Impact of facilities accredited by both adult and pediatric cardiology societies on the outcome of patients with adult congenital heart disease

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Abstract

Background: The emerging burden and need of hospital admission due to adult congenital heart disease (ACHD) will need many facilities with expertise in ACHD. Regional specialized ACHD centers are carrying this increasing patient burden. Although these centers are considered to perform better management than other institutes, their impact on outcome had not been fully evaluated.

Methods: We used the Japanese Registry of All cardiac and vascular Diseases (JROAD) and the JROAD Diagnosis Procedure Combination (DPC)/Per Diem Payment System dataset and certification data. We only analyzed adult (≥15 years old) patients with ACHD, defined by the *International Classification of Diseases, Tenth Revision,* diagnosis codes, between April 1, 2013, and March 31, 2014. We defined a "minimal essential regional ACHD (MER-ACHD) center" as an education institute accredited by adult and pediatric cardiology societies. The primary outcome is 30-day mortality. We investigated the impact of MER-ACHD centers on 30-day mortality by using generalized estimating equations.

Results: Of the 538 hospitals registered at JROAD that agreed to participate in the DPC discharge database study, 65 (12.1%) were MER-ACHD centers. Of 4,818 patients (46.8% male; age, 50.1 ± 21.4 years), 45.5% were admitted to a MER-ACHD center. Nearly half (48.1%) of the admissions were cases of atrial septal defect, followed by ventricular septal defect, tetralogy of Fallot, and congenital insufficiency of the aortic valve or bicuspid aortic valve. Multivariate analysis revealed a negative impact of emergency admission (1.051 [1.042–1.061]) and a positive impact of MER-ACHD centers (0.986 [0.973–0.999]) on 30-day mortality after adjustment of disease severity.

Conclusion: We noted the impact of MER-ACHD centers on 30-day mortality. Further investigation is needed to establish appropriate regional ACHD center criteria to deliver appropriate ACHD management.

Key words: mortality, ACHD, congenital, regional AHCD center, minimal essential

Highlights

- ✓ The patients with ACHD treated in a MER-ACHD center were better than others.
- ✓ Half of ACHD admissions were simple cases such as atrial septal defect.
- ✓ About half of the patients with ACHD were not treated in tertiary ACHD or MER-ACHD center.

Abbreviations

congenital heart disease	CHD
adult patients with CHD	ACHD
Japanese Registry of All Cardiac and Vascular Diseases	JROAD
Diagnosis Procedure Combination	DPC
Per Diem Payment System	PDPS
“Minimal essential regional ACHD center	MER-ACHD center
International Classification of Diseases	ICD

1. Background

Recent progress in initial treatment has improved the survival of children with congenital heart disease (CHD) into adulthood in up to almost 90% of cases.[1] Thus, attention now needs to be paid to how late complications are to be managed, who should care for the patients, and where care for these adult patients with CHD (ACHD) should be provided.[2] In many countries, specialist and specialized center certification systems have been developed in many countries.[2, 3] ACHD specialists should be able to manage moderate or complex cases, and specialized ACHD centers should afford prompt access for patients and referring physicians to provide diagnosis and management.[2] Although the impact of specialized care on the outcomes has been reported as a political success, many patients with ACHD are still cared for by non-ACHD specialists in non-specialized centers.[4] In fact, some reports have revealed that tertiary ACHD referral centers could cover only half of all admissions.[5] The emerging burden and need of hospital admission for ACHD might exceed the capacity of previously defined "tertiary ACHD referral centers."[6] Although regional ACHD centers were considered to cover these overflowing number of patients, according to the ACC/AHA guideline, the impact of these facilities on outcome has not been fully evaluated.[2]

The essential component of ACHD care is collaboration between adult and pediatric cardiologists.[6] Therefore, the minimum essential requirement for a hospital to convey better care is certification from both the adult and pediatric cardiology societies. In this study, we evaluated the impact of potential minimal essential regional ACHD (MER-ACHD) centers on outcome in Japan by using the national administrative dataset.

