The epidemiology of acute rheumatic fever in Northland, 2012–2017

Kate Wauchop, Anil Shetty, Catherine Bremner

ABSTRACT

AIM: The aim of the audit was to establish the accuracy and completeness of surveillance of acute rheumatic fever (ARF) in Northland for the period 2012–2017 and to compare the rates of ARF with the previous audit.

METHODS: Cases of ARF were identified and evaluated through the national surveillance database (EpiSurv), the Northland Rheumatic Fever secondary prophylaxis register and hospital discharges. Cases were included if they were Northland residents and met the ARF diagnostic criteria as per the 2014 New Zealand Heart Foundation guidelines.

RESULTS: A total of 69 ARF cases met audit criteria, an annualised incidence rate of 7.0/100,000. Ninety-three percent of cases were Māori. The 5–14 year age group had the highest rates, which also demonstrated the greatest disparity between Māori (64.5/100,000), Pacific (54.6/100,000) and non-Māori/non-Pacific (1.5/100,000). ARF was strongly associated with level of deprivation, with 87% of cases in NZDep8–10.

CONCLUSION: ARF surveillance in Northland appears comprehensive with only minor discrepancies identified between data sets. ARF rates among Northland Māori aged 5–14 years (64.5/100,000) remain similar to those seen in low-income countries. For Māori, an improvement from the previous audit of only one-fifth was seen despite the Rheumatic Fever Prevention Programme. The Ministry of Health's recent publications indicate a large improvement in ARF rates for Northland. However, these findings do not correlate with the results of this audit. This audit highlights the significant, ongoing need in Northland for ARF prevention efforts.

Acute rheumatic fever (ARF) is associated with poverty, poor access to healthcare and household crowding, now rarely seen in high-income countries.[1] ARF develops due to an abnormal autoimmune response to group A streptococcal (GAS) throat infection, commonly occurring two to three weeks after GAS pharyngitis.[2] Carditis (clinical and subclinical) occurs in approximately 85% of those with ARF, typically resulting in regurgitation of mitral and aortic valves.[3] Single or repeated episodes of ARF can result in permanent heart valve damage or rheumatic heart disease (RHD), a significant cause of morbidity and mortality throughout the world[1] and in the Māori and Pacific populations of New Zealand.[4]

Northland has an estimated population of 164,730, with just over 33% identifying as Māori.[5] Northland Māori are younger than the total Northland population (median age of 25.8 years, compared to 42.7 years) and have much higher levels of deprivation and unemployment.[5]

In 2012, a 10-year (2002–2011) retrospective audit of ARF cases demonstrated an overall annual incidence rate of 7.7/100,000 people across Northland.[6] Ninety-five percent of the cases were among Māori with a large disparity between Māori (24.8/100,000) and non-Māori (0.6/100,000). This disparity was most evident in those aged 5-14 years at 78/100,000 for Māori and 4.6/100,000 for non-Māori.[6]

The Rheumatic Fever Prevention Programme (RFPP) was established by the Ministry of Health (MoH) in 2011 to reduce the incidence of ARF cases in high-risk regions by preventing and treating GAS pharyngitis.[7] The government invested ~$65 million to support various new initiatives

in high-risk regions, following the introduction of the five-year rheumatic fever Better Public Service (BPS) national target of reducing ARF hospitalisation rates from 4/100,000 in 2010/11 to <1.4/100,000 in 2016/17.[7,8] In line with the national target, Northland District Health Board (NDHB) aimed to reduce its rate from 10.5/100,000 to 3.5/100,000 in 2016/2017. However, it also had set its own goal to eliminate ARF by 2020, ie, have rates below 2011 New Zealand Pakeha (European) levels (<0.6/100,000) through its 2011 RF Prevention Plan.[9]

The main aim of the audit was to verify the accuracy and completeness of ARF surveillance in Northland for the period 2012–2017 and to compare the rates of ARF with the previous audit following the implementation of the RFPP. Additionally, it aimed to assess best practice management of ARF cases as per the 2014 Heart Foundation Guidelines (ie, secondary penicillin prophylaxis and specialist follow up).[10]

