심실중격결손의 이해

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Definition and prevalance

- **Definition**
  - VSD = hole between ventricles; may occur at any septal site, even beneath pulmonary valve leaflets.

- **Associations**
  - May coexist with PDA, coarctation, aortic insufficiency, TOF, DORV, truncus arteriosus, transposition, AV anomalies.

Definition and prevalance

Epidemiology:

- Most common congenital heart disease (CHD) in childhood (~0.5% population).
- U.S. (2008): 31.4% of CHD cases were VSD.
- Paris cohort (2005–2008): 52% of neonates with CHD had VSD.

Maternal/Genetic Risks:

- Offspring recurrence risk: ~6% if mother has CHD.
- Familial/genetic component evident in recurrence studies.

Environmental Risks:

- Maternal alcohol or marijuana use ↑ risk of isolated VSD.
- Maternal conditions: epilepsy (carbamazepine), migraine, chronic hypertension, supraventricular tachycardia linked to higher risk.

Embryology and Pathologic Anatomy

- **Embryology of Interventricular Communication**
  - Appears during 6–8 weeks of fetal life.
  - Floor = muscular septal crest; roof = inner heart curvature.
  - Initially: double inlet to LV, double outlet from RV.
  - Interventricular communication acts as LV outlet until subaortic outlet transfers to LV.
  - Closure achieved by tissue growth from AV cushions → membranous septum.

Embryology and Pathologic Anatomy

- **Developmental Errors → VSD**
  - Failure of RV inlet transfer.
  - Failure of arterial root separation / aortic transfer.
  - Failure of interventricular communication closure.
  - Abnormal muscular septum formation (compaction defects).

Embryology and Pathologic Anatomy

- **Classification Evolution**
  - Historic numeric classification: 4 types.
  - Modern approach: emphasizes **anatomic borders**.
  - Three main phenotypic types:
    - **Juxtapulmonary**
    - **Perimembranous**
    - **Muscular**
  - Location relative to RV landmarks and septal malalignment also important.
  - Surgical relevance: border-based classification predicts **AV conduction axis location**.

Traditional “Type I–IV” Classification

- **Type I (Juxtapulmonary / Doubly Committed Outlet Defect)**
  - Beneath pulmonary valve; aortic–pulmonary fibrous continuity (conal hypoplasia).
  - Opens between limbs of septal band.
  - Posteroinferior muscular rim protects conduction axis.
  - Surgical risk ↑ if extends to central ventricular base.

Traditional “Type I–IV” Classification

- **Type II (Perimembranous / Central Defect)**
  - At central ventricular base (site of embryonic interventricular communication).
  - Fibrous continuity between aortic and tricuspid valves.
  - Conduction axis at posteroinferior margin → **high surgical risk**.
  - Most common VSD during surgery.

Traditional “Type I–IV” Classification

- **Type III (Inlet Defect / Canal-Type Variant)**
  - Malalignment between atrial and ventricular septum.
  - Associated with straddling/overriding tricuspid valve.
  - Resembles AV canal defect but with separate AV junctions.
  - Conduction axis displaced from normal Koch’s triangle → anomalous node.

Traditional “Type I–IV” Classification

- **Type IV (Muscular Defects)**
  - Exclusively muscular borders; variable geography.
  - Locations: inlet, outlet, apical, basal.
  - May appear multiple (“Swiss cheese septum”).
  - Surgical challenge: thin muscular strands between multiple defects.

Special Variants

- **Gerbode Defect**: LV → RA shunt.
  - Direct (true) form = membranous septum deficiency.
  - Indirect form = central VSD with tricuspid leaflet deficiency.
  - Rare; surgical closure needed, excellent outcomes.

Associated Pathophysiology

- **Aortic valve prolapse/regurgitation** → often with juxtapulmonary or malalignment outlet defects.
- **Outflow tract obstruction** → from septal malalignment:
  - Anterocephalad malalignment → TOF.
  - Posterocaudal malalignment → subaortic obstruction (with coarctation/arch interruption).

분류

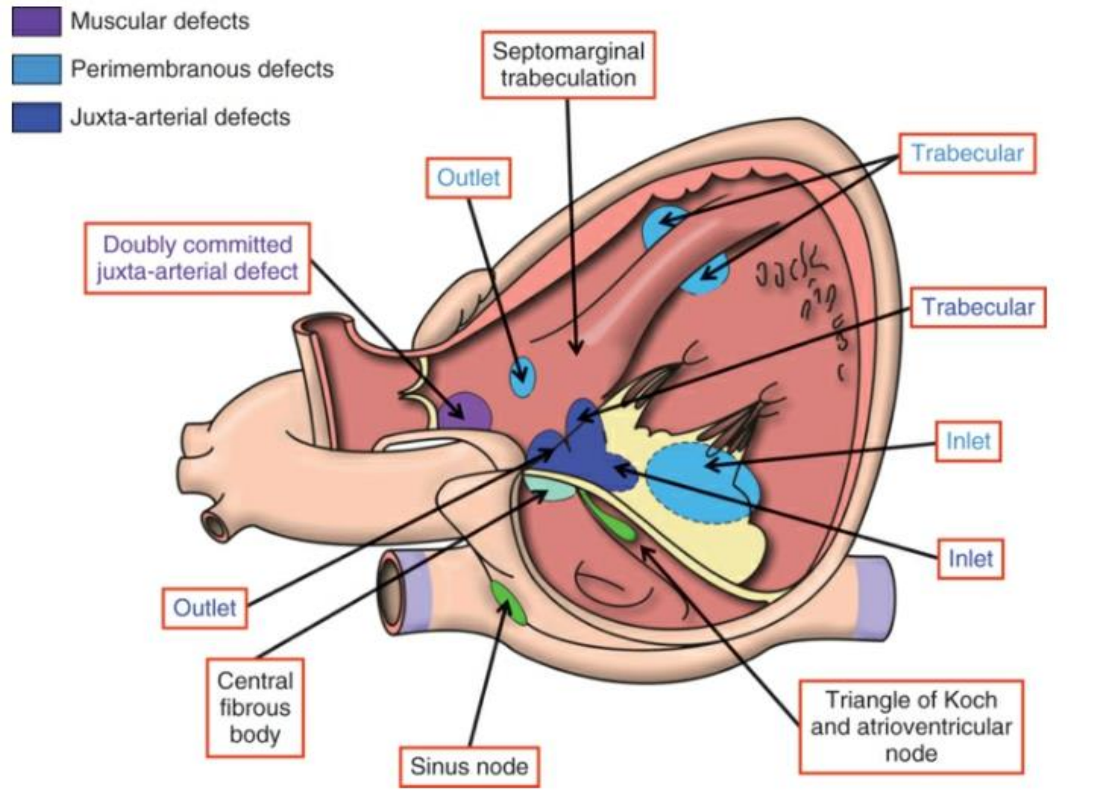

Table 35-1 Morphologic Classification of Ventricular Septal Defect

Classification	% of VSDs	Location/Borders
Perimembranous	80	Borders tricuspid valve Conduction system in posterior rim
Muscular	5	Borders all muscle Frequently multiple Conduction system remote
Doubly committed subarterial	5-10	Borders both semilunar valves Conduction system remote
Inlet septal	<5	Atrioventricular septal type Posterior position Conduction system in posterior rim

Key: *VSD*, Ventricular septal defect.

