ACUTE RHEUMATIC FEVER AND RHEUMATIC HEART DISEASE: MANAGEMENT FOR 12 MONTHS POST-DIAGNOSIS

PRACTICE GUIDELINE ©

DOCUMENT SUMMARY/KEY POINTS

- Acute Rheumatic Fever (ARF) and Rheumatic Heart Disease (RHD) present infrequently in NSW, and therefore there is little familiarity with the post-diagnosis management and prophylaxis required.
- ARF and RHD disproportionately affect the Aboriginal and Torres Strait Islander, Maori and Pacific Islander populations.
- The recurrence of ARF and progression of RHD can be prevented through antibiotic prophylaxis, education, family-centred and culturally safe support, and a trusted, consistent Primary Care Provider (General Practitioner or Aboriginal Community Controlled Health Service)

CHANGE SUMMARY

- Not applicable - new document
- **25/10/21** Minor review: Title corrected from "…Health disease" to "…Heart disease"

READ ACKNOWLEDGEMENT

- All medical, nursing and pharmacy staff working within Ambulatory Care, Infectious Diseases, Cardiology and General Medicine are to read and acknowledge they understand the contents of this document.

Approved by:	SCHN Policy Procedure and Guideline Committee	
Date Effective:	1st June 2021	**Review Period:** 3 years
Team Leader:	Fellow	**Area/Dept:** Ambulatory Care