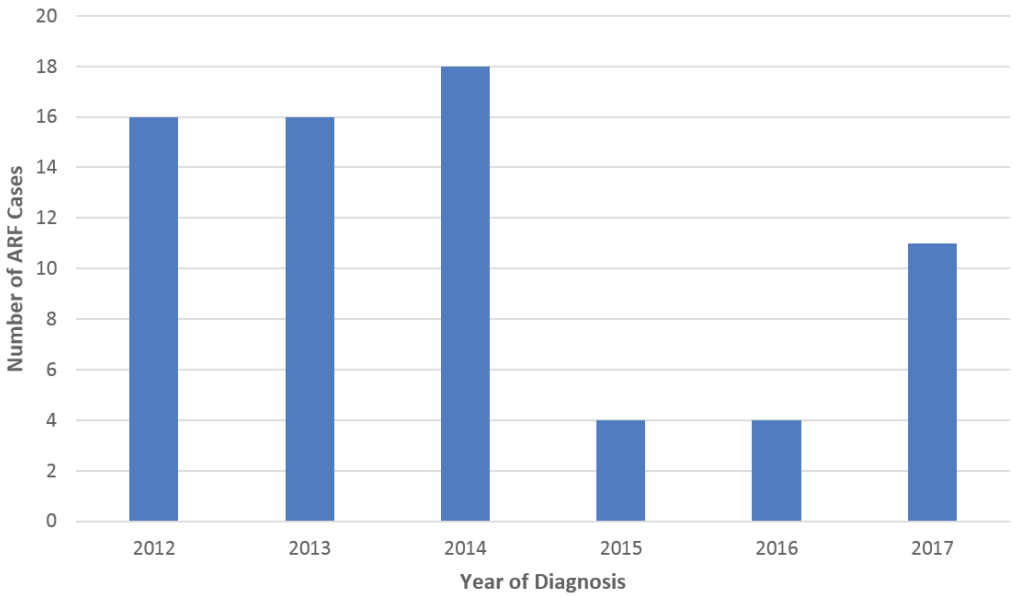
Methods

Similar to the previous audit, ARF cases (2012–2017) were identified and assessed through reviewing Northland hospitals discharges (discharge data), the benzathine penicillin prophylaxis register (RF register) and the national-notifiable disease surveillance database (EpiSurv).[6] All hospital discharges with ARF or RHD aged less than 40 years were identified using the ICD-10 codes 100, 101.0, 101.2, 101.8, 101.9, 102.0, 102.9.[11] All patients on the RF register were reviewed, along with all Northland cases notified to EpiSurv. Cases were included in the audit if they met criteria for ARF as per the 2014 Heart Foundation guidelines during the audit period and were resident in Northland.[10] The 2014 updated guidelines (as compared with the 2008 guidelines used in the previous audit) result in minimal change in diagnosis of ARF, though do include monoarthritis with-or-without the use of non-steroidal anti-inflammatories as a major criterion. In addition to clinical notes for the hospital discharges, notes from RF register and EpiSurv were also reviewed for the identified cases. Information from the notes was entered on an audit tool (MS Excel). The modified Jones criteria (which utilises echocardiography to detect subclinical carditis as a major criteria) was used to establish ARF diagnosis of 'definite', 'probable' or 'possible'.[10] The NZDep2013 deciles for the cases were assigned by their place of residence at the time of diagnosis.[12] Population numbers from Statistics New Zealand 2013 Census were utilised for rates calculations.[5] Analysis was carried out by age, prioritised ethnicity and NZDep2013 using MS Excel.

Results

Seventy cases were identified from EpiSurv with three discarded for not meeting audit criteria. One was an Auckland resident, the second diagnosed outside the audit time frame and the third was a new diagnosis of RHD without ARF during the audit time frame. Of interest, the diagnosis of RHD was in a 14-year-old with recurrent hospital admissions for severe respiratory illnesses. Of the 155 cases from the RF register, 87 were excluded for not meeting criteria (time frame/RHD). Two **further** cases of ARF were identified which were not on EpiSurv. Both were more complex diagnoses of acute on chronic RHD. The first met criteria for ARF (carditis, arthralgias, inflammatory markers and significant streptococcal serology) but also had signs of chronic RHD (tethering) on echo. This was reviewed with a paediatrician and a classification of initial ARF was made (likely acute on chronic RHD). The second case was atypical considering age and latency of recurrence. A 50-year-old who had ARF as a child presented with mono-arthritis, fevers, high inflammatory markers, significant streptococcal serology and carditis with severe valve disease. As an inpatient, two valves were replaced with histology demonstrating acute on chronic RHD. As such, the case was defined as a recurrence of ARF. One case from EpiSurv was not on the RF register, this was a case of recurrent ARF who had left Northland. Of the 91 cases identified from discharge data, 33 were excluded for not meeting audit criteria. The remaining 58 cases of ARF were duplicates of those from EpiSurv and the RF register. There were 11 cases of ARF that were not found in hospital discharge data. One case was older than 40 years. Seven cases were diagnosed in outpatient clinic. It is unclear why the remaining three cases (all hospital admissions) were not identified in discharge data (could be due to coding error).

