KEY INFORMATION

Australian guideline for the prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease

(Edition 3.3) 2025

menzies SCHOOL OF HEALTH RESEARCH

Heart Foundation

This document includes a summary of key information from each chapter of the Australian guideline for the prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (Edition 3.3) published in 2025.

References have been removed, and only selected tables and figures have been included.

To read the full Guideline go to https://rhdaustralia.org.au/arf-rhd-guidelines/.

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Introduction

FOREWORD

This edition of the Australian guideline for the prevention, diagnosis and management of acute rheumatic fever (ARF) and rheumatic heart disease (RHD) has a new focus which places people with ARF and RHD, and their families and communities, at the centre of care. To achieve a value-based healthcare system that breaks down the complex and hierarchical structures based on Western ideologies, we must look at whose values are represented.

There are many cultural and structural barriers for First Nations peoples requiring evidence-based care. Most, however, are poorly understood. If guidelines are to be successful, we need to move beyond the 'evidence base' – what is known and understood – to what we do not know. We need to understand the complex relationships between the social, cultural, political and economic situations in which people live.

Despite advances in medical treatment and management of ARF and RHD, the associated health benefits at population and community level have not been as evident for First Nations peoples as they have for non-Indigenous Australians. These challenges are more than biomedical and are driven by the social, cultural and environmental determinants of health. The Australian Institute of Health and Welfare reported that in 2022, Indigenous Australians represented almost 80% of all new RHD cases. The report also indicated that only 14% of First Nations peoples with ARF and RHD received all prescribed secondary prophylaxis injections. The healthcare system must respond to these disparities and refocus on people with this disease; acknowledging their unique culture, and the social, economic and environmental circumstances in which they live.

There is a growing interest in ethnomedicine where traditional biomedical healthcare methods are guided by Indigenous cultural beliefs and practices. Within each chapter of this guideline, the medical problems and solutions have been viewed within a socio-cultural context, with the aim of reducing the evidence-practice gap. This guideline identifies the systemic factors that drive disparities in best-practice care delivery and offers solutions. We have come a long way from the first edition, and this journey has culminated in an important balance between cultural and clinical competence.

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April 2025

SUMMARY OF IMPORTANT CHANGES IN THIS EDITION

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None

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Primordial prevention and social determinants of acute rheumatic fever

None

Primary prevention

Addition of Summary of Recommendations with GRADE Level of Evidence (Table 5.1)

Azithromycin elevated to first line oral option for treatment of sore throats (Table 5.3)

Progress of molecular point of care tools for identifying Strep A throat infections

Recommendation to use phenoxymethylpenicillin (oral penicillin) as first line treatment for sore throat during periods of rationing premix Benzylpenicillin G (Table 5.3)

Clarity that antibiotic treatment is indicated for all people (not just children) with one or more skin lesions with pus or thick crust (Table 5.4)

Diagnosis of acute rheumatic fever

Addition of Summary of Recommendations with GRADE Level of Evidence (Table 6.1)

Clarification of who may be managed in the community with suspected ARF (on medical specialist advice)

Addition of "advanced conduction abnormalities" on ECG as a minor manifestation of ARF

Dengue, chikungunya and malaria added to the differential diagnosis of polyarthritis and fever

Section Echocardiography and ARF updated in line with 2023 World Heart Federations RHD diagnosis guidelines

- Discussion related to valvulitis: minimal echocardiographic criteria diagnosis of acute rheumatic fever for pathological regurgitation removed
- Discussion related to morphological changes associated with rheumatic carditis removed

Clarification that echocardiography cannot accurately determine the timing or duration of rheumatic valve changes

Management of acute rheumatic fever

Updated medications used for acute rheumatic fever with GRADE Level of Evidence (Table 7.1)

Removal of tramadol from management of severe pain while awaiting diagnostic confirmation (Table 7.1)

New emphasis on potential disease-modifying treatment (corticosteroid) in the management of Sydenham chorea (Table 7.1)

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Updated Sydenham chorea management strategies (Table 7.6)

Diagnosis of rheumatic heart disease

Addition of Summary of Recommendations with GRADE Level of Evidence (Table 8.1)

Integration of the 2023 World Heart Federation (WHF) diagnostic morphological features for RHD (Table 8.5)

Integration of the 2023 WHF guidelines for pathological valvular regurgitation and stenosis (Table 8.6)

Integration of the 2023 WHF guidelines staging criteria (with removal of definite and borderline definitions) (Table 8.7)

Updated reference for 2023 WHF guidelines and explanation of the staging criteria

Addition of 2023 WHF echocardiographic screening criteria (Table 8.8)

Screening for rheumatic heart disease

Addition of Summary of Recommendations with GRADE Level of Evidence (Table 9.1)

Integration of the WHF 2023 echocardiographic diagnosis of rheumatic heart disease (RHD) guidelines including screening and confirmatory criteria

Integration of new evidence including randomised controlled trial data on prophylaxis of early echocardiography detected RHD

Secondary prophylaxis

Addition of Summary of Recommendations with GRADE Level of Evidence (Table 10.1)

Updated recommendations for duration of secondary prophylaxis (Table 10.3)

Addition of guidance for managing long-acting penicillin supply interruptions

Addition of technique for administering BPG injections

Updated strategies to manage injection pain (Figure 10.1)

Guidance for BPG administration in people who may be at high risk of vasovagal syncope (fainting)

Consideration of oral antibiotic prophylaxis rather than intramuscular injections for the small subset of people who may be at high risk of vasovagal syncope (fainting)

Recommendation for clinical discretion for duration of antibiotic prophylaxis after surgery in people aged over 40 years

Addition of calculation for days at risk

Management of rheumatic heart disease

Addition of Summary of Recommendations with GRADE Level of Evidence (Table 11.1)

Integration of management recommendations for all stages of RHD based on 2023 World Heart Federation guidelines, Table 11.2. Priority classification and recommended follow-up (updated 2025)

Integrated evidence from the INVICTUS trial regarding anticoagulation in AF and advanced MS (no change to recommendations)

No change to the management of each individual valve lesion or indications for referral to surgery/heart team. These appear in line with ESC and AHA/ACC valvular heart disease guidelines

Women and girls with rheumatic heart disease

Emerging importance of early access to echocardiography for pregnant women in high-risk populations

Rheumatic heart disease control programs

Updated status of RHD control programs in the Australian context

Addition of notifiable conditions (Table 13.1)

Updated notification process and consent requirements (Table 13.2)

Updated key performance targets and metrics for RHD Control Programs (Table 13.3)

New technologies

Introduction to the SubCutaneous Infusions of high dose benzathine benzylpenicillin G (SCIP) trial

Updated discussion on Strep A vaccine development

CLASSIFICATIONS OF ACUTE RHEUMATIC FEVER AND RHEUMATIC HEART DISEASE USED IN THIS GUIDELINE

Classification of ARF

Definite ARF: acute presentation which fulfils Jones diagnostic criteria for ARF.

Probable ARF: acute presentation which does not fulfil Jones diagnostic criteria for ARF, missing one major or one minor criterion or lacking evidence of preceding streptococcal infection, but ARF is still considered the most likely diagnosis.

Possible ARF: acute presentation which does not fulfil Jones diagnostic criteria for ARF, missing one major or one minor criterion or lacking evidence of preceding streptococcal infection, and ARF is considered uncertain but cannot be ruled out.

Classification of RHD

Stage A: Minimum echocardiographic criteria for RHD, may be at risk for valvular heart disease progression.

Stage B: Mild RHD, at risk for developing clinical symptoms of valvular heart disease.

Stage C: Advanced RHD, at high risk of developing clinical complications of disease.

Stage D: Advanced RHD, already has clinical complications of disease.

RHD in pregnancy risk levels

Level I: low risk of maternal mortality, low to moderate risk of morbidity (e.g. mild RHD with no mitral stenosis).

Level II: elevated risk of maternal mortality or moderately increased risk of morbidity (e.g. bioprosthetic valve or mild mitral stenosis).

Level III: further elevated risk of maternal mortality or severe morbidity (e.g. mechanical heart valve, severe asymptomatic mitral / aortic regurgitation or severe asymptomatic aortic stenosis or moderate mitral stenosis.

Level IV: extremely high risk of maternal mortality or severe morbidity (e.g. severe mitral stenosis or valve disease with pulmonary hypertension).

Types of penicillin used in ARF

Benzathine benzylpenicillin G: long-acting intramuscular formulation of penicillin.

Phenoxymethylpenicillin: short-acting oral formulation of penicillin.

LEVELS OF EVIDENCE FOR GRADE RECOMMENDATIONS

Grading of Recommendations Assessment, Development and Evaluation (GRADE) is used in this document where the level of evidence of a recommendation requires grading. The GRADE approach is an internationally recognised system for grading quality of evidence and strength of recommendations. The GRADE approach rates evidence across studies for specific clinical outcomes to link evidence-quality evaluations to recommendations in clinical guidelines.

Table 1.1. GRADE evidence grade

CODE	QUALITY OF EVIDENCE	DEFINITION
A	High	We are very confident that the true effect lies close to that of the estimate of the effect
B	Moderate	We are moderately confident in the effect estimate: the true effect is likely to be close to the estimate of the effect, but there is a possibility that it is substantially different
C	Low	Our confidence in the effect estimate is limited: the true effect may be substantially different from the estimate of the effect
D	Very low	We have very little confidence in the effect estimate: the true effect is likely to be substantially different from the estimate of effect

Table 1.2. GRADE strength of recommendations

CODE	STRENGTH OF RECOMMENDATION	IMPLICATIONS WHEN COMBINED WITH EVIDENCE GRADE
1	Strong	1A: Strong recommendation, applies to most patients without reservation. Clinicians should follow a strong recommendation unless a clear and compelling rationale for an alternative approach is present 1B: Strong recommendation, applies to most patients. Clinicians should follow a strong recommendation unless a clear and compelling rationale for an alternative approach is present 1C: Strong recommendation, applies to most patients. Some of the evidence base supporting the recommendation is, however, of low quality
2	Weak	2A: Weak recommendation. The best action may differ depending on circumstances of patients or societal values 2B: Weak recommendation. Alternative approaches likely to be better for some patients under some circumstances 2C: Very weak recommendation. Other alternatives may be equally reasonable 2D: No evidence available; expert consensus judgement

Culture and Workforce

KEY INFORMATION

- Centrality of culture is the core component of these guidelines.
- Cultural and structural competencies in healthcare are necessary to close the evidence–practice gap.
- An ethnomedical framework (respecting and incorporating traditional Indigenous medical practices) should be used to inform guideline development.
- A socioecological model (understanding the personal and environmental factors – interpersonal, community, organisational and environmental – that determine health behaviours) can highlight the complex relationships that exist for First Nations peoples.
- An adequately trained and supported First Nations workforce is the key driver for successful health programs.

Cultural awareness is different to cultural safety. Cultural awareness is awareness of one's own biases. It is a single event while cultural safety is the ongoing critical reflection of health practitioner knowledge, skills, attitudes, practising behaviours and power differentials in delivering safe, accessible and responsive healthcare free of racism and harm. The effect of cultural safety is judged by the person receiving care

Workplaces must be free of racism, culturally safe and supportive, and attractive to the First Nations health workforce. The First Nations health workforce has unique insight into the lived experiences of families and communities, support by their knowledge of cultural beliefs, practices, and protocols with First Nations peoples. Health workers possess a cultural intellect that cannot be replicated by mainstream.

Burden of acute rheumatic fever and rheumatic heart disease

KEY INFORMATION

- Since the early 1990s, acute rheumatic fever (ARF) in Australia has occurred almost exclusively in young First Nations peoples, particularly in the 5–14-year-old age group.
- During the same period, rheumatic heart disease (RHD) has predominately affected young to middle-aged First Nations peoples as a consequence of the current era of endemic ARF among this population. Older non-Indigenous people with RHD were affected during a past era of endemic ARF.
- After accounting for age difference between populations, the rate of new diagnoses among First Nations peoples in 2015–2017 was 98.7 times higher for ARF and 49.0 times higher for RHD compared with the non-Indigenous population.
- Females are more likely to be diagnosed with ARF and RHD than males.
- The number of First Nations peoples with ARF and RHD is increasing.
- Over a quarter of people diagnosed with ARF or RHD are not recorded on Australian registers. In high-risk areas, many people with RHD are only diagnosed during screening activities.
- The burden of disease often spans the majority of a person's lifetime, starting with ARF in childhood, where ongoing active engagement with the healthcare system is needed for many years and progressing in many cases to RHD and associated heart conditions during adulthood.
- People who have had ARF are more likely to have a recurrence, with one in five people having a recurrent episode of ARF within 10 years of their first.
- There is a high risk of rheumatic valvular damage from a recurrent or single severe episode of ARF; more than half of those with ARF progress to RHD within 10 years of their initial ARF episode, and more than one-third of these people develop severe RHD.
- Almost one quarter of young people initially diagnosed with mild or moderate RHD will progress to a complication within 8 years of diagnosis.
- First Nations peoples with RHD are more likely to die compared to non-Indigenous Australians with RHD; however, the death rates have decreased for all population groups over the past few decades.

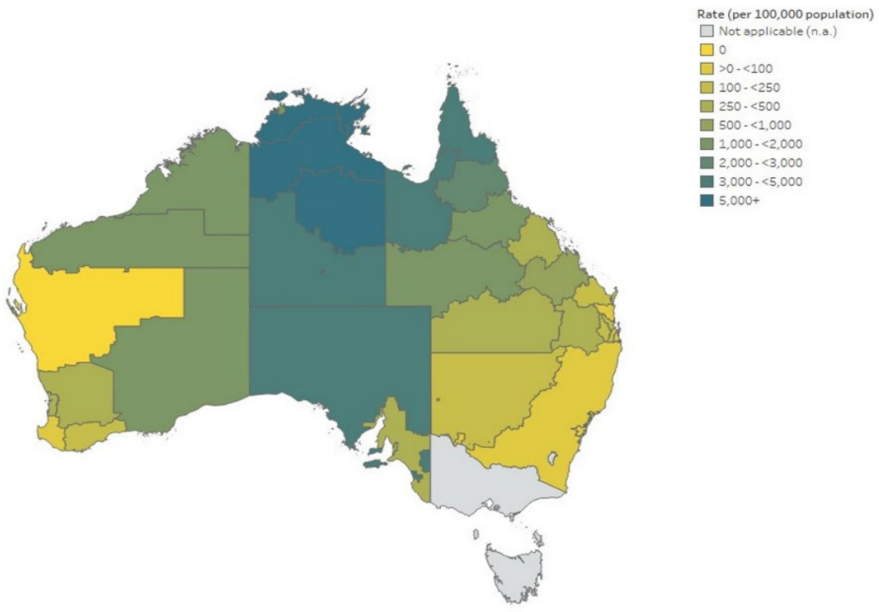


Figure 3.4. ARF and/or RHD diagnoses among First Nations people, by region of management, 31 December 2022
SOURCE: Australia Institute of Health and Welfare, 2024

Primordial prevention and social determinants of acute rheumatic fever

KEY INFORMATION

- The socioeconomic and political factors that influence people's lives can cause structural barriers and inequities in health. These social determinants of health within an Indigenous cultural context have profound impacts on health and wellbeing.
- The circumstances in which people live affect the risk of Group A streptococcus (Strep A) infections, acute rheumatic fever (ARF) and rheumatic heart disease (RHD). Household crowding and limited access to facilities to wash people, clothes and bedding increase the risk of Strep A infections, ARF and RHD.[1]
- Strep A is a human-only infection with no animal or insect hosts; therefore control strategies comprise modifications of human behaviours and environments.
- Nine Healthy Living Practices were developed in the 1980s by the Nganampa Health Council in South Australia to help prioritise what people need to live healthy lives (Table 4.1). There is evidence that the Healthy Living Practices can help reduce Strep A infections.
- While not all Healthy Living Practices apply directly to Strep A, the approach to social determinants of health should be holistic rather than disease specific.
- There are several approaches to increase access to Healthy Living Practices to reduce the development of Strep A skin and throat infections which lead to ARF and RHD.
- Interventions on living practices applied for ARF and RHD are likely to have an impact on other diseases and conditions.

Culture and workforce considerations to help address Social Determinants of Health in the context of ARF and RHD include:

Locally acceptable and feasible strategies to address the Social Determinants of Health.

Dedicated First Nations Environmental Health Workers employed in high-risk communities.

- Environmental health and housing assessment and action for people with Strep A infections.

Culturally appropriate, respectful and practical information and support for reducing risk factors for Strep A infections available where required/requested.

- Hand and body washing promoted by school, education and housing sectors.
- Provision of adequate washing hardware for people, clothes and bedding.
- Health and government services work with community groups to address the environmental and social determinants of health which drive Strep A infections.

Established cross-sector collaboration of departments and activities.

Table 4.1. Healthy Living Practices and their association with reducing Strep A infections, ARF and RHD

HEALTHY LIVING PRACTICE	ASSOCIATION WITH REDUCING STREP A INFECTIONS	NOTES
1 – Washing people	Strong	Washing of hands and bodies, particularly for children, is clearly associated with a reduction in the risk of Strep A infections.
2 – Washing clothes and bedding	Medium	Washing clothing and bedding is an important way to reduce the risk of Strep A skin infections. • Washing clothes and bedding does not directly reduce the risk of Strep A skin infections. • Washing clothes and bedding can reduce the transmission of scabies mites and lice which can cause skin itch and skin damage which lead to Strep A skin infection.
3 – Removing wastewater safely	Weak	Removing wastewater safely is important to reduce the risk of many infectious diseases. • Wastewater is not a major contributor to the spread of Strep A infections.
4 – Improving nutrition, the ability to store, prepare and cook food	Weak	Improving nutrition is important to improve many health outcomes. • Poor nutrition is not known to be a major risk factor for Strep A infection. • Strep A throat infections can spread through food which has not been cooked or stored properly. This is rare and not a major driver of ARF and RHD in Australia.
5 – Reducing the negative impacts of overcrowding	Strong	While households accommodating large family or social groups promotes health and wellbeing in many cultures, overcrowding is a major contributor to the burden of Strep A, ARF and RHD. • Efforts to reduce household overcrowding or reduce the risk of overcrowded living circumstances are important.
6 – Reducing the negative effects of animals, insects and vermin	Medium (Indirect)	Reducing the rates of skin infestation and damage from animals, insects and scabies are important for reducing the risk of Strep A skin infections. • Strep A only infects humans; dogs and insects do not directly spread Strep A infection. • Animals, insects and scabies mites can cause skin damage which increase the risk of secondary Strep A infection.
7 – Reducing the health impacts of dust	Weak	Dust does not contribute to Strep A infections and does not play a major part in reducing the risk of skin or throat infection.
8 – Controlling the temperature of the living environment	Weak	The risk of Strep A infections may be different in hot, wet weather or cold temperatures when people need to sleep close together for warmth. There can be differentials between rates of throat Strep A and skin Strep A based on temperature and humidity. • The evidence for these associations is variable and there is no clear evidence that controlling household temperature can have a significant impact on Strep A, ARF and RHD risk.
9 – Reducing hazards that cause trauma	Medium	Clean and tidy houses and yards may help reduce Strep A skin infections. • Living in a house with lots of rubbish and debris may increase the risk of skin damage through scratches or abrasions. These can become infected with Strep A.

