Australian Government
Australian Institute of Health and Welfare

AIHW

Acute Rheumatic Fever and Rheumatic Heart Disease in Australia

Blurb

This report presents data from the National Rheumatic Heart Disease data collection, collated from the acute rheumatic fever (ARF) and rheumatic heart disease (RHD) registers in Queensland, Western Australia, South Australia and the Northern Territory.

ARF and RHD are preventable diseases affecting disadvantaged populations. This report shows that ARF rates increase with remoteness, and that rates are highest in Aboriginal and/or Torres Strait Islander Australians, females and in young people aged 5–14 years.

Latest findings

1. At the end of 2017, the jurisdictional registers recorded almost 6,400 people with a past ARF and/or an RHD diagnosis. Most ARF diagnoses in 2013–2017 (94%, 1,776 diagnoses) were among Indigenous Australians.
2. The NT had the highest rate of ARF (260 per 100,000 population) among Indigenous Australians in 2013–2017.
3. The most common age for the first known ARF episode to occur was 5–14 years, with a rate of 195 per 100,000 population. In 2013–2017, 1,043 new RHD diagnoses were made amongst Indigenous Australians.
4. In 2013–2017, the rate of new RHD diagnoses was greatest in the NT (37 per 100,000 population).
5. Nearly 60% of Indigenous Australians diagnosed with RHD are under 25 when diagnosed.
6. In 2017, 36% of Indigenous Australians prescribed preventive penicillin received 80% or more of prescribed doses.

Topic/s

Primary topic

Aboriginal and/or Torres Strait Islander people

Additional topics

Rural and remote Australians, Indigenous Australians

Summary

Acute rheumatic fever (ARF) refers to an autoimmune response to infection of the upper respiratory tract (and possibly of the skin) by group A streptococcus (GAS) bacteria. ARF can affect the heart, joints, brain and subcutaneous tissues (the innermost layer of skin). While there is no lasting damage caused to the brain, joints and skin, ARF may cause lasting damage to the heart.

Rheumatic heart disease (RHD) is caused by damage to heart valves as a result of one or more ARF episodes. An affected heart valve may become scarred and stiffer, obstructing blood flow (stenosis), or it may fail to close properly, causing blood to flow backwards in the heart instead of forward around the body (regurgitation). Regurgitation due to damage to the mitral valve is common in RHD.

Jurisdictional RHD control programs and registers

Under the Rheumatic Fever Strategy, the Australian Government provides funding to support RHD control programs in 4 jurisdictions: Queensland (Qld), Western Australia (WA), South Australia (SA) and the Northern Territory (NT). These programs:

- identify people with or at risk of ARF and RHD
- promote primary prevention of ARF
- support the delivery of long-term secondary prevention treatment
- increase awareness of ARF and RHD among health professionals, and provide education about these diseases to health professionals, patients and their families and communities
- maintain disease registers for people diagnosed with ARF or RHD, and use this information to monitor health outcomes and improve control program activities.

How many people have acute rheumatic fever?

In 2013–2017, there were 1,897 diagnoses of ARF recorded, a rate of 4 per 100,000 population. Each year from 2013 to 2017, the number and rate of diagnoses increased.

In 2017, 528 individuals were diagnosed, a rate of 6 per 100,000 population, increasing from 3 per 100,000 population in 2013.

The most common age at diagnosis was 5–14 years, with 964 diagnoses (16 per 100,000 population). ARF was more common in females than males, with 56% of diagnoses (1,060) occurring in females.

How many Indigenous Australians have acute rheumatic fever?

In 2013–2017, there were 1,776 diagnoses of ARF among Indigenous Australians, a rate of 85 per 100,000 population. A higher rate of ARF diagnoses was recorded for Indigenous females (1,006 diagnoses, 96 per 100,000 population) than males (770 diagnoses, 74 per 100,000 population) and for the age group 5 to 14 (602 diagnoses, 195 per 100,000 population).

Numbers and rates of ARF amongst Indigenous Australians during 2013–2017 were consistently highest in the NT. Fifty-three per cent (954) of all diagnoses were from NT.

How many people have rheumatic heart disease?

As at 31 December 2017, there were 4,255 people living with RHD recorded on state and territory registers, of whom 87% were Indigenous Australians (3,687 diagnoses). NT has the greatest number of people living with RHD. The most common age group at diagnosis was 15–24 years, with 941 diagnoses (22%). Two-thirds of people living with RHD (65%, 2,783 diagnoses) were females.

How many Indigenous Australians have rheumatic heart disease?

In 2013–2017, there were 1,043 new diagnoses of RHD among Indigenous Australians, a rate of 50 per 100,000 population. The NT had the highest rate and greatest number of new RHD diagnoses among Indigenous Australians. The rate for Indigenous females was around two times the rate for Indigenous males and nearly 60% of new RHD diagnoses were made in people aged under 25 years.

How many Indigenous Australians are prescribed secondary prophylaxis?

Secondary prophylaxis with regular benzathine penicillin G (BPG) is the only RHD control strategy shown to be both clinically and cost effective at community and individual levels (RHD Australia, 2012). The recommended regimen to prevent recurrences of ARF and progression of RHD involves regular intramuscular injections of BPG every 21 to 28 days, for a minimum of 10 years.

In 2017 among Indigenous Australians prescribed BPG:

- 15% (394 people) received 100% or more of their prescribed doses
- 21% (548 people) received 80% to 99% of their prescribed doses

- 37% (964 people) received 50% to 79% of their prescribed doses
- 28% (724 people) received less than 50% of their prescribed doses.

Introduction

What is acute rheumatic fever?

Acute rheumatic fever (ARF) refers to an autoimmune response to infection of the upper respiratory tract—and possibly of the skin (McDonald et al. 2004)—by group A streptococcus (GAS) bacteria. 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 2015). ARF can affect the heart, joints, brain and subcutaneous tissues (inner most layer of skin) (Parnaby & Carapetis 2010) and can be extremely painful. While there is no lasting damage caused to the brain, joints and skin, ARF may cause lasting damage to the heart.

The risk of ARF recurrence is relatively high after an initial ARF episode and repeated episodes increase the likelihood of long-term heart valve damage, known as 'rheumatic heart disease' (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?

Rheumatic heart disease (RHD) is caused by damage to heart valves as a result of ARF. An affected heart valve may become scarred and stiffer, obstructing blood flow (stenosis), or it may fail to close properly, causing blood to flow backwards in the heart instead of forward around the body (regurgitation). Regurgitation due to damage to the mitral valve is the most common feature of RHD.

Symptoms of RHD include fatigue, chest pain, swelling of legs and face and shortness of breath. Diagnosis may 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 an RHD diagnosis. Many patients can remain asymptomatic despite having moderate or even severe RHD. Untreated RHD can cause arrhythmias (when the 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 (echo) to identify and monitor which valves are damaged and how badly. Management of an RHD diagnosis is complex and involves coordination of multiple services such as primary health care (for secondary prophylaxis with penicillin and monitoring of heart medications such as anticoagulation therapy), oral healthcare services, echo, specialist medical care, and other cardiothoracic and interventional cardiology services (RHD Australia 2012).

ARF and RHD are preventable diseases

RHD is a preventable and treatable disease. It is common in low- and middle-income countries (Wyber 2014, Webb 2015), and only in socioeconomically disadvantaged populations in high-income countries. Both ARF and RHD are linked with overcrowding, socioeconomic deprivation, and low levels of functioning 'health hardware' (for example toilets, showers, taps etc.) and lack of access to health care services (Webb 2015, Sims et al. 2016). Prevention measures that improve the living and environmental health conditions to the extent that ARF and RHD are no longer common in affected communities are known as primordial prevention measures. Improved living conditions and access to functional health hardware can reduce high rates of Group A streptococcal (GAS) infections and progression to ARF (Katzenellenbogen et al. 2017).

After a GAS infection, progression to ARF is preventable through early treatment. This is called primary prevention of ARF and relies on correct diagnosis and treatment within 9 days of onset of GAS throat infection (Gerber et al. 2009). People not seeking medical attention for a throat infection, or misdiagnosis, can affect timeliness of treatment, as well as a failure to prescribe treatment as recommended in clinical guidelines (RHD Australia 2012).

Secondary prevention of the progression from ARF to RHD relies on correct diagnosis of ARF, to enable commencement of regular antibiotic preventive medication. Correct diagnosis is challenging as there is no specific laboratory test for ARF. Diagnosis is based on clinical criteria outlined in the Australian modification of the Jones criteria (Appendix A), which takes into account Australia's high-risk groups, particularly Aboriginal and/or Torres Islander people (Carapetis et al. 2016). Guidelines recommend admission to hospital for clinical investigation and confirmation of the diagnosis (RHD Australia 2012).

For people with suspected or clinically confirmed ARF, benzathine penicillin G (BPG) is recommended in order to prevent further GAS infections and thereby reduce the risk of developing RHD or of RHD progression (Stollerman et al. 1955). BPG prophylaxis is clinically effective and cost-effective for RHD control at both individual and community levels (Webb 2015, Wyber 2015 and RHD Australia 2012).

Jurisdictional RHD control programs and registers

Under the Rheumatic Fever Strategy, the Australian Government provides funding to support RHD control programs in 4 jurisdictions: Queensland (Qld), Western Australia (WA), South Australia (SA) and the Northern Territory (NT). These programs:

- identify people with or at risk of ARF and RHD
- promote primary prevention of ARF
- support the delivery of long-term secondary prevention treatment

- increase awareness of ARF and RHD among health professionals, and provide education about these diseases to health professionals, patients and their families and communities
- maintain disease registers for people diagnosed with ARF or RHD, and use this information to monitor health outcomes and improve control program activities.

Information from the ARF/RHD registers in these 4 jurisdictions are compiled by the Australian Institute of Health and Welfare (AIHW) to provide information about ARF and RHD in Australia. The Australian ARF/RHD guideline describes a set of indicators recommended for monitoring ARF and RHD. This web-report presents data on those indicators that are currently able to be reported, including characteristics of people with ARF and RHD, geographic distribution of people with diagnoses, receipt of secondary prevention and use of surgical interventions.

Although an RHD control program and register also operates in New South Wales (NSW), this program is not currently covered under the Rheumatic Fever Strategy. Data from NSW are included as a standalone section.

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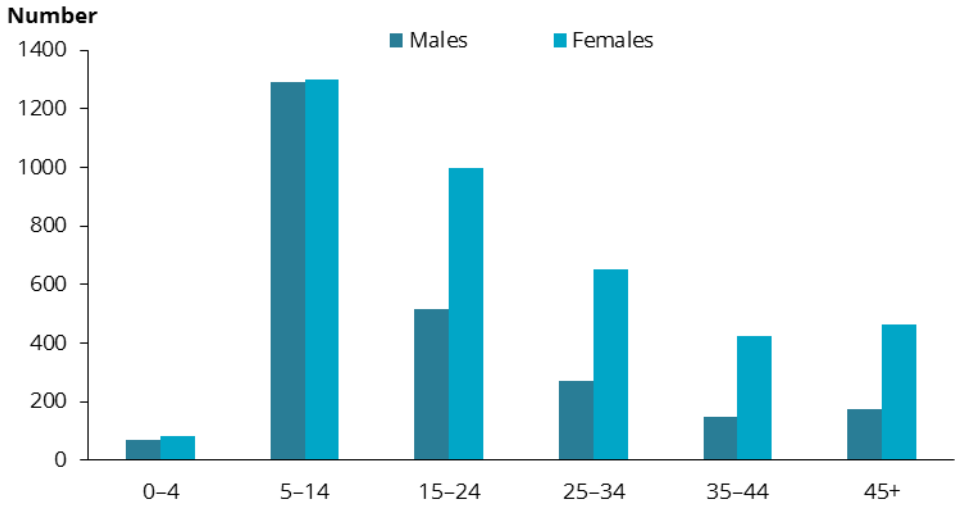
How many people have ARF and/or RHD?

