Am I Blue?
Newborn Pulse Oximetry Screening for Critical Congenital Heart Disease
“The State of the State”

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The Barbara Bush Children’s Hospital at MMC
Maine AAP Spring Conference
May 5th, 2013
Overview

- Briefly review the data to support screening
- Maine’s approach to screening
- Regional Collaboration
- Data Review to date
- What’s working, what’s not
- Legislative Initiative and Issues
Objectives

• State rationale for screening newborns for Critical Congenital Heart Disease
• Describe screening process
• Discuss materials available to support implementation of Critical Congenital Heart Disease screening at your facility
Points to leave with

• Newborn saturation screening is happening in ME
• Clinical examination misses critical CHD
• Oximetry is stable and reliable
• False positive rates are low (lower than false positive rate based on physical exam)
• Data to support this is strong
• Some lesions will not be detected
Overview

• CHD leading cause of infant death
  – 40% of all deaths from congenital defects
  – 3-7.5% of infants deaths are due to cardiac anomalies

• Failure to detect early increases the risk of circulatory collapse
  – Adverse effect on prognosis
  – Poor clinical status at time of surgery increases surgical mortality
Congenital Heart Diseases: The magnitude of the problem

**CHD:**
- 5-10 / 1,000 live births
- 1.4 cyanotic CHD / 1,000 live births
- 2 critical CHD / 1,000 live births
- 25,000 cases of CHD/yr in US
- 25% of infantile deaths
- 31% of neonatal deaths


Screening strategies for Critical Heart Disease

- Physical examination
- Fetal echocardiography
- Oximetry
  - Most critical heart disease produces some degree of cyanosis not visible to examiner
  - To date uncertainty exists regarding false positive rate and test accuracy
  - Recent meta-analysis in Lancet May 2012 provides important data in 229,421 newborn babies

- Critical heart disease
  - Duct dependent systemic or pulmonary circulation
  - Surgery required within 28 days of birth
  - Examples
    - HLHS, PA-IVS
    - Aortic Stenosis
    - TAPVR
    - Severe coarctation
Missed Critical Congenital Heart Diseases (CCHD)

<table>
<thead>
<tr>
<th>First author</th>
<th>Years</th>
<th>CCHD</th>
<th>Missed CCHD</th>
</tr>
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<tbody>
<tr>
<td></td>
<td></td>
<td>live born</td>
<td>/1,000 live births</td>
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<tr>
<td></td>
<td></td>
<td>prenatal Dx</td>
<td>postnatal Dx</td>
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<tr>
<td>Aamir [19]</td>
<td>1999–2004</td>
<td>18 94</td>
<td>0.2</td>
</tr>
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<td>9 109^{4}</td>
<td>1.0</td>
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<td></td>
<td></td>
<td>110^{6}</td>
<td>–</td>
</tr>
<tr>
<td>Meberg [23]</td>
<td>2005–2006</td>
<td>31 50^{7}</td>
<td>1.2</td>
</tr>
<tr>
<td></td>
<td></td>
<td>7 48^{8}</td>
<td>–</td>
</tr>
</tbody>
</table>

Hoffman, J. It is time for routine neonatal screening by pulse oximetry. Neonatology 2011;99:1-9
50% of deaths from CHD occur in 1st year and
50% of infantile deaths occur in 1st month of life

Boneva, R: Circulation. 2001;103:2376
Timeline of pulse oximetry screen for CCHD

- **2002**: Oximetry screen 1st clinical reports
- **2005**: Larger prospective study in NY (Koppel)
- **2008**: Large European prospective studies (Norway, Sweden, UK, Germany, Switzerland)
- **2008**: Several European countries adopt pulse-ox screen as standard of care
- **2008**: AHA comment: Evidence not sufficient
- **2008**: Proposed bill in TN to mandate screening
- **2008**: SACHDNC (HHS) recommended adding pulse-ox screen
- **2008**: AHA/AAP statement comment
- **2011**: AAP endorses HHS recommendations
- **2012**: Maine Hospitals start screening
- **2012**: 2002-2005: Large European prospective studies (Norway, Sweden, UK, Germany, Switzerland)
Workgroup convened...

