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Racial and ethnic differences in infant survival for hydrocephaly —Texas, 1999–2017

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Abstract

Background: Congenital hydrocephaly, an abnormal accumulation of fluid within the ventricular spaces at birth, can cause disability or death if untreated. Limited information is available about survival of infants born with hydrocephaly in Texas. Therefore, the purpose of the study was to calculate survival estimates among infants born with hydrocephaly without spina bifida in Texas.

Methods: A cohort of live-born infants delivered during 1999–2017 with congenital hydrocephaly without spina bifida was identified from the Texas Birth Defects Registry. Deaths within 1 year of delivery were identified using vital and medical records. One-year infant survival estimates were generated for multiple descriptive characteristics using the Kaplan–Meier method. Crude hazard ratios (HRs) for one-year survival among infants with congenital hydrocephaly by maternal and infant characteristics and adjusted HRs for maternal race and ethnicity were estimated using Cox proportional hazard models.

Results: Among 5709 infants born with congenital hydrocephaly without spina bifida, 4681 (82%) survived the first year. The following characteristics were associated with infant survival: maternal race and ethnicity, clinical classification (e.g., chromosomal or syndromic), preterm birth, birth weight, birth year, and maternal education. In the multivariable Cox proportional hazards model, differences in survival were observed by maternal race and ethnicity after adjustment for other maternal and infant characteristics. Infants of non-Hispanic Black (HR: 1.28, 95% CI:

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SUPPORTING INFORMATION

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1.04–1.58) and Hispanic (HR: 1.31, 95% CI: 1.12–1.54) women had increased risk for mortality, compared with infants of non-Hispanic White women.

Conclusions: This study showed infant survival among a Texas cohort differed by maternal race and ethnicity, clinical classification, gestational age, birth weight, birth year, and maternal education in infants with congenital hydrocephaly without spina bifida. Findings confirm that mortality continues to be common among infants with hydrocephaly without spina bifida. Additional research is needed to identify other risk factors of mortality risk.

Keywords

birth defects; epidemiology; hydrocephaly; infant; race and ethnicity; survival

1 | INTRODUCTION

Congenital hydrocephaly is characterized by excessive fluid accumulation, resulting in abnormally dilated cerebral ventricles at birth (Isaacs et al., 2018). Although congenital hydrocephaly is frequently present among infants with spina bifida, when it occurs without spina bifida the etiologies are different (Gili et al., 2022; Isaacs et al., 2018). Therefore, this study focused on congenital hydrocephaly without spina bifida (hereafter referred to as hydrocephaly, except where otherwise indicated). If left untreated, hydrocephaly leads to chronic morbidity and can be fatal (Gili et al., 2022; Isaacs et al., 2018). Prevalence of hydrocephaly differs worldwide (Isaacs et al., 2018). Total birth prevalence of hydrocephaly in North America is estimated to be approximately 5–6 cases/10,000 live births (Gili et al., 2022; Isaacs et al., 2018). In Texas, prevalence of hydrocephaly appears to be higher than national levels and is estimated to be 8 cases/10,000 live births (Texas Birth Defects Registry, n.d.).

Prior epidemiologic studies (Acakpo-Satchivi et al., 2008; Casey et al., 1997; Fernell et al., 1994; Iskandar et al., 1998; Persson et al., 2005; Vinchon et al., 2012) have reported survival estimates for hydrocephaly based on limited, clinic-based populations (e.g., a single hospital), which are subject to selection bias. A limited number of published population-based studies are available regarding hydrocephaly mortality or survival. In a recent international study, 6% of neonates with hydrocephaly died within 7 days, and infants with syndromic hydrocephaly had approximately a two times higher risk for death (Gili et al., 2022). However, the study did not assess mortality beyond 7 days. Two older studies conducted in the United States provided state-based estimates for survival among infants with hydrocephaly (Nembhard et al., 2001; Wang et al., 2011). In one study investigators reported 75% survival (one-year period) among infants born with hydrocephaly in Texas in 1995–1997. These results were before full implementation of a statewide active ascertainment registry in 1999 (Nembhard et al., 2001). Another study examined data from a passive ascertainment system in New York State. The study showed an overall 25-year survival of 83% among infants with hydrocephaly (Wang et al., 2011). Studies also report that infant survival is influenced by maternal and infant-associated risk factors, such as maternal race and ethnicity, preterm birth, low birth weight, and maternal age (Ely & Driscoll, 2019; Gili et al., 2022; Ratnasiri et al., 2020; Shi et al., 2004), including improvements in care of preterm infants in more recent years (Glinianaia et al., 2020).

