السلام عليكم ورحمة الله وبركاته

ل امنة الله

Rheumatic fever & Valvular heart diseases

By

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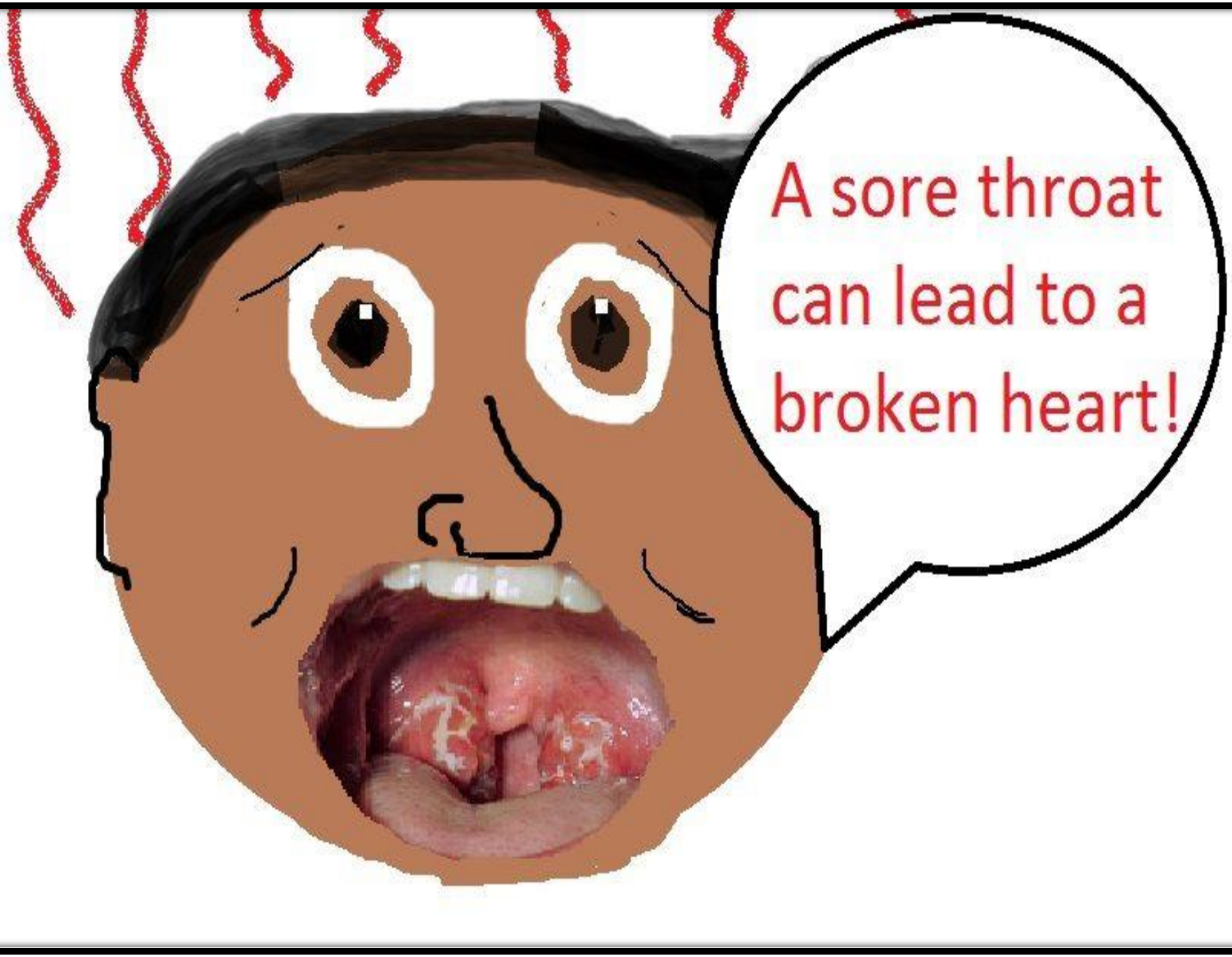
Clinical Background

- Acute rheumatic fever usually affects children and young adults between the ages of 5 - 15 years.
- Rh. Fever remains endemic in low economic countries in South Asia, Africa and India but rare in high economic countries.

Predisposing factors

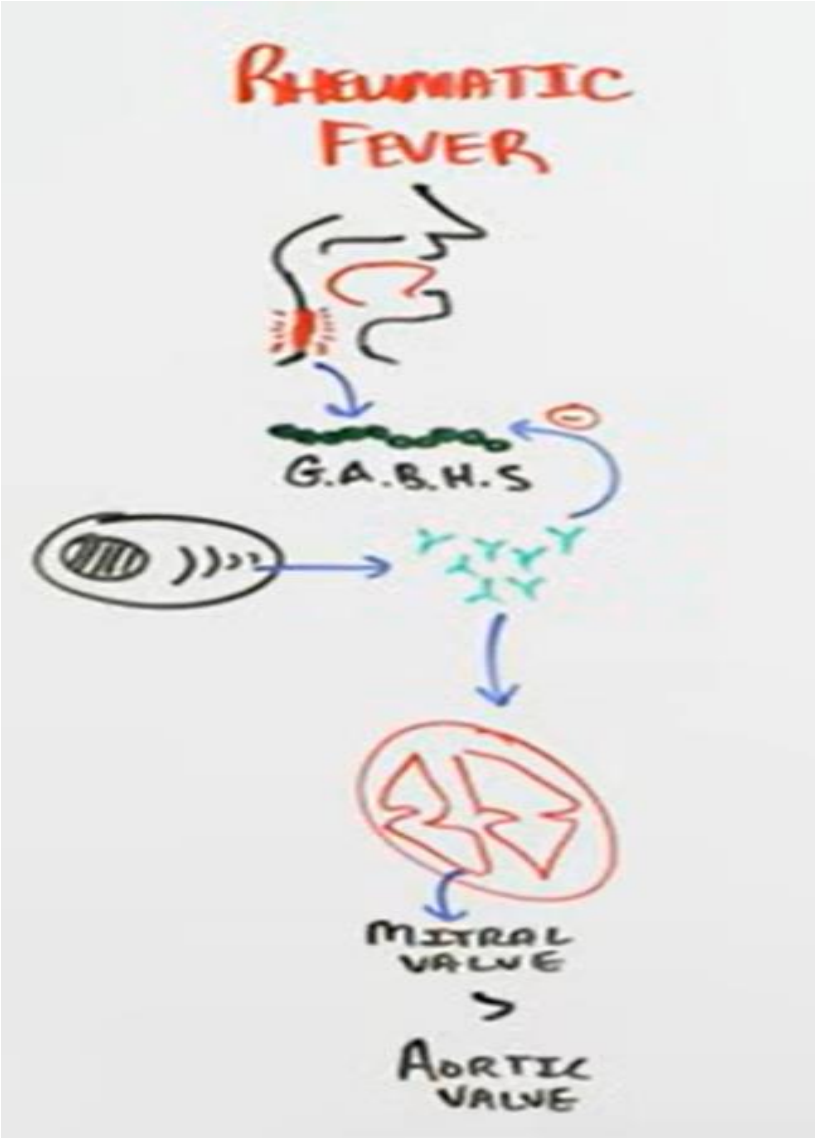
1. **Age:** between 5-15y rare below 3 or above 25y
2. **Sex:** equal. But rheumatic chorea more in females
3. Familial tendency
4. Low economic countries and Over crowding
5. Recurrent streptococcal infections


A sore throat
can lead to a
broken heart!

Pathogenesis

- **An autoimmune disease related to infection with specific strain, group A beta hemolytic Streptococci**

- **Antibodies against streptococcal antigens that cross-react with cardiac proteins as both are immunologically identical.**



Clinical Features

- Acute rheumatic fever is a multisystem disorder that usually presents with fever, anorexia and joint pain, 2–3 weeks after an episode of streptococcal pharyngitis.

- Using the Revised Jones Criteria, the diagnosis is based on two major criteria, or one major and two minor criteria, along with evidence of preceding streptococcal infection.

Clinical Features

Jones criteria for the diagnosis of rheumatic fever

Major manifestations	1. Carditis 2. Polyarthritis 3. Chorea 4. Erythema marginatum 5. Subcutaneous nodules
Minor manifestations	1. Fever 2. Arthralgia 3. Previous rheumatic fever 4. Raised ESR or CRP 5. Leukocytosis 6. First-degree atrioventricular block
Plus	**Supporting evidence of preceding streptococcal infection:** o Recent scarlet fever, o Raised antistreptolysin o titre, o Positive throat culture

Major Criteria

1- Arthritis:

- The commonest major manifestation in 75%
- Polyarticular, asymmetrical affecting big joints as knees, ankles, elbows and wrists in a fleeting manner
- The affected joint is painful, swollen, hot, red & tender with limitation of movement.

Major Criteria

2-Carditis:

- Rheumatic fever causes a **pancarditis** involving the endocardium, myocardium and pericardium to varying degrees. Its incidence declines with increasing age.

- **Symptoms may include;**
  - Breathlessness (due to heart failure or pericardial effusion),
  - Palpitations
  - Chest pain (usually due to pericarditis).

Major Criteria

Myocarditis

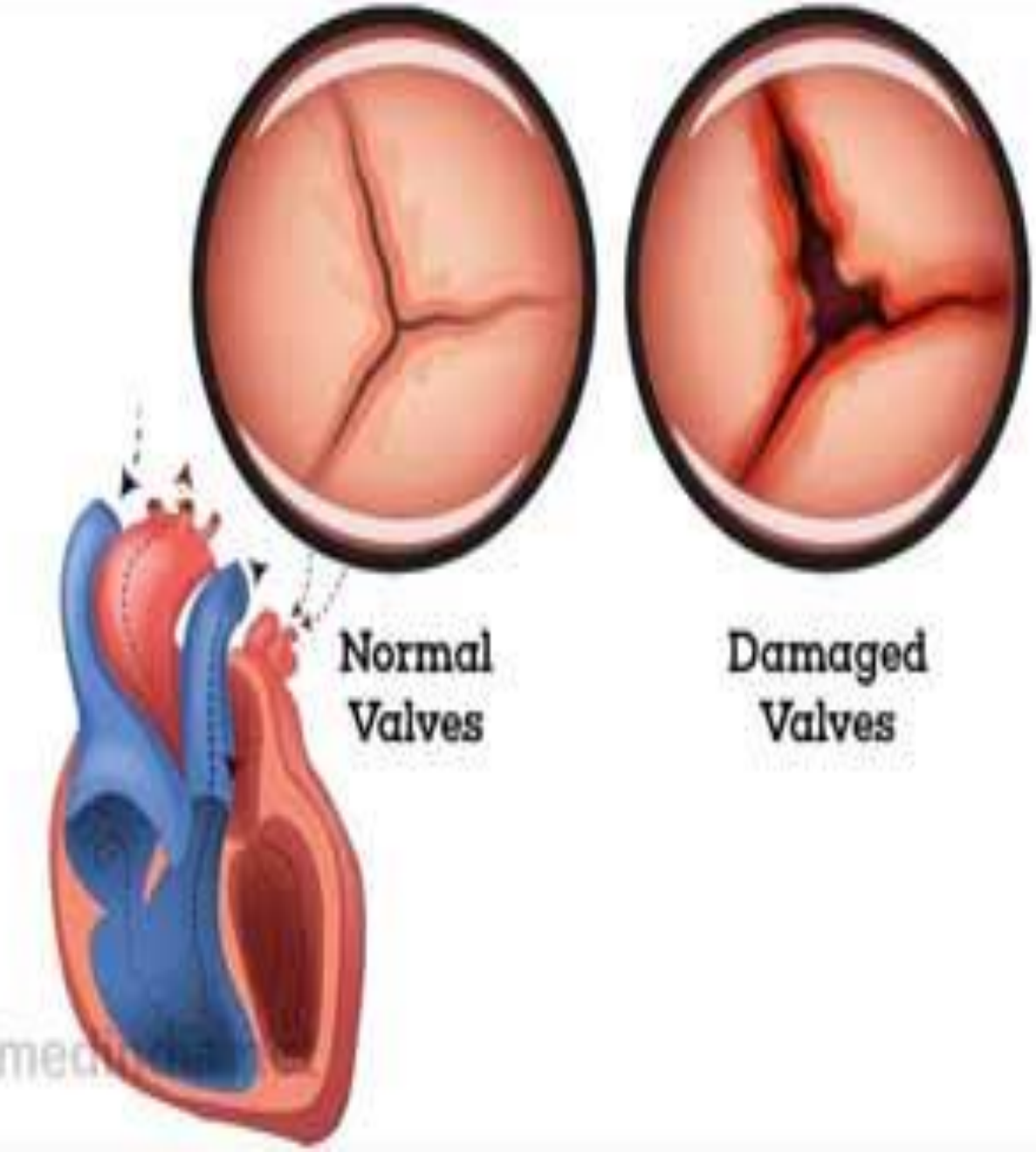
- Disproportionate tachycardia.
- Cardiac dysrhythmias.
- Cardiac enlargement & may be failure.
- Prolonged P~R more than 0.2 second.

