The cost of inaction on rheumatic heart disease: technical report

The predicted human and economic impact of rheumatic heart disease for Aboriginal and Torres Strait Islander people until 2031.

Analysis team

Jeffrey Cannon (health economist)
Judith Katzenellenbogen (epidemiologist)
Melanie Greenland (biostatistician)
Rebecca Cunneen (biostatistician)
Daniela Bond-Smith (biostatistician)

Expert advisers

Jonathan Carapetis
Lee Nedkoff
Nick de Klerk
Frank Sanfilippo
Rosemary Wyber

Design and supprot

Caroline Wise
Kate Harford

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end.rhd.cre@telethonkids.org.au

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Executive summary

- Infection with the bacterium group A *Streptococcus* ("Strep A") can cause an abnormal immune reaction called acute rheumatic fever (ARF). A severe episode of ARF or repeated Strep A infections that results in ARF recurrences can lead to damaged heart valves, known as rheumatic heart disease (RHD).

- In Australia, 94% of new ARF cases occur among Aboriginal or Torres Strait Islander people.[1]

- This report uses linked hospital admissions and mortality data for Aboriginal or Torres Strait Islander people over the last two decades to analyse the prevalence and incidence of ARF and RHD in the four Australian jurisdictions with the highest recorded burden of disease: Western Australia, South Australia, Northern Territory and Queensland. These analyses are used to determine the number of active cases at mid-2016 and to project the number of cases to occur between mid-2016 and 2031. Subsequently, modelling was used to estimate disease progression and associated medical care and cost for both the active and projected cases.

- The results indicate that:
    - 4,539 Aboriginal and Torres Strait Islander people who were alive at June 30th, 2016 had been hospitalised for ARF and/or RHD between 2001 and 2016. Forty percent of those people were under 25 years of age in 2016, and 3,420 people had been hospitalised for the diseases within the last 10 years and are therefore recommended to receive medical care. Of the people who had been hospitalised within the 10 years prior to mid-2016.
    - 110 people with RHD will die and;
    - $27 million will be spent to complete their recommended course of medical care.

- 10,212 Aboriginal and Torres Strait Islander people will develop ARF or RHD between mid-2016 and 2031 if the trend in incidence rates of ARF and RHD observed over the last several years continues. Of these people:
    - 1,370 people will need heart surgery for RHD,
    - 518 people with RHD will die, and
    - $317 million will be spent on medical care.

- This suffering is preventable; unlike the first half of the 20th century, ARF is almost unheard of in non-Indigenous Australian children in the present era. Deaths from RHD can be avoided, financial costs can be reduced, and new cases of disease prevented. This report outlines the trajectory of RHD if Australia maintains only the current levels of funding and attention to ARF and RHD.

Introduction

Strep A, ARF and RHD

People are exposed to the bacterium Group A *Streptococcus* ('Strep A') through contact with other people. Enough exposure, and susceptibility, to Strep A can result in an infection of the throat or skin, and either of those infections can lead to an abnormal immune response known as acute rheumatic fever (ARF – see Figure 1). Symptoms of ARF include sore joints, rashes, abnormal movements, fever, and heart inflammation. Most of these symptoms resolve over a few weeks, but heart damage often remains. Repeated Strep A infections can cause recurrent ARF and permanent scarring of the heart valves. Damage to the heart valves is called rheumatic heart disease (RHD).

Strep A infections among Aboriginal and Torres Strait Islander people usually begin in childhood. On average, Aboriginal children in remote communities have their first skin infection, which is predominantly caused by Strep A, at seven months of age.[2] Strep A skin infections continue throughout childhood and into adolescence. Strep A throat infections begin in later childhood, peaking between 5-14 years of age.[3]

The incidence of ARF episodes also peaks at between five and 14 years of age and then steadily declines with increasing age, whereas the prevalence of RHD steadily increases with age. The age distribution of ARF and RHD is illustrated in Figure 2. The median age at death of Aboriginal people dying with RHD in the Kimberley region is 41 years.[4]
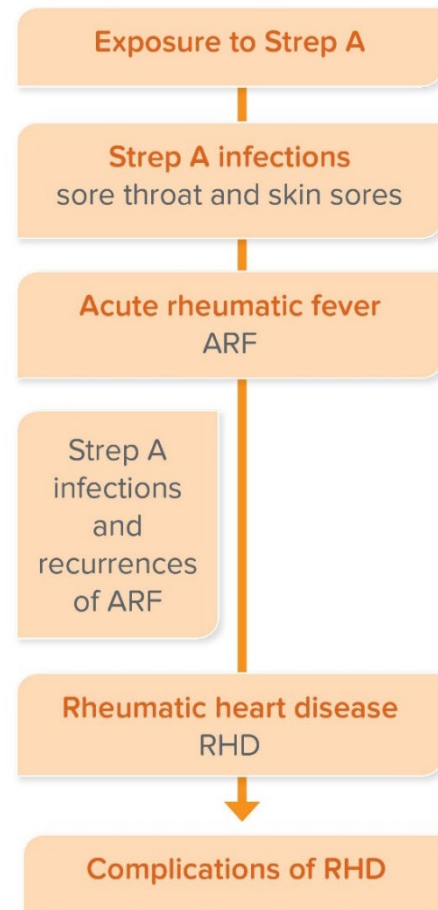
Figure 1: Causal pathway from Strep A to RHD
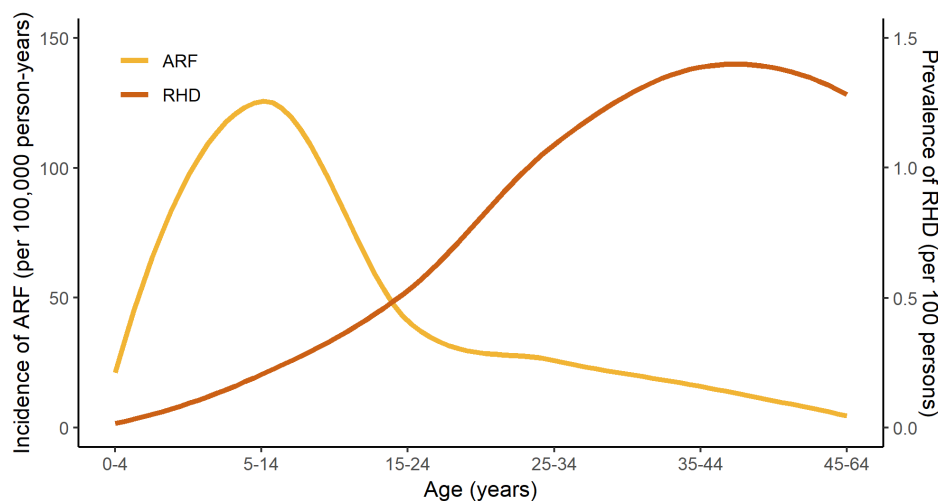
Figure 2: Incidence of ARF between 2014 and 2016 and prevalence of RHD in mid-2016, the NT, QLD, SA and WA (preliminary analysis of data from NHMRC PG 1146525)

Current clinical action

There are many ways to intervene on the causal pathway from the exposure to Strep A to the development of RHD and associated complications. A summary of these is presented in Table 1.

Table 1: Overview of prevention and management opportunities for acute rheumatic fever (ARF) and rheumatic heart disease (RHD).

Point in the disease pathway	Prevention opportunities
Strep A exposure	Improve social determinants of health (primordial prevention): • Activities to address the underlying causes of RHD such as crowded living environments, poor health-hygiene infrastructure, and barriers to seeking and accessing healthcare. By reducing the exposure of children to environmental and social risk factors, infection can be prevented, or at least limited, and the probability of developing ARF and RHD can be reduced.
Strep A infection Skin and throat infection	Primary prevention: • Prompt diagnosis and treatment of Strep A throat and skin infections to prevent the development of ARF. The risk of ARF following Strep A throat infection is reduced by 80% with antibiotic treatment.[5]
Acute rheumatic fever (ARF)	Secondary prevention: • The diagnosis and management of ARF through the delivery of regular antibiotic injections, known as secondary prophylaxis, to prevent recurrences of ARF and possible RHD disease progression. The recommended antibiotic is penicillin, which is administered by intramuscular injection every 28 days for at least decade after the first episode of ARF.
Rheumatic heart disease (RHD)	Tertiary intervention: • Advanced medical care, including cardiac surgery, to prevent people with RHD suffering further health complications. Open heart surgery in people with advanced RHD can prolong life expectancy and improve symptoms, but it is not a cure, a lifetime of medical care is still needed. These includes medication to manage heart failure and anticoagulation treatment to reduce the risk of stroke. Women with RHD require particularly close monitoring and specialist support during pregnancy due to the additional stress placed on the heart.

The current clinical action and healthcare investment towards tackling ARF and RHD in Australia has focused on secondary prevention (namely improving adherence to a secondary prophylaxis schedule) and tertiary intervention. This means preventing recurrent ARF and death due to RHD, rather than preventing and treating the Strep A infections which cause ARF and RHD.

The Australian Government has invested in the Rheumatic Fever Strategy since 2009.[6] The Rheumatic Fever Strategy has focused on secondary prophylaxis through:

1. Improved detection, monitoring, and management of ARF and RHD via State-based registers and control programs, which exist in five jurisdictions; and

2. Developing education and training resources, supporting the register and control programs via a national coordinating body (RHDAustralia).

The delivery of secondary prophylaxis is predominately provided by primary care services. Although high-quality primary care services exist in Australia, Aboriginal and Torres Strait Islander people experience barriers to

accessing primary care, as well as hospital and specialist services.[7,8] These barriers contribute to devastating outcomes:

- Studies in the Northern Territory have demonstrated that RHD causes an increased mortality rate compared to the general rate, and that the risk of mortality is multiple times higher for Aboriginal and Torres Strait Islander people with RHD compared to non-Indigenous people.[9,10]

- One study in the Northern Territory found that more than one in ten (13%) children and young adults aged five to 24 years at diagnosis died within ten years of severe RHD diagnosis.[11]

An independent, external review of the Commonwealth Government's Rheumatic Fever Strategy recommended that, in addition to the focus on secondary prevention, the strategy broaden its mandate to also address social and environmental determinants of health and primary prevention measures.[6] Such a broadening of focus would prevent ARF happening in the first place.