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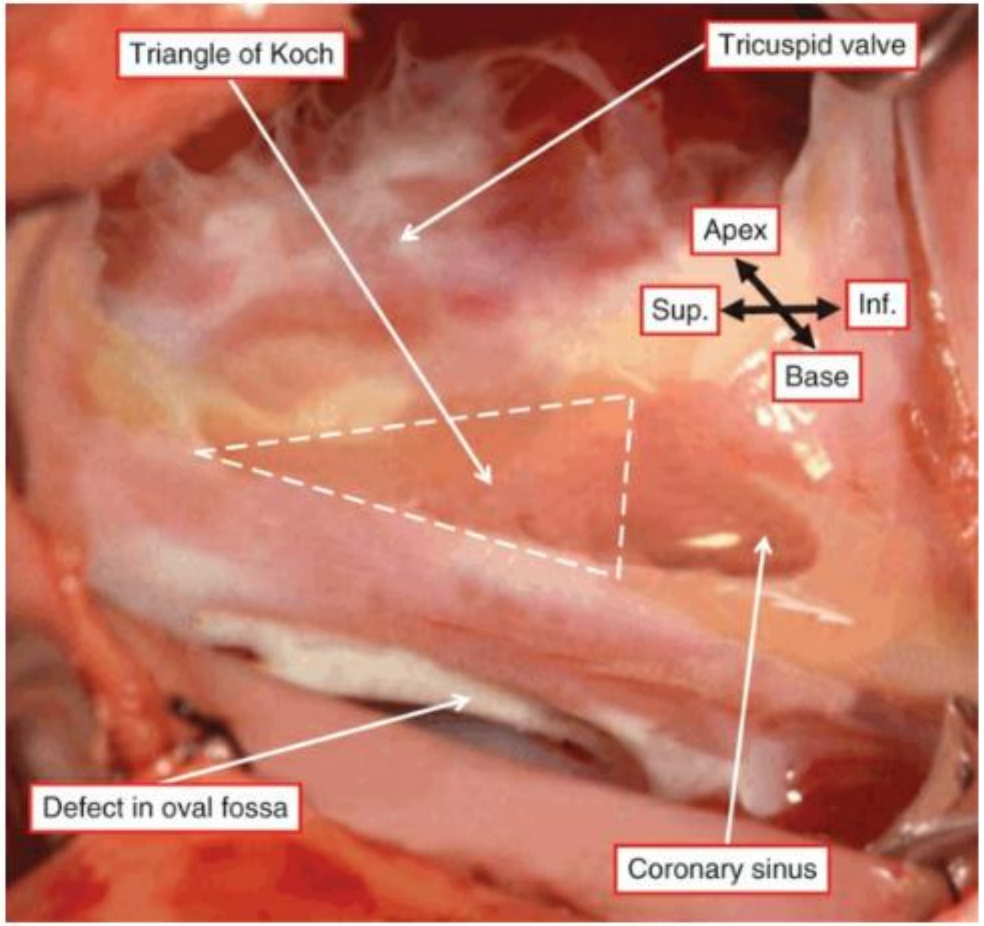
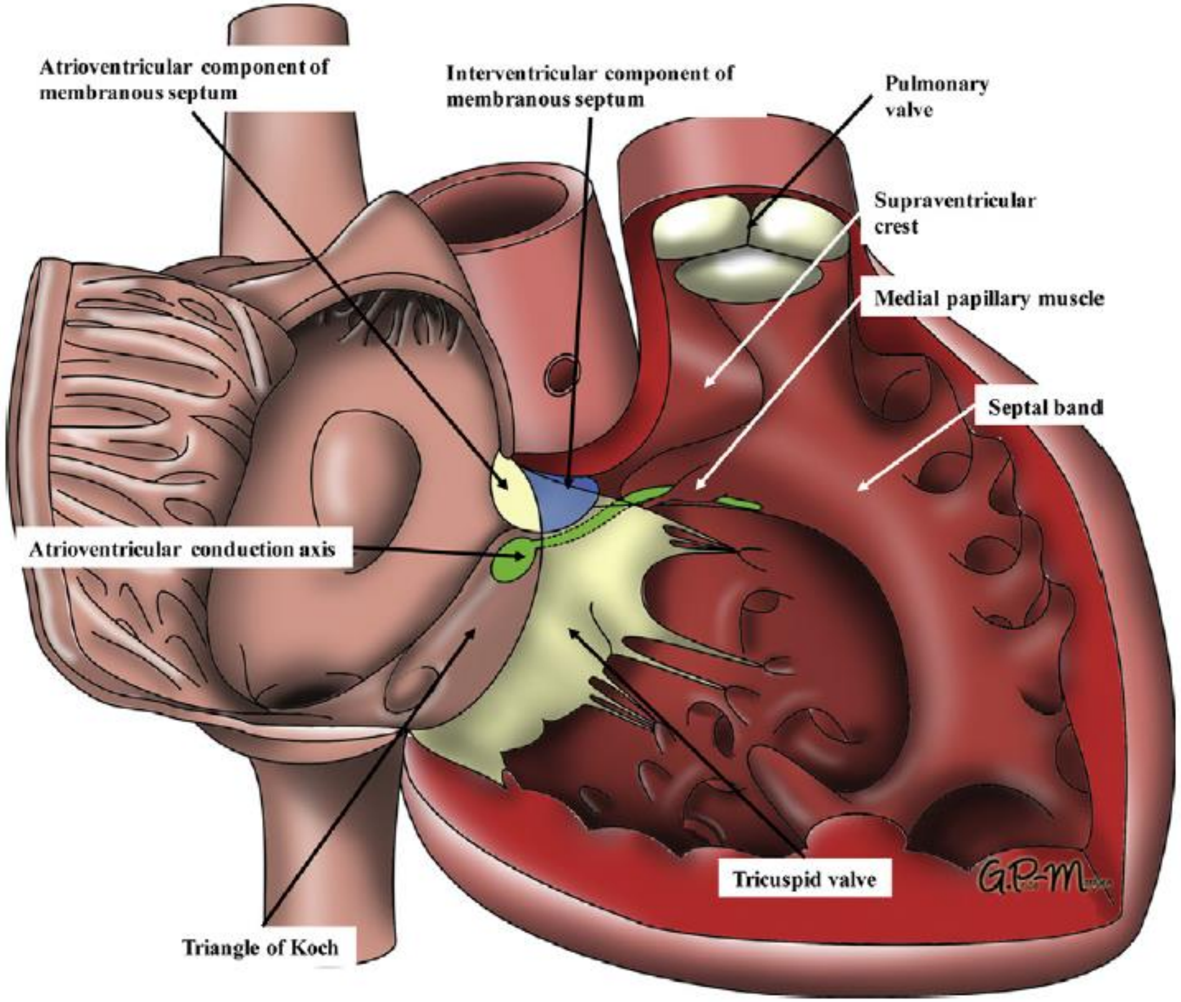
Conduction axis