Primary prevention

KEY INFORMATION

- Primary prevention of ARF aims to interrupt the link between Strep A infection and the abnormal immune response to Strep A that causes ARF by early identification and treatment of Strep A infections.
- Strep A has been shown to be associated with up to 37% of sore throats. Strep A is only one cause for tonsillitis. Strep A is present in 10% to 40% of children presenting with a sore throat.
- Treatment of the Strep A sore throat in those at risk of ARF can decrease the subsequent development of ARF by up to two-thirds.[3]
- Strep A has been shown to be associated with most impetigo episodes. Strep A impetigo is very common among First Nations children living in remote areas of Australia, with almost one in two affected at any one time. Identification, treatment and prevention of Strep A skin infections may help reduce the burden of ARF.
- Antibiotic management of sore throat differs from antibiotic management of skin sores.
- Individuals already receiving BPG secondary prophylaxis for ARF still need active treatment of sore throats or skin sores. This is necessary because the level of penicillin achieved by BPG wanes by about 7 days to reach a prophylactic level which is lower than a required treatment level. If the last BPG dose was ≥7 days ago, provide antibiotic dosing in accordance with Table 5.2 for sore throat or Table 5.3 for skin sores.
- Strep A in the throat is transmissible to others in the absence of symptoms.[6]
- All superficial Strep A infections of the skin and throat need treatment to reduce ARF risk.
- Skin sores should be covered to prevent cross-infection.
- Molecular point of care tools aid in the timely detection and treatment of Strep A in the throat and may be a useful adjunct in prevention of ARF.
- Molecular point of care testing is not needed for impetigo, as the presence of impetigo confirms Strep A is involved, and treatment of impetigo is required.

Table 5.1. Summary of recommendations with GRADE Level of Evidence

RECOMMENDATIONS	GRADE
Antibiotics should be given empirically for people at high risk of ARF presenting with sore throat	2B
Antibiotics should be given empirically to people at high risk of ARF presenting with one or more skin sores	1A
Azithromycin is the first recommended oral treatment of sore throat if IM injection is not possible	2B
Individuals already receiving BPG secondary prophylaxis still need active treatment of sore throats or skin sores	2D
Clinical scoring of sore throat is not recommended in Australia	2D
Cotrimoxazole is the first line treatment for skin sores	1A

Table 5.2. Risk groups for primary prevention of ARF

At high risk	Living in an ARF-endemic setting† First Nations peoples living in rural or remote settings First Nations peoples, and Māori and/or Pacific Islander peoples living in metropolitan households affected by crowding and/or lower socioeconomic status Personal history of ARF/RHD and aged <40 years
May be at high risk	Family or household recent history of ARF/RHD Household overcrowding (>2 people per bedroom) or low socioeconomic status Migrant or refugee from low- or middle-income country and their children
Additional considerations which increase risk	Prior residence in a high ARF risk setting Frequent or recent travel to a high ARF risk setting Aged 5–20 years (the peak years for ARF)

† This refers to populations where community ARF/RHD rates are known to be high e.g. ARF incidence >30/100,000 per year in 5–14-year-olds or RHD all-age prevalence >2/1000 (Figure 3.4)

Health staff will often be more familiar with guidelines that prevent overuse of antibiotics for sore throats. Therefore, awareness of the need for antibiotics to treat sore throats to prevent ARF is an important learning point for all health staff working with populations at high risk of ARF.

Empirical antibiotic treatment of all Australian children presenting with a sore throat is not recommended. In addition to the unwarranted inconvenience, there is cost and potential risk from adverse medication events, while increasing the pressures that promote antibiotic resistance.

People presenting with sore throat who are identified as being at high risk for ARF (Table 5.2), including people who have a history of confirmed ARF or established RHD, should be treated with antibiotics if they develop a sore throat, irrespective of other clinical features, and irrespective (at present) of any testing using rapid tests or culture for confirmation of Strep A infection.

Table 5.3. Recommended antibiotic treatment for Strep A sore throat / tonsillitis†

DRUG	DOSE		ROUTE	DURATION
All cases				
Benzathine benzylpenicillin G (BPG)‡	Weight (kg) Child: <10 10 to <20 ≥20 Adult: ≥20	Dose in units (mL)§ 450,000 units (0.9 mL) 600,000 units (1.2 mL) 1,200,000 units (2.3 mL) 1,200,000 units (2.3 mL)	Deep IM injection	Once
If IM injection not possible:				
Azithromycin	Child: 12 mg/kg up to 500 mg, daily Adult: 500 mg daily		Oral	For 5 days
Phenoxymethylpenicillin	Child: 15 mg/kg up to 500 mg, twice daily Adult: 500 mg, twice daily		Oral	For 10 days
For patients with documented hypersensitivity to penicillin e.g. rash				
Cefalexin	Child: 25 mg/kg up to 1 g, twice daily Adult: 1 g, bd			For 10 days

† Antibiotic treatment indicated for proven Strep A infection, and for people at high risk of ARF presenting with sore throat.

‡ For information on managing injection pain, see Chapter 10. Secondary Prevention. During times of rationing of premix BPG supplies due to interruption in supply, existing premix stock should not be used for the treatment of sore throats. Instead, use phenoxymethylpenicillin as first line treatment.

§ mL is only relevant for the premix product. Volumes of powdered BPG may vary.

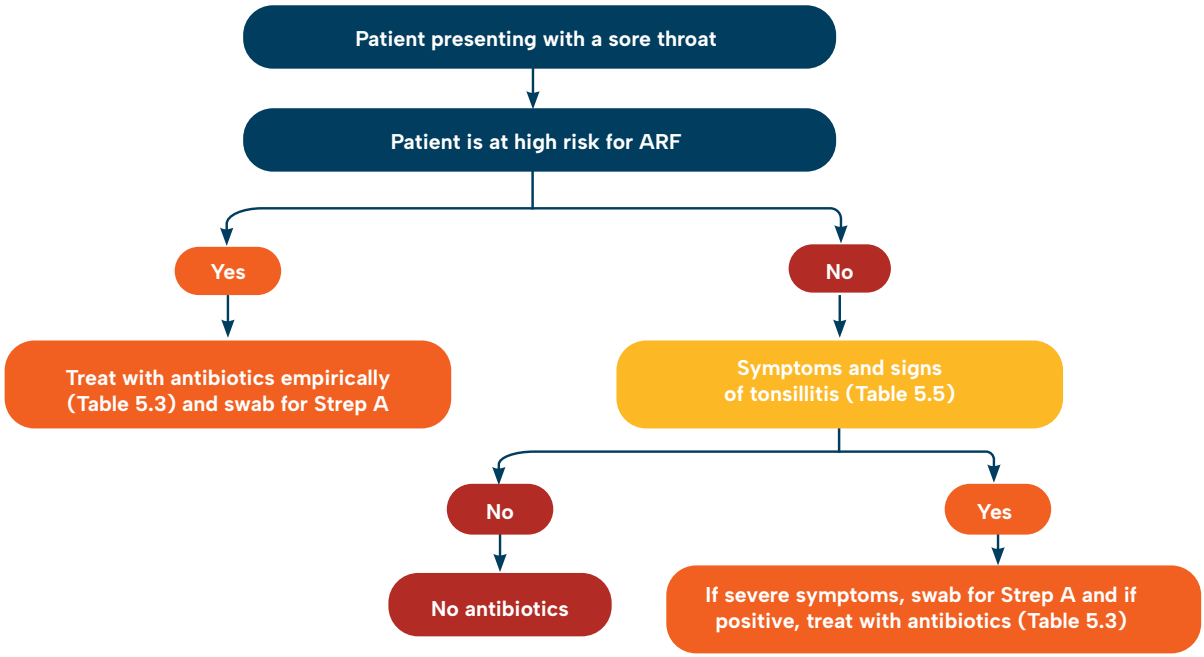


Figure 5.3. Assessment of sore throat

Table 5.4. Recommended antibiotic treatment for Strep A skin sores[†]

DRUG	WEIGHT RANGE	DOSE			ROUTE	DURATION
	For ≥1 purulent or crusted sore(s)					
Cotrimoxazole (trimethoprim / sulfamethoxazole) 4 mg/kg/dose trimethoprim component	**Weight range**	**Syrup dose (40 mg/5 mL)** [§]	**Tablet dose SS (80/400 mg)** [†]	**Tablet dose DS (160/800 mg)** [‡]	Oral	Morning and night for 3 days
	3–<6 kg	12 mg (1.5 mL)	N/A	N/A		
	6–<8 kg	24 mg (3 mL)	¼ tablet			
	8–<10 kg	32 mg (4 mL)	½ tablet			
	10–<12 kg	40 mg (5 mL)				
	12–<16 kg	48 mg (6 mL)	¾ tablet			
	16–<20 kg	64 mg (8 mL)				
	20–<25 kg	80 mg (10 mL)	1 tablet	½ tablet		
	25–<32 kg	100 mg (12.5 mL)	1 ½ tablets	¾ tablet		
	32–<40 kg	128 mg (16 mL)				
	≥40kg	160 mg (20 mL)	2 tablets	1 tablet		
Benzathine benzylpenicillin G (BPG)	Weight Child: <10 kg 10 to <20 kg ≥20 kg Adult: ≥20 kg			Dose in units (mL)[§] 450,000 units (0.9 mL) 600,000 units (1.2 mL) 1,200,000 units (2.3 mL) 1,200,000 units (2.3 mL)	Deep IM injection	Once

† Antibiotic treatment is indicated for all people with one or more lesions with pus or crust.

‡ Cotrimoxazole comes as syrup (40 mg trimethoprim/5 mL) and tablets. The tablets are single strength (SS) (80/400 mg trimethoprim/ sulfamethoxazole) or double strength (DS) (160/800 mg trimethoprim/ sulfamethoxazole). When syrup is unavailable, tablets may be crushed and dissolved in water for small children as per the table above.

§ mL is only relevant for the premix product. Volumes of powdered BPG may vary.

Diagnosis of acute rheumatic fever

KEY INFORMATION

- Diagnosis of ARF results leads to opportunity for secondary prophylaxis with benzathine benzylpenicillin G (BPG), which can prevent recurrent ARF and cumulative heart valve damage.
- Patients with sterile joint aspirates should never be treated speculatively for septic arthritis without further investigation, particularly in areas with a high ARF/RHD prevalence.
- Over-diagnosis results in the individual receiving BPG injections unnecessarily and an increased use of health system resources.
- Recurrent definite, probable or possible ARF is generally not diagnosed until more than 90 days after a previous episode of ARF, to account for prolonged or rebound symptoms related to the original episode.
- Ideally, anyone suspected to have ARF should be admitted to a hospital as soon as possible for specialist review and echocardiography. However, some people may be managed within the community on the advice of a medical specialist.
- Echocardiogram is mandatory for all people suspected and confirmed with ARF. Echocardiogram can enable a confirmation of ARF by demonstrating carditis which may not be clinically evident. It is also used to establish a baseline of cardiac status, and to determine whether valve damage (acute carditis or established RHD) is present and if so, to determine the severity.
- Electrocardiogram is also mandatory for all suspected and confirmed ARF. While first degree heart block (prolonged P-R interval) is most common, advanced conduction abnormalities (second-degree heart block, complete heart block or accelerated junctional rhythm) occur in approximately 8% of those presenting with ARF (Figures 6.9 to 6.11) and are highly specific for ARF, in the presence of other ARF diagnostic criteria.1
- An ARF diagnosis calculator app is available to support clinicians in the diagnosis of ARF.
- For each episode of suspected ARF, a final diagnosis should be reached and specified as either:
    - Definite ARF (confirmed).
    - Probable ARF (highly suspected).
    - Possible ARF (uncertain.
    - Definite ARF recurrence.
    - Probable ARF recurrence.
    - Possible ARF recurrence.
    - Not ARF.
- The final diagnosis and age of the patient determines the subsequent management recommendations, including need for and duration of secondary prophylaxis with BPG; frequency of follow-up echocardiograms; and frequency of primary care and specialist reviews (Table 10.3, Table 7.4).

Table 6.1. Summary of recommendations with GRADE Level of Evidence

RECOMMENDATION	GRADE
Anyone suspected to have ARF who is high risk for complications should be admitted to a hospital within 24–72 hours for echocardiography and specialist review.	1B
Echocardiogram is indicated for all suspected and confirmed ARF as a baseline and to determine underlying carditis or valve damage.	1A
Echocardiography is more sensitive and specific for acute rheumatic carditis than auscultation.	1A
Electrocardiogram (ECG) is indicated for all suspected and confirmed ARF.	1A
Testing CRP alone can result in a missed diagnosis of definite ARF when ESR is ≥30 mm/h but CRP is <30 mg/L.	1B
Molecular methods of Strep A detection, including rapid point-of-care tests, are more sensitive than culture.	1C
Many of the clinical features of ARF are non-specific, so a wide range of differential diagnoses should be considered.	1A
Streptococcal antibody titres are the most useful modality for detecting a recent Strep A infection if bacterial culture is negative.	1C
Streptococcal titre should be determined in the acute phase, and then in the convalescent phase 14–28 days later, with a positive result defined as a rise in titre of twofold or more.	2A
If only a single pathological specimen is available, a streptococcal titre greater than the ULN at initial testing be considered presumptive evidence of a preceding Strep A infection.	1C
Sydenham chorea alone is enough to confirm ARF provided other causes of chorea are excluded	1A
Suspected ARF (without Sydenham chorea) requires elevated serum streptococcal serology demonstrated to enable confirmation of a diagnosis of ARF.	1C
Patients with sterile joint aspirates should be investigated further, particularly in areas with a high ARF/RHD prevalence.	1B
A definite history of arthritis is sufficient to satisfy arthritis as a major manifestation of ARF.	1C
Monoarthritis is a major manifestation in high risk groups.	1C
Monoarthritis should always be considered in the differential diagnosis of ARF for people in high-risk populations.	1A
Withholding NSAIDs in patients with monoarthralgia or monoarthritis, to observe the development of polyarthritis, can also help in confirming a diagnosis of ARF.	1C
Post-streptococcal reactive arthritis does not carry a risk of carditis, and does not require secondary prophylaxis treatment	1B
Echocardiography is essential for all patients with Sydenham chorea	1A

Table 6.2. Risk groups for ARF

At high risk	Living in an ARF-endemic setting†
	First Nations peoples living in rural or remote settings
	First Nations peoples, and Māori and/or Pacific Islander peoples living in metropolitan households affected by crowding and/or lower socioeconomic status
	Personal history of ARF/RHD and aged <40 years
May be at high risk	Family or household recent history of ARF/RHD
	Household overcrowding (>2 people per bedroom) or low socioeconomic status
	Migrant or refugee from low- or middle-income country and their children
Additional considerations which increase risk	Prior residence in a high ARF risk setting
	Frequent or recent travel to a high ARF risk setting
	Aged 5–20 years (the peak years for ARF)

† This refers to populations where community ARF/RHD rates are known to be high e.g. ARF incidence >30/100,000 per year in 5–14-year-olds or RHD all-age prevalence >2/1000 (Figure 3.4).

Table 6.3. Australian criteria for ARF diagnosis

	HIGH-RISK GROUPS†	LOW-RISK GROUPS
Definite initial episode of ARF	2 major manifestations + evidence of preceding Strep A infection, or 1 major + 2 minor manifestations + evidence of preceding Strep A infection‡	
Definite recurrent§ episode of ARF in a patient with a documented history of ARF or RHD	2 major manifestations + evidence of preceding Strep A infection, or 1 major + 2 minor manifestations + evidence of preceding Strep A infection‡, or 3 minor manifestations + evidence of a preceding Strep A infection‡	
Probable or possible ARF (first episode or recurrence§)	A clinical presentation in which ARF is considered a likely diagnosis but falls short in meeting the criteria by either: • one major or one minor manifestation, or • no evidence of preceding Strep A infection (streptococcal titres within normal limits or titres not measured) Such cases should be further categorised according to the level of confidence with which the diagnosis is made: • Probable ARF (previously termed 'probable: highly suspected') • Possible ARF (previously termed 'probable: uncertain')	
Major manifestations	Carditis (including subclinical evidence of rheumatic valvulitis on echocardiogram) Polyarthritis¶ or aseptic monoarthritis or polyarthralgia Sydenham chorea†† Erythema marginatum‡‡ Subcutaneous nodules	Carditis (including subclinical evidence of rheumatic valvulitis on echocardiogram) Polyarthritis¶ Sydenham chorea†† Erythema marginatum‡‡ Subcutaneous nodules
Minor Manifestations	Fever ≥38°C§§ Monoarthralgia¶¶ ESR ≥30 mm/h or CRP ≥30 mg/L Prolonged P–R interval or advanced conduction abnormalities on ECG††† ‡‡‡	Fever ≥38.5°C Polyarthralgia or aseptic monoarthritis¶¶ ESR ≥60 mm/h or CRP ≥30 mg/L Prolonged P–R interval or advanced conduction abnormalities 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). First Nations peoples living in rural or remote settings are known to be at high risk. Data are not available for other populations but First Nations peoples living in urban settings, Māori and Pacific Islanders, and potentially immigrants from developing countries, may also be at high risk.

‡ Elevated or rising antistreptolysin O or Anti-DNase B, or a positive throat culture or rapid antigen or nucleic acid test for preceding Strep A infection.

§ Recurrent definite, probable or possible ARF requires a time period of more than 90 days after the onset of symptoms from the previous episode of definite, probable or possible ARF.

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

†† Chorea does not require other manifestations or evidence of preceding Strep A infection, provided other causes of chorea are excluded.

‡‡ Care should be taken not to label other rashes, particularly non-specific viral exanthems, as erythema marginatum.

§§ In high-risk groups, fever can be considered a minor manifestation based on a reliable history (in the absence of documented temperature) if anti-inflammatory medication has already been administered.

¶¶ If polyarthritis is present as a major criterion, monoarthritis or arthralgia cannot be considered an additional minor manifestation.

††† Advanced conduction abnormalities include second-degree heart block, complete heart block or accelerated junctional rhythm.

‡‡‡ If carditis is present as a major manifestation, a prolonged P–R interval cannot be considered an additional minor manifestation.

Table 6.4. Suggested upper limits of normal (ULN) for serum streptococcal antibody titres in children and adults

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 B: antideoxyribonuclease B, ASO: antistreptolysin O, ULN: upper limit of normal.

Table 6.5. Upper limits of normal for P–R interval

AGE GROUP (YEARS)	SECONDS
3–11	0.16
12–16	0.18
17+	0.20

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

ARF can be difficult to diagnose, and some people are at higher risk of complications. Generally, ARF and probable ARF should be initially managed in hospital to facilitate access to timely workup – most importantly echocardiography, to commence clinical management and education, and to plan follow up.

Health staff moving to, and working in, areas with high rates of ARF and RHD (e.g. remote locations) should receive appropriate training on identifying and managing people suspected to have ARF. Regular workforce education should be provided to health staff working with First Nations communities.

Table 6.7. Key points in identifying major manifestations of ARF

MANIFESTATION	POINTS FOR DIAGNOSIS
Arthritis	Most common presenting symptom of ARF Usually extremely painful Polyarthritis (or polyarthralgia) is usually asymmetrical and migratory but can be additive Monoarthritis may be a presenting feature in high-risk populations Large joints are usually affected, especially knees and ankles Should respond within three days of starting NSAID therapy, including aspirin
Sydenham chorea	Present in up to one-quarter of ARF presentations, particularly females, and predominantly in adolescence Consists of jerky, uncoordinated movements, especially affecting the hands, feet, tongue and face, disappears during sleep Neuropsychiatric features such as decreased school performance, emotional lability, anxiety and obsessional disorder may be present Echocardiography is essential for all patients with chorea
Carditis	Asymptomatic unless moderate or severe If moderate or severe, usually presents clinically as an apical holosystolic (pansystolic) murmur (MR), and/or an early diastolic murmur at the base of the heart or left sternal edge (AR) May only be detected using echocardiography (subclinical carditis)
Subcutaneous nodules	Rare, but highly specific, manifestation 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 the vertebrae
Erythema marginatum	Extremely rare, as well as difficult to detect in darker skin pigmentation; highly specific for ARF Occurs as circular patterns of bright pink macules or papules on the trunk and proximal extremities

NSAID, non-steroidal anti-inflammatory drug; MR, mitral regurgitation; AR, aortic regurgitation.

