Pathophysiology: Left To Right Shunts

Daphne T. Hsu, MD
dh17@columbia.edu

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

\[
\text{Pressure} = \text{Flow} \times \text{Resistance}
\]

\[
\text{Resistance} = \frac{\Delta \text{Pressure}}{\text{Flow}}
\]
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 = \( \Delta \text{Pressure} \)
  - Flow
Transition from Fetal to Neonatal Circulation

- Pulmonary blood flow
- Pulmonary venous return
- Left atrial pressure
- Closure Foramen Ovale
- Arterial pO₂
- Closure Ductus Arteriosus

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

• Vasocostriction
  – 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

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**Ventricular Septal Defect**

- Subaortic
- Inlet/Outlet
- Inlet (Atrio-ventricular)
- Subpulmonary
- Conoventricular/Supracristal
- Muscular
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

\[ \Delta \text{Pressure} \uparrow = \downarrow \text{Blood Flow} \]
\[ \uparrow \text{Resistance} \]

• 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 ± 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, Sidenafil
- 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