



Critical Congenital Heart Disease Screening in Tennessee (CCHD)

- ❖ Tennessee State Legislature passed a bill directing the Genetics Advisory Committee to develop a screening program using pulse oximetry to identify Critical Congenital Heart Disease (CCHD) on or before January, 1st 2013.

- ❖ Tennessee rules and regulations state that all administrators of hospitals and birthing facilities and medical personnel from non-birthing facilities will be responsible for ensuring all newborns in their facility are screened for metabolic diseases and hearing loss.
- ❖ The addition of pulse oximetry screening to rules and regulations is in process.



NBS

Newborn Screening Program

National Recommendation

❖ Screening recommended by the:

- United States Health and Human Services (HHS) Secretary's Advisory Committee
- Supported by:
 - American Academy of Pediatrics
 - American Heart Association
 - American College of Cardiology



Current Uniform Screening Panel

- ❖ Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (SACHDNC) recommends **all** states screen for 31 core conditions.
 - With the addition of CCHD, Tennessee screens for 30* of the 31 core conditions.
 - CCHD: Only point-of-care type of screening

*Severe combined immunodeficiency (SCID) is the only condition Tennessee is not screening for but we are currently evaluating testing methods.

Background on CCHD

- ❖ Congenital heart disease (CHD) is the most common birth defect – about 8 out of 1,000 births for all types of CHD, including both mild and severe atrial septal defects (ASDs) and ventral septal defects (VSDs).
- ❖ CHD accounts for nearly 1 in 4 infant deaths due to birth defects.
- ❖ **Critical Congenital Heart Disease (CCHD)** (7 well defined types – “blue babies”) accounts for 11-12 out of 10,000 births.
- ❖ In Tennessee, with around 80,000 to 90,000 births each year, we may see about 85 to 95 cases of **CCHD** each year.



CCHD Meets Standards for Universal Screening

- ❖ Signs of CCHD may not be evident (easily detected through physical exam) prior to infant being sent home from hospital.
- ❖ CCHD lesions can be detected during either the prenatal or postnatal period (a cost-effective screening method exists).
- ❖ Early detection of CCHD can potentially improve the prognosis and decrease the mortality and morbidity rate of affected infants (interventions are available and are effective).
- ❖ Survivors who present late are at greater risk for neurologic injury and subsequent developmental delay.

CCHD Meets Standards for Universal Screening

- ❖ Pulse oximetry has been studied and shown to be successful in detecting some forms of CCHD in the newborn nursery prior to discharge.
 - Physical exam (PE=current method) is shown to detect up to 50% of cases
 - Add Pulse oximetry to PE, the detection rate increases to up to 80%

Pulse Ox Targets 7 Lesions

- ❖ Screening targets 7 lesions that benefit from early detection and intervention (i.e. surgical, catheterization, or pharmacologic interventions).
- ❖ Seven specific lesions as primary targets for screening are:
 1. Hypoplastic left heart syndrome (HLHS)
 2. Tricuspid atresia
 3. Pulmonary atresia
 4. Tetralogy of Fallot (TOF)
 5. Total anomalous pulmonary venous return (TAPVR)
 6. Transposition of the great arteries (TGA)
 7. Truncus arteriosus

Who should be screened?

- ❖ Newborns who are at least 24 hours of age;
 - If early discharge is planned, screening should occur as late as possible prior to discharge
- ❖ Infants in special care nurseries (intermediate care and neonatal intensive care, etc.) should be screened at 24-48 hours of age or when medically appropriate after 24 hrs of age.

Pulse oximetry as screening method:

- A pulse oximeter is used to measure the percentage of hemoglobin in the blood that is saturated with oxygen.
- Non-invasive and **painless**
- Accurate with newer generation oximeters
- Fast (<2 min) and reliable
- Inexpensive

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

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

- ❖ The CCHD screen is a **point-of-care test**.
- ❖ Point-of-care testing refers to those tests administered outside of a laboratory but close to the site of direct delivery of medical care for a patient.
- ❖ *Interpretation* of test results and decisions around *intervention* will take place **at the hospitals and birthing facilities, at the time of the test.**

- ❖ Each birthing facility will be responsible for selecting and securing pulse oximeter equipment for screening newborns for CCHD, if appropriate equipment is not already available.
- ❖ Such equipment **must be compliant** with national standards:
 - Cleared by the FDA for use in newborns
 - Calibrated regularly based on manufacturer guidelines

Pulse Ox Probe Placement

- ❖ Select application site on the outside, fleshy area of the infant's hand or foot.
- ❖ Place the photodetector portion of the probe on the fleshy portion of the outside of the infant's hand or foot.



