Congenital Heart Disease

Tech Science Press

DOI: 10.32604/CHD.2021.014384

ARTICLE

Hypertension and Heart Failure as Predictors of Mortality in an Adult Congenital Heart Defect Population

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Received: 23 September 2020 Accepted: 05 January 2021

ABSTRACT

Early intervention to prevent premature mortality is vital for adults with congenital heart defects (CHD). Anatomic complexity and comorbid conditions are thought to contribute to CHD mortality. Since hypertension (HTN) and heart failure (HF) are the comorbid conditions among the most prevalent causes of death in the United States, and commonly accompany CHD, it is crucial to evaluate whether they are reliable predictors of mortality for adults with CHD (ACHD) independent of anatomic CHD complexity. A retrospective cross-sectional analysis of ACHD, aged 18–64, with concomitant HTN and/or HF and at least one health care encounter during 2008–2010 were assessed. Of 5,397 ACHD patients (18.3% HTN without HF, 4.4% HF without HTN, 8.3% with both), 3.0% died (n = 163) during the study period. Overall, the sample was 45.1% white, 61.4% female, and 29.0% had a complex CHD. Among those who died, 23.3% had HTN without HF, 17.2% had HF without HTN, and 42.3% had both. Crude analyses revealed that older age, male gender, black race, and having public health insurance were associated with increased mortality during the three-year study period compared to ACHD patients who were younger, female gender, white race, and covered by private health insurance. ACHD patients diagnosed with non-complex CHD lesions (i.e., shunts, valves, or shunts + valves) were at greater risk of dying compared to those with severe complex CHDs. When CHD type was assessed separately, those with valve lesions were more likely to die compared to those with complex CHD lesions. After adjustment for age, gender, race, insurance and CHD complexity, ACHD patients with HF, with or without HTN, were equally likely to die during the study period. However, ACHD patients with HF, without or without HTN, who had valve defects were more likely to die during the three-year study period compared to patients with complex CHDs.

KEYWORDS

Congenital heart defect; risk factors; hypertension; heart failure; mortality

1 Introduction

Congenital heart defects (CHDs) account for ~1% of live births [1] ranging in complexity, often requiring interventions in childhood [2]. In children, increasing anatomic complexity is associated with worse survival [3]. As children with CHD age into adulthood, they face comorbid conditions including

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cardiovascular complications such as hypertension (HTN), heart failure (HF), and myocardial infarction (MI), and have higher mortality with advancing age compared to younger patients with CHD [1,4]. Previous studies of CHD have identified heart failure as an important contributor to mortality and poor long-term outcomes in an adult CHD population [5–9]. HTN is a significant contributor to the development of heart failure [10,11] in the general population, but once heart failure develops, higher blood pressure is associated with better outcomes than low blood pressure [12–15]. The interplay of HTN and HF with race in the setting of pre-existing CHD in an adult population is unknown. Race may be associated with the increased development of HTN and HF in the general population. The Atherosclerotic Risk in Communities (ARIC) study, which examined HF across race and gender in the general population, noted a discrepancy with cardiovascular risk factors disproportionately affecting blacks more than other races [11]. The evaluation of trends in CHD mortality by race and gender has also shown disparities affecting blacks more than other races, as well as males more than females [8,16]. HF is an increasing cause of hospitalization among adults with CHD (ACHD) in the US, and ACHD patients aged 18–65 years account for a disproportionate number of hospitalizations compared to adults without CHD in the same age range [17]. While anatomic complexity of CHD is a significant contributor to mortality in children [3], HTN and HF are common cardiovascular conditions in adults in the US and often also accompany CHD in adults. While development of HF may be associated with anatomic complexity, and either HF or anatomic complexity may be associated with worse outcomes, it is not known if HF predicts mortality regardless of anatomic complexity. For this reason, the evaluation of whether HF with or without HTN are significant predictors of mortality in the context of age, race, gender and CHD anatomic complexity is imperative. Our aim is to evaluate the impact of HTN and HF, and accompanying covariates as predictors of mortality in ACHD in the context of underlying anatomic complexity. Understanding the contribution of these comorbid conditions to mortality in ACHD may help guide long-term management.

2 Materials and Methods

A de-identified analytic file, created from a linked, de-duplicated CHD repository of Georgia residents developed as part of a pilot CHD surveillance project with the Centers for Disease Control and Prevention (CDC) and Emory University, was used. Methods for that project have previously been described [18] and were approved by the Emory University Institutional Review Board (#IRB0000064051). This cross-sectional retrospective study identified 9,394 adult Georgia residents with CHD, ages 18–64 years as of January 1, 2010, who sought medical services at least once from at least one of seven Georgia healthcare facilities (Appendix A) between January 1, 2008 and December 31, 2010. All adults with CHD were diagnosed with at least one of 55 CHD-related ICD-9-CM diagnostic codes (Appendix B) and categorized by CHD anatomy [18] using a five-level classification scheme [19] that was further refined by a multi-site group of congenital heart disease experts, as a component of a larger parent project and adapted for anatomic complexity. The classification scheme included: (1) Severely complex; (2) Shunts; (3) Valves; (4) Shunts plus valves; and (5) Other CHD anomalies (Appendix B) [18]. However, due to lack of specificity and poor diagnostic accuracy determined through chart abstraction and code validation, code 745.5 used for both atrial septal defect (ASD) (a true CHD) and patent foramen ovale (PFO) (a normal condition), 1,491 patients diagnosed with 745.5 in isolation or in combination with either 746.89 or 746.9 (both defined as unspecified congenital anomaly of the heart) were excluded from the analytic cohort, and another 2,506 cases with an ‘Other’ CHD code were also excluded [20] leaving an analytic dataset with N = 5, 397. The main outcome, mortality, confirmed from Georgia Department of Public Health (GA DPH) Death Certificates for years 2008–2010 served as the dependent variable. The primary comorbid conditions, HTN and HF, were derived from the presence of at least one of 40 HTN-related (Appendix C) or one of 16 HF-related ICD-9-CM codes (Appendix D) appearing on at least one healthcare encounter during the study period; number and percentage of ACHD patients with each of

these 40 HTN and 16 HF codes are reported as part of Appendices C and D, respectively. Comorbid groupings of HTN/HF included those having at least one HTN ICD-9-CM code without having any HF ICD-9-CM codes, those having at least one HF ICD-9-CM code without any HTN codes, or those having a combination of both HTN and HF ICD-9-CM codes. We assessed the predictors of age, gender and race because they are established risk factors for CHD. Age was calculated by subtracting the patient's date of birth from 01/01/2008 and was then classified into four age groups approximating age decades as close as possible: 18–29 years, 30–39 years, 40–49 years, and 50–64 years of age. As noted, two of age intervals include patients whose ages fell outside of the age-specific 10 year or decade intervals. As such, we placed the 18 and 19 year olds with the 20–29 year old decade, and patients who were 60–64 years of age were grouped with the 50–59 year old decade. Patients who were 30–39 years of age and 40–49 years of age were retained in age-specific 10 year or decade intervals. Race was classified into the following 4 categories: White, Black, other (i.e., American Indian/Alaskan Native, Asian, Native Hawaiian/other Pacific Islander, and multi-racial), and unknown. Individuals who had a heart transplantation history were not included.

