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Research Article

Clinical features and risk assessment for cardiac surgery in adult congenital heart disease: Three years at a single Japanese center

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Adult congenital heart disease (ACHD);
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Abstract *Purpose:* The aims of our study are twofold: first, to retrospectively identify the demographic characteristics and outcomes in cardiac surgery for adult congenital heart disease (ACHD); second, to explore whether certain preoperative examinations are useful for assessing the risk of perioperative mortality and morbidity.
Methods: Ninety-two ACHD patients who underwent cardiac surgery from 2009 to 2011 were enrolled in the study. The subjects were classified into three groups based on the complexity of the ACHD. We retrospectively collected data on demographics, operations, and postoperative courses. We also collected the results of examinations performed in the three months leading up to the cardiac surgery, including exercise tolerance testing and measurement of brain natriuretic peptide (BNP).
Results: The 30-day mortality was 3.3%. A remarkable discrepancy was found between subjective assessment and the severity of exercise intolerance by exercise tolerance testing. The NYHA class was 1 or 2 in all but one of 13 patients with moderate-severe exercise intolerance and a high mortality/major complication rate (53.8%). Patients with BNP $\geqq$ 100 pg/ml had a significantly higher mortality/major complication rate than patients with BNP < 100 (34.8% vs. 11.5%, $p < 0.05$), but the sensitivity (53.3%) and positive predictive value (34.8%) were not high enough in themselves to identify patients at high risk of poor outcome.

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Conclusion: Cardiac surgery could be safely performed in most ACHD cases. Exercise tolerance testing can be useful in identifying patients at high risk of mortality or major complications. BNP can be valuable in predicting poor outcomes after cardiac surgery.

1. Introduction

The population of patients with adult congenital heart disease (ACHD) in Japan has skyrocketed over the last four decades [1]. The number of ACHD patients surpassed the number of pediatric CHD patients more than a decade ago, in 1997, and today it surpasses 400,000 [1]. The distribution of ACHD complexity has also dramatically changed over the same period [1]. A Task Force of the American College of Cardiology (ACC) categorized ACHD in three types, simple, moderate, or complex [2]. Nowadays, cases with moderate to complex ACHD make up one-third of all ACHD cases in Japan [1]. The same trends in the number of ACHD patients and distribution of ACHD complexity are taking place in other developed countries, as well [3–7]. For these reasons, anesthesiologists will be expected to provide anesthetic management for a growing number of ACHD patients, especially patients with moderate to complex CHD, who undergo cardiac surgery, non-cardiac surgery, and delivery.

Meanwhile, care facilities for ACHD in Japan are still far from ideally established. A nationwide survey revealed that half of Japan's training hospitals for board-certified cardiologists perform cardiac surgery, but only on small numbers of patients [8]. The hospitals that performed the surgery generally do so on less than 10 patients per year, and only 6 of them nationwide have conducted more than 20 cardiac surgeries per year [8]. This naturally limits the opportunities for training in the field for anesthesiologists. As a first step to provide better training, we should recognize the demographic characteristics and outcomes of cardiac surgery for ACHD and gather sufficient knowledge on risk assessment for perioperative mortality and morbidity. Our hospital has conducted an average of 30–40 cardiac surgeries per year, which makes it one of Japan's most-experienced hospitals in cardiac surgery for ACHD. As such, knowledge of the demographic characteristics and outcomes of cardiac surgery for ACHD at our hospital may be useful for recognizing the same characteristics and outcomes on a larger scale, for all of Japan or other developed countries. The aims of this study are twofold; first, to retrospectively identify the demographic characteristics and outcomes in cardiac surgery for ACHD performed at our hospital over the three-year period; second, to explore whether certain preoperative examinations are useful for assessing the risk of perioperative mortality and morbidity.

2. Methods

2.1. Patients

The study subjects were consecutive CHD patients who underwent cardiac surgery under cardiopulmonary bypass (CPB) at the age of 15 years or older at Tokyo Women's Medical University Hospital over the period from January 1, 2009 to December 31, 2011. Patients undergoing aortic valve replacement for bicuspid aortic valve lesion were excluded from this study. The patients were classified into three groups based on the complexity of the CHD according to the definition established by the ACC Task Force. This study was approved by the institutional ethics committee with a waiver of the requirement to obtain written informed consent from patients older than 20 years old and from the parents of younger patients.

