



Critical Congenital Heart Disease (CCHD) Screening in Ontario

Definition

Critical Congenital Heart Disease (CCHD):

(noun); life threatening, structural cardiac malformations, present from birth, where surgery or catheter-based intervention are typically required in the first year for survival



Incidence

In Canada, 12 in 1000 babies are born with a heart defect (CHD).
One quarter of these babies are *critical* CHD (CCHD) (3 in 1000).

- accounts for more newborn deaths than any other type of congenital defect
- represents up to 40% of all deaths from congenital defects and 3-7.5% of all infant deaths
- Unrecognized CCHD can result in sudden deterioration and death

How is CCHD identified?

Approximately **50%** of CCHD cases are identified by prenatal ultrasound.

Newborn physical assessment can detect more cases (**20-30%**).

What about the other 20-30%?

Why screen?

In the immediate newborn period, babies with CCHD can have a normal newborn exam with no heart murmur and no clinical cyanosis but most will have hypoxemia.

Pulse Oximetry Screening measures oxygenation and can identify these babies before they show signs of the disease adding a third layer of detection.

Early Detection = Better Outcome

Changes in the structure and function of the newborn heart can lead to CCHD going unrecognized (e.g. patent ductus arteriosus or PDA).

A PDA can provide enough blood flow of mixed oxygenated and unoxygenated blood to hide a critical heart defect.

A baby with CCHD is reliant on this mixing.

The crisis happens when the ductus closes, resulting in a rapid clinical deterioration with potential life-threatening consequences.

Pulse oximetry can identify low oxygen levels or a more than allowable difference between pre and post ductal measurements, both flags for identifying CCHD.

Three-layer approach



+



+



The combination of prenatal ultrasound, physical assessment and pulse oximetry screening is the best approach to identify newborns with CCHD. It should be a rare situation where CCHD is missed.

Targets of CCHD Screening

PRIMARY-

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

SECONDARY- Cardiac

- Coarctation of the aorta
- Double outlet right ventricle
- Ebstein's anomaly
- Interrupted aortic arch
- Single ventricles

SECONDARY –Non-Cardiac

- Respiratory diseases
- Sepsis
- Persistent Pulmonary Hypertension of the Newborn (PPHN)
- Delayed Transition

Responsibilities for the Screener

- Ensure **quality** screening
 - Follow the NSO recommended algorithm
 - Perform the screen adhering to best practice standards
 - Escalate a screen positive for urgent assessment by a physician*
 - Document completely and accurately
- **Educate** parents/guardians
 - CCHD screening is not mandatory
 - CCHD screening is a recommended standard of care for newborns

Pulse Oximetry Screening



Quick, painless,
non-invasive, and
cost effective



Takes only a few
minutes to
perform and
results are
available
immediately



Optimal timing **24-
48 hours of age**;
the earlier during
that time frame,
the better

The goal of pulse oximetry screening is **to increase the rate of detection prior to clinical deterioration** in affected newborns.

Best Practices

Screen well babies, in a quiet, non-fussing state, prior to any disruptive care activities (e.g. bloodwork).

Early Discharge – **If discharge occurs before 24 hours**, arrangements are to be made for CCHD screening during the recommended time frame.

NICU/SCN babies may be screened if -

- You have easy access to the right hand
- Their cardio-respiratory status is stable. (e.g. vital signs within normal range, no assisted ventilation or supplemental Oxygen)
- They are discharged at less than 7 days of age

Do NOT screen babies diagnosed with CCHD/CHD prenatally or symptomatically after birth, babies over 7 days of age or when parents/guardians decline.

