VENTRICULAR SEPTAL DEFECTS

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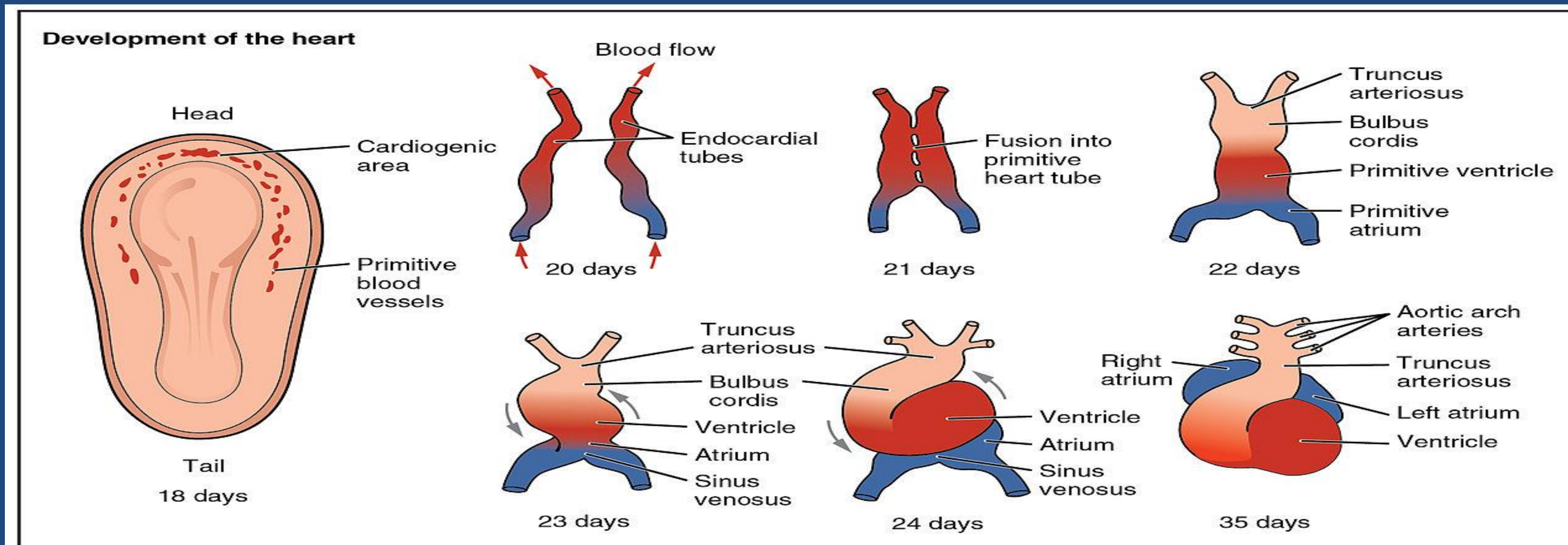
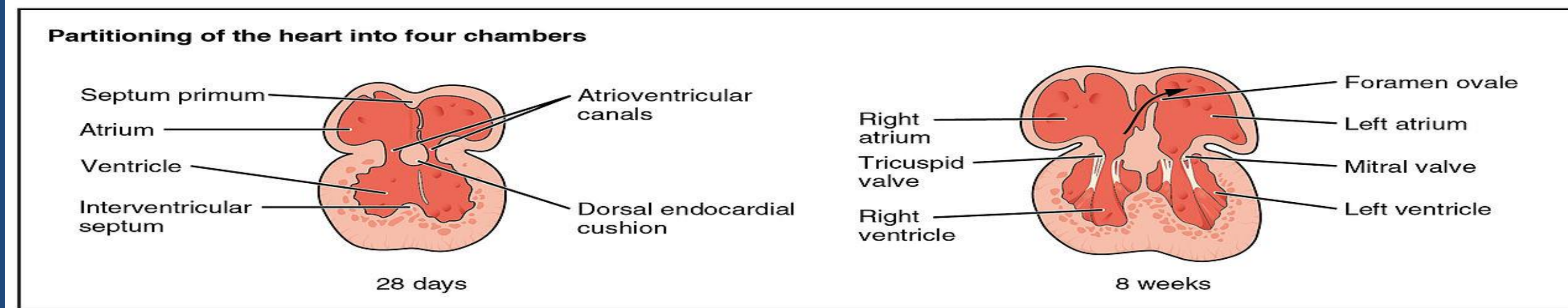
children's health | UT Southwestern Pediatric Group

LEARNING OBJECTIVES

- Review normal cardiac anatomy
- Discuss different types and location of VSD
- Understanding of SVR and PVR
- Define diagnostic tools for diagnosing VSD
- Discuss preoperative management for VSD patients
- Review surgical repair of VSD
- Discuss postoperative care and follow up
- Discuss and understand the etiology of Eisenmenger Syndrome

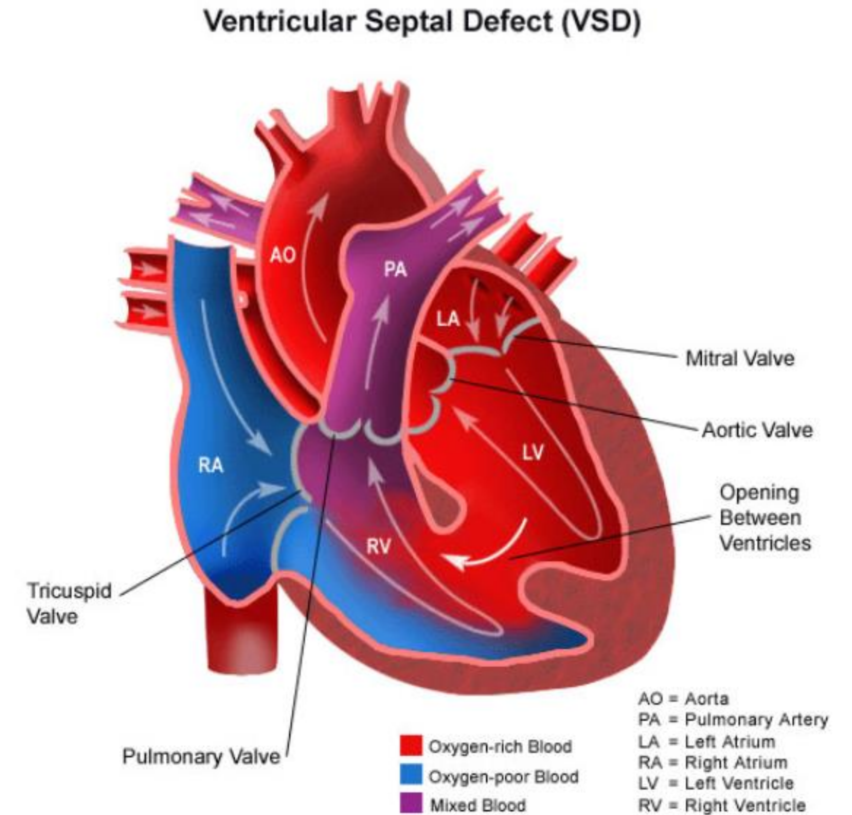
EMBRYOLOGY OF HUMAN HEART (QUICK REVIEW)





