Ontario Newborn Screening for Critical Congenital Heart Disease (CCHD)

Submitters Training Guide
PART ONE

❤ Overview of CCHD
  • What is CCHD? Incidence? Why Screen?

❤ How/Who do we screen?

Best Practices

❤ Evaluation of the Screen

PART TWO

❤ Practice Exercises
PART ONE

Did you know...

In March 2016, the Ministry of Health and Long Term Care approved the addition of Critical Congenital Heart Disease (CCHD) to Newborn Screening Ontario’s (NSO’s) newborn screening panel.

The goal is that every infant born in Ontario is offered CCHD screening by pulse oximetry.
What is CCHD?

Definition: (noun)

❤️ Critical Congenital Heart Disease (CCHD)

a term that refers to a group of serious heart defects that can affect the structure or vessels of the heart and are present from birth

❤️ CCHD prevents the heart from pumping blood effectively or reduces the amount of oxygen in the blood. Either can lead to organ damage and life-threatening complications

AAP/CDC, 2016
It is not uncommon...

Congenital Heart Disease (CHD) occurs in 12 per 1000 live births, 25% of CHD cases are critical or CCHD – require surgery or catheter intervention in the 1st year of life.

CHD remains a leading cause of infant death.

~1-2 infant deaths per year in Ontario due to undiagnosed CCHD (Ontario Coroner’s office)
Pulse Oximetry Primary Targets

- 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

The Cyanotic Seven
Detected by Pulse Oximetry

Secondary Targets: CCHDs detected by Pulse Oximetry (sometimes cyanotic):

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

Also:

- Sepsis
- Respiratory Issues
- Persistent Pulmonary Hypertension of the Newborn (PPHN)
Why screen for CCHD?

Missed or late diagnosis can be devastating...

❤️ Prenatal ultrasound detects only about half of CCHD

❤️ Unfortunately, changes in the structure and function of the newborn heart can lead to CCHD going unrecognized during the newborn hospital stay

Pulse Oximetry Newborn Screening can identify some infants with CCHD before they show signs of the condition, resulting in better outcomes for the baby
Diagnosis of CCHD

PO Screening used **IN COMBINATION** with prenatal ultrasound & postnatal physical exam, is the best approach to identify newborns with CCHD that would otherwise be missed prior to discharge.

*Estimated ~300 positive screens per year in Ontario with CCHD pulse oximetry screening implementation*
Responsibilities

❤️ Education to parents/guardians should include
  ❤️ CCHD screening is not mandatory
  ❤️ CCHD screening is a recommended standard of care

❤️ Parents/guardians have the right to decline
   ❤️ If they choose to do so, please document this on the CCHD portion of the blood spot card
Best Practices

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

❤ If discharge occurs before 24 hours, arrangements to be made for CCHD screening during the recommended time frame

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

❤ NICU/SCN/PICU babies may be screened if

❤ Cardio-respiratory status is stable and expected length of stay is less than 7 days OR at time of discharge if baby is less than 7 days of age
Special Considerations

*Do not screen:*

❤ Infants in NICU/SCN/PICU with an expected length of stay of **longer than 7 days**

❤ Infants diagnosed with CCHD/CHD prenatally or symptomatically after birth

❤ When parents/guardians decline
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 values are then compared using the algorithm or evaluation chart.
CCHD Pulse Oximetry Algorithm

Completed on well baby at 24-48 hours of age or before discharge if less than 7 days old

SCREEN

<90% in RH or F

90 - 94% in RH and F or >3% difference between RH & F

≥95% in RH or F & ≤3% difference between RH & F

Repeat SCREEN in 1 h

<90% in RH or F

90 - 94% in RH and F or >3% difference between RH & F

≥95% in RH or F & ≤3% difference between RH & F

Repeat SCREEN in 1 h

<90% in RH or F

90 - 94% in RH and F or >3% difference between RH & F

≥95% in RH or F & ≤3% difference between RH & F

SCREEN POSITIVE

SCREEN NEGATIVE

(Adapted from Kemper et al, 2011)
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Adapted from the Utah Department of Public Health
Screen Negative

Screen Negative ("Pass")

- 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...

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

❤️ 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 ("Refer")

♥ 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 next?

♥ Urgent referral to a physician for further investigation

♥ For out of hospital environments, follow your usual referral protocol as appropriate per clinical picture
Screen Positive Result?

❤️ Urgent physician referral
  • Consideration of non-cardiac pathology (e.g. infection, persistent pulmonary hypertension)
  • Referral to pediatric cardiology / echocardiography if cannot exclude CCHD

❤️ Possible transfer to another unit or hospital

❤️ Inform parents/guardians of the result

❤️ Documentation on CCHD portion of the blood spot card and forward to Newborn Screening Ontario
Remember...a screen positive does not necessarily mean CCHD...it indicates a need for further assessment

Newborn Screening Ontario will follow up on screen positives with a phone call to determine the outcome
Example One:

Term infant, 30 hours of age, initial screen

- Pre-ductal RIGHT Hand: 100%
- Post-ductal (left foot): 96%

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

Pass  Repeat  Refer
Repeat Screen one hour later:

Term infant, 31 hours of age, second screen

- Pre-ductal RIGHT Hand 98%
- Post-ductal (left foot) 96%

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.
Example Two:

Term infant, 25 hour old.

- Pre-ductal RIGHT Hand: 95%
- Post-ductal (left foot): 89%

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.
Example Three:

Term infant, 40 hours old, initial screen

- Pre-ductal RIGHT Hand: 97%
- Post-ductal (left foot): 93%

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

Term infant, 41 hours old:

- Pre-ductal RIGHT Hand: 99%
- Post-ductal (left foot): 95%

Although both values are over 95%, the difference between the two values is more than 3%.

The screen should be repeated again in one hour.
Term infant, 42 hours of age, third screen

Pre-ductal RIGHT Hand  97%

Post-ductal (left foot)  93%

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.
Example Five:

Term infant, 16 hours old

Initial screen

- Pre-ductal (right hand) 99%
- Post-ductal (left foot) 95%

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.

The timing of the screen is not appropriate.
Example Four:

Term infant, 24 hours of age, initial screen

- Pre-ductal RIGHT Hand 95%
- Post-ductal (left foot) 92%

Pass

Repeat

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.
A few last notes...

❤ 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 easily...this is good news. We screen for those who don’t.
Questions?

Thank you for your front line commitment to promoting healthy starts for Ontario’s babies!
References