The cost of inaction

Eliminating RHD means preventing new cases of disease. There is a moral imperative to provide both treatment and prevention of RHD. In particular, Australia has an obligation to address RHD as the leading cause of cardiovascular disparity between Indigenous and non-Indigenous people.[12] A bipartisan political commitment to eliminate RHD has already been made.[13]

Delivering on this commitment means understanding the resources needed to deliver best-practice medical care and to address the underlying inequities that increase the risk of ARF and RHD. This report is the starting point of that process. It outlines the methods used to estimate;

- The clinical trajectory of Aboriginal and Torres Strait Islander people who have ARF and RHD today and those who can be expected to develop it in the future and

- The resulting human and financial costs in terms of death and medical care, respectively.

The suffering caused by RHD to individuals, families, and communities is preventable. The human and economic toll could be avoided by urgent investment by Australian governments in collaborative, community-led strategies to address the environmental and socio-economic causes of Strep A infections leading to ARF and RHD and in improving treatment for established disease.

The future financial costs from ARF and RHD estimated in this report are expected to be absorbed by Australia's health system, with a continuing high human cost. With different kinds of investment, it would be possible to shift these costs to prevention – thereby saving lives, building equity, and closing the gap in health outcomes.

Report Outline

The costs of ARF and RHD were estimated in several stages of data analyses and disease modelling (Box 1). Stages 1 and 2 involved analyses of linked health data to determine the number of people who have recently had or are projected to develop ARF and/or RHD. Stages 3 and 4 modelled the disease trajectory in these people to estimate the number of people who will develop severe RHD, need valvular surgery, and/or die and the related medical care and associated financial cost.

Following this introductory section, the contents contained in this report are presented in four sections (1 to 4) based on the stages outlined in Box 1. The specific methods, results, and limitations for each stage are combined and described in each of these sections. The report then concludes with a general summary and conclusion in the final section.

Box 1: Methodological approach to estimating the cost of inaction

1. Epidemiological analyses	2. Case identification (people requiring SP)	3. Disease trajectory	4. Medical care and associated cost
1A The prevalence of ARF/RHD	**2A** N of people with a recent history of ARF/RHD (2016)	**3** Estimate the future clinical outcomes	**4** Estimate the future financial cost
1B The incidence rates of ARF/RHD	**2B** N of people to develop ARF/RHD (2016 – 2031)		

1A & 1B	Analysis of linked hospital and death data, available from July 1st, 2001 to December 31st, 2016, to determine the (A) prevalence and (B) incidence rates of ARF and of RHD.
2A	The number of living people at June 30th, 2016 who had been hospitalised for ARF or RHD in the previous 9.5 years and are recommended to receive medical care.
2B	The number of people projected to develop ARF or RHD between July 1st, 2016 and December 31st, 2031, based on existing patterns of disease incidence rates.
3	The estimated disease trajectories for all people who have a recent history of ARF/RHD (2A) or who are projected to develop ARF/RHD (2B). This includes the likelihood of progressing from ARF to RHD, from RHD to severe RHD requiring heart surgery, and from RHD to death.
4	The estimated future medical care and associated cost for each disease trajectory, including hospitalisations, secondary prophylaxis and specialist follow up, for all people who have a recent history of ARF/RHD (2A) or who are projected to develop ARF/RHD (2B).

1. The prevalence and incidence of ARF and RHD.

1.1. Data sources and methods

The prevalence and incidence of ARF and RHD were derived from routine collections of health data in the Northern Territory (NT), South Australia (SA), Queensland (QLD), and Western Australia (WA). These data comprise hospitalisation and mortality records, and the records can be linked electronically by person-based encrypted identifiers. The encrypted identifiers can be used when analysing the data to ensure that people are not counted more than once and to allow longitudinal analyses of each persons' medical history.

Data linkage was undertaken by the WA Data Linkage Branch, SA-NT DataLink, and the Queensland Statistical Analysis and Linkage Unit. SA and NT patient data were linked across these two jurisdictions to address the extensive cross-jurisdictional patient flows. Strict protocols are used to ensure individuals cannot be identified. The de-identified linked data thus allows person level analyses to ascertain the prevalence and incidence of ARF and RHD based on hospitalisations.

Hospitalisations for ARF and RHD were identified using the International Classification of Diseases, Tenth Revision, Australian Modification (ICD-10-AM) coding system. A hospitalisation for ARF was identified by the ICD-10-AM codes I00-I02. For RHD, the ICD-10-AM codes I05, I06, I08.0, I08.1, I08.3, and I09 were used to identify hospitalisations for RHD, and the total number of cases identified by these codes was scaled down to reflect their positive-predictive value based on previous work.[14,15] In order to differentiate first-ever RHD patients from those with a previous diagnosis, previous admissions were reviewed, with the full range of codes that are specified in the ICD-10-AM manual for 'RHD or other non-specified valvular disease' (I05-I09) being used to identify prior RHD hospitalisations.

Although the linked data covers all hospitalisation and mortality records in each jurisdiction, it is likely that the true prevalence and incidence of ARF is underestimated in this report. People with ARF/RHD who had not been admitted to hospital since 2001 would not be included, affecting prevalence. The guidelines recommend that all people with ARF or RHD are referred from a community medical service to a hospital but this does not always occur, as shown in a study in the NT, which found that more than 40% of ARF cases captured by the NT RHD register were not referred to hospital.[9] Further, this report does not include linked hospitalisation and mortality data on ARF and RHD from New South Wales, Victoria, Tasmania, or the Australian Capital Territory. However, indicative data from the New South Wales RHD program (Appendix A) suggests that there are relatively few cases compared to the jurisdictions included in this report.

1.2.   The prevalence of ARF and RHD

1.2.1.   Definitions

The prevalence of ARF and RHD were defined as the number of Aboriginal and Torres Strait Islander people who (a) had been hospitalised with a principal diagnosis of ARF or with a principal or secondary diagnosis of RHD in the period July 1st, 2001 to June 30th, 2016 and (b) were alive and aged under 65 years at June 30th, 2016. The dates for identification of people with ARF or RHD were chosen because data availability was consistent across all four jurisdictions for the study period, enabling comparable and contemporary analyses.

In this report, people who had been hospitalised for both ARF and RHD within the study period were counted as prevalent RHD only (i.e., people with RHD may have had a previous ARF hospitalisation, while people were counted as ARF only if they did not have a previous or subsequent RHD hospitalisation). Further, the prevalence of 'Severe RHD' was defined as people who had been admitted for RHD as well as with heart failure (ICD-10-AM I50) or at least one cardiovascular procedure of the heart valves (Table 1) during the study period. However, this definition did not capture valvular surgery in children with RHD from the NT because these procedures are performed at the Royal Children's Hospital in Melbourne, Victoria. For context, in 2014 there were six children (5 – 14 years of age) from the Northern Territory who had cardiac surgery for RHD in Melbourne.[16] Adult surgeries from NT would have been undertaken in South Australia and thus captured in the available data.

Table 2: ICD procedure codes used to identify cardiovascular procedures related to RHD.

Valvular procedure	ICD-10-AM procedure code
Repair	• 38487-00, 38456-10, 38456-11, 38456-01, 38485-01, 38483-00, 38270-02, 38270-01, 38270-03, 38480-01, 38480-00, 38480-02, 38481-01, 38481-00, 38481-02, 38475-00, 38475-01, 38477-00, 38477-01, 38485-00, 38456-16, 38456-15, 38456-17, 38456-18, 38653-05, 38653-04, 38653-06, 38653-07 • 38475-02, 38477-02* • 96222-00**
Replacement: bioprosthesis	• 38488-03, 38488-01, 38488-05, 38488-07, 38489-02, 38489-00, 38489-03, 38489-04, 38489-01, 38489-05 • 38488-09, 38488-08, 38488-10, 38488-11***
Replacement: mechanical prosthesis	• 38488-02, 38488-00, 38488-04, 38488-06

AM, Australian Modification.
*New codes added in the 4th edition (from 1st July 2004).
**New codes added in 9th edition (from 1st July 2015)
***New codes added in 8th edition (from 1st July 2013).

1.2.2.   Results: ARF and RHD

In 2016, there were 4,539 Aboriginal and Torres Strait Islander people living with ARF or RHD across the NT, SA, QLD, and WA (Table 3). This equates to more than one-percent of the Aboriginal and Torres Strait Islander population under 65 years of age in these jurisdictions; it is more than two percent of the population in the combined Northern Territory and South Australian jurisdictions. Almost two-thirds of prevalent ARF was in people who were under 25 years old in 2016. More than one-third of prevalent RHD was Severe RHD, meaning that these people had also been hospitalised for heart failure or a cardiac valvular procedure/surgery.

Table 3: Prevalence of acute rheumatic fever (ARF) and rheumatic heart disease (RHD) among Aboriginal and Torres Strait Islander people, with percent of the population with disease shown in parenthesis, by jurisdiction and age group, 30.06.2016.

