



Routine Screening for Critical Congenital Heart Disease in Healthy Newborns

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I have nothing to disclose.

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Objectives

- Rationale for Screening for Critical Congenital Heart Disease
- Discuss supporting evidence for the use of pulse oximetry to detect CCHD
- Discuss implementation of CCHD screening in Texas
 - Texas Pulse Oximetry Project

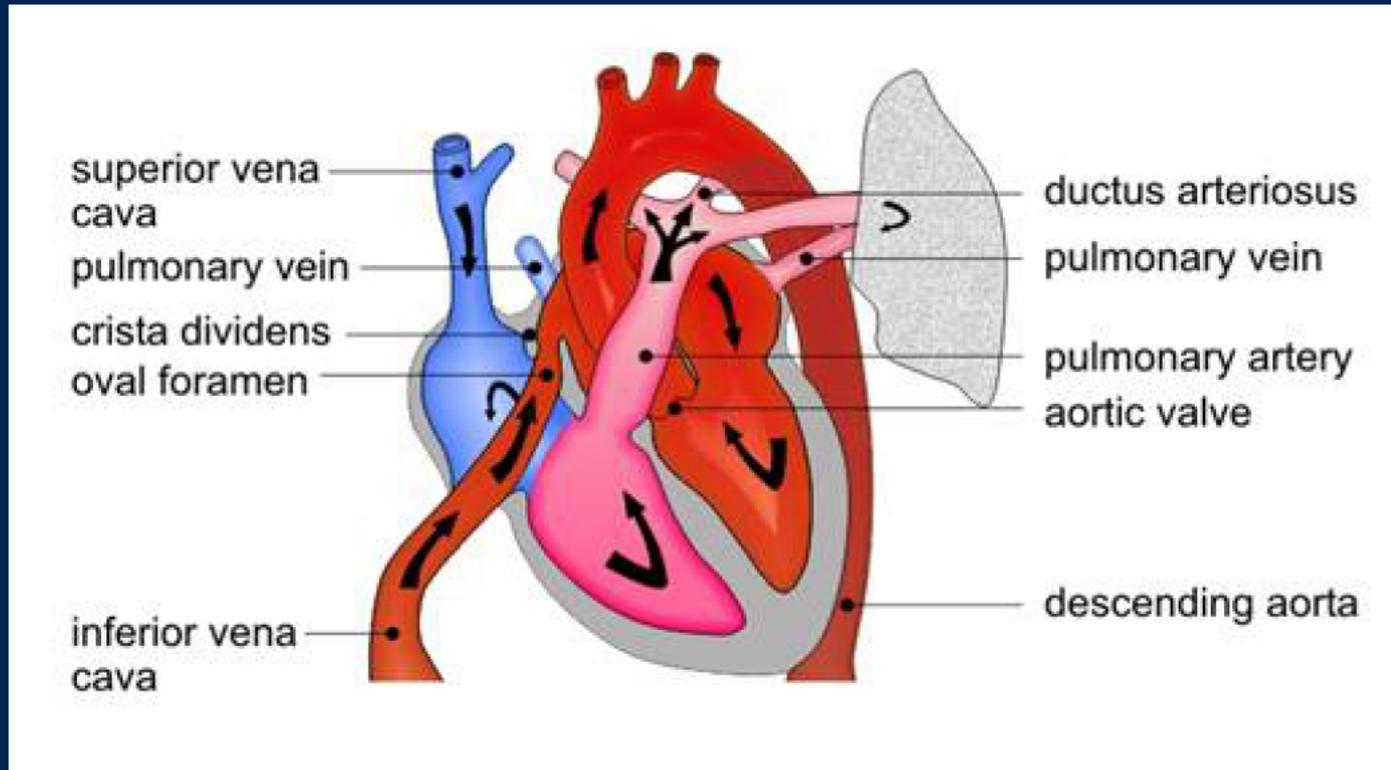
Congenital Heart Disease

- Incidence of CHD : 8-9/1000 births
- 2/1000 potentially lethal - “critical”
 - Requiring expert cardiac care and intervention in the immediate newborn period or early infancy
 - About 4800 babies are born each year with CRITICAL CHD
 - Leading cause of death in infants < 1 year old
 - About 300 infants/yr are discharged with unrecognized CCHD

Critical Congenital Heart Disease

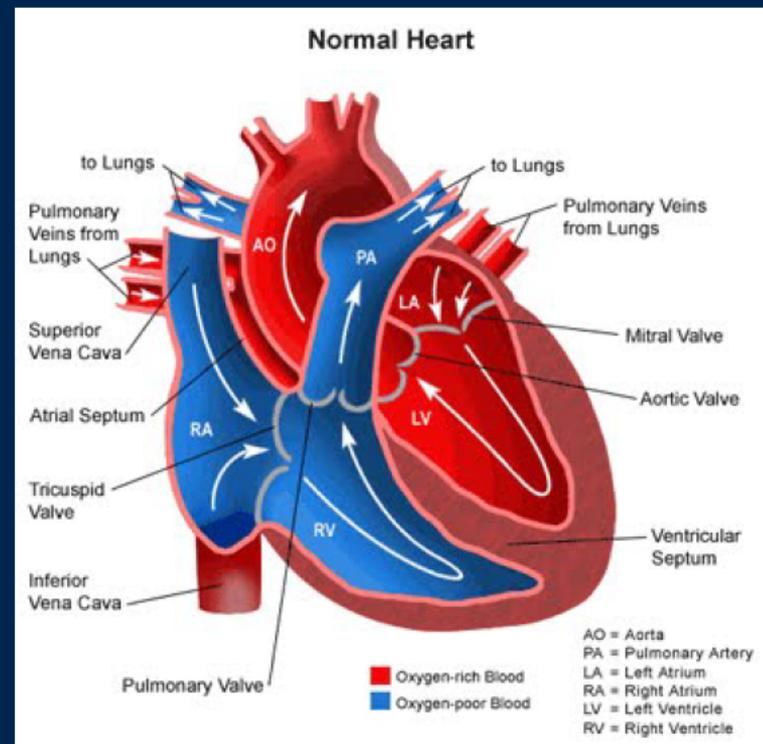
- Those CHD's that will require cardiac intervention in the newborn period or within the first year of life
 - Ductal dependent systemic circulation
 - HLHS, Coarctation, IAA, Critical AS
 - Ductal dependent pulmonary circulation
 - PA, PS and variants, TOF
 - Complex critical CHD
 - TGA, Truncus Arteriosus,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

