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ORIGINAL RESEARCH

Association of Adults With Congenital Heart Disease–Specific Care With Clinical Characteristics and Healthcare Use

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BACKGROUND: Many adults with congenital heart disease (ACHD) are cared for by non-ACHD specialists, if they receive care at all. Little is known about the differences between those who access care at an ACHD center and those who do not access ACHD-specific care.

METHODS AND RESULTS: The Oregon All Payer All Claims database was queried to identify subjects aged 18 to 65 years with an *International Classification of Diseases, Ninth Revision* (*ICD-9*) code consistent with ACHD from 2010 to 2015. ACHD center providers were identified using National Provider Identification numbers. Usage rates and percentages were calculated with person-years in the denominator, and rate ratios and odds ratios (ORs) were estimated using negative binomial and logistic regression. Only 11.7% of identified individuals (N=10 199) were seen at the ACHD center. These individuals were younger (median 36 versus 47 years; $P<0.0001$) and had higher rates of Medicaid insurance (47.8% versus 28.4%; $P<0.0001$), heart failure (31.4% versus 15.3%; $P<0.0001$), and arrhythmia (75.5 versus 49.2%; $P<0.0001$). They had more visits of all types (outpatient: 79% per year versus 64% per year [age-adjusted OR, 2.54; 99% CI, 2.24–2.88]; emergency department: 29% versus 22% per year [adjusted OR, 1.34; 99% CI, 1.18–1.52]; inpatient: 17% versus 12.0% per year [adjusted OR, 1.92; 99% CI, 1.67–2.20]). Rates of guideline-indicated annual echocardiography were low (7.7% overall, 13.4% in patients at the ACHD center).

CONCLUSIONS: Patients at an ACHD center comprise a distinct and complex group with a high rate of healthcare use and a relatively higher compliance with guideline-indicated annual follow-up. These findings underscore the importance of building and supporting robust systems for ACHD care in the United States.

Key Words: congenital heart disease ■ epidemiology ■ health services ■ quality and outcomes

The adult congenital heart disease (ACHD) population in the United States currently numbers >1.4 million people and is predicted to continue to grow in the future.[1] Efforts to study this population have been hampered by multiple factors, including a high degree of phenotype heterogeneity and a lack of a large comprehensive database with which to study the population.[2] Administrative data sets, including statewide All Payer All Claims (APAC) databases, provide a way to measure the healthcare use of patients with ACHD across payers and healthcare systems. As such, they are an appealing method of examining trends in healthcare use and delivery in this group.

Although patients with ACHD are known to be high users of both outpatient and inpatient care,[3,4] the details and drivers of this usage are still not well understood. Most previous analyses have had a limited focus, for instance, on inpatient but not outpatient visits.[4–6] Importantly, a recent study has shown significant regional variations in the demographics, comorbidities, and healthcare use of patients with ACHD,[3] underscoring the importance of obtaining and integrating data from a broad array of sources.

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CLINICAL PERSPECTIVE

What Is New?

- A minority of Oregon patients with adult congenital heart disease (ACHD) are seen at an ACHD center.
- Patients at an ACHD center are younger and more likely to be on Medicaid than patients with ACHD not seen at an ACHD center.
- Patients at an ACHD center have more outpatient, emergency department, and inpatient visits than patients not at an ACHD center, even after adjustments for age and disease complexity.

What Are the Clinical Implications?

- These data suggest that most patients with ACHD do not receive guideline-directed ACHD care. Continued work is needed to improve referral pathways and access to care for all patients.

Nonstandard Abbreviations and Acronyms

ACHD	adult congenital heart disease
APAC	All Payer All Claims

ACHD guidelines recommend assessment by an ACHD specialist for the majority of patients with congenital heart disease (CHD), with yearly or biannual follow-up recommended for those with moderate-complex CHD.[7] Despite a known association between referral to an ACHD center and decreased mortality, many US patients with ACHD do not regularly see an ACHD specialist.[7,8] Contributors to poor access to ACHD care include lack of insurance or underinsurance, a shortage of ACHD physicians in the United States, inadequate knowledge about the importance of follow-up among patients and providers, and challenges related to geography and distance to care, among others.[9,10] Although it is likely that patients who seek care at an ACHD center differ in substantial ways from patients who are cared for in the community, the nature of these differences has yet to be defined.

In this analysis, we sought to define the characteristics of the ACHD population in Oregon with a specific focus on understanding differences in care use between patients who did and did not receive care at the state's only accredited ACHD center. We used the Oregon APAC database, a large administrative data set representing ≈94% of Oregon residents.

