Advance Publication

Circulation Journal
doi:10.1253/circj.CJ-23-0537

ORIGINAL ARTICLE

In-Hospital Death and End-of-Life Status Among Patients With Adult Congenital Heart Disease

— A Retrospective Study Using the JROAD-DPC Database in Japan —

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Background: The end-of-life (EOL) status, including age at death and treatment details, of patients with adult congenital heart disease (ACHD) remains unclear. This study investigated the EOL status of patients with ACHD using a nationwide Japanese database.

Methods and Results: Data on the last hospitalization of 26,438 patients with ACHD aged ≥15 years, admitted between 2013 and 2017, were included. Disease complexity (simple, moderate, or great) was classified using International Classification of Diseases, 10th Revision codes. Of the 853 deaths, 831 patients with classifiable disease complexity were evaluated for EOL status. The median age at death of patients in the simple, moderate, and great disease complexity groups was 77.0, 66.5, and 39.0 years , respectively. The treatments administered before death to patients in the simple, moderate, and great complexity groups included cardiopulmonary resuscitation (30.1%, 35.7%, and 41.9%, respectively), percutaneous cardiopulmonary support (7.2%, 16.5%, and 16.3%, respectively), and mechanical ventilation (58.7%, 72.2%, and 75.6%, respectively). Overall, 70% of patients died outside of specialized facilities, with >25% dying after ≥31 days of hospitalization.

Conclusions: Nationwide data showed that patients with ACHD with greater disease complexity died at a younger age and underwent more invasive treatments before death, with many dying after ≥1 month of hospitalization. Discussing EOL options with patients at the appropriate time is important, particularly for patients with greater disease complexity.

Key Words: Adult congenital heart disease; End-of-life status; Nationwide database

Survival among patients with congenital heart disease (CHD) has increased due to advances in cardiovascular interventional techniques and therapy, with many patients reaching adulthood.[1] Reports indicate that in 2023 an estimated 500,000 individuals have adult CHD (ACHD) in Japan.[2] As the number of patients with ACHD has increased, the focus has primarily shifted to improving treatment outcomes, life expectancy, and quality of life.

With improvements in the care of patients with ACHD, their life expectancy has also improved. Changes in patterns of death have also been observed,[3] with higher rates of sudden death,[4] mortality from chronic heart failure exacerbations,[5,6] and admission to the intensive care unit (ICU) or emergency room 30 days before death than in patients with cancer.[7] These findings support an approach that focuses on the clinical status of patients in the last phase of life and on the circumstances surrounding their death, and are important data for developing a framework for medical and end-of-life (EOL) care for patients with ACHD. However, most studies to date have been single-center studies with small sample sizes[8,9] and limited generalizability to the growing population of patients with

Received July 23, 2023; revised manuscript received October 29, 2023; accepted November 12, 2023; J-STAGE Advance Publication released online December 9, 2023 Time for primary review: 20 days

