Routine Screening for Critical Congenital Heart Disease – Pros and Cons

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Objectives
- Explain the rationale for screening of 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

Pros/Cons
- 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
  - Potential Barriers

Congenital Heart Disease
- Incidence: 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

- 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
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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, Interrupted Aortic Arch, 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

Screening- 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

CCHD detection

- Fetal echocardiography
  - >50% detection rates for single ventricle lesions
  - <30% for 2-ventricle
    - Highly variable, limited access
- Newborn physical exam (cyanosis)
  - 4-5 grams of deoxygenated Hgb is needed to detect cyanosis
  - Most CCHD have mild desaturation to 80-95%
  - Harder to detect in darker skinned babies

CCHD detection

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

CCHD detection

- PE, EKG, chest X-ray
  - Can help to identify CCHD's
  - Lack sensitivity and specificity to detect all CCHD's
- Echocardiogram
  - High cost
  - <15% of echos ordered by PMD for suspicion of CHD showed significant CHD
  - Lack of expert personnel
  - High number of non-critical lesions – PFO’s, PDA, small VSD’s
CCHD detection

- **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 in its current usage in ICU’s, ER’s, etc

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Pulse Oximetry Screening - Evidence FOR

- Using a cut-off of 95% in the LE, Hoke et al identified 8% of infants with CCHD
  - Hoke et al. Oxygen saturation as a screening tool for critical CHD. Ped Cardiol. 2002;23:203-209

- Several investigators have suggested using pulse oximetry as a screening tool to detect CCHD in newborns prior to discharge
  - Most studies were small, with different protocols and cut-offs
  - Low false positive rate < 1%, sensitivity 60-80%

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Pulse Oximetry Screening - Evidence AGAINST

- 15,000 newborns screened at 4 hours of life
  - 5.6% had foot pulse ox < 96%, of these 99.9% were normal at re-screen
  - 31 of all NB’s had CCHD (1.9/1000 livebirths)
  - 16 were directly admitted to NICU
  - 15 went to NB nursery first and were screened
    - 3 had pulse ox < 96%, but ALL developed SSx before d/c

  Sendelbach et al. Pulse oximetry Screening at 4 hours of age to detect CHD. Pediatrics 2008;122;285

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Pulse Oximetry Screening - Evidence

- Scientific Statement from the AHA/AAP in 2009
  - Comprehensive review of all available studies
  - Found compelling reasons for newborn screening
  - Class IIb, Level of Evidence C (observational studies)
  - Called for “collaborative studies in larger populations across a range of newborn nursery systems” before universal pulse ox screening is recommended


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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%


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Current Status of Recommendations

- US Health and Human Services Secretary’s Advisory Committee on Heritable Disorders in Newborns and Children (HHS-SACHDC)
  - 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
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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-funded demonstration projects
- Opportunity for other states to learn and not have to “re-invent” the wheel

Potential Barriers- Cons
- Each state has a different process
- Several programs who do not publish their experience
- Reporting/Tracking/ QI
- Inadequate resources
- Limited U.S. evidence-based research
- Resistance from some in the medical community

SACHDNC /AAP/ACCF/AHA-2011
- Health Resource Service Administration’s Advisory Council on Heritable Diseases in Newborns and Children hosted a workshop to discuss implementation recommendations surrounding screening
- Screening protocol based on the most current evidence available

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
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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

Screening Recommendations
- Screen after 24 hours of age when infant is calm and awake
- Perform preductal (RIGHT hand) and post ductal (one FOOT), in parallel or one after the other
  - If ≤90% in either extremity, positive screen - REFER
  - If ≥95% in BOTH extremities with ≤3% difference: PASS
  - If 90-94% in EITHER or difference > 3%; REPEAT in one hour up to two times; If positive screen, then REFER

Screening in the Real World
- Feasibility of implementing pulse oximetry screening for CHD in a community hospital
  - 6700 eligible infants screened at average age 42h
  - Barriers (1.4%) : screening equipment 54%, staff (23%), infant (20%)
  - 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

TxPOP
- Develop a Needs Assessment tool
- Develop an educational plan to include curriculum and educational materials
- Target: minimum of 14 facilities in South 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
- Identify a staff person to champion CCHD screening
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TxPOP
- First meeting Aug 24-25, 2012
- UTHSCSA: Alice Gong, MD
  - Judith Livingston, MEd, MCHES
  - Rachel Turner, RN, BSN
- BCM/TCH: Charleta Guillory, MD
  - Yvette Johnson, MD
  - Elena Ocamo, MD
  - Susanna Rubio, RN, BSN
  - Tiffany Mc Kee-Garrett, MD
- Texas DSHS: Debra Freedenberg, MD, PhD
- Texas Health Institute: Liza Covel, MPH
- Valley Baptist Medical Center-Brownsville: Maria Gutierrez, MSN,APRN,NNP-BC

References


References
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Thank you!