Opinion
The Simple Test That Saved My Baby
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"ON July 10, my wife gave birth to a seemingly healthy baby boy with slate-blue eyes and peach-fuzz hair. The pregnancy was without complications. The delivery itself lasted all of 12 minutes. After a couple of days at Greenwich Hospital in Connecticut, we were packing up when a pediatric cardiologist came into the room.

We would not be going home, she told us. Our son had a narrowing of the aorta and would have to be transferred to the neonatal intensive care unit at NewYork-Presbyterian Hospital at Columbia, where he would need heart surgery."

Objectives
- Explain the rationale for screening for Critical Congenital Heart Disease (CCHD) in newborns
- Examine the evidence supporting the routine use of pulse oximetry in the Newborn Nursery to detect CCHD
- Discuss evidence-based recommendations for implementation of CCHD screening

Outline
- What is "critical" congenital heart disease?
- Why do we need to screen?
- How do we screen for critical CHD?
- Current status of screening
  - National
  - Local
  - TxPOP data

Congenital Heart Disease
- Incidence: 8-9/1000 births
- 2/1000 potentially lethal - "critical"
  - Requiring expert cardiac care and intervention in the immediate NB period or early infancy.
- In the US, about 4800 babies are born each year with CRITICAL CHD
- Leading cause of death in infants < 1 year old
Congenital Heart Disease
- Advances in surgical and interventional cardiology has improved survival over the past 30 years.
- There are an estimated 800,000 adults living with CHD.
- Survivors who present late are at greater risk for neurologic injury and subsequent development delay.
- Focus now has shifted from increasing survival to reducing morbidity.

Critical Congenital Heart Disease
- Those CHD’s that will require cardiac intervention in the newborn period or within the first year of life.
  - Ductal dependent systemic circulation
    - HLHS, Coarctation, IAA, Critical AS
  - Ductal dependent pulmonary circulation
    - PA, PS and variants, TOF
  - Complex critical CHD
    - TGA, Truncus Arteriosus, TAPVR, Single ventricle

Can we screen for CCHD?
- Screening valuable if:
  - Incidence is sufficient in the population
  - Therapy provided before onset of clinical manifestations results in an improved outcome
  - Screening identifies disease before symptoms
  - Test has acceptable sensitivity and false positive rates
  - Cost effective
    - Wilson and Junger WHO 1968 Public Health Paper

Diagnosis vs. Screening

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<thead>
<tr>
<th></th>
<th>Diagnostic</th>
<th>Screening</th>
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<tbody>
<tr>
<td>Pros</td>
<td>Fewer resources needed</td>
<td>Higher detection rate</td>
</tr>
<tr>
<td></td>
<td>Identification may be too late</td>
<td>High resource use</td>
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<tr>
<td>Cons</td>
<td>Application may be spotty</td>
<td>Adverse impact of false positives</td>
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CCHD detection – diagnostic
- Fetal echocardiography
  - >50% detection rates for single ventricle lesions
  - <30% for 2-ventricle
    - Highly variable, limited access
- Newborn physical exam
  - in nursery and in clinic
  - 4-5 grams of deoxygenated Hgb is needed to detect cyanosis
  - Most CCHD have mild desaturation to 80-95%
  - Harder in darker skinned babies
Diagnostic Process

Newborn presents in shock with murmur → Exam suggestive of CHD → Hypoplastic Left Heart

Missed Diagnosis

- Some babies can appear healthy at first
  - Some have no murmurs or cyanosis

- PE alone failed to identify 50% of CHD’s that were not detected by prenatal U/S

- Estimated 30% of infant deaths from CCHD occur before diagnosis

Missed Diagnosis of CCHD

- Up to 30 infants die annually in CA of missed or late diagnosis of CCHD at median age of 13.5 days.

CCHD Screening

- Pulse Oximetry
  - Indirectly monitors the oxygen saturation of a patient’s blood and changes in blood flow in the skin
  - Can detect mild hypoxemia without obvious cyanosis
  - Can provide continuous and immediate values
  - Non-invasive
  - Easy to use and widely available
  - Cost-effective and widely used

Pulse Oximetry Screening - Evidence

- Using a cut-off of 95% in the LE, Hoke et al identified 81% of infants with CCHD

- Many investigators have since investigated the use of pulse oximetry as a screening tool in newborns NOT known to have CHD
  - Most studies were small, with different protocols and cut-offs, at low altitude
  - Low false positive rate < 1%, sensitivity < 80%
    - Likely because hypoxemia is not present in all CCHD

