

Pathophysiology: Left To Right Shunts

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Learning Objectives

- Learn the relationships between pressure, blood flow, and resistance
- Review the transition from fetal to mature circulation
- Determine the effects of the transitional circulation on the physiology of left to right shunts
- Correlate clinical signs and symptoms with cardiac physiology

Pressure, Flow, Resistance

- **Perfusion Pressure:** Pressure gradient across vascular bed
 - Δ Mean Arterial - Venous pressure
- **Flow:** Velocity of flow across vascular bed
- **Resistance:** Opposition to flow
 - Vessel diameter
 - Vessel structure and organization
 - Physical characteristics of blood

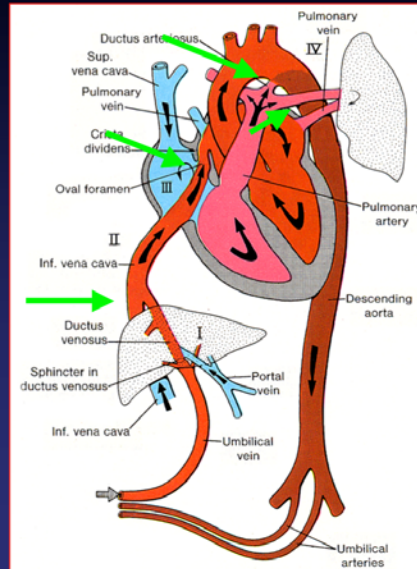
Hemodynamics

$$\text{Flow} = \frac{\Delta \text{ Pressure}}{\text{Resistance}}$$

$$\text{Resistance} = \frac{\Delta \text{ Pressure}}{\text{Flow}}$$

Fetal Circulation

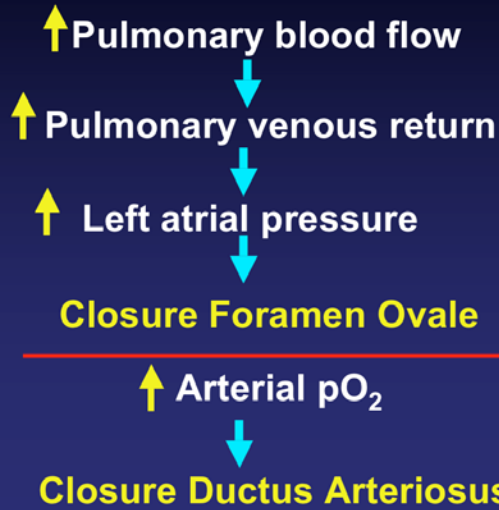
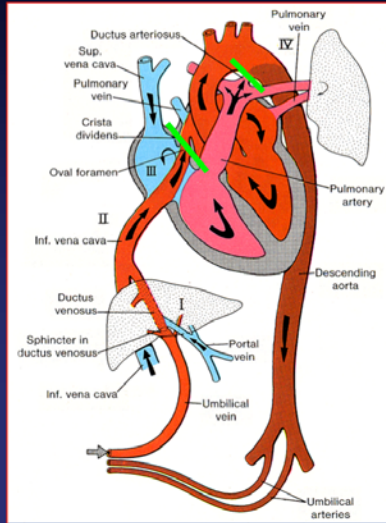
- Placenta supplies oxygenated blood via ductus venosus
- Foramen ovale directs ductus venous blood to left atrium (40%)
- Pulmonary blood flow minimal (10%)
- Ductus arteriosus allows flow from PA to descending aorta (50%)



Fetal Pulmonary Vascular Bed

- Pulmonary Pressure
 - Vasoconstriction
 - Medial wall hypertrophy
 - Pulmonary = Aortic Pressure (DA)
- Pulmonary blood flow
 - Blood bypasses the lungs via the ductus arteriosus to the aorta
 - Flow: Minimal
- Pulmonary resistance: High-Infinite
 - Resistance = $\frac{\Delta \text{Pressure}}{\text{Flow}}$

