Title: Critical Congenital Heart Disease Pulse Oximetry Screening – Screen Positive Workup Protocol

Purpose:

- To provide a guideline for evaluation and management to those performing Newborn Screening Ontario (NSO) Critical Congenital Heart Disease (CCHD) Pulse Oximetry Screening who obtain a screen positive result.

Scope or Principle:

This protocol provides a guideline for evaluation and management for Health Care Providers (HCPs) who obtain a **screen positive** CCHD screen result. A screen positive is when

1. The pulse oximetry is less than 90% in the right hand or either foot
   OR
2. The pulse oximetry is less than 95% on both extremities OR pulse oximetry difference in oxygen saturation is greater than 3% between the right hand and either foot **for 3 consecutive measures each separated by 1 hour.**

Background:

CCHD screening in asymptomatic newborns can assist in the early identification and treatment of CCHD, resulting in better outcomes for affected babies. If a screen positive result is obtained, an urgent referral or consultation to a physician is warranted. Follow-up information collected by NSO after receiving record of a screen positive result can assist in tracking the effectiveness of the screening program.

Responsibility:

All HCPs caring for newborns including but not limited to nurses and midwives, during the first days of life should be familiar with the protocol.

Definitions/Acronyms:

- NSO = Newborn Screening Ontario
- CCHD = Critical Congenital Heart Disease
- HCP = Health Care Provider
- DBS card = Dried Blood Spot card

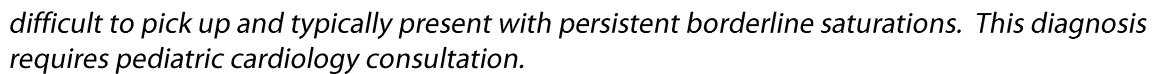
Related Documentation:

- Newborn Screening Ontario Critical Congenital Heart Disease Pulse Oximetry Screening Protocol
- Newborn Screening Ontario Critical Congenital Heart Disease Pulse Oximetry Screening – Community Screening Protocol (Non-Hospital Setting)

Protocol:

Submitter

1. Make an urgent referral to a physician upon identification of a screen positive result for assessment (recommend assessment to be within 6-8 hours).
2. Physical exam by a physician should include a 4 limb BP, femoral pulses, full vital signs and pre and post ductal saturations. Also consider an ECG and chest X-Ray to rule out other non-cardiac causes of cyanosis. If cardiac diagnosis cannot be confidently ruled out, consultation with a paediatric cardiologist or paediatrician/neonatologist for further investigation is advisable (Appendix A). *Please note some diagnoses of CCHD, specifically TAPVR, can be*



difficult to pick up and typically present with persistent borderline saturations. This diagnosis requires pediatric cardiology consultation.

3. Document the pulse oximetry values and the evaluated screen result in the infant's chart or medical record, as well as on the CCHD screen portion of the DBS card. Send the completed CCHD screen result page to NSO.

Newborn Screening Ontario

1. When a record of a screen positive result is received by NSO, a representative from NSO will contact the appropriate organization involved in the newborn's care to follow up.
2. The Diagnostic Evaluation Report Form (DERF) (see Appendix B) will be completed, documenting the interventions and outcome of the investigation (as information is available), and the file will be closed. This will complete the screening process.

