Australian Government



Australian Institute of Health and Welfare





# Acute rheumatic fever and rheumatic heart disease in Australia, 2014–2018

## 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. New South Wales register data are summarised also. The third version of the national guidelines for ARF and RHD management were released in February 2020. The 2nd version of the guidelines (released 2012) are relevant to the data in this report.

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 2018, the 4 jurisdictional registers recorded 6,802 people with a past ARF and/or an RHD diagnosis
- 2. Most ARF diagnoses in 2014–2018 (95%, 1,963 diagnoses) were among Indigenous Australians
- Among Indigenous Australians, 5–14 years was the most common age for first known ARF episode (195 per 100,000)
- 4. In 2014–2018, NT had the highest rate of ARF (291 per 100,000 population) among Indigenous Australians
- 5. In 2014–2018, 1,314 new RHD diagnoses were made among Indigenous Australians
- 6. Nearly 60% of Indigenous Australians diagnosed with RHD are under 25 when diagnosed
- 7. In 2014–2018, the rate of new RHD diagnoses in Indigenous Australians was greatest in the NT (141 per 100,000 population)

8. In 2018, 42% of Indigenous Australians prescribed preventive penicillin received 80% or more of their prescribed doses

## Topic/s

### **Primary topic**

Aboriginal and Torres Strait Islander people

### **Additional topics**

Rural and remote Australians; Indigenous Australians; Heart, stroke & vascular diseases

## 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 (Strep A) 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 can 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 can become scarred and stiffer, obstructing blood flow (stenosis), or it can fail to close properly, causing blood to flow backwards in the heart (regurgitation) instead of forward around the body. Regurgitation due to damage to the mitral valve is common in RHD.

This is the second 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, and should not be compared with those in previously published reports.

#### Jurisdictional RHD control programs and registers

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

- improve clinical care, including improved delivery of and adherence to secondary prophylaxis antibiotics
- provide education and training for health care providers, individuals, families and communities
- collect and provide agreed data annually to the AIHW for national monitoring and reporting of ARF and RHD and measuring program effectiveness in the detection and management of ARF and RHD
- maintain a dedicated state-wide patient register and recall system for ARF and RHD.

### How many people have ARF?

In 2014–2018, a total of 2,076 diagnoses of ARF were recorded in Queensland, Western Australia, South Australia and the Northern Territory, a rate of 4.4 per 100,000 population over the 5 years combined. During this period the number and rate of diagnoses increased each year, with 478 individuals diagnosed in 2018—a rate of 5.0 per 100,000 (compared to 321, or 3.5 per 100,000 in 2014).

During the same period, 1,963 ARF diagnoses were recorded among Indigenous Australians—a rate of 89 per 100,000 population over the 5 years combined. The number and rate of diagnoses increased from 300 (71 per 100,000) in 2014 to 457 (100 per 100,000) in 2018. ARF was more common among Indigenous females than males, and rates were highest among Indigenous people aged 5–14 (964 diagnoses, 195 per 100,000).

#### How many people have RHD?

As at 31 December 2018, there were 4,993 people living with RHD recorded on the 4 jurisdictional registers. Of these, nearly 9 in 10 diagnoses (4,325) were among Indigenous Australians, 1 in 3 (1,634) were aged under 25, nearly 2 in 3 (3,232) were females, and the greatest number were living in the NT (2,076).

In 2014–2018, 1,314 new RHD diagnoses were made among Indigenous Australians, a rate of 60 per 100,000 population. For this group, new RHD diagnoses were more common among Indigenous females compared to males (76 and 44 diagnoses per 100,000, respectively), and nearly 3 in 5 new diagnoses were among people aged under 25 years (773 diagnoses). The greatest number and highest rate of new diagnoses among Indigenous Australians was in the Northern Territory (525, or 141 per 100,000).

### How many people died with RHD?

In 2014–2018, 315 deaths were reported for people living with RHD (cause of death is not recorded in the registers). Of these, 248 people (79%) were Indigenous Australians, 135 (43%) were aged 45–64 years, and 88 (28%) were aged 15–44 years. The median age at death was 56 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). Between 2014 and 2018, the recommended regimen to prevent recurrences of ARF, and progression of RHD, involved regular intramuscular injections of BPG every 21–28 days, for a minimum of 10 years.

In 2018, among Indigenous Australians prescribed 4-weekly BPG:

- 23% (748 people) received 100% or more of their prescribed doses
- 19% (613) received 80% to 99% of their prescribed doses
- 32% (1,012) received 50% to 79% of their prescribed doses
- 26% (836) received less than 50% of their prescribed doses.

