Rheumatic Fever And RHD

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Lecture Outline

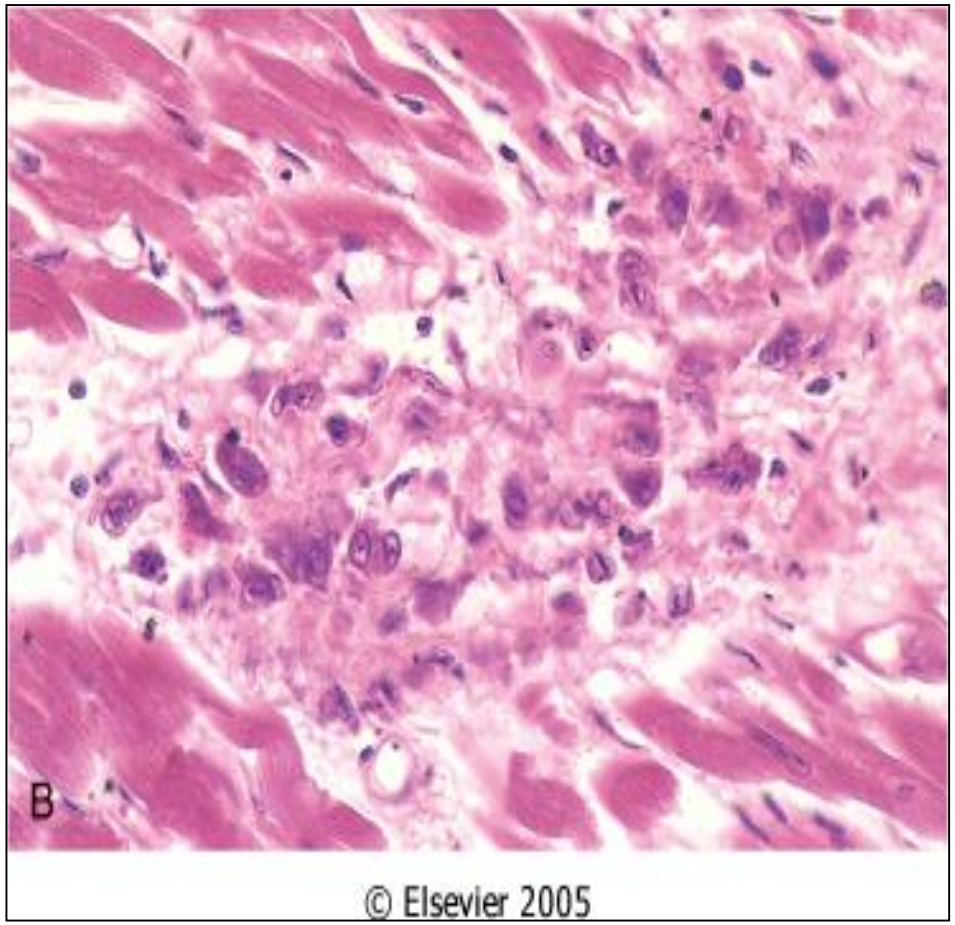
- What is ARF And RHD?
- Diagnosis
- Jones Criteria & 2015 revision
- Differential Diagnosis
- Investigations, Management
- Rheumatic Valvular Heart Disease
- Prevention

Rhuematic Fever

- Follows group A beta hemolytic streptococcal throat infection
- It represents a delayed immune response to infection with manifestations appearing after a period of 2-4 weeks
- Age 5-15 yrs
- A multisystem disease
- RHD is a long term complication og ARF
- Major effect on health is due to damage to heart valves

Pathologic Lesions

- Ashcoff nodules
- Fibrinoid degeneration of connective tissue, inflammatory cells



Global Burden of RHD

- A leading cause of CV morbidity & mortality in young people
- Total cases with RHD:20 Millions
- CHF:3Million,valve surgery required in 1Million
- Annual incidence of RF: 0.5 Million, nearly half develop carditis
- Estimated deaths from RHD: 250,000/YR
- Imposes a substantial burden on health care systems with limited budgets

Epidemiologic Background

- The incidence of RF and the prevalence of RHD has declined substantially in Europe, North America and other developed nations
- this decline has ben attributed to improved hygiene, reduced household crowding, and improved medical care

Epidemiologic Background

- The major burden is currently found in low and middle income countries (India, middle east), and in selected indigenous populations of certain developed countries (Australia and Newzealand).
- A disease of poverty and low socioeconomic status
- In underdeveloped countries RHD is the leading cause of CV death during the first five decades of life

Epidemiologic Background

- The incidence in indigenous population of Australia: 53-380 cases/100000 people/yr in 5-14 yrs age group.
- In Saudi Arabia: incidence 30 cases/100000 people/yr and prevalence 310/100000 people in 6-15 yrs age group
- Low risk population ARF incidence < 2/100000/yr (5-14 yrs) or all age prevalence of RHD <1/1000 population/yr

Diagnosis of ARF

- No single test to diagnose ARF
- The symptoms and signs are shared by many inflammatory and infectious diseases
- Accurate diagnosis is important
- Overdiagnosis will result in individuals receiving treatment unnecessarily
- Underdiagnosis may lead to recurrences of ARF causing further damage, the need for valve surgery, CHF and premature death

Diagnosis of ARF

- Diagnosis is primarily clinical and is based on a constellation of signs and symptoms, which were initially established as the Jones criteria
- In 1944 Dr. TD Jones published a set of guidelines for diagnosis of ARF “Jones Criteria”
- Subsequently Modified in 1965, 1984 and 1992by AHA
- Revised recently -2015 by AHA

