**RHEUMATIC FEVER AND HEART DISEASE**

**Introduction**

Acute rheumatic fever is due to a delayed immune response which develops in response to a group A streptococcal throat infection (typically about 3 weeks after the sore throat). It usually occurs in school-age children and may affect the brain, heart, joints, skin or subcutaneous tissue [106]. Recurrent episodes of rheumatic fever may result in the development of rheumatic heart disease, a progressive condition leading to damage, scarring and deformities of the heart valves. Surgery to repair or replace damaged valves may be required [163].

While New Zealand’s rheumatic fever rates have declined significantly during the past 30 years, they still remain higher than those of many other developed countries. Risk factors include age (school age children), ethnicity (Pacific>Māori>European), socioeconomic disadvantage and overcrowding [164]. Primary prevention focuses on the adequate treatment of streptococcal throat infections, while secondary prevention aims to ensure that those previously diagnosed with rheumatic fever receive monthly antibiotic prophylaxis, either for 10 years from their first diagnosis or until 21 years of age (whichever is longer), to prevent recurrent rheumatic fever [165].

The following section explores rheumatic fever and heart disease rates in children and young people using information from the National Minimum Dataset and Mortality Collection. The section concludes with a brief overview of policy and evidence-based review documents which consider interventions to prevent rheumatic fever and rheumatic heart disease at the population level.

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**Data Sources and Methods**

**Indicator**

1. **Acute and Semi Acute Hospital Admissions for Children and Young People Aged 0–24 Years with Acute Rheumatic Fever or Rheumatic Heart Disease listed in any of their first 15 diagnoses.**
   
   **Numerator:** National Minimum Dataset: Acute and semi-acute hospital admissions for children and young people aged 0–24 years with Acute Rheumatic Fever (ICD-10-AM I00–I02) or Chronic Rheumatic Heart Disease (I05–I09) listed in any of the first 15 diagnoses.
   
   **Denominator:** Statistics NZ Estimated Resident Population (with linear extrapolation being used to calculate denominators between Census years).

2. **Mortality from Acute Rheumatic Fever or Rheumatic Heart Disease in Children and Young People Aged 0–24 Years**
   
   **Numerator:** National Mortality Collection; Deaths in children and young people aged 0–24 years where the main underlying cause of death was Acute Rheumatic Fever or Rheumatic Heart Disease (I00–I09).
   
   **Denominator:** Statistics NZ Estimated Resident Population (with linear extrapolation being used to calculate denominators between Census years).

**Notes on Interpretation**

Note 1: Unless otherwise specified, this analysis focuses on hospital admissions for children and young people with either acute rheumatic fever or chronic rheumatic heart disease listed in any of the first 15 diagnoses (rather than on the subset of admissions where these diagnoses were listed only as the primary diagnosis). The rationale for this wider focus was the fact that many children and young people with chronic rheumatic heart disease will not be hospitalised for their heart disease per se, but rather for one of its resulting complications. For example, during 2005–2009 only 39.0% of hospitalisations for children and young people with rheumatic heart disease had this listed as the primary diagnosis, with 11.8% being admitted for pregnancy and childbirth, and 11.0% for other cardiovascular diagnoses [145].

Note 2: An acute admission is an unplanned admission occurring on the day of presentation, while a semi-acute admission (referred to in the NMDS as an arranged admission) is a non-acute admission with an admission date <7 days after the date the decision was made that the admission was necessary.

Note 3: **Appendix 3** outlines the limitations of the hospital admission data used. The reader is urged to review this Appendix before interpreting any trends based on hospital admission data.
New Zealand Distribution and Trends

New Zealand Trends
In New Zealand, hospital admissions for children and young people with acute rheumatic fever declined gradually during the early-mid 2000s, but then increased again after 2006–07. In contrast, admissions for those with rheumatic heart disease were relatively static during the mid-2000s, although a downswing in rates was evident in 2010. During 2000–2008, on average one child or young person each year died as the result of acute rheumatic fever or rheumatic heart disease (Figure 88).

