

## The epidemiology of acute rheumatic fever in Northland, 2002–2011

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### Abstract

**Aim** An audit of rheumatic fever surveillance in Northland was carried out for the period 2002–2011. The aim of the audit was to establish the accuracy and completeness of surveillance of Acute Rheumatic Fever in Northland, and to provide a robust baseline for future comparison given current rheumatic fever prevention efforts.

**Methods** Cases of acute rheumatic fever (2002–2011) were identified and evaluated through auditing Northland hospital discharges, the Northland Rheumatic Fever secondary penicillin prophylaxis register and the national EpiSurv database. Cases were included in the audit if they met diagnostic criteria according to the 2008 Heart Foundation guidelines.

**Results** A total of 114 acute rheumatic fever cases met the audit criteria, an annualised incidence of 7.7/100,000 in Northland. 95% of all cases were Māori with a large disparity between Māori (24.8/100,000) and non-Māori (0.6/100,000). Acute rheumatic fever cases were strongly associated with living in high deprivation areas. This audit noted both under- and over-notification of acute rheumatic fever.

**Conclusion** Acute rheumatic fever rates in Northland Māori children aged 5–14 (78/100,000) are similar to those seen in developing countries and nearly double the rates seen other New Zealand audits. The findings highlight the urgent need to address crowding, poverty and inequitable primary care access if rheumatic fever is to be eliminated.

Acute rheumatic fever (ARF) is a preventable disease associated with poverty, poor access to health care and crowding, and is now rare in most developed countries. It results from an abnormal autoimmune response to Group A streptococcal (GAS) pharyngitis in a susceptible individual.<sup>1</sup> Repeated episodes of ARF can result in structural damage to the heart valves, or rheumatic heart disease (RHD).

This is an important cause of premature death and significant morbidity worldwide, and in Maori and Pacific communities in New Zealand.<sup>2</sup> Rates of ARF in Northland have been historically amongst the highest in New Zealand, and disproportionately impact on Maori children.

The primary aim of this audit was to establish the accuracy and completeness of surveillance for ARF in Northland for the ten-year period 2002–2011 as a robust baseline for future comparison, given current prevention efforts. In addition, we aimed to identify patients with ARF who were not receiving best practice management as per the 2008 Heart Foundation guidelines (i.e. secondary penicillin prophylaxis and specialist follow up).<sup>3</sup>

The population of Northland is estimated at 148,470, with 29% identifying as Maori.<sup>4</sup> The Maori population is significantly younger than non-Maori (36% are aged less than 15 years, compared to 23% of non-Maori) and there are high levels of socio-economic deprivation, unemployment and one-parent families in Northland, compared with the New Zealand population.<sup>4</sup>

## Methods

ARF cases (2002-2011) were identified and evaluated through auditing Northland hospital discharges, the RF register and the national surveillance EpiSurv database. All hospital discharges with Rheumatic Heart Disease (RHD) or ARF aged less than 35 years who were diagnosed during 2002-2011 were identified using the ICD-9 and ICD-10 coding systems (ICD codes 100, 101.0, 101.2, 101.8, 101.9, 102.0, 102.9).<sup>5</sup> All patients on the RF register currently or ever receiving benzathine penicillin prophylaxis for ARF and RHD during the period 2002-2011 were reviewed, along with all Northland cases notified to EpiSurv from 2002-2011.

Cases were included in the audit if they were diagnosed with ARF and met criteria according to the 2008 Heart Foundation guidelines<sup>3</sup> during the period 2002-2011, were resident in Northland and aged less than 35 years at the time of diagnosis.

