Congenital Heart Disease

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INTRODUCTION

- CHD ~ 0.8% of live births.
- Major CHD:
  - Ventricular Septal Defect: 35%
  - Atrial Septal Defect: 7 %
  - Patent Ductus Ateriosus: 7 %
  - Coarctation of Aorta: 6 %
  - Tetralogy of Fallot: 6 %
  - Pulmonary valve stenosis: 6 %
  - Aortic valve stenosis: 5 %
  - D-Transposition of great arteries: 4 %

Congenital Heart Disease

- Etiology: Mostly unknown
- Chromosomal abnormalities can cause
  - Trisomy 21: AVSD
  - Trisomy 18: VSD
  - Trismoy 13: PDA, VSD, ASD
  - DiGeorge Syndrome: Arch, Conotruncal abnormalities
  - Turner syndrome: Coarctation of Aorta
  - Williams Syndrome: Supra-aortic stenosis, PA stenosis
  - Noonan Syndrome: Dysplastic pulmonary valve

Classification of CHD

- Divided into 2 major groups:
  - Cyanotic heart diseases.
  - Acyanotic heart diseases.
- Subdivided further according to:
  - Physical Finding
  - Chest X-ray finding
  - ECG finding
- Diagnosis is confirmed by:
  - Echo, Cardiac CT/MRI or Cardiac Catheterization.

Classification of CHD

Cyanotic Heart Disease

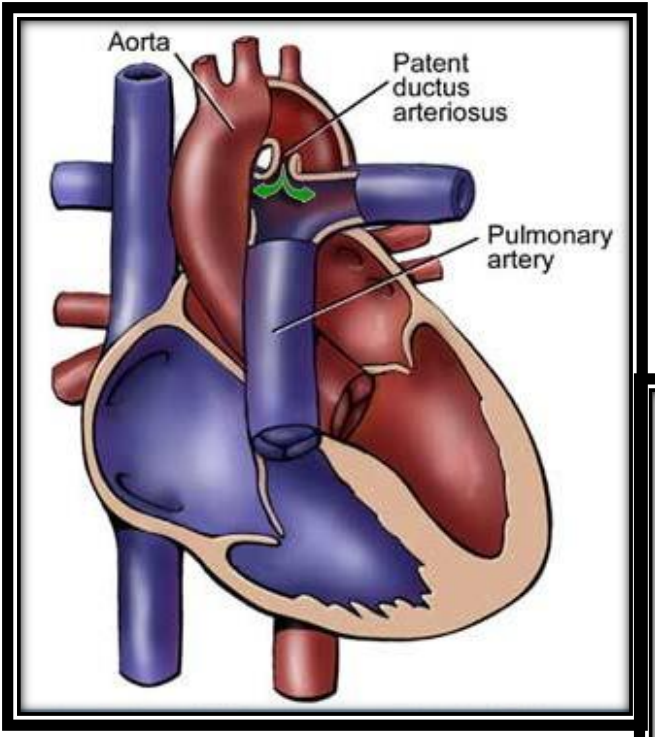
- Decreased pulmonary flow:
  - Tetralogy of Fallot
  - Tricuspid atresia
  - Other univentricular heart with pulmonary stenosis.
- Increased pulmonary flow:
  - Transposition of great arteries
  - Total anomalous pulmonary venous return.

Acyanotic Heart Disease

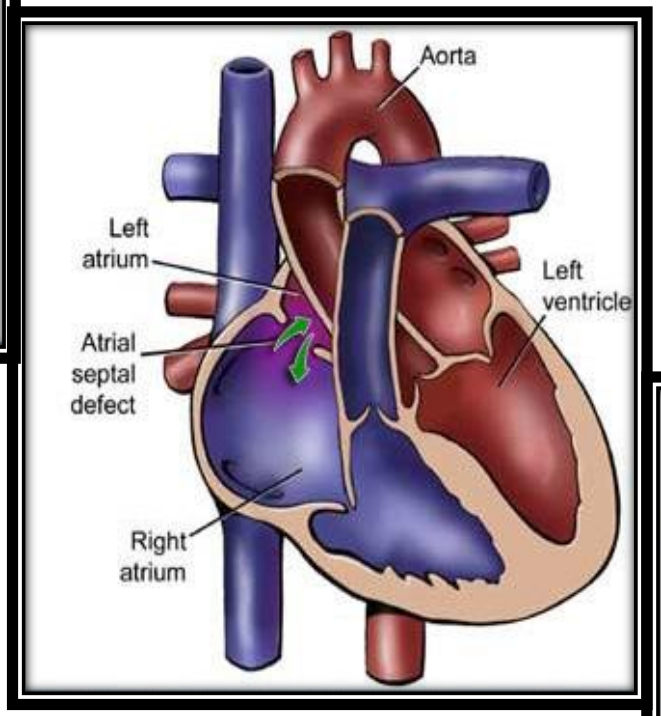
- Left – Right shunt lesions:
  - Ventricular septal defect
  - Atrial Septal Defect
  - Atrio-ventricular Septal Defect
  - Patent Ductus Arteriosus
- Obstructive lesions:
  - Aortic stenosis
  - Pulmonary valve stenosis
  - Coarctation of Aorta

