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# 16 Bulletin

# Rheumatic heart disease: all but forgotten in Australia except among Aboriginal and Torres Strait Islander peoples

## Highlights

- Acute rheumatic fever, which can lead to rheumatic heart disease, is very rare in Australia except among Aboriginal and Torres Strait Islander children and young people.
- The incidence of acute rheumatic fever among these children in regional areas of northern Australia is much higher than recently reported rates in other countries.

## Regional incidence of acute rheumatic fever in children

- The incidence of acute rheumatic fever among Aboriginal and Torres Strait Islander children aged 5–14 years in the Top End of the Northern Territory is about 250 per 100,000 and in Central Australia it is about 350 per 100,000.
- Incidence in Central Australia appears to be increasing but has remained relatively stable in the Top End of the Northern Territory.
- In 2002, Aboriginal and Torres Strait Islander children aged 5–14 years accounted for over 50% of new cases of acute rheumatic fever in these regions. There were no reported cases among the corresponding group of other Australian children.
- High incidence rates for acute rheumatic fever among Indigenous children have also been reported for the Kimberley region in Western Australia and in Far North Queensland.

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# Rheumatic heart disease

## Regional prevalence of rheumatic heart disease

- In 2002, in the Top End of the Northern Territory, the prevalence of rheumatic heart disease was just under 17 per 1,000 among Aboriginal and Torres Strait Islander peoples and less than two per 1,000 among other Australians. In Central Australia the prevalence among Aboriginal and Torres Strait Islander peoples was around 13 per 1,000 compared with less than one per 1,000 among other Australians living in the region.

## National hospitalisation and mortality

- In 2001–02, Indigenous people were six to eight times as likely to be hospitalised for acute rheumatic fever and rheumatic heart disease and nearly 20 times as likely to die from these diseases as other Australians.
- Among Indigenous people receiving heart valve surgery for acute rheumatic fever or rheumatic heart disease, nearly 45% are less than 25 years of age. By contrast, just 4% of these procedures are performed on other Australians of a similar age.

## The problem

Rheumatic heart disease is caused by the long-term damage done to the heart muscle or heart valves by acute rheumatic fever (see Box 1). Both acute rheumatic fever and rheumatic heart disease are important and preventable causes of ill health and death. They are a significant problem in the Indigenous Australian population of northern and central Australia, among children in particular. In contrast, they are extremely rare in other Australian children (Carapetis et al. 1996).

Acute rheumatic fever and rheumatic heart disease are typically associated with overcrowding, poor sanitary conditions and other aspects of social and economic disadvantage. In Australia, limited access for some population sub-groups to medical care for adequate diagnosis and treatment of these diseases is also recognised as a contributing factor to their occurrence and recurrence (Couzos & Carapetis 2003).

It is important to attend to both the social issues for Indigenous people and to manage the disease. The immediate aim in the management of these diseases is to identify those with acute rheumatic fever and, once identified, prevent the progression to rheumatic heart disease. Rheumatic heart disease can be prevented through early diagnosis and successful treatment of the acute condition. In practice it is estimated that more than 50% of 'newly diagnosed' cases of acute rheumatic fever are not actually new cases. Thus, by the time of diagnosis, rheumatic heart disease may already be established.

This bulletin describes the population patterns of acute rheumatic fever and rheumatic heart disease in Australia today using data obtained from regional registers and national databases.

## Australian registers for rheumatic heart disease

Tackling the problem of acute rheumatic fever and rheumatic heart disease is complex. Aspects of control programs may include (WHO 1995):

- a register<sup>1</sup>-based control system to track all known or suspected cases of acute rheumatic fever or rheumatic heart disease within a region;

<sup>1</sup> A register is a data file of all cases in a defined population.

- health education;
- secondary prevention (follow-up treatment with antibiotics); and
- collection of epidemiological data for monitoring.

The regional disease registers are used both in patient care and for monitoring the extent of the disease.

In terms of patient care, the regional registers can help coordinate individual patient management and improve adherence to drug treatment to prevent recurrent rheumatic fever and the associated cumulative heart valve damage. A system can be established to indicate that the monthly penicillin injection is due, and to later check that the injection has been provided. Clinic based patient information systems also have the ability to maintain care plans and service level registers for rheumatic heart disease. Adopting this approach has been associated with reduced acute rheumatic fever recurrences and reduced incidence of rheumatic heart disease (Thornley et al. 2001).

### *Box 1: Acute rheumatic fever*

#### *What causes acute rheumatic fever?*

Acute rheumatic fever is a delayed complication of an untreated throat infection with group A streptococcus (GAS) bacteria. The rate of development of acute rheumatic fever in individuals with untreated streptococcal infection is estimated to be 3% (US NLOM & NIH 2003). Those most at risk are children and young adults. It can affect the heart valves, the heart muscle and its lining, the joints and the brain.

