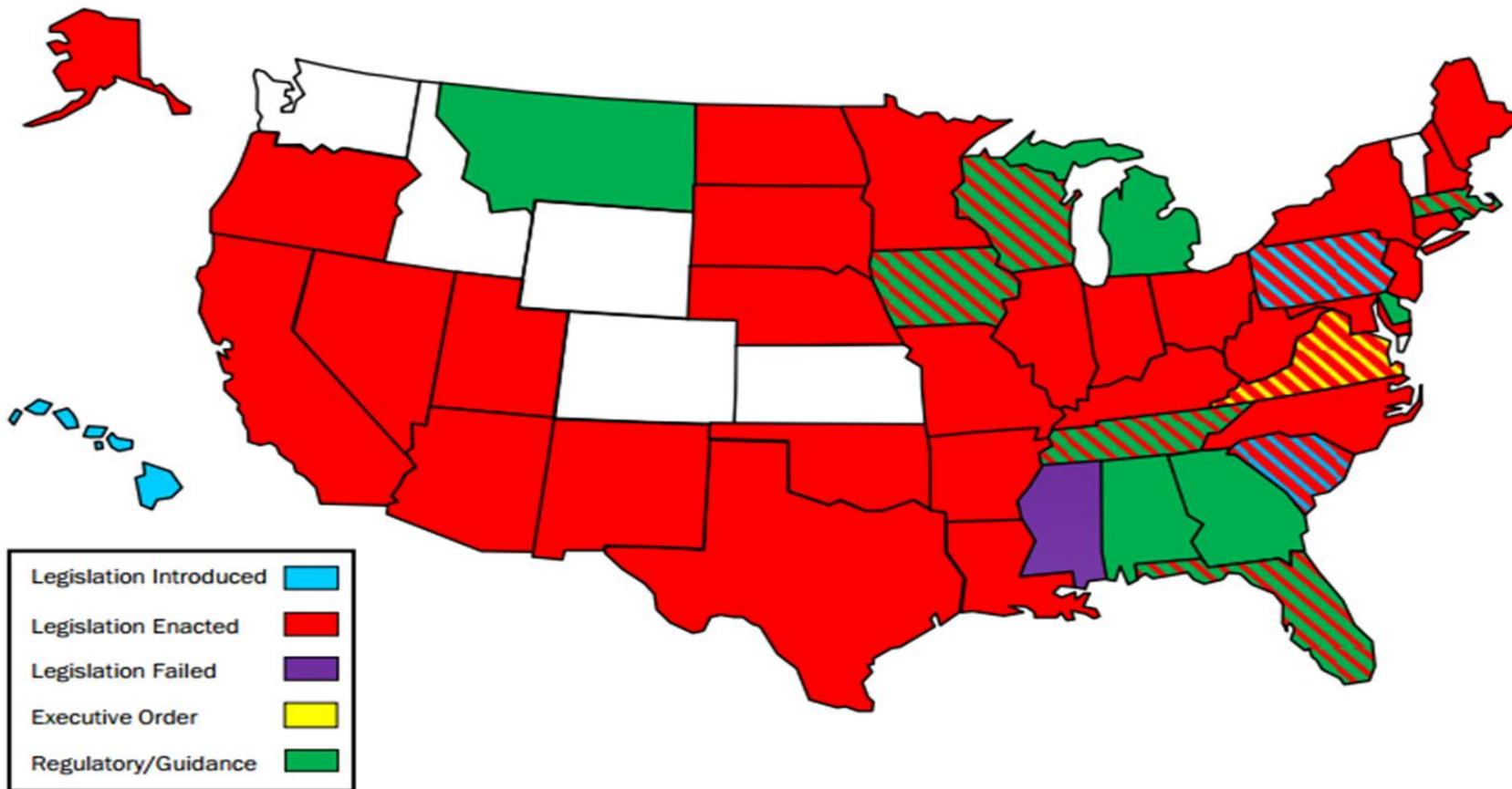


# **Critical Congenital Heart Disease (CCHD) Pulse Oximetry Screening**

Presentation to the Public Health Council  
August 2014

- In September 2011, Kathleen Sebelius, U.S. Secretary of Health and Human Services, approved the Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (SACHDNC) 2010 recommendation that all newborns be screened for critical congenital heart disease (CCHD) using pulse oximetry to prevent morbidity and mortality.
- The American Academy of Pediatrics, American Heart Association, and March of Dimes have all endorsed CCHD screening
- States have responded in a number of ways (e.g., through legislation, regulation, and pilot programs)

