Rheumatic Fever And RHD

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Lecture Outline

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

Rhuematic Fever

- It represents a delayed immune response to infection with manifestations appearing after a period of 2-4 weeks.
- Group A beta hemolytic streptococcal throat infection (***Strep. pyogenes).***
- Age 5-15 yrs
- A multisystem disease

Rheumatic Heart Disease (RHD)

- RHD is a long term complication of ARF.
- Major effect on health is due to damage to heart valves.

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 been 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.

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 further episodes of ARF causing damage, and the need for valve surgery, and or 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 1992 by AHA
- Revised recently -2015 by AHA

Modified Jones Criteria 1992

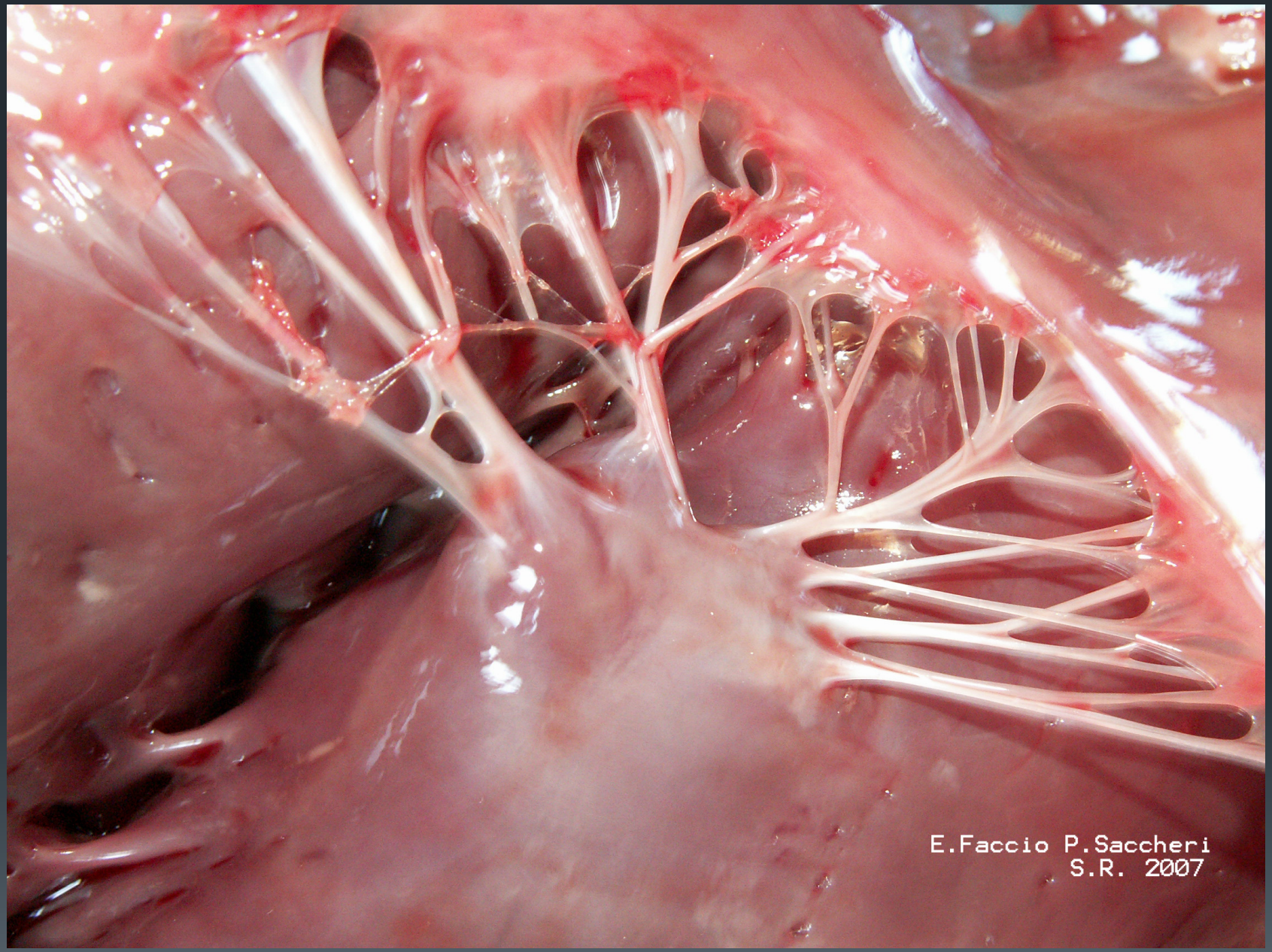
Major criteria	Minor criteria
Migratory polyarthritis	Arthralgia
Carditis	Fever
Erythema marginatum	First degree heart block
Syndenham chorea	Elevated inflammatory markers (ESR, CRP)
Subcutaneous nodules	

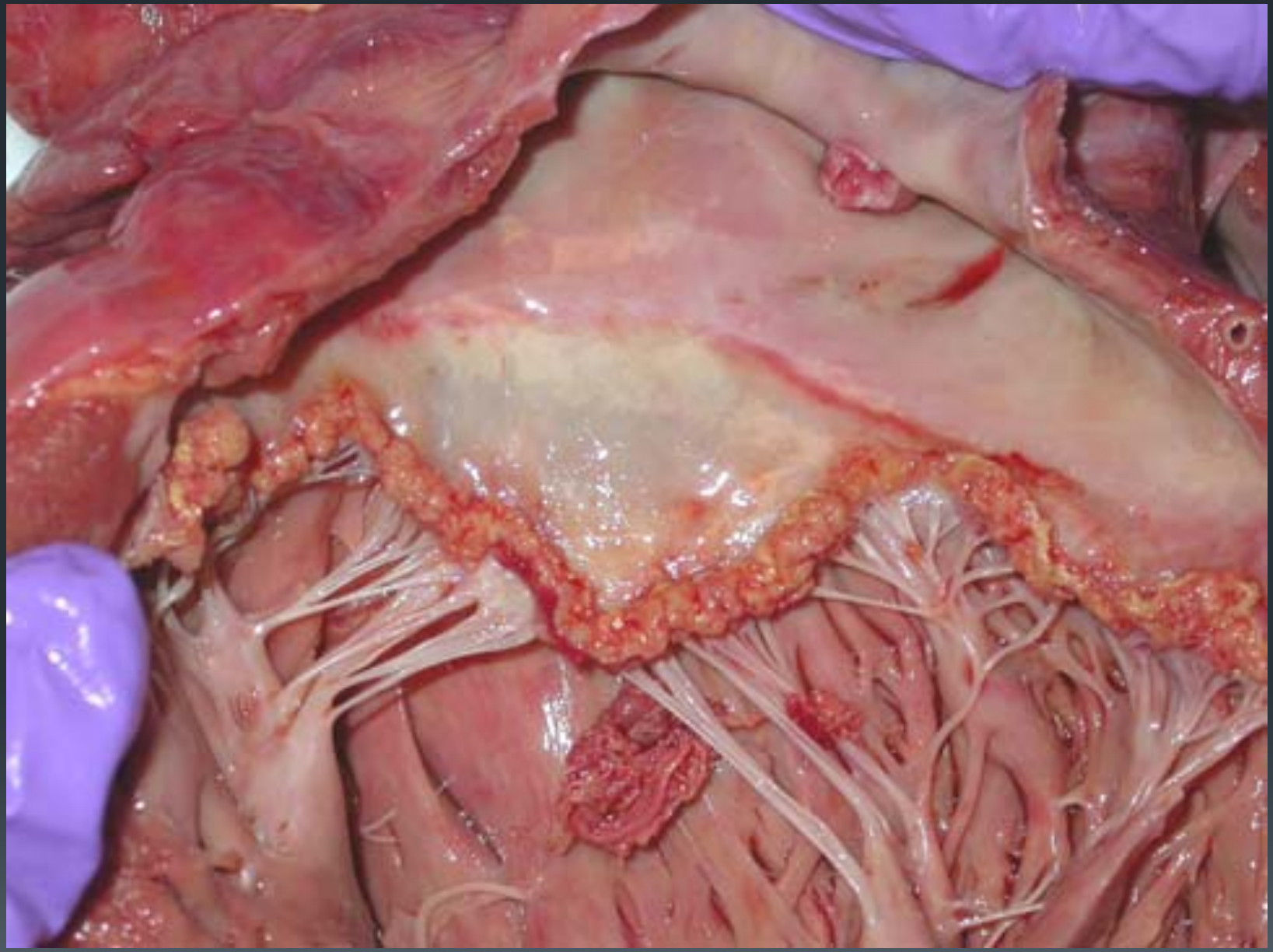
Mnemonic for Jone's criteria

- Recent Strep infection.
- J♥nes Criteria (Major Criteria)
  - J Joints (Polyartheritis).
  - ♥ Carditis (Pancarditis).
  - N Nodules.
  - E Erythema marginatum.
  - S Sydenham's chorea.
- Minor criteria:
  - Fever, ESR, Arthralgia, Long PR interval.

