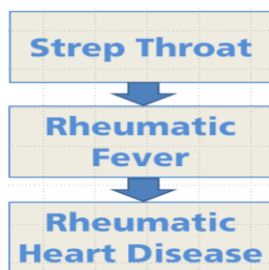


## ACUTE RHEUMATIC FEVER/RHEUMATIC HEARTDISEASE

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Rheumatic Fever is the most common acquired heart disease in children, adolescents, and young adults. In the World Health Organization 2018 report, there are 30 million people affected worldwide. And approximately 300,000 deaths per year are caused by Acute Rheumatic Fever and Rheumatic Heart Disease (ARF/RHD). Children and adolescents between the ages of 5-15 years are at highest risk. Poverty, overcrowded housing, and poor access to healthcare are risk factors, particularly for underdeveloped and developing countries.

The relationship with Group A  $\beta$ -hemolytic Streptococcus (GAS) infection and RF/RHD has been established. The actual mechanism is unknown. But it has been postulated that an autoimmune or hypersensitivity reaction to GAS produces pathogenic autoantibodies to cardiac, brain, connective and subcutaneous tissues. There is diffuse inflammation of the tissues involved.



Strep Throat presentation:

enlarged and erythematous tonsils, erythematous pharynx,  
with patchy discrete infiltrates  
fever  
headache  
nausea and vomiting  
enlarged anterior cervical lymph nodes

GAS infection presenting as Streptococcal Sore Throat (also known as Strep Throat) initiates the inflammatory process. Repeated untreated Strep Throat leads to Acute Rheumatic Fever. And ARF recurrences lead to RHD.

Accepted evidence of previous GAS infection include: throat swab and culture, rapid antigen detection test, and anti-streptolysin O antibody titer (ASOT). Recently, anti-DNAse B has been recommended, particularly if ASOT is negative.

The diagnosis of Rheumatic Fever remains to be clinical in nature. Since the conception of Jones Criteria in 1944, it has been revisited and revised at least six times. The latest revision by the American Heart Association was published in 2015.

The current update takes into consideration the population risk. The low risk population is defined as having an ARF incidence of <2 per 100,000 school-aged children (5-14 years old) per year, or an all age prevalence of RHD of ≤1 per 1,000 population per year. And, children not clearly from low-risk population are at moderate to high risk depending on their reference population. The changes are summarized in the following tables:

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

Table 1. diagnosis of ARF

Low-risk	<i>Moderate- and high-risk</i>
<b>CARDITIS</b> Clinical and/or <i>Subclinical</i>	Clinical and/or <i>Subclinical</i>
<b>ARTHRITIS</b> Polyarthritis only	Polyarthritis or <i>Monoarthritis</i> <i>Polyarthralgia</i>
<b>Chorea</b> <b>Erythema marginatum</b> <b>Subcutaneous nodules</b>	<b>Chorea</b> <b>Erythema marginatum</b> <b>Subcutaneous nodules</b>

Table 2. Major criteria; changes are in *italics*

Low-risk	<i>Moderate- and high-risk</i>
Polyarthralgia	<i>Monoarthralgia</i>
Fever (≥38.5°C)	<i>Fever (≥38°C)</i>
ESR ≥60 mm in the first hour and/or CRP ≥3.0 mg/dL	<i>ESR ≥30 mm/h</i> and/or CRP ≥3.0 mg/dL
Prolonged PR interval (unless carditis is a major criterion)	Prolonged PR interval (unless carditis is a major criterion)

Table 3. Minor criteria; changes are in *italics*

## MAJOR CRITERIA

**CARDITIS**- pancarditis; but the hallmark is valvulitis

- ▶ Apical systolic murmur (Mitral regurgitation)
  - ▶ Apical mid-diastolic murmur (Carey-Coomb's)
  - ▶ Basal diastolic murmur (Aortic regurgitation)
  - ▶ Basal systolic murmur (Tricuspid regurgitation)
- Other cardiac presentations:
- ▶ Resting tachycardia
  - ▶ Muffled heart sounds
  - ▶ Gallop rhythm
  - ▶ Pericardial friction rub
  - ▶ Congestive heart failure
  - ▶ Subclinical carditis/ Echo carditis- Echocardiographic abnormalities (based on strict criteria given by the AHA) in acute rheumatic fever in the absence of clinical carditis

## ARTHRITIS

- ▶ Most frequent & benign but least specific
- ▶ Asymmetric & migratory Inflammation
- ▶ Larger joints: knees, ankles, elbows, wrists
- ▶ Rapid healing without sequelae
- ▶

## CHOREA

- ▶ Sydenham chorea; St. Vitus' dance; Chorea minor
- ▶ Rheumatic involvement of basal ganglia & caudate nucleus
- ▶ Purposeless & involuntary movement; muscular incoordination & weakness; emotional lability & slurred speech
- ▶ Delayed manifestation & usually does not follow Jones criteria to diagnose RF
- ▶ Disappear in 1 – 2 wks, even w/o treatment

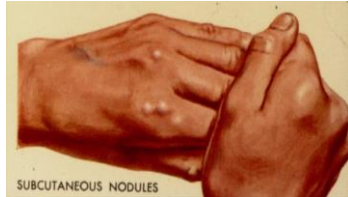
## ERYTHEMA MARGINATUM

- ▶ Distinctive non-pruritic transient rash
- ▶ Pale centers with round or serpiginous margin
- ▶ Blanches
- ▶ Trunk & proximal extremities; not the face
- ▶ Induced by heat



