



# **NORTH DAKOTA BIRTH DEFECTS MONITORING SYSTEM SUMMARY REPORT 2001–2005**

March 2010



**NORTH DAKOTA**  
DEPARTMENT *of* HEALTH



# **North Dakota Birth Defects Monitoring System Summary Report 2001–2005**

**John Hoeven**  
Governor

**Terry Dwelle, M.D., M.P.H.T.M.**  
State Health Officer

**Devaiah Muccatira**  
Research Analyst III  
State System Development Initiative Coordinator

**North Dakota Department of Health**  
Division of Children's Special Health Services  
600 E. Boulevard Ave., Dept. 301  
Bismarck, ND 58505-0200  
701.328.4963 or 1.800.755.2714  
[www.ndhealth.gov](http://www.ndhealth.gov)

**Report Prepared By**  
The North Dakota State Data Center

*Contributors*  
Jean Beaupre  
Kendra Erickson  
Ramona Danielson

**Publication Date**  
December 2009

## **Acknowledgments**

We wish to thank the Alaska Division of Public Health for allowing us to use aspects of formatting and narrative from its report, Alaska Maternal and Child Health Data Book 2005: Birth Defects Surveillance Edition with permission from the author, Janine Schoellhorn, senior epidemiologist, MCH Epidemiology Unit, and Alaska Division of Public Health.



# *Executive Summary*

---

## **Introduction**

The state of North Dakota monitors 43 major congenital anomalies, or birth defects. This report presents an analysis of birth defects in North Dakota from 2001 through 2005. The report presents the rates for the 24 birth defects that had incidences of 10 or more. In addition, 95 percent confidence intervals also are presented. Within the summary of results and the report, the 24 birth defects are divided into sections based on the anatomical site of the malformation. For purposes of comparison, North Dakota's rates are compared to those of Alaska, Colorado and Utah<sup>1</sup>.

In addition to analysis of birth defect information, information regarding population characteristics for North Dakota also is provided, including population distribution and population characteristics, as well as characteristics of live births and birth, pregnancy, and death data. Detailed incidence and prevalence for all 43 birth defects are provided in the Appendix Tables section of the Appendices. Additional state and national data, a glossary, a list of acronyms, birth defect resources, and references also are provided as appendices.

## **What Are Birth Defects?**

Birth defects are defined as abnormalities of structure, function or body metabolism that are present at birth.

The major categories of birth defects tracked in North Dakota are organized according to the anatomical site of the anomaly. The categories in which there were anomalies with 10 or more incidents in 2001 through 2005 in North Dakota include central nervous system defects (also referred to as neural tube defects), cardiovascular defects, orofacial defects, gastrointestinal defects, genitourinary defects, musculoskeletal defects, chromosomal defects and other defects. Additional categories tracked include eye defects and ear defects.

## **North Dakota Birth Defects Monitoring System**

The North Dakota Birth Defects Monitoring System (NDBDMS) was established in 2003 as a means of identifying and collecting information about all babies born with certain birth defects in North Dakota.

The surveillance method of the NDBDMS is passive case ascertainment where the surveillance program receives reports of birth defects from data sources such as birth certificates, death certificates and fetal death certificates, Medicaid claims payment and Children's Special Health Services program data. These data were provided to the North Dakota State Data Center by the North Dakota Department of Health for purposes of presenting the information in this report.

---

<sup>1</sup> Due to variability in the methods used by state birth defects surveillance systems and differences in populations and risk factors, state prevalence estimates may not be directly comparable with national estimates or those of other states.

## Highlights

### Population Characteristics

- North Dakota had 641,481 people as of July 2008, and was ranked 48<sup>th</sup> in population size in the United States.<sup>(27)</sup>
- North Dakota is predominantly white. American Indian and Alaska Natives are the state's largest minority.<sup>(27)</sup>
- Approximately 7 percent of North Dakota resident live births from 2001 to 2005 were children with low birth weight (less than 2,500 grams).
- Of mothers who gave live birth from 2001 to 2005, 8 percent were in their teens. Thirty percent of mothers were unmarried. Thirteen percent of mothers started prenatal care in their second trimester or later or did not receive prenatal care. One percent of mothers reported using alcohol during their pregnancy, and 17 percent reported using tobacco during their pregnancy.
- The number of births in North Dakota has been increasing, reaching 8,381 in 2005 (up from 7,664 in 2001). North Dakota's birth rate reached 13.05 per 1,000 residents in 2005 (up from 11.93 per 1,000 residents in 2001).
- The numbers of infant and neonatal deaths in North Dakota have fluctuated, with an annual average of 53 infant deaths and 39 neonatal deaths from 2001 through 2005.

### Major Congenital Anomalies in North Dakota

- There were 39,955 live births in North Dakota from 2001 through 2005. Of these live births, 1,240 (3 percent) had major congenital anomalies, although some live births may have one or more congenital anomaly.
- Cardiovascular birth defects were the most common type of anomaly in North Dakota from 2001 through 2005; 58 percent of congenital anomalies were cardiovascular defects. An additional 10 percent were gastrointestinal defects, and 8 percent were central nervous system defects.

### Summary of Birth Defects With 10 or More Incidences in North Dakota

#### Central Nervous System Defects (Also Called Neural Tube Defects)

- North Dakota had four central nervous system birth defects with an incidence of 10 or more from 2001 through 2005. The defects, along with North Dakota's rate per 10,000 live births, are as follows: anencephalus (2.50), hydrocephalus without spina bifida (7.76), microcephalus (8.51), and spina bifida without anencephalus (5.76).
  - The North Dakota rate for microcephalus was lower than Alaska's rate, but slightly higher than Utah's rate. North Dakota's rate of microcephalus was statistically similar to Colorado's rate.

- North Dakota's rate for anencephalus was statistically similar to the rates of Colorado and Utah. No data were provided for Alaska.
- North Dakota's rates for hydrocephalus without spina bifida and spina bifida without anencephalus were statistically similar to the rates of Alaska, Colorado, and Utah.

### **Cardiovascular Defects**

- North Dakota had 10 cardiovascular birth defects with an incidence of 10 or more from 2001 through 2005. The defects, along with North Dakota's rate per 10,000 live births, are as follows: aortic valve stenosis (2.75), atrial septal defect (62.57), coarctation of aorta (5.51), endocardial cushion defect (3.00), hypoplastic left heart syndrome (3.50), patent ductus arteriosus (32.29), pulmonary valve atresia and stenosis (13.01), tetralogy of Fallot (4.51), transposition of great arteries (4.76), and ventricular septal defect (44.55).
  - North Dakota's rate of atrial septal defect was much lower than Alaska's rate and slightly lower than Colorado's rate. North Dakota's rate was much higher than Utah's rate.
  - North Dakota's rate for patent ductus arteriosus was much lower than Alaska's rate and statistically similar to Colorado's rate. No data were provided for Utah.
  - North Dakota's rate for ventricular septal defect was much lower than Alaska's rate and much higher than Utah's rate. North Dakota's ventricular septal defect rate was statistically similar to Colorado's rate.
  - North Dakota's rates for aortic valve stenosis, coarctation of aorta, endocardial cushion defect, hypoplastic left heart syndrome, pulmonary valve atresia and stenosis, tetralogy of Fallot, and transposition of great arteries were statistically similar to the rates of Alaska, Colorado and Utah.

### **Orofacial Defects**

- North Dakota had two orofacial birth defects with an incidence of 10 or more from 2001 through 2005. The defects, along with North Dakota's rate per 10,000 live births, are as follows: cleft lip with and without cleft palate (10.01) and cleft palate without cleft lip (10.76).
  - North Dakota's rates for both cleft lip with and without cleft palate and cleft palate without cleft lip were statistically similar to the rates of Alaska, Colorado and Utah.

### **Gastrointestinal Defects**

- North Dakota had two gastrointestinal birth defects with an incidence of 10 or more from 2001 through 2005. The defects, along with North Dakota's rate per 10,000 live births, are as follows: pyloric stenosis (21.52) and rectal and large intestinal atresia/stenosis (5.76).
  - North Dakota's rate for pyloric stenosis was slightly higher than Utah's rate and statistically similar to Alaska's and Colorado's rates.
  - North Dakota's rate for rectal and large intestinal atresia/stenosis was statistically similar to the rates of Alaska, Colorado and Utah.

## **Genitourinary Defects**

- North Dakota had two genitourinary birth defects with an incidence of 10 or more from 2001 through 2005. The defects, along with North Dakota's rate per 10,000 live births, are as follows: hypospadias and epispadias (13.26) and renal agenesis/hypoplasia (4.76).
  - North Dakota's rate for hypospadias and epispadias was much lower than Alaska's and Colorado's rates and lower than Utah's rate.
  - North Dakota's rate for renal agenesis/hypoplasia was statistically similar to the rates of Alaska, Colorado and Utah.

## **Musculoskeletal Defects**

- North Dakota had two musculoskeletal birth defects with an incidence of 10 or more from 2001 through 2005. The defects, along with North Dakota's rate per 10,000 live births, are as follows: congenital hip dislocation (7.01) and diaphragmatic hernia (3.00).
  - North Dakota's rate for congenital hip dislocation was lower than Alaska's and Colorado's rates. No data were provided for Utah.
  - North Dakota's rate for diaphragmatic hernia was statistically similar to the rates of Alaska, Colorado and Utah.

## **Chromosomal Defects**

- North Dakota had one chromosomal birth defect with an incidence of 10 or more from 2001 through 2005. The defect, along with North Dakota's rate per 10,000 live births, is as follows: Down syndrome (Trisomy 21) (12.26).
  - North Dakota's rate for Down syndrome (Trisomy 21) was lower than Colorado's rate and statistically similar to Alaska's and Utah's rates.

## **Other Defects**

- North Dakota had one birth defect that fell under the category of other defects with an incidence of 10 or more from 2001 through 2005. The defect, along with North Dakota's rate per 10,000 live births, is as follows: fetal alcohol spectrum disorder (FASD) (5.26).
  - North Dakota's rate for FASD could not be compared to the rates of Alaska, Colorado and Utah because no data were provided for these states.

# Table of Contents

---

<b>Executive Summary</b> .....	<b>4-7</b>
<b>Introduction</b> .....	<b>11</b>
<b>Population Characteristics</b> .....	<b>12-19</b>
North Dakota Population Distribution .....	13
North Dakota Population Characteristics .....	14
<i>Table 1. North Dakota Population Distribution by Age, 2008</i> .....	14
<i>Table 2. North Dakota Population Distribution by Sex, 2008</i> .....	14
<i>Table 3. North Dakota Population Distribution by Race, 2008</i> .....	14
Characteristics of Live Births in North Dakota .....	15-16
<i>Table 4. Characteristics of North Dakota Resident Live Births from 2001 through 2005</i> .....	15
<i>Table 5. Education Level of North Dakota Mothers Who Gave Birth by Age Group from 2001 through 2005</i> .....	15
<i>Table 6. Characteristics of North Dakota Mothers Who Gave Birth from 2001 through 2005</i> .....	16
North Dakota Birth, Pregnancy, Fertility, and Death Data .....	17-19
<i>Table 7. North Dakota Resident Births, 2001 through 2005</i> .....	17
<i>Table 8. North Dakota Resident Pregnancies, 2001 through 2005</i> .....	17
<i>Table 9. North Dakota Resident Deaths, 2001 through 2005</i> .....	17
<i>Table 10. North Dakota Resident Birth Ratios, 2001 through 2005</i> .....	18
<i>Table 11. North Dakota Resident Pregnancy Ratios, 2001 through 2005</i> .....	18
<i>Table 12. North Dakota Resident Death Ratios, 2001 through 2005</i> .....	18
<i>Table 13. North Dakota Resident Birth Rates, 2001 through 2005</i> .....	19
<i>Table 14. North Dakota Resident Pregnancy Rates, 2001 through 2005</i> .....	19
<i>Table 15. North Dakota Population Resident Fertility Rates, 2001 through 2005</i> .....	19
<b>North Dakota Birth Defects Monitoring System (NDBDMS)</b> .....	<b>20-25</b>
NDBDMS .....	20-22
<i>Table 16. Birth Defects Monitoring System: ICD-9 Codes</i> .....	21-22
Data Collection Methodology .....	23-25
<b>Description of Birth Defects and Results</b> .....	<b>26-60</b>
Overview of Major Congenital Anomalies .....	27
Central Nervous System Defects .....	28-32
Anencephalus .....	29
Hydrocephalus without spina bifida .....	30
Microcephalus .....	31
Spina bifida without anencephalus .....	32
Cardiovascular Defects .....	33-43
Aortic valve stenosis .....	34
Atrial septal defect .....	35
Coarctation of aorta .....	36
Endocardial cushion defect .....	37
Hypoplastic left heart syndrome .....	38

Patent ductus arteriosus .....	39
Pulmonary valve atresia and stenosis .....	40
Tetralogy of Fallot.....	41
Transposition of great arteries.....	42
Ventricular septal defect.....	43
Orofacial Defects .....	44-46
Cleft lip with and without cleft palate .....	45
Cleft palate without cleft lip.....	46
Gastrointestinal Defects .....	47-49
Pyloric stenosis.....	48
Rectal and large intestinal atresia/stenosis .....	49
Genitourinary Defects.....	50-52
Hypospadias and epispadias.....	51
Renal agenesis/hypoplasia.....	52
Musculoskeletal Defects .....	53-55
Congenital hip dislocation.....	54
Diaphragmatic hernia .....	55
Chromosomal Defects.....	56-57
Down syndrome (Trisomy 21) .....	57
Other Defects .....	58-59
Fetal alcohol spectrum disorders.....	59
Additional Birth Defects Monitored by NDBDMS .....	60
<i>Table 17. Prevalence of Birth Defects Monitored in North Dakota with     Less Than 10 Cases, 2001–2005.....</i>	<i>60</i>

<b>Appendices.....</b>	<b>61-111</b>
North Dakota Birth Defects State Profile .....	62
Appendix Tables .....	63-77
<i>Appendix Table 1. Incidence and Prevalence of Reported Cases of North         Dakota Birth Defects by Race/Ethnicity, 2001–2005 .....</i>	<i>63-65</i>
<i>Appendix Table 2. Incidence of Birth Defects in North Dakota, Alaska,         Colorado, and Utah with Confidence Intervals, 2001–2005...</i>	<i>65-66</i>
<i>Appendix Table 3. Prevalence of Birth Defects in North Dakota, Alaska,         Colorado, and Utah, 2001–2005 .....</i>	<i>66-71</i>
<i>Appendix Table 4. Reportable Birth Defects Monitoring System:         ICD-10 &amp; CDC/BPA Codes.....</i>	<i>72-73</i>
<i>Appendix Table 5a. Average Prevalence of Selected Major Birth Defects –         Active Surveillance Systems, U.S., 1999–2001 .....</i>	<i>74</i>
<i>Appendix Table 5b. Average Prevalence of Selected Major Birth Defects – Passive         Surveillance Systems with Follow-Up, U.S., 1999–2001.....</i>	<i>75</i>
<i>Appendix Table 5c. Average Prevalence of Selected Major Birth Defects – Passive         Surveillance Systems without Follow-Up, U.S., 1999–2001 .....</i>	<i>76</i>
<i>Appendix Table 5d. Prevalence of Selected Birth Defects in North Dakota Between         1995-1999 and 2001-2005.....</i>	<i>77</i>
Glossary of Congenital Anomalies .....	78-81
List of Acronyms .....	82
Prevention Methods .....	83-84
North Dakota Resources .....	85-86

Highlighted Birth Defect Resources .....	87-94
Birth Defect Resources on the Internet.....	95-104
North Dakota Century Code .....	105
Acknowledgements .....	106
References (Text).....	107-109
References (Images).....	110-111

## *Introduction*

---

Birth defects are defined as abnormalities of structure, function or body metabolism that are present at birth. Birth defects can lead to mental or physical disabilities, years of life lost, illness, and long-term disabilities that are sometimes fatal. There are more than 4,000 known birth defects in the United States. Birth defects are the leading cause of infant death in the first year of life. About 70 percent of all birth defects have unknown causes; however, environmental, genetics, dietary, and personal risk factors are all possible contributors to birth defects.

A birth defect is a problem that happens while the baby is developing in the mother's body. Most birth defects happen during the first three months of pregnancy. A birth defect may affect how the body looks, works or both. It can be found before birth, at birth or anytime after birth. Most defects are found within the first year of life. Some birth defects (such as cleft lip or clubfoot) are easy to see, but others (such as heart defects or hearing loss) are found using special tests (such as X-rays, CAT scans or hearing tests). Birth defects can vary from mild to severe. Babies with birth defects may need surgery or other medical treatments, but, if they receive the help they need, these babies often lead full lives.<sup>(4)</sup>

The North Dakota Birth Defects Monitoring System (NDBDMS) was established in 2003 as a means of identifying and collecting information about all babies born with certain birth defects in North Dakota. The monitoring program analyzes data about babies born with health problems to help researchers and health-care providers learn more about preventing these problems in the future. The primary purpose of the NDBDMS is to provide a means for accurate and up-to-date tracking in order to determine if affected children have access to needed health care and other services necessary to treat their condition.

NDBDMS tracks 43 birth defects in the state using a method of passive case ascertainment (e.g., data are obtained from birth, death and fetal death certificates, Medicaid claims data and Children's Special Health Services program data). Birth defects are categorized as central nervous system defects (also referred to as neural tube defects), cardiovascular defects, eye defects, ear defects, orofacial defects, gastrointestinal defects, genitourinary defects, musculoskeletal defects, chromosomal defects and other defects. Of the 43 birth defects, 24 had incidences of 10 or more in the time period of 2001 through 2005.

For these 24 birth defects, we present a picture of the anomaly, a definition of the anomaly, and comparative data between North Dakota, Alaska, Colorado and Utah (when available). North Dakota, Colorado and Utah are all states within the same region of the National Birth Defects Prevention Network. Alaska also is included for comparison because of its sizeable proportion of American Indian/Alaska Natives, which is North Dakota's largest minority group.

The four states also are comparable in their methods of surveillance. Similar to North Dakota, Alaska uses a passive case ascertainment surveillance method. Colorado and Utah use a combination of active and passive case ascertainment.

Data for all 43 birth defects can be found in the Appendix Tables.

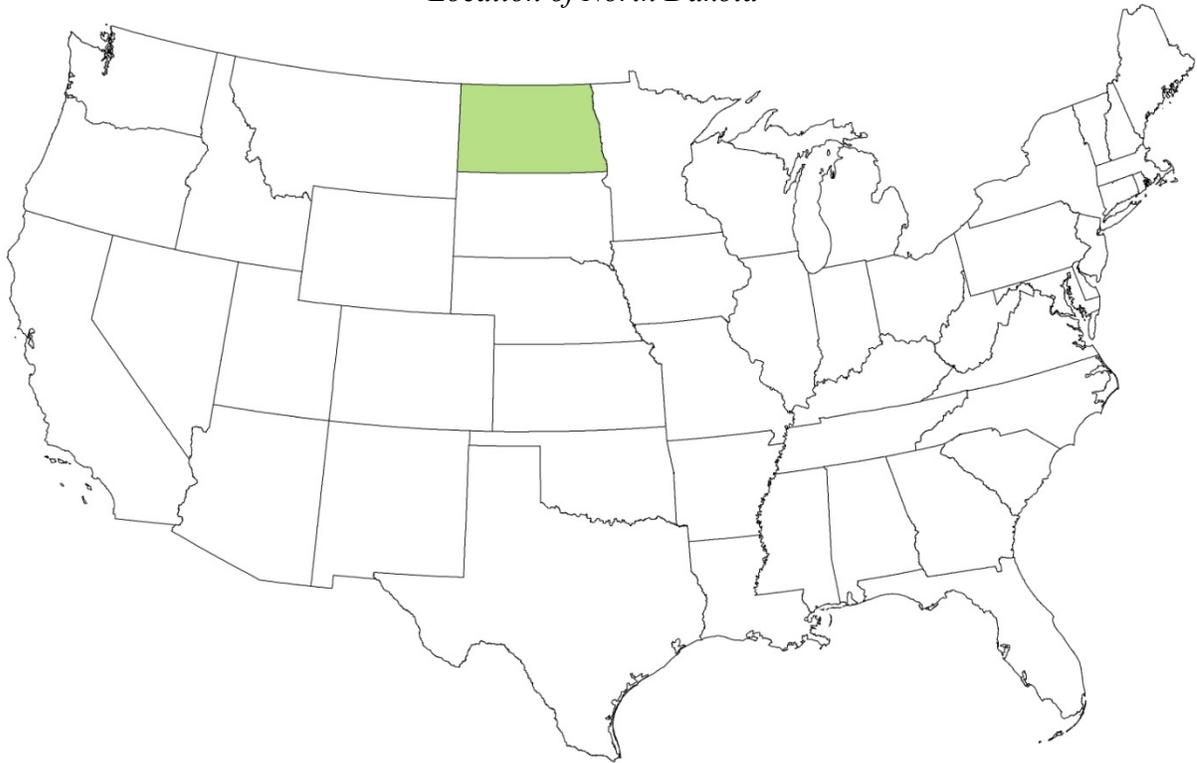
# Population Characteristics

## *North Dakota Population Distribution*

---

- Comprising approximately 2 percent of the overall land mass of the United States, North Dakota occupies 68,975.93 square miles.<sup>(26)</sup>
- Representing 0.2 percent of the population in the United States and ranking 48<sup>th</sup> among the 50 states, North Dakota's population estimate as of July 2008 was 641,481.<sup>(27)</sup>
- In North Dakota, whites accounted for 91 percent of the state's population in 2008, American Indian and Alaska Natives constituted 6 percent, blacks made up 1 percent, and Asian or Pacific Islanders made up 1 percent. Two percent of North Dakotans indicated that they were of Hispanic ethnicity.<sup>(27)</sup>
- North Dakota's median age was similar to the United States in 2008 (37.1 years and 36.8 years, respectively).<sup>(27)</sup>

*Location of North Dakota*



## North Dakota Population Characteristics

The following population characteristics describe the age, gender and racial features of North Dakota's overall population.

**Table 1. North Dakota Population Distribution by Age, 2008<sup>(27)</sup>**

Age	North Dakota Residents	
	Number	Percent
Younger than 5 years	41,896	6.5
5–9 years	36,706	5.7
10–14 years	38,826	6.1
15–19 years	48,165	7.5
20–24 years	60,084	9.4
25–34 years	80,504	12.5
35–44 years	74,409	11.6
45–54 years	94,271	14.7
55–59 years	41,099	6.4
60–64 years	31,245	4.9
65–74 years	43,583	6.8
75–84 years	32,921	5.1
85 years and older	17,772	2.8
Total population	641,481	100.0
Median age (in years)		37.1

**Table 2. North Dakota Population Distribution by Sex, 2008<sup>(27)</sup>**

Sex	North Dakota Residents	
	Number	Percent
Male	321,933	50.2
Female	319,548	49.8
Total population	641,481	100.0

**Table 3. North Dakota Population Distribution by Race, 2008<sup>(27)</sup>**

Race	North Dakota Residents	
	Number	Percent
White alone	586,272	91.4
American Indian and Alaskan Native alone	35,666	5.6
Black alone	6,956	1.1
Asian or Pacific Islander alone	5,095	0.8
Two or more races	7,492	1.2
Total population	641,481	100.1

Note: These data represent race alone or in combination with one or more other races. The numbers may add to more than the total population and the percentages may add to more than 100 percent because individuals may report more than one race.

## Characteristics of Live Births in North Dakota

Characteristics of live births are documented on an infant's birth certificate and registered as a vital record with the state of North Dakota. These characteristics include details of the infant's birth, as well as demographic, medical and behavioral factors affecting the pregnancy.

Table 4 presents information on the number of live births from 2001 through 2005 by child sex and birth weight. Tables 5 and 6 present information about maternal characteristics, including maternal education levels, race, age, use of prenatal care, reported alcohol and tobacco use during pregnancy, and marital status.

**Table 4. Characteristics of North Dakota Resident Live Births From 2001 Through 2005**

Characteristics	Live Births	
	Number	Percent
Total births	39,955	100.0
<b>Total Births by Year</b>		
2001	7,664	19.2
2002	7,755	19.4
2003	7,976	20.0
2004	8,179	20.5
2005	8,381	21.0
<b>Sex of Child</b>		
Female	19,502	48.8
Male	20,453	51.2
<b>Birth Weight</b>		
Low (less than 2,500 grams)	2,639	6.6
Normal	37,316	93.4

**Table 5. Education Level of North Dakota Mothers Who Gave Birth by Age Group From 2001 Through 2005**

Age of Mother	North Dakota Mothers' Education Level											
	High School or Lower		Some College		Bachelor's Degree		College Past Bachelor's Level		Unknown		Total	
	#	%*	#	%	#	%	#	%	#	%	#	%*
15–19 years	2,933	21.0	263	2.2	2	0.0	0	0.0	7	4.1	3,205	8.2
20–29 years	8,710	62.3	7,884	64.7	4,504	51.9	1,909	45.0	82	47.7	23,089	58.8
30–39 years	2,137	15.3	3,822	31.3	3,980	45.9	2,195	51.7	37	21.5	12,171	31.0
40–45 years	169	1.2	218	1.8	180	2.1	135	3.2	5	2.9	707	1.8
Missing/other	39	0.3	5	0.0	9	0.1	5	0.1	41	23.8	99	0.3
Total	13,988	100.1	12,192	100.0	8,675	100.0	4,244	100.0	172	100.0	39,271	100.1

\*Percentages may not equal 100.0 due to rounding.

**Table 6. Characteristics of North Dakota Mothers Who Gave Birth From 2001 Through 2005**

Characteristics	Mothers Who Gave Live Birth	
	Number	Percent*
Total mothers	39,271	100.0
<b>Maternal Race</b>		
White	33,262	84.7
American Indian and Alaskan Native	4,904	12.5
Black	515	1.3
Asian or Pacific Islander	498	1.3
Other	6	0.0
Missing	86	0.2
<b>Maternal Age</b>		
15–19 years	3,205	8.2
20–29 years	23,089	58.8
30–39 years	12,171	31.0
40–45 years	707	1.8
Missing and others	99	0.3
<b>Started Prenatal Care</b>		
First trimester	33,454	85.2
Second trimester	4,380	11.1
Later or none	890	2.3
Missing or unknown	547	1.4
<b>Self-Reported Maternal Alcohol Use During Pregnancy</b>		
Used	383	1.0
Did not use	38,568	98.2
Missing	320	0.8
<b>Self-Reported Maternal Tobacco Use During Pregnancy</b>		
Used	6,545	16.7
Did not use	32,503	82.8
Missing	223	0.6
<b>Maternal Marital Status</b>		
Married	27,595	70.3
Unmarried	11,666	29.7
Unknown	10	0.0

\*Percentages may not equal 100.0 due to rounding.

## *North Dakota Birth, Pregnancy, Fertility and Death Data*

Data regarding births, pregnancies, fertility and deaths are found in Tables 7 through 15. Birth data include number of births in Table 7, ratios in Table 10, and rates in Table 13. Pregnancy data include numbers of pregnancies in Table 8, ratios in Table 11, and rates in Table 14. Death data include number of deaths in Table 9 and ratios in Table 12. Fertility rates are presented in Table 15.

**Table 7. North Dakota Resident Births, 2001 Through 2005**

Year	Number of Resident Births			
	Total Births	Teenage Births (Ages 15–19)	Births to Unmarried Mothers	Low Weight Births (Less Than 2,500 Grams)
2001	7,664	712	2,132	528
2002	7,755	657	2,250	498
2003	7,976	634	2,276	533
2004	8,179	631	2,447	542
2005	8,381	668	2,698	538

**Table 8. North Dakota Resident Pregnancies, 2001 Through 2005**

Year	Number of Resident Pregnancies		
	Total Pregnancies*	Teenage Pregnancies (Ages 15–19)	Pregnancies Among Unmarried Women
2001	8,453	889	2,762
2002	8,572	826	2,905
2003	8,835	794	2,957
2004	9,064	806	3,168
2005	9,347	829	2,957

\*Pregnancies are resident live births plus fetal deaths (death of a fetus after 20 weeks of gestation) plus induced terminations of pregnancy.

**Table 9. North Dakota Resident Deaths, 2001 Through 2005**

Year	Number of Resident Deaths	
	Infant Deaths*	Neonatal** Deaths
2001	63	42
2002	49	33
2003	57	46
2004	45	36
2005	50	36

\*An infant death is a death to a newborn less than 1 year old.

\*\*A neonatal death is a death to an individual less than 28 days old.

**Table 10. North Dakota Resident Birth Ratios, 2001 Through 2005**

Year	Resident Birth Ratios		
	Teenage Birth Ratios (Ages 15–19)*	Birth Ratios Among Unmarried Mothers**	Low Weight Birth Ratios (Less Than 2,500 Grams)***
2001	92.90	278.18	68.89
2002	84.72	290.14	64.22
2003	79.49	285.36	66.83
2004	77.15	299.18	66.27
2005	79.70	321.92	64.19

\*Ratio is equal to teenage live births divided by total resident live births multiplied by 1,000.

\*\*Ratio is equal to live births to unmarried mothers divided by total resident live births multiplied by 1,000.

\*\*\*Ratio is equal to low weight live births divided by total resident live births multiplied by 1,000.

**Table 11. North Dakota Resident Pregnancy Ratios, 2001 Through 2005**

Year	Resident Pregnancy Ratios	
	Teenage Pregnancy Ratios (Ages 15–19)*	Pregnancy Ratios Among Unmarried Women**
2001	105.17	326.75
2002	96.36	338.89
2003	89.87	334.69
2004	88.92	349.51
2005	88.69	365.79

\*Ratio is equal to teenage pregnancies divided by total resident pregnancies multiplied by 1,000.

\*\*Ratio is equal to out of wedlock pregnancies divided by total resident pregnancies multiplied by 1,000.

**Table 12. North Dakota Resident Death Ratios, 2001 Through 2005**

Year	Resident Death Ratios	
	Infant Death Ratios*	Neonatal Death Ratios**
2001	8.22	5.48
2002	6.32	4.26
2003	7.15	5.77
2004	5.50	4.40
2005	5.97	4.30

\*An infant death is a death to a newborn less than 1 year old. Ratio is equal to infant deaths divided by total resident live births multiplied by 1,000.

\*\*A neonatal death is a death to an individual less than 28 days old. Ratio is equal to neonatal deaths divided by total resident live births multiplied by 1,000.

**Table 13. North Dakota Resident Birth Rates, 2001 Through 2005**

Year	Resident Birth Rates	
	Total Birth Rates*	Teenage Birth Rates** (Ages 15–19)
2001	11.93	27.53
2002	12.08	25.41
2003	12.42	24.52
2004	12.74	24.40
2005	13.05	25.83

\*Rate is equal to total resident live births divided by total resident population multiplied by 1,000.

\*\*Rate is equal to teenage live births divided by female teen population multiplied by 1,000.

**Table 14. North Dakota Resident Pregnancy Rates, 2001 Through 2005**

Year	Resident Pregnancy Rates	
	Total Pregnancy Rates*	Teenage Pregnancy Rates** (Ages 15–19)
2001	13.16	34.38
2002	13.35	31.94
2003	13.76	30.71
2004	14.11	31.17
2005	14.55	32.06

\*Rate is equal to total pregnancies divided by total resident population multiplied by 1,000.

\*\*Rate is equal to teenage pregnancies divided by female teen population multiplied by 1,000.

**Table 15. North Dakota Resident Fertility Rates, 2001 Through 2005**

Year	Resident Fertility Rates
2001	56.61
2002	57.29
2003	58.92
2004	60.42
2005	61.91

\*Rate is equal to total resident live births divided by female population ages 15–44 multiplied by 1,000.

