Is a rheumatic fever register the best surveillance tool to evaluate rheumatic fever control in the Auckland region?

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ABSTRACT

AIM: To determine the most accurate data source for acute rheumatic fever (ARF) epidemiology in the Auckland region.

METHOD: To assess coverage of the Auckland Regional Rheumatic Fever Register (ARRFR), (1998–2010) for children <15 years and resident in Auckland at the time of illness, register, hospitalisation and notification data were compared. A consistent definition was applied to determine definite and probable cases of ARF using clinical records. (www.heartfoundation.org.nz)

RESULTS: Of 559 confirmed (definite and probable) RF cases <15 years (median age 10 years), seven were recurrences. Of 552 first episodes, the ARRFR identified 548 (99%), hospitalisations identified 501 (91%) including four not on the register, and public health notifications identified 384 (70%). Of hospitalisation cases, 33% (245/746), and of notifications 20% (94/478) did not meet the case definition and were therefore excluded. Between 1998–2010, eight cases, initially entered as ARF on the ARRFR, were later removed once further clinical detail was available.

CONCLUSION: The ARRFR produced the most accurate information surrounding new cases of ARF (for children <15 years) for the years 1998–2010 in Auckland. This was significantly more accurate than medical officer of health notification and hospitalisation data.

Rheumatic heart disease (RHD), the long-term sequela of acute rheumatic fever (ARF), can persist for life.¹ Despite ARF being preventable, the associated morbidity and mortality continue to be a significant global burden falling largely on low-income countries.² However, it remains a significant issue in some indigenous and low-income communities in the industrialised world. The diagnosis of rheumatic fever is an estimate of probability using clinical and laboratory parameters, as there is no single diagnostic test. Internationally, the Jones criteria have been used with modifications made over time to improve specificity at the expense of sensitivity. New Zealand has led the way with the use of echocardiography to support the diagnosis.³ ⁴ A case definition with precise cut-offs for each criteria have been in place since the 1980’s with ongoing modifications⁵ (Heart Foundation of New Zealand guidelines www.heartfoundation.org.nz).

In New Zealand by the 197, ARF hospitalisation rates in children and young people had declined.⁶ However, the disease persisted and over the last 25 years, until the end of the study period 2010, national ARF hospitalisation rates had not improved.⁷ Most (80%) cases occur between 5–14 years of age, predominately in Māori and Pasifika and in lower socioeconomic areas.⁴ Rheumatic fever has been a disease legally notifiable to medical officers of health since 1986.⁸
In 2011, the New Zealand Government announced funding of a rheumatic fever prevention programme (RFPP), principally primary prevention of ARF by diagnosis and treatment of group A streptococcal pharyngitis in high-risk populations predominately in schools. A Better Public Services target was announced in 2012 to reduce rheumatic fever by two-thirds to 1.4 cases per 100,000 people by June 2017. Progress towards this target is measured using first episode ARF hospital admissions as the best currently available national estimate of the burden of ARF in New Zealand (http://www.health.govt.nz/about-ministry/what-we-do/strategic-direction/better-public-services/progress-better-public-services-rheumatic-fever-target), (http://www.health.govt.nz/our-work/diseases-and-conditions/rheumatic-fever).

A school-based intervention programme commencing in 2001 in Northland, aiming to reduce ARF rates, was implemented in high-risk communities in the North Island from 2010, including the Auckland region incrementally from 2011/2012. The intervention programme was based on the results of a randomised controlled trial of this alternative form of primary care access and a subsequent meta-analysis.9,10

Information from high-quality surveillance is essential to monitor a substantial government investment, comparing the pre-intervention (up to and including 2010) and the post-intervention phases. Currently within the Auckland region (where ~60% of New Zealand ARF occurs), there are three sources of RF surveillance data available.11 These are the Auckland Regional Rheumatic Fever Register (ARRFR) from 1981, hospitalisation data coded by non-medical staff using International Classification of Diseases (ICD) codes, and the national public health notification database from 1986.8 ARF notifications were previously recorded by the Auckland Regional Public Health Service (ARPHS) as a paper record and transferred to the national notifiable disease dataset at the Institute of Environmental Science and Research Limited (ESR). The electronic EpiSurv database for notifiable diseases was established in 2006.12 Data contributing to each rheumatic fever case in public health notifications during the period of the study (up to 2010) did not strictly adhere to the Heart Foundation of New Zealand guidelines, eg, erythrocyte sedimentation rate of a specific range, age-adjusted PR interval from electrocardiogram, etc. Thus, the application of the ARF case definition could lead to a different decision as to whether or not a case is truly ARF.

