



3. Management of acute rheumatic fever

All patients with suspected ARF (first episode or recurrence) should be hospitalised as soon as possible after the onset of symptoms. This ensures that all investigations are performed, and if necessary, the patient should be observed to confirm the diagnosis before commencing treatment.

This quick reference guide is derived from the *Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease* (2nd edn).

What is acute rheumatic fever?

Acute rheumatic fever (ARF) is an illness caused by a reaction to a bacterial infection with group A streptococcus. It causes an acute, generalised inflammatory response and an illness that targets specific parts of the body, including the heart, joints, brain and skin. Individuals with ARF are often unwell, have significant joint pain and require hospitalisation. Despite the dramatic nature of the acute episode, ARF leaves no lasting damage to the brain, joints or skin, but can cause persisting heart damage, termed 'rheumatic heart disease' (RHD). Recurrences of ARF may cause further cardiac valve damage. Hence, RHD steadily worsens in people who have multiple episodes of ARF.

Who gets ARF?

Although ARF is relatively rare in industrialised countries, in Australia it is a significant illness among Aboriginal people and Torres Strait

Islanders, particularly across northern and central Australia. Pacific Islanders, and migrants from countries with a high prevalence of RHD, are also known to be at high risk.

Problems with diagnosis and management

Several factors contribute to the barriers in diagnosis and management of ARF and RHD in Australia:

- although strategies for preventing RHD have been proven to be simple, cheap and cost-effective, they must be adequately implemented in populations at highest risk of the disease
- because ARF is rare in most metropolitan centres, the majority of clinicians will have seen very few, if any, cases of ARF
- there is variability in the management of these diseases, with minimal training and experience in the management of ARF and RHD, occasionally resulting in inappropriate management
- access to healthcare services by population groups experiencing the highest rates of ARF and RHD is often limited.

Confirming the diagnosis

Diagnostic criteria, recommended investigations and detailed information on differential diagnoses are given in the quick reference guide *Diagnosis of acute rheumatic fever*.

As the arthritis, arthralgia and fever of ARF respond to non-steroidal anti-inflammatory drugs (NSAIDs), which may prevent the full clinical manifestations becoming apparent, it is recommended that joint pain be treated with paracetamol or codeine until the diagnosis is confirmed.

There is convincing evidence that subclinical or silent rheumatic valve damage detected by echocardiography is part of the spectrum of rheumatic carditis, and should not be ignored.

Guidelines for general in-hospital care

All patients with suspected ARF (first episode or recurrence) should be discussed immediately with a paediatrician or adult physician expert in the diagnosis and management of ARF, admitted to hospital as soon as possible after the onset of symptoms, and the steps initiated to confirm the diagnosis.

While in hospital, the patient should be registered in centralised and local ARF/RHD registers.

Occasionally, when the diagnosis has already been confirmed and the patient is not unwell (e.g. mild recurrent chorea in a child with no other symptoms or signs), outpatient management may be appropriate, but only after consultation with a specialist.

Treatment

All cases

Single-dose im benzathine penicillin G (preferable) or 10 days of oral penicillin V (iv not needed; oral erythromycin if allergic to penicillin)

Arthritis and fever

Paracetamol (first line) or codeine until diagnosis confirmed

Aspirin, naproxen or ibuprofen, once diagnosis confirmed, if arthritis or severe arthralgia present

Mild arthralgia and fever may respond to paracetamol alone

Influenza vaccine for children receiving aspirin during the influenza season (autumn/winter)

Chorea

No treatment for most cases

Carbamazepine or valproic acid, if treatment necessary

Carditis/heart failure

Bed rest, with mobilisation as symptoms permit

Urgent echocardiogram

Antifailure medication:

- Diuretics/fluid restriction for heart failure
- ACE inhibitors for more severe failure, particularly if aortic regurgitation is present; glucocorticoids optional for severe carditis (in central and northern Australia, prophylaxis for opportunistic infections may be needed for those on prolonged steroid courses. Seek specialist advice)
- Digoxin, beta-blocker or electrical cardioversion if atrial fibrillation present

Valve surgery for life-threatening acute carditis (rare)