All analyses were conducted using SAS 9.4 (Cary, NC). Frequencies were conducted for all categorical variables and means and standard deviations were computed for each age in its continuous form. For bivariate analysis, crude or unadjusted odds ratios were computed using chi-square tests to analyze the association of HTN/HF comorbid conditions exposures and other comorbidities with mortality. Bivariate logistic regression analyses were performed to assess the association of mortality with the three HTN/HF primary comorbidities separately (HTN without HF, HF without HTN and both HTN and HF) and with each of the other demographic variables including age, gender, race, and insurance. Multiple logistic regression adjusted models were then conducted to determine the likelihood of dying during the three-year surveillance period for CHD patients diagnosed with HTN without HF, HF without HTN, and both HTN and HF with each model separately, controlling for age group, gender, race, insurance, CHD severity, and interaction terms. The backwards elimination (BWE) approach was applied to determine if any interactions should be included in the models and addressed collinearity by removing inter-correlated variables; SAS default criteria for variable entry and exit in the models (0.05 for SLENTRY and SLSTAY). Odds ratio estimates and Wald confidence intervals in addition to *p*-values are reported for logistic regressions. The Hosmer-Lemeshow (HL) test statistic option assessed model goodness of fit.

3 Results

For the 5,397 patients included in the analysis, mean age was 33.2 years (SD = 13.7), 61.4% were females, 45.1% were White, 24.6% were Black, 1.1% identified themselves as race 'other' which included American Indian, Asians, native Hawaiians/Pacific Islanders, and multi-racial, and 29.2% had race unknown. Patients ranged from 18–64 years of age with more than half comprising the youngest age group, 18–29 years (51.2%, n = 2,761, mean = 22.1 years, SD = 3.4), with 19.3% between 30–39 years of age (n = 1,042, mean = 34.3 years, SD = 2.9), 12.2% were between 40–49 years old (n = 661, mean 44.2 years, SD = 2.9), and 17.3% (n = 933, mean = 57.0 years, SD = 4.3) in the 50–64 years age group. Over 18% (18.3%) of patients had a diagnosis of HTN without HF, 4.4% had a diagnosis of HF without HTN, 8.3% had both HTN and HF and 3.0% of the cohort died during the three-year study period. Patients with a severely complex CHD made up 29.0% of the sample. Public health insurance coverage was the most prevalent, 47.9%, followed by 40.2% with private insurance coverage. Only 1.1% of the cohort indicated that they were uninsured or self-pay patients and health insurance coverage was not indicated for 10.8% of the sample (Tab. 1).

Table 1: Demographics of Adult Congenital Heart Defect (ACHD) Patients, 2008–2010

	Total N = 5,397	
	N	%
Mortality	163	3.0%
Hypertension without Heart Failure	985	18.3%
Heart Failure without Hypertension	238	4.4%
Both Hypertension and Heart Failure	448	8.3%
Age Group (Years)		
18–29	2,761	51.2%
30–39	1,042	19.3%
40–49	661	12.2%
50–64	933	17.3%
Gender		
Female	3.311	61.4%
Male	2,086	38.6%
Race		
White	2,436	45.1%
Black	1,326	24.6%
^Other	61	1.1%
Unknown	1,574	29.2%
Insurance		
Any Public	2,586	47.9%
Private Only	2,171	40.2%
Uninsured/Self-pay	57	1.1%
Unknown	583	10.8%
CHD Anatomic Group		
Severe Complexity	1,565	29.0%
Shunt	1,342	24.9%
Valve	2,272	42.1%
Shunt + Valve	218	4.0%
CHD Anatomic Group (Collapsed)		
Severe Complexity	1,565	29.0%
Shunt, Valve or Shunt + Valve	3,832	71.0%
Age Group (Years)	**Mean**	**SD**
18–64	33.2	13.7
18–29	22.1	3.4
30–39	34.3	2.9
40–49	44.2	2.9
50–64	57.0	4.3

Note: ^Other race includes American Indian/Alaskan Native, Asian, Native Hawaiian/Pacific Islander and multi-racial.

Tab. 2 presents the association of mortality of patients with CHD who were diagnosed with either HTN without HF, HF without HTN, or with both HTN and HF, along with several demographics. Over the three-year study period, 3.9% of those diagnosed with HTN without HF died (ns) as did 11.8% of those diagnosed with HF without HTN ($p < 0.0001$), and 15.4% of those diagnosed with both HTN and HF ($p < 0.0001$). An age-specific trend in mortality was seen during the study period with the risk of dying greatest among older age groups: 8.8% for 50–64 year olds, 3.9% for 40–49 year olds, 1.7% among 30–39 year olds and 1.3% for 18–29 year olds ($p < 0.0001$). Survival favored females (2.6%) compared to males (3.7%) ($p < 0.05$). Black patients were more likely to die (5.4%) compared to White patients (2.9%) ($p < 0.0001$), and those with public health insurance were at significantly greater risk of dying (4.7%) compared to those with private insurance coverage (1.4%) ($p < 0.0001$).

Table 2: Hypertension, heart failure, and other comorbidities by mortality for adult Congenital Heart Defects (ACHD) patients, 2008–2010

	Mortality				
	Alive		Deceased		X^2
	n = 5,234 (97.0%)		n = 163 (3.0%)		
	n	row %	n	row %	
Hypertension without Heart Failure	947	96.1%	38	3.9%	ns
Heart Failure without Hypertension	210	88.2%	28	11.8%	**65.00*****
Both Hypertension and Heart Failure	379	84.6%	69	15.4%	**255.71*****
Age Group (Years)					
18–29	2,724	98.7%	37	1.3%	**160.11*****
30–39	1,024	98.3%	18	1.7%	
40–49	635	96.1%	26	3.9%	
50–64	851	91.2%	82	8.8%	
Gender					
Female	3,225	97.4%	86	2.6%	**5.22***
Male	2,009	96.3%	77	3.7%	
Race					
White	2,365	97.1%	71	2.9%	**51.92*****
Black	1,255	94.7%	71	5.4%	
^Other	56	91.8%	<10	–	
Unknown	1,558	99.0%	16	1.0%	
Insurance					
Any Public	2,464	95.3%	122	4.7%	**49.40*****
Private Only	2,140	98.6%	31	1.4%	
Uninsured/Self-pay	57	100.0%	0	0.0%	
Unknown	573	98.3%	10	1.72%	

(Continued)

Table 2 (continued).

	Mortality				
	Alive		Deceased		X^2
	n = 5,234 (97.0%)		n = 163 (3.0%)		
	n	row %	n	row %	
CHD Anatomic Group					
Severe Complexity	1,533	98.0%	32	2.0%	**34.80*****
Shunt	1,319	98.3%	23	1.7%	
Valve	2,167	95.4%	105	4.6%	
Shunt + Valve	215	98.6%	<10	–	
CHD Anatomic Group (Collapsed)					
Severe Complexity	1,533	98.0%	32	2.0%	**7.16****
Shunt, Valve or Shunt + Valve	3,701	96.6%	131	3.4%	
Age Group (Years)	**Mean**	**SD**	**Mean**	**SD**	***t*-test**
18–64	32.8	13.5	45.4	15.0	**−11.66*****
18–29	22.1	3.4	23.0	3.1	ns
30–39	34.3	2.9	34.9	2.4	ns
40–49	44.2	2.9	44.2	3.1	ns
50–64	56.9	4.3	58.1	4.5	**−2.59****

Notes: Cell sizes <10 not reported.
^ Other race includes American Indian/Alaskan Native, Asian, Native Hawaiian/Pacific Islander and multi-racial.
*$p < 0.05$; **$p < 0.01$; ***$p < 0.0001$.

