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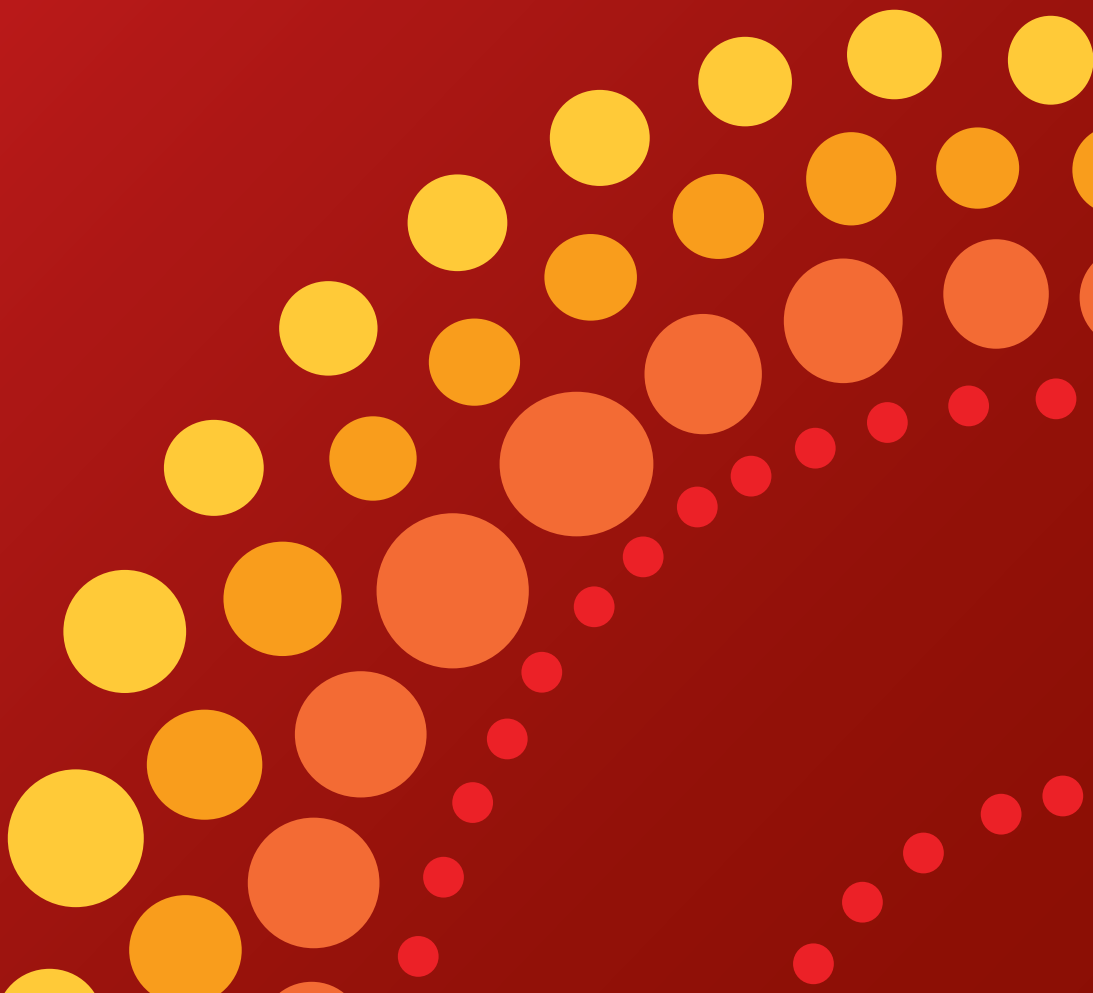
The Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition)

Quick reference guides



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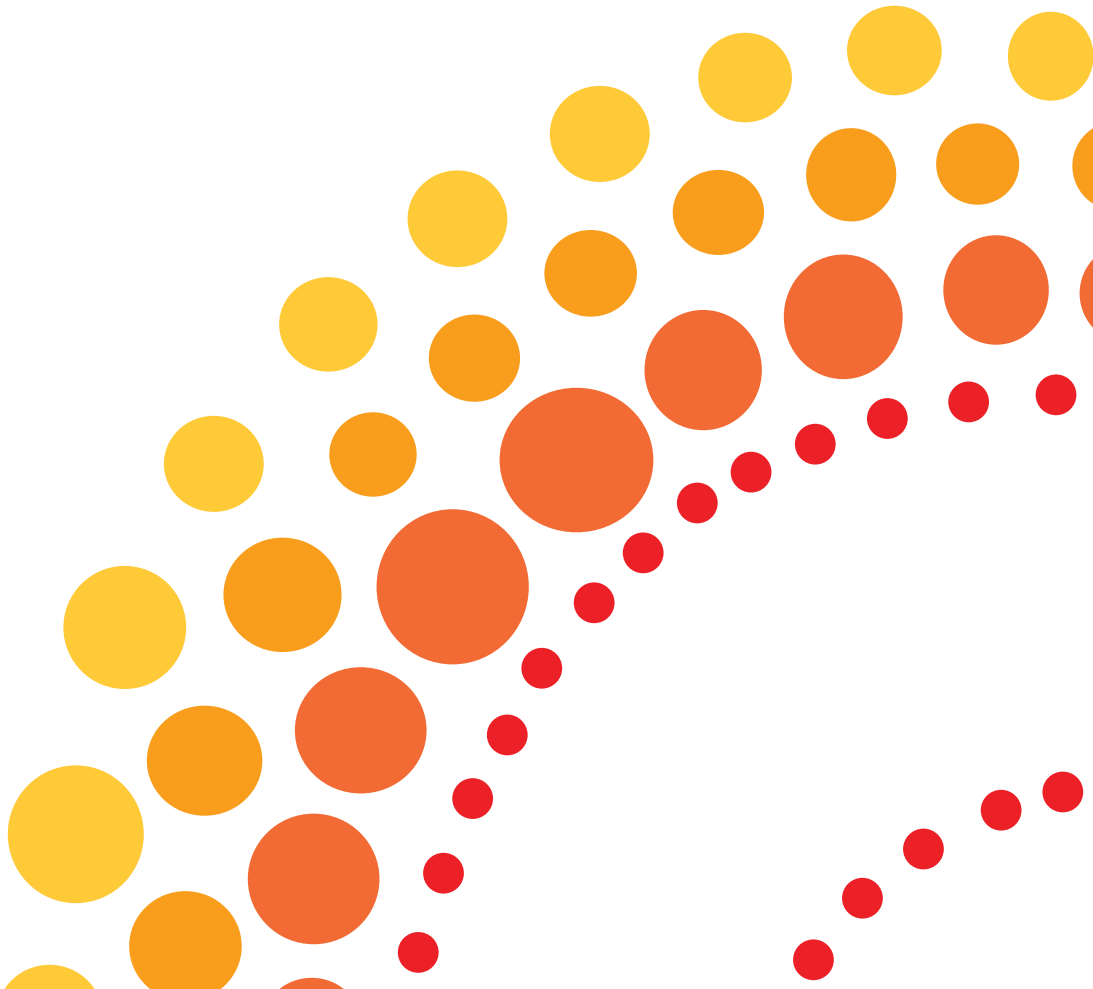

Contents

1. Primary prevention of acute rheumatic fever

The purpose of primary prevention is to limit the incidence of disease by controlling causes and risk factors. Primary prevention can either focus on an entire population or on individuals within that population who are at elevated risk (e.g. people with GAS infection).

This quick reference guide is derived from the *Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease* (2nd edn).

What is acute rheumatic fever?

Acute rheumatic fever (ARF) is an illness caused by a reaction to a bacterial infection with group A streptococcus (GAS). It causes an acute, generalised inflammatory response and an illness that targets specific parts of the body, including the heart, joints, brain and skin. Individuals with ARF are often unwell, have significant joint pain and require hospitalisation. Despite the dramatic nature of the acute episode, ARF typically leaves no lasting damage to the brain, joints or skin, but can cause persisting heart damage, termed 'rheumatic heart disease' (RHD).

Primary prevention of ARF

When an individual is exposed to GAS, the organism attaches to and colonises the pharyngeal mucosa. A process of infection incorporating an immune response is initiated, and an episode of ARF may occur 2–3 weeks later. The aim of primary prevention is to identify symptomatic GAS pharyngitis in those individuals most at risk of ARF (typically children aged 5–14 years), and eradicate the bacterium with antibiotic treatment before the immune response associated with ARF has been initiated. Studies show that ARF associated with GAS pharyngitis can be prevented if treatment is commenced within 9 days of symptoms appearing. While the association between GAS pharyngitis and ARF is well described, the role of GAS-associated skin infection remains unclear.

Antibiotic treatment of sore throats

The management of pharyngitis as a mechanism for preventing ARF is complicated by the fact that only a minority of sore throats are caused by GAS. While it is possible to treat all cases of pharyngitis with antibiotics, this exposes a significant proportion of patients to unnecessary treatment, as only 20–40% of pharyngitis episodes are associated with GAS infection; the remainder are caused by viruses or by bacteria for which antibiotic treatment is not recommended. Some treatment guidelines do suggest that people identified as being from populations at high risk of ARF (e.g. Aboriginal people and/or Torres Strait Islanders), or who have established RHD, but are not currently receiving secondary antibiotic prophylaxis, should be treated with antibiotics

if they develop pharyngitis, irrespective of other clinical features, and before confirmatory testing for GAS is available.