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ISSN-1346-9843

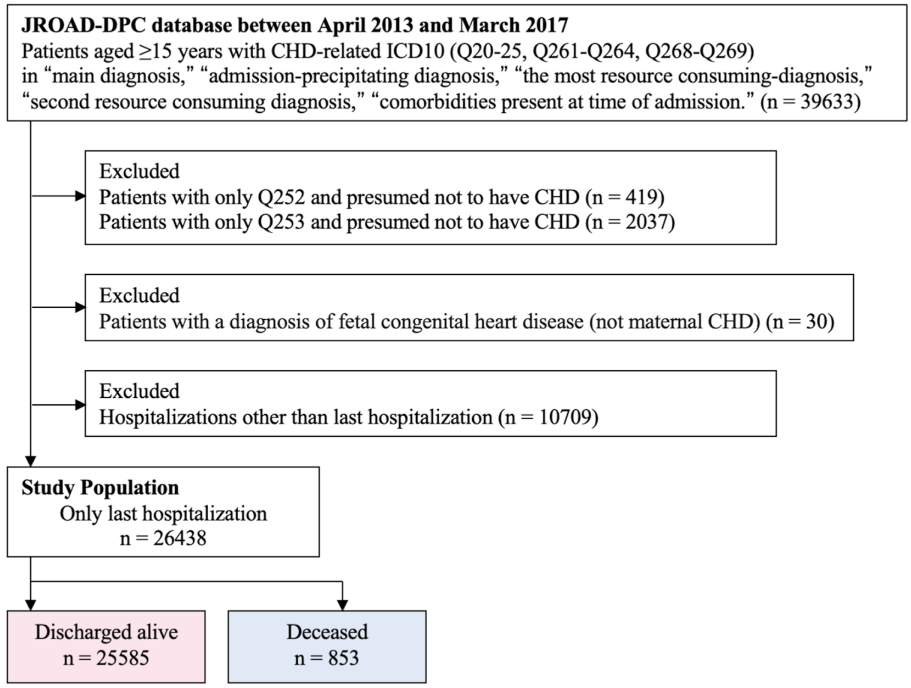

Figure 1. Study flowchart. CHD, congenital heart disease; ICD-10, International Classification of Diseases, 10th Revision; JROAD-DPC, Japanese Registry Of All cardiac and vascular Diseases-Diagnosis Procedure Combination.

ACHD. Furthermore, the differences in disease complexity among patients with ACHD have not been thoroughly examined.

The aim of this study was to clarify the EOL status of patients with ACHD, including age at death according to disease complexity, the proportion of invasive treatments (e.g., percutaneous cardiopulmonary support [PCPS] and intra-aortic balloon pumping [IABP]), and the number of ICU deaths, using a nationwide database in Japan. This investigation is expected to help patients and healthcare providers consider more realistic goals when planning EOL care.

Methods

Study Design

This retrospective cohort study was based on a nationwide database of acute care institutions in Japan.

Data Source

The Japanese Registry Of All cardiac and vascular Diseases (JROAD) was launched in 2004 by the Japanese Circulation Society to assess the status of cardiovascular disease clinical activity in Japan and improve the quality of cardiovascular disease care. In 2014, in collaboration with the National Cerebral and Cardiovascular Center, the JROAD Diagnosis Procedure Combination (DPC) database was launched with a medical payment system. Detailed descriptions of JROAD and JROAD-DPC are provided elsewhere.[10] The DPC system calculates the cost of inpatient care for acute care hospitals in Japan according to the International Classification of Diseases, 10th Revision (ICD-10) diagnoses and medical treatment. Therefore, the JROAD-DPC provides information on medical care provided by acute care hospitals specializing in cardiology. The validity of the primary diagnosis, procedure records,[11] and surgical records[12] in the DPC database is generally high.

Guidelines[13] recommend that, upon reaching adulthood, patients with CHD should continue to be followed up in specialized facilities (e.g., hospitals certified by the Japanese Society of Pediatric Cardiology and Cardiac Surgery [JSPCCS] or the Japanese Society for Adult Congenital Heart Disease). Because these facilities are acute care hospitals, data obtained from the JROAD-DPC are expected to be representative of the actual status of patients with ACHD.

Study Population

Data on adult (age ≥15 years) patients with CHD hospitalized between April 1, 2013 and March 31, 2018 at all participating hospitals were obtained from the JROAD-DPC database. The study flowchart is shown in **Figure 1**.

The identification of patients hospitalized for CHD was based on ICD-10 diagnosis codes related to CHD (Q20–25, Q261–Q264, Q268–Q269) as the "main diagnosis, admission-precipitating diagnosis, the most resource-consuming diagnosis", "second resource-consuming diagnosis", and "comorbidities present at the time of admission".

Patients with acquired heart disease and pregnant women with fetal CHD were excluded. Acquired heart disease was identified by ICD-10 codes Q252 and Q253, and the absence of another CHD code. In addition, 428 patients with the ICD-10 code Q252 (atresia of the aorta) were identified. Of these, 9 patients had CHD-related ICD-

10 codes, and 419 patients did not have a CHD-related diagnosis. Furthermore, there were 2,102 patients with the ICD-10 code Q253 (stenosis of the aorta or supravalvular aortic stenosis). Of these patients, 65 had CHD-related ICD-10 codes and 2,037 had no CHD-related ICD-10 codes. Compared with patients with CHD-related ICD-10 codes, patients without a CHD-related diagnosis were older, and it was assumed that these patients were misclassified as patients with CHD despite having acquired the disease. Therefore, we excluded patients with only ICD-10 codes Q252 and Q253 and no other CHD-related ICD-10 codes. Thirty pregnant women with a manually entered diagnosis such as "fetal heart disease" and no other CHD code were considered to have fetal rather than maternal CHD and were also excluded. To compare the characteristics between the living and deceased groups, admissions other than the last admission for each patient during the study period were also excluded.

