

Oklahoma
Birth Defects Registry
1994 - 1998 Annual Report

Introduction

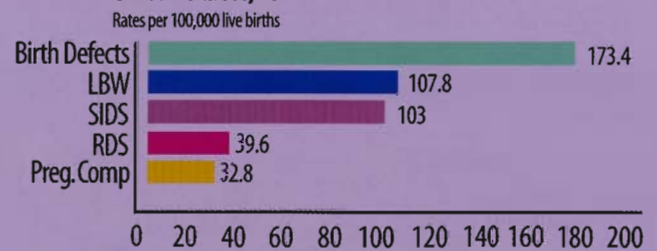
Introduction

Birth defects are the leading cause of infant mortality in the United States (Figure 1) and the fifth leading cause of years of potential life lost, contributing substantially to childhood morbidity and mortality (1).

The Oklahoma Birth Defects Registry (OBDR) is an active public health surveillance program that monitors the status of children with birth defects born to women who are Oklahoma residents, delivering in hospitals in Oklahoma or Fort Smith, Arkansas (since 1995). The OBDR operates under 63 O.S. Section 1-550.2, which gives the Oklahoma State Department of Health the responsibility for operating the registry and assuring confidentiality of the data collected. Medical record departments identify cases by use of ICD-9-CM codes for discharge diagnoses in the hospital and outpatient setting. Data abstractors collect information on all affected intrauterine fetal deaths (IUFDs) for fetuses at least 20 weeks gestation at birth, all affected live births diagnosed by 2 years of age, and identified prior to the child's sixth birthday.

Active surveillance of birth defects is limited by the hospital's accuracy in coding discharge diagnoses, and the timeliness of reporting the diagnoses to the OBDR. Children whose birth defects were identified after discharge from the birth hospital, and did not require hospital admission or receive hospital outpatient services, will not be identified by the OBDR.

Figure 1. Five Leading Causes Infant Mortality: United States, 1994



Data Analysis

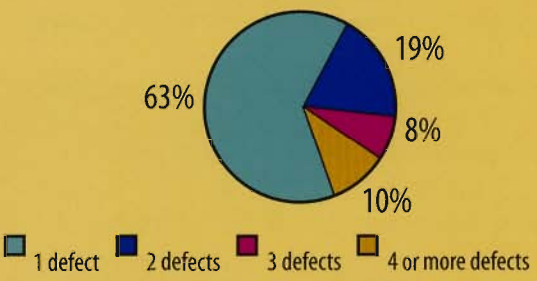
Data Analysis

Data representing a child and its parents were collected using an abstracting form developed by the OBDR. An Access database was created and the data were coded appropriately. For analysis purposes, the data were imported without confidential identifiers to SAS version 8.1. The data were checked for data entry errors and other discrepancies. Each child could have as many as seven birth defect codes listed. For data categorized by infant/fetus, an infant/fetus was included in each category for which a birth defect occurred. We calculated the prevalence of specific birth defects using a modified 6 digit ICD-9-CM code system to identify cases (2). We used the number of live births alone or the number of live births plus stillbirths for the denominator of rates due to specific code and/or classification variable. The rates presented use a constant multiplier of 1,000 or 10,000 live births (or live births plus stillbirths). The OBDR rates can be compared to national and other state birth defects rates to determine if differences in prevalence exist. The Oklahoma State Department of Health, Center for Health Statistics provided all population and vital statistics data. Analyses were obtained for gender; race; geographical region; maternal smoking, medication use and age; source of diagnosis; prenatal care; number of defects/child; and collection year as well as by grouping by specific system and type of birth defects.

Overview of Children with Birth Defects in Oklahoma

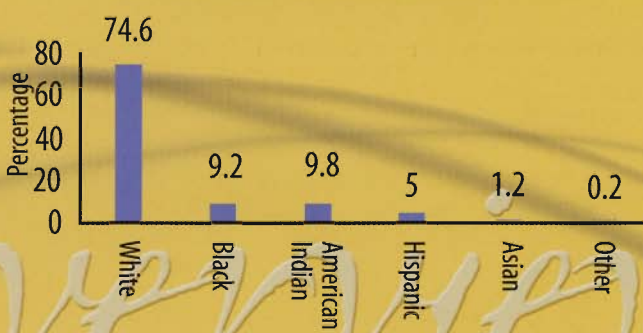
From 1994 through 1998, 8,950 children were reported to the OBDR. Approximately 63 percent of the children had one birth defect, 19 percent had two defects, eight percent had three defects, and 10 percent had four or more defects (Figure 2). Of all infants with birth defects born in Oklahoma from 1994-1998, White non-Hispanic represents the largest percentage (74.6%) (Figure 3). The remaining racial/ethnic distribution is as follows: Black non-Hispanic (9.2%), American Indian (9.8%), Hispanic (5%), Asian (1.2), and Other 0.2%.

Figure 2. Percentage of Number of Birth Defects per Child for Years 1994 - 1998 in Oklahoma



The rates of birth defects per 1,000 births have been calculated for six regions of Oklahoma (Figure 4). The overall state rate was 37.9. The rate for Oklahoma County was 42.6, and the rate for Tulsa County was 40.9, both above the state rate. The Southeast region is slightly above the state rate at 38.3. All other regions of the state fall below the state rate: 36.0 Northeast, 33.7 Southwest, and 26.6 Northwest.

Figure 3. Percentage of Infants with Birth Defects by Race for the Years 1994 - 1998 in Oklahoma



Overview

Rates

Figure 4. Rates of Infants with Birth Defects per 1,000 Births by Six Oklahoma Regions for 1994 - 1998

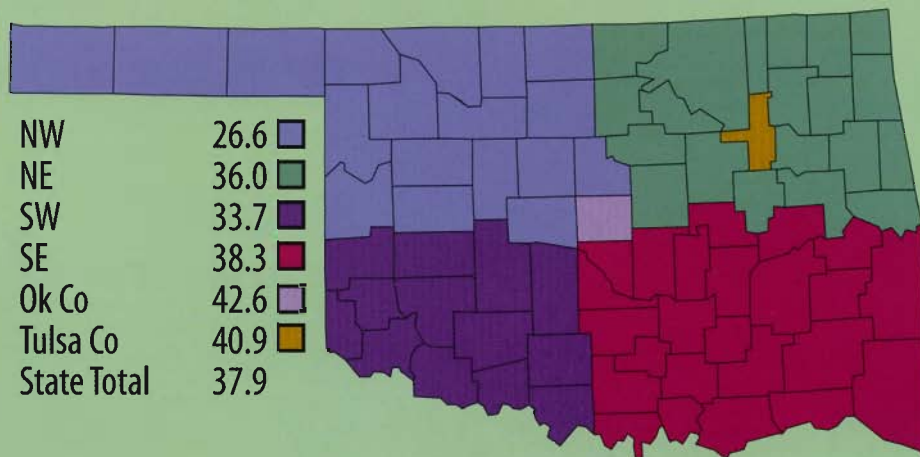
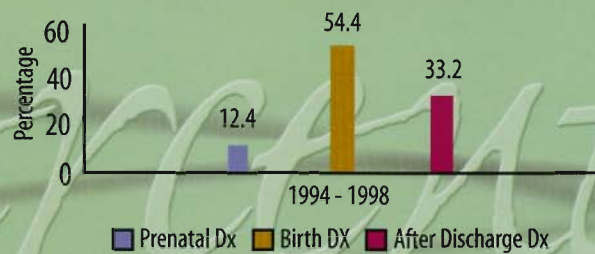


Figure 5 identifies the source of diagnosis of birth defects by three categories: prenatal diagnosis, birth diagnosis, or after discharge diagnosis from 1994 through 1998.

