



Improved Outcomes of Heart Transplantation in Adults With Congenital Heart Disease Receiving Regionalized Care

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ABSTRACT

BACKGROUND The number of adult congenital heart disease (CHD) patients undergoing heart transplantation is increasing rapidly. CHD patients have higher surgical risk at transplantation. High-volume adult CHD transplant centers may have better transplant outcomes.

OBJECTIVES This study aimed to evaluate the effect of center CHD transplant volume and expertise on transplant outcomes in CHD patients.

METHODS The authors studied heart transplantations in CHD patients age ≥ 18 years using the United Network of Organ Sharing (UNOS) database for the primary outcomes of waitlist mortality and post-transplant outcomes at 30 days and 1 year. Transplant centers were assessed by status as the highest CHD transplant volume center in a UNOS region versus all others, presence of Adult Congenital Heart Association accreditation, and adult versus pediatric hospital designation.

RESULTS Between January of 2000 and June of 2018, 1,746 adult CHD patients were listed for transplant; 1,006 (57.6%) of these underwent heart transplantation. After adjusting for age, sex, listing status, and inotrope requirement, waitlist mortality risk was lower at Adult Congenital Heart Association accredited centers (hazard ratio: 0.730; $p = 0.020$). Post-transplant 30-day mortality was lower at the highest volume CHD transplant center in each UNOS region (hazard ratio: 0.706; $p = 0.014$).

CONCLUSIONS Designated expertise in CHD care is associated with improved waitlist outcomes for CHD patients listed for transplantation. Post-transplant survival was improved at the highest volume regional center. These findings suggest a possible advantage of regionalization of CHD transplantation. (J Am Coll Cardiol 2019;74:2908-18)
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Although the vast majority of infants born with congenital heart disease (CHD) survive to adulthood, overall life expectancy is reduced in complex CHD. Heart failure is the leading cause of premature morbidity and mortality in adults with CHD (ACHD) (1-3). The efficacy of medical therapy for heart failure in CHD is not well established, so many ACHD patients require consideration for heart transplantation.

Because of complex anatomy, prior cardiac surgery, the presence of collateral vessels, and associated liver and renal disease, heart transplantation for ACHD patients portends a high surgical risk (4-8). Allosensitization increases duration and mortality risk on the waitlist for ACHD patients and is an additional risk factor for primary graft dysfunction following transplant (8-10). Therefore, ACHD patients with advanced heart failure are challenging for



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transplant programs. Although early post-transplant mortality is elevated for ACHD patients, those who survive transplant have excellent long-term post-transplant outcomes (9,11,12). Previous studies have shown that ACHD patients who are transplanted at low-volume transplant programs have increased early mortality (13). However, CHD patients have anatomic and physiologic challenges that may not be familiar, even at high-volume transplant centers, unless these centers have experience and expertise in ACHD care and ACHD surgery. The impact on ACHD expertise and transplant volume on ACHD transplant outcomes has not been studied. We therefore hypothesize that clustered CHD expertise is associated with improved transplant outcomes in ACHD.

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METHODS

PATIENT POPULATION. We studied adult heart transplant candidates age ≥ 18 years with CHD in the Scientific Registry of Transplant Recipients registry from the United Network of Organ Sharing (UNOS) who were listed for heart transplantation between January 2000 and June 2018. CHD patients were defined as those with a diagnosis of CHD as their listing diagnosis. Additionally, patients were also included if there was a diagnosis of hypoplastic left heart syndrome or if 1 of the following words were identified in the thoracic diagnosis text description: “transposition,” “congenital,” “Ebstein,” “Fontan,” “tetralogy,” “atresia,” “Noonan,” or “William.” Misspellings were sought using a strategy of searching for abbreviated forms of the aforementioned keywords. Patients with Eisenmenger syndrome and dual organ listings were excluded.

CHD TRANSPLANT CENTER AND VOLUME. Several measures were used to distinguish transplant centers in the study. Total ACHD transplant volume was calculated for each center for the duration of the study period. The highest total volume ACHD transplant center in each of the 11 UNOS regions was identified for analysis. Given the long period of the study, an additional sensitivity analysis was performed for the highest volume ACHD transplant center in the UNOS region over 2 eras of the study: period 1 was defined as 2000 to 2009 and period 2 was defined as 2010 to 2018. All transplant centers were also denoted by their Adult Congenital Heart Association (ACHA) accreditation status as of December 2018. Since 2017, ACHA accreditation has been awarded to centers with a comprehensive multidisciplinary ACHD team (14). All hospitals with current ACHA accreditation status and affiliated sites were

considered to be ACHA-accredited centers for the purposes of the study. Accreditation requires a dedicated ACHD heart failure team, which includes heart failure cardiologists and CHD surgeons. In addition, pediatric hospitals were compared with adult medical centers for the primary outcomes.