CCHD Pulse Oximetry Screening

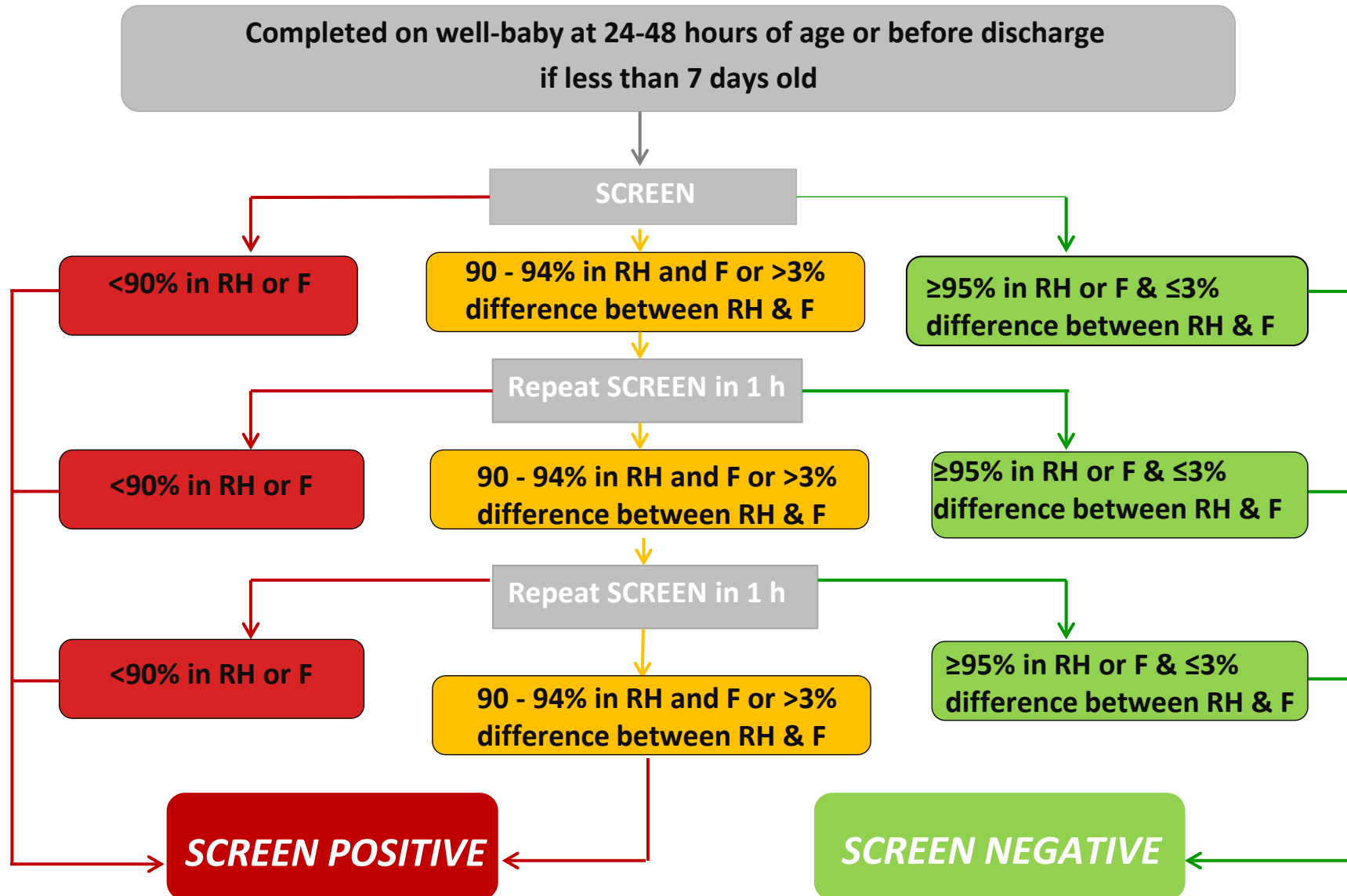
Two separate consecutive oxygen saturation measurements

Pre-ductal (**RIGHT hand**) and post-ductal (**EITHER foot**) saturations are measured in direct sequence, noting the highest value achieved during a 30 second evaluation **once a reliable signal is obtained.**

The two measurements are then evaluated using an algorithm.



CCHD Pulse Oximetry Algorithm



Evaluation

Refer

Screen Positive

Urgent referral to a physician

Repeat

in one hour, up to 3
attempts

Pass

Screen Negative

NSO Screening Evaluation Chart

		RIGHT Hand Pulse Oximetry Measurement												
Either Foot Pulse Oximetry Measurement		100	99	98	97	96	95	94	93	92	91	90	≤89	
	100	Green	Green	Green	Green	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Red
	99	Green	Green	Green	Green	Green	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Red
	98	Green	Green	Green	Green	Green	Green	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Red
	97	Green	Green	Green	Green	Green	Green	Green	Yellow	Yellow	Yellow	Yellow	Yellow	Red
	96	Yellow	Green	Green	Green	Green	Green	Green	Green	Yellow	Yellow	Yellow	Yellow	Red
	95	Yellow	Yellow	Green	Green	Green	Green	Green	Green	Green	Yellow	Yellow	Yellow	Red
	94	Yellow	Yellow	Yellow	Green	Green	Green	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Red
	93	Yellow	Yellow	Yellow	Yellow	Green	Green	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Red
	92	Yellow	Yellow	Yellow	Yellow	Yellow	Green	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Red
	91	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Red
	90	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Red
	≤89	Red	Red	Red	Red	Red	Red	Red	Red	Red	Red	Red	Red	Red