Table 6.8. Key points in identifying minor manifestations of ARF

MANIFESTATION	POINTS FOR IDENTIFICATION
Arthralgia	Suggestive of ARF if the arthralgia occurs in the same pattern as rheumatic polyarthritis (migratory, asymmetrical, affecting large joints).
Fever	Most manifestations of ARF are accompanied by fever (which can be low-grade and transient.) Oral, tympanic or rectal temperature ≥38ºC (high-risk groups) or ≥38.5ºC (low-risk groups) on/after admission or documented with a reliable history during the current illness (high-risk groups only), should be considered as fever.
Elevated acute-phase reactants	Serum CRP level of ≥30 mg/L (both high-risk and low-risk groups) or ESR of ≥30 mm/h (high-risk groups) or ≥60 mm/h (low-risk groups) meets this diagnostic criterion.
Prolonged PR interval	If a prolonged P-R interval or a more advanced conduction abnormality is detected on ECG, the ECG should be repeated daily during ARF (as often evolves in the acute process), then if still abnormal on discharge, on each follow up visit until normal. If the P-R interval or a more advanced conduction abnormality has returned to normal, ARF becomes a more likely diagnosis.

ECG, electrocardiogram; ESR, erythrocyte sedimentation rate.

Table 6.9. Differential diagnoses of common major presentations of ARF

PRESENTATION		
Polyarthritis and fever	**Carditis**	**Sydenham 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 including Dengue and Chikungunya§	Congenital heart disease	Wilson's disease
Reactive arthropathy§	Infective endocarditis	Tic disorder††
Malaria¶	Hypertrophic cardiomyopathy	Choreoathetoid cerebral palsy
Lyme disease¶	Myocarditis: viral or idiopathic	Encephalitis
Sickle cell anaemia	Pericarditis: viral or idiopathic	Familial chorea (including Huntington's)
Infective endocarditis§§		Intracranial tumour
Leukaemia or lymphoma		Lyme disease¶
Gout and pseudo-gout		Hormonal‡‡

† Gonorrhoea should be actively sought in all potentially sexually active cases. Tests for gonorrhoea include microscopy and culture and polymerase chain reaction (PCR) of joint aspirate, endocervical swab, or first-pass 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. Note that systemic lupus erythematosus occurs at a significantly higher rate in the northern Australian First Nations population than in the non-Indigenous population.

§ Ross River Virus, Barmah Forest Virus, Mycoplasma, cytomegalovirus, Epstein–Barr virus, parvovirus, chlamydia, hepatitis, rubella vaccination, and Yersinia spp. and other gastrointestinal pathogens.

¶ If these conditions occur locally, or there is a relevant travel history.

†† Tourette's syndrome and possibly including PANDAS (paediatric autoimmune neuropsychiatric disorder associated with streptococcal infection).

‡‡ Includes oral contraceptives, hyperthyroidism and hypoparathyroidism.

§§ Ensure separate sets of blood cultures are collected and stigmata of endocarditis are investigated.

Table 6.10. Uses of echocardiography in ARF

Valvulitis
Define the severity of mitral, aortic and/or tricuspid regurgitation.
Define the severity of mixed valve disease (mixed stenotic and regurgitant).
Identify subclinical evidence of rheumatic valve damage.
Visualise valvular anatomy and define mechanism of regurgitation (prolapse, flail leaflet, annular dilatation etc).
Cardiac function
Assess left ventricular size and function.
Pericarditis
Confirm the presence of a pericardial effusion.
Reveal inaudible or subclinical valvular regurgitation in the presence of a friction rub.
Exclude other causes of cardiac murmur
Identify congenital heart disease, such as bicuspid aortic valve and congenital mitral valve anomalies, as the cause for a pathological murmur.
Confirm normal valvular function and morphology in the presence of flow or innocent murmurs.

Table 6.11. Minimal echocardiographic criteria to allow a diagnosis of pathological valvular regurgitation

Pathological (at least mild) MR (all criteria must be met)	**Pathological (at least mild) AR** (all criteria must be met)
1. Seen in at least 2 views 2. In at least one view, MR jet length measures ≥1.5 cm (in individuals weighing <30 kg) or ≥2.0 cm (in individuals weighing ≥30 kg)† 3. Velocity ≥3 m/sec for one complete envelope‡§ 4. Pan-systolic jet in at least one envelope†† §	1. Seen in at least 2 views 2. In at least one view jet length ≥1 cm‡‡† 3. Velocity ≥3 m/sec in early diastole§ 4. Pan-diastolic jet in at least one envelope§

† Cut off is based on expert consensus. If weight is not available, then an age cut off at ≤10 years or >10 years can be applied)

AR, aortic regurgitation; MR, mitral regurgitation.

‡ Body of the pansystolic envelope should be ≥3m/s

§ It is reasonable to use separate Continuous Wave Doppler traces seen in different views to document the pansystolic / pan-diastolic envelope and jet velocity.

†† Given the difficulty in aligning spectral Doppler through eccentric regurgitant jets, it is reasonable to use an appearance of a jet being pan-systolic based on qualitative assessment, such as colour jet seen throughout systole.

‡‡ A regurgitant jet length should be measured from the vena-contracta to the last pixel of regurgitant colour (blue or red).

AR, aortic regurgitation; MR, mitral regurgitation.

Management of acute rheumatic fever

KEY INFORMATION

- People suspected to have acute rheumatic fever (ARF) should be referred as soon as possible for investigations (including echocardiography), treatment and education.
- Admission to a hospital with echocardiography services is generally recommended in order to facilitate correct diagnosis. Echocardiographic findings inform the management plan including recommended duration of secondary prophylaxis. A normal echocardiogram does not exclude ARF.
- While the diagnosis is uncertain, giving salicylate or non-steroidal anti-inflammatory drug (NSAID) therapy should be deferred because they might mask symptom evolution, and thereby impede correct diagnosis.
- Suspected ARF' is a term that applies during diagnostic workup. For each ARF episode, a final diagnosis should be reached and specified as either:
    - Definite ARF (initial or recurrence);
    - Probable ARF (initial or recurrence);
    - Possible ARF (initial or recurrence);
    - Not ARF.
- For definite ARF, a 'priority' grade 1 through 4 based on the presence and severity of any accompanying RHD should also be provided, using the revised priority classification (Table 7.4). The priority determines which care plan to use, including frequency of medical reviews and echocardiograms.
- The 'priority' grade and recommended follow up schedule represents best practice according to expert consensus opinion for the Australian context (GRADE 1D). These acknowledge that ongoing advocacy is needed to achieve levels of servicing to meet recommendations.
- People diagnosed with ARF should be:
    - Notified to the local Disease Control Unit or Public Health Unit in accordance with jurisdictional legislation (Table 13.1); and
    - Registered with the jurisdictional RHD Control Program, with details of their secondary prophylaxis requirements (Table 10.2).
- The pillars of management are
    - Eradication of the inciting infection using antibiotics as directed (penicillin or azithromycin for initial prevention, and penicillin or an alternative if allergic to penicillin)
    - Management of symptoms with analgesic / antipyretic agents as needed.

Table 7.1. Medications used for acute rheumatic fever

INDICATION	MEDICATION OPTIONS LISTED IN ORDER OF PREFERENCE	COMMENT	GRADE
Eradication of inciting streptococcal infection	1. Benzathine benzylpenicillin G (BPG) 1,200,000 units (child <20 kg: 600,000 units; ≥20 kg: 1,200,000 units) IMI single dose or	Streptococcal infection may not be evident by the time ARF manifests (e.g. cultures often negative) but eradication therapy for possible persisting streptococci is recommended, nonetheless. Intramuscular penicillin is preferred due to better adherence and its ongoing use in secondary prophylaxis.	1B
	2. Phenoxymethylpenicillin 500 mg (child: 15 mg/kg up to 500 mg) orally 12-hourly for 10 days		
	3. Penicillin hypersensitivity (non-severe): cefalexin 1 g (child: 25 mg/kg up to 1 g) orally, 12-hourly for 10 days	Between 3 and 30% of Group A Streptococcus isolates internationally are resistant to macrolide antibiotics (e.g. azithromycin).	
	4. Severe penicillin hypersensitivity: azithromycin 500 mg (child: 12 mg/kg up to 500 mg) orally daily for 5 days		
Initial analgesia while awaiting diagnostic confirmation: mild-moderate pain	Paracetamol 1000 mg (child 15 mg/kg) orally, 4-hourly up to a maximum of 60 mg/kg/day or 4000 mg/day	Preferred initial analgesia during diagnostic uncertainty, to avoid the masking effect that anti-inflammatory use can have on migratory joint symptoms, fever and inflammatory markers.	2D
Symptomatic management of arthritis/arthralgia after confirmation of ARF diagnosis	1. Naproxen immediate-release 250–500 mg (child 10–20 mg/kg/day) orally twice daily or	Naproxen may be safer than aspirin, and convenient due to twice daily dosing and the capability oral suspension. Ibuprofen is well tolerated and readily available but data and experience with its use are less in ARF than for naproxen. The dose of NSAIDS needed for ARF is generally higher than the dose recommended for other conditions, therefore it may be appropriate to commence at the higher dose range. Due to the rare possibility of Reye's syndrome in children, aspirin may need to be ceased during intercurrent acute viral illness, and influenza vaccination is strongly recommended.	1B
	2. Ibuprofen 200–400 mg (child 5–10 mg/kg) orally three times daily or		
	3. Aspirin adults and children 50–60 mg/kg/day orally, in four to five divided doses. Dose can be escalated up to a maximum of 80–100 mg/kg/day in four to five divided doses		
Symptomatic management of moderate to severe chorea / chorea paralytica (Table 7.6)	1. Carbamazepine 3.5 to 10 mg/kg per dose orally, twice daily 2. Sodium valproate 7.5 to 10 mg/kg per dose orally, twice daily Plus 1. Prednisone/prednisolone 1 to 2 mg/kg up to a maximum of 80 mg orally, once daily or in divided doses Plus consider intravenous immunoglobulin therapy or plasma exchange	Treatment of Sydenham chorea should be considered if movements interfere substantially with normal activities.	2B

Table 7.1. Medications used for acute rheumatic fever (continued)

INDICATION	MEDICATION OPTIONS LISTED IN ORDER OF PREFERENCE	COMMENT	GRADE
Symptomatic management of carditis	Paediatric dosing: Furosemide (frusemide) 1 to 2 mg/kg orally as a single dose, then 0.5 to 1 mg/kg (to a maximum of 6 mg/kg) orally, 6- to 24-hourly Spironolactone 1 to 3 mg/kg (initially) up to 100 mg orally, daily in 1 to 3 divided doses. Round dose to a multiple of 6.25 mg (a quarter of a 25 mg tablet) Enalapril 0.1 mg/kg orally, daily in 1 or 2 divided doses increased gradually over 2 weeks to a maximum of 1 mg/kg orally, daily in 1 or 2 divided doses, other ACE inhibitors (captopril, lisinopril, ramipril, perindopril)	Treatment of heart failure may be required in severe, acute carditis. Seek advice from a specialist cardiologist. Choice of ACE inhibitor will vary depending on the clinical situation. Seek advice from a specialist cardiologist.	1B
	Adult dosing: Furosemide (frusemide) 20–40 mg oral or intravenous as a single dose followed by 20–40 mg oral or intravenous 8–12 hourly. Ongoing dose adjustment based on clinical progression and renal function Spironolactone may be added for patients having limited or no response to loop diuretic, 12.5–25 mg spironolactone orally daily Nitrate therapy may be added for patients having limited or no response to diuretic therapy, whose systolic blood pressure is greater than 90 mmHg. Intravenous or topical glyceryl trinitrate may be used ACE inhibitor is recommended in patients with moderate or severe left ventricular systolic dysfunction, unless contraindicated	The management of acute carditis follows the same principles as the management of acute heart failure. This table gives a guide to the initial management of acute heart failure due to acute carditis in adults. Seeking advice from a specialist cardiologist early is strongly recommended.	1B
	Digoxin 15 micrograms/kg orally, as a single dose, then 5 micrograms/kg after 6 hours, then 3–5 micrograms/kg (adult: 125–250 micrograms) orally, daily	Digoxin is rarely used in the treatment of acute carditis. Seek advice from a specialist cardiologist.	2C
Disease-modifying (immunomodulatory) treatments	Prednisone/prednisolone 1 to 2 mg/kg up to a maximum of 80 mg orally, once daily or in divided doses	Considered for use in selected cases of severe carditis, despite meta-analyses in which overall benefit was not evident.	2C

IMI, intramuscular injection; NSAID, non-steroidal anti-inflammatory drug; ACE, Angiotensin-converting enzyme

Table 7.2. Priorities in managing ARF in the acute setting

ADMISSION TO HOSPITAL	
Anyone suspected to have ARF who is high risk for complications (See Chapter 6 Diagnosis of ARF, Indications for Hospitalisation).	
DETERMINE THE DIAGNOSIS	
The diagnosis is determined based on • Understanding of epidemiological risk. • History obtained from primary care staff and/or patient and their family. • Clinical observation prior to anti-inflammatory treatment: use paracetamol (first line) during this time if required for fever or joint pain. Investigations (Table 7.3). • Follow up findings o The final diagnosis may not be clear until several months after the acute episode, e.g. if Jones criteria are not met for a diagnosis of definite ARF but a follow up echocardiogram confirms rheumatic valvular changes not visible at the outset, then the diagnosis shifts from possible or probable to definite ARF.	
TREATMENT	
All cases	Provision of supportive, culturally safe care. Antibiotic management using pain avoidance techniques for delivery of intramuscular injection (Figure 10.1). Influenza vaccine – annual influenza vaccination is part of the long-term care plan but needs to be considered acutely as a strategy to reduce the risk of Reye's syndrome for children receiving aspirin.
Arthritis and fever	Paracetamol (first line) until diagnosis confirmed. Naproxen, ibuprofen or aspirin once diagnosis confirmed, if arthritis or severe arthralgia present. Mild arthralgia and fever may respond to paracetamol alone.
Sydenham chorea	No pharmacological treatment for mild cases. Anticonvulsant such as carbamazepine or sodium valproate if indicated (Table 7.1). Stepwise use of other agents as per text below (Table 7.6). Evidence base is limited.
Carditis/heart failure	Bed rest, with mobilisation as symptoms permit. Anti-failure medication as required (Table 7.1). Corticosteroids for severe carditis or pericarditis with effusion. (Tables 7.1 and 7.5). Valve surgery for life-threatening acute carditis (rare).

Table 7.2. Priorities in managing ARF in the acute setting (continued)

LONG-TERM PREVENTIVE MEASURES AND DISCHARGE PLANNING

Prepare for discharge to primary care facility and follow-up.
• Notify case to the jurisdictional ARF/RHD register (where it exists) (Table 13.1).
• Contact the patient's local primary care service and community pharmacist.
  o Provide a discharge letter to the patient or family, the primary care service and community pharmacist including information about:
  o ARF diagnosis (possible, probable, definite).
  o Priority classification of RHD if also present (Priority 1, 2 or 3) (Table 11.2).
  o A recommended care plan summary based on disease priority classification (Table 7.4).
  o Date of last BPG administration.
  o Required frequency of BPG, and the due date of next dose.
  o Date of next medical appointment.
  o Date of next echocardiogram.
  o Information about vaccinations administered in hospital.
  o Relevant contraception information and/or pregnancy planning for women.
• Arrange dental review and ongoing dental care to reduce risk of endocarditis.
Family and community engagement
• Involve family in care.
• Engage interpreters for patients and families whose first language is not English.
• Provide education that is culturally appropriate and age-appropriate.
• With consent from family, notify school (for school-aged children) to encourage support for ongoing care.
• Acknowledge the significance of a chronic disease diagnosis in childhood, including the need for linkage with peer-support networks, psychological support, ongoing education, transition care as the individual ages, and self-management support. Where indicated, engage adolescent support services (Table 11.4).

Table 7.3. Testing and monitoring of ARF in the acute setting

Investigations	Always request: • Electrocardiogram (ECG). • Echocardiogram. • Full blood count (FBC). • Erythrocyte sedimentation rate (ESR). • C-reactive protein (CRP). • Streptococcal serology (anti-streptolysin O and anti-DNase B). In relevant situations: • Throat swab. • Skin sore swab. • Blood cultures. • Synovial fluid aspirate. o Ensure sample does not clot by using correct tubes which have been well mixed and transported promptly to the laboratory. o Include request for cell count, microscopy, culture and gonococcal polymerase chain reaction (PCR). • Pregnancy test. • Creatinine test (UEC [urea, electrolytes, creatinine]) since NSAIDS can affect renal function. Tests to exclude alternative diagnoses, depending on clinical presentation and locally endemic infections: • Autoantibodies, double-stranded DNA, anti-cyclic citrullinated peptide (anti-CCP) antibodies. • Urine for Neisseria gonorrhoeae molecular test. • Urine for Chlamydia trachomatis molecular test. • Serological or other testing for viral hepatitis, Yersinia spp, cytomegalovirus (CMV), parvovirus B19, respiratory viruses, Ross River virus, Barmah Forest virus.
Clinical observations	Temperature, pulse, respiratory rate, blood pressure 4 times daily. On occasions of rapid or irregular pulse, ensure ECG is recorded. Thorough skin examination for skin sores, erythema marginatum, subcutaneous nodules. Regular assessment of joints.
Diet	Standard healthy diet. Early dietary advice if overweight (especially if in heart failure), to avoid further weight gain. Consider testing lipids, HbA1c. Weekly weight.
If clinical carditis is present	Document cardiac symptoms and signs. Include sleeping pulse in regular nursing observations (e.g. 0200 hours), as long as this can be done without waking the patient. Individuals with heart failure or severe acute valve disease should be encouraged to rest in bed and avoid exertion until symptoms are improving. Daily weight and fluid balance chart. Weekly echocardiograms in severe acute valvulitis while patient is hospitalised, if able.

Table 7.4. Priority classification and recommended follow-up (updated 2025)

DIAGNOSIS	RECOMMENDED FOLLOW-UP PLAN†
Priority 1 **All Stage D RHD**‡ High risk post-valve surgery patients§ ≥ 3 episodes of ARF within the last 5 years Pregnant women with RHD (of any severity) may be considered Priority 1 for the duration of the pregnancy Children ≤ 5 years of age with ARF or RHD	Specialist review: at least 6 monthly Echocardiogram: at least 6 monthly Medical review: at least 6 monthly Pregnant: see Figure 12.1 for care pathway Dental review: within 3 months of diagnosis, then 6 monthly
Priority 2 **Any Stage C RHD**‡ Moderate risk post-valve surgery patients§	Specialist review: 6 monthly – yearly Echocardiogram: 6 monthly – yearly Medical review: 6 monthly Dental review: within 3 months of diagnosis, then 6 monthly
Priority 3 **Any Stage A or B RHD**‡ ARF without carditis or RHD, currently prescribed secondary prophylaxis¶ Low risk post-valve surgical patients§	Specialist review: 1 – 3 yearly Echocardiogram: children ≤21 years: 1–2 yearly, >21 years: 2–3 yearly Medical review: yearly Dental review: yearly
Priority 4 **History of ARF** (possible, probable or definite) and completed secondary prophylaxis **Resolved RHD** and completed secondary prophylaxis††	Specialist review: 1 year, 3 years and 5 years post cessation of secondary prophylaxis Echocardiogram: 1 year, 3 years and 5 years post cessation of secondary prophylaxis Medical review: yearly until discharge from specialist care and then as required Dental review: yearly or as required

† Frequency should be tailored to the individual following specialist assessment. All patients should be given influenza vaccine annually and have completed pneumococcal vaccinations as per Australian Immunisation Handbook. Intervals for medical and specialist review and echocardiography are a guide and may vary for specific individuals. Medical and dental reviews may be combined with general health check-up. People with RHD require endocarditis prevention as indicated. (See Chapter 11. Management of RHD, Prevention of infective endocarditis).

‡ See Table 8.7 for definitions of RHD severity.