2. Methods

We used the administrative data and questionnaire by Ochiai et al. in 2013 to determine appropriate tertiary ACHD facilities.[7] By using this dataset, we retrospectively analyzed in detail the chief reason for admission among patients with ACHD defined by the *ICD-10* code and compared the outcomes of the patients with ACHD between hospitals with or those without the minimum requirement to be considered an adult congenital heart disease center, as defined below. We defined the primary outcome as 30-day mortality.

2.1. Data collection

The details of the Japanese Registry of All Cardiac and Vascular Diseases (JROAD) and the JROAD Diagnosis Procedure Combination/Per Diem Payment System (DPC/PDPS) project had already been described before.[8] The JROAD-DPC database covers nearly all teaching hospitals with cardiovascular beds, except for stroke participation in JROAD, to meet the Japanese Circulation Society (JCS) requirement that JCS-certified teaching hospitals should provide cardiology training for physicians who wish to be JCS board-certified cardiologists and to undertake the JCS board test.[9] This cross-sectional survey used the DPC discharge database from institutions that participated in the JROAD study. Of the 1,535 hospitals that responded to JROAD, 637 agreed to participate in the DPC discharge database study. Hospitalization records between April 1, 2013, and March 31, 2014, were collected. Operation was defined by using the surgical procedure coding K-code, which is Japan's local surgical coding to picking up surgery.[10] For example, the Norwood procedure is coded as K-587. Thirty-day mortality could be available via DPC dataset.

2.2. Inclusion criteria and definitions

We only analyzed adult (≥15 years old) patients with ACHD, defined as those whose diagnosis for admission and comorbidities at admission include *International Classification of Diseases, Tenth Revision* (*ICD-10*), diagnosis codes related to adult congenital heart disease (Congenital malformations of the circulatory system Q20–Q28). In this study, we defined a "minimal essential regional ACHD (MER-ACHD) center" as follows: (1) an accredited center for education by the Japanese Circulation Society with appropriately submitted DPC data to the JCS in 2013 (including both class A and B teaching hospitals, class A JCS-certified teaching hospitals needing >2 JCS board-certified cardiologists and 30 cardiovascular beds, and class B needing >1 JCS board-certified cardiologist and 15 cardiovascular beds) [8] and (2) a center also accredited by the Japanese Society of Pediatric Cardiology and Cardiac Surgery in 2013, including >1 board-certified pediatric cardiologist and >100 admissions, and >50 catheterizations per year.[11] Unfortunately, the DPC database did not include any Japanese Association of Children's Hospitals and Related Institutions. Therefore, we combined the questionnaire data by Ochiai et al. in the same year to confirm the centers accredited by the Japanese Society of Pediatric Cardiology and Cardiac Surgery.[11] Disease severity was classified according to the previous definitions (Supplement table).[12] We excluded patients whose main diagnosis was not classified according to the severity classification. Other variables such as Charlson comorbidity index could be used as reported in previous JROAD study.[8] The study protocol was approved by the institutional review board of St. Luke's International University (16-R080).

2.3. Statistical analysis

Categorical variables were presented as numbers and proportions, and continuous variables were presented as mean ± standard deviation (SD) or median (interquartile range). We tested the hypothesis of a significant association between MER-ACHD centers and patient outcomes using generalized estimating equations (GEE) to account for the clustering of patients within each hospital. Disease severity, Charlson score index, age, sex, chief reason of admission (ACHD or not), operation, and emergent admission were adjusted from the clinical perspective. All hypothesis tests had a two-sided significance level of 0.05. All statistical analyses were performed with RStudio version 1.0.143 (RStudio, Boston, MA).

3. Results

A MER-ACHD center was detected in 65 (12.1%) of the total 538 hospitals. Of the total 4,818 patients (46.8% male, 50.1 ± 21.4 years old), about half (2191 patients, 45.5%) were admitted to a MER-ACHD center. Simple complexity cases such as atrial and ventricular septal defects were found in more than half of the patients (Table 1, Supplement table). Simple complexity cases were frequently admitted in the non MER-ACHD center (79.2% vs 61.6%). Moderately and great complexity cases were significantly frequently admitted to a MER-ACHD center (12.1% vs 17.0%, and 8.7% vs 21.4%, respectively). On the other hand, older patients and many emergency admitted patients were statistically more frequently observed in the non-MER-ACHD centers. The comorbidity index was higher, and associated comorbidities such as myocardial infarction, congestive heart failure, peripheral vascular disease, cerebrovascular disease, dementia, diabetes, and cancer were frequently significantly observed in the non-MER-ACHD centers (Table 1).