# North Dakota Birth Defects Monitoring System

## *North Dakota Birth Defects Monitoring System*

---

The North Dakota Birth Defects Monitoring System (NDBDMS) was established in 2003 as a means of identifying and collecting information about all babies born with certain birth defects in North Dakota. The monitoring program analyzes data about babies born with health problems to help researchers and health-care providers learn more about preventing these problems in the future. North Dakota monitors 43 major congenital anomalies in the categories of central nervous system, eye, ear, cardiovascular, orofacial, gastrointestinal, genitourinary, musculoskeletal, chromosomal and other birth defects.

The primary purpose of the NDBDMS is to provide a means for accurate and up-to-date tracking in order to determine if affected children have access to needed health-care and other services necessary to treat their condition.

The ICD-9 codes found in Table 16 come from the International Statistical Classification of Diseases and Related Health Problems and are used to help identify and classify birth defects. Definitions of the birth defects can be found in the “Glossary of Congenital Anomalies” in the Appendices. ICD-10 & CDC/BPA Codes can be found in Appendix Table 4.

**Table 16. Birth Defects Monitoring System ICD-9 Codes**

<b>Birth Defect</b>	<b>ICD-9 Code</b>
<b>Central nervous system</b>	
Anencephalus	740.00
Encephalocele	742.00
Hydrocephalus without spina bifida	742.30
Microcephalus	742.10
Spina bifida	741.00
<b>Eye</b>	
Aniridia	743.45
Anophthalmia/microphthalmia	743.00, 743.10
Congenital cataract	743.30–743.34
<b>Ear</b>	
Anotia/microtia	744.01, 744.23
<b>Cardiovascular</b>	
Aortic valve stenosis	746.30
Atrial septal defect	745.50
Coarctation of aorta	747.10
Common truncus	745.00
Ebstein's anomaly	746.20
Endocardial cushion defect	745.60, 745.61, 745.69
Hypoplastic left heart syndrome	746.70
Patent ductus arteriosus	747.00
Pulmonary valve anomalies	747.30

<b>Birth Defect</b>	<b>ICD-9 Code</b>
Pulmonary valve atresia and stenosis	746.01, 746.02
Tetralogy of Fallot	745.20
Transposition of great arteries	745.10, 745.11, 745.12, 745.19
Tricuspid valve atresia and stenosis	746.10
Ventricular septal defect	745.40
<b>Orofacial</b>	
Choanal atresia	748.00
Cleft lip with and without cleft palate	749.10
Cleft palate without cleft lip	749.00
<b>Gastrointestinal</b>	
Biliary atresia	751.61
Esophageal atresia/ tracheoesophageal fistula	750.30
Hirschsprung's disease	751.30
Pyloric stenosis	750.50
Rectal and large intestinal atresia/stenosis	751.20
<b>Genitourinary</b>	
Bladder exstrophy	753.50
Hypospadias and Epispadias	752.61, 752.62
Obstructive genitourinary defect	753.20, 753.60
Renal agenesis/hypoplasia	753.00
<b>Musculoskeletal</b>	
Congenital hip dislocation	745.30, 745.31, 745.35
Diaphragmatic hernia	756.60
Gastroschisis/omphalocele	756.79
Reduction deformity, lower limbs	755.30–755.39
Reduction deformity, upper limbs	755.20–755.29
<b>Chromosomal</b>	
Down syndrome (Trisomy 21)	758.00
Edwards syndrome (Trisomy 18)	758.20
Patau syndrome (Trisomy 13)	758.10
<b>Other</b>	
Amniotic Bands	-
Fetus or newborn affected by maternal alcohol use	760.01

# *Data Collection Methodology*

---

## **Description of Passive Surveillance Method**

The North Dakota Birth Defects Monitoring Program is a passive surveillance system. Data are collected and linked from three secondary data sources: vital records information, health-care claims data, and program information, specifically, the Children's Special Health Services program in the North Dakota Department of Health.

**Security and Confidentiality:** The NDBDMS will adhere to the “Standards for Privacy of Individually Identifiable Protected Health Information” as mandated by the Health Insurance Portability and Accountability Act of 1996 (HIPAA), as well as all policies and procedures established by the North Dakota Department of Health and the North Dakota Department of Health related to release or reporting of personally identifiable protected health information.

### **Case inclusion criteria:**

- Live birth to a North Dakota resident mother or a fetal death (at less than 20 weeks) from 2001 to 2005
- Verification of a diagnosed birth defects using ICD9-CM or ICD-10CM codes from at least one of the three data sources

### **Data collected:**

- Demographic information
- Risk factors
- Service utilization
- Health-care history

Five-year rates rather than single-year rates are used to improve statistical stability of the data. As a general rule, rates based on fewer than 20 observed cases are considered unreliable and are less likely to reflect true rates than are those based on a larger number of cases.

Prevalence rates are estimates of the true prevalence, which is unknown. Caution should be used when interpreting rates based upon a small number of events. Confidence intervals were calculated for North Dakota rates. The degree of precision or certainty of a rate is reflected by the width of the confidence interval, with a wider interval indicating less precision.

The prevalence of birth defects reported by surveillance systems can vary considerably due to differences in case definition, the method of case ascertainment and the types of data sources used. Major approaches to birth defects surveillance consist of active case ascertainment and passive case ascertainment. Active case ascertainment generally involves several staff members who confirm every diagnosis that is identified through data sources.

In the passive case ascertainment approach, the surveillance program receives case reports of birth defects from data sources. The completeness of ascertainment is highly dependent on the number of types of data sources used by the surveillance program and on the consistency of case reporting from the data source. Since the case reports usually are not confirmed by the staff in a passive case ascertainment program, it is particularly important for these programs to implement quality assurance procedures aggressively. In passive surveillance, the programs should evaluate

their surveillance approaches regularly for accuracy, completeness and timeliness and should be creative in identifying strategic means of quality improvement.

In passive surveillance, the level of interaction between programs and reporting sources varies, as does the method of reporting. Many programs merge or extract pertinent information into the surveillance program's database from a source's existing database. Many use a combination of reporting methods to develop as complete an identification of birth defects cases as possible within the resources available. Regardless of the method used, operating a surveillance system that receives case reports from data sources requires the program to identify and use multiple data reporting sources, provide detailed instructions to case reporting sources, nurture the relationship between the program and the reporting data source, and evaluate the quality and difference between the data source of the case reports received.

### **Review of Data Sources Used:**

#### **1. North Dakota Division of Vital Records**

All resident birth certificate, death certificate and fetal death certificate files in 2001 through 2005 were reviewed to determine if a diagnosis of one of the selected birth defects was made on the child's birth, death or fetal death certificate.

##### **A. Birth Certificates**

In order for a determination of the presence of one of the selected birth defects to be made from the birth certificate file, an ICD-9 or ICD-10 diagnosis must be present in one or more of the congenital anomaly description columns. A congenital anomaly code is not sufficient to determine the presence of a birth defect.

##### **B. Death Certificates**

In order for a determination of the presence of one of the selected birth defects to be made from the death certificate file, an ICD-9 or ICD-10 diagnostic code for one of the selected birth defects must be present in either the *Primary Cause of Death* or the *Contributing Cause of Death* column.

##### **C. Fetal Death Certificate (< 20 weeks gestation)**

In order for a determination of the presence of one of the selected birth defects to be made from the fetal death certificate file, an ICD-9 or ICD-10 diagnostic code for one of the selected birth defects must be present in one or more of the congenital anomaly description columns or in either the *Primary Cause of Death* or the *Contributing Cause of Death* column.

#### **2. Children's Special Health Services (CSHS) Program Data**

A list of children with any of the selected diagnoses and a date of birth of January 1 to December 31, 2001, 2002, 2003, 2004, and 2005, was generated from the CSHS database. The data are captured for children born and served for each year.

A. The child's CSHS case file is reviewed to substantiate the diagnosis. Medical reports, clinic reports and any other diagnostic information are reviewed in the child's case file to substantiate the diagnosis.

### **3. Health Care Claims**

Inpatient and outpatient health-care claims are accessed through the Med Stat Data Probe system. Health-care claims available from the North Dakota Department of Human Services (Medicaid) for this report began in calendar year 2001 and ended in year 2005.

### **Master Lists of Birth Defects Registry:**

#### **1. Matched Master List**

The Matched Master List contains the child's name; unique identification number; birth, death or fetal death certificate number; and an indication as to which data source(s) contained the diagnostic information.

#### **2. Unmatched Master List**

The Unmatched Master List contains the (1) names of individuals from CSHS for whom no diagnosis has been confirmed from either CSHS case files, vital records or a health-care claim and (2) unique identification number of individuals for whom a health-care claim had been identified but no name is known.

# Description of Birth Defects and Results

# Overview of Major Congenital Anomalies

---

The major categories of birth defects tracked in North Dakota are organized according to the anatomical site of the anomaly. The categories in which there were 10 or more incidents in 2001 through 2005 in North Dakota include central nervous system defects (also referred to as neural tube defects), cardiovascular defects, orofacial defects, gastrointestinal defects, genitourinary defects, musculoskeletal defects, chromosomal defects and other defects. Eye defects and ear defects also are tracked.

There were 39,955 live births in North Dakota from 2001 through 2005. Of these live births, 1,240 (3.1 percent) had major congenital anomalies, although some babies may have one or more congenital anomalies.

The proportions of anomalies by category for all birth defects in North Dakota from 2001 through 2005 are as follows:

- Cardiovascular defects – 57.7 percent
- Gastrointestinal defects – 10.4 percent
- Central nervous system defects – 8.0 percent
- Orofacial defects – 6.7 percent
- Genitourinary defects – 6.1 percent
- Chromosomal defects – 4.8 percent
- Musculoskeletal defects – 3.6 percent
- Chromosomal defects – 1.8 percent
- Other\* defects – 1.7 percent

\*Fetus or newborn affected by maternal alcohol use; amniotic bands.

# Central Nervous System Defects

Birth defects of the central nervous system, also known as neural tube defects, are thought to be caused by interacting genetic and environmental factors. Causes of central nervous system anomalies include gene mutations, exposure of the fetus to alcohol or other toxic elements, and deficiencies of critical nutrients such as folic acid. Studies have reported increased risks of specific central nervous system anomalies among obese women and women with diabetes. Women who undergo surgery with general anesthesia early in pregnancy may be at increased risk of delivering an infant with a central nervous system anomaly.<sup>(22)</sup>

Central nervous system defects accounted for 8.0 percent of all birth defects in North Dakota from 2001 through 2005. A profile for each of the four central nervous system defects with 10 or more incidents is provided in this section.

<b>Central Nervous System Defects.....</b>	<b>28–23</b>
<i>Anencephalus</i> .....	29
<i>Hydrocephalus Without Spina Bifida</i> .....	30
<i>Microcephalus</i> .....	31
<i>Spina Bifida Without Anencephalus</i> .....	32

# Central Nervous System Defects

## ➤ Anencephalus



(A)

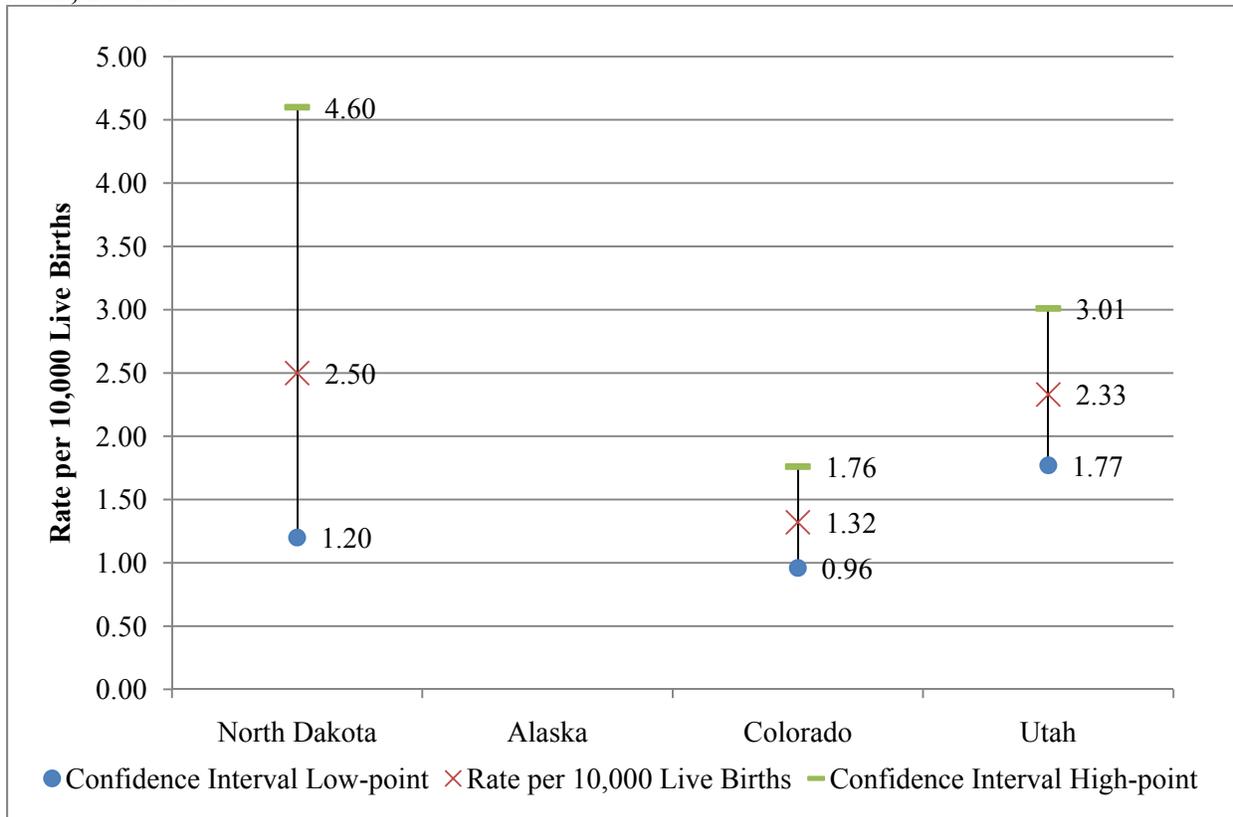
### Definition:

Anencephaly is a birth defect that affects the closing of the neural tube during pregnancy. The neural tube is a narrow channel that folds and closes during the third and fourth weeks of pregnancy to form the brain and spinal cord. Anencephaly occurs when the portion of the neural tube that forms the brain does not close. This results in the baby lacking parts of the brain, skull and scalp. Babies with this condition often are born without a forebrain (the front part of the brain) and a cerebrum (the thinking and coordinating part of the brain). The remaining brain tissue is often exposed (i.e., it is not covered by bone or skin).<sup>(2)</sup>

### Prevalence:

North Dakota has an anencephalus rate of 2.50 per every 10,000 live births. The five-year rate of anencephalus in North Dakota is statistically comparable to Colorado and Utah.

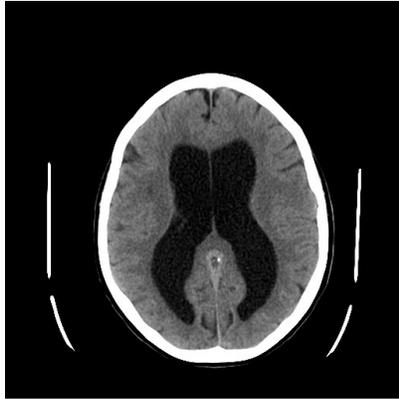
**Figure 1. North Dakota's Anencephalus Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



Note: No data were provided for Alaska.

# Central Nervous System Defects

## ➤ Hydrocephalus Without Spina Bifida



(K)

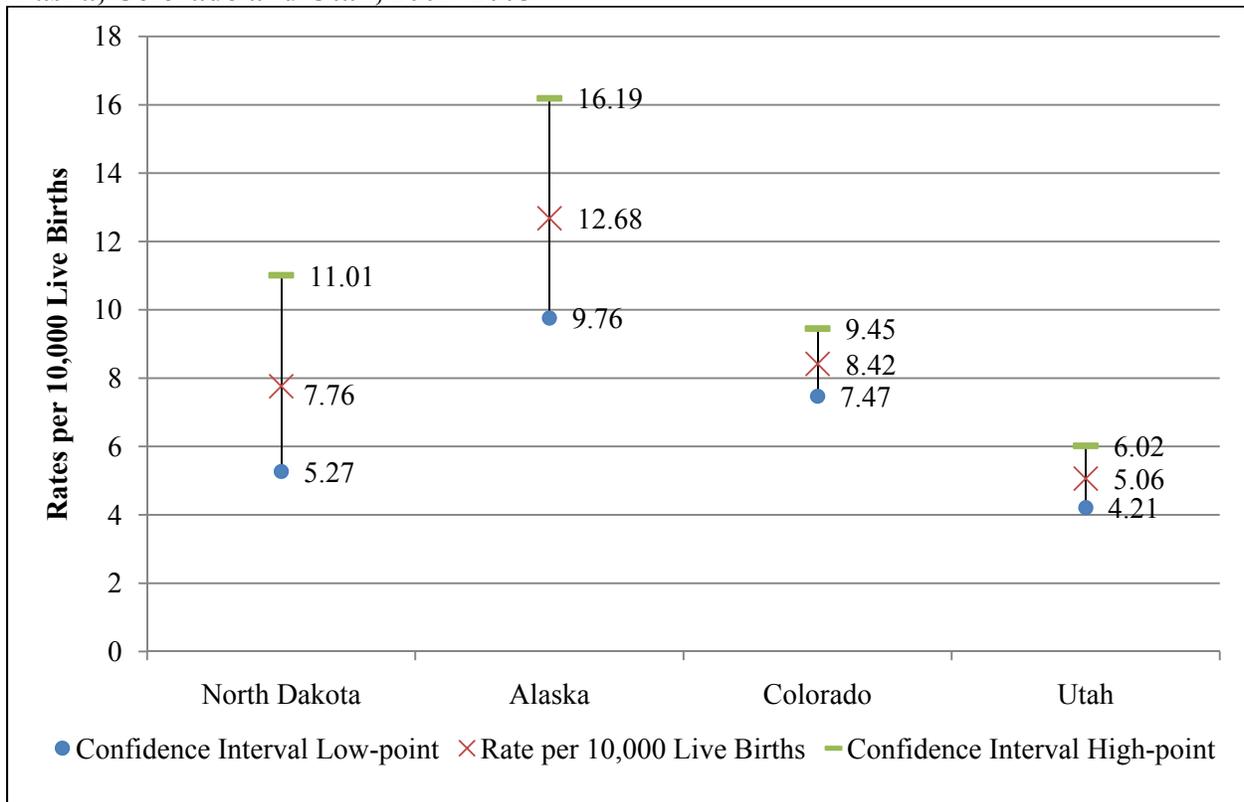
### Definitions:

Hydrocephalus is an abnormal buildup of cerebrospinal fluid (CSF) in the ventricles of the brain. The buildup of CSF can be caused by a blockage in its circulation or absorption, or when too much CSF is produced. The excess fluid causes pressure on the brain, which can result in brain damage. Hydrocephalus is treated by addressing the underlying cause or palliated through surgical insertion of a shunt to improve the flow of CSF. Advances in diagnostic imaging technology may contribute to an increasing trend in the number of diagnoses for hydrocephalus.<sup>(14)</sup>

### Prevalence:

North Dakota has a hydrocephalus rate of 7.76 per every 10,000 live births. The five-year rate of hydrocephalus in North Dakota is statistically comparable to Alaska, Colorado, and Utah.

**Figure 2. North Dakota’s Hydrocephalus Without Spina Bifida Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Central Nervous System Defects

## ➤ Microcephalus



(P)

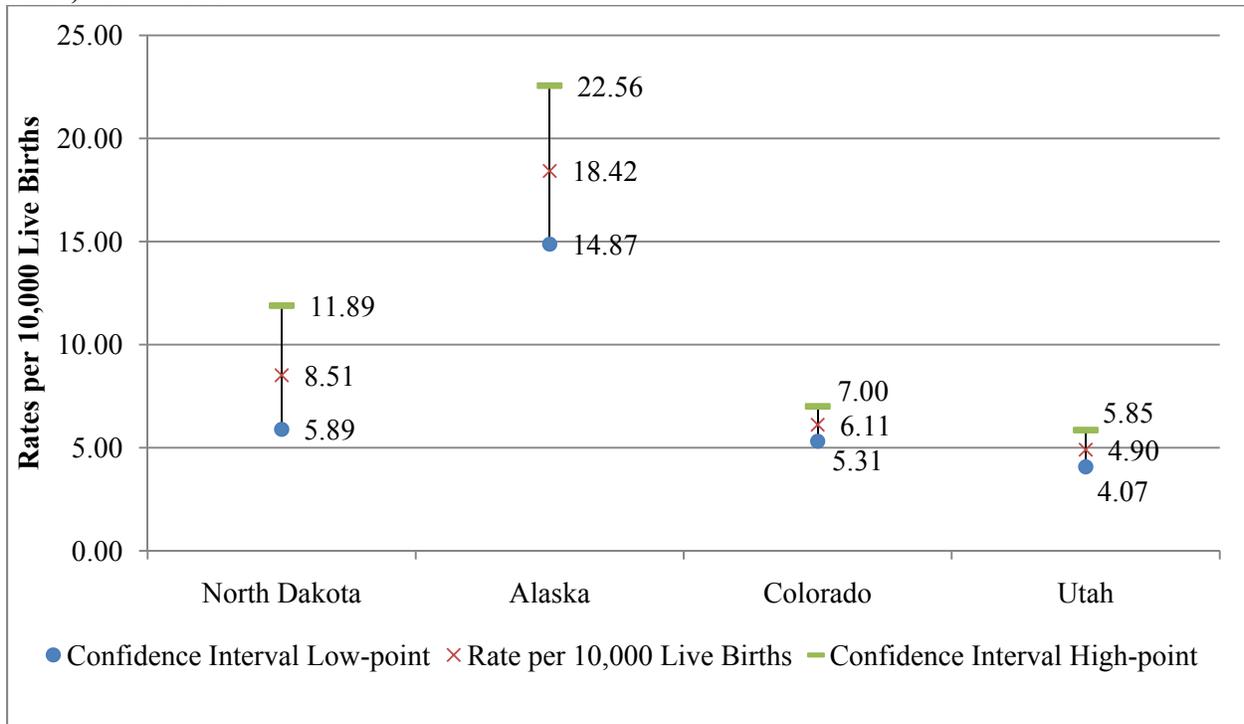
### Definition:

Congenital microcephalus refers to an infant born with a head circumference that is less than the 10th percentile for gestational age and may be caused by improper or incomplete development of the brain. While there is no physical treatment for microcephalus, pediatric neurologists and early childhood intervention teams can treat the neurological and social disabilities that often accompany the anomaly. These disabilities can include mental retardation, delayed motor functions, facial distortions, dwarfism, hyperactivity and seizures.<sup>(22)</sup>

### Prevalence:

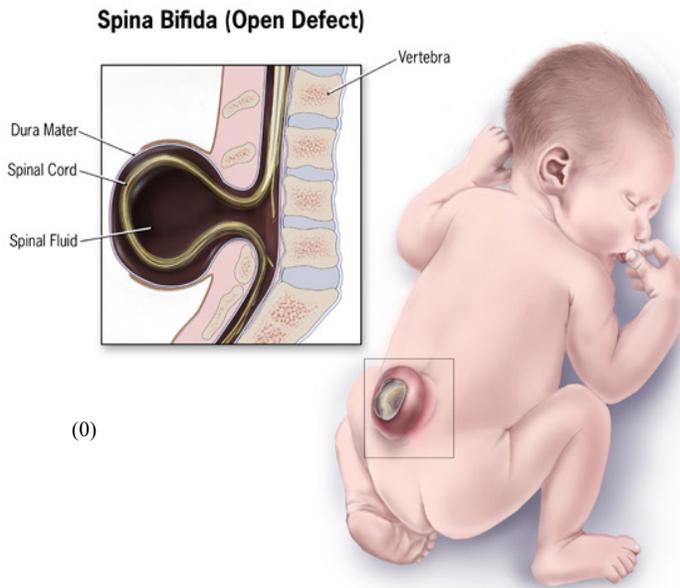
North Dakota has a microcephalus rate of 8.51 per every 10,000 live births. The five-year rate of microcephalus in North Dakota is statistically comparable to Colorado. North Dakota's rate is lower than Alaska but slightly higher than Utah.

**Figure 3. North Dakota's Microcephalus Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Central Nervous System Defects

## ➤ *Spina Bifida Without Anencephalus*



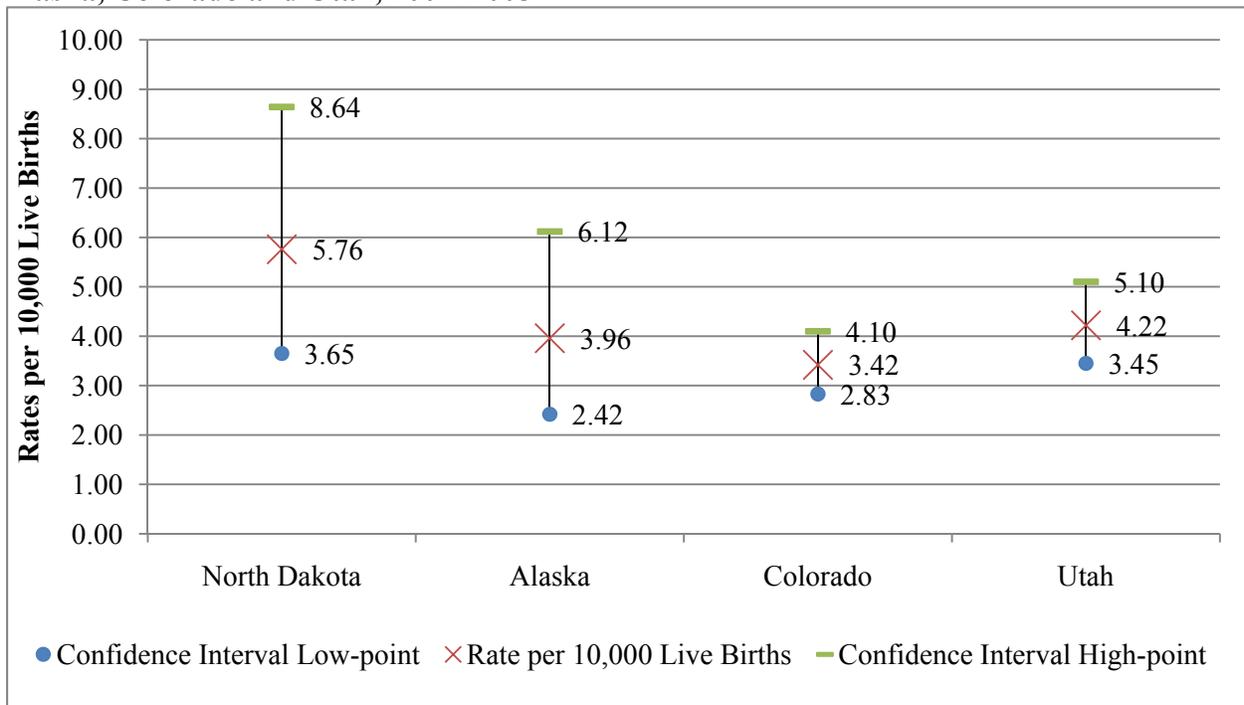
### Definition:

Spina bifida is a type of neural tube defect (NTD). The neural tube is a narrow channel that folds and closes during the third and fourth weeks of pregnancy to form the brain and spinal cord. Spina bifida happens if the portion of the neural tube that forms the spinal cord does not close completely during the first month of pregnancy.<sup>(23)</sup>

### Prevalence:

North Dakota has a spina bifida rate of 5.76 per every 10,000 live births. The five-year rate of spina bifida in North Dakota is statistically comparable to Alaska, Colorado, and Utah.

**Figure 4. North Dakota's Spina Bifida without Anencephalus Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Cardiovascular Defects

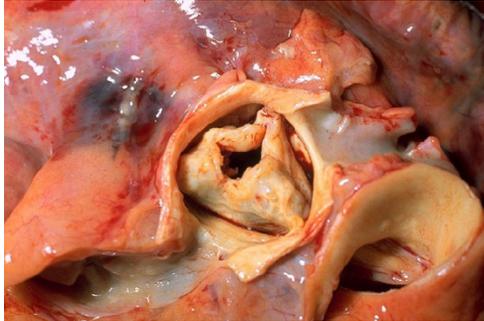
Cardiovascular birth defects affect the heart or blood vessels surrounding the heart. Cardiovascular defects are generally estimated to be present in about 1 percent of live births in the United States and are the most commonly diagnosed congenital anomalies. Cardiovascular anomalies usually result in either obstructed or abnormal blood flow to or from the heart. They range in seriousness from minor self-correcting anomalies to fatal conditions. Prevalence estimates for cardiovascular anomalies are highly influenced by the availability of modern diagnostic techniques that can identify less serious defects.<sup>(22)</sup>

Cardiovascular defects accounted for 57.7 percent of all birth defects in North Dakota from 2001 through 2005. A profile for each of the 10 cardiovascular defects with 10 or more incidents is provided in this section.

<b>Cardiovascular Defects.....</b>	<b>33–43</b>
<i>Aortic Valve Stenosis .....</i>	<i>34</i>
<i>Atrial Septal Defect.....</i>	<i>35</i>
<i>Coarctation of Aorta.....</i>	<i>36</i>
<i>Endocardial Cushion Defect.....</i>	<i>37</i>
<i>Hypoplastic Left Heart Syndrome.....</i>	<i>38</i>
<i>Patent Ductus Arteriosus .....</i>	<i>39</i>
<i>Pulmonary Valve Atresia and Stenosis.....</i>	<i>40</i>
<i>Tetralogy of Fallot.....</i>	<i>41</i>
<i>Transposition of Great Arteries.....</i>	<i>42</i>
<i>Ventricular Septal Defect.....</i>	<i>43</i>

# Cardiovascular Defects

## ➤ Aortic Valve Stenosis



(G)

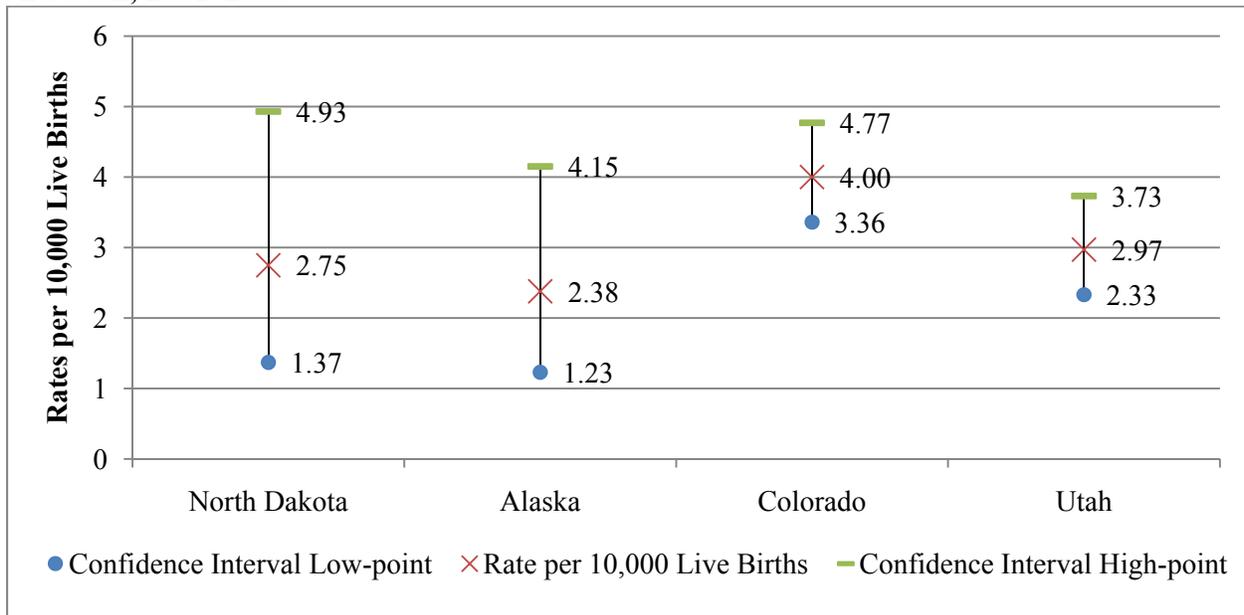
### Definition:

An aortic valve stenosis is a cardiac anomaly characterized by a narrowing or stricture of the aortic valve. This condition causes abnormal cardiac circulation and pressure in the heart during contractions. This condition can be repaired surgically in some cases.<sup>(19)</sup>

### Prevalence:

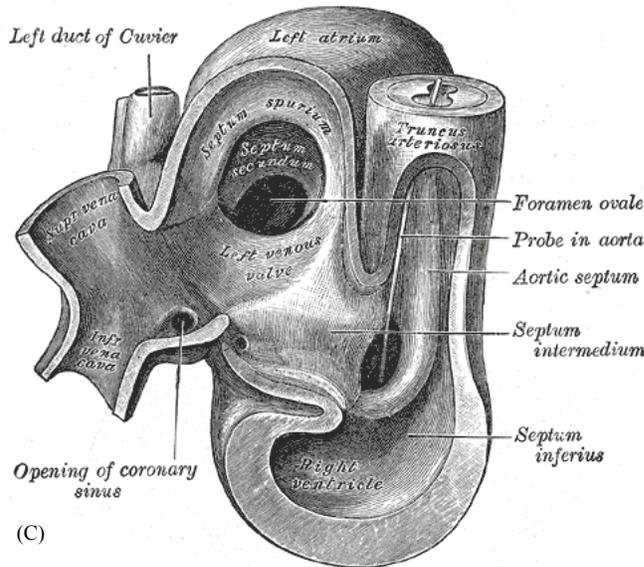
North Dakota has an aortic valve stenosis rate of 2.75 per every 10,000 live births. The five-year rate of aortic valve stenosis in North Dakota is statistically comparable to Alaska, Colorado and Utah.

