The incidence of acute rheumatic fever in New Zealand (2010-2013)

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Ethical Approval and Consultation

This study received ethical approval from the Ministry of Health Central Health and Disability Ethics Committee (reference #: 14/CEN/86). Consultation was also undertaken with the Ngai Tahu Research Consultation Committee.

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Introduction

Acute rheumatic fever (ARF) is an auto-immune disorder resulting from exposure to strains of the *Group-A Streptococcus* (GAS) bacterium. Typical features of ARF include inflammation of joints, the central nervous system and, most seriously, cardiac valve tissue – with more than half of all ARF cases expected to develop chronic rheumatic heart disease (RHD).[1]

Beyond the primary prevention of ARF, a reduction in the burden of RHD may be possible by identifying the condition while the patient is still asymptomatic and then intervening with secondary antibiotic prophylaxis, or with cardiac surgery if severe disease is detected. It has been estimated that 40% of patients who present with symptomatic RHD do not have a known history of ARF.[2] For this reason, screening high risk populations has been recommended by some groups and is an ongoing area of debate in the literature.[3 4]

If screening for RHD is beneficial, then those who are most likely to benefit are the populations who have the highest incidence of disease. In a RHD screening pilot conducted in New Zealand over the past several years, the population targeted for screening were largely those of intermediate school age (10-13 years) who attended schools in the lowest socio-economic areas.[5] This targeting was based on the strong association between deprivation and ARF incidence; however by using this approach those with other risk factors (most notably Māori or Pacific ethnicity) may not be included if they live in less deprived areas.

Targeting of RHD screening to those with the highest prevalence of ARF will increase the positive predictive value of the screening test and reduce the number needed to screen per definite RHD identified. In order to maximise the efficacy of this targeting, we need to understand more regarding: a) which groups carry the highest burden of disease; b) where they reside; and c) how many people potentially comprise these target groups. For example, Māori and Pacific ethnicity are both strongly associated with ARF incidence independent of deprivation.[6]

To this end, the current study aimed to update and build on previous work in this area [6-8] by estimating the burden of ARF across multiple demographic and geographic strata. This required the achievement of the following objectives:

1) using literature review, summarise available data regarding the distribution of ARF in New Zealand by ethnicity, age, geographic region and deprivation (NZDep);
2) collect data from hospitalisations (*National Minimum Dataset [NMDS]*, Ministry of Health) and national notifications (*EpiSurv*, Environmental Science and Research) on all new cases of

ARF (2010-2013), including encrypted patient identifier (NHI) and all other available demographic information;

3) quantify the incidence (n) and distribution of ARF separately by geographic region, prioritised ethnicity, age group and deprivation level using census data as our denominator;
4) develop a risk prediction model for the New Zealand context which will allow us to simultaneously combine the effects of our predictors (e.g. ethnicity, deprivation, etc), and to identify groups who are most at risk (and therefore most likely to benefit from RHD screening);
5) using data from the 2013 census, quantify by geographic region the number of people who belong to the target ethnicity / age / deprivation groups (as identified from the risk prediction model).

The current brief report pertains to the first three of these objectives, while the final two objectives are the subject of a separate report.[9]

Methods

Data sources and definition of cases

There are three primary sources of ARF incidence data in New Zealand: hospitalisation data (National Minimum Dataset, or NMDS), national notification data (EpiSurv, maintained by the Institute of Environmental Science and Research) and 11 regional rheumatic fever registers. Each of these data sources have their own complexities and varying levels of data completeness.[8] It is thought that hospitalisation data may overestimate the true incidence of ARF largely because of misclassification of rheumatic heart disease as acute rheumatic fever, while notification data may underestimate the incidence of ARF because of under-reporting.[8 10 11] Regional registers are thought to be the most complete list of patients with definite ARF, since their primary role is as a patient management system to assist with delivery of secondary prophylaxis. However completeness of these registers varies by region,[8] and given their primary purpose they are most likely to capture cases who are at the more severe end of the ARF spectrum.[12]

Given that the purpose of the current study was to estimate the underlying burden of ARF across multiple demographic and geographic strata – combined with the likelihood that many new ARF cases do not present to health care services at all [4] and thus will not be captured by any data source – we aimed to optimise the sensitivity of case identification. In other words, we tended towards a low

threshold for identifying an individual as a case in the first instance; and as such, our initial cohort was defined using hospitalisation (NMDS) data. Since 13% of all ARF cases who are notified to the EpiSurv database are not recorded as having a corresponding hospitalisation [8] – and thus would not be included in our initial cohort – we made the decision to augment our initial cohort with cases from the EpiSurv database.

Participants

Each new case of ARF identified between 2010-2013 were included in this study. To identify cases, we requested hospitalisation (NMDS) data from the Ministry of Health pertaining to all hospitalisations that occurred between 2010-2013 in which a primary diagnosis of ARF was made (ICD-10-AM codes: I00-I02). Secondly, we requested notification data ('EpiSurv') from the Institute of Environmental Science and Research for all new cases of ARF reported 2010-2013. The notification data was then merged (by unique patient identifier) with the hospitalisation data. In the event that a patient and associated data existed in both cohorts, a 'give way' rule was applied whereby hospitalisation data was preferred to notification data.

For any given patient, only data pertaining to the earliest ARF diagnosis between 2010-2013 was retained (i.e. one patient = one diagnosis). Since it was feasible that the diagnosis dates recorded on the hospitalisation and notification datasets might not be identical, a four-week diagnosis 'window' was employed when merging these datasets – whereby hospitalisation and notification data were assumed to refer to the same underlying ARF event provided the diagnosis dates were within four weeks of each other. The earliest recorded date of diagnosis for a given ARF case was retained as the index date for that patient.

