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## The impact of the ICD-9-CM to ICD-10-CM transition on the prevalence of birth defects among infant hospitalizations in the United States

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### Abstract

**Background**—Many public health surveillance programs utilize hospital discharge data in their estimation of disease prevalence. These databases commonly use the International Classification of Diseases (ICD) coding scheme, which transitioned from the ICD-9 clinical modification (ICD-9-CM) to ICD-10-CM on October 1, 2015. This study examined this transition's impact on the prevalence of major birth defects among infant hospitalizations.

**Methods**—Using data from the Agency for Health Care Research and Quality-sponsored National Inpatient Sample, hospitalizations during the first year of life with a discharge date between January 1, 2012 and December 31, 2016 were used to estimate the monthly national hospital prevalence of 46 birth defects for the ICD-9-CM and ICD-10-CM timeframes separately. Survey-weighted Poisson regression was used to estimate 95% confidence intervals for each hospital prevalence. Interrupted time series framework and corresponding segmented regression

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#### CONFLICT OF INTEREST

The authors have no conflict of interest relevant to this article to disclose.

#### DISCLAIMER

The findings and conclusions in this report are those of the authors and do not necessarily represent the official position of the Centers for Disease Control and Prevention.

#### DATA AVAILABILITY STATEMENT

Data from the National Inpatient Sample can be purchased from the Agency for Healthcare Research and Quality at [https://www.hcup-us.ahrq.gov/tech\\_assist/centdist.jsp](https://www.hcup-us.ahrq.gov/tech_assist/centdist.jsp).

#### 6 | SUPPORTING INFORMATION

Additional Supporting Information may be found online in the supporting information tab for this article. This supplemental material includes a table presenting the overall rates of each birth defect per 10,000 hospitalizations during the ICD-9-CM versus ICD-10-CM timeframes, separately for birth hospitalizations and all hospitalizations during the first year of life; a table presenting the immediate impact of ICD-10-CM on calculated prevalence of each birth defect per 10,000 hospitalizations during the ICD-9-CM versus ICD-10-CM timeframes; and a series of figures, one for each defect under analysis, that depict the impact of the ICD-9-CM to ICD-10-CM code transition on monthly prevalence estimates.

#### SUPPORTING INFORMATION

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

was used to estimate the immediate change in monthly hospital prevalence following the ICD-9-CM to ICD-10-CM transition.

**Results**—Between 2012 and 2016, over 21 million inpatient hospitalizations occurred during the first year of life. Among the 46 defects studied, statistically significant decreases in the immediate hospital prevalence of five defects and significant increases in the immediate hospital prevalence of eight defects were observed after the ICD-10-CM transition.

**Conclusions**—Changes in prevalence were expected based on changes to ICD-10-CM. Observed changes for some conditions may result from variation in monthly hospital prevalence or initial unfamiliarity of coders with ICD-10-CM. These findings may help birth defects surveillance programs evaluate and interpret changes in their data related to the ICD-10-CM transition.

## Keywords

birth defects; classification; coding; hospital discharge data; International Classification of Diseases; interrupted time series; National Inpatient Sample

## 1 | INTRODUCTION

Researchers and public health programs often utilize population-based hospital discharge data to assess disease incidence and prevalence, health care utilization, costs and charges, and health outcomes (Andrews, 2015; Salemi, Salinas-Miranda, Wilson, & Salihu, 2015). Most of these databases use standardized coding rubrics to report and classify medical diagnoses as well as diagnostic and therapeutic procedures. The most commonly used coding scheme is the International Classification of Diseases (ICD), which has been overseen by the World Health Organization since 1948. For morbidity coding, the US health care system uses a clinical modification of the ICD developed by the National Center for Health Statistics, Centers for Disease Control and Prevention (CDC) (Centers for Disease Control and Prevention, 2018). Periodically, the ICD is revised, which results in the need to assess the comparability of data generated before and after a revision is implemented. From 1979 until September 30, 2015, most hospital discharge data in the United States were reported using the ICD-9 clinical modification (ICD-9-CM). On October 1, 2015, disease coding for all US healthcare institutions and practitioners covered by the Health Insurance Portability and Accountability Act (HIPAA) was transitioned to the 10th revision of the ICD, clinical modification (ICD-10-CM) (Centers for Disease Control and Prevention, 2015). The ICD-10-CM provided a number of improvements in morbidity coding, including updated medical terminology, disease classification, and code specificity. The number of available codes was expanded greatly from approximately 14,000 in ICD-9-CM to over 68,000 in ICD-10-CM.

On an annual basis, the National Birth Defects Prevention Network (NBDPN) publishes state-specific birth defect counts and prevalence estimates for 47 major birth defects, which cover a wide range of organ systems (<https://www.nbdpn.org/ar.php>). Most birth defects surveillance programs in the United States rely on hospital discharge records either as a primary source of passive case-reporting or to assist in identifying cases for medical record review and case confirmation for active surveillance (Stallings et al., 2018). Among the 41

state and territorial population-based programs providing data to NBDPN, approximately 70% report using passive case-finding methodology, with or without case confirmation. Because both passive and active birth defects programs rely on surveillance data derived at least partially from hospital discharge records to estimate the burden of birth defects in their populations, it is important to determine the extent to which the prevalence of birth defects may be influenced by the coding transition from ICD-9-CM to ICD-10-CM.

A number of US studies have evaluated the financial impact or differences in incidence or prevalence of adverse health outcomes between the period before and after the transition to ICD-10-CM (Hellman, Lim, Leung, Blount, & Yiu, 2018; Inscore, Gonzales, Rennix, & Jones, 2018; Panozzo et al., 2018). However, the effect of this transition on birth defects prevalence and trends has not been explored. We leveraged nationally representative all-payer hospital discharge data from the Healthcare Cost and Utilization Project (HCUP) to examine changes and temporal trends in the prevalence of major birth defects among infant hospitalizations in the 45 months prior to and the 15 months immediately after the transition from ICD-9-CM to ICD-10-CM.

## 2 | METHODS

### 2.1 | Data source

We conducted a serial cross-sectional analysis of inpatient hospitalization data for five consecutive years (2012–2016) from the National Inpatient Sample (NIS). The NIS—a product of HCUP sponsored by the Agency for Healthcare Research and Quality, and compiled as part of a federal-state-industry partnership—is the largest publicly available all-payer inpatient database in the United States. Between 44 (in 2012) and 47 (in 2016) states have contributed discharge data from all of their non-federal, short-term general, and specialty hospitals to be compiled annually as part of the NIS (Healthcare Cost and Utilization Project, 2016). To construct the NIS each year, a systematic sampling design is used to select an approximate 20% sample of discharges—henceforth referred to as hospitalizations—from all participating states (Houchens, Ross, Elixhauser, & Jiang, 2014). The systematic sampling ensures that hospitalizations in the NIS are representative of the population on a number of key characteristics such as admission month, reason for hospitalization (represented by the assigned diagnosis-related group [DRG]) and hospital size, location, ownership, and teaching status. Hospitalization-level sampling weights are provided with the NIS to facilitate generation of national estimates of the prevalence and trends of inpatient conditions and procedures. In 2016, the NIS contained over 7.1 million records that were used to generate national estimates representing more than 35.6 million hospitalizations (97% of all hospitalizations in the United States) (Health Care Cost and Utilization Project, 2016).