Right Hand Application



Foot Application

Pulse Ox Probe Placement

- ❖ Place the light emitter portion of the probe on the top of the hand or foot. Place the photodetector directly opposite of light emitter, on the bottom of the hand or foot.



- ❖ Remember: The photodetector and emitter must be directly opposite each other in order to obtain an accurate reading.
- ❖ Secure the probe to the infant's hand or foot using the adhesive or foam tape recommended by the vendor. It is not recommended to use tape to secure probe placement.

Screening Hints

- ❖ Calm the infant prior to completing exam.
- ❖ Movement, shivering and crying can affect the accuracy of the pulse ox reading.
- ❖ Make sure right hand or either foot is clean and dry prior to testing.
- ❖ Clean probe with recommended disinfectant between each infant (dirty probes can decrease reading accuracy and spread infection).
- ❖ Substances with dark pigmentation (such as dried blood) can affect the pulse ox reading.
- ❖ The best sites for performing pulse ox on infants are around the palm and the foot. An infant pulse ox probe (not an adult pulse ox clip) should always be used for infants.

- ❖ Should be performed after the infant turns 24 hours of age, or when *medically appropriate* if the infant was born prematurely or sick in the NICU. If early discharge is planned, screening should occur as late as possible.
- ❖ Recommended that pulse oximetry screening be done in conjunction with other standard-of-care newborn screening that requires the infant be at least 24 hours of age, such as metabolic or hearing screening.

- ❖ The initial screen will be done in the foot only (left or right):
 - If the oxygen saturation in the foot is $<90\%$ the newborn fails the screen.
 - An oxygen saturation from the foot of 97-100% is an immediate pass and does not require a right hand saturation.
 - If the oxygen saturation in the foot is 90-96% the right hand will also be screened.

Screening Protocol

Guidance for Sick Infants/NICU Infants

- ❖ If the special care nursery infant...
 - 1) ...never required oxygen during their stay → proceed with screening protocol.
 - 2) ...required oxygen but has been weaned to room air → proceed with screening protocol at least 24 hours after weaning to room air.
- ❖ If infant is being discharged home on oxygen, obtain an echocardiogram (if one was not obtained during their neonatal course).
- ❖ In all cases (except infants going home on oxygen who should be echo'd), screening should occur prior to discharge from the hospital.

❖ To pass the screening:

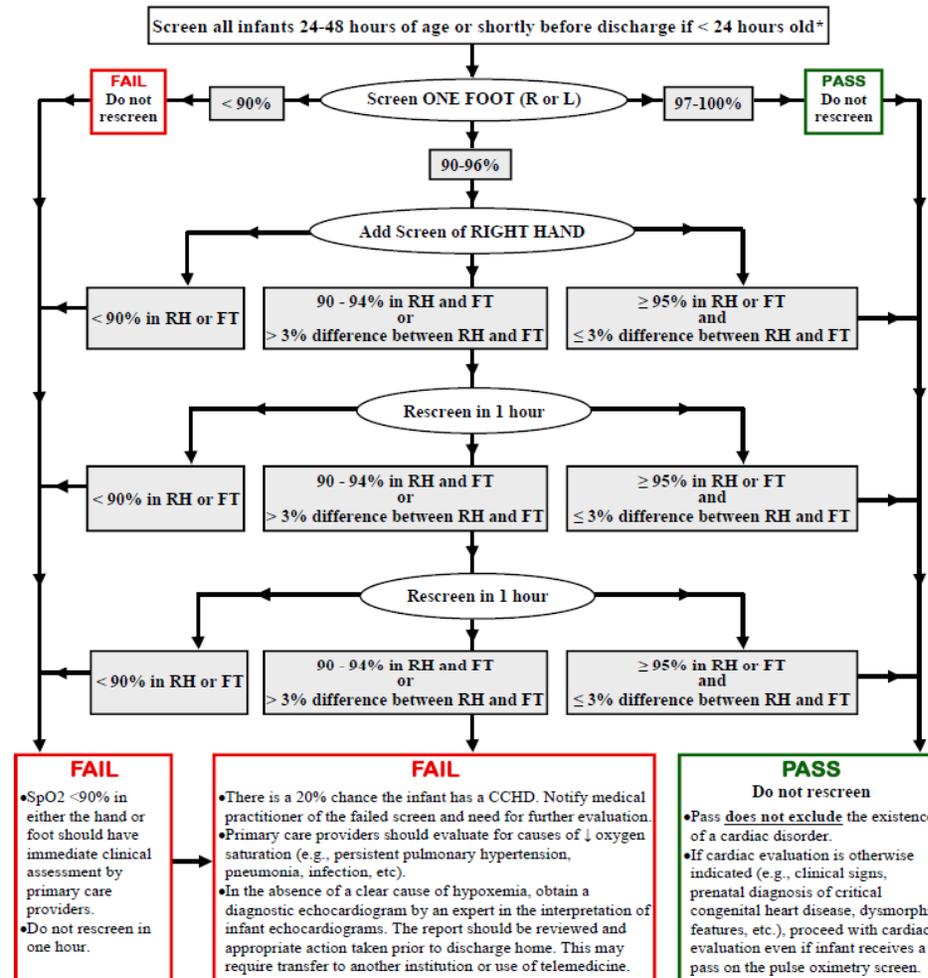
- An oxygen saturation from the foot of 97-100% is an immediate pass and does not require a right hand saturation.
- Oxygen saturation of $\geq 95\%$ on either the right hand or foot (left or right) and a difference of $\leq 3\%$ between the right hand and either foot.

