Nevada Birth Outcomes Monitoring System
Second Biennial Report
(2005-2009 Data)

Department of Health and Human Services
Division of Public and Behavioral Health
Bureau of Health Statistics, Planning, and Emergency Response
Office of Public Health Informatics and Epidemiology

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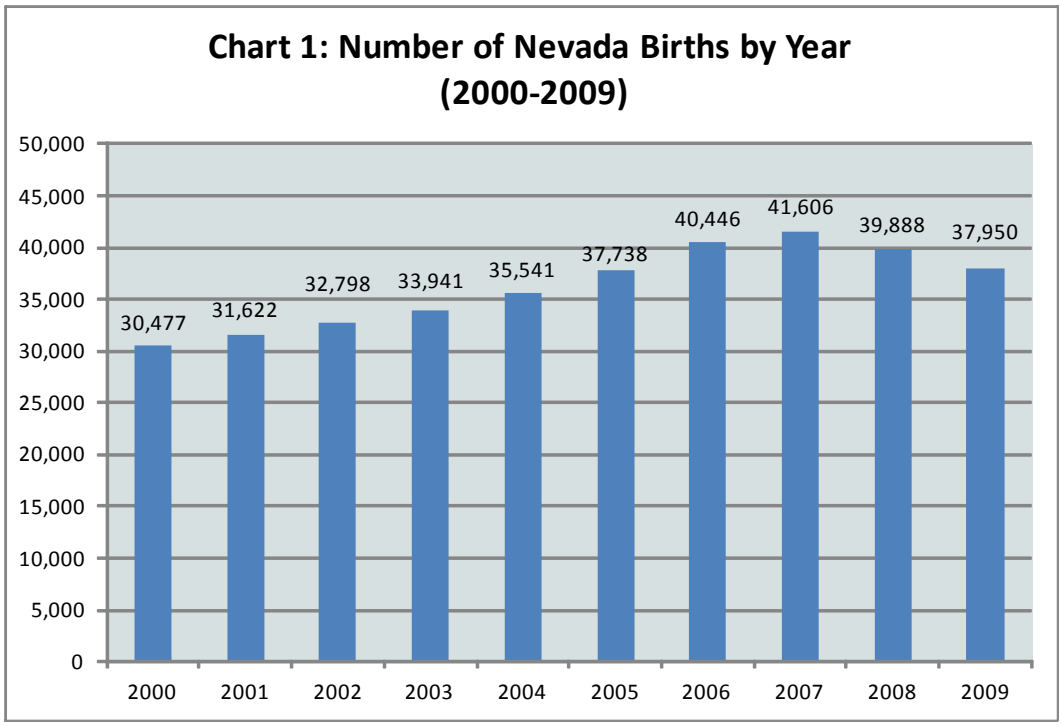


Nevada Demographics:

Nevada is the seventh largest state (geographically) in the nation and is comprised of 17 counties spread across 110,540 square miles. Nevada is a frontier state with a 2009 population estimate of 2.7 million (Nevada State Demographer) and is traditionally divided into three regions: Clark County (70.7% of the population), Washoe County (15.2% of the population), and the balance of state (14.1% of the population). Approximately 83% of Nevada's land area is under the jurisdiction of the Bureau of Land Management; the remaining 17% is under private ownership or state and local jurisdiction. Nevada has 13 Native American colonies or reservations statewide, and six military bases located in five counties.

From 1990 to 2000, Nevada had a population growth of 64% and was the fastest growing state in the nation for 19 straight years until surpassed by Arizona in 2006. In the ten year period from 2000 to 2009, it is estimated that Nevada had a 21.2% increase in population, Clark County a 28.6% increase, Washoe County a 18.0% increase, and the balance of state a 17.9% increase. Chart 1 details the increase in the Nevada birth rates from 2000 to 2009. Nevada's population is becoming more racially diverse. The American Indian, Eskimo, or Aleut population increased 29.9% in the ten-year period from 2000 to 2009, the Asian/ Pacific Islander population increased 66.4%, the Black population 39.5%, Hispanics 74.5% and whites 21.6%.

Program Overview

Authority

The authority for the Nevada Birth Outcome Monitoring System (NBOMS; formerly the Birth Defect Registry) is Nevada Revised Statutes (NRS) 442.320 mandating that, "The Health Division, in cooperation with the University of Nevada, School of Medicine, establish and maintain a statewide system for the collection and analysis of information concerning birth defects and other adverse birth outcomes."

Data Collection

Hospital information is gathered for newborns or patients who are under seven years of age and have been diagnosed with one or more birth defects. A letter is sent to the parent/guardian of each child identified with an adverse birth outcome. Parents are given the option to remove their child's name from NBOMS; however, other information will remain in the registry. Parents are advised that access to information contained in the system is limited to persons authorized by the Chief Medical Officer or the University School of Medicine, and that any information revealing the identity of a patient will remain confidential.

This information is to be collected and used only to "investigate the causes of birth defects and other adverse birth outcomes; determine, evaluate, and develop strategies to prevent the occurrence of birth defects and other adverse birth outcomes; assist in the early detection of birth defect cases and; assist in ensuring delivery of services for children identified with birth defects (NRS 442.330)."

NBOMS utilizes an "active surveillance" system to track adverse birth outcomes in Nevada. This means that hospital staff and medical professionals are not required to report birth defects, but that trained program staff regularly visit medical facilities to review records and gather data. Although authorization for this program was enacted in 1999, funds were not available to staff a full-time program position until state fiscal year (SFY) 2004. Funding was made available through the increase of birth registration fees, passed by the State Board of Health in SFY 2004. Complete data is now available for calendar year 2005, 2006, 2007, 2008, and 2009; incidence rates included in this report are based on 2005 through 2009 pooled data.

NBOMS funds one full time program person responsible for data retrieval and collection. This person maintains communication with Nevada birthing facilities and periodically receives a list of patients and births with birth defects and their related International Classification of Diseases - Clinical Modification codes (ICD-9-CM codes) . The Program Specialist reviews facility lists for eligible ICD-9-CM codes and visits each birthing facility to gather pertinent patient information from hospital records. Information reviewed includes admission records, discharge summaries, available laboratory test results, physician/nurse notes, and birth/death certificates. These data are then cross-matched with birth registration data to gather prenatal care and high risk behavior information.

Program Overview

Eligible Conditions

A birth defect is defined in NRS 442.310 as "...any structural or chemical abnormality present in a child at birth." Abnormalities due to birth complications and prematurity are also included in NBOMS. Hospital admissions are tracked for children born with birth anomalies up to seven years of age to ensure they receive needed services. Nevada's NBOMS uses criteria provided by The National Birth Defects Prevention Network for inclusion in a birth defects surveillance system. These criteria are as follows:

- Abnormal structural and chromosomal conditions identified from birth to the seventh birth date
- Born after completion of 20 weeks of gestation
- Fall under Centers for Disease Control and Prevention (CDC) and the National Birth Defect Prevention Network (NBDPN) guidelines (appropriate ICD-9-CM codes; Appendix C)
- Abortion at any stage of gestation
- Abnormalities/injuries during labor (birth complications)
- Born in Nevada
- Parents reside in the State of Nevada during the gestation period

Availability of Comparable National Data

National data is not available for comparison with Nevada's birth defect incidence rates. CDC published an article in January of 2006 using 1999 to 2001 data averaging incidence rates for 18 states (Morbidity and Mortality Weekly Report, 6 Jan. 2006). Incidence rates reported in the *2008 Congenital Malformations Surveillance Report* were determined from 2001 to 2005 data. Comparison of birth defect data is also compounded by the variety of programs across the states. Some states have passive surveillance systems that depend on reporting from external sources; therefore, this data is often less comprehensive than states, such as Nevada, utilizing active surveillance programs. Nevada program staff review hospital records and collect data from essentially all eligible cases statewide. Because of the low numbers of birth defects, the National Birth Defects Prevention Network analyzes data in five year increments to produce more reliable incidence rates. This report pools five years of birth outcome data (2005 - 2009).

Birth Defect Prevention

Although the causes of most birth defects are unknown, there are preventive measures known to reduce the risk of some birth defects. These measures promote healthy lifestyles and are much more important to the health status of the mother and infant than monitoring birth defect incidence rates. Birth defects can develop during any stage of pregnancy; however, most birth defects occur within the first three months. Any abnormalities during the first three months can cause irregular fetal growth for the remainder of the pregnancy. It is important that women of child bearing age improve and maintain their health because birth defects may occur before a woman discovers she is pregnant; planned pregnancies have a reduced risk of birth defects. It is important that women have a preconception visit with their health care provider to treat health conditions that can pose a risk in pregnancy such as diabetes or high blood pressure. The doctor can also ensure that any medications a woman takes are safe during pregnancy.

Folic Acid

Folic acid is a B vitamin that women need every day for their bodies to make healthy new cells, whether they are pregnant or not. A woman planning to become pregnant should begin taking folic acid on a daily basis as a preventative measure against birth defects. The CDC recommend a daily intake of 400 micrograms of folic acid which can reduce the risk of neural tube defects by approximately 80%. Neural tube defects are defects of the brain or spine such as spina bifida or anencephaly. Folic acid can be taken in a multivitamin or as a supplement and comes in chewable and liquid forms as well as pills. A single serving of many breakfast cereals contain the daily recommended amount of folic acid.

Fetal Alcohol Spectrum Disorders

Prenatal exposure to alcohol can cause a range of disorders (fetal alcohol spectrum disorders or FASD). Fetal alcohol syndrome (FAS) is one of the most severe disorders caused by drinking during pregnancy. FAS is one of the leading preventable causes of mental retardation and birth defects. It is recommended that a woman abstain from drinking alcohol at any time during pregnancy. There is no known safe amount of alcohol to drink while pregnant and no safe time to drink during pregnancy. Women who are sexually active and not using effective birth control should refrain from drinking because they could become pregnant and not know it for several weeks. If a woman is drinking during pregnancy, the sooner she stops the better it will be for the baby and herself.

Lymphocytic Choriomeningitis Virus (LCMV)

Lymphocytic choriomeningitis virus is carried by wild mice and can spread to household pets such as hamsters, guinea pigs, and other pet rodents through contact with wild mice. Humans can be infected with LCMV through contact with urine, blood, saliva, droppings or nesting materials of infected rodents. LCMV infection can also be contracted by inhaling dust while sweeping infected rodent droppings. Approximately 5% of adults have been infected by LCMV at some time during their lives. Some individuals show no symptoms when infected while others exhibit mild symptoms such as headache, fever chills, and muscle aches. Sometimes meningitis will occur. Should a pregnant mother contract LCMV, the infection may be passed on to the unborn child, resulting in miscarriage or severe birth defects.

