Racial/Ethnic Disparities in Risk of Early Childhood Mortality Among Children With Congenital Heart Defects

WHAT'S KNOWN ON THIS SUBJECT: Congenital heart defects (CHDs) are the leading cause of death among all infants with birth defects. Mortality and survival vary greatly and are influenced by several factors including length of follow-up, phenotype, number of co-occurring, and severity of defects.

WHAT THIS STUDY ADDS: Numerous reports have described growing disparities in infant and childhood mortality between non-Hispanic white and minority children. However, little is known about survival among minority children with CHDs. These data demonstrate racial/ethnic disparities in early childhood survival among children with CHDs.

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KEY WORDS
congenital heart defects, race/ethnicity, mortality, survival, childhood

ABBREVIATIONS
CHD—congenital heart defect
NH—non-Hispanic
TBDR—Texas Birth Defects Registry
TGA—transposition of the great arteries
VSD—ventricular septal defect
HLHS—hypoplastic left heart syndrome
ASD—atrial septal defect
RUCA—rural urban commuting area
HR—hazard ratio
CI—confidence interval

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abstract

BACKGROUND: Infants with congenital heart defects (CHDs) have increased risk of childhood morbidity and mortality. However, little is known about racial/ethnic differences in early childhood mortality.

PATIENTS AND METHODS: We conducted a retrospective cohort study with data from the Texas Birth Defect Registry on 19 530 singleton, liveborn infants with a CHD and born January 1, 1996, to December 31, 2003, to non-Hispanic (NH) white, NH black, and Hispanic women. Texas Birth Defect Registry data were linked to Texas death records and the National Death Index to ascertain deaths between January 1, 1996, and December 31, 2005. Kaplan-Meier survival estimates were computed, and hazard ratios (HRs) and 95% confidence intervals (CIs) were calculated from multivariable Cox-proportional hazard regression models to determine the effect of maternal race/ethnicity on mortality for selected CHD phenotypes.

RESULTS: After adjusting for covariates, compared with NH white children, NH black children had increased early childhood mortality risk for transposition of the great arteries (HR: 2.04 [95% CI: 1.40–2.97]), tetralogy of Fallot (HR: 1.85 [95% CI: 1.09–3.12]), pulmonary valve atresia without ventricular septal defect (VSD) (HR: 2.60 [95% CI: 1.32–5.12]), VSD (HR: 1.56 [95% CI: 1.19–2.03]), and atrial septal defect (HR: 1.34 [95% CI: 1.08–1.66]). Hispanic children had higher mortality risk for pulmonary valve atresia without VSD (HR: 1.76 [95% CI: 1.06–2.91]) and hypoplastic left heart syndrome (HR: 1.51 [95% CI: 1.13–2.02]).

CONCLUSIONS: We provide evidence that supports racial/ethnic disparities in early childhood mortality among infants with CHDs. Identifying infants with the greatest risk of early childhood mortality will facilitate development of interventions and policies to mitigate these risks. *Pediatrics* 2011;127:e1128–e1138

Birth defects are the leading cause of infant mortality in the United States. Congenital heart defects (CHDs), abnormalities in the formation of the heart and major vessels, are the most common of all birth defects (annual prevalence: 6–12 affected infants per 1000 live births[1–3]) and are the leading cause of death among all infants with birth defects. Although most infants with CHDs who are diagnosed early benefit from successful surgical repair, mortality rates can be quite high; survival rates vary greatly and are influenced by several factors including length of follow-up, phenotype, number of co-occurring defects, severity of defect, and also maternal race/ethnicity.[4–13]

Numerous study reports[14–18] and a recent technical report[19] have described growing disparities in health and health care between non-Hispanic (NH) white and minority children, especially in infant and childhood mortality. Minority children are more likely to receive lower-quality general pediatric care and have increased risk of death in infancy.[19] Racial/ethnic disparities in health and health care also exist among children with CHDs.[20] Minorities now comprise ~43% of all children in the United States, which is a 58% increase since 1990,[19] and conservative projections estimate that minorities will comprise half of US children by 2040. Therefore, even modest decreases in childhood survival rates or increases in mortality rates among minority children will cause greater burden on the health care system and adversely affect the lives of families. However, little is known about survival experiences among minority children with CHDs. We identified 4 studies in the peer-reviewed literature that were cross-sectional studies based on single-year administrative data[21,22] or national death-certificate data[20,23]; racial/ethnic disparities in mortality rates for children with CHDs were reported from all 4 of these studies. However, none was a population-based cohort study or used birth defects registry data. We conducted a population-based cohort study to determine survival rates and risk of early childhood mortality according to maternal race/ethnicity among children with CHDs.

PATIENTS AND METHODS

Study Design

We conducted a retrospective cohort study with data from the Texas Birth Defects Registry (TBDR), which is maintained by the Birth Defects Epidemiology and Surveillance Branch at the Texas Department of State Health Services. The TBDR is a population-based, active surveillance system that collects information on all structural and chromosomal birth defects diagnosed within 1 year after delivery. The TBDR began birth defects surveillance from 1996 to 1998 in increasingly larger areas of the state (35% of all resident live births in 1996, 56% in 1997, and 85% in 1998) and expanded to statewide coverage in 1999. The TBDR staff routinely visit delivery units, pediatric hospitals, and birthing centers where affected children are delivered or treated to identify and collect information for the registry.

To obtain additional demographic information, TBDR staff link registry cases to Texas birth and fetal death certificates filed with the Vital Statistics Unit of the Texas Department of State Health Services on the basis of the infant's and mother's names and dates of birth.[24] For 1996–2003 deliveries, the registry linked ≥99.0% of live-born case-infants to their Texas birth certificates. TBDR case-infants are also linked to their Texas death certificates through the death-to-birth certificate matching routinely performed by the Texas Vital Statistics Unit.

Study Population

We selected all live-born singleton infants diagnosed with a CHD within the first year of life and born January 1, 1996, to December 31, 2003, to NH white, NH black, or Hispanic women. CHDs were classified by using the British Pediatric Association extension of the *International Classification of Diseases, Ninth Edition, Clinical Modification* (ICD-9-CM) coding system, as modified by the Division of Birth Defects and Developmental Disabilities of the Centers for Disease Control and Prevention and by the TBDR. CHDs were categorized as 1 of 4 types of defects: conotruncal; right obstructive; left obstructive; or septal. Conotruncal CHDs included common truncus (745.000–745.010), transposition of the great arteries (TGA) (745.100–745.190), and tetralogy of Fallot (745.200–745.210, 746.840, or 746.000 with 745.400–745.490). Right obstructive CHDs included pulmonary valve atresia without ventricular septal defect (VSD) (746.000 without 745.400–745.490), pulmonary valve stenosis (746.010), anomalies of the tricuspid valve (746.100), and Ebstein anomaly (746.200). Left obstructive CHDs included hypoplastic left heart syndrome (HLHS) (746.700), aortic valve stenosis (746.300), and coarctation of the aorta (747.100–747.190). Septal CHDs included VSD (745.400–745.490), atrial septal defect (ASD) (745.510–745.590), and atrioventricular septal defect (745.600–745.690). Infants who had both pulmonary valve atresia and VSD were classified as having tetralogy of Fallot.

Study Variables

Information on medical, pregnancy, and sociodemographic factors was obtained from birth certificates and medical records. We included gender, gestational age (categorized as 20–36 or ≥37 completed weeks), birth weight

(<1500, 1500–2499, or ≥2500 g), and the number and nature of the defects (isolated CHD, multiple CHDs, or CHD with extracardiac defects). An extracardiac defect was defined as any of 34 major categories of genetic or structural defects that affect all organ systems except the circulatory system. Gestational age was calculated on the basis of last menstrual period taken from the medical record. When the date of the last menstrual period was missing, we substituted the clinical estimate of gestation. Following the method described by Bol et al,[25] a variable that combined categories of gestational age and birth weight was created, and the 6 resultant combinations were used in the analyses. Maternal factors included age (<20, 20–29, 30–39, or ≥40 years), race/ethnicity (NH white, NH black, or Hispanic), education (<12, 12, or >12 years), and primary residence at delivery. Primary maternal residence at delivery was defined by 2 variables: residence in a Texas-Mexico border county or not and rural versus urban area. The degree of urbanization or rurality of maternal residence at delivery was measured by using a zip-code approximation for the rural urban commuting area (RUCA) codes. The RUCA classification system uses the standard US Census Bureau urban area and place definitions in combination with traffic-flow patterns to characterize census tracts by rural and urban status. Our classification was based on primary traffic-flow categories and resulted in urban core areas (RUCA code 1), suburban areas (RUCA code 2), micropolitan areas (RUCA codes 3–6), and small town/rural areas (RUCA codes 7–10).

Ascertainment of Death

We linked cases of CHDs to Texas birth and death records to ascertain in-state deaths; we used the National Death Index at the National Center for Health Statistics to ascertain out-of-state deaths. Children were considered deceased if noted as deceased in Texas death records, the National Death Index, or the TBDR.

Statistical Analysis

We calculated descriptive statistics for the main study variables and covariates. Survival time was calculated as the time from date of birth to date of death; for infants who were not identified as deceased, survival time was calculated as the time from date of birth to the end of the study period (December 31, 2005). We computed Kaplan-Meier survival curves to describe the pattern of survival within early childhood for each type of CHD. Using proportional hazards regression (PHREG procedure in SAS 9.2 [SAS Institute, Inc, Cary, NC]), we calculated crude and adjusted hazard ratios (HRs) and 95% confidence intervals (CIs) to determine the effect of maternal race/ethnicity on early childhood survival; NH white children were used as the referent group.

The following variables were significantly associated with survival and were included as potential confounders in our models: maternal age; education; residence at delivery variables; infant gender; number of co-occurring defects; and gestational age/birth weight combinations. The statistical significance level was $P < .05$ for main effects.

The study was approved by the institutional review board at the University of South Florida, the National Death Index, and the Texas Department of State Health Services, including the use of data from Texas vital records and the TBDR.

RESULTS

During the study period, 2.35 million singleton live-born infants were born in the areas covered by the TBDR; 20 405 had at least 1 of the selected CHDs. We excluded infants not born to an NH white, NH black, or Hispanic mother ($n = 529$), diagnosed with trisomy 13 or 18 ($n = 343$), or children deceased but missing date of death ($n = 15$), which resulted in an unduplicated total of 875 excluded children (4.3%) (numbers do not add up because 12 children had >1 reason for exclusion). Our final study population included 19 530 children, 1572 (8.0%) of whom died before 1 year of age and 1826 (9.3%) of whom died during follow-up. Of these children, 67 (3.7%) did not have a death-certificate number, and 1 child had a death-certificate number that did not link to a death certificate. Thus, 1758 children had cause-of-death information; 840 (47.8%) had a CHD as the underlying cause of death, and 1046 (59.5%) had a CHD as any cause of death (includes the 840).

Approximately 26.7% of the study children were born preterm, and 21.2% were born at very low or low birth weight (Table 1). Of those who died during the study period, 39.9% were born preterm, 15.6% were born at very low birth weight, and 23.6% were born at low birth weight. Overall, after adjusting for covariates, NH black children with CHDs had a 32% increased risk of early childhood mortality compared with affected NH white children (HR: 1.32 [95% CI: 1.14–1.54]). In contrast, Hispanic children with CHDs had no increased risk overall of early childhood mortality compared with NH white children (HR: 0.96 [95% CI: 0.85–1.08]) (Table 2).

Conotruncal CHDs

As seen in Table 3, there were notable racial/ethnic disparities in early childhood survival rates for children with conotruncal defects. Among children with common truncus, only 49.1% of Hispanic and 58.3% of NH black children survived, whereas 67.0% of NH white children survived early child-

TABLE 1 Description of Infants With CHDs According to Maternal and Infant Characteristics: TBDR, 1996–2003

Characteristic	NH White		NH Black		Hispanic		Total	
	Case-Infants, *n* (%)	Deceased, *n* (%)	Case-Infants, *n* (%)	Deceased, *n* (%)	Case-Infants, *n* (%)	Deceased, *n* (%)	Case-Infants, *n* (%)	Deceased, *n* (%)
Mothers								
Maternal age								
<20 y	712 (10.0)	83 (13.1)	334 (17.8)	46 (19.2)	1800 (17.1)	183 (19.2)	2846 (14.6)	312 (17.1)
20–29 y	3572 (50.0)	321 (50.6)	986 (52.5)	123 (51.3)	5629 (53.5)	488 (51.3)	10 187 (52.2)	932 (51.0)
30–39 y	2590 (36.3)	210 (33.1)	494 (26.3)	63 (26.3)	2771 (26.4)	252 (26.5)	5855 (30.0)	525 (28.8)
≥40 y	265 (3.7)	20 (3.2)	65 (3.5)	8 (3.3)	312 (3.0)	29 (3.0)	642 (3.3)	57 (3.1)
Maternal education								
<High school	1064 (14.9)	108 (17.0)	375 (20.0)	50 (20.8)	5312 (50.5)	486 (51.1)	6751 (34.6)	644 (35.3)
High school	2231 (31.3)	200 (31.5)	757 (40.3)	98 (40.8)	3144 (29.9)	274 (28.8)	6132 (31.4)	572 (31.3)
>High school	3711 (52.0)	309 (48.7)	686 (36.5)	75 (31.3)	1801 (17.1)	153 (16.1)	6198 (31.7)	537 (29.4)
Residence in a border county								
Yes	206 (2.9)	26 (4.1)	15 (0.8)	6 (2.5)	3204 (30.5)	300 (31.5)	3425 (17.5)	332 (18.2)
No	6933 (97.1)	608 (95.9)	1864 (99.2)	234 (97.5)	7308 (69.5)	652 (68.5)	16 105 (82.5)	1494 (81.8)
RUCA								
Urban core areas	4823 (67.6)	413 (65.1)	1648 (87.7)	207 (86.3)	8533 (81.2)	765 (80.4)	15 004 (76.8)	1385 (75.8)
Suburban areas	1031 (14.4)	94 (14.8)	61 (3.2)	7 (2.9)	605 (5.8)	47 (4.9)	1697 (8.7)	148 (8.1)
Micropolitan areas	707 (9.9)	71 (11.2)	90 (4.8)	10 (4.2)	866 (8.2)	86 (9.0)	1663 (8.5)	167 (9.1)
Small town area/ rural areas	558 (7.8)	56 (8.8)	74 (3.9)	14 (5.8)	497 (4.7)	54 (5.7)	1129 (5.8)	124 (6.8)
Infants								
Gender								
Male	3653 (51.2)	343 (54.1)	943 (50.2)	115 (47.9)	5273 (50.2)	507 (53.3)	9869 (50.5)	965 (52.8)
Female	3481 (48.8)	290 (45.7)	933 (49.7)	123 (51.3)	5228 (49.7)	442 (46.4)	9642 (49.4)	855 (46.8)
Birth weight								
<1500 g	457 (6.4)	91 (14.4)	289 (15.4)	52 (21.7)	644 (6.1)	141 (14.8)	1390 (7.1)	284 (15.6)
1500–2499 g	1005 (14.1)	146 (23.0)	327 (17.4)	62 (25.8)	1427 (13.6)	223 (23.4)	2759 (14.1)	431 (23.6)
≥2500 g	5675 (79.5)	396 (62.5)	1260 (67.1)	124 (51.7)	8433 (80.2)	588 (61.8)	15 368 (78.7)	1108 (60.7)
Gestational age								
<37 wk	1850 (25.9)	240 (37.9)	686 (36.5)	119 (49.6)	2674 (25.4)	369 (38.8)	5210 (26.7)	728 (39.9)
≥37 wk	5287 (74.1)	393 (62.0)	1192 (63.4)	121 (50.4)	7833 (74.5)	583 (61.2)	14 312 (73.3)	1097 (60.1)
Birth weight/gestational age								
<37 wk, <1500 g	440 (6.2)	86 (13.6)	284 (15.1)	50 (20.8)	617 (5.9)	135 (14.2)	1341 (6.9)	271 (14.8)
<37 wk, 1500–2499 g	655 (9.2)	86 (13.6)	207 (11.0)	37 (15.4)	887 (8.4)	128 (13.4)	1749 (9.0)	251 (13.7)
<37 wk, ≥2500 g	754 (10.6)	67 (10.6)	193 (10.3)	30 (12.5)	1169 (11.1)	106 (11.1)	2116 (10.8)	203 (11.1)
≥37 wk, <1500 g	17 (0.2)	5 (0.8)	5 (0.3)	2 (0.8)	27 (0.3)	6 (0.6)	49 (0.3)	13 (0.7)
≥37 wk, 1500–2499 g	349 (4.9)	59 (9.3)	120 (6.4)	25 (10.4)	539 (5.1)	95 (10.0)	1008 (5.2)	179 (9.8)
≥37 wk, ≥2500 g	4920 (68.9)	329 (51.9)	1066 (56.7)	94 (39.2)	7261 (69.1)	482 (50.6)	13 247 (67.8)	905 (49.6)
Type of defect								
Isolated heart defect	2872 (40.2)	97 (15.3)	827 (44.0)	52 (21.7)	4483 (42.6)	141 (14.8)	8182 (41.9)	290 (15.9)
Multiple heart defects	2718 (38.1)	240 (37.9)	696 (37.0)	97 (40.4)	3892 (37.0)	388 (40.8)	7306 (37.4)	725 (39.7)
Extracardiac defects	1549 (21.7)	297 (46.8)	356 (18.9)	91 (37.9)	2137 (20.3)	423 (44.4)	4042 (20.7)	811 (44.4)

Percentages do not add up to 100% and counts do not add up to the total because of missing values.

hood. Most notably, NH black children with TGA (Fig 1) had a much lower survival rate during early childhood than NH white or Hispanic children (52.4% vs 76.9% and 71.5%, respectively). NH black and Hispanic children had poorer survival rates than NH white children for tetralogy of Fallot (73.3% and 76.0%, respectively, vs 84.2%) (Fig 2).

After adjusting for covariates, NH black and Hispanic children with common truncus had an ~88% increased risk of early childhood mortality compared with similarly affected NH white children, but the increase was not statistically significant (Table 2). NH black children with TGA had a statistically significant twofold increased risk of death in early childhood compared with similarly affected NH white children (95% CI: 1.40–2.97). NH black children with tetralogy of Fallot also had almost a twofold increased risk of death in early childhood compared with similarly affected NH white children (HR: 1.85 [95% CI: 1.09–3.21]). Hispanic children with tetralogy of Fallot had a 39% increased risk of mortality compared with NH white children, but

TABLE 2 Data From Cox-Proportional Hazards Regression Models for Risk of Early Childhood Mortality Among Children With CHDs According to Maternal Race/Ethnicity: TBDR, 1996–2003

CHD	NH White, *n* (%)[a]	NH Black			Hispanic		
		n (%)[a]	Adjusted HR (95% CI)[b]	*P*	*n* (%)[a]	Adjusted HR (95% CI)[b]	*P*
Conotruncal							
Common truncus	61 (38.1)	12 (7.5)	1.88 (0.62–5.66)	.2617	87 (54.4)	1.76 (0.88–3.49)	.1080
TGA	418 (39.0)	87 (8.1)	2.04 (1.40–2.97)[c]	.0002[c]	566 (52.9)	1.16 (0.87–1.55)	.3050
Tetralogy of Fallot	300 (39.2)	84 (11.0)	1.85 (1.09–3.12)[c]	.0215[c]	382 (49.9)	1.39 (0.92–2.10)	.1148
Right obstructive							
Tricuspid valve defects	197 (35.1)	80 (14.2)	1.41 (0.90–2.21)	.1329	285 (50.7)	0.97 (0.66–1.43)	.8806
Pulmonary valve stenosis	464 (39.7)	151 (12.9)	1.13 (0.57–2.22)	.7240	555 (47.4)	1.15 (0.68–1.96)	.6008
Pulmonary valve atresia without VSD	95 (32.8)	29 (10.0)	2.60 (1.32–5.12)[c]	.0058[c]	166 (57.2)	1.76 (1.06–2.91)[c]	.0290[c]
Ebstein anomaly	51 (31.9)	13 (8.1)	1.42 (0.43–4.70)	.5699	96 (60.0)	1.88 (0.74–4.79)	.1864
Left obstructive							
HLHS	204 (44.4)	43 (9.4)	1.06 (0.67–1.66)	.8091	213 (46.3)	1.51 (1.13–2.02)[c]	.0059[c]
Aortic valve stenosis	261 (46.6)	35 (6.3)	1.02 (0.49–2.13)	.9608	264 (47.1)	0.92 (0.56–1.51)	.7332
Coarctation of the aorta	502 (43.8)	89 (7.8)	1.12 (0.71–1.76)	.6379	554 (48.4)	0.73 (0.53–1.02)	.0632
Septal							
VSD	3559 (34.3)	869 (8.4)	1.56 (1.19–2.03)[c]	.0011[c]	5954 (57.4)	0.96 (0.79–1.18)	.7086
ASD	3347 (36.5)	937 (10.2)	1.34 (1.08–1.66)[c]	.0084[c]	4880 (53.3)	0.94 (0.80–1.11)	.4577
Atrioventricular septal defect	354 (41.5)	115 (13.5)	1.02 (0.68–1.54)	.9197	384 (45.0)	0.98 (0.71–1.37)	.9232
Total	7139 (36.6)	1879 (9.6)	1.32 (1.14–1.54)[c]	.0003[c]	10512 (53.8)	0.96 (0.85–1.08)	.4749

The follow-up period was from 1996 to 2005.

[a] Percentages may add up to >100% because of rounding.

[b] Adjusted for maternal age, education, infant gender, border county residence, RUCA, number of defects, birth weight/gestational age combinations.

[c] Statistically significant findings.

TABLE 3 Kaplan-Meier Estimates and 95% CIs for Early Childhood Survival Among Children With CHDs According to Maternal Race/Ethnicity: TBDR, 1996–2003

CHD	NH White			NH Black			Hispanic		
	Cases, *n*	Deaths, *n*	Estimate (95% CI)	Cases, *n*	Deaths, *n*	Estimate (95% CI)	Cases, *n*	Deaths, *n*	Estimate (95% CI)
Conotruncal									
Common truncus	61	20	67.0 (53.7–77.3)	12	5	58.3 (27.0–80.1)	87	44	49.1 (38.2–59.2)
TGA	418	97	76.9 (72.5–80.7)	87	41	52.4 (40.6–62.9)	566	163	71.5 (67.6–75.1)
Tetralogy of Fallot	300	46	84.2 (79.4–87.9)	84	22	73.3 (62.3–81.6)	382	92	76.0 (71.3–80.0)
Right obstructive									
Tricuspid valve defects	197	56	70.9 (63.8–76.8)	80	33	58.2 (46.5–68.3)	285	84	70.1 (64.4–75.2)
Pulmonary valve stenosis	464	32	93.0 (90.1–95.0)	151	16	88.6 (81.7–93.0)	555	50	90.7 (87.9–92.9)
Pulmonary valve atresia without VSD	95	27	72.4 (62.1–80.3)	29	15	48.3 (29.5–64.8)	166	67	59.0 (50.9–66.2)
Ebstein anomaly	51	11	77.5 (62.8–87.0)	13	5	61.5 (30.8–81.8)	96	34	64.6 (54.1–73.2)
Left obstructive									
HLHS	204	114	43.5 (36.4–50.3)	43	27	37.2 (23.1–51.3)	213	146	30.9 (24.7–37.2)
Aortic valve stenosis	261	52	79.7 (74.2–84.2)	35	10	71.4 (53.4–83.5)	264	55	79.6 (74.1–84.0)
Coarctation of the aorta	502	107	78.4 (74.4–81.7)	89	26	70.1 (59.1–78.6)	554	112	80.2 (76.6–83.3)
Septal									
VSD	3559	200	94.4 (93.6–95.1)	869	81	90.8 (88.6–92.6)	5954	348	94.1 (93.5–94.7)
ASD	3347	307	90.7 (89.7–91.7)	937	123	86.9 (84.6–88.9)	4880	493	89.9 (89.0–90.7)
Atrioventricular septal defect	354	87	75.2 (70.2–79.4)	115	35	68.8 (59.1–76.6)	384	116	69.6 (64.7–73.9)
Total	7139	634	91.1 (90.4–91.7)	1879	240	87.3 (85.7–88.7)	10512	952	90.9 (90.4–91.5)

The follow-up period was from 1996 to 2005.

the increase was not statistically significant.

Right Obstructive CHDs

NH black children with tricuspid valve defects had a notably poorer survival rate during early childhood compared with similarly affected NH white and Hispanic children (58.2% vs 70.9% and 70.1%, respectively) (Table 3). Figure 3 shows that NH black and Hispanic children with pulmonary valve atresia without VSD had lower survival rates (48.3% and 59.0%, respectively) than NH white children (72.4%). Both NH black and Hispanic children with Ebstein anomaly fared much worse during the first 5 years than similarly affected NH white children (61.5% and 64.6% vs 77.5%, respectively) (Table 3).

After adjusting for covariates, NH black children with tricuspid valve defects had a 42% increased risk of early childhood mortality compared with NH white children with tricuspid valve de-

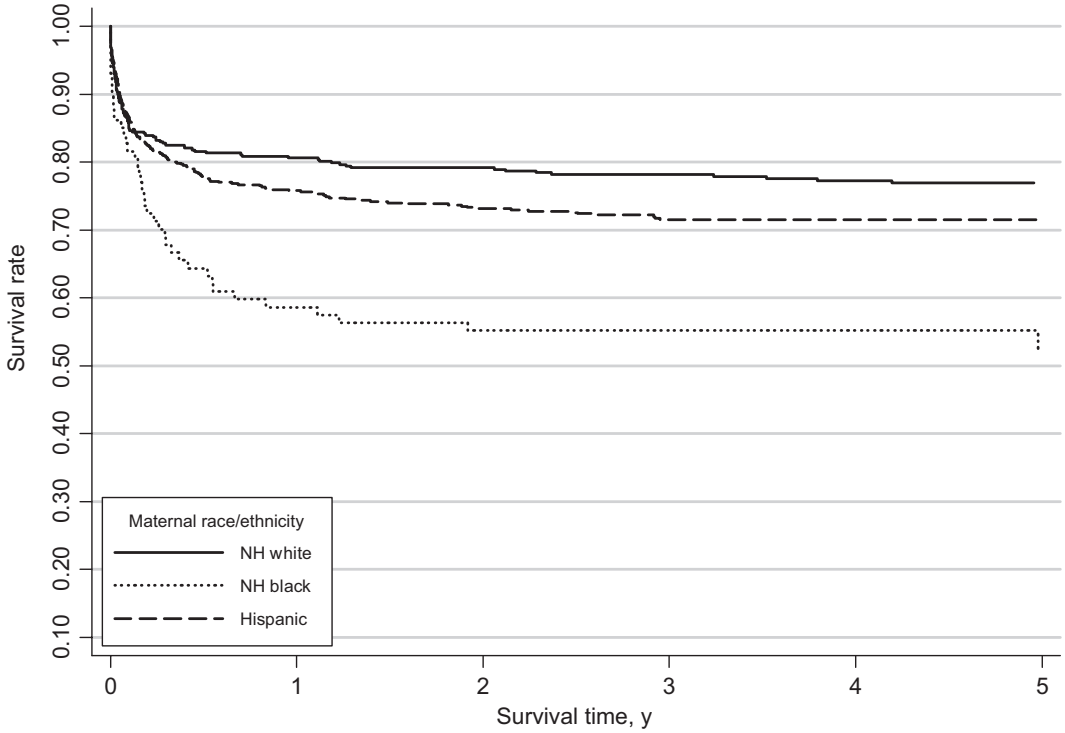

FIGURE 1
Early childhood Kaplan-Meier survival curves for infants with TGA according to maternal race/ethnicity; TBDR, 1996–2003.

fects, but the increase was not statistically significant (Table 2). NH black and Hispanic children with pulmonary valve atresia without VSD had a statistically significant 160% and 76%, increased risk of death in early childhood, respectively, after adjusting for covariates, compared with NH white children with the same CHD (95% CI: 1.32–5.12 and 1.06–2.91, respectively). Although NH black and Hispanic children with Ebstein anomaly had increased risk of mortality, the increases were not statistically significant in the adjusted analyses.

Left Obstructive CHDs

Racial/ethnic disparities in survival rates were seen among children with left obstructive CHDs (Table 3). Although survival rates were low for all children with HLHS, Hispanic children had the lowest survival rate (30.9%), followed by NH black (37.2%) and NH white (43.5%) children (Fig 4). After adjusting for covariates, Hispanic children with HLHS had a 51% greater risk of death compared with NH white children (95% CI: 1.13–2.02) (Table 2). NH black children with aortic valve stenosis and coarctation of the aorta had poorer survival rates than NH white and Hispanic children (Table 3). In contrast to the unadjusted analyses (Kaplan-Meier), NH black children with aortic valve stenosis or coarctation of the aorta had no increased risk of early childhood mortality compared with NH white children after adjusting for covariates (Table 2).

Septal CHDs

Overall, the survival rate for infants with septal defects was quite high regardless of maternal race/ethnicity; however, NH black children had a consistent pattern of having the lowest survival rate of the 3 racial/ethnic groups (Table 3). NH black and Hispanic children with atrioventricular septal defect had poorer survival rates (68.8% and 69.6%, respectively) than NH white children (75.2%) with the same defect.

As shown in Table 2, after adjusting for covariates, NH black children with VSD had a 56% increased risk and those with ASD had a 34% increased risk of early childhood mortality compared with NH white children with the same CHD (95% CI: 1.19–2.03 and 1.08–1.66, respectively).

DISCUSSION

We investigated maternal race/ethnicity and its effects on early childhood survival among children with CHDs; NH black race/ethnicity was more strongly associated with increased risk of early childhood mortality than Hispanic race/ethnicity. NH black children with TGA, tetralogy of Fallot, pulmonary valve atresia without VSD, VSD, and ASD had a statistically significant increased risk of mortality compared with NH white children with the same defects. Hispanic children with pulmo-

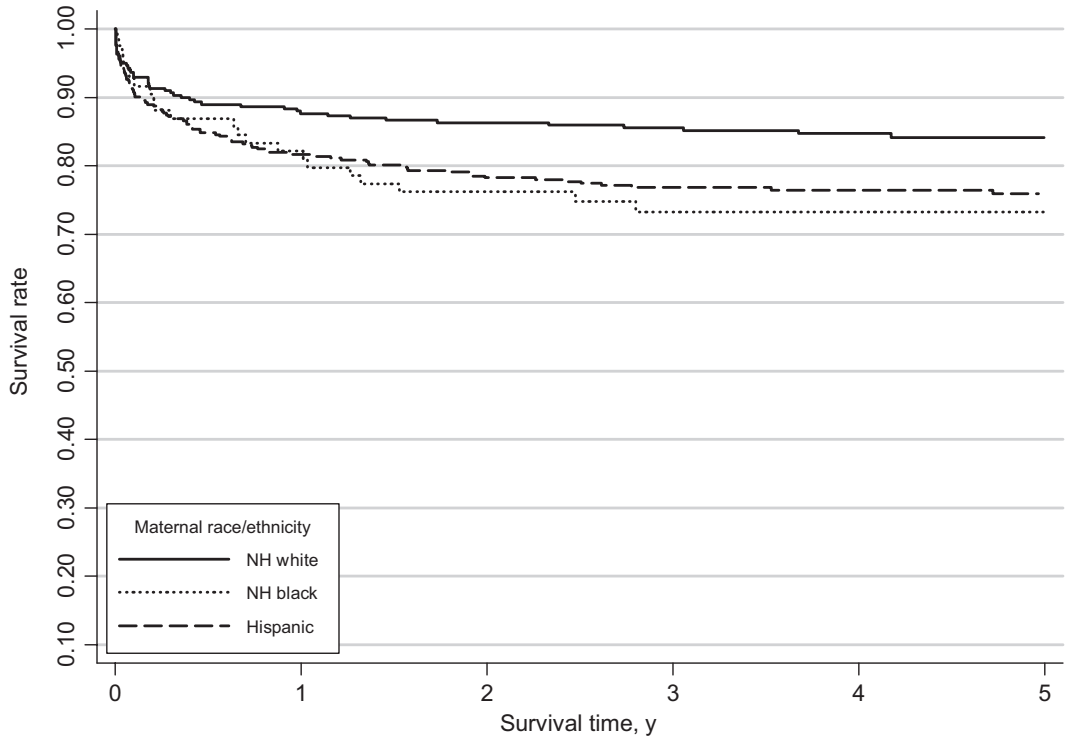

FIGURE 2
Early childhood Kaplan-Meier survival curves for infants with tetralogy of Fallot according to maternal race/ethnicity; TBDR, 1996–2003.

nary valve atresia without VSD or HLHS also had a statistically significant increased risk of mortality compared with NH white children. With inspection of the survival curves for TGA, pulmonary valve atresia without VSD, and HLHS (Figs 1–3) it is apparent that the disparity in ethnic mortality develops during months 2 to 3 of life. Because these cases require cardiac surgery during the first month, this temporal trend suggests that deficiencies in postdischarge care play a role in the increased mortality rates among minorities. Such findings indicate a need to intensify home health care and enhance communication between the cardiac center and the families.

Our findings provide additional evidence of racial/ethnic disparities in early childhood survival among children with CHD and are consistent with those of earlier reports. Using 1996 data from the Healthcare Cost and Utilization Project (HCUP) Kids' Inpatient Database (KID) for 4 states, Gonzalez et al[16] found that Hispanic children had a higher risk of dying after surgery for CHD than NH white children. Although there was no statistically significant difference in mortality rates overall between NH black and white children, there was variation in mortality rates for NH black children between states. Benavidez et al[22] used 2000 HCUP-KID data for 19 states and found that black and Hispanic children with CHD had higher odds of in-hospital mortality after CHD surgery than white children after adjusting for covariates. Another study[20] used national death-certificate data from 1979–1997 to determine trends in CHD-associated mortality in the United States. Black children had a 19% higher mortality rate from CHDs compared with NH white children and a lower decline in mortality rates over the 18-year period than NH white children. The average ages at death of CHD were also 3 to 6 times lower for black children compared with NH white children and approximately half the average age at death compared with NH white children for children with TGA, tetralogy of Fallot, VSD, and single ventricle. The infant mortality rate for VSD was higher during the 18-year study period for NH black children.

The racial/ethnic disparities we observed in early childhood survival and mortality may be caused by (1) underlying racial/ethnic biological differences, (2) differences in access to health care,[22,26] and (3) differences in cultural preferences. Biological differences that influence survival include the severity of the defect,[27–30] number of co-occurring defects,[27,31] and prevalence at live birth. In our study, we found higher mortality rates for NH black children with several types of CHDs. Defects greatly vary in severity, which may partially explain our findings. Although we were unable to evaluate the effect of severity on early childhood mortality and whether severity differed according to race/ethnicity, there is no published evidence

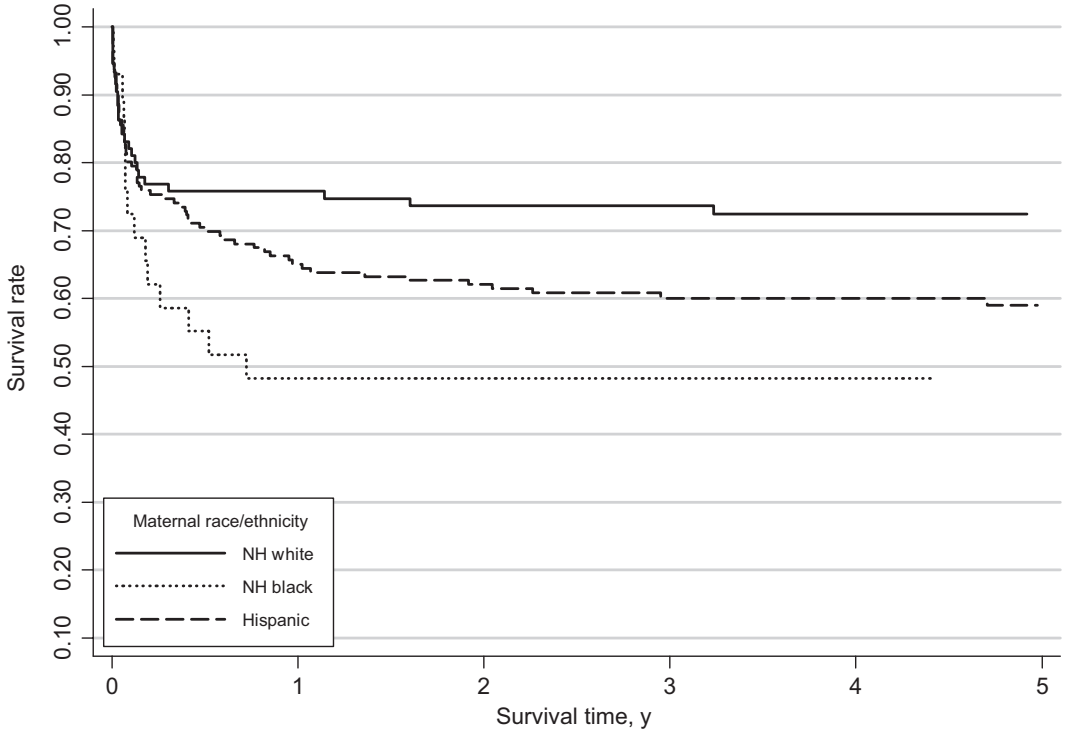

FIGURE 3
Early childhood Kaplan-Meier survival curves for infants with pulmonary valve atresia without VSD according to maternal race/ethnicity; TBDR, 1996–2003.

that suggests that defect severity differs according to maternal race/ethnicity. Our findings may also be related to racial/ethnic differences in the distribution of extracardiac defects and syndromes; however, we did adjust for the number of co-occurring defects in our analyses and excluded infants with trisomies 13 and 18.