Patient background data were obtained, including age, sex, and the length of hospital and ICU stays. Variable definitions are provided below.

Disease Complexity The disease complexity of CHD was classified as "simple", "moderate", or "great" based on the ICD-10 codes and in accordance with the classifications described in the 2008 American College of Cardiology and American Heart Association guidelines[14] (**Supplementary Table 1**). Diseases that could not be classified into these 3 groups were defined as "others". If a patient had multiple ICD-10 codes related to CHD, the disease complexity was classified based on the most complex code.[15] For patients with ICD-10 codes encompassing multiple CHD diagnoses with different disease complexity classifications, the disease complexity was classified by referring to the manually entered diagnoses. For example, diagnoses with ICD-10 code Q248 include "double-chambered right ventricle" and "single atrium single ventricle". Therefore, based on the manually entered diagnoses, "double-chambered right ventricle" was classified as "moderate" and "single atrium single ventricle" was classified as "great". The classification of patients as "others" or with different levels of complexity was reaffirmed by 2 researchers (N.A., R.O.).

Emergency Admission Emergency admission was defined using specific DPC variables.

Heart Failure Admission Hospitalization for heart failure was defined as including the ICD-10 code "I50" in either the "main diagnosis", "admission-precipitating diagnosis", or "the most resource-consuming diagnosis" in order to identify those admissions due to heart failure.[16]

CHD Admission CHD admission was defined as having a CHD-related ICD-10 diagnosis in either the "main diagnosis", "admission-precipitating diagnosis", or "the most resource-consuming diagnosis" in order to identify those admissions due to CHD.

Admission to a JSPCCS-Certified Hospital We defined JSPCCS-certified hospitals as those that had been certified by the JSPCCS as of 2017. A JSPCCS-certified hospital is a training facility for pediatric cardiologists specializing in pediatric cardiology. The admission of patients to one of these hospitals was defined as an admission to a JSPCCS-certified hospital. JSPCCS-certified hospitals were determined based on their accreditation status as of 2017 because accreditation at that time was based on past practice and was considered to reflect the quality of care at the facility during the study period.

Distance Between Home and Hospital The distance between home and hospital was calculated as the straight-line distance between the patient's home and hospital, based on zip codes.

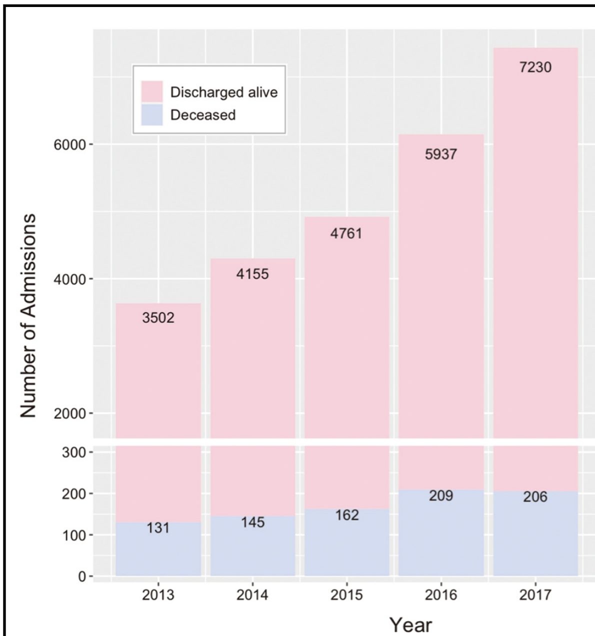

Figure 2. Annual changes in hospitalizations and deaths (n=26,438) for fiscal years 2013–2017.