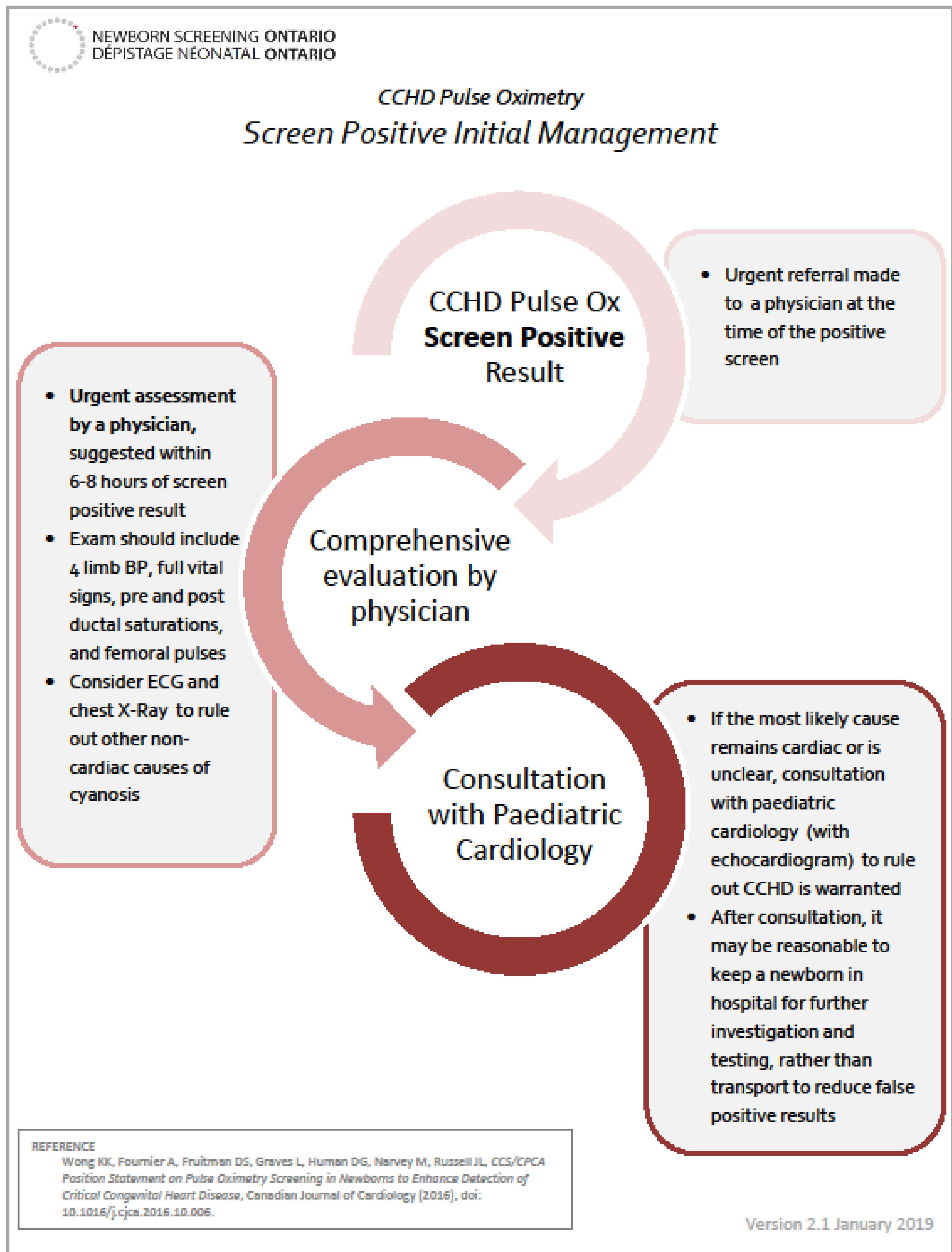
References:

- 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.aspxCCS 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.

Reviewed by:

- CCHD Disease Specific Working Group (2016/09; 2019/05)
- CCHD Midwifery Task Force (2016/10)
- CCHD Hospital Advisory Group (2016/11)

Appendix A



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CCHD Diagnostic Evaluation Report Form

Last Name: ____________ **First Name:** ____________

Health Card Number:		DOB:	
Episode Number:	NSO	Time of Birth:	
CCHD Form Number		Date of screen:	
Submitting Facility:		Time of screen(s):	
Screen performed by:		Age at first screen:	
Screen Positive for:	CCHD	Sex:	Select

Screen Results:

Initial Screen	SpO_2 R Hand		%
	SpO_2 Foot		%
First Repeat	SpO_2 R Hand		%
	SpO_2 Foot		%
Second Repeat	SpO_2 R Hand		%
	SpO_2 Foot		%