Figure 1: ARF cases per year and prioritised ethnicity Northland 2012–2017.



Of the seven cases diagnosed in outpatient clinic; one case was 12 years old with the others older than 20. Two cases (12 and 29 years old) were admitted to hospital with joints pains and received a diagnosis of ARF after discharge in rheumatology clinic follow-up. Five patients were referred directly to outpatient clinic by either general practice (GP) or emergency department (ED). Two of these cases had 'medium-to-high-risk' carditis at the time of diagnosis in clinic.

Therefore, a total of 69 ARF cases (65 initial and four recurrences) met the audit criteria. 2012 and 2013 had 16 cases each, 2014 had 18 cases, 2015 and 2016 had only four each while 2017 had 11 cases (Figure 1). The four recurrences occurred in 2012, 2013, 2014 and 2016. Of the total 69 cases, 51 were definite, eight probable and 10 possible. The annualised incidence for the six-year period from 2012–2017 was 7.0/100,000 (approx. 12 cases per year). The data from 2017 gives a most recent annual incidence of 6.7 cases/100,000.

It is important to note that one case in 2017 had a prior diagnosis of ARF in 2010 and as such was included in the previous audit. After receiving more than five years penicillin prophylaxis the diagnosis was rescinded on follow up in 2016 and penicillin discontinued. The patient then had a definite ARF diagnosis in 2017. This was reviewed with a paediatrician and a decision was made to include the case in this audit as an 'initial' diagnosis.

Ninety-three percent of all cases were Māori (n=64). Four cases (6%) were Pacific Islander (two Tongan, one Cook Islander and one Niuean) and only one case (1%) was New Zealand European (Figure 2).

Figure 2: Ethnicity of ARF cases in Northland 2012–2017.