All clinical notes were sought and reviewed of cases with an appropriate primary or secondary diagnostic code of ARF and RHD. All EpiSurv and RF register notes were reviewed. Information was extracted from case notes and entered into a standard data format. The modified Jones criteria (inclusive of echocardiographic detection of carditis in the absence of a clinical murmur as a major criteria) was used to determine ARF diagnosis of “definite”, “probable” or “possible”.<sup>3</sup>

All “possible” cases were additionally reviewed by a paediatrician to ensure adequate diagnosis. Each case was geocoded according to their place of residence at the time of diagnosis and deprivation status assigned using NZDep2006.<sup>6</sup> Population statistics were obtained from Statistics New Zealand 2006 Census of population.<sup>4</sup> Analysis was carried out by age and ethnicity.

## Results

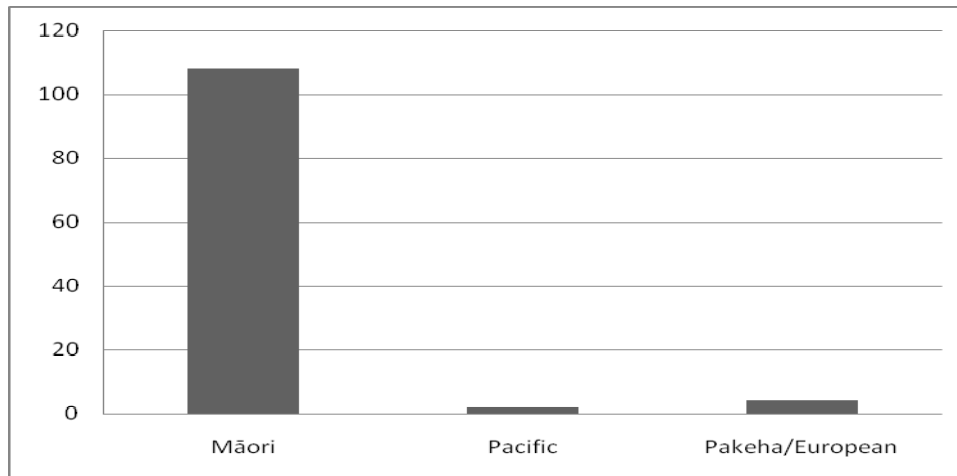
117 rheumatic fever cases (including six notified recurrences) were identified from EpiSurv. 13 cases were discarded on review, as they did not meet diagnostic criteria for acute rheumatic fever.

Of these, five were recurrences that had insufficient data and did not meet criteria for recurrence, and eight other ARF cases were excluded: five were not ARF and three were diagnosed outside of the area. One notified recurrence was actually a case of initial ARF incorrectly entered as a recurrence in EpiSurv.

157 RHD/ARF cases were identified from hospital discharge ICD coding to ensure RHD cases were not incorrectly coded as ARF and vice versa. Most were excluded as they were RHD rather than ARF, or were outside the audit criteria (for the period or age range). Ten cases met the criteria for ARF that were not duplicates with those on EpiSurv.

The RF register was compared with EpiSurv and ten cases, all duplicates with those identified from ICD coding above, were identified that were not on EpiSurv. Therefore a total of 114 ARF cases (81 “definite”, 18 “probable” and 15 “possible”) met the audit criteria, an annualised incidence rate of 7.7/100,000 (~12 cases per year). The mean annual number of cases from 2002-2006 was 9.2 (range 7-12), while in the period 2007-2011 it increased to 13.6 per year (range 7-18).

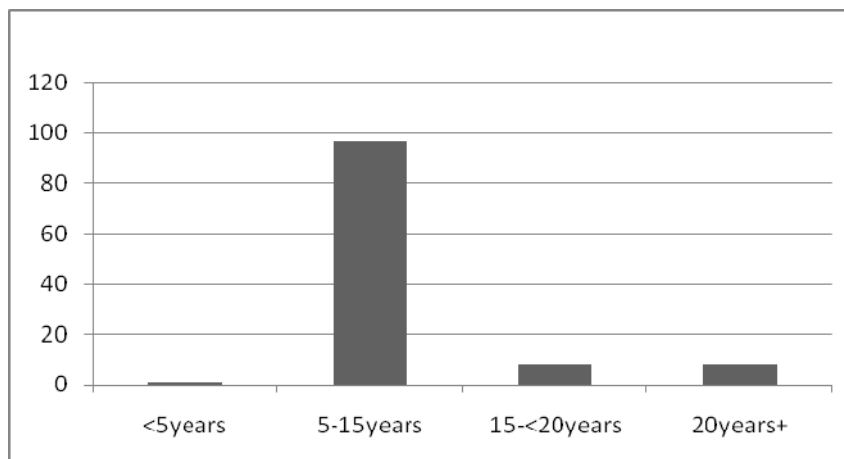
**Figure 1. Ethnicity of ARF cases in Northland 2002-2011**



