

# Rheumatic Heart Disease (RHD)

## Case Definitions

**Rheumatic Heart Disease (RHD):** damage to one or more heart valves that remains after an episode of Acute Rheumatic Fever (ARF) has resolved. Patients are classified according to their disease priority (see [classification, treatment and follow up](#)).

**Acute Rheumatic Fever (ARF):** illness caused by an immunological reaction following infection with Strep A. It can cause fever, arthritis, chorea (abnormal movements), and carditis (inflammation of the heart) (see [KAHPF Acute Rheumatic Fever protocol](#)).

**Strep A:** group A streptococcus bacteria that can infect skin or throat. Also called "Group A Strep" or "*Streptococcus pyogenes*".

**Benzathine Benzylpenicillin G (BPG):** long acting intramuscular (IM) form of penicillin. Sometimes called "LA Bicillin" or "LAB".

## Prevention

Early identification and treatment of skin and throat infections can prevent ARF/RHD. See [KAHPF "Sore Throat" and "Skin Infections" protocols](#).

Those already receiving BPG prophylaxis still need active treatment of sore throat or skin infection. If the most recent dose was  $\geq 7$  days ago then treat as per above guidelines.

## Case Detection & Diagnosis

**ARF:** everyone with confirmed or suspected ARF must have an echocardiogram to look for RHD (see [KAHPF Acute Rheumatic Fever protocol for frequency](#)).

**Murmur:** all Aboriginal people in the Kimberley with a murmur must be assessed for possible RHD.

- Adults: echocardiogram and refer to physician
- Children: refer to paediatrician for clinical assessment and possible referral to paediatric cardiology

**Cardiac failure:** consider RHD and echocardiogram in patients with unexplained symptoms of cardiac failure including dyspnoea, tachypnoea, oedema, paroxysmal nocturnal dyspnoea, or those with syncope or chest pain.

## Principles of Management

### Co-ordinated multidisciplinary team-based approach

- Refer for [Physician/Paediatrician or Cardiology](#) involvement following RHD diagnosis
- Refer to clinic / community chronic disease program champion where available
- Aboriginal Health Workers or Aboriginal Health Practitioners are a vital part of the team, providing support, education and assistance to both patients and other health professionals

### Secondary prophylaxis with BPG

- BPG secondary prophylaxis to prevent recurrences of ARF and progression to RHD (see [classification, treatment and follow](#)

[up](#) and [secondary prophylaxis](#)).

### Notification

- RHD is a notifiable disease. Ensure notification to the WA RHD Register (see [WA RHD Register and Control Program](#))

### Prevention of infective endocarditis

- Encourage good dental hygiene
- Regular dental review (as per [classification](#))
- Antibiotic prophylaxis when required (see [antibiotic prophylaxis for procedures](#))

### Monitor and manage for complications of RHD including:

- Atrial fibrillation (AF): consider rate control to prevent decompensation. Consider anticoagulation for those with an elevated CHA<sub>2</sub>DS<sub>2</sub>-VA score (see [RHD Australia 2020 guideline page 195](#)). Direct oral anticoagulants (DOACs) are appropriate, except in patients with moderate or greater mitral stenosis who require warfarin or enoxaparin
- Heart failure: see [KAHPF Heart Failure protocol](#)
- Pulmonary hypertension
- Endocarditis
- Thromboembolic events: those with atrial appendage thrombus without AF should be considered for anticoagulation
- Significant valvular disease: ensure all those with moderate to severe valve lesions have been seen by/referred to cardiology. Valve repair/replacement is typically required for high severity disease (see [RHD Australia 2020 guideline page 197 for indications](#))

### Anticoagulation for mechanical heart valves

Patients with mechanical valves usually require anticoagulation.

- In some circumstances, warfarin may still be continued in pregnancy provided dose  $<5$ mg, however seek specialist advice due to risk of embryopathy
- Target INR as per cardiothoracics/cardiology advice, but typically:
  - Mitral valve replacement (MVR) 2.5-3.5
  - Aortic valve replacement (AVR) 2-3

### Pregnant women

- Pregnancy causes a 40-50% increase in cardiac output and workload, which may exacerbate pre-existing RHD
- Recommend asking all pregnant women from high risk groups about cardiac symptoms, previous symptoms consistent with ARF, and performing cardiovascular examination. Have a low threshold for performing screening echocardiogram for RHD for these women
- All pregnant women with known RHD, previous ARF or a new murmur need an echocardiogram and early specialist involvement (see [women's health in RHD](#))
- All pregnant women with RHD can be considered priority 1 for the duration of their pregnancy

### Other

- Ensure pneumococcal / influenza vaccines up to date
- Consider [Environmental Health referral](#)

