HHS Public Access

Author manuscript

Circulation. Author manuscript; available in PMC 2024 August 15.

Published in final edited form as:

Circulation. 2023 August 15; 148(7): 575–588. doi:10.1161/CIRCULATIONAHA.123.064400.

Survival From Birth Until Young Adulthood Among Individuals With Congenital Heart Defects: CH STRONG

Karrie F. Downing, MPH,

Wendy N. Nembhard, PhD, MPH,

Charles E. Rose, PhD,

Jennifer G. Andrews, PhD,

Anthony Goudie, PhD,

Scott E. Klewer, MD,

Matthew E. Oster, MD, MPH,

Sherry L. Farr, PhD

National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention, Atlanta, GA (K.F.D., C.E.R., M.E.O., S.L.F.). Department of Epidemiology, Fay W. Boozman College of Public Health and the Arkansas Center for Birth Defects Research and Prevention, University of Arkansas for Medical Sciences, Little Rock (W.N.N.). Department of Pediatrics, University of Arizona, Tucson (J.G.A., S.E.K.). Department of Pediatrics, Center for Applied Research and Evaluation, College of Medicine, Little Rock, AR (A.G.). Children's Healthcare of Atlanta and Emory University School of Medicine, Atlanta, GA (M.E.O.).

Abstract

BACKGROUND: Limited population-based information is available on long-term survival of US individuals with congenital heart defects (CHDs). Therefore, we assessed patterns in survival from birth until young adulthood (ie, 35 years of age) and associated factors among a population-based sample of US individuals with CHDs.

METHODS: Individuals born between 1980 and 1997 with CHDs identified in 3 US birth defect surveillance systems were linked to death records through 2015 to identify those deceased and the year of their death. Kaplan-Meier survival curves, adjusted risk ratios (aRRs) for infant mortality (ie, death during the first year of life), and Cox proportional hazard ratios for survival after the first year of life (aHRs) were used to estimate the probability of survival and associated factors. Standardized mortality ratios compared infant mortality, >1-year mortality, >10-year mortality, and >20-year mortality among individuals with CHDs with general population estimates.

Correspondence to: Karrie F. Downing, MPH, 4770 Buford Hwy, Mailstop S106-3, Atlanta, GA 30341. yyx9@cdc.gov.

Disclosures

None.

The findings and conclusion in this article are those of the authors and do not necessarily represent the official position of the Centers for Disease Control and Prevention. V. Villamil replicated the analysis.

Supplemental Material

Tables S1–S4

Figures S1–S4

Supplemental Material is available at: https://www.ahajournals.org/doi/suppl/10.1161/CIRCULATIONAHA.123.064400

RESULTS: Among 11 695 individuals with CHDs, the probability of survival to 35 years of age was 81.4% overall, 86.5% among those without co-occurring noncardiac anomalies, and 92.8% among those who survived the first year of life. Characteristics associated with both infant mortality and reduced survival after the first year of life, respectively, included severe CHDs (aRR=4.08; aHR=3.18), genetic syndromes (aRR=1.83; aHR=3.06) or other noncardiac anomalies (aRR=1.54; aHR=2.53), low birth weight (aRR=1.70; aHR=1.29), and Hispanic (aRR=1.27; aHR=1.42) or non-Hispanic Black (aRR=1.43; aHR=1.80) maternal race and ethnicity. Individuals with CHDs had higher infant mortality (standardized mortality ratio=10.17), >1-year mortality (standardized mortality ratio=3.29), and >10-year and >20-year mortality (both standardized mortality ratios ≈1.5) than the general population; however, after excluding those with noncardiac anomalies, >1-year mortality for those with nonsevere CHDs and >10-year and >20-year mortality for those with any CHD were similar to the general population.

CONCLUSIONS: Eight in 10 individuals with CHDs born between1980 and 1997 survived to 35 years of age, with disparities by CHD severity, noncardiac anomalies, birth weight, and maternal race and ethnicity. Among individuals without noncardiac anomalies, those with nonsevere CHDs experienced mortality between 1 and 35 years of age, similar to the general population, and those with any CHD experienced mortality between 10 and 35 years of age, similar to the general population.

Keywords

adult; epidemiology; heart defects; congenital; mortality; survival

Congenital heart defects (CHDs) are the most common type of birth defect, affecting ≈1% of live births,[1,2] and are the leading cause of birth defect–related mortality in the United States.[3] However, due to advancements in medical and surgical management of CHDs, mortality rates have been declining,[4] and >90% of individuals with CHDs in the United States are expected to survive to adulthood.[5] Despite improvements in survival, several non-US studies suggest that those living with CHDs, especially severe CHDs, continue to experience higher rates of mortality than the general population.[3,5–10] However, due to different cultures, health care systems, and health care accessibility, survival and mortality estimates from other countries may not be transportable to the US population.

A few studies have investigated CHD survival using US data. Spector et al[11] linked patient data from the Pediatric Cardiac Care Consortium, a multicenter registry of pediatric patients undergoing cardiac surgery, to death records through 2014 from the National Death Index (NDI). They found elevated mortality (standardized mortality ratio=8.3) compared with the US general population. Another study by Oster et al[12] identified CHD cases from a single birth defects registry in metropolitan Atlanta, GA, and linked them to state and NDI records. They reported that survival up to 27 years of age differed by CHD severity (comparing critical versus noncritical CHDs) in unadjusted analyses, but they did not compare overall mortality with that of the general population. A few studies have also examined trends in CHD mortality (deaths with a CHD documented as a cause of death) using US death record data.[13–17] These studies noted that CHD mortality rates have been declining over time, and that disparities in CHD mortality persist by race, ethnicity, and sex, but they

could not include individuals with CHDs who died of other causes who did not have CHDs documented on their death certificates.

Previous literature describing survival among those with CHDs has been limited to individuals receiving surgery for their CHDs, born in a single metropolitan area, or with CHD only identified from their death certificate. Therefore, using data collected during the CH STRONG project (Congenital Heart Survey to Recognize Outcomes, Needs, and Well-Being), our objective was to assess patterns in survival from birth until young adulthood and related factors and disparities among a population-based sample of individuals with CHDs born in 3 locations in the United States.

METHODS

Requests to access the data set from qualified researchers trained in human subject confidentiality protocols may be sent to the Centers for Disease Control and Prevention (CDC) at chstrong@cdc.gov.

Study Population From Population-Based Birth Defects Surveillance Systems

Individuals with CHDs were identified through population-based birth defects surveillance systems with active case-finding methods in Arizona, Arkansas, and Atlanta, GA, for the CDC-funded CH STRONG. Detailed methods of CH STRONG have been published previously.[18] In brief, individuals born between 1980 and 1997 in the catchment area of each surveillance system were included if they had ≥1 CHD diagnosis codes (defined as 6-digit CDC-modified version of the British Paediatric Association [CDC/BPA] codes between 745.000 and 747.9XX, excluding patent foramen ovale and some non-specific codes; Table S1).

These birth defects surveillance systems also collected data on diagnoses of other congenital anomalies (hereafter referred to as noncardiac anomalies) at the time of birth and up to 1 year (Arizona), 2 years (Arkansas), and 6 years (Georgia) there-after. Those with noncardiac anomalies were further grouped into those with genetic syndromes (any CDC/BPA diagnoses codes between 758.000 and 758.999, including Down syndrome) and those with other noncardiac anomalies (all other CDC/BPA diagnosis codes outside of 745.000–745.9XX captured by the birth defect surveillance systems; eg, cleft lip-bilateral without cleft palate). In addition to diagnoses, information on year of birth, sex, birth weight, plurality, and maternal race and ethnicity were collected.

CHD Severity and Primary CHD Diagnosis

Using a previously published algorithm modified by CH STRONG clinicians for use with CDC/BPA codes,[19] CHD severity was categorized by diagnosis code classification from most to least severe as: severe; shunt + valve; shunt (without valve); valve (without shunt); or other. Codes and lesion descriptions corresponding to each category can be found in Table S1 (eg, lesions classified as severe include single ventricle, coarctation of the aorta, and tetralogy of Fallot). For some analyses, severity was further collapsed into severe CHDs, and all other categories were combined as nonsevere CHDs. Each individual was assigned a primary CHD diagnosis. For individuals with multiple CHDs in different severity categories,

the most severe defect was considered their primary diagnosis. If individuals had multiple CHDs of similar severity, CH STRONG congenital cardiologists reviewed the case diagnosis codes to determine the primary diagnosis. If the clinicians determined that the individual had >1 primary diagnosis, then the primary diagnosis was categorized as “multiple” regardless of severity of the defects.

Linkage to Death Records

Each surveillance system linked the identified individuals with CHDs to their respective state death records through December 31, 2015, with manual verification. Linkage variables included subject name, sex, date of birth, and parent name(s). To collect out-of-state deaths, Arizona additionally linked to the NDI through 2015, and Atlanta linked to the NDI through 2008. Arkansas Vital Records collected out-of-state deaths through an interstate exchange agreement. Individuals who were not linked to a death record were presumed to be alive and recruited to participate in a survey used in other CH STRONG analyses.[20–22] For those determined to be deceased, only the year of death was provided by the surveillance systems. Due to institutional review board restrictions, more detailed information on date of death could not be shared across sites for inclusion in the CH STRONG data set.