Endocarditis:

- Rheumatic valvulits affects mainly the **mitral**, less commonly the **aortic** & least commonly the, 'tricusped & the pulmonary valves.

Pericarditis:

- **Usually dry** less commonly mild effusion may occur rarely it may develop to adhesive pericarditis years later.


Normal
Valves
Damaged
Valves

Major Criteria

3-Sydenham's chorea:

- A late neurological manifestation that appears 3 months after acute rheumatic fever, in form of purposeless, involuntary movements of the hands, feet or face.
- It occurs in one-third of cases and is more common in females.
- Spontaneous recovery usually occurs within a few months.

Major Criteria

4-Subcutaneous nodules:

- It occur in 5–7% of patients.
- They are small (0.5–2 cm), firm not tender and adherent to the deeper structures
- Best felt over extensor surfaces of bone or tendons.
- They appear more than 3 weeks after the onset of other manifestations.

Major Criteria

5-Erythema marginatum:

- Erythema marginatum occurs in less than 5% of patients.
- The lesions start as red macules that appear in crops mainly on the trunk and proximal extremities but not the face.
- They fade in the centre but remain red at the margins that may coalesce or overlap.

Mnemonic: "JONES CAFE PAL"

Major Criteria

J	Joint Involvement
O	**O** looks like a heart = myocarditis
N	Nodules, subcutaneous
E	Erythema marginatum
S	Sydenham chorea

Minor Criteria

C	CRP Increased
A	Arthralgia
F	Fever
E	Elevated ESR

P	Prolonged PR Interval
A	Anamnesis of Rheumatism
L	Leukocytosis

Diagnosis

Throat cultures growing GABHS
OR
Elevated anti-streptolysin O titers

+

2 Major criteria

OR

1 Major criterion and 2 Minor criteria

Treatment

Prophylactic ttt:

1- Treatment of any streptococcal infection.
2- Tonsillectomy for chronically infected tonsils,
3- Long acting penicillin 1.200.00 u/m until the age of 25 5 years after the last attack or for life.

Curative ttt

1- Reset: Complete bed rest till ESR drops to normal
2- Diet: Restrictions of salt, light meals with plenty of proteins
3- Crystalline Penicillin 1 million u/6 h for 10 days to deal with the original septic focus

Treatment

4- Salicylates:

Indications:

1. Rh fever without carditis
2. Patients with contraindications or side effects for corticosteroids.

5- Corticosteroids:

Indications:

1. Rheumatic carditis.
2. If salicylates are not effective or not tolerated.

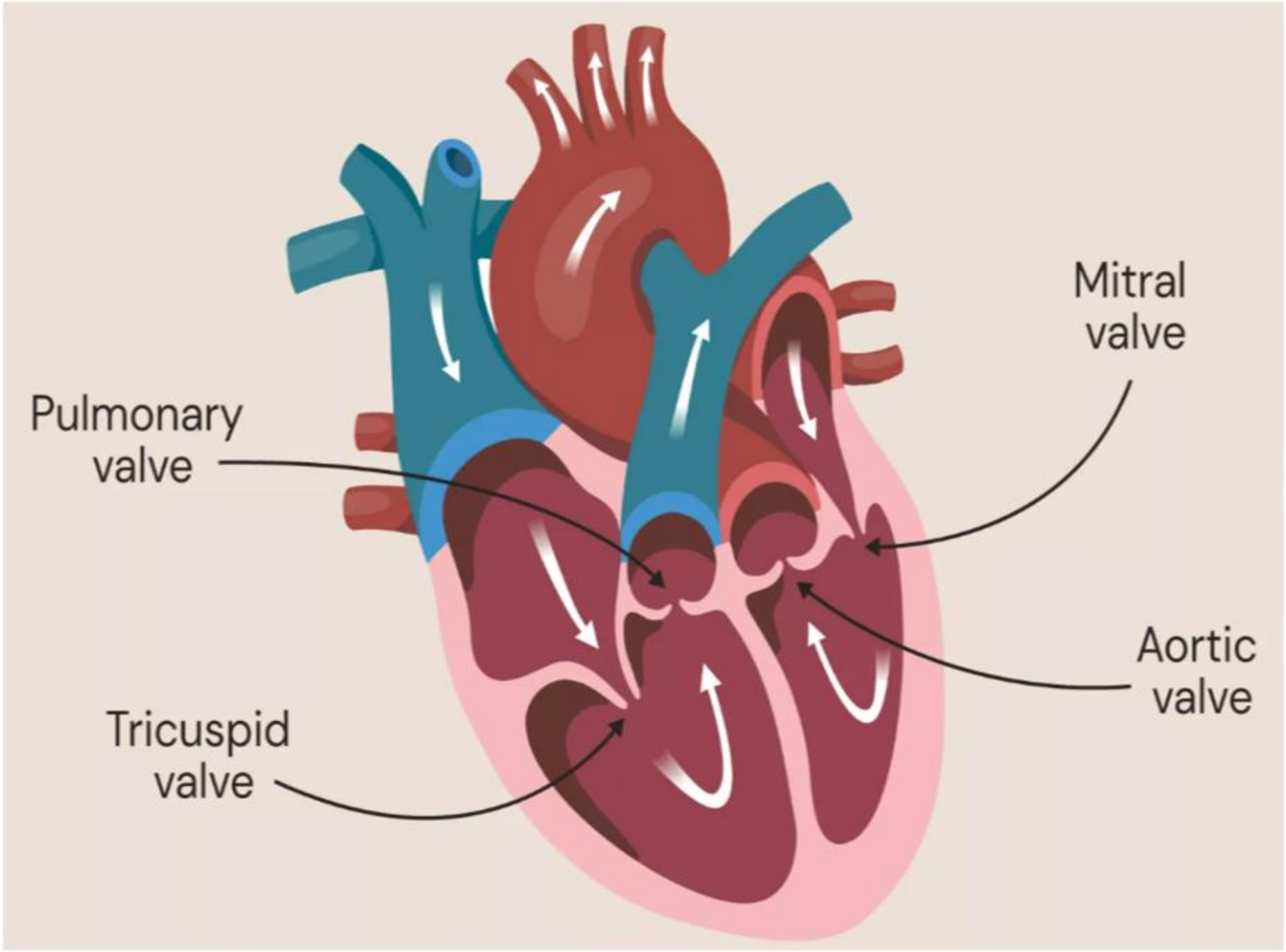
Prednisolone : 50 mg/d for 4 w. gradually diminish to 5 mg/d 'for 4w.

Diseases of the heart valves

- The heart valves **allow forward flow** of blood through the cardiac chambers when they are open and **prevent backward flow** when they are closed.

- <u>A diseased valve may lead to:</u>
  1. Narrow when opened → Obstruction of flow → **Stenosis**
  2. Leaky when closed → backward flow → **Regurgitation** or incompetence.
  3. Both → Stenosis and Regurgitation

- **Breathlessness** and **chest pain** are a common symptom of VHD.


Mitral valve
Pulmonary valve
Aortic valve
Tricuspid valve

Scheme for left sided valvular heart diseases

Aetiology of VHD

MS	AS	MR	AR
1. Rheumatic (The most common) 2. Congenital. 3. Collagen diseases : SLE, RA. 4. Relative (functional)			
		5. Infective endocarditis. 6. Surgical.	
5-Carey Coomb murmur 6 -Austin Flint murmur.	5- Calcification. 6- IHSS.	7- Mitra valve prolapse. 8- Papillary muscle Dysfunction (MI).	7. Syphilis. 8. Dissecting aorta. 9. Marfan syndrome. 10. Severe hypertension.

Scheme for left sided valvular heart diseases

Hemodynamics of VHD

1. Cardiac chambers pressure or volume overload
2. Heart Failure: Right Sided or Left Sided.
3. Cardiac output.
4. PCWP, Pulmonary congestion, Pulmonary hypertension.

- ✓ In general, any stenosis lead to pressure overload on the upstream cardiac chamber
- ✓ whereas regurgitant lesions cause volume overload.

Scheme for left sided valvular heart diseases

Clinical Picture of VHD

Clinical picture of hemodynamics PLUS:

- ✓ MS → 4 Stages. (↑LAP, P congestion , P. HTN, RSHF)
- ✓ AS → Syncope.
- ✓ AR → Peripheral signs of AR (9 signs)
  → Angina.
  → Palpitation , general throbbing.

Scheme for left sided valvular heart diseases

Cardiac examination in VHD

Inspection & palpation:

Apex:

- MS → Slapping apex (weak impulse (due to ↓ LV filling) with palpable S1 .
- AS → Sustained apex (forcible, sustained).
- AR, MR → hyperdynamic apex (forcible, non-sustained).

Pulsation in the 2nd left intercostal space in pulmonary HTN.

Signs of ventricular enlargement

Percussion :

Dullness in the 2nd left intercostal space in pulmonary HTN.

Scheme for left sided valvular heart diseases

Auscultation:

Heart sounds:

- S1 : ↑ in MS, ↓ in MR.
- S2 : ↑ Pulmonary component accentuated in pulmonary HTN

Additional sounds :

① Ejection click (due to Pulmonary HTN)

① Gallop (due to heart failure)

① Opening snap: in MS due to sudden opening of rigid cusps.

Murmur :

- Ejection Systolic → AS
- Pan systolic → MR
- Early diastolic → AR
- Mid diastolic → MS

Scheme for left sided valvular heart diseases

Complications in VHD

1. Calcification.
2. Rheumatic activity.
3. Infective endocarditis. (rare in MS)

4. Arrhythmia e.g. AF in a case of MS , heart block in calcified AS.
5. Thromboembolism: stroke.
6. LA enlargement → compression on :
   - Lung → dyspnea & cough.
   - Esophagus → dysphagia.
   - Left recurrent laryngeal nerve → hoarseness of voice.

Scheme for left sided valvular heart diseases

7. Pulmonary congestion → Hemoptysis.
8. Pulmonary infection (recurrent).
9. Pulmonary embolism (secondary to DVT)

10. RSHF.
11. LSHF except in MS.

12. Complications of surgery (artificial valves) :
    - Mechanical dysfunction.
    - Infective endocarditis.
    - Thromboembolism.
    - Hemolytic anemia.

