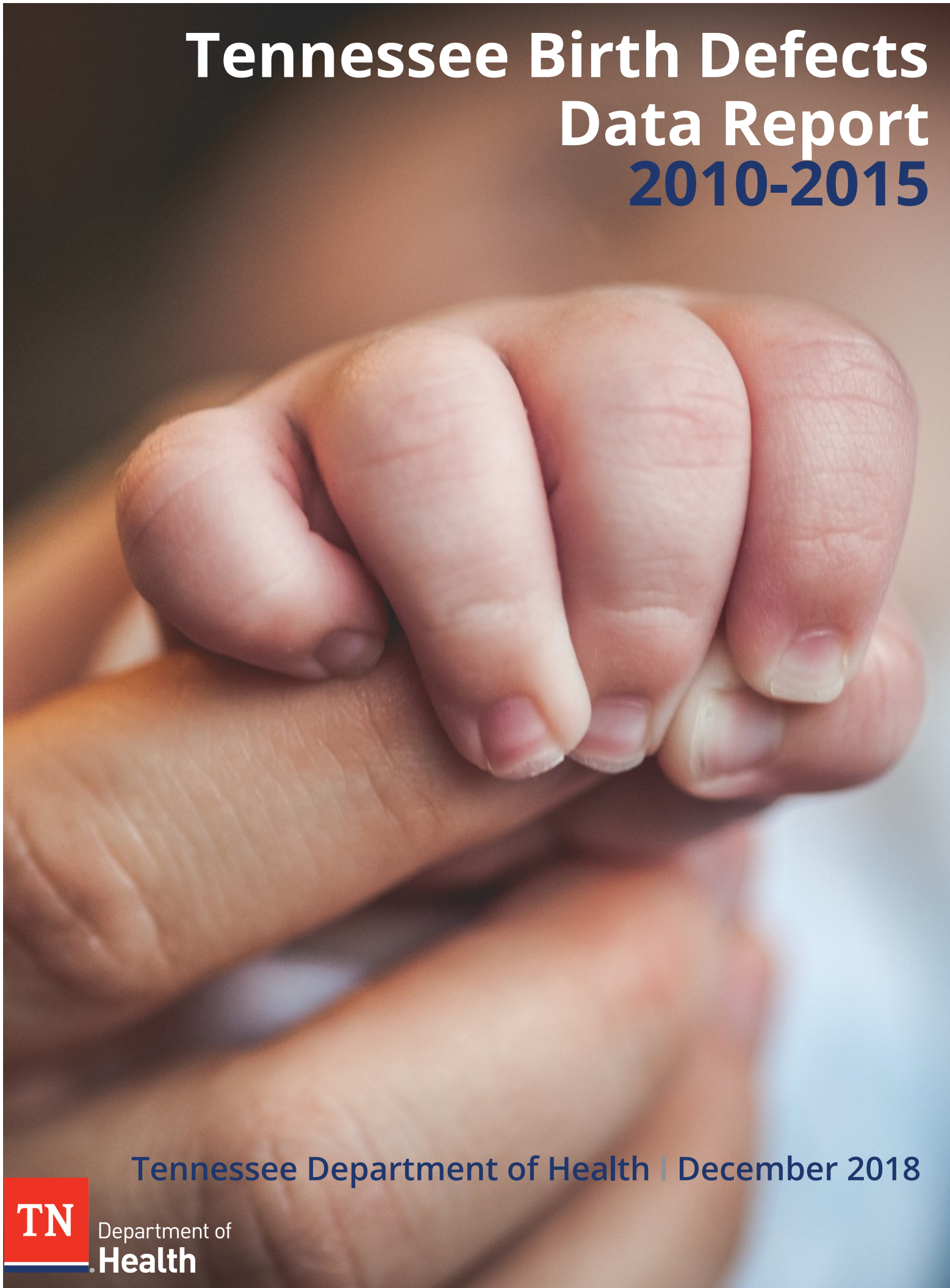
Tennessee Birth Defects Data Report 2010-2015



Tennessee Department of Health | December 2018

TN Department of Health

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Executive Summary

Birth Defects are common and costly. During the period 2010 - 2015, there were 15,325 babies (about 2,554 babies per year) diagnosed with birth defects in Tennessee. The most commonly reported birth defect was atrial septal defect (ASD), a hole or opening in the heart, followed by hypospadias, a genitourinary defect that affects males. Twenty-two percent of infant deaths in Tennessee were caused by birth defects, making it the leading cause of infant mortality. Furthermore, the CDC estimates that, in the United States, birth defects result in more than 139,000 hospital stays per year which amasses $2.6 billion in hospital costs alone.

The Tennessee Department of Health's Tennessee Birth Defects Surveillance System (TNBDSS) , as outlined in Tennessee Code Annotated§ 68-5-506, is a statewide surveillance program that identifies children with birth defects; provides information on the incidence, prevalence and trends of birth defects; informs partners and the public on birth defects and risk factors; provides guidance on prevention efforts; and, for children with specific neurologic birth defects, makes referrals for needed services such as early intervention.

This is a statewide population based birth defects report prepared by TNBDSS. The report provides details about the prevalence of 46 major birth defects and fetal alcohol syndrome for Tennessee infants born in the years 2010 through 2015. This report also includes specific information about birth defect rates by socio-demographics characteristics, known risk factors, and county of residence.

Major findings from this report include:

- Non-Hispanic Blacks had the highest prevalence of birth defects among maternal racial/ethnic groups.
- The highest prevalence of birth defects was found in the Northeast region.
- A higher prevalence of birth defects is also noted among infants of women with a 12th grade education or less and women on Medicaid compared to private insurance.
- Maternal health behaviors such as smoking and chronic health conditions such as diabetes and hypertension are associated with an increased risk of specific birth defects.

- Certain types of birth defects, especially chromosomal defects, were more common among babies who were born to mothers aged 35 years old and greater.

Key Prevention Messages:

- Birth defects surveillance programs play a key role in efforts to prevent birth defects.
- Women should see their health care providers when planning a pregnancy and begin prenatal care as early as possible.
- Women of childbearing age should consume at least 400 micrograms of folic acid every day.
- Preventing and managing chronic health conditions (like diabetes and high blood pressure) and adopting healthy behaviors before pregnancy can help prevent birth defects.
- Harmful substances (such as alcohol, tobacco, marijuana, illicit drugs) and certain medications should be avoided during pregnancy.
- It is important for women and their healthcare providers to discuss any medication use, routine vaccinations that are given before and during pregnancy, and ways to prevent infections.
- It is recommended that women plan and space pregnancies at least 18 months apart.

Introduction

What are Birth Defects?

Birth defects are changes that can affect almost any part of the body and alter how the body looks and/or functions. Birth defects are identified before birth, at birth, or after birth. Not all birth defects are the same; some are very mild while others are severe. One's life expectancy may vary depending on the severity and affected body part(s).

Why Study Birth Defects?

Birth defects are common, costly, and critical. According to the Centers for Disease Control and Prevention (CDC), an infant is born every four and half minutes with a birth defect in the United States. Nationally, nearly one out of every 33 babies is born with a birth defect and about 120,000 babies are affected by birth defects each year[1]. Birth defects cause 1 in 5 infant deaths and contribute to life-long disability. In addition to the emotional impact on affected children and their families, birth defects have financial implications for families, the healthcare system and society. Furthermore, families are often faced with missing work and subsequent wages due to medical care associated with birth defects.

Despite the prevalence and potential for significant morbidity and/or mortality, the underlying cause of most birth defects is largely unknown at the present time. This underscores the importance of birth defect surveillance, which can detect changes in the occurrence of birth defects and identify associations between exposures and birth defects. A birth defect surveillance program also plays a critical role in providing education about birth defects and risk factors, such as drinking alcohol during pregnancy, smoking during pregnancy, low blood folate levels, poorly controlled blood sugar levels in diabetic mothers, and certain maternal infections. Finally, real-time birth defect surveillance programs can ensure timely connection to key support services, such as early intervention, home visiting, care coordination, and parent support organizations.

[1] Centers for Disease Control and Prevention. Update on Overall Prevalence of Major Birth Defects–Atlanta, Georgia, 1978-2005. MMWR Morb Mortal Wkly Rep. 2008;57(1):1-5. Accessed [June 22, 2018] from https://www.cdc.gov/mmwr/preview/mmwrhtml/mm5701a2.htm.

About this Report

This is a statewide population based birth defects report prepared by the Tennessee Birth Defects Surveillance System (TNBDSS). The report provides details about the prevalence of 46 major birth defects and fetal alcohol syndrome for Tennessee infants born in the years 2010 through 2015. TNBDSS selected which birth defects to study based on national surveillance recommendations. This report also includes specific information about birth defect rates by socio-demographic characteristics, known risk factors, and county of residence. Individual birth defect counts and rates are presented in tabular form for the state overall and broken down by infant sex, maternal education, race/ethnicity, maternal age, and maternal pre-pregnancy diabetes status. Special attention is given to selected birth defects of public health significance. Birth defect rates for the thirteen Tennessee Health Department regions are also illustrated. This report also provides education on birth defects prevention, highlights prevention efforts currently underway, and suggests future directions.

In Tennessee, the most commonly reported birth defect was an atrial septal defect (ASD), a hole or opening in the upper chambers of the heart. Birth defect rates were generally equal for males and females, with the exception of the genitourinary defects such as hypospadias, which affects only males. Certain types of birth defects, especially chromosomal defects, were more common among babies who were born to mothers aged 35 years old and greater. This report highlights socio-demographic factors that are associated with an increased prevalence of birth defects. Non-Hispanic Blacks had the highest prevalence of birth defects among maternal racial/ ethnic groups. The highest prevalence of birth defects was found in the Northeast region. Higher prevalence of birth defects is also noted among infants of women with a 12th grade education or less and women on Medicaid compared to private insurance. Selected maternal health characteristics and associations with birth defects are also reviewed in this report. Babies born to mothers with pre-pregnancy diabetes are at increased risk for cardiovascular

[2] Confirmed diagnostics include: (i) fetal death cases, (ii) linked infant death cases with maternal information from Tennessee birth statistics file, (iii) linked hospital discharged cases with maternal information from Tennessee birth statistics file. The linkage is essential for confirming that the mother was Tennessee resident at the time of delivery, especially in the case of diagnoses that happened afterbirth.