1992 Modified Jones Criteria

Major criteria	Minor criteria
Migratory polyarthritis	Arthralgia
Carditis	Fever
Erythema marginatum	First degree heart block
Syndenham chorea	Elevated inflammatory markers (ESR, CRP)
Subcutaneous nodules	

Carditis

- Occurs in 50-70% of cases
- Only manifestation of ARF that leaves permanent damage
- May be subclinical
- Murmurs of MR or AR may occur in acute stage while mitral stenosis occurs in late stages
- Cardiomegaly and CHF may occur

Arthritis

- Common: present in 35-66%
- Earliest manifestation of ARF
- Large joints: The knees and ankles, shoulders, elbows
- “Migrating”, “Fleeting” polyarthritis
- Duration short < 1 week
- Rapid improvement with salicylates
- Does not progress to chronic disease

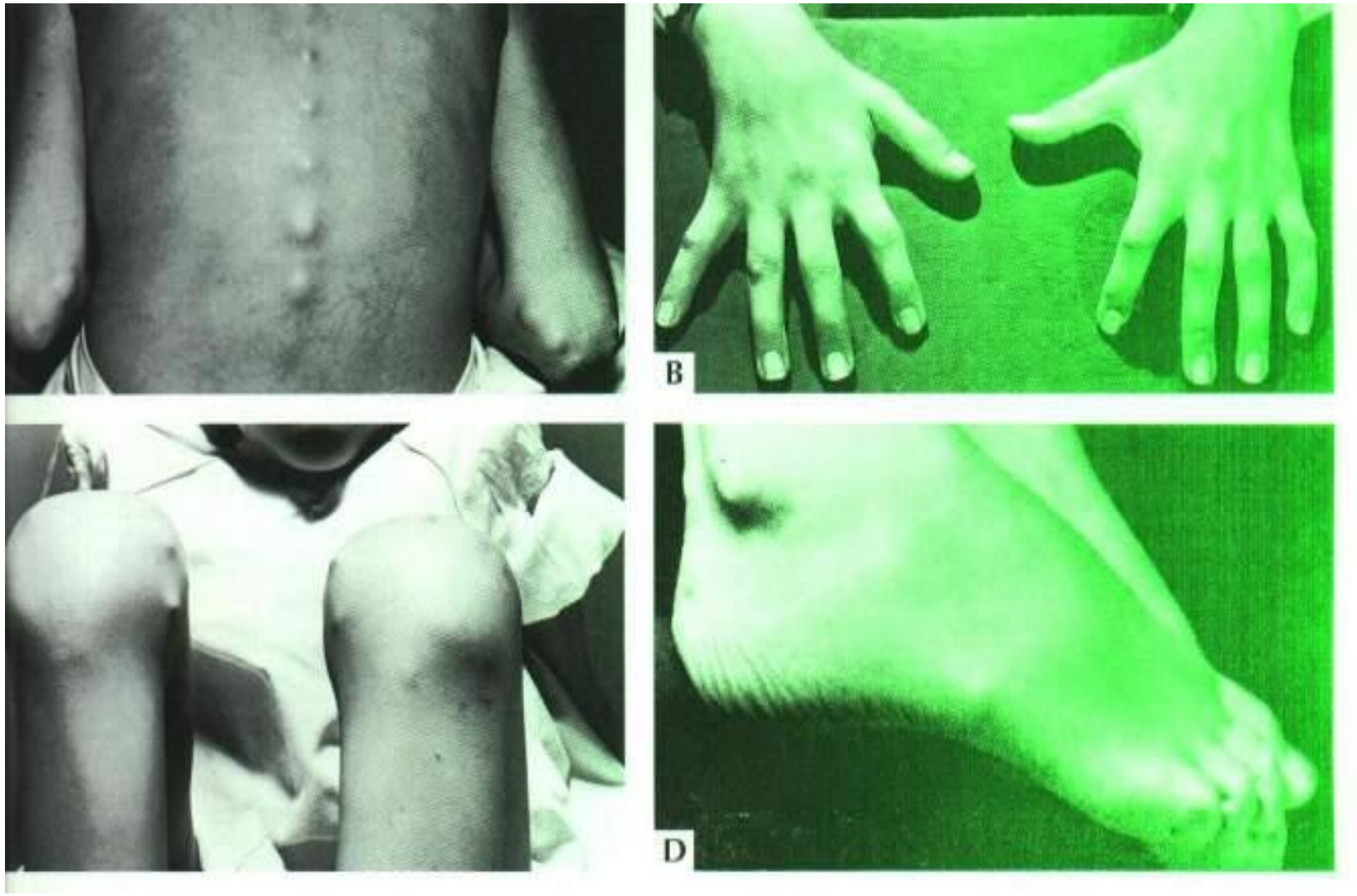
Sydenham Chorea

- Also known as Saint Vitus'dance
- Occur in 10-30%, extrapyramidal manifestation, female predominnce
- Abrupt Purposeless involuantry movements of muscles of face, neck, trunk, and limbs.
- Delayed manifestation of ARF -months
- Clinically manifest as-clumsiness, deterioration of handwriting,emotional lability or grimacing of face

