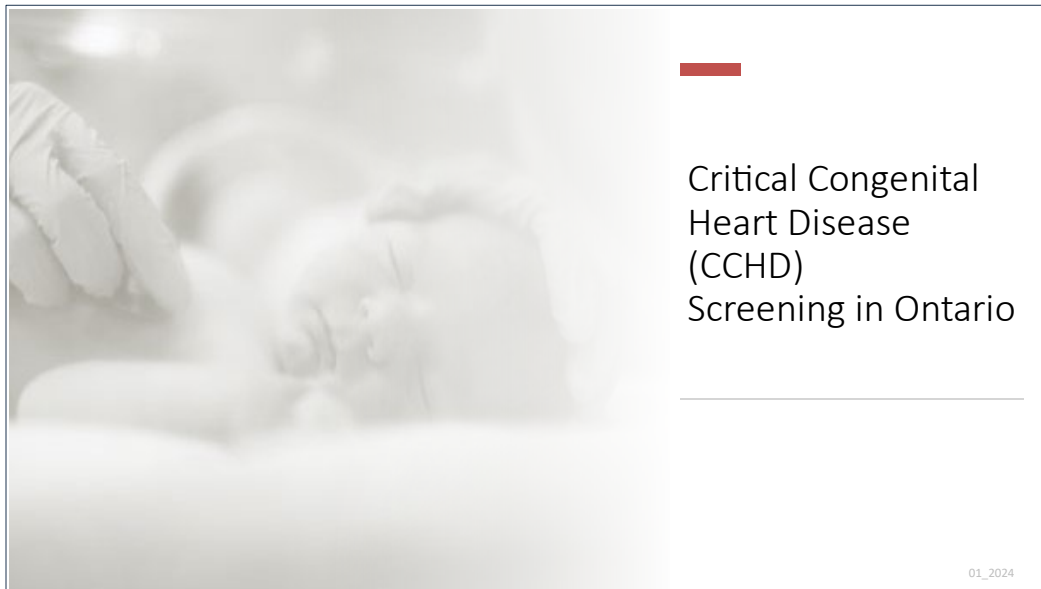





# Suggested Script for Submitters Training Module CCHD Power Point Presentation



## Suggested script to accompany Power Point presentation -

<p><b>Slide 2</b></p> <div data-bbox="253 275 631 485" style="border: 1px solid black; padding: 10px; text-align: center;"> <p>Definition</p> <p><b>Critical Congenital Heart Disease (CCHD):</b> <i>(major), life threatening, structural cardiac malformations, present from birth, where surgery or catheter-based intervention are typically required in the first year for survival</i></p>  </div>	<ul style="list-style-type: none"> <li>• CCHD is an acronym for Critical Congenital Heart Disease. It is life threatening and present from birth.</li> <li>• It involves structural malformations of the heart or major vessels of the heart which require surgery or other catheter-based intervention or procedures in the first year of life for survival.</li> <li>• It is not one singular disease...but rather a group of diseases...7 identified as primary targets for screening.</li> </ul>
<p><b>Slide 3</b></p> <div data-bbox="246 644 641 867" style="border: 1px solid black; padding: 10px;"> <p style="text-align: center;">Incidence</p> <p><small>In Canada, 12 in 1000 babies are born with a heart defect (CHD). One quarter of these babies are critical CHD (CCHD) (<b>3 in 1000</b>).</small></p> <ul style="list-style-type: none"> <li>• accounts for more newborn deaths than any other type of congenital defect</li> <li>• represents up to 40% of all deaths from congenital defects and 3-7.5% of all infant deaths</li> <li>• Unrecognized CCHD can result in sudden deterioration and death</li> </ul> </div>	<ul style="list-style-type: none"> <li>• <i>Statistics for Canada show Congenital Heart Defects or <b>CHD</b> occur in about 1 in 100 births or 12 in 1000, and about <b>25%</b> of these or about <b>3 in 1000</b> are Critical CHD</i></li> <li>• <i>Congenital <b>heart</b> defects account for more newborn deaths than any other type of congenital defect, representing up to 40% of all deaths from congenital defects and 3–7.5% of all infant deaths.</i></li> <li>• <i>Unrecognized CCHD can result in sudden deterioration and death.</i></li> <li>• <i>Most babies with CCHD now survive past infancy due to improvements in early detection, diagnosis, and treatment</i></li> </ul>
<p><b>Slide 4</b></p> <div data-bbox="233 1121 652 1356" style="border: 1px solid black; padding: 10px;"> <p style="text-align: center;">How is CCHD identified?</p> <p><small>Approximately 50% of CCHD cases are identified by prenatal ultrasound.</small></p> <p><small>Newborn physical assessment can detect more cases (20-30%).</small></p> <p style="text-align: center;"><b>What about the other 20-30%?</b></p> </div>	<ul style="list-style-type: none"> <li>• <i>Approximately 50% of CCHD cases are identified by prenatal ultrasound... very surprising!! (with some cardiologists suggesting that this number is actually quite generous!) This detection rate can vary based on several factors including the type of ultrasound technology being used, the level of expertise of the operators, the complexity of the cardiac lesions and location of the centre (a tertiary centre vs a non-tertiary centre)</i></li> <li>• <i>Another 20-30 percent of babies with CCHD are detected through physical assessment, (they present with subtle (and sometimes not so subtle) clinical symptoms) ...in the first day or so of life (prior to when they would have been screened...)</i></li> <li>• <i>This leaves us with our final 20-30% of babies with CCHD... What about those kids...</i></li> </ul>