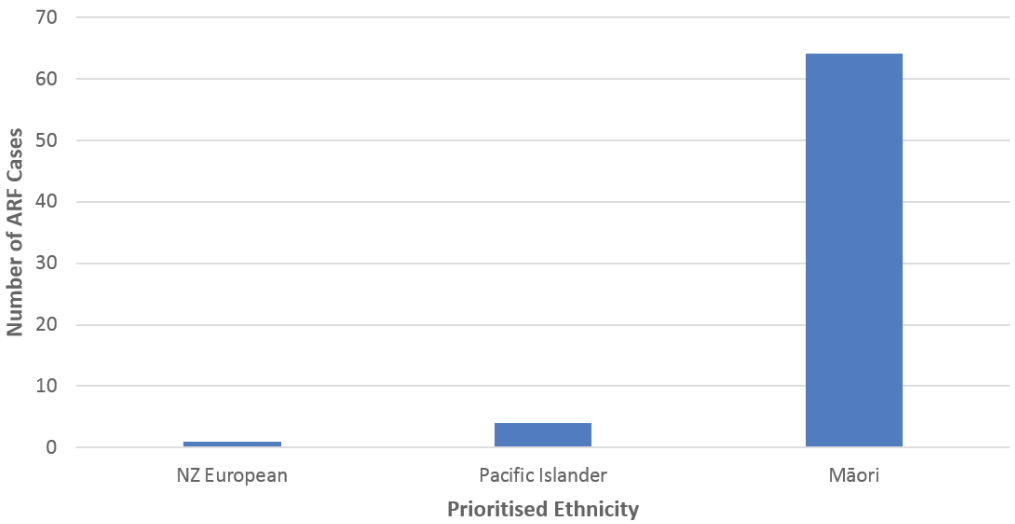


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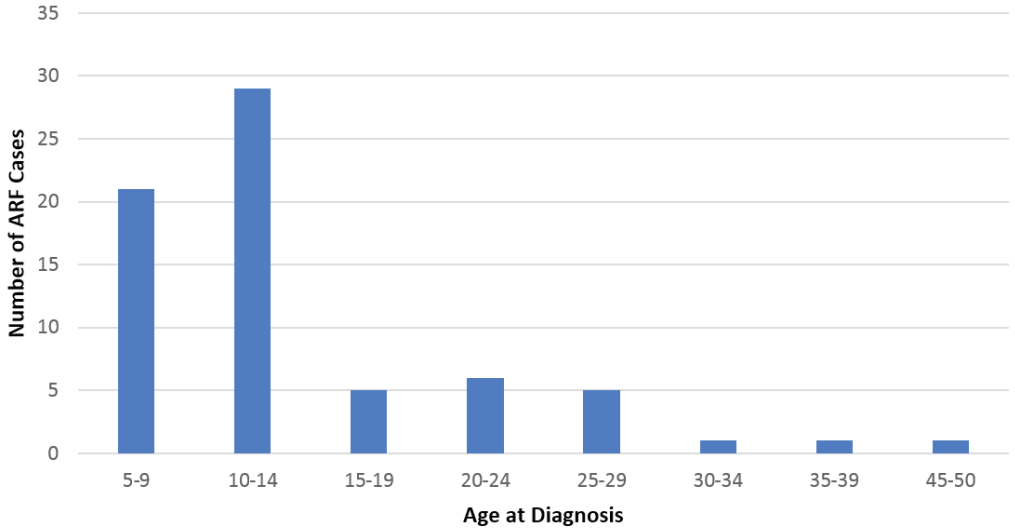
Figure 3: Age at diagnosis of ARF in Northland 2012–2017.



There was large disparity in rates between Māori (19/100,000), Pacific (22/100,000) and non-Māori/non-Pacific (0.2/100,000). All four recurrences were among Māori aged between 20 and 50 years.

The highest rates were identified in the 5–14 year age group, which also demonstrated the greatest disparity between rates in Māori (64.5/100,000), Pacific (54.6/100,000) and non-Māori/non-Pacific (1.5/100,000). Sixty-two percent (n=43) of cases were male and 38% (n=26) female. Ages ranged from 5–50 years; 72% (n=50) were aged 5–14 years (Figure 3). The mean age at ARF diagnosis was 14 years with a median age of 12 years. Ten new cases of ARF were diagnosed in patients aged 20 years or older. Of these, two demonstrated medium-high risk carditis at diagnosis. All four of the recurrences were 20 years or older and three of four patients had 'medium-to-high-risk' carditis at diagnosis.

Over half of the cases (55%, 38) lived in areas with NZDep2013 10 decile. Ninety percent of cases (62) lived in areas with NZDep2013 7–10 deciles (Table 1) (Figure 4).