Patients with shunt or valve (non-complex) CHD conditions were at significantly greater risk of mortality (3.4%) compared with those with complex CHD (2.0%, $p < 0.0001$), and these patients with non-complex lesions were also more likely to be diagnosed with HTN without HF (22.3%) or with both conditions (8.6%) compared to their complex CHD counterparts, 8.4% and 7.5%, respectively ($p < 0.0001$) (data not shown). This supplemental analysis also revealed that those with complex CHD lesions, were more likely to be diagnosed with HF without HTN (8.0%) compared to those with a shunt or valve CHD condition (3.0%) ($p < 0.0001$), and having a complex CHD was more prevalent among the youngest adults, 18–29 year olds (64.2.3%) compared to the 30–39 year olds (23.3%), the 40–49 year olds (8.9%) and the oldest age group (5.5%) ($p < 0.0001$).

An anatomic complexity-specific analysis revealed that those with valve lesions were at significantly greater risk of dying (4.6%) compared with those with a severe lesion (2.0%) or a shunt condition (1.7%) ($p < 0.0001$) (Tab. 3). Patients with a valve diagnosis were also more likely to have HTN without HF (27.3%) or both HTN and HF (11.5%) compared to those with a complex defect (8.4% and 7.5%, respectively) or a shunt (15.3% and 4.6%, respectively) ($p < 0.0001$). Valve (3.4%) and shunt patients (2.2%) were less likely to develop HF without HTN compared to patients diagnosed with a complex CHD (8.0%) ($p < 0.0001$). Among the two older age groups, 40–49 and 50–64 years olds, having a valve condition (52.9% and 69.5%), respectively) was more prevalent than having a shunt (25.4% and 21.1%, respectively) or a complex CHD (21.8% and 9.4%, respectively) ($p < 0.0001$) (row percentages not displayed in table), and although having a valve condition was more prevalent among females (54.1%)

compared to males (45.9%) ($p < 0.0001$), among males, valve lesions were more common (52.1%) compared to a complex CHD (27.9%) or a shunt diagnosis (20.0%) ($p < 0.0001$) (row percentages not displayed in table). Blacks were more likely to have a valve lesion (44.8%) compared to a shunt (28.5%) or a complex condition (26.7%) ($p < 0.001$) (row percentages not displayed in table). Lastly, those with valve lesions were more likely to be public insurance beneficiaries (47.9%) than have private insurance (40.6%), be uninsured or a self-payer (1.4%) or have an unknown insurance type (10.1%) ($p < 0.05$).

Table 3: Hypertension, heart failure, and other comorbidities by CHD anatomic group for Adult Congenital Heart Defects (ACHD) patients, 2008–2010

	CHD Anatomic Group						
	Severe Complexity		Shunt		Valve		
	n = 1,565 (40.2%)		n = 1,342 (25.9%)		n = 2,272 (43.9%)		
	n	col %	n	col %	n	col %	X^2
Mortality	32	2.0%	23	1.7%	105	4.6%	**32.00*****
Hypertension without Heart Failure							
	131	8.4%	205	15.3%	621	27.3%	**233.52*****
Heart Failure without Hypertension							
	125	8.0%	30	2.2%	76	3.4%	**67.90*****
Both Hypertension and Heart Failure							
	117	7.5%	62	4.6%	261	11.5%	**54.19*****
Age Group (Years)							
18–29	976	62.4%	696	51.9%	961	42.3%	**414.66*****
30–39	364	23.3%	292	21.8%	340	15.0%	
40–49	139	8.9%	162	12.1%	338	14.9%	
50–64	86	5.5%	192	14.3%	633	27.9%	
Gender							
Female	1,006	64.3%	942	70.2%	1,229	54.1%	**100.39*****
Male	559	35.7%	400	29.8%	1,043	45.9%	
Race							
White	713	45.6%	517	38.5%	1,091	48.0%	**46.20*****
Black	341	21.8%	364	27.1%	573	25.2%	
Other$^{\wedge}$	19	1.2%	20	1.5%	20	0.9%	
Unknown	492	31.4%	441	32.9%	588	25.9%	
Insurance							
Any Public	763	48.8%	673	50.2%	1,088	47.9%	**13.25***
Private Only	639	40.8%	497	37.0%	923	40.6%	
Uninsured/Self-pay	<10	–	13	1.0%	31	1.4%	
Unknown	154	9.8%	159	11.9%	230	10.1%	

(Continued)

Table 3 (continued).

	CHD Anatomic Group						
	Severe Complexity		Shunt		Valve		
	n = 1,565 (40.2%)		n = 1,342 (25.9%)		n = 2,272 (43.9%)		
	n	col %	n	col %	n	col %	X^2
Age Group (Years)	**Mean**	**SD**	**Mean**	**SD**	**Mean**	**SD**	**F-test**
18–64	29.0	10.1	32.4	12.9	36.8	15.4	**162.64*****
18–29	22.6	3.4	22.2	3.4	21.4	3.3	**29.71*****
30–39	34.1	2.9	34.3	2.9	34.5	3.0	ns
40–49	43.9	2.8	43.7	3.0	44.6	2.9	**6.55****
50–64	56.0	4.5	56.7	4.3	57.2	4.2	**3.48***

Notes: Cell sizes <10 not reported.
^Other race includes American Indian/Alaskan Native, Asian, Native Hawaiian/Pacific Islander and multi-racial.
*$p < 0.05$; **$p < 0.01$; ***$p < 0.0001$.

Crude logistic models of mortality revealed that the likelihood of dying during the study period was almost 5 times greater for those with HF without HTN (4.96, 95%CI 3.23–7.63), and almost double that for those with both HTN and HF (9.40, 95%CI 7.68–13.05) (Tab. 4). The two oldest age groups were more likely to die compared to the 18–29 year olds, [(40–49 year olds: 3.0, 95%CI 1.81–5.02) and (50–64 year olds: 7.10, 95%CI 4.78–10.54). Patients with non-complex CHD (shunts, valves and shunts + valves) were 1.69 times more likely to die during the study period compared to patients with complex CHDs (95%CI 1.15–2.51). When valve patients were assessed separately from those with shunts, the odds of dying increased to 2.32 times greater compared to patients with severe complexity types of CHD (95%CI 1.55–3.47). Blacks compared to whites were almost twice as likely to die (1.88, 95%CI 1.35–2.64) and those covered by public insurance were over three times more likely to die during the study period compared to those with covered by private health insurance (3.42, 95%CI 2.29–5.09) (Tab. 4).