METHODS

ACHD Diagnosis

The data that support this study will be available from the corresponding author on reasonable request. To minimize the chances of sharing data that will be could be used to identify patients, location data (zip code, hospital location) will not be shared. Because of the nature of the study, informed consent was not performed. The Oregon APAC database from January 2010 to October 2015 was queried to identify patients with an *International Classification of Diseases, Ninth Revision* (*ICD-9*) code consistent with CHD (codes 745–747; Figure). Those aged <18 years and >65 years in 2010 were excluded from further analyses. In evaluating CHD, we considered diagnosis codes from any year in the data set and all claims. We applied a modified hierarchical algorithm to (1) exclude patients who only had evidence of diagnoses with low sensitivity/specificity and (2) classify remaining patients into 1 of 13 major defect subgroups based on the codes. Other congenital heart abnormalities (746.9, 745.9, 746.89) were excluded. This algorithm was previously validated in a university hospital population, but because of the concern for low specificity in the general population, those with only the *ICD-9* code 394.0 (mitral stenosis, N=698) were omitted from the analyses. Importantly, 48% of individuals identified with this code in the sample had were aged ≥50 years, suggesting that they were unlikely to have Fontan physiology as they would have been categorized by the published algorithm.[11,12] For some analyses, these subgroups were categorized as either mild or moderate to complex (as in Table 1).[13]

Patient-Level Characteristics

Age was calculated as the difference between 2010 and year of birth (or between the calendar year of claims for usage analyses). The patient's geographic area of residence was classified as Portland metropolitan, other metropolitan, or rural using the Census-based metropolitan statistical area, which is a static variable in the APAC data set based on the most recent reported patient address. Driving distance to the ACHD center was calculated using the centroid of a similarly static zip code and queried from Google Maps at mid-day during the working week.[14] Insurance type was classified using APAC-provided categories; patients could have multiple coverage types and were counted if at least 1 claim appeared in any year with any insurance coverage. We identified ACHD center specialists at Oregon Health & Science University (the state's only accredited ACHD center) by National Provider Identifier using the provider table obtained

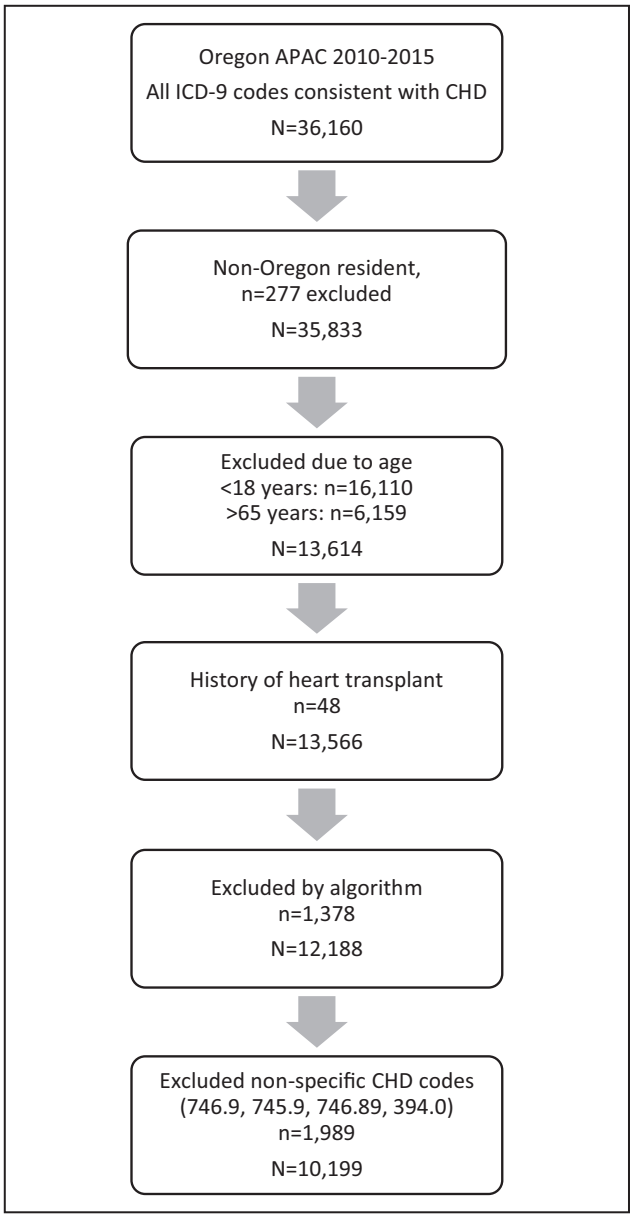

Figure. Derivation of the study cohort.
APAC indicates All Payer All Claims; CHD, congenital heart disease; and *ICD-9*, *International Classification of Diseases Ninth Revision*.