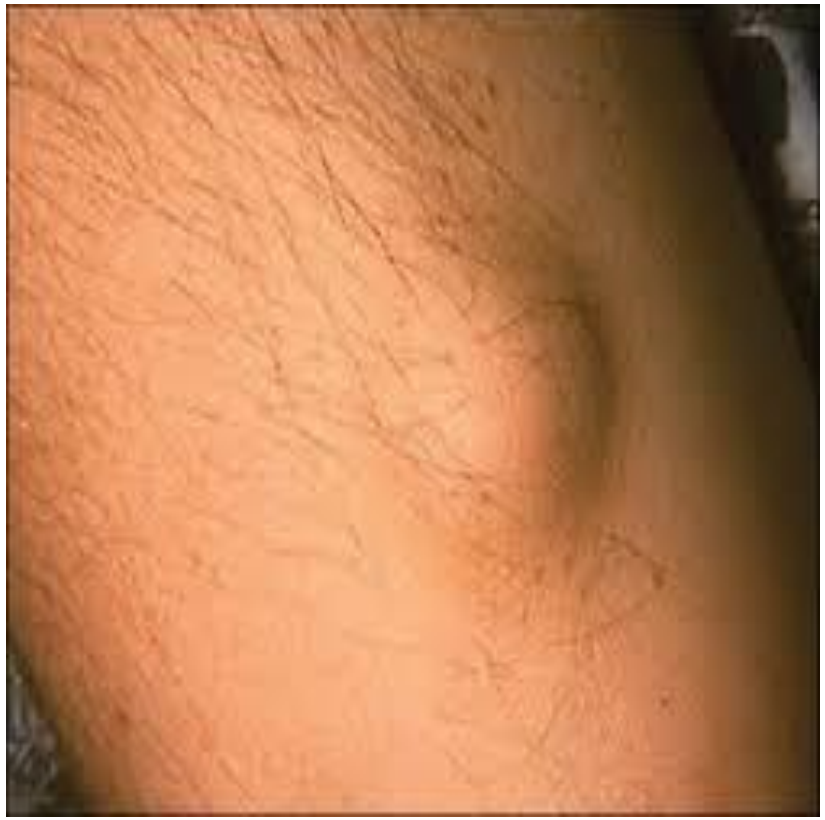
Subcutaneous Nodules

- Occur in 10%
- Usually 0.5 – 2 cm long
- Firm non-tender
- Occur over extensor surfaces of joints, on bony prominences, tendons, spine
- Short lived: last for few days
- Associated with severe carditis

Subcutaneous nodules



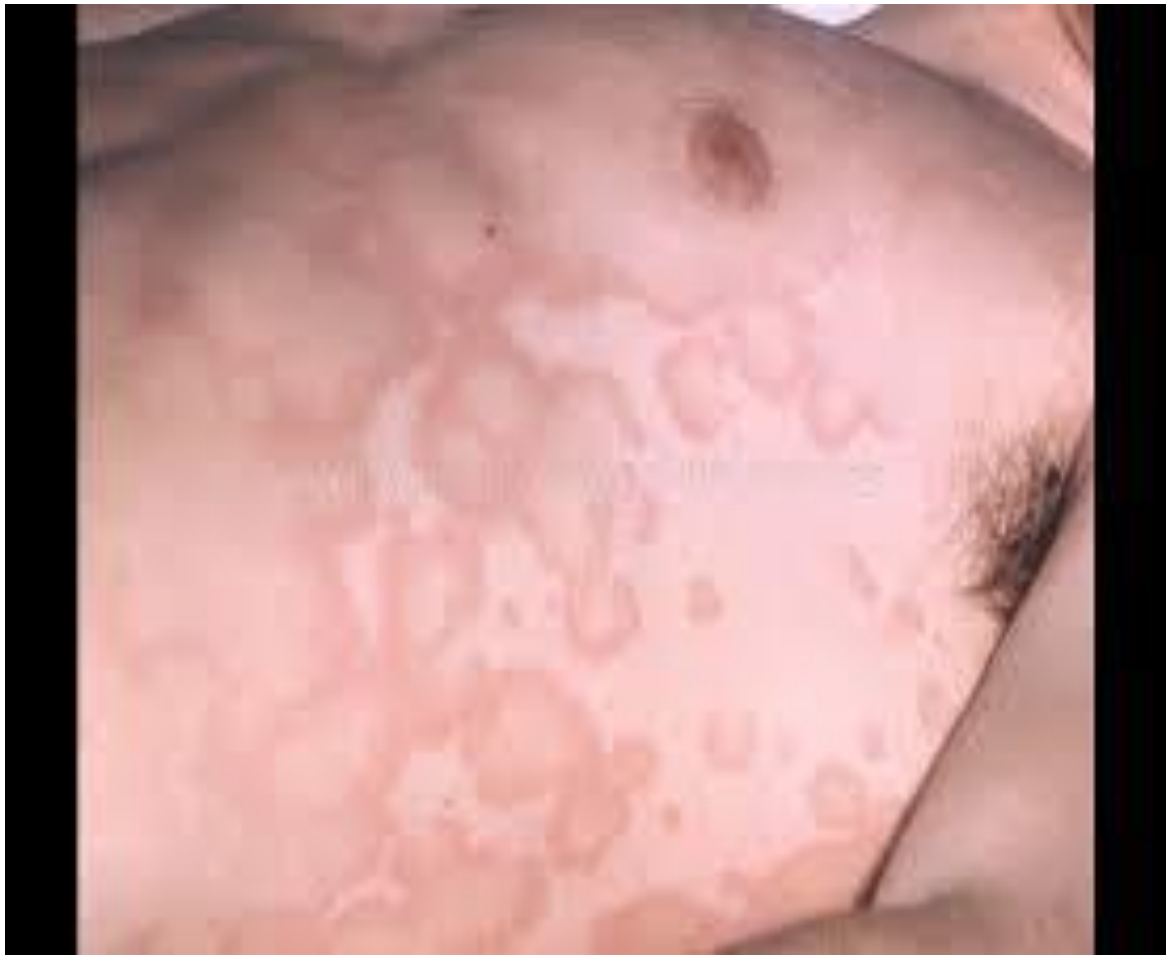
Subcutaneous Nodules



Erythema Marginatum

- Present in <6%
- Less common, but highly specific manifestation of ARF
- Reddish border, pale center, round or irregular serpiginous borders, non-pruritic, transient rash
- Occurs on trunk, abdomen or proximal limbs
- Associated with carditis

