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Screening for Critical Congenital Heart Disease in the Apparently Healthy Newborn

A presentation of *Texas Pulse Oximetry Project*:

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

WE MAKE LIVES BETTER
UT HEALTH SCIENCE CENTER
SAN ANTONIO

Texas Children's Hospital

BCM
Baylor College of Medicine

TEXAS
Department of State Health Services

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

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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
- Potential Barriers

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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 - no. in TX
  - Leading cause of death in infants < 1 year old

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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**

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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, IAA, Critical AS
  - Ductal dependent pulmonary circulation
    - PA, PS and variants, TOF
  - Complex critical CHD
    - TGA, Truncus Arteriosus, TAPVR, Single ventricle

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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**

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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

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Diagnosis vs. Screening

Diagnostic	Screening
Pros	**Pros**
Fewer resources needed	Higher detection rate
	More uniform approach
Cons	**Cons**
Identification may be too late	High resource use
Application may be spotty	Adverse impact of false positives

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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

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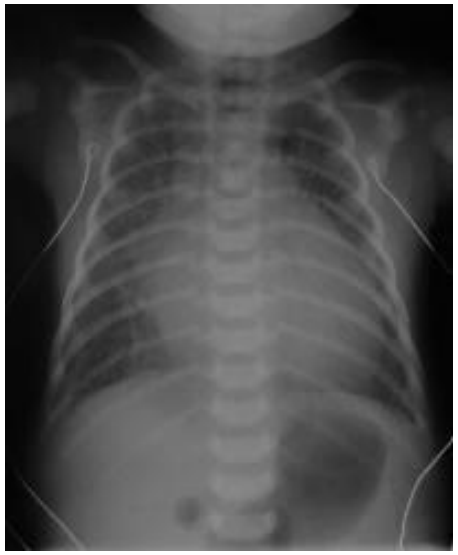
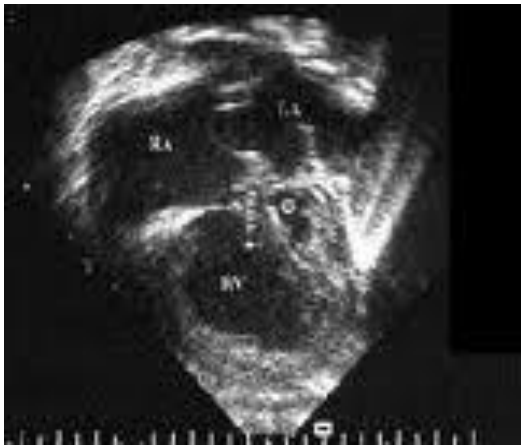
Diagnostic Process

Newborn presents in shock with murmur



Exam suggestive of CHD



Hypoplastic Left Heart

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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