Figure 88. Acute and Semi-Acute Hospital Admissions (2000–2010) and Deaths (2000–2008) from Acute Rheumatic Fever and Rheumatic Heart Disease in New Zealand Children and Young People Aged 0–24 Years

New Zealand Distribution by Age
In New Zealand during 2006–2010, hospital admissions for acute rheumatic fever were relatively infrequent during infancy, but increased rapidly during childhood, to reach a peak at 11 years of age. Hospital admissions for rheumatic heart disease also increased during childhood, to reach a peak at 12 years of age. In contrast, during 2004–2008 mortality from acute rheumatic fever or rheumatic heart disease was more common amongst those in their late teens and early twenties (Figure 89).
Figure 89. Acute and Semi-Acute Hospital Admissions (2006–2010) and Deaths (2004–2008) from Acute Rheumatic Fever and Rheumatic Heart Disease in New Zealand Children and Young People by Age

Source: Numerators: National Minimum Dataset (Acute and semi-acute admissions with Acute Rheumatic Fever or Rheumatic Heart Disease listed in any of the first 15 diagnoses) and National Mortality Collection; Denominator: Statistics NZ Estimated Resident Population

Figure 90. Acute and Semi-Acute Hospital Admissions for Acute Rheumatic Fever and Rheumatic Heart Disease in Children and Young People Aged 0–24 Years by Ethnicity, New Zealand 2000–2010

Source: Numerator: National Minimum Dataset (Acute and semi-acute admissions with Acute Rheumatic Fever or Rheumatic Heart Disease listed in any of the first 15 diagnoses); Denominator: Statistics NZ Estimated Resident Population. Note: Ethnicity is Level 1 Prioritised.

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Table 89. Acute and Semi-Acute Hospital Admissions for Acute Rheumatic Fever and Rheumatic Heart Disease in Children and Young People Aged 0–24 Years by Ethnicity, NZ Deprivation Index Decile and Gender, New Zealand 2006–2010

<table>
<thead>
<tr>
<th>Variable</th>
<th>Rate</th>
<th>Rate Ratio</th>
<th>95% CI</th>
<th>Variable</th>
<th>Rate</th>
<th>Rate Ratio</th>
<th>95% CI</th>
</tr>
</thead>
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<tr>
<td>NZ Deprivation Index Quintile</td>
<td></td>
<td></td>
<td></td>
<td>Prioritised Ethnicity</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Decile 1–2</td>
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<td>1.00</td>
<td></td>
<td>European</td>
<td>1.41</td>
<td>1.00</td>
<td></td>
</tr>
<tr>
<td>Decile 3–4</td>
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<td>2.15</td>
<td>1.31–3.53</td>
<td>Māori</td>
<td>35.6</td>
<td>25.3</td>
<td>19.4–33.0</td>
</tr>
<tr>
<td>Decile 5–6</td>
<td>4.73</td>
<td>2.89</td>
<td>1.80–4.64</td>
<td>Pacific</td>
<td>62.2</td>
<td>44.2</td>
<td>33.7–57.8</td>
</tr>
<tr>
<td>Decile 7–8</td>
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<td>7.07</td>
<td>4.58–10.9</td>
<td>Asian/Indian</td>
<td>0.65</td>
<td>0.46</td>
<td>0.20–1.07</td>
</tr>
<tr>
<td>Decile 9–10</td>
<td>41.6</td>
<td>25.4</td>
<td>16.8–38.5</td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gender</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>12.3</td>
<td>1.00</td>
<td></td>
<td>Male</td>
<td>16.4</td>
<td>1.34</td>
<td>1.18–1.51</td>
</tr>
</tbody>
</table>

Rheumatic Heart Disease 0–24 Years

| NZ Deprivation Index Quintile | | | | Prioritised Ethnicity | | | |
| Decile 1–2 | 1.14 | 1.00 | | European | 1.97 | 1.00 | |
| Decile 3–4 | 3.44 | 3.03 | 1.72–5.33 | Māori | 19.3 | 9.78 | 7.69–12.4 |
| Decile 5–6 | 3.73 | 3.27 | 1.87–5.73 | Pacific | 47.2 | 23.9 | 18.8–30.4 |
| Decile 7–8 | 7.84 | 6.89 | 4.09–11.6 | Asian/Indian | 0.87 | 0.44 | 0.21–0.91 |
| Decile 9–10 | 26.0 | 22.8 | 13.9–37.5 | | | | |
| Gender | | | | | | | |
| Female | 10.7 | 1.00 | | Male | 8.89 | 0.83 | 0.72–0.96 |