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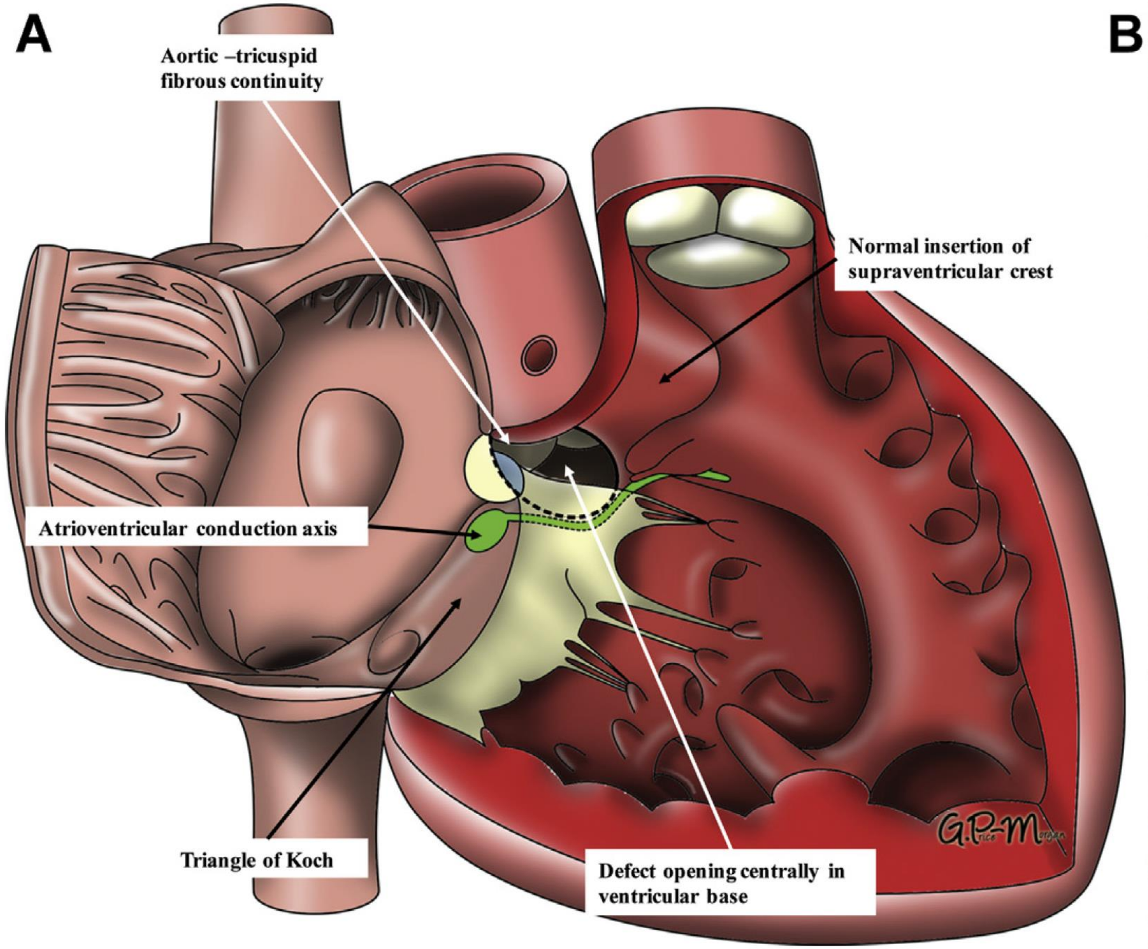
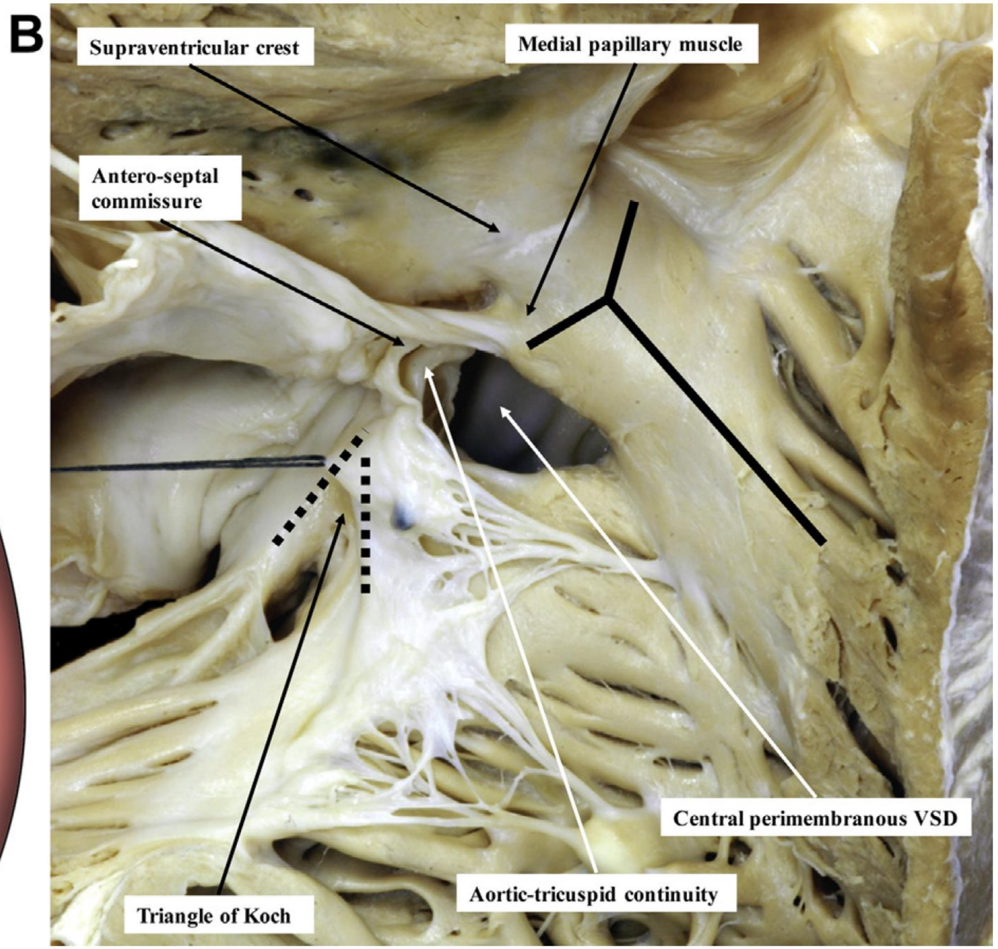
Central perimembranous defect