Birth Defect Prevention

Diabetes

Individuals with diabetes cannot properly use sugars and starches. This extra sugar collects in the blood, and can damage body organs such as the heart, eyes, and kidneys, if allowed to collect in the body too long. Three of the most common types of diabetes are Type I, Type II, and Gestational diabetes. Type I diabetes occurs when the pancreas does not make enough insulin and the body cannot utilize blood sugar; this type is treated with daily insulin shots. Type II diabetes occurs when body makes too little insulin or cannot use the insulin it makes. Type II diabetes can often be controlled by proper diet and exercise, but may also require medication in pill form, insulin or both. Gestational diabetes occurs in pregnant women who previously did not have diabetes. Often gestational diabetes can be controlled by proper diet and exercise, but in some cases insulin may be necessary. Often gestational diabetes goes away after pregnancy, but women with gestational diabetes may develop Type II diabetes later in life.

A woman who has Type I or Type II diabetes is at greater risk of having a baby with a birth defect than a woman without diabetes. The baby's organs are formed during the first two months of pregnancy, often before a woman knows she is pregnant; therefore, high blood sugar can cause serious birth defects such as those of the brain, spine, heart, or can lead to miscarriage of the developing baby. A woman with gestational diabetes has less chance of having a child with birth defects than a woman with Type I or Type II diabetes because gestational diabetes develops later in pregnancy. It is still important that a woman with gestational diabetes keep her blood sugar in control, because like those with Type I or Type II diabetes, she could have an extra large baby which could cause problems during the delivery. In addition, out-of-control blood sugar can result in high blood pressure which could lead to an early delivery and cause seizures or strokes in the mother during delivery. A pregnant woman with diabetes needs to see her doctor more often than a pregnant woman without diabetes. A pregnant woman with diabetes who keeps her blood sugar in control is more likely to have a healthy pregnancy and healthy baby.

Toxoplasmosis

Toxoplasmosis is an infection caused by a parasite (*Toxoplasma gondii*) and can be spread by cat feces; cats are infected by eating infected rodents. A pregnant woman can also be infected with toxoplasmosis by eating unwashed or unpeeled fruits and vegetables, eating under-cooked meat, handling raw meat, contaminated utensils or cutting boards, or drinking contaminated water. Most people infected with toxoplasmosis have no symptoms, but some people may feel like they have the flu with swollen lymph glands and muscle aches that last for a month or more. Severe toxoplasmosis cases can cause damage to the brain, eyes, or other organs. Most infants who are infected while still in the womb have no symptoms at birth. However, they may develop symptoms later in life such as blindness or mental retardation. A small percentage have serious eye or brain damage at birth. A woman can protect her unborn baby from toxoplasmosis by:

Birth Defect Prevention

- Avoiding changing cat litter.
- Washing hands with soap and water after exposure to soil, sand, raw meat, or unwashed vegetables.
- Cooking meat thoroughly to an internal temperature of 160^{o} with no pink in the center.
- Freezing meat for several days before cooking.
- Washing all cutting boards and knives thoroughly with soap and water.
- Washing and peeling all fruits and vegetables before eating them.
- Wearing gloves while gardening and handling sand from a sandbox and washing hands well afterwards.
- Avoiding drinking untreated water.

Phenylketonuria

Phenylketonuria (PKU) is a genetic disorder that is characterized by an inability of the body to breakdown and use phenylalanine (an essential amino acid). In classic PKU, the enzyme needed to break down phenylalanine (phenylalanine hydroxylase) is partly or entirely deficient. Without treatment, phenylalanine builds up in the bloodstream causing mental retardation. PKU occurs in approximately 1 out of 15,000 infants born in the United States. There are an estimated 3,000 to 4,000 women of childbearing age with successfully treated PKU in the United States. When women with PKU do not strictly adhere to their diet, restricting the intake of protein-containing foods, infants born to them have a 93% risk of mental retardation and a 72% risk of microcephaly. These risks result from the toxic effect of the mother's blood, not because the infant has PKU. Although women with PKU should maintain their special diet for life, it is often discontinued during adolescence. Women with PKU need to resume their special diets at least three months before pregnancy and continue the diet throughout the pregnancy. The Maternal PKU International Study found that women who keep their phenylalanine levels under control before conception or by 8 to 10 weeks of pregnancy at the latest, were as likely to have healthy babies as women without PKU.

Medications and Pregnancy

Certain medications such as thalidomide, isotretinoin (Accutane©) taken during pregnancy may cause serious birth defects in the baby and should be avoided by all pregnant women. Little is known about the safety of many other drugs. Some pregnant women must take medications to treat existing conditions such as epilepsy, asthma, or high blood pressure and it is important to know which ones are safe. The March of Dimes recommends that pregnant women:

- Do not take street drugs.
- Do not take someone else's prescription drugs.
- Take only medications prescribed for you by your health care provider who know you are pregnant.
- Check with your provider before taking any over-the-counter drugs, including aspirin.

Birth Defect Prevention

Mother's Age and Chromosomal Errors

The chances of a woman having a child with a chromosomal inheritance error increases throughout the childbearing years. Down syndrome, caused by an extra 21st chromosome, causes mental retardation and physical abnormalities and often includes heart defects. Prenatal tests are available (chorionic villus sampling or amniocentesis) that can rule out chromosomal error with a high degree of certainty. Women in their twenties who become pregnant have about a one in 1,230 chance of having a child affected by Down syndrome and by the age 40 the chance increases to one in 78. Parents who have had a child with a chromosomal abnormality should consult a genetic counselor to learn more about the causes of these birth defects and the chances that it could recur in another pregnancy.

Caffeine and Pregnancy

Caffeine is a stimulant that is naturally produced by a variety of plants. During pregnancy caffeine crosses the placenta barrier and reaches the fetus. Caffeine may decrease the blood flow to the placenta. Studies on caffeine and miscarriage have produced conflicting reports, but to be on the safe side, the March of Dimes recommends that pregnant women limit caffeine consumption to less than 200 milligrams per day; this is approximately equivalent to one 12-ounce cup of coffee. Some studies also suggest that high levels of caffeine consumption (more than 500 milligrams per day) may slightly reduce the baby's birth weight and that the baby may be more likely to have faster breathing and heart rates and sleep less during the first few days of life. The source of caffeine does not matter; the risk appears to be the same (coffee, tea, chocolate). Some medications and herbal products also contain caffeine and should be avoided.

Tobacco and Pregnancy

In addition to an increase risk of lung cancer, smoking by pregnant women can increase the risk of adverse birth outcomes. Cigarette smoke contains more than 2,500 chemicals. Of these chemicals, nicotine and carbon monoxide are known to contribute to adverse birth outcomes. Some of these adverse birth outcomes are listed in the 2004 Surgeon Generals report, *The Health Consequences of Smoking*. Women who smoke have 30% greater odds of being infertile. Smoking during pregnancy can cause the placenta to separate from the uterine wall (placental abruption) or cause the placenta to attach low on the uterine wall (placenta previa). Although placenta previa is not common, it can become serious causing heavy bleeding during pregnancy or labor. The nicotine in cigarettes reduces the amount of oxygen reaching the fetus and can reduce fetal growth. Women who smoke during pregnancy have babies with lower birth weights, often weighing less than 5.5 pounds. Women who are exposed to second hand smoke have a 20% higher odds of giving birth to a low birth weight baby than women who are not exposed to secondhand smoke. These low birth weight babies are at greater risk for childhood and adult illnesses and even death. In addition, babies whose mothers smoke before and after birth are three to four times more likely to die from SIDS. Infants exposed to secondhand smoke after birth have double the risk of sudden infant death syndrome (SIDS).

Birth Defect Prevention

Having a Healthy Pregnancy

Approximately one out of 33 babies born in the United States is born with a birth defect. Many babies born with birth defects suffer from illnesses, lifelong disabilities and social challenges. While not all birth defects can be prevented, women can take actions that significantly increase their chances of having a healthy baby. Because about half of all pregnancies are unplanned, it is important that women of childbearing age maintain healthy lifestyles. A baby's brain and spine is formed before most mothers know they are pregnant. The following bullets summarize some of the recommendations made by the CDC to increase chances of having a healthy pregnancy and a healthy baby.

- Avoid exposure to toxic substances and chemicals, such as cleaning solvents, lead and mercury, some insecticides, and paint.
- Be sure to see a doctor for prenatal care as soon as you think you are pregnant. It is important to see your doctor regularly throughout pregnancy, so be sure to keep all your prenatal care appointments.
- Do not smoke because smoking during pregnancy increases the chances of premature birth, certain birth defects, and infant death.
- Eat a healthy diet to get the nutrients you and your unborn baby need.
- Take 400 micrograms of folic acid daily, before and during the first few months of pregnancy to reduce the risk of birth defects of the brain and spine.
- Wash hands throughout the day, especially after handling raw meat or using the bathroom. This can prevent the spread of many bacteria and viruses that cause infection.
- Avoid eating undercooked meat and handling cat litter.
- Be sure to wear gloves while gardening.
- Take 30 milligrams of iron daily during your pregnancy as prescribed by your doctor to reduce the risk of anemia later in pregnancy. All women of childbearing age should eat a diet rich in iron.
- Join a support group for moms-to-be or take a class on parenting and childbirth.
- Avoid legal drugs such as alcohol and caffeine. There is no known safe amount of alcohol a woman can drink during pregnancy. Caffeine found in tea, coffee, soft drinks, and chocolate should be limited.
- Treat medical conditions such as diabetes, epilepsy, and high blood pressure.
- Ask your health care provider about prescription or over-the-counter drugs that you are taking or considering taking while pregnant because over-the-counter cough and cold remedies may contain alcohol or other ingredients that should be avoided during pregnancy.
- Avoid saunas, hot tubs, and steam rooms while you are pregnant; excessively high heat may be harmful during pregnancy.
- Get needed vaccinations before pregnancy. Vaccinations are an important concern for pregnant women. CDC has clear guidelines for the use of vaccines during pregnancy. Be sure to review the list and discuss this with your doctor.
- Avoid X rays. If you need dental work or diagnostic tests, tell your dentist or physician that you are pregnant so that extra precautions can be taken.

Report Layout

Report Layout

In this report, birth anomalies are grouped by CDC categories. Two tables are provided for each category. The first table provides Nevada incidence rate for various disorders by CDC category and the second provides unduplicated demographic information by category for Nevada. When available, national incidence rate estimates for various disorders are included in the text. Caution must be taken when comparing state and national incidence rates because national estimates may vary widely. Five years of Nevada data have been collected, so meaningful incidence rates have been calculated from the pooled number of cases (N) for the years 2005-2009. The demographic table details the percent of statewide cases per region (Clark County, Washoe County, and balance-of-state); the percent of statewide cases by sex; the percent of statewide cases by mother's age group (17 and younger, 18 to 24, 25 to 34, 35 to 44, 45 and older) and; the percent of cases statewide by type of birth (Caesarian section, C-section repeat, or vaginal). A brief explanation of common or serious birth defects is also included for each category. Appendix A provides a list of references utilized in this report. Appendix B provides a table of statistics on all birth anomalies in CDC categories (2005 through 2009). Birth anomalies are presented in the following categories: cardiovascular/respiratory, central nervous system, chromosomal, ear/eye, gastrointestinal, genitourinary, musculoskeletal, and orofacial.