In 2018, among more than 3,900 Australians prescribed BPG, there were 111 recurrent ARF episodes. The majority of these recurrences are most likely a result of delayed doses of BPG.

## Introduction

#### What is acute rheumatic fever?

Acute rheumatic fever (ARF) refers to an autoimmune response to infection of the throat and possibly of the skin by group A streptococcus (Strep A) bacteria (McDonald et al. 2004). 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 (innermost layer of skin) and can be extremely painful (Parnaby & Carapetis 2010). While there is no lasting damage caused to the brain, joints and skin, ARF may cause lasting damage to the heart. Hospitalisation is required so all necessary investigations are undertaken and to rule out other diagnoses. There is no single diagnostic test for ARF.

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.

Refer to Acute rheumatic fever for more information.

### 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 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: Diagram of the heart, emphasising the heart 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), 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, secondary prophylaxis with penicillin, 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).

Refer to Rheumatic heart disease for more information.

### Are ARF and RHD preventable diseases?

ARF and rheumatic heart disease (RHD) are both preventable diseases. They are common in low- and middle-income countries, and in socioeconomically disadvantaged populations in high-income countries (Wyber 2014, Webb 2015). Acute rheumatic fever (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). Improved living conditions and access to functional health hardware can reduce high rates of Group A streptococcal (Strep A) infections and progression to ARF (Katzenellenbogen et al. 2017). Prevention measures that improve living conditions and environmental health and address other determinants of ARF are known as primordial prevention measures.

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

Secondary prevention of the progression from ARF to RHD relies on correct diagnosis of ARF, to enable commencement of regular antibiotic preventive medication. Correct diagnosis is challenging as there is no specific single laboratory test for ARF, and it can be misdiagnosed. Diagnosis is based on clinical criteria outlined in the Australian modification of the Jones criteria (for more information on the Australian modification refer to Australian guidelines for diagnosis of ARF), 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 2020).

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

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### Jurisdictional RHD control programs and registers

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

These programs are funded to:

- improve clinical care, including improved delivery of and adherence to secondary prophylaxis antibiotics
- provide education and training for health care providers, individuals, families and communities
- collect and provide agreed data annually to the AIHW for national monitoring and reporting of ARF and RHD and measuring program effectiveness in the detection and management of ARF and RHD
- maintain a dedicated state-wide patient register and recall system for ARF and RHD.

Information from the ARF/RHD registers in these four jurisdictions is compiled by the Australian Institute of Health and Welfare (AIHW) to provide information about ARF and RHD in Australia. The <u>Australian ARF/RHD guideline</u> 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, delivery of secondary prevention and use of surgical interventions.

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 NSW register was established in 2016, with ARF and RHD becoming notifiable in NSW in 2015, and RHD being notifiable only in persons aged less than 35 years. Data from NSW are presented in detail in a standalone section.

#### Reference

RHD Australia, (ARF/RHD writing group). The 2020 Australian guideline for the prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3rd edn). Northern Territory: RHD Australia, Menzies School of Health Research.

### How many people have ARF and/or RHD?

At 31 December 2018, data from jurisdictional registers in Queensland, Western Australia, South Australia and the Northern Territory show, 6,802 living people were recorded on one or more of these jurisdictional registers. Almost 2 in 5 of these people had diagnoses of both ARF and RHD recorded (38%, 2,561 persons), around 1 in 3 had only RHD recorded (35%, 2,354), and the remainder had only ARF recorded (28%, 1,887).

Of the persons recorded on these 4 jurisdictional registers:

- 9 in 10 (89%) were Indigenous Australians
- 6 in 10 (61%) were female
- 4 in 10 (40%) were aged 5–14 when first diagnosed.



ARF and RHD were made notifiable in NSW in 2015, with RHD being notifiable only in persons aged less than 35 years. A register was established in NSW in 2016. As at 31 December 2018, 120 persons with ARF and/or RHD had been notified in NSW. Due to differences such as timeframes in operation and case ascertainment, these data are not currently comparable with those from the other jurisdictions. Data for NSW are presented in a standalone section of the report.

#### References

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## Acute rheumatic fever

This section discusses diagnoses of acute rheumatic fever (ARF) reported by Australian rheumatic heart disease (RHD) control programs between 2014 and 2018. The total number of ARF diagnoses recorded is dependent 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 is greater than the number of people affected. Cases are allocated to a jurisdiction and region based on where they were diagnosed with ARF.

It is likely that ARF diagnoses are under-reported to RHD registers in all jurisdictions. Although the 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 is likely to have 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 means some individuals are not captured in this analysis. It is difficult to determine whether increases in the number of diagnoses reflect a real increase in the number of cases occurring, improved detection and diagnosis of cases, or increases in the number of people being recorded on the registers.

