Cardiovascular Disorders

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I have no conflict of interest to disclose.

Objectives

1. Discuss the major differences between fetal circulation and extrauterine circulation and the transition to extrauterine life.
2. Differentiate between central versus peripheral cyanosis.
3. Differentiate between cyanosis that is cardiac in origin and that which is pulmonary in origin.
4. Differentiate between cyanotic versus acyanotic disease and be able to identify specific lesions as such.
5. Define and describe the anatomy, clinical manifestations, management and outcome of AV canal, coarctation of the aorta, hypoplastic left heart syndrome (HLHS), pulmonary stenosis and atresia, tetralogy of fallot (TOF), transposition of the great vessels, total anomalous pulmonary venous return (TAPVR), ventricular septal defect (VSD) and patent ductus arteriosus (PDA).
6. Discuss treatment modalities for congestive heart failure.
7. Discuss diagnostic criteria, causes and management of hypertension in the neonate.
8. Discuss the classification, presentation and management of shock.
9. Discuss the presentation and management for the most common arrhythmias among the neonatal population.
10. Discuss the presentation and management of cardiac tamponade.
11. Discuss the important aspects of a thorough cardiovascular assessment.

Introduction

- According to the CDC
  - Congenital Heart Defects (CHD) are the most common types of birth defects
  - In the US, CHDs affect nearly 1% of births per year or about 40,000/yr
  - The prevalence of some CHDs, particularly mild types, is increasing, while the prevalence of others has remained stable
  - The most common type of heart defect is a ventricular septal defect (VSD)
  - About 25% of babies with a CHD have a critical CHD – these babies generally require surgery or other procedures in the 1st year of life
  - It is estimated that about 1 million U.S. children and about 1.4 million U.S. adults are living with CHD
  - CHDs are a leading cause of birth-defect-associated illness and death
    - *CHD accounts for about 4.2% of all neonatal deaths*

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Introduction

- Survival
  - Non-critical CHD
    - *About 97% of babies born with a non-critical CHD are expected to survive to 1 year of age and about 95% of them are expected to live survive to 18 years of age*
  - Critical CHD
    - *About 75% of babies born with critical CHD are expected to survive to 1 year of age and about 69% of them are expected to survive to 18 years of age*

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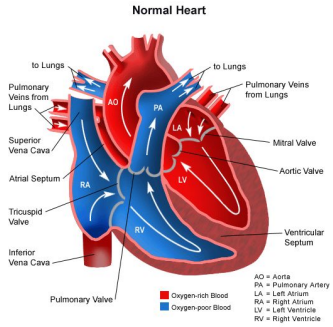
Fetal Circulation vs Extrauterine Circulation

Lets review …….



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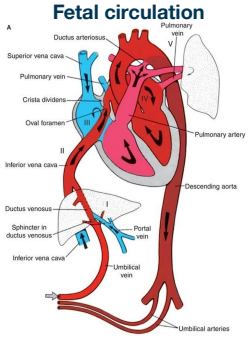
Fetal Circulation vs Extrauterine circulation

Fetal circulation



- Blood from the **lower body** and the **placenta** (better oxygenated blood) drains into the inferior vena cava (IVC); 1/3 of the IVC blood travels directly across the foramen ovale and leads to a greater O2 saturation in the cerebral and coronary vasculature (supplied by the left ventricle (LV))
- Blood from the **upper body** drains into the superior vena cava (SVC), and then mostly travels to the right ventricle (RV); blood then flows through the pulmonary arteries (PA) and the patent ductus arteriosus (PDA) to the descending aorta
  - Only ~10% of blood travels to the lungs

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Fetal circulation

In fetal circulation:

- The RV is the dominant ventricle supplying more cardiac output (CO) compared to the left ventricle (LV)
- The RV supplies the **descending** aorta (placental and lower body)
- The LV supplies the **ascending** aorta (upper body)
- The placenta receives the greatest percentage of fetal cardiac output (45%) and is the organ of **lowest** vascular resistance
- The lungs receive only a small amount of cardiac output (~5-15%)
- The PDA remains patent in utero secondary to (1) prostaglandins (PGE2), (2) prostacyclin (PGI2), and (3) thromboxane A2
  - *Of note…. PGE2 maintains patency of PDA in utero; however, PGE1 is the IV prostaglandin form administered to maintain ductal patency after birth*
- The amount of blood flow across the tricuspid valve > blood flow cross the mitral valve
  - *After birth, blood flow across these valves is equal (mostly)*