Scheme for left sided valvular heart diseases

Investigations For VHD

X ray:

- Chamber enlargement.
- Pulmonary congestion.
- Pulmonary hypertension

ECG:

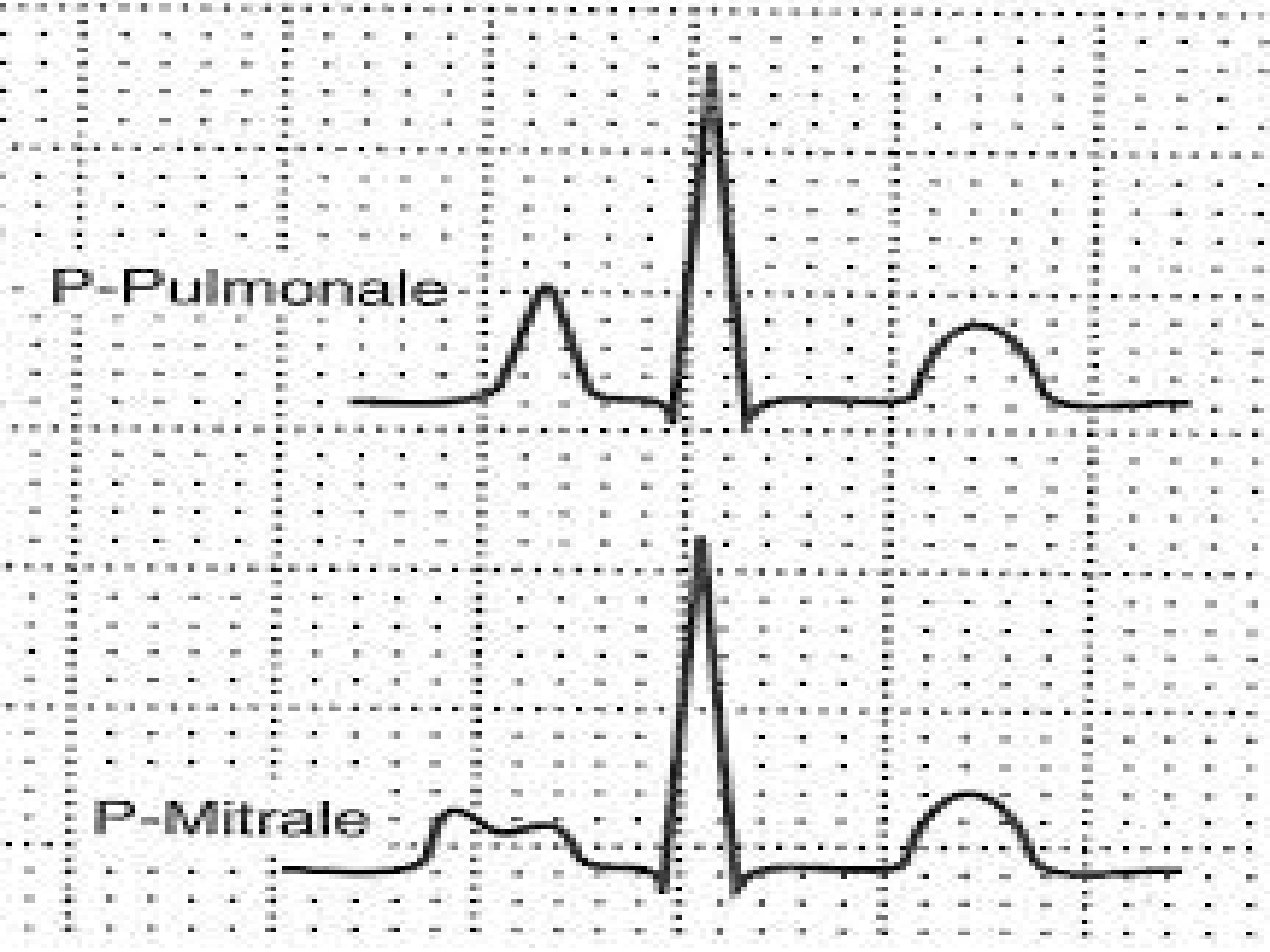
- Chamber enlargement e.g. LA → P mitrale (m shaped P wave)
- Pulmonary hypertension → P pulmonale (Peaked P wave)
- Arrhythmias

Echo & Doppler echo : (The most important)

- Chamber enlargement.
- Detect the severity of the valve lesion.

Catheterization & angiography :

- Detect the severity.
- Chamber enlargement.


P-Pulmonale
P-Mitrale

Scheme for left sided valvular heart diseases

Treatment of VHD

Medical

1- Prophylaxis against Infective Endocarditis & rheumatic activity.

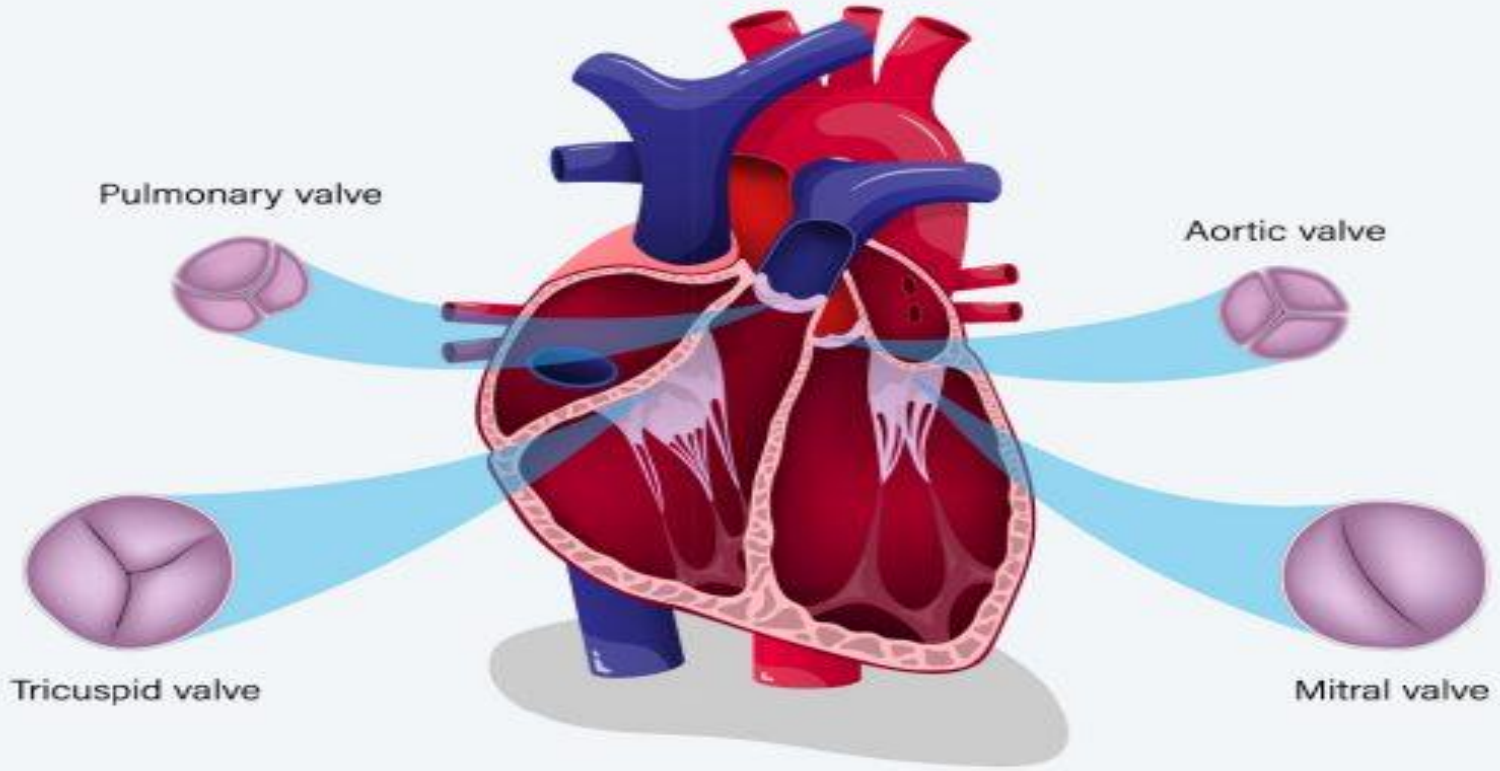
2- Treatment of complications e.g. HF, AF, infections ...

Curative

1- Balloon dilatation for stenosis especially pure MS.

2- Valvotomy: Commissurotomy for stenosis & Repair for regurge.

3- Valve replacement : Tissue or synthetic valves.


Pulmonary valve
Aortic valve
Tricuspid valve
Mitral valve

Mitral stenosis

Anatomy of Mitral valve

Site: between the LA & LV.

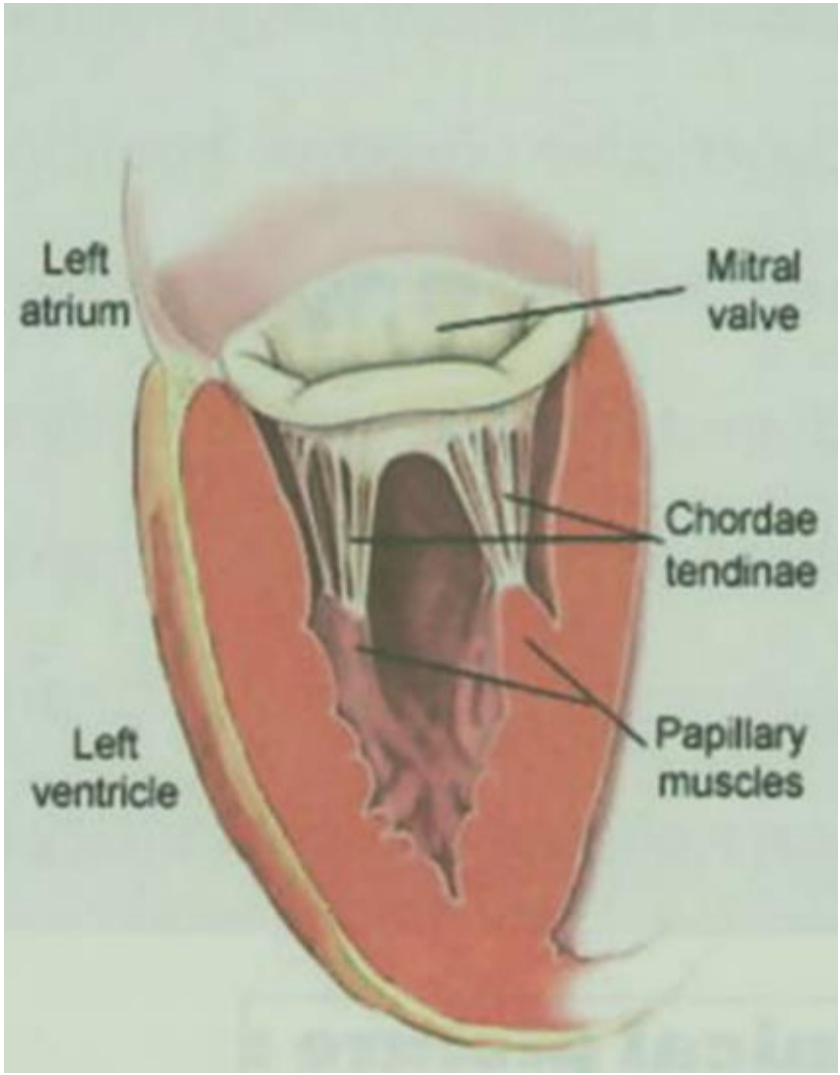
Surface area : 4 - 5 cm^2

MS if mitral valve orifice < 2.5 cm^2

if< 1 cm ~ tight MS.

Components:

- ✓ Fibrous ring.
- ✓ 2 Cusps (anteromedial & posterolateral)
- ✓ 2 Papillary muscles : arise from the ventricle, to control the cusps movement.
- ✓ Chordae tendinae : arise from papillary muscles to both cusps.