STATISTICAL ANALYSIS. Patient characteristics are displayed as mean \pm SD for continuous variables and were analyzed with an independent Student’s *t*-test. Categorical variables are reported as percent prevalence and analyzed with a chi-square test. Temporal trends are graphed by 3-year clusters. The primary waitlist outcome was death or being delisted for being too ill. The primary post-transplant outcome was graft failure, indicated by either death or retransplantation. A Fine-Gray competing risk regression analysis was used to evaluate waitlist outcomes with a competing outcome of heart transplantation. A Cox proportional hazards model was used to assess the primary outcome post-transplant. Univariate modeling was performed to identify patient variables that were associated with the primary outcomes for inclusion in multivariate modeling ($p < 0.20$ was accepted for retention). Candidate age, listing status, and inotrope requirement were identified for inclusion in a multivariate analysis model, with the addition of ischemic time and donor sex for the post-transplant analysis. All analyses are multivariate unless otherwise specified. In subsequent sensitivity analysis, the model was restricted to ACHD patients with a confirmed prior sternotomy. A p value < 0.05 was deemed statistically significant. Data analysis was performed with Stata IC, version 15 (Stata Corp, College Station, Texas). The University of Washington institutional review board approved this study.

RESULTS

The study cohort consisted of 1,746 ACHD heart transplant candidates listed for transplantation between January 1, 2000, and July 1, 2018. Of these patients, 1,006 (57.6%) patients were transplanted at 124 transplant centers during the study period. The number of ACHD patients listed for transplantation and the number of transplant recipients have both increased over time, with the greatest changes seen in the adult hospitals (Figure 1). Baseline characteristics are listed in Table 1 by highest volume center in UNOS regions versus all others. ACHD patients at highest volume centers were more likely to be inotrope dependent and have a lower body mass index. There was no significant difference in the use of intra-aortic

ABBREVIATIONS AND ACRONYMS

ACHA = Adult Congenital Heart Association

ACHD = adult congenital heart disease

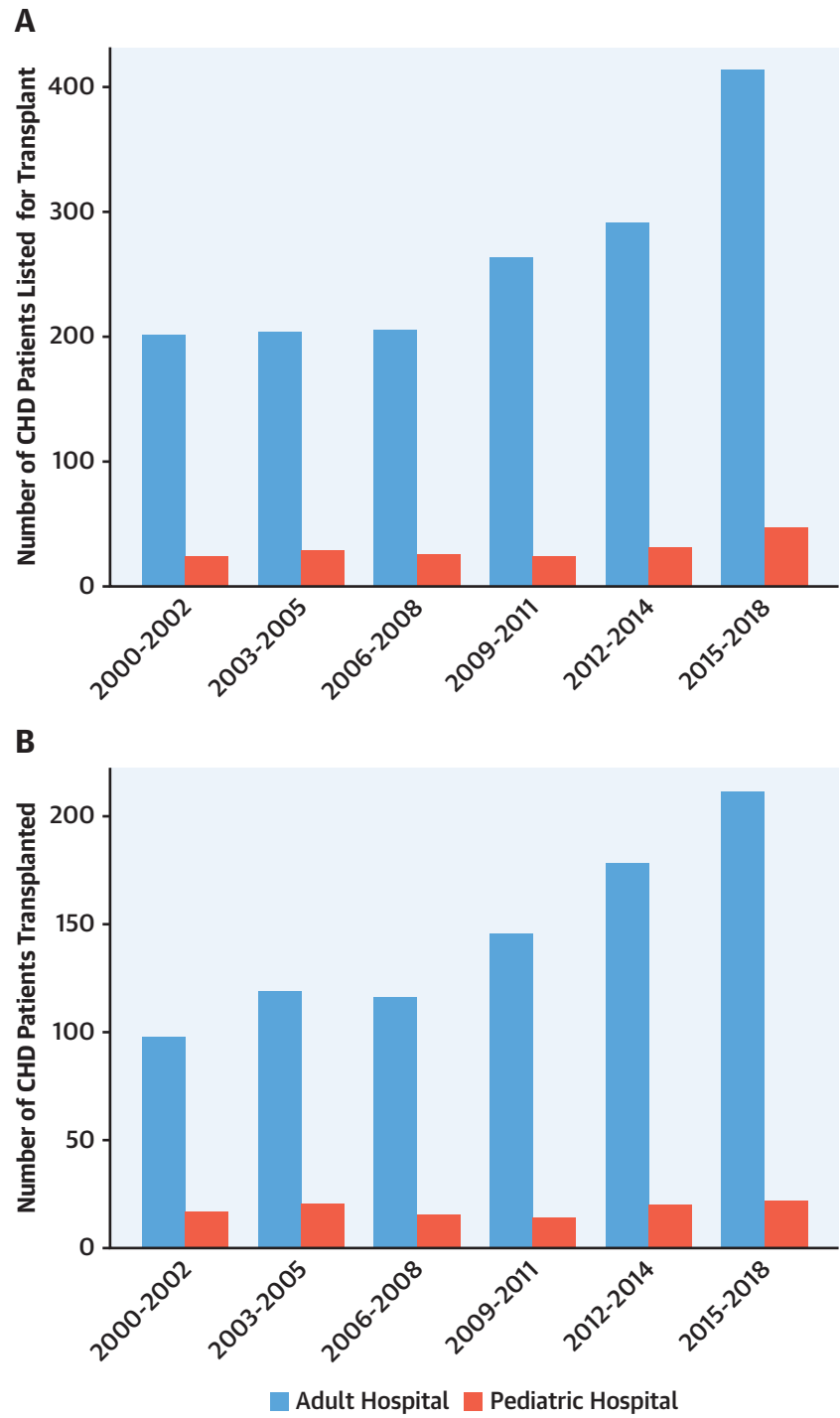
CHD = congenital heart disease

CI = confidence interval

HR = hazard ratio

UNOS = United Network of Organ Sharing

FIGURE 1 Trends in Congenital Heart Disease Patients Transplant Care Over Time



(A) Trend of patients listed for transplant over time. (B) Trends of patients receiving a transplant over time. CHD = congenital heart disease.

