Critical Congenital Heart Disease Screening: Concerns, Challenges, and Opportunities from the Clinical Perspective

Matt Oster, MD, MPH
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Concerns

• Do we really need this?

• How are we going to pay for this?

• Will this overwhelm the system?
Do we really need this?

Ailes et al. 2015
Do we really need this?

Ailes et al. 2015
How are we going to pay for this?

• Screening bundled as part of newborn care

• Further testing paid same as would be for symptomatic child
Will this overwhelm the system?

• Delayed discharges?
  – Rare
  – Parents not upset

• Excessive burden on pediatric cardiologists?
  – Absolutely not

• Unnecessary transports?
  – Exceedingly rare, and what’s “unnecessary”? 
Challenges

• What does a negative result mean?

• Why are we still missing some cases?

• How do we adapt to special settings?
What does a negative result mean?

Misinterpretation of Negative Pulse Oximetry Screening as Absence of Critical Congenital Heart Disease

“We…urge the (American) Academy (of Pediatrics) to mandate that nurseries document the cardiac conditions specifically ruled out by virtue of a negative screen on every discharge summary”
What does this mean for clinical care? Until there is a screening test for CCHD that has close to 100% sensitivity, we believe that pulse oximetry screening should be used as one additional tool to detect CCHD, but it should not preclude routine clinical examinations, nor should it be used to rule out heart disease, including any type of CCHD.
Why are we still missing some cases?

• Low sensitivity compared to other screening tests
  – Overall 50-75% (depending on definitions used for CCHD)
  – >85% if you add in clinical examination

• Determinants of hypoxemia
  – Timing of test
  – Flow across PDA
  – Severity of disease
Why are we still missing some cases?

- Timing
- Equipment

- Algorithm interpretation

- Echocardiography
How do we adapt to special settings?

- Altitude

Leuth et al. 2016.
How do we adapt to special settings?

- Out-of-hospital births

SpO$_2$ Measurement ≥ 1 hour after birth at right hand (RH) and either foot (F)

- RH or F <90%
- RH and F 90%-94% OR ΔRH-F >3%
- RH or F ≥95% AND ΔRH-F ≤3%

Repeat measurement after 1 hour

- RH and F <95% OR ΔRH-F >3%
- RH or F ≥95% AND ΔRH-F ≤3%

Repeat measurement at day 2 or 3 of infant’s life

- RH and F <95% OR ΔRH-F >3%
- RH or F ≥95% AND ΔRH-F ≤3%

Positive Screening

Narrayen et al. 2016.
How do we adapt to special settings?

- **NICU**

![Flowchart diagram](image_url)

Hu et al. 2016.
How do we adapt to special settings?

- NICU

STAGE 1: Screen ALL infants 24-48 hours of age including those on supplemental oxygen
Perform and document pulse oximetry in both RIGHT HAND and either FOOT using State/Hospital Specific Protocol

If on supplemental oxygen, implement state/hospital specific protocol with following modifications

First set of screening measurements is < 95% and consistent with clinical profile AND difference is 3 or less - DO NOT RESCREEN

First set of screening measurements is a difference of 4% or greater - RESCREEN 1 hour apart up to 2 times

If difference 3% or less - PASS
If difference 4% or greater after 3 attempts - FAIL

STAGE 2: Screen 24-48 hours after weaning to room air.

Pre-discharge screen
Performed per hospital protocol or if Stage 1 or Stage 2 screening was not done.

Opportunities

• What algorithm to use?

• What do we do with “false” positives?

• Is there something better than oxygen saturation level?
## What algorithm to use?

<table>
<thead>
<tr>
<th>Algorithm Source</th>
<th>Cutoff for Passing With First Measurement</th>
<th>Retest Criteria for Subsequent Measurements</th>
<th>Fail Criteria</th>
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</thead>
<tbody>
<tr>
<td>AAP</td>
<td>$O_2$ sat $\geq$ 95% (in either RH or F) AND [hand-foot] $O_2$ sat $\leq$ 3%</td>
<td>$O_2$ sat $&lt;$ 95% (in both RH and F) OR [hand-foot] $O_2$ sat $&gt;$ 3%</td>
<td>$O_2$ sat $&lt;$ 90% (either RH or F) OR fail retest criteria $\times$ 3</td>
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<td>New Jersey</td>
<td>$O_2$ sat $\geq$ 95% (in both RH and F) AND [hand-foot] $O_2$ sat $\leq$ 3%</td>
<td>$O_2$ sat $&lt;$ 95% (in either RH or F) OR [hand-foot] $O_2$ sat $&gt;$ 3%</td>
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<td>Tennessee</td>
<td>$O_2$ sat $\geq$ 97% (F)</td>
<td>$O_2$ sat $&lt;$ 95% (in both RH and F) OR [hand-foot] $O_2$ sat $&gt;$ 3%</td>
<td>$O_2$ sat $&lt;$ 90% (either RH or F) OR fail retest criteria $\times$ 3</td>
</tr>
</tbody>
</table>

F: either foot; $O_2$: oxygen; RH: right hand; sat: saturation.

Oster et al. 2016.
### Which algorithm to use?

<table>
<thead>
<tr>
<th></th>
<th>AAP</th>
<th>AAP Modified</th>
<th>NJ</th>
<th>NJ Modified</th>
<th>TN</th>
<th>TN Modified</th>
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</table>

Oster et al. 2015 (abs).
Which algorithm to use?

Diller et al. 2017 (abs).
What do we do with “false positives”?

- Up to 70% of “false positive” cases may have some other explanation of hypoxia:
  - Pneumonia
  - PPHN
  - Pneumothorax
  - Sepsis
  - Meconium aspiration
  - TTN requiring oxygen

Singh et al. 2013.
What do we do with “false positives”? 

• “Additional evaluation and testing of the infant should be prioritized according to the conditions most relevant for each case, and such evaluation should not be delayed while awaiting an echocardiogram. …The child should not be discharged without resolving the cause of desaturation or at least before excluding potentially life-threatening conditions. If a cause other than CCHD is identified and appropriately treated with resolution of hypoxemia, an echocardiogram might not be necessary.”

Oster et al. 2016.
Is there something better than oxygen saturation level?

de-Wahl Granelli et al. 2007.
Conclusions

• Initial concerns have been allayed

• There are still some challenges to fully implementing CCHD screening

• Opportunities exist to improve CCHD screening further
Thank you

Keep Calm and Put That Pulse-Ox On

MendedLittleHearts.org