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Comorbidities and Healthcare Utilization Among Young Adults With Congenital Heart Defects by Down Syndrome Status—Congenital Heart Survey to Recognize Outcomes, Needs, and wellbeinG, 2016–2019

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Abstract

Background: Almost half of individuals born with Down syndrome (DS) have congenital heart defects (CHDs). Yet, little is known about the health and healthcare needs of adults with CHDs and DS. Therefore, we examined comorbidities and healthcare utilization of this population.

Methods: Data were from the 2016–2019 Congenital Heart Survey to Recognize Outcomes, Needs, and well-beinG (CH STRONG), a survey of 19–38-year-olds with CHDs identified through birth defects registries in Arkansas, Arizona, and Atlanta. Outcome estimates were standardized to the CH STRONG eligible population. Multivariable Poisson regression generated adjusted prevalence ratios (aPRs) for associations between DS and each outcome, adjusting for covariates, including CHD severity.

Results: Among 1500 respondents, 9.1% had DS. Compared to those without DS, respondents with DS were more commonly male (55.5% vs. 45.0%), < 25 years old (51.8% vs. 42.7%), non-Hispanic White (72.3% vs. 69.3%), and publicly insured (77.4% vs. 22.8%; all $p < 0.05$). Of adults with CHDs and DS, 5.5% had cardiac comorbidities, 19.3% had emergency room (ER)

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Author Contributions

Vanessa I. Villamil: conceptualization, formal analysis, visualization, writing-original draft. **Karrie F. Downing:** validation, writing-review and editing. **Matthew E. Oster:** conceptualization, writing-review and editing. **Jennifer G. Andrews:** conceptualization, writing-review and editing. **Maureen K. Galindo:** writing-review and editing. **Jenil Patel:** writing-review and editing. **Scott E. Klewer:** writing-review and editing. **Wendy N. Nembhard:** writing-review and editing. **Sherry L. Farr:** conceptualization, supervision, manuscript preparation.

Supporting Information

Additional supporting information can be found online in the Supporting Information section.

visits, 6.2% had hospital admissions, and 1.2% had cost-related delays in care in the last year; 0.1 to 0.6 times lower than adults with CHDs without DS. Additionally, 26.7% had non-cardiac comorbidities (aPR = 1.25 [0.92–1.72]), most commonly sleep apnea (13.7% vs. 3.2%, aPR = 3.67 [2.02–6.67]). Receipt of cardiology care in the last 2 years was similarly low (52.7% vs. 44.7%).

Conclusions: Adults with CHDs and DS comprise a substantial percentage of adults with CHDs and have unique health and healthcare needs. Half of adults with CHDs and DS are not receiving recommended routine cardiac care.

Keywords

cardiology; congenital heart defects; Down syndrome; healthcare utilization

1 | Introduction

Congenital heart defects (CHDs) are common among infants born with Down syndrome (DS), with almost half of all infants with DS also having CHDs (Santoro and Steffensen 2021). Survival to adulthood has increased for individuals with CHDs (Moons et al. 2010; Downing et al. 2023) and individuals with DS (Centers for Disease Control and Prevention 2001; Englund et al. 2013) over the years, resulting in an increasing population of adults with CHDs and DS who may have unique health challenges and healthcare needs. Some evidence exists that adults with DS in general have lower rates of arterial hypertension (Morrison et al. 1996), myocardial infarction, and heart disease (Alexander et al. 2016), but higher rates of stroke (Sobey et al. 2015), pulmonary hypertension, diabetes (Alexander et al. 2016), and sleep apnea (Maris et al. 2016; Capone et al. 2018). The few existing studies assessing these outcomes in individuals with DS and CHDs have been based on children (Frid et al. 2002; Dawson et al. 2014) or hospitalizations (Baraona et al. 2013). Clinical guidelines exist (Tsou et al. 2020) for individuals with DS but are based mainly on expert opinion with limited scientific evidence. The guidelines contain only one weak recommendation specific to adults with DS and CHDs: periodic cardiac evaluation due to the increased risk of stroke. It is not well understood how commonly adults with CHDs and DS receive appropriate cardiac and non-cardiac healthcare and what comorbidities they most frequently experience throughout their lifespan.

