"Valvular Heart Disease" 🫀

Definition

Valvular heart disease refers to structural or functional abnormalities of cardiac valves leading to altered blood flow across the valve.

TYPES OF VALVULAR DYSFUNCTION

❑ Valvular Stenosis

Definition:
Failure of a valve to open completely, causing obstruction to forward blood flow.

Key Pathogenesis:

- Almost always due to a primary cuspal abnormality
- Usually results from chronic processes

Common Causes:

- Calcification (degenerative)
- Fibrosis and scarring (e.g., rheumatic heart disease)

📌 Exam Pearl:

Valvular stenosis is *almost never acute*.

2️⃣ Valvular Insufficiency (Regurgitation / Incompetence)

Definition:
Failure of a valve to close completely, allowing backflow of blood.

Mechanisms:

- Intrinsic cuspal disease
- Disruption of support structures (even with normal cusps)

Support structures involved:

- Valve annulus
- Chordae tendineae

- Papillary muscles
- Ventricular free wall
- Aorta (for aortic valve)

Onset:

- Acute → e.g., chordal or papillary muscle rupture
- Chronic → leaflet scarring & retraction

🔁 Comparison Table: Stenosis vs Insufficiency

Feature	Stenosis	Insufficiency
Primary defect	Failure to open	Failure to close
Flow problem	Obstructed forward flow	Backward flow (regurgitation)
Usual onset	Chronic	Acute or chronic
Main cause	Cuspal abnormality	Cusps or support structures

VALVES INVOLVED

- May involve one valve (most commonly mitral valve)
- Or multiple valves

HEART SOUNDS & MURMURS 🎧

Mechanism:

Turbulent blood flow across diseased valves → murmurs

Severe lesions may produce:

- Thrills (palpable murmurs)

Factors determining murmur characteristics:

- Valve involved
- Type of lesion (stenosis vs regurgitation)
- Severity of disease
- Phase of cardiac cycle

📌 Exam Tip:

Always mention timing (systolic/diastolic) + quality (harsh/soft).

NATURAL HISTORY & OUTCOME 🧠

Outcome depends on:

- Valve involved
- Degree of impairment
- Rate of development
- Effectiveness of compensatory mechanisms

Clinical Contrast:

- Acute aortic regurgitation (infective destruction)
  → sudden volume overload → acute cardiac failure
- Rheumatic mitral stenosis
  → slow progression over years → symptoms appear late

CONGENITAL vs ACQUIRED VALVULAR DISEASE

Congenital Valvular Disease

⭐ Bicuspid Aortic Valve (Most common)

- 2 cusps instead of 3
- Occurs in 1–2% of live births

- Cusps are unequal in size
- Larger cusp shows a midline raphe

Functional course:

- Normal early life
- With age → ↑ susceptibility to degenerative calcification → aortic stenosis

📌 Exam Favorite Line:

"Bicuspid aortic valve predisposes to early-onset calcific aortic stenosis."

Acquired Valvular Disease

- Accounts for majority of valvular pathology
- Aortic + mitral stenosis ≈ two-thirds of all valve disease

DEGENERATIVE VALVE DISEASE 🧓

Definition

Age-related disorders affecting valvular extracellular matrix (ECM) due to repetitive mechanical stress (~40 million valve cycles per year!)

Degenerative Changes

1️⃣ Calcification

- Cuspal calcification → aortic valve
- Annular calcification → mitral valve

📌 Mitral annular calcification is usually asymptomatic

⚠️ May cause conduction defects if it encroaches on conduction system

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2️⃣ ECM Alterations

Two patterns:

- Myxomatous degeneration
  → ↑ proteoglycans
  → ↓ collagen & elastin
- Fibrotic degeneration
  → thickened, scarred valve

CALCIFIC AORTIC STENOSIS 🫀

Overview

- Most common cause of aortic stenosis
- Often asymptomatic initially
- Increasing incidence with aging population

Age of Presentation

- Normal tricuspid valve → 70s–80s
- Bicuspid valve → 40s–50s

Pathogenesis (Flowchart) 🔁

Risk factors (male sex, ↑ LDL, HTN, smoking) → Lipoprotein deposition in valve → Local inflammation → Endothelial injury → Osteoblastic differentiation of valve cells → Progressive calcification → Aortic stenosis

📌 Link to Atherosclerosis:
Mechanistically similar — not just wear and tear

MORPHOLOGY 🔬

Hallmark features:

- Heaped-up calcified masses
- Located on outflow (aortic) side of cusps
- Project into sinuses of Valsalva
- No commissural fusion (important differentiator!)