After merging the hospitalisation and notification datasets, a total of n=929 unique ARF cases were identified. In order to increase the specificity of our measure, we excluded those who had a recorded history of ARF (prior to 2010) or chronic rheumatic heart disease (any time prior to the ARF diagnosis date) using a case-identification algorithm. This is the same case identification algorithm that is used by the Ministry of Health to determine ARF incidence (Appendix 1). The algorithm restricts the attribution of ARF incidence to those patients for whom: 1) ARF is the primary diagnosis only; 2) no previous primary or additional diagnoses of ARF are recorded; and 3) no previous primary or additional diagnoses of rheumatic heart disease (RHD) are recorded.[8] In order to apply this algorithm, we linked all patients in the cohort to their hospitalisation records from 1988 (the earliest year that these data are available [13]) or their birth (whichever was earlier), for evidence of previous ARF (ICD-10-AM codes: I00-I02) or chronic RHD (I05-I09). Based on this algorithm, we excluded 156 patients

from further analysis. Using hospitalisation data, we also excluded those who were recorded as being a non-New Zealand resident at the time of their ARF (n=38). Following exclusions, a final cohort of n=733 remained for further analysis (Appendix 2).

For the purposes of our incidence analysis, we accessed publicly-available New Zealand Census data in order to determine the total number of people in each demographic (ethnicity, deprivation, age) and geographic group. These stratified populations served as respective denominators for our incidence analysis. These denominators are further described in the Statistical Analysis section of this manuscript.

Variables

Ethnicity, geographic location (domicile code or Census Area Unit) and date of birth/age were determined from both the hospitalisation and notification datasets, with hospitalisation data preferred to notification data in line with our 'give way' rule. If hospitalisation data was incomplete for these variables, then notification data (if available) were used to fill these gaps.

Ethnicity. Patient ethnicity was determined using the total ethnicity approach.[14] Using this approach, patients were placed into all ethnic groups to which they were recorded as having an affiliation. In this way, a given patient could belong to any or all of Māori, Pacific or Asian ethnic groups, while those who were not recorded as having any of these three affiliations were recorded as non-Māori/Pacific/Asian (otherwise known as 'European/Other').

Age. Patient age was determined from date of birth (NMDS) or age at diagnosis (EpiSurv) data. Age was treated as both a continuous (when calculating median age) and categorical variable (age categories: 0-4, 5-14, 15-29, 30+).[8]

Geographic location: The geographic location of each patient was attributed based on the Census Area Unit where they lived at the time of ARF incidence. In urban areas, the boundaries of a Census Area Unit generally coincide with a suburb, while Census Area Units in rural areas generally span larger geographic areas.[15] Census Area Unit was determined from the domicile code attributed to the patient at time of hospitalisation using a concordance file. Notification (EpiSurv) data were used to augment Census Area Unit data in those cases where a) the domicile code mapped to non-existent or out-of-date Census Area Units or b) domicile code did not exist on the hospitalisation data but Census Area Unit did exist on the relevant notification data. Following augmentation, Census Area Unit could not be determined for a total of n=2 (0.3% of cohort) patients.

Using a concordance file,[16] Census Area Unit was then used to determine the District Health Board that a patient was residing in at the time of their diagnosis. District Health Board could not be determined for a total of n=10 (1.4% of cohort) patients.

Deprivation. Deprivation was determined using the NZDep index, which attributes level of deprivation based on the Census Area Unit where the patient resided at the time of ARF diagnosis. Incomplete Census Area Unit data (n=2, 0.3% cohort) and a lack of NZDep score availability (n=20, 2.7% of cohort) prevented attribution of NZDep in n=22 cases (3% cohort).

Rurality. Rurality was set using a modified version of the Urban/Rural Profile Classification,[17] a classification system which allows mapping of Census Area Unit down to three classifications: Urban (Main Urban Area + Satellite Urban Area); Independent Urban Area; and Rural (all rural areas). Incomplete data prevented the attribution of Urban/Rural Profile Classification in n=14 cases (1.9% of total cohort).

Statistical analysis

We quantified the incidence of ARF separately by ethnicity, age group, deprivation, rurality and geographic location (DHB and Census Area Unit). In addition to descriptive analyses, we calculated crude and age-standardised incidence rates (per 100,000) using relevant Census population data as the denominator. Data from the 2013 Census were used in all cases, with the exception of rurality for which 2006 Census data were used due to the unavailability of Urban/Rural Profile Classification denominator data for the 2013 Census.[15]

Data were collected from Statistics New Zealand for the ethnicity, age group, rurality and geographic location denominators,[18] while deprivation denominators were requested and received from the developers of the NZDep tool.[19] Age standardisation of incidence rates was performed using direct standardisation methods,[20] with the total 2013 New Zealand Census population used as the standard population.

All analyses were performed in SAS v9.3 and Microsoft Excel.

Results

A total of 929 unique cases of ARF were identified from NMDS (810 unique cases) and EpiSurv (664 unique cases) data. Following exclusions (2 cases removed due to missing NHI, 156 cases removed due to previous ARF/RHD, 38 cases removed due to non-NZ residency status), a total of 733 cases remained for further analysis.

Demographic characteristics of the total cohort are shown in Table 1. The burden of ARF was greater among males (age-standardised incidence rate: 4.7/100,000) than females (3.8/100,000), with females 20% less likely than males to be diagnosed with the disease (age-standardised relative risk [RR]: 0.80, 95% CI 0.70-0.93). The median age at diagnosis was 12 years old (range: 4-85), with those aged 5-14 approximately 50 times more likely to be diagnosed with ARF than those aged over 30(crude RR: 49.4, 95% CI 36.3-67.3). Māori (age-standardised incidence rate: 11.7/100,000) and Pacific (17.6/100,000) were substantially more likely to be diagnosed with ARF than the European/Other population (age-standardised RR: Māori 28.8, 95% CI 21.3-38.9; Pacific 43.3, 95% CI 31.9-58.7). We observed a significant deprivation gradient, whereby those residing in the most deprived deciles (NZDep 9-10) were more than 30 times more likely to be diagnosed with ARF compared to those residing in the least deprived deciles (NZDep 1-2; age-standardised RR 33.3, 95% CI 19.1-58.1). Those residing in rural areas were nearly half as likely to be diagnosed with ARF compared to those residing in urban areas (age-standardised RR: 0.58, 95% CI 0.44-0.75).