Carditis

- Occurs in 50-70% of cases
- Only manifestation of ARF that leaves permanent damage
- May be subclinical.
- Myocardium, endocardium, and pericardium.
- Tachycardia is common; its absence makes the diagnosis of myocarditis unlikely.
- Murmurs of MR or AR may occur in acute stage while mitral stenosis occurs in late stages
- Cardiomegaly and CHF may occur

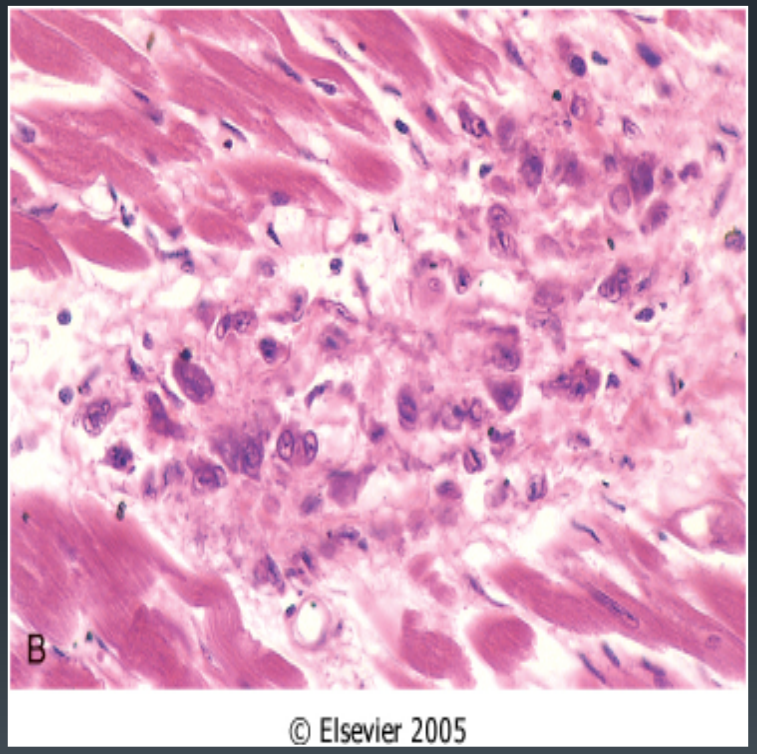

E.Faccio P.Saccheri
S.R. 2007

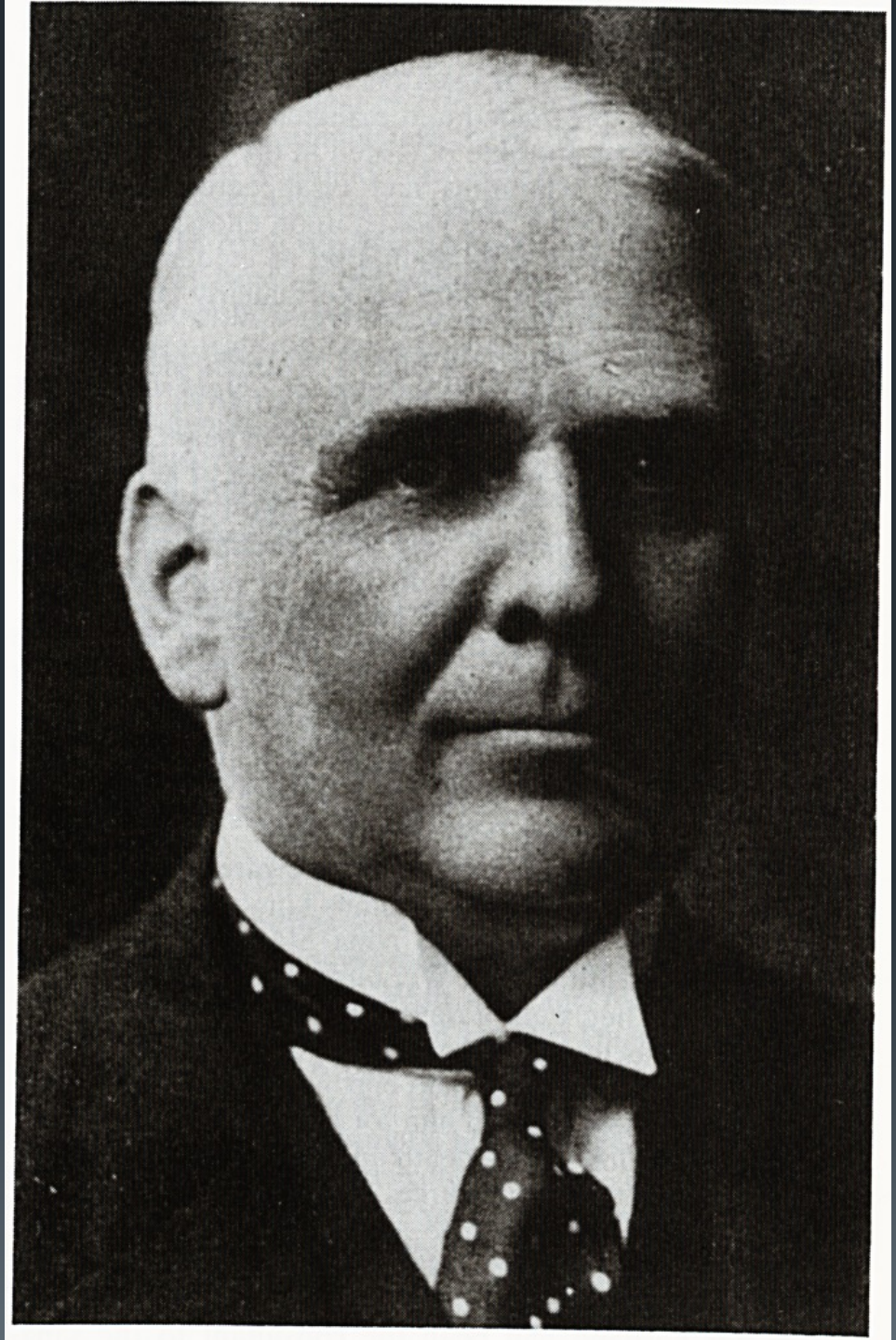


Confluent verrucae located on the atrial surface of the mitral valve consistent with acute rheumatic involvement of the heart

Pathologic Lesions

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





Carey Coombs (1879-1932)

- Described the diastolic murmur of acute rheumatic mitral valvulitis.
- Had great interest in heart disease in children.
- The first to describe myocardial infarction, credit actually went to James Herrick who published his findings in JAMA in 1912.

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

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

Revised Jones Criteria 2015

AHA Scientific Statement

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

Endorsed by the World Heart Federation

Michael H. Gewitz, MD, FAHA, Co-Chair; Robert S. Baltimore, MD, Co-Chair; Lloyd Y. Tani, MD, FAHA; Craig A. Sable, MD, FAHA; Stanford T. Shulman, MD; Jonathan Carapetis, MBBS; Bo Remenyi, MBBS; Kathryn A. Taubert, PhD, FAHA; Ann F. Bolger, MD, FAHA; Lee Beerman, MD; Bongani M. Mayosi, MBChB; Andrea Beaton, MD; Natesa G. Pandian, MD; Edward L. Kaplan, MD, FAHA; on behalf of the American Heart Association Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the Council on Cardiovascular Disease in the Young

(*Circulation*. 2015;131:000-000. DOI: 10.1161/CIR.0000000000000205.)

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. Polyarthralgia 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.

2015 Revised Jones Criteria

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

Table 6. Differential Diagnosis of Arthritis, Carditis, and Chorea

Arthritis	Carditis	Chorea
Septic arthritis (including gonococcal)	Physiological mitral regurgitation	Drug intoxication
Connective tissue and other autoimmune diseases such as juvenile idiopathic arthritis	Mitral valve prolapse	Wilson disease
Viral arthropathy	Myxomatous mitral valve	Tic disorder
Reactive arthropathy	Fibroelastoma	Choreoathetoid cerebral palsy
Lyme disease	Congenital mitral valve disease	Encephalitis
Sickle cell anemia	Congenital aortic valve disease	Familial chorea (including Huntington disease)
Infective endocarditis	Infective endocarditis	Intracranial tumor
Leukemia or lymphoma	Cardiomyopathy	Lyme disease
Gout and pseudo gout	Myocarditis, viral or idiopathic	Hormonal
Poststreptococcal reactive arthritis	Kawasaki disease	Metabolic (eg, Lesch-Nyhan, hyperalaninemia, ataxia telangiectasia)
Henoch-Schonlein purpura		Antiphospholipid antibody syndrome
		Autoimmune: Systemic lupus erythematosus, systemic vasculitis
		Sarcoidosis
		Hyperthyroidism

Investigations

- Recommended for all cases:

- CBC
- ESR
- CRP
- Blood cultures (if febrile)
- Throat swap before Abx (Group A Strep)
- Antistreptococcal serology: ASO and Anti-DNAse B titers. (repeat in 10-14 days if –ve first test)

Investigations

Recommended for all cases:

- ECG (prolonged PR or arrhythmia).
- CXR.
- Echocardiogram (2015).