The Fetal Circulation and Patent Ductus Arteriosus



What happens after birth?

- Pulmonary vascular resistance drops
- Foramen ovale closes
- Ductus arteriosus constricts



These changes may occur **AFTER** hospital discharge

Typical Scenario



Newborn presents in shock with murmur

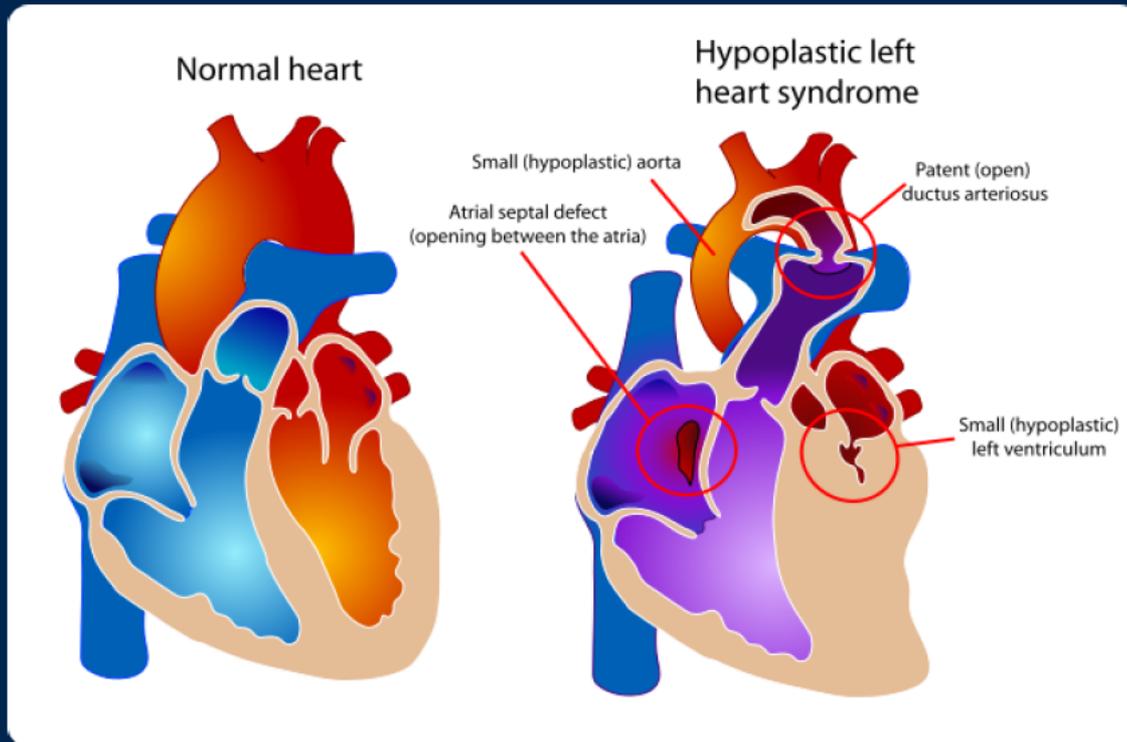


Exam suggestive of CHD



Hypoplastic Left Heart

HLHS- ductal dependent systemic circulation



Present in
extremis with
low cardiac
output and
acidosis, multi-
organ failure,
hypoxic
ischemic brain
injury

Missed Diagnosis of CCHD

- About 30 infants/yr died from late or missed CHD
 - More than 50% died at home or EC
- Median age 13.5 days
- HLHS, Coa

Table 2. Total Number of Patients in Each Group by Diagnosis

Cause of Death	No. (%) of Patients ^a		
	Study Cohort (N=898) ^b	Unknown (n=299)	Missed CCHD Diagnosis (n=152)
Aortic stenosis	31 (3.5)	19 (6.4)	12 (7.9)
Coarctation of aorta, including interrupted aortic arch	90 (10.0)	49 (16.4)	41 (27.0)
DORV and single ventricle	15 (1.7)	14 (4.7)	1 (0.7)
Hypoplastic left heart syndrome	565 (62.9)	60 (20.1)	58 (38.2)
Pulmonary atresia	30 (3.3)	22 (7.4)	8 (5.3)
Tricuspid atresia	9 (1.0)	9 (3.0)	0
TAPVR	32 (3.6)	23 (7.7)	9 (5.9)
d-Transposition of great vessels	37 (4.1)	31 (10.4)	6 (3.9)
Tetralogy of Fallot	55 (6.1)	50 (16.7)	5 (3.3)
Truncus arteriosus	34 (3.8)	22 (7.4)	12 (7.9)

Abbreviations: DORV, double outlet right ventricle; TAPVR, total anomalous pulmonary venous return.

