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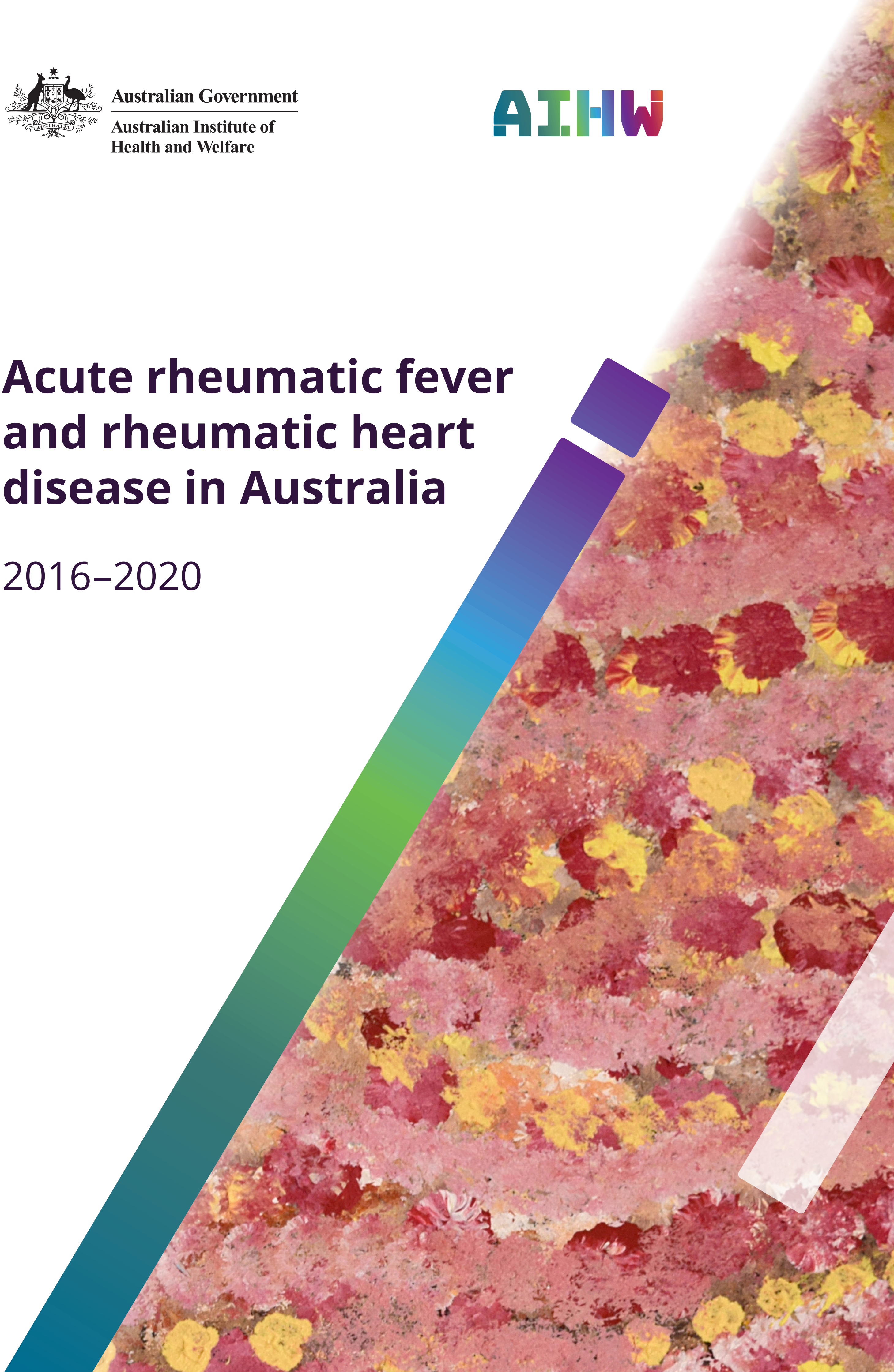
Acute rheumatic fever and rheumatic heart disease in Australia

2016–2020



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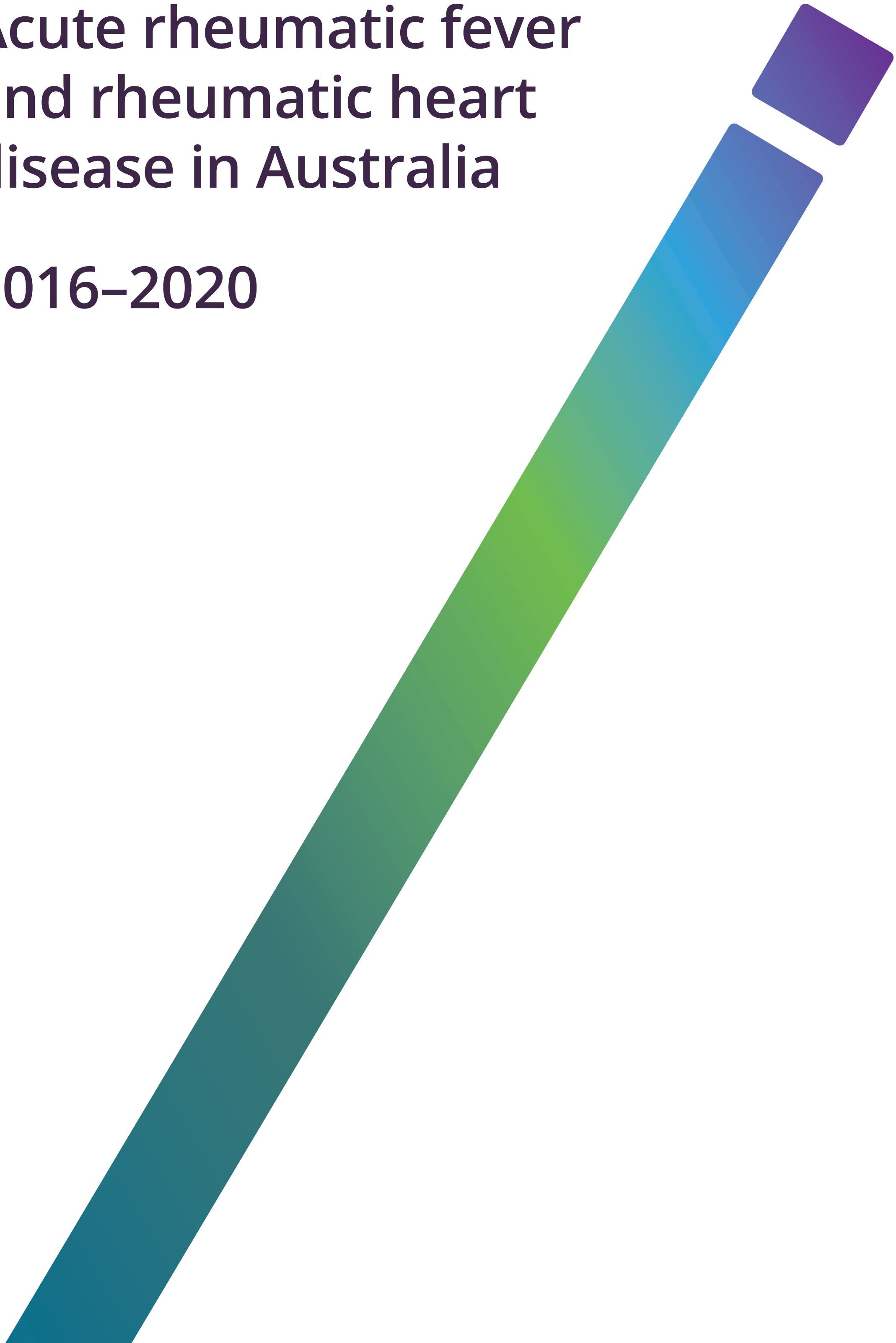

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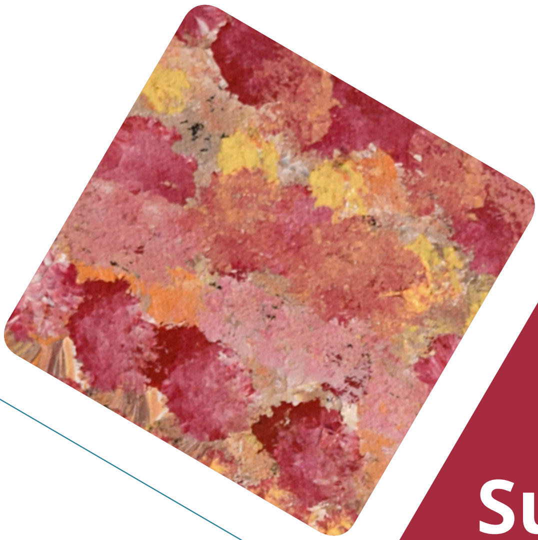


Summary

Summary

This is the fourth annual report from the National Rheumatic Heart Disease (RHD) data collection. It presents information on acute rheumatic fever (ARF) and RHD in Australia drawn from the established jurisdictional registers in New South Wales, Queensland, Western Australia, South Australia and the Northern Territory, from 2016–2020. Throughout the report, some data from New South Wales are incorporated with figures from other jurisdictions and some remain separate, depending on comparability between jurisdictions.

At 31 December 2020, there were 9,158 people living with a diagnosis of ARF and/or RHD recorded on clinical registers in New South Wales, Queensland, Western Australia, South Australia and the Northern Territory. Of these, 2,886 people (32%) had only ARF recorded, 3,197 people (35%) had only RHD recorded, and 3,075 people (34%) had both ARF and RHD recorded.

Acute rheumatic fever

In 2016–2020:

- 2,611 diagnoses of ARF were recorded in 2,380 people in New South Wales, Queensland, Western Australia, South Australia and the Northern Territory (3.0 per 100,000 population over the 5 years combined) (Supplementary ARF tables 1 and 2)
- Indigenous Australians accounted for 92% (2,392) of the diagnoses (65 per 100,000 population over the 5 years combined) (Supplementary ARF Table 2)
- the number and rate of diagnoses among Indigenous Australians increased from 424 (60 per 100,000) in 2016 to 521 (69 per 100,000) in 2020 (Supplementary ARF Table 3)
- ARF was more common among Indigenous females (1,350 cases, 74 per 100,000) than males (1,042 cases, 57 per 100,000) (Supplementary ARF Table 4b)
- the highest rate was among Indigenous Australians aged 5–14 (1,094 diagnoses, 135 per 100,000).

The median age at diagnosis among Indigenous Australians was 19 (Supplementary ARF Table 4b).

Rheumatic heart disease

Information on people with RHD in New South Wales is not comparable with that from the other jurisdictions, as it relates only to people aged under 35 at diagnosis.

Prevalence

At 31 December 2020, 6,039 people (61 per 100,000) living with RHD were recorded on the jurisdictional registers in Queensland, Western Australia, South Australia and the Northern Territory (Supplementary RHD Table 1). Of these,

- 27% (1,641) were aged under 25 (Supplementary RHD Table 2a)
- 67% (4,017) were females (Supplementary RHD Table 1)
- the highest prevalence rate was in the Northern Territory (936 per 100,000) and the greatest number was in Queensland (2,725 people) (Supplementary RHD Table 1)
- 3 in 4 diagnoses (4,696, 78%) were among Indigenous Australians (Supplementary RHD Table 2a)
- the median age among Indigenous Australians with RHD (34 years) was considerably younger than for non-Indigenous Australians (59 years) (Supplementary RHD Table 2a).

At 31 December 2020, there were 99 people living with RHD recorded on the register in New South Wales (Supplementary RHD Table 2b).

Incidence

Over the period 2016–2020 in Queensland, Western Australia, South Australia and the Northern Territory:

- 1,339 new RHD diagnoses among Indigenous Australians were reported (61 per 100,000 population) (Supplementary RHD Table 4a)
- new RHD diagnoses were more common among Indigenous females than males (80 and 43 diagnoses per 100,000, respectively) (Supplementary RHD Table 6)
- 54% of new diagnoses were among Indigenous Australians aged under 25 (754 diagnoses) (Supplementary RHD Table 6)
- the greatest number and highest rate of new diagnoses among Indigenous Australians was in the Northern Territory (557, or 146 per 100,000) (Supplementary RHD Table 4a)
- the number and rate of RHD diagnoses among Indigenous Australians increased from 222 (51 per 100,000) in 2016 to 332 (73 per 100,000) in 2018. The number and rate have since decreased to 279 and 61 per 100,000, respectively, in 2020 (Supplementary RHD Table 4a).

In 2016–2020, 58 Australians were diagnosed with RHD in New South Wales (Supplementary RHD Table 4b).

- Almost three-quarters of new cases (74%, 43 people) were in non-Indigenous Australians, with Pacific Islanders accounting for 29% (17) of all new RHD diagnoses in New South Wales (Supplementary RHD Table 5b).

Heart surgery for RHD

In 2016–2020 in Queensland, Western Australia, South Australia and the Northern Territory, 566 people underwent 595 surgical events for RHD. Most of these were Indigenous Australians, with 63% (359) of patients and 64% (380) of events being among Indigenous Australians (Supplementary RHD Table 9a).

In 2016–2020 in New South Wales, 12 people underwent 16 surgical events. Due to small numbers, the breakdown by Indigenous status could not be published (Supplementary RHD Table 9b).

Deaths among people with RHD

In 2016–2020, 476 deaths were reported among people with RHD in Queensland, Western Australia, South Australia and the Northern Territory. Of these,

- 322 people (68%) were Indigenous Australians
- 306 deaths occurred among females (64%)
- the median age at death was 52 for Indigenous males and 53 for Indigenous females, compared with 73 for both non-Indigenous males and females (Supplementary RHD Table 13).

In New South Wales, fewer than 5 deaths were reported among people with RHD.

Delivery of secondary prophylaxis to Indigenous Australians

In 2020, 4,521 Indigenous Australians were prescribed a treatment regimen to prevent recurrences of ARF, and progression to RHD, involving regular intramuscular injections of benzathine benzylpenicillin G (BPG) every 21 or 28 days (Supplementary Secondary Prophylaxis Table 1).

Proportion receiving BPG as prescribed

In 2020, among Indigenous Australians in Queensland, Western Australia, South Australia and the Northern Territory prescribed 3- or 4-weekly BPG:

- 13% (607 people) received 100% or more of their prescribed doses
- 22% (985) received 80% to 99% of their prescribed doses
- 29% (1,324) received 50% to 79% of their prescribed doses
- 36% (1,605) received less than 50% of their prescribed doses, including 418 people who received no doses (Supplementary Secondary Prophylaxis Table 1).

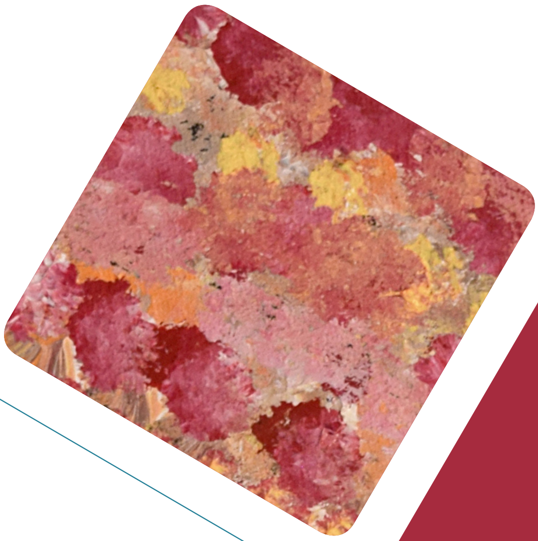
In 2020, among Indigenous Australians in New South Wales prescribed 3- or 4- weekly BPG:

- 30% (9 people) received at least 1 dose
- 70% (21) received no doses (Supplementary Secondary Prophylaxis Table 5b).

ARF recurrence rate among people on BPG

In 2020, among Indigenous Australians prescribed BPG in Queensland, Western Australia, South Australia and the Northern Territory, there were 131 ARF recurrences and a rate of 3.1 recurrences per 100 patient-years. Further information on the definition and calculation for patient-years can be found in Appendix B.

In 2020, among Indigenous Australians prescribed BPG in New South Wales, there were less than 5 ARF recurrences (Supplementary Secondary Prophylaxis Table 6).



1

1 Introduction

What is acute rheumatic fever?

Acute rheumatic fever (ARF) refers to an autoimmune response to an untreated infection of the throat and possibly skin by group A streptococcus (Strep A) bacteria (May et al. 2016; McDonald et al. 2004). Growing evidence shows that there are associations between Strep A skin infections and ARF (Bennett et al. 2019; Lorenz et al. 2021; Thomas et al. 2021; Wyber et al. 2021). Not all people who have a streptococcal infection develop ARF but, in those affected, it usually develops within 2-3 weeks of the infection (Webb et al. 2015).

ARF can affect the heart, joints, brain, and subcutaneous tissues (the innermost layers of skin) (Parnaby & Carapetis 2010). While no lasting damage is caused to the brain, joints, or skin, ARF can cause lasting damage to the heart. There is no single diagnostic test for ARF. Australian guidelines recommend hospitalisation so all necessary investigations are undertaken and to rule out other diagnoses.

The risk of ARF recurrence is relatively high after an initial episode. Repeated episodes increase the likelihood of long-term heart valve damage, known as rheumatic heart disease (RHD) (Carapetis et al. 2016). As each episode of ARF can worsen the damage to the heart, the priority in disease management is to prevent ARF recurrences using long-acting penicillin treatment, which is known as secondary prophylaxis.

What is rheumatic heart disease?

