North Dakota
Birth Defects Monitoring System

Summary Report
1995 - 1999

North Dakota
Department of Health

north dakota
department of
human services

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Introduction

The goal of the North Dakota Birth Defects Monitoring System is to determine if children born with certain birth defects have access to needed health-care and other services.

Birth defects are the leading cause of infant mortality in the United States. In addition, birth defects are the fifth leading cause of years of potential life lost and contribute substantially to childhood illness and long-term disability. Because the cause of about 70 percent of all birth defects is unknown, evaluation of environmental, genetic, dietary and personal risk factors is needed to assess their possible contribution to birth defects.

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.

The NDBDMS tracks two neural tube defects, 17 different congenital heart defects and orofacial clefts. State rates per 10,000 births for the five-year period 1995 through 1999 were compared to rates from Utah and Colorado for the same time period. Ninety-five percent confidence intervals were calculated for state rates, and the data for each birth defect were graphed.

North Dakota rates for each birth defect, except for spina bifida, were found to be statistically similar to Utah and Colorado rates. Further analysis is planned to assess if North Dakota rates of spina bifida are actually higher than expected or if other explanations can be identified.

Methods

Because of the low numbers of both resident births and occurrences of individual birth defects in North Dakota, rates for each birth defect were averaged over five consecutive years. In this report, rates per 10,000 live births are based on identified birth defects for children born during the five-year period between 1995 and 1999.

Because there is no national birth defects monitoring system, national rates for birth defects are not available. In addition, not all states monitor birth defects, and those that do have differing procedures and methodology.

In this report, rates from the NDBDMS were compared to data collected by birth defects surveillance systems in two other selected states – Colorado and Utah. These states have well-established birth defects tracking systems, as well as demographic and geographic characteristics similar to North Dakota. The Colorado and Utah rates are taken from *Teratology: The Journal of Abnormal Development*, Volume 66, Supplement 1, 2002. Ninety-five percent confidence intervals were calculated for North Dakota rates. State rates were considered statistically similar to the comparison states if those rates fell within the upper and lower confidence limit for North Dakota rates.

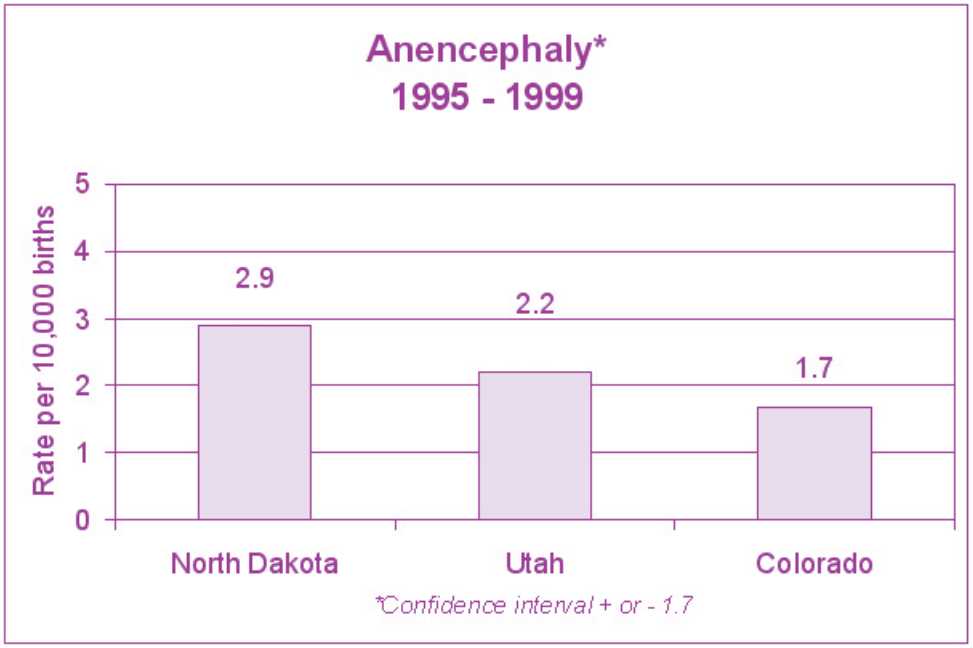
Neural Tube Defects

Neural tube defects are birth defects that result from the failure of the neural tube to close during the first month of pregnancy. The major conditions include anencephaly, spina bifida and encephalocele.

Anencephaly

Anencephaly is a congenital absence of the skull, with cerebral hemispheres completely missing or reduced to small masses attached to the base of the skull. Infants with anencephaly are unable to survive outside the womb and most are either stillborn or die shortly after birth.