Date of Publishing: 25 October 2021 8:29 AM Date of Printing: 25 October 2021

TABLE OF CONTENTS

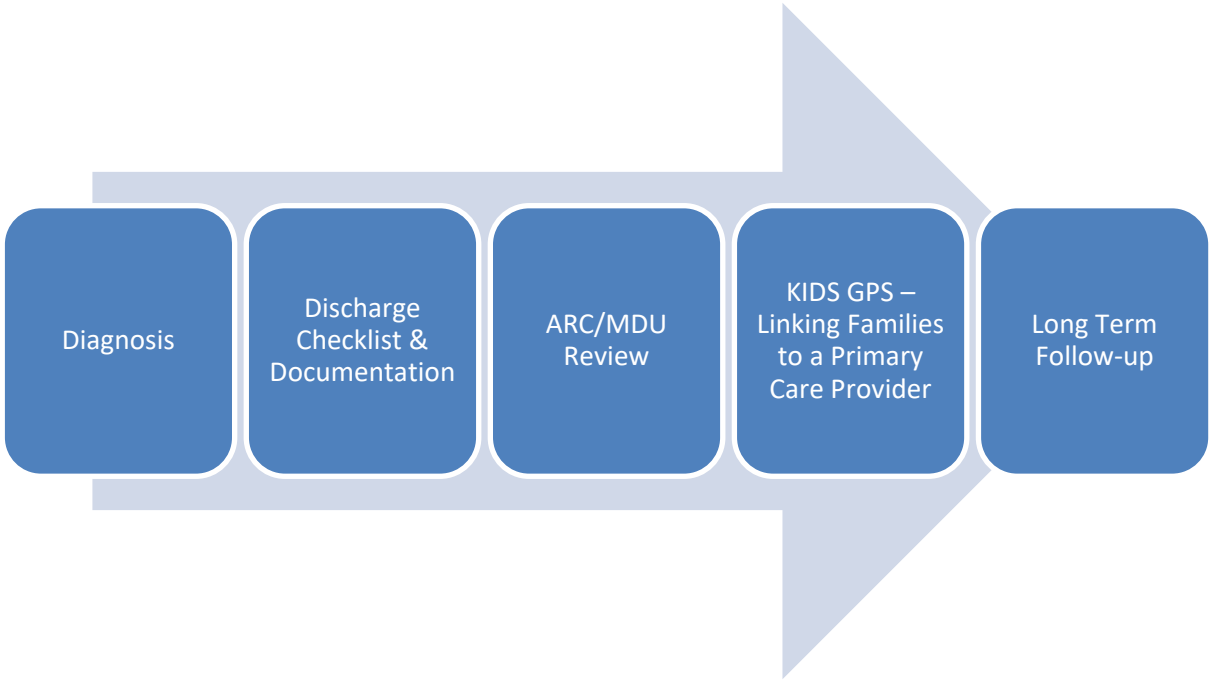
1 Aims

- To reduce the recurrence of Acute Rheumatic Fever in children by:
  - Providing culturally safe, family-centred support
  - Facilitating linkage with an appropriate Primary Care Provider for long term clinical management, prophylaxis and follow-up
  - Providing education to the patient, family and Primary Care Provider
- Scope
  - Any child seen at SCHN with Acute Rheumatic Fever or Rheumatic Heart Disease who is:
    - Newly diagnosed, or
    - Not already receiving long-term management and prophylaxis under a current Primary Care Provider with demonstrated adherence

This guideline is predominantly informed by RHD Australia's "ARF RHD Guideline: The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3rd edition)".

https://www.rhdaustralia.org.au/arf-rhd-guideline

2 Patient Flow through Ambulatory Services



3 Diagnosis

The diagnosis of Acute Rheumatic Fever (ARF) is made by the treating team in consultation with the Infectious Disease team, as well as other teams such as Orthopaedics, Rheumatology and Cardiology, as appropriate.

The diagnosis is made in accordance with the National "ARF RHD Guideline"

- Found here: https://www.rhdaustralia.org.au/arf-rhd-guideline (Chapter 6, page 72)
- **Please note:**
  - The diagnostic criteria are different for low- and high-risk populations (page 73 & 74)
  - ARF diagnosis is categorised as "Definite", "Probable" and "Possible" (page 79)
  - Differential diagnoses for consideration (page 91)

The diagnosis of Rheumatic Heart Disease (RHD) is made by the Cardiology team on the basis of an echocardiogram and the team's clinical review.

The diagnosis is made in accordance with the National "ARF RHD Guideline"

- Found here: https://www.rhdaustralia.org.au/arf-rhd-guideline (Chapter 8, page 130)

4 Discharge Checklist & Documentation

For patients presenting with ARF, please complete **ALL** below steps during the inpatient admission and document each step in the Discharge Summary. A simplified checklist is provided in the appendix for your convenience.

For patients first encountered in an outpatient setting, such as Cardiology Clinic, without concerns for ARF but only RHD, please complete steps 2-6 and document in the clinic letter or other appropriate documentation. The remainder of the steps will be completed as part of the Acute Review Clinic appointment.

1. **Commence treatment of inciting streptococcal infection with 10-day course of oral phenoxymethylpenicillin or single dose intramuscular (IM) benzathine benzylpenicillin**
   - In consultation with Infectious Diseases
   - See below table for dosing and alternatives for use in cases of penicillin hypersensitivity

Indication	Medication options listed in order of preference	Comment
Eradication of inciting streptococcal infection	1. Benzathine benzylpenicillin G (BPG) Child <20 kg: 600,000 unit Child ≥20 kg: 1,200,000 units IMI single dose	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.
	2. Phenoxymethylpenicillin Child: 15 mg/kg up to 500 mg orally 12-hourly for 10 days	
	3. Penicillin hypersensitivity (non-severe): Cefalexin Child: 25 mg/kg up to 1 g orally 12-hourly for 10 days	Between 3 and 30% of Group A Streptococcus isolates internationally are resistant to macrolide antibiotics (e.g. azithromycin).
	4. *Immediate penicillin hypersensitivity*: Azithromycin Child: 12 mg/kg up to 500 mg orally daily for 5 days	

Adapted from: RHDAustralia (ARF/RHD writing group). The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3rd edition); 2020 pp105-106 (https://www.rhdaustralia.org.au/arf-rhd-guideline)

2. Complete assessment for Carditis / RHD

- Ensure ECG, echocardiogram and Cardiology review are attended prior to discharge with a follow-up plan arranged

3. Confirm plan for secondary prophylaxis

- Standard secondary prophylaxis will be IM benzathine benzylpenicillin given every 21 days.

