Acute rheumatic fever and rheumatic heart disease in New South Wales

Surveillance Report 2018 - 2022

NSW GOVERNMENT

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Produced by:

NSW RHD Program
Communicable Diseases Branch
Health Protection NSW
Locked Mail Bag 961
St Leonards NSW 2065
Email: NSWH-RHD@health.nsw.gov.au
www.health.nsw.gov.au/infectious/pages/default.aspx

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Contents

Executive Summary

- There were 82 notified cases of acute rheumatic fever (ARF) and 90 notified cases of rheumatic heart disease (RHD) between 1 January 2018 and 31 December 2022.
- The average crude notification rate of ARF was 0.2 cases per 100,000 population per year. The ARF rate decreased from 0.3 cases per 100,000 population per year in and 2018 to 0.2 per 100,000 in 2021 and 2022. This decrease may reflect high global circulation of group A streptococcal (GAS) infection between 2016 and 2018 as evidenced by increases in invasive GAS infection, scarlet fever, and GAS pharyngitis in the United Kingdom and United States of America[1,2], and the subsequent impact of COVID-19 pandemic restrictions.
- Most ARF cases were in children and young adults (73%) and 60% of RHD cases notified were in people aged less than 25 years of age.
- According to the definitions in the *CDNA National Guidelines for Public Health Units*[3], 70% of ARF cases were classified as definite cases, 13% were probable cases, and 16% were possible cases.
- Cardiac involvement at diagnosis was reported for 37% of cases with ARF.
- The average crude notification rate for RHD was 0.5 per 100,000 per year in people aged less than 35 years of age.
- Where severity was known, 71% of RHD cases notified had severe RHD. This is likely to reflect the high proportion of RHD cases identified through review of people admitted to hospital with ARF and RHD in NSW.
- Half of ARF cases and a third of RHD cases were in people living in areas with the lowest socio-economic index in NSW at diagnosis.
- As reported by other Australian states and territories[4], Aboriginal and Torres Strait Islander people were at substantially higher risk of both ARF and RHD. The average crude notification rates in Aboriginal and Torres Strait Islander people was more than 30 times higher for ARF than in non-Indigenous people and about 10 times higher for RHD.
- NSW has a higher proportion of cases in other at-risk groups than other states and territories. People reporting Māori and Pacific Islander ethnicity represented 16% of cases of ARF and 29% of cases of RHD.

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[1] Venkatesan P. Rise in group A streptococcal infections in England. Lancet Respir Med. 2023 Feb;11(2):e16. doi: 10.1016/S2213-2600(22)00507-0. Epub 2022 Dec 19. PMID: 36549319.

[2] Kennis M, Tagawa A, Kung VM, Montalbano G, Narvaez I, Franco-Paredes C, Vargas Barahona L, Madinger N, Shapiro L, Chastain DB, Henao-Martínez AF. Seasonal variations and risk factors of Streptococcus pyogenes infection: a multicenter research network study. Ther Adv Infect Dis. 2022 Oct 19;9:20499361221132101. doi: 10.1177/20499361221132101. PMID: 36277299; PMCID: PMC9585558.

[3] Communicable Diseases Network Australia (CDNA) (2017). Acute Rheumatic Fever & Rheumatic Heart Disease: CDNA National Guidelines for Public Health Units. Version 2. Department of Health and Aged Care, Australian Government. https://www.health.gov.au/resources/publications/acute-rheumatic-fever-and-rheumatic-heart-disease-cdna-national-guidelines-for-public-health-units?language=en

[4] Australian Institute of Health and Welfare (2023) Acute rheumatic fever and rheumatic heart disease in Australia 2017–2021, catalogue number CVD 99, AIHW, Australian Government.

- Cases were distributed across both metropolitan and rural and regional local health districts (LHD) in NSW. Aboriginal and Torres Strait Islander cases were more commonly reported from rural and regional LHDs. Most cases in people reporting Māori and Pacific Island ethnicity resided in metropolitan LHDs.

ARF and RHD in Aboriginal and Torres Strait Islander people

- There were 43 cases of ARF and 33 cases of RHD in Aboriginal and Torres Strait Islander people notified in NSW between 2018 and 2022.
- The average crude notification rate of ARF in Aboriginal and Torres Strait Islander people of all ages was 3.0 cases per 100,000 population per year. The RHD rate was 3.4 per 100,000 population per year for Aboriginal and Torres Strait islander people aged less than 35 years.
- Eighty-eight per cent of ARF cases and 94% of RHD cases in Aboriginal and Torres Strait Islander people were living in rural and regional LHDs.
- The highest notification rate of ARF was in Aboriginal and Torres Strait Islander people living in Northern NSW LHD. The highest notification rates of RHD were in people living in Western NSW LHD.

Methodology

ARF in people of any age, and RHD in people aged less than 35 years have been notifiable diseases in NSW since 2 October 2015. Notifications of ARF and RHD are based on clinical reports made by doctors and supplemented by active case finding through review of hospitalisation data for hospital admissions with ICD-10 codes related to ARF and RHD. Notifications received by NSW Health are held in the Notifiable Conditions Information Management System (NCIMS). This report was produced using data extracted from NCIMS on 28 June 2023. Data was reported by onset date for ARF and notification date for RHD from 1 January 2018 to 31 December 2022. LHD was reported based on place of residence at notification.

