RACIAL/ETHNIC DISPARITIES IN MORTALITY RELATED TO CONGENITAL HEART DEFECTS AMONG CHILDREN AND ADULTS IN THE UNITED STATES

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Background: Congenital heart defects (CHD) are the most common birth defect and are a major cause of childhood illness and death. Recent progress in management of persons with CHD may have decreased CHD-related mortality.

Methods: Year 2000 US death records were used to determine CHD-related mortality by age, sex, and race/ethnicity in children and adults. CHD-related mortality was defined as all deaths with any mention of CHD on the death certificate. Age-, sex-, and racial/ethnic-specific population counts were obtained from the 2000 US Census and used as denominators in mortality rates.

Results: In 2000 there were 5441 (.23%) CHD-related deaths and CHDs were mentioned 6121 times as the underlying or contributing cause of death. In 68.4% of CHD-related deaths, CHD was the underlying cause of death. Non-Hispanic Black males had greater risk of CHD-related death than did non-Hispanic White males (RR 1.25, 95% CI 1.08–1.45). Both Hispanic males and females had lower rates of CHD-related deaths than did non-Hispanic Whites (RR .72, 95% CI .60–.85; RR .52, 95% CI .42–.65, respectively). ''Unspecified congenital malformation of the heart'' was the most common cause of death overall; however, ''malformation of the coronary vessels'' was most often a cause of death for non-Hispanic Blacks and children aged 10–19 years.

Conclusions: Racial/ethnic differences in CHD-related mortality exist in the United States. Management of CHD, access to adequate care, and misclassification in cause of death reporting on death records may explain the observed differences. (*Ethn Dis*. 2008;18:442–449)

Key Words: Mortality, Congenital Heart Defects, Disparities, Epidemiology, Race/Ethnicity

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INTRODUCTION

With an annual prevalence of 7–10 per 1,000 live births, congenital heart defects (CHD) are the most common and multifaceted family of birth defects.[1–5] CHDs have serious consequences, in terms of short- and long-term morbidity and mortality.[6,7] During the past few decades, many advances have been made in the management of CHD, including improvement in prenatal diagnosis, fetal interventions, postnatal diagnosis, and both surgical and percutaneous interventions.[8–11] Because of these advances, more children with CHD survive into adulthood.[12,13] Despite these extraordinary technical advances, previous research suggests sex and racial/ethnic disparities[14,15] in mortality among persons with CHD. These disparities have not been fully described or explored; therefore, the purpose of this study was to determine mortality related to CHD and examine age, racial/ethnic, and sex disparities in mortality related to CHD in children and adults.

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METHODS

Our study population consisted of all residents of the United States in 2000. We obtained multiple-cause mortality public-use data files from the National Center for Health Statistics, which annually compiles data from all death certificates filed in the United States. These files contain demographic and geographic information on decedents, International Classification of Disease (ICD) codes for the underlying cause of death, and up to 20 additional conditions listed on the death certificate. The ICD Tenth Revision (ICD-10) was implemented in 1999.

We determined the number of deaths for which a CHD (ICD-10 codes Q20.0–Q26.9) was listed anywhere on the death certificate. These codes include most congenital malformations of the heart and circulatory system but exclude malformations of the peripheral vascular system. We defined CHD-related deaths as all deaths that had at least one mention of a CHD on the death certificate, as either an underlying or contributing cause of death.

We analyzed several specific demographic groups in this study, including males and females, three racial/ethnic groups (non-Hispanic Whites, non-Hispanic Blacks, and Hispanics), and six age groups (<1, 1–4, 5–14, 15–19, 20–64, and ≥65 years). Age-, sex-, and racial/ethnicity-specific population counts were obtained from the 2000 US Census of Population and Housing and used as denominators for calculat-

Table 1. Leading CHD as the underlying and contributing cause of death and number of mentions on death certificates for CHD decedents, US, 2000 (*N*=5441)

Defect (IDC-10 Code)	Underlying		Contributing		Total # Mentions	
	n	%*	*n*	%*	*n*	%*
1. Unspecified congenital malformation of heart (Q24.9)	1256	20.5	470	7.7	1725	28.2
2. Hypoplastic left heart (Q23.4)	424	6.9	93	1.5	517	8.4
3. Atrial septal defect (Q21.1)	281	4.6	237	3.9	518	8.5
4. Ventricular septal defect (Q21.0)	183	3.0	289	4.7	472	7.7
5. Malformation of the coronary vessels (Q24.5)	179	2.9	105	1.7	284	4.6
6. Tetralogy of Fallot (Q21.3)	178	2.9	87	1.4	265	4.3
7. Other specified congenital malformation of the heart (Q24.8)	114	1.9	67	1.1	181	3.0
8. Other congenital malformations of cardiac septa Q21.8)	97	1.6	45	0.7	142	2.3
9. Transposition of the great vessels (complete) (Q20.3)	95	1.6	47	0.7	142	2.3
10. Insufficient aortic valve (Q23.1)	92	1.5	71	1.2	163	2.7
11. Coarctation of the aorta (Q25.1)	71	1.2	108	1.8	179	2.9
12. Atrioventricular septal defect (Q21.2)	68	1.1	60	1.0	128	2.1
13. Congenital stenosis of the aortic valve (Q23.0)	66	1.1	39	0.6	105	1.7
14. Common arterial trunk (Q20.0)	65	1.1	16	0.3	81	1.3
15. Atresia of the pulmonary artery (Q25.5)	54	0.9	77	1.3	131	2.1

