RHD Australia

RED FLAG TOOL
FOR RECOGNITION OF ACUTE RHEUMATIC FEVER (ARF)

Red flag 1:

ARF is:

- An illness caused by an autoimmune response to a bacterial infection with group A streptococcus (GAS infection)
- ARF causes an acute inflammatory response that affects the heart, joints, brain and skin. This inflammatory response leaves no lasting damage to the joints, brain and skin, however the damage to the heart (specifically the mitral and aortic valves) may remain once the episode of ARF has resolved.

This is known as rheumatic heart disease (RHD)

Red flag 2:

ARF risk groups are:

- 5–14 years old – GAS pharyngitis is common in this age group (However, ARF has also been seen in patients into their mid forties and should not be ruled out)
- Aboriginal people and Torres Strait Islanders living in rural or remote settings are known to be at high risk
- Other Aboriginal people and Torres Strait Islanders living in urban settings, Maoris and Pacific Islanders, and potentially immigrants from developing countries, may also be at high risk

Red flag 3:

ARF diagnosis:

- Is guided by the 2012 Australian criteria for the diagnosis of acute rheumatic fever
- Clinical features are divided into MAJOR and MINOR manifestations
- MAJOR manifestations are clinical features that make the diagnosis more likely
- MINOR manifestations are considered to be suggestive, but not sufficient on their own for a definite diagnosis of acute rheumatic fever.

Red flag 7:

- ARF is a notifiable condition in WA, NT and QLD. RHD is not currently notifiable anywhere in Australia.
- Whilst in hospital, the patient should be registered in centralised and local ARF/RHD registers. Please contact the Rheumatic Heart Disease Control Program in your state/territory.

For comprehensive information on ARF and RHD please refer to:

- RHDAustralia (ARF/RHD writing group), National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand. Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition). 2012
- RHDAustralia (ARF/RHD writing group), National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand. Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition) Quick reference guides. 2012

Available from: RHDAustralia

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Red flag 4:

MAJOR and MINOR manifestations of Acute Rheumatic Fever

MAJOR manifestations for high risk groups

Carditis (including subclinical evidence of rheumatic valvulitis on echocardiogram)

Points for diagnosis:

- May be asymptomatic
- May present as a murmur, chest pain (rare)
- Most common form: valvulitis (aortic and/or mitral valve)
- Severe carditis may present as heart failure

Polyarthritis or aseptic monoarthritis or polyarthralgia

Points for diagnosis:

- Most common presenting ARF symptom
- Can occur in one joint or multiple joints
- Asymmetrical, migratory
- Affects large joints: knees, ankles Less common: shoulders, hips
- Most common in children/adolescents (can present in adults)
- Responds to NSAIDs within 3 days

**ARF should always be considered as a differential diagnosis for patients presenting with arthritis in high risk populations*

Chorea

Points for diagnosis:

- Jerky, uncoordinated movements: hands, feet, tongue, face
- Other signs: milkmaids grip, spooning, pronator sign
- Movements disappear during sleep
- May affect only one side (hemichorea)
- Predominantly affects adolescent females
- Common presentation in Aboriginal and Torres Strait Islander people that present with ARF

Erythema Marginatum

Points for diagnosis:

- Extremely rare
- Presentation is highly specific for ARF
- Rash: bright pink macules or papules
- Often mistaken for ringworm
- Occurs mainly on trunk, extremities (never on face)
- Rash may persist/recur for weeks/ months after an episode of ARF

Subcutaneous nodules

Points for diagnosis:

- Firm, round, painless nodules under the skin which move freely
- ½–2 cm in size
- Appears on bony joints: elbow, wrist, knees, ankles
- Extremely rare
- Strongly associated with carditis

MINOR manifestations for high risk groups

Monoarthralgia

Points for diagnosis:

- Pain in joint movement
- No evidence of heat or swelling in the joint

Fever

Points for diagnosis:

- History or presentation with fever ≥ 38° Celsius

Raised ESR/CRP

Points for diagnosis:

- ESR ≥ 30 mm/h or
- CRP ≥ 30 mg/L

ECG changes

Points for diagnosis:

- Prolonged PR interval and other rhythm abnormalities

Red flag 5:

Using MAJOR and MINOR manifestations to make an accurate diagnosis of ARF

	For high risk groups and all other groups
Definite initial episode of ARF	**2 MAJOR** or **1 MAJOR** and 2 MINOR + evidence of a preceding **GAS infection**
Definite recurrent episode of ARF in a patient with known past ARF or RHD	**2 MAJOR** or **1 MAJOR** and 1 MINOR or 3 MINOR manifestations + evidence of a preceding **GAS infection**
Probable ARF (first episode or recurrence)	A clinical presentation that falls short by either one MAJOR or one MINOR manifestation, or the absence of streptococcal serology results, but one in which ARF is considered the most likely diagnosis. Such cases should be further categorised according to the level of confidence with which the diagnosis is made: • Highly-suspected ARF • Uncertain ARF

Red flag 6:

Treatment of suspected cases of ARF

Investigations in suspected ARF:

- All patients with suspected or confirmed ARF should undergo echocardiography to confirm or refute the diagnosis of rheumatic carditis
- **The following investigations are recommended for all cases**
  - White blood cell count
  - Erythrocyte sedimentation rate (ESR)
  - C Reactive Protein (CRP)
  - Blood cultures, if febrile
  - Electrocardiogram (if prolonged P-R interval or other rhythm abnormality, repeat in 2 weeks and again at 2 months, if still abnormal)
  - Chest X-ray, if clinical or echocardiographic evidence of carditis
  - Echocardiogram (consider repeating after 1 month, if negative)
  - Throat swab (preferably before giving antibiotics); culture for group A streptococcus
  - Antistreptococcal serology: both ASO and anti-DNase B titres, (if available) repeat 10-14 days later if first test not confirmatory
- **Tests for alternative diagnoses, depending on clinical features**
  - Repeated blood cultures, if possible endocarditis
  - Joint aspirate (microscopy and culture) for possible septic arthritis
  - Copper, ceruloplasmin, antinuclear antibody, drug screen for choreiform movements
  - Serology and autoimmune markers for arboviral, autoimmune or reactive arthritis