TITLE: RHEUMATIC HEART DISEASE KIMBERLEY CHRONIC DISEASE GUIDELINE

1. GUIDING PRINCIPLES

This guideline is applicable only to eligible Aboriginal and Torres Straight Island Health Services in remote areas of the Kimberley region of Western Australia approved under Section 100 of the *National Health Act 1953* that makes special provision for supply of medications to patients/clients.

The guideline was designed for use in conjunction with the Kimberley Standard Drug List (KSDL) which is a standardised list of medications available on the Pharmaceutical Benefits Scheme and Section 100 throughout the Kimberley, including remote area clinics.

2. GUIDELINE

2.1 Case Definition

Acute Rheumatic Fever (ARF) is an illness due to an autoimmune reaction caused by a bacterial infection with Group A Streptococcus, affecting the heart, brain, joints and skin. The acute infection often results in lasting damage to the heart valves. This is known as Rheumatic Heart Disease (RHD). People who have had ARF are more likely to have subsequent episodes, resulting in further damage to the valves. Almost all cases of ARF and resulting RHD are preventable. Australian Indigenous people are up to 8 times more likely than non-Indigenous people to require hospitalisation for ARF and RHD and 20 times more likely to die from these illnesses. The prevalence of RHD peaks in the 3rd and 4th decade, thus management and monitoring is ongoing[1].

Diagnosis of RHD is based on echocardiogram, with valve changes consistent with RHD[1].

2.2 Screening

Everyone with confirmed or suspected Acute Rheumatic Fever and / or Aboriginal and Torres Strait Islander people with a murmur need an Echocardiogram to exclude RHD[1].

The use of the term “Aboriginal” within this document refers to Australians of both Aboriginal and Torres Strait Islander people.

2.3 Priority rating[1]

PRIORITY 1 - Severe	PRIORITY 2 - Moderate	PRIORITY 3 - Mild	PRIORITY 4 - Inactive
Severe valvular disease **or** Moderate/severe valvular lesion with symptoms* **or** Mechanical prosthetic valves, tissue prosthetic valves and valve repairs including balloon valvuloplasty	• Moderate valve lesion in the absence of symptoms* and with normal LV function	ARF with no evidence of RHD clinically or on echocardiogram **or** Trivial to mild valvular disease	History of ARF (no RHD) for whom secondary prophylaxis has been ceased and who no longer require specific follow-up for ARF/RHD related conditions as determined by regional physician or paediatrician.

***Symptoms of valvular RHD include:**

- shortness of breath on exertion
- orthopnoea
- paroxysmal nocturnal dyspnoea
- syncope
- peripheral oedema
- chest pain.

2.4 Principles of Management[1, 2]

- Timely referral for echocardiography
- Timely referral of RHD to physician/paediatrician and/or cardiologist
- Prevent recurrences of ARF and RHD with penicillin prophylaxis
- Prevent Endocarditis and further damage to heart valves with good dental hygiene, and regular dental review
- Monitor and manage heart failure
- Prevent thromboembolic complications from atrial fibrillation or mechanical prosthetic valves with adequate monitoring of anticoagulation
- Ensure appropriate referral for surgical assessment. Valve repair, valvuloplasty or replacement is typically required for severe disease and may be appropriate in some cases for less severe disease.

2.5 Therapeutic Guidelines[1]

Register all patients with confirmed RHD on the WA State Register. Forms available here.

Ensure pneumococcal / influenza vaccines are up to date.

Know patient's type of valve repair /replacement:

- Mitral valve repair
- Valvuloplasty
- Bioprosthetic valve
- Mechanical valve.

Anticoagulate everyone with a mechanical valve. Aim for an INR of 2.0-3.0 for a mechanical aortic valve and an INR of 2.5-3.5 for a mechanical mitral valve.