When stratifying our demographic analysis by ethnicity (Table 1), we found similar patterns across ethnic groups in terms of gender split – whereby females (age-standardised incidence rates: Māori 10.2/100,000, Pacific 17.3/100,000, Asian 0.3/100,000, Euro/Other 0.4/100,000) were somewhat less likely to be diagnosed with ARF compared to males (Māori 13.1/100,000, Pacific 17.7/100,000, Asian 1.1/100,000, Euro/Other 0.4/100,000). The median age at diagnosis was the same for Māori and Pacific populations (12 years), similar for Asian cases (11 years) and somewhat higher for Euro/Other cases (18 years). The strong deprivation gradient was most evident for Māori and Pacific cases with 72% and 77% respectively residing in the most-deprived deciles (NZDep 9-10) at time of diagnosis, compared to 38% of Asian cases and 25% of European/Other cases. Almost all Pacific cases (97%) resided in urban areas, compared to 70% Māori, 92% Asian and 60% European/Other.

Table 3 shows the incidence of ARF across the 20 District Health Boards (DHB). In terms of absolute number of cases, Counties-Manukau DHB sustained the greatest burden of disease – with approximately 61 cases diagnosed per year in this region (age-standardised incidence rate: 11.5/100,000). Although sustaining fewer absolute cases of ARF (7 per year), Tairawhiti DHB experienced the greatest incidence of disease (13.5/100,000). In total, two-thirds of the 733 cases

observed over the study period occurred across the four northern-most DHBs (480 cases, or 65%). When stratifying this analysis by ethnicity (for Māori and Pacific populations), we observed high variability in terms of ethnicity-specific incidence rates by DHB – for example, 28/100,000 Māori living in Northland were diagnosed with ARF over our study period compared to only 5/100,000 Māori living in the Taranaki region (age-standardised incidence rates). It should be noted that due to disease rarity, incidence rates for some ethnicity/DHB strata had wide confidence intervals (Figure 2).

Table 1: *Incidence of acute rheumatic fever in New Zealand (2010-2013), by patient characteristic.*

	Total Cases		Average Cases	Incidence Rate [3]	Relative Risk [4]	
	(n)	(% total)	(n/year)	(n/100,000/year)	Crude RR (95% CI)	Age Adj. RR (95 % CI)
Total Cohort	733	100%	183 per year	4.3 (3.7-4.9)	-	-
Sex						
Female	323	44%	81 per year	3.8 (3.4-4.3)	0.83 (0.72 - 0.96)	0.8 (0.7-0.93)
Male	410	56%	103 per year	4.8 (4.3-5.3)	Ref	Ref
Age (years)						
<5	3	<1%	1 per year	0.3 (0-0.8)	0.59 (0.18 - 1.9)	-
5-14	493	67%	123 per year	21.5 (17.7-25.3)	49.5 (36.3 - 67.3)	-
15-29	193	26%	48 per year	5.7 (4.1-7.3)	13.1 (9.5 - 18.2)	-
30+	44	6%	11 per year	0.4 (0.2-0.7)	Ref	-
Median Age (Range)	12 (4 - 85)					
Ethnicity						
Māori	394	54%	99 per year	11.7 (10.6-13)	43 (31.9 - 58)	28.8 (21.3-38.9)
Pacific Island	311	42%	78 per year	17.6 (15.6-19.7)	62.7 (46.3 - 85)	43.3 (31.9-58.7)
Asian	13	2%	3 per year	0.7 (0.4-1.2)	1.8 (1 - 3.3)	1.8 (0.9-3.2)
Euro/Other	48	7%	12 per year	0.4 (0.3-0.5)	Ref	Ref
Deprivation [1]						
Lowest Deprivation: 1-2	13	2%	3 per year	0.4 (0.2-0.7)	Ref	Ref
3-4	36	5%	9 per year	1.1 (0.8-1.5)	2.8 (1.5 - 5.3)	2.8 (1.5-5.3)
5-6	53	7%	13 per year	1.7 (1.3-2.2)	4.2 (2.3 - 7.8)	4.2 (2.3-7.8)
7-8	109	15%	27 per year	3.4 (2.8-4.1)	8.8 (5 - 15.7)	8.6 (4.8-15.3)
Highest Deprivation: 9-10	500	70%	125 per year	13.1 (12-14.3)	40.3 (23.2 - 69.9)	33.3 (19.1-58.1)
Rurality [2]						
Urban	577	80%	144 per year	4.5 (4.1-4.9)	Ref	Ref
Independent Urban	80	11%	20 per year	4.5 (3.6-5.7)	0.95 (0.75 - 1.2)	1 (0.8-1.3)
Rural	62	9%	16 per year	2.6 (2-3.3)	0.58 (0.44 - 0.75)	0.58 (0.44-0.75)

[1] Defined using 2013 mapping of Census Area Unit to NZDep. [2] Defined using 2006 mapping of Census Area Unit to Urban/Rural Profile Classification. [3] Age-standardised to 2013 New Zealand Census population (with the exception of age category). 4 Age-standardised relative risk.

Table 2: *Incidence of acute rheumatic fever in New Zealand (2010-2013), by patient characteristic, stratified by ethnic group.*

