Valvular Heart Disease

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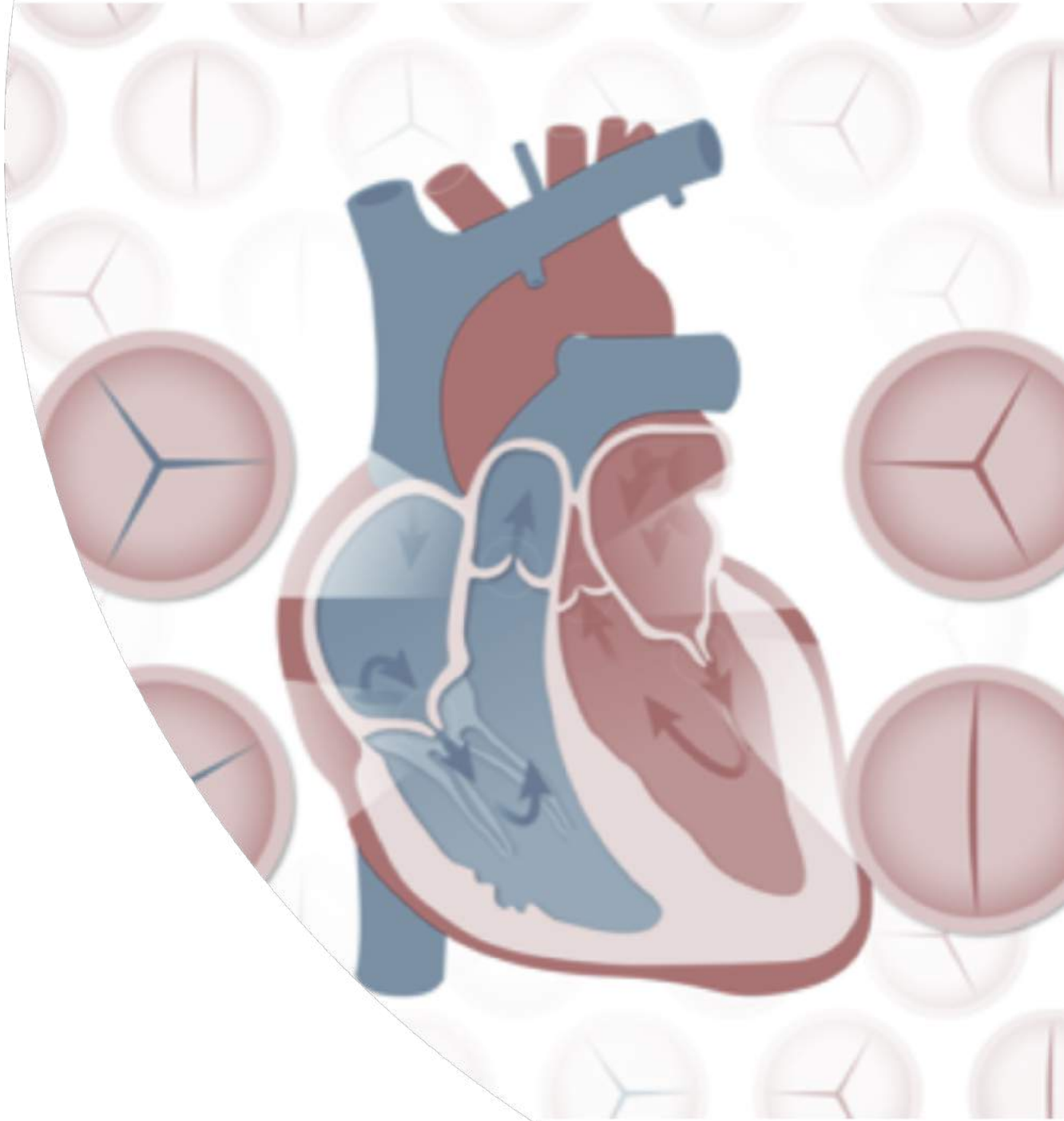


Objectives

- Describe the etiology, pathology, and natural history of valvular heart disease.
- Describe the clinical symptoms and signs of valvular heart disease.
- Explain the clinical examination findings of particular valvular problems.
- Determine the role of echocardiograms in valvular heart disease, both in diagnosis and prognosis.
- Discuss the long-term systemic consequences of valvular heart disease.
- Describe the management and identify the indications of surgical intervention for particular valvular heart diseases.

Agenda

- What are heart valves
- Anatomy
- Native and prosthetic heart valves
- Definition of valvular heart disease
- Types of pathology
- Pathophysiology
- Stages of VHD
- Diagnosis & Workup
- Prophylaxis with RF & IE.
- Overview of the main pathologies
- Management

What are heart valves?

- Cardiac structures that maintain continuous free forward blood flow without backward leakage.

- **There are four main cardiac valves divided into 2 groups:**

❑ **Atriventricular valves:**

✓ Mitral valve

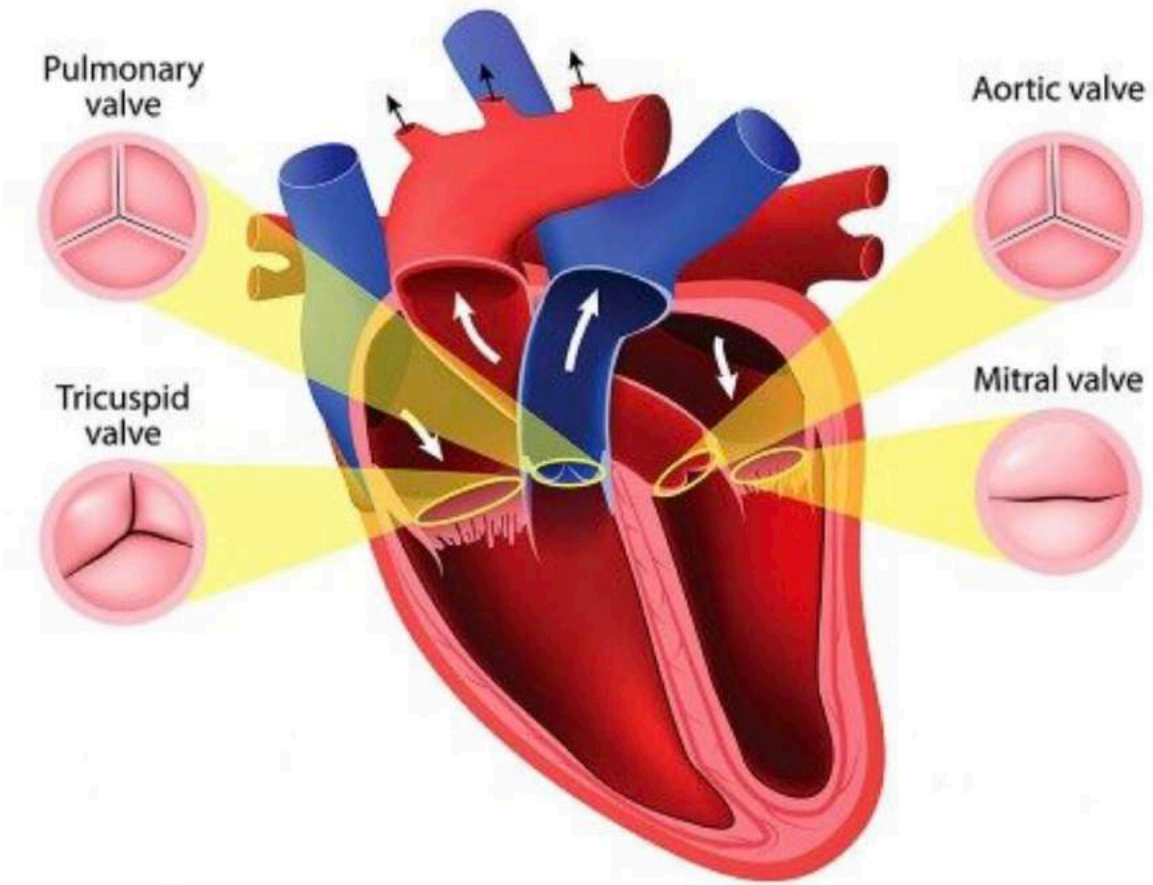
✓ Tricuspid valve

❑ **Semilunar valves**:

✓ Aortic valve

✓ Pulmonic valve

Anatomy



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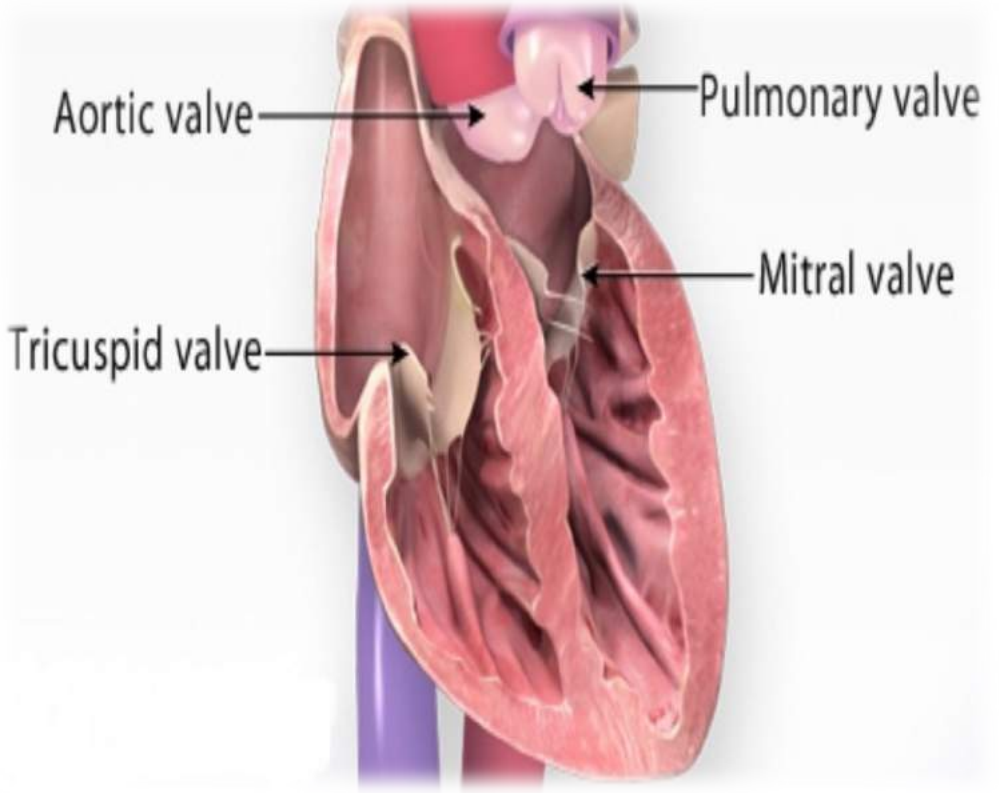
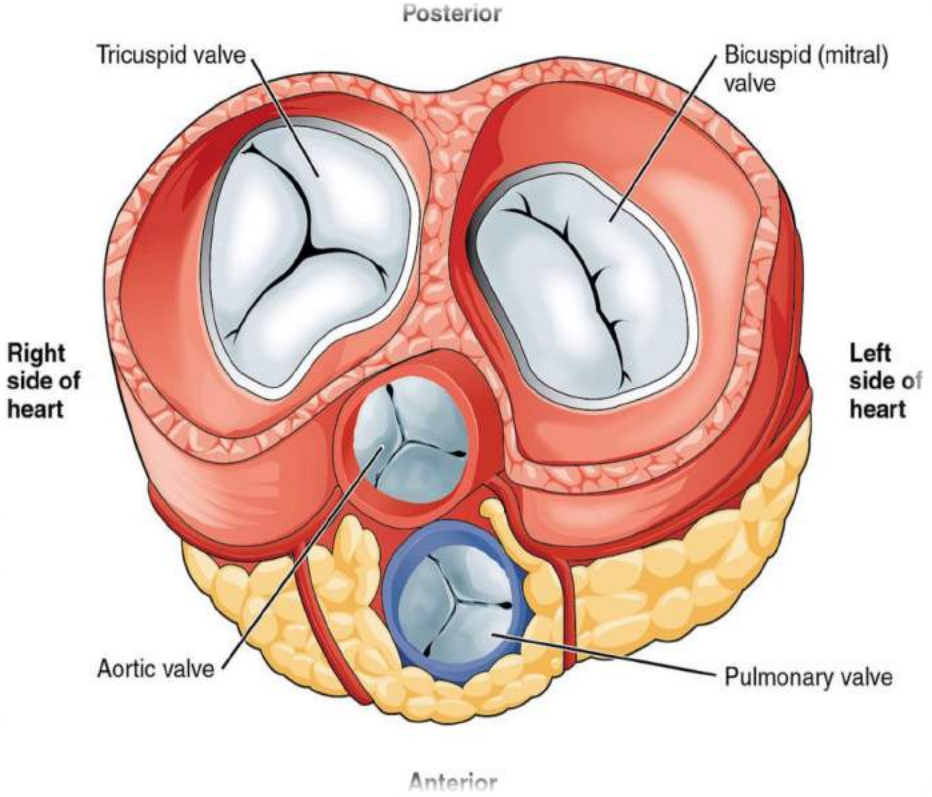
Anatomy





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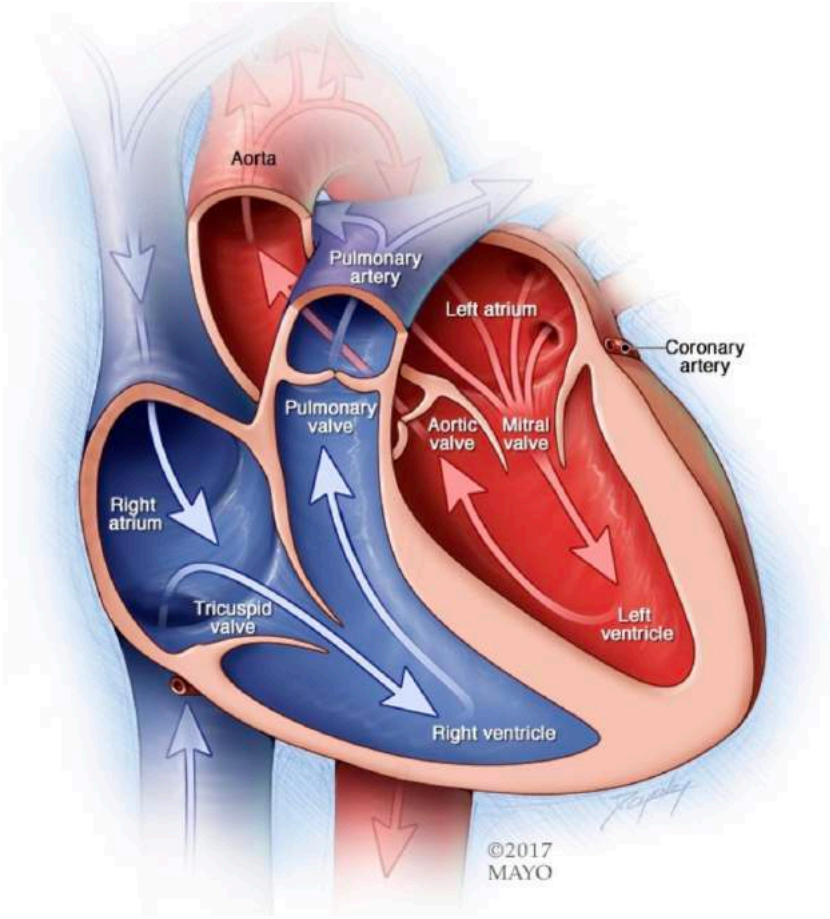
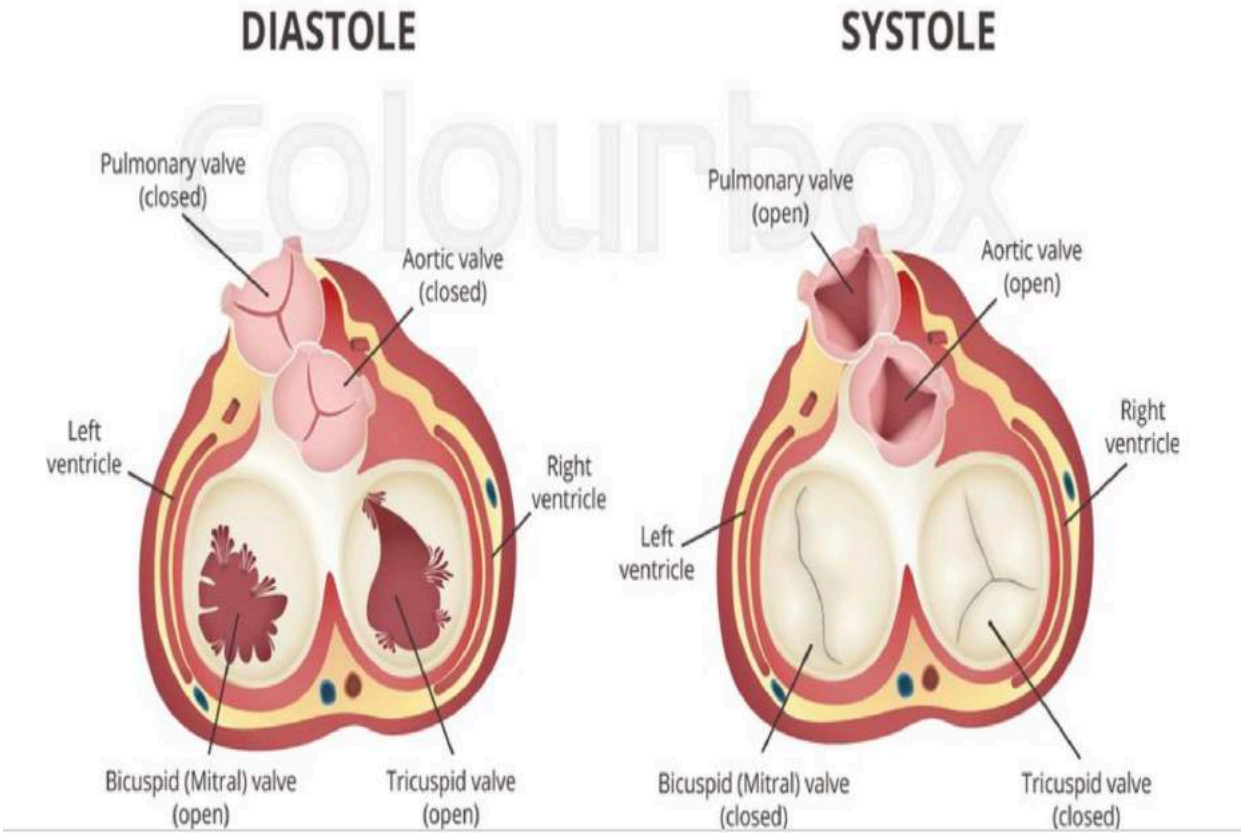
Valve function





