United States and Cuba, disease registers have been shown to facilitate follow up and effective antibiotic delivery. An Auckland study showed that the introduction of a rheumatic fever register led to a reduction in ARF rates to 0.14 per 100 patient years from 1.5 per 100 patient years (1972–1981).

In 2013 in response to the persisting high rates of ARF the New Zealand Ministry of Health released an intention to reduce the incidence of ARF hospitalisations to 0.4 cases per 100,000 population nationally by 2017. This goal is hoped to be achieved via a $24 million rheumatic fever prevention programme targeting certain high burden areas, of which Waikato is included.

This study was conducted as part of a national research project collecting retrospective data throughout New Zealand over a ten year period with the aim to gather concise, up-to-date epidemiological data.

Method

An audit of clinical notes was conducted in all Waikato DHB resident patients who were identified as having a primary or secondary diagnosis of acute and recurrent ARF in the Waikato DHB from 1 January 2002 to 31 December 2011.

To identify cases of ARF and recurrent ARF, two databases were searched and cross referenced; the hospital admission database using the ICD-10 coding system and the EpiSurv database, which is a national database of notifiable diseases held by Population Health.

The ICD-10 coding for ARF was used to identify potential cases, (ICD codes – l00, l00.0, l01.0, l01.1, l01.2, l01.8, l01.9, l02, l02.0, l02.9) and files coded as Rheumatic Heart Disease in patients aged 0–35 years were also collected and reviewed in case any of these had been coded incorrectly.
Information from identified cases was extracted from either written hospital notes, the Clinical Results Viewer (an electronic document and results viewer) or the EpiSurv database and inputted into a set template. The 1992 Jones criteria, with the New Zealand modifications (such as the use of echocardiography as evidence of carditis in absence of murmur as major criteria) was used to confirm the diagnosis of ARF and recurrent ARF. The Heart Foundation Guidelines were then used to categorise these cases into definite, probable and possible cases.

The address at diagnosis was recorded and subsequently, The New Zealand Index of Deprivation (2006) was used as an area measure of deprivation at this address. The data used for population statistical comparison was obtained from the 2006 Statistics New Zealand Census of population and dwellings.

**Results**

From 1 January 2002 to 31 December 2011 a total of 106 ARF cases and four recurrent episodes were identified as meeting the case definitions.

**Figure 1. Diagram of case inclusion for ARF**
The EpiSurv database identified 126 ARF cases, with three cases initially coded as recurrent ARF (Figure 1). These three cases after note review were found to be the first episodes of rheumatic fever, so were included as such. Of 126 cases, 21 were excluded; 3 did not meet diagnostic criteria, 3 were a proven diagnosis of arthritis, 1 case had two NHI numbers, 3 patients lived in a different area, 6 had RHD and for 5 cases there was insufficient data to confirm diagnosis. This gave a total of 105 cases of ARF.

Figure 2. Diagram of case inclusion for recurrent ARF

217 potential ARF cases were identified using the hospital discharge coding system. Sixty-six of these were duplicates of cases found from the EpiSurv data and 148 were not cases of ARF, usually being picked up on the ICD codes with either other non-RF heart or bone disease.

Only 3 cases had not been identified on the EpiSurv database. Of these previously unidentified cases, data could not be obtained for 2, so only 1 case was added to those found using EpiSurv, giving a total of 106 cases of ARF.

For recurrent ARF, 16 cases were identified by the EpiSurv Database (Figure 2). Of these, three were actually ARF cases and nine cases did not meet the stipulated
criteria because the diagnosis had been made retrospectively, there was insufficient data, or there was inadequate information or investigations conducted or recorded. Thus a total of four cases were identified.

The hospital database using the ICD-10 criteria identified 56 potential recurrent cases. Two of the cases of recurrent ARF were already identified by the EpiSurv database and ten cases were actually cases of ARF when the notes were reviewed. The remaining 44 cases had been coded inaccurately and were not episodes of recurrent ARF, the most common example was they had been referred to Waikato Hospital for heart valve surgery for RHD and coded for this. No new cases were added, giving a total of four recurrent ARF cases.