# Box 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 summarises the timeline of program and register establishment across the jurisdictions.

Refer to Jurisdictional RHD control programs and registers for more information.

30	NSW	QId	WA	SA	NT	Vic, Tas, ACT
RHD control program	2015	2009	2009	2010	1997 <sup>(a)</sup>	×
ARF/RHD register	2016	2006	2009	2012	1997	×
Definite ARF notifiable	2015	1999	2007	2016	1996	×
Probable ARF notifiable	2015	×	2015	2016	2019	×
Possible ARF notifiable	×	×	2015	2016	×	×
Confirmed RHD notifiable	2015 <sup>(b)</sup>	2018	2015	2016	2019	×

#### Table 1: Timeline of program and register establishment

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

(b) In NSW, RHD is only notifiable in persons aged less than 35 years.

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

### **Overview of ARF diagnoses**

In 2014–2018, there were 2,076 acute rheumatic fever (ARF) diagnoses recorded in Queensland, South Australia, Western Australia and Northern Territory combined (incidence rate 4.4 per 100,000 population over the 5 years combined). The number and rate of ARF diagnoses increased from 321 (3.5 per 100,000) in 2014 to 481 (5.1 per 100,000) in 2017, before stabilising. In 2018, there were 478 Australians diagnosed with ARF (5.0 per 100,000).

Among Aboriginal and Torres Strait Islander people in 2014–2018, there were 1,963 ARF diagnoses. The ARF diagnosis rate among Indigenous Australians also increased between 2014 and 2017—from 71.0 to 100.4 per 100,000 population (300 to 457 diagnoses, respectively)—but then remained stable at 100.1 per 100,000 (457 diagnoses) in 2018.



ARF diagnoses among all Australians, by state and territory and year, 2014 to 2018

Note: Data presented is for all ages.

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

## Age and sex

Of the 2,076 ARF diagnoses among all Australians:

- the most common age at diagnosis was 5–14 years, with 1,022 diagnoses (17 per 100,000 population).
- the age-specific diagnosis rate was highest in 2016 among those aged 5– 14, with 229 diagnoses (19 per 100,000).

• females accounted for more than half (1,129) of all diagnoses.

Of the 1,963 ARF diagnoses among **Indigenous Australians**:

- the highest rate of diagnosis was among those aged 5–14, accounting for half of all diagnoses (195 per 100,000 population, or 964 diagnoses)
- females accounted for 55% of diagnoses (1,085 diagnoses)
- in people aged under 15, ARF rates were generally higher among males than females
- for adults, ARF rates were generally higher among females than males.

The remaining information on ARF in this report (with the exceptions of deaths and recurrences per 100 patient-years) relates to Indigenous Australians only, due to the small number of cases occurring among non-Indigenous Australians.

#### **Region of ARF diagnosis**

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

In 2014–2018, the Northern Territory accounted for 55% of the 1,963 acute rheumatic fever (ARF) diagnoses among Aboriginal and Torres Strait Islander people (1,085 diagnoses). The rate of ARF diagnoses among Indigenous Australians also increased considerably in the Northern Territory between 2014 and 2018—from 198 to 353 per 100,000 population, respectively.

Although the rate of ARF diagnoses in 2018 was greater than that in 2014 across Queensland, South Australia and Western Australia, the increase was not consistent.

In 2014–2018, the highest rates of ARF diagnoses were reported in the Northern Territory:

- Rural Darwin with 477 per 100,000 population (300 diagnoses)
- Urban Alice Springs with 424 per 100,000 (142)
- East Arnhem with 403 per 100,000 (229).

ARF diagnoses among Indigenous Australians, by region of diagnoses, 2014–2018



## **ARF** symptoms and diagnosis

Diagnosing acute rheumatic fever (ARF) can be challenging, as there is no single diagnostic laboratory test available, with diagnosis based on clinical decisions plus supporting laboratory evidence. Diagnosis relies on identification of major and minor manifestations of the illness, as outlined by Australian guidelines (RHD Australia 2012).

#### Manifestations

Manifestations are important to assist in the diagnosis of ARF. Data on specific manifestations that can occur with ARF are reported to the National Collection by registers. This report presents information on those manifestations for which reliable data could be provided by all jurisdictions: carditis, chorea, and prolonged P-R interval. People with carditis and/or a prolonged P-R interval are more likely to sustain heart damage (and hence to develop RHD) than those without.

Global data suggest that between 30%–82% of people with their first episode of ARF experience carditis, and around 20% of children with ARF have a prolonged P-R interval (Caldas et al. 2008, RHD Australia 2020, Karacan 2010). Chorea reportedly occurs in 5%–36% of people with ARF worldwide, primarily in children and females (WHO 2014). Australian data suggest that between a quarter and one-half of Indigenous ARF patients with chorea go on to develop RHD (RHD Australia 2020).