Referral of a patient by a clinician to a rheumatic fever register has healthcare advantages, if for example the referral process is appropriately linked to the delivery of penicillin prophylaxis. Building on the New Zealand experience, this has become a worldwide recommendation.8,13 The ARRFR was established to facilitate delivery of free penicillin prophylaxis in the Auckland region by delegated authority to rheumatic fever patients by community nurses to prevent recurrences and thus further cardiac sequelae.14 Referrals are predominately made by hospital doctors of both ARF and RHD if penicillin prophylaxis is recommended. Patients/caregivers consent to receiving penicillin by delegated authority (from the prescribing doctor to community nurses) and to allow their anonymised data to be used for prospective surveillance data. Referrals are entered on initial diagnosis of ARF or RHD to ensure prophylaxis once in the community. If information subsequently becomes available (likely to be within weeks) which changes the diagnosis, the record remains on the register but is marked as ‘not RF’.

Since 1981, each ARF case referred to the ARRFR has been reviewed against diagnostic criteria and deemed definite, probable or possible cases by a single infectious diseases specialist (DL)15 (www.heartfoundation.org.nz). Penicillin prophylaxis is delivered to all definite, probable and possible ARF cases, and RHD as requested. Possible cases are those from an at-risk population that fail to meet diagnostic criteria but are considered by a clinician to be highly likely to have ARF and deserving of penicillin prophylaxis.

Consented ARF cases are referred for nurse-delivered penicillin prophylaxis, dental care, and are notified to public health. Notifications to ARPHS are sent by fax or phone and from 2006 entered into EpiSurv, which is housed at ESR under contract to the Ministry of Health.

Prior to 2009, when household contact tracing was introduced, there was no
immediate public health action in the Auckland region. Thus, time to notification after data cleaning was variable. ARF cases not hospitalised (rare cases) come from general practice or out-patients for free nurse-delivered penicillin prophylaxis. ARF cases unknown to the register from general practitioners have been solicited at intervals with minimal response.9

Given the modest case numbers of ARF, accurate diagnosis and population-based surveillance are essential, as minor fluctuations in recorded case numbers can lead to significant changes in the overall rates. The ideal assessment tool provides the application of a consistent case definition over time to provide consistent epidemiology5 (New Zealand Heart Foundation guidelines www.heartfoundation.org.nz). Presented here is an analysis of the surveillance tools available for ARF and the pre-intervention epidemiology (1998–2010) for the Auckland region.

Methods

Register, hospitalisation and notification data were compared to assess the coverage of the ARRFR during 1998–2010 for children aged less than 15 years. This age group was selected to reflect the population served by children's hospitals and the greatest portion of the case load.16 The principal focus of the primary prevention intervention is school children enrolled in primary schools (5–12 year olds).

A consistent case definition was applied to all potential first and recurrent cases of ARF from each data source using electronic and paper records as required. The criteria used were the Jones criteria modified for the New Zealand context for this time period as defined by the New Zealand guideline for rheumatic fever diagnosis, management and prevention,17 (www.heartfoundation.org.nz) and categorised as definite, probable and possible (see Appendix 1). Cases that met the definition of definite and probable are included in this review. ‘Possible’ rheumatic fever cases were excluded as these cases represent the area of greatest ambiguity in the New Zealand RF classification system. At the time the data extract was obtained, the notification data at Auckland Regional Public Health could not identify cases of RHD (chronic disease without acute inflammation) separately from ARF and so these cases could not be excluded prior to the formation of the notification dataset. Furthermore, the hospitalisation and notification data could not be defined at first cut as definite, probable or possible cases, and accordingly the exclusion of ‘possible’ cases could not be applied prior to the formation of the datasets.