Cardiovascular/Respiratory Anomalies

Table 1: Nevada Incidence Rates of Cardiovascular Anomalies (2005 Through 2009 Pooled Data)

Cardiovascular Anomalies	Cases	Rate per 10,000 Births
Aortic Valve Stenosis	41	2.1
Atrial Septal Defect	1,349	69.0
Coarctation of Aorta	118	6.0
Common Truncus	18	0.9
Ebstein's Anomaly	19	1.0
Endocardial Cushion Defect	56	2.9
Hypoplastic Left Heart Syndrome	45	2.3
Patent Ductus Arteriosis	1,196	61.2
Pulmonary Valve Atresia and Stenosis	181	9.3
Tetralogy of Fallot	113	5.8
Transposition of the Great Arteries	54	2.8
Tricuspid Valve Atresia and Stenosis	25	1.3
Ventricular Septal Defect	861	44.1
Other Cardiovascular	792	40.5
Unduplicated Cardiovascular Total	**2,823**	**144.4**
Unduplicated Other Respiratory Anomalies Total	**300**	**15.4**

Cardiovascular defects are problems with a baby's heart that are present at birth. They are the most commonly occurring birth defects and occur in about 1% of all babies born in the United States. In most cases, scientists do not know why they occur. Most heart defects obstruct the flow of blood in the heart, vessels near it, or cause blood to flow through the heart in an abnormal pattern.

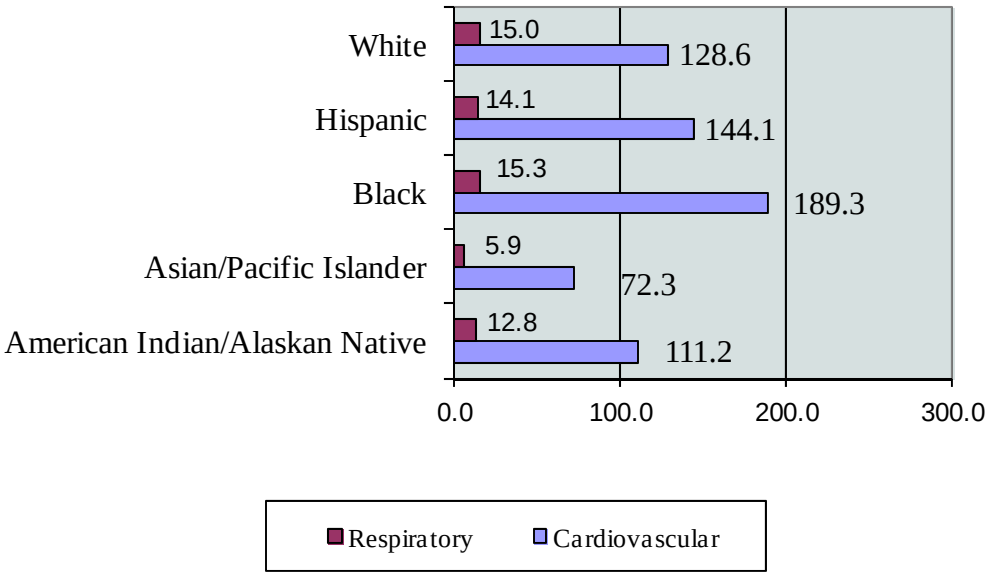
Septal defects - The most common cardiovascular birth defect occurs when holes exist in the walls between heart chambers. These holes allow the mixing of oxygen-rich blood with poor oxygenated blood, resulting in signs and symptoms of congestive heart failure. Ventricular septal defect (VSD) is the most common kind of congenital heart defect and is often referred to as the "hole-in-the-heart." The septum is the wall between the two lower chambers of the heart; 80 to 90% of cases with a very small hole close on their own in the first few months of life. Atrial septal defect (ASD) is a hole between the two upper chambers of the heart. About 80% of ASDs will close on their own in the first 18 months of life. If an ASD has not closed by age three, it is not likely to close and treatment will be necessary.

Incidence: Congenital heart defects are the most common type of birth defects. In the United States, about 40,000 infants are born with a heart defect each year.

Treatment: VSDs and ASDs that do not close on their own are repaired surgically usually with excellent outcomes.

Cardiovascular/Respiratory Anomalies

Chart 2: Prevalence of Cardiovascular/Respiratory Anomalies per 10,000 Live Births by Race/Ethnicity (2005 Through 2009 Pooled Data)



Patent ductus arteriosus (PDA) occurs soon after birth in some babies and is more common in premature babies. Before birth the aorta and pulmonary artery are connected by a blood vessel called the ductus arteriosus, which is an essential part of the fetal circulation. This passage usually closes within the first six weeks of life to allow normal flow of blood to the lungs. When the ductus arteriosus does not close after birth, blood flows directly from the aorta to the pulmonary artery which can put a strain on the heart and increase blood pressure in the lung arteries. If a patent ductus arteriosus is still present beyond the newborn period, it will generally never close on its own and surgical closure is recommended.

Incidence: PDA is considered a congenital defect in full term babies and occurs in about 8 of 1,000 live (premature) births.

Treatment: If the anomaly does not close by two years of age, surgery will be necessary to close off the vessel.

Pulmonary valve atresia and stenosis is a rare congenital heart defect where the pulmonary artery that leaves the right ventricle is not open and the right ventricle is very small. Babies with this disorder are usually full term and appear normal as the heart was not required to transport oxygen to the baby before birth. Shortly after birth the ductus arteriosis (a shunt between the aorta and pulmonary artery) closes and there is no way for blood to get to the lungs and refresh with oxygen. This produces a life-threatening situation and life saving measures are necessary.

Incidence: Pulmonary atresia/stenosis is a rare condition that occurs in about 7 to 8 children per 100,000 live births.

Cardiovascular/Respiratory Anomalies

Table 2: Nevada Demographics for Cardiovascular/Respiratory Anomalies (2005 Through 2009 Pooled Data)

*Demographic	Number Card/Resp	Percent Card/Resp
Birth Type		
Vaginal Birth	1,245/113	57.4/51.6
C Section Primary	667/87	30.7/39.7
C Section Repeat	258/19	11.9/8.7
Mother's Age		
17 and younger	83/7	3.2/2.7
18 to 24	719/79	27.5/30.7
25 to 34	1,314/126	50.2/49.0
35 to 44	490/42	18.7/16.3
45 and older	9/3	0.3/1.2
Sex		
Male	1,409/162	49.9/54.2
Female	1,413/137	50.1/45.8
Region		
Clark County	2,434/253	86.2/84.3
Washoe County	262/27	9.3/9.0
Balance-of-State	127/20	4.5/6.7
Total Unduplicated Individuals with Cardiovascular/ Respiratory Anomalies		**2,823/300**

* Demographic data is not available for all infants; therefore, the sum of individual demographic categories may not equal the total unduplicated number of individuals.

Treatment: Children with this disorder will be given a drug (prostaglandin) to keep the ductus arteriosis open so blood can go through this fetal vessel to the lungs for oxygen. Surgery will be required to repair the pulmonary valve and a stent may be used to keep the ductus arterosis; additional surgery may be needed.

Tetralogy of Fallot is a combination of four heart defects (ventricular septal defect, pulminary stenosis, right ventricular hypertrophy, and overriding aorta) that keep blood from getting to the lungs. Due to the lack of oxygen, affected babies have episodes of cyanosis and may have poor growth. This defect is usually surgically repaired around three to six months of age and most affected children live normal lives.

Incidence: Tetralogy of Fallot occurs in about 1 of 2,518 United States births annually.

Treatment: Tetralogy of Fallot is treated surgically. If the baby is small, a temporary operation may be performed with complete repair done at a later date.

Central Nervous System Anomalies

Table 3: Nevada Incidence rates of Central Nervous System Anomalies (2005 Through 2009 Pooled Data)

Central Nervous System Anomalies	Cases	Rate per 10,000 Births
Anencephalus	8	0.4
Encephalocele	12	0.6
Hydrocephalus	126	6.4
Microcephalus	71	3.6
Spina Bifida without hydrocephalus	37	1.9
Other Central Nervous System	81	4.1
Unduplicated Central Nervous System Total	344	17.6

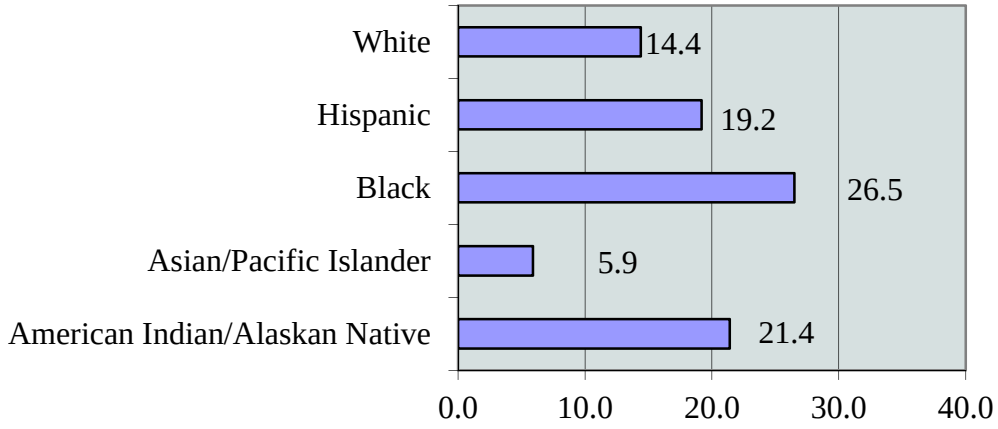
The central nervous system (CNS) consists of the brain and spinal cord. The CNS of a growing fetus starts with a simple structure called the neural groove, which folds into the neural tube, and eventually develops into the spinal cord and brain. By day 28 of conception, the neural tube should be closed and fused. If it does not close, the result is a neural tube defect. Structural defects in the brain can also be caused by the interruption of brain development into discrete, interacting regions.

