

NEURAL TUBE DEFECTS

Background

Neural tube defects (NTDs) are congenital anomalies of the brain, spine, or spinal column.¹ In the United States, around 3,000 pregnancies are affected by NTDs each year.²

NTDs occur within the first month of pregnancy, often before a woman is aware that she is pregnant. NTDs are present at birth and cause varying degrees of disability.^{1,2,3}

Forming in the first weeks after conception, the neural tube becomes the spinal cord, brain, and other adjacent structures such as the backbone. NTDs happen when the neural tube does not close properly, resulting

in a hole in the spinal column or another type of defect.¹

Three types of neural tube defects are: spina bifida, anencephaly, and encephalocele. The most common is spina bifida, and the least is encephalocele.^{1,4,5,6}

A major way to prevent NTDs is for women to take folic acid *before and very early* in pregnancy. Folic acid can be ingested through food or dietary supplements containing folic acid. Folic acid fortification became mandatory in the U.S. in 1998. Folic acid fortification has been very effective and has contributed to a dramatic decrease in NTDs.^{2,3}

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NTDs are reported to the Michigan Birth Defects Registry (MBDR) by hospitals and other reporting entities. This issue presents NTD trends for babies born in Michigan during 2007-2016.

Points of Interest

- ✦ NTDs affect around 3,000 pregnancies in the United States each year.
- ✦ Individuals with NTDs may have a variety of needs that can include health, education and lifetime support.
- ✦ Folic acid fortification has decreased NTDs dramatically.

Etiology

The cause of NTDs in most babies is unknown. Possible causes include⁷:

- ✦ Genes/genetics
- ✦ Exposure to certain foods, medication and environmental factors
- ✦ Lack of folic acid before and during pregnancy
- ✦ Exposure to alcohol, certain drugs or toxic chemicals

Neural tube defects per MBDR Reporting, 2007-2016

Occurrence

Between 2006 and 2017, the prevalence of NTDs in Michigan stayed fairly consistent with an overall prevalence of 6.6 per 10,000 live births. During the same time frame, the overall prevalence of spina bifida was 4.7 per 10,000 live births, the overall prevalence of anencephaly was 1.1 per 10,000 live births, and the overall prevalence of encephalocele was 1.1 per 10,000 live births. The prevalence rates for each of these conditions remained fairly steady over time (Figure 1).

Demographics

Between 2007 and 2016, the overall prevalence of NTDs in Michigan differed by several demographic variables (Table 1). The overall prevalence of NTDs was the highest among infants born to mothers 35 years of age or older (7.0 per 10,000 live births), infants born to mothers of white race (7.1 per 10,000 live births), infants born very preterm (34.0 per 10,000 live births), and infants born to mothers of Hispanic ethnicity (8.1 per 10,000 live births). The overall prevalence of NTDs did not differ by infant sex.

Between 2007 and 2016, the prevalence of anencephaly was the highest among infants born to mothers less than 20 years of age (1.5 per 10,000 live births), infants born very preterm (15.8 per 10,000 live births), and infants born to mothers of Hispanic ethnicity (1.5 per 10,000 live births) (Table 1). The prevalence of spina bifida was the highest among infants born to mothers 35 years of age or older (5.2 per 10,000 live births), infants born to mothers of white race (5.1 per 10,000 live births), infants born very preterm (12.5 per 10,000 live births), and infants born to mothers of Hispanic ethnicity (5.4 per 10,000 live births). Finally, the prevalence of encephalocele was the highest among infants born to mothers between 20 and 24 years of age (1.2 per 10,000 live births), infants born to mothers of black race (1.5 per 10,000 live births), infants born very preterm (6.7 per 10,000 live births). The prevalence of these individual NTDs did not differ by infant sex.

Figure 1: Three year moving prevalence rate of neural tube defects: MBDR, 2007-2016

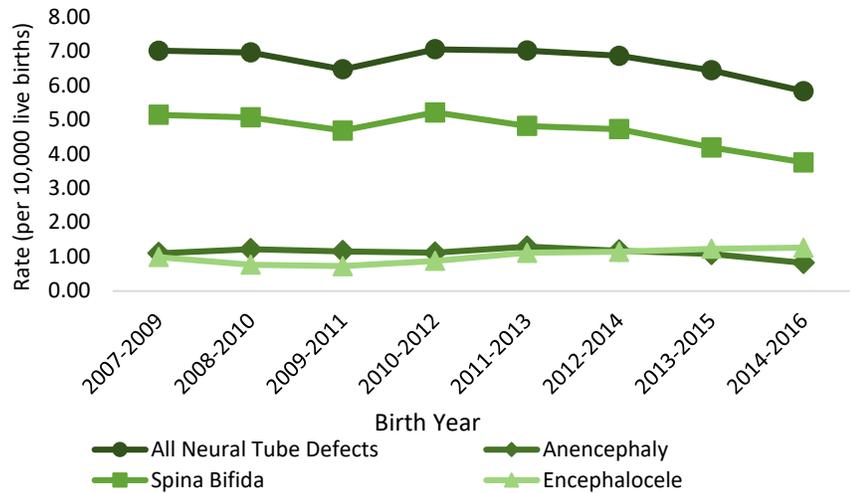


Table 1: Prevalence rate of neural tube defects stratified by selected demographic variables: MBDR, 2007-2016