VENTRICULAR SEPTAL DEFECT

- Result of abnormal development or disruption in formation of ventricular septal wall during embryology
- Simply a communication between the left and right ventricles of the heart
- Most common congenital heart disease
- As pulmonary vascular resistance decreases over the first few weeks of life a left to right shunt develops across the VSD causing increased pulmonary blood flow

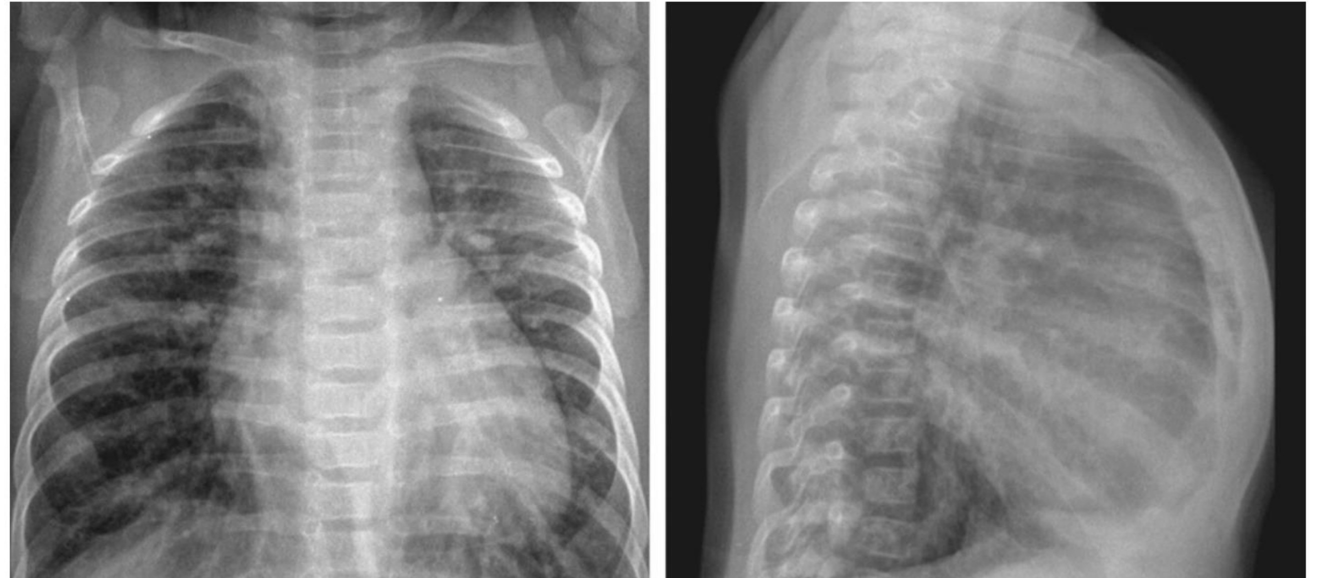


CASE STUDY ONE

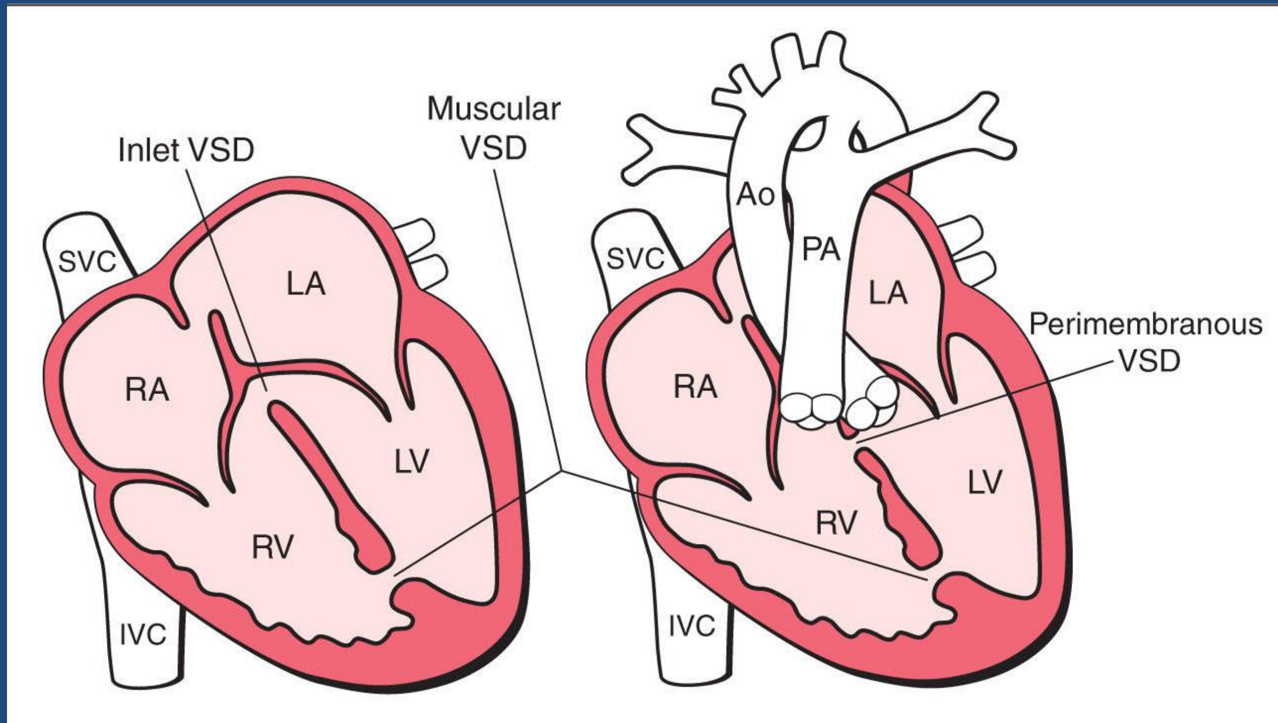
- 5-month-old presented to the clinic with persistent FTT
- Past medical history
  - Two hospital admissions for pneumonia requiring oxygen
- Physical exam
  - Small thin male tachypneic with respiratory rate in the 60s, mild subcostal retractions, clear breath sounds throughout
  - No murmur, CR <2 seconds , Pulses +2 throughout, Liver palpable 2 cm below the right costal margin

EVALUATION: WHAT DO YOU WANT DO NEXT?

- Chest X-ray
  - Cardiomegaly
  - Increased pulmonary vascular marking
- ECG
  - Biventricular hypertrophy and LAH
- ECHO
  - Position and size of defect
  - Gradient across the defect
  - LA and LV dilation
  - Mitral regurgitation