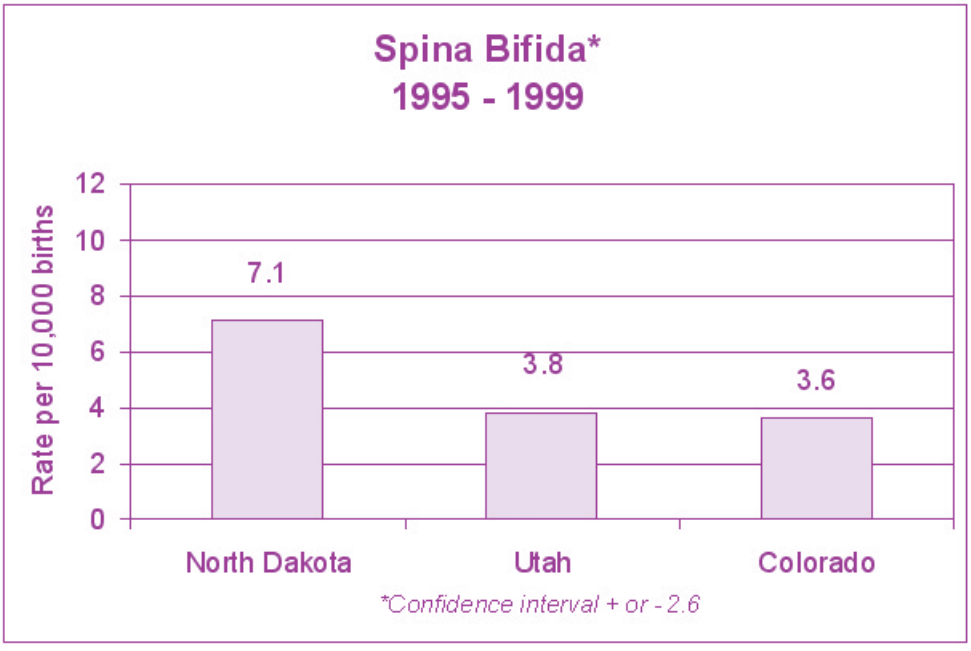
The five-year rates of anencephaly in North Dakota are statistically comparable to Utah and Colorado rates.



Spina Bifida

Spina bifida is a neural tube defect resulting from failure of the spinal neural tube to close. The spinal cord and/or meninges may or may not protrude. This usually results in damage to the spinal cord with paralysis of the involved limbs. Spina bifida includes myelomeningocele (involving both spinal cord and meninges) and meningocele (involving just the meninges).

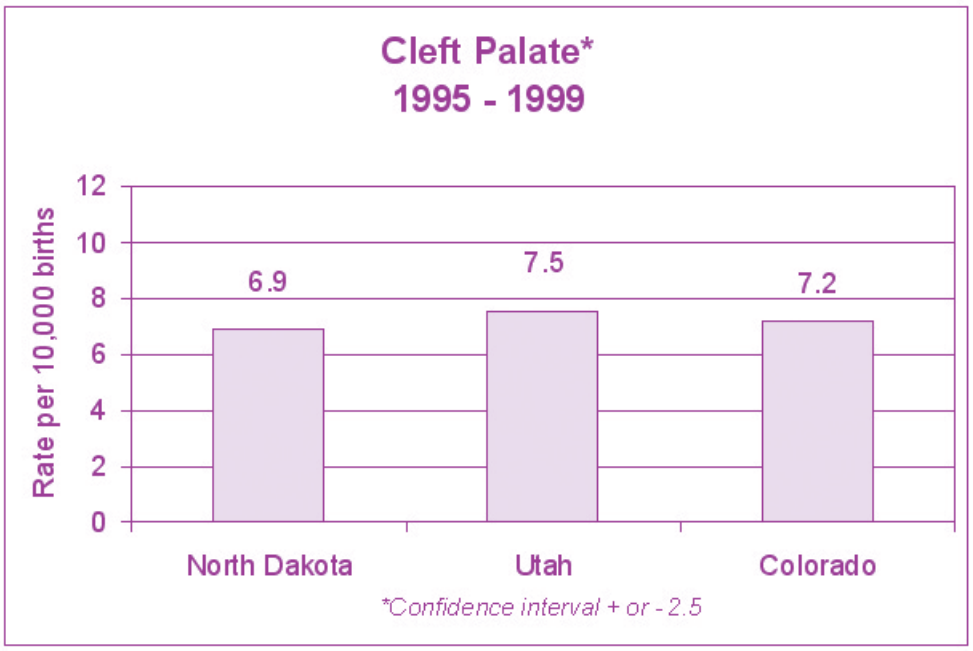
North Dakota five-year rates of spina bifida are significantly higher than comparison states despite a declining national trend since the folic acid awareness campaign was instituted in the mid-1990s. This rate is also higher than national rates reported for the same time period. Further analysis is needed to substantiate if spina bifda occurs more frequently in North Dakota.



