MICHIGAN DEPARTMENT OF COMMUNITY HEALTH Pulse Oximetry Screening for Critical Congenital Heart Disease in the HOME BIRTH COMMUNITY



Michigan Department of Community Health







2014

Critical Congenital Heart Disease Home Birth Toolkit for Implementing Pulse Oximetry Screening

Table of Contents:

Program Overview & CCHD Screening

- Introduction
- MDCH CCHD -Contact Information
- Pulse Oximetry Probe Placement & Equipment
- Pulse Oximetry "Do's & Don'ts"
- Michigan CCHD Screening Algorithm
- Screener's Tip Sheet
- Interpretation of Screening Results Graphic
- Failed CCHD Screen Follow- Up
- Letter for Emergency Room Physicians
- Submitting CCHD Screening Data

Information For Parents

- Parent Education Checklist
- Parent Education Materials
- Resources for Parents

Resources

- CCHD Overview of Defects
- Resources for Providers

CCHD Home Birth Toolkit is available for download on our website www.michigan.gov/cchd

Disclaimer:

The information included in this document is for informational and educational purposes only. The contents of this toolkit should not substitute for professional judgment, nor should the user rely solely on the information provided. Furthermore, this document does not reflect the optimal medical practice for all circumstances. Users are advised to seek professional counsel on the issues raised by consulting with medical staff on matters involving clinical practice.

Cover images courtesy of © Masimo Corporation 2011.

Program Overview and CCHD Screening



MICHIGAN CCHD SCREENING

Critical Congenital Heart Disease (CCHD) detected by pulse oximetry

Mo det	st likely to be ected:	Les: det	s likely to be ected:
0	Hypoplastic left heart syndrome	0	Coarctation of the aorta
0	Pulmonary atresia (with intact septum)	0	Double-outlet right ventricle
0	Tetralogy of Fallot	0	Ebstein anomaly
0	Total anomalous pulmonary venous return	0	Interrupted aortic arch
0	D-Transposition of the great arteries	0	Single ventricle
0	Tricuspid atresia		
0	Truncus arteriosus		

Congenital heart defects are the most common group of birth defects, affecting 9 in 1,000 newborns. *Critical* Congenital Heart Disease (CCHD) include those requiring surgery or catheter intervention in the first month of life. CCHD remains one of the most significant causes of infant death in the United States.

In 2011, pulse oximetry was recommended by the U.S. Department of Health and Human Services Secretary's Advisory Committee on Heritable Disorders in Newborns and Children as an important screening tool for detection of CCHD in asymptomatic newborns. This recommendation was subsequently endorsed by the American Academy of Pediatrics as a standard of care. As of April 1, 2014, Michigan requires screening for CCHD using pulse oximetry is required for all newborns.

This toolkit serves as a guide to help midwives and home birth attendants establish their own policies and procedures for implementing a CCHD Screening Program.

MICHIGAN CCHD SCREENING PROGRAM CONTACTS

Karen Andruszewski, BS Newborn Screening Data Manager 517-335-8532 andruszewskik@michigan.gov

Janice Bach, MS, CGC CCHD Program Director 517-335-8497 <u>bachj@michigan.gov</u>

Joan Ehrhardt, MS, CGC Birth Defects Program Coordinator 517-335-6498 <u>ehrhardtj@michigan.gov</u> Mary Kleyn, MSc Newborn Screening Epidemiologist 517-335-9296 <u>kleynm@michigan.gov</u>

Keri Urquhart, MPH, BSN, RN CCHD Nurse Educator 517-335-8135 <u>urquhartk1@michigan.gov</u>

William Young, PhD Newborn Screening Follow-up Manager 517-335-8938 youngw@michigan.gov



MICHIGAN CCHD SCREENING PULSE OX PROBE PLACEMENT

PULSE OX PROBE PLACEMENT EDUCATION

1. Select application site on the outside, fleshy area of the infant's right hand and one foot.



RH Application Site



Foot Application Site

- 2. Place the photo detector portion of the probe on the fleshy portion of the outside of the infant's right hand or foot.
- 3. Place the light emitter of the probe on the top of the right hand or foot.
- 4. Remember the photo detector and light emitter must be directly opposite each other in order to obtain an accurate reading.
- 5. Secure the probe to the infant's right hand or foot using the adhesive or foam tape recommended by the vendor. It is not recommended to use tape to secure probe placement.