Auckland Regional Rheumatic Fever Register (Figure 1)

Definite and probable ARF cases (including recurrences) aged less than 15 years, from 1998 to 2010 (inclusive) were extracted for this study. Scrutiny by the register operator in conjunction with the referring clinician occurs on a case-by-case basis. New information for an alternative diagnosis is likely within weeks of referral. Such cases will cease penicillin prophylaxis and remain on the register as a non-case. Over the study period (n=13 years), eight cases (which did not meet the extraction criteria) were labelled as ‘not RF’ by this mechanism (see Figure 1 note). The ARRFR encourages clinicians to re-refer suspected ARF recurrences to the register to ensure continuing nurse-delivered prophylaxis.

Auckland hospitalisation data

Hospitalisations for the Auckland region (Auckland District Health Board (DHB), Waitematā DHB and Counties Manukau DHB) with a diagnosis of ARF were identified using the ICD-9 coding system codes 390–392 to identify cases diagnosed prior to January 2000 and the ICD-10 codes I00–I02 (I00, I01, I01.0, I01.1, I01.2, I01.8, I01.9, I02, I02.0, I02.9) to identify cases thereafter. Cases coded with principal and all level diagnoses of ARF were included. Day cases were included; New Zealand non-residents and hospital transfers were excluded. The ICD lists were then compared to the ARRFR listings. Cases with identical National Health Index numbers (NHI’s) which had admissions within four months of a known register event were merged and deemed to be the same event. This was considered appropriate given the time course of an ARF episode.18 Cases unknown to the ARRFR were reviewed using the case definitions (www.heartfoundation.org.nz).
Auckland notification data

In order to compensate for inconsistent recording of admission dates associated with notifications and variable delays in notifications following diagnosis, several data conditions were made. For cases known to the ARRFR, notifications within two years after the register admission date were deemed to have reported the same event (provided there was only one notification and one admission). This was because notifications previously occurred in batches (as there was no public health action associated with notification) and a delay of one to two years was not uncommon in the early stages of notification. If the time lapse was greater than two years, the electronic records for all three DHBs within the Auckland region were reviewed, looking for evidence of a different hospital admission or event (to that known by the register). However, if there was more than one notification for one known register event, the patient's electronic hospital records were also reviewed regardless of the time lapse. Notified cases, unknown to the register extract, then had the case definition applied. This included review for alternative NHIs and comparison to the register for RHD events. If a case was classified as RHD on the register and there was only one notification recorded, these were deemed to be related.

Incidence

Population denominators stratified by age (5–14 years), ethnicity and domicile at time of ARF event were determined using an Excel spreadsheet tool developed within Counties Manukau DHB (‘New Zealand 21 DHBs Estimated Resident Population by CAU, 1996–2026’, K Wang, personal communication), which is based on New Zealand census and population projection data. Exact Poisson 95% confidence intervals (95% CI) and Poisson regression lines were calculated using StatsDirect V3.0 (StatsDirect Ltd).

Notification to Auckland Regional Public Health Service (ARPHS) from ARRFR

An audit of the notification to ARPHS was also undertaken. A sample year 2010 was reviewed to audit the faxing of notifications to ARPHS, a process in place since 1998.

Results

A total of 559 confirmed (definite and probable) ARF cases less than 15 years (median age 10 years) from 1998–2010 were identified, including seven recurrences (Figure 1). Of the 552 first episodes, the ARRFR identified 548 (99%), hospitalisation data identified 501 (91%), including four not on the register, and the public health notification database identified 384 (70%). Recurrences approximated one case every two years (0.53 cases per year; seven recurrences in 13 years). All seven recurrences were identified on the ARRFR, five were also identified from hospitalisation data and four also from notification data. Three of the seven cases of ARF recurrences were noted to have primary cases of ARF within the hospitalisation data extract. Three other cases were likely to have had primary ARF episodes, which predated our extract, and one case had a primary ARF episode overseas. New primary ARF episodes found from other sources and unknown to the register were added.

