

Congenital Heart Disease

Geriatric Congenital Heart Disease

Burden of Disease and Predictors of Mortality

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Objectives	The study sought to measure the prevalence, disease burden, and determinants of mortality in geriatric adults with congenital heart disease (ACHD).
Background	The population of ACHD is increasing and aging. The geriatric ACHD population has yet to be characterized.
Methods	Population-based cohort study using the Quebec Congenital Heart Disease Database of all patients with congenital heart disease coming into contact with the Quebec healthcare system between 1983 and 2005. Subjects with specific diagnoses of congenital heart disease and age 65 years at time of entry into the cohort were followed for up to 15 years. The primary outcome was all-cause mortality.
Results	The geriatric ACHD cohort consisted of 3,239 patients. From 1990 to 2005, the prevalence of ACHD in older adults remained constant from 3.8 to 3.7 per 1,000 indexed to the general population (prevalence odds ratio: 0.98; 95% confidence interval [CI]: 0.93 to 1.03). The age-stratified population prevalence of ACHD was similar in older and younger adults. The most common types of congenital heart disease lesions in older adults were shunt lesions (60%), followed by valvular lesions (37%) and severe congenital heart lesions (3%). Type of ACHD and ACHD-related complications had a minor impact on mortality, which was predominantly driven by acquired comorbid conditions. The most powerful predictors of mortality in the Cox proportional hazards model were: dementia (hazard ratio [HR]: 3.24; 95% CI: 1.53 to 6.85), gastrointestinal bleed (HR: 2.79; 95% CI: 1.66 to 4.69), and chronic kidney disease (HR: 2.50; 95% CI: 1.72 to 3.65).
Conclusions	The prevalence of geriatric ACHD is substantial, although severe lesions remain uncommon. ACHD patients that live long enough acquire general medical comorbidities, which are the pre-eminent determinants of their mortality. (J Am Coll Cardiol 2011;58:1509–15) © 2011 by the American College of Cardiology Foundation

The prevalence of adults with congenital heart disease (ACHD) is rising in the general population (1), and healthcare utilization rates are high compared with non-ACHD adults (2). Not surprisingly, the number of older adults above age 65 years with ACHD is also rising, with admission rates increasing by 30% for this group of patients over the past 10 years (3). Furthermore, mortality rates in congenital heart disease have shifted away

from the young and towards adults, with a steady increase in age at death (4).

In contrast to the younger ACHD population that has attracted research interest over the last 2 decades, the geriatric ACHD population has yet to be characterized. Extrapolation of studies on younger patients is not appropriate. First, older adults with ACHD acquire comorbid conditions that may play a significant role in their health status. Second, geriatric ACHD patients may have inherently superior resiliency, milder disease, or balanced physiology in contrast to those not surviving to an advanced age (5). Therefore, older adults with ACHD represent a distinct population for which focused studies are needed. Global disease burden is driving costs and outcomes in the United States and Canada, thus becoming relevant to policymakers and ACHD specialists. This is likely to have important implications in planning for future allocation of resources in this growing population.

Our objective was to analyze trends in numbers of older adults with ACHD over the past 15 years and to identify the determinants of mortality. In particular, the impact on

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**Abbreviations
and Acronyms****ACHD** = adult congenital heart disease**CI** = confidence interval**HR** = hazard ratio**ICD-9** = International Classification of Diseases-9th edition

mortality of ACHD lesion severity, ACHD-related complications, and acquired comorbid conditions was measured in a large population-based cohort of older adults with ACHD.

Methods**Study design and data sources.**

A population-based cohort of older adults with ACHD was extracted from the Quebec Congenital Heart Disease Database (1,4). The Quebec Congenital Heart Disease Database consists of all ACHD patients coming into contact with the healthcare system in Quebec between 1983 and 2005. The healthcare system in Quebec is a single-payer system offering comprehensive and universal access to healthcare services. Utilization of these healthcare services is tied to a unique patient identifier number that was used to cross-link the physician's services and claims database (Régie de l'Assurance Maladie du Québec), the hospital discharge database (Med-Echo), and the Quebec death registry. To protect patient confidentiality, data were transmitted with encrypted identifiers. Permission was obtained from the McGill University Health Center ethics board and government agencies responsible for privacy of access to information.