2.2. Data collection

Medical charts, anesthetic records, and operative records were retrospectively reviewed. Data on demographics (age, gender, height, body weight, diagnosis, history of previous operations, history of previous sternotomies, type of surgery, and type of procedure), preoperative status (New York Heart Association [NYHA] class and American Society of Anesthesiologists physical status [ASA-PS]), operation (operation time, CPB time, aortic cross-clamp [Ao-clamp] time, and the amount of bleeding), and postoperative course (postoperative ventilation time, length of ICU stay, length of hospitalization, mortality, and morbidity) were collected. Morbidity consists of major and minor complications. The major complications consist of mechanical ventilatory support for longer than 7 days, mechanical circulatory support, lethal ventricular arrhythmia, bleeding requiring surgical hemostasis, cardiac tamponade requiring drainage, permanent neurological deficits, and heart blocks requiring permanent pacemaker implantation. Minor complications included temporal neurological deficits, atrial arrhythmia, pleural effusion, and pneumothorax.

2.3. Preoperative examinations

The results of the following examinations were collected if the examinations were performed in the three months leading up to cardiac surgery: exercise tolerance testing (cardiopulmonary exercise testing [CPET] and 6-minute walk test [6MWT]), myocardial perfusion imaging (MPI), and measurement of brain natriuretic peptide (BNP), hemoglobin (Hb), and glomerular filtration rate (GFR).

2.4. Statistics

Data are shown as means plus-minus standard deviations or medians with ranges (minimum to maximum). The chi-square test and Mann–Whitney U test were used for comparisons between two groups and one-way ANOVA was used for comparisons among three groups. A p value of less than 0.05 was considered as statistical significance.

3. Results

Ninety-two cases underwent cardiac surgery for ACHD during the study period: 31 simple cases, 33 moderate cases, and 28 complex cases. Table 1 summarizes the demographic data of

Table 1 Complexity of CHD and type of surgery.

	Simple	Moderate	Complex	Total
No. of patients	31	33	28	92
Age (years)	40.6 ± 15.3	42.0 ± 18.1	24.8 ± 5.1[a]	36.3 ± 16.1
Body weight (kg)	57.9 ± 11.3	59.2 ± 14.4	55.2 ± 11.5	57.6 ± 12.5
Gender (male:female)	14:17	21:12	15:13	50:42
Correction	29	21	2	52 (56.5%)
Reoperation	2	12	25	39 (42.4%)
Palliation	0	0	1	1 (1.1%)

Data on age and body weight are shown as mean ± standard deviation.
[a] $p < 0.05$, vs. simple or moderate.

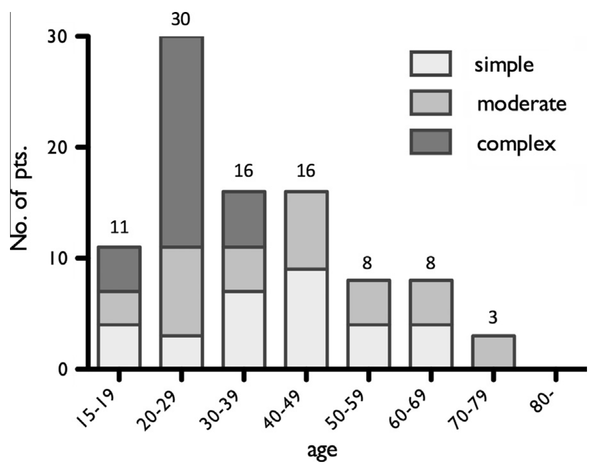

Figure 1 Distribution of age at surgery. Age at the time of surgery peaked in the patients' twenties. Most of the younger patients had moderate to severe CHD requiring reoperations following corrective surgery. Most of the elderly patients underwent corrections for simple lesions like ASD.

the three groups. The patients in the complex group were significantly younger than the patients in the other groups. Body weight and gender were similar among the three groups.

