



## 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



# Objectives

1. Discuss rationale for newborn CCHD screening.
2. Identify the steps for screening and follow-up of a newborn using the algorithm for CCHD screening.
3. Effectively explain CCHD screening results to families.

# What is Critical Congenital Heart Disease?

- A group of heart defects that causes severe and life-threatening symptoms and requires intervention within the first year of life
  - Due to hemodynamic consequences related to closing of the ductus arteriosus or dropping of pulmonary vascular resistance

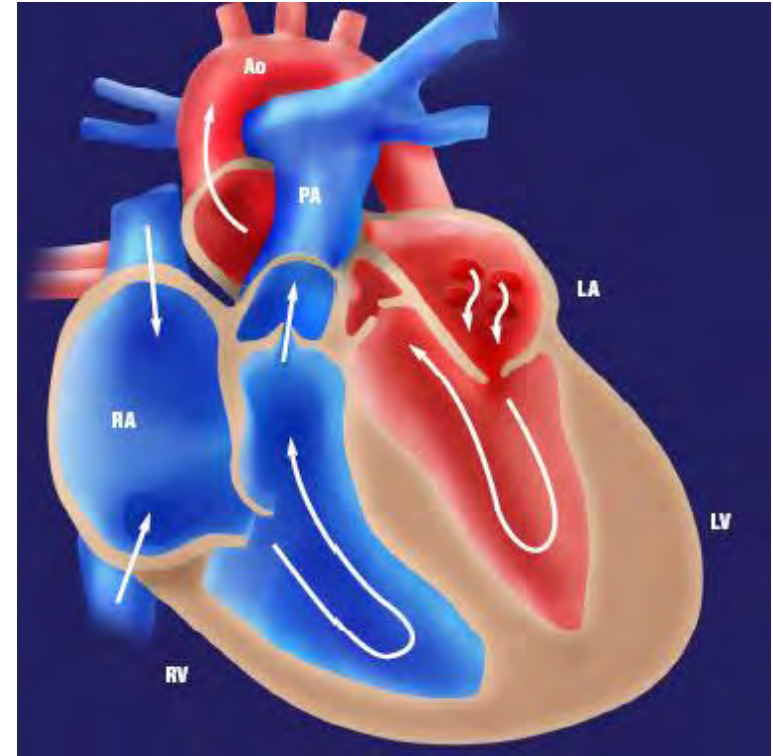
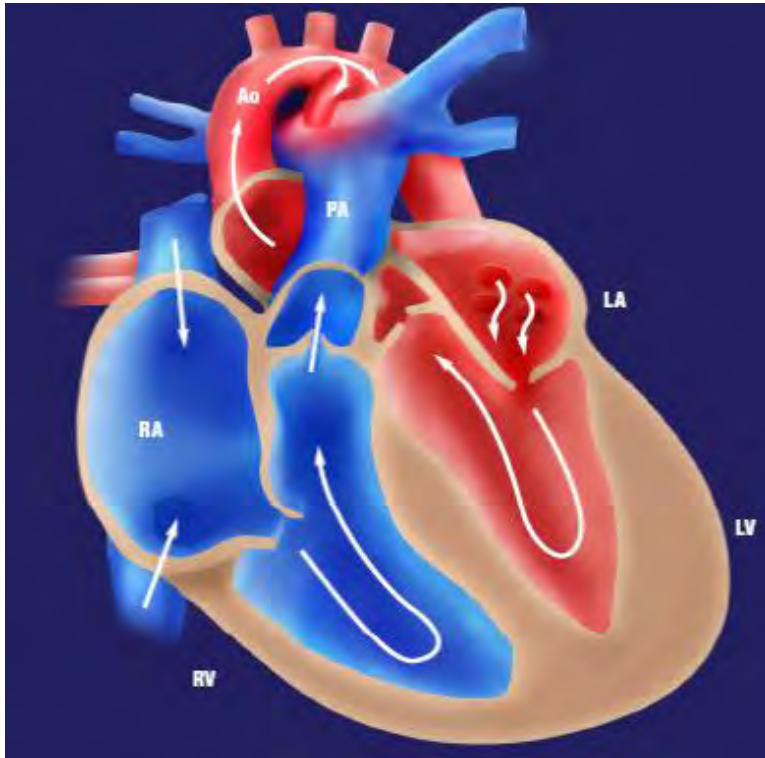
# CCHD 7