Table 4: Crude odds for the association of hypertension, heart failure and other comorbidities with mortality for Adult Congenital Heart Defect (ACHD) patients, 2008–2010

	Crude OR	95%CI	
Hypertension without Heart Failure	1.38	.95	1.99
Heart Failure without Hypertension	**4.96**	**3.23**	**7.63*****
Both Hypertension and Heart Failure	**9.40**	**7.68**	**13.05*****
Age Group (Years)			
18–29	1.00	–	–
30–39	1.29	0.73	2.28
40–49	**3.01**	**1.81**	**5.02*****
50–64	**7.10**	**4.78**	**10.54*****

(Continued)

Table 4 (continued).

	Crude OR	95%CI	
Gender			
Male	1.00	–	–
Female	**0.70**	**0.51**	**0.95***
Race^			
White	1.00	–	–
Black	**1.88**	**1.35**	**2.64*****
Insurance			
Any Public	**3.42**	**2.29**	**5.09**
Private Only	1.00	–	–
CHD Anatomic Group^			
Severe Complexity	1.00	–	–
Shunt	0.84	0.49	1.44
Valve	**2.32**	**1.55**	**3.47*****
CHD Anatomic Group (collapsed)			
Severe Complexity	1.00	–	–
Shunt, Valve or Shunt + Valve	**1.69**	**1.15**	**2.51****

Notes: 95%CIs (confidence intervals) not including '1' are in **bold** indicating odds for groups are significantly different.
^ Other & unknown race categories and shunt + value category of 4-level CHD anatomic grouping excluded from analysis due to cell sizes <10 for deceased.
*$p < 0.05$, **$p < 0.01$, ***$p < 0.001$.

Tab. 5 shows final adjusted multivariate logistic regression models that assess the association of mortality for those patients with CHD who are also diagnosed with either HF without HTN or both HTN and HF separately controlling for select demographics; odds ratios, Wald 95% likelihood confidence intervals (CI) and *p*-values are reported. These models employed the backward elimination approach (BWE) and included age group, gender, race, health insurance coverage and CHD anatomic grouping as independent predictors of mortality along with interaction terms for each of the three primary comorbid conditions (HTN without HF, HF without HTN and both) with each demographic predictor. Since the association of HTN without HF failed to predict mortality, this multivariate model and its results are not reported in Tab. 5. Model 1 includes HF without HTN as the primary comorbid predictor of mortality, while Model 2 includes having both HTN and HF.

Table 5: Adjusted odds of heart failure with and without hypertension controlling for other comorbidities and interactions on mortality for Adult CHD (ACHD) patients, 2008–2010

	Final Model 1	Final Model 2
	OR (95%CI)	
VARIABLES		
Heart Failure w/o Hypertension	**2.85 (2.09–3.61)*****	
Hypertension & Heart Failure		**2.71 (1.56–3.88)*****

(Continued)

Table 5 (continued).

	Final Model 1	Final Model 2
Age Group (Years)		
30–39 year olds *vs.* 18–29	1.38 (0.66–2.87)	0.82 (0.41–1.62)
40–49 year olds *vs.* 18–29	**2.92 (1.51–5.65)****	1.62 (0.90–2.90)
50–64 year olds *vs.* 18–29	**6.38 (3.69–11.01)*****	**2.67 (1.63–4.40)*****
Race^ Blacks *vs.* Whites	**1.75 (1.21–2.55)****	1.26 (0.81–1.96)
Insurance Any Public *vs.* Private Only	**2.22 (1.43–3.54)**	**2.34 (1.38–3.97)****
CHD Anatomic Group^		
Shunt *vs.* Severe Complexity	0.86 (0.44–1.67)	0.85 (0.38–1.92)
Valve *vs.* Severe Complexity	**1.86 (1.11–3.13)***	**2.33 (1.24–4.38)****
INTERACTIONS		
Heart Failure w/o Hypertension * Age 30–39	**0.09 (0.01–0.60)**	
Heart Failure w/o Hypertension * Age 40–49	0.36 (0.09–1.30)	
Heart Failure w/o Hypertension * Age 50–64	**0.10 (0.02–0.40)**	
Hypertension & Heart Failure * Race Blacks		**2.29 (1.06–5.05)***
Hypertension & Heart Failure * Insurance Any Public		**0.38 (0.15–0.97)***
Hypertension & Heart Failure * CHD Shunt		0.81 (0.21–3.03)
Hypertension & Heart Failure * CHD Valve		**0.25 (0.09–0.65)****

Notes: Model 1 includes heart failure w/o hypertension as primary comorbidity; Model 2 includes having both hypertension & heart failure. Gender eliminated from both models as it failed to meet minimum 0.05 criterion.
Model 1: Interactions of hypertension w/o heart failure with gender, race, insurance and CHD anatomic complexity were eliminated. Model 2: interaction of having both hypertension & heart failure with age was eliminated.
95%CIs (confidence intervals) not including '1' are in **bold** indicating odds for groups are significantly different.
^Other & unknown race categories and shunt+value category of 4-level CHD anatomic grouping excluded from analysis due to cell sizes <10 for deceased.
$^{*}p < 0.05$, $^{**}p < 0.01$, $^{***}p < 0.001$

The log odds equation used for the final adjusted models is:

Logit P (Death) = α+β1(primary comorbidity: [HF w/o HTN] or [Both HTN & HF]) + β2(Age Group)
+ β3(Gender) + β4(Race) + β5(Insurance) + β6(CHD Anatomic Group)
+ β7(primary comorbidity*Age Group) + β8(primary comorbidity*Gender)
+ β9(primary comorbidity*Race) + β10(primary comorbidity*Insurance)
+ β11(primary comorbidity*CHD Anatomic Group)

For Model 1, ACHD patients who had a diagnosis of HF without HTN were almost three times more likely to die (2.85, 95%CI 2.09–3.61), and dying was 1.75 times greater for Blacks than Whites (95%CI 1.21–2.55), and almost three times more likely for 40–49 year olds and over six times greater for 50–64 year olds compared to the youngest 18–29 age group (2.92, 95%CI 1.51–5.65 and 6.38, 95%CI 3.69–11.01, respectively). Patients covered by public health insurance were also twice as likely to die than those covered by private health insurance (2.22, CI95% 1.43–3.54). Patients with a valve lesion were at greater risk of dying during the three-year study period compared to those patients with a complex CHD (1.86, CI95% 1.11–3.13). When the combined effects of age with having a HF without

HTN diagnosis on mortality were assessed for ACHD patients, 30–39 and 50–64 year olds with HF without HTN were favored to survive during the study period compared to their younger 18–29 year old counterparts (0.09, 95%CI 0.01–0.60) and (0.10, 95%CI 0.02–0.40), respectively. The BWE approach removed all other interaction terms, except for HF without HTN * age group, for failing to meet criterion for retention in the model. The Hosmer and Lemeshow Goodness-of-Fit Test (HL GoF) revealed that the data adequately fit this adjusted model ($X^2 = 5.69$, df = 8, $p < 0.68$).

