The 2AM call from the well-baby nursery…
Your night just got a whole lot longer

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Disclosures

• I have nothing to disclose
Case 1

- You are called to a well baby nursery for a term infant who had meconium stained fluid. The baby required delivery room interventions of drying, stimulation, CPAP with PPV for stabilization. APGARs are 5/6/9. This baby did well and was transitioned to room air in the delivery room and sent to the well baby nursery. You head back to the call room to watch the Sporting KC match.

- About 20 minutes later the nurse calls you and tells you: “The baby is blue, like a blueberry but is happy and doesn’t appear in any distress. She is not having tachypnea or anything. She’s just blue.” So you order a CXR
Case 1

• You are called to a well baby nursery for a term infant who had meconium stained fluid. The baby required delivery room interventions of drying, stimulation, CPAP with PPV for stabilization. APGARs are 5/6/9. This baby did well and was transitioned to room air in the delivery room and sent to the well baby nursery. You head back to the call room to watch the Sporting KC match.

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Case 1

• Which of the following congenital heart lesions is most likely in this patient?
  • Tetralogy of Fallot
  • Hypoplastic Left Heart Syndrome
  • Pulmonary Atresia with Intact Ventricular Septum
  • Total Anomalous Pulmonary Venous Return
  • Transposition of the Great Arteries
Case 2

• You are called to the emergency department for a 5 day old baby that EMS has brought in from a PCPs office. According to the ED physician the baby was seeing their Pediatrician for their well baby follow up and was noted to be “not acting right”. The baby has had problems feeding and is 15% below birth weight. You note on exam that the baby is mottled, listless, and has poor perfusion to the lower extremities. A quick review of the baby’s birth history shows nothing abnormal, she passed her hearing screen and CCHD screens without issue.

Case 2

• Based on the diagnosis you suspect, which of the following interventions should be your FIRST step in post-natal management.
  • Initiation of prostaglandin infusion
  • Intubation and mechanical ventilation
  • Rashkind balloon septostomy
  • Extracorporeal membrane oxygenation
  • Emergent cardiac surgery
Goals and Objectives

- Know the signs and symptoms that present in an infant with CHD.
- Understand the initial management of the infant with CHD.

Outline

- Epidemiology
- Presentation of infants with CHD
- Specific CHD Lesions
- Initial management
- Summary
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Epidemiology

• Critical Congenital Heart Disease:
  • Hypoplastic left heart syndrome
  • Transposition of the great arteries
  • Tricuspid atresia, truncus arteriosus
  • Tetralogy of Fallot
  • Total anomalous pulmonary venous return
  • Pulmonary atresia
  • Interrupted aortic arch/coarctation of the aorta
  • Double outlet right ventricle
  • Epstein’s anomaly

Epidemiology

• 1% of all infants born in the US have CHD
  • Approximately 4 million infants delivered each year
  • That equates to 40,000 infants with CHD
  • Estimated that up to 70 deaths annually in the US are due to missed diagnosis

• 70-80% are not diagnosed within the first 2 days of life

Epidemiology – One year survival

• Survival*
  • 1979 – 1993: One-year survival for infants with CCHDs 67.4%.
  • 1994 – 2005: One-year survival for infants with CCHDs 82.5%
  • Non-critical CHD: One-year survival for infants 97.1%

• Higher risk of 1-year mortality for infants with
  • Earlier birth era
  • Earlier diagnosis (i.e. more severe disease)
  • Low birth weight
  • Mothers were <30 years old.

Epidemiology – Survival to adulthood

- Survival to adulthood (18 years of age)
  - Noncritical CHDs 95.4%
  - Critical CHDs 68.8%

*Oster ME, Lee KA, Honein MA, Riehle-Colarusso T, Shi M, Correa A. Temporal Trends in Survival Among Infants With Critical Congenital Heart Defects Pediatrics May 2013, 131 (5) e1502-e1508; DOI: 10.1542/peds.2012-3435

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Presentation – Delivery Room Tips

• Some reassuring information
  • A duct does not close immediately!!!!
    • Term infants → 2-3 days, sometimes 5 days.
    • Preterm infants → weeks
  • Very, very few infants with CHD are delivered in fulminate cardiovascular collapse.
    • So you have time to think, stabilize, and transfer
• Never be afraid to start Prostins.
  • If you start them and it’s not CHD, we just turn them off.

