Original Article

Longitudinal changes in ventricular function and atrioventricular valve failure following cardiac morphology after Fontan procedure

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Background: Ventricular dysfunction and atrioventricular valve (AVV) failure in Fontan patients are associated with adverse long-term outcomes; however, data on longitudinal changes and their relationship with morphology are lacking. This study aimed to describe longitudinal changes in ventricular function and AVV regurgitation and to determine the risk factors in Fontan patients who develop ventricular dysfunction and AVV failure.

Methods: We retrospectively reviewed echocardiographic images from patients who underwent Fontan procedure from 1984 to 2015. Mixed-effects model fits a unique linear regression line using serial ejection fraction (EF) and AVV regurgitation. Multivariate logistic regression was performed to find morphologic risk factors for ventricular dysfunction and AVV failure.

Results: Out of 174 patients who underwent 3,203 echocardiograms, a significant 6.2% decrease in EF was observed ($P<0.001$) over a median follow-up of 17.6 years (interquartile range, 15.3–19.2 years) post Fontan procedure. Higher prevalence of ventricular dysfunction (EF <50%) was noted in dominant right ventricle (RV) and two-ventricular (2V) morphologies compared to left ventricle (LV) ($P<0.001$). AVV failure was more common in RV and 2V morphologies as well ($P<0.001$). Notably, patients with tricuspid valve (TV) and common AVV exhibited the most pronounced AVV failure ($P<0.001$). In multivariate analysis, RV, TV and common AVV were correlated with AVV failure (hazard ratio 5.37, 8.24 and 5.43, respectively).

Conclusions: Fontan patients with long-term follow-up showed a progressive decline in ventricular and AVV function. Dominant RV, TV and common AVV were prognostic factors for predicting AVV failure. Further studies are warranted to explore and validate these findings.

Keywords: Fontan operation; functional single ventricle; atrioventricular valve (AVV); echocardiography; longitudinal study

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Introduction

The Fontan procedure, which has undergone multiple iterations, was first described in the late 1960s by Fontan and Baudet as a treatment for tricuspid atresia and is the culmination of staged palliation surgery for a wide range of functional single ventricle physiologies (1,2). Despite the significant improvement in short-term outcomes of patients with single-ventricle physiology after the Fontan operation, long-term outcomes remain suboptimal (3,4). There has been a reported burden of long-term morbidity and mortality associated with ventricular dysfunction and atrioventricular valve (AVV) regurgitation (5-7). Previous studies reported that the risk of long-term outcomes is associated with poorer ventricular performance, increased ventricular dilatation, worse strain, lower functional status, and AVV failure (5,8,9).

To determine the influence of underlying risk factors, the morphology of the dominant ventricle and AVV has been the focus of analysis, and existing clinical data have demonstrated that the progressive nature of ventricular dysfunction and AVV failure may be accompanied by right ventricular (RV) dominance or common AVV (CAVV) morphology (10-12). However, longitudinal changes in ventricular function after AVV regurgitation and their relationship with morphology remain poorly investigated (8,13). Most studies were cross-sectional in design, with restrictions on the quantity of trajectory data and a limited follow-up period (10,14). The prognosis of patients with Fontan circulation is significantly influenced by ventricular dysfunction and AVV failure. Identifying the risk factors that affect ventricular dysfunction and AVV failure allows appropriate management and vigilant monitoring, potentially optimizing the long-term outcomes of post-Fontan surgical patients.

In the present study, we aimed to assess longitudinal changes in ventricular function and AVV regurgitation after the Fontan operation and to determine the risk factors in Fontan patients who develop ventricular dysfunction and AVV failure. We present this article in accordance with the STROBE reporting checklist (available at https://jtd.amegroups.com/article/view/10.21037/jtd-2025-231/rc).

Highlight box

Key findings

- In 174 Fontan patients (median follow-up: 17.6 years), ejection fraction (EF) declined significantly (6.2%, P<0.001). Ventricular dysfunction and atrioventricular valve (AVV) failure were more common in right ventricular (RV) and two-ventricle (2V) morphologies, especially in tricuspid valve (TV) and common AVV cases. RV, TV, and common AVV were independent risk factors.