Orofacial Clefts

Cleft Palate Only (Without Cleft Lip)

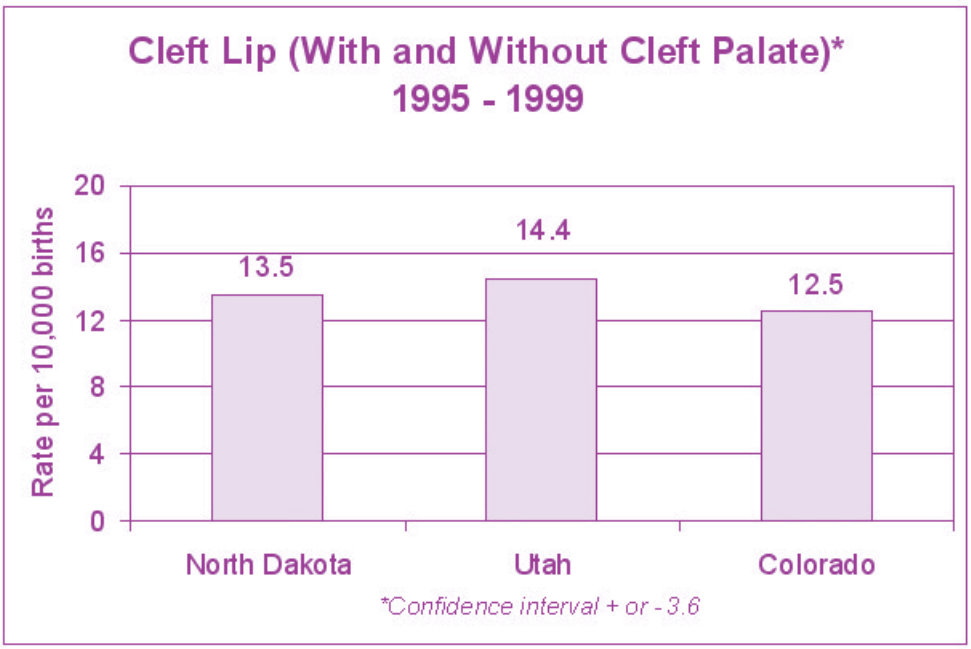
A cleft palate is the congenital failure of the palate to fuse properly, forming a grooved depression or fissure in the roof of the mouth. This defect varies in degree of severity. The fissure can extend into the hard and soft palate and into the nasal cavities. Infants with this condition have difficulty feeding. Surgical correction is begun when appropriate. Children who have cleft palates sometimes may have other kinds of birth defects, as well as hearing problems due to ear infections.



Cleft Lip (With and Without Cleft Palate)

A cleft lip is the congenital failure of the fetal components of the lip to fuse or join, forming a groove or fissure in the lip. Infants with this condition can have difficulty feeding. This condition is corrected when the infant can tolerate surgery.

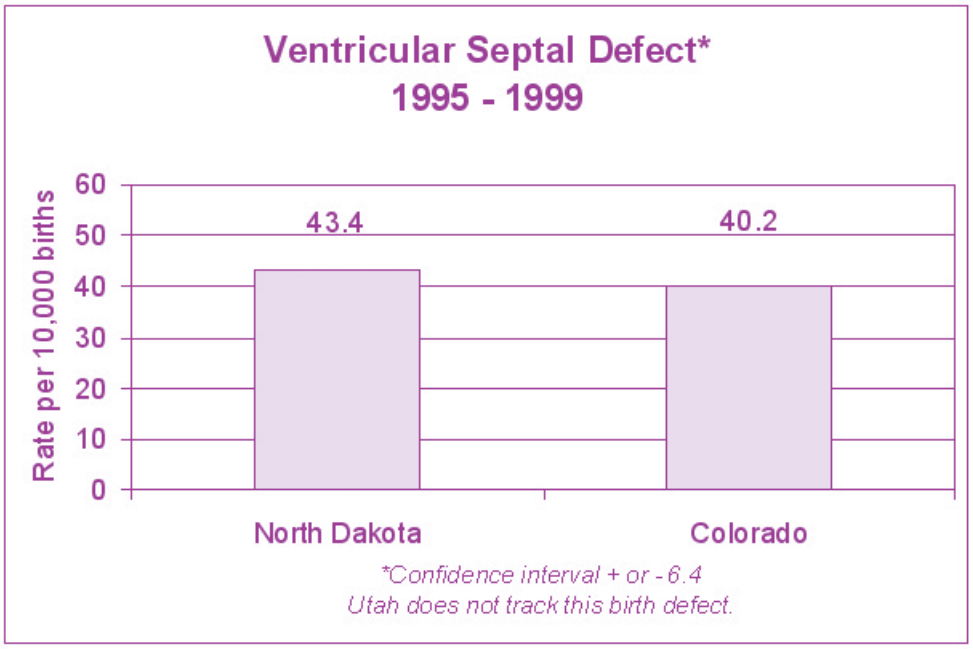
The prevalence of cleft palate (without cleft lip) and cleft lip (with and without cleft palate) in North Dakota children is statistically similar to rates for Utah and Colorado.



