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ORIGINAL RESEARCH

Adherence and Exercise Capacity Improvements of Patients With Adult Congenital Heart Disease Participating in Cardiac Rehabilitation

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BACKGROUND: As the number of adults with congenital heart disease increases because of therapeutic advances, cardiac rehabilitation (CR) is increasingly being used in this population after cardiac procedures or for reduced exercise tolerance. We aim to describe the adherence and exercise capacity improvements of patients with adult congenital heart disease (ACHD) in CR.

METHODS AND RESULTS: This retrospective study included patients with ACHD in CR at New York University Langone Rusk Rehabilitation from 2013 to 2020. We collected data on patient characteristics, number of sessions attended, and functional testing results. Pre-CR and post-CR metabolic equivalent task, exercise time, and maximal oxygen uptake were assessed. In total, 89 patients with ACHD (mean age, 39.0 years; 54.0% women) participated in CR. Referral indications were reduced exercise tolerance for 42.7% and post–cardiac procedure (transcatheter or surgical) for the remainder. Mean number of sessions attended was 24.2, and 42 participants (47.2%) completed all 36 CR sessions. Among participants who completed the program as well as pre-CR and post-CR functional testing, metabolic equivalent task increased by 1.3 (95% CI, 0.7–1.9; baseline mean, 8.1), exercise time increased by 66.4 seconds (95% CI, 21.4–111.4 seconds; baseline mean, 536.1 seconds), and maximal oxygen uptake increased by 2.5 mL/kg per minute (95% CI, 0.7–4.2 mL/kg per minute; baseline mean, 20.2 mL/kg per minute).

CONCLUSIONS: On average, patients with ACHD who completed CR experienced improvements in exercise capacity. Efforts to increase adherence would allow more patients with ACHD to benefit.

Key Words: adult congenital heart disease ■ cardiac rehabilitation ■ prevention

Congenital heart disease affects ≈1% of live births, which corresponds to 1.35 million cases annually worldwide.[1] As progress is made in the diagnosis and medical and surgical management of these conditions, more patients are surviving into adulthood.[2] Consequently, the prevalence of adult congenital heart disease (ACHD) is increasing, and in 2010, adults represented 66% of the total population with congenital heart disease, demonstrating that there are more adults than children living with congenital heart disease.[3] Despite increased survival, 4 in 5 patients with ACHD have worse cardiopulmonary function, as measured by peak oxygen uptake, compared with the general population.[4] Exercise intolerance is associated with increased hospitalization or death in patients with ACHD.[5]

Cardiac rehabilitation (CR) improves exercise capacity and is commonly prescribed for patients with a wide range of cardiovascular conditions, including coronary artery disease, peripheral artery disease, heart failure, heart transplant, and valve replacement.[6] In

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CLINICAL PERSPECTIVE

What Is New?

- Approximately half of patients with adult congenital heart disease participating in cardiac rehabilitation completed the entire 36-session program.
- Patients with adult congenital heart disease who completed cardiac rehabilitation, on average, experienced statistically significant improvements in exercise capacity, as measured by metabolic equivalent, exercise time, and maximal oxygen uptake in this heterogeneous sample.

What Are the Clinical Implications?

- Patients with adult congenital heart disease benefit from cardiac rehabilitation and should be referred after cardiac procedures or for reduced exercise tolerance.
- Opportunities exist in health care delivery for improving adherence to cardiac rehabilitation for patients with adult congenital heart disease.

Nonstandard Abbreviations and Acronyms

ACC/AHA	American College of Cardiology/ American Heart Association
ADHD	adult congenital heart disease
CR	cardiac rehabilitation
MET	metabolic equivalent task
VO_2 max	maximal oxygen uptake

clinical trials, CR has been demonstrated to decrease hospital admissions and cardiovascular mortality in patients with coronary artery disease or heart failure.[6–8] Typical CR programs consist of 36 supervised exercise sessions as well as counseling for lifestyle modification and psychosocial support. In 2018, the American College of Cardiology/American Heart Association (ACC/AHA) designated a class IIa indication (level of evidence B-NR) for using CR to improve exercise capacity in patients with ACHD.[9] However, studies of patients with congenital heart disease in CR are still limited and often included children or were performed for specific congenital heart conditions. In response to this gap in knowledge, we determined adherence and exercise capacity improvements for a heterogeneous population of patients with ACHD participating in CR at an urban academic medical center.

METHODS

This retrospective study included all patients with ACHD who participated in at least 1 CR session at New

Table 1. Baseline Characteristics of Patients With ACHD Participating in CR and Those Completing the Program