Presentation – Signs/Symptoms

• Tachypnea
  • Mild to moderate respiratory distress
  • Due to hypoxemia (PaO2) from oxygen-receptors.
  • Severe respiratory distress is triggered by hypercarbia (paCO2) from carbon dioxide-receptors.
    • Nasal flaring, grunting, retractions
• Cyanosis
  • May take hours or days to develop
  • Frequently with feeding or crying
Presentation – Signs/Symptoms

• Decreased systemic perfusion
  • Stable at birth, develops during first hours to days of life.
  • Signs include: poor feeding, diaphoresis, poor pulses, temperature instability, low capillary refill

• Tachypnea + something else…
  • Cyanosis = Consider CHD
  • Poor Perfusion = Consider CHD
  • n/a = usually primary respiratory cause

Presentation – Physical Exam

• Central cyanosis
  • Tongue, gums, buccal mucosa
  • Cyanosis is difficult to see until SpO2 < 85%*

• Pulses/Blood Pressure
  • Lower extremity pulses are easier to feel than inguinal.
  • Lower extremity BP is usually slightly higher than upper extremity
    • Right arm (prechordal) and right leg (post ductal)

• Abdominal Exam
  • Palpation of the liver
    • Hepatomegaly can be a sign of right atrial hypertension due to excessive pulmonary blood flow

*Depends of Hb level
Presentation – Physical Exam

• Murmur
  • 50% of infants have a murmur
    • Not a concern if otherwise normal exam
  • Split S2
    • Can be normal in first few days of life, as pulmonary blood flow increases
    • Suggestive of excessive pulmonary blood flow → TAPVR
• Clicks
• Bruits

• Chest Palpation
  • ↑ Parasternal thrill → d-TGA, Tricuspid atresia, or HRHS*
  • Suprasternal notch thrill → Aortic Stenosis

Presentation – Diagnostic Help

Superior, Right (Up I, Down aVF)
• Tricuspid Atresia
• Atrioventricular Canal

Superior, Left (Up I, Down AVF)
• Pulmonary Atresia w/ IVS

Inferior, Right (Down I, Up aVF)
• Normal < 1 month
• Tetrology of Fallot

Inferior, Left (Up I, Up aVF)
• Normal > 1 month
• Pulmonary Atresia w/ IVS
Outline

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Specific CHD Lesions

• Tips to understanding lesions
  • It's all plumbing
  • "Blood is stupid and lazy"
    • It always follows the path of least resistance and lowest pressure
## Summary of CHD Lesions

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Bold = Traditionally Ductal Dependence, \# = Needs catheter or operative intervention as neonate.

TOF + PS = Tetralogy of Fallot with Pulmonary Stenosis, TA = Tricuspid Atresia, HRHS = Hypoplastic Right Heart Syndrome, PA = Pulmonary Atresia, IVS = Intact ventricular septum, VS = Ventricular Septal Defect, LSMAPCAs = Long Segment Multiple Aortopulmonary Collaterals, PS = Pulmonary Stenosis, TAPVR = Total Anomalous Pulmonary Venous Return, PVS = Pulmonary Vein Stenosis, IAA = Interrupted Aortic Arch, AS = Aortic Stenosis, HLHS = Hypoplastic Left Heart Syndrome, AVC – Atrioventricular Canal, DILV = Double Inlet Left Ventricle, PDA = Patent Ductus Arteriosus, ASD = Atrial Septal Defect, D-TGA = Dextro Transposition of Great Arteries, L-TGA = Levo Transposition of Great Arteries

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Congenital Heart Disease – Right Sided Lesions

- Symptoms are dependent on degree of obstruction
  - Obstruction to pulmonary blood flow causes a R → L shunt across ASD or VSD.
  - Severe obstruction L → R shunt via PDA.
    - Pulmonary blood flow dependent on PDA
- Pre and post ductal SpO2 will be equal.
- Saturations will be low (70-85%)
- Very dark lung fields on CXR
- What makes a lesion ductal dependent?
  - Any lesion where there isn’t enough blood flow to the lungs without the duct.