All people with prosthetic valves or established RHD need antibiotic prophylaxis prior to procedures to prevent endocarditis[1]:

- all dental, oral and respiratory procedures or incision of infected skin lesions:
  - use single dose Clindamycin capsule orally one hour prior to the procedure
    - Adults: 600mg
    - Children: 15mg/kg, up to 600mg*. For patients requiring less than 150mg please refer to relevant dosing guidelines.
- Genitourinary and gastrointestinal procedures: give Vancomycin IV by slow infusion over one hour just prior to procedure:
  - Adults and children 12 years and older: 1.5g
  - Children under 12 years: 30mg/kg up to 1.5g

*See explanatory notes on medications below.

SECONDARY PROPHALAXIS TO PREVENT FURTHER ARF[1]:

- Every four weeks of Benzathine Penicillin 900mg if ≥20kg, 450mg if <20kg for most patients (see explanatory notes on medications for formulations)
- Patients at high risk of recurrent ARF should be discussed with physician for consideration of 3 weekly Benzathine Penicillin. These groups are:
  - patients who have confirmed breakthrough of ARF, despite full adherence to 4-weekly Benzathine Penicillin
  - patients with moderate or severe carditis, or a history of valve surgery, who demonstrate good adherence to four weekly injections.
- Benzathine Penicillin is superior to any oral prophylaxis, and should be used unless there is a documented severe allergy to penicillin. Patients should be carefully questioned regarding penicillin allergy prior to treatment, and if there is no unequivocal evidence for an allergy, should be referred for physician/paediatrician review, as most patients are not truly allergic.
- If hypersensitive to penicillin or absolute refusal of injection, refer to physician/paediatrician for decision on alternative medication.

Duration of Prophylaxis

- Timing of prophylaxis cessation will ultimately be decided by physician, in consultation with the patient[2].

- All patients should have an evaluation for deterioration of symptoms and an echocardiogram BEFORE stopping prophylaxis to reassess severity of RHD. Ideally echocardiogram should be performed before physician review.

Duration of Prophylaxis[1].

See description of priority grading above.
Decisions to cease secondary prophylaxis should only be made under the instruction of the regional physician based on clinical and echocardiographic assessment.

Category	Duration of Prophylaxis
All persons with ARF or RHD*	Minimum 10 years after most recent episode of ARF or until age 21 years (whichever is longer)
Status after initial period elapsed:	
No RHD	Discontinue at that time
Mild RHD	Discontinue at that time
Moderate RHD	Continue until age 35 years or 10 years after most recent episode ARF (whichever is longer)
Severe RHD	Continue until age 40 years or 10 years after most recent episode ARF (whichever is longer)

