

The Michigan Monitor

Following trends, promoting prevention
and linking families to resources

Fall 2013
Volume 6 Issue 2

Inside this issue:

| | |
|--|---|
| About congenital heart disease (CHD) | 1 |
| Screening for critical CHD (CCHD) in MI birthing hospitals | 2 |
| Michigan CCHD statistics | 2 |
| Sudden cardiac death of the young (SCDY) in MI | 3 |
| Mortality and CHD | 3 |
| MBDR quality assurance | 4 |

Congenital Heart Disease

Background

About 1 in 100 babies is born with congenital heart disease (CHD) making CHD the most common of all birth defects. This adds up to 40,000 newborns in the United States with CHD per year¹, with over 1,700 of those born in Michigan.² CHD accounts for 30% of deaths in infants with birth defects.¹



Critical Congenital Heart Disease (CCHD)

Approximately 25% of CHDs is considered **critical** because it requires intervention in the first year of life.³ CCHD may result in death or disability if not identified and treated soon after birth. Prenatal screening using ultrasound technology is an important tool for early detection of life-threatening heart disease. Alternatively, CCHD may be suspected during the postnatal period by clinical examination and/or by presentation of symptoms during the first 24 hours of life. However, both prenatal and postnatal evaluation have significant limitations in CCHD detection. As a result, affected but asymptomatic newborns may be discharged from the birthing facility with undiagnosed CCHD.⁴



Points of Interest

- ♥ Congenital heart disease (CHD) is the most common of all birth defects
- ♥ Many Michigan birthing hospitals use pulse oximetry to screen newborns for critical CHD
- ♥ CHD is one cause of sudden cardiac death in young people
- ♥ People with CHD are living longer thanks to better detection and treatment!

Pulse oximetry is a simple bedside test that measures the level of oxygen in the blood. Low blood oxygen in a newborn may be a sign of CCHD or other serious health problems. In 2011, the Secretary's Advisory Committee for Heritable Disorders in Newborns and Children recommended screening newborns for CCHD using pulse oximetry, as part of the Recommended Uniform Screening Panel. Following approval by the Secretary of the United States' Department of Health and Human Services and endorsement by the American Academy of Pediatrics, nearly all states have added CCHD screening, have legislation pending or have a pilot project in progress. Michigan began a CCHD newborn screening demonstration project in 2012. This summer, the MDCH Newborn Screening Quality Assurance Advisory Committee recommended adding CCHD to the panel of more than 50 disorders now screened in Michigan newborns. Legislative approval was received this fall. Statewide screening becomes effective April 1, 2014.

More About Newborn Screening for CCHD

Michigan— <http://www.michigan.gov/cchd/>
National— <http://1in100.org/> and <http://babysfirsttest.org/>

*Image compliments of Masimo Corporation.

Screening for CCHD in Michigan

The MDCH Newborn Screening (NBS) Follow-up Program surveyed all Michigan birthing facilities in order to learn how they may be addressing CCHD screening, identify barriers to implementation, and identify needs for training and technical assistance. Seventy-three (73) of 84 birthing facilities responded (86.9%). Screening status is shown in Figure 1. Six facilities, accounting for about 2800 of the 111,549 Michigan babies born in 2012, reported no plans to begin screening. The need to purchase new equipment was the most frequently reported barrier to screening. Figure 2. shows the clinical status and location of newborns at the time of screening.

Figure 1. Michigan birthing facility NBS CCHD screening status, 2012.

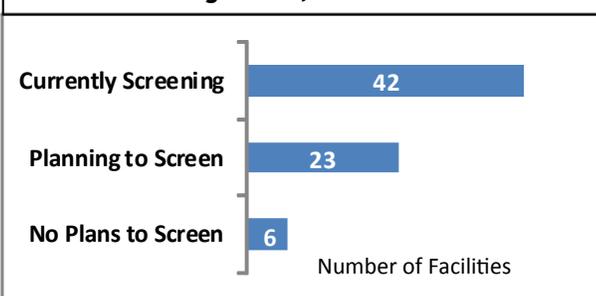
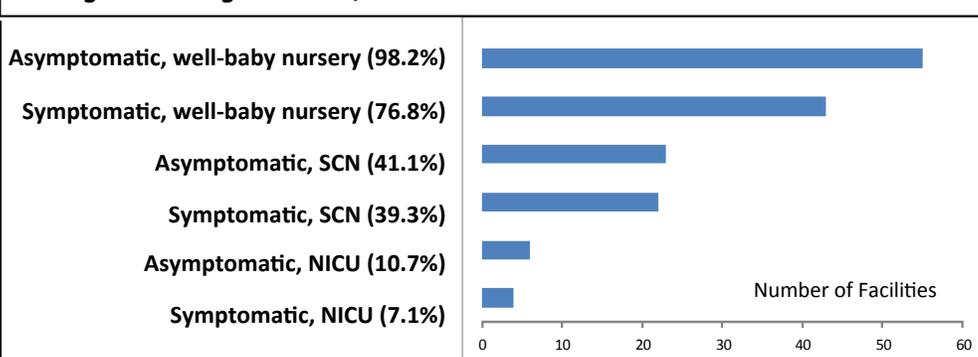


Figure 2. Clinical status and location of newborns screened for CCHD at Michigan birthing facilities, 2012.