Data from the 4 jurisdictional registers show that, at 31 December 2017, almost 6,400 living persons were recorded on one or more of the 4 jurisdictional registers. Of these:

- 9 in 10 (89%) were Indigenous Australians
- 6 in 10 (61%) were female
- 4 in 10 (41%) were aged 5–14 when registered for the first time.

The group was spilt evenly between those who only had ARF recorded, those who had only RHD recorded, and those with both ARF and RHD recorded.

Figure 1: People on ARF/RHD registers in Qld, WA, SA and the NT as at 31 December 2017, by sex and age at first registration



Note: ARF diagnoses includes all diagnostic categories and first and recurrent episodes. Refer to box 2 and 3 for more information.

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

Acute Rheumatic Fever

This section discusses diagnoses of acute rheumatic fever (ARF) reported by the RHD control programs. The total number of ARF diagnoses recorded is dependent on the reporting practices to the various RHD registers. Diagnoses made between 2013 and 2017 are the focus of this report. A person may have multiple diagnoses of ARF in their lifetime, so the number of diagnoses is greater than the number of people. This analysis focuses on the number of diagnoses.

It is likely that ARF diagnoses are under-reported to registers in all jurisdictions. Although the RHD registers in each state and territory were functional for the entire analysis period they were relatively new in some states (Box 1). Clinician awareness and reporting to the registers would have been increasing in the early years, and may still be improving. Under-diagnosis and under-reporting to the register means some individuals are not captured in this analysis.

Box 1: Acute rheumatic fever/rheumatic heart disease registers

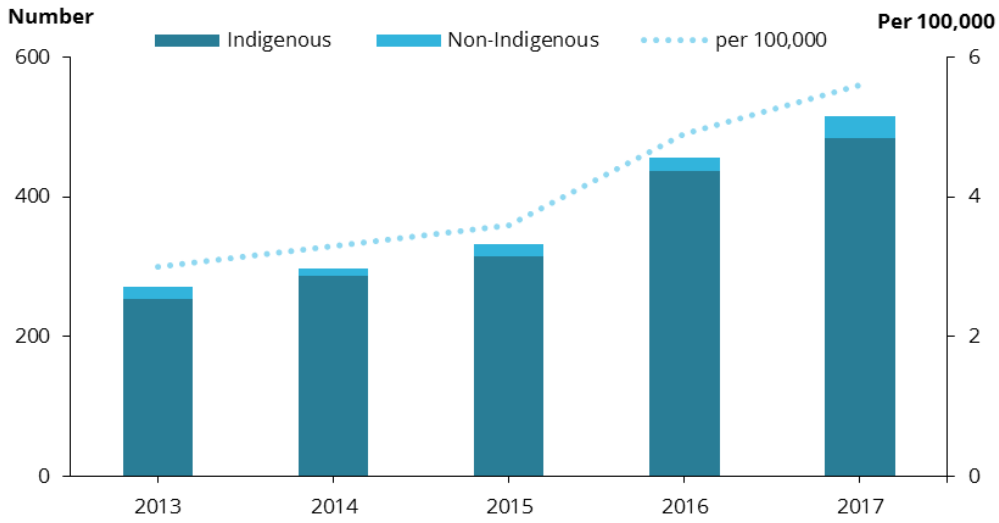
- NT RHD register has been collecting data since 1997. (The NT register incorporates information from a prior collection).
- QLD RHD register commenced in 2009 (but includes ARF data from 1999 when ARF become notifiable).
- WA RHD register commenced in 2009 (though reporting of RHD only became mandatory in 2015).
- SA RHD register commenced in 2012.

All four 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.

ARF among all Australians

There were 1,897 diagnoses of ARF recorded in 2013–2017, a rate of 4 per 100,000 population. Each year from 2013 to 2017, the number and rate of diagnoses increased. In 2017, 528 cases were diagnosed, a rate of 6 per 100,000, increasing from 3 per 100,000 in 2013.

Figure 1: Number and rate of ARF diagnoses among all Australians, 2013–2017



Notes

1. Rates are crude rates per 100,000 population.

2. ARF diagnoses includes all diagnostic categories and first and recurrent episodes. Refer to box 2 and 3 for more information.

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

In 2013–2017, the most common age at diagnosis was 5–14 years, with 964 diagnoses (16 per 100,000 population). The rate of new diagnoses increased over time in all age groups. In 2017, the highest rate was in 5–14 years, with 20 children diagnosed for every 100,000 in the population (248 diagnoses). ARF was more common in females than males, with 56% of diagnoses (1,060) occurring in females. Females had higher rates of ARF each year compared with males.

Ninety-four per cent of ARF diagnoses (1,776) in 2013–2017 were in Aboriginal and/or Torres Strait Islander Australians. The overall rate of ARF diagnoses in Indigenous Australians was 85 per 100,000 population, more than 250 times the rate among non-Indigenous Australians. The annual number and rate of ARF diagnoses among Indigenous Australians increased throughout the period.

Table 1: Number and rate of ARF diagnoses per 100,000 population, by Indigenous status, 2013–2017

Year	Indigenous (number)	Indigenous (rate per 100,000)	Non-Indigenous (number)	Non-Indigenous (rate per 100,000)	Total (number)[(a)]	Total (rate per 100,000) [(a)]
2013	254	63.5	17	0.2	276	3.0
2014	287	70.2	11	0.2	301	3.3
2015	315	75.4	17	0.2	335	3.6
2016	436	102.1	20	0.2	457	4.9
2017	484	110.9	42	0.5	528	5.6
Total	1,776	85.0	107	0.3	1,897	4.1

(a) Totals include 14 individuals with unknown Indigenous status.

Notes

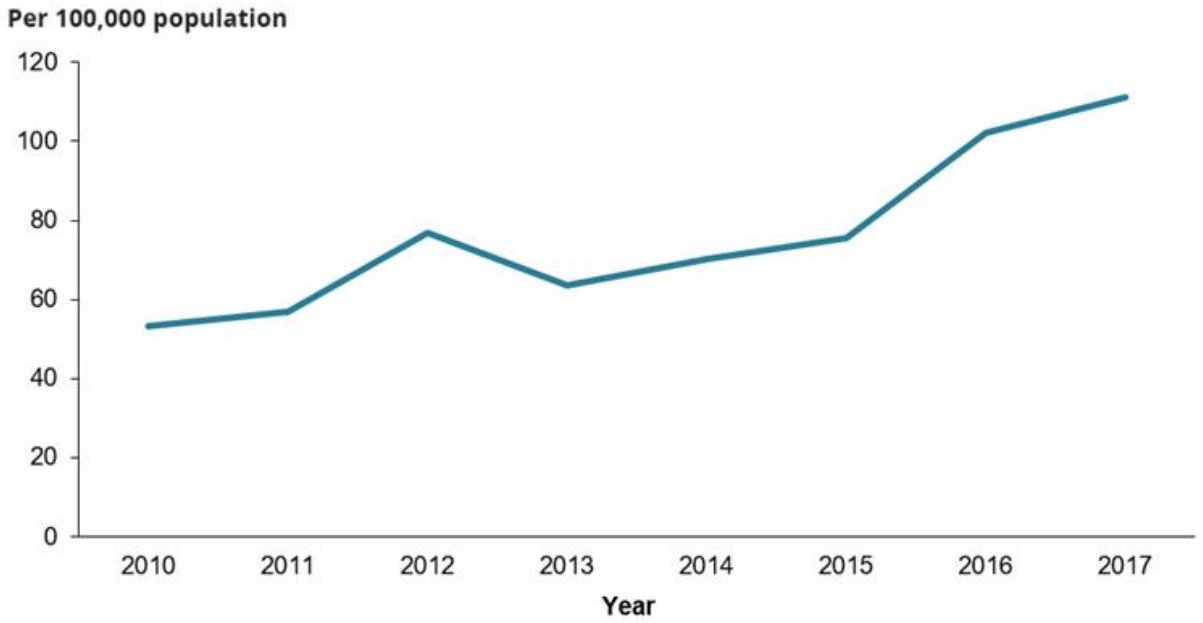
1. ARF diagnoses include all episode types and confirmation statuses.
2. Rates are crude rates per 100,000 population.

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

ARF among Indigenous Australians

The incidence rate for ARF among Indigenous Australians has increased over time from 53 per 100,000 population (200 diagnoses) in 2010 to 111 per 100,000 population (484 diagnoses) in 2017 (Figure 2).

Figure 2: Incidence of ARF among Indigenous Australians, 2010 to 2017



Note: ARF diagnoses includes all diagnostic categories and first and recurrent episodes. Refer to box 2 and 3 for more information.

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

In 2013–2017, the age and sex distribution of ARF diagnoses among Indigenous Australians follows the pattern described above for all Australians. Children aged 5–14 years had the highest rates of ARF diagnosis and constituted one-third of all people diagnosed (602 diagnoses).

More Indigenous females were diagnosed with ARF than Indigenous males, with 57% of reported Indigenous diagnoses being in females (1,006 diagnoses). ARF rates in adults were generally higher among females than males, but in children the rates were higher in males.

Table 2: Number and rate of ARF diagnoses per 100,000 population among Indigenous Australians by sex and age, 2013–2017

Age group (years)	Male (number)	Male (per 100,000)	Female (number)	Female (per 100,000)	Total (number)	Total (per 100,000)
0–4	32	25.5	30	25.1	62	25.3
5–14	478	203.1	424	185.9	602	194.6
15–24	151	70.6	274	134.2	425	101.6
25–44	104	37.6	256	90.9	360	64.5

Age group (years)	Male (number)	Male (per 100,000)	Female (number)	Female (per 100,000)	Total (number)	Total (per 100,000)
45 and over	5	2.6	22	10.3	27	6.7
Total	770	73.8	1,006	96.1	1,776	85.0

Notes

1. ARF diagnoses includes all diagnostic categories and first and recurrent episodes. Refer to box 2 and 3 for more information.
2. Rates are crude rates per 100,000 population.

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

ARF diagnoses by state and territory and region

Numbers and rates of ARF amongst Indigenous Australians during 2013–2017 were consistently highest in the Northern Territory. Fifty-three per cent (954) of all diagnoses were from NT.

The rate generally increased over time in each jurisdiction, apart from in WA, where there was no clear pattern. In the NT in 2017, 268 diagnoses were recorded, more than twice the number recorded in 2013 (127). The number of diagnoses made in QLD almost tripled over the period (from 43 to 124).

