Ventricular Septal Defects (VSDs)

AND

Atrioventricular septal defects (AVSDs)

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Case Study

A 2 mo M presents to the Emergency Department with cough and progressive increased work of breathing for the last 3 weeks. Mom reports that she noticed that he was “pulling in” at the ribs for the last 2 days and “looked tired” this morning so she brought him in to the ED. She reports that he has been taking longer to finish his bottle over the last month and that he hasn’t been gaining weight well compared to her first born. Mom suspects that she may not be producing enough breast milk for the baby to grow. His PCP had suggested that mom feeds him cow’s milk in addition to breast milk to supplement his feeds.

Case study cont…

On exam, the infant is tachypneic with the RR at 70bpm, he is tachycardic at 160bpm, BP is 95/52mmHg, SPO2 94% on RA. His weight is <5th percentile for age.

R/R: Tachypneic, subcostal and intercostal retractions, Coarse breath sounds bilaterally.

CVS: III/VI Pansystolic murmur heard at the apex

Questions:

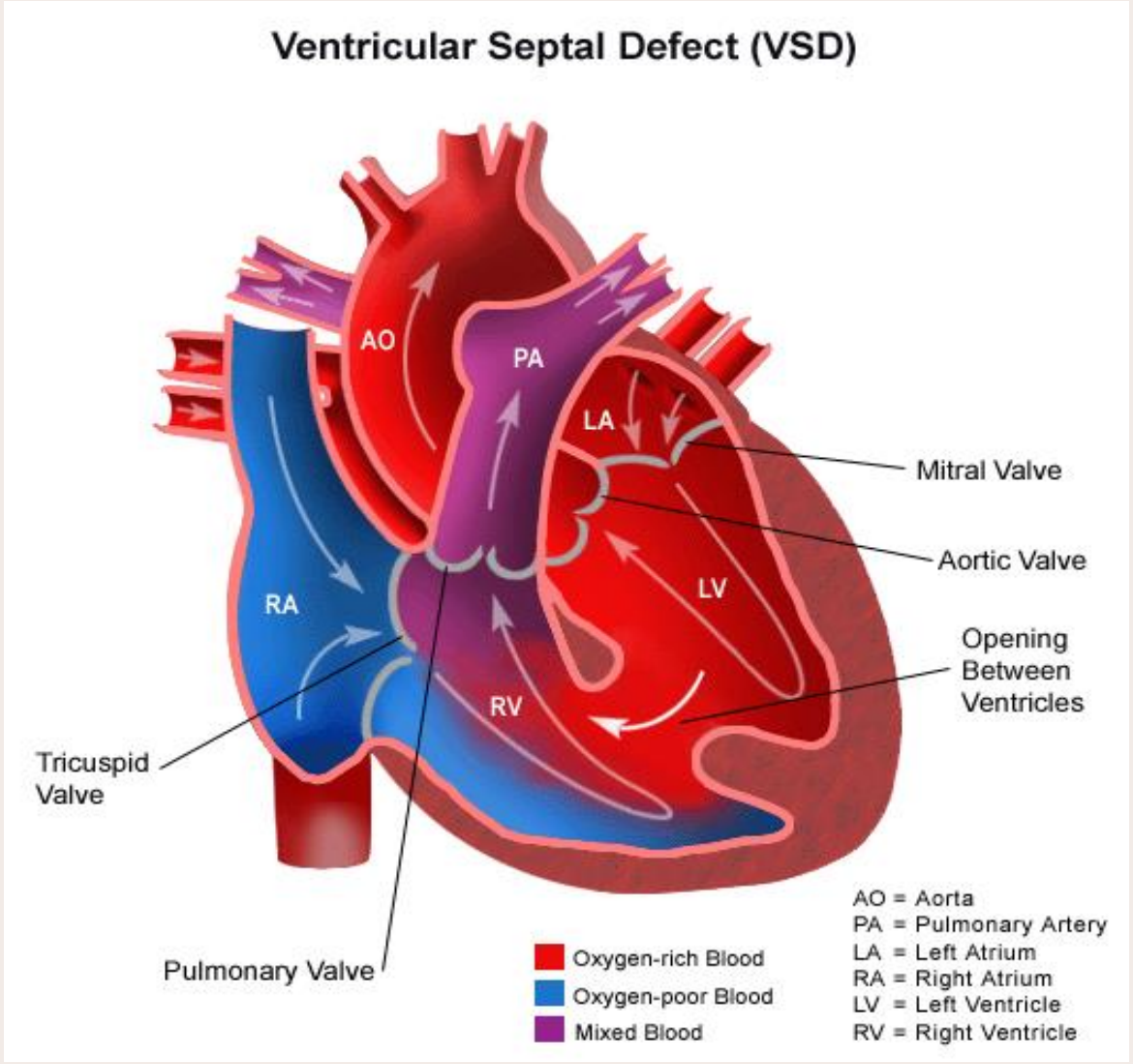
What is your differential diagnosis?

What are the lab tests and investigations that you want to order?

We will come back to these....

What is your management plan for this patient?

Ventricular Septal Defects



VSD DEFINITION:

- Ventricular Septal Defects (VSDs) are the result of abnormal development or disruption in formation of the ventricular septum during the complex process of embryological heart morphogenesis

- The pathophysiologic mechanism involves communication between left and right ventricles of the heart and very rarely between left ventricle and right atria

VSD

Most common congenital heart defect

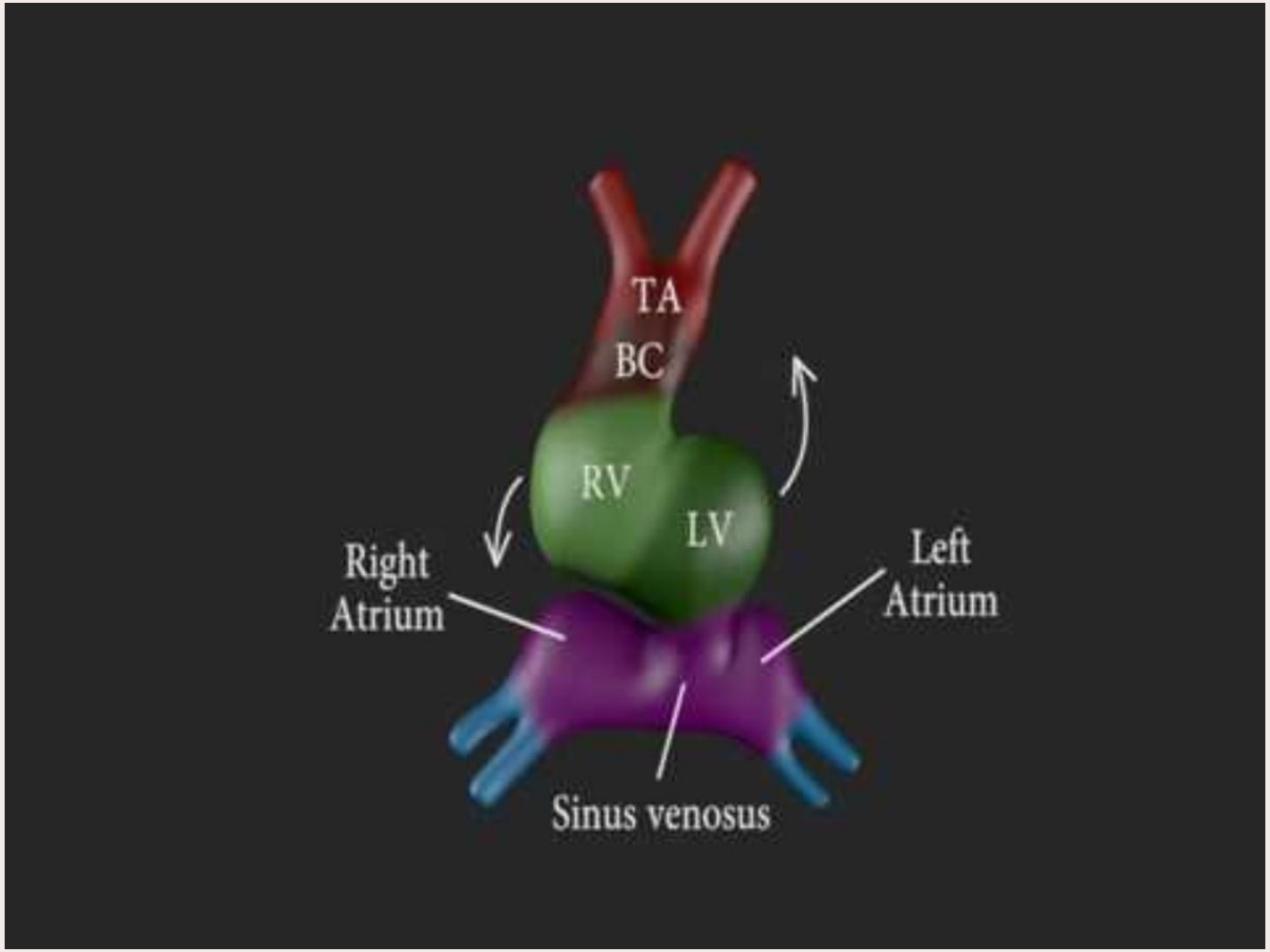
- 20% of all

A communication between the ventricles

Classified according to location along the ventricular septal wall

- Supracristal/outlet/Subpulmonary
- Perimembranous
- Muscular- anterior, mid-muscular, apical, inlet
- Inlet/AV Canal
- Gerbode Defect: LV to RA shunting
- Anterior Malaligned