Characteristic	All Participants (n=89)	Completed CR (n=42)	Did not complete CR (n=47)	*P* value
Demographics				
Age, y	39.0±12.7	39.3±12.9	38.7±12.6	0.842
Female sex	48 (54.0)	24 (57.1)	24 (51.1)	0.671
Non-White race[†]	40 (44.9)	20 (47.6)	20 (42.6)	0.674
Hispanic ethnicity	24 (27.0)	12 (28.6)	12 (25.5)	0.813
Medicare/ Medicaid	25 (28.1)	16 (38.1)	9 (19.2)	0.060
Work/school	64 (71.9)	26 (61.9)	38 (80.9)	0.060
ACHD characteristics				
Anatomic classification				
Simple	8 (9.0)	1 (2.4)	7 (14.9)	0.099
Moderate	56 (62.9)	27 (64.3)	29 (61.7)	
Complex	25 (28.1)	14 (33.3)	11 (23.4)	
Physiologic classification				
Stage A	8 (9.0)	2 (4.8)	6 (12.8)	0.134
Stage B	41 (46.1)	17 (40.5)	24 (51.1)	
Stage C	36 (40.5)	22 (52.4)	14 (29.8)	
Stage D	4 (4.5)	1 (2.4)	3 (6.4)	
Valve disease	62 (69.7)	30 (71.4)	32 (68.1)	0.819
Arrhythmia	31 (34.8)	16 (38.1)	15 (31.9)	0.657
PPM	16 (18.0)	10 (23.8)	6 (12.8)	0.269
RV dysfunction (missing 13)	22 (28.9)	14 (36.8)	8 (21.1)	0.148
Pulmonary hypertension (missing 39)	12 (24.0)	5 (21.7)	7 (25.9)	0.957
Shunt	13 (14.6)	6 (14.3)	7 (14.9)	1.000
Cyanosis	3 (3.4)	2 (4.8)	1 (2.1)	0.600
Comorbidities				
Coronary artery disease	9 (10.1)	4 (9.5)	5 (10.6)	1.000
Diabetes	4 (4.5)	1 (2.4)	3 (6.4)	0.619
Hypertension	27 (30.3)	10 (23.8)	17 (36.2)	0.252
Hyperlipidemia	19 (21.4)	7 (16.7)	12 (25.5)	0.438
Heart failure	14 (15.7)	6 (14.3)	8 (17.0)	0.778
Overweight or obese BMI	26 (29.2)	13 (31.0)	13 (27.7)	0.817
Depression	9 (10.1)	3 (7.1)	6 (12.8)	0.491
Stroke	6 (6.7)	4 (9.5)	2 (4.3)	0.415
Current/ former smoker	19 (21.4)	13 (31.0)	6 (12.8)	0.042*
Biometrics				
BMI, kg/m^2	26.6±7.2	26.9±7.6	26.4±6.9	0.745
CrCl (missing 14), mL/min	125.1±49.3	126.0±54.9	124.4±45.3	0.892
LVEF (missing 12), %	57.4±10.0	56.9±11.9	57.9±7.9	0.653
Medications				
Aspirin	53 (59.6)	25 (59.5)	28 (59.6)	1.000

(*Continued*)

Table 1. Continued

Characteristic	All Participants (n=89)	Completed CR (n=42)	Did not complete CR (n=47)	*P* value
ACE inhibitor/ ARB	35 (39.3)	18 (42.9)	17 (36.2)	0.664
β Blocker	52 (58.4)	24 (57.1)	28 (59.6)	0.833
Warfarin	14 (15.7)	7 (16.7)	7 (14.9)	1.000
DOAC	6 (6.7)	2 (4.8)	4 (8.5)	0.680
P2Y12 inhibitor	4 (4.5)	0 (0.0)	4 (8.5)	0.119
Diuretic	25 (28.1)	11 (26.2)	14 (29.8)	0.815
Statin	12 (13.5)	5 (11.9)	7 (14.9)	0.763
Pulmonary vasodilator	1 (1.1)	0 (0.0)	1 (2.1)	1.000
Referral indication				
Postprocedural	51 (57.3)	20 (47.6)	31 (66.0)	0.091
Reduced exercise tolerance	38 (42.7)	22 (52.4)	16 (34.0)	

Data are given as mean±SD or number (percentage). ACE indicates angiotensin-converting enzyme; ACHD, adult congenital heart disease; ARB, angiotensin receptor blocker; BMI, body mass index; CR, cardiac rehabilitation; CrCl, creatinine clearance; DOAC, direct oral anticoagulant; LVEF, left ventricular ejection fraction; PPM, permanent pacemaker; and RV, right ventricular.

*Statistically significant at *P* = 0.05.

†Non-White race included individuals selfidentifying as Asian, Black, or Other on clinical documentation.

York University (NYU) Langone Rusk Rehabilitation from January 2013 through December 2020. Patients were referred after cardiac procedure (surgical or catheter-based intervention within 1 year before starting CR) or for reduced exercise tolerance (based on cardiopulmonary exercise testing). Some patients with ACHD participated in >1 course of CR, but only data from the first CR course were included. This study was approved by the institutional review board of NYU Langone Health. Informed consent was not required from participants.