Acyanotic Heart Disease
Left – to- Right Shunt lesions

Left – to- Right Shunt lesions

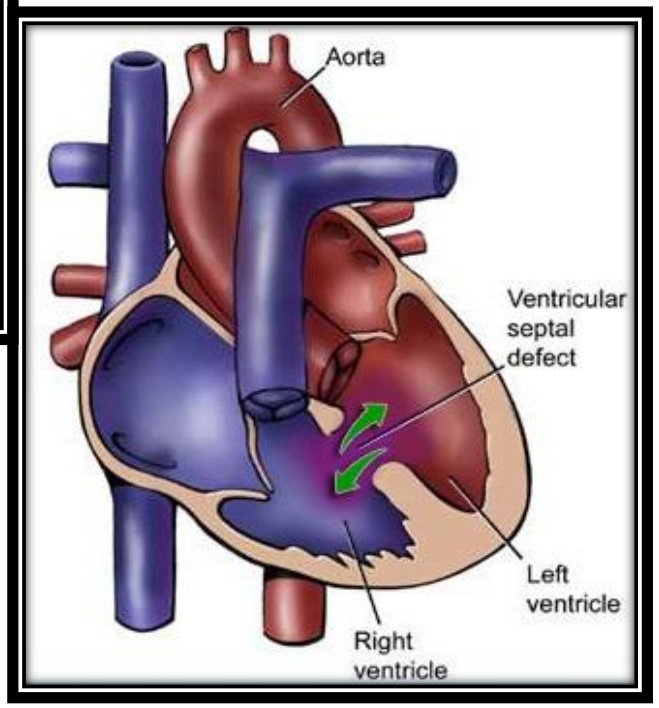


PDA

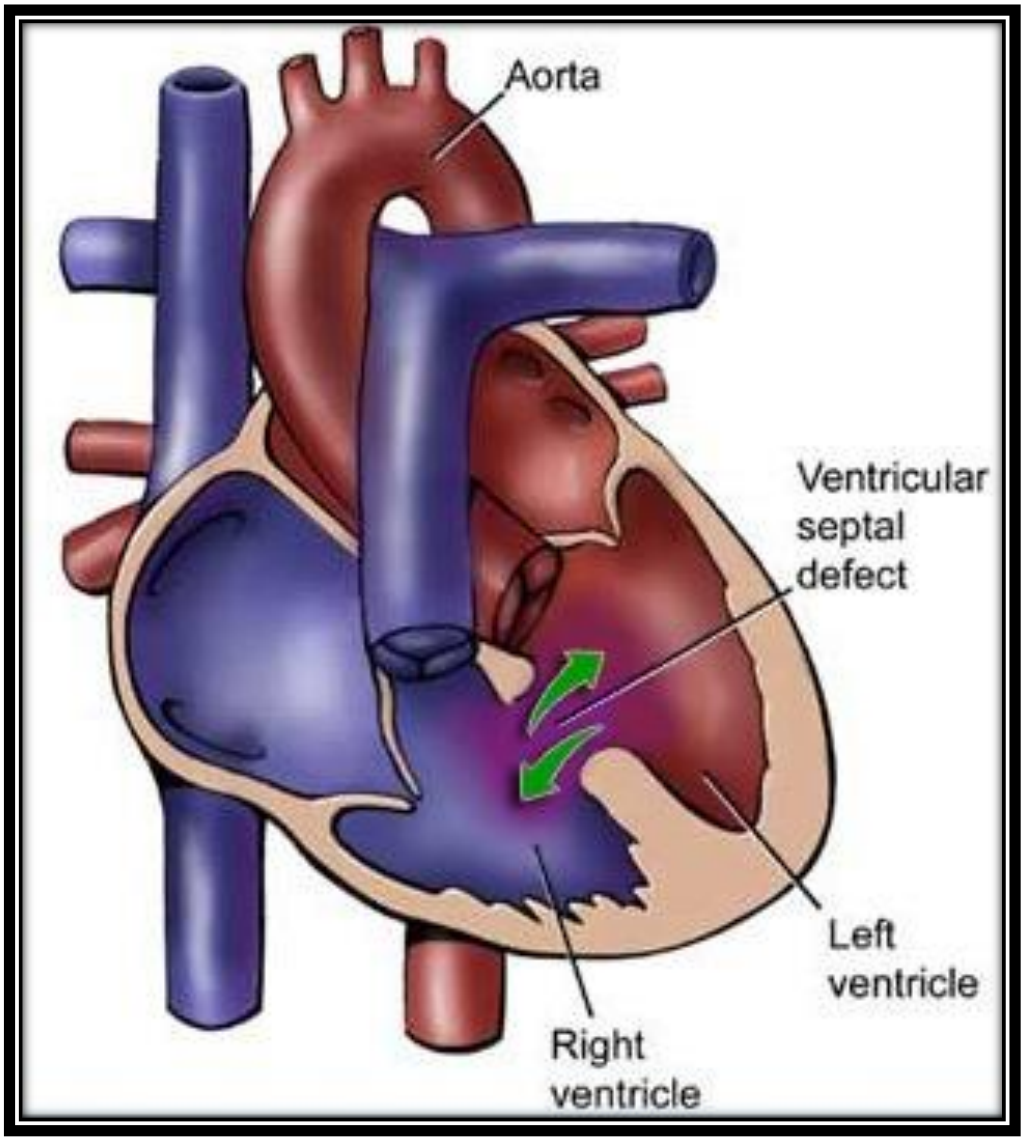


ASD

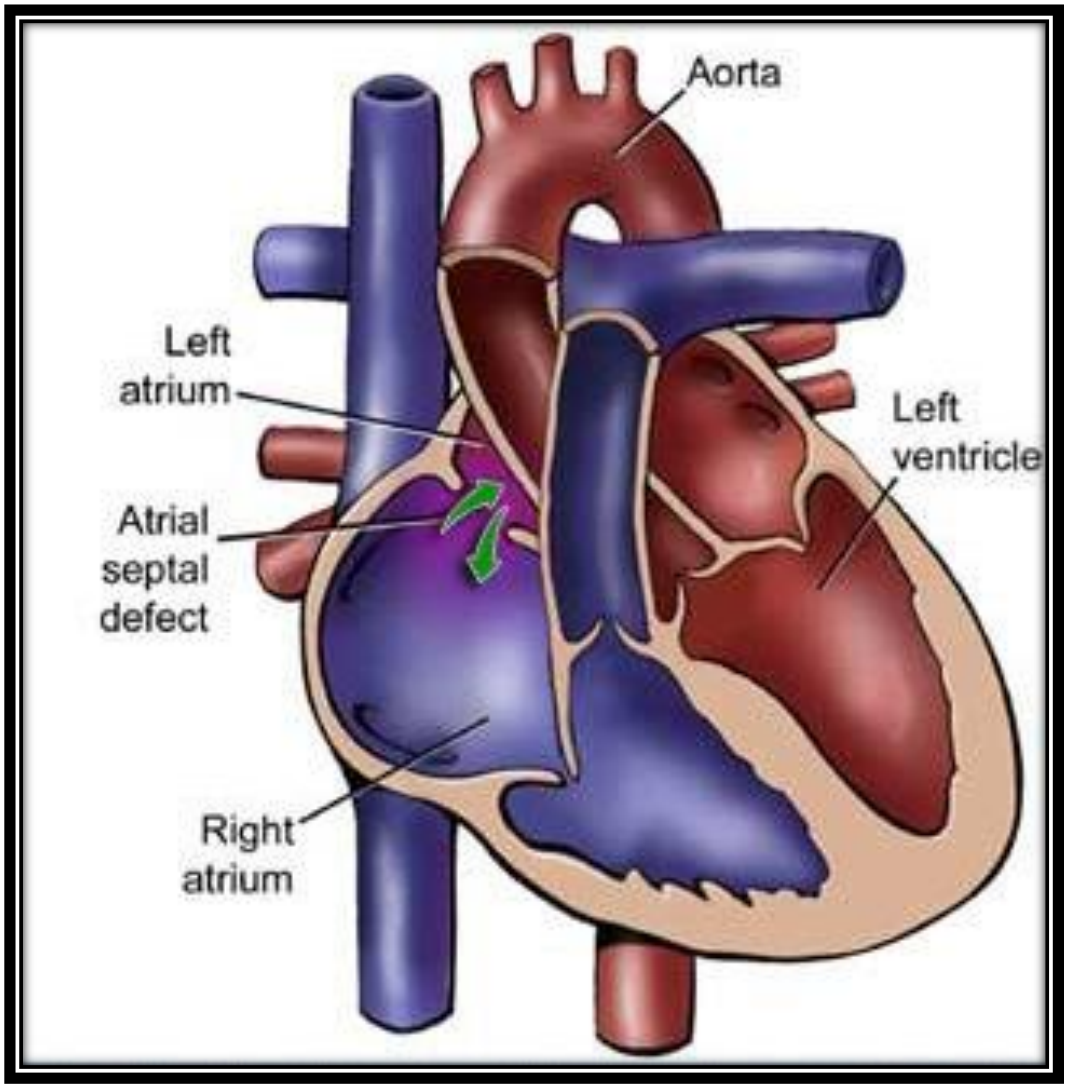
VSD



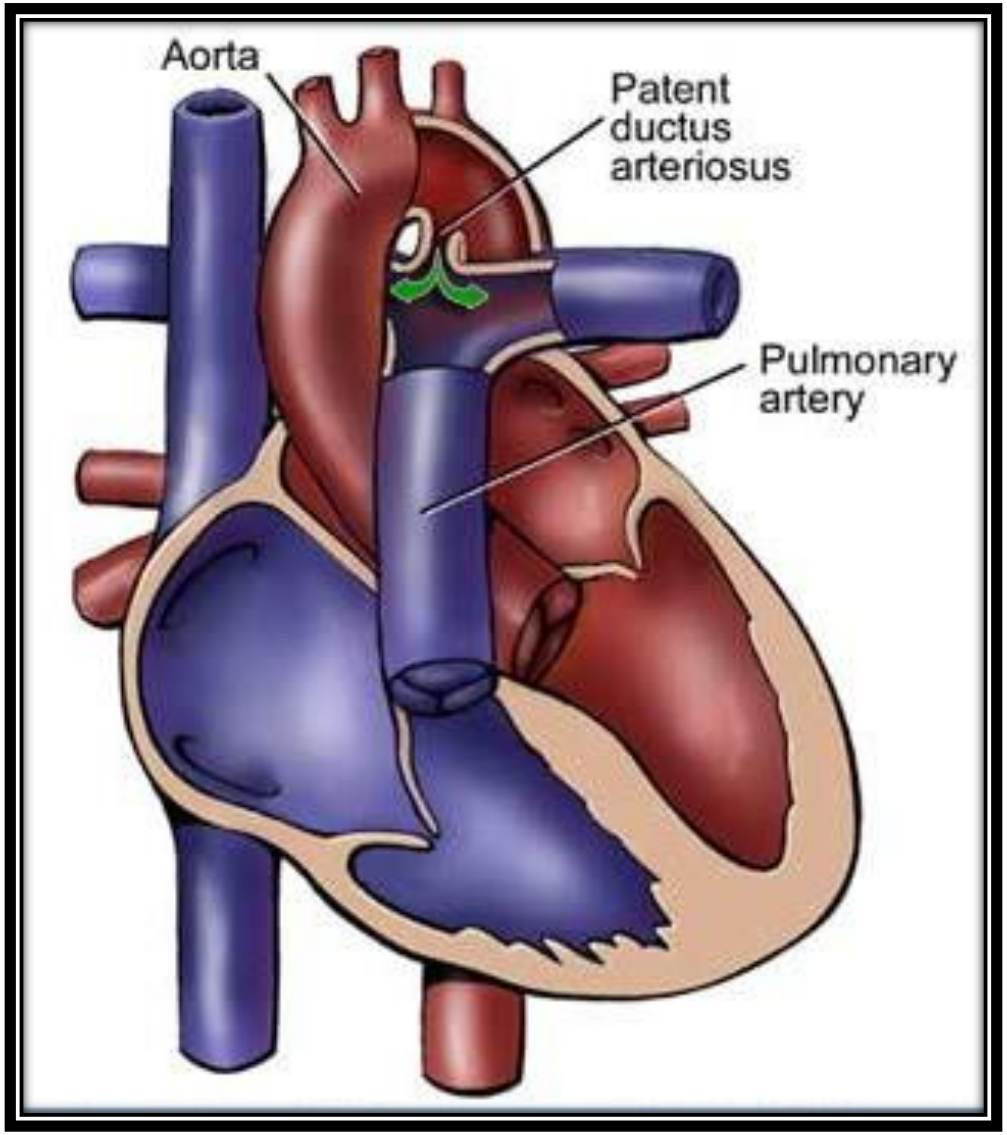
VSD


Aorta
Ventricular septal defect
Left ventricle
Right ventricle

ASD


Aorta
Left atrium
Left ventricle
Atrial septal defect
Right atrium

PDA

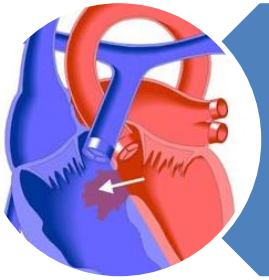


PATHOPHYSIOLOGY: L-R SHUNT

- L-R shunt toward pulmonary circulation.
  - Increased Qp:Qs ratio
    - Increased cardiac output to the pulmonary circulation (Qp)
    - Reduced of cardiac output to the systemic circulation (Qs)

PATHOPHYSIOLOGY: L-R SHUNT

VSD

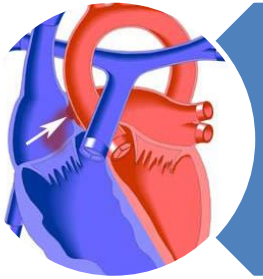


L-R shunt at ventricular level:

Dilated LA and LV

Enlarged pulmonary arteries

ASD

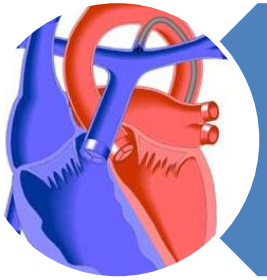


L-R shunt at atrial level:

Dilated RA and RV

Enlarged pulmonary arteries

PDA

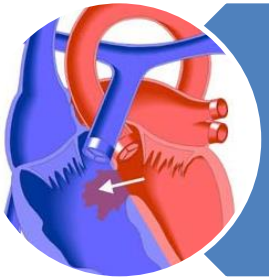


L-R shunt at artery level:

Dilated LA and LV

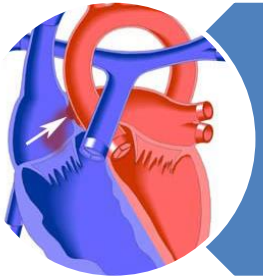
SYMPTOM: L-R SHUNT

VSD



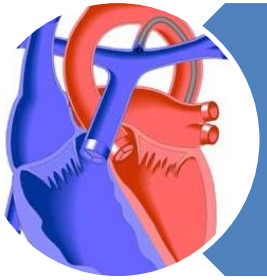
- Small VSD: Asymptomatic
- Moderate to large VSD: CHF

ASD



- Usually asymptomatic
- Older children: Activity related SOB & Fatigability
- Rare: CHF , FTT

PDA



- Small PDA: Asymptomatic
- Moderate to large PDA: CHF

CONGESTIVE HEART FAILURE

SYMPTOMS	SIGNS
• Diaphoresis • Poor feeding • Failure to thrive • Shortness of breath • Recurrent chest infection • Exercise intolerance	• Tachypnia • Tachycardia • Cardiomegally • Hepatomegally • Active precordium

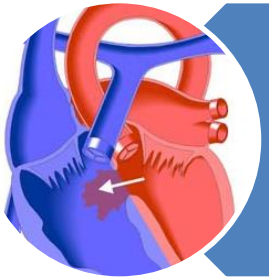
VSD

- No symptom during neonatal period
    - due to high pulmonary vascular resistance

- Symptoms of CHF started ~ 2/12 of age
  - diaphoresis, poor feeding, and failure to thrive.
  - shortness of breath, recurrent chest infection.
  - exercise intolerance.

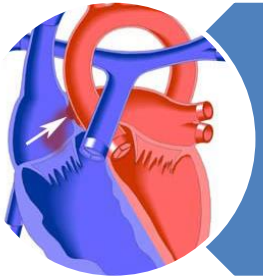
EXAMINATION: L-R SHUNT

VSD



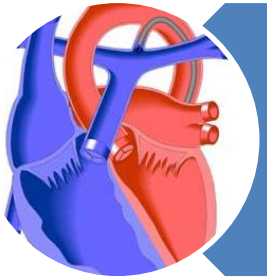
- Holosystolic murmur
- Large: mid-diastolic murmur
- Small muscular: ejection systolic murmur

ASD



- Fixed Widely splitted second heart sound
- Ejection systolic murmur
- Large: mid-diastolic murmur

PDA



- Small PDA: Silent
- Large PDA: Continuous “machinery” murmur
- Large PDA: Widened pulse pressure

INVESTIGATION: L-R SHUNT

- Diagnosis:
  - Chest X-ray:
    - Increased pulmonary vascular marking
    - +/- cardiomegally
  - ECG:
    - Small lesion: Normal
    - Mod to large: chambers enlargement
  - ECHO:
    - Confirm Diagnosis
  - Cardiac Cath: not required for Dx

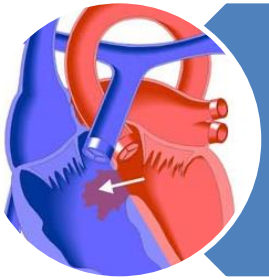
MEDICAL Rx: L-R SHUNT

Medical Mgx

- Anti-congestive therapy:
  - Diuretics
  - Digoxin.
  - After load reducing agents
- Nutritional support
- ASD: usually no medication needed

INTERVENSION: L-R SHUNT

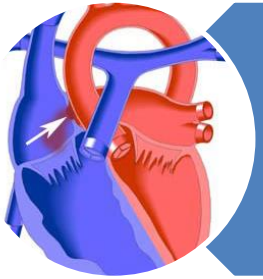
VSD



Surgical closure 4-8 months

Some types in older children can be closed by device via CATH.

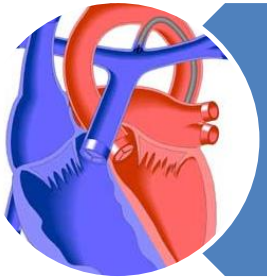
ASD



Most closed by device via CATH around 3-6 years

Some types need surgical closure.

PDA



Most closed by device via CATH

Surgery needed in premature baby and symptomtic neonate less than 6 kg.

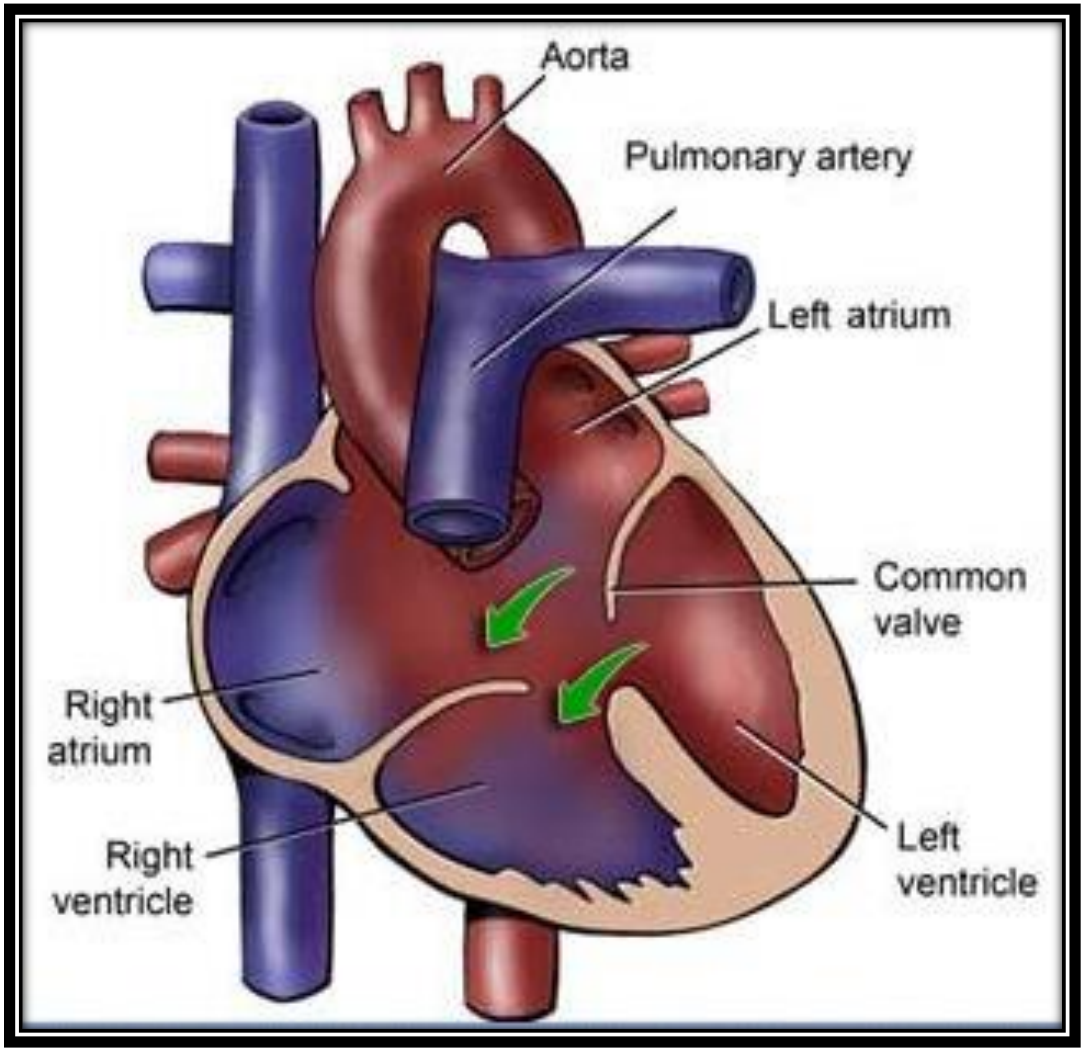
PROGNOSIS: L-R SHUNT

- Untreated VSD and PDA beyond infancy:
  - Eisenmenger's syndrome
    - Sign and symptom of CHF will disappear
    - Patient will become cyanotic
      - R-L shunt

PROGNOSIS: L-R SHUNT

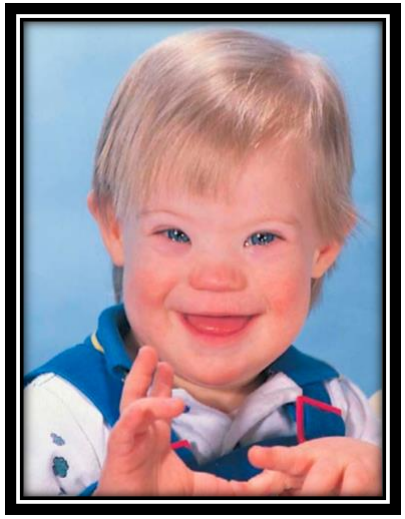
- Untreated ASD:
  - complication happened during adult Life:
    - Eisenmenger's syndrome
    - Atrial arrhythmias
    - Paradoxical embolism (rare)

AVSD


Aorta
Pulmonary artery
Left atrium
Common valve
Right atrium
Right ventricle
Left ventricle

AVSD

- Incidence: 4 % of all CHD
  - Associated with Down Syndrome (50%)
- Divided into:
  - Complete AVSD
    - ASD primum/ inlet VSD / common AV valve
    - Balanced vs. Unbalanced AVSD
  - Partial AVSD
    - ASD primum
    - No VSD



AVSD

- Pathophysiology:
  - Similar to VSD and ASD
    - left to right shunt across the atrial level
    - Left to right shunt at and ventricular level
    - In addition: AV valve regurgitation
  - Significant L-R shunting:
    - Pulmonary over-circulation
    - Increase Qp:Qs ratio.

