Rheumatic Heart Disease (RHD)

Screening

Everyone with confirmed or suspected acute rheumatic fever (ARF) AND / OR High risk population adults with a murmur need echocardiography, particularly in pregnancy, to exclude or diagnose rheumatic heart disease (RHD). High risk population children need paediatric review to determine need for further investigation including echocardiography.

Case Definition

Echocardiogram with valve changes consistent with RHD (tethering/thickening of mitral valve and/or combined mitral and aortic valve damage , see 2012 ARF/RHD National Guide and below table).

Classification	Criteria
Priority 1 - Severe RHD	Severe valvular disease or Moderate/severe valvular lesion with symptoms Or Mechanical prosthetic valves, tissue prosthetic valves and valve repairs including balloon valvuloplasty
Priority 2 - Moderate RHD	Any moderate valve lesion in the absence of symptoms and with normal LV function
Priority 3 - ARF (no RHD) or Mild RHD	ARF with no evidence of RHD or Trivial to mild valvular disease
Priority 4 - Inactive	Patients with a history of ARF (no RHD or trivial to mild valvular disease) for whom secondary prophylaxis has been ceased

Symptoms of valvular RHD:

- Shortness of breath on exertion.
- Orthopnoea, paroxysmal nocturnal dyspnoea.
- Syncope.
- Oedema.
- Chest pain

Principles of Management

- Prevent recurrences of ARF with benzathine penicillin every 21-28 days (see ARF).
- Prevent endocarditis with good dental hygiene, regular dental review, and prophylaxis when required.
- Monitor and manage heart failure (see HEART FAILURE).
- Prevent thromboembolic complications.
- Ensure appropriate management. Valve repair/replacement typically required for high severity disease.
- Timely referral for ECHO and medical/specialist review
- Identify pregnant women with RHD and manage appropriately (see RHD IN PREGNANCY).
- Always highlight RHD on referrals.

Therapeutic Protocols

Confirm diagnosis and make sure a RHD care plan has been assigned, the WA RHD Register has been notified and reports/investigations sent to them (see MANDATORY REPORTING).

Ensure pneumococcal / influenza vaccines up to date.

Anticoagulate everyone with mechanical valves. Generally aim for INR 2.5 - 3.5 but always review and be guided by cardiothoracic discharge summary.
There is currently no evidence for new oral anticoagulants (NOACs) in valvular heart disease They are therefore NOT recommended for use in RHD.

Antibiotic prophylaxis to prevent endocarditis:

Prophylaxis should be given to any patient with a valvular lesion no matter what the priority is (not needed if there is confirmed ARF but no valvular lesion).

For all dental procedures or incision of infected skin lesions use **single dose of clindamycin orally 1 hour** before procedure:

- Adults 600mg.
- Children 15-20mg/kg round up to nearest 150mg (max 600mg per dose) Refer to dosing guidelines for children requiring less than 150mg.

Secondary prophylaxis to prevent recurrent episodes of ARF:

Every 21 – 28 days Benzathine penicillin

Use

LA Bicillin 900mg/2.3ml

Dose:

Adults and children ≥ 20kg: 900mg 4 weekly.

Children < 20kg: 450mg 4 weekly.

NB: If possible hypersensitivity to penicillin refer/discuss with paediatrician or physician.

Benzathine penicillin is superior to any oral prophylaxis and should be used except when there is severe documented allergy to penicillin when oral erythromycin 250mg twice a day is indicated (all ages).

It is recommended that LA Bicillin injections are given **21 - 28 days.**

It is important that if a patient is seen between 21 and 28 days that they are given their LA Bicillin that day rather than waiting until 28 days.

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

Ensure the patient's name is recorded on the clinic list from the register and faxed through at the end of the month to (08) 9193 5260

Rheumatic Heart Disease (RHD)

DURATION OF PROPHYLAXIS:

All persons with probable ARF	Highly-suspected ARF: Minimum of 10 years after most recent suspected episode of ARF or until alternative diagnosis confirmed Uncertain ARF: 12 months after diagnosis, and then reassess (including echocardiography) at that time. If evidence of RHD at that time, manage as for highly-suspected ARF. If no evidence of RHD, consider ceasing prophylaxis
RHD Priority 3	Minimum 10 years after most recent episode of ARF or until age 21 years (whichever is longer)
RHD Priority 2	Continue until 35 years of age or 10 years after last episode of ARF (whichever is longer)
RHD Priority 1	Continue until 40 years old or 10 years after last episode of ARF (whichever is longer).