CCHD Screening should be done:

- On an infant without respiratory or cardiovascular distress
- As close to 24 hours of life as possible
- When infant is awake, comfortable, and quiet (ok for parent to hold)

MICHIGAN CCHD SCREENING PULSE OX EQUIPMENT

Use only appropriate infant probes for CCHD Screening. Use a pulse oximeter that is FDA approved for use in neonates.

Pulse oximetry equipment used for CCHD Screening:

- Must be approved by the FDA for use in newborns.
- Must be validated in lowperfusion conditions.
- Must have 2% root, meansquare accuracy.
- Must be calibrated regularly, based on manufacturer guidelines.



FDA CLEARANCE FOR PULSE OXIMETER USE IN NEONATES

http://www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfpmn/pmn.cfm

MICHIGAN CCHD SCREENING PERFORMING PULSE OX- DOS & DON'TS

PULSE OX DO's

- 1. If you are using disposable pulse ox probes, use a new, clean probe for each infant. If you are using reusable pulse ox probes, clean the probe with recommended disinfectant solution between each infant. Dirty probes can decrease the accuracy of your reading and can transmit infection. A disposable wrap can be used to secure the probe to the site.
- 2. 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.
- 3. When placing the sensor on the infant's skin, there should not be gaps between the sensor and the skin. The sides of the probe should be directly opposite each other.
- 4. Nail polish dyes and substances with dark pigmentation (such as dried blood) can affect the pulse ox reading. Assure that the skin is clean and dry before placing the probe on the infant. Skin color and jaundice do not affect the pulse ox reading.
- 5. Movement, shivering and crying can affect the accuracy of the pulse ox reading. Ensure that the infant is calm and warm during the reading. Swaddle the infant and encourage family involvement to promote comfort while obtaining the reading. If possible, conduct screening while the infant is awake.
- 6. Pulse oximeters have different confidence indicators to ensure that the pulse ox reading is accurate. Determine the confidence indicators for the pulse oximeter you are using.





MICHIGAN CCHD SCREENING PERFORMING PULSE OX- DOS & DON'TS

PULSE OX DON'Ts

- 1. Never use an adult pulse ox clip to obtain a reading for an infant. Using an adult clip on an infant will produce inaccurate results.
- 2. Blood flow is needed to obtain an accurate pulse ox reading. Never attempt to obtain a pulse ox reading on the same extremity where an automatic blood pressure cuff is located.
- 3. Bright or infrared light, including bilirubin lamps and surgical lights, can affect the accuracy of the reading. Ensure that the infant is not placed in bright or infrared light while pulse ox is being performed. You may cover the pulse ox probe with a blanket to ensure that extraneous light does not affect the accuracy of your reading.
- 4. Do not use tape to apply the pulse ox probe to the infant's skin.

PULSE OX CAUTION

- A pulse is needed to determine the oximetry reading. Pulse ox is not accurate if the patient is coding or has a cardiac arrhythmia. Remember: No pulse, no oximetry!
- 2. Pulse ox readings are not instantaneous. The oximetry reading that is displayed on the monitor is an average of readings over the past few seconds.











MICHIGAN CCHD SCREENING SCREENER TIP SHEET



Michigan Newborn Screening Tip Sheet

MICHIGAN DEPARTMENT OF COMMUNITY HEALTH

CCHD Screening for Newborn without Cardiovascular or Respiratory Distress Interpretation of Pulse Oximetry Results

Oxygen Saturation

(02 Sats; %)