VSDs: Embryology/ Cardiogenesis



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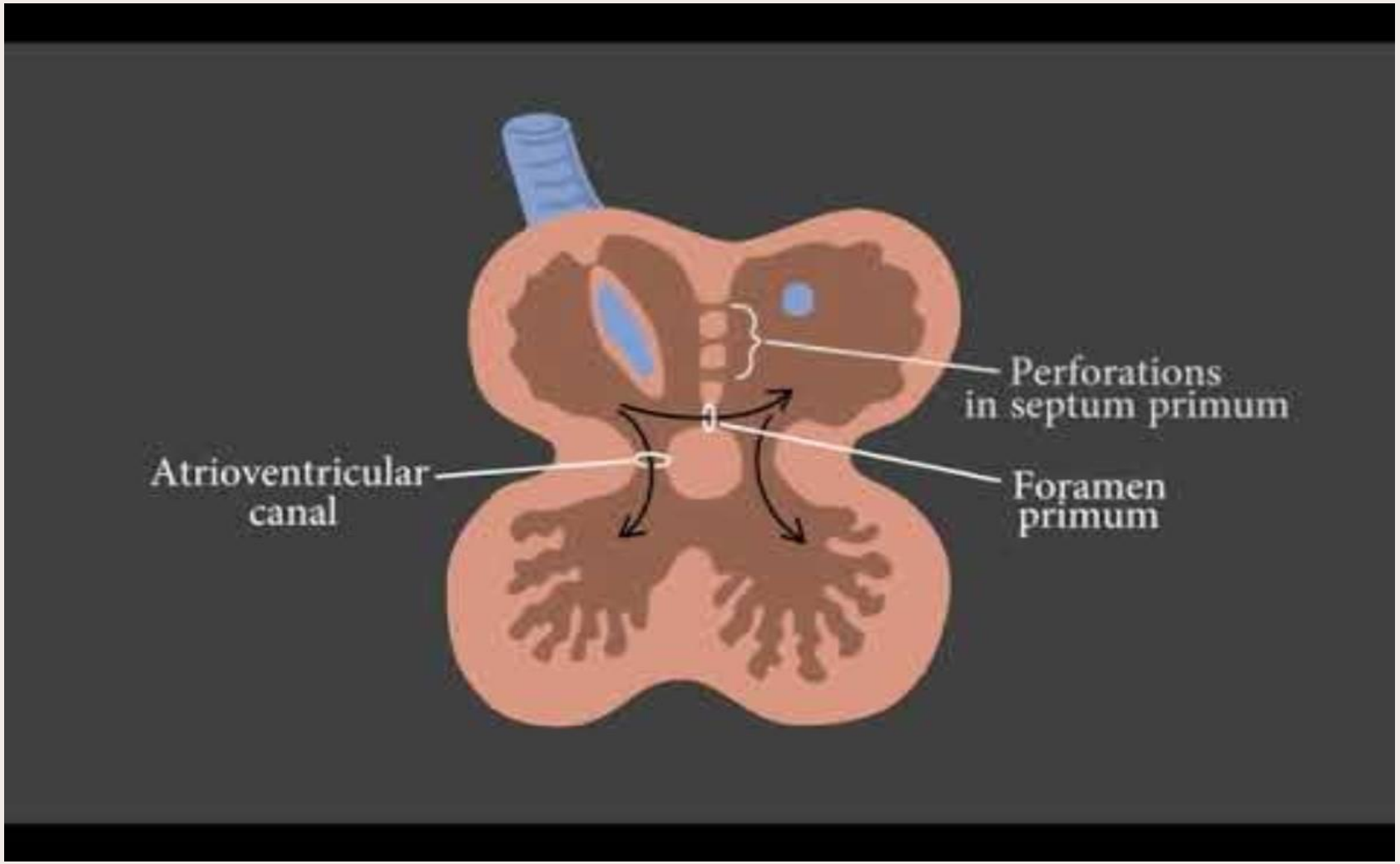


VSD: Etiology & Associated Factors

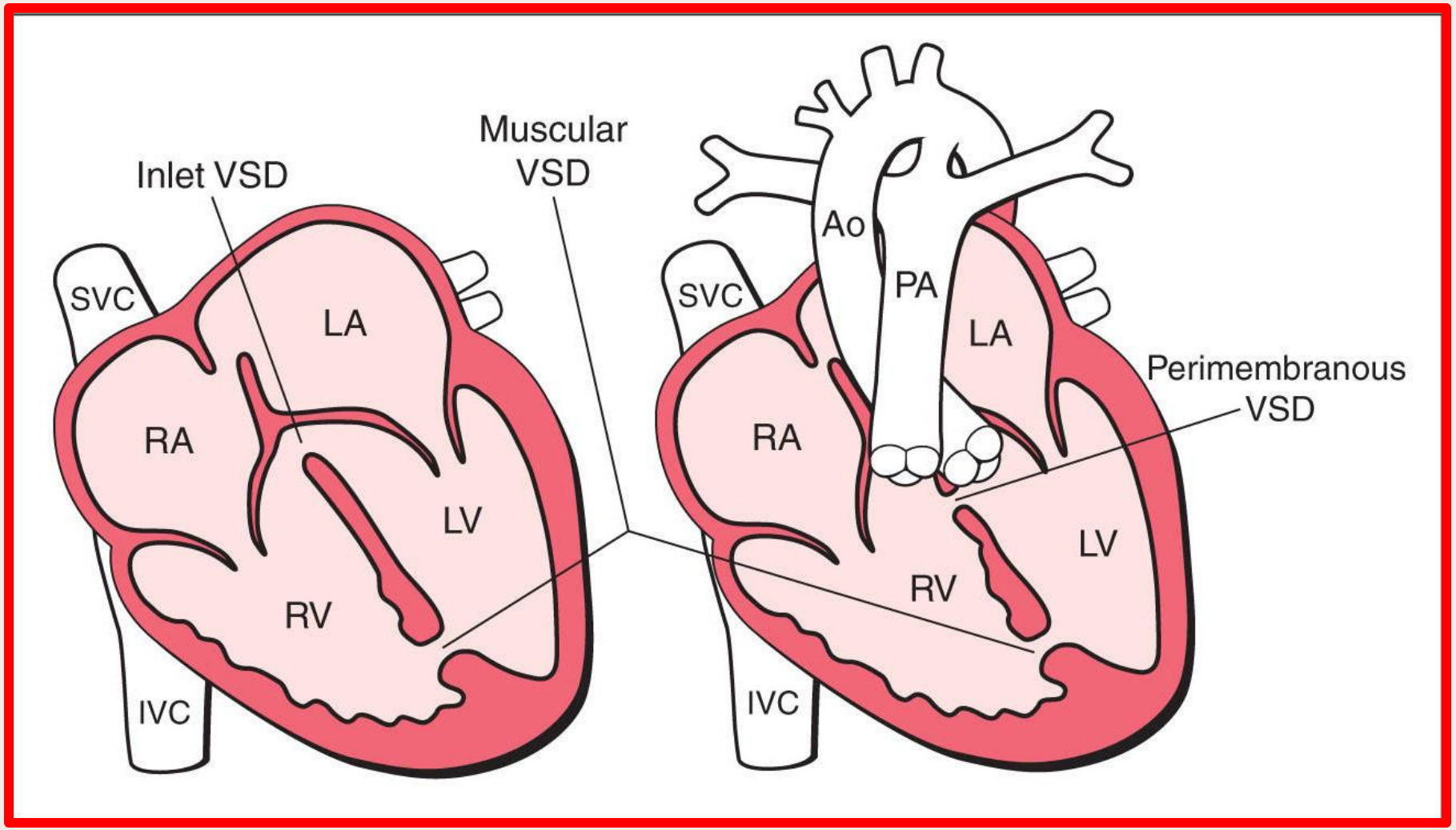
- Multifactorial
- Some types of VSDs likely arise from different developmental abnormalities: TOF (malaligned VSD)
- Maternal Infections (rubella, influenza, febrile illnesses)
- Maternal diabetes and phenylketonuria
- Maternal exposure to toxins (alcohol, marijuana, cocaine and certain medications
- Genetic factors like chromosomal abnormalities, single gene mutation and polygenic inheritance Trisomy 13, 18 and 21)

VSD Types:

The different anatomical location and hystological variations have led to various classifications of the VSDs.