Each record in the NIS contains various sociodemographic and clinical information. Most relevant to the current study are a variable that captures the principal diagnosis, up to 29 patient-level variables (25 variables in 2012–2013, and 30 variables in 2014–2016) that capture secondary diagnoses made during each hospitalization, and 15 variables capturing diagnostic and therapeutic procedures that were performed. The NIS does not contain personal identifiers that allow for linkage of multiple hospitalizations for the same person;

therefore, the unit of analysis for all NIS-based studies is the hospitalization. The NIS data can contain multiple hospitalizations for a single person.

## 2.2 | Study population and outcomes

The study sample consisted of all hospitalizations of liveborn infants between January 1, 2012 and December 31, 2016 that took place during the first year of life. Patient dates of birth are not provided in the database, so this age-based restriction was operationalized as a hospitalization in which the age (in years) at admission—a variable that is present in the NIS—was zero. For each hospitalization, we scanned all available diagnosis codes, both primary and secondary, for the presence of one of 46 specific major birth defects that were considered to (a) typically be diagnosed within the first year of life, (b) have high public health importance, and (c) be potentially responsive to prevention and intervention strategies (Stallings et al., 2018). These criteria were established as part of a collaboration between the NBDPN, CDC, and surveillance programs in the United States to publish state-level birth defects prevalence estimates annually (Mai et al., 2014). Craniosynostosis, which is also typically monitored and reported by birth defects surveillance programs, was not included due to the lack of a specific ICD-9-CM code used to identify this birth defect. Similar to Mai et al., in addition to the 46 selected defects, we also considered subcategories of two cardiac defects (pulmonary valve atresia and dextro-transposition of the great arteries), which are used by programs in monitoring critical congenital heart defects. The complete list of birth defects is provided in Table 1 along with their ICD-based code definitions. For hospitalizations in which the date of discharge was prior to October 1, 2015, birth defects were coded using ICD-9-CM diagnosis codes; for hospitalizations with date of discharge on or after October 1, 2015, birth defects were coded using ICD-10-CM. Because the data did not contain identifying information and there was no access to the actual medical records, the accuracy and severity of documented defect diagnosis codes could not be verified.

## 2.3 | Covariates

We extracted from the NIS additional patient- and hospital-level characteristics to describe the study samples during the ICD-9-CM and ICD-10-CM windows. The infant's age at admission was indicated dichotomously as during the neonatal period (0–27 days of age) or during the postneonatal period (28–364 days of age). Birth hospitalizations were defined by the presence of both a diagnosis code indicating a live birth (ICD-9-CM: V30–V39; ICD-10-CM: Z38.0–Z38.8) and a DRG code indicative of a newborn hospitalization (789–795) (Grosse, Waitzman, Yang, Abe, & Barfield, 2017). All other hospitalizations were considered to be post-birth. Race/ethnicity was defined as non-Hispanic (NH)-white, NH-black, Hispanic, NH-other, and unknown/unreported. To approximate socioeconomic status, estimates of median household income based on the zip code of the primary residence of the parents were grouped into quartiles. We also grouped the primary payer for each hospitalization into three categories: government (Medicare/Medicaid), private (commercial carrier, private health maintenance organization, and preferred provider organization), and other sources (e.g., self-pay and charity). Additional covariates included timing of the admission (weekday vs. weekend) and the infant's disposition at discharge (routine discharge home, transfer or other discharge alive, death prior to discharge). Hospital characteristics included census region (Northeast, Midwest, South, or West), number of

inpatient beds (small, medium, or large), and facility type (rural, urban nonteaching, or urban teaching).

## 2.4 | Statistical analyses

Descriptive statistics, including weighted frequencies and percentages, were used to describe the study sample during the two periods when ICD-9-CM and when ICD-10-CM were used. Survey weights were applied to yield national estimates. We then estimated the overall and monthly national hospital prevalence of each major birth defect as the number of hospitalizations during the first year of life with an ICD-based code for that defect divided by the total number of first-year hospitalizations; these were presented separately for the ICD-9-CM and ICD-10-CM timeframes. A survey-weighted Poisson regression model was used to estimate 95% confidence intervals (CI) for each hospital prevalence estimate.

To estimate changes in the hospital prevalence of birth defects in inpatient settings attributable to the ICD-9-CM to ICD-10-CM transition, we used an interrupted time series (ITS) framework and corresponding segmented regression analysis. The ITS design has been regarded as the strongest quasi-experimental design to evaluate the impact of events that take place at clearly-defined points in time, such as policies (Lieberman, Polinski, Choudhry, Avorn, & Fischer, 2016), interventions (Leopold et al., 2014), or natural disasters (Ekperi et al., 2018). In addition to its facilitation of easy-to-interpret visualization of results, we chose segmented regression for this study because it allows for investigation of the impact of the code transition on the immediate change in hospital prevalence as well as the change on the temporal trend in hospital prevalence (Penfold & Zhang, 2013). For each defect, the segmented regression model used to fit monthly hospital prevalence rates was as follows:

$$\text{Rate}_t = \beta_0 + \beta_1 * \text{time}_t + \beta_2 * \text{coding system}_t + \beta_3 * \text{time after ICD10 transition}_t + e_t$$

In this model,  $\text{Rate}_t$  is the hospital prevalence of the birth defect in month  $t$ ;  $\text{time}_t$  is a continuous variable documenting the month of analysis from 1 (January 2012) to 60 (December 2016);  $\text{coding system}_t$  is a dichotomous indicator of whether ICD-9-CM ( $\text{coding system} = 0$ ) or ICD-10-CM ( $\text{coding system} = 1$ ) was used;  $\text{time after ICD10 transition}_t$  is an indicator of the number of months after the transition from 1 (October 2015) to 15 (December 2016) and 0 for all months during the ICD-9-CM coding period; and  $e_t$  estimates the random error for each month. The two most important parameter estimates from the model as it pertains to assessing the impact of the coding transition are  $\beta_2$  and  $\beta_3$ .  $\beta_2$  estimates the immediate absolute change in the hospital prevalence following the implementation of ICD-10-CM, and  $\beta_3$  estimates the change in the slope of the temporal hospital prevalence trend following ICD-10-CM implementation. Because the occurrence of individual defects are rare events, there can be considerable variation in defect-specific hospital prevalence across months and years. To allow for a more meaningful comparison of the impact of the transition to ICD-10-CM coding across defects, we expressed the immediate impact as the percent change in the hospital prevalence rate during the ICD-10-CM timeframe relative to the average hospital prevalence rate during the ICD-9-CM period. For statistically significant findings, we further classified the immediate impact of ICD-10-

CM into six groups based on the direction (increase or decrease) and magnitude (50%, 50–99%, 100%) of the change. We used the Durbin–Watson statistic and test to examine autocorrelation, and the Dickey–Fuller unit root test to appraise seasonal fluctuations (stationarity) in the data.