Linkage to Decennial Census Data

Sites geocoded the residential address documented in each birth defect surveillance system to identify county of birth for individuals with CHDs, and then sites linked county-level information from the decennial census (1980 or 1990) occurring nearest the individual’s birth year. The following county-level information was obtained from the decennial census data and linked to each individual according to their county at birth: percentage of families with children <18 years of age in the county that were living in poverty; percentage of individuals ≥16 years of age in the county who were unemployed; percentage of individuals ≥25 years of age in the county who completed a high school degree or higher; median household income of the county; and percentage of the county population living in a rural area (as defined by the US Census Bureau at https://www.census.gov/programs-surveys/geography/guidance/geo-areas/urban-rural.html).

We created a county deprivation index for each individual using linked census information available in CH STRONG data for 4 county-level socioeconomic status indicators (percentage family poverty, unemployment, high school education attainment, and median household income). Z scores were calculated for each indicator by subtracting the mean value for all counties of birth in CH STRONG from each individual’s specific value for that indicator and dividing this by the standard deviation. Z score values for characteristics negatively associated with county deprivation (ie, median household income and high school education) were multiplied by −1 to change the direction of association, after which all 4 Z score estimates were summed to a singular county deprivation index value for that person. Finally, individuals were categorized into tertiles based on their county deprivation index value, with tertile 1 being the least deprived and tertile 3 the most deprived.

Analyses

Characteristics among individuals determined to be deceased and those presumed to be alive were described in frequencies and percentages. Year of birth was subtracted from year of death to estimate age at death in years, and individuals presumed to be alive were censored from analyses as of December 31, 2015. Kaplan-Meier survival estimates by age of death through 35 years were calculated and plotted overall and by CHD severity and primary CHD diagnosis. Estimates conditional on survival to 1, 10, and 20 years of age were also calculated and plotted overall and by CHD severity. Kaplan-Meier survival estimates were also calculated and plotted by all other included characteristics from the birth defects surveillance system for individuals who survived to at least 1 year of life.

Of CH STRONG observations, 18.2% were missing data for characteristics of interest (ie, year of death n=357; sex n=17; birth weight n=728; maternal race=1131; and any county-level census data n=580). To reduce potential bias in parameter estimation, strengthen the generalizability of the results, increase statistical power, and decrease standard errors,[23] the mice package[24] of R software was used to conduct multiple imputation by chained equations. Multiple imputation using classification and regression trees was conducted with 20 imputations over 5 iterations using all included characteristics in the analysis. The imputed data were pooled and used for adjusted analyses accounting for the multiple imputed data sets.

Using pooled imputed data, characteristics associated with death within the first year of life were assessed using multivariable Poisson models to calculate adjusted risk ratios (aRRs) and 95% CIs, overall and by CHD severity. For those who survived beyond the first year of life, adjusted hazard ratios (aHRs) and 95% CIs were calculated using Cox proportional hazards models after assessing the proportional hazards assumption by visual inspection of the log-log survival curves and evaluating the correlation between Schoenfield residuals and the ranking of individual survival times. Using directed acyclic graphs (which convey directional causal associations between variables based on previous literature and biological mechanisms) exclusively for covariate selection, the same sets of covariates were included in the Cox proportional hazards models as in Poisson models. In a supplemental analysis, we added an interaction term between birth year and race and ethnicity to the Poisson model for birth year to determine whether first-year survival differentially improved over time for non-Hispanic (NH) Black and NH White infants. For sensitivity analyses, we examined aRRs and aHRs using complete case (ie, nonimputed) analyses. We also examined aHRs stratified by CHD severity, stratified by site, and excluded those who also had noncardiac anomalies.

Similar to the methodology described and implemented in a previous publication,[11] standardized mortality ratios (SMRs) and 95% CIs were calculated using US death data from the National Vital Statistics System (NVSS) accessed through CDC WONDER, including age-, sex-, year-, race and ethnicity–, and site-specific crude death rates per 100 000 people for the catchment area of each surveillance system. SMRs were calculated for deaths in the first year of life and, separately, deaths occurring between 1 and 35 years of age by comparing observed deaths among individuals with CHDs from CH STRONG with expected deaths based on NVSS rates for the general population. Because only year

of birth and year of death were available in CH STRONG, date of birth and date of death were assumed to have occurred halfway through a given year. Expected death rates were then calculated at the individual level by multiplying the proportion of time (in half-year increments) that each individual belonged to an age-, sex-, year-, race and ethnicity– and site-specific strata by the respective mortality rate for each stratum. All strata-specific rates for a given individual were summed to obtain a person-level probability of death. All person-level probabilities were then summed to obtain an overall expected number of deaths. SMRs are presented overall by CHD severity, race and ethnicity, and sex. We also examined SMRs, excluding those who also had noncardiac anomalies. For NVSS data preceding 1999, information on Hispanic ethnicity was unavailable; therefore, we only present SMRs for Black and White races.

CH STRONG was approved by CDC and University of Arkansas for Medical Sciences institutional review boards. The University of Arizona deferred to the CDC institutional review board. SMRs were calculated using SAS 9.4 software (SAS Institute Inc., Cary, NC) and OpenEpi: Open Source Epidemiologic Statistics for Public Health.[25] All other data were analyzed using R Studio software.[26]

RESULTS

Among the 11 695 individuals born in Arkansas (42.1%), Arizona (29.1%), and Metropolitan Atlanta (28.8%) between 1980 and 1997 and identified with a CHD, 2372(20.3%) were determined to be deceased by December 31, 2015 (Table 1). Of those presumed to be alive, 29.0% had severe CHDs and 38.2% had co-occurring syndromes or noncardiac anomalies, whereas 66.1% of those deceased had severe CHDs, and 57.4% had co-occurring syndromes or noncardiac anomalies. In addition, <1% of those presumed to be alive had hypoplastic left heart syndrome as their primary CHD diagnosis, compared with 14.2% of those who were deceased. Other health and sociodemographic characteristics by vital status can be found in Table 1.

Among individuals with CHDs in CH STRONG, the largest drop in survival occurred between birth and 1 year of age, such that survival percentage at 1 year was 85.0% (Figure 1A; Table S2); when excluding those who also had noncardiac anomalies, survival percentage at 1 year was slightly higher, at 89.0%. After 1 year of age, the decline in the survival curve to 35 years became more gradual, such that survival percentage at 35 years of age was 81.4% overall (and 86.5% when excluding those who also had noncardiac anomalies). Those with severe CHDs had the sharpest decline in survival compared with those with CHDs of other severity types (Figure 1B; Table S2), which is largely driven by individuals with hypoplastic left heart syndrome (21.5% survival at 35 years of age), other single-ventricle defects (53.5% survival at 35 years of age), tricuspid atresia with pulmonary stenosis (62.9% survival at 29 years of age; no data after 29 years), and tricuspid atresia without pulmonary stenosis (61.3% survival at 35 years of age; Figure 1C; Table S2).

Among individuals who survived beyond the first year of life, the overall survival estimate at 35 years of age was 92.8% (Figure 2). Corresponding estimates among individuals who survived beyond 10 and 20 years of age were 97.2% and 98.2%, respectively. When further

restricting to individuals with severe CHDs, the probability of survival to 35 years of age was 87.7% for those who survived beyond 1 year, 96.3% for those who survived beyond 10 years, and 98.0% for those who survived beyond 20 years. When restricted to individuals with nonsevere CHDs, the probability of survival to 35 years of age was 95.3% for those who survived beyond 1 year, 97.7% for those who survived beyond 10 years, and 98.3% for those who survived beyond 20 years.

Kaplan-Meier survival curves by health and sociodemographic characteristics among those who survived beyond the first year of life are presented in Figure 3. Survival probability estimates at 35 years of age ranged from 72.2% for those with tricuspid atresia with pulmonary stenosis (Figure 3B) to 96.2% for those from a multiple birth (Figure 3G).

Adjusted associations between characteristics and death between birth and 1 year of age are described in Table 2. Compared with individuals in the shunt severity category, those with severe CHDs had the greatest risk (aRR=4.08) of death in the first year of life; however, all other severity categories also had increased risk compared with those with shunt defects (aRR range=1.29–1.76), although CIs for shunt+valve included 1.0. In addition, those with syndromes (aRR=1.83) and other noncardiac anomalies (aRR=1.54) had a higher risk of death compared with those without noncardiac anomalies, and estimates remained elevated when stratified by CHD severity. Compared with those born between 1991 and 1997, individuals born in earlier years were more likely to have died in the first year of life (aRR range=1.25–1.49); in particular, those with nonsevere CHDs born in earlier years compared with more recent years were more likely to have died in the first year of life (aRR range=1.34–1.81). These relationships between birth year and death did not substantially differ between infants with NH Black mothers and infants with NH White mothers (NH Black aRR range=1.50–1.71; NH White aRR range=1.23–1.41; interaction term $P>0.05$). Those born with a low birth weight (<2500 g) had increased risk of death in the first year of life (aRR=1.70), even when stratified by CHD severity. Those with Hispanic or NH Black mothers had increased risk of death in the first year of life (Hispanic aRR=1.27; NH Black aRR=1.43) compared with those with NH White mothers. Rurality was associated with lower risk of death for those with nonsevere CHDs (aRR=0.76 for middle and aRR=0.67 for most rural tertiles). aRR based on complete case analyses are presented in Table S3; associations using complete case analyses were stronger, in general, than those using imputed data, except for NH Black and Hispanic maternal race and ethnicity, whose associations were attenuated.