The prenatal diagnosis category indicates the diagnosis was made during the pregnancy, prior to delivery. The birth diagnosis category refers to infants that were diagnosed before they left the birth hospital. This category also includes infants transferred to a tertiary hospital from the birth hospital, and the diagnosis was made at the tertiary hospital. The after discharge diagnosis category refers to children that were diagnosed after they left the birth hospital. The diagnosis could have been made during a subsequent hospital admission, or at a hospital outpatient visit. From 1994 to 1998, the percentage of infants diagnosed in the prenatal period increased from 11.3 to 14.5 (data not shown). The percentage of infants diagnosed with birth defects at birth decreased slightly from 54.7% in 1994 to 52.2% in 1998. The percentage of birth defects diagnosed after discharge remained virtually unchanged, 33.8% in 1994 and 32.4% in 1998.

Figure 5. Percentage of Infants with Birth Defects by Source of Diagnosis for 1994 - 1998 in Oklahoma



Percentage

Overview

Overview of Mothers Giving Birth to Infants with Birth Defects

The prevalence rate of birth defects per 1,000 total births was analyzed by mother's age and the following age ranges: less than 20 years, 20 to 24 years, 25 to 34 years, and 35 years and greater (data not shown). The rate of birth defects exhibits little variability between age ranges. Mothers 35 years and greater had the highest rate of birth defects for all five years, at 42.0. Mothers 25-34 years had the lowest rate of birth defects at 37.0. Mothers less than 20 years and 20-24 years had similar rates at 38.1 and 37.9, respectively. A review of literature by Makinson found no evidence of an overall increase in congenital malformations among babies born to teenagers (3). Smoking during pregnancy has been linked to low birth weight, sudden infant death syndrome, and pre term

labor (4). Studies have also linked smoking to birth defects, though somewhat inconsistently. Khoury et al found an association between cigarette smoking and oral clefts (5). Li et al found maternal smoking was associated with urinary tract defects (4). A study by Czeizel et al indicated an association between smoking during pregnancy and the occurrence of terminal transverse limb deficiencies (6). Woods et al, in a study on maternal cigarette smoking and birth defects, found the offspring of smokers had a 56 percent increase in the frequency of cardiovascular anomalies compared to those born to nonsmokers (7). Table 1 compares the percent of mothers who had a child with a birth defect (OBDR mothers) that smoked at any time during their pregnancy, with mothers who responded to the Pregnancy Risk Assessment Monitoring System

(PRAMS) survey. PRAMS surveys women that have given birth in Oklahoma within the past six months to identify risk factors that contribute to low birth weight and infant mortality. PRAMS used data for mothers that smoked during the last trimester of their pregnancy. Mother's age group was calculated for 1994 through 1998. Consistently, for every age group, the OBDR mothers had a higher percent of smoking than the PRAMS mothers, with the exception of 35+ years of age in 1998. The higher percentage among OBDR mothers could be due to identifying smoking status at anytime during pregnancy, compared to the last three months in PRAMS. All age groups of OBDR mothers decreased their percentage of smoking from 1994 to 1998.

Table 1. Percent of Mothers Smoking During Last Trimester for OBDR and State by Mothers Age for 1994-1998 in Oklahoma

Mother Age	1994		1995		1996		1997		1998	
	OBDR %	PRAMS %	OBDR %	PRAMS %	OBDR %	PRAMS %	OBDR %	PRAMS %	OBDR %	PRAMS %
<20	33.0	26.2	31.2	27.0	28.2	21.3	38.3	30.8	32.3	26.6
20-24	29.6	23.9	27.0	26.3	29.1	18.3	27.8	15.9	26.8	22.5
25-34	25.2	20.9	22.3	19.4	22.1	18.7	20.7	18.7	20.9	11.7
35+	27.1	22.2	35.8	22.4	29.7	23.9	22.8	21.0	20.3	26.7

Overview

Use of medications during pregnancy among women delivering an infant with birth defects has steadily increased from 64.5 % in 1994 to 86.9 % in 1998 (Figure 6). Medications identified by the OBDR include prescription, over-the-counter, vitamin supplements, and herbs. The rise could be due to physicians prescribing more medications over time, increased documentation of medications in the prenatal records, or both. Some medications are known teratogens (8). Care should be taken in use of medications in the periconception period (the time just before and after conception), and during the first nine weeks of pregnancy when organogenesis is occurring. Figure 7 shows the average percentage from 1994 through 1998 of mothers taking medications by the number of medications.

For women classified by number of medications, the highest percentage (26.7) of women were in the group taking four or more.

Overview of Birth Defects

Examination of the distribution of birth defects in Oklahoma by major organ systems from 1994 through 1998 identifies cardiovascular defects as the largest category at the 29.8% (figure 8). Musculoskeletal defects comprise 24.3% of the defects, followed by gastrointestinal defects at 13.95%, and central nervous system at 8.5%.

Figure 6. Percentage of Mothers Taking Medication and Giving Birth to an Infant with Birth Defects for 1994 - 1998 in Oklahoma

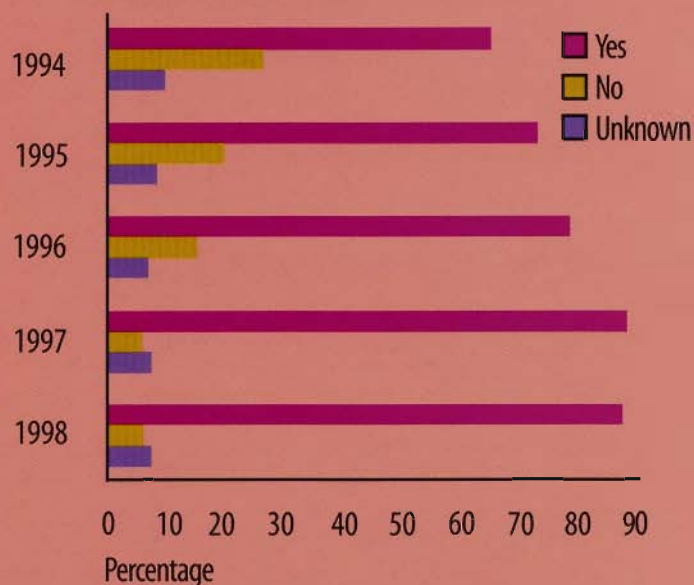
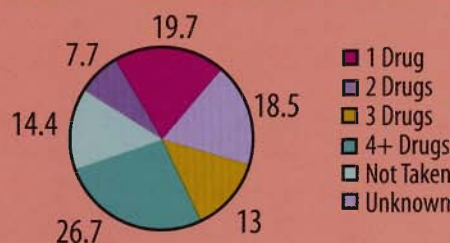


Figure 7. Percentage of Mothers Taking Medication and Giving Birth to an Infant with Birth Defects by Number of Medications Taken for 1994 - 1998 in Oklahoma



Birth Defects

Central Nervous System Defects

Figure 9 identifies the five-year rate trends for select central nervous system defects.