**Figure 5. North Dakota's Aortic Valve Stenosis Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Cardiovascular Defects

## ➤ Atrial Septal Defect



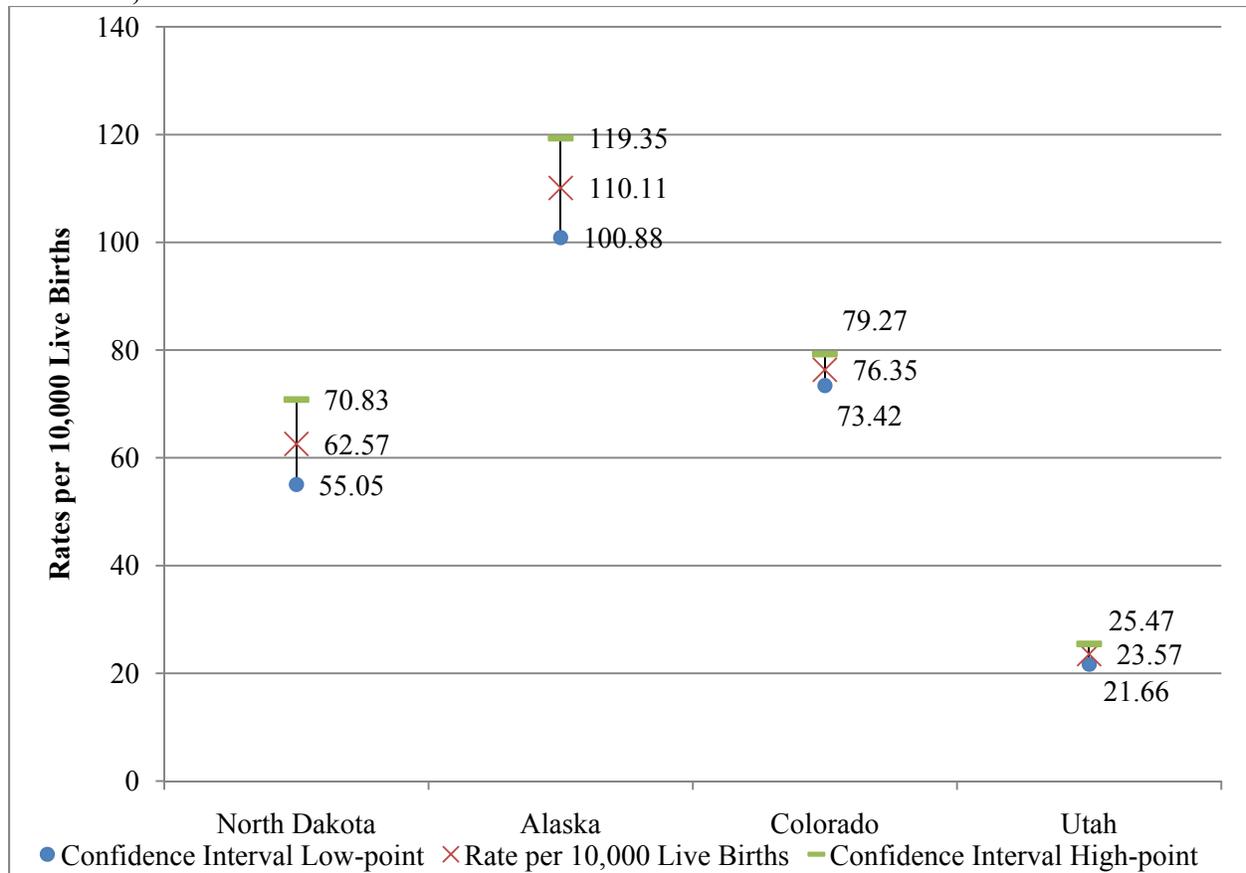
### Definition:

An atrial septal defect (ASD) is a hole (defect) in the wall (septum) that separates the two upper chambers of the heart, called atria. This hole between the heart chambers disrupts the flow of blood and oxygen to the body.<sup>(3)</sup>

### Prevalence:

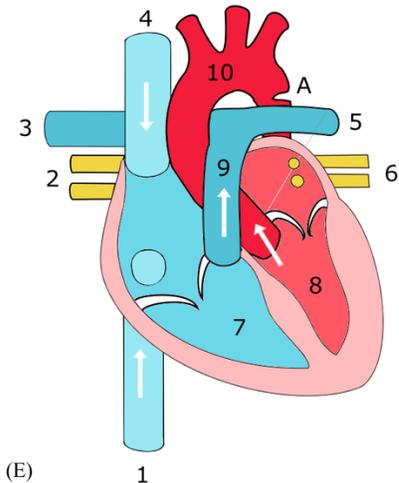
North Dakota has an atrial septal defect rate of 62.57 per every 10,000 live births. The five-year rate of atrial septal defect in North Dakota is not statistically comparable to Alaska, Colorado or Utah. North Dakota's rate is much lower than Alaska, lower than Colorado, and much higher than Utah.

**Figure 6. North Dakota's Atrial Septal Defect Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Cardiovascular Defects

## ➤ Coarctation of Aorta



### Definition:

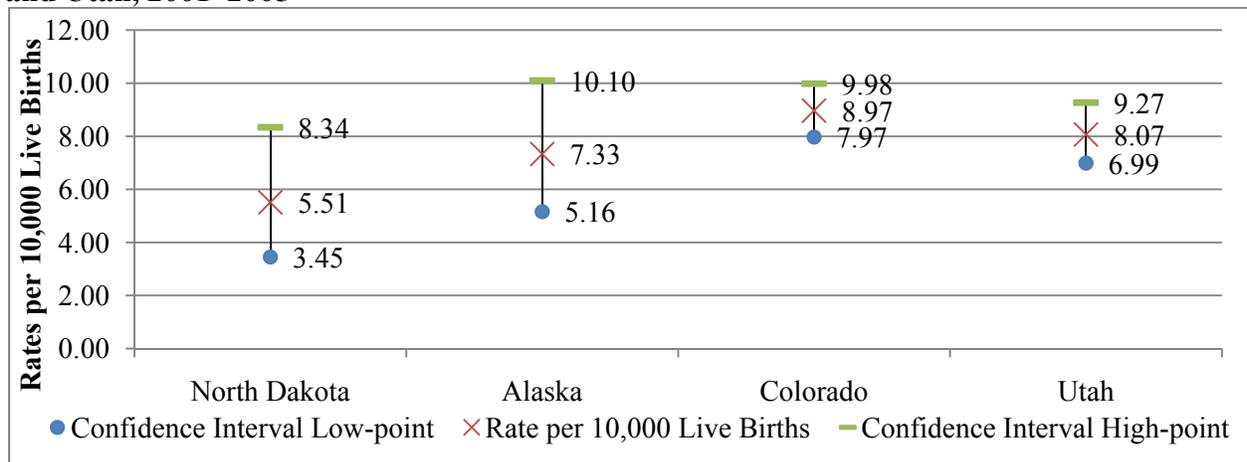
Coarctation of the aorta is localized narrowing of the aorta. This condition causes abnormal cardiac circulation and pressure in the heart during contractions. This condition can vary from mild to severe. Surgical correction is recommended even for mild defects.<sup>(13)</sup>

### Prevalence:

North Dakota has a coarctation of aorta rate of 5.51 per every 10,000 live births. The five-year rate of coarctation of aorta in North Dakota is statistically comparable to Alaska, Colorado and Utah.

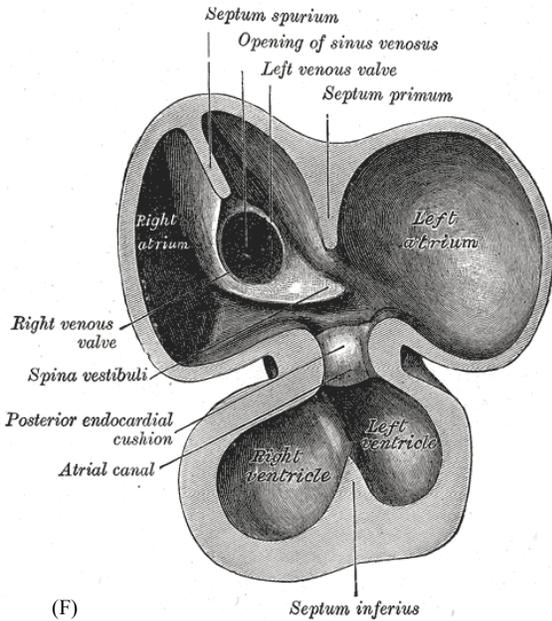
A: Coarctation (narrowing) of the aorta. 1: Inferior vena cava, 2: Right pulmonary veins, 3: Right pulmonary artery, 4: Superior vena cava, 5: Left pulmonary artery, 6: Left pulmonary veins, 7: Right ventricle, 8: Left ventricle, 9: Pulmonary artery, 10: Aorta

**Figure 7. North Dakota's Coarctation of Aorta Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Cardiovascular Defects

## ➤ Endocardial Cushion Defect



(F)

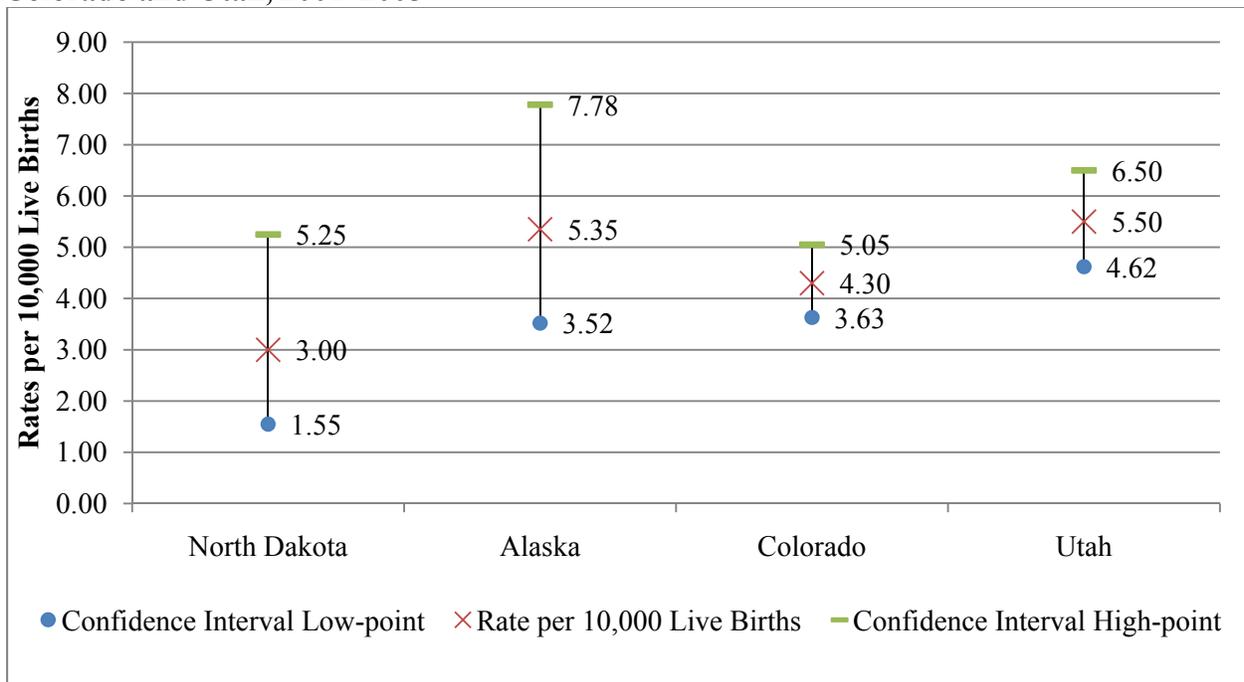
### Definition:

Endocardial cushion refers to a subset of cells in the primordial heart that play a vital role in proper heart septation. They develop on the atrioventricular canal. During development, the heart starts out as a tube. As heart development continues, this tube undergoes conformational changes and remodeling to eventually form the four-chambered heart. The endocardial cushions are a subset of cells found in the developing heart tube that will give rise to the heart's valves and septa critical to the proper formation of a four-chambered heart.<sup>(10)</sup>

### Prevalence:

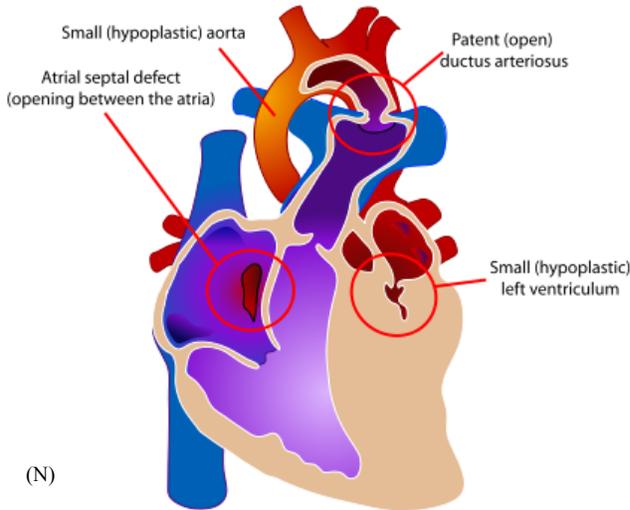
North Dakota has an endocardial cushion defect rate of 3.00 per every 10,000 live births. The five-year rate of endocardial cushion defects in North Dakota is statistically comparable to Alaska, Colorado and Utah.

**Figure 8. North Dakota's Endocardial Cushion Defect Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Cardiovascular Defects

## ➤ Hypoplastic Left Heart Syndrome



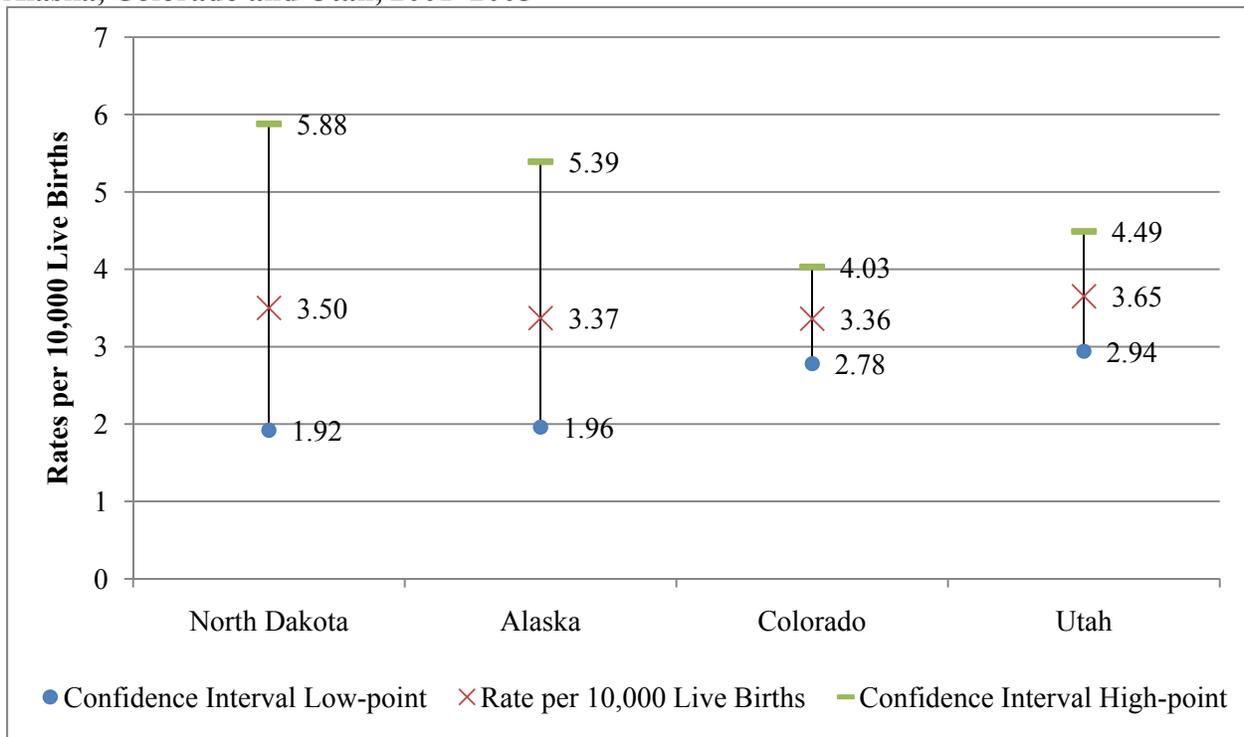
### Definition:

Hypoplastic left heart syndrome is a problem with the heart's structure that is present at birth (congenital). It is a group of related defects that, together, mean that the left side of the heart is underdeveloped. Normally, oxygen-poor blood is pumped through the right side of the heart to the lungs, where it gains oxygen and returns to the left side of the heart. The oxygen-rich blood is then pumped from the left side of the heart to the rest of the body. At birth, all babies also have two connections, or shunts, between the two sides of the heart; however, within a few days of birth these connections close.<sup>(15)</sup>

### Prevalence:

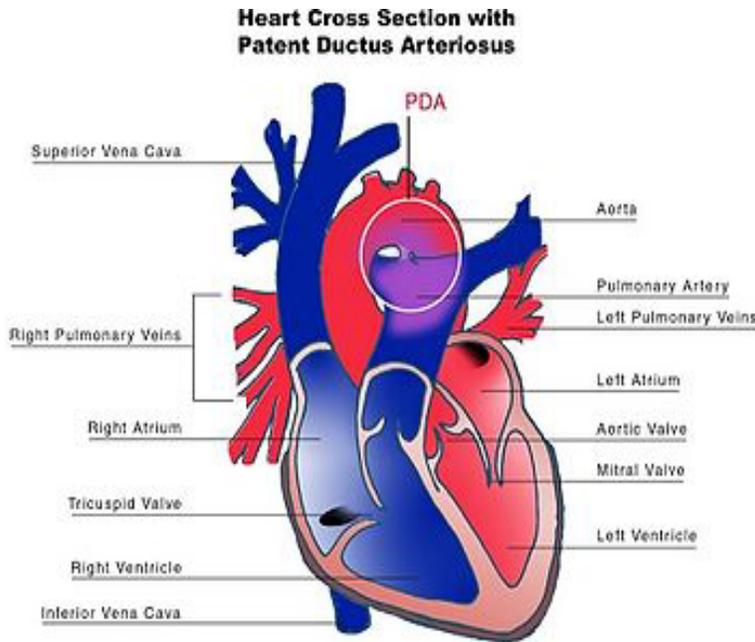
North Dakota has a hypoplastic left heart syndrome rate of 3.50 per every 10,000 live births. The five-year rate of hypoplastic left heart syndrome in North Dakota is statistically comparable to Alaska, Colorado and Utah.

**Figure 9. North Dakota's Hypoplastic Left Heart Syndrome Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Cardiovascular Defects

## ➤ Patent Ductus Arteriosus



(B)

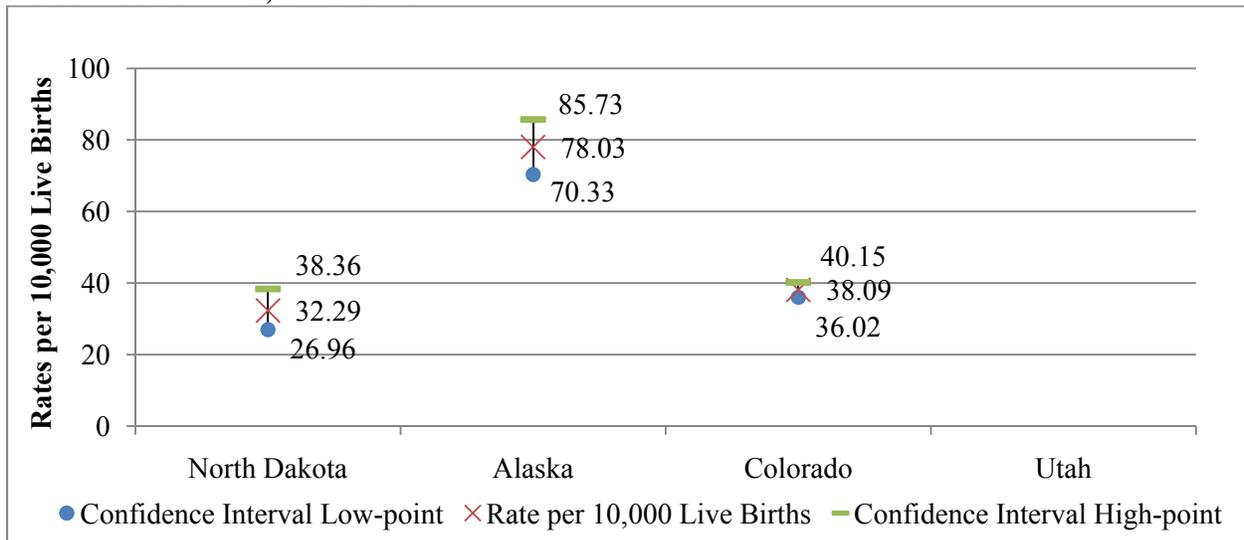
### Definition:

This congenital heart defect is characterized by persistence of the fetal blood vessel connecting the pulmonary artery and the aorta. A patent ductus arteriosus (PDA) is a condition where the ductus arteriosus fails to close within 10 days after birth. PDAs are one of the more common congenital heart defects, particularly in preterm infants. (18)

### Prevalence:

North Dakota has a patent ductus arteriosus rate of 32.29 per every 10,000 live births. The five-year rate of patent ductus arteriosus in North Dakota is statistically comparable to Colorado. North Dakota's rate is much lower than Alaska.

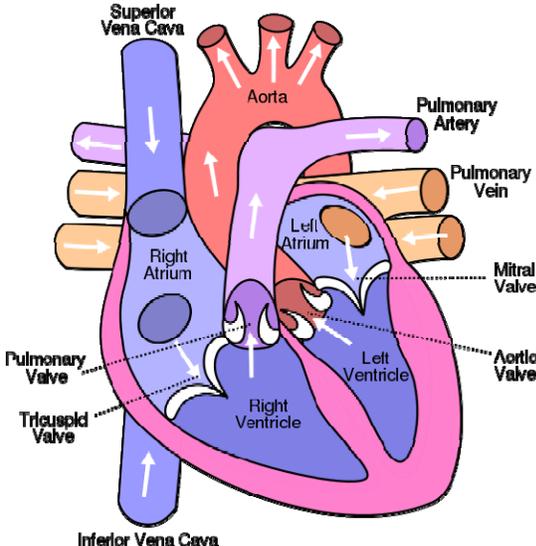
**Figure 10. North Dakota's Patent Ductus Arteriosus Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



Note: No data were provided for Utah.

# Cardiovascular Defects

## ➤ Pulmonary Valve Atresia and Stenosis

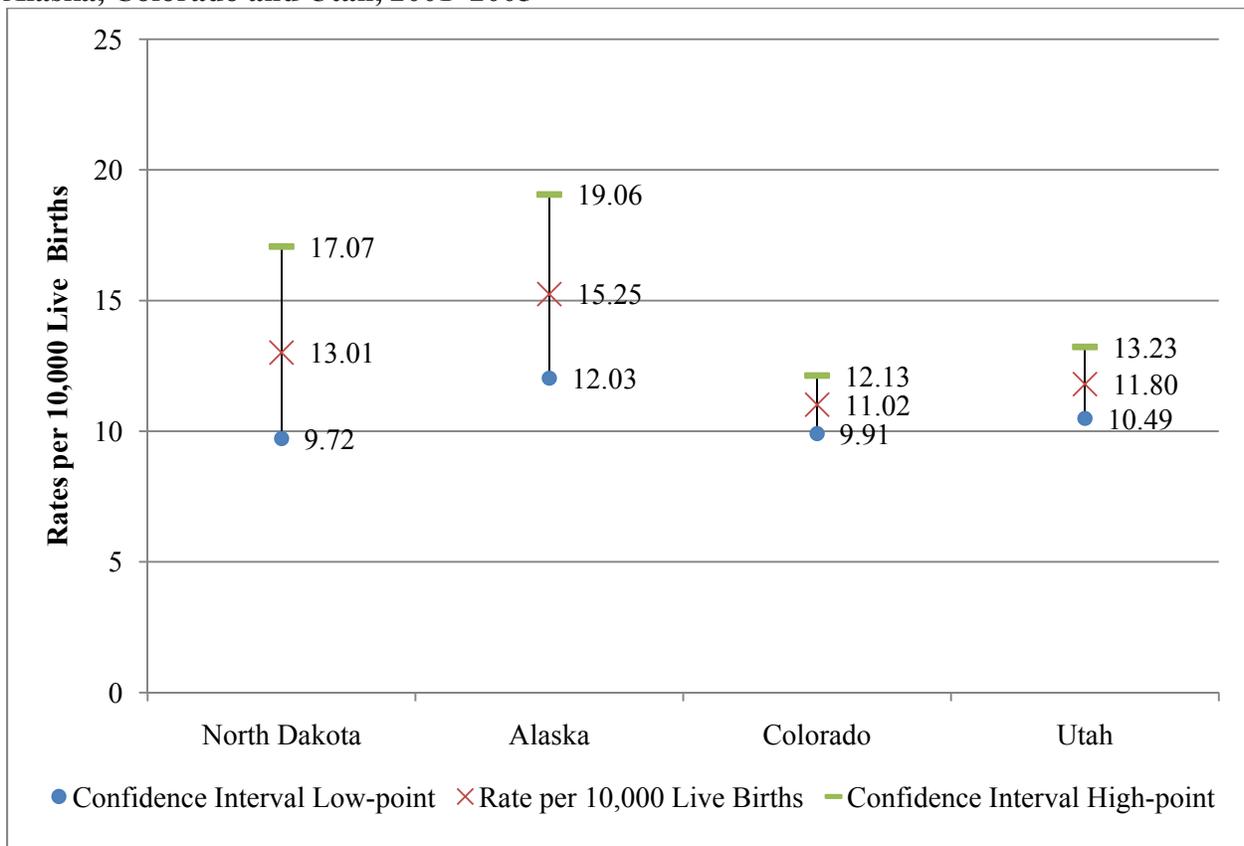


(L)

**Definition:**  
 Pulmonary valve atresia and stenosis is a congenital heart condition characterized by absence or constriction of the pulmonary valve. This condition causes abnormal cardiac circulation and pressure in the heart during contractions. This condition can vary from mild to severe. Mild forms are relatively well tolerated and require no intervention. More severe forms are surgically corrected.<sup>(13)</sup>

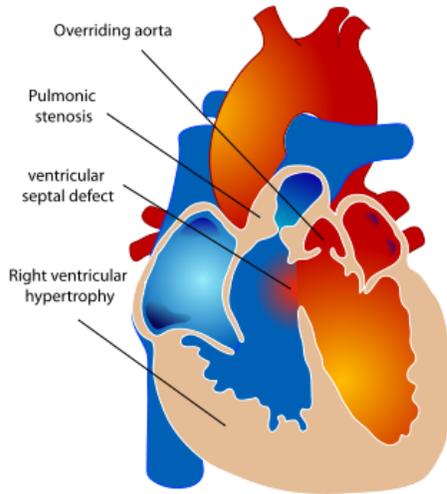
**Prevalence:**  
 North Dakota has a pulmonary valve atresia and stenosis rate of 13.01 per every 10,000 live births. The five-year rate of pulmonary valve stenosis in North Dakota is statistically comparable to Alaska, Colorado and Utah.

**Figure 11. North Dakota’s Pulmonary Valve Atresia and Stenosis Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Cardiovascular Defects

## ➤ Tetralogy of Fallot



### Definition:

Tetralogy of Fallot is a problem with the heart's structure that is present at birth. This defect changes the normal flow of blood through the heart. Tetralogy of Fallot is a combination of four defects:

- A hole in the wall between the ventricles (two lower chambers of the heart), called a ventricular septal defect.
- Narrowing of the tube that carries blood from the heart to the lungs, called pulmonary stenosis.
- The aorta (the tube that carries oxygen-rich blood to the body) grows from both ventricles, rather than from the left ventricle only.
- A thickened muscular wall of the right ventricle, called right ventricular hypertrophy.

