KEY INFORMATION

2020 Australian guideline for the prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3.2 edition, March 2022)

Contents

Chapter 1. Introduction

Classifications and definitions

Acute rheumatic fever (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.

Rheumatic heart disease (RHD)

Borderline RHD: echocardiographic features in an individual aged ≤20 years which are abnormal but do not fulfil criteria for the diagnosis of RHD.

Definite RHD: echocardiographic features in an individual of any age which are abnormal and fulfil criteria for the diagnosis of RHD.

Classification of ARF / RHD severity

Priority 1 (P1): severe RHD.

Priority 2 (P2): moderate RHD.

Priority 3 (P3): borderline RHD, mild RHD.

Priority 4 (P4): History of ARF and/or RHD, secondary prophylaxis no longer required.

RHD in pregnancy

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.

Chapter 2. Culture & Workforce

There are many cultural and structural barriers for Aboriginal and Torres Strait Islander 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.

- Centrality of culture is the core component of clinical 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 Aboriginal and Torres Strait Islander peoples.
- An adequately trained and supported Aboriginal and Torres Strait Islander workforce is the key driver for successful health programs.
- The current health workforce will contribute to closing the evidence-practice gap.

Chapter 3. Burden of ARF & RHD

- Since the early 1990s, ARF has occurred almost exclusively in young Aboriginal and Torres Strait Islander peoples, particularly in the 5-14-year-old age group.
- During the same period, RHD has predominately affected young to middle-aged Aboriginal and Torres Strait Islander peoples as a consequence of the current era of endemic ARF among this population, and it has affected older non-Indigenous people due to a past era of endemic ARF.
- Females are more likely to be diagnosed with ARF than males.
- The number of Aboriginal and Torres Strait Islander peoples affected by ARF and RHD appears to be increasing.
- 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 with ARF are prone to a further episode, with one in five people having a recurrent episode of ARF within 10 years of their first.
- There is a high risk of valvular damage (RHD) 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.
- More than one in 10 people with RHD are affected by atrial fibrillation or heart failure attributable to RHD.
- Aboriginal and Torres Strait Islander peoples with RHD were more likely to die compared to non-Indigenous Australians with RHD; however, the death rates have decreased for both population groups over the past few decades.

Chapter 4. Primordial Prevention & Social Determinants of ARF

- The socioeconomic and political factors that influence people's lives can cause structural barriers and inequalities 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, ARF and RHD. Living in overcrowded conditions and having limited access to facilities to wash people, clothes and bedding increase the risk of Strep A infections, ARF and RHD.
- 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 in order to live healthy lives. 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.

Table 4.1. Healthy living practices and their association with Strep A infections.

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, skin damage and 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	Household 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. • 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.

Chapter 5. Primary Prevention (of ARF)

- 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 throat infections. 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.
- Not everyone who gets exposed to Strep A becomes symptomatic with a sore throat. Some people may become Strep A carriers.
- Strep A has been shown to be associated with up to 82% of impetigo episodes. Strep A impetigo is very common among Aboriginal and Torres Strait Islander children living in remote areas, 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.
- Individuals already receiving BPG secondary prophylaxis 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.

Table 5.1. Risk groups for primary prevention of ARF

At high risk	Living in an ARF-endemic setting† Aboriginal and/or Torres Strait Islander peoples living in rural or remote settings Aboriginal and/or Torres Strait Islander 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

