

ACUTE RHEUMATIC FEVER



DEFINITION



- Rheumatic fever is an inflammatory process which can involve the joints, heart, skin and brain
- It is caused by antibody cross reactivity and occurs 2-3 weeks after a Group A Streptococcal infection.



BACKGROUND



- Primarily affects children between ages 5-12
- Generally occurs 2-3 weeks after Group A Streptococcal infection (strep throat or scarlet fever)
- Globally, 3% of those with an untreated streptococcal infection develop rheumatic fever
- 40% of those with Acute Rheumatic Fever develop mitral stenosis as adults

BACKGROUND



- Cutaneous streptococcal infections have not been shown to initiate Acute Rheumatic Fever.
- Strains of certain M serotypes/genotypes of streptococci have higher associations than other genotypes
- Epidemics of Acute Rheumatic Fever in Trinidad and Chile showed that streptococci causing Acute Rheumatic Fever belonged to different serotypes than those that cause Acute Glomerulonephritis.

PATHOPHYSIOLOGY

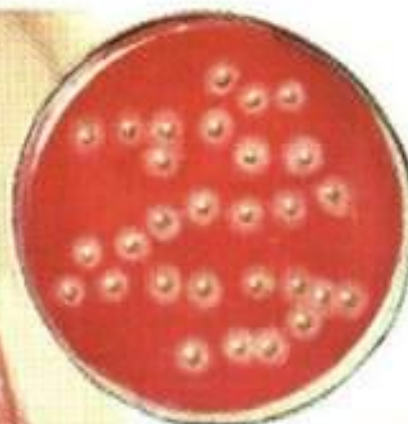
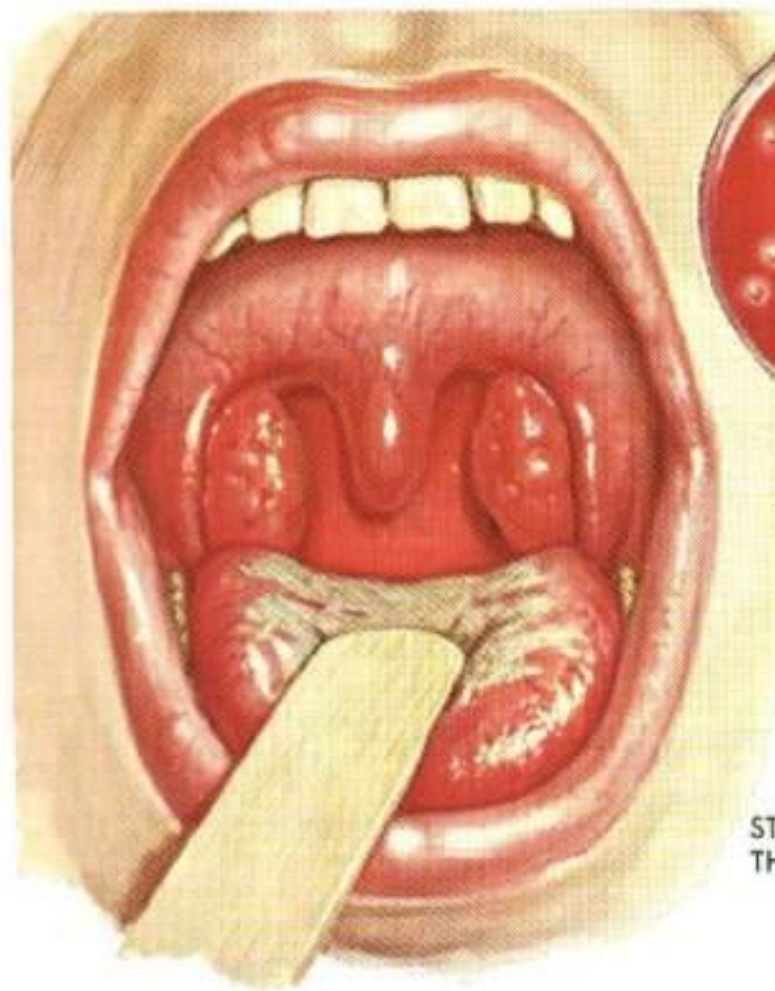