§ While post-surgical RHD is by definition severe RHD, post-surgical risk varies for individuals due to age, type of surgery, recurrence of ARF, adherence with secondary prophylaxis and other factors. Priority category for post-surgical RHD varies as listed in this Priority classification table and should be determined by specialist cardiologist/paediatrician/physician. (See Chapter 11. Management of RHD, Monitoring following valve surgery).

¶ See Table 10.2 regarding initial treatment of possible, probable and definite ARF with and without carditis. The priority table provides guidance on longer term established RHD based on Stage of disease once the acute illness has resolved.

†† A proportion of early RHD changes can resolve with no residual valve dysfunction. These cases are referred to as 'resolved RHD' and as such, may not need the longer-term follow-up required by Stage B/C/D disease.

Table 7.6. Summary of Sydenham chorea management strategies

SYDENHAM CHOREA SEVERITY	STEP-WISE MANAGEMENT	MEDICATION DOSES
All cases	Antibiotic treatment (for preceding streptococcal infection and commencement of prophylaxis).	See Table 7.1
Mild – For example: Mild involuntary movements, incoordination &/or neuropsychiatric features . Mild hypotonia or weakness. Minimal functional impairment.	Supportive measures: Calm environment, avoidance of over-stimulation, rest, education about the condition.	None
Moderate – For example: Moderate functional impairment, slightly unsteady gait, some difficulty feeding and other self-care activities. Motor impersistence (inability to maintain or sustain actions such as handgrip, tongue protrusion or outheld hand / arms). Mood change, anxiety, reduced attention, hyperactivity, obsessive compulsive behaviour. Reduced / altered speech.	Above plus: Anticonvulsant therapy usually with **carbamazepine** or **sodium valproate** (if no risk of pregnancy).	Carbamazepine 3.5 to 10 mg/kg per dose orally, twice daily (max 200mg bd) **or** **If no risk of pregnancy e.g. male or pre-pubertal female:** Sodium valproate 7.5 to 10 mg/kg per dose orally, twice daily (max 500mg bd). If significant residual symptoms, consider treatment as for severe below.
Severe – For example: Severe functional impairment, unsteady gait, significant difficulty feeding and other self-care activities. Difficulty sitting. Dysarthria (marked reduced / altered speech). Marked mood change, anx-iety, reduced attention, hy-peractivity, obsessive compulsive behaviour. Chorea paralytica (bedbound, aphasic).	Above plus: There is some evidence for **corticosteroid** use (one of prednisone, prednisolone, or methylprednisolone) in Sydenham chorea or other autoimmune encephalopathy, but the ideal dose and duration is not established. **Haloperidol** has historically been used, but the adverse effects of excessive sedation and extrapyramidal side effects mean that its use is now largely discouraged. In very unwell children, consideration is needed of intravenous **immunoglobulin** or **plasma exchange.**	Corticosteroids for example: Prednisone/prednisolone 1 to 2 mg/kg up to a maximum of 80 mg orally, once daily or in divided doses **or** Methylprednisolone 30 mg/kg/day for 3 days, max 1g/day. Intravenous immunoglobulin 2 g/kg over 5 to 7 days. Plasmapheresis e.g. for 5 days.

Discharge from hospital is a critical point in the patient journey. A health management plan should be developed before discharge, and by a multidisciplinary team which includes the hospital medical officer, nurse, First Nations Liaison Officer, primary healthcare service, and the patient and family. Where possible, the first outpatient medical appointment should be booked prior to hospital discharge.

Diagnosis of rheumatic heart disease

KEY INFORMATION

- In Australia, approximately 87% of rheumatic heart disease (RHD) occurs in First Nations peoples (depending on the dataset used).
- RHD has a female predominance of 2:1 and the prevalence peaks in the third and fourth decade of life.
- RHD should be considered in individuals from high-risk populations (Table 6.1) with reduced exercise tolerance or breathlessness noting that most RHD is asymptomatic.
- Reduced exercise tolerance or breathlessness in a pregnant woman from a high-risk population should not only be attributed to pregnancy or anaemia; consider and investigate for RHD.
- Exercise testing or stress echocardiography is recommended when severity of symptoms and echocardiographic findings are discordant.
- Transoesophageal echocardiography may help clarify valve morphology and severity to plan surgical intervention or when transthoracic echo is inconclusive.
- The first edition of the WHF guidelines on the echocardiographic diagnosis of RHD, published in 2012, have now been revised and published as the 2023 WHF guidelines.
- The 2023 WHF guidelines on echocardiographic diagnosis provide criteria to distinguish pathological RHD from physiological changes in children and adults (Table 8.6).
- The mitral valve is the most common valve involved in RHD.
- Many adult patients will have mixed or multi-valvular disease.
- Symptoms may not reflect severity of disease. Many patients will appear asymptomatic until advanced stages of disease develop.
- Patients may present with complications of valve disease including stroke, infective endocarditis, heart failure or arrhythmia.

Table 8.1. Summary of recommendations with GRADE Level of Evidence

RECOMMENDATION	GRADE
RHD should be considered in individuals from high-risk populations with reduced exercise tolerance or breathlessness.	1C
Echocardiography is the gold standard diagnostic tool for RHD and should be performed in any patient suspected of having RHD.	1A
All patients with murmurs suggestive of possible valve disease, a positive screening echocardiogram, or a history of ARF, require a formal echocardiography to confirm the diagnosis.	1A
Transoesophageal echocardiography may help clarify valve morphology and severity to plan surgical intervention or when transthoracic echo is inconclusive.	1B
In adults, transoesophageal echocardiography can help clarify severity and mechanism of valve lesions, particularly in cases of mixed and multi-valvular disease.	1B
Exercise testing or stress echocardiography is recommended when severity of symptoms and echocardiographic findings are discordant.	1B
The 2023 WHF guidelines for echocardiographic diagnosis of RHD provide criteria to distinguish pathological RHD from physiological changes in children and adults.	1B
Trivial regurgitation of the mitral or aortic valves that does not meet all four criteria for pathological regurgitation should be considered normal.	1C
Isolated morphological changes, such as valvular thickening, which occurs without pathological stenosis or regurgitation should be considered normal.	1C
Coronary angiography is indicated prior to valve surgery to exclude concurrent coronary disease requiring intervention.	1C
For First Nations peoples, invasive angiography is recommended over computed tomography in those over the age of 25 years.	2C
Right heart catheterisation may aid in clarifying valve lesion severity when echocardiographic data are inconclusive.	1C
Right heart catheterisation may be used to determine the predominant cause of pulmonary hypertension when there is clinical ambiguity.	1C

Table 8.2. Clinical features of common valve lesions

VALVE LESION	SYMPTOMS †	SIGNS	COMPLICATIONS
Mitral regurgitation (MR)	Dyspnoea on exertion Fatigue Weakness Orthopnoea, paroxysmal nocturnal dyspnoea	Mid/pan-systolic murmur at apex, radiating laterally (occasionally medially/ posteriorly) Displaced apex beat in severe MR	Congestive cardiac failure Atrial arrhythmia Pulmonary hypertension
Mitral stenosis (MS)	Exertional dyspnoea (symptoms sensitive to increase in heart rate) Orthopnoea, paroxysmal nocturnal dyspnoea Haemoptysis	Low-pitch, diastolic murmur at apex with patient in left lateral position Murmur duration correlates with severity	Atrial arrhythmia Pulmonary hypertension Systemic embolism (stroke, peripheral arterial occlusion)
Aortic regurgitation (AR)	Dyspnoea on exertion Angina Orthopnoea, paroxysmal nocturnal dyspnoea	Blowing decrescendo diastolic murmur at left sternal edge Systolic murmur due to increased flow Mitral diastolic murmur (Austin Flint) Wide pulse pressure	Congestive cardiac failure
Aortic stenosis (AS)	Dyspnoea, angina, presyncope and syncope all associated with exertion	Ejection systolic murmur over aortic region, radiating to neck Slow-rising pulse	Heart failure with preserved or reduced ejection fraction Atrial arrhythmia
Tricuspid regurgitation (TR)	Peripheral oedema Abdominal distention and discomfort	Pan-systolic murmur at left parasternal edge Elevated jugular venous pressure (JVP) with prominent V-waves. Pulsatile liver Right ventricular heave	Right-sided heart failure
Tricuspid stenosis (TS)	Fatigue Abdominal discomfort Anorexia	Soft, high-pitch diastolic murmur at left parasternal edge Abdominal ascites Hepatomegaly Giant A-waves in JVP	Anasarca Hepatomegaly and hepatic dysfunction

† Note that mild to moderate valve dysfunction is most often asymptomatic.

Table 8.3. Echocardiographic features of RHD

CHARACTERISTIC FEATURES	MARKERS OF SEVERE DISEASE
Mitral valve features • Anterior leaflet override • Thickened leaflet tips • Restricted posterior leaflet motion • Chordal thickening • Leaflet calcification • Diastolic doming of anterior leaflet with restriction of the tip ("dog leg" or "hockey stick" appearance)	Mitral regurgitation • Large central jet (>50% of LA) or eccentric wall impinging jet of variable size • CW Doppler jet is pansystolic/dense/triangular • Mitral inflow E-wave dominance (>1.2 m/s) • Vena contracta ≥0.7 cm • Regurgitant volume ≥60 mL/beat • Regurgitant fraction ≥50% • EROA ≥0.40 cm^2 Mitral stenosis: • Valve area by planimetry ≤1.5 cm^2 • Diastolic pressure half-time ≥150 ms • Mean pressure gradient ≥10 mmHg (in the absence of significant MR)
Aortic valve features • Cusp prolapse • Cusp thickening • Rolled cusp edges • Cusp restriction • Cusp fibrosis, retraction, calcification	Aortic regurgitation • Jet width ≥65% of LVOT • Vena contracta ≥0.6 cm • Pandiastolic flow reversal in the proximal abdominal aorta • Regurgitant volume ≥60 mL/beat • Regurgitant fraction ≥50% • EROA ≥0.3 cm^2 • Evidence of LV dilatation Aortic stenosis† • Aortic valve Vmax ≥4 m/s • Mean pressure gradient ≥40 mmHg • Valve area ≤1.0 cm^2
Tricuspid valve features • Leaflet thickening, calcification • Leaflet restriction, retraction • Chordal shortening	Tricuspid regurgitation • Large central jet (≥50% of RA) • Vena contracta width ≥0.7 cm • EROA ≥0.4 cm^2 • Regurgitant volume ≥45 mL/beat • CW Doppler jet is dense, triangular with early peak • Systolic flow reversal in hepatic vein Tricuspid stenosis • Mean pressure gradient ≥5 mmHg • Pressure half-time ≥90 ms • Valve area ≤1.0 cm^2

† Scenarios of low-flow, low-gradient and normal flow, low-gradient severe AS exist. Expert input is advised.
LA, left atrium; EROA, Effective regurgitant orifice area; LVOT, Left Ventricular Outflow Tract; LV, left ventricular; CW, Continuous wave; RA, right atrium.

Table 8.4. Role of cardiac investigations in the diagnosis of RHD

INVESTIGATION	ROLE
Transthoracic echocardiography† (TTE)	Baseline investigation (including screening for RHD) Assessment of valve pathology Assessment of cardiac function and chamber size Surveillance of valve pathology and cardiac function over time
Transoesophageal echocardiography (TOE)	Pre-surgical planning Anatomical assessment for valve repair Exclusion of LA thrombus and significant MR prior to percutaneous balloon mitral valvuloplasty or direct current cardioversion Assessment of valve severity when TTE non-confirmatory
Electrocardiogram†	Identify arrhythmias that may complicate RHD (e.g. atrial fibrillation) Identify structural changes of RHD (e.g. left ventricular hypertrophy, p-mitrale)
Exercise stress test	Objective assessment when valve severity discordant from symptoms
Stress echocardiogram	Objective assessment when valve severity discordant from symptoms Use in MS for assessing change in gradient and pulmonary arterial systolic pressure with exercise
Right heart catheterisation	Assessment of valve severity in cases when TTE/TOE is non-confirmatory Assessment and classification of pulmonary hypertension in setting of valvular disease
Coronary angiography	Exclude concomitant coronary disease pre-surgery (over age 25 years)
Computed tomography coronary angiogram	Exclude concomitant coronary disease pre-surgery (younger than 25 years)
Cardiac magnetic resonance imaging	Role in assessing aetiology of cardiomyopathy and quantifying chamber size and function Quantification of regurgitant volumes
Erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) and anti-streptococcal titres†	In cases of newly diagnosed RHD to exclude possible ARF episode
B-type natriuretic peptide (BNP), pro-NT BNP	Role in assessment of heart failure presentation (See NHFA/CSANZ heart failure guidelines)

† Compulsory in diagnostic work-up

Table 8.5. Diagnostic morphological features of RHD (based on WHF 2023 guidelines)

VALVE	MORPHOLOGICAL FEATURES
Mitral valve	Valve apparatus thickening category (defined by the presence of either or both): Anterior leaflet thickening†‡ Chordal thickening§ Valve mobility abnormalities category (defined by the presence of either or both): Restricted anterior or posterior leaflet motion in diastole¶ Excessive anterior leaflet tip motion during systole††
Aortic valve	Cusp thickening‡‡ Cusp prolapse Restricted cusp motion Coaptation defect in diastole

† Anterior mitral valve leaflet (AMVL) thickness should be measured during diastole at the full excursion. Measurement should be taken at the thickest portion of the leaflet, including focal thickening, beading and nodularity. Ideally, the measurement should be performed on a frame with maximal separation of chordae from the leaflet tissue.

‡ Abnormal thickening of the AMVL is age-specific and defined as follows: ≥3.0 mm for individuals aged ≤20 years, ≥4.0 mm for individuals aged 21–40 years and ≥5.0 mm for individuals aged >40 years.

§ Chordal thickening might range from individual chordae tendineae to multiple chordae tendineae fusion and calcification. The structures of papillary muscles, chordae tendineae and margins of leaflets might not be clearly distinguishable. Assessment is subjective; however, in most cases, chordal thickening is attributable to fusion of two or more chordae and is seen as an echo-bright structure near to the leaflet tip insertion. Chordal thickening is typically associated with other morphological features.

¶ Restricted leaflet motion of either the AMVL or the posterior mitral valve leaflet is usually the result of chordal shortening or fusion, commissural fusion or leaflet thickening.

†† Excessive leaflet tip motion results from elongation of the primary chords and is defined as displacement of the tip or edge of an involved leaflet towards the left atrium, resulting in abnormal coaptation and regurgitation. Excessive leaflet tip motion does not need to meet the standard echocardiographic definition of mitral valve prolapse disease, given that they refer to different disease processes. This characteristic applies only to those aged <35 years.

‡‡ In the parasternal short-axis view, the right and non-coronary aortic cusp closure line often presents as echogenic (thickened) in healthy individuals, which should be considered normal. Image optimization for valve thickness and morphology, including harmonic imaging, should be individualized on the basis of echocardiography devices and those performing echocardiograms.

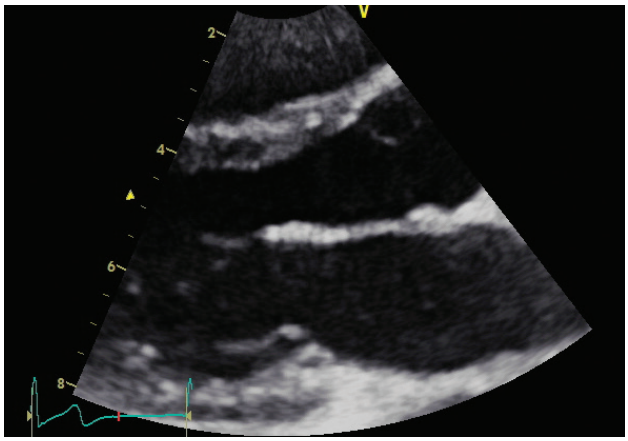


Figure 8.1a. Rheumatic mitral valve; appearance with harmonics 'on', note anterior mitral valve thickness. Harmonics should be turned off

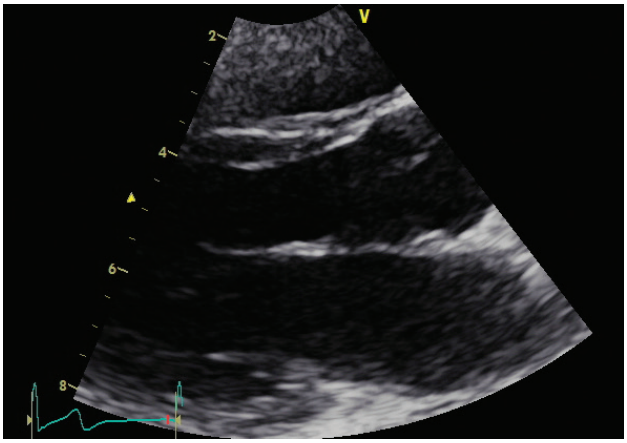


Figure 8.1b. Rheumatic mitral valve; appearance with harmonics 'off', note anterior mitral valve thickness

Table 8.6. Criteria for pathological valve dysfunction (based on WHF 2023 guidelines)

Pathological (at least mild) mitral regurgitation: (ALL criteria to be met) 1. Seen in at least two views. 2. In at least one view, jet length measures ≥1.5 cm (<30 kg) or ≥2.0 cm (≥30 kg)†‡ 3. Velocity ≥3 m/s for one complete envelope§ ¶ 4. Pan-systolic jet in at least one envelope¶ ††
Pathological (at least mild) aortic regurgitation: (ALL criteria to be met) 1. Seen in at least two views. 2. In at least one view, jet length ≥1 cm‡ 3. Velocity ≥3 m/s in early diastole¶ 4. Pan-diastolic jet in at least one envelope¶
Mitral stenosis: (ALL criteria to be met)‡‡ 1. Restricted leaflet motion with reduced valve opening 2. Mean PG ≥4 mmHg

† Cutoff is based on expert consensus. If weight is not available, then an age cutoff of ≤10 years or >10 years can be applied).

‡ A regurgitant jet length should be measured from the vena-contracta to the last pixel of regurgitant colour (blue or red).

§ Body of the pansystolic envelope should be ≥3 m/s.

¶ It is reasonable to use separate Continuous Wave Doppler traces seen in different views to document the pan-systolic / pan-diastolic envelope and jet velocity.

†† Given the difficulty in aligning spectral Doppler through eccentric regurgitant jets, it is reasonable to use an appearance of a jet being pan-systolic based on qualitative assessment, such as a colour jet seen throughout systole.

‡‡ 2D evidence of reduced mitral valve orifice area is adequate when spectral Doppler is unavailable.

Note: It is essential to exclude other common causes of mild valvular regurgitation, including mitral valve prolapse and bicuspid aortic valve, before diagnosing RHD.

PG: pressure gradient, 2D: 2 dimensional

Box 8.1. Echocardiography machine settings for confirmatory echocardiograms

- Nyquist limits for colour Doppler should be set at 50 – 70cm/s to avoid overestimation of jet length.1
- Images for the assessment of valvular and chordal thickness should be acquired with harmonics turned off and probes with variable frequency set on ≥2 MHz. Low-frequency settings and harmonics exaggerate valve and chordal thickness.
- The room should be as dark as possible for echocardiography, because it impacts on gain settings. Gain settings should be adjusted to achieve optimal resolution. Images acquired with an over-gained setting will not be suitable for objective valve thickness measurements.
- All other settings (including depth, sector size and focus) should be optimised to achieve maximal frame rate and resolution.