Transition from Fetal to Neonatal Circulation



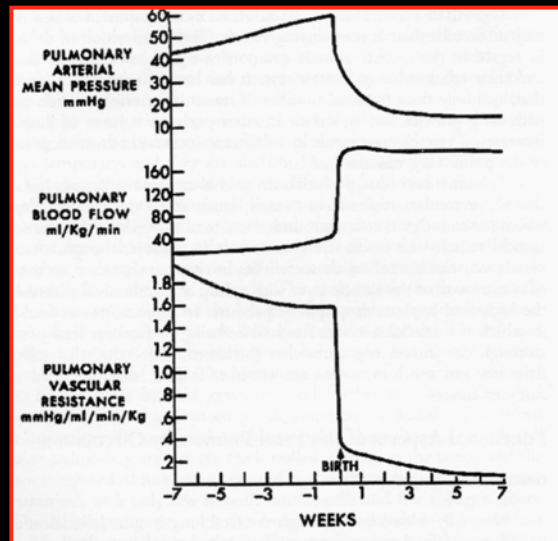
Neonatal Pulmonary Vascular Bed

- **Pulmonary Pressure**
 - Arterial vasodilation
 - Medial wall hypertrophy persists
 - **PA pressure = Aortic pressure**
- **Pulmonary Blood flow**
 - Ductus arteriosus closes
 - Neonatal cardiac output normal
 - **Pulmonary Flow = Aortic Flow**
- **PA resistance = Aortic Resistance**
 - Resistance = $\frac{\Delta \text{Pressure}}{\text{Flow}}$

Regulation of Pulmonary Vascular Tone

- **Vasconstriction**
 - Hypoxia/acidosis
 - High blood flow and pressure
 - Failure of vessel maturation (no regression of medial hypertrophy)
- **Vasodilation**
 - Improved oxygenation
 - Prostaglandin inhibition
 - Thinning of vessel media (regression of medial hypertrophy)

Pulmonary Vascular Bed: Transition from Fetal to Adult



$$R = \frac{\Delta P}{F}$$

Adult Pulmonary Vascular Bed

- **Pulmonary Artery Pressure: Low**
 - Arterial Vasodilation
 - Medial wall hypertrophy regresses
 - Pulmonary \ll Aortic pressure
 - 15 mmHg vs. 60 mmHg
- **Blood flow**
 - Pulmonary = Aortic
- **Resistance:**
 - Pulmonary \ll Aortic Resistance

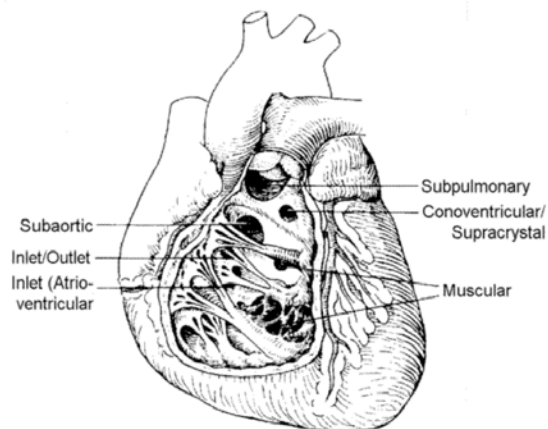
Left to Right Shunt Lesions

- **Anatomic Communication:**
Pulmonary and Systemic circulations
- **Blood flow occurs from the Systemic (Left) to the Pulmonary (Right) circulation**

