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:

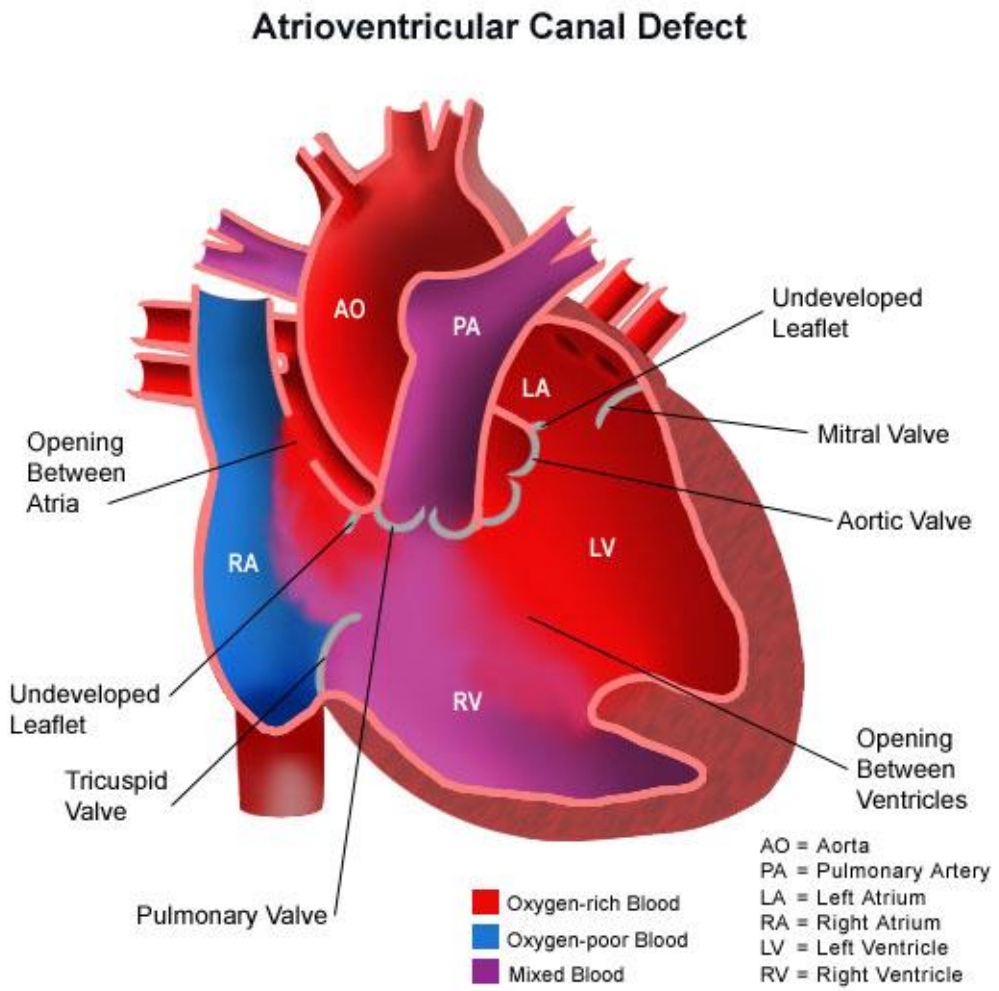
What is your differential diagnosis?

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

What is your management plan for this patient?

We will come back to these....

Atrioventricular Canal Defects

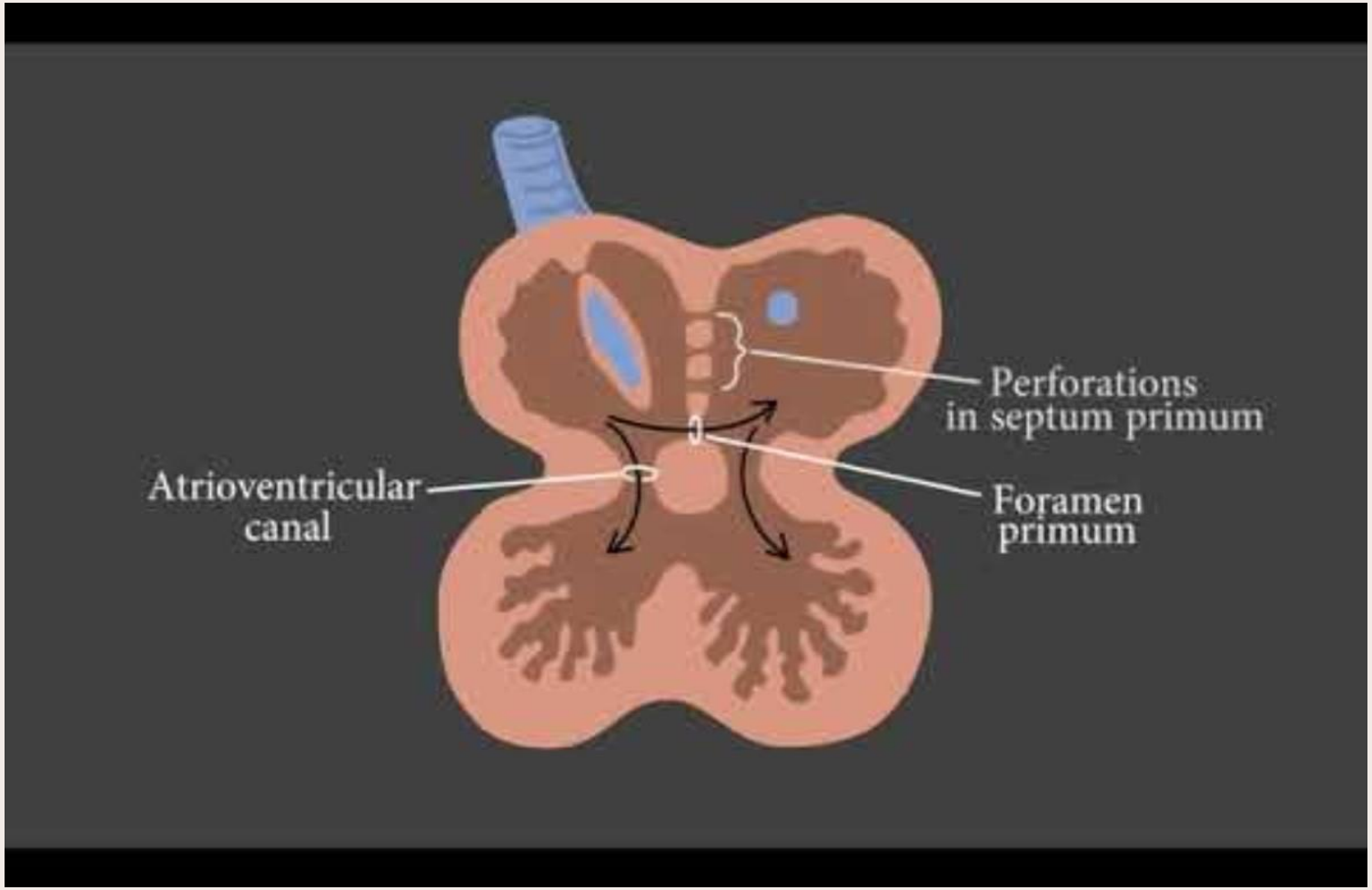


Atrioventricular Canal Defect

- Also refered as **AVSD** or **endocardial cushion defect**

- A congenital heart defect characterized by holes in the heart between the left and right receiving (atria) and the pumping chambers (ventricles)