Targeting only those people with confirmed GAS pharyngitis is an alternate strategy. If such an approach is taken, then the rapid identification of GAS in people presenting with pharyngitis is necessary to ensure that treatment is commenced within 9 days of symptom onset. While rapid diagnosis may rely on clinical features or antigen detection, the utility of these techniques in confirming or excluding GAS as the cause of pharyngitis is variable. Bacterial culture of a throat swab remains the gold standard, but is associated with an inherent delay in diagnosis, and thus treatment.

Recommended antibiotic treatment for streptococcal pharyngitis

All cases			
BPG	**Child:** *Weight (kg)* — *Dose (mg)* ≥20 — 900 15 to <20 — 675 10 to <15 — 450 6 to <10 — 337.5 3 to <6 — 225 **Adult:** 900 mg	Deep im injection	Once
If im injection not possible			
Phenoxymethylpenicillin	**Child:** 10 mg/kg up to 500 mg, bd **Adult:** 500 mg, bd	Oral	For 10 days
For patients hypersensitive to penicillin			
Erythromycin ethyl succinate	Child: 20 mg/kg up to 800 mg, bd Adult: 800 mg, bd	Oral	For 10 days

In cases of severe sore throat, procaine penicillin may be required. Refer to CARPA Manual for further information.

bd, bis die (twice daily); BPG, benzathine penicillin G; im, intramuscular injection.

Source: *CARPA standard treatment manual,* 5th ed. Rural and Remote Health 2011.

2. Diagnosis of acute rheumatic fever

An accurate diagnosis of ARF is important. Overdiagnosis results in unnecessary treatment over a long time, while underdiagnosis leads to further attacks of ARF, cardiac damage and premature death. Diagnosis remains a clinical decision, as there is no specific laboratory test.

This quick reference guide is derived from the *Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease* (2nd edn).

What is acute rheumatic fever?

Acute rheumatic fever (ARF) is an illness caused by a reaction to a bacterial infection with group A streptococcus (GAS). It causes an acute, generalised inflammatory response and an illness that targets specific parts of the body, including the heart, joints, brain and skin. Individuals with ARF are often unwell, have significant joint pain and require hospitalisation. Despite the dramatic nature of the acute episode, ARF leaves no lasting damage to the brain, joints or skin, but can cause persisting heart damage, termed 'rheumatic heart disease' (RHD). Recurrences of ARF may cause further cardiac valve damage. Hence, RHD steadily worsens in people who have multiple episodes of ARF.

Who gets ARF?

Although ARF is relatively rare in industrialised countries, in Australia it is a significant illness among Aboriginal people and Torres Strait Islanders, particularly across central and northern Australia.

Problems with diagnosis and management

Several factors contribute to the barriers in diagnosis and management of ARF and RHD in Australia:

- although strategies for preventing RHD have been proven to be simple, cheap and cost-effective, they must be adequately implemented in populations at highest risk of the disease
- because ARF is rare in most metropolitan centres, the majority of clinicians will have seen very few, if any, cases of ARF

- there is great variability in the management of these diseases, with minimal training and experience in the management of ARF and RHD occasionally resulting in inappropriate management
- access to healthcare services by population groups experiencing the highest rates of ARF and RHD is often limited.

Identifying high-risk groups

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.

ARF is predominantly a condition seen in children aged 5–14 years, although people can have recurrent episodes well into their 40s.

Comprehensive data are not available for other populations, but Aboriginal people and Torres Strait Islanders living in urban settings, and potentially immigrants from developing countries, may also be at high risk.

Diagnostic criteria for ARF

An accurate diagnosis of ARF is important, as:

- overdiagnosis will result in the individual receiving treatment unnecessarily
- underdiagnosis may lead to further episodes of ARF, cardiac damage and the need for heart valve surgery and/or premature death.

Currently, there is no diagnostic laboratory test for ARF, so diagnosis remains a clinical decision based on the identification of major and minor manifestations of the illness. The Table below outlines criteria for high- and low-risk populations in Australia.

2012 Updated Australian guidelines for the diagnosis of ARF

	High-risk groups[†]	All other groups
Definite initial episode of ARF	2 major **or** 1 major and 2 minor manifestations **plus** evidence of a preceding GAS infection[‡]	
Definite recurrent episode of ARF in a patient with known past ARF or RHD	2 major **or** 1 major and 1 minor **or** 3 minor manifestations **plus** evidence of a preceding GAS infection[‡]	
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 cases should be further categorised according to the level of confidence with which the diagnosis is made: • highly-suspected ARF • uncertain ARF	
Major manifestations	Carditis (including subclinical evidence of rheumatic valvulitis on echocardiogram) Polyarthritis[††] or aseptic mono-arthritis or polyarthralgia Chorea[§] Erythema marginatum[¶] Subcutaneous nodules	Carditis (excluding subclinical evidence of rheumatic valvulitis on echocardiogram) Polyarthritis[††] Chorea[§] Erythema marginatum[¶] Subcutaneous nodules
Minor manifestations	Monoarthralgia Fever[‡‡] ESR ≥30 mm/h or CRP ≥30 mg/L Prolonged P-R interval on ECG[§§]	Fever[‡‡] Polyarthralgia or aseptic mono-arthritis ESR ≥30 mm/h or CRP ≥30 mg/L Prolonged P-R interval on ECG[§§]

[†]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 people and Torres Strait Islanders living in urban settings, Maoris and Pacific Islanders, and potentially immigrants from developing countries, may also be at high risk. [‡]Elevated or rising antistreptolysin O or other streptococcal antibody, or a positive throat culture or rapid antigen test for GAS. [††]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. [§]Chorea does not require other manifestations or evidence of preceding GAS infection, provided other causes of chorea are excluded. [¶]Care should be taken not to label other rashes, particularly non-specific viral exanthemas, as erythema marginatum. [‡‡]Oral, tympanic or rectal temperature ≥38°C on admission, or a reliably reported fever documented during the current illness. [§§]If carditis is present as a major manifestation, a prolonged P-R interval cannot be considered an additional minor manifestation.

CRP, C-reactive protein; ECG, electrocardiogram; ESR, erythrocyte sedimentation rate.

Evidence of preceding group A streptococcal infection

All suspected cases of ARF (except those with chorea or low-grade subacute carditis) should have elevated serum streptococcal serology demonstrated.

If the initial titre is below the upper limit of normal (ULN) for age, repeat testing after 10–14 days.

In the absence of local data, it is recommended that ULN values in the following Table be used.

Upper limit of normal for serum streptococcal antibody titres

Age group	ULN (U/mL)	
(years)	ASO titre	Anti-DNase B titre
1-4	170	366
5-14	276	499
15-24	238	473
25-34	177	390
≥35	127	265

Anti-DNase, antideoxyribonuclease B; ASO, antistreptolysin O; ULN, upper limit of normal.

Manifestations of ARF

Major manifestations	
Carditis	Usually presents clinically as an apical holosystolic murmur, with or without a mid-diastolic flow murmur, or an early diastolic murmur at the base of the heart or left sternal edge
Polyarthritis	Extremely painful, affecting the large joints, especially the ankles and knees, is usually asymmetrical and migratory, but can be additive Usually responds within 3 days of starting NSAID therapy
Aseptic mono-arthritis or polyarthralgia	A major manifestation in high-risk group but a minor manifestation in other groups. Arthralgia that is migratory and asymmetrical, affecting large joints, is most indicative of ARF
Sydenham's chorea	Consists of jerky, uncoordinated movements, especially affecting the hands, feet, tongue and face, disappears during sleep Echocardiography is essential for all patients with chorea
Erythema marginatum	Extremely rare, as well as difficult to detect in Aboriginal people, but highly specific for ARF Occurs as circular patterns of bright pink macules or papules on the trunk and proximal extremities
Subcutaneous nodules	Rare, but-highly specific manifestations of ARF and strongly associated with carditis Present as crops of small, round, painless nodules over the elbows, wrists, knees, ankles, Achilles tendon, occiput and posterior spinal processes of vertebrae
Minor manifestations	
Fever	Most manifestations of ARF are accompanied by fever A reported recent history of fever or presenting fever of ≥38°C
Elevated acute phase reactants	Serum CRP level of ≥30 mg/L or ESR of ≥30 mm/h meets this diagnostic criterion
Prolonged P-R interval	If a prolonged P-R interval is detected, ECG should be repeated after 1–2 months. If it has returned to normal, ARF becomes a more likely diagnosis

CRP, C-reactive protein; ECG, electrocardiogram; ESR, erythrocyte sedimentation rate; NSAID, non-steroidal anti-inflammatory drug.

Differential diagnoses of common major presentations of ARF

The diagnosis of ARF is based on the assumption that other likely diagnoses have been excluded.

Many of the clinical features of ARF are non-specific, so a wide range of differential diagnoses should be considered.

Post-streptococcal syndromes such as PANDAS and post-streptococcal reactive arthritis may be confused with ARF, and these diagnoses should rarely, if ever, be made in high-risk populations.

Upper limits of normal of P-R interval

Age group (years)	Sec
3–12	0.16
12–16	0.18
17+	0.20

Source: Adapted from Park MK, *Pediatric cardiology for practitioners*, 2nd edn. Chicago: Year Book Medical; 1998.