3.1. Data on demographics

Age at the time of surgery was 36.3 ± 16.1 years on average and a median of 31 years (Table 1). Age distribution peaked in the patients' twenties (Fig. 1). Forty-two (45.7%) patients had a history of one or more previous operations and 3 patients had received as many as four previous operations, the maximum number received. Forty-one (44.6%) patients had a history of one or more previous sternotomies and 2 patients had received three sternotomies, the maximum number received. Thirty-nine of the surgeries were reoperations, 52 were corrective surgeries, and 1 was a bidirectional Glenn operation for palliation (Table 1). Correction for left-to-right (L–R) shunt was the most frequent procedure, accounting for roughly half of the cases (Table 2). The next most frequent, in descending order, were valve operations, Fontan revisions, and operations for right ventricular outflow tract (RVOT) lesions (Table 2).

3.2. Preoperative status

No preoperative NYHA class was identified in the patients' medical charts in 11 cases. Nine out of 81 cases (11.1%) were classified as NYHA class 3 or greater. Most of the cases (69.6%) were classified as ASA-PS 3.

3.3. Amount of bleeding

Bleeding was significantly greater in the complex group than in the simple group or moderate group (969 ± 770 ml in the complex group vs. 324 ± 198 ml and 578 ± 543 ml in the simple group and moderate group, respectively, $p < 0.05$). Patients with a previous history of one or more median sternotomies also had significantly greater bleeding than cases with no previous history of median sternotomy (826 ± 590 ml and 1183 ± 1082 ml in cases with one previous sternotomy and cases with two or more previous sternotomies, respectively, vs. 358 ± 230 ml in cases with no previous sternotomy, $p < 0.05$).

3.4. CPB time, Ao-clamp time, and operation time

CPB time was significantly longer in complex CHD than in simple CHD (Table 3). Fifteen cases were excluded from the analysis on Ao-clamp time because they received no Ao-clamps. The Ao-clamp time was significantly longer in both moderate and complex CHD than in simple CHD (Table 3).

Table 2 Type of procedure.

	References	*n*	L–R shunt lesion (%)	Valve operation (%)	RVOTR (%)	Fontan revision (%)
TWMU	This study	92	46.7	22.8	7.6	14.1
Europe	[9]	2012	42.8	3.3	11.7	2.0
Netherlands	[11]	830	26.5	23.8	12.6	2.4
USA	[10]	719	5.2	8.2	19.1	5.1
UK	[4]	474	15.0	–	42.0	1.0

Table 3 CPB time, Ao-clamp time, and operation time.

	Simple	Moderate	Complex
Operation (min)	280.5 (140–420)	342 (222–696)[b]	612 (299–1392)[a]
CPB time (min)	85.5 (43–151)	140 (50–461)	193 (56–1028)[b]
Ao-clamp (min)	37 (20–106)	88 (37–312)[b]	131 (53–194)[b]

Data on times are shown as medians and ranges.
[a] $p < 0.05$, vs. simple or moderate.
[b] $p < 0.05$, vs. simple.

Table 4 Duration of postoperative ventilation, length of ICU stay, and length of hospitalization.

	Simple	Moderate	Complex
Ventilation (h)	4 (1–17)	6 (1–872)	18 (3–338)
ICU stay (days)	2 (1–8)	4 (1–51)	8 (1–23)[a]
Hospitalization (days)	19 (12–64)	22 (16–230)	38.5 (8–87)[a]

Data on durations are shown as medians and ranges.
[a] $p < 0.05$, vs. simple.

The operation time increased significantly in a stepwise manner as the grade of complexity of CHD advanced (Table 3).

3.5. Duration of postoperative ventilatory support, length of ICU stay, and length of hospitalization

Two patients with complex ACHD who died shortly after the operation were excluded from the analysis on these parameters. The duration of postoperative ventilatory support was not significantly different among the groups (Table 4). The length of ICU stay and length of hospitalization were significantly longer in complex CHD than in simple CHD (Table 4).