Demographic information, ACHD condition characteristics, comorbidities, biometrics, cardiovascular medication use, and referral indication were extracted for each participant from the electronic health record system. ACHD condition was classified anatomically as simple, moderate, or complex, and physiologically as stage A (most healthy), B, C, or D (least healthy) based on the ACC/AHA ACHD anatomic and physiologic classification system.[9] The presence of arrhythmia, permanent pacemaker, coronary artery disease, diabetes, hypertension, hyperlipidemia, heart failure, depression, prior stroke, and smoking history was collected from clinical documentation. Body mass index and presence of cyanosis were determined on the basis of vital signs at the CR intake session. The intake form also inquired about whether participants were in school or working. Overweight or obese body mass index was defined as body mass index of >25 kg/m^2. Creatine clearance was calculated on the basis of the laboratory measurement closest in time to the first CR session. Data on left ventricular ejection fraction, right ventricular dysfunction, pulmonary hypertension, valve disease, and shunts were abstracted from the echocardiogram or cardiac magnetic resonance imaging scan closest in time to the first CR session. Laboratory and imaging results >1 year from the first CR session were excluded. Use of cardiovascular medications,

Table 2. Frequency of Congenital Conditions for Patients With ACHD in CR

Congenital heart disease condition*	Frequency	% of Patients
Anomalous coronary artery	3	3.4
Anomalous coronary artery post-repair	3	3.4
ASD	8	9.0
Atrioventricular canal post-repair and heterotaxy	1	1.1
Bicuspid aortic valve with coarctation of aorta	2	2.2
Bicuspid aortic valve	1	1.1
Bicuspid aortic valve post-AVR	9	10.1
Coarctation of the aorta	6	6.7
Congenital heart block	1	1.1
Congenital aortic valve disease post-repair	1	1.1
Congenital mitral valve disease	2	2.2
Congenital multivalve disease	1	1.1
D-TGA post-ASO	4	4.5
D-TGA post-Mustard or Senning	5	5.6
D-TGA post-Rastelli	1	1.1
Double-chamber right ventricle	1	1.1
Double-outlet ventricle s/p repair	2	2.2
Ebstein	3	3.4
Fontan procedure	7	7.9
L-TGA	1	1.1
L-TGA post-Senning/Rastelli	1	1.1
Pulmonary atresia	2	2.2
Pulmonary stenosis	3	3.4
PAPVR post-repair	3	3.4
Scimitar syndrome	1	1.1
Shone syndrome	1	1.1
TOF	14	15.7
Transitional atrioventricular canal	1	1.1
VSD	1	1.1

ACHD indicates adult congenital heart disease; ASD, atrial septal defect; ASO, arterial switch operation; AVR, aortic valve replacement; CR, cardiac rehabilitation; D-TGA, dextro-transposition of the great arteries; L-TGA, levo-transposition of the great arteries; PAPVR, partial anomalous pulmonary venous return; TOF, tetralogy of Fallot; and VSD, ventricular septal defect.

*Congenital heart disease conditions are unrepaired unless otherwise noted.

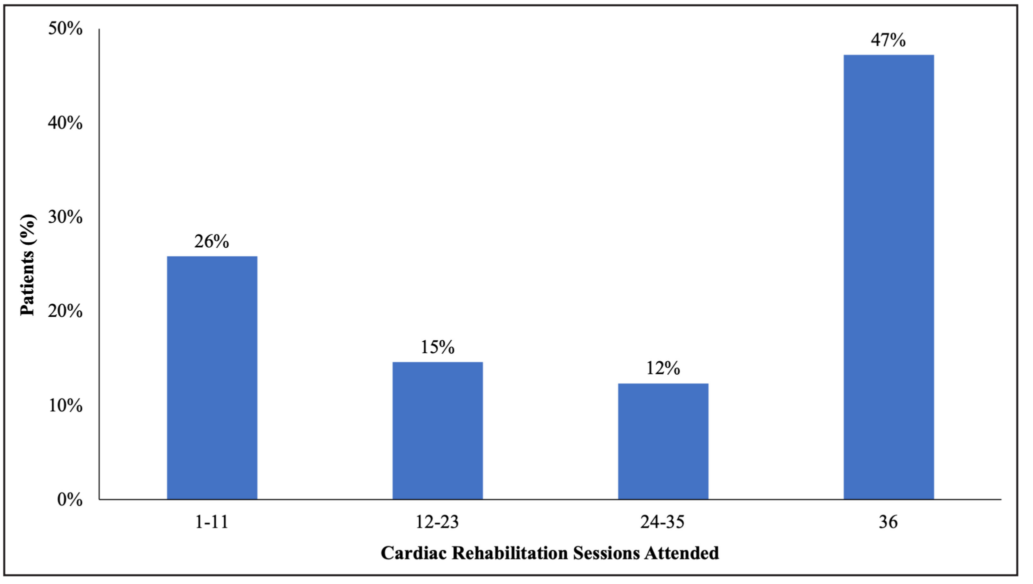

Figure 1. Distribution of patients with adult congenital heart disease by number of cardiac rehabilitation sessions attended.

including aspirin, angiotensin-converting enzyme inhibitor, angiotensin receptor blocker, β blocker, warfarin, direct oral anticoagulant, P2Y12 inhibitor, diuretic, statin, or pulmonary vasodilator, was collected from clinical documentation.

The number of CR sessions attended was abstracted and divided into groups of 1 to 11, 12 to 23, 24 to 35, or 36 sessions. Participants underwent exercise stress testing by either Bruce or modified Bruce protocol and cardiopulmonary exercise testing before and after CR. Metabolic equivalent task (MET) and exercise time were assessed by exercise stress test, whereas maximal oxygen uptake (VO_2 max) was assessed by cardiopulmonary exercise test. Not all individuals were able to complete post-CR functional testing because of patient-specific factors or logistical reasons.