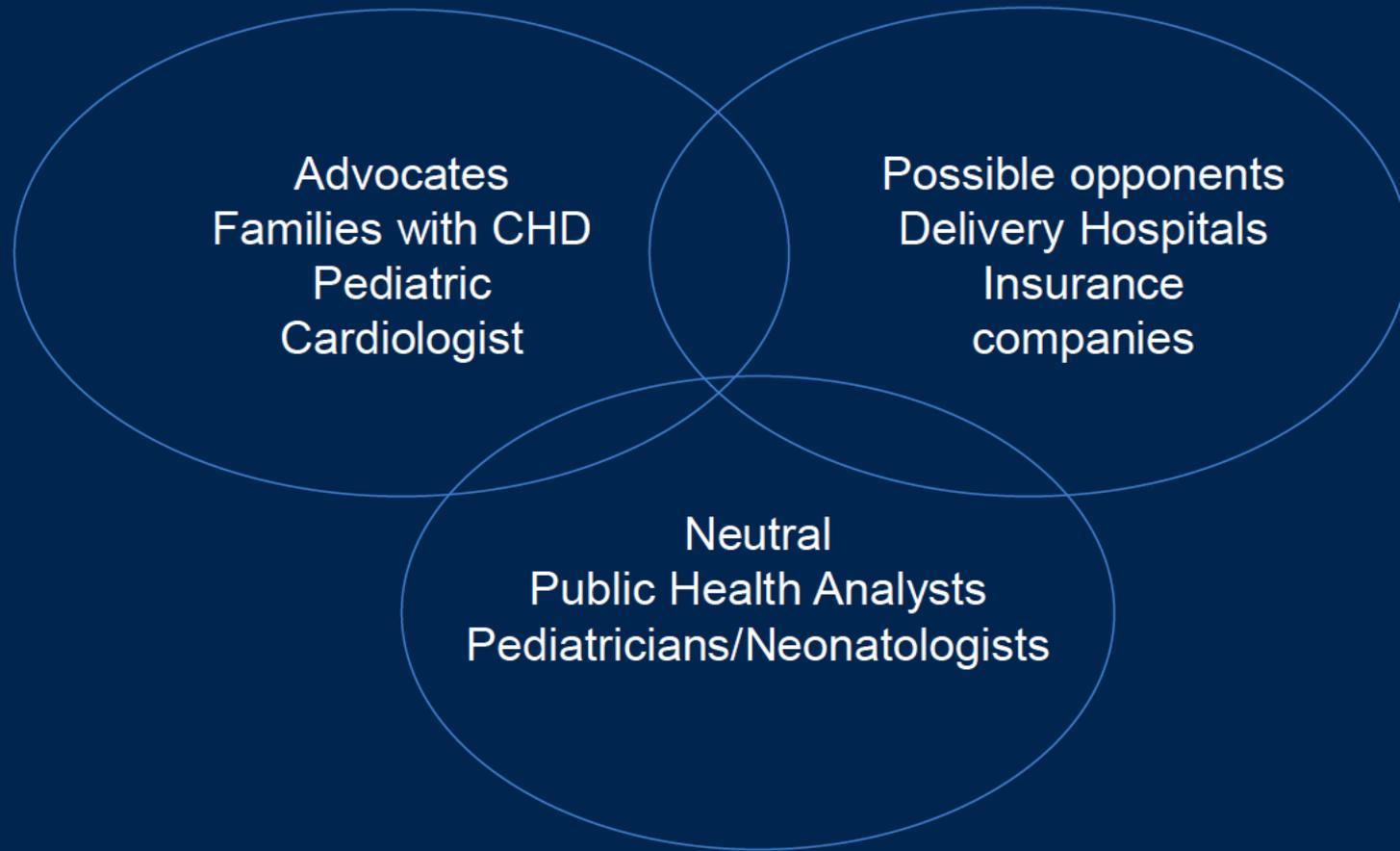
^aBecause of rounding, percentages may not total 100.

^bStudy cohort indicates the 898 patients selected by the initial selection criteria specified in the "Patient Selection" subsection of the "Methods" section.

Critical Congenital Heart Disease

- **Early detection and timely intervention can decrease morbidity and lead to better outcomes**

Interested Parties in Newborn CCHD Screening



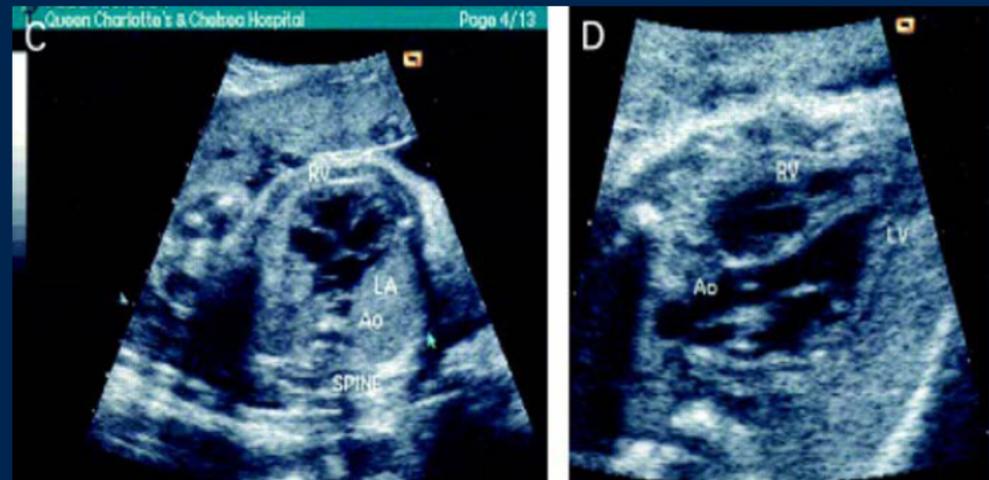
So how can we screen for CCHD?