Treatment of ARF

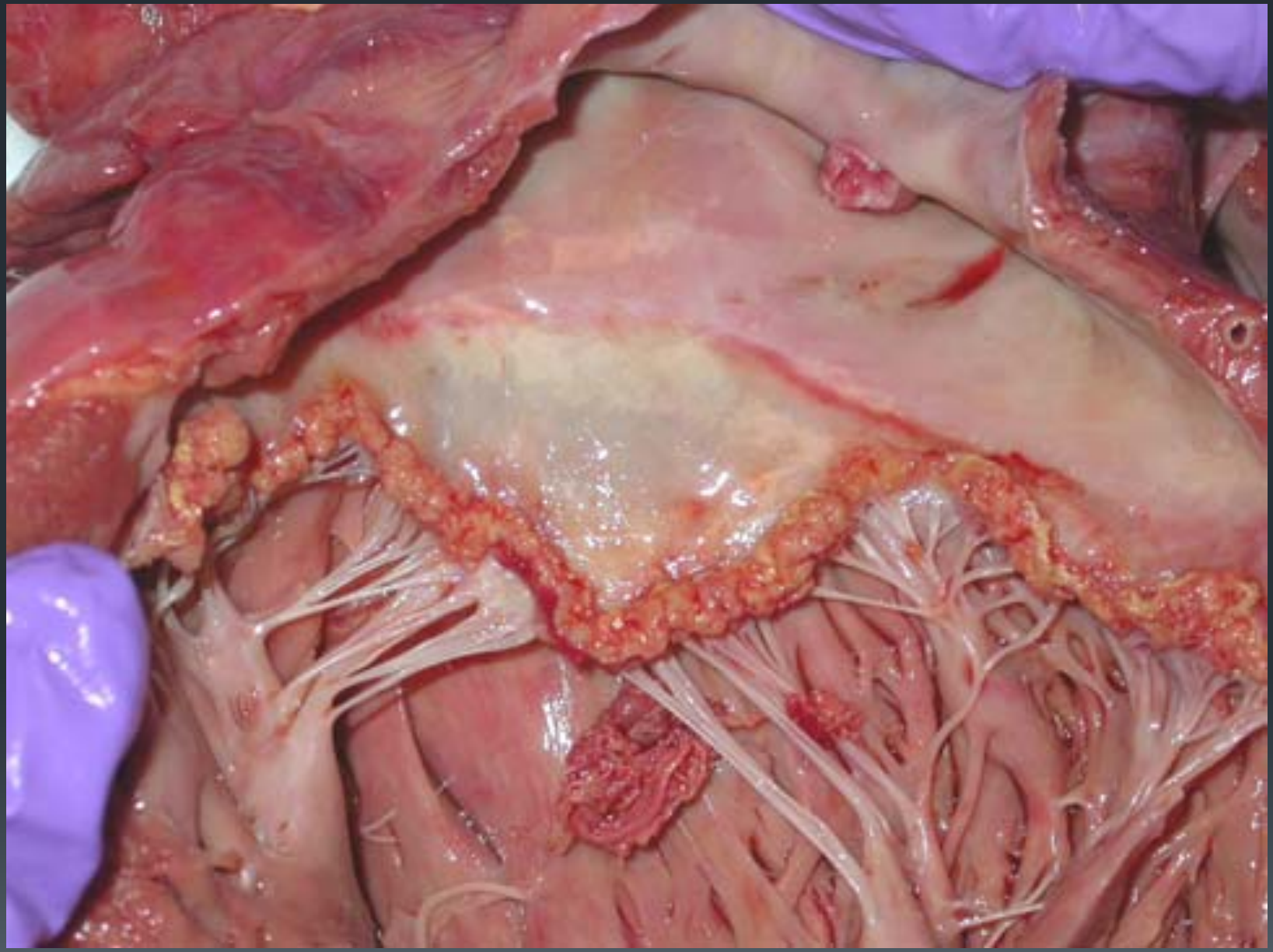
- Bed rest
- Salicylates : Aspirin
- 75-100 mg /kg/day given as 4 divided doses for 6 -8 weeks.
  Attain a blood level 20-30 mg/dl
- Penicillin: Procaine Penicillin 4 million units/day x10 days.
- Prednisolone:2mg/kg/day taper over 6 weeks, Given when there is severe carditis.
- Heart Failure Treatment: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

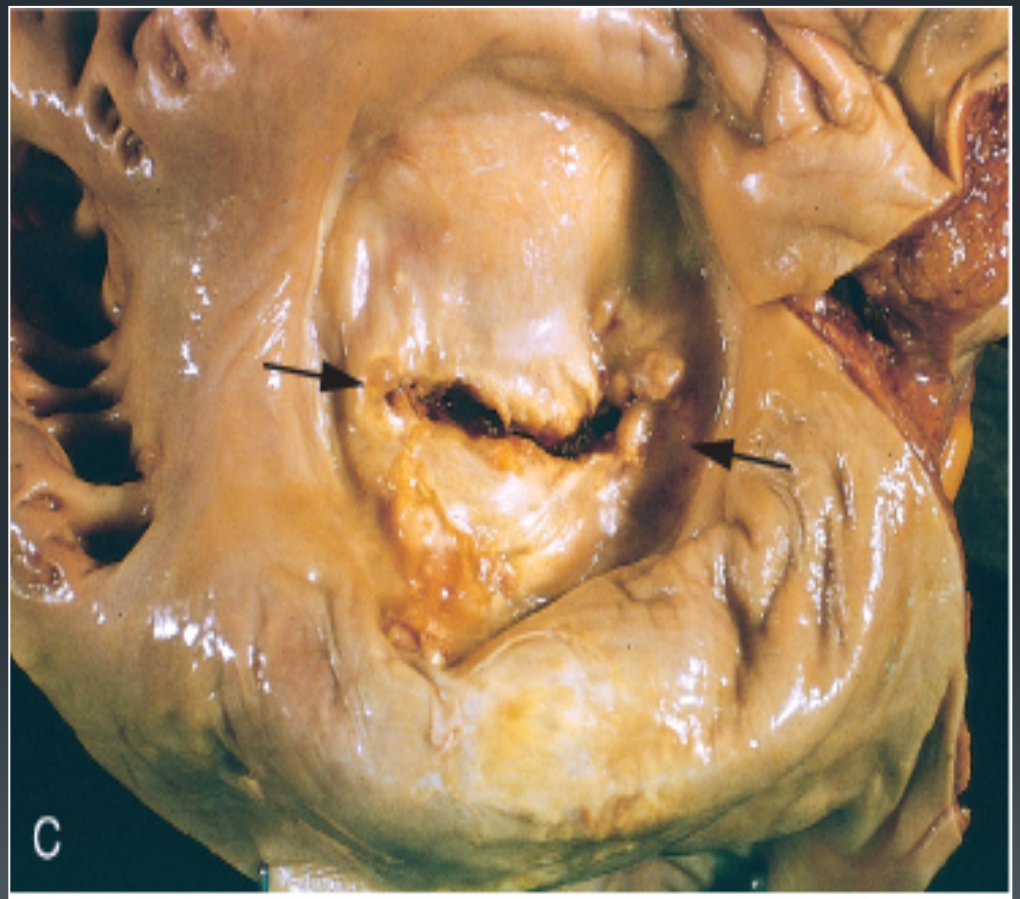
Mitral Stenosis (MS)

- The normal MVA= 4 - 5 cm^2
- In severe MS <1.5 cm^2
- High LAP
- The rise in LAP causes a similar rise in pulmonary capillaries, veins and artery.



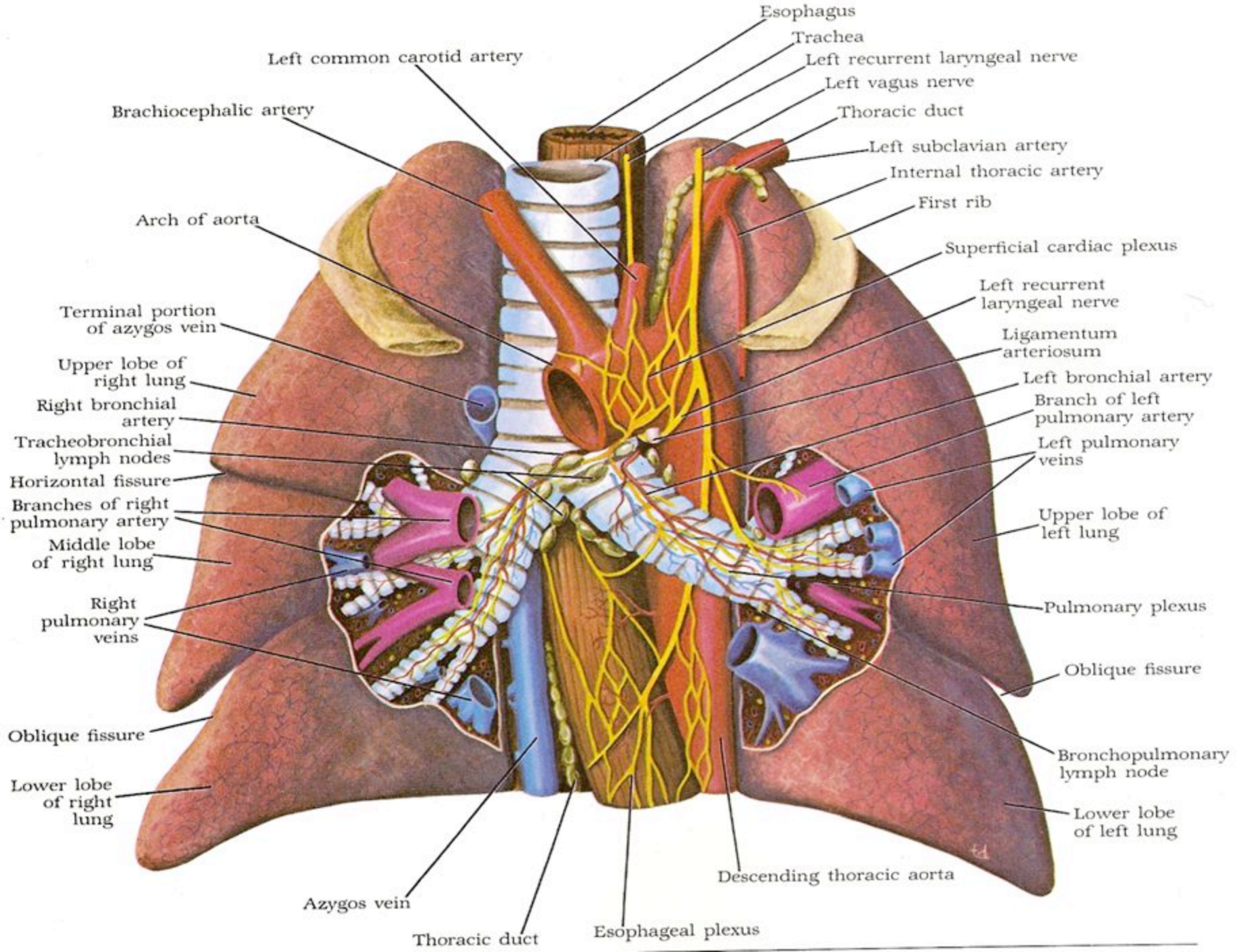
Confluent verrucae located on the atrial surface of the mitral valve consistent with acute rheumatic involvement of the heart