• Primary Care Providers, Pediatric Cardiologists and neonatologists, nurses, AAP, ACCF, AHA, ACMG, March of Dimes, Assoc. of Maternal and Child Health Programs, Association of Public Health Laboratories, the SACHDNC, parent screening advocates, state public health officials, CDC Reps, USFDA, HRSA, and NIH

• Led by William Mahle, MD* and R. Rodney Howell, MD**
  – *Led the development of the AAP/AHA statement
  – **Chair of the SACHDNC

• Meeting focused on recommendations for pulse-oximetry monitoring for CCHD including recommendations for the service infrastructure needs for f/u, and strategies for education
SACHDNC recommendations

♥Research:
♥ NIH shall fund research activities to determine the relationships among the screening technology, diagnostic processes, care provided, and then health outcomes of affected newborns with CCHD as a result of prospective newborn screening

♥Surveillance:
♥ CDC shall fund surveillance activities to monitor the CCHD link to infant mortality and other health outcomes
SACHDNC recommendations

♥ Screening Standards and Infrastructure:
  ♥ HRSA shall guide the development of screening standards and infrastructure needed for the implementation of a public health approach to point of service screening for Critical Congenital Cyanotic Heart Disease

♥ Education and Training:
  ♥ HRSA shall fund the development of, in collaboration with public health and health care professional organizations and families, appropriate education and training materials for families and public health and health care professionals relevant to the screening and treatment of CCHD
Why Screen

♥ Critical CHD has a higher frequency than other conditions that are universally screened for in the newborn nursery, including hypothyroidism and phenylketonuria

♥ Screening for critical CHD costs less than other universal newborn screenings

♥ Pulse Oximetry is an easy test to do, is painless, and a non-invasive way of measuring the oxygen saturation of hemoglobin in the arterial blood (the “5th” vital sign)
Pulse Oximetry

<table>
<thead>
<tr>
<th>Hemoglobin of 17.5 g/dL</th>
<th>83%</th>
<th>95%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abnormal Saturation</td>
<td>Visible Cyanosis</td>
<td>Abnormal Saturation</td>
</tr>
<tr>
<td>78%</td>
<td>95%</td>
<td></td>
</tr>
</tbody>
</table>

Hemoglobin of 13.5 g/dL
O₂ saturation values in patients with CCHD

Pulse oximetry screening for critical congenital heart defects in asymptomatic newborn babies: a systematic review and meta-analysis