- The seven defects classified as CCHD are:
  1. Hypoplastic Left Heart Syndrome (HLHS)
  2. Pulmonary Atresia with intact septum (PA/IVS)
  3. Tetralogy of Fallot (TOF)
  4. Total Anomalous Pulmonary Venous Return (TAPVR)
  5. Transposition of the Great Arteries (TGA)
  6. Tricuspid Atresia (TA)
  7. Truncus Arteriosus communis (TAC)

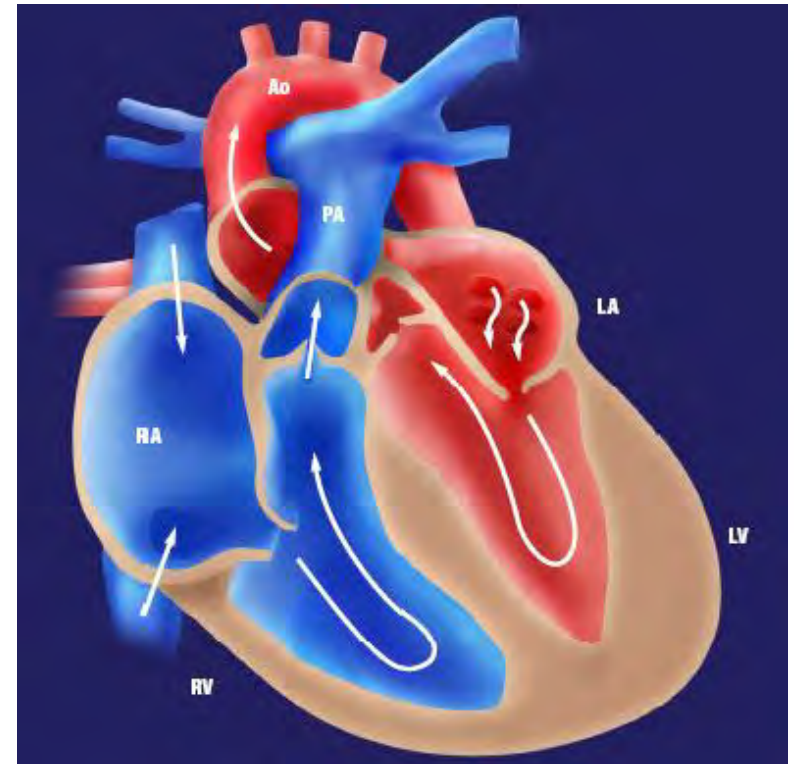
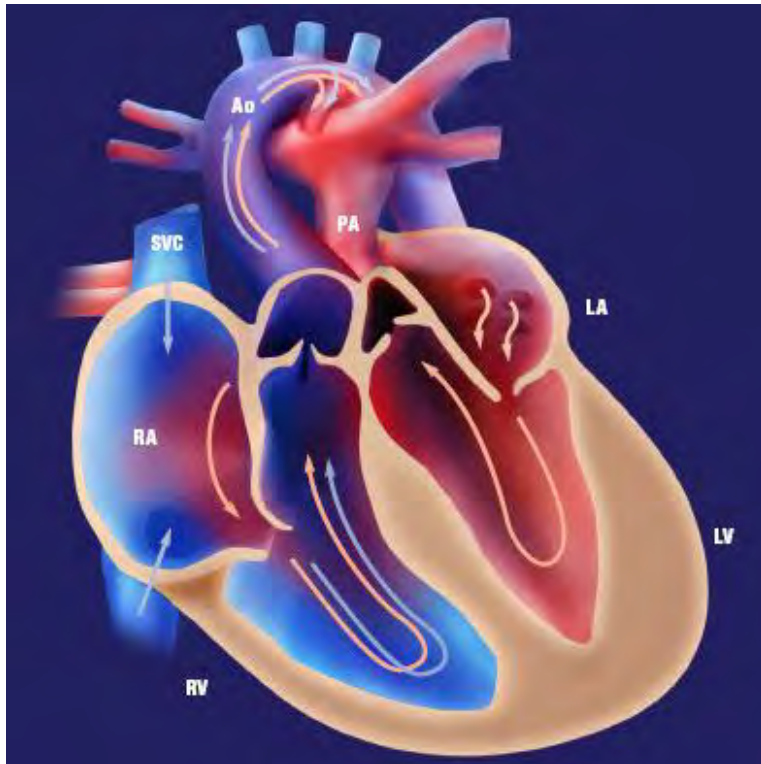
# Critical Congenital Heart Disease

