Lecture: Pediatric Cardiology-
Little Boy Blue: Lessons from Blue Babies

Jimmy Lu, MD
ACOFP FULL DISCLOSURE FOR CME ACTIVITIES

Please check where applicable and sign below. Provide additional pages as necessary.

Name of CME Activity: ACOFP 52nd Annual Convention and Scientific Seminars

Dates and Location of CME Activity: March 12-15, 2015, The Cosmopolitan Las Vegas, Nevada

Lecture: Pediatric Cardiology - Little Boy Blue: Lessons from Blue Babies

Sunday, March 15, 2015 8:00-9:00 am

Name of Faculty/Moderator: Jimmy Lu, MD

DISCLOSURE OF FINANCIAL RELATIONSHIPS WITHIN 12 MONTHS OF DATE OF THIS FORM

A. Neither I nor any member of my immediate family has a financial relationship or interest with any proprietary entity producing health care goods or services.

B. I have, or an immediate family member has, a financial relationship or interest with a proprietary entity producing health care goods or services. Please check the relationship(s) that applies:

<table>
<thead>
<tr>
<th>Organization With Which Relationship Exists</th>
<th>Clinical Area Involved</th>
</tr>
</thead>
<tbody>
<tr>
<td>Research Grants</td>
<td>Stock/Bond Holdings (excluding mutual funds)</td>
</tr>
<tr>
<td>Speakers' Bureaus*</td>
<td>Employment</td>
</tr>
<tr>
<td>Ownership</td>
<td>Partnership</td>
</tr>
<tr>
<td>Consultant for Fee</td>
<td>Others, please list</td>
</tr>
</tbody>
</table>

Please indicate the name(s) of the organization(s) with which you have a financial relationship or interest, and the specific clinical area(s) that correspond to the relationship(s). If more than four relationships, please list on separate piece of paper:

1. ________________________________________________________________
   Clinical Area: __________________________________________________

2. ________________________________________________________________
   Clinical Area: __________________________________________________

3. ________________________________________________________________
   Clinical Area: __________________________________________________

4. ________________________________________________________________
   Clinical Area: __________________________________________________

*If you checked “Speakers’ Bureaus” in item B, please continue:

- Did you participate in company-provided speaker training related to your proposed Topic?  
  Yes:  No:

- Did you travel to participate in this training?  
  Yes:  No:

- Did the company provide you with slides of the presentation in which you were trained as a speaker?  
  Yes:  No:

- Did the company pay the travel/lodging/other expenses?  
  Yes:  No:

- Did you receive an honorarium or consulting fee for participating in this training?  
  Yes:  No:

- Have you received any other type of compensation from the company? Please specify:  
  Yes:  No:

- When serving as faculty for ACOFP, will you use slides provided by a proprietary entity for your presentation and/or lecture handout materials?  
  Yes:  No:

- Will your Topic involve information or data obtained from commercial speaker training?  
  Yes:  No:

DISCLOSURE OF UNLABELED/INVESTIGATIONAL USES OF PRODUCTS

A. The content of my material(s)/presentation(s) in this CME activity will not include discussion of unapproved or investigational uses of products or devices.

B. The content of my material(s)/presentation in this CME activity will include discussion of unapproved or investigational uses of products or devices as indicated below:

I have read the ACOFP policy on full disclosure. If I have indicated a financial relationship or interest, I understand that this information will be reviewed to determine whether a conflict of interest may exist, and I may be asked to provide additional information. I understand that failure or refusal to disclose, false disclosure, or inability to resolve conflicts will require the ACOFP to identify a replacement.

Signature: __________________________________________ Date: 12/8/14

Jimmy Lu, MD

Please fax this form to ACOFP at 866-328-1835 or email to joank@acofp.org as soon as possible

Deadline: Monday, January 12, 2015
Little Boy Blue: Lessons from Blue Babies

Jimmy C. Lu, MD
Assistant Professor, Pediatric Cardiology
University of Michigan

Disclosures

- No financial relationships to disclose.
Goals

- Understand key concepts about cyanosis
- Provide a conceptual framework for cyanotic CHD
- Discuss potential interventions (medical and surgical)

Case Presentation

- You are called to a delivery at a community hospital.
- History: 24 yo G1P0 mother, serologies negative, scheduled C/S at 39 weeks. Pregnancy unremarkable. ROM clear.
- Resuscitation: Unremarkable; good resp effort, tone and HR, but the baby never really gets pink, despite BBO2.
Time out – is the baby really blue?