# Newborn Screening for Critical Congenital Heart Disease (CCHD), State Actions



\*From AAP, updated as of  
July 8, 2014

## Congenital Heart Disease (CHD)

- the most common type of birth defect in the U.S.
- occurs in approximately 9 of 1,000 live births
- A leading cause of infant deaths resulting from birth defects

## Critical Congenital Heart Disease (CCHD)

- a group of defects that cause severe and life-threatening symptoms
- requires intervention within the first days or first year of life
- accounts for approximately 25% of all cases of Congenital Heart Disease

7 critical congenital heart defects are primary screening targets based on their tendency to produce hypoxemia (abnormally low level of oxygen in the arterial blood):

- Hypoplastic left heart syndrome
- Pulmonary atresia (with intact septum)
- Transposition of the great arteries
- Truncus arteriosus
- Tricuspid atresia
- Tetralogy of Fallot
- Total anomalous pulmonary venous return

Mahle WT, Newburger JW, Matherne GP, et al., *Role of pulse oximetry in examining newborns for congenital heart disease: a scientific statement from the AHA and AAP. Circulation. 2009*

Secondary targets include defects that sometimes present with hypoxemia, but less consistently than primary targets.

For example:

- Double-outlet right ventricle
- Ebstein's anomaly
- Coartation/hypoplasia of the aortic arch
- Interrupted aortic arch
- Single ventricle

Mahle WT, Newburger JW, Matherne GP, et al., *Role of pulse oximetry in examining newborns for congenital heart disease: a scientific statement from the AHA and AAP. Circulation. 2009*

In Massachusetts, 916 cases of CCHD were identified through the Massachusetts Birth Defects Monitoring Program among live births from 2004-2009.

Of these cases, 126 (13.8%) were diagnosed after discharge from the hospital (delayed).

# Implications of Delayed Diagnoses

- Delayed diagnosis of CCHD can result in severe life threatening symptoms or death.
- Infants with undetected CCHDs are at risk of serious complications (e.g., end organ damage, motor function impairment and cognitive impairment)