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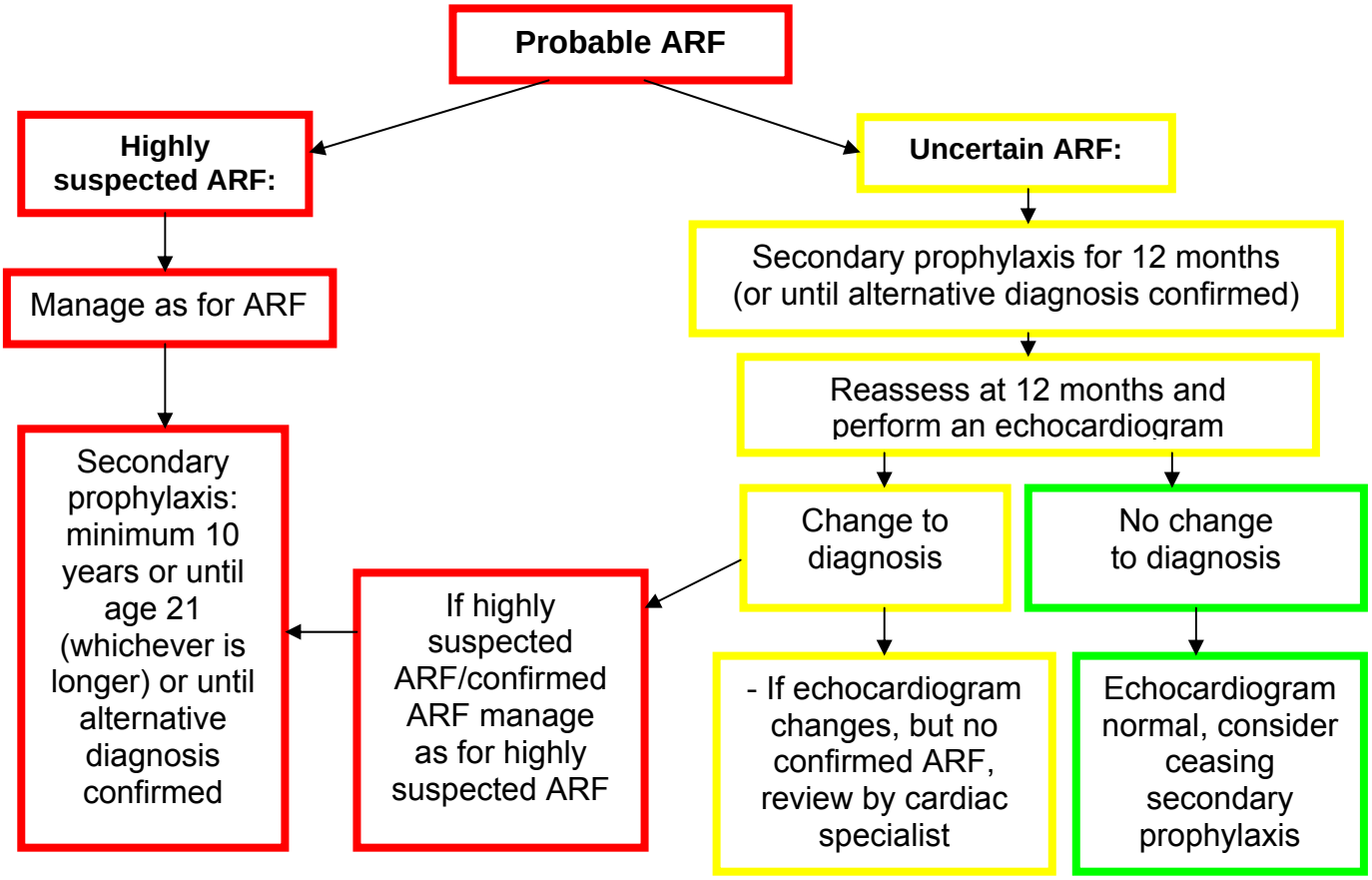
*Patients >25 years of age who are diagnosed with RHD, without any documented history of prior ARF, should receive prophylaxis until age 35 years. At this time, they should be reassessed to determine whether prophylaxis should be continued.

Duration of Secondary Prophylaxis for Patients with "Probable ARF"[1]
(see ARF guideline)



Prevent skin sores – treat the client and family early and often for scabies and impetigo[2].

2.6 Follow-Up[2]

Review frequency according to severity

- Priority 1
  - 3-4 weekly secondary prophylaxis
  - 3-6 monthly review by a doctor
  - Physician / Paediatrician review six monthly
  - Cardiology review annually
  - Dental review six monthly
  - Echocardiogram annually or more frequently as determined by physician/paediatrician
  - Influenza annually and ensure pneumococcal vaccination up to date.

- Priority 2
  - Four weekly secondary prophylaxis
  - Six monthly review by a doctor
  - Dental review 6 monthly
  - Annual echocardiogram
  - Physician / Paediatrician review annually
  - Influenza annually and ensure pneumococcal vaccination up to date.

- Priority 3
  - Monthly secondary prophylaxis
  - Annual doctor review
  - Dental review annually[1]
  - Influenza annually and ensure pneumococcal vaccination up to date
  - 2-3 yearly echocardiogram and specialist review.