ANATOMY OF VSD



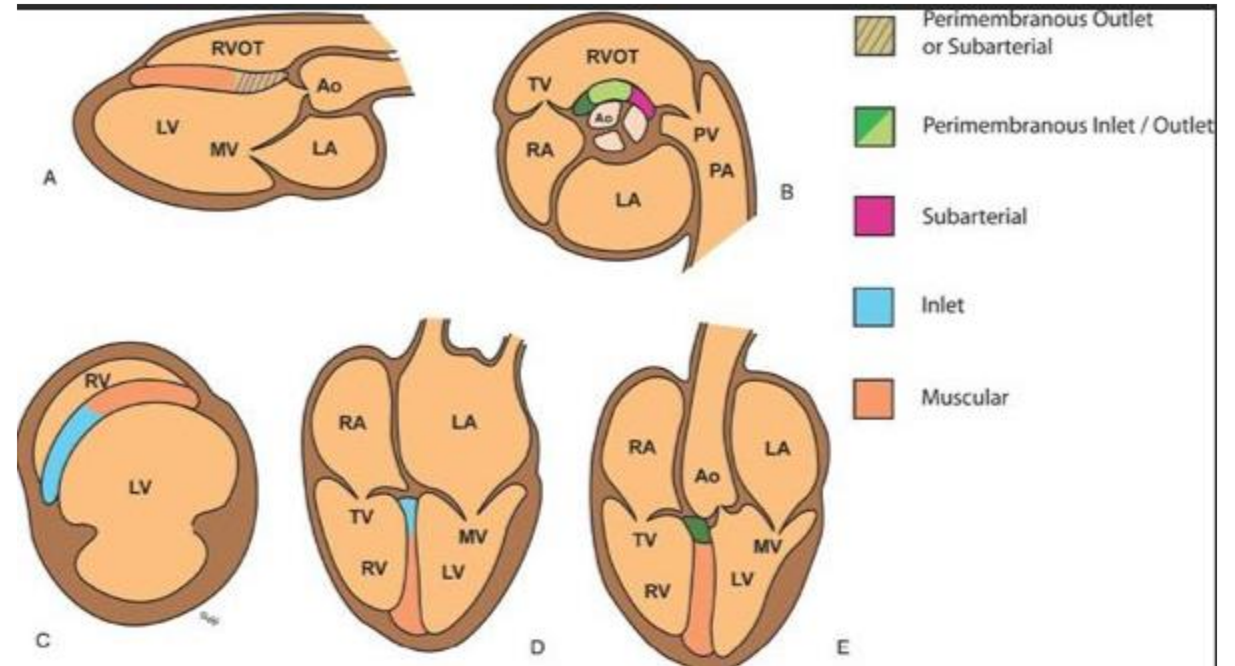
- Inlet
  - Directly beneath the tricuspid and mitral valve
- Outlet
  - Areas that gives rise the pulmonary artery and aorta
- Perimembranous
  - Directly adjacent to the membranous septum of the tricuspid valve
- Muscular
  - Anywhere on the septum between the right and left ventricle

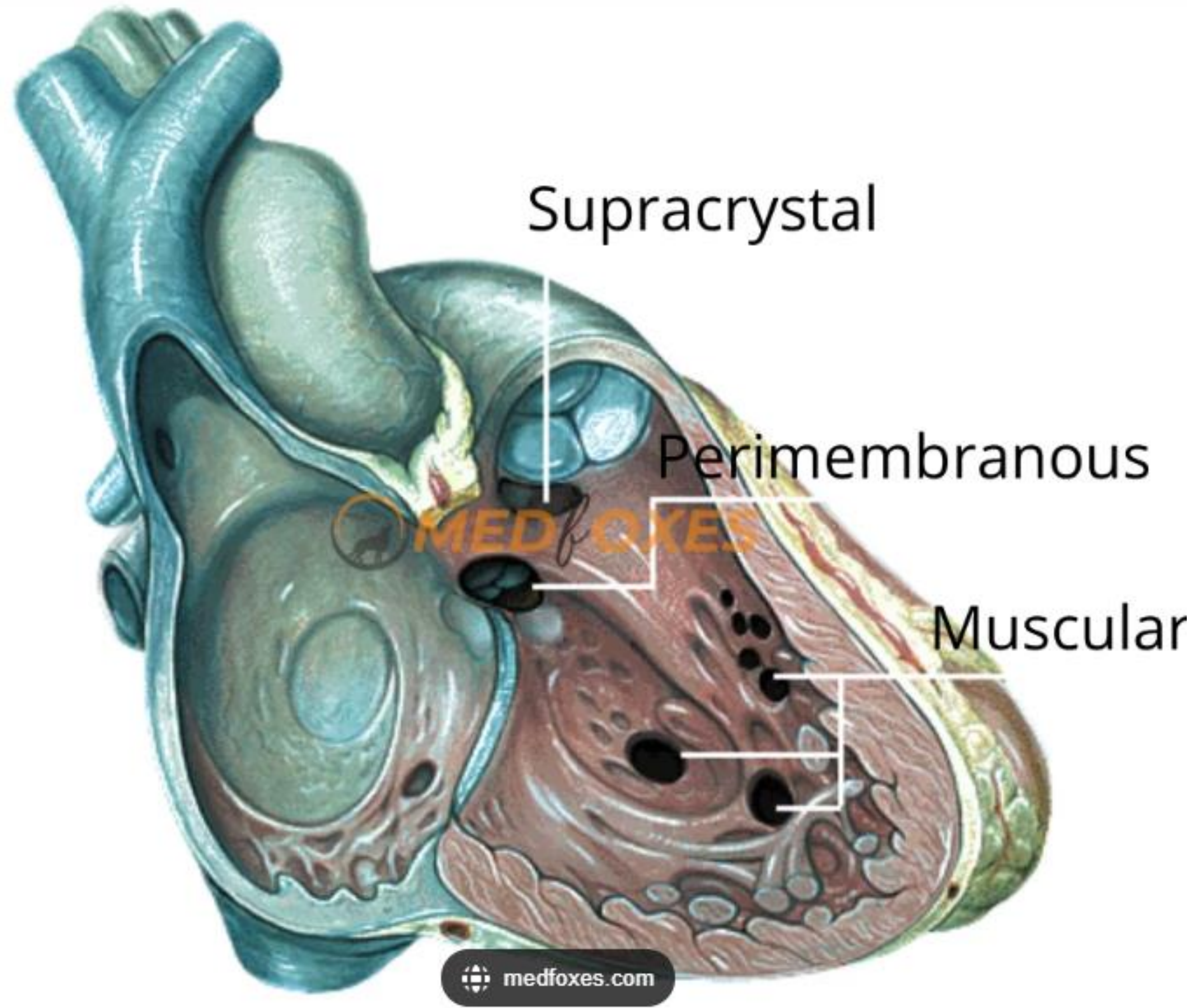
INLET VSD

Located interior to the tricuspid and mitral valve within the inlet portion of the RV septum

Associated with transitional or intermediate AV septal defects

Increased frequency with Trisomy 21 patients





OUTLET VSD

- Positioned beneath the semilunar valves without the outlet septum of the RV
- Sometimes referred as supracristal VSD
- Associated with prolapse of the aortic valve and AI