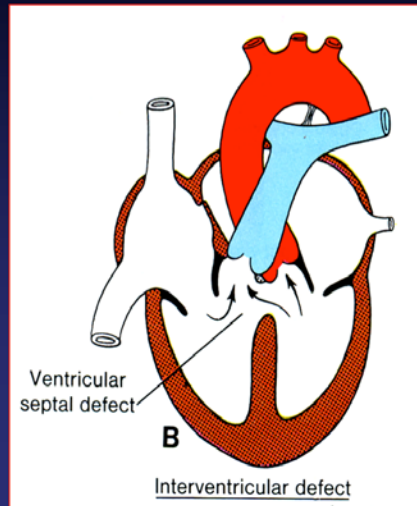
“Top 4” Left to Right Shunt Lesions

- **Ventricular Septal Defect (VSD)**
 - Left ventricle to Right ventricle
- **Persistent Patent Ductus Arteriosus (PDA)**
 - Aorta to Pulmonary artery
- **Endocardial Cushion Defect (ECD)**
 - Left ventricle to Right ventricle
 - Left atrium to Right atrium
- **Atrial Septal Defect (ASD)**
 - Left atrium to Right atrium

Ventricular Septal Defect



VSD: 2/1000 live births Determinants of L to R shunt Flow

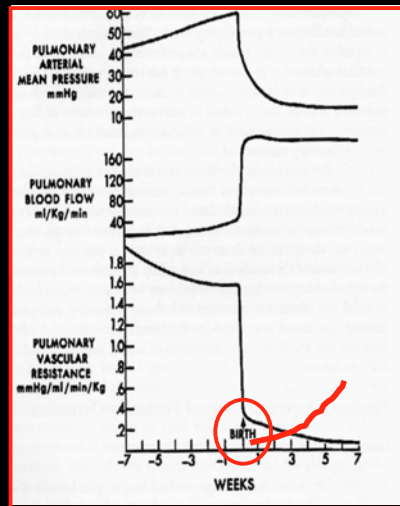


- Size of Defect
- Maturity of Pulmonary Bed
- Pressure difference between RV and LV
- Relative resistance between Pulmonary and Systemic circulations

Determinants of Left to Right Shunt

- **Small (restrictive) VSD:** L to R shunt flow limited by size of hole
- **Large (unrestrictive) VSD:** L to R shunt flow is determined by:
 - RV vs. LV pressure
 - If $RV < LV$, L to R shunt occurs
 - If $RV = LV$
 - If pulmonary $<$ aortic resistance, L to R shunt occurs

Transitional Circulation: Effects on L to R shunt in large VSD

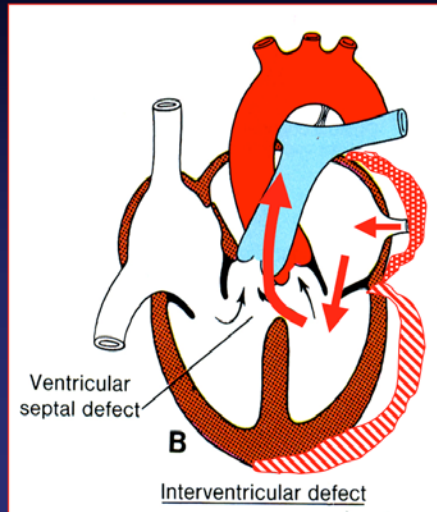


- Fetal: No shunt
- At Birth: No shunt
- Transition 1-7 weeks
 - PA/RV decreases to < LV
 - PA resistance decreases to < Systemic
 - L to R shunt increases

Large VSD: Degree of L to R Shunt

- **Large L to R shunt (normal transition)**
 - Pulmonary << Aortic Pressure
 - Pulmonary << Aortic Resistance
- **Small L to R shunt (no transition)**
 - Medial hypertrophy fails to regress
 - Damage to vessel wall from high blood flow/shear stress leads to vasoconstriction
 - Pulmonary pressure and resistance remain elevated, L to R shunt small

Large VSD: Hemodynamic Effects of Large L to R shunt



- Flow from LV to RV to Pulmonary Artery (PA)
- Increased Pulmonary Venous Return
- Increased LA size/Pressure
- Increased flow across mitral valve
- Increased LV size: Congestive HF

