Original Article

Social Independence and Lifestyles in Patients with Repaired Tetralogy of Fallot—Secondary Publication

Ryoji Shinbara, RN, MSc[1)], Hiroyuki Sawatari, PhD[2)], Keiko Yamasaki, RN, PhD[3)], Minjeong Kang, RN, PhD[4)], Ichiro Sakamoto, MD[5)], Kenichiro Yamamura, MD, PhD, MSc[6)], Hazumu Nagata, MD, PhD[6)], Hiroyuki Tsutsui, MD, PhD[5)], Hiroaki Chishaki, MD[7)], Tomotake Tokunou, MD, PhD[8)], and Akiko Chishaki, MD, PhD[3, 9)]

[1)] Department of Nursing, Kyushu University Hospital, Fukuoka, Japan
[2)] Department of Perioperative and Critical Care Management, Graduate School of Biomedical and Heath Sciences, Hiroshima University, Hiroshima, Japan
[3)] Faculty of Nursing, Department of Nursing, Fukuoka Nursing College, Fukuoka, Japan
[4)] Faculty of Nursing, Daiichi University of Pharmacy, Fukuoka, Japan
[5)] Department of Cardiovascular Medicine, Graduate School of Medical Sciences, Kyushu University, Fukuoka, Japan
[6)] Department of Pediatrics, Graduate School of Medical Sciences, Kyushu University, Fukuoka, Japan
[7)] Department of Healthcare Management, College of Healthcare Management, Fukuoka, Japan
[8)] Department of Internal Medicine, Kyushu University Beppu Hospital, Oita, Japan
[9)] Health Care Center, Fukuoka Dental College Hospital, Fukuoka, Japan

Background: The purpose of this study was to look at the current state of the adult patients with tetralogy of Fallot, the most common cyanotic congenital heart disease, to encourage social independence and healthy lifestyles.
Methods: On 186 patients, a questionnaire survey (understanding and anxiety about their heart diseases, treatments, social independence, and lifestyles) was administered. These data were also compared between patients with and without physical disability certification (a certified group and a noncertified group). Clinical data were extracted from the medical records.
Results: After excluding the cases without meeting the inclusion criteria, 112 patients (41 males, mean age 28 years) were studied. Eighty-three percent of 93 patients after excluding 19 students, were employed (66% full-time employee) , half of them lived with their parents, and 71% were concerned about their heart diseases. In terms of lifestyle, 28% were dissatisfied with the quality of their sleep. The noncertified group (n = 59) was assigned more professional tasks, whereas the certified group (n = 53) was assigned more office duties. The certified group had more regular outpatient clinic visits and dental consultations, but also had a greater experience to drink alcohol and take a sleeping pill.
Conclusion: The study patients had a relatively good job rate and a high level of social independence, despite having a variety of anxiety disorders. It was suggested that some supports for anxiety and sleep disorder issues be implemented especially in the certified group.

Keywords: tetralogy of Fallot, social independence, lifestyles

Introduction

Advancement in medicine has enabled many patients with congenital heart disease (CHD) to grow into adulthood.[1)] Those with moderate or complex CHD often face with residual problems and sequelae that need treatments in the long term. Clinical guidelines have been established for the patients with Adult Congenital Heart

Received: January 17, 2024; Accepted: June 21, 2024 J-STAGE Advance Published Online: January 17, 2025
Corresponding author: Akiko Chishaki, MD, PhD, Health Care Center, Fukuoka Dental College Hospital, 2–15–1 Tamura, Sawara-ku, Fukuoka 814–0193, Japan
E-mail: chishaki@fdcnet.ac.jp
Akiko Chishaki (https://orcid.org/0000-0003-1495-3876)
Originally published in Pediatric Cardiology and Cardiac Surgery, Vol. 38 (2022), pp. 128–139. [in Japanese] doi: 10.9794/jspccs.38.128
doi: 10.24509/jpccs.24-002

Diseases (ACHD) not only in terms of medical problems but also social problems, such as education, employment, marriage, pregnancy and child bearing, genetics, and social security.[2, 3] Moreover, social independence of the patients with ACHD has gathered recognition as an important outcome but very few studies have been reported as to this outcome in Japan. There are multiple definitions of social independence, and previous studies defined it using education level, employment status, or whether living with parents or not.[4, 5] Several studies investigated employment status and educational history of patients with ACHD, whereas only a few have dealt with anxiety and difficulty in lifestyles and social activities.

ACHD patients are confronted with continuous struggles against CHD since birth and considerable financial burden. On top of severity of their disease, it has been reported that whether they are officially registered with physical disability certificate (PDC) or not influences various aspects; the frequency of outpatient visits for follow-up, social independence such as employment, marriage and child rearing.[6–8] We expect a similar circumstance in our survey. The PDC is not necessarily approved in patients with significant physical status. Evaluation bias by the physician in charge would affect official approval. The PDC system is also managed under other non-medical and complicated factors. These issues overall seem to restrict practical lives of ACHD patients.

In this study, we conducted a cross-sectional survey on clinical characteristics, patients' knowledge and anxiety about their CHD (diagnosis and treatment), social independence, and lifestyles (smoking, drinking, and sleeping). We focused on patients with tetralogy of Fallot (TOF), the most common type of cyanotic heart disease, accounting for 11.3% of all CHD.[9] The purpose of this study was to obtain a basis for providing health guidance and patient education according to the PDC, taking "severity of cardiac disease" and "access to the social security system" into consideration. These two aspects are likely related with social independence.

Methods

Subjects

This study initially included 186 patients with TOF aged from 18 to 65 years, the working-age population, who transferred to or actually visited ACHD outpatient clinic at Kyushu University Hospital between June 2017 and March 2019. During the waiting time of the outpatient appointment, trained nurses explained them about the purpose and the content of this study, and then obtained an informed consent. We collected responses from 178 patients (collection rate 95.7%). The questionnaires were filled out by themselves without help of an accompanying person. The trained nurse explained that the answers should not be guided by anyone in case those patients needed assistance when filling out the form. Out of 178 patients, we excluded 66 patients: 53 having pulmonary atresia with ventricular septal defect (the severest form of TOF), 6 with chromosomal aberrations, 4 with intellectual disability, 2 with unrepaired TOF, and 1 as a candidate for cardiac transplantation. Thus, 112 patients with TOF (valid responses rate 62.9%) were included for the final analysis. Sixty-nine patients (61.6% of all patients for the final analysis) transitioned after attending patient education programs for transitional support at the outpatient clinic of their previous pediatric institutions.

Survey Content

1) Questionnaire Survey

This survey was conducted using a self-administered questionnaire which included the following contents: recognition of their own heart disease (diagnosis, treatment history), symptoms on physical activity with which patients are stratified according to New York Heart Association (NYHA) functional class, thoughts on transition from pediatric to adult cardiology, anxiety regarding heart problems and treatments (worsening of medical condition, complications, genetic issues, medical examinations, reoperation, timing of further treatments, specialized facilities, side effects/duration/amount of medications, medical costs, availability of social security system, whether the PDC qualified or not, life insurance, school attendance, employment, marriage, pregnancy, childbirth, child rearing, and life expectancy), and who to ask for practical advice. We also investigated the status of social independence based on previous studies,[4, 5] including employment status, household status, contents of work, educational background, and use of social security systems. Anxiety and difficulty in student life and social life (getting employed and continued employment) were assessed in relation to CHD. As for lifestyle, history of drinking and smoking, regular dental consultation, sleep status (taking of sleeping pill, sleep

latency, sleep length, and sleep satisfaction), height, and weight were investigated. Drinking habit was designated as occasional drinking, or habitual drinking (alcohol intake equivalent to Japanese sake 180 mL or more per day, 3 days or more a week). In the analysis, habitual drinking was regarded as a form of alcohol intake which would have undeniable adverse effects on health.

