Acute Rheumatic Fever

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Acute Rheumatic Fever

(Licks the Joints and Bites the Heart)

Etiology:

- ✓ **Group A beta hemolytic streptococci (GABS)**
- ✓ **Serotypes (M protein..,3,18)**
- ✓ **Rheumatogenicity**

Epidemiology:

- ✓5-15 y/o
- ✓Developing or underdeveloped countries
- ✓New outbreaks
- ✓Crowding
- ✓Low sanitary
- ✓Season
- ✓Pharyngitis but not impetigo

Pathogenesis:

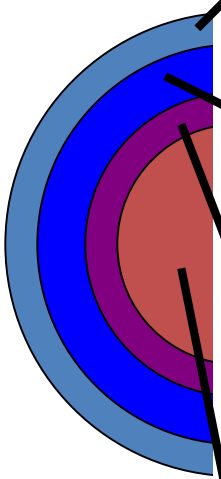
- ✓**Unknown**
- ✓**Toxic effect**
- ✓**Abnormal immune response**
- ✓**Genetic influence** (alloantigen on surface of non T-lymphocytes in 99% of ARF but 13% of controls)

Pathogenesis:

Abnormal immune response

- ✓Alteration in helper and suppressor T cells
- ✓Anti heart antibodies
- ✓IgG, IgM, C3 deposition in pericard

Streptococcal and Human Tissue Immunologic Cross Reactivity



Capsule ⇨ **Joint**

- Hyaluronic acid

Cell wall ⇨ **Myocardium**

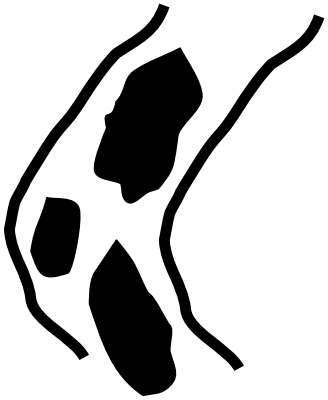
- M – protein

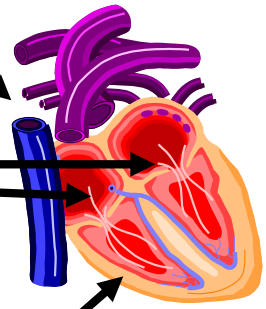
Group carbohydrate ⇨ **Valves**

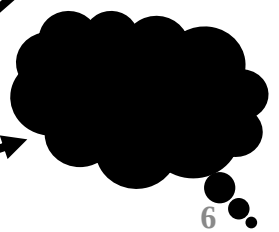
- Rhamnose
- N-acetyl glucosamine

Protoplast membrane ⇨ **Myoc.Sarcolemma, Subthalamic & Caudate nuclei**

- Protein, lipid, glucose







Clinical manifestations:

✓Polyarthritis:

- ➢The most common
- ➢Migratory polyarthritis
- ➢Tenderness
- ➢Large joints
- ➢Duration
- ➢Joint effusion and analysis
- ➢Response to ASA
- ➢*There is often an inverse relationship between the severity of arthritis and the severity of cardiac involvement.*

Poststreptococcal Reactive Arthritis:

- ✓ **Arthritis and recent evidence for GABS infection but do not fulfill the Jones criteria**
- ✓ **Arthritis course & response to Rx. like JRA**
- ✓ **About 5% acquire valvular disease without secondary prophylaxis**

Clinical manifestations:

- ✓ **Carditis:**
  - ➢ **40-80%**
  - ➢ **Mitral valve** (the most common)
  - ➢ **Pancarditis**
  - ➢ **Pericarditis**
  - ➢ **Arrhythmia**
  - ➢ **CHF**

- **Subclinical Carditis:**
  - **>90%**
  - **? Over diagnosis**

The Incidence of Carditis Depends on Age:

Age:	Carditis
< 3 Y/O	90%
3-6 Y/O	50%
14-17 Y/O	32%
> 25 Y/O	Rare

Chance of Having Normal Heart in Follow Up:

Initial Attack	5 Year Follow Up	10 Year Follow Up
No Carditis	96%	94%
CHF	40%	30%

Carditis:

- **Isolated Mitral valve disease has 76% rheumatic etiology**
- **Isolated Aortic valve disease has 13% rheumatic etiology**
- **Combined Mitral and Aortic valve disease has 97% rheumatic etiology**

Roberts: in Moss & Adams textbook, 2001 edition

- ✓ **First streptococcal pharyngitis attack cause ARF in: Epidemic = 3%, Endemic = 0.3%**
- ✓ **Second attack cause ARF in 11- 65% (after 10 years 4-8%)**
- ✓ **RHD and cardiomegaly has 43% recurrence rate**
- ✓ **RHD and no cardiomegaly has 27% recurrence rate**
- ✓ **Normal heart has 10% recurrence rate**
- ✓ **< 5% have chronic active carditis for > 6 months duration**
- ✓ **Rheumatic activity may be prolonged with rebound**