- 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

CCHD Detection

- Fetal echocardiography
 - Highly variable, limited access
 - >50% detection rates for single ventricle lesions
 - <30% for 2-ventricle



CCHD Detection

- 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%
- 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

CCHD Detection

- PE, EKG, chest X-ray (routine)
 - 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 false positive results – PFO's, PDA, small VSD's

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

History- Pulse Oximetry

- In 1935, Karl Matthes (German physician 1905–1962) developed the first 2-wavelength ear O₂ saturation meter with red and green filters (later switched to red and infrared filters)
- In 1940 Glenn Millikan and John Pappenheimer developed a working ear oximeter for use by pilots in WW2



History- Pulse Oximetry

- In 1972, Dr. Aoyagi developed the pulse oximeter using the ratio of red to infrared light absorption of pulsating components at the measuring site
- O₂ saturation: a measure of how much oxygen the blood is carrying as a percentage of the maximum it could carry
 - Dependent on Hgb, Oxygen carrying capacity

Pulse Oximetry Now

- Typically it utilizes a pair of small light-emitting diodes (LEDs) facing a photodiode through a translucent part of the patient's body, usually a fingertip or an earlobe. One LED is red, with wavelength of 660 nm, and the other is infrared, 905, 910, or 940 nm
- Absorption at these wavelengths differs significantly between oxyhemoglobin and its deoxygenated form; therefore, the oxy/deoxyhemoglobin ratio can be calculated from the ratio of the absorption of the red and infrared light.



Pulse Oximetry

- Limitations

- Poor signal

- Vasoconstriction, hypoperfusion

- Does not give oxygen content

- Anemia- high sat'n , tissue hypoxia

- CO poisoning- Hgb has higher affinity for CO

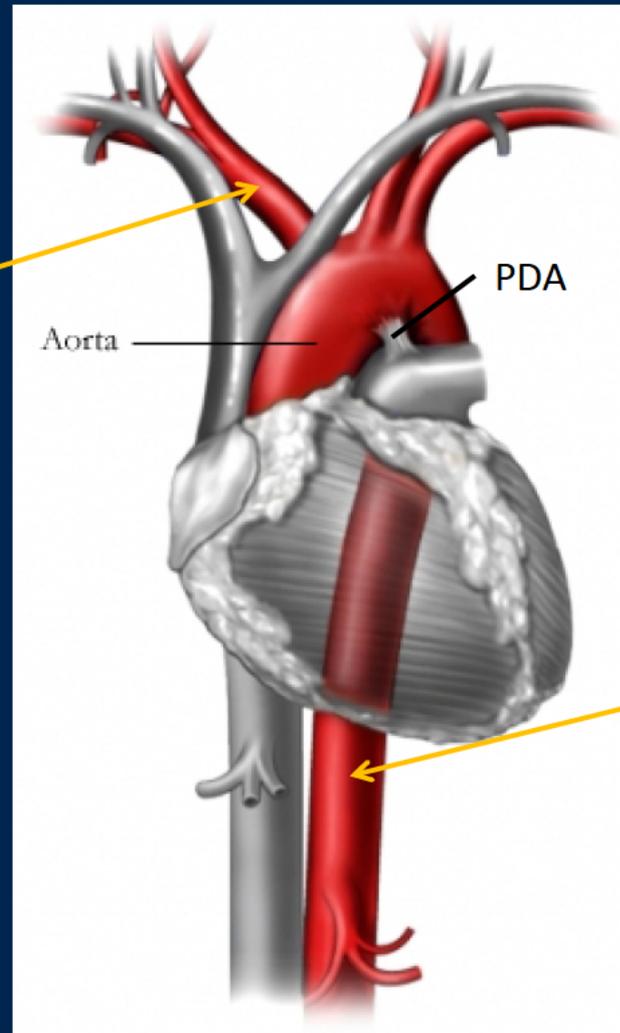
- Cyanide poisoning- high reading (but reduced oxygen extraction)