Slide 5

**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

- *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**.*
- *Low oxygen levels are frequently a sign of CCHD.*
- *Pulse oximetry measures oxygen saturation levels in the blood and can detect this subtle hypoxemia from ductal systemic or pulmonary blood flow.*
- *Mild hypoxemia is often missed through visual assessment alone...factors such as ethnicity, perfusion, skin thickness and even ambient lighting can affect one's ability to assess colour appropriately. This is a limitation for physical assessment.*
- *A Cyanotic blind spot occurs when the oxygen level is abnormal but there is no obvious visible cyanosis. This occurs around the 83-95 % range depending and makes a baby difficult to assess visually...pulse oximetry is much more reliable to assess oxygen levels.*
- *Pulse oximetry screening can identify babies before they show signs of the disease and so it adds a third layer of detection to capture those babies who may have been missed by ultrasound or physical exam.*
- *It does not replace the current approach to detecting CCHD but is used **in addition to** U/S and Physical assessment.*

Slide 6

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

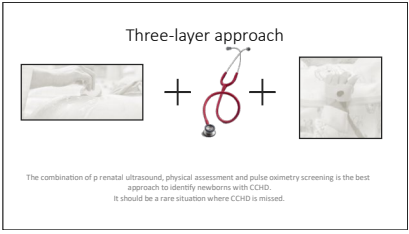
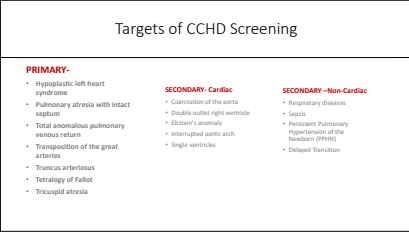
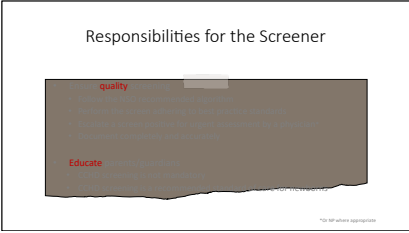
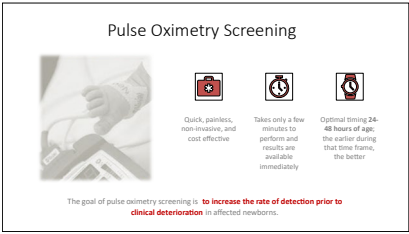
A PDA can provide enough blood flow of mixed oxygenated and unoxygenated blood to hide a critical heart defect.

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 at a more than allowable difference between pre and post ductal measurements, both flags for identifying CCHD.**

**Physiology of CCHD**

- *After birth, the lungs are now the oxygen source, the ductus arteriosus is no longer needed... we don't want the blood to bypass the lungs anymore... we want oxygenated blood from the pulmonary veins to the left ventricle to the aorta to the rest of the body...normal circulation.*
- *Constriction of the ductus arteriosus is a gradual, normal process, dependent on several factors...reduced exposure to prostaglandins (from the placenta), and rising arterial oxygen concentrations...most neonates have a PDA for the first 8 hrs of life... The mean closure time of the ductus arteriosus (DA) in full-term neonates is presumed to be about 24 hours after birth; however, its accurate time is unknown...*
- *With CCHD babies, an open or patent ductus arteriosus can provide enough blood flow to hide a critical heart defect; ... A baby with CCHD is reliant on this mixing of oxygenated and unoxygenated blood...In the case of a baby with Critical heart disease, the crisis happens when the ductus closes...*
- *Closure results in a rapid clinical deterioration and*

