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Individuals aged 1-64 years with documented congenital heart defects at healthcare encounters, five U.S. surveillance sites, 2011-2013

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Abstract

Background—Many individuals born with congenital heart defects (CHD) survive to adulthood. However, population estimates of CHD beyond early childhood are limited in the U.S.

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Conflict of interest

None.

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Disclosure

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

Supplementary materials

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Objectives—To estimate the percentage of individuals aged 1-to-64 years at five U.S. sites with CHD documented at a healthcare encounter during a three-year period and describe their characteristics.

Methods—Sites conducted population-based surveillance of CHD among 1 to 10-year-olds (three sites) and 11 to 64-year-olds (all five sites) by linking healthcare data. Eligible cases resided in the population catchment areas and had one or more healthcare encounters during the surveillance period (January 1, 2011-December 31, 2013) with a CHD-related ICD-9-CM code. Site-specific population census estimates from the same age groups and time period were used to assess percentage of individuals in the catchment area with a CHD-related ICD-9-CM code documented at a healthcare encounter (hereafter referred to as CHD cases). Severe and non-severe CHD were based on an established mutually exclusive anatomic hierarchy.

Results—Among 42,646 CHD cases, 23.7% had severe CHD and 51.5% were male. Percentage of CHD cases among 1 to 10-year-olds, was 6.36/1,000 (range: 4.33-9.96/1,000) but varied by CHD severity [severe: 1.56/1,000 (range: 1.04-2.64/1,000); non-severe: 4.80/1,000 (range: 3.28-7.32/1,000)]. Percentage of cases across all sites in 11 to 64-year-olds was 1.47/1,000 (range: 1.02-2.18/1,000) and varied by CHD severity [severe: 0.34/1,000 (range: 0.26-0.49/1,000); non-severe: 1.13/1,000 (range: 0.76-1.69/1,000)]. Percentage of CHD cases decreased with age until 20 to 44 years and, for non-severe CHD only, increased slightly for ages 45 to 64 years.

Conclusion—CHD cases varied by site, CHD severity, and age. These findings will inform planning for the needs of this growing population.

Congenital heart defects (CHD) are the most common type of birth defects and affect about 1% of U.S. births)[1,2] Due to advances in medical and surgical interventions, the majority of individuals with CHD live into adulthood[3–6]. Survival of individuals with CHD, even those with the most severe defects, has improved over time.[7,8] However, whereas birth defects surveillance programs provide valuable data on prevalence of CHD at and around the time of birth, in the U.S., population-level surveillance data on the prevalence of CHD beyond early childhood is scarce.[9]

For this reason, such estimates for the U.S. have so far largely relied on Canadian data or Canadian data extrapolated to the U.S. population.[3–5] Estimates of CHD prevalence based on Canadian data may not reflect the true population prevalence of CHD in the U.S. due to differences in sociodemographic characteristics and healthcare access and utilization. The U.S. healthcare system, which is fragmented and lacks universal health care in most states, poses a significant challenge for population-based surveillance of CHD. Individuals with less or no access to health care may not be identified through health care records. Even among insured individuals, a substantial percentage of those with CHD fall out of cardiac care.[10] In addition, following CHD patients across time and healthcare systems is difficult. All of these systemic challenges make determining U.S. population-based prevalence estimates difficult. Unsurprisingly, current knowledge of the CHD population in the U.S. is largely based on clinic-specific data or use of administrative datasets[11], each of which has its potential limitations and biases. Despite the challenges, the Centers for Disease Control and Prevention (CDC) has led a series of initiatives to improve empirical

estimates of population-based prevalence, as such estimates aid in adequately planning for the healthcare needs of individuals with CHD.[12]

From 2012 to 2015, the CDC initiated a three year, three-site pilot project to determine the feasibility of conducting population-based surveillance among adolescents and adults with CHD in the U.S.[9]. This project reported the population-based prevalence of adolescents and adults seeking healthcare with a high degree of variability, likely reflecting the access to and variation of data sources used for surveillance.[9,13,14] In 2015, the CDC funded a four-year project at five U.S. surveillance sites to conduct public health surveillance of 1 to 64-year-olds with CHD receiving health care. In this report, we describe the demographic and clinical characteristics of individuals with a documented CHD-related *International Classification of Diseases, Ninth Revision, Clinical Modification* (ICD-9-CM) diagnosis code during 2011-2013 data from these five surveillance sites.

Methods

CHD case definition

Individuals (hereafter referred to as CHD cases) were ascertained by the site-specific surveillance system if they had any ICD-9-CM CHD diagnosis codes between 745.xx to 747.xx documented in a healthcare encounter between January 1, 2011 and December 31, 2013, excluding: congenital heart block (746.86), absent/hypoplastic umbilical artery (747.5), pulmonary arteriovenous malformation (747.32), other anomalies of peripheral vascular system (747.6x), and other specified anomalies of circulatory system (747.8x). Cases with the eligible CHD diagnosis codes were categorized into five hierarchical native anatomic complexity groupings similar to previously published algorithms based on the individuals' hemodynamic severity and basic anatomy.[3,4,9] Categories are hereafter referred to as severe, shunt + valve, valve, shunt, and non-specific defects (Supplemental Table 1). If a case had a severe code and a code in one of the other categories, the case was considered severe. Cases with an isolated code of 745.5 were excluded from this analysis since secundum atrial septal defect, a CHD, cannot be distinguished by ICD codes from patent foramen ovale (PFO), a normal variant and, therefore, inclusion of 745.5 may overestimate cases with CHD.[15] Preliminary validation studies from this surveillance project showed poor positive predictive value (PPV) for non-specific CHD codes (unpublished data). Thus, cases in the non-specific defect category (all cases not categorized as severe, shunt + valve, valve, or shunt) were excluded regardless of whether their non-specific code co-occurred with 745.5, or they had > 1 non-specific code. CHD severity then was collapsed into two groups: severe CHD and non-severe CHD (shunt + valve, valve, and shunt). Severe CHD included endocardial cushion defects, interrupted aortic arch, tetralogy of Fallot, total anomalous pulmonary venous return, transposition complexes, truncus arteriosus and univentricular hearts.