Of the 106 cases of ARF identified, 36% (38) were female. The age range was 4–37 years with the mean age being 11.8 years and median age of 11 years (Figure 3). Approximately 80% of cases were aged 5–14 years, with almost 92% aged 5–19 years.

**Figure 3. Age distribution of ARF cases in the Waikato DHB, 2002–2011**

![Age distribution of ARF cases](image)

Approximately 85% (90) of cases were of Māori ethnicity, 10% (10) Pacific and 4% (1) European (Figure 4). Eighty percent (73) of Māori cases and 90% (9) of the Pacific cases were aged between 5–14 years. There was 1 case for which the ethnicity was unknown. For the purpose of statistical analysis this case was designated as Māori ethnicity based on the high probability and name of the child.
Figure 4. Incidence rates of ARF by ethnicity, Waikato DHB, 2002–2011

![Incidence rates of ARF by ethnicity](image)

The annual incidence of ARF for Māori children aged 5–14 years was 46.1 per 100,000 population, approximately fifteen times that of the general population (Table 1). Confidence intervals were calculated for incidence rates using a Poisson 95% confidence limit.

<table>
<thead>
<tr>
<th>Variables</th>
<th>Number of patients</th>
<th>Population</th>
<th>Incidence cases/100,000 population/year</th>
<th>Poisson 95% confidence limit</th>
</tr>
</thead>
<tbody>
<tr>
<td>All population</td>
<td>106</td>
<td>339,133</td>
<td>3.1</td>
<td>2.56–3.78</td>
</tr>
<tr>
<td>All 5–14 yrs</td>
<td>84</td>
<td>52,920</td>
<td>15.9</td>
<td>12.66–19.66</td>
</tr>
<tr>
<td>Māori 5–14yrs</td>
<td>74</td>
<td>16,049</td>
<td>46.1</td>
<td>36.21–57.89</td>
</tr>
<tr>
<td>Pacific 5–14yrs</td>
<td>9</td>
<td>1677</td>
<td>53.7</td>
<td>24.54–101.88</td>
</tr>
<tr>
<td>European 5–14yrs</td>
<td>1</td>
<td>25,496</td>
<td>0.39</td>
<td>0.001–2.19</td>
</tr>
</tbody>
</table>

The yearly incidence of ARF in Waikato DHB varies by year, with 19% (20) occurring in 2008, and only 4% (4) in 2003 (Figure 5).

104 cases of ARF were geocoded and analysed by the New Zealand index of deprivation, 2006. Almost 88% (91) of cases lived in an area with a decile score of 5–10, and three-quarters (76) in the most deprived areas (7–10) (Figure 6).
Using the Heart Foundation Guidelines\textsuperscript{11} to assess ARF cases, 83\% (88) were defined as a definite case, 9\% (10) a probable case and 8\% (8) a possible case.

The most common major criteria identified in cases was evidence of carditis (76\%), including evidence of subclinical carditis, and most common minor feature used was ESR (87\%) (Table 2).
Table 2. Number and percentage of cases by major and minor criteria for ARF

<table>
<thead>
<tr>
<th>Major criteria and minor criteria</th>
<th>Number of cases</th>
<th>Percentage of cases</th>
<th>No data available</th>
</tr>
</thead>
<tbody>
<tr>
<td>Carditis</td>
<td>81</td>
<td>76%</td>
<td>5</td>
</tr>
<tr>
<td>Arthritis</td>
<td>50</td>
<td>47%</td>
<td>7</td>
</tr>
<tr>
<td>Chorea</td>
<td>12</td>
<td>11%</td>
<td>4</td>
</tr>
<tr>
<td>Erythema marginatum</td>
<td>5</td>
<td>5%</td>
<td>7</td>
</tr>
<tr>
<td>Fever (&gt;38°C)</td>
<td>48</td>
<td>45%</td>
<td>17</td>
</tr>
<tr>
<td>Prolonged P-R interval</td>
<td>42</td>
<td>40%</td>
<td>16</td>
</tr>
<tr>
<td>Arthralgia</td>
<td>63</td>
<td>59%</td>
<td>7</td>
</tr>
<tr>
<td>Erythrocyte sedimentation rate (ESR)</td>
<td>92</td>
<td>87%</td>
<td>1</td>
</tr>
<tr>
<td>C-reactive protein (CPR)</td>
<td>79</td>
<td>75%</td>
<td>6</td>
</tr>
</tbody>
</table>