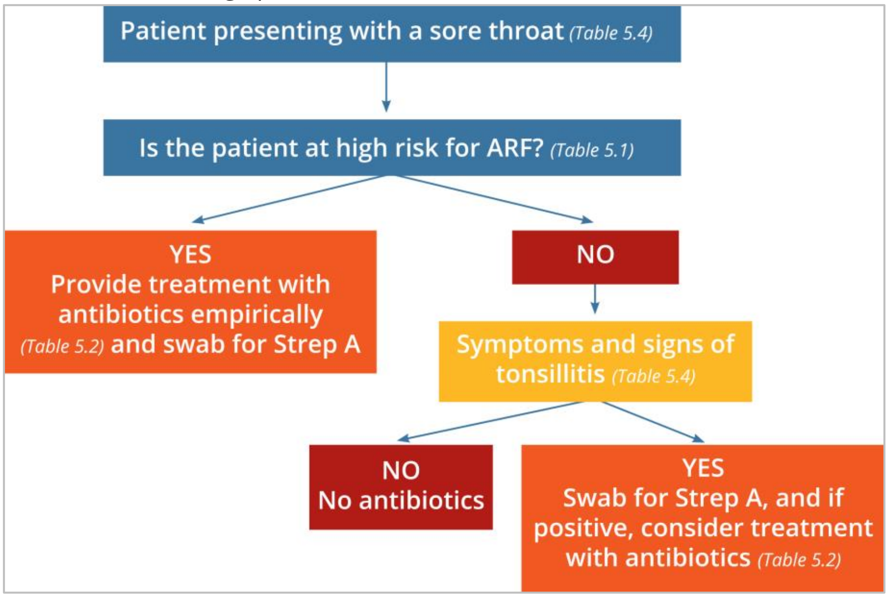


Figure 5.3. Assessment for sore throat

Table 5.4. Symptoms and signs of a sore throat / tonsillitis

Symptoms	Signs
Throat pain / sore throat	Fever (> 38°C)
Difficulty swallowing	Swollen, enlarged tonsils
Not eating as much	Erythematous tonsils with exudate
Not drinking as much	Enlarged, tender cervical lymph nodes
Croaky voice	Absence of cough
Feeling hot	

Symptoms derived from the Aboriginal Sore Throat Story (Sampson C et al, Australasian Society for Infectious Diseases Annual Scientific Meeting 2019 poster presentation). Signs derived from the Modified Centor Score.

Table 5.2. Recommended antibiotic treatment for Strep A sore throat / tonsillitis†

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

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

Table 5.3. Recommended antibiotic treatment for Strep A skin sores

DRUG	WEIGHT RANGE	DOSE			ROUTE	FREQU-ENCY
	All children with ≥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	Twice daily 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)	**Child:**				Deep IM injection	Once
	Weight			Dose in units (mL)		
	<10 kg			450,000 units (0.9 mL)		
	10 to <20 kg			600,000 units (1.2 mL)		
	≥20 kg			1,200,000 units (2.3 mL)		
	Adult:					
	≥20 kg			1,200,000 units (2.3 mL)		

† 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.

IM; intramuscular, BD; twice a day

Chapter 6. Diagnosis of acute rheumatic fever

- For each episode, 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; or
    - not ARF.
- Failure to diagnose ARF results in a missed opportunity for secondary prophylaxis with benzathine benzylpenicillin G (BPG), increasing the risk of recurrent ARF and cumulative heart valve damage.
- If clinicians are expecting to find a collection of ARF diagnostic criteria simultaneously in one individual, they are likely to be missing people with mild ARF, such as those who present with joint pain and fever only.
- 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.
- Anyone suspected to have ARF should be admitted to a hospital within 24-72 hours for echocardiography and specialist review.
- Echocardiogram is mandatory for all people with possible or confirmed 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 possible or 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.

- 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 6.1. Risk groups for ARF

At high risk	Living in an ARF-endemic setting†
	Aboriginal and/or Torres Strait Islander peoples living in rural or remote settings
	Aboriginal and/or Torres Strait Islander 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.

Table 6.2. 2020 Updated 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 on ECG†††	Fever ≥38.5°C Polyarthralgia or aseptic monoarthritis¶¶ ESR ≥60 mm/h or CRP ≥30 mg/L Prolonged P-R interval on ECG†††

† High-risk groups are those living in communities with high rates of ARF (incidence >30/100,000 per year in 5–14-year-olds) or RHD (all-age prevalence >2/1000). Aboriginal and Torres Strait Islander peoples living in rural or remote settings are known to be at high risk. Data are not available for other populations but Aboriginal and Torres Strait Islander 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 other streptococcal antibody, or a positive throat culture or rapid antigen or nucleic acid test for 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.
††† If carditis is present as a major manifestation, a prolonged P-R interval cannot be considered an additional minor manifestation.

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

Table 6.3. 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 6.4. 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.

Chapter 7. Management of ARF

- People suspected to have ARF should be referred as soon as possible for investigations (including echocardiography), treatment and education.
- Admission to a hospital with echocardiography services is strongly recommended in order to facilitate correct diagnosis. Echocardiographic findings inform the management plan including recommended duration of secondary prophylaxis.
- While the diagnosis is uncertain, giving salicylate and 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 based on the presence and severity of any accompanying RHD should also be provided, using the revised priority classification. The priority determines which care plan to use, including frequency of medical reviews and echocardiograms.
- People diagnosed with ARF should be
    - notified to the local Disease Control Unit or Public Health Unit in accordance with jurisdictional legislation.
    - registered with the jurisdictional RHD Control Program, with details of their secondary prophylaxis requirements.
- The pillars of management are eradication of the inciting infection using penicillin (or an alternative if allergic to penicillin) and 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
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*** 2. Phenoxymethylpenicillin 500 mg (child: 15 mg/kg up to 500 mg) orally 12-hourly for 10 days	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.
	3. *Penicillin hypersensitivity (non-severe)*: cefalexin 1 g (child: 25 mg/kg up to 1 g) orally, 12-hourly for 10 days 4. *Immediate penicillin hypersensitivity*: azithromycin 500 mg (child: 12 mg/kg up to 500 mg) orally daily for 5 days	Between 3 and 30% of Group A Streptococcus isolates internationally are resistant to macrolide antibiotics (e.g. azithromycin).
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.
Initial analgesia while awaiting diagnostic confirmation: severe pain	Tramadol immediate-release 50 to 100 mg (child 1 to 2 mg/kg) orally, 4-hourly up to a maximum of 400 mg/day	As above but for severe pain. Note safety warnings to avoid tramadol (or codeine) in children aged <12 years due to variable metabolism; therefore, use only when strong analgesia is essential and cautious monitoring is available.
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, up to a maximum of 1250 mg daily ***or*** 2. Ibuprofen 200-400 mg (child 5-10 mg/kg) orally three times daily, up to a maximum of 2400 mg 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.	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 is less in ARF than 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.
Symptomatic management of moderate to severe chorea *(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	Treatment of Sydenham chorea should be considered if movements interfere substantially with normal activities.