- Methemoglobinemia - low 80's

- Is not a substitute for blood gas

Pre-ductal and Post-ductal spO2

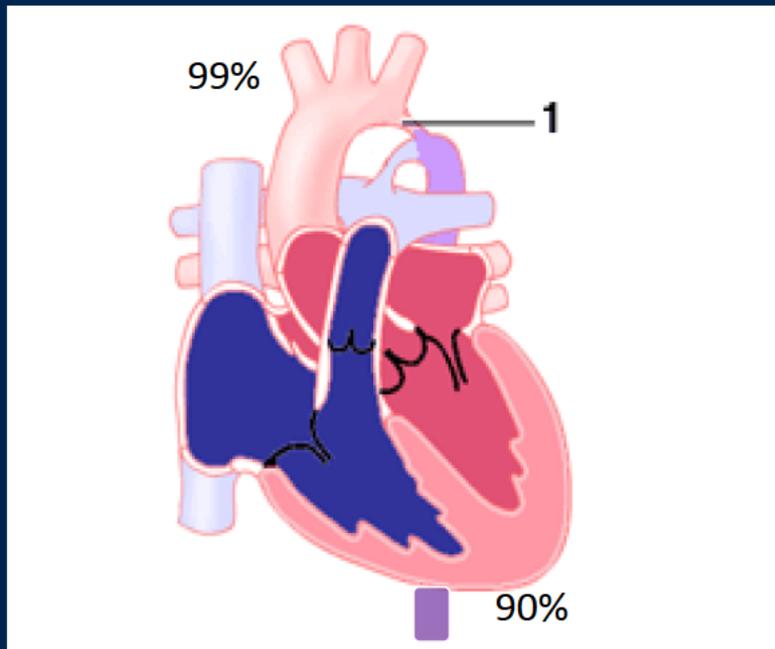
Pre-ductal



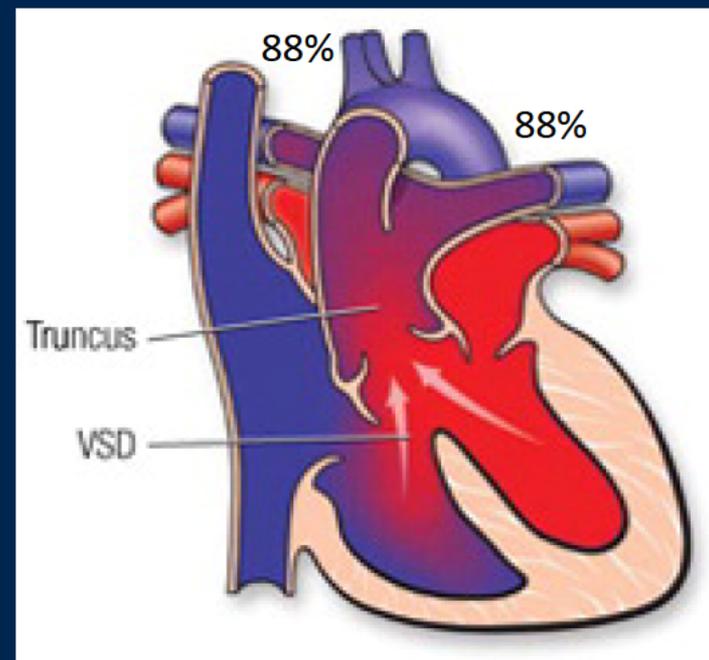
Post-ductal



Preductal and Post-ductal spO2



Coarctation of the aorta



Truncus arteriosus

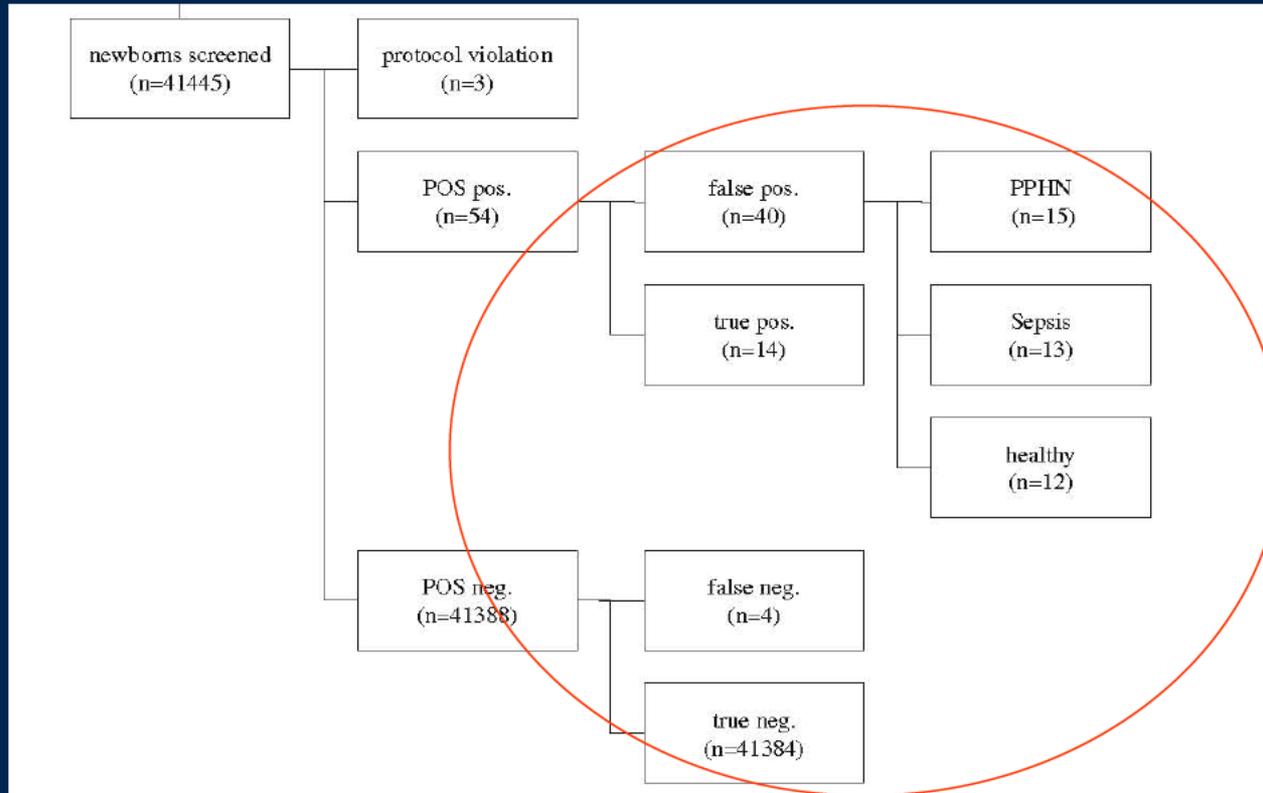
Pulse Oximetry Screening- Evidence

- Pre and post-ductal saturation to detect ductal dependent left-sided lesions
 - >2800 well baby nursery, and 32 with known CHD
 - 57 (0.02%) had abnormal test
 - 4/57 had CCHD
 - Using a cut-off of 95% in the LE, 81% of infants with CCHD were identified
 - NOT ALL were identified because not all CHD are cyanotic
- Hoke,et al, Oxygen saturation as a screening test for critical CHD. Ped Cardiol.2002.23:203-409