All statistical tests were performed with SAS version 9.4 (Cary, NC) using two-sided statistical tests and a 5% type I error rate. As this constitutes an analysis of publicly-available, deidentified hospital discharge data within the NIS database, the analyses performed for this study were deemed exempt by the Baylor College of Medicine Institutional Review Board.

## 2.5 | Human protection statement

As this constitutes an analysis of publicly-available, deidentified hospital discharge data within the National Inpatient Sample database, the analyses performed for this study were deemed exempt by the Baylor College of Medicine Institutional Review Board.

## 3 | RESULTS

Over 21 million inpatient hospitalizations of infants during the first year of life were included in this 5-year study, 89.3% of which were birth hospitalizations, and 24.9% of which took place during the 15-month ICD-10-CM coding period (Table 2). We observed few meaningful differences in patient characteristics between the ICD-9-CM coding period and the ICD-10-CM coding period with the exception of discharge disposition, which was slightly less likely to be routine (i.e., to home) in the ICD-9-CM period compared with the ICD-10-CM period (95.2% vs. 95.7%,  $p < .01$ ). However, hospitalizations during the ICD-10-CM period were slightly less likely to be from larger hospitals (52.0% vs. 55.9%,  $p < .01$ ) and more likely to be from urban teaching hospitals (67.3% vs. 59.5%,  $p < .01$ ). Table 3 presents the overall prevalence per 10,000 hospitalizations of each specific birth defect included in the study for both coding periods. We observed statistically significant differences in the overall hospital prevalence of 14 of the 46 defects between the ICD-9-CM and ICD-10-CM periods, with the hospital prevalence being higher in the ICD-10-CM period for nine defects (anotia/microtia, transposition of the great arteries, dextro-transposition of the great arteries, interrupted aortic arch, atrial septal defect, atrioventricular septal defect, biliary atresia, renal agenesis/hypoplasia, clubfoot) and lower for five defects (holoprosencephaly, common truncus, pulmonary valve atresia, tricuspid valve atresia and stenosis, cloacal exstrophy). Similar results were observed when restricting the assessment to birth hospitalizations only (data not shown).

Estimates of the immediate impact of the transition to ICD-10-CM coding on the calculated hospital prevalence of each birth defect using segmented regression models expressed as the percent change in hospital prevalence relative to the 45-month ICD-9-CM period are shown in Figure 1 and Table S1. A statistically significant decrease in the immediate hospital prevalence of five individual defects (double outlet right ventricle, esophageal atresia/tracheoesophageal fistula, congenital posterior urethral valves, cloacal exstrophy, and deletion 22q11.2) was associated with the implementation of ICD-10-CM coding, ranging in magnitude from a 23% decrease for double outlet right ventricle of the heart to a several-fold



decrease for cloacal exstrophy. The decrease in hospital prevalence of cloacal exstrophy is depicted in Figure 2. A significant increase in the immediate hospital prevalence of eight birth defects (anencephaly, dextro-transposition of the great arteries, interrupted aortic arch, hypoplastic left heart syndrome, total anomalous pulmonary venous connection, biliary atresia, diaphragmatic hernia, and clubfoot) was observed after the transition to ICD-10-CM, with 5 showing a <50% increase, anencephaly showing a 65% increase, biliary atresia showing a 132% increase, and interruption of the aortic arch (IAA) showing a sevenfold increase. The increase in hospital prevalence of biliary atresia is depicted in Figure 3. Six defects (holoprosencephaly, dextro-transposition of the great arteries, IAA, biliary atresia, cloacal exstrophy, and clubfoot) showed significant changes in both the overall and immediate hospital prevalence following the transition to ICD-10-CM coding. The hospital prevalence of the remaining 26 (56.5%) birth defects showed no significant change in either the overall (Table 3) or immediate (Figure 1, Table S1) hospital prevalence following the transition to ICD-10-CM.

IAA deserves additional mention. Immediately following implementation of ICD-10-CM, the hospital prevalence of IAA showed an abrupt increase from 1.0 per 10,000 in the ICD-9-CM period to 9.7 per 10,000 in the initial ICD-10-CM period (Figure 4, **left panel**).

However, on October 1, 2016, 1 year after its implementation, ICD-10-CM was revised to include a more specific code for IAA only (Q25.21), resulting in a subsequent hospital prevalence of IAA more similar to that in the ICD-9-CM period (Figure 4, **right panel**).

## 4 | DISCUSSION

In this article, we summarize the immediate impact of the coding transition from ICD-9-CM to ICD-10-CM, implemented on October 1, 2015, on the prevalence of specific major birth defects typically monitored by birth defects registries in the United States among infant hospitalizations. Among the 46 defects studied, statistically significant decreases in the immediate hospital prevalence of five defects and significant increases in the immediate hospital prevalence of eight defects were observed after the ICD-10-CM transition. This information can be helpful to surveillance programs in assessing changes in the prevalence of specific birth defects seen in their own data compared with that from a large nationally representative inpatient database.

Some observed changes in hospital prevalence of defects would be expected based on changes made to the ICD-10-CM codes. In ICD-9-CM, holoprosencephaly and cloacal exstrophy were each specified under a code that included a number of other defects of the nervous system and digestive system, respectively. In the more specific ICD-10-CM rubric, each defect has its own unique code, which results in a lower and more accurate estimate of hospital prevalence for each. As mentioned, the initial increase in hospital prevalence of IAA during initial ICD-10 implementation declined with the addition of a specific ICD-10-CM code for this condition 1 year later. Although significant increases in both the overall and immediate hospital prevalence for dextro-transposition of the great arteries, biliary atresia and clubfoot were observed, these are not as readily explained by difference in the codes. Transposition of the great arteries can be a complex anomaly and there is more than one type. Similarly, the term clubfoot can incorporate a variety of foot anomalies that look

similar but are different in nature. It seems reasonable that some degree of misclassification in disease coding would be expected immediately following the change to a new, more expansive and specific coding system such as ICD-10-CM. It will be important to continue to evaluate the prevalence of these conditions in this and other datasets over time as experience with use of the ICD-10-CM codes is gained. The reason for the increase in overall and immediate hospital prevalence for biliary atresia is also unclear. This finding should be verified in other data.