Mitral stenosis

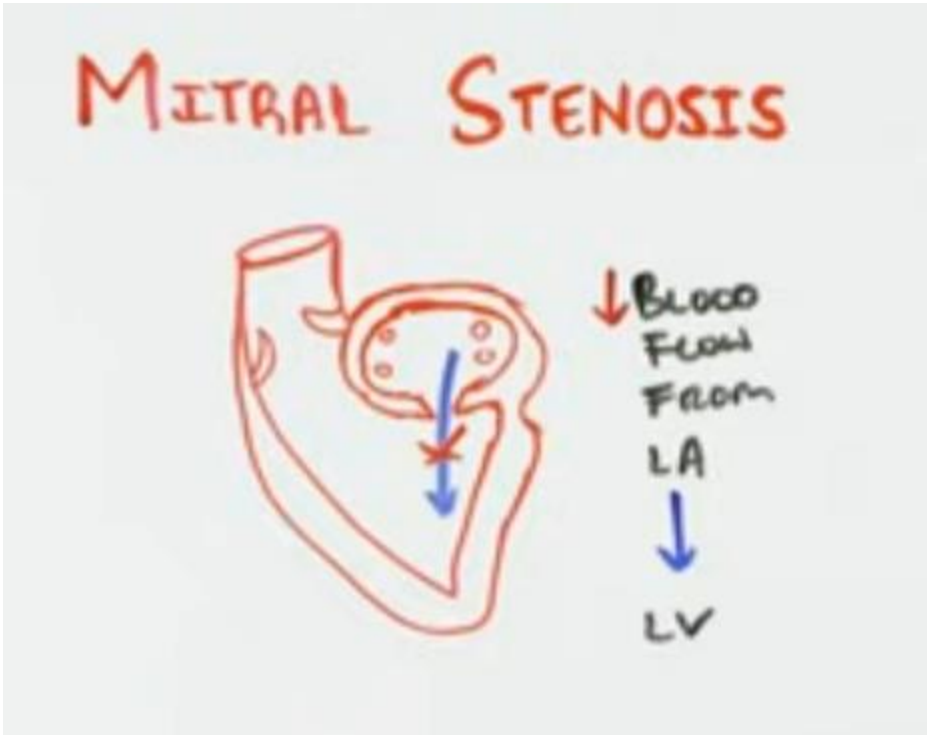
Etiology

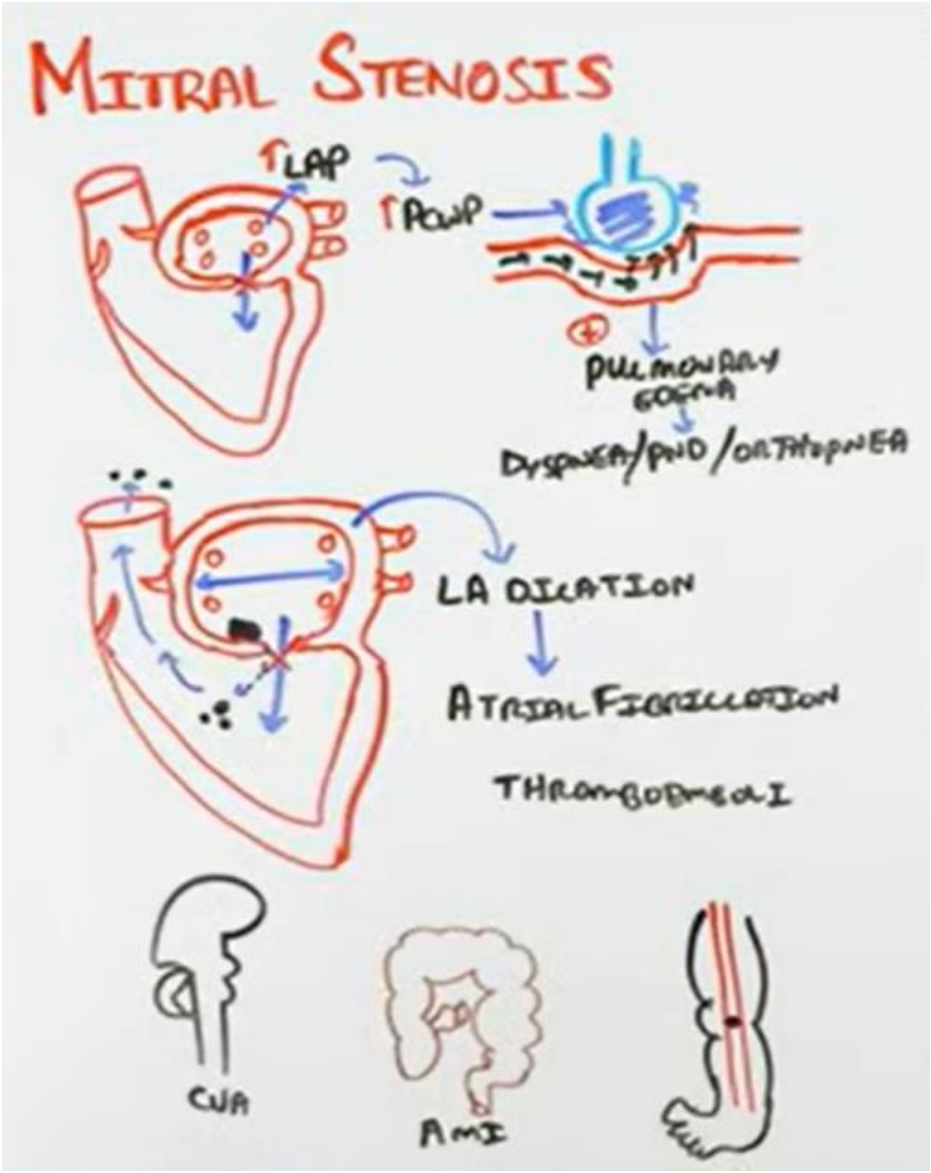
- **Rheumatic heart disease :**
  1. The commonest cause (99%).
  1. Occurs years after the original attack
  1. Usually associated with multi valvular lesions.

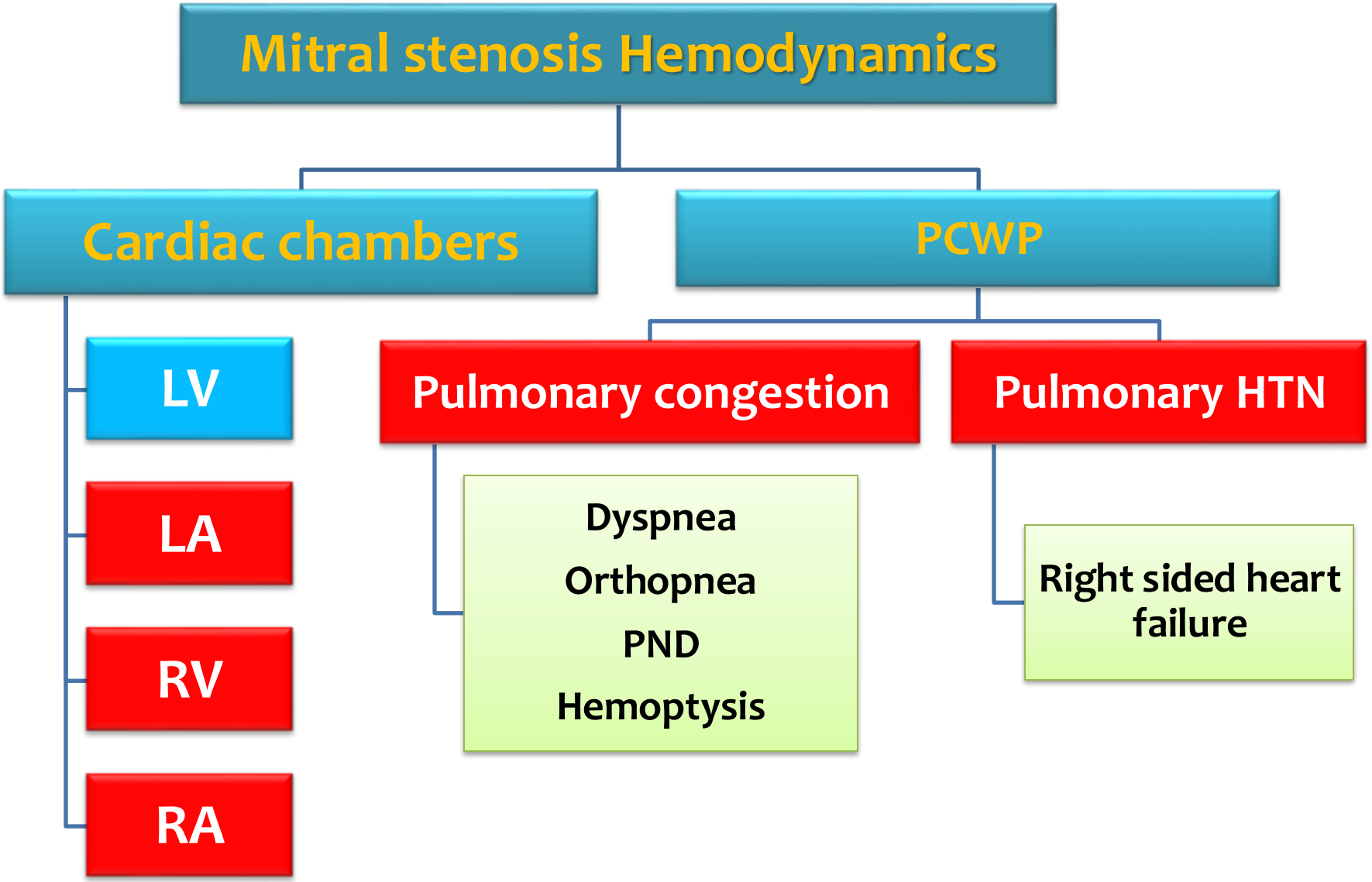
- **Relative :**
  - **Carrey coomb's murmur:** in acute stage of rheumatic fever due to edema of the cusps → transient narrowing of the mitral valve.
  - **Austin-Flint murmur:** murmur of MS in sever AR (The regurged blood during diastole interferes with opening of mitral valve).
  - ↑ **blood flow** through the mitral valve : MR, VSD, PDA.

Mitral stenosis Hemodynamics

During diastole: ↓ blood flow through the mitral valve ↑ pressure load on LA → LA dilatation → AF and Pulmonary congestion.






Mitral stenosis Hemodynamics
Cardiac chambers
PCWP
LV
LA
RV
RA
Pulmonary congestion
Pulmonary HTN
Dyspnea
Orthopnea
PND
Hemoptysis
Right sided heart failure

Mitral stenosis

4 stages

1- ↑ LA Pressure with dilatation

↓

2- Back pressure on pulmonary vein (pulmonary congestion)

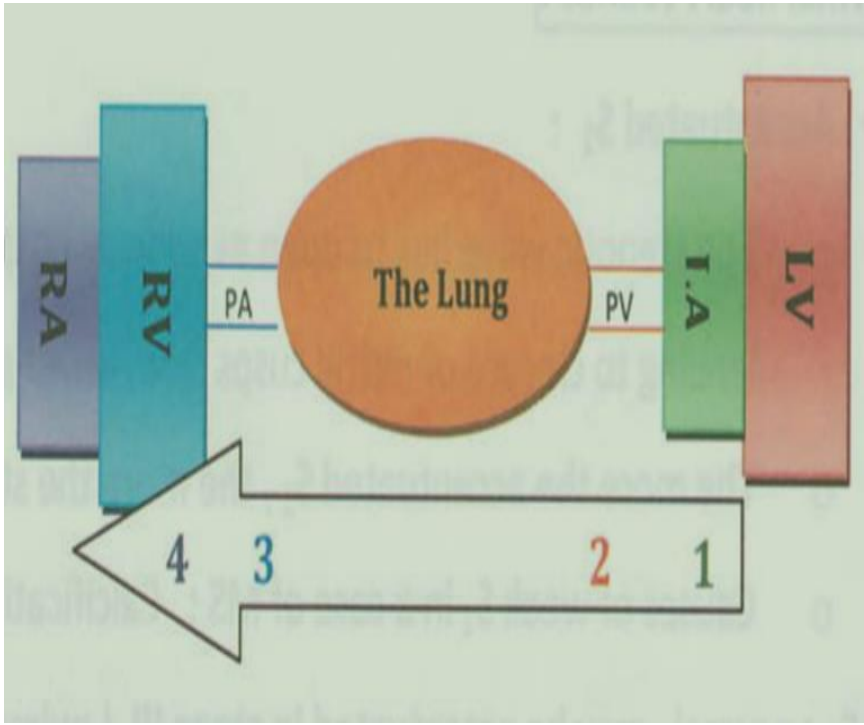
↓

3- Pulmonary hypertension

- Passive
- Constrictive → reflex VC of pulmonary arterioles
- Obstructive → PE

↓

4- RSHF



Mitral stenosis

Clinical picture :

- **Stage I :** asymptomatic (just ↑ of LA pressure)
- **Stage II :** manifestations of pulmonary congestion
- **Stage III :** manifestations of pulmonary hypertension : LCOP , Malar flush,
- **Stage IV:** manifestations of RSHF : LCOP, systemic congestion.