Statistical Analysis

Baseline characteristics of participants who completed all 36 CR sessions were compared with those of participants who did not complete the program using 2-sample *t* tests for continuous variables and Fisher exact tests for categorical variables. Given limited sample sizes, race was categorized as White versus non-White (consisting of individuals who were Black, Asian, or had race not identified on clinical documentation). For participants who completed all 36 CR sessions and both pre– and post–exercise stress test or cardiopulmonary exercise test, changes in MET, exercise time, and VO_2 max before and after CR were assessed using paired-sample *t* tests. Similarly, changes in MET, exercise time, and VO_2 max before and after CR were compared between patients with simple or moderate versus complex congenital heart conditions and between patients with physiologic stages A or B versus C or D congenital heart conditions using 2-sample *t* tests. (Certain categories in above comparisons were combined because of limited sample sizes.) Finally, changes in MET, exercise time, and VO_2 max before and after CR were also compared in a subgroup analysis after excluding patients meeting alternative indications for cardiac rehabilitation, mainly concurrent heart failure or recent valve surgery. An increase of at least 1 MET was set as the threshold for clinically significant improvement because that amount has been shown to correspond to a mortality reduction of 15%, which is comparable to the mortality benefit of common cardiovascular medications prescribed after myocardial infarction.[10] A subset of baseline characteristics, which were chosen on the basis of sample size and on likelihood of affecting exercise tolerance, of participants who improved by at least 1 MET after completing all 36 CR sessions were compared with those of participants who did not improve by at least 1 MET using Fisher exact tests. Threshold for statistical significance was set at P=0.05. Statistical analyses were performed using STATA 16.1 (Statacorp, College Station, TX).

RESULTS

Our sample included 89 consecutive patients with ACHD who participated in at least 1 CR session from January 2013 through December 2020. Mean age was 39.0±12.7 years (minimum, 19 years; maximum, 69 years), 54.0% were women, and 71.9% were in school or working. Self-identified race of patients was 55.1% White individuals, 10.1% Black individuals, 13.5% Asian individuals, and 21.3% other individuals

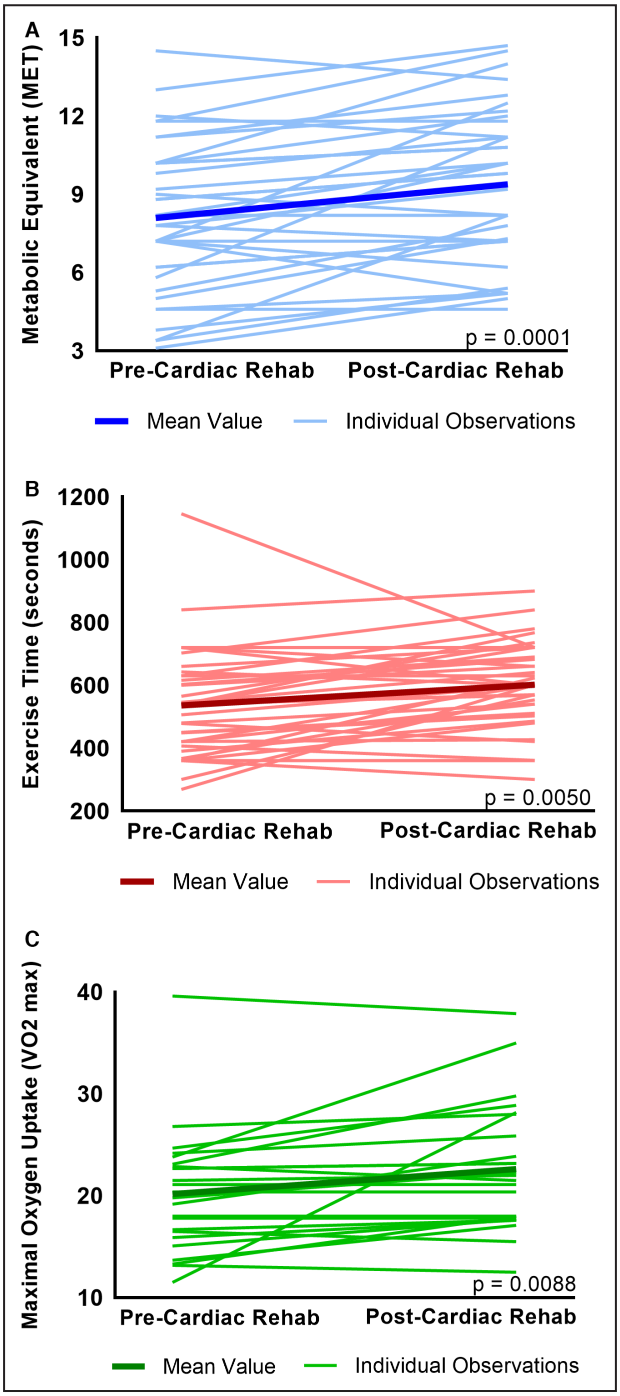

Figure 2. A, Metabolic equivalent task (MET). B, Exercise time. C, Maximal oxygen uptake (VO_2 max) before and after cardiac rehabilitation (rehab) for patients with adult congenital heart disease who completed the program.

