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Racial/Ethnic Differences in Survival of United States Children with Birth Defects: A Population-Based Study

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Abstract

Objectives—To examine racial/ethnic-specific survival of children with major birth defects in the US.

Study design—We pooled data on live births delivered during 1999-2007 with any of 21 birth defects from 12 population-based birth defects surveillance programs. We used the Kaplan-Meier method to calculate cumulative survival probabilities and Cox proportional hazards models to estimate mortality risk.

Results—For most birth defects, there were small-to-moderate differences in neonatal (<28 days) survival among racial/ethnic groups. However, compared with children born to non-

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*List of centers of the National Birth Defects Prevention Network is available at www.jpeds.com (Appendix).

The findings and conclusion in this report are those of the authors and do not necessarily represent the official position of the Centers for Disease Control and Prevention. The authors declare no conflict of interest.

Hispanic white mothers, postneonatal infant (28 days to <1 year) mortality risk was significantly greater among children born to non-Hispanic black mothers for 13 of 21 defects (hazard ratios [HRs] 1.3-2.8) and among children born to Hispanic mothers for 10 of 21 defects (HRs 1.3-1.7). Compared with children born to non-Hispanic white mothers, a significantly increased childhood (≤8 years) mortality risk was found among children born to Asian/Pacific Islander mothers for encephalocele (HR 2.6), tetralogy of Fallot, and atrioventricular septal defect (HRs 1.6-1.8) and among children born to American Indian/Alaska Native mothers for encephalocele (HR 2.8), whereas a significantly decreased childhood mortality risk was found among children born to Asian/Pacific Islander mothers for cleft lip with or without cleft palate (HR 0.6).

Conclusion—Children with birth defects born to non-Hispanic black and Hispanic mothers carry a greater risk of mortality well into childhood, especially children with congenital heart defect. Understanding survival differences among racial/ethnic groups provides important information for policy development and service planning.

Birth defects are a leading cause of infant death in the US.[1] National vital statistics data are critical to our understanding of infant mortality[2] and child and adult mortality.[3,4] However, compared with population-based birth defects surveillance systems, birth certificates have relatively poor sensitivity and specificity for the reporting of birth defects.[5] Linking population-based birth defects surveillance data to state death certificates and the National Death Index (NDI) can provide high high-quality information on both short- and long-term survival of children with birth defects.

There have been several previous studies on survival of infants with birth defects using statewide[6-14] or regional[15-21] population-based birth defects surveillance data. The use of pooled data from several surveillance systems in the US, however, has been limited to only a few studies of individual defects.[22-24] Previous literature suggests that the mortality and survival experience of children with birth defects differs by specific birth defect phenotype and by demographic factors such as maternal race/ethnicity.[12-14,25-28] Racial/ethnic disparities in infant and child mortality were found among Florida[29] and Texas infants with birth defects[25,27,28] but not among New York children (up to 25 years) with birth defects.[12]

To date, no studies using pooled population-based surveillance data have investigated the survival of children with a broad range of birth defects. A recent study using pooled data from 12 population-based birth defects surveillance programs in the US examined the relationship between race/ethnicity and occurrence of selected major birth defects.[30] Using that study population, in the current study we estimated infant and child survival by birth defect subtype and race/ethnicity among live-born individuals with selected birth defects.

Methods

Information on all live births with any of the selected major birth defects was obtained from 12 participating population-based birth defects surveillance programs: Arizona, Colorado, Florida, Georgia (5 counties of metropolitan Atlanta), Illinois, Massachusetts, Michigan, Nebraska, New Jersey, New York (excludes New York City), North Carolina, and Texas. Surveillance programs matched cases to state birth certificate records to obtain data on maternal race/ethnicity, classified as non-Hispanic white (NHW), non-Hispanic black

(NHB), Hispanic, Asian/Pacific Islander (A/PI), and American Indian/Alaska Native (AI/AN). The study protocol was reviewed and approved by the participating states' institutional review boards, as necessary.

The birth defects included in the study were spina bifida without anencephalus; encephalocele; common truncus; transposition of great arteries; tetralogy of Fallot; atrioventricular septal defect (AVSD) (and a subgroup without co-occurring Down syndrome); aortic valve stenosis; hypoplastic left heart syndrome; coarctation of the aorta; cleft palate without cleft lip; cleft lip with or without cleft palate; esophageal atresia/s tracheoesophageal fistula; pyloric stenosis; rectal, anal, and large intestinal atresia/stenosis; upper and lower limb deficiencies; diaphragmatic hernia; gastroschisis; omphalocele; and Down syndrome. States selected cases from their surveillance systems for inclusion in this analysis based on a list of specified International Classification of Diseases, 9th Revision, Clinical Modification or Centers for Disease Control and Prevention/British Pediatric Association Classification of Diseases codes that are used for annual reporting by the National Birth Defects Prevention Network.[31] The birth defects included are not mutually exclusive, and infants with multiple defects were included in each relevant birth defect category.

Each state surveillance program linked its case information to the state's death certificate data files to obtain the vital status information of the study cohort. The follow-up period for children in the study ranged from 1 (for those born at the end of 2007 followed through the end of 2008) up to 9 years (for those born in the beginning of 1999 followed through the end of 2008). Illinois and Nebraska programs only provided vital status information for the first year. If a child was deceased, participating programs provided the date of death and duration of life in days. Additional data sources used to obtain vital status information included hospital discharge files (Arizona, Texas), medical records (Arizona, Texas), and the NDI (Georgia, Michigan).

Statistical Analyses

The Kaplan-Meier product limit method was used to calculate survival probabilities (<1 day, <7 days, <28 days, <1 year, <2 years, ≤8 years) for specific defects and by maternal race/ethnicity. Greenwood method was used to calculate 95% CIs. The infant survival analysis was conducted using data from all 12 birth defects surveillance programs. For the analyses of survival beyond infancy, data for those born during 1999-2005 from 10 programs (note: Massachusetts was 2000-2007 and North Carolina was 2003-2007) were analyzed; Illinois and Nebraska were excluded from the analyses of survival beyond infancy because they did not provide vital status data beyond one year of life. Because the birth cohort for one of the participating states (New Jersey) was through 2005 only, 2005 was chosen as the latest birth year to be included for all 10 programs in the analysis. Thus, the longest possible period of follow-up was just under 9 years (infants born in the beginning of 1999 with follow-up though the end of 2008).