95% of all ARF cases were Māori (n=108), with a large disparity between rates in Māori (24.8/100,000) and non-Māori (0.6/100,000). There were two cases in Pacific children. In the 5-14 age group where the highest rates and largest disparity were found, 94% were Māori (a rate of 78.0/100,000 compared with 4.6/100,000 for non-Māori).

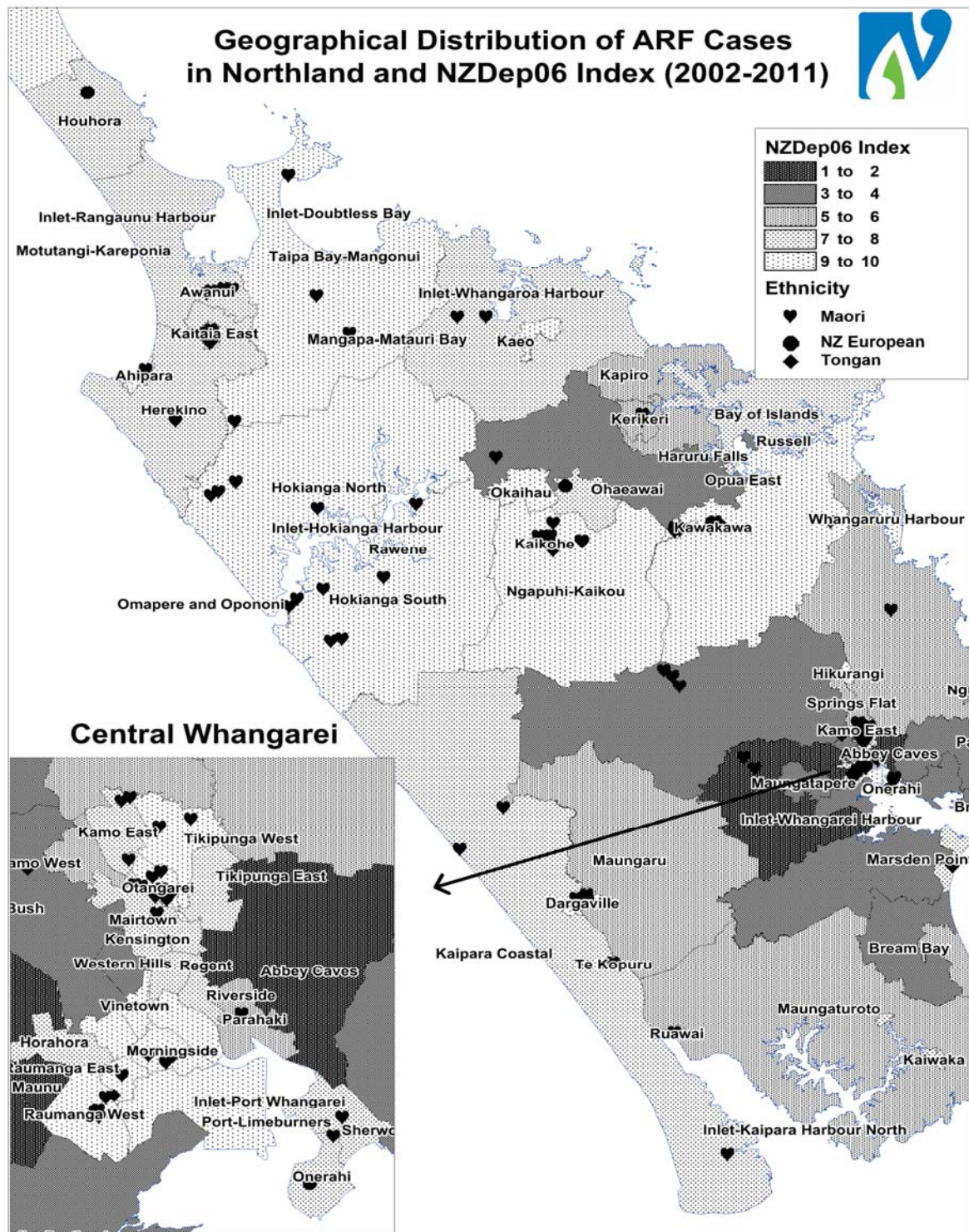
60% (n=68) of cases were male and 40% (n=46) female, with ages ranging from 4-26 years; 85% (n=97) were aged 5-<15years. The mean age was 11.4 years.