Table 3: Rate of ARF diagnoses per 100,000 population among Indigenous Australians, by state and territory, 2013–2017

Year	Qld (per 100,000)	WA (per 100,000)	SA (per 100,000)	NT (per 100,000)	Total (per 100,000)
2013	21.7	80.5	25.7	178.6	63.5
2014	36.0	57.6	37.7	200.7	70.2
2015	32.7	63.7	22.1	241.2	75.4
2016	59.6	59.4	33.7	318.0	102.1
2017	56.8	75.2	40.1	354.1	110.9
Total rate	41.8	67.3	32.0	260.0	85.0

Notes

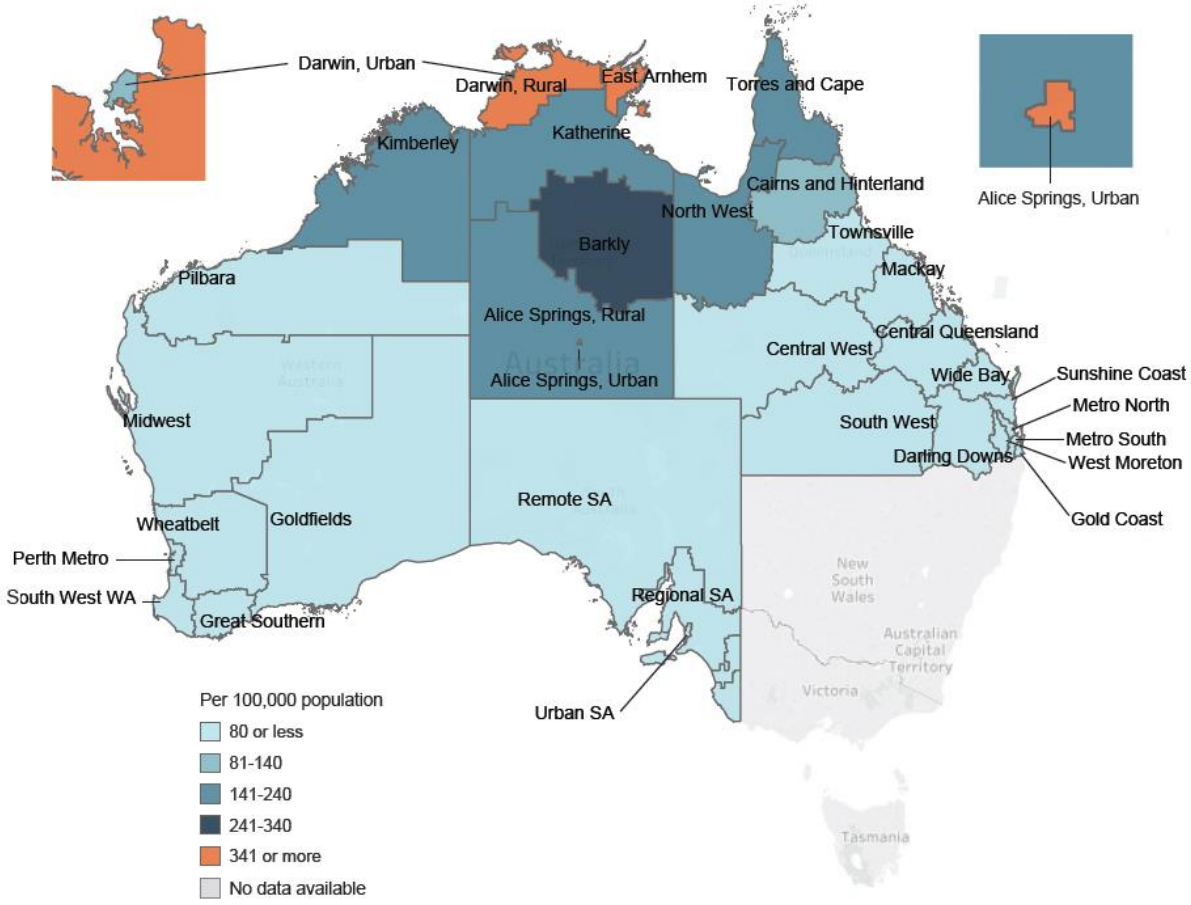
1. ARF diagnoses includes all diagnostic categories and first and recurrent episodes. Refer to box 2 and 3 for more information.
2. Rates are crude rates per 100,000 population.

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

For each ARF diagnosis, the RHD registers record the region of onset (where the patient was diagnosed with ARF) and the region of management (where the patient receives the majority of their primary health care). The place where GAS infection was acquired is not captured.

Regions which were more remote often had higher rates of ARF onset than non-remote areas. In 2013–2017, the region with the highest rate of ARF onset was Rural Darwin (391 per 100,000 population, 252 diagnoses). Urban Alice Springs (371 per 100,000 population or 124 diagnoses) and East Arnhem in the NT (357 per 100,000 population, 207 diagnoses) also had high rates.

Figure 3: Rate of ARF diagnoses per 100,000 population among Indigenous Australians, by region of onset, 2013–2017



Notes:

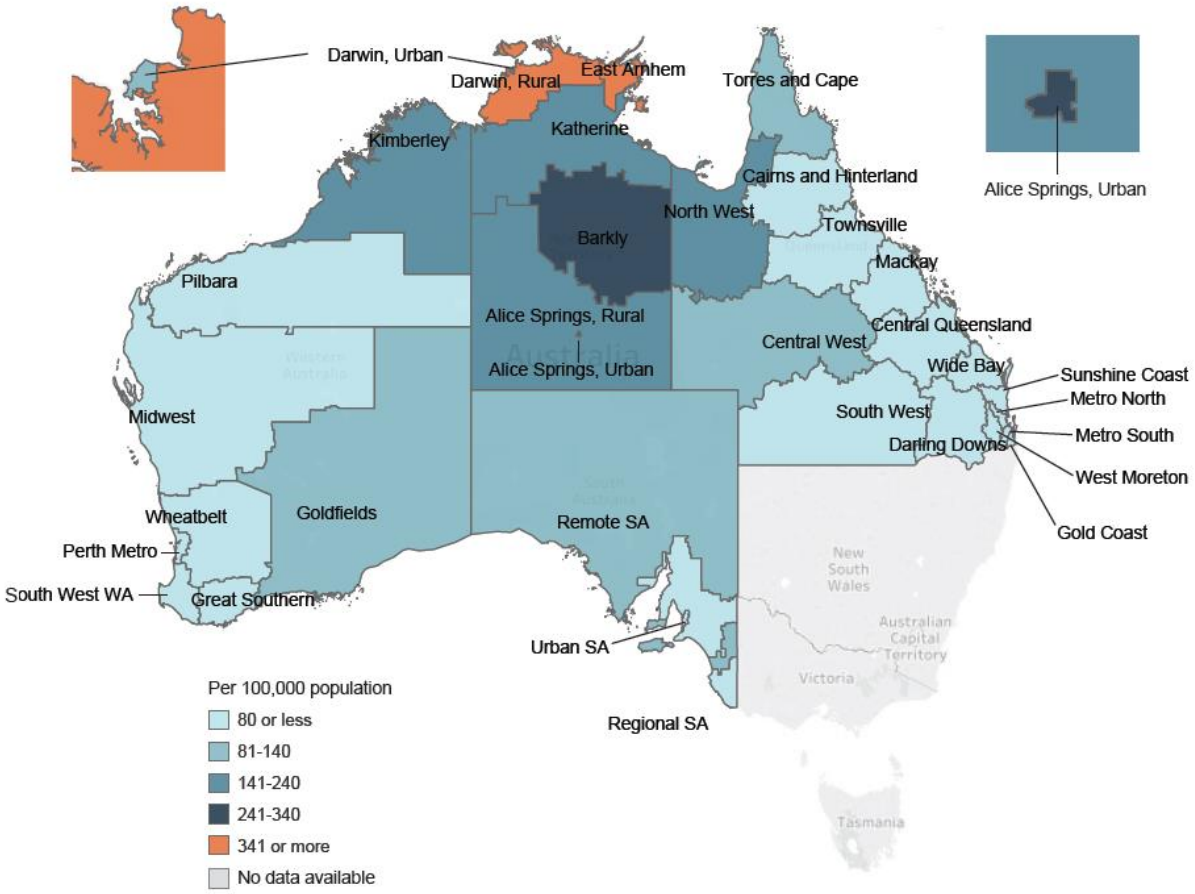
1. There are 33 regions across the 4 states and territories. Each state and territory define regions uniquely, based on their own specific health services boundaries.
2. Rates are crude rates per 100,000 population.
3. ARF diagnoses include all episode types and confirmation statuses. Refer to box 2 and 3 for more information.
4. No data are available for jurisdictions not included in the National Rheumatic Heart Disease data collection.
5. For Queensland regions, the 2016 population estimates were used to calculate rates for 2016 and 2017.
6. Perth Metro North and South in WA have been combined. Torres Strait and Cape York in Qld have been combined.

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

Over the period 2013–2017, Rural Darwin in the NT (387 per 100,000 population; 249 diagnoses) managed the greatest number of Indigenous Australians with recently

diagnosed ARF. (Note that primary health care services manage cases diagnosed prior to 2013 as well.) Rates in East Arnhem in the NT (366 per 100,000; 212 diagnoses) and the Kimberley in WA (180 diagnoses or 200 per 100,000) were also high.

Figure 4: Rate of ARF diagnoses per 100,000 population among Indigenous Australians, by region of management, 2013–2017



Notes

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5. No data are available for jurisdictions not included in the National Rheumatic Heart Disease data collection.
6. Perth Metro North and South in WA have been combined. Torres Strait and Cape York in Qld have been combined.

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

ARF among Indigenous Australians by diagnostic category

Nearly three-quarters of all ARF diagnoses among Indigenous Australians in 2013–2017 (1,284 diagnoses, 72%) were for definite ARF (see Box 2). Probable diagnoses made up 17% (296 diagnoses) and 11% of diagnoses were possible (195 diagnoses).

Between 2013 and 2017, rates for definite or probable ARF increased from 59 per 100,000 population (234 diagnoses) to 106 per 100,000 population (424 diagnoses), in line with overall increases in ARF regardless of diagnostic category. However, the proportion of definite or probable ARF diagnoses decreased from 92% in 2013 to 88% in 2017, with a corresponding increase in possible diagnoses. Older Indigenous Australians were more likely to have a definite or probable ARF diagnosis and the patterns in diagnostic category were similar in males and females.

Box 2: ARF diagnostic categories

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

- **Definite ARF:** 2 major or 1 major and 2 minor manifestations plus evidence of preceding GAS infection. Long–term preventive penicillin should commence.
- **Probable ARF:** clinical presentation falls short by either one major or one minor manifestation, or the absence of streptococcal serology results, but where ARF is the most likely diagnosis. Long-term preventive penicillin should commence.
- **Possible ARF:** Strong clinical suspicion of ARF, but insufficient signs and symptoms for diagnosis of definite or probable ARF. Preventive penicillin should commence, with a clinical review scheduled for 12 months later, to determine if it should continue long-term.

This classification applies to both first known and recurrent episodes.

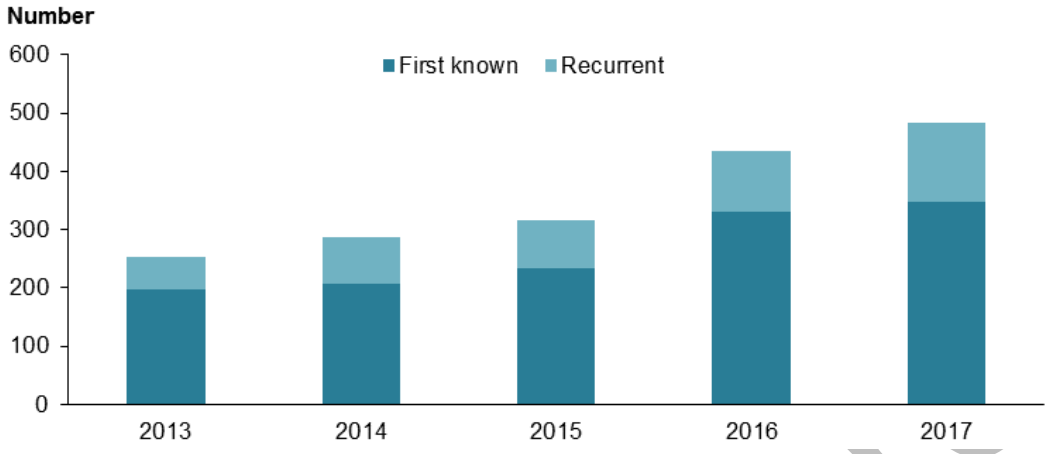
Source: RHD Australia 2012.

ARF diagnoses by recurrence category

Both first known ARF episodes and recurrent episodes (see Box 3) are preventable. After the first known ARF episode, adherence to secondary prophylaxis reduces the likelihood of a recurrence.

In 2013–2017, three-quarters of ARF diagnoses (74%, 1,316 diagnoses) were recorded as the first known diagnosis for that individual while one-quarter of diagnoses were for recurrent disease (26%, 460 diagnoses). The number and rate of first known ARF episodes increased from 49 per 100,000 population in 2013 (197 diagnoses) to 72 per 100,000 population in 2017 (348 diagnoses), in line with an overall increase in diagnoses. The proportion of ARF diagnoses categorised as first known ARF was relatively stable over this period, ranging between 72% and 78% of diagnoses each year.

Figure 5: Number of ARF diagnoses among Indigenous Australians by recurrence category, 2013–2017



Note: ARF diagnoses includes all diagnostic categories and first and recurrent episodes. Refer to box 2 and 3 for more information.