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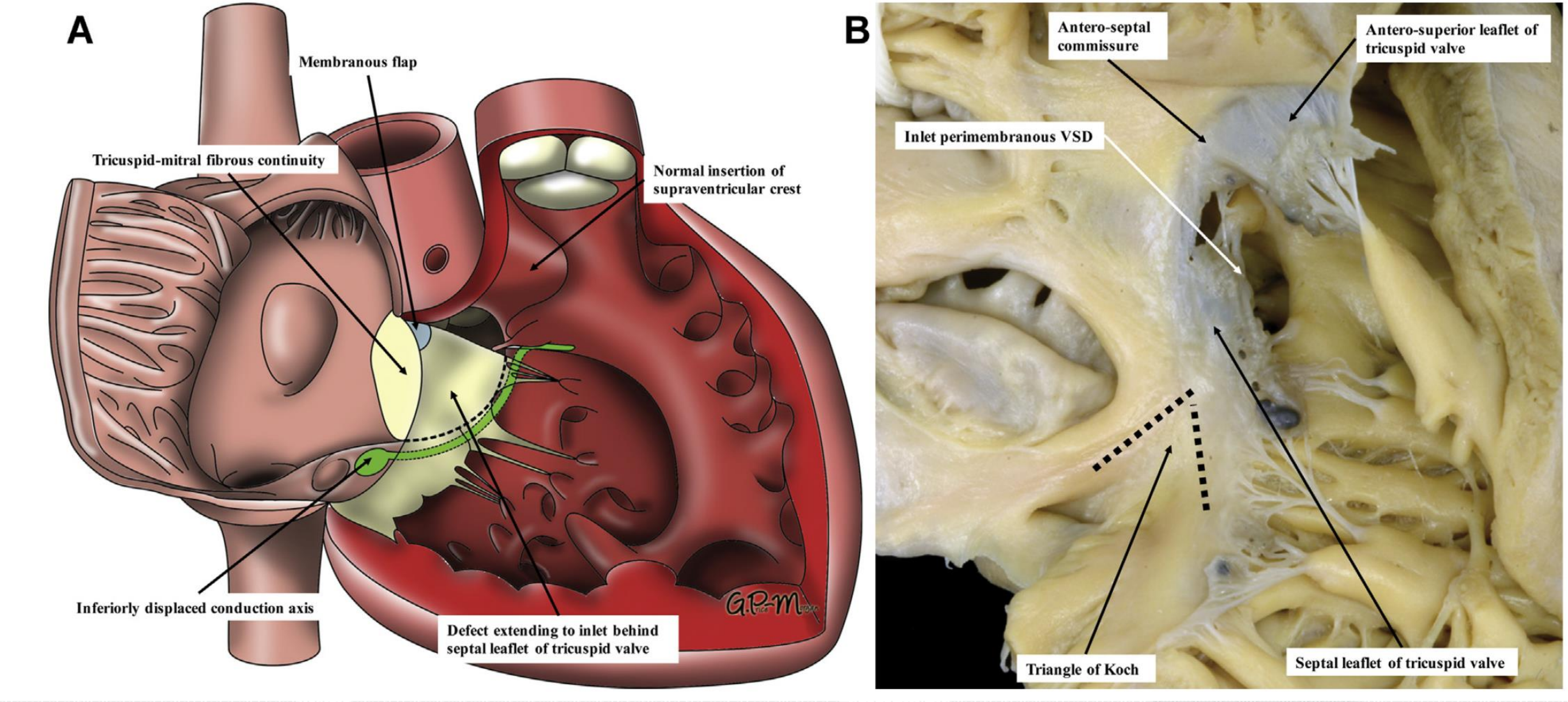
Inlet perimembranous VSD



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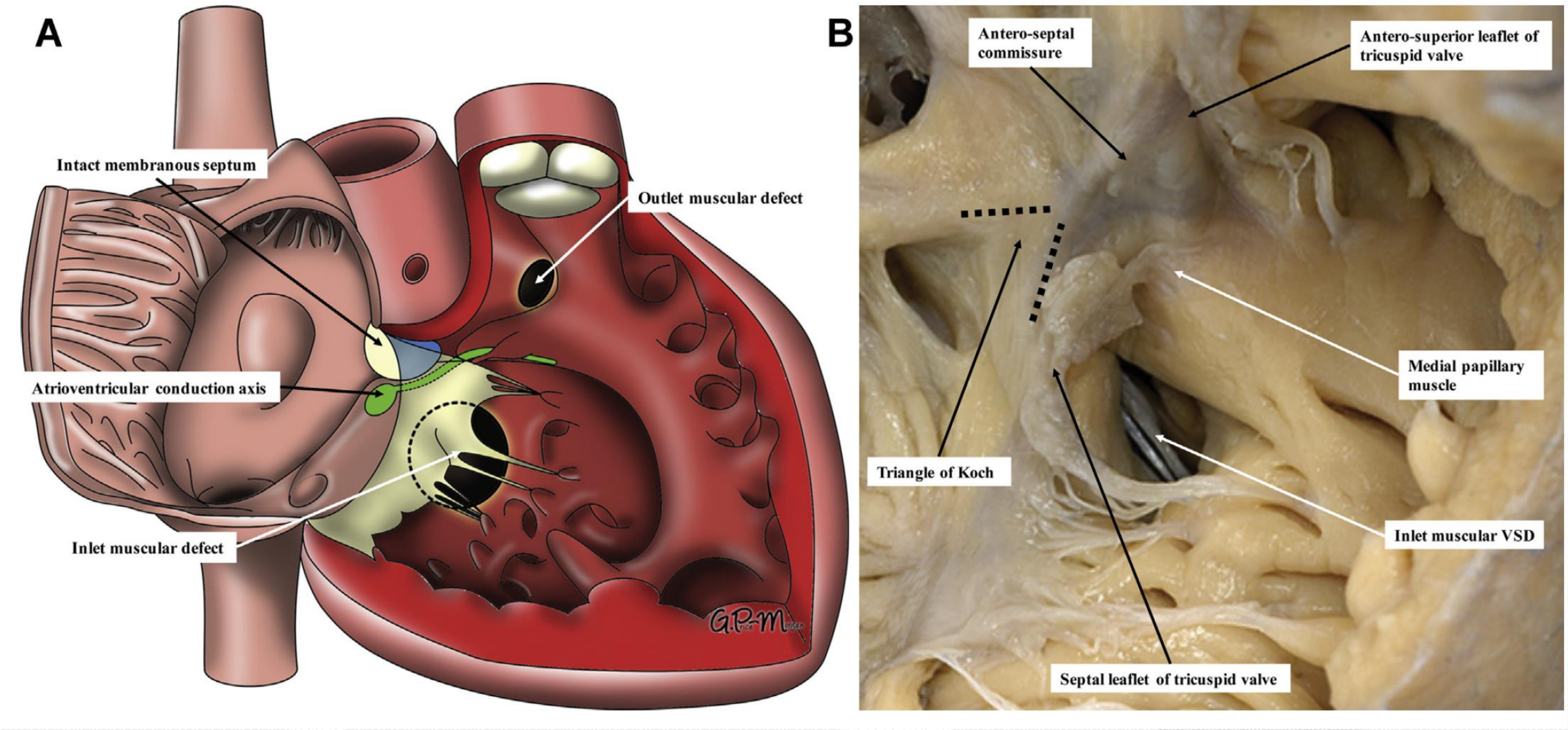
Inlet muscular defect