Study population. Inclusion criteria were age 65 years at any point during the time period of 1990 to 2005, and confirmed diagnosis of ACHD. The diagnosis of ACHD was ascertained based on a hierarchical diagnostic algorithm that has been validated in prior studies (1,4) and adapted to the current study to exclude elderly patients who had an isolated diagnostic code for ACHD without confirmation by a cardiovascular specialist or ACHD-related hospitalizations or surgeries. This algorithm contains 24 International Classification of Diseases-9th edition (ICD-9), codes for ACHD diagnosis and surgery, weighted according to the frequency of the code (multiple vs. isolated) and the physician making it (cardiovascular specialist vs. other). Uncertain diagnoses were hand-checked and audited by 2 independent observers and either excluded, validated, or reclassified. Patients with nonspecific diagnostic codes for ACHD were excluded (unspecified anomalies of the heart [ICD-9 codes 7468–7469], unspecified anomalies of the aorta [ICD-9 code 7472], unspecified anomalies of the circulation [ICD-9 code 7479]).

Predictor variables and covariates. ACHD lesions were grouped into 3 categories: severe lesions (tetralogy of Fallot, transposition of the great arteries, endocardial cushion defect, Ebstein anomaly, truncus arteriosus, univentricular heart, hypoplastic left heart syndrome), shunt lesions (atrial septal defect, ventricular septal defect, patent ductus arteriosus), and congenital regurgitant or stenotic lesions termed valvular lesions (congenital aortic stenosis, congenital aortic regurgitation, congenital mitral stenosis, congenital mitral

regurgitation, congenital tricuspid valve disease, anomalies of the pulmonary valve, aortic coarctation) (1,4). Severe lesions reflect lesions with the highest probability of being associated with cyanosis or requiring surgical intervention in early life (1,4).

ACHD-related complications and acquired comorbid conditions were analyzed (Table 1). Diagnostic codes for comorbid conditions were available from 1983 onward. For patients entering the cohort in the first year (1990), there were 7 years of prior records for ascertainment of comorbid conditions (1983 to 1990). For patients entering the cohort in later years, there were more prior records, although a fixed time period of 7 years prior to entry in the cohort was used in order to allow all patients to have equal ascertainment time. Recognizing that there may be overlap between what would be considered an ACHD complication or an acquired comorbid condition (e.g., heart failure), variables were entered individually and not grouped in the analytical model.

Outcome and censoring. The primary endpoint was all-cause mortality. Vital status was determined in the physician's services and claims database and the Quebec death registry. The Quebec civil code (section III, articles 122–128) mandates that all deaths be ascertained by law and forwarded to the physician's services and claims database. In a study of patients with prior myocardial infarction conducted in the medical claims database of Quebec, vital status was determined in 99.75% of subjects (6). Ascertainment of mortality in the Quebec Congenital Heart Disease Database has been previously published by our group (4). Patients were followed for up to 15 years (up to 80 years of age), from their respective entry into the study cohort until death or end of follow-up.

Statistical analysis. Baseline characteristics of patients with severe, shunt, and valvular lesions were compared with the chi-square test. Prevalence was measured by dividing the number of patients with a confirmed diagnosis of ACHD in the provincial ACHD database during the calendar years of 1990, 1995, 2000, and 2005 by the midyear general population estimates matched for age group and calendar year from the provincial census (Statistics Canada [7]). Prevalence was measured for the adult age group (age 18 to 64 years) and the elderly age group (age 65 years and above). The change in prevalence between the first calendar year (1990) and the last (2005) was compared using the prevalence odds ratio with an associated 95% confidence interval. The effect of predictors and covariates on all-cause mortality was measured using a Cox proportional hazards model, where time zero was age 65 years and the time axis was age (range 65 to 80 years). The proportionality of hazards was verified graphically by the log-log survival plot. The list of predictors and covariates were pre-specified and determined based on a priori knowledge. To account for possible time-dependent effects of ACHD-related complications and acquired comorbid conditions, a sensitivity analysis using a nested case-control model was performed. In such a