Disease	Age group in years						
	0-4 N (%)	5-14 N (%)	15-24 N (%)	25-34 N (%)	35-44 N (%)	45-64 N (%)	All (0-64) N (%)
Northern Territory and South Australia							
ARF	7 (0.1)	237 (1.0)	297 (1.3)	135 (0.7)	63 (0.4)	34 (0.2)	773 (0.7)
Mild/Moderate RHD	<5 (0.0)	95 (0.4)	223 (1.0)	336 (1.7)	307 (2.2)	294 (1.5)	1,257 (1.1)
Severe RHD	<5 (0.0)	11 (0.0)	51 (0.2)	122 (0.6)	144 (1.0)	255 (1.3)	586 (0.5)
Total	11 (0.1)	343 (1.4)	571 (2.5)	593 (3.1)	515 (3.6)	583 (3.0)	2,615 (2.4)
Queensland							
ARF	6 (0.0)	168 (0.3)	210 (0.5)	73 (0.2)	24 (0.1)	27 (0.1)	508 (0.2)
Mild/Moderate RHD	<5 (0.0)	39 (0.1)	76 (0.2)	103 (0.3)	82 (0.4)	83 (0.3)	387 (0.2)
Severe RHD	<5 (0.0)	11 (0.0)	37 (0.1)	52 (0.2)	65 (0.3)	141 (0.4)	307 (0.1)
Total	10 (0.0)	219 (0.5)	323 (0.7)	228 (0.7)	171 (0.7)	251 (0.8)	1,202 (0.6)
Western Australia							
ARF	<5 (0.0)	101 (0.5)	111 (0.6)	59 (0.4)	31 (0.3)	16 (0.1)	320 (0.3)
Mild/Moderate RHD	<5 (0.0)	24 (0.1)	54 (0.3)	71 (0.4)	48 (0.4)	31 (0.2)	227 (0.2)
Severe RHD	<5 (0.0)	9 (0.0)	10 (0.1)	32 (0.2)	45 (0.4)	79 (0.5)	175 (0.2)
Total	<5 (0.0)	134 (0.6)	175 (0.9)	161 (1.0)	124 (1.1)	126 (0.8)	722 (0.8)
Northern Territory, South Australia, Queensland, and Western Australia							
ARF	15 (0.0)	506 (0.5)	618 (0.7)	267 (0.4)	118 (0.2)	77 (0.1)	1,601 (0.4)
Mild/Moderate RHD	<5 (0.0)	158 (0.2)	353 (0.4)	509 (0.8)	438 (0.9)	409 (0.6)	1,870 (0.5)
Severe RHD	<5 (0.0)	32 (0.0)	98 (0.1)	206 (0.3)	254 (0.5)	475 (0.7)	1,068 (0.3)
Total	23 (0.0)	696 (0.7)	1,068 (1.3)	982 (1.5)	810 (1.7)	961 (1.4)	4,539 (1.1)

Severe RHD were defined as people with RHD and a history of hospitalisation for heart failure or at least one cardiovascular procedure of the heart valves (as defined in Table 2).

1.3.   The incidence rates of ARF and RHD

1.3.1.   Definitions

The incidence rates of ARF and RHD were calculated as the annual number of Aboriginal and Torres Strait Islander people who were hospitalised for the first time for each disease according to the following definitions:

- First hospitalisation for ARF: first hospitalisation with a principal diagnosis of ARF and no history of hospitalisation for RHD or other non-specified valvular disease (ICD-10-AM I05-I09).

- First hospitalisation for RHD with no history of ARF hospitalisation: first hospitalisation with a principal or secondary diagnosis of RHD and no history of (1) RHD or other non-specified valvular disease (ICD-10-AM I05-I09) or (2) ARF.

Annual incidence rates were calculated for two periods: a 'contemporary period' and a 'long-term period.' The contemporary period comprised all records between Jan 1st, 2014 and December 31st, 2016. This period provided the most recent annual incidence rates based on a fixed clearance period of 12.5 years (given that the hospitalisation data starts from July 1st, 2001) for detecting previous ARF and RHD hospitalisations. The long-term period included all records between Jan 1st, 2011 to December 31st, 2016. This period allowed for an analysis of temporal trends in annual incidence rates, but incidence rates were calculated using a shorter fixed clearance period of 9.5 years to exclude people with previous ARF and/or RHD hospitalisations.

Age-specific incidence rates were calculated per 100,000 population for six age groups: 0-4-years-old (yo), 5-14yo, 15-24yo, 25-34yo, 35-44yo, and 45-64yo. The population denominators for each age group were taken from the Australian Bureau of Statistics (ABS) publication entitled 'E*stimates and projections of the Aboriginal and Torres Strait Islander Australians between 2001 and 2026*'.[17]

Models to calculate trends of annual incidence rates as a function of calendar year were independently fitted for each age group and jurisdiction (except for the NT and SA which were combined) for the long-term period using linear regression. The trends in incidence rates were used to predict the number of people who will get ARF or RHD (with no history of ARF hospitalisation) between mid-2016 and 2031 (described in Section 2.2).

1.3.2.   Results: ARF

The number of people and average age-specific incidence rates of ARF between 2014 and 216 in each jurisdiction are shown in Table 4.

The annual age-specific incidence rates between 2011 and 2016, with a total of 1035 people hospitalised for first ARF during the period (~172 people per year), are shown in Figure 3. The average and annual age-specific incidence rates were highest in the NT and SA for all age-groups under 45 years old. Only the trend in ARF incidence rates for the 5-14yo age-group in the NT and SA was significant (p=0.0496), at an increasing rate of approximately 28 people with ARF per 100,000 persons per year.

Table 4: Number and average annual age-specific incidence rates of people with a first acute rheumatic fever hospitalisation between 2014-2016.

Age group (years)	Number of people (incidence rate per 100,000 person-years)			
	NT & SA	QLD	WA	Total
0-4	20 (55.6)	9 (11.4)	2 (6.3)	31
5-14	187 (260.3)	105 (72.7)	58 (93.1)	350
15-24	66 (98.8)	25 (19.7)	12 (21.0)	103
25-34	29 (51.7)	11 (12.5)	9 (19.6)	49
35-44	15 (35.5)	4 (5.8)	4 (11.9)	23
45-64	4 (7.2)	1 (1.1)	4 (8.7)	9
Total	321	155	89	565
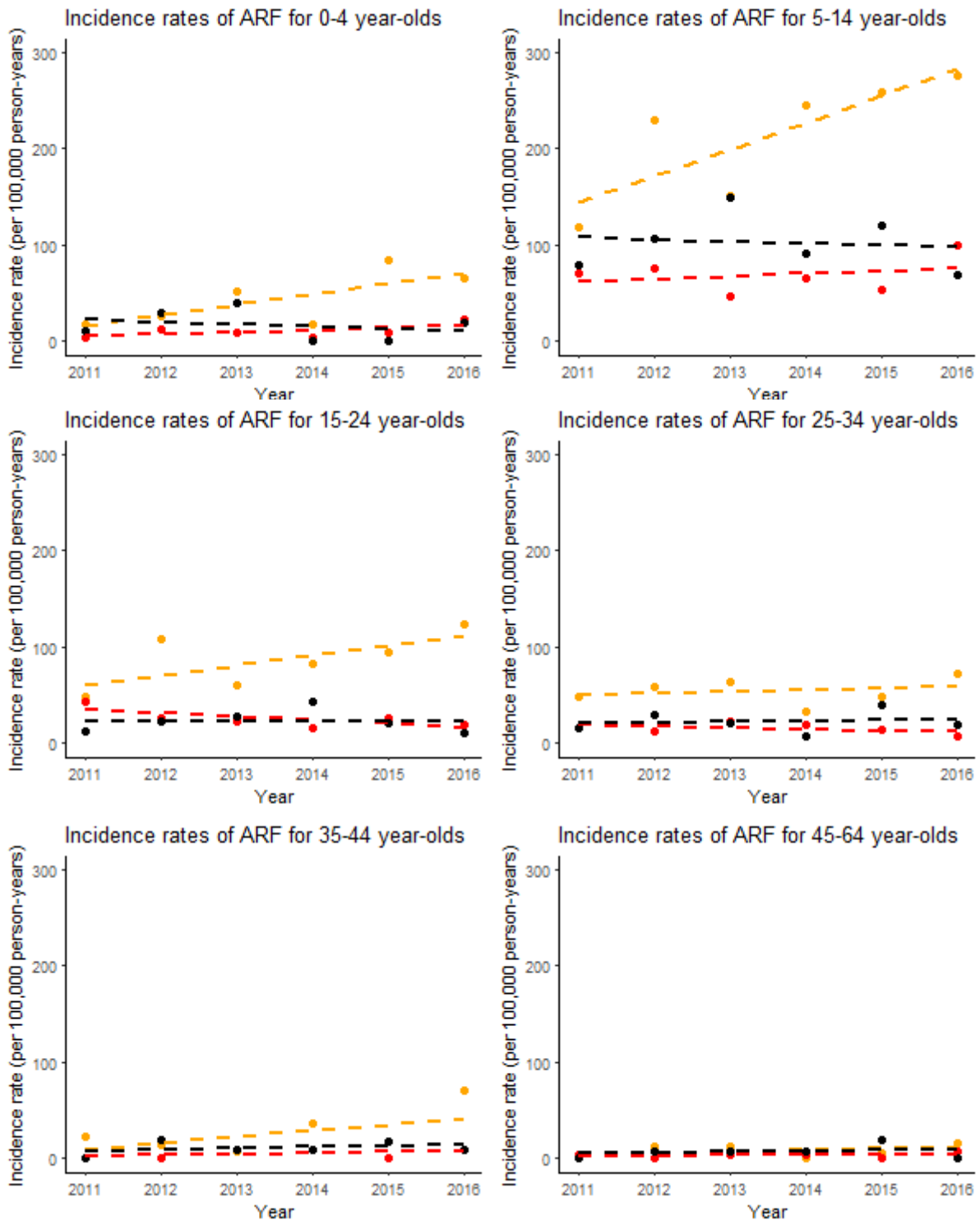
Figure 3: Age-specific incidence rates for acute rheumatic fever (ARF), 2011-2016.

Age-specific incidence rates for ARF in the NT and SA (orange), QLD (red), and WA (black). The points represent the annual incidence rates for the period 2011-2016 (9.5-year clearance period) and the dashed lines show the fitted linear trends in annual incidence rates.

