

Newborn Screening for Critical Congenital Heart Disease (CCHD)

April 29th, 2015



Webinar Objectives:

- Understand the vital importance of pulse oximetry for newborn infants.
- Identify resources to ensure that pulse oximetry screening is conducted in accordance with the recommended protocol.
- Describe reporting requirements and procedures.

Congenital Heart Defects and Critical Congenital Heart Disease

- 1 in 110 newborns in the United States has a Congenital Heart Defect, approximately 25% of these are Critical.¹
- Critical Congenital Heart Defects are those that require surgery or catheter intervention within the first year of life.
- ~28 babies per year in Montana.

¹ <http://www.cdc.gov/ncbddd/heartdefects/cchd-facts.html>

History

September 2011- CCHD screening is added to the Recommended Uniform Screening Panel for newborns.

October 2011- A work group investigated existing research and published recommendations regarding screening population, protocol, equipment, and follow up.*

*Strategies for Implementing Screening for Critical Congenital Heart Disease; Alex R. Kemper, William T. Mahle, Gerard R. Martin, W. Carl Cooley, Praveen Kumar, W. Robert Morrow, Kellie Kelm, Gail D. Pearson, Jill Glidewell, Scott D.Grosse and R. Rodney Howell *Pediatrics* 2011;128:e1259; originally published online October 10, 2011. <http://pediatrics.aappublications.org/content/early/2011/10/06/peds.2011-1317.full.pdf>

July 2014- Administrative Rules of Montana (37.57.301-37.57.320) updated including the addition of CCHD screening.

Montana Administrative Rules

37.57.301-37.57.321 INFANT SCREENING TESTS AND EYE TREATMENT

37.57.301 DEFINITIONS

37.57.304 NEWBORNS HOSPITALIZED FOR NEONATAL INTENSIVE CARE

37.57.305 NEWBORNS OTHER THAN THOSE HOSPITALIZED FOR NEONATAL INTENSIVE CARE

37.57.306 TRANSFER OF NEWBORN INFANT

37.57.307 INFANT BORN OUTSIDE HEALTH CARE FACILITY

37.57.316 REPORTING SCREENING RESULTS

37.57.320 RESPONSIBILITIES OF REGISTRAR OF BIRTH:
ADMINISTRATOR OF HEALTH CARE FACILITY

www.mtrules.org

37.57.304 NEWBORNS HOSPITALIZED FOR NEONATAL INTENSIVE CARE

.....(4) Hospitals providing neonatal intensive care are responsible for developing and implementing a protocol to ensure a newborn hospitalized for neonatal intensive care receives screening for critical congenital heart disease.

Equipment

National Work Group Recommendations*

Pulse Oximeters must:

- Be approved by the FDA for use in newborns
- Be validated in low-perfusion conditions,
- Have a 2% root, mean-square accuracy
- Be motion tolerant

*Strategies for Implementing Screening for Critical Congenital Heart Disease; Alex R. Kemper, William T. Mahle, Gerard R. Martin, W. Carl Cooley, Praveen Kumar, W. Robert Morrow, Kellie Kelm, Gail D. Pearson, Jill Glidewell, Scott D. Grosse and R. Rodney Howell *Pediatrics* 2011;128;e1259; originally published online October 10, 2011
<http://pediatrics.aappublications.org/content/128/5/e1259.full.pdf>

Montana Administrative Rule 37.57.320

- Be approved by the FDA for use in newborns
- Be motion tolerant

FDA Clearance:

<http://www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfpmn/pmn.cfm>

Key Points

- Parents are educated regarding the importance of screening, the screening procedure, and the meaning of results.
- Screening will not detect every heart defect.
- Screening may detect other conditions such as sepsis.
- Screening is not to replace customary clinical practice, evaluation, or intervention.

Newborn Screening for Critical Congenital Heart Disease

- ☑ Most babies are born healthy.
- ☑ A few babies look healthy but have rare health problems.
- ☑ Montana tests all babies because early detection of these health problems can be life-saving.



What is Critical Congenital Heart Disease?

Critical congenital heart disease (CCHD) occurs when a baby's heart or major blood vessels do not form correctly, causing a defect. There are many different types of heart defects that range from mild to severe. Babies with "critical" heart defects need urgent treatment, which may include medicine or surgery.