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## Therapeutic Protocols

### Classification, Treatment, Follow up and Duration of Prophylaxis in RHD

- Is dependent on the patient's disease priority
- Should be guided by a specialist. Ensure specialist plan is clearly documented in the medical record
- Ensure the [WA RHD Register and Control Program](#) is kept up to date with the latest patient treatment information

The following table is a guide only, and should never replace specialist advice. Individual circumstances may vary

CLASSIFICATION	TREATMENT & FOLLOW UP	DURATION OF PROPHYLAXIS
<b>Priority 1</b> <ul style="list-style-type: none"> <li>• Severe RHD of any valve</li> <li>• High risk post-valve surgical patients (risk level determined by specialist)</li> <li>• ≥3 episodes of ARF within the past 5 years</li> <li>• Pregnant women with RHD (of any severity) may be considered Priority 1 for the duration of their pregnancy</li> <li>• Children ≤ 5 years of age with ARF or RHD</li> </ul>	<ul style="list-style-type: none"> <li>• BPG every 21-28 days</li> <li>• Specialist review: at least 6 monthly</li> <li>• Echocardiogram: at least 6 monthly</li> <li>• Medical review: at least 6 monthly</li> <li>• Pregnant: see pregnancy section</li> <li>• Dental review: within 3 months of diagnosis, then 6 monthly</li> </ul>	<ul style="list-style-type: none"> <li>• If documented history of ARF: Minimum of 10 years after the most recent episode of ARF or until age 40 years (whichever is longer)</li> <li>• If no documented history of ARF: Minimum of 5 years following diagnosis of RHD or until age 40 years (whichever is longer)</li> </ul>
<b>Priority 2</b> <ul style="list-style-type: none"> <li>• Moderate RHD of any valve</li> <li>• Mild RHD involving both aortic and mitral valves</li> <li>• Moderate-risk post-valve surgery patients (risk level determined by specialist)</li> </ul>	<ul style="list-style-type: none"> <li>• BPG every 21-28 days</li> <li>• Specialist review: yearly</li> <li>• Echocardiogram: yearly</li> <li>• Medical review: 6 monthly</li> <li>• Dental review: within 3 months of diagnosis, then yearly</li> </ul>	<ul style="list-style-type: none"> <li>• If documented history of ARF: Minimum of 10 years after the most recent episode of ARF or until age 35 years (whichever is longer)</li> <li>• If no documented history of ARF and aged &lt;35 years: Minimum of 5 years following diagnosis of RHD or until age 35 years (whichever is longer)</li> <li>• If diagnosed with mild or moderate RHD aged ≥35 years (without ARF), secondary prophylaxis is not required.</li> </ul>
<b>Priority 3</b> <ul style="list-style-type: none"> <li>• Mild RHD involving only a single valve</li> <li>• ARF (probable or definite) currently prescribed secondary prophylaxis</li> <li>• Borderline RHD currently prescribed secondary prophylaxis</li> <li>• Low-risk post-valve surgery patients (risk level determined by specialist)</li> </ul>	<ul style="list-style-type: none"> <li>• BPG every 21-28 days</li> <li>• Specialist review: 1 – 3 yearly</li> <li>• Echocardiogram: children ≤21 years: 1-2 yearly, &gt;21 years: 2-3 yearly</li> <li>• Medical review: yearly</li> <li>• Dental review: yearly</li> </ul>	<ul style="list-style-type: none"> <li>• If documented history of ARF: Minimum of 10 years after the most recent episode of ARF, or until age 21 years (whichever is longer)</li> <li>• If NO documented history of ARF and aged &lt;35 years: Minimum of 5 years following diagnosis of RHD or until age 21 years (whichever is longer)</li> <li>• If diagnosed with mild or moderate RHD aged ≥35 years (without ARF), secondary prophylaxis is not required</li> </ul>
<b>Priority 4</b> <ul style="list-style-type: none"> <li>• History of ARF (possible, probable or definite) and completed secondary prophylaxis</li> <li>• Borderline RHD not on secondary prophylaxis</li> <li>• Resolved RHD and completed secondary prophylaxis</li> </ul>	<ul style="list-style-type: none"> <li>• 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)</li> <li>• Medical review: yearly until discharge from specialist care and then as required</li> <li>• Dental review: yearly or as required</li> </ul>	<ul style="list-style-type: none"> <li>• Secondary prophylaxis not required</li> </ul>

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## Antibiotic Prophylaxis for Procedures

Antibiotic prophylaxis to prevent infective endocarditis should be given to any patient with echocardiographic evidence of rheumatic heart disease undergoing any of the following procedures: (see [RHD Australia 2020 guideline page 223](#)).