Table 1 Patient Characteristics Grouped By ACHD Lesion Type

	All Lesions (n = 3,239)	Shunts (n = 1,946)	Severe (n = 82)	Valvular (n = 1,211)	p Value
Male	1,440 (44)	766 (39)	32 (39)	642 (53)	<0.0001
Atrial fibrillation	681 (21)	433 (22)	26 (32)	222 (18)	0.002
Ventricular arrhythmia	99 (3)	65 (3)	4 (5)	30 (2)	0.24
Sick sinus syndrome	145 (4)	89 (5)	5 (6)	51 (4)	0.69
Heart block	139 (4)	74 (4)	10 (12)	55 (5)	0.001
Endocarditis	31 (1)	11 (1)	0 (0)	20 (2)	0.006
Pulmonary hypertension	186 (6)	115 (6)	7 (9)	60 (5)	0.006
Erythrocytosis	61 (2)	37 (2)	1 (1)	23 (2)	0.90
Epilepsy	61 (2)	34 (2)	0 (0)	27 (2)	0.28
Diabetes	527 (16)	318 (16)	11 (13)	198 (16)	0.78
Dyslipidemia	875 (27)	480 (25)	27 (33)	368 (30)	0.001
Hypertension	1,534 (47)	914 (47)	38 (46)	582 (48)	0.82
Coronary artery disease	339 (10)	206 (11)	13 (16)	120 (10)	0.23
Myocardial infarction	216 (7)	141 (7)	9 (11)	66 (5)	0.04
Heart failure	667 (21)	392 (20)	22 (27)	253 (21)	0.32
Stroke/TIA	330 (10)	201 (10)	3 (4)	126 (10)	0.14
Chronic kidney disease	75 (2)	41 (2)	1 (1)	33 (3)	0.43
Chronic liver disease	61 (2)	29 (1)	2 (2)	30 (2)	0.13
COPD	254 (8)	158 (8)	5 (6)	91 (8)	0.69
Pneumonia	93 (3)	51 (3)	4 (5)	38 (3)	0.38
Thyroid disease	431 (13)	269 (14)	13 (16)	149 (12)	0.37
Cancer	542 (17)	317 (16)	9 (11)	216 (18)	0.19
Deep vein thrombosis	38 (1)	28 (1)	2 (2)	8 (1)	0.08
Gastrointestinal bleed	33 (1)	19 (1)	1 (1)	13 (1)	0.95
Depression	212 (7)	115 (6)	9 (11)	88 (7)	0.08
Dementia	20 (1)	15 (1)	0 (0)	5 (0.4)	0.35

Values are n (%).

COPD = chronic obstructive pulmonary disease; TIA = transient ischemic attack.

time-dependent model, the presence or absence of covariates is ascertained within a fixed time period before events (death or selection as a control) rather than before entry in the cohort. To account for potential misclassification due to errors in grouping lesions into 3 exclusive categories, a sensitivity analysis using 3 individual lesions representative of each category was performed. These representative individual lesions were tetralogy of Fallot (severe), atrial septal defect (shunt), and congenital aortic stenosis (valvular). Analyses were performed with the SAS statistical software package (version 8.02, Cary, North Carolina).

Results

Prevalence of geriatric ACHD. The Quebec Congenital Heart Disease Database consisted of 71,467 patients, of which 44,865 were adults during the study period. After excluding nonelderly patients and those with nonspecific ACHD diagnoses, the geriatric ACHD cohort consisted of 3,239 older adults age 65 years at any point between 1990 and 2005. The flow diagram depicting the derivation of the study cohort is shown in Figure 1. The prevalence of ACHD in the geriatric population was found to be 3.8 per 1,000 in 1990 and 3.7 per 1,000 in 2005, representing a constant prevalence (prevalence odds ratio: 0.98; 95% confidence interval [CI]: 0.93 to 1.03). In comparison, the

prevalence of ACHD in the nongeriatric adult population (age 18 to 64 years) was found to be 3.1 per 1,000 in 1990 and 4.2 per 1,000 in 2005, representing a 37% increase in odds (prevalence odds ratio: 1.37; 95% CI: 1.34 to 1.40). Trends in prevalence are shown in Figure 2.

Baseline characteristics of study population. Patient characteristics grouped by type of ACHD lesion are shown in Table 1. The most frequent type of ACHD lesion in older adults was shunts (60%), followed by valvular (37%) and severe lesions (3%). Severe ACHD lesions were rare, and mostly consisted of conotruncal abnormalities and endocardial cushion defects; whereas nonsevere lesions mostly consisted of atrial septal defects, ventricular septal defects, congenital aortic stenosis, and congenital mitral insufficiency. Covariates were generally well balanced between groups, although patients with severe lesions were more likely to have atrial fibrillation ($p = 0.002$), heart block ($p = 0.001$), and pulmonary hypertension ($p = 0.006$). Patients with valvular lesions were more likely to be male ($p < 0.0001$) and have had endocarditis in the past ($p = 0.006$).