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Fetal circulation

Intrauterine oxygenation

- The fetus lives in a relatively hypoxic environment because:
  - The placenta does not exchange O2 as well as the (postnatal) lungs
  - Oxygenated maternal blood 1st supplies the uterine wall with blood flowing freely around the villi, mixing with deoxygenated blood
- The highest oxygen content of the fetus is in the umbilical **veins** (pO2=27-37torr, O2 saturation 70%)
- The oxygen content in the SVC is low (pO2=12-14torr, O2 sat 40%) because of the high extraction by the brain
- Remember…blood from the IVC (better oxygenated) is diverted via the foramen oval to the left side of the heart; therefor, the O2 saturation in the brain and coronary arteries is greater than the post-ductal blood supplied to the body

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Fetal circulation

The fetus tolerated lower pO2 because:

- Fetal hemoglobin has a higher affinity for O2; therefor, a fetus can tolerate a lower pO2
- Increased O2 carrying-capacity because of elevated hemoglobin concentrations in the fetus as a result of hypoxic induction of erythropoietin
- Increased ability to utilize glucose by anaerobic metabolism

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Hemodynamic changes at birth

At delivery:

- Umbilical arteries constrict, preventing blood flow from baby to mother
- Umbilical vein remains dilated, allowing blood to flow in the direction of gravity
- A delay in cord clamping, will increase the infant's blood volume

After delivery:

- Function of gas exchange changes from the placenta to the lungs
- Systemic vascular resistance (SVR) increases due to the removal of the placenta
- Ductus venosus closes
- Expansion of lungs ⇒ **decreased** pulmonary vascular resistance (PVR) and increased pulmonary blood flow
  - *With increased pulmonary venous return, the left atrial pressure increases and becomes greater than the right atrial pressure, leading to closure of the PFO*
- PDA closes as arterial O2 saturation increases and ductus becomes less responsive to PGE

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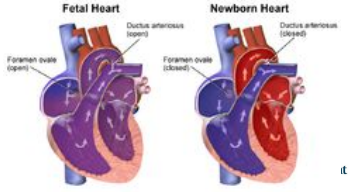
Fetal vs Newborn circulation

Fetal	Infant
▪ Low pressure system ▪ Right-to-left shunt ▪ Lungs non-functional ▪ Increased pulmonary resistance ▪ Decreased systemic resistance	▪ High pressure system ▪ Left-to-right blood flow ▪ Lungs functional ▪ Decreased pulmonary resistance ▪ Increased systemic resistance



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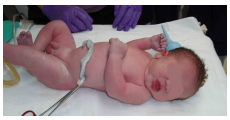
Cyanosis

Central

- Caused by reduced arterial oxygen saturation
- In newborns, mucous membranes appear blue
- Newborns normally have central cyanosis up to 5-10 minutes after birth
- Persistent central cyanosis should be evaluated

Peripheral

- Normal systemic arterial oxygen saturation and increased tissue oxygen extraction ⟹ increased concentration of reduced hemoglobin on the venous side of the capillary bed
- Typically affects the distal extremities
- In newborns, the mucous membranes remain pink



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Cyanosis

How do you differential between cyanosis that is cardiac in origin and that which is pulmonary in origin?

HYPEROXIA TEST

- Measure paO2 (preferably preductal) following administration of 100% FiO2
  - If there is **NOT** a substantial increase in PaO2, consider **cardiac** etiology
    - *And infant with a paO2 < 100 on 100% FiO2, normal PaCO2, and without severe lung disease/distress, consider cardiac disease*
  - If the paO2 is > 250 on 100% FiO2, structural heart disease is unlikely = **pulmonary** in origin
  - If the paO2 is between 100-200 on 100% FiO2, might have a complete **mixing** defect

*** May not always be able to rule out CHD with a hyperoxia test ***
When in doubt, obtain an echocardiogram

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Congenital Heart Disease

Cyanotic

5 Ts + 1

- Truncus arteriosus- 1-4%
- Transposition of the great vessels (TGA)-5-10%
- Tricuspid atresia (TA)
- Tetralogy of Fallot (TOF)- 8-10%
- Total anomalous pulmonary venous return (TAPVR)- 1-2.5%
- Hypoplastic left heart syndrome (HLHS)- 1.5%
- Others: Double outlet right ventricle (DORV), Ebstein's anomaly, single ventricle, pulmonary atresia (PA)

