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Population-based birth defects data in the United States, 2012–2016: A focus on abdominal wall defects

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DATA AVAILABILITY STATEMENT
Research data are not shared.

SUPPORTING INFORMATION
Additional supporting information may be found online in the Supporting Information section at the end of this article.

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Abstract

Background/Objectives: In this report, the National Birth Defects Prevention Network (NBDPN) examines and compares gastroschisis and omphalocele for a recent 5-year birth cohort using data from 30 population-based birth defect surveillance programs in the United States.

Methods: As a special call for data for the 2019 NBDPN Annual Report, state programs reported expanded data on gastroschisis and omphalocele for birth years 2012–2016. We estimated the overall prevalence (per 10,000 live births) and 95% confidence intervals (CI) for each defect as well as by maternal race/ethnicity, maternal age, infant sex, and case ascertainment methodology utilized by the program (active vs. passive). We also compared distribution of cases by maternal and infant factors and presence/absence of other birth defects.

Results: The overall prevalence estimates (per 10,000 live births) were 4.3 (95% CI:4.1–4.4) for gastroschisis and 2.1 (95% CI: 2.0–2.2) for omphalocele. Gastroschisis was more frequent among young mothers (<25 years) and omphalocele more common among older mothers (>40 years). Mothers of infants with gastroschisis were more likely to be underweight/normal weight prior to pregnancy and mothers of infants with omphalocele more likely to be overweight/obese. Omphalocele was twice as likely as gastroschisis to co-occur with other birth defects.

Conclusions: This report highlights important differences between gastroschisis and omphalocele. These differences indicate the importance of distinguishing between these defects in epidemiologic assessments. The report also provides additional data on co-occurrence of gastroschisis and omphalocele with other birth defects. This information can provide a basis for future research to better understand these defects.

Keywords

abdominal wall defects; birth defects; gastroschisis; NBDPN; omphalocele

1 | INTRODUCTION

Since 2000, the National Birth Defects Prevention Network (NBDPN) has published state-specific data annually for selected major structural birth defects as part of the Congenital Malformations Surveillance Report. The 2019 NBDPN Congenital Malformations Surveillance Report is comprised of three parts: (a) state-specific birth defect counts and prevalence estimates from 42 population-based birth defects surveillance programs in the United States, reporting up to 47 major birth defects enumerated in Table 1; (b) a program

directory providing information on participating state birth defect surveillance programs, including each program's case definition, surveillance methodology, sources of birth defect information, data collection procedures, and program contacts, available on pages S1-S180; and (c) a data brief highlighting data pooled from 30 surveillance programs on two abdominal wall defects, gastroschisis, and omphalocele. Given the clinical and public health importance of gastroschisis and omphalocele, these defects were chosen to provide a more detailed description of the descriptive epidemiology and the differentiating characteristics between the two defects.

2 | STATE-SPECIFIC DATA COLLECTION AND PRESENTATION OF 47 MAJOR BIRTH DEFECTS

2.1 | Data collection

A call for data, including a data dictionary with a list of the birth defects and variables to be collected, was issued in March 2019 to population-based birth defect surveillance programs in the United States. Participating state surveillance programs submitted case counts for the major structural birth defects included in Table 1 and live births occurring from January 1, 2012 through December 31, 2016. Data collected from each program included major birth defects by year of birth, maternal race/ethnicity and age at delivery, and infant sex. Maternal race/ethnicity was stratified by six US Census groups: non-Hispanic (NH) white, NH black, Hispanic, NH Asian/Pacific Islander, NH American Indian/Alaska Native, and other/unknown. Maternal age at delivery was stratified into seven categories: <20, 20–24, 25–29, 30–34, 35–39, ≥40 years and unknown. Infant sex was stratified by male, female, and unknown.

State birth defect surveillance programs were asked to submit current directory profiles with a description of their program, including any updates since the previous year. State surveillance programs submitted data to the Centers for Disease Control and Prevention (CDC) either as SAS datasets or in Microsoft Excel. Data and directory profiles were submitted to CDC for cleaning and processing. CDC worked with state programs to review and validate all submitted data and directory information. State programs approved final data tables and directory profiles prior to publication. Data analysis was performed using SAS Version 9.4 (SAS Institute, Cary, NC).

2.2 | State-specific data presentation for 47 NBDPN birth defects

State-specific data from 42 population-based birth defects surveillance programs for 2012 through 2016 are presented in two separate tables for each state program (supplement). The first table presents birth defect counts and prevalence (per 10,000 live births) by maternal racial/ethnic groups. The second table displays counts and prevalence for gastroschisis and trisomies 13, 18, and 21 by two maternal age categories (<35 years, ≥35 years) because these particular defects are known to be associated with maternal age. Maternal age was combined into two groups to ensure adequate cell counts. For all defects except congenital posterior urethral valves (CPUV), hypospadias, and Turner syndrome, prevalence was calculated as the count of cases within each stratum of a maternal or infant characteristic—regardless of pregnancy outcome (i.e., live birth, stillbirth, spontaneous/elective termination)

—divided by the total number of live births within the same stratum, and then multiplied by 10,000 (Mason, Kirby, Sever, & Langlois, 2005). For CPUV and hypospadias, the denominator was restricted to total male live births; for Turner syndrome, the denominator was restricted to total female live births.

Because of the variability between state surveillance programs, each program's tables are accompanied by footnotes describing the program's coding system, notes on defect inclusion/exclusion, data sources, birth outcomes, and case ascertainment methodology. Additional detail regarding each program's data collection methodology and potential sources of variation between programs can be found in the supplemental program directory.

2.3 | Special data brief background

The NBDPN 2019 Congenital Malformations Surveillance Report includes state-level data on gastroschisis and omphalocele from 30 state programs, providing a unique opportunity to compare the descriptive epidemiology of the two most common types of abdominal wall defects. Abdominal wall defects are a type of birth defect in which the stomach, the intestines, or other organs protrude through an opening in the abdomen. Collectively, abdominal wall defects occur in an estimated 7.4 per 10,000 live births (Benjamin & Wilson, 2014); however, birth prevalence varies by defect type. Despite differences in the presentation of these defects, gastroschisis and omphalocele shared an International Classification of Diseases, 9th Revision, Clinical Modification (ICD-9-CM) code until October 2009, which created challenges in interpreting data on these defects.