PASS
screen complete

REPEAT
In 1 hr (max 2 repeats)

REFER
Physician assessment required

Screen Negative

SpO₂ is greater than or equal to 95% in either the hand or foot, with less than or equal to 3% difference between them

What next?

No further measurements required

Inform parents/guardians of the result

Documentation on CCHD portion of the blood spot card and forward to Newborn Screening Ontario



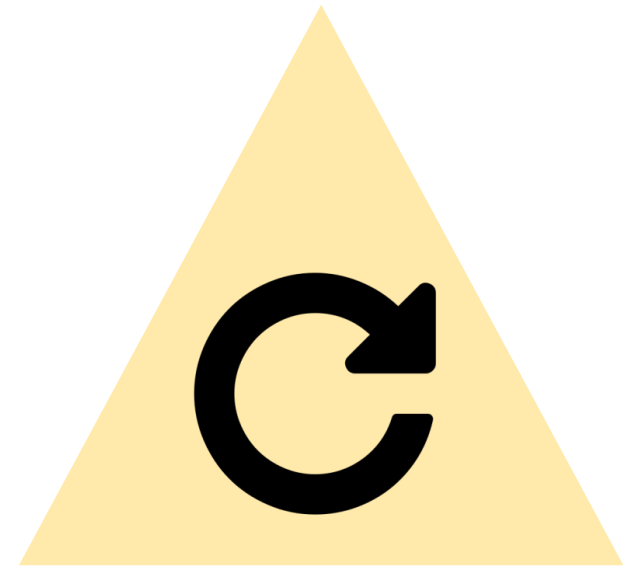
Repeat Result

The SpO₂ is less than 95% in hand AND foot (but not less than 90) or more than 3% difference between the hand and foot

What next?

The screen can be repeated twice for a total of three chances

After the third screen, you will have either a **Pass** or **Refer** result

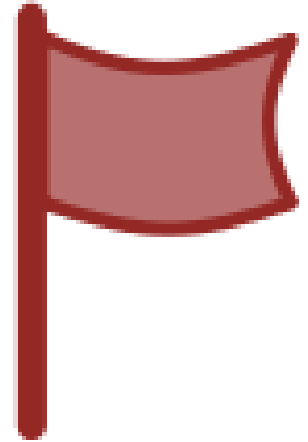


Screen Positive

SpO₂ in hand OR foot less than 90% at any time

OR

SpO₂ is less than 95% in hand AND foot or more than 3% difference on **3** separate measures, each separated by 1 hour



Remember: “Three strikes and you’re out”

Screen Positive? What now?

Urgent referral at the time of the screen positive to a physician for further investigation