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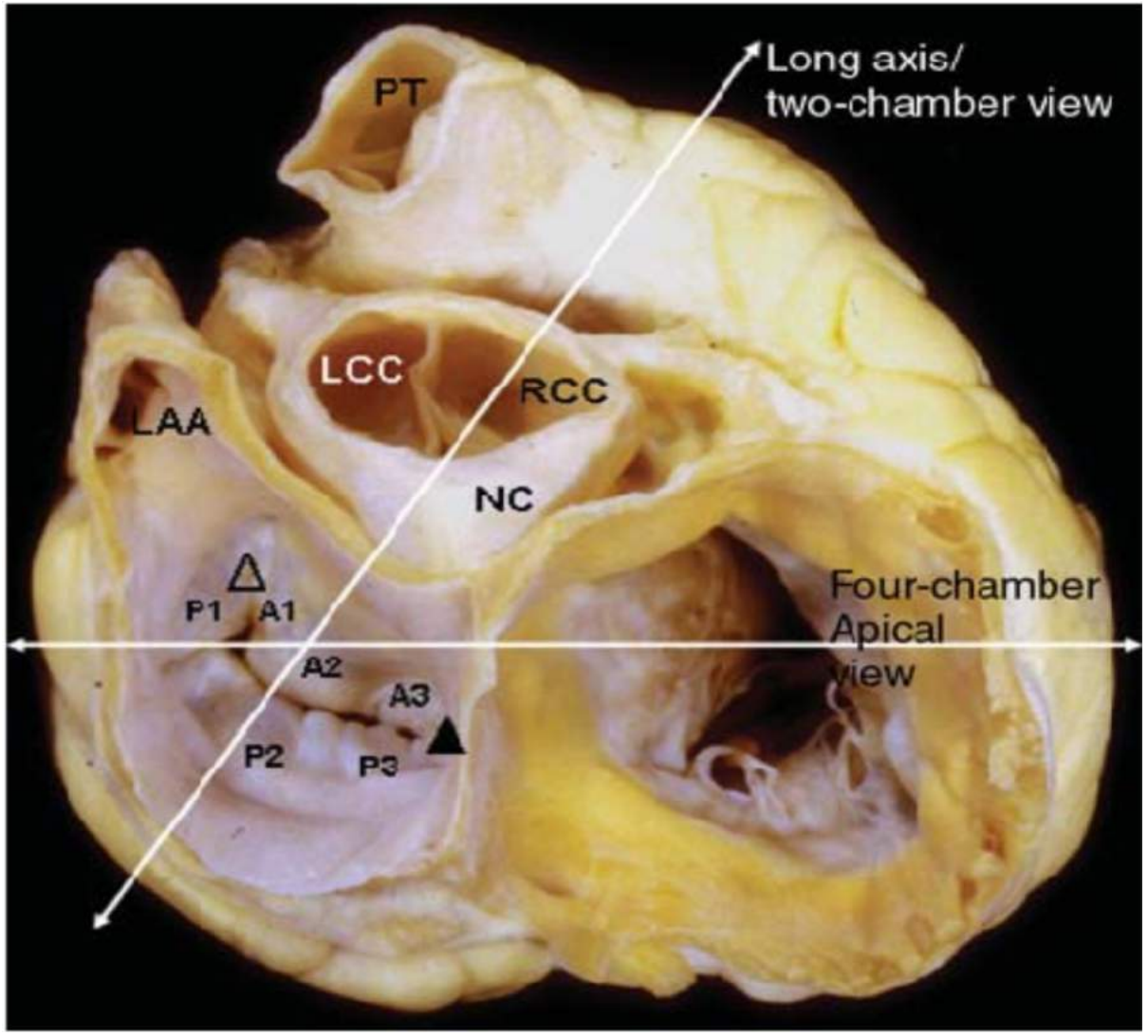
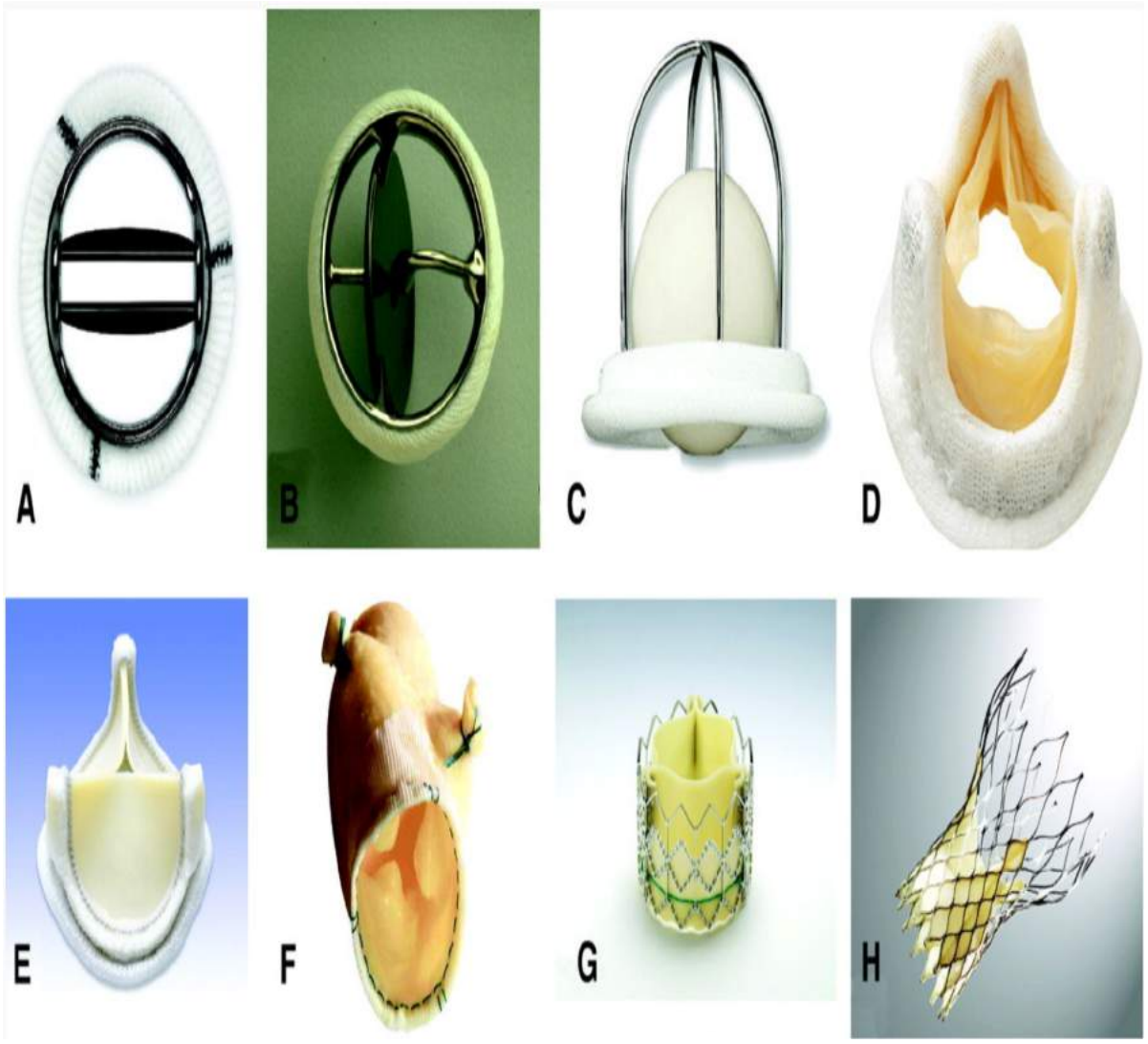
Native & prosthetic valves





What is Valvular Heart Disease (VHD) ?

Acquired or congenital cardiac abnormality of the heart valves that interfere with their normal function

Types of pathology

Stenosis

- Narrowing that leads to obstruction of flow

Regurgitation

- Backward leakage of blood

Etiology of VHD

Stenosis	Regurgitation
Congenital	Congenital
Rheumatic	Rheumatic
Degenerative / Senile	Degenerative e.g myxomatous, calcification
Drugs	Infective endocarditis
Radiation	Valve ring dilatation e.g Dilated Cardiomyopathy
	Infections e.g syphilis
	Traumatic
	Ischemia

Stages of VHD

Stage	Definition	Description
A	At risk	Patients with risk factors for development of VHD
B	Progressive	Patients with progressive VHD (Mild-Moderate) (Asymptomatic)
C	Asymptomatic Severe	Asymptomatic but reached the criteria of severe VHD C1: Asymptomatic with compensated cardiac function C2: Asymptomatic but decompensated cardiac function.
D	Symptomatic Severe	Developed symptoms secondary to VHD

Types of Presentations

Acute	Chronic
e.g Acute mitral regurgitation due to eg acute myocardial infarction acute chordea tendineae rupture	e.g Chronic mitral regurgitation due to eg RHRUMATIC fever . Mitral valve Prolapse . e.g Chronic aortic regurgitation due to eg Bicuspid Aortic valve

Hemodynamic consequences

Pressure Overload	Volume Overload
Aortic stenosis Left Ventricular hypertrophy Mitral stenosis Left Atrial hypertrophy & dilatation	Chronic mitral regurgitation Dilated left ventricle & left atria Chronic tricuspid regurgitation Dilated right ventricle & right atria

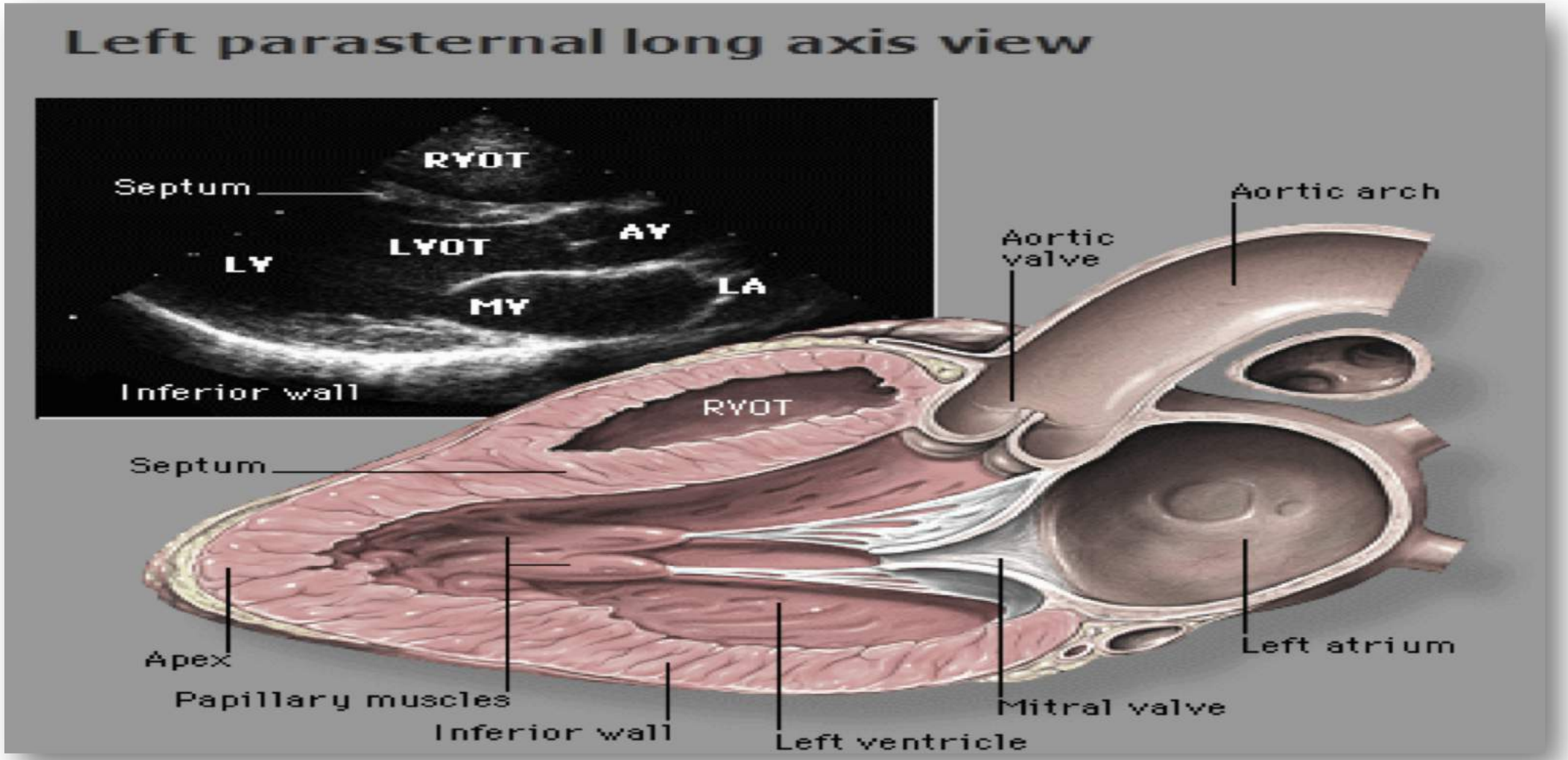
Presentation

Symptoms	Signs
Dyspnea , paroxysmal nocturnal dyspnea orthopnea .	Abnormal look (Mitral facies)
Palpitations	Abnormal pulse (AF)
Chest pain	Abnormal JVP
Dizziness, presyncope, syncope	Apex beat abnormality
Edema, ascites	Parasternal heave
Cough	Thrill
Fatigue	Abnormal heart sounds
Hemoptysis	Extra heart sounds
Symptoms of thromboembolic complications	Murmurs (systolic or diastolic)

Dx & workup

- ECG .
- CXR .
- Echocardiography
- Holter monitor.
- MRI .
- Cardiac catheterization.
- Exercise test.

Echocardiography



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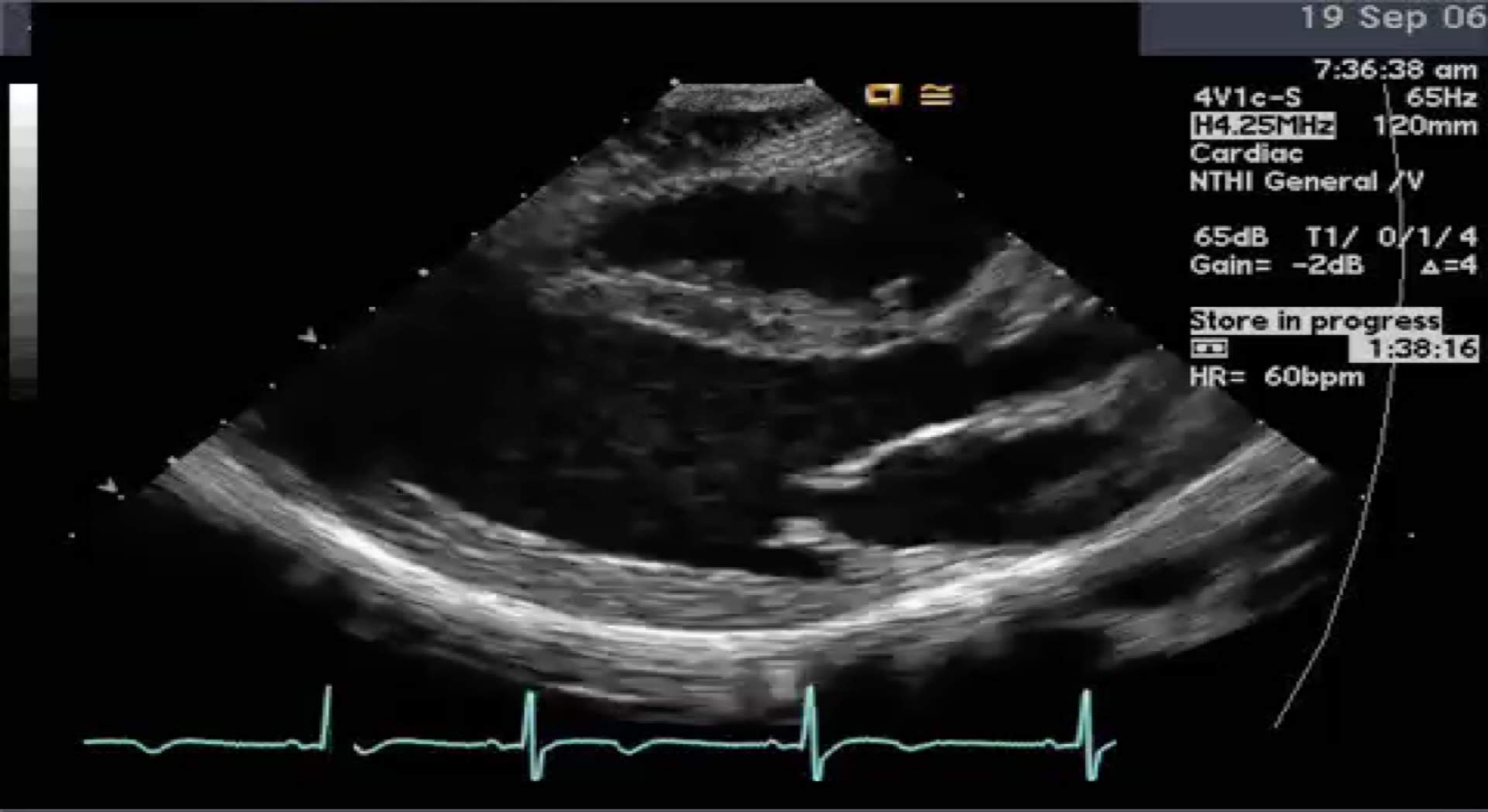
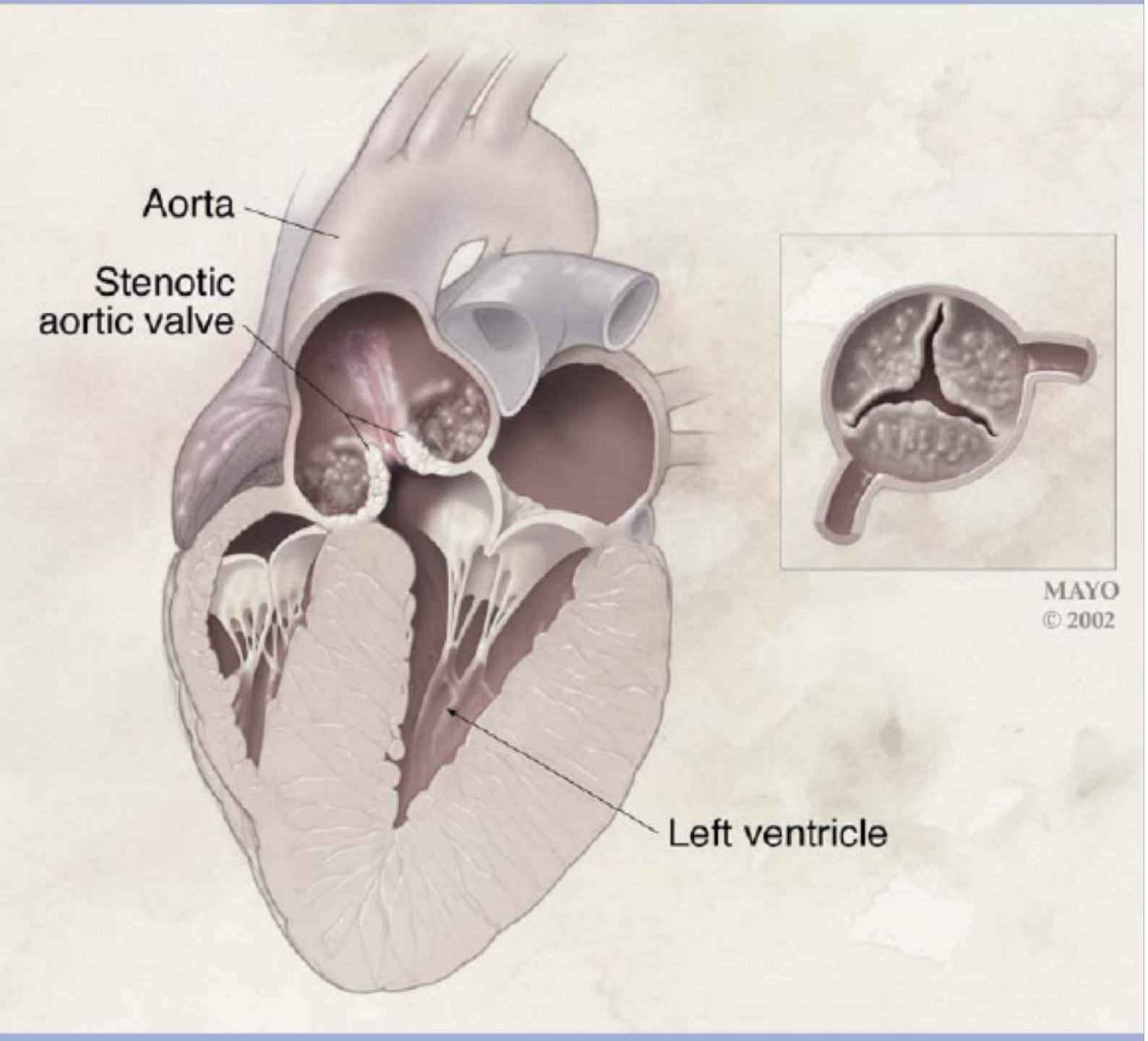
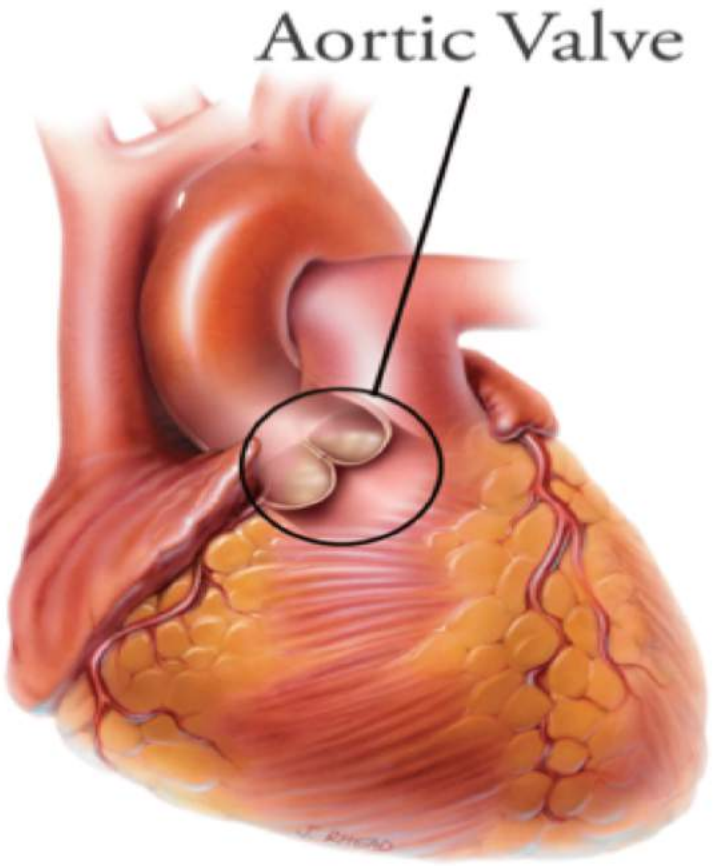

19 Sep 06
7:36:38 am
4V1c-S
65Hz
H4.25MHz
120mm
Cardiac
NTHI General /V
65dB T1/ 0/1/ 4
Gain= -2dB
Δ=4
Store in progress
1:38:16
HR= 60bpm

Overview of the main valvular pathologies

- Aortic stenosis
- Aortic insufficiency (Acute & chronic)
- Mitral stenosis
- Mitral regurgitation (Acute & chronic)
- Tricuspid and pulmonic valves
- Prosthetic valve dysfunction



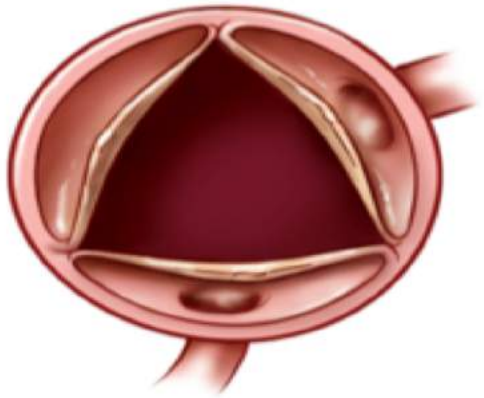
Aortic stenosis (AS)



Narrowing of the aortic valve

HEALTHY AORTIC VALVE

Open



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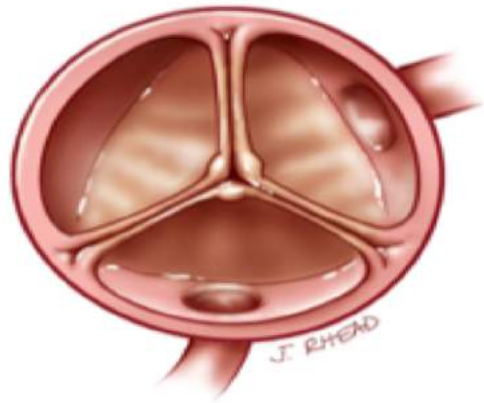
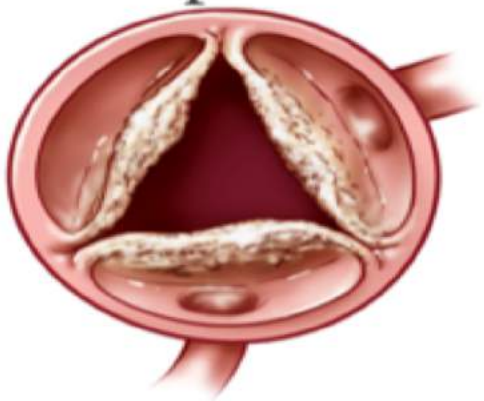


AORTIC VALVE STENOSIS

Open



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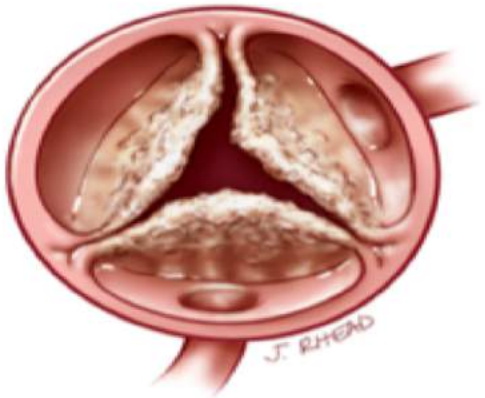


Etiology

- **Degenerative calcific AS (Most common in elderly)**
  - ✓ A degenerative condition caused by inflammation and progressive calcificationwhich limits movement of the aortic valve.
  - ✓ Most common cause (80%).
  - ✓ Affects patients later in life (>65).
  - ✓ Risk factors: hypercholesterolaemia, hypertension, smoking and diabetes.