Erythrma Marignatum



2015 Revised Jones Criteria

Revision of the Jones Criteria for the Diagnosis of Acute Rheumatic Fever in the Era of Doppler Echocardiography

A Scientific Statement From the American Heart Association

Circulation. published online April 23, 2015;

2015 Revision of Jones Criteria

1. In accordance with the degree of prevalence of ARF/RHD in the population:

- low risk populations have been defined as those with ARF incidence < 2:100000 school-age children or all age prevalence of RHD of < 1:1000 population per year
- Children not from low risk population have been considered to be at moderate or high risk

2015 Revision of Jones Criteria

2. Advocated the use of Echocardiography in all cases of confirmed or suspected ARF or RHD, to diagnose valvulitis(subclinical carditis) and has been included as a major criterion to diagnose carditis

3. Aseptic monoarthritis has been included as a major criteria in moderate or high risk population

2015 Revision of Jones Criteria

4. Polyarthritis has been recognized as a major manifestation for moderate or high risk population

5. Fever >38.5 c, ESR >60 and or CRP > 3mg/dl for low risk population, and fever >38 and ESR >30 and or CRP > 3mg/dl for moderate or high risk population

Revised Jones Criteria-2015

A. 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
B. Major criteria	
Low-risk populations*	Moderate- and high-risk populations
Carditis† • Clinical and/or subclinical	Carditis • Clinical and/or subclinical
Arthritis • Polyarthritis only	Arthritis • Monoarthritis or polyarthritis • Polyarthralgia‡
Chorea	Chorea
Erythema marginatum	Erythema marginatum
Subcutaneous nodules	Subcutaneous nodules
C. Minor criteria	
Low-risk populations*	Moderate- and high-risk populations
Polyarthralgia	Monoarthralgia
Fever (≥38.5°C)	Fever (≥38°C)
ESR ≥60 mm in the first hour and/or CRP ≥3.0 mg/dL§	ESR ≥30 mm/h and/or CRP ≥3.0 mg/dL§
Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion)	Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion)

2015 Revised Jones Criteria

A firm diagnosis requires

1) 2 Major manifestations or 1 Major and 2 Minor manifestations

and

2) Evidence of a recent streptococcal infection.

2015 Revised Jones Criteria

Evidence of Preceding GAS Infection:

1) Increased or rising ASO titer or Anti-Dnase B titer
2) A positive throat culture

Rheumatic Fever Recurrences

- Reliable past history of ARF: 2 major or 1 major and 2 minor or 3 minor manifestations sufficient for diagnosis
- Presence of antecedent streptococcal infection
- When minor manifestations only present exclude other causes.

DDX of ARF

Presentation		
Polyarthritis and fever	**Carditis**	**Chorea**
Septic arthritis (including disseminated gonococcal infection)†	Innocent murmur	Systemic lupus erythematosus
Connective tissue and other autoimmune disease††	Mitral valve prolapse	Drug intoxication
Viral arthropathy¥	Congenital heart disease	Wilson's disease
Reactive arthropathy¥	Infective endocarditis	Tic disorder‡
Lyme disease≠	Hypertrophic cardiomyopathy	Choreoathetoid cerebral palsy
Sickle cell anaemia	Myocarditis: viral or idiopathic	Encephalitis
Infective endocarditis	Pericarditis: viral or idiopathic	Familial chorea (including Huntington's)
Leukaemia or lymphoma		Intracranial tumour
Gout and pseudogout		Lyme disease≠
		Hormonal§

Investigations

Recommended for all cases
White blood cell count
Erythrocyte sedimentation rate (ESR)
C-reactive protein (CRP)
Blood cultures, if febrile
Electrocardiogram (if prolonged P-R interval or other rhythm abnormality, repeat in 2 weeks and again at 2 months, if still abnormal
Chest X-ray, if clinical or echocardiographic evidence of carditis
Echocardiogram (consider repeating after 1 month, if negative)
Throat swab (preferably before giving antibiotics): culture for group A streptococcus
Antistreptococcal serology: both ASO and anti-DNase B titres, if available (repeat 10–14 days later if first test not confirmatory)

Investigations

Investigations in suspected ARF

All patients with suspected or confirmed ARF should undergo echocardiography to confirm or refute the diagnosis of rheumatic carditis.

