

## Pediatric Congenital Heart Defects: Understanding the Basics

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<http://angiohealth.com/pregnancy/pregnancy-guide/pregnancy-health/miss-and-infections/congenital-heart-defects-know-baby-heart/>

## Learning Objectives

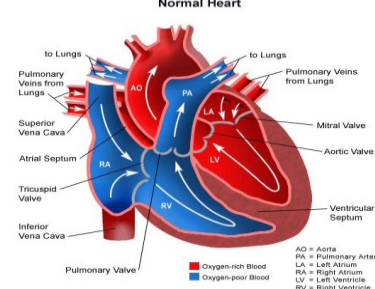
- Describe the differences between fetal and postnatal circulation
- Identify the etiology, signs and symptoms, and most common congenital heart defects
- Discuss the pharmacologic management of common congenital heart defects

POA: patent ductus arteriosus

## Disclosures

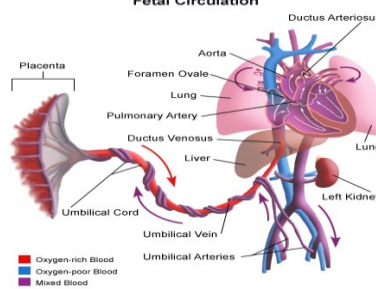
I have no financial nor affiliation based disclosures to make regarding to this lecture

## Normal Heart Physiology



AD = Aorta  
 PA = Pulmonary Artery  
 LA = Left Atrium  
 RA = Right Atrium  
 LV = Left Ventricle  
 RV = Right Ventricle

## Total Fetal Circulation

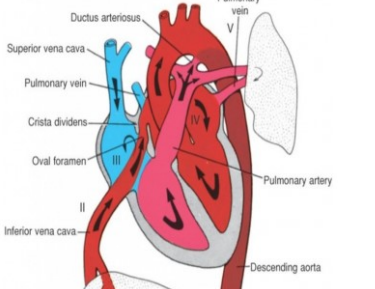


Ductus Arteriosus  
 Aorta  
 Foramen Ovale  
 Lung  
 Pulmonary Artery  
 Ductus Venosus  
 Liver  
 Left Kidney  
 Placenta  
 Umbilical Cord  
 Umbilical Vein  
 Umbilical Arteries

■ Oxygen-rich Blood  
 ■ Oxygen-poor Blood  
 ■ Mixed Blood

<http://www.stanfordchildrens.org/en/topic.do?id=9d7b2868-456d-4164-b919-a26434474842>

## Heart Fetal Circulation



Pulmonary vein  
 Ductus arteriosus  
 Superior vena cava  
 Pulmonary vein  
 Crista dividens  
 Oval foramen  
 Inferior vena cava  
 Pulmonary artery  
 Descending aorta

## Anatomical Shunts

- Ductus Venosus**
  - Bypasses liver
  - Connects umbilical vein and the inferior vena cava
- Foramen Ovale**
  - Bypasses lungs
  - Opening between the right and left atria
- Ductus Arteriosus**
  - Bypasses lungs
  - Connects pulmonary artery and the descending aorta

JPT: July-September 2004, Vol. 9, No. 3, pp. 160-178.

## Transitional Circulation

- SVR > PVR
  - Placenta removed
  - Ductus venosus closes
  - Fetal lung fluid resorbed causing air-induced lung expansion
- Pressure in RA < LA
  - Foramen ovale closes
- Flow through the ductus arteriosus L → R (aorta to PA)
  - Ductal constriction usually complete within 96 hrs

SVR: systemic vascular resistance, PVR: pulmonary vascular resistance, RA: right atrium, LA: left atrium, PA: pulmonary artery

JPT: July-September 2004, Vol. 9, No. 3, pp. 160-178.

## Congenital Heart Defects

- A defect in the structure and/or function of the heart due to abnormal development prior to birth
  - May or may not be detected prior to birth depending on severity of defect
  - CHD screening – pulse oximetry
- Over 35 different abnormalities currently identified
  - Cyanotic – resulting in “blue” skin due to lack of oxygenated blood in circulation
  - Acyanotic – “pink” skin due to adequate oxygenation of blood, but poor delivery

CHD: congenital heart defects

Genève: World Health Organization, 2011.  
JPT: July-September 2004, Vol. 9, No. 3, pp. 160-178.

