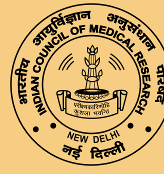


Department of Health Research
Ministry of Health and Family Welfare, Government of India

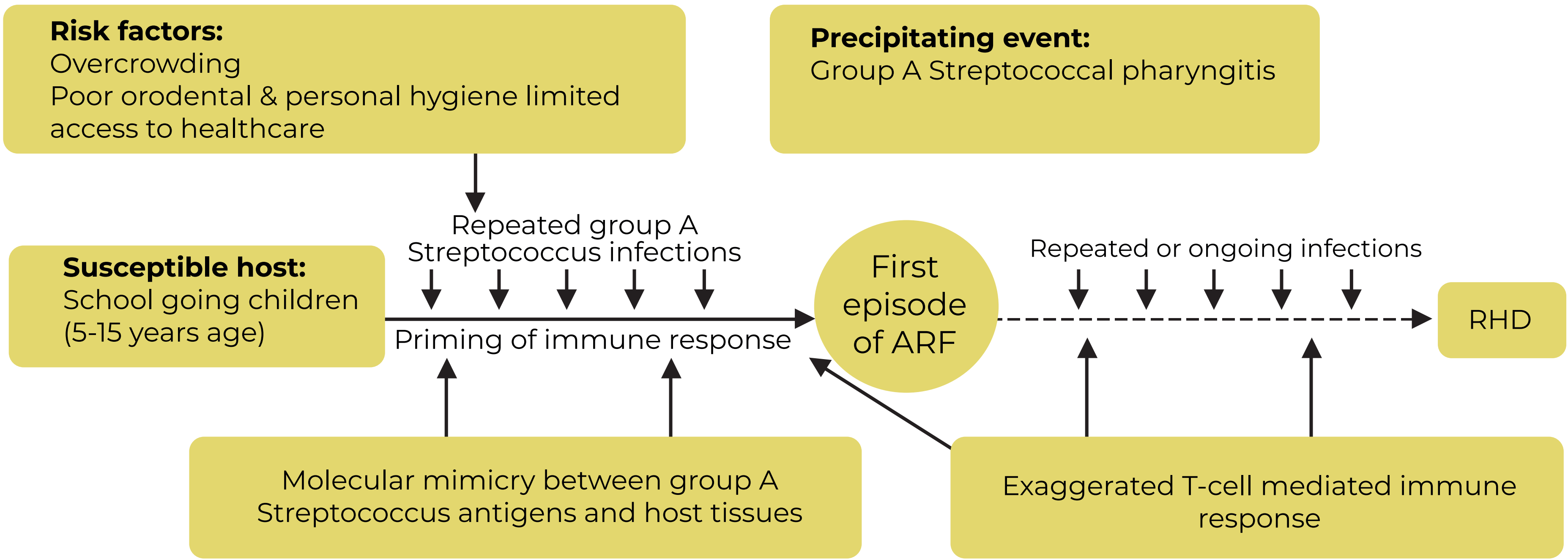
Standard Treatment Workflow (STW)

ACUTE RHEUMATIC FEVER

ICD-10-I01.9

Rheumatic fever (RF) is an acute, nonsuppurative inflammatory disease complicating untreated or partially treated Group A Streptococcus (GAS) pharyngitis