In Model 2, ACHD patients diagnosed with both HTN and HF were almost three times more likely to die (2.71, 95%CI 1.56–4.23) and the oldest age group (50–64 year olds) had significantly greater odds of dying compared to the youngest group of patients with CHD (2.67, 95%CI 1.63–4.40). Patients with CHD who also had concomitant HTN and HF diagnoses were twice as likely to die if covered by public health insurance compared to those with private health insurance coverage (2.34, 95%CI 1.38–3.97). In addition, those with valve CHD lesions had 2.33 times greater mortality compared to those with a complex CHD (95%CI 1.24–4.38). Although race was not shown to influence mortality statistically, Blacks with Concomitant HTN and HF diagnoses were 2.29 times more likely to die compared to Whites diagnosed with both HTN and HF (95%CI 1.06–5.05). Lastly, public insurance beneficiaries with both HTN and HF were favored for survival compared to private insurance beneficiaries with both diagnoses, and the same was true for HTN and HF patients with valve lesions in comparison to those with both comorbid conditions and complex CHDs [(038, 95%CI 0.15–0.97) and (0.25, 95%CI 0.09–0.65). This adjusted model also adequately fit the data (HL GoF $X^2 = 7.21$, df = 8, $p < 0.51$).

4 Discussion

As adults with CHD represent a large proportion of the CHD population [21], it is important to understand contributors to mortality in the ACHD population, which may differ from a pediatric population. Decisions regarding care are often made based on anatomic complexity alone, although more recent ACHD guidelines integrate physiology [22]. While HF is known to account for a significant proportion of deaths in patients with CHD [5,6], the predictive value of HF with and without HTN on mortality in ACHD has not been clearly elucidated in the context of CHD anatomic complexity. This cross-sectional retrospective study revealed that adults with CHD experience significant mortality regardless of their anatomic CHD complexity, with even those considered to have simple defects experiencing increased mortality. CHD patients with any HF codes accounted for 12.7% (=686/5,397) of the cohort, but comprised 59.5% (=97/163) of all those who died. The prevalence of any HTN among adults at least 18 years of age and older is 29.0% as reported by CDC 2015–2016 [23]. In the current study, the overall proportion of those with HTN was 26.6% for ages 18–64 years, which is slightly less than CDC's reported U.S. population-based prevalence. In mimicking an age-specific analysis using CDC age groupings of 18–39 years, 40–59 years and 60 and older [23] to our current data, a similar age trend for HTN was found: 13.5% for ages 18–39 years old, 53.7% for ages 40–59 year olds, and 74.3% for those 60 and older. This is comparable to U.S. HTN rates which shows increases for HTN with age from 7.5% between ages 18–39 years, to 33.2% between ages 40–59 years and 63.1% for 60 years and older [23]. While US population-based data reveals the highest prevalence of HTN among the black population (40.3%) [23], prevalence of HTN among blacks in the current study was lower at 27.0%. HF has been reported to affect 2.4% of the US population and is also seen to increase with age [24]. After the age of 79 years, 12% of the population has HF [24]. The proportion of individuals with HF in our cohort (12.7%) is higher than the published population-based prevalence. These findings emphasize the importance of ongoing surveillance for cardiac comorbidities in all patients with heart defects, as the development of HF is an important predictor of mortality in those with complex anatomy, shunt and valve lesions, and may occur at younger ages.

Unadjusted prevalence of ACHD patients diagnosed with HF without HTN and those diagnosed with both HF and HTN died approximately 5 times and almost double that rate, respectively. Overall mortality was 3.0% for this 3-year ACHD sample, and their age-related mortality was 1.3%, 1.7%, 3.9% and 8.8% for 18–29, 30–39, 40–49 and 50–64 year olds, respectively. In a supplemental age-specific analysis of mortality, when we constrained the sample only to those with HF, with or without hypertension, overall mortality in this 3-year study period showed an almost five-fold increase to 14.1% (=97/686), with mortality incrementally increasing from 23.7% (=23/97) in the youngest age group (18–29 year olds) to 46.4% (=45/97) in the oldest (50–64 year olds). Unadjusted logistic regression results showed a lower likelihood of mortality for ACHD patients who had a HF without HTN diagnosis (OR = 4.96) compared to ACHD with both comorbidities (OR = 9.40). Prior large-scale studies have noted a J- or U-shaped mortality curve in those hospitalized with HF possibly related to the worse outcomes seen in those who have HF with lower blood pressure [12,13]. The reason for this finding may be related to the proposed protective effect of blood pressure elevation in those with a HF diagnosis [25] or the known poor prognosis of hypotension [14] in association with HF. Although HTN is a significant risk factor for the development of HF, a similar J-shaped survival curve has been noted in individuals with HTN [26]. Previous studies have shown a higher mortality in men with HF compared to women [27], and have shown a possible U-shaped mortality curve based on age in adults with HF, with a higher mortality in those under 25 years of age and greater than 64 years of age, and the lowest mortality in the middle group [27]. While crude analyses favored females for survival, unlike prior studies, adjusted logistic regression did not demonstrate mortality being impacted by gender. Also, unadjusted mortality results revealed racial differences with black patients with CHD dying 1.88 times the rate of whites, and CHD severity differences for those with valve CHD lesions dying 2.32 times the rate of ACHD patients with complex CHDs.

Results from the final multivariate adjusted logistic regression model which included HF without HTN as the primary comorbidity (Model 1) revealed that Black ACHD patients were 1.75 times more likely to die compared to White ACHD patients, and that the two older age groups, 40–49 year olds and 50–64 year olds, were at risk for greater mortality compared to the youngest ACHD individuals (2.92, 95%CI 1.51–5.65 and 6.38, 95%CI 3.69–11.01, respectively). The adjusted multivariate model for ACHD patients with both HTN and HF diagnoses (Model 2) revealed a comparable increased mortality for the oldest patients (50–64 year olds) when compared to 18–29 years olds (2.67, 95%CI 1.63–4.40). Those with complex CHD tend to be younger, which may account for some of these differences in mortality across anatomic groups. Anatomic CHD group was predictive of mortality in patients with HF without HTN, suggesting less complex heart defects such as valvular disease and shunts may have a cumulative detrimental effect on heart function over time, leading to heart failure in these anatomic groups over time. The development of HF in adults with less complex heart defects may also be related to the interplay of their heart defect with development of acquired comorbid conditions. Mortality within this ACHD population may be more complex and influenced by the effects of having a concomitant HF diagnosis with CHD.

In the multivariate logistic models for those with HF codes in Models 1 and 2, older individuals with ACHD (50–64 year olds) had greater odds of dying compared to the younger age group, 18–29 years [(6.38, 95%CI 3.69–11.01) and (2.67, 95%CI 1.63–4.40), respectively]. In this study, complex heart defects accounted for a larger proportion of CHD in younger adults (29.0% = 1,565/5,397). The younger age groups were the least likely to die during the study window despite having more complex CHD and HF compared to their older age counterparts with less severe CHD and HF, suggesting age has a protective effect for those with comorbid HF. When the interactive effects of age and having HF without HTN on mortality were assessed irrespective of CHD severity, results revealed that one of the younger adult groups, 30–39 year olds, and the oldest age group, 50–64 year olds, were both favored for survival. Khairy et al. also reported a bimodal pattern of mortality which initially peaked in childhood and then

again during late adulthood, with the earlier peak perhaps relating to a contribution by children with severe unrepaired or palliated CHDs [7]. Additionally, in an aging CHD population, common comorbid conditions such as diabetes, pulmonary hypertension, atrial fibrillation and other comorbidities may influence mortality [28] and may also contribute to the development of HF.