INDICATION	MEDICATION OPTIONS LISTED IN ORDER OF PREFERENCE	COMMENT
Symptomatic management of very severe chorea / *chorea paralytica* (Table 7.6)	In addition to an anticonvulsant agent, consider adding corticosteroid: Prednisone/prednisolone 1 to 2 mg/kg up to a maximum of 80 mg orally, once daily or in divided doses	
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.
	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.
	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.
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.

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

Chapter 8. Diagnosis of RHD

- In Australia, approximately 87% of RHD occurs in Aboriginal and Torres Strait Islander 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 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 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 World Heart Federation guidelines on echocardiographic diagnosis provide criteria to distinguish pathological RHD from physiological changes in children and adults.
- 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, heart failure or arrhythmia.

Table 8.1. Clinical features of common valve lesions

VALVE LESION	SYMPTOMS	SIGNS	COMPLICATIONS
Mitral regurgitation **(MR)**	Mild-moderate: Asymptomatic 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)**	Mild-moderate: Asymptomatic 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

Table 8.2. Echocardiographic features of RHD

CHARACTERISTIC FEATURES	MARKERS OF SEVERE DISEASE
Mitral valve features • Prolapse of anterior leaflet • Thickened leaflet tips • Restricted posterior leaflet • Chordal thickening • Leaflet calcification • Diastolic doming of anterior leaflet ("dog leg" or "hockey stick" appearance)	**Mitral regurgitation** • Central jet > 40% of LA • Holosystolic eccentric jet • Vena contracta ≥ 0.7 cm • Regurgitant volume ≥ 60 mL • Regurgitant fraction ≥ 50% • ERO ≥ 0.40 cm^2 **Mitral stenosis:** • Valve area ≤ 1.5cm^2 • Diastolic pressure half-time ≥ 150 ms • Mean pressure gradient ≥ 10 mmHg
Aortic valve features • Cusp prolapse • Cusp thickening • Rolled cusp edges • Cusp restriction • Cusp fibrosis, retraction, calcification • Dilated aortic root	**Aortic regurgitation** • Jet width ≥ 65% of LVOT • Vena contracta ≥ 0.6 cm • Holodiastolic flow reversal in the proximal abdominal aorta • Regurgitant volume ≥ 60 mL/beat • Regurgitant fraction ≥ 50%; • ERO ≥ 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** • Central jet area ≥ 10.0 cm^2 • Vena contracta width ≥ 0.7 cm • CW jet density and contour: dense, triangular with early peak • Systolic flow reversal in hepatic vein **Tricuspid stenosis** • Pressure half-time ≥ 190 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; ERO, Effective regurgitant orifice; LVOT, Left Ventricular Outflow Tract; LV, left ventricular; CW, Continuous wave;

Table 8.3. Role of cardiac investigations in the diagnosis of RHD

INVESTIGATION	ROLE
Transthoracic echocardiography† (TTE)	Baseline investigation 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 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 8.4. Morphological features of RHD

VALVE	MORPHOLOGICAL FEATURES†
Mitral valve	AMVL thickening ≥3 mm (age specific)‡ Chordal thickening Restricted leaflet motion§ Excessive leaflet tip motion during systole¶
Aortic valve	Irregular or focal thickening†† Coaptation defect Restricted leaflet motion Leaflet prolapse

†Minimal diagnostic criteria to differentiate normal from rheumatic

‡ Anterior mitral valve leaflet (AMVL) thickness should be measured during diastole at full excursion. Measurement should be taken at the thickest portion of the leaflet, including focal thickening, beading, and nodularity. Measurement should be performed on a frame with maximal separation of chordae from the leaflet tissue. Valve thickness can only be assessed if the images were acquired at optimal gain settings, without harmonics and with a frequency ≥2 MHz. Abnormal thickening of the AMVL is age specific and defined as follows: ≥3 mm for individuals ≤20 years of age; ≥4 mm for individuals 21–40 years of age; ≥5 mm for individuals >40 years of age.