Mitral Stenosis



Clinical Features of MS (Symptoms)

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

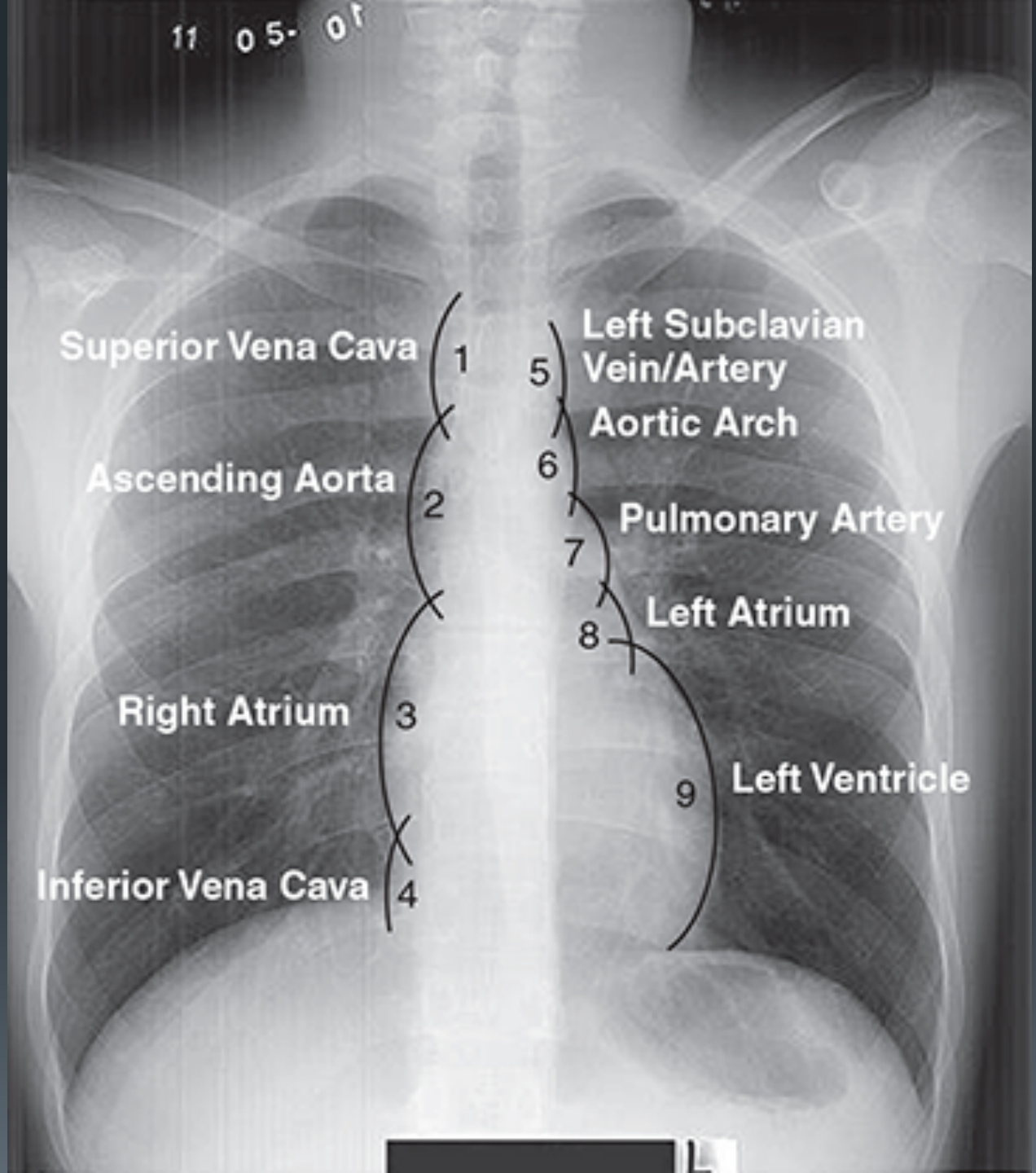

Esophagus
Trachea
Left common carotid artery
Left recurrent laryngeal nerve
Left vagus nerve
Brachiocephalic artery
Thoracic duct
Left subclavian artery
Internal thoracic artery
First rib
Arch of aorta
Superficial cardiac plexus
Left recurrent laryngeal nerve
Terminal portion of azygos vein
Ligamentum arteriosum
Upper lobe of right lung
Left bronchial artery
Right bronchial artery
Branch of left pulmonary artery
Tracheobronchial lymph nodes
Left pulmonary veins
Horizontal fissure
Branches of right pulmonary artery
Upper lobe of left lung
Middle lobe of right lung
Pulmonary plexus
Right pulmonary veins
Oblique fissure
Bronchopulmonary lymph node
Oblique fissure
Lower lobe of right lung
Lower lobe of left lung
Descending thoracic aorta
Azygos vein
Thoracic duct
Esophageal plexus

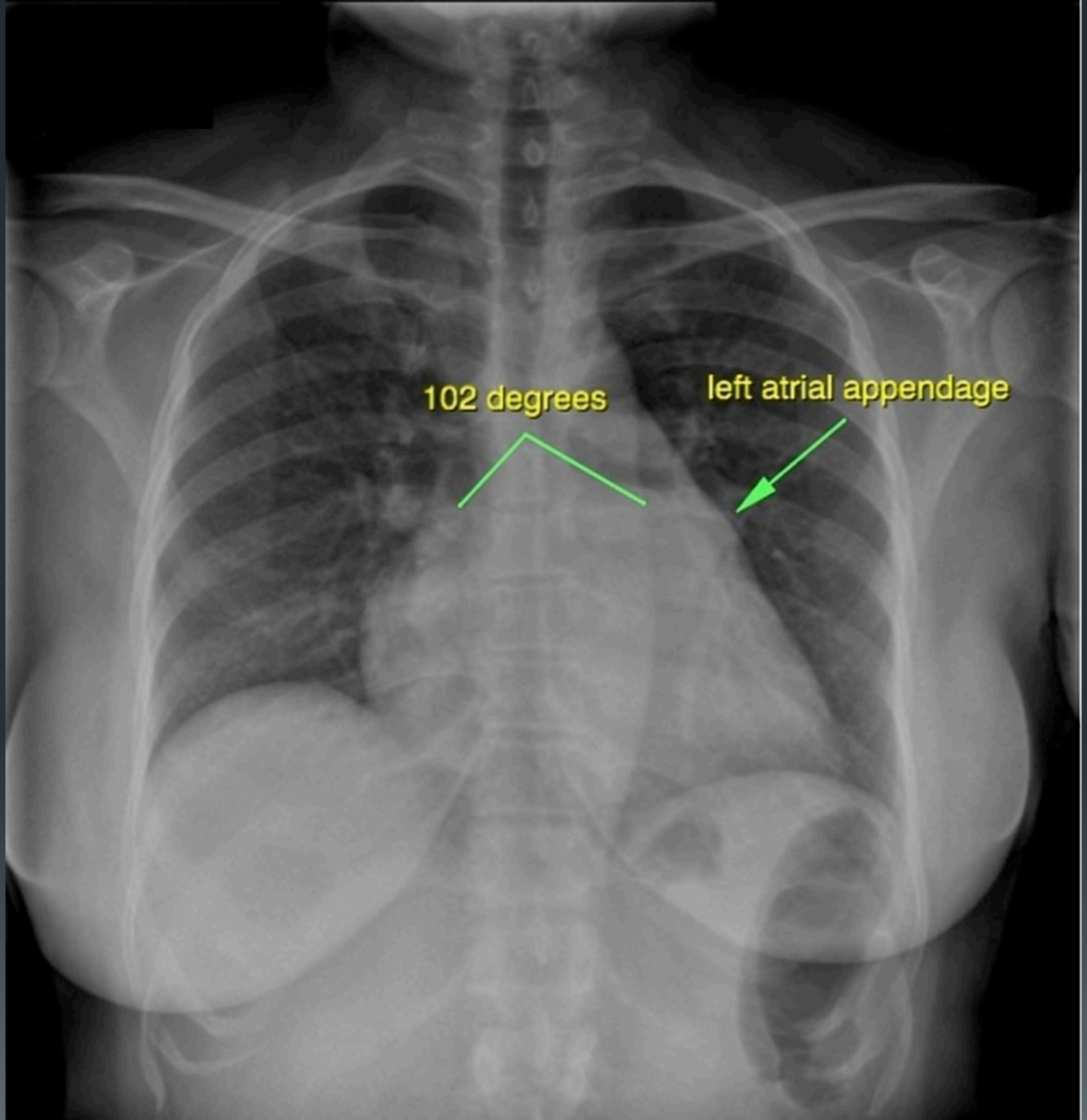
Clinical Features of MS (Physical Exam.)

