

# The Use of Pulse Oximetry in Newborn Screening for Critical Congenital Heart Disease

Melissa Carmen, MD

University of Rochester

Golisano Children's Hospital

# Objectives

- To discuss the prevalence and scope of the problem of missed or untimely diagnosis of CCHD.
- To discuss the evidence supporting the routine use of pulse oximetry in newborns to detect critical congenital heart disease.
- To discuss the benefits as well as the limitations of this type of screening.
- To present the screening protocol currently in use at Strong Memorial Hospital.

# Background

- Congenital heart disease occurs in 9 of every 1000 live births.
- Approximately ¼ of these children will have **critical congenital heart disease (CCHD)**
- Critical congenital heart disease: Congenital heart disease that requires surgery or catheter intervention in the first year of life.
- Congenital malformations are one of the leading causes of infant death in the United States and other developed nations.
- **CCHD is responsible for more deaths than any other type of congenital malformation.**

# Diagnosis of CCHD

- Currently, children with CCHD are diagnosed in the newborn nursery by a variety of mechanisms.
- Usually, diagnosis relies on physical exam findings:
  - Heart murmur
  - Tachypnea
  - Cyanosis
- CXR, pulse oximetry, ABG, EKG and ECHO are only done if the baby is symptomatic or has increased risk of CHD (family history, chromosomal anomaly, etc.)

# Diagnosis of CCHD

- Physical exam usually occurs within the first 24 h of life, and may be the only exam performed by a pediatrician or other licensed health care professional prior to discharge.
  
- May have only **one** exam prior to discharge.



# Diagnosis of CCHD



- In the US, delayed or missed diagnosis of CCHD may occur in 7 per 100,000 live births, possibly more.
- Delayed or missed diagnosis may result in the infant presenting *in extremis* with sudden and profound worsening clinical status corresponding to changes in pulmonary vascular resistance and closure of the ductus arteriosus.

# Morbidity of CCHD

- Short term: cardiorespiratory compromise, hemodynamic instability, shock
- Long term: sequelae from the short term problems and/or chronic hypoxia both causing brain injury from ischemia and reperfusion.
- More likely to have impairments in motor function, speech and language, visual-motor-perception, executive function and increased use of special services.

# Mortality Associated with CCHD

- Delayed or missed diagnosis accounts for 1-2 deaths per 100,000 livebirths
- ~10% of children with serious undiagnosed CHD die either at presentation or before their first surgery.
- For undiagnosed cyanotic heart disease, 81% of patients at autopsy died in the first 3 months of life, and 16% of those died without previous symptoms.