📌 Commissural fusion → suggests rheumatic disease, not degenerative.

CLINICAL FEATURES & HEMODYNAMICS

Hemodynamic Consequences (Flowchart)

Aortic valve narrowing → Outflow obstruction → ↑ LV systolic pressure → Concentric LV hypertrophy → ↓ Coronary perfusion → Myocardial ischemia → angina → LV dysfunction → CHF

Key Clinical Facts

- Valve orifice may be reduced by 70–80%
- LV pressures may reach ≥200 mm Hg
- Hypertrophied myocardium is ischemia-prone

Classic Triad of Severe Aortic Stenosis ⚠️

- Angina
- Syncope
- Congestive heart failure

📌 Prognosis:

Once symptoms appear → poor prognosis

Without surgery:

- 50–80% mortality within 2–3 years

ETIOLOGY OF ACQUIRED VALVULAR DISEASE 📊

Mitral Valve Disease	Aortic Valve Disease
-> Mitral Stenosis	-> Aortic Stenosis
Rheumatic scarring	Rheumatic scarring
	Senile calcific stenosis
	Calcified bicuspid valve
-> Mitral Regurgitation	-> Aortic Regurgitation
Infective endocarditis	Rheumatic disease
Mitral valve prolapse	Infective endocarditis
Papillary muscle rupture	Aortic root dilation

Chordae rupture	Syphilitic aortitis
LV dilation	Marfan syndrome
Mitral annular calcification	Ankylosing spondylitis
Fen-phen-induced fibrosis	Rheumatoid arthritis

MITRAL VALVE PROLAPSE

(Myxomatous Mitral Valve Disease) 🫀

Definition

Mitral valve prolapse (MVP) is a condition in which one or both mitral valve leaflets are abnormally floppy and balloon (prolapse) back into the left atrium during systole.

TYPES OF MITRAL VALVE PROLAPSE

❑ Primary Mitral Valve Prolapse

- Idiopathic

- Associated with myxomatous degeneration of the mitral valve
- Prevalence: 0.5%–2.4% of adults
- Can be sporadic or familial
- One of the most common valvular heart diseases

2️⃣ Secondary Mitral Valve Prolapse

- Occurs secondary to identifiable genetic disorders
- Classically associated with:
  - Marfan syndrome 🧬

📌 Exam Tip:

Always mention *Marfan syndrome* when asked about secondary MVP.

MORPHOLOGY 🔬

Gross Features

- Ballooning (hooding) of mitral leaflets into left atrium
- Leaflets are:
  - Enlarged
  - Redundant
  - Thick
  - Rubbery
- Chordae tendineae:
  - Elongated
  - Thinned
  - May rupture (important cause of acute MR)

📌 Associated valve involvement (Primary MVP):

- Tricuspid valve → 20–40%
- Aortic and pulmonic valves → less common

Histopathology 🧠

Key structural layers affected:

Valve Layer	Change

Fibrosa	Thinning (loss of tensile strength)
Spongiosa	Expansion due to ↑ myxomatous (mucoid) material

📌 These changes are identical whether MVP is:

- Primary (intrinsic ECM defect)
- Secondary (due to regurgitation from another cause)

PATHOGENESIS (Flowchart) 🔁

Extracellular matrix abnormality → Weak fibrosa layer → Excess myxomatous material in spongiosa → Leaflet redundancy & elongation → Ballooning into left atrium during systole → Mitral regurgitation

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CLINICAL FEATURES 🩺

Presentation

- Most patients are asymptomatic
- Often discovered incidentally

Symptomatic patients may have:

- Palpitations
- Dyspnea
- Atypical chest pain

Auscultatory Findings 🎧

- Mid-systolic click
  → Due to sudden tension on redundant leaflets and chordae
- May be followed by a late systolic regurgitant murmur

📌 Classic Viva Line:

"Midsystolic click is characteristic of mitral valve prolapse."