Lastly, for patients with both HF, with or without HTN, having a valve lesion was associated with a higher likelihood of death compared to those in the complex group or with shunt defects [(HF without HTN: 1.86, 95%CI 1.11–3.13) and (HTN and HF: 2.33, 95%CI 1.24–4.38)]. There are several possible explanations for this finding. The most likely explanation is a protective effect of younger age on mortality. In addition, anatomically complex CHD was defined as defects that typically require intervention or surgery in the first year of life [18,19]. In an ACHD population this may introduce survivor bias, as some children with complex CHD may not reach adulthood. Conversely, the effect of reparative surgery in early childhood may mitigate some of the risk of death in adulthood, and 'simple' shunt or valve heart defects may not capture the long-term cumulative effects of valve and shunt lesions on ventricular function, or the cumulative effects of multiple surgeries for valvular heart defects, which may lead to the development of ventricular dysfunction and HF in adulthood, particularly in the older age groups. Aortic valve disease, for example, may require repeated interventions over a lifetime [29,30], may also be associated with other left sided obstructive lesions such as coarctation of the aorta, and both aortic stenosis and regurgitation may contribute to the development of left ventricular dysfunction and HF [29,30]. The finding of increased mortality in all CHD groups warrants further study to understand contributors such as multiple surgeries over a life-time, and the potential consequences of unrepaired or late repair of shunt or valve lesions, the consequences of repeated surgeries for valvular heart defects, and reevaluation of the perception that adults with anatomically simple heart defects have a benign course requiring less frequent surveillance.

This study is limited by its cross-sectional retrospective design, use of administrative data, the inability to definitely verify diagnosis and comorbidity codes, define completeness of repairs, or measure blood pressure. Anatomic groupings are limited by use of ICD-9-CM codes, which may include cases that cross anatomic groups. Eisenmenger syndrome does not have a code in ICD-9-CM, and thus, could not be taken into account. Since our data evaluated individuals from the state of Georgia, results may not be broadly applicable to other geographic regions. In addition, our study window was only three years, and a longer study period may enable better detection of contributors to mortality from HTN. More complete race data would also allow for better assessments of comorbidities that historically are known to be more prevalent within certain racial or ethnic groups, and perhaps, shed light on racial disparities when they exist.

In conclusion, we found in a population-based cohort from Georgia, that adults with heart defects who also had HF experienced significant mortality in a 3-year period of time, and this risk for mortality was age-related. Patients with CHD valve lesions who were also diagnosed with HF, with or without HTN, were at increased risk of death. In older individuals, mortality was higher compared to younger individuals, and Black patients with CHD who had HF without HTN had higher mortality than Whites. Lastly, patients with CHD who also were diagnosed with either HF without HTN or both HTN and HF and who had public health insurance coverage had a greater risk of mortality than those covered by private health insurance carriers. Vigilance for signs and symptoms of HF in adults with all heart defects, and treatment to mitigate comorbid HF and HTN may lessen mortality in adults with CHD regardless of the anatomic complexity of the heart defect. Results from this study suggest factors in addition to anatomic complexity may more accurately predict the contributors to mortality in adults with CHD, emphasizing the need for all ACHD patients to stay in specialty care. Future studies should evaluate other comorbidities that are known to influence health outcomes.

Acknowledgement: The findings and conclusions in this report are those of the authors and do not necessarily represent the official position of the Centers for Disease Control and Prevention. The authors thank Trenton Hoffman, Sr. Data Analyst, for his expertise and assistance in preparing the analytic dataset, and Lindsey Ivey for helping confirm the references.

Data Sharing: The individual deidentified participant data cannot be shared.

Funding Statement: Cheryl Raskind-Hood and Wendy M. Book are supported by a Cooperative Agreement from the Centers for Disease Control and Prevention Cooperative Agreement, Public Health Pilot Project *Surveillance of Congenital Heart Defects (CHDs) Focusing on Adolescents and Adults;* FOA #DD12-1207.

Conflicts of Interest: The authors declare that they have no conflicts of interest to report regarding the present study.

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Appendix A: CHD Data Sources

Data Source	Description
Children's Healthcare of Atlanta (CHOA) and Sibley Heart Center	Children's Healthcare of Atlanta (CHOA) consists of pediatric facilities across Georgia dedicated to treating and providing care to children and adolescents. Ranked 4th in the country by U.S. News & World Report, Sibley Heart Center is one of the top pediatric cardiac programs with 20 outpatient locations and 40 hospitals in Georgia. Sibley offers a spectrum of cardiac programs and services from birth until the age of 21 and has multidisciplinary teams offering specialized care designed especially for children and adolescents who need treatment and management of cardiovascular conditions. Sibley has 22 outpatient locations in Georgia. Medicaid covers 57% of CHOA/Sibley patients; 4% are uninsured. In the pilot surveillance system, 40% of CHD individuals from CHOA and 26% from Sibley identified as black. Our pediatric collaborators currently care for an estimated more than 90% of children and adolescents in Georgia. https://www.choa.org/medical-services/heart-center
ECDW (Emory Clinical Data Warehouse from Emory Healthcare (EHC)) and St. Joseph's Hospital (SJH)	Emory Healthcare (EHC) is the largest health care system in Georgia, encompassing many hospitals, clinics, and local practices, including St. Joseph's Hospital (SJH) (as of 2011) and community-based specialty associates. EHC houses Georgia's only comprehensive adult CHD center, the Emory Adult Congenital Heart Center (EACHC), physically located in DeKalb County. EACHC serves a diverse population (10% uninsured, 27% government insurance, 25% over age 44). Data were obtained through querying of Emory Clinical Data Warehouse (ECDW) and electronic data capture tools. http://it.emory.edu/ClinicalResearchData/data-sources/clinical-data-warehouse/index.html
Grady Health System	Grady Health System is one of the region's premier level 1 Trauma Centers committed to improving the health and providing quality comprehensive healthcare to the underserved primarily in Fulton and DeKalb counties, as well as other metro-Atlanta counties and the state of Georgia. Grady Health manages ~700K patients/year with a majority being enrolled in either Medicare or Medicaid.* It serves an ethnically diverse and socioeconomically disadvantaged population, with 75% African Americans, 20% Hispanics and Asians, 5% Caucasians. About half have no insurance and half have public health insurance. Grady Health has 8 facilities in Atlanta, and its Cardiac Clinic provides comprehensive cardiac care housed at the main hospital in downtown Atlanta. *https://www.gradyhealth.org/about-us/
Pediatric Cardiology Services (PCS)	Pediatric Cardiology Services (PCS) is a group of certified pediatric cardiologists who specialize in the care of infants, children, and adolescents in need of high-risk cardiac care. During the CHD pilot study, there were six locations across Georgia that provided comprehensive cardiac services including cardiac evaluation with diagnostic equipment, prevention counseling, and management of cardiac problems. PCS had close affiliations with EHC, providing cardiac care to over 37K patients within the Atlanta area. At the writing of this manuscript, PCS has since been subsumed by other pediatric cardiology healthcare systems in Georgia like CHOA/Sibley.

(Continued)

Appendix A (continued).