Clinical manifestations:

- ✓ **Sydenham chorea:**
  - ➢ **Deterioration of handwriting**
  - ➢ **Emotional instability**
  - ➢ **Milkmaid's, silk fork and bag of worm signs**
  - ➢ **Late presentation**
  - ➢ **Disappear with sleep**

Clinical manifestations:

- ✓ **Subcutaneous nodule:**
  - ➢ **Not tender**
  - ➢ **Extensor surface**
  - ➢ **Associated with severe carditis (MS)**

Clinical manifestations:

- ✓ **Erythema Marginatum:**
  - ➢ **Not pathognomonic**
  - ➢ **Non tender**
  - ➢ **No itching**
  - ➢ **Associated with chronic carditis**

Erythema Marginatum:

Minor Manifestations:

- ✓ **Fever**
- ✓ **Arthralgia**

Modified Jones Criteria:

Major	Minor
Carditis	Fever
Polyarthritis, migratory	Arthralgia
Erythema marginatum	ESR, CRP
Chorea	Prolonged PR interval
Subcutaneous nodule	? Previous Hx. of ARF

Plus:

Evidence of preceding streptococcal infection

Except: Chorea, insidious or late onset carditis

Modified Jones criteria:

- ✓ 2 major
- ✓ or one major and 2 minor criteria

  plus evidence for recent streptococcal infection.

Recurrence of ARF:

- ✓ Only one major criteria
- ✓ or fever, arthralgia,
- ✓ and elevated acute phase reactants

  plus evidence for recent streptococcal infection.

Lab findings:

- ✓ **ASO titer** (acute and convalescent sera)
- ✓ **ADB**
- ✓ **AH**
- ✓ **Throat culture**
- ✓ **ECG**
- ✓ **Echocardiography**

Frequency of Elevated Antibody Titer (%) in Patients With ARF

	ASO	Anti-Dnase B	ASO & Anti Dnase B
Normal Controls	19	19	30
ARF	83	82	92

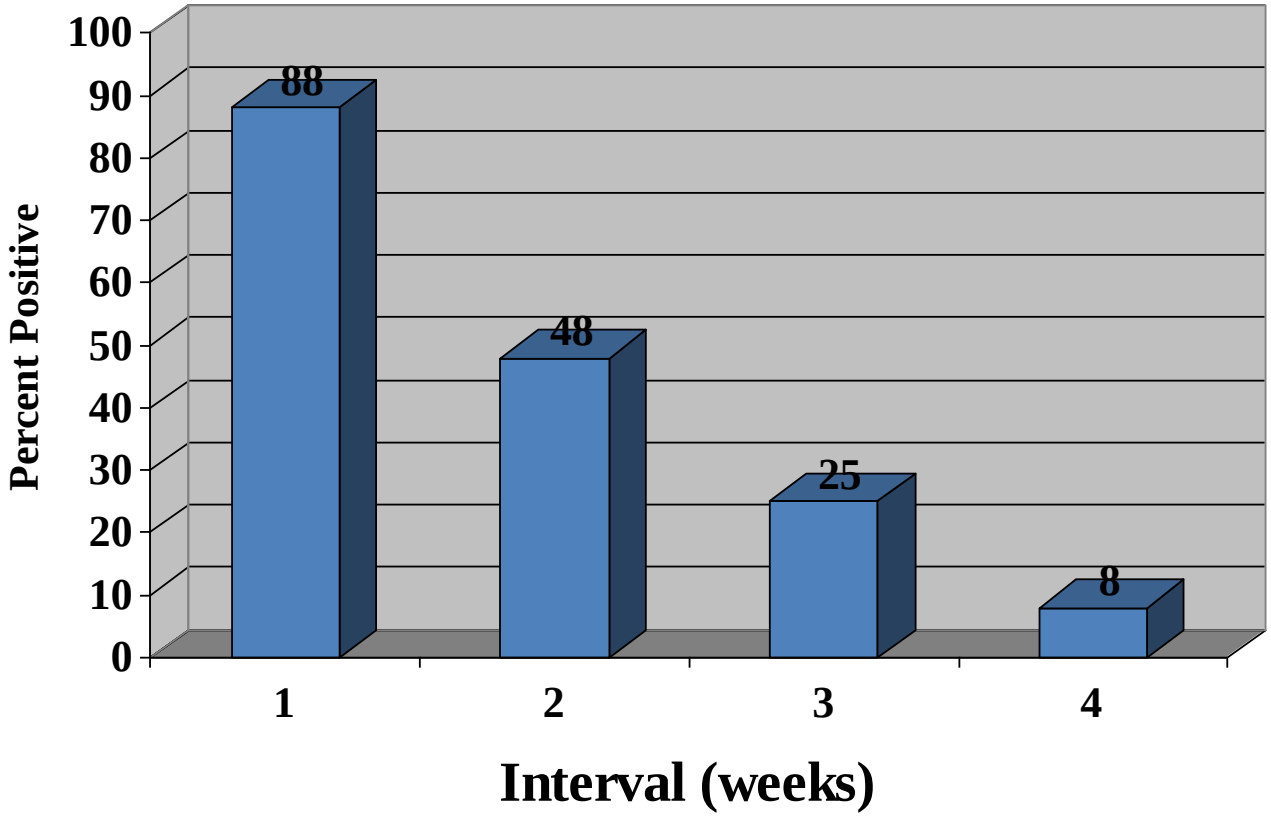
ASO Titers in Children in Tropical Countries