In general, the defects for which there was a significant immediate change in hospital prevalence following the shift to ICD-10-CM but no change in the overall hospital prevalence have considerable variation in their monthly hospital prevalence, with sharp increases and decreases in a some-what inconsistent pattern. This likely reflects the rarity of these individual defects in the general population. The hospital prevalence of some of these defects showed an initial change, either increase or decrease, with a subsequent trend toward the hospital prevalence in the ICD-9-CM period. This might reflect increasing familiarity with ICD-10-CM and specificity of coding. It will be important to continue to assess these changes as additional data accumulate over longer time periods and improved assessment of post ICD-10 trends becomes possible. In some instances, the overall hospital prevalence showed a significant change, either increase or decrease, while the immediate change was not significant. In particular, a steep decline in the hospital prevalence of pulmonary valve atresia and tricuspid valve atresia and stenosis was observed following the ICD-10-CM transition, as well as a statistically significant difference between overall hospital prevalence during the ICD-9-CM and ICD-10-CM periods; however, the immediate impact parameter estimated by segmented regression was not statistically significant. Segmented regression did detect a statistically significant decrease in the trend parameter for pulmonary valve atresia, suggesting a more gradual decrease in hospital prevalence for this defect following the ICD-10-CM transition. Reasons for significant changes in the overall prevalence for other defects, such as anotia/microtia, common truncus, atrial septal defect, atrioventricular septal defect, and renal agenesis/hypoplasia, are more difficult to interpret. The rarity of the conditions, the relatively short ICD-10-CM time period, and limited experience with use of the code may have contributed to these findings. Additional monitoring over time and comparison with findings in other data are indicated. If the change in prevalence persists, further exploration of coding practices at individual healthcare facilities by programs may be warranted.

Among the defects with no significant change in hospital prevalence, 24 exhibited a hospital prevalence during the ICD-10-CM period that was very similar (within 10%) of that for the ICD-9-CM period. These included spina bifida without anencephaly, congenital cataract, pulmonary valve atresia and stenosis, single ventricle, bladder exstrophy, gastroschisis, omphalocele, and trisomy 18, among potentially others. This is not unexpected as the codes for these defects in ICD-9-CM and in ICD-10-CM have similar inclusion criteria and specificity.



#### 4.1 | Strengths and limitations

Strengths of this analysis include the large size of the NIS database with 21 million inpatient hospitalizations of infants during the first year of life, which enables the analysis of hospital prevalence for even very rare defects. Other strengths of the data include its population-based representativeness and weighting to facilitate estimation of national hospital prevalence and trends. There are also a few limitations to consider when interpreting the data. First, there are no personal identifiers contained in the NIS database so that hospitalizations for the same infant could not be deduplicated. Therefore, infants with more than one hospitalization in which the same birth defect was diagnosed would be counted multiple times. However, we observed similar differences in overall hospital prevalence between the ICD-9-CM and ICD-10-CM periods when the analysis was restricted to birth hospitalizations (i.e., one discharge per infant). Second, we are unable to assess the relative accuracy of the assigned ICD-9-CM or ICD-10-CM codes and whether there are differences in accuracy between the two coding rubrics. Studies investigating the accuracy of ICD codes in correctly identifying birth defects to date have focused primarily on ICD-9-CM codes and found a high level of overall accuracy (>93%), but considerable variation in the positive predictive value of these codes across the specific defects investigated (Salemi et al., 2016). Third, the available time period in which to evaluate ICD-10-CM coding was shorter than that for ICD-9-CM, which likely contributed to variations in hospital prevalence estimates following the transition to ICD-10-CM and may have led to spurious statistical results. Finally, the number of variables in the NIS used to capture diagnosis codes changed from 25 in 2012–2013 to 30 in 2014–2016. Additional variables offer increased likelihood of capturing diagnoses, particularly for complex cases; therefore, it is plausible this could influence temporal trends in hospital prevalence of birth defects. However, Salemi et al. previously investigated the impact of expanding the number of diagnosis codes reported in inpatient discharge databases on the counts and rates of birth defects and observed extremely small effects of adding additional diagnosis code fields above 20 (Salemi, Rutkowski, Tanner, Matas, & Kirby, 2018).

#### 4.2 | Next steps

These findings require further exploration in other data systems over longer periods of time. Future efforts to evaluate the extent to which the prevalence and temporal trends seen in birth defects surveillance data are impacted by the ICD-10-CM transition should involve collaborations between birth defects surveillance programs and the use of comparability ratios (Rosamond et al., 2004). Although labor-intensive, comparability ratios based on coding several years of data once using the ICD-9-CM rubric and then separately using ICD-10-CM, will provide a more accurate representation of the impact of ICD-10 implementation.

### 5 | CONCLUSION

This study investigates the impact of the ICD-10-CM transition on temporal trends for major birth defects. We demonstrate differences in hospital prevalence of some birth defects between the two ICD eras, which may in part be due to changes in the codes being used under each rubric. These findings may be helpful to birth defects surveillance programs in

evaluating and interpreting changes in their data potentially related to the transition to ICD-10-CM coding. Policy makers, health care providers, public health experts, and researchers continue to rely on data from state-based birth defects surveillance programs to investigate trends and outbreaks, identify causes and risk factors, and plan for services, referrals and interventions among vulnerable populations (National Birth Defects Prevention Network, 2004). Furthermore, as ICD codes are revised periodically, it is important that surveillance programs continue to evaluate these changes and their impact on birth defect counts and prevalence rates over time.

## Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

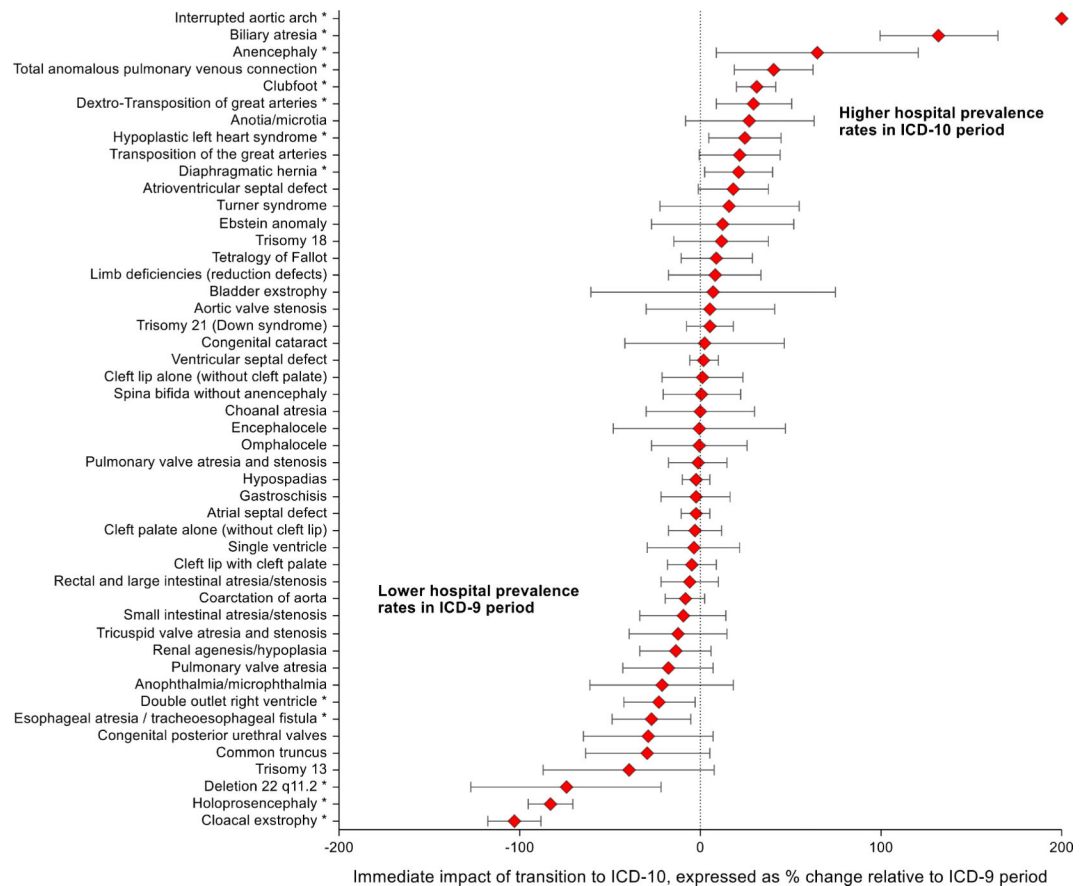
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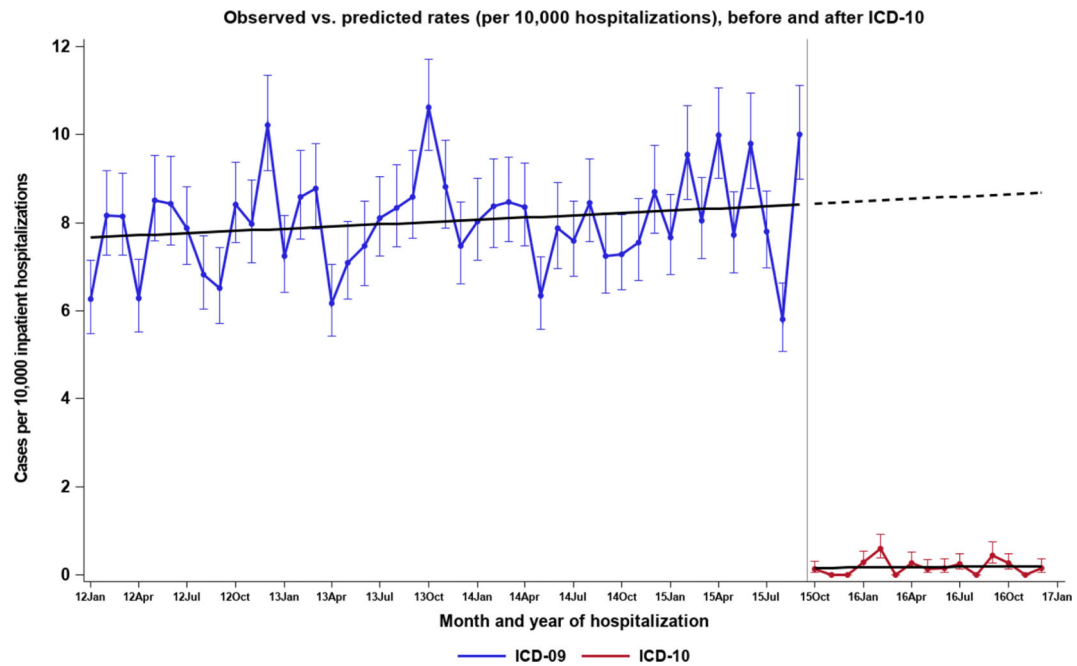
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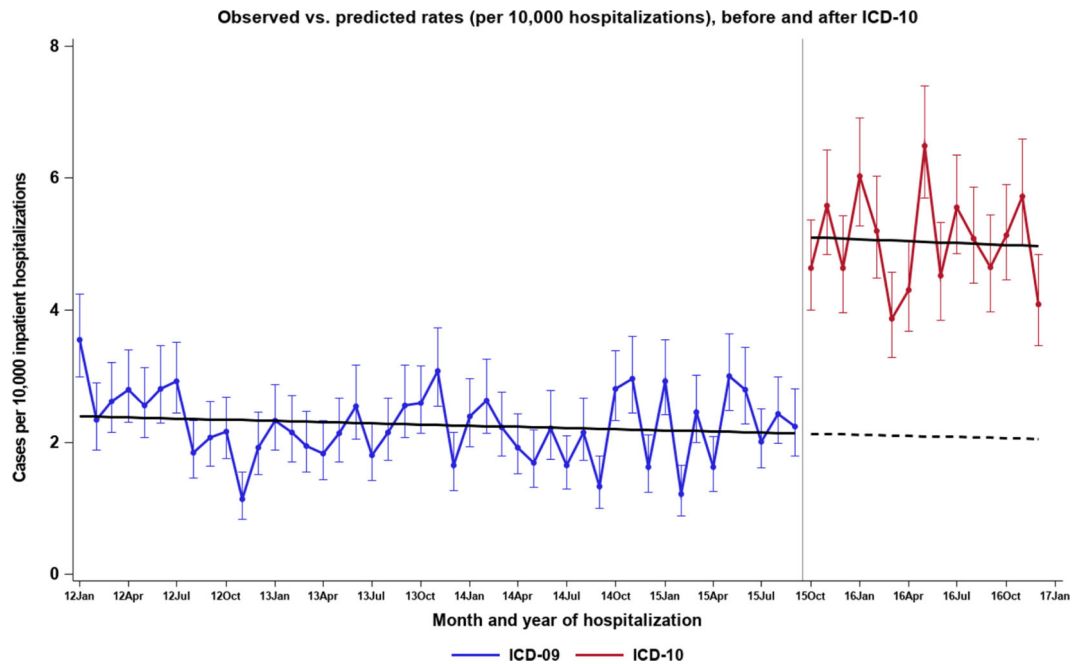
**FIGURE 1.**

Immediate impact of ICD-10-CM on calculated prevalence of each birth defect per 10,000 hospitalizations (hospital prevalence) during the ICD-9-CM versus ICD-10-CM timeframes, 2012–2016. An asterisk (\*) next to a birth defect indicates a statistically significant immediate impact of the transition from ICD-9-CM to ICD-10-CM. The marker representing the change for interrupted aortic arch appears at the maximum of the visible x-axis; however, the actual percent change relative to the ICD-9 period was 771.3 (701.9, 840.7)