Acute rheumatic fever is also suspected of being caused by streptococcal skin sores (McDonald et al. 2004). Carapetis et al. (1997a) reported that the pool of streptococcal bacteria found in skin sores comprise up to 70% of the total pool in some Indigenous communities whereas throat carriage of the bacteria in the communities may be quite low. The ongoing epidemic of scabies in Indigenous communities is thought to contribute to the high prevalence of streptococcal skin infections (Carapetis & Currie 1998).

#### *How is acute rheumatic fever diagnosed?*

There is no specific laboratory test that can diagnose acute rheumatic fever. The diagnosis is a combination of clinical and laboratory features and recent evidence of a GAS infection. Acute rheumatic fever can include a variety of signs and symptoms. Some of the more common features include fever, joint pain and swelling, and carditis (inflammation of the heart). Rare features include a movement disorder called Sydenham's chorea and specific skin disorders. Repeated episodes of rheumatic fever can cause permanent damage to the heart valves (rheumatic heart disease).

#### *Why do so many Indigenous children with acute rheumatic fever develop rheumatic heart disease?*

Rheumatic heart valve damage is the result of repeated or prolonged episodes of acute rheumatic fever in childhood and adolescence. In many Indigenous children these episodes are not being diagnosed, and are therefore not treated (Carapetis et al. 1996). The need for continued treatment over a long period compounds the problem.

#### *Can acute rheumatic fever or rheumatic heart disease be prevented?*

Recurrences of acute rheumatic fever lead to cumulative heart damage but this can be prevented by strict follow-up and monthly injections of penicillin for at least five years (and often longer) after the last episode (Mincham et al. 2002).



## Rheumatic heart disease

In terms of monitoring, the regional registers provide valuable information on the incidence and prevalence of the diseases, which is necessary to determine the extent of the problem, as well as monitoring progress on addressing the situation. It is the monitoring function of the registers that is the focus of this bulletin.

A rheumatic heart disease control program was established in the Top End of the Northern Territory in late 1997. In 2000, the program was extended to cover people living in Central Australia and was fully operational in mid 2002 (DHA 2003). Geographically, the Top End program covers the northern part of the Northern Territory including coastal and island communities and deep into the desert bordering Western Australia. It includes the Darwin urban area and three health regions: Darwin Remote, Katherine and East Arnhem. The Central Australian program covers a large area of the Northern Territory, including Alice Springs and Barkly Regions, the Anangu-Pitjantjatjara lands in South Australia, the Ngaanyatjarra lands in Western Australia, and extends to the Queensland border (CARHDSC 2002). It does not extend to the Kimberley or Pilbara regions of Western Australia. Together the two programs cover a population of over 200,000 people.

### **What the Top End and Central Australian registers tell us about incidence and prevalence**

The registers collect data related to diagnosis, hospitalisations, compliance with preventive penicillin use, clinical progress, surgery and mortality (CARHDSC 2002). This information assists clinics in managing the care of people registered with the rheumatic heart disease programs.

#### **Incidence of acute rheumatic fever**

Acute rheumatic fever is frequently under-reported because of difficulty in diagnosis and reduced awareness of the disease—therefore its true incidence is underestimated. The peak age of incidence of acute rheumatic fever is 5–14 years, but cases do occur in adults. The disease is rare in children under four years of age.

Incidence, as it is reported here from data provided by the registers, includes new and recurrent<sup>2</sup> cases.

In 2002, 58 people were diagnosed with acute rheumatic fever in the Top End of the Northern Territory—all were Australian Aboriginals, and females accounted for just over two-thirds of identified cases. Most cases (83%) were hospitalised and about a third of these (35%) were for recurrences. Aboriginal children aged 5–14 years accounted for 32 cases of acute rheumatic fever (55%)—a rate of 346 per 100,000 in this population. There were no reported cases of acute rheumatic fever among non-Indigenous children living in the area.

Data from Central Australia present a similar picture. In 2002, there were 27 cases of acute rheumatic fever. Of these, 30% (8 cases) were recurrences. The highest incidence rates were found in the 5–14 year age group (365 per 100,000).

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2 A recurrence is defined as a repeat episode of acute rheumatic fever three months or more after the last episode.

## Trends in incidence of acute rheumatic fever

Between 1989–93 and 1998–02, the incidence of acute rheumatic fever in Indigenous children aged 5–14 years in the Top End of the Northern Territory remained relatively stable with rates around 250 per 100,000 (Table 1). Falls in recurrence rates from about 40% in 1987–96 to 16% of all episodes in the late 1990s were also observed (Couzos & Carapetis 2003).