Among individuals who survived beyond the first year of life, ratios up to 35 years of age remained elevated for those with severe (aHR=3.18), shunt+valve (aHR=1.45), and valve (aHR=1.54) defects compared with those with shunt defects who also have genetic syndromes (aHR=3.06) or other noncardiac anomalies (aHR=2.53), were born with a low birth weight (aHR=1.29), and for NH Black (aHR=1.80) or Hispanic mothers (aHR=1.42; Figure 4). When stratifying by CHD severity (Figure S1), greater rurality was associated with higher aHRs for those with severe CHDs; otherwise, associations for those with severe CHDs were similar to those with nonsevere CHDs.

aHRs using complete case (nonimputed) and imputed data stratified by site and by noncardiac anomalies are displayed in Figures S2 through S4. Results were similar, with slightly wider CIs when using complete case rather than imputed data (Figure S2). The magnitude of the aHRs, in general, was consistent across sites, but with limited power; more CIs included 1.0 (Figure S3). Likewise, excluding individuals with noncardiac anomalies did not substantially affect the aHRs, although some CIs widened (Figure S4).

Comparing observed deaths in the first year of life in CH STRONG with expected deaths based on NVSS rates (Figure 5A), mortality was 10.17× greater (95% CI, 9.57–10.81) for individuals in CH STRONG than in the US population after standardizing by age group, sex, year, race and ethnicity, and site; those with severe CHDs had an SMR of 21.65, and the nonsevere CHD SMR was also elevated, at 4.87. SMRs remained elevated when stratified by race and ethnicity (NH White SMR=11.30; NH Black SMR=10.21) and sex (male SMR=9.54; female SMR=11.00). After excluding individuals with noncardiac anomalies from the CH STRONG sample, estimates remained elevated overall (SMR, 7.18 [95% CI, 6.53–7.88]) and across all strata (SMR range=2.38–18.06).

Comparing individuals who survived beyond the first year of life in CH STRONG with NVSS (Figure 5B), mortality was 3.29× greater (95% CI, 2.99–3.61) overall; by severity, those with severe CHDs had an SMR of 5.78, and those with nonsevere CHDs had an SMR of 2.17. Standardized mortality ratios remained elevated when limited to NH White individuals (SMR=2.84), NH Black individuals (SMR=4.58), males (SMR=2.49), and females (SMR=4.99). After restricting the sample to individuals with CHDs without noncardiac anomalies, mortality among those with nonsevere CHDs was similar to the general population (SMR, 1.13 [95% CI, 0.87–1.44]), but all other estimates remained elevated (overall SMR, 1.91 [95% CI, 1.62–2.23]; stratified SMRs range=1.38–3.66; Table S4). Among those who survived to 10 and 20 years of age, SMRs remained elevated overall (SMRs=1.45 and 1.54, respectively). However, after excluding individuals with noncardiac congenital anomalies, the overall SMRs at 10 and 20 years of age were null.

DISCUSSION

Among a large population-based sample of individuals with CHDs born between 1980 and 1997 in 3 US locations, the probability of survival to 35 years of age was 81.4% overall. For those who survived beyond the first year of life, the probability of survival to 18 to 35 years of age surpassed 95%. Unfortunately, infant mortality for those with CHDs was ≈0× greater overall and 22× greater for those with severe CHDs than the general population. Mortality beyond 1 year of age for individuals with CHDs was still 2 to 5× higher than in the general population, particularly among individuals who had severe CHDs and co-occurring noncardiac anomalies. After excluding individuals with co-occurring noncardiac anomalies, mortality among individuals with nonsevere CHDs who survived beyond 1 year of age and among individuals with any CHD who survived beyond 10 years of age were similar to the general population. Survival probabilities varied by defect, and characteristics associated with reduced survival included severe CHDs (including hypoplastic left heart syndrome, other single-ventricle defects, and tricuspid atresia with or without pulmonary

valve stenosis), presence of noncardiac anomalies, low birth weight, and Hispanic or NH Black maternal race and ethnicity.

Several large studies have been limited to examining mortality in US death certificate data (in which a CHD was listed as an underlying or contributing cause of death).[13–16] In general, these studies found that CHD-related mortality has declined over time, although less so for NH Black individuals. Although we similarly observed higher infant mortality in earlier birth years than in later years, we did not find strong evidence for effect modification by race in the association between infant mortality and birth year in this analysis. Other studies have investigated survival up to 1 year of age among infants with CHDs identified from a US birth defect surveillance system.[12,27–30] Excluding those with co-occurring noncardiac anomalies, their single-site probability estimates of 1-year survival for individuals with CHDs ranged from 88% to 97%, similar to our estimate of 89% using data from 3 sites. Our estimate, including those with noncardiac anomalies, was slightly below that range, at 85%.

To our knowledge, our analysis is the first to assess both conditional survival to adulthood and survival stratified by health and sociodemographic characteristics at birth among a population-based sample of US adults with CHDs. Our 35-year survival probability estimates that were conditional on 1-year survival (93% for all CHDs; 88% for severe) were similar to previously published US results on 30-year survival after cardiac surgery, with estimates ranging from 93% to 97% for nonsevere CHDs and from 65% to 86% for severe CHDs.[11] Overall, severe CHDs are commonly associated with reduced survival in the first year of life[12,29,30] and into adulthood.[11,12] Furthermore, our analysis contributes to a body of literature concluding that overall survival is lowest specifically for those with hypoplastic left heart syndrome,[27,28,31,32] and survival after 1 year of age is lowest for all single-ventricle defects.

In a previous publication comparing patients up to 32 years after CHD surgery with the population,[11] the authors found the overall SMR to be 8.3, and it remained elevated among those with nonsevere CHDs (who still required CHD surgery). Their overall estimate and their estimate after excluding chromosomal defects (SMR=7.5), respectively, fall between the SMRs we found in the first year of life (overall SMR=10.17; excluding chromosomal defects=7.18) and after the first year of life (overall SMR=3.29; excluding chromosomal defects=1.91). Our estimates differ in that we include deaths due to first CHD surgery, of which the majority occur in the first year of life.[11] Our analyses also include estimates stratified by severity, race, sex, and conditional survival beyond 1, 10, and 20 years of age. Our estimates show elevated SMRs overall; however, after excluding individuals with co-occurring noncardiac anomalies from the sample, mortality among individuals with nonsevere CHDs after 1 year of age and for individuals with any CHD after 10 years of age were similar to the general population. Another recent publication also reported that all-cause mortality rates among Danish individuals with simple CHDs who survived to at least 5 years of age were similar to matched controls without CHDs until ≈70 years of age, at which point higher mortality is driven by excess risk of comorbidity.[33]

In a previously published meta-analysis combining US and non-US mortality estimates,[34] Black race was found to be associated with mortality in the first year of life among those with severe (pooled odds ratio=1.68; 5 studies) and nonsevere CHDs (pooled odds ratio=1.62; 3 studies) and with inpatient mortality at any age among those with severe CHDs (pooled odds ratio=1.44; 5 studies). The authors reported that an association between Black race and overall mortality beyond the first year of life was less clear; their pooled odds ratio estimate was elevated at 1.47 with wide confidence intervals (0.82–2.63). The authors further reported no clear associations with Hispanic ethnicity, and studies were too heterogeneous to calculate a pooled estimate. Similarly finding that racial and ethnic disparities in CHD mortality persist through 35 years of age in the United States, Lopez et al[14] provided a comprehensive overview of literature describing upstream factors that influence disparities at the population level, such as access to proximate care; the systemic level, such as the number of adult CHD providers available or the health policies protecting insurance coverage; the institutional level, such as in referrals to quality CHD care and the existence of implicit provider bias; and the individual level, such as socioeconomic barriers to health care or even awareness of the need for continual cardiac care, which may all affect racial and ethnic differences in mortality.

Aside from CHD severity and maternal race and ethnicity, noncardiac anomalies[31,32] and low birth weight[12] have also been associated with reduced survival among individuals with CHDs in previous literature and in this analysis. Although earlier birth year has been identified as a risk factor for infant death among those with severe CHDs born between 1979 and 2005 in previous literature,[12] earlier birth years were only associated with increased infant mortality among those with nonsevere CHDs in the present analysis. Authors of the previous publication used a different definition to identify individuals with severe CHDs, which explains why our results differ from theirs (eg, they included the Ebstein anomaly and not common atrioventricular canal or aortic atresia/hypoplasia in their severe defects). Diagnostics and care of individuals with CHDs have improved over the decades,[35] which might explain the association between birth year and mortality among individuals with nonsevere CHDs. However, concurrent improvements in documentation of neonatal deaths over time might explain the lack of association among those with severe CHDs.

A few studies have found that neighborhood deprivation increases the risk of mortality for infants and children with CHDs, but they do not provide information beyond early childhood.[30,36,37] We only found an association between county deprivation and reduced survival into adulthood among those with severe CHDs who survived the first year of life, but no associations with survival during the first year of life. Different definitions of neighborhood (eg, census tract, zip code, or county) and different definitions of deprivation (eg, poverty only, median income only, or a combination of socioeconomic variables) might explain why our findings differ from others in the first year of life. After the first year of life, county rurality was associated with reduced survival for those with severe CHDs. Rurality may be indicative of proximity to specialty cardiac care,[38] another important geographic predictor of mortality[39,40] and later diagnosis and treatment.[41]

Limitations

Our analysis should be considered within the context of its limitations. Some linkages to death records may have been missed if their date of birth, sex, or name in the birth defects surveillance system had changed or did not resemble that in the death records. In addition, the Atlanta site only linked to the NDI through 2008 to identify out-of-state deaths. For these reasons, a small percentage of later deaths may have been missed. Specific dates of birth and death were not available; therefore, estimated age of death could differ from actual age by up to 1 year. We did not examine gestational age at birth or maternal age at delivery because nearly 45% of individuals were missing these data. Deprivation indicators were based on decennial census data for the person's county of birth, which was the smallest geographic unit available, but may not fully reflect deprivation in the immediate birth neighborhood. In addition, we did not have individual-level socioeconomic status. Categorization of county deprivation and rurality into tertiles is specific to their distribution in the CH STRONG data set. The NVSS site, from which general population death rates were collected for the SMRs, was assigned according to residence at the time of death certificate or census, whereas the CH STRONG site was assigned according to the state in which the patient was born. Furthermore, unlike NVSS, CH STRONG used race and ethnicity of the mother as a proxy for race and ethnicity of the individual with CHD. It is unclear how these differences may have affected the SMR.