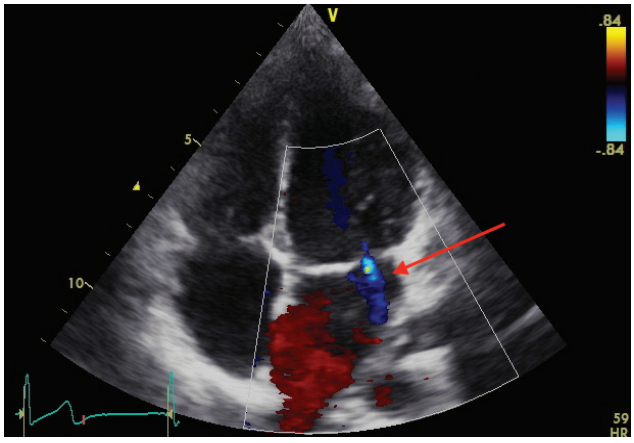


Figure 8.2. Rheumatic mitral valve; mitral regurgitant jet needs to measure at least 2 cm on colour Doppler (if patient weighs over 30 kg) to meet RHD diagnostic criteria for pathological regurgitation (red arrow). (See Table 8.6)

Table 8.7. Staging of RHD as detected by echocardiography based on WHF 2023 guidelines†‡

Minimum echocardiographic criteria for RHD (previously termed Borderline RHD) • This only applies to individuals ≤20 years old. • Echocardiographic features: The presence of mild§ mitral OR aortic regurgitation WITHOUT morphological features. • Clinical risk: for valvular heart disease progression.
Mild RHD (previously termed definite RHD) • Can apply to any age. • Echocardiographic features: Evidence of mild§ valvular regurgitation AND at least 1 morphological category in ≤20 years old and at least 2 morphological categories in >20 years old¶; OR mild regurgitation of BOTH mitral and aortic valves • Clinical risk: of progression and at risk of developing symptoms of valvular heart disease.
Advanced RHD at risk of clinical complications • Can apply to any age. • Echocardiographic features: Moderate/severe MR, moderate/severe AR, any MS or AS†† +/– associated pulmonary hypertension, dilated cardiac chambers, decreased LV systolic function. • Clinical risk: of developing clinical complications needing medical and/or surgical intervention.
Advanced RHD with clinical complications • Can apply to any age. • Echocardiographic features: Moderate/severe MR, moderate/severe AR, any MS or AS††+/– associated pulmonary hypertension, dilated cardiac chambers, decreased LV systolic function. • Clinical risk: including cardiac surgery, heart failure, arrhythmia, stroke, and infective endocarditis.

† To be applied in high-risk settings (RHD all-age prevalence is >2/1000 RHD or ARF incidence >30/100,000 per year in 5–14-year-olds) and requires other causes of valvular heart disease to have been excluded.

‡ Following the application of the confirmatory echocardiographic criteria, diagnostic categories may include a) 'Normal'; and b) 'Other' – congenital heart disease, cardiomyopathies, pericardial effusion, etc.

§ Fulfilling the confirmatory criteria for pathological regurgitation (Table 8.6) .

¶ This cut-off is based on expert consensus.

†† Aortic stenosis is defined as per international guidelines on valvular heart disease. A diagnosis of rheumatic aortic stenosis requires the exclusion of other causes including bicuspid aortic valve and degenerative calcific aortic stenosis.

MR: mitral regurgitation, AR: aortic regurgitation, MS: mitral stenosis, AS: aortic stenosis, LV: left ventricle

First Nations women may be identified in an advanced stage of disease during pregnancy. Given the importance and impact of personal and cultural supports at the time of diagnosis, there is a critical need for culturally appropriate knowledge transfer and communication. Potential language barriers should be addressed through the use of interpreters.

Table 8.8 Screening criteria using had carried ultrasound for the echocardiographic detection of RHD in individuals ≤20 years old (based on the 2023 WHF guidelines)

SCREENING CRITERIA
Mitral regurgitation (requires ALL of the following): 1. In <30 kg: MR jet length ≥1.5 cm. In ≥30 kg: MR jet length ≥2 cm†. 2. MR jet is seen in at least one view. 3. MR jet seen in ≥2 consecutive frames.
Aortic regurgitation (requires ALL of the following): 1. Any aortic regurgitation . 2. Seen in at least one view. 3. Seen in ≥2 consecutive frames.
Mitral stenosis: 1. Restricted leaflet motion with reduced valve opening.

† Cut-off is based on expert consensus. If weight is not available, then an age cut-off of <10 years or ≥10 years can be applied.

Note: Positive screen includes presence of ANY of the defined MR, AR, or MS; Negative screen is the absence of ANY of the defined MR, AR, or MS.

Screening for rheumatic heart disease

KEY INFORMATION

- Echocardiographic screening for RHD has been widely used in Australia and internationally, in research settings to estimate burden of disease, and as part of community-led initiatives aimed at early detection and treatment of RHD.
- Screening procedures have evolved over time, using different technologies and using operators with varying levels of expertise.
- Population-based screening using auscultation is not accurate for detecting undiagnosed RHD so is not recommended.
- Screening using echocardiography can accurately detect previously undiagnosed RHD.
- Echocardiographic screening for RHD meets public health criteria for community screening for disease (Table 9.2).
- The role for echocardiographic screening at community level and in pregnancy is increasingly favoured as technology evolves.
- Communities with high rates of ARF and RHD should be prioritised but are often the ones where resources and access to health care are most limited. Sustained national and regional funding and coordination are required to support echocardiographic screening and follow-up care for those at highest risk.
- Targeted screening of particular highest-risk groups (including 5 to 20 year olds in high risk settings) may be appropriate under certain circumstances; factors to take into consideration are presented in this chapter.
- Screening activities may be effectively used to estimate disease prevalence, and have the potential to improve community awareness, foster local champions and galvanise support for better RHD control.

Table 9.1. Summary of recommendations with GRADE Level of Evidence

RECOMMENDATION	GRADE
Population-based screening using auscultation is not accurate for detecting undiagnosed RHD so is not recommended.	1A
Echocardiographic active case finding can accurately detect previously undiagnosed RHD.	1A
The 2023 WHF guidelines should be applied when diagnosing RHD by echocardiography.	1A
Populations most likely to benefit from echocardiography active case finding are young people <20 years of age and pregnant women.	1C
Screening programs should prioritise communities with a high prevalence of RHD.	1B
Echocardiography detected cases of early RHD benefit from secondary prophylaxis	1A
Echocardiography active case finding for RHD should only be performed in settings with access to appropriate follow-up and management.	1B
Task-sharing echocardiography active case finding by non-expert operators is supported in a two-stage protocol under the supervision of experts.	1B
Highly abbreviated echocardiography screening protocols should only be used in settings where confirmatory echocardiography is available to diagnose RHD.	1B
Individuals with echocardiography screening detected RHD benefit from secondary prophylaxis to prevent disease progression.	1B
RHD screening programs should consider the benefits and risk to individuals, community and health services prior to implementation.	1B

Table 9.3. Considerations for screening

ETHICAL CONSIDERATIONS	
Availability of treatment	• Is there a reliable supply of BPG and a means of administering it for potentially many years? • Is immediate cardiac medical and/or surgical treatment available for severe RHD or congenital heart disease detected by screening?
Community leadership	• Has the community requested screening? • Are principles of self-determination met? • Are First Nations peoples involved in leadership of the program? • Do First Nations peoples have sovereignty of their data?
Culturally appropriate, informed consent	• Are educational resources available in local language? • Should interpreters be used and are they available? • Who is the most appropriate person to provide consent? • How is consent obtained, and what is the age of consent? • Have First Nations Health Workers and Health Practitioners been engaged in the process? • Are the potential impacts of a positive screening test able to be conveyed with the chosen consenting procedure?
WORKFORCE CONSIDERATIONS	
Resources available to conduct screening	• Who will perform the screening? • Who will perform the usual duties of that person(s) if they are assigned to screening activities? • Is additional training required? • Who will provide that training? • Will it be sustainable? • Who will maintain quality standards? • Is there support for community-based workers?
Resources available to confirm diagnosis	• Is there access to rapid cardiology review of abnormal screens? • Is there capacity within local cardiology services to review individuals with abnormal screens if needed? • How will the result and recommendations be transmitted to the patient and local health service?
Resources available to provide education	• Do local health facilities have the capacity to deliver education to people diagnosed with RHD and their families? • Is there capacity to support First Nations health staff to provide education and support? • Are specific resources available to engage adolescents?
Resources available to treat confirmed cases	• Do local health facilities have capacity to provide ongoing secondary prophylaxis? • Is there capacity within the local primary healthcare and cardiology services to provide clinical follow-up? • Is there an RHD register/control program to monitor follow-up?
ECONOMIC CONSIDERATIONS	
Additional resources will be required	• Cost of resources required will depend on screening model used • Equipment and consumables. • Staff, including training. • Travel. • Estimated number of peopel to be screened.

Cost effectiveness is affected by many variables	• Health economic analyses suggest that echocardiographic screening for RHD is cost-effective in high-risk populations in Australia.
	• Cost effectiveness will increase as the number of new cases detected per population screened increases, i.e.
	  • in high-prevalence populations (high pre-test probability).
	  • in settings with poor disease surveillance (resource-poor settings).
	  • if large cohorts can be screened at one time (e.g. large target population, high screening attendance).
	• Factors that make screening less cost effective include:
	  • High number of screens requiring cardiology review (poor specificity of the screening test/model).
	  • High travel costs associated with remoteness.†

† Combining screening activities in rural and remote areas with specialist cardiology visits may reduce costs and result in timely diagnosis and treatment planning.

Table 9.4. Models of echocardiographic screening

	(Cardiologist, Physician)	(Cardiac sonographer)		(Trained local health worker)	
		Direct support	Indirect support	Direct support	Indirect support
Screening personnel	Cardiologist	Cardiac sonographer	Cardiac sonographer	Briefly trained healthcare worker	Briefly trained healthcare worker
Diagnostic confirmation	Cardiologist	On-site cardiologist	Off-site cardiologist	On-site cardiologist	Off-site cardiologist
Availability of staff	✠	✠✠	✠✠✠	✠✠	✠✠✠
Echocardiographic equipment	Portable	Portable	Portable	Handheld	Handheld
Screening protocol	Abbreviated Full screen Confirmatory	Abbreviated Full screen Confirmatory	Abbreviated Full screen Confirmatory	Abbreviated	Abbreviated
Sensitivity of RHD detection	✠✠✠	✠✠✠	✠✠✠	✠✠	✠
Specificity of RHD detection	✠✠✠	✠✠✠	✠✠✠	✠✠	✠
Detection of congenital heart defects	✠✠✠	✠✠✠	✠✠✠	✠	–
Time to confirm diagnosis	Immediate	Immediate	Delayed	Immediate	Delayed

† Operators with limited training using handheld devices have lower sensitivity and specificity than qualified technicians using portable machines; a greater proportion will require a subsequent definitive scan by an expert, increasing the final cost.

✠: poor, ✠✠: good, ✠✠✠: excellent

Local healthcare workers have established relationships within the community and a connection with language and culture. Use of trained healthcare workers recognises these qualities and builds community capacity.

Involvement of local consumer/youth representatives helps to facilitate better access for local young people.

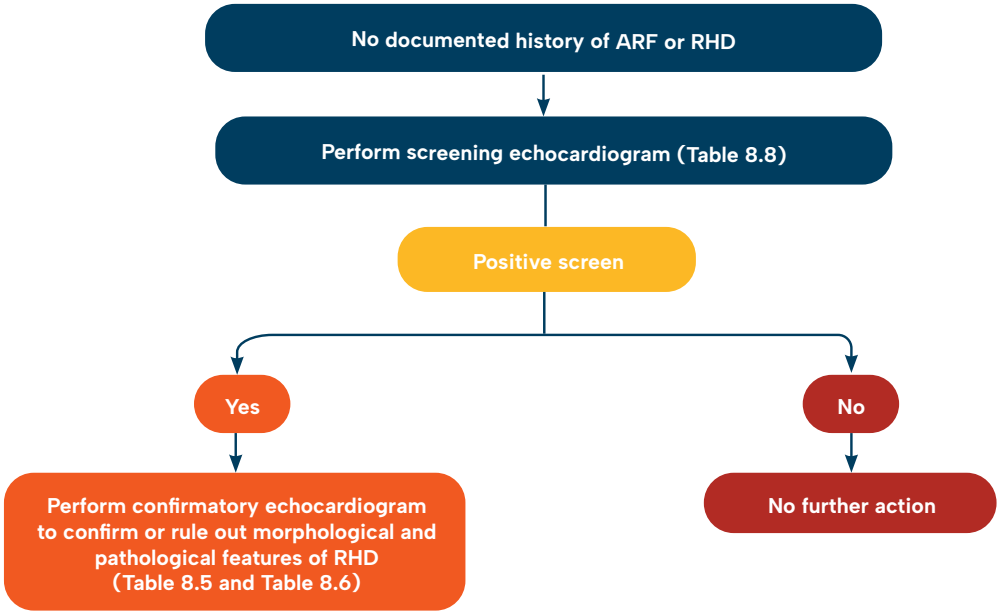


Figure 9.1. Screening pathway

Secondary prophylaxis

KEY INFORMATION

- Secondary prevention of ARF and RHD comprises secondary prophylaxis with an antibiotic (discussed here), plus holistic measures including enhance primordial and primary preventive activities targeted for individuals and households affected by ARF and/or RHD (discussed in Chapters 2, 4 and 5).
- Secondary prophylaxis of ARF is the consistent and regular administration of antibiotics to people who have had ARF or rheumatic heart disease (RHD), to prevent future group A beta-haemolytic streptococcus (Strep A) infections and recurrent ARF.
- Long-acting intramuscular benzathine benzylpenicillin G (BPG) used for ARF prophylaxis should not be confused with short-acting intravenous benzylpenicillin.
- Strep A is fully sensitive to penicillin. Failure of penicillin prophylaxis (i.e. breakthrough ARF recurrence despite receiving all prophylaxis with no days at risk) is not thought to be attributable to organism resistance but rather, to low serum or tissue concentrations of penicillin due to individual host differences in pharmacokinetic-pharmacodynamic properties.
- BPG injections should be delivered no later than 28 days after the last injection (or no later than 21 days for those prescribed a 21-day regimen) (Table 10.2).
- Starting from the first dose, administration of BPG should be patient centred, with appropriate technique and pain management to help reduce the risk of needle phobia developing, particularly in children.
- Single doses of BPG for the treatment of Strep A infection (i.e. primary prevention, Tables 5.3 and 5.4) differ slightly from regular doses of BPG for regular secondary prophylaxis of ARF. For example, small children under the age of five living in high-risk settings who frequently develop Strep A infections but rarely develop ARF, are recommended to receive weight-adjusted dosing to avoid excessively large BPG doses. A simpler strategy of two dose options with a single weight cut-off at 20 kg is used for secondary prophylaxis of ARF for pragmatic reasons (Table 10.2).

Table 10.1. Summary of recommendations with GRADE Level of Evidence

RECOMMENDATION	GRADE
Intramuscular penicillin is the most effective pharmacological strategy for ARF prevention	1B
Group A Streptococcal is fully sensitive to BPG	1A
BPG should be administered no later than 28 days after the last injection	1B
The internationally accepted standard dose of BPG for the secondary prevention of ARF in adults is 1,200,000 units	1B
Children weighing less than 20kg should receive 600,000 units of BPG to prevent recurrent ARF	1C
People who have ARF despite receiving all BPG injections on time, may be indicated for a 21-day regimen	2B
If a confirmed severe allergic reaction to penicillin is revealed, a non-beta lactam antimicrobial such as erythromycin should be used for secondary prophylaxis of ARF	1B
Caution is advised in the small subset of people who have severe pulmonary hypertension (mean pulmonary arterial pressure >50mmHg) and right ventricular failure and/or severe valve disease who are not eligible for valve replacement or repair. These patients may be prescribed oral penicillin following specialist review	2B
Penicillin and erythromycin are safe for mother and child during pregnancy and breastfeeding and should continue if indicated	1B
Oral penicillin prescribed for the same duration as BPG it is not as effective as BPG at preventing Strep A infections and subsequent recurrences of ARF	2B
BPG injections should be continued in patients receiving anticoagulation unless there is evidence of uncontrolled bleeding, or the international normalised ratio (INR) is greater than 4.5	1B
The duration of secondary prophylaxis following ARF is determined by the presence and/or severity of RHD	1A
Before ceasing secondary prophylaxis, it must be confirmed that there is no symptomatic deterioration, and that any existing valve lesions are stable	1A
Patients of all ages should have control over where and how they receive their injection	1B
Lidocaine (lignocaine) can reduce pain during injection and in the 24 hours after injection	1C
Withhold BPG pending specialist review in patients with severe pulmonary hypertension (mean pulmonary arterial pressure >50mmHg) and right ventricular failure and/or severe valve disease OR moderate RHD plus progressive symptoms suggesting progression or new complications since last echocardiogram	2C

Table 10.2. Recommended antibiotic regimens for secondary prophylaxis

ANTIBIOTIC	DOSE	ROUTE	FREQUENCY
First line			
Benzathine benzylpenicillin G (BPG)	1,200,000 units (≥20 kg) 600,000 units (<20 kg)†	Deep intramuscular injection	Every 28 days‡ Every 21 days for selected groups§
Second line (if intramuscular route is not possible or consistently declined)			
Phenoxymethylpenicillin (penicillin V)	250 mg	Oral	Twice a day
Following documented severe penicillin allergy			
Erythromycin	250 mg	Oral	Twice a day

† For children weighing less than 10 kg, a dose of 600,000 units is still generally recommended but seek paediatric advice for careful planning of the secondary prophylaxis regimen.

‡ People on 28-day regimens can be recalled from day 21 to help ensure that injections are given by day 28.

§ BPG given every 21 days may be considered for a) patients who have breakthrough ARF despite complete adherence to a 28-day regimen, or b) are at high risk of adverse consequences if ARF occurs (have severe RHD or a history of heart valve surgery).

NOTE: Amoxycillin 250mg twice daily for all ages is a pragmatic oral penicillin alternative that is preferred by some clinicians because of adequate absorption even when the stomach is not empty.