RHD refers to damaged heart valves as a result of 1 or more episodes of ARF. An affected heart valve can become scarred and/or stiff, obstructing blood flow (stenosis), or it can fail to close properly, causing blood to flow backwards in the heart instead of forward around the body (regurgitation). The mitral and aortic valves are most frequently affected. Regurgitation due to damage to the mitral valve is the most common feature of RHD.

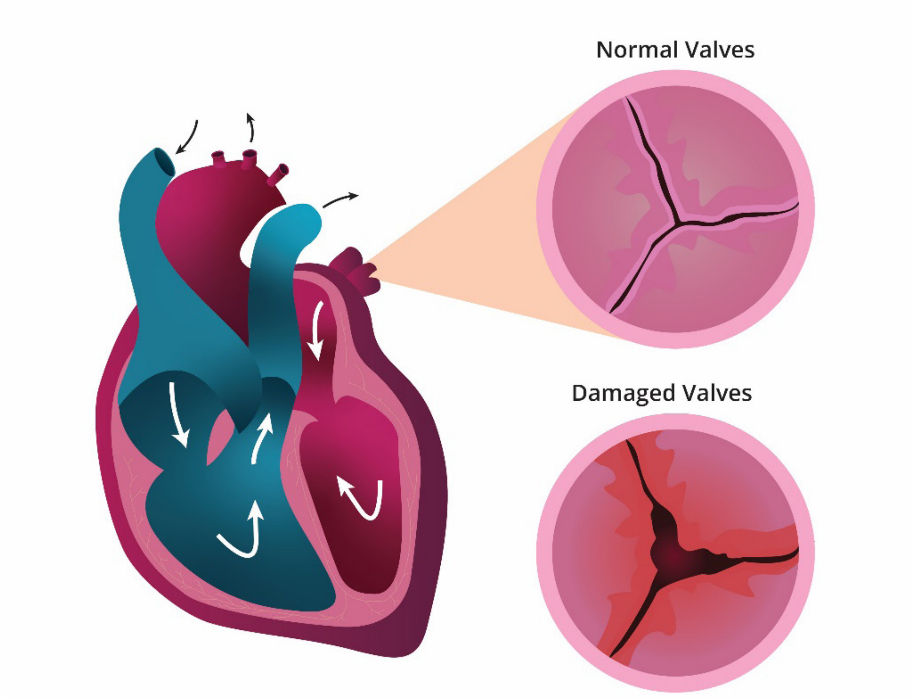
Figure 1.1 shows a diagram of the heart, comparing normal and damaged valves.

Symptoms of RHD include fatigue, chest pain, swelling of legs and face, and shortness of breath. Diagnosis can be difficult as symptoms are shared with other cardiac diseases.

The type of valve affected and severity of damage, along with a history of ARF, are important clinical indicators for RHD diagnosis. Many patients can remain asymptomatic despite having moderate or severe RHD. If left untreated, RHD can cause arrhythmias (heart beats too fast, too slow, or irregularly), stroke, endocarditis (infection of the inner lining of the heart or its valves), and complications of pregnancy, and may be fatal.

Management of RHD includes treating symptoms and preventing worsening of disease, which requires regular echocardiography to identify and monitor which valves are damaged and how badly. Management of an RHD diagnosis is complex and can involve coordination of multiple services and treatments such as primary health care, secondary prophylaxis with penicillin, monitoring of heart medications such as anticoagulation therapy, oral health care services, obstetrical and gynaecological services, echocardiography, specialist medical care, and other cardiothoracic and interventional cardiology services (RHD Australia 2012).

Figure 1.1: Diagram of the heart, comparing normal and damaged valves



Source: AIHW.

ARF and RHD are preventable diseases

ARF and RHD are both preventable diseases. They are common in low- and middle-income countries, and in socioeconomically disadvantaged populations in high-income countries (Wyber 2015; Webb et al. 2015). ARF and RHD are caused by aspects of socioeconomic disadvantage, such as household crowding, socioeconomic deprivation, low levels of functioning ‘health hardware’ (for example, toilets, showers, taps) and lack of access to health care services (Webb et al. 2015; Sims et al. 2016). Improved living conditions and access to functional health hardware can reduce high rates of Strep A infections (Katzenellenbogen et al. 2017).

Several opportunities exist to interrupt the disease pathway from Strep A infection to ARF and then RHD (Figure 1.2). Prevention measures that improve living conditions and environmental health and address eradication of group A streptococcal infections are primordial prevention measures.

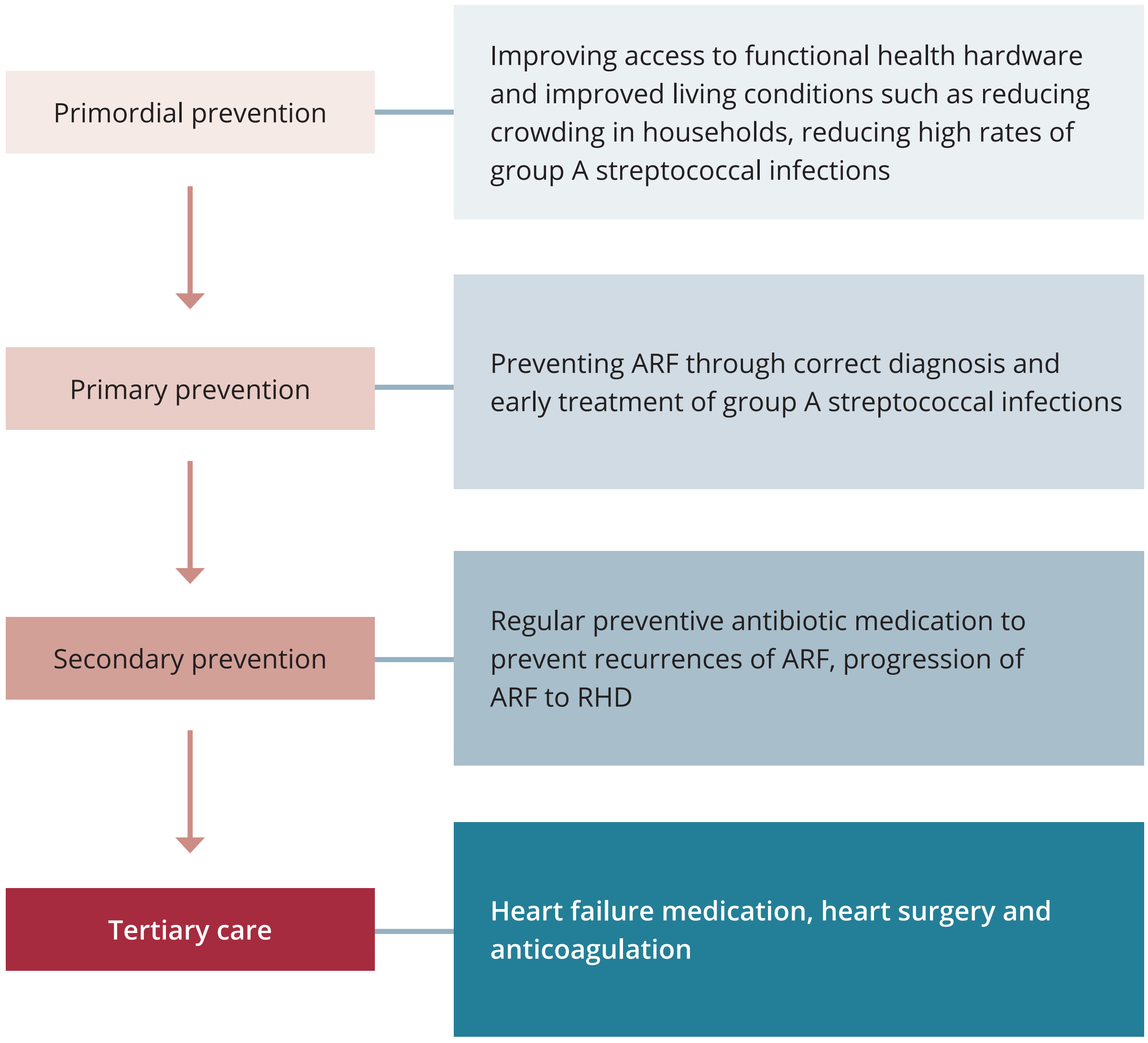
ARF is also preventable through early treatment of Strep A infections with penicillin. This is called primary prevention and relies on correct diagnosis and treatment of skin and throat infections with antibiotics as soon as possible after onset of symptoms. Timeliness of diagnosis and subsequent treatment can be negatively affected by health service access issues and delayed presentation to health services. The effectiveness of primary prevention is also compromised when the prescribed treatment does not comply with clinical guidelines (RHD Australia 2012).

Secondary prevention of the progression from ARF to RHD relies on correct diagnosis of ARF, to enable commencement of regular antibiotic preventive medication. Correct diagnosis is challenging as there is no specific single laboratory test for ARF, and it can be misdiagnosed. Diagnosis is based on clinical criteria outlined in the Australian modification of the Jones criteria, which take into account Australia’s high-risk groups (Technical information Table 4, and RHD Australia 2020 Chapter 6).

For people with suspected or clinically confirmed ARF episodes, benzathine benzylpenicillin G (BPG) is recommended every 21-28 days in order to prevent further Strep A infections and thereby reduce the risk of developing recurrent ARF. BPG prophylaxis is clinically effective and cost-effective for RHD control at both individual and community levels (Webb et al. 2015; Wyber & Carapetis 2015; RHD Australia 2020).

Tertiary care aims to slow disease progression and prevent complications associated with RHD and can include surgery to repair or replace damaged heart valves once RHD is established (Noonan 2020).

Figure 1.2: ARF and RHD prevention measures



Primordial prevention

Improving access to functional health hardware and improved living conditions such as reducing crowding in households, reducing high rates of group A streptococcal infections

Primary prevention

Preventing ARF through correct diagnosis and early treatment of group A streptococcal infections

Secondary prevention

Regular preventive antibiotic medication to prevent recurrences of ARF, progression of ARF to RHD

Tertiary care

Heart failure medication, heart surgery and anticoagulation

Championing change on rheumatic heart disease

Nearly 60 Aboriginal and Torres Strait Islander people are transforming understandings and experiences of RHD and ARF in their own communities and seeking change at the national and international level through the Champions4Change program, hosted by RHD Australia.

Champions4Change is the first program of its kind in Australia. It works to privilege and promote the voices of its champions, to support them in their lives and work, and to put culture, Country and community at the centre of responses to RHD.

The program emerged in 2018 from a review by RHD Australia on self-management of RHD. It identified a clear need for an Aboriginal and Torres Strait Islander-specific program that had self-determination and culture at its heart.

Its champions currently represent 27 communities across Australia and include people of all ages who are living with RHD and ARF, as well as parents and other family members, Aboriginal Health Practitioners, environmental health workers, education workers and others. Many have long held leadership roles in RHD and other health issues in their communities.

All are volunteers who are passionate for the cause and their communities. They work in multiple ways to support each other, and advocate for ending RHD and for health systems reform. They also design education and awareness programs for their own communities and in their own languages. Chief among their efforts is making sure that local culture, language and understandings shape local health messaging.

“Our champions are from the community. They have the cultural knowledge and the cultural protocols and lore. That means being able to provide strong health information in a culturally appropriate manner in a way that community understands.”

Vicki Wade, RHD Australia Director

Jurisdictional control programs and registers

Under the Rheumatic Fever Strategy, the Australian Government provides funding to support RHD control programs in 4 jurisdictions: Queensland, Western Australia, South Australia and the Northern Territory.

These programs are funded to support:

a. improved clinical care, including improved delivery of and adherence to secondary prophylaxis antibiotics

b. provision of education and training for health care providers, individuals, families and communities

c. collection and provision of agreed data annually to the Australian Institute of Health and Welfare (AIHW) for national monitoring and reporting of ARF and RHD and measuring program effectiveness in the detection and management of ARF and RHD

d. maintenance of a dedicated state-wide patient register and recall system for ARF and RHD.

Although an RHD control program and register also operates in New South Wales, this program is not currently covered under the Rheumatic Fever Strategy. The New South Wales register was established by the state government in 2016, with ARF and RHD becoming notifiable in the state in 2015, and RHD being notifiable only in persons aged under 35. Information on ARF and RHD diagnoses is based on notification data. Information on secondary prophylaxis is only collected for patients that consent to have this information collected on the register.

Information from the ARF/RHD registers in these 5 jurisdictions is compiled by the AIHW to provide information about ARF and RHD in Australia.

Box 1.1: Acute rheumatic fever/rheumatic heart disease registers

All jurisdictions with RHD registers have different notification and data collection practices and therefore the numbers, data quality and completeness in the RHD registers are variable. Table 1.1 summarises the timeline of program and register establishment across the jurisdictions.

Table 1.1: Timeline of program and register establishment

	NSW	SA	WA	Qld	NT	Vic, Tas, ACT
RHD control program	2015	2010	2009	2009	1997[a]	–
ARF/RHD register	2016	2012	2009	2014[b]	1997	–
Definite ARF notifiable	2015	2016	2007	1999	1996	–
Probable ARF notifiable	2015	2016	2015	1999	2019	–
Possible ARF notifiable	–	2016	2015	1999	–	–
Confirmed RHD notifiable	2015[c]	2016	2015	2018[d]	2019	–
Borderline RHD notifiable	–	2016	2015	2018	–	–

(a) The Top End Control Program was established in Darwin in 1997 and expanded in 2000 to include the whole Northern Territory.

(b) Prior to the current register, Queensland utilised the FERRET electronic patient record system for North Queensland Health facilities from 2009-2014.

(c) In NSW, RHD is notifiable only in persons aged under 35.

(d) In Queensland, RHD only became a notifiable condition on 1 September 2018.

Source: RHD Australia (ARF/RHD writing group) 2020.

About this report

This report presents information on ARF and RHD cases diagnosed or receiving treatment during 2016–2020, in the 5 jurisdictions maintaining RHD registers. Its aim is to provide an overview of ARF and RHD and so it focuses mainly on data for the combined jurisdictions. Accompanying web reports provide more detail at the individual state and territory level, and those reports are planned to be published shortly after this report.

Supplementary tables are available at: https://www.aihw.gov.au/reports/indigenous-australians/rheumatic-heart-disease-in-australia-2016-2020/data

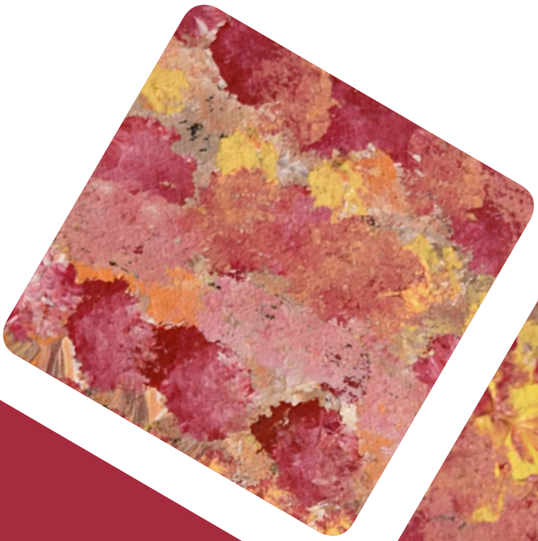
The 2020 Australian guideline for the prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease describes a set of indicators recommended for monitoring ARF and RHD. This fourth annual report presents data on those indicators that currently can be reported, including incidence and prevalence, characteristics of people with ARF and RHD, and delivery of secondary prophylaxis. It also includes data on the geographic distribution of people being managed for ARF and RHD, and the use of surgical interventions.

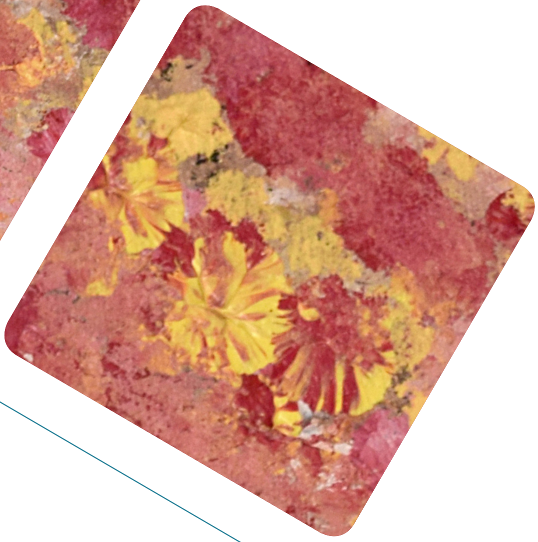
Data from NSW are presented in this report when definitions and reporting protocols are comparable to other jurisdictions, and in standalone data or figures where they are not comparable. More detail on this is provided below.

About the data in this report

The data used in this report are from the National RHD data collection, which is hosted and managed by the AIHW by collating and cleaning data from the ARF and RHD registers in the 5 jurisdictions to remove any duplications. Data in the collection are updated over time as the jurisdictional programs undertake data cleaning and quality improvement activity, so numbers in this report may not match those in previous reports. In addition, historical rates presented in this report have been calculated using revised Aboriginal and Torres Strait Islander population estimates based on the 2016 Census and should not be compared with those in previously published reports.

Throughout this report, some data from New South Wales will be incorporated with figures from other jurisdictions and some will remain separate, depending on comparability between the jurisdictions. New South Wales data were provided directly to the AIHW from the state's RHD register. ARF (all ages) and RHD (in persons under the age of 35 at diagnosis) became notifiable in New South Wales in October 2015, and the register was established in May 2016. For secondary prophylaxis, the register captures only patients who consent to their information being recorded on the register. People 35 years and older and people previously diagnosed outside New South Wales may be included on the register if it is felt worthwhile by their health practitioner. People who were under 35 at the time of RHD diagnosis remain on the register even after turning 35. Data for secondary prophylaxis were provided for 2017–2020.