Source: Numerator: National Minimum Dataset (Acute and semi-acute admissions with Acute Rheumatic Fever or Rheumatic Heart Disease listed in any of the first 15 diagnoses ); Denominator: Statistics NZ Estimated Resident Population. Note: Rate is per 100,000; Ethnicity is Level 1 Prioritised; Decile is NZDep2001.

Figure 91. Average Number of Acute and Semi-Acute Hospital Admissions for Acute Rheumatic Fever and Rheumatic Heart Disease in Children and Young People Aged 0–24 Years by Month, New Zealand 2006–2010

Source: National Minimum Dataset (Acute and semi-acute admissions with Acute Rheumatic Fever or Rheumatic Heart Disease listed in any of the first 15 diagnoses ).
New Zealand Distribution by Ethnicity, NZDep Index Decile and Gender
In New Zealand during 2006–2010, hospital admissions for acute rheumatic fever were significantly higher for males, Pacific > Māori > European and Asian/Indian children and young people and those from average-to-more deprived (NZDep decile 3–10) areas. Hospital admissions for rheumatic heart disease were significantly higher for females, Pacific > Māori > European > Asian/Indian children and young people and those from average-to-more deprived (NZDep decile 3–10) areas (Table 89). Similar ethnic differences were seen during 2000–2010 (Figure 90).

New Zealand Distribution by Season
In New Zealand during 2006–2010, hospital admissions for acute rheumatic fever and rheumatic heart disease were generally higher during the cooler months (Figure 91).

South Island Distribution and Trends
South Island DHBs vs. New Zealand
In Canterbury and Otago during 2006–2010, hospital admissions for children and young people with acute rheumatic fever and rheumatic heart disease were significantly lower than the New Zealand rate, while in the West Coast no admissions for either outcome occurred during this period, and in South Canterbury small numbers precluded a valid analysis. Rheumatic heart disease admissions in Nelson Marlborough and Southland were also significantly lower than the New Zealand rate, although small numbers precluded a valid analysis for acute rheumatic fever (Table 90).

Table 90. Acute and Semi-Acute Hospital Admissions for Acute Rheumatic Fever and Rheumatic Heart Disease in Children and Young People Aged 0–24 Years, South Island DHBs vs. New Zealand 2006–2010

<table>
<thead>
<tr>
<th>DHB</th>
<th>Number: Total 2006–2010</th>
<th>Number: Annual Average</th>
<th>Rate per 100,000</th>
<th>Rate Ratio</th>
<th>95% CI</th>
</tr>
</thead>
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<tr>
<td></td>
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<td></td>
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<tr>
<td><strong>Acute Rheumatic Fever 0–24 Years</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nelson Marlborough</td>
<td>&lt;3</td>
<td>s s</td>
<td>s</td>
<td>s</td>
<td>s</td>
</tr>
<tr>
<td>West Coast</td>
<td>0</td>
<td>0.0</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Canterbury</td>
<td>18</td>
<td>3.6</td>
<td>2.13</td>
<td>0.15</td>
<td>0.09–0.24</td>
</tr>
<tr>
<td>South Canterbury</td>
<td>&lt;3</td>
<td>s s</td>
<td>s</td>
<td>s</td>
<td>s</td>
</tr>
<tr>
<td>Otago</td>
<td>6</td>
<td>1.2</td>
<td>1.82</td>
<td>0.13</td>
<td>0.06–0.28</td>
</tr>
<tr>
<td>Southland</td>
<td>&lt;3</td>
<td>s s</td>
<td>s</td>
<td>s</td>
<td>s</td>
</tr>
<tr>
<td>New Zealand</td>
<td>1,096</td>
<td>219.2</td>
<td>14.4</td>
<td>1.00</td>
<td></td>
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<tr>
<td><strong>Rheumatic Heart Disease 0–24 Years</strong></td>
<td></td>
<td></td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Nelson Marlborough</td>
<td>9</td>
<td>1.8</td>
<td>4.33</td>
<td>0.44</td>
<td>0.23–0.85</td>
</tr>
<tr>
<td>West Coast</td>
<td>0</td>
<td>0.0</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Canterbury</td>
<td>16</td>
<td>3.2</td>
<td>1.89</td>
<td>0.19</td>
<td>0.12–0.32</td>
</tr>
<tr>
<td>South Canterbury</td>
<td>&lt;3</td>
<td>s s</td>
<td>s</td>
<td>s</td>
<td>s</td>
</tr>
<tr>
<td>Otago</td>
<td>5</td>
<td>1.0</td>
<td>1.51</td>
<td>0.15</td>
<td>0.06–0.37</td>
</tr>
<tr>
<td>Southland</td>
<td>4</td>
<td>0.8</td>
<td>2.20</td>
<td>0.23</td>
<td>0.08–0.60</td>
</tr>
<tr>
<td>New Zealand</td>
<td>747</td>
<td>149.4</td>
<td>9.79</td>
<td>1.00</td>
<td></td>
</tr>
</tbody>
</table>