[3] Matthews, TJ, et al. Infant mortality statistics from the 2013 period linked birth/infant death data set. National Vital Statistics Report. 2015. 64(9).

system birth defects, and the rates for birth defects in other organ systems are also higher in babies born to mothers with pre-pregnancy diabetes and those who did not have diabetes prior to their pregnancy.

Data Sources and Limitations

The primary data sources for this report are the Hospital Discharge Data System (HDDS) and the Birth, Death, and Fetal Death Statistical Data Systems, which are compiled, processed and stored by the Office of Vital Records and Statistics and the Office of Population Health Assessment. The HDDS contains admission-level records for all patients treated in Tennessee- licensed hospitals and their outpatient treatment and rehabilitation centers. The TNBDSS uses these records to track the 46 major birth defects and fetal alcohol syndrome. Infants' HDDS records containing diagnostic codes corresponding to the tracked birth defects are extracted, compiled, and linked with their birth certificate records. The linkages provide validity checks and add information such as maternal risk factors, demographics, and street-level geography that are not available in the HDDS. Diagnostic data are also obtained from the fetal death and death certificate data systems. For the fetal death certificate identified cases, demographic, geographic, and risk factor information are obtained from the fetal death certificate system. For the death certificate identified cases, demographic, geographic, and risk factor information are obtained from the death certificate data system. Together these sources provide statewide population-based birth defects surveillance for Tennessee.

The methodology of data collection used for this report results in a time lag for analysis, since finalization of the HDDS data occurs one year after the birth year. Additional limitations of administrative data systems involve coding. Some of the diagnostic codes used in the HDDS correspond to both the major and minor variants of a given birth defect. The previous coding system used in the HDDS, The International Classification of Diseases Revision 9 (ICD-9- CM), prevents distinguishing these differences for certain birth defects. This may have the effect of increasing rates for some of the more common birth defects, such as atrial septal defects, which are congenital heart defects, and hypospadias, a common genitourinary defect in males. Less systematically, there are simple coding errors that result in both non-cases being miscoded as having a birth defect and valid cases not being recorded as having a birth defect.

The Tennessee Birth Defects Surveillance System

According to Tennessee Code Annotated § 68-5-506, the Tennessee Department of Health (TDH) is responsible for maintaining "an ongoing program for birth defects monitoring state- wide." The goals of the birth defects registry are to report on incidence, prevalence and trends of birth defects; to provide information about potential environmental hazards associated with birth defects; to evaluate current prevention initiatives; and to provide families of children with birth defects information on public services.

Until recently, surveillance was conducted passively, primarily using data from the Hospital Discharge Data System and the Birth, Death, and Fetal Death Statistical Data Systems. An opportunity to enhance surveillance emerged after Zika virus[4] surfaced as a public health threat in the United States. In 2016, the Tennessee Department of Health (TDH) was awarded a Center for Disease Control and Prevention (CDC) Epidemiology and Laboratory Capacity grant, which has supported enhanced surveillance for specific neurologic birth defects that have been associated with Zika virus and connection to care for affected infants and their families. In January 2017, provider reporting of 23 neurologic birth defects associated with Zika was mandated by the Tennessee Department of Health. All physicians, hospitals, laboratories, healthcare providers, and other persons knowing of or suspecting a reportable disease case are responsible for reporting it to the health department. The list of reportable birth defects and the link to the reporting website can be found in Appendix A.

Monitoring birth defects is essential to ensure timely referral to services and enhance care coordination for affected children in Tennessee. Following the confirmation of a reported neurologic birth defect, referrals are made by TDH to the Tennessee Early Intervention System (TEIS), Children's Special Services (CSS) and Family Voices of Tennessee. TEIS is a voluntary educational program for families with children from birth through two years old with disabilities or developmental delays that supports families in promoting their child's optimal development, facilitates the child's participation in family and community activities,

[4] Zika virus infection during pregnancy can cause microcephaly and other neurologic birth defects. To understand more about Zika virus infection, CDC established the US Zika Pregnancy Registry and is collaborating with state, tribal, local, and territorial health departments to collect information about pregnancy and infant outcomes following laboratory evidence of Zika virus infection during pregnancy. The data collected through this registry is used to update recommendations for clinical care, plan for services for pregnant women and families affected by Zika virus, and improve efforts to prevent Zika virus infection during pregnancy.

and encourages the active participation of families by embedding strategies into family routines. The CSS program provides resources for medical and non-medical services for children with physical disabilities and special health care needs from birth to 21 years of age if certain diagnostic and financial eligibility criteria are met by the family in need. Family Voices of Tennessee, a program of the Tennessee Disability Coalition, provides emotional and educational support to the families of children with special healthcare needs, chronic illnesses or disabilities.

In addition to connecting families to needed services and monitoring the occurrence of birth defects and patterns or trends, the TN Birth Defects Surveillance System (TNBDSS) contributes to research conducted by the CDC and the National Birth Defects Prevention Network. Through collaboration with national partners, TNBDSS aim to better understand the causes of birth defects and identify strategies for reducing birth defects.

Tennessee Birth Data

In Tennessee, an average of 80,324 live births occurred to resident mothers annually during the years 2010 through 2015. During this time frame, approximately 67% of all infants born were Non-Hispanic White and 21% were Non-Hispanic Black (Table 1).

Table 1. Live Births by Maternal Race/Ethnicity, Tennessee, 2010-2015

Year	Total	Non-Hispanic White	Non-Hispanic Black	Hispanic
2010	79,345	53,288	16,489	7,121
2011	79,462	53,471	16,377	7,017
2012	80,202	54,032	16,465	6,977
2013	79,954	54,284	16,769	6,850
2014	81,609	55,375	16,941	6,982
2015	81,374	54,695	16,595	7,260

Note: Race/ethnicity categories do not sum to total as other and unknown categories are not shown.
Data Source: Tennessee Department of Health, Office of Vital Records and Statistics.

Birth Defects Prevalence in Tennessee, 2010-2015

Table 10 in Appendix B shows the case numbers and rates for the 46 major birth defects by organ system and fetal alcohol syndrome. Between January 2010 and December 2015, there were 15,325 babies diagnosed with birth defects. In addition, there were 77 infants identified with fetal alcohol syndrome during this time period. Because a baby may be diagnosed with more than one birth defect, the number of confirmed diagnosed[5] birth defects over this time period is higher – 20,758. Out of the 20,758 defects, 12,313 were cardiovascular defects which represent 59% of the total. The genitourinary system, with 3,391 defects, is the second most-affected organ system (16% of total defects). The largest single birth defect in Tennessee is atrial septal defect with a count of 7,435 or 154.3 per 10,000 live births, followed by hypospadias (n=2,580) and ventricular septal defect (n=2,415). By identifying the most common birth defects and most commonly affected organ systems, targeted prevention efforts can be developed based on known risk factors for particular birth defects.

Tables 2 and 3 show the number of cases and prevalence for selected birth defects for which national estimates are available. Table 2 features selected birth defects in Tennessee from 2010-2015, while Table 3 includes national rates from 2004-2006 (the most recent data available) for the same selected birth defects. National estimates are based on pooled data from 14 state birth defects surveillance programs. As shown in Table 2 for Tennessee, some selected conditions were more common (e.g., Down syndrome with an average of 116 cases a year for the period 2010-2015) while others were more rare (e.g. Trisomy 13 with an average of only 8 cases a year for the period 2010-2015). Down syndrome was also the most common birth defect of the selected conditions nationally for the 2004-2006 time period, with an estimated average of 6037 cases diagnosed each year in the United States. While the difference in time periods (2010-2015 vs. 2004-2006) limits the ability to make direct comparisons, it is important to identify birth defects with rates that are higher than the national average and therefore warrant particular attention.

[5] Confirmed diagnostics include: (i) fetal death cases, (ii) linked infant death cases with maternal information from Tennessee birth statistics file, (iii) linked hospital discharged cases with maternal information from Tennessee birth statistics file. The linkage is essential for confirming that the mother was Tennessee resident at the time of delivery, especially in the case of diagnoses that happened after birth.

Table 2. Frequency of Selected Birth Defects, Tennessee, 2010-2015				
Birth Defect	**Annual Number of Cases**	**Rate[1]**	**95% CI[2]**	**Frequency[3]**
Central Nervous System				
Anencephaly	12	1.54	1.21-1.93	1 in 6513
Spina bifida without anencephaly	34	4.27	3.69-4.86	1 in 2340
Encephalocele	10	1.20	0.91-1.56	1 in 8309
Eye				
Anophthalmia/microphthalmia	11	1.33	1.02-1.70	1 in 7530
Cardiovascular				
Common truncus	8	0.93	0.68-1.25	1 in 10,710
Transposition of great arteries	42	5.17	4.52-5.81	1 in 1936
Tetralogy of Fallot	48	6.02	5.32-6.71	1 in 1662
Atrioventricular septal defect	42	5.27	4.62-5.92	1 in 1897
Hypoplastic left heart syndrome	31	3.90	3.34-4.46	1 in 2564
Orofacial				
Cleft palate alone (without cleft lip)	56	6.93	6.19-7.67	1 in 1443
Gastrointestinal				
Esophageal atresia/tracheoesophageal fistula	24	3.01	2.52-3.50	1 in 3324
Rectal and large intestinal atresia/stenosis	45	5.56	4.89-6.23	1 in 1798
Musculoskeletal				
Gastroschisis	45	5.60	4.93-6.27	1 in 1785
Omphalocele	20	2.47	2.03-2.91	1 in 4050
Diaphragmatic hernia	33	4.15	3.57-4.73	1 in 2410
Chromosomal				
Trisomy 13	8	0.98	0.72-1.30	1 in 10254
Trisomy 18	14	1.72	1.37-2.13	1 in 5807
Trisomy 21 (Down syndrome)	116	14.46	13.39-15.54	1 in 691

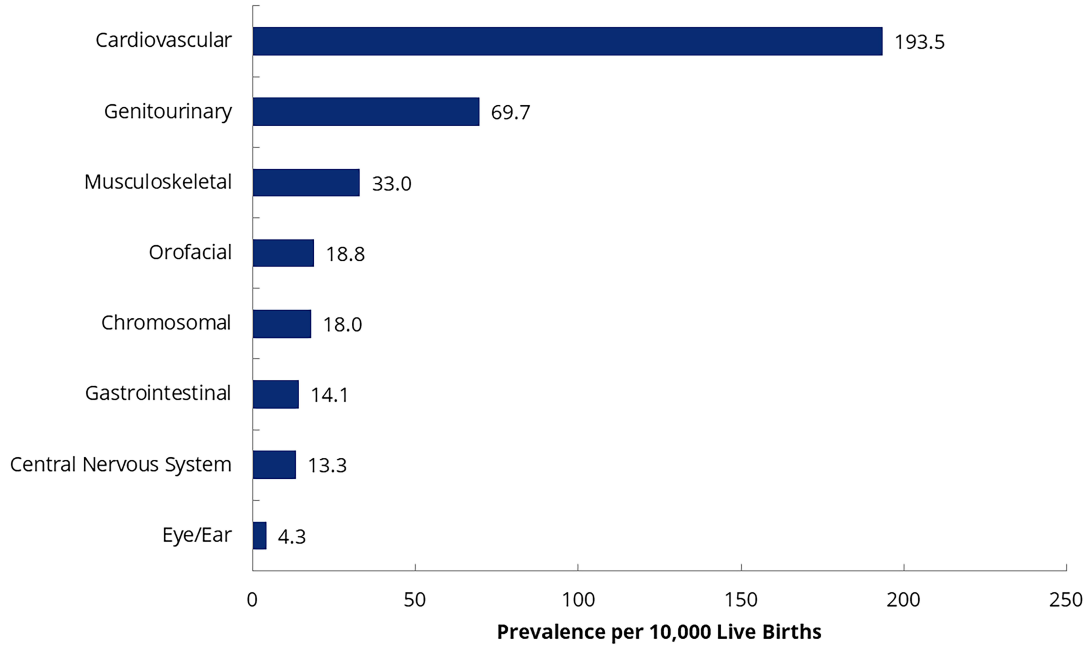
1. Rate per 10,000 Tennessee resident live births.
2. Can be interpreted as range that we are 95% confident contains the true incidence in the population. Confidence intervals for 100 cases or fewer are exact Poisson; otherwise confidence intervals are based on the normal approximation.
3. Estimated frequency of occurrence in a given number of Tennessee resident live births.
Data Source: Tennessee Department of Health, Tennessee Birth Defects Registry.

Table 3. Frequency of Selected Birth Defects, United States, 2004-2006[1]				
Birth Defect	**Annual Number of Cases**	**Rate[2]**	**95% CI[3]**	**Frequency[4]**
Central Nervous System				
Anencephaly	859	2.06	1.92-2.20	1 in 4859
Spina bifida without anencephaly	1460	3.50	3.31-3.68	1 in 2858
Encephalocele	341	0.82	0.73-0.91	1 in 12,235
Eye				
Anophthalmia/microphthalmia	780	1.87	1.73-2.01	1 in 5349
Cardiovascular				
Common truncus	301	0.72	0.63-0.81	1 in 13,876
Transposition of great arteries	1252	3.00	2.83-3.17	1 in 3333
Tetralogy of Fallot	1657	3.97	3.77-4.17	1 in 2518
Atrioventricular septal defect	1966	4.71	4.48-4.94	1 in 2122
Hypoplastic left heart syndrome	960	2.30	2.15-2.45	1 in 4344
Orofacial				
Cleft palate alone (without cleft lip)	2651	6.35	6.11-6.60	1 in 1574
Gastrointestinal				
Esophageal atresia/tracheoesophageal fistula	905	2.17	2.02-2.32	1 in 4608
Rectal and large intestinal atresia/stenosis	1952	4.68	4.45-4.91	1 in 2138
Musculoskeletal				
Gastroschisis	1871	4.49	4.28-4.69	1 in 2229
Omphalocele	775	1.86	1.73-1.99	1 in 5386
Diaphragmatic hernia	1088	2.61	2.45-2.77	1 in 3836
Chromosomal[5]				
Trisomy 13	528	1.26	1.16-1.37	1 in 7906
Trisomy 18	1109	2.66	2.50-2.81	1 in 3762
Trisomy 21 (Down syndrome)	6037	14.47	14.11-14.83	1 in 691

1. National estimates based on pooled data from 14 state birth defects surveillance programs. Estimates were standardized to the racial and ethnic distribution of the United States live birth population from 2004 through 2006. See full paper below:
Parker SE, Mai CT, Canfield MA et al: Updated national birth prevalence estimates for selected birth defects in the United States, 2004–2006. *Birth Defects Research Part A Clin Mol Teratol* 2010; 88: 1008–1016.
2. Rate per 10,000 live births.
3. Confidence intervals calculated within the assumption that the observed number of cases followed a Poisson distribution.
4. Estimated frequency of occurrence in a given number of live births.
5. Estimates for chromosomal birth defects were standardized to the United States maternal age distribution.

Figure 1 shows the prevalence rates of birth defects by organ system. Cardiovascular system defects are the most commonly diagnosed forms of birth defect in Tennessee, with a rate of 193.5 per 10,000 live births, followed by genitourinary system defects with a rate of 69.7 per 10,000 live births.

Figure 1. Prevalence of Major Birth Defects by Organ System, Tennessee, 2010-2015



Data Source: Tennessee Department of Health, Tennessee Birth Defects Registry.

Infant Mortality Data and Birth Defects in Tennessee

During the period 2010-2015, there was an average of 578 infant deaths per year (Table 4). While the infant mortality rate decreased from 2010-2013, it increased in 2014 and 2015. Non-Hispanic Black infants have a mortality rate 1.8 times higher than Non-Hispanic White infants and almost 2.5 times higher than Hispanic infants.

Table 4. Infant Deaths by Maternal Race/Ethnicity, Tennessee, 2010-2015

Year	Total		Non-Hispanic White		Non-Hispanic Black		Hispanic	
	Number	Rate[1]	Number	Rate	Number	Rate	Number	Rate
2010	626	7.9	344	6.5	228	13.8	40	5.6
2011	587	7.4	328	6.1	211	12.9	36	5.1
2012	579	7.2	337	6.2	200	12.1	26	3.7
2013	543	6.8	295	5.4	195	11.6	40	5.8
2014	562	6.9	302	5.5	211	12.5	28	4.0
2015	569	7.0	331	6.1	184	11.1	33	4.5

1. Rate per 1,000 Tennessee resident live births.
Note: Race/ethnicity categories do not sum to total as other and unknown categories are not shown.
Data Source: Tennessee Department of Health, Office of Vital Records and Statistics.

Table 5 shows the ten leading causes of infant deaths in Tennessee between 2010 and 2015. Birth defects were the leading cause of all infant deaths (22%).

Table 5. Leading Causes of Infant Death, Tennessee, 2010-2015

Rank	Cause of Death	Number of Deaths	Percent of Deaths
1	Birth defects	762	22.0
2	Preterm birth and low birthweight	541	15.6
3	Accidents	254	7.3
4	Sudden infant death syndrome (SIDS)	149	4.3
5	Maternal complications of pregnancy	118	3.4
6	Complications of placenta, cord, and membranes	88	2.5
7	Bacterial sepsis of newborn	81	2.3
8	Diseases of the circulatory system	78	2.3
9	Respiratory distress of newborn	70	2.0
10	Atelectasis (partial lung collapse)	64	1.8
10	Necrotizing enterocolitis of newborn	64	1.8
--	All other causes	1197	34.5
--	*All Causes*	*3466*	*100*

Data Source: Tennessee Department of Health, Office of Vital Records and Statistics.

Table 6 examines the top two causes of infant deaths, birth defects and prematurity/low birth weight, more closely. Among infants whose primary cause of death was a birth defect, 58% were also born premature (< 37 weeks). Preterm delivery often exacerbates the medical complications faced by infants born with major birth defect. Depending on the type of defect, infants born preterm may be at significantly greater risk of mortality compared to their counterparts delivered at term[6]. This pattern of preterm infants experiencing increased mortality has been demonstrated for neural tube defects[7], congenital diaphragmatic hernia[8], and congenital heart defects[9].

Table 6. Co-occurrence of two leading causes of infant death, Tennessee, 2010-2015

Cause of Infant Death[1]		
	Born Preterm (<37 weeks)	Born Full Term (37+ weeks)
Birth Defect	58%	42%
	Major Birth Defect Present	No Major Birth Defect Present
Preterm birth and low birth weight	2%	98%

1. Represents underlying cause of death recorded on infant's death certificate.
Note: All birth defect or preterm birth/low birth weight related deaths with corresponding birth data available were included. This represented 97 percent of all birth defect deaths and 99 percent of preterm birth/low birth weight deaths.
Data Source: Tennessee Department of Health, Office of Vital Records and Statistics; Tennessee Department of Health, Tennessee Birth Defects Registry.

6 Honein MA, Kirby RS, Meyer RE, et al. The association between major birth defects and preterm birth. Matern Child Health J 2009;13:164–75..
7 Davidoff, M. J., Petrini, J., Damus, K., Russell, R. B., & Mattison, D. Neural tube defect-specific infant mortality in the United States. Teratology 2002; 66(Suppl 1): S17–S22.
8 Cannon, C., Dildy, G. A., Ward, R., Varner, M. W., & Dudley, D. J. A population-based study of congenital diaphragmatic hernia in Utah: 1988–1994. Obstetrics and Gynecology 1996; 87(6): 959–963.
9 Tanner, K., Sabrine, N., & Wren, C. Cardiovascular malformations among preterm infants. Pediatrics 2005; 116(6): e833–e838.

Table 7 provides more detail about the gestational ages of infants who died due to birth defects. Among the infants who died due to birth defects, 42% were full-term (37+ weeks) and 26% were late-preterm (34 to <37 weeks).

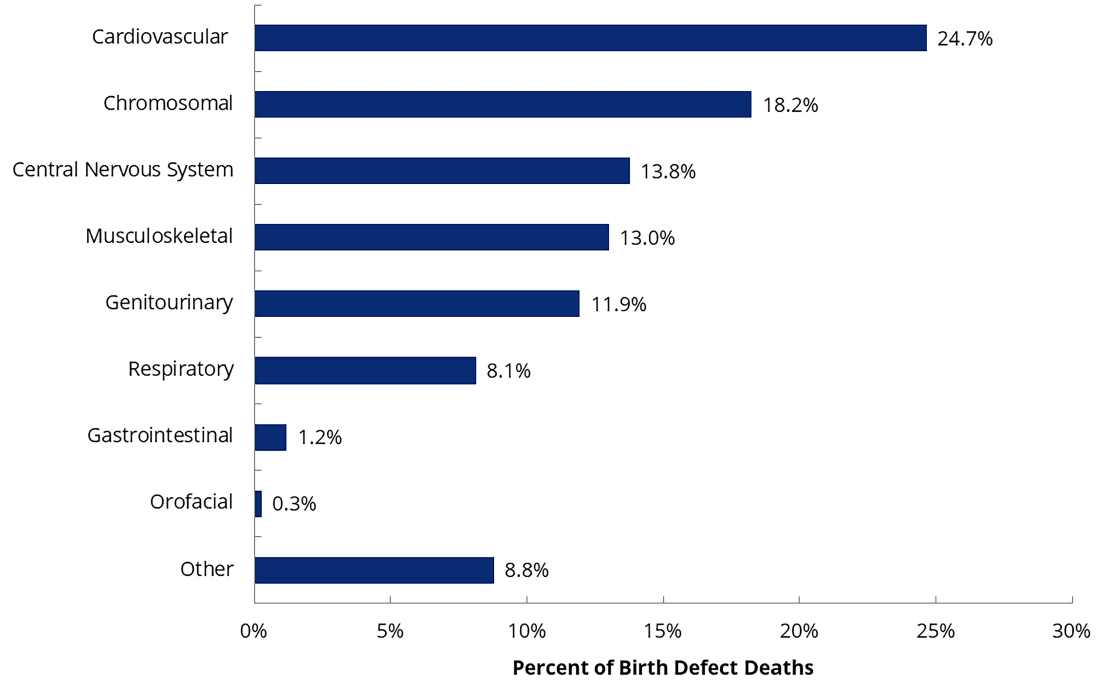
Table 7. Gestational age at birth of infant deaths caused by birth defects, Tennessee, 2010-2015

Gestational Age at Birth	Percent of Cases
<28 weeks	9
28-<32 weeks	11
32-<34 weeks	12
34-<37 weeks	26
37+ weeks	42

Note: All birth defect related deaths with corresponding birth data available were included. This represented 97 percent of all birth defect related deaths.
Data Source: Tennessee Department of Health, Office of Vital Records and Statistics.

Figure 2 demonstrates birth defect deaths by the type of defect. Heart defects were the leading cause of birth defect deaths (25% of birth defect deaths), followed by chromosomal (18%) and central nervous system (13.8%) defects. These categories may not be mutually exclusive (i.e. chromosomal birth defects may be the cause of cardiovascular and other birth defects).

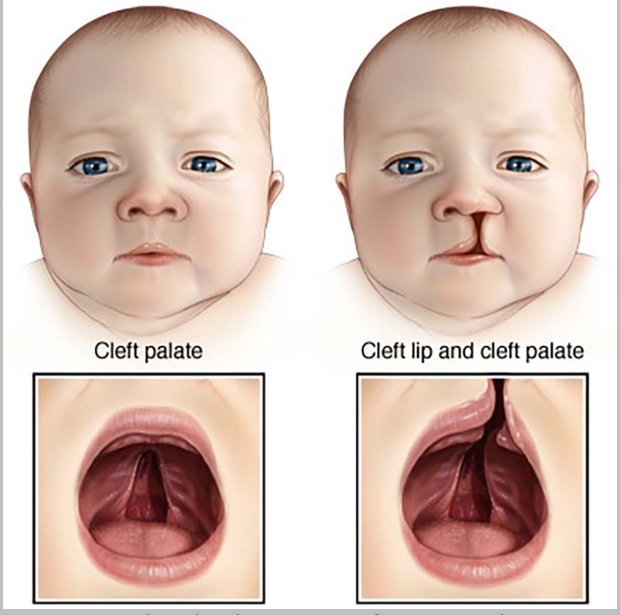
Figure 2. Birth Defect Deaths by Type of Defect, Tennessee, 2010-2015



Data Source: Tennessee Department of Health, Office of Vital Records and Statistics.

Each Tennessee Birth Defects Report will highlight a specific birth defect, with the goal of providing information on causes, risk factors, and opportunity for prevention. This year's report features orofacial clefts, which includes cleft lip and/or cleft palate.

Birth Defect Highlight: Orofacial Clefts



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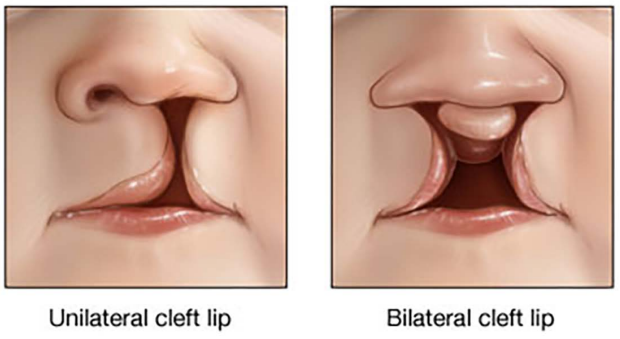
What is cleft lip and cleft palate?

Orofacial clefts include cleft lip and cleft palate, which are birth defects that occur when the structures of the mouth do not form properly during pregnancy. A cleft lip is an opening in the lip, and a cleft palate is an opening in the roof of the mouth (palate). These defects typically occur during fetal development, between four and ten weeks after conception. Orofacial clefts are often diagnosed during pregnancy by ultrasound. Management and treatment of orofacial clefts depends on the severity of the cleft, but typically involves surgical repair, dental or orthodontic care, and speech therapy.

Occurrence in the United States and Tennessee

Orofacial clefts are one of the most common types of defects. National rates are 10.63 per 10,000 live births for cleft lip with and without cleft palate and 6.35 per 10,000 live births for cleft palate alone. In Tennessee, rates for orofacial clefts are 9.94 per 10,000 live births for cleft lip with and without cleft palate and 6.93 per 10,000 live births for cleft palate alone.



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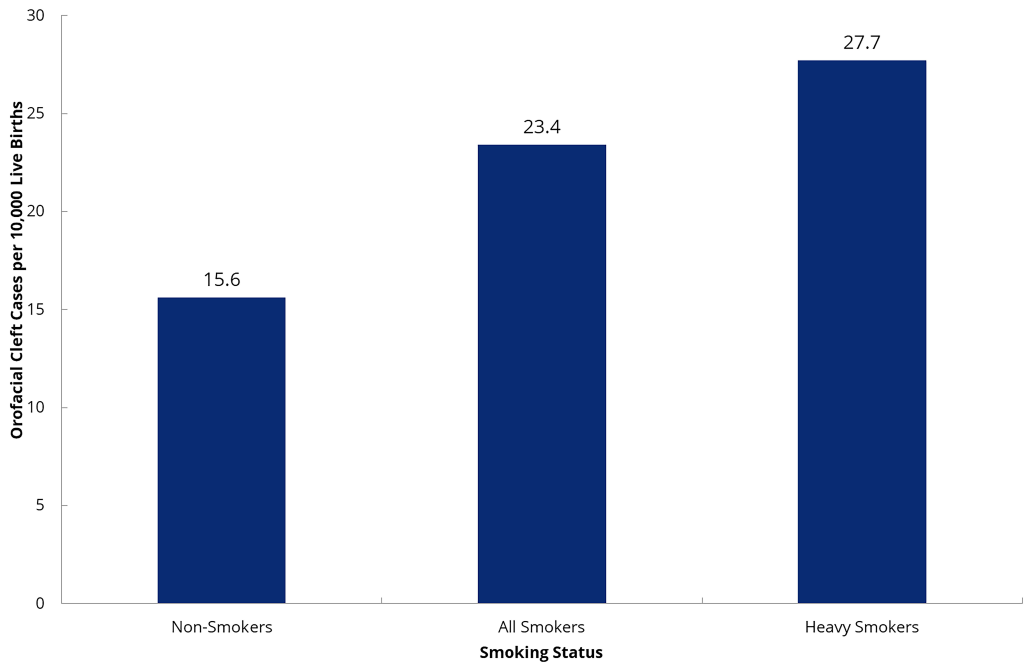
Causes and Risk Factors

Similar to other birth defects, orofacial clefts are thought to be caused by a combination of genetic changes and other factors, such as environmental exposures or certain medications taken during pregnancy. Recently, research studies have found that pregnant women who smoke are more likely to have a baby born with a cleft lip or cleft palate. Figure 3 shows the rate of orofacial clefts by maternal smoking status and intensity. There is a statistically significant difference in the rate of orofacial clefts between women who did not smoke during pregnancy and smokers.

Diabetes diagnosed before pregnancy also increases the risk of having an infant born with cleft lip with or without cleft palate. In addition, women who use certain medications used to treat seizure disorder (such as topiramate and valproic acid) during the first trimester of pregnancy have an increased risk of having a baby with cleft lip with or without cleft palate, compared to women who do not use these medications.

Birth Defect Highlight: Orofacial Clefts

Figure 3. Prevalence of Orofacial Clefts by Maternal Smoking Status, Tennessee, 2010-2015



Note: All Smokers includes women who smoked during any trimester of pregnancy. Heavy smokers includes women who smoked more than 10 cigarettes per day on average per trimester of pregnancy.
Data Source: Tennessee Department of Health, Tennessee Birth Defects Registry.

Tennessee Tobacco Cessation Resources

Baby & Me-Tobacco Free™ is an evidence based smoking cessation program that helps pregnant women quit smoking and remain tobacco-free throughout postpartum period and beyond. Pregnant women and smokers who live with the pregnant woman can enroll through contacting their local health department. Eligible women and their partners can receive counseling and diaper incentives for every month they are free of tobacco.

Tennessee Tobacco Quitline is a toll-free telephone hotline service that provides personalized support for Tennesseans who want to quit smoking or chewing tobacco.

Tennessee Tobacco QUITLINE
1-800-QUIT-NOW
1-800-784-8669
www.tnquitline.org

Birth Defects by Socio-Demographic Factors

When examining the prevalence of birth defects, it is important to consider maternal socio-demographic and health factors. Advanced maternal age is a risk factor for certain birth defects. In addition, there are racial and ethnic differences in the occurrence of certain birth defects[10]. Some health behaviors (such as smoking, alcohol use, and drug use) and health conditions (such as diabetes and hypertension) are also associated with an increased risk of specific birth defects[11].

In Tennessee, babies born to women younger than 25, women 35 years and older, women with ≤ 12th grade education, and women on Medicaid have a higher rate of birth defects (Figures 4 and 8). There are important racial/ethnic and geographical differences as well. Birth defect prevalence rates are highest for Non-Hispanic Blacks and for those living in Northeast Tennessee (Figures 7 and 9). Identifying these at-risk groups in Tennessee allows for the development of targeted prevention efforts, with the goal of reducing birth defects.

Maternal Age

Maternal age is a significant risk factor for certain types of birth defects, with advanced maternal age (defined as women who are 35 years old or older at the time of delivery) posing a higher risk for birth defects such as such as Trisomy 21 (Down Syndrome)[12]. In contrast, women younger than 20 years old are more likely to have babies born with gastroschisis[13], a birth defect of the abdominal wall, than older women.

10 Canfield MA, Mai CT, Wang Y, O'Halloran M, Marengo LK, Olney RS, Borger CL, Rutkowski R, Fornoff J, Irwin N, Copeland G, Flood TJ, Meyer RE, Rickard R, Alverson CJ, Sweatlock J, Kirby RS. The Association Between Race/Ethnicity and Major Birth Defects in the United States, 1999-2007. American Journal of Public Health. 2014.

11 https://www.cdc.gov/ncbddd/birthdefects/facts.html, accessed August 31, 2018

12 Allen EG, Freeman SB, Druschel C, et al. Maternal age and risk for trisomy 21 assessed by the origin of chromosome nondisjunction: a report from the Atlanta and National Down Syndrome Projects. Hum Genet. 2009 Feb;125(1):41-52. https://genetics.emory.edu/documents/down- syndrome/Allen%202008.pdf.

13 Jones AM, Isenburg J, Salemi JL, et al.; for the National Birth Defects Prevention Network. Increasing prevalence of gastroschisis—14 States,1995-2012. MMWR morb Mortal Wkly Rep. 2016 Jan 22;65(2):23 https: // www .cdc.gov/ mmwr /volumes/65/wr/mm6502a2.htm.

Figure 4 shows the overall prevalence of birth defects by maternal age group in Tennessee. During 2010-2015, the birth defects prevalence rates were highest among women 40 and older (480.0 per 10,000 live births), women aged 35-39 (363.6 per 10,000 live births) and women less than 20 years old (331.2 per 10,000 live births).

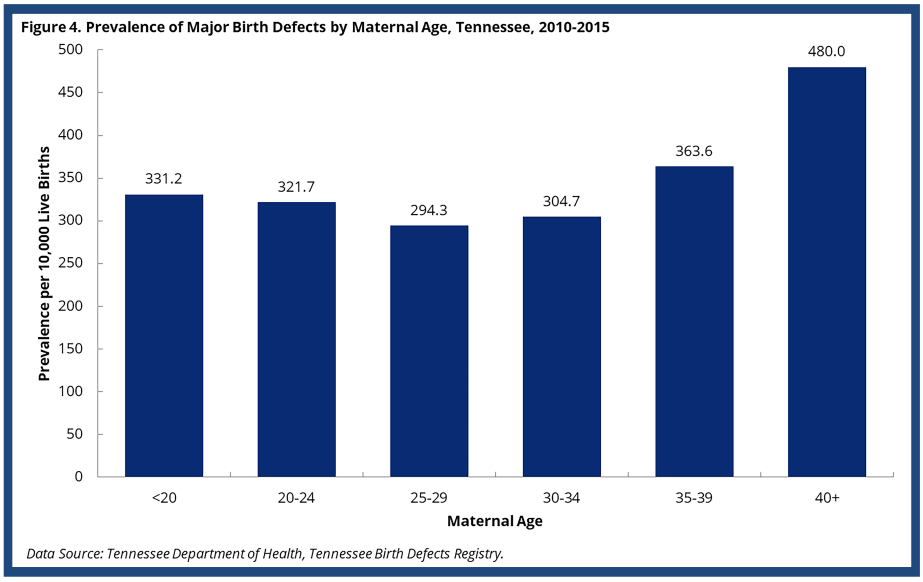


Figure 5 and Figure 6 further illustrate the significant role that maternal age plays in birth defect occurrence. Figure 5 demonstrates that children born to mothers aged 40 years and above are more likely to have a chromosomal birth defect than those born to mothers in the other age groups. In this figure, chromosomal birth defects include Trisomy 21 and Trisomy 13, which are known to be associated with advanced maternal age, as well as Deletion 22q11.2 and Turner Syndrome, which are not traditionally associated with advanced maternal age.

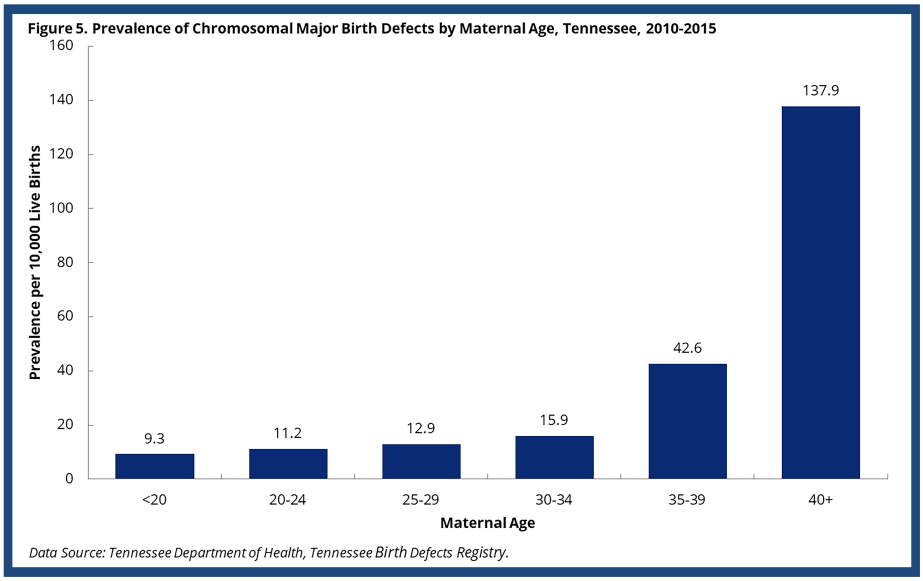
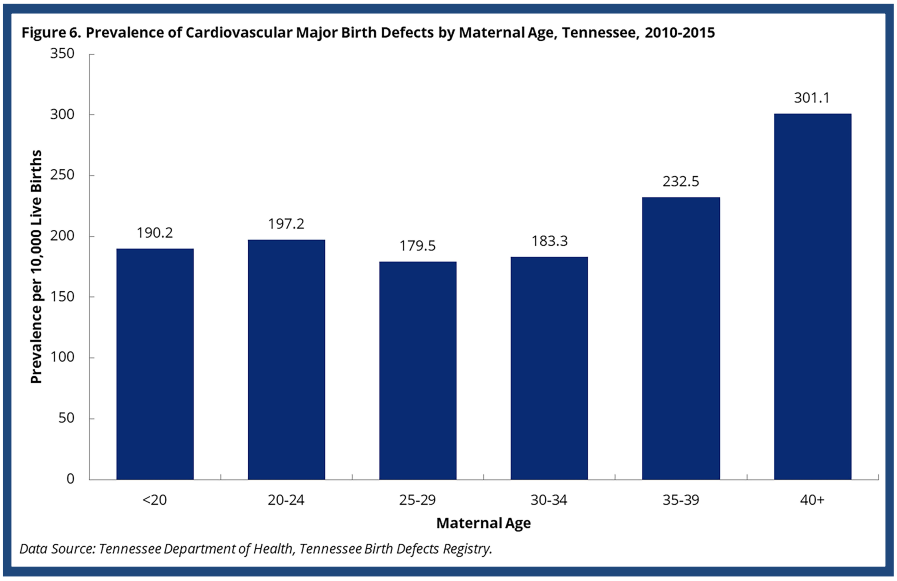
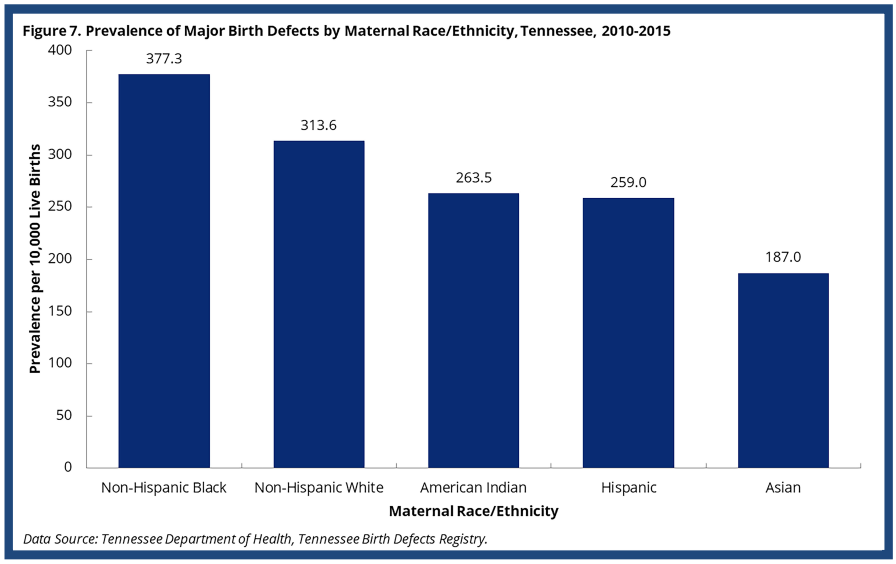


Figure 6 illustrates that the rate of cardiovascular birth defects is higher for women 35 years old or greater, as compared to younger mothers. The rate of cardiovascular birth defects among women aged 40 years old or greater (301.1 per 10,000 live births) is about 1.7 times the average rate for babies born to mothers in the age group 25-29 (179.5 per 10,000 live births).



Maternal Race and Ethnicity

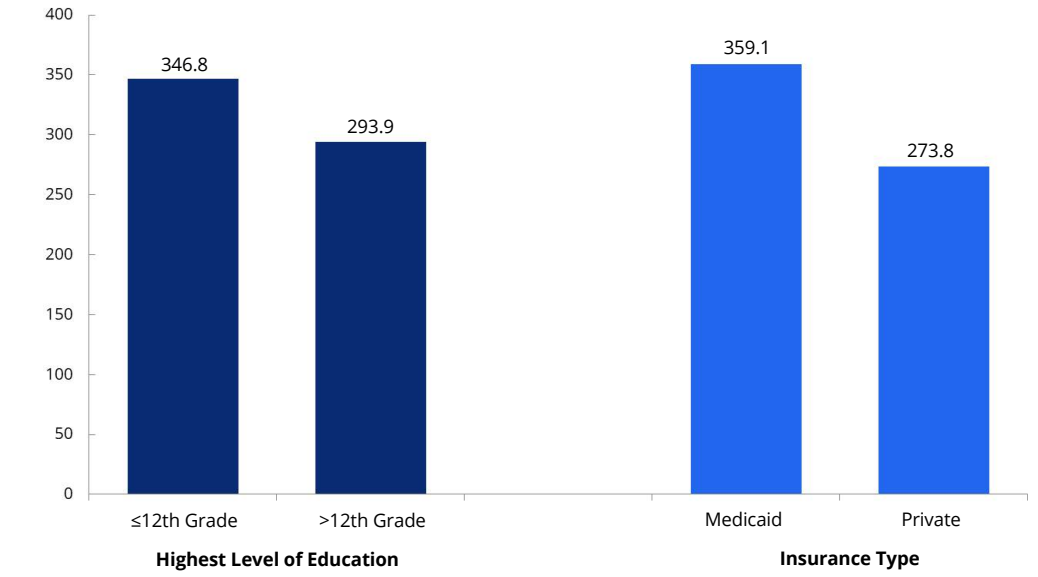
Birth defects prevalence rates are highest for Non-Hispanic Blacks (377.3 per 10,000 live births), followed by Non-Hispanic Whites and American Indians (Figure 7). The lowest prevalence rate for birth defects (187.0 per 10,000 live births) was seen among babies born to Asian women.



Other Maternal Characteristics

Birth defect prevalence rates also differed by education and insurance type. Figure 8 shows that babies born to women with ≤ 12th grade education and women on Medicaid are at a higher risk for birth defects.

Figure 8. Prevalence of Major Birth Defects by Selected Maternal Characteristics, Tennessee, 2010-2015

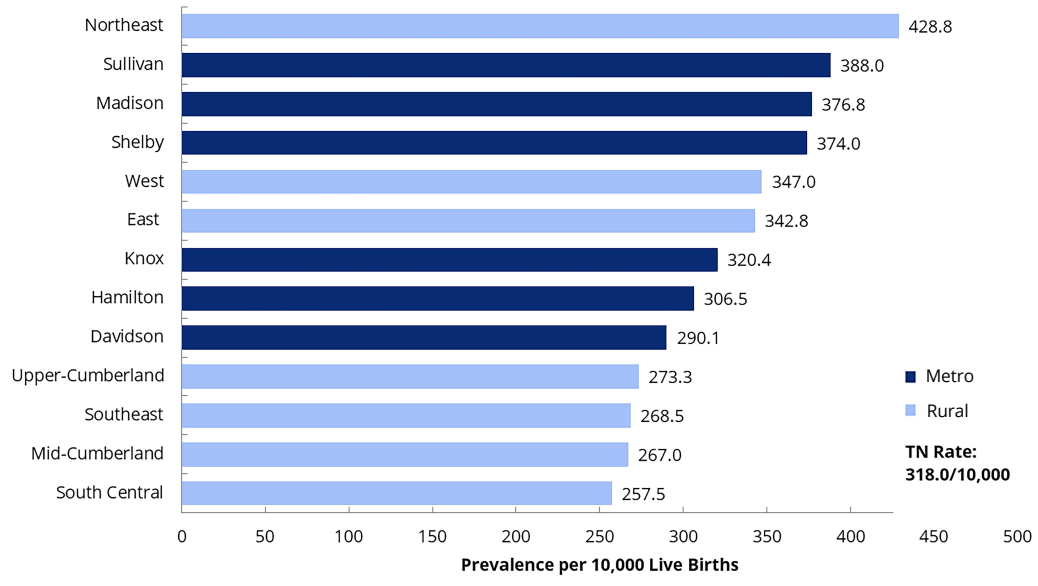


Data Source: Tennessee Department of Health, Tennessee Birth Defects Registry.

Birth Defects by Mother's Residence

Birth defects prevalence rates also differ by mother's residence. Figure 9 shows that birth defects prevalence rates are highest for those living in the Northeast region, followed by Sullivan, Madison and Shelby counties.

Figure 9. Prevalence of Major Birth Defects by Maternal Region of Residence, Tennessee, 2010-2015

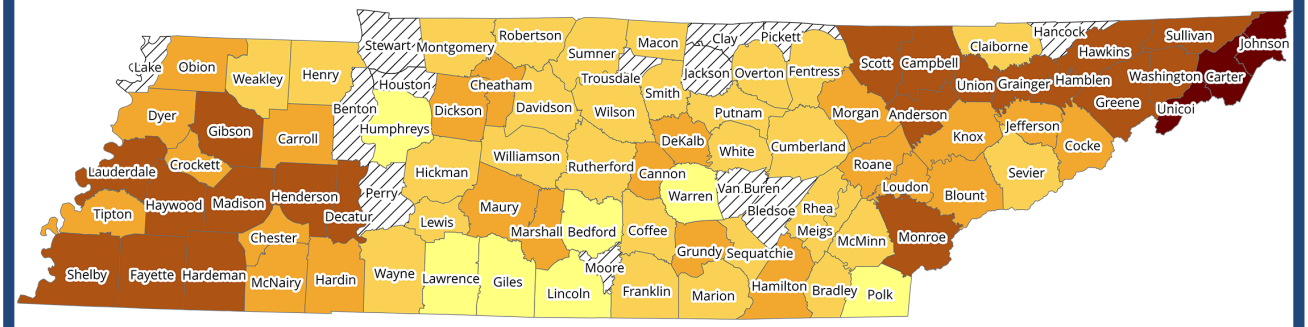


Data Source: Tennessee Department of Health, Tennessee Birth Defects Registry.

Birth Defect Rates by County

Figure 10 depicts the birth defects rate by maternal county of residence. There are variations from one county to another in terms of specific defect rates. For instance, the overall defect rate is 395.2 per 10,000 live births in Anderson County compared to 233.2 in Bedford County. The differences may reflect underlying differences in the population and variations in risk factors. Given that the numbers are generally small the differences in rates should be interpreted with caution.

Figure 10. Prevalence of Major Birth Defects by County, Tennessee, 2010-2015



Data Source: Tennessee Department of Health, Tennessee Birth Defects Registry.

Maternal Health Factors

Some health behaviors (such as smoking and drinking alcohol) and chronic health conditions (such as diabetes and hypertension) are associated with an increased risk of specific birth defects. By identifying and analyzing these risk factors, targeted prevention efforts can be developed. Table 8 illustrates the relative risk of birth defects by maternal health factors: diabetes, hypertension (high blood pressure), smoking during pregnancy, body mass index (BMI), and level of prenatal care (based on the Kotelchuck index, which combines the timing of initiation of prenatal care with the number of prenatal visits). Infants born to mothers with pre-pregnancy diabetes had almost 2.8 times the risk of birth defects compared to infants born to mothers without diabetes. In addition, infants born to mothers with gestational diabetes had about 1.4 times the risk of birth defects compared to infants born to mothers without diabetes. Maternal hypertension (pre-existing and gestational) was also associated with increased risk of birth defects. Infants born to mothers with pre-pregnancy hypertension had almost 1.9 times the risk of birth defects, while infants born to mothers with gestational hypertension had almost 1.5 times the risk of birth defects compared to infants born to mothers without hypertension. Smoking during pregnancy was also associated with increased risk of birth defects; infants born to mothers who smoked during pregnancy had 1.25 times the risk of birth defects compared to infants born to mothers who did not smoke during pregnancy. An abnormal BMI (underweight, overweight or obese) was also associated with increased risk of birth defects; the highest relative risk of birth defects in this category was for infants born to mothers who were obese, compared to mothers with normal BMI. Finally, inadequate prenatal care was associated with increased risk of birth defects. Infants born to mothers with inadequate or intermediate prenatal care had about 1.4 and 1.1 times the risk of birth defects, respectively, compared to mothers who received adequate prenatal care. All of these reported associations between maternal health factors and birth defects were found to be statistically significant, meaning that the differences are larger than would be expected by chance alone. It is important to note, however, that Table 8 presents crude relative risks not adjusted for other factors such as maternal age and race that could contribute to the observed associations.

Table 8. Relative Risk of Birth Defects by Maternal Health Characteristics, Tennessee, 2010-2015[1]			
Maternal Health Characteristic		**Relative Risk of Birth Defects[2]**	**95% CI[3]**
Diabetes	Pre-existing	2.79	2.55-3.06
	Gestational	1.41	1.33-1.49
	None	Reference	–
Hypertension	Pre-existing	1.85	1.72-2.00
	Gestational	1.48	1.40-1.57
	None	Reference	–
Pregnancy Smoking Status[4]	Smoker	1.25	1.20-1.30
	Non-Smoker	Reference	–
BMI	Underweight	1.12	1.04-1.21
	Normal	Reference	–
	Overweight	1.09	1.05-1.14
	Obese	1.28	1.23-1.33
Prenatal Care[5]	Inadequate	1.43	1.36-1.50
	Intermediate	1.13	1.05-1.21
	Adequate	Reference	–

1. Relative risks shown are crude, meaning they are not adjusted for any other factors that could contribute to the observed associations.
2. Compares the risk of birth defects in group exposed to a given maternal characteristic with the risk in the reference group for that category. Data interpretation example: infants born to mothers with pre-pregnancy diabetes had 2.79 times the risk of birth defects compared to infants born to mothers with no diabetes.
3. Can be interpreted as range that we are 95% confident contains the true relative risk for the population. Data interpretation example: we are 95% confident that the relative risk of birth defects in infants born to mothers with pre-pregnancy diabetes compared to infants born to mothers with no diabetes is between 2.55 and 3.06. Note that where 95% confidence interval does not include 1 (every instance shown), the difference is statistically significant.
4. Smokers defined as women who smoked during any trimester of pregnancy.
5. Prenatal care categories based on the Kotelchuck index, which combines the timing of initiation of prenatal care with the number of prenatal visits (adjusted for gestational age).

Maternal Diabetes

Diabetes is a chronic disease affecting an increasing number of mothers. Babies born to mothers with type I and type II diabetes are more likely to be born with central nervous system, ear/ eye, gastrointestinal, genitourinary, musculoskeletal, orofacial and cardiovascular birth defects. The 2010-2015 birth defects counts, rates, and confidence intervals for 5 organ systems by maternal pre-pregnancy diabetes are presented in Table 9. The cardiovascular system birth defect rate for mothers with pre-pregnancy diabetes was 630.4 per 10,000 live births. The corresponding figure for mothers without pre- pregnancy diabetes was 188.9. This finding suggests that babies born to mothers with pre-pregnancy diabetes are at increased risk for cardiovascular system birth defects. The defect rates for four other organ systems (central nervous, genitourinary, musculoskeletal and orofacial) are also significantly different between babies born to mother with pre-pregnancy diabetes and those whose mother did not have diabetes prior to their pregnancy. However, given that the counts in the pre-pregnancy diabetes groups are very small for these other organ systems, the findings should be interpreted with caution.

Table 9. Prevalence of Major Birth Defects by Organ System for Infants Born to Mothers with Pre-Pregnancy Diabetes Compared to Infants Born to Mothers without Pre-Pregnancy Diabetes, Tennessee, 2010-2015

Organ System	Pre-Pregnancy Diabetes		No Pre-Pregnancy Diabetes[1]		Relative Risk[3] (95% CI)[4]
	Cases	Rate[2]	Cases	Rate[2]	
Cardiovascular	316	630.4	9008	188.9	3.3 (3.0, 3.7)
Central Nervous System	20	39.9	603	12.6	3.2 (2.0, 4.9)
Genitourinary	72	143.6	3283	68.8	2.1 (1.7, 2.6)
Musculoskeletal	26	51.9	1548	32.5	1.6 (1.1, 2.4)
Orofacial	22	43.9	886	18.6	2.4 (1.5, 3.6)

1. Includes infants born to mothers with gestational (but not pre-pregnancy) diabetes.
2. Rate per 10,000 live births.
3. Compares the rate of birth defects for a given organ system amongst infants born to mothers with pre-pregnancy diabetes to the rate amongst infants born to mothers with no pre-pregnancy diabetes. Relative risks shown are crude, meaning they are not adjusted for any other factors that could contribute to the observed associations.
4. Can be interpreted as range that we are 95% confident contains the true relative rate for the population.

Data Source: Tennessee Department of Health, Tennessee Birth Defects Registry.

Other Risk Factors

Although the causal mechanisms of most birth defects are not fully understood, there are known risk factors that increase the likelihood of giving birth to a baby with a birth defect. Drinking alcohol and smoking cigarettes during pregnancy are associated with increased risk of having a baby born with a birth defect. Babies born to mothers who smoke cigarettes are more likely to be born premature and low birth weight. They are also more likely to be born with cardiovascular, orofacial, gastrointestinal, and musculoskeletal birth defects. There is no amount of alcohol that is safe to drink during pregnancy. When a pregnant woman drinks, the alcohol in her system passes from mother to baby. Drinking can also cause fetal alcohol syndrome, which is a serious condition involving growth deficiencies, facial abnormalities, central nervous system impairment, and intellectual disabilities.

Some infections that a woman can get during pregnancy can be harmful to the developing baby and can even cause birth defects. For example, Zika virus infection during pregnancy can cause microcephaly and other neurologic birth defects. Fetal exposure to rubella, a vaccine preventable illness, increases the risk of a baby being born with congenital rubella syndrome, which affects the ear/eye and cardiovascular systems. Toxoplasmosis is caused by the parasite, Toxoplasma gondii. Babies born to women with a toxoplasmosis infection are at risk for hydrocephalus which affects the central nervous system. Likewise, babies born following in utero exposure to cytomegalovirus (CMV) may have long-term health problems, such as hearing loss, vision loss, microcephaly, seizures and developmental delay.

Although not all birth defects can be prevented, avoiding the known risk factors could help to reduce one's risk of having a baby with a birth defect. A woman can reduce her risk of delivering a baby born with a birth defect or other adverse outcomes by taking precautions before and during pregnancy. The best time to start preventing birth defects is before a woman becomes pregnant. Most of the baby's vital organs and systems are formed in the first four to eight weeks of gestation, often before a woman knows she is pregnant.

Folic Acid is a B-complex vitamin that is proven to be protective against neural tube defects such as anencephalus and spina bifida, which are defects of the central nervous system. It may also provide protection against other birth defects.

To be fully effective, a woman needs to begin taking the recommended daily dose of 400 micrograms at least a full month before becoming pregnant and continue to take folic acid daily during pregnancy. For women who have had a baby with a neural tube defect in the past, the recommended daily dose of folic acid is higher. If a woman finds she is pregnant and has not been taking folic acid, it is best to start taking folic acid immediately and continue to do so thereafter.

A woman should see her medical provider when planning a pregnancy and start prenatal care as soon as she thinks that she is pregnant. A pregnant woman should work with her healthcare provider to keep chronic diseases (like diabetes) under control, avoid drinking alcohol, avoid smoking cigarettes and prevent infections as much as possible. Some easy steps to prevent infections include frequent hand-washing, cooking meat until it is well done, and staying away from people who have an infection. Another way to prevent infections is to be up-to-date with recommended vaccines before, during, and after pregnancy. Vaccines such as the measles, mumps, rubella (MMR) vaccine, which are recommended in childhood, are critical to prevent congenital infections that can cause birth defects. It's also important for people who pregnant woman may come in contact with to be vaccinated so they don't expose the pregnant woman and her baby to vaccine-preventable diseases. Likewise, vaccines such as the influenza vaccine are critical during pregnancy for the health of both mother and baby.

Occupational and environmental exposures such as radiation, certain chemicals (such as lead), and strenuous physical labor may harm the health of mother and baby. Hazardous work environments should be avoided during pregnancy. Pregnant workers, and those planning to become pregnant, should learn the safety and proper work practices of their employer. The National Institute for Occupational Safety and Health recommends using personal protective equipment, avoiding skin contact with chemicals, washing hands before eating or drinking, reviewing all workplace material safety data sheets to learn about potential hazards, leaving contaminated clothing at work, showering with soap and water before leaving, and keeping street clothes separate from work clothes to prevent contamination[14]. These practices help prevent exposure of individuals and their familial contacts to hazardous chemicals.

While there are still certain hereditary and genetic factors that cannot be avoided, there are many factors that public health staff, new mothers-to-be and health care providers can address together to reduce birth defect occurrences in infants born in Tennessee.

14. National Institute for Occupational Safety and Health (US). The Effects of Workplace Hazards on Female Reproductive Health. Department of Health and Human Services (US) 1999. 20 p. (DHHS (NIOSH) publication; no. 99-10). https://www.cdc.gov/niosh/docs/99-104/pdfs/99-104.pdf?id=10.26616/NIOSHPUB99104

Tips for a Healthy Pregnancy

Before and during pregnancy

- Consume at least 400 micrograms (mcg) of folic acid every day
- See a healthcare professional regularly
- Plan and space pregnancies at least 18 months apart
- Prevent and treat medical conditions like diabetes and hypertension
- Strive to reach and maintain a healthy weight
- Be physically active
- Eat a healthy diet that includes fruits, vegetables, whole grains, low fat dairy, and lean proteins

Avoid harmful substances

- Avoid smoking
- Avoid drinking alcohol
- Avoid drugs such as opioids, marijuana, cocaine, methamphetamines, and other "street" drugs
- Be aware of and avoid potentially harmful exposures at work and home

Talk to a healthcare provider about

- Getting a medical checkup
- Taking any medications, both prescription and over-the-counter
- Family history of medical conditions
- Vaccinations needed before, during (Flu vaccine and Tdap vaccine), and after pregnancy
- Any upcoming travel (either domestically or abroad) to discuss Zika and other risks, vaccination requirements, and the potential need for medical care in transit and at your destination

Appendix A: Reportable Birth Defects in Tennessee

Reportable Birth Defects

Brain abnormalities with and without microcephaly	
Confirmed or possible congenital microcephaly <3rd percentile	Q02
Intracranial calcifications	No specific code; may be included under Q04.8, Q04.9
Cerebral atrophy	No specific code; may be included under Q04.3
Abnormal cortical formation (e.g., polymicrogyria, lissencephaly, pachygyria, schizencephaly, gray matter heterotopia)	Q04.3, Q04.6, Q04.8
Corpus callosum abnormalities	Q04.0
Cerebellar abnormalities	No specific code; may be included under Q04.3
Porencephaly	Q04.6
Hydranencephaly	No specific code; should be included in Q04.3
Ventriculomegaly / hydrocephaly Mild or borderline Ventriculomegaly/enlargement of cerebral ventricles must have another qualifying defect to be reported.	Q03.0–Q03.9
Fetal brain disruption sequence (include: collapsed skull, overlapping sutures, prominent occipital bone, scalp rugae, etc.)	No specific code. This might be coded as microcephaly or another single brain malformation, or all the components that might be coded individually. Q02, Q04.8, Q04.9 Include the following abnormalities only if co-existing abnormalities of the brain have been diagnosed: Q67.4, Q75.8, Q75.9, Q82.8
Other major brain abnormalities, including intraventricular hemorrhage Include *in utero* IVH, only if an additional qualifying defect is present	Q04.0, Q04.3–Q04.9, Q07.00, Q07.02
Neural tube defects and other early brain malformations	
Anencephaly / Acrania	Q00.0–Q00.2
Encephalocele	Q01.0–Q01.9
Spina bifida	Q05.0–Q05.9, Q07.01, Q07.03
Holoprosencephaly / Arhinencephaly	Q04.1, Q04.2
Eye abnormalities	
Microphthalmia / Anophthalmia	Q11.0–Q11.2
Coloboma	Q12.2, Q13.0, Q14.1–Q14.8
Cataract	Q12.0
Intraocular calcifications	Q13.8, Q13.9, Q14.1–Q14.9
Chorioretinal anomalies involving the macula (e.g., chorioretinal atrophy and scarring, macular pallor, gross pigmentary mottling and retinal hemorrhage); excluding retinopathy of prematurity	No specific code. This might be coded under the affected part of the eye. Q14.1–Q14.9
Optic nerve atrophy, pallor, and other optic nerve abnormalities	Q14.2, H47.03
Consequences of central nervous system (CNS) dysfunction	
Congenital contractures (e.g., arthrogryposis, club foot, congenital hip dislocation/developmental dysplasia of the hip) **only** with associated brain abnormalities	Q65.0–Q65.9, Q66.0–Q66.9, Q68.8, Q74.3
Confirmed congenital deafness documented by postnatal testing	H90.0–H90.8, H90.A, H91.0-H91.9, Q16.0–Q16.9

The TN Birth Defects Surveillance System Reporting Portal can be found here:
https://tdhrc.health.tn.gov/redcap/surveys/?s=TDEYPYCHET

Appendix B: Birth Defects by Organ System and Fetal Alcohol Syndrome

Table 10. Cases and Prevalence of Major Birth Defects and Fetal Alcohol Syndrome, Tennessee, 2010-2015			
Birth Defect	**Number[1]**	**Rate[2]**	**95% CI[3]**
Central Nervous System			
Anencephaly	74	1.5	1.2-1.9
Encephalocele	58	1.2	0.9-1.6
Holoprosencephaly	336	7.0	6.2-7.7
Spina bifida without anencephaly	206	4.3	3.7-4.9
Total Central Nervous System Cases	642	13.3	12.3-14.4
Total Central Nervous System Defects	674	14.0	12.9-15.0
Eye/Ear			
Anophthalmia/microthalmia	64	1.3	1.0-1.7
Anotia/microtia	48	1.0	0.7-1.3
Congenital cataract	103	2.1	1.7-2.5
Total Eye and Ear Cases	208	4.3	3.7-4.9
Total Eye and Ear Defects	215	4.5	3.9-5.1
Cardiovascular			
Aortic valve stenosis	81	1.7	1.3-2.1
Atrial septal defect	7,435	154.3	150.7-157.8
Atrioventricular septal defect (Endocardial cushion defect)	254	5.3	4.6-5.9
Coarctation of the aorta	397	8.2	7.4-9.0
Common truncus (truncus arteriosus or TA)	45	0.9	0.7-1.2
Double outlet right ventricle (DORV)	149	3.1	2.6-3.6
Ebstein anomaly	80	1.7	1.3-2.1
Hypoplastic left heart syndrome	188	3.9	3.3-4.5
Interrupted aortic arch (IAA)	56	1.2	0.9-1.5
Pulmonary valve atresia and stenosis	453	9.4	8.5-10.3
Single Ventricle	84	1.7	1.4-2.2
Tetralogy of Fallot (TOF)	290	6.0	5.3-6.7
Total anomalous pulmonary venous connection (TAPVC)	67	1.4	1.1-1.8
Transposition of the great arteries (TGA)	249	5.2	4.5-5.8
Tricuspid valve atresia and stenosis	70	1.5	1.1-1.8
Ventricular septal defect	2,415	50.1	48.1-52.1
Total Cardiovascular Cases	9,324	193.5	189.5-197.4
Total Cardiovascular Defects	12,313	255.5	250.9-260.1
Orofacial			
Choanal atresia	102	2.1	1.7-2.5
Cleft lip with cleft palate	340	7.1	6.3-7.8
Cleft lip alone (without cleft palate)	139	2.9	2.4-3.4
Cleft palate alone (without cleft lip)	334	6.9	6.2-7.7
Total Orofacial Cases	908	18.8	17.6-20.1
Total Orofacial Defects	915	19.0	17.8-20.2

Continued on next page

Appendix B: Birth Defects by Organ System and Fetal Alcohol Syndrome

Birth Defect	Number	Rate	95% CI
Gastrointestinal			
Biliary atresia	62	1.3	1.0-1.6
Esophageal atresia/tracheoesophageal fistula	145	3.0	2.5-3.5
Rectal and large intestinal atresia/stenosis	268	5.6	4.9-6.2
Small intestinal atresia/stenosis	257	5.3	4.7-6.0
Total Gastrointestinal Cases	680	14.1	13.0-15.2
Total Gastrointestinal Defects	732	15.2	14.1-16.3
Genitourinary			
Bladder exstrophy	14	0.3	0.2-0.5
Cloacal exstrophy	432	9.0	8.1-9.8
Congenital Posterior Urethral Valves	69	2.8	2.2-3.5
Hypospadias	2,580	104.6	100.5-108.6
Renal agenesis/hypoplasia	296	6.1	5.4-6.8
Total Genitourinary Cases	3,359	69.7	67.3-72.1
Total Genitourinary Defects	3,391	70.4	68.0-72.7
Musculoskeletal			
Clubfoot	864	17.9	16.7-19.1
Diaphragmatic hernia	200	4.1	3.6-4.7
Gastroschisis	270	5.6	4.9-6.3
Limb deficiencies (reduction defects)	194	4.0	3.5-4.6
Omphalocele	119	2.5	2.0-2.9
Total Musculoskeletal Cases	1,592	33.0	31.4-34.7
Total Musculoskeletal Defects	1,647	34.2	32.5-35.8
Chromosomal			
Deletion 22q11.2	10	0.2	0.1-0.4
Trisomy 13	47	1.0	0.7-1.3
Trisomy 18	83	1.7	1.4-2.1
Trisomy 21 (Down syndrome)	697	14.5	13.4-15.5
Turner syndrome	34	1.4	1.0-2.0
Total Chromosomal Cases	866	18.0	16.8-19.2
Total Chromosomal Defects	871	18.1	16.9-19.3
Total Birth Defects Cases	15,325	318.0	312.9-323.1
Total Birth Defects	20,758	430.7	424.7-436.7
Fetal Alcohol Syndrome[4]	77	1.6	1.3-2.0

Note: For each organ system, the total *cases* number represents the number of infants. The total *defects* number represents the full count of diagnosed birth defects. These numbers are not equivalent because one infant can potentially be diagnosed with more than one birth defect. For example, a total of 9,324 infants were diagnosed with cardiovascular birth defects, but amongst these 9,324 infants, there were 12,313 total cardiovascular defects.

1. Number includes cases born alive and fetal deaths.
2. Rate per 10,000 live births.
3. Can be interpreted as range that we are 95% confident contains the true incidence in the population. Confidence intervals for conditions with less than 100 cases are exact Poisson; otherwise confidence intervals are based on the normal approximation.
4. Fetal alcohol syndrome cases are not included in the count for total birth defect cases/total birth defects.

Data Source: Tennessee Department of Health, Tennessee Birth Defects Registry.

Appendix C: Resources

Tennessee Resources

Tennessee Department of Health:
https://www.tn.gov/health/health-program-areas/mch-cyshcn.html

Tennessee Medical Home:
http://www.tennesseemedicalhome.com/tnaap/

Tennessee Parent to Parent Program:
http://www.tndisability.org/tennessee-parent-parent

Family Voices of Tennessee:
http://www.tndisability.org/familyvoices

Support and Training for Exceptional Parents (STEP):
https://www.tnstep.org/

Kidcentral tn:
http://www.kidcentraltn.com

Disability Pathfinder:
http://vkc.mc.vanderbilt.edu/vkc/pathfinder/

Disability Rights Tennessee:
http://www.disabilityrightstn.org/

University of Tennessee Boling Center for Developmental Disabilities:
https://www.uthsc.edu/bcdd/

Vanderbilt Kennedy Center:
https://vkc.mc.vanderbilt.edu/vkc/

Vanderbilt Consortium Leadership Education in Neurodevelopmental Disabilities (LEND)
https://www.etsu.edu/coe/efse/lend.php

Tennessee Early Intervention System (TEIS):
https://www.tn.gov/education/early-learning/tennessee-early-intervention-system-teis.html

Chattanooga Down Syndrome Society:
http://www.chattanoogadownsyndrome.org/

Clarksville Association Down Syndrome:
https://www.cadstn.org/

Down Syndrome Association of Middle Tennessee:
https://www.somethingextra.org/

Down Syndrome Association of West Tennessee:
https://dsawt.com/

Down Syndrome Awareness Group of East Tennessee:
http://dsagtn.org/

FRIENDS (Friends Reaching Inspiring Educating Neighbors about Down Syndrome):
http://dsfriends.net/

Baby & Me - Tobacco Free™:
http://www.babyandmetobaccofree.org

Tennessee Tobacco Quitline:
http://www.tnquitline.org

National Resources

CDC National Center on Birth Defects and Developmental Disabilities:
https://www.cdc.gov/ncbddd/birthdefects/index.html

National Birth Defects Prevention Network:
https://www.nbdpn.org/

March of Dimes:
https://www.marchofdimes.org/

CDC National Institute for Occupational Safety and Health:
https://www.cdc.gov/niosh/topics/repro/pregnancyjob.html/

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