ACUTE RHEUMATIC FEVER
RHEUMATIC FEVER

“Licks the joint, bites the heart”
INTRODUCTION

• Leading cause of Acquired Heart disease

• Incidence: Peak 5 – 15 yrs.
ORGANISM

Group-A beta hemolytic streptococci
DISEASES CAUSED BY GROUP A STREPTOCOCCI

- Pharyngitis
- Impetigo/pyoderma
- Pneumonia, Necrotizing fasciitis
- Rheumatic fever
- Glomerulonephritis
- Osteomyelitis
- Scarlet fever & erysipelas
- Toxic shock syndrome
RHEUMATIC FEVER

• Non suppurative complications of Group A streptococcal pharyngitis
• Latent period of 1-3 weeks
• A delayed immune response caused by antibody cross reactivity that can involve the heart, joints, skin, and brain (basal ganglia)
• Serotypes M types (1,3,5,6,18,24)
INTRODUCTION

• Untreated Group-A beta hemolytic streptococcal infection –
  is the commonest antecedent event that precipitates an attack of ARF.
EPIDEMIOLOGY

• Prevalence RHD India - 0.5 %.

• About 50% of children with ARF will suffer from RHD
EPIDEMIOLOGY

- Sex – Both sex Equally Affected
- Depends on Individual Susceptibility
- Season – Winter
- Predisposing Factors –
  1. Low Socioeconomic status
  2. Overcrowding
  3. Poor Medical Care
DRAMATIC DECLINE IN DEVELOPED COUNTRIES

A - Antibiotic coverage has increased
B - Better housing
C - Conditions (economic & health) have improved
D - Decreased bacterial virulence
E - Easy access to medical care
Jones criteria (updated in 1992)

5- Major
4- Minor

2 majors or
1 major & 2 minors
with
Evidence of (microbiologic or serologic) of recent Group A beta hemolytic streptococcal infection
“Not meant as a substitute for Judgement by clinician. Intended guidelines to restrict the diagnosis to an acceptable clinical group”
WHY JONES CRITERIA

There is no specific lab test in diagnosis of RF

Aim

• Avoid over diagnosis

• Minimize missing an opportunity for the safety net of secondary prophylaxis

• Avoid over diagnosis in ‘Judgement’
MAJORS

- Carditis
- Polyarthritis
- Erythema marginatum
- Subcutaneous nodules
- Chorea

Mnemonic

$C^2ASE$

- C – Carditis
- C – Chorea
- A – Arthritis
- S – Subcutaneous nodules
- E – Erythema marginatum
MINORS

Clinical features
• Fever
• Arthralgia (in the absence of polyarthritis)

Laboratory features
• Elevated acute phase reactants
  – Raised ESR, Raised CRP
• Prolonged PR interval
Evidence of (microbiologic or serologic) of recent Group A beta hemolytic streptococcal infection (Essential criteria)

• Raised or Rising ASO Titers
• Positive Throat Culture for GAS
• Rapid streptococcal antigen test
• Anti-DNase B, anti-hyaluronidase
• H/o Recent Scarlet Fever
3 Circumstances – Where ARF diagnosed without strict adherence to Jones criteria

• Indolent carditis may be sole manifestation
• Chorea may be the sole manifestation
• ARF Recurrence may not fulfill the Jones criteria
ARTHRITIS

- Most Common – 30 to 50%
- Large joint involvement
- Migratory Polyarthritis
- Dramatic response to Aspirin
- Sacroiliac, Temporo-mandibular and Cervical joints not involved
- **No** Permanent Sequelae
- Resolves in Six weeks
CARDITIS PRESENTATION

• **Tachycardia** out of proportion to fever
• Sleeping pulse rate raised.
CARDITIS

- 50 -60%
- Usually **Pancarditis**
- Pericarditis never occurs in isolation
- Variable Severity
- Clinical Signs
  - Pericarditis – Effusion, Rub, Pain
  - Myocarditis – Tachycardia, Arrhythmia, cardiomegaly, failure
  - Endocarditis – Murmurs
MURMUR

• High pitched apical **holosystolic murmur** radiating to axilla
  
  – – Mitral regurgitation.