Why does my baby need this test?

Most babies are born with healthy hearts, but a few are born with CCHD. Your doctor will examine your baby very carefully; however, some babies with CCHD may not have any symptoms until they are at home. Screening helps to find these babies before they go home.

How will my baby be tested?

A simple test, called pulse oximetry, is used to screen babies for CCHD. Pulse oximetry does not hurt. A sensor that measures the oxygen level in your baby's blood is placed on your baby's right hand and one foot. The screening is done when your baby is 24 hours old or older, is quiet, warm, and awake.

What if the pulse oximetry reading is low?

A healthy baby may have a low oxygen reading. Babies with low oxygen levels **may** have CCHD or they may have an infection or breathing problem. If your baby has a low oxygen reading, the doctor may order other tests to determine why the oxygen level is low. A heart ultrasound ("echo" or "echocardiogram") may be done to look for CCHD. Your baby's doctor will discuss the results of these tests with you.

What if the pulse oximetry reading is normal?

Most babies who pass the pulse oximetry screen will not have CCHD. **It is important to know that screening cannot identify every child with a heart problem.** Parents should watch for the following warning signs:

- Bluish color to the lips or skin
- Grunting
- Fast breathing
- Poor feeding
- Poor weight gain
- Sweating around the forehead- especially during feeding.

If you see any of these signs, contact your baby's doctor right away!

For More Information Contact:

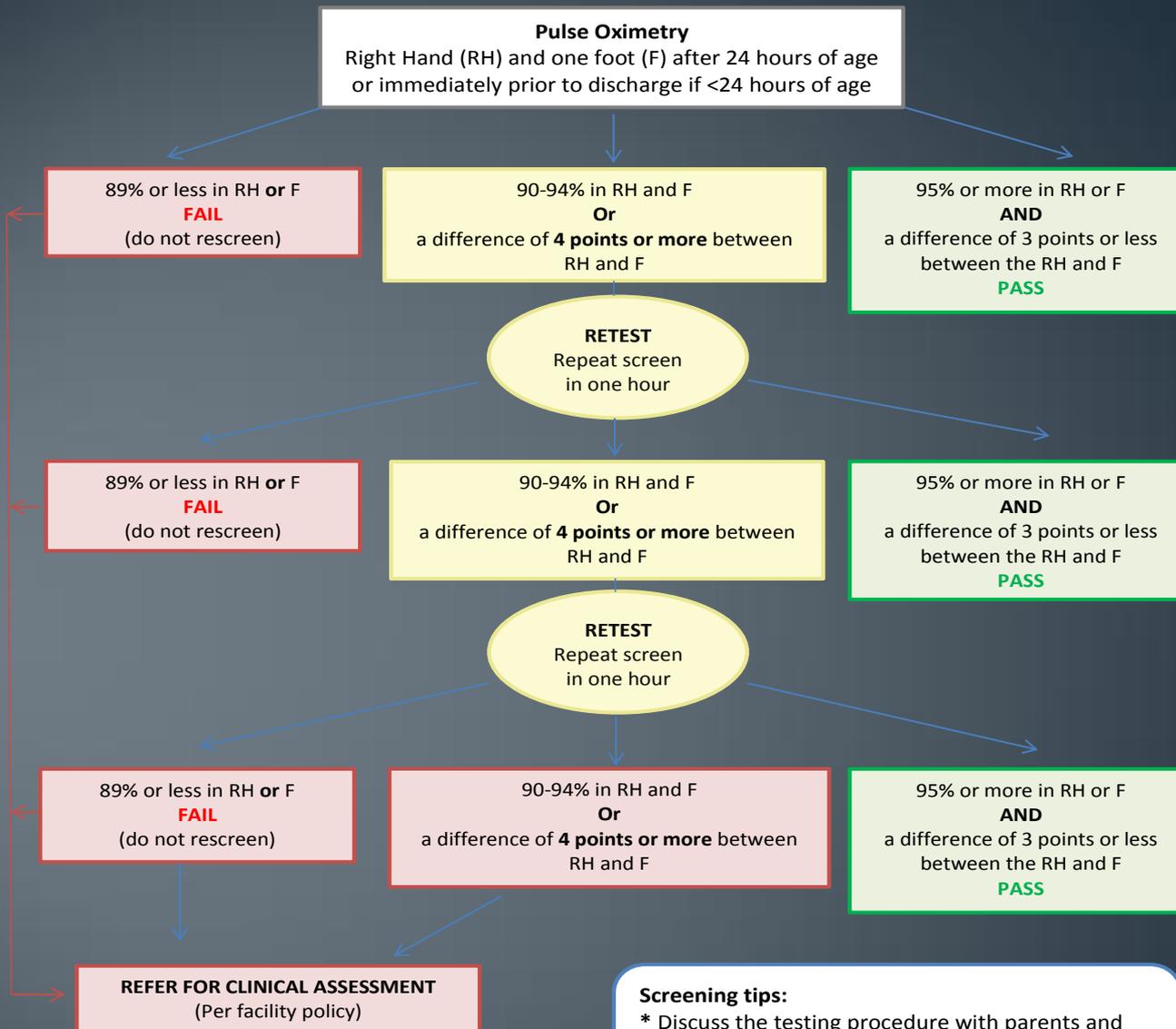
- Your health care provider,
- The Montana Newborn Screening Program at: 1-800-762-9891 or 406-444-3622, or
- Children's Special Health Services online at www.cshs.mt.gov