Antibiotic	Dose	Route	Frequency
Benzathine benzylpenicillin G (BPG)	Child <20 kg: 600,000 unit Child ≥20 kg: 1,200,000 units	Deep intramuscular injection	Every 21 days (NB. This frequency differs from the RHD Australia policy. 21 days will be used as initial frequency but may be increased to 28 days pending demonstration of compliance with treatment and infectious diseases review)

Adapted from: RHDAustralia (ARF/RHD writing group). The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3rd edition); 2020 pp167 (https://www.rhdaustralia.org.au/arf-rhd-guideline).

- See the National "ARF RHD Guideline" Chapter 10, Table 10.1, page 167 for alternatives in cases of penicillin hypersensitivity
- Timing of first IM prophylaxis dose: If oral phenoxymethylpenicillin is commenced for treatment of acute rheumatic fever, the first secondary prophylaxis IM benzathine benzylpenicillin dose will need to be administered PRIOR to the end of the 10 day oral course

- Duration: Minimum duration to be advised by Infectious Diseases/Cardiology team, total duration to be reassessed at Infectious Diseases/Cardiology follow-up
- Analgesia: IM injection is painful, multimodal analgesia is required. See the National "ARF RHD Guideline" Chapter 10, page 177 for lidocaine dosing to accompany IM injections if required

4. Refer to Acute Review Clinic (ARC) / Medical Day Unit (MDU)

- ARC (CHW) referral: Refer via Powerchart → "Orders" → "ARC Referral"
- MDU (SCH) referral: Contact MDU on 9382 0223 to book a review appointment AND submit a Request for Admission (RFA) for the date of the appointment
- Timing: To coincide with first secondary prophylaxis IM benzathine benzylpenicillin dose
- Order "Future Orders" in Powerchart for IM benzathine benzylpenicillin, IM lidocaine and oral paracetamol, for the date of the ARC / MDU review
- Order repeat inflammatory markers if required (if not normalised by discharge), for the date of the ARC / MDU review

5. Refer to Kids GPS

- Powerchart → "Orders" → "Kids GPS Referral"
- Ensure the Primary Care Provider details are correct on Powerchart under "Patient Summary" → "Providers"
- Kids GPS contact details:
  - Email: schn-kidsgps@health.nsw.gov.au
  - Phone:
    - CHW: 9845 2526
    - SCH: 9382 1714

6. Notification

- Make a notification for ARF / RHD to the local Public Health Unit
  - https://www.health.nsw.gov.au/Infectious/Forms/acute-rheumatic-fever-form.pdf
  - The Children's Hospital at Westmead:
    Western Sydney LHD Public Health Unit – Fax: 9840 3591
  - Sydney Children's Hospital at Randwick:
    South Eastern Sydney LHD Public Health Unit – Fax: 9382 8314

7. Registration

- Provide education about the RHD Register to families
  - https://www.health.nsw.gov.au/Infectious/rheumatic/Publications/rhd-information-brochure.pdf

- Obtain consent from the family for Registration on the RHD Register
  - https://www.health.nsw.gov.au/Infectious/rheumatic/Pages/consent.aspx
- If consent is provided, fax the completed registration form to the local Public Health Unit (contact details as above)

8. Refer to Cardiology Outpatient Clinic for Follow up

- The subsequent echocardiogram should occur 12 months after diagnosis at the latest, in accordance with the National "ARF RHD Guideline", Chapter 7, Table 7.4, page 114, or as directed by the Cardiology team

9. Refer to Infectious Diseases Outpatient Clinic for Follow up

- Request the first clinic appointment for 3 months after diagnosis, or as directed by the Infectious Diseases team at time of referral

10. Offer Referral to an Aboriginal Health Worker if appropriate

- Ask the family whether they identify as Aboriginal and/or Torres Strait Islander and offer referral to an Aboriginal Health Worker if appropriate

11. Offer Referral to Social Work

- Offer referral to Social Work if the family might benefit from additional support including with aspects not directly related to the diagnosis