PERIMEMBRANOUS VSD

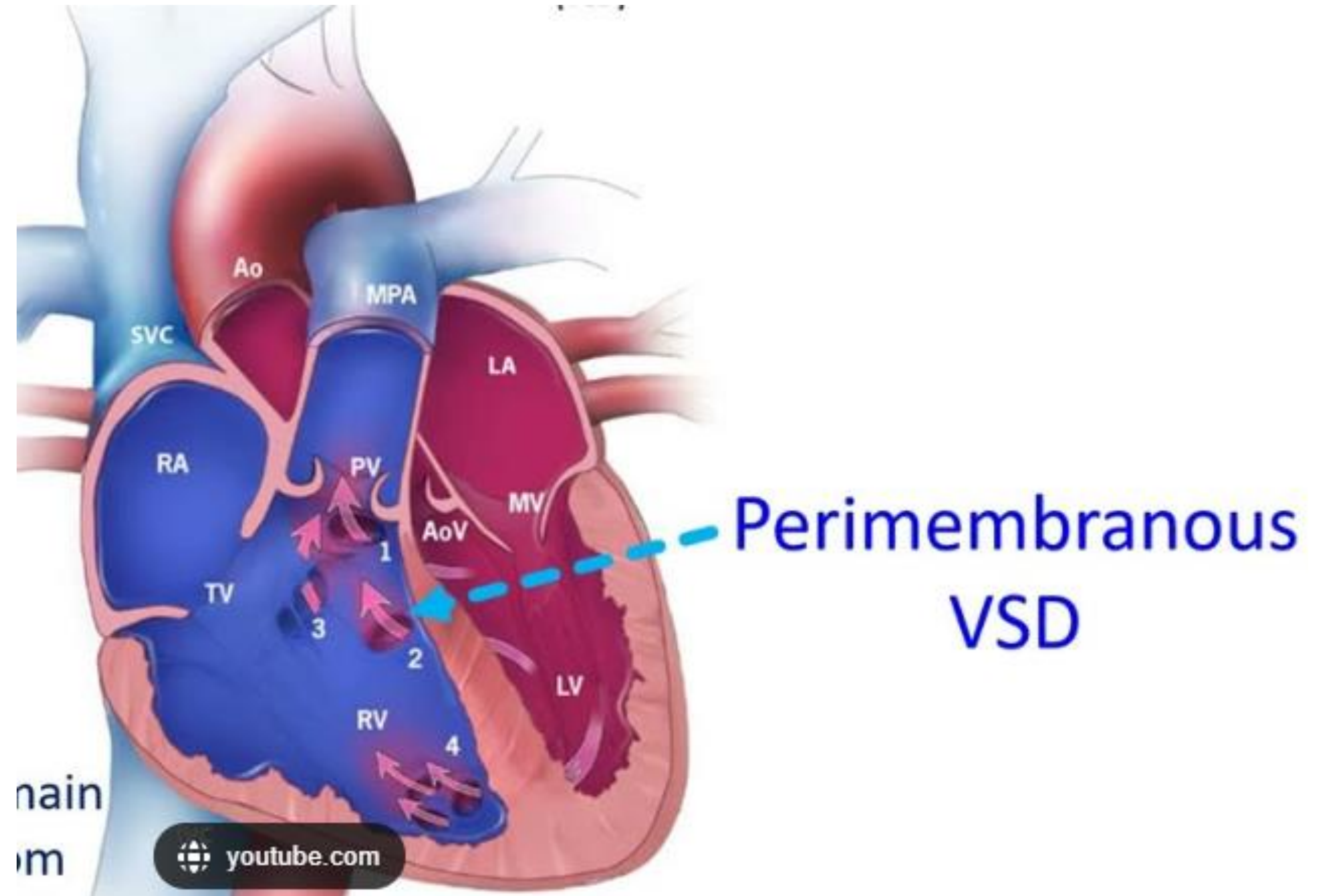
Located in membranous septum below the crista supraventricular

Most common type

Can be restrictive or closed by tricuspid valve tissues

Associated with prolapse of aortic valve cusp and AI

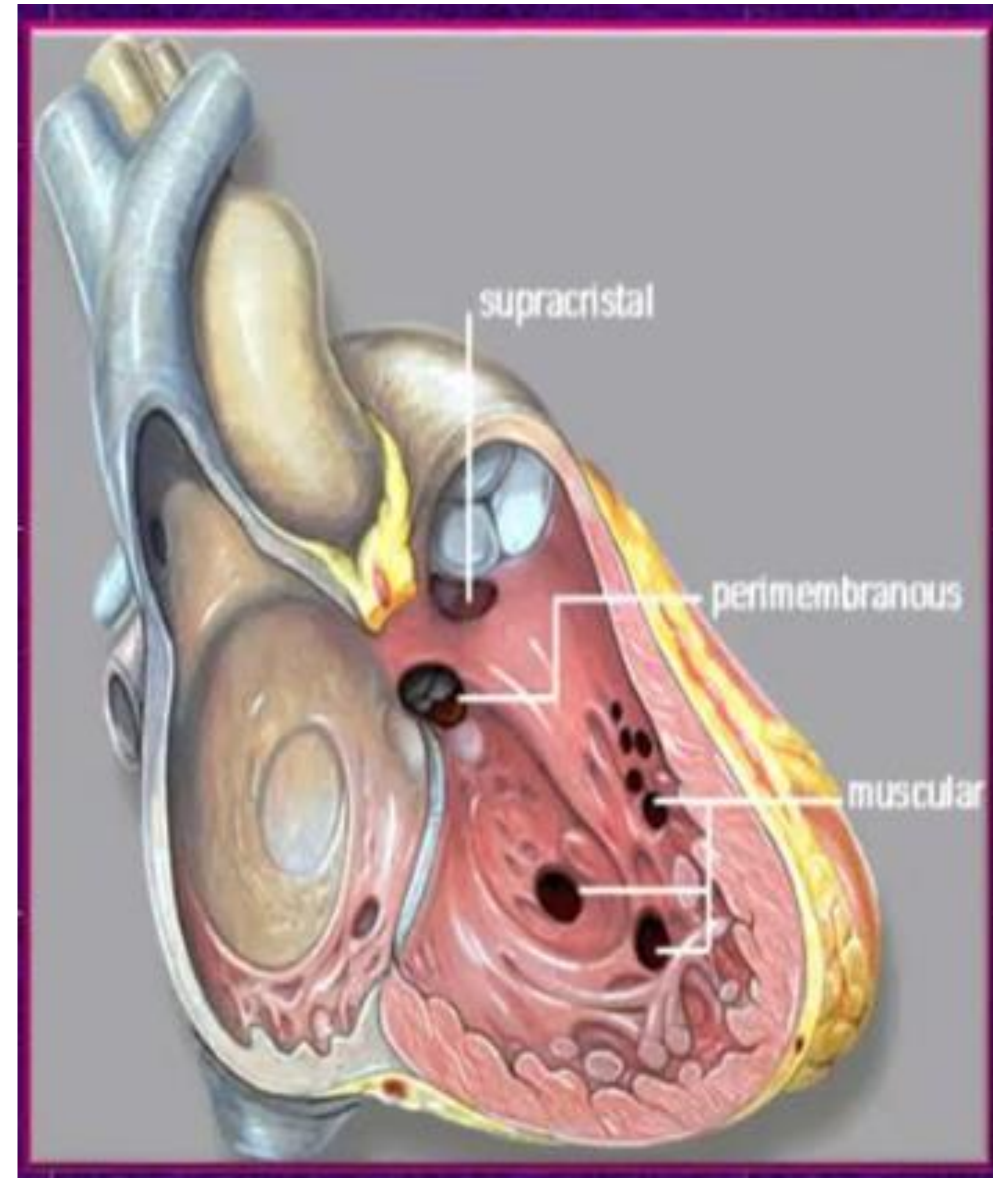
Surgical approach through the TV



MUSCULAR VSD

Located in the apical, central, anterior or outlet region of the muscular septum

More likely to close without intervention



PATHOPHYSIOLOGY

- Depends on the size of the defect
  - Restrictive or non-restrictive
- Relationship between SVR and PVR
  - PVR high at birth and then falls starting at 2 weeks
  - Low PVR leads to increased pulmonary blood flow
  - Decreasing of PVR can be delayed due to higher altitude (Zambia is 1200 meter/3900 feet)
  - If the VSD is large, there is little pressure difference

