Specialized Adult Congenital Heart Disease Care
The Impact of Policy on Mortality

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Background—Clinical guidelines recommend specialized care for adult congenital heart disease (ACHD) patients. In reality, few patients receive such dedicated care. We sought to examine the impact of specialized care on ACHD patient mortality.

Methods and Results—We examined referral rates to specialized ACHD centers and ACHD patient mortality rates between 1990 and 2005 in the population-based Quebec Congenital Heart Disease database (n=71 467). This period covers several years before and after the publication of guidelines endorsing specialized care for ACHD patients. A time-series design, based on Joinpoint and Poisson regression analyses, was used to assess the changes in annual referral and patient mortality rates. The association between specialized ACHD care and all-cause mortality was assessed in both case–control and cohort studies. The time-series analysis demonstrated a significant increase in referral rates to specialized ACHD centers in 1997 (rate ratio, +7.4%; 95% confidence interval [CI], +6.6% to +8.2%). In parallel, a significant reduction in expected ACHD patient mortality was observed after year 2000 (rate ratio, −5.0%;95% CI, −10.8% to −0.8%). In exploratory post hoc cohort and case–control analyses, specialized ACHD care was independently associated with reduced mortality (hazard ratio, 0.78; 95% CI, 0.65–0.94) and a reduced odds of death (adjusted odds ratio, 0.82; 95% CI, 0.08–0.97), respectively. This effect was predominantly driven by patients with severe congenital heart disease (hazard ratio, 0.38; 95% CI, 0.22–0.67).

Conclusions—A significant increase in referrals to specialized ACHD centers followed the introduction of the clinical guidelines. Moreover, referral to specialized ACHD care was independently associated with a significant mortality reduction. Our findings support a model of specialized care for all ACHD patients. **(*Circulation*. 2014;129:1804-1812.)**

Key Words: congenital ■ heart diseases ■ mortality ■ referral and consultation ■ treatment outcome

Despite the growing need to bring quality to congenital heart disease (CHD) care,[1] there are no data demonstrating that specialized adult CHD (ACHD) care can improve outcomes. An estimated 1 million adults were thought to be living with CHD in the United States in 2000, and this number is expected to increase.[2–5] Traditionally, the life expectancy in many infants with severe CHD was limited to months; however, advances in medical and surgical care have borne remarkable improvements in survival[6]: the median age of those alive with severe CHD has increased from 11 years in 1985 to 17 years in 2000, and the majority of children born with CHD now reach adulthood.[7,8] Accordingly, the number of adults with CHD now exceeds the number of children with CHD, and, as these patients mature into adulthood, many develop late cardiac complications.[5,7,9,10] The unique needs of this ACHD population center on the care of life-long comorbidities and have brought about the emergence of ACHD as a new area of specialized cardiovascular care.[11]

Editorial see p 1795
Clinical Perspective on p 1812

Building on the work of the Canadian Adult Congenial Heart Network established in 1992, the Canadian Cardiac Society (CCS) convened a panel of international experts for an ACHD consensus conference in 1996. Foremost among the proposals arising from this forum was a recommendation that all ACHD patients be referred to specialized ACHD centers for ongoing care.[12] Clinical guidelines to this effect were presented at the 1996 CCS conference, published in 1998, and were followed by similar proposals emerging from the 32nd Bethesda Conference in 2001.[5,12] Despite these recommendations and more than a decade later, less than one-third

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of eligible patients actively receive specialized ACHD care in Canada, where health insurance is universal.[13] In the United States, this proportion is expected to be significantly less, and care gaps and dispersion have been documented.[14,15] Despite specialized care for all ACHD patients being espoused by societal guidelines,[9,10] there is little evidence demonstrating that this model of care improves clinical outcomes.

Hence, we sought to evaluate the impact of specialized ACHD care on mortality to inform policy. Our objectives were: (1) to identify changes in the time trends of referral to specialized ACHD referral centers in Quebec over a 15-year period that included the 1998 Canadian Consensus Guideline recommendations[12]; (2) to contrast the trends in referral to specialized ACHD centers with those in ACHD mortality; and (3) to assess whether ACHD referral center care was associated with reduced mortality.

Methods

Data Sources

In Quebec, Canada, access to health care is universal and each resident is assigned a unique healthcare identifier that is recorded every time the individual accesses the healthcare system. This information is recorded in province-wide administrative databases and allows for the reconstruction of an individual's medical history in terms of diagnoses, hospitalizations, and medical services received. We merged the provincial medical claims (Regie de l'assurance Maladie du Quebec: 1983–2005), the provincial hospital discharge database (Med-Echo: 1987–2005), and the provincial death registry to develop the Québec Congenital Heart Disease Database.[7] This database contains comprehensive longitudinal, demographic, diagnostic, and therapeutic records of all CHD patient encounters with the provincial healthcare system between January 1, 1983 and December 31, 2005.[16] CHD patients were identified if they had at least 1 diagnostic code for CHD or a CHD-specific surgical procedure. All CHD patients were assigned 1 or 2 CHD diagnoses by using a previously described and validated hierarchical algorithm.[7] Manual audits of randomly generated patient samples were performed by 2 CHD specialists (A.J.M. and A.S.M.) to detect and correct discrepancies between the data sources and algorithms.

