

Surveillance of Congenital Heart Defects among Adolescents at Three U.S. Sites



George K. Lui, MD^{a,*}, Claire McGarry, MPH^b, Ami Bhatt, MD^c, Wendy Book, MD^d, Tiffany J. Riehle-Colarusso, MD, MPH^e, Julie E. Dunn, PhD^f, Jill Glidewell, MSN, MPH^e, Michelle Gurvitz, MD^g, Trenton Hoffman, MS^h, Carol J. Hogue, PhD, MPH^h, Daphne Hsu, MDⁱ, Stan Obenhaus^f, Cheryl Raskind-Hood, MPH, MS^h, Fred H. Rodriguez, III, MD^d, Ali Zaidi, MDⁱ, and Alissa R. Van Zutphen, PhD^b

The prevalence, co-morbidities, and healthcare utilization in adolescents with congenital heart defects (CHDs) is not well understood. Adolescents (11 to 19 years old) with a health-care encounter between January 1, 2008 (January 1, 2009 for MA) and December 31, 2010 with a CHD diagnosis code were identified from multiple administrative data sources compiled at 3 US sites: Emory University, Atlanta, Georgia (EU); Massachusetts Department of Public Health (MA); and New York State Department of Health (NY). The estimated prevalence for any CHD was 4.77 (EU), 17.29 (MA), and 4.22 (NY) and for severe CHDs was 1.34 (EU), 3.04 (MA), and 0.88 (NY) per 1,000 adolescents. Private or commercial insurance was the most common insurance type for EU and NY, and Medicaid for MA. Inpatient encounters were more frequent in severe CHDs. Cardiac co-morbidities included rhythm and conduction disorders at 20% (EU), 46% (MA), and 9% (NY) as well as heart failure at 3% (EU), 15% (MA), and 2% (NY). Leading noncardiac co-morbidities were respiratory/pulmonary (22% EU, 34% MA, 16% NY), infectious disease (17% EU, 22% MA, 20% NY), non-CHD birth defects (12% EU, 23% MA, 14% NY), gastrointestinal (10% EU, 28% MA, 13% NY), musculoskeletal (10% EU, 32% MA, 11% NY), and mental health (9% EU, 30% MA, 11% NY). In conclusion, this study used a novel approach of uniform CHD definition and variable selection across administrative data sources in 3 sites for the first population-based CHD surveillance of adolescents in the United States. High resource utilization and co-morbidities illustrate ongoing significant burden of disease in this vulnerable population. © 2019 The Authors. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license. (<http://creativecommons.org/licenses/by-nc-nd/4.0/>) (Am J Cardiol 2019;124:137–143)

Congenital heart defects (CHDs) are one of the most common birth defects in the United States.^{1–3} Advances in the treatment and management of infants and children with CHDs have improved survival; estimates suggest that there are over 2 million individuals with CHDs living in the United States in 2010.⁴ This population is vulnerable to

loss to follow-up from cardiology care with attrition rates of 20%, beginning at ages 6 to 12.⁵ Successful transition of care to adult specialists has been associated with improved survival, supporting the importance of specialized care for CHD patients.⁶ Suboptimal transition predisposes adolescents to delayed recognition of residual hemodynamic abnormalities and may further complicate their ongoing cardiac disease.^{7,8} Previous studies have examined the prevalence of CHDs in adolescents based on the birth prevalence of CHDs and administrative databases.^{2–4} A recent study using Canadian administrative data estimated 12.62 CHDs per 1,000 and 1.87 severe CHDs per 1,000 in adolescents aged 13 to 17.⁴ From 1985 to 2000 in Quebec, Canada, there was a 22% increase in the number of children surviving with severe CHDs.⁹ However, it is important to estimate US prevalence using US administrative databases. The goal of this study is to estimate population-based prevalence, healthcare utilization, and co-morbidities in adolescents who had a healthcare encounter through linkage of existing data from multiple sources.

^aStanford University School of Medicine, Stanford, California; ^bNew York State Department of Health, Albany, New York; ^cMassachusetts General Hospital, Boston, Massachusetts; ^dEmory University, Atlanta, Georgia; ^eCenters for Disease Control and Prevention (CDC), National Center on Birth Defects and Developmental Disabilities, Atlanta, Georgia; ^fMassachusetts Department of Health, Boston, Massachusetts; ^gBoston Children's Hospital and Brigham and Women's Hospital, Boston, Massachusetts; ^hEmory University, Rollins School of Public Health, Atlanta, Georgia; and ⁱAlbert Einstein College of Medicine, Bronx, New York. Manuscript received December 1, 2018; revised manuscript received and accepted March 21, 2019.