Presentation		
Polyarthritis and fever	**Carditis**	**Chorea**
Septic arthritis (including disseminated gonococcal infection)†	Innocent murmur	Systemic lupus erythematosus
Connective tissue and other autoimmune disease††	Mitral valve prolapse	Drug intoxication
Viral arthropathy¥	Congenital heart disease	Wilson's disease
Reactive arthropathy¥	Infective endocarditis	Tic disorder‡
Lyme disease≠	Hypertrophic cardiomyopathy	Choreoathetoid cerebral palsy
Sickle cell anaemia	Myocarditis: viral or idiopathic	Encephalitis
Infective endocarditis	Pericarditis: viral or idiopathic	Familial chorea (including Huntington's)
Leukaemia or lymphoma		Intracranial tumour
Gout and pseudogout		Lyme disease≠
		Hormonal§

†Gonorrhoea should be actively sought in all sexually-active cases. Tests for gonorrhoea include polymerase chain reaction (PCR) of joint aspirate, endocervical PCR (gonococcal and chlamydia) and microscopy, culture and sensitivity, or urine/self-collected vaginal swabs in cases where endocervical PCR is not possible. ††Includes rheumatoid arthritis, juvenile chronic arthritis, inflammatory bowel disease, systemic lupus erythematosus, systemic vasculitis and sarcoidosis. ¥Mycoplasma, cytomegalovirus, Epstein–Barr virus, parvovirus, hepatitis, rubella vaccination, and *Yersinia* spp and other gastrointestinal pathogens. ≠Lyme disease has not been confirmed in Australia or New Zealand. ‡Possibly including PANDAS (paediatric autoimmune neuropsychiatric disorder associated with streptococcal infection). §Includes oral contraceptives, pregnancy (chorea gravidarum), hyperthyroidism and hypoparathyroidism.

Investigations in suspected ARF

All patients with suspected or confirmed ARF should undergo echocardiography to confirm or refute the diagnosis of rheumatic carditis.

Other investigations are listed below.

Recommended for all cases

White blood cell count

Erythrocyte sedimentation rate (ESR)

C-reactive protein (CRP)

Blood cultures, if febrile

Electrocardiogram (if prolonged P-R interval or other rhythm abnormality, repeat in 2 weeks and again at 2 months, if still abnormal

Chest X-ray, if clinical or echocardiographic evidence of carditis

Echocardiogram (consider repeating after 1 month, if negative)

Throat swab (preferably before giving antibiotics): culture for group A streptococcus

Antistreptococcal serology: both ASO and anti-DNase B titres, if available (repeat 10–14 days later if first test not confirmatory)

Tests for alternative diagnoses, depending on clinical features

Repeated blood cultures, if possible endocarditis

Joint aspirate (microscopy and culture) for possible septic arthritis

Copper, ceruloplasmin, antinuclear antibody, drug screen for choreiform movements

Serology and autoimmune markers for arboviral, autoimmune or reactive arthritis

3. Management of acute rheumatic fever

All patients with suspected ARF (first episode or recurrence) should be hospitalised as soon as possible after the onset of symptoms. This ensures that all investigations are performed, and if necessary, the patient should be observed to confirm the diagnosis before commencing treatment.

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Who gets ARF?

Although ARF is relatively rare in industrialised countries, in Australia it is a significant illness among Aboriginal people and Torres Strait Islanders, particularly across northern and central Australia. Pacific Islanders, and migrants from countries with a high prevalence of RHD, are also known to be at high risk.

Problems with diagnosis and management

Several factors contribute to the barriers in diagnosis and management of ARF and RHD in Australia:

- although strategies for preventing RHD have been proven to be simple, cheap and cost-effective, they must be adequately implemented in populations at highest risk of the disease
- because ARF is rare in most metropolitan centres, the majority of clinicians will have seen very few, if any, cases of ARF
- there is variability in the management of these diseases, with minimal training and experience in the management of ARF and RHD, occasionally resulting in inappropriate management
- access to healthcare services by population groups experiencing the highest rates of ARF and RHD is often limited.

Confirming the diagnosis

Diagnostic criteria, recommended investigations and detailed information on differential diagnoses are given in the quick reference guide *Diagnosis of acute rheumatic fever.*

As the arthritis, arthralgia and fever of ARF respond to non-steroidal anti-inflammatory drugs (NSAIDs), which may prevent the full clinical manifestations becoming apparent, it is recommended that joint pain be treated with paracetamol or codeine until the diagnosis is confirmed.

There is convincing evidence that subclinical or silent rheumatic valve damage detected by echocardiography is part of the spectrum of rheumatic carditis, and should not be ignored.

Guidelines for general in-hospital care

All patients with suspected ARF (first episode or recurrence) should be discussed immediately with a paediatrician or adult physician expert in the diagnosis and management of ARF, admitted to hospital as soon as possible after the onset of symptoms, and the steps initiated to confirm the diagnosis.

While in hospital, the patient should be registered in centralised and local ARF/RHD registers.

Occasionally, when the diagnosis has already been confirmed and the patient is not unwell (e.g. mild recurrent chorea in a child with no other symptoms or signs), outpatient management may be appropriate, but only after consultation with a specialist.

Treatment

All cases

Single-dose im benzathine penicillin G (preferable) or 10 days of oral penicillin V (iv not needed; oral erythromycin if allergic to penicillin)

Arthritis and fever

Paracetamol (first line) or codeine until diagnosis confirmed

Aspirin, naproxen or ibuprofen, once diagnosis confirmed, if arthritis or severe arthralgia present

Mild arthralgia and fever may respond to paracetamol alone

Influenza vaccine for children receiving aspirin during the influenza season (autumn/winter)

Chorea

No treatment for most cases

Carbamazepine or valproic acid, if treatment necessary

Carditis/heart failure

Bed rest, with mobilisation as symptoms permit

Urgent echocardiogram

Antifailure medication:

- Diuretics/fluid restriction for heart failure
- ACE inhibitors for more severe failure, particularly if aortic regurgitation is present; glucocorticoids optional for severe carditis (in central and northern Australia, prophylaxis for opportunistic infections may be needed for those on prolonged steroid courses. Seek specialist advice)
- Digoxin, beta-blocker or electrical cardioversion if atrial fibrillation present

Valve surgery for life-threatening acute carditis (rare)

Nursing recordings
Temperature, pulse, respiratory rate, blood pressure 4 times daily
Sleeping pulse (e.g. 0200 hours)
If pulse >100, apical heart rate
Diet
Free fluids (if no heart failure)
Normal diet (limit extras)
Early diet advice, if overweight and in failure, to avoid further weight gain
Weekly weight
Bed rest and general care
Strict bed rest not necessary for most patients
Plan care to provide rest periods
Provide age-appropriate activities
Notify school teacher
Involve family in care
Prepare for discharge to primary care facility, and follow up
If clinical carditis present (heart murmur, heart failure, pericardial effusion, valvular damage)
Document cardiac symptoms and signs
Daily weight and fluid balance chart
Diuretics, ACE inhibitors, digoxin, if indicated; consider glucocorticoids
Anticoaglulation if atrial fibrillation present
Cardiology opinion

ACE, angiotensin-converting enzyme; im, intramuscular; iv, intravenous.

Medication used in ARF

Medication	Indication	Regimen	Duration
BPG, im	Treat streptococcal infection	900 mg (1,200,000 U) ≥20 kg 450 mg (600,000 U) <20 kg	Single dose
or Penicillin V, po	Initial treatment of streptococcal infection	**Child:** 250 mg, bd **Adolescent and Adult:** 500 mg, bd	10 days
or Erythromycin ethyl succinate, po (only if allergic to penicillin)	Initial treatment of streptococcal infection	**Child:** 20 mg/kg up to 800 mg, bd **Adult:** 800 mg, bd	10 days
or Erythromycin, po	Initial treatment of streptococcal infection	**Child:** 12.5 mg/kg up to 500 mg, bd **Adult:** 500 mg, bd	10 days
Paracetamol, po	Arthritis or arthralgia: mild or until diagnosis confirmed	60 mg/kg/day (max 4 g) given in 4–6 doses/day; may increase to 90 mg/kg/day, if needed, under medical supervision	Until symptoms relieved or NSAID started
Codeine, po	Arthritis or until diagnosis Confirmed arthralgia	0.5–1 mg/kg/dose (adults 15–60 mg/dose) 4–6 hourly	Until symptoms relieved or NSAID started
Aspirin, po	Arthritis or severe arthralgia (when ARF diagnosis confirmed)	Begin with 50-60 mg/kg/day, increasing, if needed, up to 80–100 mg/kg/day (4–8 g/day in adults) given in 4–5 doses/day If higher doses required, reduce to 50–60 mg/kg/day when symptoms improve, and cease when symptom free for 1–2 weeks Consider ceasing in the presence of acute viral illness, and consider influenza vaccine if administered during autumn/winter	Until joint symptoms relieved
Naproxen, po	Arthritis or severe arthralgia (when ARF diagnosis confirmed)	10–20 mg/kg/day (max 1250 mg) given, bd	As for aspirin
Ibuprofen, po	Arthritis or severe arthralgia (when ARF diagnosis confirmed)	30 mg/kg/day (max 1600 mg) given tds	As for aspirin
Prednisone or prednisolone, po.	Severe carditis, heart failure, pericarditis with effusion	1–2 mg/kg/day (max 80 mg); if used >1 week, taper by 20–25% per week	Usually 1–3 weeks