Treatment of ARF

- Bed rest
- Salicylates : Aspirin
  - 75-100 mg /kg/day given as 4 divided doses for 6 -8 weeks
  - Attain a blood level 20-30 mg/dl
- Penicillin: Procaine Penicillin 4 million units/day x10 days
- Prednisolone:2mg/kg/day taper over 6 weeks, Given when there is severe carditis
- Heart Failure Treatment: diuretics, ACEI

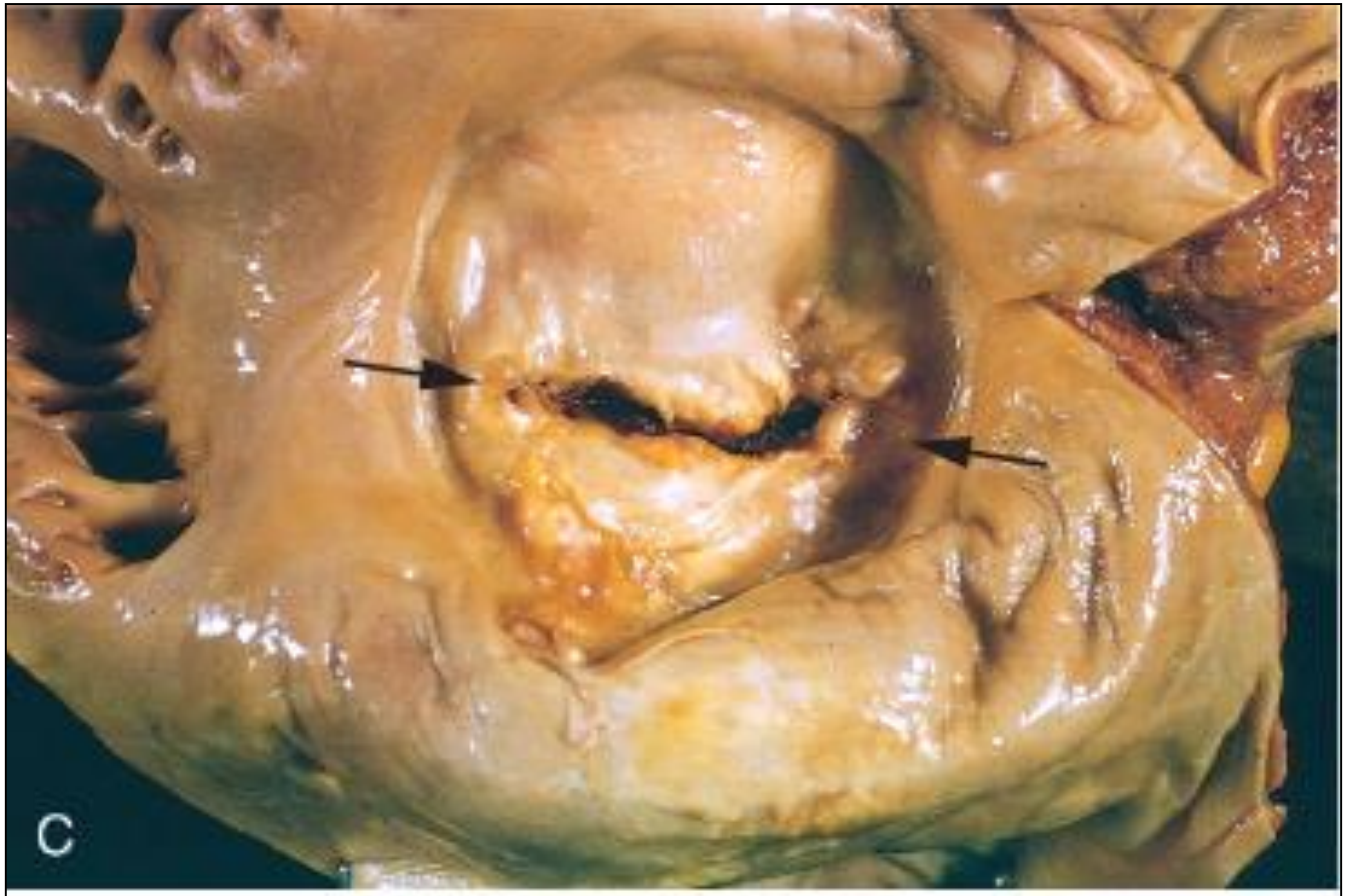
Chronic Rheumatic Heart Disease

- Most commonly in Mitral-70%
- Frequently in Aortic-40%
- Less frequently Tricuspid-10%
- Rarely pulmonary valve-2%
- Mitral Stenosis is more common in females(3:1), while males have higher incidence of Aortic Regurgitation

Mitral Stenosis

- The normal MVA= 4-5 cm2
- In severe ms <1.5 cm2
- High LAP
- The rise in LAP causes a similar rise in pulmonary capillaries, veins and artery

Mitral Stenosis



Clinical Features

- Dyspnea
- Fatigue
- Palpitation
- Hemoptysis (10%)
- Hoarseness (Ortner's syndrome)
- Dysphagia
- Storke or peripheral embolization