Multivariable analyses using Cox proportional hazards models were conducted to estimate the mortality risk, the hazard ratio (HR), for each birth defect, with adjustment for the following covariates: birth weight and gestational age (<37 weeks and <2500 g, <37 weeks

and ≥2500 g, ≥37 weeks and <2500 g, and ≥37 weeks and ≥2500 g),[22] maternal age (<35 and ≥35 years), birth period (1999-2000, 2001-2002, 2003-2005, and 2006-2007), and state surveillance program. These variables were selected because bivariate analyses indicated these factors were associated with survival ($P < .1$). Other factors, such as mother's birth country, marital status, insurance status, and method of delivery were excluded from the multivariable models because they were not available from all participating surveillance programs. SAS Version 9.2 (SAS Institute, Cary, North Carolina) was used for all statistical analyses.

Results

The study cohort contained 98 833 children born alive in 1999-2007 with at least 1 of the selected major birth defects and ascertained from the 12 state surveillance programs (Table I; available at www.jpeds.com) among approximately 14 million live births (about 39% of all live births in the US during the study period). The study cohort did not include 2007 births from Colorado, Illinois, Michigan, and Nebraska and 2006-2007 births from New Jersey because of unavailability of the vital status data; the earliest available data were 2000 for Massachusetts and 2003 for North Carolina. A total of 9997 deaths were identified in the study cohort, with 8893 (89%) occurring during infancy.

The lowest 1-day and 7-day survival probabilities were found for encephalocele (Table II). Children with hypoplastic left heart syndrome had the lowest neonatal (<28 days), infant (<1 year), and childhood (<2 years and <8 years) survival probability. Of the 21 birth defects studied, 6 (spina bifida, cleft palate, cleft lip with or without cleft palate, pyloric stenosis, gastroschisis, and Down syndrome) had >90% survival for all ages examined. At every age, children with AVSD without co-occurring Down syndrome experienced poorer survival than children with AVSD overall.

For most birth defects examined (excluding spina bifida, tetralogy of Fallot, pyloric stenosis, and Down syndrome), there were small-to-moderate (≤5%) absolute differences in neonatal survival among the 3 major racial/ethnic groups (NHW, NHB, and Hispanic); the differences were striking for common truncus, esophageal atresia/tracheoesophageal fistula, and diaphragmatic hernia (Table III). Similarly, with the exception of spina bifida, pyloric stenosis and Down syndrome, all birth defects exhibited at least a 5% difference in infant survival across the 3 major racial/ethnic groups; infants born to NHB and Hispanic mothers had consistently lower infant survival than those born to NHW mothers. Neonatal survival among infants of A/PI and AI/AN mothers generally was comparable with that of NHW mothers with the exceptions of markedly lower survival for encephalocele and hypoplastic left heart syndrome, and common truncus (AI/AN only). At least a 5% lower infant survival was found among infants of A/PI and AI/AN mothers for several defects: encephalocele, common truncus, AVSD, hypoplastic left heart syndrome, coarctation of the aorta, and omphalocele.

Similar to infant survival, with the exception of spina bifida, pyloric stenosis, upper limb deficiencies, gastroschisis, and Down syndrome, there was 5% or greater variability in early childhood survival (<2 years) among the 3 major racial/ethnic groups; the survival

probability among children born to NHB mothers was nearly universally lower than that among children born to NHW mothers, with the largest difference noted for transposition of the great arteries (Table IV). Compared with children of NHW mothers, the survival probability among children of A/PI and AI/AN mothers was substantially lower for encephalocele, common truncus, AVSD, hypoplastic left heart syndrome, and coarctation of the aorta.

Results from multivariable analysis (Table V) showed that, compared with children of NHW mothers, the overall childhood (≤8 years) mortality risk was significantly greater among children born to NHB mothers for 12 of 21 defects (HR 1.3-2.0), children born to Hispanic mothers for 8 defects (HR 1.3-1.6), children born to A/PI mothers for 4 defects (HR 1.6-2.6), and children born to AI/AN mothers for only 1 defect, encephalocele (HR 2.8). However, a significantly decreased overall mortality risk was found among children born to A/PI mothers for cleft lip with or without cleft palate (HR 0.6). Among children of A/PI mothers, a significantly increased mortality risk was found for hypoplastic left heart syndrome (HR 1.6) during the neonatal period and for transposition of great arteries (HR 3.6) and tetralogy of Fallot (HR 2.4) during early childhood (1-8 years); a significantly decreased neonatal mortality risk was found for cleft lip with or without cleft palate (HR 0.5), compared with children born to NHW mothers.

Discussion

For most of the major birth defects included in this study, we found maternal racial/ethnic differences in survival and mortality risk for all survival age groups examined. These findings are consistent with previous studies in which the authors used data from a single birth defects surveillance program.[14,25,27-29] Black-white disparities in mortality risk consistently were observed across birth defect types during the postneonatal infancy period and continued to widen in childhood for some of the more severe congenital heart defects. Racial and ethnic disparities in health often represent potential prevention opportunities, and this pattern of changing racial/ethnic disparities across early childhood for these complex conditions suggests specific age periods that could be amenable to health services and policy interventions that address improved access to and delivery of quality and timely care.

Using the same birth cohort as we did in the current study, others previously have reported racial/ethnic disparities in prevalence for several major birth defects.[30] Significantly greater risks in both overall childhood prevalence as well as increased mortality were found among children of NHB mothers for tetralogy of Fallot and AVSD and among children of AI/AN mothers for encephalocele, compared with children of NHW mothers.

Our study found that children who had AVSD without co-occurring Down syndrome had poorer childhood survival compared with children with both AVSD and Down syndrome across all racial/ethnic groups. Previous studies[32-34] have shown that children with AVSD with a normal chromosome complement had a statistically greater risk of requiring reoperation than did children with AVSD and Down syndrome. However, a recent study did not find a difference in survival between the 2 groups.[20] There is a possibility that a greater

proportion of infants with both AVSD and co-occurring Down syndrome are diagnosed prenatally compared with infants with AVSD alone.

