CCHD Pulse Oximetry Newborn Screening: State Program Perspectives

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A picture may be worth a thousand words...

... but with CCHD...

...the devil is in the details.
• Utilizes pulse oximetry to detect lower oxygen saturations often associated with ductal-dependent Critical Congenital Heart Disease (CCHD)
  • Critical = surgery or catheter intervention in first year of life

• The screen detects HYPOXEMIA
  • Associated with non-critical CHD
  • Associated with Pulmonary Conditions
    • Pneumonia
    • Persistent Pulmonary Hypertension
  • Associated with Bacterial Infections
    • Sepsis
  • Associated with CCHD
    • Originally 7 primary targets – now 12
PRIMARY TARGETS OF CCHD PULSE OXIMETRY SCREENING

• Hypoplastic Left Heart Syndrome (HLHS)
• Pulmonary Atresia
• Tetralogy of Fallot
• Total Anomalous Pulmonary Venous Return
• Transposition of the Great Arteries
• Tricuspid Atresia
• Truncus Arteriosus
• Coarctation of the Aorta
• Double-Outlet Right Ventricle
• Ebstein’s Anomaly
• Interrupted Aortic Arch
• Single Ventricle
CCHD PULSE OXIMETRY SCREENING IS....

- One of the least uniform of the conditions on the RUSP
  - States utilize various:
    - Screening Algorithms
    - Follow-up practices
    - Data collection requirements and analysis

<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>
</tr>
</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 x 3</td>
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<tr>
<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>
<td>$O_2$ sat $&lt;90%$ (either RH or F) OR fail retest criteria x 3</td>
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<tr>
<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 x 3</td>
</tr>
</tbody>
</table>

$F$, either foot; $O_2$, oxygen; RH, right hand; sat, saturation.
CCHD PULSE OXIMETRY SCREENING IS...

• Unique to all other NBS conditions
  • Pulse Oximetry Screening is the third line of defense
    • And the first two lines are getting better (though unlikely to ever be 100%)

• Other Public Health Programs are involved (e.g., Birth Defects Registries)
  • In most states, identified cases of primary CCHD targets are being reported

• Necessity of the screen itself varies by individual and location
  • Dependent upon prenatal and clinical care availability and accessibility
• Infants who may have otherwise gone home undetected have been picked up by screening
  • Many, if not most eligible infants appear to be getting screened

• Significant other diseases like PPHN and pneumonia are being detected

• The addition of CCHD screening has not appeared to “shock” the system as some had feared (anecdotal)

• Addition of CCHD has resulted in stronger relationships with other Public Health Programs like Birth Defects Registries
EXISTING CHALLENGES IN CCHD PULSE OXIMETRY SCREENING

• Data Collection: Buy-In; Timeliness; Quality; Border Babies
  • Initial screening results
  • Echocardiogram results
  • Non-cardiac findings
  • Reasons for not screening

• Uniform case definitions still being developed

• Education regarding limitations of CCHD screening

• Screening devices
  • Concerns over accuracy and precision of currently available screening devices
EXISTING CHALLENGES IN CCHD PULSE OXIMETRY SCREENING

• **Unknown best practices/algorithm**
  • Also unknown, in many cases, if algorithm is being followed correctly
  • In 2016, Minnesota had a misinterpreted algorithm in 0.6% of cases (despite building algorithm into software)

• **Infants in the NICU**

• **Out-of-Hospital Births**
  • How to incorporate algorithm into existing workflows

• **Facility versus Program roles/responsibilities**
  • Individual level QI/QA
  • System level QI/QA
  • Varying from traditional roles and responsibilities
• Support for robust data collection and analysis
  • Resources for follow-up (inclusive of long term follow-up) and quality assurance
  • Will allow for better evidence-based recommendation to improve upon current implementation efforts

• A fresh perspective
  • CCHD screening does not appear to fit into typical NBS paradigm
    • Metrics and expectations need to be different
CONCLUSION

- CCHD screening has value – just not yet quantifiable
  - Overall mortality from CCHDs appears to be going down

The question remains – as newborn screening programs – how do we best approach this screening program with the ultimate end goal of improving outcomes in mind?
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Thank you

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