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

¶ Excessive leaflet tip motion is the result of elongation of the primary chords, and is defined as displacement of an involved leaflet's tip or edge 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, as that refers to a different disease process. This feature applies to only those <35 years of age. In the presence of a flail mitral valve leaflet in young patients (<20 years of age), this single morphological feature is sufficient to meet the morphological criteria for RHD (i.e. where the criteria state 'at least two morphological features of RHD of the mitral valve', a flail leaflet in a person <20 years of age is sufficient).

†† In the parasternal short axis view, the right and non-coronary aortic cusp closure line often appears echogenic (thickened) in healthy individuals, and this phenotype should be considered as normal.

Echocardiography machine settings

1. Nyquist limits for colour Doppler should be set on maximum to avoid overestimation of jet length.
2. 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.
3. 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.
4. All other settings (including depth, sector size and focus) should be optimised to achieve maximal frame rate and resolution.

Table 8.5. 2012 World Heart Federation criteria for echocardiographic diagnosis of RHD

Echocardiographic criteria for individuals aged ≤20 years

Definite RHD (either A, B, C or D):

A) Pathological MR and at least two morphological features of RHD of the MV

B) MS mean gradient, ≥4 mmHg (note: congenital mitral valve anomalies must be excluded)

C) Pathological AR and at least two morphological features of RHD of the AV

D) Borderline disease of both the AV and MV†

Borderline RHD (either A, B, or C):

A) At least two morphological features of RHD of the MV without pathological MR or MS

B) Pathological MR

C) Pathological AR

Normal echocardiographic findings (all of A, B, C, and D):

A) MR that does not meet all four Doppler echocardiographic criteria (physiological MR)

B) AR that does not meet all four Doppler echocardiographic criteria (physiological AR)

C) An isolated morphological feature of RHD of the MV (for example, valvular thickening) without any associated pathological stenosis or regurgitation

D) Morphological feature of RHD of the AV (for example, valvular thickening) without any associated pathological stenosis or regurgitation

Echocardiographic criteria for individuals aged >20 years

Definite RHD (either A, B, C, or D):

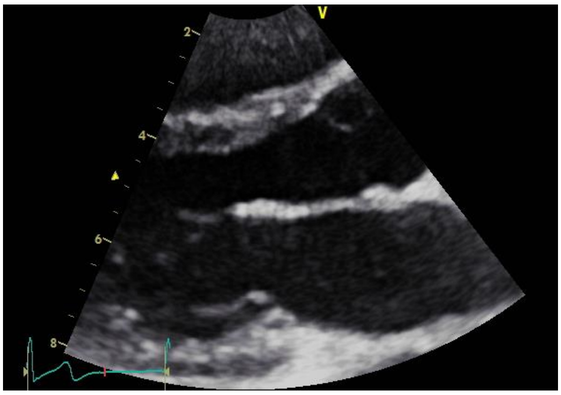
A) Pathological MR and at least two morphological features of RHD of the MV

B) MS mean gradient, ≥4 mmHg

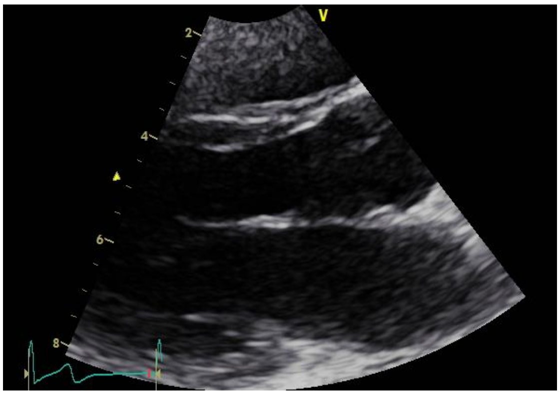
C) Pathological AR and at least two morphological features of RHD of the AV, only in individuals aged <35 years

D) Pathological AR and at least two morphological features of RHD of the MV

† Combined AR and MR in high-prevalence regions and in the absence of congenital heart disease is regarded as rheumatic.
AR, aortic regurgitation; MR, mitral regurgitation; MS, mitral stenosis.



Rheumatic mitral valve; appearance with



Rheumatic mitral valve; appearance with

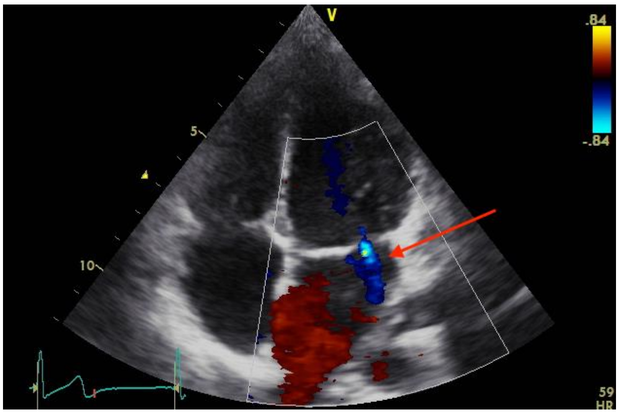
harmonics 'on', note anterior mitral valve thickness. Harmonics should be turned off.

harmonics 'off', note anterior mitral valve thickness.