Shakila Thangaratnam, Kiritea Brown, Javier Zamora, Khalid S Khan, Andrew K Ewer

- Critical defects
  - All lesions that are duct dependent and required surgery in first 28 days after birth
- 552 studies, 13 primary studies were eligible for inclusion, 229,421 newborns were included
- Variability in studies included – inclusions of antenatal dx (4/12), timing of oximetry (5/12 less than 24 hours), foot alone (60%) or in conjunction with right hand, length of f/u.
<table>
<thead>
<tr>
<th>Limb</th>
<th>Antenatal diagnosis of CHD</th>
<th>Test timing</th>
<th>Total</th>
<th>True positive</th>
<th>False positive</th>
<th>False negative</th>
<th>True negative</th>
<th>Sensitivity (%; 95% CI)</th>
<th>Specificity (%; 95% CI)</th>
<th>Likelihood ratio positive (%; 95% CI)</th>
<th>Likelihood ratio negative (%; 95% CI)</th>
<th>False-positive rate (%; 95% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Meberg et al (2008) a</td>
<td>Foot only</td>
<td>Excluded</td>
<td>&lt;24 h</td>
<td>50008</td>
<td>27</td>
<td>297</td>
<td>8</td>
<td>496/76</td>
<td>77.1% (59.9–89.6)</td>
<td>99.4% (93.3–99.5)</td>
<td>129.8% (104.9–160.6)</td>
<td>0.23% (0.13–0.43)</td>
</tr>
<tr>
<td>Bakr et al (2005) b</td>
<td>Foot and right hand</td>
<td>Excluded</td>
<td>&gt;24 h†</td>
<td>5211</td>
<td>3</td>
<td>2</td>
<td>0</td>
<td>5206</td>
<td>100.0% (29.2–100.0)</td>
<td>100.0% (99.9–100.0)</td>
<td>1823.1% (500.1–6646.1)</td>
<td>0.13% (0.01–1.67)</td>
</tr>
<tr>
<td>Arlettaz et al (2006) c</td>
<td>Foot only</td>
<td>Included</td>
<td>&lt;24 h</td>
<td>3262</td>
<td>12</td>
<td>12</td>
<td>0</td>
<td>3238</td>
<td>100.0% (73.5–100.0)</td>
<td>99.6% (94.4–99.8)</td>
<td>250.1% (142.3–439.5)</td>
<td>0.04% (0.01–0.59)</td>
</tr>
<tr>
<td>Sendelbach et al (2008) d</td>
<td>Foot only</td>
<td>Excluded</td>
<td>&lt;24 h</td>
<td>15233</td>
<td>1</td>
<td>24</td>
<td>0</td>
<td>15208</td>
<td>100.0% (2.5–100.0)</td>
<td>99.8% (99.8–99.9)</td>
<td>466.3% (191.0–1138.5)</td>
<td>0.25% (0.02–2.8)</td>
</tr>
<tr>
<td>Reich et al (2003) e</td>
<td>Foot and right hand</td>
<td>Excluded</td>
<td>&gt;24 h†</td>
<td>2114</td>
<td>0</td>
<td>4</td>
<td>0</td>
<td>2110</td>
<td>99.8% (99.5–99.9)</td>
<td>99.8% (99.8–99.9)</td>
<td>383.4% (268.8–546.9)</td>
<td>0.35% (0.21–0.57)</td>
</tr>
<tr>
<td>Koppel et al (2003) f</td>
<td>Foot only</td>
<td>Excluded</td>
<td>&gt;24 h</td>
<td>11281</td>
<td>3</td>
<td>1</td>
<td>2</td>
<td>11275</td>
<td>69.0% (14.7–94.7)</td>
<td>100.0% (100.0–100.0)</td>
<td>675.6% (839.8–54506.3)</td>
<td>0.40% (0.14–1.17)</td>
</tr>
<tr>
<td>Rosati et al (2005) g</td>
<td>Foot only</td>
<td>Excluded</td>
<td>&gt;24 h</td>
<td>5292</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>5288</td>
<td>66.7% (9.4–99.2)</td>
<td>100.0% (99.9–100.0)</td>
<td>3526.0% (424.6–29282.9)</td>
<td>0.33% (0.07–1.70)</td>
</tr>
<tr>
<td>Richmond et al (2002) h</td>
<td>Foot only</td>
<td>Included</td>
<td>&lt;24 h</td>
<td>5626</td>
<td>8</td>
<td>56</td>
<td>1</td>
<td>5561</td>
<td>88.8% (51.8–99.7)</td>
<td>99.0% (98.7–99.2)</td>
<td>89.2% (62.9–126.3)</td>
<td>0.11% (0.02–0.71)</td>
</tr>
<tr>
<td>de Wahl Granelli (2009) i</td>
<td>Foot and right hand</td>
<td>Excluded</td>
<td>&gt;24 h†</td>
<td>39821</td>
<td>19</td>
<td>68</td>
<td>10</td>
<td>39724</td>
<td>65.5% (45.7–82.1)</td>
<td>99.8% (99.8–99.9)</td>
<td>383.4% (268.8–546.9)</td>
<td>0.35% (0.21–0.57)</td>
</tr>
<tr>
<td>Riede (2010) j</td>
<td>Foot only</td>
<td>Excluded</td>
<td>≥24 h</td>
<td>41442</td>
<td>14</td>
<td>40</td>
<td>4</td>
<td>41384</td>
<td>77.8% (52.4–93.6)</td>
<td>99.9% (99.9–99.9)</td>
<td>805.5% (542.0–1197.0)</td>
<td>0.22% (0.09–0.53)</td>
</tr>
<tr>
<td>Fwer et al (2011) k</td>
<td>Foot and right hand</td>
<td>Included</td>
<td>&lt;24 h</td>
<td>20055</td>
<td>18</td>
<td>177</td>
<td>6</td>
<td>19854</td>
<td>75.0% (53.3–90.2)</td>
<td>99.1% (99.0–99.2)</td>
<td>84.9% (64.6–111.6)</td>
<td>0.25% (0.13–0.50)</td>
</tr>
<tr>
<td>Kawalec et al (2006) l</td>
<td>Foot only</td>
<td>Excluded</td>
<td>≥24 h</td>
<td>27200</td>
<td>7</td>
<td>13</td>
<td>1</td>
<td>27179</td>
<td>87.5% (47.3–99.7)</td>
<td>100.0% (99.9–100.0)</td>
<td>1830.2% (1001.3–3345.9)</td>
<td>0.13% (0.02–0.78)</td>
</tr>
<tr>
<td>Hoke et al (2002) m</td>
<td>Foot and right hand</td>
<td>Included</td>
<td>&lt;24 h</td>
<td>2876</td>
<td>4</td>
<td>53</td>
<td>0</td>
<td>2819</td>
<td>100.0% (39.8–100.0)</td>
<td>98.7% (97.6–98.6)</td>
<td>48.3% (32.6–71.7)</td>
<td>0.10% (0.01–1.40)</td>
</tr>
<tr>
<td>Summary estimate</td>
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<td></td>
<td>229421</td>
<td></td>
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</tbody>
</table>