from the Oregon APAC. Providers can have multiple (>50) identification numbers assigned internally to the APAC database arising from slight variations in how the name is punctuated or similar differences. The data set included the internal identification for APAC, National Provider Identifier, first and last names, medical license number, and Medicaid identification. After first identifying exact National Provider Identifier matches, we merged those records back to the full list on names, allowing partial matches. Poor matches were flagged and all potential matches were reviewed for accuracy. Of 315 potential matches, 24 were rejected because they had mismatched names or insufficient evidence to decide. The identifiers were stripped, and the internal identification was used to identify claims originating from ACHD center providers. We identified 189 patients who were seen by a board-certified ACHD physician in community practice; these patients were not included in the ACHD center sample as the goal of the analyses was to examine patients at the accredited ACHD center versus non-ACHD centers, and the number was small (1.8% of the total sample). Out-of-state ACHD referral centers were identified by manual review of the hospital names included in the data set; because only 5 individuals seeking care exclusively at an out-of-state ACHD center were identified, the decision was made to exclude them from further analyses. Similarly, only 89 patients seeking care exclusively at out-of-state hospitals were identified, and they were also excluded. Comorbidities were determined using the SAS version of the Clinical Classifications Software for the *International Classification of Diseases, Ninth Revision, Clinical Modification* (*ICD-9-CM*) made available by Agency for Healthcare Research and Quality.[15] Guideline-indicated annual ACHD follow-up and annual echocardiography were defined as per the 2018 American Heart Association/American College of Cardiology Guideline for the Management of Adults With Congenital Heart Disease.[16] Diagnosis groups with guideline-indicated annual ACHD follow-up were Eisenmenger syndrome/cyanotic, single ventricle/Fontan, transposition of the great arteries, conotruncal abnormalities, and Ebstein's anomaly. Diagnosis groups with guideline-indicated annual echocardiography and/or electrocardiography were Eisenmenger syndrome/cyanotic, single ventricle/Fontan, transposition of the great arteries if aged ≥30 years, and Ebstein's anomaly.

Usage Variables

Visits were classified as outpatient, emergency department (ED), or inpatient using the Health Care Group codes provided by APAC.[17] Inpatient episodes were identified using Health Care Group codes, and if multiple overlapping or consecutive (next day) date ranges existed, these were counted as a single episode. Certain hospitalizations—for bone marrow or organ transplants, perinatal conditions, observation, or chemotherapy—were identified and excluded using Healthcare Effectiveness Data and Information Set definitions.[18] Cardiac admissions were identified using *ICD-9* codes that appeared either as the primary diagnosis or as the secondary diagnosis if the primary diagnosis was an ACHD code (ie, 745–747).

Outpatient and ED visits were considered only if they occurred outside of inpatient date ranges, with the exception of ED visits resulting in inpatient admissions, which were defined as ED claims with inpatient

Table 1. ACHD Diagnoses and Location of Care

ACHD Diagnosis	Total, N (%)	ACHD Center		Other Location of Care	
		N (%)	99% CI	N (%)	95% CI
Mild disease					
Bicuspid aortic valve	2532 (24.8)	222 (8.8)	7.4–10.3	2310 (91.2)	89.7–92.6
Pulmonic stenosis	210 (2.1)	33 (15.7)	9.9–23.2	177 (84.3)	76.8–90.1
Shunt lesion	3276 (32.1)	194 (5.9)	4.9–7.1	3082 (94.1)	92.9–95.1
Moderate-complex disease					
Eisenmenger/cyanotic	58 (0.6)	25 (43.1)	26.7–60.7	33 (56.9)	39.3–73.3
Single ventricle/Fontan	375 (3.7)	93 (24.8)	19.3–31.0	282 (75.2)	69.0–80.7
TGA	343 (3.4)	128 (37.3)	30.7–44.3	215 (62.7)	55.7–69.3
Conotruncal	1666 (16.3)	291 (17.5)	15.1–20.0	1375 (82.5)	80.0–84.9
Coarctation	855 (8.4)	128 (15.0)	12.0–18.4	727 (85.0)	81.6–88.0
AVSD	71 (0.7)	6 (8.5)	2.2–20.6	65 (91.5)	79.4–97.8
Ebstein's anomaly	85 (0.8)	14 (16.5)	7.6–29.2	71 (83.5)	70.8–92.4
TAPVR/PAPVR	67 (0.7)	15 (22.4)	10.9–37.9	52 (77.6)	62.1–89.1
Subaortic stenosis	107 (1.0)	9 (8.4)	3.0–17.7	97 (91.6)	82.3–97.0
Anomalous coronary	554 (5.4)	33 (6.0)	3.7–9.0	521 (94.0)	91.0–96.3

ACHD indicates adult congenital heart disease; AVSD, atrioventricular septal defect; PAPVR, partial anomalous venous return; TAPVR, total anomalous pulmonary venous return; and TGA, transposition of the great arteries.

claims on the same or next day. Similarly, outpatient visits were counted only if they did not occur on the same day an ED visit. Multiple claims from the same day were counted as a single episode.

