Acute rheumatic fever and rheumatic heart disease in Australia, 2017–2021

Contents

Summary

This is the fifth 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 2017–2021. Throughout this report, some data from New South Wales are incorporated with figures from other jurisdictions and some remain separate, depending on comparability between jurisdictions. In this report, the terms 'Aboriginal and Torres Strait Islander people' and 'Indigenous Australians' are used interchangeably.

What is new to this report:

- All statistical information for previous years has been updated in this report. Changes between years are presented in the trend analysis in each section of the report. 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. Comparisons to the results in previous versions of the report is discouraged.
- COVID-19 impacted both the health sector and the utilisation of health services in 2020 and 2021. This could have affected results such as diagnosis rates, BPG delivery and the number of surgeries undertaken. Further information on how the pandemic affected ARF and RHD specifically is not available.
- Trend figures now show a 95% confidence Interval for each year instead of a trendline. These have been used to determine whether changes over time are statistically significant.

At 31 December 2021, there were 9,922 people living with a diagnosis of ARF and/or RHD recorded on the registers in New South Wales, Queensland, Western Australia, South Australia and the Northern Territory. Of these, 3,053 people (31%) had only ARF recorded, 3,237 people (33%) had only RHD recorded, and 3,632 people (37%) had both ARF and RHD recorded.

Acute rheumatic fever

In 2017–2021:

- 2,784 diagnoses of ARF were recorded in 2,537 people in New South Wales, Queensland, Western Australia, South Australia and the Northern Territory (3.1 per 100,000 population over the 5 years combined) (Supplementary tables 3.1 and 3.2)
- Indigenous Australians accounted for 92% (2,570) of the diagnoses (69 per 100,000 population over the 5 years combined) (Supplementary Table 3.2)
- the number and rate of diagnoses among Indigenous Australians increased from 474 (66 per 100,000) in 2017 to 582 (75 per 100,000) in 2021 (Supplementary Table 3.3)
- ARF was more common among Indigenous females (1,465 cases, 78 per 100,000) than males (1,105 cases, 59 per 100,000) (Supplementary Table 3.4b)

- the highest rate was among Indigenous Australians aged 5–14 (1,142 diagnoses, 140 per 100,000). The median age at diagnosis among Indigenous Australians was 15.4 years (Supplementary Table 3.4b)
- there were 42 deaths reported among all Australians with only an ARF diagnosis on the register (Supplementary Table 3.9a).

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 2021, 6,749 people (67 per 100,000) living with RHD were recorded on the jurisdictional registers in Queensland, Western Australia, South Australia and the Northern Territory (Supplementary Table 4.1). Of these,

- 29% (1,952) were aged under 25 (Supplementary Table 4.2a)
- 66% (4,447) were females (Supplementary Table 4.1)
- the highest prevalence rate was in the Northern Territory (984 per 100,000) and the greatest number was in Queensland (3,104 people) (Supplementary Table 4.1)
- About 3 in 4 diagnoses (5,238, 78%) were among Indigenous Australians (Supplementary Table 4.2a)
- the median age among Indigenous Australians with RHD (33 years) was considerably younger than for non-Indigenous Australians (60 years) (Supplementary Table 4.2a).

At 31 December 2021, there were 120 people living with RHD recorded on the register in New South Wales (Supplementary Table 4.2b).

Incidence

Over the period 2017–2021 in Queensland, Western Australia, South Australia and the Northern Territory:

- 1,750 new RHD diagnoses among Indigenous Australians were reported (75 per 100,000 population) (Supplementary Table 4.4a)
- new RHD diagnoses were more common among Indigenous females than males (97 and 53 diagnoses per 100,000, respectively) (Supplementary Table 4.6)
- 55% of new diagnoses were among Indigenous Australians aged under 25 (970 diagnoses) (Supplementary Table 4.6)
- the greatest number and highest rate of new diagnoses among Indigenous Australians was in the Northern Territory (740, or 193 per 100,000) (Supplementary Table 4.4a)
- the number and rate of RHD diagnoses among Indigenous Australians fluctuated from a low of 303 (68 per 100,000) in 2017 to a high of 393 (86 per 100,000) in 2018. There has been no discernible pattern (Supplementary Table 4.4a).

In 2017–2021, 64 Australians were diagnosed with RHD in New South Wales (Supplementary Table 4.4b).

- Two-thirds of new cases (67%, 43 people) were in non-Indigenous Australians, with Pacific Islanders accounting for 27% (17) of all new RHD diagnoses in New South Wales (Supplementary Table 4.5b).

Heart surgery for RHD

In 2017–2021 in Queensland, Western Australia, South Australia and the Northern Territory, 626 people underwent 666 surgical events for RHD. Most of these were Indigenous Australians, with 59% (368) of patients and 59% (396) of events being among Indigenous Australians (Supplementary Table 4.9a).

In New South Wales, data on heart surgery for RHD are only collected for patients who provide consent. In 2017–2021, 12 people underwent 12 surgical events in New South Wales. Due to small numbers, the breakdown by Indigenous status could not be published (Supplementary Table 4.9b).

Deaths reported among people on the RHD registers

In 2017–2021, 595 deaths were reported among people on the RHD registers in Queensland, Western Australia, South Australia and the Northern Territory. Of these,

- 382 people (64%) were Indigenous Australians
- 389 deaths occurred among females (65%)
- the median age at death was 51 for Indigenous males and 56 for Indigenous females, compared with 73 and 74 for non-Indigenous males and females, respectively (Supplementary Table 4.13).

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

Delivery of secondary prophylaxis to Indigenous Australians

In 2021, 4,816 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 21–28 days (Supplementary Table 5.1).

In New South Wales, information on BPG is only recorded for patients who provide consent.

Proportion receiving BPG as prescribed

In 2021, among Indigenous Australians in Queensland, Western Australia, South Australia and the Northern Territory prescribed 3- or 4-weekly BPG:

- 18% (875 people) received 100% or more of their prescribed doses
- 13% (638) received 80% to 99% of their prescribed doses
- 27% (1,319) received 50% to 79% of their prescribed doses
- 41% (1,984) received less than 50% of their prescribed doses, including 514 people who received no doses (Supplementary Table 5.1).

The proportion of Indigenous Australians receiving at least 80% of their prescribed doses increased from 36% in 2017 to 39% in 2019, but then decreased to 31% in 2021. This drop may relate to the effects of the COVID-19 pandemic on care-seeking behaviour and health service delivery.

In 2021, among Indigenous Australians in New South Wales for whom information on BPG was available:

- 28% (9 people) received at least 80% of their prescribed doses
- 25% (8 people) received no doses (Supplementary Table 5.5b).

ARF recurrence rate among people on BPG

In 2021, among 4,795 Indigenous Australians prescribed BPG in Queensland, Western Australia, South Australia and the Northern Territory, there were 163 ARF recurrences at a rate of 3.7 recurrences per 100 patient-years (Supplementary Table 5.6). Further information on the definition and calculation for patient-years can be found in Appendix B.

In 2021, among 32 Indigenous Australians in New South Wales for whom information on BPG was available, there were no ARF recurrences (Supplementary Table 5.6).

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.

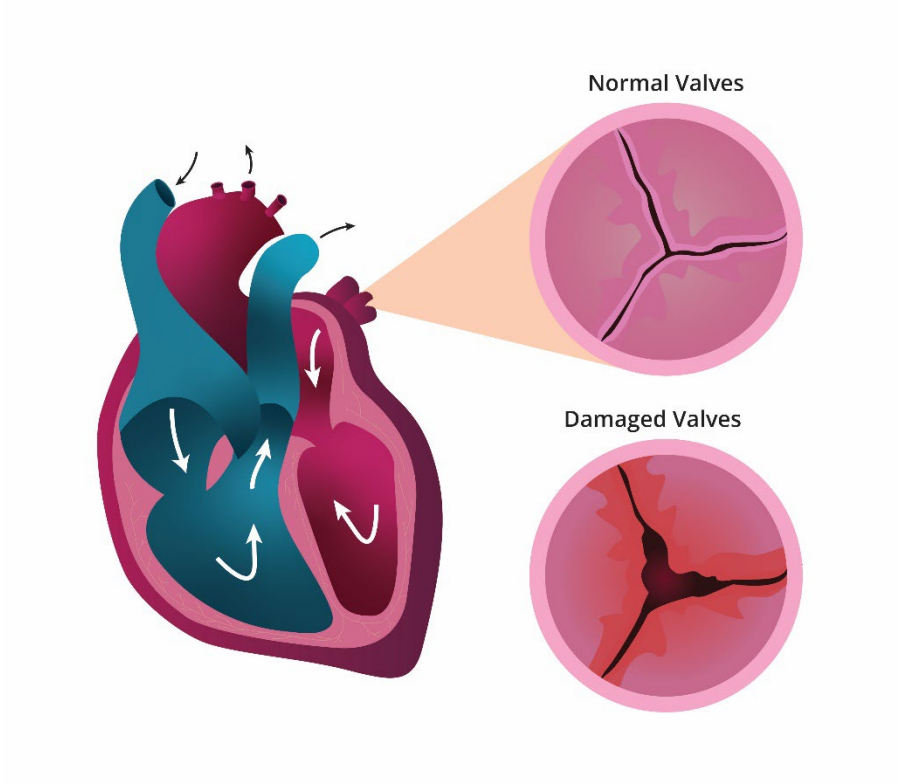
Figure 1.1 shows a diagram of the heart, comparing normal and damaged valves.

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 (RHDAustralia 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.