PATHOPHYSIOLOGY

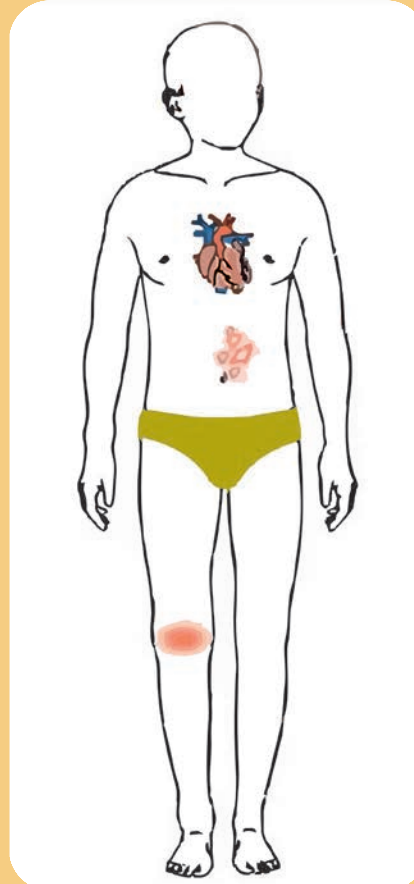


CLINICAL PRESENTATION

Arthritis (80%) – Most common manifestation
- Multiple joints
- Migratory – lasts <1 week in a joint
- Large joints – ankles, knees & wrist
- Exquisite tenderness with redness & swelling
- Prompt response to NSAIDs
- Leaves no deformity

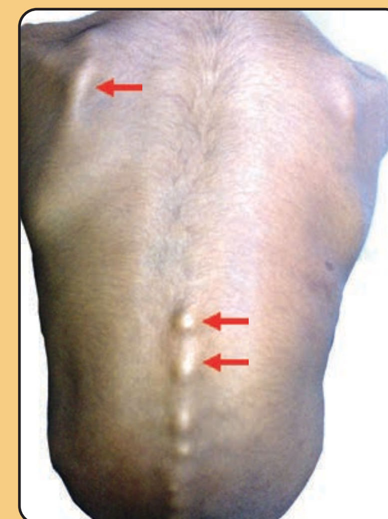
Carditis (50%) – Most devastating manifestation
- Tachycardia
- Dyspnoea
- Heart Failure
- Murmur on auscultation

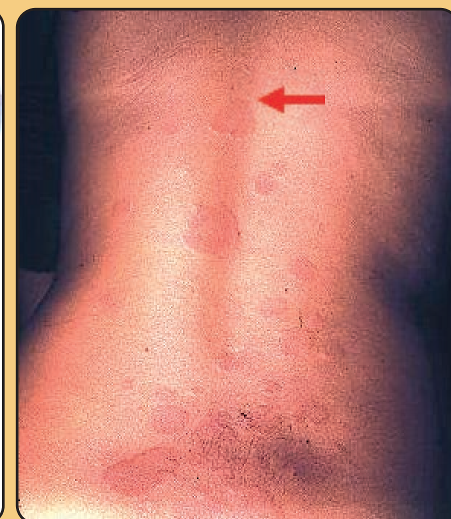
Chorea (10%) - 2-6 months after streptococcal sore throat
- Quasipurposive, involuntary movements with emotional lability
- Best seen in hands, arms, tongue and face
- Affects fine motor movement like handwriting



Subcutaneous nodules – rare
Painless, pea-sized, hard nodules
On extensor surfaces of limbs, skull and back

Erythema marginatum (5%)
Transient pink macule with fading centre
Mostly located on the trunk and limbs





DIAGNOSIS BASED ON JONES CRITERIA

For all patient populations with evidence of preceding group A streptococcal infection

Diagnosis:

Initial ARF	2 major or 1 major plus 2 minor Criteria
Recurrent ARF	2 major or 1 major and 2 minor or 3 minor Criteria
Recurrent ARF in RHD	2 minor (No major criteria needed)

Criteria

Major

Low-risk populations[a]	***Moderate and high-risk populations***[a]
Carditis (Clinical and/or subclinical)[b]	Carditis (Clinical and/or subclinical)[b]
Arthritis (Polyarthritis only)	Arthritis (Monoarthritis or polyarthritis or polyarthralgia)[c]
Chorea	Chorea
Erythema marginatum	Erythema marginatum
Subcutaneous nodules	Subcutaneous nodules

Minor

Polyarthralgia[c]	Monoarthralgia
Fever (≥38.5°C)	Fever (≥38°C)
ESR >60 mm/h and/or CRP ≥3 mg/dL	ESR >30 mm/h and/or CRP ≥3 mg/dL
Prolonged PR on ECG (for age) (unless carditis is a major criterion)	Prolonged PR on ECG (for age) (unless carditis is a major criterion)

Essential

Throat culture or antigen positive for streptococcal sore throat OR elevated ASO titers (>320 U)

[a]Low-risk populations ARF incidence ≤2/ per 100 000 school-aged children or all-age RHD prevalence of ≤1/per 1000 population per year