## Incidence

- CHDs affect nearly 1% of—or about 40,000—births per year in the United States
  - Most common defect is VSD
  - About 25% of babies have critical CHDs requiring surgery or other measures
- Males > Females
- 4.2% of all infant deaths occurred due to CHD, and 48% of CHD related deaths occur within 1-year of birth

CHD: congenital heart defects

CHD: congenital heart defects

Birth Defects Research Part A: Clinical and Molecular Teratology 94:22 (2012): 970-983.  
JPT: July-September 2004, Vol. 9, No. 3, pp. 160-178.

## Etiology

- Most common birth defect noted worldwide (1.35 million babies per year)
- Risk factors associated with CHDs
  - Maternal illness (i.e. pregestational diabetes, phenylketonuria)
  - Maternal drug exposure
  - Maternal obesity
  - Non-Hispanic black
  - Trisomy 21
  - Assisted reproductive technologies (ART)
  - Etc.

CHD: congenital heart defects

ART: assisted reproductive technologies

Visual Guide to Neonatal Cardiology, 2013 Apr 18:43.

## Signs and Symptoms of CHD

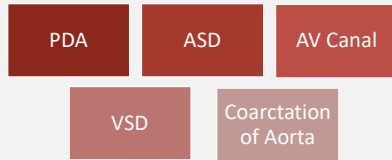
- Respiratory**
  - Tachypnea
  - Dyspnea
- Cardiovascular**
  - Tachycardia
  - Murmurs
  - Arrhythmias
- Cyanosis**
  - Mucous membranes
- Failure to thrive**
  - Poor feeding
- Hepatomegaly**
- Faint peripheral pulses**
- Decreased UOP**

CHD: congenital heart defects

CHD: congenital heart defects

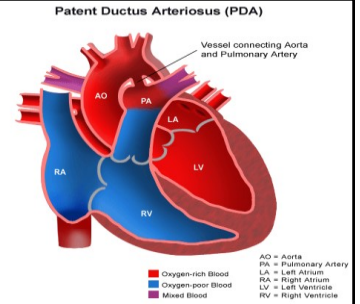
Genève: World Health Organization, 2011.  
JPT: July-September 2004, Vol. 9, No. 3, pp. 160-178.

## Acyanotic Congenital Heart Defects



PDA: patent ductus arteriosus; ASD: atrial septal defect; VSD: ventricular septal defect; AV canal: atrioventricular canal.

## Patent Ductus Arteriosus (PDA)



<http://photos.state.gov/libraries/california/online.com/Search/90302348>

## Patent Ductus Arteriosus

- Ductus Arteriosus (bypasses lungs in utero)
  - Connects pulmonary artery and descending aorta
- Remains patent in utero due to low oxygen tension and high levels of circulating PGEs
- At birth, oxygen saturation increases and PGE concentration decreases → constriction of the PDA
  - Functional closure within 96 hrs
  - Anatomic closure 2 weeks – 3 months

PGE: prostaglandin; PDA: patent ductus arteriosus

PPF: July-September 2004, Vol. 9, No. 1, pp. 160-178.  
Annals of cardiac anaesthesia, 2007; 11, 150-154.

## Patent Ductus Arteriosus

- Ductus remains patent in 3-8 per 10,000 live births worldwide
- Risk Factors:
  - Prematurity
    - 8 per 1,000
    - 80% of infants < 2200g (1 kg)
  - Respiratory distress syndrome
  - High altitude
  - Excessive fluid administration (>150 mL/kg/day)

PPF: July-September 2004, Vol. 9, No. 1, pp. 160-178.  
Annals of cardiac anaesthesia, 2007; 11, 150-154.

## Patent Ductus Arteriosus

- Ductus may be closed:
  - Pharmacologically
  - Surgical ligation
  - Trans-catheter device (coil)
- Only CHD where medications CORRECT the lesion
  - Indomethacin
  - Ibuprofen
  - Acetaminophen

PPF: July-September 2004, Vol. 9, No. 1, pp. 160-178.  
Annals of cardiac anaesthesia, 2007; 11, 150-154.

## Indomethacin

- Cyclooxygenase inhibitor
  - Inhibits prostaglandin synthesis
- Dose: 3 doses given at 12-24hr intervals (dependent on PNA)
- AE: Renal dysfunction, bleeding, NEC, hyperbilirubinemia
- No longer considered the "drug of choice"

PDA: patent ductus arteriosus; AE: adverse event; NEC: necrotizing enterocolitis

## Ibuprofen

- Cyclooxygenase inhibitor
- Now has become the "standard of care"
- May be associated with lower risk of NEC
- Dose: 10 mg/kg followed by 5 mg/kg at 24hr and 48hr post initial dose
- AE: renal dysfunction, thrombocytopenia, bleeding

AE: adverse events  
NEC: necrotizing enterocolitis

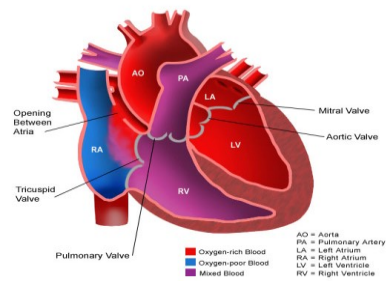
## Acetaminophen

- Blocks the peroxidase segment of prostaglandin synthetase
- FDA approved for PDA closure
- Much safer side-effect profile
  - Does not cause peripheral vasoconstriction
  - No effect on platelet aggregation
  - No effect on bilirubin displacement
- Dose: 15 mg/kg every 6 hrs (variable duration)

FDA: federal drug and food association  
PDA: patent ductus arteriosus

## Atrial Septal Defect (ASD)

### Atrial Septal Defect (ASD)



<http://homesteaders.org/wildlifedictionary.com/Search/95392346>

## Atrial Septal Defect (ASD)

- Opening in the septum between the two atria
- Blood shunts from the LA to the RA across the ASD
- Blood flow to the lungs is higher than normal
- Most patients are asymptomatic
- Surgical repair is required in most ASD
  - Left unrepaired, can lead to RV hypertrophy and CHF

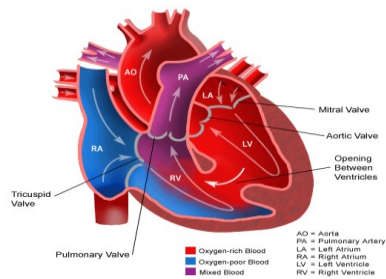


LA = left atrium, RA = right atrium  
CHF: congestive heart failure

BPPT: July-September 2004, Vol. 9, No. 3, pp. 160-174  
Annals of Cardiac Anesthesia, 2007, 9(1), 101-110

## Ventricular Septal Defect (VSD)

### Ventricular Septal Defect (VSD)



<http://homesteaders.org/wildlifedictionary.com/Search/95392346>

## Ventricular Septal Defect (VSD)

- Opening in the septum between the two ventricles
- Blood shunts from the LV to the RV across the VSD
- Blood flow to the lungs is higher than normal
- Small VSD may spontaneously close
- Moderate to large VSD require surgical repair
  - May lead to pulmonary over-circulation and CHF



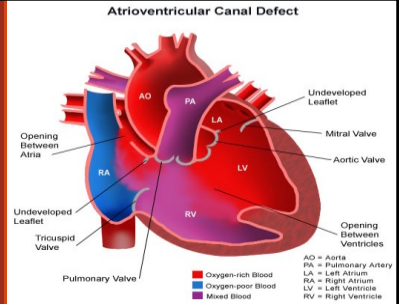
LV = left ventricle, RV = right ventricle  
CHF: congestive heart failure, VSD: ventricular septal defect

BPPT: July-September 2004, Vol. 9, No. 3, pp. 160-174  
Annals of Cardiac Anesthesia, 2007, 9(1), 101-110  
http://www.lippincott.com/lookup/author.aspx?authorid=104511&backlist=author&page=author&tab=author

## Pharmacotherapy Post-Operatively

- LV may be have mild hypertrophy
- Pulmonary vasculature requires time to heal
- Often require aggressive diuresis and blood pressure management
- Furosemide: loop diuretic (ascending loop of Henle)
  - 1-2 mg/kg/dose q6h-q24 IV/PO
- Chlorothiazide: thiazide-like diuretic (distal convoluted tubule)
  - 5-10 mg/kg/dose q6h-q24h IV
  - 10-20 mg/kg/dose q6h-q24h PO

## Atrioventricular Canal Defect (AVC)



## Atrioventricular Canal (AV Canal)

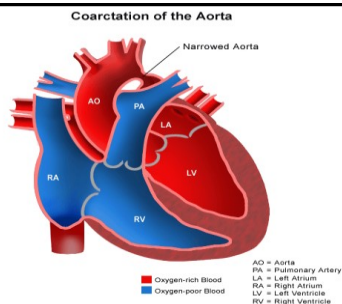
- Comprised of several abnormalities of structures inside the heart
- Atrial septal defect
- Ventricular septal defect
- Improperly formed mitral/tricuspid valve
- Blood flow to the lungs is **higher** than normal
- Decreased systemically circulated oxygenated blood

## Unbalanced Atrioventricular Canal (AV Canal)

- Left or right dominance occurs (aorta override)
- Often creates hypoplasia of one ventricle
- May require single ventricle physiologic approach
- Often results in pulmonary over circulation and damage to the pulmonary vasculature
- Afterload reduction
- Pulmonary artery band

Orlando et al. Semin. Thorac. Cardiovasc. Surg. 1997;9(1):1-5

## Coarctation of the Aorta (CoA)



## Coarctation of the Aorta (CoA)

- Narrowing or constriction of the aorta
  - LV outflow obstruction
  - Increased LV pressure
- Decreased peripheral perfusion and oxygenation
- Pulmonary congestion from high left-sided pressures
- Patient may remain asymptomatic with mild obstruction
  - Diagnosis typically made after birth

LV: left ventricle

1997;10(1):September 2004, 1(8), 6, No. 6, pp. 180-176  
Annals of cardiac anaesthesia, 2007 vol. 1, (01) 1-5

## Pharmacotherapy Post-Operatively

- LV is acclimated to pumping against coarctation
  - Often mild-moderate hypertrophy of LV
  - Autoregulation/remodeling is not immediate
- ACEi: inhibits the conversion of angiotensin I to angiotensin II
  - Captopril – approved for neonates and older (TID dosing)
  - Enalapril – approved for neonates and older (BID dosing)
  - Lisinopril – only approved for ≥ 6 years and older (once daily dosing)

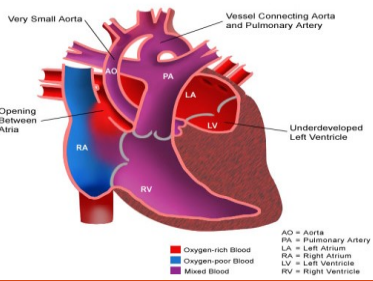
## Cyanotic Congenital Heart Defects



HLHS, hypoplastic left heart syndrome; ToF, tetralogy of Fallot; TGA, transposition of the great arteries.

## Hypoplastic Left Heart Syndrome (HLHS)

### Hypoplastic Left Heart Syndrome



<http://homechickfilms.com/wordpress/wp-content/uploads/downloads/2013/05/902284>

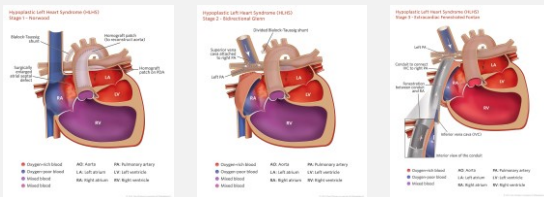
## Hypoplastic Left Heart (HLHS)

- The hearts left side (aorta, aortic valve, left ventricle, and mitral valve) are underdeveloped
- PDA IS ESSENTIAL FOR LIFE
- Three stage surgical correction
  - Norwood procedure
  - Bi-directional Glenn procedure
  - Fontan procedure
- Goal: create single ventricle physiology allowing the RV to pump oxygenated blood to the body

PDA, patent ductus arteriosus; RV, right ventricle.

JPTT, July-September 2004, Vol. 6, No. 3, pp. 100-171. *Journal of Pediatric Intensive Care*, 2007, Vol. 1, pp. 120-124.

## Hypoplastic Left Heart (HLHS)



<https://www.hopkins.com/treatments/medical-reconstruction-heart-surgery>

## Aspirin

- Inhibitor of COX-1 and COX-2 activity
  - Causes irreversible platelet inhibition
- Low-dose aspirin recommended for the prevention of thrombosis in patients with pulmonary artery shunts (i.e. s/p Norwood procedure)
  - Sano: 5 mg/kg
  - BT: 10 mg/kg
  - Heparin drip initiated immediately post-op

ST, Blomh, Tawing

## Digoxin

- Selective inhibitor of the membrane Na-K pump
  - Increases the force of contraction
  - Decreases automaticity of the AV node
- Use of digoxin at discharge s/p Norwood procedure has been associated with reduced interstage mortality
- 10 mcg/kg/day divided BID

<http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1089806/>

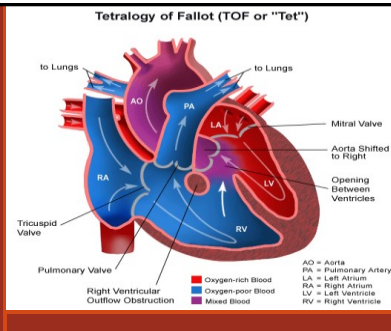
## Sildenafil

- Phosphodiesterase-5 inhibitor in smooth muscle of pulmonary vasculature
  - Results in increased cGMP concentrations and pulmonary vasculature relaxation
  - Vasodilation of the pulmonary bed
- Bi-directional Glenn creates passive blood flow from the IVC to the PAs
- High PVR can result in poor pulmonary blood flow and saturations

[cGMP, cyclic guanosine monophosphate, IVC inferior vena cava, PA pulmonary artery, PVR pulmonary vascular resistance](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1089806/)

[Mori et al. Int J Cardiol. 2004;231:122-7](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1089806/)

## Tetralogy of Fallot (TOF)



## Tetralogy of Fallot

- "Tetralogy" – 4 anatomical defects
  - VSD
  - Pulmonary artery stenosis
  - Aorta overriding both ventricles
  - RV hypertrophy
- Decreased pulmonary blood flow
- Severe obstruction leads to cyanosis at birth requiring immediate surgical repair and PGE1 therapy to maintain a patent ductus arteriosus



[VSD, ventricular septal defect, RV, right ventricle, PVR, pulmonary resistance](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1089806/)

[PFT, July-September 2004, Vol. 9, No. 3, pp. 185-178, Annals of Thoracic Medicine, 2004;9:185-195](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1089806/)

## Tetralogy of Fallot

- ToF can vary depending on severity of obstruction
- "Tet" spells or infundibular spasms
  - Hypercyanotic crisis
  - Triggered by increased oxygen demand > supply
- Signs and symptoms
  - Crying/agitation
  - Poor feeding
  - Fever



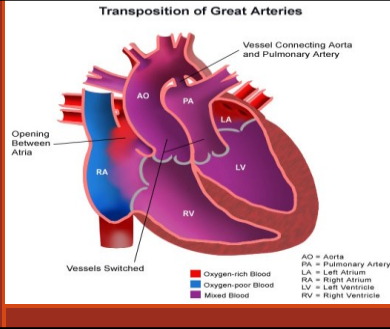
<http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1089806/>

## Treatment of "Tet" Spells

- Increase the SVR to increase pulmonary blood flow
  - Knee to chest
  - Fluid bolus
  - IV phenylephrine
- Decrease PVR to increase pulmonary blood flow
  - Supplemental oxygen
- IV / PO propranolol
  - Decrease infundibular tone
  - Increase pre-load via improved diastole

[SVR, systemic vascular resistance, PVR, pulmonary vascular resistance](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1089806/)

## Transposition of the Great Arteries (TGA)



## Transposition of the Great Arteries

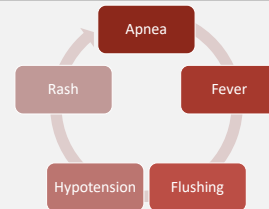
- Two parallel circuits
- Aorta arises from the RV
  - Causes deoxygenated blood to be pumped to the circulation
- Pulmonary artery arises from the LV
  - Causes oxygenated blood to be pumped to the lungs
- Not compatible with life without shunting
  - ASD and PDA
- Requires surgical correction: arterial switch

RV = right ventricle, LV = left ventricle, ASD = atrioventricular septal defect, PDA = patent ductus arteriosus

## Prostaglandin E<sub>1</sub> (PGE<sub>1</sub>)

- Alprostadil
- Synthetic prostaglandin
  - Direct effect on vascular and ductus arteriosus smooth muscle
  - Allows the ductus arteriosus to remain patent
- Short  $t_{1/2}$ 
  - Administer as continuous infusion through dedicated line
- Initial dose: 0.05 mcg/kg/min
  - Range: 0.01 – 0.4 mcg/kg/min
  - Titrate to response

## Prostaglandin E<sub>1</sub> (PGE<sub>1</sub>)



## Congenital Heart Defects

|        |        |               |                   |
|--------|--------|---------------|-------------------|
| DORV   | DILV   | TAPVR         | Ebstein's anomaly |
| AAORCA | AAOLCA | Vascular ring | PA-IVS            |

## Summary

- Described the differences between fetal and postnatal circulation
- Identified the etiology, signs and symptoms, and most common congenital heart defects
- Discussed the pharmacologic management of common congenital heart defects

PDA = patent ductus arteriosus