However, a fuller understanding of mortality risk factors is needed using current information on survival estimates.

The objectives of this study were to (1) describe population-based survival in a statewide cohort of infants born with hydrocephaly, through their first year of life, and (2) to identify independent maternal and infant characteristics that influence survival among these infants, including maternal race and ethnicity.

2 | METHODS

2.1 | Data

The Texas Department of State Health Services (DSHS) Texas Birth Defects Registry (TBDR) was established in 1993 and became statewide in 1999. TBDR is a large, active-ascertainment surveillance system. It includes diagnoses and relevant clinical information for all major birth defects documented within the first year of life and infant and maternal demographic characteristics. Methods for identifying structural birth defects and chromosomal anomalies within the first year of life have been documented (Anderka et al., 2015; Miller, 2006). Briefly, trained program staff routinely visit medical facilities in person or access the medical records online, to review medical logs (e.g., hospital unit logs and discharge listings) and other records to identify potential infants with birth defects. If an eligible birth defect is identified during pregnancy, at delivery, or during the child's first year of life, and the mother resided in Texas at delivery, the relevant medical records are abstracted.

TBDR is routinely linked to state vital event data managed by the Center for Health Statistics at DSHS. These data are derived from birth, death, and fetal death certificates, and linkage is performed using a unique identifier (i.e., birth certificate number). Aside from vital status and live birth denominators, other important information from vital records includes gestational age, maternal education, maternal residence, and supplemental demographic data.

This study used data on live-born infants delivered by women residing in Texas from 1999 (the first year the TBDR covered the entire state of Texas) to 2017. Infants with hydrocephaly were identified primarily using codes from the *International Classification of Diseases Clinical Modification* (ICD-9-CM) and the *International Classification of Diseases Clinical Modification* (ICD-10-CM). Infants meeting the case definition were then abstracted using modified specific corresponding codes from the British Pediatric Association Classification of Diseases: 742.300, 742.310, 742,320, 742.380, and 742.390. Because congenital hydrocephaly is frequently present among infants with spina bifida and its etiology is similar to hydrocephaly, infants with spina bifida were excluded from this analysis.

2.2 | Outcome and independent variables

The main outcome variable was survival time for infants born with hydrocephaly. This was calculated in days from birth until the death of an infant or until the infant was aged 1 year, with censoring at 1 year. Based on prior work (Benjamin et al., 2021, 2023; Marengo et

al., 2014, 2023; Martin-Giacalone et al., 2023; Vendola et al., 2010), infants without death records were assumed to have survived to their first birthday. Several independent variables were considered, including maternal race and ethnicity (non-Hispanic White [White], non-Hispanic Black [Black], Hispanic, and additional groups), maternal age at delivery (<35 years or ≥ 35 years), maternal education at delivery (less than high school [<12 years] high school [12 years], and more than high school [>12 years]), preterm birth (gestational age < 37 weeks or ≥ 37 weeks at delivery), birthweight (<2500 grams or ≥ 2500 grams), clinical classification for infants with hydrocephaly (isolated, chromosomal or syndromic, and multiple), and birth year (1999–2011 or 2012–2017) (Supplement A). The clinical classification of hydrocephaly was categorized as isolated cases (infants with hydrocephaly and no other birth defects), chromosomal or syndromic (infants with hydrocephaly and chromosomal or syndromic birth defects) and multiple (infants with hydrocephaly and other major birth defects other than chromosomal and syndromic) (Benjamin et al., 2022; Langlois et al., 2023). The “additional groups” race and ethnicity category included non-Hispanic Asian, non-Hispanic Native Hawaiian and Pacific Islander, non-Hispanic American Indian and Alaskan Native, and non-Hispanic with ≥2 reported races. Birth year was dichotomized before and after 2012 to specifically characterize survival in recent years.