	<p><i>potential life-threatening consequences.</i></p>
<p><b>Slide 7</b></p>  <p>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.</p>	<ul style="list-style-type: none"> <li>• <i>The combination of prenatal ultrasound, physical assessment and pulse oximetry screening is the best approach to identify newborns with CCHD.</i></li> <li>• <i>It should be a rare situation where CCHD is missed.</i></li> </ul>
<p><b>Slide 8</b></p>  <p><b>PRIMARY-</b></p> <ul style="list-style-type: none"> <li>• Hyaline membrane disease</li> <li>• Pulmonary atresia with intact septum</li> <li>• Total anomalous pulmonary venous return</li> <li>• Transposition of the great arteries</li> <li>• Truncus arteriosus</li> <li>• Tetralogy of Fallot</li> <li>• Tricuspid atresia</li> </ul> <p><b>SECONDARY-Cardiac</b></p> <ul style="list-style-type: none"> <li>• Coarctation of the aorta</li> <li>• Double outlet right ventricle</li> <li>• Ebstein's anomaly</li> <li>• Interrupted aortic arch</li> <li>• Single ventricle</li> </ul> <p><b>SECONDARY-Non-Cardiac</b></p> <ul style="list-style-type: none"> <li>• Respiratory disease</li> <li>• Sepsis</li> <li>• Persistent Pulmonary Hypertension of the Newborn (PPHN)</li> <li>• Delayed Transition</li> </ul>	<ul style="list-style-type: none"> <li>• <i>This screen measures <b>oxygenation</b></i></li> <li>• <i>Primary targets – called the “Cyanotic 7” are <b>duct dependent</b>, should be identified through screening.</i></li> <li>• <i>Secondary targets, some cardiac - not duct dependent – sometimes identified by screening.</i></li> <li>• <i>other conditions that can be ‘caught in the CCHD screening net’.</i></li> <li>• <i>This is why it is important to tell parents that the screen is not a diagnostic tool for critical heart disease, but an alert to investigate further. Other conditions can cause cyanosis and it is important to determine the cause.</i></li> </ul>
<p><b>Slide 9</b></p>  <p>Quality</p> <p>Educate</p>	<ul style="list-style-type: none"> <li>• <i>Responsibilities include quality screening, and education to families.</i></li> </ul>
<p><b>Slide 10</b></p>  <p>The goal of pulse oximetry screening is to increase the rate of detection prior to clinical deterioration in affected newborns.</p>	<ul style="list-style-type: none"> <li>• <i>Pulse Oximetry Screening is quick, painless, and non-invasive.</i></li> <li>• <i>The best result is obtained with a calm baby, with optimal timing <b>24-48 hours of age</b>; the earlier during that time frame, the better... remember the timing is important... we would expect the ductus to be closed at 24 hours...</i></li> <li>• <i>Pulse oximetry has the ability to continuously and transcutaneously monitor the functional oxygen saturation of hemoglobin in arterial blood (SaO<sub>2</sub>).</i></li> <li>• <i>Pulse oximetry is so widely prevalent in medical care that it is often regarded as a fifth vital sign.</i></li> <li>• <i>Immediate live result, in real time, accuracy to 2-4 % more accurate at higher oxygen levels, less so when oxygenation is lower...</i></li> </ul>

Slide 11

**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/CMD prenatally or symptomatically after birth, babies over 7 days of age or when parents/guardians decline.

- A quiet alert state is best for screening.
- For early discharged babies - it is the responsibility of the organization to ensure arrangements are made for the family during the recommended window
- Pulse Ox screening before 24 hours of age increases the risk of false positive because transitional cardiovascular changes that occur during the initial 24 hours of life may be incomplete (closure of the ductus arteriosus)
- While the pulse ox screen is intended for ‘well babies’, NICU/SCN babies can benefit as well, provided their cardio-respiratory status is stable and their length of stay is expected to be less than 7 days of age. There is no value in performing the screen after 7 days of age as most CCHD cases would be evident by that time.