Therefore, the objective of this study was to assess the prevalence of comorbidities and healthcare utilization among adults with CHDs and DS using data from the Congenital Heart Survey to Recognize Outcomes, Needs, and well-beinG (CH STRONG). This information may be used to inform clinical care guidelines and optimize the management of comorbidities for adults with CHDs and DS.

2 | Methods

2.1 | Data Source

Data for this analysis were from the 2016–2019 CH STRONG, a cross-sectional survey of adults 19–38 years of age, born between 1980 and 1997, and diagnosed with CHDs in early childhood, or their proxy (e.g., parent or caregiver). CHD status was based on the Ninth Revision of the International Classification of Disease with the British Pediatric

Association Classification of Diseases extension (ICD-9-CM/BPA) codes 745.0–747.X, except non-specific codes (Table S1). Eligible adults were identified through birth defects registries with active case ascertainment in Arkansas, Arizona, and metropolitan Atlanta, Georgia, tracked and traced for current contact information, and sent a survey on health and healthcare utilization. Individuals who were incarcerated or deceased at the time of survey recruitment or unable to complete the survey in English or Spanish were ineligible to participate. Among the 9312 CH STRONG-eligible participants, 6943 had an available address and were sent a survey, of whom 1656 (23.9%) responded to the survey (Farr et al. 2020). CH STRONG was funded by the CDC and approved by the Institutional Review Boards at the CDC and the University of Arkansas for Medical Sciences. The University of Arizona deferred to the CDC Institutional Review Board. A more detailed description of the CH STRONG methods has been published (Farr et al. 2020).

2.2 | Measures

DS status was based on ICD-9-CM/BPA codes between 758,000 and 758,090 in the birth defects registry. Within the CH STRONG survey, participants were asked if they had ever received a clinician diagnosis of four cardiac conditions (congestive heart failure, hypertension, myocardial infarction, and stroke) and five non-cardiac conditions (asthma, cancer, diabetes, rheumatologic disease, obstructive sleep apnea). Participants were considered to have a cardiac and/or non-cardiac comorbidity (dichotomized as 0 and ≥ 1) if they answered yes to any of the respective conditions. Healthcare utilization in the past 12 months was assessed through survey questions on the number of visits with a healthcare provider (0, 1, 2–7, and ≥ 8), number of emergency room (ER) visits (dichotomized as 0 and ≥ 1), number of hospital admissions (dichotomized as 0 and ≥ 1), number of cardiac-related hospital admissions (dichotomized as 0 and ≥ 1), and delays in care due to cost (Yes/No). Two variables were created to examine the receipt of cardiology care. The last time seeing a cardiologist (dichotomized as 0–2 years and ≥ 3 years or never) was based on self-report. Similar to a previous CH STRONG analysis (Andrews et al. 2023), being lost to follow-up for cardiology care (LTF) was determined based on an individual's self-reported last time seeing a cardiologist and the 2018 AHA/ACC guidelines for CHDs¹⁷ based on the individual's most severe diagnosis.

2.3 | Covariates

Covariates assessed included CHD severity, age group, and sex from the birth defects registry, and race/ethnicity and health insurance from the survey. CHD severity was categorized using a previously published algorithm (Glidewell et al. 2018) based on anatomy and dichotomized as severe and non-severe. Age was estimated using the mid-point of the year of birth and the date the survey was received. Race/ethnicity was categorized as Hispanic, non-Hispanic Black, non-Hispanic White, and non-Hispanic Other. Health insurance was categorized as any public, only private, and other/none.

2.4 | Statistical Analysis

Among the 1656 CH STRONG participants who returned surveys, a complete-case analysis was conducted, including individuals with information on all variables of interest. Characteristics of included and excluded individuals were compared using chi-square tests to

assess for potential non-response bias related to non-response on specific survey questions used in this analysis (Table S2).