Mortality. During a median follow-up of 6.9 years (Q1, Q3: 3.3, 10.7 years), 630 deaths were observed. The Kaplan-Meier analysis revealed a projected 15-year survival rate of 56.2%. Survival was similar regardless of ACHD

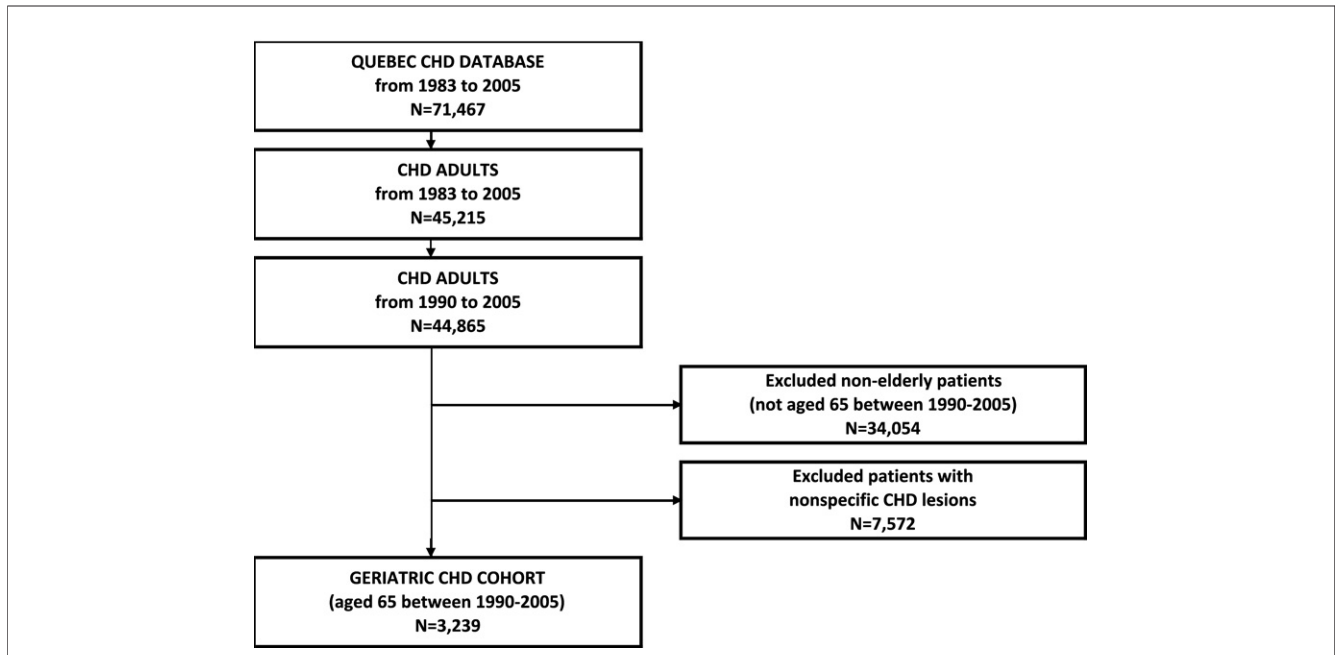


Figure 1 Flow Diagram for Derivation of Study Cohort

The geriatric CHD cohort included patients with a confirmed diagnosis of congenital heart disease (CHD) who were 65 years of age at any point between 1990 and 2005.

lesion type (54.9% in shunts, 57.7% in valvular lesions, and 66.8% in severe lesions, log-rank $p = 0.09$) (Fig. 3). After adjusting for potential confounders, ACHD lesion type remained a nonsignificant predictor of survival. Unadjusted and adjusted predictors of all-cause mortality are shown in Table 2.