Responses were obtained in three ways; respondents were asked to write down specific numbers or words according to the content of questions, to choose one from a couple of options such as "Yes" or "No," or to choose one from a five-point scale (strongly disagree, disagree, nether agree nor disagree, agree, strongly agree) according to the Likert scale.[10)]

Free description was also utilized for anxiety and any additional comments regarding disease and daily life.

2) Medical Records Survey

We retrospectively collected data from the medical records, including CHD diagnosis, previous cardiac surgery, age at this study, age at initial intracardiac repair, and medical history. To estimate how severe their heart diseases were, we investigated whether a pulmonary arterial lesion had been reoperated, whether cardiovascular events had occurred (arrhythmias and heart failure requiring treatments), plasma levels of brain natriuretic peptide (BNP), chest X-ray (cardiothoracic ratio), echocardiographic and catheterization data, status of residua and sequelae of CHD, as well as presence or absence of comorbid diseases and regular use of medication. Clinical information was basically obtained on the day of the questionnaire survey (date of survey). In addition, investigation data dated within ±1 year of the date of survey were referred for blood tests, chest X-ray, and echocardiography. Data related to residua and sequelae of CHD were based on echocardiographic and catheterization findings as well as surgical records.

Ethical Consideration

This study was approved by the Institutional Review Board/Ethics Committees of the Kyushu University Hospital and Medical Institutions (approval #2019-043). The content and the relevance of the study were explained to the respondents in writing and they signed the consent form.

Statistical Analysis

Considering that "severity of CHD" and "use of social security system" affect social independence of the patients, we divided them into two groups according to the status of acquisition of the PDC (Grade 1, 3, or 4); namely, the PDC group (certified group) versus the non-PDC group (non-certified group). We analyzed differences between these groups using Student's *t*-test or Mann–Whitney *U* test following Kolmogorov–Smirnov test. For binary data, χ^2 test was used. Continuous variables are shown as a mean ± a standard deviation in the case of a normal distribution, and categorical variables are shown as percentage. Propensity score matching (Nearest neighbor method, Caliper = 0.20) was used to adjust for confounding by age at this study in the clinical characteristics (SPSS v21.0, IBM Inc., Chicago, IL). The frequency of the questionnaire items was determined by simple descriptive statistics, and we considered a two-sided p value <0.05 as statistically significant and $0.05 \leq p$ value <0.10 as marginally significant.

Results

Among 112 patients with TOF, the certified group contained 53 patients (47.3%) and the non-certified group 59 patients (52.7%). We included 13 patients under social security systems other than the PDC; that is, specific medical expenses, rehabilitation certificate, and medical subsidies for specific pediatric chronic diseases. Their data were categorized and analyzed up to whether they are with or without the PDC qualified.

Medical Records Information

Clinical Characteristics (Table 1)

The mean age of overall patients was 28 years, 36.6% of them were male, and approximately 90% of them were in NYHA functional class I–II. In terms of cardiac surgery, 23 patients had undergone a total of 26 palliative operations. Intracardiac repair was achieved in all patients (including the Rastelli procedure in 4). In total, 52 reoperations were conducted in 47 patients for pulmonary arterial lesions. A total of 25 cardiovascular events occurred in 20 patients (17.9%), including 18 arrhythmias and 2 heart failure requiring treatments. Of the remaining 5, infective endocarditis was seen in 2, angina pectoris in 1, brain abscess in 1, and transient ischemic attack in 1. In terms of pharmacotherapy, approximately 37% of the patients were on certain medication.

Compared with the non-certified group, a mean age of the certified group was significantly higher (31.3 ±

Table 1 Clinical characteristics of total TOF patients

	All patients $N=112$	PDC (−) $n=59$	PDC (+) $n=53$	p-value
Age				
at this study, years (range: 18–55)	28.3±8.4	25.6±5.9	31.3±9.6	<0.001
at ICR, years (range: 0–22)	2.4±3.0	2.1±2.5	2.8±3.4	0.095
Male, n (%)	41 (36.6)	21 (35.6)	20 (37.7)	0.846
NYHA functional classification				
Class I–II, n (%)	98(87.5)	56 (94.9)	42 (79.2)	0.020
Class III–IV, n (%)	14 (12.5)	3 (5.1)	11 (20.8)	
Physical findings				
Height, cm	159.9±8.5	160.2±9.0	159.5±7.8	0.856
Weight, Kg	54.6±12.3	56.1±13.8	53.0±10.4	0.454
BMI, Kg/m^2	21.2±3.6	21.7±3.8	20.8±3.4	0.312
Cardiac operations				
Palliative operation before ICR, n (%)	23 (20.5)	12 (20.3)	11 (20.8)	1.000
Rastelli procedure, n (%)	4 (3.6)	2 (3.4)	2 (3.8)	1.000
Reoperation for PA lesion, n (%)	47 (42.0)	11 (18.6)	36 (67.9)	<0.001
Cardiovascular events				
Total pt. with CVE, n (%)	20 (17.9)	2 (3.4)	18 (34.0)	<0.001
Frequency of CVE occurrence				
1 time	16 (14.3)	2 (3.4)	14 (26.4)	<0.001
2 times	3 (2.7)	0 (0.0)	3 (5.7)	
3 times	1 (0.9)	0 (0.0)	1 (1.9)	
Types of CVE				
Arrhythmia, times (%)	18 (16.1)	1 (1.7)	17 (32.1)	<0.001
Heart failure, times (%)	2 (1.8)	1 (1.7)	1 (1.9)	0.939
Others, times (%)	5 (4.5)	0 (0.0)	5 (9.4)	0.016
Device treatments				
Permanent pacemaker implantation, n (%)	4 (3.6)	0 (0.0)	4 (7.5)	0.047
Medications				
Total pt. with medications, n (%)	41 (36.6)	14 (23.7)	27 (50.9)	0.003
ACE-I/ARB, n (%)	20 (17.9)	9 (15.3)	11 (20.8)	0.470
Anticoagulant/Antiplatelet drugs, n (%)	17 (15.2)	5 (8.5)	12 (22.6)	0.063
β-blockers, n (%)	9 (8.0)	1 (1.7)	8 (15.1)	0.013
Diuretics, n (%)	9 (8.0)	3 (5.1)	6 (11.3)	0.303
Others, n (%)	6 (5.4)	5 (9.4)	1 (1.7)	0.099
Laboratory data				
BNP, pg/dL	29.1±34.9	21.4±18.1	37.7±45.7	0.007
Chest X-ray ($N=111$)				
CTR, %	50.7±5.6	49.0±4.6	52.6±6.1	<0.001

Physical disability certificate (PDC) was considered to be the suitable criterion for comprehensive grouping because it reflects both disease severity and social life influencing factors. Data are expressed as number of patients (%) or mean±standard deviation. ACE-I, angiotensin converting enzyme inhibitor; ARB, angiotensin receptor blocker; BMI, body mass index; BNP, brain natriuretic peptide; CTR, cardiothoracic ratio, CVE, Cardiovascular events; ICR, intra cardiac repair; NYHA, New York Heart Association; PA, pulmonary artery; pt, patients; TOF, Tetralogy of Fallot.