	Māori			Pacific Island			Asian			Euro/Other		
	Total Cases		Incidence Rate [3]	Total Cases		Incidence Rate [3]	Total Cases		Incidence Rate [3]	Total Cases		Incidence Rate [3]
	(n)	*(%)*	*(n/100,000/year)*	*(n)*	*(%)*	*(n/100,000/year)*	*(n)*	*(%)*	*(n/100,000/year)*	*(n)*	*(%)*	*(n/100,000/year)*
Total Cohort	**394**	**54%**	**11.7 (10.6-13)**	**311**	**42%**	**17.6 (15.6-19.7)**	**13**	**2%**	**0.7 (0.4-1.2)**	**48**	**7%**	**0.4 (0.3-0.5)**
Sex												
Female	167	42%	10.2 (8.7-11.9)	147	47%	17.3 (14.6-20.4)	3	23%	0.3 (0.1-1.1)	22	46%	0.4 (0.2-0.6)
Male	227	58%	13.1 (11.4-15)	164	53%	17.7 (15.1-20.7)	10	77%	1.1 (0.6-2)	26	54%	0.4 (0.3-0.6)
Age (years)												
<5	0	0%	0 (0-0)	2	1%	1.3 (0-5)	0	0%	0 (0-0)	1	2%	0.1 (0-0.6)
5-14	271	69%	51.6 (39.3-63.9)	218	70%	80.5 (59.2-101.9)	10	77%	4.1 (0-9.1)	17	35%	1.1 (0.1-2.1)
15-29	104	26%	18 (11.1-25)	81	26%	26.4 (14.9-38)	1	8%	0.2 (0-0.9)	17	35%	0.8 (0-1.5)
30+	19	5%	1.9 (0.2-3.6)	10	3%	2.2 (0-4.9)	2	15%	0.2 (0-0.8)	13	27%	0.2 (0-0.4)
Median Age (Range)	12 (5 - 57)			12 (4 - 39)			11 (6 - 43)			18 (4 - 85)		
Deprivation [1]												
Lowest Deprivation: 1-2	4	1%	1.4 (0.5-3.9)	0	0%	-	2	15%	0.6 (0.1-2.4)	7	15%	0.3 (0.1-0.6)
3-4	19	5%	5.1 (3.2-8.2)	8	3%	5.8 (2.9-11.6)	2	15%	0.5 (0.1-2)	9	19%	0.4 (0.2-0.8)
5-6	26	7%	4.6 (3.1-6.9)	20	7%	9.3 (6-14.4)	1	8%	0.3 (0-1.8)	9	19%	0.4 (0.2-0.8)
7-8	60	16%	7.6 (5.9-9.8)	41	14%	11.6 (8.5-16)	3	23%	0.8 (0.3-2.6)	11	23%	0.7 (0.4-1.2)
Highest Deprivation: 9-10	278	72%	20.4 (18.1-23.1)	225	77%	22.7 (19.8-26)	5	38%	1.5 (0.6-3.6)	12	25%	1.2 (0.7-2.2)
Rurality [2]												0 (0-0)
Urban	272	70%	17.5 (13.3-21.6)	293	97%	29.5 (22.7-36.2)	12	92%	0.9 (0-1.9)	29	60%	0.3 (0.1-0.6)
Independent Urban	71	18%	20.2 (10.8-29.5)	5	2%	11.5 (0-31.8)	0	0%	0 (0-0)	6	13%	0.4 (0-1.1)
Rural	46	12%	13.1 (5.5-20.7)	3	1%	11.2 (0-36.5)	1	8%	3.8 (0-18.8)	13	27%	0.7 (0-1.4)

[1] Defined using 2013 mapping of Census Area Unit to NZDep. [2] Defined using 2006 mapping of Census Area Unit to Urban/Rural Profile Classification. [3] Total cohort, sex and deprivation incidence rates are age-standardised to the 2013 New Zealand Census population, using relevant denominators (e.g. total number of Maori males). Age- and ethnicity- stratified denominator data for URPC status were unavailable, and thus crude incidence rates are shown for this variable (using the total 2013 New Zealand Census population as the denominator).

Table 3: *Incidence of acute rheumatic fever in New Zealand (2010-2013), by district health board.*

	Total Cases		Average Cases	Incidence Rate [1]
	(n)	(% total)	(n/year)	(n/100,000/year)
District Health Board				
Northland	73	10%	18 per year	12.2 (9.7-15.4)
Waitemata	40	6%	10 per year	1.9 (1.4-2.6)
Auckland	52	7%	13 per year	3.1 (2.3-4)
Counties Manukau	245	34%	61 per year	11.5 (10.1-13)
Waikato	70	10%	18 per year	4.7 (3.7-5.9)
Lakes	31	4%	8 per year	7.7 (5.4-10.9)
Bay of Plenty	37	5%	9 per year	4.5 (3.2-6.2)
Tairawhiti	26	4%	7 per year	13.5 (9.2-19.9)
Taranaki	7	1%	2 per year	1.5 (0.7-3.2)
Hawkes Bay	24	3%	6 per year	3.8 (2.5-5.7)
Whanganui	8	1%	2 per year	3.4 (1.7-6.9)
MidCentral	13	2%	3 per year	2 (1.2-3.4)
Hutt	33	5%	8 per year	5.9 (4.2-8.4)
Capital and Coast	38	5%	10 per year	3.4 (2.4-4.6)
Wairarapa	0	0%	0 per year	-
Nelson Marlborough	2	<1%	1 per year	0.4 (0.1-1.8)
West Coast	1	<1%	<1 per year	1 (0.1-6.9)
Canterbury	19	3%	5 per year	1 (0.7-1.6)
South Canterbury	0	0%	0 per year	-
Southern	4	1%	1 per year	0.3 (0.1-0.9)

[1] Age-standardised to 2013 New Zealand Census population. Ordered from (approximate) geographic north to south. District health board boundaries can be viewed at: http://www.health.govt.nz/new-zealand-health-system/key-health-sector-organisations-and-people/district-health-boards/location-boundaries-map.