What is known and what is new?

- Fontan patients are at long-term risk for ventricular dysfunction and AVV failure.
- This study demonstrates a progressive deterioration in EF and AVV function after Fontan procedure using longitudinal data and identifies RV, TV, and common AVV morphological features as predictors of AVV failure.

What is the implication, and what should change now?

- Early surveillance is crucial for high-risk patients (RV, 2V). Regular echocardiographic monitoring and targeted management strategies are needed to prevent functional decline and improve outcomes.

Methods

Data collection and patient selection

This was a retrospective, longitudinal, single-center cohort study. Patients who underwent the Fontan procedure between January 1984 and December 2015 were included in this study. Patients with insufficient echocardiographic data or shorter follow-up periods <5 years were excluded. All patients in this study were treated at Severance Hospital, Yonsei University Health System, Seoul, Korea, which oversees clinical care and research activities. This study was approved by the Institutional Review Board of Yonsei University Health System (No. 2023-1068-001) and conducted in accordance with the Declaration of Helsinki and its subsequent amendments. Individual informed consent was waived due to the retrospective nature of the study using anonymized clinical data.

Echocardiography

All echocardiograms were obtained from the initial Fontan procedure until death, heart transplantation, or the last follow-up through December 2022. The ejection fraction (EF) and AVV regurgitation data were obtained using pre- and postoperative echocardiogram images, which were performed at the time of the pre-Fontan procedure, before discharge, as well as at least 6 months, and 1, 3, 5, 10, 15, and 20 years after the Fontan procedure. We approach the EF assessment, which is based on the echocardiographer's collation of multiple methods of EF measurement (M-mode or 2-dimensional echocardiography using the Quinones formula from the parasternal views or by the quantitative 2-dimensional biplane volumetric Simpson method from

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4- and 2-chamber views) (15), into an EF assessment quoted in the final impressions. To minimize interobserver variability, all EF measurements were independently reviewed by two investigators (A.Y.K., J.E.B.). When discrepancies were noted, consensus was achieved through collaborative discussion. This approach was implemented to enhance consistency and reliability in the retrospective data collection process. The final EF value used for analysis was based on this consensus evaluation, ensuring the most accurate representation of ventricular function. AVV regurgitation was assessed qualitatively and semi-quantitatively using Doppler color imaging and classified as mild, moderate, or severe. The classification was recorded as follows: 1 = trivial/mild (thin jet that extends to the atrial wall), 2 = moderate (broad jet that extends to the atrial wall), and 3 = severe (broad jet that occupies more than half of the atrial chamber). In the case of two AVVs, the AVVR was classified based on the most severe regurgitation, which is expected to have the greatest hemodynamic impact (5,16).

Term definition and outcome measures

The dominant ventricle morphology in patients who needed the Fontan procedure was categorized as RV, left ventricle (LV), or two-ventricle (2V) types. The 2V morphology was defined as adequate ventricles if the ventricular size itself did not preclude anatomic biventricular repair and two unbalanced ventricles with borderline ventricles in one of the two, but not rudimentary. All data for these patients were reexamined using available records and echocardiography image by investigators (A.Y.K., J.E.B., and J.E.H.) to confirm the adequacy of both ventricles by means of expert consensus opinion using usual assessment of ventricular size, including adequacy of AVV inflow, ventricular length, and proportion compared with the "systemic" ventricle. AVV morphology was grouped into mitral valve (MV), tricuspid valve (TV), CAVV, and two separate valve groups. To evaluate the potential impact of a surgical era on long-term outcomes, patients were stratified into three groups based on the period of their Fontan operation: 1984–1990, 1991–2000, and 2001–2015. These groupings correspond to distinct periods of surgical technique evolution and perioperative management, including the transition from atriopulmonary connections to lateral tunnels and subsequently to extracardiac conduits, as well as advancements in postoperative care. Ventricular dysfunction was defined as an EF <50% during follow-up. Ventricular dysfunction was defined as the definition by experts as the presence of single-ventricle EF <50%, regardless of dominant ventricular morphology at any point during the follow-up period (17-19). AVV failure is defined as either moderate to severe AVV regurgitation or that necessitates AVV replacement or repair.