Overall review (Figure 2)

On page 53 the three datasets are analysed in turn.

Auckland RF hospitalisations 1998–2010

Inclusions/exclusions

Of the 746 cases listed by the hospitalisation dataset, 501 cases met the stipulated criteria for definite or probable ARF and 245 cases were excluded (Figure 1). Twenty-five percent (62/245) of cases were matched to possible ARF cases known to the register and receiving penicillin prophylaxis. Eleven percent (27/245) represented duplications, i.e., more than one recorded hospitalisation for the same ARF episode. No documented record (i.e., no electronic or hard copy record could be identified for the reported event) could be located for five cases. The majority of the exclusions (151/245, 62%) were cases that did not meet the criteria and were not referred to the register for penicillin prophylaxis by a clinician. This subset included mostly cases where the diagnosis of ARF was considered but deemed to be very
Recurrences excluded from all data sources (n=7).
* Case definition, which was used on the register was applied. All potential cases from all data sources were tested against the case definition.
** Between 1998–2010 for the <15 year age group, eight cases which were initially entered as ARF on the ARFR were later labelled as ‘not ARF’ once further clinical detail (which excluded them as ARF) was available. This scrutiny occurs within weeks of initial entry to the register.
*** ‘Possible’ ARF cases were excluded (n=73).
unlikely or data was missing (97/151), RHD (20/151) and a set (34/151) were cases that had no direct association with rheumatic fever, ie, epistaxis, obstructive sleep apnoea, febrile seizures, adenotonsillectomy and counselling (some of these cases had a past history of rheumatic fever).

Accuracy of Auckland RF hospitalisation data 1998–2010 compared to the ARRFR (Figure 3)

The hospitalisation dataset identified 91% (501/552) first presentation cases and identified four cases that appeared to be unknown to the ARRFR. Nine percent (51/552) of the identified total ARF first presentations were not identified by the hospitalisation dataset as ARF, but had a verified admission date on the register. This dataset also comprised 33% (245/746) cases which were not new cases of ARF consistent with our case definition. This subset, even after excluding known possible ARF cases, contains a significant proportion which did not meet the case definition (ie, considered potential ARF but never confirmed) or were duplications (ie, patients with previous ARF events returning for follow up, had duplicate NHI codes or had previous diagnosis of ARF, which was then carried over in the ICD coding).

The four cases unknown to the ARRFR were individually reviewed. The first of these cases was known to the ARRFR from a later date and may have been receiving penicillin prophylaxis from another (but unable to be identified) source, ie, in primary care. Alternatively, this may represent an incorrectly recorded date in the ARRFR database. The second case had declined to proceed with intramuscular penicillin prophylaxis and so opted for oral penicillin prophylaxis through their general practitioner. The third case identified received penicillin prophylaxis while in hospital and then did not attend clinic appointments prior to moving out of the Auckland region, and the fourth was diagnosed in hospital very shortly before moving overseas. In the two cases moving out of region, arrangements were made for penicillin prophylaxis delivery to continue in their respective new areas.

Of the 746 hospitalisation records, 73 (10%) had a secondary diagnosis of ARF. Of the 73, six met the case definition and were known to the register, one was a true case unknown to the register and one was matched to a register ‘possible’ case. Five cases were RHD, 26 did not meet criteria for ARF (ie, unconfirmed cases), 23 were
admitted for other diagnoses, eight were duplications and three cases had no identifiable record. Accordingly, of the 501 definite and probable cases identified by the hospitalisation dataset, only seven had ARF coded as a secondary diagnosis.

**RF notifications to medical officers of health in the Auckland region 1998–2010**

**Inclusions/exclusions**

The notification dataset identified 478 cases; 384 cases identified represented definite or probable cases of ARF and 94 cases were excluded (Figure 1). Forty-nine (52%) of the excluded cases were matched to register possible cases and 37 cases (39%) were cases known to the register as RHD. Two other cases were excluded as they did not meet the criteria, and six cases represented duplications.

