Critical Congenital Heart Disease (CCHD) Newborn Screening

Georgia Newborn Screening Program
Maternal and Child Health Section
Georgia Department of Public Health

Georgia Department of Public Health

Objectives

- Provide information on newborn screening for critical congenital heart defects using pulse oximetry.

- Cover Georgia's procedures for critical congenital heart disease (CCHD) screening, follow-up for at risk infants, and result reporting.

Critical Congenital Heart Disease (CCHD)

300

Infants sent home each year in the US with undiagnosed CCHD putting them at risk for serious complications.

- Baby's First Test, 2013

CCHD is a group of heart defects that can be life threatening and require medical attention within the first few days or first year of life.

Newborn Screening Goals

1. Early education of providers and families
2. Early identification
3. Early intervention through timely referrals
4. Reduced morbidity and mortality

Why Screen for CCHD?

Newborns with CCHD typically appear normal at birth

Screening identifies CCHD before symptoms are detected

Early identification can result in early intervention & better outcomes

Factors Contributing to Missed Detection:

1. Absence of Murmurs
2. Palpable pulses from presence of PDA
3. Cyanosis (difficult to detect in many newborns, not noticeable for O2 saturation between 80-89 percent.

Early Detection of CCHD: Examination and Screening

- CCHD can be identified as a results of either prenatal ultrasound or postnatal physical exam but is missed in a small percentage of births.
- Detection rates for CCHD:
  - Prenatal ultrasound = ~25-50%
  - Postnatal newborn physical exams = ~25-50%
- Approximately 30% of the remaining infants will be undetected in the immediate newborn period.
- Some of these will present with life threatening symptoms shortly after discharge.

Pulse Oximetry Screening

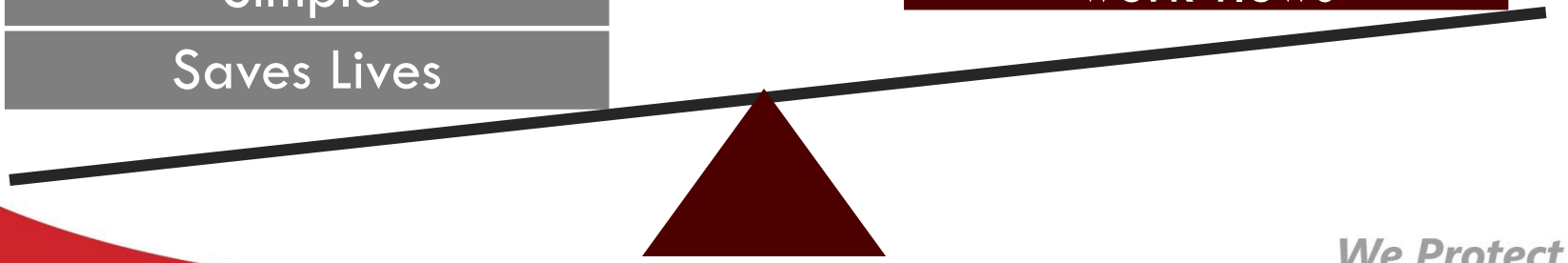
A pulse oximeter is used to measure the percentage of hemoglobin in the blood that is saturated with oxygen.

Strengths

- Inexpensive
- Non-invasive
- Painless
- Quick
- Simple
- Saves Lives

Weaknesses

- Possible false positives and negatives
- Not all types of CHD detectable
- Costly follow-up
- Requires change in work flows