Hoke et al, Oxygen saturation as a screening test for critical CHD. Ped Cardiol.2002;23:203-409
Pulse Oximetry Screening Program Saxony, Germany

Riede et al Eur J Pediatr 2009

Pulse Oximetry Screening - Evidence

- 2 separate large prospective screening of 40,000 newborns in Sweden and nearly 40,000 in Germany.
  - Sensitivity 62%, Specificity 99.8%
- A meta-analysis of pulse ox screening for CCHD in asymptomatic newborns
  - Over 220,000 NB’s
  - Overall sensitivity was 76.5%, specificity was 99.9% with a false positive rate of 0.14%


Cost of Routine Pulse Oximetry

- Includes both the direct cost of the pulse oximetry and the follow-up costs of any additional examinations and transfers.
  - At experienced centers, it took technicians only 2 minutes on average to perform screen.
  - Calculation of time in New Jersey 9 min per child
    - No new nursing or medical technician FTEs added
- 0.13%
- Cost of approximately $3-6 per asymptomatic newborn
  - Assumes reusable probe

Current Status of Recommendations

- US Health and Human Services Secretary’s Advisory Committee on Heritable Disorders in Newborns and Children (HHS-SACHDNC)
  - In 2010, recommended that CCHD be added to the newborn uniform screening panel
  - Identify newborn with structural heart defects associated with hypoxia that could have significant morbidity or mortality early in life with closing of the patent ductus arteriosus or other physiologic changes
  - 2011, Endorsed by Secretary of Health Kathleen Sibelius

Taryn’s story

- http://youtu.be/2lM8hFHUMl4

National Efforts

- Maryland first state to pass CCHD screening legislation.
- New Jersey first state to mandate universal CCHD screening- Implemented August 31, 2011.
- Other states have legislation passed, introduced or pending
  - Multi-center screening/pilots
  - HRSA sponsored demonstration projects
  - Opportunity for other states to learn and not have to “re-invent” the wheel
Texas
- HB 740 passed the Senate on 5/13/2013 and was signed into law – effective September 1, 2013.
  - Taryn Kennedy, Nash Sievers & Rex Van de Putte act. Taryn & Nash are two babies who died from CCHD and their moms worked on the legislation. Rex is the grandson of Sen. Leticia Van de Putte who passed away at 6 months of SIDS.
  - Requires all newborns of a birthing facility be screened.
Exceptions – parental refusal, transfer prior to screening, screening had previously been completed, discharge before 10 hours and referral made.

Potential Barriers
- Reporting/Tracking/ QI
- Inadequate resources
- Resistance from some in the medical community
- Screener
  - Additional work load
  - Education
- Equipment
  - Probe, machine
- Patient/Parent
  - False positives, false negatives
  - Delay in discharge
- Potential transfer to another center
- Costs and reimbursement

AAP/CDC Algorithm

CCHD Screening Protocol
- 7 primary targets
  - Hypoplastic Left Heart Syndrome
  - Pulmonary Atresia (with intact atrial septum)
  - Tetralogy of Fallot
  - Total Anomalous Pulmonary Venous Return
  - Transposition of the Great Arteries
  - Tricuspid Atresia
  - Truncus arteriosus
- 17-31% of all CHD’s

CCHD Screening Protocol
- Secondary screening targets
  - Can be just as severe but not consistently detected
  - Aortic arch atresia/hypoplasia
  - Interrupted aortic arch
  - Coarctation
  - DORV
  - Ebstein’s anomaly
  - PS, PA, AVCD
  - Other Single ventricle defects
How to Perform Screening

- Screen after 24 hours of age
- Conduct when infant is calm and awake
- Perform in preductal (RIGHT hand) and postductal (one FOOT), in parallel or one after the other
- If < 90% - positive screen, refer
- If ≥ 95% in EITHER extremity with ≤ 3% difference: PASS
- If 90 - 94% in BOTH or difference > 3%: REPEAT in 1 hour up to 2 times, then refer

How is it done?

CCHD Screening Algorithm

Pulse ox on right hand and foot after 24 hours

- 95% in right hand (RH) or foot and < 3% difference between RH and foot
  - More
  - Pass

- 90-94% in RH and foot
  - Repeat in 1 hour
  - Notify MD/NNP

- < 90% in RH or foot
  - Positive (FAIL)

If > 3% difference between RH and foot

Evaluation for Positive Screen

- Clinical Assessment
- Infectious or Pulmonary pathology should be excluded
- Complete echocardiogram
- Pediatric Cardiology referral as indicated

Managing the Positive Screen

“In the absence of other findings to explain hypoxemia, CCHD needs to be excluded on the basis of a diagnostic echocardiogram (which would involve an echocardiogram within the hospital or birthing center or transport to another institution)....”