This study was subject to several limitations. There was a potential for incomplete ascertainment of deaths possibly from missed matches of the study cohort to state death certificate files or underascertainment of out of state deaths. By potentially missing these deaths, we may have overestimated the survival probabilities. However, overall ascertainment of deaths for the 2 states that used NDI for vital status determination was not appreciably different from that of the other states.

Another limitation was the potential misclassification of birth defect diagnoses for cases obtained from the birth defects surveillance programs that rely exclusively on case reporting by physicians and hospitals (passive case ascertainment). Seven of the 12 participating programs use a passive case-finding methodology, and 4 of these 7 programs validate the accuracy of the birth defect case diagnosis through active case follow-up. The 3 programs with no case-verification protocol in place would be the most susceptible to misclassification. Sensitivity analyses showed that the estimated survival probabilities using data from all 12 surveillance programs were 3%-10% greater for 4 of the 21 defects compared with the estimated survival probabilities excluding the 3 passive surveillance programs (data not shown). The overestimate of survival for the 4 defects could be attributable to underascertainment of deaths or misclassification of noncases as cases (more likely for congenital heart defects than for encephalocele).

Additional limitations include: (1) wide 95% CIs associated with the estimated survival probabilities for several defects among A/PI and AI/AN subgroups attributable to small sample sizes; and (2) lack of data on potentially important clinical factors (eg, timing and age of the child at initial diagnosis, the severity of the defect, and whether the child had isolated or nonisolated defects), demographic factors (eg, socioeconomic status[13] and health insurance payer[29]) and hospital factors (eg, nursery care level at the hospital of delivery[35]) that are also likely to play a role. Considering these limitations and the descriptive nature of the analysis, the survival estimates presented here should be interpreted cautiously.

Despite these limitations, the survival analyses reported in this study are based on an unprecedented dataset. Pooling data from 12 birth defects surveillance programs, all of which linked their surveillance data to vital records data enabled the assembly of the largest population-based cohort of US infants with birth defects for whom survival up to age eight years could be calculated. The defect-specific sample sizes allowed for relatively precise survival estimates for most birth defects subtypes. These data also provided an opportunity to examine up to 8-year survival among less common racial/ethnic groups (ie, A/PI and AI/AN) and for selected defects among NHBs and Hispanics for which no previous survival data were available. Future investigations should focus on mortality outcomes associated with surgical intervention, co-occurring conditions requiring hospitalization or outpatient procedures, and complexity of case presentation for children with specific birth defects.

Appendix

Centers that included data for the National Birth Defects Prevention Network include:

Arizona Birth Defects Monitoring Program, Metropolitan Atlanta Congenital Defects Program, Colorado Responds to Children with Special Needs, Florida Birth Defects Registry, Illinois Adverse Pregnancy Outcomes Reporting System, Massachusetts Birth Defects Monitoring Program, Michigan Birth Defects Registry, Nebraska Birth Defects Registry, New Jersey Special Child Health Services Registry, New York State Congenital Malformations Registry, North Carolina Birth Defects Monitoring Program, and Texas Birth Defects Epidemiology, and Surveillance Branch.

Glossary

A/PI	Asian/Pacific Islander
AI/AN	American Indian/Alaska Native
AVSD	Atrioventricular septal defect
HR	Hazard ratio
NDI	National Death Index
NHB	Non-Hispanic black
NHW	Non-Hispanic white

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Table II

Overall survival probabilities and 95% CIs for infants and children with selected birth defects by survival age and birth defect category based on pooled data from 12 state birth defects surveillance programs, National Birth Defects Prevention Network, 1999-2007