_____copies of this pamphlet were printed at a cost of _____ per copy for a total cost of \$-----.

Key Points-Continued

- Optimal timing is when baby is 24 - 48 hours of age. Infants older than 48 hours of age should not be excluded. Infants under 24 hours of age are more likely to receive a false positive result.
- Infant is on room air, calm, warm, and awake.
- Screening is performed on both the right hand and either foot.



Screening tips:

- * Discuss the testing procedure with parents and answer questions.
- * Baby should be calm, warm, and awake.
- * Sensor application sites should be clean and dry.



Pulse Oximetry Screening for Critical Congenital Heart Disease (CCHD) Quick Reference Guide

Screening Basics:

- ♥ Screen all babies in well baby or intermediate care nurseries per the recommended algorithm*. Babies hospitalized for neonatal intensive care are required to be screened per facility policy.
- ♥ Optimal timing for screening is between 24-48 hours of age. However, infants older than 48 hours of age who have not previously been screened should not be excluded. Infants under 24 hours of age are more likely than those over 24 hours of age to receive a false positive result.
- ♥ The infant should be on room air, calm, warm, and awake.
- ♥ The pulse oximeter used must be approved by the FDA for use in newborns and be motion tolerant.
- ♥ Education regarding the importance of screening, the screening procedure, and the meaning of results must be provided to the infant's family.
- ♥ Screening is not a replacement for careful clinical practice, evaluation, or intervention and will not detect every baby with a heart defect.
- ♥ The full screening protocol, administrative rules, and educational resources can be accessed at: <http://dphhs.mt.gov/publichealth/cshs/NewbornScreeningPrograms/CriticalCongenitalHeartDiseaseScreening/CCHDProviderResources>

*see back of this sheet or go to the link above

Reporting Basics:

- ♥ Report results to DPHHS in conjunction with reporting of hearing screening results.
- ♥ If pulse oximetry results are out of range, fax a Failed Screen Report Form to 406-444-2750.
- ♥ If a parent declines screening, a signed waiver must be faxed to 406-444-2750.
- ♥ Reporting tutorials and forms can be found at: <http://dphhs.mt.gov/publichealth/cshs/NewbornScreeningPrograms/CriticalCongenitalHeartDiseaseScreening/CCHDProviderResources>

Out of Range Screen Follow Up:

- ♥ Out of range screens should be referred for a complete physical examination by the newborn's primary care provider.
- ♥ It is recommended that the primary care provider seek consultation with a Neonatologist or Pediatric Cardiologist to determine what additional testing may be indicated.
- ♥ Conditions other than CCHD which cause hypoxemia may be detected.