Data Source	Description
Medicaid Claims	Georgia Medicaid administrative claims for individuals with a CHD diagnosis were obtained from the Centers for Medicare and Medicaid Services (CMS) via Research Data Assistance Center (ResDAC), a CMS contractor. Medicaid is a social health care program for families and individuals with low income and resources. The state and federal governments jointly fund the program, with each state having its own criteria for determining eligibility based on state demographics and geography. In Georgia, the Medicaid program provides health care for more than 1.7 million residents with low incomes including children, pregnant women, the disabled, and the blind.* Disability claims for persons with CHDs are made to the Social Security Administration (SSA) if they meet the general disability requirements and qualify for symptomatic congenital heart disease.** Even if they do not qualify for one of the listing requirements for symptomatic congenital heart disease, they may still be approved for disability through a physical residual functioning capacity assessment, via the opinion of a licensed physician, and through evidence of emotional/psychological impairment or complications.** Medicaid files come from the Medicaid Statistical Information System (MSIS) from which Medicaid Analytic eXtract (MAX) files are constructed.^ Medicaid data include eligibility status, demographics, claims histories with diagnosis codes, procedure codes, and dates of service. Medicaid data are obtained strictly from billing records, and so, this source is considered to be administrative. *https://dch.georgia.gov/about-us/publications/annual-reports/DCH FY2011 Annual Report_web.pdf **https://kaiserfamilyfoundation.files.wordpress.com/2013/01/8048.pdf ^https://www.resdac.org/cms-data/file-family/Medicaid-Analytic-Extracts-MAX

Appendix B: Congenital Heart Defect Severity Classification [18]

Category	ICD-9-CM Code	Code Description
Severe *(If case has a severe code, regardless of presence of shunt, valve, or other codes)*	745.0	Common truncus
	745.1	Transposition of the great arteries (TGA)
	745.10	Complete TGA (dextro-TGA), not otherwise specified (NOS) or classical
	745.11	Double outlet right ventricle, or incomplete TGA
	745.12	Corrected TGA (levo-TGA)
	745.19	TGA Other
	745.2	Tetralogy of Fallot
	745.3	Single ventricle, or cor triloculare
	745.6	Endocardial cushion defect
	745.60	Endocardial cushion defect unspecified
	745.69	Endocardial cushion defect, other
	746.01	Pulmonary valve atresia or absence
	746.1	Tricuspid atresia, stenosis or absence
	746.7	Hypoplastic left heart syndrome
	747.11	Interrupted aortic arch
	747.41	Total anomalous pulmonary venous return
Shunt + Valve *(Case has shunt AND valve codes)*	A combination of the shunt/valve codes below	A combination of the shunt/valve defects below
Shunt *(Case has at least one shunt code, no valve or severe codes)*	745.4	Ventricular septal defect (VSD)
	745.5	Atrial septal defect (ASD secundum or Patent Foramen
	745.61	Ovale
	745.8	ASD primum
	745.9	Other specified defect of septal closure
	747.0	Unspecified defect of septal closure
	747.42	Patent ductus arteriosus (PDA)
		Partial anomalous venous return
Valve *(Case has at least one valve code, no shunt or severe codes)*	746.0	Anomalies of pulmonary valve
	746.00	Pulmonary valve anomaly, unspecified
	746.02	Pulmonary valve stenosis
	746.09	Pulmonary valve anomaly, other
	746.2	Ebstein Anomaly
	746.3	Aortic valve stenosis
	746.4	Aortic insufficiency or bicuspid/unicuspid aortic valve
	746.5	Mitral stenosis or mitral valve abnormalities
	746.6	Mitral insufficiency
	764.81	Subaortic stenosis
	746.83	Infundibular or subvalvar pulmonary stenosis
	747.1/747.10	Coarctation of aorta
	747.22	Atresia or stenosis of aorta
	747.3	Anomalies of pulmonary artery
	747.31	Pulmonary artery atresia, coarctation, or hypoplasia
	747.39	Anomalies of pulmonary artery, other

(Continued)

Appendix B (continued).

Category	ICD-9-CM Code	Code Description
Other Only	745.7	Cor biloculare
(Case only has one or more codes in this category)	746.8	Other specified anomalies of heart
	746.82	Cor triatriatum
	746.84	Obstructive anomalies of heart
	746.85	Coronary artery anomaly
	746.87	Malposition of heart or apex
	746.89	Other specified anomaly of heart (various types)
	746.9	Unspecified defect of heart
	747.2	Other anomalies of the aorta
	747.20	Anomalies of aorta, unspecified
	747.21	Anomaly of aortic arch
	747.29	Other anomalies of aorta, other specified
	747.4	Anomalies of great veins
	747.40	Anomalies of great veins, unspecified
	747.49	Other anomalies of great veins
	747.9	Unspecified anomalies of circulatory system
	648.5x	Congenital cardiovascular disorders in the mother
	V13.5	Personal history of (corrected) congenital malformations of heart and circulatory system

Appendix C: ICD-9-CM Codes Determining Hypertension (HTN) (40 codes)

ICD-9-CM Code	ICD-9-CM Description
401.1	Benign Hypertension
401.9	Hypertension Nos
401.0	Malignant Hypertension
403.0	Mal Hypertens Renal Dis (Begin 1980 End 1989)
403.1	Benign Hypert Renal Dis (Begin 1980 End 1989)
403.9	Hypertens Renal Dis Nos (Begin 1980 End 1989)
404.0	Mal Hypert Hrt/Renal Dis (Begin 1980 End 1989)
404.1	Ben Hypert Hrt/Renal Dis (Begin 1980 End 1989)
404.9	Hypert Hrt/Renal Dis Nos (Begin 1980 End 1989)
437.2	Hypertens Encephalopathy
402.00	Mal Hyperten Hrt Dis Nos
402.01	Mal Hypert Hrt Dis W Chf
402.10	Ben Hyperten Hrt Dis Nos
402.11	Benign Hyp Hrt Dis W Chf
402.90	Hypertensive Hrt Dis Nos

(Continued)

Appendix C (continued).

ICD-9-CM Code	ICD-9-CM Description
402.91	Hyperten Heart Dis W Chf
403.00	Mal Hyp Ren W/O Ren Fail (Begin 1989)
403.01	Mal Hyp Ren W Renal Fail (Begin 1989)
403.10	Ben Hyp Ren W/O Ren Fail (Begin 1989)
403.11	Ben Hyp Renal W Ren Fail (Begin 1989)
403.90	Hyp Ren Nos W/O Ren Fail (Begin 1989)
403.91	Hyp Renal Nos W Ren Fail (Begin 1989)
404.00	Mal Hy Ht/Ren W/O Chf/Rf (Begin 1989)
404.01	Mal Hyper Hrt/Ren W Chf (Begin 1989)
404.02	Mal Hy Ht/Ren W Ren Fail (Begin 1989)
404.03	Mal Hyp Hrt/Ren W Chf & Rf (Begin 1989)
404.10	Ben Hy Ht/Ren W/O Chf/Rf (Begin 1989)
404.11	Ben Hyper Hrt/Ren W Chf (Begin 1989)
404.12	Ben Hy Ht/Ren W Ren Fail (Begin 1989)
404.13	Ben Hyp Hrt/Ren W Chf & Rf (Begin 1989)
404.90	Hy Ht/Ren Nos W/O Chf/Rf (Begin 1989)
404.91	Hyper Hrt/Ren Nos W Chf (Begin 1989)
404.92	Hy Ht/Ren Nos W Ren Fail (Begin 1989)
404.93	Hyp Ht/Ren Nos W Chf & Rf (Begin 1989)
405.01	Mal Renovasc Hypertens
405.09	Mal Second Hyperten Nec
405.11	Benign Renovasc Hyperten
405.19	Benign Second Hypert Nec
405.91	Renovasc Hypertension
405.99	Second Hypertension Nec