Statistical Analysis

Descriptive statistics (counts and percentages) and Pearson chi-squared tests were used to compare groups defined by disease severity and specialty care with respect to patient characteristics. Usage rates were calculated with the number of events per person per calendar year in the numerator and the number of people with any claim of any type for the calendar year in the denominator, that is, events per person-year. Rate ratios (RRs) were estimated using negative binomial regression. Each person contributed 1 observation for each year they appeared in the data set. The outcome was coded as the number of events of a given type (0 or more) they experienced in that calendar year. Because no partial years were used, no offset was included in the model. Clustered variance estimates were used to account for the correlation between multiple observations from the same person. Adjusted estimates included age and disease severity (mild versus moderate to complex) in the model, and figures were divided by 100 to express rates as events per 100 patients per year for ease of interpretation.

The percentage of patients receiving at least 1 procedure in a given year was also calculated with each patient contributing 1 observation per calendar year in which they appeared in the claims data. They were counted as having a procedure if 1 or more claims were submitted for the procedure in that year. Annual testing was evaluated at the patient level, rather than year by year, and patients were counted as having annual testing if they were tested in every year they appeared in claims. Odds ratios (ORs) were calculated using logistic regression with clustered variance estimates for the repeated observations from the same individual, and adjusted estimates included age and disease severity in the model as noted previously.

Data management and descriptive statistics were completed using SAS software version 9.4 for Windows (SAS Institute, Cary, NC). Usage analyses were completed with Stata/IC software version 15 for Windows (StataCorp, College Station, TX).

This study was approved by the institutional review board at Oregon Health & Science University.

RESULTS

A total of 10 199 individuals with ACHD were identified, 53% of which were women. The majority of individuals had missing race (64%) and ethnicity (67%) data (Oregon APAC reports 59% and 72% missing, respectively, in all claims).[19] Of those with an indicated race, 85.8% of patients were White, 5.8% were Black, 2.2% were American Indian or Alaska Native, 2.3% were Asian, 0.4% were Native Hawaiian or Pacific Islander, and 3.5% of the patients were listed as other. Of the patients, 4181 (41.0%) had moderate-complex CHD, and 6018 (59.0%) had CHD of mild complexity. The

Table 2. Characteristics of the Study Population by Location of Care

	Overall	Patients at the ACHD Center	Patients Not at the ACHD Center	*P* Value
Total, n (%)	10 199 (100)	1191 (11.7)	9008 (88.3)	…
Female, n (%)	5336 (53.2)	635 (53.3)	4701 (52.2)	0.463
Age, y, n (%)				
18–24	1443 (14.1)	255 (21.4)	1188 (13.2)	<0.0001
25–34	1893 (18.6)	327 (27.5)	1566(17.4)	
35–44	1665 (16.3)	226 (19.0)	1439 (16.0)	
45–54	2256 (22.1)	215 (18.1)	2041 (22.7)	
55–65	2942 (28.8)	168 (14.1)	2774 (30.8)	
Comorbidities, n (%)				
Hypertension	5013 (49.2)	522 (43.8)	4491 (49.9)	<0.0001
Heart failure	1748 (17.1)	374 (31.4)	1374 (15.3)	<0.0001
Stroke	2142 (21.0)	228 (19.1)	1914 (21.2)	0.094
Coronary artery disease	2445 (24.0)	310 (26.0)	2135 (23.7)	0.077
Rhythm disorder	5329 (52.3)	899 (75.5)	4430 (49.2)	<0.0001
Diabetes mellitus	2907 (28.5)	305 (25.6)	2602 (28.9)	0.019
Insurance type, n (%)				
Medicaid	3126 (30.7)	569 (47.8)	2557 (28.4)	<0.0001
Medicare	939 (9.2)	48 (4.0)	891 (9.9)	<0.0001
Dual Medicaid/Medicare	989 (9.7)	170 (14.3)	819 (9.1)	<0.0001
Commercial	5255 (51.5)	480 (40.3)	4775 (53.0)	<0.0001
Self	2907 (28.5)	343 (28.8)	2634 (29.2)	0.753
Home geographic area, n (%)				
Portland metro	5203 (51.0)	653 (54.8)	4550 (50.5)	0.018
Other metro	2992 (29.3)	318 (26.7)	2674 (29.7)	
Rural	2004 (19.6)	220 (18.5)	1784 (19.8)	
Distance from ACHD center, n (%)				
<1 h	5343 (52.4)	695 (58.4)	4648 (51.6)	<0.0001
1–4 h	3827 (37.5)	445 (37.4)	3382 (37.5)	
>4 h	1029 (10.1)	51 (4.3)	978 (10.9)	

ACHD indicates adult congenital heart disease.

most common diagnosis categories represented in the sample were shunt lesions (N=3276, 32.1%), bicuspid aortic valve (N=2532, 24.6%), and conotruncal abnormalities (N=1666, 16.3%; Table 1). The most common cardiovascular comorbidities were rhythm disorders (N=5329, 52.3%) and hypertension (N=5013, 49.2%; Table 2). A total of 17.1% (N=1748) had a diagnosis of heart failure.