Study Design

A time-series design was used to assess temporal changes in specialized ACHD center referral patterns and ACHD mortality. Yearly referral and mortality rates were calculated by using the total number of ACHD patients alive in that year as the denominator, and either the number of patients referred to a specialized ACHD center or the number of patients that died in that year as the numerator.

Given that the time-series analysis showed a decrease in ACHD patient mortality concurrent with the introduction of the CCS guidelines, we further explored the impact of ACHD referral center care on mortality in 2 separate post hoc analyses. First, we performed a case–control study in which patients that died during the observation period (1988–2005) were risk set matched 1:2 by calendar year of death to patients from the underlying longitudinal cohort that were alive at the time that the patient died. Potential predictors of death, such as age, severe CHD, comorbidity, and patient use of ACHD referral center services, were measured for each matched case–control set in the 3 years before the date of death for the case. For descriptive purposes, administrative records for all deaths were reviewed, and the most probable cause of death was determined for each case.

Second, we performed a cohort study with patient follow-up starting at the time of the patient's first outpatient visit to a cardiologist after January 1, 1998. This date corresponds to the publication of the CCS guidelines that recommended referral to specialized ACHD centers.[12] Patients were classified as receiving care in an ACHD referral center or in a nonreferral center and were then followed until death or administrative censoring on December 31, 2010. Potential confounders were measured in the 3 years before the index visit.

Study Population

The CHD Quebec database includes 71 467 patients who were alive between 1983 and 2005. In this longitudinal cohort, we identified separate study populations for the analysis based on the time-series design and for those based on the case–control and cohort designs. For the time-series analysis, we first identified, for each calendar year, all ACHD patients between 18 and 65 years of age that were alive on January 1 of that year. To ensure the comparability of visit and death rates across years, the study population in each year was restricted to patients identified with CHD in the previous 7 years, because this was the maximal duration of follow-up available for patients in 1990 (Figure 1). In the case–control analysis, the study population included 2092 ACHD death cases and 4184 risk-set matched controls. The cohort study consisted of 9458 patients aged 18 to 65 years at the time of their index outpatient visit to cardiologist after January 1, 1998.

Definitions and Measurements

Specialized ACHD referral centers were defined by the use of the following criteria: (1) the expertise criterion, mandating that each center uses at least 1 imaging or interventional cardiologist and at least 2 CHD surgeons with advanced training and experience with ACHD

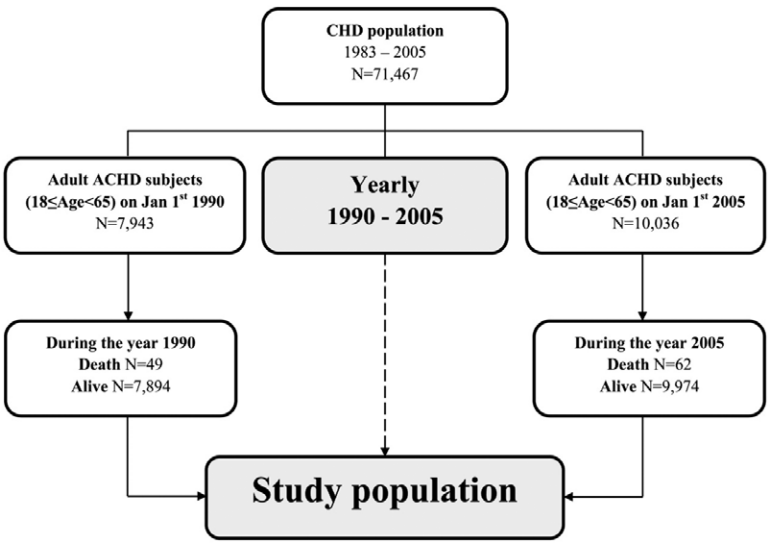

Figure 1. Derivation of the study population. In each calendar year, the number of adult congenital heart disease (ACHD) patients (18–65 years of age) alive on January 1 was assessed. In each year (1990–2005), we measured patient referral patterns and deaths. The study population in each year was restricted to those patients identified with congenital heart disease (CHD) in the previous 7 years.

patients[9,17,18]; (2) the patient volume criterion, requiring that ≥520 patients per year be seen at the center[19] and that the center was listed as a designated center in the 1998 consensus document.[12] In Quebec, 3 ACHD referral centers met these criteria.