Nine independent predictors of increased all-cause mortality were identified, all of which had been classified

as acquired cardiovascular and general medical conditions. In descending order of magnitude, these were dementia (hazard ratio [HR]: 3.24; 95% CI: 1.53 to 6.85), gastrointestinal bleed (HR: 2.79; 95% CI: 1.66 to 4.69), chronic kidney disease (HR: 2.50; 95% CI: 1.72 to 3.65), heart failure (HR: 1.98; 95% CI: 1.65 to 2.38), diabetes mellitus (HR: 1.76; 95% CI: 1.45 to 2.13), chronic obstructive pulmonary disease (HR: 1.67; 95% CI: 1.31 to 2.12), cancer (HR: 1.43; 95% CI: 1.17 to

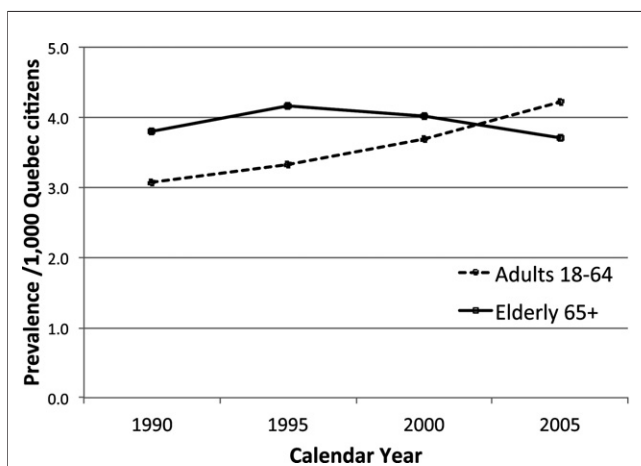


Figure 2 Trends in Prevalence of ACHD in Different Age Groups

In 2005, the prevalence of ACHD was 3.7 per 1,000 in the elderly and 4.2 per 1,000 in nonelderly adults. Using 1990 as a reference, the prevalence remained constant in the elderly (prevalence odds ratio: 0.98; 95% confidence interval [CI]: 0.93 to 1.03), whereas it increased in nonelderly adults (prevalence odds ratio: 1.37; 95% CI: 1.34 to 1.40).

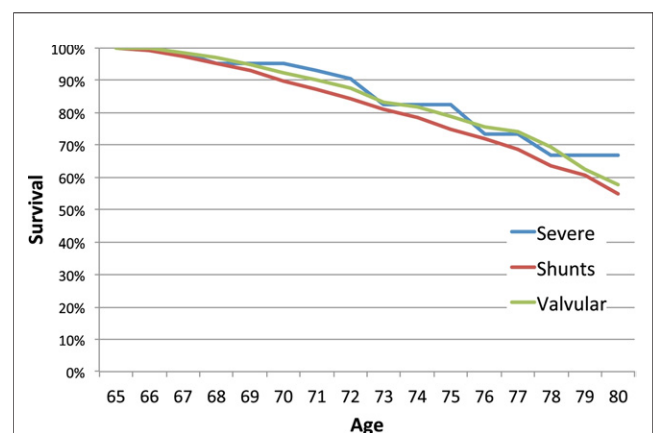


Figure 3 Kaplan-Meier Curves for Unadjusted All-Cause Mortality

The 15-year survival was 56%; 67% for patients with severe lesions, 55% for patients with shunts, and 58% for patients with valvular lesions (log-rank $p=0.09$).

1.76), myocardial infarction (HR: 1.40; 95% CI: 1.07 to 1.83), and male sex (HR: 1.33; 95% CI: 1.13 to 1.55).

Sensitivity analysis. In a sensitivity analysis comparing 3 representative individual ACHD lesions (tetralogy of Fallot, atrial septal defect, and congenital aortic stenosis), ACHD lesion type remained a nonsignificant predictor of all-cause mortality, and the Cox proportional hazards model yielded similar results. A nested case-control model, performed to better account for the time-dependency of covariates, also yielded similar results.

Discussion

To our knowledge, this is the first study to specifically address the population of older adults with ACHD. We present data on the prevalence, disease burden, and predictors of mortality in a large population-based geriatric ACHD cohort. The contemporary prevalence of geriatric ACHD was found to be 3.7 per 1,000 older adults, which is slightly lower than that in nongeriatric patients, but high enough to document that the absolute number of older adults with ACHD is substantial. ACHD patients survived long enough to acquire a significant disease burden from general medical conditions, which were the main determinants of their mortality. Dementia, chronic kidney disease, and gastrointestinal bleeding were the most powerful predictors of all-cause mortality over a 15-year period.

