



ပြည်ထောင်စုမြန်မာနိုင်ငံ၊ အထူးဒေသ(၁) ကျန်းမာရေးဌာန
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Rheumatic Fever

Standard Operating Procedure (SOP)



Union of Myanmar Special region - 1
Pediatric (SOP)

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Union of Myanmar Special region - 1
Pediatric (SOP)

Rheumatic Fever

- An inflammatory reaction in joints, skin, heart and CNS following Group A haemolytic streptococcal infection
- It is mostly seen in children aged 5-15 years and the incidence is highest in those from socially and economically disadvantaged areas

Diagnosis

Table 3.14 Revised Jones Criteria (Source: Gewitz et al, 2015)

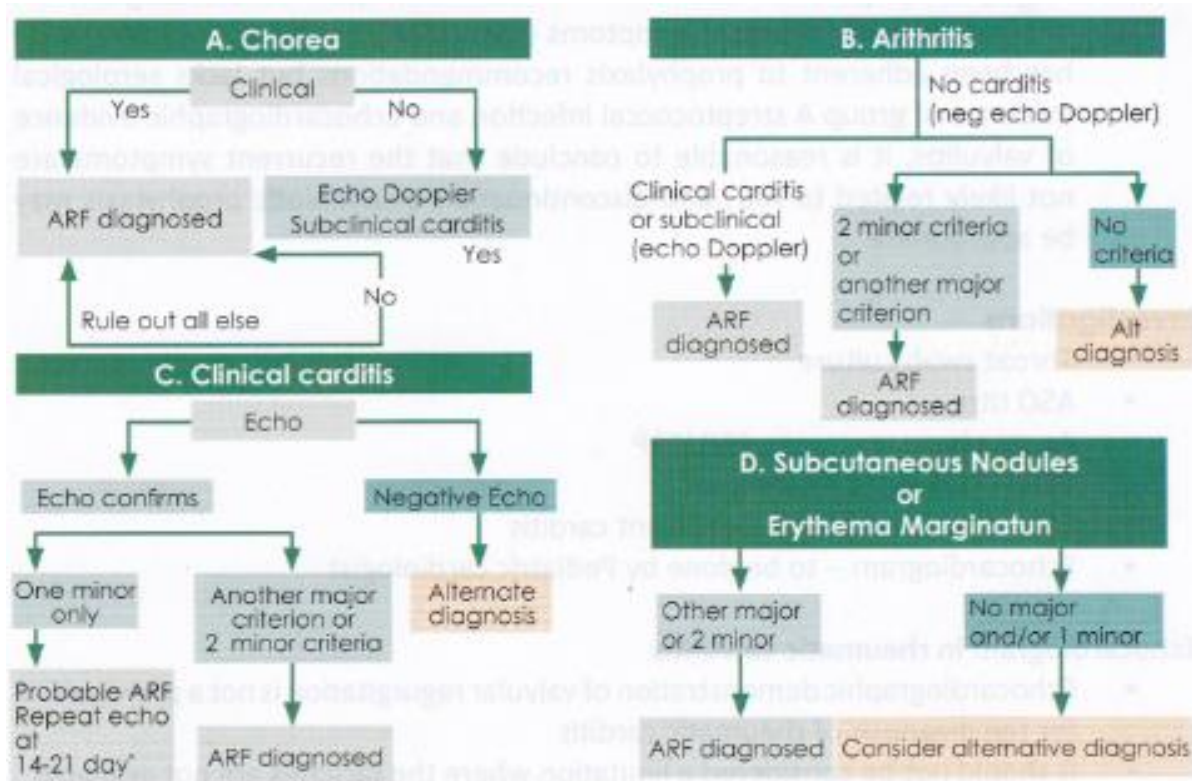
A. For all patient populations with evidence of preceding GAS infection	
Diagnosis: Initial ARF	2 Major manifestations or 1 major plus 2 minor manifestations
Diagnosis: Recurrent ARF	2 Major or 1 major and 2 minor or 3 minor
B. Major criteria	
<i>Low risk populations*</i> • Carditis ○ Clinical and/or subclinical • Arthritis ○ Polyarthritides only • Chorea • Erythema marginatum • Subcutaneous nodules	<i>Moderate and high risk populations</i> • Carditis ○ Clinical and/or subclinical • Arthritis ○ Monoarthritic or Polyarthritides ○ Polyarthralgia • Chorea • Erythema marginatum • Subcutaneous nodules
C. Minor criteria	
<i>Low risk populations*</i> • Polyarthralgia • Fever ($\geq 38.5^{\circ}\text{C}$) • ESR ≥ 20 mm in the 1st hour and/or • CRP ≥ 3.0 mg/dL • Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion)	<i>Moderate and high risk populations</i> • Monoarthralgia • Fever ($\geq 38.5^{\circ}\text{C}$) • ESR ≥ 20 mm in the 1st hour and/or CRP ≥ 3.0 mg/dL • Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion)

ARF indicates acute rheumatic fever, CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; and GAS, group A streptococcal infection.

*Low-risk populations are those with ARF incidence ≤ 2 per 100 000 school-aged children or all-age rheumatic heart disease prevalence of ≤ 1 per 1000 population per year

+Subclinical carditis indicates echocardiographic valvulitis.