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See page 143 for disclosure information.

*Corresponding author: Tel: 650-723-1663; fax: 650-725-1599.

E-mail address: glui@stanford.edu (G.K. Lui).

Methods

Overall study methods are detailed in a separate study.¹⁰ Cases of CHDs were identified using *International*

Classification of Disease version 9.0 Clinical Modification (ICD-9-CM) diagnostics codes 745.xx-747.xx, 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) (Supplemental Table 1). The CHD diagnostic codes were classified into mutually exclusive hierarchical groups similar to Marelli et al.,^{9,11} integrating both hemodynamic severity and basic anatomy: severe, shunt (excludes isolated 745.5), shunt + valve, valve, other CHDs, and isolated 745.5 (Supplemental Table 1). Isolated 745.5 is used to code both atrial septal defect (ASD) and patent foramen ovale. While ASD represents a true CHD lesion, a patent foramen ovale is a normal newborn condition that may persist in adulthood. Including 745.5 in the CHD case definition may lead to overestimation of true CHDs because most of these lesions close in the newborn period. Therefore it was considered separately. We used 3 analytic categories: (1) severe CHDs, (2) nonsevere CHDs, and (3) isolated 745.5. Severe CHDs were: endocardial cushion defects, interrupted aortic arch, tetralogy of Fallot, total anomalous pulmonary venous return, transposition complexes, truncus, and univentricular hearts. Cases with only 1 code were classified according to these 3 groups. If there were multiple codes, cases were classified as severe if they had at least 1 severe CHD code (regardless of the presence of other CHD codes). Supplemental Figure 1 illustrates the algorithm for case classification in the presence of multiple CHD codes.

Adolescents between the ages of 11 and 19 with CHD diagnosis codes who had a healthcare encounter with a CHD diagnosis code between January 1, 2008 (January 1, 2009 for MA) and December 31, 2010 and who were presumed alive as of January 1, 2010 were identified using administrative and clinical data sources. EU identified cases in 5 metropolitan Atlanta area counties (Clayton, Cobb, Dekalb, Fulton, and Gwinnett) in Georgia using Medicaid claims data and administrative and clinical data from 6 pediatric and adult care facilities. MA identified cases statewide using the MA All Payer Claims Database, and pediatric and adult care facilities. NY identified cases in 11 counties (Allegany, Cattaraugus, Chautauqua, Erie, Genesee, Monroe, Niagara, Orleans, and Wyoming in the west, and Bronx and Westchester in the south) based on administrative data from 7 pediatric cardiology clinics and hospital inpatient and outpatient data from the New York Statewide Planning and Research Cooperative System.

Descriptive variables included age, gender, height, weight, and insurance status, based on health insurance source for the encounter nearest January 1, 2010. To verify the vital status as of January 1, 2010, sites attempted to link identified cases to vital records data. Race and ethnicity are not presented because they were unknown in 50% (EU), 40% (MA), and 36% (NY) of cases.

Healthcare utilization was assessed from outpatient, inpatient, and emergency department (ED) encounters. For multiple encounters on the same day, the patient was counted as having only 1 encounter, coded using the following hierarchy: (1) inpatient, (2) emergency department, and (3) outpatient. ICD-9-CM codes unrelated to

CHD diagnoses and all Current Procedural terminology (CPT) across all encounters were grouped into categories of co-morbidities and procedures using the Clinical Classification Software (CCS) tool. Developed by the Agency for Healthcare Research and Quality, CCS is a categorization scheme which collapses thousands of ICD-9-CM and CPT codes into over 200 diagnostic and procedure categories.¹² Noncardiac diagnostic CCS categories were further collapsed into 24 broader co-morbidity groups for this project (Supplemental Table 2). The ICD-9-CM and CPT codes in the cardiac- and vascular-related procedural CCS categories were collapsed into the following project-specific cardiac procedure categories: cardiac procedures and surgeries, diagnostic imaging, and vascular procedures (Supplemental Table 3). Procedural codes not specified in these categories were considered noncardiac procedures. Cases were only reported as having a noncardiac procedure (eg, knee replacement [yes/no]); the specific noncardiac procedural codes were not reported in the final dataset.