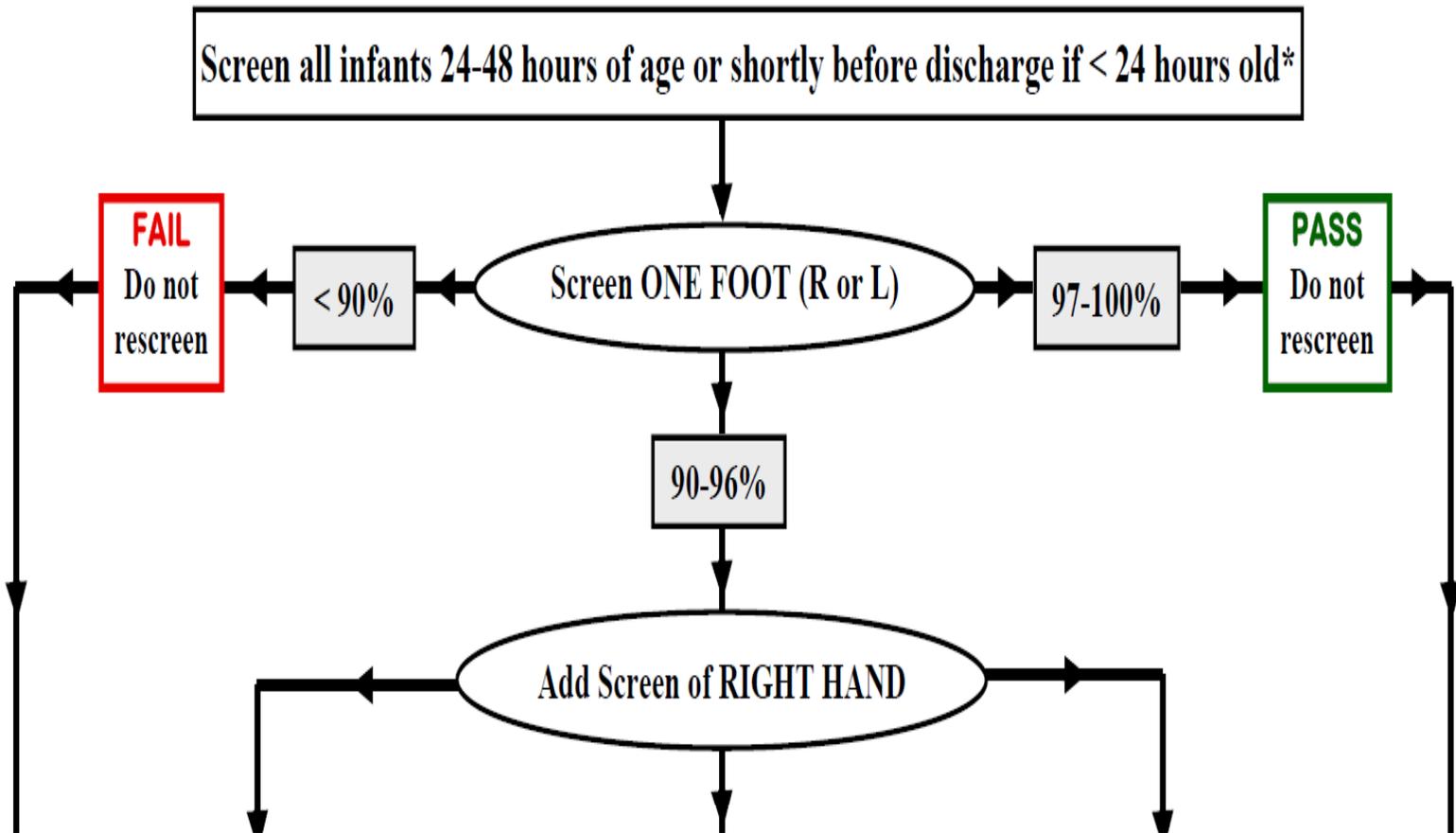
A Note of Caution

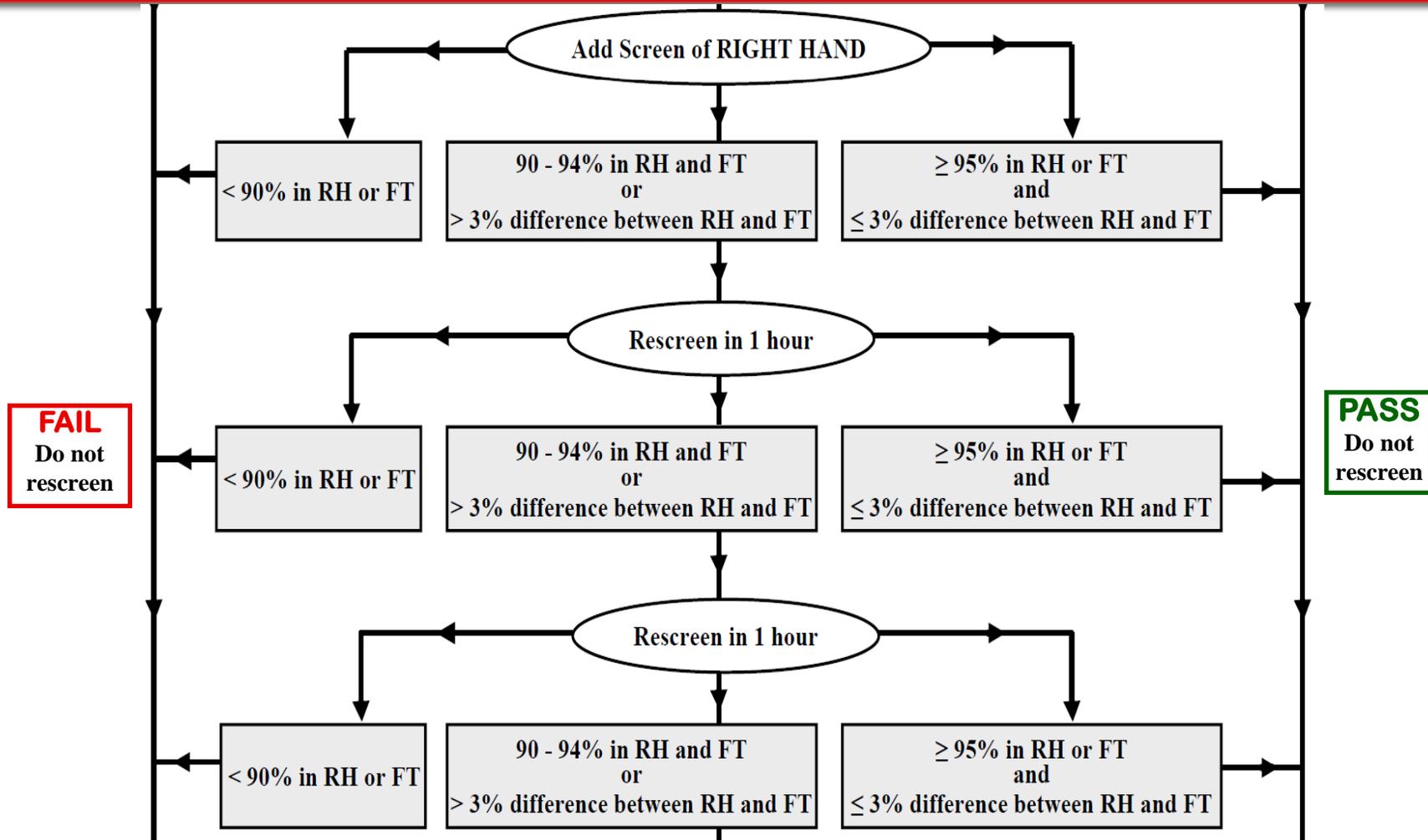
- ❖ A 'Pass' does **not** exclude the existence of a cardiac disorder.
- ❖ If cardiac evaluation is otherwise indicated (e.g., clinical signs, prenatal diagnosis of critical congenital heart disease, dysmorphic features, etc.), proceed with cardiac evaluation **even if** baby receives a pass on the pulse oximetry screen.