Detectable CCHD Defects Through Screening

Primary Targets

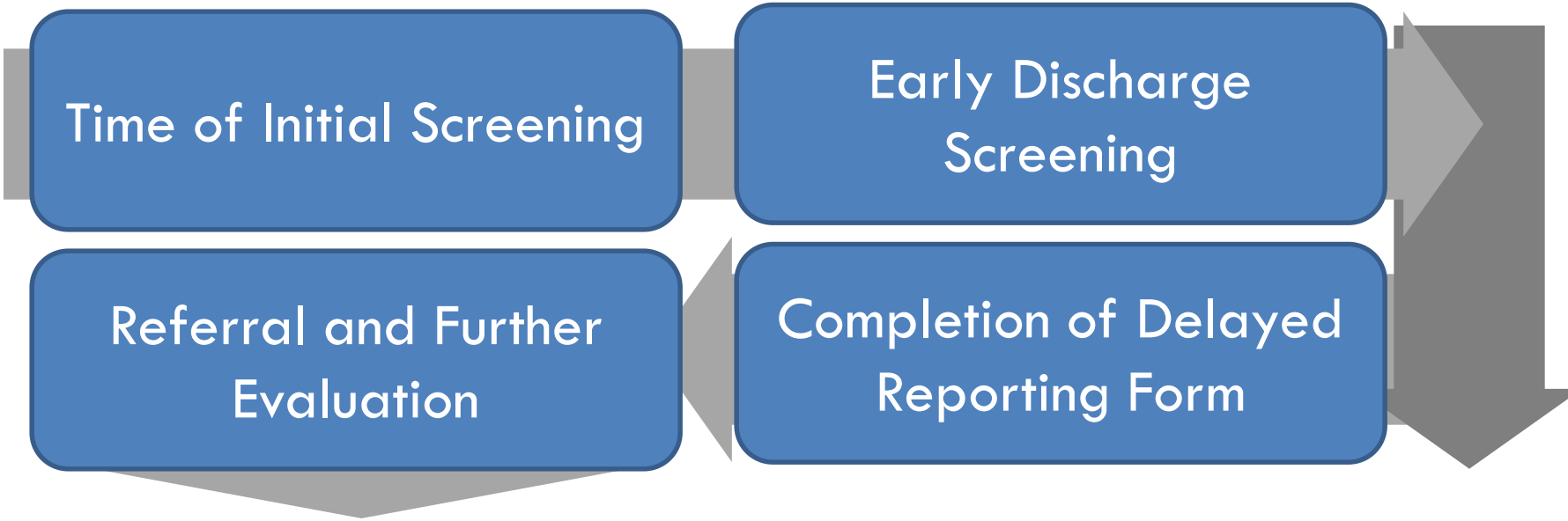
- Hypoplastic left heart syndrome
- Pulmonary atresia (with intact septum)
- Tetralogy of fallot
- Tricuspid Atresia
- Total anomalous pulmonary venous connection
- Truncus Arteriosus
- Transposition of the great vessels

Secondary Targets

- Single Ventricle
- Coarctation
- Interrupted aortic arch
- Ebstein Anomaly
- Double-outlet right ventricle
- Aortic Atresia
- Hypoplasia of aortic arch

Hospitals' Role in CCHD Screening

Georgia law requires hospitals to perform a pulse oximetry screen on all live births before discharge. Key implementation considerations include:



Equipment

- Motion-tolerant and report functional oxygen saturation
- Validated in low-perfusion conditions
- Cleared by the FDA for use in newborns
- 2% root, mean-square accuracy
- Calibrated regularly based on manufacturer guidelines
- Used with infant disposable or reusable pulse oximeter probes

Why No Adult Oximeters?

Conventional Adult Oximeter

- Does not have heart rate (HR) display with normal correlation for newborns
- Does not have stable pleth wave with motion artifact

Adult Probe

- Clips too large for testing newborns
- Gives inaccurate readings

Factors Affecting Pulse Oximetry Interpretation

- Translucency and blood flow where the measurement is taken
- Extreme low body temperatures
- Blood volume deficiency
- Exposure to strong external light while taking measurement

Pulse Oximeter Probe Placement

Place the photo-detector portion of the probe on the fleshly portion of the outside of the infant's right hand or foot

1

Right Hand Application Site

Place the light emitter portion of the probe on the top of the hand or foot.

2

Place the photo-detector directly opposite of light emitter, on the bottom of the hand or foot.

3

Foot Application Site

Additional Screening Tips

- ✓ Clean reusable probes with recommended disinfectant, as dirty probes can decrease accuracy of the reading.
- ✓ Use disposable wraps to secure sensor to right hand or foot, with no gaps between probe and infant's skin.
- ✓ Allow the pulse-ox to remain in place for at least 30 seconds before obtaining a reading.
- ✓ Ensure pleth wave on the oximeter (arterial pulse) is stable at the monitoring site and is without motion artifact.