[b]Subclinical carditis is pathological echocardiographic valvulitis

[c]Polyarthralgia should only be considered as a major manifestation in moderate-to high-risk populations after exclusion of other conditions. Joint manifestations can only be considered in either the major or minor categories but not both in the same patient

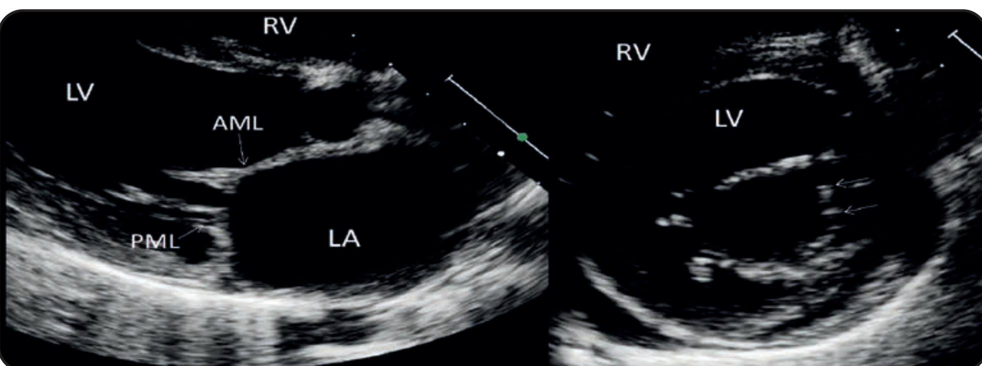
Erythema marginatum and subcutaneous nodules are 'stand-alone major criteria

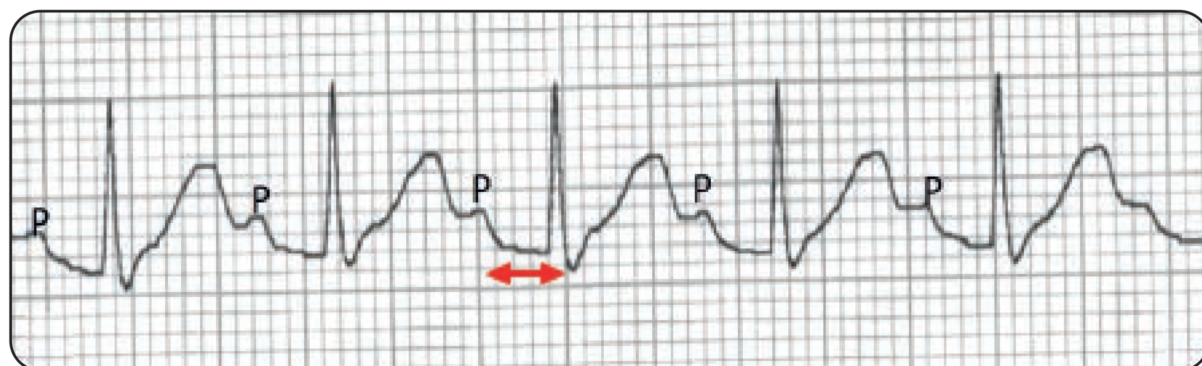
LABORATORY INVESTIGATIONS

Essential		*Optional*
TLC, DLC	Chest X ray	Throat swab antigen
ESR, CRP	Anti-streptolysin O	Throat swab culture
ECG (12 lead)	Echocardiogram	Anti DNAse-B

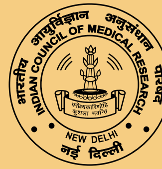
DIFFERENTIAL DIAGNOSIS

1. Pediatric autoimmune neuropsychiatric disorders (PANDAS)-autoimmune disorder
2. Post streptococcal reactive arthritis (PSRA)- small joint arthritis, poor response to NSAIDs
3. Juvenile rheumatoid arthritis
4. Infective endocarditis





Department of Health Research
Ministry of Health and Family Welfare, Government of India



icmr INDIAN COUNCIL OF MEDICAL RESEARCH
Serving the nation since 1911

Standard Treatment Workflow (STW)