9.6 years old vs. non-certified group 25.6±5.9 years old, $p<0.001$) and NYHA class III–IV was more common (20.8% vs. 5.1%, $p<0.05$). Reoperation, cardiovascular events (especially arrhythmias), and staying on regular medication were also more frequent in the certified group than in the non-certified group. Plasma level of BNP was higher and cardiothoracic ratio on chest X-ray was greater in the former than in the latter.

The types of CHD were significantly more complicated in the certified group. We examined the clinical background of the groups by adjusting for age at survey using propensity score matching and looked into whether this was confounded by the significantly higher age at survey in the certified group (Table 2). Even after the adjustment, the certified group still had a significantly higher proportion of patients with NYHA class III–IV (23.5% vs. 5.6% in the non-certified group, $p<0.05$), a higher reoperation rate (73.5% vs. 19.4%, $p<0.001$), a greater incidence of cardiovascular events (26.5% vs. 5.6%, $p<0.05$), a higher rate of staying on medications (50.0% vs. 19.4%, $p<0.01$), and a greater cardiothoracic ratio (52.3±5.2% vs. 48.4±4.3%, $p<0.001$) than the non-certified group did.

Of all patients, 80 patients (62.5%) had either residua, sequelae, or complications. There was no significant difference between the two groups with regard to these

Table 2 Clinical characteristics adjusted the age by propensity score matching

	All patients $N=70$	PDC (−) $n=36$	PDC (+) $n=34$	p-value
Age				
at this study, years (range: 18–42)	26.7±5.6	27.6±6.5	25.7±4.3	0.364
at ICR, years (range: 0–10)	2.2±2.5	2.9±2.9	1.6±1.8	0.051
Male, n (%)	26 (37.1)	13 (36.1)	13 (38.2)	0.854
NYHA functional classification				
Class I–II, n (%)	60 (85.7)	34 (94.4)	26 (76.5)	0.032
Class III–IV, n (%)	10 (14.3)	2 (5.6)	8 (23.5)	
Physical findings				
Height, cm	159.4±8.4	161.2±9.3	157.5±7.1	0.068
Weight, Kg	54.5±11.8	58.0±13.8	50.7±7.8	0.022
BMI, Kg/m^2	21.3±3.3	22.2±3.9	20.4±2.4	0.074
Cardiac operations				
Palliative operation before ICR, n (%)	12 (17.1)	6 (16.7)	6 (17.6)	0.913
Rastelli procedure, n (%)	3 (4.3)	1 (2.8)	2 (5.9)	0.522
Reoperation for PA lesion, n (%)	32 (45.7)	7 (19.4)	25 (73.5)	<0.001
Cardiovascular events				
Total pt. with CVE, n (%)	11 (15.7)	2 (5.6)	9 (26.5)	0.016
Frequency of CVE occurrence				0.097
1 time	8 (11.4)	2 (5.6)	6 (17.6)	0.097
2 times	2 (2.9)	0 (0.0)	2 (5.9)	
3 times	1 (1.4)	0 (0.0)	1 (2.9)	
Types of CVE				
Arrhythmia, times (%)	11 (15.7)	1 (2.8)	10 (29.4)	0.002
Heart failure, times (%)	1 (1.4)	1 (2.8)	0 (0.0)	0.328
Others, times (%)	3 (4.3)	0 (0.0)	3 (8.8)	0.068
Device treatments				
Permanent pacemaker implantation, n (%)	3 (4.3)	0 (0.0)	3 (8.8)	0.068
Medications				
Total pt. with medications, n (%)	24 (34.3)	7 (19.4)	17 (50.0)	0.007
ACE-I/ARB, n (%)	10 (14.3)	3 (8.3)	7 (20.6)	0.143
Anticoagulant/Antiplatelet drugs, n (%)	11 (15.7)	4 (11.1)	7 (20.6)	0.276
β-blockers, n (%)	4 (5.7)	0 (0.0)	4 (11.8)	0.034
Diuretics, n (%)	5 (7.1)	2 (5.6)	3 (8.8)	0.596
Others, n (%)	8 (11.4)	2 (5.6)	6 (17.6)	0.112
Laboratory data				
BNP, pg/dL	26.1±24.1	24.1±21.3	28.2±26.9	0.391
Chest X-ray				
CTR, %	50.3±5.2	48.4±4.3	52.3±5.2	<0.001

After adjusting the confounding factor: age, the certified group still showed severer cardiac state than that of non-certified group. Data are expressed as number of patients (%) or mean±standard deviation. Abbreviations as the same as in Table 1.

issues.

Twenty comorbidities were found in 14 patients (12.5%); liver dysfunction (7); lifestyle-related disease (diabetes mellitus (3), hypertension (2), renal dysfunction (2), hyperuricemia (2), and dyslipidemia (2)); sleep apnea syndrome (1) and insomnia (1). There was no significant difference between the two groups in terms of these comorbidities.

Thus, it was suggested that the composition of simple, moderate, and complex CHD was different between the two groups even with age-adjusted comparison.

Questionnaire Survey Information

1) Patients' Knowledge and Anxiety about Their Heart Diseases and Treatments

As shown in Table 3, diagnoses obtained from the medical records matched with those of patients' recognition of their own CHD in 109 patients (97.3%). The remaining 3 patients had misunderstood their heart diseases. Two of them had thought that they had atrial septal defect combined with ventricular septal defect, and the other one as having atrial septal defect combined with pulmonary valve stenosis.

All 112 patients had undergone intracardiac repair, and 109 of them (97.3%) correctly recognized the fact. All patients had regularly visited the cardiovascular outpatient clinic at the time of this study, but only 44 of them (39.3%) answered that they had been under regular follow-up. Twenty-eight patients (25.0%) answered that they took medication, whereas 41 patients (36.6%) were actually on medication according to the medical records. Age at recognition of their treatment (6.7±3.2

Table 3 Patients' knowledge and anxiety about the heart defects and its treatments