Location of Death The location of death was defined as the ICU if the patient was in the ICU the day before death and as the hospital ward for remaining cases of in-hospital deaths. The day before death was used in defining the location of death because, in some cases, the day after the date of death was used as the discharge date. If we had defined the location of death as the ICU only if the deceased patient was in the ICU on the day of discharge, then if the patient had died in the ICU but the day after their death was the day of discharge, the patient would be classified as having died outside the ICU (i.e., having died on the hospital ward). Therefore, we decided to use the location on the day before death to define the location of death.

Cardiac Intervention Cardiac intervention was defined as surgery and catheter intervention corresponding to the DPC procedure K codes, presented in **Supplementary Table 2**. PCPS and IABP were not included as cardiac interventions in the present study and were tabulated separately.

Statistical Analysis

To emphasize the EOL status, this study focused on the last hospitalization of eligible patients and compared the data of patients who were discharged alive with those of patients who died. First, we calculated the number of hospitalizations and deaths at all hospitals for each fiscal year during the study period. In addition, we only used the datasets from hospitals that submitted DPC data for 5 consecutive fiscal years to confirm trends in the numbers of hospitalizations and deaths over time during the study period.

Descriptive statistics were calculated for patient demographics, admission paths, facility types, and medical treatment administered during hospitalization to patients

Table 1. Comparison of Patients Discharge Alive and Those Who Died in Hospital (n=26,438)

	Discharged alive (n=25,585)	Deceased (n=853)
Age (years)	54.0 [32–70]	69.0 [45–81]
Sex		
Male	12,349 (48.3)	416 (48.8)
Female	13,236 (51.7)	437 (51.2)
Complexity		
Simple	15,525 (60.7)	429 (50.3)
Moderate	5,722 (22.4)	230 (27.0)
Great	3,598 (14.1)	172 (20.2)
Other	740 (2.9)	22 (2.6)
Length of hospital stay (days)	9 [4–18]	12 [3–32]
Emergency admission (n=26,435)		
Yes	5,871 (22.9)	705 (82.6)
Heart failure admission		
Yes	2,681 (10.5)	367 (43.0)
CHD admission		
Yes	12,814 (50.1)	185 (21.7)
Experience of ICU stay		
Yes	7,334 (28.7)	353 (41.4)
Length of ICU stay (days; n=7,687)	2 [1–4]	8 [3–14]
Cardiac intervention		
Yes	11,696 (45.7)	183 (21.5)
In-hospital CPR (n=25,537)		
Yes	1,106 (4.3)	291 (34.2)
IABP		
Yes	117 (0.5)	75 (8.8)
PCPS		
Yes	36 (0.1)	98 (11.5)
Dialysis		
Yes	324 (1.3)	63 (7.4)
Mechanical ventilation		
Yes	4,218 (16.5)	562 (65.9)
Admission to JSPCCS-certified hospital		
Yes	13,151 (51.4)	251 (29.4)
Distance between hospital and home (km; n=26,392)	10.5 [4.2–26.4]	6.0 [2.9–13.9]
No. hospitalizations within 1 year		
2	4,601 (18.0)	131 (15.4)
≥3	1,103 (4.3)	66 (7.7)

Data are presented as n (%) or median [interquartile range]. CHD, congenital heart disease; CPR, cardiopulmonary resuscitation; IABP, intra-aortic balloon pumping; ICU, intensive care unit; JSPCCS, Japanese Society of Pediatric Cardiology and Cardiac Surgery; PCPS, percutaneous cardiopulmonary support.

discharged alive and to those who died. Data for patients who died were compared by disease complexity.

Continuous variables are presented as the mean±SD for normally distributed data and as the median with interquartile range (IQR) for non-normally distributed data. Categorical data are presented as frequencies and percentages and were compared by complexity using Pearson's Chi-squared test. Continuous variables were analyzed by complexity using the Kruskal-Wallis test. Analyses were performed using SPSS version 28 (IBM Corp., Armonk, NY, USA), and the figures were created using R version 4.1.2. (R Foundation for Statistical Computing, Vienna, Austria).