2.7 Women of child bearing age[1]

- Circulatory changes during pregnancy will exacerbate any existing valvular disease.
- Discuss effective contraception use with all patients.
- Ideally, pregnancy should be planned.
- Physician / cardiology / obstetrician review **before** planned pregnancy and early in an unplanned pregnancy.
- The implications of pregnancy need to be discussed with all women with moderate to severe valve disease or with valve replacements on warfarin.
- Women requiring continuous anticoagulation (i.e. AF or mechanical valve) should not have anticoagulation ceased. Urgent discussion with physician required to determine anticoagulation regime.
- Some pregnant women may develop CCF, especially in the third trimester. ACEIs and ARBs are contraindicated in pregnancy, thus hydralazine, nitrates or nifedipine recommended if vasodilation is required, under guidance from physician.
- Digoxin or beta blockers may be used for rate control in atrial fibrillation, under guidance from physician.
- Secondary prophylaxis is safe and should be continued in pregnancy[1, 2].
- Antibiotic prophylaxis to prevent endocarditis in prolonged labour/prolonged rupture of membranes.
- Women on heparin or warfarin can breastfeed.
- All women with mitral stenosis should be referred for specialist review prior to pregnancy, as surgical intervention may be required.

2.8 Refer / Discuss

TO PHYSICIAN / PAEDIATRICIAN / CARDIOLOGIST[1,2, 3]:

- Recurrent ARF
- Endocarditis (unexplained fever)
- Worsening valve lesion or signs of cardiac failure (e.g. shortness of breath / dizziness)
- Embolus (stroke or leg pain)
- Pregnancy.
- Possible penicillin hypersensitivity/allergy or absolute injection refusal

TO OBSTETRICIAN:
If planning pregnancy or in early pregnancy[2].

3. ADDITIONAL ROLES AND RESPONSIBILITIES FOR INDIVIDUAL CLINICIANS

Physician/Cardiologist[1]:

- One to two yearly review (depending on severity).Patients with severe disease may require review six monthly.
- A patient with recurrent ARF needs to be assessed and managed by the physician/cardiologist.
- A patient with endocarditis (unexplained fever) needs to be assessed and managed by the physician/cardiologist.
- A patient with worsening valve lesion (shortness of breath / dizziness) needs to be assessed and managed by the physician/cardiologist.
- A patient planning pregnancy or pregnant needs to be assessed and managed by the physician/cardiologist.
- Utilise interpreter services, if appropriate. Call the Translating and Interpreter Service (TIS) on 13 14 50.
- Liaison with multidisciplinary team.

Paediatrician[1]:

- One to two yearly review (depending on severity).Patients with severe disease may require review six monthly.
- A patient with recurrent ARF needs to be assessed and managed by the paediatrician.
- A patient with endocarditis (unexplained fever) needs to be assessed and managed by the paediatrician.
- A patient with worsening valve lesion (shortness of breath / dizziness) needs to be assessed and managed by the paediatrician.
- Review if penicillin hypersensitivity or absolute injection refusal
- Utilise interpreter services, if appropriate. Call the Translating and Interpreter Service (TIS) on 13 14 50.
- Liaison with multidisciplinary team.

Obstetrician:

- A patient planning pregnancy or pregnant needs to be assessed and managed by the obstetrician.
- Utilise interpreter services, if appropriate. Call the Translating and Interpreter Service (TIS) on 13 14 50.
- Liaison with multidisciplinary team.

Medical Officer:

- Review interval will depend on patient's condition and stability, but patient should be reviewed by a medical officer (or nurse practitioner) at least yearly. Patients with severe RHD may require review every three months[2].
- If medication is being titrated, the patient should be reviewed at least every two weeks by a medical officer (or nurse practitioner).
- Refer patient to physician or obstetrician if they meet the requirements stated in the chronic disease guideline.

- Order blood or other tests at appropriate times according to the guideline, and the patient's clinical picture. Review of the results and arrange follow-up as per results and clinical picture.
- Give appropriate education with regard to patient's condition, co-morbidities, other risk factors, lifestyle issues and medications.
- Offer patient support with regard to the management of their condition, including self-management where appropriate, and other issues affecting lifestyle.
- Commence the patient on treatment as per clinical picture and guideline.
- Commence on, or update the patient's care plan in accordance with their clinical picture, and the guideline.
- Engage in brief interventions, and make use of the Lifescripts as appropriate.
- Utilise interpreter services, if appropriate. Call the Translating and Interpreter Service (TIS) on 13 14 50.
- Liaison with multidisciplinary team.