Country of birth and ethnicity data are based on the ABS Standard Australian Classification of Countries (1269.0). People from the Pacific Islands were defined as those from Melanesia, Micronesia, and Polynesia, excluding Papua New Guinea. Specific ethnicity that is collected in NCIMS allows people to nominate up to two ethnicities. People who report being an Aboriginal and/or Torres Strait Islander person and a distinct specific ethnicity, or those that report multiple ethnicities may be counted more than once in the data looking at priority groups.

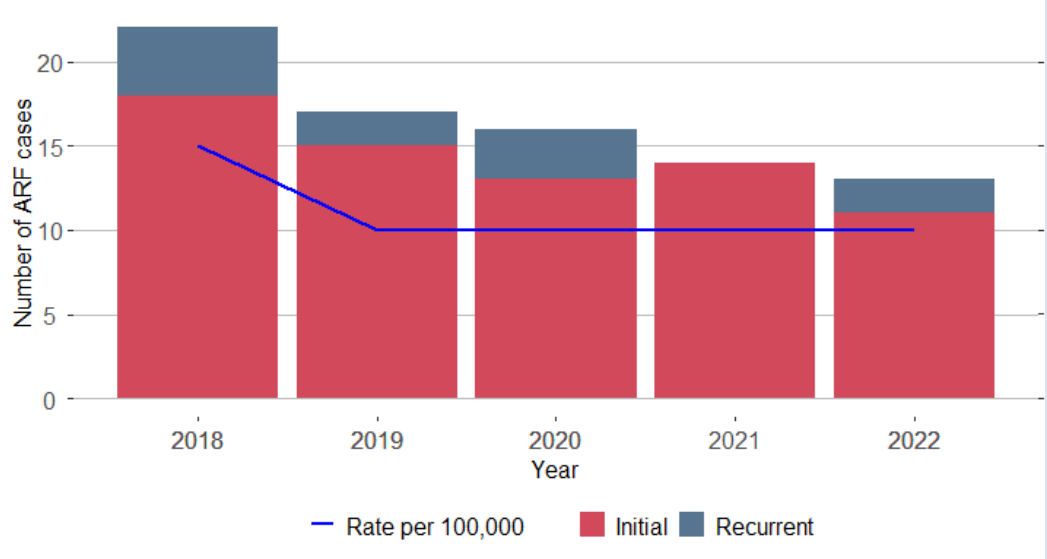
Population data including NSW mid-year population estimates, estimated populations by country of birth, population estimates by Index of Relative Socio-economic Disadvantage (IRSD) and population estimates by LHD were obtained from the Australian Bureau of Statistics (ABS) via the Secure Analytics for Population Health Research and Intelligence System (SAPHaRI). Rates for ARF were calculated using the entire population as the denominator. For RHD, rates were calculated with the same denominator limited to people aged 0-34 years.

High-risk groups are those living in communities with high rates of ARF (incidence >30/100,000 per year in people aged 5-14 years) or RHD (all-age prevalence >2/1000). In NSW, Aboriginal people and Torres Strait Islander people, people reporting Māori and Pacific Island ethnicity, and immigrants from countries with an RHD prevalence greater than 2 per 1000 population are considered high risk for ARF and RHD.

Acute Rheumatic Fever

A total of 82 cases of ARF were notified with an onset date between 1 January 2018 and 31 December 2022 (Figure 1). Of these, 11 (13%) had a previous recorded ARF diagnosis (recurrent cases), of which 3 (27%) were previously notified in NSW. In 2022, there were 11 cases with an initial (or first) diagnosis reported and 2 recurrent cases, compared to 14 initial and no recurrent cases in 2021. The median number of cases per year was 16 (range 13 to 22), and the average crude notification rate was 0.2 per 100,000 population per year (Figure 1). Cases were classified as definite, probable, or possible in line with *CDNA National Guidelines for Public Health Units*[5] (Table 1).

Figure 1: ARF cases and notification rate by year of onset, NSW, 2018 – 2022



82

Number of cases notified

Table 1: ARF cases by case classification and year, NSW, 2018 – 2022

Year	Case classification status			Total
	Definite	Probable	Possible	
2018	18	4	0	**22**
2019	10	2	5	**17**
2020	10	2	4	**16**
2021	11	1	2	**14**
2022	9	2	2	**13**
Total	**58**	**11**	**13**	**82**

0.2

Average crude rate per 100,000 populatio n

[5] Communicable Diseases Network Australia (CDNA) (2017). Acute Rheumatic Fever & Rheumatic Heart Disease: CDNA National Guidelines for Public Health Units. Version 2. Department of Health and Aged Care, Australian Government. https://www.health.gov.au/resources/publications/acute-rheumatic-fever-and-rheumatic-heart-disease-cdna-national-guidelines-for-public-health-units?language=en

Notifications by LHD

Northern NSW had the highest number of notifications between 1 January 2018 and 31 December 2022 with 19 cases, followed by Hunter New England with 18 cases (Table 2). The highest average crude notification rate was reported in Northern NSW (1.2 cases per 100,000 population) followed by Far West LHD (0.7 cases per 100,000 population).