**Figure 2. Age of ARF cases in Northland 2002-2011**



ARF cases were strongly associated with living in high deprivation areas and distributed across Northland (Figure 3). Over half (55%, 63 cases) resided in the most deprived decile (NZDep10) and 89.5% (102 cases) resided in NZDep deciles 8-10.

Figure 3. Geographical & NZ Dep distribution of ARF cases in Northland 2002–2011





The majority of cases had a definite diagnosis (n=81, 71%) and were low risk (n=92, 81%). At diagnosis 97 (85%) had carditis, 48 (42%) polyarthritis, eight (7%) had chorea and ten (9%) had erythema marginatum.

There were no cases of nodules recorded, but 48% had no data on the presence or absence of nodules. Of the ten cases of erythema marginatum, only seven were definite, with one being noted on history and two recorded as “possible”. Three of the eight children presenting with chorea were NZ European (there were only four NZ European ARF cases in the period). The commonest presentation occurring simultaneously was carditis and arthritis (51 cases, 45%).

53 (46%) of the ARF cases gave a history of a preceding sore throat; 26 (23%) had a GAS positive throat swab, with only 16 (14%) having both a sore throat and GAS positive throat swab. There were no data on pharyngitis symptoms for 20 cases, and 21 (18%) had no data on GAS.

46 (40%) had both raised plasma antistreptolysin O titres (ASOT  $\geq$  480) and anti deoxyribonuclease B titres (antiDNase B  $\geq$  660). 20 (18%) had raised ASOT only while 18 (16%) had only raised anti DNase B titres. Of those with positive serological titres, 5 grew Group C and 1 Group G streptococci on throat swab. Of the remaining 30 (26%) with low titres or no titres documented, 9 (8%) had a GAS+ swab, 3 (3%) had chorea, 3 (3%) had rising or falling titres documented and in the remainder (15, 13%), ARF was considered the most likely clinical diagnosis.<sup>3</sup>

Inflammatory markers (ESR and CRP) were both raised in 72 (63%) of ARF cases. 76 (67%) had a CRP  $>30$  and 96 (84%) an ESR  $>50$ . In all cases where ESR levels did not meet criteria, none had a raised CRP; however for those that did not have a raised CRP, 14/22 (64%) had a raised ESR. Only two ARF cases had no data for both markers.

All ARF cases were risk allocated based on the severity of carditis as determined by the Heart Foundation guidelines.<sup>3</sup> 92 (81%) were classified as low risk and 22 (19%) medium to high risk. In terms of post-diagnosis follow up, seven of the medium to high risk cases were not receiving best practice care as per HF guidelines, that is, being followed up by a cardiologist.<sup>3</sup>

Of these seven, two have moved out of the region and care has been officially transferred, one did not receive cardiology follow up due to transport issues and four had no cardiology follow up organised. In addition, only one case was documented to have had a dental review in the preceding six months.