The neural tube forms the brain and spinal cord. Failure of the neural tube to close properly, which occurs at approximately 26 to 28 days after conception, results in neural tube defects (NTDs) (8). When the neural tube fails to close at the top of the spine, anencephaly occurs. Anencephaly literally means absence of the brain. These babies are stillborn or die a few hours after birth. The overall rate of anencephaly from 1994-1998 is 3.0 per 10,000 live births and stillbirths. When the neural tube fails to close along the spine, the resulting defect is called spina bifida. Varying degrees of neurological deficit occur, depending upon the location of the defect on the spine. Children can have minimal complications, require braces and or canes to walk, or be confined to a wheelchair. They may also lack bowel and bladder control. In general, the higher the defect location along the spine, the more organ systems are affected. The overall rate of spina bifida from 1994-1998 is 6.3 per 10,000 live births and stillbirths. Figure 9 identifies the five-year trend for annual spina bifida rates.

The OSDH recommends all women of childbearing age take a multivitamin containing 400 micrograms (0.4 mg.) of folic acid, eat foods fortified with folic acid, and consume a balanced diet. This recommendation can help prevent 50 to 70 percent of NTDs (9). Hydrocephalus is the abnormal enlargement of the head caused from impaired circulation and absorption of cerebrospinal fluid (8). Treatment is through a surgical by-pass (shunt), which drains fluid into the stomach. If untreated, mental retardation results. With shunting, 80% of the children reach 5 years of age. Learning disabilities are associated with hydrocephalus. The overall rate of hydrocephalus from 1994-1998 is 9.3 per 10,000 live births and stillbirths. Figure 9 provides rate trends for individual years.

The rates for selected central nervous system defects, hydrocephalus, spina bifida and anencephaly, have decreased steadily from 1994 to 1998.

Figure 8. Percentage of Birth Defects by Major Organ System for 1994 - 1998 in Oklahoma

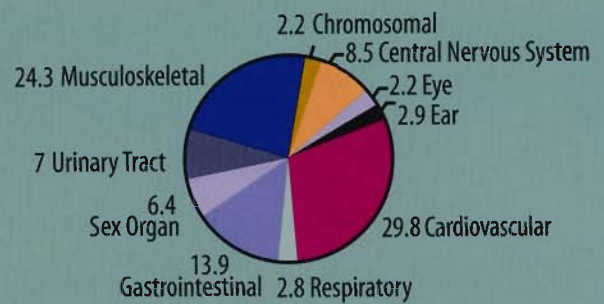
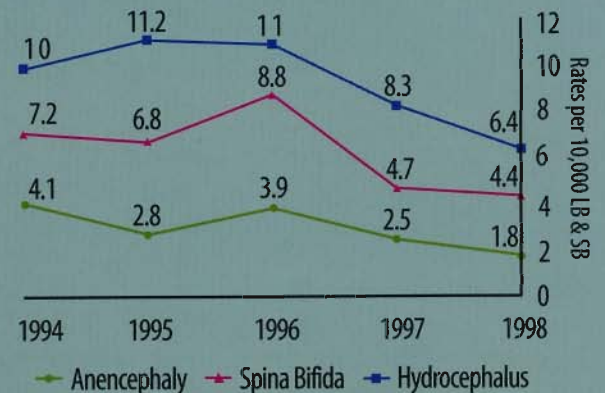


Figure 9. Rates per 10,000 Live Births and Stillbirths for Selected Central Nervous System Defects from 1994 - 1998 in Oklahoma

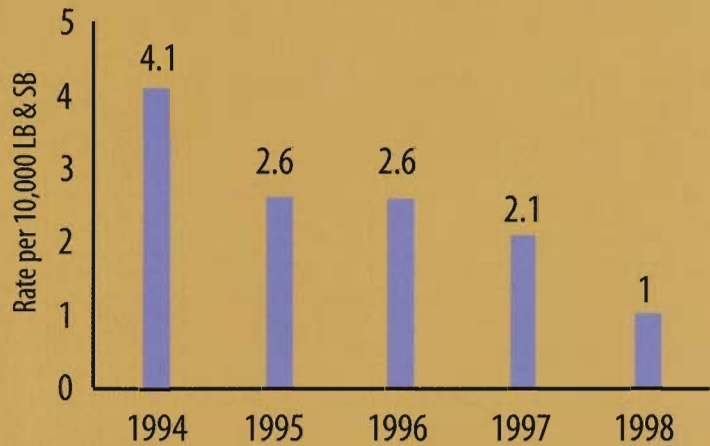


Birth Defects

Congenital Cataracts

In congenital cataracts the eye lens is opaque and can vary from insignificant to severe visual impairment. Known causes of cataracts include inheritance by dominant transmission, or teratogenic agents such as rubella virus; however, in most cases the cause is unknown (8). The rate of occurrence is undetermined. The overall rate of congenital cataracts from 1994 -1998 in Oklahoma is 2.5 per 10,000 live births and stillbirths. Refer to Figure 10 for yearly rates, and five-year trends.

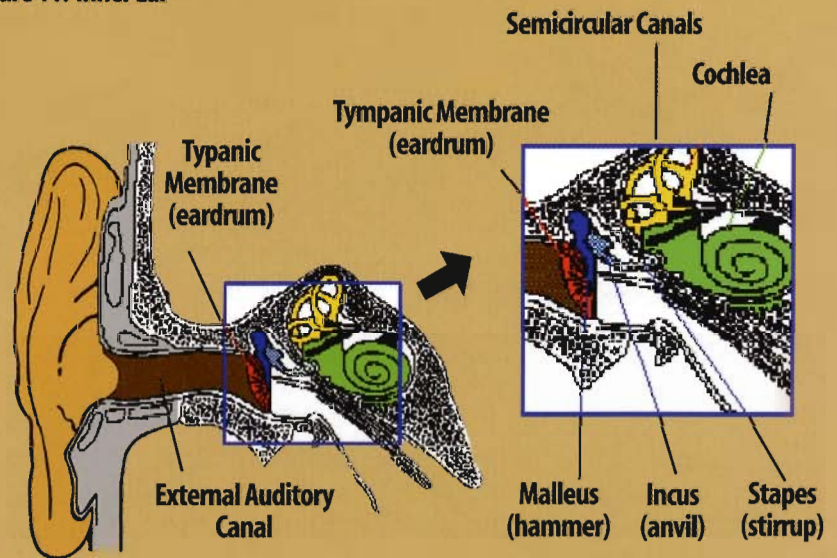
Figure 10. Rates of Congenital Cataracts per 10,000 Live Births and Stillbirths from 1994 - 1998 in Oklahoma



Anomalies of the Inner Ear

The ear begins to develop at four weeks after conception. The inner ear is the first to develop and the outer ear is the last to develop. Defects of the inner ear can cause hearing loss. Figure 11 diagrams the ear, highlighting the inner ear. Late detection of inner ear malformations can lead to consequences, such as speech delay, progressive hearing loss, and life-threatening meningitis (10). The rate for 1994-1998 in Oklahoma is 3.7 per 10,000 live births and stillbirths.