12. Provide initial education to the family

- Watch "Michael's Story" with the family and patient and address questions
  - https://www.rhdaustralia.org.au/individuals-families
- Provide written educational information
  - https://www.health.nsw.gov.au/Infectious/factsheets/Factsheets/rheumatic-heart-disease.pdf

13. Outpatient Community Dental Review

- All children in the household should have a dental review annually
- Provide information to help the family access the Child Dental Benefits Schedule
  - https://www.health.nsw.gov.au/oralhealth/Pages/child-dental-benefits-schedule.aspx
- Provide the family with the phone number for their local public dental clinic (search in ***google chrome***):
  - https://www.health.nsw.gov.au/oralhealth/Pages/call-centre-search.aspx
- NOTE: endocarditis prophylaxis is recommended for some cardiac conditions and dental procedures. If the patient has RHD please refer to the National "ARF RHD Guideline" Chapter 11, Table 11.5, page 223 and consult with the Cardiology team

14. Confirm routine vaccinations are up to date

- If not, then include in Kids GPS referral

15. **Refer all household members to their Primary Care Provider for throat swabs to assess for Group A Streptococcus carriage**
    - All household members with Group A Streptococcus found on throat swab should receive a 10 day course of oral phenoxymethylpenicillin, to be prescribed by the Primary Care Provider
16. **Review discharge plan with patient/family/carer**

5 Acute Review Clinic / Medical Day Unit

The first ambulatory review should involve a

- Medical assessment
- Review and education by a Kids GPS Care Coordinator
- Administration of the first secondary prophylaxis IM benzathine benzylpenicillin dose by nursing staff
- Aboriginal Health Worker, at the family's request

5.1 Medical Review

- The Medical Review should be undertaken by the Infectious Disease Fellow if available, otherwise by the Ambulatory Care Fellow, and should include:
    - Clinical Review
    - Review inflammatory markers if required (if not normalised by discharge)
    - Provide education to the family in collaboration with Kids GPS
    - Provide a prescription to the family for IM benzathine benzylpenicillin and lidocaine to fill and take to the first appointment with the Primary Care Provider
    - Verbal handover to the Primary Care Provider in collaboration with Kids GPS (as well as written referral letter provided to the Primary Care Provider by Kids GPS in collaboration with medical staff (template in appendix)
    - Remind the family that all household members need to be seen by the Primary Care Provider for throat swabs for Group A Streptococcus carriage
    - Ensure referrals have been made to Cardiology and Infectious Diseases for follow-up

5.2 Nursing Review

- ARC / MDU nursing staff to administer first secondary prophylaxis IM benzathine benzylpenicillin and lidocaine injection
    - Utilise the "Future Order" in Powerchart for IM benzathine benzylpenicillin, IM lidocaine and oral paracetamol

5.3 Kids GPS Review

- **Provide initial education to the family** (revisit if already completed prior to discharge)
  - Watch "Michael's Story" with the family and patient and address questions
    - https://www.rhdaustralia.org.au/individuals-families
  - Provide written information
    - https://www.health.nsw.gov.au/Infectious/factsheets/Factsheets/rheumatic-heart-disease.pdf
  - Introduce family to the Treatment Tracker App
    - https://www.rhdaustralia.org.au/treatment-tracker-app
- **Provide Care Coordination**
  - Primary Care Provider:
    - Identify whether the family have an established relationship with a Primary Care Provider (General Practitioner or Aboriginal Controlled Community Health Centre). A consistent Primary Care Provider is advisable for long term follow up and continuity of care. Kids GPS can help connect the family to a suitable local provider if a relationship is not already established
    - Check whether first appointment with the Primary Care Provider is booked to occur in 21 days for the next dose of IM benzathine benzylpenicillin to be given:
      - IF YES: provide Outreach and Handover to Primary Care Provider (see Part 6)
      - IF NOT: re-refer to ARC / MDU for second IM benzathine benzylpenicillin injection and book first appointment with the Primary Care Provider for the third IM benzathine benzylpenicillin injection
  - Specialities:
    - Confirm follow-up appointments for Cardiology and Infectious Diseases
  - Public Dental Clinic:
    - Assist the family to book an appointment for all children in the household
    - Child Dental Benefits Schedule Information: https://www.health.nsw.gov.au/oralhealth/Pages/child-dental-benefits-schedule.aspx
    - The family's local public dental clinic can be found here (search in *google chrome*): https://www.health.nsw.gov.au/oralhealth/Pages/call-centre-search.aspx
    - See note above regarding endocarditis prophylaxis (page 7)