Patient referral to specialized ACHD centers was defined differently for each study design. In the time-series analysis, patients were defined as referred to a specialized ACHD center if they had a cardiology consultation at a designated ACHD center in the year when the cross-section was taken. In the case–control study, patients were classified in 1 of the following 4 categories based on their pattern of cardiology outpatient visits between database entry and the risk-set year corresponding to each case–control set: (1) ACHD referral center care; (2) nonreferral center care; (3) mixed care (both ACHD referral and nonreferral center care); and (4) no outpatient cardiology care (used as reference category in regression models). In the cohort study, patients were defined as referred to a specialized ACHD center if their first cardiology outpatient visit after January 1, 1998 occurred at a designated ACHD referral center. Visits to ACHD referral or non referral centers were only considered if they were with a cardiologist. Noncardiology visits were not counted.

Mortality was measured yearly per 1000 ACHD population at risk by the use of date of death records. The cause of death was ascertained by using death records according to the *International Classification of Diseases, Ninth Revision* diagnostic code. CHD lesions were grouped as severe (endocardial cushion defect, tetralogy of Fallot, truncus arteriosus, transposition of the great vessels, and hypoplastic left heart syndrome/univentricular heart), and other.[7] Several a priori selected cardiac comorbid conditions were measured among ACHD patients alive in 2005, in the case–control and cohort populations, as follows: congestive heart failure,[20] pulmonary hypertension,[21] atrial fibrillation,[16] and stroke.[22] These comorbid illnesses were identified in the Quebec CHD database based on *International Classification of Diseases, Ninth Revision* diagnostic codes in the 3 years before the index time for the study populations for the case–control and cohort studies. The presence of genetic syndromes associated with CHD were identified in the database by using *International Classification of Diseases, Ninth Revision* diagnostic codes, and included Down syndrome, DiGeorge syndrome, and Turner syndrome.

Our database contains residential 3-digit postal codes. With the use of this information, income data aggregated for the 3-digit postal code and geographic area of residence (rural versus urban) was obtained by linking the postal-code conversion file to the 2006 Canadian Census data on income, earnings, housing, and shelter costs.[23,24] These 2 files were linked by using a combination of the census metropolitan area code and the census tract name.

Statistical Analysis

Descriptive statistics include proportions and medians (interquartile ranges), as appropriate. Patients were compared across different groups with χ^2 and Wilcoxon tests.

To assess changes in yearly rates of referral to specialized ACHD centers and ACHD mortality, time-series analyses were performed based on Joinpoint and Poisson regression models. Specifically, we used Joinpoint regression to identify for each outcome the time (change point) when ≥1 significant changes occurred in the trends.[25,26] Poisson regression models were used to estimate the trends in the 2 outcomes before and after the corresponding point of change estimated in the Joinpoint analyses. The Poisson models use the yearly rates of referral to specialized ACHD centers and ACHD mortality as the respective dependent variables and include an interaction with a binary indicator of the period before and after the change point to estimate the separate rate ratios (RRs) for the linear effect of calendar year in the 2 periods (before and after). A statistically significant interaction ($P \leq 0.05$) was interpreted as indicating that the change in trend was systematic and not a result of chance.

In the case–control study, the impact of ACHD referral center care on mortality was analyzed by using a multivariable conditional logistic regression model, which adjusted for the following a priori selected potential confounders: age, sex, CHD lesion type, cardiac comorbidity (congestive heart failure, atrial fibrillation, pulmonary hypertension, stroke). With the use of this model, the effect of ACHD referral center care on mortality was estimated through the adjusted odds ratios (ORs) and 95% confidence intervals (CIs).

In the cohort study, adjusted Kaplan-Meier curves were developed by the use of inverse probability of referral weighting,[27] based on the propensity score, to compare mortality in patients referred to specialized ACHD centers with the mortality of those never referred. These curves were compared with the log-rank test. Furthermore, the effect of ACHD referral center exposure on the time to death was analyzed by using multivariable Cox regression, adjusted for the same a priori selected potential confounders, associated genetic syndromes, geographic location, and average posttaxation income. From this model, we report adjusted hazard ratios (HRs), with 95% CIs.

All analyses were performed by using SAS software (SAS institute Inc SAS/STAT Software: changes and enhancement through Release 9.2, Cary, NC) and the Joinpoint Regression Program Release 3.4.3 from the National Cancer Institute.[28]

Results

Description of ACHD Population

In each year between 1990 and 2005, there were between 7000 and 11 000 patients who satisfied the study inclusion criteria: 7943 in l990, 10 195 in 1998, and 10 036 in 2005. In Table 1, we illustrate the differences in outpatient visits and clinical characteristics for the most recent cross-section of the study population (year 2005). Among 10 036 ACHD patients alive in 2005, 25% (2477) had an outpatient visit to a cardiologist and 75% (7559) did not. ACHD patients without cardiology consultations were younger and less likely to be males, have severe CHD, or have comorbid conditions than those with consultations. Among the 2477 patients with a cardiology consultation in 2005, visits occurred in an ACHD referral center in 70%, in a nonreferral center in 27%, and in both centers (mixed care) in 4.1%. In comparison with patients receiving nonreferral center care, those with ACHD referral center care were younger and more likely to have severe CHD. The highest burden of comorbid cardiac disease occurred in patients receiving mixed care. Associated genetic syndromes were observed in 3% of the overall population with equivalent distribution among groups. Patients receiving nonreferral center care had marginally lower average posttaxation income and were more likely to be rural dwellers than those referred for specialized care.