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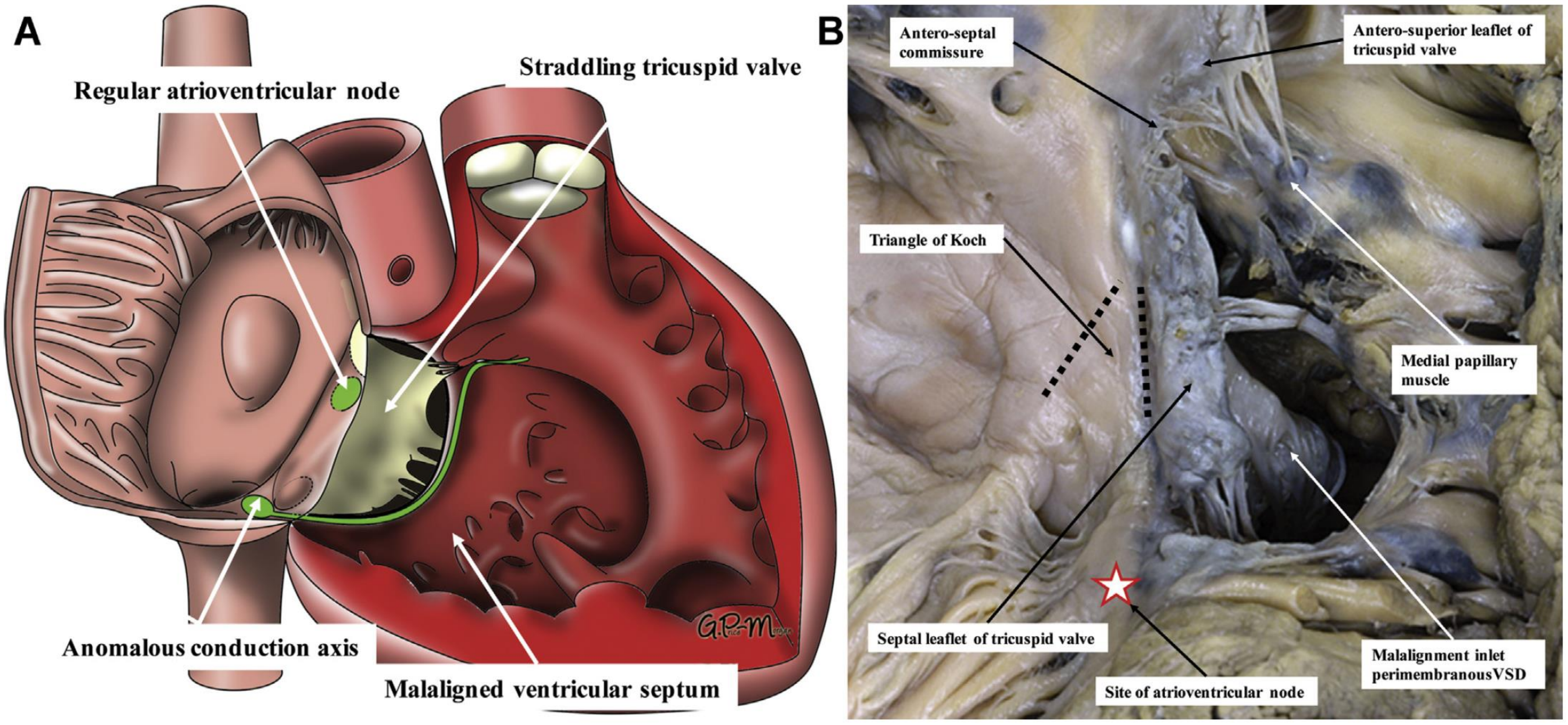
Inlet PM VSD with malalignment of septum



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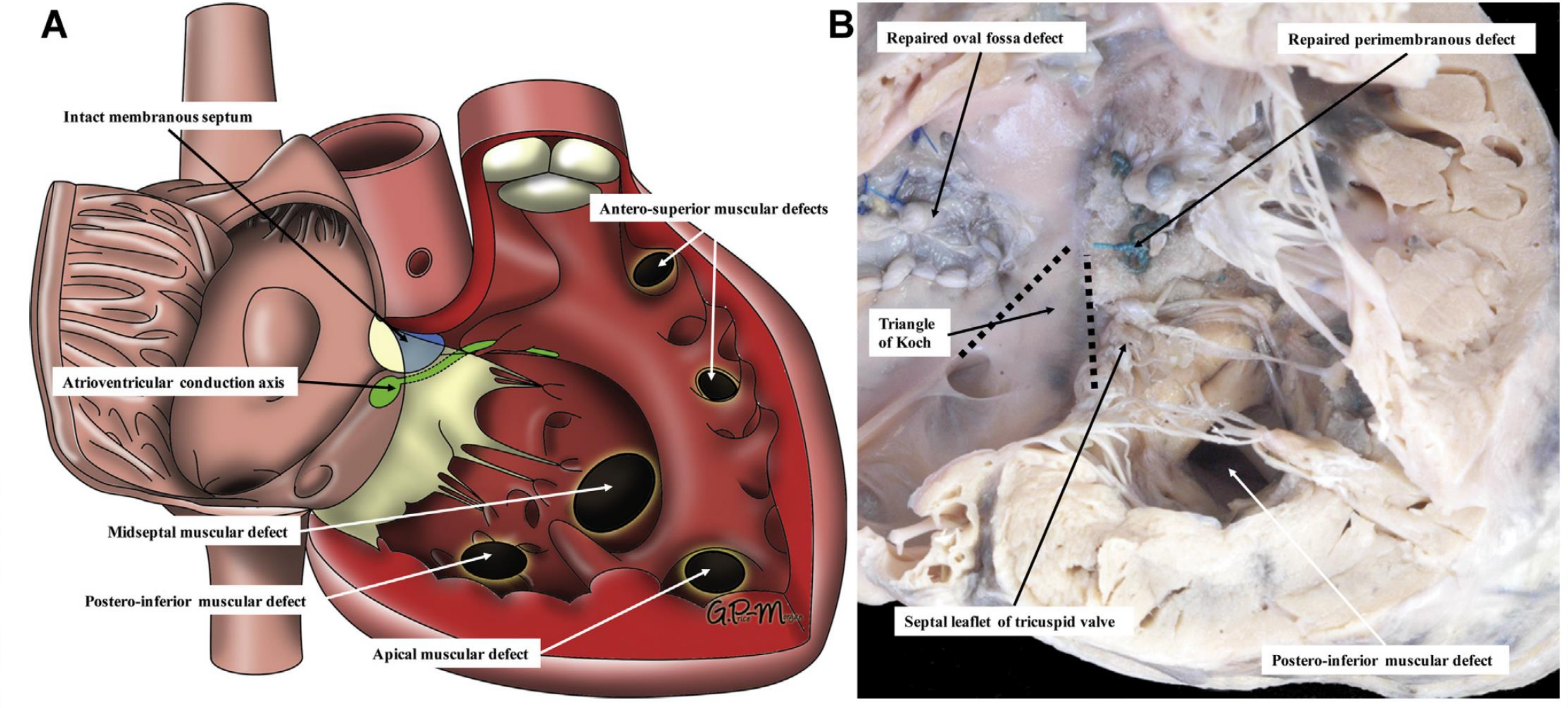
Trabecular muscular defect



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Pathophysiology

Determinants of Left-to-Right Shunt

- **Primary factors**: VSD size + pulmonary vascular resistance (PVR).
- **Other modifiers**: ventricular compliance, outflow obstructions (pulmonary/aortic).

Age-Dependent Physiology

- **At birth**: High PVR → minimal shunt; VSD often undetected.
- **Weeks–Months**: PVR falls → ↑ left-to-right shunt → audible murmur.
- **Excessive shunt**: CHF → dyspnea, poor feeding, recurrent infections, failure to thrive.

Pathophysiology

- **Progression of Pulmonary Vascular Disease**
  - Early: Pulmonary arteriolar thickening (first months).
  - 2–3 years: Irreversible pulmonary vascular obstructive disease.
  - Late: PVR > systemic resistance → shunt reversal → **cyanosis (Eisenmenger physiology)** → death if untreated.

Natural History

- **Spontaneous Closure**
  - Majority are **restrictive (<0.5 cm)**.
  - **80% close by 1 month**, mostly within 1st year; rare after 5 years.
  - Mechanisms:
    - Fibrosis of margins (hemodynamic).
    - Septal tricuspid leaflet adherence → pouch.
    - Muscular hypertrophy (common in muscular VSD).
  - Rare in adulthood (~10%).
  - Undesirable closure: **aortic valve prolapse → aortic regurgitation** (requires early surgery).