Table 2: ARF cases by LHD of residence at onset, NSW, 2018 – 2022

LHD	Number of ARF cases						Average crude rate per 100,000
	2018	2019	2020	2021	2022	Total	
Northern NSW*	0	5	4	8	2	**19**	1.2
Hunter New England	6	5	3	3	1	**18**	0.4
Western Sydney	6	2	4	0	1	**13**	0.3
South Western Sydney	3	2	2	0	4	**11**	0.2
Western NSW	4	0	0	0	1	**5**	0.4
Nepean Blue Mountains	0	2	0	1	0	**3**	0.2
South Eastern Sydney	0	0	1	1	1	**3**	0.1
Sydney	0	0	1	0	2	**3**	0.1
Northern Sydney	1	0	1	0	0	**2**	0.0
Murrumbidgee	1	1	0	0	0	**2**	0.1
Illawarra Shoalhaven	0	0	0	0	1	**1**	0.0
Far West	0	0	0	1	0	**1**	0.7
Mid North Coast	1	0	0	0	0	**1**	0.1
Central Coast	0	0	0	0	0	**0**	0.0
Southern NSW	0	0	0	0	0	**0**	0.0

19

Highest number of cases by LHD - Northern NSW

1.2

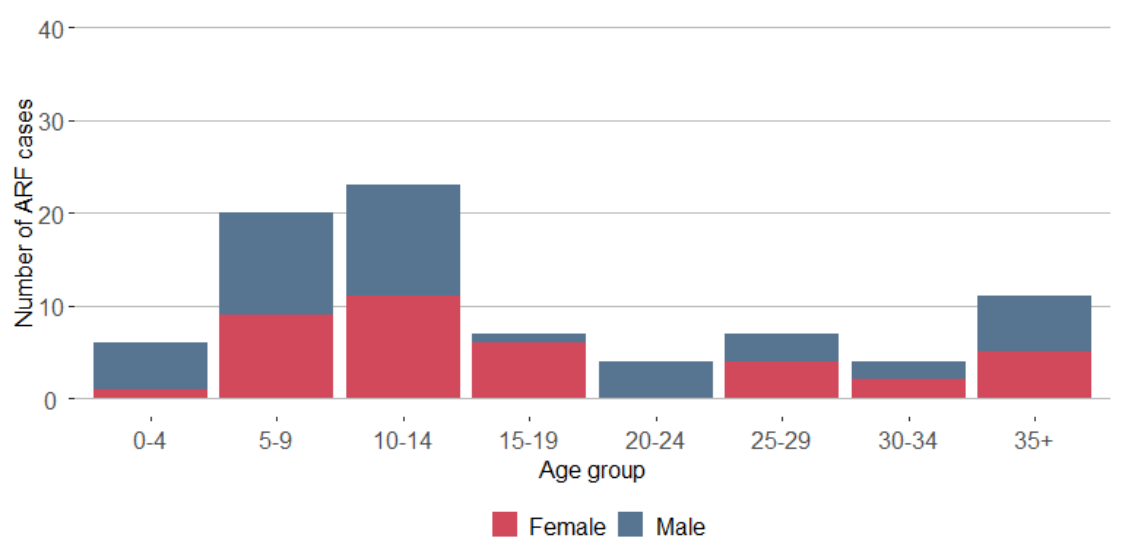
Highest average crude rate per 100,000 by LHD - Northern NSW

*A significant project to identify additional ARF/RHD cases was undertaken through Northern NSW Aboriginal Medical Services in the past 12 months.

Demographics

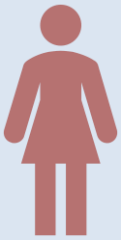
The median age at onset was 12 years (range 2 - 58) and 52% of notifications were in people aged 5-14 years (Figure 2). Males accounted for 54% of cases.

Figure 2: ARF cases by age group and sex, 2018 – 2022



54%

Male



46%

Female

Cases were more common in areas of lower socioeconomic status (Table 3).