Trends in Referral to Specialized ACHD Centers and ACHD Mortality

The observed and estimated time trends for referral to specialized ACHD centers and ACHD mortality were comparable in the early 1990s (Figure 2). In the late 1990s, these trends diverged with a significant change point noted in 1997 for the observed rates of referral to specialized ACHD centers. Specifically, the slowly progressive trend of increasing referral to specialized ACHD centers before 1997 (RR, +1.4%) was followed by a more rapid increase of 7.4% per year after 1997 (RR, +7.4%; 95% CI, +6.6% to +8.2%, $P<0.0001$). In parallel, the slow rise in ACHD patient mortality noted before 2000 (RR, +2.0%) was reversed to a reduction of −5.0% per year after 2000 (RR, −5.0%; 95% CI, −10.8% to −0.8%, $P=0.04$). For both trends, a significant interaction was observed in the Poisson model between year and

Table 1. Patient Characteristics and ACHD Care Centers in 2005

	Referral Center Care (n=1708)	Nonreferral Center Care (n=667)	Mixed Care (n=102)	No Outpatient Cardiology Care (n=7559)	*P* Value
Age, y	37.3 (26.3–50.4)	47.3 (32.6–57.5)	47.8 (35.6–56.4)	34.0 (24.4–46.8)	<0.001
Female	842 (49.3)	316 (47)	47 (46)	4711 (62)	<0.001
CHD diagnosis					
Severe CHD	540 (31.6)	99 (14.8)	23 (22.6)	1094 (14.5)	<0.001
Shunts	533 (31.2)	265 (39.7)	43 (42.2)	3233 (42.8)	<0.001
Other	635 (37.2)	303 (45.4)	36 (35.3)	3232 (42.8)	<0.001
Comorbid condition					
CHF	557 (32.6)	231 (34.6)	44 (43.1)	928 (12.3)	<0.001
PH	194 (11.4)	81 (12.1)	20 (19.6)	322 (4.3)	<0.001
AF	525 (30.7)	196 (29.4)	51 (50.0)	574 (7.6)	<0.001
Stroke	154 (9.0)	93 (13.9)	18 (17.7)	381 (5.0)	<0.001
Associated genetic syndromes	55 (3.2)	20 (3.0)	2 (1.9)	208 (2.7)	0.573
Urban domicile	1482 (86.8)	523 (78.4)	86 (84.3)	6423 (85.0)	<0.001
Average net individual income (Canadian dollars)	27 755	27 085	27 660	27 627	0.018

Data represent median and interquartile range or number and percentage. *P* values represent the differences among the groups and were calculated by using χ^2 test or analysis of variance. AF indicates atrial fibrillation; CHD, congenital heart disease; CHF, congestive heart failure; and PH, pulmonary hypertension.

a binary indicator of the period before and after the change point identified by the Joinpoint regression model ($P<0.0001$ for rates of referral to specialized ACHD centers, and $P<0.01$ for ACHD mortality rates).

The increase in referrals to specialized ACHD centers after 1997 was similar for those with severe and other CHD (severe CHD: RR +1.3% increasing to +5.6%; interaction $P=0.008$; other CHD: RR +2.1% increasing to +7.1%; interaction $P<0.0001$). In contrast, the reduction in ACHD mortality after 2000 was more marked in those with severe CHD (severe CHD: RR +4.0% decreasing to −5.0%; interaction $P=0.07$; other CHD: RR +1.8 decreasing to −5.0%; $P=0.04$).

Predictors of Death

We assessed the principal cause of death among hospitalized patients before (n=389) and after (n=165) the year 2000 (Table 2). The distribution of the causes of death was not significantly different between both periods ($P=0.74$). Congenital or cardiac causes accounted for the majority of deaths (40.6% versus 41.2%, $P=0.90$), with neoplasms and respiratory diseases the second and third most frequent causes of death.