Slide 12

**CCHD Pulse Oximetry Screening**

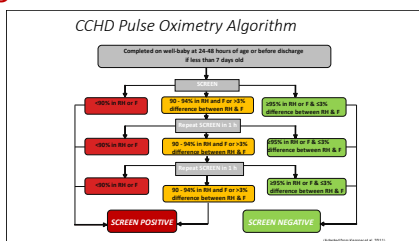
Two separate consecutive oxygen saturation measurements

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

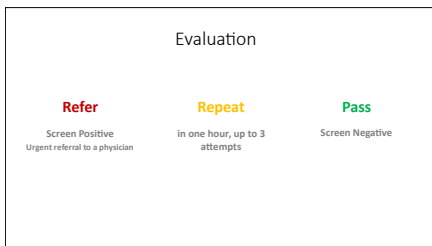
- Evaluation of 2 separate oxygen saturation measurements, one pre-ductal (the RIGHT hand) and one post-ductal (either foot) collected in direct sequence; The two saturations are then evaluated using an algorithm.
- Their numeric value and also their relationship to each other (their difference) can be an indicator of CCHD.
- Your hand and your foot should have the same (or very close) oxygen levels...this is true of healthy hearted babies after 24 hours.
- Babies with CCHD can either have overall low oxygen levels or a notable difference in the two (pre and post ductal) measurements.
- It does not matter which site is performed first (try the least disruptive to the baby)
- Turn off any bili treatment lights as they can interfere with the receptor side of the oximeter probe.
- Ensure a reliable signal is established (using confidence indicators e.g. even pleth line, regular HR audible, etc.) prior to gathering a reading.
- Once you have a reliable signal, watch the saturation value for 30 seconds, noting the highest saturation value during that time. That is your value for that site.

Slide 13



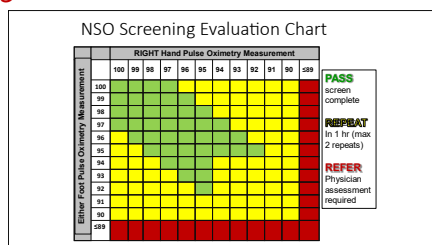
- This is the algorithm that is suggested for use for Ontario babies.
- It is a revised version of the American Academy of Pediatrics (AAP) algorithm for CCHD.
- Note it outlines the recommended window for screening as 24-48 hours.
- It offers 3 result options for each screen with a possible 2 repeat screens (3 total)

Slide 14



- The screen will have 3 options for a result on the first and second attempt.
- The third attempt will only have 2 options... PASS or REFER

Slide 15



- The chart compliments the algorithm.
- Follow the pre-ductal or Right-hand value across the top and the post-ductal or either foot value along the left side. Find the intersecting point and note the colour of the box for the result of the screen.
- Red is immediate REFER... yellow means a repeat in one hour (unless this is the third screen, then refer) and green is a pass, and no further testing is needed.
- You should have this chart (or the algorithm) handy at the time of screening for evaluation purposes.

Slide 16

Screen Negative

SpO<sub>2</sub> 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

- A screen negative or **PASS** result means the baby has at least one measurement over 95% and the difference between the two measurements is 3% or less.
- If a pass result is obtained, the blood spot card is completed, and forwarded to Newborn Screening Ontario.

Slide 17

Repeat Result

The SpO<sub>2</sub> 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.

- For an indeterminate or **REPEAT** result, both measurements are under 95% (but not less than 90%) or the difference is greater than 3%.
- The screen can be done up to 3 times, (an initial, plus 2 repeats...total is 3 attempts)
- The third attempt is the last chance, and the result will either be a pass or refer result.

Slide 18


Screen Positive

SpO<sub>2</sub> in hand **OR** foot less than 90% at any time  
**OR**  
SpO<sub>2</sub> 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"*

- A screen positive or **REFER** result indicates that either both measurements are under 95% (but not less than 90%) or the difference is greater than 3% on three attempts each separated by an hour (3 unsuccessful repeats)
- OR**
- the saturation in the hand or foot was less than 90% at some point in the screen.

Slide 19

<p>Screen Positive? What now?</p> <p><b>Urgent referral</b> at the time of the screen positive to a physician for further investigation</p> <p>Possible transfer to another unit or hospital</p> <p>Gold standard for cardiac diagnosis is the <b>echocardiogram</b></p>	
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- Once a **screen positive** result is obtained, the cause for the difference in saturations or low saturation must be determined.
- A screen positive is an indication for an urgent consultation by a physician (or NP if you have one) for further investigation.
- Screen positives at home should follow the usual protocol for referral to physician as appropriate for the clinical picture of the baby.
- physical exam by physician should include a four limb BP, femoral pulses, full vital signs and pre and post ductal saturations.
- Also consider an ECG, chest X-Ray and rule out other non-cardiac causes. If cardiac diagnosis cannot be confidently ruled out, consultation with a paediatric cardiologist or paediatrician/neonatologist for further investigation would be advisable.
- The gold standard for cardiac assessment is an ECHOCARDIOGRAM.
- The screener will complete the CCHD portion of the DBS card appropriately and forward to NSO.