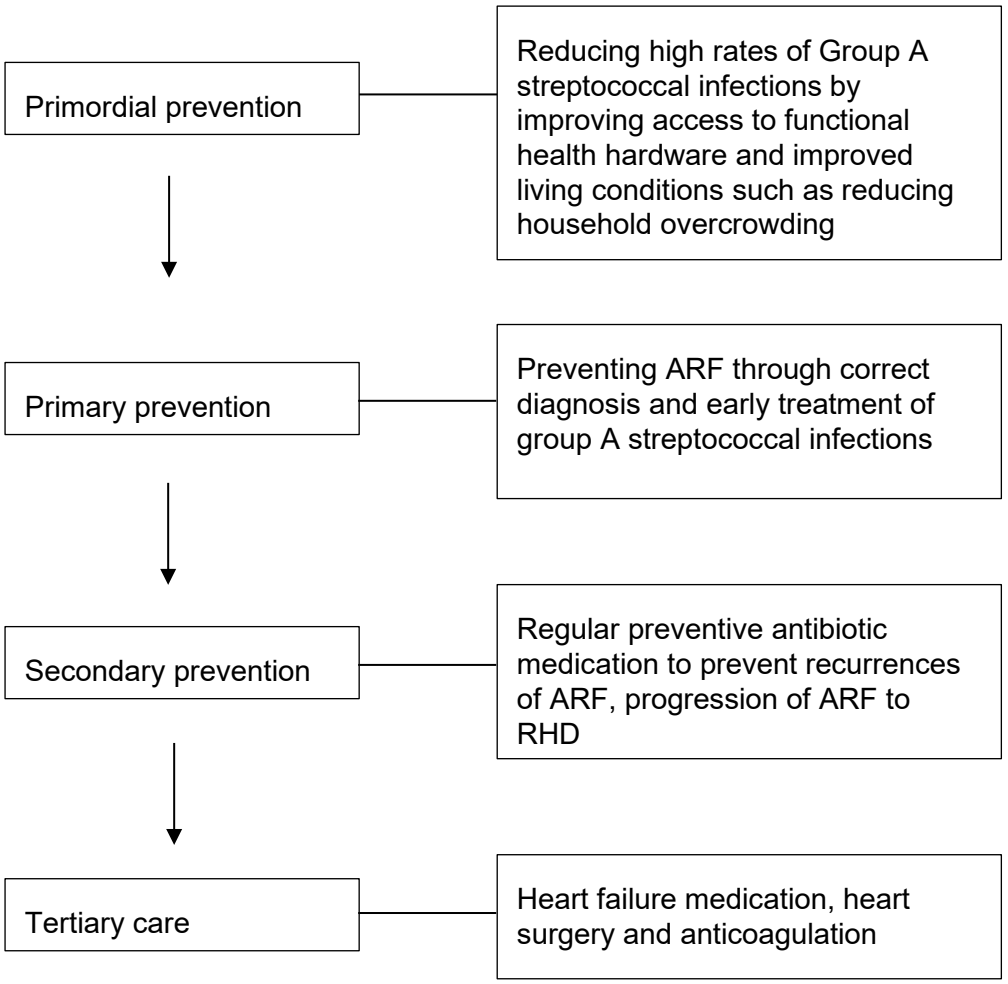
ARF is also preventable through early treatment of Strep A infections with penicillin. This is called primary prevention and relies on correct diagnosis and treatment of skin and throat infections with antibiotics as soon as possible after onset of symptoms. Timeliness of diagnosis and subsequent treatment 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 (RHDAustralia 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 RHDAustralia 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; RHDAustralia 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



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. More detailed information 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	2015	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: RHDAustralia (ARF/RHD writing group) 2020.

About this report

This report presents information on ARF and RHD cases diagnosed or receiving treatment during 2017–2021, 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. Additional web reports will provide more detail at the individual state and territory level (AIHW forthcoming 2023).

Supplementary tables are available at: https://www.aihw.gov.au/reports/indigenous-australians/arf-rhd/summary

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 (Table 1.2). This fifth 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.

Table 1.2: Key reporting indicators

1.1 Yearly ARF incidence by episode type, age group and	1.1.1 Sex 1.1.2 Ethnicity
1.2 Yearly ARF recurrences	1.2.1 Proportion of all ARF episodes 1.2.2 Rate per 100 patient-years for patients prescribed prophylaxis (both oral and BPG)
1.3 Yearly RHD point prevalence by age group and	1.3.1 Sex 1.3.2 Ethnicity 1.3.3 Severity classification
1.4 Proportion of people receiving secondary prophylaxis each year	1.4.1 80-100% 1.4.2 40-79% 1.4.3 0-39% of expected doses

Source: RHDAustralia 2020.

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. Comparison of results between different time periods should use the data presented in this report, and comparisons to the results in previous versions of the report is discouraged.

Some sections in this report present results by whether people are male or female. This may refer to either sex or gender, depending on the data source. Most current data sources do not record sex and gender as separate data items so it can be unclear which is being reported.

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 Notifiable Conditions Information System (NCIMS) and the NSW ARF and 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. RHD severity, priority, cardiac surgery, and secondary prophylaxis for people diagnosed with ARF and/or RHD are only captured on the NSW ARF and RHD Register for 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.

2 Overview of people on the registers

At 31 December 2021, there were 9,922 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:

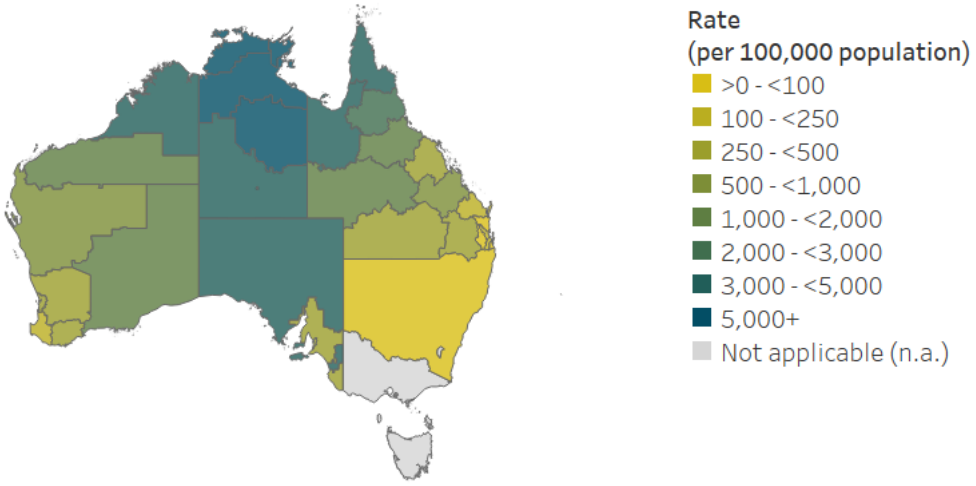
- 3,053 (31%) people had ARF only recorded; 3,237 (33%) had only RHD recorded; and 3,632 (37%) had both ARF and RHD recorded (Supplementary Table 2.1)
- Northern Territory had the highest number of people living with ARF and/or RHD (4,009, 40%), followed by Queensland (3,913, 39%), Western Australia (1,380, 14%), South Australia (419, 4.2%) and New South Wales (200, 2.0%) (Supplementary Table 2.2)
- Indigenous Australians accounted for 8,082 (81%) of the people diagnosed compared with 1,815 (18%) non-Indigenous Australians (Supplementary Table 2.2)
- more females (6,133, 62%) than males (3,789, 38%) were living with a diagnosis of ARF and/or RHD (Supplementary Table 2.3).

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. For more information on regions, see Region of Diagnosis in Chapter 3: Acute rheumatic fever. The regions with the highest rates of management for Indigenous Australians were Rural Darwin, East Arnhem, and Urban Alice Springs:

- Rural Darwin (NT), with 7,612 cases per 100,000 population (946 persons)
- East Arnhem (NT), with 6,607 per 100,000 (754 persons)
- Urban Alice Springs (NT), with 6,226 per 100,000 (430 persons, Figure 2.1) (Supplementary Table 2.4).

  Note that the region of the NT community of Numbulwar was changed from East Arnhem to Katherine (Big Rivers), and this change accounts for some of the reduction in patient numbers compared with previous results.

Figure 2.1: ARF and/or RHD diagnoses among Indigenous Australians, by region of management, at 31 December 2021



Notes

1. Crude age-specific rates per 100,000 calculated using the number of notifications of each calendar year divided by the regional population for 2021 provided by the states or territory or calculated by the AIHW, with the exception of NSW which is from the Australian Bureau of Statistics. Due to constraints in the jurisdictions, previous populations were used when new populations were unable to be supplied. NSW, SA, and NT populations have up-to-date data. Qld populations are from 2020, and WA populations are from 2019.

2. Includes all Indigenous Australians managed for ARF and/or RHD in 2021.

3. The data excludes 82 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. See Supplementary Table 2.4.

3 Acute rheumatic fever

This section discusses diagnoses of ARF reported by Australian RHD control programs between 2017 and 2021. 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

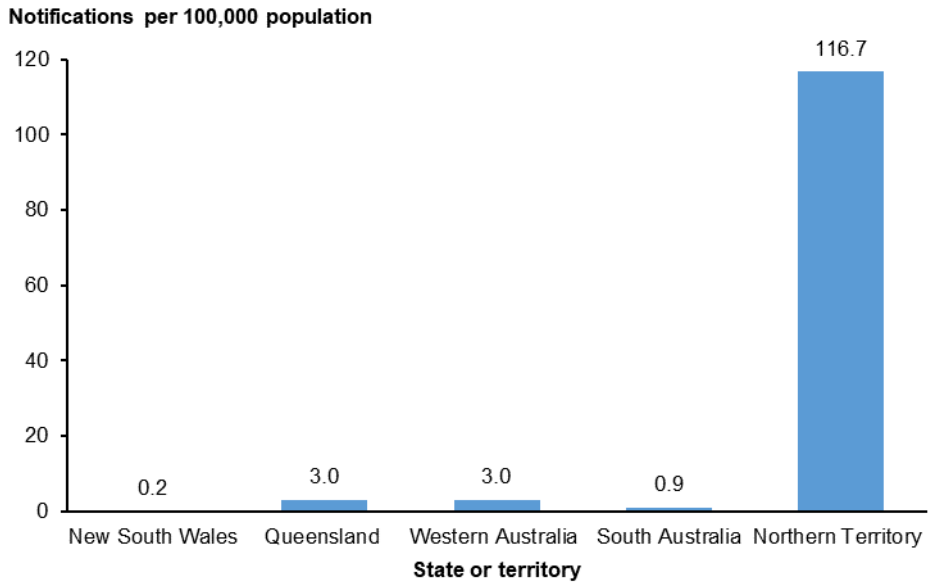
In 2017–2021:

- 2,784 diagnoses of ARF were recorded in New South Wales, Queensland, Western Australia, South Australia and the Northern Territory combined (incidence rate 3.1 per 100,000 population over the 5 years combined). These diagnoses occurred in 2,537 Australians (Supplementary tables 3.1 and 3.2)
- the Northern Territory had the highest rate of ARF diagnoses (117 per 100,000 population; 1,450 diagnoses), followed by Western Australia (3.0 per 100,000; 404 diagnoses), Queensland (3.0 per 100,000; 767 diagnoses), South Australia (0.9 per 100,000 population; 80 diagnoses) and New South Wales (0.2 per 100,000 population; 82 diagnoses) (Figure 3.1) (Supplementary Table 3.2).

