Opinion
The Simple Test That Saved My Baby
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Published: September 22, 2013

"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 New York-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 have 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

Critical Congenital Heart Disease

- Physiologic changes may occur after hospital discharge corresponding to changes in the pulmonary vascular resistance and closure of the patent ductus arteriosus.
- Present in extremis with low cardiac output and acidosis, multi-organ failure, hypoxic ischemic brain injury.
- Early detection and timely intervention can thus decrease morbidity and lead to better outcomes.

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

<table>
<thead>
<tr>
<th>Diagostic Pros</th>
<th>Screening Cons</th>
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<tbody>
<tr>
<td>Fewer resources needed</td>
<td>High resource use</td>
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<tr>
<td>Higher detection rate</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.

Chain of Detection

CCHD → Prenatal US → Symptoms → CCHD → Physical Exam → CCHD

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 CCHD
  - 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


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
  - ?????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/2IM8hFHUMI4

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
29 states have passed CCHD legislation

Potential Barriers
- States have different processes
- 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?

95% in right hand (RH) or foot and < 3% difference between RH and foot
90-94% in RH and foot
< 90% in RH or foot

CCHD Screening Algorithm

Pititi (FAIL)
Pulse ox on right hand and foot after 24 hours
or Positive (FAIL)
Repeat in 1 hour
Notify MD/NNP

Posi

PASS
Indeterminate
Remind parents that CCHD newborn screening may not find all types of problems in a baby's heart.

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)...."
Kemper et al Pediatrics 2011

• 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
  • Bradshaw, J Perinat. 2012;1-6.
  • 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

TxPOP

• 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.

**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 N=2</th>
<th>Pretest CCHD</th>
<th>Posttest CCHD</th>
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<tbody>
<tr>
<td>South Texas</td>
<td>13/6</td>
<td>73%</td>
<td>*56%</td>
<td>*42%</td>
<td>*9%</td>
<td>*3%</td>
</tr>
<tr>
<td>Houston-area</td>
<td>10/2/7</td>
<td>69%</td>
<td>*70%</td>
<td>*36%</td>
<td>*2%</td>
<td>*2%</td>
</tr>
</tbody>
</table>

*Percent incorrect

**Quality Improvement – Feb-July**
- 12,946 births in the 13 facilities
- 11,711 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)

**Positive Screens – 10 – all had ECHOs**

- 2 were in the <90% group – 1 had severe CCHD from secondary target; other had subclinical seizures
- 7 were the indeterminates 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**
References


References


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