The correlates of survival in patients with congenital heart defects vary with age. In infants and children, lesion severity and surgical results have been shown to be the primary determinants of mortality (8,9). In young and middle-aged adults, ACHD-related complications begin to play a larger role in determining outcomes; especially cyanosis, pulmonary hypertension, heart failure, and arrhythmias (10–15). In older adults, our findings suggest that acquired medical conditions have the greatest impact on mortality. These acquired medical conditions mirror the most common causes of death in the general population (16), with the addition of gastrointestinal bleeding, which is increasingly recognized as a powerful predictor of mortality in patients taking antiplatelet and anticoagulant therapies (17,18) (a substantial proportion of patients in this cohort had atherosclerotic disease and atrial fibrillation). ACHD lesion type was not an independent predictor of mortality after adjusting for confounders. ACHD-related complications were similarly not independent predictors of mortality (with the exception of heart failure, which may or may not be ACHD related).

There are at least 3 reasons for which acquired comorbid conditions and not ACHD lesion type or complications had an impact on mortality in older patients. First, the prevalence of severe lesions and/or severe complications in older adults (and consequently, the statistical power in this subset of patients) was relatively low and remained constant throughout the study period, in part as a result of the lack of effective therapeutic options in the 1920s and 1930s, when

most of this study cohort was born. With the evolution of congenital cardiac surgery, as well as sensitive diagnostic testing and greater general life expectancy, many more patients with severe ACHD (including those with very complex lesions) are expected to survive to an advanced age. This phenomenon has been observed in the nongeriatric adult population wherein the prevalence of adults with severe ACHD is rising faster than the prevalence of children with severe ACHD (prevalence odds ratio: 1.85 vs. 1.22) (1). This should translate into an increasing prevalence of older adults with severe ACHD, and possibly an increasing proportion of older adults dying from ACHD-related conditions.

Second, the number of older adults with atrial septal defects and valvular lesions was high, representing the majority of our cohort. Prior studies have shown that the most common causes of death in patients with atrial septal defects are noncardiac (19), whereas in patients with repaired bicuspid aortic valves, they tend to be perioperative or related to coronary artery disease (20). Even in an unselected population of young adults with ACHD, one-third of deaths were noncardiac in origin (21). The proportion of deaths attributable to ACHD is much higher in studies focusing on complex and relatively rare pathophysiologies such as Fontan or Eisenmenger, which may not be generalizable to ACHD patients at large. Within the small group of patients with severe lesions, the majority consisted of tetralogy of Fallot and endocardial cushion defects, for which there are well-recognized balanced and partial variants, respectively. Although higher than nonsevere lesions, the modest rates of heart failure (27%) and pulmonary hypertension (9%) suggest that many of the older adults with severe lesions in this study had either balanced or minor variants allowing them to naturally survive to an advanced age, or had operated lesions with favorable results.

Third, the number of older ACHD patients with documented cardiovascular disease and risk factors was substantial (7% prevalence of myocardial infarction as compared with an age-matched prevalence of 5% in Canada [22]), leading to a large burden of atherosclerotic disease. The burden of atherosclerotic disease in older adults with ACHD is clearly driven by age but may also be associated with ACHD. Billet et al. (23) found that adults with ACHD were more likely to have hypertension, diabetes, stroke, and chronic kidney disease (a coronary artery disease risk equivalent) than age-matched controls without ACHD. Moons et al. (24) found that at least 80% of adults with ACHD had at least 1 coronary artery disease risk factor, with the rates of hypertension and obesity being more common than the general population. The mean age in both of these studies was only 26 to 28 years, suggesting that ACHD patients may harbor risk factors for many years. It is, therefore, not surprising that by the time they reach 65 years of age, there would be a large burden of atherosclerotic disease.