The denominator for age-specific prevalence at each site was estimated by using county-level 2010 US Census data. Prevalence was defined as the number of resident cases who were alive as of January 1, 2010 who had a CHD diagnosis code in the study period (2008 (MA: 2009) to 2010) divided by the total population of the corresponding study area according to 2010 census data, reported per 1,000 individuals. Statistical tests were performed to examine differences in the distribution of demographics, healthcare utilization, and co-morbidities by CHD severity for each US site. Chi-square tests were used to compare categorical variables: age (11 to 13, 14 to 16, 17 to 19), gender, insurance status, type of healthcare visit, type of procedure, and co-morbidity categories. The Fisher's exact 2-sided p value was used when a cell size was less than 5. The student's *t* test was used to examine both differences in the average number of days in care and the average number of selected procedures by severity. All descriptive analyses were performed with SAS software (SAS/STAT software version 9.3, SAS Institute, Inc, Cary, NC). Compilation and sharing of deidentified data with the CDC was approved by each participating site's Institutional Review Board. Deidentified, deduplicated demographic, encounter, and summary data which combined and reconciled information from multiple data sources were transmitted by all 3 sites to CDC via a secure mechanism.

Results

As of January 1, 2010, the estimated adolescent prevalence of all CHDs was 4.77 (EU), 17.29 (MA), and 4.22 (NY) per 1,000 adolescents, and of severe CHDs was 1.34 (EU), 3.04 (MA), and 0.88 (NY) per 1,000 adolescents (Table 1).

In all 3 sites most CHD cases had only 1 CHD ICD-9-CM code (Table 1). The proportion of isolated 745.5 cases ranged from 6% to 12% whereas the proportion of severe CHDs in total CHDs ranged from 18% to 28% at the 3 sites. The top 5 most frequent severe CHD codes varied across the 3 sites and included the following: complete transposition of great arteries, double outlet right ventricle, endocardial cushion defect, hypoplastic left heart syndrome,

Table 1
CHD prevalence and severity among adolescents at the Emory, Massachusetts, and New York sites as of January 1, 2010

| CHD diagnosis | Emory | | Massachusetts | | New York | |
|------------------------------------|--------------|---|---------------|---|--------------|---|
| | | Prevalence based on 2010 census population (Total = 424,336)* | | Prevalence based on 2010 census population (Total = 788,402)* | | Prevalence based on 2010 census population (Total = 600,809)* |
| Total cases | 2026 | 4.77 | 13629 | 17.29 | 2537 | 4.22 |
| Severe | 567 (28.0%) | 1.34 | 2400 (17.6%) | 3.04 | 527 (20.8%) | 0.88 |
| Nonsevere | 1215 (60.0%) | 2.86 | 10438 (76.6%) | 13.24 | 1745 (68.8%) | 2.90 |
| Shunt (excludes isolated 745.5) | 528 (26.1%) | | 3477 (25.5%) | | 628 (24.8%) | |
| Shunt + valve | 78 (3.9%) | | 311 (2.3%) | | 101 (4.0%) | |
| Valve | 399 (19.7%) | | 2176 (16.0%) | | 742 (29.3%) | |
| Other | 210 (10.4%) | | 4474 (32.8%) | | 274 (10.8%) | |
| Isolated 745.5 | 244 (12.0%) | 0.58 | 791 (5.8%) | 1.00 | 265 (10.5%) | 0.44 |
| Type of CHD diagnosis | | | | | | |
| Isolated (Excludes 745.5) | 1127 (55.6%) | | 7901 (58.0%) | | 1623 (64.0%) | |
| Multiple | 655 (32.3%) | | 4937 (36.2%) | | 649 (25.6%) | |

*Prevalence per 1,000.

pulmonary valve atresia, single ventricle, and tetralogy of Fallot (Supplemental Table 4). Shunt (excluding 745.5) and valve were the largest proportion of nonsevere CHDs in NY (25% and 29%) and EU (26% and 10%), respectively. The largest categories in MA were other (33%) and shunt (26%).

EU and NY had more male than female cases in the non-severe and severe CHDs (Table 2). MA had an even distribution of male and female nonsevere and severe CHD cases. The age distribution differed by CHD severity in MA and NY data (Table 2). The largest proportion of cases were 14 to 16 years old in the severe and nonsevere groups for all 3 sites; whereas cases were more often 17 to 19 years old in the isolated 745.5 (MA 37%; NY 38%). Most cases had private/commercial insurance in EU (62%) and NY (61%), whereas Medicaid was the most frequent insurer in MA (52%).