Cause of death was not consistently captured by each site and therefore not examined in this analysis. Other US studies on CHD mortality found noncardiac anomalies, including chromosomal syndromes, cardiovascular disorders (acute myocardial infarction, cardiac arrest, heart failure, or arrythmia), injury, respiratory disease, and infection commonly colisted with CHD as causes of death before 35 years of age.

Individuals in CH STRONG were born ≥15 years before the implementation of critical newborn CHD screening policies in their respective states.[42] Therefore, our data do not reflect the effect these policies have had on improved early detection of critical CHDs and subsequent improvements in survival, although a previous article reported a resulting 33% decrease in infant deaths from severe CHDs after policy implementation.[43]

Conclusions

At least 8 in 10 individuals with CHDs born between 1980 and 1997 survived to adulthood, and survival improved substantially after the first year of life. Still, survival for those with CHDs was lower than in the general population and depended on severity of defect, co-occurring anomalies, weight at birth, and race and ethnicity. However, excluding those with noncardiac anomalies, mortality among individuals with nonsevere CHDs who survived beyond 1 year of age and among individuals with any CHD who survived beyond 10 years of age were similar to the general population. In addition, increasing access to and use of high-quality cardiac care, especially for those disproportionately affected by physical and socioeconomic barriers, may improve survival among those with CHDs and help address disparities by race and ethnicity.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

The authors thank Dr Kramer and A. Judge for developing the methods used in this analysis to calculate a county deprivation index based on the socioeconomic census data available in CH STRONG.

Nonstandard Abbreviations and Acronyms

aHR	adjusted hazard ratio
aRR	adjusted risk ratio
CDC/BPA	Centers for Disease Control and Prevention-modified version of the British Paediatric Association
CHD	congenital heart defect
CH STRONG	Congenital Heart Survey to Recognize Outcomes, Needs, and Well-Being
NDI	National Death Index
NH	non-Hispanic
NVSS	National Vital Statistics System
SMR	standardized mortality ratio

REFERENCES

1. Reller MD, Strickland MJ, Riehle-Colarusso T, Mahle WT, Correa A. Prevalence of congenital heart defects in metropolitan Atlanta, 1998–2005. J Pediatr. 2008;153:807–813. doi: 10.1016/j.jpeds.2008.05.059 [PubMed: 18657826]
2. Hoffman JIE, Kaplan S. The incidence of congenital heart disease. J Am Coll Cardiol. 2002;39:1890–1900. doi: 10.1016/s0735-1097(02)01886-7 [PubMed: 12084585]
3. Yang Q, Chen H, Correa A, Devine O, Mathews TJ, Honein MA. Racial differences in infant mortality attributable to birth defects in the United States, 1989–2002. Birth Defects Res A Clin Mol Teratol. 2006;76:706–713. doi:10.1002/bdra.20308 [PubMed: 17022030]
4. Su Z, Zou Z, Hay SI, Liu Y, Li S, Chen H, Naghavi M, Zimmerman MS, Martin GR, Wilner LB, et al. Global, regional, and national time trends in mortality for congenital heart disease, 1990–2019: an age-period-cohort analysis for the Global Burden of Disease 2019 study. eClinicalMedicine. 2022;43:101249. doi: 10.1016/j.eclinm.2021.101249 [PubMed: 35059612]
5. Moons P, Bovijn L, Budts W, Belmans A, Gewillig M. Temporal trends in survival to adulthood among patients born with congenital heart disease from 1970 to 1992 in Belgium. Circulation. 2010;122:2264–2272. doi:10.1161/CIRCULATIONAHA.110.946343 [PubMed: 21098444]
6. Greutmann M, Tobler D, Kovacs AH, Greutmann-Yantiri M, Haile SR, Held L, Ivanov J, Williams WG, Oechslin EN, Silversides CK, et al. Increasing mortality burden among adults with complex congenital heart disease. Congenit Heart Dis. 2014;10:117–127. doi: 10.1111/chd.12201 [PubMed: 25043406]

7. Oliver JM, Gallego P, Gonzalez AE, Garcia-Hamilton D, Avila P, Yotti R, Ferreira I, Fernandez-Aviles F. Risk factors for excess mortality in adults with congenital heart diseases. Eur Heart J. 2017;38:1233–1241. doi:10.1093/eurheartj/ehw590 [PubMed: 28077469]
8. Wu M-H, Lu C-W, Chen H-C, Kao F-Y, Huang S-K. Adult congenital heart disease in a nationwide population 2000–2014: epidemiological trends, arrhythmia, and standardized mortality ratio. J Am Heart Assoc. 2018;7:e007907. doi: 10.1161/JAHA.117.007907 [PubMed: 29437602]
9. Müller MJ, Norozi K, Caroline J, Sedlak N, Bock J, Paul T, Geyer S, Dellas C. Morbidity and mortality in adults with congenital heart defects in the third and fourth life decade. Clin Res Cardiol. 2022;111:900–911. doi:10.1007/s00392-022-01989-1 [PubMed: 35229166]
10. Mandalenakis Z, Rosengren A, Skoglund K, Lappas G, Eriksson P, Dellborg M. Survivorship in children and young adults with congenital heart disease in Sweden. JAMA Intern Med. 2017;177:224–230. doi:10.1001/jamainternmed.2016.7765 [PubMed: 27992621]
11. Spector LG, Menk JS, Knight JH, McCracken C, Thomas AS, Vinocur JM, Oster ME, St Louis JD, Moller JH, Kochilas L. Trends in long-term mortality after congenital heart surgery. J Am Coll Cardiol. 2018;71:2434–2446. doi:10.1016/j.jacc.2018.03.491 [PubMed: 29793633]
12. Oster ME, Lee KA, Honein MA, Riehle-Colarusso T, Shin M, Correa A. Temporal trends in survival among infants with critical congenital heart defects. Pediatrics. 2013;131:e1502–e1508. doi: 10.1542/peds.2012-3435 [PubMed: 23610203]
13. Gilboa SM, Salemi JL, Nembhard WN, Fixler DE, Correa A. Mortality resulting from congenital heart disease among children and adults in the United States, 1999 to 2006. Circulation. 2010;122:2254–2263. doi:10.1161/CIRCULATIONAHA.110.947002 [PubMed: 21098447]
14. Lopez KN, Morris SA, Sexson Tejtel SK, Espaillat A, Salemi JL. US mortality attributable to congenital heart disease across the lifespan from 1999 through 2017 exposes persistent racial/ethnic disparities. Circulation. 2020;142:1132–1147. doi: 10.1161/CIRCULATIONAHA.120.046822 [PubMed: 32795094]
15. Pillutla P, Shetty KD, Foster E. Mortality associated with adult congenital heart disease: Trends in the US population from 1979 to 2005. Am Heart J. 2009;158:874–879. doi: 10.1016/j.ahj.2009.08.014 [PubMed: 19853711]
16. Boneva RS, Botto LD, Moore CA, Yang Q, Correa A, Erickson JD. Mortality associated with congenital heart defects in the United States. Circulation. 2001;103:2376–2381. doi: 10.1161/01.cir.103.19.2376 [PubMed: 11352887]
17. Nembhard WN, Pathak EB, Schocken DD. Racial/ethnic disparities in mortality related to congenital heart defects among children and adults in the United States. Ethn Dis. 2008;18:442–449. [PubMed: 19157248]
18. Farr SL, Klewer SE, Nembhard WN, Alter C, Downing KF, Andrews JG, Collins RT, Glidewell J, Benavides A, Goudie A, et al. Rationale and design of CH STRONG: Congenital Heart Survey to Recognize Outcomes, Needs, and well-beinG. Am Heart J. 2020;221:106–113. doi: 10.1016/j.ahj.2019.12.021 [PubMed: 31986287]
19. Glidewell J, Book W, Raskind-Hood C, Hogue C, Dunn JE, Gurvitz M, Ozonoff A, McGarry C, Van Zutphen A, Lui G, et al. Population-based surveillance of congenital heart defects among adolescents and adults: surveillance methodology. Birth Defects Res. 2018;110:1395–1403. doi:10.1002/bdr2.1400 [PubMed: 30394691]
20. Downing KF, Oster ME, Klewer SE, Rose CE, Nembhard WN, Andrews JG, Farr SL. Disability among young adults with congenital heart defects: congenital heart survey to recognize outcomes, needs, and well-being 2016–2019. J Am Heart Assoc. 2021;10:e022440. doi: 10.1161/JAHA.121.022440 [PubMed: 34666499]
21. Farr SL, Downing KF, Goudie A, Klewer SE, Andrews JG, Oster ME. advance care directives among a population-based sample of young adults with congenital heart defects, CH STRONG, 2016–2019. Pediatr Cardiol. 2021;42:1775–1784. doi: 10.1007/s00246-021-02663-5 [PubMed: 34164699]
22. Oster ME, Riser AP, Andrews JG, Bolin EH, Galindo MK, Nembhard WN, Rose CE, Farr SL. Comorbidities among young adults with congenital heart defects: results from the Congenital Heart Survey to Recognize Outcomes, Needs, and well-being–Arizona, Arkansas, and Metropolitan Atlanta, 2016–2019. MMWR Morb Mortal Wkly Rep. 2021;70:197–201. doi:10.15585/mmwr.mm7006a3 [PubMed: 33571179]