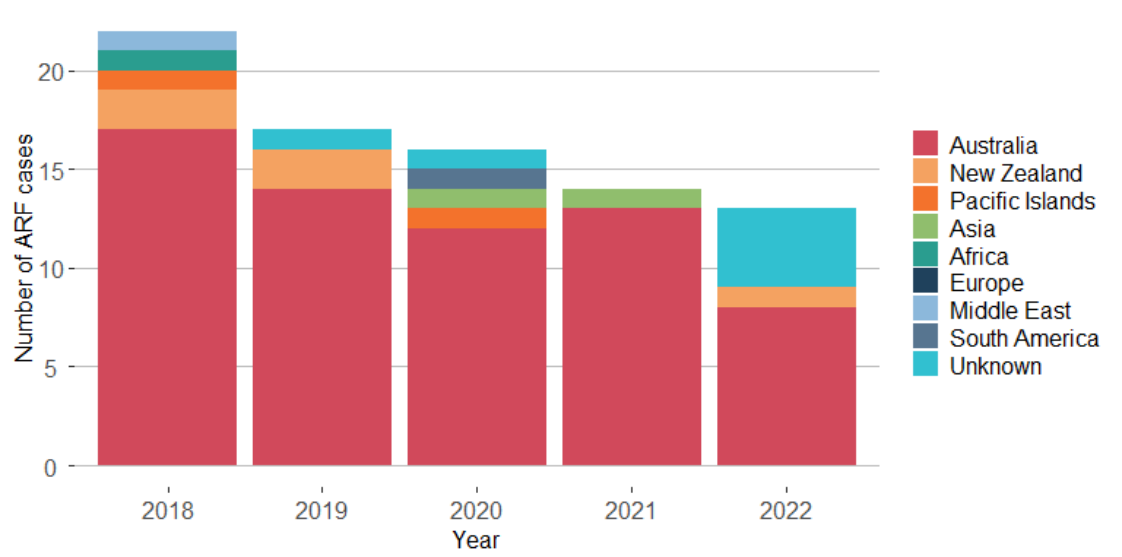
Table 3: Number of ARF cases by IRSD quintile, 2018 – 2022

	IRSD Quintile				
	1	2	3	4	5
Number of cases^	3	9	15	13	41
Crude notification rate per 100,000	0.04	0.12	0.18	0.19	0.57

^There was one notification where residence/IRSD quintile was unknown

At the time of reporting, country of birth was complete for 93% of cases. There were 16 countries reported. The most common country of birth was Australia with 64 cases (78%) (Figure 3). Due to low case numbers for other countries, cases are reported by region (Figure 3).

Figure 3: Country/region of birth by year of onset, NSW, 2018 – 2022



78%

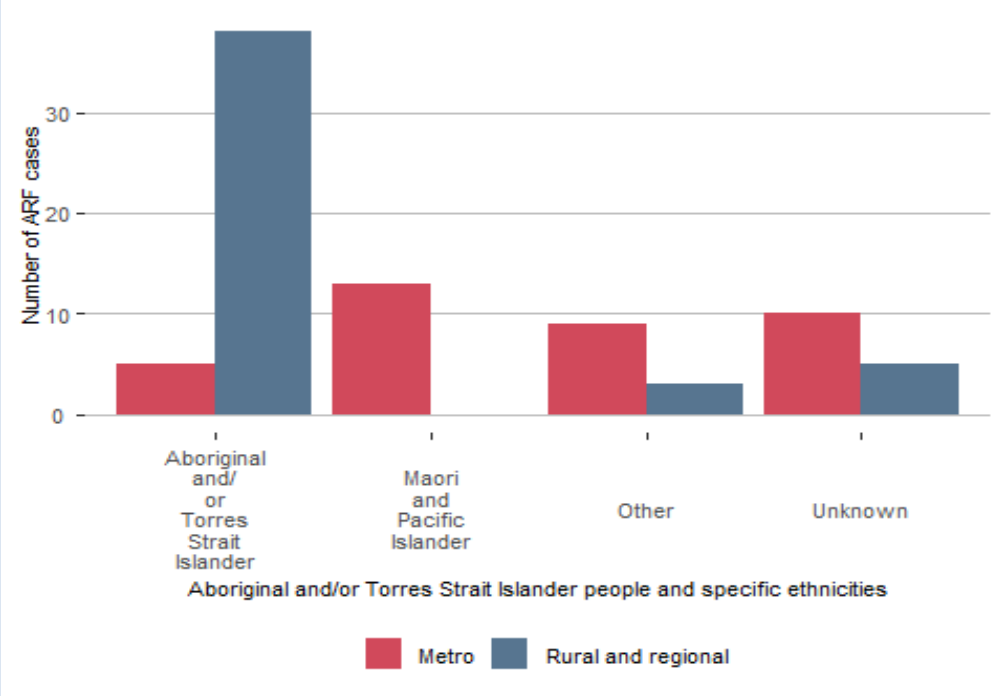
Cases born in Australia

15%

Cases born overseas

Aboriginal and Torres Strait Islander status was complete for 100% of cases, and specific ethnicity fields were completed for 80% of cases. There were 43 Aboriginal and/or Torres Strait Islander cases (52%) and 13 Māori and Pacific Islander cases (16%) (Figure 4). Aboriginal and Torres Strait Islander cases were more likely to live in rural and regional LHDs while Māori and Pacific Islander cases were more likely to live in metropolitan LHDs (Figure 4). Of the cases with ethnicity classified as 'Other' on Figure 4, eight cases (10%) were born in Australia (and did not identify as Aboriginal or Torres Strait Islander people). Ethnicities for cases who were born in Australia included Australian (63%), Middle Eastern heritage (13%), Asian heritage (25%) and European heritage (13%).

Figure 4: ARF cases in Aboriginal and/or Torres Strait Islander people and specific ethnicities by LHD of residence at disease onset, NSW, 2018 - 2022



Aboriginal and Torres Strait Islander people

During 2018-2022, 52% of notified ARF cases were in Aboriginal and Torres Strait Islander people (Table 4). The average crude notification rate from 2018 to 2022 in the Aboriginal and Torres Strait Islander population was 3.0 cases per 100,000 population per year compared to 0.1 cases per 100,000 population per year in non-Indigenous people (Table 4). The highest average crude notification rate in Aboriginal and Torres Strait Islander people was seen in Northern NSW LHD with a rate of 18.5 cases per 100,000 population per year (Table 5).