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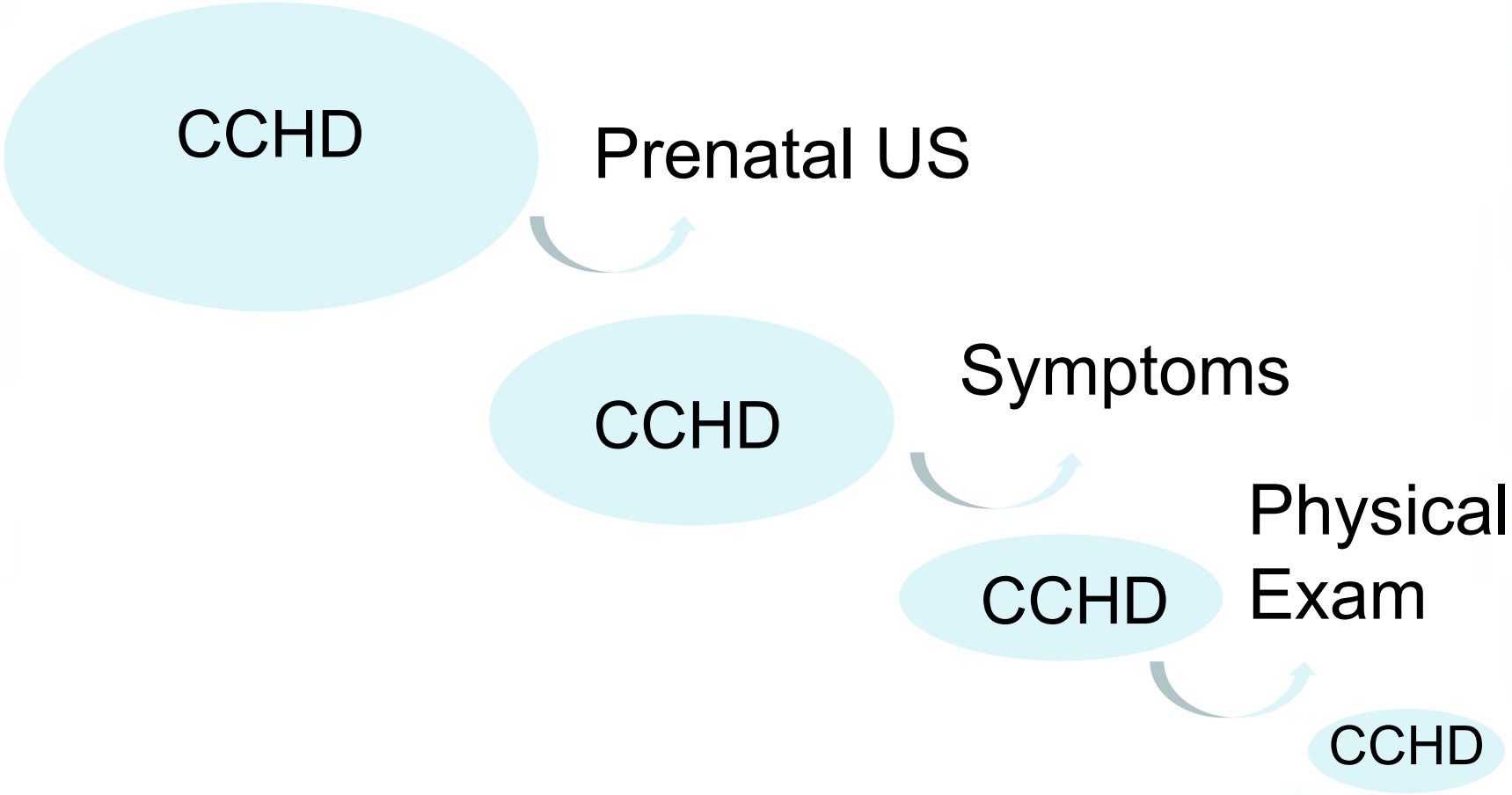
Chain of Detection



Missed Diagnosis of CCHD

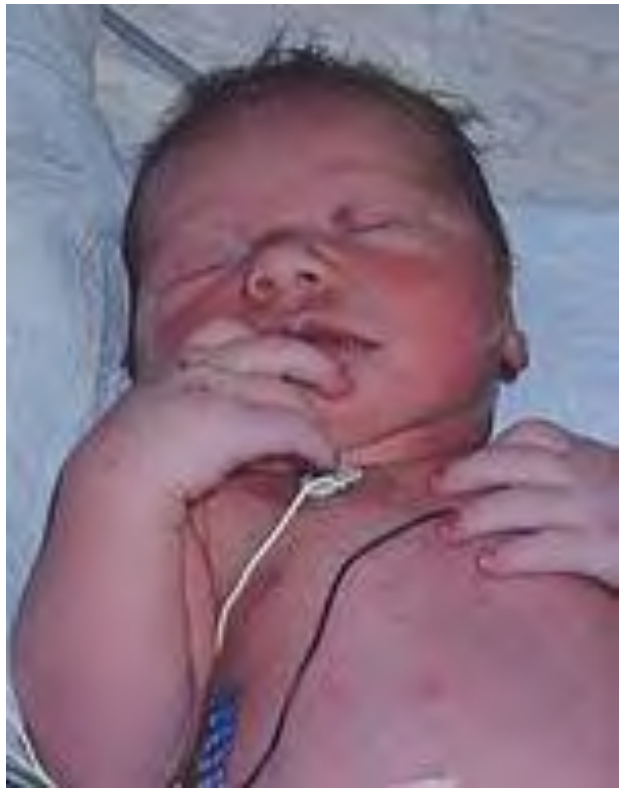
Table 2. Total Number of Patients in Each Group by Diagnosis

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.

[a] Because of rounding, percentages may not total 100.

[b] Study cohort indicates the 898 patients selected by the initial selection criteria specified in the "Patient Selection" subsection of the "Methods" section.