Birth defects	Infant survival probability* (95% CI)						Childhood (up to 8 years) survival probability† (95% CI)			
	No. live births with defects‡	No. deaths with defects‡	<1 d	<7 d	<28 d	<1 y	No. live births with defects‡	No. deaths with defects‡	<2 y	≤8 y
Central nervous system defects										
Spina bifida without anencephalus	3903	318	96.9 (96.3-97.4)	95.4 (94.7-96.0)	94.3 (93.5-95.0)	91.9 (90.9-92.7)	2704	266	90.7 (89.6-91.8)	90.2 (89.0-91.2)
Encephalocele	909	254	88.6 (86.3-90.5)	80.2 (77.5-82.6)	77.7 (74.8-80.2)	72.1 (69.0-74.9)	627	189	70.3 (66.6-73.7)	69.9 (66.1-73.3)
Congenital heart defects										
Common truncus	956	238	98.2 (97.2-98.9)	94.1 (92.5-95.5)	87.2 (85.0-89.2)	75.1 (72.2-77.7)	670	191	72.4 (68.8-75.6)	71.5 (67.9-74.8)
Transposition of great arteries	4330	705	98.7 (98.3-99.0)	95.5 (94.8-96.0)	90.9 (90.1-91.8)	83.7 (82.6-84.8)	3160	601	82.0 (80.6-83.3)	81.0 (79.6-82.3)
Tetralogy of Fallot	5208	674	99.3 (99.1-99.5)	97.2 (96.8-97.6)	94.6 (94.0-95.2)	87.1 (86.1-87.9)	3730	573	85.5 (84.4-86.6)	84.6 (83.4-85.8)
AVSD	4884	972	98.8 (98.5-99.1)	95.7 (95.1-96.2)	91.6 (90.7-92.3)	80.1 (79.0-81.2)	3523	825	78.1 (76.7-79.4)	76.6 (75.1-77.9)
AVSD (without Down syndrome)	2450	711	98.0 (97.4-98.5)	92.5 (91.4-93.5)	86.0 (84.5-87.3)	71.0 (69.1-72.7)	1810	594	69.1 (66.9-71.1)	67.2 (65.0-69.3)
Aortic valve stenosis	2646	435	99.2 (98.8-99.5)	96.3 (95.5-97.0)	91.8 (90.7-92.8)	83.6 (82.1-84.9)	1958	363	82.5 (80.7-84.1)	81.5 (79.7-83.1)
Hypoplastic left heart syndrome	2976	1334	96.9 (96.2-97.5)	87.0 (85.7-88.2)	73.1 (71.5-74.7)	55.2 (53.4-56.9)	2077	1030	52.7 (50.5-54.8)	50.4 (48.2-52.5)
Coarctation of aorta	6365	985	99.4 (99.2-99.6)	97.0 (96.5-97.4)	92.5 (91.8-93.1)	84.5 (83.6-85.4)	4543	826	82.7 (81.5-83.7)	81.8 (80.7-82.9)
Oral clefts										
Cleft palate without cleft lip	7356	660	98.3 (98.0-98.6)	96.5 (96.0-96.9)	94.8 (94.2-95.3)	91.0 (90.4-91.7)	5204	504	90.9 (90.1-91.6)	90.3 (89.5-91.1)
Cleft lip with or without cleft palate	11 862	999	97.7 (97.4-98.0)	95.3 (94.9-95.7)	94.0 (93.5-94.4)	91.6 (91.1-92.1)	8351	771	91.2 (90.6-91.8)	90.8 (90.1-91.4)
Gastrointestinal defects										
Esophageal atresia/tracheoesophageal fistula	3084	476	97.5 (96.9-98.0)	92.9 (91.9-93.8)	90.0 (88.9-91.0)	84.6 (83.2-85.8)	2192	356	84.4 (82.8-85.8)	83.8 (82.1-85.2)
Pyloric stenosis	21 233	109	100.0 (100.0-100.0)	100.0 (99.9-100.0)	99.9 (99.9-100.0)	99.5 (99.4-99.6)	15 883	110	99.4 (99.3-99.5)	99.3 (99.2-99.4)
Rectal and large intestinal atresia/stenosis	5400	702	95.9 (95.3-96.4)	92.6 (91.8-93.2)	90.9 (90.1-91.6)	87.0 (86.1-87.9)	3866	537	86.6 (85.5-87.6)	86.1 (85.0-87.2)
Musculoskeletal defects										
Upper limb deficiencies	3602	387	96.5 (95.8-97.0)	94.0 (93.2-94.8)	92.6 (91.7-93.4)	89.3 (88.2-90.2)	2527	298	88.6 (87.3-89.8)	88.2 (86.9-89.4)
Lower limb deficiencies	1913	219	94.9 (93.8-95.8)	92.9 (91.7-94.0)	91.5 (90.1-92.6)	88.6 (87.0-89.9)	1349	159	88.7 (86.8-90.2)	88.2 (86.4-89.8)
Diaphragmatic hernia	3248	1017	91.8 (90.8-92.7)	83.9 (82.6-85.1)	76.1 (74.6-77.5)	68.7 (67.1-70.3)	2174	695	68.3 (66.3-70.2)	68.0 (66.0-69.9)
Gastroschisis	3698	266	98.5 (98.0-98.8)	96.7 (96.1-97.3)	95.8 (95.1-96.4)	92.8 (91.9-93.6)	2326	183	92.3 (91.1-93.3)	92.1 (91.0-93.2)
Omphalocele	1281	367	88.7 (86.8-90.3)	82.1 (79.9-84.1)	78.5 (76.1-80.6)	71.4 (68.8-73.7)	844	243	71.4 (68.3-74.4)	71.2 (68.0-74.1)

Birth defects	Infant survival probability* (95% CI)						Childhood (up to 8 years) survival probability† (95% CI)			
	No. live births with defects‡	No. deaths with defects‡	<1 d	<7 d	<28 d	<1 y	No. live births with defects‡	No. deaths with defects‡	<2 y	≤8 y
Chromosomal defects										
Trisomy 21 (Down syndrome)	15939	944	98.9 (98.7-99.0)	98.1 (97.9-98.3)	97.2 (96.9-97.4)	94.1 (93.7-94.4)	10880	787	93.4 (92.9-93.8)	92.8 (92.3-93.2)

*Children born in 1999-2007 from all 12 states.

†Children born in 1999-2005 from 10 states; data from Illinois and Nebraska were excluded because no vital status data beyond infancy were available.

‡Children with 2 or more birth defects may be counted in multiple categories.

Table III

Survival probabilities and 95% CIs for infants with selected birth defects by survival age (<28 days, <1 year), birth defect category, and maternal race/ethnicity based on pooled data from 12 state birth defects surveillance programs, National Birth Defects Prevention Network, 1999-2007