P values are based on paired-sample *t* tests assessing pre–cardiac rehab and post–cardiac rehab values.

(including those with race not identified in clinical documentation). ACHD conditions were classified anatomically as simple for 9.0%, moderate for 62.9%, and complex for 28.1% (Table 1). The most common simple condition was atrial septal defect. The most common moderate conditions were tetralogy of Fallot, bicuspid aortic valve post-valve replacement, and coarctation of the aorta. The most common complex conditions were post-Fontan procedure and dextro-transposition of great arteries post–arterial switch operation or Mustard and Senning repair (Table 2). ACHD conditions were also classified physiologically as stage A for 9.0%, stage B for 46.1%, stage C for 40.5%, and stage D for 4.5%. Congenital heart conditions frequently involved valve disease (69.7%) or arrhythmia (34.8%). Common comorbidities affecting patients included hypertension (30.3%), overweight or obese body mass index (29.2%), and current or former smoking history (21.4%). Most patients (76.4%) received primary cardiology care at our institution, whereas the remaining patients (23.6%) were from outside academic institutions or private practice. Referral indications were post-cardiac procedure (transcatheter or surgical) for 57.3% and reduced exercise tolerance for the remaining 42.7% (Table 1).

Table 3. Baseline and Improvement in Exercise Capacity of Patients With ACHD Who Completed CR

Metric	Baseline, mean±SD	Difference (95% CI)
Metabolic equivalent	8.1±2.9	1.3 (0.7–1.9)
Exercise time, s	536.1±172.0	66.4 (21.4–111.4)
Maximal oxygen uptake, mL/kg per min	20.2±5.9	2.5 (0.7–4.2)

Sample sizes: n=36 for metabolic equivalent and exercise time, and n=24 for maximal oxygen uptake. ACHD indicates adult congenital heart disease; and CR, cardiac rehabilitation.

Patients with ACHD completed on average 24.2 CR sessions. Full adherence to all 36 CR sessions was observed for 42 participants (47.2%) (Figure 1). Although the CR program was paused from mid-March through May 2020 because of the COVID-19 pandemic, adherence was not markedly affected. When excluding participants from 2020, full adherence to the CR program was observed for 48.7% of participants. Among patients not completing CR, the following reasons were most commonly cited: personal reason (30 patients [63.8%]), work (6 patients [12.8%]), and medical reason (3 patients [6.4%]). Mean age was 39.3 years for patients who completed CR and 38.7 years for patients who did not complete the program. Being a current or former smoker was the only baseline patient characteristic associated with CR completion (Table 1).

Among the 42 participants who completed CR, exercise stress test results were available for 36 (86%), and cardiopulmonary exercise test results were available for 24 (57%). When comparing pre-CR and post-CR functional test results, statistically significant improvements were observed in MET by 1.3 (95% CI, 0.7–1.9; baseline, 8.1), exercise time by 66.4 seconds (95% CI, 21.4–111.4 seconds; baseline, 536.1 seconds), and VO_2 max by 2.5 mL/kg per minute (95% CI, 0.7–4.2 mL/kg per minute; baseline,

Table 4. Baseline and Improvement in Exercise Capacity of Patients With ACHD Who Completed CR for Simple or Moderate Versus Complex and for Stage A or B Versus Stage C or D Congenital Heart Disease Conditions Based on ACC/AHA ACHD Anatomic and Physiologic Classification System[9]

Metric	Simple/moderate		Complex		*P* value*
	Baseline, mean±SD	Difference (95% CI)	Baseline, mean±SD	Difference (95% CI)	
Metabolic equivalent	8.1±3.4	1.5 (0.6 to 2.4)	8.1±2.2	1.0 (0.3 to 1.6)	0.38
Exercise time, s	527.5±204.9	73.6 (1.5 to 145.8)	549.5±106.9	55.0 (15.7 to 94.3)	0.69
Maximal oxygen uptake, mL/kg per min	20.2±4.8	4.0 (0.3 to 7.8)	20.2±6.8	1.1 (−0.01 to 2.3)	0.09
	Stage A/B		Stage C/D		
	Baseline, mean±SD	Difference (95% CI)	Baseline, mean±SD	Difference (95% CI)	*P* value*
Metabolic equivalent	8.5±3.4	1.9 (0.5 to 3.3)	7.9±2.7	1.0 (0.4 to 1.5)	0.13
Exercise time, s	588.9±223.2	41.6 (−67.4 to 150.7)	506.2±131.3	80.4 (37.6 to 123.2)	0.41
Maximal oxygen uptake, mL/kg per min	20.9±8.5	3.5 (−1.6 to 8.5)	19.8±4.4	2.0 (0.4 to 3.6)	0.42

Sample sizes: n=22 for metabolic equivalent and exercise time among simple/moderate, n=14 for metabolic equivalent and exercise time among complex; n=11 for maximal oxygen uptake among simple/moderate, n=13 for maximal oxygen uptake among complex; n=13 for metabolic equivalent and exercise time among stage A/B, n=23 for metabolic equivalent and exercise time among stage C/D; and n=8 for maximal oxygen uptake among stage A/B, n=16 for maximal oxygen uptake among stage C/D. ACC/AHA indicates American College of Cardiology/American Heart Association; ACHD, adult congenital heart disease; and CR, cardiac rehabilitation.