1.3.3. Results: RHD with no history of ARF

The number of people and average age-specific incidence rates of RHD (with no history of RHD hospitalisation) between 2014 and 216 in each jurisdiction are shown in Table 5.

The annual age-specific incidence rates between 2011 and 2016, with a total of 1164 people hospitalised for first RHD during the period (~194 people per year), are shown in Figure 4. The age-specific incidence rates of RHD with no history of ARF hospitalisation were highest in the NT and SA for all age-groups. Only the trend in RHD incidence rates for the 15-24yo age-group in the NT and SA was statistically significant (p=0.016), at an increasing rate of approximately 9 people with RHD per 100,000 persons per year in this age group.

Table 5: Number and average annual age-specific incidence rates of people with their first rheumatic heart disease hospitalisations between 2014-2016.

Age group (years)	Number of people (incidence rate per 100,000 person-years)			
	NT & SA	QLD	WA	Total
0-4	5 (13.1)	4 (4.8)	0 (0.0)	8
5-14	24 (32.7)	22 (15.0)	5 (7.5)	50
15-24	73 (109.7)	28 (22.2)	8 (14.8)	110
25-34	93 (165.8)	30 (34.3)	13 (28.7)	136
35-44	70 (166.5)	21 (30.8)	10 (30.4)	102
45-64	102 (182.9)	72 (75.7)	18 (39.3)	192
Total	367	177	55	598
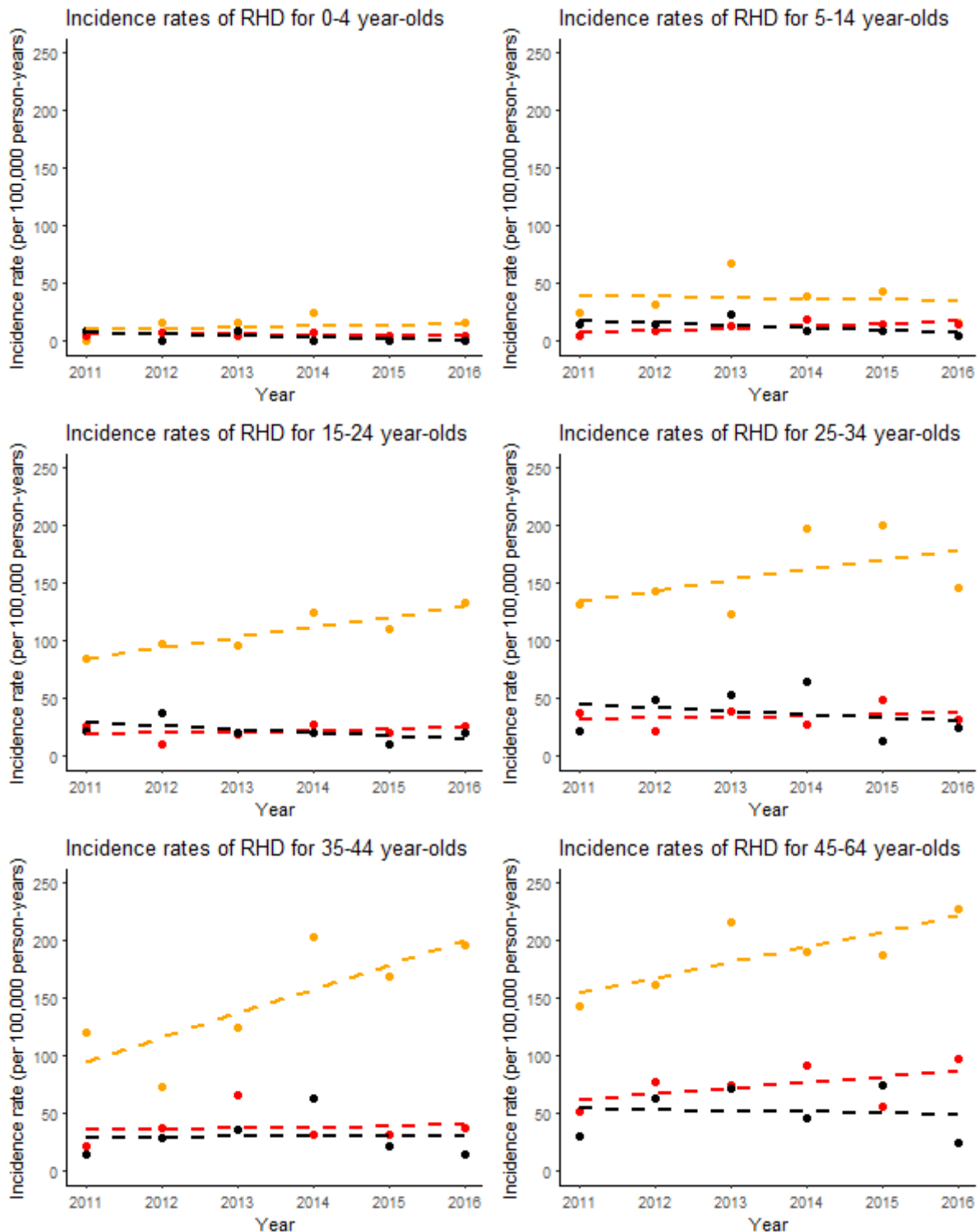
Figure 4: Age-specific incidence rates for rheumatic heart disease (RHD) with no history of acute rheumatic fever (ARF) hospitalisation, 2011-2016.

Age-specific incidence rates for RHD with no history of ARF hospitalisation in the NT and SA (orange), QLD (red), and WA (black). The points represent the annual incidence rates for the period 2011-2016 (9.5-year clearance period) and the dashed lines show the fitted linear trends in annual incidence rates.

2. The number of people with or projected to develop ARF or RHD

2.1. (A) People with a recent history of ARF or RHD

2.1.1. Definitions

People with a recent history of ARF or RHD were defined as the sub-set of prevalent cases (Section 1.2) who had been hospitalised after January 1st, 2007. This date, being 9.5 years before mid-2016, was chosen for two reasons. The first reason was that the ARF/RHD guidelines recommend a minimum period of 10 years of medical management. Thus, these people will all be on SP and a specialist follow-up plan for at least half a year from mid-2016. The second reason was that the modelled progression of ARF or RHD was limited to 10 years since diagnosis of either disease because published progression rates are limited to this time period.

2.1.2. Results: ARF and RHD with no history of ARF

Of the 4,539 people who have been hospitalised for ARF and/or RHD between 2001 and mid-2016 (Section 1.2.2), 3,420 (75%) people had been hospitalised since 2007 (Table 6). These people were included in the further analyses on the costs of ARF and RHD.

Table 6: Aboriginal and Torres Strait Islander people with a history of acute rheumatic fever (ARF) and/or rheumatic heart disease (RHD) between 01.01.2007 and 30.06.2016

Disease	Age group in years						
	0-4 N (%)	5-14 N (%)	15-24 N (%)	25-34 N (%)	35-44 N (%)	45-64 N (%)	All (0-64) N (%)
Northern Territory and South Australia							
ARF	7 (0.1)	236 (1.0)	237 (1.1)	78 (0.4)	43 (0.3)	13 (0.1)	614 (0.6)
Mild/Moderate RHD	<5 (0.0)	93 (0.4)	176 (0.8)	230 (1.2)	165 (1.2)	167 (0.9)	833 (0.7)
Severe RHD	<5 (0.0)	9 (0.0)	48 (0.2)	95 (0.5)	108 (0.8)	183 (0.9)	444 (0.4)
Total	11 (0.1)	338 (1.4)	461 (2.0)	403 (2.1)	316 (2.2)	363 (1.9)	1,892 (1.7)
Queensland							
ARF	6 (0.0)	168 (0.3)	157 (0.4)	52 (0.2)	9 (0.0)	18 (0.1)	410 (0.2)
Mild/Moderate RHD	<5 (0.0)	39 (0.1)	71 (0.2)	82 (0.3)	50 (0.2)	53 (0.2)	297 (0.1)
Severe RHD	<5 (0.0)	11 (0.0)	32 (0.1)	44 (0.1)	55 (0.2)	113 (0.3)	258 (0.1)
Total	10 (0.0)	218 (0.4)	260 (0.6)	178 (0.6)	115 (0.5)	184 (0.6)	965 (0.5)
Western Australia							
ARF	<5 (0.0)	99 (0.5)	78 (0.4)	35 (0.2)	14 (0.1)	12 (0.1)	240 (0.3)
Mild/Moderate RHD	<5 (0.0)	24 (0.1)	50 (0.3)	57 (0.4)	29 (0.3)	20 (0.1)	180 (0.2)
Severe RHD	<5 (0.0)	9 (0.0)	9 (0.0)	29 (0.2)	36 (0.3)	59 (0.4)	143 (0.2)
Total	<5 (0.0)	132 (0.6)	137 (0.7)	121 (0.8)	79 (0.7)	92 (0.6)	564 (0.6)
Northern Territory, South Australia, Queensland, and Western Australia							
ARF	15 (0.0)	503 (0.5)	472 (0.6)	165 (0.3)	66 (0.1)	43 (0.1)	1,264 (0.3)
Mild/Moderate RHD	<5 (0.0)	155 (0.2)	297 (0.3)	369 (0.6)	245 (0.5)	241 (0.4)	1,311 (0.3)
Severe RHD	<5 (0.0)	30 (0.0)	89 (0.1)	168 (0.3)	199 (0.4)	355 (0.5)	846 (0.2)
Total	23 (0.0)	688 (0.7)	858 (1.0)	703 (1.1)	510 (1.1)	638 (0.9)	3,420 (0.8)

Percent of the population with disease shown in parenthesis.

Severe RHD were defined as people with RHD and a history of hospitalisation for heart failure or at least one cardiovascular procedure of the heart valves (as defined in Table 2).