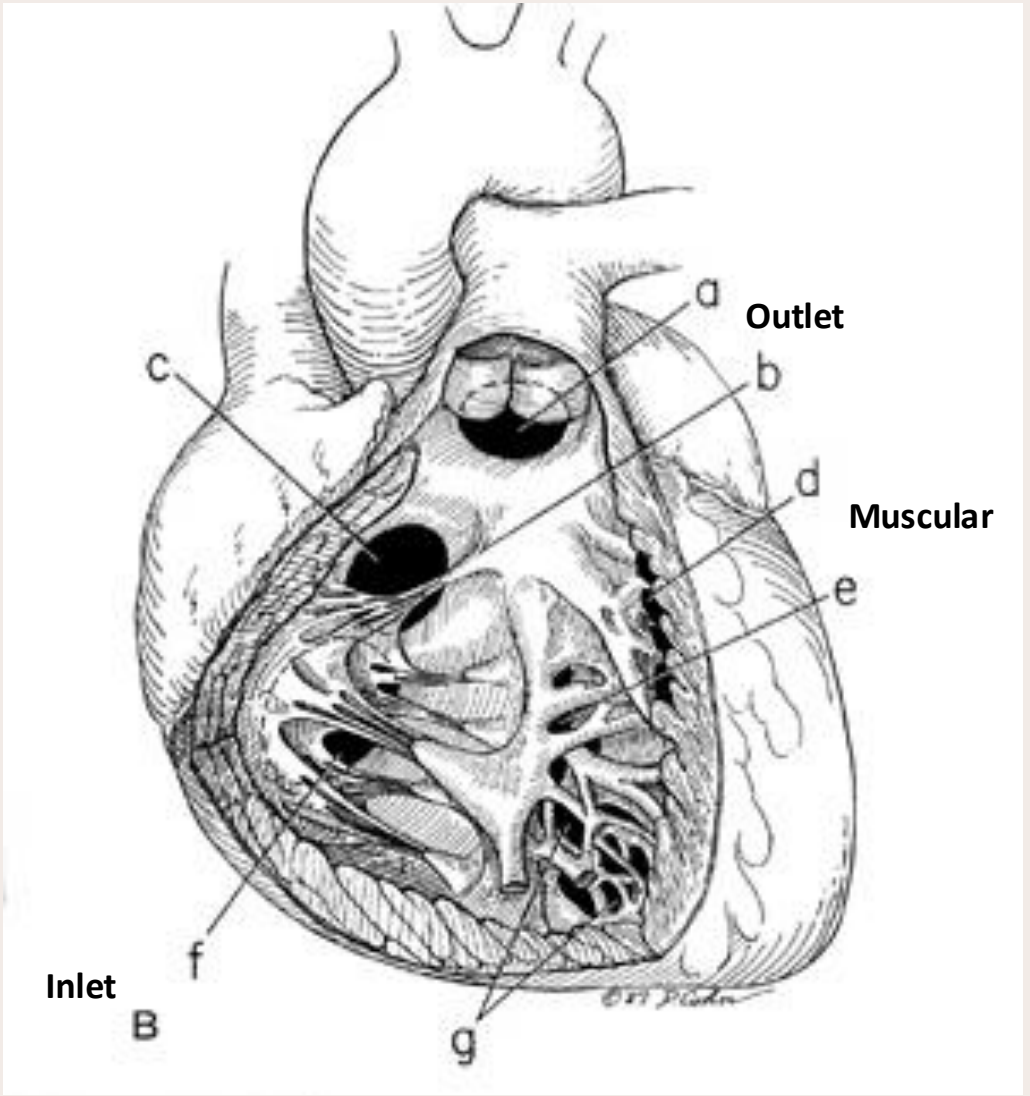
1. Perimembranous VSDs (Conoventricular)
2. Infundibular (Outlet) VSDs
3. Inlet or Atrioventricular Canal VSD
4. Muscular or Trabecular VSDs
5. Gerbode Defect: LV to RA shunting
6. Malaligned VSD: TOF

VSD Types I:



VSD Types 2

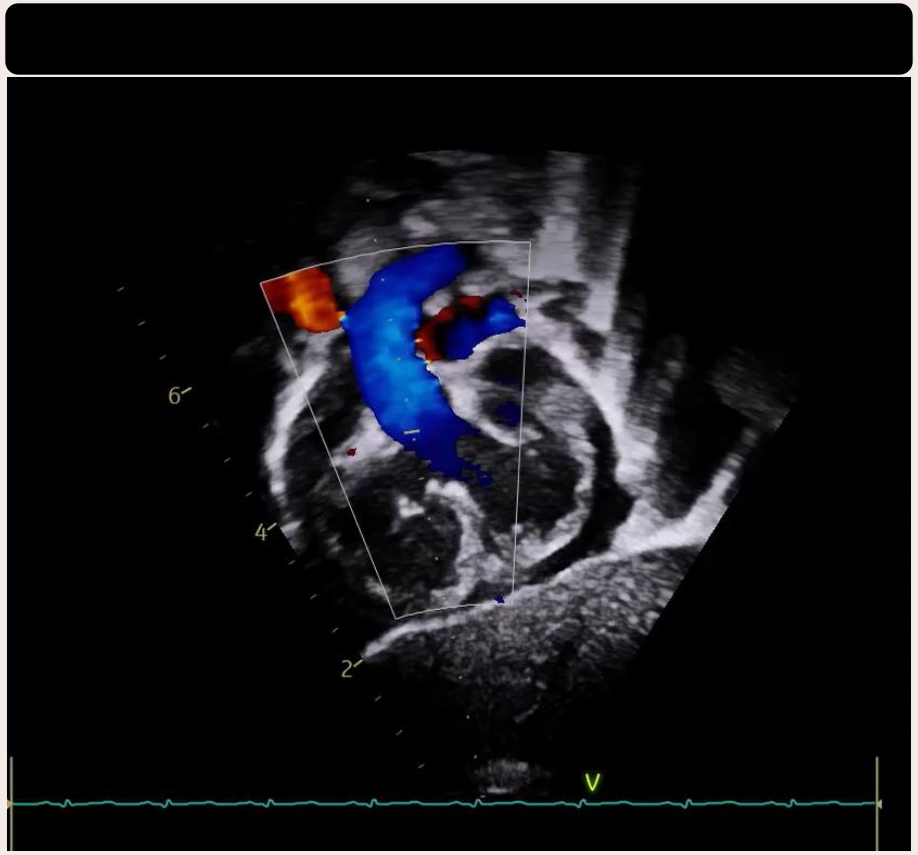
Perimembranous



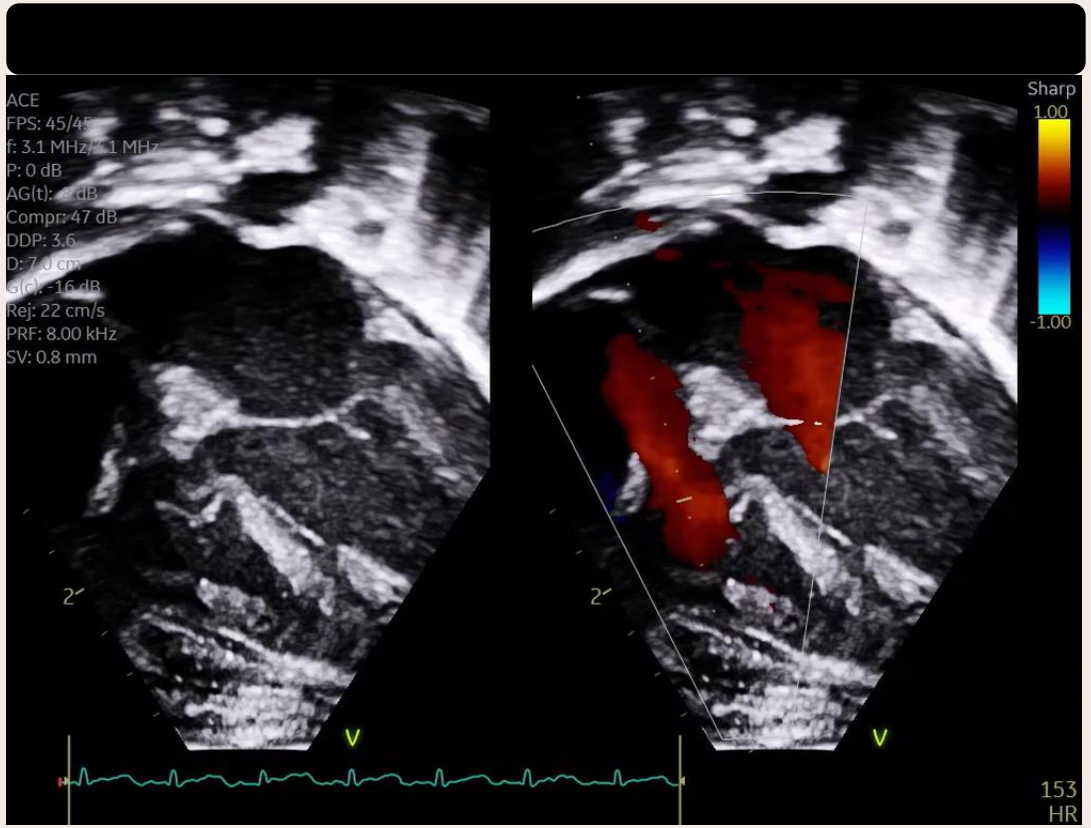
VSD Types: Perimembranous

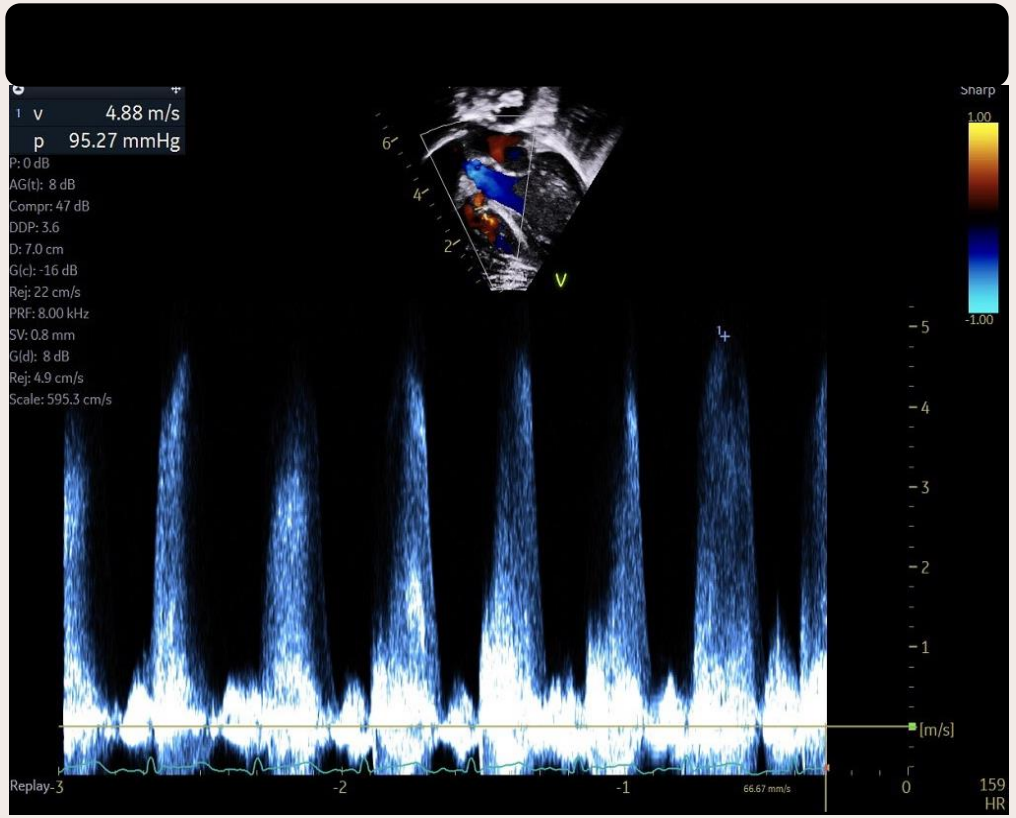
- Located in the membranous septum, below the crista supraventricularis
- Most common type of VSDs (80%)
- Can be restricted or spontaneously closed by tricuspid valve tissue
- Associated sometimes with prolapse of the aortic valve cusp and aortic insuficiency (AI)
- The amount of AI is one factor taken in consideration to determine how soon surgery is indicated
- Fixed surgically through the TV

VSD Types: Perimembranous 2



VSD Types: Perimembranous 3





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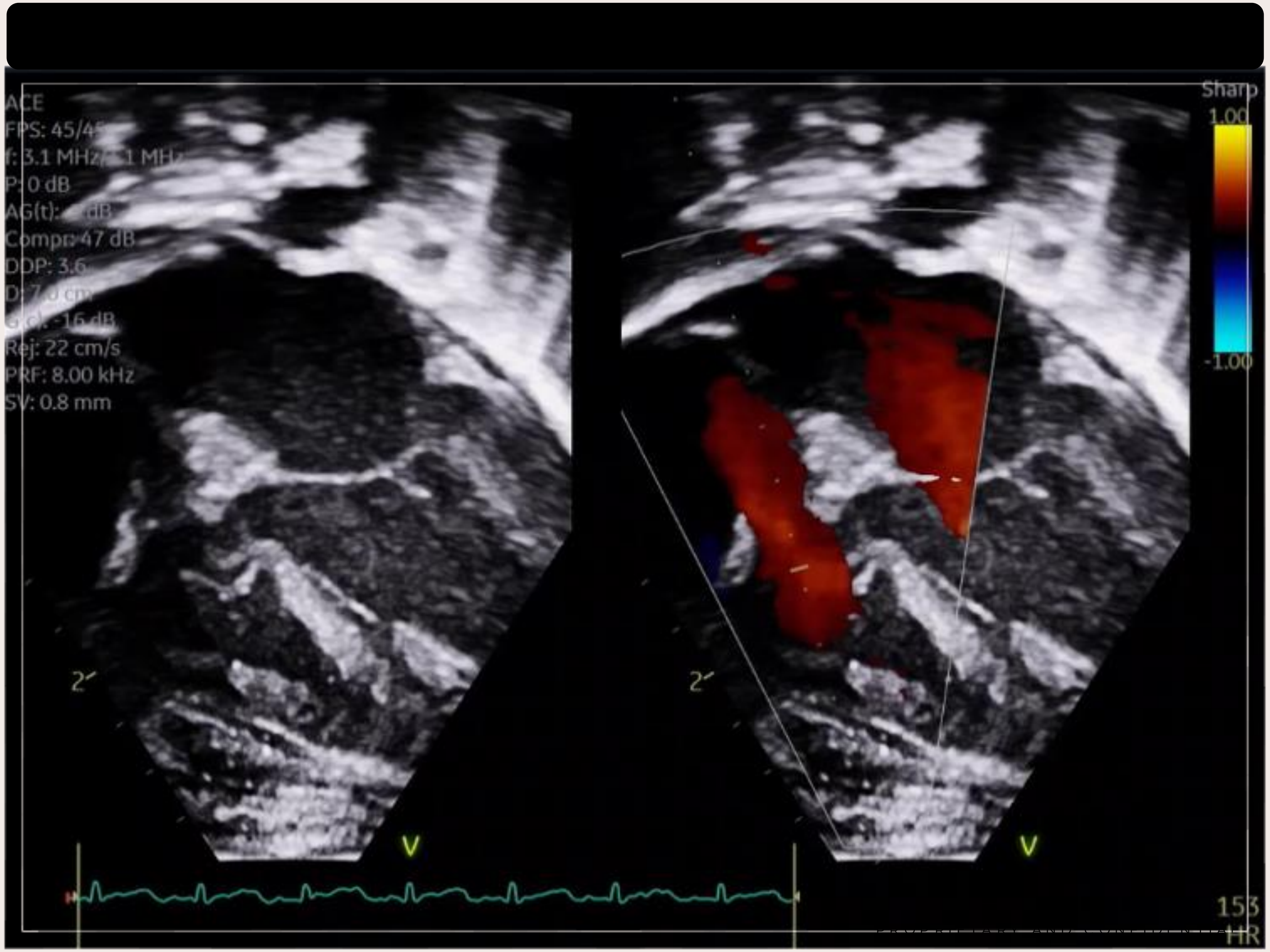
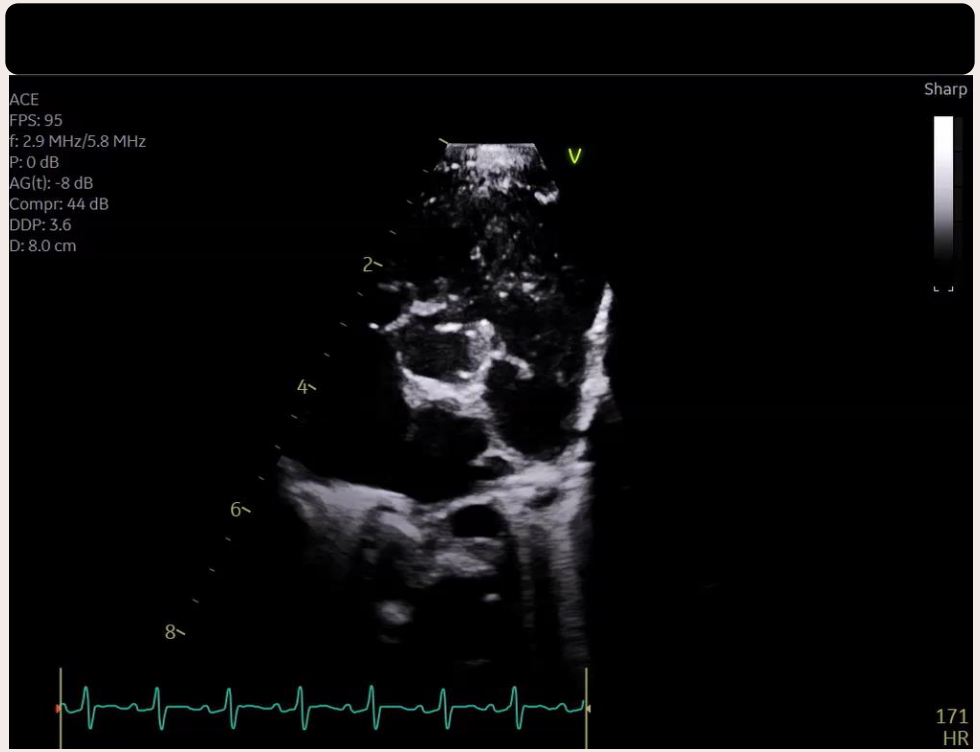
VSD Types: Perimembranous 4

