RHEUMATIC FEVER & RHEUMATIC HEART DISEASE
ACUTE RHEUMATIC FEVER

- Autoimmune consequence of infection (pharyngeal infection not the skin infection) with Group A beta haemolytic streptococcal infection

- Generalized inflammatory response affecting brains, joints, skin, subcutaneous tissues & the heart

- Modified Duckett-Jones criteria form the basis of the diagnosis of the condition
ACUTE RHEUMATIC FEVER

Supporting evidences:

- About 66% of the patients with an acute episode of rheumatic fever have a history of an upper respiratory tract infection several weeks before.

- The peak age (6-15 yrs) & seasonal incidence of acute rheumatic fever closely parallel those of GABHS infections.
Features suggestive of GABHS infection

- Patient 5 to 15 years of age
- Presentation in winter or early spring
- Fever, Headache
- Sudden onset of sore throat
- Nausea, vomiting & abdominal pain; Pain with swallowing
- Beefy, swollen, red uvula
- Soft palate petechiae (“doughnut lesions”)
- Tender, enlarged anterior cervical nodes
- Tonsillopharyngeal erythema & exudates
ACUTE RHEUMATIC FEVER

Redness & swelling of throat & tonsils;

Beefy, swollen, red uvula; Soft palate petechiae ("doughnut lesions")

Tonsillopharyngeal erythema & exudates

Sore throat: fever, white draining patches on the throat & swollen or tender lymph glands in the neck
ACUTE RHEUMATIC FEVER

Supporting evidences:

- Patients with acute rheumatic fever almost always have **serologic evidence** of a recent GABHS infection.
- Their **antibody titers** are usually considerably **higher** than those in patients with GABHS infections **without** acute rheumatic fever.
- Antimicrobial therapy against GABHS: prevents initial episodes of acute rheumatic fever &
- Long-term, continuous prophylaxis: prevents recurrences of acute rheumatic fever.
ACUTE RHEUMATIC FEVER

**Predisposing factors:**

- Family history of rheumatic fever
- Low socioeconomic status (poverty, poor hygiene, medical deprivation)
- Age: 6-15 years
EPIDEMIOLOGY

- Prevalence of Acute rheumatic fever & RHD: 0.67/1000 to 11/1000 children

- The INCIDENCE of rheumatic fever varies from 0.2 to 0.75/1000/ year in schoolchildren 5-15 years of age (2001 Govt. Census)
CLINICAL MANIFESTATIONS

- No pathognomonic clinical or laboratory finding for acute rheumatic fever
- Duckett Jones in 1944 proposed guidelines to aid in diagnosis & to limit overdiagnosis
- Jones criteria for the diagnosis of acute rheumatic fever: 2 major criteria or 1 major & 2 minor criteria along with the absolute requirement
- There are 5 major and 4 minor criteria & an absolute requirement for evidence (microbiologic or serologic) of recent GABHS infection
# Diagnosis

<table>
<thead>
<tr>
<th>Major Manifestations</th>
<th>Minor Manifestations</th>
<th>Supporting Evidence of Antecedent Group A Streptococcal Infection</th>
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<tbody>
<tr>
<td>Carditis</td>
<td>Clinical features:</td>
<td><strong>Elevated or increasing streptococcal antibody titer</strong></td>
</tr>
<tr>
<td></td>
<td>Arthralgia</td>
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<td></td>
<td>Fever</td>
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<tr>
<td>Polyarthritis</td>
<td>Laboratory features:</td>
<td>History of (&lt;45 days)</td>
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<tr>
<td></td>
<td></td>
<td>-Positive throat culture or rapid streptococcal antigen test or</td>
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<td></td>
<td></td>
<td>streptococcal sore throat or scarlet fever)</td>
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<tr>
<td>Erythema marginatum</td>
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<tr>
<td>Subcutaneous nodules</td>
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</tr>
<tr>
<td>Chorea</td>
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</tbody>
</table>
ARF & RHD

Valvulitis: whoosh swish gurgle etc.