By contrast, the incidence of acute rheumatic fever in Central Australia increased with rates on average around 250 per 100,000 in the mid 1990s rising to an average annual rate of about 350 per 100,000 in 1998–02 (Table 1). In Central Australia, there was a recorded increase in incidence among 15–24 year olds over this period, rising from 83 per 100,000 in 1995 to 187 per 100,000 in 2002. In Central Australia, between 1995 and 2002, recurrences accounted for between 23% and nearly 60% of all cases of acute rheumatic fever. More importantly, before the program began in this region (pre-2001), recurrences accounted for 40% of cases on average, but these had fallen to 26% in 2001–02 (CARHDSC 2002).

These trends need to be interpreted with caution, however. For example, in the Top End, the population estimates for Indigenous children increased dramatically (by more than 25%) in the last ten years. So although the average number of new cases over this period has increased, the age-specific incidence has remained relatively stable. In Central Australia, apparent increases in incidence may simply reflect better reporting to the register, which was quite low in the early years (less than 50% of cases), but was estimated to be nearly 90% in 2001–02 (CARHDSC 2002).

**Table 1: Incidence<sup>(a)</sup> of acute rheumatic fever among Aboriginal and Torres Strait Islander children aged 5–14 years in the Top End of the Northern Territory and in Central Australia, 1989–2002**

Year	Top End		Central Australia <sup>(b)</sup>	
	Number	Age-specific rate <sup>(c)</sup>	Number	Age-specific rate <sup>(c)</sup>
<b>1989–93</b>	<b>91</b>	<b>254<sup>(d)</sup></b>	<i>n.a.</i>	<i>n.a.</i>
1994	18	204	<i>n.a.</i>	<i>n.a.</i>
1995	13	148	8	198
1996	21	238	13	319
1997	14	159	9	222
1998	24	270	12	295
1999	23	254	18	440
2000	14	154	16	388
2001	19	210	11	265
2002	32	346	15	365
<b>1998–02</b>	<b>112</b>	<b>245</b>	<b>72</b>	<b>351</b>

*n.a.* Not available. (Acute rheumatic fever was added to the list of notifiable diseases in the Northern Territory in 1995, so notifications in Central Australia were not available before this time).

(a) Includes new and recurrent cases of acute rheumatic fever.

(b) Excludes those cases from South Australia and Western Australia that are serviced by the Alice Springs hospital, because of difficulties ascertaining denominator populations over time.

(c) Rate per 100,000 Indigenous children aged 5–14 years in each region.

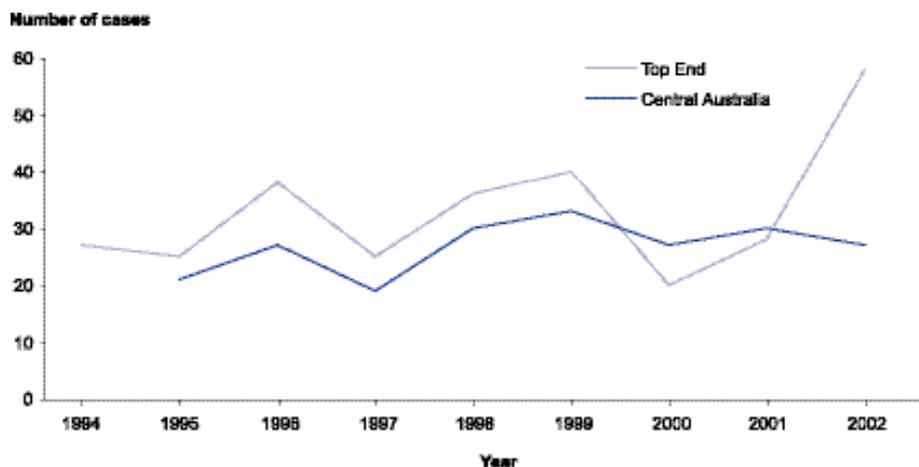
(d) Carapetis et al. (1996).

Sources: Top End Rheumatic Heart Disease Register and the Central Australian Rheumatic Heart Disease Register.

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Assessing trends in the incidence of acute rheumatic fever is also hindered by the variability in the number of new and recurrent cases from one year to the next (Figure 1). For example, in the Top End between 2001 and 2002 the number of cases nearly doubled (from 28 to 58) whereas between 1999 and 2000 the number of cases halved (from 40 to 20). To date, the number of new and recurrent cases has been less variable in Central Australia, but the numbers are too small and inconsistent to assess trends.

**Figure 1: Number of new and recurrent cases of acute rheumatic fever among Aboriginal and Torres Strait Islander peoples in the Top End of the Northern Territory and in Central Australia, all ages, 1994–2002**



Sources: Top End Rheumatic Heart Disease Register and the Central Australian Rheumatic Heart Disease Register.