**Accuracy of RF notifications to medical officers of health in the Auckland region 1998–2010 compared to the ARRFR (Figure 4)**

The notification dataset identified 70% (384/548) of cases known to the register. All cases identified were known to the ARRFR. This dataset also included 20% (94/478) which did not meet the case definition. These were predominantly RHD cases. A significant proportion, 30% (164/548) of the confirmed ARF cases, were unknown to the notification dataset.

**Auckland Regional Rheumatic Fever Register extract**

The ARRFR extract identified 99.4% (548/552) of the total ARF cases (Figure 2).

**One year audit of faxing information to ARPHS/notification database from ARRFR (2010)**

All ARF cases referred to the ARRFR in the study period should have been notified to ARPHS by fax, as ARF is a notifiable disease. This process was audited for this review for 2010. Of the ‘missing’ cases from the notification dataset, 89% (8/9) had documented evidence of a faxed form for notification sent to ARPHS for the notification database. There was no such evidence for 11% (1/9). There was no process available to verify receipt by ARPHS of faxes.

**Incidence of rheumatic fever (ARF definite and probable including chorea and recurrences) in Auckland children aged 5–14 years, for the years 1998–2010**

The following results are based on combined data from the ARRFR, notification and hospitalisation data. There was an average of 42 cases per year aged 5–14

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**Figure 3:** Auckland hospitalisation data compared to register extract for first episode ARF, 1998–2010 for under 15 year age group. Note: The ‘Excluded’ area includes five cases for which no relevant clinical record could be found.
**Figure 4:** Notification dataset compared to register extract for first episode ARF, 1998–2010 for under 15 year age group.

**Figure 5:** Cases (ARF definite and probable and chorea and recurrences) aged 5–14 years in the Auckland region from 1998 to 2010.
years in the Auckland region from 1998 to 2010, at an overall incidence of 21.9/100,000/year (95% CI 20.1–23.8). The rates for Māori (45.8/100,000, 95% CI 39.7–52.5) and Pacific (66.6/100,000, 95% CI 59.5–74.4) compared to non-Māori/non-Pacific (1.7/100,000, 95% CI 1.1–2.5) had rate ratios of 27 (95% CI 18–42) and 39 (95% CI 26–60) respectively. ARF case numbers (Figure 5) increased over the study period. Incidence of ARF in Māori and Pacific (comprising 18 and 19% of the child population respectively) increased in incidence by 51 and 35% respectively, whereas the incidence in non-Māori/non-Pacific cases more than halved.

Discussion

Key findings and implications

The ARRFR has produced high-quality surveillance data for the pre-primary prevention period for ~60% of New Zealand 5–14 year-old ARF patients over 13 years in this study. Hospitalisation and notification data are not precise tools. Clinicians are motivated to report ARF cases to the register to ensure free penicillin prophylaxis, which is delivered in Auckland by community nurses historically not linked to ARPHS. Nurses network across the region to ensure efficient penicillin prophylaxis delivery across DHB boundaries, as domicile and school may not be in the same DHB. A uniformly applied case definition ensures high-quality data.

Mean annual national hospitalisation data (2000–2009) demonstrated an all-New Zealand rate for 5–14 year olds for Māori of 40.2/100,000 and Pacific of 81.2/100,000. ARRFR data found rates for Māori of 45.8/100,000 and Pacific of 66.6/100,000. With most Pacific children living in the Auckland region, the over-estimate by hospitalisation data could be explained by our study finding. Young Māori with ARF are scattered throughout the North Island; DHB ICD coding practices beyond Auckland were not a feature of this study. Data comparisons for Māori will await further investigations. However, there is no doubt ARF remains significantly disparate, with the disease burden carried almost exclusively by Māori or Pacific children and young people who make up approximately one-third of this age group in New Zealand. Such rates in European children are recorded from the 1920’s in Auckland.
Auckland hospitalisation dataset