Natural History of Large VSD

- **Asymptomatic at birth: Pulmonary = Aortic Pressure and Resistance**
- **Signs of congestive heart failure as pulmonary pressure and resistance falls**
 - Poor feeding
 - Failure to thrive (FTT) with preserved height and low weight
 - Tachypnea
 - Diaphoresis
 - Hepatomegaly
 - Increased respiratory illness

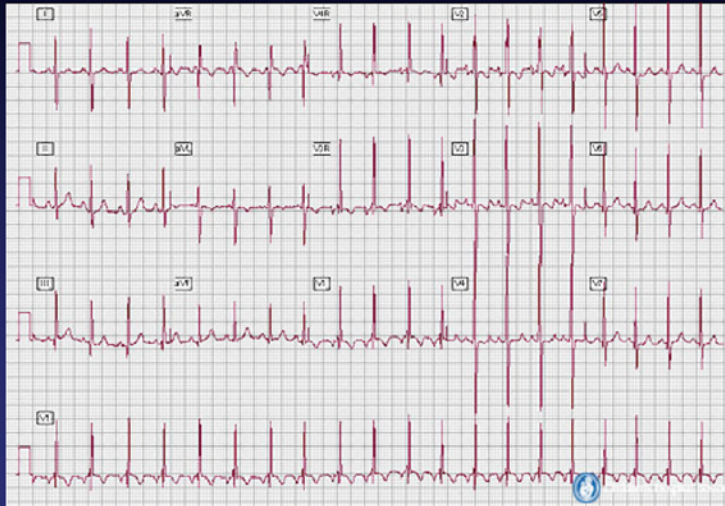
VSD: Clinical Findings

- **Holosystolic murmur loudest LLSB radiating to apex and back**
- **Mid-Diastolic rumble: Increased flow across the mitral valve**
- **LV heave: LV dilation**
- **Precordial Thrill: turbulent blood flow across VSD**
- **Heart failure: Gallop rhythm (S3), Hepatomegaly, Rales**
- **↑ Second heart sound: elevated PA pressure**

Laboratory Findings: VSD

- **EKG: LV dilatation ± RVH (if pulmonary artery pressure high)**
- **Chest x-ray: Large heart, ↑ PVM**
- **Echo: Gold Standard**
 - Location/Size of lesion
 - LA/LV size
 - Estimation RV pressure
- **Catheterization: only in cases when high PVR suspected**

Electrocardiogram: VSD

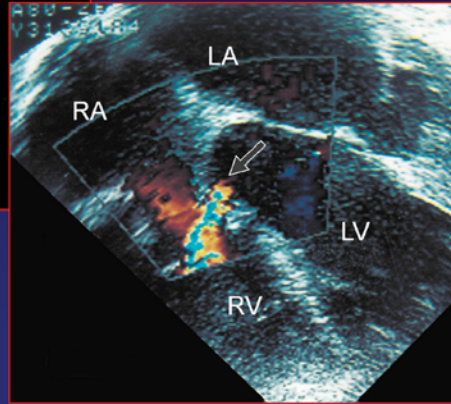
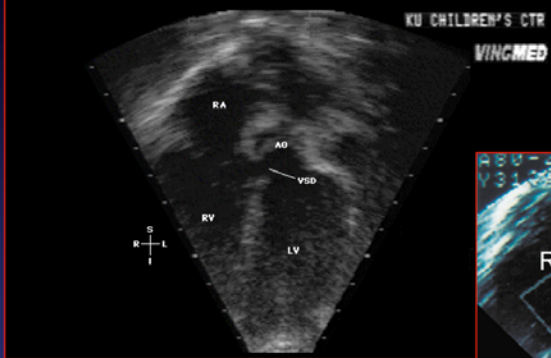