Prevalence estimates for comorbidity and healthcare utilization outcomes by DS status were standardized to the 9312 individuals in the CH STRONG eligible population by site, sex, maternal race and ethnicity, birth year, and CHD severity to provide population-based estimates and reduce nonresponse bias related to not returning a CH STRONG survey. Prevalence of demographic and healthcare characteristics by DS status was summarized.

Bivariate and multivariable Poisson regression models generated crude prevalence ratios (cPRs) and adjusted prevalence ratios (aPRs) with 95% confidence intervals (CIs) for associations between DS and each of the nine comorbidity and healthcare utilization outcomes. Models were adjusted for CHD severity, age group, sex, race/ethnicity, and health insurance. Additionally, to further assess the relationship between DS and the outcomes of interest, 10 sensitivity analyses were conducted, limiting the analytic sample to those: (1) < 25 years old, (2) 25–30 years old, (3) > 30 years old, (4) with health insurance, (5) with a shunt CHD, (6) with a primary diagnosis of ASD, VSD, or AV canal/endocardial cushion defect, (7) with any diagnosis of ASD, VSD, or AV canal/endocardial cushion defect, (8) with severe CHD, also (9) adjusting for proxy report, and (10) imputing missing data to assess whether non-response on survey questions impacted the results.

All analyses were conducted in SAS-callable SUDAAN version 11. Significance was set at $p < 0.05$ for all analyses. Multiple imputations were conducted using the SAS Proc MI procedure, with 25 imputations, using all characteristics included in the analysis. The imputed data were pooled and used for adjusted analyses; results were combined using the SAS Proc MIANALYZE procedure.

3 | Results

Of the 1656 CH STRONG respondents, 50 (3.0%) were missing demographic and healthcare characteristics, and 106 (6.4%) were missing data on comorbidities or healthcare utilization outcomes (Table S2). Therefore, the final analytic sample included 1500 (90.6%) respondents with complete data (representing 17.8% of the CH STRONG eligible population), of whom 9.1% had a DS diagnosis.

Over a third (38.7%) of adults with CHDs and DS had severe CHDs, which did not differ compared to those without DS (33.8%) (Table 1). The three most common primary CHDs among respondents with DS were atrioventricular (AV) canal/endocardial cushion defect (32.8%), atrial septal defect (ASD) (26.3%), and ventricular septal defect (VSD) (21.2%), while among those without DS were VSD (27.6%), multiple primary CHDs (13.4%), and ASD (9.0%). Comparing respondents with DS to those without, the distributions of non-severe CHD type (shunt: 56.2% vs. 41.6%), age (< 25 years old: 51.8% vs. 42.7%), sex (male: 55.5% vs. 45.0%), race/ethnicity (Hispanic: 16.1% vs. 10.1%), and insurance (any public health insurance: 77.4% vs. 22.8%) differed ($p < 0.05$ for all). Additionally, respondents with DS were more likely to have their survey completed by a proxy (91.2% vs. 13.2%, $p < 0.0001$).

Among adults with CHDs, 5.5% of those with DS had one or more cardiac comorbidities compared to 14.2% of those without DS (aPR = 0.27 [0.13–0.55], Figure 1, Table S3), with the most notable difference in reported hypertension, which was lower for adults with DS. Prevalence of having one or more non-cardiac comorbidities was 26.7% in adults with CHDs and DS, similar to that among adults without DS (21.2%). However, there was a 3.7 times increased prevalence of sleep apnea in adults with DS (13.7% vs. 3.2%). Among adults with DS, 22.8% had one visit with a healthcare provider in the past 12 months (aPR = 1.48 [1.11–1.97]), and 64.2% had between 2 and 7 visits (aPR = 1.16 [1.06–1.27]). Approximately one in five adults with DS reported ER visits in the past year (aPR = 0.57 [0.42–0.78]), 6.2% reported hospital admissions overall (aPR = 0.56 [0.33–0.95]), 0.7% reported cardiac-related hospital admissions (aPR = 0.16 [0.04–0.65]), and 1.2% reported delays in care due to cost during the 12 months before the survey (aPR = 0.14 [0.04–0.42]) (Figure 2, Table S3). No other associations were found.