# CCHD Cases by Birth Year and Timing of Diagnosis among MA Live Births

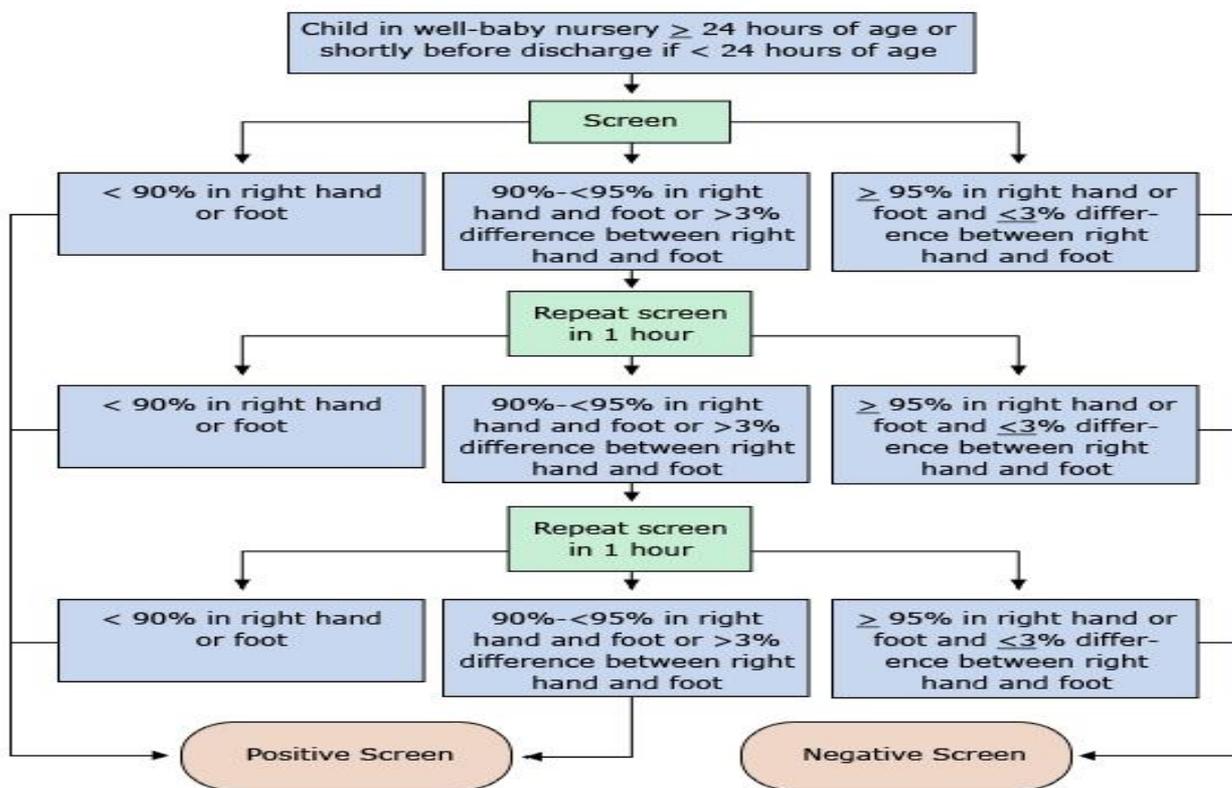
Year	N	Prenatal	Postnatal Before discharge from birth or transfer hospital	Postnatal After discharge home
		%	%	%
2004	158	44.9	38.0	17.1
2005	174	45.4	35.6	19.0
2006	164	53.0	35.4	11.6
2007	141	53.9	34.8	11.4
2008	158	55.8	32.6	11.6
2009	141	63.8	25.5	10.6
<b>Total</b>	<b>916</b>	<b>52.4</b>	<b>33.8</b>	<b>13.8</b>
		<b>(N=480)</b>	<b>(N=310)</b>	<b>(N=126)</b>

- Convened a work group to advise the Department (2012)
- Participants represented:
  - All three levels of perinatal hospital care
  - Cardiology
  - Newborn nursery and NICU (Nursing, Pediatrics and Neonatology)
  - Obstetrics
  - Professional organizations (AAP and AHA)
  - Birth defect researchers
  - Newborn screening program

- Circular letter #**13-5-590** issued 5/10/13 (clarified 6/21/13)
- Recommended maternity facilities incorporate pulse oximetry screening for CCHD into the routine assessment of newborns using guidelines recommended by the Secretary of HHS Advisory Committee
  - Requested aggregate quarterly data reporting
  - Also provided:
    - CCHD/Pulse Oximetry Screening and Reporting Factsheet
    - Quarterly Aggregate Data Reporting Form
    - CCHD Pulse Oximetry Screening Algorithm

# Critical Congenital Heart Disease Pulse Oximetry Screening Algorithm

Child in well-newborn or special care nursery for whom discharge is expected in the first week of life should be screened at 24-48 hours of age or shortly before discharge if <24 hours of age



# CCHD Initial Aggregate Reporting Results

- **16,471** infants reported screened between 7/1/13 and 6/30/14
- Represents **22.5%** of live births

**21/47 (45%)** of hospitals have reported at least once:

- 1/9 (11%) Level 3 hospitals
- 8/22 (36%) Level 2 hospitals
- 12/16 (75%) Level 1 hospitals

- Two true positive CCHD cases were identified with pulse oximetry screening between May 2013 – June 2014 (screening recommendation issued May 2013)
- Neither of these positive screens was reported to DPH, but rather were identified during Birth Defects Monitoring Program (BDMP) medical record abstraction of ascertained CCHD cases
- Both cases were diagnosed with CCHDs that are on the list of secondary screening target defects

On March 6, 2014, Governor Patrick signed into law Chapter 42 of the Acts of 2014, which adds a new section 110C to Chapter 111 of the Massachusetts General Laws.