Medication	Indication	Regimen	Duration
Frusemide, po/iv (can also be given im)	Heart failure	**Child:** 1–2 mg/kg stat, then 0.5–1 mg/kg/dose 6–24 hourly (max 6 mg/kg/day) **Adult:** 20–40 mg/dose, 6–24 hourly, up to 250–500 mg/day	Until failure controlled and carditis improved
Spironolactone, po	Heart failure	1–3 mg/kg/day (max 100–200 mg/day) in 1–3 doses; round dose to multiple of 6.25 mg (1/4 of a tablet)	As for frusemide
Enalapril, po	Heart failure	**Child:** 0.1 mg/kg/day in 1–2 doses, increased gradually over 2 weeks to a max of 1 mg/kg/day in 1–2 doses **Adult:** initial dose 2.5 mg daily; maintenance dose 10–20 mg daily (max 40 mg)	As for frusemide
Captopril, po	Heart failure	**Child:** initial dose 0.1 mg/kg/dose. Beware of hypotension. Increase gradually over 2 weeks to 0.5–1 mg/kg/doses 8 hourly (max 2 mg/kg/dose 8 hourly). **Adult:** initial dose 2.5–5 mg. Maintenance dose 25–50 mg 8 hourly	As for frusemide
Lisinopril, po	Heart failure	**Child:** 0.1–0.2 mg/kg once daily, up to 1 mg/kg/dose **Adult:** 2.5–20 mg once daily (max 40 mg/day)	As for frusemide
Digoxin, po/iv	Heart failure/ atrial fibrillation	**Child:** 15 mcg/kg start, and then 5 mcg/kg after 6 hours, then 3–5 mcg/kg/dose (max 125 mcg) 12 hourly **Adult:** 125–250 mcg daily Check serum levels	Seek advice from specialist
Carbamazepine	Severe chorea	7–20 mg/kg/day (7–10 mg/kg day usually sufficient) given tds	Until chorea controlled for several weeks, then trial off medication
Valproic acid, po	Severe chorea (may affect salicylate metabolism)	Usually 15–20 mg/kg/day (can increase to 30 mg/kg/day) given tds	As for carbamazepine

bd, *bis die* (twice daily); BPG, benzathine penicillin G; im, intramuscular; iv, intravenous; NSAID, non-steroidal anti-inflammatory drug; po, per oral; tds, *ter die sumendum* (three times daily).

Commencing long-term preventive measures

Secondary prevention

As outlined in the previous Table, penicillin is given in cases of ARF to ensure eradication of streptococci that may persist in the upper respiratory tract. As this could be considered the commencement of secondary prophylaxis, it may be advisable to use im benzathine penicillin G.

Some clinicians prefer to use oral penicillin while patients are in hospital, and to defer the im injection until there has been improvement, and patients and their families have been properly counselled about secondary prophylaxis.

Patients with a reliably-documented penicillin allergy may be treated with oral erythromycin. However, most patients labelled as being allergic to penicillin are not. It is recommended that patients with a stated penicillin allergy be investigated carefully, preferably with the help of an allergist, before being accepted as truly allergic.

Patients with probable ARF may be managed in two ways, according to the level of confidence with which the diagnosis is made:

- highly-suspected ARF: manage as for definite ARF
- uncertain ARF: in patients from high-risk groups, administer 12 months of secondary prophylaxis initially, and reassess (including echocardiography) at that time. If no evidence of recurrent ARF, and no evidence of cardiac valvular damage on echocardiography, consider ceasing secondary prophylaxis. In such cases, the residual uncertainty should be discussed with the patient, and they should be encouraged to be particularly vigilant about the treatment of sore throats, prevention and treatment of skin sores and early presentation with any symptoms of potentially recurrent ARF.

Secondary prevention is discussed in greater detail in the quick reference guide *Secondary prevention of acute rheumatic fever.*

Advice on discharge

- All patients should have a good understanding of the cause of ARF and the need to have sore throats and skin sores treated early. Family members should be informed that they are at increased risk of ARF compared to the wider community.
- Patients and families should understand the reason for long-term secondary prophylaxis, and the consequences of not receiving all recommended treatment. They should be given clear information about where to go for secondary prophylaxis, and written information on appointments for follow up with their local medical practitioner, physician/paediatrician and cardiologist (if needed).
- If there is cardiac valve damage, patients and families should be reminded of the importance of antibiotic prophylaxis for dental and other procedures to protect against endocarditis.

Contact local health staff for follow up

- The notifying medical practitioner should make direct contact with community medical staff, so that they are aware of the diagnosis, the need for secondary prophylaxis and any other specific follow up requirements.

4. Secondary prevention of acute rheumatic fever

Secondary prevention of further episodes of ARF is a priority. Secondary prophylaxis with regular benzathine penicillin G (BPG) is the only RHD control strategy shown to be effective and cost-effective at both community and population levels.

This quick reference guide is derived from the *Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease* (2nd edn).

What is acute rheumatic fever?

Acute rheumatic fever (ARF) is an illness caused by a reaction to a bacterial infection with group A streptococcus (GAS). It causes an acute, generalised inflammatory response and an illness that targets specific parts of the body, including the heart, joints, brain and skin. Individuals with ARF are often unwell, have significant joint pain and require hospitalisation. Despite the dramatic nature of the acute episode, ARF typically leaves no lasting damage to the brain, joints or skin, but can cause persisting heart damage, termed 'rheumatic heart disease' (RHD). People who have had ARF previously are much more likely than the wider community to have subsequent episodes. Recurrences of ARF may cause further cardiac valve damage. Hence, RHD steadily worsens in people who have multiple episodes of ARF.

Who gets ARF?

Although ARF is relatively rare in industrialised countries, in Australia it is a significant illness among Aboriginal people and Torres Strait Islanders, particularly across central and northern Australia. Pacific Islanders, and migrants from countries with a high prevalence of RHD, are also known to be at high risk.

Secondary prevention

Secondary prevention of further episodes of ARF is a priority. Secondary prophylaxis with regular benzathine penicillin G (BPG) is the only RHD control strategy shown to be effective and cost-effective at both community and population levels.

The appropriate duration of secondary prophylaxis is determined by age, time since the last episode of ARF and potential harm from recurrent ARF, but is likely to be 10 years or more.

While secondary prophylaxis is a proven strategy for controlling RHD, and is also simple, cheap and cost-effective, it must be adequately implemented. Persistent high rates of recurrent ARF in high-risk populations highlight the continued barriers to secondary prevention.

The effectiveness of secondary prophylaxis is impaired by factors that contribute to poor adherence to antibiotic regimens and increased incidence rates of ARF. These factors relate to overcrowded housing, poor access to health services, limited educational opportunities and poor environmental conditions. Communities with the highest rates of ARF and RHD are often the least equipped to deal with the problem.

Secondary prevention should include:

- strategies aimed at improving the delivery of secondary prophylaxis and patient care
- the provision of education
- coordination of available health services
- advocacy for necessary and appropriate resources.

Antibiotic regimens for secondary prophylaxis

Antibiotic	Dose	Route	Frequency
First line			
BPG	900 mg (1,200,000 U) ≥20 kg 450 mg (600,000 U) < 20 kg	Deep im injection	4 weekly, or 3 weekly for selected groups*
Second line (If im route is not possible or refused, adherence should be carefully monitored)			
Phenoxymethylpenicillin (Penicillin V)	250 mg	Oral	Twice daily
Following documented penicillin allergy			
Erythromycin	250 mg	Oral	Twice daily

*Three-weekly BPG may be considered for patients with moderate or severe carditis or a history of valve surgery, who demonstrate good adherence to less frequent injections, and for those who have confirmed breakthrough ARF, despite full adherence to 4-weekly BPG.

BPG, benzathine penicillin G; im, intramuscular.

Measures that may reduce the pain of BPG injections

- Use a 21-gauge needle
- Warm syringe to room temperature before using
- Allow alcohol from swab to dry before inserting needle
- Apply pressure with thumb for 10 sec before inserting needle
- Deliver injection very slowly (preferably over at least 2–3 mins)
- Distract patient during injection (e.g. with conversation)
- (The addition of 0.5–1.0 mL of 1% lignocaine is used elsewhere, but is not recommended with preloaded syringes currently available in Australia)

Factors that affect the duration of secondary prophylaxis

Factor	Implication
Age	ARF recurrence is less common between 25–40 years of age, and rare >40 years
Presence and severity of RHD	ARF recurrence could be life-threatening in people with moderate or severe RHD, or in those with a history of valve surgery
Presence of carditis during initial episode	Increases the likelihood of further cardiac damage, should a recurrence occur
Time elapsed since last episode of ARF	ARF recurrences are less common >5 years since last episode
Socioeconomic circumstances	ARF recurrences are more common in lower socioeconomic groups (particularly related to overcrowded housing)
Background risk of GAS infection and ARF within the community	ARF recurrences are more common in higher-incidence communities or settings
Adherence to treatment	Optimised adherence for a few years after the initial episode may provide greater protection from recurrences than offered by poor adherence for many years
Assessment at time of cessation of secondary prophylaxis	Evidence of moderate or greater RHD may warrant prolonged prophylaxis

Duration of secondary prophylaxis

Category	Definition of category	Duration
All persons with ARF or RHD†		Minimum 10 years after most recent episode of ARF or until age 21 years (whichever is longer).
Status after initial period elapsed:		
No RHD	No pathological mitral or aortic regurgitation, but may have minor morphological changes to mitral or aortic valves on echocardiography	Discontinue at that time#
Mild RHD	Mild mitral or aortic regurgitation clinically and on echocardiography, with no clinical evidence of heart failure, and no evidence of cardiac chamber enlargement on echocardiography	Discontinue at that time
Moderate RHD	• Any valve lesion of moderate severity clinically (e.g. mild–moderate cardiomegaly and/or mild–moderate heart failure) or on echocardiography • Mild mitral regurgitation, together with mild aortic regurgitation clinically or on echocardiography • Mild or moderate mitral or aortic stenosis • Any pulmonary or tricuspid valve lesion co-existing with a left-sided valve lesion	Continue until 35 years of age
Severe RHD	• Any severe valve lesion clinically (e.g. moderate to severe cardiomegaly or heart failure) or on echocardiography • Any impending or previous cardiac valve surgery for RHD	Continue until age 40 years, or longer*

† Patients >25 years of age who are diagnosed with RHD, without any documented history of prior ARF, should receive prophylaxis until the age of 35 years. At this time, they should be reassessed to determine whether prophylaxis should be continued. # Decisions to cease secondary prophylaxis should be based on clinical and echocardiographic assessment. * Risk of recurrence is extremely low in people aged >40 years. In some cases, for example, when the patient decides that they want to reduce even a minimal risk of recurrence, prophylaxis may be continued beyond the age of 40 years, or even for life.