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2 Overview of people on the registers

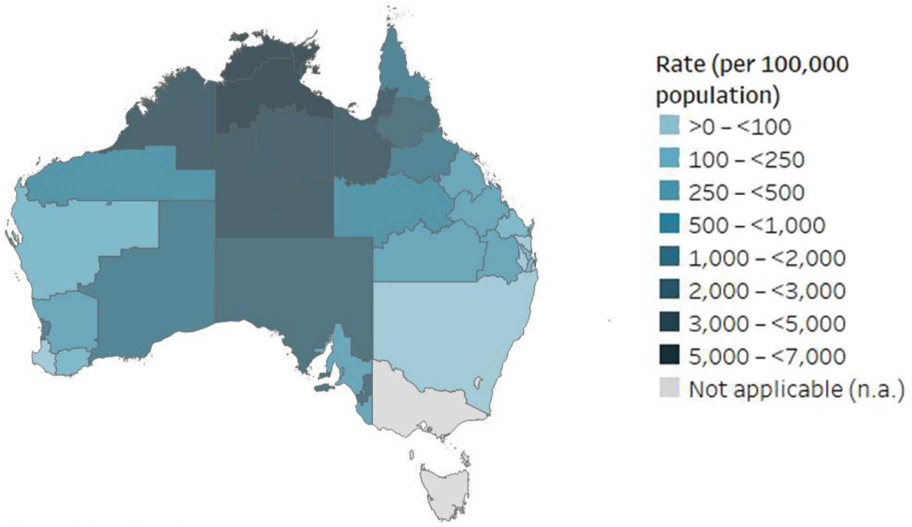
At 31 December 2020, there were 9,158 people on the RHD registers living with a diagnosis of ARF and/or RHD in New South Wales, Queensland, Western Australia, South Australia, and the Northern Territory. Of these:

- 2,886 (32%) people had ARF only recorded; 3,197 (35%) had only RHD recorded; and 3,075 (34%) had both ARF and RHD recorded (Supplementary Overview Table 1)
- the Northern Territory had the highest number of people living with ARF and/or RHD (3,805, 42%), followed by Queensland (3,622, 40%), Western Australia (1,215, 13%), South Australia (353, 3.9%) and New South Wales (163, 1.8%) (Supplementary Overview Table 2)
- Indigenous Australians accounted for 7,492 (82%) of the diagnoses compared with 1,550 (17%) diagnoses in non-Indigenous Australians (Supplementary Overview Table 2)
- more females (5,686, 62%) than males (3,470, 38%) were living with a diagnosis of ARF and/or RHD (Supplementary Overview Table 3).

For each person reported to a register, the region of management is recorded. This is the area where the patient was most recently reported to receive the majority of the primary health care for their ARF or RHD. The region of management may differ from the person's region of diagnosis and the notifying jurisdiction. The regions with the highest rates of management for Indigenous Australians were:

- Rural Darwin (NT), with 6,615 cases per 100,000 population (815 persons)
- East Arnhem (NT), with 6,332 per 100,000 (713 persons)
- Urban Alice Springs (NT), with 5,633 per 100,000 (384 persons, Figure 2.1) (Supplementary Overview Table 4).

Figure 2.1: ARF and/or RHD diagnoses among Indigenous Australians, by region of management, at 31 December 2020



Notes

1. Crude rates per 100,000 calculated using the number of notifications of each calendar year divided by the regional population for 2020 provided by the states or territory. Due to constraints in the jurisdictions, some previous populations were used when new populations could not be supplied. NSW, Qld, and NT populations are from 2020, SA populations are from 2018, and WA populations are from 2019.
2. Includes all Indigenous Australians managed for ARF and/or RHD in 2020.
3. The data excludes 510 ARF and/or RHD diagnoses nationally that had an unknown or other region of management.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

3

3 Acute rheumatic fever

This section discusses diagnoses of ARF reported by Australian RHD control programs between 2016 and 2020. The total number of ARF diagnoses recorded depends on the reporting practices to the various RHD registers. A person may have multiple diagnoses of ARF in their lifetime, so the number of diagnoses can be greater than the number of people affected. In this section of the report, cases are allocated to a jurisdiction and region based on where they were diagnosed.

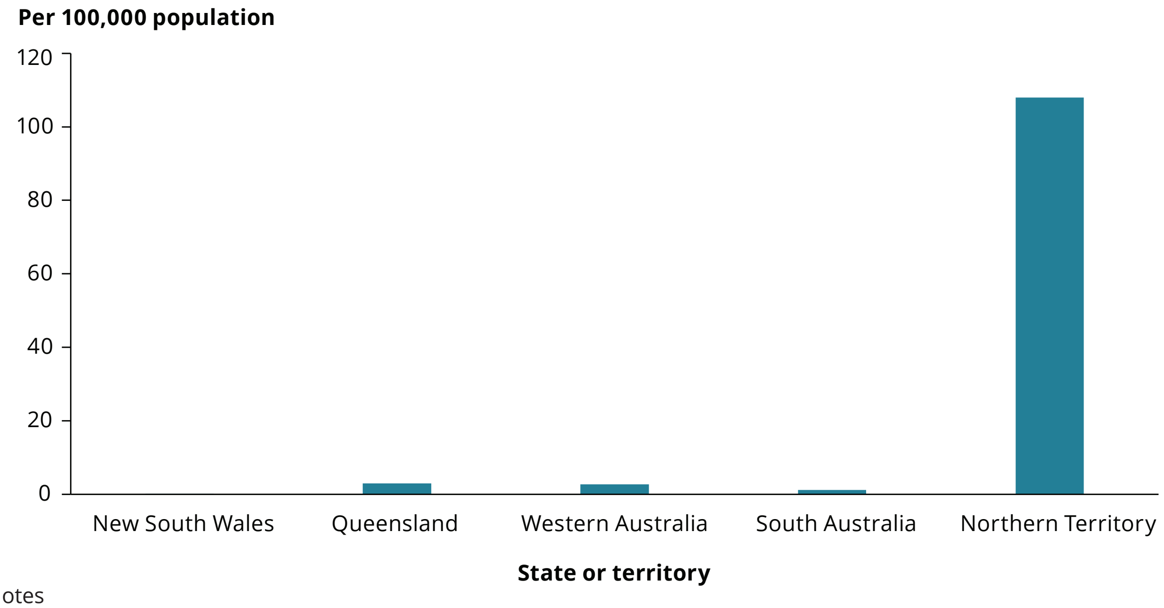
It is likely that ARF diagnoses are under-reported to RHD registers in all jurisdictions. A report from Agenson and others (2020) suggests that many cases of patients who attend the hospital for ARF or RHD are not reported to the jurisdictional registers. Although the registers in each state and territory were functional for the entire analysis period, they were relatively new in some states (see Table 1.1 in Introduction). Clinician awareness and reporting to the registers has likely increased in the years since register commencement and may also have been affected by the addition of ARF and RHD to the list of notifiable diseases at different times in the various jurisdictions. However, under-diagnosis and under-reporting to the register also mean some individuals are not captured in this analysis. It is difficult to determine whether increases in the number of notifications reflect a real increase in the number of cases occurring, improved detection and diagnosis of cases, increases in the number of people being recorded on the registers, or a combination of these.

Among all Australians

In 2016–2020:

- 2,611 diagnoses of ARF were recorded in New South Wales, Queensland, Western Australia, South Australia and the Northern Territory combined (incidence rate 3.0 per 100,000 population over the 5 years combined). These diagnoses occurred in 2,380 Australians (Supplementary ARF tables 1 and 2)
- the Northern Territory had the highest rate of ARF diagnoses (108 per 100,000 population; 1,331 diagnoses), followed by Queensland (3.0 per 100,000; 748 diagnoses), Western Australia (2.7 per 100,000; 352 diagnoses), South Australia (1.1 per 100,000 population; 100 diagnoses) and New South Wales (0.2 per 100,000 population; 80 diagnoses) (Figure 3.1) (Supplementary ARF Table 2).

Figure 3.1: ARF notifications among all Australians, by state and territory, 2016–2020



Notes

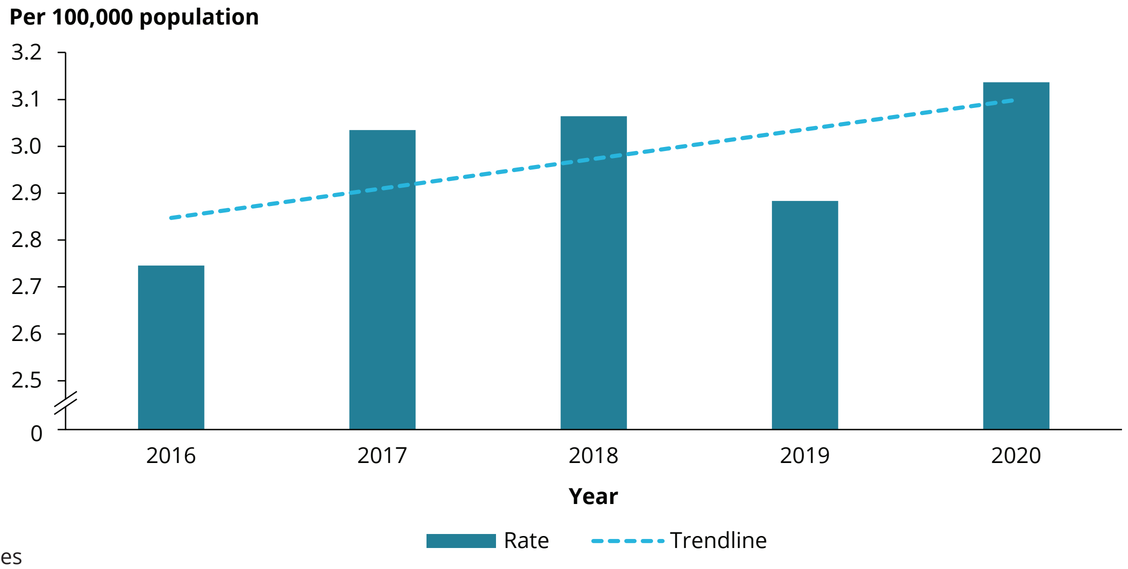
1. Crude age-specific rates per 100,000 calculated using the number of notifications of each calendar year divided by the corresponding 30 June populations of each year based on the 2016 Census.
2. ARF notifications include all recurrence categories and diagnostic categories.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Time trend

The number and rate of ARF diagnoses increased from 469 (2.7 per 100,000) in 2016 to 565 (3.1 per 100,000) in 2020. There was a decrease in cases in 2019, to 513 cases (2.9 per 100,000) (Figure 3.2) (Supplementary ARF Table 3).

Figure 3.2: ARF notifications among all Australians, by year, 2016–2020



Notes

1. ARF notifications include all recurrence categories and diagnostic categories.
2. Data for New South Wales, Queensland, Western Australia, South Australia and the Northern Territory combined.
3. Crude age-specific rates per 100,000 calculated using the number of notifications of each calendar year divided by the corresponding 30 June populations of each year based on the 2016 Census.

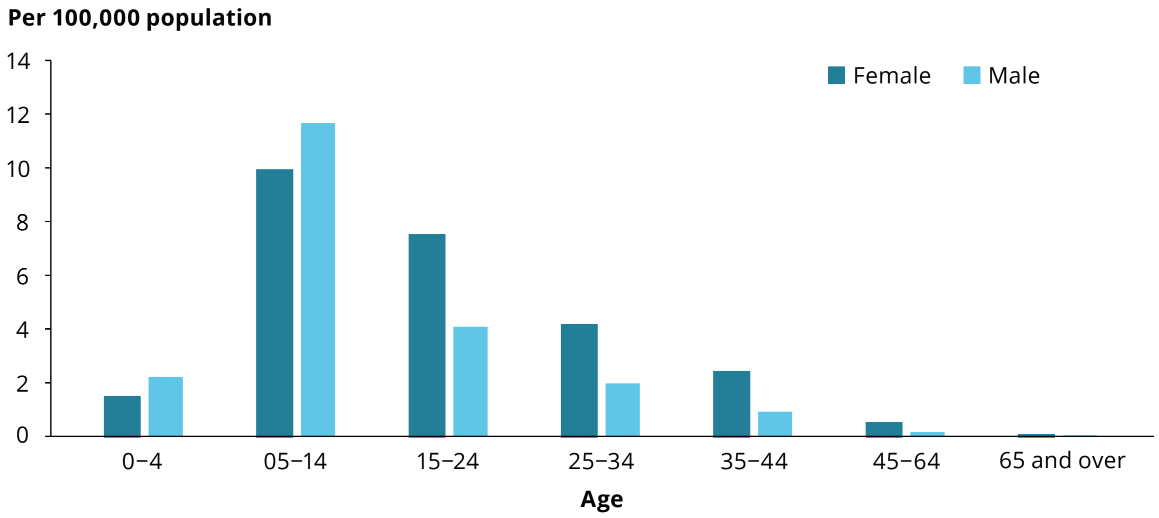
Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Age and sex

Of the 2,611 ARF diagnoses among all Australians in 2016–2020:

- the most common age at diagnosis was 5–14, with 1,200 diagnoses (11 per 100,000 population)
- there were 102 diagnoses (1.8 per 100,000) among children aged 0-4
- females accounted for 1,446 (55%) diagnoses
- in people aged under 15, ARF rates were generally higher among males than females
- for adults, ARF rates were generally higher among females than males (Figure 3.3) (Supplementary ARF Table 4a).

Figure 3.3: ARF notifications among all Australians, by sex and age group, 2016–2020



Notes

1. Data from NSW, Qld, SA, WA, and NT combined.
2. Crude age-specific rates per 100,000 calculated using the number of notifications of each calendar year divided by the corresponding 30 June populations of each year based on the 2016 Census.
3. ARF notifications include all recurrence categories and diagnostic categories.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Indigenous Australians

In 2016–2020:

- 2,392 Aboriginal and Torres Strait Islander people, in New South Wales, Queensland, Western Australia, South Australia and the Northern Territory, were diagnosed with ARF, a rate of 65 per 100,000 population
- the Northern Territory had the highest rate of diagnoses (344 per 100,000 population; 1,310 diagnoses), followed by Western Australia (65.1 per 100,000 population; 340 diagnoses), Queensland (53 per 100,000 population; 615 diagnoses), South Australia (43 per 100,000; 95 diagnoses) and New South Wales (2.3 per 100,000 population; 32 diagnoses) (Figure 3.4) (Supplementary ARF Table 2).

Figure 3.4: ARF notifications among Indigenous Australians, by state and territory, 2016–2020

Notes

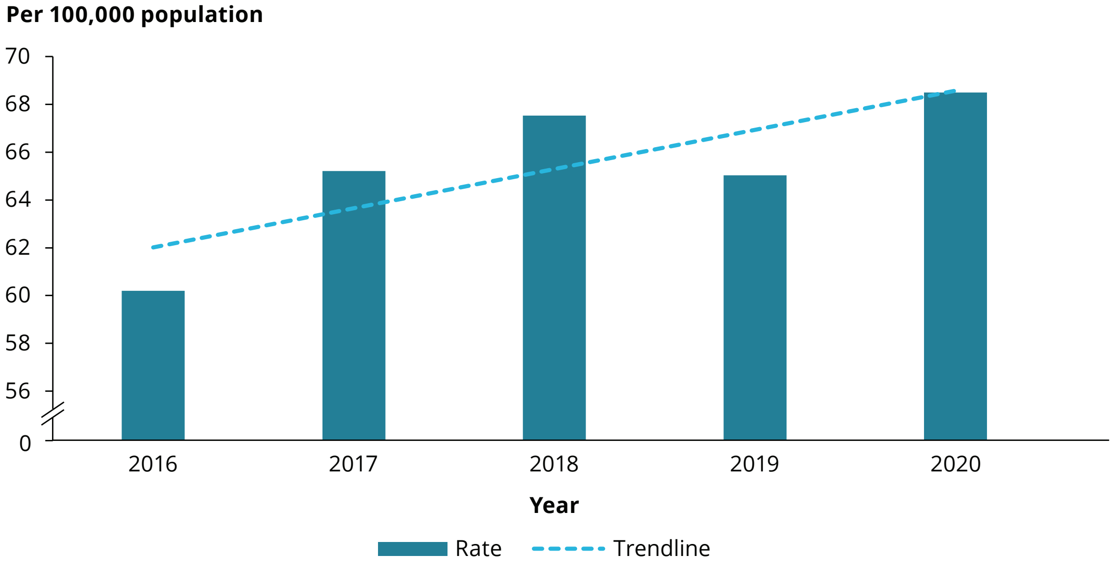
1. Crude age-specific rates per 100,000 calculated using the number of notifications of each calendar year divided by the corresponding 30 June populations of each year based on the 2016 Census.
2. ARF notifications include all recurrence categories and diagnostic categories.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Time trend

The ARF diagnosis rate among Indigenous Australians increased between 2016 and 2018 – from 60 to 68 per 100,000 population (424 to 494 diagnoses, respectively) – but then dipped to 65 per 100,000 (485 diagnoses) in 2019 before rising in 2020 to 69 per 100,000 (521 diagnoses) (Figure 3.5) (Supplementary ARF Table 3).