Location of Cardiac Care

Of the patients, 11.7% were seen at least once at the ACHD center during the study period (moderate-complex CHD, 17.7%; mild disease, 7.4%). Those seen at the ACHD center were younger with a right-skewed distribution of age with a median of 36 years versus a left-skewed distribution with a median of 47 years for patients not seen at the ACHD center (*P*<0.0001). Patients at the ACHD center were more likely to have moderate-complex CHD (62.3% versus 38.1%). The majority of patients across all diagnosis groups were not seen at the ACHD center. Patients with Eisenmenger syndrome had the highest rates of ACHD follow-up (43.1%; 99% CI, 26.7%–60.7%), and anomalous coronary artery patients had the lowest rates (6.0%; 99% CI, 3.7%–9.0%; Table 1). Of the patients seen in the ACHD center, 13.4% were seen annually during the study period, with a rate of 16.2% when the analysis was limited to those with a guideline indication for annual follow-up. In the overall cohort, 3.1% of individuals with a guideline indication for annual ACHD follow-up received such follow-up.

Patients at the ACHD center were more likely to have heart failure (31.4% versus 15.3%; *P*<0.0001) and arrhythmia (75.5 versus 49.2%; *P*<0.0001) but less likely to have hypertension (43.8% versus 49.9%; *P*<0.0001;

Table 3. Healthcare Use by ACHD Versus Non-ACHD Care

	Patients at the ACHD Center	Patients Not at the ACHD Center	Adjusted RR*
Type of visit			
Outpatient	633.20 (559.25–707.15.93)	372.05 (354.44–389.67)	2.01 (1.76–2.28)
ED	73.06 (60.79–85.34)	44.40 (41.46–47.34)	1.50 (1.25–1.80)
Inpatient	30.84 (26.32–35.37)	17.47 (16.43–18.51)	2.13 (1.81–2.50)
ED to hospital admission	2.30 (1.66–2.94)	1.23 (1.05–1.41)	2.21 (1.59–3.07)
Cardiac admission type			
All cardiac	14.55 (12.11–16.98)	7.32 (6.84–7.81)	2.59 (2.16–3.10)
Heart failure/cardiomyopathy	4.68 (3.17–6.19)	1.34 (1.10–1.59)	4.49 (3.01–6.69)
Arrhythmia	4.03 (2.93–5.14)	1.39 (1.18–1.59)	3.89 (2.83–5.34)
Valve disease	2.19 (1.55–2.84)	1.61 (1.43–1.80)	2.03 (1.47–2.81)
Coronary artery disease	1.68 (1.01–2.34)	1.47 (1.27–1.68)	1.47 (0.97–2.21)
Stroke	1.69 (1.05–2.33)	1.28 (1.11–1.45)	2.09 (1.38–3.15)
Congenital heart disease	3.00 (2.24–3.76)	0.98 (0.85–1.11)	2.85 (2.14–3.78)
Endocarditis	0.50 (0.18–0.81)	0.19 (0.12–0.26)	2.86 (1.35–6.07)
Pericarditis	0.33 (0.01–0.65)	0.18 (0.11–0.25)	2.06 (0.71–6.02)
Pulmonary hypertension	0.50 (0.20–0.79)	0.16 (0.09–0.24)	3.79 (1.76–8.17)
Cardiogenic shock	0.66 (0.34–0.99)	0.12 (0.07–0.17)	6.11 (3.05–12.21)
Cardiac arrest	0.20 (0.01–0.40)	0.06 (0.02–0.09)	4.18 (1.31–13.36)

Values are expressed as the number of events per 100 patients per year (99% CI). ED indicates emergency department; and RR, rate ratio.
*RR adjusted for age and disease complexity.

Table 2). There were no significant differences in the prevalence of stroke, coronary artery disease, and diabetes mellitus between the groups.

Patients at the ACHD center were more likely to have Medicaid (47.8% versus 28.4%; $P<0.0001$) and less likely to have Medicare (4.0% versus 9.9%) or commercial (39.4% versus 52.4%) insurance. Of the patients at the ACHD center, 58.4% lived within an hour of the center versus only 51.6% of those who did not access ACHD care (Table 2).