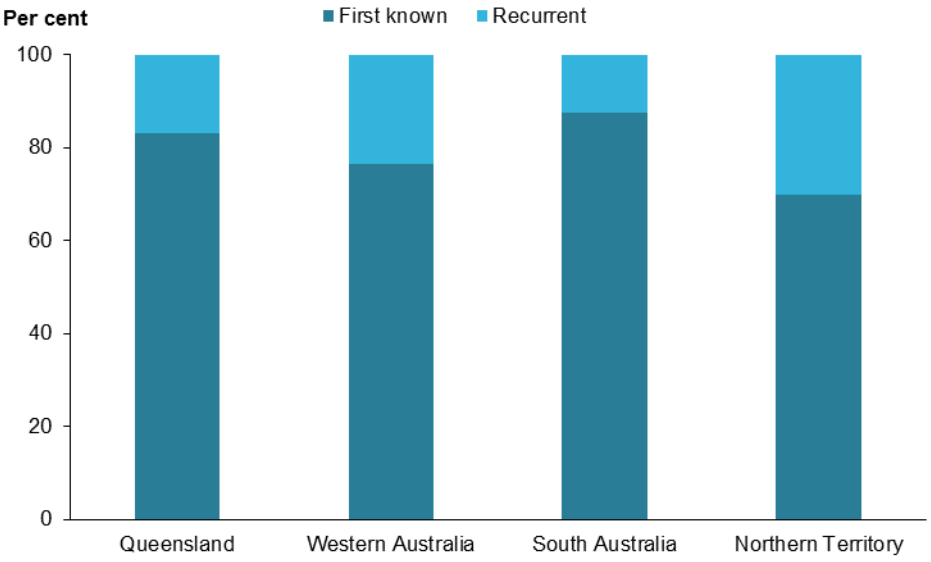
Source: AIHW analysis of National Rheumatic Heart Disease data collection.

Recurrent ARF episodes

Around one-quarter of ARF diagnoses reported were for recurrent disease. Recurrent episodes were more common in females and in older people.

In each state and territory, the majority of ARF diagnoses were categorised as the first known diagnosis. In 2013–2017, the proportion of first known ARF diagnoses in Qld was 82% (356 diagnoses). WA reported 76% of ARF diagnoses as first known (246 diagnoses) overall, though the proportion varied from year to year. In SA, 75% of diagnoses were categorised as first known (49 diagnoses). There was greater annual variation in SA, because of a smaller number of diagnoses. In NT, 70% were first known ARF diagnoses (665 diagnoses), and this was stable over the 5 years.

Figure 6: Proportion of ARF diagnoses among Indigenous Australians by recurrence category and state and territory, 2013–2017



Note: ARF diagnoses includes all diagnostic categories and first and recurrent episodes. Refer to box 2 and 3 for more information.

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

The proportion of episodes categorised as recurrent increased with age. In 2013–2017, 5% of 0–4 year olds had a recurrent diagnosis compared to 48% of those aged over 45 years. Females were more likely to have recurrent episodes, both overall (28% of diagnoses in females compared with 23% of those in males) and in all age groups except 15–24 year olds.

Box 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.
- Recurrence status: A reported ARF episode (definite, probable or possible) in an individual with known past ARF or RHD.

Deaths among all Australians with ARF

Twenty one deaths were recorded on the Registers in 2013–2017 among people who had an ARF diagnosis (these people had ARF only—people with ARF and RHD diagnoses were analysed as RHD deaths). The individuals could have died from any cause, as cause of death is not captured. Most of these deaths were of Indigenous Australians (19 deaths), and 16 deaths occurred in the NT. Among the Indigenous Australians who died, 2 were aged between 15 and 24 years and 6 were aged 25–44 years. All but one of the remainder died before turning 65.

Rheumatic heart disease

The National Rheumatic Heart Disease data collection includes information about diagnoses of rheumatic heart disease (RHD) recorded in each jurisdiction. The commencement year of each register varies, from 1997 in NT, 2009 in Qld and in WA, to 2012 in SA, though reporting of RHD may not have been mandatory in the relevant jurisdiction at the time of commencement. A person can only have one diagnosis of RHD, though they may be registered in more than one jurisdiction as they can receive care in different places. For the purpose of the national data collection, each diagnosis was assigned to only one jurisdiction.

RHD among all Australians

As at 31 December 2017, there were 4,255 (45 per 100,000 population) living persons with RHD recorded on state and territory registers. Eighty–seven per cent were Indigenous Australians (3,687 diagnoses). The NT had the highest rate of prevalent RHD diagnoses (793 per 100,000 population, 1,951 diagnoses), while SA had the lowest (11 per 100,000 population, 181 diagnoses).

Table 1: Prevalence of RHD, by Indigenous status and state and territory, 31 December 2017

State and territory	Indigenous (number)	Indigenous rate (per 100,000)	Non-Indigenous (number)	Non-Indigenous rate (per 100,000)	Total (number) [(a)]	Total rate (per 100,000) [(a)]
Qld	1,160	531.0	331	7.0	1,569	31.8
WA	522	523.6	32	1.3	554	21.5
SA	161	379.7	20	1.2	181	10.5
NT	1,844	2,436.2	107	62.8	1,951	792.8
Total	3,687	845.2	490	5.4	4,255	44.0

(a) Totals include 78 diagnoses with unknown Indigenous status.

Notes

1. For NT, RHD diagnoses have been recorded in a Register since 1997. Registers began in 2009 in Qld, in 2009 in WA and in 2012 in SA.
2. Rates are crude rates per 100,000 population.

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

There were almost twice as many females living with RHD (2,783 diagnoses, 58 per 100,000 population) than males (1,470 diagnoses, 31 per 100,000 population) as at 31 December 2017.

Table 2: Prevalence of RHD, by age and sex, 31 December 2017

Age group (years)	Male (number)	Male (rate per 100,000)	Female (number)	Female (rate per 100,000)	Total (number)	Total (rate per 100,000)
5–14	167	26.8	220	37.1	387	31.8
15–24	424	67.3	517	85.8	941	76.4
25–34	309	44.7	610	88.1	919	66.4
35–44	227	36.4	529	84.4	756	60.5
45 and over	343	18.7	907	46.5	1,250	33.0
Total	1,470	31.2	2,783	58.4	4,255	44.9

Notes

1. The grand total number includes 2 diagnoses with age not stated.
2. Rates are crude rates per 100,000 population.

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

The majority of people had mild RHD (42%, 1,766 diagnoses), followed by severe RHD (31%, 1,312 diagnoses).

New RHD diagnoses in all Australians

In this report, a 'new' RHD diagnosis is defined as one that was diagnosed between 1 January 2013 and 31 December 2017, and these will be the focus of the analysis in the next sections. In most cases, it is not possible to identify a year of onset for RHD as the condition may be asymptomatic initially. The analysis is based on year of diagnosis.

There were 1,261 reports of new RHD diagnoses in 2013–2017 (3 per 100,000 population). Australian annual diagnosis rates remained relatively steady at around 3 diagnoses per 100,000 population each year. The state and territory specific rates for 2013–2017 were 2 per 100,000 population for Queensland, 2 per 100,000 population for WA, 1 per 100,000 population for SA and 37 per 100,000 population for NT.

In 2013–2017, just over 1,000 new RHD diagnoses (83% of all diagnoses) were made among Indigenous Australians. The annual rate remained steady over time, ranging from 48 to 52 per 100,000 population. Over the 5 years, the overall rate in Indigenous Australians was around 125 times that in non-Indigenous Australians.

Table 2: Number and rate per 100,000 population of new RHD diagnoses by Indigenous status, 2013–2017

Year	Indigenous (number)	Indigenous (rate per 100,000)	Non-Indigenous (number)	Non-Indigenous (rate per 100,000)
2013	202	50.5	38	0.4
2014	214	52.3	46	0.5
2015	199	47.6	34	0.4
2016	204	47.8	46	0.5
2017	224	51.3	34	0.4
Total	1,043	49.9	198	0.4

Note: Rates are crude rates per 100,000 population

Source: AIHW analysis of National Rheumatic Heart Disease data collection

New RHD diagnoses among Indigenous Australians

The NT had the highest rate and greatest number of new RHD diagnoses among Indigenous Australians each year, with overall rates around 3 times that in the other jurisdictions.

Table 3: Rate of new RHD diagnoses per 100,000 population among Indigenous Australians by jurisdiction, 2013–2017

Year	Qld	WA	SA	NT
2013	35.8	46.8	35.9	104.1
2014	37.9	29.9	40.2	128.7
2015	32.7	29.3	34.4	121.3
2016	34.2	48.1	31.3	95.2

Year	Qld	WA	SA	NT
2017	30.7	42.1	23.6	138.7
Total rate	34.2	39.3	32.9	117.7
Total number	356	188	67	432

Note: Rates are crude rates per 100,000 population

Source: AIHW analysis of National Rheumatic Heart Disease data collection

Age and sex of new RHD diagnoses

The rate of new RHD for Indigenous females was around two times the rate for Indigenous males. Females had higher rates compared to males in all age groups apart from those aged 0–4.

Nearly 60% of cases diagnosed in 2013–2017 were less than 25 years old at diagnosis. There were 15 children aged less than 5 when diagnosed with RHD.

Table 4: Number and rate per 100,000 population of new RHD diagnoses among Indigenous Australians by sex and age, 2013–2017

Age group (years)	Male (number)	Male (rate per 100,000)	Female (number)	Female (rate per 100,000)	Total (number)	Total (rate per 100,000)
0–4	8	6.4	7	5.9	15	6.1
5–14	164	69.7	195	85.5	359	77.5
15–24	71	33.2	167	81.8	238	56.9
25–44	79	28.5	218	77.4	297	53.2
45 +	36	18.8	98	45.9	134	33.1
Total	358	34.3	685	65.4	1,043	49.9

Note: Rates are crude rates per 100,000 population

Source: AIHW analysis of National Rheumatic Heart Disease data collection

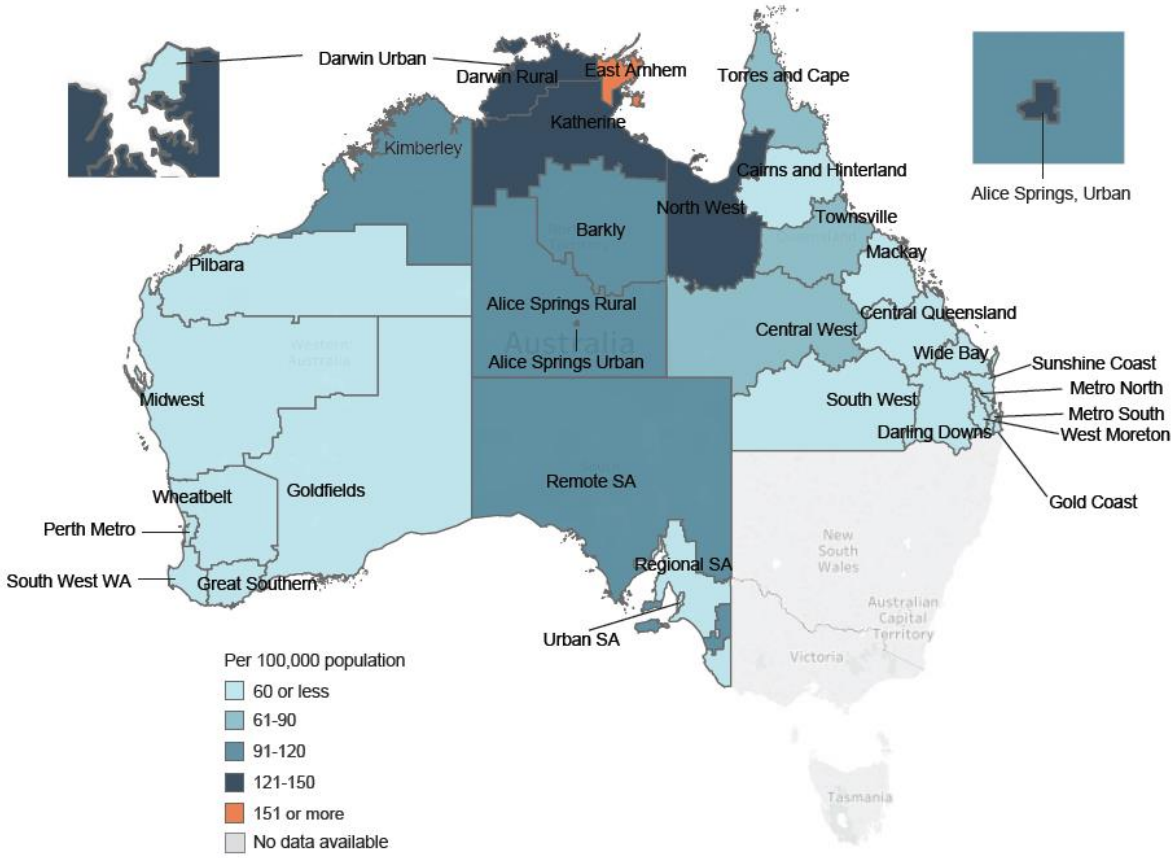
Region of management of new RHD diagnoses

For each RHD diagnosis reported to a register, the region of management (where the patient receives the majority of the primary health care for their RHD) is recorded. The region of management may be different to the region of diagnosis.