- Incidence of 2/1,000 live births with potentially lethal - “critical” heart defects
  - Requires expert cardiac care and intervention in the immediate NB period or early infancy
  - Leading cause of death in infants < 1 year old

# Transitional Circulation



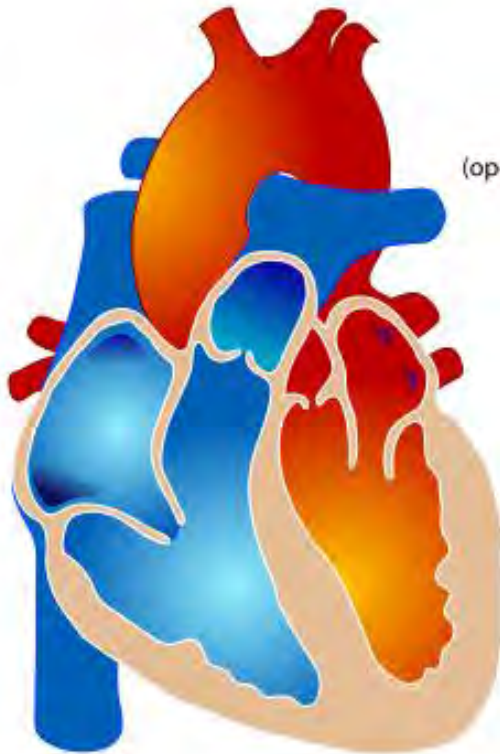
# Simple Transposition of Great Vessels



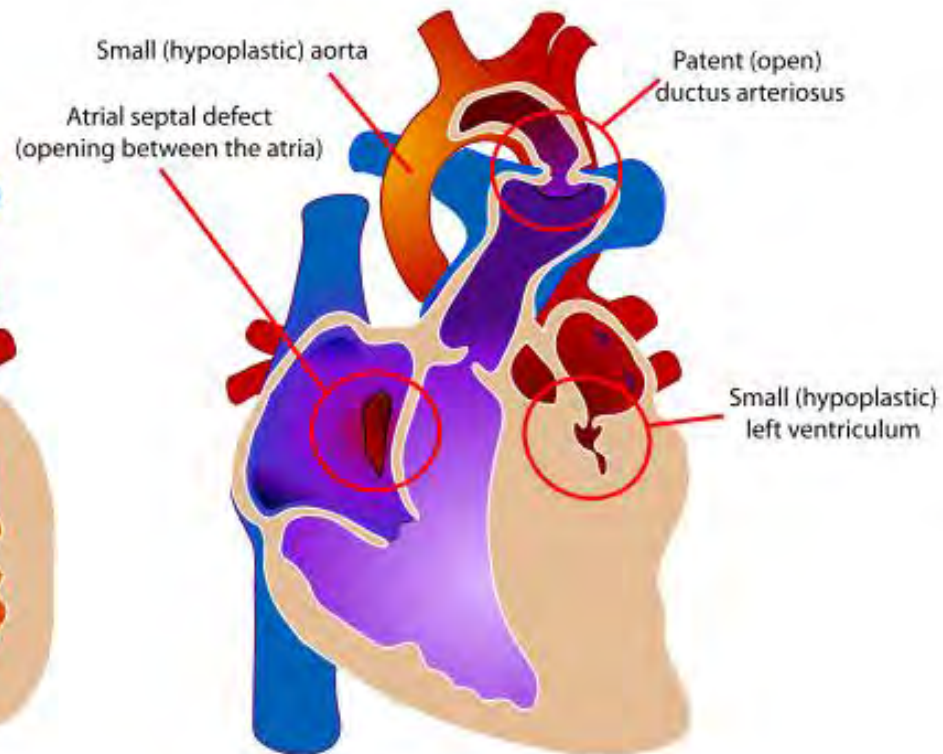


# Hypoplastic Left Heart Syndrome

Normal heart



Hypoplastic left heart syndrome





# Congenital Heart Disease vs. CCHD

- CHD is the most common birth defect.
- Approximately 8 of every 1,000 infants born in the United States each year have a form of CHD, some of which cause no or very few problems in the health and development of the child.
- CCHD comprises 25% of CHD but can bring a significant risk of morbidity and mortality if not diagnosed soon after birth.
- Failing to detect CCHD while in the newborn nursery may lead to critical events such as cardiogenic shock or death at home.