Possible transfer to another unit or hospital

Gold standard for cardiac diagnosis is the **echocardiogram**



Remember

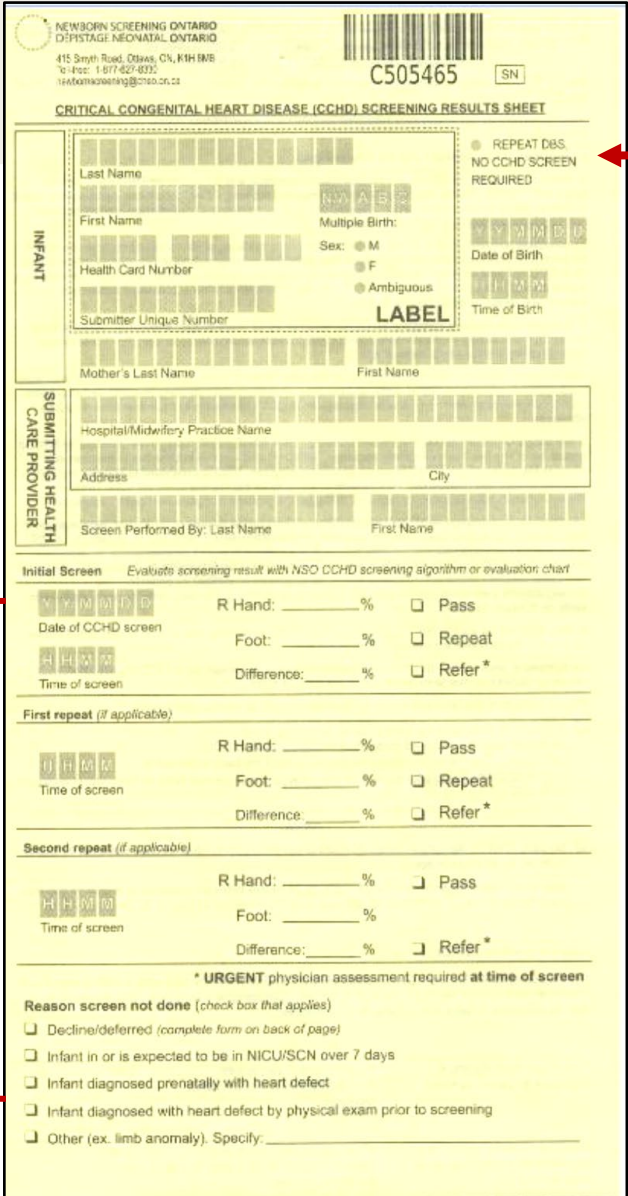
A **screen positive** does not necessarily mean the baby has CCHD.

It indicates a need for further assessment.

Documentation

CCHD documentation form - detachable part of the newborn screening blood spot form

Every baby should have either a screen result or a reason why the screen was not completed documented.



NEWBORN SCREENING ONTARIO
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C505465 SN

CRITICAL CONGENITAL HEART DISEASE (CCHD) SCREENING RESULTS SHEET

INFANT

Last Name: [Grid]
First Name: [Grid] Multiple Birth: [Y/N] [M/D]
Sex: M F Ambiguous
Date of Birth: [Y] [M] [D]
Time of Birth: [H] [M]
Health Card Number: [Grid]
Submitter Unique Number: [Grid] LABEL

SUBMITTING HEALTH CARE PROVIDER

Mother's Last Name: [Grid] First Name: [Grid]
Hospital/Midwifery Practice Name: [Grid]
Address: [Grid] City: [Grid]
Screen Performed By: Last Name: [Grid] First Name: [Grid]

Initial Screen Evaluate screening result with NSO CCHD screening algorithm or evaluation chart

Date of CCHD screen: [Y] [M] [D] [D] R Hand: _____% Pass
Foot: _____% Repeat
Time of screen: [H] [M] [M] Difference: _____% Refer *

First repeat (if applicable)

Time of screen: [H] [M] [M] R Hand: _____% Pass
Foot: _____% Repeat
Difference: _____% Refer *

Second repeat (if applicable)

Time of screen: [H] [M] [M] R Hand: _____% Pass
Foot: _____% Repeat
Difference: _____% Refer *

* URGENT physician assessment required at time of screen

Reason screen not done (check box that applies)

- Decline/deferred (complete form on back of page)
- Infant in or is expected to be in NICU/SCN over 7 days
- Infant diagnosed prenatally with heart defect
- Infant diagnosed with heart defect by physical exam prior to screening
- Other (ex. limb anomaly). Specify: _____

Indicate repeat blood spot and no CCHD screen done here.

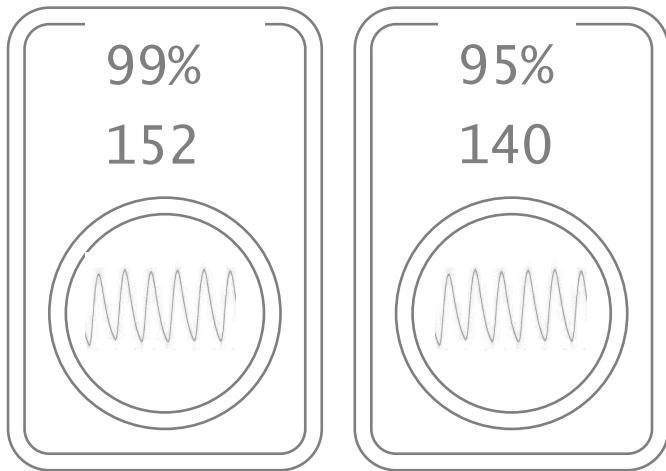
At Newborn Screening Ontario, CCHD screening forms are entered into an electronic data base evaluated through the screening algorithm for quality management.