When compared to the register extract, the hospitalisation dataset identified 91% of the confirmed ARF case load, but over-counted by including n=245 cases (245/746, 33%), which were not definite or probable ARF (http://www.health.govt.nz/system/files/documents/pages/first-episode-rheumatic-fever-hospitalisations-02-14-mar15.xlsx). The Ministry algorithm includes cases with an ARF-related ICD code as the principal diagnosis only, and excludes those with a previous diagnosis from 1998 onwards of ARF or RHD (as searched for case by case in our study) in any of their diagnostic codes. Given these differences, the Ministry algorithm improves accuracy for hospitalisation data, as it is more likely to exclude those with a previous ARF or RHD diagnosis. Under-counting by this method also occurs but is less of a concern. We detected seven confirmed (definite and probable) cases of ARF only coded as a secondary diagnosis.

Why were cases not identified by the Auckland hospitalisation dataset?

In addition, 9% of the total cases of ARF were not identified by the hospitalisation dataset. However, all cases known to the register had associated hospital admission dates recorded in the register database. Accordingly, this discrepancy is most likely attributable to incorrect coding. In some cases the diagnosis may have been suspected but not confirmed until further clinical review (eg, at outpatient clinic) was completed.

Why did cases identified from the Auckland hospitalisation dataset not meet the New Zealand RF criteria?

Coding of hospitalisations that do not meet ARF case definitions has been identified as an important issue. The ICD dataset identified cases of RHD as ARF (a coding error), unconfirmed cases of ARF (ie, ARF was part of the differential, however, the criteria were not met) as well as cases without direct association with ARF. If coding occurs promptly after discharge, missing results, eg, a repeat echocardiogram, may influence the final diagnosis. The cases excluded from the audit comprised 62%, which did not meet the set criteria. This percentage is comparable to the erroneous cases documented by Pennock et al in the Waikato review of all ages using a similar methodology (68%).

It is important to note that among the cases excluded from this audit, 25% were known to the register as possible rheumatic fever cases deemed worthy of penicillin prophylaxis. Currently, hospitalisation data does not allow for distinction between definite, probable and possible ARF. Other cases were excluded for not representing a separate ARF event, ie, double-ups of known ARFs who return to hospital for procedures, eg, a repeat echocardiogram or review. This is addressed by the MOH algorithm as above.

The concept of overestimation of the case numbers associated with hospitalisation data has previously been discussed by Oliver et al who also identified a sensitivity of 82%. As there were no chart reviews in the latter study, this is an estimate and does not take into account ARF cases hospitalised but not coded appropriately (highlighted as 9% in our review). This characteristic of dual inaccuracy (elements of over and underestimation of case numbers) of hospitalisation data was confirmed in our audit and also documented by Pennock et al.

Notification dataset

The notification dataset identified a significant number of non-cases (20%) and 30% of total ARF case load was not known to the notification database over the time of this study.

ARF became notifiable in 1986 after advocacy from the Auckland ARF clinicians to improve national data and raise awareness. Initial guidance was to include cases of RHD up to 20 years of age. Unlike most notifiable diseases where data is collected for action, in the absence of household contact tracing there was no ensuing action.

In the time period following this study, RHD has been explicitly excluded from the notification process, the capacity to capture possible ARF cases has been added and the ARF case definition updated to match the modified New Zealand Jones criteria 2015 (www.heartfoundation.org.nz). During our study period, as part of the notification process a yes/no question “does the case conform to the Jones criteria?” without a data dictionary was in place. ARF cases were...
not subsequently subject to audit to our knowledge, and therefore we suggest should be viewed with caution.

Why were cases not identified by the notification dataset?

The large number of cases missed by the notification dataset led to our audit of the referrals from ARRFR to ARPHS. The responsibility for notification to public health lies with the clinician responsible for the case. The ARRFR was set up (1981) before ARF became notifiable (1986). Clinicians who referred their patients to the ARRFR for penicillin prophylaxis were aware that registration with the ARRFR would lead to notification to ARPHS, who would then refer on to the national notification database (EpiSurv from 2006). This continued as accepted practice. The audit for 2010 showed that of the cases that were unknown to the notification dataset but known to the ARRFR, the significant majority had evidence of a faxed notification form to ARPHS (90%).