Study limitations. Our study findings need to be interpreted in light of our study design since administrative databases have certain inherent limitations. Although the Quebec Congenital Heart Disease Database contained our variables of interest, additional sources of data may have

Table 2 Predictors of All-Cause Mortality

	Deaths (n = 630)	Survivors (n = 2,609)	Univariate HR (95% CI)	Adjusted HR (95% CI)
ACHD lesion				
Severe	12 (2)	70 (3)	0.88 (0.50–1.56)	
Shunts	397 (63)	1,549 (59)	1.19 (1.02–1.40)	
Valvular	221 (35)	990 (38)	0.84 (0.72–0.99)	
Male	323 (51)	1,117 (43)	1.41 (1.20–1.65)	1.33 (1.13–1.55)
Atrial fibrillation	156 (25)	52 (20)	1.72 (1.43–2.06)	
Ventricular arrhythmia	28 (4)	71 (3)	2.19 (1.50–3.20)	
Sick sinus syndrome	27 (4)	118 (5)	1.37 (0.93–2.02)	
Heart block	33 (5)	106 (4)	1.72 (1.21–2.44)	
Endocarditis	7 (1)	24 (1)	1.82 (0.87–3.84)	
Pulmonary hypertension	45 (7)	141 (5)	2.19 (1.61–2.97)	
Erythrocytosis	18 (3)	43 (2)	2.27 (1.42–3.64)	
Epilepsy	17 (3)	44 (2)	1.74 (1.08–2.82)	
Diabetes	142 (23)	385 (15)	2.11 (1.75–2.54)	1.76 (1.45–2.13)
Dyslipidemia	112 (18)	763 (29)	0.83 (0.67–1.02)	0.74 (0.60–0.91)
Hypertension	305 (48)	1,229 (47)	1.36 (1.16–1.59)	
Coronary artery disease	97 (15)	242 (9)	2.08 (1.67–2.58)	
Myocardial infarction	70 (11)	146 (6)	2.34 (1.82–3.00)	1.40 (1.07–1.83)
Heart failure	196 (31)	471 (18)	2.54 (2.15–3.02)	1.98 (1.65–2.38)
Stroke/TIA	72 (11)	258 (10)	1.50 (1.17–1.92)	
Chronic kidney disease	32 (5)	43 (2)	4.21 (2.95–6.02)	2.50 (1.72–3.65)
Chronic liver disease	17 (3)	44 (2)	2.16 (1.33–3.50)	
COPD	79 (13)	175 (7)	2.05 (1.62–2.60)	1.67 (1.31–2.12)
Pneumonia	28 (4)	65 (2)	2.23 (1.53–3.26)	
Thyroid disease	66 (10)	365 (14)	0.97 (0.75–1.25)	
Cancer	118 (19)	424 (16)	1.59 (1.30–1.94)	1.43 (1.17–1.76)
Deep vein thrombosis	6 (1)	32 (1)	1.39 (0.62–3.11)	
Gastrointestinal bleed	15 (2)	18 (1)	4.26 (2.55–7.12)	2.79 (1.66–4.69)
Depression	35 (6)	177 (7)	0.82 (0.58–1.15)	
Dementia	7 (1)	13 (0.5)	2.62 (1.24–5.52)	3.24 (1.53–6.85)

Values are n (%).

CI = confidence interval; HR = hazard ratio; other abbreviations as in Table 1.

been useful for verification of ACHD diagnoses. Several measures were employed to minimize misclassification of ACHD diagnoses. A previously validated multilevel hierarchical algorithm was used, additional hand-checking and auditing was performed, and nonspecific ACHD codes were rejected. Furthermore, sensitivity analysis for each category of ACHD and for individual ACHD lesions yielded similar results. Patients not using the Quebec healthcare system between 1983 and 2005 were not included in the cohort. This may have led to an underestimation of the prevalence of older adults with ACHD, especially those with mild, clinically silent disease. Inclusion of these patients may have further inflated the impact of acquired comorbid conditions on mortality since patients with mild ACHD would be expected to die from ACHD-unrelated causes. Therefore, our results provide a conservative estimate of both prevalence and predictors of mortality.

Prior studies addressing the burden of ACHD have been limited by short-term follow-up and emphasis on complex lesions being cared for in highly specialized centers (23). The present study was population based, encompassing simple and complex lesions, tertiary and nontertiary care

settings, and long-term follow-up. The implications of our findings underscore the complex needs of patients with ACHD (25), and suggests that there will be a need for resources to care for this growing group of older adults with ACHD: ACHD specialists to care for the small but increasing number of severe lesions and complications, general cardiologists to care for the burden of atherosclerotic disease, and general practitioners and internists to care for the multitude of acquired comorbid conditions that these patients possess.

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