**P* values reflect comparisons between differences in each metric for simple/moderate vs complex and stage A/B vs stage C/D congenital heart disease conditions.

20.2 mL/kg per minute) (Figure 2 and Table 3). The degree of improvement based on the above metrics did not vary by congenital heart disease severity defined by ACC/AHA ACHD anatomic and physiologic classifications (Table 4). In a subgroup analysis excluding patients with ACHD meeting alternative CR indications, mainly concurrent heart failure or recent valve surgery, statistically significant improvements were observed in MET by 0.7 (95% CI, 0.2–1.3; baseline, 9.2) and exercise time by 49.1 seconds (95% CI, 12.0–86.3 seconds; baseline, 538.1 seconds). VO_2 max improved by 1.6 mL/kg per minute but did not meet statistical significance (95% CI, −0.1 to 3.3 mL/kg per minute; baseline, 21.4 mL/kg per minute) (Table S1). No statistically significant associations were observed between baseline patient characteristics and physiologic improvement of at least 1 MET after completing CR (Table 5).

DISCUSSION

Our study shows that patients with ACHD participating in CR were demographically diverse. Almost all patients had moderate or complex and physiologic stage B or C ACHD conditions. Nearly two thirds of these patients were referred after cardiac procedures, whereas the remainder were referred for reduced exercise tolerance. Only half of participants completed the entire 36-session program, but those who were fully adherent experienced statistically significant improvements in exercise capacity, as measured by MET, exercise time, and maximal oxygen uptake.

In our sample, approximately two thirds of participants with ACHD were referred to CR after surgical or transcatheter procedures. Similarly, a retrospective study based on data from a separate CR program determined that referral indications for patients with ACHD in their program were post-surgical for 61% and post-transplant for 9% of patients.[11] These trends reflect the common need for procedural intervention among patients with ACHD but may also suggest underuse of CR among patients with reduced exercise tolerance but without need for procedural intervention. An analysis of a Dutch national registry showed that 1 in 5 patients with ACHD required surgery over a 15-year period.[12] Meanwhile, reduced cardiopulmonary function is more prevalent and affects 4 in 5 patients with ACHD.[4] Participation in CR requires a multistep process (eg, physician referral and program enrollment), and there may be opportunities at each step to increase use among patients with ACHD with reduced exercise tolerance. For example, ACC/AHA recommends using cardiopulmonary exercise testing for assessing functional capacity in patients with ACHD.[9] Prior data have shown that asymptomatic patients with ACHD, despite not reporting symptoms of exercise intolerance, have significantly lower peak oxygen uptake compared with healthy patients (peak VO_2 for asymptomatic patients with ACHD versus healthy patients, 26.1±8.2 versus 45.1±8.6 mL/kg per minute; P<0.0001), which suggests that reduced exercise tolerance is underdiagnosed among patients with ACHD.[5] Therefore, application of cardiopulmonary exercise testing may enhance detection of reduced exercise tolerance, resulting in higher numbers of patients with ACHD with reduced exercise tolerance being referred to CR.

Table 5. Number of Patients With ACHD Whose Exercise Capacity Improved by at Least 1 MET Among Those Who Completed CR, by Patient Characteristic

Patient characteristic	Completed CR	Improved, n (%)	*P* value
Sex			
Women	20	12 (60.0)	0.731
Men	16	11 (68.8)	
Race			
Non-Whit†	17	9 (52.9)	0.299
White	19	14 (73.7)	
Ethnicity			
Hispanic	11	8 (72.7)	0.708
Not Hispanic	25	15 (60.0)	
BMI			
Overweight or obese	13	9 (69.2)	0.727
Normal	23	14 (60.9)	
Insurance			
Medicare/Medicaid	14	7 (50.0)	0.286
Private	22	16 (72.7)	
Work/school			
Yes	21	15 (71.4)	0.310
No	15	8 (53.3)	
Valve disease			
Yes	25	17 (68.0)	0.475
No	11	6 (54.6)	
Arrhythmia			
Yes	15	10 (66.7)	1.000
No	21	13 (61.9)	
Heart failure			
Yes	6	6 (100.0)	0.068
No	30	17 (56.7)	
Cardiovascular medications*			
≤2	21	12 (57.1)	0.484
>2	15	11 (73.3)	
Referral Indication			
Post–cardiac procedure	15	9 (60.0)	0.736
Reduced exercise tolerance	21	14 (66.7)	

ACHD indicates adult congenital heart disease; BMI, body mass index; CR, cardiac rehabilitation; and MET, metabolic equivalent task.

*Number of cardiovascular medications was summed from aspirin, angiotensin-converting enzyme inhibitor/angiotensin receptor blocker, β blocker, warfarin, direct oral anticoagulant, P2Y12 inhibitor, diuretic, statin, and pulmonary vasodilator.

†Non-White race included individuals selfidentifying as Asian, Black, or Other on clinical documentation.