Ethical Considerations

This study was approved by the Ethics Committee of Tsukuba University (Approval no. 1543-2) and was conducted in compliance with the Declaration of Helsinki. The requirement for informed consent was waived because of the retrospective anonymized nature of the data.

Results

Comparison Between Patients Discharged Alive and Those Who Died

During the 5-year study period, there were 26,438 last admissions of patients with CHD, including 853 patients who died. **Figure 2** shows the numbers of hospitalizations and deaths between 2013 and 2017. Through 2016, the number of both hospitalizations and patient deaths increased, whereas in 2017 the number of deceased patients

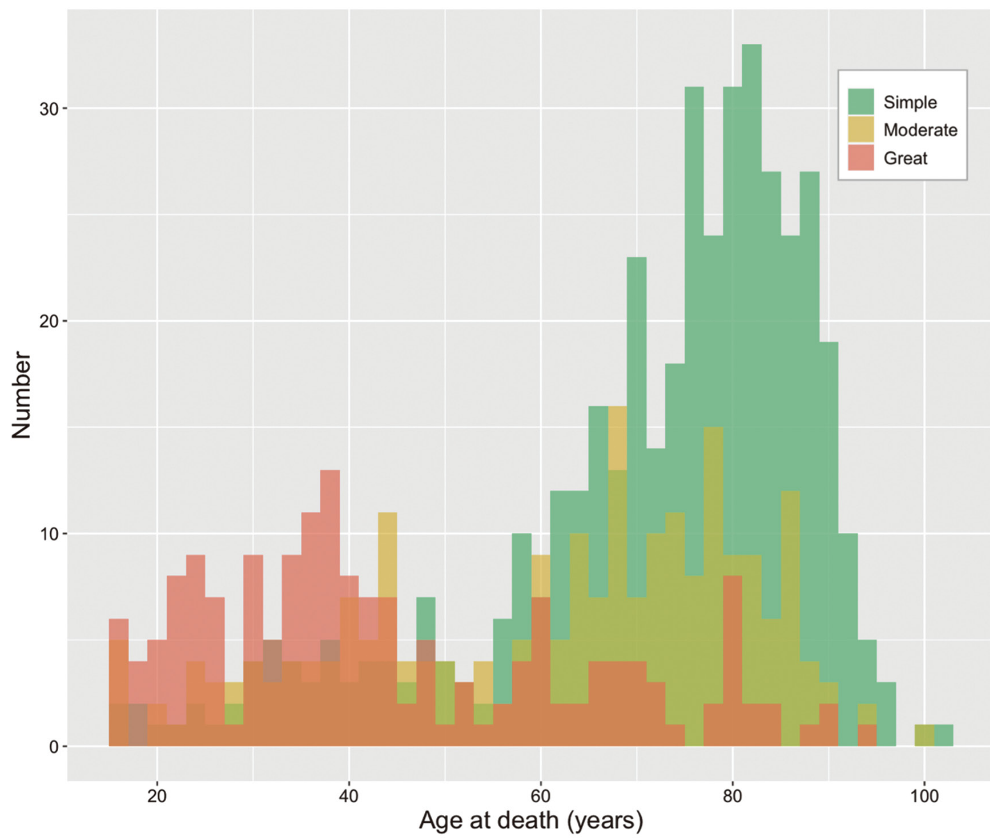

Figure 3. Age at death by disease complexity (simple, moderate, or great).

increased less compared with the number of hospitalizations. The analysis using only the datasets of hospitals that submitted DPC data for 5 consecutive fiscal years showed similar trends in the number of hospitalizations and deaths (**Supplementary Figure**).

The demographic characteristics of patients who were discharged alive and those who died are presented in **Table 1**. Compared with patients discharged alive, the patients who died were older and had higher emergency and heart failure admission rates and a lower CHD admission rate. The cardiac intervention rate was 45.7% among patients discharged alive and 21.5% among the patients who died. The patients who deceased were more commonly treated with PCPS, IABP, dialysis, and mechanical ventilation.

Characteristics of Deceased Patients According to Disease Complexity

A comparison of disease complexity among the patients who died revealed that those with greater disease complexity died at a younger age. Twenty-two patients classified as "others" were excluded from this comparison because their detailed anatomical features were unknown. **Figure 3** shows age at death by disease complexity. The median age at death was 77.0, 66.5, and 39.0 years for the simple, moderate, and great complexity groups, respectively.