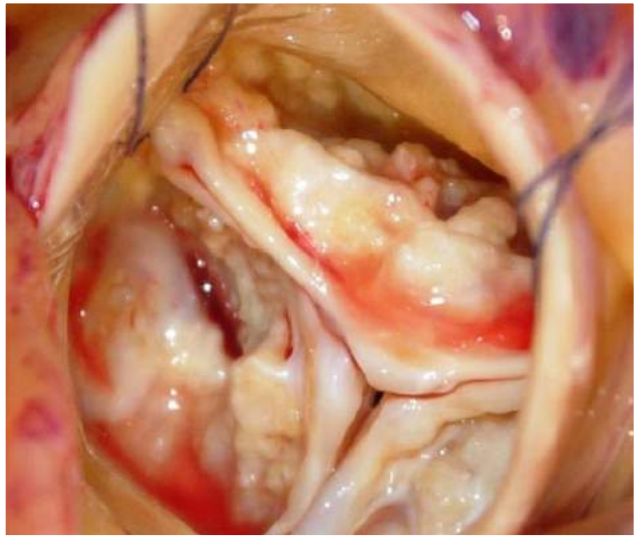
- **Congenital: Bicuspid aortic valve (Most common in young)**
  - ✓ The most common congenital abnormality of the heart occurring with a frequency of 1-2%, with males more commonly affected.
  - ✓ Valves predisposed to becoming stenotic.
  - ✓ Stenosis tends to appear at a younger age (<65).

- **Rheumatic**
- **CTD e.g Rheumatoid**

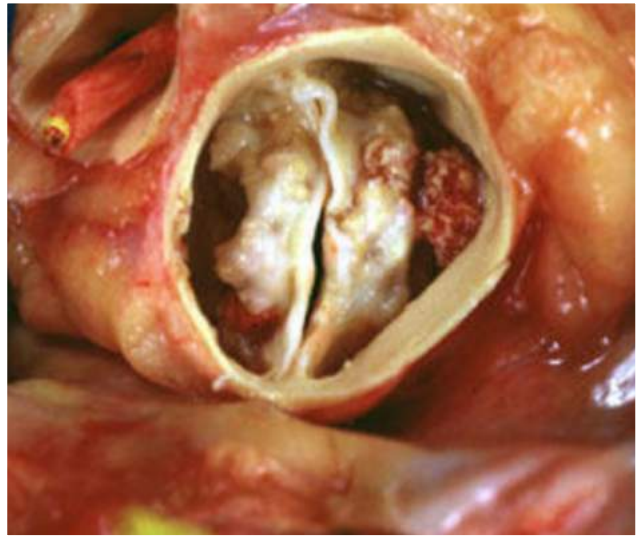
Differential Diagnosis

- Supravalvular stenosis
- Subvalvular stenosis
- Hypertrophic cardiomyopathy (HCM)

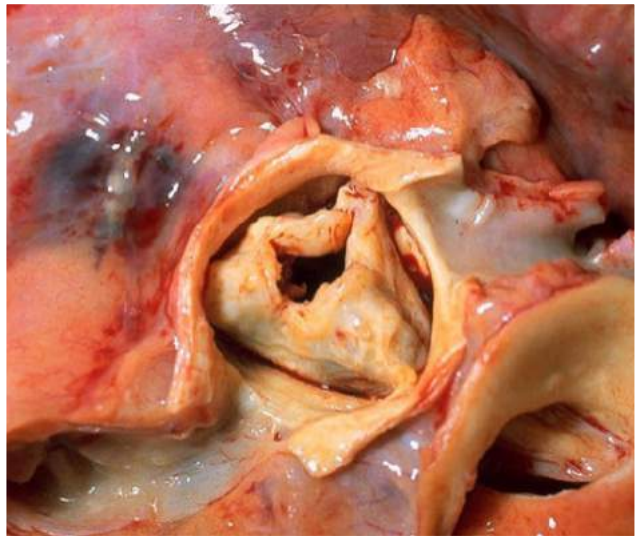
Degenerative



Bicuspid



Rheumatic



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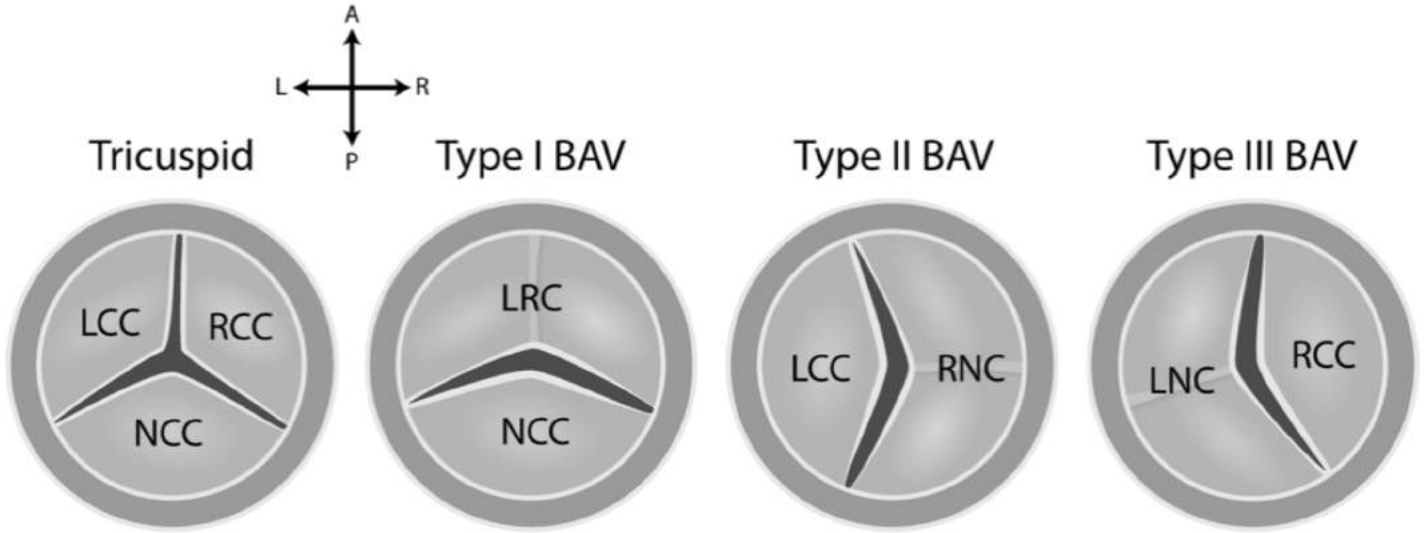
Bicuspid Aortic Valve

- 1-2% of the population
- 70-80% fusion of the right & left coronary and non-coronary leaflets
- 20-30% fusion of the right & non-coronary leaflets
- Fusion of the non coronary & left coronary leaflets is rare
- One commissure



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Bicuspid Aortic Valve

- Associated aortopathy (Medial degeneration) : aneurysm, dissection.
- Requires annual imaging if aorta > 4.5 cm
- Beta blockers in absence of significant AI

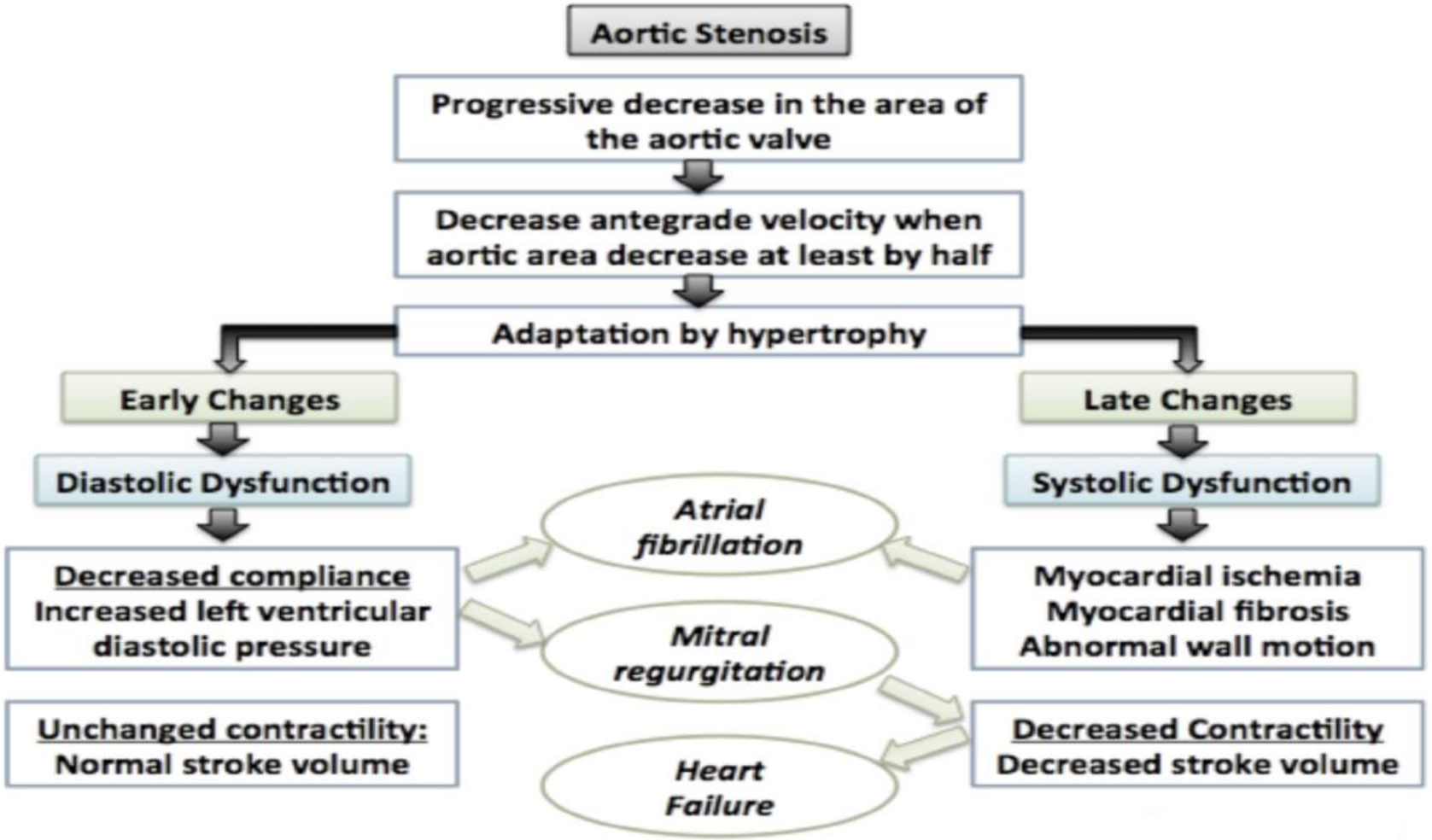
Asc A replacement if:

✓ Aorta > 5.5 cm

✓ Aorta > 4.5 cm if AVR indicated

✓ Aorta >5 cm with risk factors for dissection e.g FHX or progression of >0.5 cm/y

Pathophysiology



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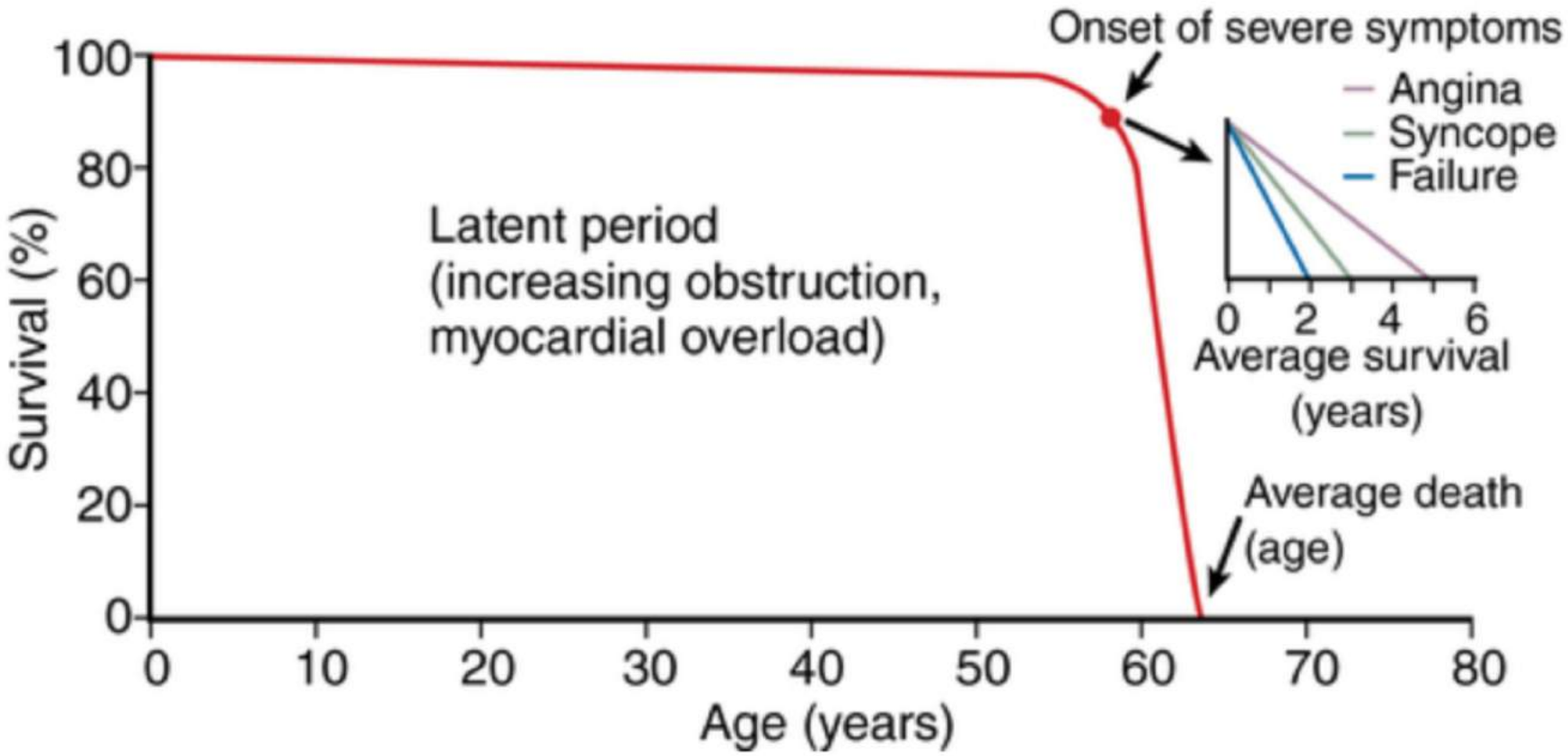
Natural history



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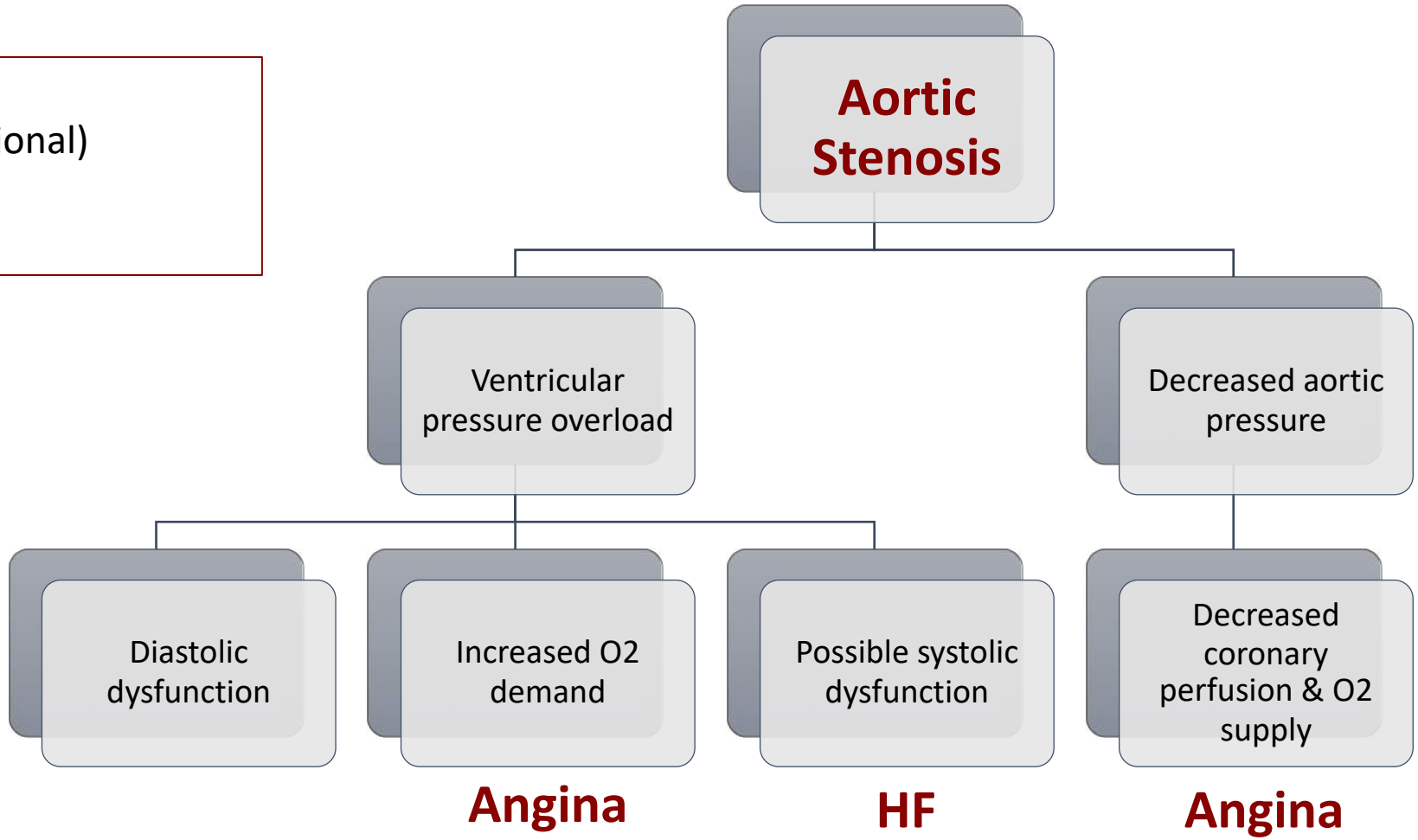
Clinical Presentation (Symptoms)