2.3 | Statistical analysis

Counts and percentages were tabulated to summarize distribution of infants with hydrocephaly by maternal and infant characteristics. The Kaplan–Meier method was used to estimate infant survival probability and to generate survival curves for hydrocephaly overall. Analyses were also stratified by maternal race and ethnicity, maternal education, maternal age, hydrocephaly classification type, preterm birth, birth weight, and birth year.

Unadjusted hazard ratios were computed using univariable Cox proportional hazards models.

Subsequently Cox proportional hazards models were used for multivariable analysis to estimate the mortality hazard ratios (HRs) for infants with hydrocephaly by maternal race and ethnicity adjusted for all other maternal and infant characteristics identified during literature review. All variables were included in a simultaneous Cox proportional hazards regression model to obtain adjusted HRs. A simultaneous regression approach was employed because no theoretical basis exists for considering one variable over another in terms of relevance to study objectives (Harrell, 2001). Schoenfeld residuals were used to test proportional hazards assumptions, and assumptions were met for main variable of interest and other covariates with exception of gestational age and less than high school category of maternal education (Supplement B). All analyses were carried out using SAS® version 9.4 (SAS Institute Inc., Cary, North Carolina, USA).

This project was approved by the DSHS Institutional Review Board. This project was reviewed by the Centers for Disease Prevention and Prevention (CDC) and was conducted consistent with applicable federal law and CDC policy (e.g., 45 C.F.R. part 46.102(l)(2), 21 C.F.R. part 56; 42 U.S.C. §241(d); 5 U.S.C. §552a; 44 U.S.C. §3501 et seq.).

3 | RESULTS

In total, 5709 infants with hydrocephaly were analyzed for delivery years 1999–2017 (Table 1). In terms of maternal race and ethnicity, 34.78% of mothers of infants were White, 13.37% were Black, 48.48% were Hispanic, and 3.37% were from additional groups. Based on clinical classification, 15.76% of infants had chromosomal or syndromic, 26.69% isolated (no co-occurring birth defects), and 53.56% multiple forms of hydrocephaly. Among infants analyzed, 42.53% were preterm (<37 weeks of gestation) and 41.01% were low birth weight (<2500 grams). In total, 62.67% of infants with hydrocephaly were born before 2012. Distribution of infants born based on maternal education was 28.51%, 30.68%, and 40.81% for <12 years, 12 years, and > 12 years of education, respectively.

Overall, infant survival for hydrocephaly was 82.83% (95% CI: 0.82–0.84) (Table 2, Figure 1). The lowest infant survival rates were observed among infants with hydrocephaly classified as chromosomal and syndromic (66.33%; 95% CI: 63.14–69.32), preterm births (78.91%; 95% CI:77.24–80.48), and low birth weight (76.29%; 95% CI: 74.52–77.96). The highest survival rates were observed among infants with isolated hydrocephaly (92.45%; 95% CI:91.01–93.67). The survival patterns in Kaplan–Meier curve showed the greatest decrease in survival occurring during neonatal period (within the first month), followed by a leveling of the survival estimates in later months (Figure 1).

In the unadjusted Cox proportional hazard model, the following maternal and infant characteristics had an increased risk of infant mortality: maternal race and ethnicity, clinical classification, gestational age, birth weight, birth year period, and maternal education (Table 3). In the multivariable Cox proportional hazards model, racial and ethnic differences in infant survival were observed after adjustment for all maternal and infant characteristics. Infants of Black (HR:1.28, 95% CI: 1.04–1.58) and Hispanic (HR:1.31, 95% CI: 1.12–1.54) women were at approximately 30% increased relative risk for death, compared with White women (Table 4).

4 | DISCUSSION

This study provides contemporary population-based infant survival estimates for hydrocephaly in Texas from an active ascertainment birth defects surveillance system. Overall survival estimates of 83% appears to be higher than the 75% estimate observed in a previous study conducted in Texas during the 1990s. The difference between the two studies might be the result of the earlier data being from an incomplete surveillance system and with relatively limited frequencies. However, this estimate seems consistent with the survival estimate (83%) provided in a 2011 New York study (1983–2006), based on passively reported data (Nembhard et al., 2001; Wang et al., 2011). Compared with population of all infants born in Texas, the excess mortality among infants with hydrocephaly is considerable (Nembhard et al., 2001).