Of the 65 facilities screening or planning to screen:

- ♥ Four (6%) had educational materials for parents if a newborn fails a screen.
- ♥ Two (3%) said they maintain a database of results. Neither could report the number of newborns screened, the number of failed screens and/or the number confirmed to have CCHD.

More hospitals have now implemented CCHD screening. Currently, nearly 89% of Michigan birthing facilities are screening or are planning to screen. The MDCH NBS Program works with the CCHD NBS Advisory Committee and supports the implementation of CCHD screening through development of a uniform screening algorithm, educational materials, reporting infrastructure, facility mini-grants and quality assurance measures.

Michigan CCHD Statistics, MBDR 2000-2010

According to Michigan Birth Defects Registry reporting, approximately 267 babies are born with CCHD each year in Michigan. Of the CCHD most likely to be identified with pulse oximetry screening, tetralogy of Fallot has the highest birth incidence per year. See below for Michigan estimates of CCHD occurrence by condition per year.

CCHD Detected by Screening

- ♥ **Common truncus (truncus arteriosus); n≈16**
- ♥ **Hypoplastic left heart syndrome; n≈47**
- ♥ **Pulmonary atresia; n≈ 24***
- ♥ **Tetralogy of Fallot; n≈57**
- ♥ **D-Transposition of the great arteries; n≈41**
- ♥ **Tricuspid atresia; n≈17***
- ♥ **Total anomalous pulmonary venous return; n≈14**

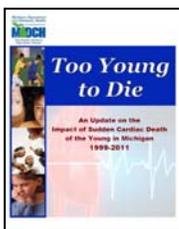
CCHD which are less likely to be detected by screening were also reported as follows:

Additional CCHD Detected

- ♥ **Coarctation of the aorta; n≈71**
- ♥ **Double-outlet right ventricle, n≈26**
- ♥ **Ebstein anomaly; n≈17**
- ♥ **Interrupted aortic arch; n≈12**
- ♥ **Single ventricle; n≈16**

Estimates are per MBDR reporting 2000-2010. Variation is based on changes in the birth rate, timely diagnosis and reporting. A child may have more than one CCHD. Revisions in diagnosis may occur due to the complexity of CCHDs. *Due to condition coding, counts include occurrences of stenosis as well as atresia.

Sudden Cardiac Death of the Young (SCDY) Surveillance and Prevention Project



MDCH first identified sudden cardiac death of the young (SCDY) as a focus for coordinated prevention efforts in 2004. A number of surveillance projects to identify SCDY trends were conducted from 2005-2011. Analysis of mortality data for the years 1999-2009 shows that CHD is the **most common cause of SCDY** in Michigan residents aged 1-19 years (Figure 3).

In addition:

- As many as 329 out-of-hospital SCDY occur each year.
- The statewide age-adjusted mortality rate is 5.5 per 100,000.
- Significant disparities exist, with more than two-thirds of the deaths occurring in males, and one-third in blacks.
- Dilated cardiomyopathy is the most commonly reported underlying cause of SCDY in children, adolescents and young adults, 1-29 years of age.
- Atherosclerotic cardiovascular disease is the most commonly reported underlying cause of SCDY at 30-39 years of age.

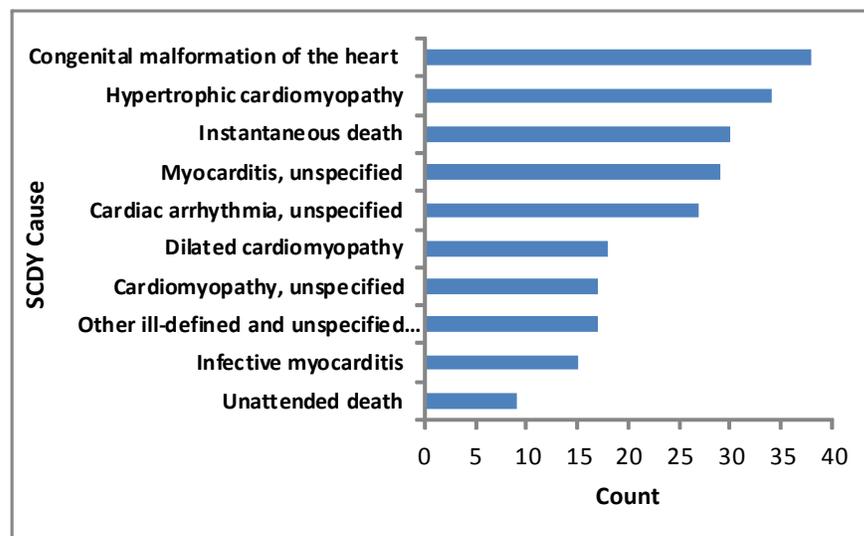
Since 2008, MDCH and multiple partners have been working on identified action steps to help prevent SCDY. In 2010, the Michigan High School Athletic Association (MHSAA) approved our recommended physical examination and medical history form that includes SCDY risk factors and symptoms. For more information about the SCDY Surveillance and Prevention Project, please visit www.michigan.gov/scdy.

