## Acute Rheumatic Fever

## *Executive summary*

## Introduction

# Acute rheumatic fever (ARF) is an auto-immune consequence of infection with the bacterium group A streptococcus (GAS). The symptoms usually appear 2-4 weeks following infection. It causes an acute generalised inflammatory response and an illness that affects only certain parts of the body, mainly the heart, joints, brain and skin.

# ARF leaves no lasting damage to the brain, joints or skin. However, the damage to the heart, or more specifically the mitral and/or aortic valves, may remain once the acute episode has resolved and these long term changes are termed rheumatic heart disease (RHD). Because of its high prevalence in developing countries, RHD is the most common form of paediatric heart disease globally. In many countries, it is the most common cause of cardiac mortality in children and adults aged less than 40 years.

### Target users

* Doctors
* Nurses

### Target area of use

* Gate clinic
* OPD
* Ward

### Key areas of focus / New additions / Changes

## This guideline is intended to support clinicians in the diagnosis and management of acute rheumatic fever in adults and children who present to CSD.

## Limitations

We do not currently have access to streptococcal antibody tests (Anti-DNAse B and Anti-Streptolysin Titre) which are highly useful investigations in acute rheumatic fever.

There is limited use of intramuscular Benzathine Penicillin G due to historical cases of severe adverse drug reaction. Throat culture is not routinely available at Keneba.

## Presenting symptoms and signs

WHO criteria for the diagnosis of rheumatic fever are based on the revised Jones criteria:

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| **Diagnostic category** | **Criteria** |
| Primary episode of ARF | 2 major OR 1 major and 2 minor manifestations **PLUS**  Evidence of a preceding GAS infection |
| Recurrent attack of ARF in a patient **without** established rheumatic heart disease | 2 major OR 1 major and 2 minor manifestations **PLUS**  Evidence of a preceding GAS infection |
| Recurrent attack of ARF in a patient **with** established rheumatic heart disease | 2 minor manifestations **PLUS**  Evidence of a preceding GAS infection |

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| **MAJOR MANIFESTATIONS** | **POINTS FOR DIAGNOSIS** |
| Arthritis | Most common presenting feature of ARF  Polyarthritis is usually asymmetrical and migratory  Monoarthritis may also be a presenting feature  Large joints are usually affected, especially the knees and ankles  Usually responds within 3 days of starting NSAID therapy |
| Chorea | Consists of jerky, uncoordinated movements especially affecting the hands, feet, tongue and face  Disappears during sleep |
| Carditis | Usually presents as a pansystolic murmur over the apex or an early diastolic murmur at the left sternal edge |
| Subcutaneous nodules | Present as crops of small, round, painless nodules over the elbows, wrists, knees, occiput and spinal processes |
| Erythema marginatum | Occurs as circular patterns of bright pink macules or papules on the trunk and proximal extremities |
| **MINOR MANIFESTATIONS** | **POINTS FOR DIAGNOSIS** |
| Arthralgia | Migratory, asymmetrical and affecting large joints |
| Fever | Most manifestations of ARF are accompanied by fever  A fever of ≥ 38 degrees meets this criteria |
| Elevated acute phase reactants | ESR (or CRP) ≥ 30 meets this criteria |

## Differential diagnosis

The following table outlines the features that can help differentiate between pharyngitis caused by GAS from that caused by a viral illness.

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| **GAS infection** | **Viral infection** |
| * Sudden onset of sore throat * Fever * Headache * Nausea, vomiting, abdominal pain * Tonsillopharyngeal inflammation * Tonsillopharyngeal exudates * Palatal petechiae * Enlarged anterior cervical lymph nodes * Scarlatiniform rash | * Conjunctivitis * Coryza * Cough * Diarrhoea * Hoarseness * Ulcerative stomatitis * Viral exanthema |

## Investigations

Patients should have the following:

* Bloods: FBC, ESR, blood culture
* Throat swab
* ECG
* Echocardiogram
* CXR (if carditis suspected)

Evidence of preceding GAS infection includes:

* Elevated P-R interval on ECG: > 0.16s (3-12 years), > 0.18s (13-16 years), >20s (≥ 17 years)
* Elevated streptococcal antibodies
* Positive throat culture
* Recent scarlet fever

## Management

### Primary prevention of ARF

# All patients presenting with a sore throat should have their throat examined.

# All children aged 4-15 years who present with a fever should have their throat examined (unless clear alternative source of their fever).

# Any patient who has features suggestive of GAS and all children aged 4-15 years who have evidence of pharyngeal inflammation/exudate should be treated with antibiotics for suspected GAS infection.

# A throat swab should be sent for culture in all patients who are being treated with antibiotics for suspected GAS infection.

### Antibiotic therapy for GAS infection

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| For patients without penicillin allergy | | | |
| Penicillin V  (Phenoxymethylpenicillin) | Oral | 7.5 mg/kg (max dose 500 mg) QDS | 10 days |
| Amoxicillin | Oral | 50 mg/kg (max dose 1000 mg) OD | 10 days |
| Benzathine penicillin G | Intramuscular | Weight < 30 kg: 450 mg (600,000 units)  Weight ≥ 30 kg: 900 mg (1,200,000 units) | One dose |
| **For patients with penicillin allergy** | | | |
| Erythromycin | Oral | 20 mg/kg (max 500 mg) BD | 10 days |

### Treatment of ARF

* **Antibiotics:** All patients diagnosed with ARF should receive a course of antibiotics to treat GAS infection (see above on antibiotics).
* **Analgesia/anti-inflammatories**: Paracetamol may be sufficient for fever and mild arthralgia. Ibuprofen (10 mg/kg, maximum 500 mg, TDS) may be given for arthralgia or Aspirin (50-60 mg/kg/day, increased if needed to 80-100 mg/kg/day, given in 4-5 divided doses). Ibuprofen and aspirin may be used for more severe arthritis and pain but should only be started after a diagnosis of ARF has been made as they may mask the manifestations used to make the diagnosis.
* **Chorea:** Can use carbamazepine (2-3 mg/kg TDS) for severe symptoms.
* **Carditis/heart failure:** Diuretics and fluid restriction, use of ACE inhibitor and Digoxin if atrial fibrillation present.

### Secondary prevention of ARF

Patients who develop recurrent episodes of ARF are at increased risk of worsening rheumatic heart disease. Therefore antibiotic prophylaxis is used in all patients who have had an episode of ARF to minimise the risk of recurrent episodes of GAS infection.

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| **For patients without penicillin allergy** | |
| Oral Penicillin V  (Phenoxymethylpenicillin) | 7.5 mg/kg (max dose 500 mg) BD |
| **For patients with penicillin allergy** | |
| Oral Erythromycin | 20 mg/kg (max dose 500 mg) BD |

## References

New Zealand Guidelines for Rheumatic Fever- Diagnosis, Management and Secondary Prevention of Acute Rheumatic Fever and Rheumatic Heart Disease: 2014 Update

The Australian Guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (second edition)

AHA Guidelines on Prevention of Rheumatic Fever and Diagnosis and Treatment of Acute Streptococcal Pharyngitis

NICE guidelines: Sore throat (acute): antimicrobial prescribing, January 2018

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