Unsatisfactory screens and referrals (screen positives) are identified.

Newborn Screening Ontario will follow up on unsatisfactory or missed screens as well as screen positives with a phone call or email to determine the clinical path and outcome.



Practice

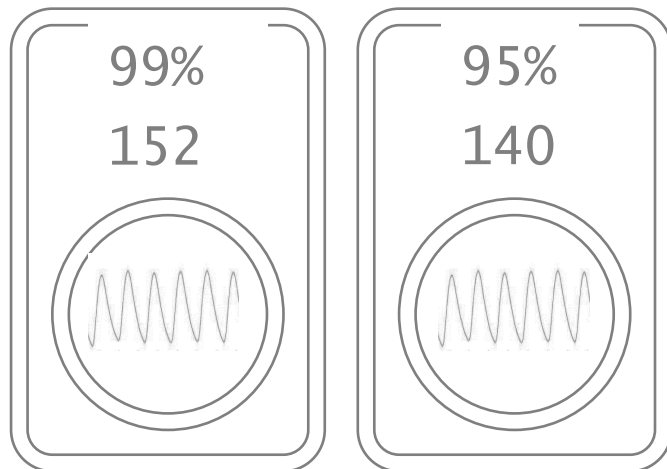


Initial screen

30 hrs old

- Pass
- Repeat
- Refer

Practice



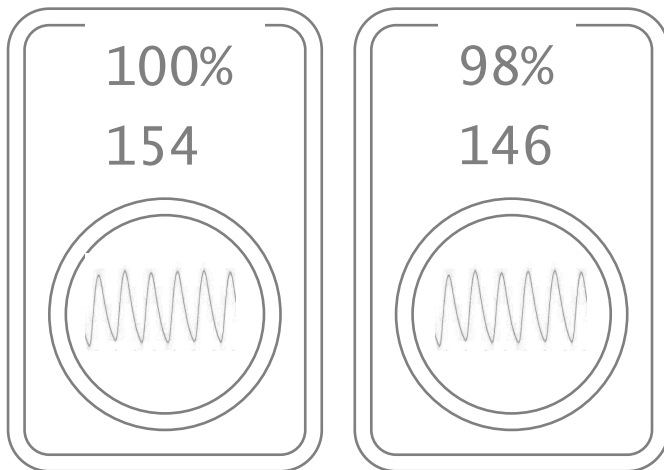
Initial screen

30 hrs old

- Pass
- Repeat
- Refer

Although one value is over 95%, the **difference between the two values is more than 3%**. The screen should be repeated in one hour

Practice

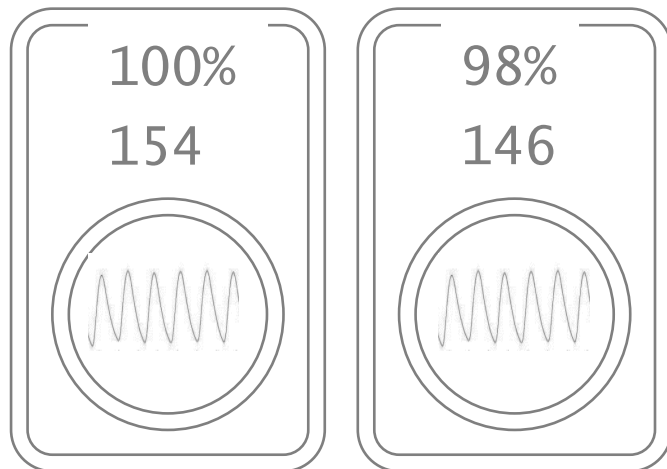


Second attempt

31 hrs old

- Pass
- Repeat
- Refer

Practice



Second attempt

31 hrs old

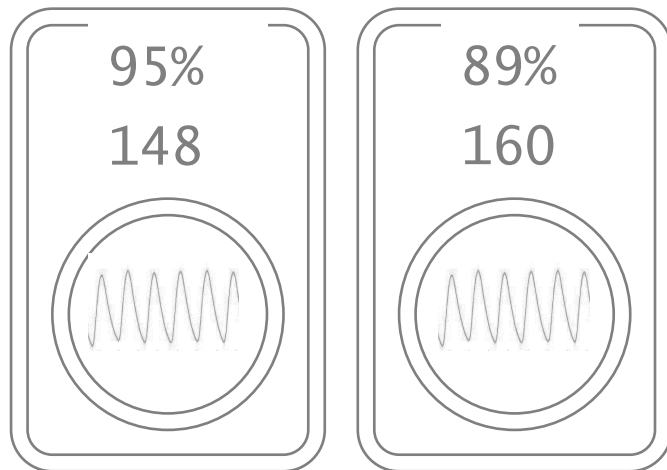
- Pass
- Repeat
- Refer

At least one value is over 95% and the difference is less than or equal to 3%.