Study population and case ascertainment

Cases aged 1-64 years were identified in Georgia (GA), North Carolina (NC) and New York (NY); cases aged 11-64 years were identified in Colorado (CO) and Utah (UT). All cases had a documented CHD-related ICD-9-CM code in one or more healthcare

encounters between January 1, 2011 and December 31, 2013 and a residential address in the site-specific catchment areas at some point in the 3-year surveillance period. The catchment areas for each site are as follows: statewide for CO, NC and UT, a 5-county metropolitan-Atlanta area for GA that included Clayton, Cobb, DeKalb, Fulton, and Gwinnett counties, and an 11-county area for NY that included Allegany, Bronx, Cattaraugus, Chautauqua, Erie, Genesee, Monroe, Niagara, Orleans, Westchester, and Wyoming counties. CO cases were identified using electronic health records (EHR) from four independent healthcare systems that serve diverse populations across the state and the Colorado All Payer Claims Database[16]. NC cases were identified using EHR data from five pediatric and adult comprehensive CHD clinical facilities. UT cases were identified using hospital discharge data and clinical data from two major health facilities with pediatric and adult cardiology specialty clinics. In GA, cases were identified using Medicaid claims data, data from six pediatric and adult clinical facilities, and an existing surgical database. NY cases were identified from three pediatric cardiology clinics, Medicaid claims data, and hospital inpatient and outpatient discharge data. Each site determined vital status by linking to state death certificates and retained this information in the surveillance dataset. Individuals that died during the surveillance period were not excluded from the surveillance system. Cases were linked and de-duplicated across participating case finding data sources at each site.

Analytic data elements

At each site, once eligible cases were identified, all available healthcare encounters, including those not related to CHD, were captured over the 3-year surveillance period. Age was defined as age at the first healthcare encounter with an eligible ICD-9-CM CHD code during the surveillance period. For this analysis, insurance type was a static, hierarchical variable (public, private, and none) during the 3-year period. I.e., if insurance documented at any healthcare encounter was Medicaid or Medicare, insurance type was assigned “public”; if it was private, other government, or other insurance, insurance type was assigned “private”; and if all encounters were self-pay or no insurance, insurance type was assigned to “none”. Insurance type was stratified by age groups, 1 to 10, 11 to 18, 19 to 64, as qualification for public insurance may vary by age. Sex (male, female, unknown), ethnicity (Hispanic, non-Hispanic, unknown), race (White, American Indian or Alaskan Native, Asian, Black or African American, Multi-racial, Native Hawaiian or Pacific Islander, unavailable/unknown), and data source type (Colorado All Payer Claims Database, clinical, hospital discharge, Medicaid, surgical data) were abstracted from the EHR or administrative data source and included in this analysis. As Down syndrome is the most common chromosomal abnormality associated with CHD[17], we also assessed co-occurring Down syndrome (yes, no; defined as having ≥ 1 ICD-9-CM diagnosis code of 758.0) with identified individuals.

Population data

The denominator for overall, age-specific, and sex-specific populations at each site was estimated by averaging 2011, 2012, and 2013 U.S. Census estimates (Supplemental Table 2).

Statistical analysis

Estimated percentage of cases at each site were calculated by dividing the number of cases by the total individuals in the general population for each site. Estimates per 1,000 population were provided overall and stratified by site, CHD severity, and age group (age group 1-10 years was reported for 5-county metropolitan-Atlanta, GA, 11-county NY, and statewide NC; all five sites reported age groups 11-18, 19-44, and 45-64 years). All analyses were performed using SAS software version 9.4 (SAS Institute Inc., Cary, NC). Compilation and sharing of de-identified data with the CDC were approved by each site's Institutional Review Board. De-identified, de-duplicated demographic, encounter, and summary-level data which combined and reconciled information from all data sources were transmitted by the five surveillance sites to CDC via a secure mechanism.

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Results

The data source types used in the surveillance system varied by site (Supplemental Table 3). Almost a third (31%) of all cases were found in two or more data sources. All sites used clinical databases, in which 75.5% of individuals were found (range across sites: 40.7%-100%). CO was the only site to use an all-payer claims data source which included all Medicaid insurance claims for the surveillance period, however the majority of cases identified in the claims dataset were also identified in clinical EHR.[16] The largest percentage of cases from GA (72.6%) and UT (99.2%) and all cases in NC were found in clinical data. For NY cases, 88.2% were found in hospital discharge data.

A total of 42,646 unique individuals with CHD-related ICD-9-CM code(s) were identified through this 5-site population-based surveillance system. The majority of the sample (across all sites) was male (51.5%), non-Hispanic (64.8%), and White (55.2%); however, race was missing for 13-34% of cases and ethnicity was missing for 3-44% of cases (Table 1). Severe CHD occurred in 23.7% of cases. Insurance type varied by age and by site. Overall, the majority of individuals residing in 5-county metropolitan-Atlanta, GA and 11-county NY had at least one encounter covered by public insurance (55.9% and 72.2%, respectively), whereas the majority of individuals in CO, NC, and UT had private insurance only (59.4%, 41.4%, and 72.8%, respectively). Public insurance was documented at one or more encounters for 61.9% of 1 to 10-year-olds (range: 43.1% in NC to 80.8% in NY), 42.7% of 11 to 18-year-olds (range: 26.7% in UT to 62.4% in NY), and 39.1% of 19 to 64-year-olds (range: 24.9% in NC to 67.6% in NY). Approximately 5.5% of cases also had Down syndrome (range across sites: 3.9%-7.4%).