Right Hand (RH) Either Foot (F) Right Hand (RH) Either Foot (F) 100 100 99 98 97 96 92 91 90 89% or lower 99 100 99 98 97 96 93 91 90 89% or lower 99 100 99 98 97 96 95 94 93 92 91 90 89% or lower 91 100 99 98 97 96 95 94 93 92 91 90 89% or lower 92 100 99 98 97 96 93 92 91 90 89% or lower 93 100 99 98 97 96 93 92 91 90 89% or lower 93 100 99 98 97 96 93 92 91 90 89% or lower 93 93 92 91	CIPC 70	ler '							3					
100 100 99 97 96 95 94 93 92 91 90 89% or lower 99 100 99 97 96 95 94 93 92 91 90 89% or lower 95 100 99 98 97 96 93 92 91 90 89% or lower 95 100 99 98 97 96 93 92 91 90 89% or lower 95 100 99 98 97 96 95 94 93 92 91 90 89% or lower 93 100 99 98 97 96 95 94 93 93 93 93% or lower 93 100 99 98 97 96 93 92 91 90 89% or lower 90 100 99 98 97 96 93 92 91	Sup	Right Hand (RH)					Either Fi	oot (F)	-					
99 100 99 97 96 95 94 93 92 91 90 89% or lower 97 100 99 98 97 96 93 92 91 90 89% or lower 96 100 99 98 97 96 93 92 91 90 89% or lower 95 100 99 98 97 96 93 92 91 90 89% or lower 95 100 99 98 97 96 93 92 91 90 89% or lower 93 100 99 98 97 96 93 92 91 90 89% or lower 93 100 99 98 97 96 93 92 91 90 89% or lower 90 100 99 98 97 96 93 92 91 90 89% or lower 90<	3	100	100	66	86	97	96	95	94	93	92	91	6	89% or lower
98 100 99 97 96 55 94 93 92 91 90 89% or lower 97 100 99 98 97 96 93 92 91 90 89% or lower 95 100 99 98 97 96 93 92 91 90 89% or lower 95 100 99 98 97 96 95 94 93 92 91 90 89% or lower 93 100 99 98 97 96 95 94 93 92 91 90 89% or lower 93 100 99 98 97 96 95 94 93 92 91 90 89% or lower 90 100 99 98 97 96 95 94 93 92 91 90 89% or lower 90 100 99 98 97		66	100	66	86	97	96	95	94	93	92	91	90	89% or lower
97 100 99 97 96 93 92 91 90 89% or lower 96 100 99 98 97 96 93 92 91 90 89% or lower 95 100 99 98 97 96 93 92 91 90 89% or lower 93 100 99 98 97 96 95 94 93 92 91 90 89% or lower 93 100 99 98 97 96 95 94 93 92 91 90 89% or lower 90 100 99 98 97 96 95 94 93 92 91 90 89% or lower 90 93 97 96 95 94 93 92 91 90 89% or lower 90 93 92 91 93 92 91 90 89% or lower </td <td></td> <td>98</td> <td>100</td> <td>66</td> <td>98</td> <td>97</td> <td>96</td> <td>95</td> <td>94</td> <td>93</td> <td>92</td> <td>91</td> <td>60</td> <td>89% or lower</td>		98	100	66	98	97	96	95	94	93	92	91	60	89% or lower
96 100 99 97 96 93 92 91 90 89% or lower 93 100 99 98 97 96 93 92 91 90 89% or lower 93 100 99 98 97 96 95 94 93 92 91 90 89% or lower 93 100 99 98 97 96 95 94 93 92 91 90 89% or lower 90 100 99 98 97 96 95 94 93 92 91 90 89% or lower 90 100 99 98 97 96 95 94 93 92 91 90 89% or lower 90 90 90 93 92 91 90 89% or lower 90 90 93 93 93 92 91 90 89% or lower <t< td=""><td></td><td>67</td><td>100</td><td>66</td><td>86</td><td>97</td><td>96</td><td>95</td><td>94</td><td>93</td><td>92</td><td>91</td><td>06</td><td>89% or lower</td></t<>		67	100	66	86	97	96	95	94	93	92	91	06	89% or lower
95 100 99 98 97 96 95 91 90 89% or lower 93 100 99 98 97 96 95 94 90 89% or lower 92 100 99 98 97 96 95 94 93 92 91 90 89% or lower 92 100 99 98 97 96 95 94 93 92 91 90 89% or lower 90 100 99 98 97 96 95 94 93 92 91 90 89% or lower 90 100 99 98 97 96 95 94 93 92 91 90 89% or lower 100 99 98 97 96 95 94 93 92 91 90 89% or lower 101 91 96 95 94 93 92		96	100	66	86	67	96	95	94	93	92	91	06	89% or lower
94 100 99 97 96 95 94 93 92 91 90 89% or lower 93 100 99 97 96 95 94 93 92 91 90 89% or lower 91 100 99 98 97 96 95 94 93 92 91 90 89% or lower 90 100 99 98 97 96 95 94 93 92 91 90 89% or lower 90 100 99 98 97 96 95 94 93 92 91 90 89% or lower 89% or lower 100 99 98 97 96 95 94 93 92 91 90 89% or lower 100 99 98 97 96 95 94 92 91 90 89% or lower 100 90 98 97		95	100	66	86	67	96	95	94	93	92	91	90	89% or lower
