

# Birth Defects Among 1999-2011 Deliveries:

Summary and Key Findings from the Texas Birth  
Defects Registry's Report of Birth Defects Among 1999-  
2011 Deliveries

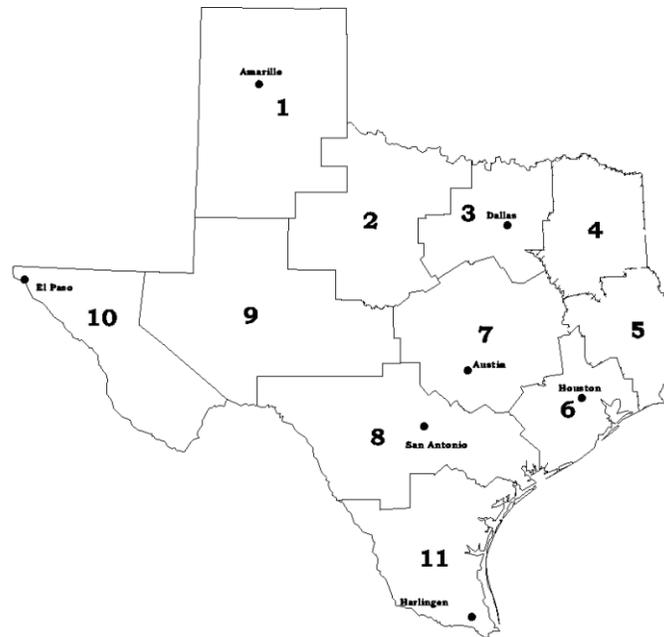
# Table of Contents

<b>Methods</b> .....	<b>2</b>
Case Definition .....	2
Data Collection .....	3
Data Analysis.....	3
Limitations.....	5
Acknowledgements.....	5
<b>Results</b> .....	<b>6</b>
Overall .....	6
Time .....	6
Location.....	8
Sex of Infant .....	9
Maternal Race/Ethnicity .....	10
Maternal Age.....	12
<b>Conclusions</b> .....	<b>14</b>
<b>References</b> .....	<b>14</b>

The Texas Birth Defect Registry’s Report of Birth Defects Among 1999-2011 Deliveries can be found at: <https://www.dshs.state.tx.us/birthdefects/Data/reports.shtm>. Requests for additional information may be sent to [BirthDefects@dshs.state.tx.us](mailto:BirthDefects@dshs.state.tx.us).

## Methods

This document discusses some of the findings in the Texas Birth Defects Registry's Report of Birth Defects Among 1999-2011 Deliveries, which presents information on selected birth defects among deliveries during 1999 through 2011 to women who were residents of Texas at the time of delivery.



### Case Definition

To be included as a case in the Texas Birth Defects Registry, all of the following criteria must be met:

- The mother's residence at the time of delivery must be in an area covered by the registry. During 1999-2011, the registry covered the entire state of Texas.
- The infant or fetus must have a structural birth defect or developmental disability monitored by the registry.
- The defect must be diagnosed prenatally or within one year after delivery. This is extended to six years of age for special cases, currently only for fetal alcohol syndrome.

The current case definition includes all pregnancy outcomes (live births, spontaneous fetal deaths, and induced pregnancy terminations) at all lengths of gestation. Prior to April 5, 2001, when the current case definition was adopted, the registry did not collect information on birth defects among fetal deaths before 20 weeks gestation. Most 1999 and much of year 2000

surveillance activities were completed at the time this case definition went into effect. As a result, the 1999 and 2000 data in the Texas Birth Defects Registry include only a very small number of fetal deaths before 20 weeks gestation.

### **Data Collection**

The Texas Birth Defects Registry uses active surveillance. This means it does not require reporting by hospitals or medical professionals. Instead, trained program staff members regularly visit medical facilities where they have the authority to review logs, hospital discharge lists, and other records. From this review, a list of potential cases is created. Starting with deliveries during 2009, we began also using Texas fetal death certificates with a congenital anomaly as the underlying cause of death (codes Q00.0 through Q99.9, International Classification of Diseases, Tenth Revision (ICD-10)) to identify potential cases. Program staff then review medical charts for each potential case identified. If the infant or fetus has a birth defect covered by the registry, detailed demographic and diagnostic information is abstracted. That information is entered into the computer and submitted for processing into the registry. Quality control procedures for finding cases, abstracting information, and coding birth defects help ensure completeness and accuracy.

Records in the birth defects registry were matched to birth certificates and fetal death certificates filed with the Vital Statistics Unit of the Texas Department of State Health Services. When a record in the birth defects registry matched a birth or fetal death certificate, and information was not missing from the matching certificate, the analysis for this report used demographic data from the birth or fetal death certificate for the following: date of delivery, mother's date of birth, mother's race/ethnicity, and mother's county of residence at the time of delivery. Information on the sex of the infant or fetus was handled a bit differently. We used the sex reported on the matching birth or fetal death certificate unless information abstracted from medical records indicated the sex was ambiguous, in which case we used the information from medical records. When a registry record did not match a birth or fetal death certificate, or when information was missing from the certificate, then this report used demographic data abstracted from medical records.