Table 4: ARF cases and average crude rate per 100,000 population in NSW by Aboriginal and/or Torres Strait Islander status, 2018 – 2022

	Year					Total/ Average
	2018	2019	2020	2021	2022	
Aboriginal and/or Torres Strait Islander cases	12	10	7	10	4	**43**
Non-Indigenous cases	10	7	9	4	9	**39**
Aboriginal and/or Torres Strait Islander rate	4.4	3.6	2.4	3.4	1.3	**3.0**
Non-Indigenous rate^	0.1	0.1	0.1	0.1	0.1	**0.1**

^Non-Indigenous rate includes those with missing or no information on Aboriginal and Torres Strait Islander status

Table 5: ARF cases and crude notification rate per 100,000 by Aboriginal and/or Torres Strait Islander status and LHD of residence at onset, 2018 – 2022

LHD	Number of ARF cases			Crude rate per 100,000	
	Aboriginal and Torres Strait Islander people	Non-Indigenous people*	Total	Aboriginal and Torres Strait Islander people	Non-Indigenous people
Northern NSW	14	5	**19**	18.5	0.3
Hunter New England	17	1	**18**	6.4	0.0
Western Sydney	2	11	**13**	2.6	0.2
South Western Sydney	1	10	**11**	1.2	0.2
Western NSW	4	1	**5**	2.4	0.1
Nepean Blue Mountains	-	3	**3**	-	0.2
South Eastern Sydney	2	1	**3**	4.6	0.0
Sydney	-	3	**3**	-	0.1
Northern Sydney	-	2	**2**	-	0.0
Murrumbidgee	2	-	**2**	2.9	-
Illawarra Shoalhaven	-	1	**1**	-	0.1
Far West	-	1	**1**	-	0.7
Mid North Coast	1	-	**1**	1.5	-
Central Coast	-	-	**0**	-	-
Southern NSW	-	-	**0**	-	-

Māori and Pacific Islander people

Between 2018 and 2022, there were 13 cases of ARF in people identifying as Māori and/or Pacific Islander, with an average crude notification rate of 2.2 cases per 100,000 population per year (Table 6). Samoan (n=8, average crude notification rate = 5.3 per 100,000), followed by Tongan (n=6, average crude notification rate = 5.3 per 100,000) were the most reported ethnicities among people identifying as Māori and/or Pacific Islander people.

Table 6: ARF cases and crude rate per 100,000 in Māori and/or Pacific Islander people, 2018 – 2022

	Year					Total/ Average
	2018	2019	2020	2021	2022	
Number of Māori/Pacific Islander cases	4	2	4	0	3	**13**
Māori/Pacific Islander rate	3.8	1.9	3.8	0.0	2.1	**2.2**

Clinical symptoms

The frequency of reported clinical symptoms and signs is shown in Table 7.

A longer duration of secondary prophylaxis therapy is recommended for people with cardiac involvement at ARF diagnosis: a minimum of 10 years for people with cardiac involvement and a minimum of 5 years for people without[6]. Thirty-seven per cent of cases diagnosed with ARF in NSW had evidence of cardiac involvement. A quarter of all cases were diagnosed with carditis and 22% had a prolonged PR interval.

Table 7: Presenting Jones criteria for ARF cases, 2018 - 2022^

Symptoms	Frequency	%
Elevated CRP	58	70.7
Fever	50	61.0
Elevated ESR	48	58.5
Polyarthralgia	40	48.8
Polyarthritis	28	34.1
Carditis	21	25.6
Prolonged P-R interval	18	22.0
Chorea	13	15.9
Other	11	13.4
Mono-arthritis, aseptic	6	7.3
Subcutaneous nodules	6	7.3
Mono-arthralgia, aseptic	3	3.7