Figure 11. Inner Ear

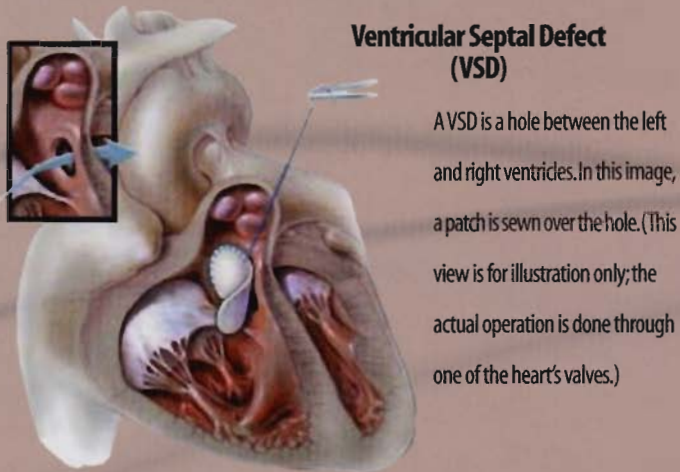
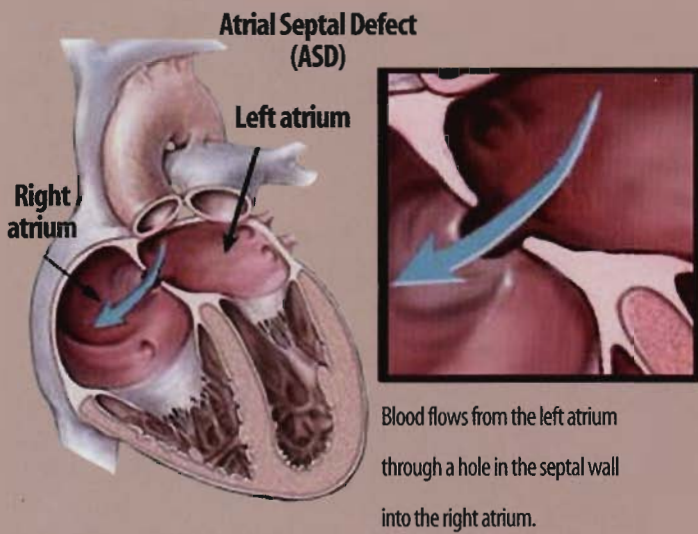
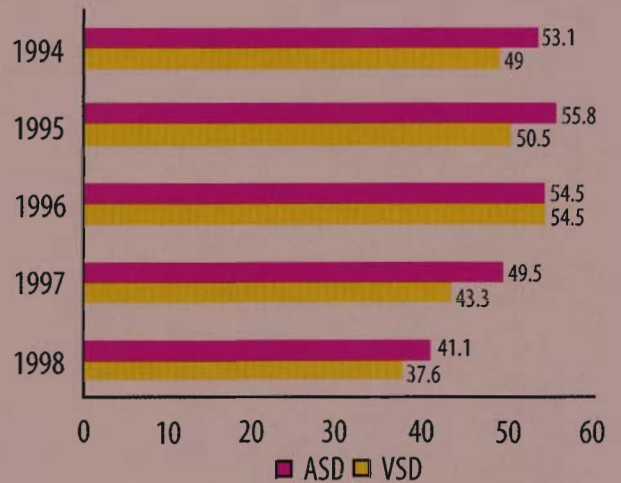


Birth Defects

Heart Defects

Congenital heart defects are present in six to eight cases per 1,000 births (8). The atrial septal defect (ASD) is a communication (hole) between the left and right atrium. The overall rate from 1994-1998 in Oklahoma is 50.6 per 10,000 live births and stillbirths (Figure 12). The ventricular septal defect (VSD) is caused by a communication (hole) between the left and right ventricles. The VSD is the most common type of heart disease, comprising 25 percent of heart defects (8). An estimated 30 and 50 percent of VSDs close spontaneously in the first year of life. The rate of VSDs from 1994-1998 in Oklahoma is 46.8 per 10,000 live births and stillbirths (Figure 12).

Figure 12. Rates of ASD and VSD per 10,000 Live Births and Stillbirths from 1994 - 1998 in Oklahoma

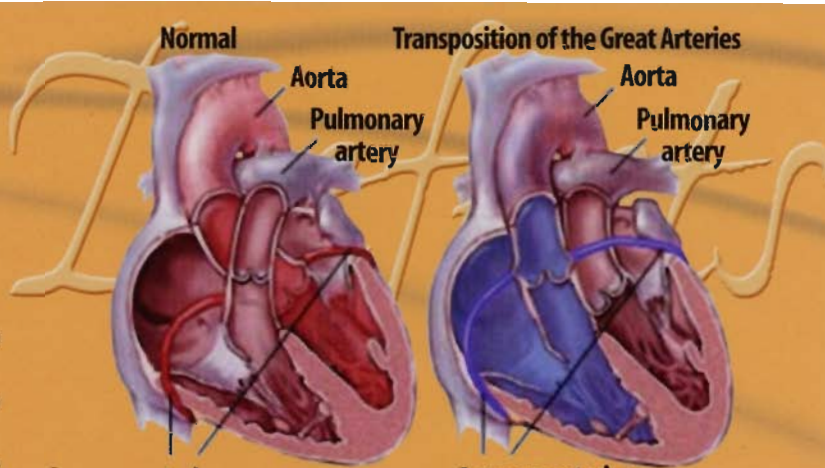


Birth

Transposition of the great arteries is the most common cause of cyanotic heart disease in newborn infants (8). The defect occurs when the aorta arises from the right ventricle (normally arises from the left ventricle) and the pulmonary trunk arises from the left ventricle (normally the right ventricle). The overall rate from 1994–1998 in Oklahoma was 4.9 per 10,000 live births and stillbirths. Refer to Figure 13 for the five-year trend and annual rates.

Coarctation of the aorta is characterized by constriction of varying lengths of the aorta. It occurs twice as often in males as females. In 70% of the cases, it is associated with a bicuspid aortic valve. From 1994–1998 in Oklahoma, the average rate was 3.6 per 10,000 live births and stillbirths. Figure 13 identifies the rate trend for the five-year period.

Hypoplastic left heart syndrome is a group of congenital anomalies consisting of hypoplasia or atresia of the left ventricle and of the aortic or mitral valve, or both. Hypoplastic left heart also includes hypoplasia of the ascending aorta. The defect causes respiratory distress and extreme cyanosis, with cardiac failure and death in early infancy. The overall rate from 1994–1998 in Oklahoma was 3.8 per 10,000 live births. Refer to Figure 13 for rate trends for the five-year trend.



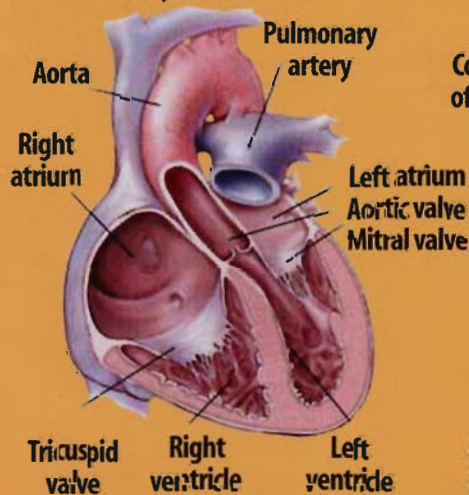
Coronary arteries

Oxygen-poor blood travels from heart's right side to the pulmonary artery and to the lungs. The blood (oxygen-rich) returns to the heart's left side and is pumped out to nourish the heart (via the coronary arteries) and the rest of the body (via the aorta).