Refer to Box 2 for more information on manifestations.

In 2014–2018, of the 1,963 ARF diagnoses among Aboriginal and Torres Strait Islander people:

- carditis was present in 19%
- prolonged P-R interval was present in 23%
- Sydenham's chorea was present in 8%
- 1 in 3 (36%) had either carditis, prolonged P-R interval, or both.



ARF symptoms at diagnosis among Indigenous Australians, by state and territory, 2014–2018

#### **Box 2: Examples of ARF manifestations**

**Arthritis:** a swollen and hot joint with pain on movement. Can involve one joint (monoarthritis) or several joints (polyarthritis), which may be affected at the same time or one after the other. The large joints such as the knees and ankles are most commonly affected. Arthritis is the most common presenting symptom of ARF, and the pain is generally greater than would be expected based on the clinical signs.

**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:** detected through electrocardiography (ECG). Refers to when the time between specific electrical features of a heartbeat is longer than expected. Often the person has no symptoms.

**Sydenham Chorea:** 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 Australian guidelines for diagnosis of ARF.

Source: RHD Australia 2020.

#### **Diagnostic categories**

In 2014–2018, of all 1,963 ARF diagnoses among Indigenous Australians:

• 1,670 diagnoses were definite or probable diagnoses

• 251 were possible diagnoses.

Refer to Box 3 for more information on ARF diagnostic categories.

Between 2014 and 2018, the rate of definite or probable ARF diagnoses increased from 65 to 80 per 100,000 population (273 to 366 diagnoses). This was in line with overall increases in ARF, regardless of diagnostic category. However, the proportion of definite or probable ARF diagnoses decreased, from 91% to 80% from 2014 to 2018—with a corresponding increase in the proportion of possible diagnoses.

#### Box 3: ARF diagnostic categories

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

**Definite ARF**, **first episode:** 2 major or 1 major and 2 minor manifestations plus evidence of preceding Strep A infection. Long-term preventive penicillin should commence.

**Definite ARF, recurrent episode:** 2 major or 1 major and 1 minor manifestations or 3 minor manifestations plus evidence of preceding Strep A 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.

Note that these definitions applied when the data in this report were collected, and have been updated in the most recent clinical guidelines (RHD Australia 2020).

Source: RHD Australia 2012.

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#### **ARF** recurrences

First known and recurrent acute rheumatic fever (ARF) episodes are preventable (Box 4). After the first known ARF episode, adherence to secondary prophylaxis reduces the likelihood of recurrence.

Among Aboriginal and Torres Strait Islander people in 2014–2018, 512 ARF episodes were recorded as recurrent disease (26%). First known ARF episodes increased from 222 diagnoses in 2014 to 338 in 2018—53 to 74 per 100,000 population, respectively. This is in line with an overall increase in diagnoses.



Note: Note: Data from NT, Qld, SA and WA Indigenous patients, prescribed BPG Source: AIHW analysis of National Rheumatic Heart Disease data collection.

### Box 4: ARF episode category 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.

Of the 512 recurrent episodes diagnosed in 2014–2018:

- 62% were in females. Females were more likely to have recurrent episodes, with 29% (316) of all episodes in females being recurrent, compared to 22% (196) in males
- among males, the greatest number of recurrences was reported in those aged 5–14
- among females, the greatest number of recurrences was reported in those aged 25–44.

#### **ARF** recurrence rates

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 were prescribed secondary prophylaxis in 2018.

In 2018:

- among more than 3,911 people who were prescribed BPG, there were 111 reported ARF recurrences
- for every 100 patient-years at risk, there were 3.1 ARF recurrences
- most recurrences were in the NT (72 recurrences)
- the rate of ARF recurrences per 100 patient-years was highest in the Northern Territory (4.3) and lowest in Queensland (1.7)
- the rate of recurrence per 100 patient-years generally decreased with age, with the highest risk among those aged 0–14 (4.9).



ARF recurrences per 100 patient years, among Indigenous Australians, by state and territory, 2018

Note: Data from NT, Qld, SA and WA Indigenous patients, prescribed BPG. Source: AIHW analysis of National Rheumatic Heart Disease data collection.

#### Deaths among all Australians with ARF

In 2014–2018, there were 21 deaths recorded among people with a diagnosis of ARF only (people with ARF and RHD diagnoses were analysed as RHD deaths). These individuals could have died from any cause, as cause of death is not captured on registers. Among Aboriginal and Torres Strait Islander people 19 deaths were recorded, with 15 deaths occurring in the Northern Territory. Three of those who died were aged 15–24 years.