Studies have shown that mortality is higher among infants and children with birth defects (Agha et al., 2006; Copeland & Kirby, 2007; Kassebaum et al., 2017), compared with those in the general population. An analysis of National Vital Statistics System and the National

Linked Birth and Death data from 1915 to 2017 revealed racial and ethnic, socioeconomic, and geographic disparities in infant mortality in the United States (Singh & Stella, 2019). Factors, including maternal race and ethnicity, maternal age, maternal education, gestational age, and birth weight were shown to be associated with mortality and survival among infants with or without birth defects (Almli et al., 2020; Lopez et al., 2018; Nembhard et al., 2010; Oster et al., 2013; Pace et al., 2018; Yang et al., 2006). Early diagnosis and treatment of birth defects such as hydrocephaly and an improved understanding of potential risk factors is crucial for infant survival.

This study provided statewide, population-based survival estimates for infants with hydrocephaly stratified by multiple factors, including maternal race and ethnicity. After accounting for multiple variables, adjusted HRs suggested that infants with hydrocephaly born to Black and Hispanic mothers are approximately 30% more likely to die during infancy, compared with infants born to White mothers. These results appear consistent with typical birth defects survival patterns (Glinianaia et al., 2020). Associations with maternal race and ethnicity might represent the role of many complex factors not included in this study. Although racial and ethnic differences in low birth weight, prematurity, and respiratory distress syndrome are hypothesized to play a role in birth defect mortality (Centers for Disease Control and Prevention [CDC], 1990), the current analyses accounted only for low birth weight and preterm birth. Given the complex landscape of factors associated with maternal race and ethnicity, to reduce infant mortality from birth defects, further work is needed to understand discrepancies that lie within social determinants associated with adverse infant outcomes.

In this study, unadjusted results showed infants with chromosomal or syndromic hydrocephaly had approximately five-times higher hazard of death, compared with infants born with isolated hydrocephaly. This finding is consistent with those from an international study concerning early neonatal hydrocephaly mortality that reported case fatality rates approximately 2.7 times higher among infants with syndromic versus nonsyndromic hydrocephaly (Gili et al., 2022). A previous Texas study also reported 88%, 63%, and 25% survival for infants with hydrocephaly with zero, one, and two co-occurring “life-threatening” defects, respectively, but did not focus on chromosomal or syndromic defects specifically (Nembhard et al., 2001).

Similarly, the unadjusted results showed low birthweight (<2500 grams) was significantly associated with higher mortality. Further, unadjusted preterm birth was associated with infant mortality among this population. Gestational age and birthweight play a crucial role in survival among infants without birth defects and might also be important among those with birth defects. For instance, Benjamin et al. reported that preterm and low birth weight contributed to 10% of deaths among all infants with birth defects in Texas (Benjamin et al., 2021). Similarly, a national-level analysis of linked birth defects and infant death records for infants born to U.S. residents during 2002–2017 reported increased mortality rates among preterm infants with birth defects born at 32–36 weeks (Almli et al., 2020). Prematurity and low-birth weight pose increased health concerns because of conditions such as premature lungs, difficulty with feeding and risk for infection, which might influence mortality hazard in infants with existing hydrocephaly.

Based on unadjusted results, the current study also reported that over the 19 years of the study's cohort (from 1999 to 2017), infants born before 2012 were at an increased risk for mortality (16% higher), compared with those born in years after 2012. These findings suggest some improvement in clinical care and wraparound services in terms of timely identification and referrals. Examples of potential improvement in prenatal and neonatal clinical care over time include advances in prenatal diagnosis, neonatal care (including intensive care, standard use of antenatal steroids, and surfactant therapy for prevention of neonatal mortality and morbidity in preterm births), early surgical interventions, extracorporeal membrane oxygenation, care centralization, and treatment and transplantation of certain organs or body systems (for those with co-occurring defects) (Glinianaia et al., 2020). Similarly, results of a meta-analyses showed statistically significant improvement in survival over time among infants with spina bifida, esophageal atresia, biliary atresia, congenital heart defects (CHD), gastroschisis, and Down syndrome with CHD, but not with encephalocele, biliary atresia with a native liver or Down syndrome without CHD (Glinianaia et al., 2020).