balloon counterpulsation, extracorporeal membrane oxygenation, or ventricular assist devices at transplant (Table 1). A wide variation of ACHD transplant volumes existed between all transplant centers. Only 3 centers performed 40 or more ACHD transplants over the span of 18 years; these centers accounted for 14.7% of all congenital transplants during this time (Figure 2). The 11 highest volume ACHD transplant centers in the UNOS region performed 34.2% of all ACHD transplants during this period compared with the other 113 centers who performed 65.8% of the ACHD transplantations. In addition, 49 transplant centers (39.5%) performed ≤ 1 heart transplantation every 5 years, accounting for 223 (12.4%) of all ACHD heart transplantation in this study cohort. Of 124 transplant centers, 45 were ACHA accredited or affiliated with ACHA-accredited hospitals. There were 97 adult medical centers and 27 pediatric hospitals. In our study cohort, 184 (10.4%) patients were listed at pediatric hospitals and 110 (6.2%) candidates were ultimately transplanted there. The mean age of ACHD candidate listing for transplant was lower at pediatric hospitals compared with adult hospitals (23.3 ± 6.5 years vs. 36.9 ± 12.0 years; $p < 0.0001$). The mean age at transplant was also lower at pediatric hospitals compared with adult hospitals (24.2 ± 6.7 years vs. 38.0 ± 12.4 years; $p < 0.0001$). There was no significant difference in the use of intra-aortic balloon counterpulsation ($p = 0.247$), extracorporeal membrane oxygenation ($p = 0.337$), or ventricular assist

TABLE 1 Baseline Characteristics at Transplant

	Highest ACHD Transplant Volume Center in UNOS Region (n = 542)	All Other Centers in UNOS Region (n = 1,041)	p Value
Age, yrs	38.8 \pm 12.3	37.6 \pm 12.5	0.177
Male	312 (57.6)	640 (61.5)	0.131
White	454 (83.8)	839 (80.6)	0.122
BMI, kg/m ²	24.5 \pm 5.1	25.6 \pm 5.3	<0.0001
Smoker	33 (6.1)	75 (7.2)	0.404
Diabetes mellitus	89 (16.4)	184 (17.7)	0.531
History of CVA	9 (1.7)	20 (1.9)	0.714
Ischemic CM	4 (0.73)	3 (0.29)	0.201
Inotropes	333 (61.5)	466 (44.8)	<0.0001
IABP	17 (3.2)	26 (2.5)	0.590
ECMO	9 (1.6)	15 (1.4)	0.879
VAD	26 (4.8)	52 (5.0)	0.863

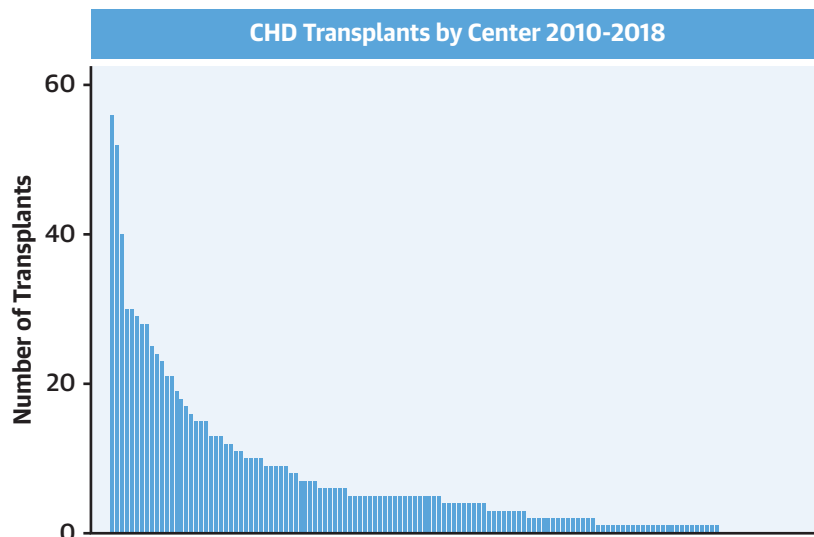
Values are mean \pm SD or n (%).

BMI = body mass index; CM = cardiomyopathy; CVA = cerebrovascular accident; ECMO = extracorporeal membrane oxygenation; IABP = intra-aortic balloon counter-pulsation; VAD = ventricular assist device.

devices ($p = 0.226$) at the time of transplantation between pediatric and adult medical centers. Prior sternotomy was noted for 71.6% of the patient sample, making up 71.4% of transplanted patients at adult hospitals and 73.6% of transplanted patients at pediatric hospitals ($p = 0.623$).