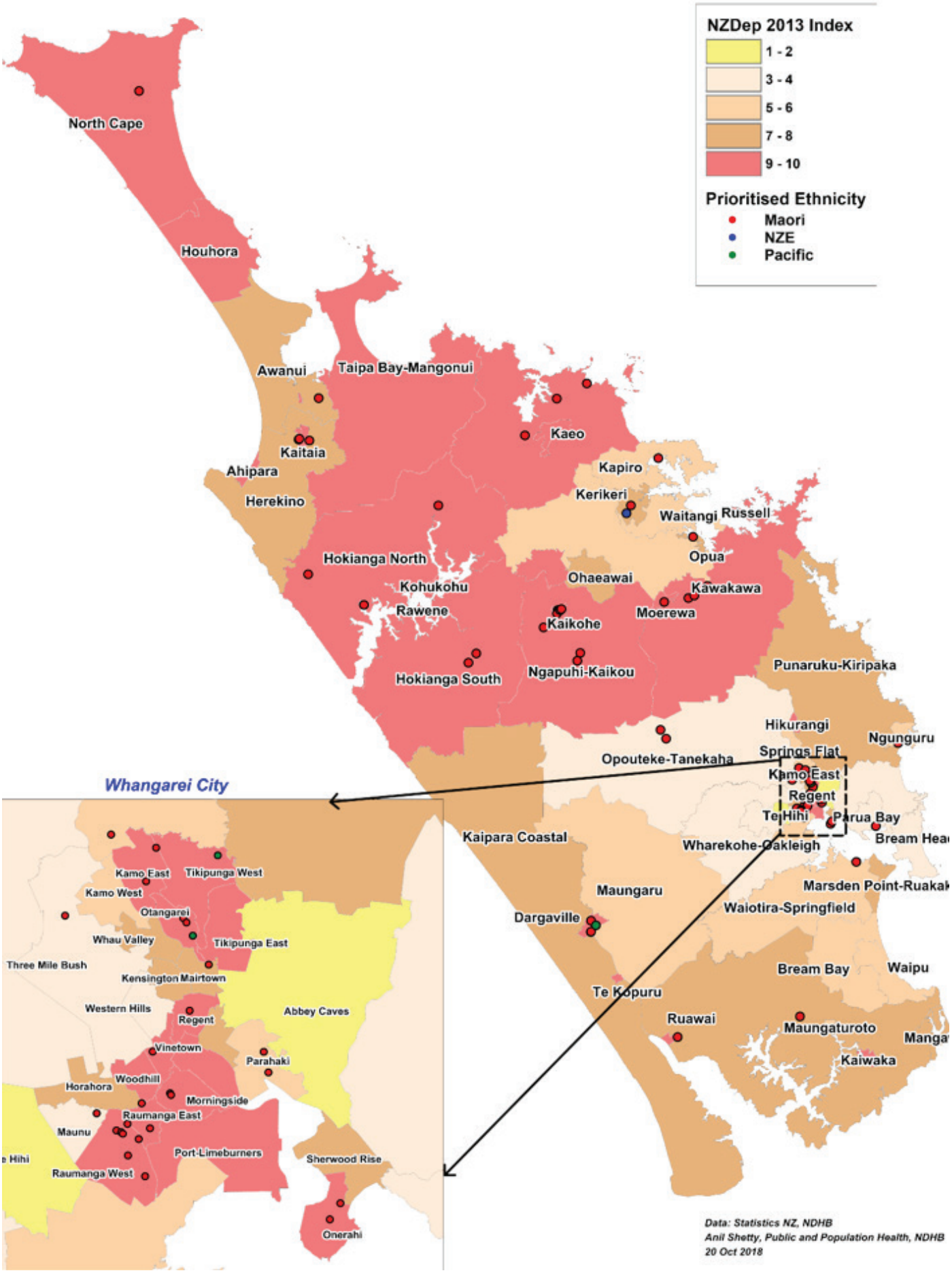
Most ARF cases were classified as 'definite' (n=51, 74%). Eighty percent (55) had carditis at diagnosis, 50% (35) polyarthritis, 13% (9) monoarthritis, 9% (6) chorea, 10% (7) erythema marginatum and 1% (1) nodules. Of the seven cases with erythema marginatum, two were recorded as 'possible erythema marginatum'. These cases were reviewed with a paediatrician and downgraded to probable and possible in accordance with other criteria met. The most common presentation occurring simultaneously was carditis and poly-arthritis (26 cases, 38%). The only New Zealand European case presented with chorea and carditis.

Of 69 audited cases, for 14 cases sore throat data was missing. Of the remaining 55 cases, 37 (67%) reported a recent sore

Table 1: Distribution of ARF cases in Northland by NZ Deprivation Index 2013 by year.

NZDep2013 Index	2012	2013	2014	2015	2016	2017	Total
1–2	1	0	0	0	0	0	1
3–4	1	0	1	0	0	0	2
5–6	2	1	0	0	1	0	4
7–8	2	0	2	1	0	3	8
9–10	10	15	15	3	3	8	54
Total	16	16	18	4	4	11	69

Figure 4: Geographical and NZ Dep distribution of ARF cases in Northland 2012–2017.



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throat. Throat swabbing data for six patients was missing. Fifty-nine percent (37/63) of the tested cases had GAS positive throat cultures. For 35 cases, data on both sore throats and throat swabs was available, and of these 23 (66%) had both a sore throat and GAS positive throat swab.