2.4 | Gastroschisis

Gastroschisis is a full-thickness abdominal wall defect in which the intestines, and occasionally other organs, protrude outside the body through an opening usually to the right of the umbilicus without a protective membranous covering (Centers for Disease Control and Prevention, 2017). The estimated prevalence of gastroschisis in the United States ranges from 2.6 per 10,000 live births (Vu, Nobuhara, Laurent, & Shaw, 2008) to 5.1 per 10,000 live births (Vo & Langlois, 2015). Gastroschisis is one of the few major birth defects with a documented increasing birth prevalence in both resource-limited and resource-rich settings. In fact, the prevalence of gastroschisis has been increasing worldwide for decades (Bugge et al., 2017; Castilla, Mastroiacovo, & Orioli, 2008; Kazaura et al., 2004; Loane, Dolk, Bradbury, & Group, 2007; Whitehall, Kandasamy, Stalewski, & Gill, 2010) as well as in the United States (B. G. Benjamin, Ethen, Van Hook, Myers, & Canfield, 2010; Chabra, Gleason, Seidel, & Williams, 2011; Collins et al., 2007; Hougland, Hanna, Meyers, & Null, 2005; Jones et al., 2016; Kirby et al., 2013; Laughon et al., 2003; Salemi et al., 2009; Salihu, Pierre-Louis, Druschel, & Kirby, 2003; Short et al., 2019; St Louis et al., 2017; Vo & Langlois, 2015; Vu et al., 2008).

Reasons for the increasing prevalence in gastroschisis are unknown; however, several risk factors are associated with this defect, especially young maternal age. An increasing prevalence of gastroschisis among younger mothers, especially under 24 years of age, has consistently been documented, and young maternal age has been recognized as one of the strongest risk factors for gastroschisis (Anderson et al., 2018; B. G. Benjamin et al., 2010;

Bugge et al., 2017; Chabra et al., 2011; Hougland et al., 2005; Jones et al., 2016; Kirby, 2017; Kirby et al., 2013; Laughon et al., 2003; St Louis et al., 2017; Williams, Kucik, Alverson, Olney, & Correa, 2005). The prevalence of gastroschisis also differs by maternal race/ethnicity, with lower prevalence among infants born to NH black mothers compared to Hispanic mothers and NH white mothers (B. G. Benjamin et al., 2010; Chabra et al., 2011; Jones et al., 2016; Kirby, 2017; Salemi et al., 2009; Williams et al., 2005).

Other maternal and infant characteristics have been suggested to be associated with gastroschisis. Specifically, mothers of infants born with gastroschisis are more likely to be underweight (i.e., BMI <18 kg/m^2; Kirby et al., 2013), nulliparous (B. G. Benjamin et al., 2010; Vu et al., 2008), and low gravida (Hougland et al., 2005). Gastroschisis prevalence has not been shown to differ significantly by infant sex (Kirby et al., 2013), however, infants with gastroschisis are more likely to be preterm and low/very low birth weight (Anderson et al., 2018; Boutros, Regier, Skarsgard, & Canadian Pediatric Surgery Network, 2009; Bugge et al., 2017). Unlike other abdominal wall defects, gastroschisis usually occurs without other birth defects or chromosomal anomalies (Feldkamp, Botto, Byrne, Krikov, & Carey, 2016; Stoll, Alembik, Dott, & Roth, 2008). Gastroschisis has a low rate of fetal deaths and terminations compared to other major birth defects, the prognosis continues to improve with advances in prenatal monitoring, and the one-year survival rate is currently 94% (Adair et al., 1996; Akhtar, Skarsgard, & Canadian Pediatric Surgery Network, 2012; Brantberg, Blaas, Salvesen, Haugen, & Eik-Nes, 2004; Kuleva, Salomon, Benoist, Ville, & Dumez, 2012; Perry et al., 2017; South, Stutey, & Meinzen-Derr, 2013; Tennant, Pearce, Bythell, & Rankin, 2010).

2.5 | Omphalocele

Omphalocele is an abdominal wall defect wherein the intestines, liver, or other organs are most often covered by a membranous sac outside of the abdomen through the umbilicus (Centers for Disease Control and Prevention, 2017). The presence or absence of the membranous sac aids in differentiating gastroschisis and omphalocele, however, the membrane can rupture in the uterus causing the differential diagnosis to depend on the cord insertion site. The cord insertion site is located in the umbilical sac in omphalocele and paraumbilical in gastroschisis.

The estimated prevalence of omphalocele ranges from 1.0 per 10,000 live births (St Louis et al., 2017) to 3.8 per 10,000 live births (Springett et al., 2014). In contrast to gastroschisis, prevalence studies of omphalocele have reported either stable rates (Allman et al., 2016; Bugge et al., 2017; Marshall et al., 2015) or modest increases over time (St Louis et al., 2017). Other studies have reported inconsistent results regarding maternal age (Agopian, Marengo, & Mitchell, 2009; Goldkrand, Causey, & Hull, 2004; Marshall et al., 2015; St Louis et al., 2017). One study examining prevalence by maternal age found a small increase in prevalence for omphalocele among mothers 29 years or younger, however, a decrease was observed for those 30 years or older (St Louis et al., 2017). Additional studies examining the prevalence for omphalocele by maternal age reported a higher prevalence among older women, 35 years or older (Marshall et al., 2015) and a higher prevalence of nonsyndromic omphalocele among women aged 20–29 years and 40 years and older (Agopian et al., 2009).

Goldkrand et al. (2004) reported no trend in maternal age among mothers whose infants had omphalocele.

Maternal risk factors associated with omphalocele include multiparity (Duong et al., 2012) and prepregnancy overweight or obese status (Waller et al., 2007). In addition, omphalocele prevalence rates have been reported to be higher for infants born to Hispanic mothers and lower for NH black mothers. Omphalocele appears to be associated with lower birth weight and small for gestational age (Agopian et al., 2009; Kirby, 2017; Kirby et al., 2013; Marshall et al., 2015). Unlike gastroschisis, the prevalence of omphalocele has been observed to differ by infant sex, with a higher prevalence in males compared with females (Agopian et al., 2009; Goldkrand et al., 2004; Kirby, 2017; Kirby et al., 2013; Marshall et al., 2015). Marshall et al. (2015) reported that cases of omphalocele occur in isolation only 22% of the time. Similarly, Benjamin and Wilson (2014) reported that roughly 80% of cases of omphalocele had an associated anomaly. Defects that occur with omphalocele primarily include those of the cardiovascular system, musculoskeletal system, gastrointestinal system, urogenital system, and central nervous system as well as chromosomal defects (Benjamin & Wilson, 2014; Bugge et al., 2017; Springett et al., 2014; Stoll et al., 2008). Approximately one-sixth of omphalocele cases also have chromosomal anomalies, which are more common in infants born to women aged 35 years and older and among male infants (Marshall et al., 2015). In conjunction with these co-occurring defects, omphalocele has been associated with a higher rate of terminations, a higher rate of fetal deaths, and lower 1-week and 1-year survival than gastroschisis (Akhtar et al., 2012; Brantberg et al., 2004; Springett et al., 2014; Tennant et al., 2010).