Healthcare resource utilization is illustrated in Figure 1. More than 95% of adolescents with CHDs had an outpatient encounter (Table 3). Inpatient encounters were more frequent in severe CHDs as compared to nonsevere. More diagnostic imaging studies were done in subjects with severe CHDs as compared to nonsevere CHDs in MA. Additionally, the percentages of cases with at least 1 cardiac procedure/surgery and vascular procedure were significantly higher in those with severe CHDs as compared to nonsevere CHDs for all 3 sites.

Common co-morbidities were grouped into relevant organ systems by ICD-9-CM diagnostic codes using CCS.¹² The frequency of co-morbidities by CHD severity for each site are shown in Supplemental Table 5. In common co-morbidities, other cardiovascular disorders (27% to 69%) and conduction and rhythm disorders (9% to 46%) were the most frequent cardiac co-morbidities seen in adolescents with any CHD at all 3 sites (Figure 2). Heart failure varied from 2% to 15%. Atherosclerotic cardiovascular risk factors such as hypertension (3% to 18%), hyperlipidemia (0.4% to 11%), and diabetes (1% to 13%) were also noted in this population. The most frequent noncardiac co-

morbidities at all 3 sites included respiratory/pulmonary (16% to 34%), infectious disease (17% to 22%), injury/trauma (9% to 22%), birth defects (non-CHDs) (12% to 23%), gastrointestinal (10% to 28%), musculoskeletal (10% to 32%), and mental health (9% to 30%). Cases with severe CHD had significantly more co-morbidities than nonsevere CHD in the following: other cardiovascular, heart failure, conduction and rhythm disorders, respiratory/pulmonary, gastrointestinal, infectious disease, and birth defects (non-CHDs) (Supplemental Table 5).

Discussion

Our study used a novel approach of uniform CHD definition across multiple administrative data sources in 3 sites for the first population-based CHD surveillance in adolescents in the United States. With US Census Bureau intercensal estimates from July 1, 2010 and this study, we estimate approximately 150,000 to 650,000 adolescents are living with CHDs in the United States.¹³ Approximately 35,000 to 115,000 adolescents have severe CHDs. The estimated adolescent prevalence was approximately 4 to 17 per 1,000 for any CHDs and 0.9 to 3 per 1,000 for severe CHDs across 3 US sites in 2010 with a healthcare encounter. Marelli et al estimated a prevalence in children of 13 per 1,000 for all CHDs and 1.76 per 1,000 for severe CHDs in 2010 in Quebec, Canada.¹¹ Extrapolating this data to the United States, prevalence was estimated to be 12.62 and 10.18 per 1,000 for all CHDs in adolescent males and females, respectively; estimates for severe CHDs were 1.87 and 1.85 per 1,000, respectively.⁴ This corresponds well to our estimates.

The observed prevalence at each site represents the adolescents who had a healthcare encounter during the surveillance period and may be an over or underestimate of the true adolescent prevalence. Important differences existed in the sites which may relate to the quality and comprehensiveness of the data used to create the estimates, such as the characteristics of the adolescent population within the

Table 2

Demographics of Adolescents with CHDs by CHD severity at the Emory, Massachusetts, and New York sites as of January 1, 2010