Natural History

- **Complications Over Time**
  - **Childhood**: CHF (dyspnea, FTT, recurrent infections).
  - **Adolescence/Adulthood**: Endocarditis, AR, arrhythmias, pulmonary hypertension, heart failure.
  - **Pulmonary vascular disease**:
    - Begins 1–2 years → irreversible damage.
    - Eisenmenger syndrome: reversal of shunt, cyanosis, RV failure (2nd–3rd decade; death by ~40 yrs).
  - **Subpulmonic stenosis**: From RV infundibular hypertrophy, mimicking TOF physiology.

Natural History

- **Endocarditis**
  - Incidence: ~0.3%/patient/year; higher in adults with unrepaired VSD.
  - Typical site: septal leaflet of tricuspid valve.
  - Signs: fever, bacteremia, recurrent infections.
  - Treatment: antibiotics → followed by surgical closure + tricuspid repair.
  - **Prophylactic closure** of restrictive VSDs remains controversial.

Natural History

- **Aortic Valve Prolapse & Regurgitation**
  - Common with outlet VSDs (juxtaarterial, muscular outlet, malaligned perimembranous).
  - Leads to ↓ shunt (leaflet prolapse) but ↑ aortic insufficiency.
  - **Early closure** advised if prolapse present, to prevent AI progression.
  - If AI already exists → repair at time of closure.

Diagnosis

- **Clinical Features**
  - **Auscultation**: Loud holosystolic murmur at LSB (louder with smaller defects).
  - Murmur weakens/shortens as PVR ↑; may disappear in severe pulmonary hypertension (loud P2).
  - Other findings: hyperactive precordium (rare), ventricular bulge, hepatomegaly, distended neck veins.

Diagnosis

Imaging & ECG

- **Chest X-ray**: Variable pulmonary vascularity, cardiomegaly, biventricular enlargement.
- **ECG**: May be normal, or show RVH, LVH, or biventricular hypertrophy.

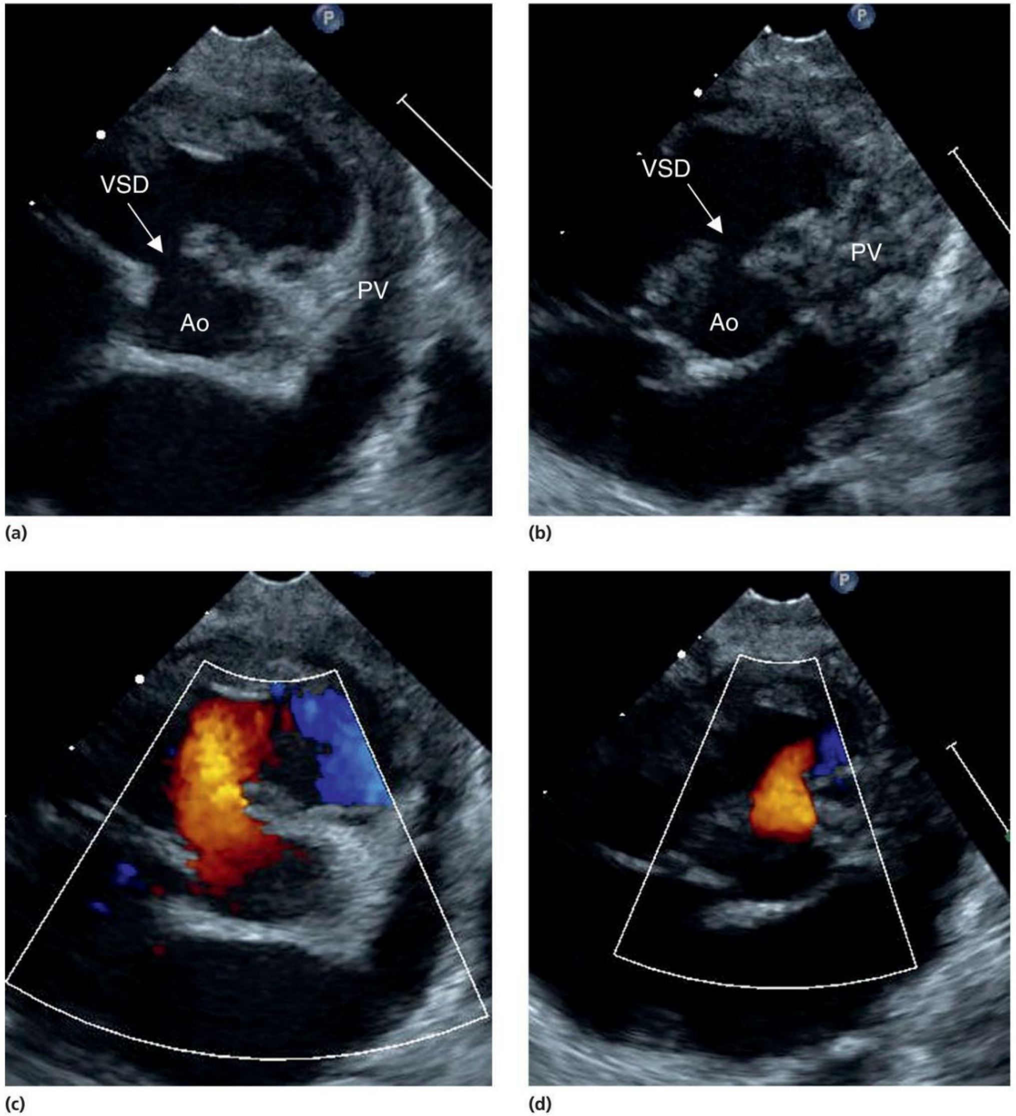
Echocardiography (Mainstay)

- Defines presence, size, location, borders.
- Assesses outflow tracts, aortic valve involvement, AV valve tension apparatus.
- Doppler → shunt direction and velocity.

Diagnosis

- **Cardiac Catheterization**
  - Now less frequently required.
  - Indicated when:
    - Older children/young adults with large VSD.
    - Need to quantify **Qp:Qs ratio**, measure **pulmonary arterial pressure & resistance**, and test **vasodilator response**.
  - Less used in infants since echo findings + symptoms are sufficient for surgical decision-making.

Echo : parasternal short-axis view



Lai, Wyman W.; Mertens, Luc L.; Cohen, Meryl S.; Geva, Tal. Echocardiography in Pediatric and Congenital Heart Disease: From Fetus to Adult (p. 255). Wiley. Kindle Edition.

Medical Management

- **Goals of Medical Management**
  - Control pathophysiologic effects of left-to-right shunt.
  - Manage increased pulmonary vascular resistance (PVR).
  - Prevent infective endocarditis (antibiotic prophylaxis)

Medical Management

- **Infants with CHF**
  - **Therapies**: Digitalis, diuretics, afterload reduction.
  - Nutritional support for poor feeding/FTT.
  - Antibiotics for recurrent pulmonary infections.
  - Supportive therapy may delay surgery or allow spontaneous closure.
  - Severe cases: assisted ventilation + inotropes as bridge to surgery.
  - Evaluate for secondary causes: subaortic stenosis, coarctation, PDA, infection.

Medical Management

- **Older Patients with Pulmonary Hypertension**
  - **Cardiac catheterization** essential:
    - Measure pulmonary arterial pressure.
    - Assess vasodilator response (O_2, amrinone, isoproterenol, nitroglycerin, nitroprusside, inhaled NO, PGE1).
  - **Favorable response** = ↑ L→R shunt and/or ↓ mean PA pressure → candidate for VSD closure.
  - Same agents useful postoperatively to reverse reversible pulmonary hypertension.