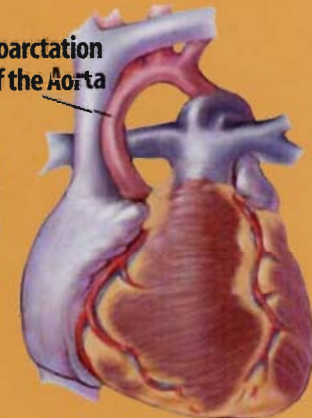
Coronary arteries

Oxygen-poor blood travels from heart's right side directly to the coronary arteries and the aorta. Because the blood does not go to the lungs for fresh oxygen, it cannot nourish the heart or the rest of the body.

Normal Baby Heart

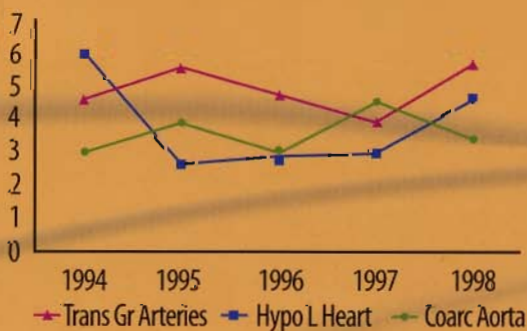


Coarctation of the Aorta

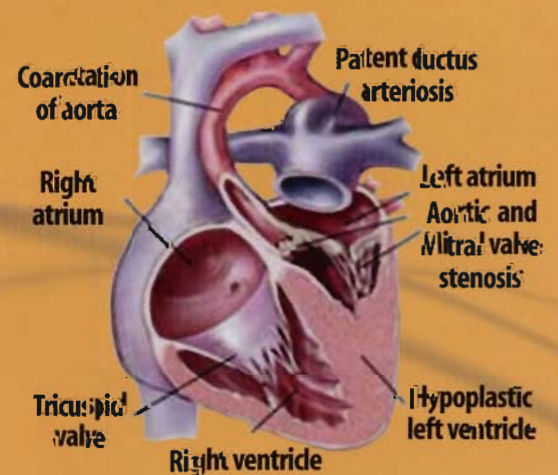


Coarctation of the aorta occurs when the aorta, the body's main artery, is pinched or narrowed, restricting blood flow to the body.

Figure 13. Rates of Transposition of the Great Arteries, Hypoplastic Left Heart Syndrome, Coarctation of the Aorta per 10,000 Live Births and Stillbirths in Oklahoma



Hypoplastic Left Heart



Birth Defects

The aortic valve is at the entrance of the aorta, where it arises from the left ventricle (11). Aortic stenosis results from the obstruction of the left ventricular outflow.

The most common aortic obstruction is in the aortic valve, where the edges of the valve may be fused or bicuspid instead of tricuspid. In approximately 65% of the cases, the aortic valve is bicuspid. In about 15% of the cases, the aortic valve is fused. In 10 to 20 % of cases, the aortic valve has one single leaflet and is dome shaped.

The combined rate for 1994-1998 in Oklahoma was 7.0 per 10,000 live births and stillbirths.

Pulmonary valve atresia results from abnormal formation of the valve leaflets during fetal cardiac development resulting in obstruction of blood flow from the right ventricle to the pulmonary artery (11). Pulmonary valve stenosis represents ten percent of congenital heart defects. The combined rate for atresia and stenosis of the pulmonary valve for all five years was 8.2 per 10,000 live births and stillbirths. Figure 14 provides the rate trends for each individual year, 1994-1998 in Oklahoma.

The tricuspid valve is between the right atrium and the right ventricle. Tricuspid valve atresia occurs when the valve between the right atrium and left ventricle fails to form during fetal development. The right ventricle is hypoplastic, with ventricular septal defect and small atrial septal defect. There is no blood flow between the right atrium and right ventricle. The average rate from 1994-1998 in Oklahoma was 5.3 per 10,000 live births and stillbirths.

Heart and Valves

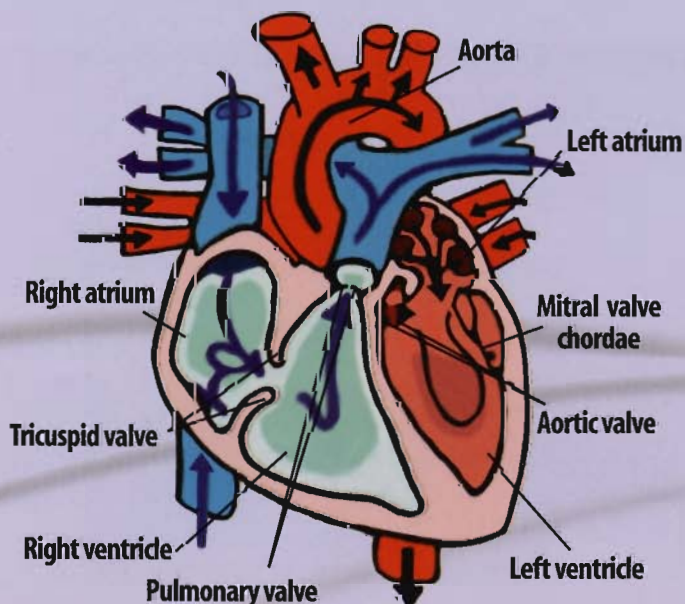
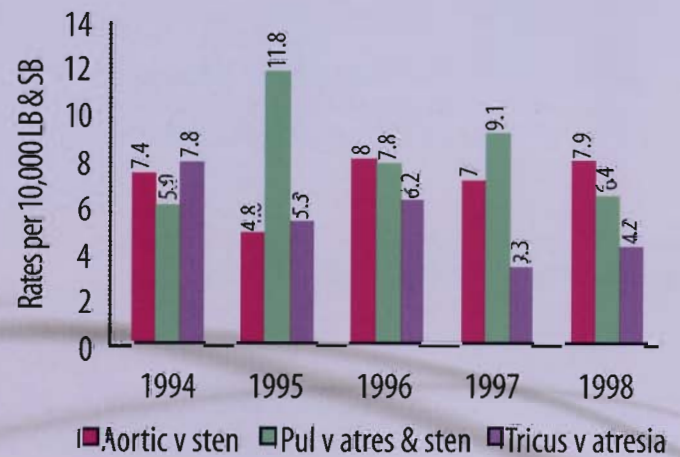


Figure 14. Rates for Selected Cardiac Valve Defects per 10,000 Live Births and Stillbirths in 1994 - 1998 in Oklahoma