	Neonatal survival probability (<28 d)					Infant survival probability (<1 y)				
Birth defects*	**NHW**	**NHB**	**Hispanic**	**A/PI**	**AI/AN**	**NHW**	**NHB**	**Hispanic**	**A/PI**	**AI/AN**
Central nervous system defects										
Spina bifida without anencephalus	94.0 (92.9-95.0)	92.8 (90.1-94.7)	95.2 (93.9-96.2)	98.4 (88.9-99.8)	94.1 (78.5-98.5)	92.0 (90.7-93.1)	88.6 (85.4-91.1)	92.7 (91.1-94.0)	98.4 (88.9-99.8)	91.2 (75.1-97.1)
Encephalocele	77.8 (73.3-81.6)	83.8 (77.8-88.2)	75.2 (69.8-79.7)	66.7 (40.4-83.4)	58.3 (27.0-80.1)	73.6 (68.9-77.7)	78.2 (71.7-83.3)	67.6 (61.9-72.6)	66.7 (40.4-83.4)	41.7 (15.2-66.5)
Congenital heart defects										
Common truncus	90.9 (88.1-93.1)	84.3 (77.5-89.2)	81.6 (76.2-85.9)	91.7 (53.9-98.8)	72.7 (37.1-90.3)	80.3 (76.7-83.5)	68.0 (59.9-74.7)	68.2 (61.9-73.6)	75.0 (40.8-91.2)	72.7 (37.1-90.3)
Transposition of great arteries	91.5 (90.3-92.6)	88.2 (85.3-90.6)	91.3 (89.5-92.8)	92.9 (87.5-96.0)	88.5 (68.4-96.1)	86.0 (84.6-87.4)	75.0 (71.2-78.4)	83.1 (80.7-85.1)	86.4 (79.9-90.9)	76.9 (55.7-88.9)
Tetralogy of Fallot	95.0 (94.1-95.7)	94.4 (92.7-95.8)	94.1 (92.7-95.3)	93.8 (89.5-96.3)	93.0 (79.9-97.7)	89.5 (88.3-90.6)	83.6 (81.1-85.9)	84.4 (82.3-86.3)	86.5 (81.1-90.5)	83.7 (68.9-91.9)
AVSD	92.9 (91.9-93.8)	90.4 (88.2-92.2)	89.4 (87.4-91.1)	88.9 (82.0-93.3)	95.7 (72.9-99.4)	84.1 (82.7-85.5)	76.4 (73.5-79.1)	74.1 (71.5-76.6)	76.2 (67.7-82.7)	60.9 (38.3-77.4)
AVSD (without Down syndrome)	87.6 (85.6-89.3)	84.6 (81.0-87.6)	84.0 (80.9-86.6)	83.3 (72.5-90.2)	93.3 (61.3-99.0)	75.6 (73.1-77.9)	67.9 (63.5-71.9)	64.9 (61.1-68.5)	68.1 (56.0-77.5)	60.0 (31.8-79.7)
Aortic valve stenosis	92.5 (91.2-93.7)	87.7 (82.6-91.3)	90.7 (88.3-92.7)	98.3 (88.6-99.8)	95.2 (70.7-99.3)	85.5 (83.7-87.1)	78.4 (72.5-83.2)	80.4 (77.2-83.2)	91.5 (80.8-96.4)	76.2 (51.9-89.3)
Hypoplastic left heart syndrome	74.0 (71.8-76.0)	73.6 (69.5-77.3)	72.2 (68.8-75.4)	60.0 (46.5-71.1)	58.8 (32.5-77.8)	57.8 (55.4-60.2)	51.5 (47.0-55.9)	52.2 (48.4-55.8)	50.0 (36.8-61.8)	41.2 (18.6-62.6)
Coarctation of aorta	92.9 (92.1-93.7)	89.7 (87.4-91.7)	92.8 (91.5-94.0)	88.7 (82.5-92.8)	92.3 (80.8-97.0)	86.3 (85.1-87.3)	77.2 (74.1-80.0)	84.4 (82.5-86.0)	80.1 (72.8-85.7)	76.9 (63.0-86.2)
Oral clefts										
Cleft palate without cleft lip	95.7 (95.1-96.3)	93.4 (91.5-95.0)	92.8 (91.6-93.9)	95.5 (92.2-97.4)	98.2 (88.2-99.8)	93.0 (92.2-93.7)	87.2 (84.7-89.4)	87.7 (86.1-89.1)	92.5 (88.6-95.1)	94.7 (84.6-98.3)
Cleft lip with or without cleft palate	95.2 (94.7-95.7)	90.3 (88.4-91.9)	92.5 (91.5-93.3)	97.0 (94.7-98.3)	92.4 (87.5-95.4)	93.5 (92.9-94.1)	84.7 (82.4-86.7)	89.7 (88.7-90.7)	94.9 (92.2-96.7)	91.3 (86.2-94.6)
Gastrointestinal defects										
Esophageal atresia/tracheoesophageal fistula	92.5 (91.2-93.6)	84.7 (80.5-88.0)	86.5 (83.9-88.8)	88.1 (77.5-93.8)	96.0 (74.8-99.4)	88.2 (86.7-89.6)	73.8 (68.9-78.0)	80.6 (77.6-83.2)	83.6 (72.3-90.6)	92.0 (71.6-97.9)
Pyloric stenosis	99.9 (99.9-100.0)	99.9 (99.5-100.0)	99.9 (99.8-100.0)	100	100	99.5 (99.4-99.6)	99.3 (98.8-99.6)	99.5 (99.3-99.6)	100	98.3 (93.5-99.6)
Rectal and large intestinal atresia/stenosis	93.3 (92.3-94.2)	89.7 (87.2-91.7)	87.5 (85.8-89.0)	88.1 (82.7-92.0)	84.6 (64.0-93.9)	90.3 (89.2-91.4)	83.4 (80.4-85.9)	82.7 (80.8-84.5)	87.1 (81.5-91.1)	84.6 (64.0-93.9)
Musculoskeletal defects										
Upper limb deficiencies	93.6 (92.4-94.6)	91.0 (88.3-93.2)	91.4 (89.5-92.9)	96.8 (90.4-99.0)	85.7 (69.0-93.8)	91.4 (90.1-92.6)	85.3 (82.0-88.1)	87.3 (85.2-89.2)	91.5 (83.7-95.7)	85.7 (69.0-93.8)
Lower limb deficiencies	92.9 (91.1-94.4)	94.4 (91.3-96.4)	87.5 (84.3-90.1)	87.2 (73.8-94.1)	81.0 (56.9-92.4)	90.7 (88.7-92.3)	89.6 (85.9-92.5)	84.0 (80.5-86.9)	87.2 (73.8-94.1)	81.0 (56.9-92.4)
Diaphragmatic hernia	76.6 (74.5-78.6)	69.7 (65.2-73.7)	78.5 (75.8-81.0)	78.3 (69.2-85.0)	78.6 (58.4-89.8)	70.6 (68.3-72.7)	59.5 (54.8-63.9)	70.2 (67.2-73.0)	69.8 (60.1-77.6)	67.9 (47.3-81.8)
Gastroschisis	95.9 (94.9-96.8)	93.3 (90.1-95.5)	96.5 (95.4-97.3)	95.2 (85.7-98.4)	91.2 (80.2-96.3)	93.0 (91.8-94.1)	89.5 (85.8-92.3)	93.5 (92.1-94.7)	90.3 (79.7-95.5)	89.5 (78.1-95.1)
Omphalocele	79.6 (76.3-82.5)	80.2 (73.9-85.1)	75.4 (70.8-79.4)	80.6 (61.9-90.8)	81.8 (44.7-95.1)	73.9 (70.3-77.1)	74.6 (67.9-80.1)	66.2 (61.2-70.7)	67.7 (48.4-81.2)	63.6 (29.7-84.5)
Chromosomal defects										

Birth defects *	Neonatal survival probability (<28 d)					Infant survival probability (<1 y)				
	NHW	NHB	Hispanic	A/PI	AI/AN	NHW	NHB	Hispanic	A/PI	AI/AN
Trisomy 21 (Down syndrome)	97.0 (96.6-97.3)	96.8 (95.9-97.5)	97.8 (97.4-98.2)	96.8 (94.8-98.0)	99.1 (93.6-99.9)	94.5 (94.0-95.0)	91.5 (90.2-92.7)	94.8 (94.2-95.4)	92.8 (90.1-94.7)	92.5 (85.6-96.2)

* Children with 2 or more birth defects may be counted in multiple categories.