Demographic	Prevalence Rate ^{1,2}			
	Total NTDs	Anencephaly	Spina Bifida	Encephalocele
Maternal Age				
<20	6.6	1.5	4.3	1.0
20-24	6.4	1.0	4.3	1.2
25-29	5.6	0.9	4.0	0.7
30-34	6.0	0.8	4.3	1.1
35+	7.0	1.0	5.2	1.0
Maternal Race				
White	7.1	1.1	5.1	1.0
Black	5.3	0.8	3.0	1.5
Other ³	5.9	0.8	4.0	1.0
Gestational Age at Birth				
Very Preterm (<32 Weeks)	34.0	15.8	12.5	6.7
Preterm (32-37 Weeks)	15.5	3.0	10.3	2.8
Not Preterm (>37 Weeks)	5.3	0.6	4.0	4.0
Maternal Ethnicity				
Hispanic	8.1	1.5	5.4	1.4
Arabic	3.9	0.7	2.1	1.2
Sex of Infant				
Male	6.7	1.0	4.6	1.2
Female	6.6	1.1	4.7	0.9

Footnotes to Table 1.

¹Prevalence rates are based on births to mothers living in Michigan at the time of delivery. Data are current through January 2016.

²Prevalence rate expressed as cases per 10,000 live births

³Includes women who do not define themselves as Black or White and includes Native American and Asian/Pacific Islander.

Types of Neural Tube Defects

Anencephaly

Anencephaly occurs when the upper part of the neural tube does not close all the way, resulting in a baby being born without the forebrain and cerebrum. The remaining parts of the brain are often not covered by bone or skin.⁵ Anencephaly is uniformly fatal. Some affected babies are stillborn. Most affected live born babies will live for a few hours or days.

Spina Bifida

Spina Bifida occurs when the neural tube does not close all the way causing damage to the spinal cord and nerves.⁴ People with spina bifida often have trouble with bowel and bladder control and walking. There is more likely to be hydrocephaly and cognitive disability and other challenges.

Encephalocele

Encephalocele occurs when the neural tube does not close completely resulting in an opening along the center of the skull from the nose to the back of the neck, but more often at the back of the head, the top of the head, or between the forehead and the nose.⁶ This results in a sac-like protrusion of brain and membranes through that opening.

Infant Mortality and Fatality

Between 2007 and 2016, the infant mortality rate (IMR) was 1.2 deaths of babies with a NTD per 10,000 live births. Furthermore, the IMR of infants with anencephaly was 0.9 per 10,000 live births, and the IMR of both spina bifida and encephalocele was 0.2 per 10,000 live births.

The case fatality rate (CFR) among infants with an NTD was 8.5 deaths per 10,000 infants born with a birth defect. Furthermore, the CFR of infants with anencephaly was 6.2 per 10,000 infants born with a birth defect, the CFR of spina bifida was 1.4 per 10,000 infants born with a birth defect, and the CFR of encephalocele was 1.2 per 10,000 infants born with a birth defect.

Stillbirth

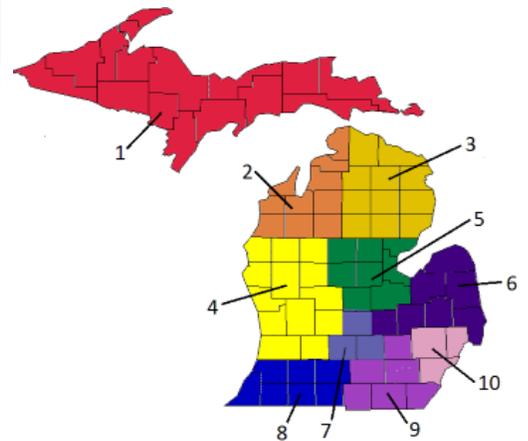
A prior analysis of [MDHHS fetal death records](#) found anencephaly or spina bifida in about 8-9 percent of recorded stillbirths that include a birth defect.

Maternal Residence

Table 3 displays the prevalence of NTDs according to the mother's resident prosperity region at the time of birth. Michigan is broken into 10 prosperity regions. The regions are defined based on shared geographic, demographic, and economic interests, and often have related public health and medical infrastructure. NTDs were found in all ten prosperity regions. Prosperity region 7 was found to have the highest NTD prevalence rate at 10.8 cases per 10,000 live births. Prosperity region 1 was found to have the lowest prevalence rate at 3.5 cases per 10,000 live births.