Figure 3.5: ARF notifications among Indigenous Australians, by year, 2016–2020



Notes

1. ARF notifications include all recurrence categories and diagnostic categories.
2. Data for NSW, Qld, WA, SA and the NT combined.
3. Crude age-specific rates per 100,000 calculated using the number of notifications of each calendar year divided by the corresponding 30 June populations of each year based on the 2016 Census.

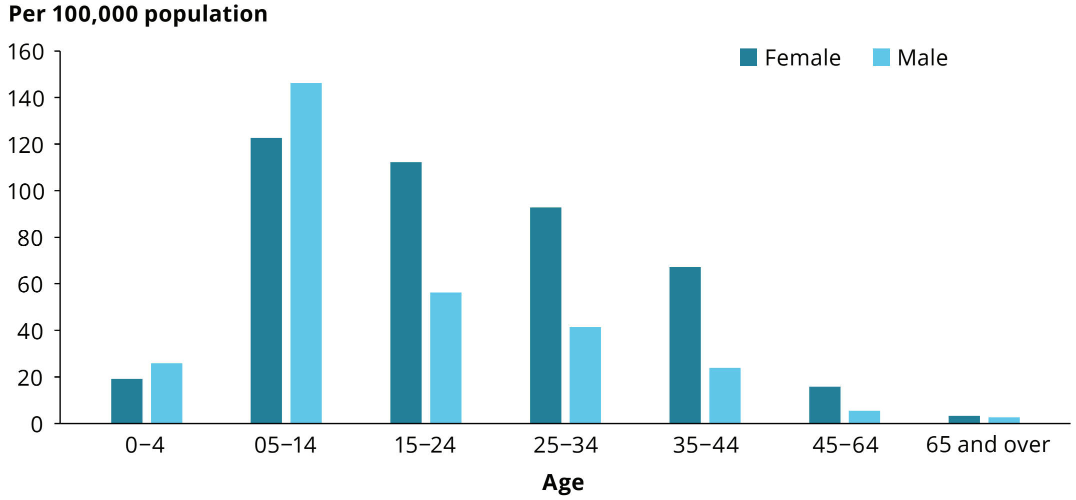
Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Age and sex

Of the 2,392 ARF diagnoses among Indigenous Australians in 2016–2020:

- the highest rate of diagnosis was among those aged 5–14, accounting for 46% of all diagnoses (135 per 100,000 population, or 1,094 diagnoses)
- there were 95 diagnoses (23 per 100,000) among children aged 0–4
- females accounted for 56% of diagnoses (1,350 diagnoses)
- in people aged under 15, rates were generally higher among males than females.
- for adults, rates were generally higher among females than males (Figure 3.6) (Supplementary ARF Table 4b).

Figure 3.6: ARF notifications among Indigenous Australians, by sex and age group, 2016–2020



Notes

1. Data from NSW, Qld, SA, WA, and NT combined.
2. Crude age-specific rates per 100,000 calculated using the number of notifications of each calendar year divided by the corresponding 30 June populations of each year based on the 2016 Census.
3. ARF notifications include all recurrence categories and diagnostic categories.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Due to the relatively small proportion of non-Indigenous Australians diagnosed with ARF, the remainder of this chapter (with the exception of the section on deaths) will focus on Indigenous Australians with ARF.

Region of diagnosis

For each ARF notification, region of diagnosis is recorded on the RHD register. In most cases the place where infection was acquired cannot be determined. ARF cases were assigned to their diagnosis state or territory, and region for this analysis.

Each state or territory defines regions uniquely, based on its own specific health services boundaries. There are 33 regions spread over Queensland, Western Australia, South Australia, and the Northern Territory. Regions do not cross state and territory boundaries.

For this reporting cycle, NSW did not provide information by region, and is considered as a whole.

In 2016–2020, the Northern Territory accounted for 55% of the 2,392 ARF diagnoses among Aboriginal and Torres Strait Islander people (1,310 diagnoses). Queensland accounted for a further 26% of cases, Western Australia for 14%, South Australia for 4.0%, and New South Wales for 1.3% (Supplementary ARF Table 2).

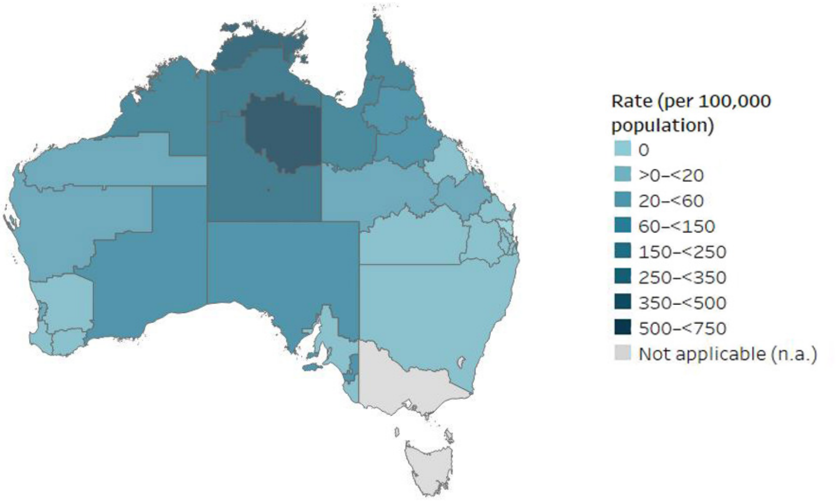
In 2016–2020, the highest 3 rates of ARF diagnoses were reported in regions in the Northern Territory:

- Urban Alice Springs with 706 per 100,000 (241 diagnoses)
- Barkly with 514 per 100,000 population (112 diagnoses)
- Rural Darwin with 496 per 100,000 population (307 diagnoses) (Figure 3.7) (Supplementary ARF Table 5).

In 2016–2020, the 3 regions with the highest numbers of ARF diagnoses were also in the Northern Territory:

- Rural Darwin with 307 diagnoses
- Urban Alice Springs with 241 diagnoses
- East Arnhem with 236 diagnoses (Figure 3.7) (Supplementary ARF Table 5).

Figure 3.7: ARF diagnoses among Indigenous Australians, by region of diagnosis, 2016–2020



Notes

1. Crude rates per 100,000 calculated using the number of notifications of each calendar year divided by the population provided by the state or territory for each region per year. Due to constraints in the jurisdictions, some previous populations were used when new populations were unable to be supplied. NSW, Qld, and NT populations have up-to-date data. SA uses the 2018 populations as a proxy for 2019 and 2020. WA uses 2019 populations as a proxy for 2020 data.
2. ARF notifications include all recurrence categories and diagnostic categories.
3. The data exclude 73 ARF diagnoses nationally that had an unknown region of diagnosis.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Symptoms and diagnosis (manifestations)

Diagnosing ARF can be challenging as there is no single diagnostic laboratory test – diagnosis is based on clinical decisions plus supporting laboratory evidence. The Jones diagnosis criteria were introduced in 1944 and have been periodically modified and updated in the Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (RHD Australia 2020). The Jones criteria specify the manifestations, counting rules and criteria which determine a diagnosis of ARF. Specific manifestations occurring in ARF that are reliably collected by jurisdictions and are related to an increased risk of RHD are presented in this report. These manifestations are: carditis, Sydenham chorea, and prolonged P-R interval (Box 3.1). People with carditis, a prolonged P-R interval and/or AV junctional arrhythmias are more likely to sustain heart damage (and hence to develop RHD) than those without.

Box 3.1: ARF manifestations associated with an increased risk of RHD

Carditis: inflammation of the heart muscle and heart tissue, including the membrane which lines the chambers of the heart and forms the surface of the heart valves (endocardium). It causes a rapid heart rate, fatigue, shortness of breath and exercise intolerance, and in ARF is associated primarily with the mitral valve. Carditis occurs in about 40%–50% of people with ARF.

Prolonged P-R interval and/or AV junctional arrhythmias: detected through electrocardiography. Refers to when the time between specific electrical features of a heartbeat is longer than expected. Often the person has no symptoms.

Sydenham chorea: a neurological disorder of childhood resulting from infection with group A beta-haemolytic streptococcus, the bacterium that causes rheumatic fever. It is characterised by involuntary movements of the hands, feet, tongue and face, which stop during sleep. This is more common in females; globally it affects up to 36% of cases, and is associated with carditis.

A complete list of major and minor manifestations of ARF is provided in the Australian guideline for prevention, management and diagnosis of ARF and RHD and in Table A1 of this report.

Source: RHD Australia 2020.

In 2016–2020, of the 2,392 ARF diagnoses among Indigenous Australians, 36% had at least 1 manifestation of carditis, prolonged P-R interval, or Sydenham chorea (Supplementary ARF Table 6). The inclusion of Sydenham chorea in this value may cause an overestimation of the number of cases at higher risk of progressing to RHD, as it is most often associated with the development of carditis rather than directly with RHD.

In 2016–2020, of all 2,392 ARF diagnoses among Indigenous Australians:

- 1,932 diagnoses were definite or probable diagnoses (Box 3.2)
- 460 were possible diagnoses (Supplementary ARF Table 7).

Note that probable ARF has been notifiable in Northern Territory only since 2019 and possible ARF is not notifiable in New South Wales or the Northern Territory (See Table 1.1 in Introduction), so these cases may not necessarily be recorded on registers.

Box 3.2: ARF diagnostic categories

There is no 1 specific diagnostic test for ARF. Instead, it is diagnosed based on medical history and a pattern of clinical features ('manifestations') as follows:

Definite ARF, first episode: 2 major or 1 major and 2 minor manifestations plus evidence of preceding Strep A infection.

Definite ARF, recurrent episode: 2 major or 1 major and 1 minor manifestations or 3 minor manifestations plus evidence of preceding Strep A infection. (The 2020 RHD Australian guideline increased the manifestation requirement to 1 major and 2 minor manifestations plus preceding Strep A to confirm diagnosis.)

Probable ARF: clinical presentation falls short by either 1 major or 1 minor manifestation, or the absence of streptococcal serology results, but where ARF is the most likely diagnosis.

Possible ARF: Strong clinical suspicion of ARF, but insufficient signs and symptoms for diagnosis of definite or probable ARF.

These definitions applied when the majority of the data in this report were collected, and have been updated in the most recent clinical guidelines. The new guidelines were released in early 2020 and, where changed, have been added in parentheses.

Source: RHD Australia 2012.

Between 2016 and 2020:

- the rate of definite or probable ARF diagnoses reached a peak of 56 per 100,000 (404 diagnoses) in 2017 and has been falling since then, to 51 per 100,000 (388 diagnoses) in 2020
- the rate of possible ARF episodes increased, from 9.1 per 100,000 (64 diagnoses) in 2016 to 18 per 100,000 (133 diagnoses) in 2020
- the proportion of ARF diagnoses which were definite or probable decreased, from 85% (360 diagnoses) in 2016 to 75% (388) in 2020 – with a corresponding increase in the proportion of possible diagnoses. This may relate to possible ARF diagnoses becoming notifiable in Western Australia and South Australia in 2015 and 2016, respectively (Supplementary ARF Table 7).

Recurrences

Box 3.3: ARF recurrence status definitions

First known episode: a reported ARF episode (definite, probable, or possible) in an individual with no known past ARF or RHD.

Recurrent episode: a reported ARF episode (definite, probable, or possible) in an individual with known past ARF or RHD.

First known and recurrent ARF episodes are preventable (Box 1.2). After the first known ARF episode, adherence to secondary prophylaxis reduces the likelihood of recurrence.

Of the 2,611 ARF diagnoses between 2016 and 2020 among all Australians in New South Wales, Queensland, Western Australia, South Australia and the Northern Territory combined, 697 (27%) were recurrent cases.

Of the 2,392 ARF diagnoses among Indigenous Australians in these jurisdictions, 663 (28%) were recurrent cases (Supplementary ARF Table 8).

Further analysis on recurrence rates among Indigenous Australians on prophylaxis can be found in 'Chapter 5: Secondary prophylaxis'.

Deaths among all Australians

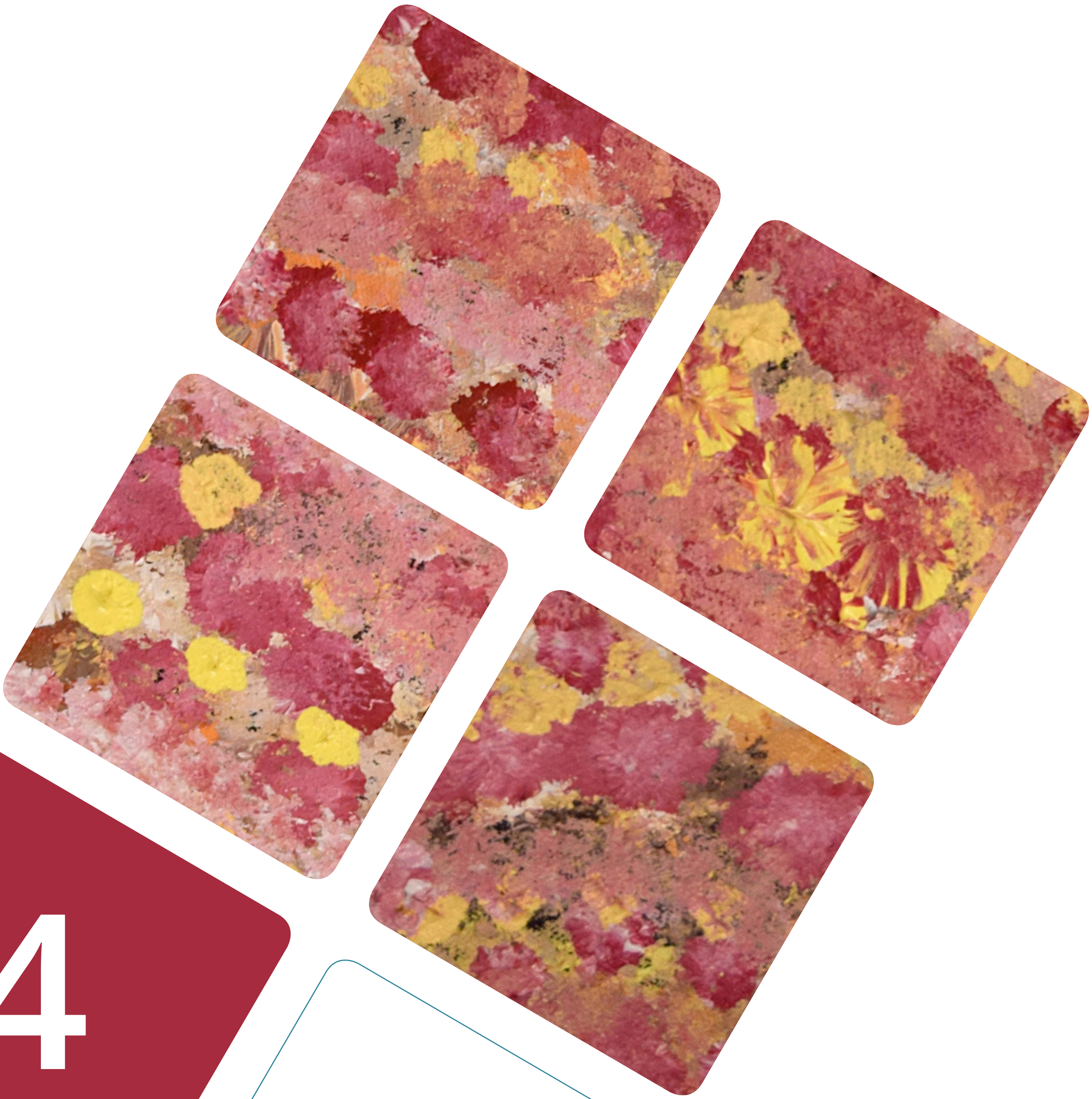
In 2016–2020, 153 deaths were recorded among people with a diagnosis of ARF only on the RHD registers (people with both ARF and RHD diagnoses were analysed as RHD deaths; Table 3.1). These individuals could have died from any cause, as detailed cause of death information is not captured on most registers. Among Aboriginal and Torres Strait Islander people, 146 deaths were recorded on these registers. The median age at death for Indigenous Australians with ARF was 40, with 86 (59%) of those who died aged under 45 (Supplementary ARF tables 9a and 9b).

Table 3.1: Deaths among people with ARF recorded on jurisdictional registers, by Indigenous status, 2016–2020

Age group	Indigenous	All Australians
0–24	20	20
25–44	66	111
45–64	n.p.	n.p.
65+	n.p.	n.p.
All ages	**146**	**153**

Note: n.p. means not publishable due to low numbers.

Source: AIHW analysis of the National Rheumatic Heart Disease data collection.



4

4 Rheumatic heart disease

The National Rheumatic Heart Disease data collection includes information about diagnoses of RHD recorded in each jurisdiction (Table 1.1). It is important to note the following:

- The total number of RHD diagnoses recorded depends on each state and territory's reporting practices, both historically and presently.
- The commencement year of each register varies, and RHD has become notifiable at different times in each jurisdiction (refer to Table 1.1).
- A person can have only 1 diagnosis of RHD, though they may be registered in more than 1 jurisdiction as they can receive care in different places. For the national data collection, each diagnosis was assigned to only 1 jurisdiction, based on location for primary health care at the time the data were submitted**.**

RHD is notifiable only in those aged under 35 at the time of diagnosis in New South Wales, but it is notifiable for all ages for the other 4 jurisdictions. As New South Wales uses different inclusion criteria it is not comparable to the other 4 jurisdictions and so results for New South Wales are shown separately.