93 100 99 97 96 95 94 93 92 91 90 89% or lower 92 100 99 98 97 96 95 94 93 92 91 90 89% or lower 91 100 99 98 97 96 95 94 93 92 91 90 89% or lower 90 100 99 98 97 96 95 94 93 92 91 90 89% or lower Nower 100 99 98 97 96 95 94 93 92 91 90 89% or lower Pass 95% or higher in right hand (RH) or either foot (F) AND difference of 3% or less between RH and F. 5 94 93 92 91 90 89% or lower Pass 95% or higher in right hand (RH) or either foot (F) AND difference of 3% or less between RH and F. 5 5 94 93 92 91 90 89% or lower Pass 95% or higher in right hand F. 6 95 94 <t< td=""><td></td><td>94</td><td>100</td><td>66</td><td>86</td><td>97</td><td>96</td><td>95</td><td>94</td><td>93</td><td>92</td><td>91</td><td>60</td><td>89% or lower</td></t<>		94	100	66	86	97	96	95	94	93	92	91	60	89% or lower
92 100 99 97 96 95 94 93 92 91 90 89% or lower 91 100 99 98 97 96 95 94 93 92 91 90 89% or lower 90 100 99 98 97 96 95 94 93 92 91 90 89% or lower 89% or lower 100 99 98 97 96 95 94 93 92 91 90 89% or lower Noveloation of streen 100 99 98 97 96 95 94 93 92 91 90 89% or lower Participation of streen 90-94% in RH and F 64 93 92 91 90 89% or lower 89% or lower in RH or F (at any time) 89% or lower in RH or F (at any time) 35% or more between RH and F. Screen PL or 31 in s. Areen: 90-94% in RH and F. Screen RH and F. Screen PL or 31 in s. 31d screen: 90-94% in RH and F or difference of 4% or more between RH and F. Screen PL or 31 in s. Screen PL or 31 in s.		93	100	66	86	97	96	95	94	63	92	91	06	89% or lower
91 100 99 97 96 95 94 93 92 91 90 89% or lower 90 100 99 98 97 96 95 94 93 92 91 90 89% or lower 89% or lower 100 99 98 97 96 95 94 93 92 91 90 89% or lower Pass 95% or lower 100 99 98 97 96 95 94 93 92 91 90 89% or lower Provide the state of the st		92	100	66	86	67	96	95	94	63	92	91	06	89% or lower
90 100 99 97 96 95 94 93 92 91 90 89% or lower NMMM 89% or lower 100 99 98 97 96 95 94 93 92 91 90 89% or lower NMMM Pass 97 96 95 94 93 92 91 90 89% or lower NMMM Pass 95% or light in right hand (RH) or either foot (F) AND difference of 3% or less between RH and F. Screen 90-94% in RH and F or difference of 4% or more between RH and F. Screen up to 3 times. B9% or lower in RH or F (at any time) 5 OR Advice between RH and F. Screen up to 3 times. OR Advice of 4% or more between RH and F. OR OR OR 93% or lower in RH or F (at any time) OR OR OR OR 93% or lower <td></td> <td>16</td> <td>100</td> <td>66</td> <td>86</td> <td>67</td> <td>96</td> <td>95</td> <td>94</td> <td>93</td> <td>92</td> <td>91</td> <td>90</td> <td>89% or lower</td>		16	100	66	86	67	96	95	94	93	92	91	90	89% or lower
89% or lower 100 99 97 96 95 94 93 92 91 90 89% or lower Pass 95% or higher in right hand (RH) or either foot (F) AND difference of 3% or less between RH and F. 90-94% in RH and F. 90-94% in RH and F. 90 89% or lower Bass 90-94% in RH and F. 90-94% in RH and F. Screen up to 3 times. 91 90 89% or lower Bass 89% or lower in RH or F (at any time) 0 89% or lower in RH or F (at any time) 31d screen. 90-94% in RH and F. Screen up to 3 times.		06	100	66	98	97	96	95	94	63	92	91	6	89% or lower
With the set of the set		89% or lower	100	66	86	67	96	95	94	93	92	91	90	89% or lower
With and F or difference of 4% or more between RH and F. Screen up to 3 times. 89% or lower in RH or F (at any time) 89% or lower in RH or F (at any time) 3rd screen: 90-94% in RH and F or difference of 4% or more between RH and F. Screen up to 3 times.		Availa	95% 0	or higher	in right !	hand (RH)	or eithe	er foot (F)	AND dif	ference o	f 3% or I	ess betw	een RH	and F.
screen 90-94% in RH and F or difference of 4% or more between RH and F. Screen up to 3 times. 89% or lower in RH or F (at any time) and screen: 90-94% in RH and F or difference of 4% or more between RH and F.	.mi	ble												
89% or lower in RH or F (at any time) 0R 3rd screen: 90-94% in RH and F or difference of 4% or more between RH and F.	ichi	e fo	76-06	1% in RH	and F or	different	ce of 4%	or more	between	RH and F		Screen u	up to 3 ti	mes.
provide and For difference of 4% or more between RH and F.	gan	r do wel	89%	or lower	in RH or	F (at any	time							
ad	.gov/	ownlo psite	3rd s	OR creen: 90	-94% in	RH and F	or differ	ence of 4	% or mot	te betwe	en RH an	đF		
		ad ccl												