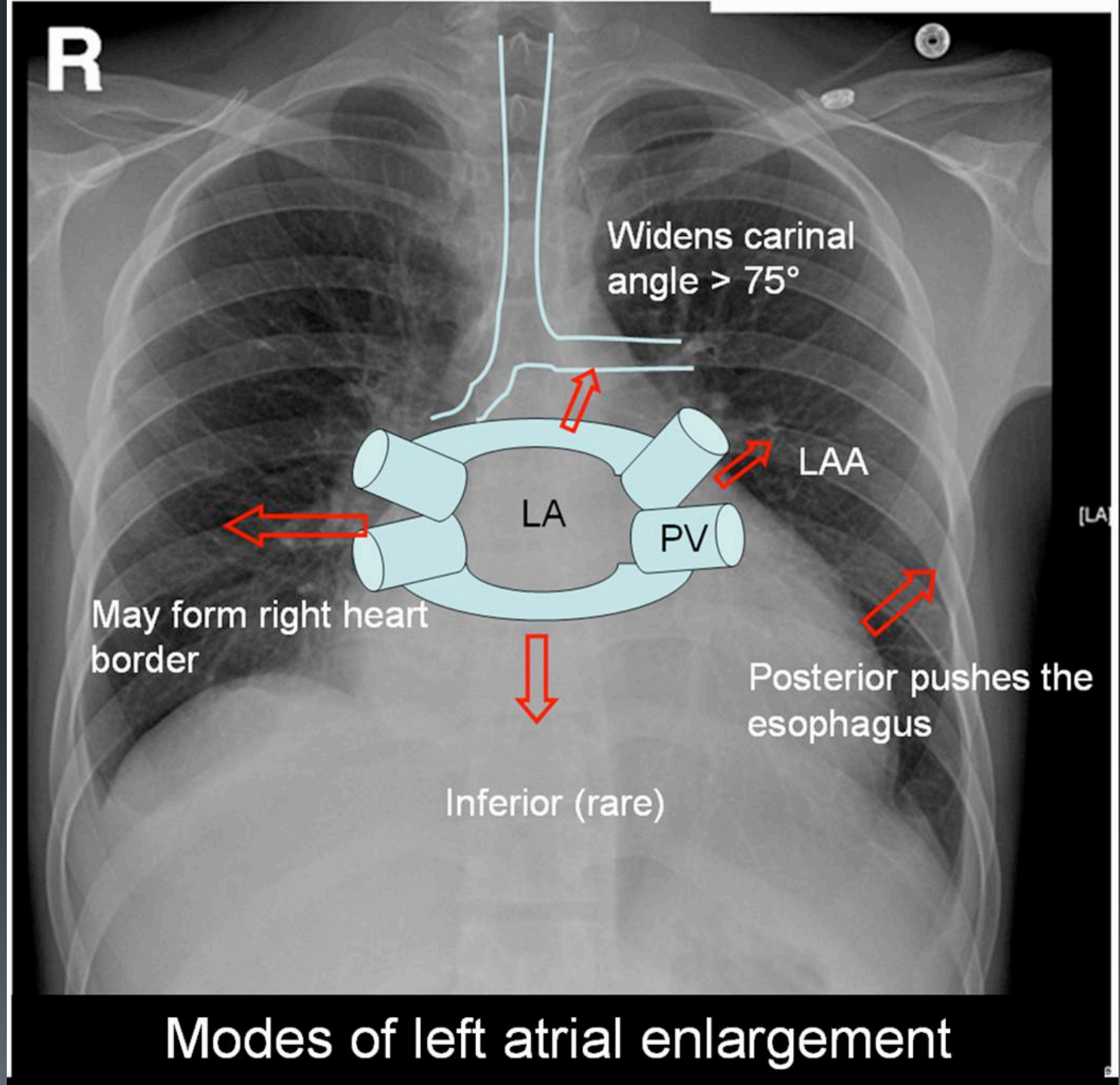
- Cyanosis (Mitral facies,malar flush)
- Tapping apex (S1)
- Parasternal heave
- Diastolic thrill
- Accentuated S1 , accentuated S2
- Opening snap
- Mid-diastolic rumble