Acyanotic

Left-to-right shunt

- Ventricular septal defect (VSD)- 16%
- Atrial septal defect (ASD)- 6-11%
- Patent ductus arteriosus- 4-10%
- Complete AV canal (endocardial cushion defect)- 2-5%
- Partial anomalous pulmonary venous return (PAPVR)

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Cyanotic Heart Defects

What makes it a cyanotic defect?

Right-to-left shunt

- Blue deoxygenated blood from the IVC/SVC flows from the right side of the heart to the left side of the heart (LA/LV/aorta) instead of to the pulmonary arteries
  - Can occur at the PDA if PPHN is present
  - Can occur as a result of a cyanotic heart defect

Pre-ductal saturations > Post-ductal saturations

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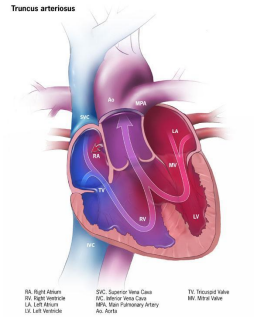
Truncus arteriosus



- Aorta, pulmonary arteries, and coronary arteries arise from single arterial blood vessel
- Truncus sits over a large VSD – complete ventricular mixing

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Truncus arteriosus

Clinical manifestations

- Mild to moderate cyanosis
- Congestive heart failure
- Wide pulse pressure
- Bounding arterial pulses
- Loud pansystolic murmur loudest at left lower sternal border
- CXR:
  - Increased heart size, increased pulmonary vascular markings
  - Right arch 30%
- Associated with DiGeorge syndrome (30%)

Management

- Management of congestive heart failure
- Early and complete surgical repair
  - Closure of the VSD
  - PA detached from truncus
  - Right ventricle to PA conduit
- PGE1??
  - Not useful because in most cases there is no PDA

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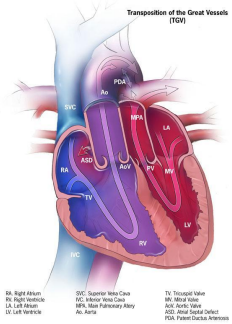
D-Transposition of the Great Arteries



- Aorta arises from the RV
- PA arises from the LV
- 2 circulations in parallel
  - Systemic venous blood returns to the right heart, and is pumped back to systemic circulation
  - Pulmonary venous blood returns to left heart, and is pumped back to the lungs
- Necessary to have a shunt, allowing communication between the 2 circulations
  - Ventricular, atrial, and/or patent ductus level
  - Best mixing at the atrial septum
  - PGE1 useful to maintain PDA
- VSD in 50%
- Pulmonary stenosis (PS) in 30-35%

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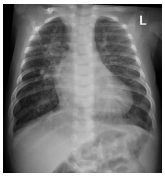
D-Transposition of the Great Arteries

Clinical manifestations

- Large, male infant
- Marked cyanosis
- "happy tachypnea"
- Congestive heart failure
- No murmur, unless VSD or PS
- CXR: **"Egg on a string"**



Management

- PGE1
- Balloon atrial septostomy
- Surgery
  - Arterial switch (during 1st 2 weeks of life)

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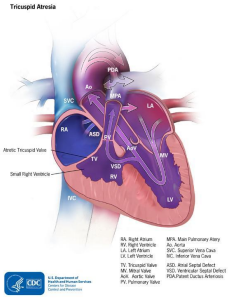
Tricuspid Atresia



- Complete absence of the tricuspid valve
- Hypoplastic right ventricle
- TA with minimal VSD:
  - Leads to poor RV development because of lack of blood flow in utero; ductal dependent; requires a shunt
- TA with VSD
  - Size of VSD determines size of RV (the greater L-R shunt across VSD, greater RV size)
- Can have pulmonary stenosis (PS) or pulmonary atresia (PA)
- Usually PDA dependent

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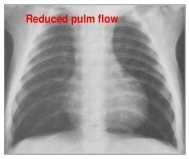
Tricuspid Atresia