Anencephaly is a serious congenital defect of CNS when the cerebrum and cerebellum are reduced or totally absent, but the hindbrain is present. It occurs due to disruptions of neural tube closure during the third and fourth weeks of development of the brain. The consequences of this defect are fetal loss, still birth, or neonatal death. Causes may be genetic, environmental, chromosomal, maternal diabetes mellitus, or deficiency of folic acid.
Incidence: Incidence rate in the United States is approximately 3.4 per 10,000 births with a gradient of increasing frequency from the west coast to east. Prevalence is higher among Whites and Hispanics than Blacks, and females are more likely to be affected than males.
Treatment: This condition results in a life span of no more than a few hours or days.

Encephalocele is a rare neural tube defect caused by failure of the neural tube to close completely during fetal development and characterized by sac-like protrusions of the brain and the membranes that cover it through openings in the skull. Usually encephaloceles are

Central Nervous System Anomalies

Chart 3: Prevalence of Central Nervous System Anomalies per 10,000 Live Births by Race/Ethnicity (2005 Through 2009 Pooled Data)



diagnosed immediately after birth, but occasionally a small encephalocele in the nasal and forehead region can go undetected. Currently most cases are identified prenatally by ultrasound. Encephaloceles are often accompanied by craniofacial abnormalities or other brain malformations such as hydrocephalus (excessive accumulation of cerebrospinal fluid in the brain), spastic quadriplegia (paralysis of the arms and legs), and microcephaly (abnormally small head).

Incidence: In the United States encephalocele has an incidence of one per 10,000 live births. Twenty-one percent of infants with encephalocele are born live, but only about half survive. Of these, about 75% having mental deficits.

Treatment: Surgery is performed to remove the sac and the abnormal neural tissue it contains. Multi-stage surgery may be needed, depending on the extent of the encephalocele.

Congenital hydrocephalus is a buildup of excess cerebrospinal fluid (CSF) within the brain that is present at birth. The excess fluid can increase pressure in the baby's brain, possibly resulting in brain damage and loss of mental and physical abilities. Prompt diagnosis and treatment is important to help limit serious long-term problems. In infancy, the most obvious indication of hydrocephalus is often the rapid increase in head circumference or an unusually large head size.

Incidence: It is estimated that 1 baby out of every 1,000 are born with the condition.

Central Nervous System Anomalies

Table 4: Nevada Demographics for Central Nervous System Anomalies (2005 Through 2009 Pooled Data)

*Demographic	Number	Percent
Birth Type		
Vaginal Birth	112	40.6
C Section Primary	138	50.0
C Section Repeat	26	9.4
Mother's Age		
17 and younger	15	4.9
18 to 24	100	32.7
25 to 34	134	43.8
35 to 44	56	18.30
45 and older	1	0.3
Sex		
Male	167	48.7
Female	176	51.3
Region		
Clark County	281	81.7
Washoe County	43	12.5
Balance-of-State	20	5.8
Total Unduplicated Individuals with CNS Anomalies		**344**

* Demographic data is not available for all infants; therefore, the sum of individual demographic categories may not equal the total unduplicated number of individuals.

Treatment: Hydrocephalus is most often treated with the surgical placement of a shunt-system. This system diverts the flow of CSF from a site within the central nervous system to another area of the body where it can be absorbed as part of the circulatory process.

Spina bifida is the most common form of neural tube defect; infants born with the milder form of spina bifida may be asymptomatic rarely having disabilities. The more severe form results in complete or partial paralysis of body parts below the opening. About 90% of children born with the worst form of spina bifida have hydrocephalus and must have a shunt inserted surgically to drain fluid. With new medical treatments and technology most people born with spina bifida live normal lives, but may have many special challenges. Studies show that administration of folic acid during the preconception and conception periods can reduce the incidence rate and severity of neural tube defects by 80%.

Incidence: Spina bifida is the most common severe birth defect, occurring in about 1 out of every 2,858 live births.

Treatment: The common spina bifida occulta typically causes no problems because the underdeveloped gap in the vertebrae is very small and the spinal cord is normal. Surgery is required for treatment of more severe forms of spina bifida with exposed neural tissue.

Chromosomal Anomalies

Table 5: Nevada Incidence Rates of Chromosomal Anomalies (2005 Through 2009 Pooled Data)

Chromosomal Anomalies	Cases	Rate per 10,000 Births
Down Syndrome (Trisomy 21)	271	13.9
Edwards Syndrome (Trisomy 18)	29	1.5
Patau Syndrome (Trisomy 13)	10	0.5
Other Chromosomal	35	1.8
Unduplicated Chromosomal Anomalies Total	**356**	**18.2**

Chromosomal abnormalities can be organized into two basic groups: numerical and structural abnormalities. Numerical abnormalities are caused by additional or missing chromosomes from a chromosome pair before or during conception. Examples of an additional chromosome (three chromosomes instead of two) are Down Syndrome (Trisomy 21), Edwards Syndrome (Trisomy 18), and Patau Syndrome (Trisomy 13). Structural abnormalities occur when there are changes of chromosome structure during chromosome replication. About one in 500 babies are born with a missing or an extra sex chromosome. Chromosomal abnormalities can be detected through different types of screening before or after birth.

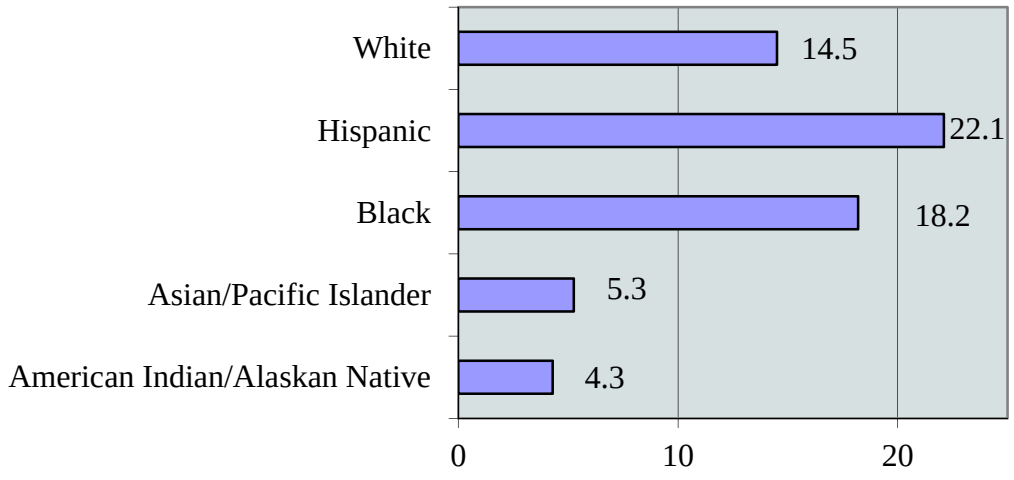
Down syndrome (Trisomy 21) is caused by the presence of three 21st chromosomes instead of two. Down Syndrome is characterized by impairment of cognitive ability, impairment of physical growth, and facial configuration. Children with Down Syndrome tend to have certain physical features such as a flat facial profile, an upward slant to the eyes, small ears, a single crease across the center of the palms, and an enlarged tongue. This condition in a newborn can usually be determined through a physical exam. Down Syndrome increases the risk of congenital heart disease, gastro-esophageal reflux disease, recurrent ear infections, obstructive sleep apnea, and thyroid dysfunctions. Approximately half of children with Down Syndrome have congenital heart defects and are prone to develop pulmonary hypertension.

Incidence: The CDC estimates that Down Syndrome occurs in one out of 691 live births in the United States.

Treatment: The number and type of associated conditions will determine treatment for children with Down Syndrome.

Chromosomal Anomalies

Chart 4: Incidence of Chromosomal Anomalies per 10,000 Live Births by Race/Ethnicity (2005 Through 2009 Pooled Data)



Edwards syndrome (Trisomy 18) is named after John Edwards who first described the disease and is the most common chromosomal abnormality after Down Syndrome. Approximately half of the cases diagnosed during the prenatal period do not survive until birth. Edwards Syndrome is caused by the presence of three 18th chromosomes instead of two. Like Down Syndrome, it occurs before conception during sex cell formation. Major clinical features of this syndrome are low birth weight, abnormally-shaped small head, small jaw, small mouth, low set of ears, and clenched fists with overlapping fingers. Babies with Edwards Syndrome may have heart defects and other malformations and the survival rate is very low through their first year of life (about 5 to 10 percent).
Incidence: Edwards syndrome has an estimated incidence rate of 1 in 3,762 live births.
Treatment: Treatment is very rarely an option, typically because of the severity of the baby's heart defects.

Patau syndrome (Trisomy 13) is caused by the presence of three 13th chromosomes and is the least common of the autosomal trisomies, after Down syndrome and Edwards syndrome. Babies are identified at birth and diagnosis is confirmed with genetic testing. Babies are small at birth and have characteristic facial appearances. Patau syndrome causes serious physical and mental abnormalities including: heart defects; incomplete brain development, unusual facial features, cleft palate or hare lip, extra fingers and toes, abnormal genitalia, spinal defects and mental retardation. Due to the seriousness of these conditions, fewer than

Chromosomal Anomalies

Table 6: Nevada Demographics for Chromosomal Anomalies (2005 Through 2009 Pooled Data)

*Demographic	Number	Percent
Birth Type		
Vaginal Birth	154	59.0
C Section Primary	84	32.2
C Section Repeat	23	8.8
Mother's Age		
17 and younger	6	1.9
18 to 24	57	18.2
25 to 34	115	36.6
35 to 44	127	40.5
45 and older	9	2.9
Sex		
Male	145	40.8
Female	210	59.2
Region		
Clark County	286	80.3
Washoe County	49	13.8
Balance-of-State	21	5.9
Total Unduplicated Individuals with Chromosomal Anomalies		**356**

* Demographic data is not available for all infants; therefore, the sum of individual demographic categories may not equal the total unduplicated number of individuals.

20% of babies with Patau syndrome survive beyond infancy. About five percent of children survive the first year and exhibit developmental and growth delays. The list of possible complications is large because so many body systems are affected by this condition.

Incidence: Trisomy 13 occurs in about 1 out of 7,906 births and the incidence increases with maternal age.

Treatment: Regardless of medical intervention, most babies will not live past the first week of life.