Regardless of the source of demographic information for this report, all diagnostic information was abstracted from medical records.

### **Data Analysis**

The Texas Birth Defects Registry's Report of Birth Defects Among 1999-2011 Deliveries presents results for 48 selected birth defects, regardless of whether the defect occurred alone or

together with others. Appendix B of the report lists the BPA codes used to define these defects, and Appendix C provides a glossary of birth defects and related terms.

Because an infant or fetus often has more than one defect, and not all monitored defects are included in these analyses, it is not meaningful to sum all diagnostic categories in the tables to obtain the total number of children with birth defects. In the data tables, totals are shown in the line labeled, “Infants and fetuses with any monitored birth defect.”

Tables in the report include the number of cases found, the estimated prevalence per 10,000 live births, and the 95% confidence interval for the prevalence. A case is an infant or fetus with the specified birth defect. Birth prevalence was calculated as follows:

$$\frac{\text{Number of cases}}{\text{Total number of live births}} \times 10,000$$

The denominators used in calculating prevalence are shown in Appendix A of the Report of Birth Defects Among 1999-2011 Deliveries.

The prevalence is an estimate of the true prevalence, which can never be known with certainty. The 95% confidence interval contains the true prevalence of a birth defect 95% of the time. A wide interval indicates the uncertainty stemming from small numbers. The report displays 95% confidence intervals based on the Poisson distribution when there are 100 or fewer cases, and based on the normal distribution when there are more than 100 cases.

We used Poisson regression to identify birth defects with statistically significant differences in prevalence between maternal age groups, maternal race/ethnic groups, and between males and females. These birth defects are marked with an asterisk in Tables 3, 4, and 5 of the Report of Birth Defects Among 1999-2011 Deliveries.

Another simpler approach to determine whether the prevalence of a particular birth defect differs between groups, for example, between males and females, is to examine the 95% confidence intervals for the prevalences. If the 95% confidence interval for the prevalence among males does not overlap with the 95% confidence interval for females, we consider the prevalences to be statistically significantly different. However, this method is more conservative and has less power than Poisson regression, and will identify fewer significant differences between groups than Poisson regression.

Readers who compare Table 2 of the Report of Birth Defects Among 1999-2011 Deliveries to previous reports may notice that the prevalence of “Infants and fetuses with any monitored birth defect” decreased slightly for the years 1999 through 2006. This occurred because we removed from the registry the records of infants and fetuses whose birth defect diagnoses were all “conditional inclusion” defects according to the registry’s birth defects code list, revision date 6/29/2007. Conditional inclusion defects are diagnoses that should only be entered into the registry if the infant or fetus has at least one birth defect diagnosis that is not a conditional inclusion defect. The decreases in the prevalence of “Infants and fetuses with any monitored birth defect” ranged from about a 3% decrease in prevalence for 1999 and 2000 to less than a 1% decrease in prevalence for 2006, compared to previous reports.

### **Limitations of these Data**

These data are subject to several limitations. First of all, the registry only includes birth defects diagnosed within one year after delivery (except for fetal alcohol syndrome), so birth defects detected after the first birthday and diagnoses that are refined after the first birthday are not in the registry. Second, we miss diagnoses that are made outside of Texas or in Texas facilities that our staff does not access at this time, such as prenatal diagnostic facilities and private physicians’ offices. Third, data are collected from medical records and as such are subject to differences in clinical practice.

### **Acknowledgements**

The Department of State Health Services continues to work on behalf of children and families affected by birth defects in Texas, and recognizes the critical contributions of families who have participated in research that one day will eliminate these conditions. We further acknowledge the dedicated efforts of the birth defects surveillance staff, who collect information on birth defects across the state.