	Denominator	All patients		PDC (−)		PDC (+)	*p*-value
Heart defects							
Correctly answer the name of heart defects, n (%)	(*N* = 112)	109 (97.3)	(*n* = 59)	57 (96.6)	(*n* = 53)	52 (98.1)	1.000
Age at recognizing the diagnoses, years (range: 0–26)	(*N* = 105)	8.8 ± 4.3	(*n* = 57)	9.7 ± 4.0	(*n* = 48)	7.7 ± 4.3	0.008
Treatments							
Treatment contents (multiple answers)	(*N* = 112)		(*n* = 59)		(*n* = 53)		
Surgery, n (%)		109 (97.3)		58 (98.3)		51 (96.2)	1.000
Follow up, n (%)		44 (39.3)		23 (39.0)		21 (39.6)	1.000
Medication, n (%)		28 (25.0)		13 (22.0)		15 (28.3)	0.512
Unavailable, n (%)		1 (0.9)		0 (0.0)		1 (1.9)	—
Age at recognizing the treatments, years (range: 0–18)	(*N* = 102)	6.7 ± 3.2	(*n* = 55)	7.3 ± 3.0	(*n* = 47)	6.1 ± 3.5	0.038
Transition of outpatient clinic from pediatrics to adult cardiology	(*N* = 112)		(*n* = 59)		(*n* = 53)		
Positive response for transition, n (%)		97 (86.6)		51 (86.4)		46 (86.8)	0.783
Negative response for transition, n (%)		14 (12.5)		8 (13.6)		6 (11.3)	
Unavailable, n (%)		1 (0.9)		0 (0.0)		1 (1.9)	—
Anxiety regarding heart defects and its treatments	(*N* = 112)		(*n* = 59)		(*n* = 53)		
Worried, n (%)		79 (70.5)		41 (69.5)		38 (71.7)	0.838
Top 5 anxiety factors (multiple answers)	(*N* = 79)		(*N* = 41)		(*N* = 38)		
Worsening of medical condition, n (%)		43 (54.4)		21 (51.2)		22 (57.9)	0.653
Surgery, n (%)		40 (50.6)		18 (43.9)		22 (57.9)	0.263
Pregnancy/delivery, n (%)		35 (44.3)		20 (48.8)		15 (39.5)	0.498
Life span, n (%)		33 (41.8)		17 (41.5)		16 (42.1)	1.000
Marriage, n (%)		21 (26.6)		7 (17.1)		14 (36.8)	0.073
Age at beginning to feel anxiety, years (range: 4–42)	(*N* = 72)	16.2 ± 6.6	(*n* = 37)	16.1 ± 6.4	(*n* = 35)	16.1 ± 6.9	0.847
Existence of adviser	(*N* = 112)		(*n* = 59)		(*n* = 53)		
Yes, n (%)		100 (89.3)		52 (88.1)		48 (90.6)	0.766
Top 5 advisers (multiple answers)	(*N* = 100)		(*n* = 52)		(*n* = 48)		
Mother, n (%)		86 (86.0)		50 (96.2)		36 (75.0)	0.044
Father, n (%)		47 (47.0)		31 (59.6)		16 (33.3)	0.022
Spouse, n (%)		26 (26.0)		11 (21.2)		15 (31.3)	0.266
Friends, n (%)		17 (17.0)		8 (15.4)		9 (18.8)	0.793
Siblings, n (%)		17 (17.0)		6 (11.5)		11 (22.9)	0.186

Data are expressed as number of patients (%) or mean ± standard deviation. Abbreviations as the same as in Table 1.

years) was younger than that of their diagnosis (8.8 ± 4.3 years, $p < 0.001$).

Admitting that all patients had transitioned from pediatric to adult cardiology department, 14 patients (12.5%) had negative impression toward the new circumstance. The reasons for this were: "I don't know the doctor in charge well" in 7, "I don't like the atmosphere of adult cardiology department" in 6, "I can't explain my disease well to the physician" in 2, "the doctor in charge do not understand me" in 1, and "others" in 4.

As many as 79 patients (70.5%) were worried about their heart disease and its treatment. Thirteen patients wrote free descriptions regarding anxiety and requests related to their heart disease and daily life. Among them, the disease-related contents were; "a decline in physical strength" in 3, "meaning of having CHD" in 2, "desire to know their medical condition in detail" in 2, and "reoperation," "arrhythmia," "insomnia," and "request to have an opportunity to share information with people having similar heart problems" in one each. Regarding daily life, the issues mentioned were "restriction in physical exercise," "pregnancy and childbirth," and "lack of consideration for employees with disabilities" in 1 each. The other two were "strain of family" and "hospital expenses and a financial co-signer."

Age at recognition of their diagnosis was significantly younger in the certified group than in the non-certified group (7.7 ± 4.3 years vs. 9.7 ± 4.0 years, $p < 0.01$), so was that of their treatment (6.1 ± 3.5 years vs. 7.3 ± 3.0 years, $p < 0.05$). There was no significant difference between the two groups in terms of previous treatments, response to transitional care, or anxiety regarding their heart disease and its treatment. Respondents in the non-certified group more frequently expressed that the parents were their advisors, comparing to those in the certified group, with a statistical significance.

2) Social Independence

As shown in Table 4, employment status was investigated in 93 excluding 19 students. Of these 93, 77 patients (82.8%) were employed and 16 patients (17.2%)

