Case Scenarios in Congenital Heart Disease

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Case #1

You are called to the newborn nursery because several newborns failed pulse oximetry screening test.
Which Saturation for Which Disease?

D-TGA
HLHS (Hypoplastic Left Heart Syndrome)
Pneumonia
PPHN (Primary Pulm. Hypertension of the Newborn)

Stolen from Dr. Sulaiman with permission
Pulse Oximetry in Critical CHD

POx in RH & RF at 24 hrs

1. $\geq 95\%$ in RH &
2. $\geq 95\%$ in RF &
3. $<4\%$ difference

Borderline?

Repeat after 1 hr

Borderline?

Repeat after 1 hr

PASS

FAIL

1. $< 90\%$ in RH or
2. $< 90\%$ in RF or
3. $>4\%$ difference

Kemper AR et al., Pediatr 2011
Detection of Critical CHD by Pulse Oximetry: Preliminary Data from the UAE

Mohamed Hamdan, MD, FAAP, FACC

Friday 20 April, 2012
Screening Protocol

Pulse oximetry of right foot $\geq 24$ hours of age

Reading $<95\%$

FAIL

Cardiology Consultation & Echo

Reading $\geq 95\%$

PASS (Discharge)
# Results

**Jan. 1st – Dec. 31st, 2011**

<table>
<thead>
<tr>
<th>Item</th>
<th>Value</th>
<th>95% C.I.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sensitivity</td>
<td>91%</td>
<td>60-95</td>
</tr>
<tr>
<td>Specificity</td>
<td>99.7%</td>
<td>99.5-99.9</td>
</tr>
<tr>
<td>PPV</td>
<td>48%</td>
<td>33-80</td>
</tr>
<tr>
<td>NPV</td>
<td>99.5%</td>
<td>99.3-99.7</td>
</tr>
</tbody>
</table>

**Inc. of CCHD:** 1.9/1000 LB (95% C.I. 0.6-5.3)

**Cost:** 28 Dhs (7 USD) per screened newborn

- True Positive: 1 (0.2/1000)
- Additional 9 positive by fetal Echo
- True positive: 2.3/1000

- False Positive: 11 (2.5/1000)

- False Negative: 0

**Median age:** 26 hrs

**Median Wt:** 2948 g

**Median LOS:** 41 hrs
Understanding Cyanotic CHD
Cyanotic CHD

❖ Cyanosis= Right to Left Shunt
  ❑ Intrapulmonary: Pneumonia
  ❑ Intracardiac: Cyanotic CHD

❖ In cyanotic CHD:
  ❑ Mixing of oxygenated and de-oxygenated blood
due to either:
    ▪ Obstructed pulm. blood flow (↓ PBF)
      (TOF, Pulmonary atresia, Tricuspid atresia, Ebstein)
    ▪ Abnormal vessel connection (↑ PBF)
      (D-TGA, TAPVR, Truncus)
## Hyperoxia Test: *Lung vs Heart?*

<table>
<thead>
<tr>
<th>Value</th>
<th>Normal</th>
<th>Cyanotic CHD</th>
<th>Lung Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>pO2</strong></td>
<td>≥ 90</td>
<td>&lt; 60</td>
<td>&lt; 60</td>
</tr>
<tr>
<td><strong>(Room Air)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>pO2</strong></td>
<td>600</td>
<td>≤ 150</td>
<td>≥ 400</td>
</tr>
<tr>
<td><strong>(100% O2)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>pCO2</strong></td>
<td>&lt; 40</td>
<td>&lt; 35</td>
<td>&gt; 45</td>
</tr>
</tbody>
</table>
Normal Heart

RA

LV

RV

LA

MPA

Ao
D-TGA: ↑ PBF
Truncus Arteriosus

Truncus: ↑ PBF

RA  RV  LA  LV  MPA  Ao

Truncal Vessel
TAPVR: Supracardiac TAPVR

- Pulm. veins
- RA
- LA
- RV
- LV
- MPA
- Ao

TAPVR: ↑ PBF
Tetralogy of Fallot

TOF: ↓ PBF

Pulm. atresia/VSD: ↓ PBF
Tricuspid Atresia

**Tricuspid atresia:**
VSD determines cyanosis
Small/absent VSD: ↓PBF
Ebstein Anomaly

4D’s
- Displacement
- Dysfunction
- Desaturation
- Dysrhythmia
Hints to Cyanotic CHD

❖ History

- Cyanosis in 1st hour: D-TGA
- No improvement with O2 (positive hyperoxia test)

❖ Exam

- Comfortable tachypnea
- Minimal or absent murmurs
- Normal perfusion & pulses
Case #2

❖ A 10 d/o male neonate presents to ER with a 3 day h/o respiratory distress, poor deeding, and hypoactivity

❖ Exam:
  • Afebrile, lethargic
  • Moderate respiratory distress
  • No murmurs
  • Weak pulses and poor perfusion
What’s your diagnosis?
Sepsis vs Cardiogenic shock
Causes of Non-traumatic Shock