Among the three sites that ascertained cases aged 1-64 years (5-county metropolitan-Atlanta, GA, 11-county NY, and statewide NC), the percentage of cases was 2.07 per 1,000; site-specific estimates ranged from 1.52 to 2.74 per 1,000 (Table 2). Among all five sites that

ascertained cases aged 11-64, the percentage of cases was 1.47 per 1,000; with site-specific estimates ranging from 1.02 to 2.18 per 1,000.

When stratified by age, for children ages 1-10 years from 5-county metropolitan-Atlanta, GA, 11-county NY, and statewide NC, the combined percentage of cases across sites was 6.36 per 1,000, but site-specific estimates ranged from 4.33 - 9.96 per 1,000 (Figure 1). Across all five sites, the percentage of cases declined with advancing age from 3.35 per 1,000 among 11 to 18-year-olds to 1.07 per 1,000 among 45 to 64-year-olds. Estimates of both severe and non-severe CHD decreased with age; however, among cases with non-severe CHD, the estimate increased slightly from 19-44 years to 45-64 years. This slight increase occurred among males only (Supplemental Figure 1).

Among 1 to 10-year-olds (based on three sites) and 11 to 64-year-olds (based on all five sites), cases were stratified by severity group (Figure 2). The highest estimates were observed for the 1 to 10-year-olds in the shunt group (2.6/1,000) and for the 11 to 64-year-olds in the valve group (0.7/1,000). Among those with non-severe CHD, the majority (52.8%) had valve defects, though the composition of the non-severe group varied by site.

Discussion

This is one of the first population-based, multi-site surveillance systems of 1 to 64-year-olds with CHD receiving health care in the U.S. This surveillance project expands upon previous U.S. population-based surveillance efforts among adolescents and adults and showed consistency across sites in estimates of individuals with CHD receiving health care.[9,13,14] At five U.S. sites, we found that one in 157 1 to 10-year-olds and one in 680 11 to 64-year-olds had CHD and received health care from 2011 to 2013, although estimates varied by surveillance site, CHD severity, and age group. These small variations by site are likely influenced by data sources utilized (eg, clinical versus administrative data), coding practices at healthcare institutions, and healthcare access and utilization. These estimates were lower than published estimates from Canada of 13 per 1,000 in children under 18 years and 6 per 1,000 in adults; however, these estimates may not be comparable to ours due to differences in healthcare systems, access to care, and insurance coverage.[3]

Overall, percentage of cases decreased from ages 1 to 10 to 19 to 44, which may reflect higher mortality rates with age among individuals with CHD compared to the general population. However, given survival has improved across all types of defects,[8] this decrease from ages 1 to 10 to 19 to 44 may also reflect individuals ages to 44 years with CHD receiving less health care during the 3-year surveillance period and having their CHD documented less frequently at healthcare encounters compared to 1 to 10-year-olds with CHD. Age 1 to 10 is when the majority of individuals with CHD are diagnosed and receive interventions. After reparative surgeries and interventions, individuals may begin to drop out of care.[10] In addition, once children reach adulthood, they may no longer have access to state insurance coverage programs only eligible to children, limiting their access to care,[18] which has been shown to be a concern for parents of adolescents with CHD from New York and Georgia.[19]

The pattern of CHD type differed by age group: among 1 to 10-year-olds, shunt defects were the most common defect type (2.6/1,000), whereas valve defects (0.7/1,000) were most common among 11 to 64-year-olds. Among males, the estimates for non-severe CHD increased slightly from 19-44 years to 45-64 years, which may be due to late manifestation of symptoms from CHD types more common in males leading to delayed diagnosis later in life, increased rates of cardiovascular disease (CVD), or increased CVD screening in males compared to females. E.g., bicuspid aortic valve is three times more common in men than women and valve pathology may not manifest until later ages.[20] In addition, acquired CVD related to changes in ventricular compliance may exacerbate CHD symptoms; CVD screening may lead to an incidental diagnosis of a non-symptomatic CHD; and/or coders may erroneously document acquired CVD as CHD. The percentage of severe CHD in the 45-64-year-old age group was lower across sites compared to the other age groups. It is possible the birth years of these cases cover a time where childhood congenital heart surgery was not available, or in the early stages, leading to lower survival in this age group.[8] Overall, 23.7% of CHD cases had severe CHD, which is higher than estimates from other countries or those modeled from Canadian healthcare data[5]. Given cases in this U.S. surveillance system were identified through healthcare encounters with a documented CHD during a three-year period, it is possible that the most complex patients may have sought care more often than less complex cases, leading to a higher proportion with severe CHD in our data.

The characteristics of individuals living with CHD in this sample differ from the general population. Almost half of individuals with CHD had public insurance at one or more healthcare encounters, compared to 25% of the U.S. general population under the age of 65 years in 2013.[21] This may reflect differences in methodology between estimates (our estimate is based on individuals receiving health care), differences in data sources used by sites, or differences in insurance eligibility requirements, such as disability status, among people with CHD compared to the general population.[21] Over 5% of individuals in our analysis had Down syndrome, which has a birth prevalence of approximately 1 in 700 births, a lifespan prevalence of 1 in 1200, and a life expectancy of 47 years.[22,23] Understanding population estimates of CHD across the lifespan, in relation to defect type, healthcare coverage, and genetic syndromes can improve planning for the needs of the growing CHD population.

This project has several strengths. This is the first population-based effort among five diverse geographic sites to conduct surveillance of CHD in the U.S. All surveillance sites adhered to a uniform project methodology, including case definition and data elements, allowing for comparison of data across sites. This project expands upon a previous population-based surveillance effort that reported adolescent and adult estimates across three sites.[9,13,14] Sites linked and deduplicated multiple clinical and/or administrative data sources; 31% of individuals were found in two or more data sources, indicating value in linking multiple data sources to ascertain healthcare encounters among cases. Despite site-specific data source variations, the total estimates and severe/non-severe stratified estimates were largely consistent across sites.