Chang et al, *Arch Pediatr Adolesc Med,* 2008

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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

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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

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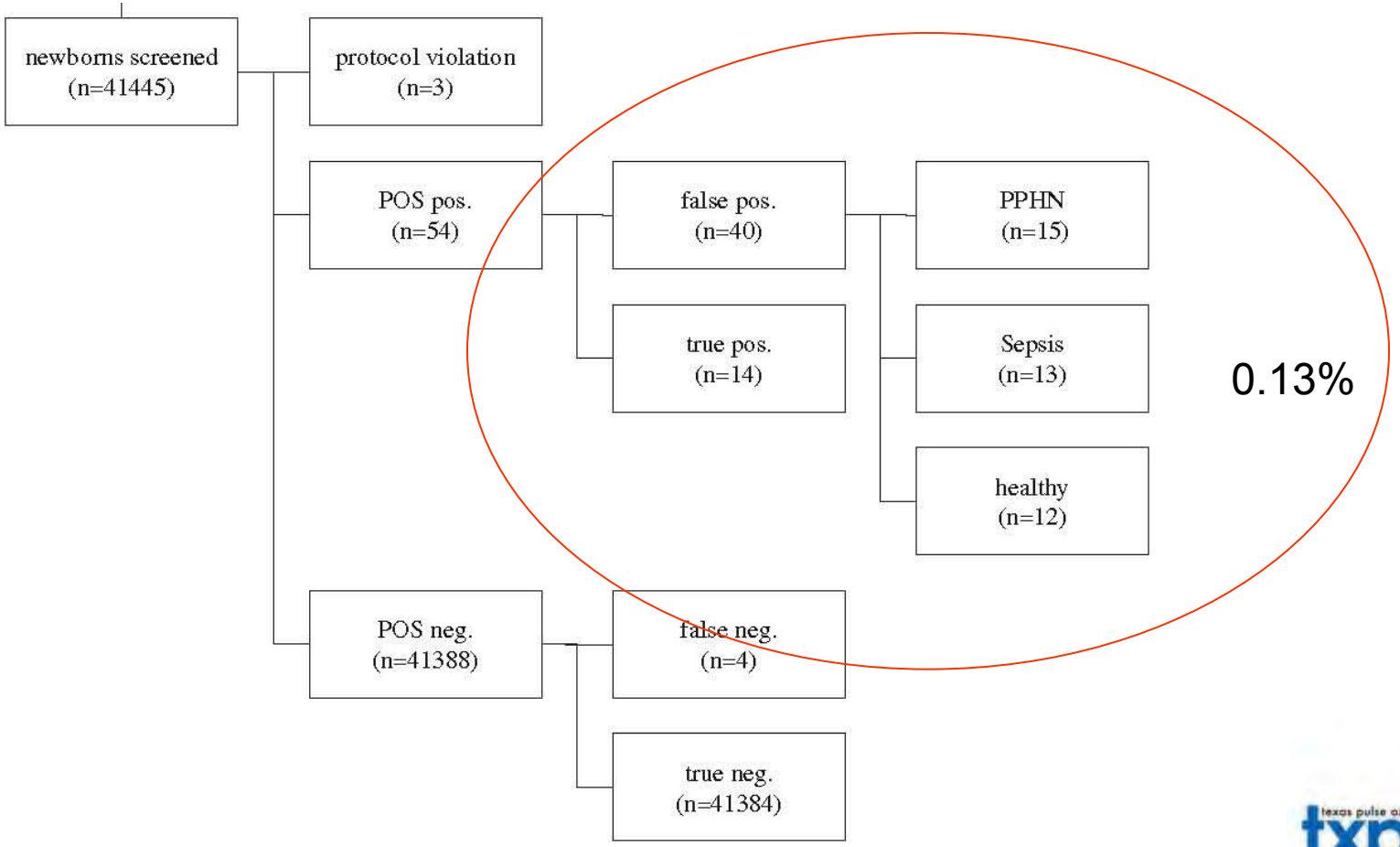
Pulse Oximetry Screening Program Saxony, Germany



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%

Thangaratinam, et al. Lancet 2012;379:2459-64

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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

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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-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

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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 sponsored demonstration projects
- Opportunity for other states to learn and not have to “re-invent” the wheel