Pulse Oximetry Screening- Evidence

- Many 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

Pulse Oximetry Screening Program Saxony, Germany



Sensitivity 78%
Specificity 99.9%
PPV 25%
NPV 99.99%

Riede et al *Eur J Pediatr* 2009

Pulse Oximetry Screening- Evidence

- A large prospective screening of 40,000 newborns in Sweden
 - 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%
 - Thangaratinam, et al. Lancet, 2012;379:2459-64

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 may take a technician 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

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

Potential Barriers

- Screener

- Additional work load
- Education

- Potential transfer to another center

- Equipment

- Probe, machine

- Costs and reimbursement

- Patient/Parent

- False positives, false negatives
- Delay in discharge

Potential Barriers

- States have different processes
- Several programs do not publish their experience
- Reporting/Tracking/ QI issues
- Inadequate resources
- Limited US evidence-based research
- Resistance from some in the medical community

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

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

National Efforts

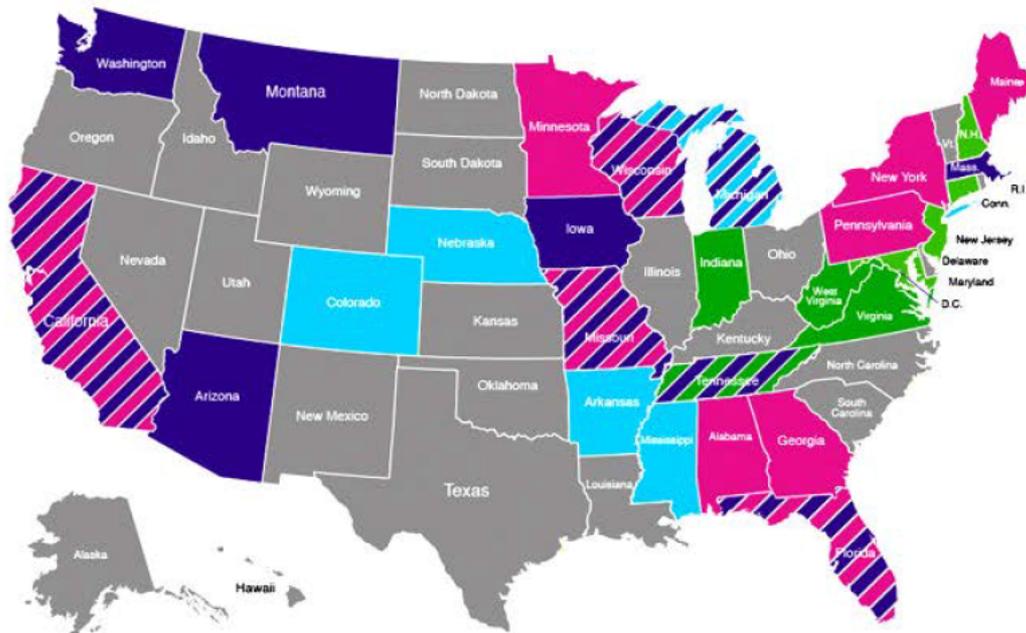
- Multi-center screening/pilots
- Health Resources and Services Administration (HRSA) sponsored demonstration projects
 - Opportunity for other states to learn and not have to “re-invent” the wheel

SACHDNC /AAP/ACCF/AHA 2012

- Health Resource Service Administration's Advisory Council on Heritable Diseases in Newborns and Children hosted a workshop to discuss implementation recommendations surrounding screening – Sept 2012
- Screening protocol based on the most current evidence available

CCHD Screening progress- Sept 2011

cchdscreeningmap.com

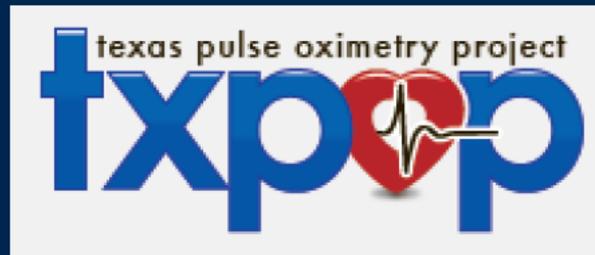


- : Legislation Passed
- : Legislation Introduced
- : Legislation Pending
- : Multi-Center Screening and/or Pilots
- : No Action States



Texas Pulse Oximetry Project- TxPOP

- A Joint Educational Initiative of The University of Texas Health Science Center at San Antonio/Department of Pediatrics, Baylor College of Medicine/Department of Pediatrics and Texas Department of State Health Services