Of the cases of ARF without chorea who had a documented history of a preceding sore throat (48), 29% (14) had a positive throat swab for *Streptococcus* bacteria. Forty-six percent (22) were negative and 25% (12) had no data to indicate whether a throat swab had been taken or not, however all cases fulfilled the criteria for evidence of streptococcal infection with raised blood titre levels.11

Of the four cases of recurrent ARF identified two were of Māori and two were of European ethnicity. The age range was 22–35 years with a mean age of 26 and median age of 23. The rate of recurrence was 0.12 per 100,000 population.

Using NZDep20 three cases lived in an area of high deprivation. In one case the recurrence happened two years after prophylaxis had stopped which was in keeping with national guidance and without a history of a preceding throat infection. For the other three cases, there was only data to show that one of the cases was compliant with prophylaxis at the time of recurrence.

**Discussion**

This study has shown that ARF still persists in the Waikato DHB area with the overall mean annual incidence for 2002–2011 at 3.1 per 100,000, which is in keeping with national rates of 3.4 per 100,000 population.5 This is in comparison to other developed countries where the rates of ARF have reduced, with annual rates reported as below 1 per 100,000 population.1

The incidence of ARF is much more prominent in the 5–14 year age group showing an incidence 15.8 per 100,000 population, which is as observed internationally and the results for this particular area in New Zealand are comparable to previous local and national data.5,12,13,21

For Māori and Pacific children aged 5–14 years the rate of AFR was 46.1 and 53.7 respectively per 100,000 population, much greater than that of European children (0.39).

Previous studies of rheumatic fever have shown similar disproportionate results.5,12,13,15 A review of the Auckland Rheumatic Fever Register 1993–1999 for 5–14 year olds described rates of 42.8 per 100,000 population for Māori, 84.9 per 100,000 population for Pacific, and 1.4 per 100,000 for European children.15 The numbers of Pacific children resident in the Waikato area are low, and only 1 European
child had ARF. This data stresses the importance of targeting the Māori population with public health initiatives.

Previous studies have also linked ARF with low socioeconomic status, and this study again corroborates this as almost three quarters of all Waikato DHB ARF cases lived in areas with the three most deprived decile scores. This link has been attributed to overcrowding, poor housing conditions and poor access to health care.¹ 7

A New Zealand study in 1996 observed that three quarters of people living in crowded houses were of Māori or Pacific origin, again highlighting this at-risk group.⁷

So far there is no clear evidence to suggest a genetic link to ARF; however family clusters of ARF have been seen which may indicate a certain predisposition to developing ARF. It is still not understood why in certain people a group A Streptococcal throat infection can lead to ARF, and not in others.¹ ² ²²

Carditis was found to be a major criteria in 76% (81) cases, chorea in 11% (12) of cases and a raised ESR was used as a minor criteria in 87% (92) of cases. This is in keeping with the epidemiology seen in Northland for the same time period.²³ In addition the Northland study also found a median age of 11.4 years, and gender split of 60% male and 40% female which mirrors our results.²³

Limitations—The Heart Foundation Guidelines¹¹ recommends that everyone with ARF be admitted to hospital; therefore using hospital notes to collect data should be adequate to collect epidemiological data of this disease. There may be cases that were diagnosed in primary care, and some cases that may not have presented at all in the acute instance, or misdiagnosed by medical professionals but later present with rheumatic heart disease. It was hoped that by using the ICD-10 codes and EpiSurv data, that the cases that did not present to hospital could be located.

Of all the potential cases, notes were not available for five cases, and in six cases information could only be found on the hospital electronic system. The ICD-10 criteria used were very broad in the hope to pick up cases that otherwise may have been missed. It is not surprising that many cases were not in fact recurrent or ARF and therefore not included in this data.

