Acute Rheumatic Fever

Case Definitions

Acute Rheumatic Fever (ARF) is an illness caused by an immunological reaction following infection with Group A Streptococcus, also known as *Streptococcus pyogenes*.

	High risk[1] groups	Low risk groups
Definite initial episode of ARF	2 major OR 1 major + 2 minor OR chorea[2] AND Evidence of preceding Group A Strep infection[3]	
Definite recurrence episode of ARF with documented previous history of ARF/Rheumatic Heart Disease (RHD)	2 major OR 1 major + 2 minor OR 3 minor OR chorea[2] AND Evidence of preceding Group A Strep infection[3] > 90 days from onset of previous episode ARF	
Probable (first episode of recurrence)	A clinical presentation that falls short by either one major or one minor manifestation OR Clinical evidence (as above) without laboratory suggestive evidence of a preceding Group A infection[2] AND where ARF is considered the most likely diagnosis	
Possible ARF (first episode or recurrence)	As for probable case, where the treating clinician has less confidence about ARF as the correct diagnosis, but other differential diagnoses have been excluded	
Major manifestations	**1. Carditis** Clinical and/or subclinical **2. Arthritis** Polyarthritis (definite history is sufficient) Aseptic monoarthritis Polyarthralgia **3. Chorea**[2] **4. Erythema marginatum** **5. Subcutaneous nodules**	**1. Carditis** Clinical and/or subclinical **2. Arthritis** Polyarthritis only **3. Chorea**[2] **4. Erythema marginatum** **5. Subcutaneous nodules**
Minor manifestations	**1. Monoarthralgia** (unless polyarthritis as a major manifestation already) **2. Fever ≥38°C** (can be based on reliable history alone) **3. ESR ≥30 mm/hr** and/or **CRP ≥30 mg/dl** **4. Prolonged PR** interval on ECG, after accounting for age variability, if no carditis present	**1. Polyarthralgia** or **aseptic monoarthritis** (unless polyarthritis as a major manifestation already) **2. Fever ≥38.5°C** **3. ESR ≥60 mm** and/or **CRP ≥30 mg/dl** **4. Prolonged PR** interval, after accounting for age variability, if no carditis present

[1] Kimberley Aboriginal population defined as high risk.

[2] Except in the case of rheumatic Sydenham's chorea, which may occur alone without other manifestations or laboratory suggestive evidence, provided other causes of chorea are ruled out. Therefore, Sydenham's chorea alone, without laboratory suggestive evidence, is sufficient evidence for a definite case, provided other causes of chorea are excluded.

[3] Evidence of preceding Group A Strep infection – elevated or rising Antistreptolysin-O or other streptococcal antibody, or a positive throat culture or rapid antigen or nucleic acid test for Group A Strep infection.

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Suggested upper limits of normal for Serum Streptococcal Antibodies		
	Upper limit of normal (U/mL)	
Age (in years)	ASOT	Anti-DMAse N
1-4	170	366
5-14	276	499
15-24	238	473
25-34	177	390
≥35	127	265

Upper limit of normal for PR-interval	
Age (in years)	Seconds
3-11	0.16
12-16	0.18
≥17	0.20

Early identification and treatment of skin and throat infections can prevent ARF/RHD. Those already receiving benzathine benzylpenicillin G (BPG) prophylaxis still need active treatment if the most recent dose was ≥7 days ago.

Refer Kimberley Clinical Guidelines: Sore Throat in Kids and Skin Infections in Children.

Accurate and timely diagnosis of ARF is critical to avoid progression to RHD:

- under-diagnosis of ARF may result in risk of recurrence
- over-diagnosis of ARF will result in a long period of unnecessary treatment

Principles of Management

Coordinated multidisciplinary team-based approach

It is strongly recommended that **ALL** children and preferably all adults with suspected ARF should be admitted to hospital for full review and work-up including echocardiography.

- **Refer all suspected cases of ARF to Kimberley Regional Physician Team/Regional Paediatrician** to guide investigation, admission, and involvement of paediatric or adult cardiology.
- General Practitioners are integral to ensuring early diagnosis and treatment of ARF to minimise heart valve damage and progression to RHD.
- Aboriginal Health Workers and Aboriginal Health Practitioners are a vital part of the team, providing support, education and assistance to patients, families and other health professionals.