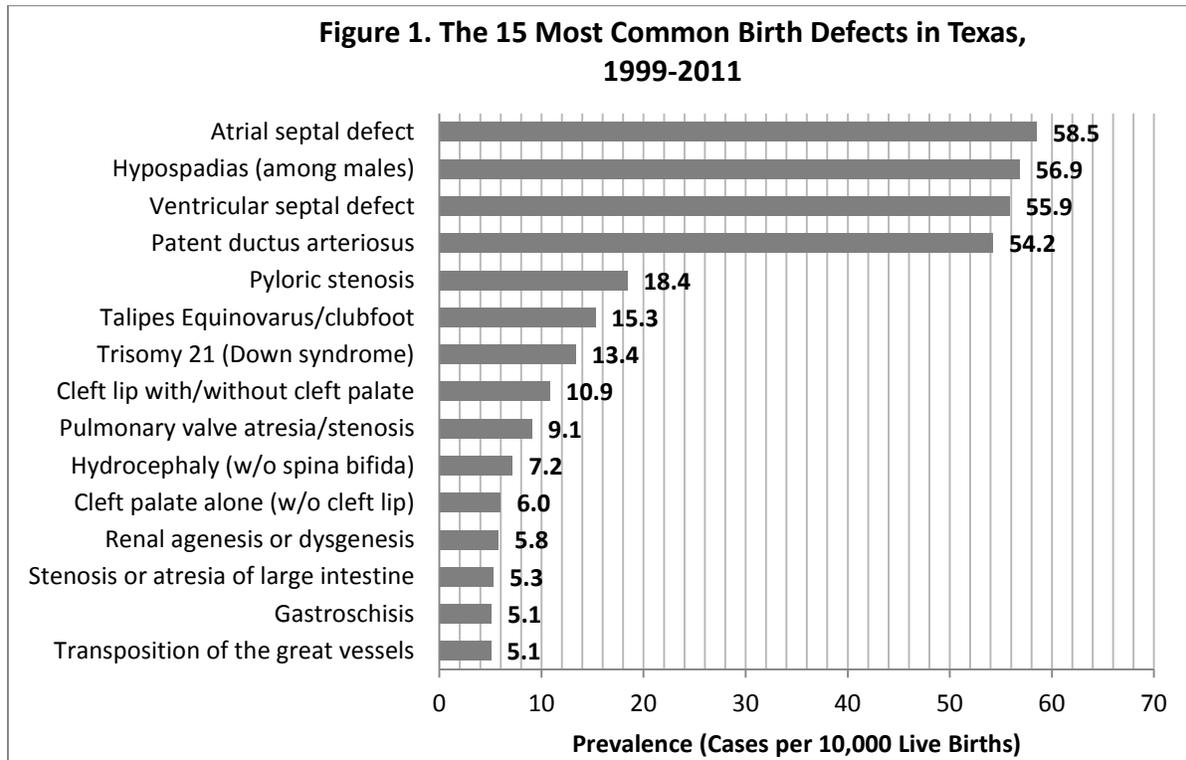
The work of the Texas Birth Defects Epidemiology and Surveillance Branch is supported by the Texas General Revenue Fund and the Title V Maternal and Child Health Block Grant Funds, Office of Title V and Family Health, Department of State Health Services. Through May 30, 2014, research activities were supported in part by Cooperative Agreement 5U01DD000494 from the Centers for Disease Control and Prevention.

## Results

While the causes of many birth defects are unknown, data from the registry show interesting patterns and trends that can give public health professionals and scientists clues about what may cause birth defects. The sections below highlight some interesting patterns in birth defects.

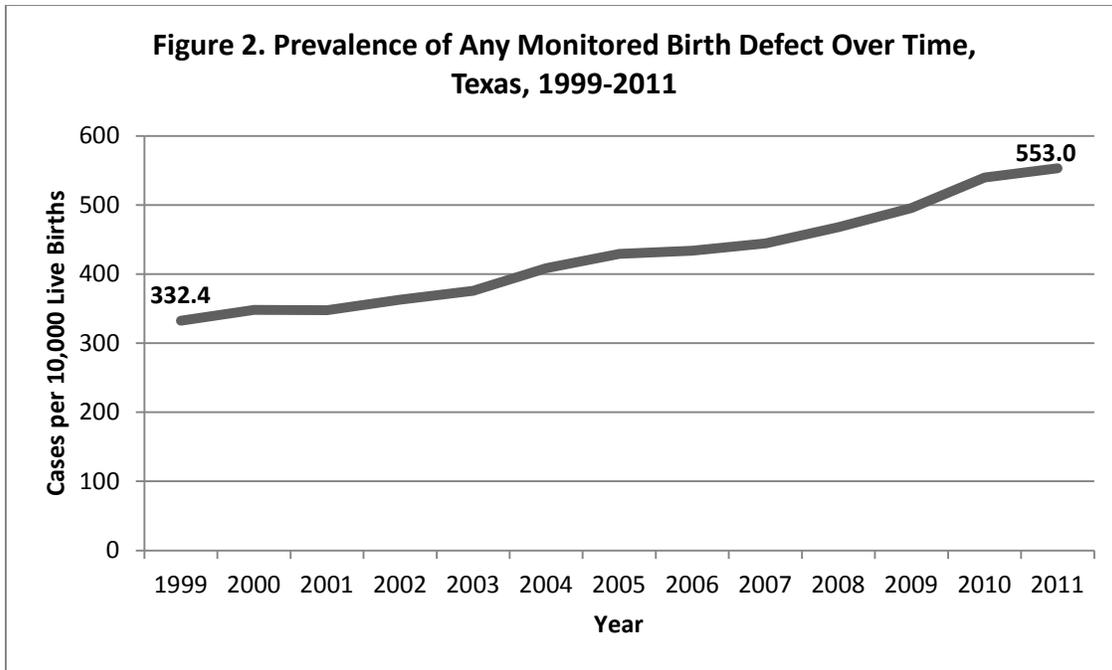
### Overall

Between 1999 and 2011, about 428 babies with a structural or chromosomal birth defect were born per every 10,000 live births (or 4.3%) to Texas residents. The most common birth defect was atrial septal defect, a heart defect with a prevalence of 59 cases per 10,000 live births (0.6% or one in every 169 births). In fact, five of the 15 leading birth defects involve the cardiovascular system.