Table 4 Social independence

	Denominator	All patients		PDC (−)		PDC (+)	*p*-value
Student, n (%)	(*N* = 112)	19 (17.0)		16 (27.1)		3 (5.7)	0.003
Employment states	(*N* = 93)		(*n* = 43)		(*n* = 50)		
Employed, n (%)		77 (82.8)		34 (79.1)		43 (86.0)	0.419
Unemployed, n (%)		16 (17.2)		9 (20.9)		7 (14.0)	
Employment status †	(*N* = 77)		(*n* = 34)		(*n* = 43)		
Regular employment, n (%)		51 (66.2)		23 (67.6)		28 (65.1)	0.816
Non-regular employment, n (%)		26 (33.8)		11 (32.4)		15 (34.9)	
Hours of work/week, hours	(*N* = 73)	36.0 ± 17.4	(*n* = 32)	37.4 ± 15.8	(*n* = 41)	34.9 ± 18.7	0.194
Type of works †	(*N* = 77)		(*n* = 34)		(*n* = 43)		
Professional /technical work, n (%)		26 (33.8)		17 (50.0)		9 (20.9)	0.007
Office work, n (%)		26 (33.8)		6 (17.6)		20 (46.5)	0.008
Sales work, n (%)		10 (13.0)		5 (14.7)		5 (11.6)	0.690
Business work, n (%)		6 (7.8)		5 (14.7)		1 (2.3)	0.044
Management, n (%)		4 (5.2)		0 (0.0)		4 (9.3)	0.068
Transportation/cleaning/packing, n (%)		2 (2.6)		0 (0.0)		2 (4.7)	0.203
Others, n (%)		3 (3.9)		1 (2.9)		2 (4.7)	0.700
Educational background †	(*N* = 77)		(*n* = 34)		(*n* = 43)		
<College, n (%)		47 (61.0)		21 (61.8)		26 (60.5)	0.908
≥College, n (%)		30 (39.0)		13 (38.2)		17 (39.5)	
Anxiety about employment or continuation of work	(*N* = 93)		(*n* = 43)		(*n* = 50)		
Worried, n (%)		56 (60.2)		24 (55.8)		32 (64.0)	0.525
Not worried, n (%)		37 (39.8)		19 (44.2)		18 (36.0)	
Top 5 anxiety factors (multiple answers)	(*N* = 56)		(*n* = 24)		(*n* = 32)		
Physical strength, n (%)		32 (57.1)		10 (41.7)		22 (68.8)	0.058
Understanding of people around for the disease, n (%)		23 (41.1)		5 (20.8)		18 (56.3)	0.013
Business content, n (%)		13 (23.2)		2 (8.3)		11 (34.4)	0.028
Working hours, n (%)		11 (19.6)		5 (20.8)		6 (18.8)	1.000
Relationships with superiors, n (%)		9 (16.1)		2 (8.3)		7 (21.9)	0.274
Existence of advisers	(*N* = 93)		(*n* = 43)		(*n* = 50)		
Yes, n (%)		66 (71.0)		30 (69.8)		36 (72.0)	1.000
No, n (%)		21 (22.6)		10 (23.3)		11 (22.0)	
Unavailable, n (%)		6 (6.5)		3 (7.0)		3 (6.0)	—
Lists of advisers	(*N* = 93)		(*n* = 43)		(*n* = 50)		
Relative, n (%)		25 (26.9)		10 (23.3)		15 (30.0)	0.612
Others, n (%)		30 (32.3)		13 (30.2)		17 (34.0)	0.807
Both relative and others, n (%)		10 (10.8)		6 (14.0)		4 (8.0)	0.492
Unavailable, n (%)		28 (30.1)		14 (32.6)		14 (28.0)	—
Resolution of the problems	(*N* = 93)		(*n* = 43)		(*n* = 50)		
Not sure, n (%)		36 (38.7)		16 (37.2)		20 (40.0)	1.000
Resolved, n (%)		30 (32.3)		16 (37.2)		14 (28.0)	0.809
Not resolved, n (%)		5 (5.4)		1 (2.3)		4 (8.0)	0.378
Unavailable, n (%)		22 (23.7)		10 (23.3)		12 (24.0)	—
Household states	(*N* = 112)		(*n* = 59)		(*n* = 53)		
Living with parents, siblings, and/or grandparents, n (%)		55 (49.1)		32 (54.2)		23 (43.4)	0.264
Living alone, n (%)		29 (25.9)		14 (23.7)		15 (28.3)	0.667
Living with spouse (partner) and/or offspring, n (%)		28 (25.0)		13 (22.0)		15 (28.3)	0.515
Anxiety and difficulty in student life and social life							
Having anxiety and difficulty, n (%)	(*N* = 112)	49 (43.8)	(*n* = 59)	21 (35.6)	(*n* = 53)	28 (52.8)	0.086
No anxiety and difficulty, n (%)		63 (56.3)		38 (64.4)		25 (47.2)	
Top 5 anxiety and difficulty factors (multiple answers)	(*N* = 49)		(*n* = 21)		(*n* = 28)		
Exercise or physical restriction, n (%)		34 (69.4)		17 (81.0)		17 (60.7)	0.128
School event, n (%)		25 (51.0)		9 (42.9)		16 (57.1)	0.322
Understanding of people around for the disease, n (%)		18 (36.7)		7 (33.3)		11 (39.3)	0.669
Education and Employment, n (%)		9 (18.4)		1 (4.8)		8 (28.6)	0.033
Compatibility of treatments and studies, n (%)		4 (8.2)		1 (4.8)		3 (10.7)	0.451
Existence of advisers	(*N* = 112)		(*n* = 59)		(*n* = 53)		
Yes, n (%)		82 (73.2)		42 (71.2)		40 (75.5)	0.821
No, n (%)		24 (21.4)		13 (22.0)		11 (20.8)	
Unavailable, n (%)		6 (5.4)		4 (6.8)		2 (3.8)	—
Top 5 advisers (multiple answers)	(*N* = 112)		(*n* = 59)		(*n* = 53)		
Mother, n (%)		71 (63.4)		37 (62.7)		34 (64.2)	1.000
Father, n (%)		29 (25.9)		17 (28.8)		12 (22.6)	0.514
Friends, n (%)		21 (18.8)		12 (20.3)		9 (17.0)	0.633
Doctor, n (%)		20 (17.9)		11 (18.6)		9 (17.0)	0.808
Teacher, n (%)		15 (13.4)		6 (10.2)		9 (17.0)	0.407
Unavailable, n (%)		6 (5.4)		4 (6.8)		2 (3.8)	—

†: Analyses of employment status and educational background were conducted in 77 patients under employments. Data are expressed as number of patients (%) or mean ± standard deviation. Abbreviations as the same as in Table 1.

unemployed. The main reasons for unemployment were poor physical conditions in 6, housewives in 4, pregnancy/child rearing in 2, and others in 4. There was no significant difference in gender in terms of the employment status. Of the 77 workers, 66% had regular employment. As for types of work, approximately 34% were in professional/technical work and another 34% in office works. Among the employed, 39% of them had a college/university or higher degree.

Of 93 non-student patients, 56 (60.2%) had anxiety about employment or continuation of work. The top 5 issues were; "physical strength" in 32 patients (57.1%), "understanding of their disease by people around" in 23 (44.1%), and "contents of work" in 13 (23.2%). Approximately 70% of patients had someone to talk to about their anxiety. Those people were their relatives (approximately 30%) or others (approximately 30%). Only 30 patients (32.3%) were able to relieve their anxiety.

About half of the patients lived with their family. Forty-nine patients (43.8%) had anxiety or difficulty in student life and/or social life due to their heart problems. Common factors of anxiety and difficulty were "restriction in physical exercise" in 34 of the 49 patients (69.4%); "distress in school events" in 25 (51.0%), "understanding of their disease by people around" in 18 (36.7%), and "uncertainty in education and employment" in 9 (18.4%). In addition, 82 patients (73.2%) had someone to talk to about these problems.

Students were significantly fewer in the certified group than in the non-certified group. The employment status was compared between the two groups (in 93 non-student patients, excluding 19 students). In the certified group, office work was commoner (46.5% vs. 17.6% in the non-certified group, $p<0.01$), professional/technical work less common (20.9% vs. 50.0%, $p<0.01$), and sales work less common as well (2.3% vs. 14.7%, $p<0.05$). There was no significant difference between the two groups in the educational backgrounds of the employees. Anxiety about "physical strength" was more frequently seen in the certified group (68.8% vs. 41.7% in the non-certified group, $p<0.10$). Concerns about "understanding of their disease by people around" and "contents of work" were significantly more frequent in the certified group (56.3% vs. 20.8% in the non-certified group, $p<0.05$, and 34.4% vs. 8.3% in the non-certified group, $p<0.05$, respectively). There was a tendency that those in the certified group feel anxiety and difficulty in student life and social life more frequently (52.8% vs. 35.6% in the non-certified group, $p<0.10$). Anxiety regarding "education and employment" was significantly more frequent in the certified group than in the other (28.6% vs. 4.8%, $p<0.05$).

3) Lifestyles

As shown in Table 5, history of drinking was present in 44 (42.3%) as an occasional drinker and in 5 (4.8%) as a habitual drinker out of 104 adult patients (aged 20 years or above, excluding 8 underage patients). Habitual drinkers were commoner among males than females (10.8% of all male patients vs. 1.5% of all female patients, $p<0.05$). There were seven daily smokers (6.7%) and another occasional smoker. Similarly to drinking, smoking was commoner among males than females (18.9% vs. 1.5%, $p<0.01$). The average interval was about 8 months for outpatient clinic, and nearly 36% of patients also had regular dental consultation. In terms of sleep, 6.3% of patients occasionally used sleeping pills, and the average sleep length was 6.6 hours. Fifty-three patients slept favorably within the recommended range of sleep length (18–64 years old; 7–9 hours),[11] while 56 slept below the favorable range and 3 above the range. Poor sleep satisfaction was observed in 27.7% of patients. Sleep latency more than 30 minutes was the case in 54.