Chest X-Ray: VSD

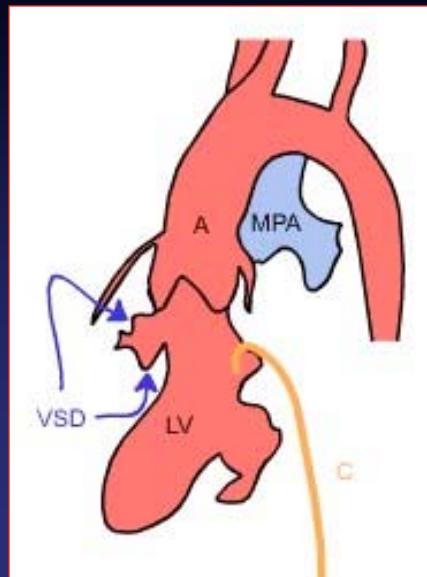


Echocardiogram: Membranous VSD

Membranous Ventricular Septal Defect, Apical 4 Chamber View, Inverted



Angiogram: VSD



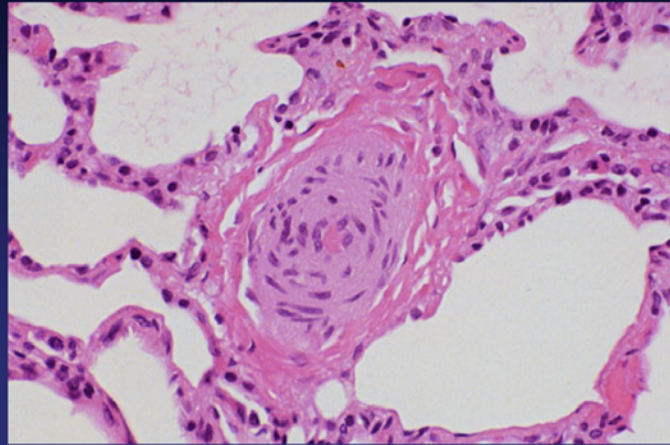
Treatment of Large VSD

- **Medical: Anticongestive Therapy**
 - Digoxin
 - Lasix
 - Increased caloric intake
- **VSD size decreases**
 - Resolution of CHF without surgery (50%)
- **Indications for VSD closure**
 - Persistent CHF with failure to thrive or other symptoms
 - Increasing pulmonary vascular resistance
 - Within first two years of life

Eisenmenger's Syndrome

- **Dr. Victor Eisenmenger, 1897**
- **Pathophysiology**
 - Medial hypertrophy of pulmonary arteries
 - Perivascular necrosis
 - Replacement of normal vascular architecture
- **High pulmonary vascular resistance**
 - Right to left shunt via VSD
 - Severe cyanosis

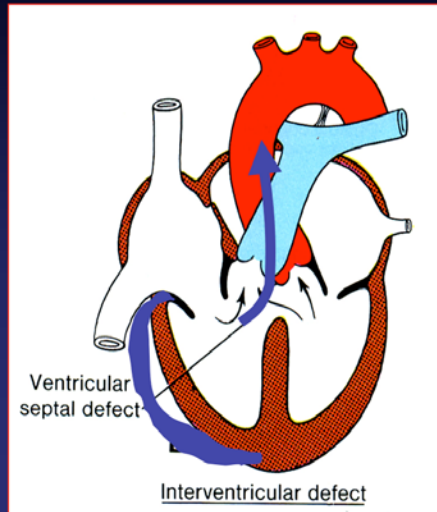
Medial Hypertrophy



Effect of Large Left to Right Shunt on Pulmonary Vascular Bed

- High pulmonary blood flow: Shear Stress
 - Medial hypertrophy
 - Endothelial damage
 - $\uparrow \Delta \text{ Pressure}$ = $\downarrow \text{ Blood Flow}$
 - $\uparrow \text{ Resistance}$
- Left to right blood shunt decreases
- Congestive heart failure improves

Eisenmenger's Syndrome R to L flow via VSD



- Pressure:
 - Pulmonary = Aortic
- Resistance
 - Pulmonary > Aorta
- RV hypertrophy
- Blood flow: RV to LV
- Cyanosis
- Normal LA/LV size