## **Rheumatic heart disease**

The National Rheumatic Heart Disease Data Collection includes information about diagnoses of rheumatic heart disease (RHD) recorded in Queensland, Western Australia, South Australia, Northern Territory and New South Wales. The total number of RHD diagnoses recorded is dependent 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).

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, based on their location for primary health care at the time the data were submitted.

As at 31 December 2018, there were 4,993 (52.1 per 100,000 population) people living with RHD recorded on registers recorded in Queensland, Western Australia, South Australia and the Northern Territory. Of these:

- 87% were Indigenous Australians (4,325 diagnoses, 948 per 100,000 population)
- 33% were aged under 25 (1,634 diagnoses)—with 5 aged under 5
- 65% were female (3,232 diagnoses)
- Northern Territory had the highest prevalence (2,076 diagnoses, 840 per 100,000).

Of those RHD diagnoses with severity status recorded, just under half (48%) had mild disease (2,301 diagnoses), while 29% had severe disease (1,408).

Older people were more likely to have severe RHD, with 47% aged 45 or over having severe disease (650 diagnoses), compared to 13% of those aged 5–14 (61 diagnoses).



#### New RHD diagnoses

In this report, a 'new' rheumatic heart disease (RHD) diagnosis is defined as one that was diagnosed between 1 January 2014 and 31 December 2018. 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.

In 2014–2018, there were 1,586 reports of new RHD diagnoses in Queensland, South Australia, Western Australia and the Northern Territory (3.4 per 100,000 population).

For the 4 jurisdictions combined, RHD diagnosis rates between 2014 and 2018 have remained relatively stable, at around 3–4 diagnoses per 100,000 population annually. During this period, diagnosis rates varied by state and territory, but in general:

- South Australia had less than 1 diagnosis per 100,000 population
- Western Australia and Queensland had 2–3 diagnoses per 100,000
- Northern Territory had 30–50 diagnoses per 100,000.



New RHD diagnoses among all Australians in Qld, WA, SA and NT, by year, 2014 to 2018

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

### **Indigenous Australians**

Of the 1,586 new RHD diagnoses among all Australians in 2014–2018, 83% (1,314) were Indigenous Australians (60 per 100,000 population). The annual combined rate in Queensland, Western Australia, South Australia and the Northern Territory decreased slightly between 2014 and 2016, but then increased from 51 to 73 per 100,000 between 2016 and 2018. During the same period, the overall diagnosis rate for Indigenous Australians was around 100 times the rate for non-Indigenous Australians.

In 2014–2018, most new RHD diagnoses among Indigenous Australians were from the Northern Territory. The rate of new diagnoses in the Northern Territory was twice that of Western Australia, nearly 4 times that of Queensland and nearly 5 times that of South Australia. In 2018, 2 in 5 (132, 40%) new RHD diagnoses were from the Northern Territory.

### Age and sex

In 2014–2018, for all new RHD cases diagnosed among Indigenous Australians:

- the rate of new RHD diagnosis for females was nearly twice that for males (76 and 44 diagnoses per 100,000 population, respectively)
- females had higher rates compared to males in all age groups, excluding those aged 0–4
- 59% were aged under 25 years at diagnosis (773 people)
- 23 children were aged under 5 when diagnosed
- the median age at diagnosis was 20 years (15 years for males and 23 years for females).

The remainder of the information on RHD in this report (with the exception of deaths) relates to Indigenous Australians, due to the relatively small number of new cases recorded among non-Indigenous Australians.



#### New RHD severity at diagnosis

In 2014–2018, of the 1,314 Indigenous Australians with new RHD diagnoses:

- 58% had mild RHD when first diagnosed (764 diagnoses)
- 25% had moderate RHD (326)
- 16% had severe RHD (214).

This distribution was similar across states and territories. Distribution also varied by age group, with relatively large proportions of severe cases in the 0–4 and 45 and over age groups (21% and 30% of cases, respectively).

New RHD diagnoses among Indigenous Australians, by age group (years) and severity status (priority level), 2014–2018



Notes

1. Totals include 8 cases where severity status was inactive.

2. RHD severity assessment is at the time of diagnosis. These data exclude 2 cases with no assessment recorded.

3. Data includes Qld, WA, SA and NT.

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

## Region of management of people with ARF and RHD

For each ARF and RHD diagnosis 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.