Drinking alcohol was significantly more frequent in the certified group (10.0% vs. non-certified group 0.0%, $p<0.05$), while there was no significant difference between the two groups in terms of smoking. The interval between outpatient clinic visits was significantly shorter in the certified group (6.8 ± 4.6 months vs. 9.2 ± 4.7 months in the non-certified group, $p<0.01$). Regular dental consultation was significantly more often in the certified group (47.2% vs. 26.3%, $p<0.05$). There were no significant differences in sleep length and sleep satisfaction between the two groups, but use of sleeping pills tended to be more common in the certified group (11.3% vs. 1.7%, $p<0.10$).

Discussion

We examined the patients' knowledge and anxiety about their heart malformation and its treatment, social independence, and lifestyles in patients with repaired TOF. Analysis was carried out as a whole, as well as in respect of with or without the PDC.

Table 5 Lifestyles and sleeping habits

	Denominator	All patients		PDC (−)		PDC (+)	*p*-value
Lifestyles							
History of drinking in adult patients †, n (%)	(*N* = 104)	5 (4.8)	(*n* = 54)	0 (0.0)	(*n* = 50)	5 (10.0)	0.017
History of smoking in adult patients, n (%)	(*N* = 104)	8 (7.7)	(*n* = 54)	4 (7.4)	(*n* = 50)	4 (8.0)	0.910
Outpatient clinics visit intervals, months (range: 1–29)	(*N* = 112)	8.1 ± 4.8	(*n* = 59)	9.2 ± 4.7	(*n* = 53)	6.8 ± 4.6	0.006
Regular dental consultation, n (%)	(*N* = 110)	40 (36.4)	(*n* = 57)	15 (26.3)	(*n* = 53)	25 (47.2)	0.029
Consultation intervals, months (range: 1–12)	(*N* = 37)	4.8 ± 3.2	(*n* = 15)	5.4 ± 2.7	(*n* = 22)	4.5 ± 3.6	0.161
Sleeping habits	(*N* = 112)		(*n* = 59)		(*n* = 53)		
Taking a sleeping pill, n (%)		7 (6.3)		1 (1.7)		6 (11.3)	0.051
Sleep latency	(*N* = 112)		(*n* = 59)		(*n* = 53)		
Sleep latency, min (range: 3–120)		24.8 ± 19.8		24.1 ± 17.6		25.7 ± 22.1	0.974
≥30 min, n (%)		54 (48.2)		29 (53.7)		25 (47.2)	0.852
Sleep length	(*N* = 112)		(*n* = 59)		(*n* = 53)		
Sleep length, hours (range: 3–10)		6.6 ± 1.3		6.7 ± 1.1		6.5 ± 1.5	0.432
Normal ‡, n (%)		53 (47.3)		29 (49.2)		24 (45.3)	0.757
Short, n, (%)		56 (50.0)		29 (49.2)		27 (50.9)	
Long, n (%)		3 (2.7)		1 (1.7)		3 (2.7)	
Sleep satisfaction	(*N* = 112)		(*n* = 59)		(*n* = 53)		
Very good n (%)		11 (9.8)		7 (11.9)		4 (7.5)	0.899
Fairly good n (%)		70 (62.5)		36 (61.0)		34 (64.2)	
Fairly bad n (%)		29 (25.9)		15 (25.4)		14 (26.4)	
Very bad n (%)		2 (1.8)		1 (1.7)		1 (1.9)	

†: Only daily drinkers were considered to be drinkers, and occasional drinkers were analyzed as not drinking. ‡: The normal sleeping length was considered as 7–9 hours / day in those of 18–64 years. Data are expressed as number of patients (%) or mean ± standard deviation. *N* = 104 was only the patients with the ages ≥20 years. Abbreviations as the same as in Table 1.

Rationale for Grouping by Physical Disability Certificate Status

Whether the PDC had been qualified or not was used as a criterion for dividing the whole patients' cohort into two groups. We considered that this criterion represents not only the severity of cardiac defects but also factors affecting a financial issue and social life. The purposes of this study were firstly to examine the differences in patients' knowledge and anxiety about the heart problems and their treatment, social independence, and lifestyles, and secondly to obtain a basis for health guidance and education of the patients. While it is certain that the certificate status affects various aspects of social life such as medical expenses and employment, there was a need to confirm whether the certified group was a medically serious group. The simple descriptive statistics of both groups in this study showed that the medical indices of the certified group were more significant in many respects, but the certified group was significantly older and had a longer clinical history. We used age-adjusted propensity score matching to examine whether the significantly older age and longer medical history of the certified group confounded the progression of cardiac disease severity. The post matching data showed that the certified group had a severer NYHA cardiac functional class and more frequently cardiovascular events, suggesting that the certified group is a cohort of patients with severer cardiac disease.

Patient's Knowledge and Anxiety about the Heart Problems and Their Treatment

The percentages of correctly answering diagnosis of their own heart defects and recognizing their surgical history were reasonably high. On the other hand, their understanding of the follow-up plan and medication were different from the actual treatment situation. Compared with a previous study,[12] the proportion of correctly recognizing their diagnosis was higher in this study, which may be due to the fact that approximately 60% of the patients had transitioned to the cardiology outpatient clinic after undergoing patient education for transitional support at their previous pediatric institutions. This suggests that education during the transition period would be effective. It could also be the case that the answers presented in a multiple-choice format might have facilitated correct recognition of disease names in the questionnaire. The fact that age at recognizing treatments was younger than age at recognizing diagnosis probably reflects the impact and memory of having experienced invasive treatments such as surgery. Understanding and remembering their heart diagnosis require intellectual development.

Although all the patients had transitioned to adult cardiology clinic, 13% of them had a negative impression

toward their transition. The reason for this was mostly anxiety about the change in therapeutic surroundings. In the transitional care, it is necessary to educate and support patients, not only to establish a system that enables a smooth transition, so that they themselves can become independent and take the initiative of continuing medical care and resolve their anxiety.[13] If the transition does not go smoothly, patients might drop out of medical treatment, their quality of life would deteriorate, and appropriate self-management could become difficult. It is considered necessary to clarify the factors that hinder patients' independence and to support them in shifting the responsibility for medical care to themselves.

Anxiety related to heart diseases and their treatment was considerable in both groups. Anxiety about marriage appeared to be more common in the certified group. It was presumed that patients in the certified group had more concern about marriage which was obviously a big life event, because their cardiac disease tended to be severer. In a study of young ACHD patients, anxiety about future health, operative scars, and the need for future reoperation caused stress in about half of the patients.[14] Psychological problems often occurred against the background of such long-term stress, and depression and anxiety have been reported in patients with CHD.[15] These psychological problems not only affect the treatment of cardiac disease and social adjustments but may also impact on their prognosis.[16] It is desirable to quantitatively assess the psychological problems of ACHD patients using established screening scales and to appropriately correspond to them. The age at which patients begin to feel anxiety also coincides with their transition from pediatric to adult cardiology, and pre-transitional education should be promoted so that transitional care is performed smoothly and trustful relationships can be established promptly between adult cardiologists and patients.