Table 2 lists the main diagnoses in >100 patients, which consists 89.3% of total admissions. Nearly half (48.1%) of the admitted patients were diagnosed as having atrial septal defect, followed by ventricular septal defect, tetralogy of Fallot, and congenital insufficiency of the aortic valve or bicuspid aortic valve (12.3%, 8.8%, and 3.3%, respectively; Figure 1).

Multivariate analysis revealed the impact of MER-ACHD centers on patient outcomes after adjustment for disease severity, Charlson score index, and other available patient characteristics (Table 3).

4. Discussion

This is the first study to evaluate the impact of MER-ACHD centers (facilities certified by the adult and pediatric cardiac societies) on 30-day mortality in patients with ACHD. After adjustment for disease severity, the MER-ACHD centers showed superior outcomes than the other centers. Our data might encourage further implementation of more regional facilities to support ACHD care.

4.1. Specialized ACHD referral center and tertiary hospitals

In 1998, the Canadian Cardiac Society (CCS) recommended that all patients with ACHD be referred to specialized ACHD referral centers, which required that the following criteria be fulfilled: (1) the expertise criterion, mandating that each center uses at least 1 imaging and/or interventional cardiologist and at least 2 CHD surgeons with advanced training and experience in ACHD; (2) the patient volume criterion, requiring that at least 520 patients per year be seen at the center and that the center is listed as a designated center in the 1998 consensus document.[13] Unfortunately, only one-third of patients could receive specialized ACHD care even after these recommendations in Canada. Furthermore, with regard to hospitalizations, half of ACHD patients were admitted at non-specialized ACHD centers, which was compatible with our study.[5] Therefore, we should admit the fact that patients with ACHD are currently not able to access specialized ACHD centers. According to the calculated estimation of the increasing number of patients with ACHD, which is expected to reach 500,000 in Japan, we cannot imagine that these patients could be appropriately cared for only by tertiary hospitals. Therefore, it is time to consider how to implement appropriate care for patients with ACHD from the perspective view of facility certification. Of course, these tertiary specialized ACHD centers will have a chief role in education. The education on ACHD care should be imparted appropriately and quickly.

4.2. Minimal essential regional ACHD centers

Regardless of the names and definition of MER-ACHD centers, as mentioned previously, we need regional hospitals to care for patients with ACHD appropriately. Regional ACHD centers were advocated at the 32nd Bethesda Conference, especially mentioned in Task Force 4, an organization of delivery systems for adults with congenital heart disease.[2] Regional ACHD centers were considered to afford prompt access to provide comprehensive diagnosis, management planning, and patient counseling. These facilities need several services such as cardiac anesthesia, operating rooms, cardiac surgery, intensive care, in-patient service, transplantation, catheterization laboratory, noninvasive imaging service, electrophysiology service, high-risk obstetrics, and cardiac pathology.

Unfortunately, in addition to the fact evidence is lacking to support these facilities to improve outcome, some criteria might not fit in other countries such as Japan, where the number of transplantations have been quite fewer.[14] Previous surveys also revealed that the essential component of ACHD care is collaboration between adult and pediatric cardiologists.[6] In Japan, ACHD specialist certification is yet to be finalized until 2020. Therefore, the presence of both pediatric and adult cardiologists might be the sole indicator to convey appropriate adult congenital heart disease management in this study period. Therefore, in this study, we define a MER-ACHD center as a facility certified by the national and pediatric cardiology societies.