Patient Selection

- **Selection Criteria for VSD Closure**
  - **Defect characteristics**: Size & type (echo ± cath).
  - **Natural history**: Likelihood of spontaneous closure vs progression.
  - **Clinical status**: Presence and severity of symptoms.
  - **Complications**: Pulmonary hypertension, CHF, endocarditis, aortic regurgitation, associated anomalies.

Large VSD

- **Timing of Closure – Infants**
  - **Indication**: Severe, intractable CHF → surgery within first 3 months.
  - If medical therapy effective → observe until ~6 months.
  - After 6 months: spontaneous closure unlikely; risk of progressive pulmonary vascular disease increases.
  - Indications:
    - PVR > 4 Wood units/m^2.
    - Qp:Qs > 2:1.

Large VSD

- **Older Patients with Large VSD**
  - May show “clinical improvement” due to ↑ PVR and ↓ L→R shunt.
  - **Not surgical candidates**:
    - Dominant R→L shunt.
    - PVR > 8 Wood units/m^2.
    - No vasodilator response.
  - Lung biopsy: no longer recommended.
  - Some centers → staged therapy: preoperative sildenafil/bosentan → surgical closure.

Large VSD

- **Advanced PAH Management**
  - Targeted drugs ↓ PVR:
    - **Prostanoids** (epoprostenol).
    - **Endothelin receptor antagonists** (bosentan, ambrisentan).
    - **PDE-5 inhibitors** (sildenafil, tadalafil).
  - Used pre- & post-op → improve operability and outcomes.
  - Clinical studies: significant ↓ PA pressure, improved functional status, low operative mortality.

Large VSD

- **Surgical Innovations – Valved Patch Closure**
  - **Unidirectional or double-flap fenestrated patch** → allows decompression during pulmonary hypertensive crises.
  - Reported series:
    - Novick technique: survival 96% (primary VSD).
    - Rao: mortality 5.6%; crises manageable.
    - Talwar: no deaths; long-term survival excellent; no cyanosis.

Large VSD

- **Special Cases**
  - VSD + severe PAH in developing countries → high morbidity/mortality due to delayed diagnosis and limited resources.
  - Rare option: **VSD closure with lung transplantation** for advanced pulmonary vascular disease.

Small VSD

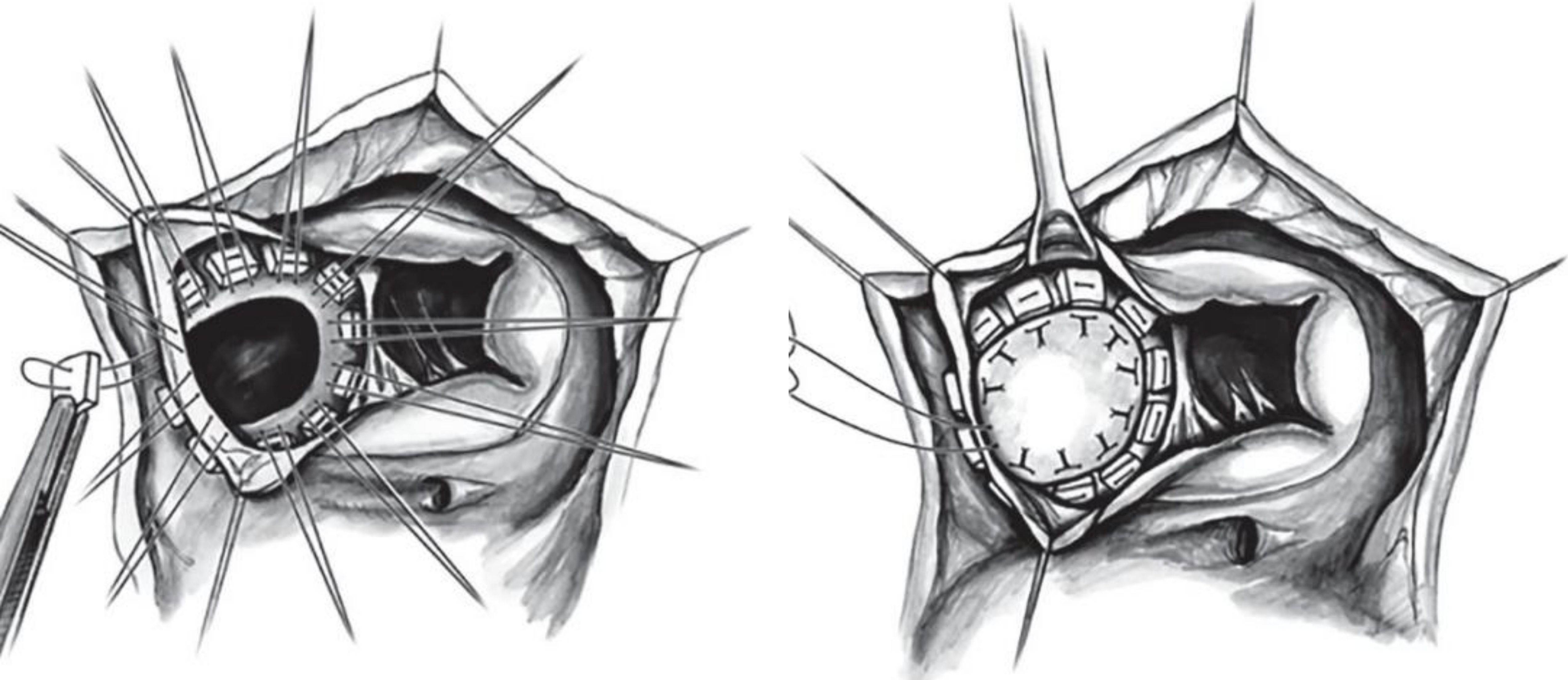
- **Management**: Usually no medical therapy; most shrink or close spontaneously.
- **Follow-up**:
  - After 1 year → periodic reassessment for physiologic/anatomic changes.
- **More liberal indications for closure** (even if Qp:Qs < 2:1):
  - Aortic valve prolapse (with or without regurgitation).
  - Prior infective endocarditis.
  - Ventricular dilation.
- **Evidence**: Large series (Backer et al., 1993) → safe closure, no operative deaths or major complications.
- **Rationale**: Surgical risk < lifetime risk of endocarditis, AR progression, TR.

Doubly Committed & Juxtaarterial VSDs

- Defects beneath pulmonary valve.
- High risk of **aortic valve prolapse & regurgitation**, especially >5 years of age.
- **Recommendation**: Early closure in all cases, regardless of shunt size.