For new RHD diagnoses in Indigenous Australians between 2013 and 2017, the region with the highest rate of RHD management was East Arnhem in the NT (179 per 100,000 population, 104 diagnoses), followed by Northwest, Qld (145 per 100,000 population, 64 diagnoses). The rates of new RHD management in Urban Alice Springs (46 diagnoses), Rural Darwin (87 diagnoses) and Katherine (73 diagnoses) in the NT were all between 130 and 140 diagnoses per 100,000 population.

Each region also manages RHD diagnoses which were diagnosed prior to 2013, so the total number of cases being managed within each region may be greater than shown here.

Figure 1: Rate of new RHD diagnoses among Indigenous Australians per 100,000 by region of management, 2013–2017



Notes:

1. There are 33 regions across the 4 states and territories. Each state and territory define regions uniquely, based on their own specific health services boundaries.
2. Rates are crude rates per 100,000 population.
3. No data are available for jurisdictions not included in the National Rheumatic Heart Disease data collection.
4. For Queensland regions, the 2016 population estimates were used to calculate rates for 2016 and 2017.
5. Perth Metro North and South in WA have been combined. Torres Strait and Cape York in Qld have been combined.

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

Prior ARF episodes among Indigenous Australians with RHD

RHD cannot occur without a preceding ARF episode. However, some people diagnosed with RHD have not had a previous ARF episode recorded. While it is possible that some of these individuals will have had an appropriately managed ARF episode which was not captured in a relevant register, it is likely many had undiagnosed ARF with no

preventative treatment. There are a range of reasons why ARF might not have been diagnosed or notified to the Register. Possible explanations include — mild or subclinical ARF not causing symptoms severe enough to present to a health service, challenges in diagnosing ARF and challenges in upskilling the workforce in a setting of high turnover of primary health care staff.

Among all Indigenous Australians with a new RHD diagnosis between 2013 and 2017, 4 in 5 (838 diagnoses) did not have a prior ARF episode recorded on the registers. This proportion varied between 71% and 85% each year, but did not vary by sex. There was some variation between states and territories, with 91% of newly registered RHD diagnoses in Qld, 85% in SA, 77% in NT and 67% in WA not having a previous ARF episode documented. In Queensland, the proportion of RHD diagnoses with no previous ARF recorded is overestimated due to the timing of notification legislation.

Children (0–14 years) and older Indigenous Australians were more likely to have had no prior ARF at the time of RHD diagnosis compared to those aged 15–34 years.

Table 5: Number and proportion of new RHD diagnoses among Indigenous Australians where no prior ARF was recorded, by age, 2013–2017

Age group (years)	Number of new RHD diagnoses	Number with no prior ARF recorded	Per cent with no prior ARF recorded
0–4	15	15	100.0
5–14	363	298	82.1
15–24	235	176	74.9
25–34	182	132	72.5
35-44	115	99	86.1
45 +	133	118	88.7
Total	1,043	838	80.4

Source: AIHW analysis of National Rheumatic Heart Disease data collection

Severity classification (priority level) among Indigenous Australians with new RHD diagnoses

Patients on the RHD Registers are categorised as having mild, moderate, or severe RHD (Priority level 3, 2, and 1, respectively), as outlined in the Australian RHD guidelines (Box 4). In this report, severity status and priority level are used interchangeably.

Severity classification is based on clinical symptoms. Classification also determines the frequency of clinical assessment and echocardiography. Patients with ARF on prophylaxis (but no RHD diagnosis) are routinely assigned a priority level of mild.

Box 4: Severity classification (priority level)

Priority 1 (Severe): Severe damage to the heart valves or moderate to severe heart valve damage with symptoms of heart failure

Priority 2 (Moderate): Any moderate heart valve damage without symptoms, and with normal left ventricle function

Priority 3 (Mild): Mild heart valve disease

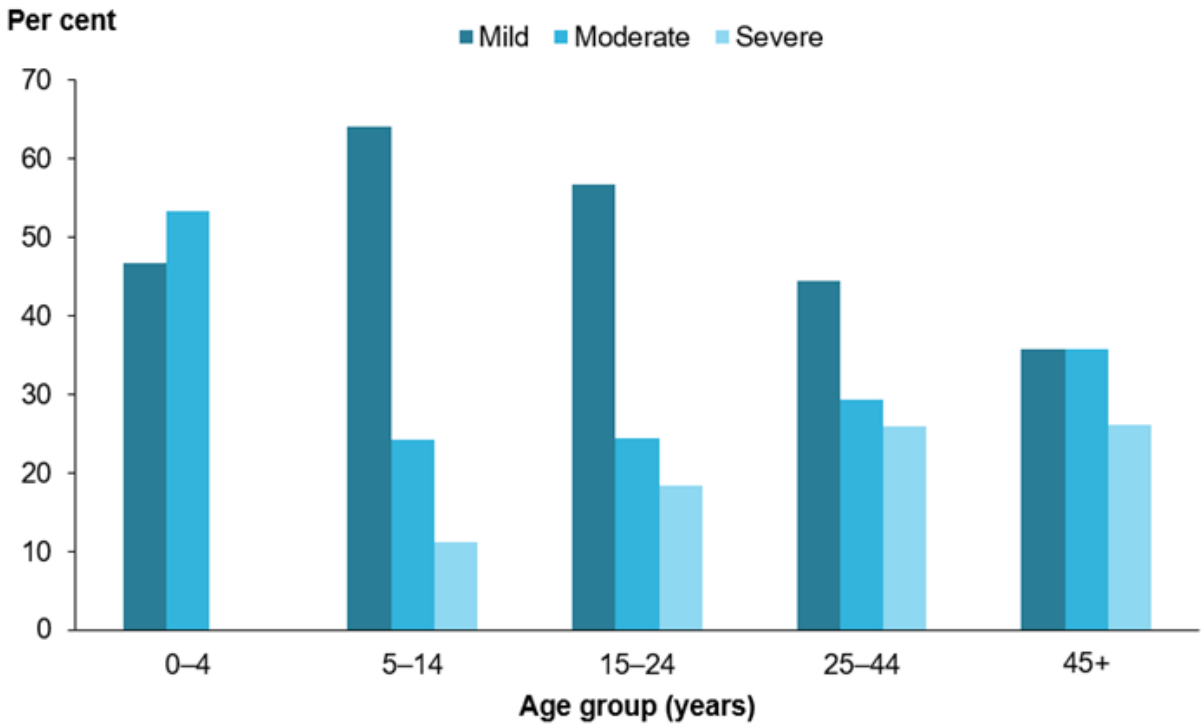
Priority 4 (inactive): Patients with a history of ARF (no RHD) for whom secondary prophylaxis has been ceased

Source: RHD Australia 2017

Severity at diagnosis

In 2013–2017, of the 1,043 Indigenous Australians with new RHD, 53% (552 diagnoses) had mild RHD when first diagnosed, while 28% (288 diagnoses) had moderate RHD and 19% (196 diagnoses) had severe disease. A very small proportion of cases (7 diagnoses) did not have RHD status recorded at diagnosis. This distribution was similar across states and territories.

Figure 2: Severity status at diagnosis among Indigenous Australians newly diagnosed with RHD, by age at 31 December 2017, 2013–2017



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

The majority of the 196 diagnoses with severe RHD at diagnosis were Indigenous females (65%, 128 diagnoses) and in those aged 25–44 years (39%, 77 diagnoses). Those who were diagnosed as adults were more likely than those diagnosed as children to have severe RHD at first diagnosis.

RHD surgery

RHD leads to damage to the heart's valves—the mitral, aortic, pulmonary and tricuspid valves. The damaged valves may need surgery so they can be replaced or repaired. The main options for surgical management of RHD are:

- Balloon valvotomy, which can be used to treat mitral stenosis. This operation can be performed by threading a deflated balloon on wires up to the heart from a cut in the groin. The narrowed mitral valve is opened by gently inflating a balloon inside the valve. The small incision reduces costs and complications compared with open surgical repair.
- Valve repair involves repairing the heart valve shape and function to allow for normal blood flow. It usually involves open heart surgery. Repair generally offers the

best outcomes for children and adults, particularly for mitral valve (RHD Australia et al. 2012, Wang et al. 2013).

- Valve replacement involves removing the damaged valve and replacing it with a mechanical prosthetic (metallic valve) or bioprosthetic valve (tissue valve from animal or human donors). This generally involves open heart surgery.

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.

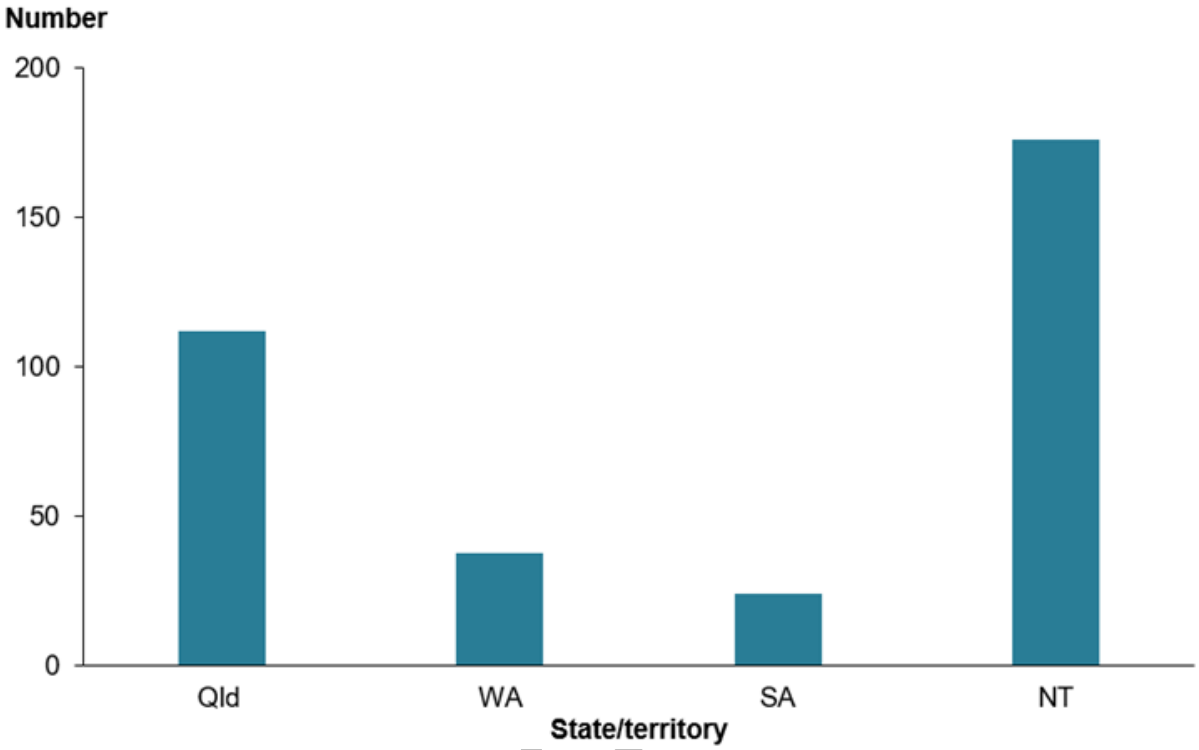
Surgery among all people with RHD

Since the commencement of record keeping by the registers, 1,725 RHD related surgeries have been recorded for 1,277 individuals. More than three-quarters (78%, 1,340) of these surgeries occurred in Indigenous Australians. The following sections focus on surgeries performed on Indigenous Australians with RHD.