Table 8.6. Criteria for pathological regurgitation

Pathological mitral regurgitation	Seen in two views In at least one view, jet length ≥ 2cm† Peak velocity ≥ 3 m/s Pan-systolic jet in at least one envelope
Pathological aortic regurgitation	Seen in two views In at least one view, jet length ≥ 1cm† Peak velocity ≥ 3 m/s Pan-diastolic jet in at least one envelope

† A regurgitant jet length should be measured from the vena contracta to the last pixel of regurgitant colour (blue or red) on unmagnified images.



Rheumatic mitral valve. Mitral regurgitant jet needs to measure at least 2 cm on colour doppler to meet RHD diagnostic criteria for pathological regurgitation (red arrow).

Chapter 9. Screening for Rheumatic Heart Disease

- Echocardiographic screening for rheumatic heart disease (RHD) has been widely used internationally, often in a research setting or to estimate burden of disease.
- Screening procedures have evolved over time, using different technologies and 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 some, but not all, of the standard public health criteria for community screening for disease. In particular, the impact of secondary prophylaxis on disease trajectory is not well defined.
- There remains insufficient evidence to support routine, population-level echocardiographic screening for RHD in Australia as a method of disease detection and control.
- Targeted screening of particular highest-risk groups 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.

Chapter 10. Secondary Prophylaxis

- Secondary prophylaxis of ARF is the consistent and regular administration of antibiotics to people who have had ARF or RHD, to prevent future group A beta-haemolytic streptococcus (Strep A) infections and the recurrence of ARF.
- Long-acting benzathine benzylpenicillin (BPG) used for ARF prophylaxis should not be confused with short-acting benzylpenicillin.
- Strep A is fully sensitive to penicillin. Failures of penicillin secondary prophylaxis (i.e. breakthrough ARF recurrence despite receiving all prophylaxis with no days at risk) are 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 21 days for those prescribed a 21-day regimen)
- Doses of BPG for the treatment of Strep A infection differ slightly from doses of BPG for regular secondary prophylaxis. 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.
- Heart valve surgery does not affect the risk of Strep A infections or recurrent ARF. Therefore, secondary prophylaxis must be continued following surgery

Table 10.1. 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 IM route is not possible or consistently declined)			
Phenoxymethylpenicillin (penicillin V)	250 mg	Oral	Twice a day
Following documented 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 regimen of secondary prophylaxis.

‡ People on 28-day regimens can be recalled from 21 days 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).

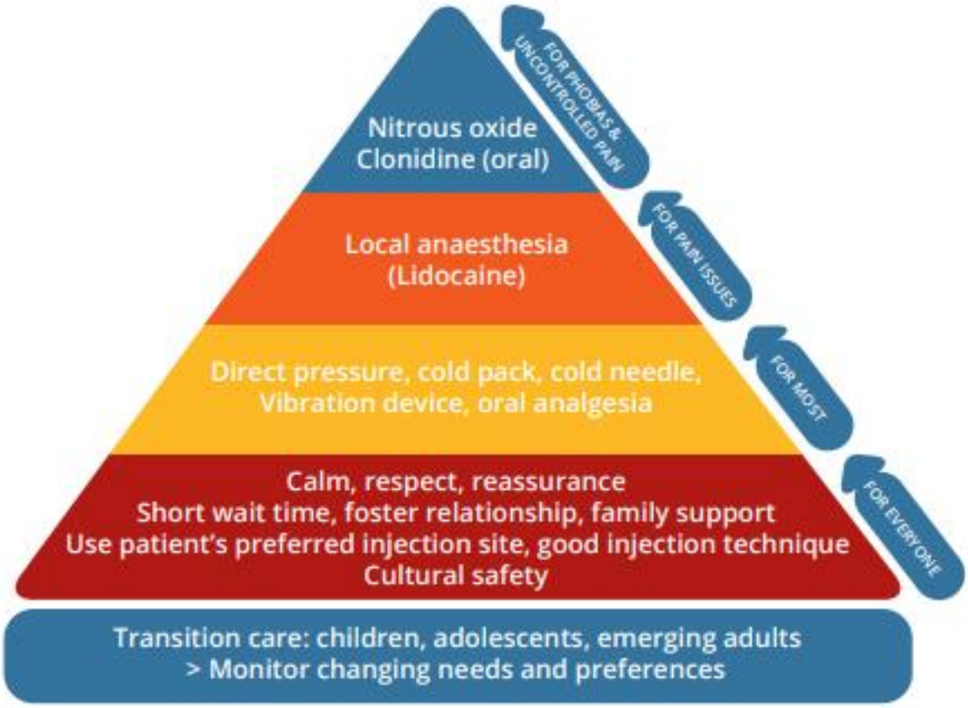


Figure 10.1 Strategies for managing injection pain, fear and distress.