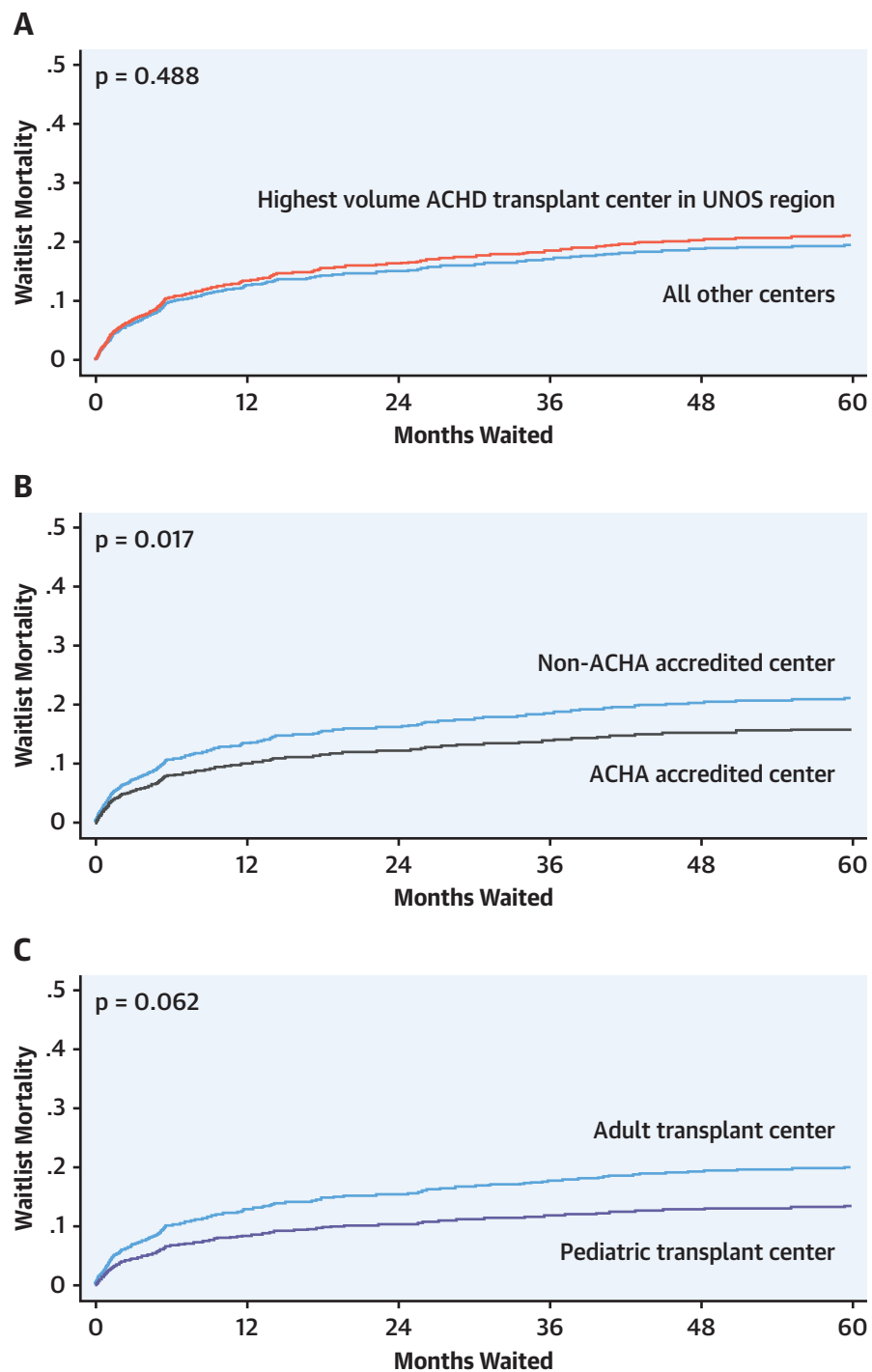
WAITLIST MORTALITY. Over the study duration, 16.5% of ACHD patients died before transplant or were removed from the waitlist for being too ill. The remainder of patients, who were not otherwise

FIGURE 2 Variation in the Number of Transplants by Each Center Over the Duration of the Study Period



X-axis plots individual centers, ranked by transplant volume. CHD = congenital heart disease.

FIGURE 3 Cumulative Incidence of Waitlist Mortality for All Listed Congenital Heart Disease Patients Age ≥ 18 Years



Unadjusted curves for waitlist mortality, derived from a cumulative incidence function from a competing risks regression model. **(A)** Highest CHD volume center in a UNOS region versus all other centers. **(B)** ACHA-accredited versus not accredited of both adult and pediatric hospitals. **(C)** Adult versus pediatric hospital. ACHA = Adult Congenital Heart Association; CHD = congenital heart disease; UNOS = United Network of Organ Sharing.