Table 10.3. Recommended duration of secondary prophylaxis, updated 2025

DIAGNOSIS	DEFINITION	DURATION OF PROPHYLAXIS	CONDITIONS FOR CEASING PROPHYLAXIS[†]	TIMING OF MEDICAL REVIEW AND ECHOCARDIOGRAPHY AFTER CESSATION[‡]
Possible ARF (no cardiac involvement)	Incomplete features of ARF with normal echocardiogram and normal ECG[§] throughout ARF episode	12 months (then reassess)	No signs and symptoms of ARF within the previous 12 months Normal echocardiogram	At 1 year
Probable ARF (no cardiac involvement)	Highly suspected ARF (with or without prolonged PR interval on ECG) with normal echocardiogram	Minimum of 5 years after most recent episode of probable ARF, or until age 21 years (whichever is longer), then reassess	No probable or definite ARF within the previous 5 years Normal echocardiogram	At 1, 3 and 5 years
Definite ARF (no cardiac involvement)	ARF with normal echocardiogram and normal ECG[§] throughout ARF episode (including with a background of Stage A)	Minimum of 5 years after most recent episode of ARF, or until age 21 years (whichever is longer), then reassess	No probable or definite ARF within the previous 5 years Normal echocardiogram	At 1, 3 and 5 years
Definite ARF (with cardiac involvement)	ARF with carditis or RHD on echocardiogram, or with atrioventricular conduction abnormality on ECG[§] during ARF episode (including with a background of Stage A)	According to relevant RHD Priority classification		
Applies only to people ≤20 years of age only (Previously borderline RHD)	**Stage A RHD** The presence of mild mitral OR aortic regurgitation WITHOUT morphological features of RHD on echocardiogram AND without a documented history of ARF	In a high-risk setting: Minimum of 2 years following diagnosis, then reassess *If Stage A RHD still present at 2 years continue for further 2 years and reassess. Consider specialist input*	No probable or definite ARF within the previous 10 years Normalisation of echocardiogram after a minimum of 2 years follow up	At 1–2 years
Priority 3 RHD[††]	Definite ARF with a prior diagnosis of Stage A RHD **Any Stage B RHD** by echocardiogram: Evidence of mild[‡] valvular regurgitation **AND** at least 1 morphological category in ≤ 20 years old and at least 2 morphological categories in > 20 years old[†] OR Mild regurgitation of BOTH mitral and aortic valves	If documented history of ARF: Minimum of 10 years after the most recent episode of ARF, or until age 21 years (whichever is longer), then reassess If NO documented history of ARF and aged <35 years:[‡‡] Minimum of 5 years following diagnosis of RHD or until age 21 years (whichever is longer), then reassess	No probable or definite ARF within the previous 10 years, no progression of RHD Stable echocardiographic features for 2 years	At 1, 3 and 5 years

Table 10.3. Recommended duration of secondary prophylaxis, updated 2025 (continued)

DIAGNOSIS	DEFINITION	DURATION OF PROPHYLAXIS	CONDITIONS FOR CEASING PROPHYLAXIS†	TIMING OF MEDICAL REVIEW AND ECHOCARDIOGRAPHY AFTER CESSATION‡
Priority 2 RHD†† §§	**Stage C RHD:** Moderate/severe MR, moderate/severe AR, any MS or AS+/– associated pulmonary hypertension, dilated cardiac chambers, decreased LV systolic function WITHOUT evidence of clinical complications including cardiac surgery, heart failure, arrhythmia, stroke, and infective endocarditis	If documented history of ARF: Minimum of 10 years after the most recent episode of ARF or until age 35 years (whichever is longer), then reassess noting that some individuals may require extended duration If no documented history of ARF and aged <35 years:‡‡ Minimum of 5 years following diagnosis of RHD or until age 35 years (whichever is longer), then reassess	No probable or definite ARF within the previous 10 years Stable echocardiographic features for 2 years	Initially every 12 months
Priority 1 RHD§§ ¶¶	**All Stage D RHD:** Moderate/severe MR, moderate/severe AR, any MS or AS+/– associated pulmonary hypertension, dilated cardiac chambers, decreased LV systolic function. **PLUS** Evidence of clinical complications including cardiac surgery,‡‡‡ heart failure, arrhythmia, stroke, and infective endocarditis	If documented history of ARF: Minimum of 10 years after the most recent episode of ARF or until age 40 years (whichever is longer), then reassess noting that some individuals may require extended duration If no documented history of ARF:††† Minimum of 5 years following diagnosis of RHD or until age 40 years (whichever is longer), then reassess	Stable valvular disease / cardiac function on serial echocardiogram for 3 years **OR** Patient or family preference to cease due to advancing age and/or end of life care	Initially every 6 months

† All people receiving secondary prophylaxis require a comprehensive clinical assessment and echocardiogram prior to cessation. Risk factors including future exposure to high Strep A burden environments need to be considered.

‡ Echocardiography may be more frequent based on clinical status and specialist review.

§ Normal ECG means no atrioventricular (AV) conduction abnormality during the ARF episode – including first-degree heart block, second degree heart block, third-degree (complete) heart block or accelerated junctional rhythm.

†† Prophylaxis may be considered for longer in women considering pregnancy who are at high risk of recurrent ARF (Table 6.2).

‡‡ If diagnosed with mild or moderate RHD aged ≥35 years (without a documented history of ARF), secondary prophylaxis is not recommended.

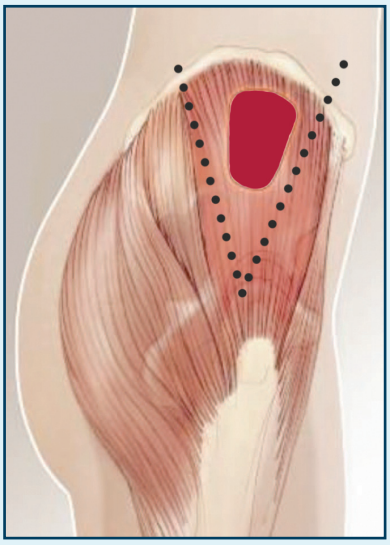
§§ Rarely, moderate or severe RHD may improve on echocardiogram without valve surgery. In these cases, the conditions for ceasing prophylaxis can change to follow the most relevant severity category. For instance, if moderate RHD improves to mild on echocardiogram, recommendations for mild RHD can then be instigated.

††† If diagnosed with severe RHD aged ≥40 years (without a documented history of ARF), specialist input is required to determine the need for secondary prophylaxis.

‡‡‡ There may be clinician discretion regarding duration of prophylaxis following surgery for people aged >40 years based on continued exposure to Strep A infections, ongoing risk of ARF recurrence, and type of valve surgery (repaired native valves are higher risk of damage; replaced mechanical valves are lower risk).

VENTROGLUTEAL SITE

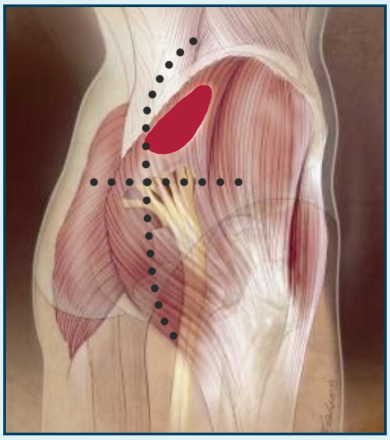
1. Place the patient in a side-lying position.
2. Using your right hand on the patient's left hip; or left hand on the patient's right hip:
    a. With the palm of your hand, locate the greater trochanter of the femur.
    b. Place your index finger towards the front or anterior superior iliac spine and fan the middle finger as far along the iliac crest as you can reach. (The thumb should always be pointed toward the front of the leg.)
3. The injection site is in the middle of the triangle between the middle and index fingers.
4. Remove your fingers prior to inserting the needle.



DORSOGLUTEAL SITE

CAUTION: Injections into the dorsogluteal muscle have been associated with sciatic nerve injury.

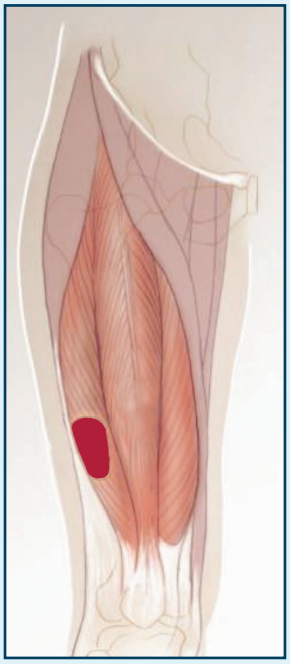
1. Place the patient in a prone (face down) position or lying on the side. Some patients may prefer standing up. Patients with valve disease at risk of cardiac decompensation must lie down (See Vasovagal syncope (fainting)).
2. The site for injection can be identified by either:
    a. dividing the buttock into four quadrants, selecting the upper outer quadrant;
    b. drawing an imaginary diagonal line from the posterior superior iliac spine to the greater trochanter. From the middle of the line move up and out.



VASTUS LATERALIS SITE

CAUTION: Some local protocols include volume restrictions for this site.

3. Place the patient in a supine (on back) or sitting position. Patients with valve disease at risk of cardiac decompensation must lie down (See Vasovagal syncope (fainting)).
4. Place one hand on patient's thigh against greater trochanter, the other hand against the lateral femoral condyle near the knee.
5. Visualise a rectangle between the hands across the thigh.
6. The correct injection site is the middle third of the anterolateral thigh.



Technique for administering BPG injections

1. Swab the skin with alcohol if it is visibly unclean.
2. Allow the skin to dry completely.
3. Apply firm pressure to the site with gloved thumb for at least 10 seconds immediately before injecting.
4. Insert using a 21 g needle into deep muscle, at an angle of 90 degrees to the skin.
5. Once inserted, apply gentle back pressure on the plunger to make sure that the needle tip is not in a blood vessel. (If blood returns into the syringe, withdraw, discard, and prepare a new syringe)
6. Inject slowly over 2–3 minutes.
7. Withdraw syringe and discard immediately into a sharps container.

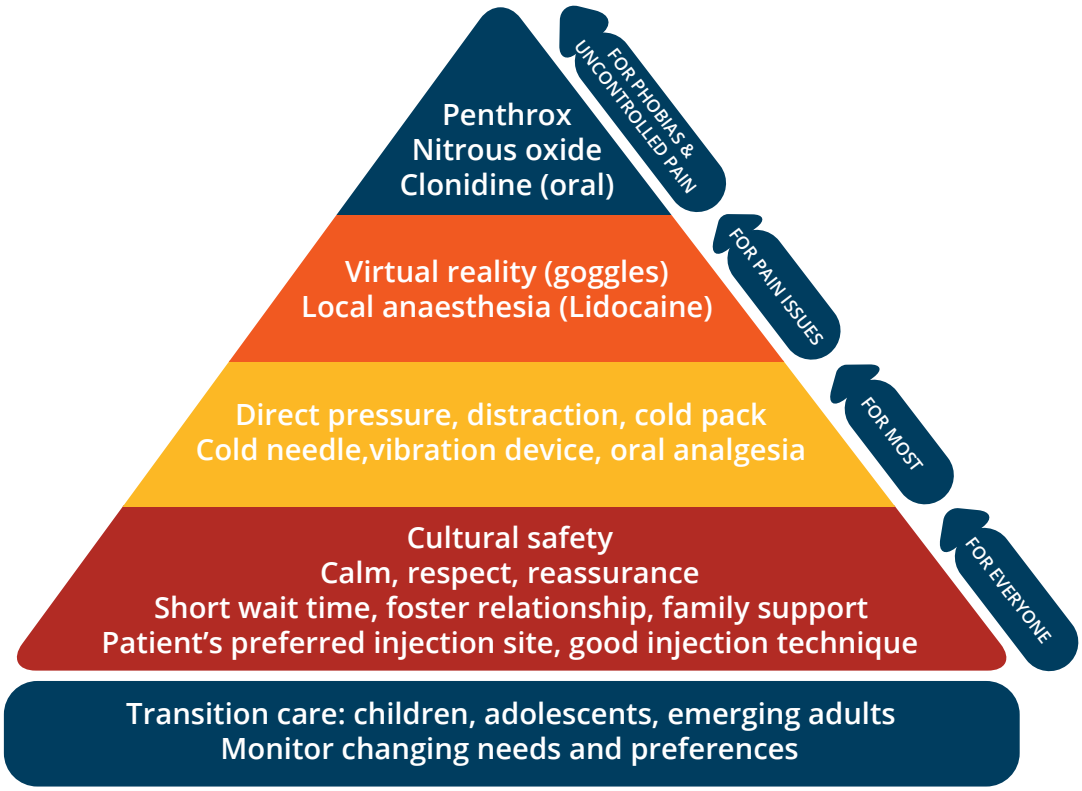


Figure 10.1. Strategies for injection managing pain, fear and distress

Box 10.2 Minimising the risk of fainting when BPG is administered

1. Prior to injection

- Check if there is any dizziness, chest pain or increasing shortness of breath. If yes:
  - Check manual pulse and blood pressure (if equipment available).
  - Withhold the BPG dose.
  - Follow usual clinical practice as per CARPA or other local guideline appropriate for the setting.
  - Arrange for transfer to the clinic if providing an outreach service.
  - Obtain medical (general practitioner) review, including phone consultation with a medical specialist where indicated.
  - If recommended by the doctor, substitute BPG with oral penicillin until the medical situation is stabilised and the doctor is confident in BPG safety (see Box 10.3).
- Provide a glass of water.
- Provide a snack if the patient is hungry.
- Provide reassurance and strategies to manage anxiety if there is anxiety about the injection.

2.During injection

- Recommend the patient lies down to receive the injection and stay resting for 5 minutes afterwards. If they don't want to lie down, respect patient autonomy but ensure they are making an informed decision. Explain that some people faint if they're standing up, but it's their choice how they get their injection.
- Use correct technique for injection delivery, and the hierarchy of strategies for managing injection pain, fear and distress (Figure 10.1).
- Use the 'Boss of my Body' checklistspecifying the patient's choice on who gives the injection, where they have it, pre-medication approaches, during needle methods, post-injection approaches.

Table 10.6. Strategies to improve the delivery of secondary prophylaxis

SUPPORT CATEGORY	EXAMPLES OF STRATEGIES
Health centre-related approaches	Health services imbed secondary prophylaxis strategies into existing programs including: • Prioritising secondary prophylaxis delivery • Ensuring that staff are skilled in injection delivery techniques • Providing care that is culturally safe • Using proactive, register-based recall systems to ensure patients are recalled for their next dose and given adequate notice • Having strategies in place to respond to overdue or missed injections • Providing options for outreach (home-based / school-based) injection delivery when feasible Liaising with other health services to help support continuation of care for patients who travel between regional areas.
Community-level approaches	Community awareness-raising events are supported Local peer support groups are established
Patient-level approaches	Self-management support with family and community engagement is provided ARF and RHD educational materials are suitable for the target audience e.g. in the patient's local language; provided in audio or video format BPG dose reminder systems are used, such as: • Smartphone application • Electronic or paper calendar Incentives are considered
Condition and therapy-specific approaches	Validated methods such as 'transition care' are used to support chronic care management from childhood through to adolescence and adulthood Non-pharmacological techniques are employed to improve the experience of BPG injection Pharmacological techniques are used when needed, and in addition to non-pharmacological approaches

Box 10.3: Checklist to review ongoing appropriateness of BPG in severe RHD

1. Can BPG be CEASED?

Ensure people who are no longer recommended for BPG have the opportunity to discuss discontinuation. Patients with Stage C or Stage D RHD may be able to cease secondary antibiotic prophylaxis at the age of 35 or 40 respectively, (Table 10.3) and according to the outcome of patient and clinician shared decision making discussions. All changes to treatment should be must be made by an RHD medical specialist.

2. Should the patient PERMANENTLY switch from injectable to oral penicillin?

Circumstances in which cardiovascular collapse may complicate a vasovagal episode include the following:

- RHD complicated by severe pulmonary hypertension (mean pulmonary arterial pressure >50mmHg) with or without right ventricular failure.
- RHD complicated by advanced (Class III, IV) and severe left ventricular systolic dysfunction function for which valvular intervention is not an option.

3. Should the patient TEMPORARILY switch from injectable to oral penicillin?

A temporary switch to oral penicillin may be needed in the days to weeks while a patient with increasing RHD severity is being stabilised (e.g. pending surgical and improved medical management). Additional circumstances in which there may be the very rare chance of cardiovascular compromise complicating a vasovagal episode include the following:

- Severe aortic insufficiency.
- Severe mitral stenosis.
- Severe aortic stenosis.
- Moderate or severe ventricular dysfunction.
- Significant or unstable symptoms (NYHA class III or IV).

4. What measures are being used to support oral penicillin prophylaxis?

People on oral penicillin may be taking other medications (e.g. anticoagulation, heart failure medications), so may be accustomed to taking medications every day. However, penicillin is taken twice per day and extra support may be needed (See Box 10.1).

Management of rheumatic heart disease

KEY INFORMATION

- Secondary prophylaxis is an integral aspect of the management of RHD.
- RHD is a notifiable disease in Western Australia, South Australia, Northern Territory, Queensland, New South Wales (for those aged <35 years) and Victoria.
- Anticoagulation in RHD:
    - Non-vitamin K antagonist oral anticoagulants (NOACs) are reasonable for patients with RHD and atrial fibrillation (AF) with an elevated CHA_2DS_2-VA score, except in those with moderate or greater mitral stenosis (Table 11.5).
    - For patients with moderate or greater mitral stenosis and atrial fibrillation, warfarin is currently the only indicated oral anticoagulant.
    - Patients with a mechanical valve prosthesis require anticoagulation with warfarin, low molecular weight heparin or unfractionated heparin.
- All patients with RHD should have access to specialist paediatric and adult cardiology services.
- Coordinated transition from paediatric to adult services is imperative for young patients with ARF and/or RHD.
- Shared decision making with patients and their families can be fostered through approaches that address power differentials between healthcare providers and patients, including culturally safe communication styles and respect for Indigenous knowledges.
- First Nations Health Workers and Health Practitioners and remote area nurses should be consulted prior to surgery when applicable to provide an understanding of the patient's personal, social, economic and cultural situation that will likely determine which surgical option is best suited to that individual.
- Early engagement of a multidisciplinary heart team is essential in determining the appropriate choice and timing of intervention for patients with RHD.
- The decision between balloon valvuloplasty, repair, bioprosthetic and/or mechanical valve replacement needs to take into consideration the age at first operation, risks of anticoagulation, adherence, future pregnancy, and durability of valve repair and prosthesis.
- Choice of valve replacement for RHD:
    - Mechanical valve: proven durability, requires lifelong anticoagulation.
    - Bioprosthetic valve: does not require lifelong anticoagulation, limited durability, may enable future valve-in-valve procedure.
- Complications of RHD include atrial fibrillation, heart failure, infective endocarditis, thromboembolic events, pulmonary hypertension, prosthetic valve thrombosis and death.
- Mixed and multi-valvular disease is common in RHD and requires more frequent surveillance and follow-up.
- Regular oral healthcare and education may reduce the long-term risk of infective endocarditis (IE) for patients with RHD.
- All people with RHD require IE antibiotic prophylaxis prior to high-risk procedures.

Of vital importance in Australia is the inclusion of First Nations Health Workers and Health Practitioners, nurses, Liaison Officers, and educators who are familiar with patients' social and cultural backgrounds. Early engagement of the heart team at dedicated centres of excellence is essential in determining the appropriate choice and timing of intervention for patients with RHD.