Performing CCHD Screening

Time of Screening
(per AAP Guidelines)

- Greater than 24 hours of age
- If discharged before 24 hours old, screen as close to 24 hours as possible

Environment for testing

- Quiet, comforting
- Avoid noise and harsh lights
- Babies should be warm, quiet, alert, not crying or moving

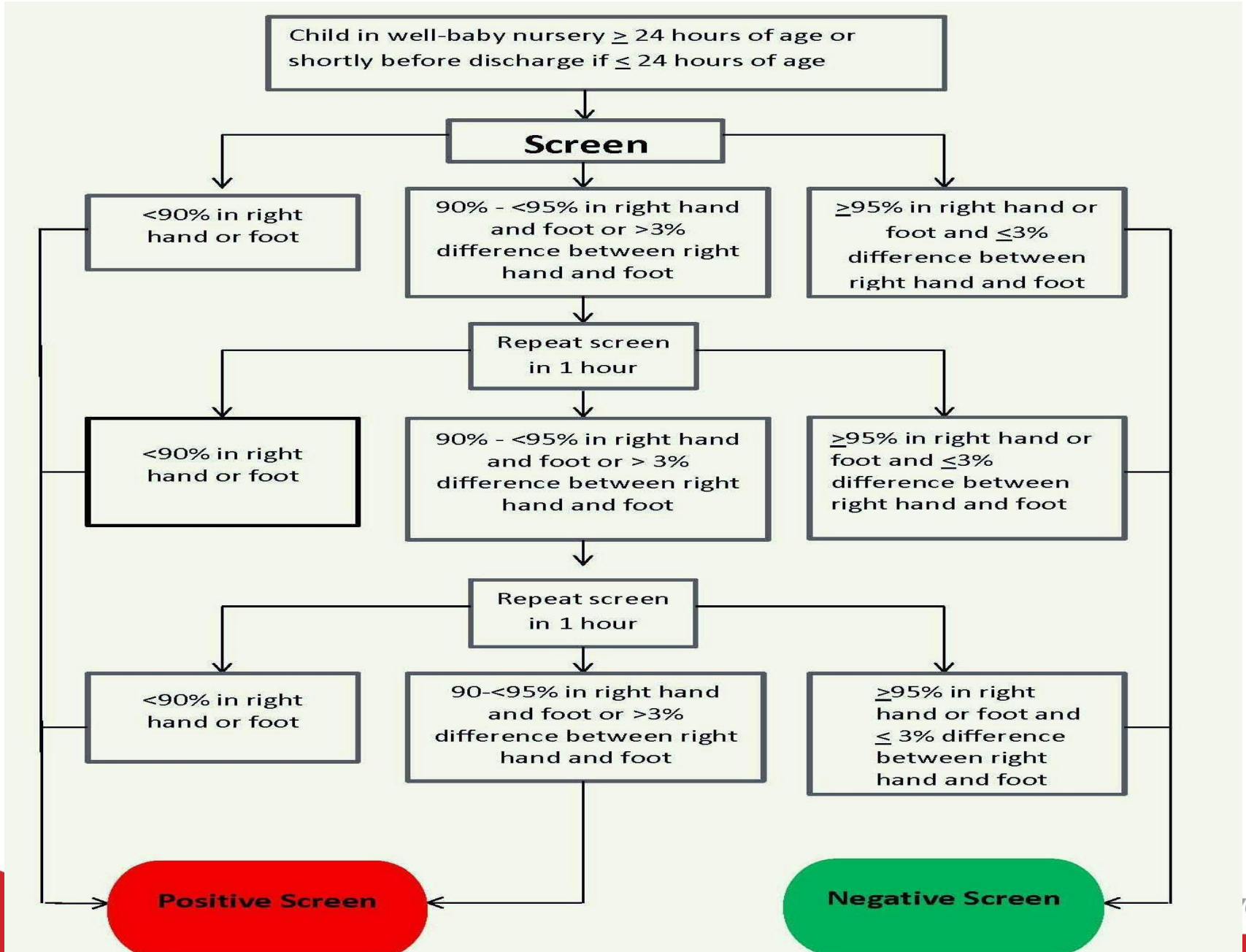
Assess for:

- Hypothermia
- Presence/adequacy of pulses
- Phototherapy
- Presence of dried blood, foot print ink, betadine solution

Other Screening Tests

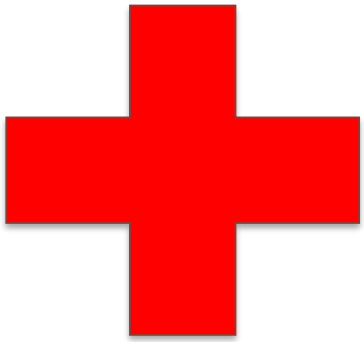
- Perform prior to painful heel-stick procedures
- Document results on NBS card
- For delayed reporting, complete delayed reporting form. Fax to NBS Program.