Chang et al. Arch Pediatr Adolesc Med. 2008 Oct;162(10):969-74

MEDICINE *of* THE HIGHEST ORDER



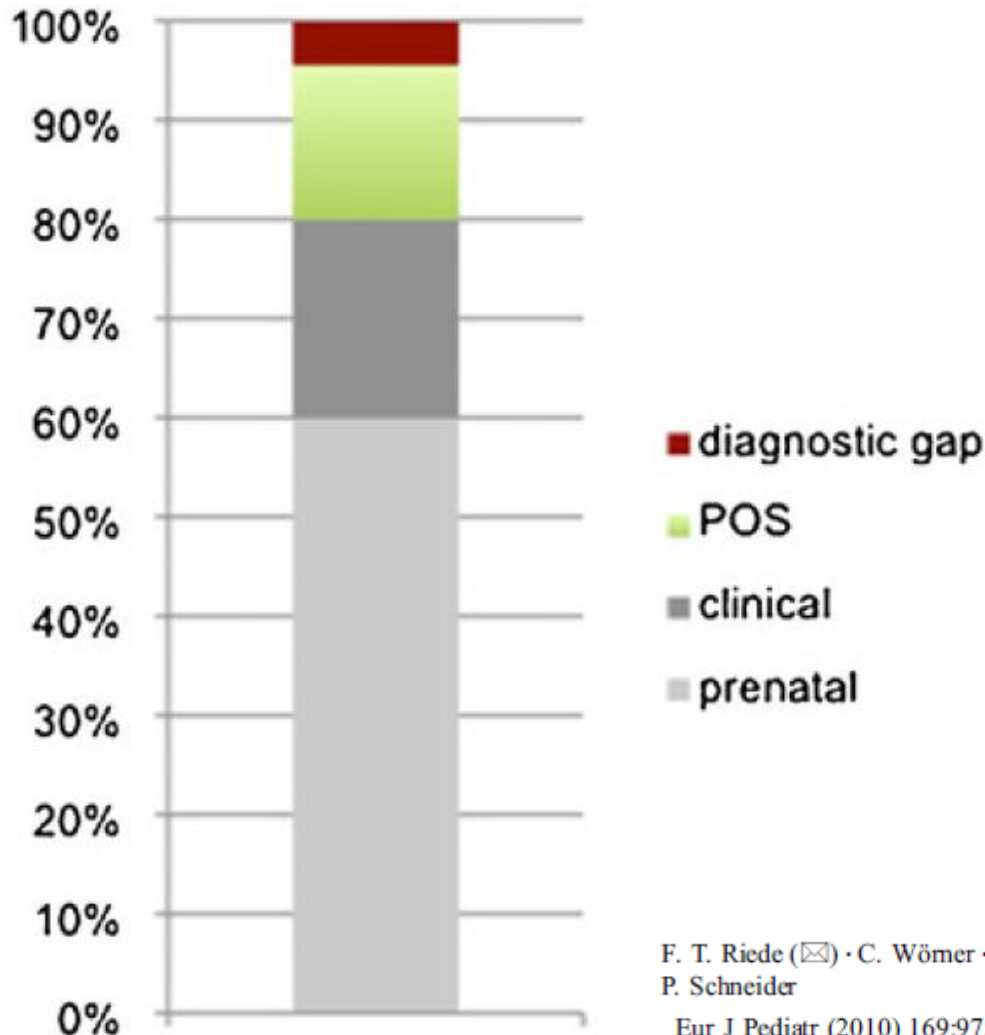
# Pulse Oximetry Can Identify Babies with CCHD

- Prospective, multicenter study performed in Saxony, Germany from 2006-2008.
- 41,445 newborns >37 weeks and no prenatal CHD diagnosis, were screened at 24-72 hours of age.
- If lower extremity sats <95% and confirmed after 1 hour, complete clinical exam and echo were performed.
- Aim was to reduce the diagnostic gap in CCHD in daily clinical routine in a variation of care centers.

F. T. Riede (✉) · C. Wömer · I. Dähnert · M. Kostelka ·  
P. Schneider

Eur J Pediatr (2010) 169:975–981

# Reduction in the “Diagnostic Gap”



- Incidence of CCHD 0.186% (90 out of 48,348 study population)

- Pulse Oximetry Screening:
  - Sensitivity 77.78%
  - Specificity 99.9%
  - PPV 25.93%
  - NPV 99.99%