Prevalence among all Australians

RHD in Queensland, Western Australia, South Australia and the Northern Territory

At 31 December 2020, 6,039 people (61 per 100,000 population) were recorded as having RHD on registers in Queensland, Western Australia, South Australia, and the Northern Territory (Supplementary RHD Table 1). Of these:

- 78% were Indigenous Australians (4,696 diagnoses, 991 per 100,000 population)
- 27% were aged under 25 at 31 December 2020 (1,641 diagnoses) – with fewer than 5 aged under 5
- 67% were female (4,017 diagnoses)
- the Northern Territory had the highest prevalence rate (2,305 diagnoses, 936 per 100,000) (Supplementary RHD tables 1 and 2a).

Priority status for RHD diagnoses in Queensland, Western Australia, South Australia and the Northern Territory

An individual's priority status (Table 4.1) determines the recommended care plan and schedule given their clinical and personal needs. This status may change over time as their condition and needs change. Priority definitions changed from the 2012 guidelines to the 2020 guidelines and some people now require ongoing management that they did not need based on the 2012 guidelines. Both definitions are explained in Table 4.1. The time of application of these changes may vary between jurisdictions.

Table 4.1: Definitions of RHD priority status

	2012 Guideline	2020 Guideline
Priority 1	• Severe valvular disease or • Moderate/severe valvular lesions with symptoms or • Mechanical prosthetic valves; tissue prosthetic valves & valve repairs including balloon valvuloplasty	• Severe RHD of any valve or • High risk post-valve surgical patients or • ≥ 3 episodes of ARF within the last 5 years or • Pregnant women with RHD (of any severity) may be considered Priority 1 for the duration of the pregnancy or • Children ≤ 5 years of age with ARF or RHD
Priority 2	Any moderate valve lesion in the absence of symptoms and with normal LV function	Moderate RHD of any valve or Mild RHD involving both aortic and mitral valves or Moderate risk post-valve surgical patients
Priority 3	• ARF with no evidence of RHD or • Trivial to mild valvular disease	• Mild RHD involving only a single valve or • ARF (probable or definite), currently prescribed secondary prophylaxis or • Borderline RHD currently prescribed secondary prophylaxis or • Low risk post-valve surgical patients
Priority 4	• Patients with a history of ARF (no RHD) for whom secondary prophylaxis has been ceased	• History of ARF (possible, probable or definite) and completed secondary prophylaxis or • Borderline RHD not on secondary prophylaxis or • Resolved RHD and completed secondary prophylaxis

Source: Adapted from The Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease 2020.

Of the 3,895 people diagnosed with RHD with a priority status recorded for their most recent evaluation:

- 22% were priority 1 (864 people), 24% were priority 2 (914), 49% were priority 3 (1,892), and 225 people (5.8%) were priority 4
- older people were more likely to have priority 1 RHD, with 35% aged 45 or over having priority 1 disease (285 people), compared with 13% of those aged 0–14 (87 people).

Too few people aged 0–4 diagnosed with RHD had a priority status recorded to make conclusions about priority status for that age group. By the 2020 guideline, these are all priority 1, but they may have been assessed before that guideline went into effect (Supplementary RHD Table 3a).

RHD in New South Wales

At 31 December 2020, 98 people (2.4 per 100,000 population) were recorded on the New South Wales register. This may not be representative of all Australians with RHD in New South Wales as RHD is notifiable only in those under 35 at the time of diagnosis. Of these:

- 31% (30 diagnoses, 14.5 per 100,000 population) were Indigenous and 67% (66 cases, 1.6 per 100,000) were non-Indigenous
- the median age on the register at the end of 2020 was 22 (Supplementary RHD Table 2b).

Priority status of RHD diagnoses in New South Wales

Thirteen diagnoses of RHD had a current priority status recorded. About half (54%, 7 cases) were priority 3 (Supplementary RHD Table 3b).

Incidence of RHD in 2016–2020

In this report, incidence (a 'new' RHD diagnosis) is defined as one that was diagnosed between 1 January 2016 and 31 December 2020. In most cases, it is not possible to identity a year of onset for RHD as the condition may be asymptomatic initially. The analysis is based on year of diagnosis.

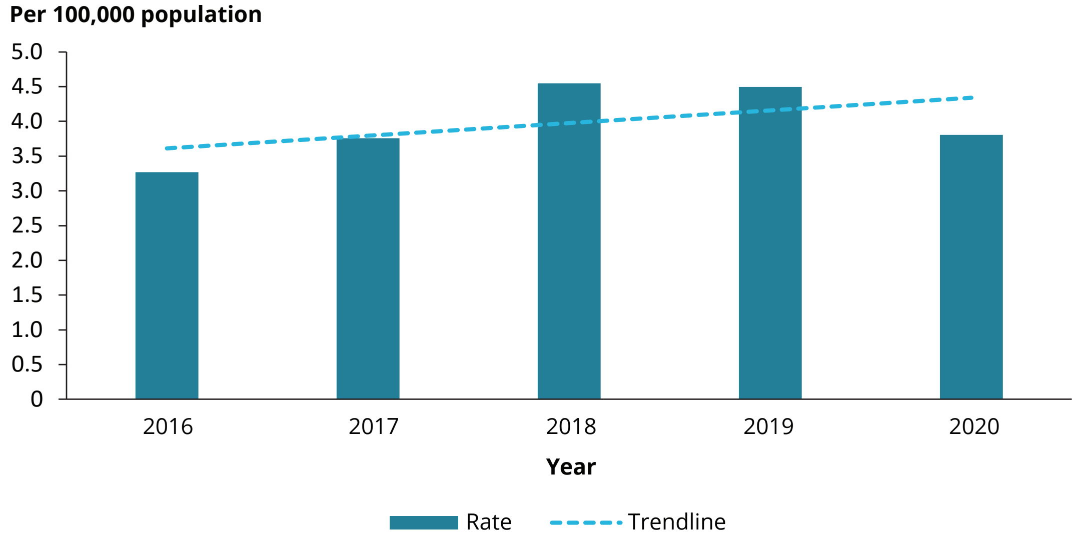
New diagnoses in Queensland, Western Australia, South Australia and the Northern Territory

All Australians

In 2016–2020:

- there were 1,910 reports of new RHD diagnoses in Queensland, South Australia, Western Australia and the Northern Territory (4.0 per 100,000 population)
- diagnosis rates increased between 2016 and 2018 from 3.3 to 4.5 diagnoses per 100,000. Since then, the rate has fallen to 3.8 per 100,000 in 2020 (Figure 4.1)
- diagnosis rates varied by state and territory. The Northern Territory had 47 diagnoses per 100,000, followed by Queensland (3.8 diagnoses per 100,000) and Western Australia (2.4 diagnoses per 100,000). South Australia had less than 1 diagnosis per 100,000 (Figure 4.2) (Supplementary RHD Table 4a).

Figure 4.1: New RHD diagnoses, all Australians in Qld, WA, SA and NT, by year, 2016–2020



Notes

1. Crude age-specific rates per 100,000 calculated using the number of notifications of each calendar year divided by the corresponding 30 June populations of each year based on the 2016 Census.
2. 25 people with an unknown or blank Indigenous status are included in All Australians.
3. Data from Qld, WA, SA and NT combined.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Figure 4.2: New RHD diagnoses, all Australians, by state and territory, 2016–2020

Note: Crude age-specific rates per 100,000 calculated using the number of notifications of each calendar year divided by the corresponding 30 June populations of each year based on the 2016 Census.

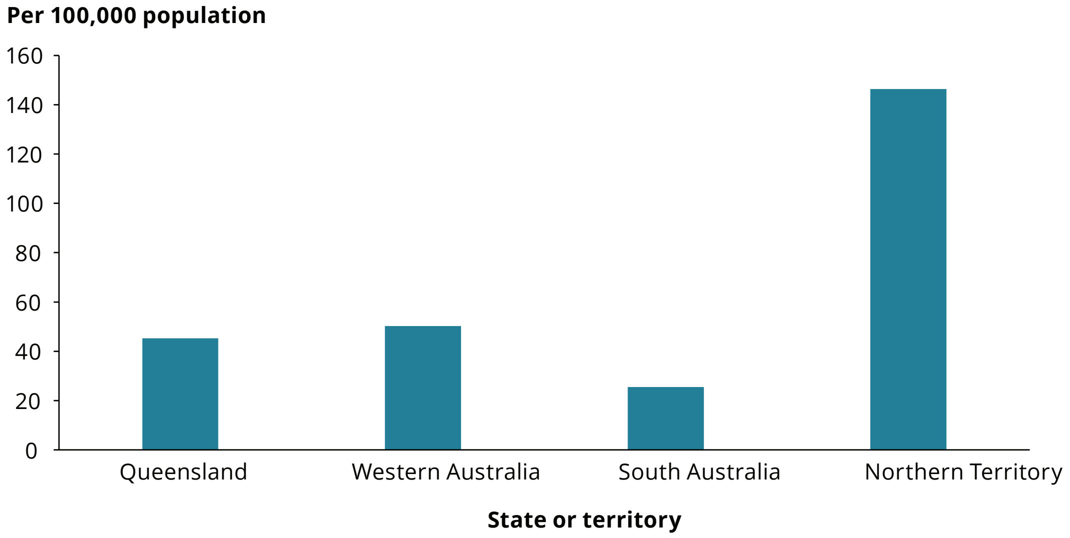
Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Indigenous Australians

In 2016–2020:

- of the 1,910 new RHD diagnoses among all Australians in Queensland, Western Australia, South Australia and the Northern Territory, 73% (1,399) were Indigenous Australians (61 per 100,000 population)
- the overall diagnosis rate for Indigenous Australians was more than 15 times the rate for all Australians (61 per 100,000, compared with 4.0 per 100,000, respectively)
- the Northern Territory had the highest rate of new RHD diagnoses among Indigenous Australians (146 per 100,000 population) followed by Western Australia (50 per 100,000; Figure 4.3), Queensland (45) and South Australia (26)
- the annual combined rate increased from 51 to 73 per 100,000 (222 diagnoses to 332 diagnoses) between 2016 and 2018, then decreased to 59 per 100,000 in 2020 (279 diagnoses) (Figure 4.4). It is possible that the decrease is related to reduced access to echocardiography as a result of COVID-19 restrictions, and is not necessarily reflective of a true reduction in disease incidence (Supplementary RHD tables 4a and 5a).

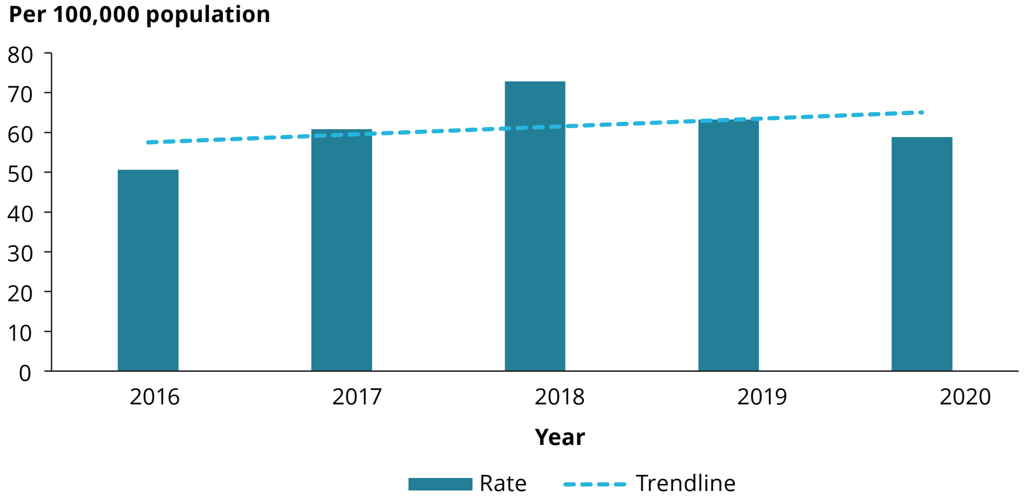
Figure 4.3: New RHD diagnoses, Indigenous Australians, by state and territory, 2016–2020



Note: Crude age-specific rates per 100,000 calculated using the number of notifications of each calendar year divided by the corresponding 30 June populations of each year based on the 2016 Census.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Figure 4.4: New RHD diagnoses, Indigenous Australians, Qld, WA, SA and NT, by year, 2016–2020



Notes

1. Crude age-specific rates per 100,000 calculated using the number of notifications of each calendar year divided by the corresponding 30 June populations of each year based on the 2016 Census.
2. Data from Qld, WA, SA and NT combined.

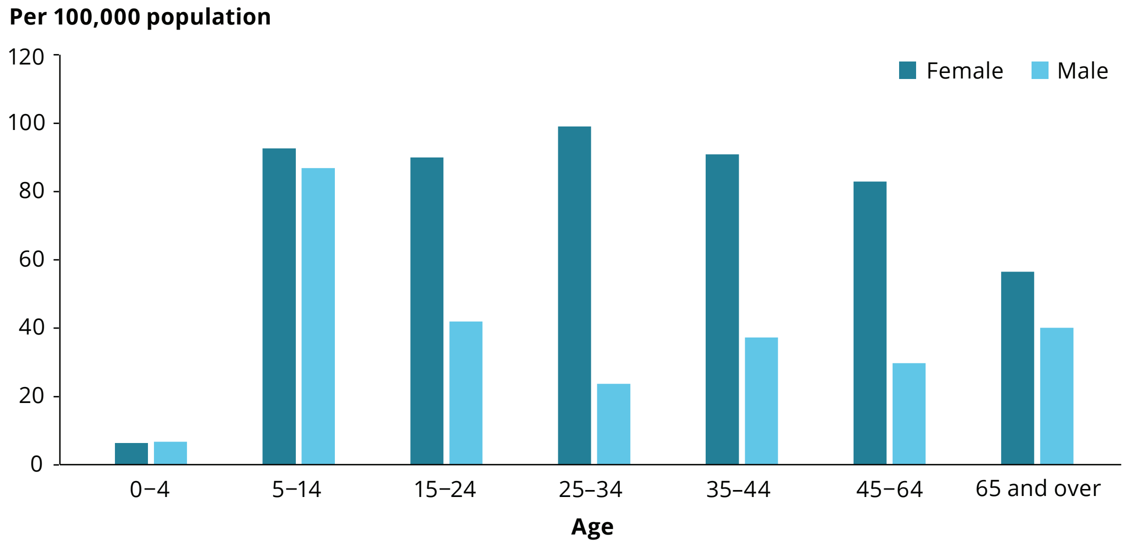
Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Sex and age

In 2016–2020, for all new RHD cases diagnosed among Indigenous Australians:

- the rate of new RHD diagnosis for females was 1.9 times that for males (43 and 80 diagnoses per 100,000 population, respectively)
- females had higher rates than males in all age groups, excluding those aged 0–4 which had too few cases to draw conclusions
- 54% were aged under 25 at diagnosis (754 people)
- 17 children were aged under 5 and 451 children were aged 5–14 at the time of RHD diagnosis (Figure 4.5)
- The median age at diagnosis for Indigenous Australians was 23 (age 15 for males and 27 for females), compared with age 54 for non-Indigenous Australians (Supplementary RHD tables 5a and 6).

Figure 4.5: New RHD diagnoses, Indigenous Australians, by age group and sex, 2016–2020



Notes

1. Crude age-specific rates per 100,000 calculated using the number of notifications of each calendar year divided by the corresponding 30 June populations of each year based on the 2016 Census.
2. Data from Qld, WA, SA and NT combined.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Severity of RHD at time of diagnosis among Indigenous Australians

Severity is collected at the time of diagnosis and can be categorised as severe, moderate, mild, or borderline, determined by a cardiologist. Table 4.2 lists the definitions of each status. In previous reports, severity and priority were combined to reflect the patient's current status and that was called severity. As such, previous reports do not have comparable data related to severity. Borderline RHD data were not available for this report.

For this report, Queensland was unable to provide severity of RHD at diagnosis.

In 2016–2020, of the 630 Indigenous Australians with severity recorded at a new RHD diagnoses:

- 56% had mild RHD when first diagnosed (353 diagnoses)
- 30% had moderate RHD (191)
- 14% had severe RHD (86) (Supplementary RHD Table 7).

Table 4.2: Definitions of RHD severity status

Severe	Severe valvular disease Or moderate/severe valvular lesions with symptoms Or mechanical prosthetic valves; tissue prosthetic valves & valve repairs including balloon valvuloplasty
Moderate	Any moderate heart valve damage without symptoms, and with normal left ventricle function.
Mild	Trivial to mild valvular disease
Borderline	Individual aged equal to or less than 20 years at diagnosis And at least one of the following: • At least two morphological features of RHD of the MV without pathological MR or MS • Pathological MR • Pathological AR
ARF only/No RHD	ARF with no evidence of RHD

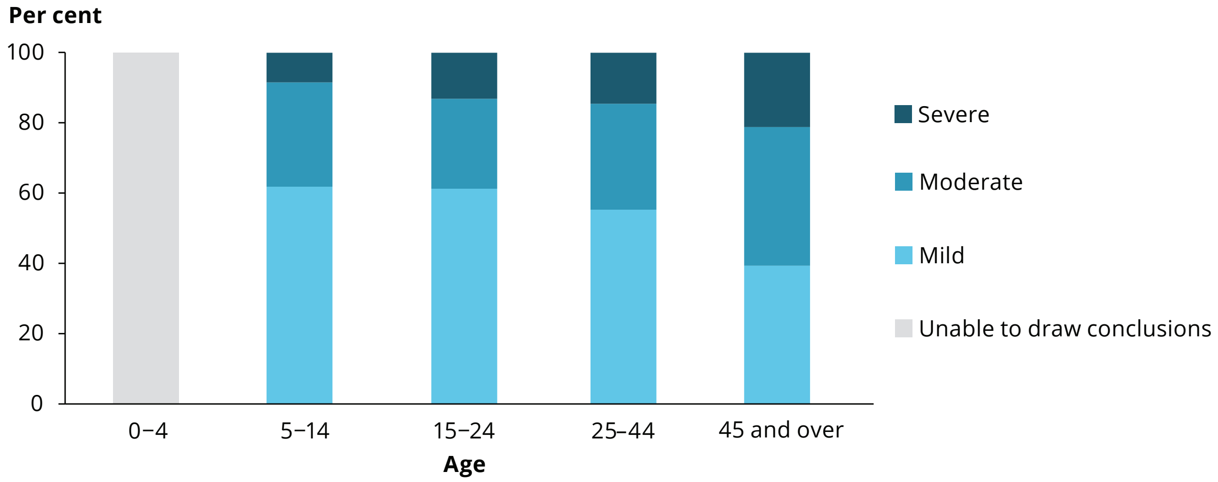
By state or territory:

- Western Australia, South Australia and the Northern Territory had similar distributions of cases, with the majority of cases being mild at diagnosis
- severe cases were the least common in all 3 jurisdictions, but the Northern Territory had the lowest proportion: 10% at diagnosis (Supplementary RHD Table 7).