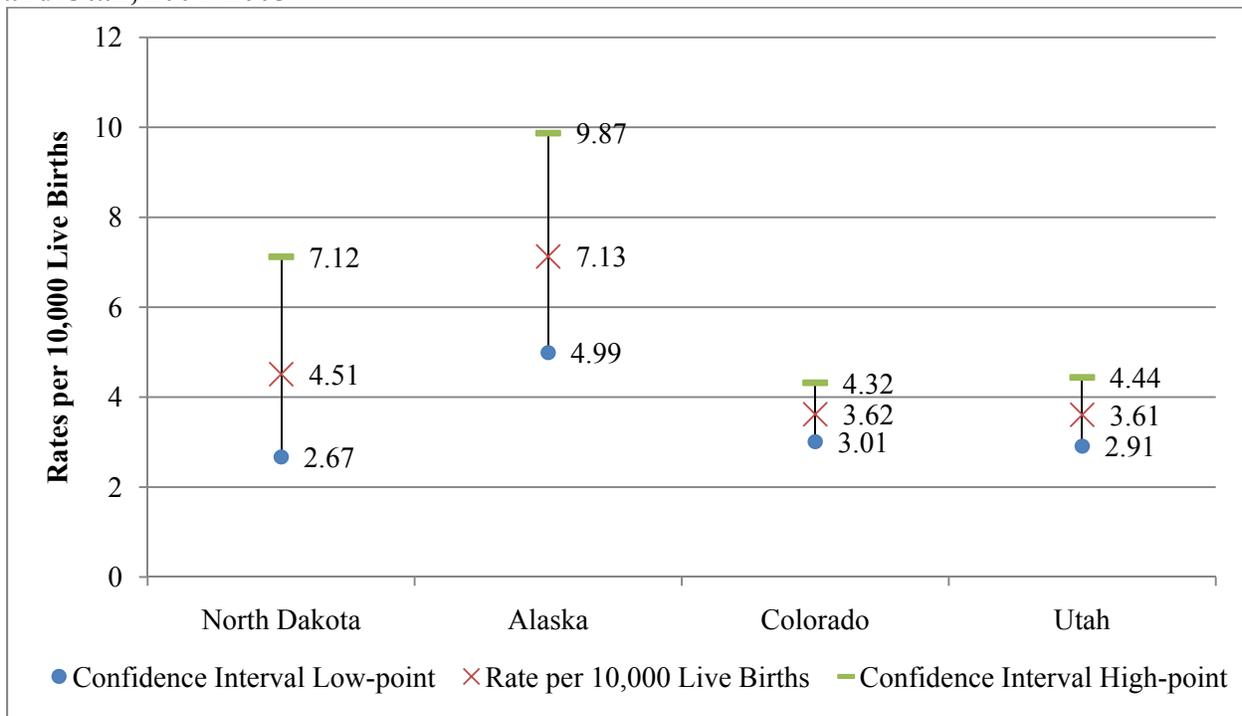
Infants and young children with tetralogy of Fallot often have blue- or purplish-looking skin color, called cyanosis, because of oxygen-poor blood.<sup>(24)</sup>

(Q)

### Prevalence:

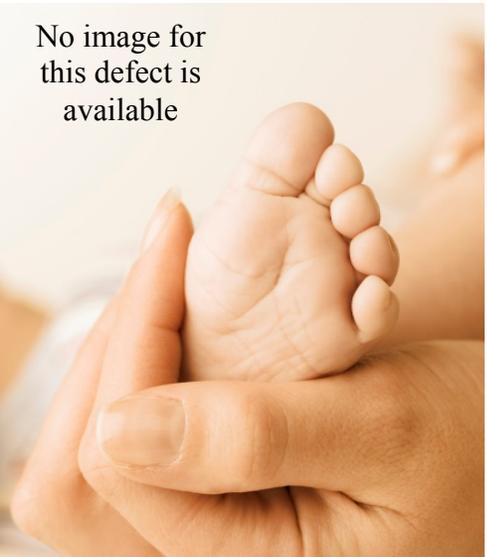
North Dakota has a tetralogy of Fallot rate of 4.51 per every 10,000 live births. The five-year rate of tetralogy of Fallot in North Dakota is statistically comparable to Alaska, Colorado and Utah.

**Figure 12. North Dakota's Tetralogy of Fallot Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Cardiovascular Defects

## ➤ Transposition of Great Arteries



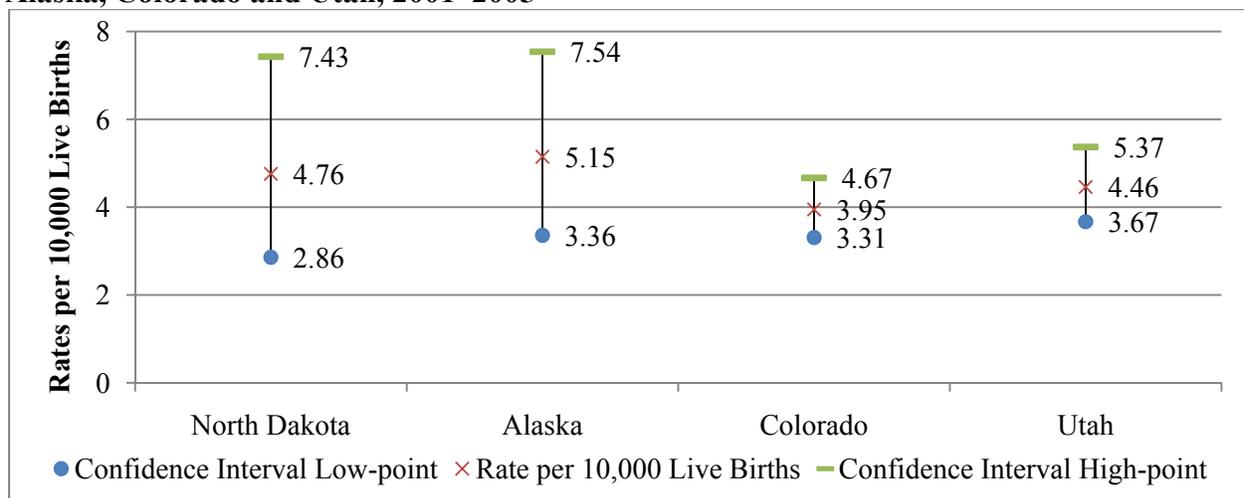
### Definition:

Transposition of great arteries (TGA) is a heart condition that is present at birth and often is called a congenital heart defect. TGA occurs when the two main arteries going out of the heart – the pulmonary artery and the aorta – are switched in position, or “transposed.” Normally, blood returning to the heart from the body is pumped from the right side of the heart through the pulmonary artery to the lungs. There, it receives oxygen and returns to the left side of the heart. Then, the oxygen-rich blood is pumped from the left side of the heart through the aorta to the body. In TGA, blood returning from the body bypasses the lungs and is pumped back out to the body. This occurs because the main connections are reversed. The pulmonary artery, which normally carries oxygen-poor blood from the right side of the heart to the lungs, now arises from the left side and carries oxygen-rich blood returning from the lungs back to the lungs. The aorta, which normally carries blood from the left side of the heart to the body, now arises from the right side and carries oxygen-poor blood back out to the body. The result of transposition of these two vessels is that too little oxygen is in the blood that is pumped from the heart to the rest of the body.<sup>(25)</sup>

### Prevalence:

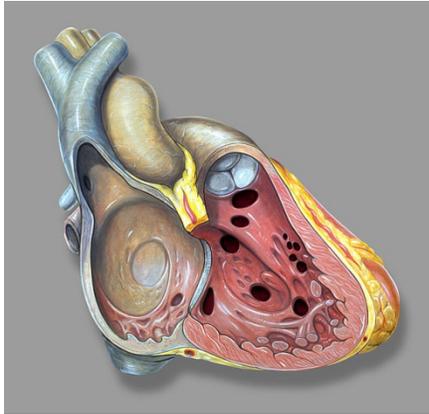
North Dakota has a transposition of great arteries rate of 4.76 per every 10,000 live births. The five-year rate of transposition of great arteries in North Dakota is statistically comparable to Alaska, Colorado and Utah.

**Figure 13. North Dakota’s Transposition of Great Arteries Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Cardiovascular Defects

## ➤ *Ventricular Septal Defect*



(R)

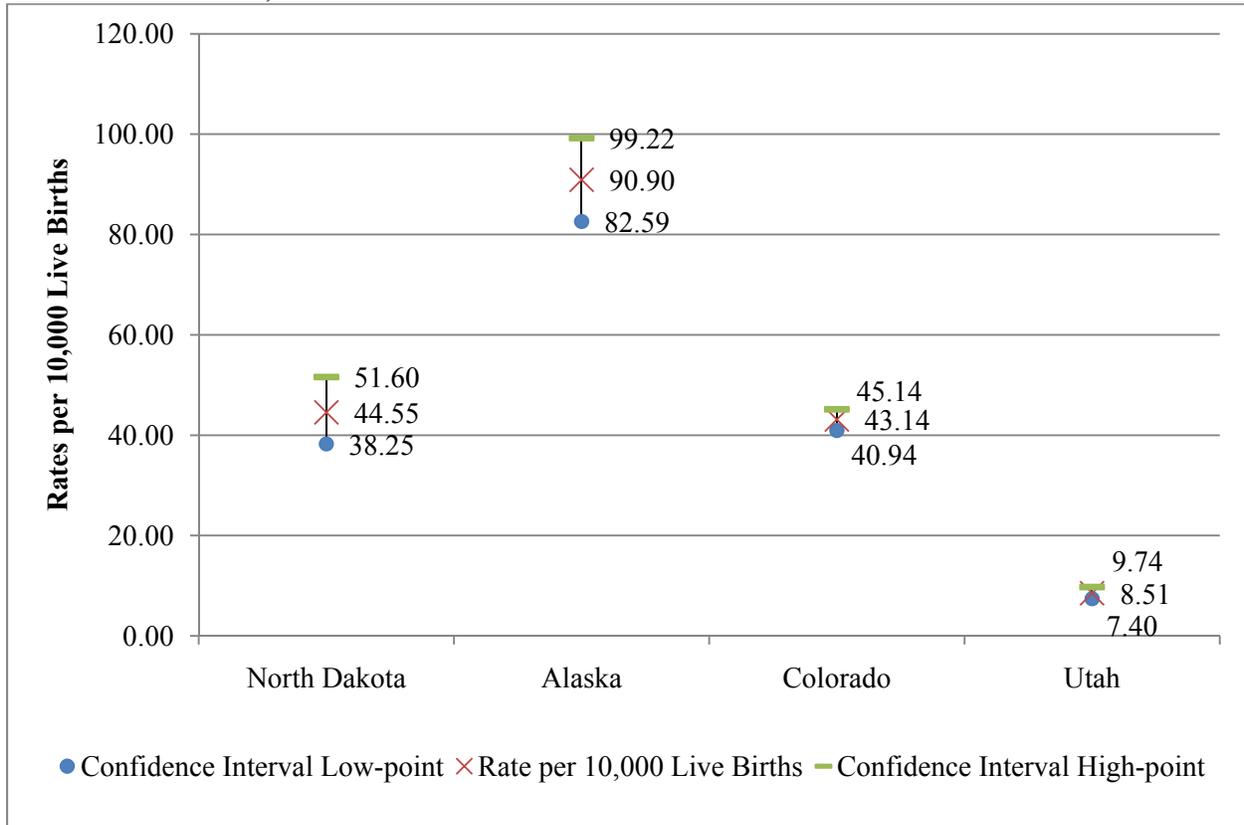
### Definition:

A ventricular septal defect (VSD) is a heart condition that is present at birth and often is called a congenital defect. A VSD occurs when there is a hole (defect) in the wall (septum) that separates the two lower chambers of the heart, called ventricles. This hole between the heart chambers disrupts the flow of blood and oxygen to the body.<sup>(28)</sup>

### Prevalence:

North Dakota has a ventricular septal rate of 44.55 per every 10,000 live births. The five-year rate of ventricular septal defect is statistically similar to Colorado. North Dakota's rate is much lower than Alaska and much higher than Utah.

**Figure 14. North Dakota's Ventricular Septal Defect Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Orofacial Defects

---

Orofacial clefts are birth defects where the mouth or the roof of mouth does not form properly. A cleft is a separation in a body structure, often resulting from the failure of tissues to grow together properly. Oral facial clefts may involve the lip, the roof of the mouth (hard palate) or the soft tissue in the back of the mouth (soft palate).<sup>(17)</sup>

Orofacial defects accounted for 6.7 percent of all birth defects in North Dakota from 2001 through 2005. A profile for the two orofacial defects with 10 or more incidents is provided in this section.

<b>Orofacial Defects</b> .....	<b>44-46</b>
<i>Cleft Lip With and Without Cleft Palate</i> .....	45
<i>Cleft Palate Without Cleft Lip</i> .....	46

# Orofacial Defects

## ➤ Cleft Lip With and Without Cleft Palate



Baby with cleft lip

(D)

### Definition:

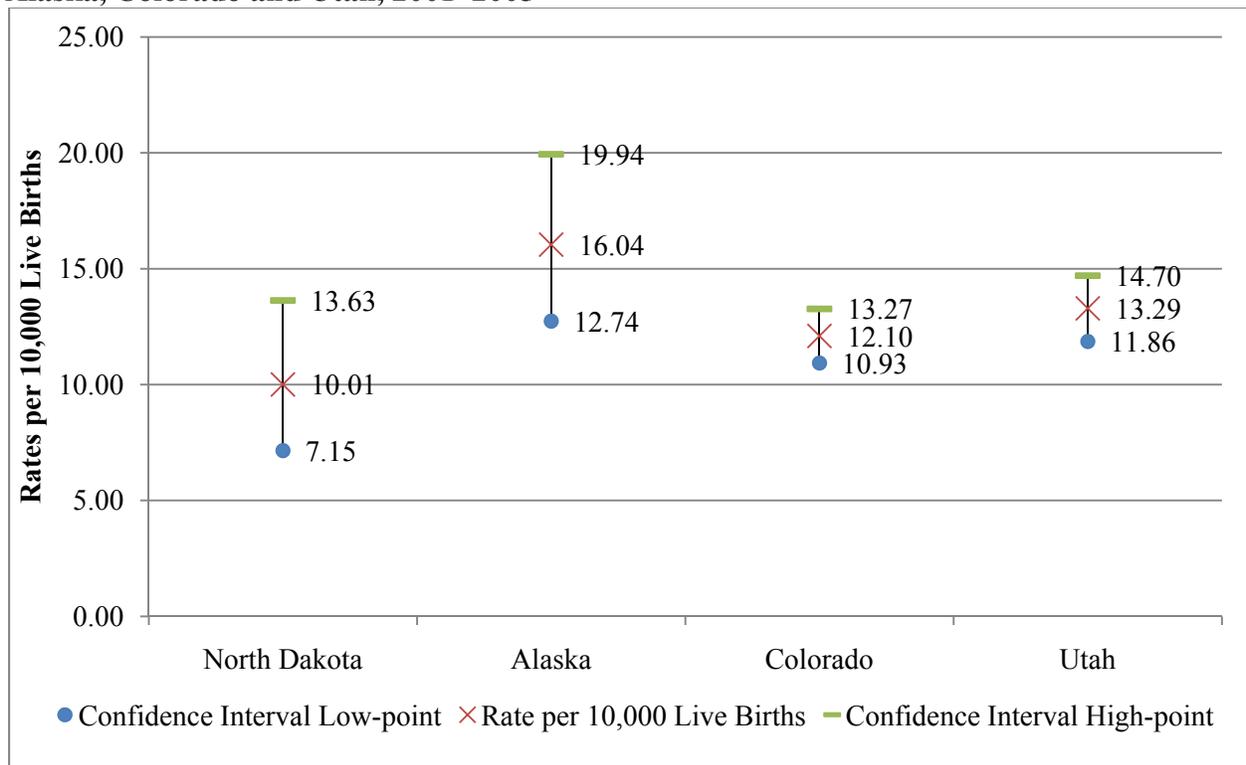
A cleft lip is an opening in the upper lip. The lip is formed early in pregnancy (at about four to seven weeks of pregnancy). The opening in the lip can be a small slit in the lip or a large opening that goes through the lip into the nose. A cleft lip can be on one or both sides of the lip, or more rarely, can be in the middle. Children with a cleft lip also can have a cleft palate.

Surgery to repair the cleft lip usually is done in the first few months of life, although more surgeries are sometimes needed later in life.<sup>(8)</sup>

### Prevalence:

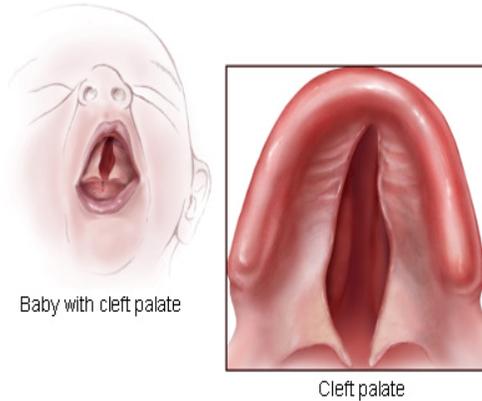
North Dakota has a cleft lip with and without cleft palate rate of 10.01 per every 10,000 live births. The five-year rate of cleft lip with and without cleft palate in North Dakota is statistically comparable to Alaska, Colorado and Utah.

**Figure 15. North Dakota's Cleft Lip with and without Cleft Palate Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Orofacial Defects

## ➤ Cleft Palate Without Cleft Lip



### Definition:

A cleft palate is an opening in the roof of the mouth, called the palate. The palate is formed early in pregnancy (at about six to nine weeks of pregnancy). A cleft palate can occur when the two sides of the palate do not come together correctly. In some children, both the hard (front) and soft (back) parts of the palate are open. In other children, only part of the palate is open.

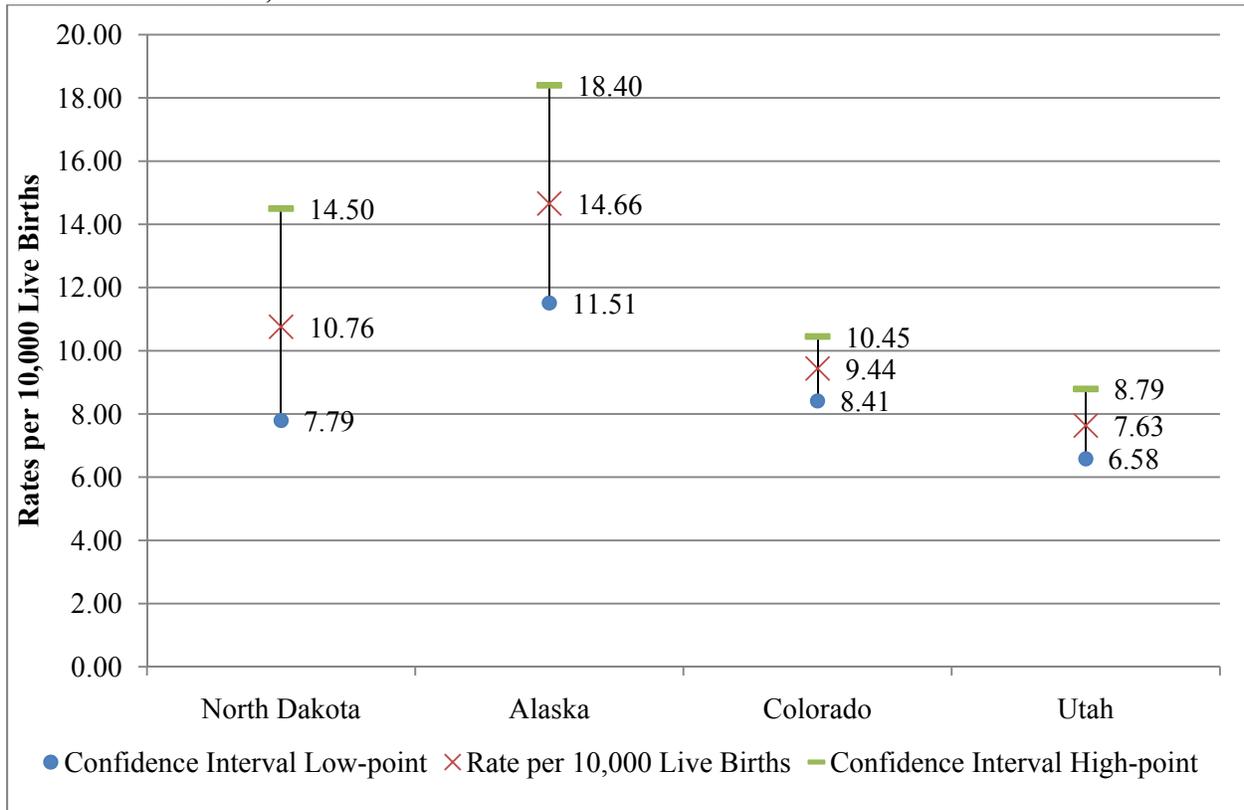
Surgery to repair a cleft palate usually is done in the first year of life, although more surgeries are sometimes needed later in life.<sup>(8)</sup>

(D)

### Prevalence:

North Dakota has a cleft palate without cleft lip rate of 10.76 per every 10,000 live births. The five-year rate of cleft palate without cleft lip in North Dakota is statistically comparable to Alaska, Colorado and Utah.

**Figure 16. North Dakota's Cleft Palate without Cleft Lip Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Gastrointestinal Defects

---

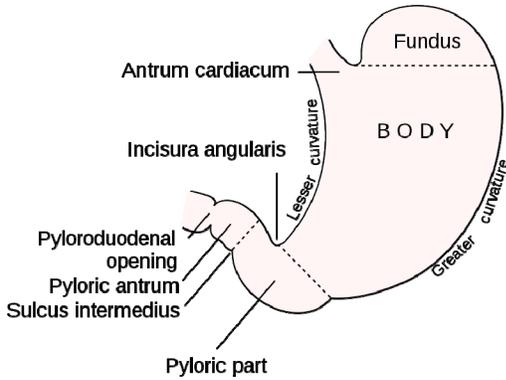
Gastrointestinal defects occur along the gastrointestinal tract (the mouth to the anus) and include defects of the stomach and intestines.<sup>(5)</sup>

Gastrointestinal defects accounted for 10.4 percent of all birth defects in North Dakota from 2001 through 2005. A profile for the two gastrointestinal defects with 10 or more incidents is provided in this section.

<b>Gastrointestinal Defects</b> .....	<b>47-49</b>
<i>Pyloric Stenosis</i> .....	48
<i>Rectal and Large Intestinal Atresia/Stenosis</i> .....	49

# Gastrointestinal Defects

## ➤ Pyloric Stenosis



### Definition:

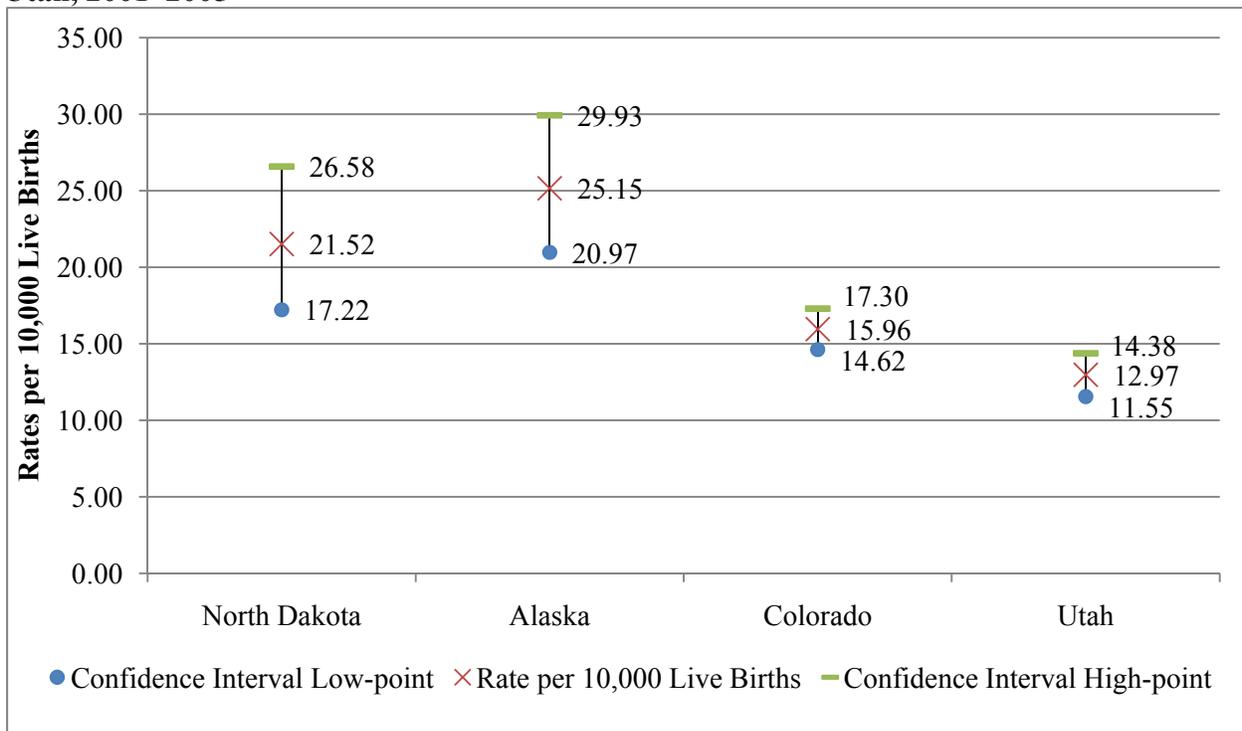
Pyloric stenosis is a condition where a thickening of muscles narrows the opening of the stomach into the small intestine. Hindering or preventing the stomach from emptying, pyloric stenosis causes external symptoms such as vomiting, diarrhea, dehydration and failure to gain weight. One of the most common causes of gastrointestinal obstruction in infants, it is usually not diagnosed at birth, but within the first three to 12 weeks of life. The condition must be repaired surgically.<sup>(20)</sup>

(M)

### Prevalence:

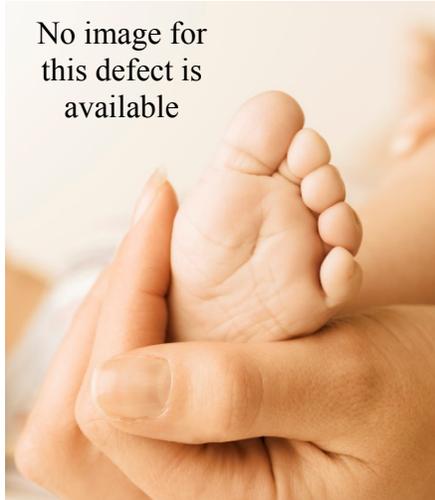
North Dakota has a pyloric stenosis rate of 21.52 per every 10,000 live births. The five-year rate of pyloric stenosis in North Dakota is statistically comparable to Alaska and Colorado. North Dakota's rate is slightly higher than Utah.

**Figure 17. North Dakota's Pyloric Stenosis Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Gastrointestinal Defects

## ➤ Rectal and Large Intestinal Atresia/Stenosis



No image for this defect is available

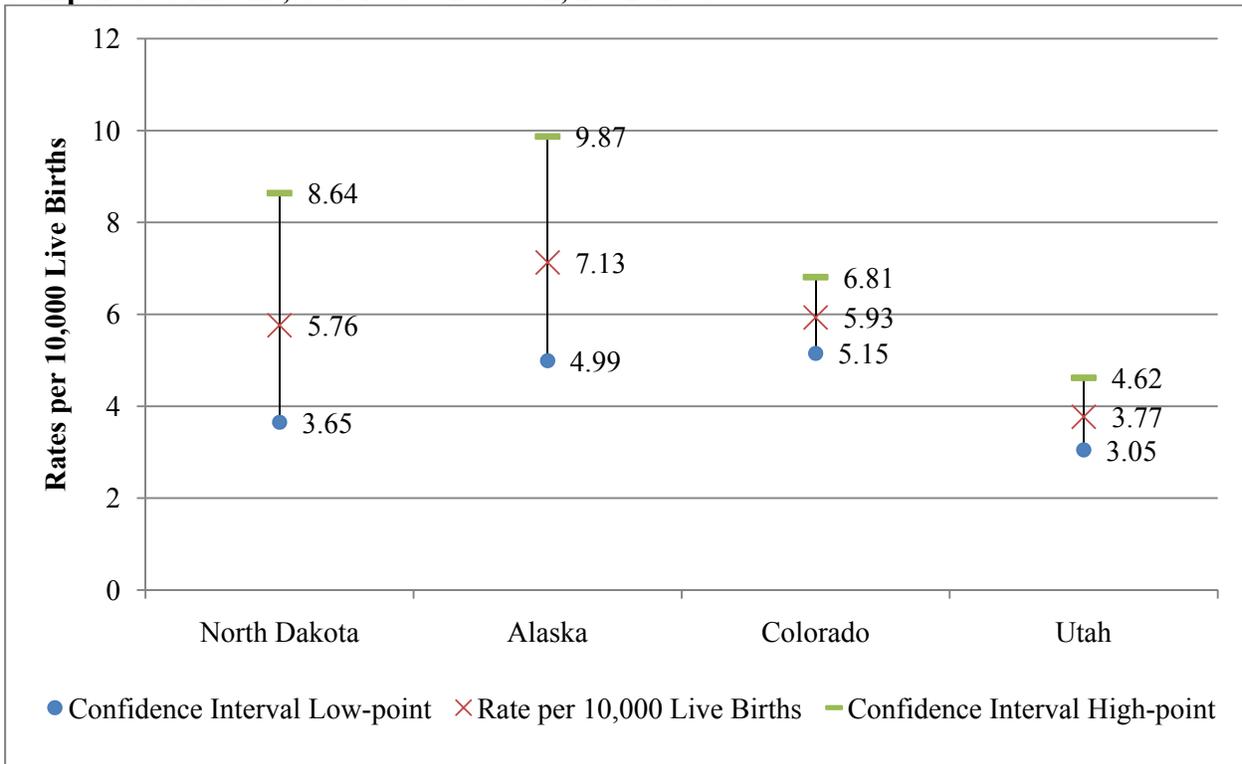
**Definition:**

Rectal and large intestinal atresia/stenosis is a congenital absence, closure or constriction of the large intestine, rectum or anus.<sup>(22)</sup>

**Prevalence:**

North Dakota has a rectal and large intestinal atresia/stenosis rate of 5.76 per every 10,000 live births. The five-year rate of rectal and large intestinal atresia/stenosis in North Dakota is statistically comparable to Alaska, Colorado and Utah.

**Figure 18. North Dakota’s Rectal and Large Intestinal Atresia/Stenosis Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Genitourinary Defects

---

Genitourinary anomalies are congenital malformations of the urinary tract and reproductive system. As a group, these anomalies are relatively common and include both rare, life-threatening anomalies and less severe but more common anomalies that may be corrected surgically.<sup>(22)</sup>

Genitourinary defects accounted for 6.1 percent of all birth defects in North Dakota from 2001 through 2005. A profile for the two genitourinary defects with 10 or more incidents is provided in this section.