Mitral stenosis

Murmur:

- **Site:** best heard at the apex.
- **Propagation :** No propagation (localized)
- **Timing:** Mid diastolic with pre systolic accentuation due to atrial contraction .
- **Character:** rumbling, low pitched murmur.
- **Relation to respiration & position:** ↑ with expiration & ↓ in left lateral position.
  - Left sided heart murmurs are ↑on expiration .
  - Right sided heart murmurs are ↑on inspiration .

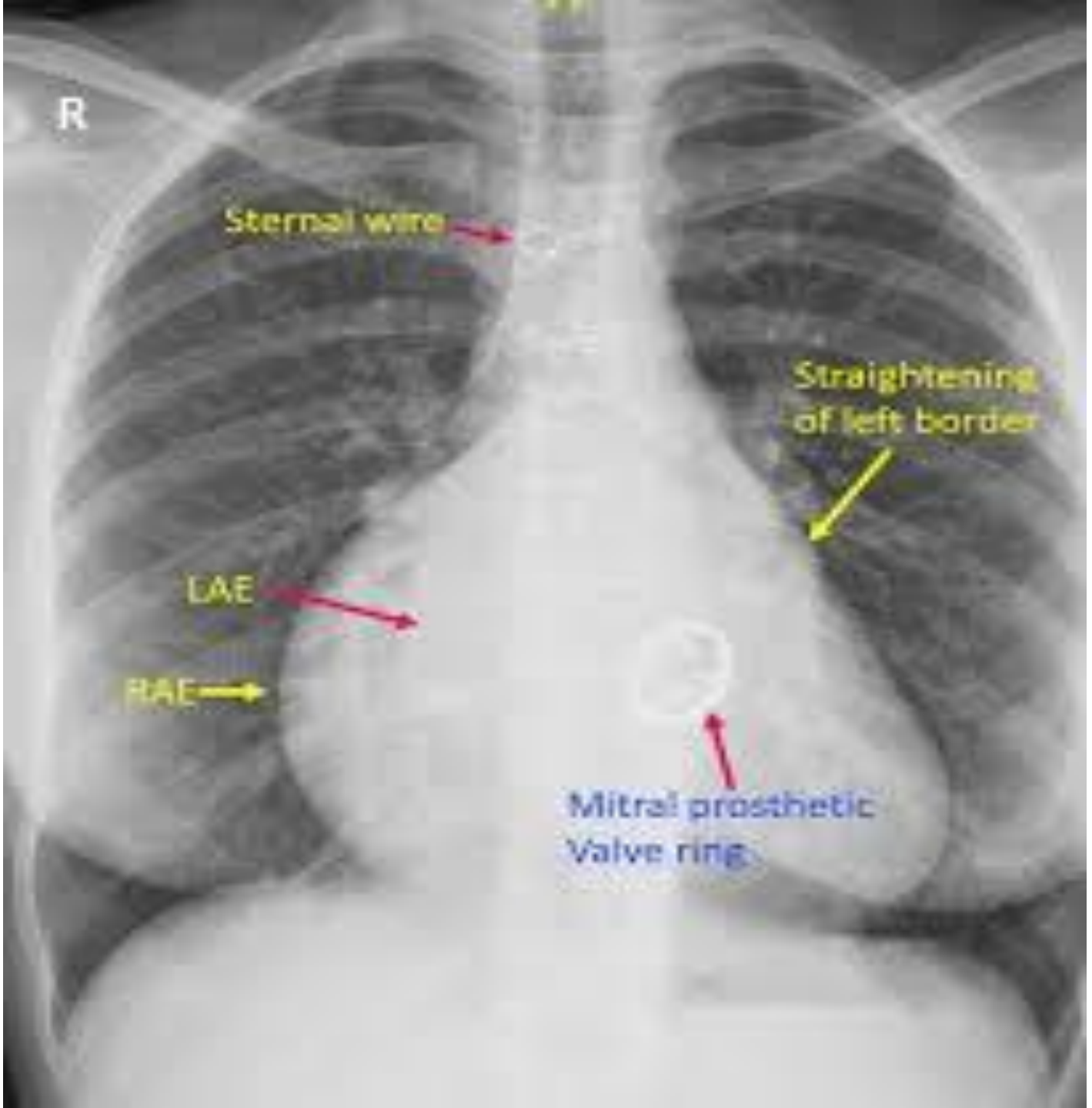
Mitral stenosis

Complications: as
Investigations: as
Treatment: as

Indications of valve replacement :

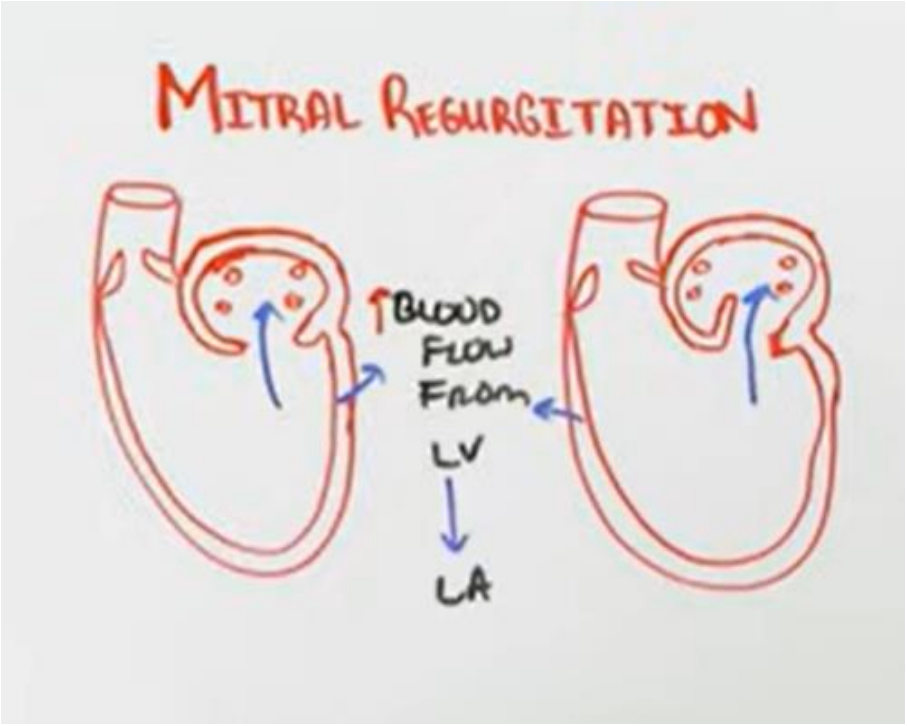
1. Calcification
2. Associated MR
3. Tight MS (surface area < l cm^2 or severe manifestations)
4. Recurrent stenosis after balloon dilatation or valvotomy .

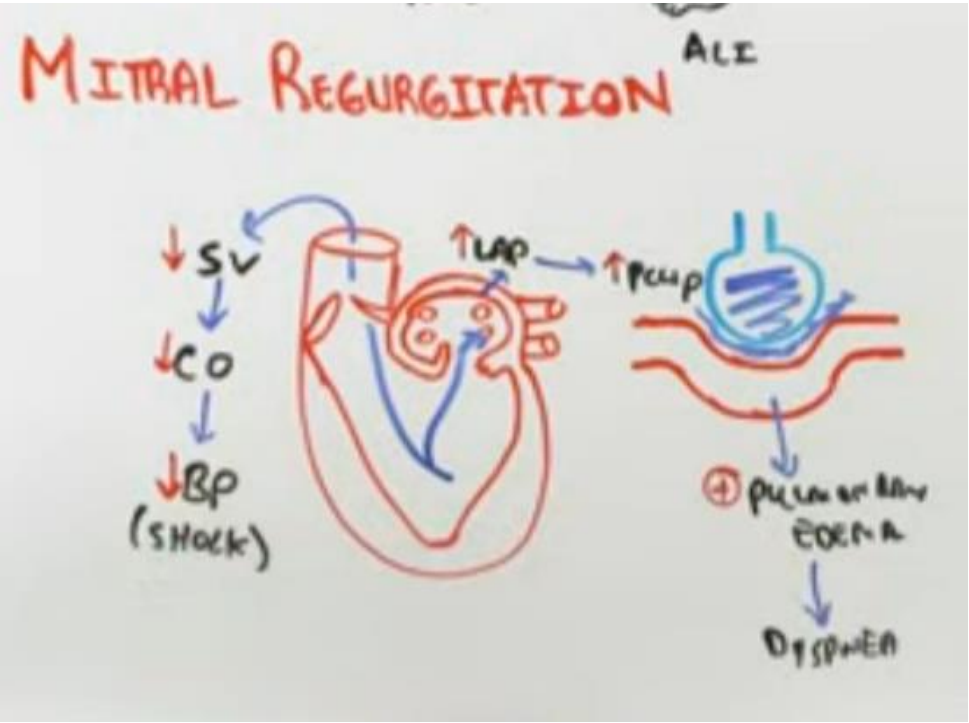

PA
L

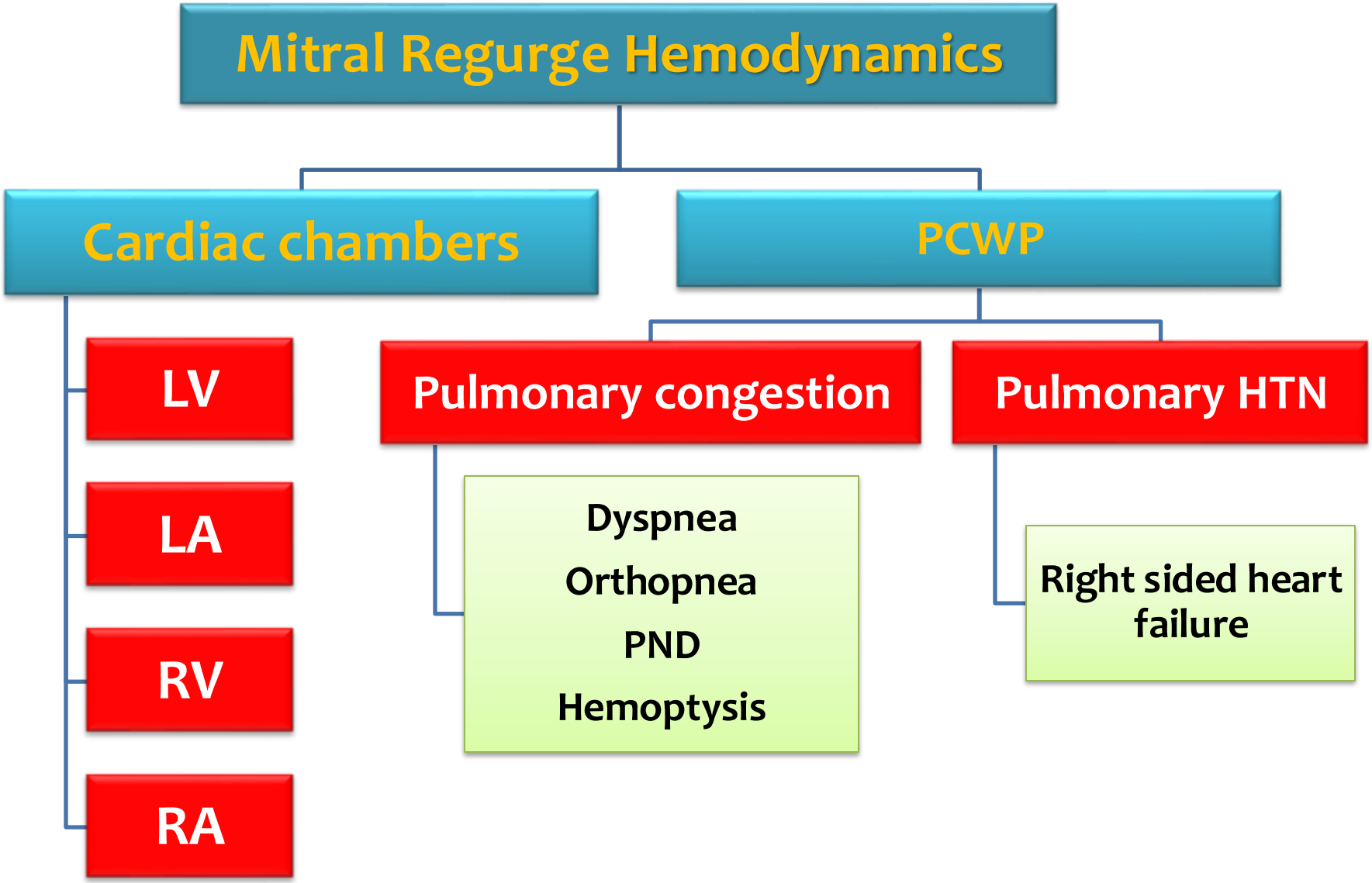

R
Sternal wire
Straightening of left border
LAE
RAE
Mitral prosthetic Valve ring

Mitral Regurge Hemodynamics

- **During systole:** A part of blood regurgitates from LV to LA → LA dilatation and pulmonary congestion.
- **During diastole:** ↑ blood flow through the mitral valve → volume overload on LV → LV enlargement then failure.