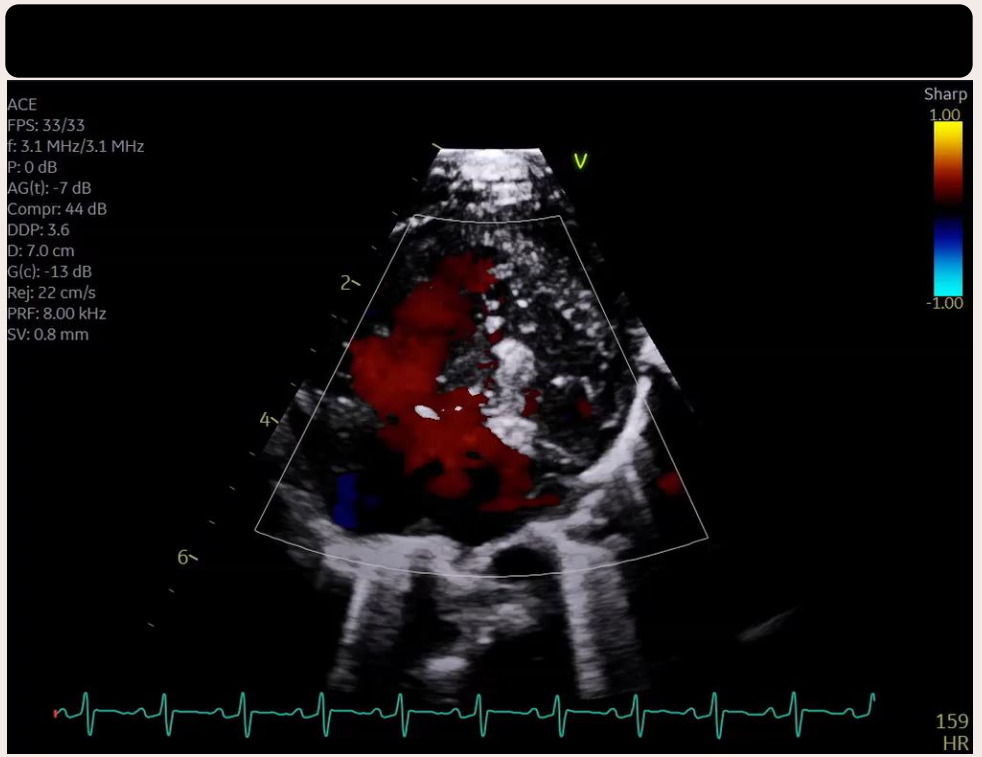


VSD Types: Infundibular (Outlet) or Supracristal

- Positioned beneath the semilunar valves (aortic and pulmonary) within the outlet septum of the right ventricle, above the crista supraventricularis
- Rare type of VSD, also referred as Supracristal (5 – 10%)
- Associated in some cases with prolapse of the aortic valve and aortic valve regurgitation/ insufficiency
- The spontaneous closure of these type of VSDs is extremely rare

VSD Types: Infundibular (Outlet) or Supracristal 2

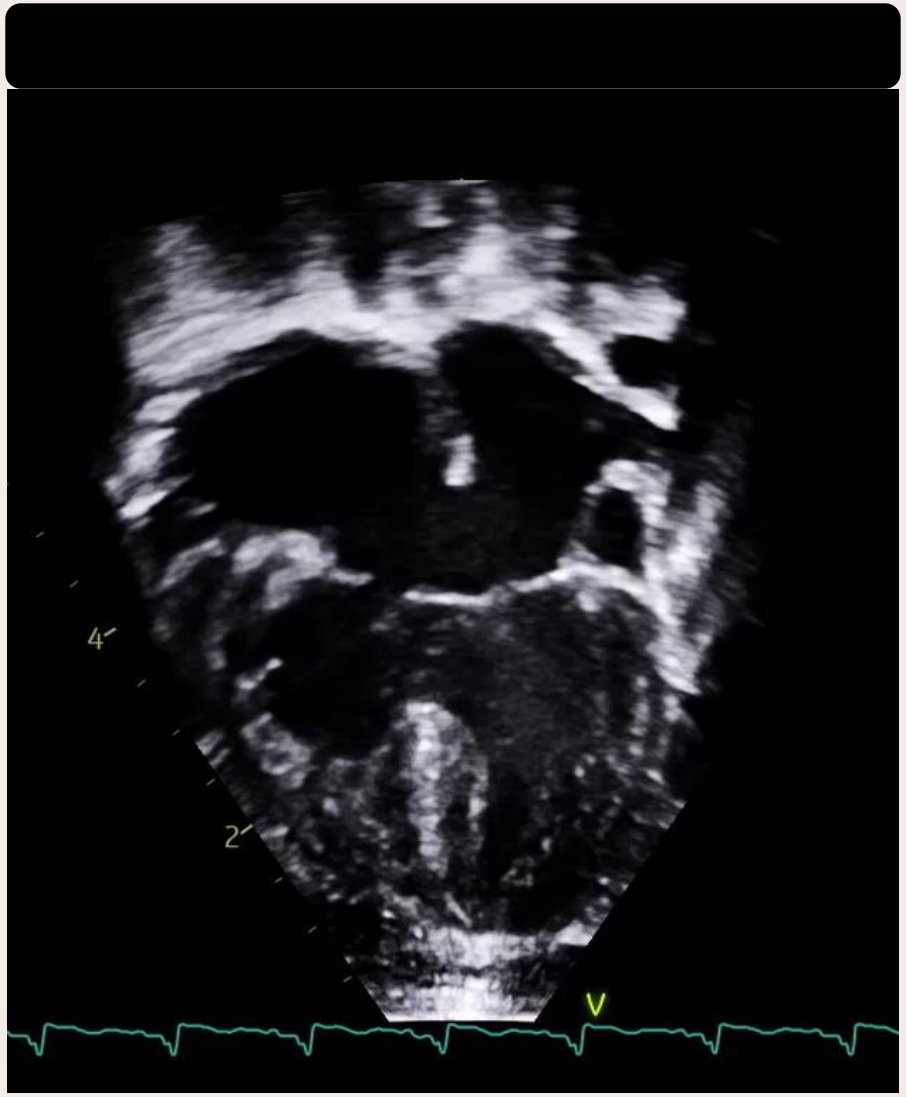


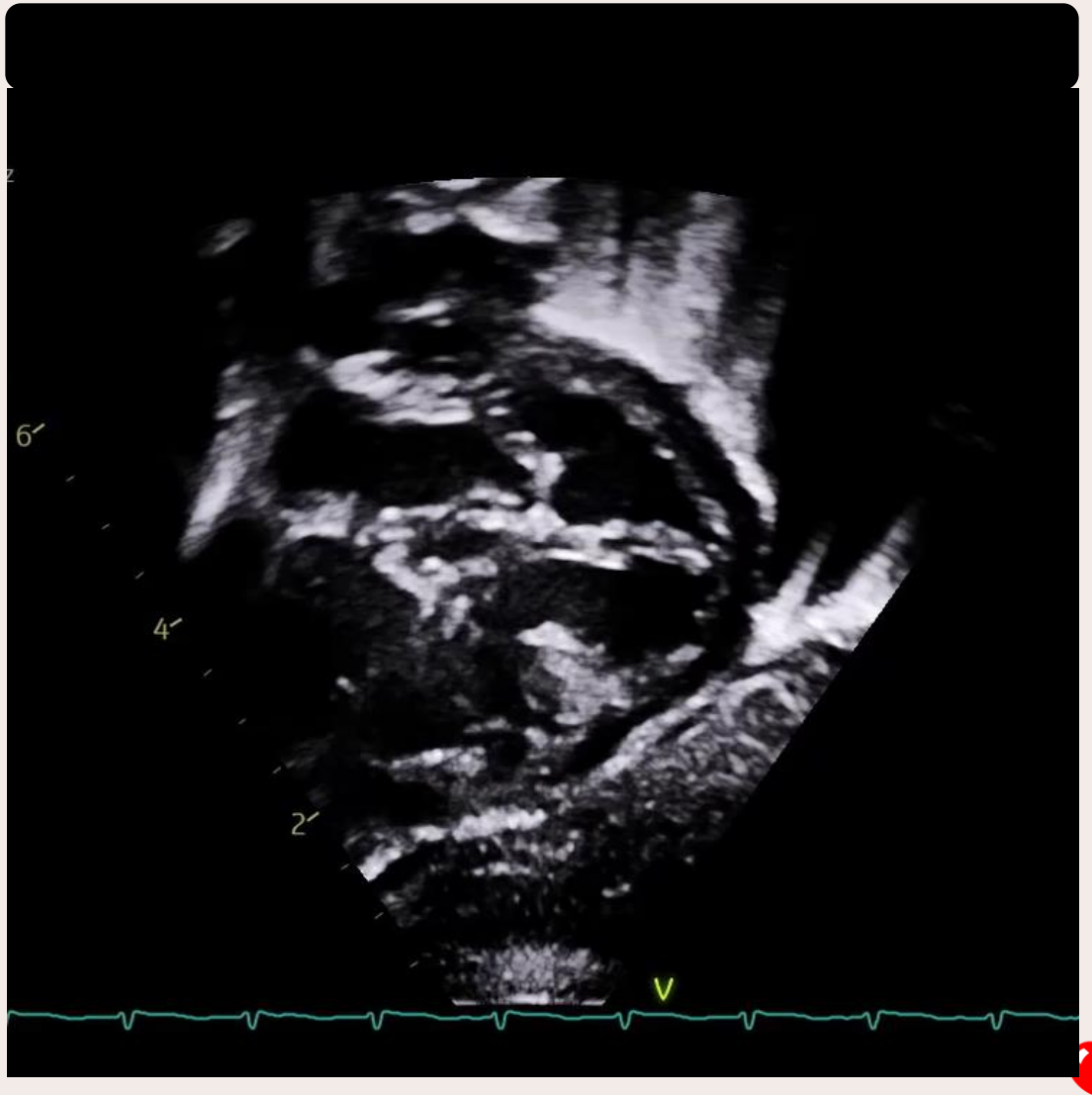


VSD Types: Inlet or Atrioventricular Canal

- Located inferior to the inlet valves (tricuspid and mitral valve) within the inlet portion or the right ventricular septum
- More frequently seen on Trisomy 21 patients
- Often associated with common, transitional or intermediate atrioventricular septal defects

VSD Types: Inlet or Atrioventricular Canal 2





VSD Types: Muscular or Trabecular

- Located in the apical, central, anterior or outlet regions of the muscular septum
- More likely to spontaneously close on their own