CLINICAL PRESENTATION OF VSD

Elevated PVR

- R → L shunting
- No murmur
- No symptoms of heart failure

Decreasing PVR

- Increased L→ R shunting
- Increased pulmonary blood flow
- Tachypnea
- Poor feeding
- Decreased exercise tolerance

PHYSIOLOGY OF A LARGE VSD

- Nonrestrictive to pressure
  - RV and LV pressures equalize
  - Flow depends upon pulmonary vascular resistance and systemic vascular resistance
  - Pulmonary vascular bed exposed to systemic BP and ↑ blood flow. Over time the pulmonary vascular endothelium undergoes changes that if prolonged can become irreversible. Prolonged exposure and associated changes are referred to as Eisenmenger's syndrome.
  - LA/LV volume loaded, high LVEDP → poor function, CHF, pulmonary edema

PREOP MEDICAL MANAGEMENT

- Focus on management of congestive heart failure
  - FTT management (Infant):
    - If breast fed, add olive or coconut oil 2-3 times a day (like a medication). Feed every 3 hours including overnight.
    - If formula fed or pumping , increase calories by adding extra formula (can to 22kcal, 24kcal, 27kcal, & 30kcal)
  - FTT management (Older child):
    - Try to eat 1-2 snacks a day
    - Try to eats more red meats and eggs
    - Increase seed, nuts, legumes, pasta, rice, or carbohydrate intake
    - Use cheese or peanut butter when available
    - Cook using real butter or oil
  - Medical management
    - Furosemide
    - Digoxin
    - Spironolactone

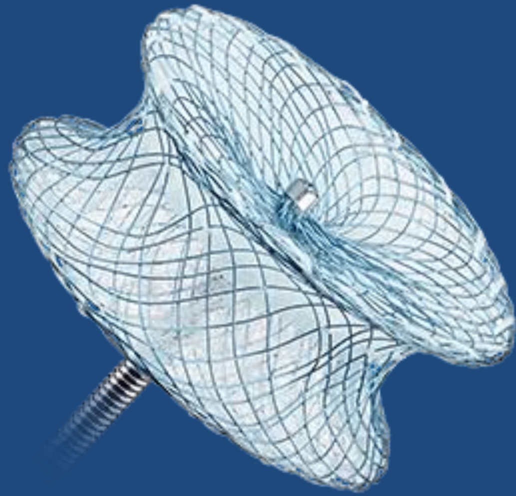
PREOP MEDICAL MANAGEMENT

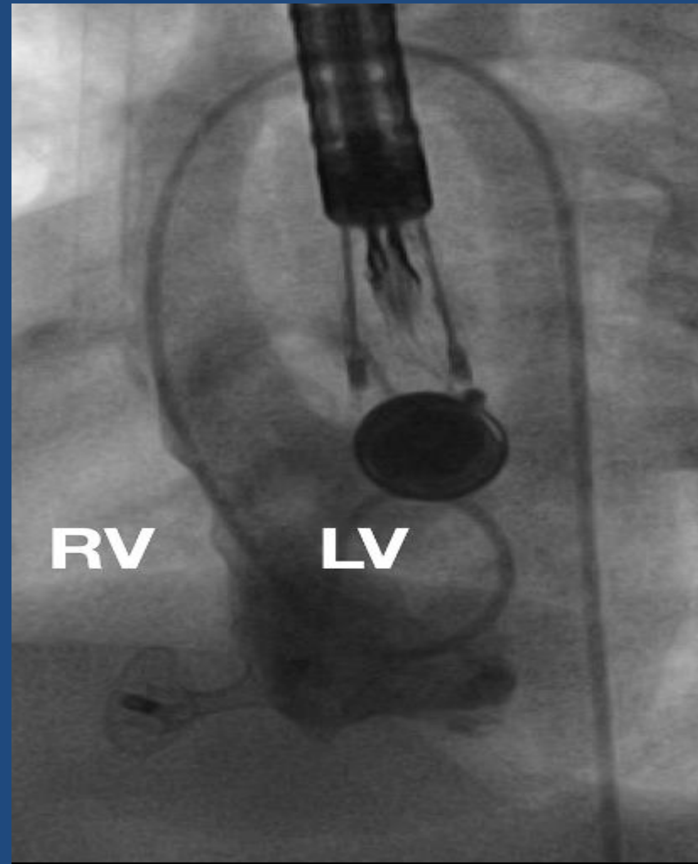
- If child is older (over 3-4 years), may need preop cath to assess PVR.
  - If PVR high, cath will assess if PVR is reversible (ie oxygen). If PVR responsive to oxygen, consider sildenafil or bosentan for vasodilation.
  - If not reversible, may not be surgical candidate.
- Preoperative Testing includes:
  - Chest Xray
  - Type and crossmatch
  - Electrolytes, BUN, and creatinine
  - CBC with diff
  - Urinalysis
  - EKG (within 45 days)
  - Obtain Preop H&P for surgical clearance
  - Preoperative parental teaching (defect, surgical plan, postop plan, & postop course)
  - Child life preparation for older patients

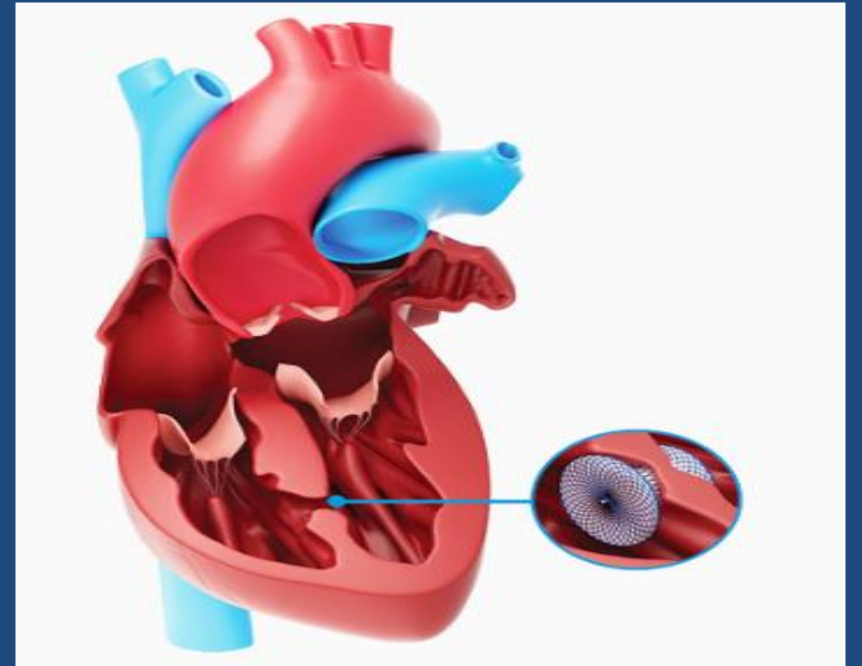
CANDIDATES FOR TRANSCATHETER VSD DEVICE CLOSURE

- Minimum weight of 5 kg
- Adequate space between the defect and the atrioventricular valves
- Complications
  - vascular injury
  - infection
  - conduction disturbances (heart block)
  - residual shunt
  - hemolysis
  - device embolization