AVSDs: Embryology/ Cardiogenesis



AVSDs: Etiology & Risk Factors

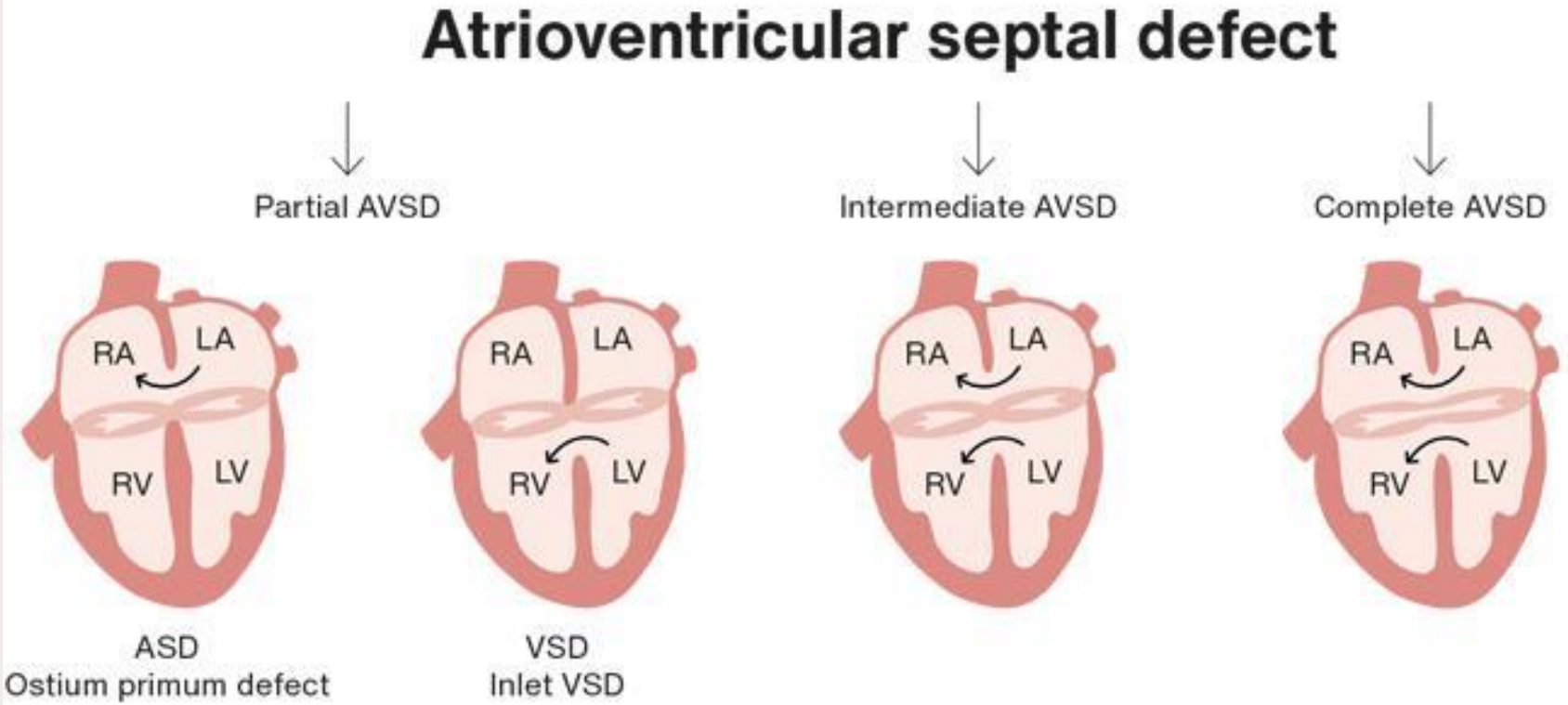
- Atrioventricular Septal Defect (AVSD) arise from the abnormal development of endocardial cushions during early embryogenesis
- The endocardial cushions play an important role in formation of the lower portion of the atrial septum and upper portion of the ventricular septum, as well as formation on the inlet valves (Tricuspid and mitral)
- Syndromic conditions such as Down Syndrome are strongly associated with these types of congenital heart defects, about 4 – 5%
- Maternal diabetes and obesity are risk factors associated with AVSDs

AVSD: Associated lesions

- 7-25% association with other cardiac defects
- Heterotaxy
- TOF
- DORV
- Transposition of great arteries
- PDA

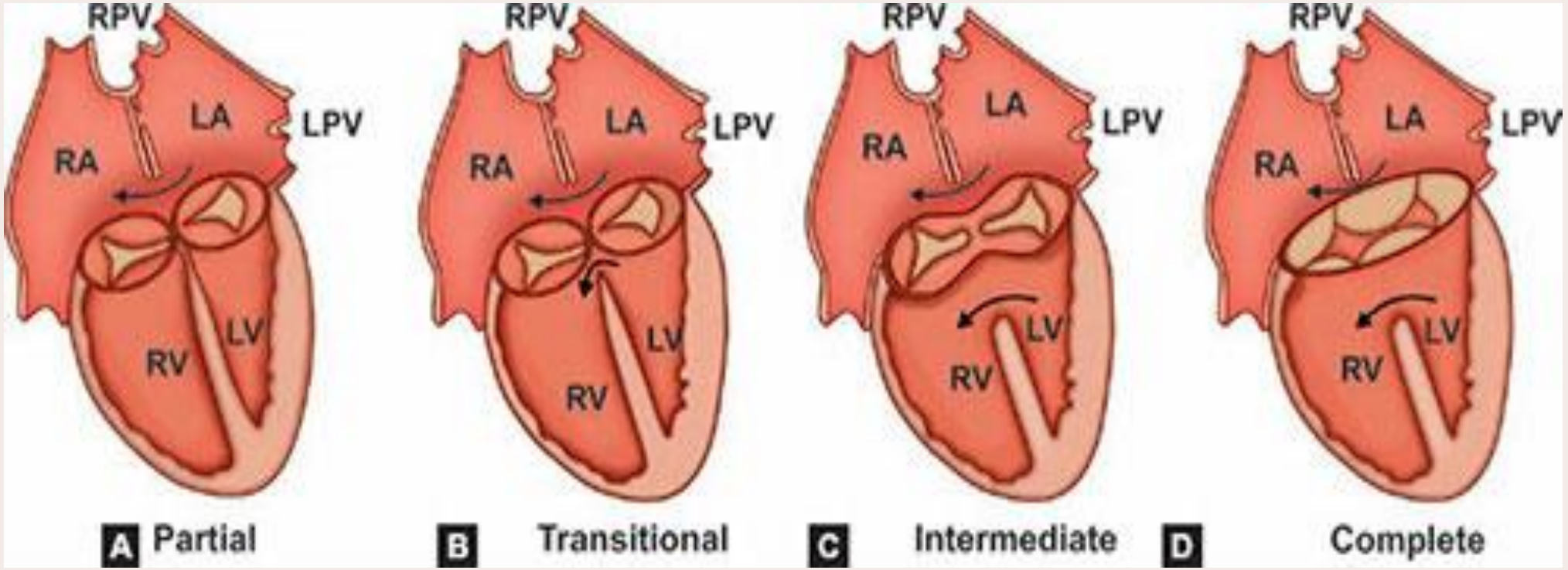
AVSD Types:

FIGURE 1 Classification of atrioventricular septal defect.