Remote Nurse Practitioner:

The scope of practice of the remote nurse practitioner is defined by:

- The ordering of pathology, radiology and referral for routine specialist review as outlined within this guideline.
- The management of ongoing care of the client whose pathology and radiology results are within normal limits, which includes the provision of a further 12 month prescription for the medications outlined within this guideline.
- All clients must be reviewed by a medical officer annually.
- Review interval will depend on client's condition and stability, but patient should be at least yearly (unless patient has been seen by a medical officer). Patients with severe RHD may require review every three months[2].
- Give appropriate education with regard to their condition, co-morbidities, other risk factors, lifestyle issues and medications.
- Offer patient support with regard to the management of their condition, including self-management where appropriate, and other issues affecting lifestyle.
- Commence the patient on treatment as per clinical picture and guideline.
- Commence on, or update the patient's care plan in accordance to their clinical picture, and the guideline.
- Engage in brief interventions, and make use of the Lifescripts as appropriate.
- Discuss any concerns about patient and / or treatment with medical officer, or relevant specialist.
- Timely and appropriate referral of patient.
- Commence on, or update the patient's care plan in accordance with their clinical picture, and the guideline.
- Utilise interpreter services, if appropriate. Call the Translating and Interpreter Service (TIS) on 13 14 50.
- Liaison with multidisciplinary team.

Remote Area Nurse:

- Review four weekly for prophylaxis, or according to patient's regime[1].
- Three monthly observations / assessment.
- Give appropriate education with regard to patient's condition, co-morbidities, other risk factors and lifestyle issues.
- Offer patient support with regard to the management of their condition, including self-management where appropriate, and other issues affecting lifestyle.
- The taking of bloods / urine as ordered by the medical officer or nurse practitioner.
- Engage in brief interventions, and make use of the Lifescript[6] as appropriate.
- Liaison with multidisciplinary team.

Aboriginal Health Worker

- Review four weekly for prophylaxis, or according to patient's regime[1].
- Three monthly observations / assessment.
- Give appropriate education with regard to patient's condition, co-morbidities, other risk factors and lifestyle issues.
- Offer patient support with regard to the management of their condition, including self-management where appropriate, and other issues affecting lifestyle.
- The taking of bloods / urine as ordered by the medical officer or nurse practitioner.
- Engage in brief interventions, and make use of the Lifescripts as appropriate.
- Liaison with multidisciplinary team.

4. EXPLANATORY NOTES ON CHOICE OF MEDICATIONS

CLINDAMYCIN
As clindamycin liquid is difficult to access, it is recommended to give clindamycin tablets to children at dosages of 150mg tablet increments, up to maximum 600mg. It is recommended to round *up* to the nearest 150mg.

5. REFERENCES

1. RHD Australia, (ARF/RHD writing group), National Heart Foundation and Cardiac Society of Australia and New Zealand, Australian Guideline for Prevention, Diagnosis and Management for Acute Rheumatic Fever and Rheumatic Heart Disease (2nd Edition) 2012.
   Level 1-4 evidence, in accordance to the NHMRC guidelines.
2. Queensland Health, Royal Flying Doctor Service of Australia (Queensland Section) and Apunipima Cape York Health Council, Chronic Disease Guidelines, 3rd edition, 2010. Cairns.
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3. CARPA (2009) CARPA Standard Treatment Manual, 5th edition. Alice Springs Central Australian Rural Practitioners Association Inc.
4. Kimberley Aboriginal Medical Services Council and WA Country Health Service (WACHS) Kimberley. Kimberley Standard Drug List. 6th ed. 2010.
5. Commonwealth Consolidated Acts. *National Health Act 1953* - Sect 100 [Internet] 2010 [cited 2011 August 3].