TABLE 2 Risk of Waitlist Mortality or Delisting for Being too Ill

	Waitlist Mortality Rate, %	Univariate HR (95% CI)	p Value	Multivariate HR (95% CI)	p Value
Highest volume CHD transplant center in UNOS region vs. all other transplant centers in the same UNOS region	18.3 vs. 16.5	1.091 (0.852-1.397)	0.488	1.062 (0.827-1.366)	0.635
ACHA-accredited center vs. non-ACHA-accredited center	13.4 vs. 17.9	0.730 (0.563-0.946)	0.017	0.730 (0.559-0.951)	0.020
Pediatric hospital listing vs. adult hospital listing	10.9 vs. 17.1	0.645 (0.407-1.022)	0.062	0.654 (0.405-1.056)	0.083

Hazard of the outcome of waitlist mortality or being delisted for being too ill, derived from a Fine-Gray competing risk regression model with the competing outcome of heart transplantation. ACHA = Adult Congenital Heart Association; CHD = congenital heart disease; CI = confidence interval; HR = hazard ratio; UNOS = United Network of Organ Sharing.

transplanted, includes those who were still waiting, deemed medically unsuitable for transplant, transferred care out of the transplant center, or were delisted for improvement in their medical condition. Listing for transplantation at the highest volume ACHD transplant center in the UNOS region did not affect waitlist mortality (hazard ratio [HR]: 1.062; 95% confidence interval [95% CI] 0.827 to 1.366; $p = 0.635$) (Figure 3A). ACHA accreditation of the listing center was associated with a lower mortality on the waitlist (HR: 0.730; 95% CI: 0.559 to 0.951; $p = 0.020$) (Table 2, Figure 3B), and although there was a trend toward the association of heart transplant listing at a pediatric hospital being associated with lower waitlist mortality, this did not reach statistical significance when compared with adult transplant centers (multivariate HR: 0.654; 95% CI: 0.405 to 1.056; $p = 0.083$) (Table 2, Figure 3C). In our sensitivity analysis of prior sternotomy patients, those with a history of a prior sternotomy had a significantly lower waitlist mortality at pediatric hospitals (HR: 0.539; 95% CI: 0.299 to 0.971; $p = 0.040$) and ACHA-accredited hospitals (HR: 0.697; 95% CI: 0.504 to 0.962; $p = 0.028$), but not at the highest volume ACHD transplant center in the UNOS region (HR: 0.847; 95% CI: 0.597 to 1.203; $p = 0.355$).

POST-TRANSPLANT OUTCOMES. The rate of graft failure was 10.7% in the first month and 16.9% in the first year in the study cohort. Overall graft failure decreased on average by 12.4% (odds ratio: 0.883; 95% CI: 0.859 to 0.908) each year over the duration of the study. ACHD patients transplanted at the highest volume ACHD transplant center in the UNOS region had a lower risk of graft failure at 30 days (HR: 0.706; 95% CI: 0.535 to 0.932; $p = 0.014$) and at 2 years (HR: 0.658; 95% CI: 0.443 to 0.978; $p = 0.038$) (Table 3, Figure 4A). In the sensitivity analysis evaluating the 2 9-year periods separately, ACHD transplants in the highest volume ACHD transplant center in the UNOS region were not associated with decreased 1-year post-transplant graft failure between 2000 and 2009 (HR: 0.802; 95% CI: 0.599 to 1.613; $p = 0.947$), but was associated with decreased 1-year post-transplant graft failure between 2010 and 2018 (HR: 0.554; 95% CI: 0.318 to 0.963; $p = 0.036$).

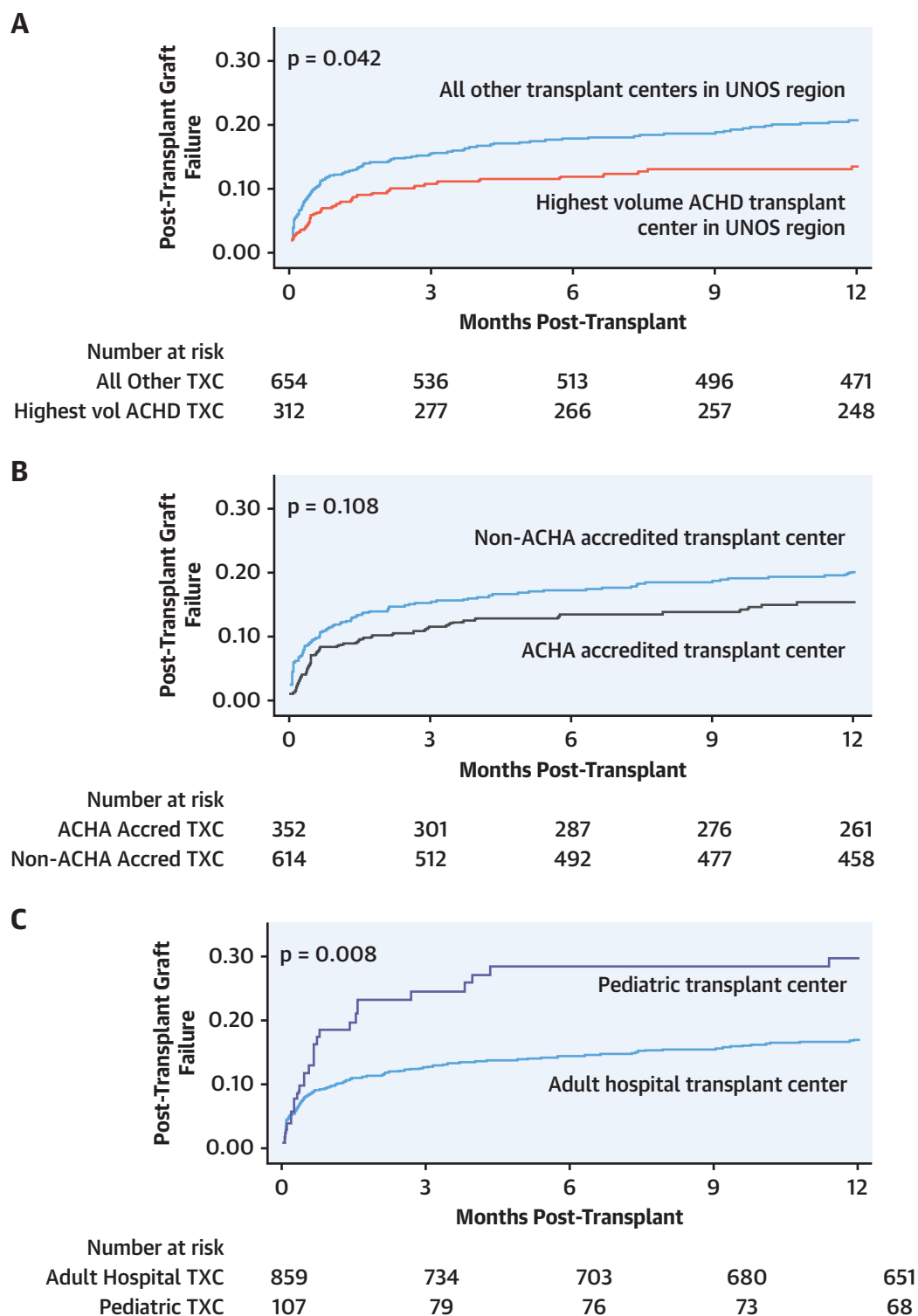
ACHA accreditation did not affect post-transplant outcomes at 30 days (HR: 0.871; 95% CI: 0.681 to 1.115; $p = 0.273$) (Table 3) or at 1 year (HR: 0.741; 95% CI: 0.522 to 1.052; $p = 0.094$) (Figure 4B). In both univariate and multivariate analyses, ACHD patients who received their transplant at a pediatric hospital had a significantly increased risk of graft failure