Ear/Eye Anomalies

Table 7: Nevada Incidence Rates of Ear/Eye Anomalies (2005 Through 2009 pooled data)

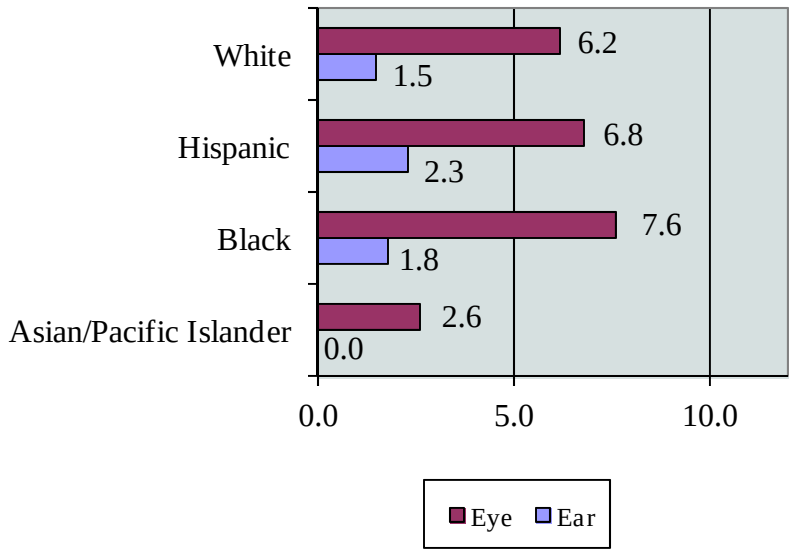
Ear/Eye Anomalies	Cases	Rate per 10,000 Births
Anotia/Microtia	6	0.3
Other Ear	28	1.4
Unduplicated Ear Total	**32**	**1.6**
Aniaridia	0	0.0
Anophthalmia/Micropthalmia	17	0.9
Congenital Cataract	12	0.6
Other Eye	110	5.6
Unduplicated Eye Total	**129**	**6.6**

Anotia refers to the congenital absence of the external ear (auricle) most often with a narrowing or absence of the external auditory canal. **Microtia** is a condition in which the auricle is malformed and there is usually a narrowing or absence of the external auditory canal. This is different from "small ear" in which the ear is normally shaped, but smaller than normal as in Down syndrome. Over 80% of the cases are unilateral (one ear). Anotia/microtia can occur independently or as part of a syndrome (anotia/microtia is an isolated condition in about 65% of cases). Surgical correction is the preferred treatment for both microtia and anotia. These disorders are more common among the Japanese and Navajo Indian populations, but unusual in the Hispanic population. They are also more common in males.
Incidence: Anotia/microtia occur in about 1 out of 6,000 to 12,000 births.
Treatment: Due to variations in this condition, treatment plans will vary for each individual. Surgery may consist of the reconstruction of the external ear and/or the creation of ear canals so sound can travel to the middle and inner ear.

A congenital cataract is cloudiness in the lens of the eye that is present at birth or develops shortly after birth. The lens is located at the front of the eye and focuses light and images on the retina at the back of the eye. This lens is essential for vision, and if cloudy, vision will be blurry. In the case of newborn infants, a cataract causes the immature visual system to be deprived of normal development and, if left untreated, permanent visual loss may occur. Congenital cataracts may affect one or both eyes.
Incidence: Congenital cataracts are quite rare, about 3 in 10,000 births.
Treatment: Surgical extraction of the affected lens is the most effective treatment for most cases. Once the lens is removed, it may be replaced by a lens placed in the eye, but in most

Ear/Eye Anomalies

Chart 5: Incidence of Ear/Eye Anomalies per 10,000 Live Births by Race/Ethnicity (2005 Through 2009 Pooled Data)



cases this does not happen and glasses or contacts will be required. If no complications occur, and children use their corrective lenses, children with congenital cataracts can have near normal vision.

Aniridia is a rare congenital absence or partial absence of the iris. Though not entirely absent, all that remains of the iris, the colored part of the eye, is a collar of tissue around the pupil of the eye. The muscles that open and close the pupil are entirely missing. There are many variations of aniridia and vision may be limited or greatly reduced. Parents will often detect this condition when the baby is very sensitive to light and the pupil of the eye seems very large. Other deformities are often present and glaucoma frequently develops before adolescence. Pinhole contact lenses and/or sunglasses and optical aids can be used to control glare. If glaucoma develops, medical and/or surgical treatment may help, but long-term prognosis is poor.

Incidence: This anomaly is very rare and occurs in about 1 out of 60,000 to 90,000 births.

Treatment: In the short term, children with aniridia do very well, though they need to be carefully followed by an ophthalmologist throughout life so complications can be diagnosed an treated promptly. There is no cure and individuals with aniridia will have to adapt to reduced vision. Vision typically gets worse with age and cataract surgery may be required and glaucoma frequently develops.

Ear/Eye Anomalies

Table 8: Nevada Demographics for Ear/Eye Anomalies (2005 Through 2009 Pooled Data)

*Demographic	Number Ear/Eye	Percent Ear/Eye
Birth Type		
Vaginal Birth	14/51	58.3/54.8
C Section Primary	10/37	41.7/39.8
C Section Repeat	0/5	0.0/5.4
Mother's Age	**Number**	**Percent**
17 and younger	1/5	3.2/4.4
18 to 24	10/25	32.3/21.9
25 to 34	17/68	54.9/59.7
35 to 44	3/16	9.7/14.0
45 and older	0/0	0/0
Sex	**Number**	**Percent**
Male	19/81	59.4/63.3
Female	13/47	40.6/36.7
Region	**Number**	**Percent**
Clark County	25/106	78.1/82.2
Washoe County	6/15	18.8/11.6
Balance-of-State	1/8	3.1/6.2
Total unduplicated individuals with Ear/Eye Anomalies		**32/129**

* Demographic data is not available for all infants; therefore, the sum of individual demographic categories may not equal the total unduplicated number of individuals.

Microphthalmia is a disorder in which one or both eyes are abnormally small, while **anophthalmia** is the absence of one or both eyes. These disorders develop during pregnancy and can be associated with other birth defects. Causes of these conditions may include genetic mutations and abnormal chromosomes. Environmental factors such as exposure to X-rays, chemicals, drugs, pesticides, toxins, radiation, or viruses may increase the risk of anophthalmia or microphthalmia, but research is not conclusive.

Incidence: These are rare conditions and occur in about 2 out of 10,000 live births.

Treatment: There is no treatment for severe anophthalmia or microphthalmia that will create a new eye or restore vision; however, some less severe forms of microphthalmia may benefit from medical or surgical treatments. Children with eye defects are at increased risk for detached retina or glaucoma.

Gastrointestinal Anomalies

Table 9: Nevada Incidence Rates of Gastrointestinal Anomalies (2005 Through 2009 Pooled Data)

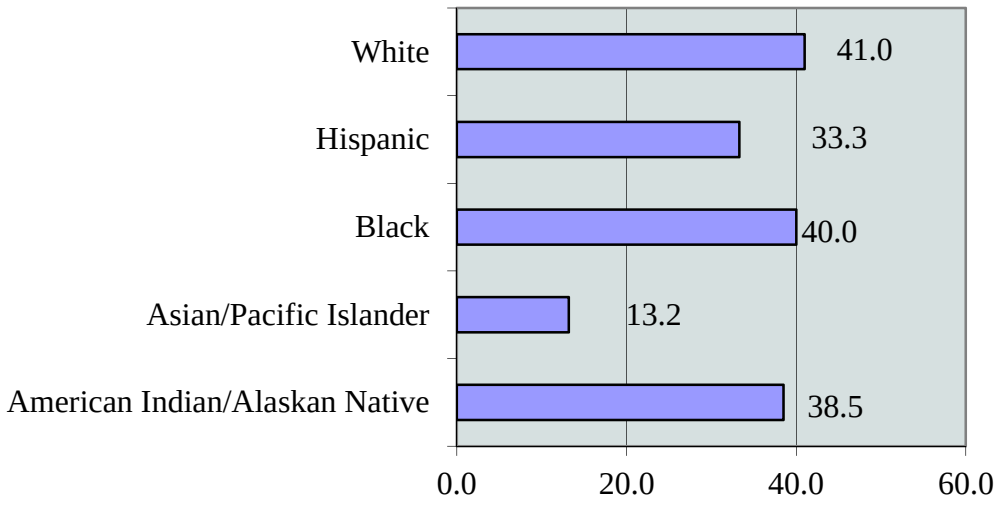
Gastrointestinal Anomalies	Cases	Rate per 10,000 Births
Biliary Atresia	13	0.7
Esophageal Atresia/Tracheoesophageal Fistula	43	2.2
Hirshsprung's Disease (congenital megacolon)	40	2.0
Small Intestine Atresia/Stenosis	70	3.6
Rectal and Large Intestinal Atresia/Stenosis	71	3.6
Other Gastrointestinal Anomalies	226	11.6
Unduplicated Gastrointestinal Total	**752**	**38.5**

Gastrointestinal (GI) anomalies are structural defects that can occur at any point along the gastrointestinal tract, which includes the esophagus, stomach, small and large intestines, rectum, and anus. GI malformations result in some type of intestinal obstruction that frequently cause feeding difficulties, distention, and vomiting shortly after birth. Early identification, immediate postnatal evaluation, and timely intervention decrease neonatal mortality and increase the chance of long-term infant survival.

Congenital hypertrophic pyloric stenosis is the narrowing of stomach into the small intestine (pylorus). This abnormality is not present at birth but develops within a few days or weeks after birth. The pyloric valve regulates the emptying of the stomach contents into the small intestine. Projectile vomiting occurs two to four weeks after birth due to failure of the stomach to empty. Babies will be very hungry after vomiting and persistent vomiting can cause dehydration and electrolyte imbalance which can be life-threatening.
Incidence: This anomaly occurs in approximately 3 out of every 1,000 live births and is four times more common in males. It is more common in Whites, compared to Blacks, and rarely seen in Asians.
Treatment: Initially infants will need intravenous fluids to bring their hydration and electrolytes back to normal. Babies who receive timely diagnoses and undergo surgery have excellent outcomes.

Gastrointestinal Anomalies

Chart 6: Prevalence of Gastrointestinal Anomalies per 10,000 Live Births by Race/Ethnicity (2005 Through 2009 Pooled Data)



Hirschsprung's disease occurs when the lower part of the large intestine is partly or totally immobilized due to faulty development of nerve cells located in the intestine. Most babies have just a short section of intestine affected so this disease if often termed "short segment disease." Only about 15% of the cases have large sections of the bowel affected and the entire large intestine is affected in about 5% of the cases. Digested food particles are completely or partially trapped and the affected part of the colon becomes enlarged. With surgical removal of the affected part of the colon, prognosis is good.
Incidence: Hirschsprung's disease occurs in about 1 out of 6,300 live births.
Treatment: The first stage of treatment is called a temporary colostomy that brings a functioning loop of the large intestine to the surface. An opening in the skin is created for the bowel to drain and heal. The affected portion of the large intestine is removed and the colostomy closed in the second stage of surgery. Children who are healthy and have a small segment of bowel affected may have a one-stage surgical repair.