## Prevalence of rheumatic heart disease

In 2002, there were 696 people with rheumatic heart disease in the Top End of the Northern Territory; 92% of these were Aboriginal and Torres Strait Islander peoples (643 cases). About twice as many cases were females (65%) compared with males (35%). Rheumatic heart disease was present in 54 children aged 5–14 (8% of all cases), all of whom were Indigenous Australians. Overall, the prevalence of rheumatic heart disease among Aboriginal and Torres Strait Islander peoples in the Top End was 17 per 1,000 in 2002, but rates were highest among 25–44 year olds where nearly 3% of the Indigenous Australian population in this age group is affected by the disease (Table 2).

In 2002, there were 283 cases of rheumatic heart disease in Central Australia. Of these, 267 cases (94%) were reported by Aboriginal and Torres Strait Islander peoples. This represents a prevalence rate of around 13 per 1,000 among Indigenous Australians compared with less than one per 1,000 among other Australians. Over two-thirds of cases of rheumatic heart disease in Central Australia were in the 15–44 year age group. The highest prevalence occurred among 15–24-year-old Aboriginal and Torres Strait Islander peoples (19 per 1,000, or almost 2% of this population) (Table 2).

**Table 2: Prevalence<sup>(a)</sup> of rheumatic heart disease in the Top End of the Northern Territory and in Central Australia, 2002**

Age (years)	Top End		Central Australia	
	Indigenous Australians	Other Australians	Indigenous Australians	Other Australians
5–14	5.8	0.2	7.6	0.0
15–24	19.8	0.3	18.6	0.5
25–44	29.4	0.8	15.8	0.4
45+	18.6	0.8	14.9	1.4
Total	16.6	1.7	12.5	0.6

(a) Per 1,000 population in each region.

Sources: Top End Rheumatic Heart Disease Register and the Central Australian Rheumatic Heart Disease Register.

### Trends in prevalence of rheumatic heart disease

Despite a stabilising of trends in the incidence of acute rheumatic fever in the Top End of the Northern Territory, the reported prevalence of chronic rheumatic heart disease continues to increase. In 2002, there were 17 cases per 1,000 Indigenous Australians, compared with nine per 1,000 in 1995. This increase is likely to be due to an improvement in reporting and case finding, and better awareness of the condition and its symptoms, rather than an actual rise in the number of cases.

It is too early to assess trends from the Central Australian register.

## National data on acute rheumatic fever and rheumatic heart disease

### Hospitalisation

Acute rheumatic fever is a serious disease and usually requires hospitalisation. Richmond & Harris (1998) reported hospitalisation rates in the Kimberley region of Western Australia as high as 80% for new cases of acute rheumatic fever and 73% for recurrences. In the Northern Territory, it is recommended that all cases of acute rheumatic fever be hospitalised for proper assessment.

Reflecting the incidence and prevalence of acute rheumatic fever and rheumatic heart disease obtained from the Top End of the Northern Territory and the Central Australian registers, the national hospitalisation rate for these diseases is much higher among Indigenous Australian males and females than among other Australians (Table 3). In 2001–02, the rate of hospitalisation for acute rheumatic fever and rheumatic heart disease among Indigenous Australian males was six times as high, and among Indigenous Australian females was eight times as high, as the rates among other Australians. These rate ratios were substantially higher than for other cardiovascular diseases.

In 2001–02, there were 341 hospitalisations in Australia for either acute rheumatic fever or rheumatic heart disease among those identified as Indigenous. This represents 15% of all hospitalisations for these diseases. The remaining 85% are likely to be for treatment of rheumatic heart disease among other Australians. These data cannot separate initial and subsequent hospitalisations for the same individual.

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**Table 3: Hospitalisations among Indigenous patients for principal diagnosis of the major cardiovascular diseases<sup>(a)</sup>, by sex, 2001–02**

	Males			Females		
	Number	Rate <sup>(b)</sup>	Ratio of the rate for Indigenous Australians compared with the rate for other Australians <sup>(c)</sup>	Number	Rate <sup>(b)</sup>	Ratio of the rate for Indigenous Australians compared with the rate for other Australians <sup>(c)</sup>
Acute rheumatic fever and rheumatic heart disease	125	0.5	5.7	216	0.9	8.1
Coronary heart disease	1,568	15.9	1.4	1,243	11.6	2.2
Heart failure	516	5.9	2.4	507	5.7	3.1
Stroke	238	2.8	1.2	304	3.4	1.9
Peripheral vascular disease	90	1.3	0.7	50	0.6	0.7
Other diseases of the circulatory system	951	8.7	0.9	1,028	8.3	1.0
<b>Total</b>	<b>3,488</b>	<b>35.0</b>	<b>1.3</b>	<b>3,348</b>	<b>30.6</b>	<b>1.7</b>

(a) Data are for public and most private hospitals. Disease categories are based on ICD-10-AM codes I00–I02 for acute rheumatic fever; I05–I09 for rheumatic heart disease; I20–I25 for coronary heart disease; I50 for heart failure; I60–I69 and G45 for stroke; I70–I74 for peripheral vascular disease and I26–I28, I30–I49, I51–I52 and I75–I99 for other diseases of the circulatory system.