Slide 20

Remember

A **screen positive** does not necessarily mean the baby has CCHD.  
It indicates a need for further assessment.

- A screen positive does not necessarily mean the baby has CCHD.
- It indicates a need for further assessment.


Slide 21

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.

Indicate **screened** spot and no CCHD screen done here.




- The documentation of CCHD screening is completed on the yellow form, the last page of the newborn screening form.
- Every baby should have a CCHD screen result OR a reason the screen was not done

Slide 22

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

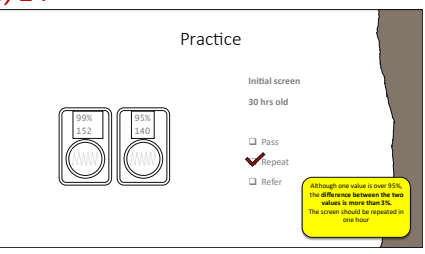
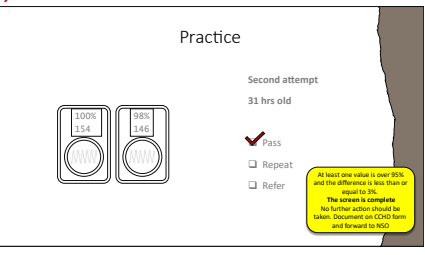
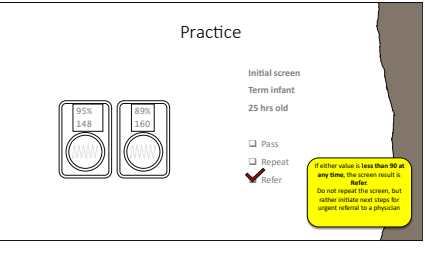
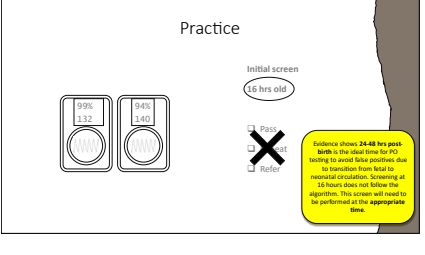
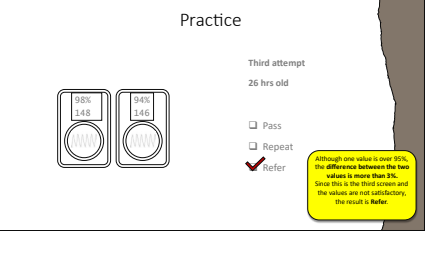
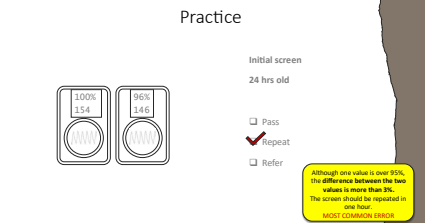
Unsatisfactory screens and referrals (screen positives) are identified.

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.



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



<p><b>Slide 23,24</b></p> 	<ul style="list-style-type: none"> <li>Review this using the algorithm/chart</li> </ul>
<p><b>Slide 25,26</b></p> 	<ul style="list-style-type: none"> <li>Review this using the algorithm/chart</li> </ul>
<p><b>Slide 27,28</b></p> 	<ul style="list-style-type: none"> <li>Review this using the algorithm/chart</li> </ul>
<p><b>Slide 29,30</b></p> 	<ul style="list-style-type: none"> <li>Review this using the algorithm/chart</li> <li><b>Please note with this example attention to the timing of the screen</b></li> <li>Screens performed before the 24-hour mark have a much higher false positive rate</li> </ul>
<p><b>Slide 31,32</b></p> 	<ul style="list-style-type: none"> <li>Review this using the algorithm/chart</li> </ul>
<p><b>Slide 33,34</b></p> 	<ul style="list-style-type: none"> <li>Review this using the algorithm/chart</li> <li><b>This is the most common error in evaluating CCHD screens. This is not a pass.</b></li> </ul>



Slide 35,36

Practice

Initial screen  
25 hrs old

95%	126	92%	136
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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.

- Review this using the algorithm/chart
- Although this result meets the criteria for a CCHD Screen pass, the saturation values are not ideal for a well baby.
- **This is the lowest result that is still considered a pass.**
- **It is important to pay attention to the clinical picture of this baby.**

Slide 37

#Goals

Standardization  
Access  
Quality

- NSO oversees the CCHD screening program for Ontario so that every baby has the same opportunity for quality screening, based on best practice standards.

Slide 38

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 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.

Slide 39

The End.

Questions?