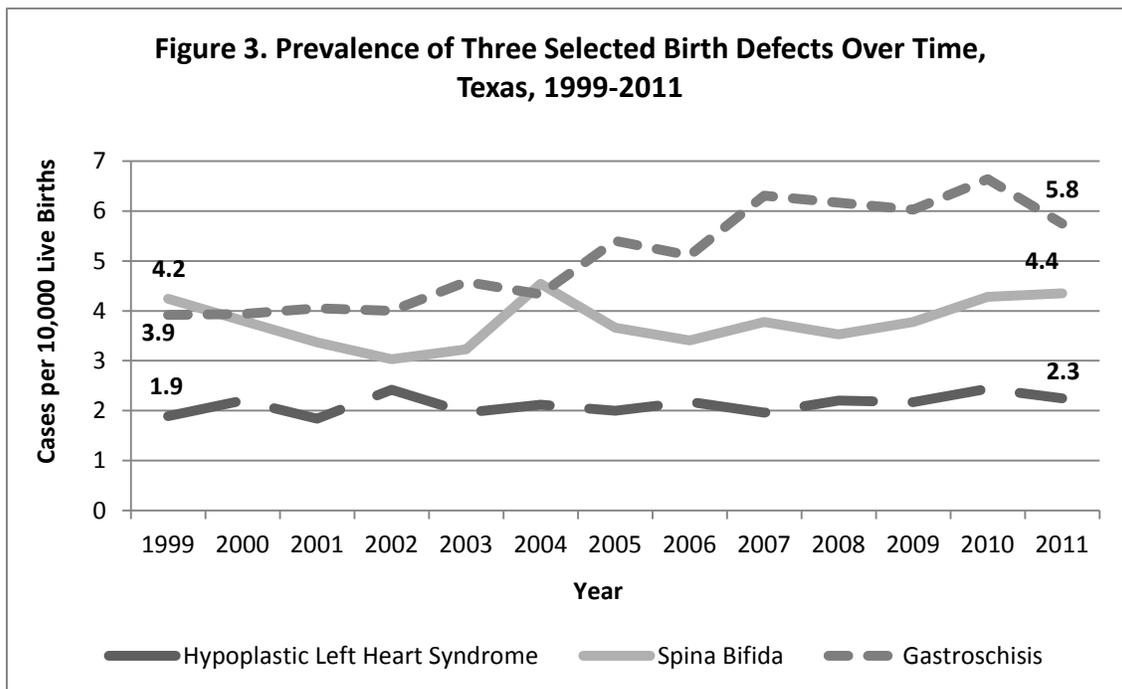


### Time

Overall, 3.3% (332.4 cases per 10,000 live births) of all babies born in 1999 were born with one or more structural malformations or chromosomal anomalies. By 2011, this rose to 5.5% (553.0 cases per 10,000 live births). Figure 2 on the next page shows the prevalence of any monitored birth defect from 1999 to 2011.



Although the overall prevalence of birth defects is increasing with time, not all birth defects follow this trend. The chart below shows different time trends from 1999 to 2011 for three selected birth defects: hypoplastic left heart syndrome, spina bifida, and gastroschisis.



Langlois *et al.* (2011) recently studied time trends for birth defects in Texas from 1999-2007. In this analysis, the authors found a 3.6% increase in the prevalence of total birth defects per year. Increases remained significant after adjustment for demographic characteristics. Larger upward trends were observed in defects that had been rated as more susceptible to diagnostic variation, suggesting that the overall increases may not be due to increased occurrence, but rather due to increased awareness, documentation in health care facilities, and detection of certain birth defects before the first birthday.