(b) Per 1,000 population. Hospitalisation rates have been directly age-standardised to the total Australian population as at 30 June 2001.

(c) The rate of hospitalisations for patients identified as Indigenous divided by the hospitalisation rate among other Australians.

Source: AIHW National Hospital Morbidity Database.

It is likely that these are underestimates of the rates of hospitalisation for circulatory diseases, including acute rheumatic fever and rheumatic heart disease, as identification of Indigenous status is incomplete in the national hospital morbidity database. Similarly, comparisons of hospitalisation rates between states and territories have not been presented because regional differences are affected by the variation in Indigenous identification among state and territory hospital data collections (AIHW 2003).

## Surgical procedures

Acute rheumatic fever, particularly recurrences of the disease, can cause damage to the heart valves. A defective heart valve is one that fails to fully open or close. Historically, Aboriginal and Torres Strait Islander peoples in the Northern Territory with severe rheumatic heart disease have undergone heart valve replacement surgery with mechanical devices at a late stage in their illness. However, techniques of valve repair have been shown to improve health outcomes compared with valve replacement (Carapetis et al. 1997b).

Recent national data show that there were 106 heart valve procedures among Indigenous Australian patients in 2001–02 where acute rheumatic fever or rheumatic heart disease was the principal diagnosis during hospitalisation (Table 4). Mitral and aortic valve replacements comprised nearly 60% of these, and repairs comprised 23%. The situation among other Australians was similar, where mitral and aortic valve replacements comprised 67% and valve repairs comprised 15%. Consistent with the incidence and prevalence of these diseases, about two-thirds were performed on females.

Among patients undergoing these procedures, 45% of Indigenous Australian patients were less than 25 years old compared with just 4% of other Australians of a similar age.

As more than one procedure can be performed at each hospital visit, and the data do not indicate repeat hospitalisations for the same individual, it is difficult to estimate the proportion of individuals with acute rheumatic fever or rheumatic heart disease who have undergone heart valve procedures.

**Table 4: Heart valve procedures for principal diagnosis of acute rheumatic fever or rheumatic heart disease(a), by Indigenous status, 2001–02**

Heart valve procedure	Indigenous Australians		Other Australians	
	Number	Proportion of all heart valve procedures %	Number	Proportion of all heart valve procedures %
Replacement of mitral valve	42	39.6	435	35.4
Replacement of aortic valve	20	18.9	390	31.7
Repair of mitral or aortic valve	24	22.6	178	14.5
Other heart valve procedures	20	18.9	227	18.5
<b>Total</b>	<b>106</b>	<b>100</b>	<b>1,230</b>	<b>100</b>

(a) Acute rheumatic fever refers to ICD-10 codes I00–I02 and rheumatic heart disease to ICD-10 codes I05–I09.

Source: AIHW National Hospital Morbidity Database.

## Deaths

Similar to differences in hospitalisation rates, Indigenous Australians are much more likely to die from acute rheumatic fever and rheumatic heart disease than other Australians. Although the numbers of deaths were higher among other Australian males and females than among Indigenous Australians over the period 2000–02, the death rates for Indigenous Australian males and females were 17 and 21 times the respective death rates in other Australians (Table 5).

**Table 5: Standardised mortality ratios where acute rheumatic fever or rheumatic heart disease(a) was the underlying cause of death, by Indigenous status and by sex, 2000–02**

	Indigenous Australians		Other Australians	
	Number of deaths	Standardised mortality ratio <sup>(b)</sup>	Number of deaths	Standardised mortality ratio <sup>(b)</sup>
Males	18	16.6	95	1.0
Females	37	21.1	164	1.0

(a) Acute rheumatic fever refers to ICD-10 codes I00–I02 and rheumatic heart disease to ICD-10 codes I05–I09.

(b) The standardised mortality ratio has been presented in this table, instead of death rates, due to the small number of deaths in the Indigenous Australian population. For further information see the Methods section.

Note: All deaths are based on year of registration for usual residents of Queensland, South Australia, Western Australia and the Northern Territory.

Source: AIHW National Mortality Database.