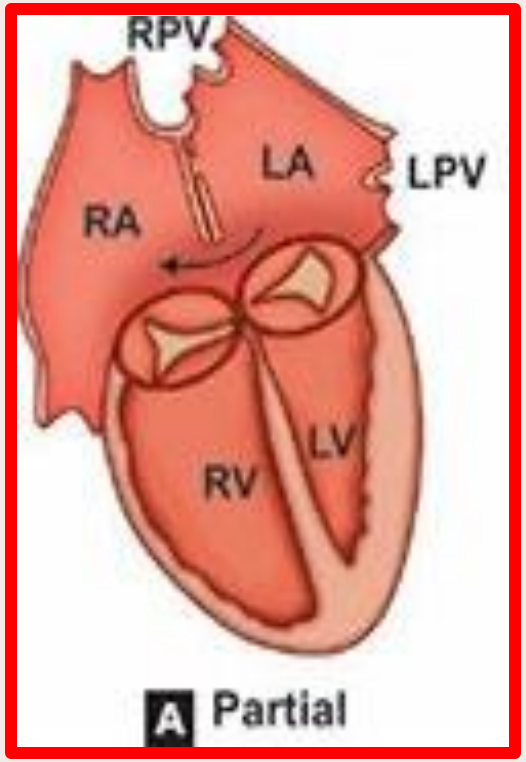
ASD = atrial septal defect; AVSD = atrioventricular septal defect: LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle; VSD = ventricular septal defect.