Surgery in Indigenous Australians with RHD

In 2013–2017, 322 Indigenous Australians with RHD underwent 350 surgery events. For analysis purposes, a surgical event was included regardless of the year of RHD diagnosis, acknowledging that jurisdictions have been collecting data for different lengths of time (Box 1). The majority of people had one surgery while 23 people had up to 4 surgeries. Around half of these surgeries were in cases from the NT (176 surgical events), and one-third (112) from Qld.

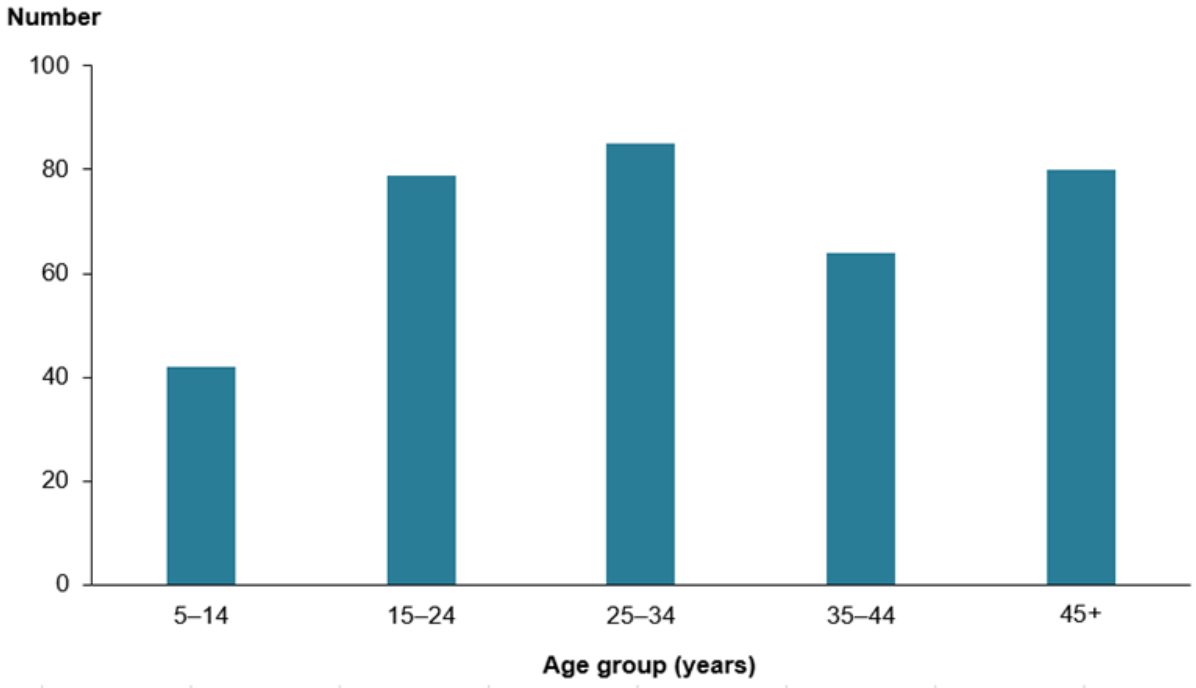
Figure 3: Number of surgical events among Indigenous Australians with RHD, by state and territory, 2013–2017



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

Just over three quarters of all surgical events among Indigenous Australians with RHD occurred in people aged less than 45 years (270 surgical events), of which 42 were in children aged 5–14 (12%). Only 23% of surgical events were for those aged 45 and over (80 surgical events). Two-thirds of all surgical events (234) were in females. Across age categories, between 64% and 70% of surgery events occurred in females. The sex distribution of surgical events is similar to the distribution of people living with RHD.

Figure 4: Number of surgical events among Indigenous Australians with RHD, by age group, 2013–2017



Notes

1. There were no people who received surgery for RHD when aged 0–4 during 2013–2017.
2. This shows the age at surgery of the 350 surgical events 2013–2017. Individuals who had more than one surgery are included multiple times as an individual could have more than one surgery in the period.

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

In 2013–2017, during the 350 surgical events amongst Indigenous RHD cases, there were 477 individual procedures performed. There were 89 people who had 2 different surgical procedures, 18 who had 3 and 9 who had between 4 and 6 procedures. Multiple procedures could occur in one surgery, or over multiple surgeries.

Nearly 300 valve replacement procedures (62%) occurred. Among these, replacement with a mechanical valve was more common than with a bioprosthetic valve. A quarter of procedures involved the aortic valve (122 procedures).

Table 6: Proportion of surgical procedures by surgery type and valve among Indigenous Australians, 2013–2017

Procedure type	Mitral valve	Aortic valve	Tricuspid valve	Total per cent	Total number
Repair, including open valvuloplasty	26.9	11.5	73.2	28.5	136
Replacement—mechanical valve	37.9	40.2	7.1	34.8	166
Replacement—bioprosthetic valve	24.2	42.6	12.5	27.5	131
Percutaneous balloon valvuloplasty	4.0	0.8	1.8	2.9	14
Other or unknown type[(a)]	7.0	4.9	5.4	6.3	30
Total per cent	62.5	25.6	11.7	100.0	477
Total number	298	122	56	. .	477

(a) Includes 28 not stated or unknown surgery types, 1 surgery of the pulmonary valve and 2 Ross procedure surgeries.

Note: The NT data groups valvuloplasties with repairs.

Source: AIHW analysis of National Rheumatic Heart Disease data collection

Surgery for RHD

322 Indigenous Australians underwent surgery for RHD in 2013–2017. Three-hundred and fifty surgical events were recorded, comprising 477 individual procedures. Surgery was most common in people less than 45 years and in females.

References

RHD Australia, (ARF/RHD writing group), National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand 2012. The Australian guideline for the prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edn). Northern Territory: RHD Australia, Menzies School of Health Research.

Wang Z, Zhou C, Gu H, Zheng Z & Hu S 2013. Mitral valve repair versus replacement in patients with rheumatic heart disease. J Heart Valve Dis 22(3):333–9.

Deaths among all Australians with RHD

In 2013–2017, there were 275 deaths reported among people with RHD in the registers. Of these, 221 (80.3%) were in Indigenous Australians. These people could have been diagnosed at any time since RHD Registers have been operational, but had died between 2013 and 2017. This analysis was not restricted by cause of death—people with RHD may have died of any cause. Cause of death is not captured on Registers.

The majority of deaths (67%) occurred in NT, which has the greatest number of RHD diagnoses recorded.

Table 7: Number and rate of deaths among Indigenous Australians with RHD by year of death and state and territory, 2013–2017

Year	Queensland	Western Australia	South Australia	Northern Territory
2013	n.p.	5	n.p.	40
2014	n.p.	n.p.	5	20
2015	11	n.p.	n.p.	29
2016	7	8	—	29
2017	11	n.p.	n.p.	30
Total number	37	24	12	148
Total rate per 100,000 population	3.6	0.3	5.9	40.3

Note: Rates are crude rates per 100,000 population.

Source: AIHW analysis of National Rheumatic Heart Disease data collection

Nearly 50% of the Indigenous RHD cases who died in 2013–2017 were 45–64 years old (108 deaths), and just over one-quarter (61 deaths) were in cases 25–44 years old. Thirteen deaths (5.9%) were in young adults aged 15–24 years. The median age of death was 50 years. In line with overall rates of RHD, around two-thirds of deaths (145 deaths) were in female RHD cases.

Secondary prophylaxis

Secondary prophylaxis with regular benzathine penicillin G (BPG) is the only RHD control strategy shown to be both clinically and cost effective at community and individual levels (RHD Australia 2012). The recommended regimen to prevent recurrences of ARF and progression of RHD involves regular intramuscular injections of BPG every 21 to 28 days, for a minimum of 10 years. This treatment aims to provide penicillin in the blood over a period of 3 to 4 weeks, providing protection against GAS infections (Wyber 2013). As the penicillin concentration wanes, the individual's susceptibility to subsequent GAS infection and to recurrent ARF will increase.

Delivery of BPG every 28 days is challenging for health services, affected individuals and their families. In remote Indigenous communities, a major factor contributing to low levels of prophylaxis delivery is the availability and acceptability of health services. Personal factors include such as injection refusal, pain caused by injections. Lack of knowledge and understanding of ARF and RHD may negatively influence adherence to secondary prophylaxis but are often not the major contributing factors (Parnaby & Carapetis 2010).

References

Parnaby M & Carapetis J 2010. Rheumatic fever in Indigenous Australian children. Journal of Paediatrics and Child Health 46:527–33.

RHD Australia (ARF/RHD writing group), National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand 2012. The Australian guideline for the prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edn). Northern Territory: RHD Australia, Menzies School of Health Research.

Wyber R, Taubert K, Marko S & Kaplan EL 2013. Benzathine penicillin G for the management of RHD, concerns about quality and access, and opportunities for intervention and improvement. Global Heart 8(3):227–234.

Secondary prophylaxis in Indigenous Australians

This section focuses on BPG adherence amongst Indigenous Australians who were prescribed BPG on a 28-day regimen before 31 January 2017, who did not cease BPG before 31 December 2017, and who received at least one dose in 2017. If a person had more than one diagnosis (for example, of ARF and of RHD), they were included in the analysis only once. Those on a different regimen or an alternative treatment (not BPG) were excluded.

In total 3,372 people received any prophylaxis in 2017. Qld had 1,041 people on preventive penicillin in 2017 and there were 582 in WA, 118 in SA and 1,631 NT. Of these, 2,630 people were eligible for inclusion in further adherence calculations. They were located in NT (1,240), QLD (871), WA (438) and SA (81). Because they were on a 28 day

schedule for the entire year, this group were able to receive at least 13 doses of BPG. Of these:

- 15% (394 people) received 100% or more of their prescribed doses
- 21% (548 people) received 80% to 99% of their prescribed doses
- 37% (964 people) received 50% to 79% of their prescribed doses
- 28% (724 people) received less than 50% of their prescribed doses.

For all states and territories, the most common level of adherence was receiving 50% to 79% of prescribed doses.

Adherence of 100% is the gold standard for all people on prophylactic treatment. Some people may receive more than 13 doses due to their particular schedule. The proportion of Indigenous Australians receiving 13 or more doses of BPG in one year was 23% in NT (280 people), 33% SA (27 people), 7% in Qld (62 people) and 6% in WA (25 people).

At a population level, the proportion of people receiving at least 80% of their scheduled doses is used as an indicator of adherence which is likely to protect against ARF recurrences. In 2017, 36% of Indigenous Australians (942 people) received at least 80% of doses. The proportion of people achieving at least 80% adherence was greatest in SA (63%, 51 people), the NT (46%, 573 people), followed by 27% in WA (118 people) and 23% in Qld (200 people).

Younger Indigenous Australians (aged 15–24 years) had generally lower adherence than other age groups, with almost one-third of people of this age receiving less than half of their prescribed doses. Almost half (46%) of those aged 5–14 and more than half (52%) of those aged 45 and over received at least 80% of their prescribed doses.