Among its provisions is the requirement that on or before January 1, 2015, hospitals with maternal and newborn services and birth centers adopt protocols for pulse oximetry screening for congenital heart defects on all newborns prior to discharge, in accordance with Department regulations.

- Other provisions in chapter 42 direct DPH to develop regulations in consultation with the Perinatal Advisory Committee (“PAC”) and to consider other tests that may be at least as accurate, widely available, and cost effective as pulse oximetry.
- DPH staff met with the PAC at its meeting on April 3, 2014. PAC members suggested that the term “critical congenital heart disease” be used instead of the statutory term “congenital heart defects” as it is consistent with nationally accepted terminology.

- The proposed CCHD amendments to the two relevant sections of the hospital regulation, 105 CMR 130.601 and 130.616, and the birth center regulation in its entirety, 105 CMR 142.000, were approved by the Perinatal Advisory Committee.
- The CCHD amendments will be available for public comment in time for filing the final amendments with the Secretary of the Commonwealth before the statutory deadline of January 1, 2015.

Amendment language adds a definition and statutory mandate for CCHD:

- Critical Congenital Heart Disease means a group of defects that cause severe and life-threatening symptoms and require intervention within the first days or first year of life.
- Each birth center shall adopt protocols for screening newborns for critical congenital heart disease with pulse oximetry or another test approved by the Department in accordance with Department guidelines. Such protocols shall provide that the screening shall not be performed if the parent or guardian of the newborn infant objects to the screening based upon the sincerely held religious beliefs of the parent or guardian.
- Several minor technical changes are also included

## Pulse Oximetry

- A readily available, noninvasive, and painless technology that measures the percentage of oxygen saturation of hemoglobin in arterial blood.
- Infants with heart or lung problems may have lower readings.
- Can detect hypoxemia even without obvious cyanosis (appearance of a blue or purple coloration of the skin or mucous membranes due to the tissues near the skin surface having low oxygen saturation).

# Pulse Oximetry

- ♥ Pulse oximetry measures the amount of oxygen in the blood
- ♥ Non-invasive and painless test





Pulse oximetry screening does not replace a complete history and physical examination, which sometimes can detect a CCHD before the development of low levels of oxygen in the blood.

Pulse oximetry screening, therefore, should be used along with the physical examination.

An infant may require transfer for an echocardiogram, cardiology consultation or other intervention based on pulse oximetry, physical examination or both. 21

Pulse oximetry screening has some limitations:

- Increase in false positive readings when screening is done before 24 hours of age.
- During crying, feeding, and sleep, healthy newborns can have periodic or sustained desaturations below 95%
- Other known causes of false positive pulse oximetry readings include inappropriate probe placement, partial detachment and presence of other conditions.

Mahle, W.T., Newburger, J.W., Matherne, G.P., Smith, F.C., Hoke, T.R., Koppel, R., ... Grosse, S.D. (2009). Role of pulse oximetry in examining newborns for congenital heart disease: A scientific statement from the AHA and AAP. *Pediatrics*, 124(2), 823–836

Kemper, AR et al. (2011) “Strategies for implementing screening for critical congenital heart disease.” *Pediatrics*  
doi:10.1542/peds.2011-1317.

Recent studies have shown that the cost of pulse oximetry screening and follow up can cost as little as \$5 and to take as little as 5 minutes or less of staff time (Kemper et al., 2011).

However, associated costs include: staff time conducting the screening, staff education and training, maintaining equipment, tracking results, follow-up on positive test results; verifying positive screening results, and treatment costs.

## Current DPH Factsheet includes:

- Who should be screened
- Algorithm (how to screen)
- Follow-up for newborns who have positive screens
- Aggregate data to report to DPH

## Areas of Focus:

- Screening technology – compliance with national standards
- Who should do screening
- Staff training
- Data to be recorded at site
- Individual data to report to DPH
- Payment for screening
- Special populations (preterm infants, infants born at home, NICU)
- Educational materials for families

## Example of an Available Resource:

- The Children's National Medical Center has developed an evidence-based Congenital Heart Disease Screening Program Toolkit that includes additional information about pulse oximetry as well as resources for educating and training providers responsible for the screening.
- The toolkit can be obtained **free of charge** at:  
<http://www.childrensnational.org/PulseOx/request-tool-kit.aspx>