* Calculated by dividing the number of deaths by the total number of mentions (6,121) on the death certificates

ing mortality. We calculated mortality separately for demographic groups defined by age, sex, and race/ethnicity and per 100,000 population. In some cases, we present age-adjusted mortality; for those rates, the direct method of age-adjustment was used with the 2000 US population as the standard. Infant mortality was calculated as the number of deaths before the first birthday divided by the number of live births in the United States in 2000 (per 10,000 live births). Rate ratios (RR) and *P* values were also calculated. *P* values$<$.05 were considered significant, and SAS version 9.1 (SAS Institute, Inc, Cary, NC) was used for the analyses.

The Office of Research Integrity and Compliance, the institutional review board at the University of South Florida, reviewed and approved the study.

RESULTS

In 2000, 5441 (0.23%) CHD-related deaths occurred in the US. Some had more than one CHD mentioned as the underlying or contributing cause of death on the death certificates (6121 total mentions). In 68.4% of CHD-related deaths, the CHD was the underlying cause of death. ''Unspecified congenital malformation of the heart'' (UCMH) was the CHD most frequently the underlying (20.5%) or contributing (7.7%) cause of death among CHD-related deaths (Table 1). Aside from UCMH, the five CHDs that were most frequently the underlying cause of death were hypoplastic left heart syndrome, atrial septal defect, ventricular septal defect, malformation of the coronary vessels, tetralogy of Fallot, and ''other specified congenital malformations of the heart.''

Differences in Mortality by Race/Ethnicity

There were significant differences in CHD-related pattern of death by race/ethnicity. Although no racial/ethnic differences were observed in overall mortality, mortality rates differed by age group (Table 2). Infant mortality was 1.45 times as high (95% CI 1.31–1.60) among non-Hispanic Blacks than among non-Hispanic Whites. CHD-related mortality was also higher for non-Hispanic Black children aged 1–4 (RR 2.09, 95% CI 1.54–2.80), 10–14 (RR 1.76, 95% CI 1.08–2.79) and 15–19 years (RR 1.90, 95% CI 1.29–2.77) than for their non-Hispanic White counterparts. Hispanic children aged 1–4 years also had higher rates than did non-Hispanic Whites (RR 1.40. 95% CI 1.02–1.89). However, mortality for non-Hispanic Blacks and Hispanics was similar to that seen among non-Hispanic Whites for persons aged 20–64 years.

Racial/ethnic differences in the underlying and contributing causes of death were also observed. Although UCMH was the CHD that accounted for the most deaths for all groups, non-Hispanic Blacks and Hispanics were 20% more likely to have it as an underlying cause of death than non-Hispanic Whites (Table 3). Non-Hispanic Blacks and Hispanics were less likely to die from atrial septal defect than were non-Hispanic Whites. Non-Hispanic Blacks were two times more likely to have malformation of the coronary vessels as the underlying cause of death than were non-Hispanic Whites. Among Hispanics, hypoplastic left heart syndrome was more often the underlying cause of death.

Differences in Mortality by Sex and Race/Ethnicity

Males

The greatest differences we observed were among males and females by race/

Table 2. Age-adjusted mortality rates*, rate ratios and 95% confidence intervals for congenital heart defect-related deaths by age, sex and race/ethnicity, US, 2000 (*N*=5441)

