

# Acute rheumatic fever and rheumatic heart disease in the Kimberley: using hospitalisation data to find cases and describe trends

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As in other remote areas of Australia, Aboriginal people in the Kimberley region of north-west Western Australia suffer from unacceptably high rates of acute rheumatic fever (ARF) and rheumatic heart disease (RHD), with an estimated prevalence of at least 1.3%.<sup>1</sup> RHD contributes to cardiac valve damage, invasive heart surgery, heart failure and early death,<sup>2,3</sup> with devastating effects on children and young adults in their most productive years.<sup>4,5</sup>

Almost all cases of RHD and its associated morbidity and mortality are preventable.<sup>6</sup> Improved living conditions during the 20th Century dramatically reduced the burden of ARF in non-Aboriginal Australians to the point where it is now rarely seen.<sup>6,7</sup> The ultimate solution to ARF and RHD in Aboriginal communities is the primordial prevention of the social and environmental determinants of recurrent Group A Streptococcus (GAS) infection, particularly poverty and household crowding.<sup>8</sup> While prevention should remain the long-term objective, much can be achieved to reduce complications in the short to medium term, particularly by reducing the number of recurrences of ARF that an individual suffers.

ARF recurrences can be reduced with regular antibiotic prophylaxis to prevent reinfection with GAS.<sup>9,10</sup> This secondary prophylaxis is also associated with regression of heart disease<sup>11,12</sup> and has been shown to reduce the severity of RHD<sup>11,13</sup> and mortality.<sup>14</sup> This prophylaxis is usually delivered in the form of a four-weekly benzathine penicillin injection.<sup>6</sup> Register-based RHD control

## Abstract

**Objective:** To describe the epidemiology of hospitalisations due to acute rheumatic fever (ARF) and rheumatic heart disease (RHD) in the Kimberley region of Western Australia (WA) and use these data to improve completeness of the WA RHD Register.

**Methods:** Retrospective analysis of Kimberley regional hospitalisation data for hospitalisations coded as ARF/RHD from 01/07/2002 to 30/06/2012, with individual follow-up of those not on the register. Annual age-standardised hospitalisation rates were calculated to determine hospitalisation trend.

**Results:** There were 250 admissions among 193 individuals. Of these, 53 individuals (27%) with confirmed or probable ARF/RHD were not on the register. Males were less likely to be on the register (62% versus 79% of females,  $p < 0.01$ ), as were those hospitalised with ARF without heart involvement (68% versus 87% of other ARF diagnoses,  $p < 0.01$ ). ARF/RHD hospitalisation rates decreased by 8.8% per year ( $p < 0.001$ , rate ratio = 0.91, 95%CI 0.87–0.96).

**Conclusions and implications:** Using hospitalisation data is an effective method of identifying cases of ARF/RHD not currently on the register. This process could be undertaken for initial case finding in areas with newly established registers, or as regular quality assurance in areas with established register-based programs. Reasons for the observed decrease in hospitalisation rates remain unclear and warrant further investigation.

**Key words:** Acute rheumatic fever, rheumatic heart disease, Aboriginal health

programs are designed to improve delivery of this prophylaxis by assisting health care providers to record administration and recall individuals who have missed doses.<sup>15</sup> These programs also assist with the co-ordination of ongoing care and provide valuable epidemiological information. Best practice guidelines recommend offering all patients with a history of ARF/RHD the opportunity to register with a RHD control program.<sup>16</sup> The effectiveness of such a program depends on the accuracy of its database, how well it is maintained and how well the information is disseminated.<sup>17</sup>

A register-based RHD control program was established in Western Australia (WA) in 2009, and new and recurrent cases of ARF have been notifiable to the Western Australian Notifiable Infectious Diseases Database (WANIDD) since 2008.<sup>18</sup> The register currently uses a passive case finding technique that encourages clinicians to consent all patients with confirmed or probable ARF/RHD to the register. As of March 2013, this process had resulted in 516 individuals being added to the register, 70% (364) of whom were from the Kimberley (unpublished data from the WA RHD control program).