- Dental procedures involving manipulation of the gingival or periapical tissue, or perforation of oral mucosa
- Incision of infected skin lesions
- Invasive respiratory tract, ear, nose and throat procedures to treat established infection
- Tonsillectomy, adenoidectomy
- Gastrointestinal or genitourinary procedures where surgical antibiotic prophylaxis is required or for established infection

### Use single dose of oral Amoxicillin, 60mins before the procedure:

- Adults 2g
- Children 50mg/kg up to 2g

**Note:** If penicillin allergy alternatives include:

- Cephalexin 2g (child:50mg/kg up to 2g) or
- Clindamycin 600mg (child 20mg/kg up to 600mg)

## Secondary Prophylaxis

**Administration of BPG:** for more detail please see [RHD Australia 2020 guideline](#) pages 170-180.

### Use LA- Bicillin (BPG) Every 21 – 28 days:

- Adults and children  $\geq 20$ kg: 1,200,00 units/2.3ml – IM
- Children  $< 20$ kg: 600,000 units/1.2ml - IM

If a patient is seen between 21 and 28 days they should be given their BPG that day rather than waiting until 28 days.

Patients who have confirmed breakthrough ARF, despite full adherence to 4-weekly BPG, should be considered for 3 weekly injections. Discuss with the physician or paediatrician.

### Note:

BPG is superior to any oral prophylaxis and should be used except when there is severe documented allergy to penicillin. In this case, use:

Oral erythromycin 250mg twice a day (all ages).

If possible hypersensitivity to penicillin refer/discuss with paediatrician or physician prior to administration.

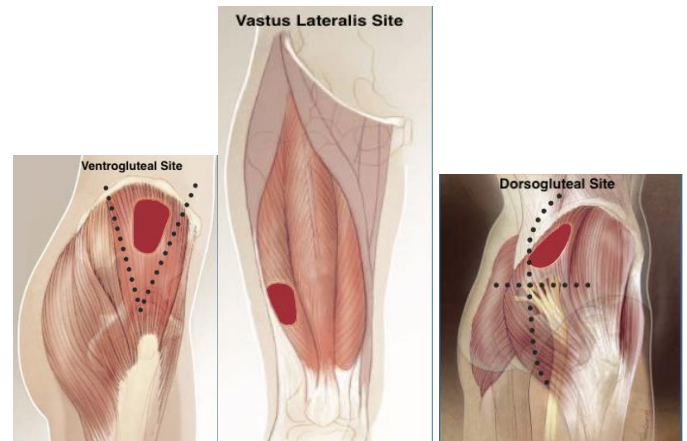
### Notification:

Ensure the [WA RHD Register and Control Program](#) is informed every time the patient receives BPG. For sites using MMEx, the register will be notified automatically if the patient is assigned an RHD care plan, and BPG is marked as **administered** correctly on MMEx.

## Site of injection

### Ventrogluteal site is emerging as the preferred site of injection

- Vastus lateralis site (lateral thigh) is an acceptable site
- Dorsogluteal site has a risk of sciatic nerve injury
- Deltoid is not recommended



(Courtesy of Australian RHD guidelines 3<sup>rd</sup> edition 2020)

## Analgesia for BPG injections

### Non-pharmacological strategies

- Inject slowly
- Distraction technique with videos
- Minimal waiting time for injection
- Allow alcohol swab to dry
- Firm pressure to site for 10 seconds before injecting
- Ice pack to site before injecting
- Vibrating ice pack (e.g. Buzzy bee) if available
- Other distracting stimuli to skin
- Refrigerating needle prior to injection

### Pharmacological strategies (see [RHD Australia 2020 guideline](#) pages 175 – 180 for guidance with patient selection)

- Oral paracetamol before injection, and as needed afterwards
- Local anaesthetic cream/spray applied prior to injection
- Lidocaine/lignocaine can be injected with BPG in patients experiencing moderate to severe pain during or following BPG injection (*N.B. Lidocaine is not funded as an s100 medication and there may be extra cost to the clinic to use it*)
  - Process - Ensure no contraindications (see [RHD Australia 2020 guideline](#) pages 177 – 178 for contraindications and considerations). Prescribe lidocaine. Draw up required dose of BPG into a 3ml syringe. With a new drawing up needle, draw up 0.5ml of 1% lidocaine into the tip of the same syringe. Do not mix (keep lidocaine in the tip). Push plunger to expel air. Attach IM needle and administer injection.
- Nitrous oxide during the injection procedure. 50% nitrous and 50% oxygen. Prescription only. Not for children  $<4$ y.o.
- Oral clonidine can be considered prior to injection in highly distressed patients where other techniques have failed. This should only be done in a monitored primary or tertiary care setting. Seek specialist advice. (See table 10.4 on page 180 of [RHD Australia 2020 guideline](#))

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## Women's Health in RHD

### Pre-pregnancy

#### Contraception

- In high risk adolescents and women who may need future cardiac intervention or surgery, long-acting reversible contraception such as an Intrauterine Contraceptive Device or an etonorgestrel implant should be encouraged.
- Oestrogen containing contraceptives are associated with a higher risk of thrombosis and should be avoided
- Consider tubal ligation in high risk disease if women agree they do not want more children.