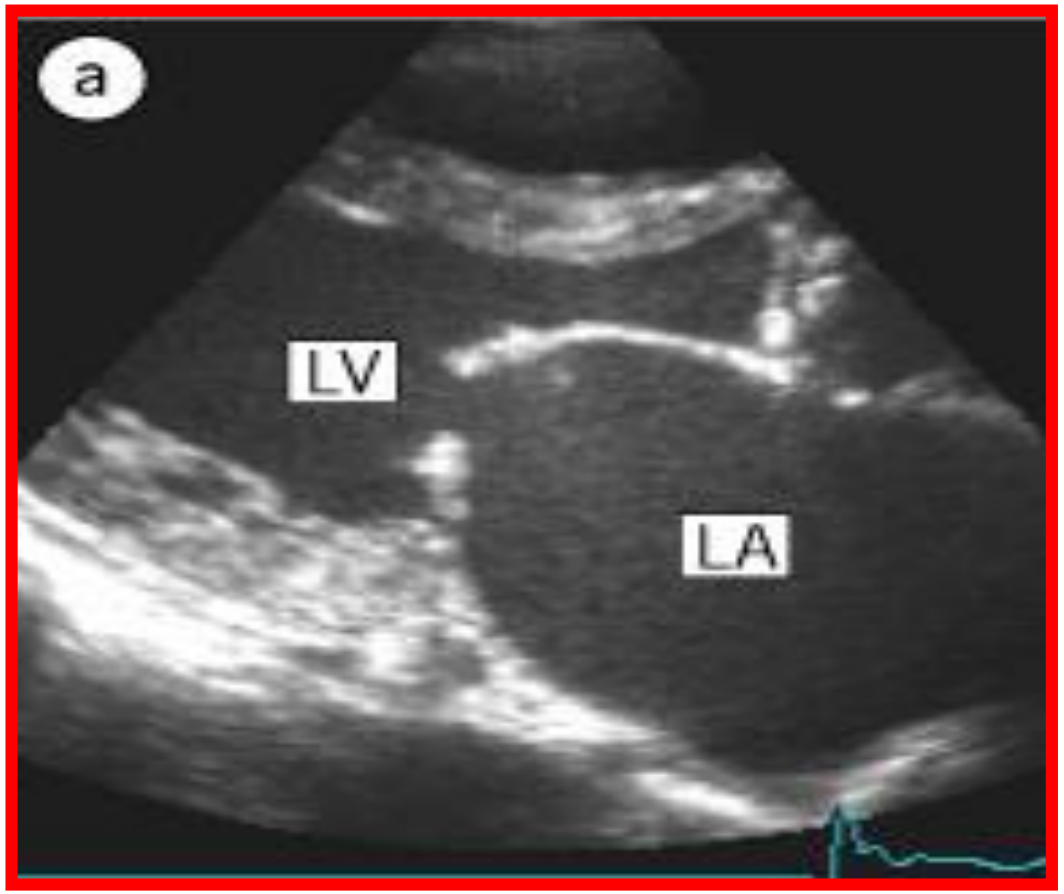
Clinical Features

- Cyanosis (Mitral facies,malar flush)
- Tapping apex (S1)
- Parasternal heave
- Diastolic thrill
- Accentuated S1 , accentuated S2
- Opening snap
- Mid-diastolic rumb**le**

Investigations

- CXR
  - Straightening of the left heart border
  - Double density
  - Kerley B lines , CA in MV
- ECG: LAE, P Mitrale ,RV dominance
- Echodoppler

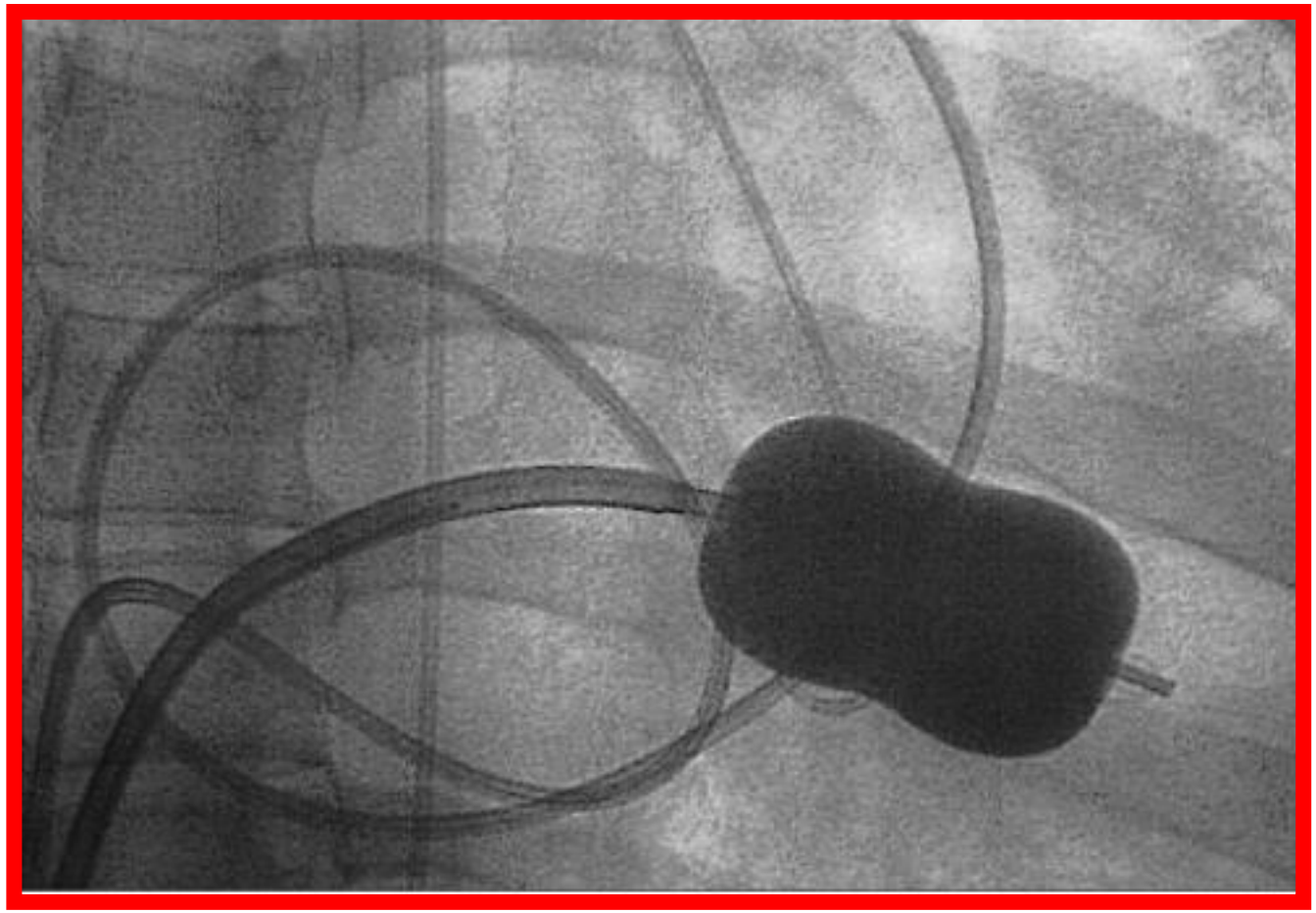
Echo In Mitral Stenosis



Management

- B-Blockers ,CCB
- Digoxin (AF)
- Warfarin
- Balloon Valvuloplasty
- Mitral valve replacement