<b>Genitourinary Defects</b> .....	<b>50-52</b>
<i>Hypospadias and Epispadias</i> .....	<i>51</i>
<i>Renal Agenesis/Hypoplasia</i> .....	<i>52</i>

# Genitourinary Defects

## ➤ *Hypospadias and Epispadias*



(l)

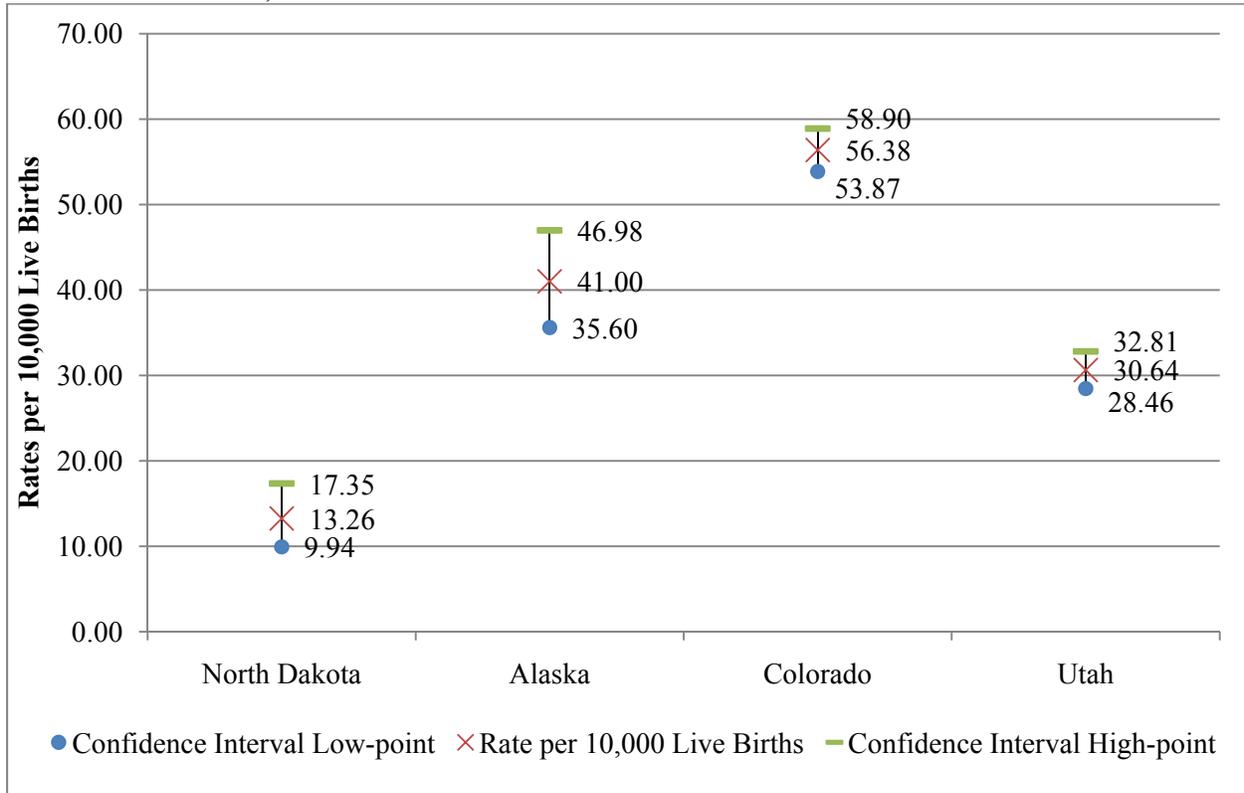
### Definition:

Hypospadias is a birth defect among boys in which the opening of the urethra is located somewhere along the underside of the penis instead of at the tip. Epispadias is when the urethra ends in an opening on the upper aspect (the dorsum) of the penis. Females can also have this type of congenital malformation. Epispadias of the female may occur when the urethra develops too far anteriorly, exiting in the clitoris or even more forward. The urethra is the tube that carries urine from the bladder to the outside of the body. This defect occurs when the urethra does not complete its development during the pregnancy.<sup>(16, 11)</sup>

### Prevalence:

North Dakota has a hypospadias and epispadias rate of 13.26 per every 10,000 live births. The five-year rate of hypospadias and epispadias in North Dakota is not statistically comparable to Alaska, Colorado and Utah. North Dakota's rate is much lower than Alaska and Colorado and lower than Utah.

**Figure 19. North Dakota's Hypospadias and Epispadias Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Genitourinary Defects

## ➤ Renal Agenesis/Hypoplasia



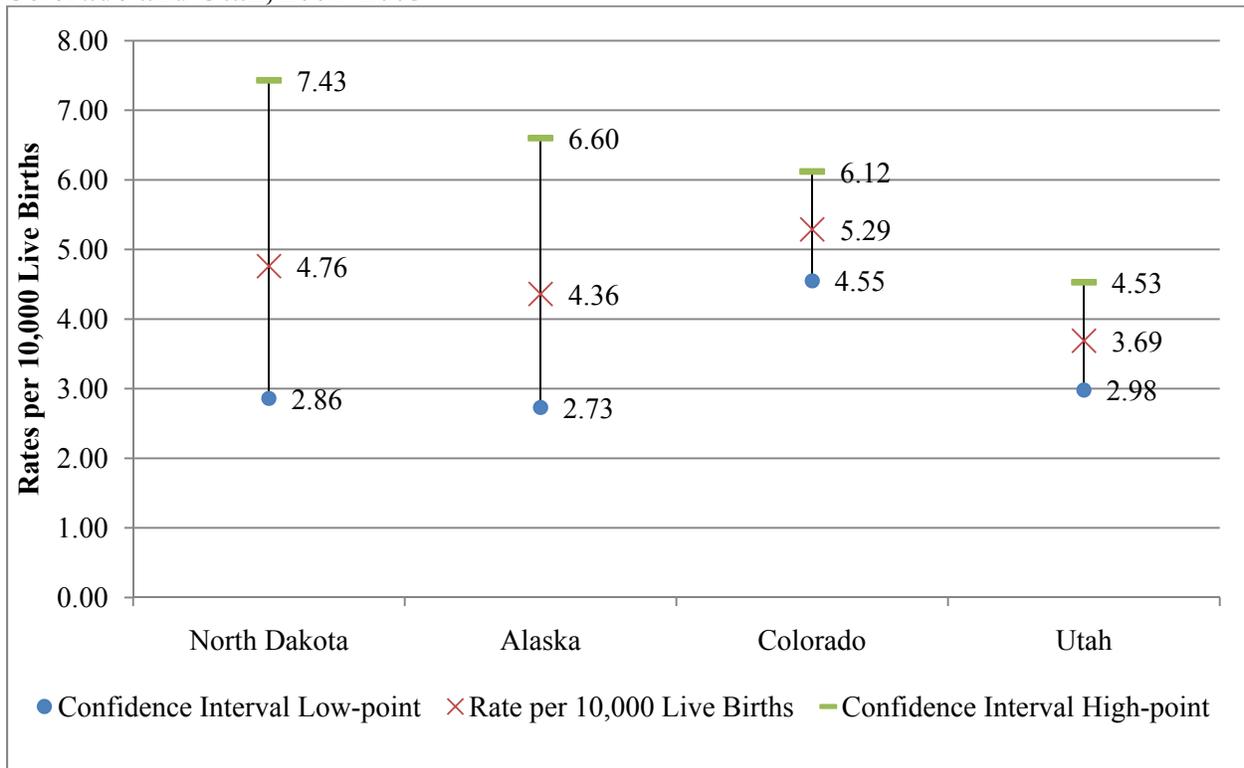
### Definition:

Renal agenesis is the name given to a congenital absence of one or both kidneys. The kidneys develop between the fifth and 12th week of fetal life, and by the 13th week they are normally producing urine. When the embryonic kidney cells fail to develop, the result is called renal agenesis. It is often detected on fetal ultrasound because there will be a lack of amniotic fluid (called oligohydramnios). When bilateral (both kidneys are absent), this condition is not compatible with survival; in fact, 40 percent of babies with bilateral renal agenesis will be stillborn, and if born alive, the baby will live only a few hours.<sup>(21)</sup>

### Prevalence:

North Dakota has a renal agenesis/hypoplasia rate of 4.76 per every 10,000 live births. The five-year rate of renal agenesis/hypoplasia in North Dakota is statistically comparable to Alaska, Colorado and Utah.

**Figure 20. North Dakota's Renal Agenesis/Hypoplasia Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Musculoskeletal Defects

---

Musculoskeletal anomalies include diverse congenital anomalies of the limbs, abdominal wall and diaphragm. Major skeletal anomalies occur when one or more parts of a limb are missing or abbreviated (reduction deformities of the arms and legs) or when the hip joint capsule is so relaxed that it dislocates at birth (congenital hip dislocation). Abdominal wall anomalies are formed early in gestation when the wall fails to close properly, causing part of the gut to protrude outside the abdomen (gastroschisis or omphalocele). A diaphragmatic hernia occurs when there is an incomplete separation of the thorax (containing the heart and lungs) from the abdomen (containing the gastrointestinal organs). Most musculoskeletal defects are repaired surgically.<sup>(22)</sup>

Musculoskeletal defects accounted for 3.6 percent of all birth defects in North Dakota from 2001 through 2005. A profile for the two musculoskeletal defects with 10 or more incidents is provided in this section.

<b>Musculoskeletal Defects</b> .....	<b>53-55</b>
<i>Congenital Hip Dislocation</i> .....	54
<i>Diaphragmatic Hernia</i> .....	55

# Musculoskeletal Defects

## ➤ Congenital Hip Dislocation

No image for this defect is available



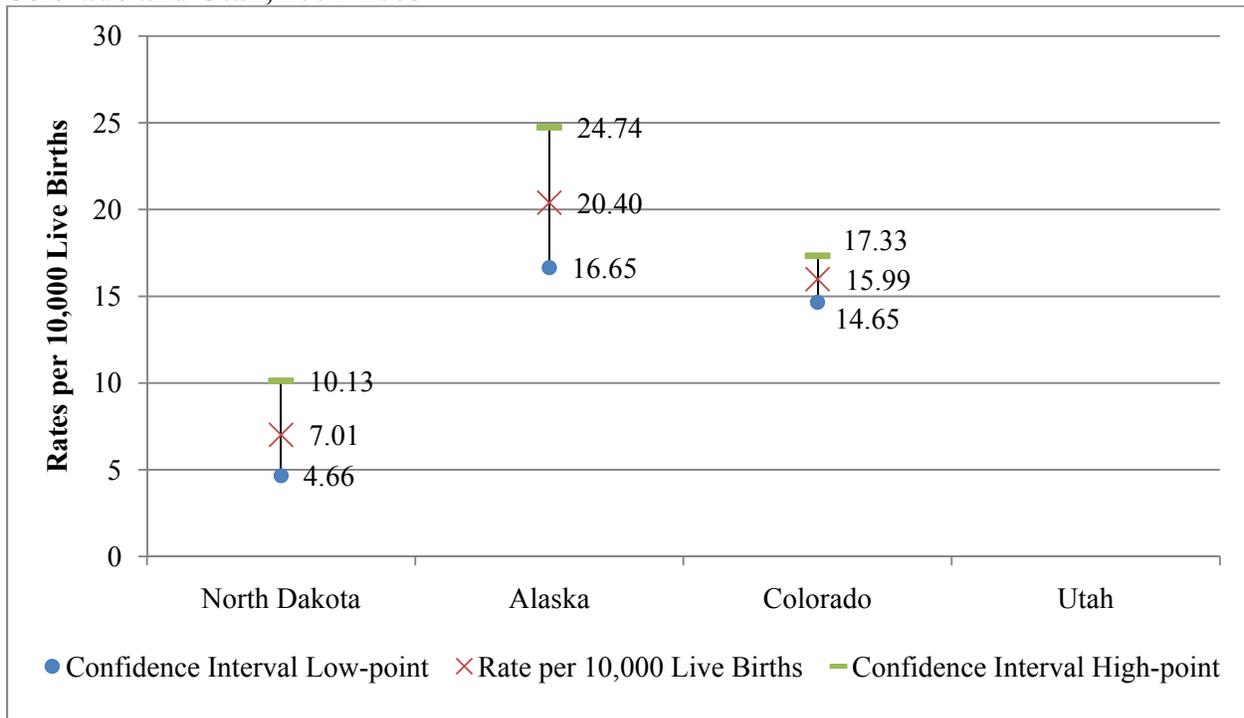
### Definition:

This congenital defect dislocation refers to a condition in which one or both of the hips are dislocated at birth. This often occurs due to improper formation of components of the joint or due to loose ligaments and muscles involved in hip movement. The actual dislocation usually occurs postpartum. If congenital hip dislocation is diagnosed in infancy, treatment with bracing is usually successful. As age at diagnosis increases, the condition becomes harder to treat with braces and may necessitate surgery and body casting.<sup>(22)</sup>

### Prevalence:

North Dakota has a congenital hip dislocation rate of 7.01 per every 10,000 live births. The five-year rate of congenital hip dislocation in North Dakota is not statistically comparable to Alaska or Colorado. North Dakota's rate is lower than Alaska and Colorado.

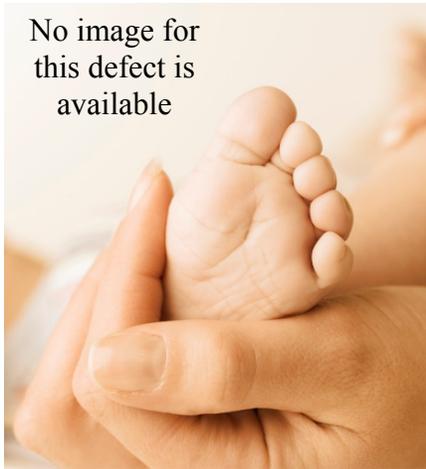
**Figure 21. North Dakota's Congenital Hip Dislocation Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



Note: No data were provided for Utah.

# Musculoskeletal Defects

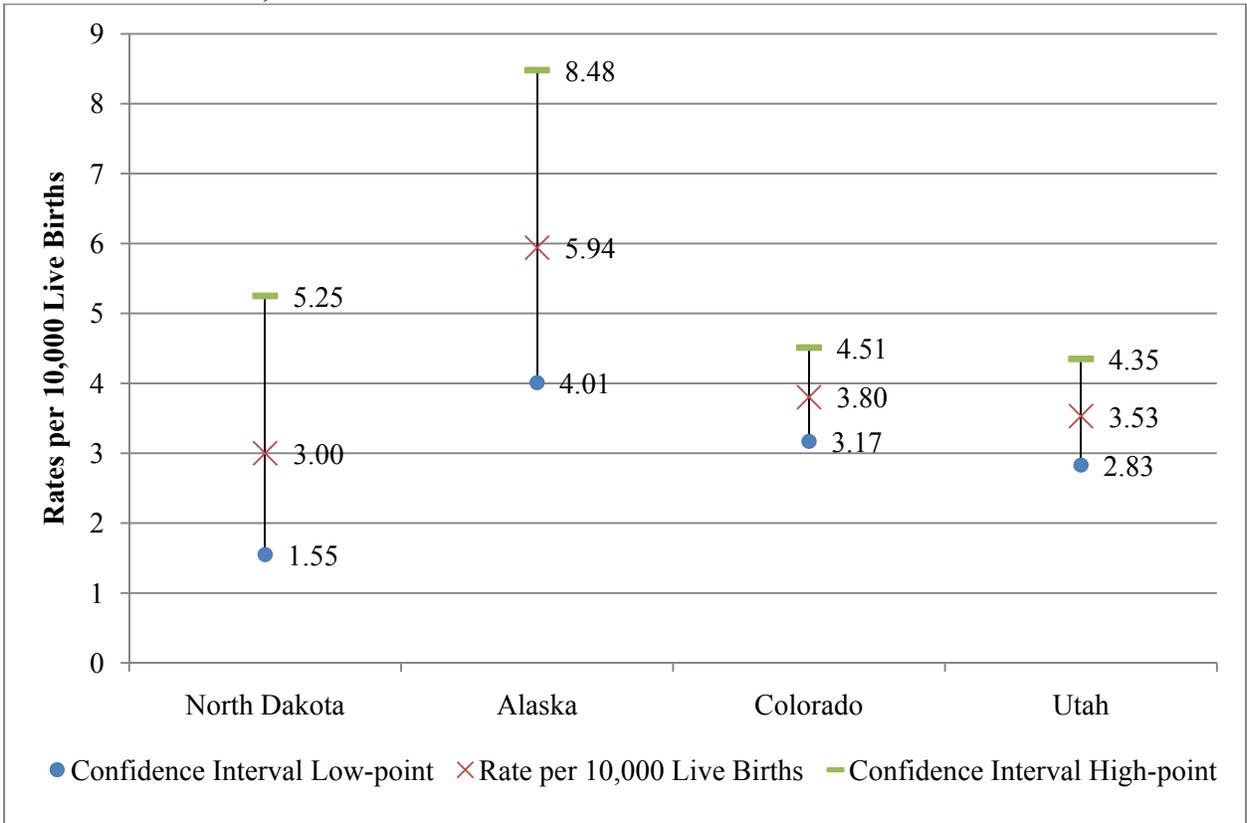
## ➤ *Diaphragmatic Hernia*



**Definition:**  
 A diaphragmatic hernia is a congenital defect of the muscular diaphragm resulting in herniation of the abdominal contents into the chest.<sup>(22)</sup>

**Prevalence:**  
 North Dakota has a diaphragmatic hernia rate of 3.00 per every 10,000 live births. The five-year rate of diaphragmatic hernia in North Dakota is statistically comparable to Alaska, Colorado and Utah.

**Figure 22. North Dakota’s Diaphragmatic Hernia Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Chromosomal Defects

---

Birth defects categorized as chromosomal anomalies refer to those that are caused by abnormal numbers of chromosomes or deletions or damage to the structure of the chromosome. Chromosomal anomalies usually occur when the sperm and egg are developing, before the egg is fertilized. A trisomy is a common type of chromosomal anomaly. Humans have 22 matched pairs of autosomal chromosomes plus the pair that determines sex. Trisomy occurs when an infant has an extra copy of a chromosome, forming a triad instead of a pair. A characteristic syndrome results, depending on which chromosome pair was affected.<sup>(22)</sup>

Chromosomal defects accounted for 4.8 percent of all birth defects in North Dakota from 2001 through 2005. A profile for the chromosomal defect with 10 or more incidents is provided in this section.

<b>Chromosomal Defects</b> .....	<b>56–57</b>
<i>Down Syndrome (Trisomy 21)</i> .....	<i>57</i>

# Chromosomal Defects

## ➤ Down Syndrome (Trisomy 21)



(J)

### Definition:

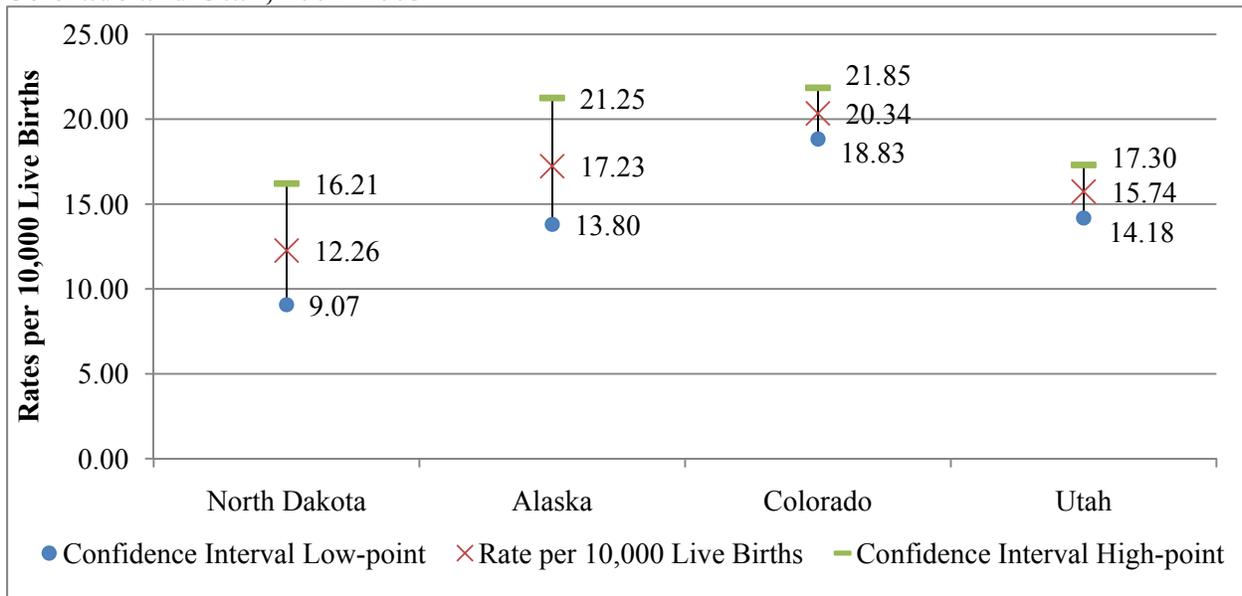
Down syndrome is a condition in which a baby is born with an extra chromosome. Chromosomes are small “packages” of genes in the body. They determine how a baby’s body forms during pregnancy and how, as the baby grows in the womb and after birth, the baby’s body functions. Normally, a baby is born with 46 chromosomes. Babies born with Down syndrome have an extra copy of one of these chromosomes. This extra copy changes the body’s and brain’s normal development and causes mental and physical problems for the baby.

Even though people with Down syndrome might have some physical and mental features in common, symptoms of Down syndrome can range from mild to severe. Usually, mental development and physical development are slower in people with Down syndrome than in those without it.<sup>(9)</sup>

### Prevalence:

North Dakota has a Down syndrome (Trisomy 21) rate of 12.26 per every 10,000 live births. The five-year rate of Down syndrome (Trisomy 21) in North Dakota is statistically comparable to Alaska and Utah. North Dakota’s rate is lower than Colorado.

**Figure 24. North Dakota’s Down Syndrome (Trisomy 21) Birth Rate Compared to Alaska, Colorado and Utah, 2001–2005**



# Other Defects

---

Other birth defects in North Dakota with 10 or more cases in 2001 through 2005 include fetal alcohol spectrum disorders (fetus or newborn affected by maternal alcohol use).

Other defects (i.e., fetal alcohol spectrum disorder and amniotic bands) accounted for 1.7 percent of all birth defects in North Dakota from 2001 through 2005.

<b>Other Defects</b> .....	<b>58–59</b>
<i>Fetal Alcohol Spectrum Disorder</i> .....	<i>59</i>

## Other Defects

### ➤ Fetal Alcohol Spectrum Disorder



(H)

#### Definition:

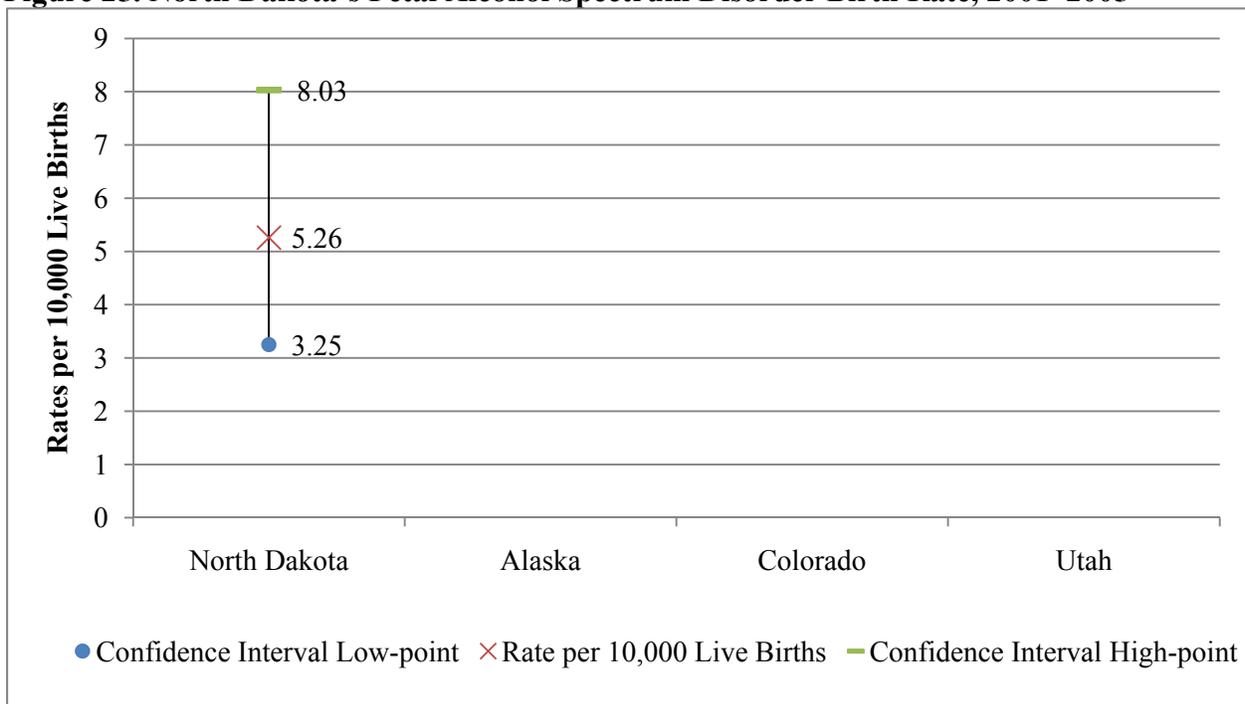
Fetal alcohol spectrum disorder (FASD) describes a continuum of permanent birth defects caused by maternal consumption of alcohol during pregnancy which includes, but is not limited to, fetal alcohol syndrome (FAS).

Over time, as it became apparent through research and clinical experience that a range of effects (including physical, behavioral and cognitive) could arise from prenatal alcohol exposure, the term FASD was developed to include FAS as well as other conditions resulting from prenatal alcohol exposure. There are a number of other subtypes with evolving nomenclature and definitions based on partial expressions of FAS, including partial fetal alcohol syndrome (PFAS), alcohol-related neurodevelopmental disorder (ARND), alcohol-related birth defects (ARBD), and fetal alcohol effect (FAE).<sup>(12)</sup>

#### Prevalence:

North Dakota has a fetal alcohol spectrum disorder rate of 5.26 per every 10,000 live births.

Figure 25. North Dakota's Fetal Alcohol Spectrum Disorder Birth Rate, 2001–2005



Note: Data are for the category of “fetus or newborn affected by maternal alcohol use.” No data were provided for Alaska, Colorado or Utah.

# Additional Birth Defects Monitored by NDBDMS

Among the 43 birth defects monitored by the North Dakota Birth Defect Monitoring System (NDBDMS), 19 birth defects had fewer than 10 cases in North Dakota in 2001 through 2005. The prevalence and 95 percent confidence intervals for these 19 birth defects are shown in Table 17.

Definitions of these birth defects can be found in the “Glossary of Congenital Anomalies” in the Appendices.

**Table 17. Prevalence of Birth Defects Monitored in North Dakota With Fewer Than 10 Cases, 2001–2005**

Birth Defect	Prevalence		
	Rate Reported Per 10,000	Confidence Interval, Lower 95%	Confidence Interval, Upper 95%
Amniotic bands	0.25	0.01	1.81
Aniridia	0.25	0.01	1.81
Anophthalmia/microphthalmia	1.00	0.27	2.56
Anotia/microtia	0.50	0.06	1.81
Biliary atresia	1.00	0.27	2.56
Bladder exstrophy	0.25	0.01	1.81
Choanal atresia	0.00	0.00	0.92
Common truncus	0.50	0.06	1.81
Congenital cataract	1.50	0.55	3.27
Ebstein’s anomaly	0.75	0.15	2.19
Encephalocele	0.25	0.01	1.81
Esophageal atresia/tracheoesophageal fistula	2.25	1.03	4.28
Hirschsprung’s disease (congenital megacolon)	1.75	0.70	3.61
Obstructive genitourinary defect	0.50	0.06	1.81
Reduction deformity, lower limbs	0.50	0.06	1.81
Reduction deformity, upper limbs	0.75	0.15	2.19
Tricuspid valve atresia and stenosis	1.25	0.41	2.92
Trisomy 13 (Patau syndrome)	1.25	0.41	2.92
Trisomy 18 (Edwards syndrome)	1.25	0.41	2.92

Note: Appendix Tables 1, 2 and 3 list actual numbers of cases as well as prevalence and confidence intervals. However, due to the small numbers, we encourage caution when drawing conclusions from these numbers.

# Appendices

# North Dakota Birth Defects State Profile<sup>(6)</sup>

Birth defects state profile – North Dakota		January 2009
<p><b>About 1 out of every 33 babies is born with a major birth defect.</b></p>	<p><b>Birth defects cause one in five deaths among infants less than a year old.</b></p>	<p><b>Birth defects lead to \$2.5 billion per year in hospital costs alone in the U.S.</b></p>

Defects	North Dakota <sup>†</sup>		US <sup>‡</sup>	
	Annual no. of cases	Birth prevalence*	Annual no. of cases	Birth prevalence*
<b>Central nervous system</b>				
Anencephalus	2	2.50	1,009	2.51
Spina bifida without anencephalus	5	5.76	1,477	3.68
<b>Cardiovascular</b>				
Transposition of great arteries	4	4.76	1,901	4.73
Tetralogy of Fallot	4	4.51	1,574	3.92
Atrioventricular septal defect (also known as endocardial cushion defect)	2	3.00	1,748	4.36
Hypoplastic left heart syndrome	3	3.50	975	2.43
<b>Orofacial</b>				
Cleft lip with and without cleft palate	8	10.01	4,209	10.47
Cleft palate without cleft lip	9	10.76	2,567	6.39
<b>Musculoskeletal</b>				
Upper limb defect	<1	0.75	1,521	3.79
Lower limb defect	<1	0.50	763	1.90
Gastroschisis	--	--	1,497	3.73
<b>Chromosomal</b>				
Down syndrome	6	8.01	5,132	12.78

**Selected birth defects counts and birth prevalence, North Dakota and US**

\* per 10,000 live births

<sup>†</sup> estimates based on pooled data from birth years 2001-2005

<sup>‡</sup> estimates based on pooled data from birth years 1999-2001

-- No data available

*Note: Due to variability in the methods used by state birth defects surveillance systems and differences in populations and risk factors, state prevalence estimates may not be directly comparable with national estimates or those of other states.*

**Preventing birth defects**

- The causes of about 70% of birth defects are unknown.
- Many birth defects happen during early pregnancy, often before a woman knows she is pregnant.
- Addressing health risks and behaviors before pregnancy can reduce the risk of poor birth outcomes, including some birth defects.
- All women who could become pregnant should take 400 micrograms of folic acid every day to help prevent serious defects of the baby's brain and spinal cord.

**North Dakota's Birth Defect Surveillance System**

The North Dakota Birth Defects Monitoring System was established in 2003 as a means of identifying and collecting information about babies born with certain birth defects in North Dakota. The North Dakota Birth Defects Monitoring program is a passive surveillance system. Data are collected and linked from three secondary data sources. Because of the low numbers of both resident births and occurrences of individual birth defects in North Dakota, rates for each birth defect are averaged over five consecutive years to improve statistical stability of the data.

**Program information:**

Devaiah Muthappa Muccatira, MS  
 North Dakota Birth Defects Monitoring System  
 E-mail: [dmuccatira@nd.gov](mailto:dmuccatira@nd.gov)

Tamara Lynn Gallup-Millner, RN, MPA  
 North Dakota Birth Defects Monitoring System  
 E-mail: [tgallupmillner@nd.gov](mailto:tgallupmillner@nd.gov)

Website: <http://www.health.state.nd.us/cshs/docs/birth-defects-report.pdf>

**How birth defects data are used in North Dakota**

Data from the North Dakota Birth Defects Monitoring System is used to: 1) report incidence and prevalence of birth defects, 2) increase awareness of birth defects and identified risk factors, 3) help researchers and health-care providers learn more about preventing future problems, and 4) assure that children born with birth defects have access to needed health-care and other services.