AAP-endorsed CCHD Screening Algorithm



Using the Screening Tool

A positive screen = *"fail"*



1. Any oxygen saturation value <90% (right hand or either foot)
2. Oxygen saturation value ≤95% in both extremities on 3 different measurements, each separated by one hour ***or***
3. A >3% difference in oxygen saturation between the right hand and foot on 3 measurements each separated by one hour.

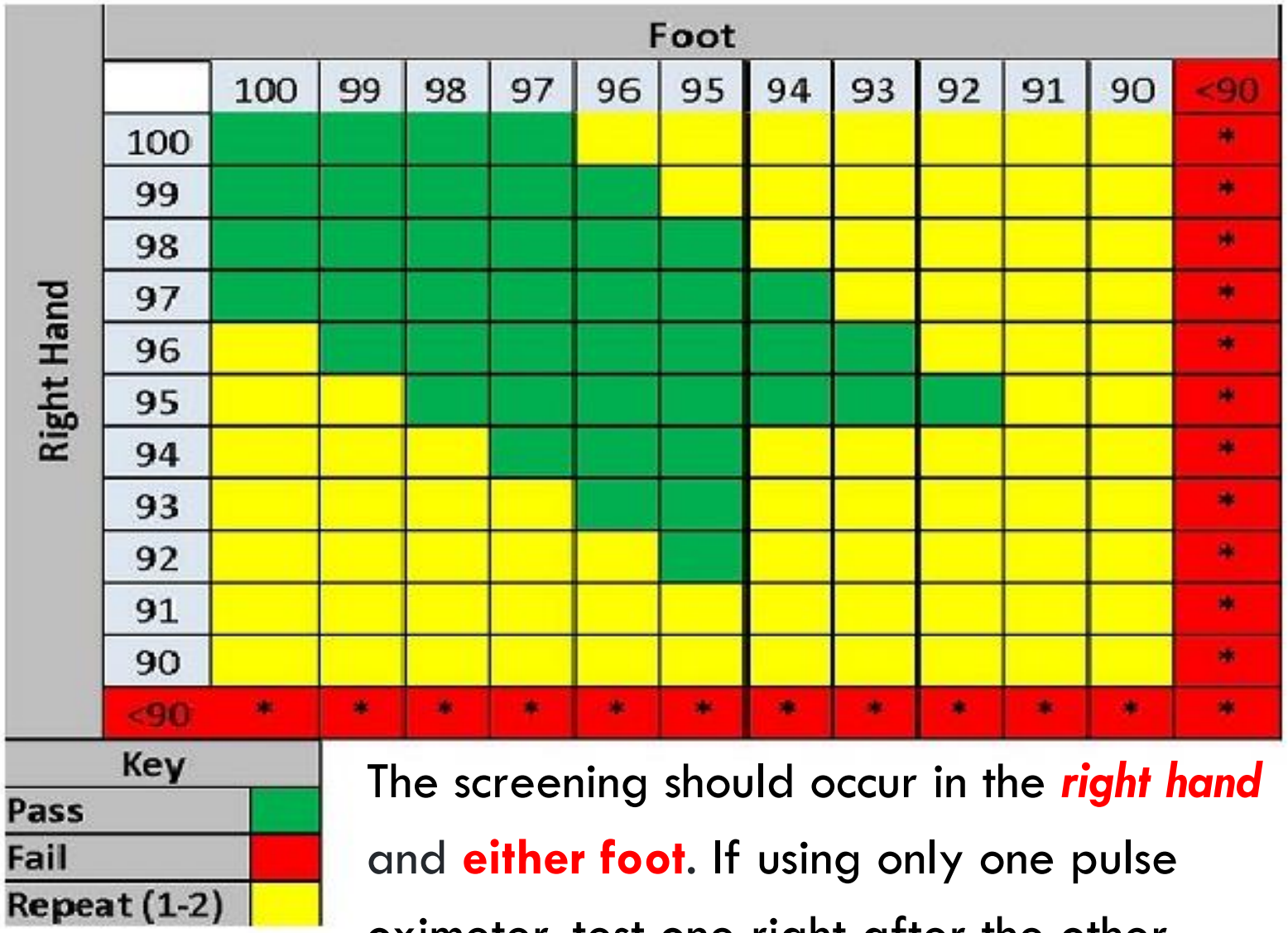
A negative screen = *"pass"*

Any oxygen saturation value that is ≥ 95% in either extremity ***and*** ≤ 3% difference in oxygen saturation between the upper and lower extremity.

Calculating Pulse Oximetry Values

- **Use of calculator**
- **AAP/CHOA CCHD APP:** http://pulseoxtool.com/index.php
- **Use of Pulse Oximetry Grid**
  1. The combined values **from the right hand and either foot must be used** in order to identify a pass, fail or requires re-screening:
  2. The pulse oximeter values for the right hand are located in the column on the left side of the grid.
  3. The rest of the grid contains the pulse oximeter values for either foot.
  4. Obtain values for right hand and either foot
  5. If value falls in **"green"** section, no action is needed.
  6. If value falls in "yellow" or **"red"** section, **Action is needed.** Refer to the AAP – endorsed CCHD screening algorithm to determine the action required.

Pulse Oximetry Calculation Tool

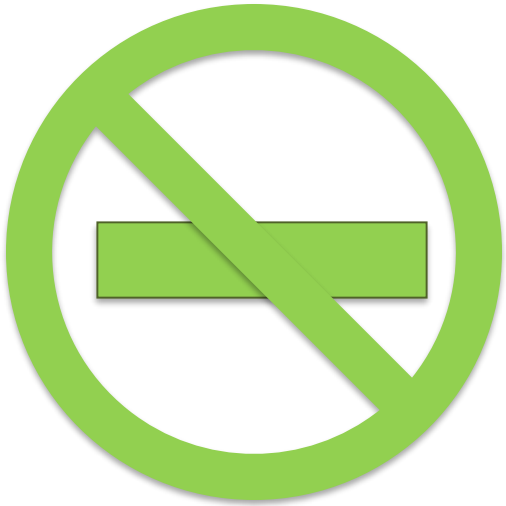


The screening should occur in the ***right hand*** and **either foot**. If using only one pulse oximeter, test one right after the other.

Screening Results

Factors that can lead to false positive results:

- Lung disease
- Sepsis
- Screened too early



Factors that can lead to false negative results:

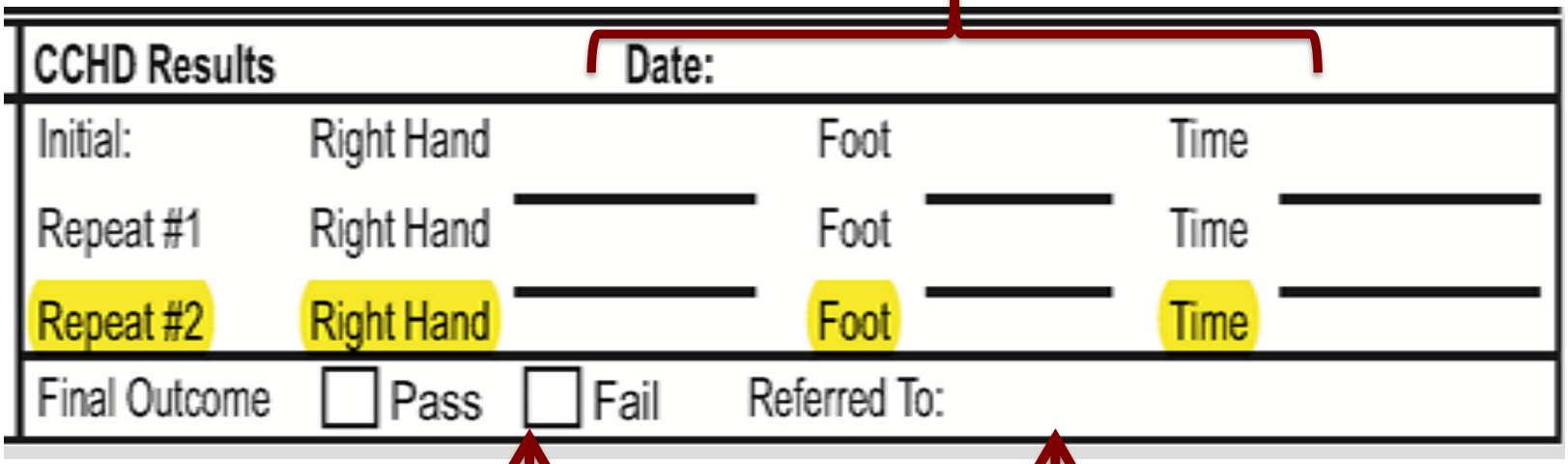
- Not all CHD defects detected through pulse oximetry screening