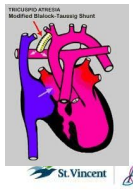
Clinical manifestations

- Cyanosis at birth
- Moderate systolic murmur and single S2
- May develop CHF
- CXR:
  - Minimal cardiomegaly
  - Decreased pulmonary vascular markings



Management

- PGE1
- Surgical repair usually consists of modified BT shunt
  - Gore-Tex tube that attaches a section of the aorta to the pulmonary artery to increase pulmonary blood flow
  - Palliative procedure



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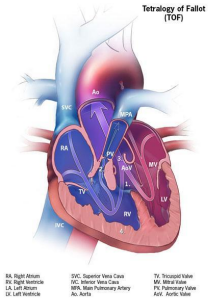
Tetralogy of Fallot (TOF)



RA, Right Atrium
RV, Right Ventricle
LA, Left Atrium
LV, Left Ventricle
SVC, Superior Vena Cava
IVC, Inferior Vena Cava
MPA, Main Pulmonary Artery
Ao, Aorta
TV, Tricuspid Valve
MV, Mitral Valve
PV, Pulmonary Valve
AoV, Aortic Valve

- 4 components:
  - Pulmonary stenosis
  - Large VSD
  - Overriding aorta
  - Right ventricular hypertrophy

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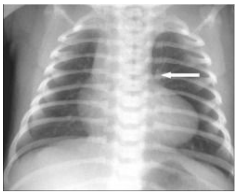
Tetralogy of Fallot (TOF)

Clinical manifestations

- "Cyanotic" TOF
  - "Blue Tet"
  - Cyanosis (mild-severe); depends on degree of RV outflow tract obstruction
  - Typically, no CHF
- "Acyanotic" TOF
  - "Pink Tet"
  - Large VSD, mild RV outflow tract obstruction
  - Usually present with CHF from large L-R ventricular shunt
- Harsh murmur of PS

CXR

- Heart size usually normal
- "boot shaped" heart



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Tetralogy of Fallot (TOF)

Tet spell

- Rapid and deep respirations
- Irritability
- Cyanosis
- Decreasing intensity of heart murmur
- Hypoxic spell often precipitated by stooling/crying
  - Increased PVR, decreased SVR
  - Increased R-L shunt → cyanosis
  - Decreased PBF
  - Decreased pO2, acidosis, increased pCO2

Management

- Knee-chest position
- Morphine (affects CNS and breaks cycle of hypoxia and agitation)
- Oxygen
- Vasoconstrictors to increase SVR
- Propranolol or Esmolol – decreases "spasm" of RVOT
- Expand intravascular volume with fluid boluses to dilate RV and create retrograde flow

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Tetralogy of Fallot (TOF)

Management

- PGE1 ("blue TET")
- BT shunt
- Complete surgical repair with VSD closure and relief of RVOT obstruction

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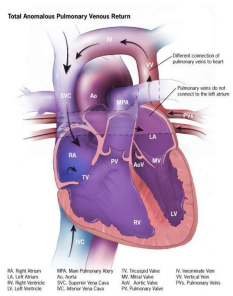
Total Anomalous Pulmonary Venous Return (TAPVR)



- All pulmonary veins with fully oxygenated blood follow an abnormal route returning to RA instead of LA
- Secundum ASD or PFO is required for R-L shunt to provide systemic blood flow
- 2 types:
  - Obstructed: when pulmonary venous blood takes a route below the diaphragm; infracardiac (subdiaphragmatic)
  - Unobstructed: cardiac and supracardiac

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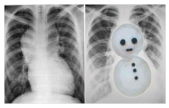
Total Anomalous Pulmonary Venous Return (TAPVR)

Clinical manifestations

- Varying degrees of cyanosis
- Obstructive:
  - Severe CHF
  - Moderate to severe cyanosis
  - Surgical emergency
- Unobstructed:
  - Mild to moderate CHF
  - Mild to moderate cyanosis
  - Surgery in 1st 6 months
- CXR
  - Moderate to marked cardiomegaly
  - "snowman" sign (with supracardiac type)



Management

- Management of CHF
- Require an ASD
- Surgery – re-implantation of pulmonary veins and closure of ASD
  - Urgent for obstructed type
- PGE1 not helpful
  - Could make the patient worse by worsening pulmonary edema