2.2. (B) People projected to develop ARF or RHD

2.2.1. Definitions

The number of Aboriginal and Torres Strait Islander people projected to develop ARF, a, or RHD, r, for age group i = (0-4, 5-14, 15-24, 25-34, 35-44, 45-64) was calculated as

$$\tilde{a}_i = c_i + j_i t, \text{ or}$$
$$\tilde{r}_i = d_i + k_i t$$

Where;

c and d = the mean of the annual age-specific ARF and RHD incidence rates per population, respectively, from the contemporary period (2014-2016),

j and k = the gradients of the linear trend in annual age-specific ARF and RHD incidence rates, respectively, from the long-term period (2011-2016), and

t = time, with 0, 1, 2, ..., 15 representing the calendar years 2016, 2017, 2018, ..., 2031.

Projections for each year were adjusted for the population size, which was based on the Australian Bureau of Statistics (ABS) estimates and projections of the Aboriginal and Torres Strait Islander Australians between 2001 and 2026.[17] As the ABS does not currently provide any Aboriginal projections beyond 2026, a conservative assumption of no population growth was made for the projected population between 2027 and 2031 using the ABS's projected population at 2026 for each of those years.

2.2.2. Results: ARF and RHD with no history of ARF

The number of people projected to develop ARF and RHD with no history of ARF hospitalisation in NT and SA, QLD, and WA between mid-2016 and the end of 2031 was 4885 and 5327 respectively (Table 7). Cumulative projections for each jurisdiction by year and age-group are shown in Figure 5.
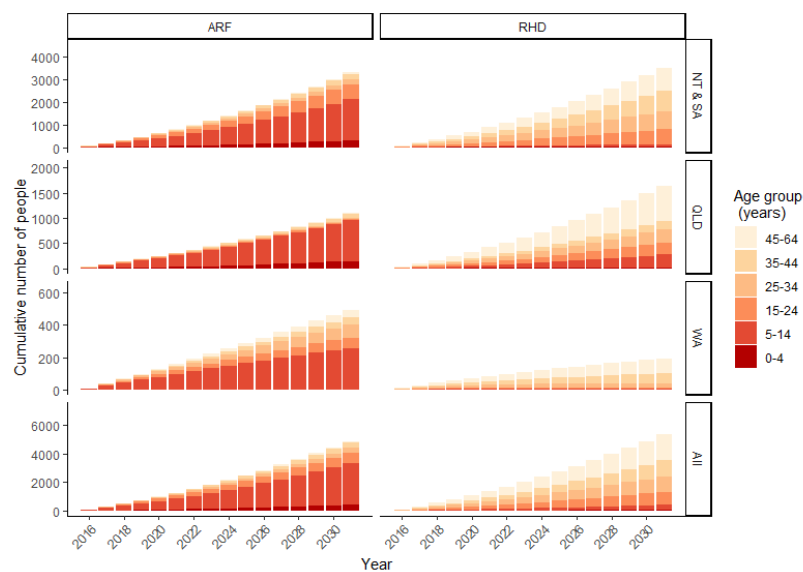
Figure 5: The cumulative number of first projected hospitalisations for ARF and RHD.

Table 7: Projected number of Aboriginal and Torres Strait Islander people who will develop acute rheumatic fever (ARF) and rheumatic heart disease (RHD), with no history of ARF, between 1.07.2016 and 31.12.2031

Disease	Age group in years						
	0-4	5-14	15-24	25-34	35-44	45-64	All (0-64)
	N	N	N	N	N	N	N
Northern Territory and South Australia							
ARF	295	1817	659	223	234	63	3291
RHD	44	96	653	799	888	1024	3504
Total	339	1913	1312	1022	1122	1086	6795
Queensland							
ARF	145	818	24	20	68	24	1100
RHD	12	261	237	263	160	696	1630
Total	158	1079	261	284	228	720	2729
Western Australia							
ARF	1	256	65	80	47	46	494
RHD	0	3	8	24	68	89	192
Total	1	259	74	104	114	135	686
Northern Territory, South Australia, Queensland, and Western Australia							
ARF	441	2891	748	323	349	132	4885
RHD	57	360	899	1086	1116	1809	5327
Total	498	3251	1647	1410	1465	1941	10211

3. Disease trajectory

3.1. Model of ARF and RHD progression

A state–transition model was developed to predict the progression (transition) between health states related to ARF and RHD in people over time (Figure 6). Health states were defined as people living with ARF, RHD, or Severe RHD that required surgery and people who die after the onset of ARF and/or RHD. Transitions between the health states were modelled in yearly increments for the duration of recommended medical care (secondary prophylaxis and specialist follow-up). This started at the year of last hospitalisation for people with a history of ARF and/or RHD prior to mid-2016 (Section 2.1.2) or the year that people are projected to develop ARF or RHD for onset occurring between mid-2016 and 2031 (Section 2.2.2). People who were predicted to develop RHD between mid-2016 and 2031 were divided between Mild and Moderate RHD and Severe RHD at diagnosis, with 11-12% having Severe RHD.[11] The distribution of severity in existing cases was based on their hospitalisation history of heart failure or valvular procedure (Section 2.1.2).

For the duration of recommended medical care, a proportion of people will remain in their current health state and the other proportion will transition to a more severe health state, as depicted by the arrows in Figure 6; thus, people who die will remain in the mortality state for the duration of time modelled. The proportion of people who will transition to a more severe health state is governed by the transition probabilities shown in Table 8. For example, 27% of people will progress from ARF to RHD within their first year of diagnosis, and a further 17% of people will progress between 1 and less than 5 years since diagnosis of ARF (44% in total within 5 years of ARF diagnosis).

Finally, there were several assumptions that were made in the model because of limited or no available data. These assumptions were:

1. Progression in people with a recent history of ARF prior to mid-2016 was limited to Mild/Moderate RHD (i.e., progression to Severe RHD with or without surgery was not modelled for this group), and progression in people with existing RHD prior to mid-2016 was limited to mortality. These limitations were made because only the date of last hospitalisation was known, not the date of disease onset on which the time-dependent transition probabilities are based. As a result, this assumption underestimates the estimated number of people who will develop Severe RHD, will require one or more valvular surgeries, or will die.

2. Projected cases of RHD were based on the observed incidence rates of RHD with no history of ARF hospitalisations. This was done to prevent double counting the RHD cases that are modelled to progress from ARF.

3. The duration of medical care and risk of progression between each health state (i.e. ARF to RHD, RHD to Severe RHD with surgery, or RHD to mortality) was limited to 10 years. This limitation was made because the risk of progression beyond 10 years of disease onset has not been quantified, and it results in an underestimate of the true costs of disease.

4. The risk of Severe RHD in the absence of surgical intervention was only modelled at the time of diagnosis. This assumption was made to limit the complexity of the model, and it underestimates the true number of people who will develop Severe RHD. This does not affect the estimates of

people who will develop Severe RHD requiring surgery (i.e., the number of people who will need surgery).

5. Recurrences of ARF were not modelled. This assumption was also made to simplify the model and is consistent with the preference to underestimate the true cost of ARF and RHD.

6. Mortality in ARF cases was not modelled as death from isolated ARF outside the context of RHD is considered a rare event.[18]
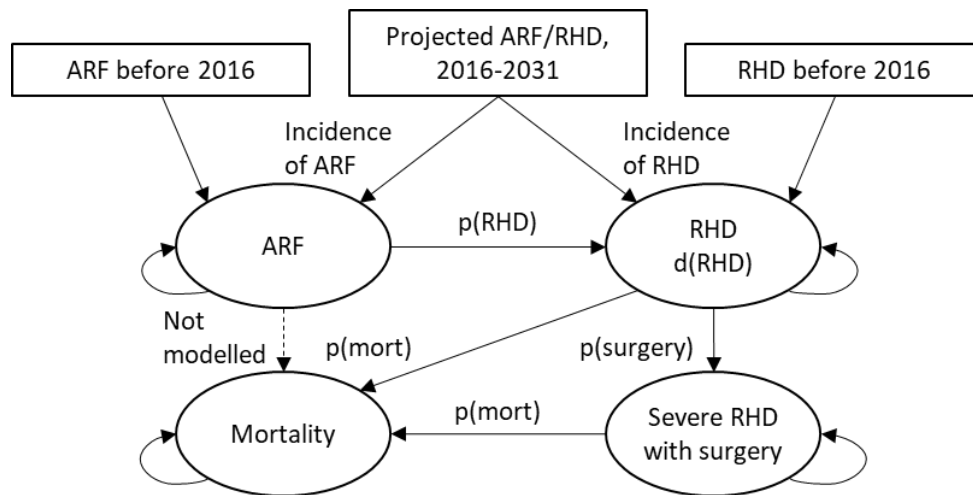
Figure 6: Model of disease states and transition possibilities for ARF and RHD

d(RHD), distribution of RHD severity at diagnosis; p(RHD), p(surgery), and p(mort), probabilities of progressing to RHD, Severe RHD requiring surgery, and mortality – with corresponding parameter estimates in Table 10.

Table 8: Time-dependent transition probabilities used in the state-transition model to estimate ARF and RHD trajectory over time.

Probability of disease progression within specified period (years)	Probability estimates*			
	Age range			
	0-64 years	0-14 years	15-24 years	25-64 years
p(RHD): progression from ARF to RHD[9]				
0-1 years	0.27			
1-5 years	0.17			
5-10 years	0.08			
p(surgery): progression from RHD to surgery[11]**				
0-1 years		0.06	0.09	0.07
1-5 years		0.09	0.03	0.06
5-10 years		0.05	0.02	0.04
p(death): progression from RHD to death[9,11] ***				
0-1 years		0.00	-	0.01
1-5 years		0.01	0.03	0.03
5-10 years		0.01	0.03	0.07

*Probability estimates represent the proportion of people transitioning over the whole period specified.