# Current Assessment of Newborns for Heart Defects

- Physical Exam
  - Blood pressures of upper and lower extremity
  - Cyanosis
  - Auscultation
  - Palpation of pulses

Physical exam might detect <50%

# CCHD Detection

- Pulse Oximetry –
  - A readily available, noninvasive, and painless technology that measures the percentage of oxygen saturation of hemoglobin in arterial blood.
  - Normal reading is 95-100%
  - Infants with **heart or lung** problems may have lower readings
  - Can detect mild hypoxemia without obvious cyanosis

# Pulse Oximetry Screening - Evidence

- 2009 scientific statement from AHA/AAP
  - Comprehensive review of all available studies
  - Compelling reasons for newborn screening
  - Called for “collaborative studies in larger populations across a range of newborn nursery systems” before universal pulse ox screening is recommended

Mahle, et al. Circulation 2009; 120: 447-458.

# Pulse Oximetry Screening - Evidence

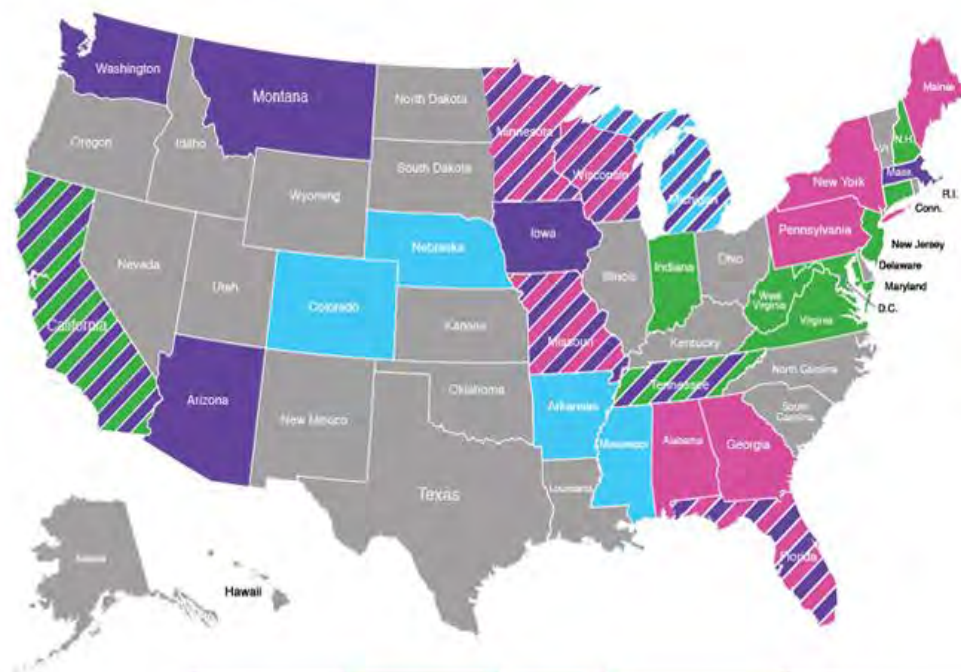
- Two separate large prospective screening of 40,000 newborns in Sweden and nearly 40,000 in Germany
- A meta-analysis of pulse ox screening for CCHD in asymptomatic newborns, involving over 220,000 newborns
- High level of sensitivity and specificity of testing

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

- In September 2010, the US Department of Health and Human Services (HHS) Secretary's Advisory Committee on Heritable Disorders in Newborns and Children recommended that screening for CCHD using pulse oximetry be added to the uniform panel of newborn screens.
- In 2011, US DSHS Secretary Kathleen Sebelius formally accepted this recommendation.

# National Movement Toward a Public Health Mandate

cchdscreeningmap.com



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





# Performing Pulse Oximetry for CCHD Screening

- The baby should be at least 24 hours of age.
  - early screening can increase false positive results
- Screening can be done in conjunction with the hearing screen and the newborn blood spot screen.
- The baby should be awake and quiet.

# Performing Pulse Oximetry (cont)

- Place probe on the infant's right hand (preductal) and either foot (post-ductal); apply to clean dry skin.
- Light emitter portion of the probe should be on top of the hand or foot with the photodetector directly opposite.