By age group:

- there was a relatively large proportion of severe cases in the 45 and over age group (24%, 28 diagnoses)
- the proportion of mild cases was highest in young people, with 62% (102 diagnoses) of 5–14 year olds having mild RHD at diagnosis
- there were too few cases among those aged 0–4 to draw conclusions regarding severity at diagnosis (Figure 4.6) (Supplementary RHD Table 7).

Figure 4.6: New RHD diagnoses, Indigenous Australians, by age group and severity status at diagnosis, 2016–2020



Notes

1. RHD severity assessment is at the time of diagnosis. These data exclude 54 cases with no assessment recorded.
2. Data includes WA, SA and NT. Queensland was unable to provide severity data for this report.
3. Age group 0–4 had too few cases to draw conclusions about severity at diagnosis

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Indigenous Australians with no documented previous ARF episode

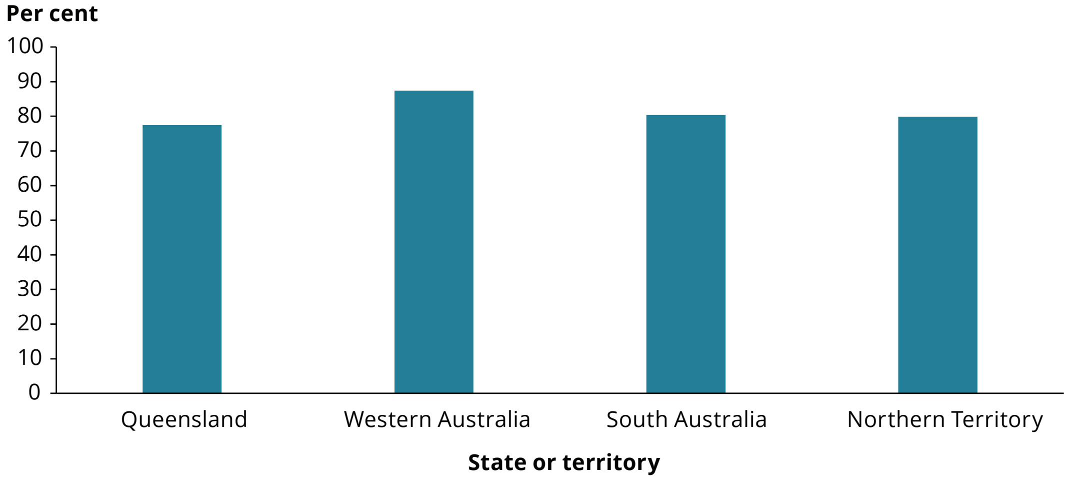
RHD occurs only in someone who has had ARF, but some people with RHD have no recorded previous ARF episode on state and territory registers. ARF might not be notified to a register for various reasons, such as being diagnosed before the relevant register began; being diagnosed prior to the condition being notifiable; the person being diagnosed in a jurisdiction that does not have a register, or the episode was never diagnosed.

Among all Indigenous Australians with a new RHD diagnosis in 2016–2020, four-fifths (80%, 1,125 diagnoses) did not have a previous ARF episode recorded on the registers or were diagnosed with RHD within 90 days of their recorded first ARF episode. Of these:

- the proportion was similar in females (81%) and males (79%)
- the proportion varied across the jurisdictions, being highest in Western Australia (87%) and lowest in Queensland (78%) (Figure 4.7)
- the proportion was lowest in those 15–24 and then increased with age.

These data show that in many cases, RHD could not have been prevented by secondary prophylaxis, and highlight the importance of primordial and primary prevention (Supplementary RHD Table 8).

Figure 4.7: New RHD diagnoses, Indigenous Australians with no previous ARF recorded, by state or territory, 2016–2020



Note: A person is considered to have a previous ARF diagnosis if they were diagnosed with ARF more than 90 days before their RHD diagnosis.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

New RHD diagnoses in New South Wales

All Australians

In 2016–2020:

- there were 58 reports of new RHD diagnoses in New South Wales (0.3 per 100,000 population)
- there were more diagnoses of RHD in non-Indigenous Australians (43 diagnoses, 74%) than Indigenous Australians (15 diagnoses, 26%).
- Pacific Islanders made up the largest proportion of cases in a known risk group (29%, 17 diagnoses)
- cases were evenly distributed by sex with 29 diagnoses each
- the 25–34 age group had the most diagnoses (23 diagnoses, 40%). RHD is not notifiable in NSW in people aged 35 or over at the time of diagnosis
- RHD diagnosis rates have fluctuated between 0.1 and 0.5 per 100,000 cases, likely due to low numbers (Supplementary RHD tables 4b and 5b).

Indigenous Australians

Of the 58 new RHD diagnoses among all Australians in New South Wales in 2016–2020:

- 26% (15) were Indigenous Australians (1.6 per 100,000 population)
- the overall diagnosis rate for Indigenous Australians was 5 times the rate for all Australians (1.6 compared with 0.3 per 100,000)
- the annual rate for Indigenous Australians fluctuated between 1.1 and 3.3, due to the small number of diagnoses. There were too few cases among Indigenous Australians to draw meaningful conclusions about change over time (Supplementary RHD tables 4b and 5b).
- Five Indigenous Australians recorded a severe status. There were too few cases to draw meaningful conclusions.
- Nine-tenths (87%, 13 diagnoses) of Indigenous Australians did not have a previous ARF episode recorded on the registers. There were too few cases to identify patterns with sex or age (Supplementary RHD Table 8b).

Surgery among Indigenous Australians with RHD

For analysis purposes, a surgical event was included regardless of the year of RHD diagnosis, acknowledging that the years for which jurisdictions have been collecting data vary.

Refer to Table 1.1 for more information.

RHD leads to structural damage to the heart valves – most commonly the mitral valve. The aortic, pulmonary and tricuspid valves can also be affected. Surgery may be needed to replace or repair diseased valves. Common surgeries include: balloon valvotomy, used to reopen narrowed valves; valve repair, which reconstructs and reshapes heart valves to allow for normal blood flow; and valve replacement, where the damaged valve is replaced with a mechanical or bioprosthetic valve. Surgery may include prolonged hospitalisation, isolation from family, and ongoing regular monitoring after replacements. An individual may have surgical events more than once on damaged valves, and may have multiple procedures in one surgical event – that is, multiple valves repaired or replaced in a single surgery.

These figures reflect only those surgeries that were recorded in the registers, and may not include all RHD-related surgery undertaken. However, comparison with data from the National Hospital Morbidity Database suggests that most RHD surgeries among Indigenous Australians in Queensland, Western Australia, South Australia, and the Northern Territory are recorded on the registers (AIHW unpublished analysis). NSW was not included in this previous analysis.

Surgery in Queensland, Western Australia, South Australia and the Northern Territory

In 2016–2020, 566 people diagnosed with RHD and living in Queensland, Western Australia, South Australia, or the Northern Territory had surgery. Of those, almost 2 in 3 (63%; 359 people) were Indigenous Australians who underwent 380 surgical events (Supplementary RHD Table 9a). Of these Indigenous Australians:

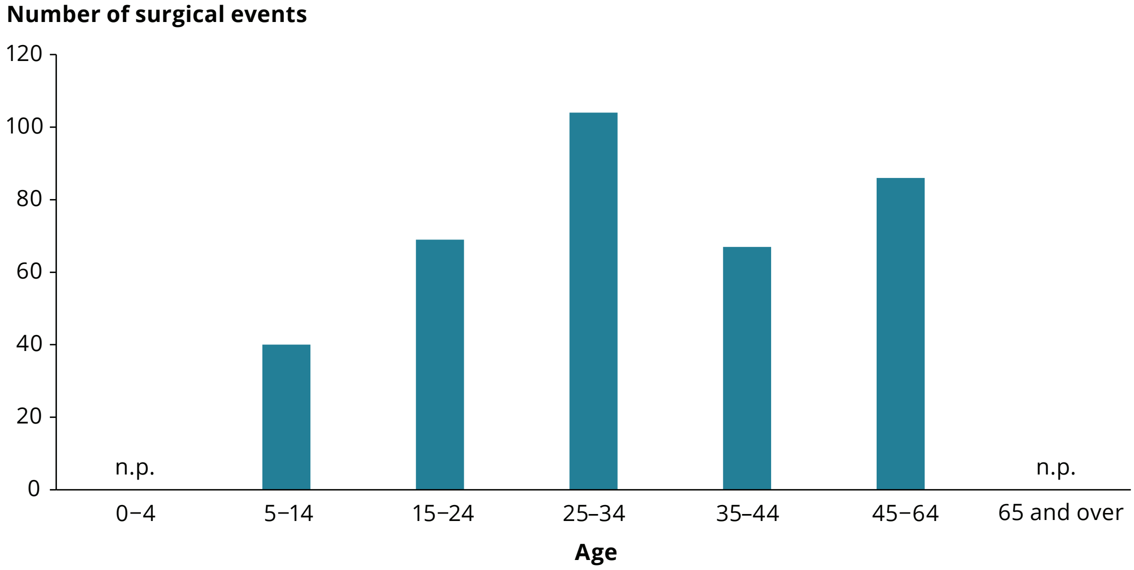
- 339 had surgery once and 20 had at least 2 surgeries (Supplementary RHD Table 10)
- 173 (48%) were from the Northern Territory
- 95 (27%) were from Queensland
- 242 (67%) were female (Supplementary RHD Table 11).

On average, people had their first surgery 6.8 years after RHD diagnosis (Supplementary RHD Table 11).

In 2016–2020, of the 380 surgical events among 359 Indigenous Australians:

- most surgical events occurred among those aged 25–34, who had 104 surgical events (27%), followed by those aged 45–64, with 86 surgical events (23%)
- children aged 5–14 with RHD had 40 surgical events.
- Fewer than 5 children aged under 5 years had surgery for RHD (Figure 4.8) (Supplementary RHD Table 11).

Figure 4.8: Surgical events among Indigenous Australians with RHD, by age group, 2016–2020



Notes

1. There were fewer than 5 surgical events for RHD among those aged 0–4 during 2016–2020.
2. This shows the age at surgery of the 380 RHD-related surgical events for Indigenous Australians during 2016–2020. Individuals who had more than 1 surgery during this period are included multiple times.
3. Data includes Indigenous Australians from Qld, WA, SA and NT combined.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Surgery in New South Wales

In New South Wales in 2016–2020, 12 people underwent 16 RHD surgical events and fewer than 5 of these were among Indigenous Australians.

As such, there are too few cases to draw any meaningful statistical conclusion (Supplementary RHD Table 9b).

All-cause deaths among all Australians with RHD

This analysis was not restricted by cause of death – people with RHD may have died of any cause. Detailed cause of death information is not captured on most registers.

All-cause deaths in Queensland, Western Australia, South Australia and the Northern Territory

All Australians

In 2016–2020 in Queensland, Western Australia, South Australia and the Northern Territory, 476 deaths were reported among people registered as having RHD (Table 4.3). Of these deaths:

- 189 deaths occurred in Northern Territory (40%) (Supplementary RHD Table 12)
- 306 deaths occurred among females (64%)
- 192 deaths occurred among people aged 65 and over (40%)
- the median age of death for males was 60 and for females, 61 (Supplementary RHD Table 13).

Indigenous Australians

In 2016–2020, 322 deaths (68% of all RHD deaths) were reported among Indigenous Australians registered as having RHD. Of these deaths:

- the highest number occurred in the Northern Territory (177 deaths, 55%) (Supplementary RHD Table 12)
- none occurred in Indigenous Australians aged under 15 (Supplementary RHD Table 13).

Table 4.3: Deaths among Indigenous Australians recorded on RHD jurisdictional registers, 2016–2020

Age group	Number	Per cent
0–24	20	6.2
25–44	92	28.6
45–64	134	41.6
65+	76	23.6
All ages	**322**	**100.0**

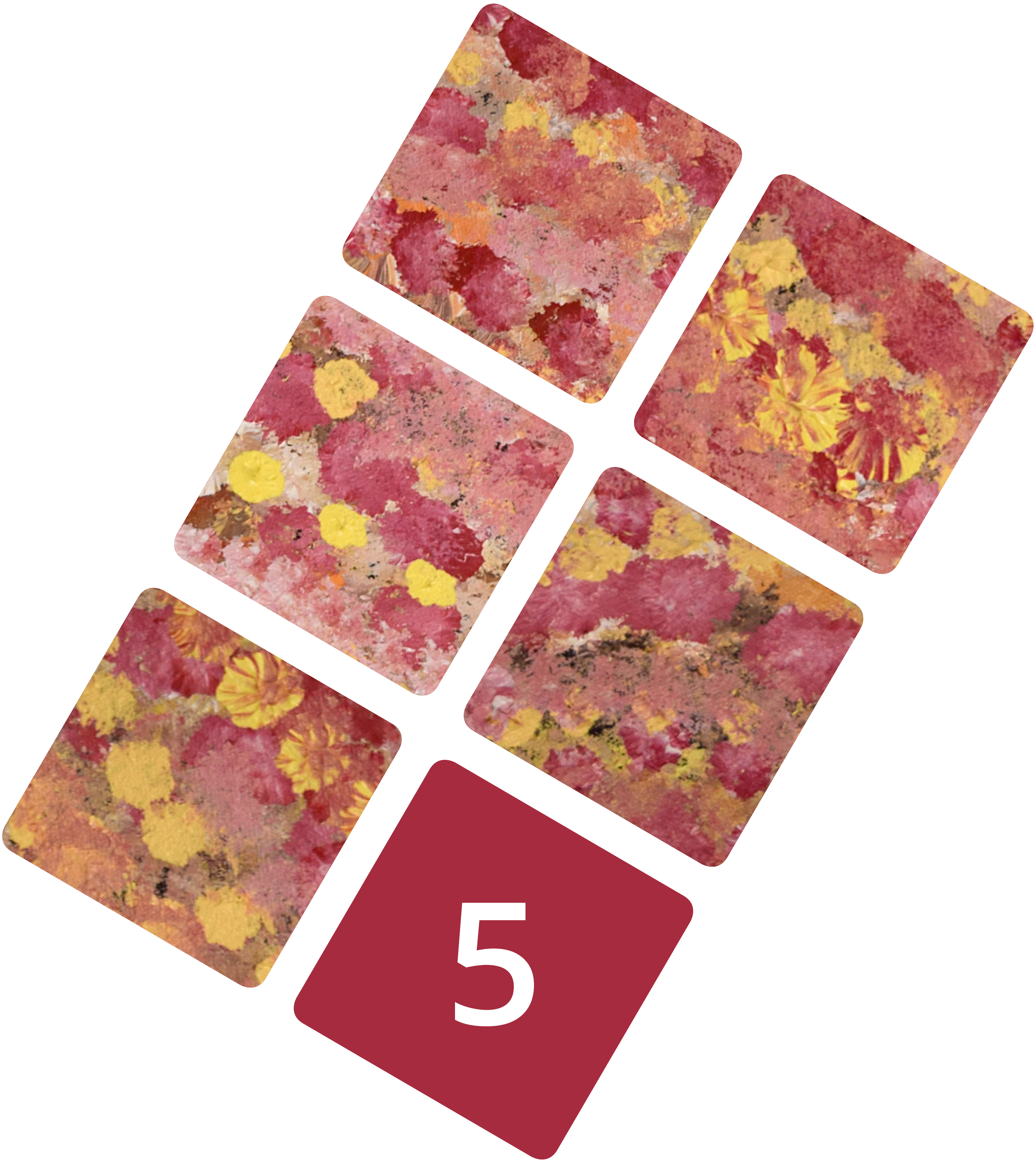
Source: AIHW analysis of the National Rheumatic Heart Disease data collection.

The median age of death was 52 for Indigenous males and 53 for Indigenous females. In comparison, the median age at death for non-Indigenous Australians with RHD was 73 for both males and females. Indigenous Australians with RHD who died during 2016–2020 had lived with their diagnosis for a median of 11 years (Supplementary RHD tables 12 and 13).

All-cause deaths in NSW

There were fewer than 5 deaths among people with RHD in New South Wales, so further analyses cannot be conducted. The small number of deaths is likely due to RHD not being notifiable for those 35 and over at the time of diagnosis and the register being relatively new. Few people aged 35 or over are included on the register.