Classical triad:
1. Syncope (exertional)
2. Angina
3. Dyspnoea



Syncope

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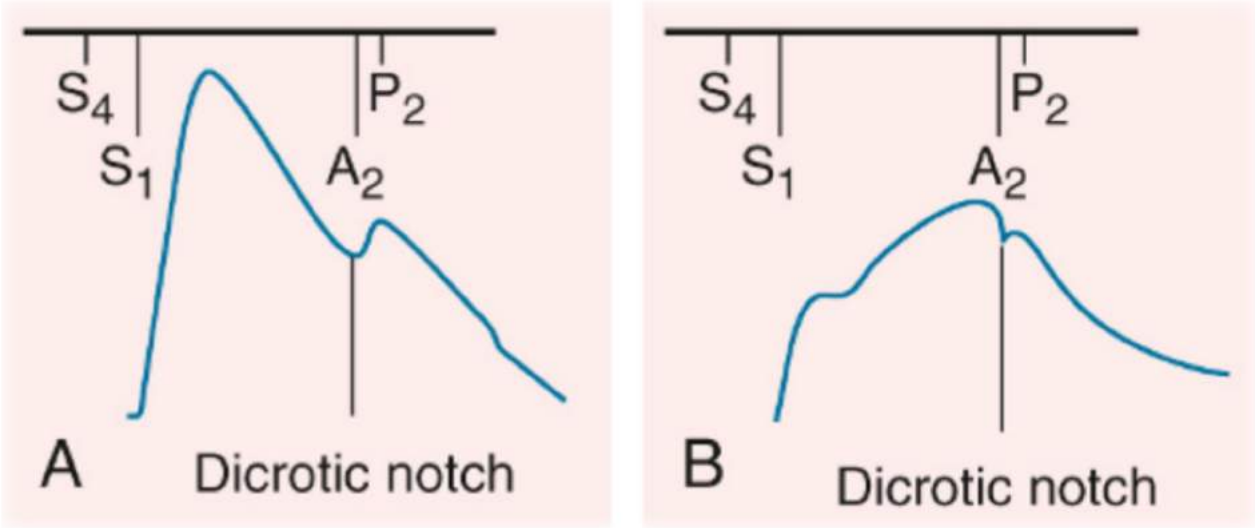
Physical exam & clinical signs:

- **Pulse:**

✓ Low volume, slow-rising, delayed upstroke carotid pulse ***(pulsus parvus et tardus)***

✓ Brachioradial delay



Physical exam & clinical signs:

- **BP:** Narrow pulse pressure
- **Apex:** Sustained beat
- **HS:** Soft S2, reversed splitting, single S2.
- **Extra-HS:** S4

Physical exam & clinical signs:

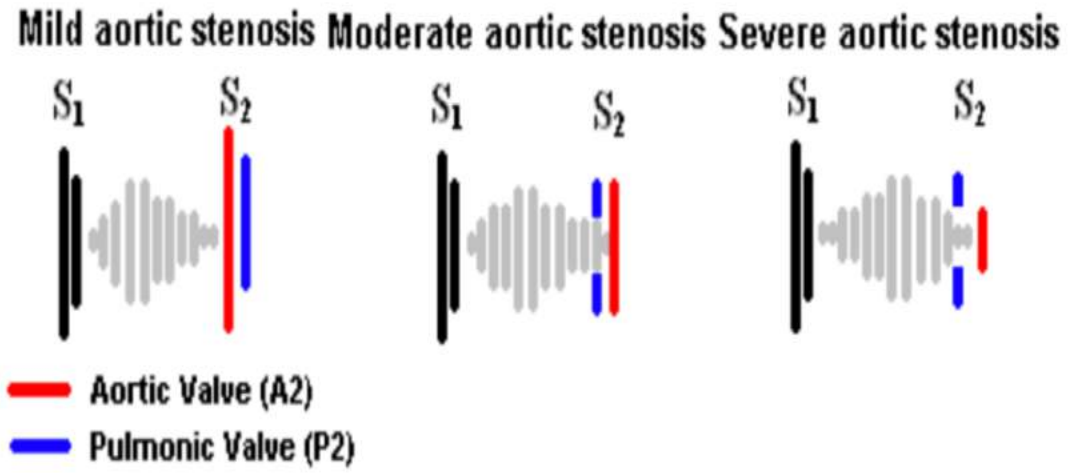
- **Murmurs:**

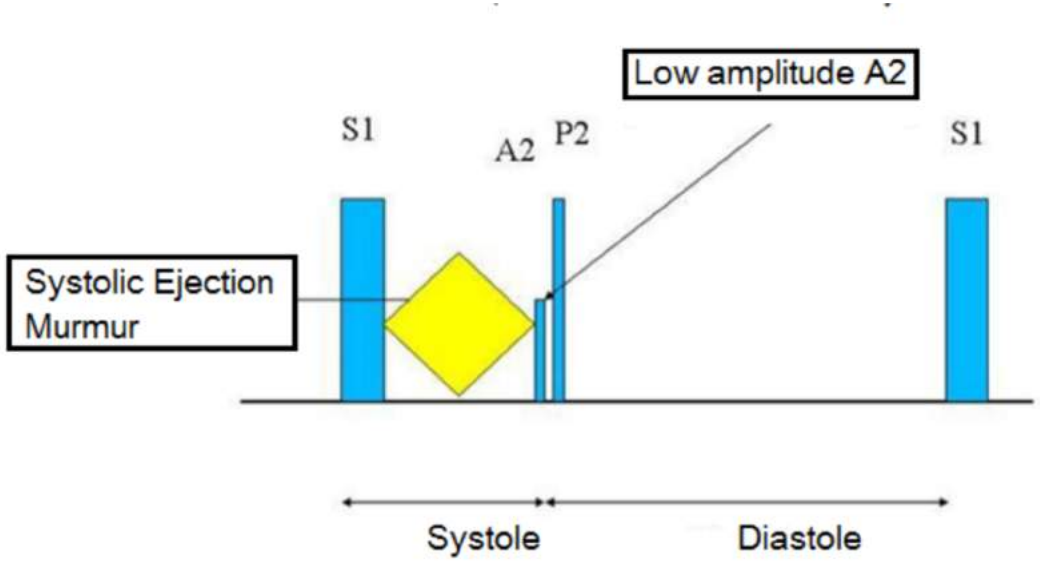
✓ Loud mid- to late-peaking ejection systolic murmur in the right intercostal space radiating to carotids

✓ Gallavardin phenomenon:

High-pitched musical components of the murmur of aortic stenosis heard at the apex

HS & Murmur of AS





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Investigations & Diagnosis

- Bedside assessment & observation of syndromic features
- Blood pressure
- ECG
  - ✓ Left ventricular hypertrophy (deep S-waves in V1 and V2, tall R-waves in V5 and V6) & strain pattern
  - ✓ Normal axis or LAD
  - ✓ LAE

Investigations & Diagnosis

Blood tests:

- CBC
- U&Es
- Lipid profile
- Coagulation profile

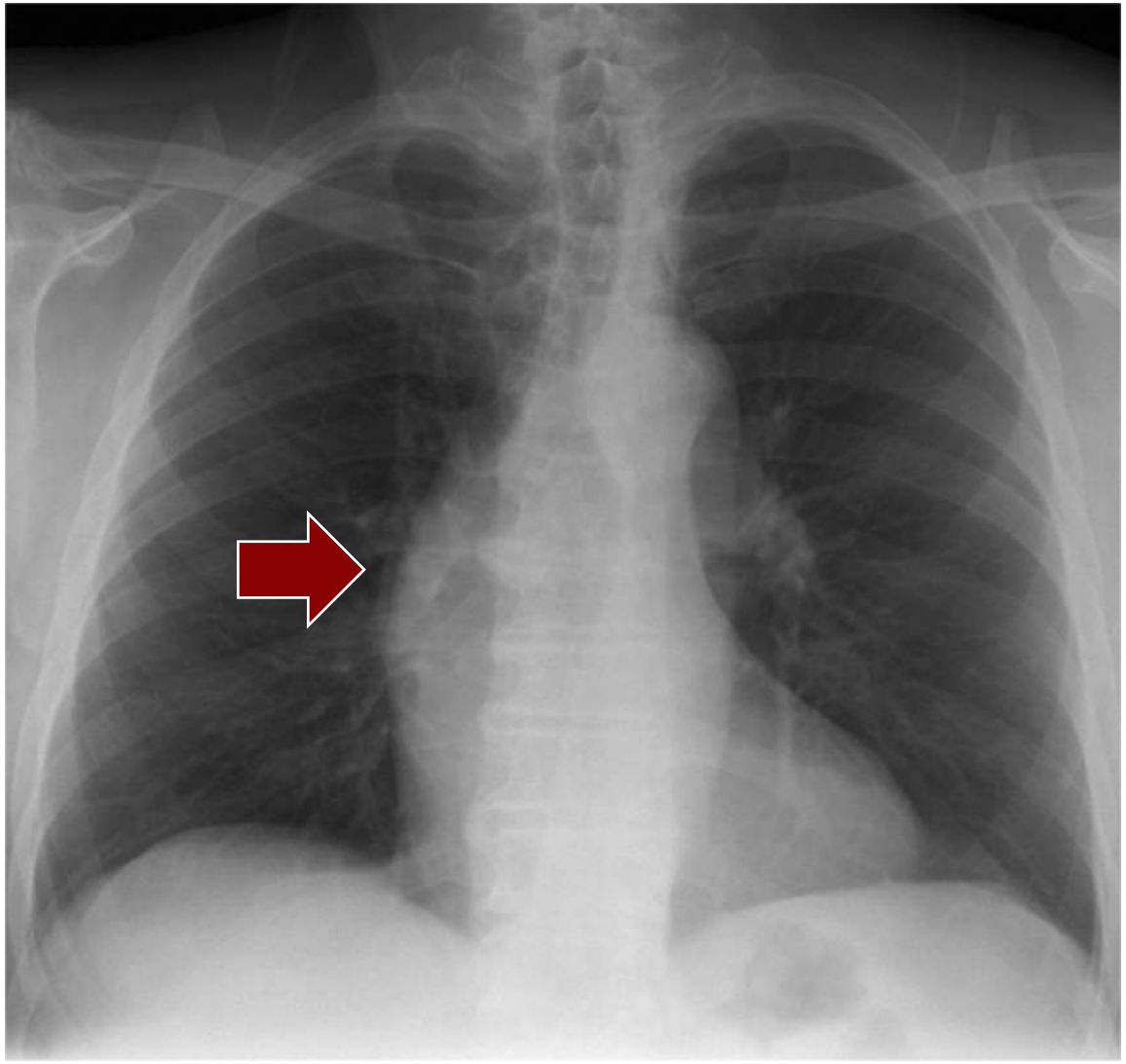
Imaging:

- Echocardiogram
  - Allows assessment the valve area, ejection fraction and ventricular hypertrophy, see below.
- CXR
  - Typically demonstrates a small heart; cardiomegaly occurs if heart failure develops.
  - Dilated ascending aorta.

Special:

- Cardiac MRI
- Cardiac catheterisation
- ECG exercise stress testing
  - May be used in asymptomatic patients. A positive test may be indicated by the onset of symptoms, ECG changes, or an abnormal BP response.

CXR



Echo

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Management

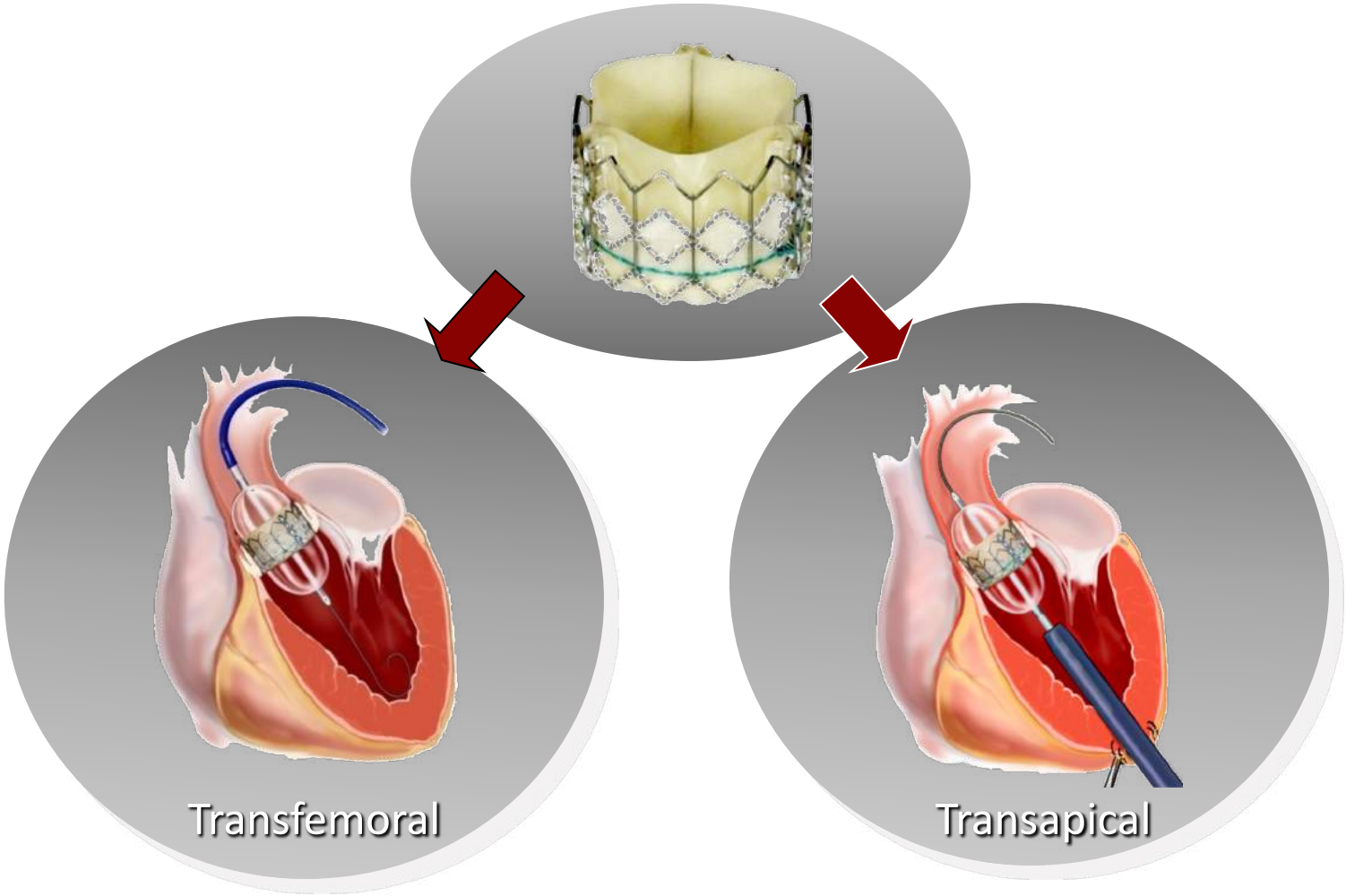
Depends on:

- Severity of the stenosis
- Presence of symptoms

Severe symptomatic cases need surgical (SAVR) or percutaneous (TAVR) intervention

HTN is common (Cautious use of vasodilators due to afterload reduction)

TAVR
Transfemoral (TF) and Transapical (TA)



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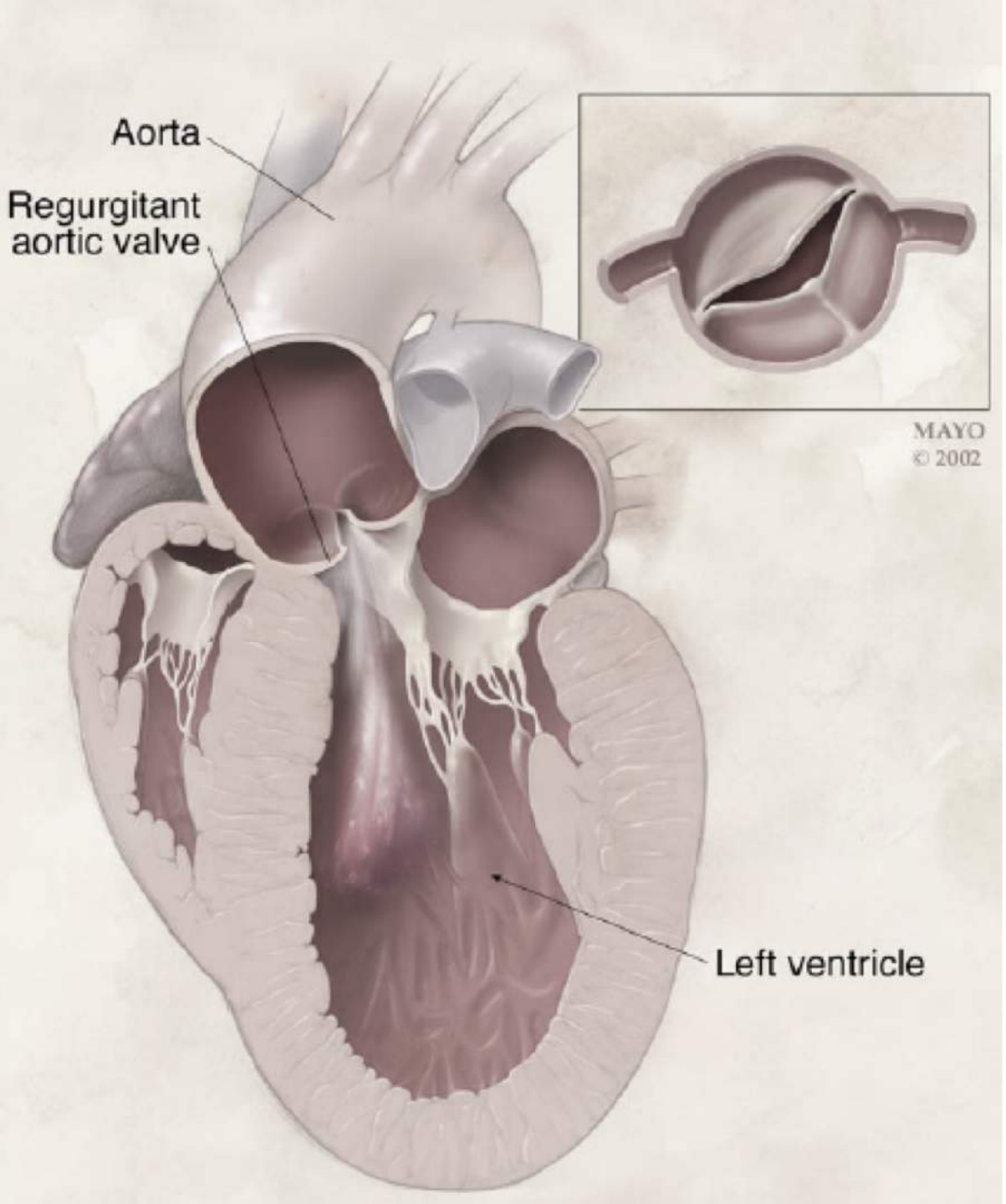

Aortic Regurgitation/Insufficiency AR/AI

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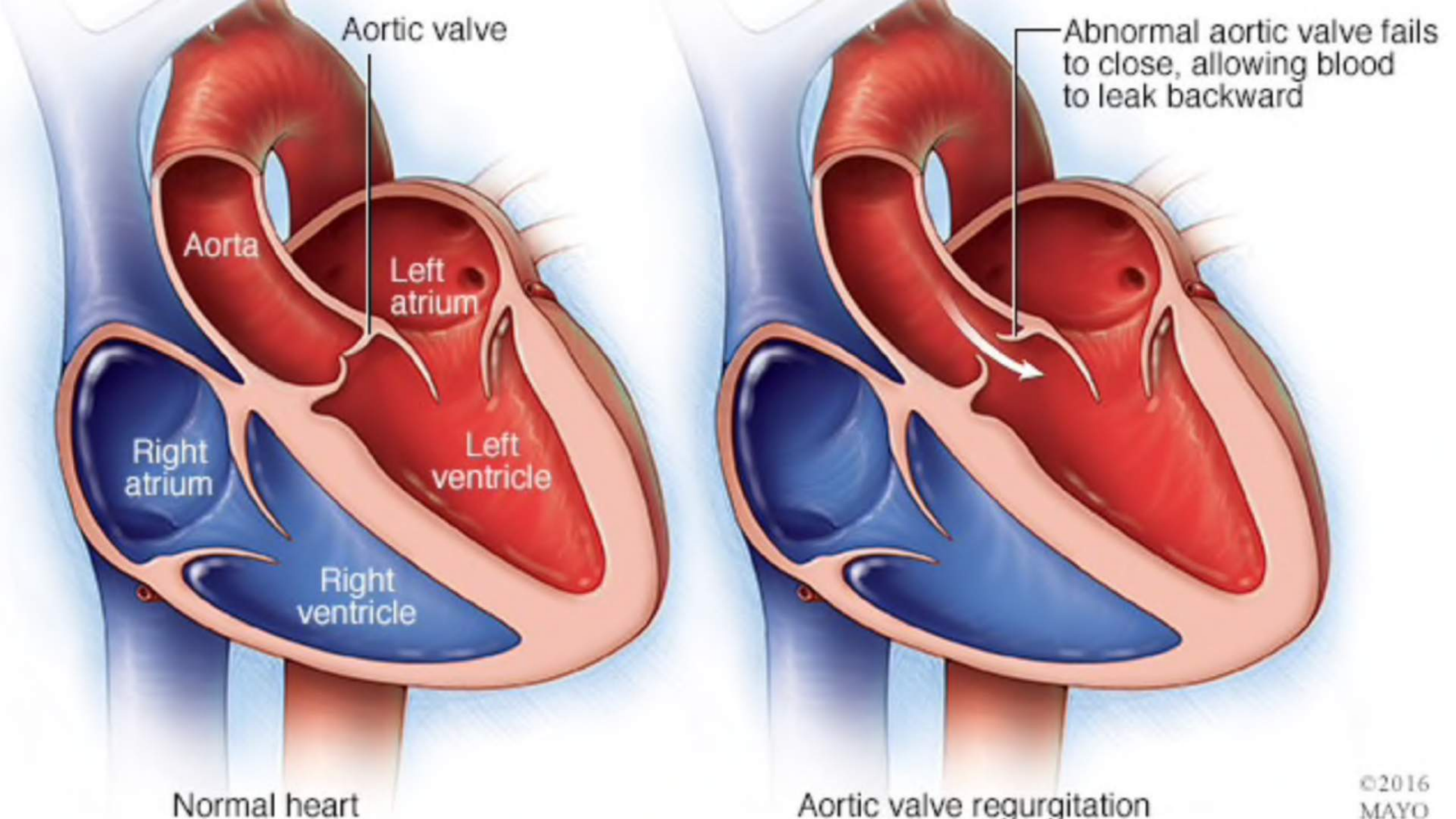
Aortic valve
Aorta
Left atrium
Right atrium
Left ventricle
Right ventricle
Abnormal aortic valve fails to close, allowing blood to leak backward
Normal heart
Aortic valve regurgitation
©2016 MAYO

Aortic Regurgitation/Insufficiency (AR/AI)

- Aortic regurgitation results from an incompetent aortic valve causing a regurgitant flow of blood in diastole.
- Usually presents 4^{th}-6^{th} decades of life.
- Affects males 3 times more commonly than women.
- Severe disease is seen in < 1% of the population.
- The most common causes are degenerative disease & congenital bicuspid valve.