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Given the limitations of passive case finding, it is likely that the register is not a complete representation of all people with ARF/RHD in the Kimberley. An audit of a remote Northern Territory community found that 19% of eligible patients were not on the Territory-wide register seven years after its inception.<sup>17</sup> One of the central requirements for a good RHD control program is an effective method for finding new cases.<sup>15,19</sup> Current guidelines recommend that patients with ARF are admitted for investigation and management<sup>6</sup> and most patients diagnosed with ARF/RHD in WA are hospitalised at some point during their care. Hospitalisation data are therefore a potentially useful source for active case finding for the RHD Register.

In addition, hospitalisation data can provide information on trends in incidence of ARF/RHD. According to WANIDD notification data, there has been a steady increase in ARF notifications in WA since 2009,<sup>18</sup> including in the Kimberley: 1 in 2009, 7 in 2010, 18 in 2011 and 43 in 2012 (email communication from Dr Carole Reeve, FAFPHM, Kimberley Population Health Unit, January 2013). It is unclear whether this increase is due to increased practitioner awareness and education or represents a true increase in disease.

The objectives of this study were twofold. First, to use hospitalisation data to actively find cases for the WA RHD Register and, second, to describe the epidemiology of hospitalisations due to ARF and RHD in Kimberley hospitals over a 10-year period.

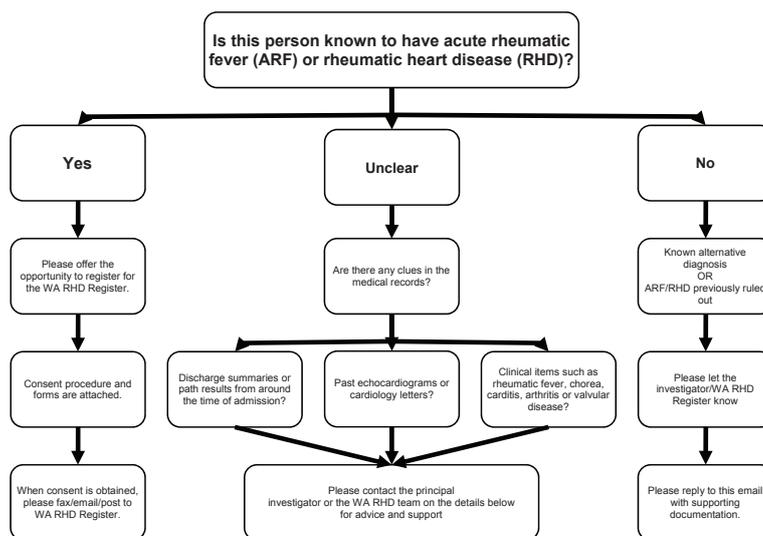
### Methods

The WA Country Health Service provided hospitalisation data from all six Kimberley hospitals from 1 July 2002 to 30 June 2012. Hospitalisations that had been assigned an ARF/RHD International Classification of Diseases version 10 (ICD-10) code for the primary diagnosis were included (see Table 1).

Variables extracted were: primary diagnosis, sex, Aboriginality, hospital record number and discharge date. For each hospitalisation, the unique medical record number (UMRN), patient name, primary care provider and deceased status were obtained from the regional admissions database (HCARE Kimberley) and manually added to the data set.

Data cleaning checked for missing variables, duplicates and anomalies. If an individual had two admissions with the same ICD code within a 3-month period, the second admission was excluded.

Figure 1: Instructions sent to primary care providers of individuals identified for follow up.



### Active case finding

Hospitalisation data were manually cross-referenced with the WA RHD Register using UMRN. The hospital separation diagnoses of those individuals who were already on the WA RHD Register were not further examined, their diagnoses having been previously confirmed as part of the notification and consent process of the WA RHD Register.

Deceased individuals were excluded from follow-up. Individuals who were not already on the register and who were not recorded as deceased were identified for follow-up.