- Alternative strategies
  - Keep child until evaluation can be performed
  - Transfer to advanced nursery (without cardiac inpatient service)
  - Transfer to center with advanced cardiac care

Screening in the Real World

- Feasibility of implementing pulse oximetry screening for CHD in a community hospital
  - 6745 eligible infants screened at average age 42h
    - 9 positive – 1 had CCHD
- Barriers (1.4%):
  - screening equipment 54%
  - staff 23%
  - infant 20%
  - family 4%
- Physician and Nurse “champions” important to successful implementation
Texas Pulse Oximetry Project: A Joint Educational Initiative.

Goal: Develop an appropriate implementation strategy for screening of CCHD using pulse oximetry as a potential public health mandate.

- Develop and provide educational programs and materials
- Funding: Texas Department of State Health Services' Children's Outreach Heart Program

Devised and implemented Needs Assessment of clinical sites.

Developed an educational plan to include curriculum and educational materials.

Target: 13 facilities in South Texas and Southeast Texas representing an array of birthing facilities ranging from the rural hospital with limited resources to the large metropolitan medical centers with access to multiple resources.

Identified a nurse champion at each facility to champion CCHD screening.

Project Timeline

- Develop TxPOP proposal and project team
- Execute TxDSHS contracts
- Conduct needs assessments; lit review
- Design curriculum
- Plan/apply for CNE credits
- Produce educational materials
- Prepare and conduct Train the Trainer
- Implement trainings at hospitals
- Additional trainings at hospitals
- Collect and report data to TxPOP
- Aggregate data for 6 months; to TxDSHS
- Draft and circulate comprehensive report
- Finalize and submit report to TxDSHS

Quality Improvement – Feb-July

- 12,946 births in the 13 facilities
- 11,721 newborn nursery admissions
- 11,289 CCHD newborn screens
- 96% of babies admitted to the newborn nursery received a CCHD screen during the recommended time frame (between 24 hours and discharge).
- Babies not admitted to the newborn nursery after birth (approximately 1,235)
- Transfers out of newborn nursery prior to CCHD screen (249)
- Screens performed prior to 24 hours (38)

Nurse CNE trainings

<table>
<thead>
<tr>
<th>No nurses/hospitals</th>
<th>Pre-test</th>
<th>Post-test</th>
<th>Pretest algorithm questions N=2</th>
<th>Posttest algorithm questions CCHD N=2</th>
<th>Pretest CCHD</th>
<th>Posttest CCHD</th>
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<tbody>
<tr>
<td>South Texas</td>
<td>113/6</td>
<td>73%</td>
<td>73%</td>
<td>91%</td>
<td>*56%</td>
<td>*16%</td>
</tr>
<tr>
<td>Houston area</td>
<td>102/7</td>
<td>69%</td>
<td>94%</td>
<td></td>
<td>*70%</td>
<td>*16%</td>
</tr>
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*Percent incorrect

Positive Screens – 10 – all had ECHOs
False Positive rate of 0.079%

- 2 were in the <90% group – 1 had severe CCHD from secondary target; other had subclinical seizures
- 7 were the indeterminate of 90-95% X 3
- 1 had >3% difference

- 32 babies from the 13 facilities had ECHOs

Only 3 transfers from initial facility, all within same zipcode
TAPVR

Toolkit
• [http://txpeds.org/txpop](http://txpeds.org/txpop)

Continued issues
• How to ensure 100% of newborns are screened? – home deliveries excluded
• Who is responsible for reporting and follow-up, an important element in assuring that all babies are screened?
• What are the reimbursement mechanisms for screening which has its costs in nursing time, probes, and education materials for families?

Continued issues
• What are the reimbursement mechanisms for positive screens?
• How to track implementation compliance to ensure all babies are screened and treated appropriately?
• How to identify referral centers that will accept timely transfer throughout the state?
• How do we screen for babies admitted to NICUs?

Thank you!
References


7) Congenital heart disease (CHD) in the newborn: Presentation and screening for critical CHD. Carolyn A. Altman, MD; Wolters Kluwer Health, Official reprint from UpToDate; Literature review current through 2012