Prevalence estimates were similar, and associations remained when analyses were limited to individuals < 25 years old, 25–30 years old, > 30 years old, with health insurance, with a shunt CHD, with a primary diagnosis of ASD, VSD, or AV canal/endocardial cushion defect, with any diagnosis of ASD, VSD, or AV canal/endocardial cushion defect, with severe CHD, after adjusting for proxy report, and when imputing to account for missing data (Tables S4–S13). However, for most sensitivity analyses, CIs were wider due to lower sample sizes and reduced power, and some aPRs could not be calculated due to insufficient data. Imputing data did not change the estimated aPRs by more than eight-hundredths of a decimal.

4 | Discussion

Our findings indicate that DS is common among young adults with CHDs—over 9% of young adults with CHDs in the general population are also diagnosed with DS. Previous studies estimated the prevalence to be between 5% and 8%; however, these studies were among children requiring cardiac operations or individuals of all ages receiving CHD-related healthcare (Reller and Morris 1998; Evans et al. 2014; Glidewell et al. 2021). This analysis is one of the first to assess comorbidities and healthcare utilization among adults with CHDs and DS in the U.S. We found that one in four adults with CHDs and DS had a non-cardiac comorbidity, similar to adults with CHDs without DS, and about one in 20 had cardiac comorbidities, an estimate 75% lower than that of adults with CHDs without DS. Adults with CHDs and DS were less likely to report ER visits and hospital admissions in the last 12 months. They also reported fewer delays in care due to cost and more visits with a healthcare provider in the last 12 months. These findings persisted when stratified by CHD severity, CHD type, age group, health insurance status, when adjusted for proxy report, and when multiple imputation was used to account for non-response. Overall, these results show that adults with CHDs and DS comprise a substantial percentage of adults with CHDs and have a unique demographic and health profile, even when compared to adults with the same CHD types.

Some evidence suggests that cardiac and non-cardiac outcomes differ among the general population of individuals with and without DS; however, most studies have not stratified

results by CHD status. Adults with DS are more likely than those without DS to experience low blood pressure (Morrison et al. 1996); and, among young adults with CHD, we similarly found a lower prevalence of hypertension. Studies have also shown that rates of myocardial infarction and heart disease are the same or lower in individuals with DS compared to those without (Tsou et al. 2020); similar to our findings among individuals with CHD. Strokes may occur more often and at younger ages in individuals with DS; however, we found no difference by DS status in the low prevalence of stroke in young adults with CHD. A previous study of hospitalized patients (Sobey et al. 2015) found higher prevalence of diabetes and sleep apnea among adults with DS, compared to those without DS; similarly, our findings in adults with CHD showed sleep apnea was over three times higher in adults with DS than those without, however, the prevalence of diabetes did not differ statistically.

Our results also show that those with DS have better healthcare access, and insurance status may partly explain this finding. A previous study assessing Medicaid enrollment and service use among adults with DS found that, compared to adults without developmental disabilities, adults with DS were more consistently enrolled in Medicaid (Rubenstein et al. 2023). Similarly, in our study, adults with CHDs and DS were more likely to have some form of public health insurance compared to adults with CHDs without DS and were less likely to report delaying care due to cost. Additionally, a higher proportion of adults with CHDs and DS in CH STRONG reported multiple visits with a healthcare provider in the past year, which may indicate better access to care, compared to adults with CHDs without DS. Little information exists on the receipt of cardiology care among adults with DS and CHDs. A study based on Medicaid data found that over 90% of adolescents and adults with CHDs and DS had an echocardiogram in a 5-year period between 2006 and 2010 (Jensen et al. 2021). Our study found that only half of the adults with CHDs and DS had seen a cardiologist in the last 2 years and over a third were LTF for cardiology care based on their specific CHDs, similar to adults with CHDs without DS. These results suggest that a large percentage of individuals with CHDs, including those with DS, are not receiving the recommended routine cardiac evaluation to improve their cardiac health and decrease their risk of stroke (Tsou et al. 2020; Stout et al. 2019).