Their primary care providers were initially contacted by telephone. An email followed with instructions on how to proceed (see Figure 1). If the individual was known by the primary care provider to have had ARF, the primary care provider was instructed to follow WA RHD’s usual procedures for confirmation of diagnosis using the 2012 Updated Australian Guidelines for the Diagnosis of ARF (see Table 2) and consent to the register. If the individual was known by the primary care provider to have RHD (documented diagnosis in patient’s clinical records), the researchers

Table 1: Included acute rheumatic fever and rheumatic heart disease ICD codes .

ICD code	ICD description
<b>Acute rheumatic fever codes</b>	
I00.	Rheumatic fever without mention of heart involvement
I01.0	Acute rheumatic pericarditis
I01.1	Acute rheumatic endocarditis
I01.2	Acute rheumatic myocarditis
I01.8	Other acute rheumatic heart disease
I02.0	Rheumatic chorea with heart involvement
I02.9	Rheumatic chorea without heart involvement
<b>Rheumatic heart disease codes</b>	
I05.1	Rheumatic mitral insufficiency
I06.0	Rheumatic aortic stenosis
I06.1	Rheumatic aortic insufficiency
I06.2	Rheumatic aortic stenosis with insufficiency
I06.9	Rheumatic aortic valve disease
I07.0	Tricuspid stenosis
I07.1	Tricuspid insufficiency
I09.0	Rheumatic myocarditis
I09.1	Rheumatic disease endocardium unspecified
I09.2	Chronic rheumatic pericarditis
I09.8	Other specified rheumatic heart disease
I09.9	Rheumatic heart disease unspecified

accepted a statement from the primary care provider and asked them to consent the individual to the register.

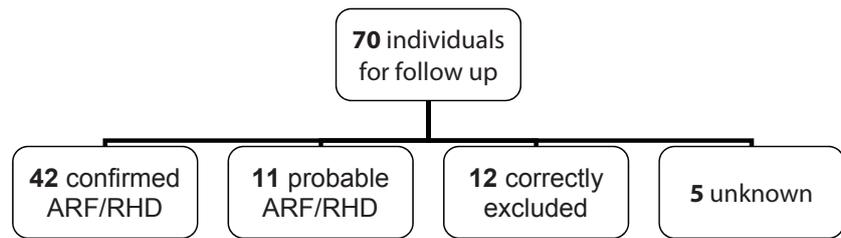
If an individual's diagnosis was unclear to the primary care provider, the researchers offered supported examination of the primary care records to determine if there was any evidence of ARF/RHD. Evidence of ARF was taken as an episode that met the criteria for diagnosis according to the 2012 Updated Australian Guidelines (see Table 2). These were divided into 'definite' and 'probable' cases as per the guidelines. Evidence for RHD was taken as documentation of RHD as a clinical problem, past history of receiving ongoing benzathine penicillin prophylaxis or evidence of RHD on echocardiogram report. Those individuals who had a clear alternative diagnosis (such as syphilitic aortitis) and no documented history of RHD were excluded. If the diagnosis remained unclear following supported examination of the primary care records, the hospitalisation records were requested from the relevant hospital and the study team reviewed these with the primary care provider using the same inclusion and exclusion criteria. If the diagnosis remained unclear, the case was judged 'unknown'.

### Analysis

Statistical analyses were conducted using Epi Info™ version 7 and Microsoft Excel™; for calculation of standardised rates and trend analysis, Rates Calculator version 9.5.5 (Epidemiology Branch, WA Department of Health) was used. Descriptive analyses of hospitalisation data were conducted by place, person and time. Patients on the register and not on the register were compared by demographic characteristics, diagnosis and time period. The two-sided chi-square test was used to compare categorical variables with values considered significant at the 0.05 level. Population data were extracted from the Rates Calculator; this program uses Estimated Resident Populations developed by the Australian Bureau of Statistics for the census year 2006, and estimates derived using Aboriginal birth and death data for non-census years and 2011. The Australian 2001 ERP was used for the calculation of age-standardised hospitalisation rates by means of direct standardisation. Poisson regression was used for trend analysis.