Despite studies on the prevalence of gastroschisis and omphalocele and the increasing prevalence of gastroschisis, the etiology of these defects remains poorly understood. Moreover, given the prevalence and clinical impact of these defects, further research is needed to gain insight into the etiology and differences reported in the birth prevalence by various maternal and infant characteristics for these two defects. While a few large-scale population-based studies have investigated the prevalence of these defects by maternal race/ethnicity, maternal age, and infant sex (Anderson et al., 2018; Gong et al., 2016; Jones et al., 2016; Kirby et al., 2013; Loane et al., 2007; Marshall et al., 2015; Salihu et al., 2003; St Louis et al., 2017; Vu et al., 2008), many of these studies focused on gastroschisis or omphalocele alone and did not examine the differentiating characteristics between the two defects. Therefore, we examined the birth prevalence and descriptive epidemiology of gastroschisis and omphalocele using population-based data (2012–2016) submitted to the NBDPN.

2.6 | Special data brief methods

As a special call for data for the 2019 NBDPN Annual Report, state programs were invited to report expanded data on gastroschisis and omphalocele. The specific codes for gastroschisis are 756.73 (ICD-9-CM), Q79.3 (ICD-10-CM), and 756.71 (CDC/BPA). Specific codes for omphalocele include 756.72 (ICD-9-CM), Q79.2 (ICD-10-CM), and 756.70 (CDC/BPA), listed in Table 1. Data requested included case-level information by year of birth, maternal race/ethnicity, maternal age at delivery, infant sex, pregnancy

outcome, birth weight, maternal prepregnancy body mass index (BMI), gestational age at delivery, plurality, and co-occurring birth defects. Programs were also asked to submit information on any co-occurring birth defects they collected for these cases, including major and minor defects; however, this report presents co-occurring birth defects grouped by organ system, as specified by ICD-9-CM codes (740–759—congenital anomalies) and ICD-10-CM codes (Q00-Q99—congenital malformations, deformations, and chromosomal abnormalities), and excludes any birth defects codes outside these ranges. Cases were limited to those with a gestational age ≥20 weeks to ensure data quality and uniformity and BMI information was limited to live births due to data availability. Cases with diagnosis codes for both gastroschisis and omphalocele were excluded from the analyses when the correct diagnosis could not be confirmed. The data submission process and review followed a similar approach to the main annual report data submission. CDC obtained a non-research determination for the project.

Analyses were performed using SAS 9.4 (Cary, NC). The 95% confidence intervals were calculated using exact Poisson methodology for prevalence estimates and exact binomial for percentages (Daly, 1992). Prevalence estimates are reported as the number of cases per 10,000 live births. Analyses of co-occurring birth defects were limited to programs with active case ascertainment methodology to increase quality of data.

3 | RESULTS

A total of 30 state and territorial birth defects surveillance programs, covering a total of 12,563,163 births from 2012 to 2016, provided expanded data for gastroschisis and omphalocele. Table 2 presents count and prevalence for each defect by case ascertainment methodology, maternal race/ethnicity, maternal age at delivery, and infant sex. Pooling data from all 30 participating surveillance programs, the overall prevalence estimates (per 10,000 live births) were 4.3 for gastroschisis (5,349 cases) and 2.1 for omphalocele (2,601 cases).

Differences in prevalence by case ascertainment methodology were observed for both defects but were more pronounced for gastroschisis compared to omphalocele. Both defects showed a higher prevalence (per 10,000 live births) among active case ascertainment programs compared to passive programs (4.7 vs. 3.9 for gastroschisis and 2.2 vs. 1.9 for omphalocele, respectively).

Variations in prevalence were observed among maternal racial/ethnic groups (Table 2), with infants born to NH American Indian or Alaska Native mothers having the highest prevalence (per 10,000 live births) of gastroschisis (7.2) and infants born to NH black mothers having the highest prevalence of omphalocele (3.2).

Prevalence estimates (per 10,000 live births) for gastroschisis were highest among infants born to younger mothers, with infants born to mothers age 20 years and younger having the highest prevalence (15.5) followed by infants born to mothers ages 20–24 years (8.5). Prevalence for gastroschisis decreased by ascending maternal age group, with infants born to mothers age 35 years and above having the lowest prevalence (0.7). For omphalocele the

highest prevalence (per 10,000 live births) was observed among infants born to mothers age 40 years and above (4.1).

Prevalence estimates (per 10,000 live births) did not vary by infant sex for gastroschisis. However, omphalocele was more prevalent among males compared to females (2.2 vs. 1.9, respectively; Table 2).

Table 3 shows a comparison of case distribution by case ascertainment methodology, maternal race/ethnicity, maternal age, maternal prepregnancy BMI, infant sex, gestational age at delivery, infant birth weight, plurality, and birth outcome for the two abdominal wall defects. Both active and passive surveillance programs reported a similar proportion of cases of gastroschisis, but passive programs reported a higher proportion of omphalocele cases compared to active programs (51.5 vs. 48.5%, respectively). When comparing case distribution among maternal racial/ethnic groups, gastroschisis was more common in infants born to NH white (53.4 vs. 47.8%) and Hispanic (28.5 vs. 20.9%) mothers, while omphalocele was more common in infants born to NH black mothers (24.1 vs. 11.8%). Some of the most divergent case proportions were seen among maternal age groups, with a higher proportion of cases concentrated in the 24 years and below age groups for gastroschisis compared to omphalocele, while omphalocele cases were more common in the 25 years and above age groups. Gastroschisis was more common in infants of mothers in the underweight and normal BMI categories in comparison to omphalocele, while omphalocele was more common in infants of mothers in the overweight and obese categories. Both defects showed a higher proportion of cases among males compared to females. Cases of omphalocele were more common in early preterm deliveries (20–33 weeks gestation) and gastroschisis cases were more frequent in late preterm and full term deliveries (Table 3). Both defects had relatively high proportions of low birth weight, with omphalocele having a larger proportion of very low birth weight compared to gastroschisis (14.2 vs. 5.4%, respectively) and gastroschisis having a larger proportion of low birth weight (52.1 vs. 24.2%, respectively). Omphalocele cases had a higher proportion of both plural births (twins or multiples; 6.0% vs. 2.2%, respectively) and non-live births (19.1 vs. 5.6%) compared to gastroschisis.

Table 4 shows co-occurring birth defects by organ system for the abdominal wall defects for 15 surveillance programs with active case ascertainment. Co-occurring birth defects were observed twice as often among children with omphalocele compared to those with gastroschisis (71.8 vs. 33.6%, respectively). Cardiovascular defects represented the most common co-occurring group of birth defects for both defects (11.9% of gastroschisis cases and 44.9% of omphalocele cases). A moderate proportion of omphalocele cases had co-occurring central nervous system (15.2%), ear/face/neck (14.1%), renal (16.8%), musculoskeletal (16.5%), limbs (16.5%), other musculoskeletal (16.0%), and chromosomal (21.7%) defects.