AVSD

- Clinical Features:
  - Usually asymptomatic at neonatal period
    - Due to high pulmonary vascular resistance
    - Baby may have slightly lower oxygen saturation
  - Symptoms of CHF started at few months of age
    - Diaphoresis
    - Poor feeding
    - Failure to thrive.
    - Shortness of breath
    - Recurrent chest infection.
    - Exercise intolerance.

AVSD

- Clinical Features:
  - Physical Examination:
    - Feature of Down Syndrome
    - Tachypnia
    - Tachycardia
    - Active precordium
    - Murmur:
    - Pan-systolic (holosystolic) murmur
    - Hepatomegaly

AVSD

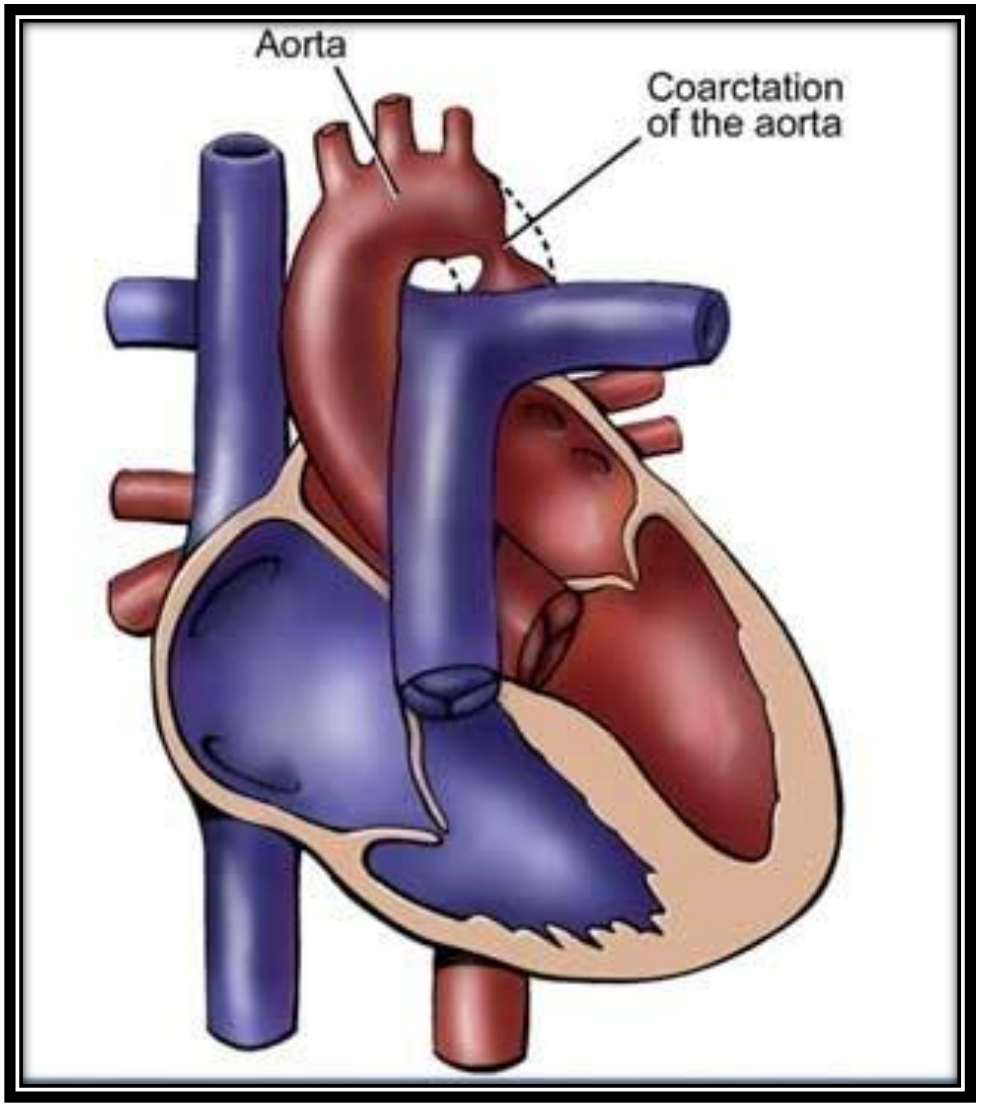
- Diagnosis:
  - Chest X-ray:
    - Increased pulmonary vascular marking
    - Cardiomegaly
  - ECG:
    - Left Axis deviation with RVH is very suggestive of AVSD
  - ECHO:
    - Confirm Diagnosis
  - Cardiac Cath: not required for Dx

AVSD

- Treatment:
  - Medical Rx:
    - Anti-congestive therapy:
    - Nutritional suppor
  - Surgical closure for complete VSD:
    - Usually done before 6 months of age to ovoid development of Eisenmenger's syndrome.
      - Balanced AVSD: Biventricular repair
      - Unbalanced AVSD: may need single ventricular repair

Acyanotic Heart Disease obstructive lesion

Coarctation of Aorta (CoA)



Coarctation of Aorta (CoA)

- Incidence: 5-7 % of all CHD
  - Associated with Turner syndrome in female
  - Arch interruption: seen in DiGeorge syndrom
- Can be: Discrete or Diffuse
- Can be mild to severe

PATHOPHYSIOLOGY: CoA

CRTICAL CoA

- Spontaneous PDA closure
  - Obstruction of blood flow to distal arch
  - Hypotension and Shock
  - Acute increase of LV after-load
  - LV dysfunction
- "DUCT DEPENDENT CHD"

MILD CoA

- Collateral vessels develop overtime
- Flow maintained between proximal and distal aorta
- Present later on life

CLINICAL PRESENTATION: CoA

CRTICAL CoA

- Presented 2-3 wks of life
  - Sign of CHF
  - Circulatory collapse
  - Shock
  - Death
- **"DUCT DEPENDENT CHD"**

MILD CoA

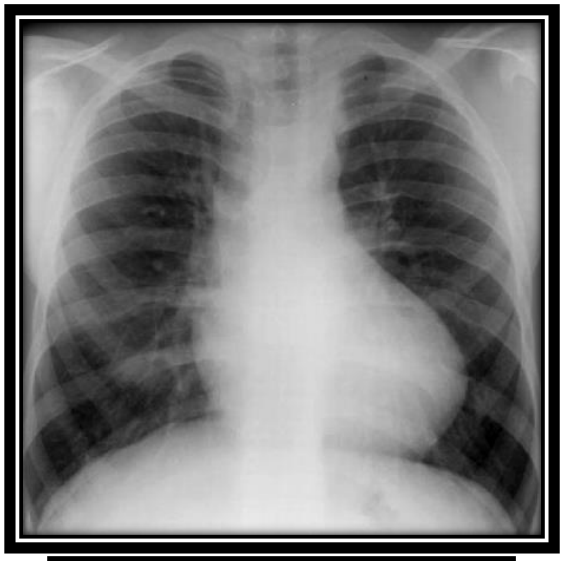
- Present later on life
  - Murmur
  - Chronic hypertension
  - Headache
  - Fatigue
  - Stroke
    - » Rupture cerebral aneurysm

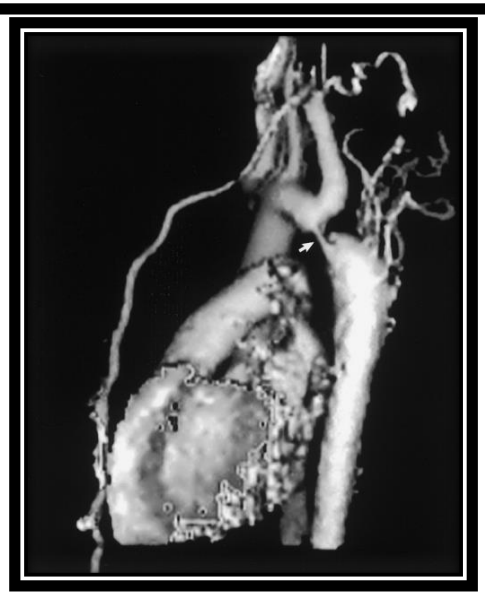
Clinical Features: CoA

- Physical Examination:
  - Differential cyanosis (severe CoA in newborn)
  - Signs of cardiac shock
  - Reduced or absent femoral pulses
  - Brachio- femoral delay
  - BP in lower limb lower than upper Limb BP
  - Murmur:
    - Ejection systolic murmur at the back
    - Continues murmur due to collateral at the back

DIAGNOSIS: CoA

- Chest X-ray:
  - Cardiomegaly
  - Prominent aortic knob
  - Rib notching
    - » Due to d of collateral vessels
    - » Rarely seen before age of 10 years
- ECG:
  - Neonate: RV hypertrophy
  - Older children: LV hypertrophy
- ECHO:
  - Usually will establish the diagnosis
- May need Cardiac CT /MRI