Source: Numerator: National Minimum Dataset (Acute and semi-acute admissions with Acute Rheumatic Fever or Rheumatic Heart Disease listed in any of the first 15 diagnoses); Denominator: Statistics NZ Estimated Resident Population. Note: s: suppressed due to small numbers.

South Island Trends
In the South Island during 2000–2010, small numbers made trends in hospital admissions for children and young people with acute rheumatic fever and rheumatic heart disease difficult to interpret (Figure 92).
Figure 92. Acute and Semi-Acute Hospital Admissions for Acute Rheumatic Fever and Rheumatic Heart Disease in Children and Young People Aged 0–24 Years, South Island DHBs vs. New Zealand 2000–2010

Source: Numerator: National Minimum Dataset (Acute and semi-acute admissions with Acute Rheumatic Fever or Rheumatic Heart Disease listed in any of the first 15 diagnoses); Denominator: Statistics NZ Estimated Resident Population
Summary

In New Zealand, hospital admissions for children and young people with acute rheumatic fever declined gradually during the early-mid 2000s, but increased again after 2006–07. In contrast, admissions for those with rheumatic heart disease were relatively static during the mid-2000s, although a downswing in rates was evident in 2010. During 2006–2010, acute rheumatic fever and heart disease admissions were both relatively infrequent during infancy, but increased rapidly during childhood, to reach a peak at 11-12 years of age. Acute rheumatic fever admissions were significantly higher for males, Pacific > Māori > European and Asian/Indian children and young people and those from average-to-more deprived (NZDep decile 3–10) areas, while rheumatic heart disease admissions were significantly higher for females, Pacific > Māori > European > Asian/Indian children and young people and those from average-to-more deprived (NZDep decile 3–10) areas.

In Canterbury and Otago during 2006–2010, hospital admissions for children and young people with acute rheumatic fever and rheumatic heart disease were significantly lower than the New Zealand rate, while in the West Coast no admissions for either outcome occurred during this period, and in South Canterbury small numbers precluded a valid analysis. Rheumatic heart disease admissions in Nelson Marlborough and Southland were also significantly lower than the New Zealand rate, although small numbers precluded a valid analysis for acute rheumatic fever.

Local Guidelines and Evidence-Based Reviews Relevant to the Prevention and Management of Rheumatic Fever

The primary prevention of rheumatic fever focuses on the adequate treatment of streptococcal throat infections, while secondary prevention aims to ensure that all children and young people previously diagnosed with rheumatic fever receive monthly antibiotic prophylaxis. In New Zealand, while there are no Government policy documents which focus solely on rheumatic fever, the National Heart Foundation has developed a set of guidelines for the primary and secondary prevention of rheumatic fever. These are reviewed in Table 91, along with a range of other reviews and guidelines which consider these issues in the overseas context.