4 | DISCUSSION

The overall prevalence estimates presented in this data brief for both gastroschisis and omphalocele are consistent with those previously reported in the literature (Mai et al., 2015;

Springett et al., 2014; St Louis et al., 2017; Vo & Langlois, 2015; Vu et al., 2008). Variations in prevalence by maternal race/ethnicity, maternal age at delivery, maternal prepregnancy BMI, infant sex, and infant birth weight also largely reflect previously reported estimates (Anderson et al., 2018; Kirby, 2017; Kirby et al., 2013). Our results support the findings of Marshall et al. (2015), who reported an increase in omphalocele in infants of older mothers. Finally, our comparison of co-occurring birth defects by organ system supports previous research showing a higher prevalence of co-occurring defects for omphalocele cases compared to gastroschisis, with cardiovascular defects and chromosomal defects showing a particularly large difference between the two abdominal wall defects (Feldkamp et al., 2016; Stoll et al., 2008).

Notably, these birth defects previously shared an ICD-9-CM diagnosis code. Our results, and the work of others, highlight the important differences between gastroschisis and omphalocele. These differences indicate the importance of distinguishing between these two defects in epidemiologic assessments. In fact, there are often varying and sometimes opposite risk factors for each defect, particularly maternal age and prepregnancy BMI. Omphalocele prevalence estimates are roughly one-half those of gastroschisis, but the omphalocele prevalence may be underestimated due to the higher proportion of fetal deaths and terminations of omphalocele cases compared to gastroschisis (Akhtar et al., 2012; Brantberg et al., 2004; Kuleva et al., 2012; Perry et al., 2017; Springett et al., 2014). In addition, infants with omphalocele are more likely to be early preterm deliveries and have very low birth weights compared to infants with gastroschisis (Anderson et al., 2018; Bugge et al., 2017; Feldkamp et al., 2016; Marshall et al., 2015). Despite the increase in prevalence of gastroschisis that has been well-documented in the literature, it is clear that omphalocele also presents a large burden for families and the healthcare system.

This report must be considered in the light of certain limitations. To increase the reliability of our data, we restricted our case definition to all birth outcomes with 20 weeks or more completed gestation. As a result, we know that we will have missed earlier fetal deaths and terminations. Given that omphalocele cases are more likely to occur in conjunction with more severe defects such as chromosomal defects (Akhtar et al., 2012; Brantberg et al., 2004; Springett et al., 2014), we anticipate that this may affect our estimates for omphalocele more than our estimates for gastroschisis. We also eliminated any cases with diagnosis codes for both defects, as we could not determine which defect was correct for these cases. These exclusions may also affect our estimates.

Variations in case ascertainment methodology and anomalies collected by the different state/territorial programs could contribute to variations in the quantity and range of co-occurring defects reported. We attempted to limit this by restricting our analyses of co-occurring defects to only include data from surveillance programs with active case-finding methodologies. The birth defects we evaluated for co-occurrence were also limited to the congenital anomalies range of ICD-9-CM codes and the congenital malformations, deformations, and chromosomal abnormalities range of ICD-10-CM. As a result, diagnoses that potentially may be considered birth defects outside these ranges are excluded. However, by restricting the code range the co-occurring data presented are more consistent across programs.

5 | CONCLUSION

The 2019 NBDPN Congenital Malformations Surveillance Report includes data from 42 state and territorial population-based birth defect surveillance programs, reporting up to 47 major structural birth defects. The report continues to provide important and updated information on the occurrence of these birth defects in the United States. This year's data brief on abdominal wall defects, using pooled surveillance data from 30 states, provides population-based prevalence estimates along with data on co-occurrence with defects by organ system for a cohort of about 12.5 million births. The updated prevalence estimates for gastroschisis and omphalocele along with comparison of case distribution among maternal and infant factors can provide a basis for future epidemiologic studies to better understand these birth defects.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

REFERENCES

Adair C, Rosnes J, Frye A, Burrus D, Nelson L, & Veille J (1996). The role of antepartum surveillance in the management of gastroschisis. International Journal of Gynaecology and Obstetrics, 52(2), 141–144. [PubMed: 8855092]

Agopian A, Marengo L, & Mitchell LE (2009). Descriptive epidemiology of nonsyndromic omphalocele in Texas, 1999–2004. American Journal of Medical Genetics. Part A, 149A(10), 2129–2133. 10.1002/ajmg.a.33000 [PubMed: 19760656]

Akhtar J, Skarsgard ED, & Canadian Pediatric Surgery Network. (2012). Associated malformations and the "hidden mortality" of gastroschisis. Journal of Pediatric Surgery, 47(5), 911–916. [PubMed: 22595571]

Allman R, Sousa J, Walker MW, Laughon MM, Spitzer AR, & Clark RH (2016). The epidemiology, prevalence and hospital outcomes of infants with gastroschisis. Journal of Perinatology, 36(10), 901–905. 10.1038/jp.2016.99 [PubMed: 27388940]

Anderson JE, Galganski LA, Cheng Y, Stark RA, Saadai P, Stephenson JT, & Hirose S (2018). Epidemiology of gastroschisis: A population-based study in California from 1995 to 2012. Journal of Pediatric Surgery, 53(12), 2399–2403. 10.1016/j.jpedsurg.2018.08.035 [PubMed: 30266482]

Benjamin B, & Wilson GN (2014). Anomalies associated with gastroschisis and omphalocele: Analysis of 2825 cases from the Texas birth defects registry. Journal of Pediatric Surgery, 49(4), 514–519. 10.1016/j.jpedsurg.2013.11.052 [PubMed: 24726103]

Benjamin BG, Ethen MK, Van Hook CL, Myers CA, & Canfield MA (2010). Gastroschisis prevalence in Texas 1999–2003. Birth Defects Research. Part A, Clinical and Molecular Teratology, 88(3), 178–185. 10.1002/bdra.20642

Boutros J, Regier M, Skarsgard ED, & Canadian Pediatric Surgery Network. (2009). Is timing everything? The influence of gestational age, birth weight, route, and intent of delivery on outcome in gastroschisis. Journal of Pediatric Surgery, 44(5), 912–917. 10.1016/j.jpedsurg.2009.01.026 [PubMed: 19433169]

Brantberg A, Blaas H, Salvesen K, Haugen S, & Eik-Nes S (2004). Surveillance and outcome of fetuses with gastroschisis. Ultrasound in Obstetrics & Gynecology, 23(1), 4–13. [PubMed: 14970991]

Bugge M, Drachmann G, Kern P, Budtz-Jorgensen E, Eiberg H, Olsen B, … Nielsen IM (2017). Abdominal wall defects in Greenland 1989–2015. Birth Defects Research, 109(11), 836–842. 10.1002/bdr2.1025 [PubMed: 28464537]

Castilla EE, Mastroiacovo P, & Orioli IM (2008). Gastroschisis: International epidemiology and public health perspectives. American Journal of Medical Genetics. Part C, Seminars in Medical Genetics, 148C(3), 162–179. 10.1002/ajmg.c.30181

Centers for Disease Control and Prevention. (11 21, 2017). Learn about specific birth defects. Retrieved from https://www.cdc.gov/ncbddd/birthdefects/types.html.