## Appendix Tables

**Appendix Table 1. Incidence and Prevalence of Reported Cases of North Dakota Birth Defects by Race/Ethnicity, 2001–2005**

Birth Defect	Number and <i>Prevalence</i> ** of Birth Defects						
	All births	Race/Ethnicity					
		Non-Hispanic white	Non-Hispanic black	Hispanic	Asian or Pacific Islander	American Indian and Alaskan Native	Unknown or other
*Amniotic bands	1 0.25	1 0.30	0 0.00	0 0.00	0 0.00	0 0.00	0 0.00
Anencephalus	10 2.50	9 2.72	0 0.00	0 0.00	0 0.00	1 2.02	0 0.00
*Aniridia	1 0.25	1 0.30	0 0.00	0 0.00	0 0.00	0 0.00	0 0.00
*Anophthalmia/ microphthalmia	4 1.00	2 0.60	0 0.00	0 0.00	0 0.00	2 4.04	0 0.00
*Anotia/microtia	2 0.50	2 0.60	0 0.00	0 0.00	0 0.00	0 0.00	0 0.00
Aortic valve stenosis	11 2.75	9 2.72	2 38.46	0 0.00	0 0.00	0 0.00	0 0.00
Atrial septal defect	250 62.57	170 51.29	30 576.92	0 0.00	0 0.00	46 92.93	4 0.00
*Biliary atresia	4 1.00	3 0.91	0 0.00	0 0.00	0 0.00	1 2.02	0 0.00
*Bladder exstrophy	1 0.25	0 0.00	0 0.00	0 0.00	0 0.00	1 2.02	0 0.00
*Choanal atresia	0 0.00	0 0.00	0 0.00	0 0.00	0 0.00	0 0.00	0 0.00
Cleft lip with and without cleft palate	40 10.01	25 7.54	4 76.92	0 0.00	0 0.00	10 20.20	1 0.00
Cleft palate without cleft lip	43 10.76	36 10.86	2 38.46	0 0.00	0 0.00	5 10.10	0 0.00
Coarctation of aorta	22 5.51	15 4.53	3 57.69	0 0.00	0 0.00	4 8.08	0 0.00
*Common truncus	2 0.50	1 0.30	0 0.00	0 0.00	0 0.00	1 2.02	0 0.00
*Congenital cataract	6 1.50	4 1.21	1 19.23	0 0.00	0 0.00	1 2.02	0 0.00
Congenital hip dislocation	28 7.01	21 6.34	2 38.46	0 0.00	0 0.00	5 10.10	0 0.00
Diaphragmatic hernia	12 3.00	9 2.72	0 0.00	0 0.00	0 0.00	3 6.06	0 0.00
Down syndrome (Trisomy 21)	49 12.26	38 11.47	2 38.46	0 0.00	0 0.00	9 18.18	0 0.00
*Ebstein's anomaly	3 0.75	2 0.60	1 19.23	0 0.00	0 0.00	0 0.00	0 0.00

Birth Defect	Number and Prevalence** of Birth Defects						
	All births	Race/Ethnicity					
		Non-Hispanic white	Non-Hispanic black	Hispanic	Asian or Pacific Islander	American Indian and Alaskan Native	Unknown or other
*Encephalocele	1	1	0	0	0	0	0
	0.25	0.30	0.00	0.00	0.00	0.00	
Endocardial cushion defect	12	9	1	0	0	2	0
	3.00	2.72	19.23	0.00	0.00	4.04	
*Esophageal atresia/ tracheoesophageal fistula	9	6	1	0	0	2	0
	2.25	1.81	19.23	0.00	0.00	4.04	
Fetus or newborn affected by maternal alcohol use	21	12	0	0	0	9	0
	5.26	3.62	0.00	0.00	0.00	18.18	
*Hirschsprung's disease (congenital megacolon)	7	5	1	0	0	1	0
	1.75	1.51	19.23	0.00	0.00	2.02	
Hydrocephalus without spina bifida	31	24	1	0	0	6	0
	7.76	7.24	19.23	0.00	0.00	12.12	
Hypoplastic left heart syndrome	14	13	0	0	0	1	0
	3.50	3.92	0.00	0.00	0.00	2.02	
Hypospadias and epispadias	53	42	3	0	0	8	0
	13.26	12.67	57.69	0.00	0.00	16.16	
Microcephalus	34	25	1	0	0	8	0
	8.51	7.54	19.23	0.00	0.00	16.16	
*Obstructive genitourinary defect	2	2	0	0	0	0	0
	0.50	0.60	0.00	0.00	0.00	0.00	
Patent ductus arteriosus	129	77	18	0	0	32	2
	32.29	23.23	346.15	0.00	0.00	64.65	
Pulmonary valve atresia and stenosis	52	44	5	0	0	0	3
	13.01	13.28	96.15	0.00	0.00	0.00	
Pyloric stenosis	86	54	6	0	0	26	0
	21.52	16.29	115.38	0.00	0.00	52.53	
Rectal and large intestinal atresia/stenosis	23	10	3	0	0	10	0
	5.76	3.02	57.69	0.00	0.00	20.20	
*Reduction deformity, lower limbs	2	0	1	0	0	1	0
	0.50	0.00	19.23	0.00	0.00	2.02	
*Reduction deformity, upper limbs	3	1	1	0	0	1	0
	0.75	0.30	19.23	0.00	0.00	2.02	
Renal agenesis/hypoplasia	19	18	0	0	0	1	0
	4.76	5.43	0.00	0.00	0.00	2.02	

Birth Defect	Number and Prevalence** of Birth Defects						
	All births	Race/Ethnicity					
		Non-Hispanic white	Non-Hispanic black	Hispanic	Asian or Pacific Islander	American Indian and Alaskan Native	Unknown or other
Spina bifida without anencephalus	23	19	0	0	0	4	0
	5.76	5.73	0.00	0.00	0.00	8.08	
Tetralogy of Fallot	18	15	1	0	0	2	0
	4.51	4.53	19.23	0.00	0.00	4.04	
Transposition of great arteries	19	14	0	0	0	5	0
	4.76	4.22	0.00	0.00	0.00	10.10	
*Tricuspid valve atresia and stenosis	5	4	0	0	0	1	0
	1.25	1.21	0.00	0.00	0.00	2.02	
*Trisomy 13 (Patau syndrome)	5	4	0	0	0	1	0
	1.25	1.21	0.00	0.00	0.00	2.02	
*Trisomy 18 (Edwards syndrome)	5	4	0	0	0	1	0
	1.25	1.21	0.00	0.00	0.00	2.02	
Ventricular septal defect	178	133	15	0	0	27	3
	44.55	40.13	288.46	0.00	0.00	54.55	
<b>Total Live Births</b>	<b>39,955</b>	<b>33,143</b>	<b>520</b>	<b>796</b>	<b>501</b>	<b>4,950</b>	<b>45</b>

\*Indicates incidence of fewer than 10 anomalies in 2001–2005.

\*\*Prevalence is rate per 10,000 live births.

**Appendix Table 2. Incidence of Birth Defects in North Dakota, Alaska, Colorado and Utah, 2001–2005**

Birth Defect	Number of Birth Defects			
	North Dakota	Alaska	Colorado	Utah
*Amniotic bands	1	NA	NA	35
Anencephalus	10	0	45	58
*Aniridia	1	<6	5	4
*Anophthalmia/microphthalmia	4	24	57	39
*Anotia/microtia	2	15	77	74
Aortic valve stenosis	11	12	137	74
Atrial septal defect	250	556	2,612	587
*Biliary atresia	4	9	33	16
*Bladder exstrophy	1	<6	10	4
*Choanal atresia	0	20	57	37
Cleft lip with and without cleft palate	40	81	414	331
Cleft palate without cleft lip	43	74	323	190
Coarctation of aorta	22	37	307	201
*Common truncus	2	10	33	24
*Congenital cataract	6	22	101	63

Birth Defect	Number of Birth Defects			
	North Dakota	Alaska	Colorado	Utah
Congenital hip dislocation	28	103	547	NA
Diaphragmatic hernia	12	30	130	88
Down syndrome (Trisomy 21)	49	87	696	392
*Ebstein's anomaly	3	<6	30	14
*Encephalocele	1	7	37	23
Endocardial cushion defect	12	27	147	137
*Esophageal atresia/tracheoesophageal fistula	9	12	173	64
Fetus or newborn affected by maternal alcohol use	21	NA	NA	NA
*Hirschsprung's disease (congenital megacolon)	7	35	75	43
Hydrocephalus without spina bifida	31	64	288	126
Hypoplastic left heart syndrome	14	17	115	91
Hypospadias and Epispadias	53	207	1,929	763
Microcephalus	34	93	209	122
*Obstructive genitourinary defect	2	192	1,129	486
Patent ductus arteriosus	129	394	1,303	NA
Pulmonary valve atresia and stenosis	52	77	377	294
Pyloric stenosis	86	127	546	323
Rectal and large intestinal atresia/stenosis	23	36	203	94
*Reduction deformity, lower limbs	2	21	67	44
*Reduction deformity, upper limbs	3	8	130	158
Renal agenesis/hypoplasia	19	22	181	92
Spina bifida without anencephalus	23	20	117	105
Tetralogy of Fallot	18	36	124	90
Transposition of great arteries	19	26	135	111
*Tricuspid valve atresia and stenosis	5	<6	56	35
*Trisomy 13 (Patau syndrome)	5	7	89	42
*Trisomy 18 (Edwards syndrome)	5	12	159	81
Ventricular septal defect	178	459	1,476	212
Total births	39,955	50,493	342,127	249,059

\*Indicates incidence of fewer than 10 anomalies in 2001–2005.

Note: NA = Data not available.

**Appendix Table 3. Prevalence of Birth Defects in North Dakota, Alaska, Colorado and Utah with Confidence Intervals, 2001–2005**

State	Prevalence**	95% Confidence Intervals of Prevalence	
		Lower	Upper
<b>*Amniotic bands</b>			
North Dakota	0.25	0.01	1.81
Alaska	NA	NA	NA
Colorado	NA	NA	NA
Utah	1.41	0.98	1.95
<b>Anencephalus</b>			
North Dakota	2.50	1.20	4.60
Alaska	NA	NA	NA
Colorado	1.32	0.96	1.76
Utah	2.33	1.77	3.01
<b>*Aniridia</b>			
North Dakota	0.25	0.01	1.81
Alaska	NA	NA	NA
Colorado	0.15	0.05	0.34
Utah	0.16	0.04	0.41
<b>*Anophthalmia/microphthalmia</b>			
North Dakota	1.00	0.27	2.56
Alaska	4.75	3.05	7.07
Colorado	1.67	1.26	2.16
Utah	1.57	1.11	2.14
<b>*Anotia/microtia</b>			
North Dakota	0.50	0.06	1.81
Alaska	2.97	1.66	4.90
Colorado	2.25	1.78	2.81
Utah	2.97	2.33	3.73
<b>Aortic valve stenosis</b>			
North Dakota	2.75	1.37	4.93
Alaska	2.38	1.23	4.15
Colorado	4.00	3.36	4.77
Utah	2.97	2.33	3.73
<b>Atrial septal defect</b>			
North Dakota	62.57	55.05	70.83
Alaska	110.11	100.88	119.35
Colorado	76.35	73.42	79.27
Utah	23.57	21.66	25.47
<b>*Biliary atresia</b>			
North Dakota	1.00	0.27	2.56
Alaska	1.78	0.82	3.38
Colorado	0.96	0.66	1.35
Utah	0.64	0.37	1.04
<b>*Bladder exstrophy</b>			
North Dakota	0.25	0.01	1.81
Alaska	NA	NA	NA

State	Prevalence**	95% Confidence Intervals of Prevalence	
		Lower	Upper
Colorado	0.29	0.14	0.54
Utah	0.16	0.04	0.41
<b>*Choanal atresia</b>			
North Dakota	0.00	0.00	0.92
Alaska	3.96	2.42	6.12
Colorado	1.67	1.26	2.16
Utah	1.49	1.05	2.05
<b>Cleft lip with and without cleft palate</b>			
North Dakota	10.01	7.15	13.63
Alaska	16.04	12.74	19.94
Colorado	12.10	10.93	13.27
Utah	13.29	11.86	14.70
<b>Cleft palate without cleft lip</b>			
North Dakota	10.76	7.79	14.50
Alaska	14.66	11.51	18.40
Colorado	9.44	8.41	10.45
Utah	7.63	6.58	8.79
<b>Coarctation of aorta</b>			
North Dakota	5.51	3.45	8.34
Alaska	7.33	5.16	10.10
Colorado	8.97	7.97	9.98
Utah	8.07	6.99	9.27
<b>*Common truncus</b>			
North Dakota	0.50	0.06	1.81
Alaska	1.98	0.95	3.64
Colorado	0.96	0.66	1.35
Utah	0.96	0.62	1.43
<b>*Congenital cataract</b>			
North Dakota	1.50	0.55	3.27
Alaska	4.36	2.73	6.60
Colorado	2.95	2.40	3.59
Utah	2.53	1.94	3.24
<b>Congenital hip dislocation</b>			
North Dakota	7.01	4.66	10.13
Alaska	20.40	16.65	24.74
Colorado	15.99	14.65	17.33
Utah	NA	NA	NA
<b>Diaphragmatic hernia</b>			
North Dakota	3.00	1.55	5.25
Alaska	5.94	4.01	8.48
Colorado	3.80	3.17	4.51
Utah	3.53	2.83	4.35
<b>Down syndrome (Trisomy 21)</b>			
North Dakota	12.26	9.07	16.21
Alaska	17.23	13.80	21.25

State	Prevalence**	95% Confidence Intervals of Prevalence	
		Lower	Upper
Colorado	20.34	18.83	21.85
Utah	15.74	14.18	17.30
<b>*Ebstein's anomaly</b>			
North Dakota	0.75	0.15	2.19
Alaska	NA	NA	NA
Colorado	0.88	0.59	1.25
Utah	0.56	0.31	0.94
<b>*Encephalocele</b>			
North Dakota	0.25	0.01	1.81
Alaska	1.39	0.56	2.86
Colorado	1.08	0.76	1.49
Utah	0.92	0.59	1.39
<b>Endocardial cushion defect</b>			
North Dakota	3.00	1.55	5.25
Alaska	5.35	3.52	7.78
Colorado	4.30	3.63	5.05
Utah	5.50	4.62	6.50
<b>*Esophageal atresia/tracheoesophageal fistula</b>			
North Dakota	2.25	1.03	4.28
Alaska	2.38	1.23	4.15
Colorado	5.06	4.33	5.87
Utah	2.57	1.98	3.28
<b>Fetus or newborn affected by maternal alcohol use</b>			
North Dakota	5.26	3.25	8.03
Alaska	NA	NA	NA
Colorado	NA	NA	NA
Utah	NA	NA	NA
<b>*Hirschsprung's disease (congenital megacolon)</b>			
North Dakota	1.75	0.70	3.61
Alaska	6.93	4.83	9.64
Colorado	2.19	1.72	2.75
Utah	1.73	1.25	2.33
<b>Hydrocephalus without spina bifida</b>			
North Dakota	7.76	5.27	11.01
Alaska	12.68	9.76	16.19
Colorado	8.42	7.47	9.45
Utah	5.06	4.21	6.02
<b>Hypoplastic left heart syndrome</b>			
North Dakota	3.50	1.92	5.88
Alaska	3.37	1.96	5.39
Colorado	3.36	2.78	4.03
Utah	3.65	2.94	4.49
<b>Hypospadias and epispadias</b>			
North Dakota	13.26	9.94	17.35
Alaska	41.00	35.60	46.98

State	Prevalence**	95% Confidence Intervals of Prevalence	
		Lower	Upper
Colorado	56.38	53.87	58.90
Utah	30.64	28.46	32.81
<b>Microcephalus</b>			
North Dakota	8.51	5.89	11.89
Alaska	18.42	14.87	22.56
Colorado	6.11	5.31	7.00
Utah	4.90	4.07	5.85
<b>*Obstructive genitourinary defect</b>			
North Dakota	0.50	0.06	1.81
Alaska	38.03	32.84	43.80
Colorado	33.00	31.08	34.92
Utah	19.51	17.78	21.25
<b>Patent ductus arteriosus</b>			
North Dakota	32.29	26.96	38.36
Alaska	78.03	70.33	85.73
Colorado	38.09	36.02	40.15
Utah	NA	NA	NA
<b>Pulmonary valve atresia and stenosis</b>			
North Dakota	13.01	9.72	17.07
Alaska	15.25	12.03	19.06
Colorado	11.02	9.91	12.13
Utah	11.80	10.49	13.23
<b>Pyloric stenosis</b>			
North Dakota	21.52	17.22	26.58
Alaska	25.15	20.97	29.93
Colorado	15.96	14.62	17.30
Utah	12.97	11.55	14.38
<b>Rectal and large intestinal atresia/stenosis</b>			
North Dakota	5.76	3.65	8.64
Alaska	7.13	4.99	9.87
Colorado	5.93	5.15	6.81
Utah	3.77	3.05	4.62
<b>*Reduction deformity, lower limbs</b>			
North Dakota	0.50	0.06	1.81
Alaska	4.16	2.57	6.36
Colorado	1.96	1.52	2.49
Utah	1.77	1.28	2.37
<b>*Reduction deformity, upper limbs</b>			
North Dakota	0.75	0.15	2.19
Alaska	1.58	0.68	3.12
Colorado	3.80	3.17	4.51
Utah	6.34	5.39	7.41
<b>Renal agenesis/hypoplasia</b>			
North Dakota	4.76	2.86	7.43
Alaska	4.36	2.73	6.60

State	Prevalence**	95% Confidence Intervals of Prevalence	
		Lower	Upper
Colorado	5.29	4.55	6.12
Utah	3.69	2.98	4.53
<b>Spina bifida without anencephalus</b>			
North Dakota	5.76	3.65	8.64
Alaska	3.96	2.42	6.12
Colorado	3.42	2.83	4.10
Utah	4.22	3.45	5.10
<b>Tetralogy of Fallot</b>			
North Dakota	4.51	2.67	7.12
Alaska	7.13	4.99	9.87
Colorado	3.62	3.01	4.32
Utah	3.61	2.91	4.44
<b>Transposition of great arteries</b>			
North Dakota	4.76	2.86	7.43
Alaska	5.15	3.36	7.54
Colorado	3.95	3.31	4.67
Utah	4.46	3.67	5.37
<b>*Tricuspid valve atresia and stenosis</b>			
North Dakota	1.25	0.41	2.92
Alaska	NA	NA	NA
Colorado	1.64	1.24	2.13
Utah	1.41	0.98	1.95
<b>*Trisomy 13 (Patau syndrome)</b>			
North Dakota	1.25	0.41	2.92
Alaska	1.39	0.56	2.86
Colorado	2.60	2.09	3.20
Utah	1.69	1.22	2.28
<b>*Trisomy 18 (Edwards syndrome)</b>			
North Dakota	1.25	0.41	2.92
Alaska	2.38	1.23	4.15
Colorado	4.65	3.95	5.43
Utah	3.25	2.58	4.04
<b>Ventricular septal defect</b>			
North Dakota	44.55	38.25	51.60
Alaska	90.90	82.59	99.22
Colorado	43.14	40.94	45.14
Utah	8.51	7.40	9.74

\*Indicates incidence of fewer than 10 anomalies in 2001–2005.

\*\*Prevalence is rate per 10,000 live births.

Note: Confidence intervals are calculated using rate and prevalence. NA = Data not available.

**Appendix Table 4. Reportable Birth Defects Monitoring: ICD-10 & CDC/BPA Codes**

Birth Defect	ICD-10 Code	CDC/BPA Codes
<b>Central nervous system</b>		
Anencephalus	Q000	740.00–740.10
Spina bifida	Q050	741.00–741.99 (exclude 740.0–740.10)
Encephalocele	Q010	743.30–742.39 (exclude 741.00–741.99)
Hydrocephalus without spina bifida	Q030	742.00–742.09
Microcephalus	Q020	742.10
<b>Eye</b>		
Anophthalmia/microphthalmia	Q110	743.00–743.10
Congenital cataract	Q120	743.32–743.326
Aniridia	Q130	743.42
<b>Ear</b>		
Anotia/microtia	Q170	744.01, 744.21
<b>Cardiovascular</b>		
Common truncus	Q200	745.00–745.01
Transposition of great arteries	Q203	745.10–745.19 (exclude 745.13–745.18)
Tetralogy of Fallot	Q213	745.20–745.21, 746.84, 747.310
Ventricular septal defect	Q210	745.40–745.490 (exclude 745.498, 745.487)
Atrial septal defect	Q211	745.51–745.59 (exclude 745.50)
Endocardial cushion defect	Q212	745.60–745.69, 745.487
Pulmonary valve atresia and stenosis	Q220–Q221	746.00–746.01
Tricuspid valve atresia and stenosis	Q224	746.10, 746.106 (exclude 746.105)
Ebstein's anomaly	Q225	746.20
Aortic valve stenosis	Q230	746.30
Hypoplastic left heart syndrome	Q234	746.70
Patent ductus arteriosus	Q250	747.00
Coarctation of aorta	Q251	747.10–747.19
Pulmonary valve anomalies	Q255–Q257	
<b>Orofacial</b>		
Cleft palate	Q350	749.00–749.09
Cleft lip	Q360	749.10–749.29
Cleft lip and cleft palate	Q370	
<b>Gastrointestinal</b>		
Esophageal atresia/tracheoesophageal fistula	Q390	750.30–750.35
Rectal and large intestinal atresia/stenosis	Q420	751.20–751.24
Pyloric stenosis	Q400	750.51

<b>Birth Defect</b>	<b>ICD-10 Code</b>	<b>CDC/BPA Codes</b>
Hirschsprung's disease	Q431	751.30–751.34
Biliary atresia	Q445	751.65
<b>Genitourinary</b>		
Renal agenesis/hypoplasia	Q600	753.00
Bladder exstrophy	Q641	753.50
Obstructive genitourinary defect	Q620	753.20-29–753.60-69
Hypospadias and epispadias	Q540, Q640	752.60–752.627 (exclude 752.610, 752.621)
<b>Musculoskeletal</b>		
Reduction deformity, upper limbs	Q710	755.20–755.29
Reduction deformity, lower limbs	Q720	755.30–755.39
Gastroschisis/omphalocele	Q793	756.71
Congenital hip dislocation	Q650	754.30
Diaphragmatic hernia	Q790	756.610–756.617
<b>Chromosomal</b>		
Down syndrome (Trisomy 21)	Q900	758.10–758.19
Patau syndrome (Trisomy 13)	Q914–Q917	758.00–758.09
Edwards syndrome (Trisomy 18)	Q910–Q913	758.20–758.290
<b>Other</b>		
Fetus or newborn affected by maternal alcohol use	Q860	760.71

**Appendix Table 5a. Average Prevalence of Selected Major Birth Defects – Active Surveillance Systems, U.S., 1999–2001<sup>(7)</sup>**

Birth Defects	Active Surveillance Systems (N=11)*		
	Cases	Prevalence	95% Confidence Interval
<b>Central nervous system defects</b>			
Anencephalus	675	2.60	2.41–2.81
Spina bifida without anencephalus	969	3.74	3.51–3.98
Encephalocele	248	0.96	0.84–1.08
<b>Eye defects</b>			
Anophthalmia/microphthalmia	542	2.09	1.92–2.27
<b>Cardiovascular defects</b>			
Truncus arteriosus (aka common truncus)	212	0.82	0.71–0.93
Transposition of great arteries	1,229	4.74	4.48–5.01
Tetralogy of Fallot	1,000	3.85	3.62–4.10
Atrioventricular septal defect (aka endocardial cushion defect)	1,120	4.32	4.07–4.58
Hypoplastic left heart syndrome	619	2.39	2.21–2.58
<b>Orofacial defects</b>			
Cleft palate	1,629	6.28	5.98–6.59
Cleft lip with and without cleft palate	2,733	10.54	10.15–10.94
<b>Gastrointestinal defects</b>			
Esophageal atresia/tracheoesophageal fistula	606	2.34	2.16–2.53
Rectal and large intestinal atresia/stenosis	1,255	4.84	4.58–5.11
Reduction defect, upper limbs	996	3.84	3.61–4.09
Reduction defect, lower limbs	496	1.91	1.75–2.09
<b>Musculoskeletal defects</b>			
Gastroschisis	991	3.82	3.59–4.07
Omphalocele	537	2.07	1.90–2.25
Diaphragmatic hernia	757	2.92	2.72–3.13
<b>Chromosomal defects</b>			
Down syndrome (Trisomy 21)	3,357	12.94	12.51–13.39
Trisomy 13	338	1.30	1.17–1.45
Trisomy 18	594	2.29	2.11–2.48

Note: Prevalence per 10,000 live births.

\*Eleven U.S. states with active birth defects surveillance systems, number of live births for 1999–2001 = 2,594,127

**Appendix Table 5b. Average Prevalence of Selected Major Birth Defects – Passive Surveillance Systems With Follow-Up, U.S., 1999–2001<sup>(7)</sup>**

Birth Defects	Passive Surveillance Systems with Follow-Up (N=4)*		
	Cases	Prevalence	95% Confidence Interval
<b>Central nervous system defects</b>			
Anencephalus	124	0.84	0.71–1.00
Spina bifida without anencephalus	325	2.21	1.98–2.46
Encephalocele	93	0.63	0.52–0.77
<b>Eye defects</b>			
Anophthalmia/microphthalmia	115	0.78	0.65–0.94
<b>Cardiovascular defects</b>			
Truncus arteriosus (aka common truncus)	96	0.65	0.53–0.80
Transposition of great arteries	560	3.80	3.50–4.13
Tetralogy of Fallot	593	4.03	3.72–4.37
Atrioventricular septal defect (aka endocardial cushion defect)	393	2.67	2.42–2.95
Hypoplastic left heart syndrome	318	2.16	1.94–2.41
<b>Orofacial defects</b>			
Cleft palate	916	6.22	5.83–6.64
Cleft lip with and without cleft palate	1,208	8.21	7.76–8.68
<b>Gastrointestinal defects</b>			
Esophageal atresia/tracheoesophageal fistula	377	2.56	2.32–2.83
Rectal and large intestinal atresia/stenosis	589	4.00	3.69–4.34
Reduction defect, upper limbs	342	2.32	2.09–2.58
Reduction defect, lower limbs	213	1.45	1.27–1.66
<b>Musculoskeletal defects</b>			
Gastroschisis	332	2.26	2.03–2.51
Omphalocele	208	1.41	1.23–1.62
Diaphragmatic hernia	291	1.98	1.76–2.22
<b>Chromosomal defects</b>			
Down syndrome (Trisomy 21)	1,797	12.21	11.66–12.79
Trisomy 13	129	0.88	0.74–1.04
Trisomy 18	206	1.40	1.22–1.60

Note: Prevalence per 10,000 live births.

\*Four U.S. states with passive birth defects surveillance systems with follow-up, number of live births for 1999–2001 = 1,471,987

**Appendix Table 5c. Average Prevalence of Selected Major Birth Defects – Passive Surveillance Systems Without Follow-Up, U.S., 1999–2001<sup>(7)</sup>**

Birth Defects	Passive Surveillance Systems without Follow-Up (N=7)*		
	Cases	Prevalence	95% Confidence Interval
<b>Central nervous system defects</b>			
Anencephalus	241	1.10	0.97–1.25
Spina bifida without anencephalus	718	3.29	3.06–3.54
Encephalocele	141	0.65	0.55–0.76
<b>Eye defects</b>			
Anophthalmia/microphthalmia	217	0.99	0.87–1.14
<b>Cardiovascular defects</b>			
Truncus arteriosus (aka common truncus)	156	0.73	0.62–0.85
Transposition of great arteries	779	3.57	3.33–3.83
Tetralogy of Fallot	916	4.20	3.94–4.48
Atrioventricular septal defect (aka endocardial cushion defect)	681	3.12	2.90–3.37
Hypoplastic left heart syndrome	548	2.51	2.31–2.73
<b>Orofacial defects</b>			
Cleft palate	980	4.49	4.22–4.78
Cleft lip with and without cleft palate	1,757	8.05	7.69–8.44
<b>Gastrointestinal defects</b>			
Esophageal atresia/tracheoesophageal fistula	513	2.35	2.16–2.56
Rectal and large intestinal atresia/stenosis	814	3.73	3.48–4.00
Reduction defect, upper limbs	379	1.74	1.57–1.92
Reduction defect, lower limbs	225	1.03	0.91–1.18
<b>Musculoskeletal defects</b>			
Gastroschisis	N/A	N/A	N/A
Omphalocele	N/A	N/A	N/A
Diaphragmatic hernia	632	2.90	2.68–3.13
<b>Chromosomal defects</b>			
Down syndrome (Trisomy 21)	2,579	11.82	11.37–12.29
Trisomy 13	197	0.90	0.79–1.04
Trisomy 18	358	1.64	1.48–1.82

Note: Prevalence per 10,000 live births. N/A = not applicable, share an ICD-9 code and cannot be distinguished by systems relying solely on administrative data sources.

\*Seven U.S. states with passive birth defects surveillance systems without follow-up, number of live births for 1999–2001 = 2,181,579

**Appendix Table 5d. Prevalence of Selected Birth Defects in North Dakota Between 1995-1999 and 2001-2005**

<b>Birth Defect</b>	<b>1995-1999</b>	<b>2001-2005</b>
Anencephaly	2.9	2.5
Spina Bifida	7.1	5.7
Cleft Palate	6.9	10.7
Cleft Lip (With and Without Cleft Palate)	13.5	10.0
Ventricular Septal Defect	43.4	44.5
Atrial Septal Defect	37.3	62.5
Hypoplastic Left Heart Syndrome	2.9	3.5
Coarctation of the Aorta	5.4	5.5
Tetraology of Fallot	5.4	4.5
Endocardial Cushion Defect	3.4	3.0
Transposition of the Great Arteries/Vessels	2.7	4.7
Common Truncus	2.0	0.5
Aortic Valve Stenosis	4.2	2.7
Tricuspid Valve Atresia	1.0	1.2

## *Glossary of Congenital Anomalies*<sup>(22 – unless noted)</sup>

---

- **Amniotic bands**<sup>(1)</sup>
  - (ABS) also called amniotic constriction band syndrome is a set of congenital birth defects believed to be caused by entrapment of fetal parts (usually a limb or digits) in fibrous amniotic bands while in utero.
- **Anencephaly**
  - Congenital absence of the skull and brain.
- **Aniridia**
  - Congenital absence of the iris of the eye.
- **Anophthalmia**
  - The absence of one or both eyes. These rare disorders develop during pregnancy and can be associated with other birth defects.
- **Anotia**
  - Occurs when there is a complete absence of the auricle (external ear, also called the pinna) and auditory canal.
- **Aortic valve stenosis**
  - Congenital heart defect characterized by aortic valve narrowing reducing the flow of blood.
- **Atrial septal defect**
  - Congenital heart defect characterized by one more openings in the atrial septum (wall between the right and left atria). The opening between the heart chambers disrupts the flow of blood and oxygen to the body.
- **Biliary atresia**
  - Congenital absence of the ducts in the biliary tract.
- **Bladder exstrophy**
  - Congenital exposure of the bladder mucosa caused by incomplete closure of the anterior bladder wall and the abdominal cavity.
- **Choanal atresia**
  - This is a congenital anomaly in which a bony or membranous formation blocks the passageway between the nose and the pharynx. This defect is usually repaired surgically after birth.
- **Cleft lip**
  - Congenital defect of the upper lip in which there is incomplete closure.
- **Cleft palate**
  - Congenital defect in the closure of the palate, the structure that separates the nasal cavities and the back of the mouth. May involve the soft palate, hard palate or alveolus (gum).
- **Coarctation of the aorta**
  - Congenital heart defect characterized by narrowing of the descending aorta.
- **Common truncus**
  - Congenital heart defect characterized by a single great arterial trunk, instead of a separate aorta and pulmonary artery. Commonly known as truncus arteriosus.
- **Congenital cataract**
  - An opacity (clouding) of the lens of the eye.
- **Congenital hip dislocation**
  - Congenital dislocation of one or both hips.

- **Diaphragmatic hernia**
  - Congenital defect of the muscular diaphragm resulting in herniation of the abdominal contents into the chest.
- **Down syndrome (Trisomy 21)**
  - Distinctive and common chromosome abnormality syndrome caused by an extra copy of chromosome 21. Can be complete (Trisomy 21), attached to another chromosome (translocation), or mixed with cells containing normal chromosomes (mosaic).
- **Ebstein's anomaly**
  - Congenital heart defect characterized by downward displacement of the tricuspid valve into the right ventricle.
- **Encephalocele**
  - Congenital defect of the skull resulting in herniation of the brain.
- **Endocardial cushion defect**
  - Congenital heart defect characterized by a combined atrial and ventricular septal defect and common atrioventricular valve (instead of distinct tricuspid and mitral valves).
- **Epispadias**
  - Congenital defect of the genitals where the opening of the urethra is located on the upper side of the penis in boys and between the clitoris and labia in girls.
- **Esophageal atresia/tracheoesophageal fistula**
  - The narrowing or incomplete formation of the esophagus. This is usually a surgical emergency. Frequently associated with a *tracheoesophageal fistula*.
- **Fetal alcohol spectrum disorders (fetus or newborn affected by maternal alcohol use)<sup>(13)</sup>**
  - Fetal alcohol spectrum disorders (FASDs) are a group of conditions that can occur in a person whose mother drank alcohol during pregnancy. These effects can include physical problems and problems with behavior and learning.
- **Gastroschisis**
  - Congenital opening of the abdominal contents. Can be distinguished from omphalocele by location usually to the right of the umbilicus.
- **Hirschsprung's disease (congenital megacolon)**
  - Congenital aganglionic megacolon (enlarged colon) due to absent nerves in the wall of the colon.
- **Hydrocephalus**
  - Accumulation of fluid within the spaces of the brain. Can be congenital or acquired.
- **Hypoplastic left heart syndrome**
  - Congenital heart defect characterized by extreme smallness of left-sided structures. Classically, aortic valve/mitral valve atresia or marked hypoplasia, ascending aorta and left ventricle hypoplasia.
- **Hypospadias**
  - Congenital defect of the penis in which the urethral opening is on the underside of the penis.
- **Microcephaly**
  - Small head, with corresponding smallness of the brain.