F. T. Riede (✉) · C. Wömer · I. Dähnert · M. Kostelka · P. Schneider

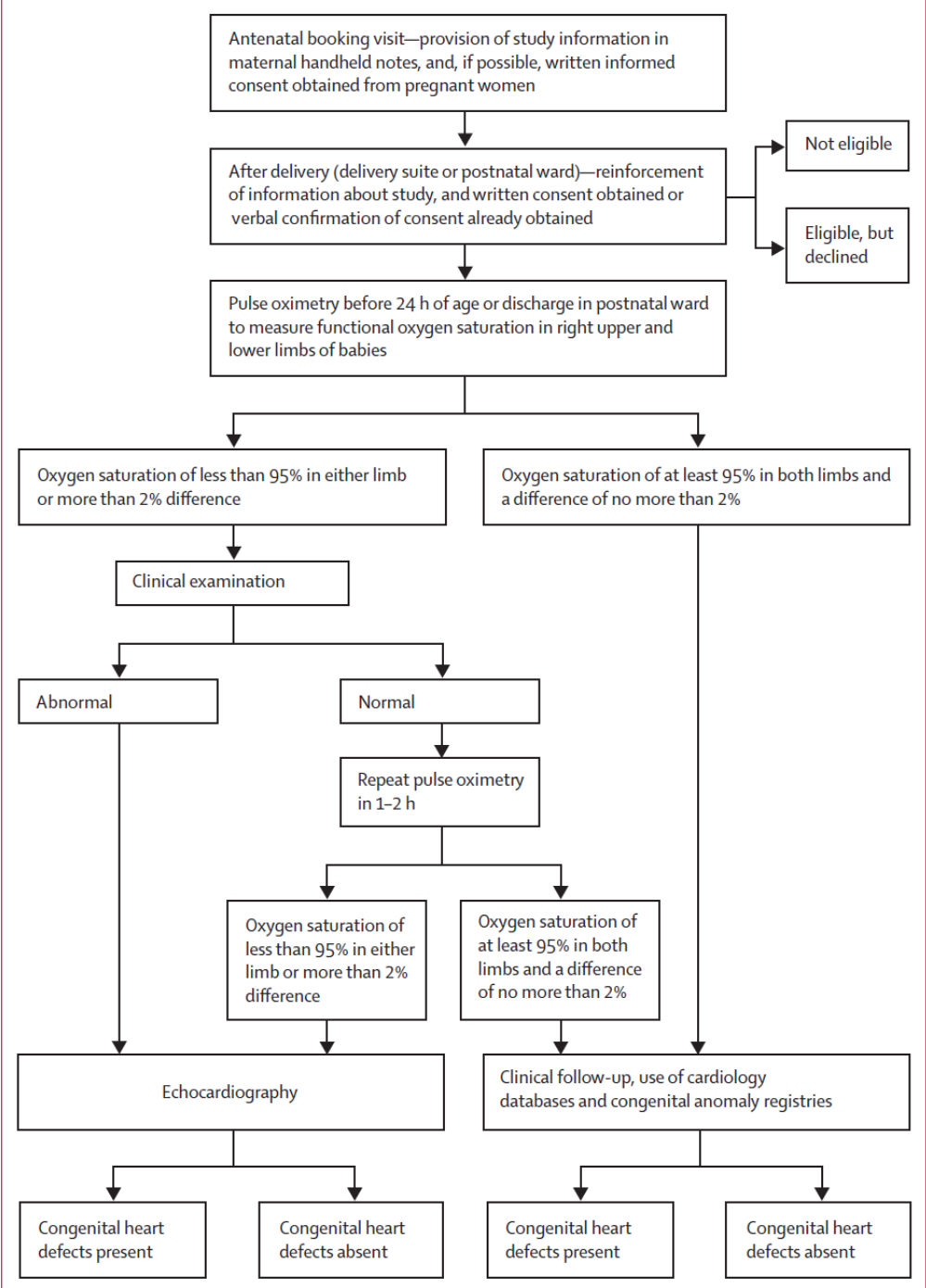
Eur J Pediatr (2010) 169:975–981

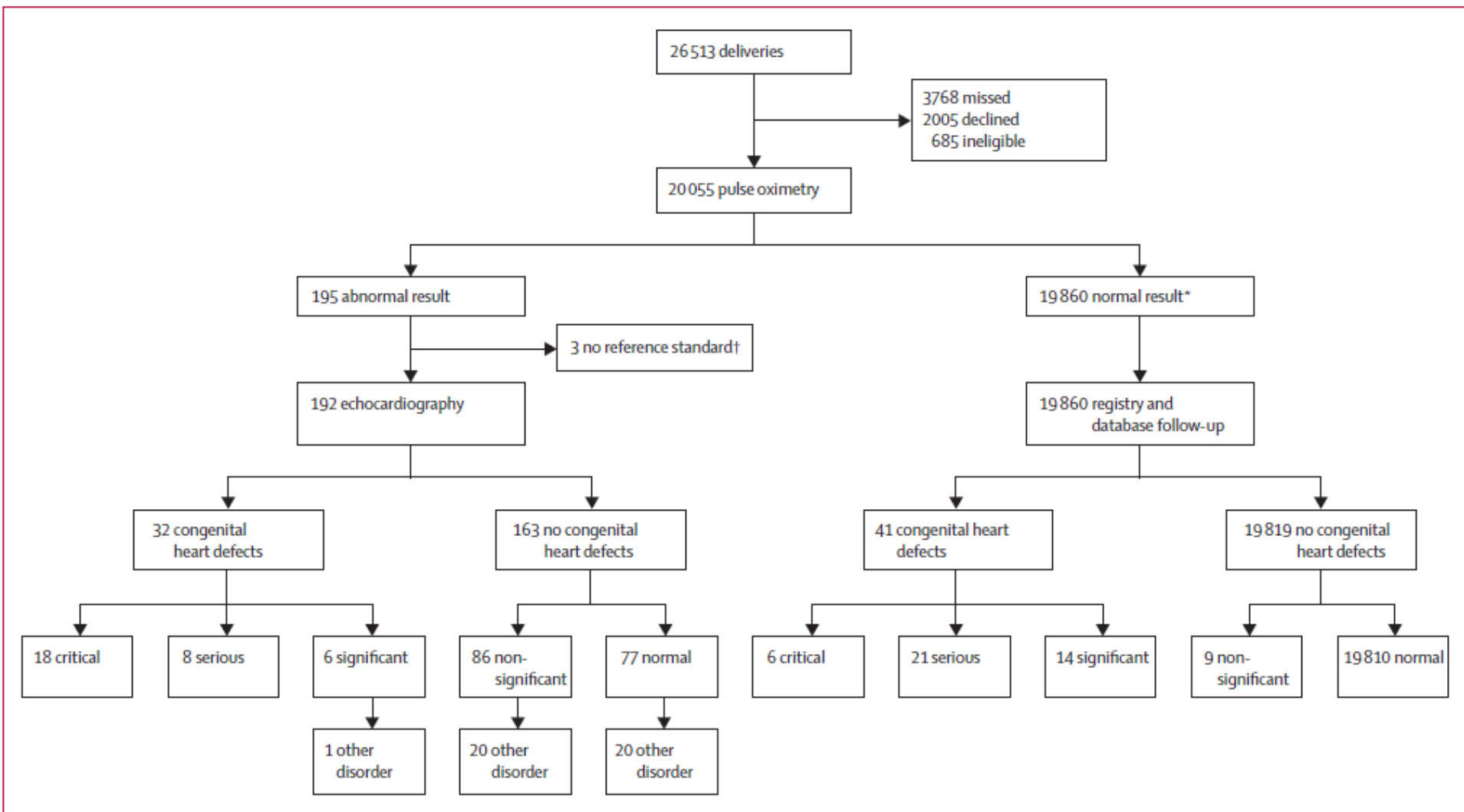
# More supporting data from the UK

- Studied infants >34 weeks at birth in six maternity units in the UK.
- Screening protocol: <95% in both RH and foot or >2% difference were abnormal. Median age of screen was 12 hrs of life.
- Two cohorts were defined: the first included all recruited babies and the second cohort excluded those with suspected CHD
- Included prenatal CCHD diagnoses but excluded babies that had symptoms suggestive of cardiac disease before screening with oximetry.
- Focused on critical ECHO findings
- Followed up babies through use of cardiology databases, regional and national registries.

Ewer et al. Lancet. 2011;378(9793):785-794.

MEDICINE *of* THE HIGHEST ORDER





**Figure 2: Trial profile**

\*Includes 78 babies who missed some or all stages of the index test after the first stage of pulse oximetry; these were followed up as per reference standard for normal result and have been confirmed as having no congenital heart defects. †Followed up as per reference standard for normal result and have been confirmed as having no congenital heart defects.

	Critical cases alone	All major cases
True positives	18	26
False negatives	6	27
False positives	177	169
True negatives	19 854	19 833
Sensitivity	75.00% (53.29–90.23)	49.06% (35.06–63.16)
Specificity	99.12% (98.98–99.24)	99.16% (99.02–99.28)
Positive predictive value	9.23% (5.56–14.20)	13.33% (8.90–18.92)
Negative predictive value	99.97% (99.93–99.99)	99.86% (99.80–99.91)

Data are number or percentage (95% CIs).