Chabra S, Gleason CA, Seidel K, & Williams MA (2011). Rising prevalence of gastroschisis in Washington state. Journal of Toxicology and Environmental Health. Part A, 74(5), 336–345. 10.1080/15287394.2011.534424 [PubMed: 21240733]

Collins SR, Griffin MR, Arbogast PG, Walsh WF, Rush MR, Carter BS, … Cooper WO (2007). The rising prevalence of gastroschisis and omphalocele in Tennessee. Journal of Pediatric Surgery, 42(7), 1221–1224. 10.1016/j.jpedsurg.2007.02.012 [PubMed: 17618884]

Daly L (1992). Simple SAS macros for the calculation of exact binomial and Poisson confidence limits. Computers in Biology and Medicine, 22(5), 351–361. [PubMed: 1424580]

Duong HT, Hoyt AT, Carmichael SL, Gilboa SM, Canfield MA, Case A, … National Birth Defects Prevention Study. (2012). Is maternal parity an independent risk factor for birth defects? Birth Defects Research. Part A, Clinical and Molecular Teratology, 94(4), 230–236. 10.1002/bdra.22889 [PubMed: 22371332]

Feldkamp ML, Botto LD, Byrne JLB, Krikov S, & Carey JC (2016). Clinical presentation and survival in a population-based cohort of infants with gastroschisis in Utah, 1997–2011. American Journal of Medical Genetics. Part A, 170A (2), 306–315. 10.1002/ajmg.a.37437 [PubMed: 26473400]

Goldkrand JW, Causey TN, & Hull EE (2004). The changing face of gastroschisis and omphalocele in Southeast Georgia. The Journal of Maternal-Fetal & Neonatal Medicine, 15(5), 331–335. 10.1080/14767050410001699875 [PubMed: 15280125]

Gong TT, Wu QJ, Chen YL, Jiang CZ, Li J, Li LL, … Huang YH (2016). Evaluating the time trends in prevalence of exomphalos in 14 cities of Liaoning province, 2006 to 2015. Scientific Reports, 6, 32901 10.1038/srep32901 [PubMed: 27604427]

Hougland KT, Hanna AM, Meyers R, & Null D (2005). Increasing prevalence of gastroschisis in Utah. Journal of Pediatric Surgery, 40(3), 535–540. 10.1016/j.jpedsurg.2004.11.026 [PubMed: 15793731]

Jones AM, Isenburg J, Salemi JL, Arnold KE, Mai CT, Aggarwal D, … Honein MA (2016). Increasing prevalence of Gastroschisis—14 States, 1995–2012. MMWR. Morbidity and Mortality Weekly Report, 65(2), 23–26. 10.15585/mmwr.mm6502a2 [PubMed: 26796490]

Kazaura MR, Lie RT, Irgens LM, Didriksen A, Kapstad M, Egenaes J, & Bjerkedal T (2004). Increasing risk of gastroschisis in Norway: An age-period-cohort analysis. American Journal of Epidemiology, 159(4), 358–363. 10.1093/aje/kwh051 [PubMed: 14769639]

Kirby RS (2017). The prevalence of selected major birth defects in the United States. Seminars in Perinatology, 41(6), 338–344. 10.1053/j.semperi.2017.07.004 [PubMed: 29037343]

Kirby RS, Marshall J, Tanner JP, Salemi JL, Feldkamp ML, Marengo L, … National Birth Defects Prevention Network. (2013). Prevalence and correlates of gastroschisis in 15 states, 1995 to 2005. Obstetrics and Gynecology, 122(2 Pt 1), 275–281. 10.1097/AOG.0b013e31829cbbb4 [PubMed: 23969795]

Kuleva M, Salomon L, Benoist G, Ville Y, & Dumez Y (2012). The value of daily fetal heart rate home monitoring in addition to serial ultrasound examinations in pregnancies complicated by fetal gastroschisis. Prenatal Diagnosis, 32(8), 789–796. [PubMed: 22653694]

Laughon M, Meyer R, Bose C, Wall A, Otero E, Heerens A, & Clark R (2003). Rising birth prevalence of gastroschisis. Journal of Perinatology, 23(4), 291–293. 10.1038/sj.jp.7210896 [PubMed: 12774135]

Loane M, Dolk H, Bradbury I, & Group EW (2007). Increasing prevalence of gastroschisis in Europe 1980–2002: A phenomenon restricted to younger mothers? Paediatric and Perinatal Epidemiology, 21(4), 363–369. 10.1111/j.1365-3016.2007.00820.x [PubMed: 17564594]

Mai CT, Riehle-Colarusso T, O'Halloran A, Cragan JD, Olney RS, Lin A, … National Birth Defects Prevention Network (2012). Selected birth defects data from population-based birth defects surveillance programs in the United States, 2005–2009: Featuring critical congenital heart defects

targeted for pulse oximetry screening. Birth Defects Research. Part A: Clinical and Molecular Teratology, 94(12), 970–983.

Mai CT, Isenburg J, Langlois PH, Alverson CJ, Gilboa SM, Rickard R, … Kirby RS (2015). Population-based birth defects data in the United States, 2008 to 2012: Presentation of state-specific data and descriptive brief on variability of prevalence. Birth Defects Research. Part A, Clinical and Molecular Teratology, 103(11), 972–993.

Marshall J, Salemi JL, Tanner JP, Ramakrishnan R, Feldkamp ML, Marengo LK, … National Birth Defects Prevention Network. (2015). Prevalence, correlates, and outcomes of Omphalocele in the United States, 1995–2005. Obstetrics and Gynecology, 126(2), 284–293. 10.1097/AOG.0000000000000920 [PubMed: 26241416]

Mason CA, Kirby RS, Sever LE, & Langlois PH (2005). Prevalence is the preferred measure of frequency of birth defects. Birth Defects Research. Part A, Clinical and Molecular Teratology, 73(10), 690–692. 10.1002/bdra.20211

Perry H, Healy C, Wellesley D, Hall N, Drewett M, Burge D, & Howe D (2017). Intrauterine death rate in gastroschisis following the introduction of an antenatal surveillance program: Retrospective observational study. The Journal of Obstetrics and Gynaecology Research, 43(3), 492–497. [PubMed: 28165177]

Salemi JL, Pierre M, Tanner JP, Kornosky JL, Hauser KW, Kirby RS, & Carver JD (2009). Maternal nativity as a risk factor for gastroschisis: A population-based study. Birth Defects Research. Part A, Clinical and Molecular Teratology, 85(11), 890–896. 10.1002/bdra.20612

Salihu HM, Pierre-Louis BJ, Druschel CM, & Kirby RS (2003). Omphalocele and gastroschisis in the state of New York, 1992–1999. Birth Defects Research. Part A, Clinical and Molecular Teratology, 67(9), 630–636. 10.1002/bdra.10113

Short TD, Stallings EB, Isenburg J, O'Leary LA, Yazdy MM, Bohm MK, … Reefhuis J (2019). Gastroschisis trends and ecologic link to opioid prescription rates—United States, 2006–2015. MMWR. Morbidity and Mortality Weekly Report, 68(2), 31–36. 10.15585/mmwr.mm6802a2 [PubMed: 30653484]