3.6. Mortality and morbidity

The 30-day mortality was 3.3% and the overall mortality was 5.4%. Forty-two complications, 23 of them major and 19 of them minor, were detected in 33 patients (35.9%). The most frequently observed complications were atrial tachyarrhythmia ($n = 9$), respiratory failure requiring mechanical ventilatory support ($n = 6$), low cardiac output requiring mechanical circulatory support (ECMO or IABP; $n = 4$), and acute renal failure requiring hemodialysis ($n = 4$). The complication rate was significantly higher in moderate or complex CHD than in simple CHD (39.4% and 50.0% in moderate and complex CHD, respectively, vs. 19.4% in simple CHD, $p < 0.05$).

3.7. Preoperative examinations

Eighteen cases underwent exercise tolerance testing, 5 by CPET and 13 by 6MWT. In the CPET, exercise tolerance was graded as follows based on the peak oxygen consumption (VO_2) or the slope of the regression line (VE/VCO_2) between minute ventilation (VE) and carbon dioxide production (VCO_2): normal ($VO_2 > 25$ ml/kg/min, or $VE/VCO_2 < 30$), mild intolerance ($20 < VO_2 \leqq 25$ ml/kg/min, or $30 \leqq VE/VCO_2 < 40$), moderate intolerance ($15 < VO_2 \leqq 20$ ml/kg/min, or $40 \leqq VE/VCO_2 < 50$), or severe intolerance ($VO_2 \leqq 15$ ml/kg/min, or $VE/VCO_2 \geqq 50$). In the 6MWT, exercise tolerance was graded as follows based on the distance or maximum decrease in oxygen saturation (SpO_2): normal (distance $\geqq$ 600 m), mild intolerance (450 $\leqq$ distance $<$ 600 m), moderate intolerance (300 $\leqq$ distance $<$ 450 m, or maximum decrease of 5–10%), or severe intolerance (distance $<$ 300 m, or maximum decrease of $\geqq$ 10%). A remarkable discrepancy was found between the patients' subjective assessment (NYHA class) and the severity of exercise intolerance by the CPET or 6MWT (Fig. 2). The NYHA class was 1 or 2 in all but one of 13 patients with moderate-severe exercise intolerance and a high mortality/major complication rate (53.8%), whereas 5 patients graded with normal or mild intolerance had a major complication rate of 20.0% without any mortality.

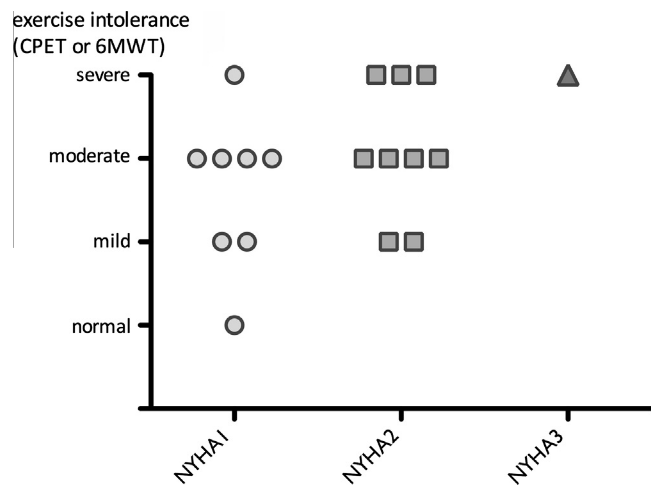

Figure 2 Relationship between NYHA class and results of exercise tolerance testing. An obvious discrepancy was observed between severity in exercise tolerance testing and NYHA class in our series. Even asymptomatic or mild symptomatic patients had moderate to severe exercise intolerance.

Myocardial perfusion was evaluated in 18 cases. Myocardial damage was detected in 8 patients with complex CHD (Table 5A). Of these 8, 7 patients had either a history of coronary transplantation or a pathophysiology of systemic right ventricle (RV), tetralogy of Fallot (TOF), or single ventricle (SV) (Table 5B). Four of the 8 patients with myocardial damage had minor complications, but none died or suffered major complications (Table 5B).