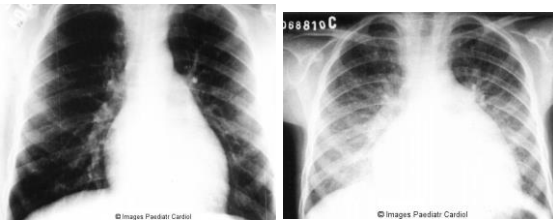
## SUBCUTANEOUS NODULES

- ▶ Small, firm, painless, freely movable & transient
- ▶ Extensor surface of elbows, knees, & wrists; scalp & spinal areas
- ▶ Often seen with carditis



## OTHER LABORATORY FINDINGS:

- ▶ **CBC**
  - ▶ Anemia ; Leukocytosis
- ▶ **ECG**
  - ▶ Sinus tachycardia
  - ▶ 1° AV block (also a minor criterion)
  - ▶ Possible chamber enlargement
  - ▶ Rarely 2° AV block, low voltages, ST-T wave changes
- ▶ **CXR**



- ▶ Mild carditis – No cardiomegaly
- ▶ Moderate carditis – mild cardiomegaly
- ▶ Severe carditis – cardiomegaly with severe pulmonary congestion or edema

Rheumatic Heart Disease (RHD), on the other hand, is chronic valvar heart disease as a sequelae of ARF and its recurrences. It denotes permanent valvar damage due to fibrosis.

RHD is more likely to develop following ARF if:

- ▶ The initial episode of ARF was severe.
- ▶ The heart was affected with ARF.
- ▶ ARF occurred at a young age.
- ▶ There has been recurrent ARF.

50% of people with RHD do not remember having ARF.

Valve **Regurgitation** suggests that heart valves:

- ▶ Are thickened and stick against the walls of the heart
- ▶ Leak (the blood flows backwards over the valve)

Valve **Stenosis** suggests that heart valves:

- ▶ Become stiff
- ▶ Do not allow blood to flow through easily (restricted forward flow)
- ▶ Occurs at least 5 years after initial ARF

*Mitral regurgitation* - a pansystolic murmur heard loudest at the apex and radiating laterally to the axilla

- ▶ ECG-Normal; LAE, LVH
- ▶ CXR-Normal; LAE, LVH; Pulmonary congestion
- ▶ Echo-Thickened valve; Dilated MV annulus; ECG-Normal;

LAE, LVH

*Mitral stenosis* - a low-pitched, diastolic rumble heard best at the apex with the bell of the stethoscope and with the person lying in the left lateral position.

- ▶ ECG- Normal; LAE, RVH
- ▶ CXR- LAE, RVH; Pulmonary congestion; Dilated MPA
- ▶ Echo- Thickened fixed leaflets; Small MVA

*Aortic regurgitation* - a diastolic blowing decrescendo murmur best heard at the left sternal border with the person sitting up and leaning forward in full expiration.

- ▶ ECG- LVH; Strain pattern
- ▶ CXR- LVH; Dilated aorta
- ▶ Echo-Thickened leaflets with prolapse

*Aortic stenosis* - a loud, low pitched mid-systolic ejection murmur best heard in the aortic area, radiating to the neck.

*Tricuspid regurgitation*

- ▶ related to pulmonary hypertension and RV failure

*Tricuspid stenosis*

- ▶ Rare

*Pulmonary regurgitation*

- ▶ related to pulmonary hypertension

*Pulmonary stenosis*

- ▶ very rare

## MEDICAL MANAGEMENT

### Prophylaxis

- ▶ 1<sup>o</sup> - prevents 1<sup>st</sup> episode of ARF; treatment of Gas infection  
Penicillin, oral, for 10 days  
Or if allergic to Penicillin- Erythromycin or Azithromycin
- ▶ 2<sup>o</sup> - prevents recurrences of RF  
Benzathine Penicillin 1.2 M U q 21 days  
or Penicillin, oral, 250 mg, BID  
or if allergic to Penicillin- Erythromycin
- ▶ Duration of 2<sup>o</sup> prophylaxis
  - ▶ Arthritis - at least 5 years or age 21 years whichever is longer (if recurrence free)
  - ▶ Carditis - at least 10 years or age 21 years whichever is longer (recurrence free & no residual heart disease)
  - ▶ RHD- for life

### Other medications

- ▶ Anti-inflammatory agents: 6 – 8 weeks
  - ▶ ASA - 100 mg/kg/day
  - ▶ Prednisone - 2 mg/kg/day
- ▶ Anti-CHF – inotrope, preload unloader, afterload unloader
- ▶ Anticoagulant
- ▶ Anti-Arrhythmics

## SURGICAL MANAGEMENT

- ▶ MR
  - ▶ Valvuloplasty; annular plication; valve replacement
- ▶ MS
  - ▶ Commisurotomy; valve replacement
- ▶ AR
  - ▶ Valvuloplasty; valve replacement
  - ▶

## SUPPORTIVE MANAGEMENT

- ▶ Complete bed rest & modified activity
- ▶ Attention to diet
- ▶ Fluid balance

## **FOLLOW-UP MANAGEMENT**

- ▶ Education
- ▶ Adherence to secondary prophylaxis
- ▶ Regular clinical assessment and follow-up echocardiography
- ▶ Management of cardiac failure
- ▶ Management of atrial fibrillation
- ▶ Dental care and Infective endocarditis prophylaxis
- ▶ Family planning referral (for women)
- ▶ Vaccination
- ▶ Appropriate surgical intervention

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