Congenital Heart Disease – Right Sided Lesions

- Tetralogy of Fallot (TOF)
  - VSD, Overriding Aorta, PS or PA, Right Ventricular Hypertrophy
  - Wide Spectrum
    - “Pink Tet” (Single VSD) has good saturations
      - Not ductal dependent (Tet Spells)
    - TOF + PA (Severely hypoxic)
      - Ductal dependent
  - Cyanosis is related to degree of RVOT obstruction
    - More obstruction of blood flow to lungs, the more cyanotic.
  - Exam findings:
    - Palpable ventricular impulse
    - Single S2

https://radiopaedia.org/articles/tetralogy-of-fallot

Potentially ductal dependent
Congenital Heart Disease – Right Sided Lesions

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Congenital Heart Disease – Right Sided Lesions

• Tricuspid Atresia
  • Plate like material separating right atrium from right ventricle.
    • Due to lack of flow can have hypoplastic right ventricle and hypoplastic pulmonary valve and artery.
  • Complete mixing in the left atrium
    • Must have ASD to survive.
  • Pulmonary blood flow from either PDA or VSD
    • TA with a small VSD and PS ARE ductal dependent
    • TA with large VSD and minimal PS are NOT.
• Exam findings:
  • Hepatomegaly
  • Single S2
  • Right upper axis deviation on EKG.
Congenital Heart Disease – Right Sided Lesions

• Pulmonary Stenosis → Pulmonary Atresia
  - Cyanosis within a few hours of birth
  - Can have a stenotic valve, normal pulmonary annulus with plate of tissue obstructing, or can have complete absence of pulmonary artery.
  - Can have VSD, IVS, or LSMAPCAs
    - VSD: cyanosis
    - IVS: cardiovascular collapse with PDA closure
  - Right ventricle feeds right coronaries
    - Fistulization from high RV pressures
  - Need catheter or surgical intervention
  - Exam findings:
    - Single S2
    - Blowing systolic murmur at LLSB (TR)

Congenital Heart Disease – Right Sided Lesions

• Ebstein’s Anomaly of the Tricuspid Valve
  - Downward displacement of tricuspid valve
  - Atrialization of the right ventricle
  - Typically a right to left shunt across PFO
    - Based on the degree of TR
    - Severe TR causes the lesion to be ductal dependent
  - EKG 20% can have WPW
  - Exam findings
    - Multiple systolic clicks
    - Holosystolic murmur

Ductal Dependent Lesion
Congenital Heart Disease – Right Sided Lesions

- Oxygen
- EKG
  - Can help identify potential lesions (left axis deviation)
- When in doubt, start Prostins
- Ductal closure presents with:
  - Severe cyanosis
  - Acidosis

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Congenital Heart Disease – Left Sided Lesions

- Symptoms vary with degree of obstruction and ductal status
  - Open ductus
    - Mild cyanosis, pulmonary over circulation
  - Closing ductus
    - Hypotension, poor perfusion, cyanosis, acidosis, tachypnea

- Pre and post ductal SpO2 will be different
  - Pre-ductal > 90%, Post-ductal 80-85%

- What makes a lesion ductal dependent?
  - Any lesion where there isn’t enough blood flow to systemic circulation without the duct.

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Congenital Heart Disease – Left Sided Lesions

- Aortic Stenosis (mild → critical)
  - Mild to moderate is well tolerated
    - Routine follow up is all that is needed.
  - Severe stenosis presents with shock
    - This is ductal dependent
      - Similar to HLHS, but less common
      - Acidosis
      - Multiorgan failure
  - Location of stenosis is important as well
    - Supravalvular = Williams Syndrome
  - Can occur with coarctation of aorta
  - Exam findings:
    - Tachypneic
    - S3 gallop
    - Upper and lower extremity saturation difference.