This project is subject to several limitations. The data are from five distinct surveillance sites and stratified and total estimates may not be generalizable to other U.S. sites, although, as

stated above, estimates were similar across sites. The estimates from each site represents individuals who had a healthcare encounter with a documented ICD-9-CM code for CHD during the three-year surveillance period and may not accurately estimate the population of individuals with CHD. Individuals with CHD residing within site catchment areas may not have received care during the study's three year-period, may not have had their CHD documented at their healthcare encounters, or may have sought care that was not captured in the sites' data sources, thereby under-estimating in the true prevalence of individuals living with CHD. Also, CHD-related ICD-9-CM codes in cardiology clinic data show good sensitivity and specificity[24], whereas CHD-related ICD-9-CM codes in administrative data have a lower PPV[15,25,26] However, the accuracy of CHD-related ICD-9-CM codes in administrative data improves with greater CHD severity and with the exclusion of non-specific CHD codes[25] and code 745.5 in isolation. [15] To improve upon these known limitations with CHD-related ICD-9-CM codes, we excluded cases with non-specific CHD codes (Supplemental Table 1) and those with an isolated code of 745.5, which may result in an underestimation of prevalence. A preliminary validation study of the current data showed that the PPV for total cases was approximately 80% after excluding isolated 745.5 and non-specific CHD codes; thus, one out of five cases in our surveillance system may not have CHD. Although data from all available sources were used, overall race data were missing for 12% to 33% of cases and ethnicity data were missing for 3% to 44% of cases, limiting our ability to understand differences and further explore previously documented disparities in morbidity and mortality by race and ethnicity[27].

Conclusions

This is one of the first population-based, multi-site surveillance systems of 1 to 64-year-olds with CHD receiving health care. Despite site-specific data source variations, the total estimate and those stratified by CHD severity were largely consistent across sites. Results from this surveillance project show a substantial number of individuals with CHD residing in the five sites, with diversity by CHD severity, age, race/ethnicity, sex, Down syndrome, and insurance coverage. These three years of population-based healthcare data in 5 sites for individuals with CHD across the lifespan will provide information on healthcare encounters and procedures, comorbidities, mortality, demographic characteristics, and health insurance by CHD severity. Project findings from this analysis, as well as future ones, will inform planning for the needs of the growing population of individuals living with CHD.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Abbreviations

ASD	atrial septal defect
CHD	congenital heart defect
ICD-9-CM	International Classification of Disease-9th Revision-Clinical Modification
PFO	patent foramen ovale
CO	Colorado
GA	Georgia
NC	North Carolina
NY	New York
UT	Utah

References

1. Reller MD, Strickland MJ, Riele-Colarusso TJ, et al. Prevalence of congenital heart defects in metropolitan Atlanta, 1998-2005. J Pediatr 2008;153(6):807–13. [PubMed: 18657826]
2. Canfield MA, Honein MA, Yuskiv N, et al. . National estimates and race/ethnic-specific variation of selected birth defects in the United States, 1999-2001. Birth Defects Res A Clin Mol Teratol 2006;76(11):747–56. [PubMed: 17051527]
3. Marelli AJ, Ionescu-Ittu R, Mackie AS, et al. . Lifetime prevalence of congenital heart disease in the general population from 2000 to 2010. Circulation 2014;130(9):749–56. [PubMed: 24944314]
4. Marelli AJ, Ionescu-Ittu R, Mackie AS, et al. . Congenital heart disease in the general population: changing prevalence and age distribution. Circulation 2007;115(2):163–72 [PubMed: 17210844]
5. Gilboa SM, Devine OJ, Kucik JE, et al. . Congenital Heart Defects in the United States: Estimating the Magnitude of the Affected Population in 2010. Circulation 2016;134(2):101–9. [PubMed: 27382105]
6. van der Bom T, Bouma BJ, Meijboom FJ, et al. . The prevalence of adult congenital heart disease, results from a systematic review and evidence based calculation. Am Heart J 2012;164(4):568–75. [PubMed: 23067916]
7. Oster ME, Lee KA, Honein MA, et al.. Temporal Trends in Survival Among Infants With Critical Congenital Heart Defects 2013;131(5):e1502–8.
8. Spector LG, Menk JS, Knight JH, et al. . Trends in Long-Term Mortality After Congenital Heart Surgery. J Am Coll Cardiol 2018;71(21):2434–46. [PubMed: 29793633]
9. Glidewell J, Book W, Raskind-Hood C, et al. . Population-based surveillance of congenital heart defects among adolescents and adults: surveillance methodology. Birth Defects Res 2018;110(19):1395–403. [PubMed: 30394691]
10. Gurvitz M, Valente AM, Broberg C, et al. . Prevalence and predictors of gaps in care among adult congenital heart disease patients: HEART-ACHD (The Health, Education, and Access Research Trial). J Am Coll Cardiol 2013;61(21):2180–4. [PubMed: 23542112]
11. Tompkins R, Khan A. ACHD Care in the United States: Complex Patients, Even More Complex Health Care System. J Am Coll Cardiol 2020;76(2):183–5. [PubMed: 32646568]