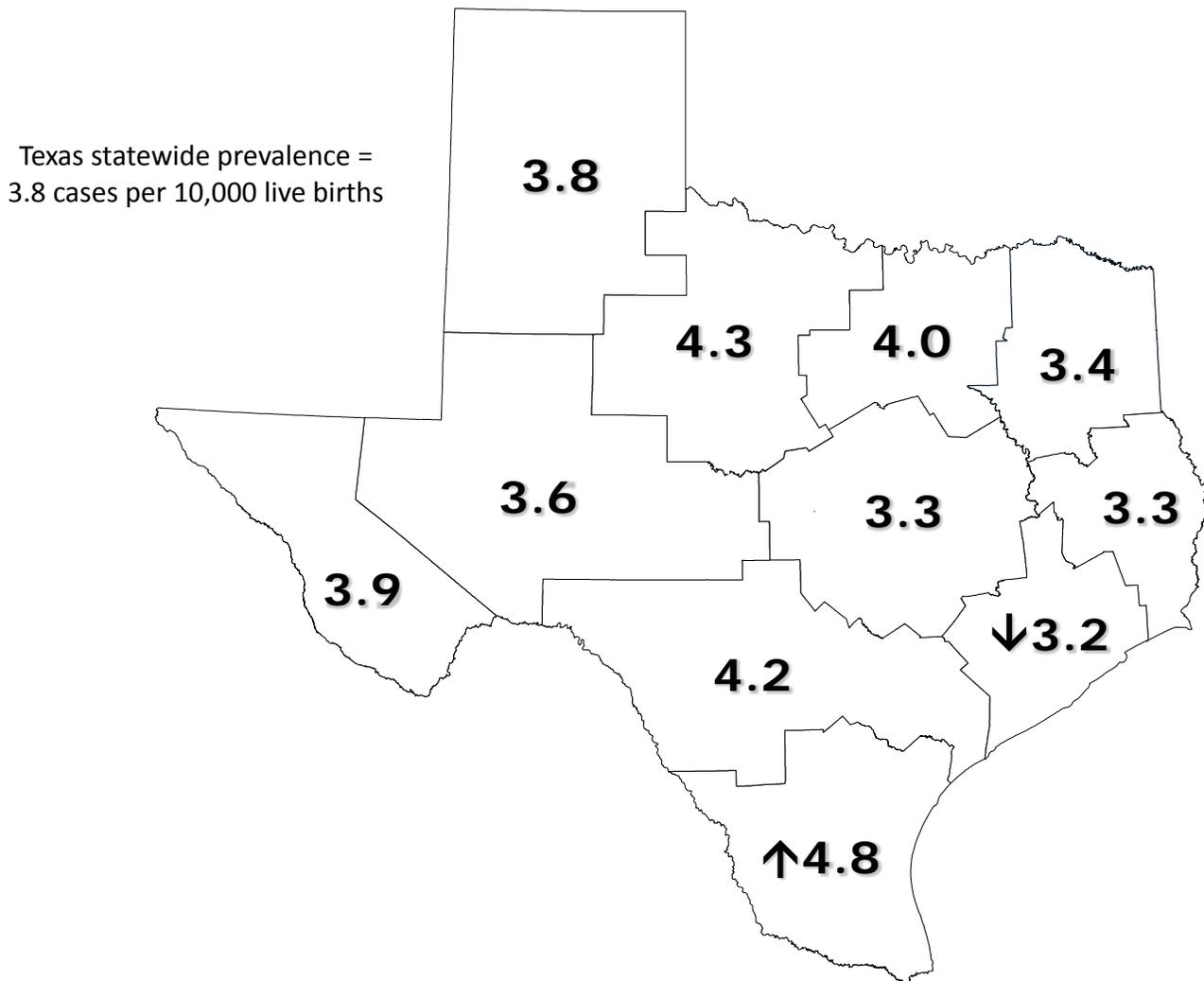
**Location:**

The prevalence of some birth defects varies by region of the state. The state is divided into 11 health service regions (Region 1 through 11) in the following general areas:

- Region 1 (Panhandle)
- Region 2 (Abilene/Wichita Falls)
- Region 3 (Dallas/Fort Worth)
- Region 4 (Northeast Texas)
- Region 5 (Southeast Texas)
- Region 6 (Houston/Galveston)
- Region 7 (Austin/Waco)
- Region 8 (San Antonio)
- Region 9 (West Texas)
- Region 10 (El Paso/Far West Texas)
- Region 11 (South Texas)

The map on the next page shows one such example for spina bifida. The region shown with a downward arrow on the map had a statistically significantly lower birth prevalence of spina bifida (3.2 cases per 10,000 live births) compared to the overall prevalence of spina bifida in the state (3.8 cases per 10,000 live births). Additionally, the region shown with an upward arrow had a significantly higher prevalence of spina bifida (4.8 cases per 10,000 live births) compared to the overall prevalence of spina bifida in the state.