Table IV

Survival probabilities and 95% CIs for children with selected birth defects by survival age (<2 years, ≤8 years), birth defect category, and maternal race/ ethnicity based on pooled data from 10 state birth defects surveillance programs,* National Birth Defects Prevention Network, 1999-2005

	Early childhood survival probability (<2 y)					Childhood survival probability (≤8 y)				
Birth defects†	**NHW**	**NHB**	**Hispanic**	**A/PI**	**AI/AN**	**NHW**	**NHB**	**Hispanic**	**A/PI**	**AI/AN**
Central nervous system defects										
Spina bifida without anencephalus	91.1 (89.4-92.5)	86.3 (82.1-89.5)	91.6 (89.6-93.2)	97.7 (84.9-99.7)	92.9 (74.3-98.2)	90.5 (88.8-91.9)	85.7 (81.5-89.0)	91.1 (89.0-92.7)	97.7 (84.9-99.7)	92.9 (74.3-98.2)
Encephalocele	73.6 (67.8-78.5)	77.3 (69.5-83.4)	65.5 (58.5-71.6)	45.5 (16.7-70.7)	30.0 (7.1-57.8)	72.8 (67.0-77.8)	76.6 (68.7-82.7)	65.5 (58.5-71.6)	45.5 (16.7-70.7)	30.0 (7.1-57.8)
Congenital heart defects										
Common truncus	77.4 (72.8-81.4)	69.3 (59.3-77.3)	63.2 (55.6-69.9)	71.4 (25.8-92.0)	71.4 (25.8-92.0)	76.4 (71.7-80.4)	68.3 (58.3-76.4)	63.2 (55.6-69.9)	71.4 (25.8-92.0)	57.1 (17.2-83.7)
Transposition of great arteries	85.0 (83.2-86.6)	70.3 (65.5-74.5)	81.8 (79.0-84.2)	82.6 (74.6-88.3)	72.2 (45.6-87.4)	84.2 (82.4-85.8)	68.8 (64.0-73.1)	80.9 (78.1-83.4)	79.3 (71.0-85.5)	72.2 (45.6-87.4)
Tetralogy of Fallot	88.7 (87.3-90.0)	81.9 (78.6-84.7)	82.0 (79.3-84.4)	83.7 (76.5-88.8)	77.4 (58.4-88.5)	87.8 (86.3-89.1)	81.4 (78.1-84.3)	81.5 (78.8-83.8)	80.1 (72.6-85.8)	77.4 (58.4-88.5)
AVSD	83.3 (81.5-84.8)	72.4 (68.6-75.8)	71.3 (68.1-74.2)	71.3 (61.0-79.3)	61.1 (35.3-79.2)	81.6 (79.8-83.2)	71.2 (67.4-74.7)	70.0 (66.7-73.0)	69.1 (58.7-77.4)	61.1 (35.3-79.2)
AVSD (without Down syndrome)	74.9 (72.0-77.6)	63.8 (58.5-68.7)	62.1 (57.5-66.3)	63.2 (49.3-74.2)	54.5 (22.9-78.0)	73.1 (70.1-75.8)	62.6 (57.3-67.5)	59.9 (55.3-64.2)	59.6 (45.8-71.0)	54.5 (22.9-78.0)
Aortic valve stenosis	84.8 (82.6-86.7)	77.3 (69.8-83.2)	78.2 (74.3-81.5)	88.4 (74.3-95.0)	80.0 (50.0-93.1)	83.9 (81.7-85.8)	76.7 (69.0-82.6)	76.8 (72.9-80.2)	86.0 (71.6-93.5)	80.0 (50.0-93.1)
Hypoplastic left heart syndrome	56.1 (53.3-58.9)	48.4 (43.1-53.5)	48.8 (44.2-53.3)	47.4 (31.0-62.1)	30.0 (7.1-57.8)	54.0 (51.1-56.8)	45.8 (40.5-51.0)	46.5 (41.9-50.9)	42.1 (26.4-57.0)	30.0 (7.1-57.8)
Coarctation of aorta	84.7 (83.2-86.0)	74.8 (70.9-78.3)	82.0 (79.6-84.0)	79.4 (70.5-85.9)	72.2 (54.5-84.0)	84.0 (82.5-85.3)	74.1 (70.1-77.6)	80.8 (78.4-82.9)	78.5 (69.5-85.2)	72.2 (54.5-84.0)
Oral clefts										
Cleft palate without cleft lip	92.7 (91.7-93.6)	87.5 (84.5-90.0)	87.7 (85.8-89.4)	90.8 (85.6-94.2)	100	92.3 (91.3-93.2)	87.0 (83.9-89.5)	87.0 (85.0-88.7)	90.8 (85.6-94.2)	93.3 (80.7-97.8)
Cleft lip with or without cleft palate	93.2 (92.4-93.9)	84.9 (82.1-87.3)	89.1 (87.8-90.3)	94.6 (91.3-96.7)	90.8 (84.7-94.5)	92.8 (92.0-93.5)	84.0 (81.1-86.4)	88.6 (87.3-89.8)	94.6 (91.3-96.7)	90.8 (84.7-94.5)
Gastrointestinal defects										
Esophageal atresia/tracheoesophageal fistula	87.3 (85.3-89.0)	75.0 (69.1-80.0)	82.0 (78.5-85.0)	82.6 (68.2-90.9)	84.2 (58.7-94.6)	86.7 (84.8-88.5)	73.8 (67.8-78.8)	81.5 (77.9-84.5)	82.6 (68.2-90.9)	84.2 (58.7-94.6)
Pyloric stenosis	99.4 (99.3-99.6)	99.4 (98.7-99.7)	99.3 (99.0-99.5)	100	99.0 (93.0-99.9)	99.3 (99.2-99.5)	99.4 (98.7-99.7)	99.2 (98.9-99.4)	100	99.0 (93.0-99.9)
Rectal and large intestinal atresia/stenosis	90.4 (89.0-91.6)	81.8 (78.0-84.9)	82.2 (79.9-84.3)	86.5 (79.7-91.2)	85.0 (60.4-94.9)	90.1 (88.7-91.3)	81.1 (77.4-84.3)	81.7 (79.3-83.8)	85.8 (78.9-90.6)	75.0 (50.0-88.7)
Musculoskeletal defects										
Upper limb deficiencies	90.6 (88.8-92.1)	85.9 (81.8-89.1)	86.6 (84.1-88.8)	95.1 (85.5-98.4)	84.6 (64.0-93.9)	89.9 (88.1-91.4)	85.6 (81.5-88.9)	86.6 (84.1-88.8)	95.1 (85.5-98.4)	80.8 (59.8-91.5)
Lower limb deficiencies	91.3 (88.9-93.2)	89.8 (85.2-93.0)	83.2 (79.1-86.6)	90.0 (72.1-96.7)	84.6 (51.2-95.9)	91.0 (88.6-92.9)	88.9 (84.2-92.3)	82.9 (78.8-86.4)	90.0 (72.1-96.7)	76.9 (44.2-91.9)
Diaphragmatic hernia	70.2 (67.4-72.9)	58.2 (52.3-63.6)	69.8 (66.1-73.1)	71.8 (59.8-80.8)	62.5 (40.3-78.4)	70.2 (67.4-72.8)	57.2 (51.3-62.6)	69.8 (66.1-73.1)	70.4 (58.3-79.6)	62.5 (40.3-78.4)
Gastroschisis	92.7 (91.0-94.1)	89.9 (84.6-93.4)	92.7 (90.8-94.2)	90.5 (76.6-96.3)	89.5 (74.3-95.9)	92.5 (90.8-93.9)	89.9 (84.6-93.4)	92.4 (90.5-94.0)	90.5 (76.6-96.3)	89.5 (74.3-95.9)
Omphalocele	73.2 (68.6-77.2)	75.4 (67.0-81.9)	67.2 (61.2-72.5)	76.2 (51.9-89.3)	83.3 (27.3-97.5)	73.2 (68.6-77.2)	75.4 (67.0-81.9)	66.4 (60.4-71.7)	76.2 (51.9-89.3)	83.3 (27.3-97.5)
Chromosomal defects										