Esophageal atresia, with or without tracheoesophageal fistula, is a congenital disorder of the feeding passage connecting the mouth with the stomach. The esophagus does not connect to the stomach because of a missing segment. The esophagus ends in a pouch so nothing the baby swallows gets to the stomach. A tracheoesophageal fistula is a connection between the breathing tube (trachea) and the swallowing tube (esophagus). These tubes are not supposed to be

Gastrointestinal Anomalies

Table 10: Nevada Demographics for Gastrointestinal Anomalies (2005 Through 2009 Pooled Data)

*Demographic	Number	Percent
Birth Type		
Vaginal Birth	293	61.3
C Section Primary	152	31.8
C Section Repeat	33	6.9
Mother's Age		
17 and younger	27	4.8
18 to 24	182	32.0
25 to 34	265	46.7
35 to 44	89	15.7
45 and older	4	0.7
Sex		
Male	498	66.3
Female	253	33.7
Region		
Clark County	592	78.7
Washoe County	94	12.5
Balance-of-State	66	8.8
Total Unduplicated Individuals with Gastrointestinal anomalies		**752**

* Demographic data is not available for all infants; therefore, the sum of individual demographic categories may not equal the total unduplicated number of individuals.

connected, but when a tracheoesophageal fistula is present, food or milk can get into the lungs. This can cause breathing problems or even pneumonia. These problems are not thought to be inherited, but occur when the esophagus and trachea develop in the embryo. About 1 in 4,000 babies has one or both of these problems. Many babies with this condition will be born prematurely and they will often have other congenital problems. Technological advances in neonatal intensive care units and special surgical techniques have greatly improved the outlook for children with this disorder. In most cases the infant will survive but will require long-term follow-up.

Incidence: This disorder occurs in approximately 2 out of 10,000 births.

Treatment: The extent of surgery necessary to correct this disorder depends on the size of the gap between the two ends of the esophagus. In some cases, if the gap is small both ends can be connected, the fistula repaired and the baby will do well. With larger gaps, more extensive surgery will be required.

Genitourinary Anomalies

Table 11: Nevada Incidence Rates of Genitourinary Anomalies (2005 Through 2009 Pooled Data)

Genitourinary Anomalies	Cases	Rate per 10,000 Births
Bladder Exstrophy	6	0.3
Epispadias	4	0.2
Hypospadias	375	19.2
Obstructive Genitourinary Defect	534	27.3
Renal Agenesis/Hypoplasia	75	3.8
Undescended Testicles	438	43.4
Other Genitourinary	393	20.1
Total Genitourinary Anomalies	**1,669**	**85.4**

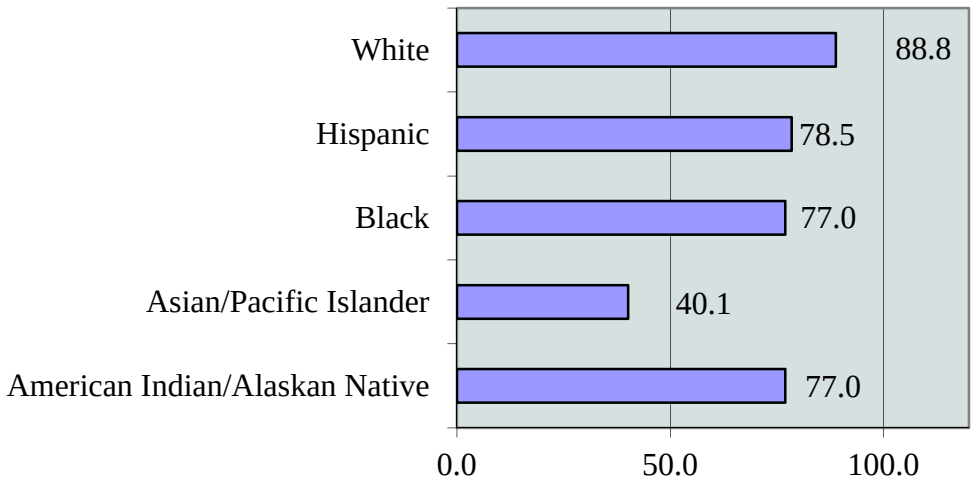
Genitourinary defects are the most common congenital anomalies affecting as many as one in ten babies. Most genitourinary anomalies occur in association with other defects, especially musculoskeletal anomalies. Some of these anomalies are minor and cause no symptoms, others can cause pain, urinary tract infections, and kidney disease. Genital anomalies may cause sexual dysfunction or infertility.

Renal agenesis is the absence of one or both kidneys. **Renal hypoplasia** refers to the presence of an abnormally small, but functional kidney(s). In renal agenesis, kidneys fail to form during the process of fetal development. Renal agenesis is often detected on fetal ultrasound because there will be a lack of amniotic fluid. Urine is needed to form amniotic fluid, which is essential for fetal lung expansion and development. Babies with one kidney can lead a normal life, but when both kidneys are absent the fetus cannot survive, resulting in still birth or death within an hour of birth.
Incidence: According to CDC, the 2006 incidence rate of renal agenesis is 4.4 per 10,000 births; one in 6000 is born with bilateral renal agenesis (both kidneys missing) , and 1 in 550 is born with a single kidney.
Treatment: Bilateral renal agenesis is fatal, but if one kidney is present (unilateral renal agenesis) the child will develop normally.

Obstructive genitourinary defect is the narrowing or absence of urinary tract structures within the urinary tract. The severity depends on the degree and place of obstruction. The most common place of obstruction is between the ureter and kidney at the place where

Genitourinary Anomalies

Chart 7: Prevalence of Genitourinary Anomalies per 10,000 Live Births by Race/Ethnicity (2005 Through 2009 Pooled Data)



they join. Urine does not easily pass through and it backs up in the renal system. The second most common obstruction occurs where the ureter enters the bladder and usually involves one kidney. The extent to which obstructive genitourinary defects will affect the development of the fetus depends on the severity of the obstruction. In newborn babies with high obstruction, the lungs will not develop properly due to the lack of amniotic fluid. If the obstruction is in both kidneys the outlook is poor because hydronephosis (swelling of the kidneys) is typically severe. Although the cause of obstructive genitourinary defect is unknown, these disorders run in families, suggesting a genetic influence.
Incidence: This disorder occurs in approximately 1 out of 350 babies.
Treatment: It is important to maximize renal function as soon as possible by surgically removing the renal blockage.

Hypospadias is a common birth defect of the penis that effects about one percent of baby boys. The urethra does not extend to the tip of the penis, but opens somewhere on the underside. The cause of hypospadias is not known but it is slightly more common in some families. Although rarely (1 out of 5,000,000) girls may have a misplaced urethra that opens into the vagina, the term hypospadias generally refers to the condition in boys.
Incidence: This is a common disorder that occurs in about 1 out of every 300 baby boys.
Treatment: Affected boys should not be circumcised because the foreskin is used in surgi-

Genitourinary Anomalies

Table 12: Nevada Demographics for Genitourinary Anomalies (2005 Through 2009 Pooled Data)

*Demographic	Number	Percent
Birth Type		
Vaginal Birth	838	62.4
C Section Primary	397	29.5
C Section Repeat	109	8.1
Mother's Age		
17 and younger	68	4.4
18 to 24	473	30.1
25 to 34	789	50.5
35 to 44	227	14.5
45 and older	4	0.3
Sex		
Male	1,390	83.3
Female	278	16.7
Region		
Clark County	1,362	81.6
Washoe County	229	13.7
Balance-of-State	78	4.7
Total Unduplicated Individuals with Genitourinary anomalies		**1,669**

* Demographic data is not available for all infants; therefore, the sum of individual demographic categories may not equal the total unduplicated number of individuals.

cal repair. The surgery is usually performed between six and 18 months of age.

Undescended testis is a common birth defect in which the testis located in the fetal abdomen do not descend to the scrotum. In the womb, the testis remain in the abdomen and descend shortly before or just after birth. The testis usually descend on their own within the first few months, but if not may require treatment by a doctor. The scrotal location keeps the testis cooler than the core body temperature, which is important for the development of the testicle and sperm production. Undescended testicles cause increased risk of testicular cancer, which is highly curable when detected early.

Incidence: Undescended testis may be present in three to four percent of boys at birth, and there is even higher incidence in premature infants. About three-fourths of undescended testicles will descend within the first three months of life.

Treatment: Treatment of undescended testicles is recommended before one year of age. The most effective treatment is outpatient surgery. Most undescended testicles are associated with a

Musculoskeletal Anomalies

Table 13: Nevada Incidence Rates of Musculoskeletal Anomalies (2005 Through 2009 Pooled Data)

Musculoskeletal Anomalies	Cases	Rate per 10,000 Births
Congenital Hip Dislocation	113	5.8
Diaphragmatic Hernia	53	2.7
Gastroschisis/Omphalocele	97	5.0
Limb Deficiencies (Reduction Defects)	60	1.3
Other Musculoskeletal	1,141	58.4
Total Musculoskeletal Anomalies	**1,585**	**75.0**

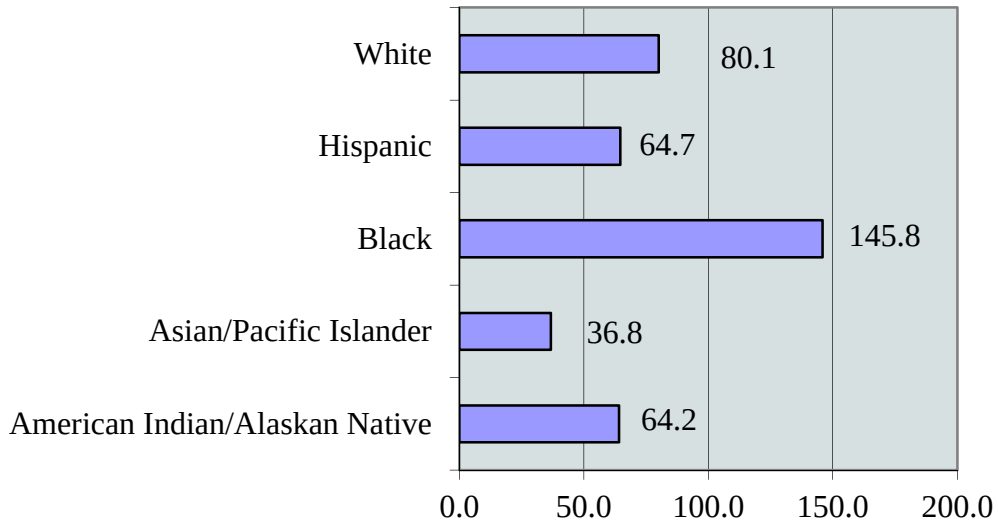
Diaphragmatic hernia is the protrusion of abdominal organs into the thoracic cavity through an abnormal opening in the diaphragm. In the most common type of diaphragmatic hernia, Bochdalek hernia, the diaphragm may not develop properly and the abdominal organs may become trapped in the chest cavity as the diaphragm is forming. In the less common Morgagni diaphragmatic hernia, the tendon in the middle of the diaphragm does not develop properly. In both cases, normal development of the diaphragm and digestive tract does not occur. Congenital diaphragmatic hernias are very serious disorders, but with advances in neonatal care and surgical techniques, survival is now greater than 80%.
Incidence: Diaphragmatic hernias occur in about 1 out of 2,000 to 5,000 live births, and boys are more likely to be affected. Bochdalek hernias make up about 90% of all cases.
Treatment: Diaphragmatic hernias are life-threatening disorders and require care in a neonatal intensive care unit. When the baby's condition improves, the hernia will be repaired surgically.