Etiology

Valvular (aortic valve cusps)	Non-valvular (aortic root)
Infectious: IE, RF	Marfan
Congenital: BAV, Marfan	syphilis
Inflammatory: SLE, RA, Behcet	Ankylosing spondylitis
Degenerative: Myxomatous AV, calcific AV	Idiopathic aortitis
Trauma	Ehler danlos syndrome
Postaortic valvuloplasty	Aortic dissection
Drug induced: Fenfluramine	Trauma
	VSD
Acute	Chronic
AD	Calcific degeneration
IE	Aortic root dilatation
Rupture of AV leaflet (trauma)	RF
PV dysfunction	Autoimmune diseases/CTD

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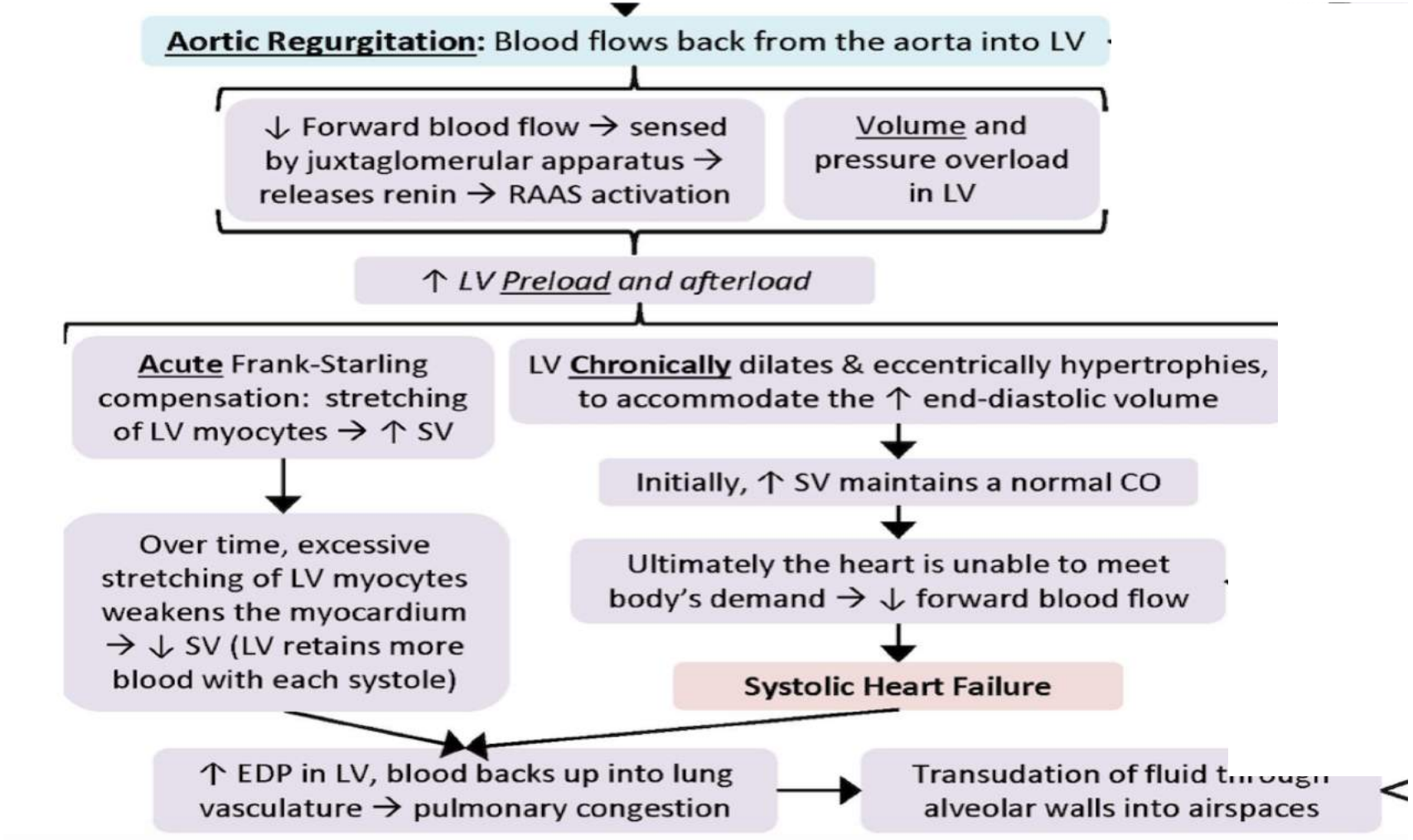
Pathophysiology



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Acute AI

- A medical emergency
- The compensatory changes seen in chronic disease do not have time to develop
- *Presents with pulmonary oedema & cardiogenic shock.*

- **2 Main consequences:**

1. Reduced coronary flow - the coronaries fill predominantly during diastole, regurgitant flow at this time reduces filling. Results in angina or in severe cases myocardial ischaemia.
2. Increased EDP- causes increased pulmonary pressures with resulting pulmonary oedema and dyspnoea. In severe cases, cardiogenic shock may occur.

Chronic AI

- Patients may remain asymptomatic for many decades.

- **Develops slowly with compensatory changes:**
  - ✓ Increase in the left ventricular end-diastolic volume (essentially the preload).
  - ✓ Increased stroke volume compensating for regurgitant flow supported by the ventricular hypertrophy to maintain ejection fraction, with a greater preload leading to greater contractility *(frank- starling law)*
  - ✓ Eventually further increases in preload cannot be met by greater contractility and heart failure develops.

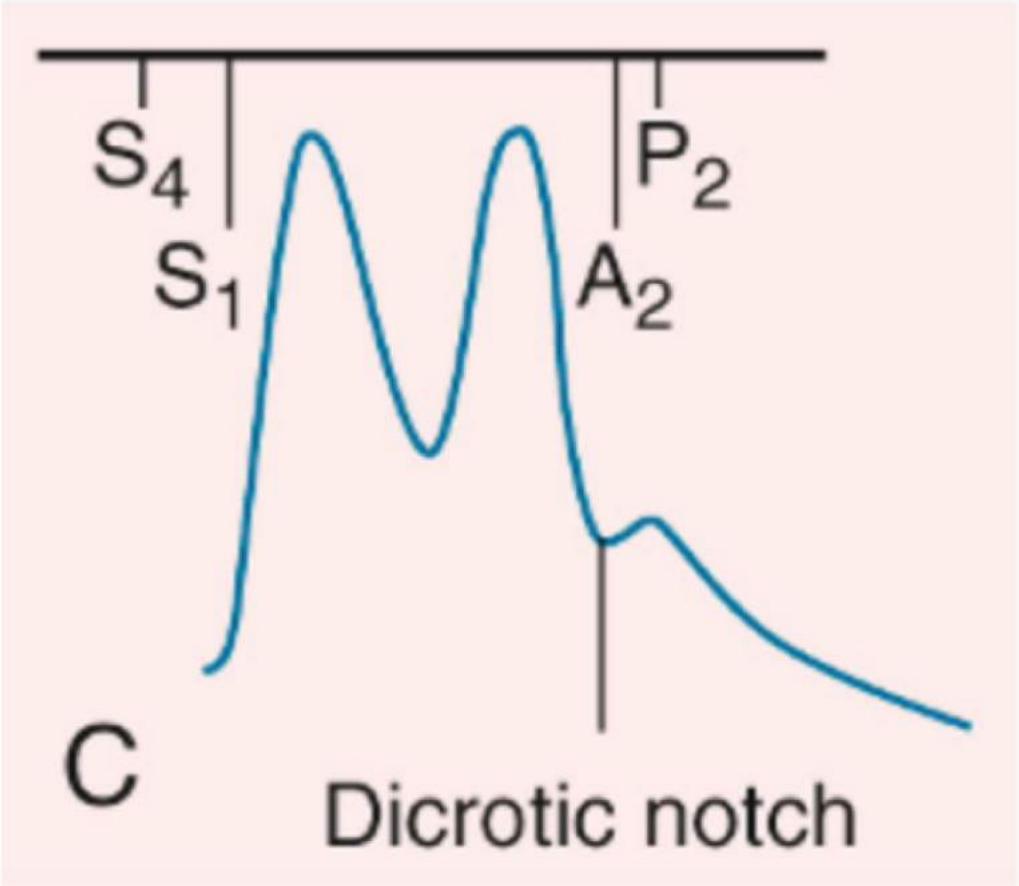
Clinical Presentation (Acute AI)

Symptoms	Signs
Dyspnea	Cardiogenic shock & Heart failure: Hypotension, tachycardia, elevated JVP…etc
Chest pain	Peripheral signs of chronic AI are usually absent
Symptoms of low cardiac output & HF	S3+ Murmur is early, short, faint and may be absent

Clinical Presentation (Chronic AI)

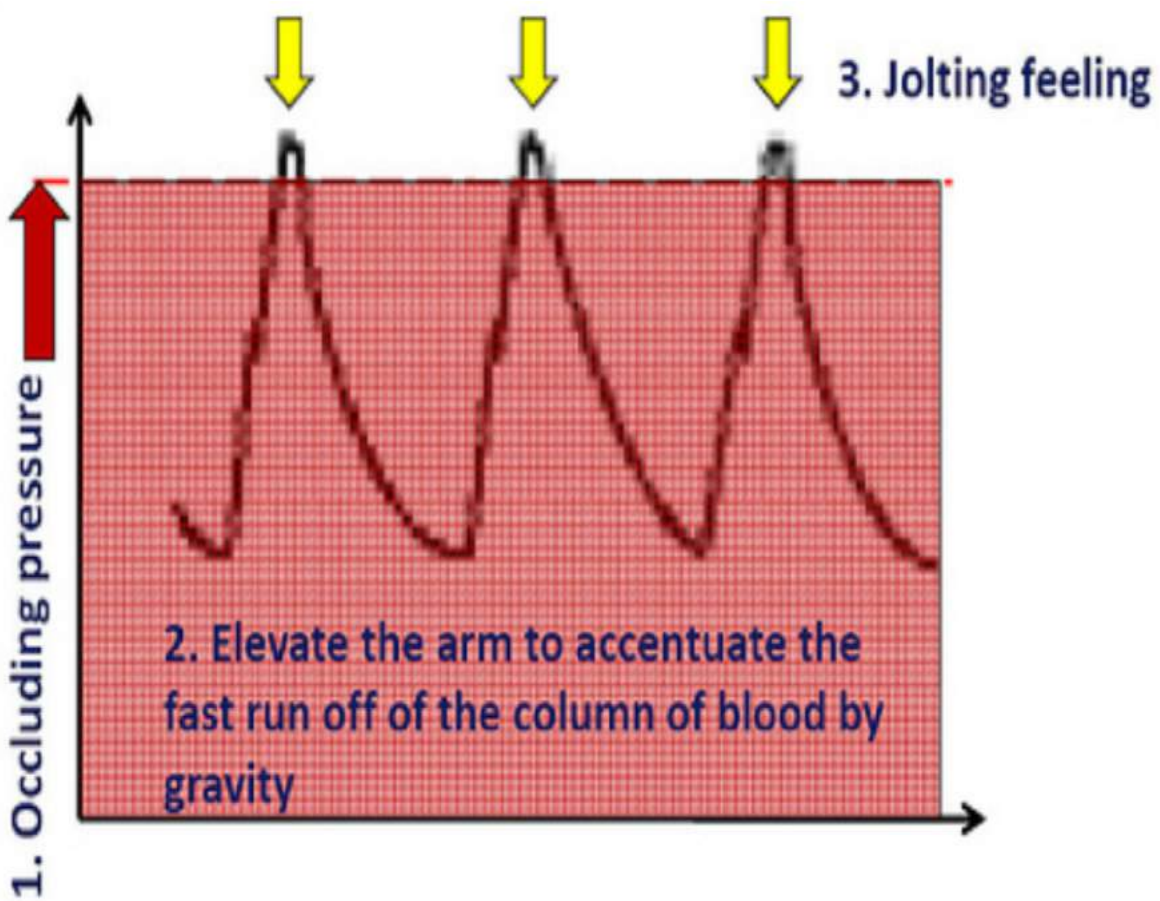
Symptoms	Signs
Dyspnea	**Pulse:** Pulsus Bisferience/ collapsing/ water hammer
Chest pain	**BP:** Elevated with wide pulse pressure
	Apex: Diffuse & Displaced
	HS: Soft S1 & S2
	Murmurs: Decrescendo early Diastolic & Austin flint murmur

Pulsus Bisferiens



Water- hammer /collapsing pulse

(Large volume sudden collapsing pulse)



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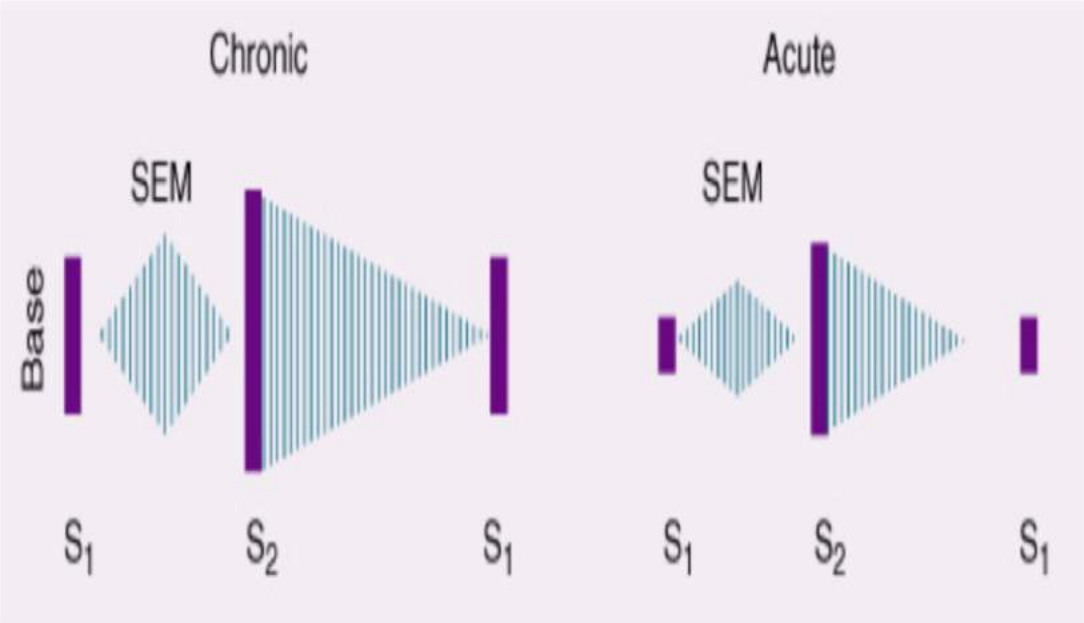
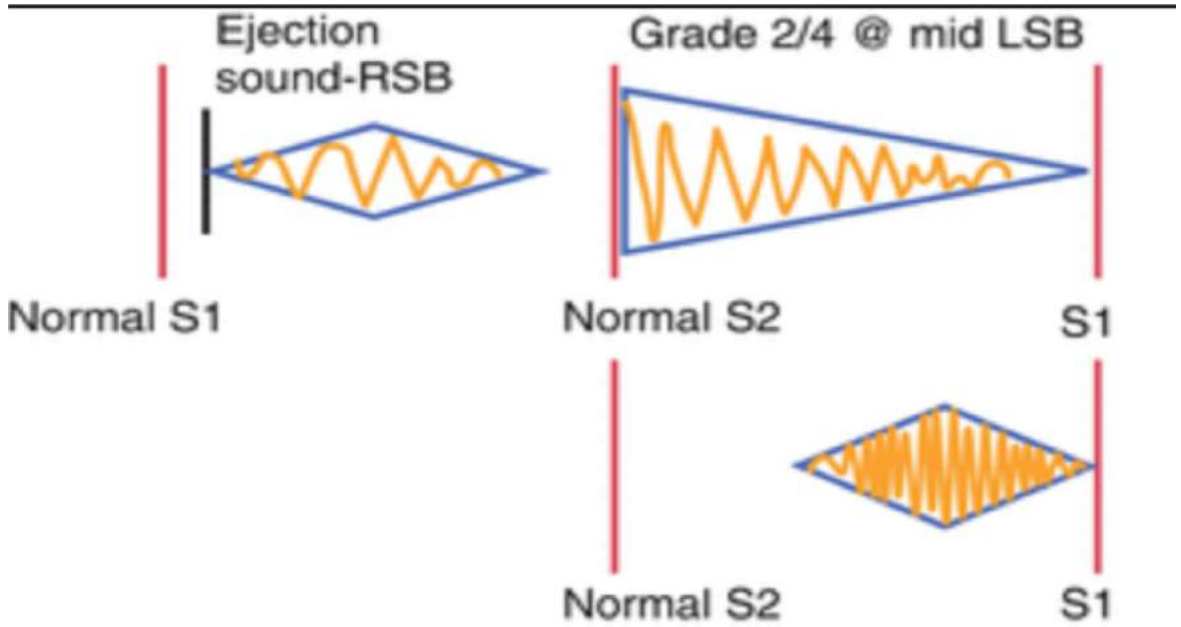
Decrescendo Diastolic murmur



Austin- flint murmur



Peripheral signs of AR

The relevance of these signs in clinical practice today is questionable:

- **De Musset's** - systolic nodding of the head.
- **Quincke's** - capillary pulsation of nail beds.
- **Corrigan's sign** – abrupt distension with prominent pulse then rapid collapse.
- **Traube's (pistol shot femoral)** – systolic & diastolic bruit in the femoral artery
- **Duroziez's** - systolic bruit in the FA with proximal compression and diastolic sound with distal compression using the stethoscope.
- **Müller's** - systolic pulsation of uvula.
- **Hill's sign** – SBP in legs > 20 mmHg higher than SBA in arms.

Investigations & diagnosis

- Bedside assessment & observation of syndromic features
- Blood pressure
- ECG
  - Left ventricular hypertrophy (deep S-waves in V1 and V2, tall R-waves in V5 and V6).
  - Left ventricular strain may be seen in severe disease.
- Bloods
- FBC
- U&Es
- Cholesterol
- Clotting

Investigations & diagnosis

- **Imaging**

A. Echocardiogram

  - Allows visualisation of the origin of regurgitant jet and its width, detection of aortic valve pathology and ventricular hypertrophy.

B. CXR

  - May demonstrate cardiomegaly.
  - Dilated ascending aorta.
  - Calcification may be seen.