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Image Credit: [http://cardinalglennon.congenital.org/?id=aorticstenosis](http://cardinalglennon.congenital.org/?id=aorticstenosis)
Image Credit: [http://www.childrenshospital.org/conditions-and-treatments/conditions/a/aortic-valve-stenosis](http://www.childrenshospital.org/conditions-and-treatments/conditions/a/aortic-valve-stenosis)
Congenital Heart Disease – Left Sided Lesions

• Hypoplastic Left Heart Syndrome
  • Aortic valve atresia, mitral valve atresia, hypoplastic proximal aortic arch, and coarctation.
  • The most common CHD that presents with inadequate systemic perfusion after birth.
  • Entire systemic cardiac output is dependent on ductal flow
• Presentation:
  • Symptoms develop a few hours after birth
    • Poor feeding
    • Tachycardia
    • Cyanosis (SPO2 80 - 90%)
    • Severe respiratory distress
    • Poor systemic perfusion
• Exam findings:
  • Hyperactive precordium
  • Hepatomegaly
  • S3 can be heard

Ductal Dependent Lesion

Congenital Heart Disease – Left Sided Lesions

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Looks a lot like Sepsis!!
Congenital Heart Disease – Left Sided Lesions

• Hypoplastic Left Heart Syndrome
  • Start Prostins immediately
  • Allow permissive hypercapnea
  • Do not give oxygen or iNO
  • Can give blood (Hct > 45%)

Ductal Dependent Lesion

Congenital Heart Disease – Left Sided Lesions

• Interrupted Aortic Arch and Coarctation of Aorta
  • Ends of a spectrum from obstruction to complete discontinuity.

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<td>Part of HLHS</td>
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<td>Type B is most common</td>
<td>Most common site is at the PDA insertion and origin of left subclavian</td>
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• Presentation
  • Not usually cyanotic or tachypneic at birth
    • Symptoms develop with anatomic closure of the PDA
    • Cyanosis, severe respiratory failure, acidosis, poor perfusion

• Exam findings:
  • Delay between upper and lower extremity pulses
    • Dorsalis pedis and posterior tibial are easier to feel, femoral are deep pulses and more challenging.
  • Blood pressure differences (right upper extremity and a lower extremity).
Congenital Heart Disease – Left Sided Lesions

- Interrupted Aortic Arch and Coarctation of Aorta
  - Treatment: Start Prostins ASAP

Ductal Dependent Lesion

- Judicious use of oxygen
  - It can make things worse
- Do NOT start iNO
- Echocardiogram, EKG is not really helpful
- Prostaglandins
- Lot of duct leads to
  - Hypotension, Acidosis, Shock
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Ductal Dependent Lesion

Congenital Heart Disease – Mixing Lesions

- Major connections between right and left heart circulation
  - Can have bidirectional shunting
  - Both circulations mix resulting in lower saturations.
  - Blood follows the path of least resistance
    - PVR starts to drop at day three of life, nadir at 6 weeks post GA
      - This can lead to pulmonary overcirculation, failure to thrive, and congestive heart failure.
- Management centers around balancing two circulations (Qp:Qs)
  - SpO2 > 90 too much blood returning to the lungs
  - SpO2 75-85% is typically a good place to be with these infants
- No mixing lesion is ductal dependent
Congenital Heart Disease – Mixing Lesions

• Total Anomalous Pulmonary Venous Return
  • Supracardiac (55%)
    • Can present with obstruction when the left bronchus and left pulmonary artery.
    • Presents with moderate distress.
  • Cardiac (27%)
    • Connects to coronary sinus or right atrium
  • Infracardiac (13%)
    • Generally presents with severe extremis.
    • Obstruction of the descending vertical vein at the diaphragm or liver.