Social Independence

According to overseas studies, the employment rate of ACHD patients was 59–66%, the non-employment rate 10–18%, and those with a college/university or higher degree was reported as 19–46%.[17–19] Compared with the national standard value of regular employment rate 81% for healthy men in Japan,[20] the regular employment rate of male patients with TOF in the present study was as low as 66%, while that of female TOF patients was equivalent to that of healthy women. In a previous study of ACHD patients with the PDC, the employment rate was reported as 41%; probably, ACHD patients with various degrees of severity of disease were included.[21] As shown in this study, patients with TOF had a relatively high employment rate and regular employment rate compared with those with other severe CHD. Still, these rates were lower than similar figures in their peers without heart disease. The employment rate is affected by the socio-economic status in each country, family circumstance, and severity of cardiac disease. In general, regular employment rate is high in men, and a high non-regular employment rate in women in most countries.[22] It should be noted that regular employment rate is low among male patients with TOF.

In addition, profession/technical work and sales work were significantly less common in the certified group. Instead, office work was more frequent. It is suggested that patients with no PDC have a relatively mild form of heart disease and that they can choose their type of work from a broader range. On the other hand, those with more complex CHD would choose office work posing less physical burden.

Lifestyles

A previous study in ACHD patients with a mean age of 30 years showed that the drinking rate was 22% and the smoking rate 18%.[23] In 2018 in our country, 15% of men and 9% of women in the general population drank alcohol at the level of amount that would increase the risk of lifestyle-related diseases. The smoking rate was 29% among men and 8% among women.[24] In comparison with these previous figures, habitual drinking and smoking were low in patients of the present study. Attention should be paid to slightly high prevalence of habitual drinking in the certified group; particularly in terms of lifestyle-related diseases. Not a few patients had difficulty in getting to sleep, and patients in the certified group tended to take more sleeping pills. These may accord with higher alcohol consumption in the certified group, since a traditional and popular behavior of Japanese people to cope with insomnia had been "drinking alcohol."[25] Although depression and anxiety were not accurately assessed in this study, it was possible that patients in the certified group had more habitual drinking driven by strong anxiety related to impaired physical strength, poor understanding of their disease, contents

of work, and uncertainty in education and employment. It is also an enormous stress to face constantly with chronic diseases.

Because of improving life prognosis in ACHD patients, the prevalences of cardiovascular risk factors have increased such as hypertension, diabetes mellitus, obesity, chronic renal disease, and peripheral arterial disease.[26] It is a major emerging agenda to encourage and support patients to promote behavior modification against drinking and smoking, paying attention to cardiac complications in the future caused by lifestyles. Worsening of underlying cardiac problems and cardiovascular events should be prevented.

Furthermore, one of the reasons why regular dental consultation was frequent on top of regular visit to cardiovascular outpatient clinic in the certified group may be related to the system by which their individual payment for medical expense is limited relatively small. Among cardiovascular events, infective endocarditis could occur as a result of bacteremia caused by poor oral hygiene.[27] In postoperative TOF patients, the incidence of endocarditis was reported as 1.3% during a long-term observation period of 30 years.[28] Although the incidence is not markedly high, both morbidity and mortality are considerably high once endocarditis occurs. Therefore, regular dental consultation should be recommended.

There are no reports on sleep disorders in patients with CHD. In those with chronic heart failure, it has been reported that 76% of patients experienced sleep-disordered breathing.[29] Difficulty in getting to sleep increases the risk of developing hypertension by 1.96 folds, and nocturnal awakening does by 1.88 folds.[30] The patients in this study were young and hypertension was uncommon. Taking sleeping pills were commoner in the certified group. This group of patients tended to have more complex cardiac disease, impaired physical strength, poor understanding of their disease, and anxiety regarding contents of work, education, and employment. Their relying on sleeping pills might be related to these issues. Further investigation is needed for sleep disorders by means of quantitative methods.

Study Limitations and Outlook for the Future

The PDC is a multifactorial qualification. Whether a patient has this certificate approved or not was used as a scale for analysis. This is one of the limitations of the current study. A simpler index such as classification according to CHD severity or gender could have been chosen from a medical perspective. By using the PDC status for dividing the cohort into two, socio-economic factors were compounded with other medical aspects. This made it impossible to discuss simple causal relationship. Moreover, this study included a few patients who did have a severer form of CHD but did not have the PDC approved. Even with such limitations, we have gained a new point of view on the actual conditions of medical care in patients with CHD. The PDC facilitated access to medical care because it reduced the burden of medical expenses, and the outpatient consultation rate of ACHD patients, either medical or dental, was likely influenced by whether this certificate had been approved or not.

Another limitation was related to selection of patients; those with a severer form of disease, such as pulmonary atresia with ventricular septal defect and intellectual disability, were excluded. Namely, the results of the present study are based on TOF patients with relatively stable cardiac function. It should be noted also that the majority of them underwent support education programs for transition at outpatient clinics. In addition, the fact that this study was conducted at a single center cannot be ignored. The results of survey on social independence and lifestyles may have been affected by the greater proportion of students in the non-certified group. It is necessary to enroll more participants for further analysis. The groups should be examined with age and proportion of students matched. As for lifestyles, dietary habits and physical exercise have not been investigated in detail. Future study should be conducted on the factors that disturb social independence of TOF patients, particularly in terms of lifestyle. It is necessary to consider physical disability, psychological problems, and social issues from multiple perspective and to seek for specific support measures.

Overview and Conclusions

We hypothesized that whether the PDC was qualified in a patient with CHD or not should be associated with significance of her/his heart disease and would affect the financial aspect (e.g. medical expenses) and the social aspect (e.g. employment). We examined the clinical characteristics, anxiety, social independence, and life-

style in TOF patients with and without the PDC.

There was no significant difference between the two groups in terms of intensity or composition of anxiety about their heart disease. They began to get worried about their conditions during the transitional period. No significant difference between the two groups in the employment rate and regular employment rate, whereas there were more office workers in the certified group. No significant difference between the two groups in the proportion of patients who felt anxiety about their work. In contrast, patients in the certified group felt anxiety more about marriage, physical fitness, contents of work, and understanding of their disease by people around. In addition, those in the certified group visited medical facilities including dentistry regularly, whereas they tended to drink alcohol and/or take sleeping pills. We presume that this is a response to cope with their anxiety. Those in the non-certified group had less frequent access to medical facilities. They rely on parents as advisors to resolve anxiety about their heart problems; this is an intense role for the parents and indicates a strong dependence on family.

Smooth transition to adult cardiology outpatient clinic is essential to reduce anxiety of adult TOF patients. Multidisciplinary/multiprofessional collaboration is also necessary among nurses, clinical psychologists, and medical doctors. Such collaboration would enable continuous lifestyle guidance providing mental and physical support and inducing behavior change. In order to provide an appropriate work environment for patients with cardiac disease, it is necessary to establish a system in which healthcare and employment support are more closely coordinated. The employment system for persons with disabilities and employment support centers can be utilized, based on precise information from the healthcare provider. For those in the non-certified group, social intervention and support by third-party organization were also considered necessary to reduce the burden on the family. It is mandatory to establish a society that accepts people with heart malformation without discrimination.