VSD Types: Muscular or Trabecular 2

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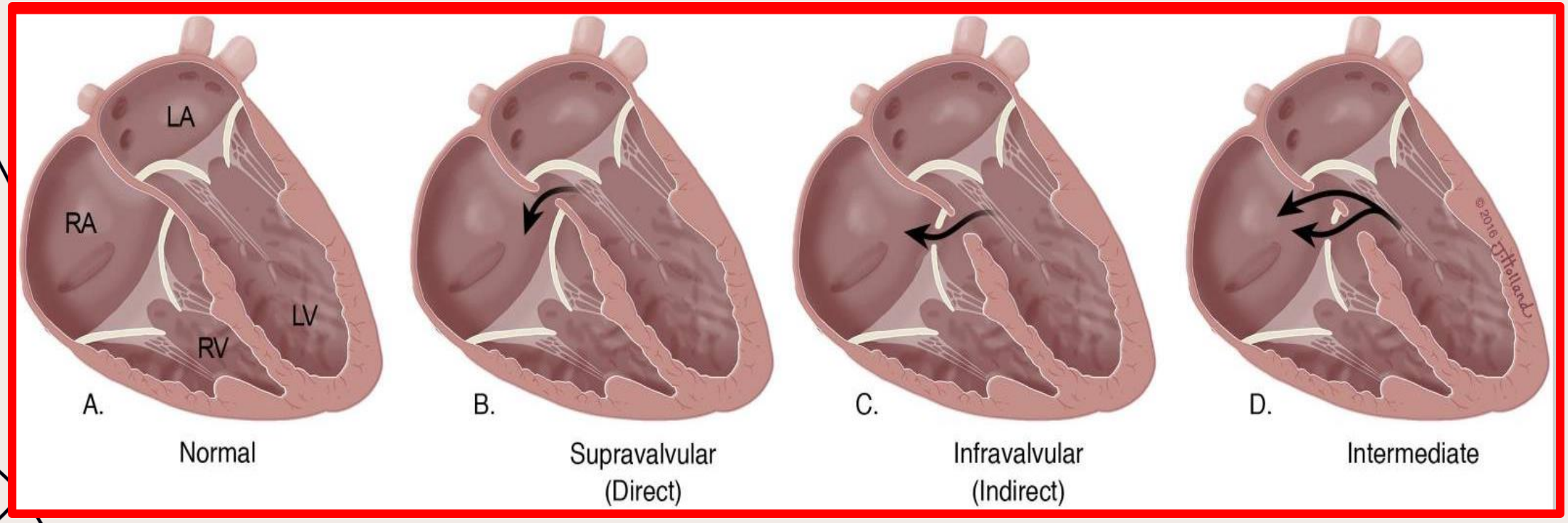
VSD Types: Gerbode Defect

- a high-velocity shunt flow from the left ventricle to the right atrium, often during systole
- **Congenital** (present at birth) or **Aquired** after heart surgery (AOV or MV replacement, AVSD repair…)
- Leads to dilation of the right atria

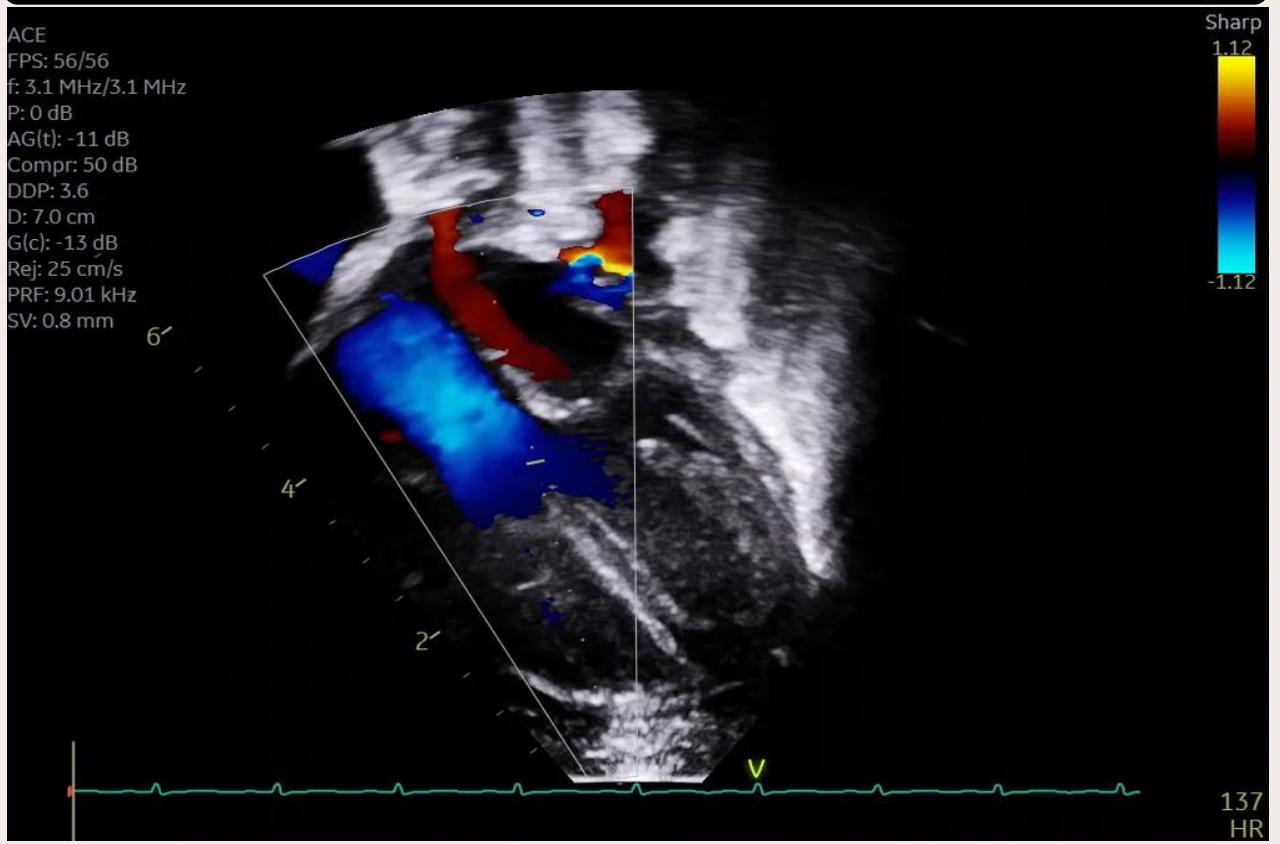
VSD Types: Gerbode Defect 2



VSD Types: Anterior Malaligned

- a type of heart defect where the conal septum (a part of the ventricular septum) is displaced forward, often towards the right ventricle outflow tract (RVOT).
- It can lead to right ventricular outflow obstruction
- Commonly associated with Tetralogy of Fallot, sometimes with Pulmonary Atresia/ VSD