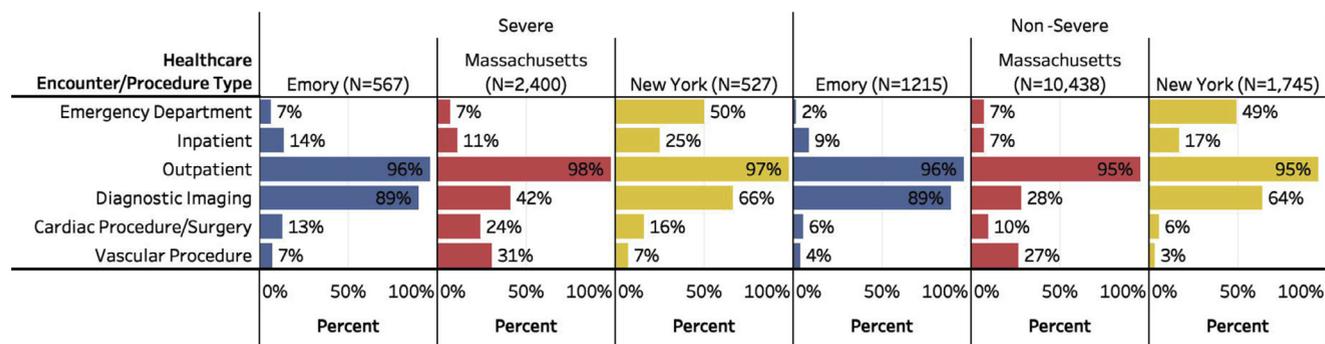
| Demographic characteristic | Emory | | | p value* |
|---|----------------------------|--------------------------------|------------------------------|----------|
| | Severity of CHD diagnosis | | | |
| | Severe (total = 567) | Nonsevere CHD (Total = 1215) | Isolated 745.5 (total = 244) | |
| Age (on January 1st, 2010) | | | | 0.40 |
| 11 - 13 | 160 (28.2%) | 346 (28.5%) | 61 (25.0%) | |
| 14 - 16 | 245 (43.2%) | 488 (40.2%) | 112 (45.9%) | |
| 17 - 19 | 162 (28.6%) | 381 (31.3%) | 71 (29.1%) | |
| Sex | | | | 0.02 |
| Male | 296 (52.2%) | 692 (56.9%) | 117 (48.0%) | |
| Female | 271 (47.8%) | 523 (43.1%) | 127 (52.1%) | |
| Missing | — | — | — | |
| Insurance status (not mutually exclusive) | | | | |
| Self-pay/uninsured | 0 (0.0%) | 5 (0.4%) | 0 (0.0%) | |
| Private/commercial | 345 (60.9%) | 768 (63.2%) | 151 (61.9%) | |
| Medicaid | 244 (43.0%) | 433 (35.6%) | 92 (37.7%) | |
| Other (includes medicare/SSI) | 4 (0.7%) | 6 (0.5%) | 1 (0.4%) | |
| Unknown/unavailable | 22 (3.9%) | 34 (2.8%) | 5 (2.1%) | |
| | Massachusetts [#] | | | |
| Demographic characteristic | Severity of CHD diagnosis | | | p value* |
| | Severe (total = 2400) | Non-severe CHD (total = 10438) | Isolated 745.5 (total = 791) | |
| Age (on January 1st, 2010) | | | | < 0.001 |
| 11 - 13 | 787 (32.8%) | 3252 (31.2%) | 214 (27.1%) | |
| 14 - 16 | 883 (36.8%) | 4045 (38.8%) | 282 (35.7%) | |
| 17 - 19 | 730 (30.4%) | 3141 (30.1%) | 295 (37.3%) | |
| Sex | | | | < 0.001 |
| Male | 1188 (49.5%) | 5279 (50.6%) | 343 (43.4%) | |
| Female | 1212 (50.5%) | 5159 (49.4%) | 448 (56.6%) | |
| Missing | — | — | — | |
| Insurance status (not mutually exclusive) | | | | |
| Self-pay/uninsured | 0 (0.0%) | 0 (0.0%) | 0 (0.0%) | |
| Private/commercial | 592 (24.7%) | 2344 (22.5%) | 124 (15.7%) | |
| Medicaid | 1489 (62.0%) | 5107 (48.9%) | 550 (69.5%) | |
| Other (includes medicare/SSI) | * | 26 (0.3%) | * | |
| Unknown/unavailable | 333 (13.9%) | 2998 (28.7%) | 116 (14.7%) | |
| | New York | | | |
| Demographic characteristic | Severity of CHD diagnosis | | | p value* |
| | Severe (total = 527) | Non-severe CHD (total = 1745) | Isolated 745.5 (total = 265) | |
| Age (on January 1st, 2010) | | | | 0.02 |
| 11 - 13 | 162 (30.7%) | 416 (23.8%) | 70 (26.4%) | |
| 14 - 16 | 188 (35.7%) | 693 (39.7%) | 94 (35.5%) | |
| 17 - 19 | 177 (33.6%) | 636 (36.5%) | 101 (38.1%) | |
| Sex | | | | < 0.001 |
| Male | 298 (59.2%) | 849 (55.0%) | 98 (40.8%) | |
| Female | 205 (40.8%) | 696 (45.0%) | 142 (59.2%) | |
| Missing | 24 | 200 | 25 | |
| Insurance status (not mutually exclusive) | | | | |
| Self-pay/uninsured | 52 (9.9%) | 175 (10.0%) | 42 (15.9%) | |
| Private/commercial | 307 (58.3%) | 1071 (61.4%) | 171 (64.5%) | |
| Medicaid | 85 (16.1%) | 234 (13.4%) | 50 (18.9%) | |
| Other (includes medicare/SSI) | 22 (4.2%) | 47 (2.7%) | 6 (2.3%) | |
| Unknown/unavailable | 128 (24.3%) | 431 (24.7%) | 47 (17.7%) | |