In 2018, there were 5,440 Indigenous Australians being managed for ARF or RHD by health services in Queensland, Western Australia, South Australia and

the Northern Territory. This number includes all persons who were either diagnosed, assessed, receiving prophylaxis or undergoing surgery for RHD between 1 January 2016 and 31 December 2018 (reflecting the recommended frequency of evaluation every 2 years for patients with mild disease). The regions with the highest rates of management were:

- Rural Darwin (NT), with 5,305 cases per 100,000 population (659 persons)
- East Arnhem (NT), with 4,759 per 100,000 (542 persons)
- Katherine (NT), with 3,675 per 100,000 (407 persons).

Management rates in remote areas tended to be higher than those in less remote areas.

Rate of new RHD diagnoses among Indigenous Australians, by region of managment, 2014-2018



Note: Data only available for Queensland, Western Australia, South Australia and Northern Territory in 2014–2018. Source: AIHW analysis of National Rheumatic Heart Disease data collection.

## RHD surgery

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 Jurisdictional RHD control programs and registers for more information.

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. 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.

Since the commencement of registers, there were 1,864 RHD related surgeries recorded for 1,432 individuals. Of these surgeries, 76% occurred in Indigenous Australians (1,416). These figures reflect only those surgeries that were reported to the registers, and may not necessarily include all RHD-related surgery undertaken.

#### **Box 5: Surgical procedures**

#### **Balloon valvotomy**

Can be used to treat mitral stenosis. 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 the mitral valve (RHD Australia 2020, 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.

In 2014–2018, 349 Aboriginal and Torres Strait Islander people underwent 364 surgical events. Of these individuals:

- 336 had surgery once, 12 had surgery twice, 1 had 3 or more surgeries
- 185 were from the Northern Territory (53%)
- 100 were from Queensland (29%)
- 236 were female (68%).

The sex distribution of people undergoing RHD surgery is similar to the distribution of people living with RHD.

# Figure 2: Number of Indigenous Australians who had RHD surgery in 2014–2018



Chart: aihw.gov.au

Note: Data only available for Qld, WA, SA and NT.

Chart: AIHW. Source: AIHW analysis of the National Rheumatic Heart Disease Data Collection.

#### Surgery by age

In 2014–2018, of the 364 surgical events among Indigenous Australians, most occurred among those aged:

- 25-44 years, with 152 surgeries (42%)
- 45 and over, with 89 surgeries (24%).

During the same period, no children aged less than 5 had surgery for RHD.





2. This shows the age at surgery of the 492 surgical events 2014–2018. Individuals who had more than one surgery are included

multiple times as an individual could have more than one surgery in the period.

3. Data includes Qld, WA, SA and and NT. Source: AIHW analysis of National Rheumatic Heart Disease data collection.

### Surgery type

More than one procedure can be performed during a surgical event, and more than one valve can be involved.

During the 364 surgical events for Indigenous Australians in 2014–2018, 494 individual procedures were performed. The mitral valve was most frequently

operated on, with 310 procedures (63%). During the same period, 309 procedures (63%) involved valve replacement. Among these, replacement with a mechanical valve was more common than with a bioprosthetic valve. Almost one-third of procedures (156) involved repairing a valve.



Surgical procedures by surgery type and affected valve among Indigenous Australians with RHD, 2014–2018

Notes

1. Totals include 2 surgeries on other heart valves.

2. Other surgery types include Ross procedure and heart transplant and other procedures.

3. Data from NT, QId, SA and WA combined.

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

#### References

RHD Australia, (ARF/RHD writing group). The 2020 Australian guideline for the prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3rd 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. Journal of Heart Valve Disease. 22(3):333–9.

#### Deaths among all Australians with RHD

In 2014–2018, there were 315 deaths reported among people with RHD in the registers. Of these deaths:

- 248 deaths were Indigenous Australians (79%)
- 159 deaths occurred in Northern Territory (50%)
- 208 deaths occurred among females (66%)—similar to overall rates of RHD
- 135 deaths occurred among people aged 45–64 (43%)
- 56 years was the median age of death (similar for males and females).

Among Indigenous Australians with RHD who died, almost half (116) were aged 45–64 and 1 in 3 (84) were aged 15–44. Indigenous Australians with RHD who died during 2014–2018 had lived with their diagnosis for a median of 11 years.

These people could have been diagnosed at any time since RHD registers have been operational, but had died between 2014 and 2018. Of the people that died, 42% (132) had been diagnosed with RHD in or after 2008.

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.