South A, Stutey K, & Meinzen-Derr J (2013). Metaanalysis of the prevalence of intrauterine fetal death in gastroschisis. American Journal of Obstetrics & Gynecology, 209(2), 114.e111–114.e113. [PubMed: 23628262]

Springett A, Draper ES, Rankin J, Rounding C, Tucker D, Stoianova S, … Morris JK (2014). Birth prevalence and survival of exomphalos in England and Wales: 2005 to 2011. Birth Defects Research. Part A, Clinical and Molecular Teratology, 100(9), 721–725. 10.1002/bdra.23301

St Louis AM, Kim K, Browne ML, Liu G, Liberman RF, Nembhard WN, … National Birth Defects Prevention Network. (2017). Prevalence trends of selected major birth defects: A multi-state population-based retrospective study, United States, 1999 to 2007. Birth Defects Research, 109(18), 1442–1450. 10.1002/bdr2.1113 [PubMed: 28905502]

Stoll C, Alembik Y, Dott B, & Roth MP (2008). Omphalocele and gastroschisis and associated malformations. American Journal of Medical Genetics. Part A, 146A(10), 1280–1285. 10.1002/ajmg.a.32297 [PubMed: 18386803]

Tennant P, Pearce M, Bythell M, & Rankin J (2010). 20-year survival of children born with congenital anomalies: A population-based study. Lancet, 375(9715), 649–656. [PubMed: 20092884]

Vo LU, & Langlois PH (2015). Time trends in prevalence of gastroschisis in Texas, 1999 to 2011: Subgroup analyses by maternal and infant characteristics. Birth Defects Research. Part A, Clinical and Molecular Teratology, 103(11), 928–940. 10.1002/bdra.23438

Vu LT, Nobuhara KK, Laurent C, & Shaw GM (2008). Increasing prevalence of gastroschisis: Population-based study in California. The Journal of Pediatrics, 152(6), 807–811. 10.1016/j.jpeds.2007.11.037 [PubMed: 18492521]

Waller DK, Shaw GM, Rasmussen SA, Hobbs CA, Canfield MA, Siega-Riz AM, … National Birth Defects Prevention Study. (2007). Prepregnancy obesity as a risk factor for structural birth defects. Archives of Pediatrics & Adolescent Medicine, 161(8), 745–750. 10.1001/archpedi.161.8.745 [PubMed: 17679655]

Whitehall JS, Kandasamy Y, Stalewski H, & Gill A (2010). Perinatal demography of gastroschisis in North Queensland. Journal of Paediatrics and Child Health, 46(12), 749–753. 10.1111/j.1440-1754.2010.01833.x [PubMed: 21166913]

Williams LJ, Kucik JE, Alverson CJ, Olney RS, & Correa A (2005). Epidemiology of gastroschisis in metropolitan Atlanta, 1968 through 2000. Birth Defects Research. Part A, Clinical and Molecular Teratology, 73(3), 177–183. 10.1002/bdra.20114

TABLE 1

Birth defects for the National Birth Defects Prevention Network (NBDPN) annual report by disease classification codes

Birth defects	ICD-9-CM codes	ICD-10-CM codes	CDC/BPA codes
Central nervous system			
Anencephaly	740.0–740.1	Q00.0-Q00.1	740.00–740.10
Spina bifida without anencephaly	741.0, 741.9 w/o 740.0–740.1	Q05.0-Q05.9, Q07.01, Q07.03 w/o Q00.0-Q00.1	741.00–741.99 w/o 740.00–740.10
Encephalocele	742.0	Q01.0-Q01.9	742.00–742.09
Holoprosencephaly	742.2	Q04.2	742.26
Eye			
Anophthalmia/microphthalmia	743.0, 743.1	Q11.0-Q11.2	743.00–743.10
Congenital cataract	743.30–743.34	Q12.0	743.32
Ear			
Anotia/microtia	744.01, 744.23	Q16.0, Q17.2	744.01, 744.21
Cardiovascular			
Common truncus (truncus arteriosus or TA)	745.0	Q20.0	745.00 (excluding 745.01)
Transposition of the great arteries (TGA)	745.10, 745.12, 745.19	Q20.3, Q20.5	745.10–745.12, 745.18–745.19
Dextro-transposition of great arteries (d-TGA)[a]	745.10	Q20.3	745.10, 745.11, 745.18, 745.19
Tetralogy of Fallot (TOF)	745.2	Q21.3	745.20–745.21, 747.31
Ventricular septal defect	745.4	Q21.0	745.40–745.49 (excluding 745.487, 745.498)
Atrial septal defect	745.5	Q21.1	745.51–745.59
Atrioventricular septal defect (endocardial cushion defect)	745.60, 745.61, 745.69	Q21.2	745.60–745.69, 745.487
Pulmonary valve atresia and stenosis	746.01, 746.02	Q22.0, Q22.1	746.00, 746.01
Pulmonary valve atresia[a]	746.01	Q22.0	746.00
Tricuspid valve atresia and stenosis	746.1	Q22.4	746.100, 746.106 (excluding 746.105)
Tricuspid valve atresia[a]	746.1	Q22.4	746.100
Ebstein anomaly	746.2	Q22.5	746.20
Aortic valve stenosis	746.3	Q23.0	746.30
Hypoplastic left heart syndrome	746.7	Q23.4	746.70
Coarctation of aorta	747.10	Q25.1	747.10–747.19
Total anomalous pulmonary venous connection (TAPVC)	747.41	Q26.2	747.42
Single ventricle	745.3	Q20.4	745.3

Birth defects	ICD-9-CM codes	ICD-10-CM codes	CDC/BPA codes
Interrupted aortic arch (IAA)	747.11	Q25.2, Q25.4	747.215–747.217, 747.285
Double outlet right ventricle (DORV)	745.11	Q20.1	745.13–745.15
Orofacial			
Cleft palate alone (without cleft lip)	749.0	Q35.1-Q35.9	749.00–749.09
Cleft lip alone (without cleft palate)	749.1	Q36.0-Q36.9	749.10–749.19
Cleft lip with cleft palate	749.20–749.25	Q37.0-Q37.9	749.20–749.29
Choanal atresia	748.0	Q30.0	748.00
Gastrointestinal			
Esophageal atresia/tracheoesophageal fistula	750.3	Q39.0-Q39.4	750.30–750.35
Rectal and large intestinal atresia/stenosis	751.2	Q42.0-Q42.9	751.20–751.24
Biliary atresia	751.61	Q44.2-Q44.3	751.65
Small intestinal atresia/stenosis	751.1	Q41.0-Q41.9	751.10–751.19
Genitourinary			
Renal agenesis/hypoplasia	753.0	Q60.0-Q60.6	753.00–753.01
Bladder exstrophy	753.5	Q64.10, Q64.19	753.50
Hypospadias	752.61	Q54.0-Q54.9 (excluding Q54.4)	752.60–752.62 (excluding 752.61 and 752.621)
Congenital posterior urethral valves	753.6	Q64.2	753.60
Cloacal exstrophy	751.5	Q64.12	751.555
Musculoskeletal			
Gastroschisis[b]	756.73	Q79.3	756.71
Omphalocele[b]	756.72	Q79.2	756.70
Diaphragmatic hernia	756.6	Q79.0, Q79.1	756.610–756.617
Limb deficiencies (reduction defects)	755.2–755.4	Q71.0-Q71.9, Q72.0-Q72.9, Q73.0-Q73.8	755.20–755.49
Craniosynostosis	No specific code	Q75.0	756.00–756.03
Clubfoot	754.51, 754.70	Q66.0, Q66.89	754.50, 754.73 (excluding 754.735)
Chromosomal			
Trisomy 13	758.1	Q91.4-Q91.7	758.10–758.19
Trisomy 21 (Down syndrome)	758.0	Q90.0-Q90.9	758.00–758.09
Trisomy 18	758.2	Q91.0-Q91.3	758.20–758.29
Turner syndrome	758.6	Q96.0-Q96.9	758.60–758.69
Deletion 22 q11.2	758.32	Q93.81	758.37