MICHIGAN CCHD SCREENING INTERPRETATION OF PULSE OXIMETRY RESULTS

1-May-13 MDCH

-Journal of Neonatology Research 2012(2)2: 96-101.

Lakshmins

MICHIGAN CCHD SCREENING-HOME BIRTHS FAILED CCHD SCREEN- FOLLOW-UP



Updated October 2014

MICHIGAN CCHD SCREENING-HOME BIRTHS FAILED CCHD SCREEN- FOLLOW-UP LETTER FOR EMERGENCY ROOM PHYSICIANS

This letter should be sent with a parent of newborn that failed the CCHD Screen and is being taken to an Emergency Room for follow-up



RICK SNYDER GOVERNOR STATE OF MICHIGAN DEPARTMENT OF COMMUNITY HEALTH LANSING

NICK LYON

on our website www.michigan.gov/cchd

Newborn Screening Program for Critical Congenital Heart Disease

Michigan Hospital Emergency Room Provider:

This infant was born at home on ___/__/

The newborn is presenting in your Emergency Department for follow-up because at approximately twenty-four hours of life he/she was screened for Critical Congenital Heart Disease (CCHD) using pulse oximetry and did not pass the screening.

The screening results were:

Date/Time of Screen	Right Hand (RH)	Foot	Difference	Result (Pass, Fail, Rescreen)
	_	1		-
		20 20 20		

As part of your initial triage on the infant we are asking that you re-check the pulse ox on the infant's **Right Hand** and **One Foot**. Using the limits defined by the algorithm (see attached), determine if the infant needs further follow-up (pulse ox of 94% or below, or a difference of 4% or greater between the readings).

The Michigan public health code mandates that all newborns be screened for CCHD using pulse oximetry as close to twenty-four hours of life as possible. Infants should be screened using the Michigan recommended screening algorithm.

Infants who fail the screen in the hospital are immedialty referred for an evaluation, often including follow up with a pediatric cardiologist/neonatologist and a possible echocardiogram for diagnosing CCHD.

If follow up is needed, upon admitance to the emergency room please consider consulting a pediatric cardiologist or neonatologist.

During the hours of 7 am-7 pm you can contact a representative from the MDCH Newborn Screening program with questions @ 517-242-0529.

More information about the screening is available at our website <u>www.michigan.gov/cchd</u>

Keri Urquhart, RN, BSN CCHD Nurse Educator

MICHIGAN CCHD SCREENING DATA REPORTING-HOME BIRTHS

CCHD Screening data reporting can be done in the homebirth community using one of the following methods:

1. CCHD Data Reporting Form for Homebirths

This paper form (available on our website) can be submitted by mail or fax to MDCH. To request blank forms be sent to you by mail, please call 1.866.673.9939.