Ethics approval for this study was granted by WA Country Health Service Research Ethics Committee and the Western Australian Aboriginal Health Ethics Committee.

Figure 2: Outcomes of follow up of patients not on WA RHD Register.



It had the support of the Kimberley Aboriginal Health Planning Forum Research Subcommittee.

### Results

#### Active case finding

From 1 July 2002 to 30 June 2012 there were 296 hospitalisations among 233 individuals. Twenty-three individuals were known to be deceased and 140 were already included on the WA RHD Register, with 70 people remaining for follow-up.

After follow-up with primary care provider and hospital admission notes, 42 (60%)

individuals were confirmed to have ARF/RHD and 11 (16%) were probable cases that had not been further investigated (Figure 2). Twelve people (17%) were found to have been correctly excluded from the register; five were initially thought to be cases but following further investigation were found not to meet the diagnostic criteria, and seven had been incorrectly assigned the ICD code. These seven had no evidence of ARF/RHD and had confirmed alternative diagnoses (rheumatic polyarthritis, perimembranous septal defect, alcoholic cardiomyopathy, syphilitic aortitis, juvenile arthritis, ischaemic heart disease, pulmonary hypertension).

Table 2: 2012 Updated Australian guidelines for the diagnosis of ARF.<sup>6</sup>

	High-risk groups <sup>a</sup>	All other groups
Definite initial episode of ARF	2 major or 1 major and 2 minor manifestations plus evidence of a preceding GAS infection <sup>a</sup>	
Definite recurrent episode of ARF in a patient with known past ARF or RHD	2 major or 1 major and 1 minor or 3 minor manifestations plus evidence of a preceding GAS infection <sup>b</sup>	
Probable ARF (first episode or recurrence)	A clinical presentation that falls short by either one major or one minor manifestation, or the absence of streptococcal serology results, but one in which ARF is considered the most likely diagnosis. Such cases should be further categorised according to the level of confidence with which the diagnosis is made:	
	<ul style="list-style-type: none"> <li>highly-suspected ARF</li> <li>uncertain ARF</li> </ul>	
Major manifestations	Carditis (including subclinical evidence of rheumatic valvulitis on echocardiogram) Polyarthritis <sup>c</sup> or aseptic mono-arthritis or polyarthralgia Chorea <sup>d</sup> Erythema marginatum <sup>e</sup> Subcutaneous nodules	Carditis (excluding subclinical evidence of rheumatic valvulitis on echocardiogram) Polyarthritis <sup>c</sup> Chorea <sup>d</sup> Erythema marginatum <sup>e</sup> Subcutaneous nodules
Minor manifestations	Monoarthralgia Fever <sup>f</sup> ESR $\geq$ 30 mm/h or CRP $\geq$ 30 mg/L Prolonged P-R interval on ECG <sup>g</sup>	Fever <sup>f</sup> Polyarthralgia or aseptic mono-arthritis ESR $\geq$ 30 mm/h or CRP $\geq$ 30 mg/L Prolonged P-R interval on ECG <sup>g</sup>

a High-risk groups are those living in communities with high rates of ARF (incidence  $>30/100,000$  per year in 5–14 year olds) or RHD (all-age prevalence  $>2/1000$ ). Aboriginal people and Torres Strait Islanders living in rural or remote settings are known to be at high risk. Data are not available for other populations, but Aboriginal people and Torres Strait Islanders living in urban settings, Maoris and Pacific Islanders, and potentially immigrants from developing countries, may also be at high risk.

b Elevated or rising antistreptolysin O or other streptococcal antibody, or a positive throat culture or rapid antigen test for GAS.

c A definite history of arthritis is sufficient to satisfy this manifestation. Note that if polyarthritis is present as a major manifestation, polyarthralgia or aseptic mono-arthritis cannot be considered an additional minor manifestation in the same person.

d Chorea does not require other manifestations or evidence of preceding GAS infection, provided other causes of chorea are excluded.

e Care should be taken not to label other rashes, particularly non-specific viral exanthemas, as erythema marginatum.

f Oral, tympanic or rectal temperature  $\geq 38^{\circ}\text{C}$  on admission, or a reliably reported fever documented during the current illness.

g If carditis is present as a major manifestation, a prolonged P-R interval cannot be considered an additional minor manifestation.