Patients at the ACHD center were more likely to have outpatient (633.2 versus 372.1 visits per 100 patients per year; RR, 2.01; 99% CI, 1.76–2.28), ED (73.1 versus 44.4 visits per 100 patients per year; RR, 1.50; 99% CI, 1.25–1.80), and inpatient visits (30.8 versus 17.5 admissions per 100 patients per year; RR, 2.13; 99% CI, 1.81–2.50), even after adjustments for age and CHD complexity (Table 3). As compared with patients not at the ACHD center, patients at the ACHD center were more likely to have a cardiac admission (14.6 versus 7.3 admissions per 100 patients per year; RR, 2.59; 99% CI, 2.16–3.10). The highest rate of admission was among patients at the ACHD center with moderate-complex CHD: 15.5% of patients per year (99% CI, 13.3–17.7) versus 11.1% (99% CI, 10.3–12.0) in those with moderate-complex CHD not seen in the ACHD center (Table 4). Heart failure/cardiomyopathy and endocarditis admissions were more common in patients at the ACHD center, among other diagnoses (Table 3).

Cardiac Procedures and Diagnostic Testing

Cardiac catheterization was more commonly performed in patients at the ACHD center than in those who were not patients at the ACHD center (7.1% versus 3.9% per year; OR, 2.41; 99% CI, 2.01–2.88; Table 4). Similarly, electrophysiology studies/ablations and pacemakers/defibrillator implantations were more commonly performed in patients at the ACHD center (electrophysiology study/ablation, 1.1% versus 0.7% [OR, 2.01; 99% CI, 1.34–3.00]; pacemaker/defibrillator, 0.8% versus 0.5% [OR, 2.24; 99% CI, 1.45–3.45]).

Patients at the ACHD center were more likely to have had at least 1 echocardiogram during the study period (95 versus 71%; OR, 8.78; 99% CI, 6.24–12.35). Only a small percentage of individuals had annual echocardiography, even at the ACHD center (11.7%; 99% CI, 9.3–14.1). When the analysis was limited to disorders for which an annual echocardiography is guideline indicated, only a minority of the overall sample had an annual echocardiography. The percentage was higher in those patients seen in the ACHD center (13.4%; 99% CI, 7.4–19.3) versus those not seen in the ACHD center (5.2%; 99% CI, 2.7–7.8), with an adjusted OR of 2.67 (99% CI, 1.28–5.58; Table 4). Of the patients at the ACHD center, 16.1% had guideline-indicated annual electrocardiograms versus 7.0% of other patients (adjusted OR, 2.70; 99% CI, 1.39–5.25).

Table 4. Healthcare Use by Disease Complexity and ACHD Care

	Overall			Moderate-Severe Complexity		
	Patients at the ACHD Center	Patients Not at the ACHD Center	Age-Adjusted OR	Patients at the ACHD Center	Patients Not at the ACHD Center	Age-Adjusted OR
Outpatient	79.26 (77.37–81.16)	63.78 (62.86–64.70)	2.54 (2.24–2.88)	79.05 (76.67–81.44)	62.39 (60.89–63.88)	2.65 (2.26–3.11)
Emergency	29.09 (26.72–31.47)	22.42 (21.67–23.18)	1.34 (1.18–1.52)	30.21 (27.71–33.26)	23.25 (21.96–24.55)	1.41 (1.19–1.66)
Inpatient						
All cause	16.96 (15.18–18.73)	11.95 (11.42–12.47)	1.92 (1.67–2.20)	15.50 (13.34–17.67)	11.15 (10.27–12.02)	1.95 (1.61–2.36)
Cardiac primary	9.262 (8.03–10.50)	5.79 (5.46–6.12)	2.16 (1.84–2.54)	8.86 (7.29–10.42)	5.12 (4.59–5.66)	2.40 (1.91–3.01)
Cardiac contributor	13.63 (12.04–15.21)	8.53 (8.09–8.96)	2.22 (1.91–2.58)	12.89 (10.89–14.88)	7.84 (7.12–8.57)	2.39 (1.94–2.93)
Cardiac procedures						
Catheterization	7.07 (6.01–8.13)	3.90 (3.64–4.16)	2.41 (2.01–2.88)	7.30 (6.00–8.59)	4.09 (3.66–4.52)	2.49 (2.00–3.12)
EP study/ablation	1.10 (0.70–1.51)	0.68 (0.56–0.79)	2.01 (1.34–3.00)	1.12 (0.59–1.64)	0.53 (0.37–0.69)	2.37 (1.36–4.12)
Pacemaker/implantable cardioverter defibrillator	0.81 (0.49–1.13)	0.44 (0.36–0.53)	1 (1.26–2.88)	1.03 (0.57–1.49)	0.41 (0.27–0.54)	3.08 (1.77–5.34)
Diagnostic testing						
Echocardiography	48.04 (46.07–50.01)	26.73 (26.05–27.42)	2.69 (2.46–2.95)	49.28 (46.77–51.79)	24.56 (23.42–25.69)	2.98 (2.65–3.36)
Electrocardiograpy	47.14 (44.92–49.36)	33.04 (32.25–33.82)	2.16 (1.95–2.39)	49.04 (46.30–51.79)	30.97 (29.65–32.28)	2.58 (2.26–2.94)
Annual testing						
Echocardiography	11.67 (9.27–14.07)	5.89 (5.26–6.53)	2.02 (1.55–2.64)	11.86 (8.80–14.92)	5.67 (4.65–6.69)	2.05 (1.44–2.92)
Electrocardiography	14.86 (12.21–17.52)	8.50 (7.75–9.26)	2.10 (1.65–2.66)	15.09 (11.71–18.48)	7.97 (6.78–9.16)	2.29 (1.67–3.15)
Guideline-indicated annual*						
Echocardiography	…	…	…	13.36 (7.41–19.31)	5.22 (2.65–7.79)	2.67 (1.28–5.58)
Electrocardiography	…	…	…	16.31 (9.70–22.56)	7.03 (4.08–9.98)	2.70 (1.39–5.25)