**Figure 4. Prevalence of Spina Bifida, in Cases per 10,000 Live Births, Texas, 1999-2011**



**Sex of Infant:**

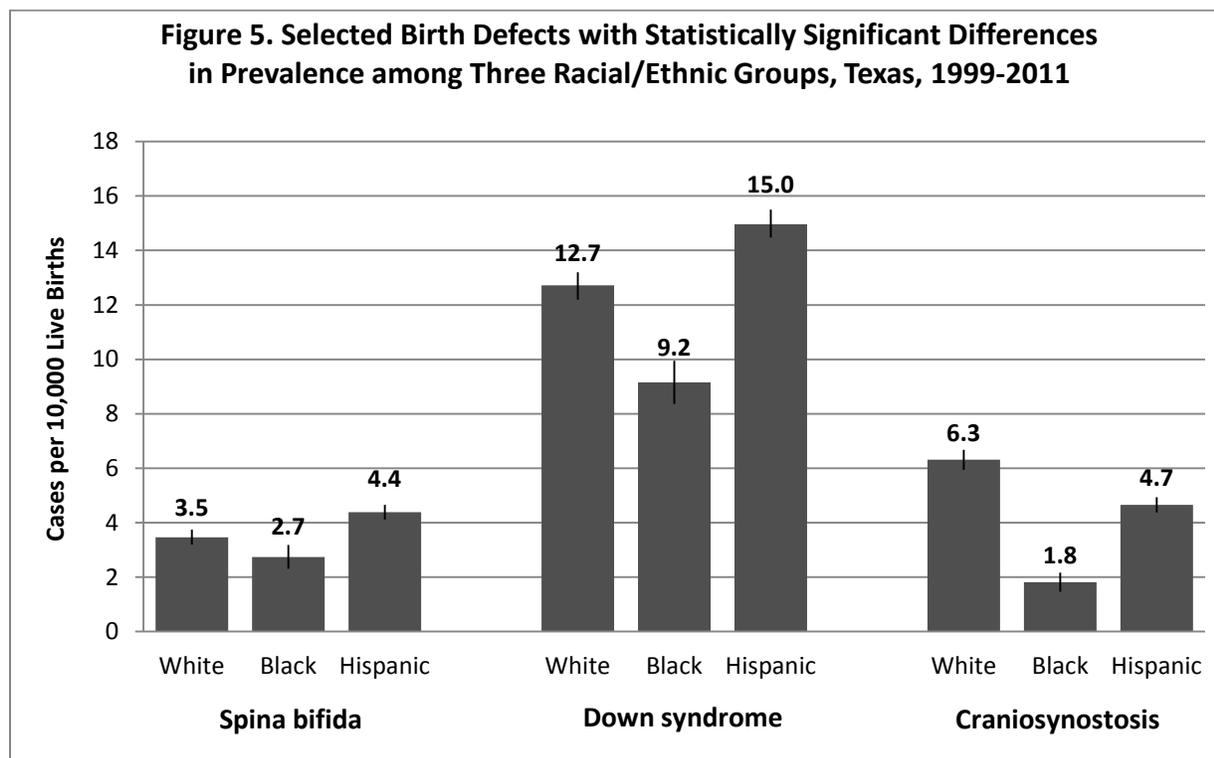
While the causes of many birth defects are unknown, the sex of the infant seems to play a role in the prevalence of some birth defects. Many birth defects tend to occur more often in males or more often in females, and there are a few birth defects that show very large differences in prevalence between the two sexes. For example, pyloric stenosis (which is an excessive narrowing of the opening between the bottom of the stomach and the small intestine) occurs about 6.6 times per 10,000 live births in females, whereas in males, it occurs about 29.8 times per 10,000 live births. This means that boys are about 4.5 times more likely to be born with pyloric stenosis. However, females are about 2.7 times more likely to be born with congenital hip dislocation (due to improper formation of the hip socket), and males are about 1.5 times more likely than females to be born with cleft lip with or without cleft palate.

Of the 48 birth defects that are shown in the Report of Birth Defects Among 1999-2011 Deliveries (not including any monitored birth defect), 22 conditions are more likely to occur in males than females, and 11 conditions are more likely to occur in females than males; the prevalence of the remaining 15 birth defects were similar in both sexes. Overall, boys are 1.4 times more likely than girls to be born with any monitored birth defect (i.e., one or more major structural malformations or chromosomal disorders).

**Maternal Race/Ethnicity:**

There are also some significant differences in the prevalence of certain birth defects across maternal racial/ethnic groups. Non-Hispanic White mothers are 9% more likely than Non-Hispanic Black mothers and 7% more likely than Hispanic mothers to have a baby with any monitored birth defect.

Some birth defects have statistically significant differences in prevalence across all three racial/ethnic groups. Spina bifida, Down syndrome, and craniosynostosis (shown below) each have a prevalence that is higher in one racial/ethnic group and lower in another.



There are other birth defects with statistically significant differences in prevalence between Non-Hispanic Whites, Non-Hispanic Blacks, and Hispanics. The table on the next page shows birth defects which have higher or lower prevalence in one racial/ethnic group when compared to the other two groups.

**Table 1. Birth defects that have statistically significantly higher or lower prevalence among births to mothers of selected racial/ethnic groups, compared to the other two racial/ethnic groups, Texas, 1999-2011<sup>1</sup>**