AntiDNAse B data was missing for one patient. Twenty cases (29%) had both raised anti deoxyribonuclease B (antiDNAse B >/= 660) and plasma antistreptolysin O titres (ASOT >/= 480). Twenty-six cases (38%) had only raised ASOT and 9 (13%) had only raised AntiDNAse B. Thirteen cases (19%) did not have either streptococcal (strep) serology significantly elevated. For two of these cases, strep serology rose and fell during the illness, indicating streptococcal exposure. Two further cases presented with chorea and carditis (thus giving a definite diagnosis without serology). The remaining nine cases without elevated strep serology were treated as 'possible' ARF based on clinical suspicion.

Both inflammatory markers (CRP ≥30 and ESR ≥50) were elevated in 41 (62%) cases. Six cases had an elevated ESR but not CRP, while three cases had an elevated CRP but not ESR. One case had no data on CRP but had a raised ESR and three cases had no data on ESR but had raised CRPs. Fifteen cases (23%) were negative for both markers.

ARF cases with carditis were designated as either 'low-risk' (n=41, 80%) or 'medium-to-high-risk' (n=14, 20%) as per the Heart Foundation Guidelines.[10] Of the four recurrences, three had 'medium-to-high-risk' RHD. Of the total 'medium-to-high-risk' patients, nine were receiving appropriate outpatient surveillance. Three patients had moved out of Northland and two had been lost to follow-up. A total of eight cases (both 'low' and 'medium-to-high-risk' patients) were identified as lost to outpatient follow up, however seven of them were receiving penicillin prophylaxis. The substantial burden of ARF/RHD is profoundly demonstrated by the 14 'medium-to-high-risk' patients with significant valve disease and multiple valve repairs and replacements (Table 2).

In terms of dental follow-up, dental review data was available for only 43/69 cases. Of these, 67% (29/43) had had a dental review in the last 12 months, 14 had not. Only four of the 14 'medium-to-high-risk' cases were documented to have had a dental review in the last year.

Table 2: Current known status of RHD among 'medium-to-high-risk' cases.

Age at admission	Current known status of RHD
6	Mitral + aortic valve repair. Now mild MR, mild-moderate MS, moderate AR
8	Moderate MR + mild MS. Trace AR + TR
8	Moderate AR
9	Moderate MR & AR. Minor TR
12	Aortic + mitral valve repair with Cosgrove ring. Currently moderate AS + AR + moderate MS
13	Mild AR, mild-mod MR. Aortic root mildly dilated
13	Mitral valve repair. Now trivial MR + mild MS. Aortic root dilatation + mild LV dysfunction. Minor TR
15	Aortic valve replacement (bioprosthetic) & mitral annuloplasty
16	St Judes aortic + mitral valve replacements + tricuspid valve repair
20*	Moderate-severe AR + Moderate MR
20[X]	Moderate severe MR, moderate AR, mild-mod TR, moderate pulmonary hypertension
22*	Moderate MR, markedly dilated LA, moderately dilated LV
25[X]	Mitral valve repair and annuloplasty. Now moderate MR
50*	Mitral + aortic mechanical valve replacements

Key: MR = mitral regurgitation, MS= mitral stenosis, AR= aortic regurgitation, TR= tricuspid regurgitation, LA = left atrium, LV = left ventricle *= recurrence ARF [X]= diagnosed in outpatient clinic.

Discussion

The incidence rate of ARF cases for total Northland population has slightly decreased (7.7/100,000/year to 7/100,000/year) as compared to the previous audit, however, they are well above the BPS target incidence rate.[8] Similarly, ARF rates for Northland Māori children aged 5–14 years have also slightly decreased (78.0/100000/year to 64.5/100,000/year). Nevertheless, they still remain well above rates for Māori documented nationally (2011, 34/100,000/year), in the Waikato (2002–2011, 46.1/100,000/year) and in Tairāwhiti (1997–2009, 59/100,000/year).[13–15] Although Pacific people make only 2% of Northland population, 6% of the cases, with an incidence rate of 54.6/100,000/year among Pacific children aged 5–14 years, show the inequitable burden of ARF cases among Northland sub-population groups. The Māori and Pacific rates contrast drastically with the rates for non-Māori/non-Pacific aged 5–14 years (1.5/100,000/year). In the previous audit, total rates were only completed for Māori and non-Māori (ie, Pacific were included in Non-Māori).[6] Hence it is difficult to compare the rates of Pacific and non-Māori/non-Pacific populations. While annual rates were much reduced for 2015 and 2016 compared with the three years prior, the low absolute numbers of the audit require cautious interpretation, especially given the increased case numbers in 2017. It is possible that these low numbers were as a result of random variation as opposed to effective public health campaigns.