TABLE 3 Risk of Post-Transplant Graft Failure

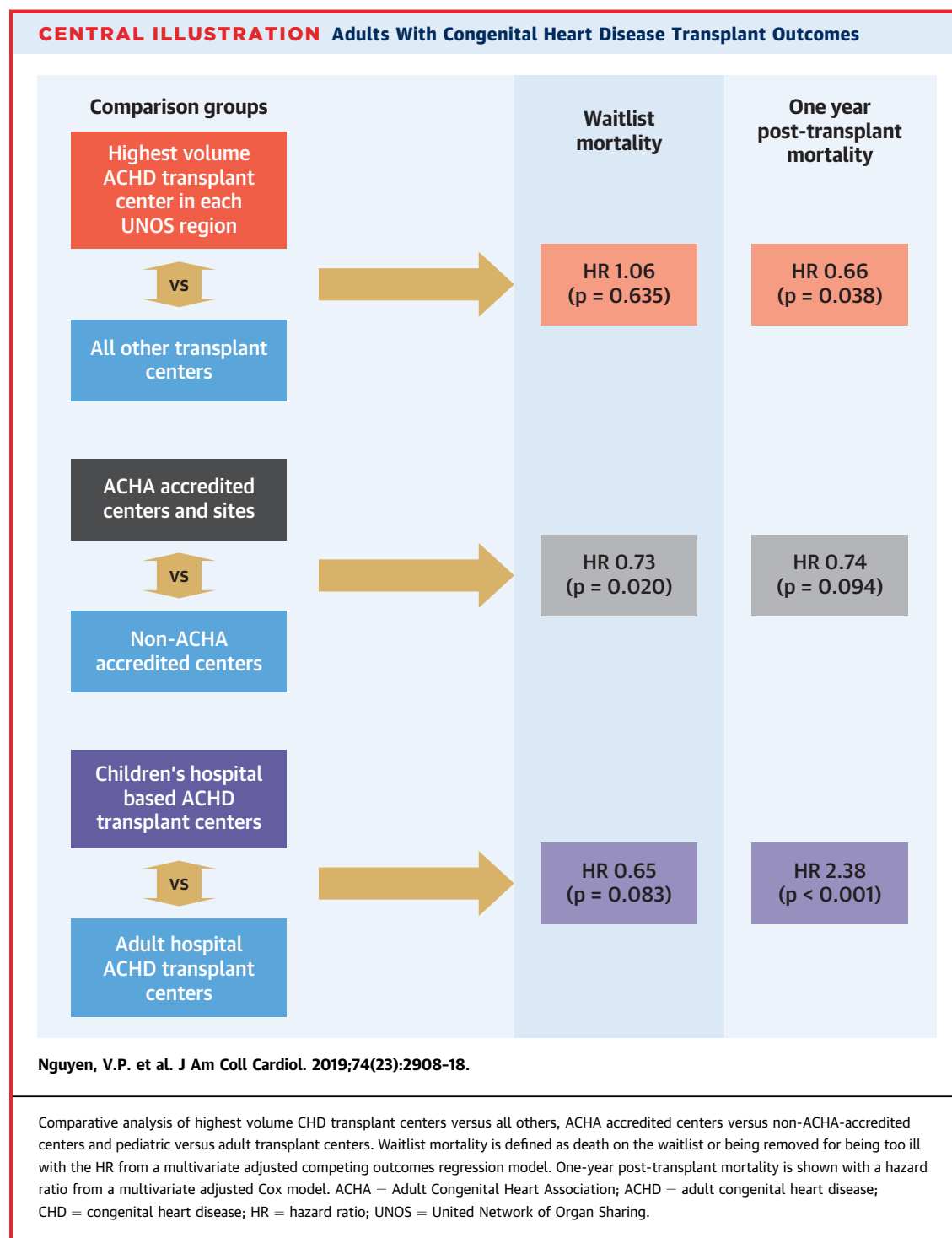
	Univariable HR (95% CI)	p Value	Multivariable HR (95% CI)	p Value
Top UNOS region hospital vs. all other hospitals in region				
30-day mortality	0.767 (0.591-0.995)	0.046	0.706 (0.535-0.932)	0.014
1-yr mortality	0.677 (0.464-0.986)	0.042	0.658 (0.443-0.978)	0.038
ACHA accredited center vs. non-ACHA-accredited center				
30-day mortality	0.911 (0.719-1.154)	0.439	0.871 (0.681-1.115)	0.273
1-yr mortality	0.759 (0.541-1.062)	0.108	0.741 (0.522-1.052)	0.094
Pediatric hospital listing vs. adult hospital listing				
30-day mortality	1.545 (1.129-2.114)	0.007	1.553 (1.097-2.198)	0.013
1-yr mortality	1.750 (1.157-2.649)	0.008	2.38 (1.480-3.830)	<0.0001