Documenting Results on NBS Card

All pulse oximetry screening results must be entered on the card for all screens done.

CCHD screening section:

Enter results for right hand, foot and time of screening in the correct spaces



Check the box for final outcome (pass or fail)

Enter either hospital or cardiologist name in the referred to box for follow-up

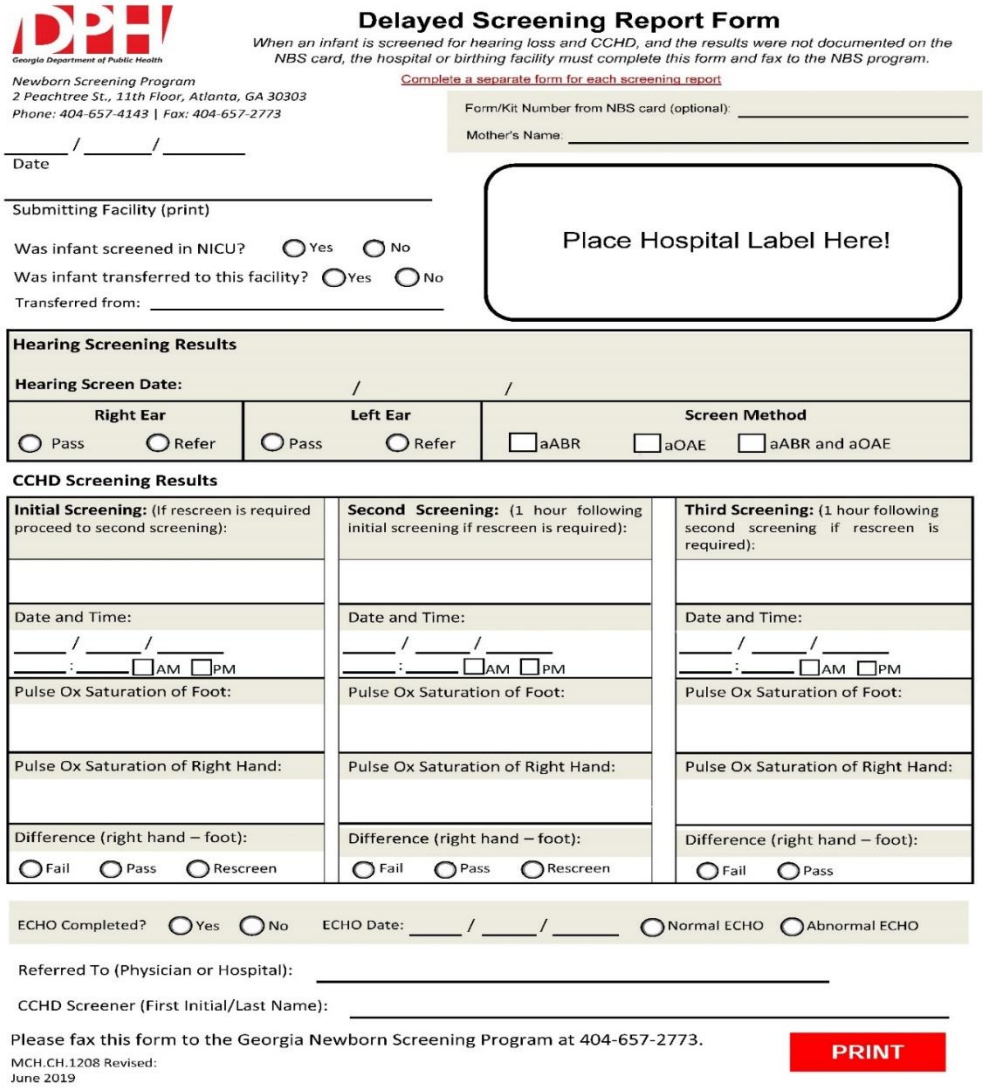
Delayed Reporting

If CCHD screening results **are not** available and the bloodspot is ready to be shipped.