Mitral Regurge Hemodynamics
Cardiac chambers
PCWP
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Pulmonary congestion
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Orthopnea
PND
Hemoptysis
Right sided heart failure

Mitral Regurge

Murmur:

- **Site:** best heard at the apex.
- **Propagation :** to axilla
- **Timing:** Pansystolic murmur. (plateau).
- **Character:** blowing, high pitched.
- **Relation to respiration & position:** ↑ with expiration & ↓ in left lateral position.

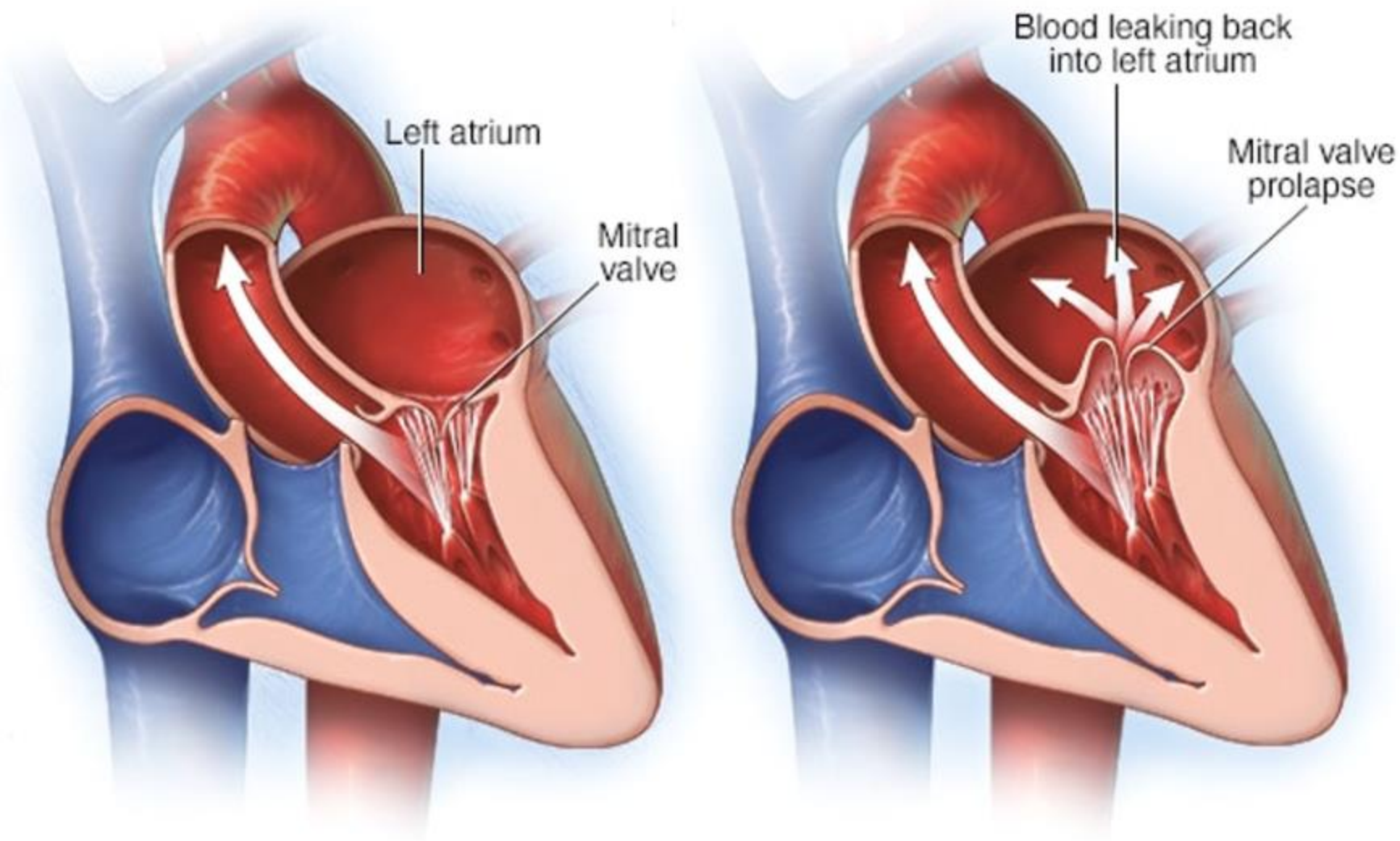

L
Semi-Upright

Mitral Valve Prolapse

Definition: Prolapse of one or both cusps of mitral valve into LA during systole.

Etiology

1. **Idiopathic:** in most cases, more common in young female.
2. **Connective tissue diseases :**
   - ① Marfan syndrome.
   - ① SLE.
   - ① Polyarthritis nodosa.
3. **Muscle disorders:** Duchenne myopathy, Myotonia dystrophy.
4. **Congenital heart diseases:** e.g. ASD
5. **Acquired heart diseases :** MI, post mitral valve surgery.



Typical heart

Mitral valve prolapse with regurgitation

Mitral Valve Prolapse

Clinical Picture

- Asymptomatic in most cases
- Atypical chest pain:
  - The most common symptom.
  - Usually it is left inframammary & stabbing.
  - Sometimes it is severe substernal aching pain.

- Palpitation : due to abnormal ventricular contraction & arrhythmias.

- Dizziness or Fainting .
- Fatigue
- Shortness of breath

Mitral Valve Prolapse

Cardiac examination :

- The most common sign is a **mid-systolic click**, which is produced by the sudden prolapse of the valve & the tension of the chordate tendineae.
- This may be followed by a **late systolic murmur** due to some regurgitation.
- With more regurgitation, the murmur becomes Pansystolic.

Mitral Valve Prolapse

Investigations : Echo is diagnostic

Treatment:

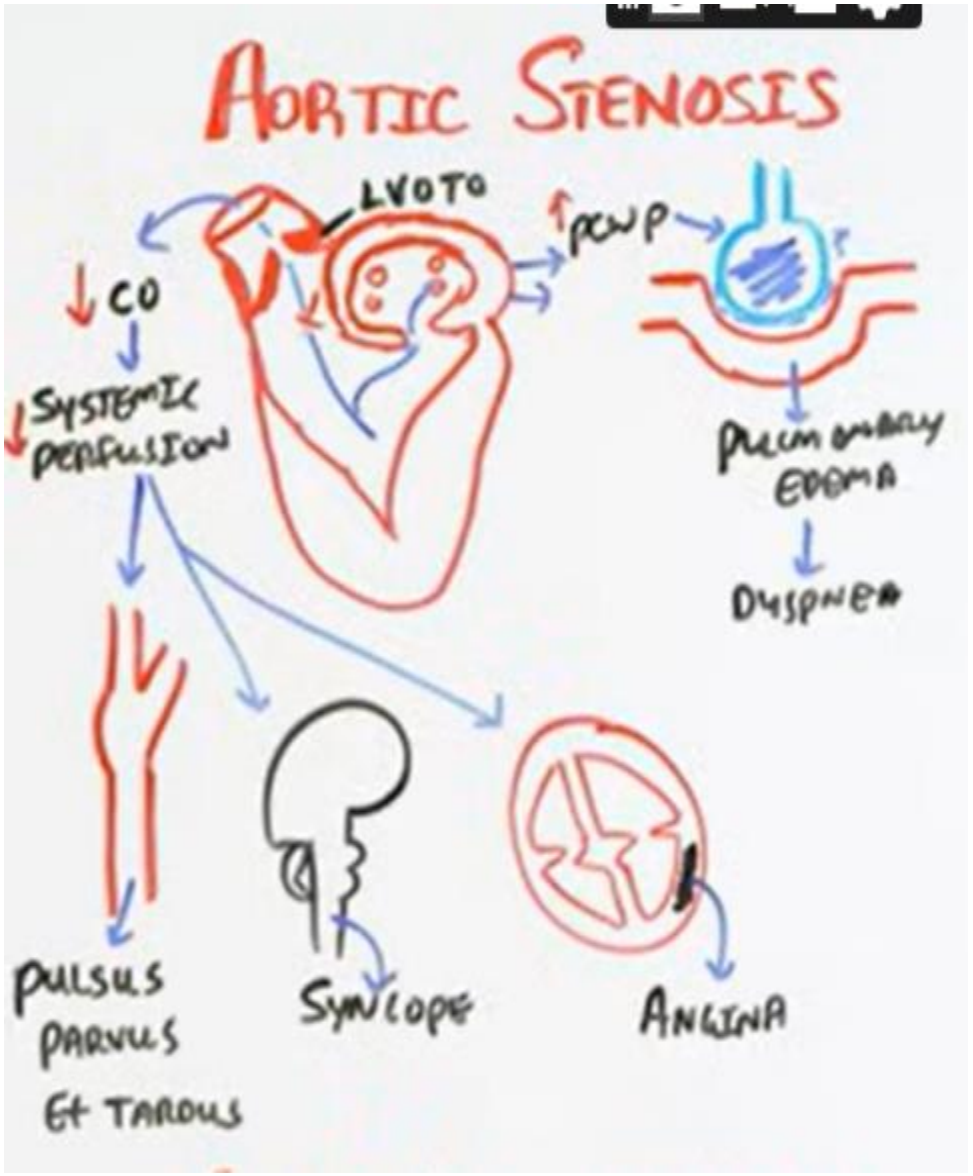
- Reassurance.
- B-blocker e.g. propranolol.
- Valve replacement in severe cases