23. Dong Y, Peng C-YJ. Principled missing data methods for researchers. SpringerPlus. 2013;2:222. doi: 10.1186/2193-1801-2-222 [PubMed: 23853744]
24. Buuren S van, Groothuis-Oudshoorn K. mice: Multivariate imputation by chained equations in R. J Stat Softw. 2011;45:1–67. doi:10.18637/jss.v045.i03
25. Dean AG, Sullivan KM, Soe MM. OpenEpi: open source epidemiologic statistics for public health, Version. Accessed April 6, 2013. https://www.OpenEpi.com
26. Team RC. R: A language and environment for statistical computing. Vienna, Austria: R Foundation for Statistical Computing; 2021. https://www.R-project.org/
27. Nembhard WN, Salemi JL, Ethen MK, Fixler DE, DiMaggio A, Canfield MA. Racial/ethnic disparities in risk of early childhood mortality among children with congenital heart defects. Pediatrics. 2011;127:e1128–e1138. doi:10.1542/peds.2010-2702 [PubMed: 21502234]
28. Cleves MA, Ghaffar S, Zhao W, Mosley 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. doi: 10.1002/bdra.10119 [PubMed: 14703791]
29. Pace ND, Oster ME, Forestieri NE, Enright D, Knight J, Meyer RE. Sociodemographic factors and survival of infants with congenital heart defects. Pediatrics. 2018;142:e20180302. doi: 10.1542/peds.2018-0302
30. Kucik JE, Cassell CH, Alverson CJ, Donohue P, Tanner JP, Minkovitz CS, Correia J, Burke T, Kirby RS. Role of health insurance on the survival of infants with congenital heart defects. Am J Public Health. 2014;104:e62–e70. doi: 10.2105/AJPH.2014.301969
31. Lynn MM, Salemi JL, Kostelyna SP, Morris SA, Tejtel SKS, Lopez KN. Lesion-specific congenital heart disease mortality trends in children: 1999 to 2017. Pediatrics. 2022;150:e2022056294. doi: 10.1542/peds.2022-056294
32. Lynn MM, Salemi JL, Meath CJ, Dolgner SJ, Morris SA, Sexson Tejtel SK, Lopez KN. Lesion-specific mortality due to congenital heart disease in U.S. adults from 1999 to 2017. Birth Defects Res. 2022;114:725–745. doi: 10.1002/bdr2.2044 [PubMed: 35593518]
33. El-Chouli M, Meddis A, Christensen DM, Gerds TA, Sehested T, Malmborg M, Phelps M, Bang CN, Ahlehoff O, Torp-Pedersen C, et al. Lifetime risk of comorbidity in patients with simple congenital heart disease: a Danish nationwide study. Eur Heart J. 2022;44:741–748. doi: 10.1093/eurheartj/ehac727
34. Tran R, Forman R, Mossialos E, Nasir K, Kulkarni A. Social determinants of disparities in mortality outcomes in congenital heart disease: a systematic review and meta-analysis. Front Cardiovasc Med. 2022;9:829902. doi:10.3389/fcvm.2022.829902 [PubMed: 35369346]
35. Liu A, Diller G-P, Moons P, Daniels CJ, Jenkins KJ, Marelli A. Changing epidemiology of congenital heart disease: effect on outcomes and quality of care in adults. Nat Rev Cardiol. 2023;20:126–137. doi:10.1038/s41569-022-00749-y [PubMed: 36045220]
36. Siffel C, Riehle-Colarusso T, Oster ME, Correa A. Survival of children with hypoplastic left heart syndrome. Pediatrics. 2015;136:e864–e870. doi:10.1542/peds.2014-1427 [PubMed: 26391936]
37. Anderson BR, Fieldston ES, Newburger JW, Bacha EA, Glied SA. Disparities in outcomes and resource use after hospitalization for cardiac surgery by neighborhood income. Pediatrics. 2018;141:e20172432. doi:10.1542/peds.2017-2432
38. Sommerhalter KM, Insaf TZ, Akkaya-Hocagil T, McGarry CE, Farr SL, Downing KF, Lui GK, Zaidi AN, Van Zutphen AR. Proximity to pediatric cardiac surgical care among adolescents with congenital heart defects in 11 New York counties. Birth Defects Res. 2017;109:1494–1503. doi:10.1002/bdr2.1129 [PubMed: 29152921]
39. Kaltman JR, Burns KM, Pearson GD, Goff DC, Evans F. Disparities in congenital heart disease mortality based on proximity to a specialized pediatric cardiac center. Circulation. 2020;141:1034–1036. doi:10.1161/CIRCULATIONAHA.119.043392 [PubMed: 32078376]
40. Udine ML, Evans F, Burns KM, Pearson GD, Kaltman JR. Geographical variation in infant mortality due to congenital heart disease in the USA: a population-based cohort study. Lancet Child Adolesc Health. 2021;5:483–490. doi: 10.1016/S2352-4642(21)00105-X [PubMed: 34051889]

41. Hill GD, Block JR, Tanem JB, Frommelt MA. Disparities in the prenatal detection of critical congenital heart disease. Prenat Diagn. 2015;35:859–863. doi: 10.1002/pd.4622 [PubMed: 25989740]

42. Glidewell J, Grosse SD, Riehle-Colarusso T, Pinto N, Hudson J, Daskalov R, Gaviglio A, Darby E, Singh S, Sontag M. Actions in support of newborn screening for critical congenital heart disease: United States, 2011–2018. MMWR Morb Mortal Wkly Rep. 2019;68:107–111. doi:10.15585/mmwr.mm6805a3 [PubMed: 30730872]

43. Abouk R, Grosse SD, Ailes EC, Oster ME. Association of US state implementation of newborn screening policies for critical congenital heart disease with early infant cardiac deaths. JAMA. 2017;318:2111–2118. doi:10.1001/jama.2017.17627 [PubMed: 29209720]

Clinical Perspective

What Is New?

- Eight in 10 individuals with congenital heart defects (CHDs) born between 1980 and 1997 in Arizona, Arkansas, and Georgia survived to 35 years of age, with disparities by CHD type and severity, noncardiac anomalies, birth weight, and maternal race and ethnicity.
- Survival for individuals with CHDs was lower than in the general population and depended on CHD severity, co-occurring noncardiac anomalies, birth weight, and race and ethnicity.
- Among individuals without noncardiac anomalies, mortality among individuals with nonsevere CHDs who survived beyond 1 year of age and among individuals with any CHD who survived beyond 10 years of age were similar to the general population.

What Are the Clinical Implications?

- On the basis of data from 3 US sites, most individuals with CHDs survive to young adulthood.
- Individuals with severe CHDs (with or without co-occurring noncardiac anomalies) and those with nonsevere CHDs with co-occurring anomalies continue to have reduced survival after the first year of life compared with the general population and may benefit from increased monitoring, earlier identification, and intervention for medical concerns.
- Improving access to and use of congenital cardiac care, especially for those disproportionately affected by physical and socioeconomic barriers, may improve survival among those with CHDs and help address disparities by race and ethnicity.