Pericarditis: rrrub rrrub

Myocarditis: LUB DUB gub

erythema marginatum
subcutaneous nodules

Arthritis /
arthralgias

Sydenham's chorea (St. Vitus's dance)

Aschoff Nodule

beta-hemolytic
streptococci from the throat

high se rate

sterile vegetations

requirement

First episode or Recurrence without established heart disease: 2 major criteria or 1 major & 2 minor criteria & the absolute requirement

Recurrence with established heart disease: 2 minor criteria and the absolute requirement

established
MAJOR MANIFESTATIONS
Migratory Polyarthritis

- Most common (75%)
- Involves larger joints: the knees, ankles, wrists & elbows
- Rheumatic joints: hot, red, swollen & exquisitely tender (friction of bedclothes is uncomfortable)
- The pain can precede & can appear to be disproportionate to the other findings
Migratory Polyarthritis

- The joint involvement is characteristically *migratory* in nature
- *Monoarticular arthritis* is unusual unless anti-inflammatory therapy is initiated prematurely, aborting the progression of the migratory polyarthritis
If a child with fever and arthritis is suspected of having acute rheumatic fever: **withhold** salicylates & observe for migratory progression

A dramatic response to even small doses of salicylates is another characteristic feature of the arthritis

Rheumatic arthritis is typically **not deforming**
Migratory Polyarthritis

- Arthritis; earliest manifestation of acute rheumatic fever
- Correlate temporally with peak antistreptococcal antibody titers
- An inverse relationship between the severity of arthritis & the severity of cardiac involvement
Carditis

- Carditis & chronic rheumatic heart disease: **most serious manifestations** of acute rheumatic fever
- Account for essentially all of the associated morbidity and mortality
- Occurs in 50% of patients
- Rheumatic carditis: **pancarditis** with active inflammation of myocardium, pericardium & endocardium
- **Acute rheumatic carditis**: tachycardia out of proportion to fever & cardiac murmurs, with or without evidence of myocardial or pericardial involvement
Carditis

- Consists of either isolated mitral valvular disease or combined aortic & mitral valvular disease
- Valvular insufficiency: characteristic of both acute & convalescent stages of acute rheumatic fever
- Mitral regurgitation: a high-pitched apical holosystolic murmur radiating to the axilla
- In patients with significant mitral regurgitation-associated with an apical mid-diastolic murmur of relative mitral stenosis
- Aortic insufficiency: a high-pitched decrescendo diastolic murmur at the upper left sternal border
Valvular stenosis: appears several years or even decades after the acute illness.

However, in developing countries where acute rheumatic fever often occurs at a earlier age, mitral stenosis & aortic stenosis may develop in young children.

*Moderate to severe rheumatic carditis: cardiomegaly & congestive heart failure with hepatomegaly & peripheral & pulmonary edema*

Myocarditis &/or pericarditis *without evidence* of endocarditis: rarely due to rheumatic heart disease.
Echocardiographic findings: pericardial effusion, decreased ventricular contractility & aortic &/or mitral regurgitation

The major consequence of acute rheumatic carditis is chronic, progressive valvular disease
During an episode of ARF, valve changes can be minor and are still able to regress.

After recurrent episodes of ARF, thickening of subvalvar apparatus, chordal thickening and shortening and progression to permanent valve damage is evident.
Chorea

- St. Vitus’ dance
- Sydenham chorea: 10-15% of patients with acute rheumatic fever
- Often in prepubertal girls (8-12 yrs)
- A long latency period (1-6 mo) between streptococcal pharyngitis & the onset of chorea
- Neuropsychiatric disorder
- Neurologic signs: choreic movement & hypotonia
- Psychiatric signs: emotional lability, hyperactivity, separation anxiety, obsessions & compulsions
Chorea