In addition, many of the measures previously reviewed in the context of respiratory and infectious diseases are likely to have a significant impact on rheumatic fever rates. These include:

1. **Generic Approaches to Infectious and Respiratory Disease:** Table 46 on Page 166
2. **The Prevention of Second Hand Smoke Exposure:** Table 47 on Page 168
3. **Interventions Aimed at Housing and Household Crowding:** Table 48 on Page 170
4. **Interventions to Improve Breastfeeding:** Table 27 on Page 107
Table 91. Local Guidelines and Evidence-Based Reviews Relevant to the Prevention and Management of Acute Rheumatic Fever and Rheumatic Heart Disease

<table>
<thead>
<tr>
<th>New Zealand Guidelines</th>
</tr>
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These guidelines provide evidence-based guidance on the best practice for the diagnosis and management of acute rheumatic fever, for secondary prophylaxis (prevention of repeat attacks), and also for the standard of care that should be available to all people in New Zealand including those who are members of high-risk populations. Evidence is graded according to the system used in the National Heart Foundation of Australia Rheumatic Fever Guidelines (level 1 evidence is that obtained from a systematic review of a number of RCTs) and recommendations are graded based on the quality of the evidence on which they are based. A summary of the N.Z. guidelines has been published as:


The purpose of these guidelines is to provide evidence-based guidance for the diagnosis and treatment of Group A streptococcal sore throats in people aged 3 to 45 years to help ensure that those at highest risk of developing rheumatic fever receive the correct diagnosis and treatment while at the same time minimising unnecessary investigations and antibiotic use in those who are at the lowest risk. The levels of evidence and grades of recommendations used are adapted from the National Health and Medical Research Council levels of evidence and the U.S. National Institute of Health clinical guidelines. A summary of the N.Z. guidelines has been published as:


These guidelines state that treating Group A Streptococcal sore throats reduces the rate of subsequent rheumatic fever and that school and community based programmes for detection and treatment are effective. There is a role for Māori and Pacific health care providers, school-based sore throat programmes and for primary health care reforms to improve access to health care for the people at highest risk of developing rheumatic fever. Household crowding is associated with an increased risk of developing rheumatic fever and some, but not all studies show a link with poverty. There is no convincing evidence of a genetic susceptibility to rheumatic fever or an association with skin infections (which can also be caused by Group A streptococcal infection). These guidelines use the same evidence grading system as the earlier National Heart Foundation guidelines.


The introduction to these guidelines states that “There has never been a prospective clinical placebo-controlled trial of antibacterial prophylaxis in individuals with cardiac risk undergoing a potentially bacteraemia-producing procedure.” It is noted the 2007 American Heart Association recommendations advise prophylaxis only for those having dental procedures while the U.K. National Institute for Clinical Excellence recommendations advise no prophylaxis for anyone, for any procedure. The New Zealand guidelines recommend prophylaxis for people with rheumatic valvular heart disease and emphasise the importance of good oral health for all people at risk of endocarditis.

**Systematic and Other Reviews from the International Literature**


This comprehensive review of the literature presents recommendations for prevention under the headings of Socio-economic factors, Biological factors, Lifestyle factors and Healthcare systems and services. The authors were members of the writing group responsible for the New Zealand guidelines and much of the material in this review is also in the third of the guidelines.
Antibiotics are of limited benefit in treating sore throat unless patients have positive throat swabs for group A beta-haemolytic streptococci (GABHS). Seventeen RCTs (5352 participants) were included in this review. Sixteen compared penicillin with another antibiotic and one compared clindamycin with ampicillin. All of the trials were conducted in high income countries where the risk of rheumatic fever is low and they do not provide information on the effectiveness of different antibiotics for the prevention of complications (rheumatic fever and post-streptococcal glomerulonephritis). The authors found that there was insufficient evidence for clinically meaningful differences between antibiotics used to treat GABHS. They conclude that considering these results together with the low cost of penicillin and the lack of penicillin resistance by GABHS, penicillin can still be recommended as a first choice antibiotic. They state that there is a need for studies in specific communities at high risk for complications.