The screen is complete

No further action should be taken. Document on CCHD form and forward to NSO

Practice



Initial screen

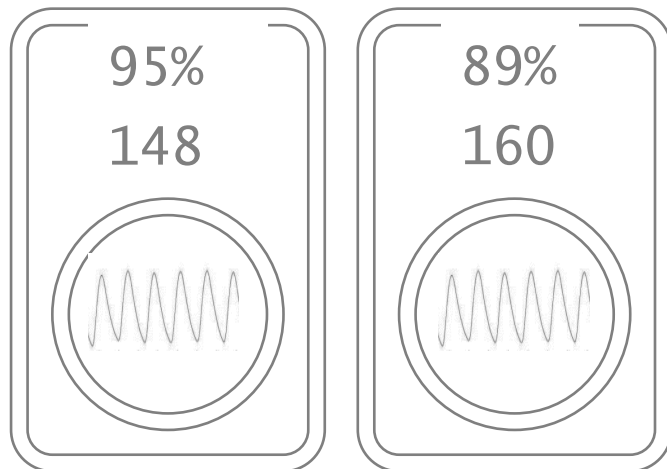
25 hrs old

Pass

Repeat

Refer

Practice



Initial screen

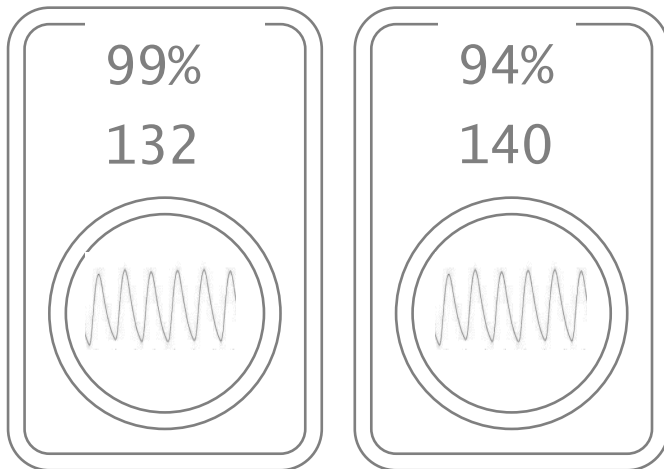
25 hrs old

- Pass
- Repeat
- Refer

If either value is **less than 90 at any time**, the screen result is **Refer**.

Do not repeat the screen, but rather initiate next steps for urgent referral to a physician