- Begins with emotional lability & personality changes (poor school performance)
- Replace in 1-4 weeks by characteristic spontaneous, purposeless movement of chorea (lasts 4-8 months) followed by motor weakness
- Exacerbation by stress & disappearing with sleep are characteristic
- Elevated titers of “antineuronal antibodies” against basal ganglion tissues have been found in over 90% of patients
Clinical maneuvers to elicit features of chorea include:

1. Demonstration of **milkmaid's grip** (irregular contractions of the muscles of the hands while squeezing the examiner's fingers)
2. Spooning and pronation of the hands when the patient's arms are extended
3. Wormian darting movements of the tongue upon protrusion
4. Examination of handwriting to evaluate fine motor movements
Chorea

- Diagnosis: based on clinical findings with supportive evidence of GABHS antibodies
- In patients with a long latent period: antibody levels may have declined to normal
- SUBCLINICAL CARDITIS - 30%
- Although the acute illness is distressing, **chorea rarely, if ever, leads to permanent neurologic sequelae**
Erythema Marginatum

- A rare (<3% of patients with acute rheumatic fever) but characteristic rash of acute rheumatic fever
- It consists of erythematous, serpiginous, macular lesions with pale centers that are not pruritic
- It occurs primarily on the trunk & extremities, not on the face & it can be accentuated by warming the skin
Subcutaneous Nodules

- A rare (≤1% of patients with acute rheumatic fever) finding
- Consist of firm nodules approximately 1 cm in diameter along the extensor surfaces of tendons near bony prominences
- A correlation between the presence of these nodules & significant rheumatic heart disease
MINOR MANIFESTATIONS
Clinical:
- 1. Arthralgia (in the absence of polyarthritis as a major criterion)
- 2. Fever (typically temperature ≥102°F & occurring early in the course of illness)

Laboratory minor manifestations:
- 1. Elevated acute-phase reactants (C-reactive protein, erythrocyte sedimentation rate, polymorphonuclear leukocytosis)
- 2. Prolonged PR interval on electrocardiogram (1st degree heart block)
An absolute requirement for the diagnosis of acute rheumatic fever is supporting evidence of a recent GABHS infection.
Recent Group A Streptococcus infection

- Hallmarks of GAS sore throat:
  - High fever, tender anterior cervical lymph nodes
  - Close contact with infected person
  - Strawberry tongue, petechiae on palate
  - Excoriated nares (crusted lesions) in infants
  - Tonsillar exudates in older children
  - Abdominal pain

**GOLD STANDARD: POSITIVE THROAT CULTURE**
Recent Group A Streptococcus Infection

- Acute rheumatic fever typically develops 2-4 wk after an acute episode of GABHS pharyngitis at a time when clinical findings of pharyngitis are no longer present & only 10-20% of the throat culture or rapid streptococcal antigen test results are positive

- Therefore, evidence of an antecedent GABHS infection is usually based on elevated or increasing serum antistreptococcal antibody titers
Recent Group A Streptococcus infection

1. ASO titre:
   - well standardized
   - elevated in 80% of patients with ARF
   - ASO titre of 333 Todd unit in children & 250 Todd unit in adults are considered elevated

2. Antideoxyribonuclease B titre:
   - ≥240 Todd unit in children & ≥120 Todd unit in adults
3. Slide agglutination test (Streptozyme):
   - Detect antibodies against 5 different GABHS antigens
   - Rapidly, relatively simple to perform & widely available
   - Less standardized & less reproducible than other tests and should not be used as a diagnostic test for evidence of an antecedent GAS infection
Recent Group A Streptococcus infection

- Single antibody measured: 80-85% of patients have an elevated titer
- If 3 different antibodies (antistreptolysin O, anti-DNase B, antihyaluronidase) measured: 95-100% have an elevation
- Therefore in suspected ARF clinically: perform multiple antibody tests
- Diagnosis of ARF should not be made in patients with elevated or increasing streptococcal antibody titers who do not fulfill the Jones criteria
- True for younger, school-aged children having GABHS pyoderma or GABHS pharyngitis
## DIFFERENTIAL DIAGNOSIS