Number and Percent of Patients with Hypertension (HTN) Who Have These 40 Codes

ICD-9-CM Code	ICD-9-CM Description	# patients w/code	% patients w/code
401.9	Hypertension Nos	1226	85.6%
401.1	Benign Hypertension	541	37.8%
401.0	Malignant Hypertension	116	8.1%
403.91	Hyp Renal Nos W Ren Fail (Begin 1989)	82	5.7%
402.10	Ben Hyperten Hrt Dis Nos	76	5.3%
403.9	Hypertens Renal Dis Nos (Begin 1980 End 1989)	71	5.0%
403.90	Hyp Ren Nos W/O Ren Fail (Begin 1989)	64	4.5%
402.90	Hypertensive Hrt Dis Nos	59	4.1%
402.91	Hyperten Heart Dis W Chf	26	1.8%
403.11	Ben Hyp Renal W Ren Fail (Begin 1989)	24	1.7%
402.11	Benign Hyp Hrt Dis W Chf	19	1.3%
403.01	Mal Hyp Ren W Renal Fail (Begin 1989)	11	0.8%
402.00	Mal Hyperten Hrt Dis Nos	10	0.7%
404.91	Hyper Hrt/Ren Nos W Chf (Begin 1989)	10	0.7%
405.99	Second Hypertension Nec	8	0.6%
403.00	Mal Hyp Ren W/O Ren Fail (Begin 1989)	7	0.5%
404.93	Hyp Ht/Ren Nos W Chf & Rf (Begin 1989)	7	0.5%
437.2	Hypertens Encephalopathy	6	0.4%
404.9	Hypert Hrt/Renal Dis Nos (Begin 1980 End 1989)	5	0.3%
405.09	Mal Second Hyperten Nec	5	0.3%
403.1	Benign Hypert Renal Dis (Begin 1980 End 1989)	4	0.3%
403.10	Ben Hyp Ren W/O Ren Fail (Begin 1989)	4	0.3%
405.19	Benign Second Hypert Nec	4	0.3%
405.91	Renovasc Hypertension	4	0.3%
404.92	Hy Ht/Ren Nos W Ren Fail (Begin 1989)	3	0.2%
405.01	Mal Renovasc Hypertens	3	0.2%
404.1	Ben Hypert Hrt/Renal Dis (Begin 1980 End 1989)	2	0.1%
402.01	Mal Hypert Hrt Dis W Chf	2	0.1%
404.01	Mal Hyper Hrt/Ren W Chf (Begin 1989)	2	0.1%
404.90	Hy Ht/Ren Nos W/O Chf/Rf (Begin 1989)	2	0.1%
404.00	Mal Hy Ht/Ren W/O Chf/Rf (Begin 1989)	1	0.1%
404.03	Mal Hyp Hrt/Ren W Chf/Rf (Begin 1989)	1	0.1%
404.10	Ben Hy Ht/Ren W/O Chf/Rf (Begin 1989)	1	0.1%

(Continued)

(continued).

ICD-9-CM Code	ICD-9-CM Description	# patients w/code	% patients w/code
404.11	Ben Hyper Hrt/Ren W Chf (Begin 1989)	1	0.1%
404.13	Ben Hyp Hrt/Ren W Chf & Rf (Begin 1989)	1	0.1%
405.11	Benign Renovasc Hyperten	1	0.1%
403.0	Mal Hypertens Renal Dis (Begin 1980 End 1989)	0	0.0%
404.0	Mal Hypert Hrt/Renal Dis (Begin 1980 End 1989)	0	0.0%
404.02	Mal Hy Ht/Ren W Ren Fail (Begin 1989)	0	0.0%
404.12	Ben Hy Ht/Ren W Ren Fail (Begin 1989)	0	0.0%

Note: # patients with at least one of these 40 ICD-9-CM HTN-related codes = 1433

Appendix D: ICD-9-CM Codes Determining Heart Failure (HF) (16 codes)

ICD-9-CM Code	ICD-9-CM Description
428.0	Congestive Heart Failure
428.1	Left Heart Failure
428.9	Heart Failure Nos
398.91	Rheumatic Heart Failure
428.20	Unspecified Systolic Heart Failure (Begin 2002)
428.21	Acute Systolic Heart Failure (Begin 2002)
428.22	Chronic Systolic Heart Failure (Begin 2002)
428.23	Acute on Chronic Systolic Heart Failr (Begin 2002)
428.30	Unspecified Diastolic Heart Failure (Begin 2002)
428.31	Acute Diastolic Heart Failure (Begin 2002)
428.32	Chronic Diastolic Heart Failure (Begin 2002)
428.33	Acute On Chronic Diastolic Heart Failr (Begin 2002)
428.40	Unspec Cmbined Syst & Dias Heart Failr (Begin 2002)
428.41	Acute Cmbined Syst & Dias Heart Failr (Begin 2002)
428.42	Chron Cmbined Syst & Dias Heart Failr (Begin 2002)
428.43	Acu Chro Combi Syst & Dias Hrt Failr (Begin 2002)

(Continued)

Number and Percent of Patients with Heart Failure (HF) Who Have These 16 Codes

ICD-9-CM Code	ICD-9-CM Description	#Patients w/code	%Patients w/code
428.0	Congestive Heart Failure	395	57.6%
428.1	Left Heart Failure	178	25.9%
428.9	Heart Failure Nos	130	19.0%
428.21	Acute Systolic Heart Failure (Begin 2002)	116	16.9%
428.22	Chronic Systolic Heart Failure (Begin 2002)	106	15.5%
428.23	Acute on Chronic Systolic Heart Failr (Begin 2002)	95	13.8%
428.32	Chronic Diastolic Heart Failure (Begin 2002)	39	5.7%
428.33	Acute On Chronic Diastolic Heart Failr (Begin 2002)	33	4.8%
428.41	Acute Cmbined Syst & Dias Heart Failr (Begin 2002)	33	4.8%
428.31	Acute Diastolic Heart Failure (Begin 2002)	31	4.5%
428.43	Acu Chro Combi Syst & Dias Hrt Failr (Begin 2002)	29	4.2%
428.20	Unspecified Systolic Heart Failure (Begin 2002)	26	3.8%
428.30	Unspecified Diastolic Heart Failure (Begin 2002)	25	3.6%
428.42	Chron Cmbined Syst & Dias Heart Failr (Begin 2002)	22	3.2%
398.91	Rheumatic Heart Failure	14	2.0%
428.40	Unspec Cmbined Syst & Dias Heart Failr (Begin 2002)	6	0.9%

Note: # patients with at least one of these 16 ICD-9-CM HF-related codes = 686.