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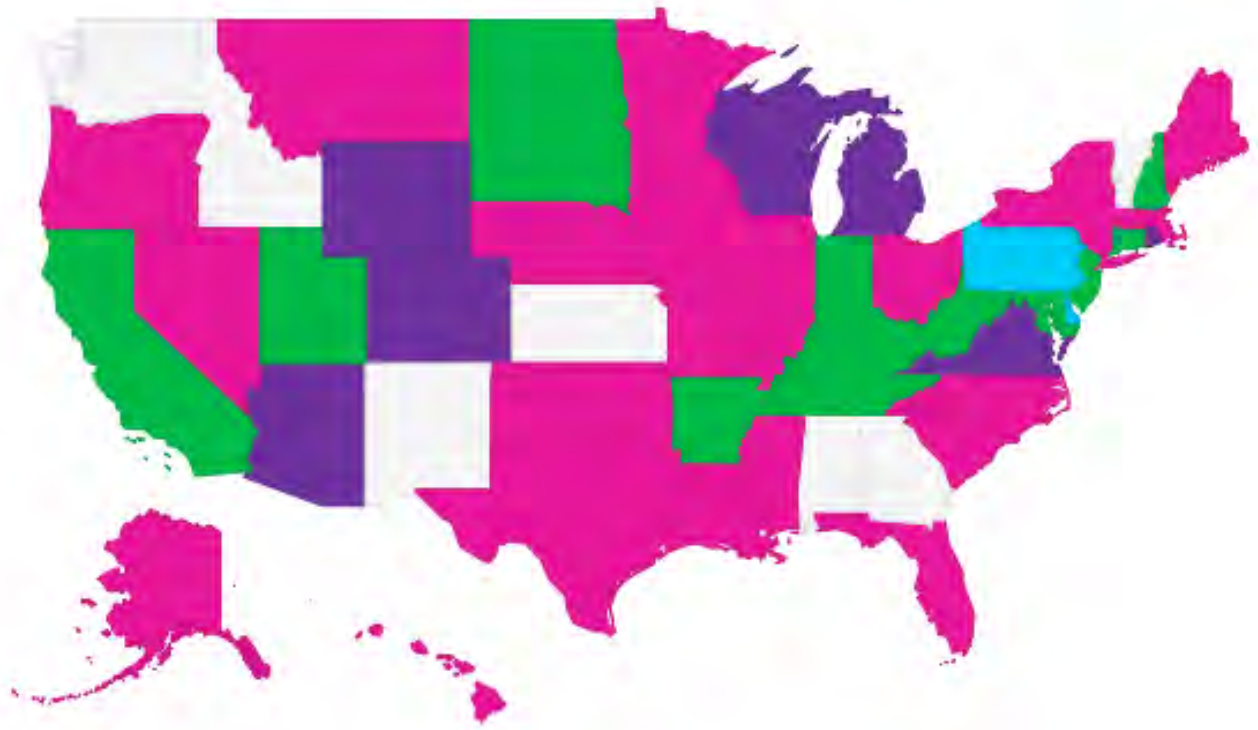
Newborn CCHD Screening Progress

Click on a state for additional details.