**FIGURE 2.**

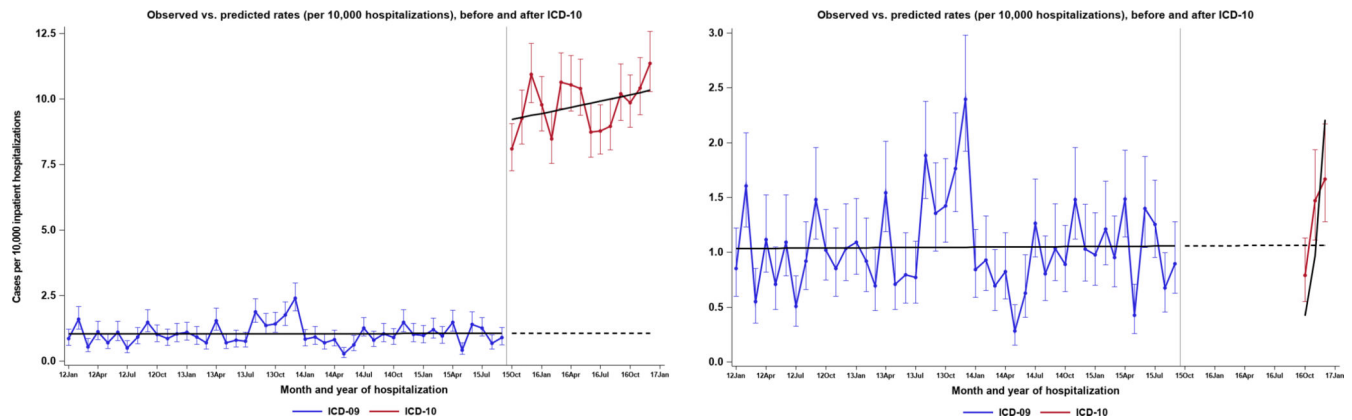
Monthly hospital prevalence estimates for cloacal exstrophy, which demonstrated an abrupt decrease following transition from ICD-9-CM to ICD-10-CM. The blue line, markers, and error bars represent the observed monthly rates during the ICD-9-CM period, with 95% confidence intervals. The red line, markers, and error bars represent the observed monthly rates during the ICD-10-CM period, with 95% confidence intervals. The solid black lines represent the estimated temporal trend during the ICD-9-CM and ICD-10-CM periods, respectively. The dotted lines represent the temporal trend during the ICD-10-CM period that would have occurred if the ICD-9-CM period trend had continued during the ICD-10-CM period



**FIGURE 3.**

Monthly hospital prevalence estimates for biliary atresia, which demonstrated an abrupt increase following transition from ICD-9-CM to ICD-10-CM. The blue line, markers, and error bars represent the observed monthly rates during the ICD-9-CM period, with 95% confidence intervals. The red line, markers, and error bars represent the observed monthly rates during the ICD-10-CM period, with 95% confidence intervals. The solid black lines represent the estimated temporal trend during the ICD-9-CM and ICD-10-CM periods, respectively. The dotted lines represent the temporal trend during the ICD-10-CM period that would have occurred if the ICD-9-CM period trend had continued during the ICD-10-CM period





**FIGURE 4.**

Monthly hospital prevalence estimates for interrupted aortic arch (IAA), which transitioned from a defect-specific ICD-9-CM code to a non-specific ICD-10-CM code on October 1, 2015 (left panel), and back to an IAA-specific ICD-10-CM code on October 1, 2016 (right panel). The blue line, markers, and error bars represent the observed monthly rates during the ICD-9-CM period, with 95% confidence intervals. The red line, markers, and error bars represent the observed monthly rates during the ICD-10-CM period, with 95% confidence intervals. The solid black lines represent the estimated temporal trend during the ICD-9-CM and ICD-10-CM periods, respectively. The dotted lines represent the temporal trend during the ICD-10-CM period that would have occurred if the ICD-9-CM period trend had continued during the ICD-10-CM period

**Table 1**  
Major birth defects, disease classification codes, and key differences between coding systems

Birth defect	ICD-9-CM codes	ICD-10-CM codes	Differences
Central nervous system			
Anencephaly	740.0–740.1	Q00.0–Q00.1	ICD-10-CM specifies inclusion of acephaly; ICD-9-CM does not mention acephaly
Encephalocele	742.0	Q01.0–Q01.9	ICD-10-CM includes Arnold–Chiari type III; ICD-9-CM does not mention Arnold–Chiari type III
Holoprosencephaly	742.2	Q04.2	ICD-10-CM has a specific code for holoprosencephaly; the ICD-9-CM code also includes other reduction deformities of the brain such as absence, agenesis, aplasia, or hypoplasia of part of the brain, agyria, arhinencephaly, or microgyria
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	ICD-10 has separate codes for Arnold–Chiari malformation without spina bifida; ICD-9-CM does not,
Eye			
Anophthalmia/microphthalmia	743.0, 743.1	Q11.0–Q11.2	None
Congenital cataract	743.30–743.34	Q12.0	None
Ear			
Anotia/microtia	744.01, 744.23	Q16.0, Q17.2	None
Cardiovascular	745.0	Q20.0	ICD-10-CM excludes aortic septal defect; ICD-9-CM includes it.
Primary critical congenital heart defects			
Common truncus			
Hypoplastic left heart syndrome	746.7	Q23.4	None
Pulmonary valve atresia and stenosis	746.01, 746.02	Q22.0, Q22.1	None
Pulmonary valve atresia <sup>a</sup>	746.01	Q22.0	None
Tetralogy of Fallot	745.2	Q21.3	ICD-10-CM has a separate code for pentalogy of Fallot; ICD-9-CM includes it with tetralogy of Fallot.
Total anomalous pulmonary venous connection	747.41	Q26.2	ICD-10-CM has a separate code for unspecified anomalous pulmonary venous connection; ICD-9-CM does not
Transposition of the great arteries	745.10, 745.12, 745.19	Q20.3, Q20.5	ICD-10-CM includes ventricular inversion, levotransposition, and dextrotransposition of aorta; ICD-9-CM does not specifically mention these conditions
Dextro-transposition of great arteries <sup>a</sup>	745.10	Q20.3	ICD-10-CM includes dextrotransposition of aorta; the ICD-9-CM does not specifically mention this condition
Secondary critical congenital heart defects			
Coarctation of aorta	747.10	Q25.1	ICD-10-CM has a separate code for hypoplasia of aorta; ICD-9-CM includes it under coarctation of aorta
Double outlet right ventricle	745.11	Q20.1	ICD-10-CM has a separate code for dextrotransposition of the aorta ICD-9-CM includes it under double outlet right ventricle
Birth defect	ICD-9-CM codes	ICD-10-CM codes	Differences

Birth defect	ICD-9-CM codes	ICD-10-CM codes	Differences
Ebstein anomaly	746.2	Q22.5	None
Interrupted aortic arch <sup>b</sup>	747.11	Q25.2, Q25.4 (eff. 10/16) Q25.21	From 10/1/15 to 9/30/16, ICD-10-CM did not mention interrupted aortic arch. A specific code was added on 10/1/2016.
Single ventricle	745.3	Q20.4	ICD-10-CM includes this under double inlet ventricle; ICD-9-CM does not mention double inlet ventricle
Tricuspid valve atresia and stenosis	746.1	Q22.4	None
Other congenital heart defects Aortic valve stenosis	746.3	Q23.0	ICD-10-CM excludes aortic valve stenosis in the setting of hypoplastic left heart syndrome; ICD-9-CM does not specify this exclusion
Atrial septal defect	745.5	Q21.1	ICD-10-CM specifies more individual atrial septal defect types
Atrioventricular septal defect	745.60, 745.61, 745.69	Q21.2	None
Ventricular septal defect	745.4	Q21.0	ICD-10 has a separate code for Eisenmenger's defect; ICD-9-CM includes it under ventricular septal defect
Orofacial			
Choanal atresia	748.0	Q30.0	None
Cleft lip alone (without cleft palate)	749.1	Q36.0-Q36.9	None
Cleft lip with cleft palate	749.2	Q37.0-Q37.9	None
Cleft palate alone (without cleft lip)	749.0	Q35.1-Q35.9	None
Gastrointestinal Biliary atresia	751.61	Q44.2-Q44.3	None
Esophageal atresia/tracheoesophageal fistula	750.3	Q39.0-Q39.4	None
Rectal and large intestinal atresia/stenosis	751.2	Q42.0-Q42.9	None
Small intestinal atresia/stenosis	751.1	Q41.0-Q41.9	None
Genitourinary			
Bladder exstrophy	753.5	Q64.10, Q64.19	None
Cloacal exstrophy	751.5	Q64.12	ICD-10-CM has a specific code for cloacal exstrophy; ICD-9-CM includes this in other anomalies of intestine, which includes a myriad of other defects (e.g., megaloappendix, microcolon, transposition of appendix, colon, or intestine).
Congenital posterior urethral valves	753.6	Q64.2	ICD-10-CM has a specific code for congenital posterior urethral valves; the ICD-9-CM code includes other atresia and stenosis of bladder neck and stricture of urethral meatus.
Hypospadias	752.61	Q54.0-Q54.9, excl.	None
Renal agenesis/hypoplasia	753.0	Q60.0-Q60.6	None
Musculoskeletal Clubfoot	754.51, 754.70	Q66.0, Q66.89	ICD-10-CM includes talipes equinovarus and other specified congenital deformities of the feet; ICD-9-CM describes talipes equinovarus and talipes, unspecified
Diaphragmatic hernia	756.6	Q79.0, Q79.1	None
Gastroschisis	756.73	Q79.3	None