Critical Congenital Heart Disease Screening Table

| Right Hand | Either Foot | | | | | | | | | | | <90 |
|------------|-------------|-----|-----|-----|-----|-----|-----|-----|-----|-----|-----|-----|
| 100 | 100 | 99 | 98 | 97 | 96 | 95 | 94 | 93 | 92 | 91 | 90 | <90 |
| 99 | 100 | 99 | 98 | 97 | 96 | 95 | 94 | 93 | 92 | 91 | 90 | <90 |
| 98 | 100 | 99 | 98 | 97 | 96 | 95 | 94 | 93 | 92 | 91 | 90 | <90 |
| 97 | 100 | 99 | 98 | 97 | 96 | 95 | 94 | 93 | 92 | 91 | 90 | <90 |
| 96 | 100 | 99 | 98 | 97 | 96 | 95 | 94 | 93 | 92 | 91 | 90 | <90 |
| 95 | 100 | 99 | 98 | 97 | 96 | 95 | 94 | 93 | 92 | 91 | 90 | <90 |
| 94 | 100 | 99 | 98 | 97 | 96 | 95 | 94 | 93 | 92 | 91 | 90 | <90 |
| 93 | 100 | 99 | 98 | 97 | 96 | 95 | 94 | 93 | 92 | 91 | 90 | <90 |
| 92 | 100 | 99 | 98 | 97 | 96 | 95 | 94 | 93 | 92 | 91 | 90 | <90 |
| 91 | 100 | 99 | 98 | 97 | 96 | 95 | 94 | 93 | 92 | 91 | 90 | <90 |
| 90 | 100 | 99 | 98 | 97 | 96 | 95 | 94 | 93 | 92 | 91 | 90 | <90 |
| <90 | <90 | <90 | <90 | <90 | <90 | <90 | <90 | <90 | <90 | <90 | <90 | <90 |

PASS 85% or higher in right hand or either foot AND difference of 3% or less between right hand and either foot.

RESCREEN 90-94% in right hand and either foot or difference of 4% or more between right hand and either foot.

Rescreen in 1 hour. If third screening is still in the yellow, it is a fail and should be reported to the MD.

FAIL 89% or lower in right hand or either foot (at any time) OR 3rd Screen: 90-94% in right hand and either foot or difference of 4% or more between right hand and either foot. Failed screening should be reported to the MD.

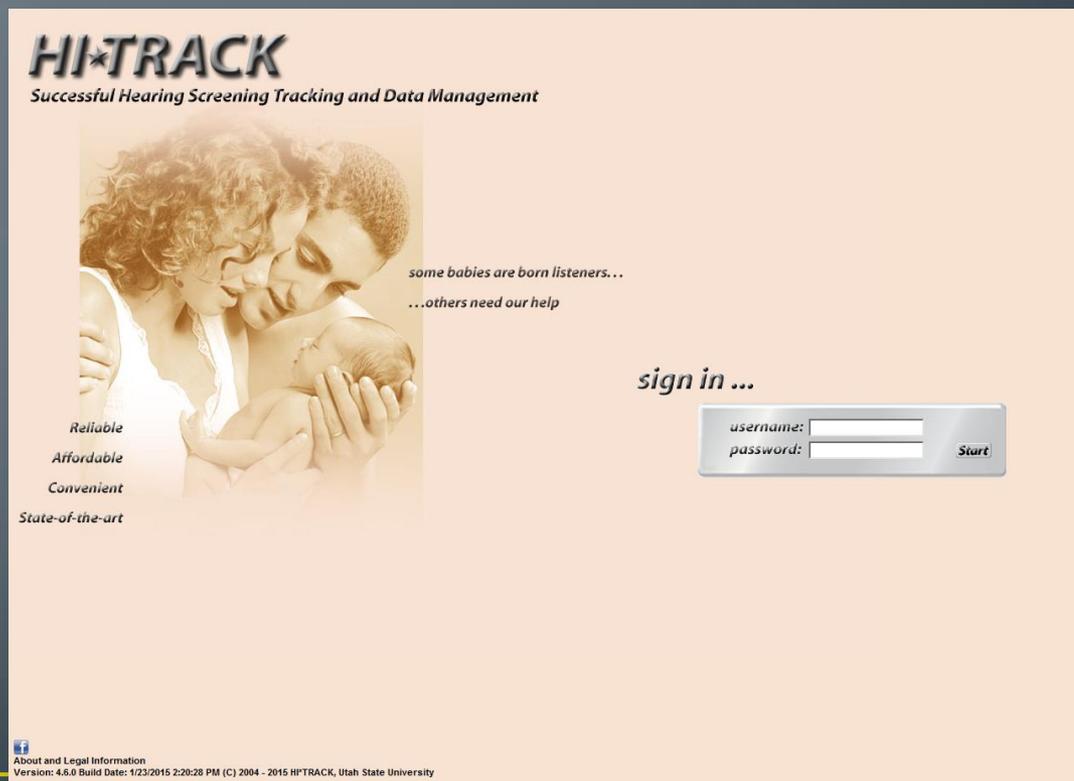
Adapted from *Strategies for Implementing Screening for Critical Congenital Heart Disease*
Kemper A., et al, Pediatrics 2011(128)5: e1259 - 1267.