Statistical analysis

Basic demographic and clinical information were presented as median and interquartile range (IQR) or mean ± standard deviation for continuous variables after testing the normality via the Shapiro-Wilk test and as numbers or percentages for categorical variables. Continuous variables were compared using two-sample *t*-tests or Mann-Whitney *U*-tests, and categorical variables were compared using Chi-square test. Linear mixed-effects regression models composed of fixed and random effects that fit a linear regression line for each participant were used to assess the longitudinal change in EF with irregularly spaced time points. To assess changes in AVV regurgitation in Fontan patients stratified by dominant ventricle and AVV morphology, we treated AVV regurgitation grade as an ordinal variable to examine its progression across different morphology types. Cox proportional hazards regression analyses were used to identify the independent risk factors for ventricular dysfunction and AVV failure. Spearman's rank correlation test was used to assess correlation between ventricular dysfunction and AVV failure. Statistical analyses were performed using SAS (version 9.4; SAS Institute Inc., Cary, North, USA) and R version 4.0.4 (R Foundation for Statistical Computing, Vienna, Austria).

Results

Demographic patient characteristics and operative findings

A total of 174 patients who underwent the Fontan procedure, and 3,203 echocardiographic examinations were included. The baseline characteristics and anatomic diagnoses are shown in *Table 1*. The average age at the time of Fontan procedure was found to be 5.6±4.3 years and 77/174 (44.3%) patients were female. The dominant ventricular morphology was LV in 85 (50%), RV in 50 (29.4%), and 2V morphology in 35 (20.6%). Regarding AVV morphology, 71 patients (41.8%) had MV, 37 (21.8%) had CAVV, 32 (18.8%) were TV group, and 30 (17.6%) had with separate AVV morphologies. Among the primary diagnoses, tricuspid atresia was the most prevalent, accounting for 23% (n=40) of the cases. The median

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Table 1 Baseline demographic and operative characteristics in patients

Factors	Values (N=174)
Age at Fontan procedure (years)	5.6 (3.5–8.9)
Female	77/174 (44.3)
Follow-up periods (years)	17.6 (15.3–19.2)
Primary diagnosis	
DORV	46/174 (26.4)
TA	40/174 (23.0)
PA	27/174 (15.5)
HLHS	10/174 (5.7)
AVSD	15/174 (8.6)
TGA	13/174 (7.5)
DILV	9/174 (5.2)
Ebstein's anomaly	6/174 (3.4)
Others	8/174 (4.6)
Dextrocardia	24/174 (13.8)
Isomerism	
Left	8/174 (4.6)
Right	18/174 (10.3)
Fenestration	90/174 (51.7)
Era of Fontan	
1984–1997	22/174 (12.6)
1998–2007	79/174 (45.4)
2008–2022	73/174 (42.0)
Fontan type	
APC	6/174 (3.4)
LT	63/174 (36.2)
ECC	97/174 (55.7)
Kawashima	8/174 (4.6)
Dominant ventricle†	
LV	85/170 (50.0)
RV	50/170 (29.4)
2V	35/170 (20.6)
AVV morphology‡	
Mitral valve	71/170 (41.8)
CAVV	37/170 (21.8)
Tricuspid valve	32/170 (18.8)
Two valves	30/170 (17.6)

All data are presented as n/N (%) or median (interquartile range). †, four patients have been classified as undetermined in the dominant ventricle type. ‡, four patients have been classified as undetermined in the valve morphology type. APC, atriopulmonary connection; AVSD, atrioventricular septal defect; AVV, atrioventricular valve; CAVV, common atrioventricular valve; DILV, double inlet left ventricle; DORV, double outlet right ventricle; ECC, extracardiac conduit; HLHS, hypoplastic left heart syndrome; LT, lateral tunnel; LV, left ventricle; PA, pulmonary atresia; RV, right ventricle; TA, tricuspid atresia; TGA, transposition of the great arteries; 2V, two-ventricular morphology.

follow-up period for the entire cohort was 17.6 years (IQR, 15.3–19.2 years). Each patient has had 18.4 times of echocardiography during follow-up. Transplantation and takedown free survival after Fontan were 98.8%, 98.0%, and 96.5% at 5, 10, 15 years, respectively.