The Snowman sign on CXR
### Congenital Heart Disease – Mixing Lesions

- **Total Anomalous Pulmonary Venous Return - Obstruction**
  - In utero the anomalous vein connects to a widely patent ductus venosus
  - At birth, this starts to narrow and close causing obstruction
  - Presents in extremis.
    - Severe respiratory distress, tachypnea, intercostal retractions, nasal flaring, grunting.
    - Saturations in mid-80s
    - Decreased pulses in all extremities
    - Narrow pulse pressure
    - Split second heart sound
    - In a cyanotic infant a split S2 nearly always indicates TAPVR
  - Prostins may dilate ductus venosus
  - Immediate surgical intervention – Need Surgeon!!!
    - Catheter procedure
    - ECMO

**Image Credit: Neonatal Cardiology**

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<td>******</td>
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</tbody>
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- **TOF**
- **TOF + PA**
  - Obstructed TAPVR
  - IAA*
  - AVC
  - D-TGA
- **TA → HRHS#**
  - PVS
  - Coarctation*
  - Truncus
  - L-TGA
- **PS#**
  - AS#
  - TAPVR
- **PA + IVS#**
  - PVS
  - HLHS*/ Shones
  - DILV
- **PA + VSD**
- **PA + LSMAPCAs**
  - PDA
  - VSD
- **Ebtstein’s Anomaly**
  - ASD

**Legend:**
- TOF + PS = Tetralogy of Fallot with Pulmonary Stenosis, TA = Tricuspid Atresia, HRHS = Hypoplastic Right Heart Syndrome, PA = Pulmonary Atresia, IVS = intact ventricular septum, VS = Ventricular Septal Defect, LSMAPCAs = Long Segment Multiple Aortopulmonary Collaterals, PS = Pulmonary Stenosis, TAPVR = Total Anomalous Pulmonary Venous Return, PVS = Pulmonary Vein Stenosis, IAA = Interrupted Aortic Arch, AS = Aortic Stenosis, HLHS = Hypoplastic Left Heart Syndrome, AVC = Atrioventricular Canal, DILV = Double Inlet Left Ventricle, PDA = Patent Ductus Arteriosus, ASD = Atrial Septal Defect, D-TGA = Dextro Transposition of Great Arteries, L-TGA = Levo Transposition of Great Arteries
- Bold = Traditionally Ductal Dependence, # = Needs catheter or operative intervention as neonate.
Congenital Heart Disease – Parallel Circulation

• Adequate mixing at the atrial level, Prostins may not be needed.
  • Prostins will never hurt baby.
  • When in doubt start before an ECHO
• Oxygen may help
  • Target Preductal SpO2 > 80%

• d - Transposition of the Great Arteries
  • Most common CHD diagnosed in 1st week of life
  • Associated with VSD (25%) and coarctation
  • Presentation
    • Profoundly cyanotic at birth
    • Little to know respiratory distress
    • Post ductal SpO2 > Preductal
    • R → L ductal flow gets oxygenated blood to systemic circulation.
    • Equally low pre and post ductal saturations
      • Adequate mixing at the level of the atria
  • Exam findings
    • Single S2 heart sound
    • Holosystolic murmur if VSD is present
    • Systolic ejection click if PS is present
  • CXR
    • Egg of on string


Ductal Dependent Lesion

Congenital Heart Disease – Parallel Circulation

• d - Transposition of the Great Arteries
  • Most common CHD diagnosed in 1st week of life
  • Associated with VSD (25%) and coarctation
  • Presentation
    • Profoundly cyanotic at birth
    • Little to know respiratory distress
    • Post ductal SpO2 > Preductal
    • R → L ductal flow gets oxygenated blood to systemic circulation.
    • Equally low pre and post ductal saturations
      • Adequate mixing at the level of the atria
  • Exam findings
    • Single S2 heart sound
    • Holosystolic murmur if VSD is present
    • Systolic ejection click if PS is present
  • CXR
    • Egg of on string

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Congenital Heart Disease – Parallel Circulation