What is the Good News?

Mortality from CHD is Declining!

Due to improved diagnostic capabilities, surgical techniques, and treatment, those born with CHD are living longer. A recent Centers for Disease Control and Prevention study entitled "Mortality Resulting From Congenital Heart Disease Among Children and Adults in the United States, 1999-2006" was published in the American Heart Association's *Circulation* journal.⁵ Mortality from CHD in the United States was found to be declining among all ages. Unfortunately, disparities persist. Among the deaths due to CHD in this study period, nearly half were in infants. Males were more likely to die from CHD than females, and non-Hispanic blacks more likely than non-Hispanic whites. This study highlights that individuals with CHD need access to appropriate medical care throughout their lifetimes.

Figure 3. Top causes of SCDY among Michigan Residents, Ages 1-19 years, 1999-2009.



Includes those who died out of the hospital, or in an emergency department, or were dead on arrival to an emergency department, and had either a cardiac-related or ill-defined condition reported as the underlying cause of death on the death certificate.

Remember to:

- ◇ Determine results of CCHD screening in newborns
- ◇ Use the MHSAA Physical Exam/Medical History form for children and youth
- ◇ Identify family history of heart symptoms
- ◇ Refer when indicated

Following trends, promoting prevention and linking families to resources

For more information or to receive future issues, please contact:

Birth Defects Program,
Lifecourse Epidemiology and Genomics
Division

201 Townsend St, CV-4

Lansing, MI 48913

Phone: Toll Free 1-866-852-1247

E-mail: BDRFollowup@michigan.gov

Website: www.michigan.gov/birthdefectsinfo

Acknowledgments

We would like to thank the Birth Defects Steering Committee for input and support.

MBDR Quality Assurance Efforts

Concurrent internal monitoring assures that incoming reports are screened for missing and invalid information as they are processed into the registry.

- MBDR staff compare demographic information on birth defects reports with that in birth and death records. The MBDR is linked with other public health program datasets, such as Newborn Screening, which helps to assure that the MBDR is as complete and accurate as possible.
- Reporting facilities are monitored for method, accuracy, and completeness of case reporting. Unreported cases are identified and submitted to the MBDR. Subsequently, education and technical support are provided to ensure reporting facilities are in compliance with legislative mandates.
- Retrospective facility audits are conducted every three to four years to assess statewide performance in the reporting of birth defects and to identify opportunities for improvement.

Besides quality improvement for birth defects reporting, an audit with on-site chart review allows for further investigation into issues affecting quality of life for children with birth defects, such as patterns of referral to needed services and access to coordinated, comprehensive medical care.

State and National Resources

- ♥ MDCH CCHD Newborn Screening Demonstration Program - www.michigan.gov/cchd
- ♥ MDCH Cardiovascular Health, Nutrition, and Physical Activity - <http://www.michigan.gov/mdch/>
- ♥ Children's Special Health Care Services (CSHCS) - www.michigan.gov/cshcs
- ♥ *Early On*® Michigan - <https://1800earlyon.org/>
- ♥ Michigan Child Death Review - <http://keepingkidsalive.org/>
- ♥ American Heart Association - <http://www.heart.org>
- ♥ CDC Congenital Heart Defects - <http://www.cdc.gov/ncbddd/heartdefects/index.html>
- ♥ Congenital Heart Information Network - <http://tchin.org/>
- ♥ Mended Little Hearts - <http://www.mendedlittlehearts.org/>
- ♥ Hearts of Hope Michigan - <http://www.heartsofhopemi.org/>

References

1. Centers for Disease Control and Prevention (CDC). 2013. Screening for Critical Congenital Heart Defects. *Pediatric Genetics*. Available from <http://www.cdc.gov/ncbddd/pediatricgenetics/cchdscreening.html#ref>.
2. Michigan Birth Defects Registry, Division for Vital Records & Health Statistics, Michigan Department of Community Health. Reports processed through March 15, 2012.
3. American Heart Association Statistical Fact Sheet, 2013 Update. Congenital Cardiovascular Defects.
4. Mahle WT, et al. Role of Pulse Oximetry in Examining Newborns for Congenital Heart Disease: A Scientific Statement from the American Heart Association and American Academy of Pediatrics. *Circulation* 2009;120:447-458;doi:10.1161/CIRCULATIONAHA.109.192576.
5. Gilboa SM, Salemi JL, Nembhard WN, Fixler DE, Correa A. Mortality resulting from congenital heart disease among children and adults in the United States, 1999 to 2006. *Circulation*. 2010;122:2254-63.

Suggested Citation

Mobley M, Simmons L, Quarshie E, Ehrhardt J, Bach J. Michigan Department of Community Health. *Michigan Monitor*. Volume 6, Issue 2. Fall 2013.