Improving adherence to secondary prophylaxis

A variety of factors, mainly sociological, combine to limit the efficacy of secondary prophylaxis. A major reason for poor adherence in remote Aboriginal and Torres Strait Islander communities is the availability and acceptability of health services, rather than personal factors, such as injection refusal, pain of injections or a lack of knowledge or understanding of ARF and RHD.

Adherence is improved when patients feel a sense of personalised care and 'belonging' to the clinic, and when recall systems extend beyond the boundaries of the community.

Organisational approaches to secondary prophylaxis (including the use of registers) are outlined in the information sheet *RHD control programs*.

Strategies to promote continuing adherence include:

- routine review and care planning (see below)
- recall and reminder systems
- having local staff members dedicated to secondary prophylaxis and coordinating routine care
- supporting and utilising the expertise, experience, community knowledge and language skills of Aboriginal health workers
- improving staff awareness of the diagnosis and management of ARF and RHD
- taking measures to minimise staff turnover
- implementing measures to reduce the pain of injections.

Procedures requiring endocarditis prophylaxis for patients with RHD

Infective endocarditis is a dangerous complication of RHD including those with prosthetic valves. People with prosthetic valves or established RHD, should receive antibiotic prophylaxis prior to procedures expected to produce bacteraemia (see below).

Dental, oral and respiratory tract procedures	
Dental extractions	Endodontic surgery and instrumentation
Periodontal procedures	Placement of orthodontic bands
Dental implant placement	Intraligamentary local anaesthetic injections
Gingival surgery	Tonsillectomy/adenoidectomy
Initial placement of orthodontic appliances	Rigid bronchoscopy
Surgical drainage of dental abscess	Surgery involving the bronchial mucosa
Maxillary or mandibular osteotomies	Sclerotherapy of oesophageal varices
Surgical repair or fixation of a fractured jaw	Dilatation of oesophageal stricture

Antibiotic	Dose
For patients on long-term penicillin therapy, hypersensitive to penicillin or who have taken penicillin or a related beta-lactam antibiotic more than once in the last month:	
Clindamycin	(Child: 15 mg/kg, up to 600 mg) 600 mg orally as 1 dose 1 hour before procedure
If unable to take orally	
Clindamycin	(Child: 15 mg/kg, up to 600 mg) 600 mg iv, over at least 20 mins just before procedure
Or vancomycin	(Child less than 12 years: 30 mg/kg up to 1.5 g) 1.5 g iv by slow infusion, over at least 1 hour just prior to procedure
Or lincomycin	(Child: 15 mg/kg, up to 600 mg) 600 mg iv, over 1 hour before procedure
Or teicoplanin	(Child: 10 mg/kg, up to 400 mg) 400 mg iv, just before the procedure or im 30 mins before procedure
For patients not on long-term penicillin therapy, not hypersensitive to penicillin and who have not taken penicillin or a related beta-lactam antibiotic more than once in the last month:	
Amoxycillin	(Child: 50 mg/kg up to 2 g) 2 g orally as 1 dose 1 hour prior to the procedure
Or amoxycillin/ ampicillin	(Child: 50 mg/kg up to 2 g) 2 g iv just prior to procedure or im 30 min prior to procedure

im, intramuscular; iv, intravenous.

Genitourinary and gastrointestinal procedures

- Surgery of the intestinal mucosa or biliary tract (except for endoscopy, biopsy and percutaneous endoscopic gastrostomy)
- Endoscopic retrograde cholangiography
- Prostate surgery
- Cystoscopy and urethral dilatation
- Vaginal delivery in the presence of infection, or prolonged labour or prolonged rupture of membranes
- Surgical procedures of the genitourinary tract in the presence of infection (e.g. urethral catheterisation, uterine dilatation and curettage, abortion, sterilisation and placement or removal of intrauterine contraceptive devices)

Antibiotic	Dose
Vancomycin	(Child <12 years: 30 mg/kg, up to 1.5 g) 1.5g iv by slow infusion, over at least 1 hour just prior to procedure
Or teicoplanin	(Child: 10 mg/kg up to 400 mg) 400 mg iv just prior to procedure

iv, intravenous.

Recommended routine review and management plan for ARF and RHD

Classification	Criteria*	Review and management plan	Frequency†
Priority 1 (severe)¥	Severe valvular disease **or** moderate/severe valvular lesion with symptoms **or** mechanical prosthetic valves, tissue prosthetic valves and valve repairs, including balloon valvuloplasty	Secondary prophylaxis (BPG)	3–4 weekly
		Doctor review	3–6 monthly
		Cardiologist/physician/paediatrician review	3–6 monthly
		Influenza vaccination	Yearly
		Echocardiography	3–6 monthly
		Dental review	Within 3 months of diagnosis, then 6 monthly thereafter
		Pneumococcal vaccination	Refer to *Immunisation handbook*
		Endocarditis prophylaxis	As required Refer to *Therapeutic Guidelines: Antibiotics 2010*
Priority 2 (moderate)	Any moderate valve lesion in the absence of symptoms, and with normal left ventricular function	Secondary prophylaxis (BPG)	4-weekly
		Doctor review	6-monthly
		Influenza vaccination	Yearly
		ECG (optional)	Yearly
		Cardiologist/physician/paediatrician review	Yearly
		Echocardiography	Yearly
		Dental review	Within 3 months of diagnosis, then 6 monthly
		Pneumococcal vaccination	Refer to *Immunisation handbook*
		Endocarditis prophylaxis	As required Refer to *Therapeutic Guidelines: Antibiotics 2010*
Priority 3 (mild)	ARF with no evidence of RHD **or** trivial to mild valvular disease	Secondary prophylaxis (BPG)	4 weekly
		Doctor review	Yearly
		Echocardiography	Children: 2 yearly‡ Adults: 2–3 yearly‡
		Dental review	Yearly

Classification	Criteria*	Review and management plan	Frequency†
		Endocarditis prophylaxis	As required Refer to *Therapeutic Guidelines: Antibiotics 2010*
Priority 4 (inactive)	Patients with a history of ARF (no RHD) for whom secondary prophylaxis has been ceased	Medical review	Yearly
		Dental review	Yearly
		Cardiologist/physician/paediatrician review	As referred with new symptoms
Additional considerations	Following valve surgery	Medical assessment ECG Chest radiograph Echocardiography Full blood count Urea, creatinine, electrolytes INR, if indicated	3–4 weeks' post-discharge
	Missed doses of BPG	Patient should be contacted if they have not presented within 3 days of due injection	
	Patient travelling to another community when injection due	Consideration should be given to bringing forward the date of injection to 2–3 weeks, or arrangements made with other service providers in advance	

*Serial echocardiographic assessments are required in the long-term management of RHD as an essential tool in determining the progress of cardiac damage and the optimal timing of surgery. Therefore, risk stratification should be based on clinical and echocardiographic findings (Grade D). †Review frequency should be determined according to individual needs and local capacity. Most critically, the frequency of review should become more frequent in the event of symptom onset, symptomatic deterioration or a change in clinical findings. ¥Any patient with severe valvular disease or moderate to severe valvular disease with symptoms should be referred for cardiological and surgical assessment as soon as possible. ‡In patients with no evidence of valvular disease on echocardiography, who have no documented ARF recurrences, good adherence to secondary prophylaxis and no cardiac murmurs on examination at follow up appointments, echocardiography may not be needed as frequently.

BPG, benzathine penicillin G; ECG, electrocardiogram; INR, international normalised ratio.

5. Management of rheumatic heart disease

The fundamental goal in the long-term management of RHD is to prevent ARF recurrences, and therefore, prevent the progression of RHD, and in many cases allow for the resolution of heart disease.

This quick reference guide is derived from the *Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease* (2nd edn).

What is acute rheumatic fever?

Acute rheumatic fever (ARF) is an illness caused by a reaction to a bacterial infection with group A streptococcus. It causes an acute, generalised inflammatory response and an illness that targets specific parts of the body, including the heart, joints, brain and skin. Individuals with ARF are often unwell, have significant joint pain and require hospitalisation. Despite the dramatic nature of the acute episode, ARF typically leaves no lasting damage to the brain, joints or skin, but can cause persisting heart damage, termed 'rheumatic heart disease' (RHD).

What is RHD?

RHD is damage to the heart that remains after the acute ARF episode has resolved. It is caused by an episode or recurrent episodes of ARF, where the heart has become inflamed; the heart valves remain stretched and/or scarred, and normal blood flow is interrupted. Recurrences of ARF may cause further valve damage, leading to worsening of RHD. Preventing recurrences of ARF by using prophylactic treatment with penicillin is therefore of great importance in controlling RHD.

Who gets RHD?

In Australia, the vast majority of people with RHD are Aboriginal people and Torres Strait Islanders, many of whom live in remote areas of central and northern Australia. Pacific Islanders, and migrants from high-prevalence countries, are also at high risk.

Best practice in RHD management

It is difficult and expensive for Aboriginal people and Torres Strait Islanders to travel to major centres for cardiac services, which are often hospital based. Although specialist outreach services are improving in many regions, access to specialist care is suboptimal in rural and remote areas.