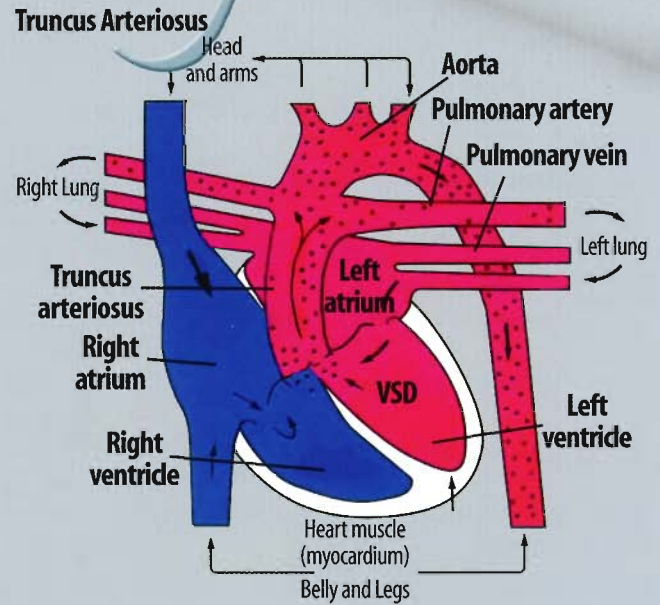


Birth Defects

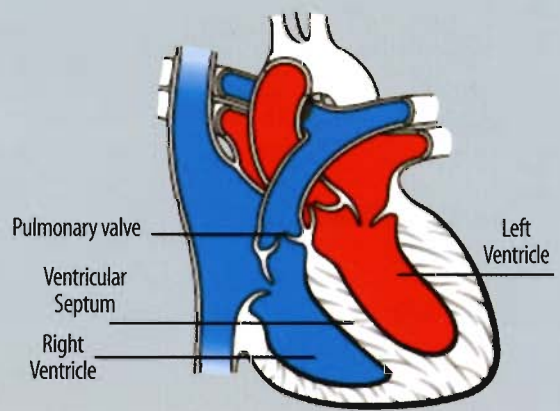
Truncus arteriosus occurs when the truncus arteriosus fails to divide into the aorta and the pulmonary trunk. The combined rate for 1994-1998 was 1.0 per 10,000 live births and stillbirths in Oklahoma.

Tetralogy of Fallot is a combination of four congenital heart defects: pulmonary stenosis, ventricular septal defect, dextroposition of the aorta, and right ventricular hypertrophy. It is the most common cyanotic congenital heart defect, approximately 9% of all heart defects (11). The rate for 1994-1998 was 3.9 per 10,000 live births and stillbirths in Oklahoma.

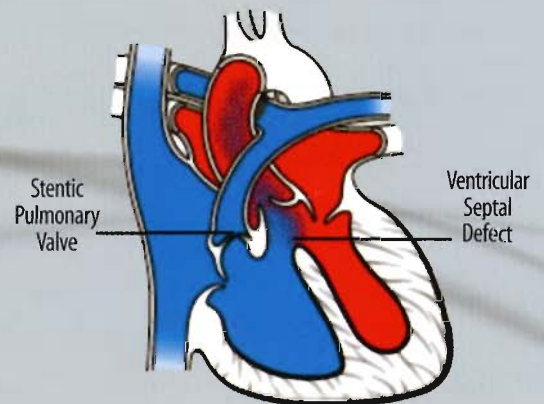
Endocardial cushion defect is thought to be caused by abnormal development of the embryonic atrioventricular endocardial cushions. There is usually a large interatrial and/or interventricular communication present. Approximately 15% of Down syndrome patients have some form of endocardial cushion defect. The average rate for 1994-1998 was 3.8 per 10,000 live births and stillbirths in Oklahoma.



Tetralogy of Fallot
Normal



Defect



Birth Defects

Respiratory Tract Defects

Choanal atresia is the congenital bony or membranous occlusion of one or both choanae (nasal passage) due to failure of the embryonic bucconasal membrane to rupture.

The newborn infant gets into respiratory distress whenever trying to breathe with the mouth closed. When crying, the infant can breathe because the mouth is open. Feeding is a problem, because the infant has to stop feeding to breathe. The combined rate for 1994 through 1998 is 2.2 per 10,000 live births and stillbirths.

Laryngomalacia may become obvious within the first two weeks of life, with noisy inspiratory breathing. The epiglottis, which protects the airway when the child feeds, also partially obstructs the airway during breathing. The partial obstruction is the source of "noise" with breathing. This type of "noisy breathing" is known as stridor and has a high pitched, harsh quality. The stridor is usually more prominent when the infant is lying on his/her back, crying, feeding, excited or has a cold. Laryngomalacia is usually a self-limited process that the infant will "outgrow" between 12 and 18 months of age.

Surgery to trim the epiglottis may be necessary in certain cases.

The average rate for 1994 - 1998 is 12.4 per 10,000 live births and stillbirths.

Cleft Lip and Palate

Cleft lip can be incomplete or complete clefts of the upper lip, and may extend into the hard and soft palate. In some cases, the cleft may include the nose of the affected side.

Cleft palate can include cleft of the soft palate, cleft of the soft and hard palates, or submucous cleft palate. In addition to needing plastic surgery to repair the opening, these children may have problems with feeding, hearing, speech, and psychological development as they grow. In Oklahoma, the rate from 1994-1998 for both cleft lip and palate was 7.8 per 10,000 live births and stillbirths (rate includes syndromes). Figure 15 identifies the rates for cleft lip, cleft palate, and both from 1994-1998 for live births and stillbirths.

Figure 15 identifies the rates for cleft lip, cleft palate, and both from 1994-1998 for live births and stillbirths.

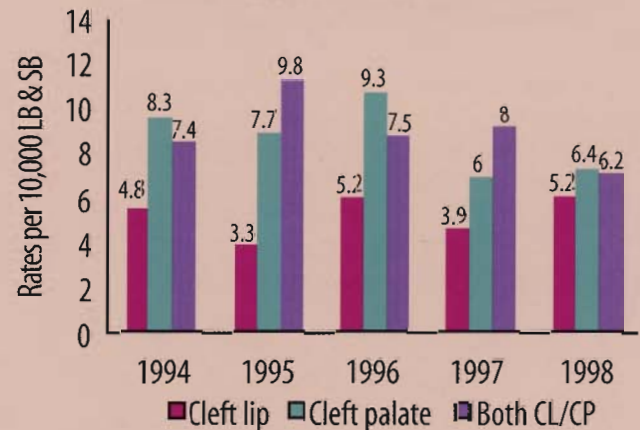
Cleft Palate



Cleft Lip



Figure 15. Rates for Cleft Lip and/or Cleft Palate per 10,000 Live Births and Stillbirths for 1994 - 1998 in Oklahoma



Birth Defects

Gastrointestinal Defects

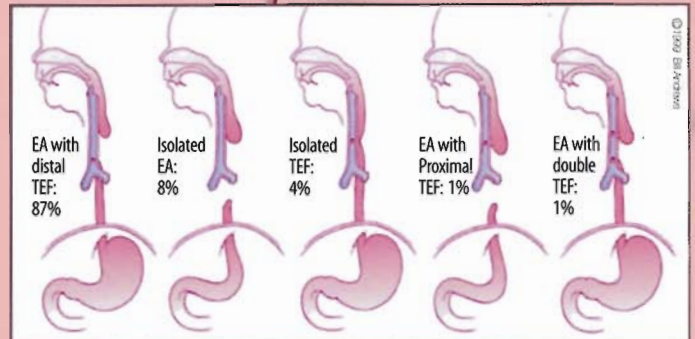
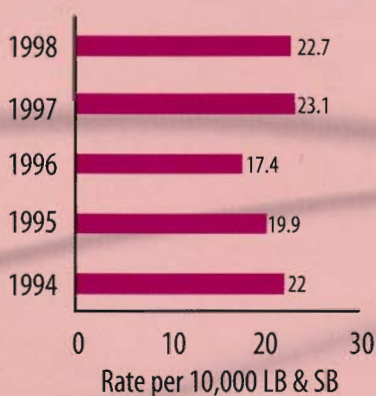
Esophageal atresia is a defect where the esophagus does not connect to the stomach, but ends in a pouch. When this is present, nothing the baby swallows reaches the stomach.