• An apical mid diastolic murmur.

• High pitched **decrescendo diastolic** murmur - upper sternal border
  
  – - Aortic regurgitation.
CARDITIS SEQUELAE (CHRONIC)

Mitral insufficiency

• Some loss of valvular substance

• Shortening & thickening of Chordae tendinae
Mitral stenosis

- Takes longer duration to develop after an attack of ARF
- Fibrosis of mitral ring, commissural adhesions
- Contracture of the valve leaflets, chordae & papillary muscles
- Opening snap, low pitched, rumbling mitral diastolic murmur with pre systolic accentuation ending in loud first sound
Aortic insufficiency

- Sclerosis of aortic valve- distortion & retraction of the cup
- Characteristic early diastolic murmur
- An apical pre systolic murmur (Austin flint)
SYDENHAM’S CHOREA

• 10 – 15 % of patients
• Usually delayed/often sole manifestation of ARF
• Involuntary movements of the face and limbs,
  – Muscle weakness
  – Disturbances of speech and gait,
  – Poor scholastic performance
SYDENHAM’S CHOREA

• Milkmaid's grip
  – irregular contractions of the muscles of the hands while squeezing the examiner's fingers
• Spooning and pronation of the hands when the patient's arms are extended *(St. Vitus Dance)*
• Wormian movements of the tongue upon protrusion *(Jack in the Box)*
• Handwriting to evaluate fine motor movements
SYDENHAM’S CHOREA

• Facial grimacing
• Emotional liability
• Exacerbated by stress
• Disappear at sleep
• Rarely leads to permanent neurological sequelae
CHOREA

• Seen more in females

• Minimum 3 months after sore throat
ERYTHEMA MARGINATUM

• Occurs < 10% of Patients
• Macular non pruritic rash
• Serpiginous border, raised edges, central clearing
• Most common on trunk
• Never seen on face
• Evanescent, warmth accentuates lesion
ERYTHEMA MARGINATUM
Erythema marginatum on the trunk, showing erythematous lesions with pale centers and rounded or serpiginous margins
Closer view of erythema marginatum in the same patient
SUBCUTANEOUS NODULES

• Seen in around 5 %
• Small, pea sized, 0.5 to 2 cms in diameter
• Firm, mobile, PAINLESS
• Seen over the extensor surface of WRIST, ELBOW, SPINE
• Usually seen in individuals with long standing carditis
SUBCUTANEOUS NODULE
Subcutaneous nodule on the extensor surface of elbow of a patient with acute rheumatic fever
<table>
<thead>
<tr>
<th>Condition</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Polyarthritis</td>
<td>50 - 75 %</td>
</tr>
<tr>
<td>SC Nodule</td>
<td>&gt; 95 %</td>
</tr>
<tr>
<td>Chorea</td>
<td>60 - 75 %</td>
</tr>
</tbody>
</table>

Half are inapparent
DIFFERENTIAL DIAGNOSIS

• Arthritis- Rheumatoid arthritis (JRA)
  - SLE
  - Reactive arthritis –
    Shigella, Salmenolosis, Yersenia
  - Lyme’s disease
  - Chikungunya

• Carditis- viral myocarditis, & Pericarditis
  Infective endocarditis
  Congenital heart lesions

• Chorea - Huntington chorea
  Wilson disease
  Tics
INVESTIGATIONS

• Tests reflecting tissue inflammation
  TC, DC, ESR, CRP (acute phase reactants)

• Tests to Prove STREP Infection
  – ASO Titer, AntiDNAase, Streptozyme, Anti Streptokinase
  – Throat Culture
INVESTIGATION