The implementation of guidelines for RHD has major implications for Aboriginal and Torres Strait Islander healthcare services, especially in rural and remote regions. In addition to access to appropriate primary care services, best practice for RHD requires:

- secondary prevention with penicillin prophylaxis
- adequate monitoring of anticoagulation therapy in patients with atrial fibrillation and/or mechanical prosthetic valves
- access to oral healthcare
- access to echocardiography
- access to a specialist physician, paediatrician and/or cardiologist, preferably the same specialist, for regular follow up visits
- access to cardiothoracic and interventional cardiology services.

The fundamental goal in the long-term management of RHD is to prevent ARF recurrences, and therefore, prevent the progression of RHD, and in many cases allow for the resolution of heart disease.

Valvular lesions in RHD

Specific valvular lesions in chronic RHD include:

- *mitral regurgitation*, in which volume overload of the left ventricle (LV) and left atrium occurs. In more severe cases, this may result in a progressive decline in systolic contractile function
- *mitral stenosis*, where progressive obstruction to left ventricular inflow develops, due to fibrosis and partial fusion of the mitral valve leaflets
- *aortic regurgitation*, where left ventricular volume overload occurs, and there is an increase in left ventricular end-diastolic volume, eventually leading to left ventricular contractile dysfunction in more severe cases
- *aortic stenosis*, which results from fibrosis and fusion of the valve cusps, causing progressive obstruction to left ventricular outflow
- *tricuspid regurgitation*, maybe secondary to left sided rheumatic valve disease or reflect inflammatory rheumatic involvement
- *tricuspid stenosis*, uncommon but causes obstruction to right ventricle inflow (RV).

In patients with multiple valve lesions, management usually focuses on the most severe valve lesion.

Key points in the management of rheumatic mitral regurgitation

Symptoms	May be asymptomatic for many years Exertional dyspnoea and fatigue
Examination	Pan-systolic murmur at LV apex
Echocardiography	Overriding or prolapse of AMVL Thickened 'dog leg' AMVL, especially if associated with mitral stenosis Retrograde colour (mosaic) regurgitant jet into left atrium, often posteriorly directed Severity graded by area of colour regurgitant jet in left atrium LV chamber dimensions enlarged if moderate or greater MR Assess LV systolic function
Cardiac catheterisation	Only to exclude coronary artery disease
Medical management	In chronic, stable MR (regardless of severity), there is no role for vasodilators, diuretics or ACE inhibitors unless clinical heart failure is present
Indications for surgery	Moderate / severe MR: 1. NYHA FC II-IV symptoms **OR** 2. Impaired LV systolic function EF <60 % **OR** 3. LVESD ≥ 40 mm in adults or enlarged LVSED Z-score in children **OR** 4. PAS hypertension >50 mmHg **OR** 5. New onset atrial fibrillation
Choice of operation	Mitral valve repair operation of choice Mitral valve replacement with biological or mechanical prosthesis Avoid mechanical prostheses, if concerns about warfarin adherence or future pregnancy

ACE, angiotensin-converting enzyme; AMVL, anterior mitral valve leaflet; EF, ejection fraction; LV, left ventricle; LVESD, left ventricular end-systolic diameter; MR, mitral regurgitation; NYHA FC, New York Heart Association Functional Class; PAS, pulmonary artery systolic.

Key points in the management of rheumatic mitral stenosis

Symptoms	May be asymptomatic Exertional dyspnoea, fatigue, palpitations
Examination	Low-pitched mid diastolic 'rumble' at LV apex
Echocardiography	Thickened restricted 'dog leg' AMVL Restricted posterior leaflet Measure mean mitral diastolic gradient from continuous-wave Doppler signal Calculate MVA from slope of Doppler mitral inflow velocity Calculate PAS pressure
Cardiac catheterisation	Only to exclude coronary artery disease
Atrial fibrillation	Common Rate control using beta-blockers or digoxin Consider cardioversion, if recent onset Need anticoagulation to prevent thromboembolic complications
Medical management	Diuretics (e.g. frusemide, spironolactone) are only indicated in patients with symptomatic pulmonary venous congestion or pulmonary oedema All symptomatic patients should be referred for cardio-surgical assessment
Indications for intervention	Symptoms NYHA FC II–IV MVA <1.5 cm^2 **OR** PAS pressure >50 mmHg
Procedure of choice	PBMV by high-volume operator/centre Mitral valve repair or replacement if morphology is not suitable for PBMV (e.g. valve is heavily calcified) or if moderate or greater MR is present

AMVL, anterior mitral valve leaflet; LV, left ventricle; MR, mitral regurgitation; MVA, mitral valve area; NYHA FC, New York Heart Association Functional Class; PAS, pulmonary artery systolic; PBMV, percutaneous balloon mitral valvuloplasty.

Key points in the management of rheumatic aortic regurgitation

Symptoms	May be asymptomatic for many years Exertional dyspnoea and fatigue
Signs	Diastolic blowing and/or decrescendo murmur at left sternal border, usually associated with systolic ejection murmur
Echocardiography	Retrograde diastolic regurgitant colour jet in LVOT and LV chamber Area of jet in LVOT correlates with severity LV chamber dimensions enlarged, if moderate or greater aortic regurgitation Associated mitral valve disease is common Pan-diastolic reversed diastolic flow in descending thoracic aorta, if moderate/severe aortic regurgitation (Doppler) Assess LV systolic function
Cardiac catheterisation	Only to exclude coronary artery disease
Medical management	All symptomatic patients should be commenced on an ACE inhibitor and referred for cardio-surgical evaluation Consider ACE inhibitors or vasodilator therapy with dihydropyridines (e.g. nifedipine) in asymptomatic patients with moderate or greater aortic regurgitation , especially if systolic hypertension is present
Indications for surgery	Moderate/severe aortic regurgitation with symptoms NYHA FC II–IV Asymptomatic moderate/severe aortic regurgitation if: • LVEF <55% **OR** • LVESD ≥55 mm **OR** • LVEDD >70 mm **OR** • Enlarged LVESD or LVEDD Z-score (in children only)
Choice of surgery	1. Bioprosthetic or homograft valve replacement: • no requirement for anticoagulation if in sinus rhythm • limited durability in younger patients 2. Mechanical valve replacement: • anticoagulation is required 3. Aortic valve repair: • many centres have limited experience 4. Ross procedure (replacement of the aortic valve with a pulmonary autograft and replacement of the pulmonary valve with a homograft): • only in selected cases with experienced surgeons

ACE, angiotensin-converting enzyme; LV, left ventricle; LVEDD, left ventricular end-diastolic diameter; LVEF, left ventricular ejection fraction; LVESD, left ventricular end-systolic diameter; LVOT, left ventricular outflow tract; NYHA FC, New York Heart Association Functional Class.

Key points in the management of rheumatic aortic stenosis

Symptoms	May be asymptomatic Exertional dyspnoea, angina, syncope
Signs	Low-pitched, systolic ejection murmur in aortic area
Echocardiography	Thickened, restricted aortic valve leaflets Measure peak and mean systolic gradient from Doppler velocity across aortic valve Assess left ventricular systolic function
Cardiac catheterisation	Only to exclude coronary artery disease
Medical management	Medical therapy is not indicated in asymptomatic patients Symptomatic patients require surgery and do not benefit from medical therapy
Indications for surgery	Symptoms plus mean systolic gradient > 40-50 mmHg or AVA <1.0 cm^2 Impaired cardiac function (EF < 50%) plus mean systolic gradient > 40-50 mmHg or AVA <1.0 cm^2
Choice of surgery	Bioprosthetic or homograft valve replacement: • limited durability • no requirement for long-term anticoagulation if in sinus rhythm Mechanical valve replacement: • long-term anticoagulation is required

AVA, aortic valve area; EF, ejection fraction.

Key points in the management of rheumatic tricuspid regurgitation

Symptoms	Exertional dyspnoea and fatigue, usually secondary to left sided rheumatic valve disease
Examination	Elevated jugular venous pressure with prominent v wave in jugular pulse Pansystolic murmur left sternal border Hepatomegaly, may be pulsatile Ascites Peripheral oedema
Echocardiography	Thickened leaflets Retrograde colour jet into right atrium Severity graded by area of colour jet Dilated IVC Retrograde flow in hepatic veins Right ventricular chamber enlargement if moderate or greater TR
Medical Management	Symptoms are generally related to the left sided valve lesions Diuretics (e.g. frusemide, spironolactone) are only indicated in patients with symptomatic right and/or left heart failure Note: Usually impossible to distinguish rheumatic from non-rheumatic tricuspid valve regurgitation clinically or by echocardiogram
Indications for surgery	Moderate/severe TR usually in association with symptomatic MVD Progressive symptomatic right heart failure
Choice of surgery	Tricuspid valvuloplasty Tricuspid valve replacement with mechanical or biological prosthesis if valvuloplasty not possible

IVC, inferior vena cava; MVD, mitral valve disease; TR, tricuspid regurgitation.

Key points in the management of rheumatic tricuspid stenosis

Symptoms	Usually secondary to left sided rheumatic valve disease
Examination	Elevated jugular venous pressure Prominent a wave in jugular pulse Presystolic and mid diastolic murmur at the left sternal border
Echocardiography	Thickened, restricted tricuspid valve leaflets with doming Diastolic gradient measured across tricuspid valve as per MS
Medical management	Symptoms are generally related to the left sided valve lesions Diuretics (e.g. frusemide, spironolactone) are only indicated in patients with symptomatic right and/or left heart failure
Indications for surgery	Moderate/severe TS in association with symptomatic MVD Progressive right heart failure
Choice of surgery	Percutaneous balloon valvuloplasty or surgical commisurotomy operation of choice Tricuspid valve replacement with mechanical or biological prosthesis if repair or PBTV not possible

MS, mitral stenosis; MVD, mitral valve disease; PBTV, percutaneous balloon tricuspid valvuloplasty; TS, tricuspid stenosis.