Table 11.1. Summary of recommendations with GRADE Level of Evidence

RECOMMENDATION	GRADE
MITRAL VALVE DISEASE	
ACE inhibitors and beta-blockers are recommended in adults with mitral regurgitation and left ventricular systolic impairment.	1A
For adults with mitral regurgitation and hypertension, anti-hypertensive agents should be used, including early use of ACE inhibitors.	1A
Patients with severe mitral regurgitation and preserved LV systolic function who are symptomatic should be automatically referred for surgical management.	1B
Patients with asymptomatic significant mitral regurgitation and any of the following; LVEF <60%, LVESD >40 mm, new onset AF, new PASP >50 mmHg or child with enlarged indexed heart size should be referred for consideration of valve intervention.	1B
Patients with significant MR, favourable anatomy, and good adherence with secondary prophylaxis, who do not meet the above criteria for intervention may be considered for early surgery in centres with low perioperative mortality and high rates of successful mitral valve repair.	2C
The operation of choice for dominant or pure rheumatic MR is mitral valve repair.	1B
A reduction in heart rate may reduce symptoms of MS, even in patients in sinus rhythm.	1C
Diuretics may be used for symptomatic relief in patients with pulmonary congestion or right heart dysfunction secondary to significant mitral stenosis.	1C
The indication for mitral stenosis intervention is progressive symptoms associated with documented evidence of severe MS (Adults: mitral orifice area ≤1.5 cm^2, trans-mitral pressure half-time ≥150 ms, mean trans-mitral gradient ≥10 mmHg. Children: mitral orifice area ≤1.5 cm^2, PASP ≥50 mmHg).	1A
Asymptomatic patients with mitral stenosis may be considered for intervention if is a history of thromboembolism, paroxysmal AF or significant pulmonary hypertension (PASP >50 mmHg).	2B
Exercise stress echocardiography can help evaluate asymptomatic mitral stenosis or discordant echocardiography parameters. Limited exercise tolerance for age as well as significant elevation in trans-mitral mean gradient (>15 mmHg) or pulmonary artery systolic pressure (>60 mmHg) may indicate the need for intervention.	2B
The treatment of choice for dominant or pure mitral stenosis is percutaneous balloon mitral valvuloplasty.	1A
In the relatively few patients who are not suitable for percutaneous balloon mitral valvuloplasty, every effort should be made to repair the mitral valve, rather than replace it, especially if patients are in sinus rhythm.	1C
Mitral valve replacement may be necessary in heavily calcified valves, especially with sub-valvular involvement, or in those with significant mixed mitral valve disease.	1B
AORTIC VALVE DISEASE	
Antihypertensive therapy is recommended in adult patients with significant aortic regurgitation and systemic hypertension.	1B
Vasodilator therapy may aid in symptoms in patients with significant symptomatic aortic regurgitation with or without impaired LV systolic function.	2B
Patients with symptomatic severe aortic regurgitation should be referred for surgery, regardless of left ventricular systolic function.	1B
Asymptomatic patients with severe aortic regurgitation and reduced systolic function (LVEF <50%) should be referred as soon as possible for valve surgery.	1B
Surgery should be considered in asymptomatic patients with severe aortic regurgitation and preserved left ventricular systolic function with severely dilated left ventricle (Adults: LVEDD >70 mm, LVESD >50 mm. Children: LVESD Z score >+4.0).	2B
Patients with asymptomatic aortic stenosis and hypertension should be treated as per standard guidelines, with frequent monitoring for side effects.	1B
Antihypertensives or diuretics should be used cautiously and with appropriate haemodynamic monitoring in patients with severe aortic stenosis who are normotensive, have clinical heart failure or have small LV cavity size.	1C

Table 11.1. Summary of recommendations with GRADE Level of Evidence (continued)

RECOMMENDATION	GRADE
Aortic valve replacement is recommended for severe symptomatic aortic stenosis (mean pressure gradient ≥40 mmHg, aortic valve area ≤1 cm^2, Vmax ≥4 m/sec).	1B
Patients with aortic stenosis with a moderate gradient with severely reduced aortic valve area should have further imaging including transoesophageal echocardiogram (TOE), CT or invasive haemodynamics to determine whether there is low flow, low gradient severe aortic stenosis.	2B
In patients with significantly impaired LV systolic function, dobutamine stress echo may help determine true severe aortic stenosis from pseudo-severe aortic stenosis.	2B
Patients with asymptomatic critical / very severe aortic stenosis, defined by a mean pressure gradient >50 mmHg or Vmax >5.5 m/sec, can be considered for surgery.	1C
TRICUSPID VALVE DISEASE	
Symptomatic relief of volume overload in severe tricuspid regurgitation is provided through use of diuretic therapy (frusemide, spironolactone).	1C
Severe symptomatic primary tricuspid regurgitation should be treated with surgical intervention.	1C
Severe tricuspid stenosis should be treated with surgery if symptomatic or in the setting of left-sided valve surgery.	1C
Intervention for minimally symptomatic severe primary or secondary tricuspid regurgitation may be warranted in the setting of progressive right ventricular dysfunction to prevent irreversible impairment.	2B
Functional progressive tricuspid regurgitation of moderate severity may be considered for surgery to coincide with a planned left-sided valve procedure.	2C
MIXED VALVE DISEASE	
In the setting of mixed or multi-valve disease, clinical symptoms and the nature of the predominant lesion should dictate the medical management and timing of cardiac intervention.	1C
Earlier surgery is preferred to avoid post-operative left ventricular dysfunction in patients with mixed valve disease.	2C
ATRIAL FIBRILLATION	
A CHA_2DS_2-VA score of ≥2 points is associated with significantly elevated risk of thromboembolic event that can be reduced with the use of therapeutic anticoagulation.	1A
Anticoagulation should be considered in individuals with a score of 1.	1B
Anticoagulation is not recommended in individuals with a score of 0.	2B
Patients with atrial fibrillation and moderate or severe mitral stenosis (regardless of CHA_2DS_2-VA score) or mechanical valve prostheses should be treated with warfarin.	1B
ANTICOAGULATION MANAGEMENT	
For patients with severe mitral stenosis in sinus rhythm and no history of atrial fibrillation, anticoagulation is indicated if there is a history of thromboembolic event or thrombus is visualised within the left atrium or left atrial appendage.	1B
Anticoagulation should be considered in those with severe mitral stenosis in sinus rhythm with significant spontaneous echo-contrast seen within the left atrium on echocardiography or significantly dilated left atrium.	2C
NOAC use in patients with significant mitral stenosis may be considered for specific cases where warfarin is contraindicated, and adherence has been demonstrated.	2D
Lifelong warfarin remains the only option for anticoagulation following implantation of a mechanical valve replacement.	1A
Patients receiving a bioprosthetic valve replacement may be treated with anticoagulation (warfarin) for the first one to three months post-surgery, as this has been demonstrated to be the highest risk period for thromboembolic events.	1C
For patients with thrombosis affecting a bioprosthetic valve, anticoagulation should be trialled before considering repeat surgery.	1C
Urgent surgery is recommended in critically ill patients with acute mechanical valve thrombosis.	1C

Table 11.1. Summary of recommendations with GRADE Level of Evidence (continued)

RECOMMENDATION	GRADE
For patients with acute mechanical valve thrombosis in locations where surgery is not immediately available or considered too high risk, thrombolysis should be considered.	2C
INFECTIVE ENDOCARDITIS PREVENTION	
Antibiotic prophylaxis is recommended for people with RHD undergoing specific dental, dermatological, musculoskeletal, respiratory, ENT, gastrointestinal and genitourinary procedures at high risk of bacteraemia that is associated with endocarditis.	1C
Amoxicillin is still appropriate for endocarditis prophylaxis in patients receiving long-term BPG injections for secondary prevention of ARF.	2D
All people with ARF and RHD need regular dental review to reduce the risk of infective endocarditis.	1C
Patients requiring cardiac intervention for RHD need a comprehensive dental consultation prior to surgery.	1C

Table 11.2. Priority classification and recommended follow-up (updated 2025)

DIAGNOSIS	RECOMMENDED FOLLOW -UP PLAN†
Priority 1	
All Stage D RHD‡	Specialist review: at least 6 monthly
High risk post-valve surgery patients§	Echocardiogram: at least 6 monthly
≥3 episodes of ARF within the last 5 years	Medical review: at least 6 monthly
Pregnant women with RHD (of any severity) may be considered Priority 1 for the duration of the pregnancy	Pregnant: see Figure 12.1 for care pathway
Children ≤5 years of age with ARF or RHD	Dental review: within 3 months of diagnosis, then 6 monthly
Priority 2	
Any Stage C RHD‡	Specialist review: 6 monthly – yearly
Moderate risk post-valve surgical patients§	Echocardiogram: 6 monthly – yearly
	Medical review: 6 monthly
	Dental review: within 3 months of diagnosis, then 6 monthly
Priority 3	
Any Stage A or B RHD‡	Specialist review: 1 – 3 yearly
ARF without carditis or RHD, currently prescribed secondary prophylaxis¶	Echocardiogram: children ≤ 21 years: 1–2 yearly, > 21 years: 2–3 yearly
Low risk post-valve surgical patients§	Medical review: yearly
	Dental review: yearly
Priority 4	
History of ARF (possible, probable or definite) and completed secondary prophylaxis	Specialist referral: 1 year, 3 years and 5 years post cessation of secondary prophylaxis
Resolved RHD and completed secondary prophylaxis††	Echocardiogram: 1 year, 3 years and 5 years post cessation of secondary prophylaxis
	Medical review: yearly until discharge from specialist care and then as required
	Dental review: yearly or as required

† Frequency should be tailored to the individual following specialist assessment. All patients should be given influenza vaccine annually and have completed pneumococcal vaccinations as per Australian Immunisation Handbook. Intervals for medical and specialist review and echocardiography are a guide and may vary for specific individuals. Medical and dental reviews may be combined with general health check-up. People with RHD require endocarditis prevention as indicated. (See Prevention of infective endocarditis).

‡ See Table 10.3 for definitions of RHD severity.

§ While post-surgical RHD is by definition severe RHD, post-surgical risk varies for individuals due to age, type of surgery, recurrence of ARF, adherence with secondary prophylaxis and other factors. Priority category for post-surgical RHD varies as listed in this Priority classification table and should be determined by specialist cardiologist/paediatrician/physician. (See Monitoring following valve surgery).

¶ See Table 7.1 regarding initial treatment of possible, probable and definite ARF with and without carditis. Table 10.3 provides guidance on longer term established RHD based on Stage of disease once the acute illness has resolved.

†† A proportion of early RHD changes can resolve with no residual valve dysfunction. These cases are referred to as 'resolved RHD' and as such may not need the longer-term follow-up required by Stage B/C/D disease.

NOTE: For Staging of RHD see Table 8.7. Staging of RHD as detected by echocardiography based on WHF 2023 guidelines.

Table 11.3. Summary of medical and surgical management options for specific advanced valve disease

VALVE DISEASE	MEDICAL THERAPY	INDICATIONS FOR CONSIDERATION OF INTERVENTION & REFERRAL TO HEART TEAM	VALVE INTERVENTION
Mitral Regurgitation (MR)	ACE inhibitor, beta-blocker and diuretic therapy in setting of heart failure. Antihypertensive medication in setting of hypertension.	Symptomatic severe MR Asymptomatic severe MR and: • LVEF ≤60% or • LVESD ≥40 mm or • New-onset AF or • New PASP ≥50 mmHg or • Child with enlarged indexed heart size	Valve repair (preferred intervention). If unable to be repaired, surgical valve replacement: • Bioprosthetic valve or • Mechanical valve
Mitral Stenosis (MS)	Beta-blockers (AF or sinus rhythm) or ivabradine (sinus rhythm) for symptom relief. Diuretics if evidence of pulmonary oedema/ congestion. Anticoagulation with warfarin if AF or high-risk features for thromboembolism present (See Monitoring anticoagulation).	Symptomatic severe MS Asymptomatic severe MS and: • Significantly elevated trans-mitral gradient or elevated PASP on EST or • New PASP ≥50 mmHg or • New-onset AF or • Cardio-embolic stroke	Percutaneous balloon mitral valvuloplasty (PBMV) if anatomically suitable. Closed or open surgical mitral valvotomy. Surgical valve replacement if not suitable for PBMV: • Bioprosthetic valve or • Mechanical valve
Aortic Regurgitation (AR)	Vasodilator therapy with ACE inhibitor, angiotensin receptor blocker or dihydropyridine calcium channel antagonist for symptom relief. Antihypertensive medication in setting of hypertension.	Symptomatic severe AR Asymptomatic severe AR and: • LVEF ≤50% or • LVESD >50 mm or • LVEDD >70 mm or • Child with enlarged indexed heart size	Aortic valve repair, if technically feasible. Surgical valve replacement: • Mechanical valve or • Bioprosthetic valve or • Homograft valve or • Ross procedure
Aortic Stenosis (AS)	Antihypertensive medication in setting of hypertension Cautious use of diuretic and afterload reduction in those with heart failure	Symptomatic severe AS Asymptomatic severe AS and: • LVEF <50% or • Abnormal EST or • Mean PG ≥60 mmHg or • Vmax ≥5 m/s or • PASP ≥60 mmHg	Surgical valve replacement or transcatheter valve replacement Decision based on surgical risk, age, anatomical assessment and heart team opinion
Tricuspid Regurgitation (TR)	Diuretic therapy for symptom relief from right heart failure and congestion	Severe primary TR Symptomatic severe secondary TR and absence of severe RV or LV dysfunction or severe pulmonary hypertension Asymptomatic or mildly symptomatic severe secondary TR with evidence of progressive RV dilatation or dysfunction Secondary moderate TR with annular dilatation in patients presenting for left-sided valve procedure	Valve repair / annuloplasty (preferred intervention) Surgical valve replacement: • Bioprosthetic valve or • Mechanical valve
Tricuspid Stenosis (TS)	Diuretic therapy for symptom relief from right heart failure and congestion	Symptomatic severe TS	Surgical valve replacement: • Bioprosthetic valve or • Mechanical valve

PASP: Pulmonary artery systolic pressure, AF: Atrial fibrillation, EST: Exercise stress test, LVEF: Left ventricular ejection fraction, LVEDD: Left ventricular end-diastolic diameter, LVESD: Left ventricular end-systolic diameter, PG: Pressure gradient, RV: Right ventricle.

Table 11.4. Standards for quality healthcare for adolescents

Adolescents' health literacy	**Standard 1**. The health facility implements systems to ensure that adolescents are knowledgeable about their own health, and they know where and when to obtain health services. *Communication needs to be in the young person's first language.*
Community support	**Standard 2**. The health facility implements systems to ensure that parents, guardians and other community members and community organisations recognise the value of providing health services to adolescents and support provision and use of services by adolescents. *Some communities have strong and active youth programs.*
Appropriate package of services	**Standard 3.** The health facility provides a package of information, counselling, diagnostic, treatment and care services that fulfills the needs of all adolescents. Services are provided in the facility and through referral linkages and outreach. *Where possible, regular BPG injections should be provided through outreach with consideration of school, work, and family commitments.*
Providers' competencies	**Standard 4**. Healthcare providers demonstrate the cultural and technical competence required to provide effective health services to adolescents. Both healthcare providers and support staff respect, protect and fulfill adolescents' rights to information, privacy, confidentiality, non-discrimination, non-judgmental attitude and respect.
Facility characteristics	**Standard 5**. The health facility has convenient operating hours, a welcoming, safe, friendly and clean environment, and maintains privacy and confidentiality. It has the equipment, medicines, supplies and technology needed to ensure effective service provision to adolescents.
Equity and non-discrimination	**Standard 6.** The health facility provides quality services to all adolescents irrespective of their ability to pay, age, sex, marital status, education level, ethnic origin, sexual orientation or other characteristics. *There needs to be reference to men's and women's business and services that reflect this.*
Data and quality improvement	**Standard 7.** The health facility collects, analyses and uses data on service utilisation and quality of care, disaggregated by age and sex, to support quality improvement. Health facility staff are supported to participate in continuous quality improvement.
Adolescents' participation	**Standard 8.** Adolescents are involved in the planning, monitoring and evaluation of health services and in decisions regarding their own care, as well as in certain appropriate aspects of service provision.

This table has been adapted to include First Nations cultural considerations.

Box 11.1. Factors to consider in selecting the nature and timing of valve interventions

Age at first operation, continued growth in children.

For women – future pregnancy and associated risk (Table 12.1).

Patient preference.

Adherence and adherence to regular secondary prophylaxis.

Access to anticoagulation monitoring and medications.

Adherence with medical therapy and anticoagulation.

Access to specialist follow-up – especially if previous valve repair.

Presence and severity of mixed and multi-valve disease.

Acceptability of redo surgery.

Comorbidities that would preclude patients from redo surgery.

Appropriateness for redo surgery using percutaneous valve-in-valve options.

Secondary indication for anticoagulation (e.g. atrial fibrillation).

Contraindications to anticoagulation (e.g. prior significant bleeding complications or bleeding conditions).

Access to primary healthcare services.

Shared decision making with patients and their families can be fostered through approaches that address power differentials between healthcare providers and patients, including culturally safe communication styles and respect for Indigenous knowledge.

If a valve is not able to be repaired, a decision between bioprosthetic and mechanical valve prosthesis needs to be made before the patient undergoes surgery, with consideration of adherence, geography, access to specialist follow-up, and cultural factors.

When discussing valve choice with First Nations peoples, the following factors remain important:

- Family support.
- Patient preference and lifestyle.
- Culturally appropriate communication.
- Interpreters being used where English is not the preferred language.
- Involvement of the patient's local health care providers with knowledge of the patient and available health care services.

Table 11.5. The CHA_2DS_2VA score is used to determine thromboembolic risk and guide use of anticoagulation in patients with non-valvular atrial fibrillation

CRITERIA	POINTS†
Age	65–74yrs = 1, ≥75yrs = 2
Congestive heart failure	1
Hypertension	1
Stroke/ transient ischaemic attack/ thromboembolic event	2
Vascular disease	1
Diabetes mellitus	1

† A score of ≥2 in the setting of non-valvular atrial fibrillation is an indication for anticoagulation. Anticoagulation should be considered in individuals with a score of 1. Anticoagulation is not recommended in individuals with a score of 0–1.

Table 11.6. Cardiac conditions and procedures for which infective endocarditis prophylaxis is recommended

Endocarditis prophylaxis is recommended ONLY for patients with the following cardiac conditions who are undergoing a procedure listed below.† ‡

CARDIAC CONDITIONS	PROCEDURES¶
Prosthetic cardiac valve, including transcatheter-implanted prosthesis or homograph Prosthetic material used for cardiac valve repair, such as annuloplasty rings and chords Previous infective endocarditis Congenital heart disease but only if it involves: • unrepaired cyanotic defects, including palliative shunts and conduits • repaired defects with residual defects at or adjacent to the site of a prosthetic patch or device (which inhibit endothelialisation) Rheumatic heart disease in all populations	**Dental procedures.** Only those involving manipulation of the gingival or periapical tissue or perforation of the oral mucosa (e.g. extraction, implant placement, biopsy, remove of soft tissue or bone, subgingival scaling and root planing, replanting avulsed teeth). **Dermatological and musculoskeletal procedures.** Only those involving infected skin, skin structures or musculoskeletal tissues. **Respiratory tract or ear, nose and throat procedures.** Only for tonsillectomy or adenoidectomy, or invasive respiratory tract or ear, nose and throat procedures to treat an established infection (e.g. drainage of abscess). **Genitourinary and gastrointestinal tract procedures.** Only if surgical antibiotic prophylaxis is required or for patients with an established infection.

Adapted from Australian Therapeutic Guidelines: Prevention of Infective Endocarditis, 2019.

† Endocarditis prophylaxis is not recommended for patients with forms of valvular or structural heart disease not listed in this table, including patients with mitral valve prolapse, septal defects or cardiac implantable electronic devices.

‡ Patients with a heart transplant who have developed cardiac valvulopathy may also be at high risk of adverse outcomes from endocarditis. Consult with patient's cardiologist for specific recommendations.

¶ Endocarditis prophylaxis is not recommended for procedures other than those listed above. However, surgical prophylaxis may be indicated if endocarditis prophylaxis is not.

Table 11.7. Antibiotics for infective endocarditis prophylaxis

DRUG	ROUTE	TIME BEFORE PROCEDURE
For endocarditis prophylaxis, use:		
Amoxicillin 2 g *(child: 50 mg/kg up to 2 g)*	oral	60 minutes before the procedure
If oral administration is not possible, use:		
Amoxicillin 2 g *(child: 50 mg/kg up to 2 g)*	intramuscular	30 minutes before the procedure, or
Amoxicillin 2 g *(child: 50 mg/kg up to 2 g)*	intravenous	within 60 minutes before the procedure, or
Ampicillin 2 g *(child: 50 mg/kg up to 2 g)*	intramuscular	30 minutes before the procedure, or
Ampicillin 2 g *(child: 50 mg/kg up to 2 g)*	intravenous	within 60 minutes before the procedure
For patients with delayed non-severe hypersensitivity to penicillins, cefalexin can be used in most cases,† use:		
Cefalexin 2 g *(child: 50 mg/kg up to 2 g)*	oral	60 minutes before the procedure
If oral administration is not possible, use:		
Cefazolin 2 g *(child: 30 mg/kg up to 2 g)*	intramuscular	30 minutes before the procedure, or
Cefazolin 2 g *(child: 30 mg/kg up to 2 g)*	intravenous	within 60 minutes before the procedure
For patients with immediate (severe or non-severe) or delayed severe hypersensitivity to penicillins,† use:		
Clindamycin‡ 600 mg *(child: 20 mg/kg up to 600 mg)*	oral	60–120 minutes before the procedure
If oral administration is not possible, use:		
Clindamycin‡ 600 mg *(child: 20 mg/kg up to 600 mg)*	intravenous	within 120 minutes before the procedure

† See Therapeutic Guidelines: Antimicrobial hypersensitivity / Management of patients reporting hypersensitivity to penicillins.

‡ There is some evidence that moxifloxacin may be used as an alternative to clindamycin for patients with immediate (severe) or non-severe or delayed hypersensitivity to penicillins, but this has not been validated.