5



5 Secondary prophylaxis

Secondary prophylaxis refers to the antibiotics given to people who have been diagnosed with ARF and/or RHD to prevent further Strep A infections, thereby reducing the risks of developing ARF again and of developing or worsening RHD. Secondary prophylaxis with regular benzathine benzylpenicillin G (BPG) is the only RHD control strategy shown to be both clinically and cost effective at community and individual levels (Webb et al. 2015; Wyber & Carapetis 2015; RHD Australia 2020), and needs to be complemented with other primordial and primary prevention activities to eliminate RHD.

BPG is routinely recommended every 28 days to maintain prolonged, low-level benzylpenicillin concentrations. A 21-day antibiotic regimen may be considered by a medical specialist for a small proportion of patients who have breakthrough ARF despite receiving the 28-day regimen, or are at high risk of adverse consequences if ARF occurs (RHD Australia 2020).

In New South Wales, details of patients prescribed or administered prophylaxis are recorded on the register only if they have consented to be included. At 31 December 2020, 48% of Indigenous Australian patients had consented to have their prophylaxis data recorded on the register. Prophylaxis data in New South Wales are available only from 2017 onwards. This means that they are not comparable to data from the other 4 jurisdictions, and are reported separately below. Due to the COVID-19 pandemic, NSW was not able to consistently follow-up with all providers of secondary prophylaxis during 2020.

Delivery to Indigenous Australians

Proportion of doses delivered is calculated as a proportion of the scheduled 13 doses per year for patients on a 28-day BPG regime, and 17 doses for patients on a 21-day regime. Patients who commenced part-way through the year have been included with an adjusted expected number of doses. Patients who should have been on BPG but did not receive a dose in 2020 were also included in the analysis.

BPG delivery in Queensland, Western Australia, South Australia and the Northern Territory

There were 4,521 Aboriginal and Torres Strait Islander people eligible for inclusion in calculations about BPG delivery in 2020. They were located in the Northern Territory (2,282), Queensland (1,371), Western Australia (720), and South Australia (148). Of these:

- 13% (607 people) received 100% or more of their prescribed doses
- 22% (985) received 80% to 99% of their prescribed doses
- 29% (1,324) received 50% to 79% of their prescribed doses
- 36% (1,605) received less than 50% of their prescribed doses, including 418 people who did not receive any doses (Table 5.1) (Supplementary Secondary Prophylaxis Table 1).

In general, a lower proportion of doses of prophylaxis was received in 2020 than in previous years. This may be due to the impact of COVID-19 on the health sector and use of health services.

Table 5.1: BPG delivery level in 2020

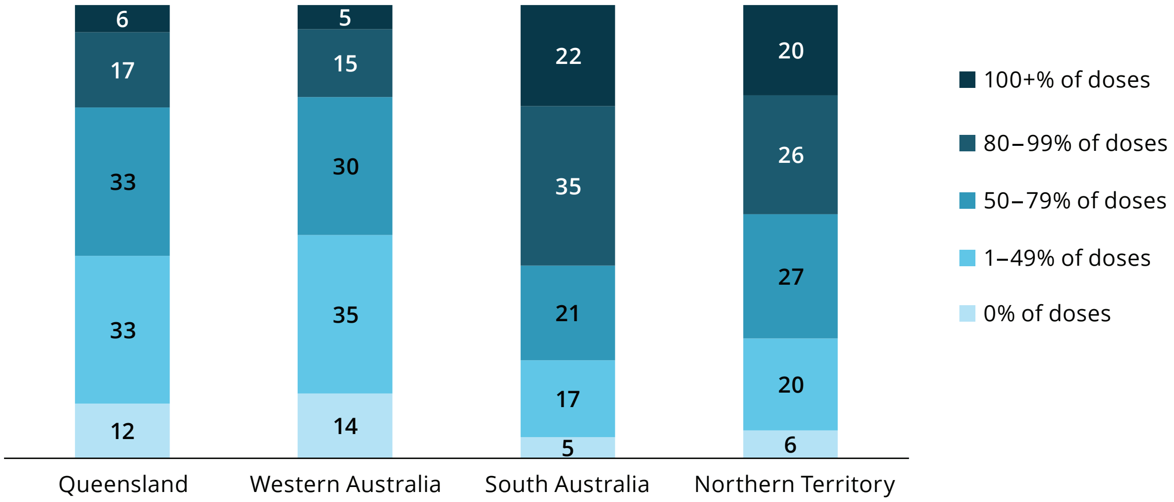
	Number	Per cent
Qld, WA, SA, NT		
0% of doses	418	9.2
1–49% of doses	1,187	26
50–79% of doses	1,324	29
80–99% of doses	985	22
100%+ of doses	607	13
NSW		
0% of doses	21	70
1–100%+ of doses	9	30

Source: AIHW analysis of the National Rheumatic Heart Disease data collection.

In 2020, 35% of Indigenous Australians (1,592 people) received at least 80% of doses. The proportion of people who received at least 80% of prescribed doses in each jurisdiction was:

- 57% in South Australia (85 people)
- 23% in Queensland (310)
- 20% in Western Australia (146)
- 46% in the Northern Territory (1051) (Figure 5.1; Supplementary Secondary Prophylaxis Table 1).

Figure 5.1: Proportion of doses received by Indigenous Australians with ARF and/or RHD on a BPG regime, by state and territory, 2020



Notes

1. People on 21-day BPG can have more than 17 doses in a year or people on 28-day BPG can have more than 13 doses in one year, therefore 100% of doses is defined as 100%+ of doses.
2. This analysis includes people who were prescribed prophylaxis for the whole of 2020, as well as those on BPG for part of the year only.
3. Data include Qld, WA, SA and NT. Does not include cases managed in NSW.

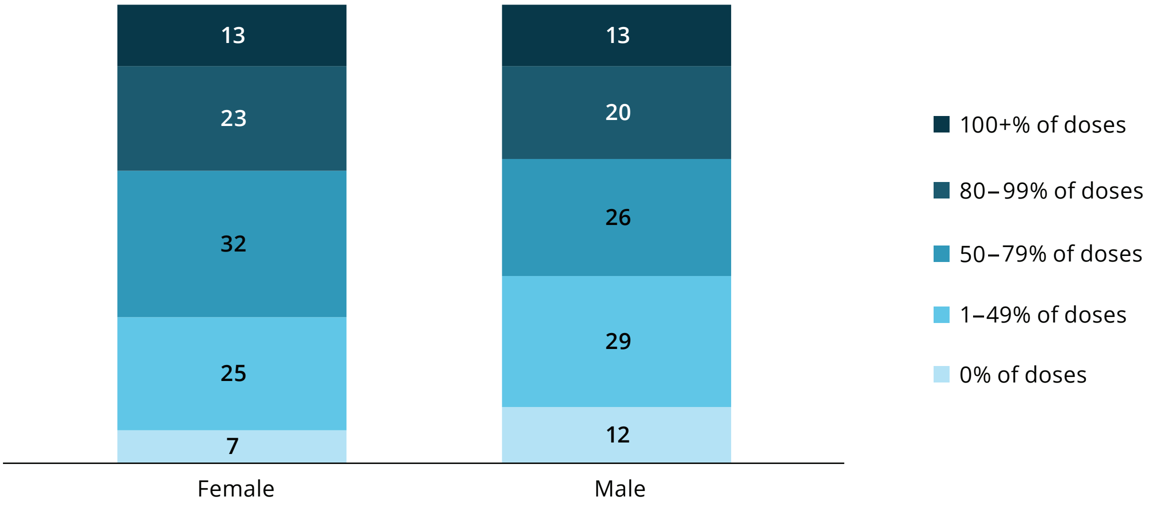
Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Age and sex

In 2020, among Indigenous Australians prescribed prophylaxis:

- more males received fewer than 40% of their prescribed doses (34%) than females (27%) and more females received at least 80% of doses than males (36% and 34%, respectively) (Figure 5.2) (Supplementary Secondary Prophylaxis Table 2).
- delivery was highest among people aged 5–14, with 50% receiving at least 80% of doses (Figure 5.3) (Supplementary Secondary Prophylaxis Table 3).

Figure 5.2: Proportion of doses received by Indigenous Australians with ARF and/or RHD on a BPG regime, by sex, 2020

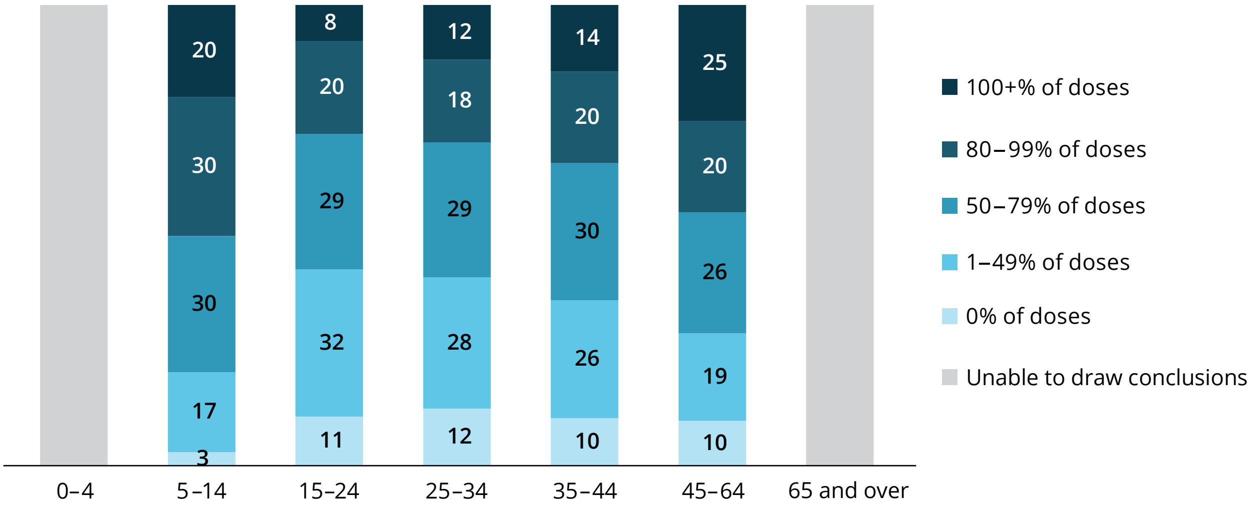


Notes

1. People on 21-day BPG can have more than 17 doses in a year or people on 28-day BPG can have more than 13 doses in one year, therefore 100% of doses is defined as 100%+ of doses.
2. This analysis includes people who were prescribed prophylaxis for the whole of 2020, as well as those on BPG for part of the year only.
3. Data include Qld, WA, SA and NT. Does not include cases managed in NSW.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Figure 5.3: Proportion of doses received by Indigenous Australians with ARF and/or RHD on a BPG regime, by age group, 2020



Notes

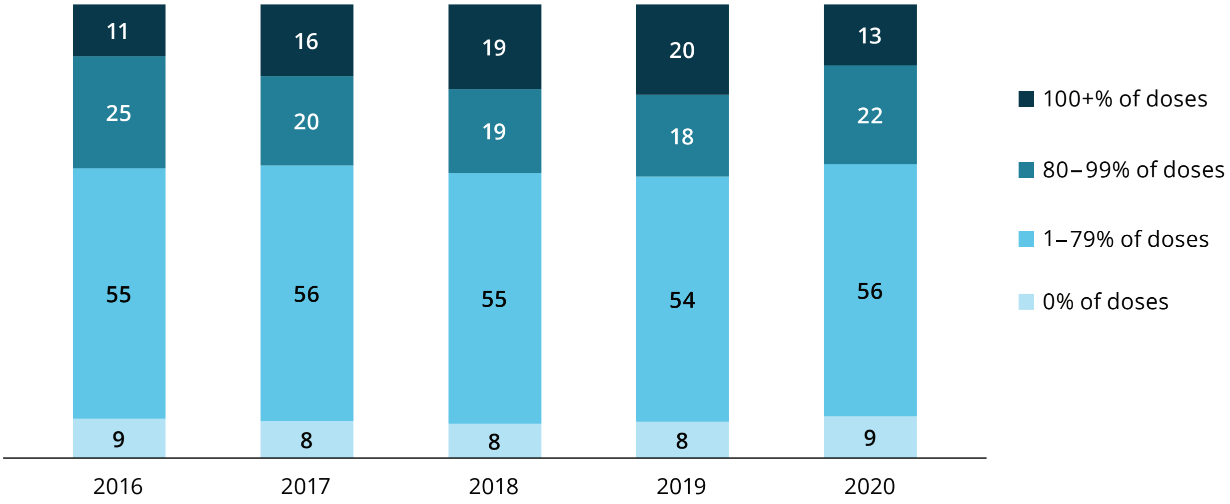
1. People on 21-day BPG can have more than 17 doses in a year or people on 28-day BPG can have more than 13 doses in one year, therefore 100% of doses is defined as 100%+ of doses.
2. This analysis includes people who were prescribed prophylaxis for the whole of 2020, as well as those on BPG for part of the year only.
3. Data include Qld, WA, SA and NT. Does not include cases managed in NSW.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Time trend

The proportion of Indigenous Australians receiving at least 80% of their prescribed doses increased from 36% in 2016 to 38% in 2019 (Figure 5.4). However, there was a decrease in 2020 when only 35% of Indigenous Australians prescribed BPG in 2020 received at least 80% of their doses. In 2020, 9.2% did not receive any of their prescribed doses (Supplementary Secondary Prophylaxis Table 4). The decrease in those receiving at least 80% of their prescribed doses could be due to the impact of COVID-19 on individuals and health services as a result of the lockdowns.

Figure 5.4: Proportion of doses received by Indigenous Australians with ARF and/or RHD on a BPG regime, by year, 2016–2020



Notes

1. People on a 21-day BPG can have more than 17 doses in a year or people on a 28-day BPG can have more than 13 doses in one year, therefore 100% of doses is defined as 100%+ of doses.
2. This analysis includes people who were prescribed prophylaxis for the whole of the relevant year, as well as those on BPG for part of the year only.
3. Data include Qld, WA, SA and NT. Does not include cases managed in NSW.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

BPG delivery in New South Wales

Thirty Indigenous Australians in New South Wales were prescribed secondary prophylaxis during 2020. Of these, 30% had one or more doses and 70% had zero doses recorded (Supplementary Secondary Prophylaxis Table 5b).

ARF recurrence rates among Indigenous Australians who were prescribed BPG

Adherence to secondary prophylaxis reduces the likelihood of recurrence. Trends in the number of recurrent ARF episodes among people prescribed secondary prophylaxis may be used to monitor the effectiveness of ARF and RHD program implementation. Recurrence rates are calculated using the rate per 100 patient-years. Further information on patient-years can be found in Appendix B.

ARF recurrence rates in Queensland, Western Australia, South Australia and the Northern Territory

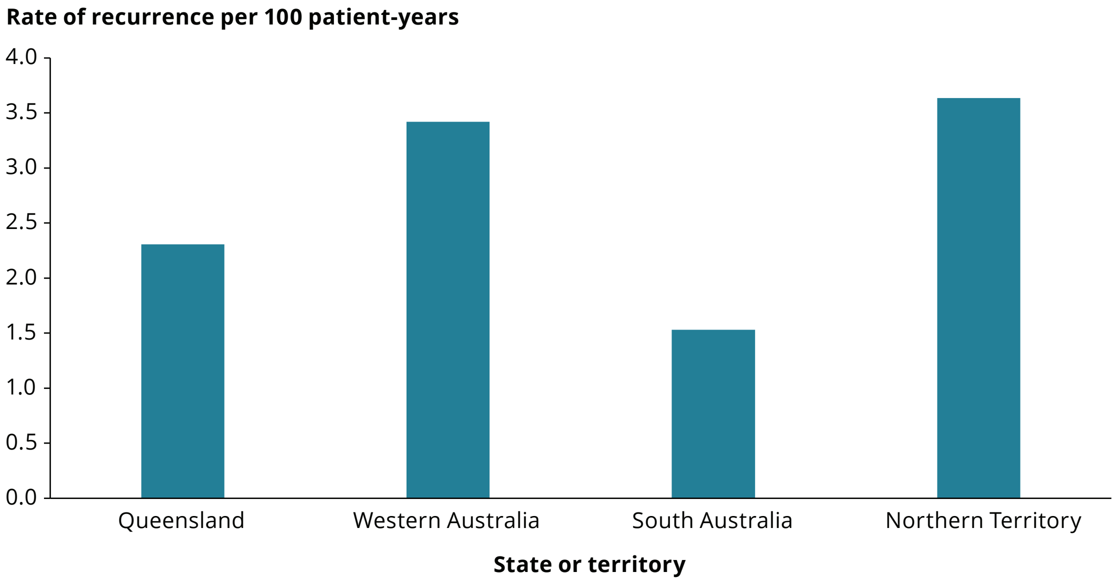
At 31 December 2020:

- among 4,504 Indigenous Australians who had been prescribed BPG, 131 ARF recurrences were reported (3.1 recurrences per 100 patient-years) (Table 5.2)
- most recurrences were in the Northern Territory (76 recurrences)
- the rate of ARF recurrences per 100 patient-years was highest in the Northern Territory (3.6) where 46% of people received at least 80% of prescribed BPG doses; the recurrence rate was lowest in South Australia (1.5) where 57% of people received at least 80% of prescribed BPG doses (Figure 5.5)
- the rate of recurrence per 100 patient-years generally decreased with age, with the highest risk among those aged 5–14 (4.5). There were no recurrences in those aged under 5 and too few in those aged 65 and over to draw conclusions. There was an increase in recurrence rate for those aged 45–64 (2.9), likely due to small numbers (Figure 5.6) (Supplementary Secondary Prophylaxis Table 6).