COMPLICATIONS ⚠️

Seen in ~3% of patients

- Hemodynamically significant mitral regurgitation
- Congestive heart failure
- Chordal or leaflet rupture
- Infective endocarditis
- Ventricular arrhythmias → sudden cardiac death
- Thromboembolism → stroke or systemic infarction

RHEUMATIC VALVULAR DISEASE ❤️‍🔥

Overview

Rheumatic fever is an acute, immune-mediated, multisystem inflammatory disease that follows group A β-hemolytic streptococcal infection (usually pharyngitis).

Rheumatic heart disease (RHD) is the cardiac manifestation of rheumatic fever.

📌 Key Concept:

All layers of the heart are involved, but valvular damage determines prognosis.

EPIDEMIOLOGY 🌍

- Rare in high-income countries
- Still most common acquired heart disease in:
  - Children
  - Young adults

  in low-income countries

PATHOGENESIS 🧠 (High-Yield)

Immune-Mediated Mechanism (Flowchart)

Group A streptococcal infection → Antibodies & T cells against M protein → Molecular mimicry with cardiac proteins → Cross-reactive immune response → Complement activation & macrophage recruitment → Cytokine release → Acute inflammation (Aschoff bodies)

→ Healing with fibrosis → Chronic rheumatic heart disease

📌 Important Exam Point:

Streptococci are absent from cardiac lesions.

MORPHOLOGY

ACUTE RHEUMATIC FEVER 🔬

Pathognomonic Lesion: Aschoff Body

- Foci of inflammation consisting of:
  - T lymphocytes
  - Plasma cells
  - Activated macrophages (Anitschkow cells)

Anitschkow cells:

- Abundant cytoplasm
- Nucleus with central wavy chromatin
  → "Caterpillar cell" 🐛

Pancarditis (All 3 Layers Involved)

- Pericardium:
  - Fibrinous pericarditis
  - Usually resolves without sequelae
- Myocardium:
  - Scattered Aschoff bodies → myocarditis
- Endocardium (Valves):
  - Fibrinoid necrosis along lines of closure
  - Small vegetations (1–2 mm) called verrucae

CHRONIC RHEUMATIC HEART DISEASE 🫀

Structural Changes

- Replacement of Aschoff bodies by fibrous scars
- Valves show:
  - Leaflet thickening
  - Commissural fusion

- Shortened, thickened, fused chordae tendineae

📌 Classic Gross Appearance:

"Fish-mouth" or "buttonhole" mitral stenosis

FUNCTIONAL CONSEQUENCES ⚙️

Valve Involvement Frequency

- Mitral valve alone → 70%
- Mitral + aortic valves → 25%
- Tricuspid → less common
- Pulmonic → almost never involved

Hemodynamic Progression (Flowchart) 🔁

Mitral stenosis → ↑ Left atrial pressure → Left atrial dilation → Atrial fibrillation → Mural thrombus formation → Systemic embolization → Pulmonary

congestion → Pulmonary hypertension → Right ventricular hypertrophy → Right-sided heart failure

📌 Important Note:

In pure mitral stenosis, the left ventricle remains normal.

FINAL EXAM TAKEAWAYS 📌

- MVP → floppy valve, midsystolic click, usually benign
- Rheumatic heart disease → only cause of acquired mitral stenosis
- Fish-mouth valve + Aschoff bodies = diagnostic clues

ACUTE RHEUMATIC FEVER

Clinical Features 🩺

Age Distribution

- Most common: Children 5–15 years
- ~20% of first attacks occur in adults

📌 Age-based pattern:

- Children → Carditis predominates
- Adults → Arthritis predominates

CARDIAC MANIFESTATIONS 💓

Principal clinical manifestation: Carditis

Clinical Signs of Carditis

- Pericardial friction rub
- Arrhythmias
- Myocarditis → may be severe

Hemodynamic Consequences (Flowchart) 🔁

Myocardial inflammation → Cardiac dilation → Functional mitral regurgitation → Reduced cardiac output → Congestive heart failure (CHF)

📌 Mortality:

- <1% die during the acute phase

SYSTEMIC FEATURES 🌡️

Symptom Onset

- Begins 2–3 weeks after streptococcal infection
- Initial features:
  - Fever
  - Migratory polyarthritis

Migratory Polyarthritis (Classic Description)

- Affects large joints
- One joint involved at a time
  → Pain & swelling for days
  → Spontaneous resolution
  → No residual deformity

📌 Exam Phrase:

"Migratory polyarthritis with complete recovery"

LABORATORY FINDINGS 🧪

- Throat cultures: Negative at symptom onset
- Evidence of recent streptococcal infection:
  - ↑ Anti-streptolysin O (ASO) titers
  - ↑ Anti-DNase B

📌 Important Concept:

Disease is immune-mediated, not due to active infection.