Table 10.2. Recommended duration of secondary prophylaxis

DIAGNOSIS	DEFINITION	DURATION of PROPHYLAXIS	CONDITIONS for CEASING PROPHYLAXIS[†]	TIMING of ECHO-CARDIOGRAPHY 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	Highly suspected ARF with normal echocardiogram	Minimum of 5 years after most recent episode of probable ARF, or until age 21 years (whichever is longer)	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	Minimum of 5 years after most recent episode of ARF, or until age 21 years (whichever is longer)	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	According to relevant RHD severity		
Borderline RHD (this diagnosis applies to people ≤20 years of age only)	Borderline RHD on echocardiogram without a documented history of ARF	In a high-risk setting: Minimum of 2 years following diagnosis of borderline RHD *If borderline 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	Medical review and repeat echocardiogram at 1-2 years after diagnosis, and 1-2 years after stopping secondary prophylaxis
Mild RHD[¶]	Echocardiogram showing: Mild regurgitation or mild stenosis of a single valve OR Atrioventricular conduction abnormality on ECG[§] during ARF episode	If documented history of ARF: Minimum of 10 years after the most recent episode of ARF, or until age 21 years (whichever is longer) 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)	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

DIAGNOSIS	DEFINITION	DURATION of PROPHYLAXIS	CONDITIONS for CEASING PROPHYLAXIS[†]	TIMING of ECHO-CARDIOGRAPHY AFTER CESSATION[‡]
Moderate RHD[††§§]	Echocardiogram showing: Moderate regurgitation or moderate stenosis of a single valve OR Combined mild regurgitation and/or mild stenosis of one or more valves Examples: • Mild mitral regurgitation and mild mitral stenosis • Mild mitral regurgitation and mild aortic regurgitation	If documented history of ARF: Minimum of 10 years after the most recent episode of ARF or until age 35 years (whichever is longer) 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)	No probable or definite ARF within the previous 10 years Stable echocardiographic features for 2 years	Initially every 12 months
Severe RHD[§§¶¶]	Echocardiogram showing: Severe regurgitation or severe stenosis of any valve OR Combined moderate regurgitation and/or moderate stenosis of one or more valves Examples: • Moderate mitral regurgitation and moderate mitral stenosis • Moderate mitral stenosis and moderate aortic regurgitation OR Past or impending valve repair or prosthetic valve replacement[‡‡‡]	If documented history of ARF: Minimum of 10 years after the most recent episode of ARF or until age 40 years (whichever is longer) If no documented history of ARF:[†††] Minimum of 5 years following diagnosis of RHD or until age 40 years (whichever is longer)	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 and accelerated junctional rhythm.

†† Prophylaxis may be considered for longer in women considering pregnancy who live in high-risk circumstances for ARF.

‡‡ If diagnosed with mild or moderate RHD aged ≥35 years (without ARF), secondary prophylaxis is not required.

§§ 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.

¶¶ Risk of ARF recurrence is low in people aged ≥40 years, however, lifelong secondary prophylaxis is usually recommended for patients who have had, or are likely to need, heart valve surgery.

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

‡‡‡Priority classification is variable, see *Table 7.4* and *Table 11.2* for clarification

Chapter 11. Management of RHD

- Secondary prophylaxis is an integral aspect of the management of RHD.
- RHD is a notifiable disease in Western Australia, South Australia, Northern Territory and Queensland. RHD is notifiable for people aged less than 35 years in Queensland.
- 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 RHD.
- Aboriginal Health Workers, Aboriginal Health Practitioners and remote-area nurses should be consulted prior to surgery 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.
- 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.
- Aboriginal and Torres Strait Islander peoples with RHD should receive IE antibiotic prophylaxis for high-risk procedures.
- The decision between 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, thromboembolic events, pulmonary hypertension, prosthetic valve thrombosis and death.

Anticoagulation in RHD

- Non-vitamin K antagonist oral anticoagulants (NOACs) are appropriate for patients with RHD and atrial fibrillation with an elevated CHA2DS2-VA score, except in those with moderate or greater mitral stenosis
- 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, clexane or heparin.