# Performing Pulse Oximetry (cont)

- Turn on Pulse Oximeter
- Connect to probe
- Wait for pleth wave (arterial pulse) to stabilize, indicating perfusion to the site being monitored and with no motion artifact
- Assess HR correlation
- Assess saturation reading
- Document

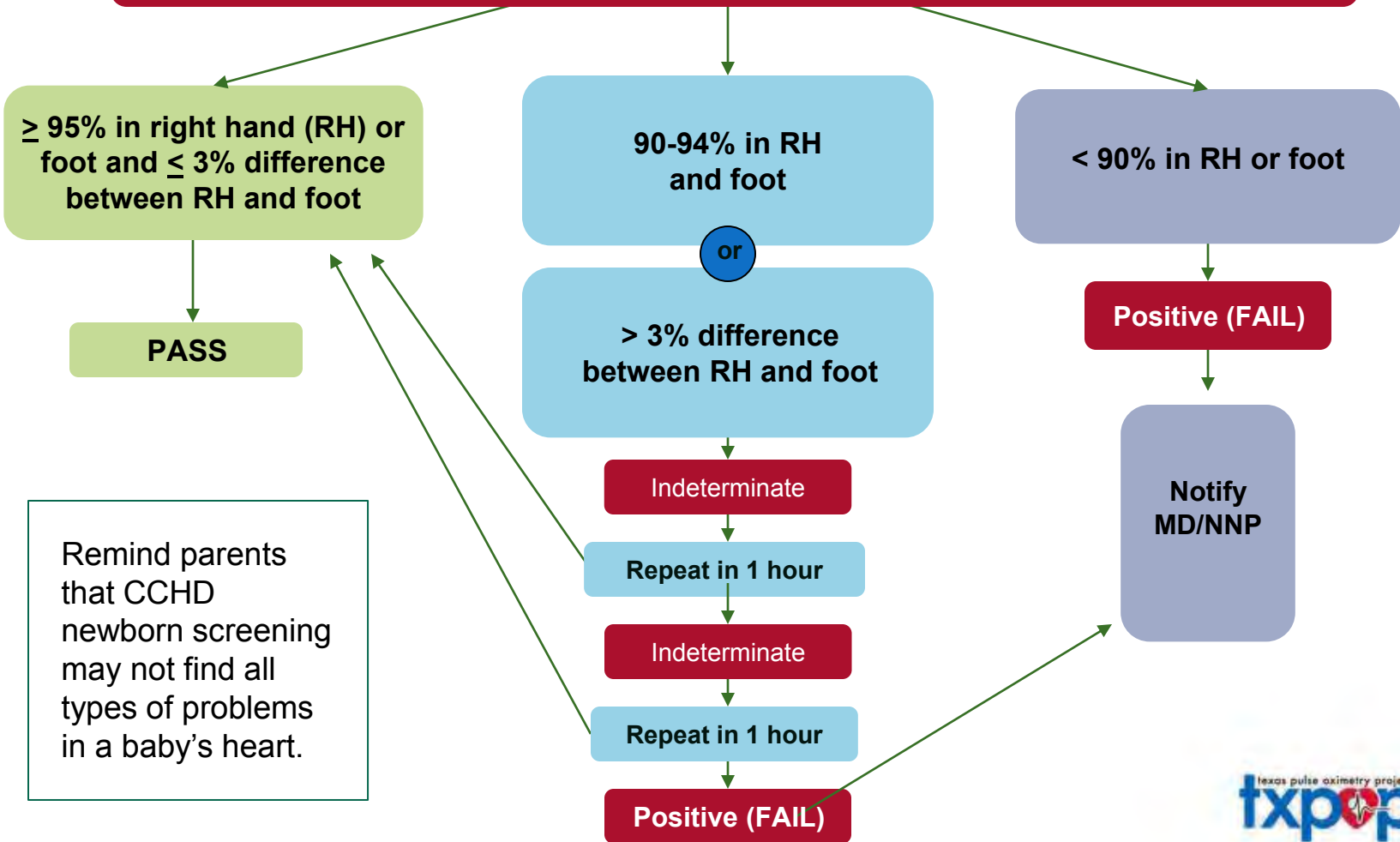


# About Pulse Oximeters

- Pulse Oximeter – standardized, neonatal, motion-tolerant, approved by FDA
- Considerations:
  - Readings are not instantaneous and are an average of readings over a few seconds. It may take up to 90 seconds to get an accurate reading.
  - Will **NOT** detect every form of CHD

# CCHD Screening Algorithm

Pulse ox on right hand and foot after 24 hours



# About Pulse Ox Results

- Threshold for a **positive** screening result relates to both the **absolute reading** as well as the **difference** between the 2 extremities (Right hand/preductal and either foot/postductal saturations).



# About Pulse Ox Results (cont)

- Saturations  $< 90\%$  is a **positive** screen and should receive immediate evaluation.