- d - Transposition of the Great Arteries
  - Most common CHD diagnosed in 1st week of life
  - Associated with VSD (25%) and coarctation
- Presentation
  - Profoundly cyanotic at birth
  - Little to know respiratory distress
  - Post ductal SpO2 > Preductal = ductal dependent
    - R → L ductal flow gets oxygenated blood to systemic circulation.
  - Equally low pre and post ductal saturations
    - Adequate mixing at the level of the atria
- Exam findings
  - Single S2 heart sound
  - Holosystolic murmur if VSD is present
  - Systolic ejection click if PS is present
- CXR
  - Egg of on string

Outline

- Epidemiology
- Presentation of infants with CHD
- Specific CHD Lesions
- Initial management
- Summary
Initial Management

- Oxygen
  - Avoid in left sided obstructive lesions
  - Potent vasodilator, can increase pulmonary overcirculation
    - ↓ UOP, ↓ systemic perfusion (worsening pulses, poor capillary refill, acidosis).
  - Can precipitate ductal closure
  - "If they get better with oxygen keep using it, if they get worse then stop."
  - Saturations in 70s-80s are acceptable for cardiac infants.

- Echocardiogram and/or EKG

- CXR
  - Boot shaped heart → TOF
  - Egg on string → TGA
  - Snowman → TAPVR

Initial Management

- Prostaglandin E1
  - Infants with ductal dependent lesions present with profound cyanosis with little respiratory distress.
    - Present between day of life 3 – 14 with severe cardiogenic shock
  - Indications for PGE1
    - ↓ pulmonary blood flow → right sided obstructive lesions
    - ↓ systemic blood flow → left sided obstructive lesions
  - Dose (0.02mcg/kg/min)
    - Continuous infusion
    - Don’t bolus, they will go apneic
  - What to watch for after starting prostins
    - Hypotension
    - Apnea/hypoventilation
    - Fever
    - Cutaneous flushing
**Where Prostins don’t help.**

- D-TGA with PFO (no ASD or VSD)
  - Insufficient mixing at atrial level, emergent Rashkind Septostomy
- Obstructed TAVPR
  - Immediate surgical correction is the only treatment
- HLHS with intact atrial septum
  - Insufficient mixing at atrial level, emergent Rashkind Septostomy

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**Case 1**

- You are called to a well baby nursery for a term infant who had meconium stained fluid. The baby required delivery room interventions of drying, stimulation, CPAP with PPV for stabilization. APGARs are 5/6/9. This baby did well and was transitioned to room air in the delivery room and sent to the well baby nursery. You head back to the call room to watch the Sporting KC match.
Case 1

• You are called to a well baby nursery for a term infant who had meconium stained fluid. The baby required delivery room interventions of drying, stimulation, CPAP with PPV for stabilization. APGARs are 5/6/9. This baby did well and was transitioned to room air in the delivery room and sent to the well baby nursery. You head back to the call room to watch the Sporting KC match.

• About 20 minutes later the nurse calls you and tells you: “The baby is blue, like a blueberry but is happy and doesn’t appear in any distress. She is not having tachypnea or anything. She’s just blue.” So you order a CXR.
Case 1

- Which of the following congenital heart lesions is most likely in this patient?
  - Tetralogy of Fallot
  - Hypoplastic Left Heart Syndrome
  - Pulmonary Atresia with Intact Ventricular Septum
  - Total Anomalous Pulmonary Venous Return
  - Transposition of the Great Arteries

Case 2

- You are called to the emergency department for a 5 day old baby that EMS has brought in from a PCP's office. According to the ED physician the baby was seeing their Pediatrician for their well baby follow up and was noted to be "not acting right". The baby has had problems feeding and is 15% below birth weight. You note on exam that the baby is mottled, listless, and has poor perfusion to the lower extremities. A quick review of the baby's birth history shows nothing abnormal, she passed her hearing screen and CCHD screens without issue.
Case 2

• Based on the diagnosis you suspect, which of the following interventions should be your FIRST step in post-natal management.
  • Initiation of prostaglandin infusion
  • Intubation and mechanical ventilation
  • Rashkind balloon septostomy
  • Extracorporeal membrane oxygenation
  • Emergent cardiac surgery

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