Gastroschisis is the herniaton of abdominal contents (small intestine and/or portions of the large intestine) into the amniotic fluid through an opening in the abdomen. Gastroschisis does not involve the umbilical cord and herniation usually occurs to the right of the naval. This defect can be detected by ultrasound before birth. The affected organs become vulnerable to damage during and after birth. Since gastroschisis exposes the fetal intestines to the amniotic fluid and they are unprotected during pregnancy, there is an increased risk for third trimester complications.
Incidence: Gastroschisis occurs in about 2 out of 10,000 live births.
Treatment: Therapy is directed toward nutrition, surgical closure of the defect, and prevention of infection.

Omphalocele is a malformation in which variable amounts of abdominal contents pro-

Musculoskeletal Anomalies

Chart 8: Incidence of Musculoskeletal Anomalies per 10,000 Live Births by Race/Ethnicity (2005 Through 2009 Pooled Data)



trude into the base of the umbilical cord. As the intestines grow and get longer they project from the abdomen into the umbilical cord, but normally return into the abdomen by the eleventh week of pregnancy. If this fails to happen, an omphalocele is present. More than half of all infants born with an omphalocele have other defects; prognosis depends largely on the presence of other associated anomalies.

Incidence: Omphalocele occurs in about 1 out of every 5,386 live births and about 50% will have other congenital defects.

Treatment: Small omphaloceles are repaired immediately to prevent infection or tissue damage, but larger omphaloceles may require gradual reduction by enlarging the abdominal cavity to accommodate the intestines.

Congenital hip dislocation, also called developmental dysplasia of the hip, is due to abnormal formation of the hip during fetal development causing an unstable hip socket. This disorder is sometimes identified at birth, but often later in infancy. The hip joint has a shallow socket that is prone to dislocate as the baby grows and most babies have a looseness of the ligaments that hold the hip joint in place. All babies are examined for hip dysplasia after birth and during early visits to the doctor.

Incidence: This condition occurs in about 1 out of 100 live births, but approximately 1 out of 800 babies have hips that dislocate.

Musculoskeletal Anomalies

Table 14: Nevada Demographics for Musculoskeletal Anomalies (2005 Through 2009 Pooled Data)

*Demographic	Number	Percent
Birth Type		
Vaginal Birth	694	54.3
C Section Primary	501	39.2
C Section Repeat	83	6.5
Mother's Age		
17 and younger	57	3.8
18 to 24	499	33.1
25 to 34	705	46.7
35 to 44	243	16.1
45 and older	5	0.3
Sex		
Male	836	52.8
Female	748	47.2
Region		
Clark County	1,272	80.2
Washoe County	244	15.4
Balance-of-State	69	4.4
Total Unduplicated Individuals with Musculoskeletal anomalies		**1,585**

* Demographic data is not available for all infants; therefore, the sum of individual demographic categories may not equal the total unduplicated number of individuals.

Treatment: Treatment techniques keep the ball of the thigh bone (femur) tightly positioned against the hip joint for two months so the joint will form a normal cup joint and the head of the femur will be round. Surgery may be necessary later in life if the joint does not develop properly.

Reduction deformity is the congenital absence of a limb or part of a limb and includes a shortened limb. This can be caused if the mother is exposed to certain drugs before birth and can be detected before birth by ultrasound. Women who take folic acid during pregnancy have a 30 to 40% reduced risk of having a child with a limb deformity.

Incidence: Upper limbs are affected more than lower limbs with an estimated incidence of 34 per 100,000 live births compared to deformities of the lower limbs at about 14 per 100,000 live births.

Orofacial Anomalies

Table 15: Nevada Incidence Rates of Orofacial Anomalies (2005 Through 2009 Pooled Data)

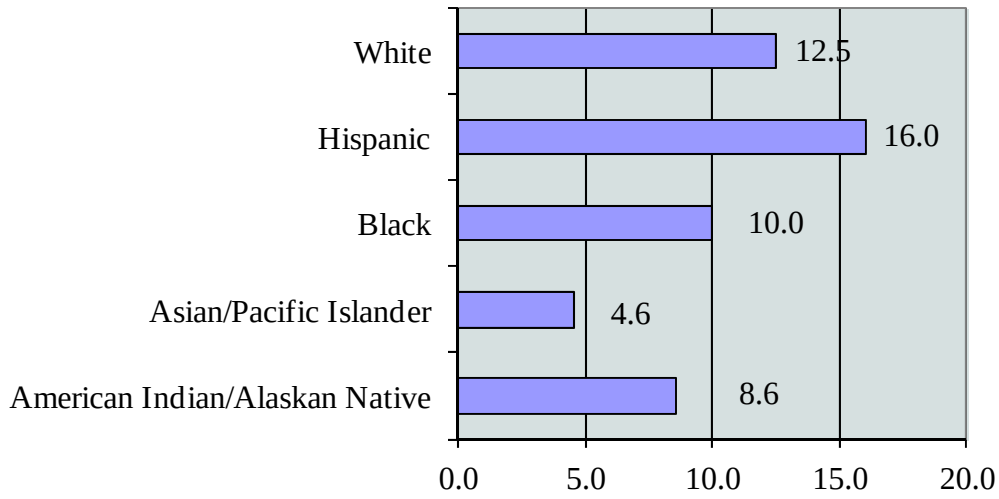
Orofacial Anomalies	Cases	Rate per 10,000 Births
Anomalies of the Skull and Face Bones	128	6.5
Cleft Lip without Cleft Palate	131	6.7
Cleft Palate without Cleft Lip	77	3.9
Cleft Lip with Cleft Palate	51	2.6
Choanal Atresia	22	1.1
Total Orofacial Anomalies	**267**	**13.7**

Oral-facial clefts (cleft lip and/or cleft palate) are facial deformities caused by the failure of fetal tissues to properly come together to form the lips and the roof of the mouth (palate). These facial features are formed early in pregnancy (about 4 to 7 weeks). Some babies only have a cleft lip, but many have a cleft palate as well. Cleft palate can also occur without cleft lip (isolated cleft palate). Cleft lip/palate and isolated cleft palate are considered separate birth defects. Doctors don't know exactly why babies develop a cleft lip or cleft palate, but it is believed to be a combination of genetic (inherited) and environmental factors such as (drugs, illnesses, alcohol or tobacco use by the mother during pregnancy). A child with a cleft lip or palate tends to be more susceptible to colds, hearing loss, and speech defects. CDC reports cleft lip and/or cleft palate as one of the six major types of birth defects affecting about 6,800 babies in the United States each year. This number represents oral-facial clefts not accompanied by other birth defects (isolated oral-facial clefts). About 20% of children with oral-facial clefts have cleft lip only, 30% have cleft palate only, and 50% are born with cleft lip and cleft palate. Ear infections, feeding difficulty, speech problems, and dental problems accompany cleft lip and palate.

A cleft lip can vary from a small slit in the upper lip to a complete opening extending to the nose. The upper gum may also be involved and cleft lip can occur on one or both sides; more rarely it can be in the middle.
Incidence: Cleft lip and palate occur in about 1 out of every 1,000 babies and slightly more in males than females.
Treatment: Treatment plans are unique to the individual; treatment is done by an interdisciplinary team. Surgery to correct a cleft lip and palate is done in stages. Lip repair is done about three months of age and the palate will be repaired once the child's facial structures have grown bigger so that tissue can successfully be pulled together. If gums are involved dental work and or oral surgery may also be necessary.
A cleft palate (without cleft lip) is an opening in the roof of the mouth (the palate). The pal-

Orofacial Anomalies

Chart 9: Incidence of Orofacial Anomalies per 10,000 Live Births by Race/Ethnicity (2005 Through 2009 Pooled Data)



ate is formed at about 6 to 9 weeks of pregnancy and a cleft occurs when the two sides of the palate do not come together correctly. In some children both the hard palate (front of the mouth) and soft palate (back of the mouth) are open and in others only part of the palate is open.

Incidence: Cleft palate alone occurs in about 1 out of 1,574 babies and is less common than the combined cleft lip and palate. It affects females more than males.

Treatment: As with cleft lip and palate treatment will be unique to the individual's needs and done with an interdisciplinary team. Children with these defects may need further surgery and dental work as they grow.

Choanal atresia is a congenital disorder where the back of the nasal passage (choana) is blocked by a membrane or boney tissue. This obstruction fails to rupture when it typically does on about the 38th day of fetal development. Approximately 60% of reported cases are unilateral (one-sided) with a right-sided predominance. Bilateral choanal atresia (both sides) is life-threatening because the baby will not be able to breath during feeding. Surgery is required to correct the defect. Most unilateral cases are isolated anomalies, but approximately 50% of bilateral cases are associated with other craniofacial syndromes and skull based defects including encephalocele.

Incidence: Choanal atresia is the most common nasal abnormality and occurs in about 1 out

Orofacial Anomalies

Table 16: Nevada Demographics for Orofacial Anomalies (2005 Through 2009 Pooled Data)

*Demographic	Number	Percent
Birth Type		
Vaginal Birth	124	63.9
C Section Primary	52	26.8
C Section Repeat	18	9.3
Mother's Age		
17 and younger	9	3.9
18 to 24	86	37.2
25 to 34	97	42.0
35 to 44	38	16.5
45 and older	0	0.0
Sex		
Male	119	44.6
Female	148	55.4
Region		
Clark County	201	75.3
Washoe County	49	18.4
Balance-of-State	17	6.4
Total Unduplicated Individuals with Orofacial Anomalies		**267**

* Demographic data is not available for all infants; therefore, the sum of individual demographic categories may not equal the total unduplicated number of individuals.

of every 7,000 – 10,000 births.

Short term treatment: A small plastic tube will be surgically inserted in the nostril to create an airway for the baby until more extensive surgery can be done.

Long term treatment: Surgery will be necessary, perhaps involving both a nose-and-throat specialist as well as a plastic surgeon, to create a functioning airway and to obtain a good cosmetic appearance of the nose.