Time trend

- The number and rate of ARF diagnoses generally increased from 534 (3.1 per 100,000) in 2017 to 612 (3.4 per 100,000) in 2021. There was a decrease in cases in 2019, to 518 cases (2.9 per 100,000) (Figure 3.2) (Supplementary Table 3.3). However, the confidence intervals suggest that none of these changes were statistically significant.

Figure 3.1: ARF notifications among all Australians, by state and territory, 2017–2021

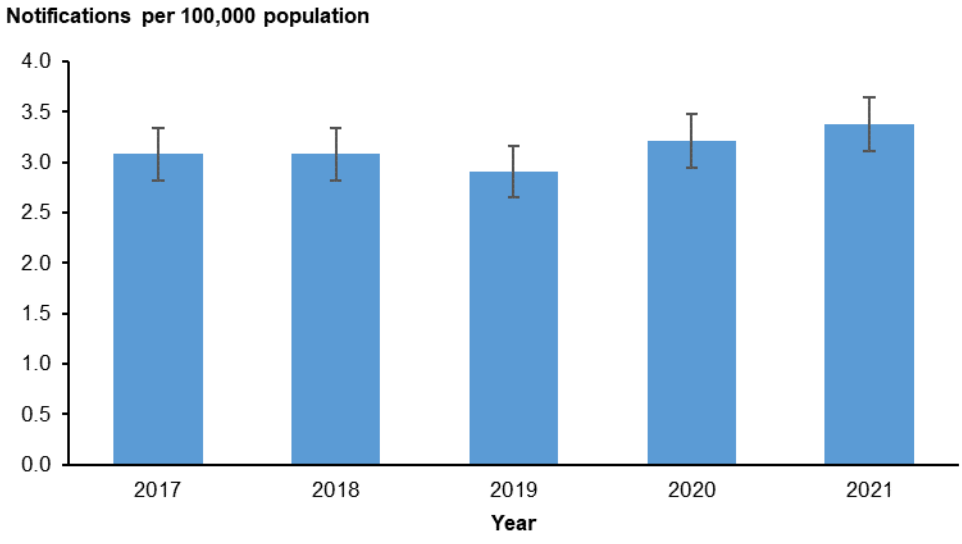


Notes

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.

Figure 3.2: ARF notifications among all Australians, by year, 2017–2021



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.
4. Whiskers show 95% confidence intervals around the rate estimate. This describes the range of values within which we can be 'confident' the true value lies.

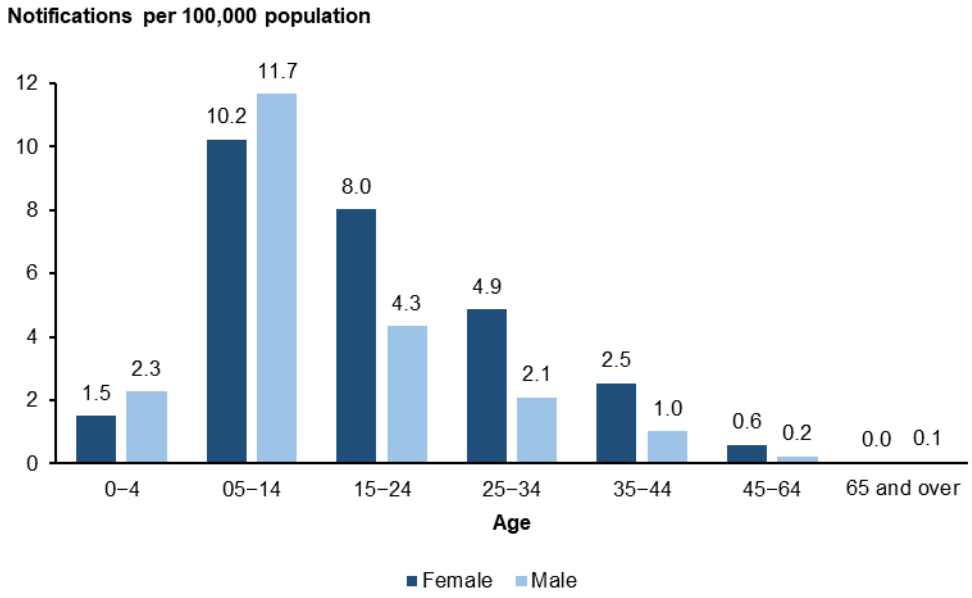
Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Age and sex

Of the 2,784 ARF diagnoses among all Australians in 2017–2021:

- the most common age at diagnosis was 5–14, with 1,238 diagnoses (11 per 100,000 population)
- there were 104 diagnoses (1.9 per 100,000) among children aged 0-4
- females accounted for 1,571 (56%) diagnoses
- in people aged under 15, ARF rates were higher among males than females
- for adults, ARF rates were higher among females than males (Figure 3.3) (Supplementary Table 3.4a).

Figure 3.3 ARF notifications among all Australians, by sex and age group, 2017–2021



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.

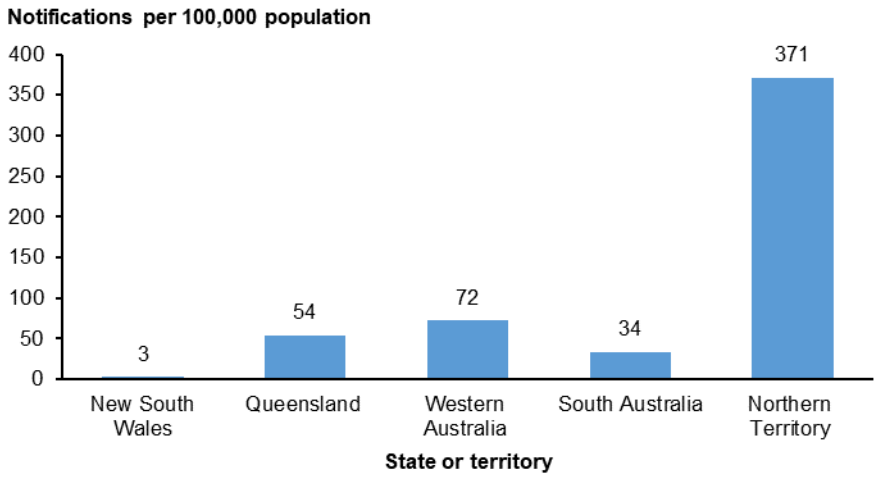
Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Indigenous Australians

In 2017–2021:

- 2,570 diagnoses of ARF were reported in Aboriginal and Torres Strait Islander people in New South Wales, Queensland, Western Australia, South Australia and the Northern Territory, a rate of 69 per 100,000 population
- the Northern Territory had the highest rate of diagnoses (371 per 100,000 population; 1,426 diagnoses), followed by Western Australia (72 per 100,000 population; 385 diagnoses), Queensland (54 per 100,000 population; 644 diagnoses), South Australia (34 per 100,000; 76 diagnoses) and New South Wales (2.8 per 100,000 population; 39 diagnoses) (Figure 3.4) (Supplementary Table 3.2).

Figure 3.4: ARF notifications among Indigenous Australians, by state and territory, 2017–2021



Notes

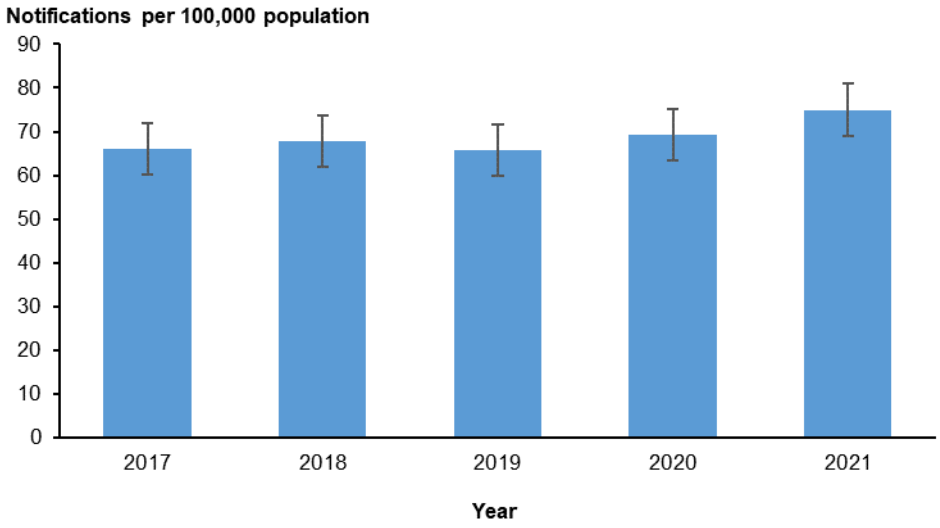
1. Crude age-specific rates per 100,000 calculated using the number of notifications of each calendar year divided by the corresponding 30 June populations of each year based on the 2016 Census.
2. ARF notifications include all recurrence categories and diagnostic categories.

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

Time trend

There was no significant change in the ARF diagnosis rate from 2017 to 2021. The ARF diagnosis rate among Indigenous Australians increased from 66 to 75 per 100,000 population over the period (474 to 582 diagnoses, respectively), but there was a dip to 66 per 100,000 (579 diagnoses) in 2019, followed by an increase in ARF diagnosis in 2020 and 2021 (Figure 3.5) (Supplementary Table 3.3). The ARF diagnosis rates in 2020 and 2021 might be affected by use of health services due to the impact of COVID-19 on the health sector.

Figure 3.5: ARF notifications among Indigenous Australians, by year, 2017–2021



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.
4. Whiskers show 95% confidence intervals around the rate estimate. This describes the range of values within which we can be 'confident' the true value lies.