In the case–control study, ACHD referral center care was associated with lower odds of death, independent of comorbidity, age, sex, and CHD severity (OR, 0.82; 95% CI, 0.68–0.97). An increased burden of comorbid disease (OR, 3.62; 95% CI, 3.13–4.18), older age (OR, 1.98; 95% CI, 1.89–2.08),

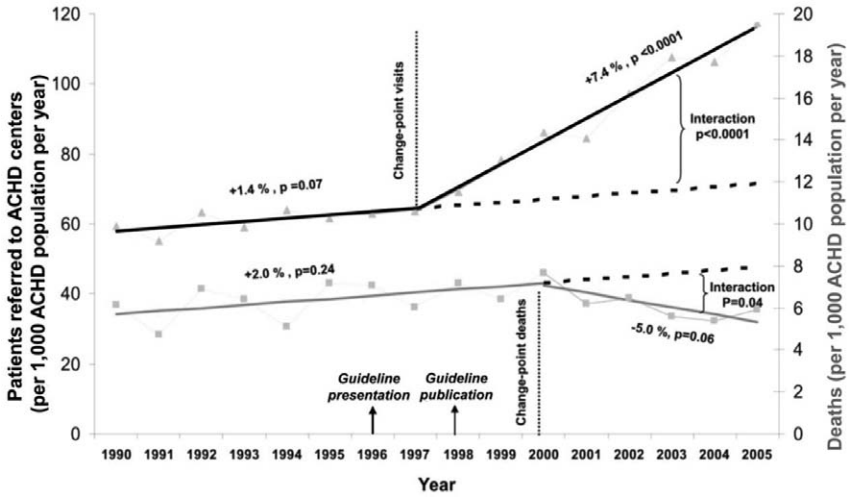

Figure 2. Time-series analysis: referral to specialized ACHD centers and ACHD patient mortality. Time-series analysis illustrating observed specialized ACHD center referral (black line) and ACHD mortality (grey line) per 1000 ACHD population per year, between 1990 and 2005. The dashed lines indicate expected trends after the change points identified by Poisson regression, and the black or grey lines represent the observed trends. ACHD indicates adult congenital heart disease.

Table 2. Principal Causes of Death

Principal Cause of Death *	Before Year 2000 (n=389)	After Year 2000 (n=165)	*P* Value
Diseases of congenital anomalies of the circulatory system	158 (40.6)	68 (41.2)	0.90
Neoplasms	70 (18.0)	30 (18.2)	0.94
Diseases of respiratory system	32 (8.2)	18 (10.9)	0.31
Infectious and parasitic diseases	19 (4.9)	5 (3.0)	0.33
Diseases of other systems	110 (28.3)	44 (26.7)	0.70

Data represent number and percentage.
*P value for trend in the distribution of cause of death = 0.74.

severe CHD (OR, 1.93; 95% CI, 1.55–2.41), and male sex (OR, 1.48; 95% CI, 1.31–1.68) were associated with an increased risk of death (Figure 3).

ACHD referral center care was also associated with a similar protective effect in the cohort study, both in the crude and adjusted analyses. Adjusted Kaplan-Meier estimates of mortality demonstrate that, in comparison with patients with nonspecialized care, those with ACHD referral center care had significantly improved survival (log rank $P<0.001$; Figure 4). After adjustment for age, sex, CHD severity, comorbidity, associated genetic syndromes, geographic location, and estimated individual net income, the hazard of death was 22% lower in patients whose first visit was to a specialized ACHD center in comparison with those whose first visit was to a nonreferral center (HR, 0.78; 95% CI, 0.65–0.94). Other predictors of mortality included male sex (HR, 1.59; 95% CI, 1.33–1.91), age (HR, 1.47; 95% CI, 1.36–1.58), congestive heart failure (HR, 3.07; 95% CI, 2.52–3.74), pulmonary hypertension (HR, 2.78; 95% CI, 1.87–4.14), stroke (HR, 1.60; 95% CI, 1.17–2.19), atrial fibrillation (HR, 1.56, 95% CI 1.26–1.94), and associated genetic syndromes (HR, 3.45; 95% CI, 2.05–5.80). Urban domicile and average net individual income were not associated with ACHD mortality.

When patients were stratified according to CHD severity, however, we found that the protective effect of ACHD referral center care observed for the overall CHD population was predominantly driven by the subgroup of patients with severe CHD (severe: HR, 0.38; 95% CI, 0.22–0.67; other: HR, 0.82; 95% CI, 0.67–0.99; Figure 5).

Discussion

In this population-based analysis, we examined the impact of specialized care on ACHD patient mortality. We observed a significant reduction in ACHD mortality rates concurrent with a parallel increase in referral to specialized ACHD centers, following the introduction of national consensus guidelines. Moreover, we found that ACHD referral center care was associated with a significant reduction in mortality, independent of age, sex, and comorbidity, and specifically in those with severe CHD. To our knowledge, this is the first study to analyze the relationship between specialized ACHD care and mortality.