Congenital Heart Defects

Ventricular Septal Defects

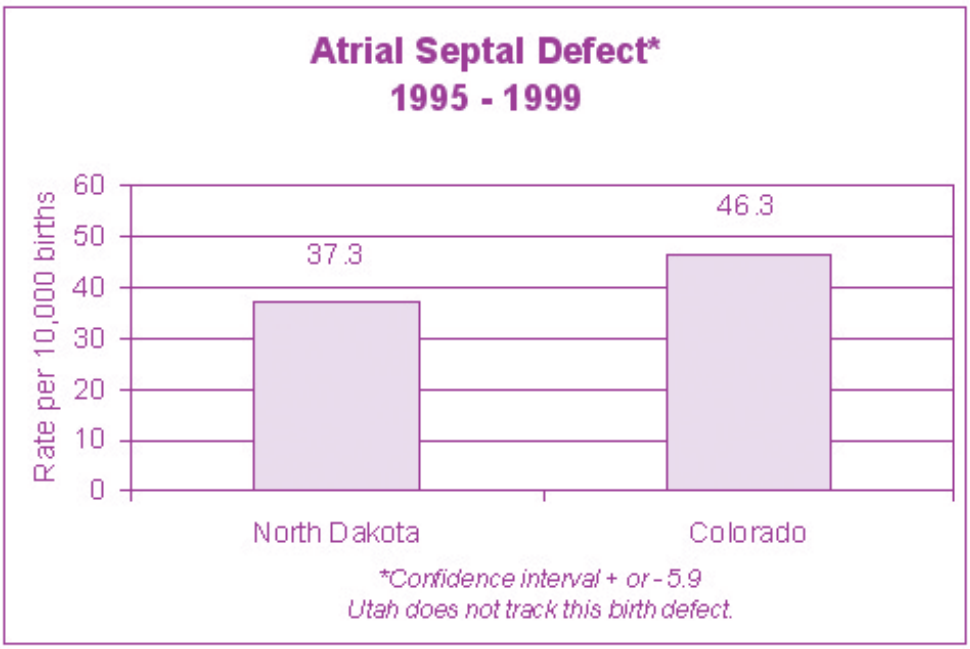
Ventricular septal defects (VSDs) are congenital cardiac malformations in which there are one or several openings in the ventricular septum (muscular and fibrous wall between the right and left ventricle or right and left lower chambers of the heart). These openings, which vary in size, allow oxygenated and unoxygenated blood to mix. The openings may resolve without treatment or may require surgery.



Atrial Septal Defects

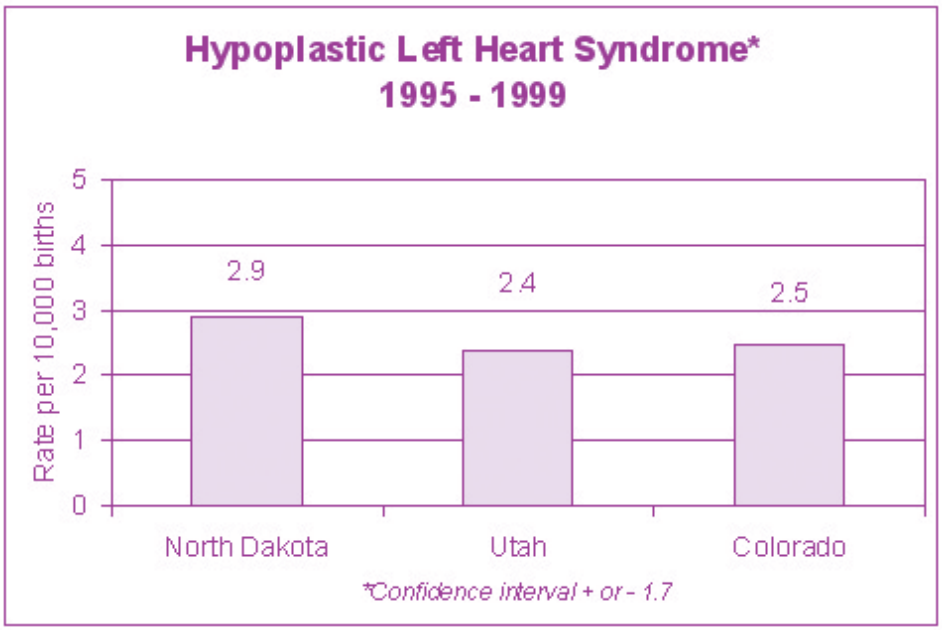
Atrial septal defects (ASDs) are congenital cardiac malformations in which there are one or several openings in the atrial septum (muscular and fibrous wall between the right and left atria). These openings, which vary in size, allow oxygenated and unoxygenated blood to mix. The openings may resolve without treatment or may require surgery. ASDs also are called ostium secundum defects.