[#] Massachusetts cannot report frequencies under 12.

* Chi-square p value compares age group and the severity of the CHD diagnosis.

surveillance areas (ie, insurance coverage, referral patterns, and access to healthcare), and variations in coding practices in sites. MA had the highest prevalence estimate at 17 per 1,000 for all CHDs. The prevalence estimate for MA was

obtained using a comprehensive all claims database in a state with near universal healthcare, which may account for the higher estimate. This prevalence is similar to a prevalence for any CHDs of 16.7 per 1,000 children and



*Percent of cases with at least one healthcare encounter/procedure

Figure 1. Healthcare utilization in adolescents with CHDs by severity and site (excluding isolated 745.5).

adolescents in a study from South Carolina using data from both Medicaid and the State Health Plan that cover 99% of children and adolescents in the state.¹⁴ In contrast, the prevalence estimates from EU and NY were based on a combination of clinical, hospital administrative, and insurance databases. NY only had access to encounters of Medicaid recipients through hospital and cardiology clinic administrative databases. Therefore, other healthcare encounters in the Medicaid population were missed (eg, a primary care visit). In addition, neither EU nor NY geographic sites have universal healthcare coverage. These

factors may have resulted in an underestimation of the true prevalence of CHDs in EU and NY.

The proportion of insurance type also varied by site. Private/commercial insurances were more frequent in EU and NY while Medicaid was the most common in MA. The proportion of CHD cases insured by Medicaid in NY appears small in comparison to the other sites and may reflect differences in the administrative databases used by this site. The healthcare needs of uninsured patients may be underestimated in part due to financial barriers which limited healthcare utilization and therefore ascertainment in this study.

Table 3

Selected encounter type by CHD severity at Emory (2008-2010), Massachusetts (2009-2010), and New York (2008-2010) sites

| Encounter type | Emory | | | Massachusetts | | | New York | | |
|---|-------------|---------------|----------|---------------|---------------|----------|-------------|---------------|---------|
| | Severe | Nonsevere CHD | p value | Severe | Nonsevere CHD | p value | Severe | Nonsevere CHD | p value |
| Total cases | 567 | 1215 | | 2400 | 10438 | | 527 | 1745 | |
| Emergency department visits | | | | | | | | | |
| Cases with at least one visit in ED | 37 (6.5%) | 23 (1.9%) | < 0.001 | 158 (6.6%) | 678 (6.5%) | 0.88 | 261 (49.5%) | 851 (48.8%) | 0.76 |
| Average number of ED visits per person with at least one visit | 1.7 | 1.7 | 0.99* | 1.6 | 1.7 | 0.59* | 2.6 | 2.7 | 0.47* |
| Inpatient | | | | | | | | | |
| Cases with at least one day of inpatient care | 80 (14.1%) | 108 (8.9%) | < 0.001 | 262 (10.9%) | 755 (7.2%) | < 0.001 | 131 (24.9%) | 290 (16.6%) | <0.001 |
| Average number of days in inpatient care with at least one day | 7.3 | 10.4 | 0.31* | 4.8 | 3.4 | 0.04* | 12.3 | 11.8 | 0.83* |
| Outpatient (clinic and non-ED outpatient visits) | | | | | | | | | |
| Cases with at least one day of outpatient care | 543 (95.8%) | 1162 (95.6%) | 0.84 | 2342 (97.6%) | 9948 (95.3%) | < 0.001 | 513 (97.3%) | 1654 (94.8%) | 0.01 |
| Average number of days in outpatient care with at least one day | 14.2 | 11.5 | 0.21* | 4.8 | 3.7 | < 0.001 | 4.0 | 2.1 | <0.001* |
| Cardiac imaging | | | | | | | | | |
| Cases with at least one procedure | 507 (89.4%) | 1076 (88.6%) | 0.56 | 997 (41.5%) | 2969 (28.4%) | < 0.001 | 346 (65.7%) | 1115 (63.9%) | 0.46 |
| Average # of procedures among those who had at least one | 10.4 | 6.6 | < 0.001* | 4.8 | 2.9 | < 0.001* | 8.1 | 4.9 | <0.001* |
| Diagnostic cardiac procedures/surgeries | | | | | | | | | |
| Cases with at least one procedure | 71 (12.5%) | 69 (5.7%) | < 0.001 | 575 (24.0%) | 1058 (10.1%) | < 0.001 | 86 (16.3%) | 106 (6.1%) | <0.001 |
| Average # of procedures among those who had at least one | 5.2 | 5.1 | 0.92* | 2.8 | 2.0 | < 0.001* | 7.1 | 5.7 | <0.001* |
| Vascular procedures | | | | | | | | | |
| Cases with at least one procedure | 40 (7.1%) | 54 (4.4%) | 0.02 | 744 (31.0%) | 2795 (26.8%) | <.001 | 37 (7.0%) | 50 (2.9%) | <0.001 |
| Average # of procedures among those who had at least one | 2.1 | 2.6 | 0.58* | 2.6 | 2.0 | <.001* | 3.8 | 2.3 | 0.05* |