- ❖ The newborn fails the screening if the:
 - Oxygen saturation is $<90\%$ (hand or foot) at anytime during screening.
 - Oxygen saturations are $<95\%$ in **both** the hand and the foot (left or right) or there is a $>3\%$ difference between the hand and foot on three measures separated by one hour.
 - A newborn with a failed screening should be referred for additional evaluation.
- ❖ If the results are "fail", it means that the baby's test results showed low levels of oxygen in the blood, which can be a sign of a CCHD. This does not *always* mean that the baby has a CCHD. It just means that more testing is needed.

Protocol for Critical Congenital Heart Disease (CCHD) Screening Tennessee Department of Health



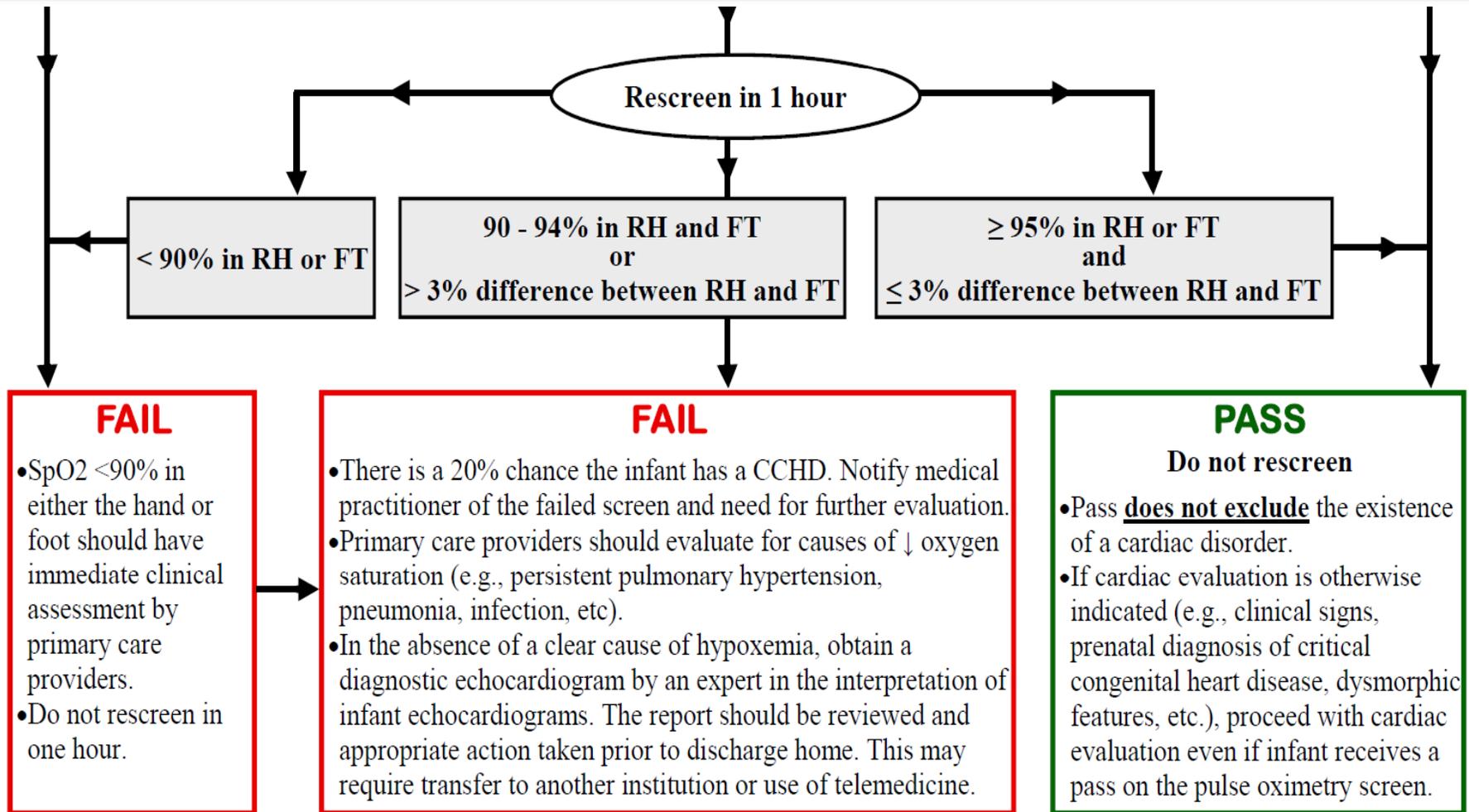




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Newborn Screening Program

Algorithm



Quick Reference Chart

Chart for identifying infants with > 3% difference between the right hand and foot:

		FOOT											
		100	99	98	97	96	95	94	93	92	91	90	<90
RIGHT HAND	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

Right hand screening not needed if foot saturation is 97-100%

❖ Filling in data fields on form (front of filter card)

PULSE OXIMETRY (>24 hours of age)

SEE BACK OF FORM FOR SCREENING INSTRUCTIONS

Initial O2 Screen Date/Time:

____/____/____ @ (____:____) ^{MIL}_{TIME}

Did both RH and foot need to be tested? (____)Y (____)N

Final Result: Passed (____) Failed (____)

Referred to Cardiology: (____)Y (____)N

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Newborn Screening Program

Instructions on Back of Filter Card

- ❖ Perform screen at 24-48 hours of age or shortly before discharge if <24 hours old.
- ❖ Screen foot only (either foot) and document date and time in area for initial screen:
 - If 97 - 100%: Mark PASSED on form. No further testing needed.
 - If <90%: Mark FAILED.
 - If 90-96%: Add the screening of Right Hand
- ❖ If both RH and Foot need to be screened, note it on the collection form:
Mark PASSED if:
 - $\geq 95\%$ in either extremity with a $\leq 3\%$ difference between the two measures.



Instructions on Back of Filter Card

Mark FAILED if:

- <90% in either the RH or the foot at anytime.
- <95% in both the RH and the foot or a >3% difference between RH and foot on three measures each separated by one hour.

Example:

- ✓ If measurements are <95% in both the RH and foot or there is a >3% difference between the two:
 - Perform a second screen in one hour
- ✓ If second measurements are <95% in both the RH and foot or >3% difference between the RH and foot
 - Perform a third screen in one hour
- ✓ If third measurements are <95% in both the RH and foot or >3% difference between the RH and foot
 - Then mark FAILED

Form Completion (Why it matters?)

❖ Filling in data fields on form

PULSE OXIMETRY (>24 hours of age)

SEE BACK OF FORM FOR SCREENING INSTRUCTIONS

Initial O2 Screen Date/Time:
 _____/_____/_____ @ (_____:_____) ^{MIL} _{TIME}

Did both RH and foot need to be tested? (___)Y (___)N

Final Result: Passed (___) Failed (___)

Referred to Cardiology: (___)Y (___)N

Quality Review
– How many tests are being done **after** 24-hours of life?

How many infants pass with foot-only?

Were any CCHD infants missed by the screen?

What is the follow-up?

Screening Packet for Hospitals

❖ The CCHD Screening Educational Packet includes information about:

1. Overview of Screen
2. Training recommendations
3. Helpful hints when screening newborns
4. Knowledge assessment tool
5. Competency Evaluation examples
6. Reporting form instructions
7. Provider Fact Sheet
8. Parent Fact Sheet (English and Spanish)
9. Resources

- ❖ Once identified, babies with a CCHD should be seen by cardiologists and receive specialized care and treatment that could prevent death or disability early in life.
- ❖ Treatment can include medications and surgery.
- ❖ All hospitals, birthing centers and midwives should have a written plan in place as part of their protocol to guide them if a newborn fails the CCHD screen.