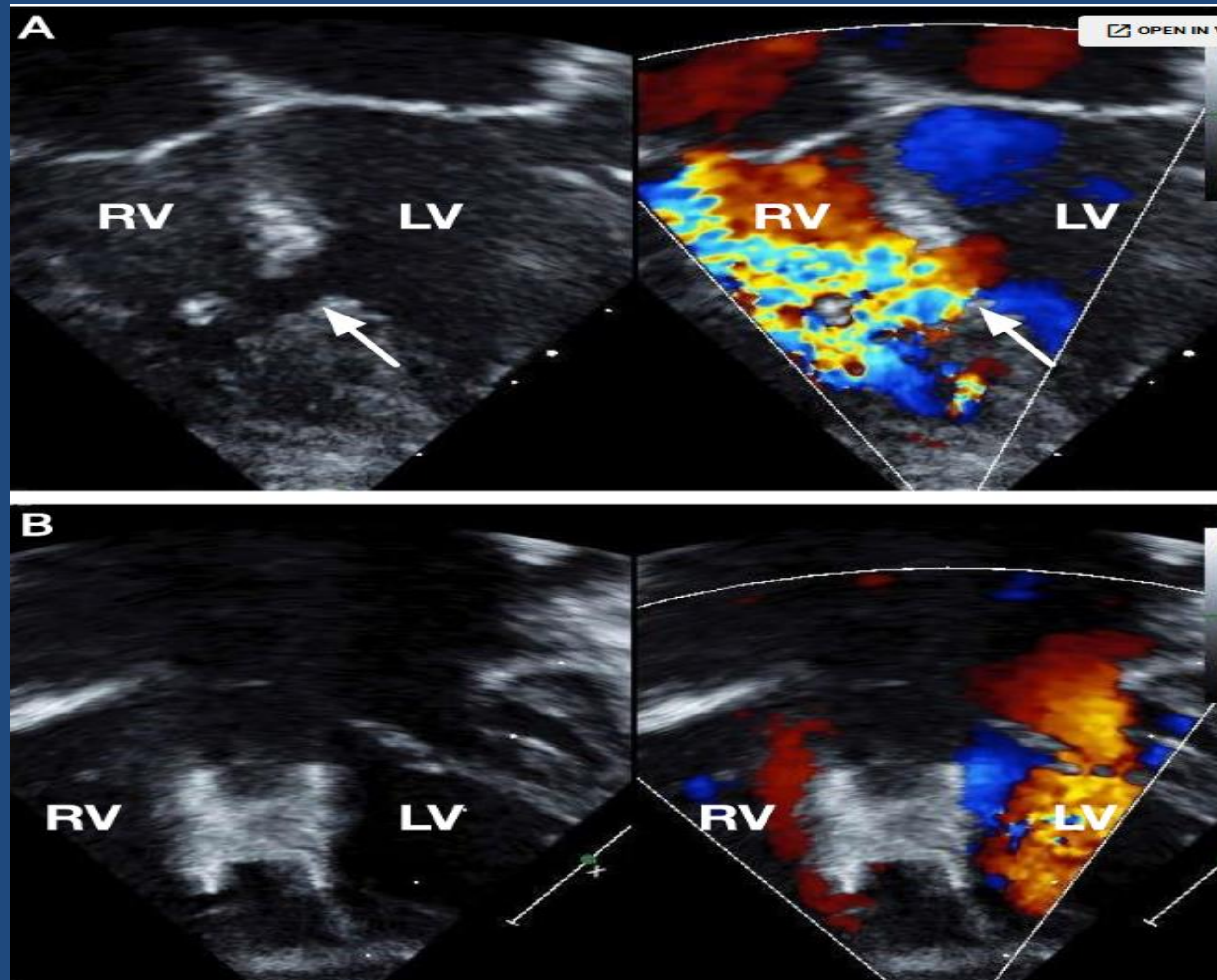
VSD CATHETER DEVICE CLOSURE





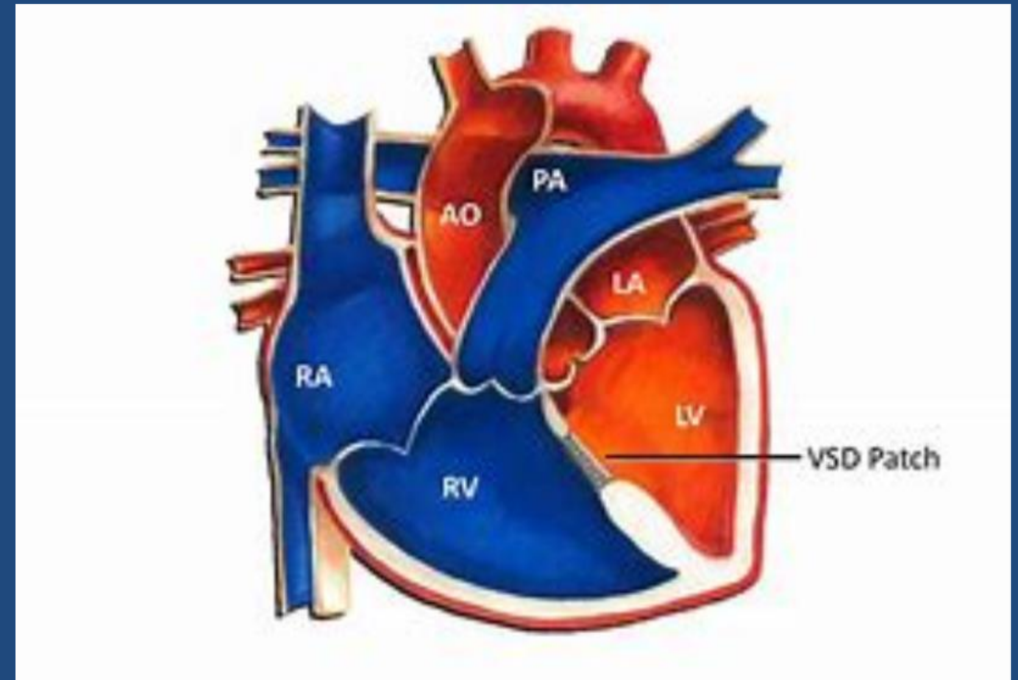


INTRAOPERATIVE DEVICE VSD CLOSURE



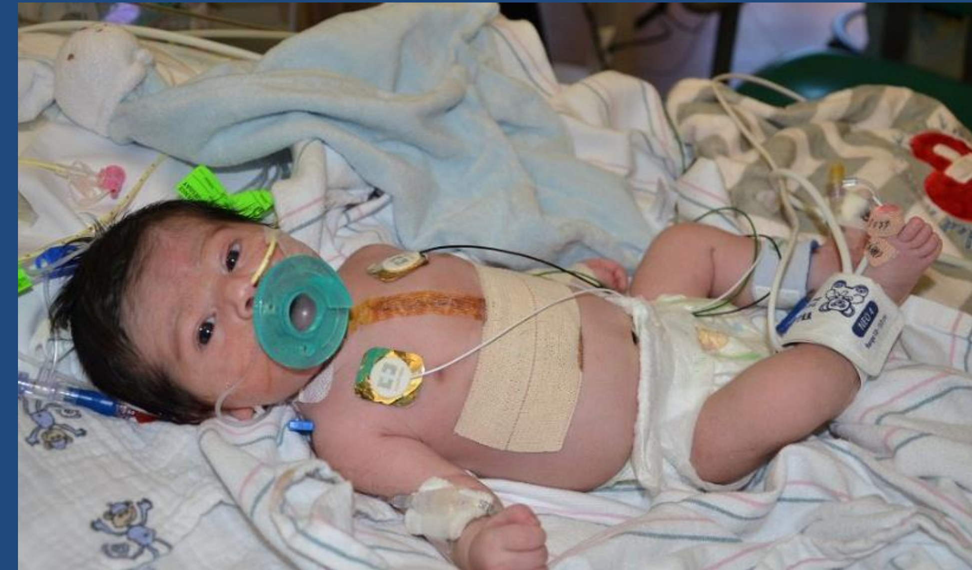
SURGICAL MANAGEMENT

- Prefer age 4-6 months
- Surgical Details
  - Use Cardiocel (bovine pericardium). Can use Dacron or Goretex patch material
  - Total time is 4 hours
    - 1st hour: Induction, PIV, A-line, EJ, intubation, foley, and monitors
    - 2nd hour: Preop TEE, chest prep, make incision, and place on CPB
    - 3rd hour: Surgical repair (45 minutes)
    - 4th hour: Place chest tube, pacer wires, and close the chest



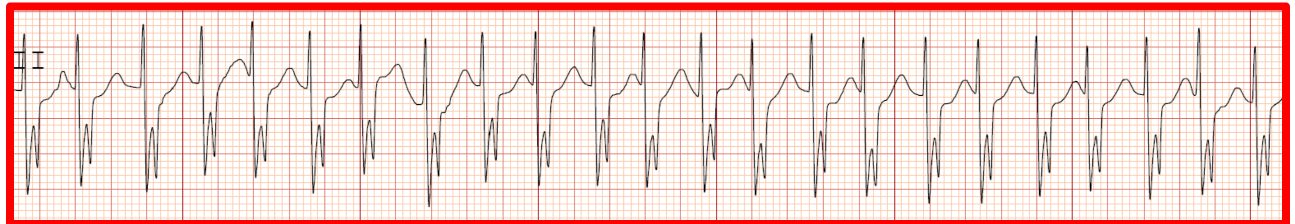
POSTOPERATIVE CARE

- Transferred to CICU
  - Try to extubate in OR or right after arrival to CICU
  - On inotropes and sedation
- POD 1-2:
  - Remove foley, CVL, A-line, CT/PW if able, & transfer to pediatric ward
- Goals for Discharge: (Typically in the hospital 2-4days)
  - Eating well & gaining weight
  - Have bowel movement
  - All tubes and line out
  - Obtain postop echo
  - Parents comfortable with care



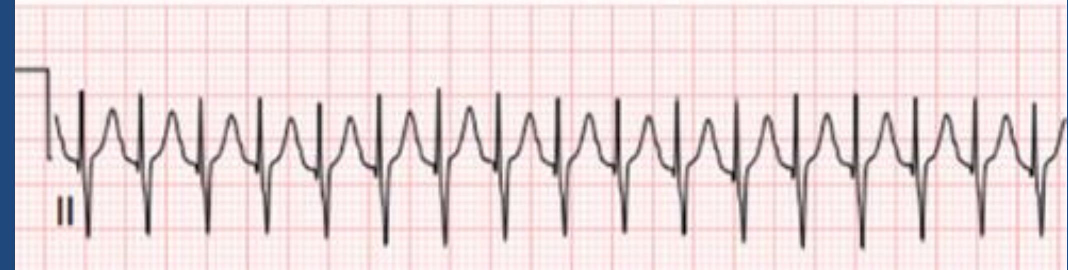
CASE STUDY CONTINUED

- Patient went to the operating room today for closure of VSD
- No complications intraoperatively
- Returned to the CICU intubated on low dose adrenaline and milrinone
- 4 hours post-operatively the nurse calls you to the bedside for tachycardia, decreased urine output, and decreased mixed venous



JUNCTIONAL ECTOPIC TACHYCARDIA

- Life threatening arrythmia originating from the atrioventricular junction or bundle of HIS
- This is a fast rhythm
- Patient can become unstable