Secondary prophylaxis should be initiated as soon as possible for all suspected cases of ARF

- Refer to Kimberley Clinical Guideline: RHD for further instructions about administration.
- Alternatively , refer to Administering BPG poster.

Notification

- **ARF is a notifiable disease** – all clinicians involved in the diagnosis and/or treatment of ARF are mandated to notify the WA RHD Register and Control Program (RHD Register) of every case diagnosis, new episode, information about patients being treated including secondary prophylaxis, any changes in patients' details.
- All diagnostic tests and medical specialist reports must be forwarded as per the mandatory reporting requirements.
- For notification of a new case – complete and forward a notification form to the RHD Register.
- Patients need to be advised of how their information will be used as per patient advice.
- Pregnancy should be notified to the RHD Register as early as possible.

Environmental Health

- Offer an environmental health referral to all patients with newly diagnosed ARF to minimise risk of future episodes.
- Contact your local service provider. Refer: referral form.

Further detail

For more details see below, Refer / Discuss

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Differential diagnosis to consider		
Arthritis	**Carditis**	**Chorea**
▪ Septic arthritis (including gonococcal infection) ▪ Connective tissue and other autoimmune disease ▪ Viral and reactive arthropathy ▪ Arbovirus ▪ Sickle cell anaemia ▪ Leukaemia or lymphoma ▪ Gout and pseudo-gout	▪ Innocent murmur ▪ Mitral valve prolapse ▪ Congenital heart disease ▪ Hypertrophic cardiomyopathy ▪ Myocarditis: viral or idiopathic ▪ Pericarditis: viral or idiopathic ▪ Kawasaki's disease	▪ Systemic lupus erythematosus ▪ Drug intoxication ▪ Wilson's disease ▪ Tic disorder ▪ Choreoathetoid cerebral palsy ▪ Encephalitis ▪ Familial chorea (including Huntington's) ▪ Intracranial tumour ▪ Hormonal (including pregnancy and OCP) ▪ Metabolic disorder ▪ Vasculitis ▪ Sarcoidosis
▪ Infective endocarditis		

Investigations to consider		
Arthritis	**Carditis**	**Chorea**
▪ Joint aspirate for MC&S/ crystals ▪ Joint Xray ▪ STI screen (syphilis/HIV serology, chlamydia/ gonorrhoea MCS and PCRs, consider throat swab) ▪ Autoimmune antibodies testing (ANA, ENA, RF, anti-CCP, dsDNA, C3/C4, Urine ACR) ▪ Mycoplasma serology	▪ Respiratory virus testing ▪ HIV serology	▪ TFT ▪ PTH ▪ BHCG ▪ Copper level ▪ Ceruloplasmin ▪ Autoimmune and vasculitis screen (ANA, ENA, RF, anti-CCP, dsDNA, C3/C4, ANCA, Urine ACR + microscopy) ▪ Neuroimaging and CSF analysis can be considered if diagnosis unclear
▪ Blood cultures ▪ Arbovirus testing ▪ CMV/EBV serology ▪ Viral hepatitis serology A, B, C		

Therapeutic Protocols

Investigations:

- Microbiology – swab throat and any infected skin sores; blood cultures if temperature ≥ 38°C. Ideally BEFORE administering antibiotics.
- Bloods – FBE, ESR, CRP, ASOT, anti-DNase B, UEC (baseline renal function).
- ECG
- Joint effusions – if significant, aspirate for MC&S, cell count and differential and crystals. In paediatric population refer to orthopaedic team.
- Echo, as soon as possible and preferably within a month of diagnosis. Discuss with Kimberley Regional Physician Team/Paediatrician/Cardiologist if strong suspicion for carditis. If initial echo unremarkable, repeat within four to six weeks or if new findings on examination.
- Investigations as above for differentials as appropriate.

Management:

1. Give BPG IMI stat
   Dose:
   - Adults and children ≥20kg: LA-Bicillin 1.2 million units
   - Children <20kg: LA-Bicillin 600,000 units

   If hypersensitivity to penicillin discuss with physician/paediatrician

2. Arthritis/arthralgia:
   First line:
   - Oral paracetamol (+/- codeine OR tramadol if >12 years old)
   - Adults and children ≥45kg: 500mg-1,000mg QID
   - Children <45kg: 15mg/kg every 4-6 hours. (Max 60mg/kg (up to 4g)/day QID)

Avoid using NSAIDs if diagnosis is unclear as it may mask symptoms hence make ARF diagnosis more difficult.