Temporal changes in ventricular function and AVV regurgitation

Analysis of the serial echo image revealed a gradual decline in EF over a period of 15 years, with a decrease of approximately 6.2% (P<0.001) (*Figure 1A*). When comparing changes in EF over time on the dominant ventricle morphology, the RV and 2V morphology showed a tendency to demonstrate a more pronounced decrease in EF compared to the LV (P=0.42, *Figure 1B*, Table S1). AVV regurgitation progressively increased over time (P<0.001) as well. When assessing the serial changes in AVV regurgitation over time according to the dominant ventricle type, RV and 2V exhibited a greater increase (P<0.001, *Figure 2A*). In terms of valve morphology type, as time progressed, AVV regurgitation increased more in the TV, CAVV, and MV than in the two valves (P=0.009, *Figure 2B*).

Morphologic risk factors for ventricular dysfunction and AVV failure

We analyzed the characteristics comparison and cumulative incidence of ventricular dysfunction, defined as a single-ventricle EF <50% and the occurrence of AVV failure at any point during the follow-up period after the Fontan procedure (*Tables 2,3* and *Figure 3*). During the follow-up period, a total of 54 (28.7%) patients experienced ventricular dysfunction with similar baseline characteristics, except for a higher proportion of dominant ventricle morphology in RV (*Table 2*). In the Kaplan-Meier analysis, the occurrence rates of ventricular dysfunction over 15 years after surgery were 48.1% in the RV, 34.6% in the 2V, and 17.3% in the LV, based on the dominant ventricle morphology. The RV had the highest occurrence rate of ventricular dysfunction, and the 2V had a higher occurrence rate compared to the LV (*Figure 3A*) (log-rank P<0.001). However, no risk factors, including dominant ventricular morphology and Fontan type, were identified in the multivariate analysis (Table S2).

A total of 27 patients (15.5%) experienced AVV failure. Based on the dominant ventricle type, the incidence rates of AVV failure were 48.1% in the 2V, 37.0% in the RV, and 14.8% in the LV. The RV and 2V had higher occurrence

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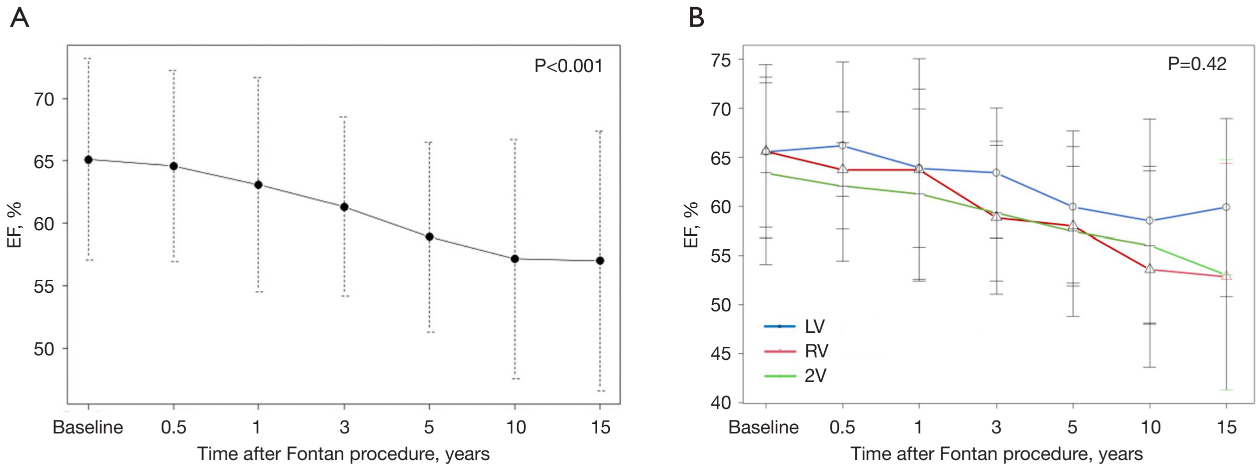

Figure 1 Changes in EF in patients who underwent the Fontan procedure. Changes are shown (A) according to postoperative periods and (B) stratified by dominant ventricle morphology. EF, ejection fraction; LV, left ventricle; RV, right ventricle; 2V, two-ventricular morphology.