12. Connor JA, Gauvreau K, Jenkins KJ. Factors associated with increased resource utilization for congenital heart disease. Pediatrics 2005;116(3):689–95. [PubMed: 16140709]
13. Gurvitz M, Dunn JE, Bhatt A, et al. . Characteristics of Adults With Congenital Heart Defects in the United States. J Am Coll Cardiol 2020;76(2):175–82. [PubMed: 32646567]
14. Lui GK, McGarry C, Bhatt A, et al. . Surveillance of Congenital Heart Defects among Adolescents at Three U.S. Sites. Am J Cardiol 2019;124(1):137–43. [PubMed: 31030970]
15. Rodriguez FH 3rd, Ephrem G, Gerardin JF, et al. . The 745.5 issue in code-based, adult congenital heart disease population studies: Relevance to current and future ICD-9-CM and ICD-10-CM studies. Congenit Heart Dis 2018;13(1):59–64. [PubMed: 29266726]
16. Crume TL, Duca LM, Ong T, et al. . Population-level surveillance of congenital heart defects among adolescents and adults in Colorado: Implications of record linkage. Am Heart J 2020;226:75–84. [PubMed: 32526532]
17. Pierpont ME, Brueckner M, Chung WK, et al. . Genetic Basis for Congenital Heart Disease: Revisited: A Scientific Statement From the American Heart Association. Circulation 2018;138(21):e653–711. [PubMed: 30571578]
18. Centers for Medicare & Medicaid Services, C.f.M.a.C.S., Division of Quality and Health Outcomes. Medicaid and CHIP Beneficiaries at a Glance. 2020; Available from: https://www.medicaid.gov/medicaid/quality-of-care/downloads/beneficiary-ataglance.pdf.
19. Gaydos LM, Sommerhalter K, Raskind-Hood C, et al. Health Care Transition Perceptions Among Parents of Adolescents with Congenital Heart Defects in Georgia and New York. Pediatr Cardiol 2020;41(6):1220–30. [PubMed: 32500288]
20. Kong WK, Regeer MV, Ng A, et al. . Sex Differences in Phenotypes of Bicuspid Aortic Valve and Aortopathy: Insights From a Large Multicenter, International Registry. Circ Cardiovasc Imaging 2017;10(3).
21. Smith J, Medalia CHealth Insurance Coverage in the United States: 2013. 2014 [cited 2020 July 28, 2020]; Available from: https://www.census.gov/library/publications/2014/demo/p60-250.html.
22. Mai CT, Isenburg JL, Canfield MA, et al. . National population-based estimates for major birth defects, 2010-2014. Birth Defects Res 2019;111(18):1420–35. [PubMed: 31580536]
23. Presson AP, Partyka G, Jensen KM, et al. . Current estimate of Down Syndrome population prevalence in the United States. J Pediatr 2013;163(4):1163–8. [PubMed: 23885965]
24. Broberg C, McLarry J, Mitchell J, et al. . Accuracy of administrative data for detection and categorization of adult congenital heart disease patients from an electronic medical record. Pediatr Cardiol 2015;36(4):719–25. [PubMed: 25428778]
25. Khan A, Ramsey K, Ballard C, et al. . Limited Accuracy of Administrative Data for the Identification and Classification of Adult Congenital Heart Disease. J Am Heart Assoc 2018;7(2). doi:10.1161/JAHA.117.007378.
26. Riehle-Colarusso TJ, Bergersen L, Broberg CS, et al. . Databases for Congenital Heart Defect Public Health Studies Across the Lifespan. J Am Heart Assoc 2016;5(11).
27. Lopez KN, Morris SA, Sexson Tejtel SK, et al. , US Mortality Due to Congenital Heart Disease Across the Lifespan from 1999-2017 Exposes Persistent Racial/Ethnic Disparities. Circulation 2020;142:1132–47. [PubMed: 32795094]

Clinical perspectives

This is one of the first population-based, multi-site surveillance systems of 1 to 64-year-olds with CHD receiving health care. Results from this surveillance effort show a substantial number of individuals with CHD residing in the five U.S. sites, with diversity in CHD severity, age, race/ethnicity, sex, Down syndrome, and insurance coverage. These three years of population-based healthcare data in five U.S. sites for individuals with CHD across the lifespan will provide information on healthcare encounters and procedures, comorbidities, mortality, demographic characteristics, and health insurance by CHD severity. Project findings from this analysis will aid in planning for the needs of the growing population of individuals living with CHD.