Acknowledgment

The authors are deeply grateful to the subjects of this study who participated in this questionnaire survey and to the doctors of the Department of Cardiology and the Department of Pediatrics of Kyushu University Hospital. This study was supported by Grant-in-Aid for Scientific Research B; #18H03083, and grants 15ek0109123h0001, 16ek0109123h0002, and 17ek0109123h0003 from the Japan Agency for Medical Care and Research.

Conflicts of Interest

The authors declare no conflict of interest.

Author Contribution

R.S. and H.S. did statical analysis, R.S. and A.C. drafted the study protocol, conceived the paper, interpreted the data, and wrote the paper. K.Y., M.K., I.S., K.Y., H.N., H.T., H.C., and T.T. were involved in data interpretation and critical revision of the paper. All authors were involved in the final approval of the manuscript and the decision to publish the study results.

References

1) Moons P, Bovijn L, Budts W, et al: Temporal trends in survival to adulthood among patients born with congenital heart disease from 1970 to 1992 in Belgium. Circulation 2010; **122**: 2264–2272
2) Akagi T, Ikeda T, Ichikawa H, et al: Guidelines for Management of Congenital Heart Diseases in Adults (JCS 2017). Available from http://www.j-circ.or.jp/guideline/pdf/JCS2017_ichida_h.pdf [accessed June 12, 2021]. (in Japanese)
3) Baumgartner H, Bonhoeffer P, De Groot NM, et al: ESC Committee for Practice Guidelines (CPG): The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC): ESC guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J 2010; **31**: 2915–2957
4) Akiyama N, Ochiai R, Higaki T, et al: Characteristics of adults with congenital heart disease living apart from parents. Nihon Shoni Junkanki Gakkai Zasshi 2020; **36**: 313–320 (in Japanese)
5) Niwa K, Tateno S, Tatebe S, et al: Education and insurability of adults with congenital heart disease. Pediatr Cardiol Card Surg 2003; **19**: 69–71 (in Japanese)
6) Ochiai R, Ikeda Y, Kato H, et al: Relationship between social independence and psychological profiles in adult congenital heart disease patients with Person's certificate. Pediatr Cardiol Card Surg 2012; **28**: 258–265 (in Japanese)
7) Niwa K, Tateno S, Tatebe S, et al: Social concern and independence in adults with congenital heart disease. J Cardiol 2002; **39**: 259–266 (in Japanese)
8) Akagi T, Hidaka Y, Himeno W, et al: Current status and problems of social independence in adult patients with congenital heart disease: Factors inhibiting independence and differences between men and women in marriage and parenthood. Pediatr Cardiol Card Surg 2003; **19**: 72–74 (in Japanese)
9) Ichida F: Epidemiology of congenital heart disease. Pedi-

atr Cardiol Card Surg 2010; **26**: 2–3 (in Japanese)
10) Likert R: A technique for the measurement of attitudes. Arch Psychol 1932; **22**: 1–55
11) Hirshkowitz M, Whiton K, Albert SM, et al: National Sleep Foundation's sleep time duration recommendations: Methodology and results summary. Sleep Health 2015; **1**: 40–43
12) Moons P, Van Deyk K, Marquet K, et al: Individual quality of life in adults with congenital heart disease: A paradigm shift. Eur Heart J 2005; **26**: 298–307
13) van Rijen EH, Utens EM, Roos-Hesselink JW, et al: Psychosocial functioning of the adult with congenital heart disease: A 20–33 years follow-up. Eur Heart J 2003; **24**: 673–683
14) Zomer AC, Vaartjes I, Uiterwaal CS, et al: Social burden and lifestyle in adults with congenital heart disease. Am J Cardiol 2012; **109**: 1657–1663
15) Ochiai R, Ikeda Y, Kato H, et al: Parents' Association of Heart Disease Children: Social independence of adult congenital heart disease patients in Japan. Pediatr Int 2017; **59**: 675–681
16) Fernandes SM, Verstappen A, Clair A, et al: Adult Congenital Heart Association and the Adult Congenital Cardiac Care Associate Research Group: Knowledge of life-long cardiac care by adolescents and young adults with congenital heart disease. Pediatr Cardiol 2019; **40**: 1439–1444
17) Yokoya S, Ochiai R, Kobayashi N, et al: Recommendations on transitional care for patients with childhood onset diseases. Pediatr Cardiol Card Surg 2014; **118**: 98–106
18) Jackson JL, Gerardo GM, Daniels CJ, et al: Perceptions of disease-related stress: A key to better understanding patient-reported outcomes among survivors of congenital heart disease. J Cardiovasc Nurs 2017; **32**: 587–593
19) Kovacs AH, Saidi AS, Kuhl EA, et al: Depression and anxiety in adult congenital heart disease: Predictors and prevalence. Int J Cardiol 2009; **137**: 158–164
20) Kourkoveli P, Rammos S, Parissis J, et al: Depressive symptoms in patients with congenital heart disease: Incidence and prognostic value of self-rating depression scales. Congenit Heart Dis 2015; **10**: 240–247
21) Statistics Bureau: Ministry of Internal Affairs and Communications: Basic results of the 2015 National Census of Employment Status. http://www.stat.go.jp/data/kokusei/2015/kekka.html [accessed June 12, 2021] (in Japanese)
22) Organisation for Economic Co-Operation and Development: OECD Employment Outlook 2017. OECD Publishing, 2017
23) Caruana M, Grech V: Lifestyle habits among adult congenital heart disease patients in Malta. Congenit Heart Dis 2016; **11**: 332–340
24) Statistics Bureau: Ministry of Health, Labour and Welfare: Overview of the results of the National Health and Nutrition Survey. https://www.mhlw.go.jp/content/10900000/000584138.pdf [accessed June 12, 2021] (in Japanese)
25) Uchiyama M: Characteristics of sleep in the Japanese: Based on the results of the International Sleep Epidemiology Study. J Clin Exper Med 2003; **205**: 529–532 (in Japanese)
26) Agarwal S, Sud K, Menon V: Nationwide hospitalization trends in adult congenital heart disease across 2003-2012. J Am Heart Assoc 2016; **5**: e002330
27) Nakatani S, Mitsutake K, Hozumi T, et al: Committee on Guideline for Prevention and Management of Infective Endocarditis, Japanese Circulation Society: Current characteristics of infective endocarditis in Japan: An analysis of 848 cases in 2000 and 2001. Circ J 2003; **67**: 901–905
28) Morris CD, Reller MD, Menashe VD: Thirty-year incidence of infective endocarditis after surgery for congenital heart defect. JAMA 1998; **279**: 599–603
29) Oldenburg O, Lamp B, Faber L, et al: Sleep-disordered breathing in patients with symptomatic heart failure: A contemporary study of prevalence in and characteristics of 700 patients. Eur J Heart Fail 2007; **9**: 251–257
30) Suka M, Yoshida K, Sugimori H: Persistent insomnia is a predictor of hypertension in Japanese male workers. J Occup Health 2003; **45**: 344–350