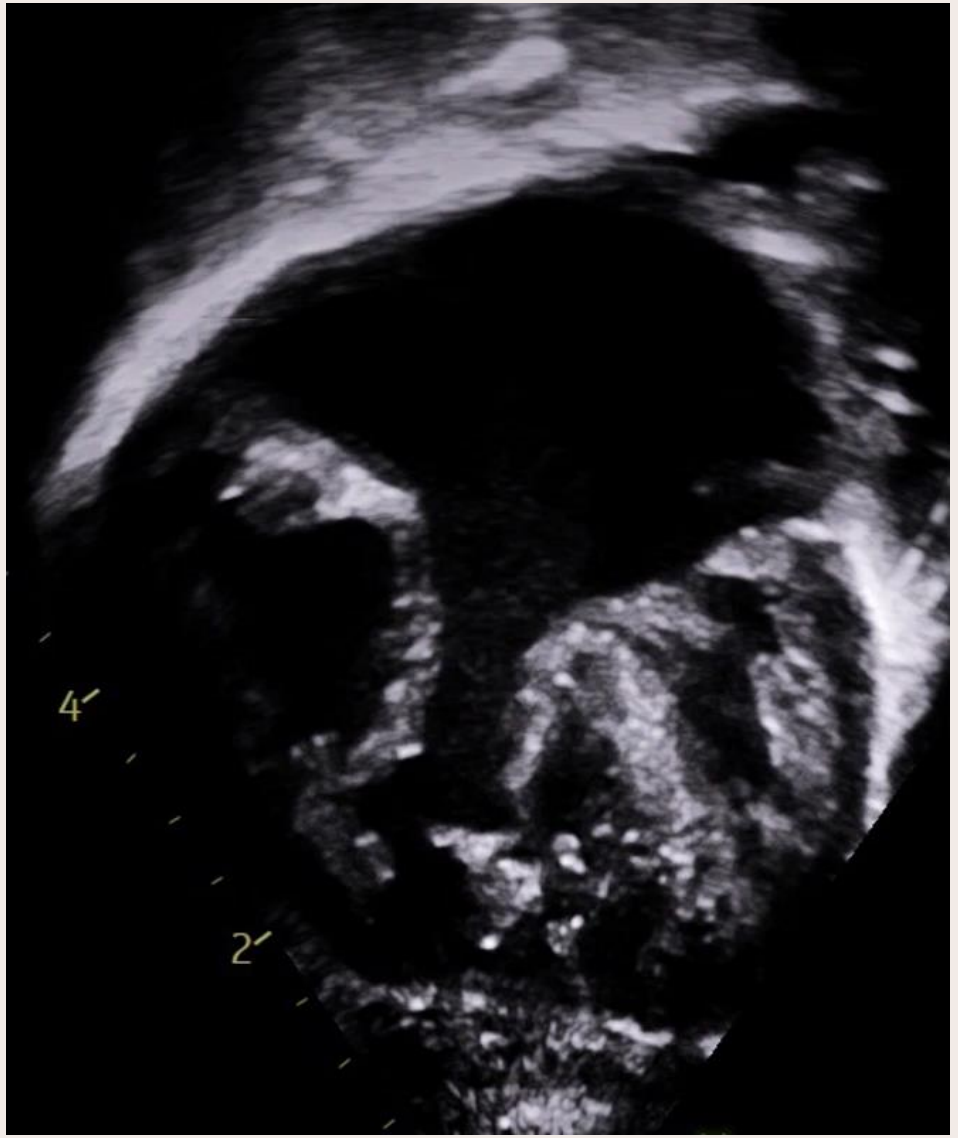
AVSD Types

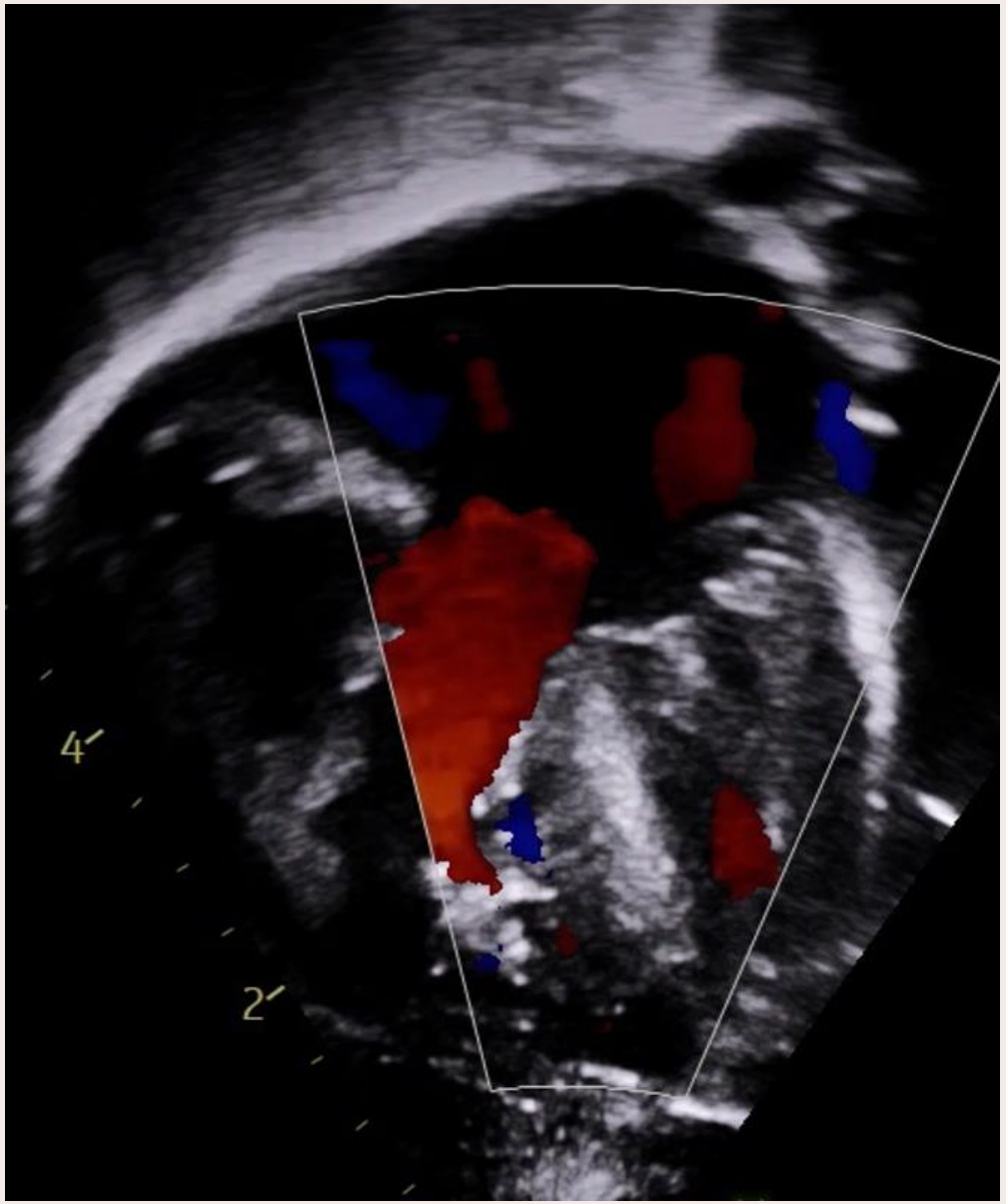


AVSD Types: Partial

- A congenital heart defect described by an atrial septal defect in the primum portion of the atrial septum, two separate atrioventricular (inlet valves) located at the same level and cleft mitral valve

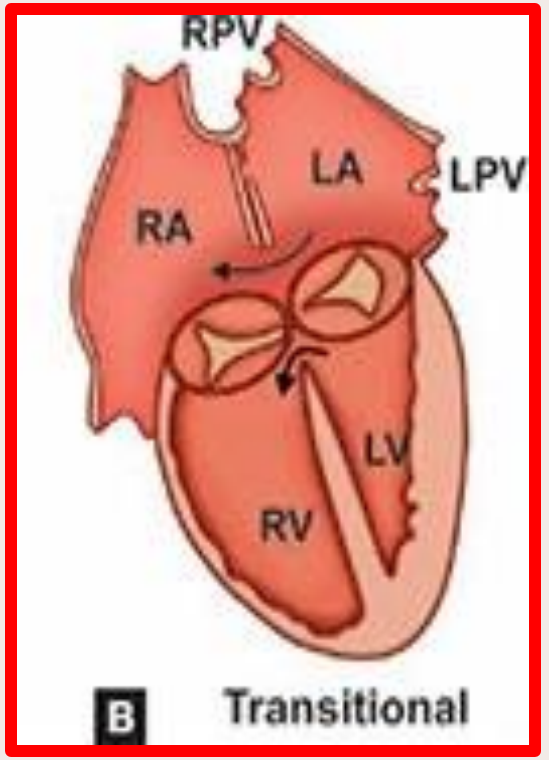






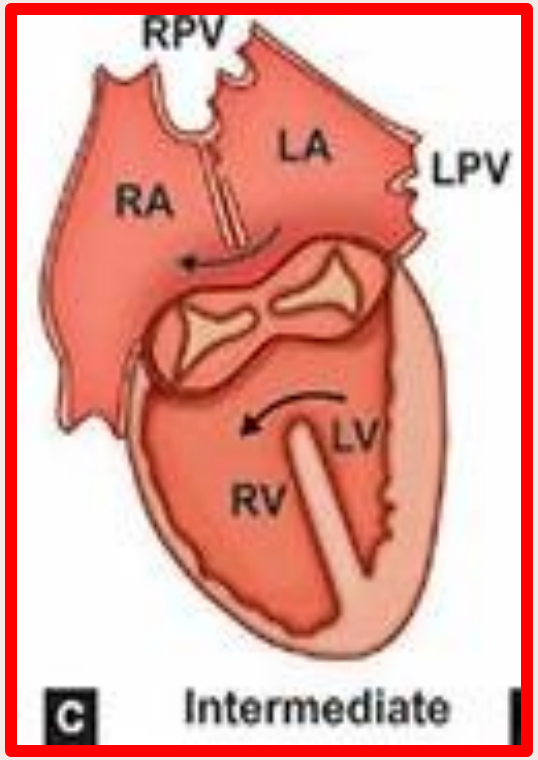
AVSD Types: Transitional

- A congenital heart defect described by an atrial septal defect in the primum portion of the atrial septum, two separate atrioventricular (inlet) valves located at the same level, cleft mitral valve and a small inlet VSD



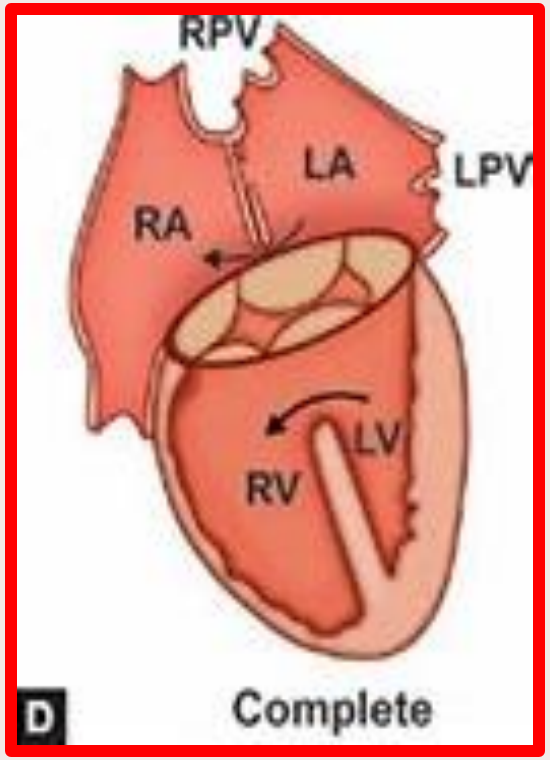
AVSD Types: Intermediate

- A rare congenital heart defect described by a mix of features found in partial and complete AVSD: primum ASD, larger VSD and common atrioventricular valve (AVV)