Texas Pulse Oximetry Project- TxPOP

- 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

CCHD Screening Protocol -CDC

- 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

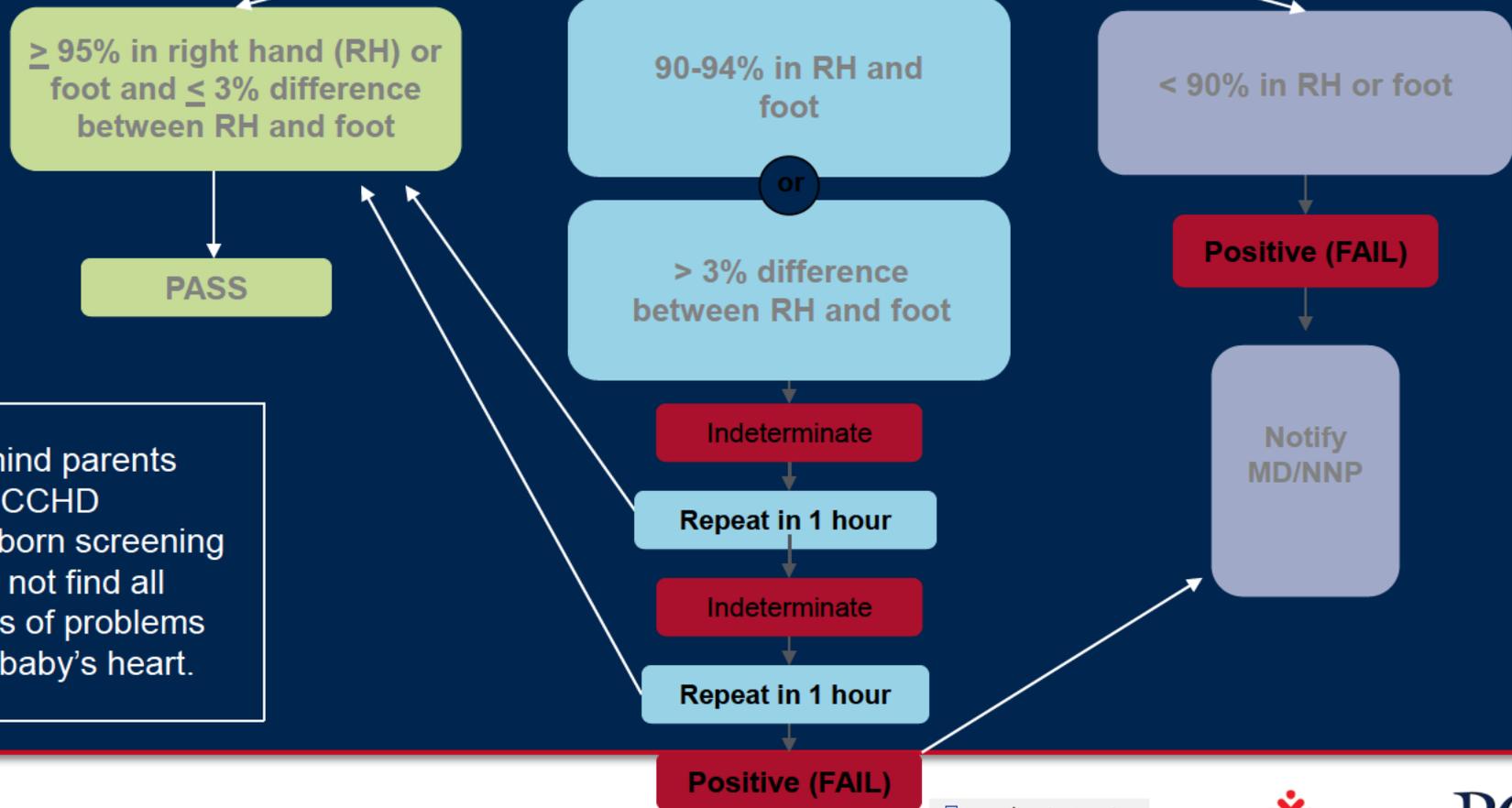
- 17-31% of all CHD's

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

CCHD Screening Algorithm

Pulse ox on right hand and foot after 24 hours



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

How to Perform Screening

- Screen after 24 hours of age
 - Conduct when infant is calm and awake



How to Perform Screening

- Perform in preductal (RIGHT hand) and postductal (one FOOT), in parallel or one after the other
 - If $< 90\%$ - **POSITIVE screen**, refer
 - If $\geq 95\%$ in EITHER extremity with $\leq 3\%$ difference: **PASS**
 - If 90 - 94% in BOTH or difference $> 3\%$: **REPEAT** in 1 hour up to 2 times, then refer