Comparison of rates over time may be misleading. The 2002 Metropolitan Atlanta Congential Defects Program report indicated that "rates for VSD and ASD have risen in recent years mainly due to more widespread use of sophisticated technology to detect these defects." North Dakota rates of VSD and ASD are statistically similar to Colorado rates. Utah did not report VSD and ASD rates for 1995 through 1999.



Hypoplastic Left Heart Syndrome

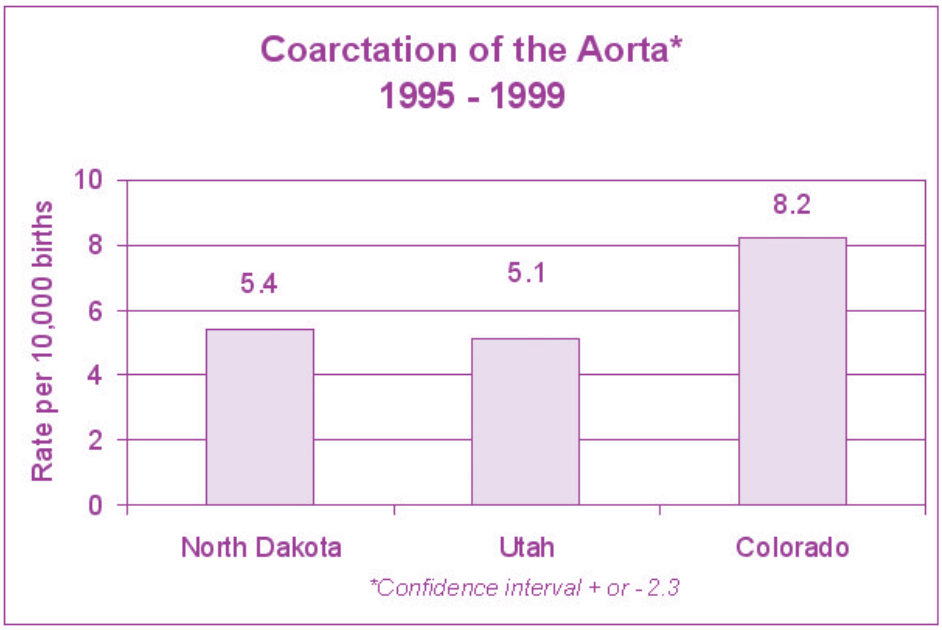
Hypoplastic left heart syndrome is an atresia, or marked hypoplasia, of the aortic opening or valve, with hypoplasia of the ascending aorta and defective development of the left ventricle (with mitral valve atresia). This condition can be repaired surgically in a series of three procedures over a period of one year. Transplantation is also a treatment. If not treated, this condition is usually fatal in the first month of life.



Coarctation of the Aorta

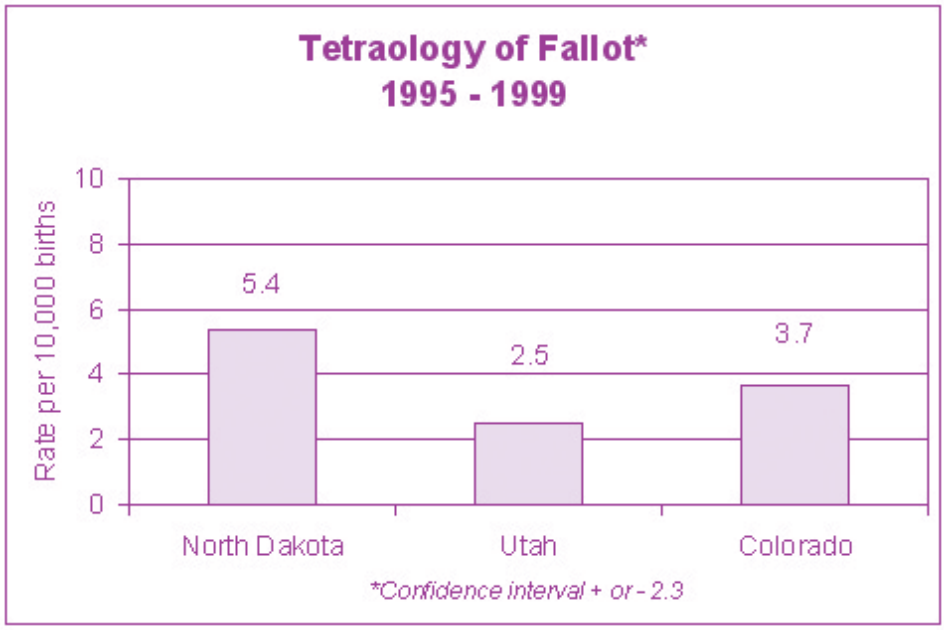
Coarctation of the aorta is a localized narrowing of the aorta. This condition causes abnormal cardiac circulation and pressure in the heart during contractions and varies from mild to severe. Surgical correction is recommended even for mild defects.