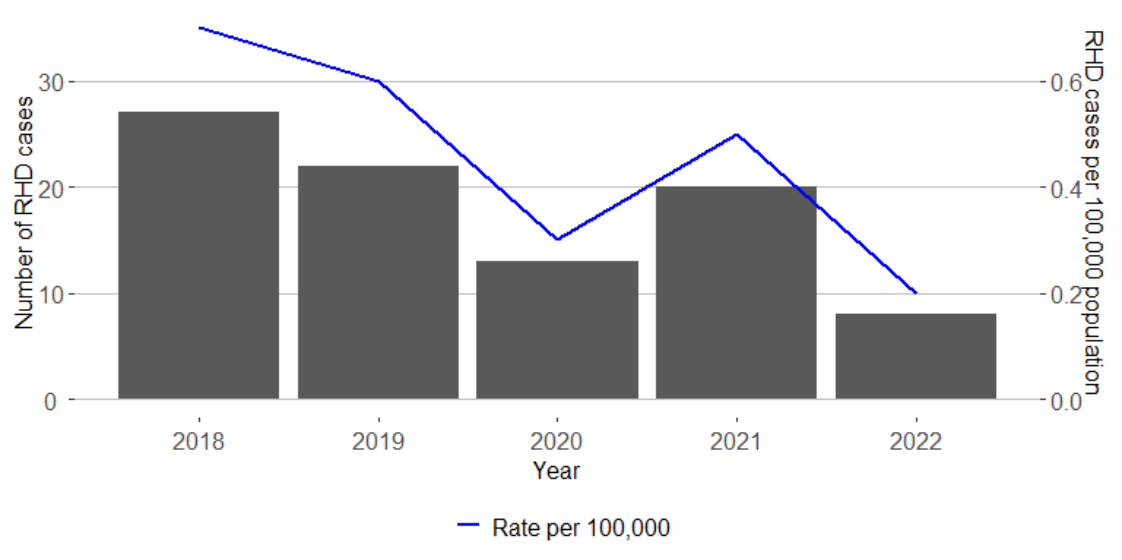
^ARF diagnosis requires the presence of multiple criteria, except in the case of chorea

[6] RHDAustralia (ARF/RHD writing group). The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3.2 edition, March 2022); 2020

Rheumatic Heart Disease

There were 90 cases of RHD were notified in people aged less than 35 years with a diagnosis date between 2018 and 2022 (Figure 5). There were eight cases reported for 2022, compared to 20 cases in 2021. The median number of cases per year was 20 (range 8 to 27), and the average crude notification rate was 0.5 cases per 100,000 population per year (Figure 5).

Figure 5: RHD cases and crude notification rate by year, NSW, 2018 – 2022



90

Number of cases

0.5

Average crude rate per 100,000 Population

Notifications by LHD

Western Sydney LHD had the highest number of cases of RHD between 2018 and 2012 with 26 cases followed by South Western Sydney LHD with 15 cases (Table 8). The highest average crude notification rate was reported in Western NSW LHD (1.9 cases per 100,000 population per year) followed by Western Sydney (1.0 cases per 100,000 population per year).

Table 8: RHD cases by LHD of residence at diagnosis, NSW, 2018 – 2022

LHD	Number of RHD cases						Average crude rate per 100,000
	2018	2019	2020	2021	2022	Total	
Western Sydney	5	6	6	6	3	**26**	1.0
South Western Sydney	6	2	1	5	1	**15**	0.6
Western NSW	2	4	2	3	1	**12**	1.9
Hunter New England	4	2	1	1	0	**8**	0.4
South Eastern Sydney	4	1	1	2	0	**8**	0.4
Mid North Coast	2	2	0	0	0	**4**	0.4
Illawarra Shoalhaven	0	0	0	2	2	**4**	0.2
Northern NSW	1	1	0	0	1	**3**	0.7
Northern Sydney	1	1	0	0	0	**2**	0.2
Sydney	1	0	1	0	0	**2**	0.1
Murrumbidgee	1	0	1	0	0	**2**	0.3
Central Coast	0	1	0	1	0	**2**	0.4
Nepean Blue Mountains	0	1	0	0	0	**1**	0.1
Far West	0	0	0	0	0	**0**	0.0
Southern NSW	0	0	0	0	0	**0**	0.0

26

Highest number of cases by LHD - Western Sydney

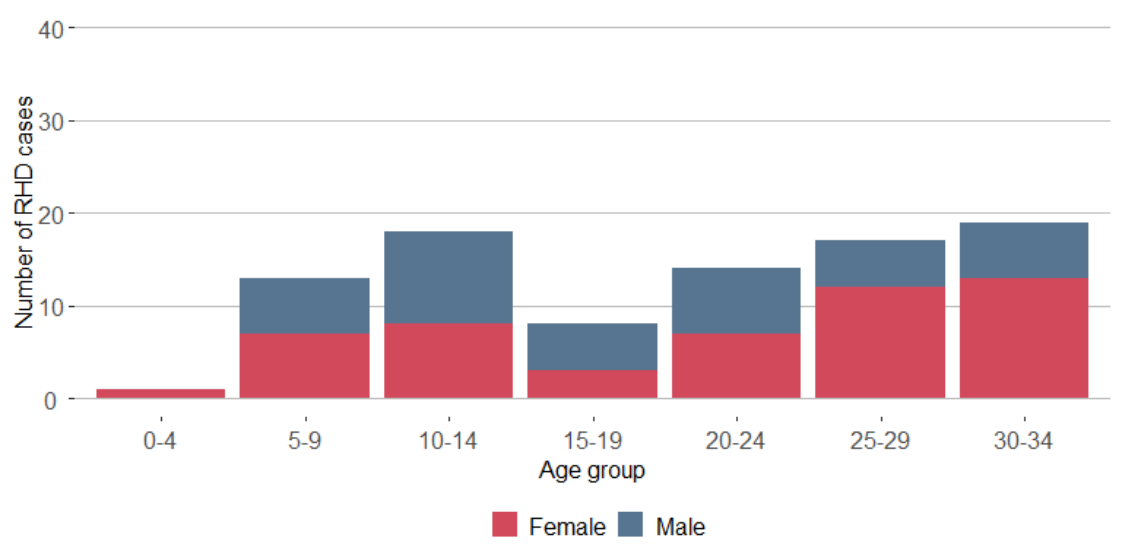
1.9

Highest average crude rate per 100,000 by LHD - Western NSW

Demographics

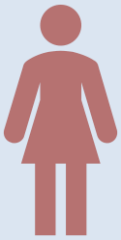
The median age at notification was 21 years (range 3-34) and 34% of cases were in people aged 5-14 years (Figure 6). Males accounted for 43% of cases.