As there was no specific action, eg, contact tracing, by public health in the Auckland region as outlined above, it appears notifications did not necessarily get nationally notifiable. Thus, the notification data is incomplete. This has been rectified from the beginning of the primary prevention programme in 2011.

Why were there cases identified on the notification dataset that did not meet the criteria?

The notification dataset contained 20% of cases which did not meet the criteria. Of this selection, approximately half were possible ARF cases known to the register. The remaining cases were predominantly RHD cases. Until recently, notification guidance still included notification of cases of RHD aged less than 20 years; however, there was at the time of our study no means of distinguishing these from definite acute cases in the national database.

ARRFR

The prime purpose of the ARRFR, free nurse-delivered penicillin prophylaxis, has been highly successful in ensuring prevention of recurrences with high adherence. The first evaluation of the ARRFR revealed 22% of ARF cases on the ARRFR for 5–14 year olds were for recurrences with the higher likelihood of worsening heart disease. This has been reduced to 1% (7/555) in this current audit.

Limitations and considerations

Within the auditing process there were a number of limitations that were encountered.

The under 15 year age group was selected as a starting point to determine the most effective surveillance data source, as this would capture the majority of first presentation ARF cases. Further work on the over 15 year age group in the Auckland region is underway.

Currently there is no diagnostic test for ARF and so it remains a diagnosis of probability based on criteria specifically modified for the New Zealand context. A consistent case definition has been applied over time in Auckland in the ARRFR with appropriate updates. As far as possible, information was sought from case files to support or refute the diagnosis of ARF. There may be limitations in the data recorded and available on patient records, which could have prevented some cases from meeting the stipulated criteria. In a small number of cases, patients were awaiting laboratory tests for confirmation of diagnosis, which subsequently did not appear on record. Accordingly, these cases had to be excluded as they did not meet the case definition.

The incomplete recording of admission dates in the notification dataset created difficulty in associating notifications with specific admissions and/or register events. This led to data conditions being imposed as listed in the methodology. As notification on suspicion of a new suspected ARF case is now required by the Ministry of Health within seven days, these difficulties are likely to be less significant moving forward (http://www.nsfl.health.govt.nz/apps/nsfl.nsf/pagesmh/508). Each case is re-reviewed with emerging data to ensure each case is a true case.

A review of the ICD-coded cases of RHD could have potentially increased the ARF case load identified. The inaccuracy of coding raises the possibility that some cases of ARF may have been coded as RHD on the ICD coding system. These cases would not have been identified by the methodology.
implemented in this review. However, a review of RHD-coded ICD cases in the Tairāwhiti region for all ages by Moore et al found that the numbers of cases of ARF miscoded as RHD were minimal (3/122, 2%) over a three-year period.26

**Relationship to other research**

Oliver et al have suggested the use of the notification dataset as a basis for the implementation of a national register.27 The results of this audit conflict with that recommendation at least for the Auckland region for the time period (1998–2010) of our study. An appropriate and consistent definition (ie, the New Zealand RF criteria) had not been implemented at the time of notification and therefore historical case accuracy remains a significant issue. In 2014, the Ministry of Health reviewed and published updated notification guidelines, which were reflected in modifications in the national notification database, which might lead to higher quality of surveillance data going forward, although the consistent application of the appropriate case definition is not considered in a recent publication.28 Quality of the retrospective or baseline comparison data for the period prior to the government intervention will remain low unless considerable effort is made to rectify this.

Oliver and others used capture-recapture methodology to assess the accuracy of the currently available New Zealand ARF surveillance data.23 This approach has been used with many infectious diseases.29-31 The authors failed to appreciate the difference between their cited examples with clean cut diagnoses, mostly laboratory based, and the complexities of ARF/RHD diagnoses. In addition, allowing for the changing epidemiology by year of age the likelihood of ARF in a patient over 15 years of age is much more likely to be a recurrence of ARF and therefore unable to be prevented by the Government’s primary prevention strategy. In addition, they suggested that cases identified by the notification and hospitalisation datasets that are unknown to registers may indicate that register data may be incomplete. Within the Auckland region, (60% of New Zealand ARF cases), a review of these cases revealed that these ‘additional cases’ reflected significant inaccuracies of these databases, as a significant number did not meet the case definition even if including ARF cases deemed as possible cases are considered.