	MALES		FEMALES			TOTAL	
Age group	*n*	MR*	*n*	MR*	RR‡¶ (95% CI**)	*n*	MR*
< 1 yrs	1313	6.79†	1223	6.62†	0.97 (0.90–1.05)	2536	6.70†
1–4 yrs	160	2.04	126	1.68	0.82 (0.65–1.05)	286	1.86
5–9 yrs	40	0.38	43	0.43	1.13 (0.72–1.78)	83	0.40
10–14 yrs	58	0.55	47	0.47	0.85 (0.57–1.27)	105	0.51
15–19 yrs	106	1.02	54	0.55	0.54 (0.38–0.75)	160	0.79
20–64 yrs	806	0.98	592	0.71	0.72 (0.65–0.81)	1398	0.84
65+ yrs	310	2.15	445	2.16	1.00 (0.87–1.16)	755	2.16
Total	1480	10.87	1307	9.24	0.85 (0.79–0.91)	2787	1.00
	NH-WHITE		NH-BLACK			HISPANIC	
	n	MR*	*n*	MR*	RR§ (95% CI)	*n*	MR*	RR§(95% CI)
Sex								
Males	1074	1.14	227	1.43	1.25 (1.08–1.45)	145	0.82	0.72 (0.60–0.85)
Females	1016	1.03	164	0.93	0.90 (0.76–1.07)	91	0.54	0.52 (0.42–0.65)
Age group								
< 1 yrs	1464	6.20†	542	8.97†	1.45 (1.31–1.60)	530	6.50†	1.05 (0.95–1.16)
1–4 yrs	138	1.53	70	3.20	2.09 (1.54–2.80)	63	2.14	1.40 (1.02–1.89)
5–9 yrs	53	0.43	17	0.54	1.26 (0.69–2.22)	11	0.30	0.69 (0.33–1.36)
10–14 yrs	65	0.50	27	0.88	1.76 (1.08–2.79)	10	0.32	0.63 (0.29–1.23)
15–19 yrs	96	0.75	41	1.43	1.90 (1.29–2.77)	14	0.44	0.59 (0.31–1.03)
20–64 yrs	1081	0.99	201	0.86	1.05 (0.90–1.22)	97	0.49	0.49 (0.39–0.61)
65+ yrs	657	2.42	45	0.83	0.34 (0.25–0.47)	41	2.36	0.97 (0.69–1.34)
Total	2090	11.40	401	11.13	0.98 (0.88–1.09)	236	6.83	0.60 (0.52–0.69)
Sex and age								
Males								
< 1 yrs	772	6.37†	269	8.77†	1.38 (1.19–1.58)	272	6.53†	1.02 (0.89–1.18)
1–4 yrs	72	1.56	46	4.14	2.65 (1.80–3.90)	35	2.32	1.49 (0.97–2.26)
5–9 yrs	23	0.36	10	0.63	1.73 (0.74–3.79)	5	0.27	0.75 (0.22–1.99)
10–14 yrs	36	0.54	15	0.97	1.80 (0.91–3.34)	5	0.31	0.57 (0.17–1.45)
15–19 yrs	59	0.90	31	2.14	2.37 (1.48–3.72)	12	0.71	0.79 (0.79–1.48)
20–64 yrs	624	1.08	106	1.17	1.08 (0.87–1.33)	63	0.61	0.56 (0.43–0.73)
65+ yrs	260	2.15	19	1.79	0.83 (0.83–1.33)	25	3.44	1.60 (1.02–2.42)
Total	1074	1.14	227	1.43	1.25 (1.08–1.45)	145	0.82	0.71 (0.60–0.85)
Females								
< 1 yrs	692	6.01†	273	9.18†	1.53 (1.32–1.76)	258	6.46†	1.07 (0.93–1.24)
1–4 yrs	66	1.51	24	2.23	1.48 (0.87–2.39)	28	1.94	1.28 (0.80–2.03)
5–9 yrs	30	0.50	7	0.46	0.91 (0.34–2.11)	6	0.34	0.68 (0.23–1.65)
10–14 yrs	29	0.46	12	0.80	1.72 (0.80–3.48)	5	0.32	0.70 (0.21–1.82)
15–19 yrs	37	0.59	10	0.71	1.20 (0.53–2.44)	2	0.13	0.22 (0.03–0.88)
20–64 yrs	457	0.78	85	0.82	1.05 (0.82–1.33)	34	0.36	0.46 (0.31–0.65)
65+ yrs	397	2.32	26	1.51	0.65 (0.42–0.97)	16	1.59	0.69 (0.39–1.13)
Total	1016	1.03	164	0.93	0.90 (0.76–1.07)	91	0.54	0.53 (0.42–0.65)

* Rates are per 100,000 population
** CI = confidence interval
† Infant mortality rates are per 10,000 live births
‡ The referent group is males
§ The referent group is non-Hispanic Whites
¶ RR = rate ratio

ethnicity. Significant differences were seen in mortality race/ethnicity among male decedents. Non-Hispanic Black males had the highest mortality (1.43 vs 1.14 for non-Hispanic Whites and .82 per 100,000 for Hispanics, Table 2). Forty-two percent of CHD-related mortality occurred during infancy for non-Hispanic White boys, in contrast to 54.2% for non-Hispanic Black boys and 65.2% for Hispanic boys. Non-Hispanic Black males had the highest mortality for all age groups except among persons aged >65 years. For children <1 year of age, non-Hispanic Blacks were 1.38 times more likely to have a CHD-related death than were non-Hispanic White infants (95% CI 1.19–1.58). For children aged 1–4, non-Hispanic Black boys had rates 2.65 times higher (95% CI 1.80–3.90) and Hispanic boys had rates 1.49 times

Table 3. CHD listed as the underlying and contributing causes of death for CHD decedents by ethnicity, US, 2000

	Underlying Cause of Death						Contributing Cause of Death					
	NH-White* (*n=3554*)		NH-Black (*n=933*)		Hispanic (*n=766*)		NH-White (*n=3554*)		NH-Black (*n=933*)		Hispanic (*n=766*)	
	n	%[a]	*n*	%[a]	*n*	%[a]	*n*	%[a]	*n*	%[a]	*n*	%[a]
1. Unspecified CMH†	761	21.4	235	25.2‡	197	25.7‡	1060	29.8	305	32.7	280	36.6‡
2. Hypoplastic left heart	245	6.9	76	8.2	90	11.8‡§	301	8.5	94	10.1	107	14.0‡§
3. Atrial septal defect	219	6.2	34	3.6‡	22	2.9‡	398	11.2	61	6.5‡	47	6.1‡
4. Ventricular septal defect	130	3.7	24	2.6	21	2.7	325	9.1	75	8.0	57	7.4
5. Malformation of the coronary vessels	108	3.0	53	5.7‡	12	1.6‡§	176	5.0	73	7.8‡	24	3.1‡§
6. Tetralogy of Fallot	122	3.4	29	3.1	20	2.6	169	4.8	46	4.9	40	5.2
7. Other specified CMH	78	2.2	21	2.3	14	1.8	128	3.6	30	3.2	20	2.6
8. Other congenital malformations of cardiac septa	78	2.2	9	1.0‡	6	0.8‡	116	3.3	12	1.3‡	8	1.0‡
9. Transposition of the great vessels			10	1.1‡	11	1.4	103	2.9	17	1.8‡	16	2.1
10. Insufficient aortic valve	76	2.1	4	0.4‡	12	1.6§	137	3.9	9	1.0‡	15	2.0‡
11. Coarctation of the aorta	44	1.2	12	1.3	14	1.8	105	3.0	28	3.0	36	4.7‡
12. Atrioventricular septal defect	47	1.3	13	1.4	6	0.8	84	2.4	23	2.5	15	2.0
13. Congenital stenosis of the aortic valve	50	1.4	4	0.4‡	9	1.2	80	2.3	7	0.8‡	14	1.8
14. Common arterial trunk	38	1.1	14	1.5	11	1.4	46	1.3	19	2.0	14	1.8
15. Atresia of the pulmonary artery	28	0.8	17	1.8‡	8	1.0	68	1.9	38	4.1‡	19	2.5

* NH = Non-Hispanic.
† CMH = congenital malformations of the heart.
‡ $P<.05$ (significantly different from NH-White) Z-test for difference between proportions.
§ $P<.05$ (significantly different from NH-Black) Z-test for difference between proportions.
[a] Calculated by dividing the number of deaths by the number of decedents in that racial/ethnic group.

higher than non-Hispanic White boys (95% CI .97–2.26). Non-Hispanic Black boys aged 15–19 had rates 2.37 times (95% CI 1.48–3.72) higher than did non-Hispanic Whites. Fewer than 15 decedents were recorded among non-Hispanic Black and Hispanic boys aged 5–9 and 10–14 years old and Hispanic boys 15–19 years old; therefore estimates in these age groups are unstable. Hispanic men aged 20–64 were .56 times less likely to have a CHD-related death than were non-Hispanic White men of the same age (95% CI .43–.73). However, Hispanic men >65 years of age were 1.60 times more likely to have a CHD-related death than were non-Hispanic Whites of similar age (95% CI 1.02–2.42).

During infancy, leading underlying causes of death were the same for non-Hispanic White, non-Hispanic Black, and Hispanic boys with CHD (UCMH and hypoplastic left heart syndrome) (Table 4). UCMH was more often the underlying cause of death for non-Hispanic White and Hispanic boys aged <5 than for non-Hispanic Black boys of the same age. For boys aged 5–9, UCMH was the leading CHD cause of death for all racial/ethnic groups, but data in these age groups were sparse. Although UCMH was the underlying cause of death for 80% of these boys aged 10–14, data were sparse (<15 decedents).

Females

Forty percent of CHD-related mortality occurred during infancy for non-Hispanic White girls, in contrast to 62.5% for non-Hispanic Black and 73.9% for Hispanic girls. Non-Hispanic Black infant girls were more likely to have a CHD-related death than were non-Hispanic White infant girls (RR 1.53, 95% CI 1.32–1.76). A similar pattern was seen for non-Hispanic Black girls aged 1–4 years, but it was not significant (RR 1.48, 95% CI .87–2.39). For children aged <5, Hispanics had a greater proportion of deaths (81.9%) than did non-Hispanic White (44.4%) and non-Hispanic Black (68.0%) girls. Hispanic adolescent girls aged 15–19 were .22 times as likely to have a CHD-related death as non-Hispanic White teen girls (95% CI .03–.88). However, overall Hispanic females had lower CHD-related mortality than did non-Hispanic White females (RR .53, 95% CI .42–.65).

UCMH and hypoplastic left heart syndrome were the leading causes of death for non-Hispanic White, non-Hispanic Black, and Hispanic infant girls with CHD (Table 5). UCMH was the leading cause of death for all girls aged 1–4, regardless of race/ethnicity. Among girls aged 10–14, UCMH was the underlying cause of death for 58% of CHD deaths among non-Hispanic Black girls. Data were sparse (<15 decedents) in the 10–14 and 15–19 age groups for non-Hispanic Blacks, which limits our ability to make inferences for these age groups. Overall, the contribution of UCMH to mortality among females decreased as age increased after 19 years for all ethnic groups.

Table 4. Top three underlying causes of death on death certificates for male CHD decedents in each age group by ethnicity, US, 2000

	Non-Hispanic White *(n=1846)*		Non-Hispanic Black *(n=496)*		Hispanic *(n=417)*	
Age	**Defect (ICD-10 Code)**	**%**	**Defect (ICD-10 Code)**	**%**	**Defect (ICD-10 Code)**	**%**
Infant	1. Unspecified CMH† (Q24.9)	27.9	1. Unspecified CMH † (Q24.9)	24.2	1. Unspecified CMH† (Q24.9)	26.1
	2. Hypoplastic left heart (Q23.4)	18.3	2. Hypoplastic left heart (Q23.4)	13.8	2. Hypoplastic left heart (Q23.4)	16.5
	3. Trisomy 18 (Q91.3)	3.4	3. Trisomy 18 (Q91.3)	6.0	3. Trisomy 18 (Q91.3)	5.1
1–4 yrs	1. Unspecified CMH (Q24.9)	38.9	1. Unspecified CMH (Q24.9)	26.1	1. Unspecified CMH (Q24.9)	45.7
	2. Tetralogy of Fallot (Q21.3)	8.3	2. Hypoplastic left heart (Q23.4)	8.7	2. Ventricular septal defect (Q21.0)	5.7
	3. Hypoplastic left heart (Q23.4)	4.2	3. Stenosis of pulmonary artery (Q25.6)	6.5	3. Hypoplastic left heart (Q23.4)	5.7
	3. Atrioventricular septal defect (Q21.2)	4.2	3. Trisomy 21 (Q90.9)	6.5		
5–9 yrs	1. Unspecified CMH (Q24.9)	38.9	1. Unspecified CMH (Q24.9)	60.0*	1. Unspecified CMH (Q24.9)	40.0*
	2. Other specified (Q24.8)	8.7	2. –		2. –	
	3. –		3. –		3. –	
10–14 yrs	1. Unspecified CMH (Q24.9)	44.4	1. Malformation coronary vessels (Q24.5)	33.3*	1. Unspecified CMH (Q24.9)	80.0*
	2. –		2. Unspecified CMH (Q24.9)	33.3*	2. Double inlet ventricle (Q20.4)	20.0*
	3. –		3. –		3. –	
15–19 yrs	1. Unspecified CMH (Q24.9)	27.1	1. Malformation coronary vessels (Q24.5)	35.5	1. Double outlet right ventricle (Q20.1)	16.7*
	2. Transposition of great vessels (Q20.3)	8.5	2. Unspecified CMH (Q24.9)	35.5	2. Unspecified CMH (Q24.9)	16.7*
	3. Trisomy 21 (Q90.9)	5.1	3. –		3. –	
20–64 yrs	1. Unspecified CMH (Q24.9)	19.2	1. Unspecified CMH (Q24.9)	17.9	1. Insufficient aortic valve (Q23.1)	14.3
	2. Tetralogy of Fallot (Q21.3)	6.9	2. Malformation coronary vessels (Q24.5)	17.0	2. Unspecified CMH (Q24.9)	12.7
	3. Malformation coronary vessels (Q24.5)	6.4	3. –		3. Malformation coronary vessels (Q24.5)	11.1
65+ yrs	1. Atrial septal defect (Q21.1)	21.5	1. Atrial septal defect (Q21.1)	31.6*	1. Atrial septal defect (Q21.1)	31.3
	2. Ventricular septal defect (Q21.0)	10.4	2. –		2. –	
	3. Acute myocardial infarction (I21.9)	8.5	3. –		3. –	

* Total *n* in age group < 20.
† CMH = congenital malformations of the heart.

DISCUSSION

Our study updates previous research[14] and describes CHD-related mortality in the United States. We described the annual contribution of CHD to mortality during childhood and adulthood and the causes of death among CHD decedents by age, sex, and race/ethnicity. As expected, CHD-related mortality was highest during infancy and for children aged 1–9 and then dropped precipitously and remained stable throughout adulthood. Although CHD-related deaths were rare, we observed racial/ethnic disparities in CHD-related mortality and causes of death, particularly between non-Hispanic Blacks and non-Hispanic Whites. Disparities were most evident for non-Hispanic Black boys aged 1–4 and 15–19, who experienced more than a twofold increase in mortality compared with non-Hispanic Whites boys of similar age. The disparity was smaller (1.5-fold) between non-Hispanic Black girls aged 1–4 and 10–14 and non-Hispanic White girls in the same age groups. In contrast, we observed no significant differences in CHD-related

Table 5. Top three underlying causes of death on death certificates for female CHD decedents in each age group by ethnicity, US, 2000

	Non-Hispanic White *(n=1708)*		Non-Hispanic Black *(n=437)*		Hispanic *(n=349)*	
Age	**Defect (ICD-10 Code)**	**%**	**Defect (ICD-10 Code)**	**%**	**Defect (ICD-10 Code)**	**%**
Infant	1. Unspecified CMH† (Q24.9)	25.0	1. Unspecified CMH† (Q24.9)	27.1	1. Unspecified CMH† (Q24.9)	27.1
	2. Hypoplastic left heart (Q23.4)	13.7	2. Hypoplastic left heart (Q23.4)	12.5	2. Hypoplastic left heart (Q23.4)	15.9
	3. Trisomy 18 (Q91.3)	7.2	3. Trisomy 18 (Q91.3)	7.0	3. –	
1–4 yrs	1. Unspecified CMH (Q24.9)	33.3	1. Unspecified CMH (Q24.9)	58.3	1. Unspecified CMH (Q24.9)	50.0
	2. –		2. Tetralogy of Fallot	8.3	2. Trisomy 21 (Q90.9)	7.1
	3. –		3. –		3. –	
5–9 yrs	1. Unspecified CMH (Q24.9)	26.7	1. Conjoined twins (Q89.4)	28.6*	1. –	–*
	2. Tetralogy of Fallot (Q21.3)	10.0	2. –		2. –	
	3. Hypoplastic left heart (Q23.4)	10.0	3. –		3. –	
10–14 yrs	1. Unspecified CMH (Q24.9)	31.0	1. Unspecified CMH (Q24.9)	58.3*	1. –	–*
	2. Ventricular septal defect (Q21.0)	10.3	2. Malformation of coronary vessels (Q24.5)	16.7*	2. –	
	3. Malformation coronary vessels (Q24.5)	10.3	3. –		3. –	
15–19 yrs	1. Unspecified CMH (Q24.9)	40.5	1. Unspecified CMH (Q24.9)	40.0*	1. –	–*
	2. Malformation coronary vessels (Q24.5)	8.1	2. Malformation of coronary vessels (Q24.5)	30.0	2. –	
	3. Ventricular septal defect (Q21.0)	5.4	3. –		3. –	
	3. Other congenital malformation of cardiac septa (Q21.8)	5.4				
20–64 yrs	1. Unspecified CMH (Q24.9)	19.9	1. Unspecified CMH (Q24.9)	17.7	1. Unspecified CMH (Q24.9)	26.5
	2. Other congenital malformation of cardiac septa (Q21.8)	8.5	2. Malformation of coronary vessels (Q24.5)	11.8	2. Other congenital malformation of cardiac septa (Q21.8)	8.8
	3. Trisomy 21 (Q90.9)	6.6	3. Atrial septal defect (Q21.1)	8.2	3. –	
65+ yrs	1. Atrial septal defect (Q21.1)	27.0	1. Atrial septal defect (Q21.1)	19.2	1. Atrial septal defect (Q21.1)	24.0*
	2. Acute myocardial infarction, (I21.9)	10.6	2. Unspecified CMH (Q24.9)	7.7	2. Acute myocardial infarction (I21.9)	20.0*
	3. Ventricular septal defect (Q21.0)	6.8	3. Hypertensive heart no heart failure (I11.9)	11.5	3. –	

* Total *n* in age group < 20.
† CMH = congenital malformations of the heart.

mortality between non-Hispanic Whites and Hispanics.

The racial/ethnic disparities observed in our study have several possible explanations. Since mortality rates are determined by both the incidence of disease and the case-fatality associated with it, non-Hispanic Blacks may have had higher CHD-related mortality because of higher incidence of CHD or higher CHD case-fatality rates compared with non-Hispanic Whites. Unfortunately, true CHD incidence is difficult to determine because of early spontaneous miscarriages, fetal deaths, and elective terminations. Thus, "prevalence at live birth" is often used as a proxy. No information is available on racial/ethnic differences in rates of spontaneous miscarriages or fetal deaths for CHD-affected pregnancies. Prenatal diagnosis of CHD can influence elective termination and prevalence at live birth.[16] Live birth prevalence estimates do not show a higher rate among Blacks than among Whites for specific CHDs that were the most frequent underlying causes of death in our study, except for tetralogy of Fallot[17] or for atrial septal defect.[1] Another explanation, that case severity is greater among Blacks than among Whites, is also not supported by the literature.

Although previous research is not extensive, differences in case-fatality is another possible explanation for the racial disparities observed in our study. Survival of infants with CHD has greatly improved during the past three

As expected, CHD-related mortality was highest during infancy and for children aged 1–9 and then dropped precipitously and remained stable throughout adulthood.

to four decades, with significant decreasing trends in perioperative mortality.[18] Case-fatality rates however, vary by length of followup and by type of CHD.[18–20] In one study, non-Hispanic Black children had significantly higher risk of death after CHD surgery than did non-Hispanic Whites, but the risk was not present for all Blacks and was highly variable by state and region.[21]

Another possible explanation for a difference in case-fatality is racial/ethnic disparities in access to medical care. Recent studies indicate racial/ethnic disparities in access to pediatric and adult care and treatment received even when care is obtained.[22–24] Blacks may be less likely than Whites to have adequate care or have their CHD diagnosed, treated, or repaired early in childhood. Some studies suggest that prenatal diagnosis of CHD leads to better management, earlier surgical intervention, and improved short- and long-term outcomes for children with CHD.[25–27]

A strength of our study is use of national multiple-cause mortality data to determine mortality rates and the ability to assess both the underlying and contributing causes of death. A limitation of our study is our use of cross-sectional data rather than birth cohort data. Thus, we were only able to calculate CHD-related mortality in 2000 rather than actual survival rates. As such, our data do not adjust for the potential time trend, period, or cohort effects. Moreover, our results are also influenced by temporal variations in incidence, prevalence at live birth, and treatment. There is also the potential effect of decreased age at operation and changes in surgical and management techniques[18] over time.

Another limitation of our study is our ascertainment of cause of death information from death certificates rather than medical records or autopsy reports. Although cause of death information is collected on highly structured and standardized forms in the United States, validation studies have documented improper completion of death certificates and discrepancies between cause of death reported on death certificates and cause of death recorded on medical records and autopsy reports.[28–31] Validation studies show inconsistent results for the sensitivity, specificity, and positive predictive value between death certificates and medical records for cause of death.[28,32,33] There is also potential for misclassification in the assignment of the underlying and contributing causes of death (ie, differentiating between dying *from* a condition and dying *with* a condition).

Another concern in this study is whether or not there is racial/ethnic differential ascertainment of causes of death for persons with CHD, which would explain the differences we observe in CHD-related mortality rates. We are unaware of any previous studies that have demonstrated racial/ethnic differential ascertainment and documentation of CHD-related causes of death. Thus, it is difficult to determine if the current findings are accurate estimates of the true death rates for non-Hispanic Whites and non-Hispanic Blacks. Nevertheless, our study is an attempt to understand this complex issue.

Conclusions

The implications of our findings are mixed. Mortality and causes of death among children and adults with CHD in the United States have not been well studied at the population level. CHD continues to be an incident illness, producing substantial infant and childhood mortality. We have replicated racial/ethnic disparities described elsewhere. [14] These racial/ethnic differences in CHD mortality present a wholly different and more challenging conundrum. The differences we observed may be largely due to poor reporting of cause of death information on death certificates; however, since CHD has complex roots, these findings may also arise from a host of other unexplored areas such as differential nutritional health in the parents, disparities in prenatal obstetrical services, and access to sophisticated fetal ultrasound. More studies are needed to better understand differences among ethnic groups regarding CHD-related mortality.

REFERENCES

1. Botto LD, Correa A, Erickson JD. Racial and temporal variations in the prevalence of heart defects. *Pediatrics*. 2001;107(3):e32–e39.
2. Carlgren LE, Ericson A, Kallen B. Monitoring of congenital cardiac defects. *Pediatr Cardiol*. 1987;8(4):247–256.
3. Fernandes ED, Kadivar H, Hallman G, Reul GJ, Ott DA, Cooley DA. Congenital malformations of the coronary arteries: The Texas Heart Institute experience. *Ann Thorac Surg*. 1992;54:732–740.
4. Hanna EJ, Nevin NC, Nelson J. Genetic study of congenital heart defects in Northern Ireland (1974–1978). *J Med Genet*. 1994;31(11): 858–863.
5. Yoon PW, Olney RS, Khoury MJ, Sappenfield WM, Chavez GF, Taylor D. Contribution of birth defects and genetic diseases to pediatric hospitalizations. A population-based study. *Arch Pediatr Adolesc Med*. 1997;151(11): 1082–1083.
6. Nuutinen M, Koiyu M, Rantakallio P. Long-term outcome for children with congenital heart defects. A study from 1 year birth cohort born in 1966 in Northern Finland. *Arct Med Res*. 1989;48:175–184.
7. Tegnander E, Williams W, Johansen OJ, Blaas HG, Eik-Nes SH. Prenatal detection of heart defects in a non-selected population of 30,149 fetuses—detection rates and outcome. *Ultrasound Obstet Gynecol*. 2006;27:252–265.
8. Armstrong BEM. Congenital cardiovascular disease and cardiac surgery in childhood: Part 2. Acyanotic congenital heart defects and interventional techniques. *Curr Opin Cardiol*. 1995;10(1):68–77.

9. Cohen D. Surgical management of congenital heart disease in the 1990's. *Am J Dis Chil*. 1992;146:1447–1452.
10. Perolo A, Prandstraller D, Ghi T, et al. Diagnosis and management of fetal cardiac anomalies: 10 years of experience at a single institution. *Ultrasound Obstet Gynecol*. 2001;18(6):615–618.
11. Tweddell JS, Hoffman GM. Postoperative management in patients with complex congenital heart disease. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu*. 2002;5:187–205.
12. Eskedal L, Skjalg Hagemo P, Eskild A, Aamodt G, Seiler KS, Thaulow E. Survival after surgery for congenital heart defects: does reduced early mortality predict improved long-term survival? *Acta Paediatrica*. 2005;94(4): 438–443.
13. Wernovsky G, Wypij D, Jonas RA, et al. Postoperative course and hemodynamic profile after the arterial switch operation in neonates and infants : a comparison of low-flow cardiopulmonary bypass and circulatory arrest. *Circulation*. 1995;92(8):2226–2235.
14. Boneva RS, Botto LD, Moore CA, Yang Q, Correa A, Erickson JD. Mortality associated with congenital heart defects in the United States: trends and racial disparities, 1979–1997. *Circulation*. 2001;103(19):2376–2381.
15. Seifert HA, Howard DL, Silber JH, Jobes DR. Female gender increases the risk of death during hospitalization for pediatric cardiac surgery. *J Thorac Cardiovasc Surg*. 2007; 133(3):668–675.
16. Stoll C, Alembik Y, Dott B, Roth MP. Impact of prenatal diagnosis on livebirth prevalence of children with congenital anomalies. *Ann Genet*. 2002;45(3):115–121.
17. Pradat P, Francannet C, Harris JA, Robert E. The epidemiology of cardiovascular defects, part I: a study based on data from three large registries of congenital malformations. *Pediatr Cardiol*. 2003;24(3):195–221.
18. Morris CD, Menashe VD. 25-year mortality after surgical repair of congenital heart defect in childhood. A population based cohort. *JAMA*. 1991;266(24):3447–3452.
19. Cleves MA, Ghaffar S, Zhao W, Mosely BS, Hobbs CA. First-year survival of infants born with congenital heart defects in Arkansas (1993–1998): a survival analysis using registry data. *Birth Defects Res A Clin Mol Teratol*. 2003;67:662–668.
20. Nembhard WN, Waller DK, Sever LE, Canfield MA. Patterns of first-year survival among infants with selected congenital anomalies in Texas, 1995–1997. *Teratology*. 2001; 64(5):267–275.
21. Benavidez OJ, Gauvreau K, Jenkins KJ. Racial and ethnic disparities in mortality following congenital heart surgery. *Pediatr Cardiol*. 2006;27(3):321–328.
22. Flores G, Fuentes-Afflick E, Barbot O, et al. The health of Latino children: urgent priorities, unanswered questions, and a research agenda. *JAMA*. 2002;288:82–90.
23. Ronsaville DS, Hakim RB. Well child care in the United States: racial differences in compliance with guidelines. *Am J Public Health*. 2000;90:1436–1443.
24. Furth SL, Garg PP, Neu AM, Hwang W, Fivush BA, Powe NR. Racial differences in access to the kidney transplant waiting list for children and adolescents with end-stage renal disease. *Pediatrics*. 2000;106(4):756–761.
25. Bonnet D, Coltri A, Butera G, et al. Detection of transposition of the great arteries in fetuses reduces neonatal morbidity and mortality. *Circulation*. 1999;99(7):916–918.
26. Fuchs IB, Muller H, Abdul-Khaliq H, Harder T, Dudenhausen W, Henrich W. Immediate and long-term outcomes in children with prenatal diagnosis of selected isolated congenital heart defects. *Ultrasound Obstet Gynecol*. 2007;29:38–43.
27. Tworetzky W, McElhinney DB, Reddy VM, Brook MM, Hanley FL, Silverman NH. Improved surgical outcome after fetal diagnosis of hypoplastic left heart syndrome. *Circulation*. 2001;103(9):1269–1273.
28. Coady SA, Sorlie PD, Cooper LS, Folsom AR, Rosamond WD, Conwill DE. Validation of death certificate diagnosis for coronary heart disease: the Atherosclerosis Risk in Communities (ARIC) Study. *J Clin Epidemiol*. 2001;54(1):40–50.
29. Gillum RF. On death certification and coding for ischemic heart disease. *Am J Epidemiol*. 1984;119(4):650.
30. Johansson LA, Westerling R, Rosenberg HM. Methodology of studies evaluating death certificate accuracy were flawed. *J Clin Epidemiol*. 2006;59(2):125–131.
31. Kircher T, Nelson J, Burdo H. The autopsy as a measure of accuracy of the death certificate. *N Engl J Med*. 1985;313(20):1263–1269.
32. Goraya TY, Jacobsen SJ, Belau PG, Weston SA, Kottke TE, Roger VL. Validation of death certificate diagnosis of out-of-hospital coronary heart disease deaths in Olmsted County, Minnesota. *Mayo Clin Proc*. 2000;75(7): 681–687.
33. Iribarren C, Crow RS, Hannan Mstat PJ, David R, Luepker RV. Validation of death certificate diagnosis of out-of-hospital sudden cardiac death. *Am J Cardiol*. 1998;82(1): 50–53.

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