Nursing recordings

Temperature, pulse, respiratory rate, blood pressure 4 times daily

Sleeping pulse (e.g. 0200 hours)

If pulse >100, apical heart rate

Diet

Free fluids (if no heart failure)

Normal diet (limit extras)

Early diet advice, if overweight and in failure, to avoid further weight gain

Weekly weight

Bed rest and general care

Strict bed rest not necessary for most patients

Plan care to provide rest periods

Provide age-appropriate activities

Notify school teacher

Involve family in care

Prepare for discharge to primary care facility, and follow up

If clinical carditis present (heart murmur, heart failure, pericardial effusion, valvular damage)

Document cardiac symptoms and signs

Daily weight and fluid balance chart

Diuretics, ACE inhibitors, digoxin, if indicated; consider glucocorticoids

Anticoagulation if atrial fibrillation present

Cardiology opinion

ACE, angiotensin-converting enzyme; im, intramuscular; iv, intravenous.

Medication used in ARF

Medication	Indication	Regimen	Duration
BPG, im	Treat streptococcal infection	900 mg (1,200,000 U) \geq 20 kg 450 mg (600,000 U) <20 kg	Single dose
or Penicillin V, po	Initial treatment of streptococcal infection	Child: 250 mg, bd Adolescent and Adult: 500 mg, bd	10 days
or Erythromycin ethyl succinate, po (only if allergic to penicillin)	Initial treatment of streptococcal infection	Child: 20 mg/kg up to 800 mg, bd Adult: 800 mg, bd	10 days
or Erythromycin, po	Initial treatment of streptococcal infection	Child: 12.5 mg/kg up to 500 mg, bd Adult: 500 mg, bd	10 days
Paracetamol, po	Arthritis or arthralgia: mild or until diagnosis confirmed	60 mg/kg/day (max 4 g) given in 4–6 doses/day; may increase to 90 mg/kg/day, if needed, under medical supervision	Until symptoms relieved or NSAID started
Codeine, po	Arthritis or until diagnosis Confirmed arthralgia	0.5–1 mg/kg/dose (adults 15–60 mg/dose) 4–6 hourly	Until symptoms relieved or NSAID started
Aspirin, po	Arthritis or severe arthralgia (when ARF diagnosis confirmed)	Begin with 50–60 mg/kg/day, increasing, if needed, up to 80–100 mg/kg/day (4–8 g/day in adults) given in 4–5 doses/day If higher doses required, reduce to 50–60 mg/kg/day when symptoms improve, and cease when symptom free for 1–2 weeks Consider ceasing in the presence of acute viral illness, and consider influenza vaccine if administered during autumn/winter	Until joint symptoms relieved
Naproxen, po	Arthritis or severe arthralgia (when ARF diagnosis confirmed)	10–20 mg/kg/day (max 1250 mg) given, bd	As for aspirin
Ibuprofen, po	Arthritis or severe arthralgia (when ARF diagnosis confirmed)	30 mg/kg/day (max 1600 mg) given tds	As for aspirin
Prednisone or prednisolone, po.	Severe carditis, heart failure, pericarditis with effusion	1–2 mg/kg/day (max 80 mg); if used >1 week, taper by 20–25% per week	Usually 1–3 weeks