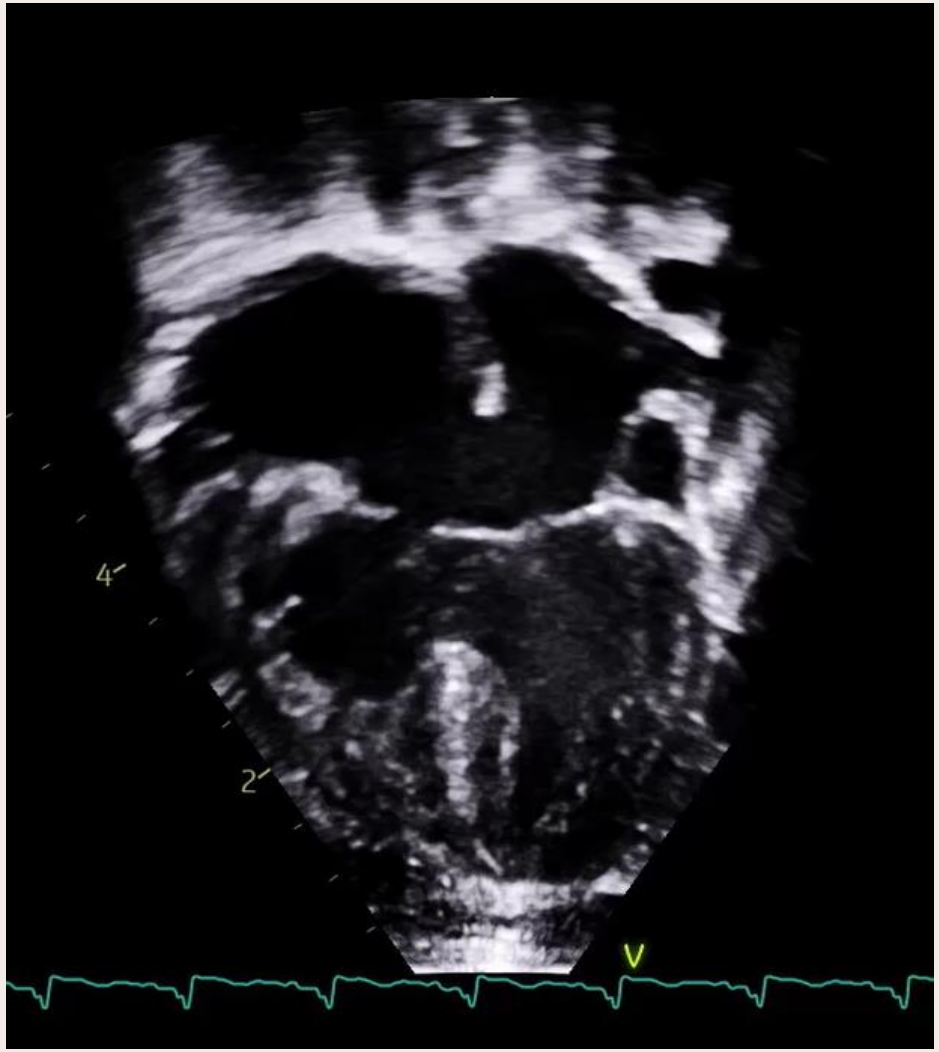


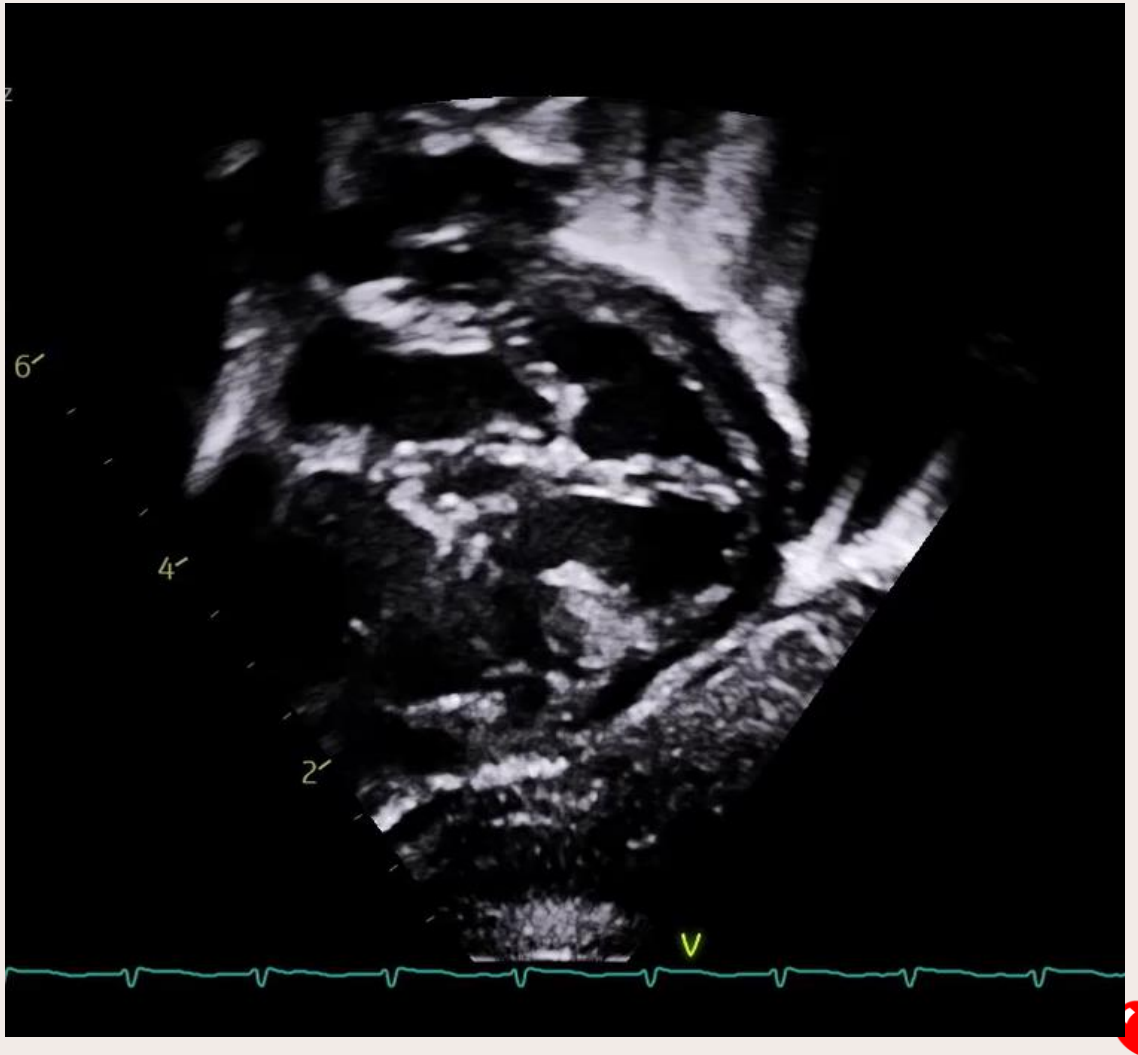
AVSD Types: Complete

- A congenital heart defect described by a mix of features found in partial and complete AVSD: primum ASD, large VSD and common atrioventricular valve (AVV)