Table 5.2: ARF recurrences in 2020

	Number	Rate per 100 patient-years
Qld, WA, SA, NT	131	3.1
NSW	n.p.	9.1

Figure 5.5: ARF recurrences per 100 patient-years, Indigenous Australians, by state and territory, 2020

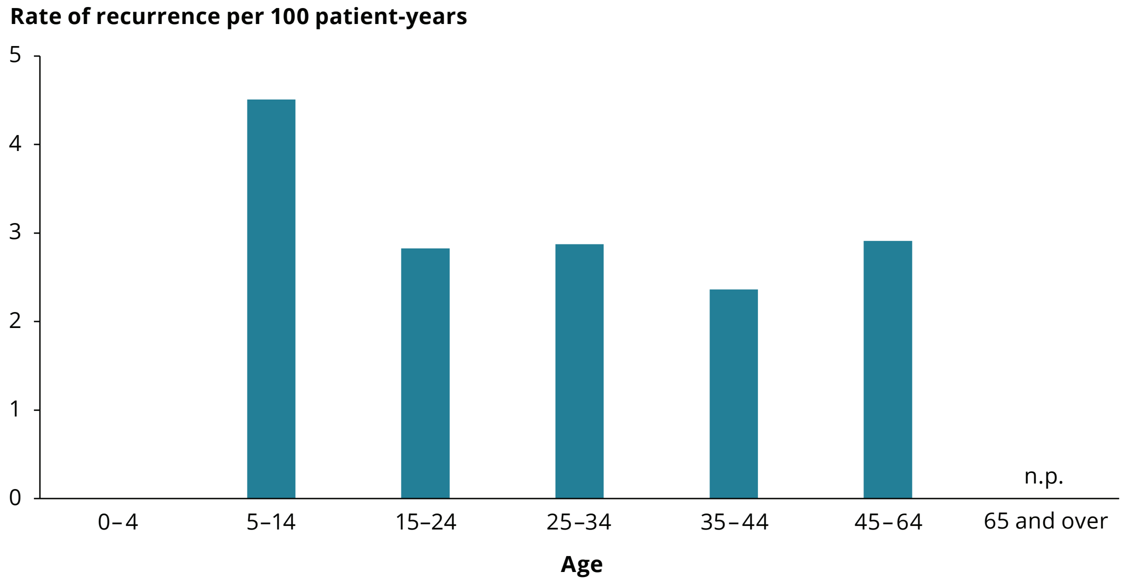


Notes

1. Includes people who were prescribed BPG before or during 2020.
2. Data from Qld, WA, SA and NT.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Figure 5.6: ARF recurrences per 100 patient-years, Indigenous Australians, by age group, 2020



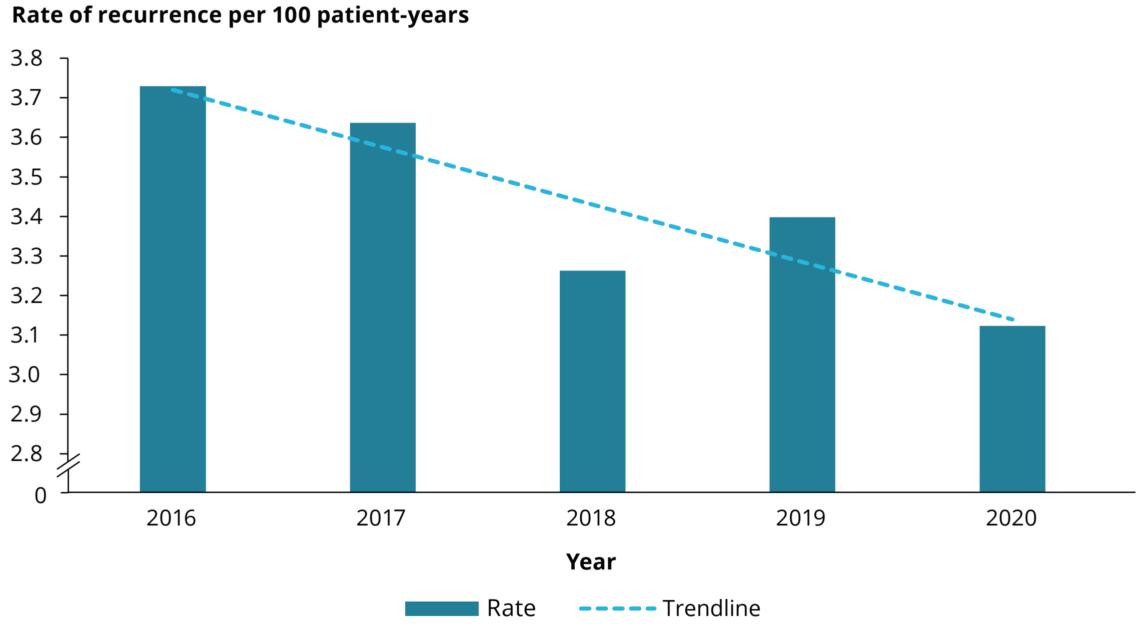
Notes

1. Includes people who were prescribed BPG before or during 2020.
2. Data from Qld, WA, SA and NT.
3. There were no cases in those under 5 and too few 65 and older to calculate meaningful recurrence rates.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

From 2016 to 2020 for Queensland, Western Australia, South Australia, and the Northern Territory, the ARF recurrence rate per 100 patient-years among Indigenous Australians prescribed BPG decreased from 3.7 to 3.1 (Figure 5.7) (Supplementary Secondary Prophylaxis Table 7).

Figure 5.7: Rate of recurrence, Indigenous Australians with ARF and/or RHD on BPG regime, by year, 2016–2020



Notes

1. This analysis includes people who were prescribed prophylaxis for the whole of the relevant year, as well as those on BPG for part of the year only.
2. Data includes Qld, WA, SA and NT. Does not include cases managed in NSW.

Source: AIHW analysis of National Rheumatic Heart Disease data collection.

ARF recurrence rates in New South Wales

Secondary prophylaxis data are provided in New South Wales only if an individual consents to be included. These data are available only from 2017 to 2020.

In 2020, among 30 Indigenous Australians who were prescribed BPG, there were less than 5 reported ARF recurrences (9.1 recurrences per 100 patient-years) (Supplementary Secondary Prophylaxis Table 6). The number of recurrences each year is too small to draw meaningful conclusions about change over time.

Appendix A: Data Quality Statement

The National Rheumatic Heart Disease data collection, held by the AIHW, contains data on diagnoses of ARF and RHD in Australia. It is a collation of data from ARF/RHD clinical registers held by certain states and territories in which ARF and/or RHD are notifiable diseases. In 2020, ARF and RHD were notifiable in 5 jurisdictions (New South Wales, Queensland, Western Australia, South Australia, and the Northern Territory), although these became notifiable at different times in different jurisdictions. In New South Wales, RHD cases are notifiable only in people aged under 35 years. Diagnoses of notifiable diseases are required by law to be reported to state and territory health authorities, to enable ongoing monitoring and support public health responses.

This is the fourth annual report from the National RHD data collection. It presents information on ARF and RHD in Australia drawn from the established jurisdictional registers. Data in the collection are updated over time as the jurisdictional programs undertake data cleaning and quality improvement activity, so numbers in this report may not match those in previous reports. In addition, rates presented in this report have been calculated using the revised Aboriginal and Torres Strait Islander population estimates based on the 2016 Census (ABS 2019), and should not be compared with those in previously published reports.

In Western Australia, South Australia and the Northern Territory, the ARF/RHD control programs are funded by the Australian Government Department of Health. In Queensland, the ARF/RHD control programs are funded by both the Australian Government Department of Health and the Queensland Government. A state-funded ARF/RHD register commenced in New South Wales in 2016, with notifications starting in late 2015. Data about ARF and RHD diagnoses are not currently collected by jurisdictional health departments in the Australian Capital Territory, Victoria or Tasmania.

The current Northern Territory RHD register has been collecting data since 1997. The South Australian RHD register commenced in 2012, and the Queensland register commenced in 2009, as did the West Australian register. The Queensland register incorporates information from 1999 onwards, from the prior collection in the Ferret database, and transitioned to the current register in 2012. The Northern Territory register incorporates information from a prior collection. All states have different notification and data collection practices and therefore the numbers, data quality and completion in the RHD registers are variable. In particular, in South Australia, only RHD cases aged under 50 are recorded on the register, except when they are from a high-risk population group. For some jurisdictions, consent must be sought from a patient before they are included in the register. Generally, notification and register data are maintained in separate systems and are not linked.

The registers include demographic and clinical information about people with ARF and/or RHD. Records are made of the first known ARF episode and recurrent episodes and diagnoses are classified as definite, probable or possible diagnoses. Data are collected about diagnoses' preventive treatment and episode type, level of confirmation, level of severity at diagnosis and when clinical monitoring activities or surgery are performed.

While the registers have comprehensive data, gaps remain in the availability, quality and collection. Some key performance indicators on echocardiograms, ethnicity, detection methods, wait times for surgery and deaths due to surgery could not be reported due to poor data quality or variation in collection across state and territories. Risk factor information about people in the registers, such as adverse events and living conditions, are not currently collected in any register. These data would assist in monitoring ARF and RHD epidemiology and program evaluation.

Appendix B: Calculations of ARF recurrences per 100 patient-years

ARF recurrence rate per 100 patient-years is the number of ARF recurrent events per 100 patient-years during the period that a person is prescribed prophylaxis and, therefore, at risk of ARF recurrence. The time prescribed prophylaxis is used to determine time at risk of ARF recurrence because a person is prescribed prophylaxis if they have been previously diagnosed with ARF and/or RHD and could therefore have an ARF recurrence. The numerator is the number of recurrences. The denominator of the rate is calculated by adding the time prescribed prophylaxis of all patients, where each patient's exposure time is defined as days spent in a pre-determined time period (that is, a year), ended only by events such as death or the end of the prescription period. The rate is then divided by the total number of days per year to get the value for each patient-year and then multiplied by 100.

Appendix C: Technical information

Table C1: The 2020 Australian guideline for the diagnosis of ARF (modified Jones criteria)

	High-risk groups(a)	All other groups
Definite initial episode of ARF	2 major or 1 major and 2 minor manifestations plus evidence of a preceding Strep A infection(b)	2 major or 1 major and 2 minor manifestations plus evidence of a preceding Strep A infection(b)
Definite recurrent episode of ARF in a patient with known past ARF or RHD	2 major or 1 major and 2 minor or 3 minor manifestations plus evidence of a preceding Strep A infection(b) In the 2012 guidelines, 2 major or 1 major and 1 minor or 3 minor manifestations plus evidence of a preceding Strep A infection(b)	2 major or 1 major and 2 minor or 3 minor manifestations plus evidence of a preceding Strep A infection(b) In the 2012 guidelines, 2 major or 1 major and 1 minor or 3 minor manifestations plus evidence of a preceding Strep A infection(b)
Probable ARF (first episode or recurrence)	A clinical presentation that falls short by either 1 major or 1 minor manifestation, or the absence of streptococcal serology results, but one in which ARF is considered the most likely diagnosis. Such diagnoses should be further categorised according to the level of confidence with which the diagnosis is made: • Highly suspected ARF • Uncertain ARF	A clinical presentation that falls short by either 1 major or 1 minor manifestation, or the absence of streptococcal serology results, but one in which ARF is considered the most likely diagnosis. Such diagnoses should be further categorised according to the level of confidence with which the diagnosis is made: • Highly suspected ARF • Uncertain ARF
Major manifestations	Carditis (including subclinical evidence of rheumatic valvulitis on echocardiogram) Polyarthritis(c) or aseptic mono-arthritis or polyarthralgia Chorea(d) Erythema marginatum(e) Subcutaneous nodules	Carditis (including subclinical evidence of rheumatic valvulitis on echocardiogram) In the 2012 guidelines, Carditis (excluding subclinical evidence of rheumatic valvulitis on echocardiogram) Polyarthritis(c) Chorea(d) Erythema marginatum(e) Subcutaneous nodules
Minor manifestations	Monoarthralgia Fever(f) ESR≥30mm/h or CRP ≥30 mg/L Prolonged P-R interval on ECG(g)	Fever(f) Polyarthralgia or aseptic mono-arthritis ESR ≥60 mm/h (ESR ≥30 mm/h in the 2012 guidelines) or CRP ≥30 mg/L Prolonged P-R interval on ECG(g)

CRP = C-reactive protein   ECG = electrocardiogram   ESR = erythrocyte sedimentation rate   Strep A = group A streptococcus

(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 and Torres Strait Islander people 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 Strep A.

(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 Strep A 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 ≥38.5°C (≥38°C in the 2012 guidelines) 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.

Source: RHD Australia 2020.

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The Department of Health funds the AIHW to report on rheumatic heart disease control strategies for Queensland, Western Australia, South Australia and the Northern Territory. The New South Wales Government funds that state's control program.

Abbreviations

ABS	Australian Bureau of Statistics
ACT	Australian Capital Territory
AIHW	Australian Institute of Health and Welfare
ARF	acute rheumatic fever
BPG	benzathine benzylpenicillin G
NSW	New South Wales
NT	Northern Territory
Qld	Queensland
RHD	rheumatic heart disease
SA	South Australia
Strep A	group A streptococcal infection
Tas	Tasmania
Vic	Victoria
WA	Western Australia
WHO	World Health Organization

Symbols

—	nil or rounded to zero
. .	not applicable
n.p.	not publishable because of small numbers, confidentiality or other concerns about the quality of the data
≥	greater than or equal to
≤	less than or equal to

Glossary

Aboriginal and/or Torres Strait Islander: A person of Aboriginal and/or Torres Strait Islander descent who identifies as an Aboriginal and/or Torres Strait Islander. See also **Indigenous**.

acute rheumatic fever (ARF): An acute, serious disease that affects mainly children and young adults, and can damage the heart valves, the heart muscle and its lining, the joints, and the brain. It is brought on by a reaction to a throat or skin infection by group A streptococcal bacteria. Now very rare in the non-Indigenous population, it is still at unacceptably high levels among Indigenous Australians living in remote areas. Also referred to as rheumatic fever.

aortic valve: Valve between the left ventricle and the aorta in the heart.

associated cause(s) of death: A cause(s) listed on the Medical Certificate of Cause of Death, other than the underlying cause of death. They include the immediate cause, any intervening causes, and conditions that contributed to the death but were not related to the disease or condition causing death.

bioprosthetic valve: A prosthetic valve made from human or animal donor tissue. Used in patients with rheumatic heart disease who require surgery.

cause(s) of death: All diseases, morbid conditions or injuries that either resulted in or contributed to death – and the circumstances of the accident or violence that produced any such injuries – that are entered on the Medical Certificate of Cause of Death. Causes of death are commonly reported by the underlying cause of death. See also **associated cause(s) of death** and **underlying cause of death**.

group A streptococcus (Strep A) infection: Caused by bacteria known as group A (beta-haemolytic) streptococcus, a common infection that can cause sore throats (pharyngitis), scarlet fever or impetigo (skin sores).

health hardware: The physical equipment necessary for healthy, hygienic living within homes or communities. The term has been used to describe safe electrical systems, toilets, showers, taps, kitchen cupboards and benches, stoves, ovens and fridges collectively.

mechanical valve: A long-lasting valve made of durable materials. Used in patients with rheumatic heart disease who require surgery. Requires lifelong anticoagulant medication.

mitral valve: Valve between the left atrium and the left ventricle in the heart.

Indigenous: A person of Aboriginal and/or Torres Strait Islander descent who identifies as an Aboriginal and/or Torres Strait Islander. Used interchangeably with Aboriginal and/or Torres Strait Islander in this report. See also **Aboriginal and/or Torres Strait Islander**.

Non-Indigenous Australians: People who have declared that they are not of Aboriginal or Torres Strait Islander descent. Compare with **Indigenous**.

pulmonary valve: Valve between the right ventricle and the pulmonary artery in the heart.

region of management: The regional health boundaries are defined by each jurisdiction. For some jurisdictions, the regions align with other standard geographic classifications such as remoteness categories but for other jurisdictions the regions are state-specific areas.

rheumatic heart disease (RHD): An acquired chronic disease referring to damaged heart valves caused by earlier episode(s) of acute rheumatic fever.

tricuspid valve: Valve between the right atrium and the right ventricle in the heart.

underlying cause of death: The disease or injury that initiated the train of events leading directly to death, or the circumstances of the accident or violence that produced the fatal injury. See also associated cause(s) of death.

valvotomy: An operation that opens up a stenosed (unnaturally narrow) heart valve and allows it to function properly. Used in patients with rheumatic heart disease who require surgery.

Valve repair: Reconstruction, restoration of diseased native valve tissue. An alternative to valve replacement and used in patients with rheumatic heart disease who require surgery.

valve replacement: Replacement of one or more of the heart valves with either an artificial valve or a bioprosthesis. Used in patients with rheumatic heart disease who require surgery. See also bioprosthetic valve.

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Acute rheumatic fever (ARF) and rheumatic heart disease (RHD) are preventable diseases disproportionately affecting Aboriginal and/or Torres Strait Islander Australians living in regional and remote areas. Prevalence rates were highest in females and young people aged 5–14. These data highlight the important role of jurisdictional control programs and registers in New South Wales, Queensland, Western Australia, South Australia, and the Northern Territory.

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