Table 1: Proportion of Indigenous Australians on BPG by adherence level and age, 2017

Adherence level	0–4 years	5–14 years	15–24 years	25–44 years	45 years and over	Total %
1%–49% adherence	—	16.3	32.5	31.7	14.0	27.5
50%–79% adherence	—	37.3	38.5	34.8	34.0	36.7
80%–99% adherence	33.3	27.9	17.9	19.5	20.0	20.8
100%+ adherence	66.7	18.5	11.1	14.0	32.0	15.0

Adherence level	0–4 years	5–14 years	15–24 years	25–44 years	45 years and over	Total %
Total %	100.0	100.0	100.0	100.0	100.0	100.0

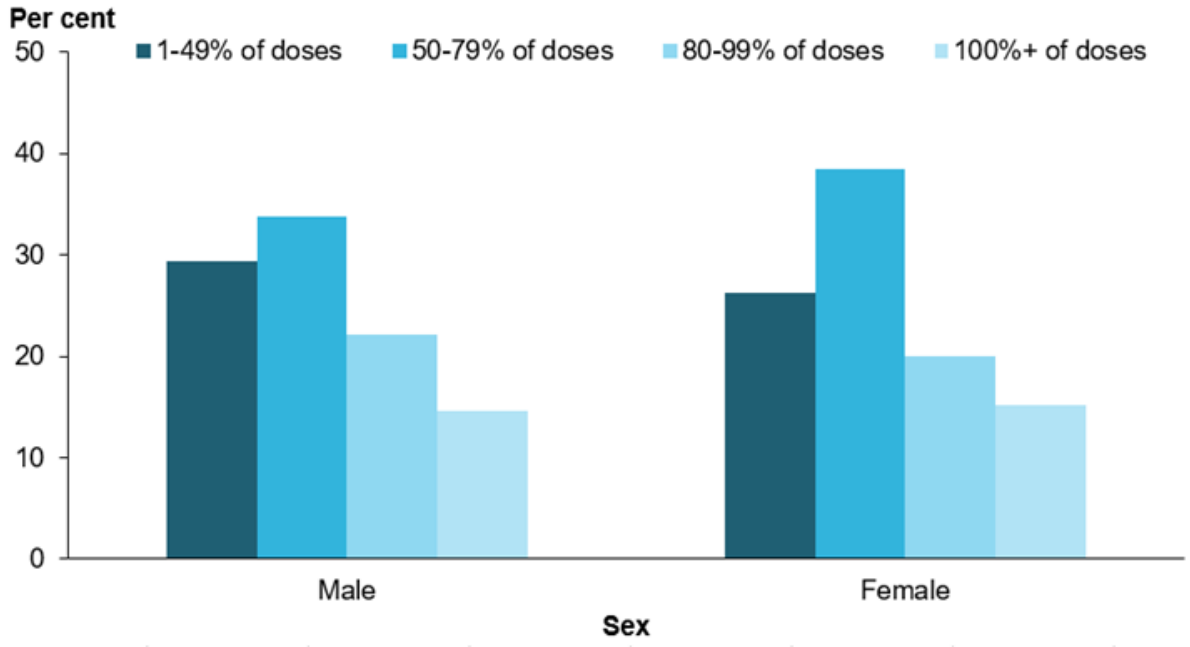
Notes

1. People on BPG can have more than 13 doses in one year, therefore 100% of doses is defined as 100%+ of doses.
2. This analysis only includes people who were prescribed prophylaxis for the whole of 2017.

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

The adherence pattern was similar for males and females on prophylaxis in 2017. Thirty-seven per cent of males (382 people) and 35% of females (561 people) achieved at least 80% adherence. Around one quarter of males (29%) and females (26%) received 50% or less of their prescribed doses.

Figure 1: Proportion of Indigenous Australians on BPG by adherence level and sex, 2017



Notes

1. People on BPG can have more than 13 doses in one year, therefore 100% of doses is defined as 100%+ of doses.
2. This analysis only includes people who were prescribed prophylaxis for the whole of 2017.

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

ARF recurrences per 100 patient years among all Australians

ARF recurrences per 100 patient years

The measure of ARF recurrences per 100 patient years is used to account for the different amounts of time people who have had an ARF episode are at risk of having a recurrent episode. It enables comparison of the total number of days after the first ARF episode a person is at risk of recurrence. This analysis only includes people who are on preventative treatment.

In 2017, among more than 3,600 people who were prescribed BPG, there were 132 reported ARF recurrences. For every 100 patient years at risk, there were 3.9 ARF recurrences. The recurrence rate per 100 patient years was slightly higher among females than males, and slightly higher among Indigenous than non-Indigenous Australians.

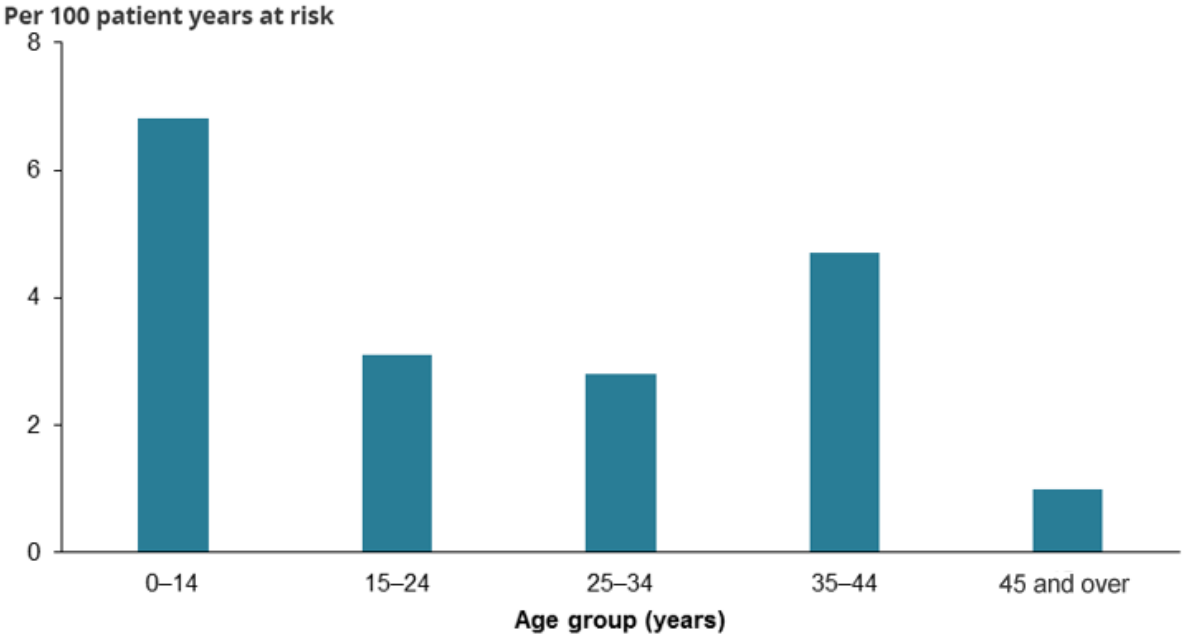
Table 4: Number and rate of ARF recurrences per 100 patient years for people prescribed BPG, by sex and by Indigenous status, 2017

	Number of recurrences	Recurrences per 100 patient years
Sex		
Male	52	3.7
Female	80	4.0
Indigenous status		
Indigenous	126	3.9
Non-Indigenous	6	3.6
Total	132	3.9

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

The rate of recurrence generally decreased with age. For children, the risk of recurrence per 100 patient years in 2017 was 6.8, compared with 0.9 in those aged 45 and over.

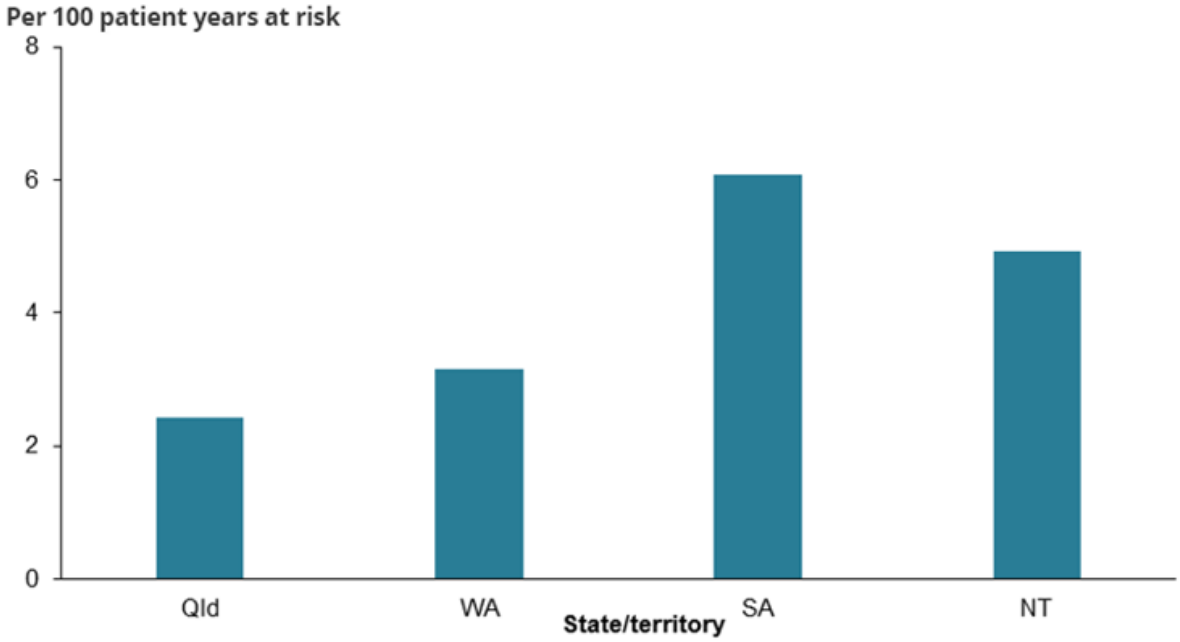
Figure 7: ARF recurrences by per 100 patient years and age group, for people prescribed BPG, 31 December 2017



Note: Age is age at 31 December 2017.
Source: AIHW analysis of National Rheumatic Heart Disease data collection.

There were 27 recurrences in QLD, 18 in WA, 7 in SA and 80 in the NT in 2017. The rate per 100 patient years of ARF recurrence was highest in SA and lowest in QLD.

Figure 8: ARF recurrences per 100 patient years, by state and territory, 2017



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

References

RHD Australia, (ARF/RHD writing group), National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand, 2012. The Australian guideline for the prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edn). Northern Territory: RHD Australia, Menzies School of Health Research.

Wyber R, Taubert K, Marko S and Kaplan EL. 2013. Benzathine penicillin G for the management of RHD, concerns about quality and access, and opportunities for intervention and improvement. Global Heart 8(3):227–234.

RHD control program and register in NSW

Data analysed for this report were provided directly to AIHW from the New South Wales (NSW) Rheumatic Heart Disease (RHD) Register. This register is funded by NSW Health. ARF and RHD became notifiable in NSW in October 2015, and the Register was established in May 2016, and captures patients notified with ARF and RHD where the individual provides consent to be incorporated into the Register. Whilst RHD is only notifiable in persons under the age of 35 years, people older than 35 years may be included on the Register if is it is felt worthwhile by their health practitioner.

Due to the short duration of the NSW Register's existence, data on ARF and RHD notifications have been provided for 1 October 2015 to 31 December 2017. Data for secondary prophylaxis was provided for 2017. Data have not been analysed by year of diagnosis. Rates have been averaged for the period 1 October 2015 to 31 December 2017. Data for 2015 were annualised. The rates for NSW are not comparable to the data provided by Qld, WA, SA and the NT, in the previous sections.