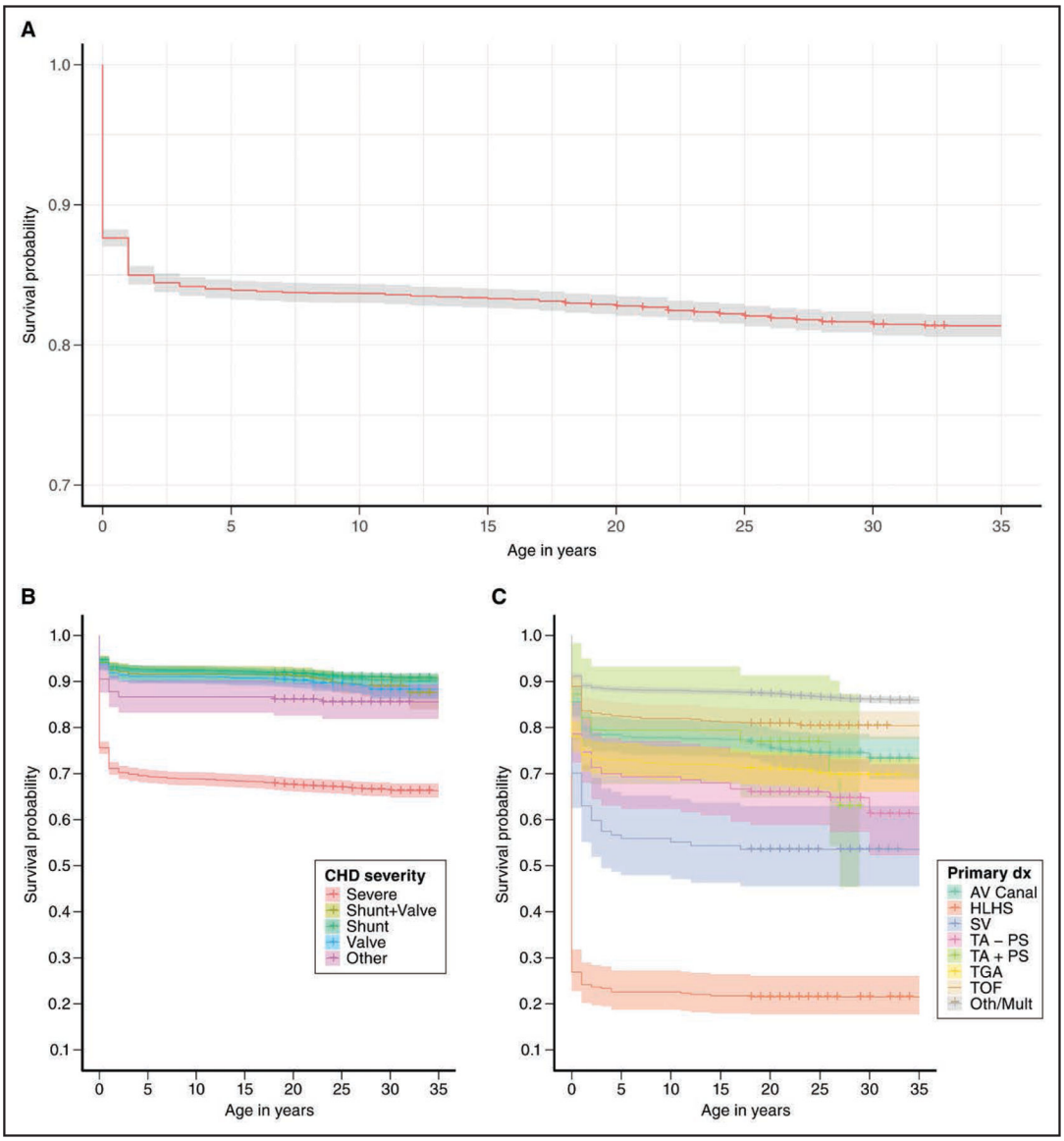

Figure 1. Survival for individuals with CHD born between 1980 and 1997 overall (A), by CHD severity (B), and by primary CHD diagnosis (C). CH STRONG.

AV indicates atrioventricular; CH STRONG, Congenital Heart Survey To Recognize Outcomes, Needs, and Well-Being; CHD, congenital heart defect; HLHS, hypoplastic left heart syndrome; Oth/Mult, other/multiple; Sh+V, shunt+valve; SV, single ventricle; TA – PS, tricuspid atresia without pulmonary stenosis; TA + PS, tricuspid atresia with pulmonary stenosis; TGA, transposition of the great arteries; and TOF, tetralogy of Fallot.

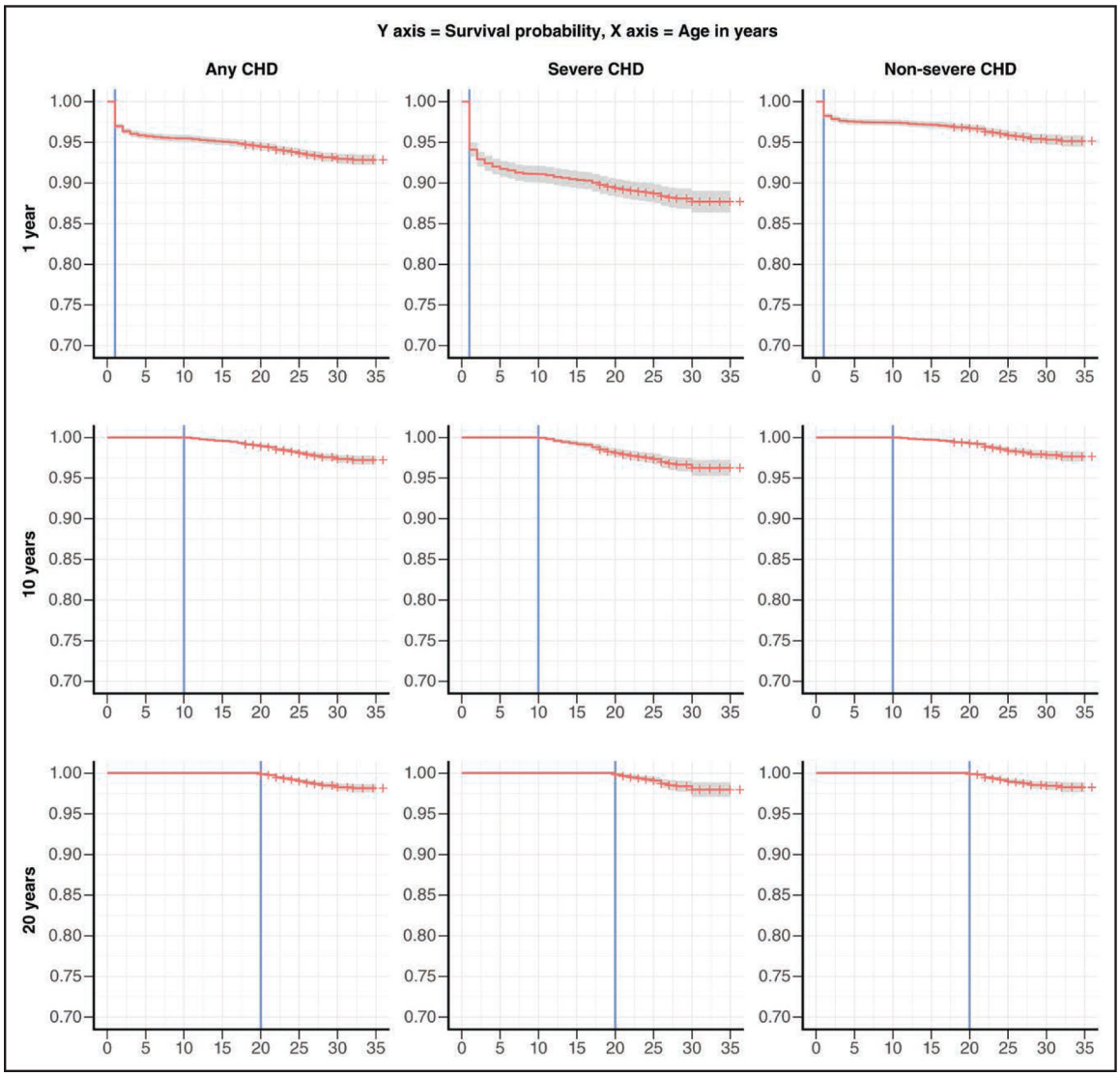

Figure 2. Survival beyond ages 1, 10, and 20 years for individuals with CHDs born between 1980 and 1997. CH STRONG.

CH STRONG indicates Congenital Heart Survey to Recognize Outcomes, Needs, and Well-Being; and CHD, congenital heart defect.

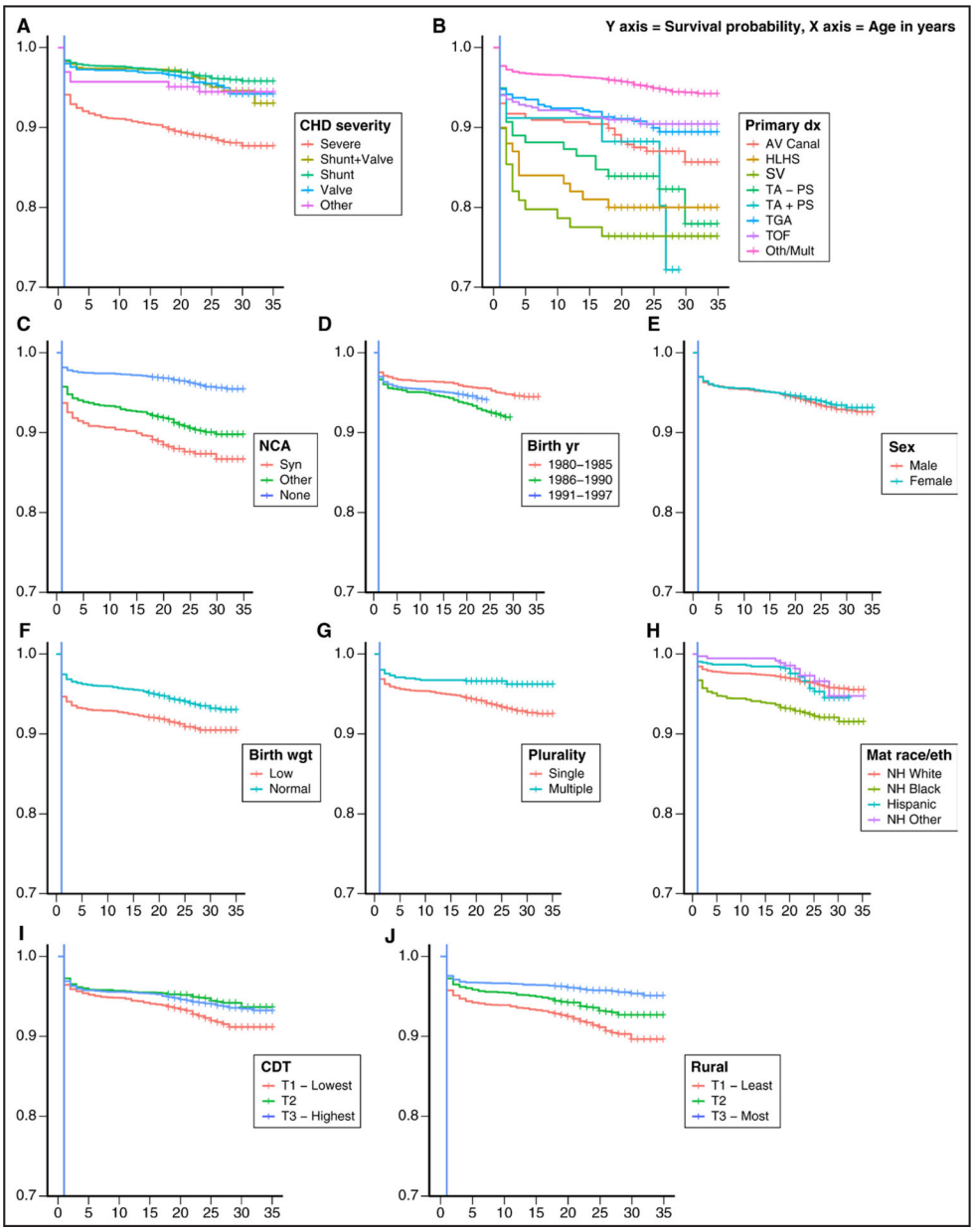

Figure 3. Survival beyond the first year of life for individuals with CHDs born between 1980 and 1997 by health and sociodemographic characteristics. CH STRONG.