Birth defect	ICD-9-CM codes	ICD-10-CM codes	Differences
Limb deficiencies (reduction defects)	755.2	Q71.0-Q71.9	ICD-10-CM includes lobster-claw hand; ICD-9-CM has a separate code for it
Omphalocele	756.72	Q79.2	None
Chromosomal			
Deletion 22 q11.2	758.32	Q93.81	None
Turner syndrome	758.6	Q96.0-Q96.9	ICD-10-CM has a separate code for gonadal dysgenesis; ICD-9-CM includes it under Turner syndrome
Trisomy 13	758.1	Q91.4-Q91.7	None
Trisomy 18	758.2	Q91.0-Q91.3	None
Trisomy 21 (Down syndrome)	758.0	Q90.0-Q90.9	None

Abbreviations: ICD-9-CM, International Classification of Diseases, Ninth Edition, Clinical Modification; ICD-10-CM, International Classification of Diseases, Tenth Edition, Clinical Modification.

<sup>a</sup>These defect subgroups are used for screening of critical congenital heart defects.

<sup>b</sup>Interrupted aortic arch was analyzed two ways: the first using the nonspecific codes implemented with the first roll-out of ICD-10-CM on October 1, 2015, and the second using the specific code added on October 1, 2016.

Breakdown of the study sample by selected patient and hospital characteristics, stratified by ICD-9-CM versus ICD-10-CM timeframes, 2012–2016<sup>a</sup>

Table 2

Birth defect	ICD-9-CM January 1, 2012–September 30, 2015		ICD-10-CM October 1, 2015–December 31, 2016	
	N <sup>a</sup>	% <sup>a</sup>	N <sup>a</sup>	% <sup>a</sup>
All hospitalizations	15,955,519	100.0	5,279,301	100.0
Age at admission				
Neonatal (0–27 days)	14,873,149	93.2	4,951,791	93.8
Postneonatal (28–364 days)	1,082,265	6.8	327,485	6.2
Sex				
Male	8,255,079	51.7	2,725,638	51.6
Female	7,691,734	48.2	2,550,463	48.3
Race/ethnicity				
White, non-Hispanic	7,540,704	47.3	2,447,878	46.4
Black, non-Hispanic	2,080,450	13.0	703,034	13.3
Hispanic	2,894,120	18.1	953,519	18.1
Asian/Pacific Islander, non-Hispanic	769,745	4.8	274,975	5.2
Native American, non-Hispanic	115,930	0.7	36,010	0.7
Other, non-Hispanic	972,640	6.1	312,145	5.9
Unknown	1,581,930	9.9	551,740	10.5
Median household income				
Lowest quartile	4,503,720	28.2	1,525,573	28.9
Second quartile	3,957,124	24.8	1,271,069	24.1
Third quartile	3,846,145	24.1	1,291,449	24.5
Highest quartile	3,393,790	21.3	1,135,714	21.5
Primary payer				
Medicare/medicaid	7,567,744	47.4	2,494,553	47.3
Private	7,213,179	45.2	2,390,143	45.3
Other <sup>c</sup>	1,174,595	7.4	394,604	7.5
Type of hospitalization				
Birth	14,227,929	89.2	4,735,851	89.7
Post-birth	1,727,590	10.8	543,450	10.3

Birth defect	ICD-9-CM January 1, 2012–September 30, 2015		ICD-10-CM October 1, 2015–December 31, 2016	
	N <sup>a</sup>	% <sup>a</sup>	N <sup>a</sup>	% <sup>a</sup> <sup>b</sup> p
Timing of admission				
Weekday	12,672,594	79.4	4,196,176	79.5
Weekend	3,282,915	20.6	1,083,119	20.5
Discharge disposition				<.01
Routine (home)	15,195,399	95.2	5,052,001	95.7
Transfer, home health, AMA	698,430	4.4	205,170	3.9
Died	59,180	0.4	19,035	0.4
US census region				.96
Northeast	2,593,934	16.3	844,395	16.0
Midwest	3,410,253	21.4	1,119,640	21.2
South	6,107,259	38.3	2,056,462	39.0
West	3,844,073	24.1	1,258,803	23.8
Hospital bed size				<.01
Small	2,303,400	14.4	920,360	17.4
Medium	4,728,915	29.6	1,611,532	30.5
Large	8,923,204	55.9	2,747,409	52.0
Hospital type				<.01
Rural	1,553,798	9.7	464,709	8.8
Urban, nonteaching	4,902,580	30.7	1,263,817	23.9
Urban, teaching	9,499,141	59.5	3,550,774	67.3

Abbreviations: AMA, against medical advice; ICD-9-CM, International Classification of Diseases, Ninth Edition, Clinical Modification; ICD-10-CM, International Classification of Diseases, Tenth Edition, Clinical Modification.

<sup>a</sup>Weighted to estimate national frequency; sum of all groups may not add up to the total and percentages may not add to 100% due to missing data.

<sup>b</sup>p-value calculated from a Rao-Scott chi-square test.

<sup>c</sup>Includes self-pay, no charge, and other payers.