#### Pre-conception planning

- Birthing on country if desired should be considered, but may not always be safe depending on the severity of the RHD (defer to local hospital practices)
- Perform clinical examination, functional assessment, echocardiogram and consider exercise testing
- Address iron deficiency, diabetes, cervical screening, vaccination status, gum disease
- Plan early cardiology review if:
  - High risk cardiac disease
    - Symptomatic moderate, or severe RHD (aortic or mitral stenosis)
    - Asymptomatic, clinically significant mitral stenosis
  - Prior cardiac event (heart failure, TIA, stroke, arrhythmia)
  - Mechanical valve or anticoagulation-requiring cardiac disorder
  - Reduced LVEF, pulmonary hypertension
    - Recommend against pregnancy if severe valvular disease, however decision lies with patient

### During pregnancy

#### General principles

- Multidisciplinary team (ensure early referral to [physician](#), [cardiologist](#), [obstetrics](#))
- Address issues as for pre-conception planning
- Encourage good adherence to BPG injections as required
- Cease teratogenic medications
- Monitor and reassess early if new or progressive dyspnoea, orthopnea, reduced exercise tolerance, presyncope or syncope, tachycardia or oedema

#### Birth planning

- Local physician/cardiologist/obstetrician should decide on the safety of local delivery
- Vaginal delivery is usually recommended, unless the patient is on oral anticoagulation, is in preterm labour, has severe heart failure or severe pulmonary hypertension
- Infective endocarditis prophylaxis should be provided as per the above recommendation

#### Termination

- May be medically indicated in women with severe disease and at high risk of pregnancy or birth complications. Seek specialist advice and ensure patient support available

#### Patients requiring anticoagulation (discuss with specialist team)

- Consider continuing warfarin in mechanical valves unless dose >5mg (higher doses > risk of embryopathy)

- If not appropriate to continue warfarin, consider Low Molecular Weight Heparin (LMWH) 1mg/kg BD (note: dose adjustments required in renal disease)
- If the patient takes a DOAC, consider changing to LMWH

## Refer/Discuss

### The WA RHD Register and Control Program

**Both ARF and RHD are notifiable diseases as per WA health regulations**

All patients with suspected or confirmed RHD should be known to the [Western Australian Rheumatic Heart Disease Register and Control Program](#).

- Phone: 1300 622 745
- Fax: (08) 65530899
- Email: [RHDRegister@health.wa.gov.au](mailto:RHDRegister@health.wa.gov.au)

For notification of a new case – complete and send this [ARF and RHD notification form](#) to the email address above.

The register can also be contacted to help find recent health events for any RHD patient.

The register must be provided a copy of **all diagnostic tests (including echocardiogram reports), and specialist (cardiologist, paediatrician or physician) letters within 30 days from date of report**, and notified of **all secondary prophylactic BPG events that occur, for every patient**

### Kimberley Specialist Teams: available for local advice

#### Kimberley Regional Physician Team

- Phone: Broome Hospital switch – 9194 2222
- Email: [krpt@health.wa.gov.au](mailto:krpt@health.wa.gov.au)
- MMEX - Kimberley regional physician team

#### Kimberley Regional Paediatric Team

- Phone: Broome Hospital switch – 9194 2222
- Direct on call phone: 0427988570
- Email: [kimberley.paediatrics@health.wa.gov.au](mailto:kimberley.paediatrics@health.wa.gov.au)

#### Kimberley Regional Obstetric Team

- Phone: Broome Hospital switch – 9194 2222

### Visiting Cardiology Services

#### Perth Cardiovascular Institute

(Adults only. Liaise with regional paediatric team for children)

- Phone: (08) 6314 6820
- Email: [Kimberley@perthcardio.com.au](mailto:Kimberley@perthcardio.com.au)

## Resources

[The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease \(3rd edition\)](#)

RHD Australia Website: <https://www.rhdaustralia.org.au>

Download the APP – 'ARF RHD Guideline' from the [App store](#) or [Google Play](#)