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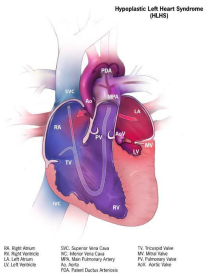
Hypoplastic Left Heart Syndrome (HLHS)



- Mitral valve hypoplasia/atresia
- Hypoplastic left ventricle
- Aortic hypoplasia/atresia
- Hypoplastic aortic arch
- Need ASD for survival

****** PDA dependent ******

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Hypoplastic Left Heart Syndrome (HLHS)

Clinical manifestations

- Timing and severity of symptoms dependent on:
  - Presence of PDA
  - Adequacy of left-right atrial flow
  - Relative PVR and SVR
- Immediately after birth:
  - Minimal symptoms: mild cyanosis and mild tachypnea
- After PDA closes:
  - CHF and decreased peripheral pulses, metabolic acidosis with renal & GI hypo-perfusion progressing to shock
- Murmur **may** be present
- CXR:
  - Increased pulmonary vascular markings
  - Increased heart size

Management

- PGE1 to maintain PDA
- Minimal oxygen supplementation (to maintain elevated PVR)
- Hypoventilation (hypercarbia associated with elevated PVR)
- Inotropic support (Dopamine/Dobutamine)
- Treat CHF
- Balloon atrial septostomy may be needed if absence of ASD
- Staged surgical repair
- May require heart transplant

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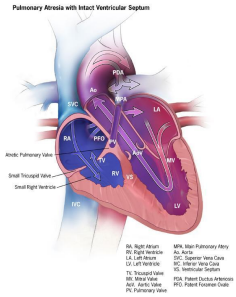
Hypoplastic Left Heart Syndrome (HLHS)

Staged repair

1. Norwood procedure
   - Usually in the first 2 weeks of life
   - Most complex surgery of all 3 surgeries
   - Create a "new" aorta and connect it to the right ventricle
   - Place a BT (Blalock-Taussig) shunt
     - One branch of the subclavian artery or carotid artery is separated and connected to the pulmonary artery in order to increase pulmonary blood flow
     - The right ventricle can pump blood to both the lungs and the rest of the body
   - After surgery, baby may appear bluish because there is still mixing of oxygen-rich and oxygen-poor blood
2. Bi-directional Glenn shunt procedure
   - Performed around 4-6 months of age
   - Connect the PA and the SVC returning oxygen-poor blood from the **upper** part of the body to the heart
     - *Reduces the work of the right ventricle by allowing blood returning from the body to flow directly to the lungs*
3. Fontan Procedure
   - Performed between 18 months to 3 years of age
   - Connect the PA and the IVC returning oxygen-poor blood from the lower part of the body to the heart
   - Once this procedure is complete, oxygen-rich and oxygen-poor blood no longer mix
     - *NO more blue skin!*

Pulmonary Stenosis/Atresia



- Absence or narrowing of the pulmonary valve
- Requires an ASD or PFO with PDA to survive

***** PDA dependent*****

Pulmonary Stenosis/Atresia

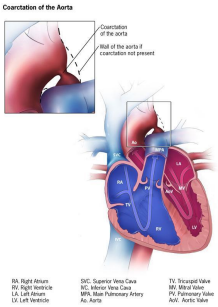
Clinical manifestations

- Severe cyanosis at birth
- Single S2
- typically no murmur, may have systolic murmur from TR
- CXR:
  - Normal or increased heart size
  - Decreased pulmonary vascular markings

Management

- PGE1 immediately
- surgical repair:
  - BT shunt
  - RVOT reconstruction +/- BT shunt

Coarctation of the aorta



- Defect in which the aorta is narrower than usual
- Considered a critical heart defect
- Ductal dependent
- Interrupted aortic arch is the most severe form (DiGeorge)
- Males > females (2:1)
- Common in Turner's syndrome

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Coarctation of the aorta

Clinical manifestations

- Decreased femoral pulses
- Most with cyanosis
- Lower blood pressure in the lower extremities than in the upper extremities
  - *Systolic differential BP gradient >10mmHg between arms and legs*
- Systolic ejection murmur, loudest at left interscapular area in back
- CXR
  - Increasing heart size with greater severity of coarctation