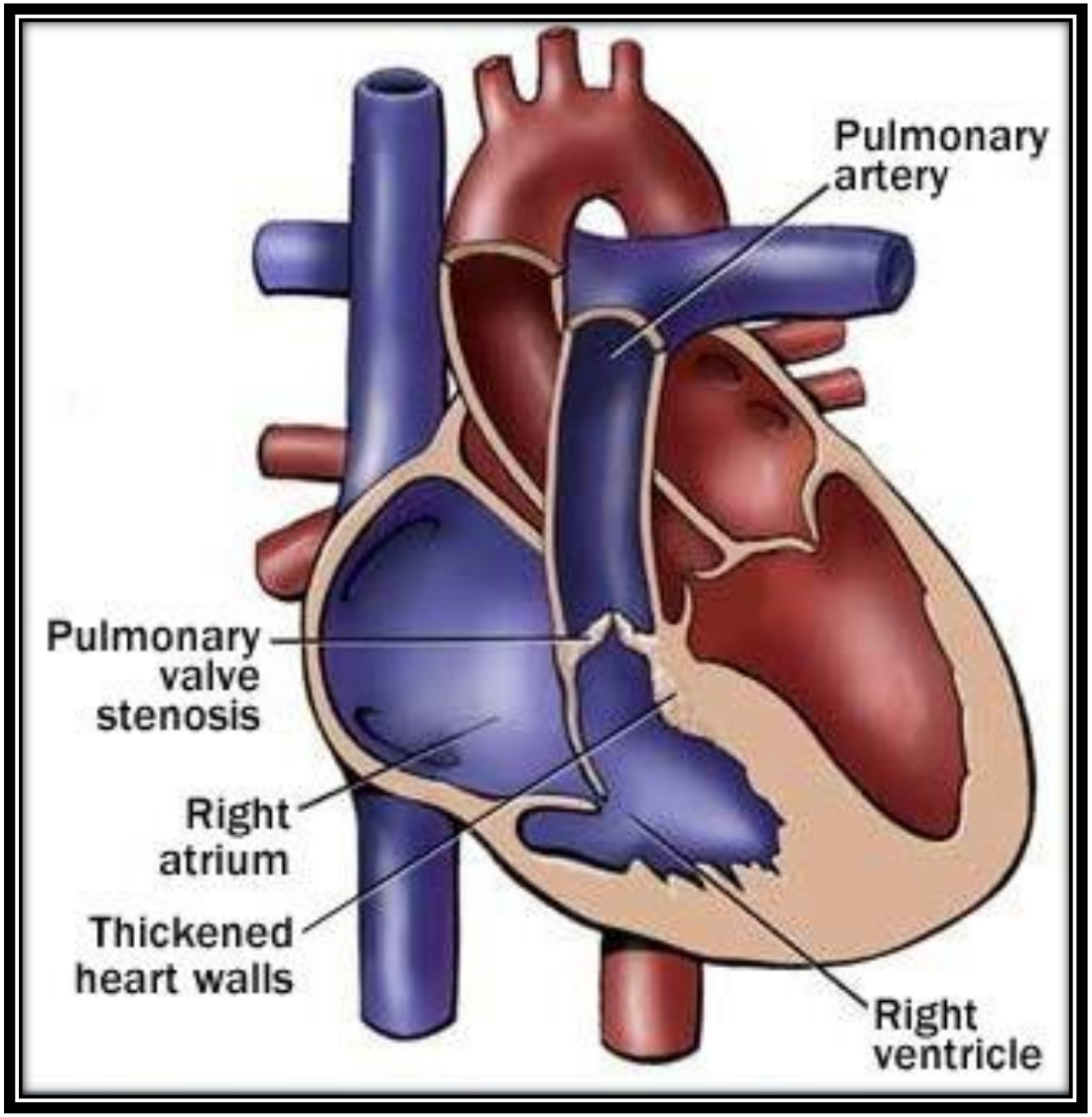


TREATMENT: CoA

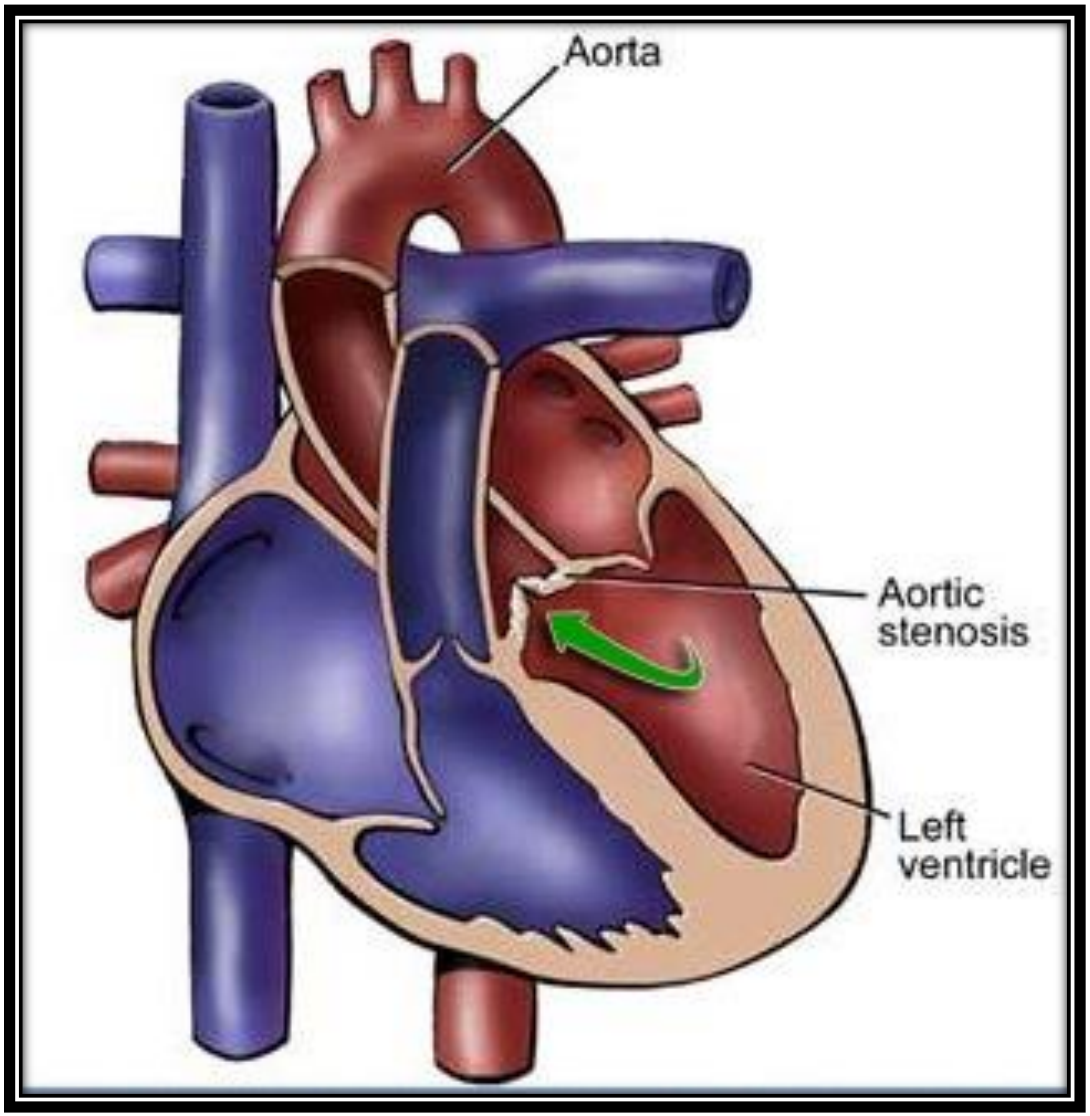
- Critical CoA
  - "Duct Dependent CHD"
    - Prostaglandin E2 to keep PDA open
- Surgery is the primary intervension

- Trans-catheter balloon angioplasty +/- stent:
  - Recurrent CoA
  - Primary intervention: Discrete CoA in older children

Pulmonary Valve Stenosis

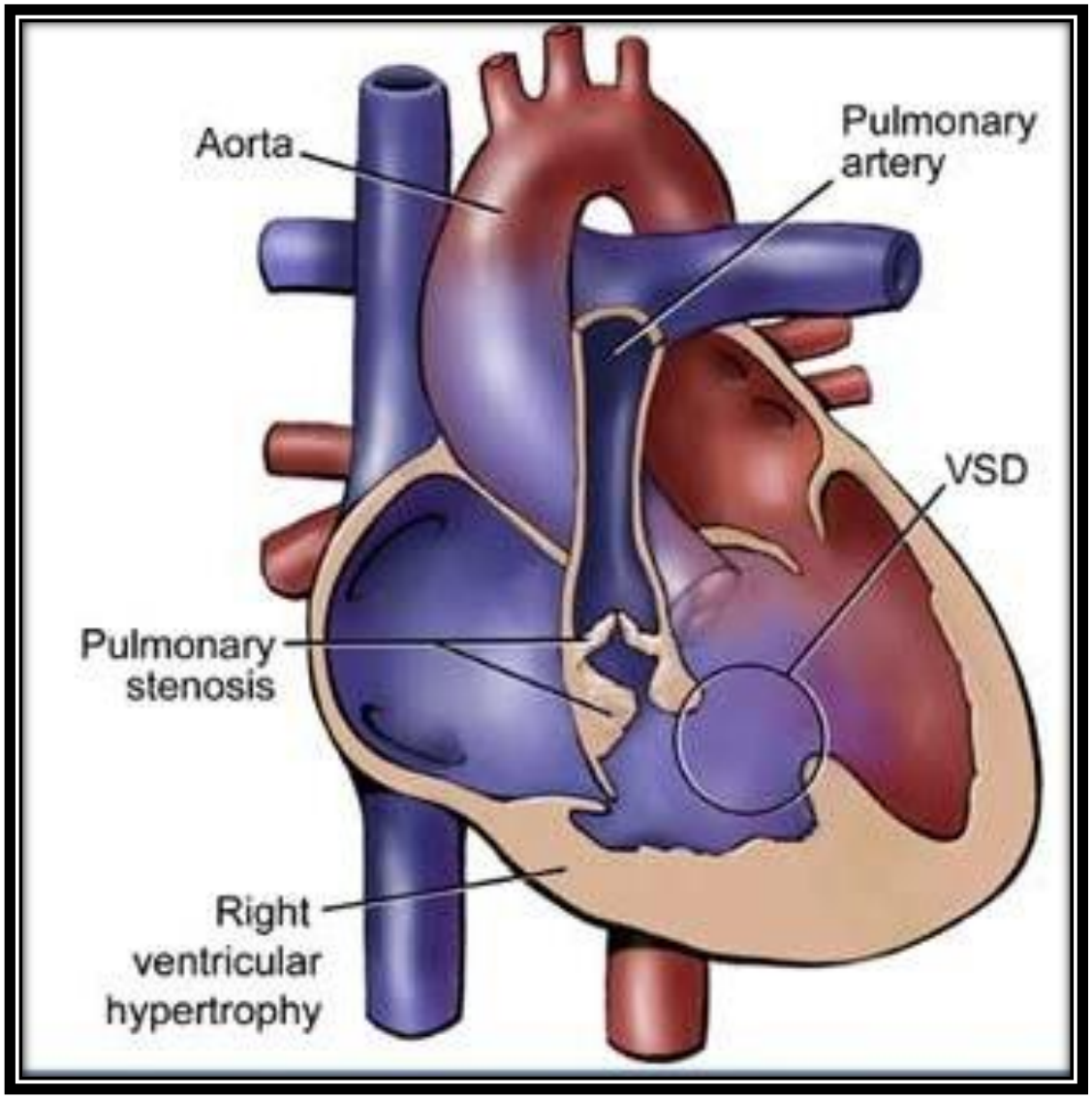


Aortic Valve Stenosis



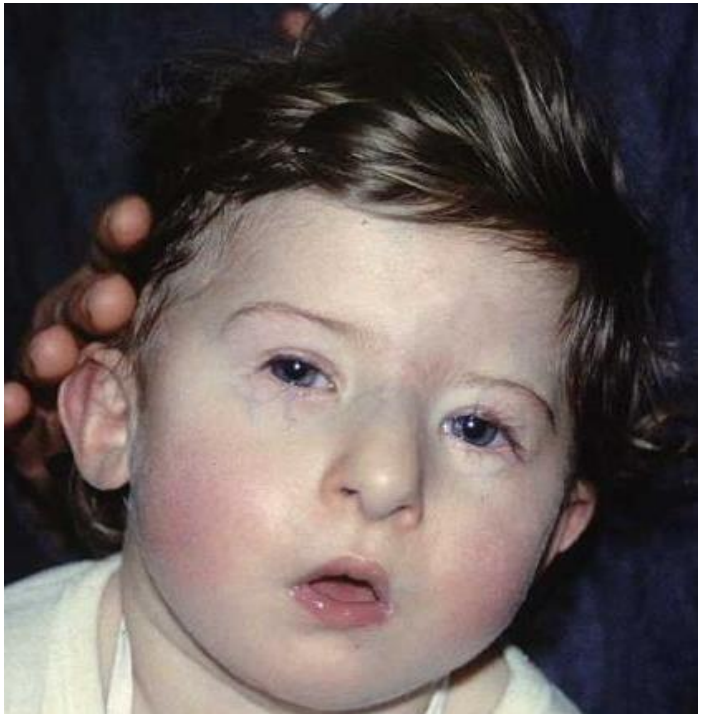
Cyanotic Heart Disease

Tetralogy of Fallot



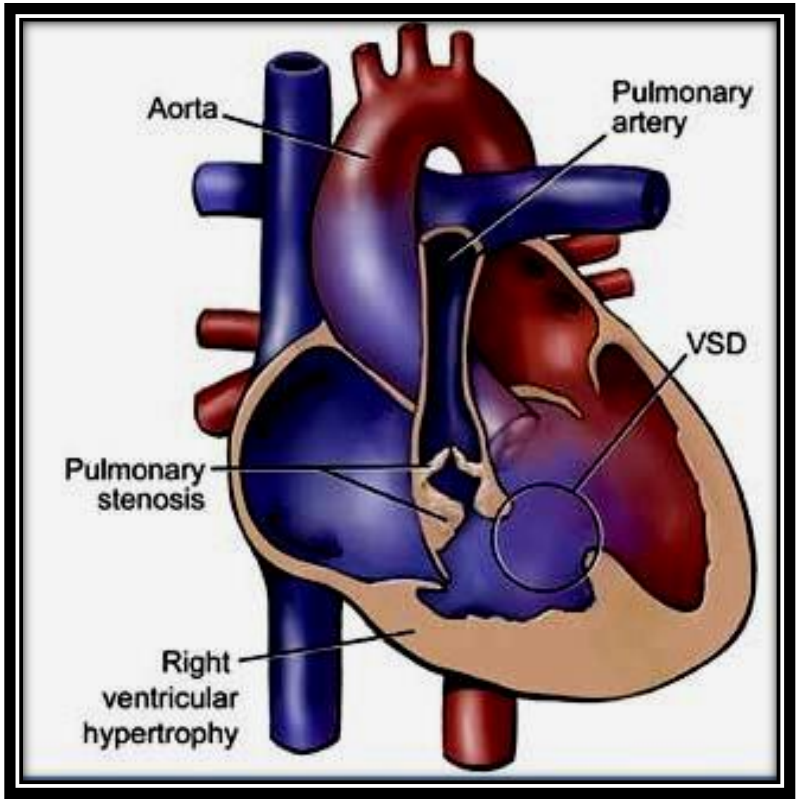
Tetralogy of Fallot

- Most common cyanotic CHD
  - Incidence: 6 % of all CHD
- Can be associated with
  - DiGeorge Syndrome



Tetralogy of Fallot

- Four basic components
  - Large VSD
  - Pulmonary stenosis (PS)
  - Overriding aorta
  - RV hypertrophy



CLINICAL FEATURES: TOF

- Depend on the severity of PS
  - Most newborn:
    - Asymptomatic
    - Ejection systolic murmur on routine discharge exam
    - Initially have mild cyanosis which progress with time:
      - Might present with hypercyanotic spells "tet spell" if delayed intervention