- **Provide Resources to the Family**
  - Provide “Kids GPS Management Plan for ARF / RHD” in hard copy to the family
  - Email “ARF / RHD Education Email to Families” (template in appendix)

6 Kids GPS Outreach to Primary Care Provider

6.1 Role of Kids GPS

To provide handover to the Primary Care Provider in collaboration with medical staff.

- To provide a written handover to the Primary Care Provider including:
  - Email / Fax “ARF / RHD Handover to Primary Care Provider” (template in appendix)
  - Email / Fax “Kids GPS Management Plan for ARF / RHD”
- Aim to attend the first appointment with the patient / family and Primary Care Provider, either in person or virtually, if required
- Contact the Primary Care Provider by phone following every IM penicillin injection appointment to ensure attendance
- If there are concerns for adherence with IM penicillin prophylaxis / appointment attendance, the Kids GPS Care Coordinator should discuss this directly with the Ambulatory Care Fellow in the first instance. The Ambulatory Care Fellow will escalate these concerns as appropriate for the individual patient

6.2 Role of Primary Care Provider

- Undertake IM injections of benzathine benzylpenicillin and lidocaine every 21 days as per the “Kids GPS Management Plan for ARF / RHD”
- Undertake throat swabs for the entire household for Group A Streptococcus and prescribe oral phenoxymethylpenicillin if positive
- Ensure all children in the household have an annual dental review (Kids GPS Care Coordinator can assist in arranging this at the local public dental clinic)
- Provide a referral to the Infectious Diseases team & Cardiology team
- Ensure routine vaccinations are up-to-date
- Contact Kids GPS Care Coordinator if the patient does not attend every 21 days for routine penicillin injections. These patients will be discussed directly with the Ambulatory Care Fellow in the first instance (as above).

7 12 month SCHN Review & Long Term Follow-up

- Follow up with the Cardiology team for an echocardiogram and clinical review at 12 months post-diagnosis or as specified by the team
- Follow up with the Infections Diseases team at intervals specified by the team
- Kids GPS may be able to assist with appointment coordination where appropriate
- Final handover to the Primary Care Provider should be completed at the end of the initial 12 months of treatment and sub-specialty review. The Kids GPS Care Coordinator may choose to remain involved with patients on a case-by-case basis if there is particular medical or psychosocial complexity

8 References

1. RHD Australia (ARF/RHD writing group). The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3rd edition); 2020 https://www.rhdaustralia.org.au/arf-rhd-guideline
2. Acute Rheumatic Fever (ARF) and Rheumatic Heart Disease (RHD), NSW Government Health. https://www.health.nsw.gov.au/Infectious/rheumatic/Pages/default.aspx

9 Appendices

9.1 ARF / RHD Discharge Checklist

To complete and document in discharge summary

9.2 ARF / RHD Information Sheet

To print and provide to patient

9.3 ARF / RHD Notification Form

To complete and fax to local Public Health Unit (fax number above)

9.4 ARF / RHD Registration Form

To complete and fax to local Public Health Unit (fax number above)

9.5 NSW RHD Registration Information Sheet

To print and provide to patient

9.6 NSW RHD Registration Consent Forms

To complete, be signed by the patient, and fax to local Public Health Unit (fax number above)

9.7 Template: ARF / RHD Education Email to Families

Kids GPS Care Coordinator to email families following initial ARC / MDU Review

9.8 Template: ARF / RHD Handover to Primary Care Provider

Kids GPS Care Coordinator to fax / email to Primary Care Provider following initial ARC / MDU Review