Hazard ratio of post-transplant graft failure as defined by mortality or retransplantation, derived from a Cox proportional hazards model. Abbreviations as in Table 2.

FIGURE 4 Post-Transplant Cumulative Hazard for Graft Failure for Transplanted Patients With Congenital Heart Disease



Unadjusted cumulative hazard function for graft failure as derived from the Cox proportional hazards model. **(A)** Highest CHD volume center in a UNOS region versus all other centers. **(B)** ACHA-accredited versus non-ACHA-accredited center. **(C)** Pediatric transplant center versus adult transplant center. ACHA = Adult Congenital Heart Association; CHD = congenital heart disease; TXC = transplant center; UNOS = United Network of Organ Sharing; Vol = volume.



within the first 30 days (HR: 1.553; 95% CI: 1.097 to 2.198; p = 0.013) (Table 3, Figure 4) and at 1 year (HR: 2.380; 95% CI: 1.480 to 3.830; p < 0.0001) (Figure 4) when compared with adult transplant centers. In a sensitivity analysis of patients with a

history of a prior sternotomy, the higher 30-day mortality risk at pediatric hospitals persisted (HR: 1.638; 95% CI: 1.050 to 2.521; p = 0.029). The effect of transplant at the highest volume ACHD transplant center in the UNOS region was no longer

significant (HR: 0.733; 95% CI: 0.520 to 1.032; $p = 0.076$) and it remained nonsignificant for ACHA-accredited centers (HR: 0.801; 95% CI: 0.530 to 1.211; $p = 0.292$).

DISCUSSION

Our study demonstrates that CHD expertise and ACHD transplant volume is associated with improved waitlist and post-transplant outcomes in ACHD patients ([Central Illustration](#)). Waitlist mortality for ACHD transplant candidates was lower at ACHA-accredited transplant centers than nonaccredited centers. In ACHD patients with prior sternotomy, waitlist mortality was lower at pediatric transplant centers than at adult transplant centers. Patients who underwent heart transplant at adult transplant centers and at the highest volume ACHD transplant center in the UNOS region had a lower risk of graft failure following transplantation. Although this retrospective analysis of the UNOS registry cannot be able to distinguish between center-level and patient-level factors as a cause of these differential mortality risks, our findings suggest that expertise in congenital heart anatomy and management is important for the care of these complex CHD patients with advanced heart failure.

ACHA-accredited comprehensive care centers had the lowest waitlist mortality. These centers are required to have a multidisciplinary team with multiple board-certified ACHD cardiologists and must include dedicated ACHD heart failure specialists, congenital heart surgeons, and comprehensive diagnostic and interventional services by specialists in CHD. Further sensitivity analyses demonstrated lower waitlist mortality at pediatric centers in adult patients with a prior sternotomy and thus presumed higher complexity of disease; this finding suggests that pediatric CHD expertise in complex ACHD benefits these highly complex patients. Of the patients studied, 28.6% did not have a prior sternotomy. The database used does not provide detail about the anatomic diagnoses of these patients. However, these may include patients with congenitally corrected transposition of the great arteries, Ebstein anomaly, aortic coarctation, and other diagnoses that may still benefit from an input from a CHD team. Dedicated CHD expertise and infrastructure may improve outcomes of ACHD patients with advanced heart failure who are listed for transplantation, and these results further validate the 2018 ACHD guideline recommendation to refer these patients to comprehensive ACHD care centers ([15,16](#)).

Transplant mortality was lowest when performed at centers with high-volume ACHD transplant

programs, especially in the more contemporary period with growing surgical experience. The cumulative hazard of 1-year graft failure was 4.6% at the highest volume centers within UNOS regions compared with 8.5% in the rest of the transplant centers, suggesting that if all ACHD patients were transplanted at the highest volume ACHD transplant centers in the UNOS region, there could be a potential 3.9% reduction in the 1-year graft failure rate, which would have saved 41 patients in this study from post-transplant graft failure. ACHD patients have high surgical risk and often require aorta and pulmonary artery reconstruction as well as venous or arterial rerouting techniques, which may be more familiar to experienced congenital heart surgeons.