**Table 3: Accuracy of pulse oximetry in full cohort (n=20 055)**

# Early Testing is Associated with Higher False Positive Rate

- Median time of testing in this study was at 12 hours of life.
- False positive rate of 80/1000 babies tested
- Of those who did not have CCHD (163 patients with abnormal screen)
  - 86 had non-critical heart disease
  - 20 had other illness causing low sat that needed attention (e.g. slow transition, infection)
  - 57 out of 21,000 babies who tested positive were well (**2.7/1,000**)
- Raises possibility that waiting until after 12 hours to perform test may reduce the likelihood of detecting babies with slow transition

# 2011 AAP Recommendations

- Found sufficient evidence to begin screening for CCHD in well-baby and intermediate-care nurseries.
- Provided an algorithm that designed to detect infants with CCHD as early as possible while minimizing the risk of false positive results.
- Discussed the eventual progression of pulse ox screening to be as routine as newborn hearing and genetic disease screens, with future involvement of public health agencies.



# AAP's Primary Targets for Screening

- Hypoplastic Left Heart Syndrome
- Pulmonary Atresia
- Tetralogy of Fallot
- Total Anomalous Pulmonary Venous Return
- Transposition of Great Arteries
- Tricuspid Atresia
- Truncus Arteriosus

# Routine Pulse Oximetry Screening

## Pros

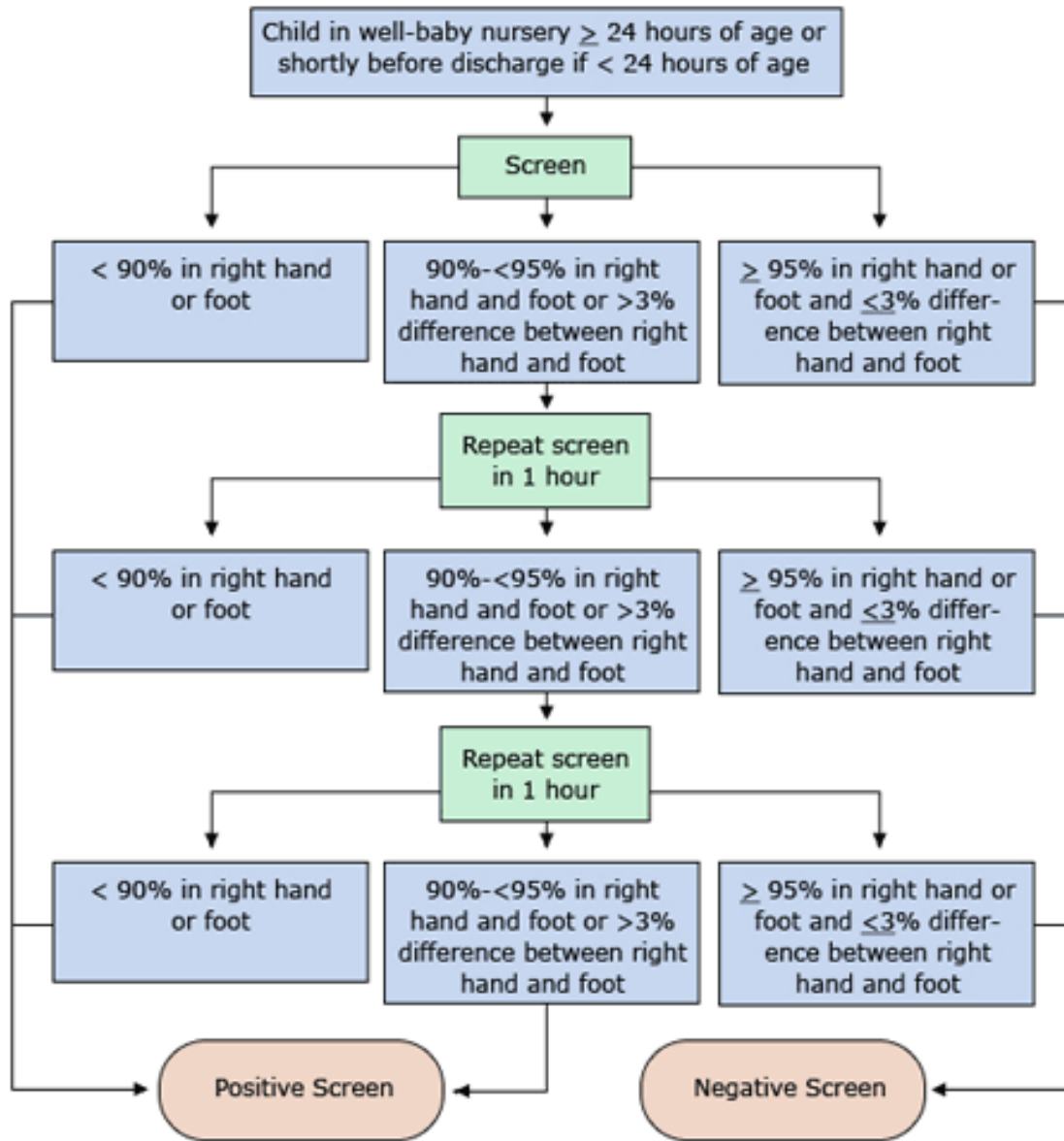
- Earlier detection of CHD, earlier treatment
- Decrease in mortality
- Detection of other conditions (PPHN, PNx, PNA, early sepsis)
- Low risk of harm, non-invasive
- Cost?