During note review, several issues were identified. Firstly, some clinicians were not aware of the NZ interpretation of the Jones criteria and publications of NZ specific criteria¹¹ that allows for subclinical carditis and monoarthritis, with some patients not undergoing echocardiograms and therefore not able to fulfil criteria for definite rheumatic fever. Secondly, there was variation by the clinician in regards to the diagnosis of arthritis and/or arthralgia, and blood serology being requested when throat swabs were negative for GAS. Medical practitioners should be knowledgeable or at least know how to access the guidelines on the diagnostic criteria for ARF to ensure accurate and prompt diagnosis, and prevent further cardiac damage from occurring.

Implications—Of all the cases included in this study, only half (48%) had a documented preceding sore throat, and in a fifth (18%) there was no data regarding sore throats. The Heart Foundation has developed a pathway to treat sore throats in New Zealand,¹¹ however for this to be effective, the general population must know the importance and implications of a sore throat so to seek medical attention, and
medical practitioners must be aware of this literature to actively seek the history and adequately treat the sore throat.

There is evidence to show treatment GAS pharyngitis with 10 days of penicillin reduces the rate of ARF. A recent randomized controlled trial assessed the primary prevention of ARF with early treatment of sore throats in school based children in South Auckland.

The introduction of a free nurse-led service providing oral penicillin for the treatment of Streptococcus A pharyngitis produced a 21–28% reduction in ARF rates over a 3-year period. This study was included in a meta-analysis of the primary prevention of ARF with treatment of a Strep. throat, which showed a relative risk of 0.41 (95%CI: 0.23–0.70) and a true treatment effect of approximately 60%. Hence as part of the Rheumatic Fever Prevention Programme (RFPP) there is a school-based throat swab initiative within the high-risk communities.

A sore throat can be seen as an insignificant ailment to the general population, and so the RFPP also identifies the need to raise community and health sector awareness. Current action is being taken to encourage families of children with sore throats to seek early treatment and to train health care professionals in sore throat management in high risk areas.

The rate of recurrence of ARF in this study was 0.12 per 100,000 population which is similar to rates seen in Auckland since 1981 with a functioning Rheumatic Fever Register. The numbers of ARF identified by the ICD-10 criteria and the Public Health EpiSurv, were almost exact, with just one further case added by using the ICD-10 criteria.

Establishment of a national Rheumatic Fever Register has been shown nationally and internationally to reduce recurrent rates, and would also serve to provide excellent epidemiological data on the disease.

**Conclusion**—The annual incidence of ARF for the overall Waikato DHB population was 3.1 per 100,000 population which highlights a clear health need to be addressed by the RFPP. High risk groups have been identified as children aged 5–14 years, Māori and Pacific ethnicity, and living in lower socioeconomic status.

To reach the ARF incidence targets proposed by the RFPP of 0.4 cases per 100,000 population by 2017, there would need to be a significant reduction of incidence of over 80% in the Waikato area. This is proposed to be achieved via multiple interventions such as addressing housing issues, sore throat clinics and promotion of community and clinician awareness, however there will need to be a substantial impact by the RFPP to meet this aim.

The data produced in this study can be used as baseline data prior to the RFPP interventions taking place, and the Hamilton EpiSurv database can then be used to assess effectiveness and trends in the coming years with continuing rigorous application of the case definitions.
Competing interests: Nil.

Author information: Victoria Pennock, Paediatric Registrar, Waikato District Health Board, Hamilton; Anita Bell, Public Health Physician, Waikato District Health Board, Hamilton; Te Aro Moxon, Paediatric Registrar, Auckland District Health Board, Auckland; Peter Reed, Statistician, Auckland District Health Board, Auckland; Fraser Maxwell, Paediatrician, Waikato District Health Board, Hamilton; Diana Lennon, Professor of Population of Children and Youth, Department of Paediatrics, The University of Auckland

Correspondence: Dr Victoria Pennock, Waikato District Hospital, Pembroke Street, Private Bag 3200, Hamilton 3420, New Zealand. Email: victoria.pennock@waikatodhb.health.nz

References:


