Cardiovascular Pathophysiology: Right to Left Shunts

aka Cyanotic Lesions

Ismee A. Williams, MD, MS
iib6@columbia.edu
Pediatric Cardiology

What is Cyanosis?

- Bluish discoloration of skin that occurs when the amount of deoxygenated hemoglobin ≥ 5 g/dL in capillaries
- Central Cyanosis: decreased systemic oxygen delivery
- Peripheral Cyanosis: increased oxygen extraction by tissue

Learning Objectives

- To discuss the hemodynamic significance of right to left shunts
- To describe the common cyanotic cardiac lesions in the newborn
- To understand the different causes of cyanosis: obstruction to pulmonary blood flow vs mixing

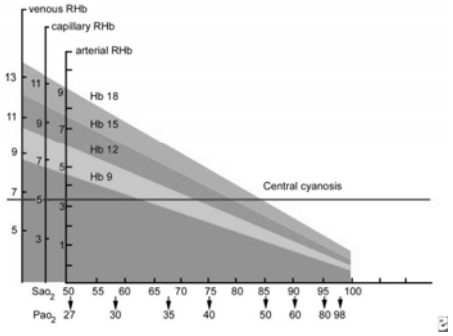
Factors affecting detection of Cyanosis

- **Total hemoglobin concentration** affects the level of O2 saturation at which cyanosis is observed
  - Hgb conc = 9 g/dL, need an O2 Sat of 67% to have 3-5 g/dL of reduced hemoglobin and ***see cyanosis***
  - Hgb conc = 20 g/dL, see cyanosis at O2 Sat of 85%
  - ***Decreased O2 sat may not be recognized in the setting of anemia***
- **Skin pigmentation**
- **Factors that shift the oxygen dissociation curve** to the left result in oxygen binding more tightly to Hgb and decreased release to the tissue at a given O2 tension (PO2)
  - Therefore, will be harder to ***see cyanosis*** (get 5 g/dL of deoxygenated Hgb) at any given PO2

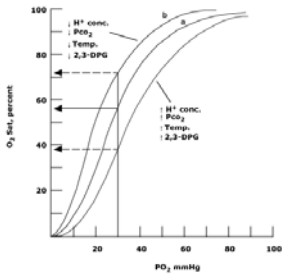
Importance of Congenital Heart Disease

- Incidence 6 to 8 per 1000 births
- 15% are ***life threatening***
- 25% are discharged without diagnosis
- 1/3 have cyanosis

Won't see cyanosis if anemic



Detecting Cyanosis



- Shift to the left (harder to see cyanosis): hyperventilation, hypothermia, and low 2,3 diphosphoglycerate, fetal Hgb
- Shift to the right (easier to see cyanosis): acidosis, fever, or increased adult hemoglobin

Cardiac Causes of Cyanosis

- Decreased/obstructed pulmonary blood flow
- Systemic and Pulmonary venous Mixing

Causes of Cyanosis

- Pulmonary causes (most common)
- Hemoglobin problems
- Poor perfusion (sepsis)
- PPHN
- Cardiac causes

Decreased Pulmonary Blood Flow

- Obligatory intracardiac right to left shunting
- Pulmonary blood flow is provided by an alternative path – usually the ductus arteriosus
- Very cyanotic

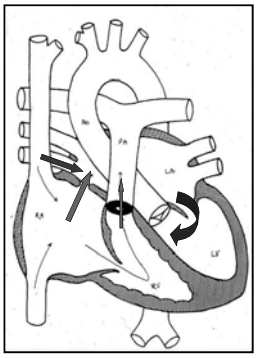
Persistent Pulmonary Hypertension of the Newborn

- Used to be called Persistent Fetal Circulation
- Abnormal pulmonary vasoconstriction or failure to “relax” leads to right to left shunting at the foramen ovale and the ductus arteriosus
- Profound cyanosis
- Associated with neonatal asphyxia, maternal infection
- Apgar scores are low
- Usually self-limited with NO and ECMO treatment

Cardiac Lesions causing cyanosis due to decreased pulmonary blood flow

- Pulmonary stenosis
- Pulmonary atresia
- Tricuspid atresia
- Tetralogy of Fallot

Pulmonary Stenosis



- Location of obstruction varies:
- RV outflow
- Pulmonary Valve Most common
- Main Pulmonary Artery

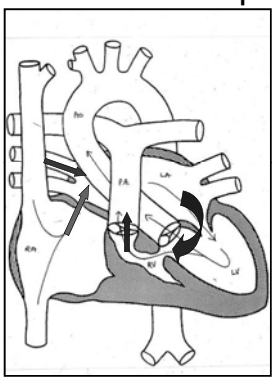
Pulmonary atresia

- 3% of CHD (0.041 per 1000 live births)
- Size of the RV varies
- PE: cyanosis, no systolic ejection murmur (no flow)
  - may have holosystolic murmur at LLSB associated with tricuspid regurgitation
- CXR: black lungs
- Treatment depends on "flavor" of PA/IVS
  - balloon of pulmonary valve if RV size adequate
  - aortico-pulmonary shunt to increase pulmonary blood flow
  - staged surgery to a Fontan if RV too small
  - Heart transplant if RV dependent coronary sinusoids

Pulmonary Stenosis

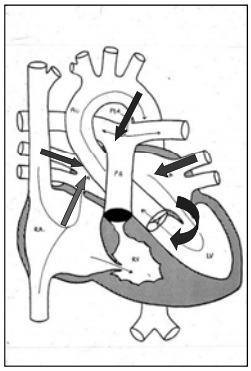
- 25-30% of CHD
  - Isolated PS in 8-10% of CHD
- Hemodynamic consequence: pressure overload and hypertrophy of the RV
- PE: cyanosis, systolic ejection murmur at LUSB
- Tx: Balloon vs surgery

Tricuspid Atresia



- Obligatory right to left shunt at the PFO
- Typically have a VSD that allows blood into the RV and out the PA
  - Obstruction to pulmonary flow related to size of VSD
- Hypoplastic right ventricle

Pulmonary Atresia

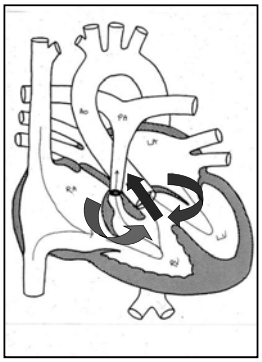


- Obligate right to left flow across the foramen ovale
- Pulmonary blood flow supplied by the ductus arteriosus *"ductal dependent"*

Tricuspid Atresia

- 3% of CHD (0.056 per 1000 live births)
- 25% have transposed great vessels and problems with aortic/systemic blood flow
- PE: systolic murmur, cyanosis
- Tx: staged surgery to a Fontan

Tetralogy of Fallot



Single defect: anterior malalignment of the interventricular septum

- VSD
- Aortic override
- Pulmonary Stenosis
- RVH

Mixing of Systemic and Pulmonary Venous Return

- No obstruction to pulmonary blood flow
  - Pulmonary flow may be greater than normal
- See both right to left AND left to right intracardiac shunting
- Associated with pulmonary HTN and ventricular failure
- Cyanosis typically less intense than with pulmonary obstruction

Tetralogy of Fallot

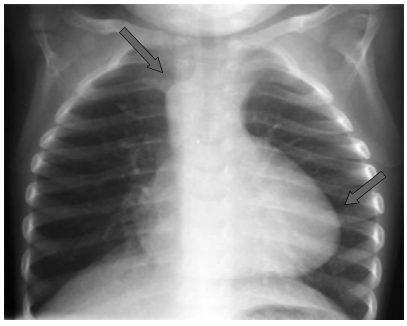
- 3.5-9% of CHD (0.26-0.8 per 1000 live births)
- Commonly associated with other defects
  - DiGeorge Syndrome in 25%
- Degree of pulmonary obstruction varies
- Symptoms depend on amount of obstruction to pulmonary blood flow
  - cyanosis, ***tet spells***
- PE: systolic ejection murmur at LUSB
- Tx: Surgical repair of VSD and PS

Cyanosis due to Mixing

- Truncus arteriosus
- Total anomalous pulmonary venous return (TAPVR)
- Transposition of the Great Arteries (TGA)

- Mixing with Heart Failure
  - HLHS, Aortic stenosis, coarctation

Boot shaped heart = TOF

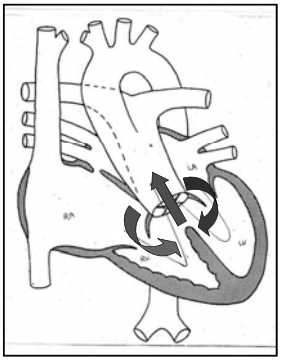


Upturned cardiac apex due to RVH

Right aortic arch

Lungs hyperinflated (black) due to decreased blood flow

Truncus Arteriosus

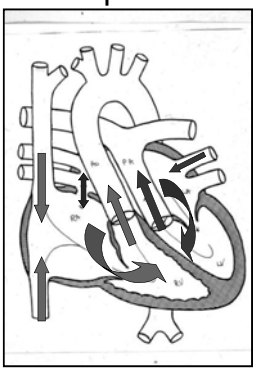


- Aorta and pulmonary artery not separate
- Single vessel gives rise to aorta, coronaries, and pulmonary arteries
- VSD always present
- Systemic and pulmonary venous blood mix at the ventricular level

Truncus arteriosus

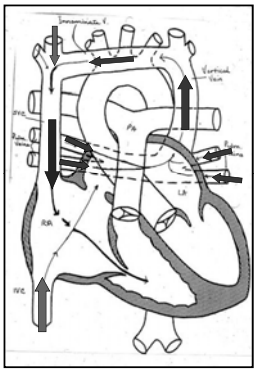
- 1-2.5% of CHD (0.08 per 1000 live births)
- Truncal valve usually very dysplastic
- Commonly associated lesions
  - Coronary anomalies, interrupted aortic arch
  - 25% DiGeorge
- PE: cyanosis and murmur of regurgitation
- High risk to develop pulm HTN over time
- Tx: surgical repair in infancy

Transposition of the Great Arteries



- Great arteries are "switched"
- Systemic venous return goes back to the body
- Pulmonary venous return goes back to the lungs
- Survival dependent on mixing between the two parallel circulations

TAPVR



- Pulmonary veins return to the right heart
  - Via supracardiac, intracardiac, or infradiaphragmatic path
- Pulmonary venous blood mixes with systemic venous blood at the atrial level
- Obligatory right to left shunt at atrial level to support systemic flow

Transposition of the Great Arteries

- Most common cyanotic CHD (0.22 per 1000 live births)
- Fetal circulation allows mixing
- Problems after birth
- Mixing via PFO/ASD, VSD (1/3), or PDA
- PE: severe cyanosis, no murmur
- Tx: balloon atrial septostomy to maximize mixing at the atrial level
  - surgical arterial switch

TAPVR

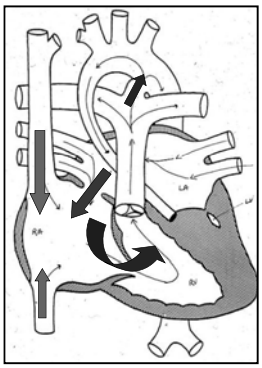
- 2-3% of CHD (0.058 per 1000 live births)
- Failure of the left atrium to incorporate the pulmonary veins during development
- Obstruction to pulmonary venous flow is common
  - Can occur at different levels
  - Most common in infradiaphragmatic TAPVR
  - Leads to pulmonary congestion and death
- PE: cyanosis, respiratory distress, CXR white out with small heart
- Tx: no PGE, surgical repair

Cyanosis due Mixing with Heart Failure

- Obstruction to systemic outflow, mixing, cyanosis, poor perfusion
- Depend on PDA to supply systemic blood flow
- As PDA closes, see poor perfusion, acidosis, death

- Hypoplastic left heart syndrome (HLHS)
- Critical valvar Aortic Stenosis
- Interrupted aortic arch/Coarctation of the Aorta

HLHS



- Left side of the heart too small/absent
- Classic form is mitral and aortic atresia
- Pulmonary venous blood shunts left to right at PFO and mixes with systemic venous return
- Blood going out the RV into the PA passes through the PDA to feed the body

Hyperoxia Test: Heart vs Lungs?