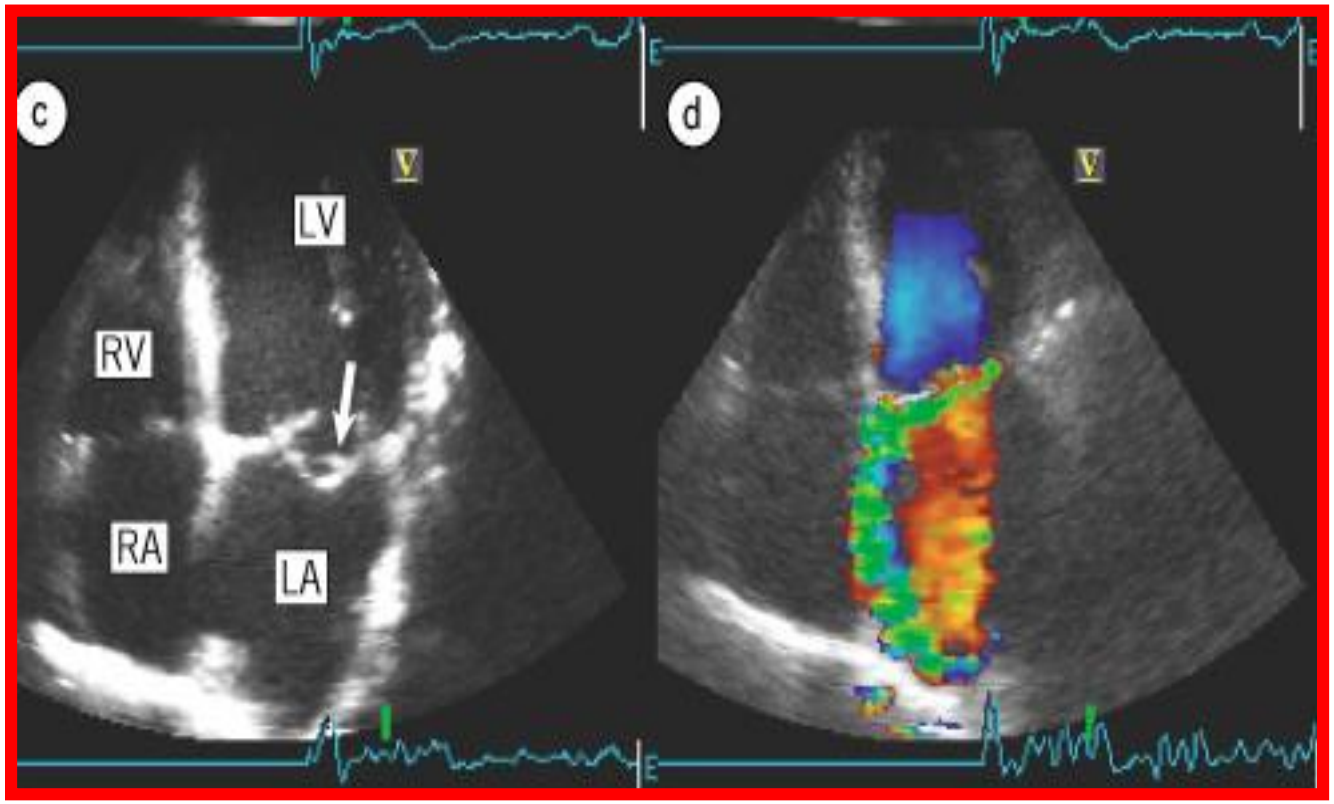
BMV



Mitral Regurgitation

- Asymptomatic
- Dyspnea , orthopnea, PND
- Displaced PMI, Thrill
- Soft S1,
- Pansystolic murmur
- Treatment is surgical