ewborn Screening Kit Number	Midwife/Birth Attendant	
aby's First Name B	aby's Last Name	Birth Order 🛛 🗸 B, C, D
aby's Date of Birth	Baby's Medical Record Number	
lother's First Name	Mother's Last Name	
Pulse Ox Reading 1	Pulse Ox Reading 2	Pulse Ox Reading 3
Date Time	Date Time	Date Time
Perfusion	Perfusion	Perfusion
Index	Index	Index
Foot Sat%	Foot Sat%	Foot Sat%
Right Hand Sat%	Right Hand Sat%	Right Hand Sat%
Difference	Difference	Difference
Outcome* Rescreer	Outcome*	Outcome*
Fail		
Completed	Completed	Completed
ther Reason	Other Reason	Other Reason
ot Completed	Not Completed	Not Completed
eason Not Completed: Cardiac/Respiratory	Distress, Transfer, Parent Refusal, Death, Prior post	tnatal diagnosis of CCHD, Referred
follow-up Diagnose	d prenatally	downoo

2. Online eReports Module

This online reporting system requires a secure log-in from the State of Michigan. Birth attendants will be able to sign into the module and enter individual screening results. User manual and more information are available on the website, or for more information and to request access please call 1.866.673.9939.

Information For Parents



"As a mother of a child born with a CCHD I am thankful there is a screening tool for newborns. Just knowing that children just like my daughter can be given a chance at life by catching their defects soon after birth shows the importance of screening. It gives these children their chance to fight and early diagnosis is key."

> Jenny Lincoln Mom to Aly Jean and Parent Advocate for CCHD screening www.alyjeansspecialheart.com

MICHIGAN CCHD SCREENING CHECKLIST FOR INFORMING PARENTS

INFORM THE PARENT:

- The purpose of screening is to detect serious heart defects in babies.
- The baby will be screened after 24 hours of life (or as close to as possible).
- The pulse ox test will be done on the baby's right hand and one foot, if possible.
- The pulse ox test is not painful and only takes a few minutes when the baby is quiet, warm, and not moving.

- It is possible that a baby with a heart problem may have a normal pulse ox reading.
- They have the right to decline screening.
- They may ask questions at the time of screening or anytime before or following the screening.

MICHIGAN CCHD SCREENING PARENT FACT SHEETS

Newborn Heart Pulse Oximetry Screening For Critical Congenital Heart Disease (CCHD)

The goal of newborn screening is to find babies who have serious medical conditions that need urgent treatment. Most babies are born with no serious health problems. But for those who do have a serious health problem, newborn screenin

Educational Materials are available in bulk supply from MDCH by request.

What is CCHD?

Critical congenital heart disease (CCHD) occurs not form properly. There are many types of he "critical" heart defect is one that needs urgen the baby. Treatment can include medical and

Why screen for CCHD?

Most babies are born with healthy hearts. carefully by a doctor before they go home until later. Screening can find babies with

How is screening for CCHD don

Pulse oximetry is used to screen babies A small sensor is placed on a baby's rig blood. Your baby's doctor or nurse wil done for well newborns sometime du baby is warm and quiet. If a baby is c longer. It may need to be repeated.

What happens if CCHD scre

Most babies will have enough oxy may have CCHD. Other condition to have a low blood oxygen level. will check the baby carefully. An "heart echo") is done to look for hospital or doctor's office. It is heart echo shows problems, th

What should parents v

Most babies who pass the pu parents to know that newb signs that all parents should breathing, poor feeding, ar please contact your baby

> For question Michigan Ne Toll-free: 1 newbornscr Michigan N

Newborn Pulse Oximetry Screening For Critical Congenital Heart Disease (CC What does a low result mean?

What is a pulse oximetry screen? Pulse oximetry (also called "pulse ox") is a simple test to measure oxygen in the blood. Pulse ox is done using a small sensor placed on the baby's right hand and one foot. Pulse ox looks for low oxygen levels in the blood. Screening is a way to find critical heart conditions in newborns.

Your baby has a low pulse ox screen. Now what?

A low pulse oximetry result does not mean your baby has critical congenital heart disease (CCHD). It means more tests are needed.

A low blood oxygen level means that your baby

may have critical congenital heart disease (CCHD) or other problems. You will discuss plans for more testing with your baby's medical team. Different types of tests may be needed, for example:

- Heart ultrasound (also called
- "echocardiogram" or "heart echo") Blood tests
- Chest x-ray
- Tests for infection

What is a heart echo?

ichigan Newborn Screening Parent Fact Sheet A heart echo is an ultrasound of the heart. It uses sound to make a picture of the heart. The picture will be read by a doctor. You will discuss results and next steps with your baby's medical team. For the safety of your baby it is important to follow the medical team's instructions for testing and follow-up.