### ARTHRITIS

<table>
<thead>
<tr>
<th>Condition</th>
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<tbody>
<tr>
<td>Rheumatoid arthritis</td>
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<tr>
<td>Reactive arthritis <em>(Shigella, Salmonella, Yersinia)</em></td>
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<tr>
<td>Serum sickness</td>
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<tr>
<td>Sickle cell disease</td>
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<tr>
<td>Malignancy</td>
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<tr>
<td>Systemic lupus erythematosis</td>
</tr>
<tr>
<td>Lyme disease <em>(Borreliia burgdorferi)</em></td>
</tr>
<tr>
<td>Gonococcal infection <em>(N.gnorrhoeae)</em></td>
</tr>
</tbody>
</table>
# Differential Diagnosis

<table>
<thead>
<tr>
<th>Carditis</th>
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<tbody>
<tr>
<td>Viral myocarditis</td>
</tr>
<tr>
<td>Viral pericarditis</td>
</tr>
<tr>
<td>Infective endocarditis</td>
</tr>
<tr>
<td>Kawasaki disease</td>
</tr>
<tr>
<td>Congenital heart disease</td>
</tr>
<tr>
<td>Mitral valve prolapse</td>
</tr>
<tr>
<td>Innocent murmurs</td>
</tr>
</tbody>
</table>
## Differential Diagnosis

<table>
<thead>
<tr>
<th>CHOREA</th>
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<tbody>
<tr>
<td>Huntington chorea</td>
</tr>
<tr>
<td>Wilson disease</td>
</tr>
<tr>
<td>Systemic lupus erythematosus</td>
</tr>
<tr>
<td>Cerebral palsy</td>
</tr>
<tr>
<td>Tics</td>
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<tr>
<td>Hyperactivity</td>
</tr>
</tbody>
</table>
DIFFERENTIAL DIAGNOSIS

- Patients with infective endocarditis: present with both joint and cardiac manifestations.

- These patients can usually be distinguished from patients with acute rheumatic fever by blood cultures & the presence of associated findings (hematuria, splenomegaly, splinter hemorrhages).
- **Bed rest**

- **Antibiotic Therapy:**
  - 10 days of orally administered *penicillin* or *erythromycin* or a single intramuscular injection of *benzathine penicillin* to eradicate *GABHS* from the upper respiratory tract
  - Afterwards, the patient should be started on long-term antibiotic prophylaxis
TREATMENT

- **Anti-inflammatory Therapy:**
  - Anti-inflammatory agents (salicylates, corticosteroids) should be withheld if arthralgia or atypical arthritis is the only clinical manifestation of presumed acute rheumatic fever.
  - Acetaminophen can be used.
  - Patients with typical migratory polyarthritis & with carditis without cardiomegaly or congestive heart failure:
    - Treatment with oral salicylates, 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 4-8 wk.
Patients with carditis & cardiomegaly or congestive heart failure:

- treatment with corticosteroids
- Prednisone 2 mg/kg/day in 4 divided doses for 2-6 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 to complete 12 wk of therapy
Supportive therapies for patients with moderate to severe carditis include digoxin, fluid & salt restriction, diuretics & oxygen.

The cardiac toxicity of digoxin is enhanced with myocarditis.
Sydenham Chorea

- Occurs after the resolution of the acute phase of the disease
- Anti-inflammatory agents are usually not indicated
- Sedatives: phenobarbital (16-32 mg every 6-8 hr PO) is the drug of choice
- If phenobarbital is ineffective, then haloperidol (0.01-0.03 mg/kg/24 hr divided bid PO) or chlorpromazine (0.5 mg/kg every 4-6 hr PO) should be initiated
- Long-term antibiotic prophylaxis
PREVENTION

PRIMARY-10 days course of penicillin therapy; about 30% of patients with acute rheumatic fever do not recall a preceding episode of pharyngitis

SECONDARY-Secondary prevention is directed at preventing acute GABHS pharyngitis in patients at substantial risk of recurrent acute rheumatic fever
SECONDARY PREVENTION

Who should receive prophylaxis?