North Dakota rates of both hypoplastic left heart syndrome and coarctation of the aorta are statistically similar to rates for Utah and Colorado.



Tetralogy of Fallot

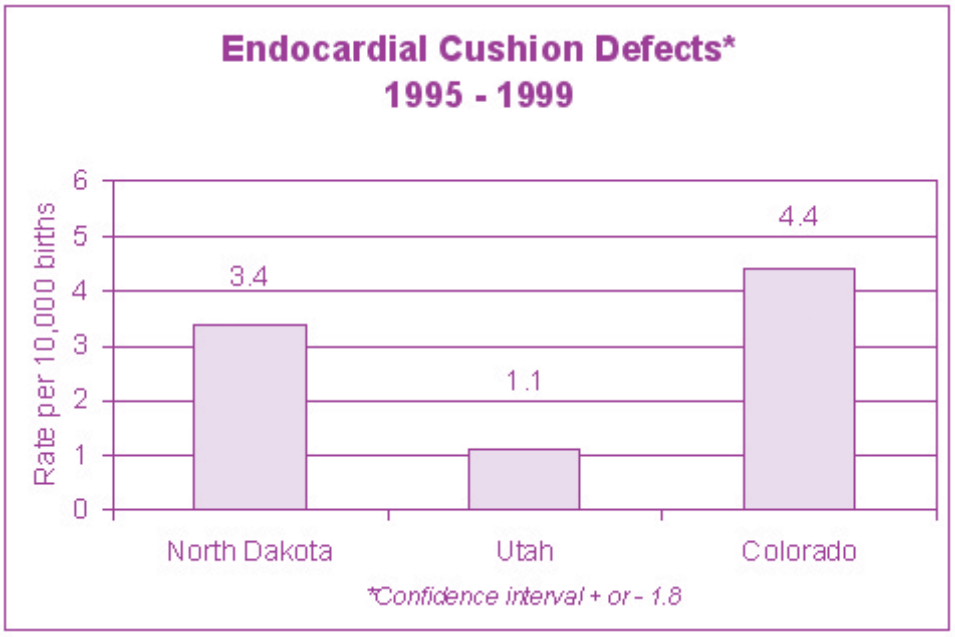
Tetralogy of Fallot is a congenital cardiac anomaly consisting of four defects: ventricular septal defect, pulmonary valve stenosis or atresia, displacement of the aorta to the right, and hypertrophy of the right ventricle. The condition is corrected surgically.



Endocardial Cushion Defect

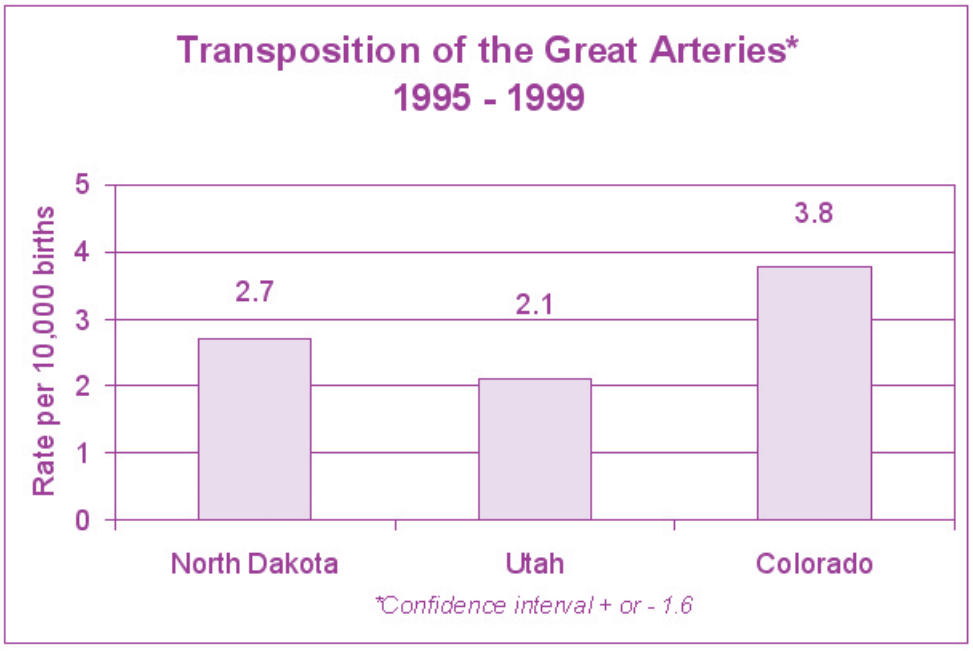
Endocardial cushion defects are a variety of septal defects (malformations of the walls separating the two atria and two ventricles of the heart) resulting from imperfect fusion of the endocardial cushions in the embryonic heart.