CHD = congenital heart defect. *Studies by Hoke and colleagues and Reich and colleagues excluded from the analysis. †Mean age at testing >24 h after birth.

Table: Accuracy estimates of primary studies for pulse oximetry in the detection of critical congenital heart defects in newborn babies.
Timing and method of testing affects false positive rate

<table>
<thead>
<tr>
<th>Test Duration</th>
<th>Method</th>
<th>Sensitivity</th>
<th>Specificity</th>
<th>False pos rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Test &lt; 24 hrs</td>
<td>Foot only</td>
<td>88.9-100%</td>
<td>89.2-99.6</td>
<td>0.4-1%</td>
</tr>
<tr>
<td>Test &lt; 24 hrs</td>
<td>Hand &amp; foot</td>
<td>60-100%</td>
<td>99.4-100%</td>
<td>0-0.6%</td>
</tr>
<tr>
<td>Test &gt; 24 hrs</td>
<td>Foot only</td>
<td>77-100%</td>
<td>98.2-99.1</td>
<td>0.9-1.8%</td>
</tr>
<tr>
<td>Test &gt; 24 hrs</td>
<td>Hand &amp; foot</td>
<td>65.5-100%</td>
<td>99.8-100%</td>
<td><strong>0-0.2%</strong></td>
</tr>
</tbody>
</table>

Variability in whether prenatal dx was included.
Pulse oximetry as screening method

- Pulse oximetry measures the amount of $O_2$Hgb in the arterial blood
- Based on differential absorption of $O_2$Hgb and RHgb
- Coupled with ability to separate pulsatile from non-pulsatile components
- Non-invasive and painless
- Accurate with newer generation oximeters
- “Motion resistant” (SET) technology
- Fast (<2 min) and reliable
- Inexpensive (~ $4 per baby)
- Peripheral perfusion index (PPI)
Oxygen Sat is as simple as this
Pulse Oximetry

• Perform after 24hrs of age
• Infant should be awake
• Place Massimo probe on right hand followed by either foot. (performed either in parallel or in sequence)
• Obtain saturation and follow algorithm
• Remember to make sure you have a strong consistent pleth for accurate results
Pleth: the good, the bad, and the ugly

- Normal Signal
- Low Perfusion
- Noise Artifact
- Motion Artifact
Pulse Oximetry

Research has been conducted nationally and internationally to determine standards for best-practice for screening of critical CHD using pulse oximetry.

Research shows that the highest sensitivity (true positives) and highest specificity (true negatives) is associated with screening the right hand and one foot, using a cut-off of 95% or a 3% difference between the two.