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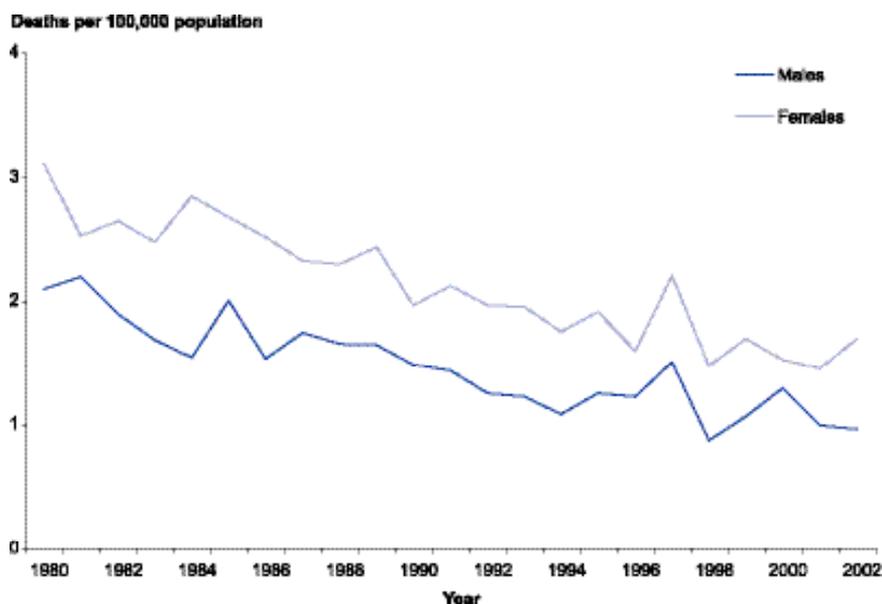
The majority of deaths (about 90%) for acute rheumatic fever and rheumatic heart disease among other Australians occur in those aged 55 years and over, and reflect a time (pre-1960s) when acute rheumatic fever was more common among children in the general population.

## Trends

Trends in death rates have been assessed for the whole population because until fairly recently identification of Indigenous status on death certificates has been poor.

Between 1980 and 2002, the national death rates for rheumatic heart disease fell on average by 3–4% per year for both males and females (Figure 2). Death rates are higher among females than males (1.7 per 100,000 in females compared with 1.0 per 100,000 in males in 2002) reflecting the higher incidence and prevalence of acute rheumatic fever and rheumatic heart disease in females.

**Figure 2: Trends in death rates among the total population where the underlying cause of death was rheumatic heart disease, by sex, 1980–02**



### Notes

1. Rheumatic heart disease refers to ICD-9 codes 393–398 and ICD-10 codes I00–I09.
2. Death rates have been directly age-standardised to the total Australian population as at 30 June 2001.
3. All deaths are based on year of registration.

Source: AIHW National Mortality Database.

## International comparisons

Much is written about the incidence and prevalence of acute rheumatic fever and rheumatic heart disease in developing countries. The World Health Organization (WHO) estimated that in 1994 acute rheumatic fever and rheumatic heart disease affected 12 million people, mostly children, with 400,000 deaths annually (WHO 1995). For developing countries it has also been estimated that acute rheumatic fever and rheumatic heart disease are responsible for almost half of the cardiovascular disease in all age groups and are leading causes of death in the first five decades of life (Limbu & Maskey 2002). By contrast, the dramatic decline in the incidence of acute rheumatic fever from 100–200 cases per 100,000 in the early 1900s (Limbu & Maskey 2002) to current estimates of 0.2–0.5 per 100,000 is indicative of the trend in most developed countries (Carapetis et al. 1996). In 1980, however, WHO noted an incidence rate of 100 per 100,000 of acute rheumatic fever or rheumatic heart disease among the younger age groups of socially disadvantaged populations in developed countries.

Despite the much lower incidence and prevalence of acute rheumatic fever and rheumatic heart disease in developed countries, they remain important causes of morbidity and mortality in some parts of Australia and in similar countries (Table 6). In New Zealand, acute rheumatic fever and rheumatic heart disease continue to be major health problems, particularly among Maori and Pacific Islander peoples. For the period 1995–00, the annual incidence of acute rheumatic fever was 2.8 per 100,000, an increase of 12% over that reported in 1990–95. The annual rate among those aged 5–14 years was 13.8 per 100,000 (Thornley et al. 2001). In a review by Franks (2002), acute rheumatic fever incidence rates of between 100 to 200 per 100,000 among Samoan children in Hawaii and Maori and Pacific Islander children in Auckland aged less than 20 years were reported throughout the 1980s.

**Table 6: International comparisons of acute rheumatic fever incidence**

Country	Period	Age range	Incidence <sup>(a)</sup>	Source
New Zealand (all population groups)	1995–00	5–14	14	Thornley et al. (2001)
Samoan children in Hawaii	1987	0–19	206	Reported in Franks (2002)
Maori children in Auckland	1988	5–15	125	Reported in Franks (2002)
Pacific Islander children in Auckland	1988	5–15	114	Reported in Franks (2002)
<i>Australian data – Aboriginal and Torres Strait Islander children</i>				
Top End of the Northern Territory	1998–02	5–14	245	Top End Rheumatic Heart Disease Register
Central Australia	1998–02	5–14	351	Central Australian Rheumatic Heart Disease Register
Kimberley region in Western Australia	1988–92	5–14	375	Richmond & Harris (1998)
Far North Queensland	1995	5–14	161	Streeton & Hanna (1995)

(a) Annual incidence rates per 100,000 population.