Birth defects†	Early childhood survival probability (<2 y)					Childhood survival probability (≤8 y)				
	NHW	NHB	Hispanic	A/PI	AI/AN	NHW	NHB	Hispanic	A/PI	AI/AN
Trisomy 21 (Down syndrome)	93.9 (93.3-94.5)	90.6 (88.8-92.0)	94.1 (93.2-94.8)	92.0 (88.4-94.4)	89.7 (80.5-94.7)	93.2 (92.5-93.8)	89.8 (88.0-91.3)	93.7 (92.8-94.5)	91.3 (87.7-93.9)	89.7 (80.5-94.7)

*Illinois and Nebraska were excluded from these analyses because vital status data beyond infancy were not available.

†Children with 2 or more birth defects may be counted in multiple categories.

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Table V

Adjusted* HRs for children with selected birth defects by survival age, birth defect category and maternal race/ethnicity based on pooled data from 12 state birth defects surveillance programs, National Birth Defects Prevention Network, 1999-2007

	Neonatal period (<28 d)‡				Postneonatal infancy period (28 d to <1 y)‡				Childhood (1 to ≤8 y)§				Overall childhood (≤8 y)§			
Birth defects†	**NHB**	**Hispanic**	**A/PI**	**AI/AN**	**NHB**	**Hispanic**	**A/PI**	**AI/AN**	**NHB**	**Hispanic**	**A/PI**	**AI/AN**	**NHB**	**Hispanic**	**A/PI**	**AI/AN**
Central nervous system defects																
Spina bifida without anencephalus	0.9	0.8	0.2	0.8	1.6	1.2	0.0	1.2	3.4¶	0.9	0.0	0.0	1.3	1.0	0.2	0.7
Encephalocele	0.7	1.2	1.7	1.9	1.2	2.0	0.0	4.8¶	1.0	0.7	0.0	0.0	0.7	1.3	2.6¶	2.8¶
Congenital heart defects																
Common truncus	1.5	1.7¶	0.9	3.3	1.4	1.2	1.5	0.0	1.3	0.7	0.0	19.7¶	1.2	1.6¶	1.1	2.0
Transposition of great arteries	1.0	0.9	0.8	1.0	2.1¶	1.5¶	1.1	2.3	2.6¶	1.4	3.6¶	3.8	1.6¶	1.2	1.3	1.5
Tetralogy of Fallot	1.0	1.1	1.1	1.3	1.8¶	1.7¶	1.2	1.4	1.2	1.1	2.4¶	1.1	1.4¶	1.4¶	1.6¶	1.3
AVSD	1.2	1.2	1.6	0.4	1.6¶	1.7¶	1.6	3.5¶	1.5	1.4	2.1	0.0	1.5¶	1.5¶	1.8¶	1.8
AVSD (without Down syndrome)	1.0	1.0	1.5	0.4	1.3¶	1.6¶	1.4	2.4	1.3	1.5	2.1	0.0	1.3¶	1.3¶	1.6¶	1.5
Aortic valve stenosis	1.5	1.1	0.2	0.3	1.2	1.3	0.8	2.1	3.3¶	2.0	2.0	0.0	1.5¶	1.3	0.7	0.8
Hypoplastic left heart syndrome	0.9	1.0	1.6¶	1.3	1.3¶	1.3¶	0.8	1.3	2.0¶	1.3	2.8	0.0	1.1	1.1	1.4	1.6
Coarctation of aorta	1.3	0.9	1.4	0.9	1.8¶	1.2	1.2	2.0	2.3¶	1.6	1.4	1.7	1.5¶	1.1	1.3	1.3
Oral clefts																
Cleft palate without cleft lip	1.2	1.4¶	1.0	0.3	1.9¶	1.7¶	1.0	0.8	0.6	1.4	0.0	2.8	1.4¶	1.4¶	1.1	0.5
Cleft lip with or without cleft palate	1.2	1.2¶	0.5¶	1.2	2.4¶	1.5¶	1.0	0.6	1.7	1.2	0.4	2.1	1.3¶	1.3¶	0.6¶	1.1
Gastrointestinal defects																
Esophageal atresia/tracheoesophageal fistula	1.9¶	1.6¶	1.6	0.5	2.8¶	1.5	1.1	0.9	1.2	0.7	0.0	3.4	2.0¶	1.4¶	1.3	1.2
Pyloric stenosis	1.7	1.1	0.0	0.0	1.2	1.3	0.0	3.6	0.0	0.8	0.0	0.0	1.0	1.4	0.0	1.2
Rectal and large intestinal atresia/stenosis	1.1	1.3¶	1.4	1.2	1.6¶	1.5¶	0.3	0.0	1.9	1.6	1.0	9.0¶	1.4¶	1.4¶	1.2	1.5
Musculoskeletal defects																