Best outcomes are found when physical examination is paired with pulse oximetry screening.
Child in well-infant nursery 24-48 h of age or shortly before discharge if <24 h of age

Screen

- <90% in RH or F
- 90% - <95% in RH and F or >3% difference between RH and F
- ≥95% in RH or F and ≤3% difference between RH and F

Repeat screen in 1 h

- <90% in RH or F
- 90% - <95% in RH and F or >3% difference between RH and F
- ≥95% in RH or F and ≤3% difference between RH and F

Repeat screen in 1 h

- <90% in RH or F
- 90% - <95% in RH and F or >3% difference between RH and F
- ≥95% in RH or F and ≤3% difference between RH and F

Positive screen

Negative screen
MMC Algorithm
Educational Materials

Critical Congenital Heart Disease Screening Program

The Barbara Bush Children's Hospital
At Maine Medical Center
Educational Materials

Heart Smart Videos

• Available in Spanish, Simplified Chinese, Russian, French and Arabic
• Grant Funded from Baby's First Test

Video for Providers:
http://www.youtube.com/watch?v=Lif7kSgHfkw

Video for Families:
http://www.youtube.com/watch?v=o2CHeMRNdGg
Abnormal?

<table>
<thead>
<tr>
<th></th>
<th>NI</th>
<th>Abn</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>88%</td>
<td>89%</td>
</tr>
<tr>
<td>2</td>
<td>95%</td>
<td>91%</td>
</tr>
<tr>
<td>3</td>
<td>97%</td>
<td>93%</td>
</tr>
<tr>
<td>4</td>
<td>98%</td>
<td>95%</td>
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<tr>
<td>5</td>
<td>96%</td>
<td>94%</td>
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<td>Abnormal?</td>
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<td>Abn</td>
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</tr>
<tr>
<td>Right hand 88%; foot 89%?</td>
<td>NI</td>
<td>X</td>
</tr>
<tr>
<td>Right hand 95%; foot 91%?</td>
<td></td>
<td>X</td>
</tr>
<tr>
<td>Right hand 97%; foot 93%?</td>
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<td>X</td>
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<tr>
<td>Right hand 98%; foot 95%?</td>
<td></td>
<td>X</td>
</tr>
<tr>
<td>Right hand 96%; foot 94%?</td>
<td></td>
<td>X</td>
</tr>
</tbody>
</table>
Abnormal

- Right hand 88%; foot 89%?
- Right hand 95%; foot 91%?
- Right hand 97%; foot 93%?
- Right hand 98%; foot 95%?
- Right hand 96%; foot 94%?
Abnormal Result

- Oxygen Sat <90% no need to repeat
  - infant transferred to a NICU for evaluation
- O2 sat 90%-94% in both extremities: 3 measures 1 hour apart (algorithm) OR
- >3% difference in O2 sat between Rt hand and foot on 3 measures 1hr apart

Keep in mind- infant may NOT have heart disease!
<table>
<thead>
<tr>
<th>Abnormal?</th>
<th>NL</th>
<th>Abn</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right hand 88%; foot 89%?</td>
<td>X</td>
<td></td>
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<tr>
<td>Right hand 95%; foot 91%?</td>
<td>X</td>
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<tr>
<td>Right hand 97%; foot 93%?</td>
<td>X</td>
<td></td>
</tr>
<tr>
<td>Right hand 98%; foot 95%?</td>
<td>X</td>
<td></td>
</tr>
<tr>
<td>Right hand 96%; foot 94%?</td>
<td>X</td>
<td></td>
</tr>
</tbody>
</table>
Abnormal Result

• Oxygen Sat <90% no need to repeat
  – infant transferred to a NICU for evaluation

• O2 sat 90%-94% in both extremities: 3 measures 1 hour apart (algorithm)

• OR >3% difference in O2 sat between Rt hand and foot on 3 measures 1hr apart

Keep in mind- infant may NOT have heart disease!
Abnormal result

• Notify Primary Care Provider
• Physical exam
  – Auscultation of heart and lung sounds
  – 4 extremity blood pressures
  – Capillary refill
  – Femoral pulses
  – ? Chest X-Ray?
  – ? Continuous saturation monitoring?
  – ? Blood Gas?
Abnormal result, “What’s next?”