Figure 6: RHD cases by age and sex, 2018 – 2022



43%

Male



57%

Female

Cases were more common in areas of lower socioeconomic status (Table 9).

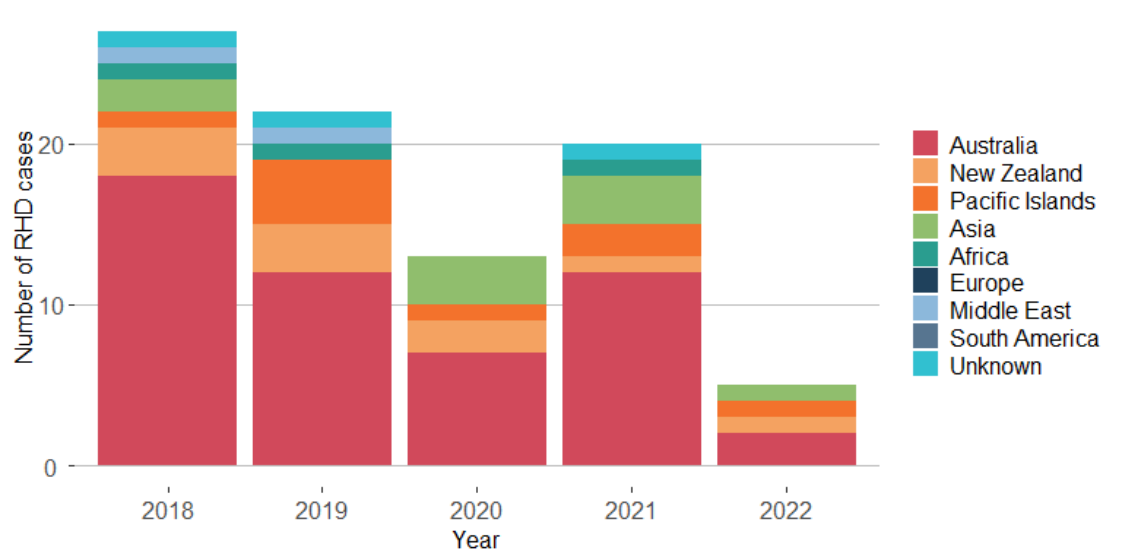
Table 9: RHD cases by IRSD quintile, 2018 – 2022

	IRSD Quintile				
	1	2	3	4	5
Number of cases^	5	16	28	12	28

^There was one notification where residence/IRSD quintile was unknown

At the time of reporting, country of birth was complete for 99% of cases. There were 20 countries reported. The most common country of birth was Australia with 51 cases (57%) (Figure 7). Due to low case numbers for other countries, cases are reported by region (Figure 7).