• X ray chest
• ECG
• ECHO
• Doppler
• Blood culture
• Catheterization studies
Chest radiograph of an 8 year old patient with acute carditis before treatment
Chest radiograph of an 8 year old patient with acute carditis after treatment
• ECG - Prolonged PR interval,

  2nd or 3rd degree blocks

  ST depression

  T inversion

• 2D Echo Cardiography - valve edema

  Mitral regurgitation, LA & LV dilatation,

  Pericardial effusion

  Decreased contractility
MANAGEMENT

• Bed rest
• Eradication of Streptococci
• Anti inflammatory therapy
• Treatment of CCF
• Treatment of Chorea
• Prevention of Recurrences
• Surgical – Acute and Chronic
ARTHRITEIS

• Bed rest
• Aspirin only for 4-6 Weeks
• Local measures
CARDITIS

• Carditis alone - ASPIRIN Only

• Carditis with mild Cardiomegaly - ASPIRIN Only

• Carditis in failure - ASPIRIN with STEROIDS and decongestive measures

• Bedrest

• Low sodium diet
SYDENHAM’S CHOREA - TREATMENT

• Anti inflammatory agents usually not required
• Phenobarbitol 15-30 mg tid or qid oral - drug of choice
• Haloperidol 0.01- 0.03mg/kg/24hrs bd oral
• Chlorpromazine 0.5mg/kg every 4-6 hrs. oral
PREVENTION IN RF

**Primordial** Preventing Strep Throat - vaccine ?

**Primary** Treating Strep Throat infection

**Secondary** Preventing Rheumatic recurrence by chemoprophylaxis

**Tertiary** Treating RHD
PRIMARY PROPHYLAXIS

Treating Streptococcal sore throat with Antibiotics

1. Oral Penicillin
2. Procaine Penicillin IM
3. Benzathine Penicillin
4. Oral Erythromycin
PRIMARY PROPHYLAXIS

Vulnerable children from 5 to 15 yrs. with pharyngitis

Oral
- Penicillin 250-500mg bd/tds 10 days
- Erythromycin 20-40 mg/kg/day tds/qid- 10 days
- First generation Cephalosporin- 10 days
- Azithromycin 12mg/kg/day single dose – 5 days max-500mg/day

Parenteral
- < than 27kg single dose IM Benzathine penicillin 6,00,000 U
- > than 27kg single dose IM Benzathine penicillin 1,20,0000 U

Therapy instituted before 9th day of symptoms of acute Pharyngitis
SECONDARY PROPHYLAXIS

• To prevent recurrences
  – Benzathine Penicillin once in 3 weeks IM
  – Oral Penicillin daily
  – Erythromycin daily
  – Sulfadiazine daily
SECONDARY PROPHYLAXIS

<table>
<thead>
<tr>
<th>Route</th>
<th>Antibiotic</th>
<th>Dose</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>IM</td>
<td>Benzathine Penicillin</td>
<td>1200000 U</td>
<td>Every 3rd wk.</td>
</tr>
<tr>
<td>Oral</td>
<td>Penicillin V</td>
<td>250mg</td>
<td>BD daily</td>
</tr>
<tr>
<td>Oral</td>
<td>Erythromycin</td>
<td>250mg</td>
<td>BD daily</td>
</tr>
<tr>
<td>Oral</td>
<td>Sulphadiazine</td>
<td>500mg to 1000mg</td>
<td>BD daily</td>
</tr>
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HOW LONG TO GIVE

RF; No Carditis 5 years from last Episode or till 21 years

RF; Carditis;

No residual RHD 10 years from last episode; or till 25 years

RF Carditis; RHD 10 years from last episode; or till 40 years / lifelong
NEW MODES OF TREATMENT

? IVIG

?? Valproate for chorea

? Anti- cytokines - adjuvants

?? Other NSAIDS
IS IT RHEUMATIC FEVER?

If you experience any of these symptoms, SEE YOUR DOCTOR!!

- Sore Throat
- Skin Sores
- Swollen Joints
THANKYOU