CRP, C-reactive protein; ECG, electrocardiogram; ESR, erythrocyte sedimentation rate.

The diagnoses for five people (7%) were not determined as researchers could not locate the individuals and no hospital notes were available. No individual had refused the opportunity to consent to the register.

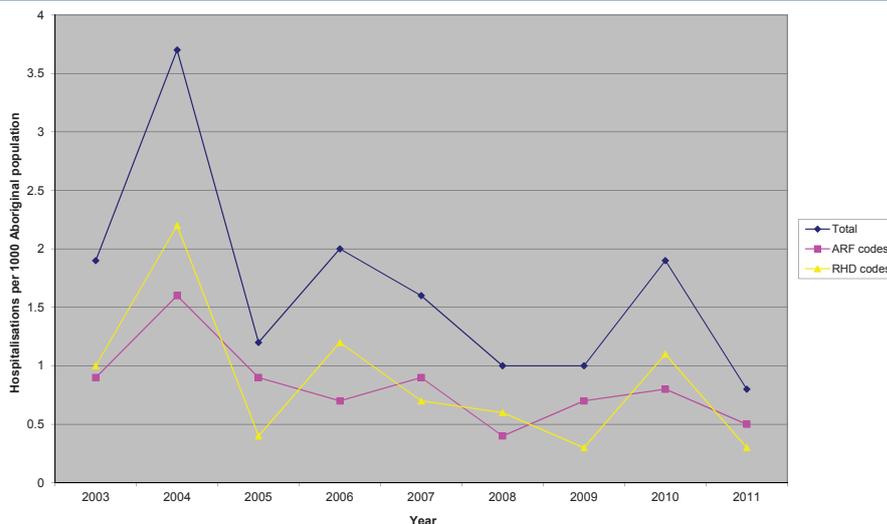
Locations were not able to be established for 19 (36%) of the 53 individuals who had confirmed or probable ARF/RHD and were not on the register. Of the 53, only 24 (45%) were known to have ARF/RHD by their primary care provider.

**Analysis**

One hundred and ninety-three people with confirmed or probable ARF/RHD (who were living at the time of analysis) were hospitalised a total of 250 times over the 10-year period of the study. The majority of these individuals (119, 62%) were female and all except one was Aboriginal. The median age of those hospitalised was 24 years (interquartile range: 15-34). Sixty per cent of the hospitalisations (155/250) were for an ARF-related diagnosis, representing an initial or recurrent episode of acute rheumatic fever.

There was a decrease of 8.8% per year in the age-standardised rate (ASR) of

**Figure 3: Age standardised rates of hospitalisations for ARF/RHD in the Kimberley by ICD code.**



hospitalisations for confirmed and probable ARF/RHD over the nine calendar years in the study period (see Figure 3). This was a statistically significant decrease ( $p < 0.001$ , rate ratio = 0.91, 95%CI 0.87–0.96). The decrease was sustained across both ARF diagnoses and RHD diagnoses.