- **Special**

  - ✓ Cardiac MRI
  - ✓ Cardiac catheterisation
  - ✓ ECG exercise stress testing

Management

- **Acute AR:**

Emergency AVR

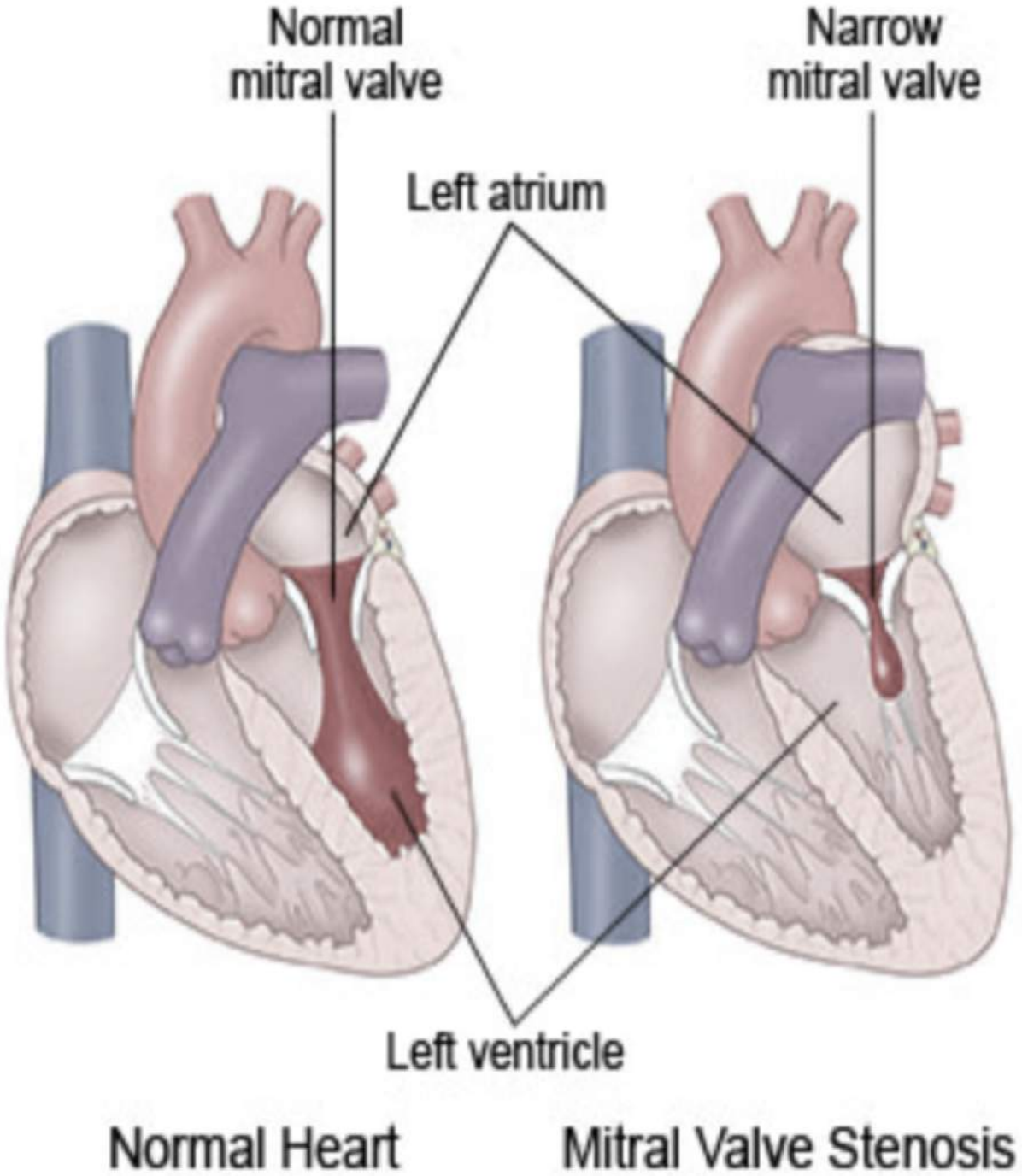
- **Chronic AR:**

A. Medical treatment:

- Treat HTN
- ACE inhibitors/ARBs are reasonable in patients with severe symptomatic AR and/or LV dysfunction when surgery is not performed because of comorbidities.

B. Surgical treatment:

- AVR



Mitral Stenosis (MS)

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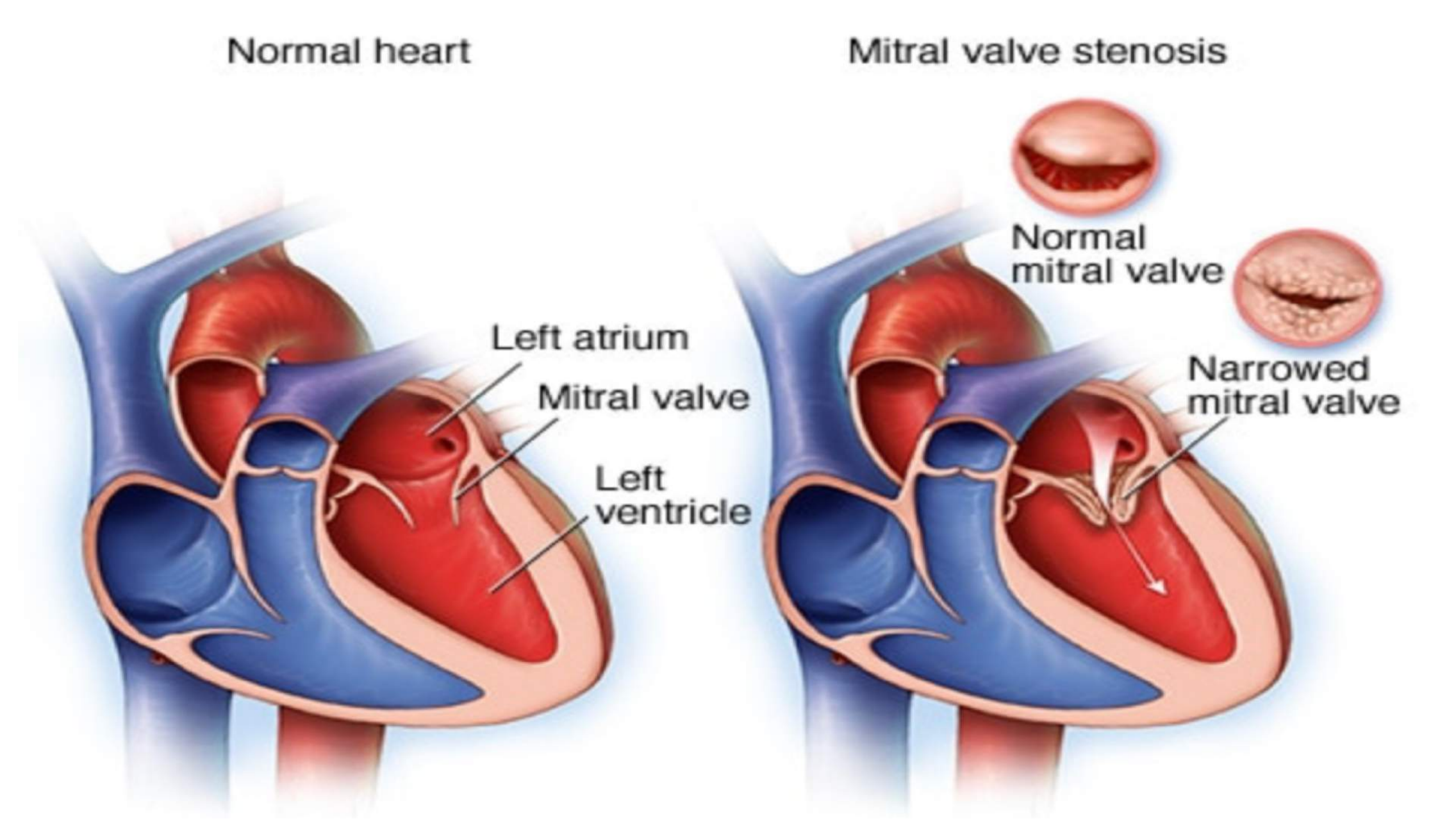
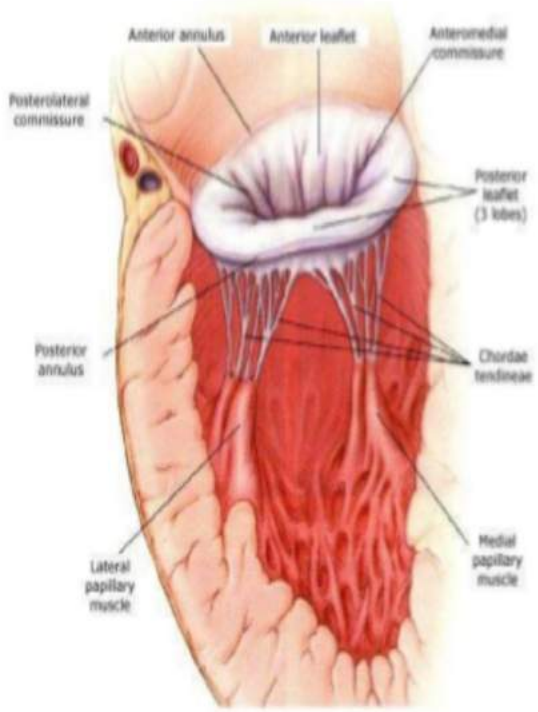

Normal heart
Left atrium
Mitral valve
Left ventricle
Mitral valve stenosis
Normal mitral valve
Narrowed mitral valve

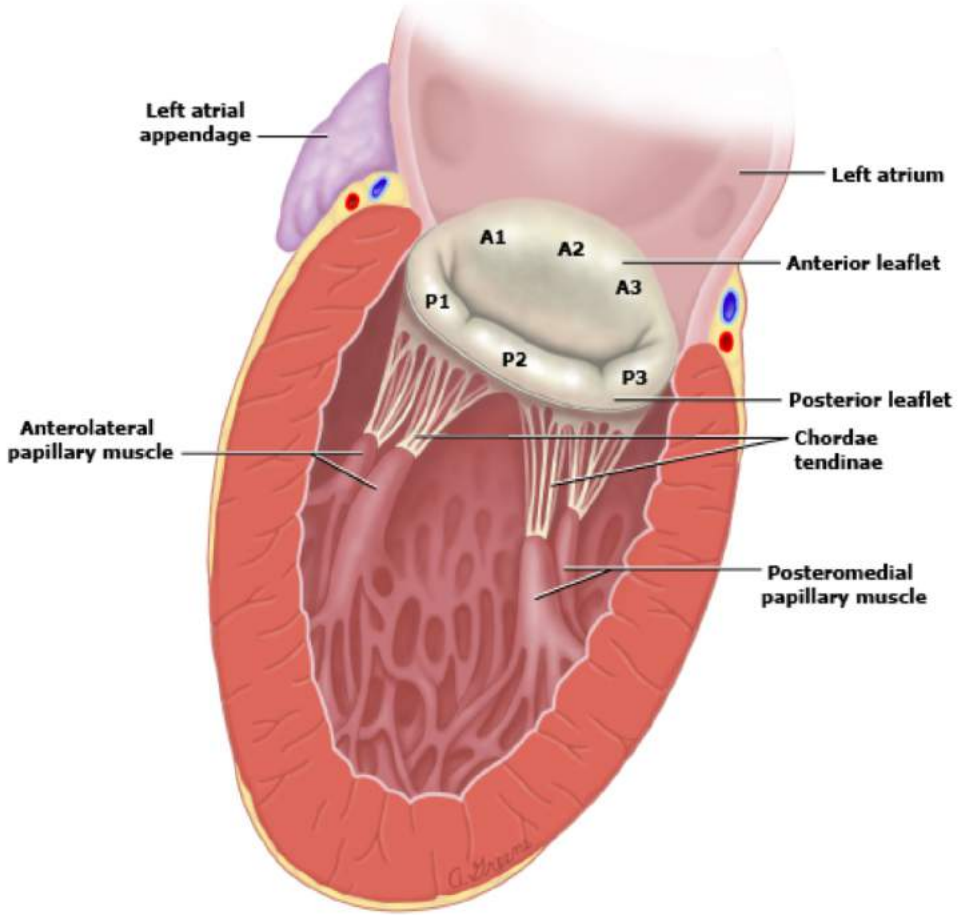
Mitral Valve Structure



- The mitral valve connects the left atrium (LA) and the left ventricle (LV).
- The mitral valve opens during diastole to allow the blood flow from the LA to the LV.
- During ventricular systole, the mitral valve closes and prevents backflow to the LA.

The mitral apparatus is composed of

1. the left atrial wall
2. the annulus
3. the leaflets
4. the chordae tendineae
5. the papillary muscles
6. the left ventricular wall



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Etiology

Rheumatic (leading cause of MS)
25% isolated
40% associated MR
35% AV disease
6% TV disease

Congenital
(Shone syndrome)

CTD RA/SLE

Drug induced e.g methysergide

Radiation induced

Rare conditions include Fabry's disease, Whipple's disease, mucopolysaccharidosis, Carcinoid with lung mets or PFO

Calcific (age related)

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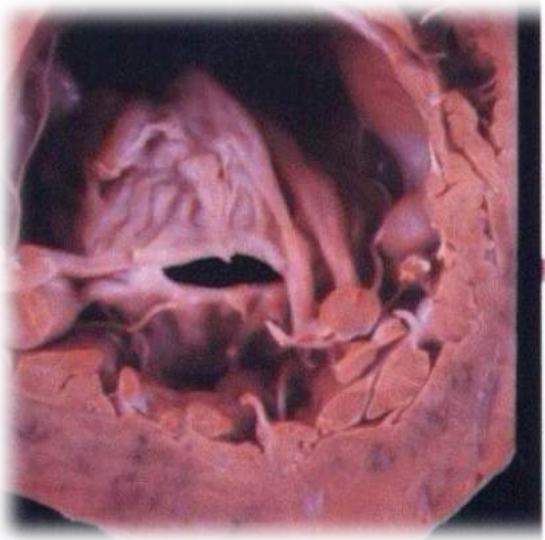
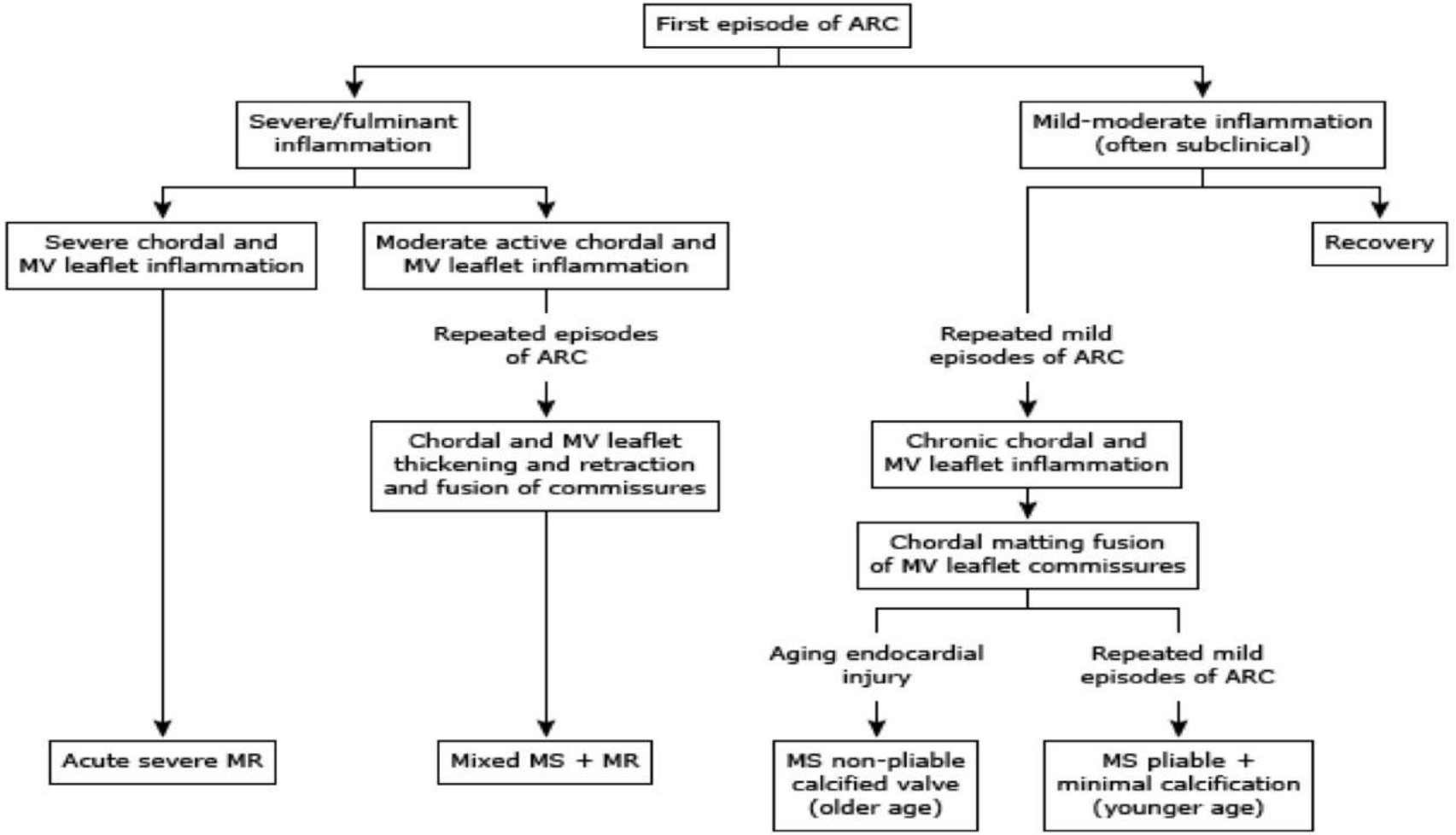


Pathophysiology



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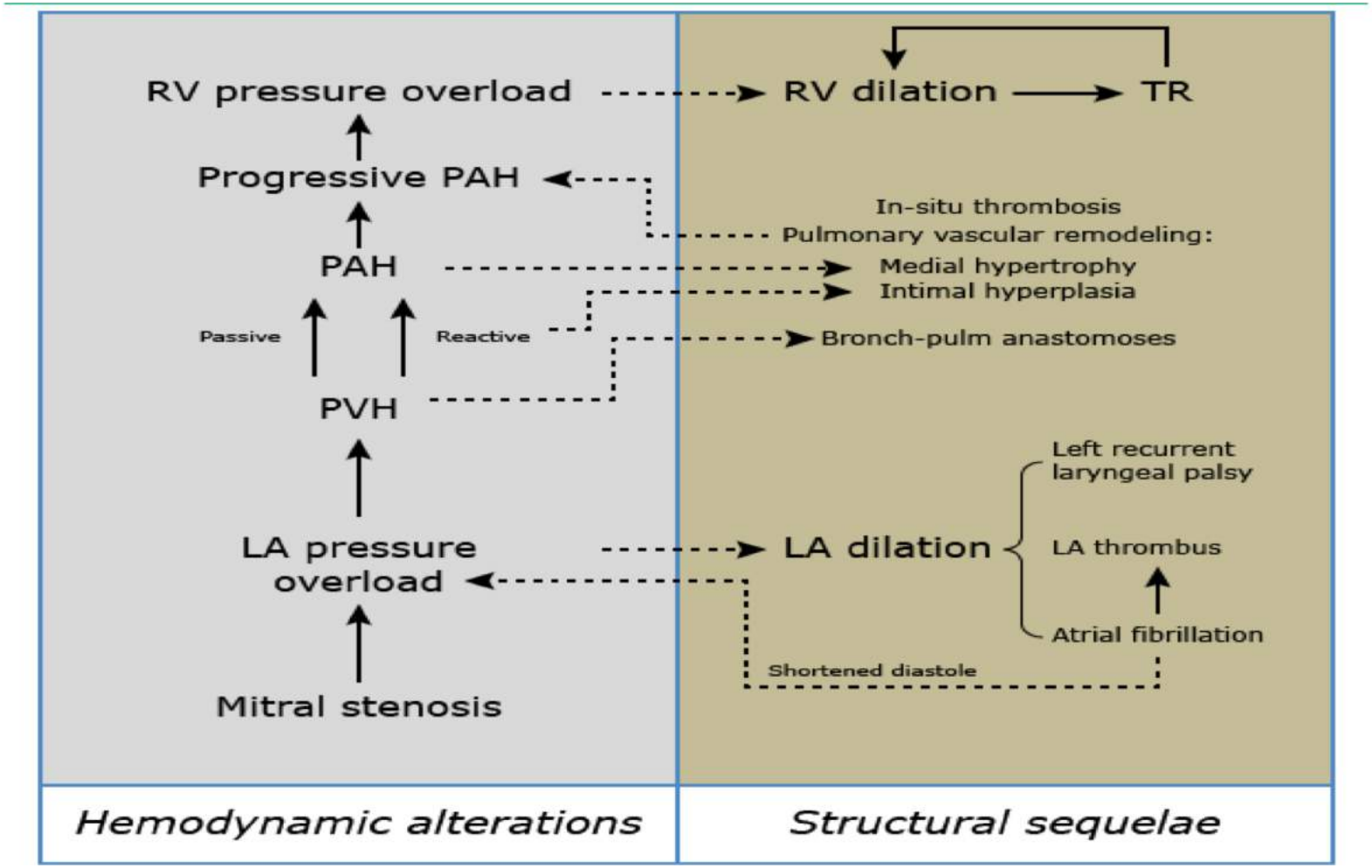
Hemodynamic consequences



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Natural history

- Progressive lifelong disease
- Slow and stable initially then progressive acceleration years later
- Long latent period post RF (years)

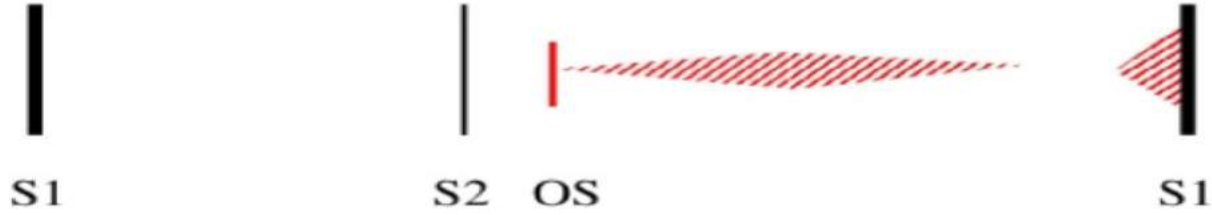
Clinical presentation

Symptoms

- Dyspnea
- Poor exercise tolerance
- Orthopnea
- PND
- PHT symptoms: RHF, hemoptysis
- Palpitation
- Peripheral embolism secondary to AF
- Ortner’s syndrome recurrent laryngeal nerve compression by dilated LA