Michigan Resources and Support

For questions contact: Michigan Newborn Screening Program Toll-free: 1-866-673-9939 Email: newbornscreening@michigan.gov

Michigan Newborn Screening Follow-up, 201 Michigan Department of Co

What is CCHD?

Critical congenital heart disease (CCHD) that the heart or major blood vessels did form correctly before birth. The heart ma work well. A baby with CCHD may not ha enough oxygen in the blood. The body nee oxygen to grow and be healthy.

CCHD may or may not run in families.

What problems can CCHD cause? CCHD is different for each child. CCHD needs urgent care. Some babies with CCHD may

- Fast, difficult breathing
- Grunting sounds with breaths Bluish coloring of lips
- Poor eating
- Extreme sleepiness Heart murmur

What if my child has CCHD?

Children with CCHD should see their regular doctor and a doctor who specializes in children with heart disease (called a "pediatric cardiologist"). Your child's medical team will talk to you about treatment options, if needed.

Treatment for CCHD varies. It can include:

- Medicine
- Medical procedures Surgery

Prompt and careful treatment helps most children with CCHD live the healthies



MICHIGAN CCHD SCREENING SUGGESTED RESOURCES FOR FAMILIES

Support Groups and Online Resources

American Heart Association Children's Heart Association Children's Heart Foundation Congenital Heart Information Network Congenital Heart Defects.com Kids with Heart PediHeart 1 in 100 Mended Little Hearts Hearts of Hope

www.amhrt.org www.heartchild.info www.childrensheartfoundation.org www.tchin.org www.congenitalheartdefects.com www.kidswithheart.org www.pediheart.net www.1in100.org www.mendedlittlehearts./org www.heartsofhopemi.org

Books for Parents

"Heart Defects in Children: What Every Parent Should Know" by Cheryl J. Wild "Heart of a Child" by Catherine A. Neill, Edward B. Clark and Carleen Clark "It's My Heart" by Children's Heart Foundation "The Heart of a Mother" by Anna Marie Jaworski and Judy Norwood

Books for Children with Heart Defects

"Blue Lewis and Sasha the Great" by Carol Donsky Newell "Matty's Heart" by C.J. Hribal "Nathan's Special Heart" by Jessica Ennis (available as a virtual book) "Pump the Bear" by Gisella Olivo Whittington

Books for Siblings

"Cardiac Kids" by Vicci Elder "My Brother Needs an Operation" by Anna Marie Jaworski & Sarah Lualdi Moran "When Molly was in the Hospital" by Debbie Duncan





CRITICAL CONGENITAL HEART DEFECTS:

Pulse oximetry screening is **most likely** to detect seven of the CCHDs. These seven main screening targets are:

D-transposition of the Great Arteries

A heart in which the two main arteries carrying blood away from the heart are reversed. In a normal heart the blood flows in a cycle: bodyheart-lungs-heart-body. When a dtransposition occurs, the blood pathway is impaired because the two arteries are connecting to the wrong chambers in the heart.

Tetralogy of Fallot

A heart defect that features four problems. They are: a hole between the lower chambers of the heart; an obstruction from the heart to the lungs; the aorta (blood vessel) lies over the hole in the lower chambers; and the muscle surrounding the lower right chamber becomes overly thickened.

Total anomalous pulmonary venous return (TAPV)

A defect in the veins leading from the lungs to the heart. In TAPV, the blood does not take the normal route from the lungs to the heart and out to the body. Instead, the veins from the lungs attach to the heart in abnormal positions and this problem means that oxygenated blood enters or leaks into the wrong chamber.

Truncus Arteriosus

When a person has one large artery instead of two separate ones to carry blood to the lungs and body. In a normal heart, the blood follows in a cycle: body-heart-lungs-heart-body. When a person has a truncus arteriosus, the blood leaving the heart does not follow this path. It has only one vessel, instead of two separate ones for the lungs and body. With only one artery, there is no specific path to the lungs for oxygen before returning to the heart to deliver oxygen to the body.

Hypoplastic Left Heart Syndrome (HLHS)

An underdeveloped left side of the heart. The aorta and left ventricle are too small and the holes in the artery and septum did not properly mature and close.

Pulmonary Atresia

A non-existent pulmonary valve, so that the only blood receiving oxygen is the blood that is diverted to the lungs through openings that normally close during development.