- **Microphthalmia**
  - Congenital smallness of the eye globe; a disorder in which one or both eyes are abnormally small.
- **Microtia**
  - Congenital smallness or maldevelopment of the external ear, with or without absence or narrowing of the external auditory canal. Consists of malformation or hypoplasia (decreased growth) of the auricle. This malformation may range from a small but otherwise normal external ear to an external ear with major structural changes. The external auditory meatus (canal) also may be narrow or absent.
- **Obstructive genitourinary defect**
  - Congenital narrowing or absence of the urinary tract structure at any level. Severity often depends upon the level of the obstruction.
- **Omphalocele**
  - Congenital opening of the abdominal wall with protrusion of the abdominal contents. Can be distinguished from gastroschisis by location within umbilical ring.
- **Patent ductus arteriosus**
  - Congenital heart defect characterized by persistence of the fetal blood vessel connecting the pulmonary artery and the aorta.
- **Pulmonary valve atresia/stenosis**
  - Congenital heart defect characterized by absence (or narrowing) of the pulmonary valve or pulmonary artery itself.
- **Pyloric stenosis**
  - A congenital narrowing of the opening of the stomach into the small intestine.
- **Rectal and large intestinal atresia/stenosis**
  - Congenital absence, closure or constriction of the large intestine, rectum or anus.
- **Reduction deformity, lower limbs**
  - Congenital absence of a portion of or an entire limb (legs).
- **Reduction deformity, upper limbs**
  - Congenital absence of a portion of or an entire limb (arms).
- **Renal agenesis/hypoplasia**
  - Congenital absence of the kidney.
- **Spina bifida**
  - Neural tube defect with protrusion of the spinal cord and/or meninges.
- **Tetralogy of Fallot**
  - Congenital heart defect composed of ventricular septal defect, pulmonary stenosis or atresia, displacement of the aorta to the right, and hypertrophy of right ventricle.
- **Transposition of great vessels (arteries)**
  - Congenital heart defect in which the aorta rises from the right ventricle and the pulmonary artery arises from the left ventricle (opposite of normal).
- **Tricuspid valve atresia and stenosis**
  - Congenital heart defect characterized by the absence (or narrowing of) of the tricuspid valve.
- **Trisomy 13 (Patau syndrome)**
  - Chromosome abnormality caused by an extra chromosome 13.
- **Trisomy 18 (Edwards syndrome)**
  - Chromosomal abnormality caused by an extra chromosome 18.

- **Ventricular septal defect**
  - Congenital heart defect characterized by one or several openings in the ventricular septum.

## *List of Acronyms*

---

<b><u>MCH</u></b>	Maternal and Child Health
<b><u>PRAMS</u></b>	Pregnancy Risk Assessment Monitoring System
<b><u>ICD-9</u></b>	International Classification of Disease Version 9
<b><u>ICD-10</u></b>	International Classification of Disease Version 10
<b><u>CDC/BPA</u></b>	Centers for Disease Control and Prevention (CDC)/ British Pediatric Association (BPA)
<b><u>NBDPN</u></b>	National Birth Defects Prevention Network
<b><u>ASD</u></b>	Atrial septal defect
<b><u>BSD</u></b>	Ventricular septal defect
<b><u>PDA</u></b>	Patent ductus arteriosus
<b><u>FASD</u></b>	Fetal alcohol spectrum disorder
<b><u>FAS</u></b>	Fetal alcohol syndrome
<b><u>ARND</u></b>	Alcohol-related neurodevelopment disorders
<b><u>ARBD</u></b>	Alcohol-related birth defects
<b><u>CP</u></b>	Cleft palate without cleft lip
<b><u>CLP</u></b>	Cleft lip with or without cleft palate
<b><u>RP</u></b>	Relative prevalence
<b><u>OGD</u></b>	Obstructive genitourinary defect
<b><u>CNS</u></b>	Central nervous system
<b><u>NTD</u></b>	Neural tube defect
<b><u>CI</u></b>	Confidence interval
<b><u>CSF</u></b>	Cerebrospinal fluid
<b><u>CHD</u></b>	Congenital hip dislocation

## *Prevention Methods*

---

By taking precautions before and during pregnancy, a woman can reduce her risk of delivering a baby born with a birth defect or other adverse outcome.

A woman should start planning for the health of her baby before she becomes pregnant. During the first three to eight weeks after conception, many of the baby's vital organs and systems are being formed. By the time most women know they are pregnant, their baby's development is well underway, and some birth defects already may have occurred.

While there is never a guarantee for a healthy baby, the following list of preventive measures can increase a women's chance of having a healthy pregnancy and a healthy baby.

- **Talk with your health-care provider.**  
It is a good idea to talk with a health-care professional before becoming pregnant. During this time, a health-care provider can identify any health risks a woman may be facing and can work with her to address them before she becomes pregnant. It is important to have conditions such as diabetes, epilepsy and high blood pressure under control before becoming pregnant. If there is history of an inherited or genetic disorder, consultation with a genetic counselor may be recommended.
- **Consume folic acid.**  
Several studies have shown that women who take a daily multivitamin with 400 micrograms of folic acid before and during pregnancy decrease the risk by as much as 70 percent that their baby will be born with a neural tube defect. Consuming folic acid also may prevent other birth defects such cleft lip/cleft palate and some congenital heart defects. For adults, folic acid may offer protection from illnesses such as heart disease and colon cancer.
- **Eat a healthy diet.**  
Women and their developing babies can benefit from good nutritional habits before and during pregnancy. It is highly recommended that all women eat a well-balanced and varied diet and take a multivitamin daily.
- **Exercise regularly.**  
Regular exercise can benefit a woman's body by increasing overall strength and creating a healthy environment in which her baby can develop. Talk with a health-care provider to determine an appropriate exercise level.
- **Achieve an ideal weight.**  
The preconception period is an excellent time to achieve an ideal weight. Women who start their pregnancies underweight or overweight may have problems. A woman who is overweight at the time of conception is more likely to develop high blood pressure and diabetes during pregnancy. A woman who is underweight is more likely to deliver a low birth-weight baby.
- **Avoid smoking.**  
Women should avoid smoking during pregnancy and should limit exposure to secondhand smoke. Smoking during pregnancy is associated with an increased risk of

miscarriage and stillbirth, SIDS (sudden infant death syndrome) and low birth weight. In addition, children exposed to smoke may have behavior problems, learning difficulties and an increased risk for respiratory problems and asthma.

- **Avoid alcohol.**

The harmful effects of alcohol on an unborn baby's growth and development are numerous. Fetal alcohol syndrome (FAS) is the most severe, creating physical, mental and behavioral problems in infants. Alcohol consumption during pregnancy is the leading cause of preventable mental retardation among infants.

- **Avoid illicit drugs.**

Research has shown that in-utero exposure to illicit drugs can cause direct toxic effects on a developing baby, as well as create fetal and maternal dependency. The baby may experience withdrawal prenatally when drugs are withdrawn from a dependent mother, or after delivery.

- **Limit exposure to environmental hazards.**

Pregnant women should limit exposure to toxic substances and chemicals. They also should avoid eating undercooked meat and handling cat litter, as these activities may lead to an infection known as toxoplasmosis, which can seriously harm a developing fetus. A few foods – including certain types of fish, some soft cheeses and ready-to-eat meats – also may pose a risk during pregnancy.

- **Discuss medications.**

A woman should discuss all medications that she is taking – whether prescription or over-the-counter – with a pharmacist or health-care provider, as these may not be appropriate to use during pregnancy.

- **Check immunizations.**

It is important for women to check her immunization history before pregnancy. If she is not immune to chickenpox and rubella or if she has not received her hepatitis B series, she should talk with her health-care provider about her risks.

## *North Dakota Resources*

---

**Children's Special Health Services (CSHS)** is a program that helps children with special health-care needs and their families. Individuals are encouraged to call CSHS for free health-care information. CSHS can also help with referrals to health-care providers and special programs for children with chronic health conditions.

Children's Special Health Services  
North Dakota Department of Health  
600 E. Boulevard Ave., Dept. 301  
Bismarck, ND 58505-0200  
Phone: 701.328.2436  
Toll-free: 800.755.2714  
Fax: 701.328.1645  
E-mail: [dohchshsadm@nd.gov](mailto:dohchshsadm@nd.gov)  
[www.ndhealth.gov/CSHS/](http://www.ndhealth.gov/CSHS/)

**Family Voices** is health-care information and education program which includes the Parent to Parent Project.

Family Voices  
PO Box 163  
312 2nd Ave. W.  
Edgeley, ND 58433  
Phone: 701.493.2634  
Toll-free: 888.522.9654  
Fax: 701.493.2635  
E-mail: [fvnd@drtel.net](mailto:fvnd@drtel.net)  
[www.fvnd.org/](http://www.fvnd.org/)

**Genetic Services**  
School of Medicine and Health Sciences Room 5910  
501 N. Columbia Road STOP 9037  
Grand Forks, ND 58201  
Phone: 701.777.4277 or 701.772.5301  
[www.med.und.nodak.edu/pediatrics/genetics/](http://www.med.und.nodak.edu/pediatrics/genetics/)

**Meritcare**  
Meritcare Broadway  
737 Broadway N.  
Fargo, ND 58102  
Phone: 701.234.2362  
[www.merticare.com](http://www.merticare.com)

**March of Dimes Birth Defects Foundation**

North Dakota Chapter

1330 Page Drive, Ste. 102

Fargo, ND 58103

Phone: 701.235.5530

[www.marchofdimes.com/northdakota/northdakota.asp](http://www.marchofdimes.com/northdakota/northdakota.asp)

## *Highlighted Birth Defect Resources*

---

The information and resources listed here are intended for educational use only and are provided solely as a service. The information provided should not be used for diagnosing or treating a health problem or disease and is not a substitute for professional care. These resources do not constitute an endorsement of these organizations or their programs by the North Dakota Department of Health and the National Birth Defects Prevention Network (NBDPN) and none should be inferred. The North Dakota Department of Health or NBDPN is not responsible for the content of the individual organization web pages/resources found at these links.

### **I. Highlighted Resources for Women and Health Care Professionals**

[http://www.nbdpn.org/current/2009MSWord/BDPM2009\\_HighlightedResources.doc](http://www.nbdpn.org/current/2009MSWord/BDPM2009_HighlightedResources.doc)

#### **1. What To Expect at Your Prenatal Visits:**

<https://www.med.uvm.edu/vchip/Downloads/PrenatalCarePoster.doc>

- This poster provides women with information about expectations at their prenatal visits to their physician. It was developed through funding from the March of Dimes and the Vermont Department of Health.

#### **2. A Healthy Baby Is Worth the Weight:**

<http://www.healthy-baby.org/HowMuchWeight.htm>

- This is Colorado's Healthy Women Campaign website. It provides information for women about appropriate weight gain and what happens to the weight during pregnancy. It also has a body mass index (BMI) calculator for women to use to track their BMI before and during pregnancy. Women can use the calculator to determine their BMI before they are pregnant. Then, women can use the BMI chart to figure out how much weight to gain during pregnancy.

#### **3. Preconception Screening and Counseling Checklist:**

[http://www.marchofdimes.com/files/preconception\\_tool\\_ed.pdf](http://www.marchofdimes.com/files/preconception_tool_ed.pdf)

- This March of Dimes one-page checklist can be used by health providers to initiate discussions about preconception care. This list offers a series of essential questions that help providers develop good clinical management plans. Patients are given the checklist at the reception desk and are asked to fill it out before seeing the provider.

#### **4. Preconception Check List – March of Dimes:**

[http://www.marchofdimes.com/printableArticles/19583\\_4182.asp](http://www.marchofdimes.com/printableArticles/19583_4182.asp)

- This March of Dimes resource can assess preconception health with all female patients of childbearing age during routine checkups. It is a way of encouraging healthy habits that can affect pregnancy outcome. The March of Dimes, in conjunction with the Greater Adirondack Perinatal Network, has developed two unique screening and counseling tools for health-care providers to use with their patients.

**5. Nutrition Tools and Resources:**

<https://www.med.uvm.edu/vchip/TB1+BL.asp?ContentItemID=10154&SiteAreaID=669>

- This resource is produced by the Collaborating to Improve Maternal, Child, Youth, and Family Healthcare in Vermont through Innovation and Excellence by The University of Vermont College of Medicine. The web page contains nutritional resources for health professionals to use in their practice and patient education.

**6. Fitness for Two – March of Dimes:**

[http://www.marchofdimes.com/printableArticles/14332\\_1150.asp](http://www.marchofdimes.com/printableArticles/14332_1150.asp)

- This March of Dimes website provides health professionals and women information about how to keep fit during pregnancy. It answers questions women have about keeping fit during their pregnancy.

**7. Are You Ready Physically?:**

[http://www.marchofdimes.com/printableArticles/173\\_14005.asp](http://www.marchofdimes.com/printableArticles/173_14005.asp)

- This March of Dimes web page provides women with the essential activities that they can do to get healthy before becoming pregnant. It includes “What you need to know before you get pregnant” and “10 steps to getting healthy before pregnancy.”

**8. March of Dimes Product Catalog:**

[http://www.marchofdimes.com/printableArticles/2222\\_2282.asp](http://www.marchofdimes.com/printableArticles/2222_2282.asp)

- This March of Dimes web page provides fact sheets for patients and health professionals. Additional fact sheets appear on the March of Dimes website and on the Spanish website. It also contains a link to the order form and product catalog. It is of particular interest for addressing topics related to preconception health and pregnancy.

**9. Healthy Pregnancy Resources – March of Dimes Product Catalog:**

[http://www.marchofdimes.com/professionals/2222\\_2262.asp](http://www.marchofdimes.com/professionals/2222_2262.asp)

- The March of Dimes complete line of prenatal materials covers everything expectant parents need to know about the growth and development of their baby and how to take good care of themselves during these special months. Two resources of particular interest are **Eating Healthy** (English and Spanish) and **I Want My 9 Months** (flyer/poster – English and Spanish).

**10. Baby Your Baby:**

<http://www.babyyourbaby.org/duringpregnancy/weightgain.htm>

- Baby Your Baby is Utah Health Department’s website that provides information to address weight gain during a woman’s pregnancy and following the birth of the baby. The site has an online BMI calculator to assist a woman in determining the recommended amount of weight that she should gain during her pregnancy. Used by permission by the staff in the Maternal Child Health Division at the Utah Health Department.

### **11. BMI – Body Mass Index:**

<http://www.cdc.gov/nccdphp/dnpa/bmi/>

- The Centers for Disease Control and Prevention provides this website for general information about the body mass index (BMI) and links to BMI calculators for adults, children and teens. There is also information regarding the BMI number interpretation, nutrition and weight resources. Other topics such as nutrition, physical activity, overweight and obesity are available. The site content is through the Division of Nutrition, Physical Activity and Obesity, National Center for Chronic Disease Prevention and Health Promotion.

### **12. Adult BMI Calculator – Adults – English:**

<http://www.cdc.gov/nccdphp/dnpa/bmi/>

- The Centers for Diseases Control and Prevention’s website is an online calculator that provides BMI and corresponding BMI weight status categories for adults age 20 years and older. The content source is the Division of Nutrition, Physical Activity and Obesity, National Center for Chronic Disease Prevention and Health Promotion.

### **13. BMI for Children and Teens – English:**

[http://www.cdc.gov/nccdphp/dnpa/bmi/childrens\\_BMI/about\\_childrens\\_BMI.htm](http://www.cdc.gov/nccdphp/dnpa/bmi/childrens_BMI/about_childrens_BMI.htm)

- The website provides information about the body mass index (BMI) relating to children and adolescents. The BMI is used as a screening tool to identify possible weight problems for children. CDC and the American Academy of Pediatrics (AAP) recommend the use of BMI to screen for overweight in children beginning at age 2.

### **14. Learn About Body Mass Index (BMI):**

<http://www.nhlbi.nih.gov/health/public/heart/obesity/wecan/learn-it/bmi-chart.htm>.

- The National Institutes of Health (NIH) provides information about various topics related to health. The Learn It looks at ways to enhance children’s health and nutrition. The Learn About Body Mass Index (BMI) provides easy-to-read information for both adults and children regarding the BMI, its usefulness and how to calculate a BMI. It is also printable in a PDF for use by health professionals and families.

## **II. Resources Related to Obesity, Diabetes and Pregnancy**

### **1. Healthy Eating During Pregnancy (You & Your Baby):** Dr. Laura Riley and Stacey Nelson, M.S., R.D., L.D.N.

- This book provides women with a guide to eating well and staying fit during pregnancy. The book is available in paperback.

### **2. Obesity, Diabetes, and Links to Congenital Defects: A Review of the Evidence and Recommendations for Intervention:** By E. Albert Reece. *The Journal of Maternal-Fetal & Neonatal Medicine*, Vol. 21, Issue 3, 2008.

- This is a review of the literature, primarily epidemiologic studies, linking obesity and obesity-related metabolic disturbances in pregnant women to a range of birth defects. The author found that obesity and diabetes are a growing problem in the

U.S. population. It was noted that the problem is especially important among women of childbearing age since the dual effect of obesity along with diabetes impacts the unborn baby – although, each impacts it independently.

3. **Screening for Gestational Diabetes Mellitus: U.S. preventive Services Task Force Recommendation Statement:** *Annals of Internal Medicine*. May 20, 2008 Vol. 148, No. 10, pp 759-765.
  - This is an update of the 2003 U.S. Preventive Services Task Force recommendations about screening for gestational diabetes. The task force concludes that the current evidence is insufficient to assess the balance of benefits and harms of screening for gestational diabetes mellitus, either before or after 24 weeks' gestation.
  
4. **A Prospective Study on the Risk of Congenital Defects Associated with Maternal Obesity and Diabetes Mellitus:** By Lynn L. Moore, Martha R. Singer, Loring M. Bradlee, et al. In *Epidemiology*, 11 (6):689-694, November 2000.
  - This study looked at the effects of maternal obesity and diabetes mellitus on the risk of congenital defects. The results suggest that obesity and diabetes mellitus may act synergistically in the pathogenesis of congenital anomalies. The defects were primarily craniofacial or musculoskeletal.
  
5. **Advances in Understanding the Molecular Causes of Diabetes-Induced Birth Defects:** By Mary R. Loeken. In *Journal of the Society for Gynecologic Investigation*, 2006; 13:2-10.
  - This is a review article regarding the molecular causes of birth defects from diabetic pregnancy, with a focus on neural tube defects – using a mouse model. The conclusion is that excel glucose metabolism by embryos resulting from maternal hyperglycemia disturbs a complex network of biochemical pathways, leading to oxidative stress. Impaired embryo gene expression resulting from oxidative stress, and consequent apoptosis or disturbed organogenesis, can be a general mechanism to explain diabetic embryopathy. While this is an excellent review, the content requires a deeper level of understanding and, as such, not useful for the non-medical reader.
  
6. **Insensitivity to Insulin and Obesity: The Underlying Cause:** By Robert Eckel and Scott Grundy. In *Diabetes Voice*, May 2006, Vol. 51, Special Issue, pp 28-30.
  - The authors examine the underlying causes of obesity and insensitivity to insulin. The report addresses the relationship between the level of obesity and its relating degree of impact on the body's insensitivity to insulin. Asians are reported to have a higher predisposition to abdominal obesity; thus, this population is at increased risk for diabetes and heart disease.

### III. Resources for Education by Health Professionals

1. **Risks of Being Overweight for Women of Reproductive Age:**  
<http://www.ci.minneapolis.mn.us/dhfs/reproductiveweight.pdf>
  - This is a Research Brief by the Minneapolis Department of Health and Family Support, dated January 2008. It provides easy-to-read information about weight, the body mass index (BMI) and the impact of weight gain on women and their babies. It also provides strategies for health professionals.
2. **A Call to Action: Obesity and Pregnancy – Women’s Health Policy Brief:** By Laura Riley, M.D., Massachusetts General Hospital Winter/Spring 2006
  - It was prepared by the Women’s Health Coordinating Council at Massachusetts General Hospital. It includes awareness and education about obesity in women.
3. **January is Birth Defects Prevention Month...but any month is the month to prevent birth defects:** By Hema Joshi and Debra L. Thompson. *Georgia Epidemiology Report*. January 2006, Vol. 22, No. 01. Published by the Georgia Department of Human Resources, Division of Public Health.
4. **WEBTREATS: Diet, Weight Management and Obesity:**
  - This list, prepared by ACOG Resource Center Librarians from other sources, is provided for information only. Referral to these sites does not imply the endorsement of The American College of Obstetricians and Gynecologists of the organization or their contents, expressed views, programs, or political activities. Refer to the WEBTREAT on Exercise and Physical Fitness for related links. [pvh/mash](http://pvh/mash) 3/19/2008. For more information, contact [resources@acog.com](mailto:resources@acog.com)
5. **Pregnancy-Related Nutrition:** By Geraldine S. Berry, Colette L. Zyrkowski, Linda D. Clark, and Stella Yu. Reproductive Health of Women is a part of *From Data to Action* and CDC’s *Public Health Surveillance for Women, Infants, and Children*.
  - This report addresses the nutritional issues for women during pregnancy from a public health perspective. The demonstrated use of data provides readers with insight of data interpretation and use.

### IV. Resources Related to Obesity and Prepregnancy

1. **Prepregnant Obesity and Risks of Selected Birth Defects in Offspring:** Brief Report. By Gary M. Shaw and Suzan L. Carmichael. Found in *Epidemiology*, 19 (4):616-620, July 2008. Copyright 2008 Lippincott Williams & Wilkins, Inc.
  - This study examined the impact of prepregnant obesity on several birth defects, especially neural tube defects. The conclusion indicated that the data did not fully support earlier findings with respect to the relationships of obesity with anencephaly and spina bifida.

2. **Trends in pre-pregnancy obesity in nine states, 1993-2003:** By Shin Y. Kim, Patricia M. Dietz, Lucinda England, Brian Morrow, and William M. Callaghan. *Obesity*. Vol. 15, April 2007, pp. 986-993.
3. **Prevalence of Overweight and Obesity in the United States, 1999-2004:** <http://jama.ama-assn.org/cgi/content/full/295/13/15490> By Cynthia L. Ogden, Margaret D. Carroll, Lester R. Curtin; et al. *Journal of American Medical Association*, April 5, 2006, Vol. 295, No. 13, pp. 1549-1555.
4. **Prepregnancy Obesity as a Risk Factor for Structural Birth Defects:** By Kim D. Waller, Gary M. Shaw, et al. – National Birth Defects Prevention Study in *Archives of Pediatric & Adolescent Medicine*. Vol. 161 (No. 8), August 2007: 745-750.
  - This population-based study describes the relation between maternal obesity, overweight and underweight status and 16 categories of structural birth defects.
  - The results involving nearly 15,000 women from eight states found abnormalities of the spine, heart, arms, legs and abdomen, building on previous research that showed heart and spine defects. The greatest risk was for spina bifida.
  - Obese women faced double the risk of having babies with spina bifida than women of healthy weight. With spina bifida, the most common disabling birth defect in the United States, the spinal column fails to close properly. That often leads to leg paralysis, learning difficulties and other serious problems. Very heavy women also were 60 percent more likely to have babies born with a rare defect in which abdominal organs protrude through the belly button; 40 percent more likely to have heart defects; 36 percent more likely to have shortened arms or legs; and at least 20 percent more likely to have any of several gastrointestinal deformities. The research is part of the National Birth Defects Prevention Study involving women who were pregnant between October 1997 and December 2002.
5. **Weight Control: Eating Right and Keeping Fit:** [ACOG Education Weight Control Eating Right and Keeping Fit.mht](#) [ISSN 1074-8601]
  - This ACOG site promotes this educational pamphlet addressing maintaining a healthy weight and the factors affecting weight. To order patient education pamphlets in packs of 50, call 800.762.2264 or order online at [sales.acog.org](http://sales.acog.org). Requests for authorization to make photocopies should be directed to the Copyright Clearance Center, 222 Rosewood Drive, Danvers, Mass. 01923.

## V. Resources Related to Obesity and Pregnancy

1. **Maternal Obesity and Risk of Neural Tube Defects: A Metaanalysis:** By Sonja A. Rasmussen, Susan Y. Chu, Shin Y. Kim, et al. *American Journal of Obstetrics & Gynecology*, June 2008, pp 611-619.
  - This is a metaanalysis of published evidence on the relationship between maternal obesity and the risk of neural tube defects (NTDs). The findings support that maternal obesity is associated with an increased risk for an NTD-affected pregnancy.

**2. Birth Defects Research:**

<http://www.cdc.gov/ncbddd/bd/research.htm>

- This CDC website contains updated information regarding birth defects and overviews of related research. It discusses the National Birth Defects Prevention Study, U.S.-China Collaborative Project and other CDC specific research.

**3. Is Maternal Obesity a Risk Factor for Anencephaly and Spina Bifida?:** By Margaret L. Watkins, Kelley S. Scanlon, Joseph Mulinare and Muin J. Khoury. *Epidemiology*, September 1996, Vol. 7, No. 5, pp. 507-512.

**4. Obesity during Pregnancy Threatens Health of Both Mother and Fetus, March of Dimes Says:** [http://www.marchofdimes.com/printableArticles/10651\\_12183.asp](http://www.marchofdimes.com/printableArticles/10651_12183.asp)

- WHITE PLAINS, N.Y., JUNE 9, 2004 – The rising obesity rate in the United States is a risk to the health of pregnant women and their babies, the March of Dimes said today. The report is published by the March of Dimes Task Force on Nutrition and Optimal Human Development.

**5. Maternal Obesity and Risk for Birth Defects:** By Margaret L Watkins, Sonjy A. Rasmussen, Margaret Honein, Lorenzo D. Botto and Cynthia Moore. In *Pediatrics* 2003;111; 1152-1158.

- This is a research study sponsored by the Division of Birth Defects and Developmental Disabilities, National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention, Atlanta, Ga. Contact [cdc.gov](http://cdc.gov). PMID: 12728129 (Pub Med – indexed for MEDLINE).
- This study was based on data from births in a five-county area of Atlanta from January 1993 to August 1997. Results indicated that obese women were more likely to have babies with spina bifida, omphalocele, heart defects or multiple anomalies. Women who were overweight were more likely to have babies with heart defects or multiple anomalies.

**6. Risk of Neural Tube Defect-affected Pregnancies among Obese Women:** By Shaw, GM, Velie, EM, and Schaffer, D. *Journal of American Medical Association* 1996; 275(14): 1093-1096. Also, noted in *Neural Tube Defects and Obesity* from California Birth Defects Monitoring Program, April 1999.

<http://www.cbtmp.org/pdf/ntdsobesity.pdf>.

- This study was done by the California Birth Defects Monitoring Program to examine the impact of obesity on the occurrence of babies born in California with neural tube defects. The results found that neural tube defects are more common in pregnancies to obese women.

**7. Maternal Obesity and Pregnancy – Weight Matters:**

[http://www.marchofdimes.com/files/MP\\_MaternalObesity040605.pdf](http://www.marchofdimes.com/files/MP_MaternalObesity040605.pdf).

(Medical Perspective on Prematurity) Prepared by the Office of the Medical Director April 6, 2005.

- This March of Dimes document discusses the fact that obesity impacts a woman's pregnancy; however, it is a modifiable risk factor that could improve birth outcomes.

8. **Overview of Clinical Perspectives and Mechanism of Obesity:** In *Birth Defects Research Part A-Clinical and Molecular Teratology* 73 (7): 470-471. (2005 July).
9. **Maternal Obesity and Neonatal Mortality According to Subtypes of Preterm Birth:** By Ellen A. Nohr, Michael Vaeth, Bodil H. Bech, et al. *Obstetrics & Gynecology*, 2007; 110:1083-1090.
  - This article evaluates the association between pre-pregnancy body mass index (BMI) and neonatal mortality while accounting for the timing of delivery and subtypes of preterm birth.
10. **Teratology Public Affairs Committee Position Paper: Maternal Obesity and Pregnancy:** By the Public Affairs Committee of the Teratology Society. Published in *Birth Defects Research (Part A): Clinical and Molecular Teratology* 76: 73-77 (2006). Copyright by 2006 Wiley-Liss, Inc.
  - This article has a list of references by condition. See Table 2 on page 74.
11. **The Implications of Maternal Overweight and Obesity on the Course of Pregnancy and Birth Outcomes:** By Anna-Maria Siega-Riz and Barbara Laraia. *Maternal Child Health Journal* (2006). 10:S153-S156.
  - This article is an overview of the complications associated with maternal overweight and obesity, including diabetes, pre-eclampsia, c-section, and birth defects. Included also is an overview of weight trends among women and prevention studies targeting adolescents and women prior to pregnancy.

## VI. Resources about Medication-Related Risk of Birth Defects

1. **Popular blood pressure drugs may increase risk of birth defects**  
By Jan Friedman. *New England Journal of Medicine*. 2006; Vol. X: pp xx (June 9, 2006).
2. **Major Congenital Malformations after First-Trimester Exposure to ACE Inhibitors:** By William O. Cooper, Sonia Hernandez, Patrick G. Arbogast, et al. *New England Journal of Medicine*, 2006, 354; 23, 2443-2451.
  - The use of angiotensin-converting-enzyme (ACE) inhibitors during the second and third trimesters of pregnancy was already known for their association with increased risk of birth defects. This study demonstrates the association between exposure to ACE inhibitors during the first trimester of pregnancy only and the risk of congenital malformations, especially those of the cardiovascular system and central nervous system.

## *Birth Defect Resources on the Internet*

---

The information and resources listed here are intended for educational use only and are provided solely as a service. The information provided should not be used for diagnosing or treating a health problem or disease and is not a substitute for professional care. These links do not constitute an endorsement of these organizations or their programs by the North Dakota Department of Health (ND DOH) or the National Birth Defects Prevention Network (NBDPN) and none should be inferred. The ND DOH or the NBDPN is not responsible for the content of the individual organization web pages found at these links.

**NOTE:** The Internet resources related to weight and nutrition are marked with an asterisk (\*).

### **I. Birth Defects Research and Prevention Information**

[http://www.nbdpn.org/current/2009MSWord/BDPM2009\\_InternetResources.doc](http://www.nbdpn.org/current/2009MSWord/BDPM2009_InternetResources.doc)

#### **1. International Clearinghouse for Birth Defects Surveillance and Research:**

<http://www.icbdsr.org>

- This site is dedicated to sharing data, news and views on the research, monitoring and prevention of congenital malformations. It provides information about the Clearinghouse, membership directory, publications/papers, and birth defects links for professionals and general audiences.

#### **2. March of Dimes (MOD) Foundation:**

<http://www.marchofdimes.com>

- This site contains a wealth of information about MOD, birth defects and infant health statistics. There are also additional links to other sources of birth defects data.