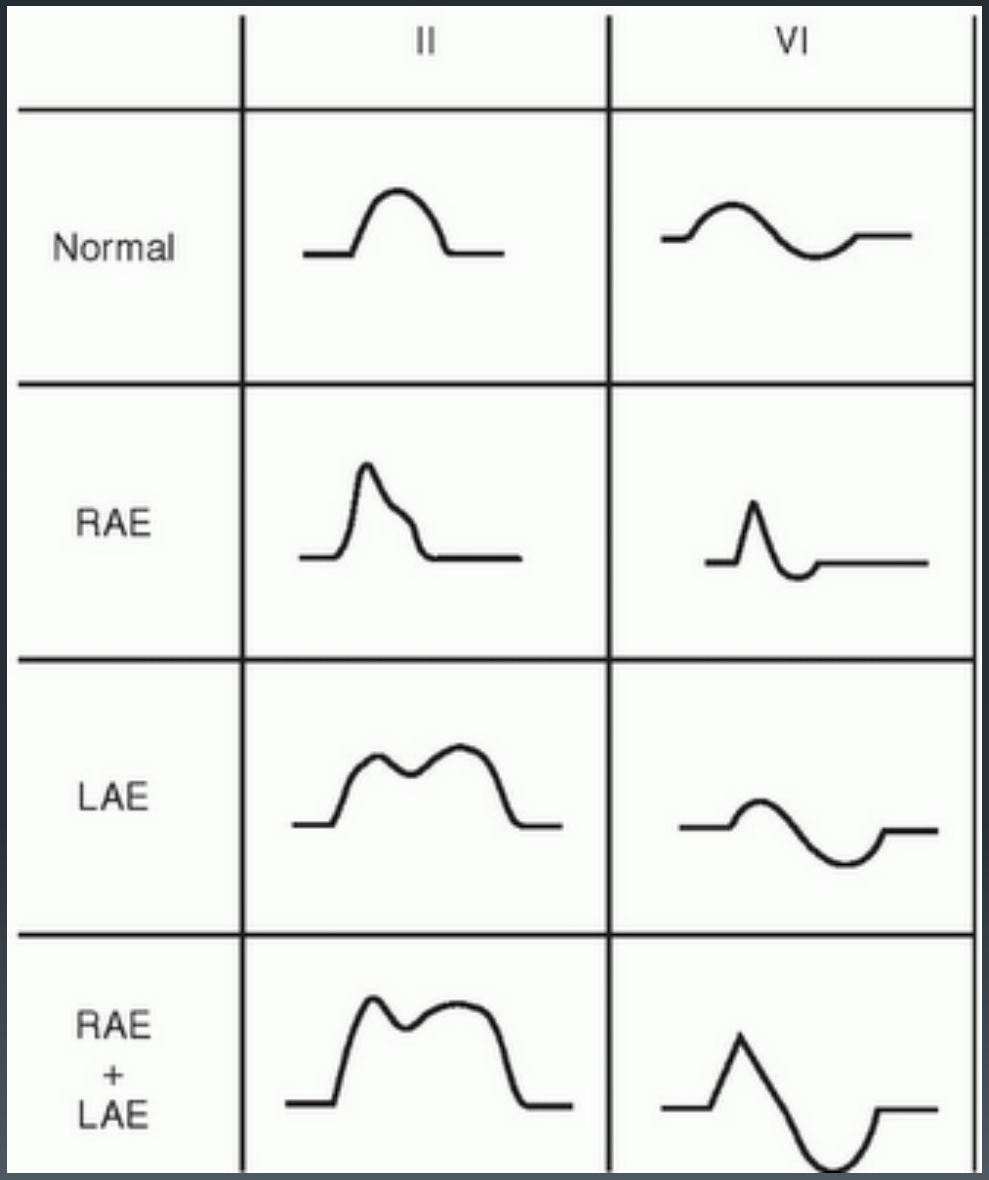
Investigations

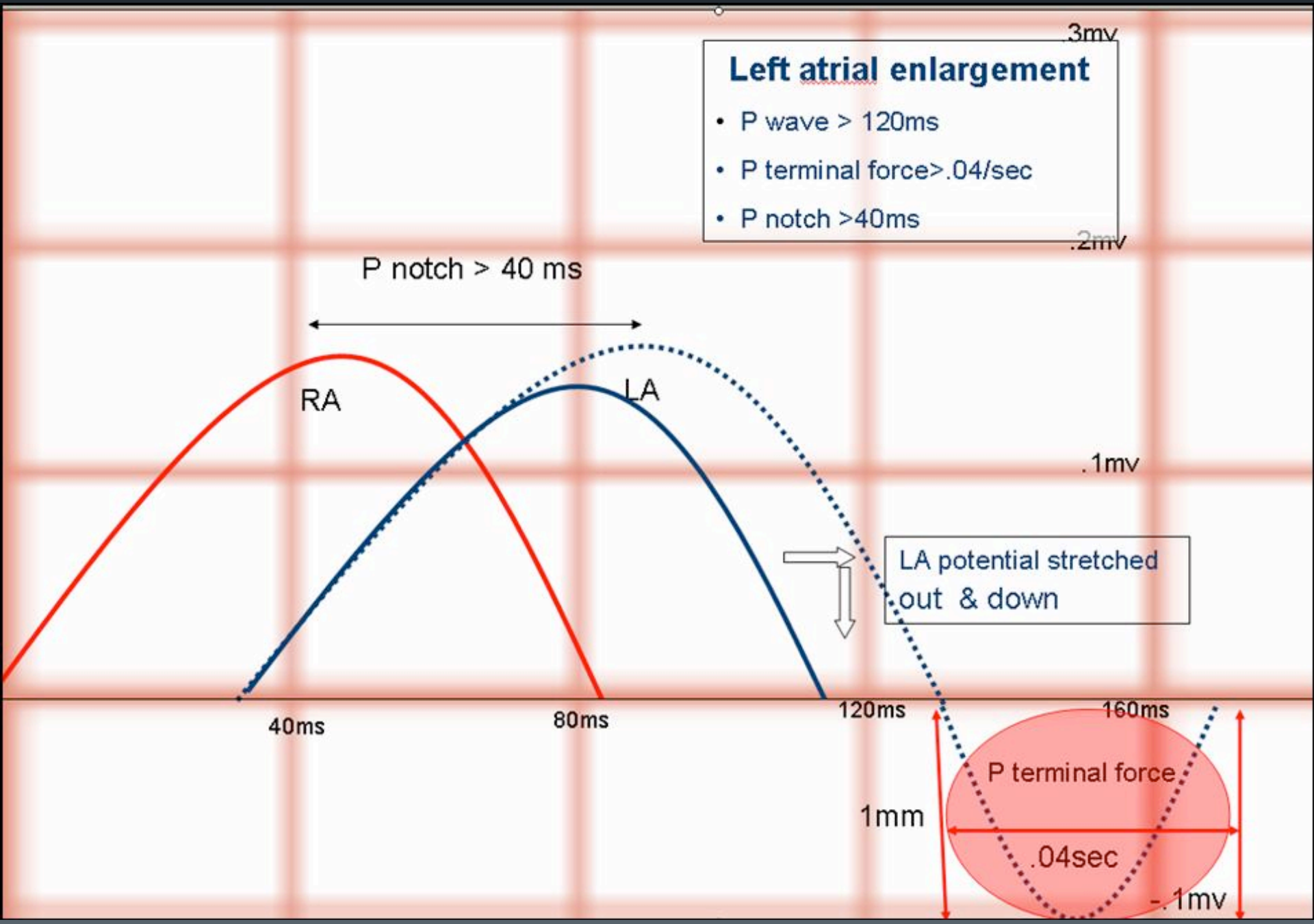
- CXR
  - Straightening of the left heart border
  - Double density
  - Kerley B lines , CA in MV
- ECG:
  - LAE, P Mitrale ,RV dominance
- Echodoppler


Superior Vena Cava
1
Ascending Aorta
2
Right Atrium
3
Inferior Vena Cava
4
5
Left Subclavian Vein/Artery
Aortic Arch
6
Pulmonary Artery
7
Left Atrium
8
9
Left Ventricle