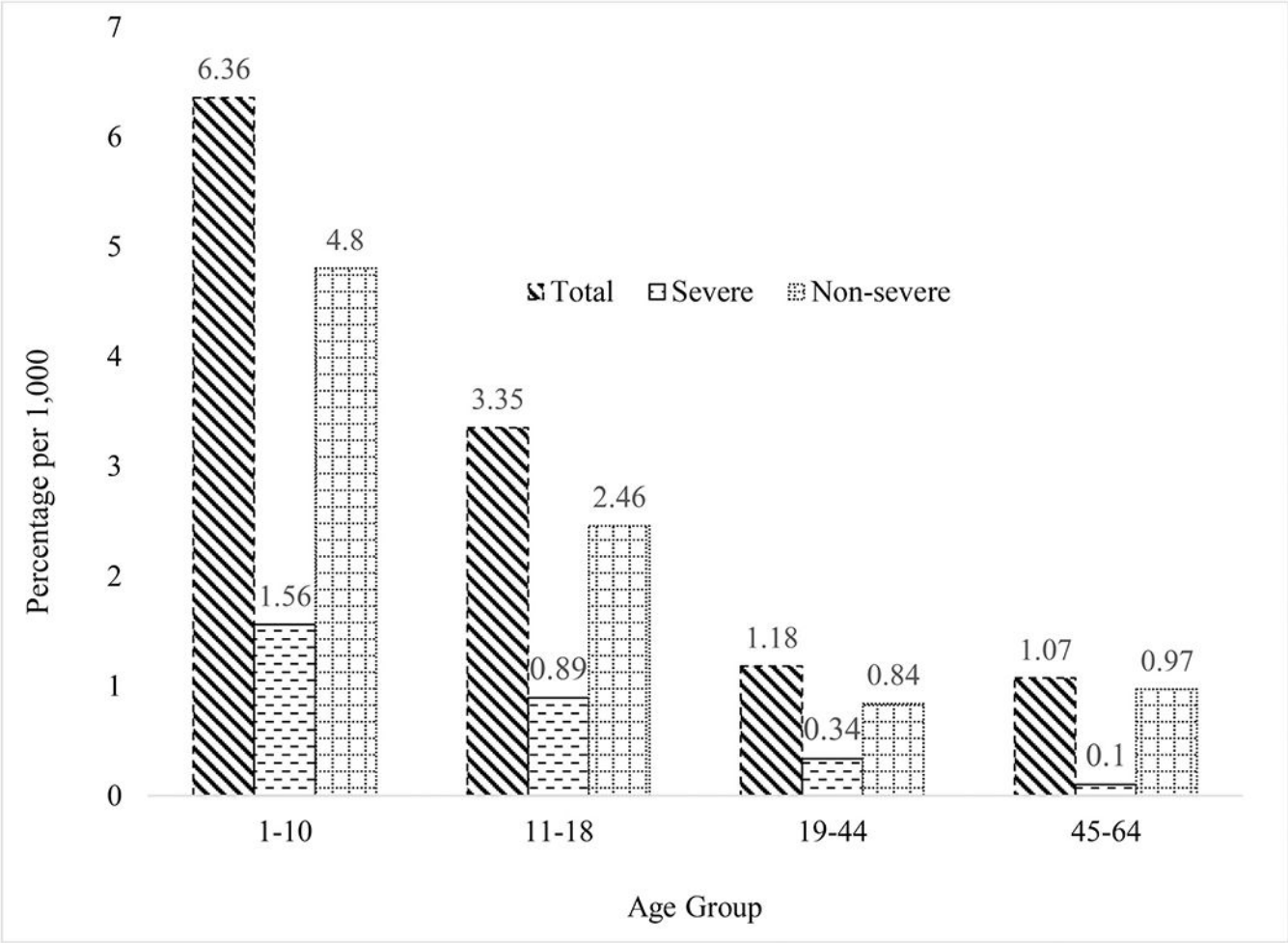

Figure 1.

Percentage by age group of 1 to 64-y-olds with a CHD-related ICD-9-CM code* per 1,000 individuals, total and by CHD severity†, 5 sites‡, 2011-2013.

*Excluding isolated 745.5, 745.5 co-occurring with a non-specific CHD code, or ≥ 1 non-specific CHD code. †Non-severe = shunt + valve, valve, and shunt categories combined. ‡Estimate for 1 to 10-year-olds based on 3 sites: 5-county metropolitan-Atlanta, GA, 11 counties in NY state, statewide in NC; 11-18, 19-44, and 45-64-y-old estimates based on 5 sites: statewide in CO, 5-county metropolitan-Atlanta, GA, 11 counties in NY state, statewide in NC, and statewide in UT.

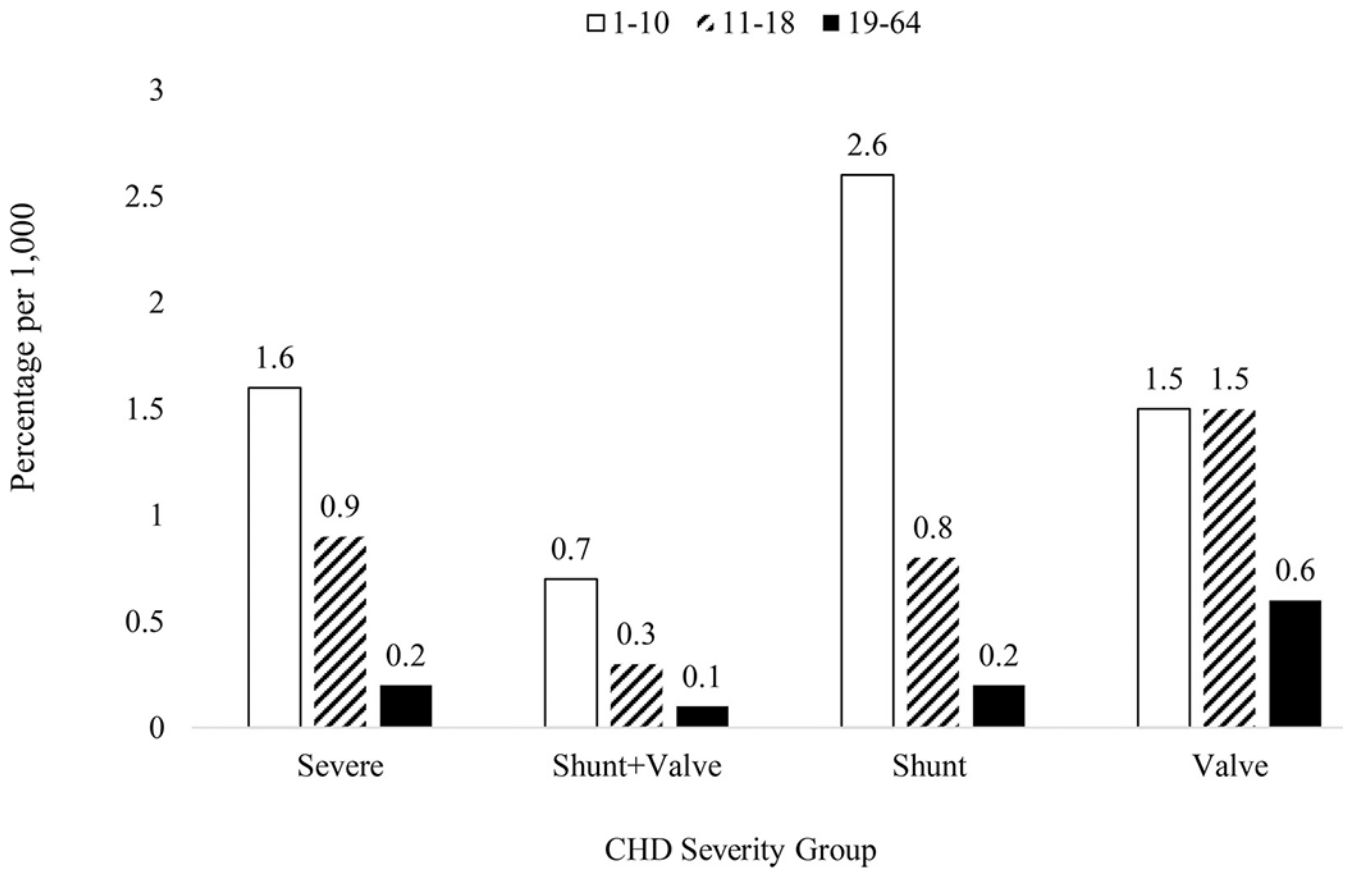

Figure 2.