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

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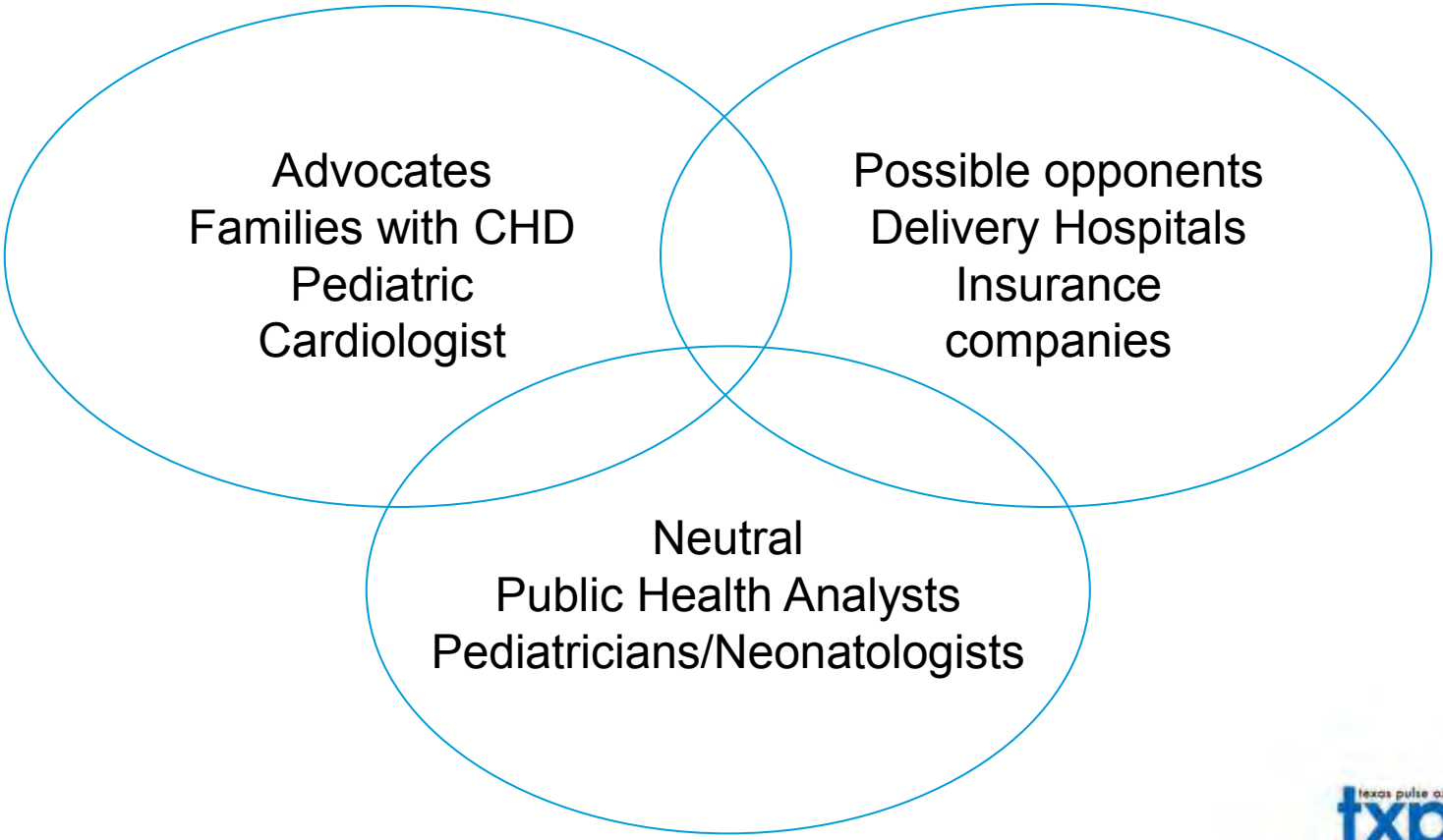
Potential Barriers

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

Potential Barriers

- 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

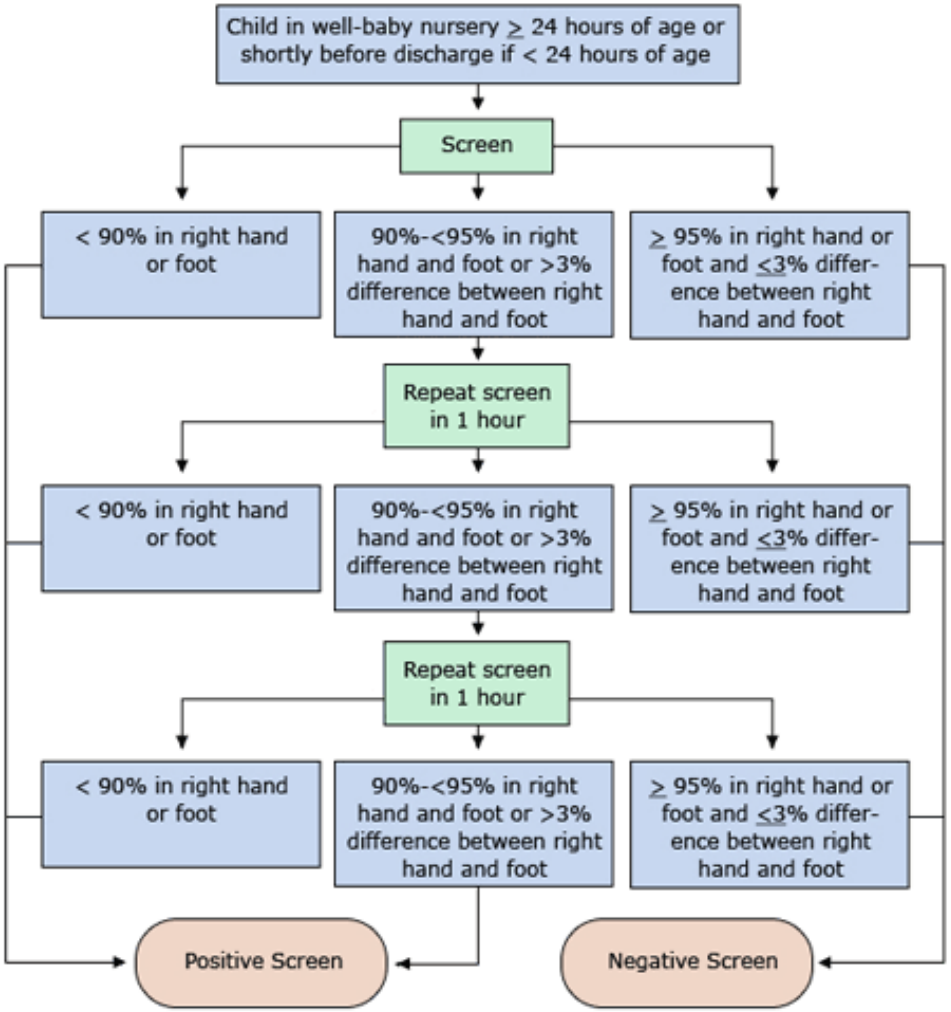
Interested Parties in Newborn Oximetry Screening