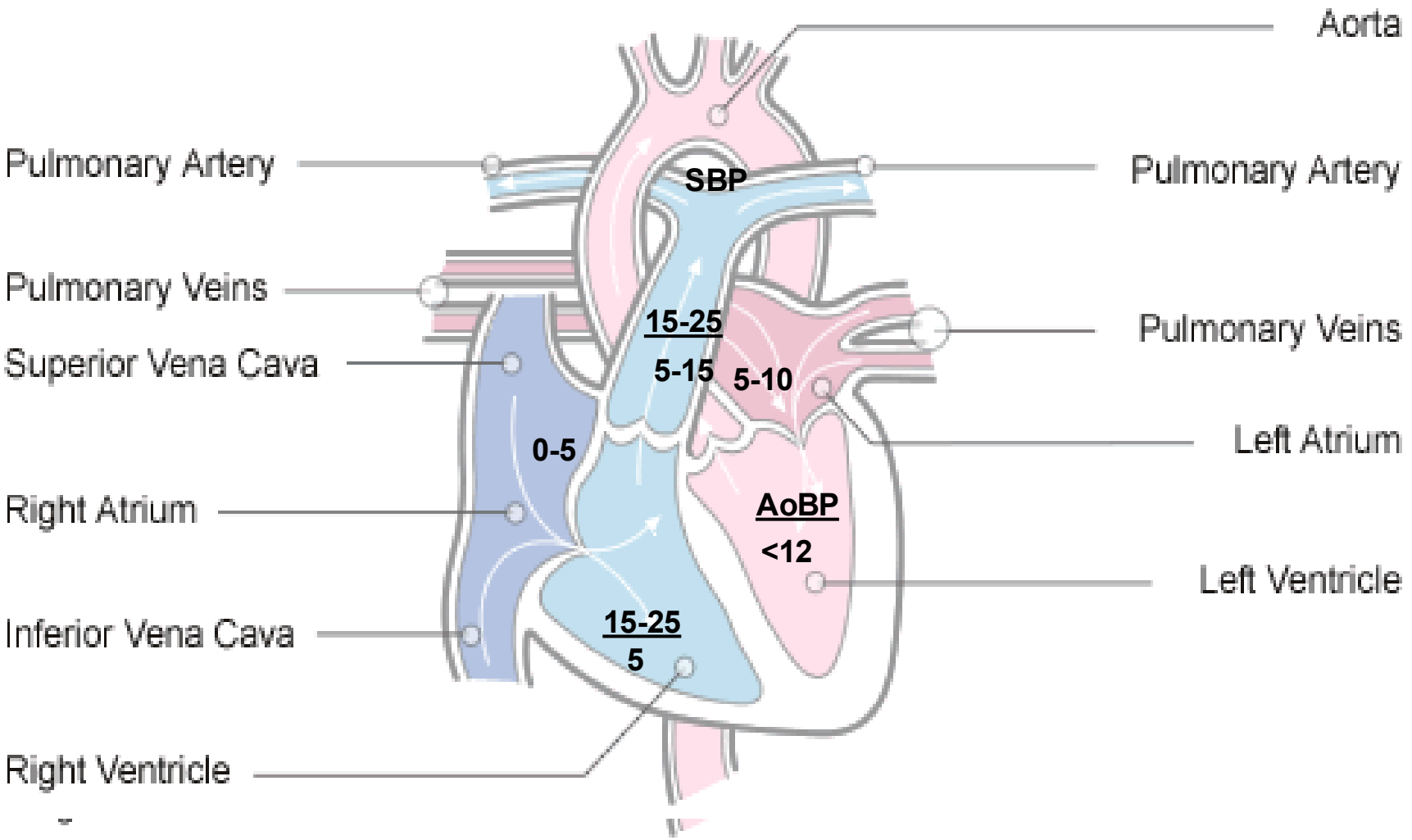


VSD Types: Inlet or Atrioventricular Canal

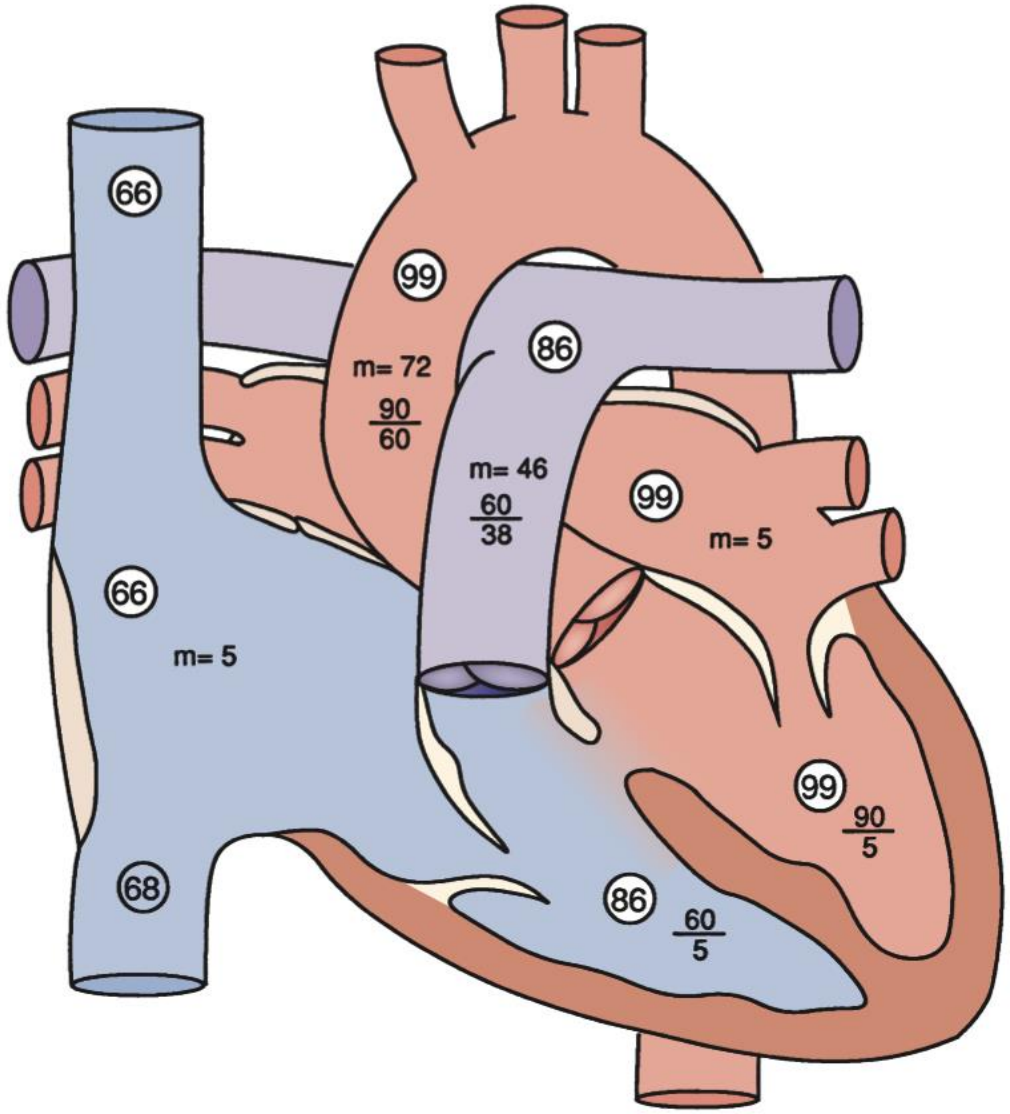




Normal Intracardiac Pressures



Pressure and oximetric changes in VSD



AVSD

- Hemodynamic alterations depend on:
  - ASD / VSD size. If significant ASD size, cyanosis can be present due to “mixing".
  - Competency of valves (regurgitation)
  - PVR – Pulmonary Vascular Resistance
    - Newborn – PVR increased so minimal shunting of blood to right side of body
    - 2 – 3 months resistance falls – Left to right shunting increases so pulmonary blood flow increases

AVSD: Diagnostics

- Echo
  - Primary diagnostic tool
- CXR
  - Cardiomegaly
- EKG
  - Prolonged PR interval
  - RVH

AVSD: Management

- Medications
  - Digoxin/Diuretics/ACE inhibitors
- Nutrition
  - Increased caloric density
  - Feeding tube
- Respiratory support
- Surgical repair

Down Syndrome

- ~ 50% have CHD
  - 40% are AVC
- Higher ratio of PVR: SVR
- May develop PH quicker
- Chronic nasopharyngeal obstruction
- Hypoventilation
- Sleep apnea