Management

- PGE1 if severe coarctation
- Surgery with end-to-end anastomosis

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Left-to-Right Shunts

- ❑ Ventricular septal defect (VSD)
- ❑ Patent ductus arteriosus (PDA)
- ❑ AV canal (endocardial cushion defect)

Left-to-right shunt → red oxygenated blood flows from the left side of the heart to the right side (lung)

- *Results in fluid overload in the lungs and CHF*

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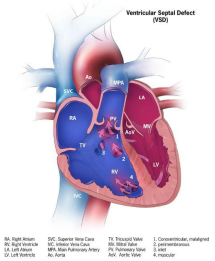
Ventricular Septal Defect (VSD)



RA: Right Atrium
RV: Right Ventricle
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SVC: Superior Vena Cava
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MPA: Main Pulmonary Artery
Ao: Aorta
TV: Tricuspid Valve
MV: Mitral Valve
PV: Pulmonary Valve
AoV: Aortic Valve
1. Conoventricular, malaligned
2. perimembranous
3. inlet
4. muscular

- Most common CHD
- Most common cause of CHF after 2 weeks of life
- **Symptoms** dependent on degree of PVR and size of VSD:
  - Small: typically asymptomatic
  - Mod-Lg: as PVR decreases, may see poor feeding, increased respiratory infections → poor growth, developmental delay
  - Harsh holosystolic ejection murmur heard over left lower sternal border
- Management:
  - Spontaneous closure
  - Treat CHF
  - Surgery with direct closure when significant L-R shunting with severe CHF, poor growth, or increased PA pressures

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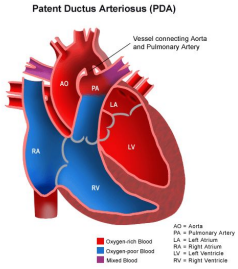
Patent Ductus Arteriosus (PDA)



- Closure occurs as PaO2 increases with increased contracting mediators and decreased dilating mediators
- Functional closure occurs in most full-term infants by 48 hours of age
  - Preterm infants may not have functional closure until days to weeks
- Left-to-Right shunting increases as PVR decreases

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Patent Ductus Arteriosus (PDA)

Clinical manifestations

- Machinery-type murmur loudest at left upper sternal border
- Small: usually asymptomatic
- Mod-Lg:
  - Bounding peripheral pulses
    - *Palmar pulses*
  - Widened pulse pressure
  - Hyperdynamic precordium
  - Symptoms of CHF
- In premature infants:
  - Decreased lung compliance
  - Difficulty weaning from ventilator
- CXR:
  - Increased heart size, increased pulmonary blood flow

Management

- Fluid restriction
- Maintain adequate hematocrit
- Indomethacin or Ibuprofen
- Surgical ligation

***** Early management improves outcomes*****

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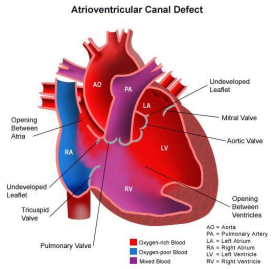
Atrioventricular Canal Defect (AV Canal)



- Majority of patients with AV canal have trisomy 21
- Associated with PDA and TOF
- Developmental abnormality of endocardial cushion (endocardial cushion defect)
- Left-to-right shunt dependent on PVR →
  - Lower PVR → greater L-R shunt

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Atrioventricular Canal Defect (AV Canal)

Clinical manifestations

- Symptoms based on combination of ASD and VSD as well as degree of AV valve insufficiency
- CHF (volume overload) and cyanosis
- Systolic murmur, loudest at lower left sternal border
- CXR:
  - Increased heart size

Management

- Treat CHF
- Limit the amount of oxygen delivered →
  - O2 is a pulmonary vasodilator → decreased PVR → increased L-R shunt
- Surgical ASD or VSD closure
- Surgical construction of 2 separate AV valves

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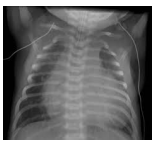
Congestive Heart Failure (CHF)

Clinical Manifestations

- S & S resulting from inability of the heart to meet the demands of the body
- Hydrops if severe fetal CHF
- Symptoms include:
  - Tachypnea
  - Tachycardia
  - Hepatosplenomegaly
  - Weak peripheral pulses
- CXR
  - Increased heart size
  - Increased pulmonary vascular markings