CLINICAL FEATURES: TET SPELL

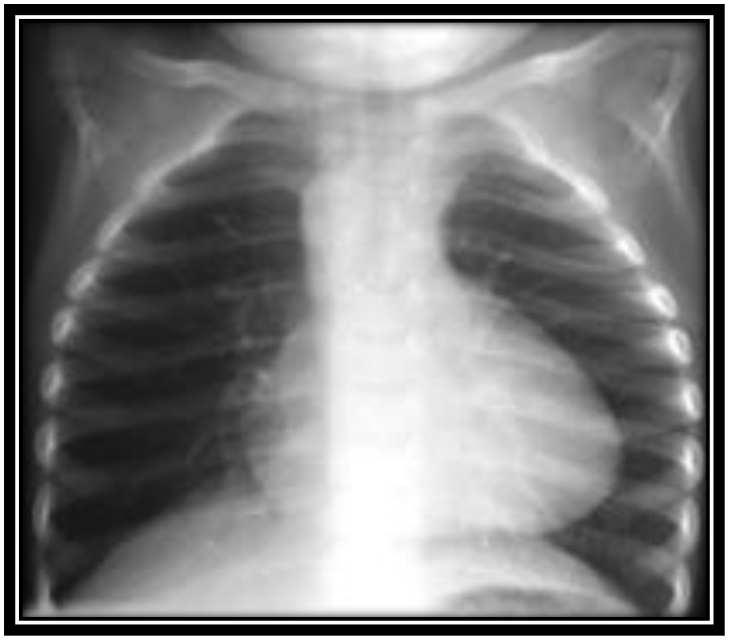
- Usually occur around 9-12 months of age
  - Episodes of acute and severe cyanosis
  - RX:
    - Medical emergency
      - Reduced anxiety “Keep child in his mother lab”
      - Knee-to-chest position
      - Give oxygen
      - Sedation with morphine
      - IV fluid
      - Beta Blocker
      - Phenylephrine
    - Might require emergency surgical intervention

CLINICAL FEATURES: TOF

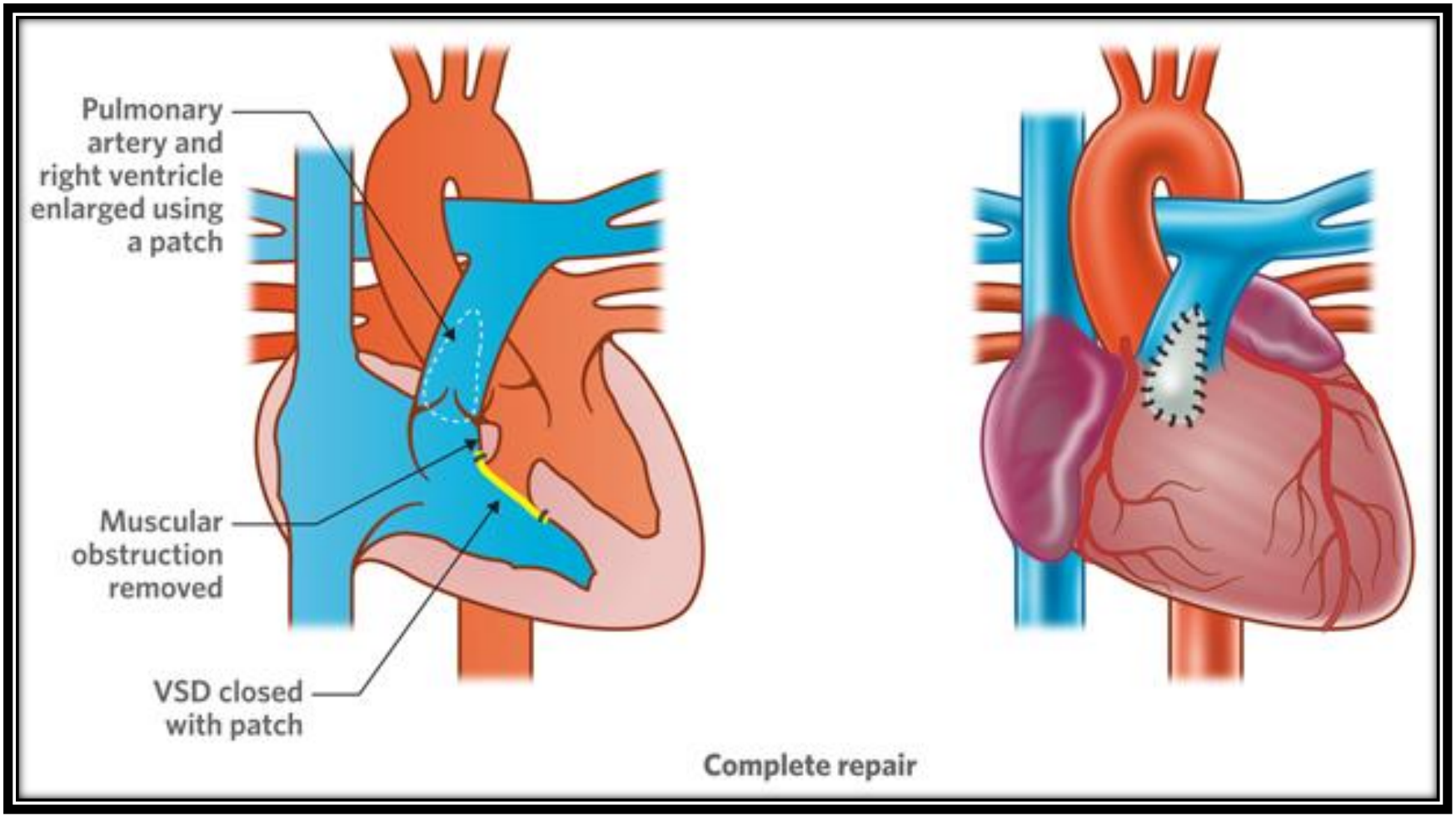
- **Newborn with severe PS or pulmonary atresia**
  - **Severe cyanosis when PDA close**
  - **“Duct dependent CHD”**
  - **Need IV prostaglandin E2**

INVESTIGATION: TOF

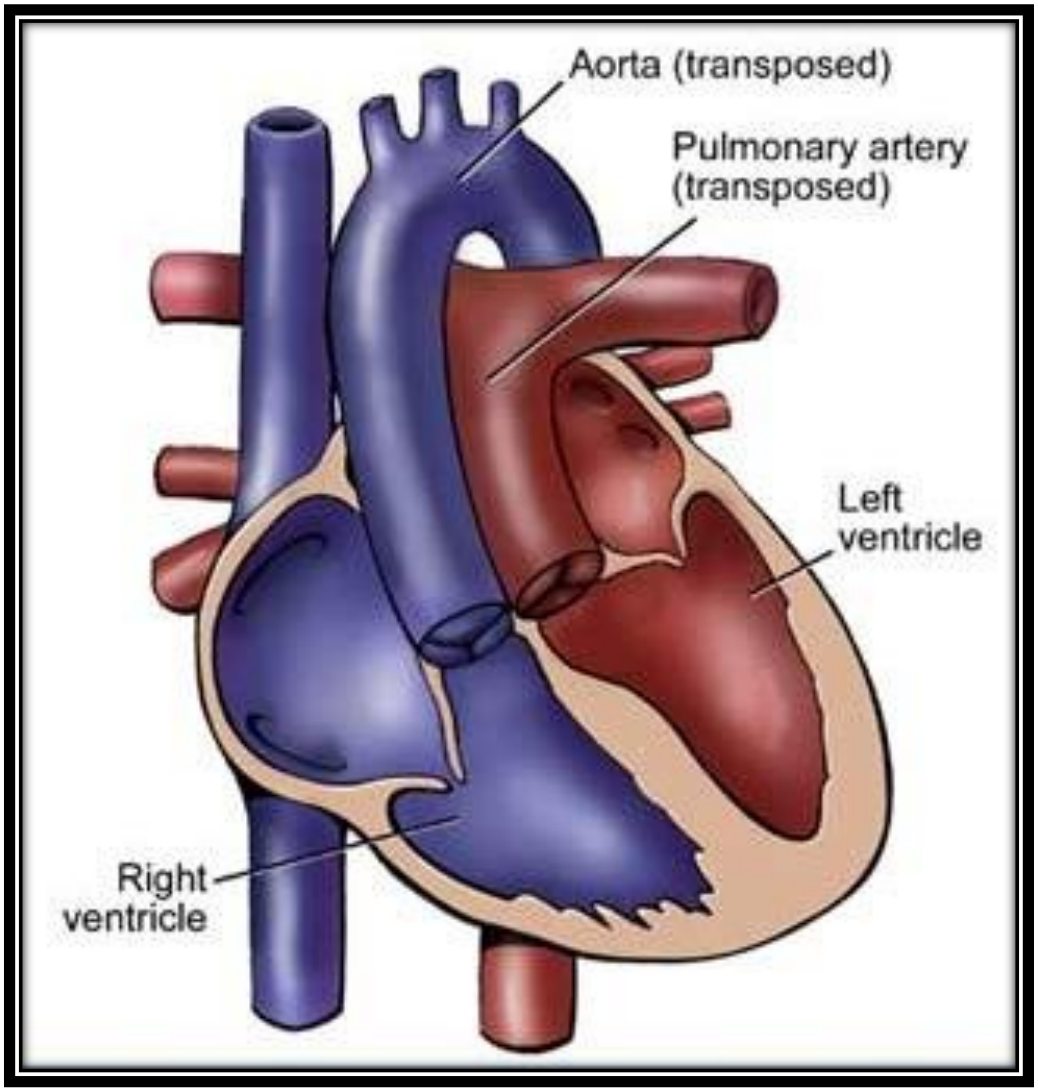
- CHEST X-RAY:
  - “boot-shaped heart”
- ECG: RVH
- ECHO: confirm diagnosis
- CT/MRI rarely needed



TREATMENT: TOF



Transposition of the Great Arteries



D-TGA

- Incidence: 4 % of all CHD
- Most common CHD presented with cyanosis at birth.
- More common in male
- Higher incidence in infant of diabetic mother

PATHOPHYSIOLOGY: D-TGA

- In Normal heart:
  - Pulmonary and systemic circulations are in series
- In D-TGA:
  - Pulmonary and systemic circulations are in parallel

PATHOPHYSIOLOGY: D-TGA

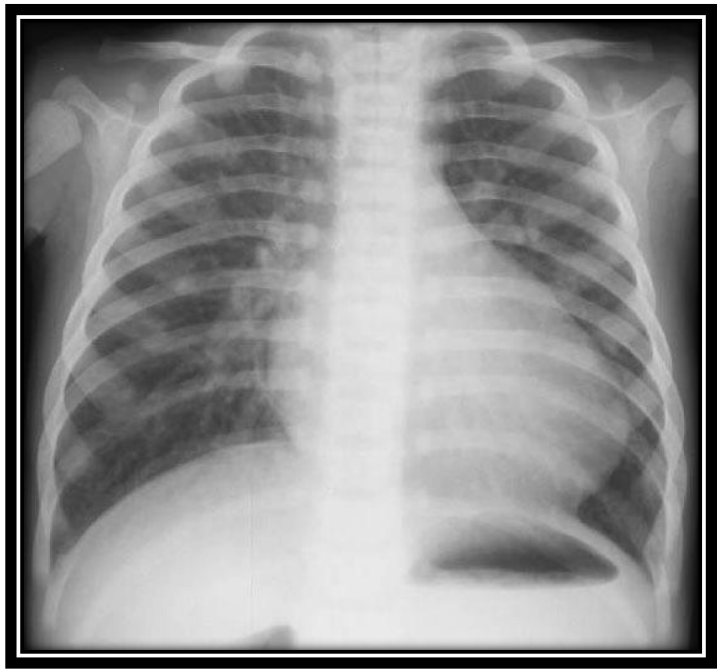
- Mixing of oxygenated and deoxygenated blood can occur at three levels:
  - Atrial level via ASD/PFO (most important)
  - Great arteries level via PDA
  - Ventricular level via VSD (if present)

PRESENTATION: D-TGA

- Severely Cyanosis after birth
- "**Duct Dependent CHD**"
- "**Reverse differential cyanosis**" if Pulm HTN
- No signs of respiratory distress
- Single second heart sound
- Typically: no murmur
- Hyperoxic test: **FAIL**

INVESTIGATION: D-TGA

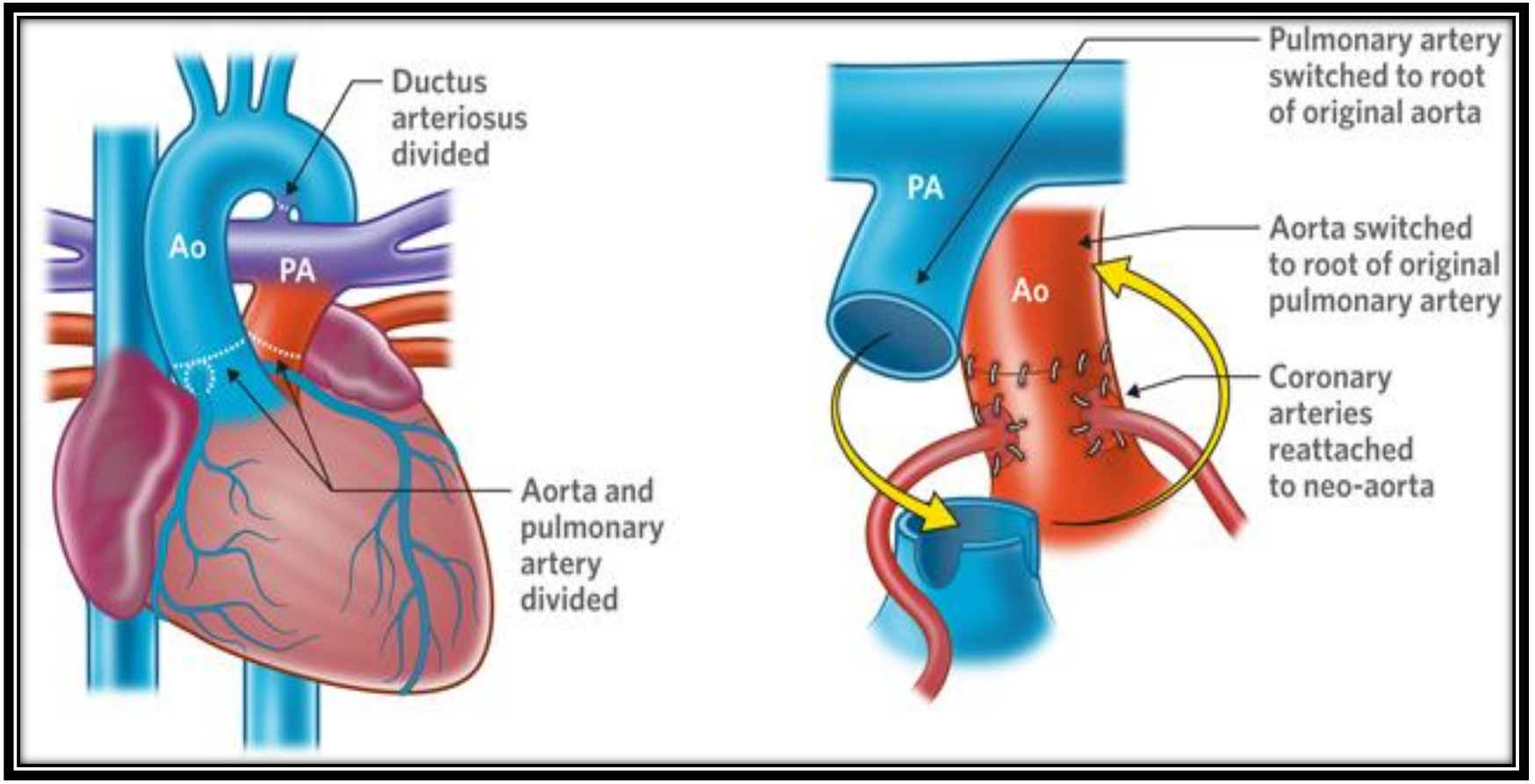
- Chest X-ray:
  - “egg on a string” appearance
- ECG:
  - Typically normal
- ECHO: confirm diagnosis
- Cardiac Cath:
  - For septestomy
  - +/- coronary arteries anatomy