Figure 1: *Age-standardised incidence of acute rheumatic fever (2010-2013), by deprivation quintile and ethnic group.*

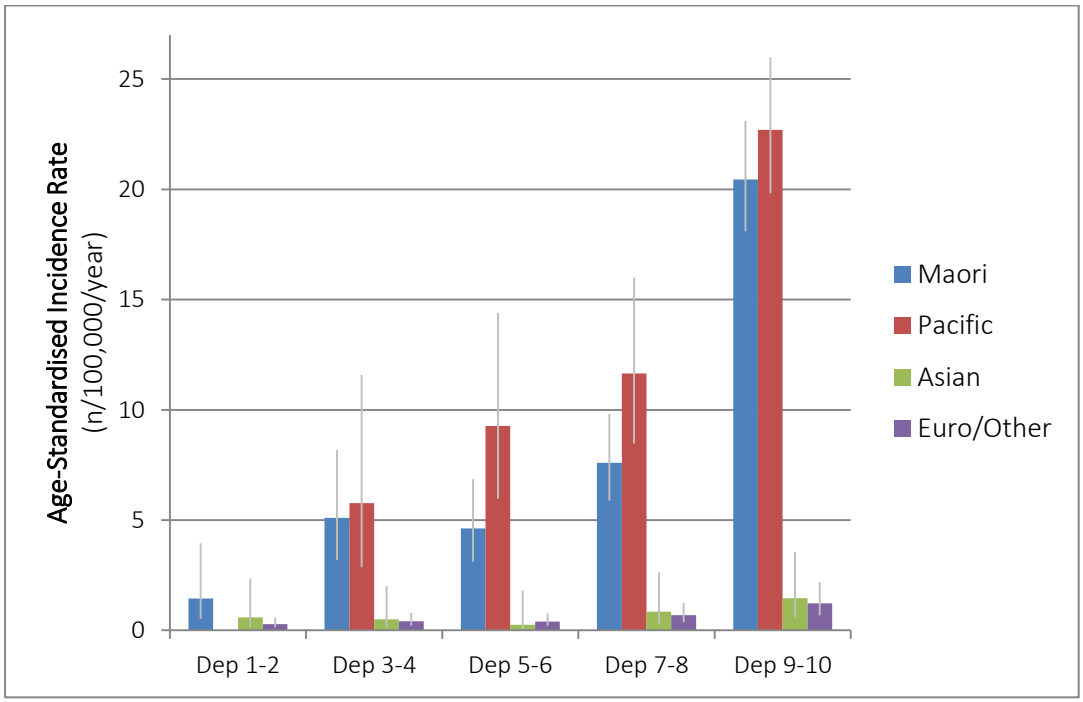
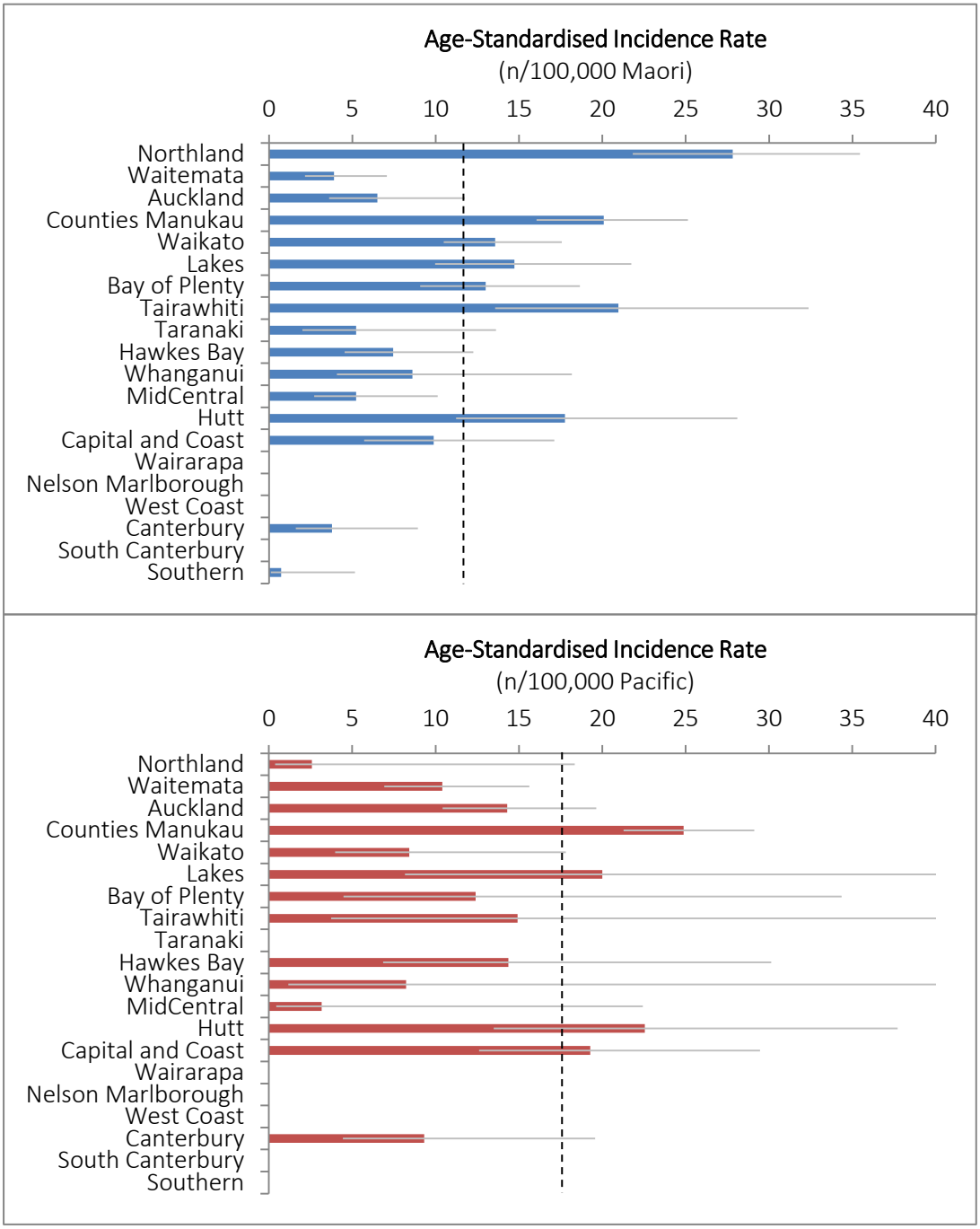


Figure 2: *Age-standardised incidence of acute rheumatic fever (2010-2013) by district health board [1], for Māori and Pacific populations. Dashed lines demark national incidence rates for the given ethnic group.*



[1] Ordered from (approximate) geographic north to south. District health board boundaries can be viewed at: http://www.health.govt.nz/new-zealand-health-system/key-health-sector-organisations-and-people/district-health-boards/location-boundaries-map.