Tricuspid Atresia

A missing tricuspid valve in the heart so blood cannot flow from the body into the heart in the normal way. The blood is not being properly refilled with oxygen and it does not complete the normal cycle of body -heart– lungs-heart-body.



These defects are **less likely** to be detected through pulse oximetry screening:

Coarctation of the Aorta

A narrowing of the major artery (the aorta) that carries blood to the body. This narrowing affects blood flow where the arteries branch out to carry blood along separate vessels to the upper and lower parts of the body. CoA can cause high blood pressure or heart damage.

Ebstein Anomaly

A malformed heart valve that does not properly close to keep the blood flow moving in the right direction. Blood may leak back from the lower to upper chambers on the right side of the heart. As a result, the right atrium becomes enlarged. If the tricuspid regurgitation (leak) is severe enough, congestive heart failure can result. This syndrome also is commonly seen with an atrial septal defect, or ASD (a hole in the wall dividing the two upper chambers of the heart).

Double-outlet right Ventricle

In double outlet right ventricle, something goes wrong during the formation of the heart and both great arteries are attached to the right ventricle. No arteries, or only a part of the aorta, are attached to the left ventricle. In some cases, because more blood than normal is flowing into the right ventricle, this heart defect means that too much blood is pumped to the lungs. Over time this uncontrolled flow can damage the lungs and heart, and heart failure can result.

Interrupted aortic arch

An absence or discontinuation of a portion of the aortic arch. IAA is classified by the site of the interruption, and is thought to be a result of faulty development of the aortic arch system during the fifth to seventh week of fetal development. This defect is almost always associated with a large ventricular septal defect (VSD).

Single ventricle

The term "single ventricle anomaly" is purposely non-specific. It is used to describe a group of cardiac defects that may differ quite dramatically from each other but share the common feature that only one of the two ventricles is of adequate functional size. Because of this feature, the ultimate plan for reconstruction is actually quite similar for most of these anomalies. All will generally undergo staged reconstructive procedures.



References:

American Heart Association. (2013). About Congenital Heart Defects. Available from

http://www.heart.org/HEARTORG/Conditions/CongenitalHeartDefects/A boutCongenitalHeartDefects

Cincinnati Children's Hospital. (2013). Heart Institute Encyclopedia: Congenital Heart Defects. Available from

**Images and Video of Defects available online from above resources. **



http://www.cincinnatichildrens.org/patients/child/encyclopedia/defects/default/

MICHIGAN CCHD SCREENING SUGGESTED RESOURCES FOR PROVIDERS

Michigan Department of Community Health

www.michigan.gov/cchd

American Academy of Pediatrics

www.aap.org AAP Strategies for Implementing Screening http://pediatrics.aappublications.or g/content/128/5/e1259.full.html

American Heart Association <u>www.aha.org</u>

Children's National Medical Center-Washington DC*

(Videos available for parents and providers) www.childrensnational.org

Children's Hospital of Philadelphia (CHOP)*

www.chop.edu

Atlanta Children's Hospital *

(CCHD Mobile App available) www.pulseoxtool.com

Cincinnati Children's Hospital * www.cincinnatichildrens.org

Center for Disease Control and Prevention <u>www.cdc.gov</u>

Congenital Heart Information Network* www.tchin.org

Baby's First Test* (Videos available for parents and providers) www.babysfirsttest.org

*Inclusion on the list does not necessarily imply endorsement, nor do we guarantee the accuracy of the information contained on these sites. Always consult your institution and its physicians with questions and concerns.

MICHIGAN Treatment Centers for CCHD:

Children's Hospital of Michigan#

DMC DETROIT MEDICAL CENTER

Children's Hospital of Michigan 3901 Beaubien Detroit, MI 48201 (313) 745-KIDS 1-888-362-2500 www.childrensdmc.org



Helen DeVos Children's hospital

OF SPECTRUM HEALTH

Helen DeVos Children's Hospital Congenital Heart Center 100 Michigan Street NE, Floor 10, Grand Rapids, Michigan 495031 616-267-9150 866.989.7999 www.helendevoschildrens.org



University of Michigan C.S. Mott Children's Hospital

Congenital Heart Center at C.S. Mott Children's Hospital 1540 East Hospital Drive Floor 11 Ann Arbor MI 48109 1-877-308-9111 www.mottchildren.org