Abbreviations: ICD-9-CM, International Classification of Diseases, 9th Revision, Clinical Modification; ICD-10-CM, International Classification of Diseases, 10th Revision, Clinical Modification; CDC/BPA, Centers for Disease Control and Prevention/British Pediatric Association Classification of Diseases.

[a] These subcategories of the broader reported anomalies (transposition of the great arteries, pulmonary valve atresia and stenosis, and tricuspid valve atresia and stenosis) are presented given the interest in monitoring critical congenital heart defects targeted for pulse oximetry screening (Mai et al., 2012).

[b] Gastroschisis and omphalocele are the focus of this year's data brief.

TABLE 2

Abdominal wall defect counts, prevalence, and 95% confidence intervals for 30 US population-based surveillance programs,[a] 2012–2016 (prevalence per 10,000 live births)

	Gastroschisis			Omphalocele		
Variable	**Count**	**Prevalence**	**95% CI[b]**	**Count**	**Prevalence**	**95% CI[b]**
Total[c]	5,349	4.3	4.1–4.4	2,601	2.1	2.0–2.2
Case ascertainment methodology[a]						
Active case finding	2,677	4.7	4.6–4.9	1,262	2.2	2.1–2.4
Passive case finding	2,672	3.9	3.7–4.0	1,339	1.9	1.8–2.0
Maternal race/ethnicity						
White, non-Hispanic	2,857	4.3	4.2–4.5	1,242	1.9	1.8–2.0
Black, non-Hispanic	632	3.2	3.0–3.5	627	3.2	2.9–3.4
Hispanic	1,524	5.0	4.8–5.3	543	1.8	1.6–1.9
Asian or Pacific Islander, non-Hispanic	105	1.6	1.3–1.9	80	1.2	1.0–1.5
American Indian or Alaska Native, non-Hispanic	71	7.2	5.6–9.1	18	1.8	1.1–2.9
Maternal age (*years*)						
<20	1,331	15.5	14.7–16.4	203	2.4	2.1–2.7
20–24	2,397	8.5	8.1–8.8	593	2.1	1.9–2.3
25–29	1,085	3.0	2.8–3.2	642	1.8	1.6–1.9
30–34	354	1.1	1.0–1.2	629	1.9	1.7–2.0
35–39	116	0.7	0.6–0.9	359	2.3	2.1–2.5
40+	24	0.7	0.4–1.0	152	4.1	3.5–4.9
Infant sex						
Male	2,715	4.2	4.1–4.4	1,405	2.2	2.1–2.3
Female	2,612	4.3	4.1–4.4	1,152	1.9	1.8–2.0

[a] Contributing programs (total live births = 12,563,163) by case-finding methodology: Active case-finding: Arizona, Arkansas (2012–2015), California, Delaware, Georgia (Metropolitan Atlanta Congenital Defects Program), Iowa, Louisiana, Massachusetts, Minnesota, North Carolina, Oklahoma, Puerto Rico, South Carolina, Texas, Utah. Passive case-finding: Florida, Illinois, Kansas, Kentucky, Maryland, Michigan, Missouri, Nebraska, New Jersey, New York, Ohio, Oregon, Rhode Island, Tennessee, Vermont.

[b] CI: Confidence interval calculated using exact Poisson methodology.

[c] Cases were included when gestational age was greater than or equal to 20 completed weeks gestation. For select states, ascertainment area differed from that reported in the supplemental tables. Counts across subgroups may not add to the total due to other/unknown categories (not shown).

TABLE 3

Abdominal wall defect counts, percentages, and 95% confidence intervals for 30 US population-based surveillance programs,[a] 2012–2016

	Gastroschisis (*n* = 5,349[b])			Omphalocele (*n* = 2,601[b])		
Variable	**Count**	**Percent**	**95% CI[c]**	**Count**	**Percent**	**95% CI[c]**
Case ascertainment methodology[a]						
Active case finding	2,677	50.0	48.7–51.4	1,262	48.5	46.6–50.5
Passive case finding	2,672	50.0	48.6–51.3	1,339	51.5	49.5–53.4
Maternal race/ethnicity						
White, non-Hispanic	2,857	53.4	52.1–54.8	1,242	47.8	45.8–49.7
Black, non-Hispanic	632	11.8	11.0–12.7	627	24.1	22.5–25.8
Hispanic	1,524	28.5	27.3–29.7	543	20.9	19.3–22.5
Asian or Pacific Islander, non-Hispanic	105	2.0	1.6–2.4	80	3.1	2.4–3.8
American Indian or Alaska Native, non-Hispanic	71	1.3	1.0–1.7	18	0.7	0.4–1.1
Maternal age (years)						
<20	1,331	24.9	23.7–26.1	203	7.8	6.8–8.9
20–24	2,397	44.8	43.5–46.2	593	22.8	21.2–24.5
25–29	1,085	20.3	19.2–21.4	642	24.7	23.0–26.4
30–34	354	6.6	6.0–7.3	629	24.2	22.5–25.9
35–39	116	2.2	1.8–2.6	359	13.8	12.5–15.2
40+	24	0.4	0.3–0.7	152	5.8	5.0–6.8
Maternal prepregnancy body mass index (BMI)[d]						
Underweight	332	6.9	6.2–7.7	65	2.9	2.3–3.7
Normal	2,733	56.9	55.5–58.3	840	37.8	35.7–39.8
Overweight	1,046	21.8	20.6–23.0	543	24.4	22.6–26.3
Obese	496	10.3	9.5–11.2	629	28.3	26.4–30.2
Infant sex						
Male	2,715	50.8	49.4–52.1	1,405	54.0	52.1–55.9
Female	2,612	48.8	47.5–50.2	1,152	44.3	42.4–46.2
Gestational age (*weeks*)						
20–23 weeks	57	1.1	0.8–1.4	127	4.9	4.1–5.8