## Cons

- False reassurance if negative screen
- False Positives
- Parental and staff anxiety
- Elevated use of healthcare
- Cost?

# Current Screening Protocol at Strong Memorial Hospital

# Algorithm for Pulse Oximetry Screening for CCHD



# Implementation of screening at SMH

- URM Tool Kit – Happy to share
  - Parent Handout
    - English
    - Spanish
  - Letter to PCPs
  - Educational Materials for staff and inhouse providers
  - Documentation tool

Congenital Heart Disease Screening Program:

# Parents and Guardian Frequently Asked Questions



## ♥ What is pulse oximetry?

Pulse oximetry (ox-eh-mah-tree) is a simple and painless test that measures how much oxygen is in the blood. Another term for pulse oximetry is "pulse ox."

## ♥ How is pulse ox performed?

The pulse ox is placed by a sticky strip, like a band-aid™, with a small red light, or "probe," on the baby's h

## ♥ Why is pulse oximetry used?

Pulse ox is used to measure how much oxygen is in the blood. Pulse ox is routinely used and can be used to monitor an infant's oxygen level during a procedure or treatment. It can also be helpful in determining if an infant's heart and lungs are healthy.

Pulse ox can also help to identify babies with low levels of oxygen in their blood that may have serious heart problems. A doctor or nurse practitioner may ask for more testing such as an ultrasound of the heart, or echocardiogram (or "echo") when a low pulse ox reading is identified. The echo will screen for a serious problem in the structure of the heart or the blood flow through the heart. Pulse ox can identify a baby with serious CHD before he or she leaves the newborn nursery.

## ♥ Can the pulse ox test hurt my child?

The pulse ox test is non-invasive and painless.

## ♥ What is congenital heart disease (CHD)?

Programa de evaluación de enfermedades cardíacas congénitas:

# Parents and Guardian

## Preguntas más frecuentes



### ♥ ¿Qué es la oximetría de pulso?

La oximetría de pulso es una prueba simple y no dolorosa que mide cuánto oxígeno existe en la sangre. También conocido como “pulse ox” en inglés

### ♥ ¿Por qué se usa la oximetría de pulso?

La oximetría de pulso se usa para medir cuánto oxígeno existe en la sangre. La oximetría de pulso es una prueba rutinaria y se usa para monitorizar el nivel de oxígeno del infante durante un procedimiento o tratamiento.

También puede servir para determinar si el corazón y los pulmones del infante están sanos. La oximetría de pulso también puede ayudar a identificar a los bebés que tengan bajos niveles de oxígeno en la sangre que puedan tener graves problemas cardíacos. Un médico o enfermero/a practicante puede pedir más pruebas como ultrasonidos del corazón o ecocardiograma (o eco) cuando se identifique una medida baja de la oximetría de pulso.