±Polyarthralgia which should only be considered as a major manifestation in moderate to high-risk populations after exclusion of other causes, As in past versions of the criteria, erythema marginatum and subcutaneous nodules are rarely "stand-alone" major criteria. Additionally, joint manifestations can only be considered in either the major or minor categories but not both in the same patient



B, C and D required evidence for GAS infection

Figure 4.5. Diagnosis strategy for acute rheumatic fever

**Subclinical carditis can be considered. Alt indicates alternative; ARF, acute rheumatic fever; echo, echocardiography; GAS, group A streptococcal; and neg, negative*

Rheumatic Fever Recurrences

- With a reliable past history of ARF or established RHD, and in the face of documented group A streptococcal infection, 2 major or 1 major and 2 minor or 3 minor manifestations may be sufficient for a presumptive diagnosis
- When minor manifestations alone are present, the exclusion of other more likely causes of the clinical presentation is recommended before a diagnosis of an ARF recurrence is made

Possible Rheumatic Fever

1. *Probable ARF is defined as 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.*
2. *Where there is genuine uncertainty, it is reasonable to consider offering 12months of secondary prophylaxis followed by reevaluation to include a careful history and physical examination in addition to a repeat echocardiogram*
3. *In a patient with recurrent symptoms (particularly involving the joints) who has been adherent to prophylaxis recommendations but lacks serological evidence of group A streptococcal infection and echocardiographic evidence of valvulitis, it is reasonable to conclude that the recurrent symptoms are not likely related to ARF, and discontinuation of antibiotic prophylaxis maybe appropriate*

Investigations

- *Throat swab culture*
- *ASO titre*
- *Acute phase reactants-ESR/CRP*
- *ECG-Prolonged PR interval*
- *CXR-Cardiomegaly in significant carditis*
- *Echocardiogram-to be done by Pediatric cardiologist*

Echocardiogram in rheumatic valvulitis

- *Echocardiographic demonstration of valvular regurgitation is not a prerequisite for the diagnosis of rheumatic carditis*
- *It should not be considered a limitation where the facilities are not available.*
- *The strict application of diagnostic criteria is mandatory to demonstrate pathological valvular regurgitation.*
- *Subclinical valvular regurgitation detected by echocardiography should not be included as an evidence of a major manifestation of carditis.*
- *Echocardiography can only play a limited role in cases of recurring Rf, unless a previous echocardiographic study is available for comparison.*

Management

- *No specific treatment*
- *Symptomatic combined with suppressive therapy*

General

- *Bed rest during the acute phase, until disease activity ceases (normal temperature and pulse rate)*
- *Patients without carditis can be ambulatory in 2-3 weeks*
- *It may be continued for 2-3 months if carditis present*

Patients with typical migratory polyarthrititis and carditis without cardiomegaly or congestive heart failure

- *Aspirin (Salicylates) 100 mg/kg/24 hours divided qid Po for 3-5 days, followed by 75 mg/kg/24 hours divided qid Po for 4 weeks*

Patients with carditis and cardiomegaly or congestive heart failure or pericarditis with effusion

- *Prednisolone-2mg/kg/24 hours in 4 divided doses for 2-3 weeks, followed by dose tapering of 5 mg/24 hours every 2-3 days*
- *Start Aspirin on tapering prednisolone at 75 mg/kg/day in 4 divided doses x 6 weeks*
- *Other measures: Fluid and salt restriction, diuretics, ACEIs and digoxin (use with caution since toxicity is enhanced in myocarditis)*

Treatment of Chorea

- *Primary treatment → elimination of streptococcal infection*
- *Secondary treatment → long term penicillin prophylaxis*
- *Symptomatic treatment → depends on impairment/disability*
- *Mild disability Rest without any medications*
May needs sedatives-Diazepam, Phenobarbitone
- *Severe disability with impairment in activities of daily living*
 1. *Neuroleptics - Haloperidol (0.025 mg/kg/day in divided doses going up to a maximum of 0.05 mg/kg/day in divided doses)*
 2. *Antiepileptics - Carbamazepine (10 mg/kg/day), Valproate (20 mg/kg/2day) duration 2-4 months*
 3. *Immunomodulation - 4-6 weeks course of oral prednisolone 2 mg/kg daily)*

Anti Streptococcal Therapy

- *Oral Penicillin V 250 mg QID for 10 days OR*
- *Oral Erythromycin 250 mg QID for 10 days (for penicillin hypersensitive) OR*
- *A single intramuscular injection of Benzathine penicillin to eradicate group A streptococcus from the upper respiratory tract*
- *After this, long term secondary prevention should be started*

Secondary Prophylaxis of Rheumatic Fever

- IM Benzathine Penicillin 0.6 mega units (<30 kg)
mega units (>30 kg) every 3 to 4 weeks OR
- Oral Penicillin V 250 mg twice daily OR
- Oral Erythromycin 250 mg twice daily if allergic to Penicillin

Table 3.15 Secondary prophylaxis of rheumatic fever

Category	Duration
ARF without carditis	5 years or until 21 years of age whichever is longer
ARF with carditis without residual valvular disease (Clinical/echo)	10 years or adulthood whichever is longer
ARF with carditis and residual valvular disease (Clinical/echo)	At least 10 years since last episode and at least until 40 years of age. Sometimes, life long prophylaxis