DIAGNOSIS: JONES CRITERIA 📋

Diagnostic Requirement

Evidence of recent streptococcal infection

➕

2 Major OR 1 Major + 2 Minor criteria

MAJOR (JONES) CRITERIA ⭐

1. Carditis
2. Migratory polyarthritis (large joints)
3. Subcutaneous nodules
4. Erythema marginatum (annular rash)
5. Sydenham chorea
    - Involuntary, purposeless, rapid movements 🧠

MINOR CRITERIA

- Fever
- Arthralgia
- ECG changes (e.g., prolonged PR interval)
- Elevated acute-phase reactants (ESR, CRP)

📌 Exam Tip:

Sydenham chorea may appear late and sometimes as the only manifestation.

RECURRENT RHEUMATIC FEVER 🔁

Immunologic Basis (Flowchart)

Initial streptococcal infection → Immune memory formation → Subsequent streptococcal exposure → Exaggerated immune response → Recurrent rheumatic fever → Progressive valvular damage

📌 Key Point:

Cardiac damage is cumulative with each recurrence.

CHRONIC RHEUMATIC HEART DISEASE 🫀

Latency

- Often clinically silent for years to decades
- Symptoms appear late due to progressive valve scarring

CLINICAL FEATURES OF CHRONIC RHD

Cardiac Manifestations

- Cardiac murmurs
- Cardiac hypertrophy & dilation
- Congestive heart failure

Rhythm & Thromboembolic Complications

- Atrial fibrillation (especially in mitral stenosis)
- Left atrial mural thrombi
  → Systemic embolization (stroke, infarcts)

Additional Risks ⚠️

- Scarred valves → ↑ risk of infective endocarditis

📌 Prognosis:

- Highly variable

- Mitral valvuloplasty / valve replacement has markedly improved outcomes

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INFECTIVE ENDOCARDITIS (IE) 🦠❤️

Definition

Infective endocarditis is a microbial infection of the endocardium, most commonly involving heart valves, resulting in vegetations composed of thrombotic debris and microorganisms, often with destruction of underlying tissue.

SITES OF INFECTION

- Native heart valves
- Prosthetic valves
- Endocardium
- Aorta

- Aneurysmal sacs
- Prosthetic devices

ETIOLOGY 🧪

Common Causes

- Bacteria → vast majority of cases

Rare Causes

- Fungi
- Rickettsiae (Q fever)
- Chlamydial species

CLASSIFICATION OF INFECTIVE ENDOCARDITIS

Basis of Classification

- Tempo & severity of disease
- Virulence of organism
- Presence of pre-existing heart disease

📌 Clear separation between types is not always possible.

TYPES OF INFECTIVE ENDOCARDITIS

1️⃣ Acute Infective Endocarditis ⚠️

- Rapid onset
- Highly destructive infection
- Occurs on normal or abnormal valves
- High morbidity & mortality
- Poor prognosis even with treatment

📌 Typical organisms: Highly virulent (e.g., *Staphylococcus aureus*)

2️⃣ Subacute Infective Endocarditis 🐢

- Insidious onset
- Prolonged course (weeks to months)
- Usually occurs on previously abnormal valves

- Better prognosis
- Most patients recover with antibiotics

📌 Typical organisms: Less virulent (e.g., *Streptococcus viridans*)

ACUTE vs SUBACUTE IE (Quick Comparison) 📊

Feature	Acute IE	Subacute IE
Onset	Sudden	Insidious
Course	Rapid, destructive	Prolonged
Valve status	Normal or diseased	Usually diseased
Organism virulence	High	Low
Prognosis	Poor	Better

📌 Exam Line:

> "Rheumatic heart disease predisposes to infective endocarditis due to chronically scarred valves."