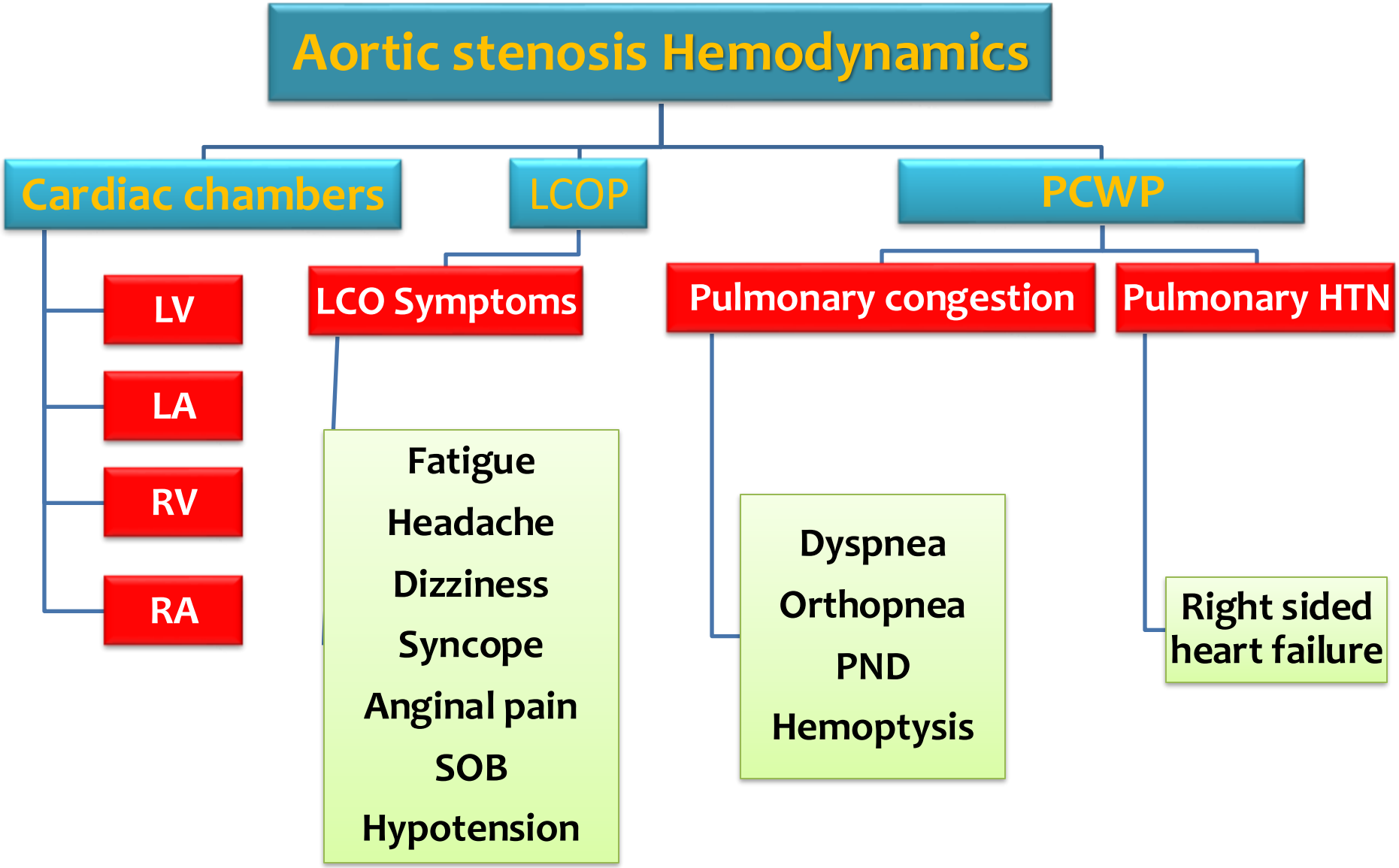
Aortic stenosis Hemodynamics

During systole:

There is obstruction of LV outflow results in:

- o LCOP.
- o Pressure overload on LV leading to LVH.




Aortic stenosis Hemodynamics
Cardiac chambers
LCOP
PCWP
LV
LA
RV
RA
LCO Symptoms
Pulmonary congestion
Pulmonary HTN
Fatigue
Headache
Dizziness
Syncope
Anginal pain
SOB
Hypotension
Dyspnea
Orthopnea
PND
Hemoptysis
Right sided heart failure

Aortic Stenosis

Murmur:

- **Site:** maximum over A1 area (2nd right intercostal space).
- **Propagation :** neck (carotid arteries) & apex.
- **Timing:** Ejection (mid) systolic murmur (diamond-shaped, crescendo decrescendo).
- **Character:** Harsh but may be soft in relative AS.
- **Relation to respiration & position:** ↑ with expiration & ↓ in left lateral position.
  - Left sided heart murmurs are ↑on expiration .
  - Right sided heart murmurs are ↑on inspiration .

Aortic stenosis

Treatment: as

Indications of valve replacement :

1. Valve area < 0.8 cm2
2. Systolic pressure gradient across the aortic valve> 50 mm Hg.
3. Severe symptoms.

► Balloon dilatation & aortic Valvotomy (associated with a high early restenosis rate)

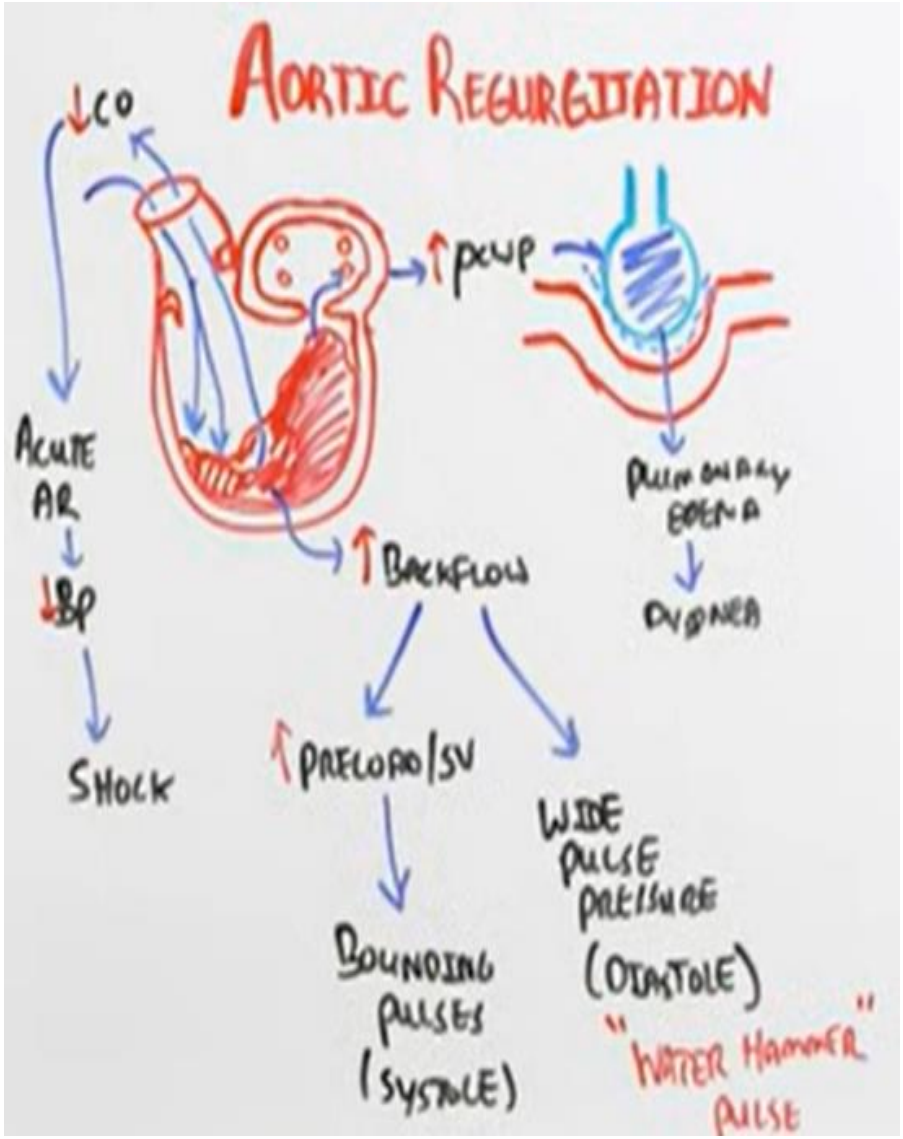
Aortic Regurge Hemodynamics

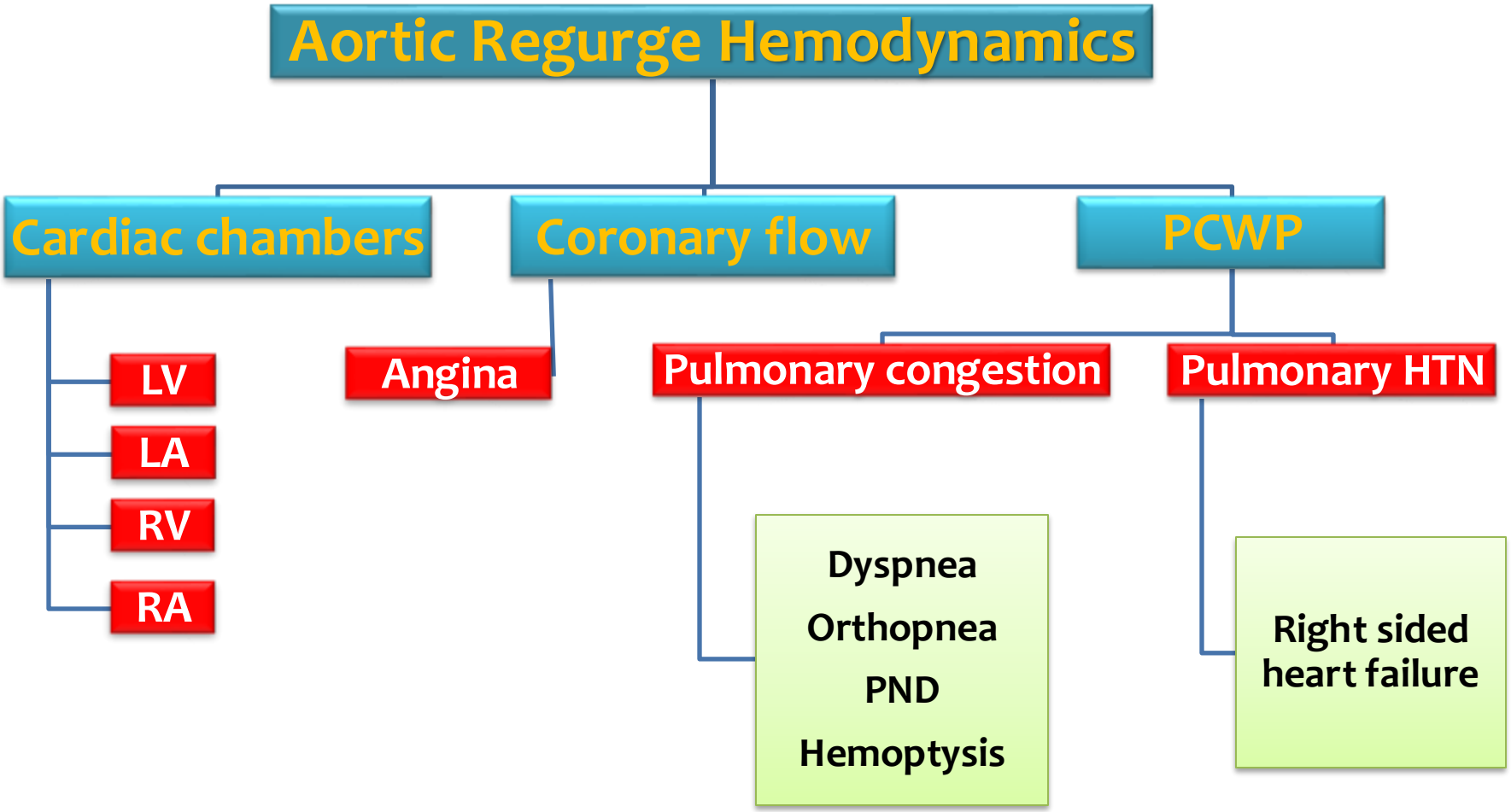
During diastol:

There is regurgitation of blood from the aorta to the LV leading to:

- ► Volume overload on the LV.
- ► ↓ Coronary blood flow → Angina.
- ► ↑Blood in LV → ↑LV stroke volume ↑ Systolic BP which is compensated by peripheral VD

- ► ↓ Diastolic BP : due to peripheral VD & regurgitation of blood during diastole.




Aortic Regurge Hemodynamics
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Aortic Regurge

Clinical picture

1. General throbbing : due to transmitted arterial pulsation.

2. Angina due to :
   - Diastolic BP ↓ coronary blood flow.
   - LV hypertrophy ↑ 02 demand.

3. Manifestations of LSHF: Pulmonary congestion & LCOP.

Aortic Regurge

Peripheral signs of AR : (due to big pulse volume)

- De Musset sign : nodding of the head.
- Corrigan's sign : Marked visible carotid pulsation.
- Systolic thrill over the carotid artery.