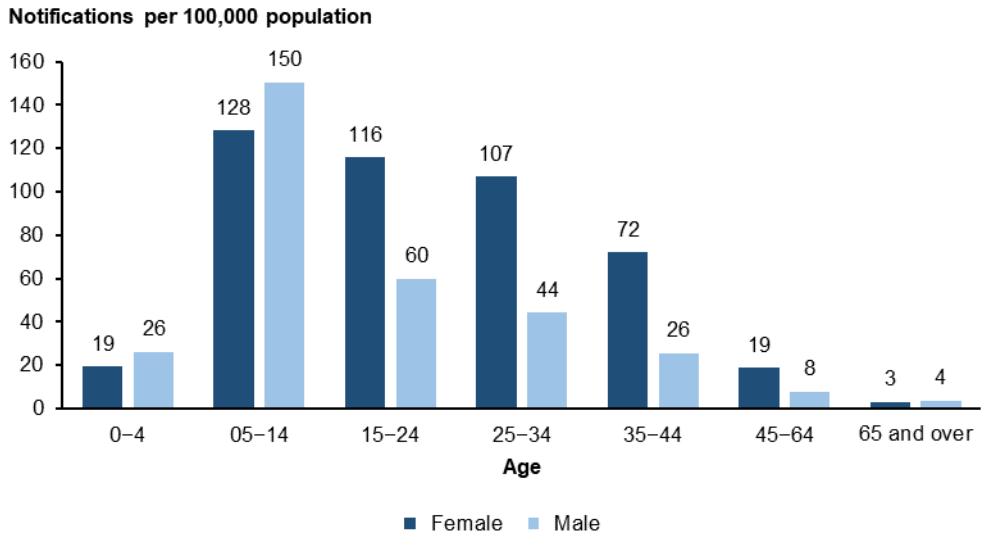
Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Age and sex

Of the 2,570 ARF diagnoses among Indigenous Australians in 2017–2021:

- the highest rate of diagnosis was among those aged 5–14, accounting for 44% of all diagnoses (140 per 100,000 population, or 1,142 diagnoses)
- there were 97 diagnoses (23 per 100,000) among children aged 0–4
- females accounted for 57% of diagnoses (1,465 diagnoses)
- in people aged under 15, rates were higher among males than females.
- for adults, rates were higher among females than males (Figure 3.6) (Supplementary Table 3.4b).

Figure 3.6: ARF notifications among Indigenous Australians, by sex and age group, 2017–2021



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. This represents the location of the health service where the person first presented with symptoms of ARF, although a formal diagnosis may be made in a hospital elsewhere. 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 2017–2021, the Northern Territory accounted for 55% of the 2,570 ARF diagnoses among Aboriginal and Torres Strait Islander people (1,426 diagnoses). Queensland accounted for a further 25% of cases, Western Australia for 15%, South Australia for 3.0%, and New South Wales for 1.5% (Supplementary Table 3.2).

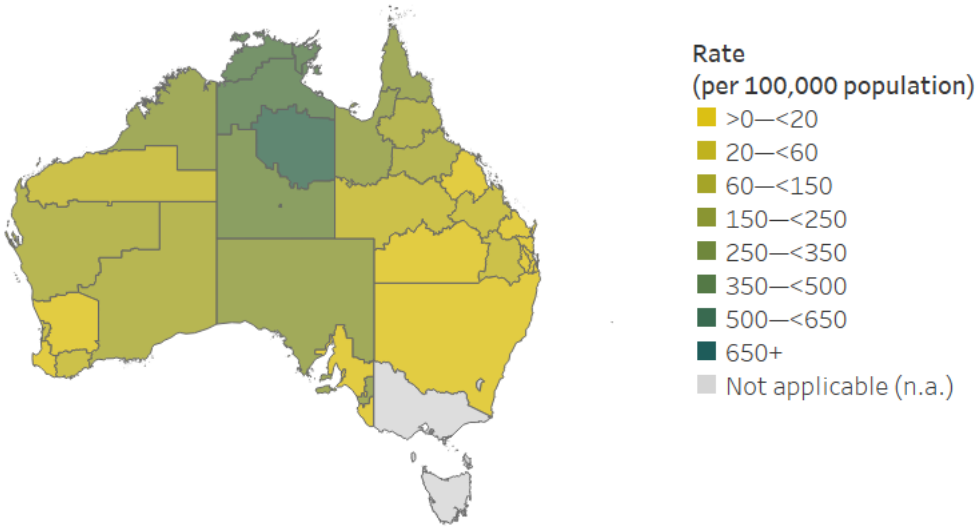
In 2017–2021, the highest 3 rates of ARF diagnoses were reported in regions in the Northern Territory:

- Urban Alice Springs with 830 per 100,000 (286 diagnoses)
- Barkly with 609 per 100,000 population (134 diagnoses)
- Rural Darwin with 465 per 100,000 population (290 diagnoses) (Figure 3.7) (Supplementary Table 3.5).

In 2017–2021, the 3 regions with the highest numbers of ARF diagnoses were also in the Northern Territory:

- Rural Darwin with 290 diagnoses
- Urban Alice Springs with 286 diagnoses
- East Arnhem with 220 diagnoses (Figure 3.7) (Supplementary Table 3.5).

Figure 3.7: ARF diagnoses among Indigenous Australians, by region of diagnosis, 2017–2021



Notes

1. Crude rates (per 100,000 population) are calculated using the number of diagnoses for the 5-year period, divided by the population provided by the state or territory for each region, with the exception of NSW which is from the Australian Bureau of Statistics. NSW, SA, and NT populations have up-to-date data. WA uses 2019 data as a proxy for 2020 and 2021 data. Qld uses 2020 data as a proxy for 2021 data.

2. ARF notifications include all recurrence categories and diagnostic categories.

3. The data exclude 101 ARF diagnoses nationally where the region and state were unknown or diagnosed overseas.

Source: AIHW analysis of National Rheumatic Heart Disease data collection. See Supplementary Table 3.5.

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 (RHDAustralia 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 C1 of this report.

Source: RHDAustralia 2020

In 2017–2021, of the 2,570 ARF diagnoses among Indigenous Australians, 36% had at least 1 manifestation of carditis, prolonged P-R interval, or Sydenham chorea (Supplementary Table 3.6). 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 2017–2021, of all 2,570 ARF diagnoses among Indigenous Australians:

- 2,054 diagnoses (80%) were definite or probable diagnoses (Box 3.2)
- 516 were possible diagnoses (Supplementary Table 3.7).

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'). 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. They are 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 RHDAustralia 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: clinical presentation falls short by either 1 major or 1 minor manifestation, or the absence of streptococcal serology results, but where ARF is suspected.

Source: RHDAustralia 2012.

Between 2017 and 2021:

- the rate of definite or probable ARF diagnoses was 57 per 100,000 (411 diagnoses) in 2017, decreasing to 52 per 100,000 (396 diagnoses) in 2020 and then increasing to 59 per 100,000 in 2021
- the rate of possible ARF episodes increased, from 8.8 per 100,000 (63 diagnoses) in 2017 to 16 per 100,000 (127 diagnoses) in 2021

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 (Figure 1.2). After the first known ARF episode, adherence to secondary prophylaxis reduces the likelihood of recurrence.

Of the 2,570 ARF diagnoses among Indigenous Australians, regardless of prophylaxis prescription, , 728 (28%) were recurrent cases in these jurisdictions (Supplementary Table 3.8).

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

Deaths among people with a history of ARF (no RHD)

In 2017–2021, 42 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, 37 deaths were recorded on these registers. The median age at death for Indigenous Australians with ARF was 43, with half (51%) of those who died being aged under 45 (Supplementary tables 3.9a and 3.9b).

Table 3.1: Deaths among people with ARF recorded on jurisdictional registers, by Indigenous status, 2017–2021

Age group	Indigenous	All Australians
0–24	n.p.	n.p.
25–44	n.p.	n.p.
45–64	12	14
65+	6	8
All ages	**37**	**42**

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

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

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 2021, 6,749 people (67 per 100,000 population) were recorded as having RHD on registers in Queensland, Western Australia, South Australia, and the Northern Territory (Supplementary Table 4.1). Of these:

- 78% were Indigenous Australians (5,238 diagnoses, 1,083 per 100,000 population)
- 29% were aged under 25 at 31 December 2021 (1,952 diagnoses)—with fewer than 5 aged under 5
- 66% were female (4,447 diagnoses)
- the Northern Territory had the highest prevalence rate (2,451 diagnoses, 984 per 100,000) (Supplementary tables 4.1 and 4.2a).

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 and recommended follow-up

	2012 Guideline definition	2020 Guideline definition	2020 Guideline recommended follow-up plan[(a)(b)]
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	• Specialist review: at least 6 monthly • Echocardiogram: at least 6 monthly • Medical review: at least 6 monthly • Dental review: within 3 months of diagnosis, then 6 monthly • Follow-up plan for pregnant women depends on severity of 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	• Specialist review: yearly • Echocardiogram: yearly • Medical review: 6 monthly • Dental review: within 3 months of diagnosis, then 6 monthly
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	• Specialist review: 1-3 yearly • Echocardiogram: 1-2 yearly up to 21 years; 2-3 yearly if over 21 • Medical review: yearly • Dental review: yearly Borderline cases should have medical review and ECG 1-2 years after diagnosis and 1-2 years after ceasing secondary prophylaxis
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	• Specialist referral and Echocardiogram 1, 3 and 5 years after ceasing secondary prophylaxis (or after diagnosis, for borderline cases not receiving BPG) • Medical review: yearly until discharge from specialist care, then as required • Dental review: yearly or as required

Echocardiogram (an ultrasound of the heart, used to diagnose and monitor heart problems)

(a) Frequency of follow-up is a guide only and should be tailored to specific individuals following specialist assessment.

(b) All patients should receive an influenza vaccine annually and pneumococcal vaccines as recommended by the National Immunisation Program schedule.

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

Of the 4,345 people diagnosed with RHD with a priority status recorded for their most recent evaluation, 22% were priority 1 (965 people), 22% were priority 2 (963), 48% were priority 3 (2,105), and 7% (315) were priority 4 (Supplementary Table 4.3).

RHD in New South Wales

At 31 December 2021, 120 people (1.5 per 100,000 population) with RHD 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:

- 33% (40 diagnoses, 14 per 100,000 population) were Indigenous and 66% (79 cases, 1.0 per 100,000) were non-Indigenous
- the median age of people on the register at the end of 2021 was 24.7 (Supplementary Table 4.2b).

Priority status of RHD diagnoses in New South Wales

7 diagnoses of RHD had a current priority status recorded. There were too few to include further breakdowns.

Incidence of RHD in 2017–2021

In this report, incidence (a 'new' RHD diagnosis) is defined as one that was diagnosed between 1 January 2017 and 31 December 2021. 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.