Some differences in baseline characteristics among the ACHD care groups are noteworthy. First, younger patients were found among the ACHD referral center and no cardiology follow-up groups. This observation is likely explained by younger patients with more severe CHD being followed at an ACHD referral center, whereas younger patients with less severe CHD may be less likely to comply with follow-up.[29] Second, we observed a higher proportion of female patients among those without ongoing ACHD care (62%). Previous studies have demonstrated that, although females are more likely to be compliant with follow-up,[29] they account for a smaller proportion of cardiology-related healthcare encounters than males, likely reflecting the more severe CHD associated with male sex.[30,31]

Clinical practice guidelines are "systematically developed statements to assist practitioner and patient decisions about appropriate healthcare for specific clinical circumstances."[32] Guideline implementation aims to enhance patient care by

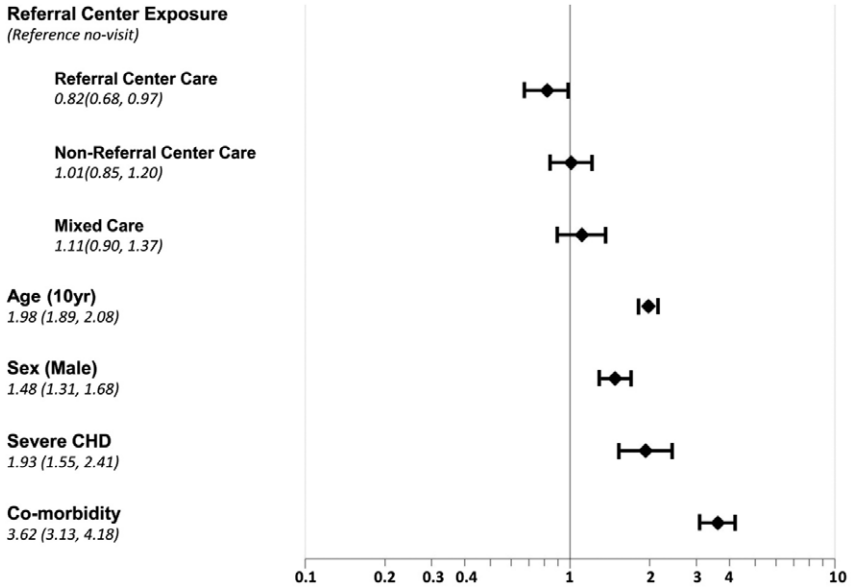

Figure 3. Case–control study: independent predictors of ACHD mortality. Plot showing odds ratios for mortality in ACHD patients. ACHD indicates adult congenital heart disease; and CHD, congenital heart disease.

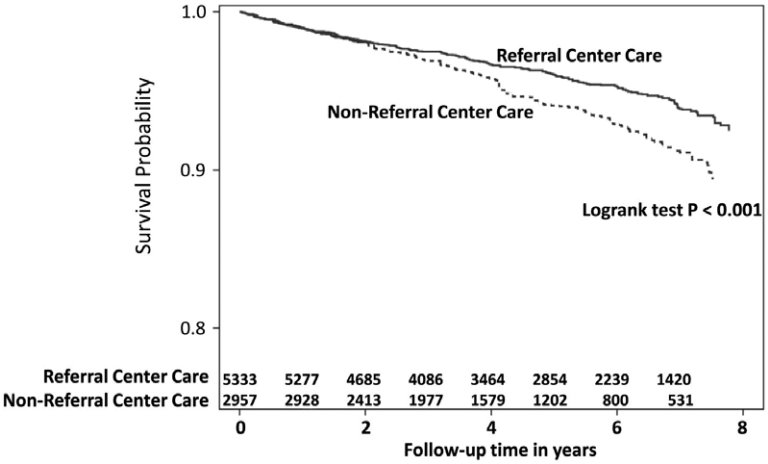

Figure 4. Cohort study: adjusted Kaplan-Meier survival curves. Adjusted Kaplan-Meier survival curves in patients with ACHD referral center care (solid line) and those with nonreferral care (dashed line). ACHD indicates adult congenital heart disease.

reducing the practice variability and through the appropriate use of medical advances.[33] However well intentioned, the impact of guidelines on modifying physician behavior appears inconsistent.[34] Importantly, we observed a significant change in physician behavior relating to the referral of ACHD patients for specialized care following publication of the CCS ACHD guidelines in 1998.[12] Although referral to specialized ACHD centers was steadily increasing before 1998, the observed acceleration thereafter was considerable (+1.4% to +7.4%). Practice guidelines from the major cardiology societies support the referral of ACHD patients to specialized centers based on accumulating evidence of lower patient mortality associated with high-volume institution care across multiple clinical conditions and care settings.[9,10,35–38] The practice-makes-perfect hypothesis introduced by Luft et al[39] explains how high-volume specialized centers improve patient outcomes: increased volume improves physician experience and skill, stimulates research and affords for economies of scale that facilitate the acquisition of specialized equipment, and promotes service development. Such advantages are particularly evident in complex patient groups,[40] such as those with ACHD that require high-risk surgical and transcatheter interventions. The impact of specialized ACHD care on patient outcomes has not been formally appraised, however.