This study assessed prevention of rheumatic fever through treatment of streptococcal pharyngitis in school- and/or community-based programmes by doing a meta-analysis of relevant RCTs or before-and-after studies. Data from 6 studies which met the inclusion criteria were pooled in a meta-analysis to give a relative risk of 0.41 (95% CI 0.23 – 0.70) for the interventions. The authors state that this result indicates that a school and/or community based programmes could be expected to decrease the number of cases of acute rheumatic fever by about 60%.


This review considered evidence for the efficacy of treatment of acute group A beta hemolytic streptococcus (GABHS) pharyngitis with two to six days of newer oral antibiotics compared to the standard treatment of 10 days of oral penicillin. The review included 20 RCTs with a total of 13102 cases. The authors concluded that three to six days of oral antibiotics had comparable efficacy to 10 days of penicillin however they noted that the primary reason for 10 days of penicillin is for prevention of rheumatic fever and they state “in areas where the prevalence of rheumatic fever is still high our results must be interpreted with caution.”


In this commentary the authors outline their concerns about the above review particularly the criteria chosen to indicate successful treatment and the heterogeneity of the trials included. They state that group A Streptococcus is the only common cause of acute pharyngitis requiring antibiotic treatment, it is highly sensitive to penicillin and that 10 days of treatment remains the gold standard.


This review assessed the benefits of antibiotics for sore throat. Sixteen of the 27 trials included (10101 participants) assessed the benefits of antibiotics in reducing the incidence of rheumatic fever within two months. A meta-analysis of these 16 studies gave a rheumatic fever risk ratio of 0.27 (95% CI 0.12 -0.60) for antibiotics vs. placebo. Meta-analyses looking at penicillin and pre-1975 studies separately gave similar results to the meta-analysis of all antibiotic studies together. (There were no cases of rheumatic fever in the post 1975 studies.) There was no distinction made in this review between adults and children. The authors concluded that antibiotic use may be justified in areas where rheumatic fever is common but that in other places practitioners need to weigh the modest symptom reductions against the hazards of antibiotic treatment.


This review included 10 random or quasi-randomised trials with a total of 7665 participants, generally considered to be of poor methodological quality. All were conducted between 1950 and 1961 and 8 of them involved young men in U.S. military bases. The results of the meta-analysis showed an overall protective effect for antibiotics in the prevention of rheumatic fever following sore throat (with or without confirmation of group A streptococcal infection) (RR 0.32, 95% CI 0.21 – 0.48). When only the 9 trials including intramuscular penicillin were included in the meta-analysis the protective effect was greater (an 80% reduction, RR 0.20, 95% CI 0.11 – 0.36). The authors state that their findings support the view that treating cases of suspected streptococcal pharyngitis with antibiotics is an effective and safe way to prevent rheumatic fever in children in poor socioeconomic conditions where rheumatic fever is common.


Recurrent (secondary) episodes of rheumatic fever are associated with a high risk of developing chronic rheumatic heart disease and also of worsening already existing rheumatic heart disease. Continuing treatment with penicillin can prevent recurrent attacks. This review included 9 studies (3008 participants in total), which the authors considered to be of generally poor quality, investigating various preventive penicillin regimens and formulations. Based on the findings from four trials (1098 participants) the authors concluded that intramuscular penicillin seemed to be more effective than oral penicillin in preventing rheumatic fever recurrence and streptococcal throat infections. There was limited evidence that two-weekly or three-weekly injections were more effective than four weekly injections (one trial for each).
Relevant New Zealand Publications and Websites


The section of this conference presentation entitled: Rheumatic Fever and Rheumatic Heart Disease in New Zealand (Nigel Wilson) provides a useful outline of the topic and some of the New Zealand studies and initiatives. It explains that it is very important to ensure that national campaigns to educate the whole population that most sore throats do not require antibiotics and thus limit unnecessary prescribing (because most sore throats are due to viral infections) do not undermine efforts to encourage identification and treatment of streptococcal sore throats which can lead to rheumatic fever in vulnerable Māori and Pacific people.

The following publications and websites provide information on a variety of New Zealand studies and initiatives:


