Tennessee Birth Defects Data Report 2011-2016

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Executive Summary

Birth defects are common, costly, and critical. 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. Between years 2011 and 2016, there were 16,138 babies (approximately 2,690 babies per year) diagnosed with birth defects in Tennessee. The most commonly reported birth defects during this period were atrial septal defect (ASD), a hole or opening in the heart, and hypospadias, a genitourinary defect that affects males. Birth defects account for 22% of the infant deaths in Tennessee, making it the leading cause of infant mortality.

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 annual population-based surveillance report provides details on the prevalence of 47 major birth defects and fetal alcohol syndrome for Tennessee infants born in the years 2011 through 2016. This report also includes specific information about birth defect rates by socio-demographics characteristics, known risk factors, region and county of residence.

Major findings from this report include:

- 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.
- The highest prevalence of birth defects was found in the Northeast region.
- Non-Hispanic Black babies had the highest prevalence of birth defects among maternal racial/ethnic groups.

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. Folic acid supplementation should begin months before becoming pregnant.
- 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, and 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?

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, about 120,000 babies (nearly one in 33 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. 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

The Tennessee Birth Defects Data Report is a statewide population-based birth defects report prepared by the Tennessee Birth Defects Surveillance System (TNBDSS). This report provides details about the prevalence of 47 major birth defects[2] and fetal alcohol syndrome for Tennessee infants born in the years 2011 through 2016. 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 and known risk factors. Individual birth defect counts and rates are presented in tabular form for the state overall. Data are also broken down by maternal education, race/ethnicity, age, and maternal health characteristics, such as pre- pregnancy diabetes and smoking during pregnancy. Special attention is given to selected birth defects of public health significance. This report provides education on birth defects prevention, highlights current prevention efforts, 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. 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. Additionally, babies born to mothers with pre-pregnancy diabetes are at increased risk for cardiovascular, central nervous system, genitourinary, musculoskeletal, and orofacial birth defects compared to babies born to mothers who did not have diabetes. These findings also underscore the impact of social and maternal health factors such as education, income, environment, and prevalence of chronic disease on health outcomes.

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 after birth.

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. TNBDSS uses these records to track the 47 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 fetal death cases, demographic, geographic, and risk factor information are obtained from the fetal death certificate system. For infant death 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 prior to October 2015, 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 defect, which is a congenital heart defect, 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[3] surfaced as a public health threat in the United States. In 2016, the Tennessee Department of Health (TDH) was awarded an Epidemiology and Laboratory Capacity grant from Centers for Disease Control and Prevention (CDC), 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, healthcare 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,

3. Zika virus infection during pregnancy can cause microcephaly and other neurologic birth defects. To understand more about Zika virusinfection, 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, 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 80,559 live births occurred to resident mothers annually during the years 2011 through 2016. 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, 2011-2016

Year	Total	Non-Hispanic White	Non-Hispanic Black	Hispanic
2011	79,462	53,454	16,370	7,017
2012	80,202	54,018	16,462	6,977
2013	79,954	54,251	16,764	6,850
2014	81,609	55,345	16,927	6,982
2015	81,374	54,621	16,571	7,260
2016	80,755	53,725	16,212	7,628

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, 2011-2016

Table 9 in Appendix B shows the case numbers and rates for the 47 major birth defects[4] by organ system and fetal alcohol syndrome. Between January 2011 and December 2016, there were 16,138 babies diagnosed with birth defects. In addition, there were 69 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 birth defects (21,807) over this time period is higher. Out of the 21,807 defects, 13,088 were cardiovascular defects which represent 60% of the total. The genitourinary system, with 3,403 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 8,057 or 166.7 per 10,000 live births, followed by hypospadias (n=2,648) and ventricular septal defect (n=2,444). 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.

Birth Defects in Tennessee versus the United States

Table 2 shows the rate and frequency for 30 birth defects for which there is updated national prevalence data available. This table features Tennessee data from 2011-2016 for the selected birth defects, while national rates are listed for these same birth defects from the years 2010-2014. National estimates are based on pooled data from 39 state birth defects surveillance programs. Of the 30 birth defects listed, rates in Tennessee are higher for 22 of the birth defects. When examining the cardiovascular birth defects, in particular, there are notable differences between rates in Tennessee and the United States. Rates for Single Ventricle and Double Outlet Right Ventricle are approximately 2 times higher in Tennessee than the national average. In addition, rates for Coarctation of the Aorta and Hypoplastic Left Ventricle are 1.5 times and 1.4 times, respectively, higher than the national rates. It is important to identify birth defects with rates that are higher than the national average as these warrant particular attention.

4. 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 for the U.S. and Tennessee.				
Birth Defect	**Tennessee 2011-2016**		**United States[1] 2010-2014**	
	Rate[2]	**Frequency[3]**	**Rate**	**Frequency**
Central Nervous System				
Anencephaly	1.82	1 in 5,493	2.15	1 in 4,647
Encephalocele	1.26	1 in 7,924	0.95	1 in 10,502
Spina bifida without anencephaly	4.59	1 in 2,177	3.63	1 in 2,758
Eye				
Anophthalmia/microphthalmia	1.41	1 in 7,108	1.91	1 in 5,243
Cardiovascular				
Atrioventricular septal defect	5.85	1 in 1,708	5.38	1 in 1,859
Coarctation of the aorta	8.59	1 in 1,165	5.57	1 in 1,795
Common truncus	0.95	1 in 10,508	0.64	1 in 15,696
Double outlet right ventricle	3.21	1 in 3,118	1.67	1 in 5,997
Ebstein anomaly	1.72	1 in 5,824	0.77	1 in 13,047
Hypoplastic left heart syndrome	3.64	1 in 2,746	2.60	1 in 3,841
Interrupted aortic arch	1.94	1 in 5,142	0.62	1 in 16,066
Pulmonary valve atresia and stenosis	10.26	1 in 975	9.51	1 in 1,052
Single ventricle	1.57	1 in 6,360	0.75	1 in 13,351
Tetralogy of Fallot	6.21	1 in 1,611	4.61	1 in 2,171
Total anomalous pulmonary venous connection	1.30	1 in 7,672	1.28	1 in 7,809
Transposition of great arteries	5.15	1 in 1,941	3.71	1 in 2,695
Tricuspid valve atresia and stenosis	1.43	1 in 7,005	1.68	1 in 5,938
Orofacial				
Cleft lip with cleft palate	6.58	1 in 1,520	6.40	1 in 1,563
Cleft lip alone (without cleft palate)	2.79	1 in 3,580	3.56	1 in 2,807
Cleft palate alone (without cleft lip)	6.74	1 in 1,483	5.93	1 in 1,687
Gastrointestinal				
Esophageal atresia/tracheoesophageal fistula	3.14	1 in 3,180	2.41	1 in 4,144
Rectal and large intestinal atresia/stenosis	5.17	1 in 1,933	4.46	1 in 2,242
Musculoskeletal				
Clubfoot	18.89	1 in 529	16.87	1 in 593
Diaphragmatic hernia	4.01	1 in 2,492	2.79	1 in 3,591
Gastroschisis[4]	5.42	1 in 1,845	4.94	1 in 2,025
Limb deficiencies (reduction defects)	4.14	1 in 2,417	5.15	1 in 1,943
Omphalocele	2.63	1 in 3,806	2.40	1 in 4,175

Continued on next page

Chromosomal[4]				
Trisomy 13	0.99	1 in 10,070	1.49	1 in 6,717
Trisomy 18	1.63	1 in 6,118	3.43	1 in 2,918
Trisomy 21 (Down syndrome)	14.85	1 in 673	15.74	1 in 635

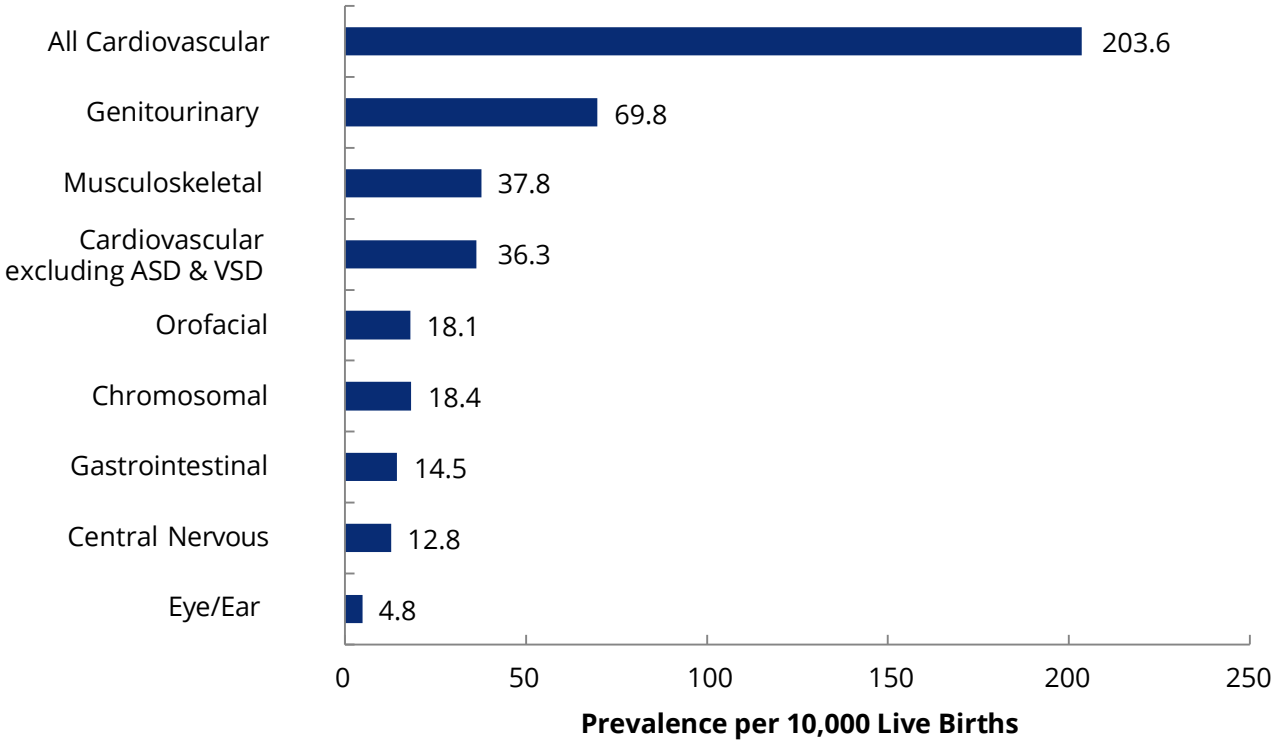
1. National estimates based on pooled data from 39 state birth defects surveillance programs. Estimates were standardized to the racial and ethnic distribution of the United States live birth population from 2010 through 2014. See full paper: Mai CT, Isenburg JL, Canfield MA et al: National population-based estimates for major birth defects, 2010–2014. *Birth Defects Research* 2019: 1–16.
2. Rate per 10,000 live births.
3. Estimated frequency of occurrence in a given number of live births.
4. Estimates for gastroschisis and the three chromosomal birth defects were standardized to the United States maternal age distribution.

Prevalence of Major Birth Defects by Organ System

Figure 1 shows the prevalence rates of birth defects by organ system. Cardiovascular system defects are the most commonly diagnosed birth defects in Tennessee, with a rate of 203.6 per 10,000 live births, followed by genitourinary system defects with a rate of 69.8 per 10,000 live births. In this figure, cardiovascular birth defects excluding atial septal defect (ASD) and ventricular septal defect (VSD) were also analyzed. An ASD is a hole in the wall (septum) that divided the two upper chambers of the heart.[5] ASDs are common (accounting for 10-15% of congenital heart defects) and often spontaneously resolve during infancy or early childhood.[6] A VSD is a hole in the septum that seperates the two lower chambers (ventricles) of the heart.[7] VSDs can be classified by the size of the hole in the septum (small, medium, or large); the size of the defect influences which signs and symptoms, if any, are present.[8] Most small VSDs spontaneously close during the first two years of life. However, babies with large VSDs may have symptoms, such as shortness of breath, fast breathing, sweating, tiredness while feeding, or poor weight gain. Because many ASDs and VSDs spontaneously close, the rate of cardiovascular birth defects excluding ASDs and VSDs was also examined and found to be 36.3 per 10,000 live births.

5. National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention. https://www.cdc.gov/ncbddd heartdefects/atrialseptaldefect.html
6. Vick, G.W. & Bezold, L.I. Isolated atrial septal defects (ASDs) in children: Classification, clinical features, and diagnosis. In: UpToDate, Post, TW (Ed), UpToDate, Waltham, MA, 2019.
7. Fulton, D. R. & Saleeb, S. Isolated ventricular septal defects in infants and children: Anatomy, clinical features, and diagnosis. In: UpToDate, Post, TW (Ed), UpToDate, Waltham, MA, 2019.
8. National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention. https://www.cdc.gov/ncbddd/ heartdefects/ventricularseptaldefect.html

Figure 1: Prevalence of Major Birth Defects by Organ System, Tennessee, 2011 - 2016



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

Infant Mortality Data and Birth Defects in Tennessee

During the period 2011-2016, there was an average of 572 infant deaths per year (Table 3). While the infant mortality rate decreased from 2011-2013, the rate increased from 2014 -2016. Non-Hispanic Black infants have a mortality rate approximately 2 times higher than Non-Hispanic White infants and more than 2.5 times higher than Hispanic infants.

Table 3. Infant Deaths by Maternal Race/Ethnicity, Tennessee, 2011-2016

	Total		Non-Hispanic White		Non-Hispanic Black		Hispanic	
Year	Number	Rate[1]	Number	Rate	Number	Rate	Number	Rate
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	194	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	182	11.0	33	4.5
2016	597	7.4	345	6.4	195	12.0	38	5.0

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 4 shows the ten leading causes of infant deaths in Tennessee between 2011 and 2016. Birth defects were the leading cause of all infant deaths (22%), followed by preterm birth/low birthweight (15%).

Table 4. Leading Causes of Infant Death, Tennessee, 2011-2016

Rank	Cause of Death	Number of Deaths	Percent of Deaths
1	Birth defects	742	22
2	Preterm birth and low birthweight	513	15
3	Accidents	245	7
4	Sudden infant death syndrome (SIDS)	161	5
5	Maternal complications of pregnancy	120	3
6	Complications of placenta, cord, and membranes	91	3
7	Bacterial sepsis of newborn	85	2
8	Diseases of the circulatory system	76	2
9	Atelectasis (partial lung collapse)	75	2
10	Respiratory distress of newborn	68	2
--	All other causes	1,261	37
--	*All Causes*	*3,437*	*100*

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

Table 5 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, 57% were also born premature (< 37 weeks). Preterm delivery often exacerbates the medical complications faced by infants born with major birth defects. Depending on the type of defect, infants born preterm may be at significantly greater risk of mortality compared to their counterparts delivered at term.[9] This pattern of preterm infants experiencing increased mortality has been demonstrated for neural tube defects,[10] congenital diaphragmatic hernia,[11] and congenital heart defects.[12]

Table 5. Co-Occurrence of Two Leading Causes of Infant Death, Tennessee, 2011-2016

Cause of Infant Death[1]		
	Born Preterm (<37 weeks)	Born Full Term (37+ weeks)
Birth Defect	57%	43%
	Major Birth Defect Present	No Major Birth Defect Present
Preterm birth and low birthweight	3%	97%

1. Represents underlying cause of death recorded on infant's death certificate.
Note: All birth defect or preterm birth/low birthweight related deaths with corresponding birth data available were included. This represented 98 percent of all birth defect deaths and 98 percent of preterm birth/low birthweight deaths.

Data Source: Tennessee Department of Health, Office of Vital Records and Statistics; Tennessee Department of Health, Tennessee Birth Defects Registry.

9. MA, Kirby RS, Meyer RE, et al. The association between major birth defects and preterm birth. Matern Child Health J 2009;13:164–75.
10. 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.
11. 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.
12. Tanner, K., Sabrine, N., & Wren, C. Cardiovascular malformations among preterm infants. Pediatrics 2005; 116(6): e833–e838.

Table 6 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 6. Gestational age at birth of infant deaths caused by birth defects, Tennessee, 2011-2016

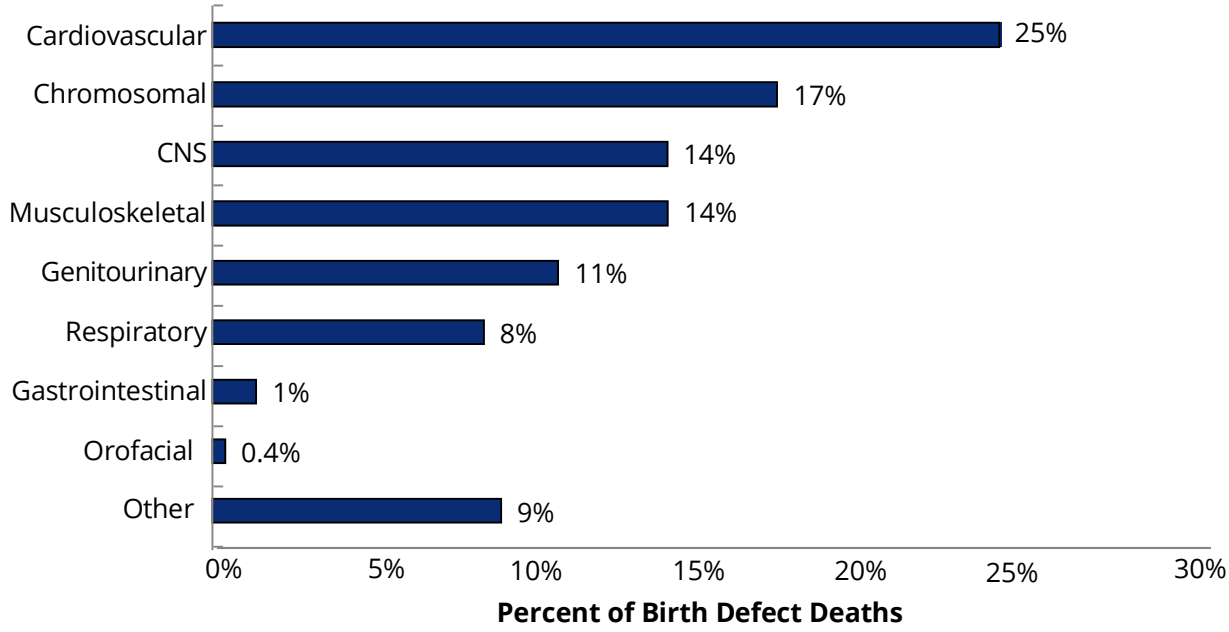
Gestational Age at Birth	Percent of Cases
<28 weeks	10
28-<32 weeks	11
32-<34 weeks	11
34-<37 weeks	26
37+ weeks	42

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 (17%) and central nervous system (14%) 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, 2011-2016.



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

Each Tennessee Birth Defects Report will highlight a specific birth defect, with the goal of providing information on causes, risk factors, and opportunities for prevention. This year's report features birth defects commonly associated with pre-pregnancy diabetes.

Diabetes During Pregnancy

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 2011-2016 birth defects counts, rates, and confidence intervals for 5 organ systems by maternal pre-pregnancy and gestational diabetes are presented in Table 7. The cardiovascular system birth defect rate for mothers with pre-pregnancy diabetes was 661.0 per 10,000 live births and 312.8 per 10,000 live births for mothers with gestational diabetes. The corresponding figure for mothers without pre- pregnancy diabetes was 198.6. This finding suggests that babies born to mothers with pre-pregnancy and gestational 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 7. Prevalence of Major Birth defects by Organ System for Infants Born to Mothers with Pre-Pregnancy Diabetes, Mothers with Gestational Diabetes, and Mother with No Diabetes, 2011-2016.

Organ System	Pre-Pregnancy Diabetes			Gestational Diabetes			No Diabetes		
	Cases	Rate[1]	Relative Rate[2]	Cases	Rate	Relative Rate	Cases	Rate	Relative Rate
Cardiovascular	344	661.0	3.5	875	312.8	1.6	8623	191.5	Reference
Central Nervous System	20	38.4	3.1	38	13.6	1.1	558	12.4	Reference
Genitourinary	77	148.0	2.2	231	82.6	1.2	3064	68.1	Reference
Musculoskeletal	35	67.3	1.8	123	44.0	1.2	1668	37.1	Reference
Orofacial	23	44.2	2.5	54	19.3	1.1	799	17.7	Reference

1. Rate per 10,000 live births.

2. Compares the rate of birth defects for a given organ system amongst infants born to mothers with pre-pregnancy diabetes (or gestational diabetes) to the rate amongst infants born to mothers with no diabetes. Relative risks shown are crude, meaning they are not adjusted for any other factors that could contribute to the observed associations.

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

Diabetes During Pregnancy

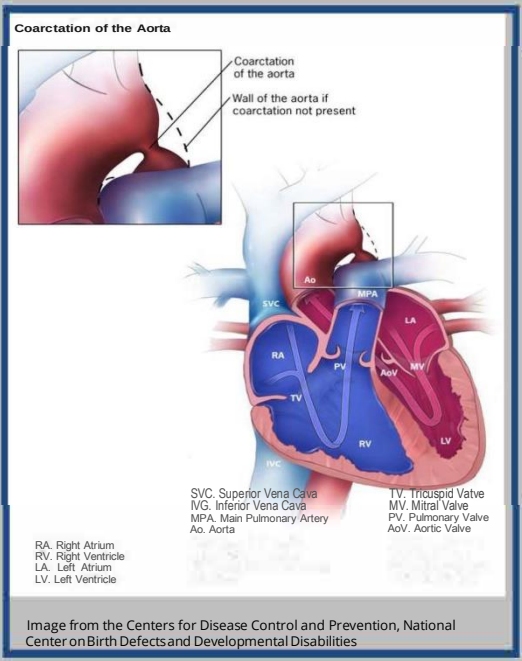


Image from the Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Disabilities

What is Coarcation of the Aorta?

According to the Centers for Disease Control, a Coarcation (pronounced koh-ark-TEY-shun) of the aorta is a birth defect in which a part of the aorta, the tube that carries oxygen-rich blood to the body, is narrower than usual. This may lead to the inability of the heart to pump blood properly throughout the body. The defect may be identified through Tennessee's mandatory Newborn Screening pulse oximetry test of all newborns born in a birthing facility. Surgery can correct the condition. In Tennessee during the years 2011-2016, women with pre-existing diabetes were 2.5 times more likely to give birth to a baby with this defect, compared to women without pre-existing diabetes.

Diabetes Prevention

- **Reach a healthy weight.** Losing even a small amount of weight can prevent or delay diabetes for some people.
- **Stay physically active.** Adults should exercise at least 2 hours and 30 minutes each week (or about 30 minutes per day on most days) of moderate-intensity physical activity such as brisk walking or dancing, swimming or bicycling. Children should get at least an hour of physical activity each day.
- **Choose healthy foods.** Choose fresh vegetables, fruits, and whole grains. Avoid foods that are high in sugar, salt, and fat.
- **Quit smoking.** Smoking raises people's blood sugar, cholesterol, and blood pressure. Quitting can lower risk for heart attack, stroke, nerve damage, and kidney disease. The Tennessee Tobacco Quit Line provides personalized support for Tennesseans who want to quit smoking or chewing tobacco. More information can be found at: https://www.tnquitline.org.

Tennessee Department of Health Resources

- **The National Diabetes Prevention Program** is an evidence-based lifestyle change program for preventing type-2 diabetes. More information can be found at: https://www.tn.gov/health/health-program-areas/mch-diabetes/d/diabetes-prevention-program.html
- **Take Charge of Your Diabetes (the Diabetes Self-Management Program)** is a six-week workshop that provides tools for living a healthy life for people with diabetes. More information can be found at: https://www.tn.gov/health/health-program-areas/mch-diabetes/d/take-charge-of-your-diabetes.html

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.[13] 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.[14]

In Tennessee, babies born to 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 Trisomy 21 (Down Syndrome).[15] In contrast, women younger than 20 years old are more likely to have babies born with gastroschisis, [16] a birth defect of the abdominal wall, than older women.

Figure 4 shows the overall prevalence of birth defects by maternal age group in Tennessee. During 2011-2016, the birth defects prevalence rates were highest among women 40 and older (504.7 per 10,000 live births), followed by women aged 35-39 (382.3 per 10,000 live births) and women less than 20 years old (342.6 per 10,000 live births).

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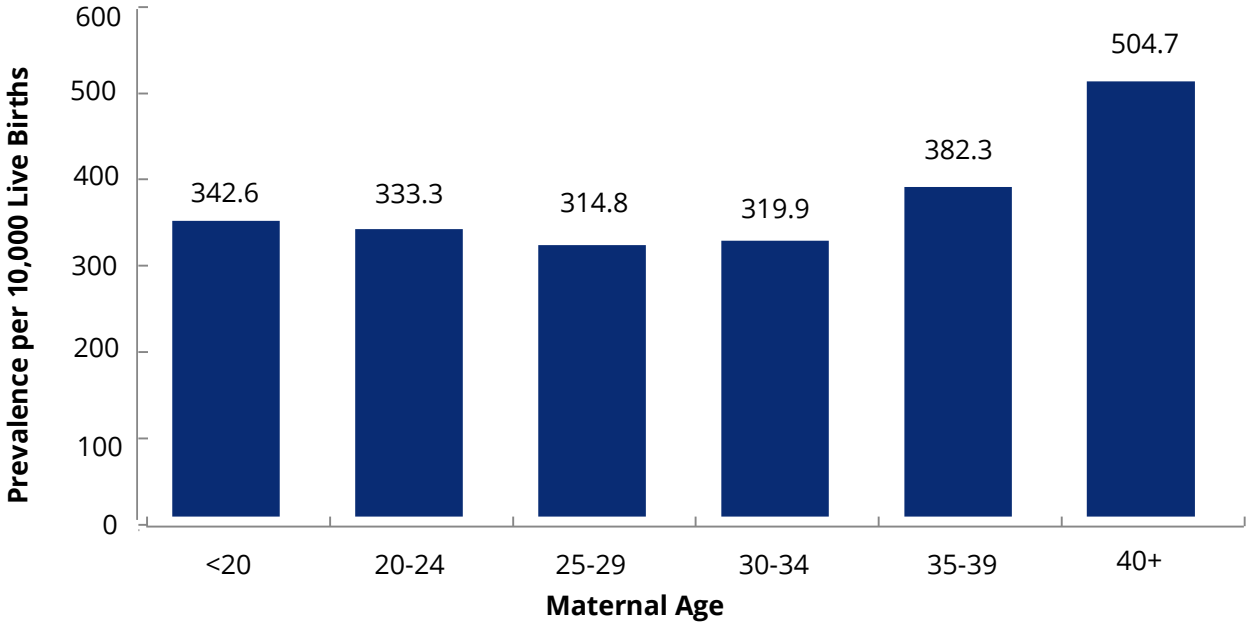
13. Canfield MA, Mai CT, Wang Y, O'Halloran M, Marengo LK, Olney RS, Borger CL, Rutkowski R, Fornoff J, Irwin N, Copeland G, Flood TJ, Meyer RE, Rickard R, Alverson CJ, Sweatlock J, Kirby RS. The Association Between Race/Ethnicity and Major Birth Defects in the United States, 1999 - 2007. American Journal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

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

15. 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.

16. 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. Prevalence of Major Birth Defects by Maternal Age, Tennessee, 2011-2016



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

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 5. Prevalence of Chromosomal Major Birth Defects by Maternal Age, Tennessee, 2011-2016

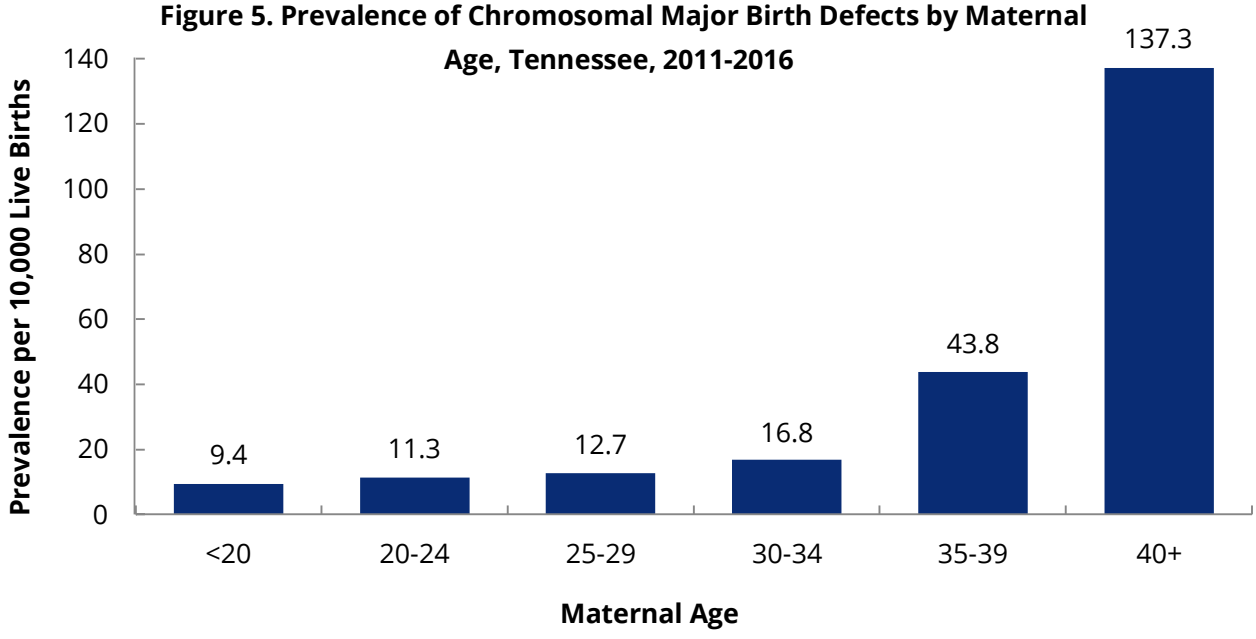
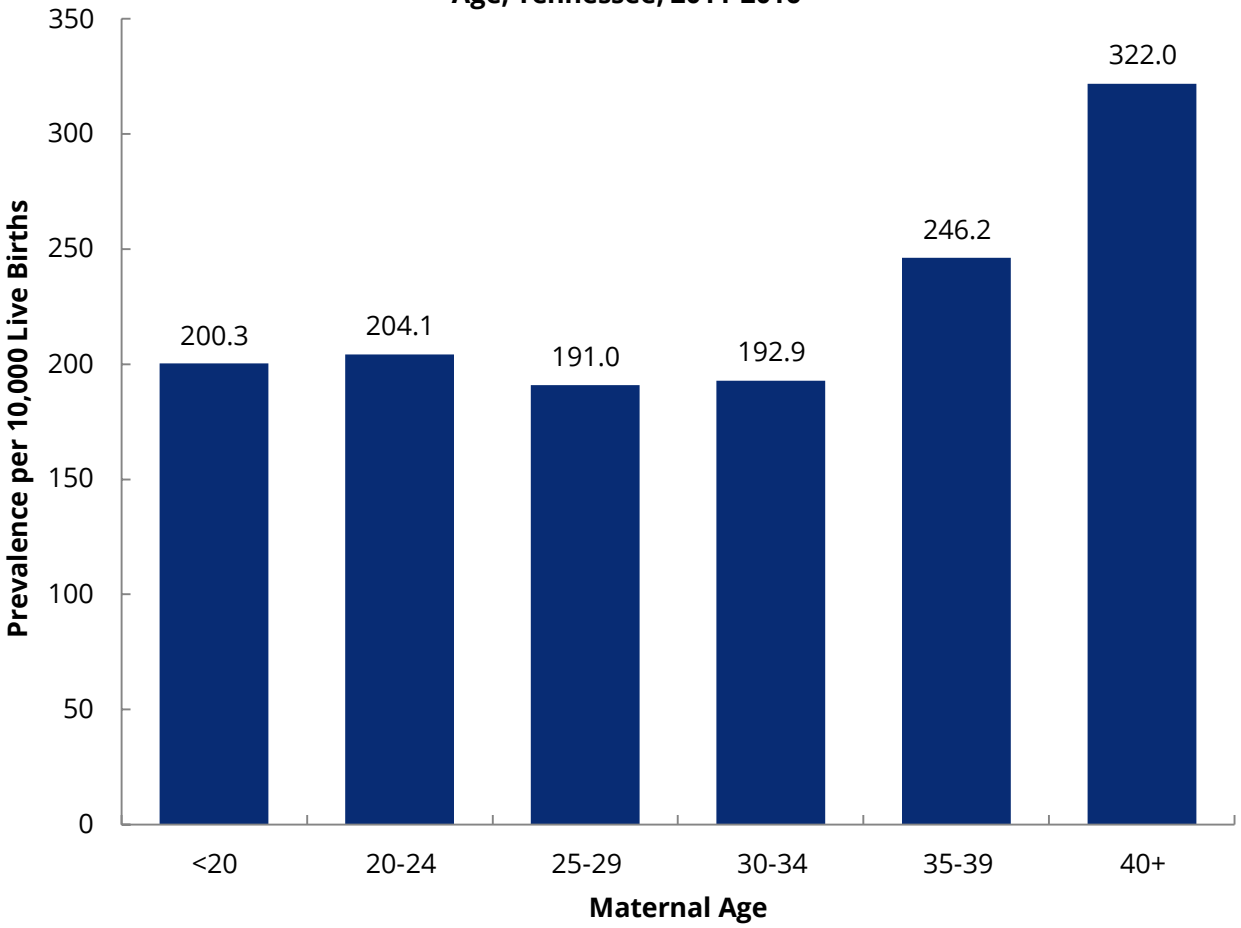


Figure 6. Prevalence of Cardiovascular Major Birth Defects by Maternal Age, Tennessee, 2011-2016



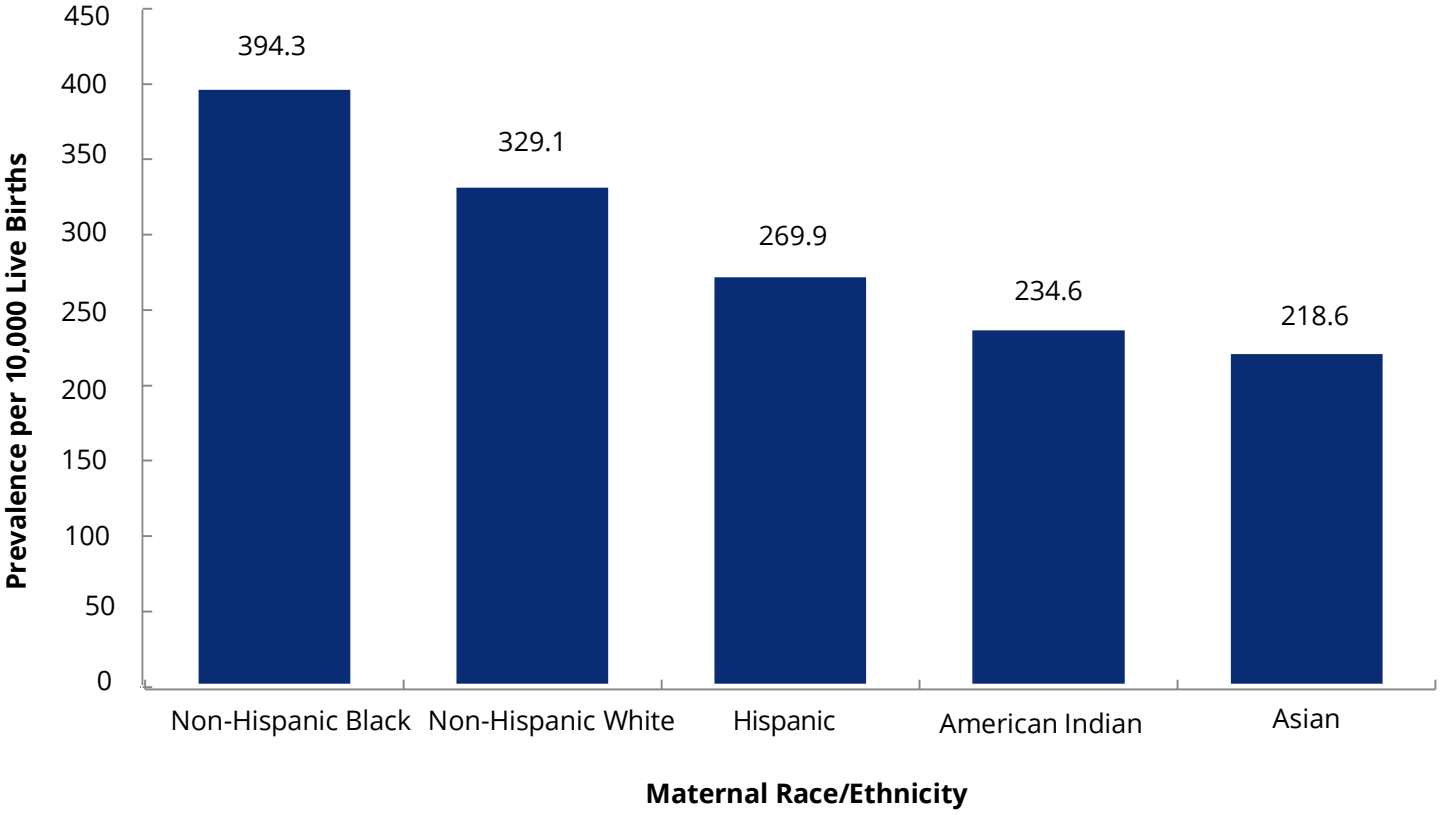
Data Source: Tennessee Department of Health, Tennessee Birth Defects Registry.

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 (322 per 10,000 live births) is about 1.7 times the average rate for babies born to mothers in the age group 25-29 (191 per 10,000 live births).

Maternal Race and Ethnicity

Birth defects prevalence rates are highest for Non-Hispanic Blacks (394.3 per 10,000 live births), followed by Non-Hispanic Whites and Hispanic women (Figure 7). The lowest prevalence rate for birth defects (218.6 per 10,000 live births) was seen among babies born to Asian women.

Figure 7. Prevalence of Major Birth Defects by Maternal Race/Ethnicity, Tennessee, 2011-2016

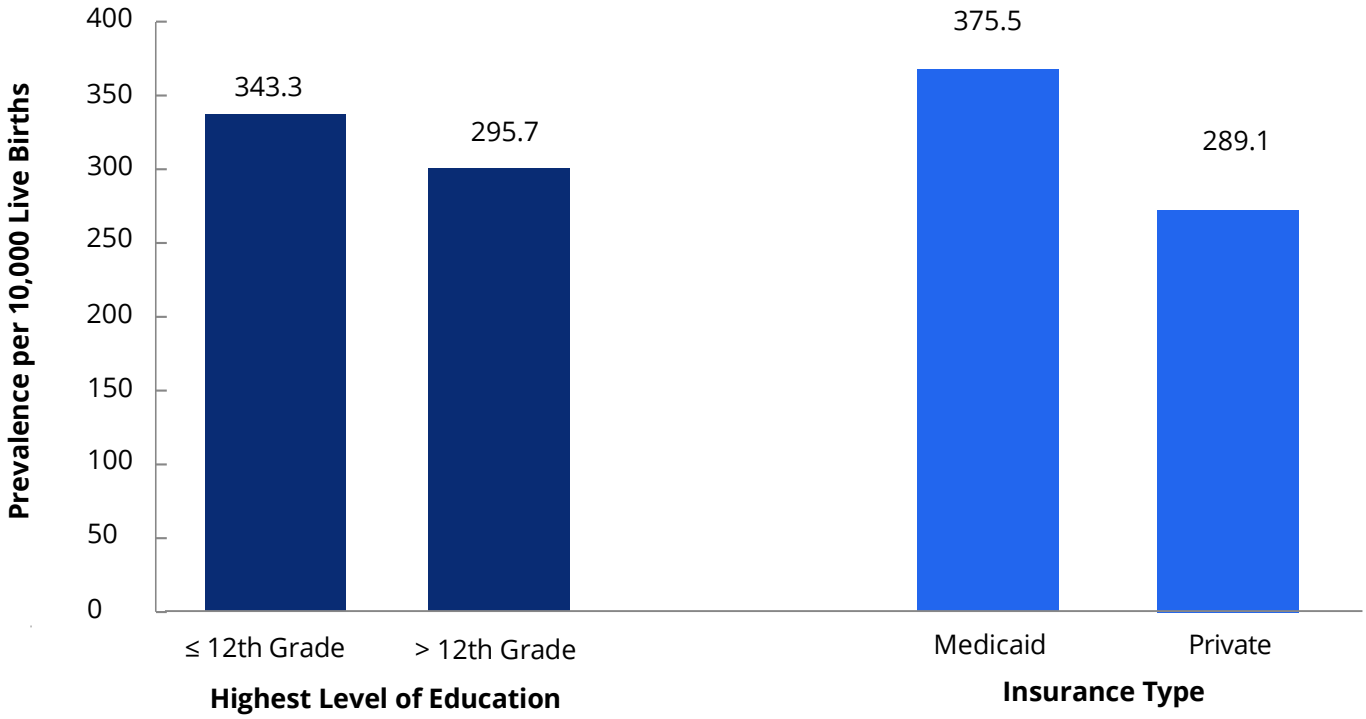


Data Source: Tennessee Department of Health, Tennessee Birth Defects Registry.

Other Maternal Characteristics

Birth defect prevalence rates also differed by education and insurance type. Figure 8 shows that babies born to women with ≤ 12 grade education and women on Medicaid, which highlights the influence of social determinants of health (such as education and income levels) on health outcomes.

Figure 8. Prevalence of Major Birth Defects by Selected Maternal Characteristics, Tennessee, 2011-2016

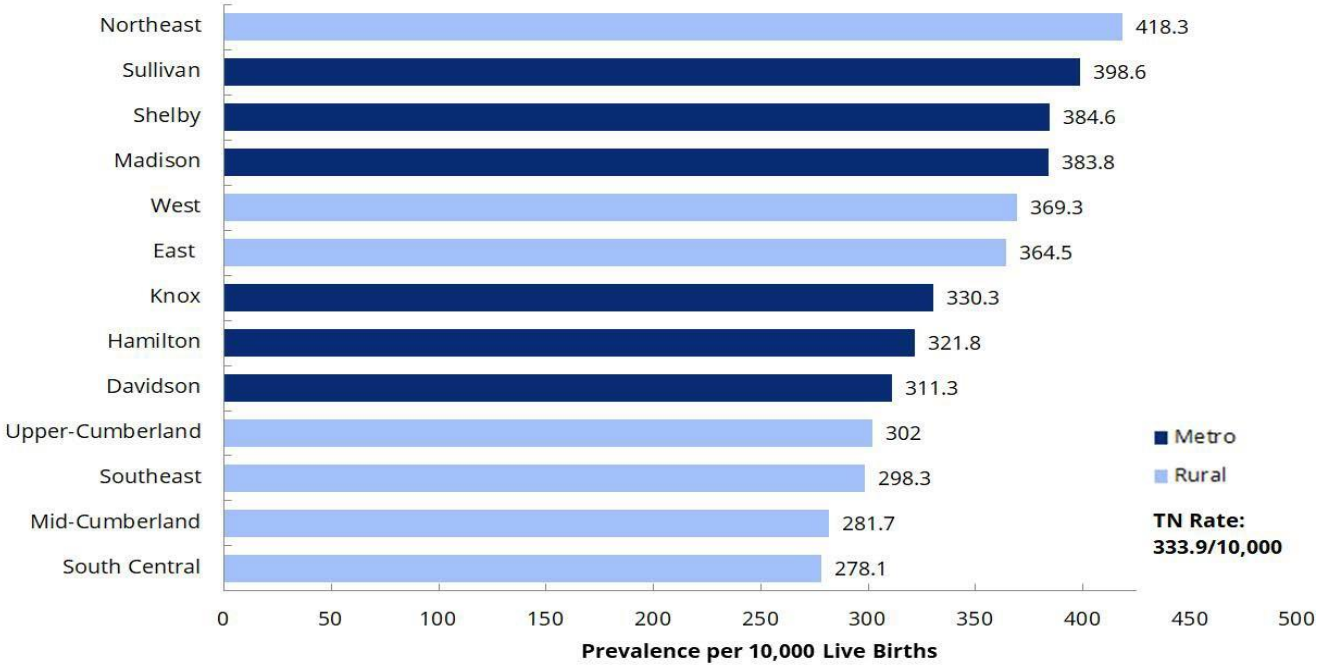


Data Source: Tennessee Department of Health, Tennessee Birth Defects Registry.

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, Shelby and Madison counties.

Figure 9. Prevalence of Major Birth Defects by Maternal Region of Residence, Tennessee, 2011-2016

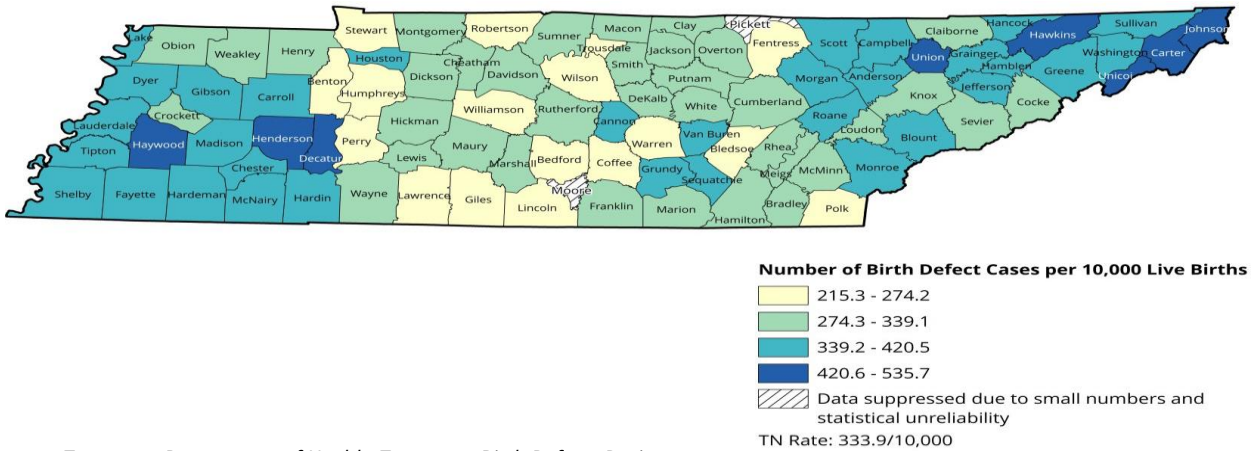


Data Source: Tennessee Department of Health, Tennessee Birth Defects Registry.

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 535.7 per 10,000 live births in Unicoi County compared to 215.3 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, 2011-2016



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 more than 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 1.2 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 1.2 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, 2011-2016[1]

Maternal Health Characteristic		Relative Risk of Birth Defects[2]	95% CI[3]
Diabetes	Pre-existing	2.81	2.57-3.06
	Gestational	1.42	1.34-1.50
	None	Reference	–
Hypertension	Pre-existing	1.89	1.76-2.03
	Gestational	1.43	1.35-1.51
	None	Reference	–
Pregnancy Smoking Status[4]	Smoker	1.24	1.19-1.29
	Non-Smoker	Reference	–
BMI	Underweight	1.10	1.02-1.19
	Normal	Reference	–
	Overweight	1.07	1.03-1.12
	Obese	1.27	1.23-1.32
Prenatal Care[5]	Inadequate	1.44	1.38-1.52
	Intermediate	1.16	1.08-1.23
	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).

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.

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 understand these risks and work with their employers to assure safety measures are in place.

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.[17] These practices help prevent exposure of individuals and their familial contacts to hazardous chemicals.

Preventing Birth Defects

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 and Tdap vaccines are critical during pregnancy for the health of both mother and baby.

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.

17. 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 cerebralventricles 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 9. Cases and Prevalence of Major Birth Defects and Fetal Alcohol Syndrome, Tennessee, 2011-2016

Birth Defect	Number[1]	Rate[2]	95% CI[3]
Central Nervous System			
Anencephaly	88	1.8	1.5-2.2
Encephalocele	61	1.3	1.0-1.6
Holoprosencephaly	281	5.8	5.1-6.5
Spina bifida without anencephaly	222	4.6	4.0-5.2
Total Central Nervous System Cases	618	12.8	11.8-13.8
Total Central Nervous System Defects	652	13.5	12.5-14.5
Eye/Ear			
Anophthalmia/microthalmia	68	1.4	1.1-1.8
Anotia/microtia	61	1.3	1.0-1.6
Congenital cataract	114	2.4	1.9-2.8
Total Eye and Ear Cases	233	4.8	4.2-5.4
Total Eye and Ear Defects	243	5.0	4.4-5.7
Cardiovascular			
Aortic valve stenosis	82	1.7	1.3-2.1
Atrial septal defect	8,057	166.7	163.0-170.4
Atrioventricular septal defect (Endocardial cushion defect)	283	5.9	5.2-6.5
Coarctation of the aorta	415	8.6	7.8-9.4
Common truncus (truncus arteriosus or TA)	46	1.0	0.7-1.3
Double outlet right ventricle (DORV)	155	3.2	2.7-3.7
Ebstein anomaly	83	1.7	1.4-2.1
Hypoplastic left heart syndrome	176	3.6	3.1-4.2
Interrupted aortic arch (IAA)	94	1.9	1.6-2.4
Pulmonary valve atresia and stenosis	496	10.3	9.4-11.2
Single Ventricle	76	1.6	1.2-2.0
Tetralogy of Fallot (TOF)	300	6.2	5.5-6.9
Total anomalous pulmonary venous connection (TAPVC)	63	1.3	1.0-1.7
Transposition of the great arteries (TGA)	249	5.2	4.5-5.8
Tricuspid valve atresia and stenosis	69	1.4	1.1-1.8
Ventricular septal defect	2,444	50.6	48.6-52.6
Total Cardiovascular Cases	9,842	203.6	199.6-207.7
Total Cardiovascular Defects	13,088	270.8	266.1-275.5
Orofacial			
Choanal atresia	101	2.1	1.7-2.5
Cleft lip with cleft palate	318	6.6	5.9-7.3
Cleft lip alone (without cleft palate)	135	2.8	2.3-3.3
Cleft palate alone (without cleft lip)	326	6.7	6.0-7.5
Total Orofacial Cases	876	18.1	16.9-19.3
Total Orofacial Defects	880	18.2	17.0-19.4

Continued on next page

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

Birth Defect	Number	Rate	95% CI
Gastrointestinal			
Biliary atresia	110	2.3	1.9-2.7
Esophageal atresia/tracheoesophageal fistula	152	3.1	2.6-3.6
Rectal and large intestinal atresia/stenosis	250	5.2	4.5-5.8
Small intestinal atresia/stenosis	247	5.1	4.5-5.7
Total Gastrointestinal Cases	703	14.5	13.5-15.6
Total Gastrointestinal Defects	759	15.7	14.6-16.8
Genitourinary			
Bladder exstrophy	12	0.2	0.1-0.4
Cloacal exstrophy	371	2.1	1.7-2.5
Congenital Posterior Urethral Valves	72	2.9	2.3-3.7
Hypospadias	2,648	107.1	103.0-111.2
Renal agenesis/hypoplasia	300	6.2	5.5-6.9
Total Genitourinary Cases	3,372	69.8	67.4-72.1
Total Genitourinary Defects	3,403	70.4	68.0-72.8
Musculoskeletal			
Clubfoot	913	18.9	17.7-20.1
Diaphragmatic hernia	194	4.0	3.4-4.6
Gastroschisis	262	5.4	4.8-6.1
Limb deficiencies (reduction defects)	200	4.1	3.6-4.7
Omphalocele	127	2.6	2.2-3.1
Craniosynostosis[4]	191	11.8	10.1-13.5
Total Musculoskeletal Cases	1,826	37.8	36-39.5
Total Musculoskeletal Defects	1,887	39.0	37.3-40.8
Chromosomal			
Deletion 22q11.2	11	0.2	0.1-0.4
Trisomy 13	48	1.0	0.7-1.3
Trisomy 18	79	1.6	1.3-2
Trisomy 21 (Down syndrome)	718	14.9	13.8-15.9
Turner syndrome	39	1.7	1.2-2.3
Total Chromosomal Cases	891	18.4	17.2-19.6
Total Chromosomal Defects	895	18.5	17.3-19.7
Total Birth Defects Cases	16,138	333.9	328.6-339.1
Total Birth Defects	21,807	451.2	445-457.3
Fetal Alcohol Syndrome[5]	69	1.4	1.1-1.8

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. Includes cases from only 2015 and 2016. Prior to 2015, craniosynostosis cases could not be identified. Rate is calculated using live births from 2015 and 2016.
5. 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:
https://www.tnaap.org/programs/tennessee-medical-home/tennessee-medical-home-overview

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

Family Voices of Tennessee:
https://www.familyvoicestn.org/

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:
https://dsagtn.org/

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

Understand Your Child's Diagnosis of Down Syndrome:
https://vkc.mc.vanderbilt.edu/assets/files/resources/DS%20Guide%20for%20Parents.pdf

Understand Your Patient's Diagnosis of Down Syndrome:
https://vkc.mc.vanderbilt.edu/assets/files/resources/DS%20Guide%20for%20Doctors.pdf

Baby & Me - Tobacco Free™:
https://www.tn.gov/health/health-program-areas/fhw/baby-me-tobacco-free.html

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

Take Charge of Your Diabetes:
https://ag.tennessee.edu/fcs/Pages/Health/TakeChargeOfYourDiabetes.aspx

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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