- More competent MV/LAVV
- Additional septal material

AV Canal Surgical Repair

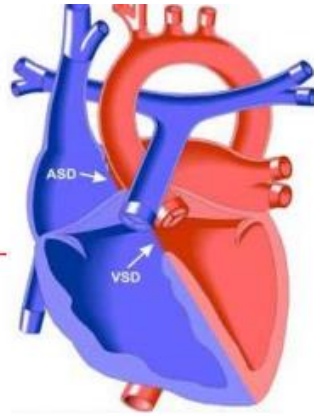
Timing of surgical repair:

- Size of infant
- Amount of CHF and pulmonary overload
- Typically 3-8 months

Surgery:

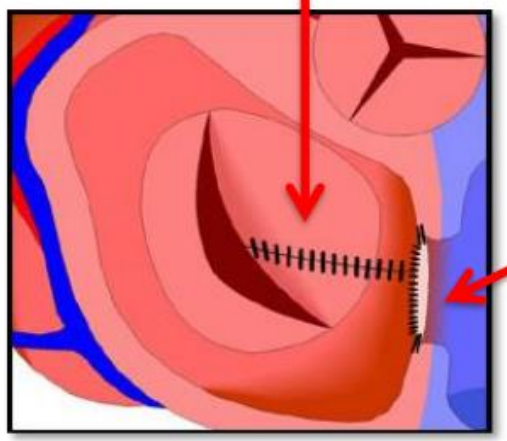
- Closure of ASD
- Patch closure of VSD
- Valve repair and division (determined by number of leaflets on common valve)
- Extent of valve involvement crucial