Values are expressed as the percent of patients per pear (99% CI) and age-adjusted OR. ACHD indicates adult congenital heart disease; EP, electrophysiology; and OR, odds ratio.
*Guideline-indicated annual testing group includes the following: Eisenmenger syndrome/cyanotic, single ventricle/Fontan, transposition of the great arteries if aged ≥30 years, and Ebstein's anomaly.

DISCUSSION

The major findings of this investigation are 3-fold. First, despite guidelines recommending regular follow-up of patients with ACHD at the ACHD center, only about a tenth of patients with ACHD in Oregon are managed at an accredited ACHD center. Second, there are important differences between the patients managed at the ACHD center and those at non-ACHD centers, including a higher burden of complex cardiac comorbidities, a higher use of Medicaid insurance, and higher use of healthcare services for those at the ACHD center. Third, only a small minority of patients with complex ACHD receive guideline-indicated annual imaging and follow-up, suggesting a need for improvement in systems of and access to care for those with ACHD.

To our knowledge, this is the first population-level comparison of US patients seen at an ACHD center versus not. By using an administrative data set that is nearly comprehensive of the state's population, we were able to obtain data on a large number of patients with ACHD accessing care across the health system. Unlike many previous analyses, which were limited to inpatient services,[4] patients with commercial insurance,[20] or select patient populations or procedures,[21,22] our analysis includes the most comprehensive population of ACHD in any state to date and encompasses the majority of care delivered in the state of Oregon between 2010 and 2015. As such, it provides a robust assessment of care usage in this complex population, with the caveat that our data include only individuals who are accessing health care.

In this study, patients at the ACHD center had nearly twice as many outpatient and inpatient visits as patients not at the ACHD center, even after adjustments for age and disease complexity. Importantly, a higher percentage of patients at the ACHD center had complex cardiac comorbidities such as heart failure and arrhythmia, and admissions for heart failure, cardiogenic shock, and cardiac arrest were more common in patients who were seen at the ACHD center. The higher complexity of the ACHD center cohort is likely a contributor to the higher rate of resource use among that population.

A notable strength of this study is the large proportion (≈94%) of the Oregon population that is included in the Oregon APAC database. The topography of Oregon presents a significant challenge for the delivery of subspecialty care, and high rates of income inequality and poverty and low levels of educational attainment are also important factors that contribute to suboptimal health outcomes in the state.[23] In contrast to the recent publication by Gurvitz et al,[3] which focused on patients receiving care near major population centers in the eastern half of the United States, our study includes data from a large western state with a significant population that is rural and/or remote to ACHD care. As such, it reflects an important and understudied segment of ACHD.

Importantly, patients at the ACHD center were more likely to live close to the ACHD center, suggesting that patients who live farther away encounter challenges in accessing care. This finding implies that further work is needed to address geographic disparities in ACHD care. This is of essential importance, as ≈45% of the US population lives >1 hour from an ACHD center, with an even higher proportion in rural states.[9] Further research is necessary to help identify strategies to mitigate geographic challenges and to ensure equal quality and access to care between rural and urban populations.

Patients receiving care at the ACHD center were significantly more likely to have Medicaid as opposed to commercial insurance, suggesting that there are important socioeconomic differences between these groups. Patients with ACHD, especially those with complex CHD, face educational and workplace-related challenges, with a lower rate of job participation than those without CHD.[24] A recent study demonstrated that adults who had CHD surgery in childhood have poorer neurodevelopment outcomes in adulthood, even among a cohort with mild complexity CHD.[25] These differences, among other factors, have important implications for work and socioeconomic outcomes. The fact that Medicaid patients, who may be more likely to face challenges in traveling for specialty care, comprise a larger proportion of the ACHD center population may reflect differential referral patterns for individuals with Medicaid as opposed to commercial insurance and/or a hesitation of community-based centers to care for patients with CHD on Medicaid. The higher burden of Medicaid patients also has important financial implications for ACHD centers, which shoulder a higher financial burden for the care of this population.