Physical exam

- **Face:** Mitral faces pink purple plaques on cheeks (due to systemic VC)
- **Pulse:** regular (sinus), irregular with AF, low volume
- **Apex:** Tapping beat (palpable S1)
- **JVP:** Prominent a wave in sinus rhythm
- **HS:** Loud S1 when leaflets are still pliable
- **Extra HS:** Opening snap (OS) early in diastole, can mimic a split second heart sound. caused by thickened valve leaflets as they open (Earlier OS indicates severe MS)
- **Murmur:** **L**ow pitched mid diastolic rumble at the apex with presystolic accentuation
- No S3
- Signs of PHTN



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Mitral faces

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Investigations & Diagnosis

- Bedside assessment & observation of syndromic features.
- Bloods
- FBC
- U&Es
- Cholesterol
- Clotting
- **ECG:**

AF, LAE, RVH

Investigations & Diagnosis

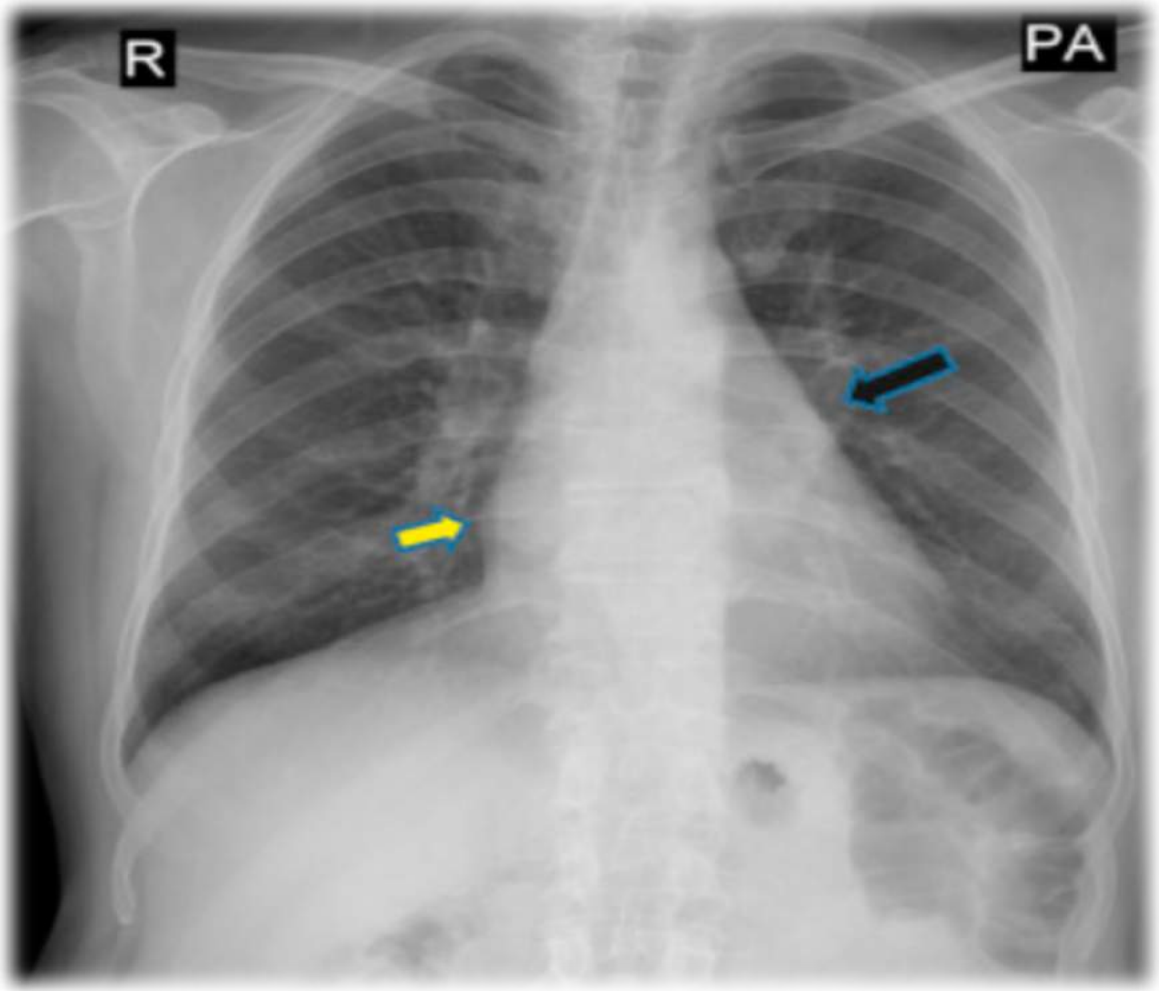
Imaging

1. Echocardiogram
    - Gold standard
2. CXR:
    - LAE: straitening of the left heart border
    - Pulmonary congestion and prominent PA
    - Calcified MV

Special

- Cardiac catheterisation
- ECG exercise stress testing

CXR with MS



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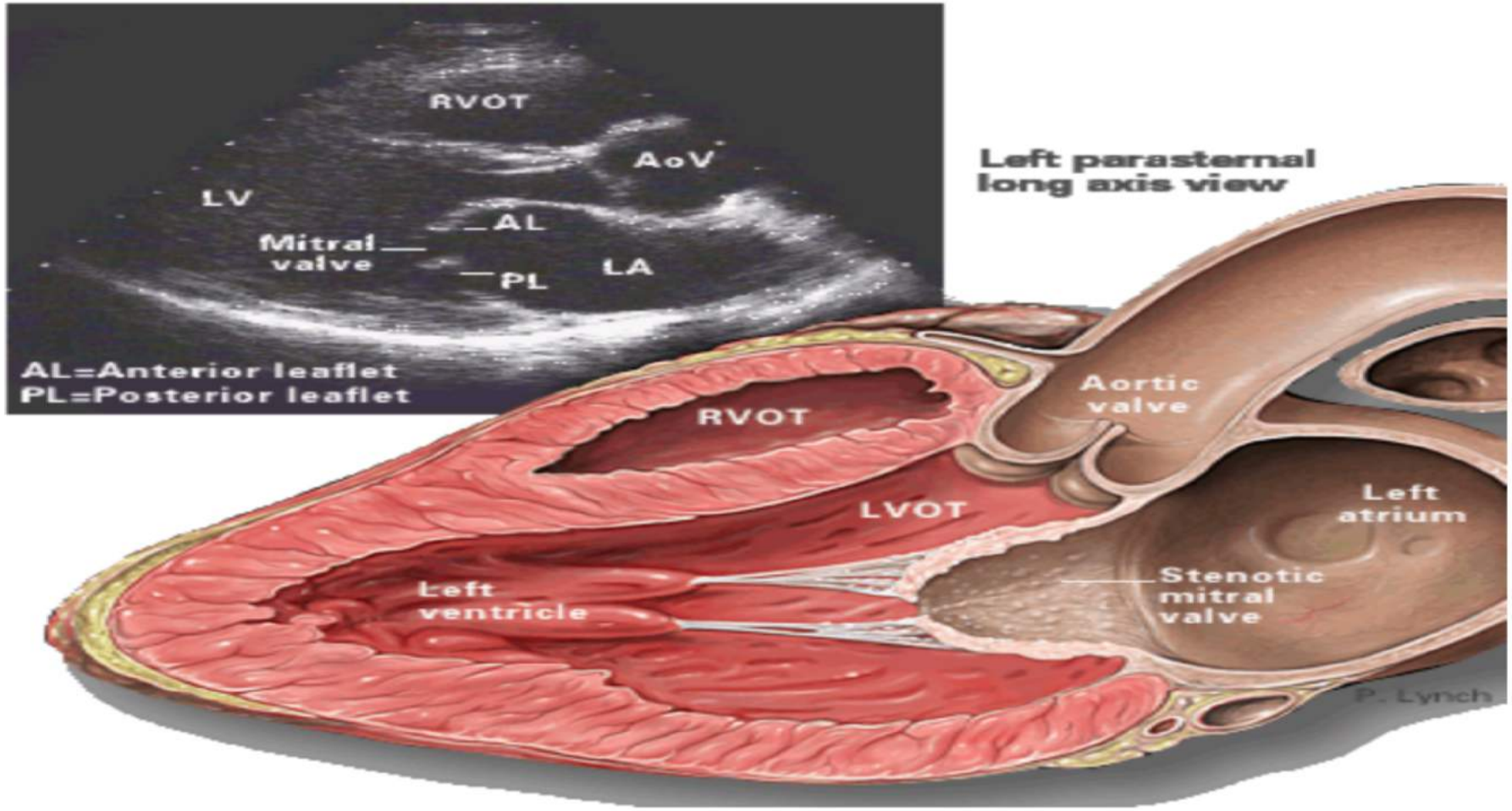
Echo (MS)



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Management

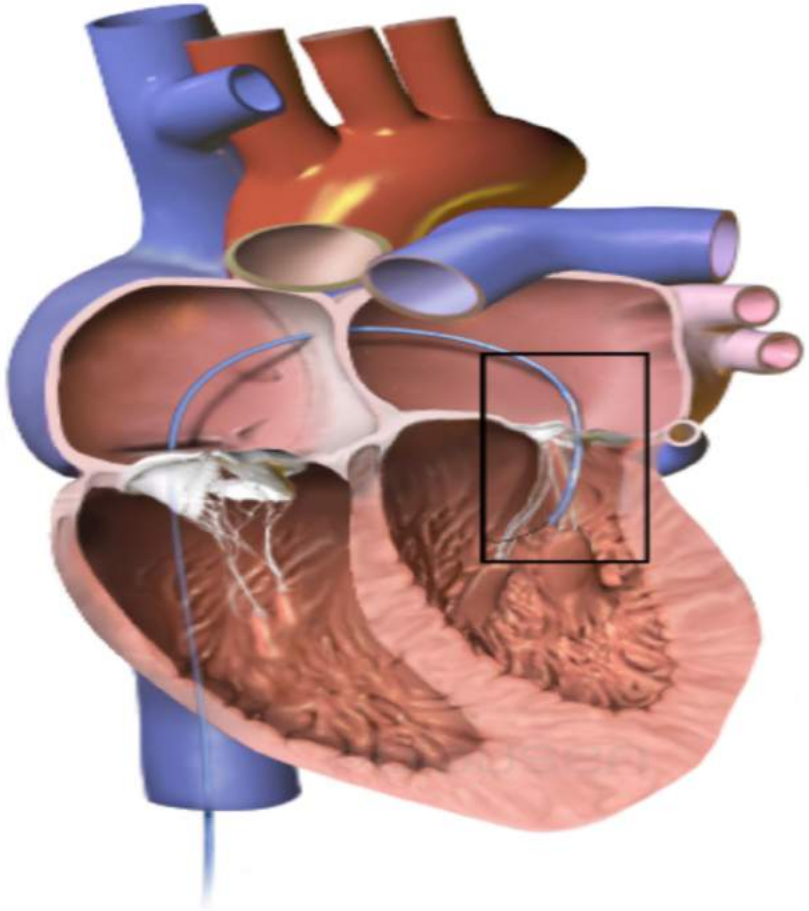
Medical

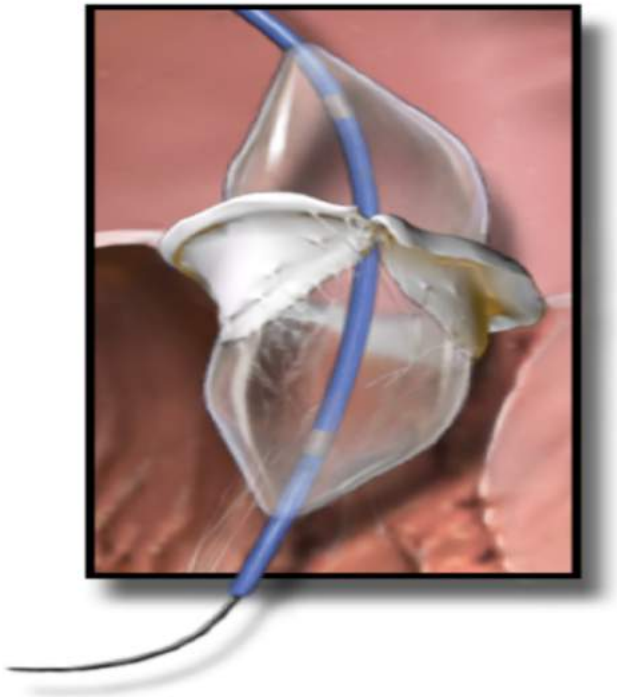
- Diuretics
- BB
- Anticoagulation

Percutaneous or Surgical intervention

- Mitral balloon valvuloplasty (percutaneous mitral balloon commissurotomy, PMBC)
- MVR

Mitral Balloon Valvuloplasty





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Mitral Valve Replacement (MVR)

Mechanical Valve

Tissue Valve

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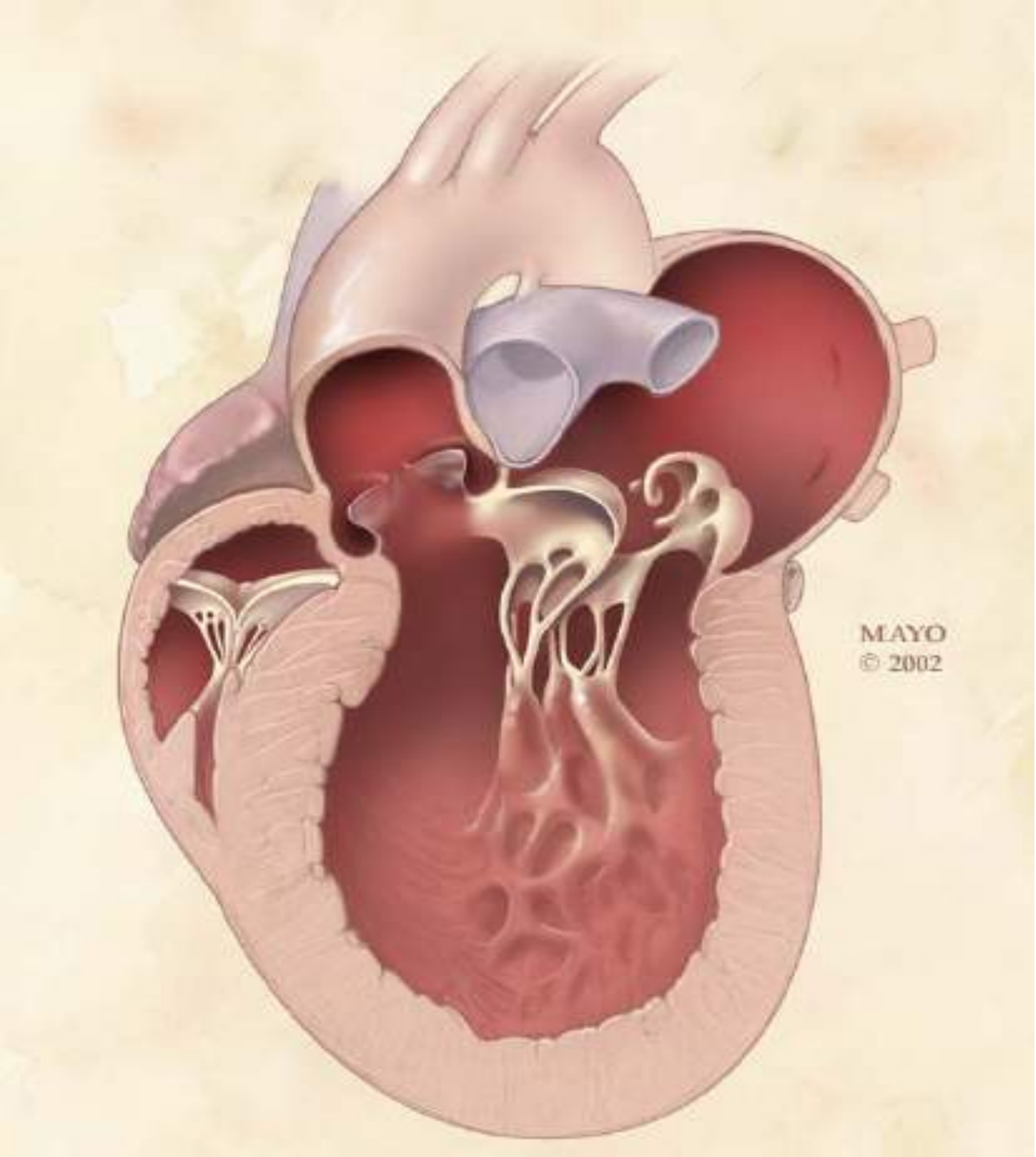

Mitral Regurgitation (MR)

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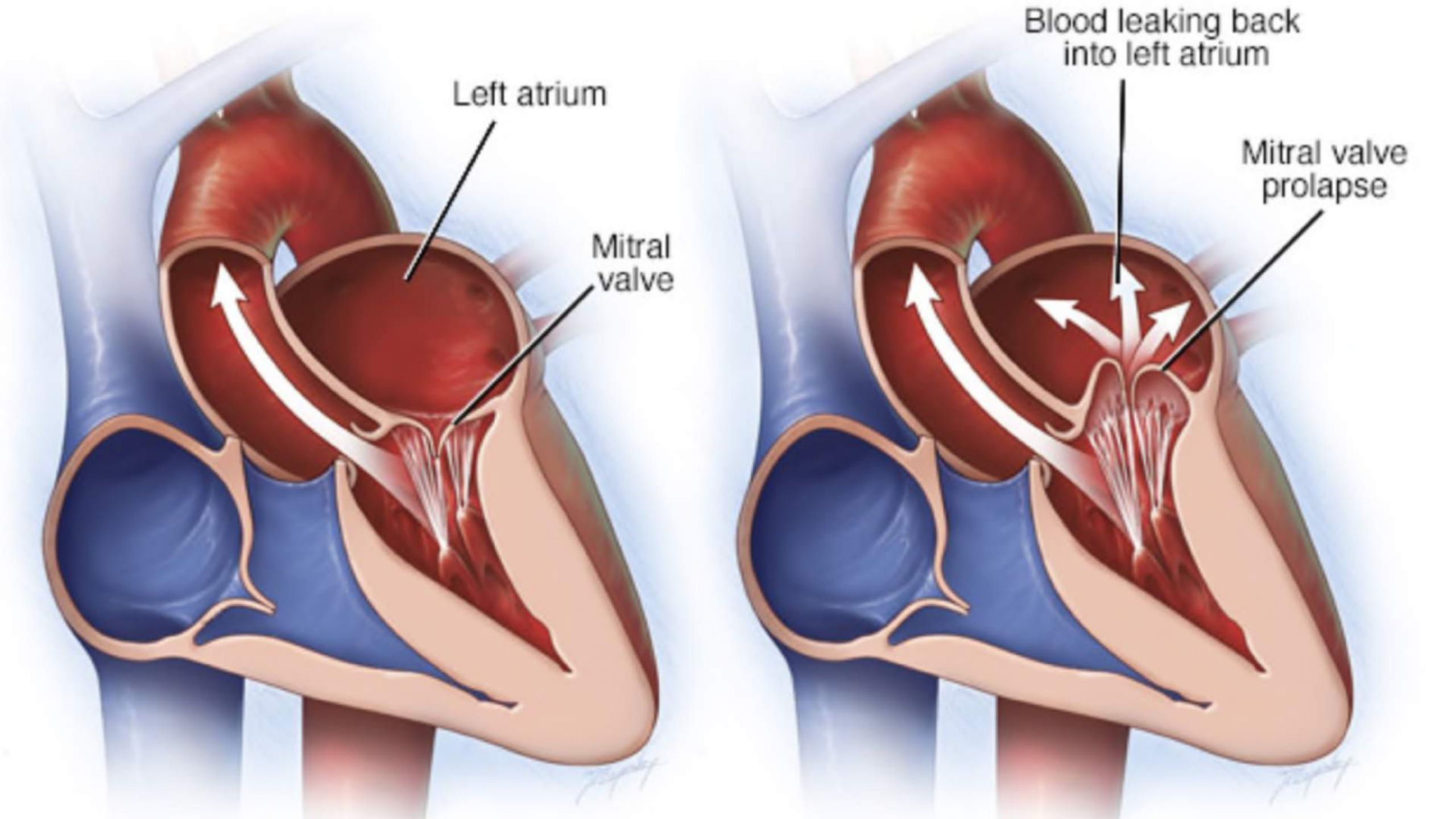
Left atrium
Mitral valve
Blood leaking back into left atrium
Mitral valve prolapse

Etiology

Acute MR	Chronic Primary MR
Endocarditis	Myxomatous (MVP)
Papillary muscle rupture (post MI)	Rheumatic Fever
Trauma	Endocarditis (Healed)
Chordal rupture/leaflet flail (MVP,IE)	Mitral annular calcifications
	Congenital (Cleft,AV canal)/ HOCM with SAM
	Radiation
	Chronic secondary MR
	Ischemic (LV remodeling)
	Dilated Cardiomyopathy