Evaluation for Positive Screen

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

Echocardiography

“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

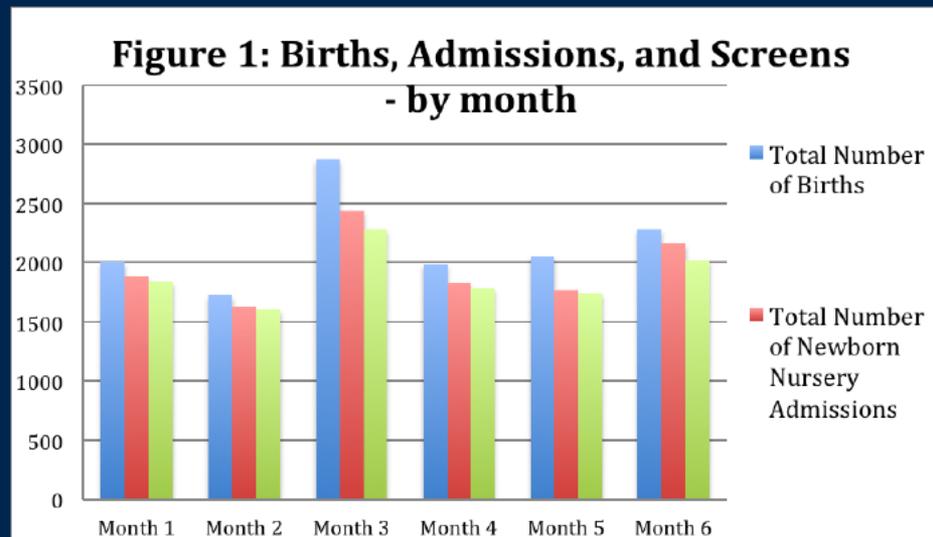
Results

- Between Feb-July 2013

- 12,946 births in 13 participating hospitals

- 11,711 newborn nursery admissions

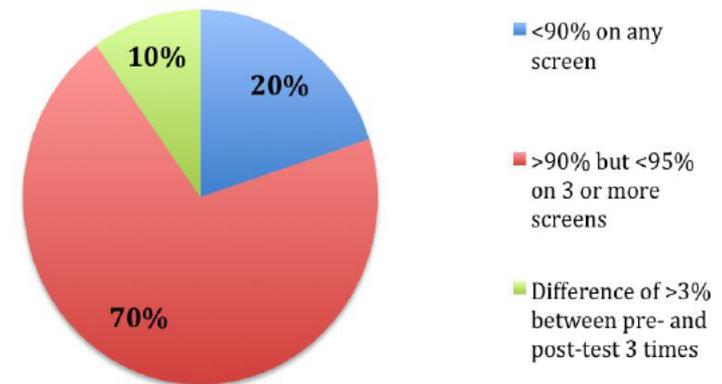
- 96% were screened between 24h- discharge



Preliminary Results

- 10 positive screens
 - 0.08 %
 - No confirmed CCHD
- 32 (0.2%) received echos
 - More echo's were ordered than positive screening tests
- Update: 1 patient transferred to TCH in Oct 2013

Figure 2: Positive Screens, by pathway



Preliminary Results



•Costs

-Screening

- Time: 5-10 minutes per patient, incl documentation
- Estimated \$5 nursing time depending on hourly rate

-Supplies

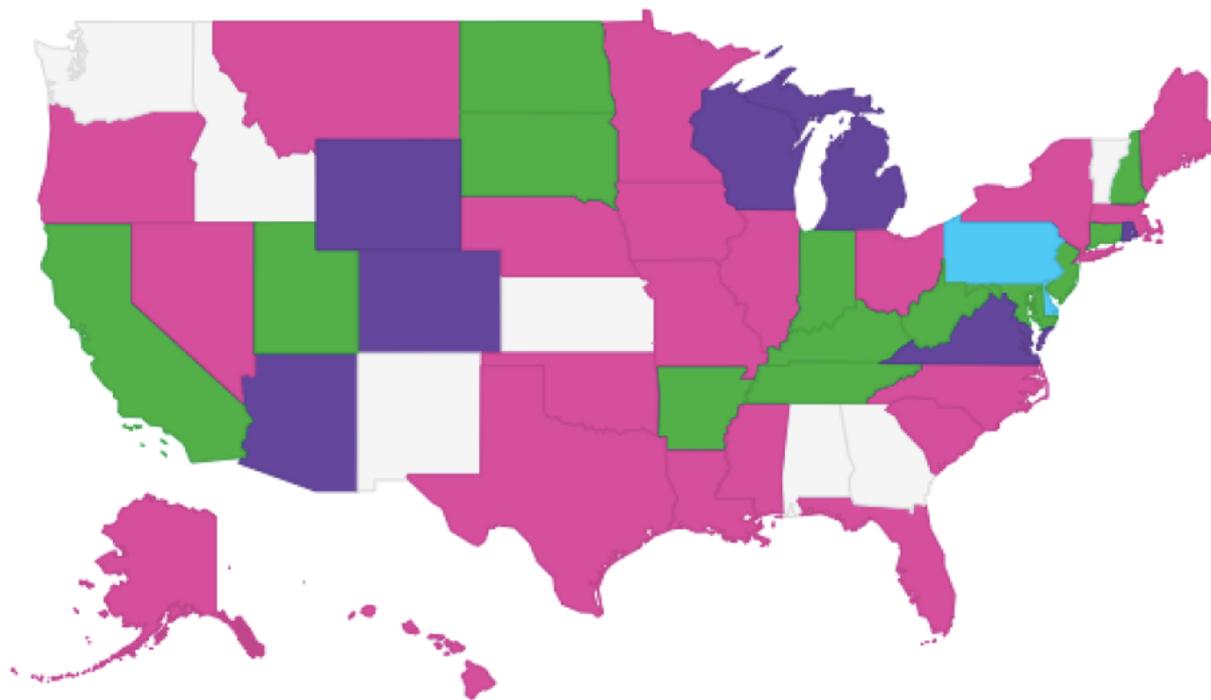
- Single probes: \$10-16 per probe
- Reusable: \$195-229 per probe

-Unknown costs

- Transport, Confirmatory testing (echo,etc)

Newborn CCHD Screening Progress

Click on a state for additional details.



Legislation Introduced Legislation Enacted Regulatory Addition to NBS Panel Multi-Center Screening or Pilot Project

TxPOP available resources

- PowerPoint presentations for nurses and for physicians
- Wall poster with algorithm to display in newborn nurseries for reference
- Laminated algorithm card to post beside pulse oximeters for reference
- Brochure, in English and Spanish, for all families of newborns
- Brochure, in English and Spanish, for families with a positive screen
- Sample physician order
- Sample newborn nursery policy
- Sample screening logs

TxPOP available resources

- Pre/Post-test instruments
- 4-minute testimonial video, “Taryn’s Story”
- 30-second public service announcement “Lifesaving Newborn Screen”
- Wall poster for display in prenatal classes to educate families
- The TxPOP ToolKit is featured on the Texas Pediatric Society web site <http://txpeds.org/txpop>
- All materials are downloadable free of charge for reproduction by anyone interested in CCHD NBS education

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Thank you!