Tracheoesophageal fistula results in a fistula (abnormal communication) connecting the trachea and the esophagus, allowing stomach contents to get into the lungs. This can cause breathing problems, and even pneumonia. The rate for esophageal atresia and tracheoesophageal fistula in Oklahoma was 3.0 per 10,000 live births and stillbirths from 1994 -1998.

After inguinal hernia, congenital hypertrophic pyloric stenosis is the most common condition requiring surgery during the first few months of life. Pyloric stenosis is the narrowing of part of the stomach (the pylorus) that leads into the small intestines (8). This narrowing occurs because the muscle around the pylorus has grown too large. Most babies with pyloric stenosis begin to vomit during the second to third week of life. The cause of the increase in the size of the circular muscle of the pylorus is unknown. The defect occurs in 1 of every 500 births. Males are affected four - five times more often than females. The rate of pyloric stenosis for 1994 -1998 is 21.1 per 10,000 live births and stillbirths in Oklahoma.

Figure 16 identifies the individual rates for each year for the five-year period.

Figure 16. Rates of Pyloric Stenosis per 10,000 Live Births and Stillbirths for 1994 - 1998 in Oklahoma



cross section of pylorus

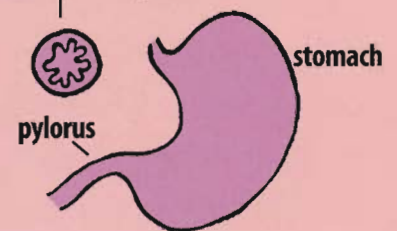


Diagram of normal stomach and pylorus. Note the cross-section showing normal pyloric opening.

cross section of pylorus

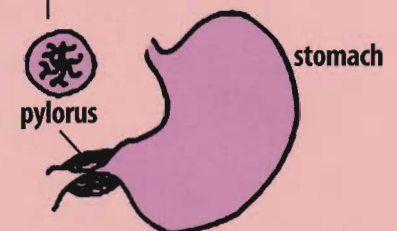
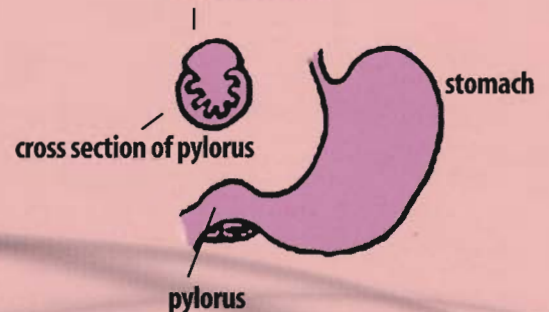


Diagram of stomach with pyloric stenosis. Note the cross-section showing how the pyloric opening is very narrowed.

area of incision



Birth Defects

Also known as congenital megacolon, Hirschsprung disease accounts for 33% of all neonatal obstructions of the colon (8). A portion of the colon lacks ganglion cells, preventing peristalsis and movement of the intestinal contents. As a result, the enlarged colon (megacolon) forms above (proximal to) the segment lacking ganglion cells. Males are four times more likely to be affected than females.

The overall rate for Hirschsprung disease from 1994 through 1998 was 2.2 per 10,000 live births and stillbirths in Oklahoma. Figure 17 identifies the yearly trend rates from 1994-1998.

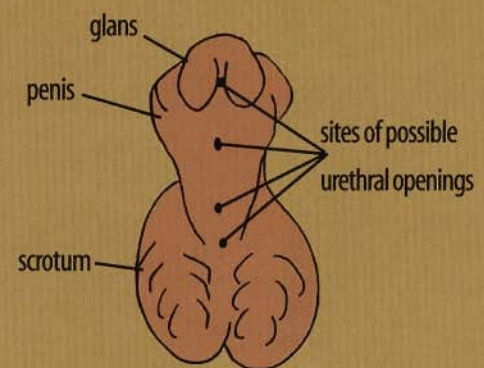
Figure 17. Rate of Hirschsprung Disease per 10,000 Live Births & Stillbirths from 1994 - 1998 in Oklahoma



Genital Organ Defects

The most common anomaly of the penis, hypospadias is characterized by the urethra opening on the underside of the penis (8). The four types of hypospadias identify where the urethral opening occurs: glandular (underside of the glans penis), penile (the underside of the body of the penis), penoscrotal (at the junction of the penis and scrotum), perineal (between the unfused halves of the scrotum).

Glandular hypospadias represents 80 percent of the cases. The incidence is 1 in 300 male infants. The average rate of hypospadias was 30.0 per 10,000 live births and stillbirths from 1994-1998 in Oklahoma. Data from the Metropolitan Atlanta Congenital Defects Program identified a rate of 40 per 10,000 total births.



Birth Defects

Urinary Tract Defects

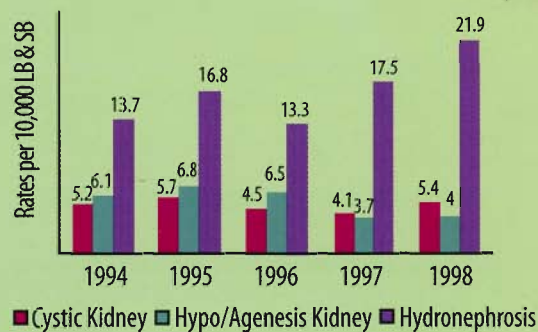
Polycystic kidney and multicystic dysplastic kidney are two types of cystic kidney disease. Polycystic kidney disease (PKD) is a genetic disorder characterized by the growth of numerous cysts filled with fluid in the kidneys. PKD cysts can reduce kidney function, leading to kidney failure. A multicystic dysplastic kidney is the result of abnormal fetal development of the kidney, where multiple cysts that vary in size replace the kidney. There is little or no normal function to this kidney. Historically, it was one of the most common causes of abdominal mass as discovered by physical examination. Currently with the use of prenatal ultrasound, most of these are discovered prior to birth. The vast majority of multicystic kidneys occur on one side (left side). Occasionally it may occur on both sides, but this is incompatible with life, primarily because the kidney is responsible for producing the amniotic fluid, which is vital to lung development. In most cases this condition isn't cause for alarm, since one properly functioning kidney is sufficient to provide the normal amount of amniotic fluid. The incidence between males and females is approximately equal. This condition is most common in the Caucasian population. The rate of cystic kidney disease in Oklahoma from 1994-1998 was 5.4 per 10,000 live births and stillbirths. Figure 18 identifies the rates for each individual year.

Renal unilateral agenesis is fairly common, occurring more often in males than females (8). The infant appears normal at birth, and often causes no symptoms, because the other kidney compensates. The condition is not clinically significant unless the one kidney becomes infected.

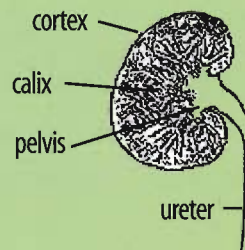
Bilateral renal agenesis is not compatible with life. Approximately 40 percent are stillborn; those infants born alive die within four hours of birth. The rate for bilateral and unilateral agenesis, dysplasia, and hypoplasia of the kidney from 1994-1998 is 5.0 per 10,000 live births and stillbirths. Figure 18 identifies the individual yearly rates for the five-year period.