**Progression within 0-1 years was assumed to occur at diagnosis of RHD, ensuring an underestimate of disease costs. Further, the Cannon *et al.*[11] provided these risk estimates for RHD diagnosed at 5-14 years old, 15-24 years old, and at a combined 5-24 years old; the combined 5-24 year-old risk estimates were used for the progression and distribution of severity in 25-64 year-old people in this model as these estimates reflect the midpoints between the stratified estimates.

***The risk of mortality in 25-64-year-old people was based on all-age mortality reported by He *et al*, 2016.[9]

3.2.   (A) The modelled disease trajectory for people with a recent history of ARF and RHD

Of the 1,264 people with a recent history of ARF prior to mid-2016, 190 (15%) were estimated to progress to RHD. This progression to RHD comprised 97, 57, and 36 people from the NT and SA, QLD, and WA respectively.

It was estimated that 110 (5.1%) of the 2,156 people who have had a hospitalisation for RHD prior to mid-2016 will die within 10 years of that hospitalisation date (Table 9). The number of people estimated to die in the NT and SA, QLD, and WA was 63, 30, and 17 respectively.

Table 9: Predicted mortality for people with a recent history of RHD

Age at 30.06.2016	People with RHD N	Projected deaths* N (% of people with RHD)
0-4	8	0 (0.0)
5-14	185	2 (1.1)
15-24	386	12 (3.1)
25-34	538	30 (5.5)
35-44	444	28 (6.3)
45-64	595	38 (6.5)
Total	2156	110 (5.1)

*Deaths limited to 10 years since the date of last RHD hospitalisation.

3.3. (B) The modelled disease trajectory for people projected to develop ARF or RHD with no history of ARF

Disease trajectory in people projected to develop ARF

Of the 4,885 projected to develop ARF (Section 2.2.2), 2,535 people are estimated to progress to RHD within 10 years of their ARF diagnosis (Table 10). Of these people, 284 are estimated to be diagnosed with Severe RHD, and 462 will be diagnosed with or progress to Severe RHD requiring surgery. Thus, a total of 746 people who are projected to develop ARF between mid-2016 and 2031 are estimated to progress to Severe RHD.

It is estimated that 93 people will die after developing ARF between mid-2016 and 2031 and progressing to RHD.

Disease trajectory in people projected to develop RHD with no hospitalised history of RHD

There were 5326 people who are projected to develop RHD between mid-2016 and 2031. Of these people, 605 are estimated to be diagnosed with Severe RHD, and 908 will be diagnosed with or progress to Severe RHD requiring surgery (Table 11). Thus, a total of 1514 people who were projected to develop RHD between mid-2016 and 2031 are estimated to progress to Severe RHD.

It is estimated that 470 people will die after developing RHD between mid-2016 and 2031.

Table 10: Number of people who are projected to develop ARF by 2031 and who are predicted to progress to RHD, severe RHD, severe RHD requiring surgery, and/or death

	SA /NT	QLD	WA	Total
	N (%)	N (%)	N (%)	N (%)
Develop ARF	3291 (100)	1100 (100)	494 (100)	4885 (100)
Progress to RHD	1708 (52)	571 (52)	256 (52)	2535 (52)
Mild/Mod RHD	1517 (46)	508 (46)	227 (46)	2251 (46)
Severe RHD	191 (6)	63 (6)	29 (6)	284 (6)
Progress to severe RHD with surgical intervention	308 (9)	108 (10)	46 (9)	462 (9)
Total number of people who will develop severe RHD	499 (15)	171 (16)	75 (15)	746 (15)
Progress to death from RHD	65 (2)	15 (1)	13 (3)	93 (2)

Table 11: Number of people who are projected to develop RHD (with no history of ARF hospitalisation) by 2031 and who are predicted to progress to severe RHD and/or death.

	SA /NT	QLD	WA	Total
	N (%)	N (%)	N (%)	N (%)
Develop RHD with no history of RHD	3504 (100)	1630 (100)	192 (100)	5326 (100)
Mild/Mod RHD	3105 (89)	1445 (89)	170 (89)	4721 (89)
Severe RHD	399 (11)	185 (11)	22 (11)	605 (11)
Progress to severe RHD with surgical intervention	593 (17)	282 (17)	33 (17)	908 (17)
Total number of people who will develop severe RHD	992 (28)	466 (29)	55 (29)	1514 (28)
Progress to death from RHD	318 (9)	133 (8)	19 (10)	470 (9)

4. Medical care and associated costs

4.1. Medical care

In this report, medical care included two main components: (1) inpatient hospitalisation for ARF or RHD and (2) annual disease management, which included secondary prophylaxis and specialist follow-up over a 10-year period post-hospitalisation. Medical care was estimated for the modelled disease trajectory occurring after mid-2016 for all people with a history of ARF or RHD hospitalisation between 2007 and mid-2016 (Section 3.2) and for all people projected to develop ARF or RHD between mid-2016 and 2031 (Section 3.3).

The methods for estimating the medical care for the several scenarios of modelled disease trajectories that could occur in people with a recent history or projected to develop ARF and/or ARD are illustrated in Figure 7. For people with a recent history of ARF or RHD:

- Medical care comprised annual disease management occurring after mid-2016 for the remainder of the 10-year period starting from the date of last hospitalisation (Trajectories A and B).

- If the disease progressed in severity, medical care comprised an inpatient hospitalisation for disease progression, and 10 years of annual disease management in addition to the disease management occurring between mid-2016 and the date of disease progression (Trajectory C).

For people projected to develop ARF or RHD:

- Medical care comprised inpatient hospitalisation for the acute episode of ARF or RHD and annual disease management (Trajectories D and E).

- If the disease progressed in severity, medical care comprised the initial and subsequent inpatient hospitalisation, and the annual disease management between hospitalisations and 10 years after the last hospitalisation, (Trajectory F) unless death occurs (Trajectory G).

- These trajectories and medical care requirements could occur for anyone projected to develop ARF or RHD up to 31.12.2031 (Trajectory H)
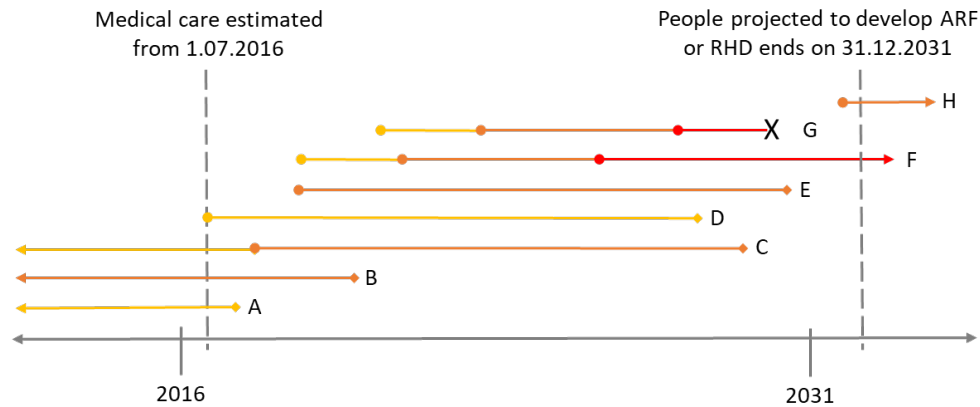
Figure 7: Medical care for example trajectories of ARF (yellow), RHD (orange), and Severe RHD requiring valvular surgery (red).

Dots represent hospitalisation, diamonds represent end of annual disease management, X represents death. See text for further description.

4.2. Treatment costs

Treatment costs for the estimated medical care were based on previous estimates and are shown in the adapted Table 12.[11] In summary, the cost of an inpatient admission was applied to all people projected to develop or progress to ARF and/or RHD. The inpatient cost of cardiac surgery included pre-operative medical and dental management, post-operative intensive care, and interstate travel (e.g., from the NT to SA for adults and to Victoria for children) to the centre that performed the surgery. For the proportion of people living in remote areas, inpatient costs also included adjustments for the provision of care, using the Independent Hospital Pricing Authority remoteness price adjustment, and travel for the patient and accompanying escort by emergency transfer.

Annual disease management included four-weekly administration of long acting antibiotic (benzathine penicillin G, Bicillin L-A®) by a nurse and 1-4 reviews by a general practitioner and consultations by paediatrics, cardiology (including use of echocardiograms), and dentist. The frequency of these consultations ranged from six monthly to two yearly, depending on the severity of RHD.[19] For the proportion of patients living in remote communities, disease management costs also included return travel and accommodation for the patient and accompanying escort for specialist visits.

Several simplifying assumptions have been made to estimate costs, each intended to underestimate rather than overestimate the true cost. These assumptions were:

1. Adherence to secondary prophylaxis and follow up care is 50% the recommended schedule

2. Moderate RHD was assumed to cost the same as Mild RHD because the distinction in disease severity in this report could not be ascertained.

3. Progression, and therefore medical care and costs, from ARF to Severe RHD (with or without valvular surgery) was not estimated for people hospitalised with ARF and not RHD before mid-

2016. Similarly, progression from Mild/Moderate RHD to Severe RHD (with or without valvular surgery) was not estimated for people hospitalised with RHD before mid-2016.

4. The adverse outcomes and costs of anticoagulation after valvular replacement were not included.

5. The costs associated with RHD care in pregnancy were not included

6. The health-related costs to community medical services to follow-up patients was not included, including outreach services and approaches.

Table 12: Summary of ARF/RHD costs at 2015 prices, Australia.