Birth defects[†]	Neonatal period (<28 d)[‡]				Postneonatal infancy period (28 d to <1 y)[‡]				Childhood (1 to ≤8 y)[§]				Overall childhood (≤8 y)[§]			
	NHB	Hispanic	A/PI	AI/AN	NHB	Hispanic	A/PI	AI/AN	NHB	Hispanic	A/PI	AI/AN	NHB	Hispanic	A/PI	AI/AN
Upper limb deficiencies	1.0	1.1	0.4	1.2	2.1[¶]	1.7[¶]	2.3	0.0	0.9	0.9	0.0	3.7	1.1	1.1	0.5	1.5
Lower limb deficiencies	0.7	1.2	1.6	1.2	1.6	1.5	0.0	0.0	4.5[¶]	0.9	0.0	7.6	1.0	1.3	1.2	1.1
Diaphragmatic hernia	1.2	0.8[¶]	0.8	0.6	1.7[¶]	1.4[¶]	1.3	1.4	3.5[¶]	0.7	1.3	20.4[¶]	1.4[¶]	0.9	0.8	0.8
Gastroschisis	1.5	0.9	0.9	2.3	1.2	1.1	1.7	0.7	1.4	1.5	0.0	0.0	1.2	1.1	1.2	1.7
Omphalocele	1.0	1.0	0.8	0.8	1.1	1.5	2.0	4.1	1.0	1.6	0.0	0.0	0.9	1.1	0.7	0.8
Chromosomal defects																
Trisomy 21 (Down syndrome)	1.0	0.8	0.9	0.3	1.9[¶]	1.2	1.5	2.5[¶]	1.9[¶]	0.8	1.4	0.8	1.4[¶]	1.0	1.1	1.4

[*] Adjusted for: birth weight and gestational age, maternal age, birth period, and state surveillance program; NHW was used as the reference group.

[†] Children with 2 or more birth defects may be counted in multiple categories.

[‡] Children born in 1999-2007 from all 12 states.

[§] Children born in 1999-2005 from 10 states; data from Illinois and Nebraska were excluded because no vital status data beyond infancy were available.

[¶] Statistically significant; $P < .05$.

Table I

Summary of the study cohort (children with selected birth defects) by participating state birth defects surveillance programs and maternal race/ethnicity, National Birth Defects Prevention Network, 1999-2007

					Number of live births, total deaths and infant deaths* and childhood deaths† by race/ethnicity														
		Total‡			NHW			NHB			Hispanic			A/PI			AI/AN		
State	Birth cohort years§	Live births	Total deaths	Infant deaths	Live births	Infant deaths	Child-hood deaths	Live births	Infant deaths	Child-hood deaths	Live births	Infant deaths	Child-hood deaths	Live births	Infant deaths	Child-hood deaths	Live births	Infant deaths	Child-hood deaths
Arizona	1999-2007	5526	726	632	2334	252	234	130	22	20	2413	284	251	120	18	13	410	52	48
Colorado	1999-2006	4992	512	455	2983	271	252	156	30	27	1496	140	137	109	8	7	33	6	¶
Florida	1999-2007	16 745	1444	1202	9494	555	493	2883	351	307	3952	257	209	262	25	23	43	¶	¶
Georgia**	1999-2007	3190	331	303	1420	99	83	1003	135	120	571	50	50	113	10	10	6	¶	¶
Illinois	1999-2006	7039	938	938	4038	492	-	1121	212	-	1562	191	-	283	42	-	9	0	-
Massachusetts	2000-2007	3222	246	223	2288	129	104	228	35	28	374	38	30	163	9	6	12	0	0
Michigan	1999-2006	9071	861	724	6845	455	463	1314	138	145	529	54	56	222	13	18	37	¶	¶
North Carolina	2003-2007	4978	630	565	3059	292	204	880	138	80	831	101	61	90	19	10	117	15	8
Nebraska	1999-2006	1750	95	95	1408	76	-	71	¶	-	201	8	-	31	¶	-	26	¶	-
New Jersey	1999-2005	5006	442	406	2699	161	172	689	94	110	1253	109	115	309	34	36	¶	0	0
New York††	1999-2007	9429	796	705	7168	513	472	756	87	76	1142	81	69	217	17	18	48	¶	¶
Texas	1999-2007	27 885	2976	2645	10 783	870	761	2132	330	271	14 173	1375	1126	605	52	49	67	6	¶
Total	1999-2007	98 833	9997	8893	54 519	4165	3238	11 363	1575	1184	28 497	2688	2104	2524	249	190	811	91	74

*The number of live births, total deaths (≤8 years) and infant deaths (<1 year) were determined using the cohort including all children with birth defects born in 1999-2007 from 12 states.

†The number of childhood deaths (infant deaths plus deaths beyond infancy; ≤8 years) was determined using the cohort including children with birth defects born in 1999-2005 from 10 states; data from Illinois and Nebraska were excluded because there were no vital status data available beyond infancy.

‡The total includes the “other” racial/ethnic group.

§The birth cohort years vary by state because of the availability of the birth defect data and vital status of the cases.

¶Data were suppressed when the number of observations was ≤5.

**Georgia includes 5 counties of metropolitan Atlanta.

††New York State excludes New York City.