Patients with documented history of rheumatic fever, including those with isolated chorea & those without evidence of rheumatic heart disease MUST receive prophylaxis.
## SECONDARY PREVENTION

- **For how long?**

<table>
<thead>
<tr>
<th>CATEGORY</th>
<th>DURATION</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rheumatic fever <strong>without carditis</strong></td>
<td>At least for 5 yr or until age 21 year, whichever is longer</td>
</tr>
<tr>
<td>Rheumatic fever <strong>with carditis but without residual heart disease</strong></td>
<td>At least for 10 yr or well into adulthood, whichever is longer</td>
</tr>
<tr>
<td>Rheumatic fever <strong>with carditis &amp; residual heart disease</strong></td>
<td>At least 10 yr since last episode &amp; at least until age 40 yr; sometime lifelong</td>
</tr>
</tbody>
</table>
What method of prophylaxis should be used?

<table>
<thead>
<tr>
<th>DRUG</th>
<th>DOSE</th>
<th>ROUTE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Penicillin G benzathine</td>
<td>600,000 U for children, ≤27 kg; 1.2 million U for children &gt;27 kg, every 3 wk</td>
<td>Intramuscular</td>
</tr>
<tr>
<td>Penicillin V</td>
<td>250 mg, twice a day</td>
<td>Oral</td>
</tr>
<tr>
<td>Sulfadiazine or sulfisoxazole</td>
<td>0.5 g, once a day for patients ≤60 lb; 1.0 g, once a day for patients &gt;60 lb</td>
<td>Oral</td>
</tr>
</tbody>
</table>

For people who are allergic to penicillin and sulfonamide drugs

| Macrolide or azalide        | Variable                                                            | Oral               |
RHEUMATIC HEART DISEASE

- Rheumatic involvement of the valves & endocardium
- The valvular lesions begin as small **verrucae** composed of fibrin and blood cells along the borders of one or more of the heart valves
- The **mitral valve** is affected most often, followed in frequency by the **aortic valve**; **right-sided** heart manifestations are rare
- At the end of inflammation: verrucae disappear & leave scar tissue
- Repeated attacks of rheumatic fever: new verrucae form near the previous ones & the mural endocardium & chordae tendineae become involved
MITRAL INSUFFICIENCY

Backflow of blood from the LV to the LA during systole
MITRAL INSUFFICIENCY

**Pathophysiology:**

- **Loss of valvular substance** & shortening & thickening of the chordae tendineae
- Because of the high volume load & inflammatory process, the left ventricle becomes enlarged
- The **left atrium dilates** as blood regurgitates into this chamber
- **Increased left atrial pressure results in pulmonary congestion & symptoms of left-sided heart failure**
MITRAL INSUFFICIENCY

Pathophysiology:

- Spontaneous improvement usually occurs with time, even in patients with severe MR at the onset.
- More than half of patients with acute mitral insufficiency no longer have the mitral murmur 1 yr later.
- With severe chronic MR, pulmonary arterial pressure becomes elevated, the right ventricle & atrium become enlarged, & right-sided heart failure subsequently develops.
MITRAL INSUFFICIENCY

Clinical manifestations:

- Exertion Dyspnea (exercise intolerance), fatigue
- Mild disease: NO signs of heart failure
- Severe mitral insufficiency: signs of left sided heart failure
- The heart is enlarged, with a forcible & hyperkinetic apical left ventricular impulse & often an apical systolic thrill
- Soft S1
MITRAL INSUFFICIENCY

Clinical manifestations:

- The 2nd heart sound may be accentuated if pulmonary hypertension is present.
- A 3rd heart sound is generally prominent.
- A holosystolic murmur is heard at the apex with radiation to the axilla.
- A short mid-diastolic rumbling murmur is caused by increased blood flow across the mitral valve as a result of the insufficiency.
MITRAL INSUFFICIENCY