- Pulse : Water hammer pulse.
- Capillary pulsations : pressing on the nail tip → moving red line.

- Pistol shots : systolic femoral sound due to sudden distension of collapsed artery.
- Hill's sign : The difference between systolic BP in LL & UL> 50 mmHg. (Normally SBP in LL> UL by 10 - 20 mmHg)

Aortic Regurge

Murmur:

- **Site:** Best heard over the left lower sternal border, around the 3rd (A2 area) and 4th intercostal spaces.
- **Propagation :** To apex
- **Timing:** Early diastolic.
- **Character:** Soft blowing, decrescendo.
- **Relation to respiration & position:** ↑ with expiration & with leaning forward.

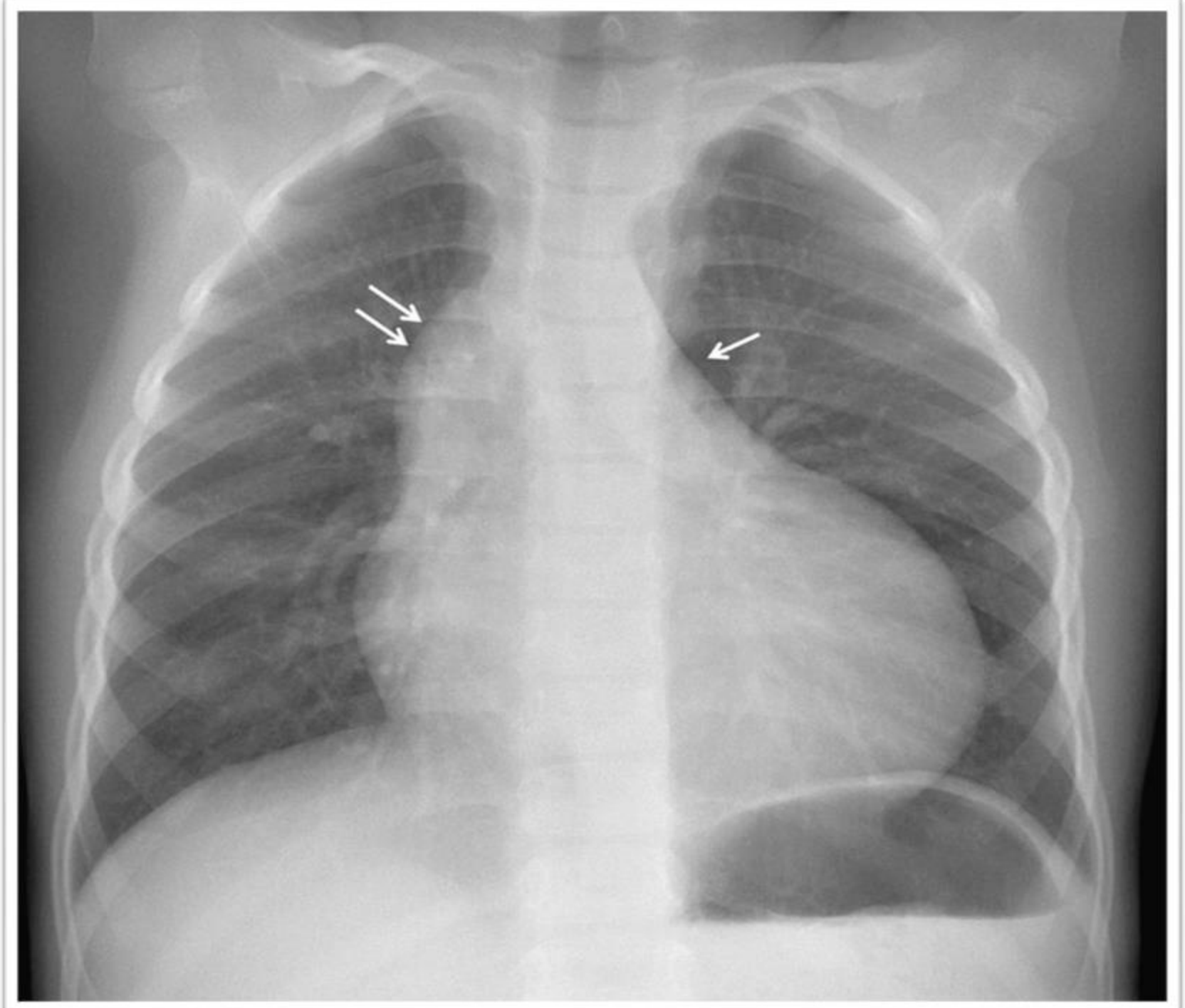
Aortic Regurge

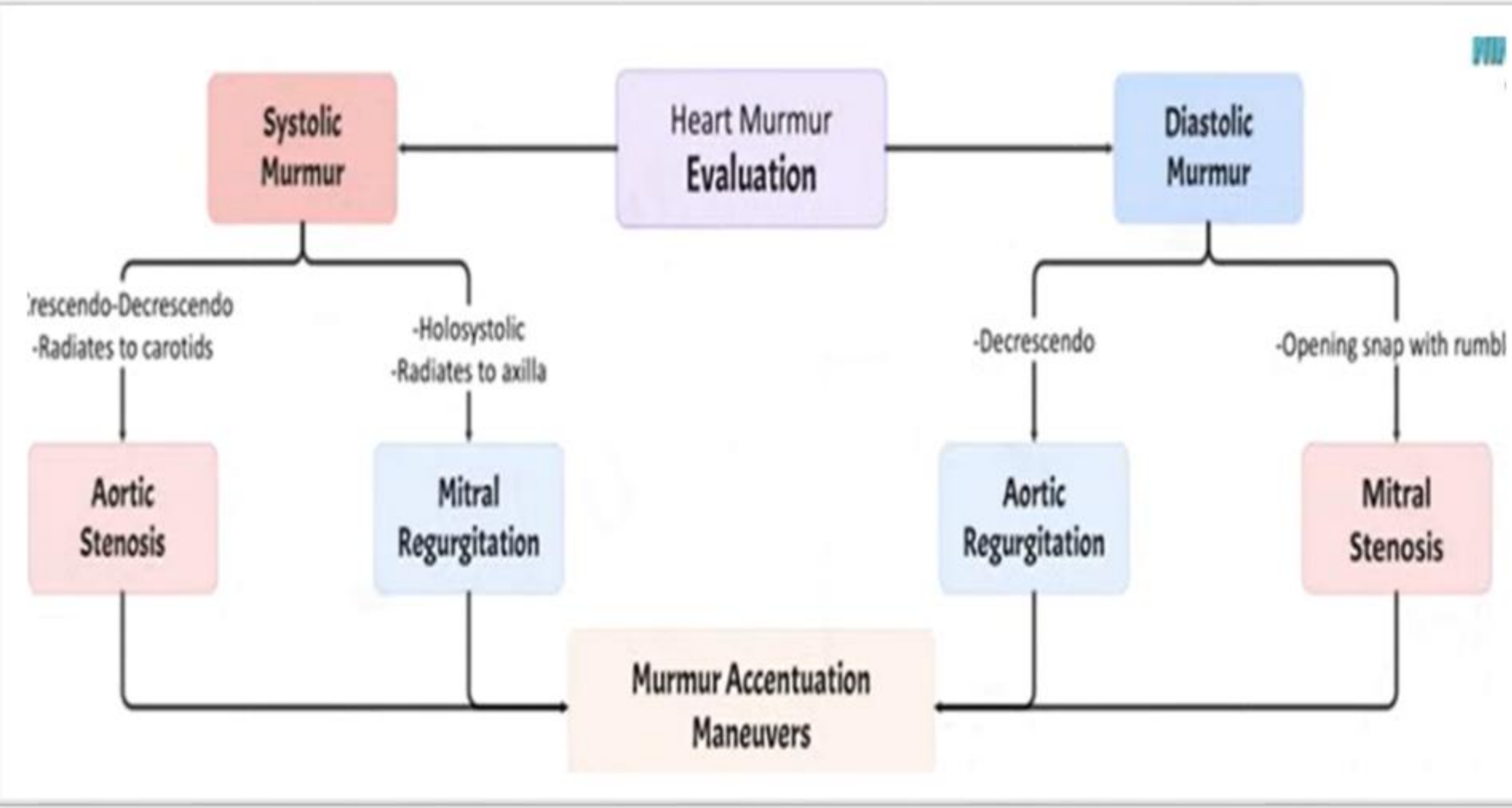
Investigations :

- **X ray:** LVE & dilated aorta (prominent right border)
- **ECG:** LVE.
- **Echo:** LVE, detects the severity of the valve lesion.
- **Catheterization:** Detects the severity of the valve lesion.

Treatment :

- **Medical:** As scheme.
- **Surgical:** Valve replacement in severe cases with LV dysfunction.




Heart Murmur Evaluation
Systolic Murmur
Diastolic Murmur
rescendo-Decrescendo
-Radiates to carotids
-Holosystolic
-Radiates to axilla
-Decrescendo
-Opening snap with rumbl
Aortic Stenosis
Mitral Regurgitation
Aortic Regurgitation
Mitral Stenosis
Murmur Accentuation Maneuvers

Tricuspid Stenosis

Etiology :

- It's usually rheumatic in origin & usually associated with mitral or aortic valve diseases.

Hemodynamics : obstruction of tricuspid valve leading to :

- ↑ RA pressure → RA enlargement & systemic congestion.
- ↓ RV filling → ↓ COP.

Tricuspid Stenosis

Clinical picture:

- ► Symptoms of LCOP and systemic congestion.
- ► Signs of LCOP:
  - ✓ Cold hands with weak pulse
  - ✓ ↓ Systolic BP
  - ✓ Peripheral cyanosis.
- ► Signs of systemic congestion:
  - ✓ Congested pulsating neck vein with systolic expansion and giant A wave.
  - ✓ Enlarged tender pulsating liver with mild jaundice.
  - ✓ Ascites and LL edema.
- ► Signs of RA & RV enlargement.
- ► Mid diastolic presystolic murmur at lower left sternal border, increases by inspiration.

Tricuspid Regurge

Etiology :

- TR is usually functional resulting from RVE → dilatation of tricuspid ring.

Hemodynamics :

During systole, part of blood regurgitates from RV to RA causing:

- ↑RA pressure → RA enlargement & systemic congestion.
- ↓ RV output → LCOP.
- RV enlargement then failure.

Tricuspid Regurge

Clinical picture:

- Symptoms and signs of LCOP and systemic congestion.
- Systolic thrill over tricuspid area.
- Pansystolic murmur over tricuspid area & propagated to the apex, and increases by inspiration.

Pulmonary Stenosis

Etiology :

- ► Anatomy :
  - Valvular : the most common type (80 %) .
  - Subvalvular (Infundibular)
  - Supravalvular: rare.

Hemodynamics :

During systole: ↓ blood flow through the pulmonary valve → ↑

Pressure overload on RV leading to :

- ► LCOP.
- ► RV enlargement then failure. & systemic congestion.
- ► ↓ Right CO to the lung → lung oligemia → predispose to TB.

Pulmonary Stenosis

Clinical picture:

- ► Symptoms and signs of LCOP and systemic congestion.
- ► Systolic thrill over pulmonary area.
- ► Weak pulmonary component of S2 with wide splitting.
- ► Ejection click in valvular type.
- ► Murmur : ejection systolic murmur on pulmonary area.

Pulmonary Stenosis

Treatment:

- Prophylaxis against infective endocarditis
- Treatment of RSHF
- Surgical : in severe PS, " the pressure gradient across the pulmonary valve : > 50
  - Valvular type : valvotomy or replacement.
  - Subvalvular type : resection of infundibulum.

Systolic Murmurs

DD of Systolic Murmurs

1. AS
2. PS
3. MR
4. TR.
5. VSD
6. PDA
7. Coarctation of aorta

"All of the above 7 lesions may propagate to the apex."