

## 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**
  - $\Delta$  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

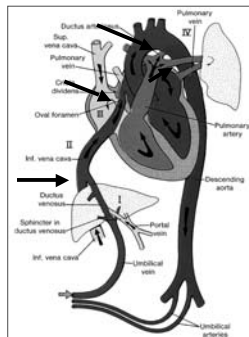
### Hemodynamics

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

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

### Fetal Circulation

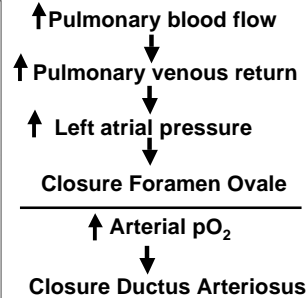
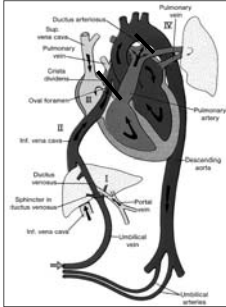
- Placenta supplies oxygenated blood via ductus venosus
- Foramen ovale directs ductus venos blood to left atrium (40%)
- Pulmonary blood flow minimal (10%)
- Ductus arteriosus allows flow from PA to descending aorta (50%)



### 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



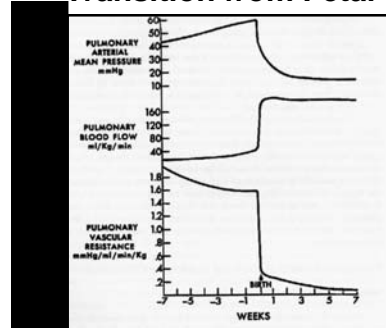
## Neonatal Pulmonary Vascular Bed

- **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

- **Vasoconstriction**
  - 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 << Aortic pressure
    - 15 mmHg vs. 60 mmHg
- **Blood flow**
  - Pulmonary = Aortic
- **Resistance:**
  - Pulmonary << Aortic Resistance

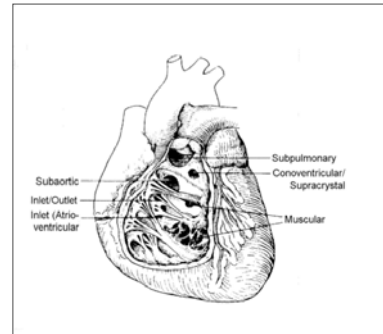
## Left to Right Shunt Lesions

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

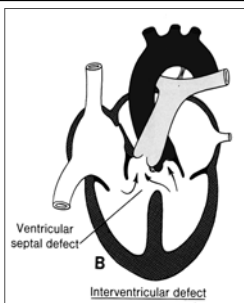
## “Top 4” Left to Right Shunt Lesions

- **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

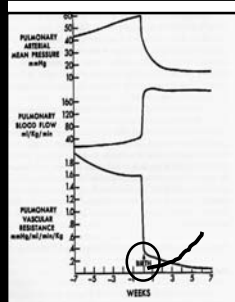


- Size of Defect
- Maturity of Pulmonary Bed
- Pressure difference between RV and LV
- Relative resistance between Pulmonary and Systemic circulations

### 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

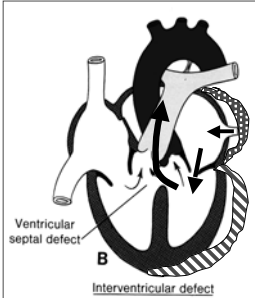


- Fetal: No shunt
- At Birth: No shunt
- Transition 1-7 weeks
  - PA/RV decreases to < LV
  - PA resistance decreases to < Systemic
  - L to R shunt increases