Factors associated with being on the register are shown in Table 3. Males were significantly

less likely to be on the register after being hospitalised for ARF or RHD. Specific ICD codes made it more likely that an individual was on the register; almost all of those hospitalised with an ICD code that indicated rheumatic fever with heart involvement or rheumatic chorea were on the register (88% and 93% respectively). Those individuals with an admission coded as rheumatic fever

**Table 3: Epidemiology of individuals hospitalised for ARF/RHD in the Kimberley.**

	On register			Not on register			p-value	Total No.
	No.	%	95% CI	No.	%	95% CI		
<b>Total patients</b>	142	61%		91	39%			233
<b>Living patients<sup>a</sup></b>	140	67%		70	33%			210
<b>Living patients with confirmed/probable ARF/RHD<sup>b</sup></b>	140	73%	66%-79%	53	27%	21%-34%		193
Female	94	79%	72%-86%	25	21%	14%-28%	$p=0.006$	119
Aboriginal	140	73%	67%-79%	52	27%	21%-33%	$p=0.137$	192
<b>Age<sup>c</sup></b>								
0-14	42	74%	62%-85%	15	26%	15%-38%	$p=0.414$	57
15-29	64	75%	66%-85%	21	25%	15%-34%	$p=0.227$	85
30-44	22	63%	47%-79%	13	37%	21%-53%	$p=0.060$	35
45+	12	75%	53%-97%	4	25%	3%-47%	$p=0.467$	16
<b>ICD code grouping<sup>c</sup></b>								
I00 Rheumatic fever without heart involvement	54	68%	58%-79%	25	32%	21%-42%	$p=0.010$	79
I01 Rheumatic fever with heart involvement	29	85%	73%-97%	5	15%	2%-27%	$p=0.056$	34
I02 Rheumatic chorea	11	92%	75%-108%	1	8%	-8%-25%	$p=0.088$	12
I05-I09 Rheumatic heart disease	46	68%	56%-79%	22	32%	21%-44%	$p=0.134$	68
<b>Number of admissions during study period</b>								
1 admission	106	69%	62%-77%	47	31%	23%-38%	$p=0.022$	153
>1 admission	34	85%	74%-96%	6	15%	4%-26%		40
<b>Year of discharge<sup>d</sup></b>								
Before RHD Register establishment	80	67%	57%-74%	42	33%	26%-43%		122
After RHD Register establishment	60	86%	76%-93%	11	14%	7%-24%	$p=0.002$	71

*a* includes patients who were confirmed to be living as at 31 March 2013

*b* includes patients who were confirmed to be living as at 31 March 2013 and who were found to have confirmed or probable ARF/RHD after follow up

*c* By first hospitalisation in study period

*d* By last hospitalisation in study period

without heart involvement, or with one of the rheumatic heart disease codes (e.g. valvular disease), were less likely to be on the register (69% and 68% respectively). Almost all (86%) of the patients who had their last admission after the establishment of the WA RHD Register in 2009 or who were admitted more than once for ARF/RHD were on the register (86% and 85% respectively). There were no differences seen by either patient address or hospital location as to whether the patient was on the register.

## Discussion

This study found that using hospitalisation data is effective at finding cases of ARF/RHD not currently included on the WA RHD Register. The process of consent for those patients whose whereabouts are known to the register is straightforward and can be facilitated by the WA RHD Control Program team. Given that this study found that no patients had refused consent when offered the option to be added to the WA RHD Register and that register-based control programs have been shown to increase awareness of and adherence to secondary prophylaxis,<sup>20,22</sup> it is hoped that this study will ultimately contribute to a decrease in ARF recurrences in the Kimberley.

There are plans to repeat this process periodically in the Kimberley. It is clear that there are a number of patients who are being lost to follow-up between hospitalisation and return to primary care. The Kimberley is particularly vulnerable to this due to the high turnover of staff and the transient place of residence of many of the region's patients.<sup>23</sup>

This method can be easily replicated in other regions and there are plans to repeat the study in the other high prevalence regions of WA. Additionally, this process could be undertaken by RHD control programs in other Australian and New Zealand jurisdictions as a quality assurance exercise to review the completeness of their registers and ensure patients are not being lost to follow-up.