Atrioventricular Canal
Surgical Management

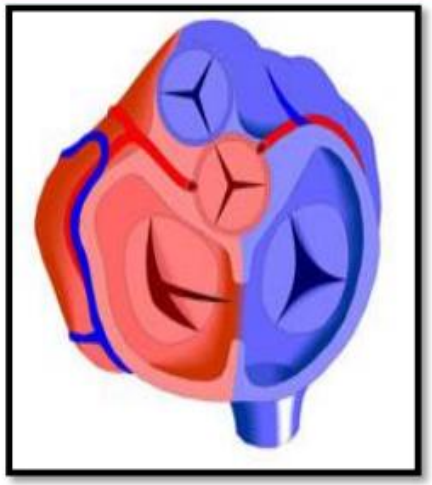


Partial / Transitional AVC

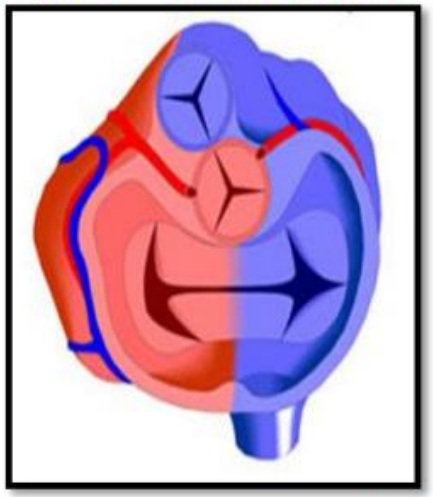
Suture repair of mitral valve cleft



ASD Patch



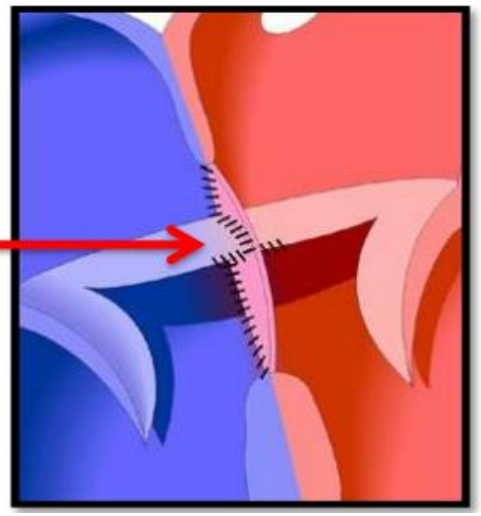
Complete Balanced AVC



Patch septal defects

Reconstruction of AV valves

- L AVV
- R AVV



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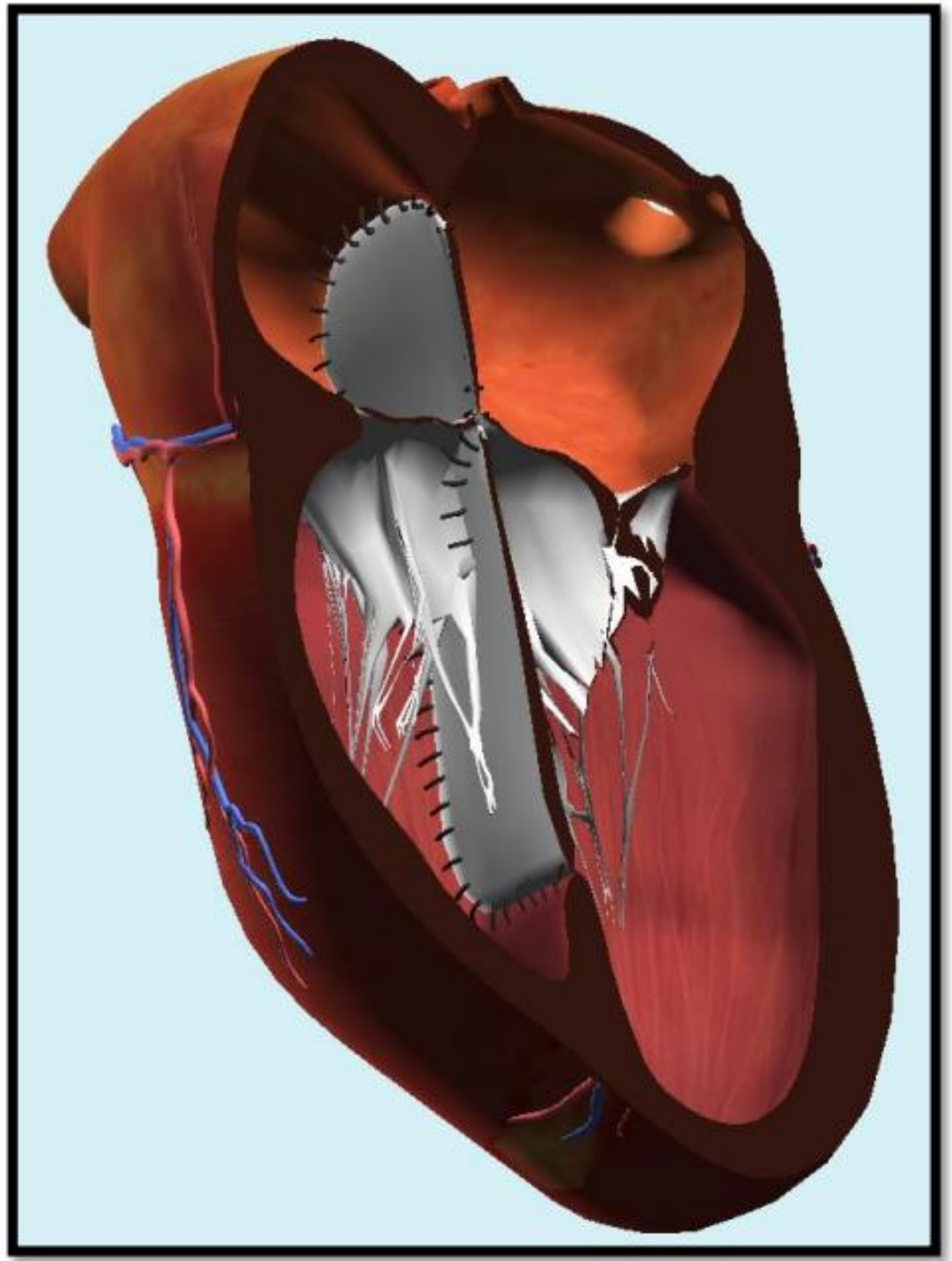
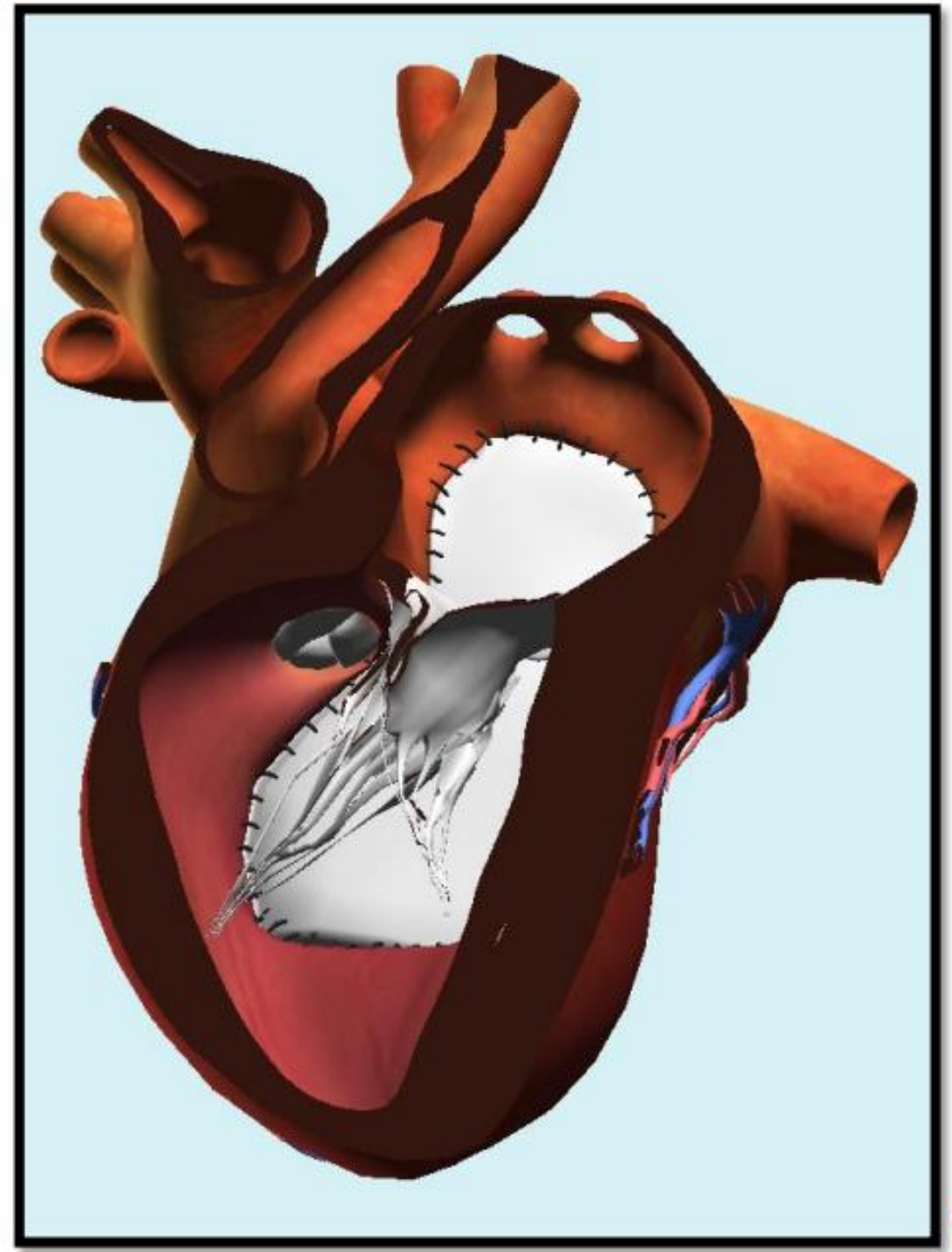
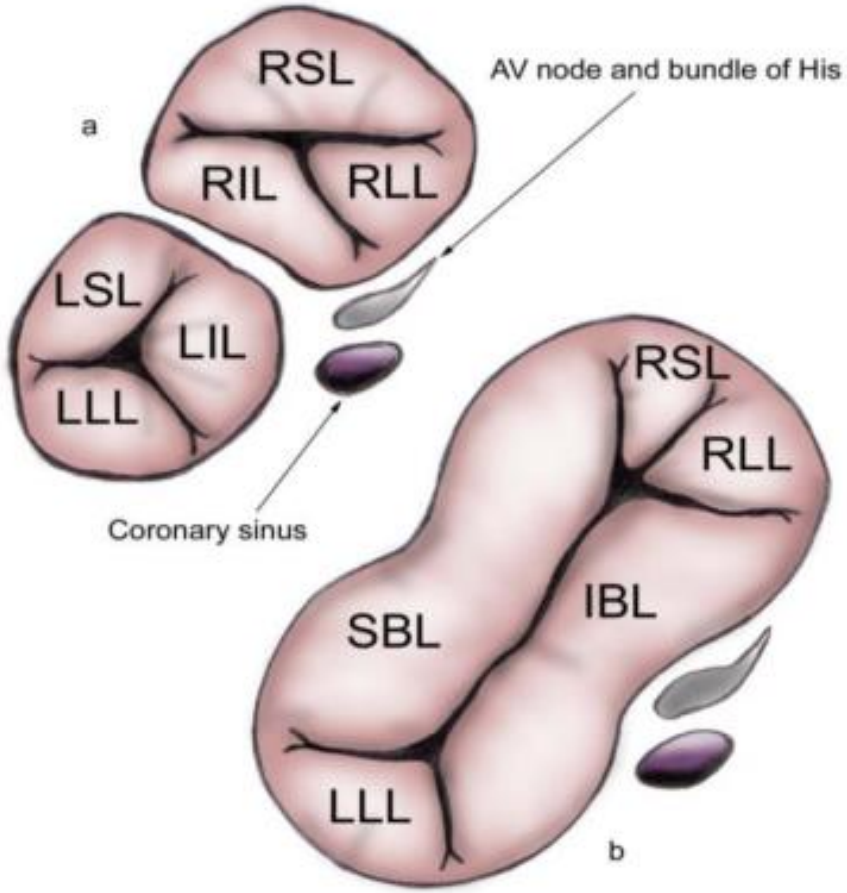




AVSD: Post Operative Concerns

- Low cardiac output
- Arrhythmias
  - Heart block
  - Junctional ectopic tachycardia
- AVV regurgitation
- Pulmonary artery hypertension
- Congestive heart failure



AVSD: Management cont..

- Vasoactive support
- Afterload reduction
- Steroids- refractory hypotension
- Diuretics

AVSD: Outcomes

- Morbidity
  - 25% await reoperation
- Mortality
  - Uncomplicated repair <2%
  - Significant MR- 4%
  - Highly inconsistent
  - PAH- affects repair after 6mo (earlier in Down syndrome)

Case Study cont..

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Questions:

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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


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