SACHDNC /AAP/ACCF/AHA

- 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

AAP/CDC Algorithm



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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

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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

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How is it done?

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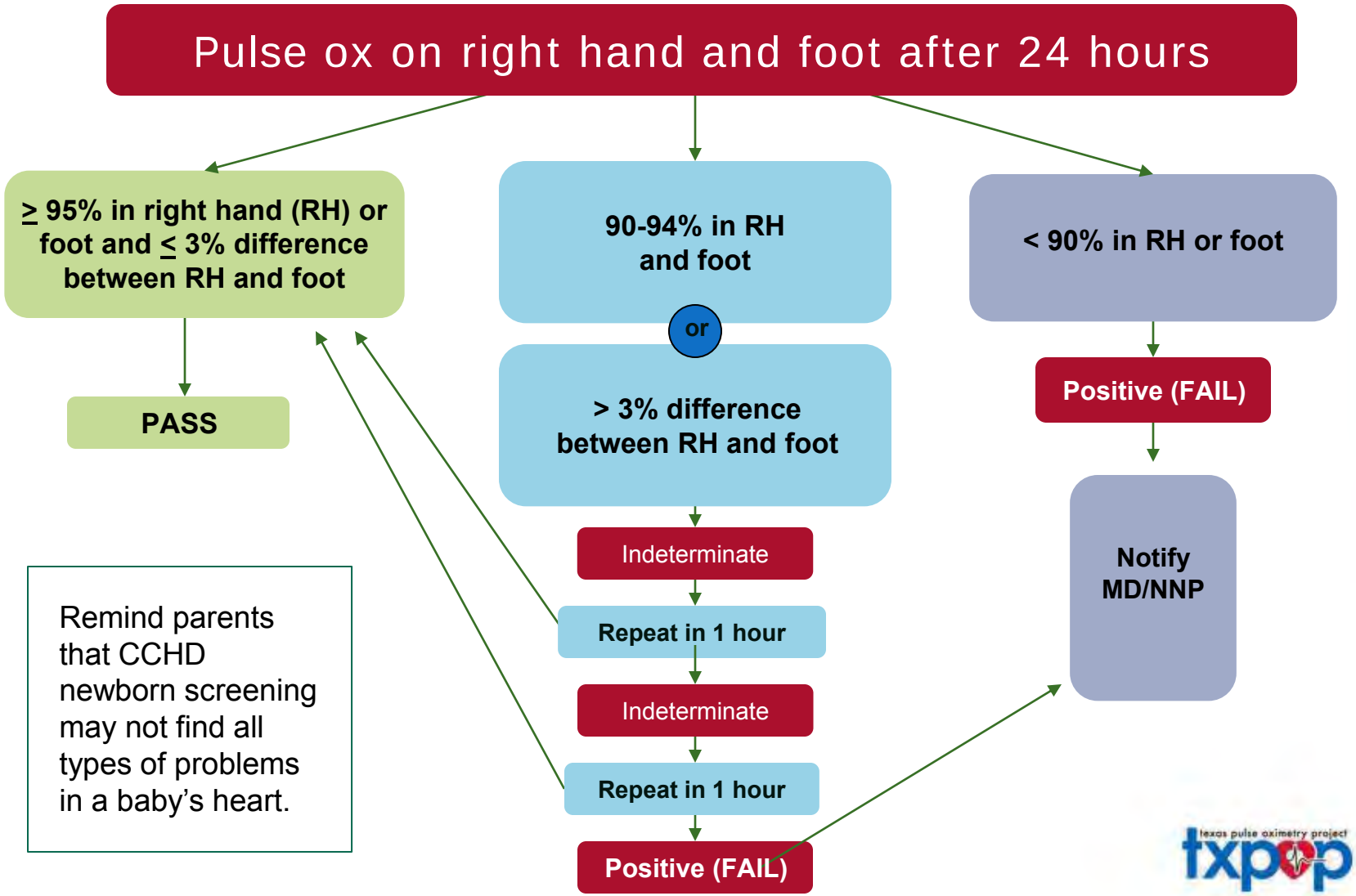
CCHD Screening Algorithm



