Recognizing Congenital Heart Disease in the Newborn

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David M Bush, MD, PhD has no relevant financial relationships with commercial interests to disclose.

Opening Thoughts...

“Too much light often blinds gentlemen of this sort. They cannot see the forest for the trees.”
Musarion [1768], Canto II

Christoph Martin Wieland (1733-1813)

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Where We Are Going....

- Tools of the Trade
- Identifying Cardiac Physiologies
  - Pulmonary Blood Flow-Dependent Cyanosis
  - Pulmonary Blood Flow-Independent Cyanosis
  - Pulmonary Vascular Resistance-Dependent Shunting
  - Ventricular Compliance-Dependent Shunting
  - Failing Cardiac Output States
  - Pulmonary Venous Obstruction
- Putting It All Together

Tools of the Trade

- The most important tool
- The adjunctive tools
  - Vital Signs
  - Pulse Oximetry
  - Chest Radiograph
  - Screening Echocardiography

Pulmonary Blood Flow-Independent Cyanosis
*a.k.a. “Transposition Physiology”*

*“The problem is inadequate atrial mixing...”* William Rashkind

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Pulmonary Blood Flow-Independent Cyanosis
*a.k.a. – “Transposition Physiology”*

**Presentation:**
- Asymptomatic to tachypneic
- Timing dependent upon amount of mixing (Birth – Two Weeks)
- LOOK for signs of increased work of breathing

**Chart Radiograph:**
- Classic finding – “normal” pulmonary vascular markings
- Generally normal heart size
- Associated anomalies rare

**Objective Findings:**
- Possible increased respiratory rate
- ALWAYS DECREASED PULSE OX
- Possible signs of respiratory distress
- Murmur occasionally present

**Acute Management:**
- Oxygen rarely wrong...
- Starting Prostaglandin E1 rarely wrong...

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Pulmonary Blood Flow-Dependent Cyanosis

*a.k.a. – “Tetralogy Physiology”*

Presentation:
- Asymptomatic to tachypneic
- Timing dependent upon amount of pulmonary blood flow (Birth – Two Weeks)
- LOOK for signs of increased work of breathing

Chest Radiograph:
- Classic finding – decreased pulmonary vascular markings
- Generally normal heart size (exception: Ebstein anomaly)
- Occasional associated anomalies

Objective Findings:
- Possible increased respiratory rate
- ALWAYS DECREASED PULSE OX
- Possible signs of respiratory distress
- Murmur frequently present

Acute Management:
- Sometimes nothing...
- Oxygen rarely wrong...
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“Once I realized the problem was inadequate pulmonary blood flow, the solution was simple”

Helen Taussig
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Pulmonary Venous Obstruction  
*a.k.a. – “Obstructed Veins Physiology”*

**Presentation:**
- Progression, refractory tachypnea / cyanosis
- Time course over hours
- CONSIDER in infant with progressive worsening course with...

**Objective Findings:**
- Tachypnea, tachycardia
- Refractory, low PULSE OX
- Progressive signs of respiratory distress
- Murmur may be present late

**Chest Radiograph:**
- Classic finding – progressive increase in PVM until “white-out”
- Generally normal heart size
- Infrequent associated anomalies

**Acute Management:**
- SURGICAL EMERGENCY
- Intubate, paralysis, sedate...
- Paradoxical worsening with Prostaglandin E, initiation...

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Objective Findings:
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• Refractory, low PULSE OX
• Progressive signs of respiratory distress
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Acute Management:
• Surgical Emergency
• Intubate, paralyze, sedate...
• Paradoxical worsening with Prostaglandin E, initiation...
Pulmonary Venous Obstruction

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- SURGICAL EMERGENCY
- Intubate, paralyze, sedate...
- Paradoxical worsening with Prostaglandin E1 initiation...

---

Left Cardiac Output Obstructive Lesions

*a.k.a.* – “Failing Cardiac Output Physiology”

**Presentation:**
- Progressive tachypnea, lethargy
- Timing dependent upon ductal size (hours - days)
- LOOK for signs gradual cardiac decompensation

**Chest Radiograph:**
- Findings often non-specific – variable pulmonary vascular markings, mild cardiomegaly
- Rare associated anomalies

**Objective Findings:**
- Increased respiratory & heart rate
- PULSE OX may be normal
- Signs of respiratory and cardiac distress
- Murmur frequently present

**Acute Management:**
- Oxygen only to keep sats in 70s...
- Start Prostaglandin E1 quickly...
- Sometimes intubate, paralyze and sedate...
- Sometimes inotropes...

---

"Once I realized the problem was inadequate pulmonary blood flow, the solution was simple"

*Helen Taussig*

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- Increased respiratory & heart rate
- **Pulse Ox may be normal**
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**Acute Management:**
- Oxygen only to keep sats in 70s...
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**Ventricular Compliance-Dependent Shunting**  
*a.k.a. – “ASD Physiology”*

**Presentation:**  
- Asymptomatic to tachypneic  
- Sometimes mimicking infection  
- Timing dependent upon degree of shunt (Birth – Years)  
- LOOK for signs of increased work of breathing

**Chest Radiograph:**  
- Classic finding – increased pulmonary vascular markings  
- Increased heart size  
- Occasional associated anomalies

**Objective Findings:**  
- Possible increased respiratory rate  
- In to slight decrease PULSE OX  
- Possible signs of respiratory distress  
- Murmur “diagnostic” – split S2, pulmonary flow murmur

**Acute Management:**  
- Often nothing...  
- Diuretics...

---

“Once I realized the problem was inadequate pulmonary blood flow, the solution was simple”  
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Timing dependent upon degree of shunt (Birth – Years)
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**Chest Radiograph:**
- Classic finding – increased pulmonary vascular markings
- Increased heart size
- Occasional associated anomalies

**Objective Findings:**
- Possible increased respiratory rate
- May or slight decrease PULSE OX
- Possible signs of respiratory distress
- Murmur “diagnostic” – split S2, pulmonary flow murmur

**Acute Management:**
- Often nothing...
- Diuretics...

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Pulmonary Vascular Resistance-Dependent Shunting

*a.k.a. – “VSD Physiology”*

"Once I realized the problem was inadequate pulmonary blood flow, the solution was simple"
Helen Taussig

**Presentation:**
- Asymptomatic to tachypneic
- Timing dependent upon amount of pulmonary blood flow (Birth – Months)
- LOOK for signs of increased work of breathing, early fatigue

**Chest Radiograph:**
- Classic finding – increased pulmonary vascular markings
- Increased heart size
- Occasional associated anomalies

**Objective Findings:**
- Possible increased respiratory rate
- Usually normal PULSE OX
- Possible signs of respiratory distress
- Murmur frequently present, often diagnostic

**Acute Management:**
- Diuretics...
- Afterload Reduction...
- Digoxin...
- Intubation, paralysis, sedation...

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Acute Management:
• Diuretics...
• Afterload Reduction...
• Digoxin...
• Intubation, paralysis, sedation...

Pulmonary Vascular Resistance-Dependent Shunting
a.k.a. – “VSD Physiology”

Ferguson EC, Krishnamurthy A, Oldham SA. Classic imaging signs of congenital cardiovascular abnormalities. Radiographics. 27 (5): 1323-34.
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Putting It All Together

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<th>TGA Physiology</th>
<th>Obstructed Veins</th>
<th>Low Cardiac Output</th>
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<th>VSD Physiology</th>
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Concluding Thoughts
• Use the multiple “versions of the truth…”
• Paint the picture…
• Treat the physiology… and re-assess…

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