AV indicates atrioventricular; CDT, county deprivation tertile; CH STRONG, Congenital Heart Survey to Recognize Outcomes, Needs, and Well-Being; CHD, congenital heart defect; dx, diagnosis; eth, ethnicity; HLHS, hypoplastic left heart syndrome; Mat, maternal; NH, non-Hispanic; NCA, noncardiac congenital anomalies; Oth/Mult, other/multiple; SV, single ventricle; Syn, syndromes; T, tertile; TA − PS, tricuspid atresia without pulmonary stenosis; TA + PS, tricuspid atresia with pulmonary stenosis; TGA, transposition of the great arteries; TOF, tetralogy of Fallot; and wgt, weight.

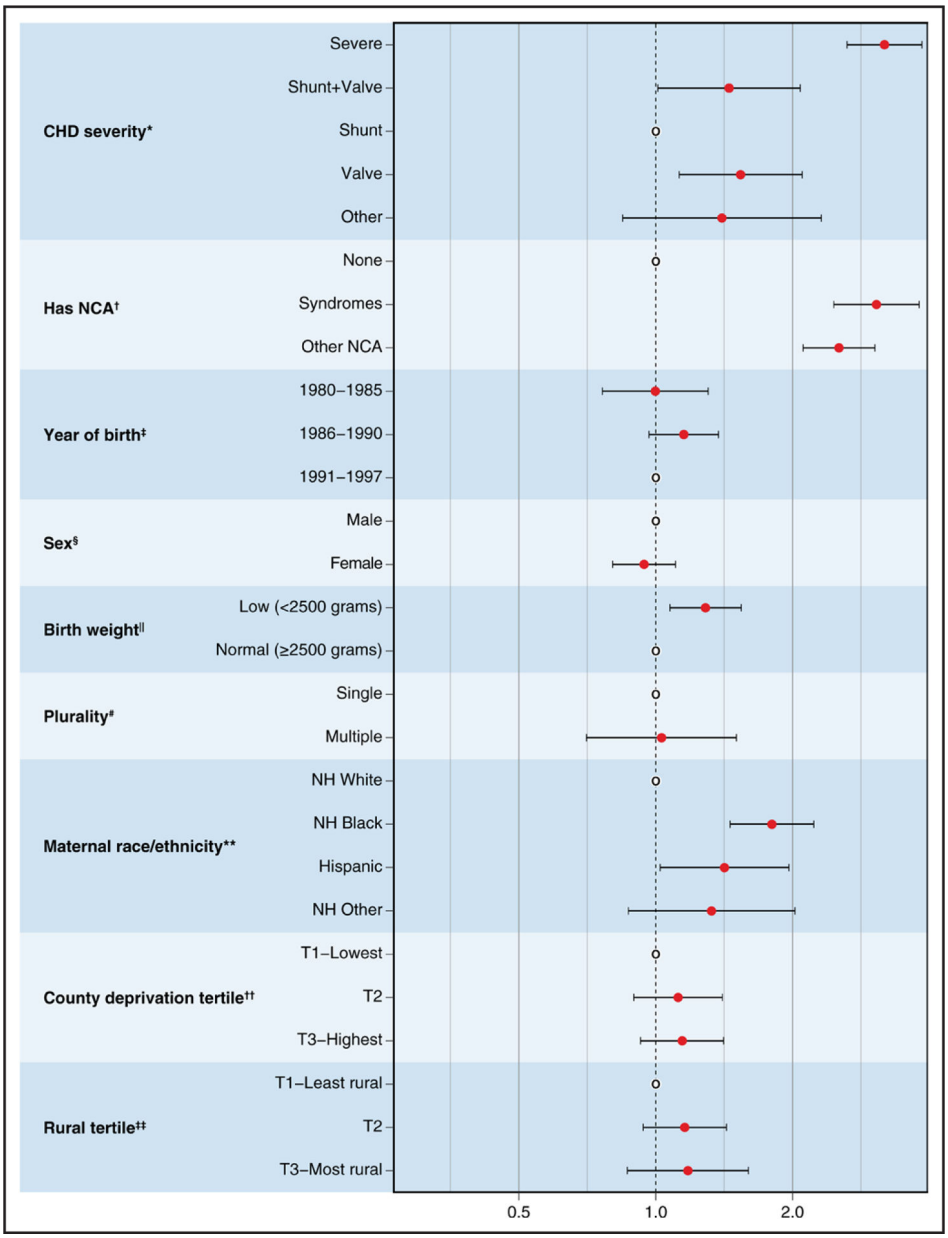

Figure 4. Adjusted hazards ratios up to 35 years of age for individuals with CHDs born between 1980 and 1997, conditional on survival beyond the first year of life. CH STRONG multiple imputed data.

CH STRONG indicates Congenital Heart Survey to Recognize Outcomes, Needs, and Well-Being; CHD, congenital heart defect; NCA, noncardiac congenital anomalies; NH, non-Hispanic; and T, tertile. O denotes reference group. *Model includes CHD severity, NCA, birth year, site, sex, maternal race and ethnicity, and county deprivation tertile at birth. †Model includes NCA, birth year, site, sex, plurality, maternal race and ethnicity, and county deprivation tertile at birth. ‡Model includes birth year and site. §Model includes sex and birth year. ‖Model includes birth weight, NCA, birth year, site, sex, plurality, maternal race and ethnicity, and county deprivation tertile of birth county. #Model includes plurality, birth year, site, sex, and maternal race and ethnicity. **Model includes maternal race and

ethnicity, birth year, and site. ††Model includes county deprivation tertile of birth county, birth year, site, and maternal race and ethnicity. ‡‡Model includes rurality of birth county, birth year, site, maternal race and ethnicity, and county deprivation tertile at birth.

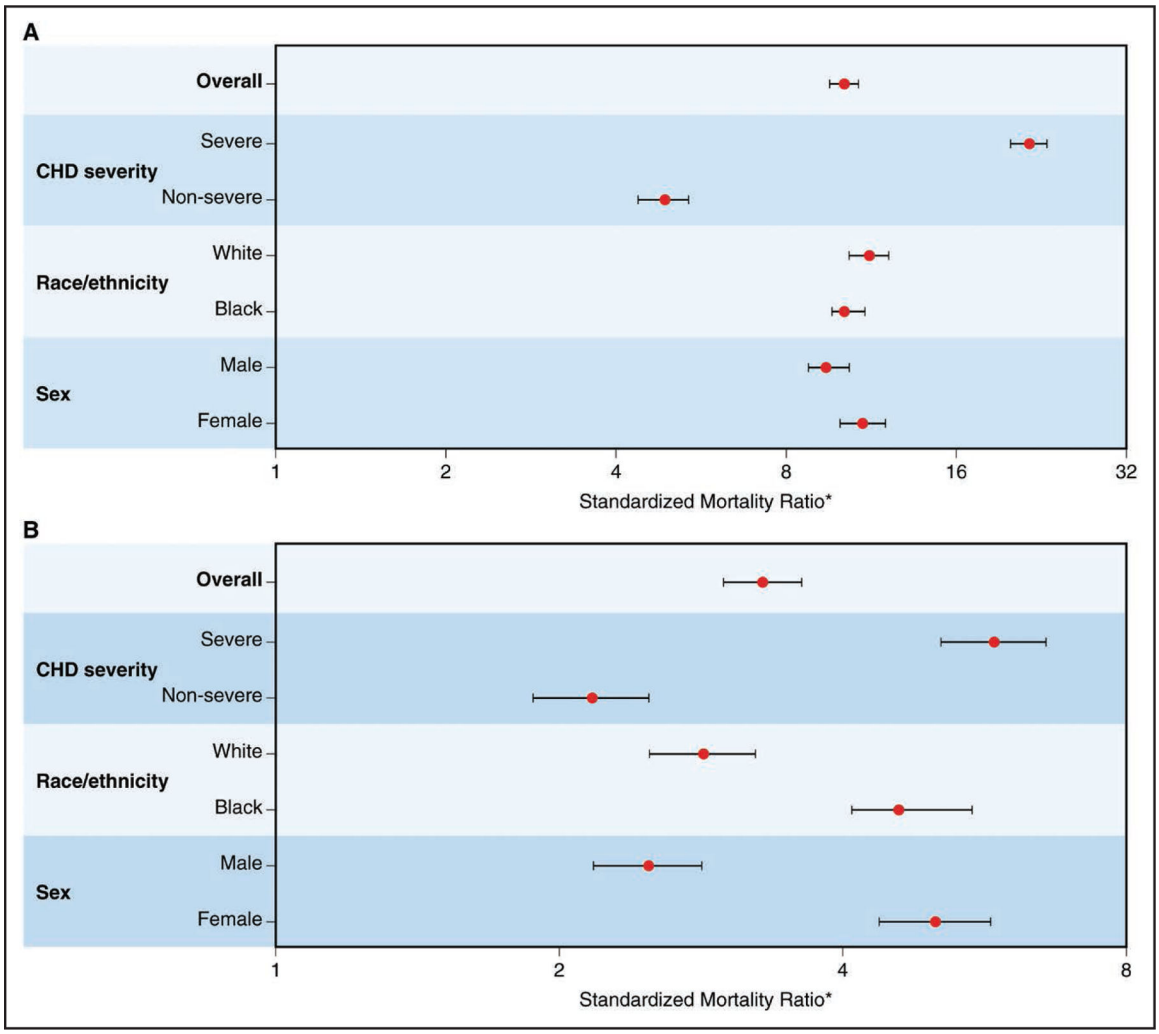

Figure 5. Standardized mortality ratios in the first year of life (A) and beyond the first year of life (B) for individuals with CHDs compared with age group-, sex-, year-, race and ethnicity–, and site-matched data from the general US population, CH STRONG, and National Vital Statistics System.