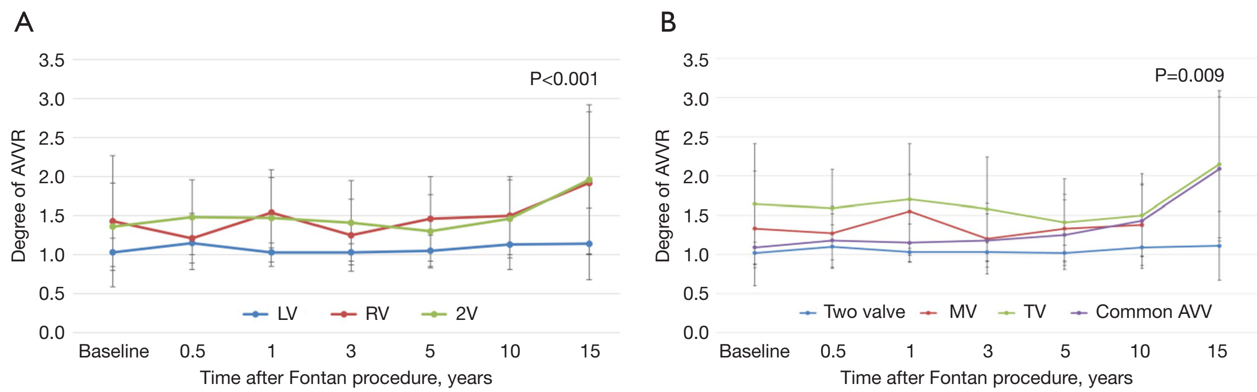

Figure 2 Changes in AVV regurgitation in patients who underwent the Fontan procedure stratified by dominant ventricle morphology and AVV morphology. Changes are stratified by (A) dominant ventricle morphology and (B) AVV morphology. AVV, atrioventricular valve; AVVR, atrioventricular valve regurgitation; LV, left ventricle; MV, mitral valve; RV, right ventricle; TV, tricuspid valve.

rates of AVV failure compared to the LV (*Figure 3B*) (log-rank P<0.001). The incidence rates of AVV failure based on AVV morphology type over 15 years after surgery were 38.5% in the TV, 42.3% in the CAVV, 7.7% in the MV, and 11.5% in valve morphologies (*Table 3*). The Kaplan-Meier curve based on valve morphology type revealed that the TV and CAVV had a higher occurrence rate compared to the two valve and MV (*Figure 3C*) (log-rank P<0.001). After all variables adjustment, the RV morphology [HR: 5.37, 95% confidence interval (CI): 1.60–18.04, P=0.007] in dominant ventricle, TV (HR: 8.24, 95% CI: 3.53–22.74, P<0.001) and CAVV (HR: 5.43, 95% CI: 1.52–19.41, P=0.009) morphology in AVV were correlated with AVV failure, while the adjusted HR for the 2V could not be accurately calculated due to a lack of patient numbers (Table S3).

Discussion

In this study, we analyzed 3,203 longitudinal echocardiographic data on ventricular function and AVV function tracked in 174 individual patients for a median of 17.6 years, with the longest follow-up being 23 years after the Fontan procedure. Our main findings were as follows: (I) ventricular function decreased and AVVR increased after more than 15 years following the Fontan procedure; (II) AVV failure is