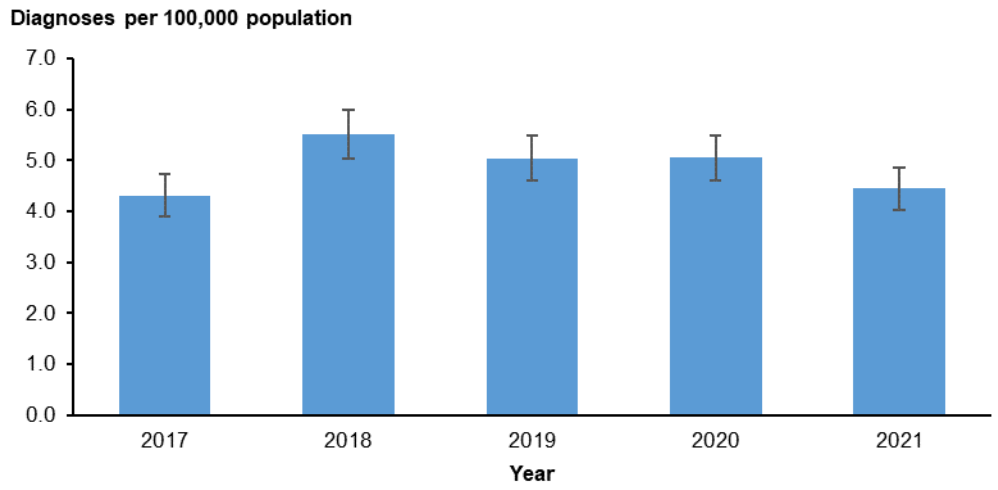
New diagnoses in Queensland, Western Australia, South Australia and the Northern Territory

All Australians

In 2017–2021:

- there were 2,381 reports of new RHD diagnoses in Queensland, South Australia, Western Australia and the Northern Territory (4.9 per 100,000 population)
- there was no discernible consistent trend in new RHD diagnosis from 2017 to 2021. Diagnosis rates increased between 2017 and 2018 from 4.3 to 5.5 diagnoses per 100,000. Since then, the rate has fallen to 4.5 per 100,000 in 2021 (Figure 4.1)
- diagnosis rates varied by state and territory. The Northern Territory had 61 diagnoses per 100,000 people, followed by Queensland (4.7 diagnoses per 100,000) and Western Australia (2.5 diagnoses per 100,000). South Australia had 1.0 diagnosis per 100,000 (Figure 4.2) (Supplementary Table 4.4a).

Figure 4.1: New RHD diagnoses, all Australians in Qld, WA, SA and NT, by year, 2017–2021

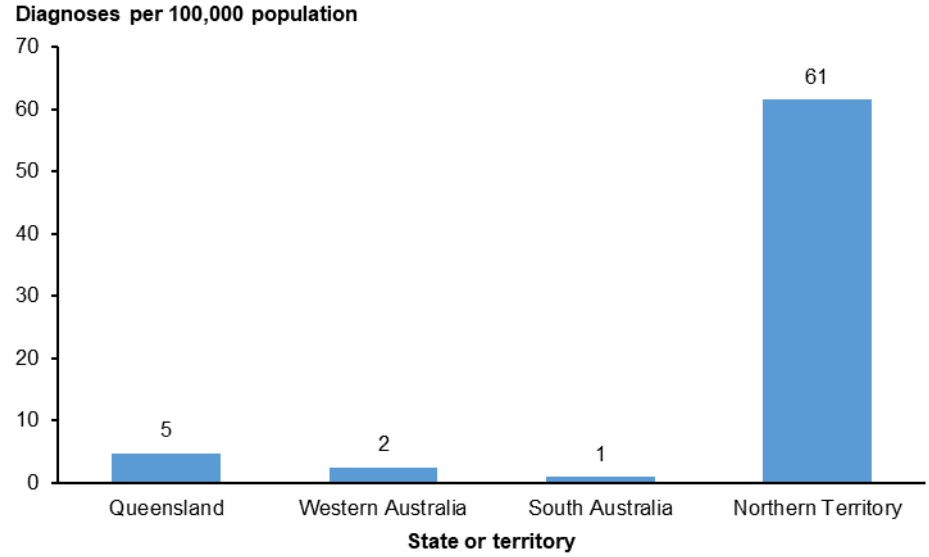


Notes

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. 12 people with an unknown or blank Indigenous status are included in All Australians.
3. Data from Qld, WA, SA and NT combined.
4. Whiskers show 95% confidence intervals around the rate estimate. This describes the range of values within which we can be 'confident' the true value lies.

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

Figure 4.2: New RHD diagnoses, all Australians, by state and territory, 2017–2021



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.

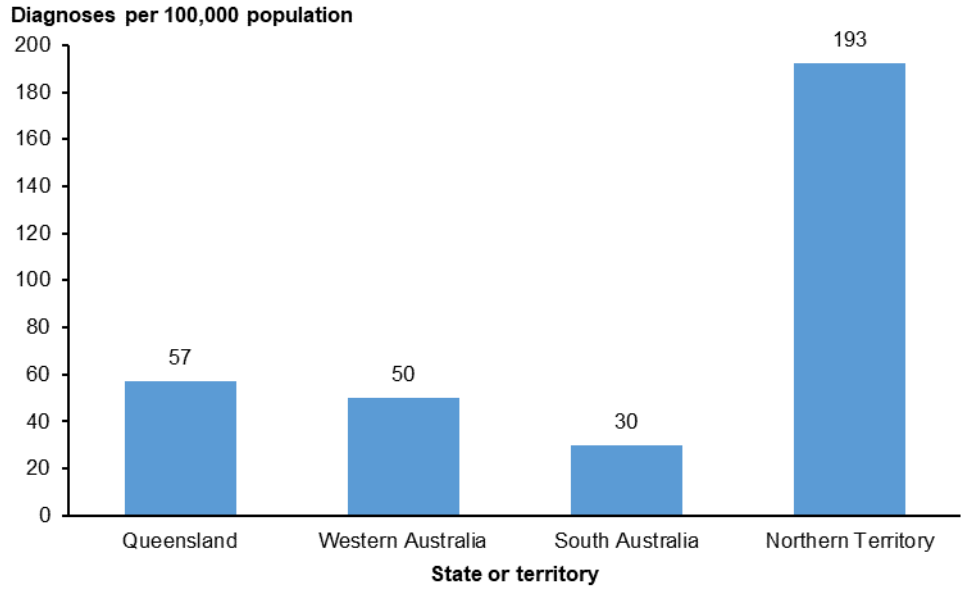
Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Indigenous Australians

In 2017–2021:

- of the 2,381 new RHD diagnoses among all Australians in Queensland, Western Australia, South Australia and the Northern Territory, 74% (1,750) were Indigenous Australians (75 per 100,000 population)
- the overall diagnosis rate for Indigenous Australians was 15 times the rate for all Australians (75 per 100,000, compared with 4.9 per 100,000, respectively)
- the Northern Territory had the highest rate of new RHD diagnoses among Indigenous Australians (193 per 100,000 population) followed by Queensland (57 per 100,000; Figure 4.3), Western Australia (50) and South Australia (30)
- the annual combined rate fluctuated between from 68 and 86 per 100,000 (303 diagnoses to 393 diagnoses) with no discernible pattern (Figure 4.4) (Supplementary tables 4.4a and 4.5a).

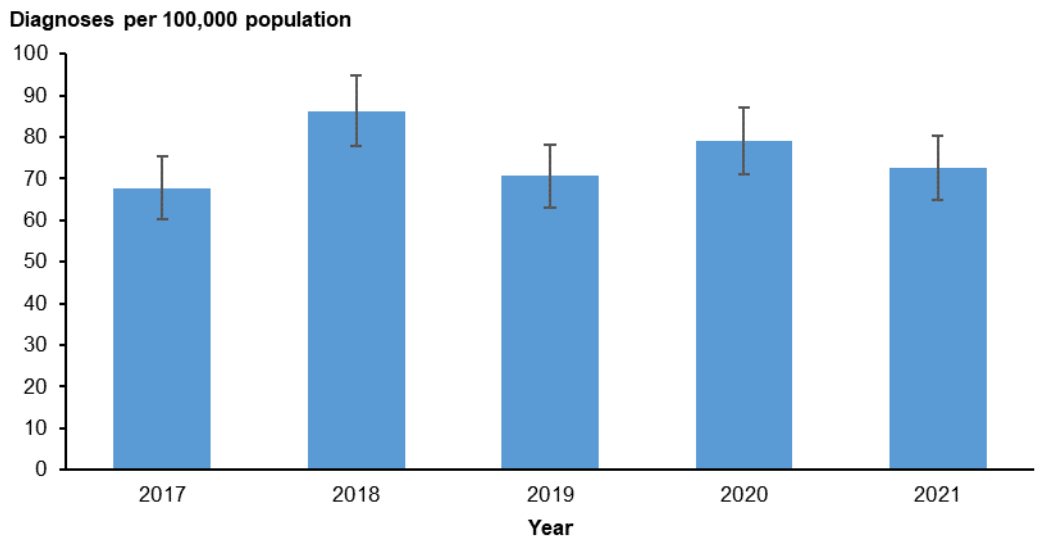
Figure 4.3: New RHD diagnoses, Indigenous Australians, by state and territory, 2017–2021



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, 2017–2021



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.

3. Whiskers show 95% confidence intervals around the rate estimate. This describes the range of values within which we can be 'confident' the true value lies.

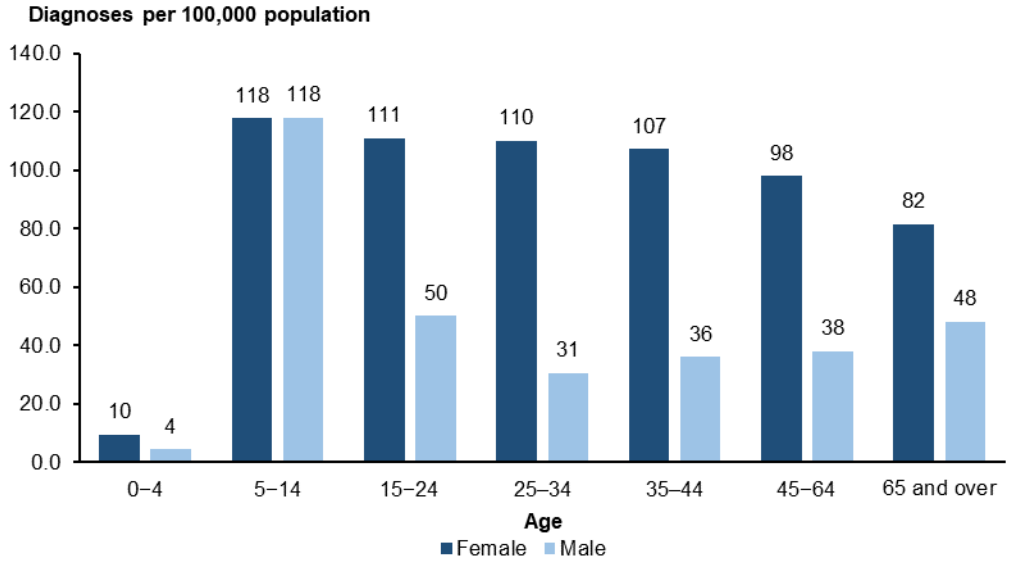
Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Sex and age

In 2017–2021, for all new RHD cases diagnosed among Indigenous Australians:

- the rate of new RHD diagnosis for females was 1.8 times that for males (97 and 53 diagnoses per 100,000 population, respectively)
- females had higher rates than males in all age groups, excluding those aged 5–14
- 55% were aged under 25 at diagnosis (970 people)
- 18 children were aged under 5 and 597 children were aged 5–14 at the time of RHD diagnosis (Figure 4.5)
- The median age at diagnosis for Indigenous Australians was 21 (15 for males and 26 for females), compared with age 56 for non-Indigenous Australians (Supplementary tables 4.5a and 4.6).