- Associated with higher morbidity and mortality due to the refractory nature and can progress to heart failure or sudden death
- Diagnosed by EKG



TREATMENT OF JET

Decrease catecholamines

- Cooling core temperatures 35C
- Decreasing inotropes such as adrenaline and dopamine

Medication

- Amiodarone
- Dexmedetomidine (Precedex)

Overdrive pacing

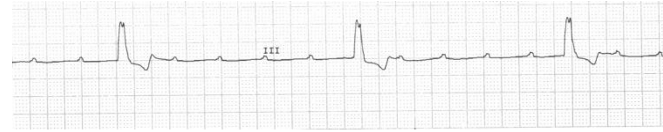
- Used to establish AV synchrony

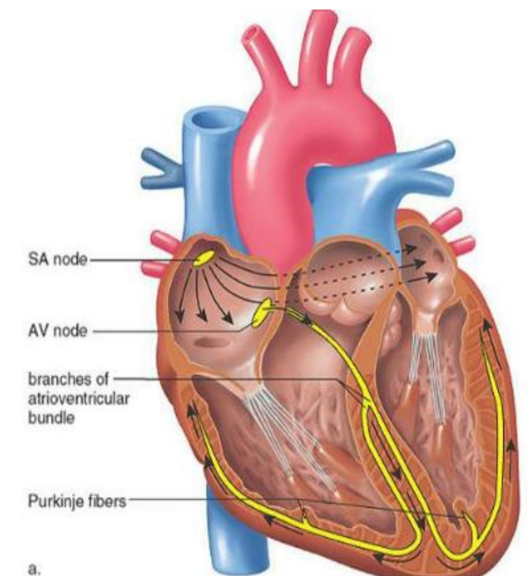
Increased automaticity = no cardioversion

Treatment goal is to restore AV conduction

SURGICAL HEART BLOCK

- Injury to the conduction system during sewing of the patch
- 5% incidence with VSD closure
- Early postop, there is edema from surgery. Wait 10 days 2 weeks to see if this reverses
- 1% permanent and would need a pacemaker system placed





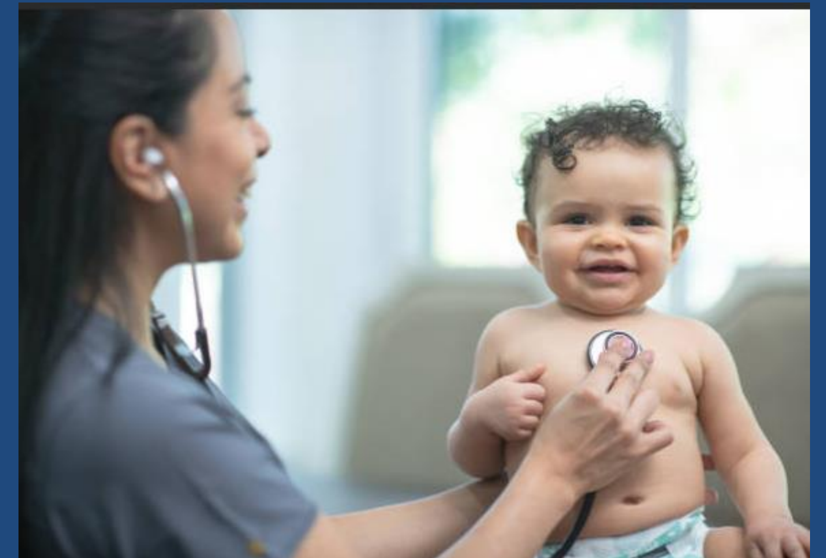
STERNAL PRECAUTIONS

Sternal precautions for 6 weeks for sternotomy:

- No lifting, holding, or swinging under the arms.
- No climbing or pulling up on furniture.
- No running where they can trip and fall and injure the sternum.
- No playground equipment, such as slides, swings, or spinning rides
- No riding in the front seat of a vehicle. Should be secured in a car seat.
- Can take a bath or shower, clean incision and chest tube sites with clean soapy water, rinse and pat dry until completely healed.
- Do NOT submerge the incision under water until completely healed (usually 6 weeks).
- Do NOT apply any lotions, creams or silicone products on incision until completely healed to prevent premature opening of incision
- Please keep incision covered from the sun for two summers with UV protected shirt to prevent Keloid scarring.