Percentage of 1 to 10, 11 to 18 and 19 to 64-y-olds with a CHD-related ICD-9-CM code* per 1,000 individuals, by CHD severity group, 5 sites†, 2011-2013.

*Excluding isolated 745.5, 745.5 co-occurring with a non-specific CHD code, or ≥1 non-specific CHD code. †Estimate for 1 to 10-y-olds based on 3 sites: 5-county metropolitan-Atlanta, GA, 11 counties in NY state, statewide in NC; 11-18, 19-44, and 45-64-y-old estimates based on 5 sites: statewide in CO, 5-county metropolitan-Atlanta, GA, 11 counties in NY state, statewide in NC, and statewide in UT.

Table 1.

Clinical and demographic characteristics of 1 to 64-year-olds with a CHD-related ICD-9-CM code*, overall and by surveillance site, 2011-2013.

	CO N (%)	Metropolitan-Atlanta, GA N (%)	11-county NY N (%)	NC N (%)	UT N (%)	Total N (%)
Total	6,511	8,312	10,801	12,618	4,404	42,646
Severity						
Severe	1,356 (20.8)	2,312 (27.8)	2,340 (21.7)	3,116 (24.7)	984 (22.3)	10,108 (23.7)
Non-severe, combined	5,155 (79.2)	6,000 (72.2)	8,461 (78.3)	9,502 (75.3)	3,420 (77.7)	32,538 (76.3)
Shunt + valve	404 (7.8)	800 (13.3)	926 (10.9)	901 (9.5)	309 (9.0)	3,340 (10.3)
Shunt	1,330 (25.8)	2,809 (46.8)	3,696 (43.7)	3,409 (35.9)	766 (22.4)	12,010 (36.9)
Valve	3,421 (66.4)	2,391 (39.9)	3,839 (45.4)	5,192 (54.6)	2,345 (68.6)	17,188 (52.8)
Age Group						
1-10 y	N/A	4,937 (59.4)	4,496 (41.6)	5,478 (43.4)	N/A	14,911 (35)
11-64 y	6,511 (100)	3,375 (40.6)	6,305 (58.4)	7,140 (56.6)	4,404 (100)	27,735 (65)
11-18 y	1,988 (30.5)	1,546 (45.8)	1,823 (28.9)	2,456 (34.4)	1,725 (39.2)	9,538 (34.4)
19-44 y	2,656 (40.8)	1,239 (36.7)	2,642 (41.9)	2,830 (39.6)	1,700 (38.6)	11,067 (39.9)
45-64 y	1,867 (28.7)	590 (17.5)	1,840 (29.2)	1,854 (26)	979 (22.2)	7,130 (25.7)
Race						
American Indian or Alaskan Native	176 (2.7)	39 (0.5)	36 (0.3)	109 (0.9)	N/A	360 (0.8)
Asian	219 (3.4)	229 (2.8)	240 (2.2)	177 (1.4)	N/R	867 (2)
Black or African American	194 (3)	2,444 (29.4)	2,304 (21.3)	2,456 (19.5)	24 (0.5)	7,422 (17.4)
Multi-racial	196 (3)	90 (1.1)	506 (4.7)	137 (1.1)	15 (0.3)	944 (2.2)
Native Hawaiian or Pacific Islander	24 (0.4)	N/R	N/R	10 (0.1)	N/A	45 (0.1)
White	3,518 (54)	3,253 (39.1)	5,291 (49)	8,137 (64.5)	3,352 (76.1)	23,551 (55.2)
Unavailable/Unknown	2,184 (33.5)	2,253 (27.1)	2,417 (22.4)	1,592 (12.6)	1,011 (23)	9,457 (22.2)
Ethnicity						
Hispanic	951 (14.6)	552 (6.6)	3,535 (32.7)	981 (7.8)	329 (7.5)	6,348 (14.9)
Non-Hispanic	4,098 (62.9)	5,408 (65.1)	6,930 (64.2)	9,038 (71.6)	2158 (49)	27,632 (64.8)
Unavailable/Unknown	1,462 (22.5)	2,352 (28.3)	336 (3.1)	2,599 (20.6)	1,917 (43.5)	8,666 (20.3)
Sex						
Male	3,374 (51.8)	4,192 (50.4)	5,300 (49.1)	6,718 (53.2)	2,368 (53.8)	21,952 (51.5)
Female	3,133 (48.1)	4,120 (49.6)	5,500 (50.9)	5,900 (46.8)	2,036 (46.2)	20,689 (48.5)