What to Do?

- Ship the bloodspot specimen once it is dried.
- Complete the "Delayed CCHD Screening Form" once CCHD screening is performed.
- Fax a copy of the delayed CCHD screening form to: Newborn Screening Program at 404-657-2773.
- Place original copy in medical record.

Delayed CCHD Reporting Form



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Delayed Screening Report Form

When an infant is screened for hearing loss and CCHD, and the results were not documented on the NBS card, the hospital or birthing facility must complete this form and fax to the NBS program.

Complete a separate form for each screening report

Newborn Screening Program
2 Peachtree St., 11th Floor, Atlanta, GA 30303
Phone: 404-657-4143 | Fax: 404-657-2773

Form/Kit Number from NBS card (optional): _____

Mother's Name: _____

_____ / _____ / _____
Date

_____
Submitting Facility (print)

Was infant screened in NICU? ◯ Yes ◯ No

Was infant transferred to this facility? ◯ Yes ◯ No

Transferred from: _____

Place Hospital Label Here!

Hearing Screening Results

Hearing Screen Date: / /

Right Ear	Left Ear	Screen Method
◯ Pass ◯ Refer	◯ Pass ◯ Refer	☐ aABR ☐ aOAE ☐ aABR and aOAE

CCHD Screening Results

Initial Screening: (If rescreen is required proceed to second screening):	**Second Screening:** (1 hour following initial screening if rescreen is required):	**Third Screening:** (1 hour following second screening if rescreen is required):
Date and Time: _____ / _____ / _____ _____ : _____ ☐ AM ☐ PM	Date and Time: _____ / _____ / _____ _____ : _____ ☐ AM ☐ PM	Date and Time: _____ / _____ / _____ _____ : _____ ☐ AM ☐ PM
Pulse Ox Saturation of Foot:	Pulse Ox Saturation of Foot:	Pulse Ox Saturation of Foot:
Pulse Ox Saturation of Right Hand:	Pulse Ox Saturation of Right Hand:	Pulse Ox Saturation of Right Hand:
Difference (right hand – foot): ◯ Fail ◯ Pass ◯ Rescreen	Difference (right hand – foot): ◯ Fail ◯ Pass ◯ Rescreen	Difference (right hand – foot): ◯ Fail ◯ Pass

ECHO Completed? ◯ Yes ◯ No ECHO Date: _____ / _____ / _____ ◯ Normal ECHO ◯ Abnormal ECHO

Referred To (Physician or Hospital): _____

CCHD Screener (First Initial/Last Name): _____

Please fax this form to the Georgia Newborn Screening Program at 404-657-2773.

PRINT

MCH.CH.1208 Revised:
June 2019

Pediatric Echocardiology and Referral Resources

Children's Healthcare of Atlanta, Sibley Heart Center:
404-256-2593

Georgia Pediatric Cardiology: 678-289-1988

Pediatric Cardiology Services: 770-995-6684

Savannah Children's Heart Center: 912-988-5050

GRU Pediatric Cardiology Services: 706-721-8522

Resources

- Baby's First Test
- Heart Smart Videos
- Centers for Disease Control and Prevention
- Children's National Medical Center
- Mended Hearts

References

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Acknowledgements

The Georgia Newborn Screening Program thanks the following organizations for their contributions and support in creating this document and working together to help save the lives of Georgia's youngest residents:

- American Academy of Pediatrics, Georgia Chapter
- American Heart Association of Georgia
- Academy of Family Practice, Georgia Chapter
- Children's National Medical Center, Washington, DC
- Baby's First Test
- Georgia Hospital Association
- March of Dimes of Georgia
- Minnesota Department of Public Health
- Sibley Heart Center