In the United States, acute rheumatic fever is rare. It may also be under-reported because of a lack of awareness of the disease among younger doctors (Wolfe 2000). Despite the low number of new cases, acute rheumatic fever and rheumatic heart disease remain prevalent in the community. In 2001, they were responsible for the deaths of 3,489 Americans (70% of these were female and 30% male). In 2001, the death rates per 100,000 population for white males were 0.9, for black males 0.7, for white females



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1.5 and for black females 1.3 (AHA 2003). These rates are similar to the death rates among the total Australian population (less than two per 100,000 in 2002) but are considerably lower than the death rates among Aboriginal and Torres Strait Islander peoples, who are up to 20 times as likely to die from acute rheumatic fever and rheumatic heart disease as other Australians.

Australian data presented here and elsewhere show much higher incidence rates for new and recurrent episodes of acute rheumatic fever and prevalence rates of rheumatic heart disease among Indigenous Australians than those reported for other countries. The Top End register indicated an annual incidence rate of acute rheumatic fever of 245 per 100,000 among 5–14 year olds between 1998 and 2002, and the Central Australia register a rate of 351 per 100,000 over the same period. Similarly high results have been published for the Kimberley region in Western Australia where the incidence was 375 per 100,000 among Aboriginal and Torres Strait Islander children aged 5–14 years between 1988 and 1992 (Richmond & Harris 1998) with no evidence of any downward trend throughout the 1990s (Ramsay 2000). In Far North Queensland, Streeton & Hanna (1995) reported an annual incidence of acute rheumatic fever of 161 per 100,000 Aboriginal and Torres Strait Islander children aged 5–14 years.

### Discussion

The data from regional rheumatic heart disease registers presented in this bulletin confirm the very high incidence and prevalence of acute rheumatic fever and rheumatic heart disease among the Indigenous Australian population compared with other Australians and other regions of the world. However, ascertaining trends is more problematic.

Early indications are that the trend in incidence of acute rheumatic fever among 5–14 year olds has been fairly stable in recent years in the Top End of the Northern Territory, but that it is increasing in Central Australia. However, the relatively small number of cases compared with other diseases, difficulties in diagnosis, substantial changes in Indigenous Australian population numbers over time, and yearly fluctuations in incidence make it difficult to confirm a trend in the short term. In addition, the relatively recent establishment of the Central Australia register will identify residual cases of both diseases thus artificially inflating incidence in the early years.

National hospital data presented here reflect the disparity in the occurrence of acute rheumatic fever and rheumatic heart disease between the Indigenous Australian population and other Australians, with Indigenous Australian people accounting for 15% of all hospitalisations for these conditions in 2001–02. In addition, hospitalisation rates among Indigenous Australian males are six times as high, and among Indigenous Australian females are eight times as high, as the rates among other Australians. However, since these diseases among the total population are rare, one might expect the Indigenous Australian hospitalisation rates compared with those for other Australians to be even higher. The lower than expected differences between the rates may be due to poorer identification of Indigenous Australians in hospital records, lower than desirable rates of hospitalisation among Indigenous Australian people and/or hospitalisation for rheumatic heart disease among older non-Indigenous Australians who had acute rheumatic fever as children.

The number of heart valve surgery procedures also reflects the impact of acute rheumatic fever and rheumatic heart disease on the Indigenous Australian population, with nearly half of Indigenous Australian people undergoing surgery for these diseases aged less than 25 years compared with just 4% of other Australians of a similar age.

Unfortunately the data do not indicate the procedures performed on individual patients or the severity of rheumatic heart disease at the time of hospitalisation. The data do show, however, that when Indigenous Australian people are hospitalised for rheumatic heart disease they are less likely to undergo heart valve procedures (about one procedure for every three people hospitalised) compared with other Australians being hospitalised for this condition (about one in two). Coupled with the significantly younger average age at death in the Northern Territory due to acute rheumatic fever or rheumatic heart disease (36 years among Indigenous Australian people compared with 67 years among other Australians in 1996 and 1997) (Carapetis & Currie 1999), this suggests that there may be scope for improved treatment for Indigenous Australian people with these diseases, but further investigation is required.

As identification of Indigenous status in both hospital records and on death certificates has only recently begun to improve, national trends in morbidity and mortality from rheumatic heart disease among the Indigenous Australian community cannot be ascertained. Based on the register data, there has been an apparent rise in the reported prevalence of rheumatic heart disease in the Top End of the Northern Territory, but this is likely to be attributable to improved reporting and better awareness of the condition. The fall in the death rate for rheumatic heart disease among the total Australian population in the past 20 years also masks the ongoing problem in the Indigenous Australian population.