	CO N (%)	Metropolitan-Atlanta, GA N (%)	11-county NY N (%)	NC N (%)	UT N (%)	Total N (%)
Unavailable/Unknown	N/R[†]	N/A	N/R	N/A	N/A	N/A
Insurance type [≠]						
1-64 y						
Any Public	2,490 (38.2)	4,645 (55.9)	7,801 (72.2)	4,332 (34.3)	1,153 (26.2)	20,421 (47.9)
Private (no public)	3,870 (59.4)	3,564 (42.9)	2,931 (27.1)	5,223 (41.4)	3,207 (72.8)	18,795 (44.1)
None	15 (0.2)	23 (0.3)	N/A	63 (0.5)	12 (0.3)	117 (0.3)
Unknown	136 (2.1)	80 (1)	65 (0.6)	3,000 (23.8)	32 (0.7)	3,313 (7.8)
1-10 y						
Any Public	N/A	3,243 (65.7)	3,633 (80.8)	2360 (43.1)	N/A	9,236 (61.9)
Private (no public)	N/A	1676 (33.9)	851 (18.9)	1830 (33.4)	N/A	4,357 (29.2)
None	N/A	17 (0.3)	12 (0.3)	44 (0.8)	N/A	73 (0.5)
Unknown	N/A	N/R	N/A	1244 (22.7)	N/A	1,245 (8.3)
11-18 y						
Any Public	940 (47.3)	727 (47)	1,137 (62.4)	804 (32.7)	460 (26.7)	4,068 (42.7)
Private (no public)	1,006 (50.6)	810 (52.4)	674 (37)	1,022 (41.6)	1,251 (72.5)	4,763 (49.9)
None	39 (2)	N/A	12 (0.7)	29 (1.2)	N/R	91 (1)
Unknown	N/R	N/R	N/A	601 (24.5)	11 (0.6)	616 (6.5)
19-64 y						
Any Public	1,550 (34.3)	675 (36.9)	3,031 (67.6)	1,168 (24.9)	693 (25.9)	7,117 (39.1)
Private (no public)	2,864 (63.3)	1,078 (58.9)	1,406 (31.4)	2,371 (50.6)	1,956 (73)	9,675 (53.2)
None	93 (2.1)	53 (2.9)	44 (1)	135 (2.9)	16 (0.6)	341 (1.9)
Unknown	16 (0.4)	23 (1.3)	N/R	1,010 (21.6)	14 (0.5)	1,064 (5.8)
Down Syndrome						
Yes	263 (4.0)	619 (7.4)	743 (6.9)	539 (4.3)	173 (3.9)	2,337 (5.5)
No	6,248 (96.0)	7,693 (92.6)	10,058 (93.1)	12,079 (95.7)	4,231 (96.1)	40,309 (94.5)

[*] Excluding isolated 745.5, 745.5 co-occurring with a non-specific CHD code, or ≥ 1 non-specific CHD code.

[†] N/R = Not Reportable; cell sizes less than 10 have been omitted.

[≠] Public defined as Medicaid, Medicare; Private defined as private, other government, or other insurance; None defined as self-pay/uninsured. Unknown defined as unavailable, unknown, or no insurance was checked.

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Table 2.

Age group percentages of 1 to 64-year-olds with a documented CHD-related ICD-9-CM code in at least one healthcare encounter* per 1,000, total and by CHD severity†, 5 sites‡, 2011-2013.

	Colorado		Metropolitan-Atlanta, GA		11-County New York		North Carolina		Utah		Total	
	N	**Percentage (1/1000)**	**n**	**Percentage (1/1000)**	**N**	**Percentage (1/1000)**	**n**	**Percentage (1/1000)**	**n**	**Percentage (1/1000)**	**N**	**Percentage (1/1000)**
Total, 1-64 y	N/A	N/A	8,312	2.66	10,801	2.74	12,618	1.52	N/A	N/A	31,731	2.07
Severe			2,312	0.74	2,340	0.59	3,116	0.38			7,768	0.51
Non-severe, combined			6,000	1.92	8,461	2.15	9,502	1.15			23,963	1.56
Total, 1-10 y	N/A	N/A	4,937	9.96	4,496	7.71	5,478	4.33	N/A	N/A	14,911	6.36
Severe			1,310	2.64	1,023	1.75	1,321	1.04			3,654	1.56
Non-severe, combined			3,627	7.32	3,473	5.95	4,157	3.28			11,257	4.80
Total, 11-18 y	1,988	3.64	1,546	3.97	1,823	3.57	2,456	2.39	1,725	4.70	9,538	3.35
Severe	522	0.96	456	1.17	474	0.93	643	0.62	448	1.22	2,543	0.89
Non-severe, combined	1,466	2.68	1,090	2.80	1,349	2.64	1,813	1.76	1,277	3.48	6,995	2.46
Total, 19-44 y	2,656	1.39	1,239	0.91	2,642	1.65	2,830	0.83	1,700	1.56	11,067	1.18
Severe	672	0.35	456	0.34	691	0.43	929	0.27	469	0.43	3,217	0.34
Non-severe, combined	1,984	1.04	783	0.58	1,951	1.22	1,901	0.56	1,231	1.13	7,850	0.84
Total, 45-64 y	1,867	1.36	590	0.67	1,840	1.47	1,854	0.72	979	1.73	7,130	1.07
Severe	162	0.12	90	0.10	152	0.12	223	0.09	67	0.12	694	0.10
Non-severe, combined	1,705	1.25	500	0.57	1,688	1.35	1,631	0.63	912	1.61	6,436	0.97
Total, 11-64 y	6,511	1.70	3,375	1.29	6,305	1.88	7,140	1.02	4,404	2.18	27,735	1.47
Severe	1,356	0.35	1,002	0.38	1,317	0.39	1,795	0.26	984	0.49	6,454	0.34
Non-severe, combined	5,155	1.35	2,373	0.91	4,988	1.49	5,345	0.76	3,420	1.69	21,281	1.13

*Excluding isolated 745.5, 745.5 co-occurring with a non-specific CHD code, or ≥ 1 non-specific CHD codes.

†Non-severe = shunt + valve, valve, and shunt categories combined.

‡Estimate for 1 to 10-year-olds based on 3 sites: 5-county metropolitan-Atlanta, GA, 11 counties i n NY state, statewide in NC; 11-18, 19-44, and 45-64-year-old estimates based on 5 sites: statewide in CO, 5-county metropolitan-Atlanta, GA, 11 counties in NY state, statewide in NC, and statewide in UT.