6. Rheumatic heart disease in pregnancy

Ideally, patients with known rheumatic valvular disease should be properly assessed before pregnancy. Discussion regarding fertility planning should be undertaken with all women with more than mild valvular disease, even if immediate pregnancy is not planned.

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What is acute rheumatic fever?

Acute rheumatic fever (ARF) is an illness caused by a reaction to a bacterial infection with group A streptococcus. It causes an acute, generalised inflammatory response and an illness that targets specific parts of the body, including the heart, joints, brain and skin. Individuals with ARF are often unwell, have significant joint pain and require hospitalisation. Despite the dramatic nature of the acute episode, ARF typically leaves no lasting damage to the brain, joints or skin, but can cause persisting heart damage, termed 'rheumatic heart disease' (RHD). Recurrences of ARF may cause further cardiac valve damage. Thus, RHD steadily worsens in people who have multiple episodes of ARF.

What is RHD?

RHD is damage to the heart that remains after the acute ARF episode has resolved. It is caused by an episode or recurrent episodes of ARF, where the heart has become inflamed; the heart valves remain stretched and/or scarred, and normal blood flow is interrupted. Recurrences of ARF may cause further valve damage, leading to worsening of RHD. Preventing recurrences of ARF by using prophylactic treatment with penicillin is therefore of great importance in controlling RHD.

Who gets RHD?

In Australia, the vast majority of people with RHD are Aboriginal people and Torres Strait Islanders, many of whom live in remote areas of central and northern Australia. Pacific Islanders, and migrants from high-prevalence countries, are also at high risk.

Pregnancy in women with RHD

Normal pregnancy is associated with a 30–50% increase in blood volume, reduction in systemic vascular resistance and corresponding increase in cardiac output. These changes begin during the first trimester, peaking at 28–30 weeks of pregnancy, and are then sustained until term. The increase in blood volume is associated with an increase in heart rate by 10–15 beats per min. Because of the hyperdynamic circulation, innocent, soft mid-systolic murmurs are common during pregnancy, particularly along the left sternal border. These circulatory changes of pregnancy will exacerbate any pre-existing valvular disease. Sometimes RHD, especially mitral stenosis, is first diagnosed during pregnancy or soon after delivery when a woman develops symptoms, usually dyspnoea.

Assessment of women with RHD

Ideally, patients with known rheumatic valvular disease should be properly assessed before pregnancy. Discussion regarding fertility planning should be undertaken with all women with more than mild valvular disease, even if immediate pregnancy is not planned. Assessment should include a full history and examination, and an echocardiogram. If patients are already symptomatic, due to significant rheumatic valvular disease, serious consideration should be given to interventional therapy or surgery prior to pregnancy to avoid life-threatening complications, which may occur in these patients. In these patients, the use of contraception with a low failure rate (etonogestrel implant; Implanon, Organon International, Oss, the Netherlands) should be strongly encouraged if there is a risk of pregnancy, while more definitive treatment is being undertaken.

Risk factors

The predictors of increased maternal and fetal risk in the pregnant patient with rheumatic valvular disease are:

- reduced left ventricle (LV) systolic function
- significant aortic or mitral stenosis
- moderate or severe pulmonary hypertension
- a history of heart failure
- symptomatic valvular disease before pregnancy
- atrial fibrillation, especially when anticoagulation is required
- pregnant women with mechanical valves are a very high-risk group, in whom all anticoagulation options carry maternal and/or fetal risks.

Management of women with RHD

Women with more than mild RHD should be identified as having a higher than normal risk of complications in pregnancy, and should receive antenatal care at an appropriate referral centre with an experienced obstetrician, in collaboration with an obstetric physician and/or cardiologist. The most severe cases should be seen at a referral centre with cardiology and intensive care facilities. Discussions with the woman regarding timing, nature and site of planned delivery should occur before or early in pregnancy.

During pregnancy, women with valvular heart disease should be reviewed regularly by a cardiac specialist, and cardiac status should be reviewed whenever there is a change in symptoms. A multidisciplinary approach to management is an important principle for care of the pregnant patient with rheumatic valvular disease. It is often necessary to advise women with heart disease to cease work earlier in pregnancy for medical reasons, as cardiac demands increase significantly as pregnancy proceeds. Most women with valvular heart disease become more symptomatic in the third trimester.

Key points in the management of pregnancy in women with RHD

Predictors of increased maternal and fetal risk	Decreased LV systolic function Significant aortic and mitral stenosis Moderate or severe pulmonary hypertension Heart failure Symptoms before pregnancy Mechanical valve prostheses Atrial fibrillation requiring warfarin
Cardiac assessment	Early comprehensive assessment with echocardiography to assess valves and LV function Plan multidisciplinary management
Mitral/aortic regurgitation	Usually well tolerated Treat medically with diuretics, vasodilators (no ACE inhibitors/angiotensin II receptor blockers) for clinical heart failure
Mitral stenosis	Mild to moderate mitral stenosis: manage medically moderate to severe mitral stenosis (MVA <1.5 cm^2)—consider PBMV during late second trimester, if patient remains symptomatic and PAS pressure >50 mmHg Beta-blockers or digoxin for rate control of atrial fibrillation
Aortic stenosis (rare)	Mild to moderate aortic stenosis: well-tolerated. Diuretics for heart failure Consider PTAV if severe symptoms Beta-blockers or digoxin for rate control of atrial fibrillation. Avoid cardiac surgery, as high risk of fetal loss
Mechanical/ prosthetic valves and anticoagulation in pregnancy	High maternal and fetal risk Risk of warfarin embryopathy in first trimester Embryopathy may be avoided if warfarin dose ≤5 mg
Choice of 3 antithrombotic regimens	1. LMWH throughout pregnancy, weight-adjusted dose with anti-Xa level monitoring 2. Warfarin throughout pregnancy, if can keep warfarin ≤5 mg, e.g. INR 2–3 in aortic prosthesis, sinus rhythm; change to LMWH or unfractionated heparin at 36 weeks 3. LMWH until 13 weeks, and then warfarin and aspirin until 36 weeks; change to LMWH or UFH until labour. Monitor anti-Xa levels with LMWH
Labour	Haemodynamic monitoring: non-invasive, if mild to moderate valve disease Antibiotic prophylaxis, if prolonged labour and/or ruptured membranes Aim for short second stage and multidisciplinary management approach, with low threshold for obstetric intervention

ACE, angiotensin-converting enzyme; anti-Xa, antifactor Xa; INR, international normalised ratio; LMWH, low-molecular weight heparin; LV, left ventricle; MVA, mitral valve area; PAS, pulmonary artery systolic; PBMV, percutaneous balloon mitral valvuloplasty; PTAV, percutaneous transluminal aortic valvuloplasty; UFH, unfractionated heparin.

7. Rheumatic heart disease control programs

Secondary prophylaxis of ARF in someone who is known to have had ARF is the only RHD control strategy shown to be effective and cost-effective at both individual and population levels. Effective RHD management involves regular clinical follow up, with specialist review and echocardiography.

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What is acute rheumatic fever?

Acute rheumatic fever (ARF) is an illness caused by a reaction to a bacterial infection with group A streptococcus. It causes an acute, generalised inflammatory response and an illness that targets specific parts of the body, including the heart, joints, brain and skin. Individuals with ARF are often unwell, have significant joint pain and require hospitalisation. Despite the dramatic nature of the acute episode, ARF typically leaves no lasting damage to the brain, joints or skin, but can cause persisting heart damage, termed 'rheumatic heart disease' (RHD). Recurrences of ARF may cause further cardiac valve damage. Hence, RHD steadily worsens in people who have multiple episodes of ARF.

What is RHD?

RHD is damage to the heart that remains after the acute ARF episode has resolved. It is caused by an episode or recurrent episodes of ARF, where the heart has become inflamed; the heart valves remain stretched and/or scarred, and normal blood flow is interrupted. Recurrences of ARF may cause further valve damage, leading to steady worsening of RHD. Preventing recurrences of ARF by using secondary prophylaxis treatment with penicillin is therefore of great importance.

Who gets RHD?

In Australia, the vast majority of people with RHD are Aboriginal people and Torres Strait Islanders, many of whom live in remote areas of central and northern Australia. Pacific Islanders, and migrants from high-prevalence countries, are also at high risk.

How RHD can be controlled

Secondary prophylaxis of ARF in someone who is known to have had ARF is the only RHD control strategy shown to be effective and cost-effective at both individual and population levels. The recommended method is a four-weekly benzathine penicillin G (BPG) injection.

The appropriate duration of secondary prophylaxis is determined by a number of factors, including age, time since the last episode of ARF and potential harm from recurrent ARF. For most individuals, the duration of secondary prophylaxis is at least 10 years.

Effective RHD management involves regular clinical follow up, with specialist review and echocardiography.

Problems with the control of RHD

While strategies for controlling RHD have been proven to be simple, cheap and cost-effective, they must be adequately implemented in the populations at highest risk of the disease.

Persistent high rates of recurrent ARF in high-risk populations highlight the continued barriers to secondary prevention.

Organised approaches are needed to increase the effectiveness of the secondary prevention of ARF and management of RHD. This should include strategies aimed at improving the delivery of secondary prophylaxis and patient care, the provision of education, coordinating available health services and advocacy for necessary and appropriate resources.