ACUTE RHEUMATIC FEVER

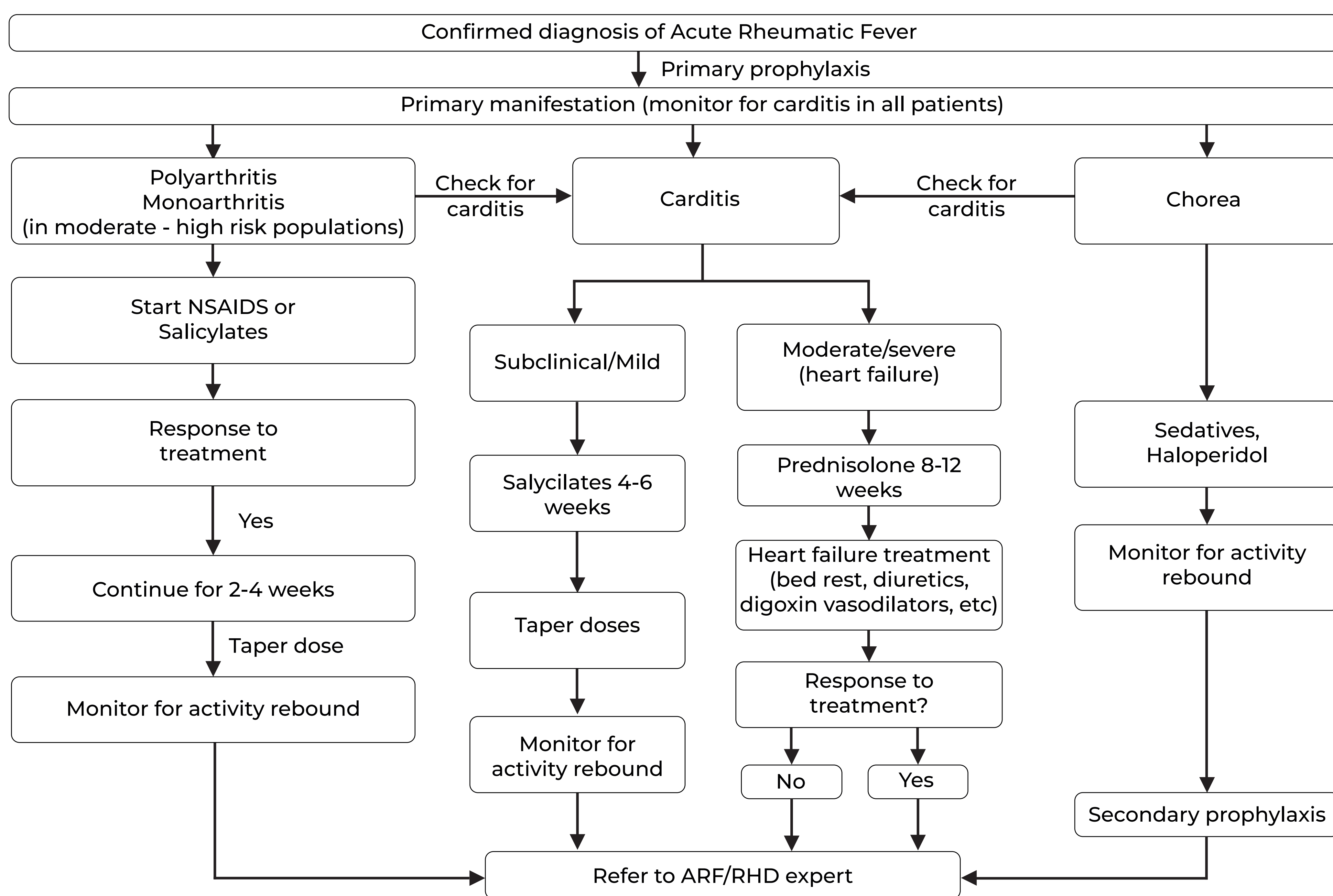
(Continued)

MANAGEMENT

Primary prophylaxis (to Eradicate streptococcus)

Agent	Dose	Duration
Benzathine penicillin (Penicillin G)	≤27kg 6,00,000U >27kg 12,00,000U	Once
or		
Phenoxymethyl penicillin(Penicillin V)	≤27kg 250mg/dose <27kg 500mg/dose	10 days
For individuals allergic to penicillin		
Amoxicillin Erythromycin	25-50mg/kg/day divided into 3 doses (maximum 1g/day) 20-40mg/kg/day divided into 2-4 doses (maximum 1g/day)	10 days