Table 3: Prevalence of NTDs by prosperity region of maternal residence: MBDR, 2007-2016

Region	Number of Cases	Prevalence Rate ^{1,2}
1	10	3.5
2	17	5.6
3	11	6.2
4	156	7.8
5	34	5.5
6	71	7.3
7	57	10.8
8	64	6.7
9	59	6.0
10	291	6.2
Total	770	6.7



Footnotes to Table 2.

¹Prevalence rates are based on births to mothers living in Michigan at the time of delivery. Data are current through January 2016.

²Prevalence rate expressed as cases per 10,000 live births.

*Regions approximate prosperity region boundaries.

Good News

New treatments and research keep improving the outlook for people with spina bifida.

Fetal Surgery

Fetal surgery may now be an option when spina bifida is diagnosed prenatally, by about 26 weeks gestation. A multicenter study called the [Management of Myelomeningocele Study \(MOMS\)](#) found that many babies

with fetal surgery were less likely to have hydrocephalus and had better development overall than babies who had spina bifida repair after birth.

Better Care

People with spina bifida are likely to have multiple surgeries, among other ongoing special health care needs. The Centers for Disease Control and Prevention (CDC) manages the [National Spina Bifida Patient Registry](#) to better

understand the health challenges of people with spina bifida and to identify and share best practices.

The [Spina Bifida Association \(SBA\)](#) provides information, support and resources for all affected by spina bifida. The SBA's *Guidelines for the Care of People with Spina Bifida* were created by an expert panel and are available [online](#).

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The Michigan Monitor is online at:

www.michigan.gov/mchepi

MBDR reporting:

<http://www.michigan.gov/mbdr>

*Give us your
feedback!*

Folic Acid for NTD Prevention

Every woman can improve her chances of having a healthy baby by taking folic acid *before* and *very early* pregnancy. Folic acid, vitamin B9, is found in many foods (as folate) and in many dietary supplements. The US Preventive Services Task Force (USPSTF) recommends a *daily* supplement with 400 to 800 micrograms (mcg) of folic acid for all women of childbearing age.⁸

Folic acid fortification of enriched grain products became mandatory in the US in 1998. Makers of ready-to-eat cereals may fortify products with up to 400 mcg of folic acid per serving. The mandate for fortification did not include corn masa flour, however, leaving many Hispanic women and their babies with less protection. The US Food and Drug Administration (FDA) approved voluntary fortification of corn masa flour in 2016.⁹

Folic acid fortification has led to an estimated 35 percent decrease in

Resources for Health Professionals and Families

Michigan

- Children's Special Health Care Services (CSHCS) Program: www.michigan.gov/cshcs
- Early On®, Michigan's early intervention system: www.1800earlyon.org
- Family to Family Information Center: <http://f2fmichigan.org>

National

- Centers for Disease Control and Prevention, National Center on Birth Defects and Disabilities: www.CDC.gov/NCBDDD/birthdefects
- March of Dimes: <http://www.marchofdimes.org/>
- National Birth Defects Prevention Network (NBDPN): www.nbdpn.org
- Spina Bifida Association: <http://spinabifidaassociation.org>

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6. Centers for Disease Control and Prevention. Facts about Encephalocele. 2017.
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8. U.S. Preventive Services Task Force. Final Recommendation Statement Folic Acid for the Prevention of Neural Tube Defects: Preventative Medication. 2017.
9. United States Food and Drug Administration. Food Additives Permitted for Direct Addition to Food for Human Consumption; Folic Acid. 2016.
10. Crider KS, Qi YP, Devine O, Tinker S, Berry RJ. Modeling the impact of folic acid fortification and supplementation on red blood cell folate concentrations and predicted neural tube defect risk in the United States: have we reached optimal prevention? . *The American Journal of Clinical Nutrition*. 2018;107(6):1027-1034.

NTDs. Nonetheless, analysis of National Health and Nutrition Examination Study (NHANES) data supports taking a daily multivitamin with folic acid to further lower NTD risk.¹⁰

Women who have had an NTD affected pregnancy have a higher chance to have another. To lower the risk, they should take **4,000 mcg of folic acid** every day – available by prescription – when planning a subsequent pregnancy.

Help spread the word about the importance of folic acid and other birth defects prevention strategies by sharing these tips from the National Birth Defects Prevention Network: [*Best for You. Best for Baby.*](#)

MBDR Coding Updates

Updates for ICD-10-CM reportable conditions for 2018-2019 include a total of 25 new codes. These include 16 new prenatal exposure condition codes which are now on the list. If you have questions regarding reporting, contact Lorrie Simmons at 517-335-9197 or

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