Management

- Medical management:
  - Diuretics
  - Digoxin
  - ACE inhibitors- reduces afterload

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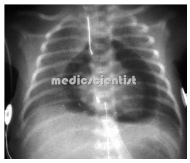
Cardiac Tamponade

- Pressure on the heart that occurs when blood or fluid builds up in the space between the heart and the outer covering sac of the heart
  - Prevents the ventricles from expanding fully and reduces cardiac output
- S & S:
  - Tachypnea
  - Tachycardia
  - Muffled heart sounds
  - Weak or absent peripheral pulses
  - Hypotension
- Management:
  - Evacuation of blood/fluid by pericardiocentesis



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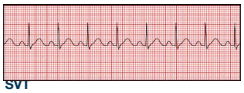
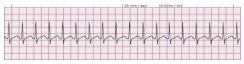
Supraventricular Tachycardia (SVT)

- Most common pediatric symptomatic arrhythmia
  - Up to 1/250 infants
- S & S:
  - HR > 220bpm with little variation in rate
  - Absent p waves
  - QRS wave usually normal
- If left untreated, can lead to CHF
- **Sinus tachycardia**



- **SVT**



Management

- If stable:
  - Vagal maneuvers (ice to face)
  - Adenosine (0.1mg/kg) rapid IV
- If unstable:
  - Synchronized cardioversion
    - *0.5-2 joules/kg*
- Long-term therapy:
  - Digoxin
  - Propranolol

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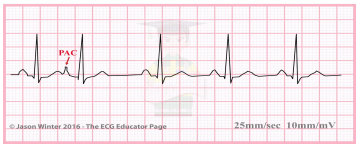
Premature Atrial Contraction (PAC)

- Common in newborns
- Typically benign
- Early p wave (looks different than normal p wave), usually normal QRS
- Premature beat originates in the atrium and leads to contraction before sinus node
- May be associated with hyperthyroidism, CHD, cardiomyopathy, central line irritation of right artium

Premature Atrial Contraction (PAC)



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Shock

Blood flow to tissues is inadequate to meet tissue metabolic requirements leading to tissue hypoxia, metabolic acidosis, irreversible cellular changes, and cellular death

Hypovolemic	Distributive	Cardiogenic
▪ **Most common in neonates** ▪ Decreased blood volume	▪ Inadequate intravascular volume secondary to vasodilation	▪ Cardiac failure
▪ Decreased ventricular filling and decreased stroke volume ▪ Decreased cardiac output	▪ Normal circulating blood volume but insufficient for adequate cardiac filling	▪ Impaired filling ▪ Impaired ventricular emptying ▪ Impaired contractility
S&S: ▪ Decreased U.O. ▪ Decreased BP ▪ Increased HR (premature infants may have decreased HR)	S&S: ▪ Decreased U.O. ▪ Increased HR ▪ Decreased BP ▪ Bounding pulses	S&S: ▪ Decreased U.O. ▪ Increased HR ▪ Decreased BP ▪ CHF/PE ▪ Hepatomegaly ▪ Cardiomegaly
Causes: ▪ Severe hemorrhage ▪ Severe fluid loss ▪ Sepsis (capillary leak into 3rd space)	Causes: ▪ Sepsis ▪ Anaphylaxis ▪ Vasodilators ▪ Toxins	Causes: ▪ Metabolic (hypocalcemia, hypoglycemia) ▪ CHD ▪ Cardiac tamponade ▪ Severe perinatal depression ▪ Arrhythmias, myocarditis, cardiomyopathy, MI ▪ Sepsis (decreased contractility)

Hypertension

- Hypertension is defined as > 2 S.D. above normal values for age and weight
- Many different causes of HTN in neonates:
  - Vascular: renal artery thrombosis (r/t umbilical lines), renal vein thrombosis, coarctation of aorta
  - Renal
  - Endocrine: congenital adrenal hypoplasia, hyperthyroidism
  - Neurologic: IVH, hydrocephalus, meningitis, drug withdrawal, seizures
  - Pulmonary: BPD
  - Drugs: corticosteroids
  - Other: fluid overload, pain

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Hypertension

Evaluation

- Determine how BP was taken
  - Invasive vs cuff
  - Which extremity
    - LE BP > UE BP
  - Proper cuff size
  - Activity state of infant
- History of umbilical artery line
  - Increased risk of renovascular HTN and thrombosis
- Pain or agitation
- History of BPD
- Medications
- Labs:
  - Cr, BUN
  - Electrolytes
  - UA/culture