Anti-inflammatory therapy & supportive care



Clinical Manifestation	Treatment Schedule	Duration
Moderate/Severe carditis	Prednisolone 2mg/kg/day once daily (Aspirin while tapering Prednisolone)	8-12 WKS
Mild carditis	Aspirin 75-100mg/day divided into 4 doses	2-4 WKS
Polyarthritis	Aspirin 75-100mg/day divided into 4 doses or Naproxen 10-20mg/kg/day	2-4 WKS
Chorea	Carbamazepine 4-10mg/kg/day or Valproic acid 20-30mg/kg/day or Haloperidol 2-6mg/day	Variable depending upon the need of the patient

Secondary prophylaxis

Category of Patient	Duration
Patients without carditis	5 years after the last ARF episode or until 21 years age (whichever is longer)
Patients with carditis but no RHD	10 years after the last acute episode or until 25 years age (whichever is longer)
Patients with RHD who have undergone valve surgery (repair or replacement)	At least until 40 years age (preferably lifelong)

Agent	Dose	Route
Benzathine penicillin (Penicillin G)	≤27kg 6,00,000U >27kg 12,00,000U	Intramuscular
or		
Phenoxymethyl penicillin(penicillin V)	250mg twice daily	Oral
For individuals allergic to penicillin		
Erythromycin	250mg twice daily	Oral

ABBREVATIONS

ARF: Acute Rheumatic Fever
ASO: Antistreptolysin O
CRP: C-reactive protein
DLC: Differential Leukocyte Count
ECG: Electrocardiogram
ESR: Erythrocyte Sedimentation Rate
NSAIDs: Non-Steroidal Anti-Inflammatory Drugs
PANDAS: Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections
RHD: Rheumatic Heart Disease
TLC: Total Leukocyte Count

REFERENCES

1. Gewitz MH, Baltimore RS, Tani LY, Sable CA, Shulman ST, Carapetis J, Remenyi B, Taubert KA, Bolger AF, Beerman L, Mayosi BM, Beaton A, Pandian NG, Kaplan EL; American Heart Association Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the Council on Cardiovascular Disease in the Young. Revision of the Jones Criteria for the diagnosis of acute rheumatic fever in the era of Doppler echocardiography: a scientific statement from the American Heart Association. Circulation. 2015 May 19;131(20):1806-18. doi: 10.1161/CIR.0000000000000205. Epub 2015 Apr 23. Erratum in: Circulation. 2020 Jul 28;142(4):e65. PMID: 25908771.
2. Kumar RK, Antunes MJ, Beaton A, Mirabel M, Nkomo VT, Okello E, Regmi PR, Reményi B, Sliwa-Hähnle K, Zühlke LJ, Sable C; American Heart Association Council on Lifelong Congenital Heart Disease and Heart Health in the Young; Council on Cardiovascular and Stroke Nursing; and Council on Clinical Cardiology. Contemporary Diagnosis and Management of Rheumatic Heart Disease: Implications for Closing the Gap: A Scientific Statement From the American Heart Association. Circulation. 2020 Nov 17;142(20):e337-e357. doi: 10.1161/CIR.0000000000000921. Epub 2020 Oct 19. Erratum in: Circulation. 2021 Jun 8;143(23):e1025-e1026. PMID: 33073615.
3. Handbook on prevention and control of rheumatic fever and rheumatic heart diseases. Directorate General of Health Services. Government of India 2015. accessed online on July 18, 2023.

INJECTABLE PENICILLIN IS SAFE; ALLERGY IS UNCOMMON

This STW has been prepared by national experts of India with feasibility considerations for various levels of healthcare system in the country. These broad guidelines are advisory, and are based on expert opinions and available scientific evidence. There may be variations in the management of an individual patient based on his/her specific condition, as decided by the treating physician. There will be no indemnity for direct or indirect consequences. Kindly visit the website of ICMR for more information: (**icmr.gov.in**) for more information. ©Indian Council of Medical Research, Ministry of Health & Family Welfare, Government of India.