Table 11.1 The CHA_2DS_2-VA 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 11.2. Priority classification and recommended follow-up

DIAGNOSIS	RECOMMENDED FOLLOW-UP PLAN†
Priority 1 Severe RHD‡ High risk post-valve surgical 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 Moderate RHD‡ Moderate risk post-valve surgical patients§	Specialist review: yearly Echocardiogram: yearly Medical review: 6 monthly Dental review: within 3 months of diagnosis, then 6 monthly
Priority 3 Mild RHD‡ ARF (probable or definite) without 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
Borderline RHD currently prescribed secondary prophylaxis	Medical review: 1-2 years after diagnosis, and 1-2 years after ceasing secondary prophylaxis Echocardiogram: 1-2 years after diagnosis, and 1-2 years after ceasing secondary prophylaxis
Priority 4 History of ARF (possible, probable or definite) and completed secondary prophylaxis Borderline RHD not on secondary prophylaxis Resolved RHD and completed secondary prophylaxis	Specialist referral and echocardiogram: 1 year, 3 years and 5 years post cessation of secondary prophylaxis *(or following diagnosis in the case of Borderline RHD not on 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 10.2* 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*).

Table 11.3. Summary of medical and surgical management options that may be considered 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*** • LVEDD >70 mm ***or*** • LVESD >50 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

VALVE DISEASE	MEDICAL THERAPY	INDICATIONS FOR CONSIDERATION OF INTERVENTION & REFERRAL TO HEART TEAM	VALVE INTERVENTION
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

(Grading of evidence-based recommendation available in relevant valve disease section)

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 11.5. 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 11.6. 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.

Chapter 12. Women & Girls with RHD

- Effective multidisciplinary, community-centred care is age-appropriate, encompasses reproductive health as well as cardiac and other health care and continues through the lifespan.
- Many women with RHD can safely conceive and have children. Women with mild RHD may be able to birth on Country.
- Pre-conception diagnosis of RHD allows optimisation of management including surgical management, before pregnancy.
- Recommended contraceptives are long-acting reversible contraceptives (intra-uterine contraceptive device or etonogestrel implant such as Nexplanon). 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 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). An approach to balancing risks and benefits is provided.
- 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 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.
- When low molecular weight heparin is used in pregnancy to replace warfarin, monitoring of anti-Xa levels and appropriate dose adjustment is essential.

- 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 with RHD

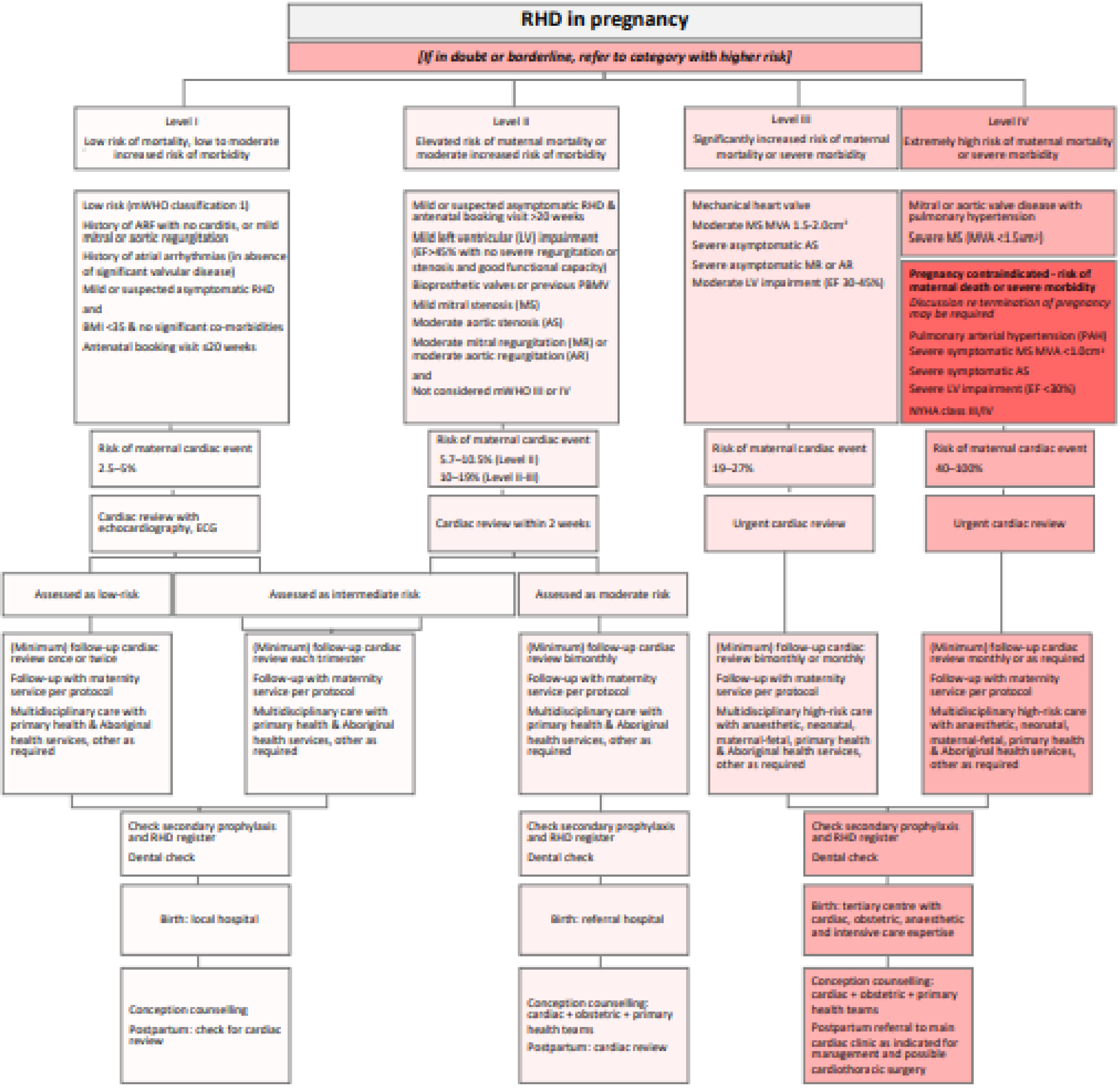
ASPECT	DETAILS	GRADE
TRANSITION TO ADULT CARDIAC CARE & PRECONCEPTION CARE		
Transition to adult cardiac care	Begins at adolescence. Include paediatric and adult cardiology teams, family planning, primary health services with the adolescent girl and her family.	1C 2B
Reproductive health & contraception	Refer to obstetrician/gynaecologist and/or family planning clinic (may be done through Aboriginal and Torres Strait Islander child and family programs) as relevant. Promote effective contraception for all girls and women, especially if pregnancy poses a risk. 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, Aboriginal health service, 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