Organisational approaches to RHD control

A coordinated control program is the most effective approach in improving adherence to secondary prophylaxis of ARF and the clinical follow up of people with RHD.

Central to coordinated control programs at individual, community and national levels are registers of people with RHD or a history of ARF. Register-based programs improve case detection, increase adherence to secondary prophylaxis, reduce recurrences of ARF and decrease hospitalisations from ARF/RHD.

Registers also provide a mechanism for monitoring patient movements, orienting staff to ongoing care requirements, identifying individuals with poor adherence to long-term therapy and monitoring the success of programs and changes in disease epidemiology.

RHD control programs aim to:

- identify and register new cases of ARF and RHD
- improve the uptake of and adherence to secondary prophylaxis
- increase awareness of the diagnosis and management among healthcare providers
- improve clinical care and follow up in line with best practice
- support education and health promotion for individuals, families and the community
- promote primary prevention, aimed at preventing initial episodes of ARF
- use data to monitor patient outcomes and improve program strategies.

Recommended elements of RHD control programs

- Commitment from national, regional and local services, particularly to ensure long-term funding and governance support.
- An effective advisory committee that includes cardiologists, paediatricians, general practitioners, physicians, epidemiologists, nurses, public health practitioners, Aboriginal health service organisations and relevant community representatives.
- A dedicated coordinating team.
- An electronic patient register that contains data elements that support quality patient management, as well as any internal and external reporting requirements.
- A commitment to partnerships between clinicians and public health services in order to support the needs of people with ARF/RHD and the community.
- Prioritisation of primary and secondary antibiotic prophylaxis delivered within the framework of primary healthcare.
- Planning and advocacy for a stable supply of BPG, and established plans for established secondary prophylaxis in the event of supply reductions.

- A commitment to partnerships between clinicians and public health services in order to support the needs of people with ARF/RHD and the community.
- Education for health practitioners and health workers, and supported education for the community, those with the disease and their families.
- Activities guided by locally-relevant, evidence-based guidelines.
- Legislation and/or regulations warranting the notification of ARF/RHD, which is supported by public health surveillance activities at the state or territory level.
- A priority system that ensures that services are delivered to those at highest risk.
- A mechanism for monitoring the delivery of secondary prophylaxis and ongoing care.
- Evaluation of patient management and program activities.

Improving the uptake of and adherence to secondary prohylaxis in primary care

Evaluate the local health service environment to identify specific barriers to injection delivery. Based on the outcome of the evaluation, the following strategies may be useful:

- identify local, dedicated staff members responsible for the delivery of secondary prevention and the coordination of routine care
- focus on improving relationships between health staff and patients/families
- support and utilise the expertise, experience, community knowledge and language skills of Aboriginal health workers
- develop and implement recall and reminder systems (based on a local ARF/RHD register, where established) to accommodate the high mobility of individuals and groups
- ensure that recall systems extend beyond community boundaries
- establish networks for timely communication between health clinics
- use a centralised coordinator and register to assist in monitoring movement
- minimise staff turnover in remote and rural primary healthcare centres and regional hospitals, or minimise the impact of staff turnover, where possible
- promote the importance of secondary prophylaxis in preventing recurrent ARF, and the development or worsening of RHD
- improve the quality and delivery of ongoing health education and support for staff, patients and families
- implement measures to reduce pain of injections, where indicated
- base routine care on standardised evidence-based guidelines.

Surveillance

Passive surveillance of ARF usually depends on case identification from healthcare providers. Historically, this has underestimated the burden of disease, due to inaccuracies and incompleteness. In under resourced settings, problems with passive surveillance are exacerbated by the high turnover of staff, and lack of awareness of ARF and RHD.

Ideally, active surveillance should be used to augment passive surveillance. This entails establishing mechanisms to identify new cases of ARF and RHD, and to update information about existing cases. This could include mechanisms allowing access to hospital separation data, echocardiography reports, specialist review correspondence, primary healthcare clinic information and notifiable disease databases.

Where possible, these processes should be automated (e.g. with regular downloads of information regarding patients admitted to hospital with a diagnosis of ARF or RHD).

When active surveillance is established, an initial apparent increase in the prevalence of RHD is expected, primarily due to the detection and recording of existing cases, rather than the appearance of new cases. Similarly, improved access to specialist care may also result in greater rates of valvular surgery in the initial years after commencing a program.

When establishing surveillance systems for ARF/RHD control, a range of issues should be considered, including:

- defining the target population and high-risk groups requiring surveillance
- establishing a process for information flow from a range of potential data sources (e.g. case reporting, data collection instruments, data transmission and handling)
- formulating the essential data elements to be collected
- ethical and privacy legislation requirements, including consent
- data management (e.g. the most appropriate format for storing the data)
- proposed process and timeliness of data analysis
- dissemination and targets for the feedback of results
- needs of healthcare providers for individual patient and epidemiological information
- continuing refinement and evaluation of the surveillance system.

RHD registers

Register-based programs:

- improve case detection
- increase adherence to secondary prophylaxis
- reduce recurrences of ARF
- decrease hospitalisations from ARF/RHD.

Some programs have all-relevant patient data entered into a centralised register. Others choose to have a subset of data (e.g. recording of individual doses for secondary prophylaxis) entered only into the local register.

Where provision of secondary prophylaxis is not entered into a central register, local health staff should have clear guidelines on identifying and managing patients overdue for secondary prophylaxis.

Screening for RHD

RHD control programs should coordinate screening programs to detect previously undiagnosed RHD in high-risk populations, wherever possible. Ideally, screening should be undertaken at a time when the program has been established, and newly-identified cases are able to be managed within a supportive framework.

Although RHD prevalence is highest in adults, they are difficult to screen, and screening of school-aged children is therefore recommended (e.g. cardiac auscultation at school entry, and again at 10 years of age).

If time and other resources allow, consideration should be given to conducting more intensive screening programs, in which children of all ages are reviewed, and attempts are also made to examine children who miss school-based screening.

Legislated notification of ARF/RHD

ARF is a notifiable condition in Western Australia, the Northern Territory and Queensland.

RHD is not currently notifiable anywhere in Australia.

Indicators for evaluating ARF/RHD control programs

Control programs for ARF/RHD should be evaluated against criteria for routine care and key epidemiological objectives (see below).

Consideration should be given to assessing the delivery of specialist cardiology services, the availability and accessibility of echocardiography, referral practices and structures, transportation for patients and support and follow up processes.

Key Performance Indicators

1	Epidemiology		
	1.1	Yearly age-specific incidence rates of all episodes, and of first episodes of ARF according to sex (refer to 1.1.1) and ethnicity (refer to 1.1.2)	• 0-4 • 5-14 • 15-24 • 25-34 • 35-44 • >44yrs
		1.1.1 sex	• Male • Female • Indeterminate • Not stated/inadequately described
		1.1.2 ethnicity	• Aboriginal but not Torres Strait Islander origin • Torres Strait Islander, but not Aboriginal origin • both Aboriginal and Torres Strait Islander origins • Maori • other Pacific Islanders • other • unknown
	1.2	Proportion of all recorded ARF episodes classified as recurrences	
	1.3	Rates of ARF recurrences per 100 patient-years	
	1.4	Number of deaths and age-standardised rates of mortality due to ARF and RHD in the previous calendar year by ethnicity (refer to 1.1.2)	
	1.5	Yearly age-specific (refer to 1.1) and overall incidence of RHD by ethnicity (refer to 1.1.2) and broken down by method found and presented	• all recorded RHD cases • cases classified as mild • cases classified as moderate • cases classified as severe
	1.6	Yearly age-specific (refer to 1.1) prevalence of RHD, by ethnicity (refer to 1.1.2)	• all recorded RHD cases • cases classified as mild • cases classified as moderate • cases classified as severe
	1.7	Proportion of newly registered cases of ARF or RHD with an initial recorded diagnosis being established RHD (rather than ARF)	

2	**Requirement and uptake of secondary prophylaxis**		
	2.1	Proportion of all people indicated for secondary prophylaxis* who are registered to receive benzathine penicillin G (BPG)	
	2.2	Median percentage of all scheduled BPG doses actually delivered	
	2.3	Proportion of people indicated for BPG secondary prophylaxis who received <50%, 50-79%, and ≥80% of scheduled doses in the previous calendar year	
3	**Quality of management**		
	3.1	Proportion of all registered ARF and RHD cases classified as mild, moderate, severe and inactive	
	3.2	Proportion of people classified as moderate or severe RHD who had an echocardiogram within the previous 6 months, 1 year, and 1-2 years	
	3.3	Number of cases, and proportion of total cases indicated for cardiac surgery, who have been waiting <6 months, 6-11 months, 12-23 months, or 24+ months	
	3.4	Number and type of surgical procedures performed during the previous calendar year by the following:	
		3.4.1 age group	(refer to 1.1)
		3.4.2 ethnicity	(refer to 1.1.2)
	3.5	Number (and proportion) of people who died in the previous calendar year within 28 days and 1 year of undergoing rheumatic cardiac surgery by the following:	
		3.5.1 age group	(refer to 1.1)
		3.5.2 ethnicity	(refer to 1.1.2)

* If denominator of those indicated for prophylaxis not known, use people with a history of ARF within the last 10 years OR ARF and RHD and aged < 21 years OR aged ≥21 years with moderate or severe RHD

RHD Australia

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The Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition)

Quick reference guides include:

- Primary prevention of ARF
- Diagnosis of ARF
- Management of ARF
- Secondary prevention of ARF
- Management of RHD
- RHD in pregnancy
- RHD control programs

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