Practice

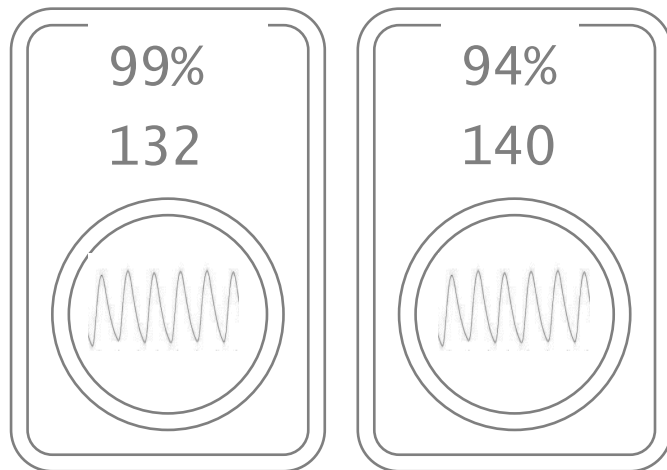


Initial screen

16 hrs old

- Pass
- Repeat
- Refer

Practice



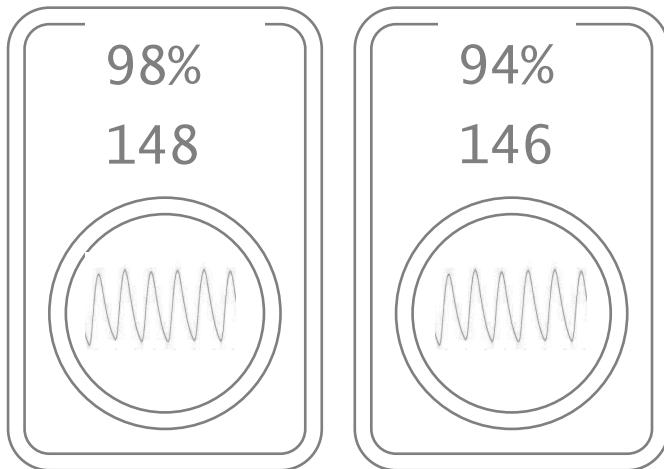
Initial screen

16 hrs old

- Pass
- Repeat
- Refer

Evidence shows **24-48 hrs post-birth** is the ideal time for PO testing to avoid false positives due to transition from fetal to neonatal circulation. Screening at 16 hours does not follow the algorithm. This screen will need to be performed at the **appropriate time**.

Practice

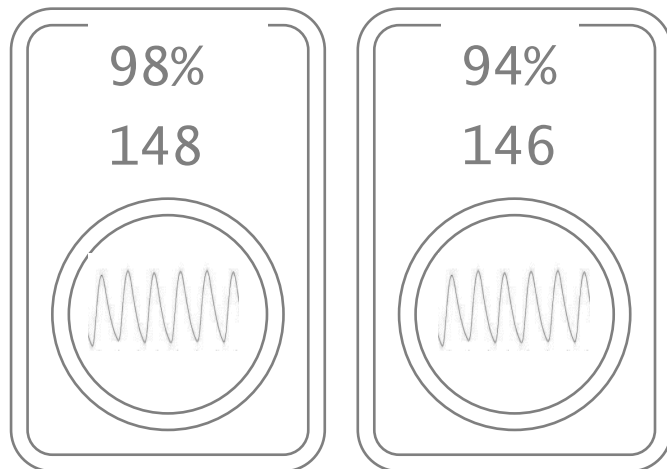


Third attempt

26 hrs old

- Pass
- Repeat
- Refer

Practice



Third attempt

26 hrs old

Pass

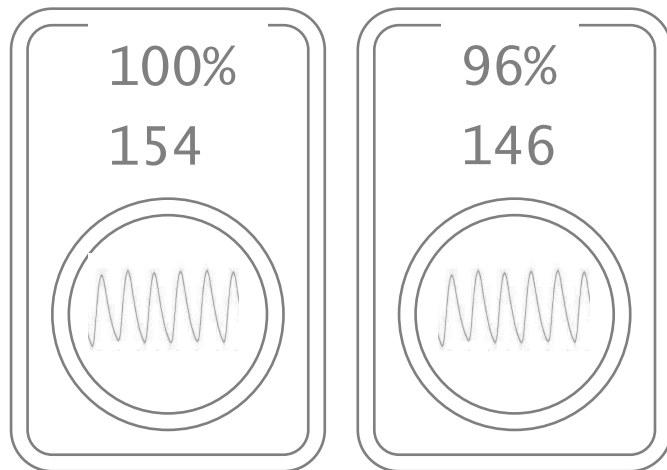
Repeat

Refer

Although one value is over 95%, the **difference between the two values is more than 3%**.

Since this is the third screen and the values are not satisfactory, the result is **Refer**.