Appendix A:
Selected References

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Appendix B:
Incidence Rates of Birth Anomalies Arranged in CDC Categories

Appendix B: Incidence Rates of Infants Born in Nevada with Birth Anomalies - Arranged in CDC Categories (Pooled Data 2005 through 2009)

Category	Anomaly	Cases	Incidence Per 10,000 Live Births
Anomalies of the Face and Neck	Anomalies of the Face and Neck	20	1.0 (0.8-1.3)
Anomalies of the Skin	Anomalies of the Skin	28	1.4 (1.2-1.7)
Cardiovascular	AVSD (Endocardial Cushion Defect)	56	2.9 (2.5-3.2)
	Aortic Valve Stenosis	41	2.1 (1.8-2.4)
	Atrial septal defect	1,349	69.0 (67.1-70.9)
	Coarctation of the Aorta	118	6.0 (5.5-6.6)
	Common Truncus	18	0.9 (0.7-1.1)
	Double outlet right ventricle (DORV)	31	1.6 (1.3-1.9)
	Ebstein's Anomaly	19	1.0 (0.7-1.2)
	Hypoplastic Left Heart syndrome	45	2.3 (2.0-2.6)
	Interupted Aortic Arch (IAA)	19	1.0 (0.7-1.2)
	Other Cardiovascular	792	40.5 (39.1-42.0)
	Patent Ductus Arteriosus	1,196	61.2 (59.4-63.0)
	Pulmonary Valve Atresia and Stenosis	181	9.3 (8.6-9.9)
	Single Ventricle	24	1.2 (1.0-1.5)
	Tetralogy of Fallot	113	5.8 (5.2-6.3)
	Total Anomalous Pulmonary Venous Return (TAPVR)	17	0.9 (0.7-1.1)
	Transposition of the Great Arteries	54	2.8 (2.4-3.1)
	Tricuspid Valve Artresia and Stenosis	25	1.3 (1.0-1.5)
	Ventricular Septal Defect	861	44.1 (42.6-45.6)
Central Nervous System	Anencephalus	8	0.4 (0.3-0.6)
	Encephalocele	12	0.6 (0.4-0.8)
	Holoprosencephaly	71	3.6 (3.2-4.1)
	Hydrocephalus without Spina Bifida	126	6.4 (5.9-7.0)
	Microcephalus	71	3.6 (3.2-4.1)
	Other Central Nervous System	81	4.1 (3.7-4.6)
	Spina Bifida without Anencephalus	37	1.9 (1.6-2.2)

Appendix B: Incidence Rates of Infants Born in Nevada with Birth Anomalies Arranged in CDC Categories (Pooled Data 2005 through 2009)

Category	Anomaly	Cases	Incidence Per 10,000 Live
Chromosomal	Deletion 22 q11.2	4	0.2 (0.1-0.3)
	Other Chromosomal	35	1.8 (1.5-2.1)
	Trisomy 13 (Patau Syndrome)	10	0.5 (0.3-0.7)
	Trisomy 18 (Edwards Syndrome)	29	1.5 (1.2-1.8)
	Trisomy 21 (Down Syndrome)	271	13.9 (13.0-14.7)
	Tuner's Syndrome	10	0.5 (0.3-0.7)
Ear	Anotia/Microtia	6	0.3 (0.2-0.4)
	Other Ear	28	1.4 (1.2-1.7)
Eye	Anophthalmia/Micropthalmia	17	0.9 (0.7-1.1)
	Congenital Cataract	12	0.6 (0.4-0.8)
	Other Eye	110	5.6 (5.1-6.2)
Gastrointestinal	Biliary Atresia	13	0.7 (0.5-0.8)
	Cloacal Exstrophy	321	16.4 (15.5-17.3)
	Esophagel Atresia/Tracheoesophageal Fistuala	43	2.2 (1.9-2.5)
	Hirshsprung's Disease (Congenital Megacolon)	40	2.0 (1.7-2.4)
	Other Gastrointestinal Anomalies	226	11.6 (10.8-12.3)
	Rectal and Large Intestinal Atresia/Stenosis	71	3.6 (3.2-4.1)
	Small Intestine Atresia/Stenosis	70	3.6 (3.2-4.0)
Genitourinary	Bladder Exstrophy	6	0.3 (0.2-0.4)
	Epispadias	4	0.2 (0.1-0.3)
	Hypospadias	375	19.2 (18.2-20.2)
	Obstructive Genitourinary Defect	534	27.3 (26.1-28.5)
	Other Genitourinary	393	20.1 (19.1-21.1)
	Renal Agenesis/Hypoplasia	75	3.8 (3.4-4.3)
	Undescdended Testicles	438	434.1 (413.4-454.9)

Appendix B: Incidence Rates of Infants Born in Nevada with Birth Anomalies Arranged in CDC Categories (Pooled Data 2005 through 2009)

Category	Anomaly	Cases	Incidence Per 10,000 Live
Musculoskeletal	Anomalies of the Skull and Face Bones (Craniosynostosis)	128	6.5 (6.0-7.1)
	Clubfoot	199	10.2 (9.5-10.9)
	Congenital Hip Dislocation	113	5.8 (5.2-6.3)
	Diaphragmatic Hernia	53	2.7 (2.3-3.1)
	Gastroschisis/Omphalocele	97	5.0 (4.5-5.5)
	Limb Deficiencies (Reduction Defects)	60	3.1 (2.7-3.5)
	Other Musculoskeletal	1,141	58.4 (56.7-60.1)
Orofacial	Choanal Atresia	22	1.1 (0.9-1.4)
	Cleft Palate without Cleft Lip	77	3.9 (3.5-4.4)
	Cleft lip with Cleft Palate	51	2.6 (2.2-3.0)
	Cleft lip without Cleft Palate	131	6.7 (6.1-7.3)
Other	Other/Multiple	93	4.8 (4.3-5.3)
Respiratory	Respiratory	311	15.9 (15.0-16.8)

Appendix C:
ICD-9-CM and CDC/BPA Codes for CDC Designated Disorders

Appendix C: ICD-9-CM and CDC/BPA Codes for CDC Designated Disorders

Birth Defects	DEFECTCD	ICD-9-CM Codes	CDC/BPA Codes
Central Nervous System			
Anencephalus	1	740.0 – 740.1	740.00 – 740.10
Spina bifida without anencephalus	2	741.0, 741.9 w/o 740.0 - 740.1	741.00 – 741.99 w/o 740.00 – 740.10
Encephalocele	4	742.0	742.00 – 742.09
Holoprosencephaly	73	742.2	742.26
Eye			
Anophthalmia/microphthalmia	6	743.0, 743.1	743.00 – 743.10
Congenital cataract	7	743.30 – 743.34	743.32
Ear			
Anotia/microtia	9	744.01, 744.23	744.01, 744.21
Cardiovascular			
Common truncus (truncus arteriosus or TA)	10	745.0	745.00 (excluding 745.01)
Transposition of the great arteries (TGA)	11	745.10, .12, .19	745.10 – 745.12, 745.18 – 745.19
Dextro-Transposition of great arteries (d -TGA) – for CCHD screening	11.4	745.10	745.10, 745.11,745.19
Tetralogy of Fallot (TOF)	12	745.2	745.20 – 745.21, 747.31
Ventricular septal defect	14	745.4	745.40 – 745.49 (excluding 745.487, 745.498)
Atrial septal defect	15	745.5	745.51 – 745.59
Atrioventricular septal defect (endocardial cushion defect)	16	745.60, .61, .69	745.60 – 745.69, 745.487
Pulmonary valve atresia and stenosis	17	746.01, 746.02	746.00, 746.01
Pulmonary valve atresia – for CCHD screening	17.4	746.01	746.00
Tricuspid valve atresia and stenosis	18	746.1	746.100, 746.106 (excluding 746.105)
Tricuspid valve atresia– for CCHD screening	18.4	746.1	746.100
Ebstein anomaly	19	746.2	746.20

*ICD-9-CM Codes = International Classification of Diseases - Clinical Modification Codes
**BPA Codes = British Pediatric Association Codes

Appendix C: ICD-9-CM and CDC/BPA Codes for CDC Designated Disorders

Birth Defects	DEFECTCD	ICD-9-CM Codes	CDC/BPA Codes
Cardiovascular Continued			
Aortic valve stenosis	20	746.3	746.30
Hypoplastic left heart syndrome	21	746.7	746.70
Coarctation of aorta	23	747.10	747.10 – 747.19
Total anomalous pulmonary venous connection (TAPVC)	24	747.41	747.42
Single ventricle	70	745.3	745.3
Interrupted aortic arch (IAA)	71	747.11	747.215 – 747.217
Double outlet right ventricle (DORV)	72	745.11	745.13 – 745.15
Orofacial			
Cleft palate only (without cleft lip)	26	749.0	749.00 – 749.09
Cleft lip only (without cleft palate)	68	749.1	749.10 – 749.19
Cleft lip with cleft palate	69	749.20-749.25	749.20 – 749.29
Choanal atresia	28	748.0	748.00
Gastrointestinal			
Esophageal atresia/tracheoesophageal fistula	29	750.3	750.30 – 750.35
Rectal and large intestinal atresia/ stenosis	30	751.2	751.20 – 751.24
Biliary atresia	33	751.61	751.65
Small intestinal atresia/stenosis	67	751.1	751.10 – 751.19
Genitourinary			
Renal agenesis/hypoplasia	34	753.0	753.00 – 753.01
Bladder exstrophy	35	753.5	753.50
Hypospadias	50	752.61	752.60 – 752.62 (excluding 752.61 and 752.621)
Congenital posterior urethral valves	65	753.6	753.6
Cloacal exstrophy	66	751.5	751.555

*ICD-9-CM Codes = International Classification of Diseases - Clinical Modification Codes
**BPA Codes = British Pediatric Association Codes

Appendix C: ICD-9-CM and CDC/BPA Codes for CDC Designated Disorders

Birth Defects	DEFECTCD	ICD-9-CM Codes	CDC/BPA Codes
Musculoskeletal Continued			
Gastroschisis	40	756.73 (as of 10/1/09; previous years, it was in a shared code 756.79 with omphalocele)	756.71
Omphalocele	41	756.72 (as of 10/1/09; previous years, it was in a shared code 756.79 with gastroschisis)	756.70
Diaphragmatic hernia	43	756.6	756.60 – 756.62
Limb deficiencies (reduction defects)	62	755.2 – 755.4	755.20 – 755.49
Craniosynostosis	63	No specific code	756.00 – 756.03
Clubfoot	64	754.51, 754.70	754.50, 754.73 (excluding 754.735)
Chromosomal			
Trisomy 13	44	758.1	758.10 – 758.19
Trisomy 21 (Down syndrome)	45	758.0	758.00 – 758.09
Trisomy 18	46	758.2	758.20 – 758.29
Turner syndrome	60	758.6	758.60 – 758.69
Deletion 22 q11.2	61	758.32	758.37

*ICD-9-CM Codes = International Classification of Diseases - Clinical Modification Codes
**BPA Codes = British Pediatric Association Codes

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