**Imaging studies:**
- **ECG:** prominent bifid P waves, signs of left ventricular hypertrophy & associated right ventricular hypertrophy if pulmonary hypertension is present
- **X-rays:** prominence of the left atrium & ventricle; congestion of perihilar vessels, a sign of pulmonary venous hypertension
- **2 D ECHO:** enlargement of the left atrium & ventricle & Doppler studies demonstrate the severity of the mitral regurgitation
MITRAL INSUFFICIENCY

Complications:
- cardiac failure
- chronic mitral insufficiency - right ventricular failure
- atrial and ventricular arrhythmias
MITRAL INSUFFICIENCY

Management:

- **Medical:**
  - *Prophylaxis* against recurrences of rheumatic fever
  - Treatment of heart failure, arrhythmias and infective endocarditis
  - Afterload-reducing agents (ACE inhibitors or angiotensin receptor blockers):
    - reduce the regurgitant volume & preserve left ventricular function
Management:

- **Surgical:**
  - For patients who despite adequate medical therapy have persistent heart failure, dyspnea with moderate activity & progressive cardiomegaly, often with pulmonary hypertension
  - Valve repair surgery preferred over valve replacement
MITRAL STENOSIS

- Obstruction of LV inflow that prevents proper filling during diastole
- Normal MV Area: 4-6 cm²
- Transmitral gradients & symptoms begin at areas less than 2 cm²
Pathophysiology:

- From fibrosis of the mitral ring, commissural adhesions & contracture of the valve leaflets, chordae & papillary muscles
- It takes 10 years or more for the lesion to become fully established
MITRAL STENOSIS

Pathophysiology:

- Significant mitral stenosis results in increased pressure, enlargement & hypertrophy of the left atrium, pulmonary venous hypertension, increased pulmonary vascular resistance & pulmonary hypertension.
- Right ventricular hypertrophy & right atrial dilatation ensue & are followed by right ventricular dilation, tricuspid regurgitation & clinical signs of right-sided heart failure.
MITRAL STENOSIS

Clinical manifestations:

- Correlation between symptoms & the severity of obstruction
- Patients with mild lesions: asymptomatic
- More severe degrees of obstruction: exercise intolerance & dyspnea
- Critical lesions: orthopnea, paroxysmal nocturnal dyspnea, & overt pulmonary edema, as well as atrial arrhythmias
MITRAL STENOSIS

Clinical manifestations:

- Pulmonary hypertension: right ventricular dilatation-functional tricuspid insufficiency, hepatomegaly, ascites & edema
- Hemoptysis: rupture of bronchial or pleurohilar veins or by pulmonary infarction
Clinical manifestations:

- Jugular venous pressure is increased in severe disease with heart failure
- Prominent "a" wave in jugular venous pulsations: Due to pulmonary hypertension & right ventricular hypertrophy
- Mild disease: heart size is normal, tapping apex
- Severe mitral stenosis: moderate cardiomegaly
- Cardiac enlargement massive: atrial fibrillation & heart failure
- A parasternal right ventricular lift is palpable when pulmonary pressure is high
MITRAL STENOSIS

Clinical manifestations:

- **Auscultatory findings:**
  - Loud 1st heart sound,
  - An opening snap of the mitral valve, and
  - A long, low-pitched, rumbling mitral diastolic murmur with presystolic accentuation at the apex
  - Murmur absent in patients with significant heart failure
Clinical manifestations:

- A holosystolic murmur secondary to tricuspid insufficiency
- Pulmonary hypertension: pulmonic component of the 2nd heart sound is accentuated
- An early diastolic murmur: associated AR or pulmonary valvular insufficiency secondary to pulmonary hypertension
MITRAL STENOSIS

Imaging studies:

- **ECG**: prominent & notched P waves & varying degrees of right ventricular hypertrophy, Atrial fibrillation
- **X-rays**: Left atrial enlargement & prominence of the pulmonary artery & right-sided heart chambers; calcifications may be noted in the region of the mitral valve
- Severe obstruction is associated with a redistribution of pulmonary blood flow so that the apices of the lung have greater perfusion (the reverse of normal)
MITRAL STENOSIS