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	Check if woman needs 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. Safe in pregnancy for mother and baby so should continue where prescribed.	1A
Mechanical heart valves & anticoagulation	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

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 Aboriginal and Torres Strait Islander women, include the Aboriginal 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 of 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 when 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	RHDAustralia Treatment Tracker reminder app. 'Sharing a Heartbeat' Parts 1 & 2 short films and posters – for women and health services. RHDAustralia 'e-learning modules modules (including RHD in Pregnancy).	

RHD Australia

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



Adapted with permission from Regitz-Zagrosek (2018), and Sliwa (2014)[1,2]

Abbreviations: Modified World Health Organization (mWHO); 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/aortic/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 and/or alcohol dependency.

Risk of maternal cardiac event: according to modified World Health Organization classification of maternal cardiovascular risk adapted from Regitz-Zagrosek (2018).[1]

Chapter 13. RHD Control Programs

An Aboriginal and Torres Strait Islander workforce should be embedded into the RHD control program, to provide guidance and support to program activities, and to help translate health promotion into culture and practice. Further consideration should be given to partnering with local Aboriginal and Torres Strait Islander health programs and organisations to facilitate disease control across social and cultural pathways.

- Comprehensive RHD control programs which span action on the social and environmental determinants of health, and primary and secondary prevention of ARF, can provide effective approaches to reducing the burden of RHD.
- A key aim of RHD control programs is to maintain a register and recall system for secondary prophylaxis and clinical management.
- RHD control programs:
    - 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	1997†	2009	2009	2010	2015	✘
ARF/RHD Register	1997	2006	2009	2012	2016	✘
Confirmed (definite) ARF notifiable	1996	1999	2007	2016	2015	✘
Probable ARF notifiable	2019	✘	2015	2016	2015	✘
Possible ARF notifiable	✘	✘	2015	2016	✘	✘
Confirmed RHD notifiable	2019	2018	2015	2016	2015‡	✘
Borderline RHD notifiable	✘	2018	2015	2016	✘	✘

† 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.

Table 13.2. Processes for notification and inclusion on registers, as at December 2019

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.[23]	Notification – consent not required. Register – informed, opt-in consent.
SA	1. Medical practitioner notifies the SA Communicable Disease Control Branch by telephone or by completing and submitting a notification form within three days of suspecting or confirming a diagnosis (online form option available). ***and*** 2. Medical practitioner notifies the SA RHD Register by telephone, or by completing and submitting a notification form.[24]	Consent not required.†
QLD	Medical practitioners, medical superintendents (or delegates) notify the QLD RHD Register and Control Program by completing and submitting an ARF notification form or RHD notification form.	Consent not required.‡
NT	Medical practitioner notifies the relevant Public Health Unit at first suspicion, by completing and submitting a notification form.	Consent not required.
WA	Medical 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.§

†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.

Chapter 14. New Technologies

- 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.
- Research is underway to determine whether a penicillin depot (implant) may be a future reality. Development is dependent on answering existing knowledge gaps relating to lowest effective dose of penicillin against Strep A and ideal route of delivery.
- Funding initiatives, including a 2019 Australian Government grant, aim to fast-track development of a Strep A vaccine, with the goal of being able to commence field trials to assess efficacy and safety within 5 years from 2019. Challenges with Strep A vaccine development 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 (measurable 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.