BNP was measured in 84 cases. NYHA class 3 patients had significantly higher BNP than NYHA class 1 or 2 patients (Fig. 3). Patients with BNP equal to or greater than 100 had a significantly higher mortality/major complication rate than patients with BNP less than 100, but the sensitivity (53.3%)

Table 5 Myocardial perfusion imaging: detection of myocardial damage and the backgrounds of patients with myocardial damage.

A. detection of damage

Complexity	Myocardial damage –	Myocardial damage +
Simple	2	0
Moderate	4	0
Complex	4	8

B. backgrounds of patients with myocardial damage

Case	Age	Operation	Previous operation or diagnosis	Mortality Morbidity
1	19	AVR	Post-Jatene	No
2	21	PA angioplasty	Post-Jatene	No
3	23	TCPC conversion	APC for TA	No
4	26	TVR	Post-Senning	Minor
5	26	SAS resection	Post-DSO	Minor
6	32	TVR	c-TGA	No
7	34	TCPC conversion	APC for SLV	Minor
8	37	Re-Rastelli	Post-Rastelli	Minor

AVR, aortic valve replacement; PA, pulmonary artery; TCPC, total cavo-pulmonary connection; APC, atrio-pulmonary connection; TA, tricuspid atresia; TVR, tricuspid valve replacement; SAS, subaortic stenosis; DSO, double switch operation; c-TGA, corrected transposition of the great arteries; SLV, single left ventricle.

and positive predictive value (34.8%) of high BNP were still too low to reliably identify patients at high risk of poor outcome after cardiac surgery for ACHD (34.8% in patients with BNP ≧ 100 pg/ml vs. 11.5% in patients with BNP < 100 pg/ml, $p < 0.05$).

Hb values were measured in all 92 cases. Hb was significantly greater in complex CHD than in simple or moderate CHD (simple CHD 13.5 ± 1.9 g/dl, moderate CHD 13.8 ± 1.6 g/dl vs. complex CHD 15.9 ± 3.0 g/dl, $p < 0.05$). The mortality/major complication rate did not significantly differ among the groups graded as anemia (Hb < 13 g/dl, 8.0%), normocythemia (13 ≦ Hb < 18 g/dl, 20.7%), and polycythemia (Hb ≧ 18 g/dl, 42.9%).

GFR calculated in 68 cases did not differ among the three groups. The mortality/major complication rate did not significantly differ among the groups graded as normal renal function (GFR ≧ 90 ml/min/1.73 m^2, 22.6%), mildly impaired renal function (60 ≦ GFR < 90 ml/min/1.73 m^2, 13.3%), and moderately impaired renal function (30 ≦ GFR < 60 ml/min/1.73 m^2, 16.7%).

4. Discussion

4.1. Data on demographics

Age at the time of surgery at our institute was similar to those in other reports from developed countries [4,9–11]. The relatively early age distribution peak, in the patients' twenties, must be attributable to the improved survival rates after surgery for complex defects. Most of the patients at the age distribution peak required reoperations for sequelae or residues following corrective surgery they underwent in childhood. In contrast, elderly patients primarily underwent closures for atrial septal defects (ASDs) or other L–R shunt lesions detected in adulthood.

The prevalence of reoperations, number of previous operations, and number of previous sternotomies have varied from study to study [4,6,9,11,12]. More experienced institutes were likely to treat patients who had undergone multiple previous operations or sternotomies. About one-third of the patients reported from the Mayo Clinic underwent three or more reoperations, and early mortality for reoperation rose to an extremely high level in patients who had undergone five or more sternotomies [13]. Only 11 of our patients (12% of all cases) underwent three or more reoperations. None of our patients received as many as five sternotomies, although 44.6% (41 cases) actually required re-sternotomy. Hence, our patients underwent markedly fewer reoperations and re-sternotomies than the patients at the Mayo Clinic.