Figure 4.5: New RHD diagnoses, Indigenous Australians, by age group and sex, 2017–2021



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, or mild, as 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 is a distinct diagnosis from RHD; data on diagnoses of borderline RHD were not available for this report.

In 2017–2021, of the 1,498 Indigenous Australians with severity recorded at a new RHD diagnoses:

- 49% had mild RHD when first diagnosed (740 diagnoses)
- 32% had moderate RHD (473)
- 19% had severe RHD (285) (Supplementary Table 4.7).

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	ARF with no evidence of RHD *Or* trivial to mild valvular disease
Borderline	Individual ages 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

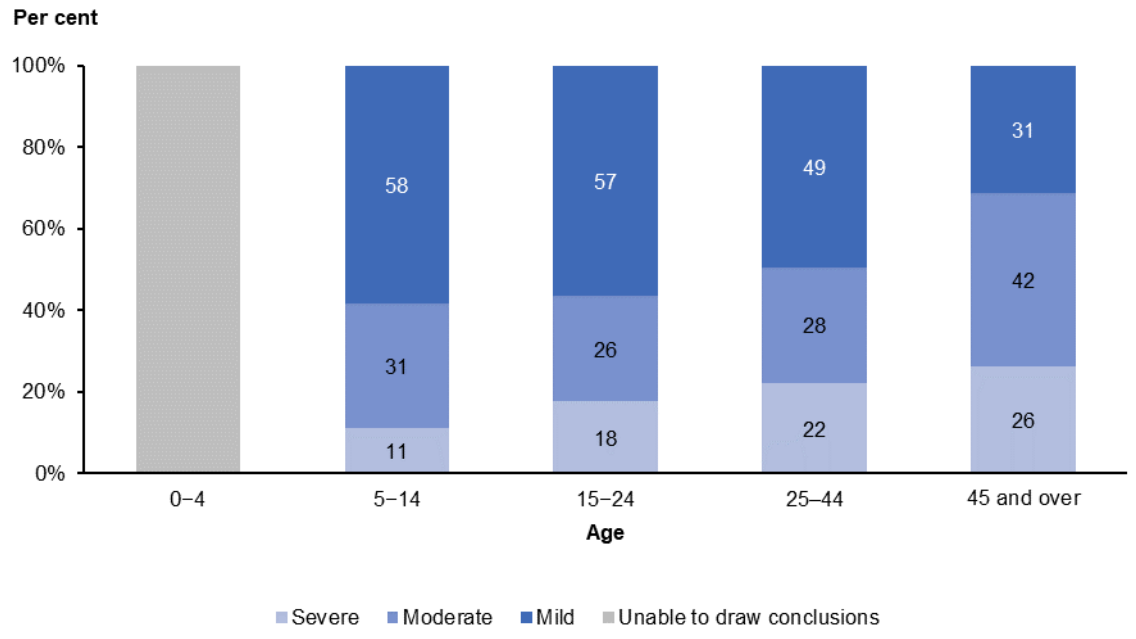
By state or territory:

- Queensland, Western Australia, South Australia and the Northern Territory had similar distributions of cases, with mild severity at diagnosis being the most common
- severe cases were the least common in all 4 jurisdictions, but Queensland had the lowest proportion: 16% at diagnosis (Supplementary Table 4.7).

By age group:

- there was a relatively large proportion of severe cases in the 45 and over age group (26%, 87 diagnoses)
- the proportion of mild cases was highest in young people, with 58% (264 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 Table 4.7).

Figure 4.6: New RHD diagnoses, Indigenous Australians, by age group and severity status at diagnosis, 2017–2021



Notes

1. RHD severity assessment is at the time of diagnosis. These data exclude 252 cases with no assessment recorded.
2. Data includes Qld, WA, SA and the NT.
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 (for example, because the patient did not seek medical care, or their symptoms were not recognised as possibly being ARF).

Among all Indigenous Australians with a new RHD diagnosis in 2017−2021, four-fifths (79%, 1,390 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 (79%) and males (80%)
- the proportion varied across the jurisdictions, being highest in Western Australia (91%) and lowest in Queensland (73%) (Figure 4.7)
- the proportion was lowest in those aged 25–34
- there was no clear change in the proportion of people without a history of ARF between 2017 and 2021.

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 Table 4.8a).

Figure 4.7: New RHD diagnoses, Indigenous Australians with no previous ARF recorded, by state or territory, 2017–2021

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 2017–2021:

- there were 64 reports of new RHD diagnoses in New South Wales (0.2 per 100,000 population)
- there were more diagnoses of RHD in non-Indigenous Australians (43 diagnoses, 67%) than Indigenous Australians (21 diagnoses, 33%).
- Pacific Islanders made up the largest proportion of cases in a known risk group (17 diagnoses, 27%)
- cases were more likely to be female (37 cases, 58%) than male (27 cases, 42%)
- the 25–34 age group had the most diagnoses (28 diagnoses, 44%). RHD is not notifiable in NSW in people aged 35 or over at the time of diagnosis
- annual RHD diagnosis rates have fluctuated between 0.1 and 0.3 diagnoses per 100,000 population, likely due to low numbers (Supplementary tables 4.4b and 4.5b).

Indigenous Australians

Of the 64 new RHD diagnoses among all Australians in New South Wales in 2017–2021:

- 33% (21) were Indigenous Australians (1.5 per 100,000 population)
- the overall diagnosis rate for Indigenous Australians was 9.3 times the rate for all Australians (1.5 compared with 0.2 per 100,000)

- the annual rate for Indigenous Australians fluctuated between 0.7 and 2.6, due to the small number of diagnoses. There were too few cases among Indigenous Australians to draw meaningful conclusions about change over time (Supplementary tables 4.4b and 4.5b).
- Most (95%, 20 diagnoses) 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 Table 4.8b).

In New South Wales, severity information is recorded only for patients who consent to join the Register. Most Indigenous cases where severity was recorded had moderate or severe RHD. This may not reflect the severity distribution among all notified cases.

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 and anti-coagulant medication 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 2017–2021, 626 people diagnosed with RHD and living in Queensland, Western Australia, South Australia, or the Northern Territory had surgery. Of those, 3 in 5 (59%; 368 people) were Indigenous Australians who underwent 396 surgical events (Supplementary Table 4.9a). Of these Indigenous Australians:

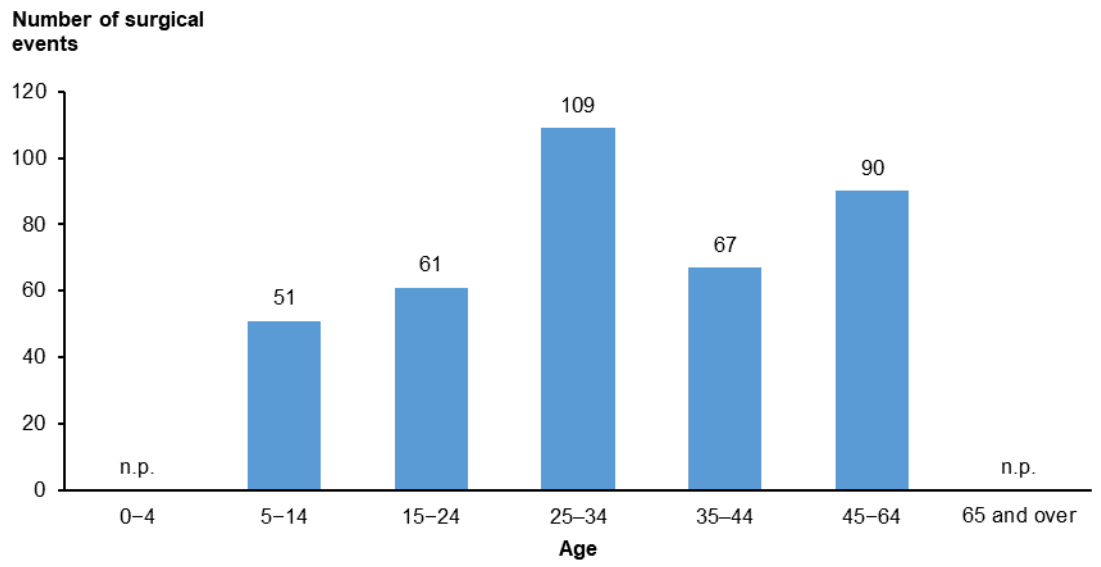
- 93% (342 patients) had surgery once and 7.1% (26 patients) had at least 2 surgeries (Supplementary Table 4.10)
- 182 (49%) were from the Northern Territory
- 102 (28%) were from Queensland
- 250 (68%) were female (Supplementary Table 4.11).

On average, people had their first surgery 7.0 years after RHD diagnosis (Supplementary Table 4.11).

In 2017–2021, of the 396 surgical events among 368 Indigenous Australians:

- most surgical events occurred among those aged 25–34, who had 109 surgical events (28%), followed by those aged 45–64, with 90 surgical events (23%)
- children aged 5–14 with RHD had 51 (13%) surgical events.
- Fewer than 5 children aged under 5 years had surgery for RHD (Figure 4.8) (Supplementary Table 4.11).

Figure 4.8: Surgical events among Indigenous Australians with RHD, by age group, 2017–2021



Notes

1. There were fewer than 5 surgical events for RHD among those aged 0–4 during 2017–2021.
2. This shows the age at surgery of the 396 RHD-related surgical events for Indigenous Australians during 2017–2021. 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, surgery is data is only provided for those who consent to the Register. Consent is further discussed in the Secondary Prophylaxis chapter. In 2017–2021, 12 people underwent 12 RHD surgical events and fewer than 5 of these were among Indigenous Australians.

As such, there are too few cases to undertake further analysis (Supplementary Table 4.9b).