Overall prevalence of each birth defect per 10,000 hospitalizations (hospital prevalence) during the first year of life, stratified by ICD-9-CM versus ICD-10-CM timeframes, 2012–2016

Table 3

Birth defect	ICD-9-CM January 1, 2012–September 30, 2015		ICD-10-CM October 1, 2015–December 31, 2016	
	N <sup>a</sup>	Rate (95% CI)	N <sup>a</sup>	Rate (95% CI)
Central nervous system Anencephaly	1,070	0.7 (0.6, 0.7)	400	0.8 (0.7, 0.8)
Encephalocele	1,800	1.1 (1.1, 1.2)	660	1.3 (1.2, 1.3)
Holoprosencephaly	14,380	9.0 (8.9, 9.2)	880	1.7 (1.6, 1.8)
Spina bifida without anencephaly	9,715	6.1 (6.0, 6.2)	3,005	5.7 (5.5, 5.9)
Eye	2,275	1.4 (1.4, 1.5)	780	1.5 (1.4, 1.6)
Anophthalmia/microphthalmia				
Congenital cataract	1,855	1.2 (1.1, 1.2)	570	1.1 (1.0, 1.2)
Ear	2,520	1.6 (1.51, 1.6)	1	195 2.3 (2.12, 2.4)
Anotia/microtia				
Cardiovascular	2,090	1.3 (1.3, 1.4)	520	1.0 (0.9, 1.1)
Primary critical congenital heart defects				
Common truncus				
Hypoplastic left heart syndrome	11,320	7.1 (7.0, 7.2)	4,175	7.9 (7.7, 8.2)
Pulmonary valve atresia and stenosis	17,915	11.2 (11.1, 11.4)	5,840	11.1 (10.8, 11.3)
Pulmonary valve atresia <sup>c</sup>	4,430	2.8 (2.7, 2.9)	1,125	2.1 (2.0, 2.3)
Tetralogy of Fallot	16,045	10.1 (9.9, 10.2)	5,615	10.6 (10.4, 10.9)
Total anomalous pulmonary venous connection	3,470	2.2 (2.1, 2.2)	1,370	2.6 (2.5, 2.7)
Transposition of the great arteries	7,855	4.9 (4.8, 5.0)	3,185	6.0 (5.8, 6.2)
Dextro-transposition of great arteries <sup>c</sup>	6,445	4.0 (3.9, 4.1)	2,990	5.7 (5.5, 5.9)
Secondary critical congenital heart defects	17,050	10.7 (10.5, 10.8)	5,285	10.0 (9.7, 10.3)
Coarctation of aorta				
Double outlet right ventricle	7,720	4.8 (4.7, 4.9)	2,710	5.1 (4.9, 5.3)
Ebstein anomaly	2,230	1.4 (1.3, 1.5)	665	1.3 (1.2, 1.4)
Interrupted aortic arch <sup>d</sup>	1,670	1.0 (1.0, 1.1)	5,145	9.7 (9.5, 10.0)
Single ventricle	4,085	2.6 (2.5, 2.6)	1,655	3.1 (3.0, 3.3)
Tricuspid valve atresia and stenosis	3,780	2.4 (2.3, 2.4)	965	1.8 (1.7, 1.9)
Other congenital heart defects	2,965	1.9 (1.8, 1.9)	1,035	2.0 (1.8, 2.1)

Birth defect	ICD-9-CM January 1, 2012–September 30, 2015		ICD-10-CM October 1, 2015–December 31, 2016	
	N <sup>a</sup>	Rate (95% CI)	N <sup>a</sup>	Rate (95% CI)
<b>Birth defect</b>				<i>b</i> <i>p</i>
Aortic valve stenosis	306,605	192.2 (191.5, 192.8)	111,950	212.1 (210.8, 213.3)
Atrial septal defect	12,040	7.5 (7.4, 7.7)	4,850	9.2 (8.9, 9.4)
Atrioventricular septal defect	106,990	67.1 (66.7, 67.5)	37,910	71.8 (71.1, 72.5)
Ventricular septal defect	3,990	2.5 (2.4, 2.6)	1,320	2.5 (2.4, 2.6)
Orofacial				
Choanal atresia	6,760	4.2 (4.1, 4.3)	2,135	4.0 (3.9, 4.2)
Cleft lip alone (without cleft palate)	17,595	11.0 (10.9, 11.2)	5,620	10.6 (10.4, 10.9)
Cleft lip with cleft palate	17,495	11.0 (10.8, 11.1)	5,240	9.9 (9.7, 10.2)
Cleft palate alone (without cleft lip)	3,605	2.3 (2.2, 2.3)	2,660	5.0 (4.9, 5.2)
Gastrointestinal				
Biliary atresia	6,810	4.3 (4.2, 4.4)	2,300	4.4 (4.2, 4.5)
Esophageal atresia/tracheoesophageal fistula	14,400	9.0 (8.9, 9.2)	4,870	9.2 (9.0, 9.5)
Rectal and large intestinal atresia/stenosis	9,540	6.0 (5.9, 6.1)	2,845	5.4 (5.2, 5.6)
Small intestinal atresia/stenosis	745	0.5 (0.4, 0.5)	250	0.5 (0.4, 0.5)
Genitourinary				
Bladder exstrophy	12,810	8.0 (7.9, 8.2)	95	0.2 (0.1, 0.2)
Cloacal exstrophy	2,725	1.7 (1.6, 1.8)	940	1.8 (1.7, 1.9)
Congenital posterior urethral valves	58,355	36.6 (36.3, 36.9)	19,110	36.2 (35.7, 36.7)
Hypospadias	9,935	6.2 (6.1, 6.4)	3,740	7.1 (6.9, 7.3)
Renal agenesis/hypoplasia	26,785	16.8 (16.6, 17.0)	12,045	22.8 (22.4, 23.2)
Musculoskeletal				
Clubfoot	7,640	4.8 (4.7, 4.9)	2,555	4.8 (4.7, 5.0)
Diaphragmatic hernia	9,860	6.2 (6.1, 6.3)	3,020	5.7 (5.5, 5.9)
Gastroschisis	6,190	3.9 (3.8, 4.0)	2,190	4.1 (4.0, 4.3)
Limb deficiencies (reduction defects)	4,175	2.6 (2.5, 2.7)	1,455	2.8 (2.6, 2.9)
Omphalocele	1,210	0.8 (0.7, 0.8)	355	0.7 (0.6, 0.7)
Chromosomal				
Deletion 22 q11.2	1,760	1.1 (1.1, 1.2)	705	1.3 (1.2, 1.4)
Turner syndrome	1,810	1.1 (1.1, 1.2)	520	1.0 (0.9, 1.1)
Trisomy 13	3,865	2.4 (2.3, 2.5)	1,370	2.6 (2.5, 2.7)
Trisomy 18	40,515	25.4 (25.1, 25.6)	14,220	26.9 (26.5, 27.4)
Trisomy 21 (Down syndrome)				

*Note:* ICD-9-CM period, hospitalizations in which the discharge was made between January 1, 2012 and September 30, 2015. ICD-10-CM period, hospitalizations in which the discharge was made between October 1, 2015 and December 31, 2016.

<sup>a</sup>Weighted to estimate national frequency.

<sup>b</sup>*p*-value calculated from a Rao-Scott chi-square test.

<sup>c</sup>These defect subgroups are used for screening of critical congenital heart defects.

<sup>d</sup>Interrupted aortic arch was analyzed two ways: the first using the nonspecific codes implemented with the first roll-out of ICD-10-CM on October 1, 2015, and the second using the specific code added on October 1, 2016. What is presented is the original change made October 1, 2015.