To conclude, in addressing the purpose of this bulletin—to describe patterns of acute rheumatic fever and rheumatic heart disease in Australia today—the importance of register data in estimating the incidence and prevalence of these diseases is clear. However, gaps remain in these data. There are regions in Australia that are known to have a high incidence of these diseases but are not covered by the registers. Without similar quality register-based data from these areas it is difficult to extrapolate regional incidence and prevalence rates to national rates, although national hospital and mortality data highlight the extent of the problem, particularly among Indigenous Australians.



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## **Abbreviations**

AHA	American Heart Association
AIHW	Australian Institute of Health and Welfare
CARHDSC	The Central Australian Rheumatic Heart Disease Steering Committee
DHA	Australian Government Department of Health and Ageing
ICD-9	International Classification of Diseases, 9th revision
ICD-10	International Classification of Diseases, 10th revision
ICD-10-AM	International Classification of Diseases, 10th revision Australian Modification
SMR	Standardised Mortality Ratio
U.S. NLOM & NIH	United States National Library of Medicine & National Institutes of Health
WHO	World Health Organization

## Methods

### Incidence and prevalence

In this bulletin, incidence includes the number of new and recurrent cases of acute rheumatic fever over a given period of time, and prevalence refers to the number of cases of rheumatic heart disease present in the population at a given time. Incidence and prevalence data have been obtained from the Top End and Central Australian Rheumatic Heart Disease Registers.

### Rates

#### Age-specific rates

Age-specific rates were calculated by dividing the number of events (in this case incidence and prevalence) occurring in each specified age group by the mid-year estimated resident population for the corresponding age group.

#### Age-standardised rates

Age-standardised rates for hospitalisations and deaths were used to remove the influence of age when comparing populations with different age structures. This was done by applying age-specific rates to a standard population. The 2001 Australian population has been used as the standard population in these analyses.

#### *Direct age standardisation*

Direct age standardisation is the most common method of age standardisation, and is used in this bulletin for hospitalisation and death data. This method is generally used when the populations under study are large and the age-specific rates are reliable.

#### *Indirect age standardization and the standardised mortality ratio*

Due to the small number of deaths from acute rheumatic fever and rheumatic heart disease creating uncertainty about the stability of age-specific rates, indirect age standardisation has also been used. This effectively removes the influence of the age structure, but does not provide a measure of prevalence in terms of a rate. Rather, the summary measure is a comparison of the number of observed cases compared with the number expected if the age-specific prevalence rates of the standard population are applied to the study population.

An SMR of one indicates the same number of observed cases that were expected (suggesting rates between populations are similar). A result greater than one indicates more cases than expected. A result less than one indicates fewer cases than expected. For example, if there are ten times as many deaths as expected then the rate of deaths can be assumed to be ten times that of the comparison population.

In this bulletin, the indirect method has been used for comparing death rates between Indigenous Australians and other Australians.



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## Data sources and data quality issues

### Data sources

#### The Top End and Central Australian Rheumatic Heart Disease Registers

Both registers are integral to the Top End and Central Australian Rheumatic Heart Disease Programs and have been developed in keeping with WHO recommendations (WHO 1995) for regions with high incidence of rheumatic heart disease. Both registers include data related to diagnosis, hospitalisations, compliance with preventive penicillin, clinical progress, surgery, and mortality.

The Top End program became fully operational in June 1998 and the Central Australian register in June 2002. Confidentialised data from the registers were provided to AIHW for inclusion in this bulletin. Estimates of incidence from these Central Australian register, exclude cases from South Australia and Western Australia because of difficulties ascertaining denominator populations.

#### AIHW National Hospital Morbidity Database

This database is held at the AIHW and contains demographic, diagnostic, procedural and length of stay information on episodes of care for patients admitted to hospital. The data are supplied to the AIHW by the state and territory health authorities. The database can be used to determine the number of hospitalisations for a particular condition or procedure. It is not possible to count patients individually.

#### AIHW National Mortality Database

This database is held at the AIHW and contains information on the cause of death as supplied by the medical practitioner certifying the death or by a coroner. Registration of deaths is the responsibility of the state and territory Registrars of Births, Deaths and Marriages. Registrars provide the information recorded on the death certificates to the Australian Bureau of Statistics for coding of cause of death and compilation into aggregate statistics.

#### Identification of Indigenous status

The hospitalisation and death rates for the Indigenous Australian population obtained from the National Hospital Morbidity Database and the National Mortality Database are likely to be underestimates because identification of Indigenous status in hospital data collections and on death certificates is in need of improvement.

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