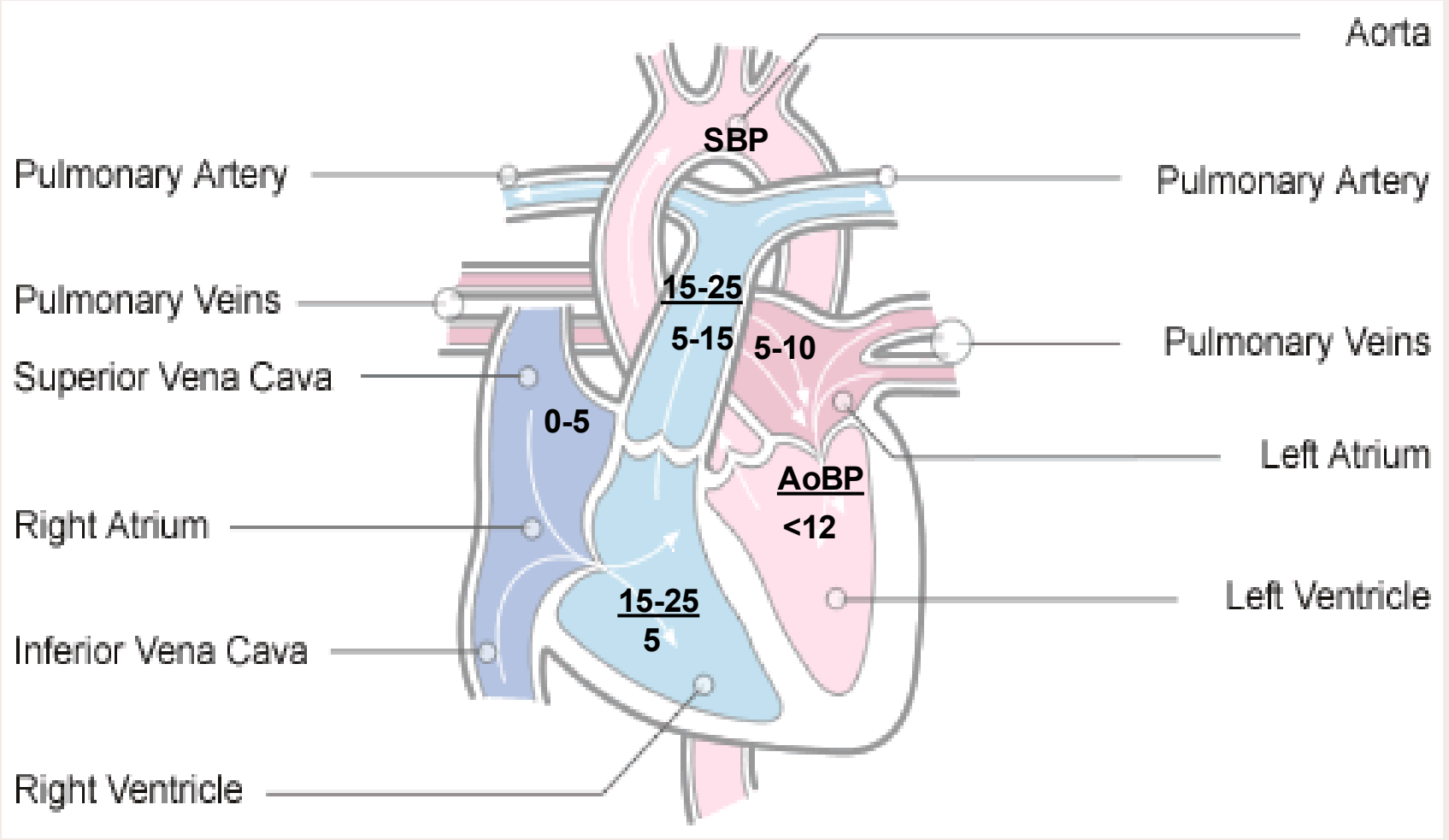
VSD Types: Anterior Malaligned 2



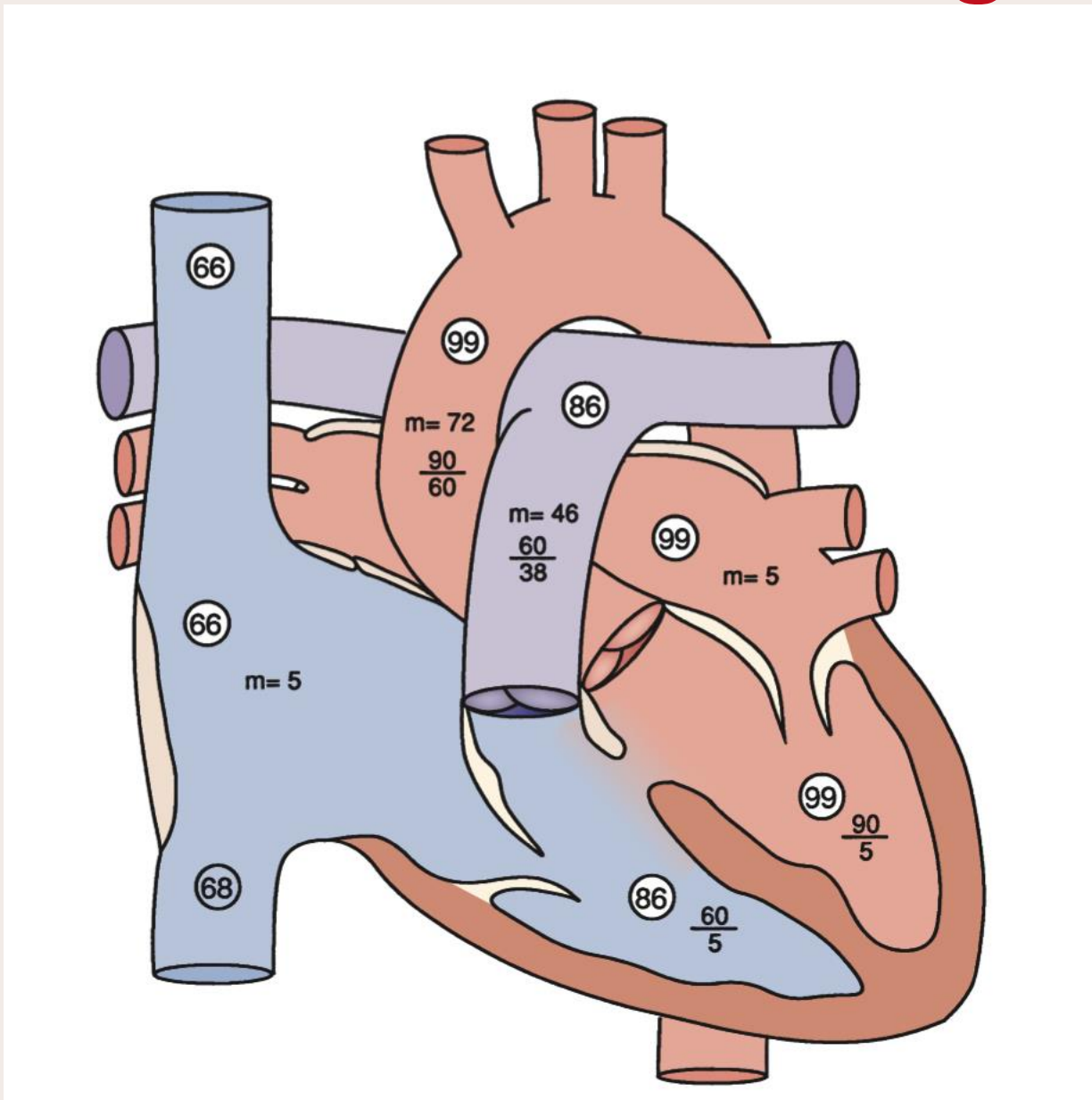
VSD:Associate Defects

- Atrial septal defect (ASD)
- Patent ductus arteriosus (PDA)
- Coarctation of the aorta (COA)
- Subaortic stenosis,
- Transposition of the great arteries (TGA)
- Tetralogy of Fallot (TOF)
- Truncus arteriosus (TA)
- Double outlet right ventricle (DORV)

Normal Intracardiac Pressures



Pressure and oximetric changes in VSD



Pathophysiology:

Dependent on:

- Size of the defect:
  - Restrictive vs Non-restricive:

- Relationship between SVR and PVR:
  - PVR initially high at birth, falls between 2 to 6 weeks leading to increased pulmonary blood flow

**These factors determine the magnitude of flow, which determines the presentation.

Principles of Blood Flow

Qp:Qs

Qp= Pulmonary artery- pulmonary vein

Qs= Systemic artery-systemic vein

Normal heart 1:1 but how do we get that??

A hole of significant size will equalize pressure between chambers

VSD Clinical Manifestations

At Birth

- High PVR
  - Restricts the L to R shunting to due higher pressure right side
  - No murmur
  - No signs and symptoms of CHF

Later

- As PVR decreases
  - L to R shunting
  - More blood flow to lungs
  - Murmur
  - Signs and symptoms of CHF become evident
- 4 – 12 weeks
  - ↑RR
  - Poor feeding
  - Diaphoretic
- Older child
  - ↓ in exercise tolerance
  - Delayed growth/development

As a result of Increased blood flow to the lungs

- Tachypnea
- CHF symptoms
    - Dyspnea on exertion (Taking longer to feed, sweating with feeds)
    - Dyspnea at rest.
    - Failure to thrive.

Physical exam:

- Pansystolic murmur localized to 2nd and 3rd ICS.
- Diastolic murmur -Qp:Qs >2:1

VSD Diagnostics

CXR

- Cardiomegaly
- Increased pulmonary vascular markings

Echocardiogram

- Position and size of defect
- Gradient across defect
- LA and LV dilation
- +/- Mitral regurgitation
- Gerbode defects (Erroneous PH diagnosis)

EKG

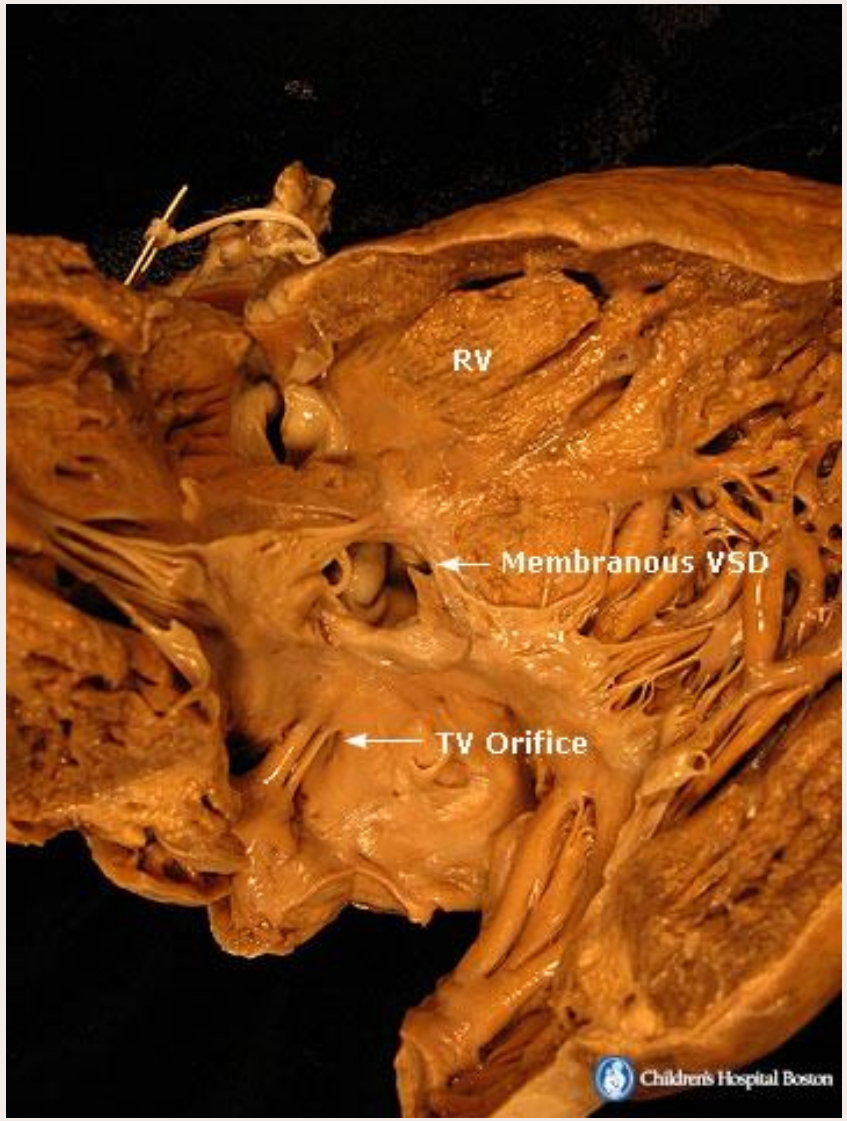
- Large VSD- Biventricular hypertrophy and LAH
- Not specific for VSDs, usually suggest the HD state. Tall upright T waves in V1 and V2 (sys RVP), LVH