Evaluation for Positive Screen

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

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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

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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

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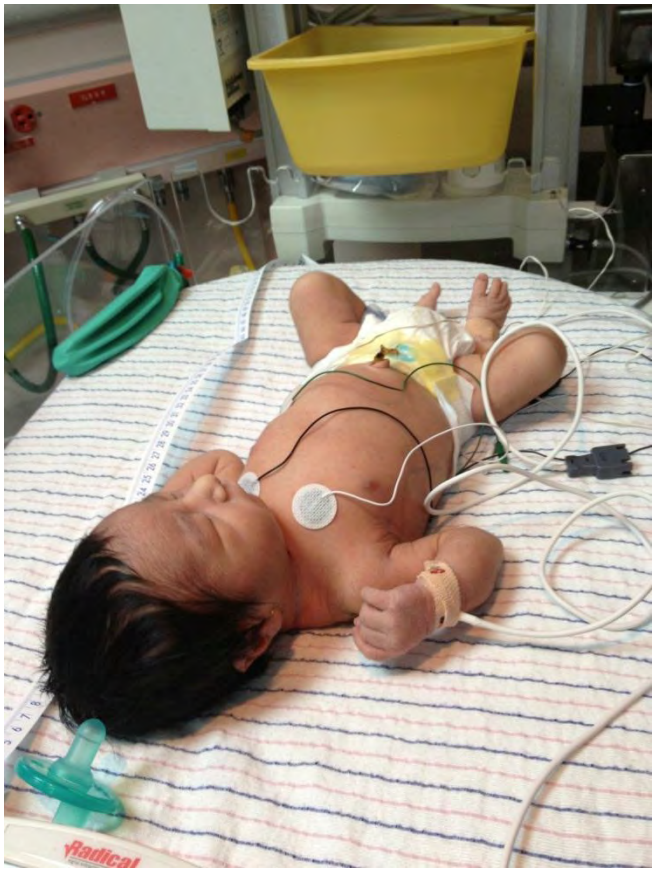
TxPOP

- 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

- 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 staff person at each facility to champion CCHD screening