### 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

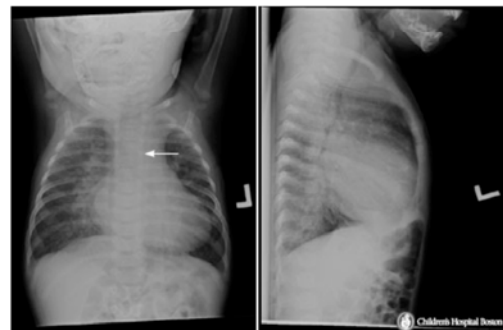
## Laboratory Findings: VSD

- 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

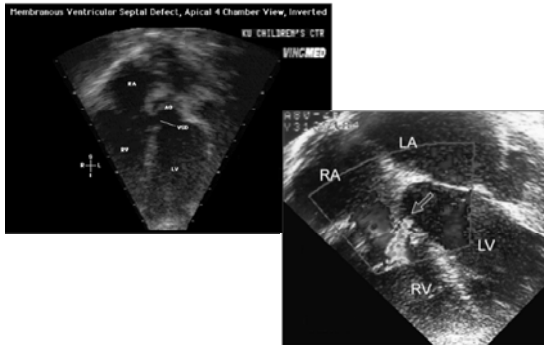
## Electrocardiogram: VSD



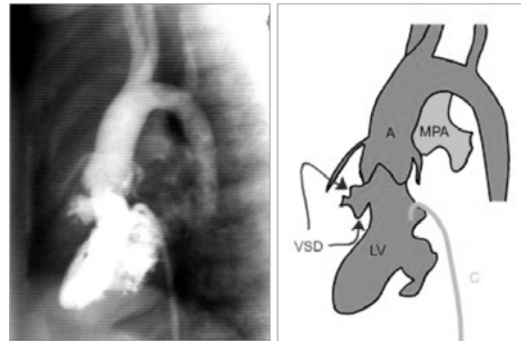
## Chest X-Ray: VSD



### Echocardiogram: Membranous VSD



### Angiogram: VSD



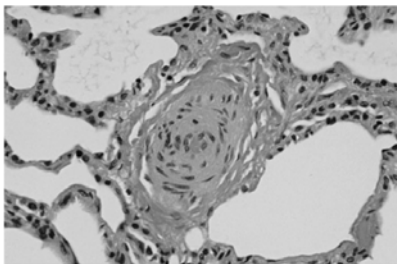
### Treatment of Large VSD

- 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

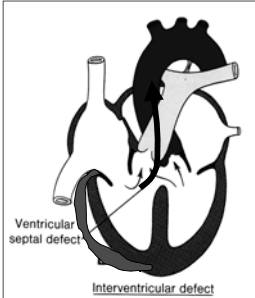
### Medial Hypertrophy



### Effect of Large Left to Right Shunt on Pulmonary Vascular Bed

- High pulmonary blood flow: Shear Stress
  - Medial hypertrophy
  - Endothelial damage
- $$\frac{\uparrow \Delta \text{ Pressure}}{\uparrow \text{ Resistance}} = \downarrow \text{ Blood Flow}$$
- 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

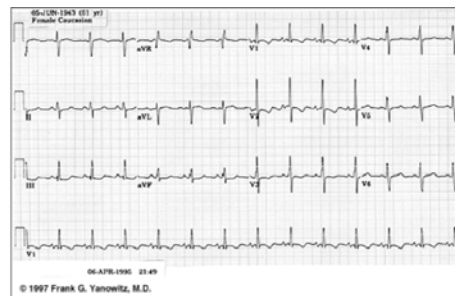
## Clinical Picture: Eisenmenger's

- 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

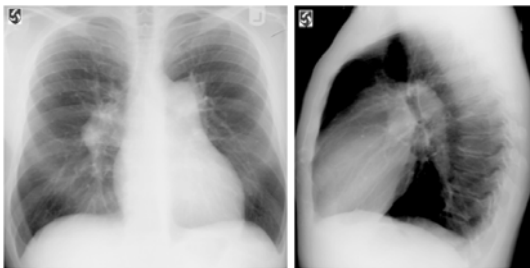
## Lab findings: Eisenmenger's

- No LV volume overload
- High RV pressure overload
- EKG: RVH ± 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
  - PGI<sub>2</sub>, Sildenafil
- Inotropic support
  - Right heart failure
- Transplant
  - Heart-Lung
  - Lung transplant, heart repair

## Patent Ductus Arteriosus (PDA)

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- 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

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

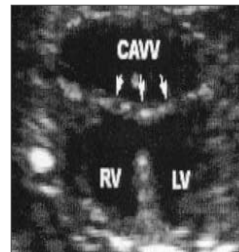
## PDA Coil Closure

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## Endocardial Cushion Defect

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

## Management: ECD

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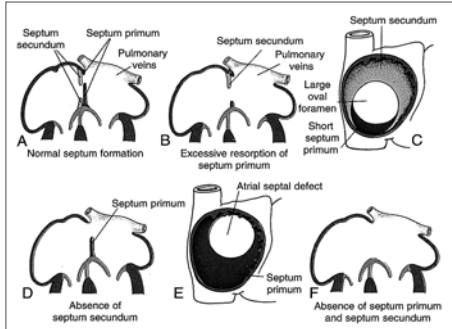
- Closure always indicated
- Timing of surgery (elective by 6 mos.)
  - Heart Failure
    - Large left to right shunt
    - Mitral insufficiency
  - Pulmonary hypertension
- Surgical repair
  - ASD, VSD closure
  - Repair of AV-Valves

## Summary: VSD, PDA and ECD

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- 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