It is therefore noteworthy that, in parallel with the increasing rates of referral to specialized ACHD centers, we observed a significant deviation from the projected mortality curve of our ACHD population (+2.0% to −5.0%). This mortality reduction was first noted in the year 2000, 2 years following the publication of the CCS guidelines. The time lag between referral and the protective effect associated with specialized ACHD is multifactorial and complex. The determinants of the protective effect of specialized care likely reflect the balance between the patient-level needs, the availability of specialized medical and interventional care, and factors governing the implementation and access to care. Nonetheless, our findings are consistent with the notion that expert care can significantly impact patient outcomes, as has been demonstrated with other areas of specialized care delivery.[41,42]

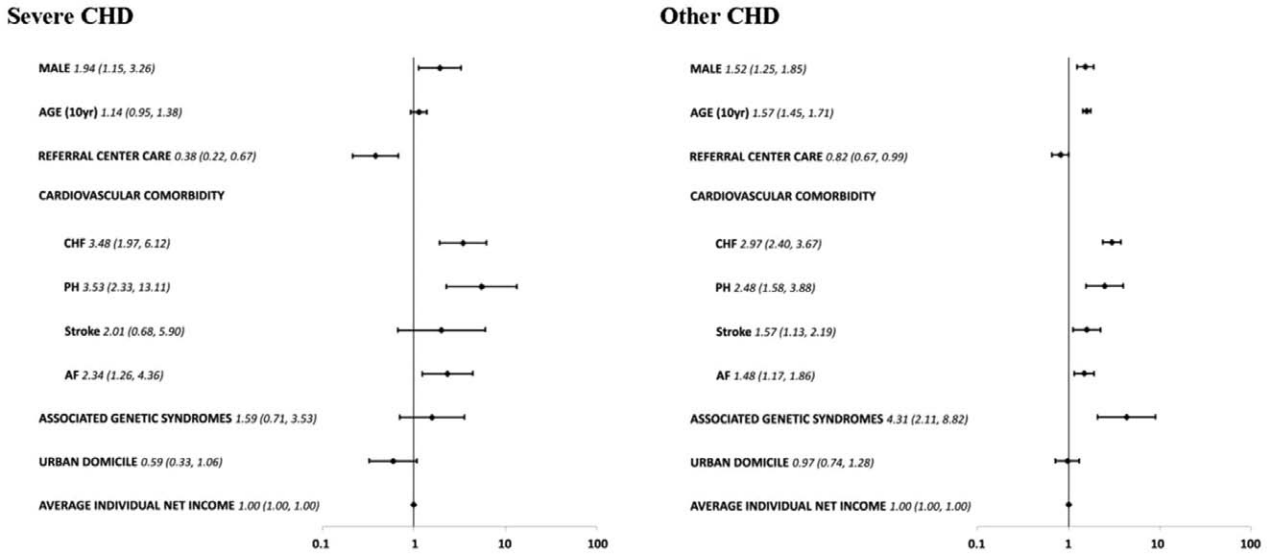

Figure 5. Cohort study: predictors of ACHD mortality. Plot showing hazard ratios for mortality in ACHD patients stratified according to disease severity: severe CHD (**left**); other CHD (**right**). ACHD indicates adult congenital heart disease; AF, atrial fibrillation; CHD, congenital heart disease; CHF, congestive heart failure; and PH, pulmonary hypertension.

In the current study, specialized ACHD care was independently associated with reduced ACHD patient mortality when tested in both the case–control and cohort studies. This observation was robust despite a higher incidence of severe CHD in those treated in ACHD referral centers. Advances in diagnosis and surgical treatment of CHD are expected to have impacted survival, and, although these advances were potentially available to both ACHD referral and nonreferral centers, the increased accessibility to expertise and application of tertiary and quaternary care by caregivers specialized in ACHD can explain the protective effect of ACHD referral centers. This, in fact, constitutes the rationale for the recommendations that all ACHD patients be assessed in a specialized ACHD center at least once, and that patients with moderate or complex CHD be followed periodically or consistently in specialized ACHD centers.[5,9,10] Our study provides new data that support these recommendations.