PATHOGENESIS 🦠🫀

Predisposing Cardiac Conditions

IE may occur on normal valves, but pre-existing cardiac abnormalities greatly increase risk.

Common substrates:

- Rheumatic heart disease
- Mitral valve prolapse (now the most common predisposing factor)
- Bicuspid aortic valve
- Calcific valvular stenosis

📌 Decline in rheumatic disease → MVP has become the leading risk factor.

Prosthetic & Iatrogenic Factors

- Prosthetic heart valves → 10–20% of IE cases
- Pacemaker leads

- Indwelling vascular catheters

Role of Endothelial Injury

Sterile platelet–fibrin thrombi form at sites of:

- Endocardial damage by turbulent flow ("jets")
- Foreign material (catheters, pacemaker lines)

These serve as niduses for bacterial adhesion.

Host Risk Factors ⚠️

- Neutropenia
- Immunodeficiency
- Malignancy
- Diabetes mellitus
- Alcohol abuse
- Intravenous drug use

📌 These factors increase both risk and severity of IE.

MICROBIOLOGY 🧪

Most Common Causative Organisms (Worldwide)

1. Staphylococci
2. Streptococci
3. Enterococci

Organisms by Clinical Setting

Community-Acquired IE

- Streptococcus viridans (50–60%)
- Normal oral flora
- Typically infects previously damaged valves
- Causes subacute IE

Healthcare-Associated IE & IV Drug Users

- Staphylococcus aureus

- Highly virulent
- Infects normal or abnormal valves
- Causes acute IE

📌 Exam Pearl:

S. aureus is now the most common cause of IE in high-income countries.

Other Organisms

- Enterococci
- HACEK group (oral commensals): Haemophilus, Actinobacillus, Cardiobacterium, Eikenella, Kingella
- Rare: Gram-negative bacilli, fungi

Culture-Negative Endocarditis

- ~10% of cases
- Causes:

- Prior antibiotic therapy
- Fastidious organisms

SOURCE OF BACTEREMIA 🔁

Portal of Entry (Flowchart)

→ Bacteremia

→ Dental / surgical procedures

→ IV drug use

→ Infected indwelling catheters

→ Occult oral or GI source

→ Trivial mucosal injuries

→ Seeding of damaged endocardium

→ Infective endocarditis

📌 Recognition of high-risk situations allows antibiotic prophylaxis.

MORPHOLOGY 🔬

Valvular Lesions

- Friable, bulky vegetations
- Composed of:
  - Fibrin
  - Inflammatory cells
  - Microorganisms

Valves Commonly Involved

- Aortic valve
- Mitral valve
- Tricuspid valve → common in IV drug users

Complications of Vegetations ⚠️

- May be single or multiple

- May involve more than one valve
- Can erode into myocardium → ring abscess

Embolic Phenomena (Flowchart) 🔁

Friable vegetation → Fragmentation → Septic emboli →
Lodgement in distant organs → Septic infarcts →
Infected arterial wall → Mycotic aneurysm

📌 Vegetations are highly embolic due to friability.

CLINICAL FEATURES 🩺

General Features

- Fever → most consistent sign
- Malaise
- Weight loss

Acute vs Subacute Presentation

Acute IE

- Rapid onset
- High fever
- Chills
- Severe systemic toxicity

Subacute IE

- Insidious onset
- May lack fever (especially elderly)
- Fatigue
- Weight loss
- Flu-like illness
- Splenomegaly common

Cardiac Findings 🎧

- Murmurs in ~90% of patients with left-sided IE

Peripheral Stigmata of IE 🖐️👁️

Due to microemboli and immune phenomena:

- Petechiae
- Splinter hemorrhages (nail beds)
- Roth spots (retinal hemorrhages)
- Janeway lesions → painless, erythematous lesions on palms/soles
- Osler nodes → painful nodules on fingertips

📌 Viva Tip:

Janeway = painless | Osler = painful

Diagnostic Confirmation ✅

- Positive blood cultures
- Echocardiography showing vegetations

PROGNOSIS & COMPLICATIONS ⚠️

Early Complications (Weeks)

- Glomerulonephritis → immune complex deposition → hematuria, albuminuria, renal failure
- Septicemia
- Arrhythmias (suggest myocardial extension)
- Systemic embolization

Prognostic Factors

- Causative organism
- Extent of valvular destruction
- Presence of embolic or renal complications

📌 Untreated IE is usually fatal.