CH STRONG indicates Congenital Heart Survey to Recognize Outcomes, Needs, and Well-Being; and CHD, congenital heart defects. *All corresponding *P* values <0.001.

Table 1.

Health and Sociodemographic Characteristics at Birth for Individuals With Congenital Heart Defects in 3 US Birth Defect Surveillance Systems Between 1980 and 1997, by Vital Status as of 2015, CH STRONG

Characteristics	Presumed alive (n=9323)	Deceased (n=2372)	Total (N=11 695)
Total, n (%)	9323 (79.7)	2372 (20.3)	11 695 (100)
Congenital heart defect severity, n (%)			
Severe	2705 (29.0)	1567 (66.1)	4272 (36.5)
Shunt+valve	839 (9.0)	111 (4.7)	950 (8.1)
Shunt	4328 (46.4)	483 (20.4)	4811 (41.1)
Valve	1142 (12.2)	154 (6.5)	1296 (11.1)
Other	309 (3.3)	57 (2.4)	366 (3.1)
Has noncardiac congenital anomalies,* n (%)			
Syndromes	977 (10.5)	475 (20.0)	1452 (12.4)
Other	2584 (27.7)	887 (37.4)	3471 (29.7)
None	5762 (61.8)	1010 (42.6)	6772 (57.9)
Primary congenital heart defect diagnosis, n (%)			
Atrioventricular canal	337 (3.6)	136 (5.7)	473 (4.0)
Hypoplastic left heart syndrome	80 (0.9)	337 (14.2)	417 (3.6)
Single ventricle	68 (0.7)	72 (3.0)	140 (1.2)
Tricuspid atresia without pulmonary stenosis	97 (1.0)	64 (2.7)	161 (1.4)
Tricuspid atresia with pulmonary stenosis	28 (0.3)	13 (0.5)	41 (0.4)
Transposition of the great arteries	416 (4.5)	211 (8.9)	627 (5.4)
Tetralogy of Fallot	532 (5.7)	157 (6.6)	689 (5.9)
Other/multiple	7765 (83.3)	1382 (58.3)	9147 (78.2)
Year of birth, n (%)			
1980–1985	1348 (14.5)	364 (15.3)	1712 (14.6)
1986–1990	2647 (28.4)	875 (36.9)	3522 (30.1)
1991–1997	5328 (57.1)	1133 (47.8)	6461 (55.2)
Sex,† n (%)			
Male	4796 (51.5)	1263 (53.4)	6059 (51.9)
Female	4515 (48.5)	1104 (46.6)	5619 (48.1)
Birth weight,‡ n (%)			
Low (<2500 g)	2037 (23.6)	848 (36.5)	2885 (26.3)
Normal (≥2500 g)	6606 (76.4)	1476 (63.5)	8082 (73.7)
Plurality, n (%)			
Single	8495 (91.1)	2272 (95.8)	10 767 (92.1)
Multiple	828 (8.9)	100 (4.2)	928 (7.9)
Maternal race and ethnicity,§ n (%)			
Non-Hispanic White	6062 (66.1)	819 (58.8)	6881 (65.1)

Characteristics	Presumed alive (n=9323)	Deceased (n=2372)	Total (N=11 695)
Non-Hispanic Black	1946 (21.2)	489 (35.1)	2435 (23.0)
Hispanic	813 (8.9)	53 (3.8)	866 (8.2)
Non-Hispanic Other	349 (3.8)	33 (2.4)	382 (3.6)
County deprivation tertile,[//] n (%)			
T1: lowest deprivation	3579 (40.8)	1159 (49.3)	4738 (42.6)
T2	2211 (25.2)	478 (20.3)	2689 (24.2)
T3: highest deprivation	2975 (33.9)	713 (30.3)	3688 (33.2)
Rural tertile,[//] n (%)			
T1: least rural	3153 (36.0)	1163 (49.5)	4316 (38.8)
T2	2472 (28.2)	630 (26.8)	3102 (27.9)
T3: most rural	3140 (35.8)	557 (23.7)	3697 (33.3)

CDC/BPA indicates Centers for Disease Control and Prevention-modified version of the British Paediatric Association; CH STRONG indicates Congenital Heart Survey To Recognize Outcomes, Needs, and Well-Being; and T, tertile.

[*]Noncardiac congenital anomalies include all CDC/BPA codes collected by the birth defects surveillance systems outside of 745.000 to 747.999. Syndromes include CDC/BPA codes between 758.000 and 758.999.

[†]Data on sex missing in 17.

[‡]Data on birth weight missing in 728.

[§]Data on maternal race and ethnicity missing in 1131.

[//]County-level census data missing in 580.

Table 2.

Adjusted Risk Ratios for Death During the First Year of Life for Individuals With CHDs Born Between 1980 and 1997, CH STRONG Multiply Imputed Data

	Any CHD (n=11 695)		Severe CHDs (n=4272)		Nonsevere CHDs (n=7423)	
Characteristic	**aRR**	**95% CI**	**aRR**	**95% CI**	**aRR**	**95% CI**
CHD severity *						
Severe	4.12	3.60–4.72				
Shunt+valve	1.28	0.97–1.70				
Shunt	—	—				
Valve	1.27	1.00–1.63				
Other	1.76	1.24–2.49				
Has noncardiac congenital anomalies †						
Syndromes	2.33	1.94–2.79	1.07	0.85–1.35	6.46	4.64–8.97
Other	1.71	1.47–1.99	1.2	0.99–1.44	3.31	2.47–4.45
None	—	—	—	—	—	—
Year of birth ‡						
1980–1985	1.49	1.27–1.74	0.98	0.81–1.19	1.80	1.34–2.41
1986–1990	1.24	1.11–1.38	1.11	0.98–1.27	1.31	1.07–1.61
1991–1997	—	—	—	—	—	—
Sex §						
Male	—	—	—	—	—	—
Female	0.93	0.84–1.03	1.07	0.95–1.21	0.94	0.78–1.13
Birth weight ‖						
Low (<2500 g)	1.70	1.53–1.90	1.63	1.42–1.87	2.88	2.36–3.51
Normal (≥2500 g)	—	—	—	—	—	—
Plurality #						
Single	—	—	—	—	—	—
Multiple	0.90	0.70–1.17	0.99	0.71–1.37	1.03	0.67–1.57
Maternal race and ethnicity **						
Non-Hispanic White	—	—	—	—	—	—
Non-Hispanic Black	1.42	1.22–1.64	1.38	1.15–1.65	1.68	1.29–2.18
Hispanic	1.28	1.05–1.57	1.28	1.02–1.60	1.49	1.05–2.12
Non-Hispanic other	1.24	0.95–1.60	1.43	1.08–1.90	1.10	0.63–1.92
County deprivation tertile ††						
T1: least deprivation)	—	—	—	—	—	—
T2	0.98	0.85–1.13	0.99	0.84–1.17	0.97	0.75–1.25
T3: most deprivation	0.99	0.87–1.14	1.02	0.87–1.21	0.90	0.70–1.15
Rural tertile ‡‡						

Characteristic	Any CHD (n=11 695)		Severe CHDs (n=4272)		Nonsevere CHDs (n=7423)	
	aRR	95% CI	aRR	95% CI	aRR	95% CI
T1: least rural	—	—	—	—	—	—
T2	0.94	0.82–1.08	1.07	0.90–1.26	0.76	0.58–0.99
T3: most rural	0.94	0.77–1.14	1.09	0.87–1.36	0.67	0.46–0.98

aRR indicates adjusted risk ratio; CH STRONG, Congenital Heart Survey to Recognize Outcomes, Needs and Well-being; CHD, congenital heart defect; and T, tertile; — indicates reference group.

[*] Model includes CHD severity, noncardiac congenital anomalies, birth year, site, sex, maternal race and ethnicity, and county deprivation tertile of birth county.

[†] Model includes noncardiac congenital anomalies, birth year, site, sex, plurality, maternal race and ethnicity, and county deprivation tertile of birth county.

[‡] Model includes birth year and site.

[§] Model includes sex and birth year.

[‖] Model includes birth weight, noncardiac congenital anomalies, birth year, site, sex, plurality, maternal race and ethnicity, and county deprivation tertile of birth county.

[#] Model includes plurality, birth year, site, sex, and maternal race and ethnicity.

[**] Model includes maternal race and ethnicity, birth year, and site.

[††] Model includes county deprivation tertile of birth county, birth year, site, and maternal race and ethnicity.

[‡‡] Model includes rurality of birth county, birth year, site, maternal race and ethnicity, and county deprivation tertile of birth county.