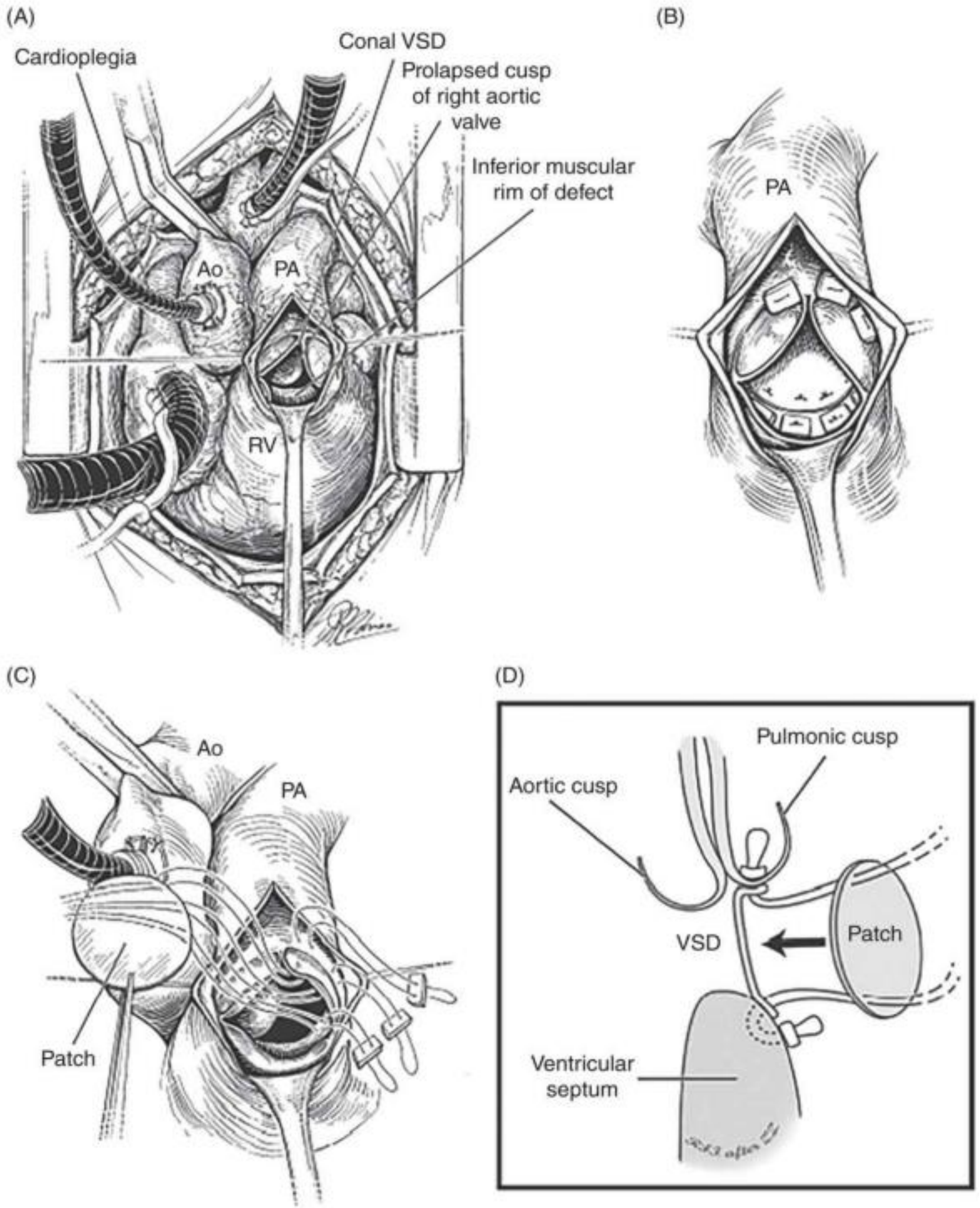
Surgical Considerations

- **Preoperative imaging**: Defines defect location, exposure options, conduction pathway risks, and closure technique (especially when valve leaflets form part of defect border).
- **Operative approaches**:
  - Right atrial
  - Transpulmonary
  - Transaortic
  - Right ventricular
  - Left ventricular
- **Complex / multiple VSDs**: May require **combined approach**.



Transatrial Closure of Perimembranous or Central Ventricular Septal Defect:

Operative view of the doubly committed and juxtaarterial (conal) ventricular septal defect (VSD)



Results

Scully et al. (2000–2006, n=215)

- Median age: 10 months; 50% had CHF/FTT pre-op.
- Mortality: 0.5% operative, 0.9% late.
- No heart block, no reoperation for residual VSD.
- 26% developed RBBB; no valve-related complications.

Hoffman et al. (1968–1980, n=176)

- Survival: 92% alive >20 years, NYHA I.
- Mortality: 4% late deaths (mostly RV hypertrophy/arrhythmia).
- Reoperation in 10% (few for residual VSD).
- Pacemaker: 6 patients (early postop or >15 yrs later).

Results

- **Natural History of Small, Unoperated VSDs (Soufflet, n=220; median f/u 6 yrs)**
  - Mortality: 1% (sudden death, end-stage HF).
  - Endocarditis: 4%.
  - Surgical closure required: 7.5%.
  - Spontaneous closure: 4%.
  - Increased risk of **PAH** with persistent open perimembranous VSD.

Results

- **Conduction & Ventricular Function**
  - **RBBB common** after repair (up to ~50%).
  - **Pederson study**:
    - Post-op systolic long axis function reduced in all patients.
    - Diastolic dysfunction more prominent in those with RBBB.
  - **Karadeniz study**:
    - RBBB associated with ↓ RV fractional area change → RV dysfunction risk.

Complications

- **Anatomic risk structures**: conduction system, tricuspid & aortic valves.
- **Arrhythmias**: transient common; RBBB frequent (esp. transventricular).
- **Heart block**: permanent CHB ~1–2% (pacemaker needed).
- **Valve injury**: aortic or tricuspid regurgitation (suture/leaflet injury).
- **Residual shunt**: <5%; reoperation if Qp:Qs > 1.5:1.
- **Bypass-related risks**: neurologic injury with deep hypothermia/circulatory arrest → continuous CPB preferred.

Pulmonary Artery Bands

- **Historical use**:
  - Protect lungs from unrestricted flow.
  - Delay corrective surgery until older age.
  - Indications: Swiss-cheese VSDs, complex anatomy (uncertain repair strategy), univentricular physiology with excessive pulmonary flow.

- **Limitations/Complications**:
  - Significant morbidity/mortality.
  - Band migration → branch PA distortion.
  - Band erosion → PA lumen injury.
  - Pulmonary valve distortion.
  - Subannular ventricular hypertrophy.

- **Current role**:
  - Transient stage before definitive closure in small infants.
  - Palliative procedure for multiple VSDs.
  - **Primary repair preferred** when feasible.

Transcatheter/ Transventricular Device Closure

- **Muscular VSDs**:
  - Advantage: Good visualization of apical/anterior defects.
  - Avoids CPB & cross-clamp.
  - Rare risk of heart block.
  - Risks: device embolization, hemolysis, chordal/papillary muscle injury.
  - Useful when multiple or hard-to-access defects.
- **Perimembranous VSDs**:
  - **Major issue**: High risk of complete heart block (5.7–22%).
  - Late-onset CHB possible → pacemaker dependence, risk of cardiomyopathy & shortened lifespan.
  - Other complications: AI, TR, hemolysis, embolization, endocarditis.
  - Device pressure on LV margin = direct risk to His bundle.
  - Considered **unacceptable risk** by most clinicians today.

Transcatheter/ Transventricular Device Closure

Surgical Closure

- Avoids femoral vessel cannulation.
- Allows correction of additional lesions (ASD, coarctation, multiple VSDs).
- Operative mortality now near zero in uncomplicated cases.
- CHB after surgery: ~1–2% (lower than device closure).

Current Limitations of Device Closure

- Young children/infants (<8–10 kg) with CHF, FTT, PAH are **poor candidates**.
- Long-term safety of large metallic devices in the heart remains uncertain.

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Thank You

For your attention