Compared to U.S. adults with CHDs without DS, our findings suggest that adults with CHDs and DS had lower rates of hospitalization—about half that of adults with CHD without DS. Our findings add to those of an older U.S. study (Baraona et al. 2013) of admissions for adults with CHDs occurring from 1998 to 2009, which found that hospitalizations of adults with CHDs and DS represented a decreasing proportion of admissions, were less likely to include cardiac procedures or surgery, and had lower hospital charges compared to adults with CHDs without DS.

Previous studies have also noted similar or better health-related outcomes, such as survival, when comparing individuals with CHDs and DS to those with CHDs without DS. Among individuals with CHD born 1980–1997, the underlying population for CH STRONG, 85.1% of infants born with CHD and DS survived to 25 years of age, which was similar to the 83.2% of infants born with CHD and no genetic syndromes (Downing et al. 2024). A previous study assessing in-hospital death among children undergoing surgery for CHDs found that children with DS were more likely to survive to discharge than children without

DS, though there were specific procedures (i.e., placement of a systemic to pulmonary shunt, the bidirectional Glenn shunt procedure) that resulted in higher mortality for children with DS (Evans et al. 2014). Additionally, among a cohort of individuals undergoing congenital heart surgery identified in a national clinical database, those with DS were not at increased risk of post-surgical mortality, although they had a prolonged length of inpatient stay associated with respiratory infection complications (Fudge Jr et al. 2010). These studies have speculated that similar or better survival outcomes may be associated with subtle differences in cardiac structure between individuals with DS and those without (Evans et al. 2014; Fudge Jr et al. 2010). In our study, limiting the analysis to individuals with ASD, VSD, and AV canal/endocardial cushion defect did not impact the results, yet differences in CHDs between groups may still exist. Another study examining longer-term survival of children with CHDs and DS after congenital heart surgery found that DS was a predictor for 30-year all-cause mortality, but not a predictor of CHD-related mortality, possibly due to other co-occurring conditions (Peterson et al. 2021).

It is possible that diagnostic overshadowing (the misattribution of signs and symptoms of a comorbid condition to an underlying disability) or differences in self-identification of symptoms may contribute to lower rates of reported comorbidities in adults with CHD and DS. However, it is unknown how often diagnostic overshadowing may occur for people with DS.

4.1 | Study Limitations

Self-report is both a strength and a limitation of this study, as it may lead to inaccurate information being reported; however, it allows for the inclusion of a sizeable sample of the population with CHDs who may often be missed in studies relying on medical record data due to falling out of cardiology care. Additionally, the CH STRONG survey response rate was 23.9%, and 9.4% of survey respondents were excluded from this analysis due to missing data on specific variables. Survey respondents excluded from the analysis differed from included respondents by health insurance coverage and reported delays in care due to cost. However, there were no differences in prevalence of DS among respondents and non-respondents or between excluded and included respondents (Farr et al. 2020). Therefore, non-response may limit the generalizability of prevalence estimates for survey items but likely had no substantial effects on prevalence ratios. Lastly, these results are specific to individuals with CHD born in the three regions included in CH STRONG who survived to adulthood; generalizability to other areas or to children may be limited.

5 | Conclusions

Individuals with DS make up a substantial proportion of young adults with CHDs and have unique health outcomes and patterns of healthcare utilization. In this study among young adults with CHDs and DS, one in 20 had cardiac comorbidities, one in four had non-cardiac comorbidities, and they had low rates of cardiology care. Cardiac comorbidities, ER visits, and hospitalizations for adults with CHDs and DS differed from those with CHDs without DS, possibly as a result of differences in insurance status and access to care. Differences in CHD type among adults with DS may also impact outcomes, although this was not found in

our study. Continued understanding of the needs of people with CHDs and DS may inform clinical guidelines, ensure that their health needs are met throughout their lifespan, and help individuals, their families, and caregivers plan for the future.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

This study was funded by the CDC and approved by the Institutional Review Boards of the CDC and the University of Arkansas for Medical Sciences. The University of Arizona deferred to the CDC Institutional Review Board.