Strengths of this study include new information for an under published topic, regarding the survival of infants with hydrocephaly in Texas. This analysis used statewide data collected through a large public health surveillance system with active ascertainment of birth defects, accounting for approximately 1 in 10 annual births in the United States (Agha et al., 2006). Finally, this study used a substantial sample size, resulting in relatively precise estimates for survival estimates and HRs.

Study limitations include reliance on state-level vital records, although our prior work suggests that only a limited number of infant deaths might occur out of state for infants with birth defects born in Texas (Nembhard et al., 2001). Assumptions about proportionality of hazards held for main variable of interest (race/ethnicity) and other covariates with exception of gestational age and less than high school category of maternal education. This analysis included some infants who might have had hydrocephaly that developed early in infancy rather than in utero. However, hydrocephaly that was likely secondary to another select documented anomaly was excluded from the analysis. This study did not conduct separate analyses for infant survival and for each confounding maternal and infant characteristics, which might have addressed bias introduced by intermediate variables in the causal pathway. Additional clinical variables that were not available (e.g., full diagnostic details, treatment strategies, and other clinical information) might shed additional light on mortality risk in future studies. In this analysis racial and ethnic groups other than NH White, NH Black, and Hispanic were collapsed into one additional groups category due to small sizes. Therefore, this may not be a full examination of all racial/ethnic differences in infant survival by maternal race/ethnicity. Although research on causal pathways has increased, most cases of hydrocephaly remain genetically unexplained from a clinical diagnosis and treatment standpoint and can play a role in infant survival (Allington et al., 2021; Tully & Dobyns, 2014). Therefore, results of this study should be interpreted cautiously.

5 | CONCLUSIONS

This study confirms that mortality continues to be common among infants with hydrocephaly in a Texas cohort, which highlights the need for new avenues to work toward identification of modifiable mortality risk factors among affected infants. As a next step in this direction, the study identified racial and ethnic differences in infant survival for hydrocephaly and potential differences by clinical classification, gestational age, birth weight, birth year, and maternal education. Some of these potential associations might identify candidate risk factors that can be considered for future hypothesis-testing analytic approaches, such as time analyses. Such further research is expected to ultimately help elucidate the underlying causal pathways involved and might translate to clinical strategies for improving infant survival for hydrocephaly.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

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

The data that supported this study are available upon application to the Texas Birth Defects Registry and Texas Department of State Health Services Institutional Review Board.