Table 2 presents demographic characteristics of the patients who deceased according to disease complexity. Emergency hospitalization rates exceeded 70% for all disease complexity groups; heart failure admission was more common in the simple complexity group (49.9%), and CHD admission was more common in the great complexity group (39.0%).

Admission to the ICU occurred in 45–50% of patients with any complexity level. The median ICU stay was 7 days for patients in the simple disease complexity group, 6 days for patients in the moderate disease complexity, and 10.5 days for patients in the great disease complexity group. Approximately 20–30% of patients with any complexity level died in the ICU.

Cardiac intervention was most common in the moderate disease complexity group (33.9%). The treatments administered before death among patients in the simple, moderate, and great complexity groups included cardiopulmonary resuscitation (CPR; 30.1%, 35.7%, and 41.9%, respectively), PCPS (7.2%, 16.5%, and 16.3%, respectively), dialysis (7.5%, 7.8%, and 7.0%, respectively), and mechanical ventilation (58.7%, 72.2%, and 75.6%, respectively).

The percentage of patients admitted to a JSPCCS-certified hospital increased with increasing disease complexity. The median distance between the hospital and patient residences was greatest in the great disease complexity group (9.8 km).

The number of hospitalizations to the same hospital within the year before death was highest among patients in the great disease complexity group, with 36.1% of patients undergoing ≥2 hospitalizations. The number of days from admission to death did not differ according to disease com-

Table 2. Characteristics of Deceased Patients According to Disease Complexity

	Simple (n=429; 51.6%)	Moderate (n=230; 27.7%)	Great (n=172; 20.7%)	P value
Age (years)	77.0 [65.0–84.0]	66.5 [45.0–78.0]	39.0 [30.5–60.5]	<0.001[A]
Sex				
Male	191 (44.5)	117 (50.9)	95 (55.2)	0.042[B]
Female	238 (55.5)	113 (49.1)	77 (44.8)	
Length of hospital stay (days)	13 [3–30]	11.5 [3–31]	14 [4–36]	0.401[A]
Emergency admission				
Yes	373 (86.9)	171 (74.3)	141 (82.0)	<0.001[B]
Heart failure admission				
Yes	214 (49.9)	78 (33.9)	68 (39.5)	<0.001[B]
CHD admission				
Yes	52 (12.1)	62 (27.0)	67 (39.0)	<0.001[B]
ICU stay				
Yes	163 (38.0)	111 (48.3)	72 (41.9)	0.039[B]
Length of ICU stay (days; n=353)	7 [3–14]	6 [3–14]	10.5 [5.5–14]	0.044[A]
Location of death				
Ward	345 (80.4)	156 (67.8)	135 (78.5)	0.001[B]
ICU	84 (19.6)	74 (32.2)	37 (21.5)	
Cardiac intervention				
Yes	73 (17.0)	78 (33.9)	28 (16.3)	<0.001[B]
In-hospital CPR				
Yes	129 (30.1)	82 (35.7)	72 (41.9)	0.019[B]
IABP				
Yes	34 (7.9)	26 (11.3)	13 (7.6)	0.281[B]
PCPS				
Yes	31 (7.2)	38 (16.5)	28 (16.3)	<0.001[B]
Dialysis				
Yes	32 (7.5)	18 (7.8)	12 (7.0)	0.950[B]
Mechanical ventilation				
Yes	252 (58.7)	166 (72.2)	130 (75.6)	<0.001[B]
Admission to JSPCCS-certified hospital				
Yes	82 (19.1)	70 (30.4)	89 (51.7)	<0.001[B]
Distance between hospital and home (km; n=849)	4.9 [2.6–10.9]	6.0 [3.1–14.9]	9.8 [5.0–24.8]	<0.001[A]
No. readmissions within 1 year				
2	59 (13.8)	36 (15.7)	34 (19.8)	<0.001[B]
≥3	22 (5.1)	15 (6.5)	28 (16.3)	
Time from admission to death				
Death on admission	23 (5.4)	10 (4.3)	11 (6.4)	0.384[B]
24 h	51 (11.9)	37 (16.1)	17 (9.9)	
2–7 days	85 (19.8)	46 (20.0)	33 (19.2)	
8–30 days	163 (38.0)	78 (33.9)	55 (32.0)	
≥31 days	107 (24.9)	59 (25.7)	56 (32.6)	