- Peripheral cyanosis: normal in babies
- Hands and feet, perioral

Peripheral cyanosis
Peripheral cyanosis

- Related to poor circulation
- Venous desaturation

Central cyanosis

- Requires 3-5 g/dl of desaturated Hgb
- Hgb 18: cyanotic at ~80%
- Arterial desaturation
- Well-vascularized regions (lips, mucous membranes) are blue despite good blood flow
So how am I supposed to figure this out?

blog.babyheartscreening.com

AHA/AAP Scientific Statement

Role of Pulse Oximetry in Examining Newborns for Congenital Heart Disease
A Scientific Statement From the American Heart Association and American Academy of Pediatrics

William T. Mahle, MD, FAHA, FAAP, Chair; Jane W. Newburger, MD, MPH, FAHA, FAAP; G. Paul Matherne, MD, FAHA, FAAP; Frank C. Smith, MD; Tracey R. Hoke, MD, FAAP; Robert Koppel, MD, FAAP; Samuel S. Gidding, MD, FAHA, FAAP; Robert H. Beekman III, MD, FAHA, FAAP; Scott D. Grosse, PhD;
on behalf of the American Heart Association Congenital Heart Defects Committee of the Council on Cardiovascular Disease in the Young, Council on Cardiovascular Nursing, and Interdisciplinary Council on Quality of Care and Outcomes Research; and the American Academy of Pediatrics Section on Cardiology and Cardiac Surgery, and Committee on Fetus and Newborn

Pediatrics 2009; Circulation 2009
Pulse oximetry screening

- Estimated 1200 additional newborns with critical CHD could be detected\(^1\)
  - Estimated 20 deaths averted per year
- Cost of screening similar to other NBS\(^2\)

Where is the baby blue?

- Pre-ductal (right arm)
  - Blood coming from the heart is blue

Where is the baby blue?

- Post-ductal (or drop from pre-ductal)
  - High PVR (e.g. PPHN)
  - Ductal-dependent systemic flow
    - Interrupted aortic arch
    - Hypoplastic left heart syndrome
Back to the Case: Physical Exam

- Vitals: afebrile, HR 160s, RR 60s
- Sats in 70s, both pre- and post-ductal.
- General: mildly tachypneic, but relatively comfortable.
- HEENT: nondysmorphic, palate intact
- Resp: Clear bilaterally. No grunting.
- CV: Mildly tachycardic but regular. No murmur. 2+ femoral pulses. CR<2 sec.
- Abd: Soft, no hepatomegaly.
- Skin: cyanotic lips, trunk
Next move?

Specialists’ favorite tricks:
1. Stall
2. Blame someone else’s organ system

Hyperoxia test: Can I blame the lungs?

- ABG on room air
  - Elevated CO$_2$: hypoventilation or obstruction, e.g. primary pulmonary or CNS problem
- ABG (R radial) on 100% oxyhood
  - PaO$_2$ < 120: shunt
  - Disclaimer: you might not want to try 100% FiO2 on certain babies
**Hyperoxia test**

- Small decrease in PaO₂ → large decrease in O₂ sats
- Increase PaO₂ → minimal inc O₂ sats
- Plenty to lose, not much to gain → hard to compensate

**Hyperoxia test (continued)**

- Pink + Blue = Blue
Back to the Case

**Summary of findings:**
- cyanotic newborn
- tachypneic but not retracting
- clear BS, CXR with nl to inc lung markings but no focal pulmonary findings
- desaturated, not responsive to O2, and failed hyperoxia test
- no murmur

**Differential diagnosis?**

---

**Cyanotic Congenital Heart Disease (If you’re studying for boards)**

- D-TGA
- Tricuspid atresia
- Truncus arteriosus
- Tetralogy of Fallot
- TAPVC
If you actually care about physiology...

In a normal heart:
- Blue blood from the body is sent to the lungs
- Pink blood from the lungs is sent to the body

How can blue blood get to the body?