NB: *Patients >25 years of age who are diagnosed with RHD, and without any documented history of prior ARF, should receive prophylaxis until the age of 35. At this time, they should be reassessed to determine whether prophylaxis should be continued.*

NB: Decisions to cease prophylaxis should be based on specialist and echocardiographic assessment.

Follow up

Review frequency according to PRIORTY:

Classification	Review & management plan
Priority 1 (severe)	3-4 weekly: LA Bicillin 3-6 monthly: GP review 6 monthly: specialist review (cardiology/ physician/ paed) dental review, Echo Yearly: influenza vaccine and check pneumococcal vaccination up to date. Educate patient about RHD/ARF PRN: Endocarditis prophylaxis
Priority 2 (moderate)	3-4 weekly: LA Bicillin 6 monthly: GP and dental review Yearly: Echo, Specialist review (cardiology/physician/paed), Influenza vaccine and check pneumococcal vaccination. Educate patient about RHD/ARF PRN: Endocarditis prophylaxis
Priority 3 (mild)	3-4 weekly: LA Bicillin Yearly: GP review, Dental review, Influenza vaccine, check pneumococcal vaccination, educate patient about RHD/ARF 2-3 yearly: Echo and specialist review
Priority 4 (inactive)	Yearly: GP and dental review

Women of Child Bearing Age

Pregnancies should be planned given the increased risks associated with RHD in pregnancy.

Contraception should be discussed with all women who have moderate or severe RHD

Physician / cardiology review before planned pregnancy and early in unplanned pregnancy.

RHD in Pregnancy

The circulatory changes of pregnancy will exacerbate any pre-existing valvular disease. Sometimes RHD is first diagnosed during pregnancy or soon after delivery when a woman develops symptoms, usually dyspnoea.

- Arrange echo at first visit and again in the third trimester.
- Ensure influenza and pneumococcal vaccinations are current.
- Monitor for signs of heart failure at each visit.
- LAB is safe in pregnancy – make sure it is continued
- If on warfarin discuss options for anticoagulation in pregnancy with physician/cardiologist
- Cease meds that are tetratogenic – e. g. ACE
- Referral early to physician/cardiologist/obstetrician (high risk clinic). They will decide whether safe to deliver locally
- Discuss contraception early (especially if current pregnancy was unplanned)
- Prophylaxis for endocarditis is required for surgical termination.

Rheumatic Heart Disease (RHD)

Refer/ Discuss

TO PHYSICIAN / PAEDIATRICIAN /CARDIOLOGIST

Recurrent ARF.

Endocarditis (unexplained fever).

Worsening valve lesion (shortness of breath / dizziness).

Thromboembolism (stroke or DVT).

Penicillin hypersensitivity or absolute injection refusal

Pregnancy (plus refer to OBSTETRICIAN as well).

Reporting to the RHD Register

ARF/RHD is a notifiable disease. Under WA Health (*Rheumatic Heart Disease Register of Western Australia*) Regulations 2015 you must report this diagnosis to the WA RHD Register - see forms in MMEx or online http://www.public.health.wa.gov.au/cproot/2848/2/acute_rheumatic_fever_notification_form_june2014.pdf

Patient consent is no longer required for notification and failure to notify can result in fines.

Completed notifications can be sent to the register by either fax or email

- fax to 9193 5260 or
- email RHDRegister@health.wa.gov.au

You must also provide a copy of each diagnostic test (including an echocardiogram) within 15 days for a new diagnosis and 30 days for a recurrence.

Ongoing reports from cardiologists/physicians/ paediatricians and surveillance echoes also need to be sent through to the register as they occur.

Patients who have had more than one episode require a new notification to be made for EACH new episode.

Pregnancy should be notified to the register in the third trimester.

For further information please call the WA RHD Register on 1300 622 745 or email RHDRegister@health.wa.gov.au or view http://ww2.health.wa.gov.au/Articles/U_Z/WA-rheumatic-heart-disease-register

Resources

RHD Australia Website – www.rhdaustralia.org.au

Download the app – "ARF / RHD Guideline" from Google Play or the App Store or follow the link from the RHD Australia Website