Adherence to CR is a recognized barrier to the effectiveness of the program. A dose-response relationship exists between the number of sessions attended and patient outcomes. In patients with coronary artery disease, increased number of sessions attended is associated with lower mortality, cardiac events, and hospitalizations.[13,14] A prior study based on our institutional data found that percentages of participants completing CR is estimated to be 42.9% among patients with heart failure and 54.9% among patients with coronary artery disease.[15] This current study revealed that 47.2% of patients with ACHD completed the program, which is similar to the percentages of patients with heart failure and coronary artery disease, despite patients with ACHD being younger with fewer comorbidities. Prior studies have shown that female sex and increased comorbidities are associated with lower adherence to CR, whereas the effects of age and socioeconomic status are mixed.[16] Proposed solutions for increasing CR adherence include material or financial rewards and home-based CR.[17]

Our study demonstrates that patients with ACHD who complete CR experience statistically significant improvements in exercise capacity. In our sample, MET increased by 1.3, and exercise time increased by 1.1 minutes, which are comparable to an increase in MET of 1.6 and in exercise time of 1.35 minutes in a study of adolescent and adult patients with congenital heart disease.[11] Similarly, we also observed an increase in peak VO_2 of 2.5 mL/kg per minute, which is comparable to an increase in peak VO_2 of 1.96 mL/kg per minute from a meta-analysis of patients with ACHD in center- and home-based CR programs.[18] If these physiologic improvements are sustained over time, CR will likely lead to improved patient outcomes, especially because a 1-MET increase in exercise capacity has been associated with a decrease in mortality of ≈15% in a healthy adult population.[10,19]

Our sample was diverse in both demographic and clinical characteristics as well as type and severity of congenital heart disease conditions, which reinforces the external validity of the results. The degree of improvement in physiologic parameters did not differ by congenital heart disease severity, as defined by ACC/AHA ACHD anatomic and physiologic classifications, although the limited sample size may have lacked the statistical power to detect such differences. Patients with ACHD with existing heart failure or recent valve surgery are already indicated for CR for those reasons, but even when excluding those patients, statistically significant improvements were observed in MET and exercise time but not in VO_2 max, although this analysis was also limited by the low statistical power of the small sample size. These findings suggest that all patients with ACHD who complete CR may benefit from the program.

There are several limitations to our study. Given limitations in data availability, we were unable to answer broader questions on use (eg, the percentage of patients with ACHD eligible for CR who were referred), which may be associated with social determinants of health. Only 57% of patients who completed CR also completed pre-cardiopulmonary exercise testing and post-cardiopulmonary exercise testing, which may have introduced ascertainment bias when assessing improvements in VO_2 max. Because of limited sample size, we were unable to perform regression analyses to control for baseline characteristics when examining which baseline characteristics were associated with CR completion and improvement in exercise capacity. Because CR in general does not involve long-term follow-up, we were unable to determine if improvements in exercise capacity were sustained over time. Our CR program also consists of patients with ACHD from outside of our health care system, and because outside records were not available, we were unable to analyze the effect of CR on subsequent outcomes, such as hospital admissions and mortality.

In conclusion, this study of patients with ACHD in CR demonstrates that although adherence remains an issue similar to patients referred to CR for other indications, patients with ACHD who complete the program experience improvements in exercise capacity. Further research is required to determine how adherence can be improved among this younger CR population and whether exercise capacity improvements are associated with reductions in hospitalizations and mortality in patients with ACHD.

ARTICLE INFORMATION

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Disclosures

None.