*T test was used to calculate p value.

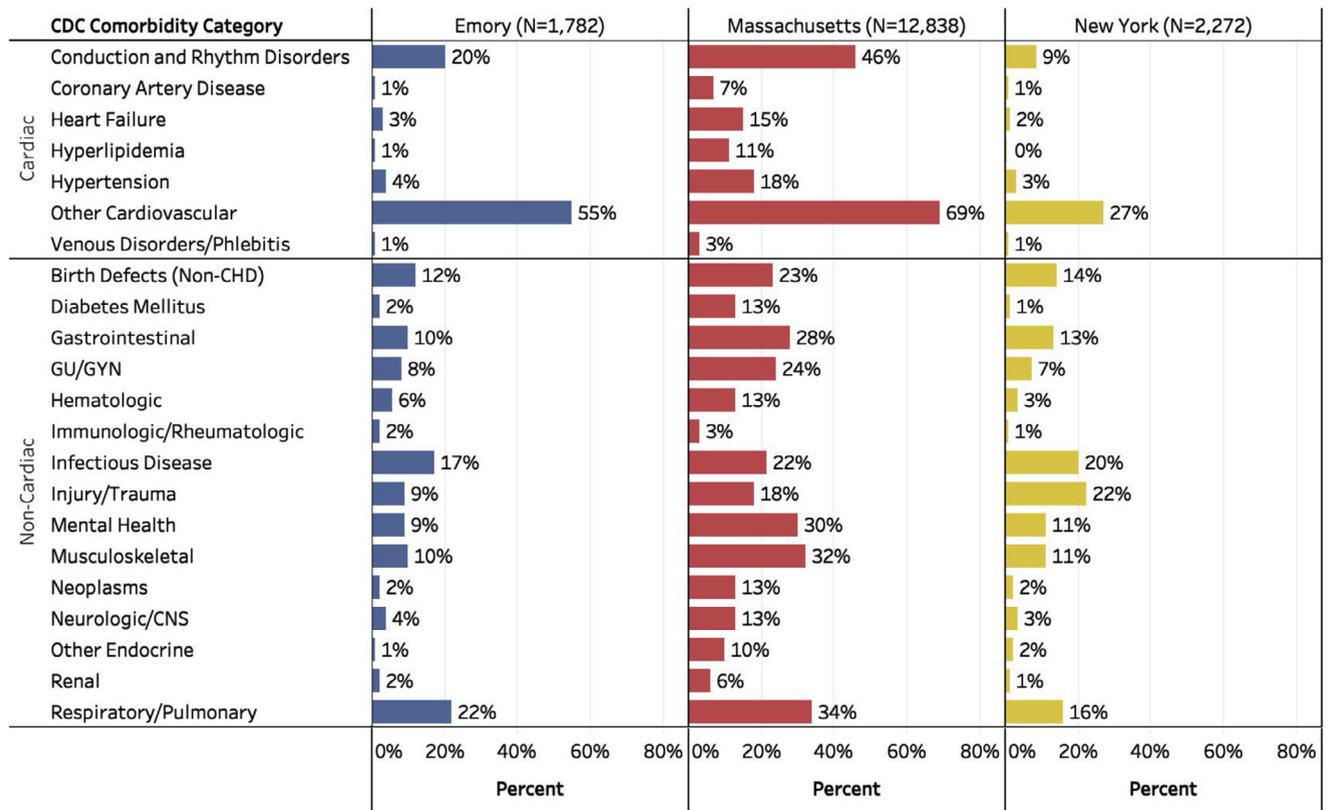


Figure 2. Comorbidities in adolescents with CHDs (excluding isolated 745.5).