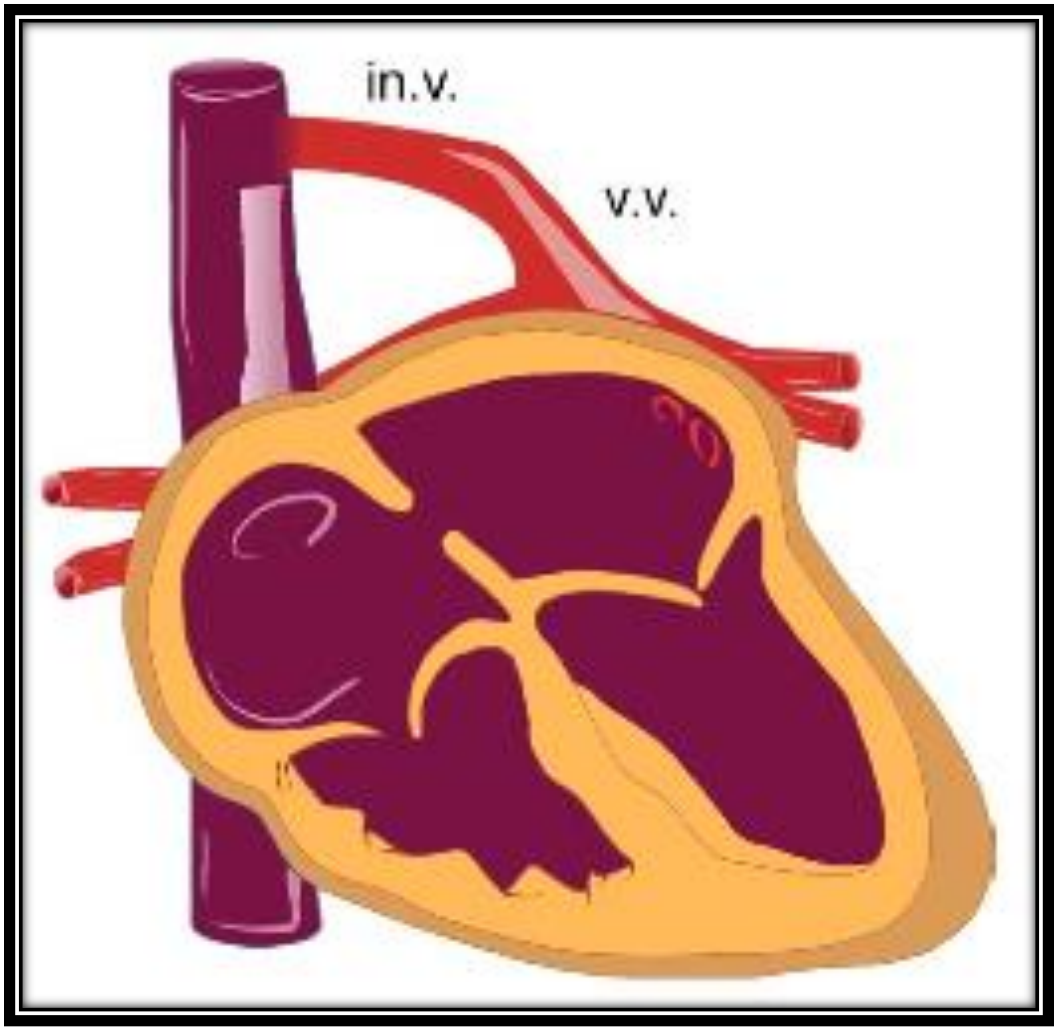
MANAGEMENT: D-TGA

- Supportive:
  - Prostaglandin E2
  - Balloon atrial septostomy (for better mixing)

MANAGEMENT: D-TGA

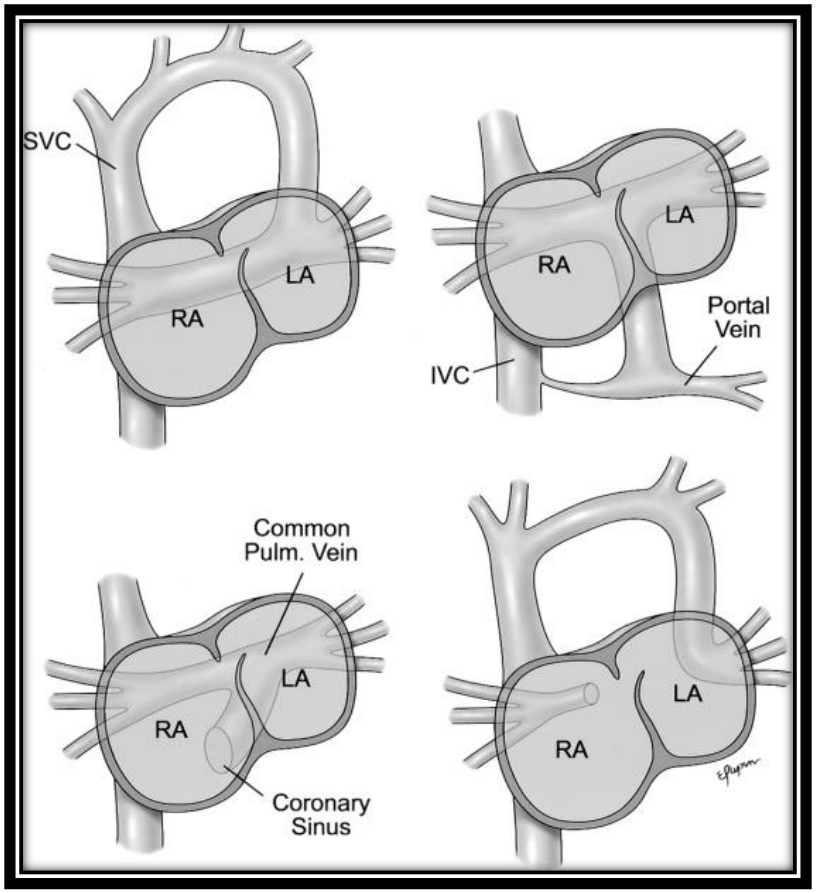


Total Anomalous Pulmonary Venous Return : TAPVD



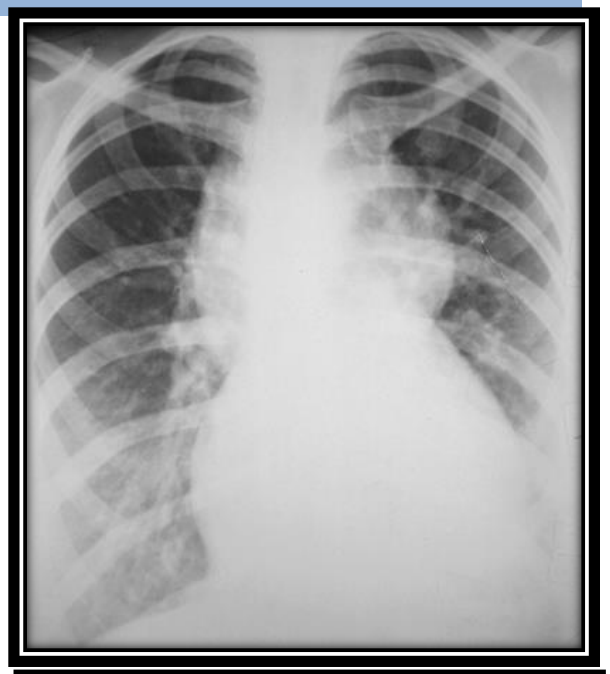
TAPVD

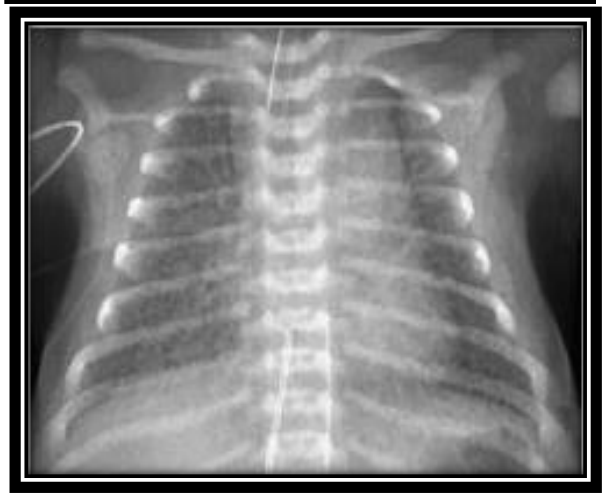
- All 4 pulmonary veins returns to the right atrium
- Can be:
  - Supracardiac (50%)
  - Cardiac (25%)
  - Infracardiac (20%)
  - Mixed (5%)
- Can be:
  - Obstructed TAPVR
  - Non-obstructed TAPVR



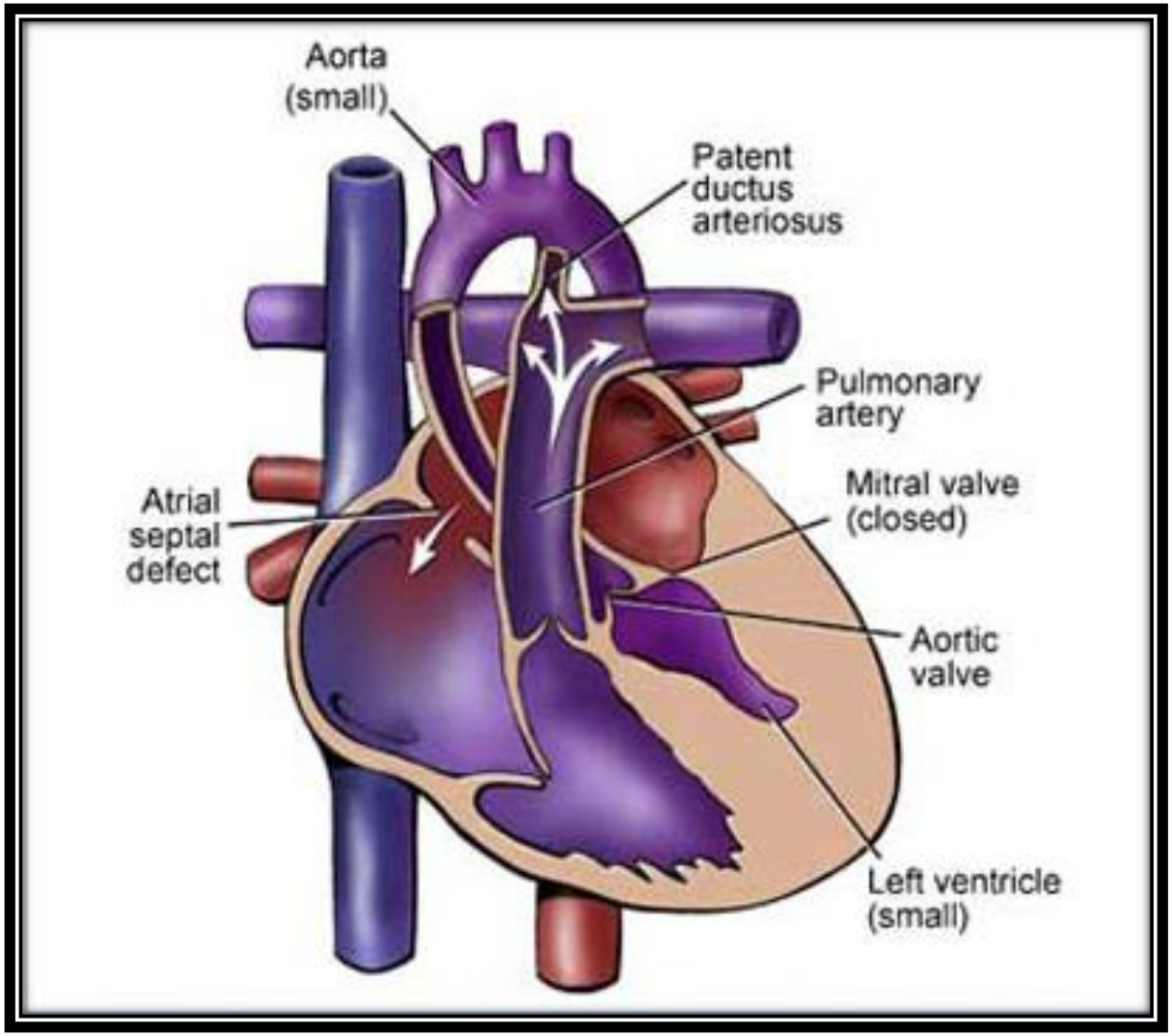
PRESENTATION: TAPVD

- Clinical Feature:
  - Cyanosis at birth
- Diagnosis:
  - Chest X-ray:
    - Increased pulmonary vascualr markings
    - "**Figure of eight**" in obstructed supracardiac TAPVR
  - ECG: RVH
  - ECHO: Confirm Dx
  - Cardiac CT/MI: may be need