Out of Range (Failed) Screen Follow Up

- Complete physical evaluation by the baby's primary care provider.
- Have a plan in place *before* this situation occurs.

Reporting Screening Results

- Reported in conjunction with hearing screening results through HiTrack.
- Pass, Fail, Not Screened, Refused, Comments



HI*TRACK
Successful Hearing Screening Tracking and Data Management



some babies are born listeners...
...others need our help

Reliable
Affordable
Convenient
State-of-the-art

sign in ...

username:
password: **Start**

 About and Legal Information
Version: 4.6.0 Build Date: 1/23/2015 2:20:28 PM (C) 2004 - 2015 HPTRACK, Utah State University

Reporting Screening Results-Cont.

- ANY baby that fails also needs the Failed Screen Report Form.
 - <http://dphhs.mt.gov/Portals/85/publichealth/documents/cshs/FailedPulseOximetryScreen.pdf>
 - Include as much information as possible
- A signed waiver of newborn screening for those families declining.

Fax to DPHHS at 406-444-2750

Newborn Pulse Oximetry Screening For Critical Congenital Heart Disease Failed Screen Reporting Form

Date _____ Facility _____ MRN _____

| | | |
|--|--------------|--------------------------------|
| Name (last, first) | DOB | Time of Birth (military) |
| Gestational age (weeks) | Birth Weight | Gender |
| Was a 2 nd trimester ultrasound performed? Yes No Don't Know | | Infant's Primary Care Provider |

| Screening Information | First Pulse Ox Screen | Second Pulse Ox Screen (if indicated) | Third Pulse Ox Screen (if indicated) |
|-----------------------|-----------------------|--|---|
| Right hand | % | % | % |
| Foot | % | % | % |
| Age in hours | Hrs | Hrs | Hrs |

Was an echocardiogram performed? Y N Don't Know
If yes- date _____ Facility _____

Echocardiogram reviewed by: _____

Was telemedicine used to review this echocardiogram? Y N Don't know

Was the patient transferred? Y N
If yes- Where? (Facility name) _____ Date of transfer _____

Findings (please list all diagnoses and include ICD9 codes):

Comments: _____

Person completing form: _____



FAX COMPLETED FORM TO:
Montana Newborn Screening Program
 Fax 406-444-2750
 For questions call 406-444-3622

Resources

- Heart atlas- visuals and descriptions of heart defects
<http://www.wisconsinshine.org/heart-atlas/>
- Video for parents: <http://www.babysfirsttest.org/newborn-screening/conditions/critical-congenital-heart-disease-cchd>
- Video for providers: <http://www.babysfirsttest.org/newborn-screening/educational-resources>
- Montana resources: www.cshs.mt.gov
- National Work Group Recommendations:
<http://pediatrics.aappublications.org/content/early/2011/10/06/peds.2011-1317.full.pdf>

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Pediatrics 2011;128:e1259; originally published online October 10, 2011.

- **Altitude:**
<http://pediatrics.aappublications.org/content/133/3/e561>

Feasibility of Critical Congenital Heart Disease Newborn Screening at Moderate Altitude: Jason Wright, MD^{a,b}, Mary Kohn, MD^b, Susan Niermeyer, MD, MPH, FAAP^{a,b}, and Christopher M. Rausch, MD^{a,b} *Pediatrics* Vol. 133 No. 3 March 1, 2014 pp. e561 -e569 (doi: 10.1542/peds.2013-3284)

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