ARF in NSW

Between October 2015 and December 2017, there were 43 reported ARF diagnoses in NSW. Nineteen (44%) ARF diagnoses were reported among Indigenous residents, 18 of which were classified as definite or probable episodes. Fourteen (33%) ARF diagnoses were in Pacific Islander people and 1 diagnosis in a person who was both Indigenous and from another high risk group. ARF rates were greater in females than males, and in the 5–14 year olds compared to other age groups. Less than 5 episodes were reported as recurrent ARF.

RHD in NSW

There are 44 known residents of NSW living with RHD and who were diagnosed at less than 35 years of age, as at December 2017. Of these, 13 (30%) identified as Indigenous Australians, 19 (43%) identified as Pacific Islanders, and 1 person identified as both Indigenous and from another high risk group. Among the Indigenous Australians, 10 were female and 8 were aged 5–14 years.

Secondary prophylaxis in NSW

In NSW, secondary prophylaxis adherence has been calculated for all patients on the NSW Register who were prescribed BPG during 2017. Adherence was calculated as a proportion of the scheduled 13 doses 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 2017 are also included in the data.

There were 26 people in NSW prescribed secondary prophylaxis during 2017. Of these, 9 received 100% or more of their prescribed doses and a further 3 received 80% to 99% of their prescribed doses. Fifteen people on treatment were Indigenous Australians.

Technical information

ARF and RHD key performance indicators

Key performance indicator	Data included in report
1. Epidemiology	
1.1. ARF incidence, by confirmation status (Definite/Probable/Possible), age group and	
1.1.1. Ethnicity (count and %)	Ethnicity not included due to incomplete or inconsistent data capture
1.1.2. Sex	Included
1.1.3. Indigenous status	Included
1.1.4. Jurisdiction and Indigenous status	Included
1.2. ARF recurrences, by	
1.2.1. Ethnicity and age group (count and %)	Ethnicity not included due to incomplete or inconsistent data capture
1.2.2. Sex and age group (rate)	Included, separately
1.2.3. Indigenous status and age group (rate)	Age group (count and rate)
1.2.4. Jurisdiction, Indigenous status and age group (rate)	Jurisdiction (count and rate)
1.2.5. Proportion of all ARF episodes by jurisdiction	Included
1.2.6. Rate per 100 patient-years for patients prescribed prophylaxis (both oral and BPG) by jurisdiction	Included
1.3. ARF and/or RHD deaths, by	
1.3.1. Ethnicity (count and %)	Ethnicity not included due to incomplete or inconsistent data capture
1.3.2. Sex and age group (rate)	Sex and age group (count, proportion) separately
1.3.3. Indigenous status and age group (rate)	Not available
1.3.4. Jurisdiction and age group (rate)	Jurisdiction only
1.3.5. Direct or indirect attribution (count and %)	Not available
1.4. RHD point prevalence, by age group and	
1.4.1. Ethnicity (count and %)	Ethnicity not included due to incomplete or inconsistent data capture
1.4.2. Sex (rate)	count, proportion (not by age group)
1.4.3. Indigenous status (rate)	count, proportion (not by age group)
1.4.4. Jurisdiction and Indigenous status (rate)	count, proportion (not by age group)
1.4.5. Severity classification (rate)	count, proportion
1.5. RHD incidence, by	
1.5.1. Ethnicity and age group (count and %)	Ethnicity not included due to incomplete or inconsistent data capture
1.5.2. Sex and age group (rate)	Included, separately
1.5.3. Indigenous status and age group (rate)	Indigenous status only
1.5.4. Jurisdiction, Indigenous status and age group (rate)	Jurisdiction only

1.5.5. Severity classification and age group (count and %)	Included
1.5.6. Severity classification and detection method (count and %)	Detection method not available due to poor data quality
1.5.7. Previous notification of ARF (definite or probable/possible) and age group (count and %)	Not included due to poor data quality
1.5.8. Severity classification if no previous notification of ARF and age group (count and %)	Included, not by age group.
2. Requirement and uptake of secondary prophylaxis	
2.1. BPG secondary prophylaxis administration, by adherence category and	
2.1.1. Ethnicity and age group (count and mean %)	Ethnicity not included due to incomplete or inconsistent data capture
2.1.2. Sex and age group (count and mean %)	Included, separately
2.1.3. Indigenous status and age group (count and mean %)	Not available
2.1.4. Jurisdiction and age group (count and mean %)	Jurisdiction only
2.1.5. Median percentage of all scheduled doses	Not available
3. Quality of management	
3.1. Priority levels of ARF/RHD cases, by age group and	
3.1.1. Ethnicity (count and %)	See measures for RHD severity
3.1.2. Sex (count and %)	
3.1.3. Indigenous status (count and %)	
3.1.4. Jurisdiction and Indigenous status (count and %)	
3.2. Echocardiograms performed within designated timeframes for priority level 1 and 2 RHD cases	
3.2.1. Priority level 1 within the previous: 6 months, 7-12 months, >12 months, No echo recorded (count and %)	Not included due to incomplete or inconsistent data capture
3.2.2. Priority level 2 with the previous: 12 months, 13-24 months, >24 months, No echo recorded (count and %)	
3.3. Waiting times of RHD cases accepted for cardiac surgery	
3.3.1. Wait times: < 6 months, 7-12 months, 13-24 months, >24 months, N/A (count and %)	Not included due to incomplete or inconsistent data capture
3.4. Surgical procedures performed, by type and	
3.4.1. Ethnicity (count and %)	Ethnicity not included due to incomplete or inconsistent data capture
3.4.2. Sex (count and %)	Included, not by type
3.4.3. Age group (count and %)	Included, not by type
3.4.4. Indigenous status (count and %)	Included, not by type
3.4.5. Jurisdiction (count and %)	Included, not by type
3.5. Deaths within 30 days and 1 year post rheumatic cardiac surgery, by	
3.5.1. Ethnicity (count and %)	Ethnicity not included due to incomplete or inconsistent data capture
3.5.2. Sex (count and %)	Not available
3.5.3. Age group (count and %)	Not available
3.5.4. Indigenous status (count and %)	Not available
3.5.5. Jurisdiction (count and %)	Not available

Additional measures:
RHD incidence by region of management
RHD incidence by region of onset
ARF incidence by region of management
ARF incidence by region of onset

Data elements

National RHD Data Collection Data elements

Data element name	Definition
Person with or at risk of RHD	
Record linkage key	A unique code that enables two or more records belonging to the same individual to be brought together.
Date of birth	The date of birth of the case.
Sex	The biological distinction between male and female, as represented by a code.
Indigenous status	Whether a case identifies as being of Aboriginal or Torres Strait Islander origin.
Non-Indigenous target group	A case's membership of specific ethnic or regional origin groups that are identified as high risk groups in guidelines for priority screening and treatment for acute rheumatic fever and rheumatic heart disease in Australian primary health care settings.
Date of death	The date of death of the case.
Jurisdiction	The state or territory which first diagnosed/notified the episode.
Region of management	The location of health services in a defined geographic or administrative area where the case is being managed.
ARF diagnosis and notification	
Notification date	The date that a health professional notified an incident case of a disease to the relevant jurisdictional authority.
Region at ARF diagnosis	The location of health services in a defined geographic or administrative area where the case was diagnosed.
Recurrence status	Whether an episode of ARF for a given case is the first episode or a recurrence.
Confirmation status	The confirmation status of an incident case of ARF.
Detection method	How an incident case of a disease notified to the jurisdictional registers.
RHD diagnosis	
RHD diagnosis date	The date on which a case was diagnosed with rheumatic heart disease.
Prophylaxis prescription	
Prophylaxis prescribed date	The date on which a case is prescribed prophylactic treatment.
Prophylaxis prescription date	The date on which a health professional recommends that prophylactic antibiotics are no longer required.
Antibiotic agent	The antibiotic agent prescribed to a case for secondary prevention of acute rheumatic fever.
Frequency of IM antibiotic treatment	The prescribed frequency of intramuscular antibiotic treatment to be administered to a case.
Antibiotic administration	

Antibiotic administration date	The date of administration of intramuscular antibiotic to a case for secondary prevention of acute rheumatic fever.
RHD surgery	
RHD surgical procedure date	The date of surgery for repair or replacement of heart valve(s) in treatment of rheumatic heart disease.
Heart valve procedure date	
Heart valve identifier	The particular heart valve repaired or replaced in surgery for treatment of rheumatic heart disease.
RHD surgical procedure date	The date of surgery for repair or replacement of heart valve(s) in treatment of rheumatic heart disease.
RHD status	
RHD status assessment date	The date on which a clinical assessment is made of the status of a case's rheumatic heart disease.
RHD priority status	The priority status of rheumatic heart disease in a person as determined on the RHD status assessment date.

Glossary

Aboriginal and/or Torres Strait Islander: people who identified themselves, or were identified by another household member, as being of Aboriginal and/or Torres Strait Islander origin. 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 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.

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

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

health hardware: the physical equipment necessary for health, 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.

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

Indigenous: used interchangeably with Aboriginal and/or Torres Strait Islander in this report.

Non-Indigenous Australians: includes people who did not identify as being of Aboriginal and/or Torres Strait Islander origin, and people for whom information on their Indigenous status was not available. Compare with non-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.

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 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.

Abbreviations & symbols

ABS	Australian Bureau of Statistics
AIHW	Australian Institute of Health and Welfare
ARF	acute rheumatic fever
BPG	Benzathine penicillin G
GAS	Group A streptococcal infection
NT	Northern Territory
Qld	Queensland
RHD	rheumatic heart disease
RFDS	Royal Flying Doctor Service
SA	South Australia
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

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 2017, ARF was notifiable to state health departments in five Australian jurisdictions (Qld, WA, SA, NT and New South Wales), while RHD was notifiable in three (NSW, WA and SA). In NSW, RHD cases are only notifiable in people aged less than 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.

In Queensland, Western Australia, South Australia and the Northern Territory the ARF/RHD control programs are funded by the Australian Government Department of Health. A state-funded ARF/RHD register has commenced in New South Wales (NSW) in 2015, but data are not available for this report. Data about ARF and RHD diagnoses are not collected by jurisdictional health departments in the Australian Capital Territory, Victoria or Tasmania.

The current NT RHD register has been collecting data since 1997. The SA RHD register commenced in 2012, the QLD RHD register commenced in 2009, as did the WA register. The Qld Register incorporates information from 1999 onwards, when ARF became notifiable. For NT, the 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 SA, RHD cases are recorded on the Register are aged less than 50, 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, 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 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 are not currently collected in any Register. These data would assist in monitoring ARF and RHD epidemiology and program evaluation.

Appendix A

Table 1: The 2012 Australian guidelines for the diagnosis of ARF

	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 GAS infection[b]	2 major or 1 major and 2 minor manifestations plus evidence of a preceding GAS infection[b]
Definite recurrent episode of ARF in a patient with known past ARF or RHD	2 major or 1 major and 1 minor or 3 minor manifestations plus evidence of a preceding GAS infection[b]	2 major or 1 major and 1 minor or 3 minor manifestations plus evidence of a preceding GAS infection[b]
Probable ARF (first episode or recurrence)	A clinical presentation that falls short by either one major or one minor manifestation, or the absence of streptococcal serology results, but one in which ARF is considered the most likely diagnosis. Such 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 one major or one minor manifestation, or the absence of streptococcal serology results, but one in which ARF is considered the most likely diagnosis. Such 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) Polyarthritis[c] Chorea[d] Erythema marginatum[e] Subcutaneous nodules

Minor manifestations	Monoarthralgia Fever[f] ESR[h] ≥30mm/hor CRP(h) ≥30 mg/L Prolonged P-R interval on ECG[g]	Fever[f] Polyarthralgia or aseptic mono-arthritis ESR(h) ≥30 mm/h or CRP(h) ≥30 mg/L Prolonged P-R interval on ECG[g][h]

CRP = C-reactive protein

ECG = electrocardiogram

ESR = erythrocyte sedimentation rate

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

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

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

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

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

(f) Oral, tympanic or rectal temperature ≥38°C on admission, or a reliably reported fever documented during the current illness.

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

Source: RHD Australia 2012.