El eco revisaría serios problemas de la estructura del corazón o el flujo de sangre en el corazón. El oxímetro de pulso puede identificar a un bebé con una seria ECC antes de retirarse de la unidad neonatal.



**Date:** February 17, 2012

**Regarding:** Congenital Heart Disease Screening Program

**Dear Colleague,**

We are pleased to inform you that we will be implementing the Congenital Heart Disease Screening Program (CHDSP) in our newborn nurseries beginning in March 2012. The CHDSP involves the use of pulse oximetry as a screening tool to detect critical congenital heart disease (critical CHD) in the newborn nursery. This letter will provide an overview of recommended guidelines.

#### Background and Significance

Early detection of critical CHD can potentially decrease mortality and morbidity of affected infants. Health and Human Services Secretary Kathleen Sebelius endorses screening for critical CHD as part of the recommended uniform screening panel. The American Heart Association, American Academy of Pediatrics and American College of Cardiology also support pulse oximetry screening of newborns.

#### CHDSP Screening Guidelines

The CHDSP will add pulse oximetry to detect critical CHD to the panel of standard-of-care screening tests (metabolic, hearing and bilirubin screening) done on newborns prior to discharge from the hospital. All newborns should be screened after 24 hours of age by performing pulse oximetry on the right hand and one foot. Screening test results are interpreted as follows:

- **Pass** – A newborn will be considered to have passed the screening test (i.e. - unlikely to have critical CHD) if the oxygen saturation is  $\geq 95\%$  in either extremity and the difference between right hand and foot saturation is  $\leq 3\%$ . For these newborns, no additional evaluation is required unless signs or symptoms of CHD are present.
- **Not Pass** – A newborn will be considered to have not passed the test (i.e. - may have critical CHD) and the baby's inpatient medical provider (Attending or NP) will be notified if the oxygen saturation is:
  1. Less than 90% in the hand or foot at any time **or**
  2. Less than 95% in both the hand and foot on three measures each separated by one hour **or**
  3. There is a  $> 3\%$  difference between the right hand and foot on three measures each separated by one hour



If a baby does not pass the screening test and does not have an apparent pulmonary cause of low oxygen saturation, an echocardiogram should be obtained to rule out structural cardiac defects. Referral for cardiac evaluation and neonatology consultation can be made by contacting the NICU at Golisano Children's Hospital at URMC (585-275-2198). Because infants who do not pass the critical CHD screening test have abnormal oxygen saturations, direct referral to Pediatric Cardiology on an outpatient basis is not recommended.

We are looking forward to working with you to implement the CHDSP in our newborn nurseries. Please contact us with any questions or concerns.



**Critical Congenital Heart Disease Screening Documentation at 02/14/12 1905 for Test, Nicubpa**

**Request Cosign by:** (none)

**User taken by:** URMIC INPATIENT, REGISTERED NURSE

**New Value:** Date: 2/14/2012 Time: 1905

Previous Values (as of: 02/14/12 1946)

CCHD Upper Extremity

Minutes between Upper and lower readings

Right Arm HR

Right Arm SPO2

No data filed  
No data filed  
90 % at 02/14/12 1905

CCHD Lower Extremity

Lower Extremity

Lower Extremity HR

Lower Extremity SPO2

No data filed  
No data filed  
90 % at 02/14/12 1905

CCHD Result

Attempt Number

SPO2 Difference

Result

2 (calculated) at 02/14/12 1905  
0 % (calculated) at 02/14/12 1905  
Repeat Screen in 1 hour (calculated) at 02/14/12 1905

**Accept** **Cancel**

# Implementation at SMH

- Implemented March 1<sup>st</sup> 2012
  - No true positive test results.
  - No false positive test results.

# Costs

Time!

Purchase and maintenance of screening equipment

AAP Published estimates: \$5-10 per infant (compare to \$30 per infant hearing screen)

Treatment: Echos, Telemedicine, Transport, Prolonged hospitalizations

BUT!!...