- Does the baby need to stay in NBN as opposed to Mom’s room?
- Will baby need an ECHO?
- Will the baby be transported or discharged?
- Who tells the parents?
Maine sites with Pediatric Cardiologists
HRSA Grant

• 3 years
  – June 1, 2012-May 31, 2015

• Total National funding
  – 2,100,000.00
  – 7 regional screening programs
    • New England Genetics Collaborative
    • ME, NH, VT, RI, CT
  – 300,000 per region

• Personnel-data entry, analysis, education

• Meetings-twice per year
HRSA Grant

• Goals and Objectives:
  – Develop a plan to incorporate CCHD screening methods at birthing facilities
  – Develop guidelines to collect and report results
  – Establish guidelines for screening follow-up and reporting
  – Develop education programs for providers and families
HRSA Grant

• Goals and Objectives (cont.):
  – Develop methods for quality assurance, outcomes analyses (including costs), and public health monitoring
  – Establish baseline data for each screening facility and develop ongoing data collection, analysis, and reporting methods
  – Establish state level health information exchange systems (requires legislation in Maine)
  – Prepare a project evaluation that includes next steps and identified best practices.
Legislation
LD 460
Maine Statistics Estimations

• Approximately 13,000 babies per year
• Incidence of 8:1000
• Anticipate 104 cases per year of CHD
• Of those- 25 cases per year of CCHD
  – Prenatal Diagnosis (~10-16)
  – Positive Screens (~ 13-20)
  – Undetected by Screening (~ 1-4)
• False Positives- (~25 babies per year)
Maine Implementation and Data

• Tertiary Care
  – EMMC (January 2012 implemented)
    • No CCHD diagnosed by pulse oximetry
      – 1 false negative (coarctation)
      – others ill or prenatally diagnosed
  – MMC (May 2012 implemented)
    • No CCHD diagnosed by pulse oximetry
      – ~1800 babies screened
      – Three abnormal screens
        » ASD, PFO, false positive
      – 2 parent refusals
Status of Implementation

• Already Implemented:
  – Farmington
  – Waldo County General
  – Bridgton
  – Mercy
  – Calais
  – Cary
  – CMMC
  – Down East Community
  – Goodall
  – Houlton
  – Maine General-both campuses
  – Penobscot Bay
  – SMMC
  – St. Mary’s
  – The Aroostook Medical Center
  – York

• In Process:
  – Mayo Regional
  – Mid Coast
  – Miles
  – Mount Desert
  – Northern MMC
  – Penobscot Valley
  – Redington Fairview
  – Rumford
  – Stephens Memorial

Approximately 2/3 of babies born in Maine are currently screened by pulse oximetry
Screening Stories from the Field

- Centers screening
  - both arms and neither leg
  - Q 15 minutes after birth X 1 hour and then one arm at 24 hours
  - results with greater than 3% difference evaluated by pediatric provider but not referred to cardiology
  - after Prenatal diagnosis of CHD already made
Points to leave with

• Newborn saturation screening is happening in ME; most centers following the AAP guideline
• Clinical examination alone misses critical CHD
• Oximetry is stable and reliable
• False positive rates are low (lower than false positive rate based on physical exam)
• Data to support this is strong
• Some lesions still will not be detected
If you remember nothing else...

• Right hand and either foot, over 24 hours
• Know the cutoffs (< 90, or ≥ 95%)
• Pay attention to the 3% difference
• Remember your exam is still important
  – Auscultation; femoral pulses (4 ext if needed)
  – CXR
  – Keep in mind, “it might not be the heart”
  – EKG- likely not necessary unless requested by pediatric cardiology
Selected References

• www.childrensnational.org/pulseox

• Strategies for Implementing Screening for Critical Congenital Heart Disease, Kemper, MD et al. Pediatrics 2011 Nov;128(5): e1-9

• Pulse oximetry screening for critical congenital heart defects in asymptomatic newborn babies. Lancet 2012. Thangaratinam et al.

• Endorsement of Health and Human Services Recommendation for Pulse Oximetry Screening for Critical Congenital Heart Disease. AAP Section on cardiology and cardiac surgery executive committee, January 2012.