Recent publications from the MoH indicate that Northland had achieved its BPS target with an incidence of 3.5/100,000/year (six cases) in 2017[16]. However, it is important to note that the MoH's ARF/RHD case definitions differ greatly from the current and previous audits.[6,10] The MoH's definitions include only the hospitalised cases of first episodes of ARF/RHD with ARF as the principal diagnosis and exclude cases diagnosed in primary care and EDs.[10] Excluding cases that were not hospitalised can lead to undercounting of the burden of ARF/RHD in our communities.

The MoH has contracted the Institute of Environmental Science and Research Limited (ESRL) to reconcile ARF/RHD cases across New Zealand bi-annually. ESRL use hospitalisation (as defined by the BPS target) and EpiSurv case notification data and the case definitions in the Communicable Disease Control Manual.[17–19] Both the MoH and ESRL case definitions differ from Northland's audits. Dissimilarity in the MoH, ESRL and Northland audits' ARF/RHD case definitions has resulted in three different sets of case numbers and rates for Northland. The authors believe that it is imperative to have consistent case definitions to accurately measure the burden of ARF/RHD. Underestimation of the burden might have future funding implications on high-risk communities like Northland.

Seven cases were diagnosed through outpatient clinics and not through hospital admissions (discharges). Although two of these cases were in follow up to hospital admissions, the remaining five were referred directly from GP or ED to clinic.

Ten new cases were 20 years or older with two having 'medium-to-high-risk' carditis at the time of review/diagnosis. The four recurrences were all adults aged 20 years or older and 75% of these (3/4) had 'medium-to-high-risk' carditis at diagnosis. These cases raise the concern that adults presenting with ARF may have unnecessarily delayed diagnosis of ARF with the resulting development of RHD.

This audit only adds further to the present evidence that ARF in New Zealand is commonly seen in areas of high deprivation.[4,13–15] Fifty-five percent of cases were in NZDep 10 areas and 87% were in NZDep 8–10.

The previous audit commented that ESR may be a more reliable positive minor manifestation than CRP.[6] However, this was not demonstrated in this audit—six cases (10%) had an elevated ESR and not CRP, three cases (5%) had an elevated CRP and not ESR. In accordance with the National Guidelines, both ESR and CRP should be checked when investigating ARF.

Overall, there has been considerable improvement in the notification of ARF/RHD cases to EpiSurv. The previous audit identified 10 cases that were not notified to EpiSurv,[6] however, during this audit period only two cases were not notified to EpiSurv. The cases were more complex, likely reflecting acute on chronic ARF. Twenty percent of patients had 'medium-to-high-risk' carditis in accordance with 20%

noted in the previous audit. The burden of ARF/RHD is most evident among this group with multiple valve repairs and replacements. This group appeared to be difficult to contact with over one-third either missing their appointment, moving out of Northland or being lost to follow-up. Dental review documentation was difficult to gather and overall rates of review were poor.

ARF is a preventable disease. This repeat audit demonstrates that despite significant public health campaigns, little improvement has been seen for Northland Māori and despite low absolute numbers, Northland Pacific are also inequitably affected. Ninety-three percent of cases remain among Māori and the very small improvement in rates for Māori (one-fifth) is seen in the context of an extremely low incidence rate for non-Māori/non-Pacific. It appears that adults may be at an increased risk of a delayed diagnosis with the resulting risk of developing RHD. This highlights the difficulty public health campaigns often have in targeting the most at-risk populations. While further funding is being provided for Northland following the completion of the RFPP in 2017,[7,8] it appears that significant ongoing investment is required to reduce the burden of ARF in Northland. It seems unlikely that Northland will achieve its 2011 goal of eliminating ARF by 2020 without vast improvements in housing, income inequities and access to primary care. It is also imperative to have consistent case definitions to assess the true burden of ARF/RHD and to inform future directions (preventative and funding measures) to reduce the incidence of ARF/RHD in Northland.

Competing interests:
Nil.

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