Item		Cost ($AU, 2015)
Acute costs		Per episode
Inpatient		
ARF	Child	11,243
	Adult	12,023
RHD, non-surgical	Child	10,985
	Adult	9,113
RHD, surgical	Child	69,755
	Adult	67,080
ARF/RHD management (per year)		Per year
Severity of RHD*	Mild	1,907
	Severe	3,650

*Mild RHD is assumed for patients with ARF and for mild/moderate RHD
Modified from the Supplemental Material A, Table S7, in Cannon *et al.*[20]

4.3.   (A) The estimated future medical cost of ARF and RHD for people with a recent history of disease

The estimated cost of medical care from July 1st, 2016 to the end of recommended follow-up for people with a history of ARF or RHD since January 1st, 2007 was $26.6 million to the health system, comprising $24.8 million in annual disease management cost and $1.7 million in hospitalisations for people with ARF estimated to progress to RHD (Table 13). The medical care cost was relatively evenly distributed across the age groups, but these costs did not include progression from Mild/Moderate RHD to Severe RHD requiring surgery, which is likely to affect people in the older age groups.

Table 13: The estimated future cost of medical care from 01.07.2016 for Aboriginal and Torres Strait Islander people with a history of acute rheumatic fever (ARF) and/or rheumatic heart disease (RHD) between 01.01.2007 and 30.06.2016

Medical management	Age group in years						
	0-4	5-14	15-24	25-34	35-44	45-64	All (0-64)
	Cost ($AU)	Cost ($AU)	Cost ($AU)	Cost ($AU)	Cost ($AU)	Cost ($AU)	Cost ($AU)
Northern Territory and South Australia							
Disease management	106,285	2,515,121	2,859,579	2,831,528	2,154,028	2,925,170	13,391,712
Hospitalisation*	17,749	446,517	259,979	87,169	52,012	18,763	882,188
Total	124,034	2,961,638	3,119,558	2,918,697	2,206,040	2,943,933	14,273,900
Queensland							
Disease management	98,524	1,533,044	1,610,847	1,337,109	939,928	1,730,955	7,250,406
Hospitalisation*	17,310	270,096	144,654	59,863	9,255	16,224	517,401
Total	115,834	1,803,140	1,755,501	1,396,971	949,183	1,747,179	7,767,808
Western Australia							
Disease management	18,117	987,768	828,941	900,761	704,174	789,182	4,228,941
Hospitalisation*	5,982	171,084	77,352	40,092	20,508	12,011	327,029
Total	24,099	1,158,852	906,293	940,853	724,682	801,193	4,555,970
Northern Territory, South Australia, Queensland, and Western Australia							
Disease management	222,925	5,035,933	5,299,366	5,069,398	3,798,129	5,445,307	24,871,059
Hospitalisation*	41,042	887,697	481,985	187,123	81,775	46,997	1,726,619
Total	263,967	5,923,630	5,781,352	5,256,521	3,879,904	5,492,304	26,597,678

*Hospitalisation for non-surgical RHD for people estimated to progress from ARF.

4.4. (B) The estimated medical costs for people projected to develop ARF or RHD

People expected to develop ARF and RHD will incur costs to the health system at the time of diagnosis (hospital admission) and for each year that they are receiving secondary prophylaxis management. Additional costs are incurred if ARF recurs or progresses to RHD, requiring further hospitalisation and protracted secondary prophylaxis, and if valvular surgery is required. The aggregated cost of these components for all people projected to develop ARF or RHD (without a history of ARF hospitalisation) between mid-2016 and 2031 is summarised as a cumulative total by year in Figure 8.
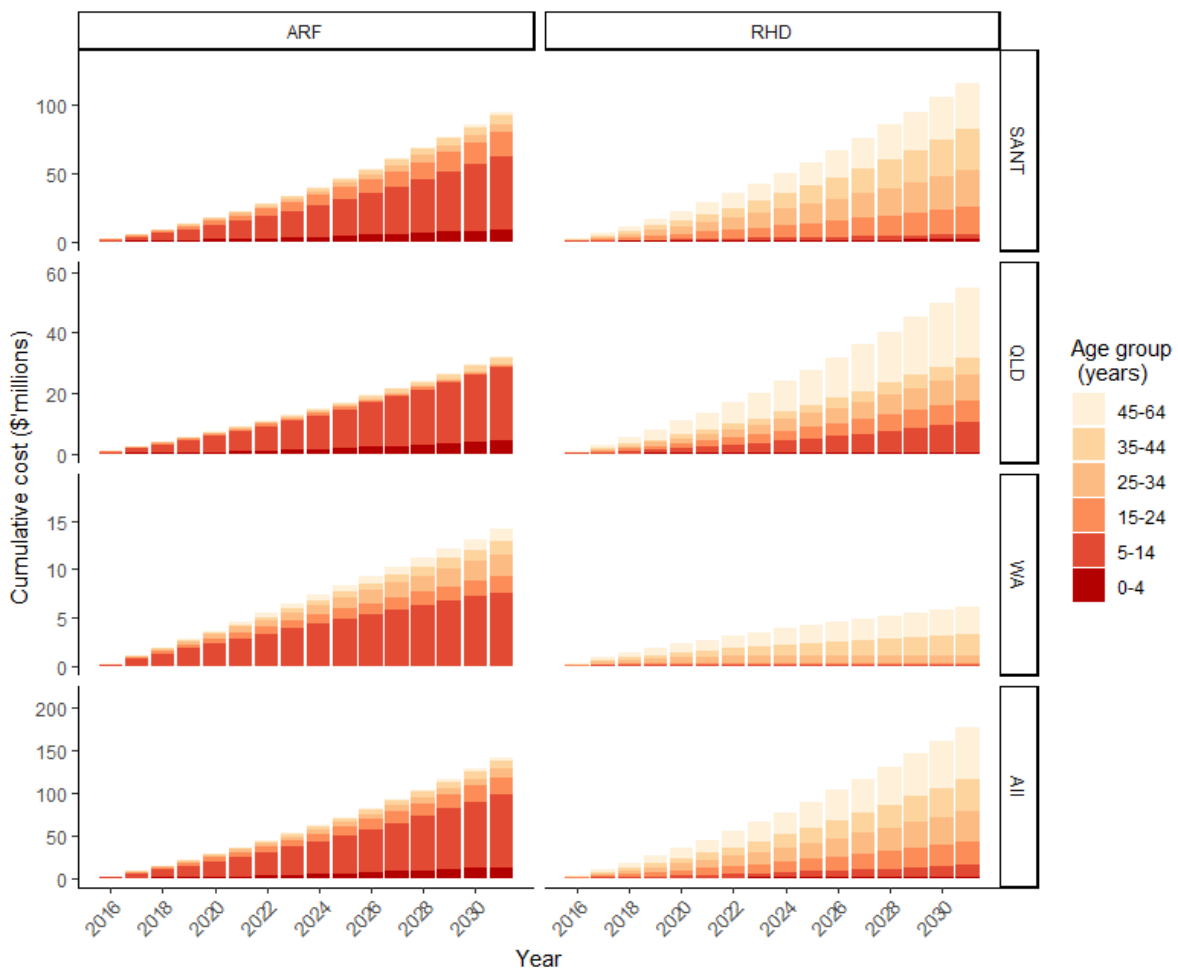
Figure 8: The estimated cumulative cost of medical care for cases of ARF or RHD projected to occur between mid-2016 and 2031.

The estimated cost of medical care for people projected to develop ARF between mid-2016 and 2031 totalled $141 million (Table 14). This included $58 million in long-term disease management and $83 million in hospitalisations, and the majority of the total cost ($85 million, 60.6%) is estimated to be incurred by children 5-14-years-old. For people projected to develop RHD with no history of ARF hospitalisation, the estimated cost of medical care was $170 million (Table 15). In total, a cost of $317

million was estimated for the medical care of all people projected to develop ARF and RHD between mid-2016 and 2031 (Table 16).

Table 14: The estimated future cost of medical care from 01.07.2016 for Aboriginal and Torres Strait Islander people projected to develop acute rheumatic fever (ARF) between 01.07.2016 and 31.12.2031.

Medical management	Age group in years						
	0-4	5-14	15-24	25-34	35-44	45-64	All (0-64)
	Cost ($AU)	Cost ($AU)	Cost ($AU)	Cost ($AU)	Cost ($AU)	Cost ($AU)	Cost ($AU)
Northern Territory and South Australia							
Disease management	3,522,750	21,702,473	7,582,033	2,587,716	2,712,665	726,102	38,833,739
Hospitalisation	5,180,514	31,915,400	10,006,902	3,622,105	3,796,999	1,016,347	55,538,267
Total	8,703,264	53,617,873	17,588,935	6,209,821	6,509,664	1,742,449	94,372,006
Queensland							
Disease management	1,736,299	9,771,151	279,813	234,562	784,509	276,930	13,083,264
Hospitalisation	2,553,381	14,369,339	369,302	328,324	1,098,101	387,627	19,106,075
Total	4,289,681	24,140,490	649,115	562,886	1,882,611	664,557	32,189,339
Western Australia							
Disease management	9,812	3,053,936	752,467	921,856	542,078	529,069	5,809,217
Hospitalisation	14,429	4,491,082	993,119	1,290,350	758,763	740,554	8,288,297
Total	24,241	7,545,017	1,745,585	2,212,206	1,300,841	1,269,623	14,097,513
Northern Territory, South Australia, Queensland, and Western Australia							
Disease management	5,268,861	34,527,559	8,614,313	3,744,134	4,039,252	1,532,101	57,726,220
Hospitalisation	7,748,324	50,775,821	11,369,323	5,240,779	5,653,863	2,144,528	82,932,639
Total	13,017,186	85,303,380	19,983,635	8,984,913	9,693,115	3,676,629	140,658,859

Table 15: The estimated future cost of medical care from 01.07.2016 for Aboriginal and Torres Strait Islander people projected to develop rheumatic heart disease (RHD) between 01.07.2016 and 31.12.2031.