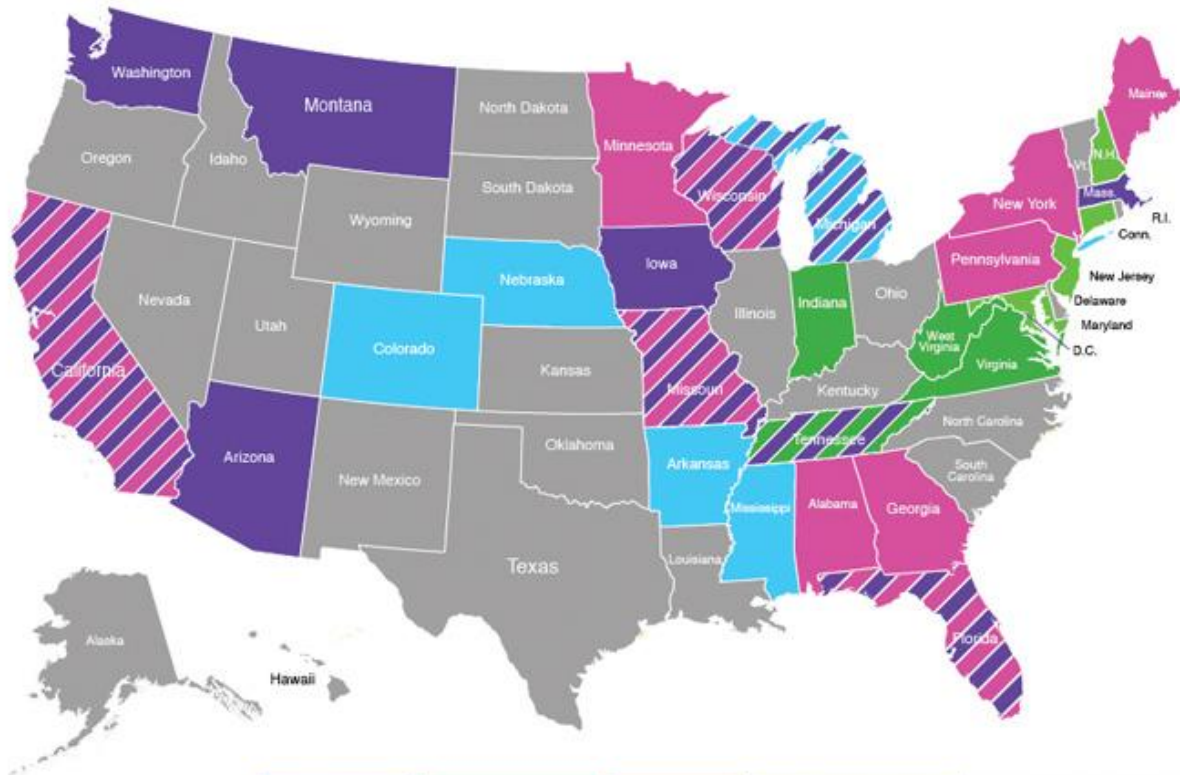
Avoided cost of care from one case of complications from circulatory collapse resulting from an undiagnosed CCHD may exceed the cost of screening two thousand newborns.

# Estimated Cost to start

- Pulse ox monitors (approx. \$6,000 each at most)
- Pulse ox probes (approx. \$20 each, 20 uses/probe)
- Pulse ox adhesives (approx. \$4 each, 1 per baby)
- CXR, EKG, ABG, ECHO, Pediatric Cardiology Consult, follow up appointments
- SMH: 3,000 births/year – 900 NICU admits = 2,100 babies/year that would require screening.
- Estimated cost ~ \$22,500 with 2 pulse ox monitors, NOT including further workup, consults, possible prolonged hospitalization or transfer to NICU, or future follow-up appointments

# Legislation to Promote Newborn Screening for CCHD

cchdscreeningmap.com



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



# Future Directions

NYS Dept of Health plans to integrate this screening with metabolic and hearing screening and track implementation, data collection, and diagnostic outcomes.

# Resources

CDC

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

Baby's First Test

<http://www.babysfirsttest.org/newborn-screening/conditions/critical-congenital-heart-disease-cchd>



UNIVERSITY *of*  
ROCHESTER  
MEDICAL CENTER

MEDICINE *of* THE HIGHEST ORDER