Imaging

- Renal U/S
- Head U/S (r/o IVH)
- Echocardiogram

Management

- Treat underlying cause
- Antihypertensive medications
  - ACE inhibitors (captopril)
  - Beta blockers (labetalol, propranolol)
  - Ca channel blockers (amlodipine)
  - Vasodilators (hydralazine)

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PPHN
Persistent Pulmonary Hypertension

- Recall:
  - Pulmonary vascular resistance (PVR) is high in utero which aids in shunting oxygenated umbilical venous blood to the aorta (Right-to-Left)
  - After delivery, expansion of lungs leads to **decreased** pulmonary vascular resistance (PVR) and increased pulmonary blood flow
- PPHN results when there is failure of this natural drop in PVR

PVR > SVR → **Right-to-left shunt**

- Shunting of deoxygenated blood away from the lungs into systemic circulation, leading to cyanosis
- Shunting occurs across PDA or PFO
  - *As a result, post-ductal saturations will be lower than pre-ductal (by as much as 10-15%)*
- May present immediately after delivery of within the first few days

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PPHN

PPHN Etiology

- Any process that causes hypoxia can lead to increased PVR
  - *Meconium aspiration syndrome*
  - *Sepsis*
  - *Congenital pneumonia*
  - *Idiopathic/abnormal vascular development*
  - *Hypoxic ischemic encephalopathy*
  - *Intrinsic lung disease*
  - *Congenital diaphragmatic hernia*
    - *Underdevelopment of lung tissue and pulmonary vasculature*
  - *Pulmonary hypoplasia*
  - *Prematurity*

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PPHN

Clinical manifestations

- Hypoxia
- Pre and post ductal saturation splitting (10-15%)
  - **Pre > Post**
- Tachypnea
- Cyanosis
- Respiratory distress
- Acidosis
- Hypotension
- Labile oxygen saturations

Diagnostic studies

- CXR
  - Dark lung fields d/t decreased pulmonary blood flow
  - CDH
  - Pneumonia
  - MAS
- Echocardiogram
  - Right ventricular hypertrophy
  - Atrial septal flattening
  - Right-to-left shunting across PDA or PFO
  - Elevated pulmonary artery pressure
  - Tricuspid regurgitation
  - Supra-systemic pulmonary pressures

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PPHN

Management

- Oxygen
  - *Goal is to keep saturations > 95% to improve pulmonary vascular resistance*
  - *Oxygen is a vasodilator*
- Surfactant
  - *RDS, MAS*
- Avoid respiratory or metabolic acidosis
  - *Goal pH > 7.25*
  - *Acidosis contributes to PPHN*
- Inhaled nitric oxide
  - *Pulmonary vasodilator*
- Low stimulation environment
- Sedation
- Blood pressure support (preload/output)
  - *Fluid*
  - *Inotropes – Dopamine/Dobutamine*
    - ***Goal: Systemic BP > pulmonary BP***
- ECMO
  - *OI > 35-40*

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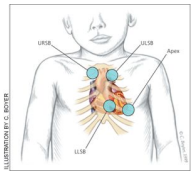
Cardiovascular Assessment
Physical Exam

- Auscultate
  - Heart sounds
    - ***S1****: closure of mitral and tricuspid valves*
    - ***S2****: closure of aortic and pulmonary valve*
  - Murmurs
    - *Note location, timing, grade and intensity (grade I-VI)*



- Inspect
  - Precordial activity
- Pulse pressure
  - Difference between systolic and diastolic BP
  - Wide pulse pressure
    - PDA, (left-to-right)
- Blood pressure
  - 4 extremity blood pressure- should not be > 10mmHg difference between UE and LE
  - Invasive blood pressure monitoring (UAC/PAL)
- Peripheral pulses
- Capillary refill
  - Normal < 3 sec
- Assess respiratory status
- Pulse oximetry

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Cardiovascular Assessment

Diagnostic tools

- Vital signs
- Labs: electrolytes, Cr/BUN, UA/culture, BNP
- CXR: assess heart size and pulmonary vascular markings
- EKG
- Echocardiogram

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Thank you

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