Hypoplastic Left Heart Syndrome

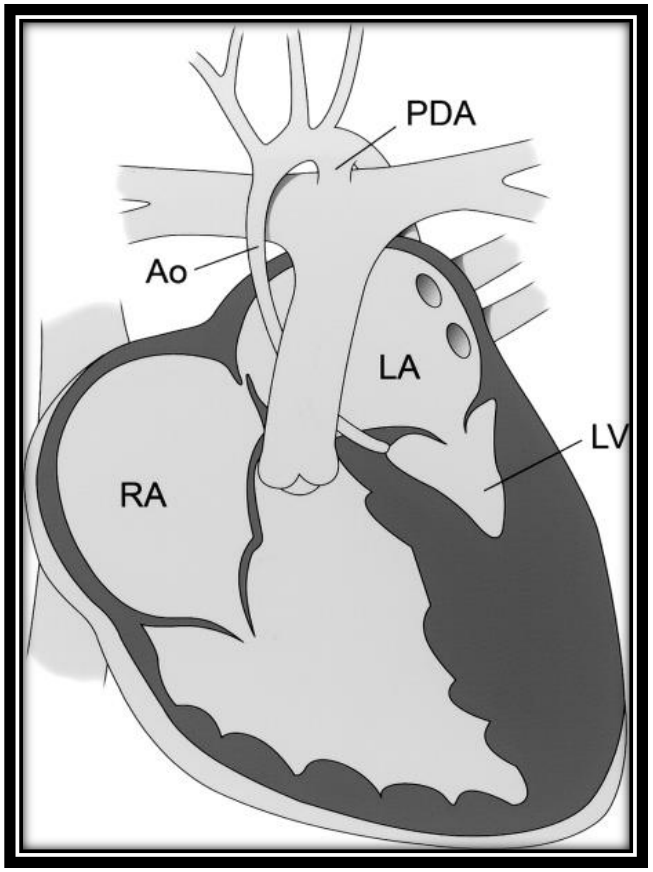


HLHS

- HLHS: one of the most severe form of CHD
  - High morbidity and mortality
- Incidence: 1-2 % of all CHD
- multiple level of obstruction at left heart structures.
  - Mitral stenosis to mitral atresia
  - Variable degree of LV hypoplasia
  - Aortic stenosis to aortic atresia
  - Variable degree of ascending aorta hypoplasia

PATHOPHYSIOLOGY: HLHS

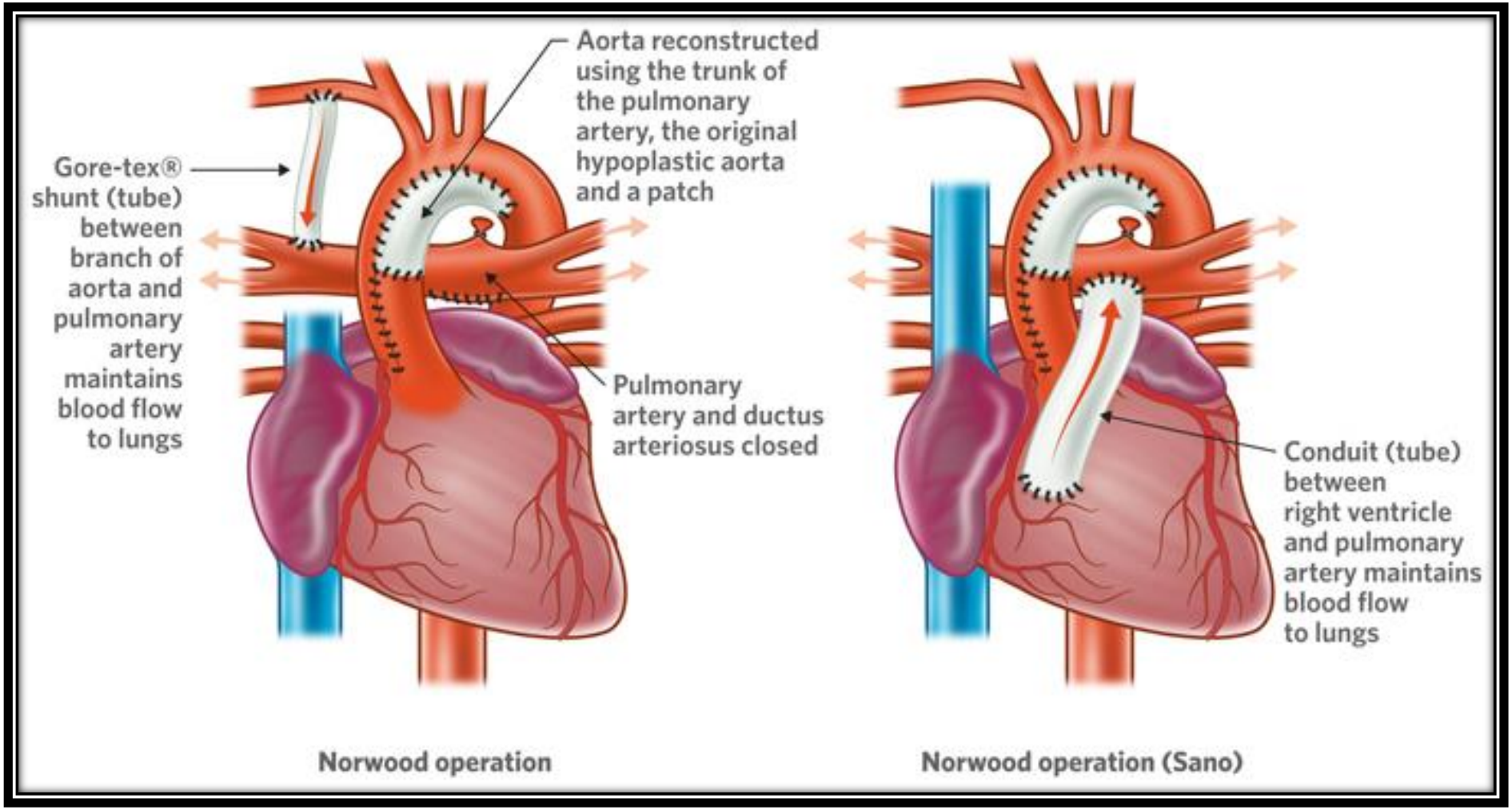
- No adequate flow across aortic valve to ascending aorta
- Relies on retrograde PDA flow to:
  - Brain
  - Coronary arteries
- Need ASD/PFO to shunt blood from LA to RA.



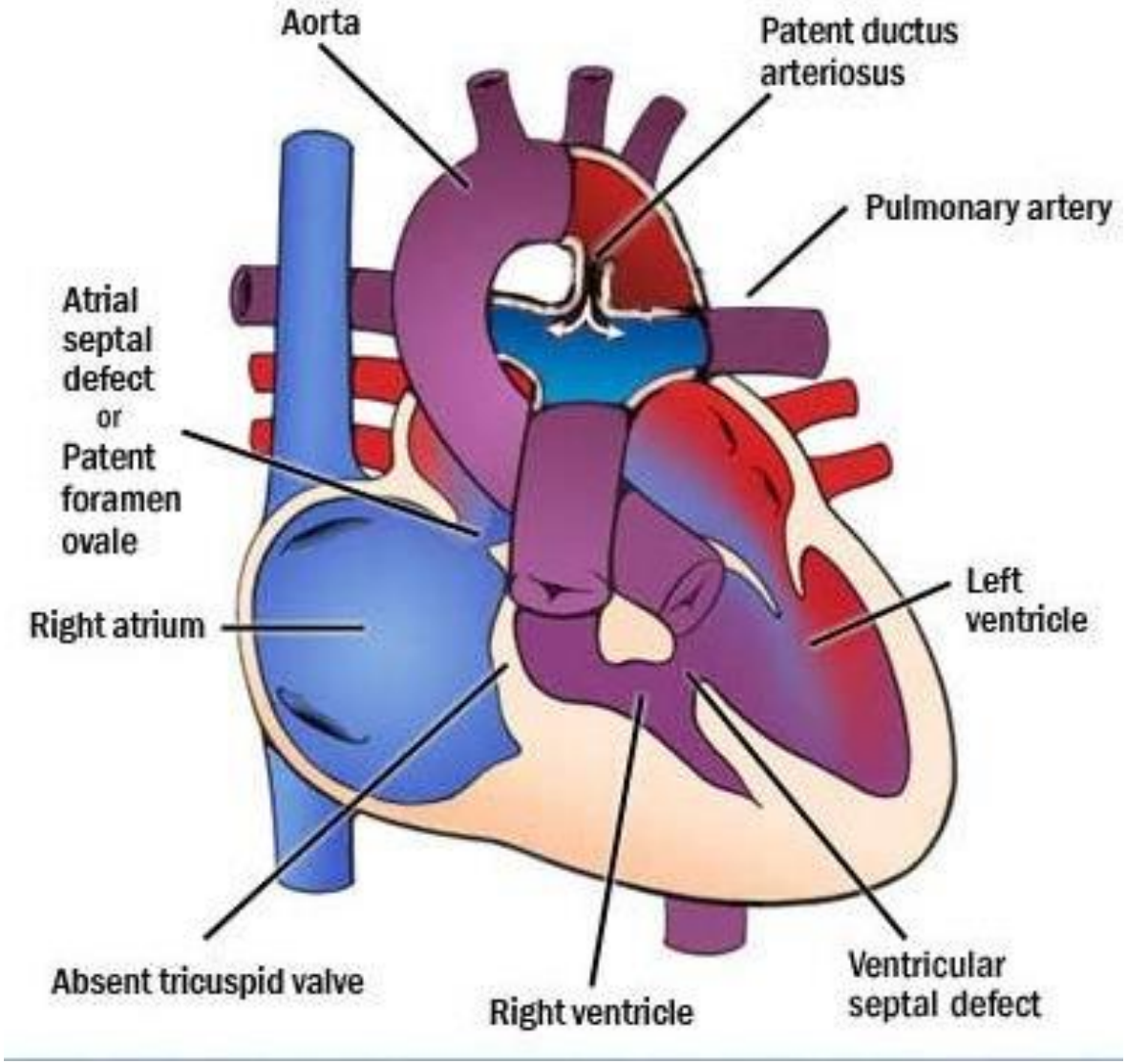
PRESENTATION: HLHS

- At birth: Cyanosis
- At 2-4 week of life:
  - Respiratory distress
  - Poor pulses/perfusion
  - Signs of cardiac shock