Medication	Indication	Regimen	Duration
Frusemide, po/iv (can also be given im)	Heart failure	Child: 1–2 mg/kg stat, then 0.5–1 mg/kg/dose 6–24 hourly (max 6 mg/kg/day) Adult: 20–40 mg/dose, 6–24 hourly, up to 250–500 mg/day	Until failure controlled and carditis improved
Spironolactone, po	Heart failure	1–3 mg/kg/day (max 100–200 mg/day) in 1–3 doses; round dose to multiple of 6.25 mg (1/4 of a tablet)	As for frusemide
Enalapril, po	Heart failure	Child: 0.1 mg/kg/day in 1–2 doses, increased gradually over 2 weeks to a max of 1 mg/kg/day in 1–2 doses Adult: initial dose 2.5 mg daily; maintenance dose 10–20 mg daily (max 40 mg)	As for frusemide
Captopril, po	Heart failure	Child: initial dose 0.1 mg/kg/dose. Beware of hypotension. Increase gradually over 2 weeks to 0.5–1 mg/kg/doses 8 hourly (max 2 mg/kg/dose 8 hourly). Adult: initial dose 2.5–5 mg. Maintenance dose 25–50 mg 8 hourly	As for frusemide
Lisinopril, po	Heart failure	Child: 0.1–0.2 mg/kg once daily, up to 1 mg/kg/dose Adult: 2.5–20 mg once daily (max 40 mg/day)	As for frusemide
Digoxin, po/iv	Heart failure/ atrial fibrillation	Child: 15 mcg/kg start, and then 5 mcg/kg after 6 hours, then 3–5 mcg/kg/dose (max 125 mcg) 12 hourly Adult: 125–250 mcg daily Check serum levels	Seek advice from specialist
Carbamazepine	Severe chorea	7–20 mg/kg/day (7–10 mg/kg day usually sufficient) given tds	Until chorea controlled for several weeks, then trial off medication
Valproic acid, po	Severe chorea (may affect salicylate metabolism)	Usually 15–20 mg/kg/day (can increase to 30 mg/kg/day) given tds	As for carbamazepine

bd, *bis die* (twice daily); BPG, benzathine penicillin G; im, intramuscular; iv, intravenous; NSAID, non-steroidal anti-inflammatory drug; po, per oral; tds, *ter die sumendum* (three times daily).

Commencing long-term preventive measures

Secondary prevention

As outlined in the previous Table, penicillin is given in cases of ARF to ensure eradication of streptococci that may persist in the upper respiratory tract. As this could be considered the commencement of secondary prophylaxis, it may be advisable to use im benzathine penicillin G.

Some clinicians prefer to use oral penicillin while patients are in hospital, and to defer the im injection until there has been improvement, and patients and their families have been properly counselled about secondary prophylaxis.

Patients with a reliably-documented penicillin allergy may be treated with oral erythromycin. However, most patients labelled as being allergic to penicillin are not. It is recommended that patients with a stated penicillin allergy be investigated carefully, preferably with the help of an allergist, before being accepted as truly allergic.

Patients with probable ARF may be managed in two ways, according to the level of confidence with which the diagnosis is made:

- highly-suspected ARF: manage as for definite ARF
- uncertain ARF: in patients from high-risk groups, administer 12 months of secondary prophylaxis initially, and reassess (including echocardiography) at that time. If no evidence of recurrent ARF, and no evidence of cardiac valvular damage on echocardiography, consider ceasing secondary prophylaxis. In such cases, the residual uncertainty should be discussed with the patient, and they should be encouraged to be particularly vigilant about the treatment of sore throats, prevention and treatment of skin sores and early presentation with any symptoms of potentially recurrent ARF.

Secondary prevention is discussed in greater detail in the quick reference guide *Secondary prevention of acute rheumatic fever*.

Advice on discharge

- All patients should have a good understanding of the cause of ARF and the need to have sore throats and skin sores treated early. Family members should be informed that they are at increased risk of ARF compared to the wider community.
- Patients and families should understand the reason for long-term secondary prophylaxis, and the consequences of not receiving all recommended treatment. They should be given clear information about where to go for secondary prophylaxis, and written information on appointments for follow up with their local medical practitioner, physician/paediatrician and cardiologist (if needed).
- If there is cardiac valve damage, patients and families should be reminded of the importance of antibiotic prophylaxis for dental and other procedures to protect against endocarditis.

Contact local health staff for follow up

- The notifying medical practitioner should make direct contact with community medical staff, so that they are aware of the diagnosis, the need for secondary prophylaxis and any other specific follow up requirements.



RHDAustralia is an initiative of the Menzies School of Health Research, in partnership with James Cook University and Baker IDI. Funded by the Australian Government Department of Health and Ageing.

The Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition)

Quick reference guides include:

- Primary prevention of ARF
- Diagnosis of ARF
- Management of ARF
- Secondary prevention of ARF
- Management of RHD
- RHD in pregnancy
- RHD control programs

RHDAustralia

Ph: 08 8922 8196

Email: info@rhdaustralia.org.au

www.rhdaustralia.org.au