We found that a large number of centers perform ACHD transplant with very low frequency. In our study, the majority of transplant centers that perform ACHD transplantation averaged <1 ACHD transplant per year and, furthermore, almost 40% of transplant centers averaged <1 transplant every 4 to 5 years. It may therefore not be surprising that transplant outcomes were better when performed at the highest volume ACHD transplant centers within each UNOS region, which averaged 1.9 (± 0.8) ACHD transplants per year. We recognize that it may be difficult or not logistically possible to implement such a regionalization structure but the data suggest that ACHD post-transplant outcomes are improved at high-volume centers.

Whereas CHD patients make up a small minority of adult cardiology patients, they compose the majority of cardiac pathology seen at pediatric hospitals. Although most cardiology patients eventually transition their care to an adult cardiology center, a minority of patients continue their heart failure management at pediatric care centers. Transplant recipients at pediatric hospitals were on average 14 years younger than recipients at an adult hospital. The waitlist mortality for CHD patients with prior sternotomies was lower at pediatric hospitals, but the risk of graft failure was higher in patients transplanted at pediatric hospitals. Despite the younger average age at transplant, the reasons for increased graft failure rates at pediatric hospitals are unclear. Our data do not account for patient factors that may have compelled the care team to retain the patient in pediatric care, such as high disease complexity or genetic syndromes. In addition, the small number of pediatric transplant centers in our sample may have skewed our results: the experience of few centers will have a large effect size on overall pediatric outcomes. Given the lack of specific diagnosis information in the UNOS database, further study in this area is needed to

define the patient factors associated with care in an adult versus pediatric hospital.

Despite lower age and less traditional cardiovascular risk factors, CHD transplant recipients experience a high early post-transplant mortality resulting from prior thoracic surgeries, longer than average ischemic times, and a high prevalence of sensitization. Hence, controversies surrounding the timing of transplantation in CHD patients are ongoing and the use of this scarce resource in this high-risk population is not uniformly accepted (17,18). Although the immediate post-surgical outcomes may be worse given the surgical complexity, published results from experienced centers have shown excellent long-term outcomes (11,19-22). In a UNOS analysis by Menachem et al. (13), high overall heart transplant volumes were associated with improved transplant outcomes in congenital patients, and 30-day post-transplant mortality was 9.0% in the highest quartile volume centers of all heart transplants. We further show that adult CHD patients who are age ≥ 18 years may have even better post-transplant outcomes when managed by high-volume ACHD transplant centers with a 30-day mortality of 7.1% at the highest volume ACHD transplant center within each UNOS region and 7.9% at ACHA-accredited transplant centers. Thus, CHD-specific expertise in terms of ACHD transplant volume and ACHA accreditation has added value on top of high-volume heart transplant experience. Careful thought must be put into development of a robust infrastructure in the health care system for this growing adult population, and although it may be inconvenient from a travel and cost perspective, regionalizing advanced heart failure and transplant care for CHD patient may ensure a consistent high standard of care for this patient population.

STUDY LIMITATIONS. This was a retrospective study of a database registry that is subject to errors of data input and variable selection. The UNOS database lacks information on anatomy and disease complexity of ACHD patients who are purely stratified by having a CHD diagnosis, limiting our assessment of lesion-specific risk and mortality, and there is likely a high variability of pathology within our study cohort. Although the majority of patients with CHD were noted to have a prior sternotomy, we are not able to verify that the remainder of the cohort has not had cardiac surgery or the congenital diagnosis that did not warrant a childhood repair. There is no ideal method for the study of regionalization, so we therefore stratified the center experience in multiple ways to comment on an overarching hypothesis on center expertise. In our study, we clustered the center

volume and experience over the duration of the study period, and although we recognize that center expertise may change from year to year, we chose to sum the total volume for our analysis to reflect institutional history and experience. A sensitivity analysis was performed to ensure validity of this method in the most recent time period. In addition, ACHA accreditation was applied for the entire study period even though we chose to capture the status of ACHA accreditation at the end of 2018. Because the accreditation program was first established in 2017 and programs have gradually been reaching certification status over the past several years, results of ACHA accreditation analyses should therefore not be interpreted in isolation. In addition, ACHA accreditation likely under-represents programs with multidisciplinary expertise because the program was established just 2 years ago and our results may therefore underestimate the difference in outcomes. We were not able to account for the confounding effect of specific providers on the outcomes of interest.

CONCLUSIONS

Significant variation in congenital heart transplant volume and experience exists between centers. Center CHD expertise, as with ACHA-accredited centers, is associated with lower waitlist mortality for adult CHD patients. Finally, heart transplants at the highest ACHD volume center in a UNOS region were associated with a significantly lower risk of graft failure. Our study supports the development of regionalization models of transplant care for these high-risk ACHD patients.

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PERSPECTIVES

COMPETENCY IN SYSTEMS-BASED PRACTICE: Adults with congenital heart disease awaiting cardiac transplantation at centers accredited by the Adult Congenital Heart Association have lower mortality rates and better post-transplant outcomes than those at lower volume centers.

TRANSLATIONAL OUTLOOK: Further efforts are needed to expand access of patients with congenital heart disease to centers with special expertise and regionalized systems of transplant care.

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