Improved outcomes in accredited hospitals by several societies have already been reported in other cardiology areas, which were comparable with those attained in this study.[15] This improvement could be explained via several mechanisms. First, cooperation with adult and pediatric cardiologists itself could improve outcome in this area.[16]

Second, hospital volume/case volume effect could be considered. Approximately 12.1% of all hospitals cover 45.5% of patients, which depicts the fact that the MER-ACHD centers cover more patients/institutes than the non-MER-ACHD centers. The number of patient admissions clearly differ between the two types of facilities. The MER-ACHD centers cover an average of 33.7 patients (2191/65) per year, in contrast to the 5.5 patients (2191/65) per year covered by the non MER-ACHD centers. This type of volume effect, that is, “the larger number of patients the hospital manages, the better the outcome,” has been already validated in many areas.[15, 16] Our data also revealed that emergent admissions considered as high-risk on the basis of the multivariate analysis results were frequently observed in the non-MER-ACHD centers. A multi-disciplinary team is required to manage patients with ACHD who especially require emergency care.[17, 18] Limited data revealed that more than half of emergencies in patients with congenital heart disease required cooperation with another specialized department, which might help us understand our results.[16][19] In addition, as shown in Table 1, the Charlson comorbidity index is not different, but comorbidities are frequently observed in non-MER-ACHD centers, which also showed the high-risk tendency in non-MER-ACHD centers. It is reasonable in emergent situations that patients cannot easily choose the hospital; therefore, selectively higher-risk patients tend to visit non-MER-ACHD centers. However, after adjustment of these conditions, we found the impact of MER-ACHD centers on outcome, which is reasonably caused by the case-volume effect. In summary, as compared with MER-ACHD centers, non-MER-ACHD centers face a relatively small number of high-risk patients, which might result in a non-favorable outcome. Further investigation might be needed to validate this type of volume effect.

Our definition of a MER-ACHD center was quite minimal; thus, previously defined regional ACHD centers in previous literatures might deliver better outcomes. Only a small number of hospitals (<10) were accredited as specialized ACHD referral centers. In Japan, we could only select 9 hospitals, and in Quebec, 3.[7, 20] As mentioned earlier, the number of specialized ACHD referral centers is too small to manage the increasing number of ACHD patients. In Japan, patients can access any hospital they want; therefore, we should improve outcomes all over the country. As the British Cardiac Society Working Party suggested, appropriate ACHD care should be spread more promptly.[21]

5. Limitations

Although we performed a serial analysis, our study has several limitations. First, because of the nature of the administrative data, we could not obtain the patients' characteristics such as ejection fraction and laboratory data; only the diagnosis was readily available. Therefore, we might not have been able to appropriately adjust potential confounding factors to evaluate outcomes. Furthermore, in this study, non-MER-ACHD centers cover higher-risk patients such as those requiring emergency admissions. Of course, we have already adjusted such variables, but we cannot adjust unmeasured potential confounding factors. We need further validation using other datasets such as registries. Second, the detailed collaboration between pediatric and adult cardiologists in the MER-ACHD centers was unclear. We simply used certification data from each cardiology society. Therefore, adult and pediatric cardiologists might not actually collaborate in some institutions. Unfortunately, no data exists that support the detailed communication between them.

Most reports related to the hospital impact on outcomes did not consider the detailed communication as well.[22] Finally, we did not include pediatric hospitals. We have to admit the fact that many pediatric hospitals might cover many complex ACHD cases that could not be successfully transferred to adult institutes yet. This type of selection bias could not be denied. Our results could be applicable but not for future potential highly complex cases treated in pediatric hospitals.

6. Conclusion

We revealed the positive impact of MER-ACHD centers on 30-day mortality. Further investigation is needed to determine the appropriate regional ACHD centers to deliver proper ACHD management.

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Figure Legend

Figure 1. Diagnoses in ≥100 admissions.