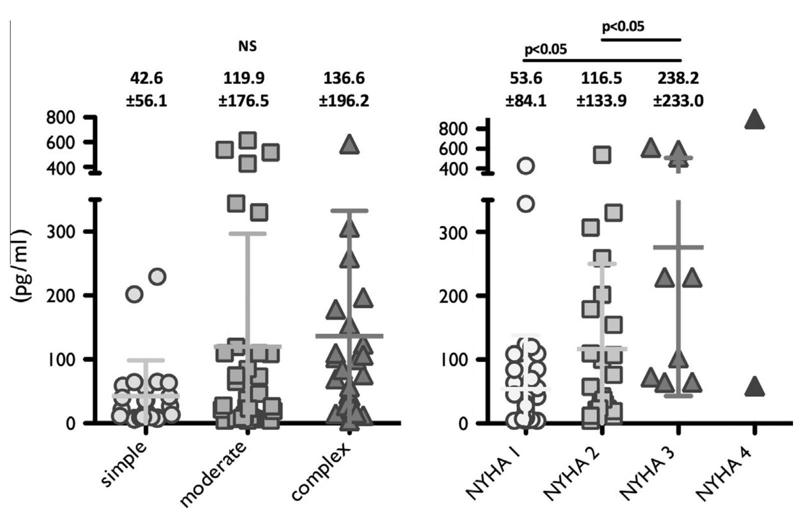

Figure 3 Distribution of preoperative BNP. BNP levels did not differ among the three groups classified based on the complexity of CHD. NYHA class 3 patients had significantly higher BNP than NYHA class 1 or 2 patients. Data are shown as mean ± standard deviation.

The types of surgery have also varied among institutes, though operations for L–R shunt, valve operations, operations for RVOT lesions, and Fontan revisions accounted for most surgeries for ACHD at many institutes. The proportions of these procedures were heterogeneously distributed, presumably in accordance with the types of corrective surgeries performed at each institute in the past. At our hospital, for example, these same four procedures accounted for more than 90% of the ACHD operations, and the proportion of Fontan revisions was especially high because of the many atrio-pulmonary connections we had conducted a decade earlier. One clinical feature specific to ACHD is a high proportion of operations for RVOT lesions. Srinathan et al. reported a dramatic increase in the number of pulmonary valve replacements and conduit replacements for RVOT lesions during 13 years of observation [4].

Our ACHD patients could be divided into two major groups: younger patients requiring reoperations following corrective surgery for complex lesions and elderly patients with simple CHD, most typically ASD. The younger patients typically underwent operations for valvular lesions or RVOT lesions, or Fontan revisions, for moderate to complex CHD.

4.2. Preoperative status

NYHA class 3 or 4 patients made up only about 10% in our series, about half of the proportion reported from other developed countries (slightly higher than 20%) [9,11,13]. This difference helps our patients look relatively well-controlled before the operations. Yet cardiologists widely recognize the clear discrepancy between the NYHA class and severity of exercise intolerance in ACHD patients. Diller et al. found that peak oxygen consumption, one of the most reliable parameters for evaluating exercise tolerance, was likely to be severely depressed even in asymptomatic ACHD patients [14]. Our exercise tolerance results on the small fraction of patients studied (18 cases) supported this discrepancy. The asymptomatic patients with impaired exercise tolerance in our study also tended to have a high mortality/major complication rate after cardiac surgery. We thus concluded that NYHA class alone, a subjective assessment by the patients themselves, was of relatively little value for assessing the risk of cardiac surgery for ACHD and was especially weak at identifying patients at high risk of mortality or morbidity.

4.3. Amount of bleeding

Complex CHD or a history of previous median sternotomy coincided with increased bleeding in our study. Yet all but 2 of our simple CHD patients underwent a first median sternotomy, whereas all but 1 of our complex CHD patients had undergone one or more sternotomies before. Hence, we were unable to estimate the impacts of complex CHD and a history of sternotomy on the amount of bleeding independently. In terms of surgical procedures, most of the patients in the simple CHD group underwent procedures that could be approached via right atriotomy alone, while many patients with moderate to complex CHD underwent procedures involving high-pressurized cardiac chambers or large vessels with longer CPB support. These differences must have also affected our results.

4.4. CPB time, Ao-clamp time, and operation time

The CPB time, Ao-clamp time, and operation time in the three groups in our series appeared to be the same or a bit longer than those in other reports, when matched by the classification for ACHD complexity [4,12]. This supports the assessment that our surgery was standard.