Outcome

- Antibiotic therapy ± valve replacement
- Significantly reduces mortality

FINAL EXAM SUMMARY 📌

- MVP is now the most common predisposing lesion
- *S. aureus* → acute, aggressive IE
- *S. viridans* → subacute IE on damaged valves
- Friable vegetations → emboli → mycotic aneurysms
- Fever + murmur + embolic signs = think IE

🫀 Noninfected Vegetations 🫀

Nonbacterial Thrombotic Endocarditis (NBTE)

Definition:

NBTE is characterized by the deposition of sterile thrombi on cardiac valves, most commonly occurring in patients with an underlying hypercoagulable state. Unlike infective endocarditis, these vegetations are noninfectious and nondestructive.

🔬 Key Morphological Features

- Vegetations:
  - Small, friable thrombi
  - Size: ~ 1–5 mm
  - Sterile (no microorganisms)
- Valve involvement:
  - Usually on previously normal valves
  - No significant valve destruction
- Inflammation:
  - Absent or minimal, explaining easy embolization

⚠️ Etiology & Predisposing Conditions

NBTE is strongly associated with hypercoagulable states.

Most common cause:

- 🧬 Underlying malignancy, especially mucinous adenocarcinomas
  - Mechanism: circulating mucin and tumor-derived procoagulants

Other predisposing conditions:

- Chronic disseminated intravascular coagulation (DIC)
- Hyperestrogenic states (e.g., pregnancy, OCP use)
- Endocardial trauma
    - e.g., indwelling intravascular catheters
- Severe systemic illness with debility or wasting

🧠 Pathogenesis (Flowchart)

Underlying hypercoagulable state → Endothelial surface becomes prothrombotic → Deposition of platelets + fibrin on valve surface → Formation of small, sterile vegetations → Minimal/no inflammation → Friable vegetations prone to embolization 🚨

🚑 Clinical Significance

- Local valvular dysfunction:
    - Usually trivial or absent
- Major complication:

- ⚡ Systemic embolization
  - Brain → cerebral infarcts
  - Heart → myocardial infarction
  - Other organs → splenic, renal infarcts
- Important exam point:
  - NBTE vegetations can act as a nidus for bacterial colonization, leading to secondary infective endocarditis

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📊 NBTE vs Infective Endocarditis (High-Yield Table)

Feature	NBTE	Infective Endocarditis
Cause	Hypercoagulable state	Infection (bacteria/fungi)
Vegetations	Sterile	Infected
Valve destruction	❌ Absent	✅ Present

Inflammation	Minimal/none	Prominent
Embolization	Very common	Common
Systemic signs	Usually absent	Fever, sepsis

Endocarditis in Systemic Lupus Erythematosus

Libman-Sacks Endocarditis 🧠🩺

Definition:

A special form of NBTE seen in patients with systemic lupus erythematosus (SLE), characterized by sterile vegetations associated with immune-mediated inflammation.

🔬 Pathogenesis (Flowchart)

SLE → Immune complex deposition on valve surface → Local inflammation → Fibrinoid necrosis of valve tissue → Sterile vegetations form → Healing with fibrosis → Valve deformity resembling chronic rheumatic heart disease

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🧩 Morphological Features

- Vegetations:
  - Sterile but associated with inflammation
- Location (very important):
  - Either surface of valve leaflets
  - Chordae tendineae
  - Atrial or ventricular endocardium
- Histology:
  - Immune complex deposition
  - Fibrinoid necrosis
  - Subsequent fibrosis

⚠️ Clinical Importance

- May cause significant valvular deformity
- Lesions can mimic chronic rheumatic heart disease
- Increased risk of thromboembolism
- Can be complicated by superimposed infective endocarditis

🧬 Related Condition (Exam Favorite ⭐)

- Antiphospholipid antibody syndrome
  - Can produce similar sterile vegetations
  - Also associated with hypercoagulability and embolic events

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📝 One-Line Exam Pearls

- NBTE: Sterile, noninflammatory vegetations → embolization risk
- Most common NBTE cause: Mucinous adenocarcinoma

- Libman-Sacks: Immune complex-mediated NBTE in SLE, can affect both sides of valves.

<-- The End -->