Country	% ASO titer Over 200 units
Thailand	17.7
Pakistan	18.4
Burma	37.2
Mongolia	52.3
Algeria	36.4
Kenya	40.6
Nigeria	53.3

Antibody response:

- **Anti-A carbohydrate test reaches a peak 1 month after GABS infection and declines to normal levels about 2 years thereafter, except in patient with persistent rheumatic MR which will be maintained for several years.**

Frequency of Positive Throat Culture Following Untreated Streptococcal Pharyngitis



Differential diagnosis:

- ✓ **JRA**
- ✓ **SBE**
- ✓ **Connective tissue disorders**

Differential diagnosis:

ARF	JRA
Large joints	Small joints
Migratory arthritis	Additive arthritis
<7 days, Max 6 weeks arthritis	**>7 days, at least 6 weeks arthritis**
Very tender	Painful
Red joints	Pallor over the joints
ASA effective in 24-48 hr	ASA effective in 5-7 days
No damage (? Jaccoud's)	Joint damage possible
Synovial fluid <10000 WBC	**Synovial fluid >20000 WBC**
> Endocarditis	> Pericarditis

Treatment:

- ✓ **Penicillin**
- ✓ **ASA (**first sign of toxicity is hyperventilation**)**
- ✓ **Prednisolone**
- ✓ **Rx. For CHF**
- ✓ **Bed rest**
- ✓ **Rx. Of Chorea** (Phenobarbital (is choice) 16-32 mg every 6-8 hr PO, chlorpromazine 0.5 mg/kg every 4-6 hr PO, diazepam, haloperidol 0.01-0.03 mg/kg/24 hr divided bid PO, valproate, vitamine E 50 mg/2 wk)
- ✓ **SBE prophylaxis (**not penicillin**)**

- **Patients with typical migratory polyarthritis and those with carditis without cardiomegaly or congestive heart failure should be treated with oral salicylates.**
- **The usual dose of aspirin is 100 mg/kg/day in 4 divided doses PO for 3-5 days, followed by 75 mg/kg/day in 4 divided doses PO for 4 wk.**
- **There is no evidence that nonsteroidal antiinflammatory agents are any more effective than salicylates.**

- **Patients with carditis and cardiomegaly or congestive heart failure should receive corticosteroids.**
- **The usual dose of prednisone is 2 mg/kg/day in 4 divided doses for 2-3 wk followed by a tapering of the dose that reduces the dose by 5 mg/24 hr every 2-3 days.**
- **At the beginning of the tapering of the prednisone dose, aspirin should be started at 75 mg/kg/day in 4 divided doses for 6 wk.**
- **Supportive therapies for patients with moderate to severe carditis include digoxin, fluid and salt restriction, diuretics, and oxygen.**
- **The cardiac toxicity of digoxin is enhanced with myocarditis.**

Surgical Treatment:

- ✓**MR:**
  - ✓**Functional class III or IV**
  - ✓**LVSD >26 mm/m^2**
  - ✓**LVDD >40 mm/m^2**
  - ✓ **SF < 31%**
- ✓**AI:**
  - ✓**Functional class III or IV**
  - ✓**LVSD >55 mm**
  - ✓**lower limit of normal SF & EF**

Prevention:

- ✓ **Primary prophylaxis**
- ✓ **Secondary prophylaxis**
  - ➢ **3 or 4 weeks 600.000-1.200.000 units Benzathine Penicillin**
- ✓ **No vaccine available**

Duration of secondary prophylaxis? AHA 2009

- **Rheumatic fever without carditis 5 yr or until 21 yr of age, whichever is longer.**
- **Rheumatic fever with carditis but without residual heart disease (no valvular disease) 10 yr or until 21 yr of age, whichever is longer.**
- **Rheumatic fever with carditis and residual heart disease (persistent valvular disease) 10 yr or until 40 yr of age, whichever is longer, sometimes lifelong prophylaxis**