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Supplemental Material

Table S1

REFERENCES

1. van der Linde D, Konings EE, Slager MA, Witsenburg M, Helbing WA, Takkenberg JJ, Roos-Hesselink JW. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. *J Am Coll Cardiol*. 2011;58:2241–2247. doi: 10.1016/j.jacc.2011.08.025
2. Triedman JK, Newburger JW. Trends in congenital heart disease: the next decade. *Circulation*. 2016;133:2716–2733. doi: 10.1161/CIRCULATIONAHA.116.023544
3. Marelli AJ, Ionescu-Ittu R, Mackie AS, Guo L, Dendukuri N, Kaouache M. Lifetime prevalence of congenital heart disease in the general population from 2000 to 2010. *Circulation*. 2014;130:749–756. doi: 10.1161/CIRCULATIONAHA.113.008396
4. Kempny A, Dimopoulos K, Uebing A, Moceri P, Swan L, Gatzoulis MA, Diller GP. Reference values for exercise limitations among adults with congenital heart disease. Relation to activities of daily life – single centre experience and review of published data. *Eur Heart J*. 2012;33:1386–1396. doi: 10.1093/eurheartj/l461
5. Diller GP, Dimopoulos K, Okonko D, Li W, Babu-Narayan SV, Broberg CS, Johansson B, Bouzas B, Mullen MJ, Poole-Wilson PA, et al. Exercise intolerance in adult congenital heart disease: comparative severity, correlates, and prognostic implication. *Circulation*. 2005;112:828–835. doi: 10.1161/CIRCULATIONAHA.104.529800
6. McMahon SR, Ades PA, Thompson PD. The role of cardiac rehabilitation in patients with heart disease. *Trends Cardiovasc Med*. 2017;27:420–425. doi: 10.1016/j.tcm.2017.02.005
7. Anderson L, Thompson DR, Oldridge N, Zwisler AD, Rees K, Martin N, Taylor RS. Exercise-based cardiac rehabilitation for coronary heart disease. *Cochrane Database Syst Rev*. 2016;2016:CD001800. doi: 10.1002/14651858.CD001800.pub3
8. O'Connor CM, Whellan DJ, Lee KL, Keteyian SJ, Cooper LS, Ellis SJ, Leifer ES, Kraus WE, Kitzman DW, Blumenthal JA, et al. Efficacy and safety of exercise training in patients with chronic heart failure: HF-ACTION randomized controlled trial. *JAMA*. 2009;301:1439–1450.
9. Stout KK, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS, Colman JM, Crumb SR, Dearani JA, Fuller S, Gurvitz M, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association task force on clinical practice guidelines. *Circulation*. 2019;139:e698–e800. doi: 10.1161/CIR.0000000000000603
10. Franklin BA, Brinks J, Berra K, Lavie CJ, Gordon NF, Sperling LS. Using metabolic equivalents in clinical practice. *Am J Cardiol*. 2018;121:382–387. doi: 10.1016/j.amjcard.2017.10.003
11. Sarno LA, Misra A, Siddeek H, Kheiwa A, Kobayashi D. Cardiac rehabilitation for adults and adolescents with congenital heart disease: extending beyond the typical patient population. *J Cardiopulm Rehabil Prev*. 2020;40:E1–E4. doi: 10.1097/HCR.0000000000000482
12. Zomer AC, Verheugt CL, Vaartjes I, Uiterwaal CS, Langemeijer MM, Koolbergen DR, Hazekamp MG, van Melle JP, Konings TC, Bellersen L, et al. Surgery in adults with congenital heart disease. *Circulation*. 2011;124:2195–2201. doi: 10.1161/CIRCULATIONAHA.111.027763
13. Martin BJ, Hauer T, Arena R, Austford LD, Galbraith PD, Lewin AM, Knudtson ML, Ghali WA, Stone JA, Aggarwal SG. Cardiac rehabilitation attendance and outcomes in coronary artery disease patients. *Circulation*. 2012;126:677–687. doi: 10.1161/CIRCULATIONAHA.111.066738
14. Doll JA, Hellkamp A, Thomas L, Ho PM, Kontos MC, Whooley MA, Boyden TF, Peterson ED, Wang TY. Effectiveness of cardiac rehabilitation among older patients after acute myocardial infarction. *Am Heart J*. 2015;170:855–864. doi: 10.1016/j.ahj.2015.08.001
15. Bostrom J, Searcy R, Walia A, Rzucidlo J, Banco D, Quien M, Sweeney G, Pierre A, Tang Y, Mola A, et al. Early termination of cardiac rehabilitation is more common with heart failure with reduced ejection fraction than with ischemic heart disease. *J Cardiopulm Rehabil Prev*. 2020;40:E26–E30. doi: 10.1097/HCR.0000000000000495
16. Ruano-Ravina A, Pena-Gil C, Abu-Assi E, Raposeiras S, van't Hof A, Meindersma E, Bossano Prescott EI, Gonzalez-Juanatey JR. Participation and adherence to cardiac rehabilitation programs. A systematic review. *Int J Cardiol*. 2016;223(436–443):436–443. doi: 10.1016/j.ijcard.2016.08.120
17. Rubin R. Although cardiac rehab saves lives, few eligible patients take part. *JAMA*. 2019;322:386–388. doi: 10.1001/jama.2019.8604
18. Li X, Chen N, Zhou X, Yang Y, Chen S, Song Y, Sun K, Du Q. Exercise training in adults with congenital heart disease: a systematic review and meta-analysis. *J Cardiopulm Rehabil Prev*. 2019;39:299–307. doi: 10.1097/HCR.0000000000000420
19. Kodoma S, Saito K, Tanaka S, Maki M, Yachi Y, Asumi M, Sugawara A, Totsuka K, Shimano H, Ohashi Y, et al. Cardiorespiratory fitness as a quantitative predictor of all-cause mortality and cardiovascular events in healthy men and women: a meta-analysis. *JAMA*. 2009;301:2024–2035. doi: 10.1001/jama.2009.681

SUPPLEMENTAL MATERIAL

Table S1. Baseline and improvement in exercise capacity of adult congenital heart disease patients who completed cardiac rehabilitation, excluding patients meeting alternative indications for cardiac rehabilitation*

Metric	Baseline	Difference (95% CI)
Metabolic Equivalent (MET)	9.2±2.1	0.7 (0.2 – 1.3)
Exercise Time (seconds)	538.1±122.5	49.1 (12.0 – 86.3)
Maximal Oxygen Uptake (mL/kg/min, VO2 max)	21.4±6.3	1.6 (-0.1 – 3.3)

Sample sizes: n=21 for metabolic equivalent and exercise time, n=16 for maximal oxygen uptake.
*Alternative indications for which patients were excluded were mostly clinical diagnosis of chronic heart failure or recent valve surgery.