General Discussion

The current study builds-on and updates previous evidence regarding the distribution of acute rheumatic fever incidence in New Zealand over a four-year period. Many of the observations we report are neither new nor unique – but rather a re-telling of a previously-described profound inequity between population sub-groups.[6 7]

While acute rheumatic fever (ARF) is uncommon in the general population, it differentially affects some population sub-groups over others. More than 9 out of every 10 cases occur among Māori or Pacific New Zealanders, with Māori nearly 30 times more likely to sustain ARF than the European/Other population – and Pacific more than 40 times as likely (Table 1). We also noted that those residing in the most deprived areas were more than 30 times as likely to sustain ARF compared to those residing in the least deprived areas (adjusted RR: 33.33, 95% CI 19.12-58.11). Rurality appeared to have a somewhat protective effect – with those living in rural areas nearly half as likely to sustain ARF compared to those living in urban areas (adjusted RR: 0.58 95% CI 0.44-0.75).

Since Māori and Pacific New Zealanders are more likely to reside in areas of high deprivation compared to other ethnic groups,[21] it is intuitive to conflate the highly-differential patterns of ARF incidence with level of deprivation – particularly given the likely role of poverty-related exposures in the aetiology of this disease. However, when stratifying disease incidence by deprivation level, we found that Māori and Pacific New Zealanders are substantially more likely to be affected by this disease regardless of NZDep decile – suggesting that while deprivation is certainly an exposure of great importance, it is unlikely to be the sole explanatory factor for this ethnic inequity. As suggested by Jaine et al., it is more likely that deprivation is a proxy for other associated exposures, such as overcrowding.[7]

We also note wide variation in the relative burden of disease within Māori and Pacific populations by geographic region – with Māori living in Northland (age-standardised incidence rate: 27.8/100,000 Māori) some four times more likely to develop this disease than Māori living in Auckland (6.5/100,000), and some five times more likely than those living in Taranaki (5.2/100,000). Likewise for Pacific peoples, those living in Counties-Manukau (24.9/100,000 Pacific peoples) were observed to be three times more likely to develop ARF than Pacific peoples living in Waikato (8.4/100,000) or Canterbury (9.3/100,000; Figure 2). It should be noted that these estimates are, in several instances, based on a small number of RF cases – and thus the observed regional variation by ethnicity could be purely due to chance. However, such patterning could plausibly be related to region-specific risk factors such as climate – or more generic ones that may apply to multiple regions, such as poverty

and over-crowding. This cannot be inferred from our findings, and thus requires further fine-grained investigation.

Limitations

This study has strengths and weaknesses. A major strength is the high-quality nature of the national-level data employed, drawn from two unique sources (i.e. hospitalisation and notification data). We do, however, note that there are inherent weaknesses with using administrative data to identify new cases of acute rheumatic fever. The ARF dataset is influenced by factors causing it to both under- and over-count the true number of cases. ARF is a syndromic diagnosis, and clinically important disease has a wide spectrum of symptoms from mild (even asymptomatic) to severe. Cases can be missed if these individuals: a) did not seek medical attention for their symptoms, b) did not have their (likely mild) symptoms recognised as ARF when they did present to health care services, or c) were neither admitted to hospital nor notified to a medical officer of health, despite being diagnosed with ARF. There is evidence that this case under-ascertainment is large: for example, less than half (41%) of the 1,016 RHD cases under 20 years old diagnosed between 1997-2010 had previously been admitted to hospital and diagnosed with ARF.[22] This undercount of the true number of ARF cases over our study period would thus make the rates of disease that we have reported here conservative.

There are also factors leading to some over-count of the true number of cases. Evidence from regional registers – which serve as vital patient management tools – suggests that national-level data over-estimate the number of diagnosed ARF cases.[11 23] In this scenario, our dataset would over-count the number of ARF cases that occurred between 2010-2013. Along this line, we also note that recent evidence suggests that rates of ARF in New Zealand appear to have reduced in the years 2014 and 2015 [24] – meaning that the rates of disease reported here may have attenuated somewhat since the end of the study period.

We must also consider the possibility that at least some of those cases diagnosed among older age groups (e.g. >30 years; 6% of all ARF cases [Table 1]) are in fact recurrences of cases that occurred earlier in life – cases that were either a) not diagnosed and reported to the central data repositories employed in the current study, or b) occurred prior to 1988 (when NHI use became universal for hospitalisation records). Such a scenario would result in an over-counting of cases in older age groups – with an associated increase in stratum-specific rates of disease in that age group.

Application of study findings to estimates of future disease burden

This report describes the likely burden of acute rheumatic fever for the period 2010 to 2013. While the patterns we describe – particularly with respect to ethnicity and deprivation – largely echo those described in previous decades,[6 7] we make no inferences or projections regarding the likely future burden of disease based on the estimates provided here. We note that the Ministry of Health has reported [24] a reduction in the number of ARF cases between 2014 and 2015. Whether this apparent reduction in disease burden is a real phenomenon – catalysed by interventions such as the national throat-swabbing programme – or a transient phenomenon remains to be seen, and will only be confirmed in retrospect.

Conclusions

In line with findings from previous cohorts, we observed that the burden of acute rheumatic fever in New Zealand is highly-differential by population sub-group. ARF is almost exclusively a disease of Māori and Pacific New Zealanders, with these populations more likely to be affected by ARF than other ethnic groups regardless of level of deprivation. We observed substantial geographic variation in the burden of ARF within Māori and Pacific peoples – a finding which suggests that ethnicity is not an ARF risk factor in silo, but rather a proxy for a multitude of risk factors (including deprivation and/or overcrowding) that disproportionately affect Māori and Pacific peoples.