All-cause deaths recorded among people with a history of 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 2017–2021 in Queensland, Western Australia, South Australia and the Northern Territory, 595 deaths were reported among people registered as having RHD (Table 4.3). Of these deaths:

- 320 deaths occurred in Queensland (54%) (Supplementary Table 4.12)
- 389 deaths occurred among females (65%)
- 265 deaths occurred among people aged 65 and over (45%)
- the median age of death was 60 years for males and 63 years for females (Supplementary Table 4.13).

Indigenous Australians

In 2017–2021, 382 deaths (64% of all RHD deaths) were reported among Indigenous Australians registered as having RHD. Of these deaths:

- the highest number occurred in the Northern Territory (192 deaths, 50% of all Indigenous deaths) (Supplementary Table 4.12)
- none occurred in Indigenous Australians aged under 15 (Supplementary Table 4.13).

Table 4.3: Deaths among Indigenous Australians recorded on RHD jurisdictional registers, 2017–2021

Age group	Number	Per cent
0–24	22	6.6
25–44	102	27
45–64	158	41
65+	100	26
All ages	**382**	**100**

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

The median age of death was 51 for Indigenous males and 56 for Indigenous females. In comparison, the median age at death for non-Indigenous Australians with RHD was 73 for non-Indigenous males and 74 for non-Indigenous females. Indigenous Australians with RHD who died during 2017–2021 had lived with their diagnosis for a median of 11.4 years (Supplementary tables 4.12 and 4.13).

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 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; RHDAustralia 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 (RHDAustralia 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 2021, 40% of Indigenous Australian patients had consented to have their prophylaxis data recorded on the register. 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 2021.

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 2021 were also included in the analysis.

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

There were 4,816 Aboriginal and Torres Strait Islander people eligible for inclusion in calculations about BPG delivery in 2021. They were located in the Northern Territory (2,447), Queensland (1,350), Western Australia (833), and South Australia (186). Of these:

- 18% (875 people) received 100% or more of their prescribed doses
- 13% (638) received 80% to 99% of their prescribed doses
- 27% (1,319) received 50% to 79% of their prescribed doses
- 41% (1,984) received less than 50% of their prescribed doses, including 514 people who did not receive any doses (Table 5.1) (Supplementary Table 5.1).

In general, a lower proportion of doses of prophylaxis was received in 2021 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 2021

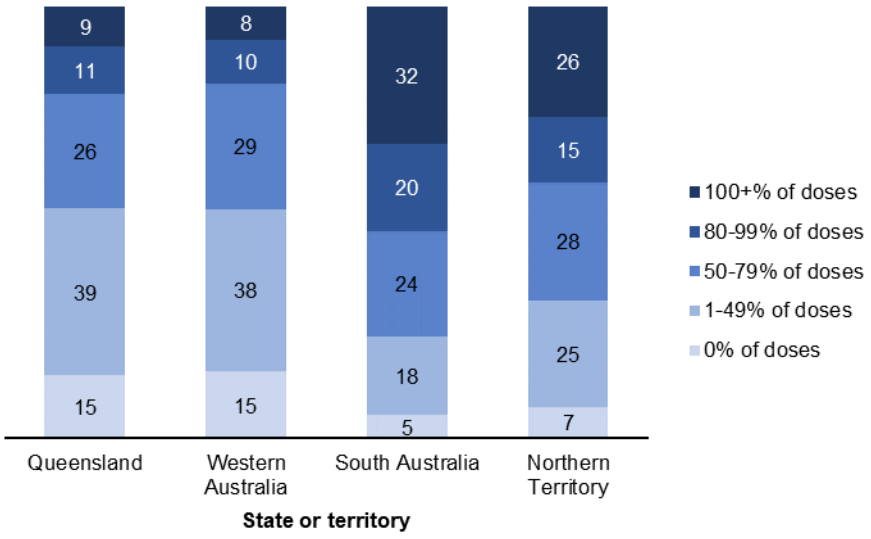
	Proportion of doses delivered	Number	Per cent
Qld, WA, SA, NT			
	0% of doses	514	10.7
	1–49% of doses	1,470	30.5
	50–79% of doses	1,319	27.4
	80–99% of doses	638	13.2
	100%+ of doses	875	18.2
NSW			
	0% of doses	8	25.0
	1–39% of doses	10	31.3
	40–79% of doses	5	15.6
	80–100%+ of doses	9	28.1

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

In 2021, 31% of Indigenous Australians (1,513 people) received at least 80% of doses. The proportion of people who received at least 80% of prescribed doses in each jurisdiction was:

- 20% in Queensland (274 people)
- 18% in Western Australia (148)
- 52% in South Australia (97)
- 41% in the Northern Territory (994) (Figure 5.1; Supplementary Table 5.1).

Figure 5.1: Proportion of doses received by Indigenous Australians with ARF and/or RHD on a BPG regime, by state and territory, 2021



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 2021, 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.

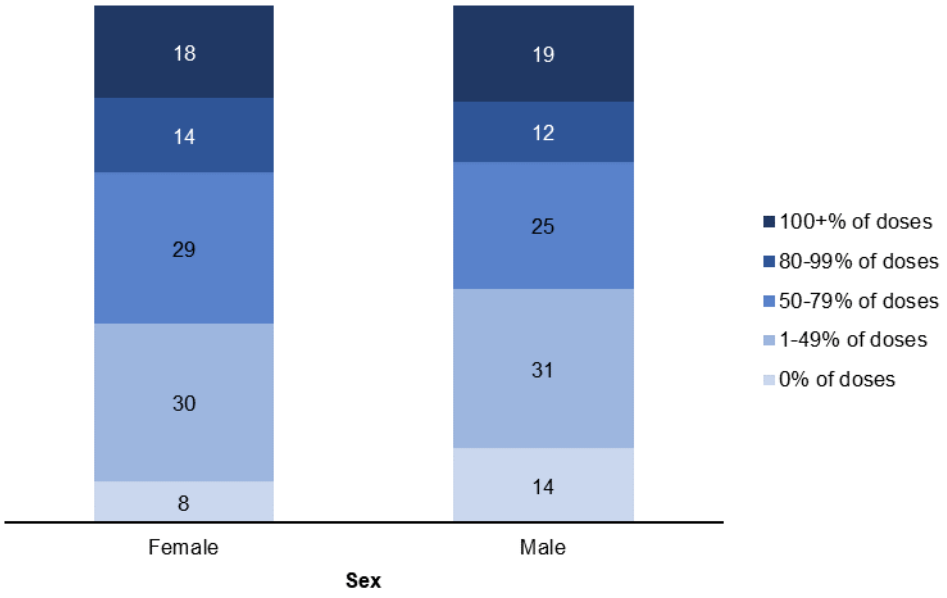
Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Age and sex

In 2021, among Indigenous Australians prescribed prophylaxis:

- more males received fewer than 40% of their prescribed doses (39%) than females (32%) and slightly more females received at least 80% of doses than males (32% and 30%, respectively) (Figure 5.2) (Supplementary Table 5.2).
- delivery was highest among people aged 5–14 with 48% receiving at least 80% of doses (Figure 5.3) (Supplementary Table 5.3).

Figure 5.2: Proportion of doses received by Indigenous Australians with ARF and/or RHD on a BPG regime, by sex, 2021

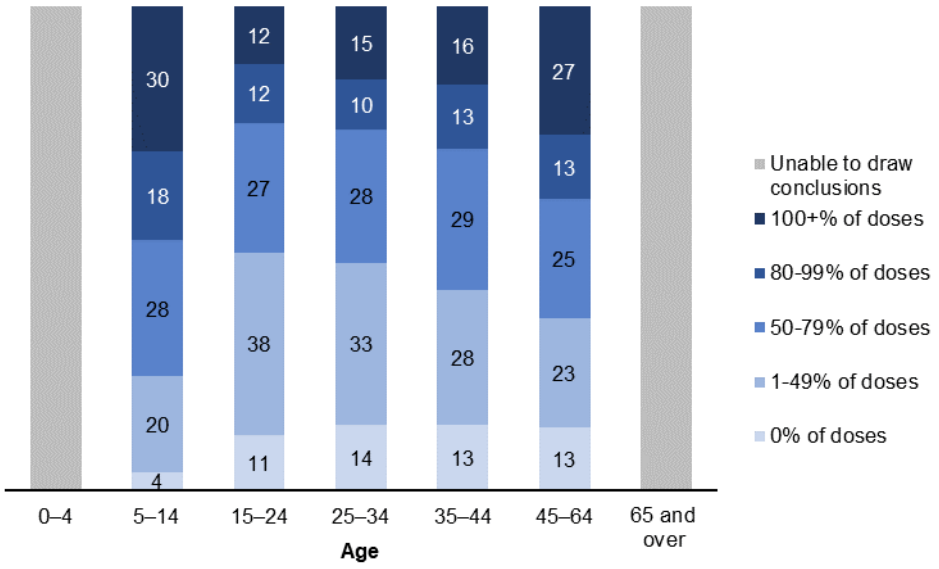


Notes

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 2021, 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, 2021



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 2021, as well as those on BPG for part of the year only.

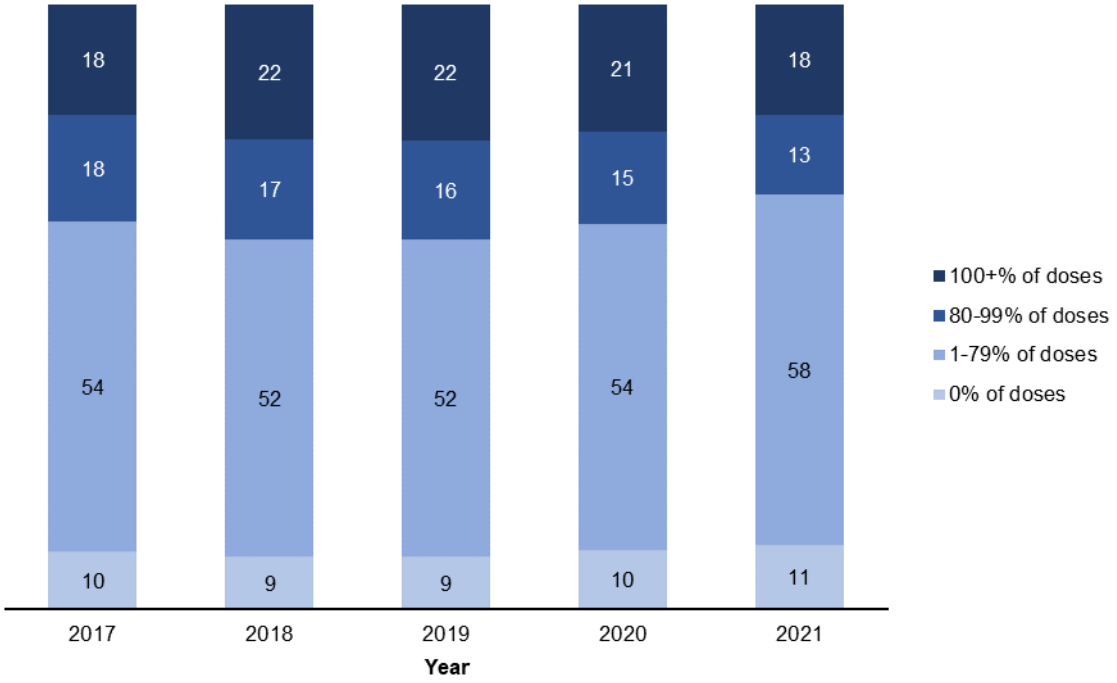
3. Data include Qld, WA, SA and NT. Does not include cases managed in NSW.