**Conclusions**

In the Auckland region this 1998–2010 audit prior to the primary prevention intervention demonstrates that the ARRFR provided the highest-quality data for monitoring ARF in the under 15 year age group. Notification and hospitalisation datasets were demonstrated to have significant inaccuracies and are not precise surveillance tools. Notification data for this time-period did not identify 30% of the ARF cases. Hospitalisation data included 33% of cases, which were not new cases of ARF in keeping with the case definition (and missed 9% of cases). Historical comparisons pre and post the intervention using these sources will be limited in their estimate of the effect. As ARF case numbers are relatively small, using the most accurate data source for evaluation is imperative.
Appendix

Table 1: New Zealand Rheumatic Fever Criteria adapted from Heart Foundation of New Zealand Guidelines.

<table>
<thead>
<tr>
<th>Diagnosis categories</th>
<th>Criteria</th>
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<tbody>
<tr>
<td>Definite ARF</td>
<td>Two major or one major and two minor manifestations plus evidence of a preceding GAS infection*</td>
</tr>
<tr>
<td>Probable ARF</td>
<td>One major and two minor with the inclusion of evidence of a preceding GAS infection* as a minor manifestation (Jones, 1956)³²</td>
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<tr>
<td>Possible ARF</td>
<td>Clinical suspicion of ARF, but insufficient signs and symptoms to fulfil diagnosis of definite or probable of ARF</td>
</tr>
<tr>
<td>Recurrence**</td>
<td>Two major or one major and two minor or several** minor plus evidence of a preceding GAS infection* (Jones, 1992)³³</td>
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**Criteria**

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<tr>
<th>Major</th>
<th>Carditis—including ECHO evidence alone***</th>
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<tr>
<td></td>
<td>Polyarthritisᵀ</td>
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<tr>
<td></td>
<td>Chorea</td>
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<td></td>
<td>Erythema marginatum</td>
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<td></td>
<td>Subcutaneous nodules</td>
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<tr>
<td>Minor</td>
<td>Fever ≥38°C</td>
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<td></td>
<td>ESR ≥50 CRP ≥50</td>
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<td></td>
<td>Polyarthralgiaᵀ</td>
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<td></td>
<td>Prolonged P-R interval on ECG: age-adjusted definitions#</td>
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</table>

Source: Lennon et al.¹ Note: ARF: acute rheumatic fever; CRP: C reactive protein; ECHO: echocardiogram; ESR: erythrocyte sedimentation rate; GAS: group A streptococcal.

* Elevated or rising antistreptolysin O or other streptococcal antibody is sufficient for a diagnosis of definite ARF. A positive throat culture or rapid antigen test for GAS alone is less secure, as 50% of those with a positive throat culture will be carriers only. Therefore, a positive culture alone demotes a case to probable or possible ARF.

** Most cases of recurrence fulfil the Jones criteria. However, in some cases (such as new carditis on previous RHD) it may not be clear. Therefore to avoid under-diagnosis, a presumptive diagnosis of rheumatic recurrence may be made where there are several minor manifestations and evidence of a preceding GAS infection in a person with a reliable history of previous ARF or established RHD.

*** Acceptance of ECHO evidence of carditis as a major criterion is a modification to the Jones (1992) update.

ᵀ Other causes of arthritis/arthralgia should be carefully excluded, particularly in the case of monoarthritis, eg, septic arthritis (including disseminated gonococcal infection), infective or reactive arthritis and autoimmune arthropathy (eg, juvenile chronic arthritis, inflammatory bowel disease, systemic lupus erythematosus, systemic vasculitis and sarcoidosis). Note that if polyarthritis is present as a major manifestation, polyarthralgia cannot be considered an additional minor manifestation in the same person.

# When carditis is present as a major manifestation (clinical and/or echocardiographic), a prolonged P-R interval cannot be considered an additional minor manifestation in the same person.
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