TAPVR



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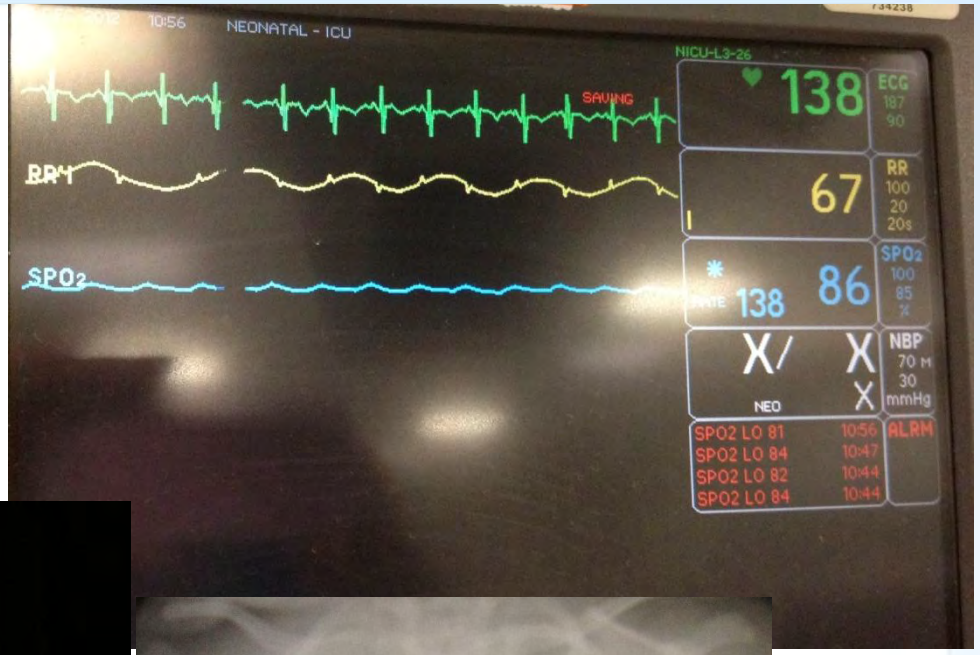
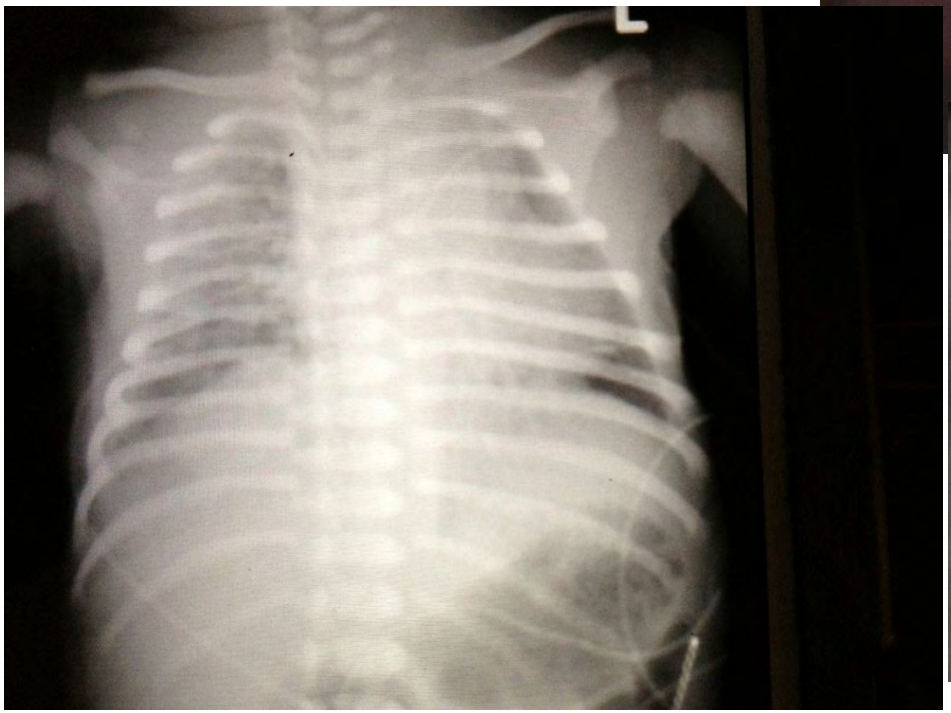
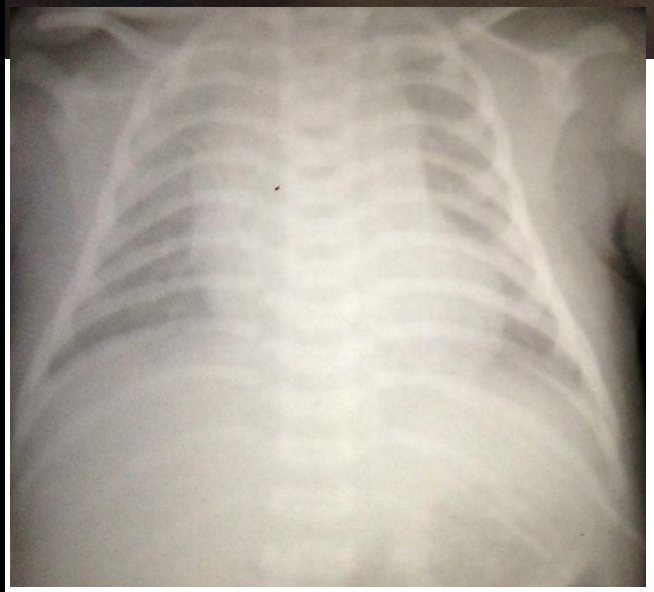
pneumonia







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Video

- http://youtu.be/2lM8hFHUMI4

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