Medical management	Age group in years						
	0-4	5-14	15-24	25-34	35-44	45-64	All (0-64)
	Cost ($AU)	Cost ($AU)	Cost ($AU)	Cost ($AU)	Cost ($AU)	Cost ($AU)	Cost ($AU)
Northern Territory and South Australia							
Disease management	611,099	1,121,841	8,449,150	10,415,285	12,272,111	13,605,258	46,474,744
Hospitalisation	1,045,417	2,260,953	11,848,997	16,045,871	17,834,371	20,561,755	69,597,364
Total	1,656,516	3,382,794	20,298,147	26,461,156	30,106,482	34,167,013	116,072,108
Queensland							
Disease management	119,063	3,687,759	3,007,173	3,340,195	2,021,656	9,207,939	21,383,784
Hospitalisation	289,336	6,142,284	4,291,464	5,287,610	3,222,103	13,981,897	33,214,694
Total	408,399	9,830,043	7,298,637	8,627,804	5,243,759	23,189,835	54,598,478
Western Australia							
Disease management	0	35,713	98,125	290,724	806,725	1,062,714	2,294,001
Hospitalisation	0	66,880	151,570	489,741	1,358,975	1,790,202	3,857,368
Total	0	102,593	249,696	780,465	2,165,700	2,852,915	6,151,369
Northern Territory, South Australia, Queensland, and Western Australia							
Disease management	730,162	4,845,312	11,554,448	14,046,204	15,100,492	23,875,910	70,152,528
Hospitalisation	1,334,753	8,470,118	16,292,031	21,823,222	22,415,448	36,333,853	106,669,426
Total	2,064,915	13,315,430	27,846,480	35,869,426	37,515,940	60,209,764	176,821,954

Table 16: The estimated future cost of medical care from 01.07.2016 for Aboriginal and Torres Strait Islander people projected to develop acute rheumatic fever (ARF) and/or rheumatic heart disease (RHD) between 01.07.2016 and 31.12.2031.

Medical management	Age group in years						
	0-4	5-14	15-24	25-34	35-44	45-64	All (0-64)
	Cost ($AU)	Cost ($AU)	Cost ($AU)	Cost ($AU)	Cost ($AU)	Cost ($AU)	Cost ($AU)
Northern Territory and South Australia							
Disease management	4,133,849	22,824,314	16,031,183	13,003,001	14,984,776	14,331,360	85,308,483
Hospitalisation	6,225,931	34,176,353	21,855,899	19,667,976	21,631,370	21,578,102	125,135,631
Total	10,359,780	57,000,667	37,887,082	32,670,977	36,616,146	35,909,462	210,444,114
Queensland							
Disease management	1,855,362	13,458,909	3,286,986	3,574,757	2,806,165	9,484,869	34,467,048
Hospitalisation	2,842,718	20,511,623	4,660,766	5,615,934	4,320,204	14,369,524	52,320,769
Total	4,698,080	33,970,533	7,947,752	9,190,691	7,126,369	23,854,392	86,787,817
Western Australia							
Disease management	9,812	3,089,648	850,592	1,212,580	1,348,803	1,591,783	8,103,217
Hospitalisation	14,429	4,557,962	1,144,689	1,780,091	2,117,738	2,530,756	12,145,665
Total	24,241	7,647,610	1,995,281	2,992,671	3,466,541	4,122,539	20,248,882
Northern Territory, South Australia, Queensland, and Western Australia							
Disease management	5,999,023	39,372,871	20,168,761	17,790,338	19,139,744	25,408,011	127,878,748
Hospitalisation	9,083,078	59,245,938	27,661,354	27,064,001	28,069,312	38,478,382	189,602,064
Total	15,082,101	98,618,809	47,830,115	44,854,339	47,209,056	63,886,393	317,480,813

Summary and limitations

This report offers the best available data on the current and future burden of ARF and RHD in Australia, and it represents the first quantification of the human and financial cost of RHD into the future. It demonstrates that at least 4,539 Aboriginal and Torres Strait Islander people are living with the effects of ARF or RHD in Australia (3,420 of those should be receiving medical care) and, without immediate action, 10,212 additional people will develop ARF or RHD by 2031 (Figure 9). To provide medical care to the people with a recent history of ARF or RHD and the people projected to develop these diseases, Australia will need to spend at least $344 million, which includes the cost of more than 1,370 heart operations. Tragically, 628 people currently with or who will develop RHD between mid-2016 and 2031 will die. Most of the people affected by ARF and RHD will be young and in the prime of their lives.
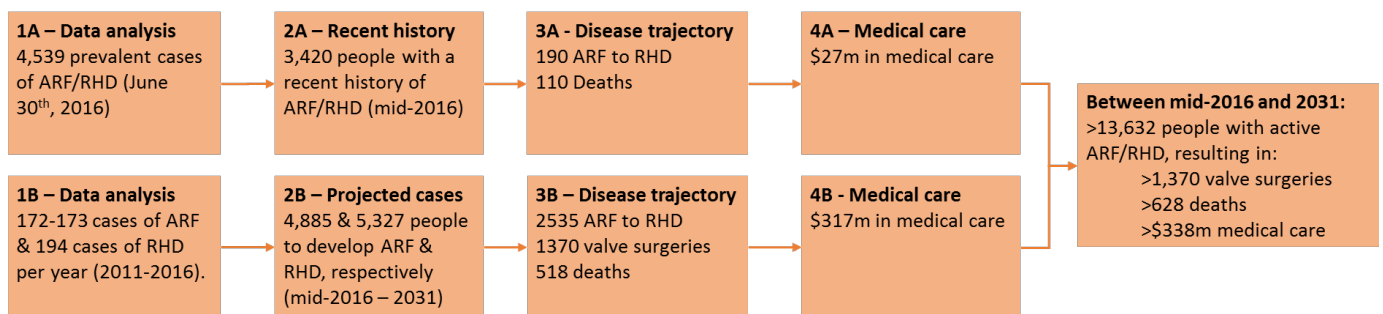

Figure 9: Summary of the estimated costs of acute rheumatic fever and rheumatic heart disease for Indigenous people in the Northern Territory, South Australia, Queensland, and Western Australia with a recent history of disease and those projected to develop it between mid-2016 and 2031.

The best data and analytic tools have been used to model and predict the future costs of ARF and RHD. However, there were several limitations:

- People with ARF or RHD were identified from hospital data. As not all cases are hospitalised, people with a new diagnosis of ARF or RHD who were not hospitalised will not have been counted in this report, leading to an underestimate of both the human and financial costs of these diseases.

- We assumed that there would be no population growth between 2027 and 2031 because ABS estimates for that time period are not available. Thus 5 years of population growth will further increase our estimates.

- Projected cases of RHD were based on the observed incidence rates of RHD without a history of ARF hospitalisation, rather than the observed incidence rates of RHD with and without a history of ARF hospitalisation. This approach was taken to prevent double counting cases of RHD that were modelled to progress from the projected ARF cases. Because these are projected cases, rather than observed cases, it is not possible to validate the accuracy of this approach in estimating the true number of future RHD cases (with and without a history of ARF).

- The estimates of the risk of disease progression over time were taken from external sources that derived such estimates from register data, which included cases diagnosed in outpatient settings. The date of ARF or RHD onset in this report was assumed to be the date of first

hospitalisation for each condition, and the progression of disease started from this date. This may underestimate the health and associated economic outcomes as hospitalised cases are likely to be more severe at 'assumed' date of onset than those captured by the jurisdictional control programmes.

- Deaths among people with RHD are all-cause mortality, not deaths attributed directly to RHD

- Women with RHD are at risk of serious complications and require close medical supervision due to the physical demands on the heart during pregnancy and birth. This analysis has not been performed by gender, which means differences in the medical costs and experiences between men and women living with RHD are not made explicit.

- The financial cost of ARF and RHD were limited to the costs of hospitalisations and annual disease management, namely secondary prophylaxis and specialist follow-ups, from the public health sector perspective. The analysis did not account for the complex and costly realities of cross-cultural communication, the cost of translators, the administrative and transport requirements of follow-up for people who move frequently, the medical challenges of managing comorbid disease that may be worsened by ARF and RHD, and the potential adverse outcomes and costs of anticoagulation for people who have had a heart valve replacement. Similarly, it did not account for losses in education, employment, or social contributions including childrearing or sharing cultural knowledge, all of which may far exceed direct financial expenditure and contribute to the social inequality between Aboriginal and Torres Strait Islander and non-Indigenous Australians

- Extrapolating from past events may not be a reliable means of prediction. For example, the cause of increasing rates of ARF is unclear. It is possible that greater awareness of the disease means the correct diagnosis is being made more often, and/or that more children are living in circumstances which drive ARF and RHD.

Acknowledging these limitations, the most conservative estimates have been made throughout this report. The cumulative effect of these conservative choices means that predictions are likely to represent an under-estimate of ARF and RHD costs.

Conclusion

This report confirms that ARF and RHD cause enormous human and financial costs. These data contribute to the compelling economic, social and moral imperative to act to end RHD in Australia. The toll is inequitable between Indigenous and non-Indigenous people, but it is not inevitable. New cases of ARF are almost unheard of among non-Indigenous people demonstrating that these diseases are entirely preventable and that the costs are avoidable and unnecessary. Resources could be better deployed on preventative rather than reactionary measures. Such measures could include reducing crowded living to reduce exposure and improving recognition, diagnosis, and treatment of Strep A throat and skin infections. Measures that result in a reduction in ARF and RHD would improve health and possibly social equity for Aboriginal and Torres Strait Islander people in Australia.

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Appendix A

Supplementary data from the NSW RHD Register

Table 17: Cases notified with ARF and/or RHD by ethnicity, NSW 1 October 2015 - 19 September 2018

Ethnicity	ARF	RHD	ARF&RHD
Aboriginal and Torres Strait Islander	27	14	5*
Māori and Pacific Islander	10	9	12
Other	12	7	0
Unknown	1	7	0
Total	50	37	17

* Includes one case also identifying as Māori and Pacific Islander

ARF without a diagnosis of RHD; RHD without a diagnosis of ARF; ARF&RHD, cases notified with both ARF and RHD.