4.5. Duration of postoperative ventilatory support, length of ICU stay, and length of hospitalization

The length of ICU stay and length of hospitalization in the three groups tended to be longer than in the other reports, when matched by the classification for ACHD complexity [4,9,12]. Most of the complex CHD patients who required longer ICU stays and hospitalizations in our series underwent TCPC conversion and had a extremely high mortality/major complication rate. Hence, the higher proportion of TCPC conversion in our series was a major contributing factor to the longer ICU stays and hospitalizations in our study. Other factors such as country differences in health insurance systems might also have affected these parameters.

4.6. Mortality and morbidity

The overall mortality was clearly higher in our series than in other reports or the database of the Society of Thoracic Surgeons in the United States, which reported mortality in the range from 1.3% to 3.0% [6,9–13]. One series from the UK reported a remarkably high mortality of 6.3% [4]. Yet when they divided their 13-year study period into three equal time periods, the mortality in the last period was 3.4%, which was comparable to the rates in the other reports mentioned above [4]. Of the 5 cases who died in our study, 2 underwent TCPC conversion, 1 underwent TCPC completion after bidirectional Glenn operation, and 1 underwent grafting for aneurysmal change in the ascending aorta after Fontan completion with Damus–Kaye–Stansel anastomosis. Hence, the overall mortality in our study was increased by the high proportion of SV physiology patients, a subgroup of patients who often died.

Unlike mortality, the morbidity in our series (35.9%) was comparable with that in other reports (approximate range: 20–40%) [4,9,11,12].

4.7. Preoperative examinations

We found an obvious discrepancy between results of exercise tolerance testing and NYHA class in our series. Even asymptomatic or mildly symptomatic patients had moderate-severe exercise intolerance. When Diller et al. monitored ACHD patients after exercise tolerance testing, they observed a stepwise increase in mortality or hospitalization by grade of exercise intolerance [14]. Their patients with severe exercise intolerance had a rate of mortality and/or hospitalization of about 50% after 3 years of follow-up, about triple the rate of their patients with normal tolerance [14]. Although we evaluated exercise tolerance in only a small number of patients, 13 patients with moderate-severe intolerance had a high mortality/major complication rate (53.8%), whereas the other 5 patients with normal tolerance or mild intolerance had a low complication rate of 20.0% without mortality. The 6MWT was a less reliable quantitative assessment of exercise tolerance [15]. The guidelines for the 6MWT from the American Thoracic Society state that the results of the 6MWT should be considered a complement to CPET, but not a replacement [16]. Some reports, however, indicated the distance in 6MWT was correlated with peak VO_2 in CPET [17–19]. Our 13 cases with moderate-severe exercise intolerance were free of orthopedic disorders and had normal physical growth (excluding 1 short-stature female undergoing TCPC conversion), so we defined the severity of exercise intolerance simply by the walking distance or decrease in SpO_2 during the test, based on the reported relationships between the distance and peak VO_2 [18,19]. No definition for grading the severity of exercise intolerance by 6MWT has been established. Hence, our definition for 6MWT grading might be suboptimal for evaluating exercise tolerance objectively. Our results suggest that preoperative exercise tolerance testing is useful for identifying patients at high-risk of mortality or major complications. Exercise tolerance testing, especially objective testing by CPET, can thus be recommended for risk assessment in cardiac surgery for ACHD.

Perfusion mismatch can induce silent myocardial ischemia and subsequent myocardial damage, which in turn can lead to RV failure in the pathophysiologies of TOF, systemic RV, or single RV. Myocardial perfusion has been evaluated in various types of ACHD [20–23]. These evaluations have covered corrected transposition of the great arteries (TGA) [20] and postoperative states after the Senning operation for TGA [23], arterial switch operation for TGA [21], Ross operation [21], and Fontan operation [22]. Myocardial perfusion was maintained at or above the normal level at rest in every study [20–23]. Yet the elevation of coronary perfusion after adenosine administration fell short of the normal value in all of the pathophysiologies studied [20–23]. Coronary reserve, that is, the ratio of perfusion after adenosine infusion to perfusion at rest, was smaller than normal as a consequence, even in the pathophysiologies in which the perfusion at rest was greater than normal [20–23]. There has been no direct evidence that these characteristics in coronary perfusion cause silent myocardial ischemia and subsequent myocardial damage. These findings are strongly suggestive, however, of at least a causal relationship in these types of ACHD.