The racial/ethnic differences we observed in early childhood survival could also be a result of lack of timely access to good-quality health care. Factors associated with access to good-quality health care include age at operation,[32,33] place of residence (ie, urban versus rural location), hospital volume, surgical case volume,[33] and socioeconomic factors.[21,34] Racial/ethnic differences in access to pediatric care and treatment received when care is obtained have been well documented.[35–37] For example, minority children on waiting lists for heart transplants have higher wait-list mortality rates than NH white children.[38,39] Mahle et al[40] showed that NH black children had a lower 5-year transplant graft survival rate than white children; the median survival time was ~6 years lower than that for white children. NH black children also had a higher median age at transplant (~5 years older) and were also more likely to have an HLA antigen mismatch.[40] Although it is plausible that lack of access to good-quality health care may cause black children to be less likely to have their CHD diagnosed early in infancy than white children, studies have revealed no statistically significant differences between black and white children in the age at diagnosis of CHDs or age at surgical repair for infants with CHDs.[34,41] Moreover, the age at operation has not been shown to affect health outcomes.[34,42] Thus, a racial/ethnic differential in age at diagnosis or surgical repair of CHD is an unlikely explanation for our findings.

NH black adults and children also have lower rates of surgical cardiovascular procedures than NH white adults and children, possibly because of socioeconomic status, insurance coverage, access to services, transportation, knowledge and understanding of the procedures, and patient trust and comfort issues.[43–46] Racial/ethnic differences in the quality of care received may also occur because of differences within a medical facility according to race/ethnicity and between medical facilities (those that serve primarily minority versus primarily white populations).[26,47] In general, white and black patients receive care at different medical institutions; black patients are treated more often at hospitals that have higher mortality rates, which possibly suggests lower quality of care.[22,26,48] Racial/ethnic disparities in access to good-quality care may also be the result of differences in health insurance; type of health insurance has a significant effect on access to care among children.[33,49,50] For example, among children with special

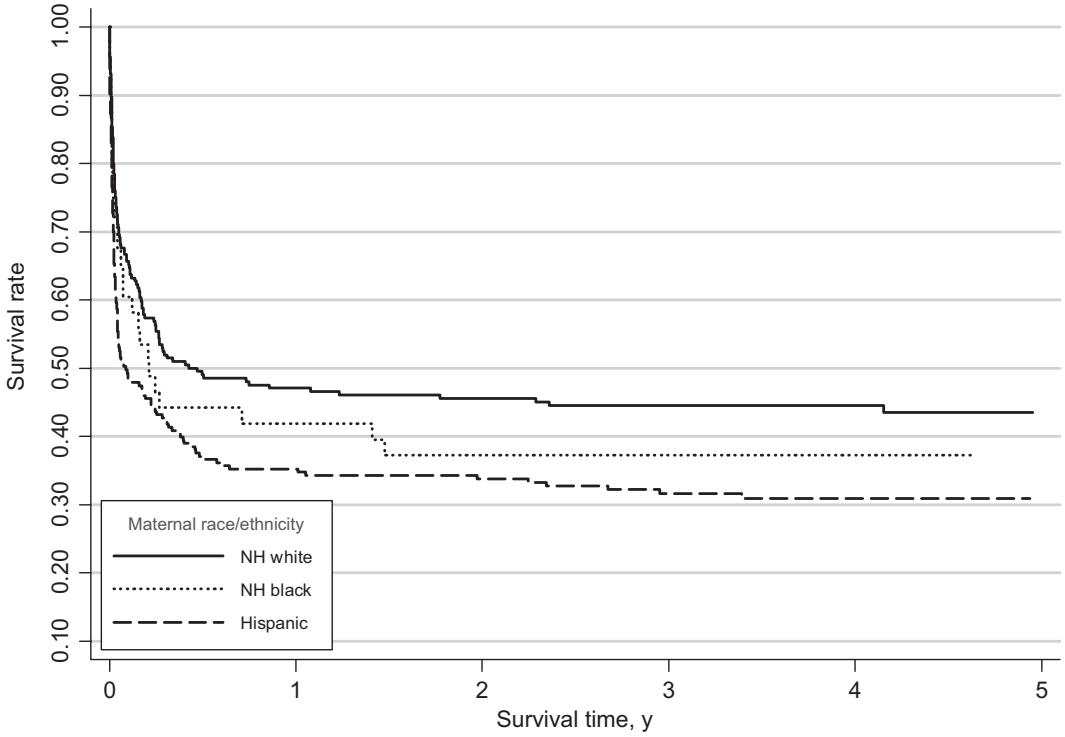

FIGURE 4
Early childhood Kaplan-Meier survival curves for infants with HLHS according to maternal race/ethnicity; TBDR, 1996–2003.

health care needs, black children are more likely to not have a regular clinician, are less likely to have a usual source of care at a physician's private office or health maintenance organization office,[51] and have reported higher levels of dissatisfaction with care.[52]

Other important influences on early childhood survival are cultural factors and preferences.[21,34] Differences in prenatal diagnosis of defects that result in a bias favoring the termination of more severely affected fetuses may partially explain our findings. NH black and Hispanic children are less likely to have their defect detected prenatally[53] and less likely to terminate their pregnancies if a defect is detected.[54]

Strengths of our study include its population-based cohort design, a large number of live births, and data from a defined multiethnic population ascertained by an active surveillance system with high sensitivity. However, our study has some potential limitations. We may have had incomplete ascertainment of (1) deaths and (2) cases of birth defects for years when the registry covered limited regions of the state; however, it is unclear how these missing data may have affected our findings. Another limitation was our inability to adjust for the potential effects of racial/ethnic differences in the type of treatment provided to the child, preferences in medical treatments received, health insurance/type of payment for services, and surgical complications.

CONCLUSIONS

Our study contributes to a growing body of literature that indicates that NH black and Hispanic children with specific phenotypes of CHDs have poorer survival rates in early childhood than NH white children. Future investigations should determine if there are racial/ethnic differences in access to care among children with CHDs, the severity of CHDs or defect subtypes, the types of treatments selected, and underlying causes of death. Elucidation of these factors will facilitate development of public health and tertiary prevention strategies to address this important health outcome.

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