VSD Complications

- Aortic valve prolapse and aortic valve insufficiency
- Subaortic membrane
- Double Chamber RV
- Congestive heart failure (CHF) due to left atrial and left ventricular dilation
- Failure to thrive due to poor feeding
- Pulmonary hypertension due to lung congestion related to increase blood flow
- Endocarditis

VSD Facts

- Spontaneous closure
  - 30-40% of all VSDs may close
    - small defects
    - muscular defects
  - Usually within 1st year of life
- Medical management
  - Diuretics
  - Digoxin
  - Afterload reduction
  - Nutrition



Who gets VSD surgery?

- Significant left to right shunting (QP:QS 2:1)
- Pts with aortic regurgitation or AoV cusp prolapse
- Infants with CHF and FTT unresponsive to medical management
- Infants with evidence of high pulmonary vascular resistance (PVR)
- Infants who respond to medical therapy may be operated on between 12-18 months- up to 5yr

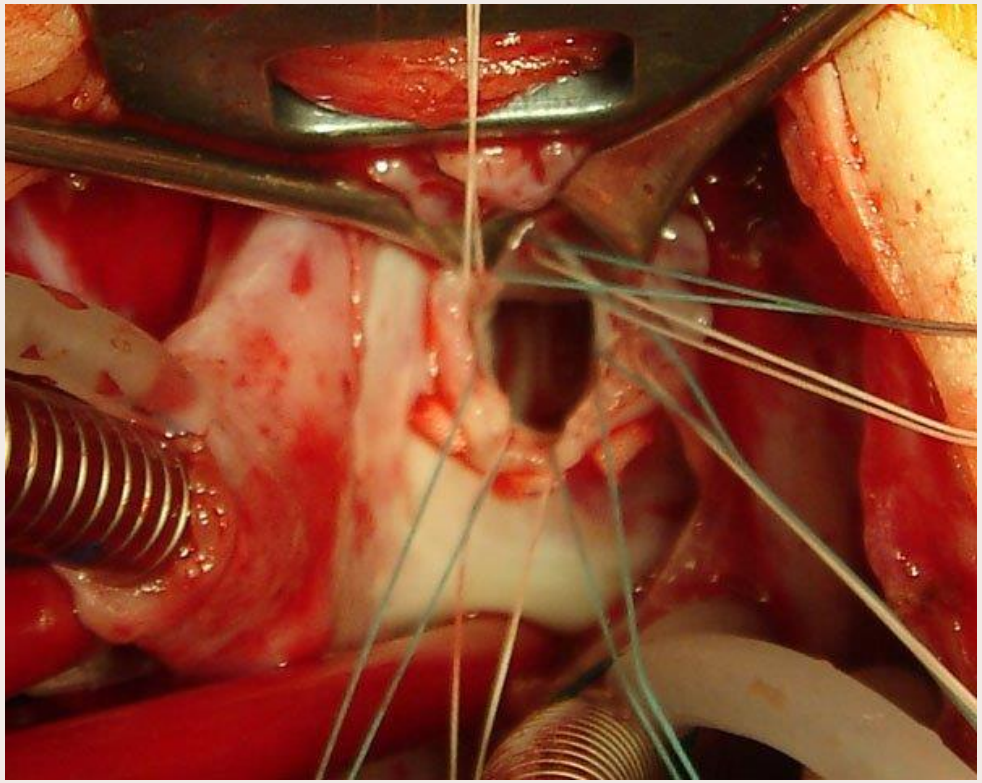
Eisenmenger's complex

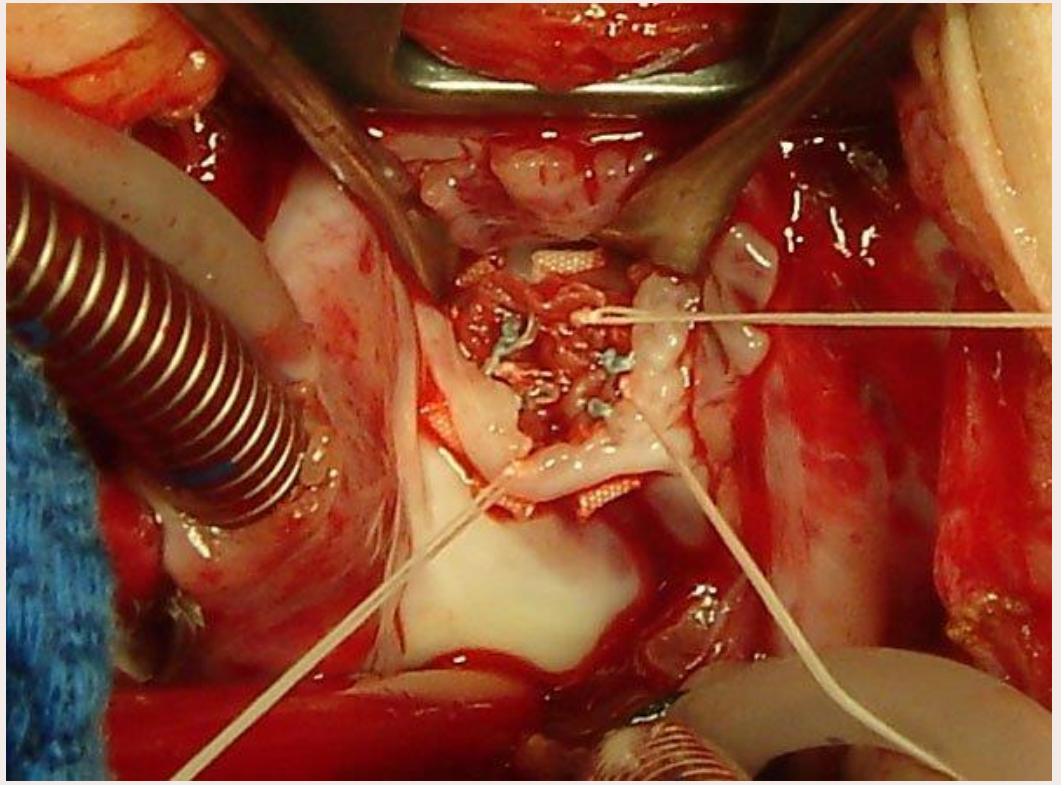
PVR>SVR- Reversing the shunt and causing cyanosis

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

VSD: How is the surgery done?

- Cardiopulmonary bypass
- Via right atrial incision-anatomy inspected through tricuspid valve
- Ventriculotomy
- Dacron or PTFE patch
- Transcatheter device





VSD: Surgical Considerations

- Usually short ICU stay
- Hypertension
- Consider degree of CHF pre-op
  - PAH crisis
- Arrhythmias
  - Transient heart block
    - Usually resolve 24-48hrs days due to decreased swelling, at CMCD 14 day no resolution = pacemaker
  - Junctional ectopic tachycardia (JET), Supraventricular tachycardia (SVT)

VSD: Outcomes

- Residual defect 20-33%
- Further surgical intervention- 5.5%
- Mortality
  - Isolated VSD- <1%
  - Multiple- 5-10%

Case Study continued

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Case study continued….

On exam, the infant is tachypneic with the RR at 70bpm, he is tachycardic at 160bpm, BP is 95/52mmHg, SPO2 94% on RA. His weight is <5th percentile for age.

R/R: Tachypneic, subcostal and intercostal retractions, Coarse breath sounds bilaterally.

CVS: III/VI Pansystolic murmur heard at the apex

Questions:

What is your differential diagnosis?

What are the lab tests and investigations that you want to order?

What is your management plan for this patient?

A patient in Zambia...

- 6month Trisomy 21, Balanced AV-Canal
- Medications: Furosemide 5mg PO TDS, Spironolacton 6.25mg PO BD, Levothyroxine 25mcg PO BD
- Reported Symptoms: None
- Family HIstory: None
- Exam: Features consistent with Trisomy 21, subcostal recessions and transmitted breath sounds, Grade III systolic murmur loudest at LSB, hepatomegaly
- Vitals: P: 141b/m, BP 92/53mmHg, RR 27b/m, 96% RA, Weight 25% on Trisomy 21 chart
- EKG: Normal Sinus rhythm, PRI 142msec; Rate 150b/m, mean frontal QRS axis –81*, Biventricular hypertrophy
- CXR: Enlarged cardiac shadow with increased cardiothoracic ratio 0.7 and increased pulmonary vascular markings
- Echo: Complete balanced AVC with multiple cleft and consequent AVVR and mild pericardial effusion

thank you

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