Of the 8 patients in whom myocardial damage was detected preoperatively in our series, seven were consistent with one of the abovementioned conditions. None of these 8 patients died or developed major complications, so MPI alone was not useful for identifying patients at high risk of poor outcome after cardiac surgery for ACHD. We propose, however, that myocardial perfusion should be evaluated in patients at high risk of myocardial damage, specifically, in patients with TOF, systemic RV, SV, or a history of coronary transplantation, to detect the presence or extent of myocardial damage. Recognition of the presence of myocardial damage can be helpful in tailoring hemodynamic management through the selection of cardiovascular-acting agents or strategies for myocardial protection.

Bolger et al. demonstrated a significant stepwise increase in BNP with increasing NYHA class or with advancing ventricular dysfunction in ACHD patients [24]. BNP was elevated to about 55 pmol/L in their patients of NYHA class 3 or 4, to 70 pmol/L in their patients with moderate ventricular dysfunction, and to 250 pmol/L in their patients with severe ventricular dysfunction [24]. Our study also demonstrated a stepwise elevation of BNP with increasing NYHA class, and the range of BNP in each class was very similar to that reported by Bolger et al. Hsu and colleagues found perioperative BNP to be predictive of mortality and morbidity after repair or palliation surgery for CHD [25–27]. In our patients, preoperative BNP alone was not highly predictive of mortality or major complications. While our results demonstrate no utility of preoperative BNP for detecting the presence of ventricular dysfunction or myocardial damage, they do suggest that preoperative BNP is a potentially valuable predictor of poor outcome after cardiac surgery in ACHD.

In a report from Dimopoulos et al., anemic ACHD patients (Hb < 13 g/dl) had threefold higher mortality than non-anemic ACHD patients during a median follow-up of 47 months at a tertiary center [28]. Dimopoulos' group also found, by as early as the first year of a 4.1-year (median) follow-up of ACHD, that patients with moderate or greater renal dysfunction (GFR < 60 ml/min/1.73 m^2) had fivefold mortality, and patients with mild renal dysfunction ($60 \leqq$ GFR < 90 ml/min/1.73 m^2) had significantly higher mortality, compared with patients with normal renal function (GFR $\geqq 90$ ml/min/1.73 m^2) [29]. Neither anemia nor renal dysfunction affected the mortality/major complication rate after cardiac surgery in the present study. While we believe these factors to be less predictive of poor outcomes after cardiac surgery, more data from a larger number of patients will be necessary to confirm this.

Our study is subject to several limitations. Anesthesiologists must closely consider the measures used to assess the risk of intraoperative hemodynamic instability preoperatively. Here, however, we made no attempt to evaluate intraoperative hemodynamic instability, although we did assess the usefulness of preoperative examinations for predicting the outcomes after cardiac surgery for ACHD. In addition, exercise tolerance testing or MPI was performed in a small population of patients with a high frequency of complex ACHD to elucidate the predictive values of these examinations in detecting patients at high risk of poor outcomes. We await further studies with larger numbers of patients, with more widely varying pathophysiologies, from simple to complex ACHD lesions, and with assessments of intraoperative hemodynamic instability in addition to outcomes after cardiac surgery.

In conclusion, this retrospective review demonstrated that cardiac surgery could be safely performed in most ACHD cases. In ACHD with severe complexity, especially SV physiology, however, the risks of bleeding, mortality, and morbidity were markedly increased and the ICU stay and hospitalization were markedly prolonged. A single preoperative examination reliable in predicting poor outcome after cardiac surgery for ACHD has yet to be established. Exercise tolerance testing may be one of the most useful examinations for identifying patients at high risk of mortality or major complications. MPI should be applied to patients with high-risk pathophysiologies of abnormal coronary perfusion, in order to detect the presence and extent of myocardial damage. BNP can also be valuable in predicting poor outcomes after cardiac surgery.

Research support

None.

Conflict of interest

None.

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