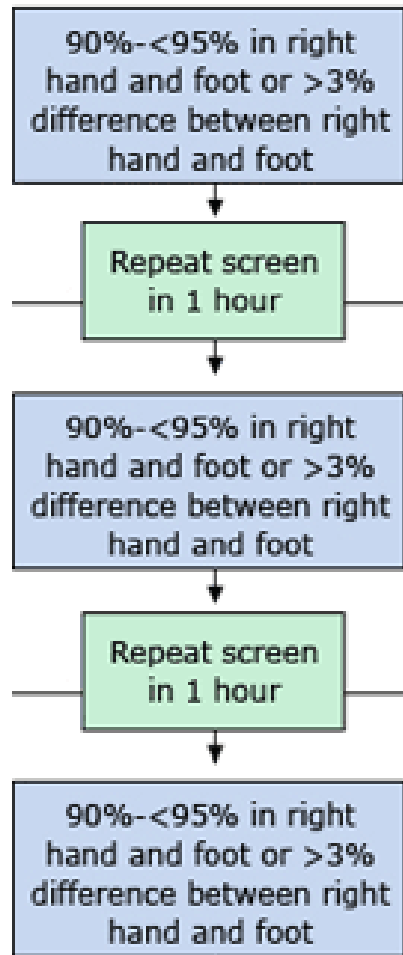


# About Pulse Ox Results (cont)

- $\geq 95\%$  in either extremity with a  $\leq 3\%$  difference between the upper and lower extremity is considered a **pass**, and screening ends.



# About Pulse Ox Results



- Repeated measurements (up to 2) 1 hour apart should be performed in cases in which the saturations are **between 90 and 94% or > 3% difference** between right hand and foot.

# In the event of a positive screen

- Evaluation by a physician
- Critical congenital heart disease needs to be excluded with a diagnostic echocardiogram and interpretation by a pediatric cardiologist.
- Infectious and pulmonary causes of hypoxemia should also be excluded.

# Documentation

- Physician Order: CCHD screening before discharge
- Patient Record: CCHD screen results with pulse ox saturations of right hand and foot
  - Pass
  - Positive (Notify MD)
- Log Book or EMR:
  - Log all screened results
  - Log all the POSITIVE results in the CCHD log book with f/u documentation



# Talking With Families

- Pass screen:
  - Explain purpose of screening: to help detect some serious heart problems in well appearing babies
  - Explain the screening process
    - Screening is performed after baby is 24 hours old when baby is quiet, warm, and awake
    - A sensor is placed on the baby's right hand and one foot; the sensor is inside a sticky strip, like a BandAid™ that can be snugly wrapped around hand and foot
    - It is painless

# Talking With Families

- Explain the screening process
  - The sensor is connected to the pulse oximeter machine which shows the level of oxygen in the baby's bloodstream;
  - Low levels of oxygen can mean there is a heart or lung/respiratory problem
  - Even with a passing screen result, it is possible that there could still be a heart problem
  - Remind parents about signs and symptoms to watch for that could suggest heart disease
  - Stress the importance of keeping PMD appointments



# Talking With Families

- Positive screen
  - If the baby has a positive result, inform the family that the doctor will be called in immediately to examine the baby
  - Advise the family of the next steps
    - Echocardiogram
    - Possible transfer for echo or cardiology consultation
    - Possible intervention depending on echo findings

# Resources

Children's National Medical Center videos for health care providers and families at <http://www.childrensnational.org/pulseox/>

Genetic Alliance – Baby's First Test  
Heart Smart Screening

<http://www.babysfirsttest.org/newborn-screening/conditions/critical-congenital-heart-disease-cchd> - (includes video for parents)

National Center for Birth Defects and Developmental Disabilities/Division of Birth Defects and Developmental Disabilities/CDC – Fact Sheets for Critical Congenital Heart Disease (English/Spanish);

<http://www.cdc.gov/ncbddd/pediatricgenetics/cchdscreening.html>

Pulse oximetry Screening Online application calculator

<http://pedspulseox.com/>

# References

American Academy of Pediatrics, Section on Cardiology and Cardiac Surgery Executive Committee. Endorsement of Health and Human Services Recommendation for Pulse Oximetry Screening for Critical Congenital Heart Disease. *Pediatrics*, Vol. 128, No. 5, Nov. 2011; 190-193.

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Sebelius K. Letter to R. Rodney Howell, MD (Internet), 2011 (USDHHS Secretary of Health and Human Services Kathleen Sebelius accepts the Secretary's Advisory Committee on Heritable Disorders in Newborns recommendation to add CCHD to uniform screening panel)