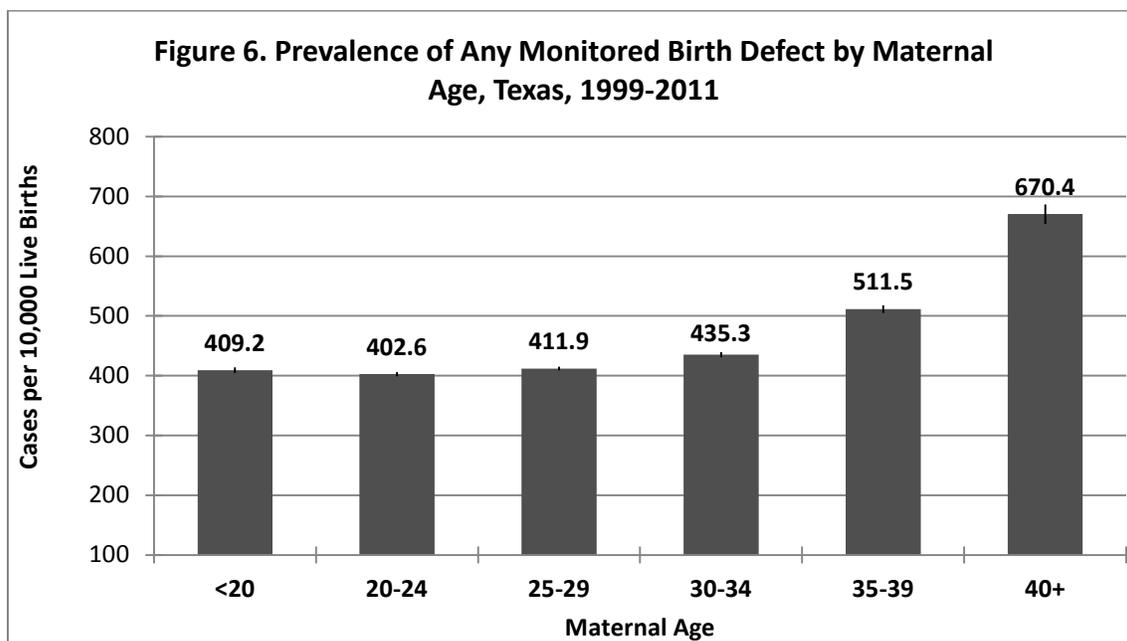
<b>Maternal Race/Ethnicity</b>	<b>Higher Prevalence<sup>2</sup></b>	<b>Lower Prevalence<sup>2</sup></b>
<b>Non-Hispanic Whites</b>	Cleft Palate (without Cleft Lip) Craniosynostosis Hypospadias Tracheoesophageal fistula/esophageal atresia	Atrial Septal Defect
<b>Hispanics</b>	Anencephaly Spina Bifida Anotia/Microtia Down syndrome Ventricular Septal Defect Patent Ductus Arteriosus Gastroschisis	Hypospadias Epispadias Hirschsprung disease Atrioventricular septal defect (endocardial cushion defect)
<b>Non-Hispanic Blacks</b>	Hirschsprung disease	Ventricular Septal Defect Aortic Valve Stenosis Coarctation of the Aorta Cleft Palate (without Cleft Lip) Cleft Lip with or without Cleft Palate Pyloric Stenosis Stenosis or Atresia of the Large Intestine, Rectum, or Anal Canal Congenital Hip Dislocation Craniosynostosis Gastroschisis Down syndrome Spina Bifida Anotia/Microtia

<sup>1</sup>Table adapted from Key Findings: Differences in Major Birth Defects | Birth Defects | NCBDDD | CDC. Retrieved from <http://www.cdc.gov/ncbddd/birthdefects/features/raciaethnicdifferences.html>

<sup>2</sup>There was a statistically significant higher or lower prevalence compared to the other two racial/ethnic groups (e.g., Non-Hispanic Whites compared to Hispanics and Non-Hispanic Blacks).

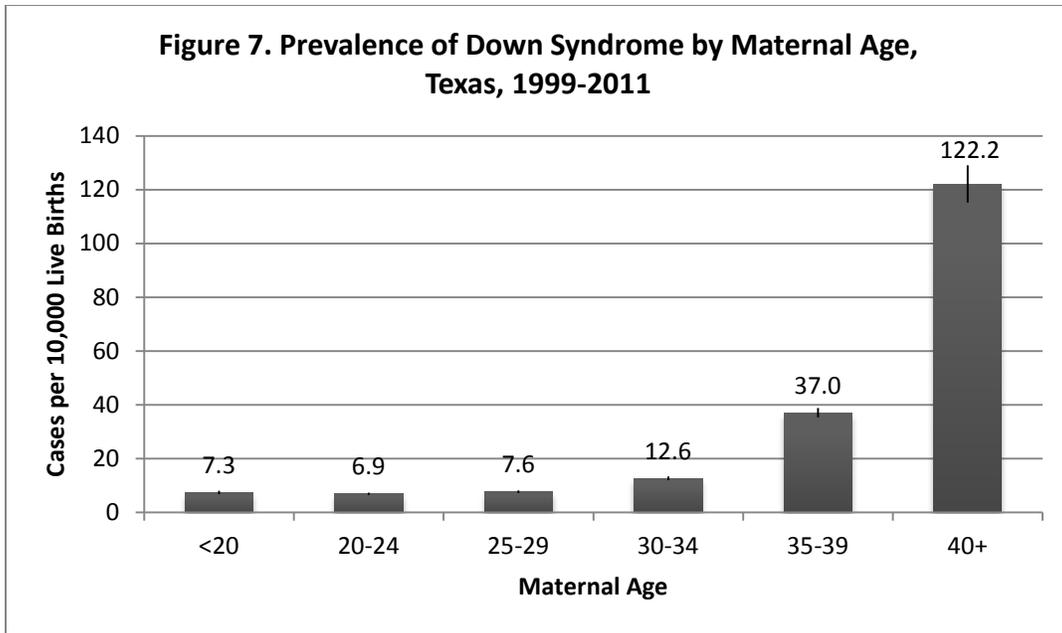
### Maternal Age:

In Texas from 1999-2011, the prevalence of any monitored birth defect in babies born to women 40 years of age or older was significantly higher than in babies born to women under 40. Further, the prevalence of birth defects in babies born to women aged 35-39 was higher than in those born to women under 35, and the prevalence of birth defects in babies born to women aged 30-34 was higher than in those born to women under 30 years of age.

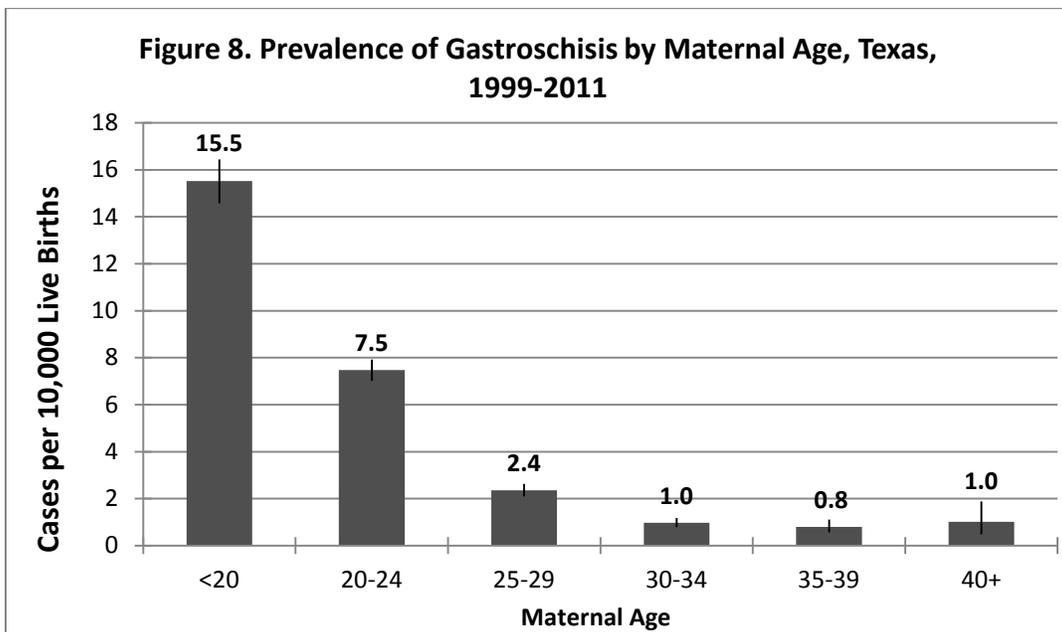


In fact, older mothers (age 40+) were about 1.7 times more likely than mothers aged 24 or younger to have a baby with a birth defect. Of the 13 circulatory and heart defects shown in the Report of Birth Defects Among 1999-2011 Deliveries, 11 had significant differences in prevalence between younger and older mothers. For example, mothers aged 40+ were 5.8 times more likely than mothers aged 24 or younger to have an infant with atrioventricular septal defect (a heart septum defect). Additionally, mothers aged 40+ were about 2.9 times more likely than mothers aged 24 or younger to have an infant with patent ductus arteriosus (a condition where a blood vessel in the heart fails to close soon after birth as it is supposed to).

One of the largest differences in the prevalence of birth defects in babies born to younger mothers versus older mothers occurred in Trisomy 21 (Down syndrome). The prevalence of Trisomy 21 in babies born to mothers aged 40+ was about 122 cases per 10,000 live births; whereas, the prevalence in babies born to mothers aged 20 or younger was about 7 cases per 10,000 live births. This is shown on the chart on the next page.



While many defects were more likely to occur among births to older mothers, there were some defects, such as pyloric stenosis, that were more common among births to younger mothers. Women less than 20 years of age were about twice as likely to have a baby with pyloric stenosis compared to women aged 40+. Gastroschisis is another birth defect with a notably higher prevalence among births to younger women than older women. As shown in the chart below, there were about 16 babies with gastroschisis per 10,000 live births to women less than 20 years of age and only one baby with gastroschisis per 10,000 live births to women 40 years of age or older.



## Conclusions

While the causes of many birth defects are unknown, the data provided by the Texas Birth Defects Registry inform public health professionals regarding significant differences in the prevalence of birth defects across various characteristics such as the sex of the infant, mother's age, and mother's race/ethnicity. This provides information on which groups experience the highest rates of birth defects as well as clues as to what their causes may be.

## References

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