References

1. Carapetis JR, Steer AC, Mulholland EK, et al. The global burden of group A streptococcal diseases. Lancet Infectious Diseases 2005;**5**(11):685-94.
2. Carapetis JR, Currie BJ, Mathews JD. Cumulative incidence of rheumatic fever in an endemic region: a guide to the susceptibility of the population? . Epidemiology and Infection 2000;**124**(239–244).
3. Marijon E, Mirabel M, Celermajer DS, et al. Rheumatic heart disease. The Lancet 2012;**379**(9819):953-64.
4. Roberts K, Colquhoun S, Steer A, et al. Screening for rheumatic heart disease: Current approaches and controversies. Nature Reviews Cardiology 2013;**10**(1):49-58.
5. Webb RH, Wilson NJ, Lennon DR, et al. Optimising echocardiographic screening for rheumatic heart disease in New Zealand: not all valve disease is rheumatic. Cardiology in the Young 2011;**21**(04):436-43.
6. Milne RJ, Lennon DR, Stewart JM, et al. Incidence of acute rheumatic fever in New Zealand children and youth. Journal of Paediatrics and Child Health 2012;**48**(8):685-91.
7. Jaine R, Baker M, Venugopal K. Epidemiology of acute rheumatic fever in New Zealand 1996-2005. Journal of Paediatrics and Child Health 2008;**44**(10):564-71.
8. Oliver J, Pierse N, Baker M. Estimating rheumatic fever incidence in New Zealand using multiple data sources. Epidemiol Infect 2015;**43**(1):167-77.
9. Gurney JK, Stanley J, Baker MG, et al. Estimating the risk of acute rheumatic fever in New Zealand by age, ethnicity and deprivation. Epidemiol Infect 2016;**144**(14):3058-67.
10. Is a rheumatic fever register the best way to evaluate the government programme to control rheumatic fever? . The Paediatric Society of New Zealand 64th Annual Scientific Meeting; 2012; Auckland, New Zealand.
11. Robin A, Mills C, Tuck R, et al. The epidemiology of acute rheumatic fever in Northland, 2002-2011. New Zealand Medical Journal 2013;**126**(1373):46-52.
12. Reményi B, Wilson N, Steer A, et al. World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease-an evidence-based guideline. Nature Reviews Cardiology 2012;**9**(5):297-309.
13. MoH. National Minimum Dataset Data Mart Dictionary. Wellington: Ministry of Health, 2010.
14. Cormack D, Robson C. Classification and output of multiple ethnicities: considerations for monitoring Māori health. Wellington: Te Rōpū Rangahau Hauora a Eru Pōmare, University of Otago, Wellington, 2010.
15. Statistics New Zealand. Area unit: classification and coding process 2015 [Available from: http://www.stats.govt.nz/methods/classifications-and-standards/classification-related-stats-standards/area-unit/classification-and-coding-process.aspx.
16. Statistics New Zealand. Geographic area files 2015 [
17. Robson B, Purdie G, Cormack D. Unequal Impact II: Māori and Non-Māori Cancer Statistics by Deprivation and Rural-Urban Status, 2002-2006. Wellington: Ministry of Health, 2010.
18. Statistics New Zealand. 2013 New Zealand Census: Data tables 2015 [Available from: http://www.stats.govt.nz/Census/2013-census/data-tables.aspx.
19. University of Otago. Socioeconomic Deprivation Indexes: NZDep and NZiDep: Department of Public Health, University of Otago, Wellington; [Available from: http://www.otago.ac.nz/wellington/departments/publichealth/research/hirp/otago020194.html.
20. MoH. Standardising rates of disease. Wellington: Ministry of Health, 1998.
21. Salmond C, Crampton P. Development of New Zealand's Deprivation Index (NZDep) and its uptake as a national policy tool. Canadian Journal of Public Health 2012;**103**(S2):S7-S11.
22. Oliver J, Pierse N, Baker M. Improving rheumatic fever surveillance in New Zealand. . Wellington, New Zealand Ministry of Health, 2013.

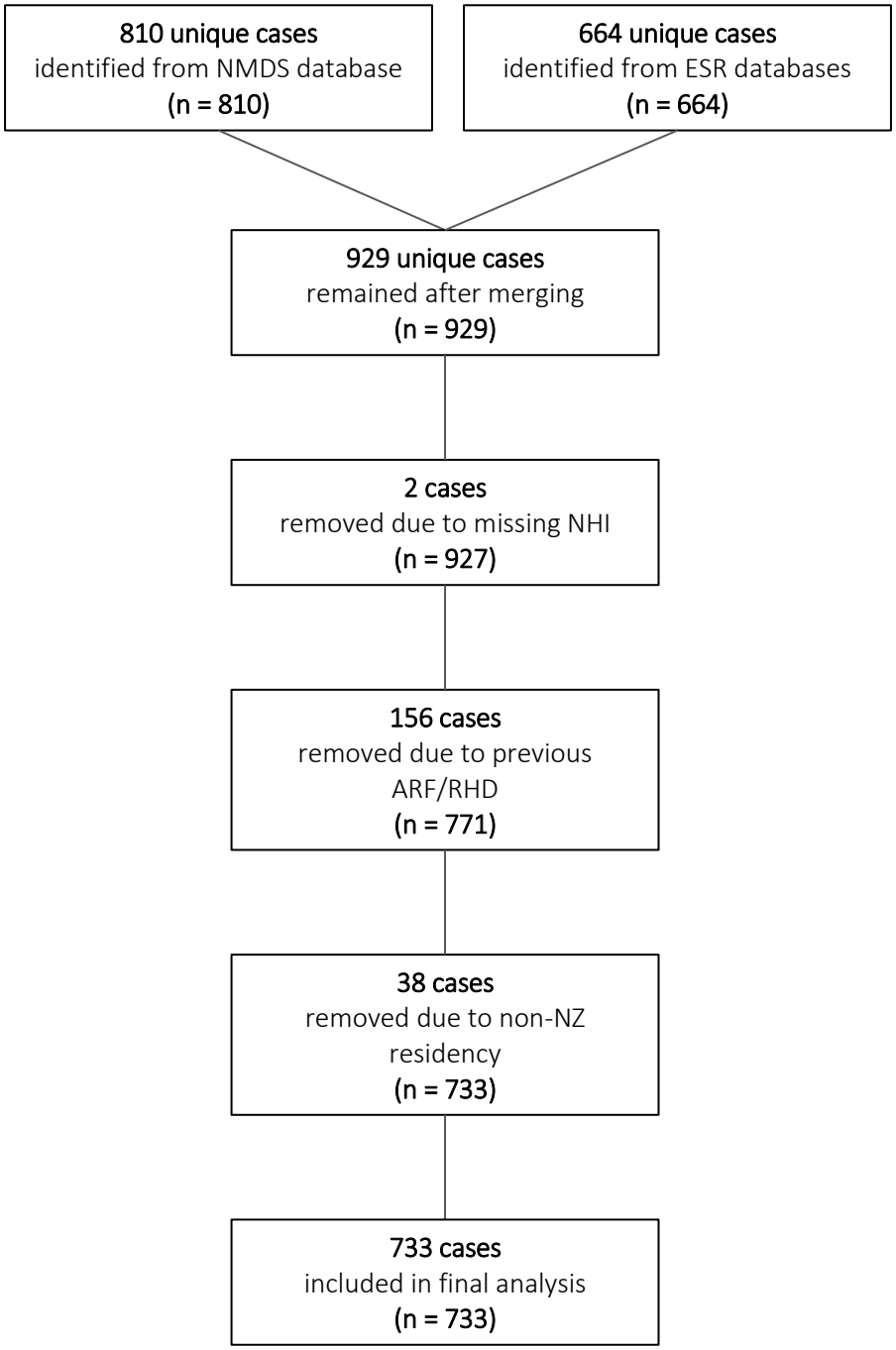
23. Siriett V, Crengle S, Lennon D, et al. The epidemiology of rheumatic fever in the Tairawhiti/Gisborne region of New Zealand: 1997-2009. The New Zealand medical journal 2012;**125**(1365):8-15.
24. Ministry of Health. Progress on the Better Public Services rheumatic fever target Wellington, New Zealand2015 [Available from: http://www.health.govt.nz/about-ministry/what-we-do/strategic-direction/better-public-services/progress-better-public-services-rheumatic-fever-target.