- It can’t get to the lungs.
- It mixes in the heart before it leaves.
- The blue blood goes out the wrong door.
Blue blood can’t get to the lungs

- Right-sided obstruction
  - Abnormal tricuspid valve (tricuspid atresia*, Ebstein)
  - Abnormal pulmonary valve (critical PS, PA-IVS, TOF)
- PPHN
- Note: Need right-to-left shunt for cyanosis (ASD/PFO, VSD, PDA)

What would these kids look like?

- Blue?
  - Duh (that’s the point of this talk)
  - May not be visibly apparent
    - May not get really blue until the PDA is closing
- CXR?
  - Decreased pulmonary markings
- EKG?
  - Tricuspid atresia has a characteristic left axis deviation (superior axis)
Can I give oxygen?

- Not enough blood to the lungs
- Oxygen dilates pulmonary vessels, increases blood flow
  - Should not hurt
  - (may not help)

Special case: Hypercyanotic spells

Little Boy Blue, come blow your horn,
The sheep’s in the meadow, the cow’s in the corn
Where is that boy who looks after the sheep?
Under the haystack fast asleep.
Will you wake him? Oh no, not I,
For if I do he will surely cry.
You wouldn’t like me when I’m angry.

Tetralogy of Fallot

- VSD and RVOT obstruction
- Blood takes the path of least resistance
Tetralogy of Fallot – “Tet” spells

- Baseline blueness can worsen
  - Infundibular spasm
  - Increased PVR
  - Drop in systemic resistance

Tetralogy of Fallot – “Tet” spells

- Baseline blueness can worsen
  - Infundibular spasm
  - Increased PVR
  - Drop in systemic resistance

- Call for help
  - Calm the baby
  - Oxygen
  - Knee to chest position
How can blue blood get to the body?

- It can’t get to the lungs.
- It mixes in the heart before it leaves.
- The blue blood goes out the wrong door.

The blood mixes before it leaves

- **Pink** + blue = blue
- Mix in the right atrium (e.g. TAPVC)
- Mix in a single ventricle (e.g. HLHS)
- Mix in a single artery (Truncus arteriosus)
- Note: If it mixes left to right (and goes to the lungs), it’s not cyanotic
What would these kids look like?

- **Blue?**
  - Maybe not that blue (lots of PBF)
  - HLHS: sats often in 90s, with crazy tachypnea
- **CXR?**
  - Normal to increased pulmonary markings
- **EKG?**
  - Not very helpful

What would these kids look like?

- **Blood takes the path of least resistance**
  - Lots of PBF → lots of work for the heart
  - Blue because of mixing
What would these kids look like?

- Wet lungs, lots of work for the heart → congestive heart failure
  - Tachypnea
  - Tachycardia
  - Hepatomegaly
  - Cardiomegaly

Special case: single ventricle

- Blood can go to the lungs or the body
  - Lots of pulmonary flow: pink, poor perfusion
  - Lots of systemic flow: blue, better perfusion
Special case: TAPVC

- Lots of PBF
  - Sats often >90%
  - Can have CHF, or decompensate with resp illness
- Obstructed pulmonary veins
  - Blue AND shock
  - Surgical emergency

How can blue blood get to the body?

- It can’t get to the lungs.
- It mixes in the heart before it leaves.
- The blue blood goes out the wrong door.
Blue blood goes out the wrong door

- Transposition of the Great Arteries (D-TGA)
  - Blue blood from the RV goes out the aorta
  - Note: there must be some connection between the circuits for survival

CXR in Cyanotic CHD

**Dec pulm markings**
- Tricuspid atresia*
- Ebstein’s anomaly
- Critical pulmonary stenosis
- Pulmonary atresia with intact ventricular septum
- Tetralogy of Fallot
  - (PPHN)

**NI to ↑ pulm markings**
- TAPVC
  - Small heart if obstruction
- Single ventricle
- Truncus arteriosus
- D-TGA

* Some forms of tricuspid atresia actually have increased pulmonary blood flow

Think before giving O2!
Cyanotic cheat sheet

- Blue with black lungs: right heart obstruction
- (Blue) with CHF: mixing lesion (single ventricle, unobstructed TAPVC, maybe D-TGA)
- Blue with shock: obstructed total veins
EKG

What do we do?