The observation that <1 in 3 ACHD patients actively receives care at a specialized center suggests that the need for delivery of appropriate health care to ACHD patients remains largely unmet.[13] Specialized ACHD centers should provide a range of services specifically geared toward meeting the needs of this patient population. Physician expertise is key to delivering quality ACHD care such that ACHD subspecialty training programs have emerged to address the shortage of manpower in this emerging field of cardiology.[43,44]

In 2005, more than three-quarters of ACHD patients did not have an outpatient cardiology review. Although direct comparisons among studies and age groups may be misleading, this figure compares poorly with the 47% of pediatric and 61% of young adult transition (18–22 years of age) CHD patients that do not attend for follow-up visits.[29] This observation would support the notion that lapses in care are not limited to pediatric CHD patients transitioning to ACHD sites, and it confirms care gaps to be among the greatest challenges to optimizing ACHD care.[14,29,45,46] Although it is easy to surmise that complex ACHD patients should receive care in regional centers, patients with more straightforward defects may also require specialist care under certain circumstances. Hence, expert consensus recommends that all ACHD patients be seen at least once in a specialized center, with further follow-up subsequently titrated on an individualized basis.[9,10] In the current study, the reduction in ACHD mortality associated with specialized ACHD care was driven by the subgroup of patients with severe CHD. Clearly, these patients have the most to gain from the high-level expertise concentrated at regional ACHD centers. It is therefore concerning that we observed care gaps in 15% of patients deemed to have severe CHD lesions in 2005. Ongoing initiatives to minimize care gaps and optimize referral to specialized ACHD centers are focused on greater patient engagement and education and enhanced awareness of access to available resources for patients and physicians.[14]

Increasing geographical distance between the patient and the ACHD referral center has also been identified as a predictor of care gaps. Hence, mixed care between the local nonreferral and regional ACHD referral center is a model of care with potential advantages for both patient and healthcare institution.[14] We elected to assess the impact of mixed care separate from patients that received ACHD referral center care because of the emerging importance of this care model. Ultimately, our analysis was limited by small patient numbers (n=102), but it is noteworthy that this care model was used infrequently among our study population and appeared to be reserved for patients with the greatest burden of disease. Further study is required to validate the impact of mixed care on care gaps and patient outcome.

Limitations

The study findings should be interpreted in light of the study design. This retrospective analysis used administrative databases that are prone to misclassification of disease. However, the algorithms applied to the Quebec CHD database have been published and validated for both large population analyses and specific subgroups of patients.[7,16,20–22,47] We minimized misclassification by using all available data for a given subject, including inpatient, outpatient, procedural, and provider information and by performing manual audits of random samples of the original 71 467 subjects. Misclassification of exposure is also possible, although the definition of a ACHD referral center in the 1998 CCS consensus statement was justifiably specific.[12] Although selection bias in patients followed in ACHD referral centers should be considered, it is unlikely to have accounted for the enhanced survival given that these patients had more severe CHD than those treated in nonreferral centers. Further research into the utility of sharing the care of ACHD patients between the local hospital and regional ACHD referral center is warranted, because we were not able to evaluate this management strategy owing to the limited number of patients receiving this model of care in our study. Our study was not designed to identify determinants of ACHD specialized care associated with its protective effect. However, this important question should be addressed in future studies. Our database contains residential 3-digit postal codes but no individual-level geographic or socioeconomic data. We approximated the average net individual income according to geographic location using the 3-digit postal code directly linked to income data from the 2006 Canadian Census. Although having direct individual level socioeconomic data would have been more specific, this approach using Canadian Census data has been previously validated.[48] Although our results may reasonably by generalized to the rest of Quebec, variations in healthcare systems may impact the applicability of these findings to other countries. However, Canadian guidelines closely espouse their US equivalents, and the standards of care for ACHD patients are similar between these nations.[49,50]

Conclusions

In this large population-based study, a significant rise in referral to specialized ACHD centers was observed concurrent with national consensus guidelines. Importantly, a significant reduction in mortality was associated with referral to an ACHD center and was most pronounced in patients with severe forms of CHD. Our findings are expected to inform policies recommending and supporting the promotion of specialized care for ACHD patients.

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Disclosures

None.

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CLINICAL PERSPECTIVE

There are expected to be over a million adults living with congenital heart disease (CHD) in the United States. Clinical guidelines recommend specialized care for this complex patient population burdened with life-long morbidity. Despite the growing need to bring quality to CHD care, there are no data demonstrating that specialized adult CHD (ACHD) care can improve outcomes. In this population-based analysis, we examined the impact of specialized care on ACHD mortality. We examined referral rates to specialized ACHD centers and ACHD patient mortality rates between 1990 and 2005 by using the population-based Quebec Congenital Heart Disease database that includes 71 467 patients. Concurrent with the guideline publication recommending specialized care for ACHD patients, we showed a significant increase in referral rates to specialized ACHD centers associated with a parallel significant reduction in ACHD patient mortality. Independent of age, sex, and comorbidity, specialized ACHD care was associated with reduced odds of death, an effect predominantly driven by patients with severe CHD. To our knowledge, this is the first study to analyze the relationship between specialized ACHD care and mortality. Our findings support the guidelines recommending specialized care for all ACHD patents as a means of improving quality of care and outcomes for this growing patient population.