Data Availability Statement

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.

Abbreviations:

APR	Adjusted Prevalence Ratio
BPA	British Pediatric Association
CDC	Centers for Disease Control and Prevention
CHDs	congenital heart defects
CH STRONG	Congenital Heart Survey to Recognize Outcomes, Needs, and well-being
DS	Down syndrome
ICD	International Classification of Disease
LTF	lost to follow-up

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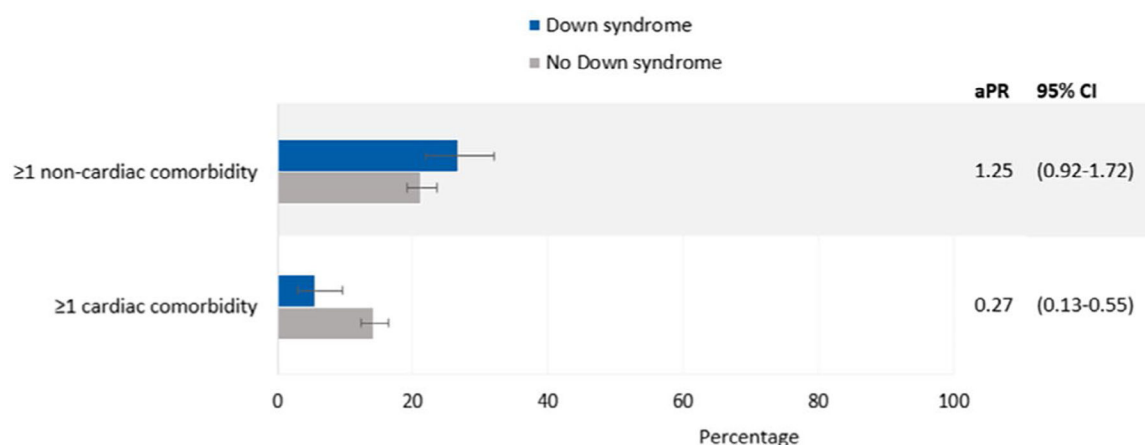


FIGURE 1 |

Comorbidities among adults with congenital heart defects, by Down syndrome status: CH STRONG, 2016–2019. Cardiac comorbidities include congestive heart failure, hypertension, myocardial infarction, and stroke. Non-cardiac comorbidities include asthma, cancer, diabetes, rheumatologic disease, and obstructive sleep apnea. Prevalence percentages were standardized to the 9312 individuals in the CH STRONG eligible population by site, sex, race/ethnicity, age group, and CHD severity to reduce nonresponse bias. Prevalence ratios were adjusted for CHD severity, age group, sex, and race. aPR = adjusted prevalence ratio of comorbidity among people with Down syndrome compared to people without Down syndrome; CH STRONG = Congenital Heart Survey to Recognize Outcomes, Needs, and wellbeinG; CI = confidence interval.