There are two main limitations to this case finding method. First, the researchers were unable to locate a number of the identified patients. This was primarily due to time and staff limitations, however, the WA RHD Control Program plans to dedicate resources to continue the tracing and follow-up of these individuals. Second, this study will not have identified all of those eligible to be on the WA RHD Register in the Kimberley. The study

only reviewed hospitalisation data. Although current Australian guidelines recommend that all patients who have an episode of ARF be hospitalised,<sup>6</sup> some individuals who had subclinical presentations, who were clinically very stable or who were misdiagnosed would not have been hospitalised and therefore would not be found through this method. The study also only covered 10 years of data. This period was selected in order to find patients who are most at risk of missing out on secondary prophylaxis. Current guidelines recommend all persons with ARF/RHD should continue secondary prophylaxis for a minimum of 10 years after their last episode of ARF.<sup>6</sup> Patients who did not have an episode of ARF between 2002 and 2012 or who were clinically stable may not have been hospitalised during the study period and therefore would not be included. There may be some benefit to extending the study period further into the past when the process is repeated.

Additionally, this study excluded deceased patients from the epidemiological analysis, which will lead to a slight under-estimate of hospitalisations. The vast majority of the deceased patients were not on the register. As the primary aim of the study was to identify individuals eligible to be on the register, resources were not devoted to following up deceased patients to confirm their diagnoses.

This study has identified that certain groups are at risk of not being consented to the RHD Register. The literature reports that males in Aboriginal communities have lower rates of uptake of secondary prophylaxis than females.<sup>24</sup> To our knowledge, this study is the first to report that they are also significantly less likely than females to be included on a register; however, this may not be generalisable to other jurisdictions' registers that routinely use active case finding techniques. Other at risk groups include patients who were hospitalised with ARF without heart involvement or with RHD, and patients who were last hospitalised prior to the implementation of the WA RHD Register in 2009. This project provides support for further education of primary care providers and hospital staff around the benefits of being on the register and the groups who are particularly vulnerable to being overlooked.

Given the increase in notifications to WANIDD for ARF since 2007 (email communication from Dr Carole Reeve, FAFPHM, Kimberley Population Health Unit, January 2013), the decrease in the rate of hospitalisations

for ARF/RHD was unexpected. Moreover, a recent examination of the RHD control program database in neighbouring Northern Territory did not find a decrease in ARF/RHD incidence.<sup>3</sup> It is unclear why this decrease in hospitalisations in the Kimberley occurred. Optimal secondary prophylaxis coverage in primary care settings would lead to an expected decrease in hospitalisations; however, a 2007 audit of ARF/RHD management in the region reported that only 14.7% of individuals received  $\geq 80\%$  of recommended doses.<sup>1</sup> Other possible explanations include more accurate population data over time,<sup>25</sup> greater awareness of notification processes among clinicians due to the implementation of the RHD Register,<sup>26</sup> changes in admissions processes or a true decrease in disease in at risk children.

It should also be acknowledged that this dataset contains all hospitalisations and not just first hospitalisations, and so may lack comparability with ARF notifications data. Nevertheless, the observed decrease in hospitalisation rates is reassuring and further investigation into its true cause is warranted.

Register-based RHD control programs remain the mainstay intervention for controlling and preventing recurrent ARF and subsequent RHD in high incidence settings, however, they can only be effective if they have accurate and complete information. This study demonstrates an effective process for identifying cases that are not currently on the register, and could be undertaken as an initial case finding exercise in areas with newly established registers, or as a regular quality assurance process in areas with established register-based programs. Until the primordial causes of Aboriginal and Torres Strait Islander health inequality are addressed, primary and secondary prevention programs such as RHD control programs offer the best chance at closing the gap between Aboriginal and non-Aboriginal health outcomes.

We acknowledge the contribution of the Aboriginal people of the Kimberley region to this project. We acknowledge the staff working in primary health across the Kimberley for their assistance with locating patients and records and for their ongoing clinical care of the patients involved. Thanks to the staff of the WA RHD Control Program for their follow-up of individuals who have not yet been located. Thanks also to Vicki Kern for her assistance with data extraction and to the medical records staff at Broome, Derby,

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