- **ABC’s**
  - A/B: Patient a little tachypneic, but not in distress
  - C: Baby needs access, preferably central → umbilical lines

- **Call cardiology**

- **Consider a potentially life-saving medication...**
Prostaglandin (E₁)

- If you’re thinking about using prostaglandin, DO IT
  - It will not hurt (unless you have obstructed TAPVC)
  - It may save the baby’s life

Prostaglandin (E₁)

- **Mechanism: keep the ductus open**
  - Lesions with decreased PBF: increases PBF via L → R shunt
  - Single ventricle: allows 1 ventricle to feed both body and lungs
  - Poor mixing (D-TGA): encourages mixing between the 2 circuits
Prostaglandin (E₁)

- **Dose:** 0.03-0.1 mcg/kg/min (infusion)
  - Newborn: 0.03 mcg/kg/min to keep duct open
  - Higher dose if the duct is closed/closing
- **Side Effects:** hypotension, apnea, hyperthermia

**What else can you do?**

Blue blood  Pink blood

![Shaker](image1.png)  ![Martini Glass](image2.png)
What else can you do?

- **Make the blue blood less blue (SvO2)**
  - Transfuse (keep Hct >40)
  - Give fluids
  - Decrease metabolic demand (sedate, paralyze)

- **Make the pink blood pinker**
  - O2 (if not total mixing/single ventricle)
  - If the kid gets more tachypneic, a little pinker and has worse perfusion, you may have guessed wrong

What else can you do?

- **Change the recipe (Qp:Qs)**
  - Prostaglandin
  - O2, NO increase Qp (PPHN)
  - Decreased FiO2 (inhaled nitrogen) decrease Qp
What will cardiology do (after the echo)?

- **Nothing**
  - Stable sats >75%, not ductal dependent
  - Some RVOT obstruction (eg TOF), unobstructed TAPVC \(\rightarrow\) repair later

- **Increase PBF**
  - Balloon valvuloplasty for PS or PA-IVS
  - Modified BT shunt for stable PBF

- **Single ventricle pathway**
- **Repair (truncus, D-TGA) vs. palliation**

---

The punchline – D-TGA

- **Pulmonary valve arises from the LV**
- **Parallel circuits, not in series**
  - Problem: lack of mixing
- Can be easily missed on routine prenatal ultrasound
Clues in diagnosis

- Most common cyanotic CHD in the newborn
  - Boys > girls (3:1)
  - Associated with maternal diabetes
  - Note: tetralogy of Fallot is the most common cyanotic CHD

- Presentation: Cyanosis from birth
  - May present with CHF in first week, if good mixing (ASD, PDA)

Clues in diagnosis

- Exam: loud single S2, usually no murmur
  - Pulmonary valve posterior, often do not hear closure

- CXR: narrow mediastinum
  - PA and aorta more in line; may not see aortic knob
  - Cardiomegaly \(\rightarrow\) “egg on a string”
  - Normal to increased pulmonary vascularity
Management

- Ensure mixing
  - Prostaglandin
  - Connects the two circuits
Management – All about that BAS

- **Ensure mixing**
  - Balloon atrial septostomy if necessary
  - Done by a cardiologist at bedside or cath lab

---

Post Balloon Atrial Septostomy
Definitive repair

- Arterial switch (Jatene)
  - Coronary arteries must be reimplanted
  - Risk of coronary problems (stenosis, etc)
  - Potential pulmonary artery stenosis

From Park, Pediatric Cardiology for Practitioners, 5th Ed. 2008

Take-home points: CHD math facts

- Pulse ox > physical exam → newborn screening
- Pink + Blue = Blue → hyperoxia test
- Blue + CHF = fear of oxygen
- Blue + shock = emergency
- No prostaglandin = big trouble
Take-home points: What can you do?

- Screen with pulse oximetry
- Recognize a hypercyanotic spell
- Start the diagnostic process
  - Hyperoxia test
  - CXR

---

Take-home points: What can you do?

- Think through first management steps
  - Think about when to give oxygen
  - Remember prostaglandin
  - Support: ABCs, fluids, blood
- Call for help
Where is the baby blue?

- Post-ductal higher
  - D-TGA with arch obstruction/pulm HTN