The North Dakota five-year rate of Tetralogy of Fallot and endocardial cushion defects is statistically similar to rates for Colorado and Utah.



Transposition of the Great Arteries/Vessels

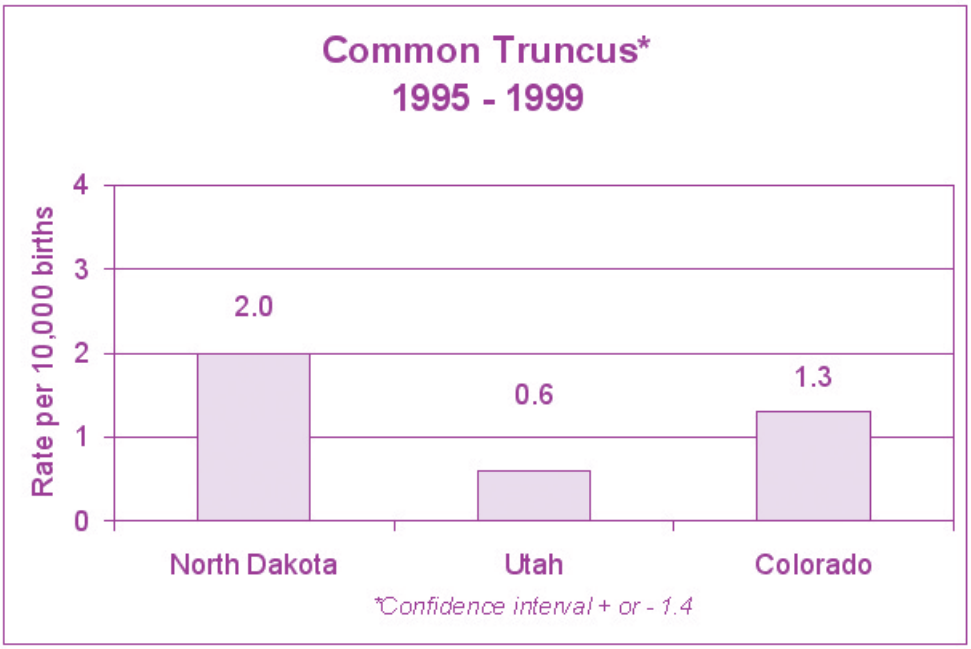
Transposition of the great arteries/vessels is a congenital malformation in which the aorta arises from the right ventricle and the pulmonary artery from the left ventricle (opposite of normal). As a result, the venous return from the peripheral circulation is recirculated without being oxygenated in the lungs. When this condition is not associated with other cardiac defects and not corrected, it is fatal.



Common Truncus

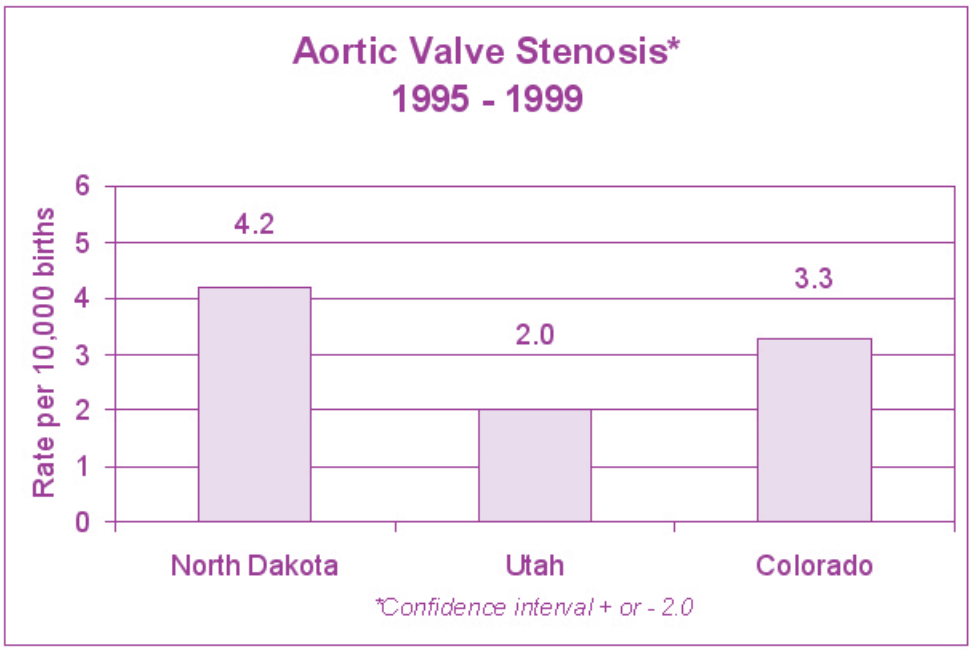
Common truncus ateriosus is a congenital heart defect in which the common arterial trunk fails to divide into the pulmonary artery and aorta. This is corrected surgically.

