Catching Blue Babies: Critical Congenital Heart Defects in Newborns at Moderate Altitude

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Pediatric Heart Defects

• Critical Congenital Heart Defects (CCHD)
  – 1 in every 4 newborns with a heart defect
  – 7,200 newborns in the US every year
  – Cardiac procedure within 1\textsuperscript{st} year of life
Newborn Screening for CCHD

• CCHD symptoms:
  – Low oxygen saturations → cyanosis
  – Blue baby syndrome

• Screening using pulse oximetry technique (!!!)

Colorado School of Public Health
CCHD Screening at Altitude

• Partial pressure of $O_2$ decreases with increasing altitude $\rightarrow$ physiological issue

• Screening algorithm based on studies at sea level $\rightarrow$ efficacy issue

• Limited information on newborns with CCHD at altitude and their screening results

• Secretary HHS requested additional studies
Research Questions

• What are pulse oximetry results of newborns with CCHD born at altitude?
  – Delineate pulse oximetry values

• How did they respond to screening algorithm?
  – Performance of AAP algorithm at altitude
Methodology

- Retrospective cohort of *true positives*
  - Using data from medical charts to recreate screening scenario
- Term/near-term newborn, 7 dx of CCHD, treated at Children’s, 2003 – 2013
  - i2b2 → 343 cases
  - No electronic chart, > 48-hr admission, < 24-hr deceased or had cardiac surgery
  - Descriptive statistics → 158 cases
  - Demography, risk factors, O₂ saturations at 24-, 28-, 36-, 48-hr, screening results
Demography

- Male → 64%
- White/Caucasian → 69%
- Non-Hispanic/Latino → 64%

- KS, MT, NE, NM, TX, WY
  - Transferred to Children’s for higher care within 24-hr after birth

78% from CO
Risk Factors and Diagnosis

• Family history of CCHD, genetic disorder, and maternal diabetic status are **not** risk factors in our cases

• Top 3 diagnosis:
  - Hypoplastic Left Heart Syndrome (70%);
  - Transposition of Great Arteries (30%);
  - Tetralogy of Fallot (18%)

• Children’s Hospital CO treated ~30 cases/yr
## Comparative $O_2$ Saturation Values

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Altitude</th>
<th>Statistics</th>
<th>Preductal (%)</th>
<th>Postductal (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>CCHD</td>
<td>Moderate</td>
<td>Average</td>
<td>87.1 ± 7.2</td>
<td>87.8 ± 6.3</td>
</tr>
<tr>
<td>CCHD</td>
<td>Moderate</td>
<td>Range</td>
<td>70.0 – 100.0</td>
<td>67.0 – 100.0</td>
</tr>
<tr>
<td>Healthy</td>
<td>Moderate</td>
<td>Average</td>
<td>97.2 ± 1.9</td>
<td>97.2 ± 2.1</td>
</tr>
<tr>
<td>Healthy</td>
<td>Moderate</td>
<td>Range</td>
<td>88.0 – 100.0</td>
<td>88.0 – 100.0</td>
</tr>
<tr>
<td>Healthy</td>
<td>Sea Level</td>
<td>Average</td>
<td>98.3 ± 1.4</td>
<td>98.9 ± 1.6</td>
</tr>
<tr>
<td>Healthy</td>
<td>Sea Level</td>
<td>Range</td>
<td>94.7 – 100.0</td>
<td>94.7 – 100.0</td>
</tr>
</tbody>
</table>

(Samuel et al., 2013, Acta Paediatrica)  
(Wright et al., 2014, Pediatrics)
Screening the Positives

1st Screen

- Passed Screen (no concern for CCHD)
- Re-screened (at next time point)
- Failed Screen (concern for CCHD)
• 29 out of 149 false negatives (19.5%)
  – 80.5% screen capture rate
  – Comparable to national data (76.5%)

(Thangaratinam et al., 2012, Lancet)
CCHD Treatments

• Prostaglandin (PGE) & O₂ supplementation
  – 72% given PGE; 54% given O₂ (158 cases)
  – Comparable to false negatives (29 cases)

• Failed screening even after treatments:
  – Newborns at altitude have more severe defects
  – Current screening algorithm is sufficiently applicable and effective at altitude
In Summary

• First comprehensive review of newborns already diagnosed CCHD
  – Novel approach
  – Delineate average and range of O₂ saturations
  – Evaluate performance of current screening algorithm performed at altitude
Implication & Future Direction

• 80.5% capture rate of current screening algorithm performed at altitude
  – Helpful in moving forward with CCHD screening efforts in CO

• Combine with data of true negatives to get a complete assessment of the performance of screening algorithm at altitude
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