## Secondary prophylaxis

Secondary prophylaxis with regular benzathine penicillin G (BPG) is the only rheumatic heart disease (RHD) control strategy shown to be both clinically and cost effective at community and individual levels (Webb 2015, Wyber & Carapetis 2015, RHD Australia 2020). According to the clinical guidelines in place when the data in this report were collected, the recommended regimen to prevent recurrences of acute rheumatic fever (ARF) and progression of RHD involves regular intramuscular injections of BPG every 21 to 28 days, for a minimum of 10 years (RHD Australia 2012). (The 2020 guidelines now state a minimum of 5 years since the most recent ARF episode.) This treatment aims to provide penicillin in the blood over a period of 3 to 4 weeks, providing protection against Strep A infections (Wyber 2013). As the penicillin concentration wanes, the individual's susceptibility to subsequent Strep A 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 such as injection refusal, pain caused by injections or a 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

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.

RHD Australia, (ARF/RHD writing group). The 2020 Australian guideline for the prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3rd edn). Northern Territory: RHD Australia, Menzies School of Health Research.

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

Webb RH, Grant C, Harnden A. 2015. Acute Rheumatic Fever. British Medical Journal 351(8017).

Wyber R & Carapetis J. 2015. Evolution, evidence and effect of secondary prophylaxis against rheumatic fever. Journal of Practice of Cardiovascular Sciences 1(1) 9–14.

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.

### Secondary prophylaxis among Indigenous Australians

This section focuses on BPG adherence amongst Indigenous Australians who were prescribed BPG on a 28-day regimen and who received at least one dose in 2018. 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 (i.e. not BPG) were excluded.

There were 3,209 Aboriginal and Torres Strait Islander people eligible for inclusion in calculations about BPG delivery in 2018. They were located in Northern Territory (1,604), Queensland (966), Western Australia (528), and South Australia (111). Of these:

- 23% (748 people) received 100% or more of their prescribed doses
- 19% (613) received 80% to 99% of their prescribed doses
- 32% (1,012) received 50% to 79% of their prescribed doses
- 26% (836) received less than 50% of their prescribed doses.

The delivery of prophylaxis was similar in 2017 and 2018, despite more people being prescribed BPG. The proportion of people receiving at least 80% of their scheduled doses is used as an indicator of prophylactic delivery at a level which is likely to protect against ARF recurrences (RHD Australia 2012). Data from the NT have shown that there is very minimal benefit when adherence is lower than 40% (de Dassel et al. 2018). In 2018, 19% of Indigenous Australians received fewer than 40% of their scheduled doses.

In 2018, 42% of Indigenous Australians (1,361 people) received at least 80% of doses. The proportion of people achieving at least 80% adherence was:

- South Australia (63%, 70 people)
- Northern Territory (53%, 848)
- Queensland (30%, 291)
- Western Australia (29%, 125).

Delivery of 100% of all the prescribed doses in a year is the gold standard for all people on prophylactic treatment. If someone is on treatment for an entire year, they should have at least 13 doses delivered. In 2018, the proportion of Indigenous Australians receiving 100% of their prescribed doses for the year, was:

- 33% (531 people) in Northern Territory
- 36% (40) in South Australia
- 12% in Queensland (114) and Western Australia (63).

#### Sex and age

In 2018, among Indigenous Australians prescribed prophylaxis:

- adherence was generally lower among those aged 15–44—27% receiving less than half their prescribed doses, and 42% receiving at least 80%
- among those aged 0–14 and 45 and over, more than half received at least 80% of prescribed doses
- 11 of the 16 children aged under 5, received 80% or more of their prescribed doses
- adherence was similar for males and females on prophylaxis
- around 40% of males and females received 80% or more of their prescribed dose
- 28% of males and 25% of females received less than 50% of their prescribed doses.

Adherence level of Indigenous Australians with ARF and/or RHD on a 28-day BPG regime, by state and territory, 2018



Notes

People on BPG can have more than 13 doses in one year, therefore 100% of doses is defined as 100%+ of doses.
This analysis only includes people who were prescribed prophylaxis for the whole of 2018.
Data includes QId, WA, SA and NT. Does not include cases managed in NSW.

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

### References

de Dassel JL, de Klerk N, Carapetis JR & Ralph AP 2018. How many doses make a difference? An analysis of secondary prevention of rheumatic fever and rheumatic heart disease. Journal of the American Heart Association 7(24): e010223.

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.

## **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 2018. During this period the NSW register recorded 61 diagnoses of ARF and 52 diagnoses of RHD.

The data presented below are for the 3 full calendar years 2016–2018 only. Data for secondary prophylaxis were provided for 2018. The data for NSW are not comparable to the data provided by Qld, WA, SA and the NT, in the previous sections.

#### **ARF in NSW**

In 2016–2018, there were 52 reported ARF diagnoses in NSW. Of these, 19 (37%) ARF diagnoses were reported among Aboriginal and Torres Strait Islander people, 16 (31%) diagnoses were in Pacific Islander people and 7 (13%) diagnoses were in people from other high risk groups. ARF rates were greater in males than females, and in 0–14 year olds compared to other age groups. Less than 10% of episodes were reported as recurrent ARF.