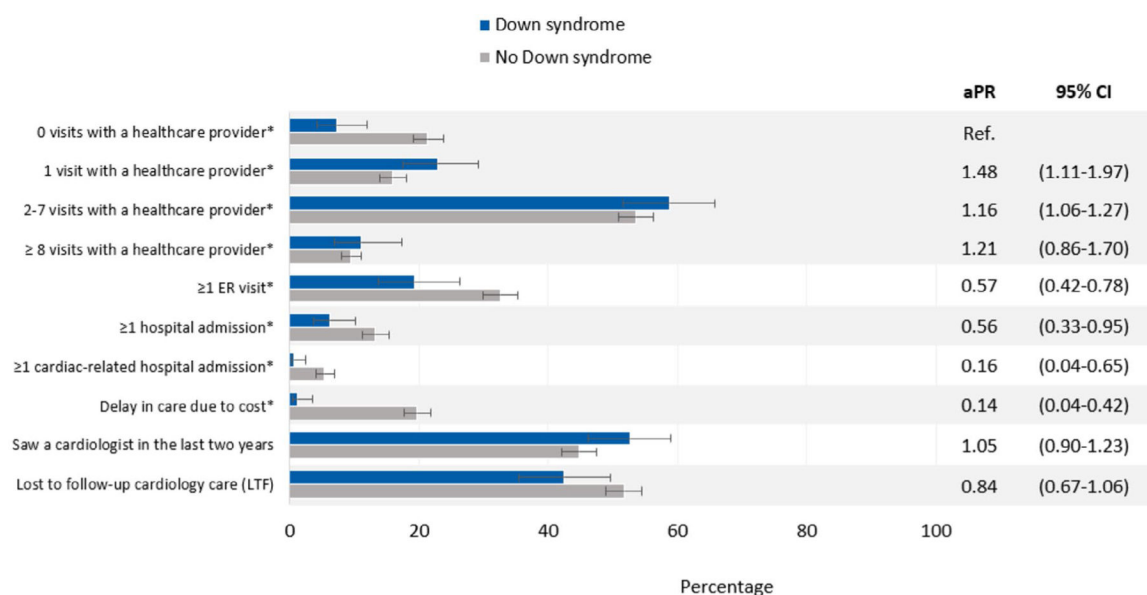


FIGURE 2 |.

Healthcare utilization among adults with congenital heart defects, by Down syndrome status: CH STRONG, 2016–2019. Prevalence percentages were standardized to the 9312 individuals in the CH STRONG eligible population by site, sex, race/ethnicity, age group, and CHD severity to reduce nonresponse bias. Prevalence ratios adjusted for CHD severity, age group, sex, race/ethnicity, and health insurance. *During the past 12 months before the survey. aPR = adjusted prevalence ratio; CH STRONG = Congenital Heart Survey to Recognize Outcomes, Needs, and wellbeinG; CI = confidence interval; ER = emergency room.

Demographic and healthcare characteristics of respondents with congenital heart defects (CHDs) by Down syndrome diagnosis: CH STRONG, 2016–2019.

TABLE 1 |

Characteristic	Down syndrome			No Down syndrome			Chi-square <i>p</i>
	<i>N</i>	%		<i>N</i>	%		
CHD severity							
Severe	53	38.7	461	33.8	0.25		
Non-severe	84	61.3	902	66.2			
Non-severe CHD type							
Valve	0	0.0	161	11.8	< 0.0001		
Shunt	77	56.2	567	41.6			
Shunt + Valve	6	4.4	122	9.0			
Other	1	0.7	52	3.8			
Age group (years) ^a							
< 25	71	51.8	582	42.7	0.02		
25–30	54	39.4	540	39.6			
> 30	12	8.8	241	17.7			
Sex							
Female	61	44.5	750	55.0	0.02		
Male	76	55.5	613	45.0			
Race/Ethnicity							
Hispanic	22	16.1	137	10.1	0.01		
NH Black	13	9.5	183	13.4			
NH White	99	72.3	944	69.3			
Other	3	2.2	99	7.3			
Health insurance							
Any public	106	77.4	311	22.8	< 0.0001		
Only private	17	12.4	735	53.9			
Other/none	14	10.2	317	23.3			
Respondent type ^b							
Proxy report	125	91.2	178	13.2	< 0.0001		

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Characteristic	Down syndrome		No Down syndrome		Chi-square <i>p</i>
	<i>N</i>	%	<i>N</i>	%	
Self-report	12	8.8	1166	86.8	

Note: Severe CHD = International Classification of Disease version 9.0 Clinical Modification Codes (745.0, 745.1, 745.10, 745.11, 745.12, 745.19, 745.2, 745.3, 745.6, 745.60, 745.69, 746.01, 746.1, 746.7, 747.11, 747.41).

^a Ages ranged from 19 to 38.

^b Respondent type had an additional 19 missing responses.