Appendices

Appendix 1: *Algorithm for determining new ARF cases from hospitalisation data*. *Document provided by Ministry of Health on 13/05/2014.*

ICD codes used:	ICD-10-AM diagnosis codes: I00, I01, I02 (Acute rheumatic fever)
	ICD 9 CM-A diagnosis codes: 390, 391, 392 (Acute rheumatic fever)
	ICD-10-AM diagnosis codes: I05-I09 (Chronic rheumatic heart disease)
	ICD 9 CM-A diagnosis codes: 393-398 (Chronic rheumatic heart disease)
Inclusions:	Principal diagnoses (Acute rheumatic fever) only
	Overnight admissions
	Day-case admissions
Exclusions:	Previous acute rheumatic fever diagnosis (principal and additional) from 1988
	Previous chronic rheumatic heart disease diagnosis (principal and additional) from 1988
	New Zealand non-residents
Transfers:	Transfers with a principal diagnosis of acute rheumatic fever are counted as one acute rheumatic fever hospitalisation episode
Timeframe:	Trends from 2002 onwards

Appendix 2: *Patient inclusion/exclusion flow chart.*



Appendix 3: *Incidence of acute rheumatic fever, by Census Area Unit. Table restricted to those CAUs with greater than four (4) cases over the study period.*

		Total Cases		Incidence Rate [1]
		(n)	(% total)	(n/100,000/year)
Census Area Unit	District Health Board			
Burbank	*Counties Manukau*	13	2%	96.7 (0-201.9)
Viscount	*Counties Manukau*	10	1%	62.8 (0-140.7)
Harania West	*Counties Manukau*	10	1%	51.1 (0-114.4)
Weymouth West	*Counties Manukau*	9	1%	53.6 (0-123.6)
Kaikohe	*Northland*	8	1%	51.1 (0-121.9)
Rongomai	*Counties Manukau*	8	1%	45 (0-107.4)
Clover Park	*Counties Manukau*	8	1%	48 (0-114.4)
Aorere	*Counties Manukau*	8	1%	37 (0-88.2)
Otahuhu West	*Auckland*	7	1%	35.5 (0-88.1)
Clendon South	*Counties Manukau*	7	1%	35.3 (0-87.7)
Aotea	*Waikato*	7	1%	58.7 (0-145.8)
Otara North	*Counties Manukau*	6	1%	89.9 (0-233.8)
Favona South	*Counties Manukau*	6	1%	45.6 (0-118.5)
Harania East	*Counties Manukau*	6	1%	29.8 (0-77.4)
Leabank	*Counties Manukau*	6	1%	28.4 (0-73.9)
Pukekohe North	*Counties Manukau*	6	1%	16.8 (0-43.6)
Ngaruawahia	*Waikato*	6	1%	29.3 (0-76.1)
Kingsley-Chatham	*Hawke's Bay*	6	1%	50 (0-129.9)
Raumanga West	*Northland*	5	1%	44.3 (0-122)
Waimumu North	*Waitemata*	5	1%	21.8 (0-59.9)
Wymondley	*Counties Manukau*	5	1%	105.5 (0-290.4)
Otara East	*Counties Manukau*	5	1%	28.3 (0-77.8)
Otara South	*Counties Manukau*	5	1%	37.8 (0-104.2)
Donegal Park	*Counties Manukau*	5	1%	18.5 (0-50.9)
Mangere South	*Counties Manukau*	5	1%	17.6 (0-48.3)
Clendon North	*Counties Manukau*	5	1%	44 (0-121.3)
Papakura East	*Counties Manukau*	5	1%	20.7 (0-57.1)
Cannons Creek North	*Capital and Coast*	5	1%	39.9 (0-109.9)
Cannons Creek East	*Capital and Coast*	5	1%	34.8 (0-95.8)
Waitangirua	*Capital and Coast*	5	1%	31.1 (0-85.5)

[1] Crude incidence rate presented due to small numbers of patients when stratified by age and CAU.