- Cardiac lesions typically have fully saturated pulmonary venous blood
  - High FiO2 has little effect on PO2 and O2 Sat
- Pulmonary lesions typically have pulmonary venous desaturation
  - Higher FiO2 increases pulmonary venous oxygen levels and PO2 and O2 Sat
- Administer 100% FiO2 for 10 minutes and compare the PO2 at baseline and after oxygen
  - PO2 > 150 mm Hg = pulmonary cause
  - PO2 < 150 mm Hg = cardiac cause

HLHS

- 0.16-0.27 per 1000 live births
- Severe form of single ventricle
- PE: no murmur, cyanosis, poor pulses
- Tx: PGE, Surgery: Norwood, Glenn, Fontan

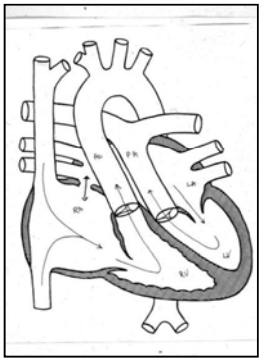
Case presentation

- Called to the nursery to evaluate a 3.5 kg product of a NSVD born at 39 wk GA to a 35 yo G2P1
- APGARS 9 and 9
- At four hours of life RN noted the infant appeared “dusky”
- Central cyanosis, no tachypnea, no murmur
- O2 Sat = 70%, PO2 = 40 mm Hg on RA, and O2 Sat = 82%, PO2 = 50 mm Hg on 100% FiO2
- CXR NL

Evaluation of the cyanotic newborn

- History: family hx, prenatal testing, peripartum information
- Vital Signs: HR, RR, O2 sat, 4 ext BP
- Physical exam: observation of skin, movement, respirations, palpation and ausculation of chest, palpation of femoral pulses, capillary perfusion
- Laboratory testing: ABG, CBC, BLCx, CXR, EKG, Echo

Transposition of the Great Arteries



- Prostaglandin E1
- Emergent balloon atrial septostomy
- O2 sat increases to 85%
- Arterial switch operation next day

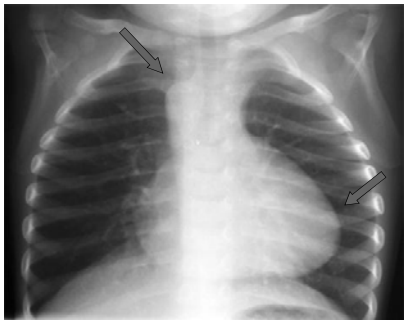
Case presentation

- Called to the nursery to evaluate a 3.5 kg product of a NSVD born at 39 wk GA to a 35 yo G2P1
- APGARS 9 and 9
- Murmur heard on discharge exam
- No tachypnea, loud SEM at LUSB
- O2 Sat = 90%
- CXR?
- Echo?

Case presentation

- Get a call from an outside pediatrician
- 10 day old infant with grunting and poor perfusion – presumed sepsis
- APGARS 9 and 9, no prenatal US
- In ER: Grey infant, O2 Sat = 90%, no femoral pulses, no murmur
- Echo?

Boot shaped heart = TOF

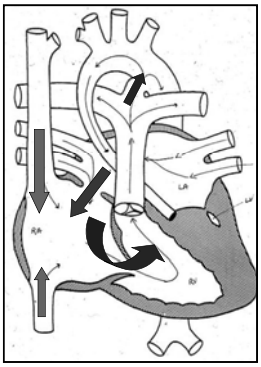


Upturned cardiac apex due to RVH

Right aortic arch

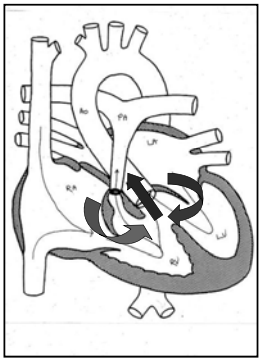
Lungs hyperinflated (black) due to decreased blood flow

HLHS



- Prostaglandin E1
- Pressors
- Intubate FiO2 21%
- Sedate and hypoventilate
- Norwood Stage I when stable

Tetralogy of Fallot



Educate parents about tet spells

Genetic testing for DiGeorge

Frequent follow up to check O2 sat

Plan elective surgical repair at 4 - 6 months

Summary

- Cyanosis when 3-5 gm/dl of desaturated Hgb – hard to see if anemic
- Mutliple causes
- Cardiac causes are EMERGENCIES
- Decreased pulmonary blood flow vs Mixing
- Prostoglandin E2 to keep ductus arteriosus OPEN