- Exact mechanism of how Group A streptococcal infection causes Acute Rheumatic Fever is unknown however it is believed to be caused by a cross reactivity of antibodies
- Suggested Theories
 - Toxic effects of streptococcal products (streptolysin S or O) which then cause direct tissue injury
 - Serum Sickness-like reaction mediated by antigen-antibody complexes
 - Autoimmune phenomenon

PATHOPHYSIOLOGY



- More support for an autoimmune phenomenon (Type II hypersensitivity reaction)
- During strep infection, antigen presenting cells present bacterial antigen to helper T cells. These helper T cells then activate B cells to induce production of antibodies against the Streptococcal cell wall. These antibodies can also interact with other cells in the body (for example, myocardium or joints, etc) producing the symptoms responsible with acute rheumatic fever



RHEUMATIC FEVER

HEMOLYTIC STREPTOCOCCI

IMMUNOLOGIC REACTION?
RHEUMATIC FEVER
10 TO 14 DAYS LATER
(MUCH LONGER GAP
FOR CHOREA)

STREPTOCOCCAL
THROAT INFECTION

F. Netter M.D.
© CIBA



JOINTS



HEART



SKIN



NERVOUS
SYSTEM
(CHOREA)

PATHOGENESIS



- Most patient have elevated antibody titers to at least one streptococcal antibody
 - Streptolysin O
 - Hyaluronidase
 - Streptokinase

CLINICAL MANIFESTATIONS



- If initial complaint is polyarthrititis, disease generally has more abrupt onset compared to if initial presentation is with myocarditis.
- Arthritis occurs in 75% of initial attacks

CLINICAL MANIFESTATIONS (JOINTS)



- Arthralgias and arthritis (may be migratory)
- Warm, swollen, tender joints
- Usually involves the knees, ankles, elbows and wrists
- Lasts 2-3 weeks

Arthralgias and arthritis



CLINICAL MANIFESTATIONS



- **Subcutaneous Nodules:** usually associated with severe carditis and occur several weeks after onset.
 - Firm, painless nodules (up to 2cm) found over bony surfaces and tendons
 - Occur near elbows, knees, wrists, achilles tendon, vertebral joints
 - Usually persist for 1-2 weeks

SUBCUTANEOUS NODULES



SUBCUTANEOUS NODULES



CLINICAL MANIFESTATIONS



- Erythema Marginatum: nonpruritic, painless erythematous rash on trunk and/or proximal extremities
 - Macular lesions with raised margins and central clearing
 - May last from weeks to months

ERYTHEMA MARGINATUM



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CLINICAL MANIFESTATIONS



- Sydenham's Chorea: neurologic disorder with muscular weakness, emotional lability and involuntary, uncoordinated, purposeless movements
 - Disappear during sleep
 - Mainly occur in hands, feet and face
 - Sensation intact
 - Lasts 2-4 months

Sydenham's Chorea



DIFFERENTIAL DIAGNOSIS



- Poststreptococcal reactive arthritis: is non-migratory
- Rheumatoid Arthritis
- SLE
- Infective endocarditis
- Sickle Cell disease
- Drug reactions
- TB
- Lyme Disease
- Serum Sickness

DIAGNOSIS



- JONES CRITERIA

TREATMENT



- Anti-inflammatory Agents
- Antibiotics
- Prophylaxis

ANTI-INFLAMMATORY AGENTS



- Aspirin 4-8grams/day for adults
- Continue anti-inflammatory therapy until ESR or CRP are normal
- May need steroids if there is cardiac involvement to help prevent sequelae such as mitral stenosis
- Corticosteroids, if indicated, are given at prednisone 2mg/kg/day for 2 weeks and then tapered

ANTIBIOTICS



- Penicillin for at least 10 days
- Penicillin 500mg BID-TID
- Can use erythromycin for PCN allergic patients (given at 40mg/kg/day given in 2-4 doses/day)