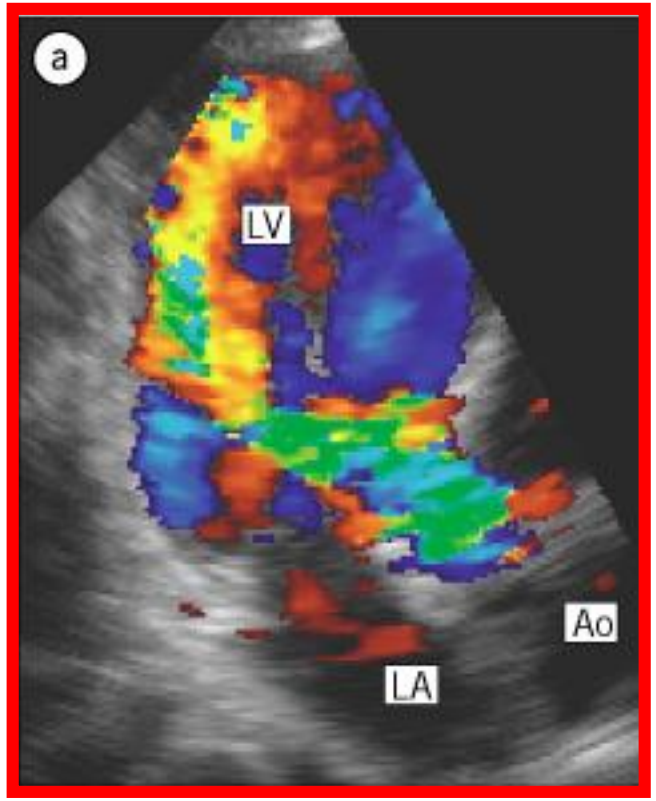
ECHO



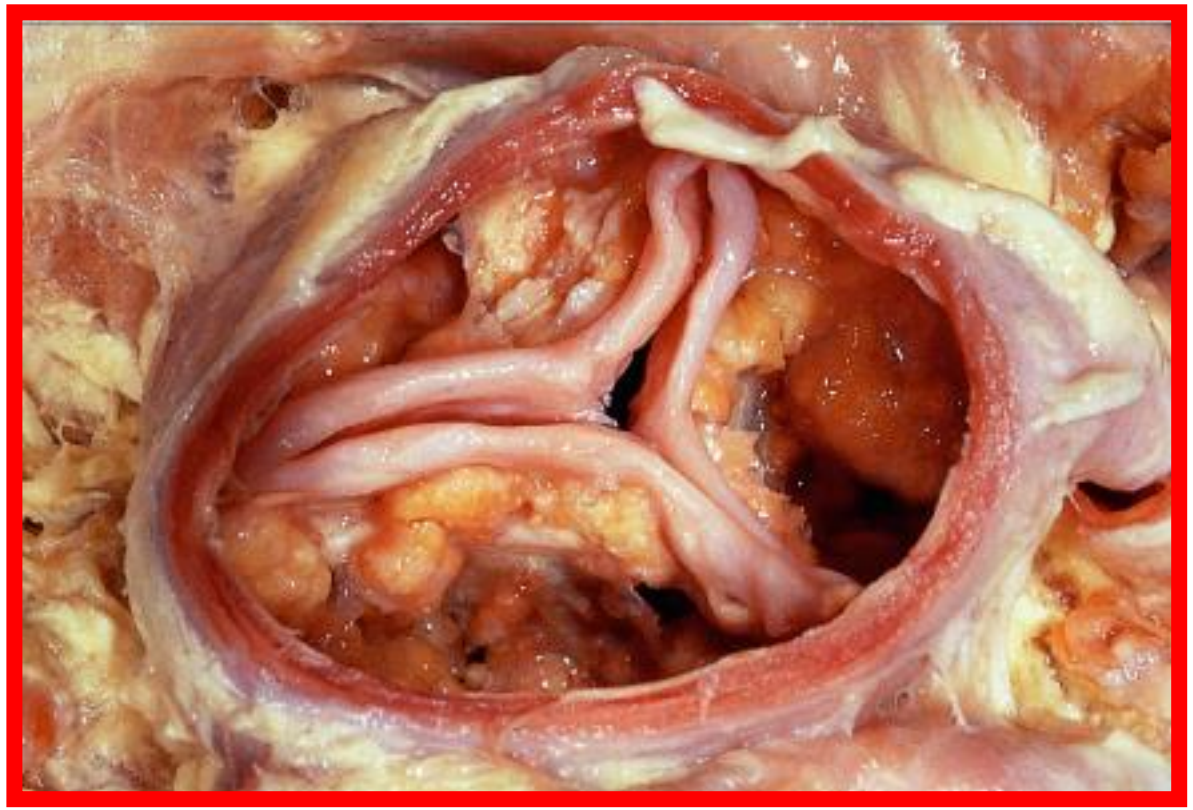
Aortic Regurgitation

- Water-hammer / collapsing pulse
- Wide pulse pressure
- Corrigan's sign
- De Musset sign
- Muller sign
- Quincke's pulse
- Hill's sign

ECHO



Aortic Stenosis



Symptoms

- Angina
- Syncope
- Dyspnea

Signs

- Arterial Pulse wave form : Plateau
  - Small (Parvus)
  - Slow rise (Tardus)
- Sustained not displaced PMI
- Systolic thrill
- S4

Signs

- Late peaking of murmur
- Single S2 : Soft or absent A2
- Paradoxical splitting of S2

Aortic Valve Disease

Treatment:

- Aortic valve Replacement
- Transcathter Aortic Valve Replacement

Secondary Prevention of Rheumatic Fever (Prevention of Recurrent Attacks)

Agent	Dose	Mode
Benzathine penicillin G	1 200 000 U every 4 weeks*	Intramuscular
or		
Penicillin V	250 mg twice daily	Oral
or		
Sulfadiazine	0.5 g once daily for patients 27 kg (60 lb 1.0 g once daily for patients >27 kg (60 lb)	Oral

For individuals allergic to penicillin and sulfadiazine

Erythromycin	250 mg twice daily	Oral

*In high-risk situations, administration every 3 weeks is justified and recommended

Duration of Secondary Rheumatic Fever Prophylaxis

Category	Duration
Rheumatic fever with carditis and residual heart disease (persistent valvar disease*)	10 y since last episode or until age 40y ,(which-ever is longer), sometimes life long prophylaxis
Rhumatic fever with carditis But no residual VHD	10 yrs or until age 21yrs (whichever is longer)
Rheumatic fever without carditis	5 y or until age 21 y, (whichever is longer)

*