Figure 7: Country/region of birth of RHD cases, NSW, 2018 - 2022



58%

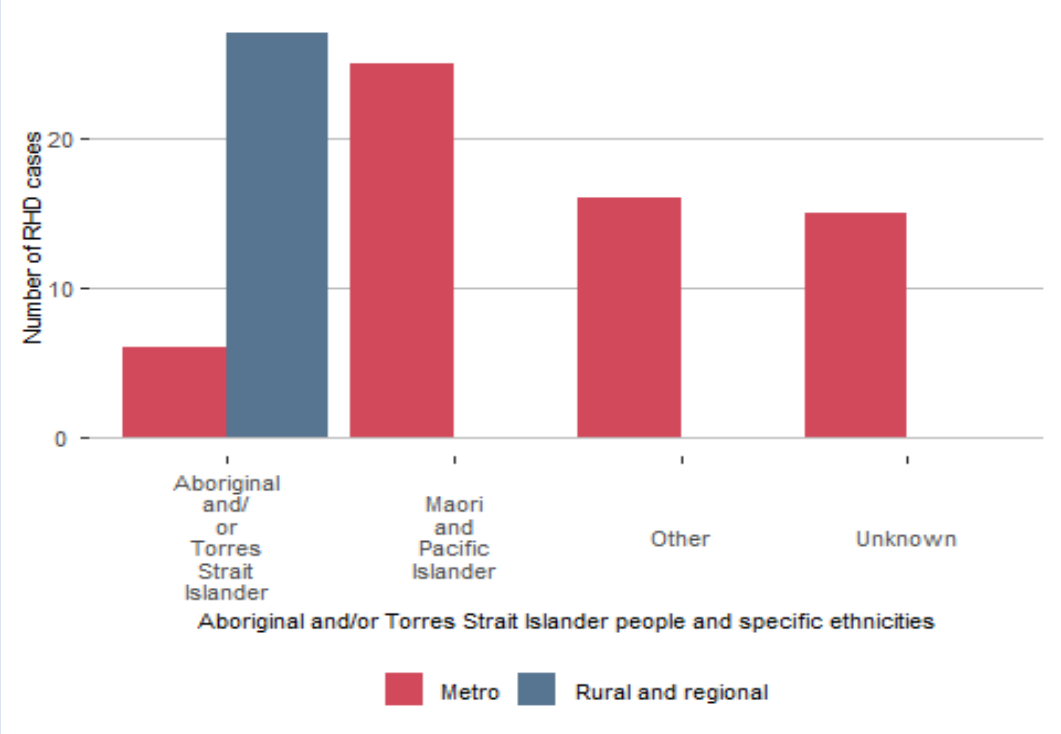
Cases born in Australia

40%

Cases born overseas

Data for Aboriginal and Torres Strait Islander status was complete for 100% of cases, and specific ethnicity data was complete for 83% of cases. There were 33 Aboriginal and/or Torres Strait Islander cases (37%) and 24 Māori and Pacific Islander cases (27%). Aboriginal and Torres Strait Islander cases were more likely to live in rural and regional LHDs while Māori and Pacific Islander cases were more likely to live in metropolitan LHDs (Figure 8). Of the cases with ethnicity classified as 'Other' on Figure 8, three cases (21%) were born in Australia (and did not identify as having Aboriginal and/or Torres Strait Islander background), with two identifying as being of Asian heritage and one identifying as of Australian heritage.

Figure 8: RHD cases in Aboriginal and/or Torres Strait Islander people and specific ethnicities by LHD of residence at disease onset, NSW, 2018 - 2022



Aboriginal and Torres Strait Islander people

In NSW, 37% of RHD cases occurred in Aboriginal and Torres Strait Islander people (Table 10). The average notification rate from 2018 to 2022 in the Aboriginal and Torres Strait Islander people was 3.4 cases per 100,000 population per year compared to 0.3 cases per 100,000 population per year in non-Indigenous people (Table 10). The highest average crude notification rate in the Aboriginal and Torres Strait Islander people was in Western NSW LHD with a rate of 15.2 cases per 100,000 population per year (Table 11).

Table 10: RHD cases and crude notification rate per 100,000 population in NSW by Aboriginal and/or Torres Strait Islander status, 2018 – 2022

	Year					Total
	2018	2019	2020	2021	2022	
Aboriginal and/or Torres Strait Islander cases	11	9	5	6	2	**33**
Non-Indigenous cases	16	13	8	14	6	**57**
Aboriginal and/or Torres Strait Islander rate	5.9	4.7	2.6	3.1	1.0	**3.4**
Non-Indigenous rate^	0.5	0.4	0.2	0.4	0.2	**0.3**

^Non-Indigenous rate includes those with missing or no information on Aboriginal and Torres Strait Islander status

Table 11: RHD cases and crude notification rate per 100,000 by Aboriginal and/or Torres Strait Islander status and LHD of residence at onset, 2018 - 2022

LHD	Number of RHD cases			Crude notification rate per 100,000	
	Aboriginal and Torres Strait Islander people	Non-Indigenous^ people	Total	Aboriginal and Torres Strait Islander people	Non-Indigenous people
Western Sydney	1	25	**26**	2.6	1.4
South Western Sydney	-	15	**15**	-	0.9
Western NSW	12	-	**12**	15.2	-
South Eastern Sydney	1	7	**8**	4.3	0.4
Hunter New England	8	-	**8**	5.6	-
Illawarra Shoalhaven	2	2	**4**	5.1	0.3
Sydney	-	4	**4**	-	0.3
Mid North Coast	3	-	**3**	9.4	-
Nepean Blue Mountains	1	1	**2**	2.6	0.2
Northern Sydney	-	2	**2**	-	0.1
Murrumbidgee	2	-	**2**	5.7	-
Northern NSW	2	-	**2**	5.9	-
Central Coast	1	-	**1**	2.8	-
Far West	-	-	**0**	-	-
Southern NSW	-	-	**0**	-	-

^Non-Indigenous includes those with missing or no information on Aboriginal and Torres Strait Islander status

Māori and Pacific Islander people

Between 2018 and 2022, there were 26 cases of RHD in people under the age of 35 years identifying as Māori and/or Pacific Islander, with an average crude notification rate of 7.84 cases per 100,000 population (Table 12). Samoan (n = 10) followed by Tongan (n = 6) were the most reported ethnicities among people identifying as Māori and/or Pacific Islander people.

Table 12: RHD cases and crude notification rate per 100,000 in Māori and/or Pacific Islander people, 2018 – 2022

	Year					Total/ Average
	2018	2019	2020	2021	2022	
Number of Māori/Pacific Islander cases	7	6	5	6	2	**26**
Māori/Pacific Islander rate	10.5	9.0	7.5	9.0	3.0	**7.84**

Severity

The severity of RHD was recorded for 69 cases (77%), for those individuals 49 were classified as severe, 13 as moderate and 7 as mild. Four cases notified between 2018 and 2022 died, three had the cause of death recorded as RHD, for the other it was unknown.