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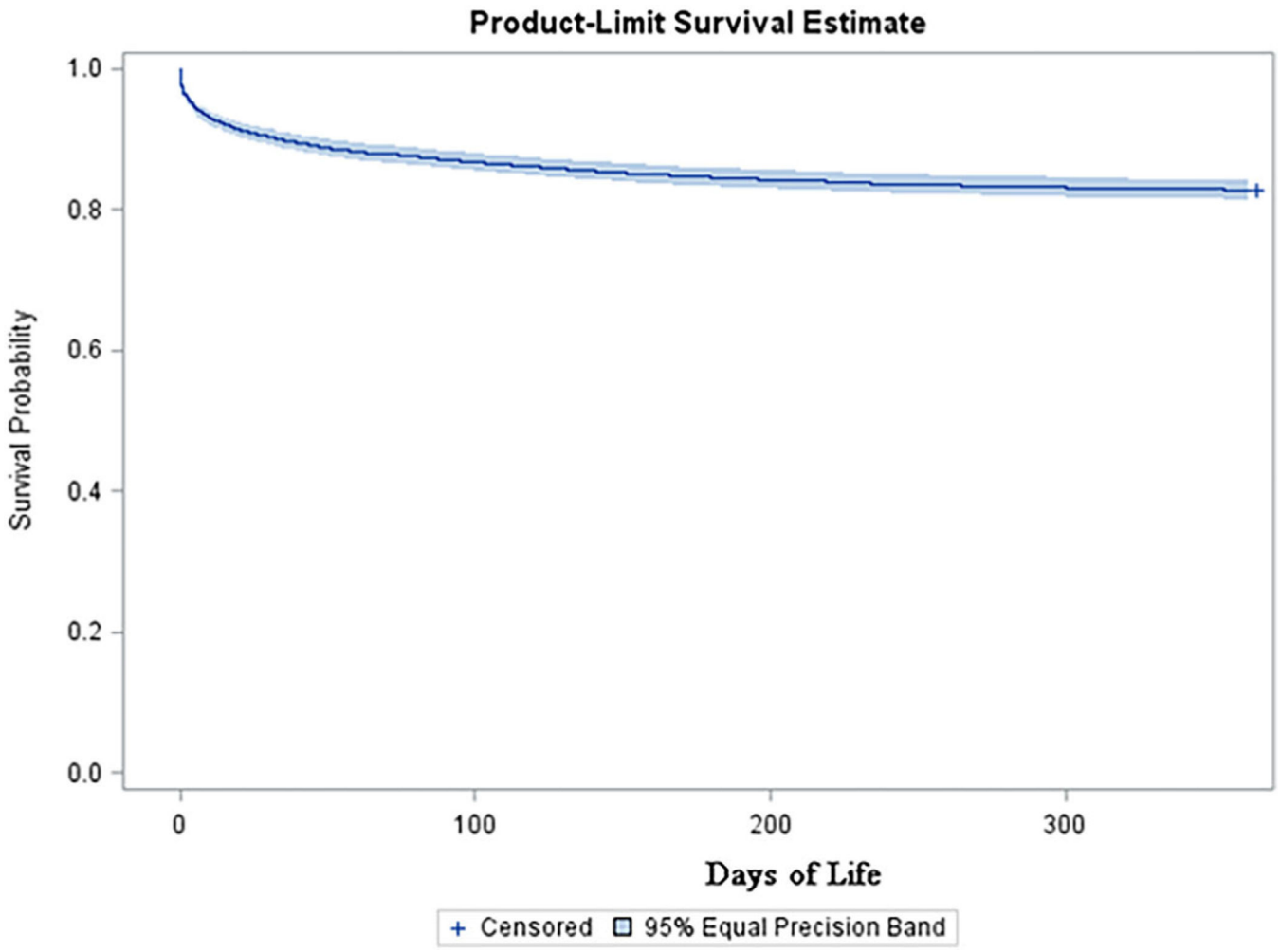

FIGURE 1.

One-year Kaplan–Meier survival estimates for infants born with hydrocephaly without spina bifida, Texas, 1999–2017. Modeled one-year survival estimates among infants born with hydrocephaly with censoring at 364 days.

TABLE 1

Distribution of infants born with hydrocephaly without spina bifida and selected cofactors, stratified by classification category, Texas, 1999–2017[a] (*N* = 5709).

Variable	Categories	Frequency	Percent
Maternal race and ethnicity	Non-Hispanic White	1983	34.78
	Non-Hispanic Black	762	13.37
	Hispanic	2764	48.48
	Additional groups[b]	192	3.37
Classification	Chromosomal and syndromic	900	15.76
	Isolated	1524	26.69
	Multiple	3058	53.56
Gestational age	Preterm <37 weeks	2428	42.53
	Term ≥37 weeks	3281	57.47
Birth weight	Low birth weight < 2500 grams	2341	41.01
	Normal birth weight ≥ 2500 grams	3368	58.99
Birth year	<2012	3578	62.67
	≥2012	2131	37.33
Maternal education	<12 years	1579	28.51
	12 years	1699	30.68
	≥12 years	2260	40.81
Maternal age	<35 years	4890	85.65
	≥35 years	819	14.35

[a] Only live-born infants with hydrocephaly without spina bifida were included in the analysis. Not all maternal and infant characteristics categories add up to the total because of missing data.

[b] Additional Groups race and ethnicity category included non-Hispanic Asian, non-Hispanic Native Hawaiian and Pacific Islander, non-Hispanic American Indian and Alaskan Native, and non-Hispanic two or more races.