Practice

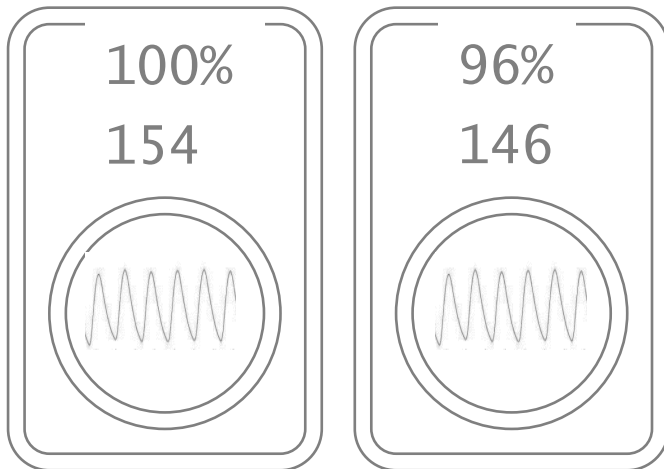


Initial screen

24 hrs old

- Pass
- Repeat
- Refer

Practice



Initial screen

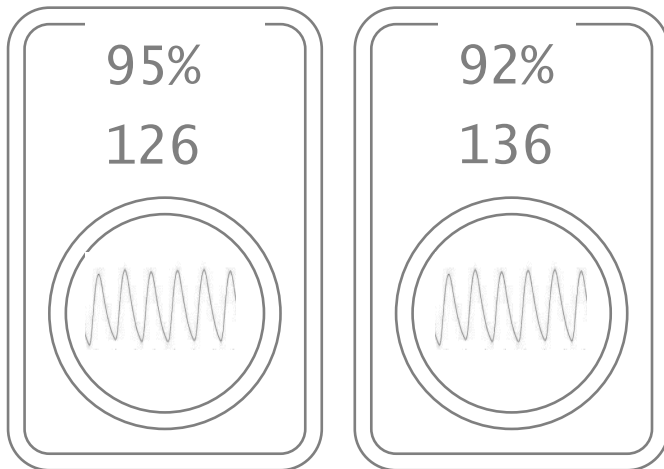
24 hrs old

- Pass
- Repeat
- Refer

Although one value is over 95%, the **difference between the two values is more than 3%**. The screen should be repeated in one hour.

MOST COMMON ERROR

Practice

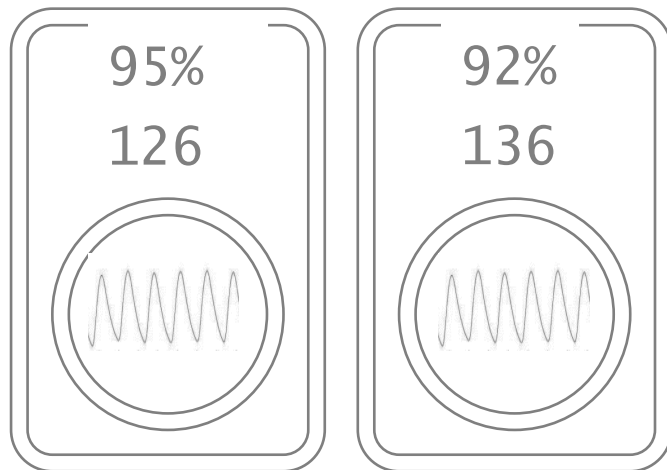


Initial screen

25 hrs old

- Pass
- Repeat
- Refer

Practice



Initial screen

25 hrs old

- Pass
- Repeat
- Refer

Although this result meets the criteria for a CCHD Screen **pass**, the saturation values are not ideal for a well baby. It is important to **pay attention to the clinical picture** of this baby.



#Goals

Standardization

Access

Quality

*The purpose of the screen is to detect oxygen saturation issues potentially related to CCHD. However, it is important **to never ignore the rest of the clinical picture.***

Remember most babies will pass the CCHD screen on the first attempt easily...this is good news.

We screen for those who don't.





The End.

Questions?

- American Academy of Pediatrics, Newborn Screening for CCHD, *Answers and Resources for Primary Care Pediatricians*; (2016) retrieved from <https://www.aap.org/en-us/advocacy-and-policy/aap-health-initiatives/PEHDIC/Pages/Newborn-Screening-for-CCHD.aspx> CCS document
- Center for Disease Control, *Screening for Critical Congenital Heart Defects*, (2016) retrieved from <http://www.cdc.gov/ncbddd/heartdefects/cchd-facts.html>
- Kemper AR, Mahle WT, Martin GR, et al. *Strategies For Implementing Screening For Critical Congenital Heart Disease*. *Pediatrics*. 2011;128(5):e1259-e1267. doi:10.1542/peds.2011-1317.
- Utah Public Health Department, *CCHD Toolkit*, (2016) retrieved from <http://www.health.utah.gov/cchd/>
- Wong KK, Fournier A, Fruitman DS, Graves L, Human DG, Narvey M, Russell JL, *CCS/CPCA Position Statement on Pulse Oximetry Screening in Newborns to Enhance Detection of Critical Congenital Heart Disease*, *Canadian Journal of Cardiology* (2016), doi: 10.1016/j.cjca.2016.10.006.