DISCHARGE INSTRUCTIONS AFTER VSD REPAIR

- No lifting over 5 lbs for 6 weeks
- Return to school in usually 2-3 weeks.
- No physical education class for 6 weeks.
- Allow to change classes before bell rings
- Have a book buddy to carry books
- Follow up with Cardiologist in 2-3 weeks



CASE STUDY THREE

- 14 year old male presents with exercise intolerance , cyanosis, and hemoptysis
- Past medical history: Murmur heard as a baby but no follow up

EISENMENGER'S SYNDROME

Who is at risk?

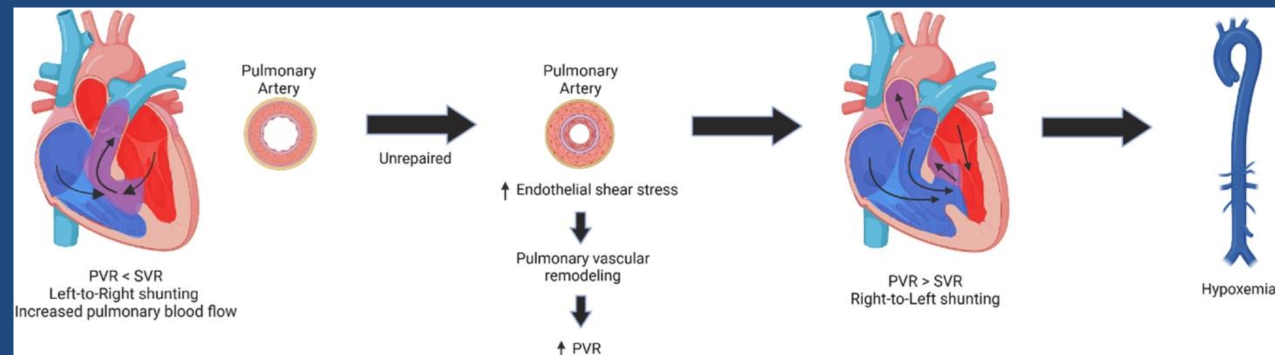
- Long standing left → right shunts
- Large non- restrictive VSD and or PDA
- Large secundum ASD
- Persistent AVC or truncus

PVR>SVR- Reversing the shunt and causing cyanosis

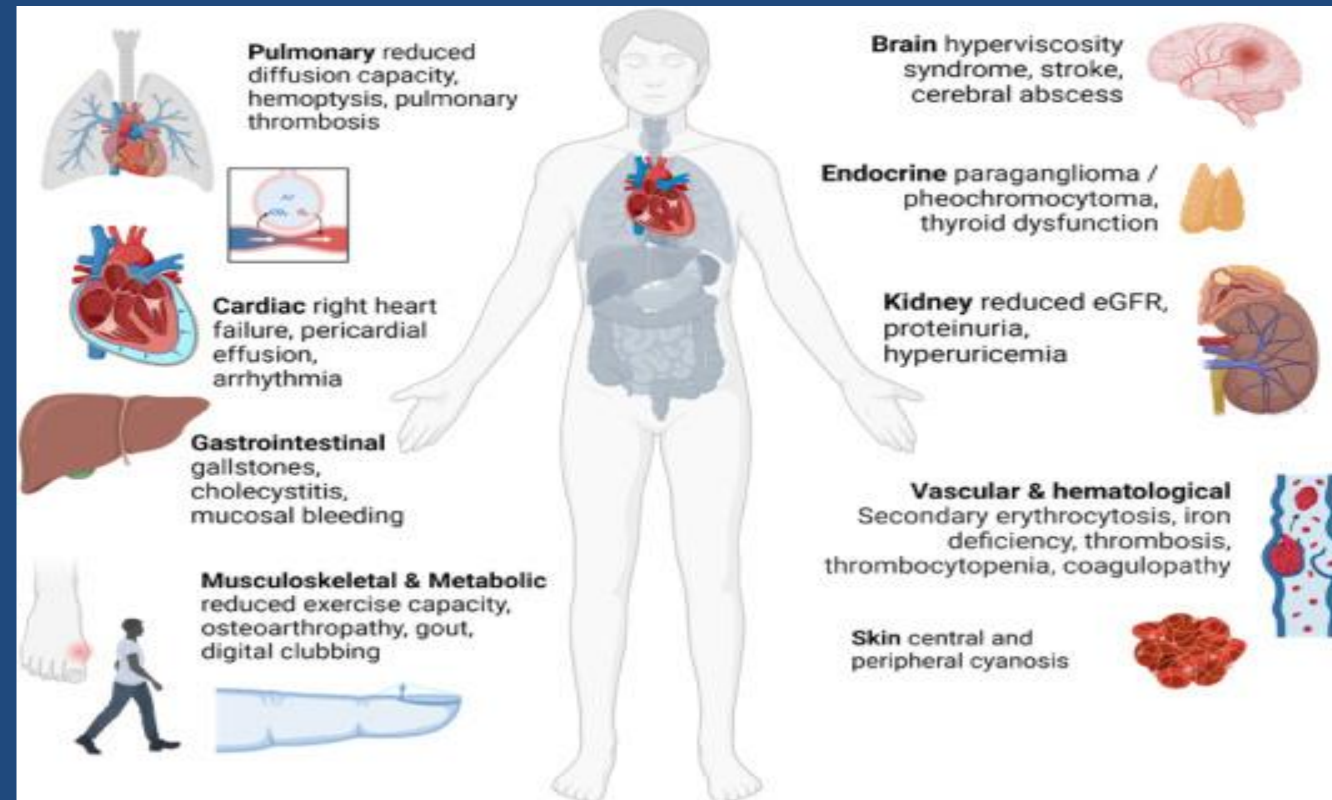
Increases risk of death 10-12 fold and carries 42% 10 yr survival

PATHOPHYSIOLOGY: EISENMENGER'S SYNDROME

- Left to right shunting causes increase in Qp and/or increased PA pressures
- Flow is inversely related to resistance and is directional proportion to pressure
- Ohm's Law: Q (flow) = P (pressure) /R (resistance)
- Pulmonary arterioles undergo morphology changes that are difficult to reverse or irreversible
- Increase PVR created bidirectional shunting
- Tunica medial proliferation and endothelial dysfunction



PHYSIOLOGIC SEQUELAE OF EISENMENGER'S SYNDROME AS A CONSEQUENCE OF CHRONIC HYPOXEMIA



QUESTIONS