Women and girls with rheumatic heart disease

KEY INFORMATION

- Effective multidisciplinary, community-centred care that is age-appropriate, encompasses reproductive health as well as cardiac and other health care and continues through the lifespan is imperative.
- Many women with RHD can safely conceive and have children. Women with mild RHD who are well-managed during their pregnancy may be able to birth on Country.
- Pre-conception diagnosis of RHD allows optimisation of management including surgical management, before pregnancy.
- There is a low threshold for performing echocardiography as part of antenatal care for any woman at high risk of RHD.
- Recommended contraceptives are long-acting reversible contraceptives (intra-uterine contraceptive device or etonogestrel implant such as Implanon). Oestrogen-containing contraceptives are associated with elevated risk of thrombosis and should be avoided.
- Women with RHD contemplating pregnancy or who are pregnant require coordinated health care. Aim to avoid multiple appointments incurring high travel costs and requiring time away from children and from community.
- Anticoagulation is needed for all women and girls with mechanical prosthetic valves to prevent valve thrombosis, stroke and other thromboembolic disease and may be needed for atrial fibrillation depending on thromboembolic risk assessment. All anticoagulants pose risks in pregnancy. Risks to the mother include both antepartum and post-partum haemorrhage. Risks to the fetus include teratogenicity and stillbirth (warfarin). Similarly, prosthetic valve thrombosis carries a high risk of mortality. An approach to balancing risks and benefits is provided in these guidelines.
- When low molecular weight heparin is used in pregnancy to replace warfarin, monitoring of anti-Xa levels and appropriate dose adjustment is essential.
- Women with valve lesions posing problems in pregnancy (moderate or greater mitral stenosis, severe mitral or aortic regurgitation, severe aortic stenosis, pulmonary hypertension or heart failure) are at high risk with elevated chance of cardiac events during pregnancy and adverse fetal outcomes. They require specialist care and close monitoring.
- A left ventricular ejection fraction of <30% or reduced systolic function with New York Heart Association (NYHA) class III/IV symptoms is associated with high risk of maternal morbidity or mortality, and pregnancy is strongly discouraged.
- A pregnant or post-partum woman at higher risk of or diagnosed with RHD who presents with breathlessness, orthopnoea, wheeze or worsening fatigue should be investigated with an echocardiogram as a matter of priority.
- Normal vaginal delivery is preferred. Epidural anaesthesia – after appropriately-timed short-term cessation of anticoagulation – may be indicated to reduce tachycardia and hypertension that can precipitate acute heart failure during delivery.

Table 12.1. Summary – Care pathways for women and girls with RHD

ASPECT	DETAILS	GRADE
TRANSITION TO ADULT CARDIAC CARE & PRECONCEPTION CARE		
Transition to adult cardiac care	Begins at adolescence. Include paediatric and adult cardiology teams, family planning, primary health services with the adolescent girl and her family.	1C 2B
Reproductive health & contraception	Refer to obstetrician/gynaecologist and/or family planning clinic (may be done through First Nations child and family programs) as relevant. Promote effective contraception to help plan safe timing of pregnancies. Avoid oestrogen-containing contraceptives.	1C 1B
Preconception care (PCC) & planning pregnancy	Full assessment and echocardiogram. Assess co-morbidities. Check vaccination status, rubella/varicella immunity and cervical screening. Review medications, especially warfarin or ACE inhibitors/angiotensin receptor blockers (ARBs). Consider a wallet card with RHD alert and key points related to care requirements and medications.	1C 1A 2C
Surgery & other interventions pre-pregnancy	Consider choices (prosthetic type/repair/PBMV) in context of future pregnancy and associated risk. Discussion with adolescent/woman, her family and appropriate primary health services together with specialist. Pre-pregnancy intervention recommended in patients with asymptomatic severe or symptomatic mitral stenosis (MS), symptomatic severe aortic stenosis (AS) or symptomatic severe valve disease.	1C 1B
DURING PREGNANCY		
Diagnosis of RHD in pregnancy	Attentive history-taking and careful cardiovascular examination. Low threshold for echocardiogram and cardiac referral in at-risk populations.	1C 1C
Integrated care	Includes cardiac (or obstetric physician), obstetric, anaesthetic, midwifery, primary health teams, First Nations health services, Māori, Pacific Islanders or refugee health workforce support (other disciplines/sectors as relevant) with women and family. Incorporate Birthing on Country models of care principles.	1C

Table 12.1. Summary – Care pathways for women and girls with RHD (continued)

ASPECT	DETAILS	GRADE
Cardiac risk assessment & general principles of care	Clinical risk assessment at booking and as required during pregnancy. Baseline echocardiography at booking and as required during pregnancy according to risk *(Figure 12.1)*. Anaesthetic assessment. Treatment in specialised centres by a multi-disciplinary pregnancy heart team for high-risk patients. Appropriate anticoagulation regimen where relevant. Interpreter services as required. Dental review. Assessment of social circumstances. Facilitate access to care depending on individual needs. Develop comprehensive birth plan as early as possible. Review/modify as needed. Discuss contraception: identify women who may desire tubal ligation at caesarean section or intrauterine device insertion at time of delivery.	1C 1C 1C 1C 2B 2B 2C 2C 1C 2C
Identify as high risk	Prior cardiac event (heart failure, transient ischaemic attack, stroke, arrhythmia) or events during pregnancy. Decreased left ventricular systolic function. Moderate or severe aortic and/or mitral stenosis. Pulmonary hypertension (PH). Mechanical valve prostheses or cardiac disorder requiring anticoagulation. Current heart failure or arrhythmia.	
RHD Register	Ensure the woman is on RHD Register in relevant jurisdictions. If not (or if not sure), contact RHD Register.	2B
Secondary prophylaxis	Determine ongoing need to be on secondary prophylaxis (usually 3–4 weekly benzathine benzylpenicillin G [BPG] injection) to prevent further rheumatic fever infection. If she is currently on regimen, check when next injection/oral antibiotic is due. Secondary prophylaxis is safe in pregnancy for mother and baby so should continue where prescribed.	1A
Mechanical heart valves & anticoagulation	Associated with high maternal and fetal risk. Discussion early in first trimester but ideally preconception to avoid warfarin in early pregnancy. Risk of warfarin embryopathy in first trimester. Risk of warfarin fetopathy in second and third trimesters. Highest risk of maternal thromboembolic complications with poor adherence to anticoagulation and/or monitoring, lack of appropriate multidisciplinary expertise especially when transitioning between different anticoagulant therapies.	1A 1C 1A 1A 1C

Table 12.1. Summary – Care pathways for women and girls with RHD (continued)

ASPECT	DETAILS	GRADE
Red flags	Symptoms and signs requiring urgent medical assessment: • new onset or progressive breathlessness or cough • need to sleep sitting up (orthopnoea) • significant reduction in exercise tolerance • syncope or presyncope (light headedness) • persistently fast heart rate (tachycardia) • wheeze and/or leg oedema	1C
LABOUR & BIRTH		
Labour & birth	Multi-disciplinary team approach (for First Nations women, include the First Nations Liaison Officer). Individualised birth plan taking account of cardiovascular and obstetric issues. Vaginal birth recommended unless obstetric and/or cardiovascular conditions preclude. Requirement for intrapartum intensive or invasive monitoring should be individualised depending on severity of underlying valvular disease. Follow anticoagulation protocol where relevant. Routine antibiotic prophylaxis for bacterial endocarditis not recommended and antibiotics should be given as per local obstetric indications. Aim for early epidural analgesia as tachycardia or hypertension may not be well tolerated because of maternal valvular disease. Oxytocin: administer slowly by infusion in third stage of labour. Avoid ergometrine in severe RHD, unless life-threatening bleeding.	2B 1C 2C 1A 1A 2C 2C 2C
POST-PARTUM & POST-DISCHARGE		
Post-partum	Consider need for diuretic therapy to assist with haemodynamic shifts post-partum. Follow anticoagulation protocol where relevant. Investigate post-partum/ post-discharge dyspnoea or new-onset cough promptly. Encourage breastfeeding and review safety of cardiac medications with lactation. Discuss family planning and contraception.	2C 1A 1A 1C 2C
Post-discharge	Follow-up cardiac review according to priority. Clinical communication follow-up with primary health services/GP/Aboriginal Medical Service and other relevant services. Maintain high degree of suspicion for presentation of dyspnoea.	1C 2C 1C
Information for health services & women	'Sharing a Heartbeat' Parts 1 & 2 short films and posters – for women and health services. eLearning modules (including Women and girls with RHD and RHD in Pregnancy).	

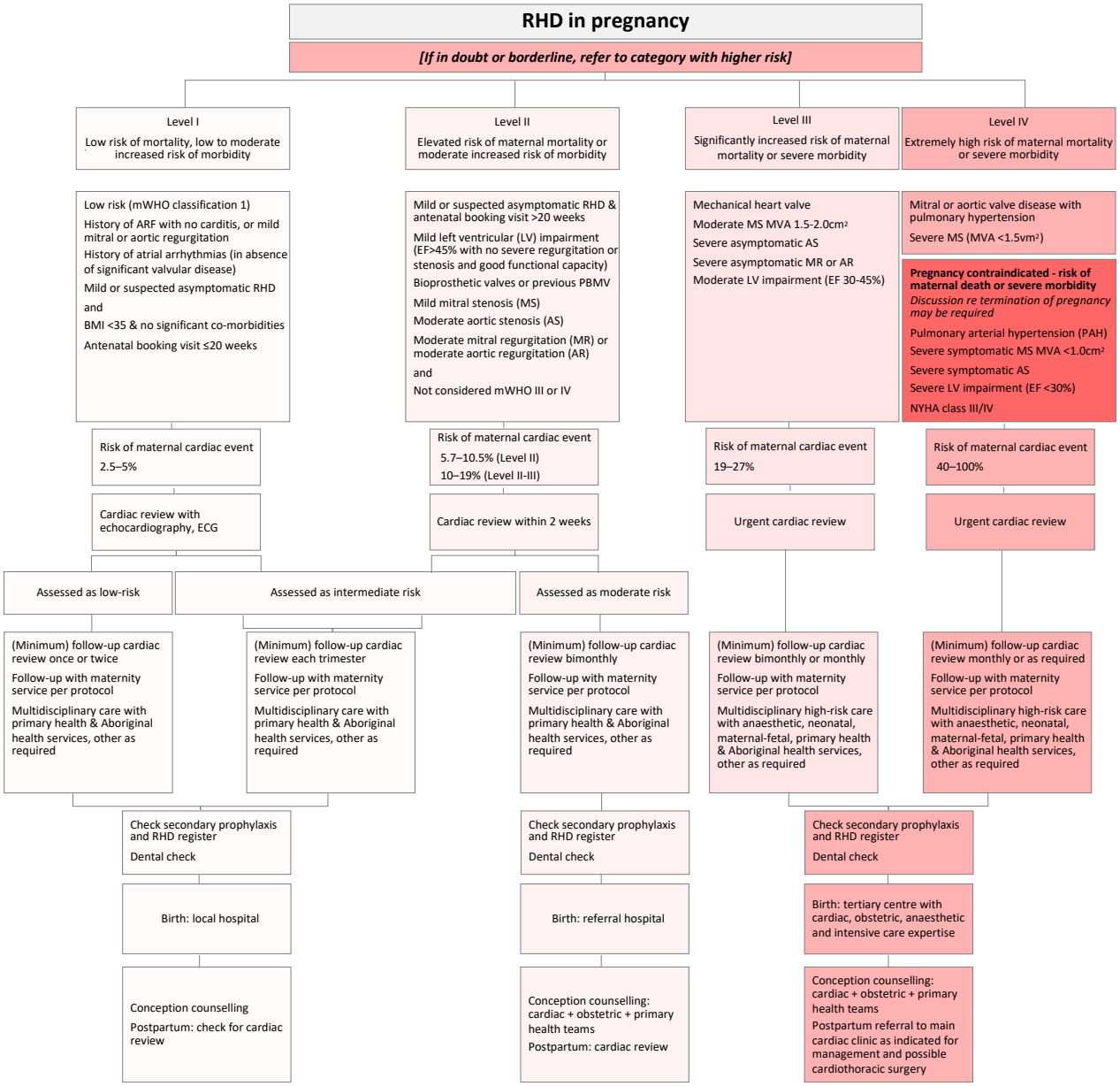


Figure 12.1. Care pathways and referral algorithm for pregnant women with RHD

Adapted with permission from Regitz-Zagrosek (2018), and Sliwa (2014).

NOTE: this and other guidelines addressing RHD during pregnancy are mostly based on case series and observational studies, often part of broader studies of all-valvular or all cardiovascular disease. Existing literature on preconception and reproductive health care is predominantly focused on congenital heart disease rather than RHD. Specific research to test the evidence is required to strengthen the rigour of recommendations, better understand the effects of pregnancy and choose the best individualised plan for ongoing care.

Abbreviations: Modified World Health Organization (mWHO), a system of classifying cardiac abnormalities in pregnancy; mitral regurgitation (MR); aortic regurgitation (AR); tricuspid regurgitation (TR); left ventricular (LV); pulmonary hypertension (PH); aortic stenosis (AS); mitral stenosis (MS); pulmonary arterial hypertension (PAH); mitral valve area (MVA); ejection fraction (EF).

Mild RHD: MVA >2 cm^2 AND EF=50-70% AND mitral or aortic or tricuspid regurgitation = none or mild AND no AS.

PAH: LV filling pressure <15 mmHg & pulmonary vascular resistance >3 Wood units

Significant co-morbidities include diabetes, BMI >35, chronic kidney disease, drug or alcohol dependency.

Risk of maternal cardiac event: according to modified World Health Organization classification of maternal cardiovascular risk adapted from Regitz-Zagrosek (2018).

Birthing on Country – why it matters for women with RHD

Birthing on Country addresses the integral connection between birthing, land (country) and place of belonging for First Nations women. The term is often not well understood. The Birthing on Country position statement describes it as '…a metaphor for the best start in life for First Nations babies and their families' which provides an appropriate transition to motherhood and parenting, and an integrated, holistic and culturally appropriate model of care for all.

Birthing on Country models can be described as maternity services that are designed, developed, delivered and evaluated with and for First Nations women that encompass the following:

- Community based and governed.
- Provide for inclusion of traditional practices.
- Involve connections with land and country.
- Incorporate a holistic definition of health.
- Value First Nations culture as well as other ways of knowing and learning; and
- Encompass risk assessment and service delivery and are culturally competent.

These principles underpin care pathways that promote optimal outcomes for women with RHD and their babies and should be incorporated into all aspects of care.

Birthing on Country models can be incorporated into any setting,[13] including urban as well as regional and remote Australia.

First Nations Mothers and Babies health services are Australian jurisdiction-based programs where midwives partner with First Nations Health Workers and Health Practitioners to provide culturally appropriate holistic support and care throughout pregnancy. They are known by various names across Australia: NSW Aboriginal Maternal Infant Health Services; SA Aboriginal Maternal and Infant Care; NT Top End Midwifery Group Practice; NT Central Australia Midwifery Group Practice; Qld Birthing in Our Country; WA Boodjari Yorgas Midwifery Group Practice.

Secondary prophylaxis (BPG injections, oral penicillin and erythromycin) is safe during pregnancy and breastfeeding and should continue if indicated. The importance of continuing secondary prophylaxis during a time of higher risk should be discussed with the woman and her family prior to a planned pregnancy, or as soon as possible during an unplanned pregnancy.

Rheumatic heart disease control programs

KEY INFORMATION

- Comprehensive RHD control programs which address the social and environmental determinants of health and the primary and secondary prevention of ARF, can be effective in reducing the burden of RHD.[1,2,3]
- RHD control programs:
    - Maintain register and recall systems to support secondary prophylaxis delivery and clinical management.
    - Support patient care by maintaining a skilled health workforce, promoting culturally appropriate care, supporting education and health promotion for patients and communities, and working with patients and primary healthcare staff to optimise delivery of secondary prophylaxis.
    - Promote primary prevention aimed at preventing initial episodes of ARF.
    - Provide jurisdiction-wide data for epidemiological reporting.

Table 13.1. Evolution of ARF and RHD notification and RHD program establishment in Australia

	NT	QLD	WA	SA	NSW	VIC	TAS, ACT
RHD Control Program established	1997†	2009	2009	2010	2015		
ARF/RHD Register established	1997	2006	2009	2012	2016		
Confirmed (definite) ARF notifiable	1996	1999	2007	2016	2015	2023	
Probable ARF notifiable	2019	2018	2015	2016	2015	2023	
Possible ARF notifiable		2018	2015	2016	2015	2023	
RHD Stage B, C and D notifiable	2019	2018	2015	2016	2015‡	2023	
RHD Stage A notifiable		2018	2015	2016		2023	

† The Top End Control Program was established in Darwin in 1997, and expanded in 2000 to include the whole NT.
‡ Notification of RHD only in persons aged less than 35 years.
§ Notification commenced from 31 July 2023.

Table 13.2. Processes for notification and inclusion on registers, April 2025

JURISDICTION	NOTIFICATION PROCESS	PATIENT CONSENT
NSW	Medical Practitioner or hospital CEO notifies the NSW Public Health Unit by telephone, or by completing and submitting a notification form.	Notification – consent not required. Register – informed, opt-in consent.
SA	Medical Practitioner notifies the SA Communicable Disease Control Branch by telephone or by completing and submitting a Notifiable Conditions reporting form within three days of suspecting or confirming a diagnosis (online form option available), AND Medical Practitioner notifies the SA RHD Register by telephone, or by completing and submitting a notification form.	Consent not required.†
QLD	Medical Practitioners, medical superintendents (or delegates) notify Queensland Health by completing and submitting an ARF notification form or RHD notification form which is recorded in the Notifiable Conditions System. Cases are referred to the Qld RHD Register.	Consent not required.‡
NT	Medical Practitioner notifies the relevant Public Health Unit at first suspicion, by contacting the NT RHD Control Program in Darwin or Alice Springs.	Consent not required for notification. (Consent requested for sharing information)
WA	Medical Practitioner or Nurse Practitioner notifies the WA RHD Register and Control Program by completing and submitting a notification form together with copies of diagnostic tests (including echocardiogram) and copies of each medical specialist's report (secure file transfer options available).¶	Consent not required.§
VIC	Medical Practitioner notifies the Victorian Department of Health by completing and submitting a notification via the Notification Portal. The notification is allocated to the relevant Public Health Unit for follow up.	Consent not required.

†Consent was required for all patients prior to 2019. From 2019 an opt-out option is available.

‡ Consent was required for patients with RHD registered prior to 2018

§ Notification required within 30 days of the medical specialist report.

¶ An individual can request in writing to the Chief Health Officer that there only be limited disclosure of identifying information on the register.

New technologies

KEY INFORMATION

- This chapter reviews research underway in Australasia which aims to discover better alternatives to benzathine benzylpenicillin G (BPG), develop a Strep A vaccine, and develop a diagnostic test for ARF.
- Promising data from Phase I trials has led to Phase II trials of delivery of high-dose subcutaneous infusions to make secondary prophylaxis hurt less and last longer.
- Development of a better BPG formulation is dependent on answering existing knowledge gaps relating to lowest effective dose of penicillin against Strep A and ideal route of delivery.
- Major national and international initiatives, including the Australian Strep A Vaccine Initiative (ASAVI), aim to fast-track development of a Strep A vaccine, with the goal to commence field trials including sites in Australia as soon as possible. Strep A vaccine challenges include the need to cover hundreds of different Strep A types and to avoid immune complications that could trigger ARF-like outcomes.
- Diagnostic tests for autoimmune disease usually rely on disease-specific antibodies and other immune markers such as complement levels, but no diagnostic test for ARF has yet been discovered. Research is underway to determine if biomarkers (molecules, genes, immune or other markers which can identify a disease process) measurable in blood may be discoverable which distinguish ARF from non-ARF presentations. If a distinguishing biomarker profile is discovered, then it may be possible to develop an ARF diagnostic test suitable for use in clinical diagnostic laboratories.

Community engagement is critically important to ensure that First Nations peoples are actively engaged in decisions about priorities and directions for research. Research into ARF and RHD must align with community needs, with consultation of community members about project design and implementation.

Australian guideline for the prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (Edition 3.3), 2025

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