The abnormal & dilated LV causes Pap muscle displacement which in turn results in leaflet tethering with associated annular dilation that prevents coaptation

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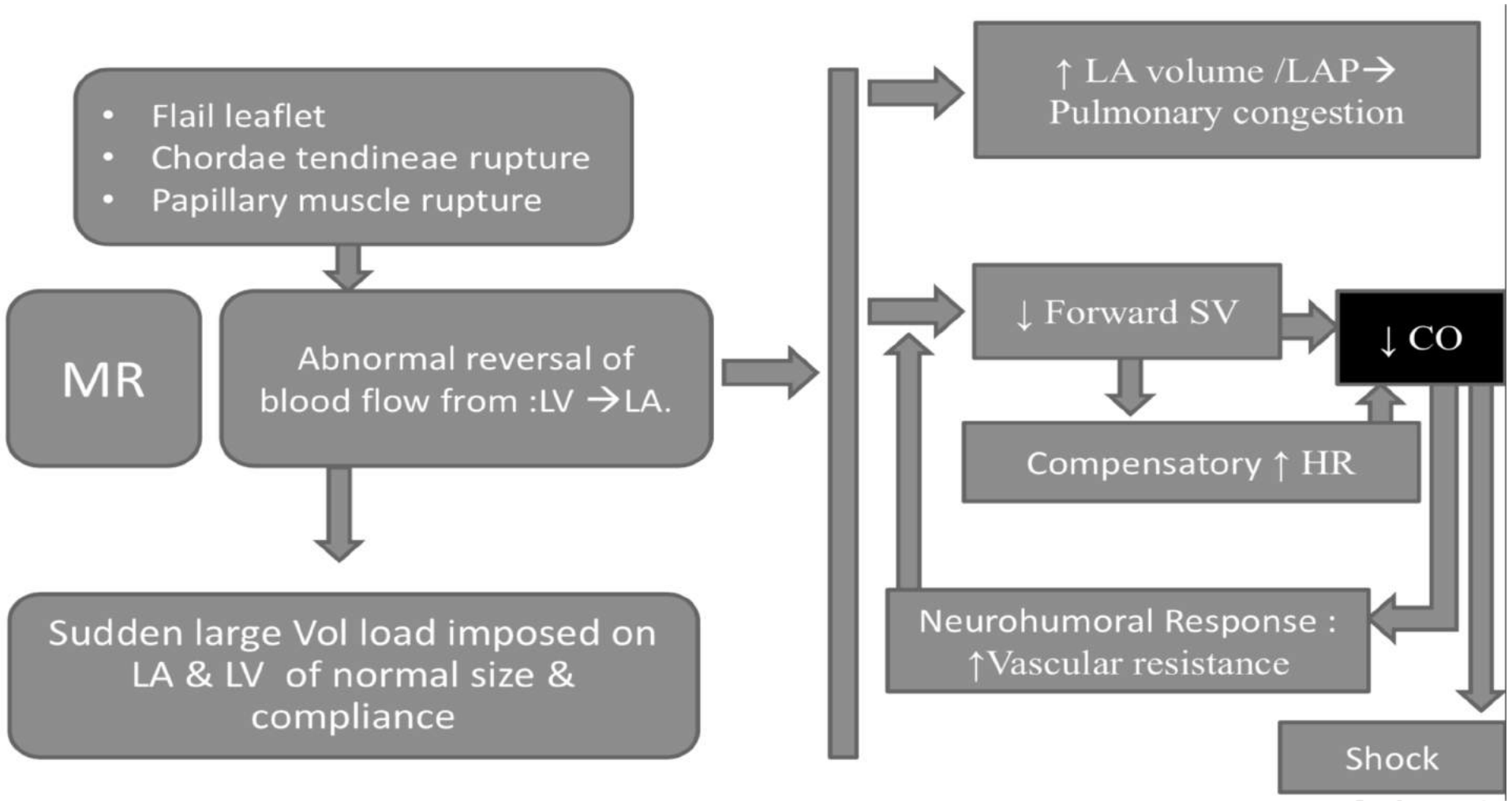
Acute MR Pathophyiology



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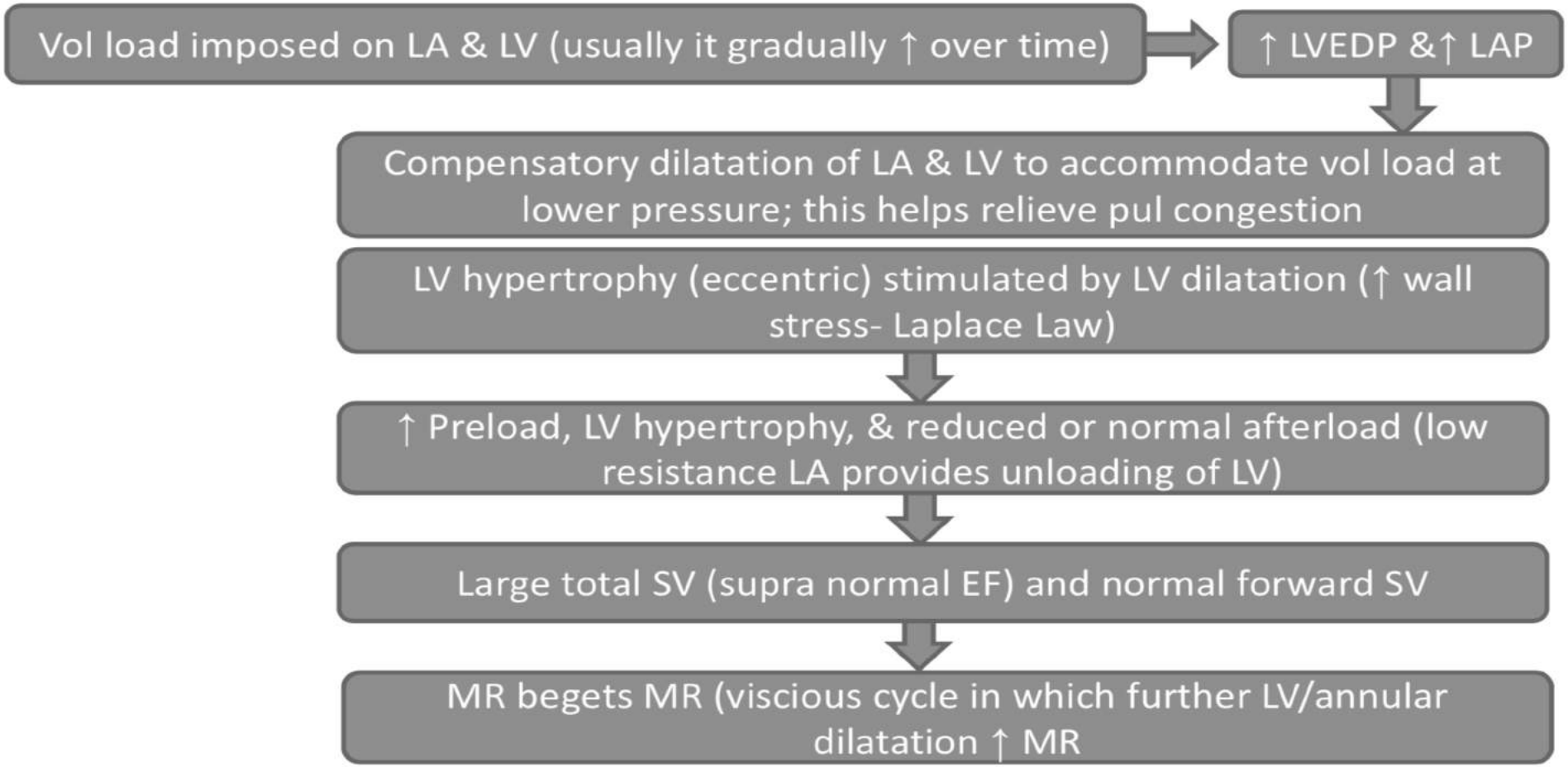
Chronic MR Pathophysiology



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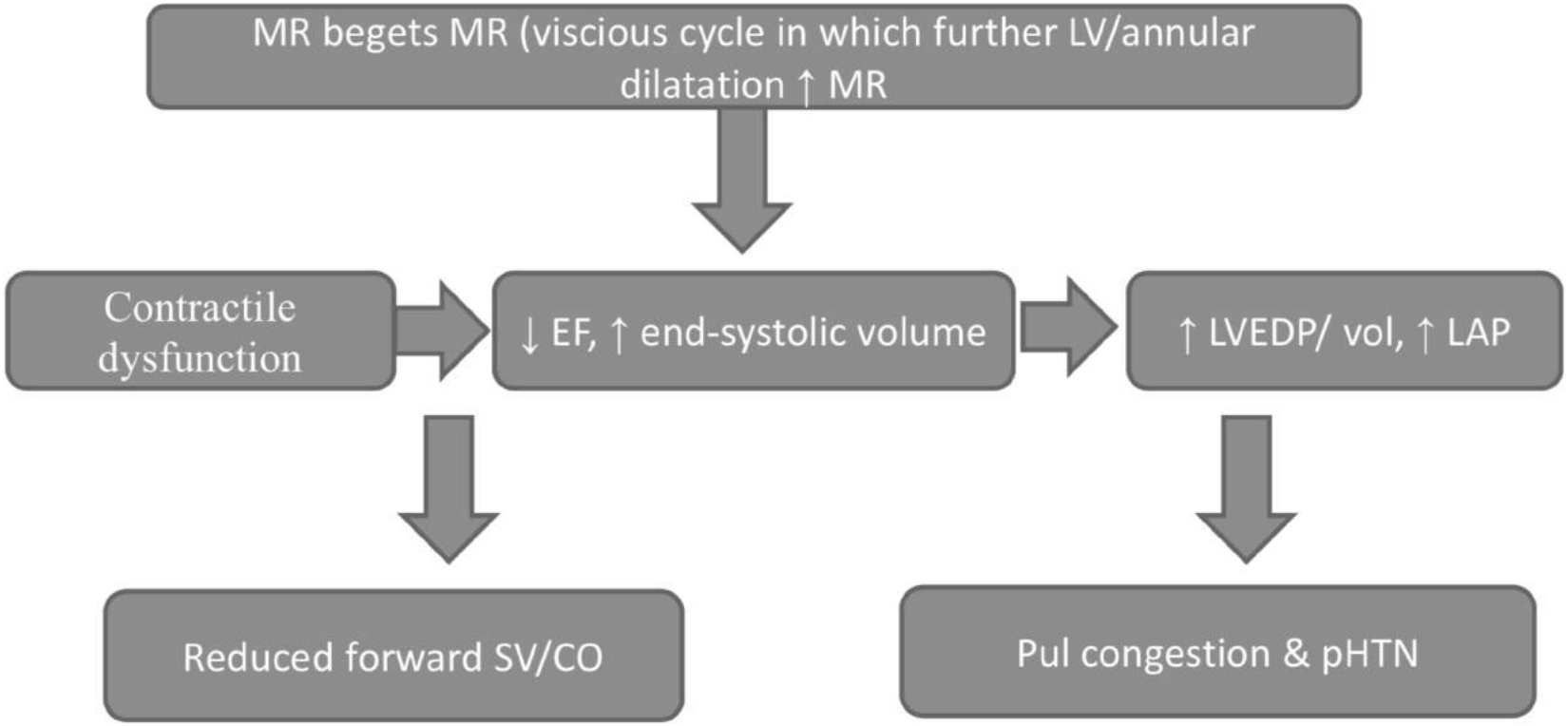
Chronic MR pathophysiology



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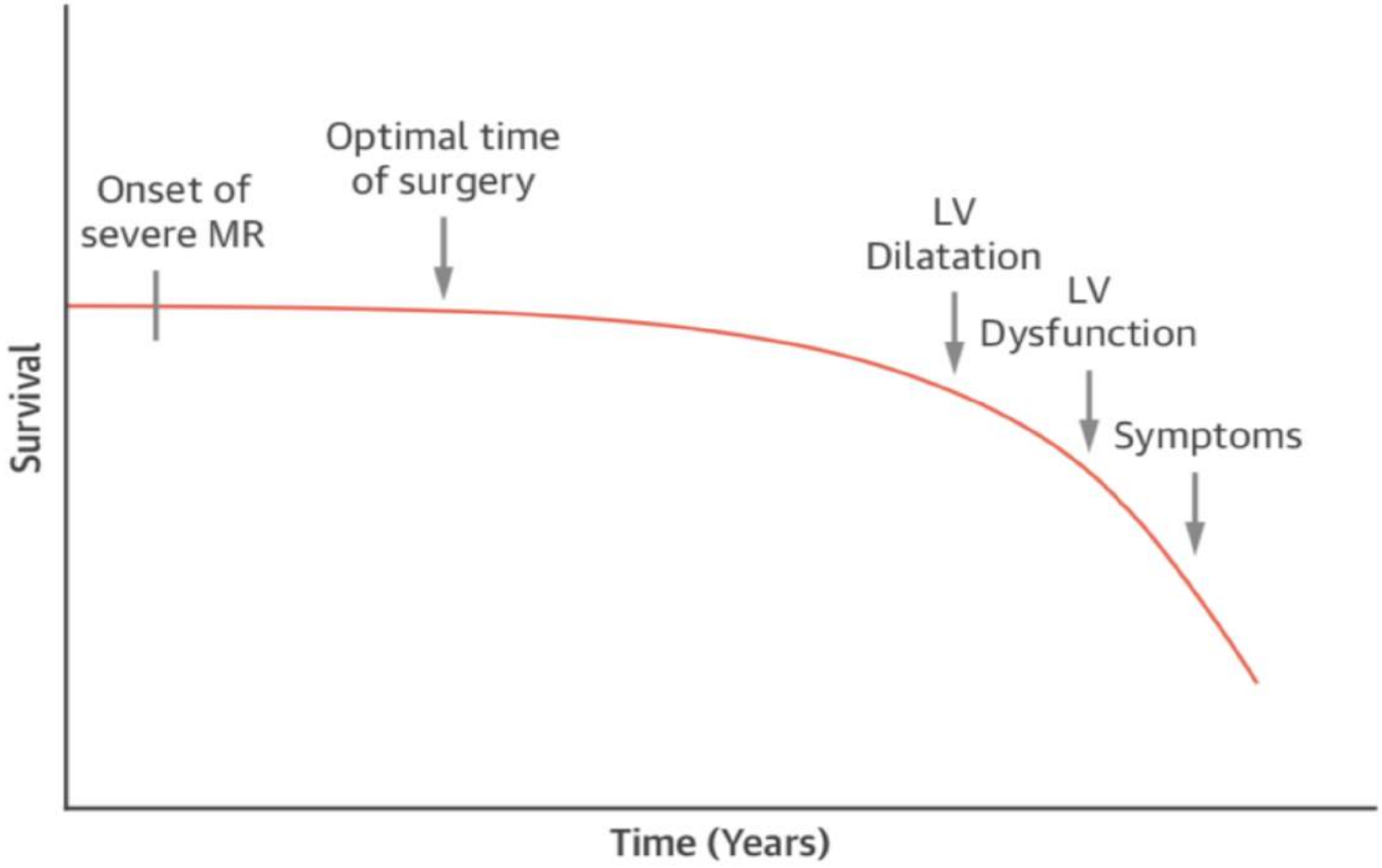
Natural History of Primary MR



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Clinical Presentation

Symptoms:

Acute MR:

- ✓ Decompensated HF symptoms: Dyspnea, orthopnea, PND)
- ✓ Low cardiac output state
- ✓ Cardiogenic shock

Chronic MR:

- ✓ Initially asymptomatic
- ✓ HF symptoms (Dyspnea, orthopnea, PND, LL edema)
- ✓ Decreased exercise tolerance
- ✓ Palpitation with AF if present
- ✓ PHTN symptoms if present

Physical exam & Clinical signs:

Pulse: Large Volume Collapsing

JVP: prominent V wave

Apex: Diffuse with Lateral displacement +/- palpable thrill

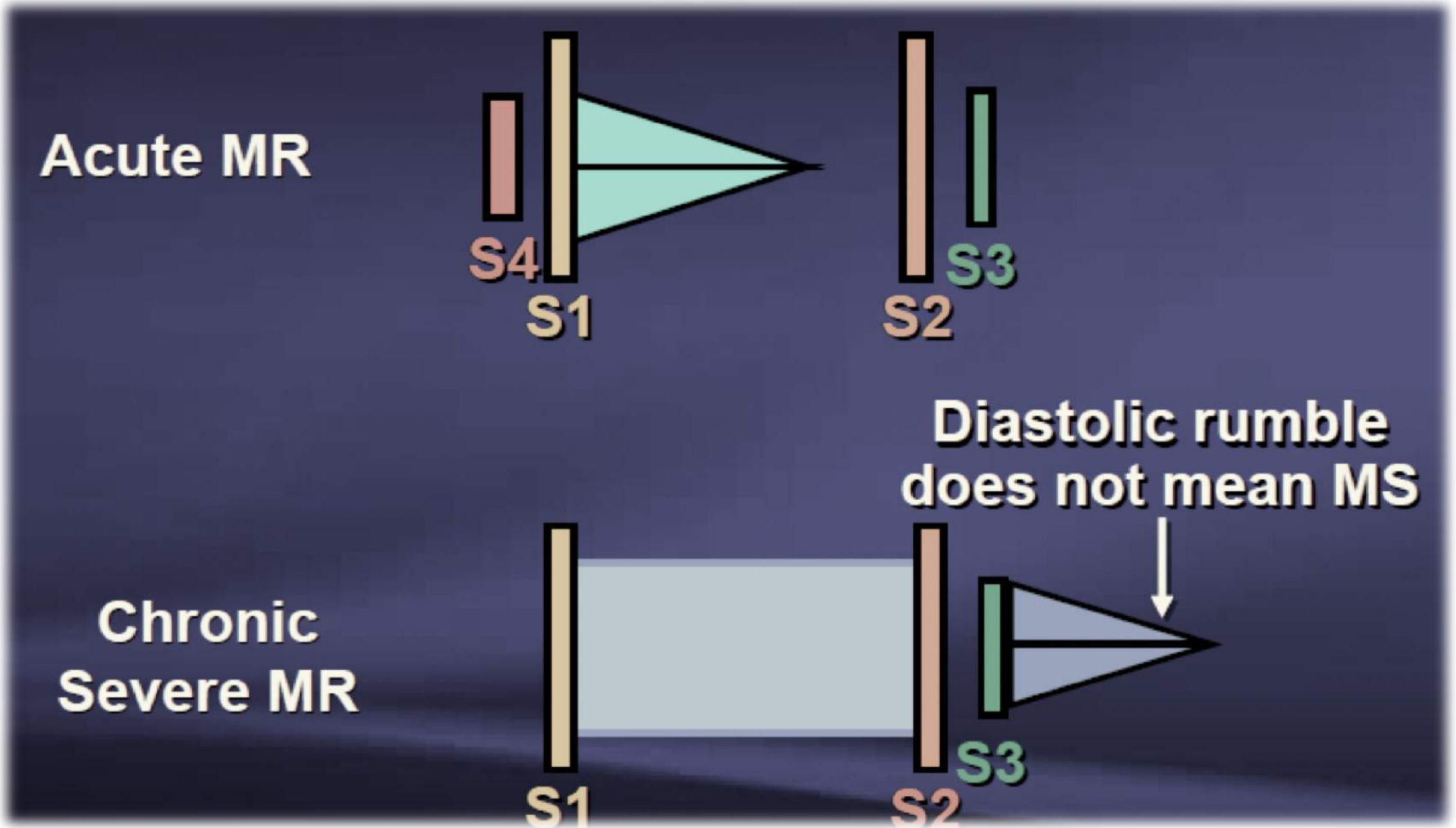
HS: Normal or Soft S1, S2 physiological split or wide split due to premature AV closure.

Extra HS: S3

Murmurs: Pansystolic (holosystolic) louder at the apex & radiates to axilla

Acute vs Chronic MR

Acute MR	Chronic NR
Tachycardia (sinus)	Normal HR (sinus AF may be present)
Low BP	N or high BP
S4	S3
Short early systolic	Holosystolic



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Investigations & Diagnosis

- Bedside assessment & observation of syndromic features.
- Bloods
- FBC
- U&Es
- Cholesterol
- Clotting
- ECG:

LAE, LVH, PHT findings, AF

Investigations & Diagnosis

Imaging

- Echocardiogram
- CXR

Special

- Cardiac catheterisation
- Exercise test

Management

Medical:

- HF medications with LV dysfunction if surgical intervention not possible.

- **Surgical/Percutaneous intervention:**
- Mitral valve repair or replacement.
- Mitral clip

Other Valve pathologies

- Tricuspid stenosis/regurgitation.
- Pulmonary stenosis/regurgitation
- Prosthetic valve dysfunction

Take home message

Over the lifetime of a patient with valvular heart disease (VHD), the most important aspects of medical care are:

- An accurate diagnosis of the cause and severity of VHD
- Measures to prevent further valve dysfunction through prevention of rheumatic fever and endocarditis
- Education about the natural history of disease, including anticipated type and timing of symptom onset
- Interval medical evaluation and imaging to monitor disease progression
- Prompt recognition and treatment of associated cardiac conditions, including (AF), HTN, (CAD), IE, and aortic dilation
- Optimal timing of surgical or transcatheter intervention to correct or ameliorate valve dysfunction.

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Thank You