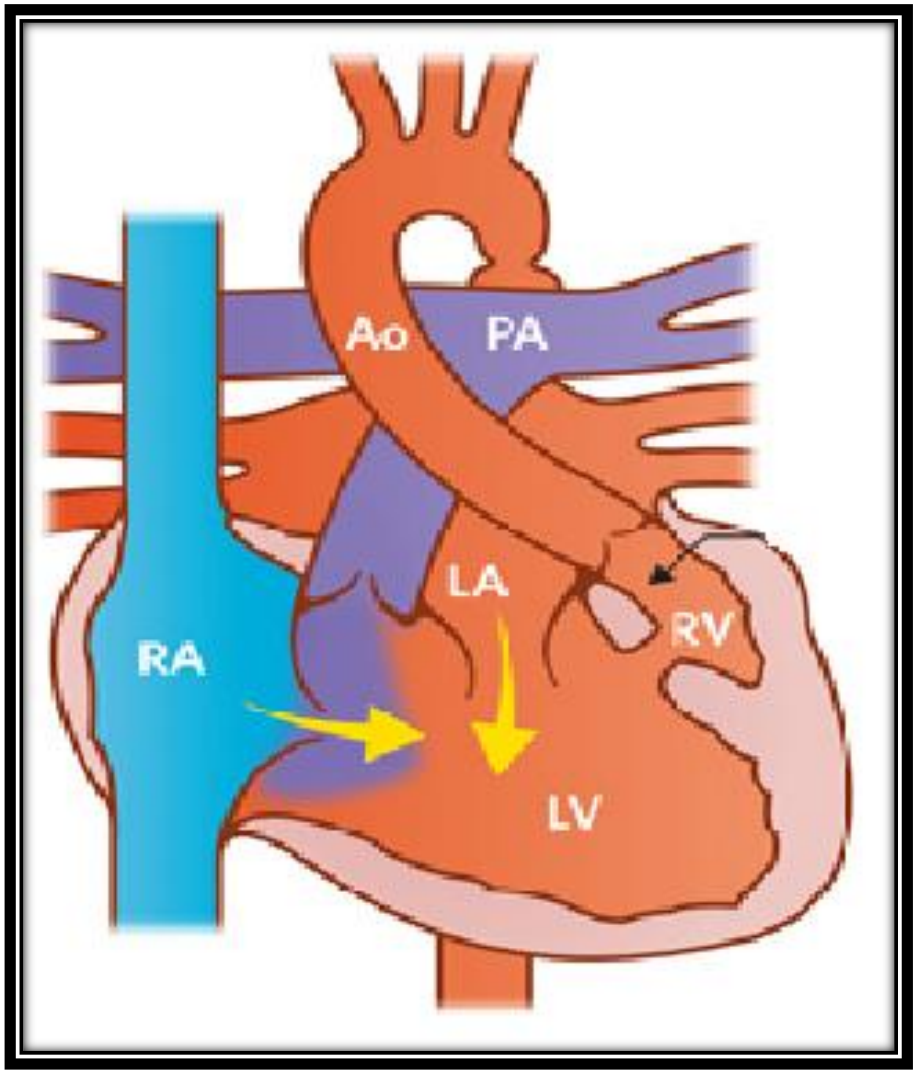
TREATMENT: HLHS



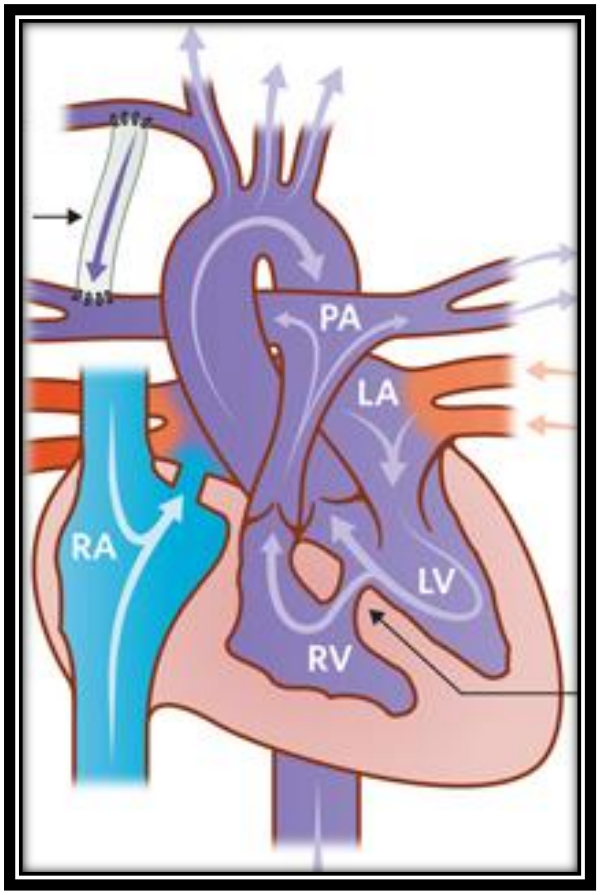
Tricuspid Atresia



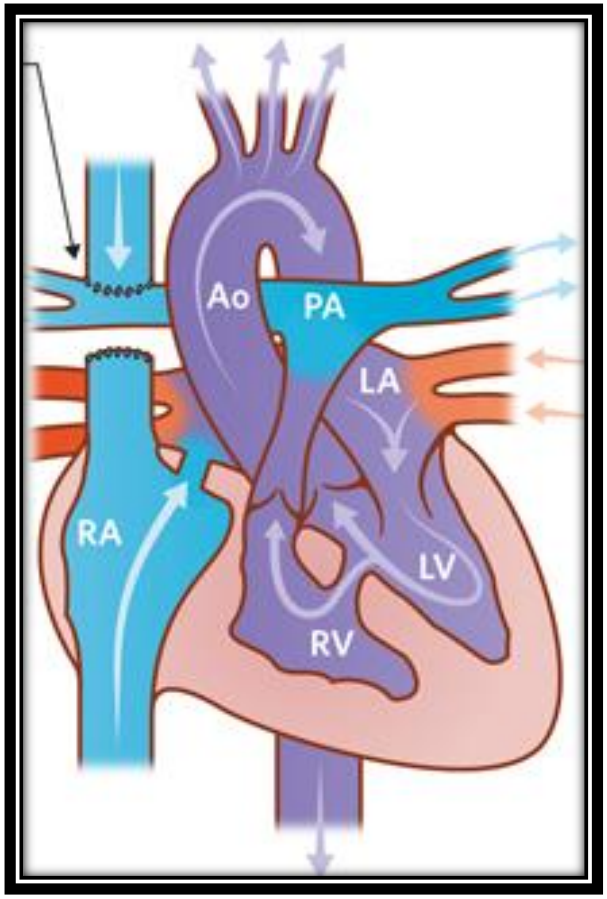
Single Ventricle



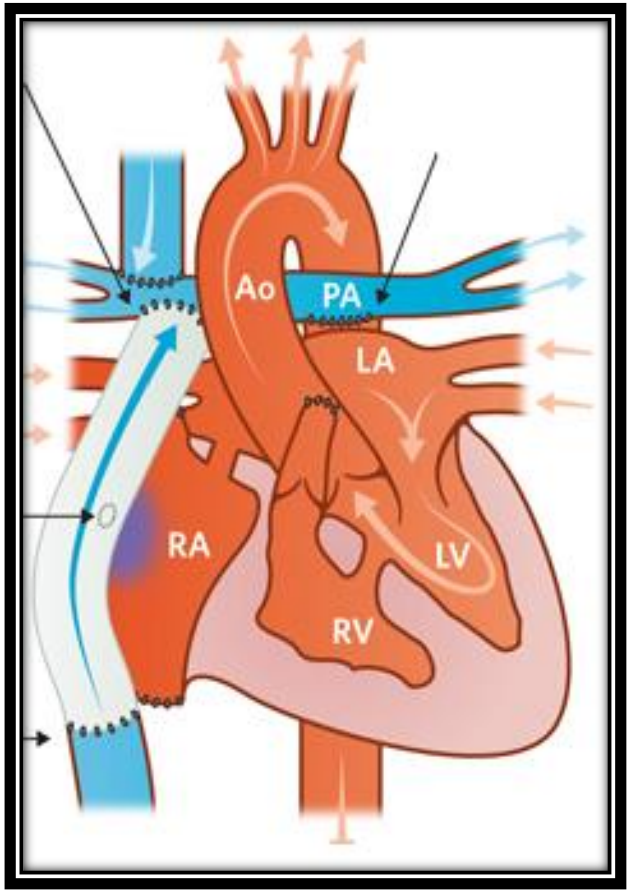
STAGED SURGERY UNIVENTRICULAR HEART



BT SHUNT

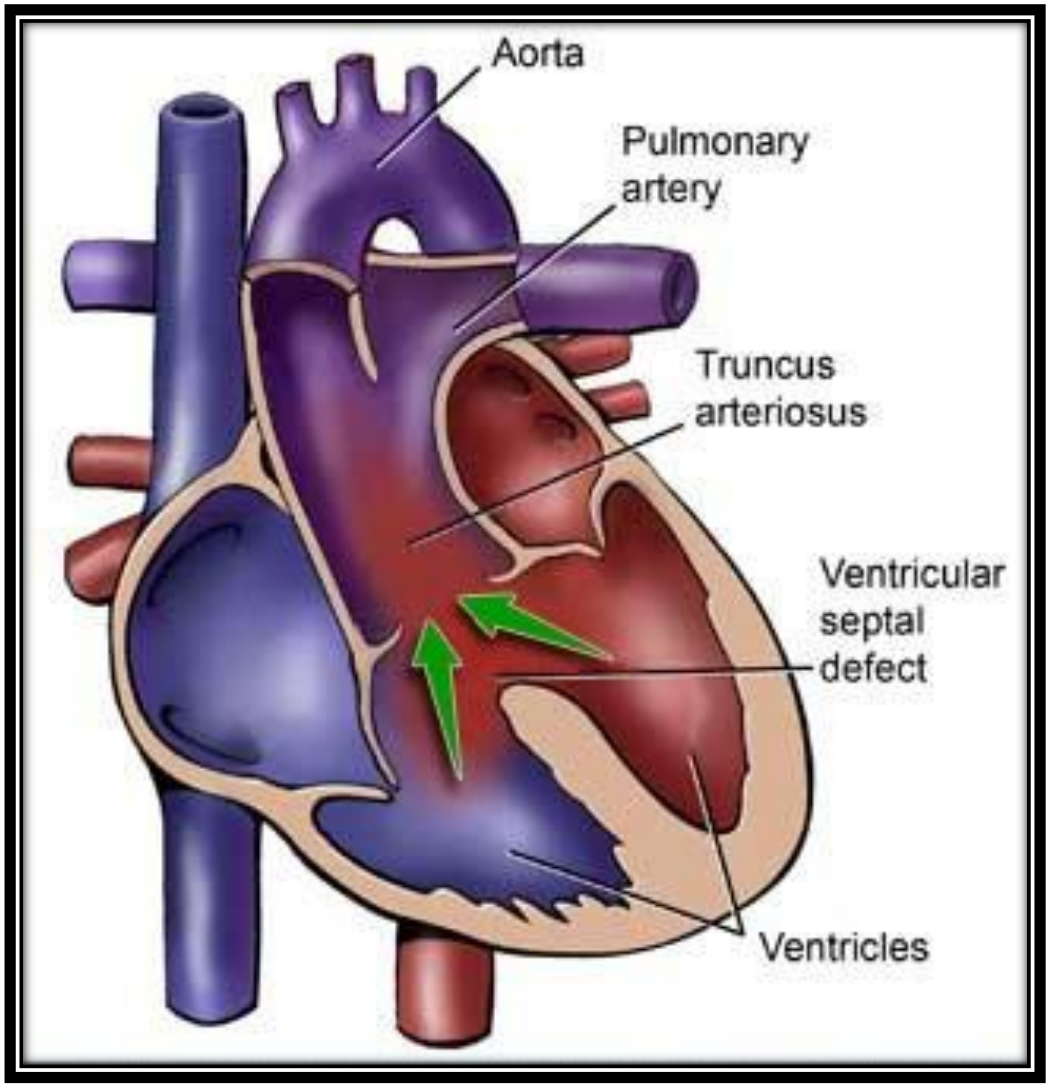


GLENN

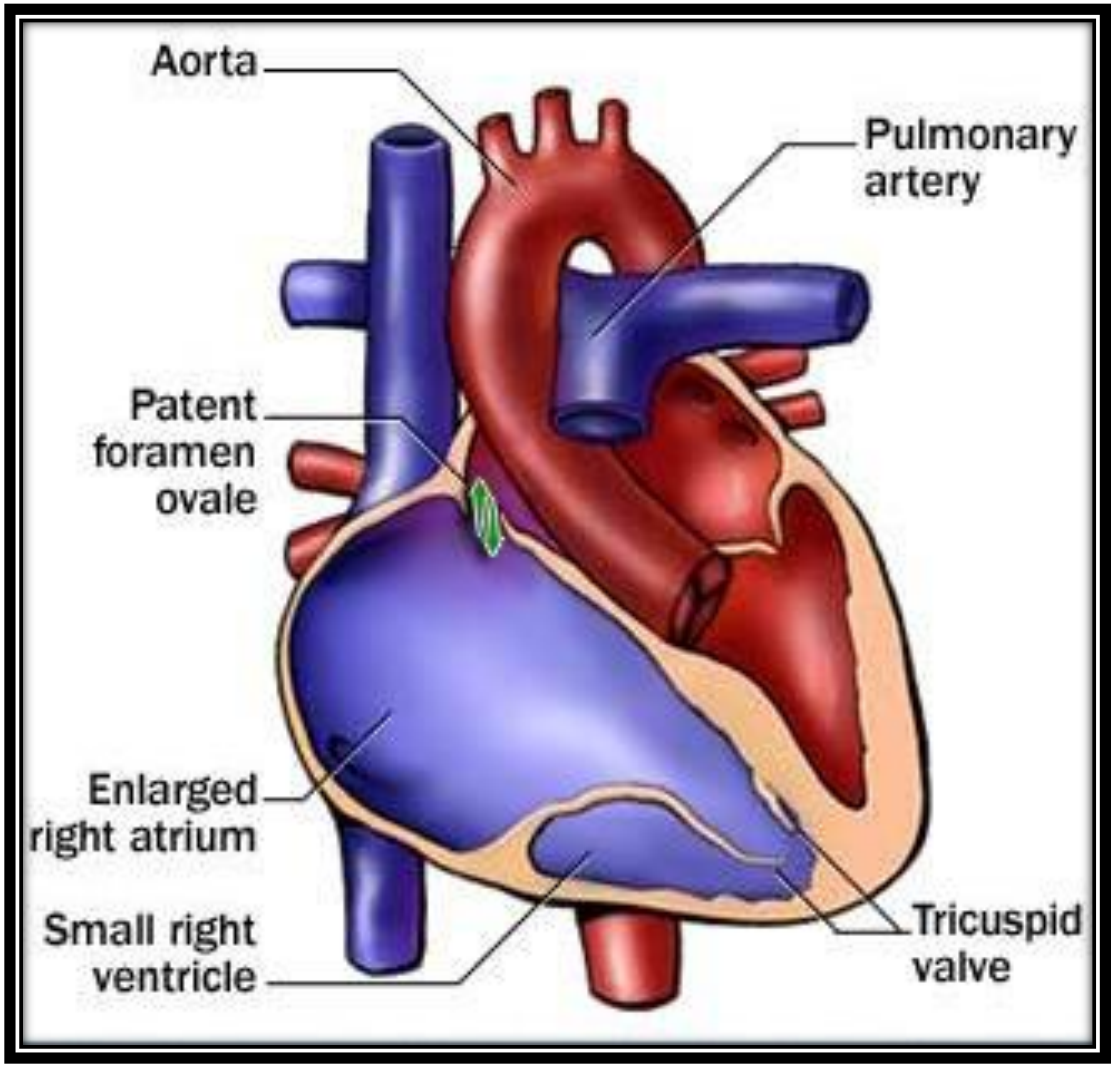


FONTAN

Truncus Arteriosus



Ebstein's Anomaly



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