Imaging studies:

- **2D ECHO**: thickening of the mitral valve, distinct narrowing of the mitral orifice during diastole and left atrial enlargement
- Doppler can estimate the transmitral pressure gradient
- Cardiac catheterization quantitates
  - Diastolic gradient across the mitral valve
  - Allows for the calculation of valve area
  - Assesses the degree of elevation of pulmonary arterial pressure
MITRAL STENOSIS

Management:
- Medical:
  - Mild & moderate MS: anticongestive measures (digoxin & diuretics)
  - Atrial fibrillation: digoxin; procainamide for conversion to sinus rhythm in hemodynamically stable patients
  - chronic AF warfarin
  - IE prophylaxis
  - percutaneous mitral balloon valvotomy: failure to thrive with repeated respiratory infections
**MITRAL STENOSIS**

**Management:**

- Surgical: indicated in patients with clinical signs & hemodynamic evidence of severe obstruction.
- or ANY SYMPTOMATIC Patient with NYHA Class III or IV Symptoms.
- or Asymptomatic moderate or severe MS with a pliable valve.
Management:

- Surgical valvotomy or balloon catheter mitral valvuloplasty
- Balloon valvuloplasty is indicated for symptomatic, stenotic, pliable, noncalcified valves of patients without atrial arrhythmias or thrombi
AORTIC INSUFFICIENCY

- Leakage of blood into LV during diastole due to ineffective coaptation of the aortic cusps
- Regurgitation of blood leads to volume overload with dilatation & hypertrophy of the left ventricle
AORTIC INSUFFICIENCY

Pathophysiology:
- Combined pressure AND volume overload
- Compensatory Mechanisms: LV dilation & LV hypertrophy
- Progressive dilation leads to heart failure
AORTIC INSUFFICIENCY

**Clinical manifestations:**

- Symptoms are unusual except in severe aortic insufficiency
- The large stroke volume & forceful left ventricular contractions result in **palpitations**
- **Sweating and heat intolerance** are related to excessive vasodilation
- Dyspnea on exertion can progress to orthopnea and pulmonary edema
- Nocturnal attacks with sweating, tachycardia, chest pain, & hypertension
AORTIC INSUFFICIENCY

Clinical manifestations:
- Wide pulse pressure with bounding peripheral pulses
- Systolic blood pressure elevated & diastolic pressure is lowered
- Severe aortic insufficiency: enlarged heart with a left ventricular apical heave
- Diastolic thrill unusual
- Murmur begins immediately with the 2nd heart sound & continues until late in diastole over the upper & midleft sternal border with radiation to the apex and upper right sternal border
AORTIC INSUFFICIENCY

Clinical manifestations:

- It has a high-pitched blowing quality & is easily audible in full expiration with the diaphragm of the stethoscope placed firmly on the chest & the patient leaning forward.

- An aortic systolic ejection murmur is frequent because of the increased stroke volume.

- An apical presystolic murmur (Austin Flint murmur) resembling MS is sometimes heard (due to the large regurgitant aortic flow in diastole preventing the mitral valve from opening fully).
**Auscultatory and peripheral findings in severe AR**

<table>
<thead>
<tr>
<th>Sign</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Austin Flint murmur</td>
<td>Low-pitched apical mid-diastolic rumble</td>
</tr>
<tr>
<td>Corrigan pulse</td>
<td>High-amplitude, abruptly collapsing pulse</td>
</tr>
<tr>
<td>Duroziez sign</td>
<td>To-and-fro bruit over femoral artery</td>
</tr>
<tr>
<td>Hill sign</td>
<td>$&gt;40$ mm Hg$_\Delta$ between popliteal and brachial pressures</td>
</tr>
<tr>
<td>Mayne sign</td>
<td>$&gt;15$ mm Hg drop in SBP with arm elevation pressure</td>
</tr>
<tr>
<td>Traube sign</td>
<td>Loud systolic “pistol shots” over the femoral artery</td>
</tr>
<tr>
<td>Quincke pulse</td>
<td>Exaggerated reddening and blanching of nail beds</td>
</tr>
<tr>
<td>Mueller sign</td>
<td>Visible pulsations of the uvula</td>
</tr>
<tr>
<td>de Musset sign</td>
<td>Visible bobbing of the head</td>
</tr>
</tbody>
</table>
AORTIC INSUFFICIENCY