Ten ARF cases were not notified and were identified via ICD coding and the RF register (five definite, 3 probable and two possible cases); all have received/are receiving secondary penicillin prophylaxis. Four cases were identified as lost to follow up, and twelve (11%) ARF cases transferred out of the region during 2002-2011. Eight of these cases are now residing in Auckland.

## Discussion

ARF rates in Northland Māori aged 5-14 (78.0/100000) are similar to those seen in developing countries and nearly double the rates seen in Auckland Māori children during 1993-1999 (41.2/100000) and in the Waikato, 1998-2004 (39.6/100000).<sup>7,8</sup>

Tairāwhiti, a region with a similarly high proportion of Māori to Northland, has also documented high rates (total population 7.6/100,000 and 5-14 years 59/100,000 for Māori children 5-14 years).<sup>9</sup> Of additional concern, our audit indicates that cases in Northland show an increasing trend over the last 5 years.

As a result of improvements in socioeconomic conditions and primary health care access, ARF and RHD have almost been eradicated from developed nations around the world. High rates of ARF have been shown to be associated with socioeconomic deprivation.<sup>10</sup> In Northland, the pattern of ARF cases correlates closely with socioeconomic deprivation, with 55% of ARF cases in Northland living in the most deprived decile (NZDep10).

This audit noted both under- and over-notification of ARF. There were also gaps in the clinical reporting and follow up of ARF. This contributed to most recurrences being excluded, and a lack of data in some key clinical areas, as well as gaps in data on school attended at diagnosis. Lack of systematic use of diagnostic criteria as per the Heart Foundation Guidelines was also noticed.<sup>3</sup>

Differentiating between major and minor manifestations in the diagnostic assessment was not always well done. Repeat streptococcal titres at 10-14 days were often not performed when initial titres were low. This is important as it is estimated that 50% of the population are colonised with GAS and rising or elevated streptococcal titres are important to diagnose “definite” ARF.<sup>3</sup> In addition, the clinical diagnosis of polyarthralgia was commonly hard to distinguish from polyarthritis in clinical notes. The inability to weight-bear should prompt a diagnosis of arthritis.<sup>3</sup>

Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) are sensitive but non-specific tests that can be elevated in any inflammatory condition.<sup>11</sup> The use of CRP in the diagnosis of ARF was noted as early as 1958.<sup>12</sup>

The audit suggests that ESR is more reliable as a positive minor manifestation in the diagnosis of ARF. In cases where the CRP was not raised, 64% had an ESR >50, while in cases where the ESR was not raised there was no associated rise in CRP.

In the past ESR has been commonly used to monitor ARF. The current guidelines suggest all patients should also have a CRP checked.<sup>3</sup> As CRP rises and falls faster than ESR this may be useful in uncomplicated cases of carditis to confirm the resolution of inflammation in those who have a prolonged elevated ESR.<sup>11</sup> CRP could aid in determining the duration of bed rest in low risk patients.

The majority of ARF cases in this audit were low risk suggesting of ARF is being promptly detected in our setting. However the nearly 20% who were medium/high risk require regular cardiology follow up and this was not always assured. Referral for regular dental review was also poorly documented. Those patients who were not benefiting from best practice have been identified and will be followed up by the Northland Public Health rheumatic fever team.

ARF is preventable. The audit findings highlight the urgent need to address crowding, poverty and inequitable primary care access if rheumatic fever is to be eliminated. Current school-based “sore throat” projects are important for primary prevention of ARF, by enhancing access to timely diagnosis and management of GAS pharyngitis.

However, to reach the national goal of reducing ARF by two-thirds in 5 years, greater efforts -in Northland and nationally - will be required.<sup>13</sup>

This will include improving housing quality and reducing crowding, addressing inequities in household incomes, reducing disparities in access to primary care and increasing awareness of the disease in those communities most affected.

**Competing interests:** Nil.

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**Acknowledgements:** Thanks to Drs Anil Shetty and Maya Williams for their support in data collation and analysis.

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