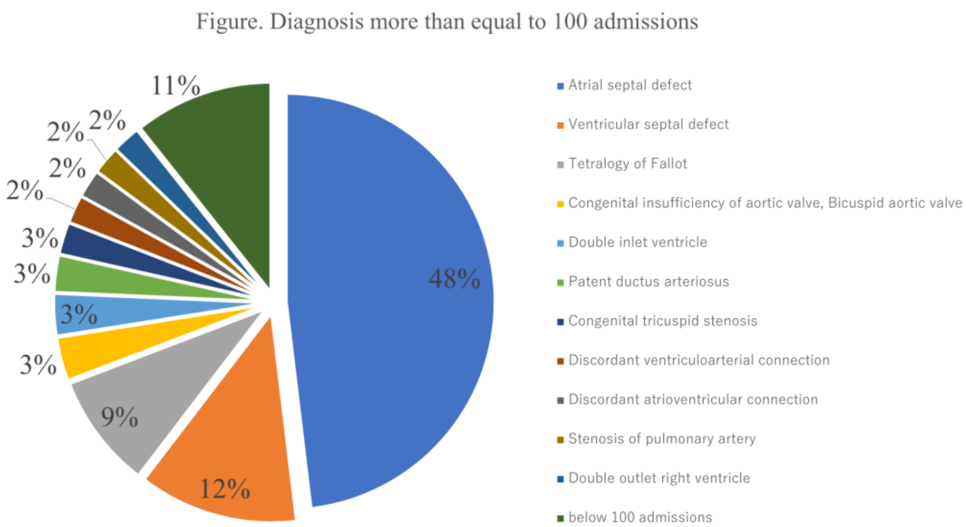

Table.1 Baseline characteristics

	non-MER ACHD centres (n=473)	MER ACHD centres (n=65)	*p* -value
Patient number	2627	2191	
Patient characteristics			
Age, y	55.9±20.3	43.1±20.6	<0.001
Male ,n (%)	1211 (46.1)	1043 (47.6)	0.300
Emergent admission ,n (%)	796 (30.3)	417 (19.0)	<0.001
Ambulance usage ,n (%)	258 (9.8)	103 (4.7)	<0.001
Pregnancy ,n (%)	12 (0.7)	39 (1.8)	<0.001
height, cm	158.4±9.9	159.9±9.8	<0.001
weight, kg	56±13	55.2±12.6	0.029
BMI	22.2±4.1	21.5±4	<0.001
Cost, median(IQR) yen	709424(253047-2000000)	886052(314234-1900000)	<0.001
Operation ,n (%)	1085 (41.3)	1053 (48.1)	<0.001
Severity (complexity)			
Simple complexity, n(%)	2081(79.2)	1349(61.6)	<0.001
Moderate complexity ,n (%)	317(12.1)	373(17.0)	
Great complexity ,n (%)	229(8.7)	469(21.4)	
Outcome			
LOS, median(IQR) days	9(3-18)	8(4-16)	0.280
Total_death, n(%)	77 (2.9)	34 (1.6)	0.001
Death within 30days, n(%)	62 (2.4)	22 (1.0)	<0.001
Comorbidities			
Myocardial infarction ,n (%)	59 (2.2)	23 (1.0)	0.001
Congestive heart failure ,n (%)	1297 (49.4)	998 (45.5)	0.008
Peripheral vascular disease ,n (%)	127 (4.8)	43 (2.0)	<0.001
Cerebrovascular disease ,n (%)	149 (5.7)	92 (4.2)	0.020
Dementia ,n (%)	16 (0.6)	1 (<0.1)	0.001
Chronic pulmonary disease ,n (%)	73 (2.8)	58 (2.6)	0.780
Rheumatic disease ,n (%)	21 (0.8)	16 (0.7)	0.780
Peptic ulcer disease ,n (%)	70 (2.7)	75 (3.4)	0.120
Mild liver disease ,n (%)	70 (2.7)	70 (3.2)	0.280
Diabetes without chronic complication ,n (%)	238 (9.1)	105 (4.8)	<0.001
Diabetes with chronic complication ,n (%)	53 (2.0)	22 (1.0)	0.005
Hemiplegia or paraplegia ,n (%)	2 (0.1)	1 (<0.1)	0.670
Renal disease ,n (%)	64 (2.4)	45 (2.1)	0.370
Cancer ,n (%)	92 (3.5)	26 (1.2)	<0.001
Moderate or severe liver disease ,n (%)	5 (0.2)	1 (<0.1)	0.160
Metastatic cancer ,n (%)	5 (0.2)	2 (0.1)	0.370
AIDS/HIV ,n (%)	0 (0.0)	1 (<0.1)	0.270
Charlson Comorbidity Index, median(IQR)	1(0-1)	1(0-1)	<0.001