N = 480

- Respiratory: 37%
- CNS: 17%
- Cardiac: 12%
- Septic: 10%
- Other: 24%

Updated American College of Critical Care Medicine–Pediatric Advanced Life Support Guidelines for Management of Pediatric and Neonatal Septic Shock

Relevance to the Emergency Care Clinician

Niranjan Kissoon, MD,* Richard A. Orr, MD,† and Joseph A. Carcillo, MD†

Abstract: Shock is a major preventable cause of morbidity and mortality in children referred to emergency care. The recently updated American College of Critical Care Medicine guidelines for the management of newborns and children with septic shock emphasize the role of emergency care in improving survival and functional outcomes. Implementation of these guidelines of stepwise use of fluids, antibiotics, and, if necessary, inotropes within the first hour of admission to the emergency department can reduce mortality and neurological morbidity risks 2-fold. Therapies should be goal directed to maintain age-specific threshold heart rates and blood pressure as well as a capillary refill of less than 3 seconds or 2 seconds or less. Inotropes should be delivered through peripheral intravenous or intrasosseous access when central access is unavailable because delay in inotrope delivery can greatly increase mortality risks. Emergency care systems should be organized to facilitate recognition, triage, and treatment of shock in the first hour. Emergency departments should be stocked with ready access to antibiotics, fluids, and inotrope infusions, and clinicians should be trained in the delivery of goal-directed fluid, antibiotics, and inotrope therapies in the first hour of resuscitation. For newborns, in addition to fluids, antibiotics, and inotropes, a prostaglandin infusion should be available within 10 minutes if duct-dependent congenital heart disease is a possibility.
A Neonate with Shock

- **Cardiogenic shock more likely than septic shock**
  - In a neonate with shock: *think ductus*

- **Almost always: left heart obstruction**
  - Coarctation of the aorta
  - Aortic stenosis
  - Hypoplastic left heart syndrome
Coarctation of the aorta

HLHS

Aortic stenosis
Effect of PGE1 on the Ductus

- Pulm. Artery
- PDA
- Desc. Aorta
Management

❖ Supportive (PALS/ APLS)
  ❑ ABC
  ❑ Fluids
  ❑ Inotropes
  ❑ Intubation

❖ Specific (PGE₁)
  ❑ Life saving
    - Even in doubt: Wouldn’t harm even if the diagnosis is septic shock
    - 0.01 mcg/kg/min infusion
    - Double the dose if no improvement
    - Watch for apnea
    - Other S.E.: flushing, jitteriness, hypoglycemia

❖ Morbidity correlates with duration & severity of acidosis
### Impact of Prenatal Diagnosis on Survival and Early Neurologic Morbidity in Neonates with Hypoplastic Left Heart Syndrome

2001;107:1277-82

<table>
<thead>
<tr>
<th>Variable</th>
<th>Prenatal Dx</th>
<th>Postnatal Dx</th>
<th>( p )</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>n=79</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Delivery in advanced nursery</td>
<td>99%</td>
<td>55%</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>DC to home before dx</td>
<td>9%</td>
<td>99%</td>
<td>0.004</td>
</tr>
<tr>
<td>Age at PGE1</td>
<td>1 day</td>
<td>2 days</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Age at hospital admission</td>
<td>1 day</td>
<td>6 days</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Base deficit</td>
<td>-4</td>
<td>-11</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Age at surgery</td>
<td>4 days</td>
<td>8 days</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

**N= 216**
Impact of Prenatal Diagnosis on Survival And Early Neurologic Morbidity in Neonates with Hypoplastic Left Heart Syndrome 2001;107:1277-82

<table>
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<tr>
<th>Variable</th>
<th>Prenatal Dx n=79</th>
<th>Postnatal Dx n=137</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Preoperative seizure</td>
<td>1.3%</td>
<td>5.1%</td>
<td>0.26</td>
</tr>
<tr>
<td>Operative coma</td>
<td>1.3%</td>
<td>8.1%</td>
<td>0.06</td>
</tr>
<tr>
<td>Overall neurologic event</td>
<td>15%</td>
<td>26%</td>
<td>0.06</td>
</tr>
<tr>
<td>Overall death</td>
<td>24%</td>
<td>28%</td>
<td>0.63</td>
</tr>
</tbody>
</table>
Take Home Message

❖ Understand the value of pulse oximetry

❖ In neonate with shock: Think ductus
SAVE THE DATE

THE 8th INTERNATIONAL PEDIATRIC CONFERENCE

of Columbia University Medical Center / New York-Presbyterian Hospital
&
KidsHeart Medical Center
(Abu Dhabi - Dubai - Al Ain)

11 - 13 October 2019

Danat Al Ain Resort
(InterContinental Hotel)
Al Ain, UAE

www.kidsheart.ae