Hydronephrosis is a "stretching" or dilation of the inside, or collecting part, of the kidney and is often a result of a blockage in the ureter where it joins the kidney. In infants, the amount of hydronephrosis may appear greater than the actual degree of blockage due to the "elasticity" of the young tissues. The hydronephrosis depends on the extent of the blockage and the amount of stretching of the kidney. Hydronephrosis ranges from mild, moderate to severe. The rate of hydronephrosis for the five-year period was 16.8 per 10,000 live births and stillbirths. Refer to Figure 18 for yearly rates.

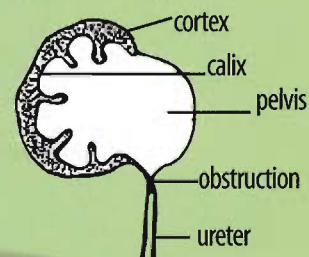
Figure 18. Rates for Selected Urinary Tract Defects per 10,000 Live Births and Stillbirths for 1994 - 1998 in Oklahoma



Normal Kidney



Hydronephrosis



Birth Defects

Musculoskeletal Defects

Clubfoot describes a range of foot and ankle abnormalities. The defects can be mild or severe and can affect one or both feet. Talipes equinovarus is the most common type of clubfoot. The sole of the foot is turned inward and the foot is inverted (8). The overall rate of clubfoot in Oklahoma from 1994 - 1998 was 20.0 per 10,000 live births and stillbirths.

Limb reduction defects involve missing tissue or bone in any part of a limb or limbs. Limb reduction defects can range in severity from missing fingers and toes to the complete absence of one or both arms and/or legs. There is no known specific cause for reduction defects. Genetic factors, environmental (such as thalidomide), and vascular disruption could all contribute (8). Figure 19 identifies the rate trends of upper and lower limb reduction defects in Oklahoma from 1994 - 1998. The overall rates for the five-year period for upper and lower defects were 4.0 and 2.0 per 10,000 live births and stillbirths, respectively.

Diaphragmatic hernia is an abnormal opening in the diaphragm that allows part of the abdominal organs to migrate into the chest cavity in fetal life. The organs could include the stomach, small intestine, spleen, part of the liver, and the kidney. The lungs fail to develop properly, which may cause life-threatening breathing problems. The defect usually occurs on the left side (8). The rate of diaphragmatic hernia from 1994 - 1998 in Oklahoma was 3.3 per 10,000 live births and stillbirths. Figure 20 identifies the rate trends for the five-year period.

Figure 19. Rates of Upper and Lower Limb Reduction Defects per 10,000 Live Births and Stillbirths from 1994 - 1998 in Oklahoma

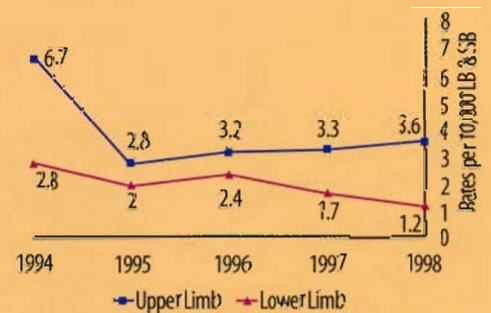
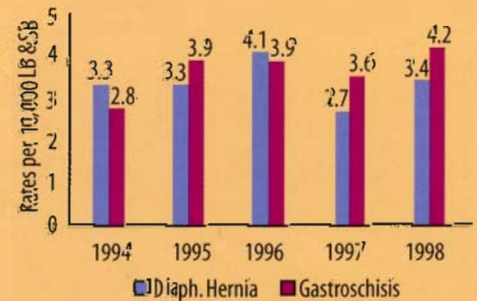
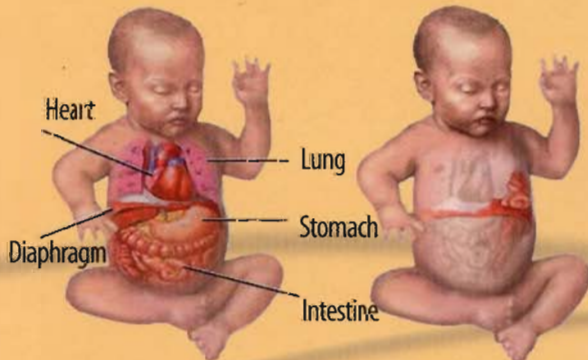


Figure 20. Rates of Diaphragmatic Hernia and Gastroschisis per 10,000 Live Births and Stillbirths from 1994 - 1998 in Oklahoma



Normal Diaphragm

Diaphragmatic Hernia



Intestine protruding through hole in diaphragm

Birth Defects

Gastroschisis, a ventral abdominal wall defect, occurs during the fourth week of pregnancy (8). The abdominal contents protrude through the defect into the amniotic cavity and the amniotic fluid. The defect usually occurs on the right side, and is more common in males than in females. The rate of gastroschisis in Oklahoma from 1994 – 1998 was 3.7 per 10,000 live births and stillbirths. Figure 20 identifies the trends in rates for gastroschisis from 1994-1998.

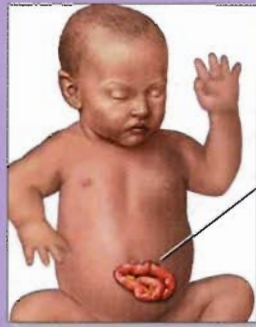
Chromosome Defects

Trisomy 21 (Down syndrome) is the presence of an additional chromosome 21 (8). In 60 percent of the cases it is caused by nondisjunction in a maternal first meiotic division. The occurrence increases with maternal age. Some of the common characteristics of Down syndrome are: poor muscle tone, slanting eyes with folds of skin at the inner corners (called epicanthal folds), hyper flexibility (excessive ability to extend the joints), short broad hands with a single crease across the palm on one or both hands, broad feet with short toes, flat bridge of the nose, short low-set ears, short neck, small head, small oral cavity (enlargement of tongue in relationship to size of mouth), short, high-pitched cries in infancy. The rate of Down syndrome from 1994 – 1998 in Oklahoma was 12.1 per 10,000 live births and stillbirths. Figure 21 identifies the rate trends for Down syndrome for the five-year period.

Trisomy 18 (Edwards syndrome) is the presence of an additional chromosome 18 (8). The usual cause is nondisjunction of chromosomes during meiosis. As with Down syndrome, the risk increases with maternal age. Characteristics include mental deficiency, growth retardation, prominent occiput, short sternum, ventricular septal defect, micrognathia (small chin), low-set malformed ears, flexed digits, hypoplastic nails, and rocker-bottom feet. Patients usually die early in infancy. The rate in Oklahoma from 1994-1998 per 10,000 live births and stillbirths was 1.7.

Trisomy 13 (Patau syndrome) is caused by an additional chromosome 13 (8). As with Trisomy 21 and 18, the usual cause is meiotic nondisjunction of chromosomes, and the risk increases with maternal age. Characteristics include mental deficiency, severe central nervous malformations, sloping forehead, malformed ears, scalp defects, microphthalmia, cleft lip and/or palate, polydactyly (extra digits). The majority of infants die by 3 months of age. The number of cases identified in Oklahoma from 1994-1998 was five, too few to calculate a rate.

Gastroschisis



Intestine protruding through abdominal wall defect

Figure 21. Rates of Down Syndrome per 10,000 Live Births and Stillbirths for 1994 - 1998 in Oklahoma



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