#### **3. National Birth Defects Prevention Network (NBDPN):**

<http://www.nbdpn.org>

- NBDPN is a national organization of individuals at the local, state and national level working in birth defects surveillance, research and prevention. This site offers organizational information, newsletters, a listserv and related links to state birth defects monitoring programs and other national organizations.

#### **4. Spina Bifida Association (SBA):**

<http://www.sbaa.org>

- The mission of SBA is to promote the prevention of spina bifida and to enhance the lives of all affected. This site offers downloadable patient education materials and folic acid tutorials for professionals.

## II. Birth Defects Databases

### 1. **GeneTests:**

<http://www.geneclinics.org>

- GeneTests is a medical knowledge base that relates genetic testing to the diagnosis, management and genetic counseling of individuals and families with specific inherited disorders. Its website contains a database of genetic diseases with summaries and diagnostic information, clinical descriptions, management details, resources and references.

### 2. **Medical Genetics, University of Kansas Medical Center:**

<http://www.kumc.edu/gec/geneinfo.html>

- This website contains a surplus of information from many national and international birth defects and genetics/birth defects organizations (including hyperlinks). Additionally, the site also contains referral information for those wishing to seek a health-care professional concerning a birth defect or genetic disorder.

## III. Family Support Groups/Information for Families

### 1. **The American College of Obstetricians and Gynecologists\*:**

<http://www.acog.org/>

- You can search the ACOG's public website to find electronic educational pamphlets. Enter *weight during pregnancy* in the Search box and you will find several titles including "*Healthy Women: Promoting Good Health Before Pregnancy*," "*Exercise During Pregnancy*" and "*Nutrition During Pregnancy*."
- The following links also offer electronic educational pamphlets:
  - Exercise and Fitness:  
[http://www.acog.org/publications/patient\\_education/bp045.cfm](http://www.acog.org/publications/patient_education/bp045.cfm)
  - Weight Control:  
[http://www.acog.org/publications/patient\\_education/bp064.cfm](http://www.acog.org/publications/patient_education/bp064.cfm)
  - Healthy Eating:  
[http://www.acog.org/publications/patient\\_education/bp130.cfm](http://www.acog.org/publications/patient_education/bp130.cfm)
  - Getting in Shape After Your Baby Is Born:  
[http://www.acog.org/publications/patient\\_education/bp131.cfm](http://www.acog.org/publications/patient_education/bp131.cfm)

### 2. **American Pregnancy Association\*:**

<http://www.americanpregnancy.org/index.htm>

- This site provides information for both women planning to become pregnant and pregnant women. It includes the following topics: planning and preparing for your baby; pregnancy wellness, such as pregnancy weight gain, pregnancy nutrition and exercise during pregnancy; and pregnancy complications.

### 3. **About Pregnancy and Childbirth\*:**

[http://pregnancy.about.com/od/healthduringpreg/index\\_r.htm](http://pregnancy.about.com/od/healthduringpreg/index_r.htm)

- This website features information about various subjects for pregnant women and moms to be, including eating disorders, thyroid, allergies, antidepressants, and alcohol.

**4. About Pediatrics:**

[http://pediatrics.about.com/od/parentingtips/a/05\\_fam\\_history.htm](http://pediatrics.about.com/od/parentingtips/a/05_fam_history.htm)

- This article provides a brief introduction for parents to the risk of disease and family history. The links included in the website provide visitors with additional information about topics such as creating a medical family tree to help determine genetic predisposition to certain diseases and genetic traits; prenatal and pediatric genetics screening questionnaires; *My Family Health Portrait* tool as developed by the U.S. Surgeon General's Family History Initiative; and information from the Mayo Clinic on family history and the role it plays in health and disease.

**5. Genetic Alliance: Advocacy, Education & Empowerment:**

<http://www.geneticalliance.org>

- Genetic Alliance is a national coalition of consumers, professionals and genetic support groups who voice the common concerns of children, adults and families living with and at risk for genetic conditions. This site contains membership information and a searchable member directory, useful health resources, newsletters, and other publications.

**6. Family Village:**

<http://www.familyvillage.wisc.edu>

- This site integrates information and resources on the Internet for people with disabilities and their families. It includes informational resources about specific diagnoses, communication connections, adaptive products, technology, adaptive recreational activities, education, worship, health issues, and disability-related media and literature.

**7. Internet Resources for Special Children (IRSC):**

[http://orsaminore.dreamhosters.com/handy/links/uk\\_various.html](http://orsaminore.dreamhosters.com/handy/links/uk_various.html)

- IRSC is an online resource dedicated to children with disabilities and their families. Its website supports a directory of links to a variety of subject areas including cognitive disabilities, diseases, conditions and support/assistance.

**8. March of Dimes (MOD) – Before You're Pregnant\*:**

<http://www.marchofdimes.com/pnhec/173.asp>

- This webpage focuses on emotional and lifestyle issues before pregnancy, such as physically ready, weight-loss surgery and questions before pregnancy.

**9. Keep Kids Healthy:**

[http://www.keepkidshealthy.com/inside\\_pediatrics/family\\_history.html](http://www.keepkidshealthy.com/inside_pediatrics/family_history.html)

- This is a parent-focused site that contains a wealth of resources about family history in relation to diseases. It helps increase awareness of the role family history plays in both risk of disease and the promotion of health. The useful links direct visitors to information about family history as presented by the Centers for Disease Control and Prevention (CDC), the National Society of Genetic Counselors and the American Medical Association.

#### **10. Medline Plus – Pregnancy\*:**

<http://www.nlm.nih.gov/medlineplus/pregnancy.html>

- This helpful site provides information for women who are pregnant or who are planning to become pregnant about issues including staying healthy during pregnancy, prenatal care, nutrition, exercise and eating disorders.

#### **11. Our Kids:**

<http://www.our-kids.org>

- This website is designed to provide information and support for caregivers and family members with disabled children, as well as others who work with children with special needs. Its sections include organizational information, support staff and caregiver resources.

#### **12. Special Child:**

<http://www.specialchild.com>

- This link is dedicated to providing support and information to parents and caregivers of children with special needs. This website includes family issues, success stories, and useful information for parents and caregivers, including a section on tips for caregivers.

#### **13. The Arc of the United States:**

<http://www.thearc.org>

- This organization works to promote services and supports for people with mental retardation and other developmental disabilities. Its website offers fact sheets and other resources pertaining to disabilities, including FAS and other alcohol-related conditions.

#### **14. The Association of Maternal and Child Health Programs\*:**

<http://www.amchp.org/>

- This organization supports state maternal and child health programs and provides national leadership on issues affecting women and children. Under the Maternal and Child Health Topics link, there is information about weight and obesity, physical activity, and nutrition during pregnancy, as well as many other topics.

### **IV. Government Agencies**

#### **• CDC, National Center on Birth Defects and Developmental Disabilities:**

<http://www.cdc.gov/ncbddd/>

- This website provides information regarding various divisions, branches and offices in birth defects and developmental disabilities, as well as publications, health topics A-Z, employment opportunities and links to the CDC home page.

#### **• CDC Pregnancy Information\*:**

[http://www.cdc.gov/ncbddd/pregnancy\\_gateway/](http://www.cdc.gov/ncbddd/pregnancy_gateway/)

- This site provides information for before, during and after pregnancy. On the “Before Pregnancy” link, there is a Planning Section that includes topics such as preconception care and women’s health and a Preventing Problems Section that

focuses on various topics, including diabetes, infections, vaccinations, smoking, alcohol and hypertension.

- **CDC Pregnancy-Planning Education Program:**  
<http://www.cdc.gov/ncbddd/pregnancy/>
  - Learn what you need to know now to have a safe pregnancy and healthy baby with CDC's online education program available for women who are planning to become pregnant. By enrolling at [www.ihealthrecord.org](http://www.ihealthrecord.org), an interactive health record, you can receive health information via email every other week for three months as you prepare for pregnancy.
- **CDC Folic Acid Now:**  
<http://www.cdc.gov/ncbddd/folicacid>, <http://www.cdc.gov/ncbddd/folico/default.htm> (Spanish)
  - This site provides information about the importance of folic acid in the prevention of birth defects. Also featured are an online CDC folic acid publication order form, an FAQ section, folic acid fact sheets, and a quiz. The Spanish version features questions and publications sections for the Hispanic population.
- **CDC Science Ambassador Program:**  
<http://www.cdc.gov/excite/ScienceAmbassador/ScienceAmbassador.htm>
  - Follow the "Lesson Plans" link to "Standards Aligned Lesson Plans & Activities" to access a collection of health and science lesson plans. The lesson plans have been correlated to state and national standards and were developed to reach middle and high-school audiences. Topics include birth defects, neural tube defects, cystic fibrosis, alcohol and pregnancy, folic acid, cytogenetics, and epidemiology.
- **CDC Overweight and Obesity\*:**  
<http://www.cdc.gov/nccdphp/dnpa/obesity/resources.htm>
  - This website includes information for general obesity prevention (including women of childbearing age). Topics include BMI, diabetes, nutrition, physical activity, and weight loss programs.
- **CDC Nutrition for Everyone\*:**  
[http://www.cdc.gov/nccdphp/dnpa/nutrition/nutrition\\_for\\_everyone/healthy\\_weight/index.htm](http://www.cdc.gov/nccdphp/dnpa/nutrition/nutrition_for_everyone/healthy_weight/index.htm)
  - Learn about nutrition, exercise and lifestyle changes to maintain a healthy weight. This site is appropriate for everyone, including non-pregnant women of childbearing age. Follow the links to find information about nutrition basics, fruits and vegetables, the food pyramid, and weight control.
- **Education Resource Organization Directory:**  
[http://wdcrobcolp01.ed.gov/Programs/EROD/org\\_list.cfm?category\\_ID=SHE](http://wdcrobcolp01.ed.gov/Programs/EROD/org_list.cfm?category_ID=SHE)
  - This site lists the State Directors of Children with Special Health Care Needs under "Organizations by Type."

- **National Guideline Clearinghouse (NGC):**  
<http://www.guideline.gov>
  - NGC is a public resource for evidence-based clinical practice guidelines and an initiative of the Agency for Healthcare Research and Quality, U.S. Department of Health and Human Services. NGC's mission is to provide physicians, nurses, other health professionals, health-care providers, health plans, integrated delivery systems, purchasers and others with an accessible mechanism for obtaining objective, detailed information about clinical practice guidelines and to further their dissemination, implementation and use.
  
- **National Institutes of Health:**  
<http://www.nih.gov>
  - This website provides updates about the latest health and scientific research conducted by NIH. The "Health Information Index" has a subject-word guide to diseases and conditions under investigation at NIH. This site also provides access to online catalogs, journals and grant funding information.
  
- **New Jersey Department of Health and Senior Services:**  
<http://www.state.nj.us/health/>
  - This website provides information for women of childbearing age regarding preconceptional health and pre-pregnancy planning information.
  
- **Maryland Department of Health and Mental Hygiene:**  
<http://www.dhmh.state.md.us/>
  - This web site features information that women of childbearing age should consider before having a baby including the physical and emotional factors, work and home hazards, exercise and nutrition.
  
- **MMWR article, April 21, 2006-55(RR06) 1-23:**  
<http://www.cdc.gov/mmwr/preview/mmwrhtml/rr5506a1.htm>
  - This MMWR articles offers 10 recommendations made the CDC/ATSDR Preconception Care Group and Select Panel to improve preconceptional health and preconceptional health care in the United States. These recommendations include preconceptional doctor visits and providing additional care and interventions to those women who have been affected by adverse pregnancies and have identified health risks such as heart disease and diabetes.
  
- **Georgia Department of Human Resources:**  
<http://health.state.ga.us/programs/women/pregnancyplanning.asp>
  - Preconceptional health care information is provided on this website, including the importance of pregnancy planning and information for women 35 and older.

## V. Health-Care Professionals and Scientific Researchers

### 1. **American Academy of Pediatrics (AAP):**

<http://www.aap.org>

- This link provides online access to AAP's recommendations, research findings and policy statements, which convey helpful tips and health information for parents and providers on all child health topics.

### 2. **The American College of Obstetricians and Gynecologists\*:**

<http://www.acog.org/>

- The following are citations for the ACOG Committee Opinions, published in the Green journal:
  - *The overweight adolescent: prevention, treatment, and obstetric–gynecologic implications.* ACOG Committee Opinion No. 351. American College of Obstetricians and Gynecologists. *Obstet Gynecol* 2006;108:1337–48.
  - *Obesity in Pregnancy:* ACOG Committee Opinion No. 315. American College of Obstetricians and Gynecologists. *Obstet Gynecol* 2005;106:671-5.
  - *The Role of the Obstetrician-Gynecologist in the Assessment and Management of Obesity:* ACOG Committee Opinion No. 319. American College of Obstetricians and Gynecologists. *Obstet Gynecol* 2005;106:895-9.

### 3. **American Family Physician:**

<http://www.aafp.org/afp/20020615/2507.html>

- This website provides a comprehensive peer-reviewed cover article on preconceptional health care. It includes a patient handout on preconceptional health, a preconceptional health-care checklist, and information about various elements of preconceptional health, including promotion of folic acid, assessment of genetic risks, carrier screening by ethnicity, congenital infections, environmental toxins, risk of prescription drug use during pregnancy, management of chronic maternal illnesses during pregnancy, exercise and nutrition, and psychosocial factors.

### 4. **California Preconception Care Initiative:**

[http://www.marchofdimes.com/california/4949\\_8258.asp](http://www.marchofdimes.com/california/4949_8258.asp)

- MOD provides links to a variety of resources that have demonstrated success in increasing the understanding and use of preconceptional care strategies in California. This website includes an executive summary on the importance of seeking preconceptional care, fact sheets in English and Spanish about folic acid, diabetes and pregnancy, infections and immunizations, genetic counseling, domestic violence, smoking, and healthy lifestyle choices. There is also a fact sheet on billing codes for preconceptional care that have been used successfully in California.

### 5. **HuGE Net: The Human Genome Epidemiology Network:**

<http://www.cdc.gov/genomics/hugenet/default.htm>

- The HuGE Net represents a collaboration of individuals and organizations from diverse backgrounds that are committed to the development and dissemination of population-based human genome epidemiologic information. The website provides information about the HuGE Net and peer-reviewed synopses of

epidemiologic aspects of human genes, prevalence of allelic variants in different populations, population-based disease risk information, gene-environment interaction, and quantitative data on genetic tests and services.

**6. Institute for Child Health Policy:**

<http://www.ichp.ufl.edu/>

- The Institute for Child Health Policy, a statewide Institution of Florida's State University System, was established in October 1986. Given the substantial changes in both the financing and organization attendant to the growth of managed health care, the Institute has focused its attention on children in managed care with special emphasis on children with special health-care needs. Issues of access, utilization, cost, quality and family involvement are principal areas of interest for the policy/program development, health-services research, and evaluation programs.

**7. MOD: Genetics & Your Practice Curriculum:**

<http://www.marchofdimes.com/gyponline/index.bm2>

- This website includes a curriculum designed for health-care professionals and customized for those working with preconception/prenatal, infant/children, and adolescent/adult client types. It provides practical information and resources to assist busy professionals in integrating genetics into their patient care. A comprehensive array of information is provided under three topic areas: genetic testing and screening; family health and social history; and referral to genetic services.

**8. MOD: Preconception Curriculum:**

<http://www.mombaby.org/beforeandbeyond/>

- The Select Panel for Preconception Health and Health Care of the U.S. Centers for Disease Control and Prevention (CDC) has created a new website: Before, Between & Beyond Pregnancy. This website is designed for physicians providing health care to women who may become pregnant. Its curriculum includes a series of PowerPoint slide sets, a core module (which should be viewed first), and a separate module for each of the following four medical specialties: obstetrics/gynecology, internal medicine, family medicine and pediatrics. There are also presentation notes included, which make this an educational tool that may also be used as teaching presentations for residents, fellows, nurses and primary care practitioners in clinical conferences, didactic sessions or Grand Rounds.

**9. MOD: Weight Matters\*:**

[http://www.marchofdimes.com/professionals/14480\\_1930.asp](http://www.marchofdimes.com/professionals/14480_1930.asp)

- This site provides information for women who are pregnant or may become pregnant. It includes information about weight, nutrition and diet recommendations.

**10. The National Academies – Board on Children Youth and Families\*:**

<http://www.bocyf.org/053006.html>

- This site offers information from a May 2006 workshop on the Impact of Pregnancy Weight on Maternal & Child Health. Different presentations are available with topics including biological determinants of gestational weight gain,

maternal weight as a predictor of offspring health, and recommended practices and policies for clinicians.

**11. National Center for Biotechnology Information — Online Mendelian Inheritance in Man (OMIM):**

<http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=OMIM>

- This database is a catalog of human genes and genetic conditions and offers textual information, pictures and reference information. It also contains numerous links to NCBI's Entrez database of MEDLINE articles and sequence information.

**12. National Council on Folic Acid (NCFA):**

<http://www.folicacidinfo.org>

- NCFA is a partnership of more than 80 national organizations and associations, state folic acid councils, and government agencies whose mission is to improve health by promoting the benefits and consumption of folic acid. NCFA is managed by the National Healthy Mothers, Healthy Babies Coalition ([www.hmhb.org](http://www.hmhb.org)) through a cooperative agreement with CDC. NCFA's goals are to reduce folic-acid preventable birth defects by recommending that women of childbearing age take 400 micrograms of synthetic folic acid daily from fortified foods and/or supplements, in addition to consuming food folate from a varied diet; and to communicate and promote emerging and new science about folic acid, especially relating to maternal and child health.

**13. National Society of Genetic Counselors (NSGC):**

<http://www.nsgc.org>

- NSGC is the leading voice, authority and advocate for the genetic counseling profession. The "Consumer Information" section of its website explains the role of genetic counselors. The site also provides contact information for genetic counselors for those seeking a referral, both nationally and internationally.

**14. Perinatology.com:**

<http://perinatology.com>

- This site provides information for women who are pregnant or are planning to become pregnant. Topics include infections, medications, and radiation and chemical exposure. The site also serves as a resource for health-care professionals by providing abstracts from various peer-review journals that focus on perinatology.

**15. State University of New York, Albany, School of Public Health:**

<http://www.albany.edu/sph/coned/preconcept.htm>

- This website offers a free Women's Health Grand Rounds web cast on preconceptional care. The goal of this program is to improve health-care providers' knowledge, understanding and practice of preconceptional care as a means of improving pregnancy outcomes for women and children. The conference builds on a Preconceptional Care Curriculum developed for March of Dimes. Continuing education credits may be obtained after viewing the web cast and completing an evaluation. Other resources available on the site include a preconceptional screening and counseling checklist, as well as a set of PowerPoint slides that accompany the presentation.

**16. Organization for Teratology Information Services (OTIS):**

<http://www.otispregnancy.org>

- Site offers information about exposures to possible harmful substances during pregnancy. Contains a list of state contacts for teratology information, fact sheets, special projects and links.

**17. Surgeon General's Family History Initiative:**

<http://www.hhs.gov/familyhistory>

- Offers an online family history tool that the user may complete and download.

**18. Teratology Society:**

<http://www.teratology.org>

- The Teratology Society website provides information about teratology, the study of factors interfering with abnormal development and causing birth defects. It offers membership information and links to numerous birth defects sites. It also features a teratology discussion forum.

**19. Wisconsin Association for Perinatal Care:**

<http://www.perinatalweb.org/index.php>

- This association provides leadership and education for improved perinatal health outcomes of women, infants and their families. Online resources include the *Becoming a Parent* leaflet and parent booklet; a preconception checklist; preconceptional care position paper; information for professionals about screening for prenatal and postpartum depression; and fact sheets about women with asthma, depression, diabetes and epilepsy. The site also includes a position statement about the importance of folic acid. Additional publications include a *Health Care Provider's Guide to Preconceptional Risk Assessment* and a video, *Becoming a Parent: Things to Think About*.

## *North Dakota Century Code*

---

Available at [www.legis.nd.gov/cencode/t23c41.pdf](http://www.legis.nd.gov/cencode/t23c41.pdf).

**1. 23-41-04. Birth report of child with special health care needs made to department.**

Within three days after the birth in this state of a child born with a visible congenital deformity, the licensed maternity hospital or home in which the child was born, or the legally qualified physician or other person in attendance at the birth of the child outside of a maternity hospital, shall furnish the department a report concerning the child with the information required by the department.

**2. 23-41-05. Birth report of child with special health care needs - Use - Confidential.**

The information contained in the report furnished to the department under section 23-39-04 concerning a child with a visible congenital deformity may be used by the department for the care and treatment of the child pursuant to this chapter. The report is confidential and is solely for the use of the department in the performance of its duties. The report is not open to public inspection nor considered a public record.

## *Acknowledgements*

---

- Carmell Barth, North Dakota Department of Health, Division of Vital Records
- Janine Schoellhorn, Alaska Department of Health and Social Services
- Sue Burns, North Dakota Department of Health, Children's Special Health Services
- Tammy Gallup-Millner, North Dakota Department of Health, Children's Special Health Services

### *Technical Consultants*

- Mark Canfield, Ph.D., Texas Department of State Health Services
- Peter Langlois, Texas Department of State Health Services
- Russel Rickard, Colorado Department of Health
- Stephen Pickard, Ph.D., North Dakota Department of Health

## References (Text)

---

- (1) Amniotic Band. (2009 October 13). Retrieved December 4, 2009, from Wikipedia website: [http://en.wikipedia.org/wiki/Amniotic\\_band\\_syndrome](http://en.wikipedia.org/wiki/Amniotic_band_syndrome)
- (2) Anencephaly. (2009, March 11). Retrieved June 17, 2009, from Centers for Disease Control and Prevention website: <http://www.cdc.gov/ncbddd/birthdefects/Anencephaly.htm>
- (3) Atrial Septal Defect. (2009, March 11). Retrieved June 17, 2009, from Centers for Disease Control and Prevention website: <http://www.cdc.gov/ncbddd/birthdefects/AtrialSeptalDefect.htm>
- (4) Basic Facts About Birth Defects. (2005, October 5). Retrieved November 20, 2009, from Centers for Disease Control and Prevention website: <http://www.cdc.gov/ncbddd/bd/facts.htm>
- (5) Birth Defects Glossary: Gastrointestinal defects. Retrieved November 24, 2009, from California Birth Defects Monitoring Program website: <http://www.cbtmp.org/glossary.htm#g>
- (6) Birth defects states profile – North Dakota. (2009, January). Retrieved June 17, 2009, from National Birth Defects Prevention Network website: [http://www.nbdpn.org/current/2009pdf/StateProfiles/ND\\_2009.pdf](http://www.nbdpn.org/current/2009pdf/StateProfiles/ND_2009.pdf)
- (7) Canfield, M.A., et al. (2006). National Estimates and Race/Ethnic-Specific Variation of Selected Birth Defects in the United States, 1999–2001. *Birth Defects Research (Part A): Clinical and Molecular Teratology*, 76, 747-756.
- (8) Cleft Lip/Cleft Palate. (2009, March 11). Retrieved June 17, 2009, from Centers for Disease Control and Prevention website: <http://www.cdc.gov/ncbddd/bd/cleft.htm>
- (9) Down Syndrome. (2009, March 11). Retrieved June 17, 2009, from Centers for Disease Control and Prevention website: <http://www.cdc.gov/ncbddd/birthdefects/DownSyndrome.htm>
- (10) Endocardial Cushion Defects. (2007, January 23). Retrieved November 18, 2009, from Wikipedia website: [http://en.wikipedia.org/wiki/Endocardial\\_cushions](http://en.wikipedia.org/wiki/Endocardial_cushions)
- (11) Epispadias. (2009, November 24). Retrieved December 4, 2009, from Wikipedia website: <http://en.wikipedia.org/wiki/Epispadias>
- (12) Fetal Alcohol Spectrum Disorder. (2009, December 1). Retrieved December 2, 2009, from Wikipedia website: [http://en.wikipedia.org/wiki/Fetal\\_Alcohol\\_Spectrum\\_Disorder](http://en.wikipedia.org/wiki/Fetal_Alcohol_Spectrum_Disorder)
- (13) Glossary of Birth Defects Terms. (2006, September 28). Retrieved November 20, 2009. Texas Department of State Health Services. <http://www.dshs.state.tx.us/birthdefects/glossary.shtm>

- (14) Hydrocephalus. (2006). Retrieved June 10, 2009, from Lucina Foundation website: <http://www.lucinafoundation.org/birthdefects-hydrocephalus.html>
- (15) Hypoplastic Heart Syndrome. (2009, March 11). Retrieved June 17, 2009, from Centers for Disease Control and Prevention website: <http://www.cdc.gov/ncbddd/birthdefects/HypoplasticLeftHeartSyndrome.htm>
- (16) Hypospadias. (2009, December 1). Retrieved December 4, 2009, from Wikipedia website: <http://en.wikipedia.org/wiki/Hypospadias>
- (17) Orofacial. Retrieved November 20, 2009, from Utah Department of Health website: <http://health.utah.gov/birthdefect/defects/orofacial.html>
- (18) Patent ductus arteriosus (PDA). (2007, December 27). Retrieved June 17, 2009, from MayoClinic.com website: <http://www.mayoclinic.com/health/patent-ductusarteriosus/DS00631>
- (19) Percutaneous Aortic Valve Replacement. (2009, March 19). Retrieved June 18, 2009, from Mercy Angiography website: <http://www.mercyangiography.co.nz/PatientInfo/OurProcedures/PAVR.aspx>
- (20) Pyloric stenosis. (2008, August 23). Retrieved June 17, 2009, from MayoClinic.com website: <http://www.mayoclinic.com/health/pyloric-stenosis/DS00815>
- (21) Renal Agenesis/Hypoplasia. (2005, November). Retrieved June 17, 2009, from Minnesota Department of Health website: <http://www.health.state.mn.us/divs/fh/mcshn/bd/renal.htm>
- (22) Schoellhorn K. J., & Beery A.L. *Alaska Maternal and Child Health Data Book 2005: Birth Defects Surveillance Edition*. Anchorage, AK. Maternal and Child Health Epidemiology Unit, Section of Women's Children's and Family Health, Division of Public Health, Alaska Department of Health and Social Services, May 2006.
- (23) Spina Bifida. (2009, March 11). Retrieved June 17, 2009, from Centers for Disease Control and Prevention website: <http://www.cdc.gov/ncbddd/birthdefects/SpinaBifida.htm>
- (24) Tetralogy of Fallot. (2009, March 11). Retrieved June 17, 2009, from Centers for Disease Control and Prevention website: <http://www.cdc.gov/ncbddd/birthdefects/TetralogyOfFallot.htm>
- (25) Transportation of great arteries. (2009, March 11). Retrieved June 17, 2009, from Centers for Disease Control and Prevention website: <http://www.cdc.gov/ncbddd/birthdefects/TranspositionGreatArteries.htm>
- (26) U.S. Census Bureau, Census 2000 Summary File 1, Table GCT-PH1. Retrieved November 24, 2009, from American Factfinder website: <http://factfinder.census.gov/>

- (27) U.S. Census Bureau, Population Estimates Program, 2008 Population Estimates, Table GCT-T1-R. Retrieved November 24, 2009, from American Factfinder website:  
<http://factfinder.census.gov/>
  
- (28) Ventricular septal defect. (2009, March 11). Retrieved June 17, 2009, from Centers for Disease Control and Prevention website:  
<http://www.cdc.gov/ncbddd/birthdefects/VentricularSeptalDefect.htm>

## *References (Images)*

---

- (A) Anencephaly. (2009, March 11). Retrieved June 17, 2009, from Centers for Disease Control and Prevention website: <http://www.cdc.gov/ncbddd/birthdefects/Anencephaly-graphic.htm>
- (B) BrownCow. Patent Ductus Arteriosus. (2003, March 3). Retrieved November 9, 2009, from Wikipedia website: [http://en.wikipedia.org/wiki/File:Patent\\_ductus\\_arteriosus.jpg](http://en.wikipedia.org/wiki/File:Patent_ductus_arteriosus.jpg)
- (C) Capan, Manco. Atrial Septal Defect-en. (2008, February 21). Retrieved November 16, 2009, from Wikipedia website: [http://en.wikipedia.org/wiki/File:Atrial\\_septal\\_defect-en.png](http://en.wikipedia.org/wiki/File:Atrial_septal_defect-en.png)
- (D) Cleft Lip and Cleft Palate. (2009, April 13). Retrieved September 30, 2009 website: <http://www.cdc.gov/ncbddd/bd/cleft.htm>
- (E) Coarctation of Aorta. (2006, May 23). Retrieved November 9, 2009, from Wikipedia website: <http://en.wikipedia.org/wiki/File:Coarctation.png>
- (F) Endocardial Cushion. (2007, January 23). Retrieved November 9, 2009, from Wikipedia website: <http://en.wikipedia.org/wiki/File:Gray465.png>
- (G) Ewing, Edwin P. Jr. Aortic Valve Stenosis. (1973). Retrieved November 9, 2009, from Centers for Disease Control and Prevention's Public Health Image Library website: <http://phil.cdc.gov/phil/details.asp>
- (H) Fetal Alcohol Syndrome. (2005, February). Retrieved December 2, 2009, from Wikipedia website: <http://en.wikipedia.org/wiki/File:FASkid.gif>
- (I) Hypospadias and Epispadias. (2009, August 7). Retrieved November 9, 2009, from Wikipedia website: <http://en.wikipedia.org/wiki/File:Hypospadias3.JPG>
- (J) Kay, Rob. Down Syndrome. (2004, April 16). Retrieved November 18, 2009, from Wikipedia website: <http://en.wikipedia.org/wiki/File:Drill.jpg>
- (K) Monfils, Lucien. Hydrocephalus. (2002, July 22). Retrieved November 9, 2009, from Wikipedia website: <http://en.wikipedia.org/wiki/File:Hydrocephalus.jpg>
- (L) Pulmonary Valve Atresia. (2006, June 2). Retrieved November 18, 2009, from Wikipedia website: [http://en.wikipedia.org/wiki/File:Diagram\\_of\\_the\\_human\\_heart\\_\(cropped\).svg](http://en.wikipedia.org/wiki/File:Diagram_of_the_human_heart_(cropped).svg)
- (M) Pyloric Stenosis. (2006, December 1). Retrieved November 9, 2009, from Wikipedia website: <http://en.wikipedia.org/wiki/File:Gray1046.svg>
- (N) Ruiz, Mariana. Hypoplastic Left Heart Syndrome. (2006 June 12). Retrieved November 16, 2009, from Wikipedia website: [http://en.wikipedia.org/wiki/File:Hypoplastic\\_left\\_heart\\_syndrome.svg](http://en.wikipedia.org/wiki/File:Hypoplastic_left_heart_syndrome.svg)

- (O) Spina Bifida. (2009, March 11). Retrieved June 17, 2009, from Centers for Disease Control and Prevention website: <http://www.cdc.gov/ncbddd/birthdefects/SpinaBifida-graphic.htm>
- (P) Stillwell, Allison. Microcephalus. (2007, August 5). Retrieved November 16, 2009, from Wikipedia website: <http://en.wikipedia.org/wiki/File:IMGP2147.JPG>
- (Q) Tetralogy of Fallot. (2006, June 12). Retrieved November 18, 2009, from Wikipedia website: [http://en.wikipedia.org/wiki/File:Tetralogy\\_of\\_Fallot.svg](http://en.wikipedia.org/wiki/File:Tetralogy_of_Fallot.svg)
- (R) Ventricular Septal Defect. (2008, February 22). Retrieved December 7, 2009, from Wikipedia website: [http://en.wikipedia.org/wiki/File:Ventricular\\_septal\\_defect-en.png](http://en.wikipedia.org/wiki/File:Ventricular_septal_defect-en.png)