#### **RHD in NSW**

There are 52 known residents of NSW living with RHD who consented to be listed on the register as at December 2018; 43 of these cases were diagnosed between 2016 and 2018. Of these newly diagnosed cases, 10 (23%) identified as Indigenous Australians, 17 (40%) identified as Pacific Islander people, and 4 (9%) were from other high risk groups. More than half (24) of cases were females and just over half (22) were aged 15–34 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 2018. 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 2018 are also included in the data. There were 34 people in NSW prescribed secondary prophylaxis during 2018. Of these, 1 received 100% or more of their prescribed doses and a further 8 received 80–99% of their prescribed doses. Of all people prescribed secondary prophylaxis in 2018, 17 were Indigenous Australians.

## **Technical information**

### Australian guidelines for diagnosis of ARF

# Table 1: The 2012 Australian guidelines 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 <sup>(b)</sup>	2 major or 1 major and 2 minor manifestations plus evidence of a preceding Strep A infection <sup>(b)</sup>	
Definite recurrent episode of ARF in a patient with known past ARF or RHD	2 major or 1 major and 1 minor or 3 minor manifestations plus evidence of a preceding Strep A infection <sup>(b)</sup>	2 major or 1 major and 1 minor or 3 minor manifestations plus evidence of a preceding Strep A infection <sup>(b)</sup>	
Probable ARF (first episode or recurrence)	A clinical presentation that falls short by either one major or one minor manifestation, or the absence of streptococcal serology results, but one in which ARF is considered the most likely diagnosis. Such 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	Carditis (excluding subclinical evidence of	

	rheumatic valvulitis on	rheumatic valvulitis on
	Polyarthritis <sup>(c)</sup> or aseptic mono-arthritis or	Polyarthritis <sup>(c)</sup>
	polyarthralgia	Chorea <sup>(d)</sup>
	Chorea <sup>(d)</sup>	Erythema marginatum <sup>(e)</sup>
	Erythema marginatum <sup>(e)</sup>	Subcutaneous nodules
	Subcutaneous nodules	
Minor manifestations	Monoarthralgia	Fever <sup>(f)</sup>
	Fever <sup>(f)</sup>	Polyarthralgia or aseptic
	$ESR^{(h)} \ge 30 mm/h \text{ or}$	
	CRP(h) ≥30 mg/L	ESR(h) ≥30 mm/h or CRP(h) ≥30 mg/L
	Prolonged P-R interval	
	on ECG <sup>(9)</sup>	on ECG <sup>(g)(h)</sup>

CRP = C-reactive protein

ECG = electrocardiogram

ESR = erythrocyte sedimentation rate

#### Strep A = group A streptococcus

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

### Key reporting indicators

# Acute rheumatic fever and rheumatic heart disease key reporting indicators, 2020 guidelines

Indicator	Recommended disagreggations
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	1.4.1 80-100%
secondary propriyaxis each year	1.4.2 40-79% 1.4.3 0-39%

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

## **National RHD Data Collection: Data elements**

Persons with or at risk of RHD				
Data element name	Definition			
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 no	tification	
Data element name	Definition	
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		
Data element name	Definition	
RHD diagnosis date	The date on which a case was diagnosed with rheumatic heart disease.	
Prophylaxis prescript	ion	
Data element name	Definition	
•		

Prophylaxis prescribed date	The date on which a case is prescribed prophylactic treatment.	
Prophylaxis prescription end 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	y of IM The prescribed frequency of intramuscular antibiotic treatment to be administered to a case.	
Antibiotic administrat	cion	
Data element name	Definition	
Antibiotic administration date	The date of administration of intramuscular antibiotic to a case for secondary prevention of acute rheumatic fever.	
RHD surgery		
Data element name	e Definition	
RHD surgical The date of surgery for repair or replacement of he valve(s) in treatment of rheumatic heart disease.		
Heart valve procedure	e	
Data element name	Definition	
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		
Data element name	Definition	
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.	

#### 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 2018, ARF was notifiable to state health departments in five Australian jurisdictions (Qld, WA, SA, NT and NSW), 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.

This is the second 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 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 in 2015, but data are not available for the full period (2014–2018) included in this report. Data about ARF and RHD diagnoses are not currently 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.

#### Reference

ABS (Australian Bureau of Statistics) 2019. Estimates and projections, Aboriginal and Torres Strait Islander Australians, 2006 to 2031. ABS cat. no. 3238.0. Canberra: ABS.

### Abbreviations & symbols

#### Abbreviations

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

#### 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 (Strep A):** 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 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.