Was the infant symptomatic: ☐ Yes ☐ No ☐ Unknown

Was the baby referred based on (check all that apply): ☐ Screening results ☐ Clinical status

Referral within 6-8 hours? ☐ Yes ☐ No Assessment started within 6-8 hours? ☐ Yes ☐ No

✓ **Decision for care** *(check all that apply and complete relevant information)*:

✓			
☐	Infant brought to hospital	Date/Time of admission:	
☐	Care provided in hospital, no transfer	Date/time of decision:	
☐	Transfer within same hospital	Date/time of transfer:	
☐	Transfer to another hospital	Date/time of transfer:	

If transferred, specify transfer to:

Transported by (check one): ☐ Transport team ☐ Parents/Guardians
☐ Ambulance w/o transport team ☐ Other, specify: ____________

Diagnostic Evaluation *(check all that apply and complete relevant information)*:

		Date/time of referral (YYYY/MM/DD HHMM)	Date/time of assessment (YYYY/MM/DD HHMM)	Name of Practitioner
☐	Nurse practitioner			
☐	Family doctor			
☐	Emergency physician			
☐	Paediatrician/Neonatologist			
☐	Paediatric cardiologist			

Investigations *(check all that apply and complete relevant information)*:

✓	Investigation	Date	Findings/Comments
☐	4 limb BP		
☐	EKG		
☐	Chest XRAY		
☐	Physical Examination		
☐	Pre and Post Ductal Pulse Ox		

☐ **Echocardiogram** **Date:** ______ **Findings:** ______

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NEWBORN SCREENING ONTARIO
DÉPISTAGE NÉONATAL ONTARIO

CHEO
NSO is located at the Children's Hospital of Eastern Ontario

CCHD Diagnostic Evaluation Report Form

✓ **Interventions** *(check all that apply)*:

- ☐ Oxygen
- ☐ Prostaglandin infusion
- ☐ Non-invasive positive pressure ventilation
- ☐ Intubation and ventilation
- ☐ IV antibiotics
- ☐ Monitoring
- ☐ None

✓ **Definitive Diagnosis** *(complete date and check one)*:

Date diagnosis made:

Primary targets	Incidental Findings: Secondary targets (Classic)	Incidental Findings: Secondary targets (Untargeted disease)
☐ *Hypoplastic left heart syndrome*	☐ *Coarctation of the aorta*	*Cardiac, Other (specify):* ☐ *structural defect* ☐ *arrhythmia* ☐ *other*
☐ *Pulmonary atresia w/ intact septum*	☐ *Double outlet right ventricle*	
☐ *Tetralogy of Fallot*	☐ *Ebstein anomaly*	☐ *Other, specify: (ventricular hypertrophy, PDA, thrombosis,)*
☐ *Total anomalous pulmonary venous return*	☐ *Interrupted aortic arch*	
☐ *Truncus arteriosus*	☐ *Single ventricle*	☐ ***No disease identified*** *Persistent low saturations, no definitive diagnosis*
☐ *Transposition of the great arteries*	☐ *Pulmonary disease, non infectious*	**Other**
☐ *Tricuspid atresia*	☐ *Infection (eg sepsis, pneumonia)*	☐ ***No disease identified*** *Normal exam, normal saturations*
	☐ *Persistent Fetal Circulation (includes pulmonary hypertension, delayed transition)*	***Infant deceased prior to diagnosis*** *Date of death:* ________ *Cause of death:*
	☐ *PPHN*	

✓ **Plan for Care** *(complete date and check one)*:

Date of Plan of Care decision:

- ☐ Discharge
- ☐ Continue to follow with no treatment
- ☐ Treatment recommended or initiated
  - ☐ Surgical
  - ☐ Medicine
  - ☐ Other:

Comments:			
FORM COMPLETED BY:		**Date:**	
Contact Number:			

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