Unless indicated otherwise, data are presented as n (%) or median [interquartile range]. [A]Kruskal-Wallis test. [B]Pearson's Chi-squared test. Abbreviations as in Table 1.

plexity; however, the highest percentage of deaths occurred in the great disease complexity group, ≥31 days after admission.

Discussion

This is the first study to use nationwide data to describe the EOL status of patients with ACHD. Our primary findings were that: (1) patients with greater disease complexity died at a younger age; (2) patients with greater disease complexity underwent more invasive treatments during the last admission; and (3) 70% of patients died outside of specialized facilities, with >25% dying after ≥31 days of hospitalization.

Greater Disease Complexity Associated With Earlier Age at Death

Advances in the diagnosis, intervention, surgical management, and care of patients with CHD have led to an increase in the number of adults with CHD.[2,17] The present study found that Japanese patients with ACHD, particularly those with greater disease complexity, were younger at the time of death compared with the general population. In 2017, the average life expectancy in Japan was 81 and 87 years for men and women, respectively,[18] whereas the average age of patients with heart failure in Japan is 78 years.[19] Furthermore, the average age of death among patients with heart failure in 7 European countries is reportedly 83 years.[20] Despite medical advances, the life expectancy of patients with ACHD is extremely short compared with that of the general population and patients with heart failure.

Studies conducted in other countries have reported a young age at the time of death among patients with ACHD.[21–25] In the UK, the median age at death of 524 patients with CHD was reportedly 47 years (IQR 34–65 years),[21] whereas in Belgium the median age at death of 390 patients was reportedly 55 years (IQR 40–73 years).[25] The median age at death in the present study was 69 years (IQR 45–81 years), which is higher than that reported in other studies. One possible reason for this may be differences in disease complexity in participants among the studies.[22,25] Although the facilities included in the present study were limited to acute care hospitals in Japan, the distribution of disease complexity may reflect the overall situation in Japan.

In a study of patients' expectations of life expectancy, young patients with ACHD tended to estimate their life expectancy to be longer, with 87% of patients with moderate disease complexity and 68% of patients with great disease complexity estimating their life expectancy to be ≥55 years.[26] In general, the overall health condition is more stable at a younger age and the patient may not face rapid disease deterioration. However, the median age of death in the present study was 39 years in the great disease complexity group. Sixty-one percent of patients with ACHD request information on estimated life expectancy.[27] Information on prognosis should be provided to patients according to the currently available evidence, the patient's physical condition, and the patient's preferences.

Patients With Greater Disease Complexity Underwent More Invasive Treatment Before Death

We compared patients who died according to disease complexity and found that the greater the complexity, the higher the percentage of patients requiring in-hospital CPR, PCPS, or mechanical ventilation. CPR, mechanical ventilation and IABP, PCPS, and dialysis are reportedly more frequently performed in patients with cardiac disease before death than in patients with cancer.[28] Although the treatment approaches for cardiac disease and cancer are different, the reasons for the high rate of invasive treatment in patients with cardiac disease may be that mechanical treatment is a component of heart failure treatment[29] and that the prognosis for heart failure is uncertain.

Patients with CHD are at higher risk of sudden death and are more likely to undergo life-saving invasive treatments.[4] One study reported that 44% of patients who died were mechanically ventilated at the time of death and that 52% died during resuscitation.[23] A study on patient wishes for advance care planning involving adolescents with heart disease showed that 92% of patients wanted to be involved in decisions about their EOL,[30] whereas 55% of patients could not decide what treatment they wished to receive in the event of serious illness.[31] Certainly, it is difficult for patients to think about the type of invasive treatment they would like to receive at the EOL. However, it may be possible to consider "a good death"[32] (a painless death, with dignity preserved) and think about how they would like to live in the event of serious illness. The current EOL status revealed in the present study, with younger age at death and patients with ACHD with more complex disease undergoing more invasive treatment, may serve as a resource for patients and healthcare providers when considering patients with ACHD, advance care planning, and EOL.