Few studies have examined healthcare utilization in the adolescent population.¹⁵ For all 3 sites, adolescents with severe CHDs had significantly higher resource utilization including inpatient hospitalizations, cardiac and vascular procedures/surgeries compared to those with nonsevere CHDs. We found that 5% to 25% of adolescents with severe CHDs were hospitalized during the surveillance period. Additionally, 13% to 24% of severe CHD and 6% to 10% of nonsevere CHD cases underwent a cardiac procedure/surgery, and cardiac imaging use was high in all CHDs. This is one of the first studies to present the high imaging and procedure use in an adolescent CHD population, indicating their ongoing significant resource utilization.

Co-morbidities add to the complexity of care and contribute to the increased resource utilization in this population. There is an increasing prevalence of co-morbidities as patients' age.^{16,17} Hypertension, diabetes, obesity, smoking, peripheral arterial disease, and chronic kidney disease increased significantly in patients with CHDs from 2003 to 2012 in an analysis of the Nationwide Inpatient Sample.¹⁸ Even in this young adolescent population, there was evidence of hypertension, diabetes, hyperlipidemia, and even atherosclerotic coronary artery disease. The prevalence of hypertension and diabetes appear to be higher than that of the general adolescent population (0.8% and 3%, respectively).^{19,20}

Noncardiac co-morbidities may play an increasingly important role in the long-term outcome of CHD patients.²¹ Our study further contributes to these findings with associated respiratory/pulmonary, infectious disease, gastrointestinal, and musculoskeletal co-morbidities.

Additionally, these co-morbidities are not limited to the physical domain but also expand to mental health co-morbidities, which were identified in 9% to 30% of adolescent CHD patients in this cohort. These results are consistent with previous literature that has suggested approximately one third of adults with CHD have mood or anxiety disorders.²²

Limitations of this study include the administrative data which can be prone to misclassification.²³ Our ICD-9-CM diagnosis code classification scheme for CHDs was not validated in the administrative data at the 3 sites. We primarily followed the categorization of severe versus nonsevere as described in Marelli et al.⁹ However, there were some changes made to attempt more accurate classification, including the addition of the following ICD-9-CM codes in the severe CHDs group: double outlet right ventricle (747.11), pulmonary valve atresia or absence (746.01), primum ASD (746.1), and total anomalous pulmonary venous return (747.41).

Our estimates are for the prevalence of patients with CHDs who had a healthcare encounter during the years 2008 (2009 for MA) – 2010. Therefore, individuals with CHDs who did not obtain care in the surveillance period were not included. Healthcare encounters that may have occurred outside of each site's geographic area would also have been missed. Additionally, the administrative record may not have documented a CHD diagnosis code during an encounter (eg, if a patient's CHD was believed to be fully repaired). We expect that records of patients with severe CHDs are more likely to include a CHD code, whereas those with nonsevere CHDs are less likely to be coded if

the CHD does not impact their current care. Thus, we may have underestimated the nonsevere CHD population more than the severe CHD population. It is possible that some patients' records were not deduplicated despite best efforts to identify individual patients only once. Also, patients with CHD diagnostic codes may have been miscoded and actually have acquired heart disease or no heart disease at all. The estimated prevalence of adolescents with CHDs varied by CHD severity and geographical site and deserves further study in other states. The use of uniform CHD definitions and variable selection allowed a comparison of surveillance data across 3 sites. The high resource utilization and co-morbidities seen in the adolescents with CHDs illustrate the ongoing significant burden of disease and highlight the importance of ensuring continuity of care for this vulnerable population.

Disclosures

The authors have no conflicts of interest to declare.

Supplementary materials

Supplementary material associated with this article can be found in the online version at <https://doi.org/10.1016/j.amjcard.2019.03.044>.

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