Once ARF diagnosis is confirmed:

- Naproxen 250-500mg (child 5-10mg/kg/day) orally twice daily, max 1,250mg; OR
- Ibuprofen 20-400mg (child 5-10mg/kg) orally three times daily, max 2400mg; OR
- Aspirin adults and children 50-60mg/kg/day orally in 4-5 divided doses, can be escalated to 80-100mg/kg/day in 4-5 divided doses.

(In children cease during intercurrent viral illness due to possibility of Reye's Syndrome)

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Many patients need anti-inflammatory therapy for only one to two weeks (i.e., anti-inflammatory therapy can be stopped at two weeks if the patient is pain-free with improved inflammatory markers). Some may have "rebound phenomenon" and require longer course up to four to six weeks, however, dose can be reduced after initial one to two weeks. Discuss with local paediatrician/physician if any concerns.

3. Chorea
    - Discuss treatment with Kimberley Regional Physician Team/Paediatrician.
4. Carditis
    - All patients with suspected carditis require admission.
    - Discuss with Kimberley Regional Physician Team/ Paediatrician/Cardiologist

Primary Care Follow Up

Follow up should be organised within one week post initial diagnosis/consultation if managed as an outpatient OR one week post discharge from hospital if initially managed as inpatient.

At the follow up appointment:

- Confirm ARF diagnosis, detailing major and minor manifestations. Each case should be assigned as definite/probable/possible (unless already done).
- Assign a care plan according to priority group. Refer to Kimberley Clinical Guideline: RHD for information on priority allocation. This will determine duration of secondary prophylaxis and follow up and required including frequency.
- Repeat full examination, noting any new findings/developing carditis.
- Repeat ECG each visit (until normalised).
- Repeat ESR/CRP once or twice weekly (until normal result for at least one month).
- Repeat ASOT and anti-DNase B 10-14 days after initial titres (if initial titres inconclusive).
- Organise repeat echo in four to sic, if normal – repeat in one year, if abnormal -discuss with cardiologist/physician/paediatrician.
- Ensure RHD Register has been notified.
- Arrange dental review and ongoing dental care to reduce risk of endocarditis.
- Ensure follow up within three months with physician/cardiologist/paediatrician has been arranged (preferably after echo has been done).
- Note last date of LAB and organise ongoing administration every three to four weeks. Refer to Kimberley Clinical Guideline: RHD for more detail.
- Ensure all immunisations are up to date, including pneumococcal and annual influenza.

Refer/Discuss

Aboriginal Environmental Health

Referral form
Find your local service via the referral form.
OR, contact Kimberley Population Health Unit (KPHU):
KPHU.envhealth@health.wa.gov.au

WA RHD Register and Control Program

Phone: 1300 622 745
Fax: (08) 6553 0899
RHDRegister@health.wa.gov.au

Kimberley Regional Physician Team

Available for local advice:
Phone: (08) 9194 2222
krpt@health.wa.gov.au
MMEx: Kimberley Regional Physician Team
Outpatient referrals via e-Referrals or local specialist clerk

Kimberley Regional Paediatric Team

Phone: (08) 9194 2222
On call: 0427 988 570
kimberley.paediatrics@health.wa.gov.au
Outpatient referrals via e-Referrals or local specialist clerk

Kimberley Regional Obstetric Team

Phone: (08) 9194 2222
krog@health.wa.gov.au
Outpatient referrals via e-Referrals or local specialist clerk

Visiting Cardiology Service, Perth Cardiovascular Institute

Adults only. Liaise with regional paediatric team for children
Phone: (08) 6314 6833
Kimberley@perthcardio.com.au
Outpatient referrals via e-Referrals or local specialist clerk

Resources

- Kimberley Clinical Guidelines (KAHPF)
- National Aboriginal Community Controlled Health Organisation (NACCHO)
- National Guidelines and ARF and RHD Guidelines app (RHD Australia)

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