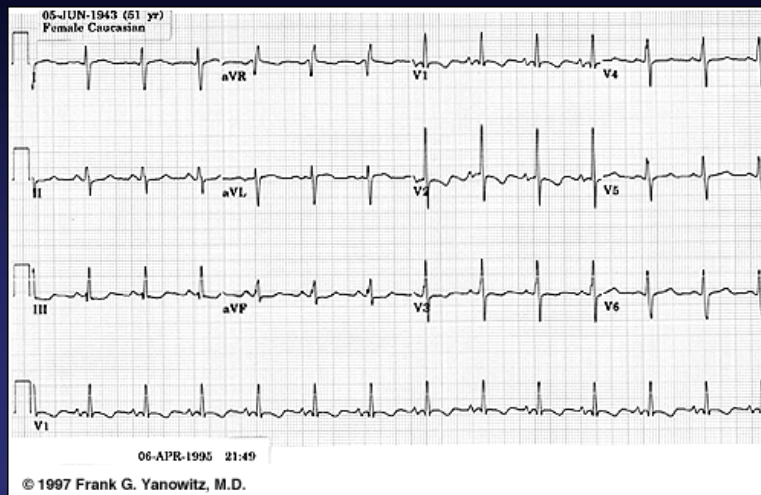
Clinical Picture: Eisenmenger's

- Rare disease in modern era
- Clinical improvement of heart failure in infancy due to decreased left to right shunt
- Clinical presentation: young adulthood
 - Exercise Intolerance
 - Cyanosis
 - Clubbing
 - No systolic murmur
- Elevated PA pressure/resistance
 - Second heart sound increased
 - RV heave (RV hypertension)
 - Pulmonary insufficiency murmur

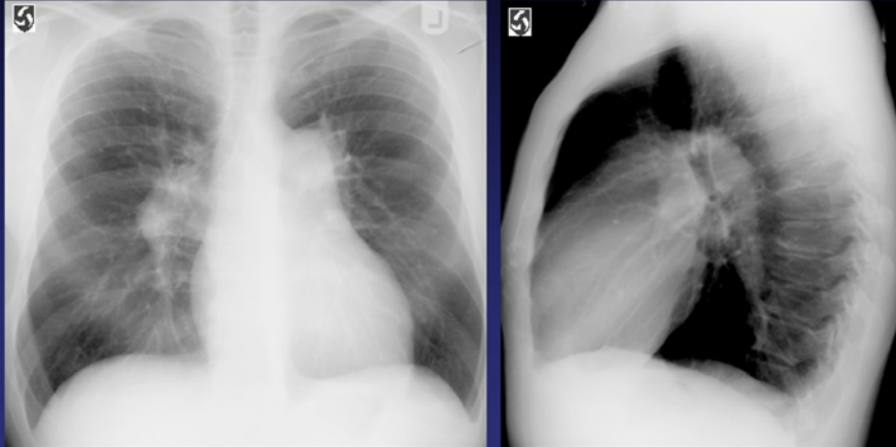
Lab findings: Eisenmenger's

- No LV volume overload
- High RV pressure overload
- EKG: RVH \pm strain
- Echo: RV hypertrophy, right to left shunt at VSD
- Chest x-ray: Clear lung fields, prominent PA segment, small heart

EKG: Eisenmenger Syndrome



Chest X-Ray: Eisenmenger Syndrome



Management

- **Do NOT close VSD**
 - No left to right shunt: No heart failure
 - Shunt is right to left through VSD
 - VSD must stay open to decompress high pressure RV and prevent RV failure
- **Pulmonary vasodilators**
 - Calcium channel blocker
 - PGI₂, Sildenafil
- **Inotropic support**
 - Right heart failure
- **Transplant**
 - Heart-Lung
 - Lung transplant, heart repair

Patent Ductus Arteriosus (PDA)