North Dakota rates of transposition of the great arteries and common truncus are similar to comparison states.



Aortic Valve Stenosis

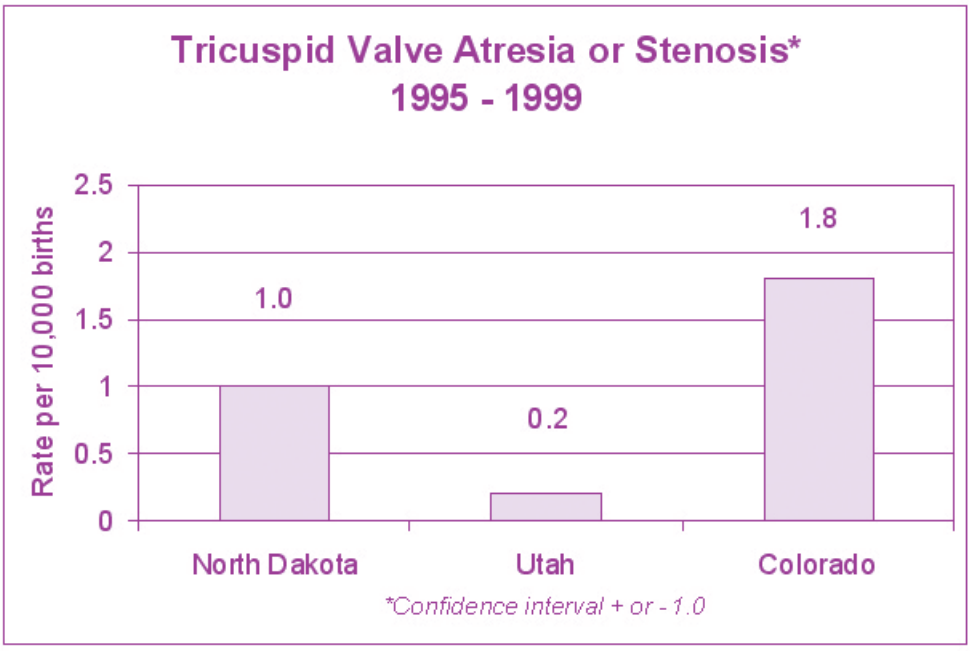
Aortic valve stenosis is a cardiac anomaly characterized by a narrowing or stricture of the aortic valve. This condition causes abnormal cardiac circulation and pressure in the heart during contractions.



Tricuspid Valve Atresia or Stenosis

Tricuspid valve atresia or stenosis is a congenital cardiac condition characterized by the absence or constriction of the tricuspid valve. The opening between the right atrium and the right ventricle is absent or restricted, and normal circulation is not possible. This condition often is associated with other cardiac defects and is corrected surgically depending upon the severity.

North Dakota rates of aortic valve stenosis and tricuspid valve atresia or stenosis are statistically similar to Utah and Colorado rates.



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 a 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 as 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 a woman 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.

Technical Notes

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 Human Services.

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 Human Services 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 1994 or later
- 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.

Resources

March of Dimes Birth Defects Foundation
North Dakota Chapter
1815 S. University Drive, Ste. C
Fargo, N.D. 58103-4941
Phone: 701.235.5530
Fax: 701.235.8725
E-mail: ND407@marchofdimes.com

Children's Special Health Services
North Dakota Department of Human Services
600 E. Boulevard Ave., Dept. 325
Bismarck, N.D. 58505-0269
Phone: 701.328.2436
Toll-free: 800.755.2714 (in-state)
TTY: 701.328.3480
Fax: 701.328.1645
E-mail: dhscshs@state.nd.us

North Dakota Genetics Program
Department of Pediatrics
University of North Dakota School of Medicine and Health Sciences
P.O. Box 9037
Grand Forks, N.D. 58202-9037
Phone: 701.777.4243

North Dakota Family to Family Network
Center for Rural Health
University of North Dakota School of Medicine and Health Sciences
501 N. Columbia Road
P.O. Box 9037
Grand Forks, N.D. 58202-9037
Phone: 701.777.2359 or 888.434.7436
Fax: 701.777.2353
E -mail: NDF2F@medicine.nodak.edu

Family Voices of North Dakota, Inc.
P.O. Box 163
Edgley, N.D. 58433
Phone: 701.493.2634
Toll-free: 888.522.9654
Fax: 701.493.2635
E-Mail: feist@daktel.com

Sources

- Metropolitan Atlanta Congenital Defects Program, 2002
- National Birth Defects Prevention Network: www.nbdpn.org
- National Center of Birth Defects and Developmental Disabilities: www.cdc.gov/ncbddd
- North Carolina Birth Defects Monitoring Program Surveillance Report, October 2000
- *Teratology: The Journal of Abnormal Development,* Volume 66, Supplement 1, 2002

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