Imaging studies:

- **ECG:** signs of left ventricular hypertrophy & strain with prominent P waves in severe cases
- **X-rays:** Enlargement of the left ventricle & aorta
Imaging studies:

- **2 D ECHO:**
  - A large left ventricle & diastolic mitral valve flutter or oscillation caused by regurgitant flow hitting the valve leaflets
- **Doppler studies** demonstrate the degree of aortic runoff into the left ventricle
- **Magnetic resonance angiography** can be useful in quantitating regurgitant volume
- **Cardiac catheterization** is necessary only when the echocardiographic data are equivocal
AORTIC INSUFFICIENCY

Management:
- Mild and moderate lesions are well tolerated. Unlike mitral insufficiency, aortic insufficiency does not regress.
- Medical:
  - Afterload reducers (ACE inhibitors or angiotensin receptor blockers)
  - Prophylaxis against recurrence of acute rheumatic fever
  - IE prophylaxis
AORTIC INSUFFICIENCY

Management:

- Surgical: Definitive Treatment
- Surgical intervention (valve replacement) should be carried out well in advance of the onset of heart failure, pulmonary edema, or angina, when signs of decreasing myocardial performance become evident as manifested by increasing left ventricular dimensions on the echocardiogram.
Management:

- Surgery is considered when early symptoms are present, ST-T wave changes are seen on the electrocardiogram, or evidence of decreasing left ventricular ejection fraction is noted.
- ANY Symptoms at rest.
- Asymptomatic treatment if: EF drops below 50% or LV becomes dilated.
TRICUSPID VALVE DISEASE

- Primary tricuspid involvement: rare
- Tricuspid insufficiency: secondary to right ventricular dilatation resulting from unrepaired left-sided lesions
- **Signs:** prominent pulsations of the jugular veins, systolic pulsations of the liver & a blowing holosystolic murmur at the lower left sternal border that increases in intensity during inspiration
- Signs of tricuspid insufficiency decrease or disappear when heart failure produced by the left-sided lesions is successfully treated
- Tricuspid valvuloplasty may be required in rare cases
PULMONARY VALVE DISEASE

- Pulmonary insufficiency usually occurs on a functional basis secondary to pulmonary hypertension & is a late finding with severe mitral stenosis.
- The murmur (Graham Steell murmur) is similar to that of aortic insufficiency, but peripheral arterial signs (bounding pulses) are absent.
- The correct diagnosis is confirmed by two-dimensional echocardiography and Doppler studies.
SUMMARY

- Rheumatic heart disease is the only truly preventable chronic heart condition
- **Primary prevention:** Penicillin for suspected strep sore throat
- **Secondary prevention:** Penicillin prophylaxis
Ensuring that patients understand their disease, are informed regarding their future and receive secondary prophylaxis

EDUCATION

✓ Health education is critical at all levels

✓ Lack of parental awareness of the causes and consequences of ARF/RHD is a key contributor to poor adherence amongst children on long-term prophylaxis
How can we reduce the pain associated with IM Penicillin?

- Use a 23-gauge needle- deeper is better
- Local pressure to area for 10 secs
- Warm syringe to room temperature
- First allow alcohol to dry or use ethylchloride spray
- Deliver injection very slowly(over 2-3mins)
- Distraction techniques
- Good rapport with the case, is a significant aid to injection comfort, compliance and understanding
- Use 0.5-1ml of 1% lignocaine. Reduces pain significantly and excellent for younger patients