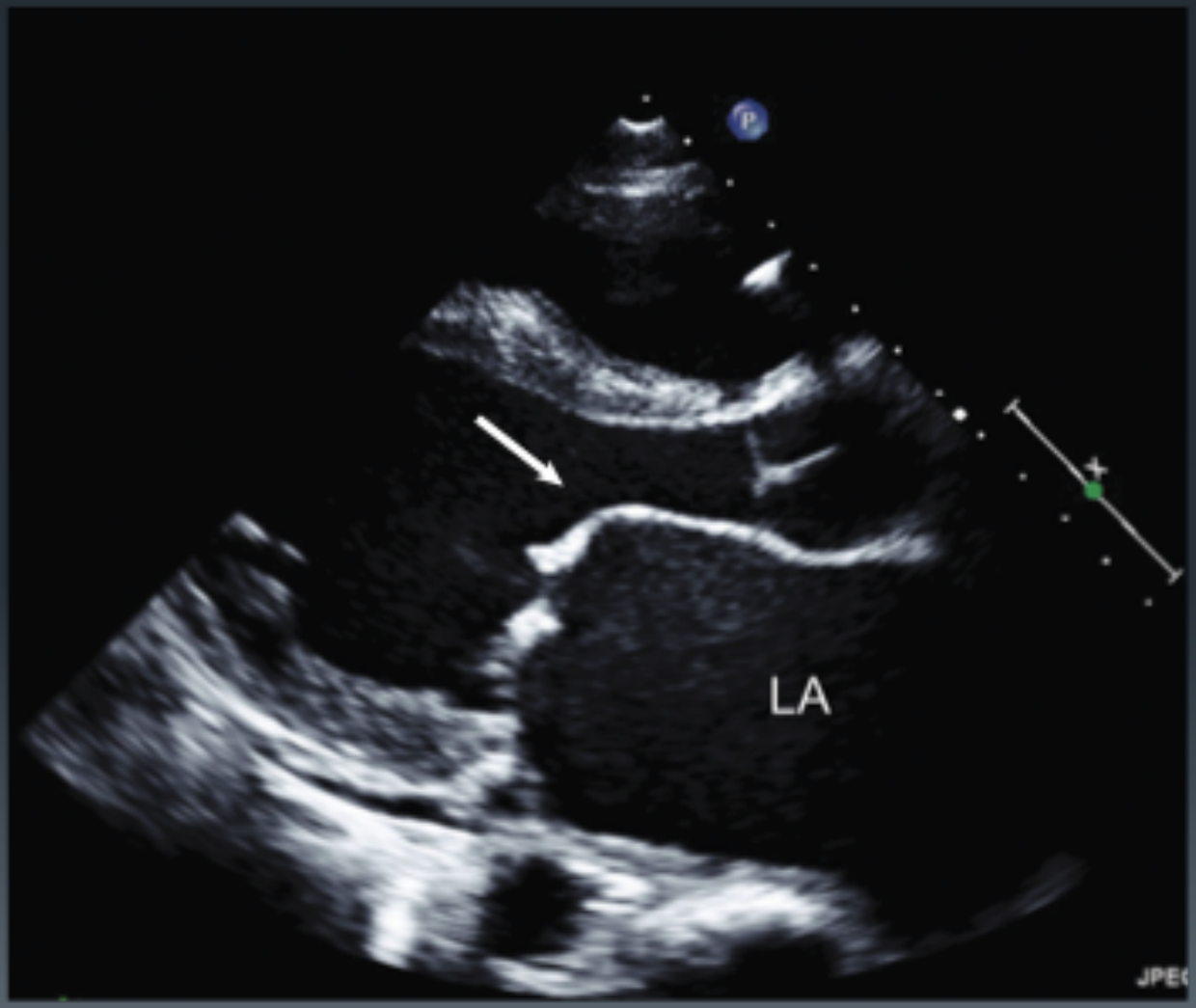

102 degrees
left atrial appendage

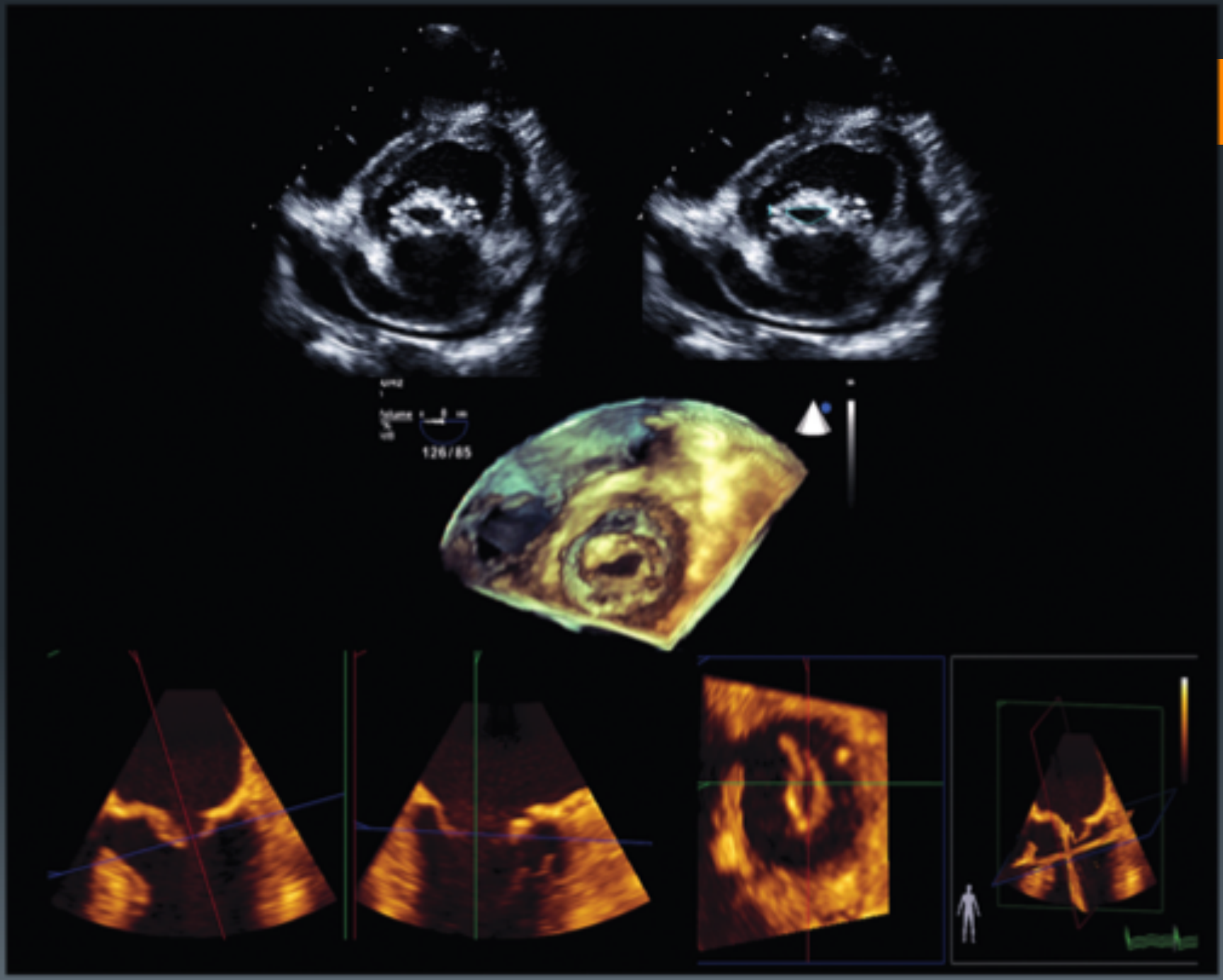

R
Widens carinal angle > 75°
LAA
LA
PV
May form right heart border
Posterior pushes the esophagus
Inferior (rare)
Modes of left atrial enlargement

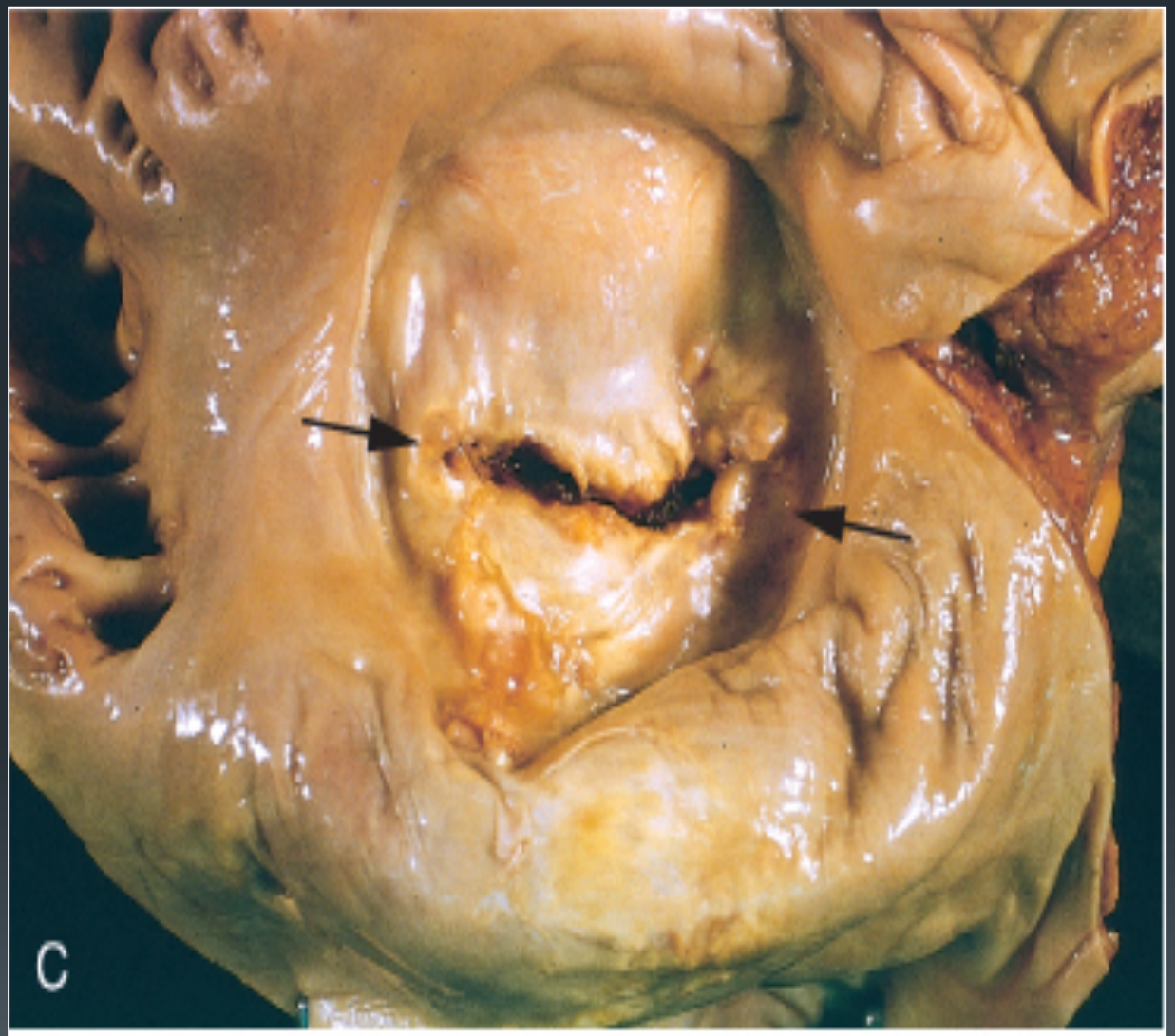

II
VI
Normal
RAE
LAE
RAE
+
LAE

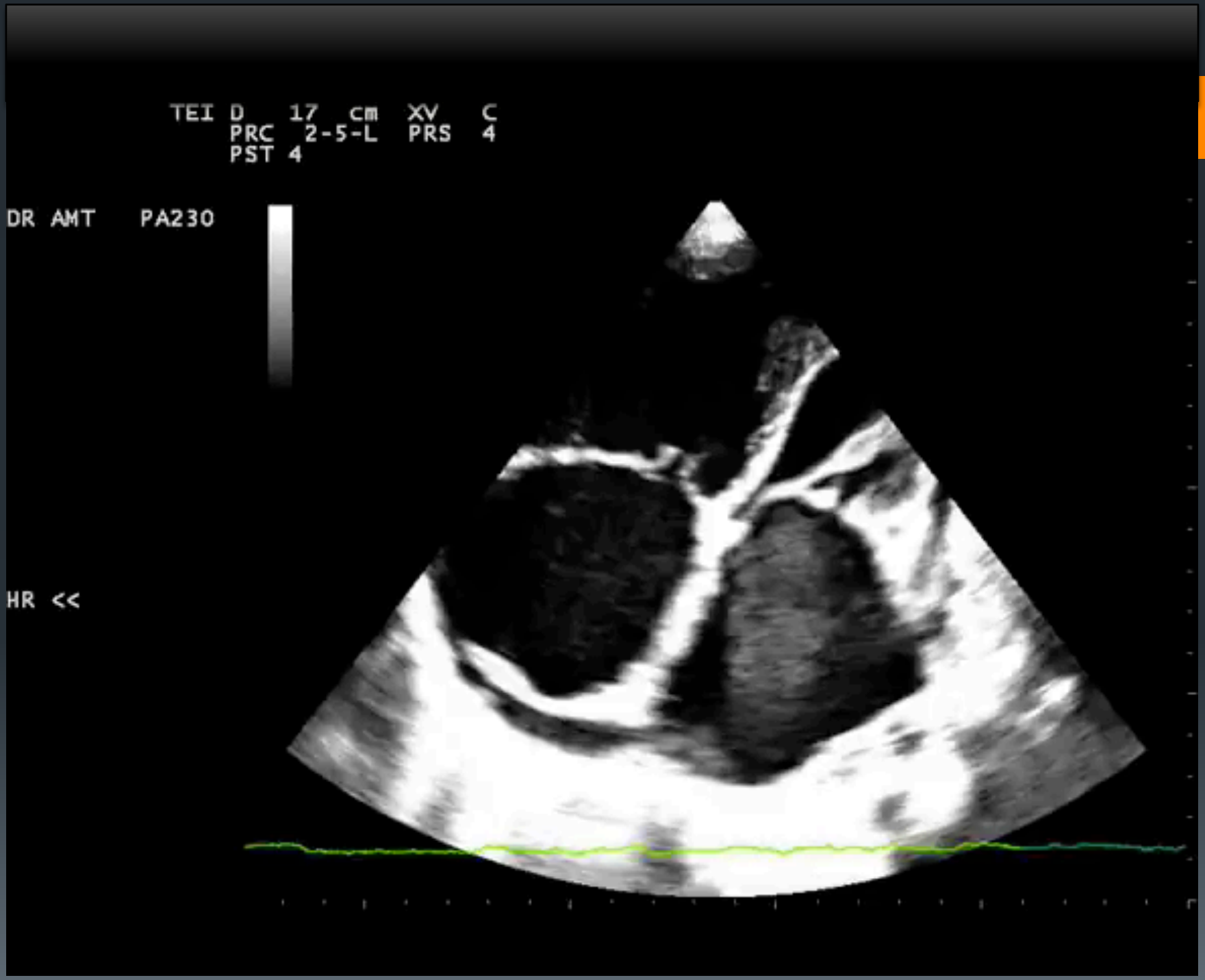

Left atrial enlargement
• P wave > 120ms
• P terminal force>.04/sec
• P notch >40ms
.3mv
.2mv
P notch > 40 ms
RA
LA
.1mv
LA potential stretched
out & down
40ms
80ms
120ms
160ms
P terminal force
1mm
.04sec
-.1mv