Follow-Up for Surveillance

- ❖ A letter will be generated on newborns that were identified as failing the screen and/or if they were referred to a cardiologist.
- ❖ Letters will be sent to the Primary Care Provider listed on the NBS filter form.
- ❖ The follow-up form will ask for:
 - Final Diagnosis
 - Treatment Date
 - Cardiologist

Dr. Provider:

This infant received a pulse oximetry test at the hospital and either failed the O2 screen and/or was referred to a cardiologist. As the infant's medical provider, we need your assistance in collecting data to determine the effectiveness of the screening and to ensure that the family has received all available assistance. Please complete the following form and return to the Tennessee Department of Health, Newborn Screening Follow-Up Program at Fax 615-532-8555.

Initial Pulse Oximetry Screening:

Performed on: 02/28/2012 Final Result: FAILED Referred to Cardiology: YES

Discharge from Hospital Date: _____ / _____ / _____

If Critical Congenital Heart Disease (CCHD) has been confirmed:

Cardiologist: _____ Phone Number: _____

Diagnosis Date: _____ / _____ / _____ Treatment Date: _____ / _____ / _____

Prenatally Diagnosed? Yes No

Final Diagnosis:

- | | |
|--|--|
| <input type="checkbox"/> Hypoplastic Left Heart Syndrome | <input type="checkbox"/> Pulmonary Atresia |
| <input type="checkbox"/> Tetralogy of Fallot | <input type="checkbox"/> Truncus Arteriosus |
| <input type="checkbox"/> Tricuspid Atresia | <input type="checkbox"/> Total Anomalous Pulmonary Venous Return |
| <input type="checkbox"/> Transposition of the Great Arteries | <input type="checkbox"/> Coarctation of the Aorta |
| <input type="checkbox"/> Other: _____ | |

If Child Expired: Date of Death: _____ / _____ / _____

If determined not to be CCHD:

Pulmonology Etiology Infectious Disease Etiology Other Etiology None (normal)

Comments _____

Signature: _____ Notified NBS f/u: _____ / _____ / _____

- ❖ Reports will be sent to the hospitals informing them of the number of newborns who had a metabolic screen submitted to the State Laboratory without the oxygen saturations being documented on the form.
 - 84,533 births in Tennessee in 2010:
 - ✓ 99.4% had a metabolic blood screen submitted
 - ✓ 97% had a hearing screen performed
- ❖ There is a system set up using vital records, hospital discharge codes and reports from cardiologists to identify any missed cases of CCHD.

WEST TENNESSEE

- LeBonheur Children's Hospital
50 N. Dunlap Street
Lobby Level
Memphis, TN 38103
(901) 866-8817
- Pediatric Cardiology, PC
805 Estate Place #1
Memphis, TN 38120
(901) 287-4150
- Dr. Dane Douglas
6401 Poplar Avenue #402
Memphis, TN 38119
(901) 682-7774

MIDDLE TENNESSEE

- Children's Hearts
1919 Charlotte Ave Suite 230
Nashville, TN 37203
(615) 321-8549
- Vanderbilt Pediatric Heart
Institute 2200 Children's Way
Nashville, TN 37232
(615) 322-7447
- Pediatric Cardiology,
Winchester Pediatrics
155 Hospital Road Suite E
Winchester, TN 37398
(931) 962-0672

EAST TENNESSEE

- Tri-City Pediatric Cardiology
2312 Knob Creek Road
Suite 208
Johnson City, TN 37604
(423) 610-1099
- East Tennessee Pediatric Cardiology
2001 Highland Avenue
Suite B
Knoxville, TN 37916
(865) 971-6897
- ETSU Pediatric Cardiology
325 N. State of Franklin Road
Johnson City, TN 37604
(423) 439-7320
- Knoxville Pediatric Cardiology
2100 Clinch Avenue
Knoxville, TN 37916
(865) 522-0420
- TC Thompson Children's Hospital
910 Blackford Street
Chattanooga, TN 37403
(423) 778-6180

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Newborn Screening Program

Thank you!

❖ **Thank you** for being the frontlines of an incredible system to provide a strong early childhood system in Tennessee.

Every infant born in Tennessee deserves a great start to life!

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Newborn Screening Program

Contact Information

❖ Newborn Screening Follow-up Program

- Phone: (615) 532-8462
(855) 202-1357
- Fax: (615) 532-8555