TABLE 2

Survival estimates (%, with 95 CI) for infants born with hydrocephaly without spina bifida and selected cofactors, Texas, 1999–2017 (N = 5709).[a]

Variable	Categories	Infants No.	Deaths No.	Survival estimate (%)	95% CI
Overall		5709	980	82.83	81.83–83.78
Maternal race and ethnicity	Non-Hispanic White	1983	271	86.33	84.74–87.77
	Non-Hispanic Black	762	145	80.97	78.00–83.58
	Hispanic	2763	538	80.53	79.00–81.96
	Additional groups[b]	192	26	86.46	80.75–90.57
Classification	Chromosomal and Syndromic	900	303	66.33	63.14–69.32
	Isolated	1523	115	92.45	91.01–93.67
	Multiple	3058	519	83.03	81.65–84.31
Gestational age	Preterm <37 weeks	2428	512	78.91	77.24–80.48
	Term ≥37 weeks	3280	468	85.73	84.49–86.88
Birth weight	Low birth weight < 2500 grams	2341	555	76.29	74.52–77.96
	Normal birth weight ≥ 2500 grams	3367	425	87.38	86.21–88.45
Birth year	<2012	3578	642	82.06	80.76–83.28
	≥2012	2130	338	84.13	82.51–85.62
Maternal education	<12 years	1579	317	79.92	77.86–81.82
	12 years	1698	287	83.10	81.23–84.80
	≥12 years	2260	344	84.78	83.23–86.20
Maternal age	<35 years	4889	829	83.04	81.96–84.07
	≥35 years	819	151	81.56	78.73–84.05

[a]Only live-born infants with hydrocephaly without spina bifida were included in the analysis. The estimates are Kaplan–Meier survival estimates. Not all maternal and infant characteristics categories add up to the total because of missing data.

[b]Additional Groups race and ethnicity category included non-Hispanic Asian, non-Hispanic Native Hawaiian and Pacific Islander, non-Hispanic American Indian and Alaskan Native, and non-Hispanic two or more races.

TABLE 3

Unadjusted hazard ratios (indicating relative risk for death) for infants with hydrocephaly without spina bifida and selected cofactors, Texas, 1999–2017.

Variable	Categories	Hazard ratio	95% CI	*p* value
Maternal race and ethnicity	Non-Hispanic White (Referent)	1.00	—	—
	Non-Hispanic Black	1.42	1.16–1.74	<.001
	Hispanic	1.47	1.27–1.70	<.001
	Additional groups[a]	0.98	0.66–1.47	.94
Classification	Chromosomal and syndromic	5.17	4.17–6.41	<.001
	Isolated (referent)	1.00	—	—
	Multiple	2.37	1.93–2.90	<.001
Gestational age	Preterm <37 weeks	1.58	1.39–1.79	<.001
	Term ≥37 weeks (referent)	1.00	—	—
Birth weight	Low birth weight < 2500 grams	2.04	1.80–2.31	<.001
	Normal birth weight ≥ 2500 grams	1.00	—	—
Birth year	>2012	1.15	1.01–1.31	0.04
	≥2012 (referent)	1.00	—	—
Maternal education	<12 years	1.34	1.15–1.57	<.001
	12 years	1.12	0.96–1.31	0.17
	>12 years (referent)	1.00	—	—
Maternal age	<35 years (referent)	1.00	—	—
	≥35 years	1.09	0.92–1.30	0.32

[a]Additional groups race and ethnicity category included non-Hispanic Asian, non-Hispanic Native Hawaiian and Pacific Islander, non-Hispanic American Indian and Alaskan Native, and non-Hispanic two or more races.

TABLE 4

Adjusted hazard ratios (indicating relative risk for death) for infants with hydrocephaly without spina bifida and race and ethnicity, Texas, 1999–2017.[a]

Variable	Categories	Hazard ratio	95% confidence limit	*p* value
Race and Ethnicity	Non-Hispanic White (Referent)	1.00	—	—
	Non-Hispanic Black	1.28	1.04–1.58	.02
	Hispanic	1.31	1.12–1.54	<.001
	Additional groups[b]	0.95	0.63–1.42	.78

[a]Adjusted for classification, gestational age, birth weight, birth year, maternal education, and maternal age.

[b]Additional groups race and ethnicity category included non-Hispanic Asian, non-Hispanic Native Hawaiian and Pacific Islander, non-Hispanic American Indian and Alaskan Native, and non-Hispanic two or more races.