Only a minority of patients with ACHD accessed care at the ACHD center, a finding that is supported by prior studies demonstrating a high prevalence of gaps in care for those with ACHD.[10] In addition, in contrast to children with CHD, a high proportion of care of adults with CHD, including inpatient care, is known to occur at non-ACHD centers. This presents a significant challenge to the delivery of high-quality care for this specialized population and may contribute to adverse outcomes.[8] Importantly, the current supply of trained ACHD physicians is not adequate to meet the needs of the entire ACHD population.[26] Systems-based solutions are necessary, including, but not limited to, identifying which individuals are most in need of ongoing ACHD care and building partnerships with non-ACHD providers to improve their ability to deliver high-quality care for this population.

The overall compliance with annual follow-up was low in this cohort even for those managed at the ACHD center. This remained true after excluding those diagnoses for which current guidelines do not mandate annual follow-up and imaging. Patients at the ACHD center were significantly more likely to receive guideline-indicated annual imaging than patients not at the ACHD center. This finding suggests that ACHD-center providers may have relatively higher adherence to guideline-indicated care. However, the overall rates were low, even among patients at the ACHD center, suggesting that there is significant room for improvement in adherence to care.

Limitations

There are several important limitations of this analysis. First, there are inherent challenges in the use of administrative data sets, most notably the inability to adjudicate ACHD diagnoses by review of the medical record. Billing codes, especially those for mild forms of CHD, are inaccurate in some cases.[11,12,27] This inaccuracy is related to a combination of factors, including a lack of specificity of some codes, miscoding/misclassification of patients, and a lack of understanding of CHD by individuals entering the codes. For instance, our study revealed a smaller than expected number of individuals with atrioventricular septal defects, perhaps because of misclassifications as other shunt lesions. Importantly, *ICD-9* codes do not allow for the distinction between different types of CHD surgeries. For instance, we are unable to differentiate between patients with dextro-transposition of the great arteries who had an arterial switch procedure and those who had an atrial switch procedure. This has important implications for recommendations about follow-up, including frequency of imaging.

We took several steps to maximize accuracy in the selection of our study cohort. First, we excluded individuals age >65 years, as they have a lower probability of having actual CHD.[12] Second, we used a well-validated algorithm to categorize ACHD diagnoses and eliminated nonspecific billing codes where possible. Third, we performed many of our analyses in groups known to have a higher likelihood of having ACHD, including moderate-complex CHD and patients at the ACHD center.

A previous analysis from our group demonstrated that *ICD-9* codes more accurately identify ACHD when they originate from ACHD providers.[11,12] The ability to identify patients seen by ACHD providers is an important strength of our article, as it allowed us to identify a subpopulation with a high likelihood of actual CHD and a relatively low likelihood of inaccurate or erroneous coding. Importantly, however, the accuracy of ACHD billing codes in the general population is unknown, and therefore we cannot exclude the possibility that some of the differences in care usage seen in this study are related to a higher rate of miscoding of CHD in the non-ACHD center population.

This study was limited to individuals who had at least 1 healthcare claim during the study period. Individuals who did not access the healthcare system are not included in the data set, even if they were insured. Although >40% of adults with CHD experience a gap in cardiac care,[28] the vast majority retain some contact with the healthcare system.[10] Therefore, although this study cannot be considered comprehensive of the entire Oregon ACHD population, it likely represents a sizable percentage of individuals with ACHD in the state.

Although we were able to identify Oregon ACHD specialists, we were not able to reliably identify non-ACHD cardiology visits. Therefore, we were unable to examine differences in patients seen by ACHD and non-ACHD cardiologists. Importantly, this study cannot be interpreted as a comparison of different types of cardiology care as we were unable to determine what percentage of the non-ACHD follow-up group were seen by other types of cardiologists.

CONCLUSIONS

This study demonstrates that there are important differences in characteristics between adults with CHD cared for at an accredited ACHD center and those who do not access ACHD-specific care. Despite being younger in age, the ACHD center population has a higher rate of major cardiovascular comorbidities and of inpatient and outpatient healthcare use. The ACHD center population is also distinct with respect to payer mix, with nearly half of the patients at the ACHD center using Medicaid insurance. Importantly, patients at the ACHD center in Oregon are more likely to receive guideline-indicated follow-up and imaging, although overall rates are low.

Our findings underscore the importance of analyzing and improving systems of care for patients with ACHD in the United States. Although ACHD-specific care is guideline indicated, many US patients with ACHD do not access it. Further research is needed to identify strategies to recruit and retain these patients into the appropriate care location as well as to examine the association between ACHD-specific care and patient outcomes. Finally, our analysis suggests that the ACHD center population is both more medically complex and relatively less financially lucrative for hospital systems. This needs to be considered when developing policies to expand and maintain the ACHD workforce over time.

ARTICLE INFORMATION

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