Echo In Mitral Stenosis



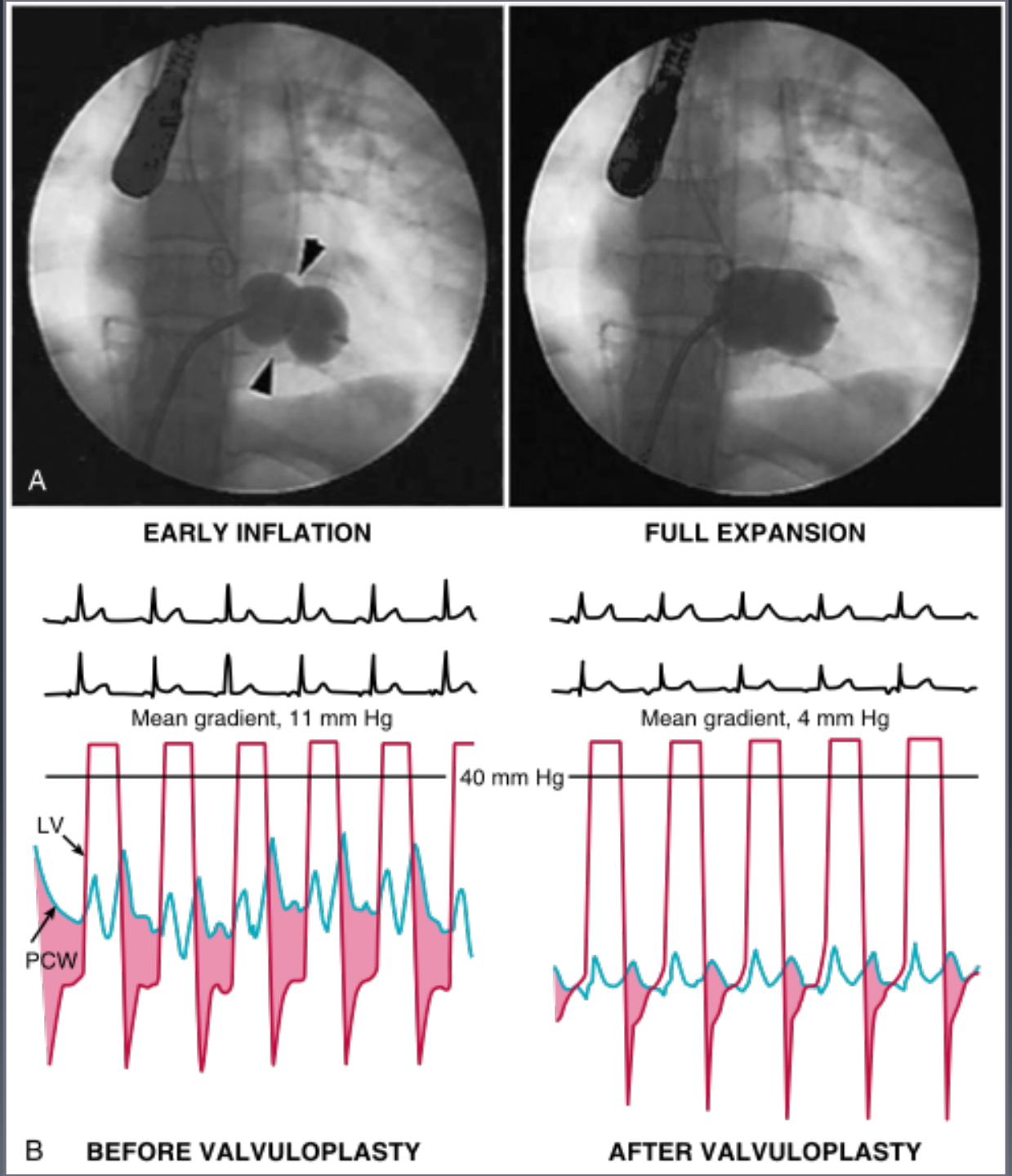

126/85




TEI D 17 cm XV C
PRC 2-5-L PRS 4
PST 4
DR AMT PA230
HR <<

Management

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


A
EARLY INFLATION
FULL EXPANSION
Mean gradient, 11 mm Hg
Mean gradient, 4 mm Hg
40 mm Hg
LV
PCW
B
BEFORE VALVULOPLASTY
AFTER VALVULOPLASTY

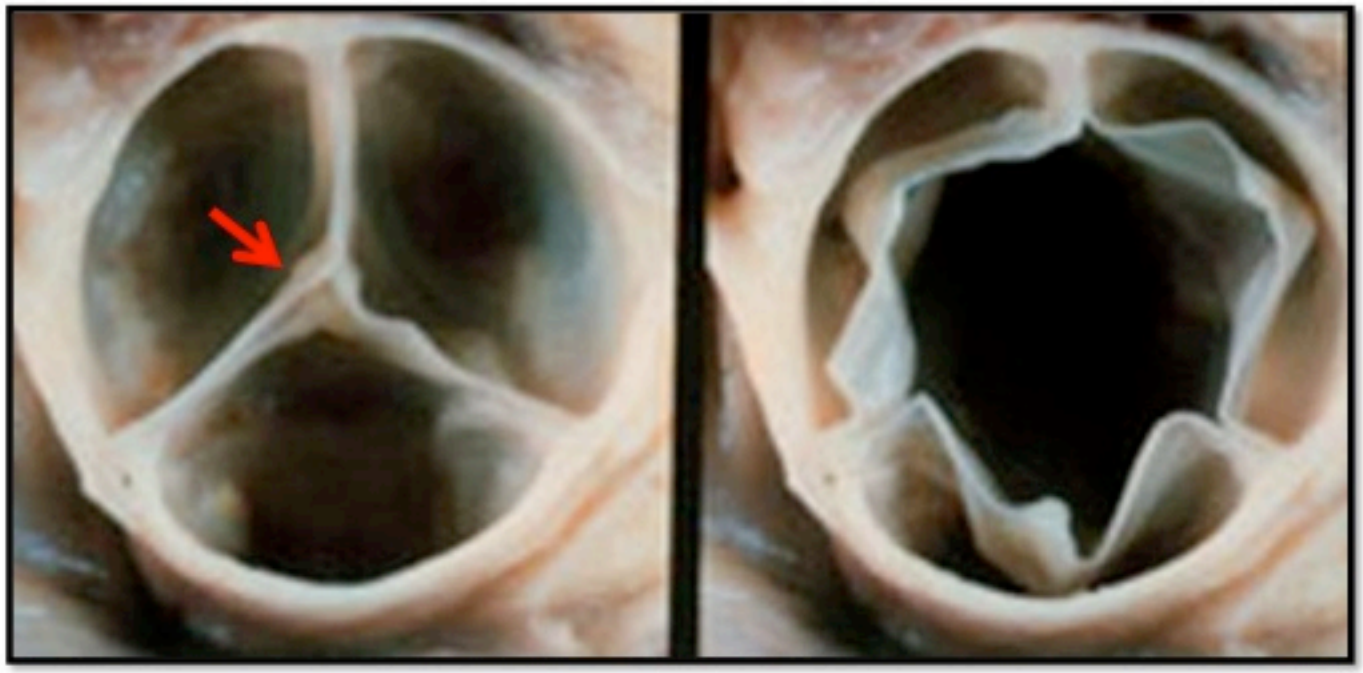
Mitral Regurgitation (MR)

- Asymptomatic
- Dyspnea , orthopnea, PND
- Displaced PMI, Thrill
- Soft S1,
- Pansystolic murmur
- Diagnosis confirmed and staged by echo.
- Treatment is surgical

Aortic Regurgitation (AR) signs

- Water-hammer / collapsing pulse
- Wide pulse pressure
- Corrigan's sign
- De Musset sign
- Muller sign
- Quincke's pulse
- Hill's sign
- Diagnosis confirmed and staged by echo.

Normal Aortic valve



Symptoms of AS

- Angina
- Syncope
- Dyspnea

Signs of Severe AS

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

Signs of Sever AS

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

Aortic Valve Disease

Treatment:

- Aortic valve Replacement (SAVR)
- Transcathter Aortic Valve Replacement (TAVI)

Secondary Prevention of Rheumatic Fever

Agent	Dose	Mode
Penicillin G	1.2 Million Units every 4 weeks (every 3 weeks in high risk situations)	IM
Penicillin V	250 mg twice daily	Oral
Sulfadiazine	1 g once daily	Oral
For individuals allergic to penicillin and sulfadiazine		
Erythromycin	250 mg twice daily	Oral

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.
Rheumatic fever with carditis but NO residual valvular heart disease.	10 y since last episode or until age 21y (which ever is longer).
Rheumatic fever without carditis	5 y since last episode or until age 21y (which ever is longer).

Good Luck ….
Qustions