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 2017 to 39% in 2019 (Figure 5.4). However, there was a decrease since 2020 and only 31% of Indigenous Australians prescribed BPG in 2021 received at least 80% of their doses. In 2021, 11% did not receive any of their prescribed doses (Supplementary Table 5.4). 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, availability and accessibility of clinics, and concern around being exposed to COVID-19.

Figure 5.4: Proportion of doses received by Indigenous Australians with ARF and/or RHD on a BPG regime, by year, 2017–2021



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

As noted above, information on secondary prophylaxis in New South Wales is only recorded for patients who consent to join the Register. This information was available for 32 Indigenous Australians during 2021. Of these, 28% received at least 80% of their scheduled doses (Supplementary Table 5.5b).

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

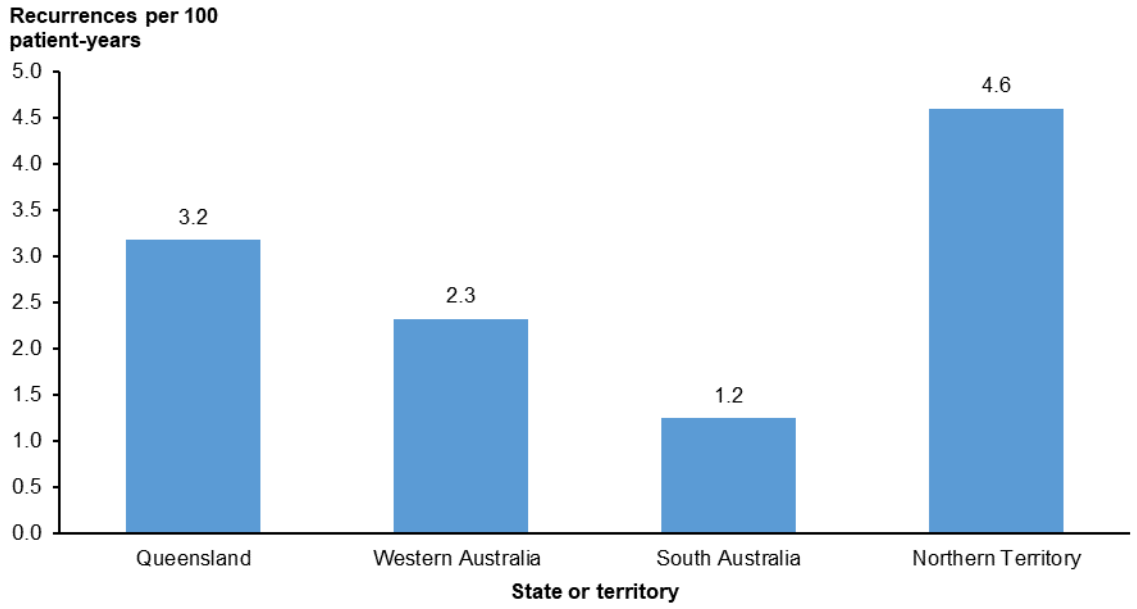
At 31 December 2021:

- among 4,795 Indigenous Australians who had been prescribed BPG, 163 ARF recurrences were reported (3.7 recurrences per 100 patient-years) (Table 5.2)
- the greatest number of recurrences was in the Northern Territory (102 recurrences)
- the rate of ARF recurrences per 100 patient-years was highest in the Northern Territory (4.6) and lowest in South Australia (1.2) (Figure 5.5)
- the rate of recurrence per 100 patient-years generally decreased with age, with the highest risk among those aged 5–14 (5.3). There were too few recurrences in those aged under 5 and aged 65 and over to draw conclusions. (Figure 5.6) (Supplementary Table 5.6).

Table 5.2: ARF recurrences in 2021

	Number	Rate per 100 patient-years
Qld, WA, SA, NT	163	3.7
NSW	0	0

Figure 5.5: ARF recurrences per 100 patient-years, Indigenous Australians, by state and territory, 2021

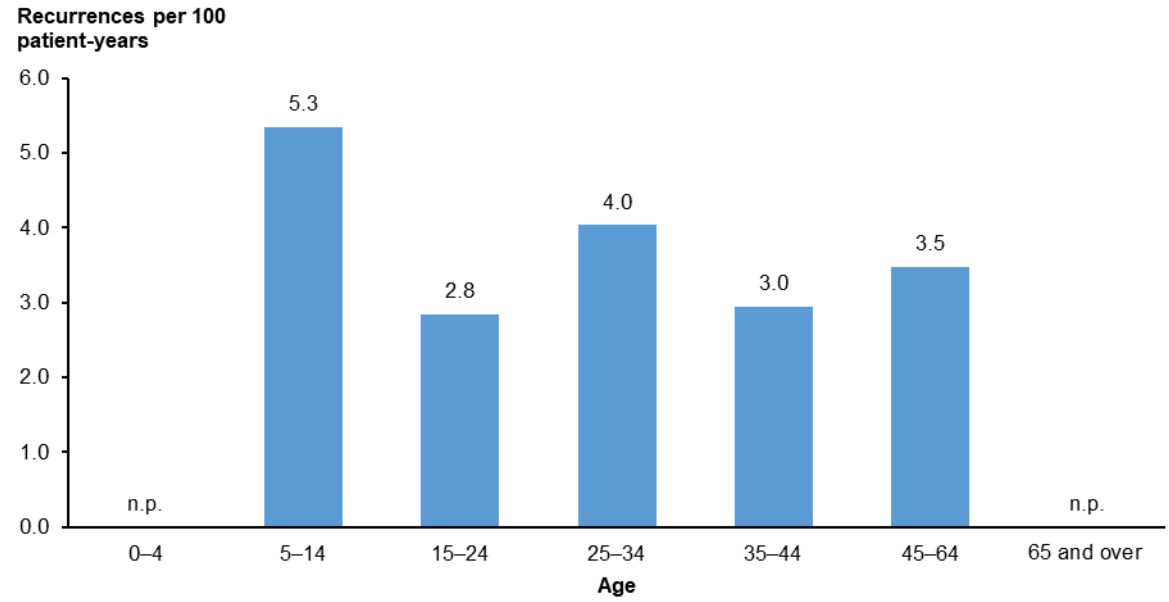


Notes

1. Includes people who were prescribed BPG before or during 2021.
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, 2021



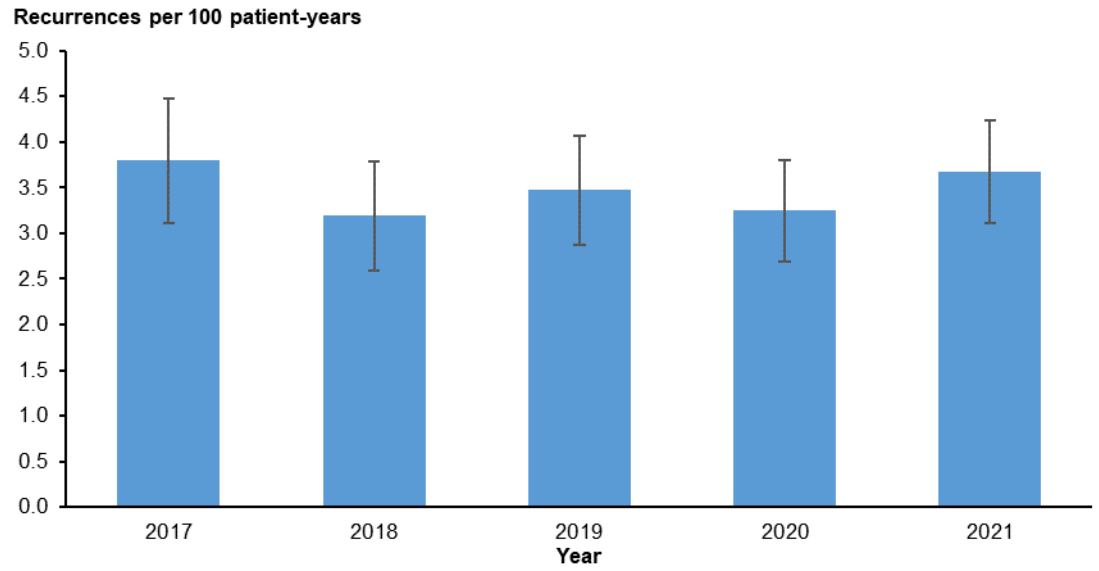
Notes

1. Includes people who were prescribed BPG before or during 2021.
2. Data from Qld, WA, SA and NT.
3. There were too few cases in those under 5 and 65 and older to calculate meaningful recurrence rates.

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

From 2017 to 2021 for Queensland, Western Australia, South Australia, and the Northern Territory, the ARF recurrence rate per 100 patient-years among Indigenous Australians prescribed BPG fluctuated around 3.5 recurrences per 100 patient-years (Figure 5.7) (Supplementary Table 5.7).

Figure 5.7: Rate of recurrence, Indigenous Australians with ARF and/or RHD on BPG regime, by year, 2017–2021



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.
3. Whiskers show 95% confidence intervals around the rate estimate. This describes the range of values within which we can be 'confident' the true value lies.

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

ARF recurrence rates in New South Wales

In 2021, among the 32 Indigenous Australians for whom information on BPG was available, there were no reported ARF recurrences (0 recurrences per 100 patient-years) (Supplementary Table 5.6). 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 2021, 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 fifth 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 and Aged Care. In Queensland, the ARF/RHD control programs are funded by both the Australian Government Department of Health and Aged Care 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, people of Maori and Pacific Islander heritage, 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: RHDAustralia 2020.

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The Department of Health and Aged Care 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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