- Communication between Aorta and Pulmonary Artery
- L to R shunt depends on pulmonary artery pressure and resistance
- Continuous murmur (flow occurs in systole and diastole)
- Congestive heart failure

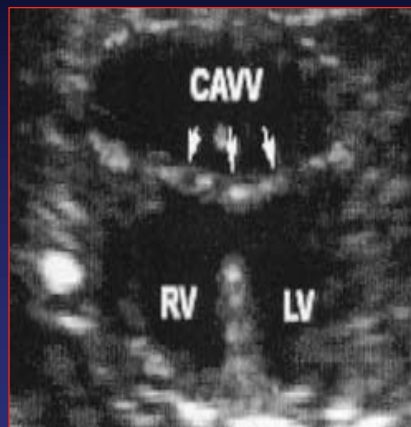
Management: PDA

- **Indications for Closure**
 - CHF/failure to thrive
 - Pulmonary hypertension
- **Closure Methods**
 - Surgical ligation
 - Transcatheter closure
 - Coil
 - Device

PDA Coil Closure



Endocardial Cushion Defect



- Atrial Septal Defect (Primum)
- VSD
- Common Atrioventricular Valve

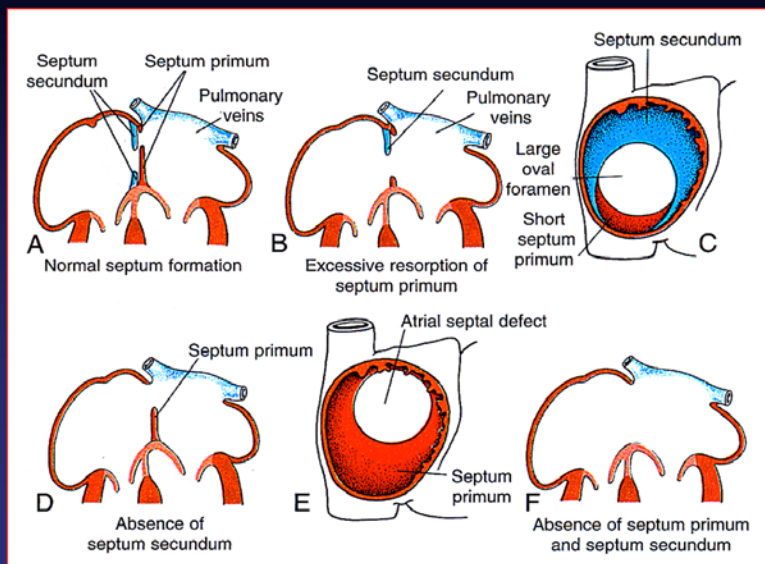
Management: ECD

- **Closure always indicated**
- **Timing of surgery (elective by 6 mos.)**
 - Heart Failure
 - Large left to right shunt
 - Mitral insufficiency
 - Pulmonary hypertension
- **Surgical repair**
 - ASD, VSD closure
 - Repair of AV-Valves

Summary: VSD, PDA and ECD

- **Asymptomatic in fetus and neonate**
- **Progressive increase in L to R shunt from 3-8 weeks of life as pulmonary pressure and vascular resistance decreases**
- **Indications for intervention**
 - Congestive heart failure: FTT
 - Pulmonary vascular disease
- **End stage: Eisenmenger's syndrome**

Atrial Septal Defect (ASD)



Manifestations of ASD

- **L to R shunt between left and right atria**
 - Increasing L to R shunt with age as LV compliance decreases and LA pressure increases
- **Survival less than age-matched population (5th-6th decade)**
 - Arrhythmias
 - Right heart failure

Management ASD

- **Indications for closure**
 - RV volume overload
 - Pulmonary hypertension
 - Thrombo-embolism
- **Closure method**
 - Surgical
 - Device
 - Cardioseal
 - Amplatzer septal occluder

Amplatzer Septal Occluder