Seventy Percent of Patients Died Outside of Specialized Facilities and >25% of Patients Died After ≥31 Days of Hospitalization

Among the patients who were admitted to JSPCCS-certified hospitals, 51.4% were discharged alive, whereas 29.4% died. Although it is recommended that patients with CHD be referred to a specialized facility,[13] it is likely that patients cannot be admitted to such a facility because of life-saving priorities in hospitalization prior to death. However, it was also found that patients who died had been admitted to JSPCCS-certified hospitals with greater disease complexity, and more complex patients were admitted to more distant hospitals. This indicates that only a limited number of facilities can provide treatment to patients with greater disease complexity. The certification of facilities specializing in ACHD started in 2019, and a system of care is currently being developed. As the number of ACHD-specialized facilities increases, the proportion of patients who die in specialized facilities may increase. However, reinforcing the network with specialized facilities may increase the proportion of patients who die in non-specialized hospitals. The development of the care system and the location of deaths should be evaluated over time.

Although the JROAD-DPC is an acute care hospital database, >25% of patients with any disease complexity died ≥31 days after admission. A previous study from the US showed that, compared with patients with cancer, patients with ACHD were more likely to be admitted to the hospital 30 days before death and to the ICU or emergency department.[7] Prolonged hospitalization is expected to increase the burden of medical costs. In patients with cancer, financial problems were reportedly associated with a psychological burden on the bereaved family members.[33] In the field of ACHD research, the burden of medical costs at the EOL and the psychological burden on bereaved families should also be investigated.

Study Limitations

First, the JROAD-DPC database is specific to acute care hospitals in Japan and carries an inherent selection bias because it includes only patients with CHD who were admitted to affiliated facilities. However, considering that 80% of patients in Japan die in hospitals, and younger patients with CHD are more likely to die in hospitals,[18] this database is considered to be representative of the majority of patients with ACHD. Furthermore, the JROAD-DPC database lacks comprehensive information on diagnosis, including detailed anatomical conditions (e.g., single right or left ventricle), comorbidities other than CHD, laboratory data, and the cause of death. In addition, DPC data do not include personal identifiable information; therefore, hospitalizations to other facilities cannot be tracked. To obtain a more detailed follow-up of the outcomes of all patients with CHD, it is essential to establish a registry and conduct studies that integrate medical records.

Second, the validity of the diagnosis and complexity of the classification of patients with CHD in the JROAD-DPC database is yet to be verified. In the present study disease complexity was classified using a highly reliable method based on previous studies.[15] Future validation studies should be conducted to identify patients using validated and highly reliable methods.

Third, the numbers of both hospital admissions and deceased patients increased until 2016, although in 2017 the number of deceased patients increased less compared with the number of hospitalizations. To our knowledge, no new treatments were introduced that significantly improved the prognosis of patients with ACHD in Japan during this period; changes in the number of deaths beyond 2018 and the underlying causes of these changes should be examined in future studies.

Conclusions

Based on the JROAD-DPC data, patients with ACHD with greater disease complexity die at a younger age, undergo more invasive treatments before death, and many die after a month or more of hospitalization. Discussing EOL options with patients at the appropriate time is important, particularly for patients with greater disease complexity.

Acknowledgments

The authors thank the participants of the JROAD-DPC for their contributions and the JROAD-DPC registry facilities for maintaining the registry.

Sources of Funding

This study was supported by the fifth Strategic Research Promotion Project of Yokohama City University (to R.O.).

Disclosures

The authors declare that there are no conflicts of interest.

IRB Information

This study was approved by the Ethics Committee of Tsukuba University (Approval No. 1543-2) and was conducted in accordance with the Declaration of Helsinki.

Data Availability

The deidentified participant data will not be shared. The JROAD-DPC database provided confidential information; therefore, the dataset cannot be made public. We only analyzed data from the National Cerebral and Cardiovascular Center.

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Supplementary Files

Please find supplementary file(s);
https://doi.org/10.1253/circj.CJ-23-0537