MER-ACHD, minimal essential regional adult congenital heart disease; BMI, body mass index; LOS, length of stay; AIDS, Acquired immune deficiency syndrome; HIV, Human Immunodeficiency Virus; IQR, interquartile range

Table.2 Diagnosis more than equal to 100 admissions

ICD-10 code	Diagnosis	The number of admissions, n(%)
Q211	Atrial septal defect	2318(48.1)
Q210	Ventricular septal defect	591(12.3)
Q213	Tetralogy of Fallot	422(8.8)
Q231	Congenital insufficiency of aortic valve, Bicuspid aortic valve	157(3.3)
Q204	Double inlet ventricle	153(3.2)
Q250	Patent ductus arteriosus	136(2.8)
Q224	Congenital tricuspid stenosis	115(2.4)
Q203	Discordant ventriculoarterial connection	103(2.1)
Q205	Discordant atrioventricular connection	102(2.1)
Q256	Stenosis of pulmonary artery	100(2.1)
Q201	Double outlet right ventricle	100(2.1)

Table.3 Multivariate analysis to predict 30-day mortatlity

Variables	Estimate (95%CI)
(Intercept)	0.993(0.968-1.018)
MER-ACHD	0.986(0.973-0.999)
Disease severity	1.003(0.998-1.009)
Charlson Comorbidity Index	1.002(0.998-1.006)
Age	1.000(1.000-1.001)
Male	0.998(0.991-1.005)
ACHD admission	1.000(0.992-1.008)
Operation	1.002(0.995-1.01)
Emergent admission	1.051(1.042-1.061)

MER-ACHD, minimal essential regional ACHD center; ACHD, adult congenital heart disease; CI, confidence interval

Supplement table.1 The definition of disease complexities

- Types of Adult Patients With Congenital Heart Disease of Great Complexity
  - Conduits, valved or nonvalved
  - Cyanotic congenital heart (all forms)
  - Double-outlet ventricle
  - Eisenmenger syndrome
  - Fontan procedure
  - Mitral atresia
  - Single ventricle (also called double inlet or outlet, common or primitive)
  - Pulmonary atresia (all forms)
  - Pulmonary vascular obstructive diseases
  - Transposition of the great arteries
  - Tricuspid atresia
  - Truncus arteriosus/hemitruncus
  - Other abnormalities of atrioventricular or ventriculoarterial connection not included above (i.e., crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)
- Types of Adult Patients With Congenital Heart Disease of Moderate Complexity
  - Aorto-left ventricular fistulae
  - Anomalous pulmonary venous drainage, partial or total
  - Atrioventricular canal defects (partial or complete)
  - Coarctation of the aorta
  - Ebstein’s anomaly
  - Infundibular right ventricular outflow obstruction of significance
  - Ostium primum atrial septal defect
  - Patent ductus arteriosus (not closed)
  - Pulmonary valve regurgitation (moderate to severe)
  - Pulmonic valve stenosis (moderate to severe)
  - Sinus of Valsalva fistula/aneurysm
  - Sinus venosus atrial septal defect
  - Subvalvar or supravalvar aortic stenosis (except HOCM)
  - Tetralogy of Fallot
  - Ventricular septal defect with
  - Absent valve or valves
  - Aortic regurgitation
  - Coarctation of the aorta
  - Mitral disease
  - Right ventricular outflow tract obstruction
  - Straddling tricuspid/mitral valve
  - Subaortic stenosis
- Types of Adult Patients With Simple Congenital Heart Disease
  - Native disease
    - Isolated congenital aortic valve disease
    - Isolated congenital mitral valve disease (e.g., except parachute valve, cleft leaflet)
    - Isolated patent foramen ovale or small atrial septal defect
    - Isolated small ventricular septal defect (no associated lesions)
    - Mild pulmonic stenosis
  - Repaired conditions
    - Previously ligated or occluded ductus arteriosus
    - Repaired secundum or sinus venosus atrial septal defect without residua

Repaired ventricular septal defect without residua