	Gastroschisis (*n* = 5,349[b])			Omphalocele (*n* = 2,601[b])		
Variable	**Count**	**Percent**	**95% CI[c]**	**Count**	**Percent**	**95% CI[c]**
24–27 weeks	59	1.1	0.8–1.4	102	3.9	3.2–4.7
28–33 weeks	565	10.6	9.8–11.4	368	14.1	12.8–15.5
34–36 weeks	2,510	46.9	45.6–48.3	530	20.4	18.8–22.0
37+ weeks	2,096	39.2	37.9–40.5	1,428	54.9	53.0–56.8
Birth weight (*grams*)						
Very low birth weight (less than 1,500 g)	291	5.4	4.8–6.1	370	14.2	12.9–15.6
Low birth weight (1,500–2,499 g)	2,785	52.1	50.7–53.4	629	24.2	22.5–25.9
Normal birth weight (≥2,500 g)	2,170	40.6	39.2–41.9	1,426	54.8	52.9–56.8
Plurality						
Singleton	5,146	96.2	95.7–96.7	2,400	92.3	91.2–93.3
Twin or multiple	117	2.2	1.8–2.6	157	6.0	5.2–7.0
Pregnancy outcome[e]						
Live births	3,181	94.3	93.4–95.0	1,237	80.7	78.7–82.7
Non-live births	190	5.6	4.9–6.5	293	19.1	17.2–21.2

[a]Contributing programs by case-finding methodology: Active case-finding: Arizona, Arkansas (2012–2015), California, Delaware, Georgia (Metropolitan Atlanta Congenital Defects Program), Iowa, Louisiana, Massachusetts, Minnesota, North Carolina, Oklahoma, Puerto Rico, South Carolina, Texas, Utah. Passive case-finding: Florida, Illinois, Kansas, Kentucky, Maryland, Michigan, Missouri, Nebraska, New Jersey, New York, Ohio, Oregon, Rhode Island, Tennessee, Vermont.

[b]Cases were included when gestational age was greater than or equal to 20 completed weeks gestation. For select states, ascertainment area differed from that reported in the supplemental tables. Counts across subgroups may not add to the total due to other/unknown categories (not shown).

[c]CI: Confidence interval calculated using exact binomial methodology.

[d]Includes live birth cases from: Arizona (2014–2016), California, Delaware, Florida, Georgia (Metropolitan Atlanta Congenital Defects Program), Illinois, Iowa, Kansas, Kentucky, Louisiana, Maryland, Massachusetts, Michigan, Minnesota, Missouri, Nebraska, New Jersey (2015–2016), New York, North Carolina, Ohio, Oregon, Puerto Rico, Rhode Island, South Carolina, Tennessee, Texas, Utah, Vermont (Total gastroschisis cases = 4,802; Total omphalocele cases = 2,224).

[e]Includes programs who ascertain pregnancy outcomes including non-live births: Arizona, Arkansas (2012–2015), California, Delaware, Georgia (Metropolitan Atlanta Congenital Defects Program), Illinois, Iowa, Kansas, Kentucky, Massachusetts, Missouri, Nebraska, North Carolina, Oklahoma, Puerto Rico, Rhode Island, South Carolina, Texas, Utah, Vermont (Total gastroschisis cases = 3,374; Total omphalocele cases = 1,529).

TABLE 4

Co-occurring birth defects by organ system for abdominal wall defects from 15 US population-based active case-finding surveillance programs,[a] 2012–2016

	Gastroschisis (*n* = 2,677[b])			Omphalocele (*n* = 1,262[b])		
ICD[c] code groups	**Count**	**Percent**	**95% CI[d]**	**Count**	**Percent**	**95% CI[d]**
Congenital anomalies (740–759)/congenital malformations, deformations and chromosomal abnormalities (Q00-Q99)[e]	898	33.6	31.8–35.4	904	71.8	69.2–74.3
Central Nervous System (740–742, Q00–07)	105	3.9	3.2–4.7	191	15.2	13.2–17.3
Eye (743, Q10–15)	29	1.1	0.7–1.6	87	6.9	5.6–8.5
Ear, Face, Neck (744, Q16–18)	69	2.6	2.0–3.3	177	14.1	12.2–16.1
Cardiovascular (745–747, Q20–28)	318	11.9	10.7–13.2	565	44.9	42.1–47.7
Respiratory (748, Q30–34)	26	1.0	0.6–1.4	113	9.0	7.5–10.7
Orofacial Clefts (749, Q35–37)	21	0.8	0.5–1.2	76	6.0	4.8–7.5
Upper Gastrointestinal (750, Q38–40)	37	1.4	1.0–1.9	94	7.5	6.1–9.1
Lower Gastrointestinal (751, Q41–45)	277	10.4	9.2–11.6	169	13.4	11.6–15.4
Genital (752, Q50–56)	112	4.2	3.5–5.0	153	12.2	10.4–14.1
Renal (753, Q60–64)	167	6.2	5.4–7.2	211	16.8	14.7–18.9
Musculoskeletal (754, Q65–68)	129	4.8	4.0–5.7	208	16.5	14.5–18.7
Limbs (755, Q69–74)	69	2.6	2.0–3.3	208	16.5	14.5–18.7
Other Musculoskeletal (756, Q75–79)	59	2.2	1.7–2.8	201	16.0	14.0–18.1
Skin (757, Q80–84)	26	1.0	0.6–1.4	68	5.4	4.2–6.8
Chromosomal (758, Q90–99)	18	0.7	0.4–1.1	273	21.7	19.4–24.1
Trisomy 13	5	0.2	0.1–0.4	65	5.2	4.0–6.5
Trisomy 18	2	0.1	0.0–0.3	139	11.0	9.4–12.9
Trisomy 21 (Down Syndrome)	0	0.0	0.0–0.1	12	1.0	0.5–1.7
Turner Syndrome	0	0.0	0.0–0.1	25	2.0	1.3–2.9
Deletion 22q 11.2	1	0.0	0.0–0.2	3	0.2	0.0–0.7
Other (759, Q85–89)	26	1.0	0.6–1.4	158	12.5	10.8–14.5

[a] Active case-finding programs: Arizona, Arkansas (2012–2015), California, Delaware, Georgia (Metropolitan Atlanta Congenital Defects Program), Iowa, Louisiana, Massachusetts, Minnesota, North Carolina, Oklahoma, Puerto Rico, South Carolina, Texas, Utah.

[b] Cases were included when gestational age was greater than or equal to 20 completed weeks gestation. For select states, ascertainment area differed from that reported in the supplemental tables. Totals include unknown/other.

[c] International Classification of Diseases.

[d] CI: Confidence interval calculated using exact binomial methodology.

[e] Birth defects that fall outside the 740–759 range for ICD-9-CM and/or CDC/BPA or outside the Q00-Q99 range for ICD-10-CM were not examined. Cases are counted separately for each organ system (categories are not mutually exclusive).