Table 2 Comparison of characteristics in relation to ventricular dysfunction

Factors	Ventricular dysfunction (EF <50%)		
	Yes (N=54)	No (N=120)	P value
Age (years)	6.9±7.6	5.1±5.6	0.09
Female	20/54 (37.0)	57/120 (47.5)	0.20
Fontan type			0.12
APC	3/54 (5.6)	3/120 (2.5)	
LT	16/54 (29.6)	47/120 (39.2)	
ECC	30/54 (55.6)	67/120 (55.8)	
Kawashima	5/54 (9.3)	3/120 (2.5)	
Fenestration	28/54 (51.9)	62/120 (51.7)	0.98
Dominant ventricle†			<0.001
LV	9/52 (17.3)	76/118 (64.4)	
RV	25/52 (48.1)	25/118 (21.2)	
2V	18/52 (34.6)	17/118 (14.4)	
AVV morphology‡			<0.001
Mitral valve	9/51 (17.6)	62/119 (52.1)	
Tricuspid valve	17/51 (33.3)	15/119 (12.6)	
CAVV	19/51 (37.3)	18/119 (15.1)	
Two valve	6/51 (11.8)	24/119 (20.2)	
Isomerism			0.47
Left	4/54 (7.4)	4/120 (3.3)	
Right	6/54 (11.1)	12/120 (10.0)	
Dextrocardio	12/54 (22.2)	12/120 (10.0)	0.03

Values are shown as n/N (%) or mean ± standard deviation. †, four patients have been classified as undetermined in the dominant ventricle type; ‡, four patients have been classified as undetermined in the valve morphology type. APC, atriopulmonary connection; CAVV, common atrioventricular valve; ECC, extracardiac conduit; EF, ejection fraction; LT, lateral tunnel; LV, left ventricle; RV, right ventricle; 2V, two-ventricular morphology.

Table 3 AVV failure comparison of baseline characteristics

Factors	AVV failure		
	Yes (N=27)	No (N=147)	P value
Age (years)	6.5±6.6	5.5±6.3	0.47
Female	17/27 (63.0)	60/147 (40.8)	0.03
Fontan type			0.04
APC	0/27 (0.0)	6/147 (4.1)	
LT	8/27 (29.6)	55/147 (37.4)	
ECC	15/27 (55.6)	82/147 (55.8)	
Kawashima	4/27 (14.8)	4/147 (2.7)	
Fenestration	13/27 (48.1)	77/147 (52.4)	0.69
Dominant ventricle†			<0.001
LV	4/27 (14.8)	81/143 (56.6)	
RV	10/27 (37.0)	40/143 (28.0)	
2V	13/27 (48.1)	22/143 (15.4)	
AVV morphology‡			<0.001
Mitral Valve	2/26 (7.7)	69/144 (47.9)	
Tricuspid valve	10/26 (38.5)	22/144 (15.3)	
CAVV	11/26 (42.3)	26/144 (18.1)	
Two valves	3/26 (11.5)	27/144 (18.8)	
Isomerism			<0.001
Left	4/27 (14.8)	4/147 (2.7)	
Right	8/27 (29.6)	10/147 (6.8)	
Dextrocardio	6/27 (22.2)	18/147 (12.2)	0.22

Values are shown as n/N (%) or mean ± standard deviation. †, four patients have been classified as undeterminated in the dominant ventricle type; ‡, four patients have been classified as undeterminated in the valve morphology type. APC, atriopulmonary connection; AVV, atrioventricular valve; CAVV, common atrioventricular valve; ECC, extracardiac conduit; LT, lateral tunnel; LV, left ventricle; RV, right ventricle; 2V, two-ventricular morphology.

associated with TV and CAVV morphologies.

Our findings are consistent with those of previous cross-sectional studies. Moon *et al.* (10) found that the dysfunction of a single ventricle and AVVR increases over time and adversely affects the clinical outcomes of the Fontan procedure, with an inferior survival rate, especially in a single RV. However, in this patient cohort, longitudinal changes are likely to be more informative, as they could provide important information on how the ventricle adapts to changes in loading conditions after Fontan circulation is completed. There is a scarcity of data concerning longitudinal changes in ventricular function post-Fontan operation. When available, these data tend to focus on a small subset of patients (13,20) and are often based on a limited number of follow-up time points (8). The strength of our study is that it analyzed over 3,000 instances of

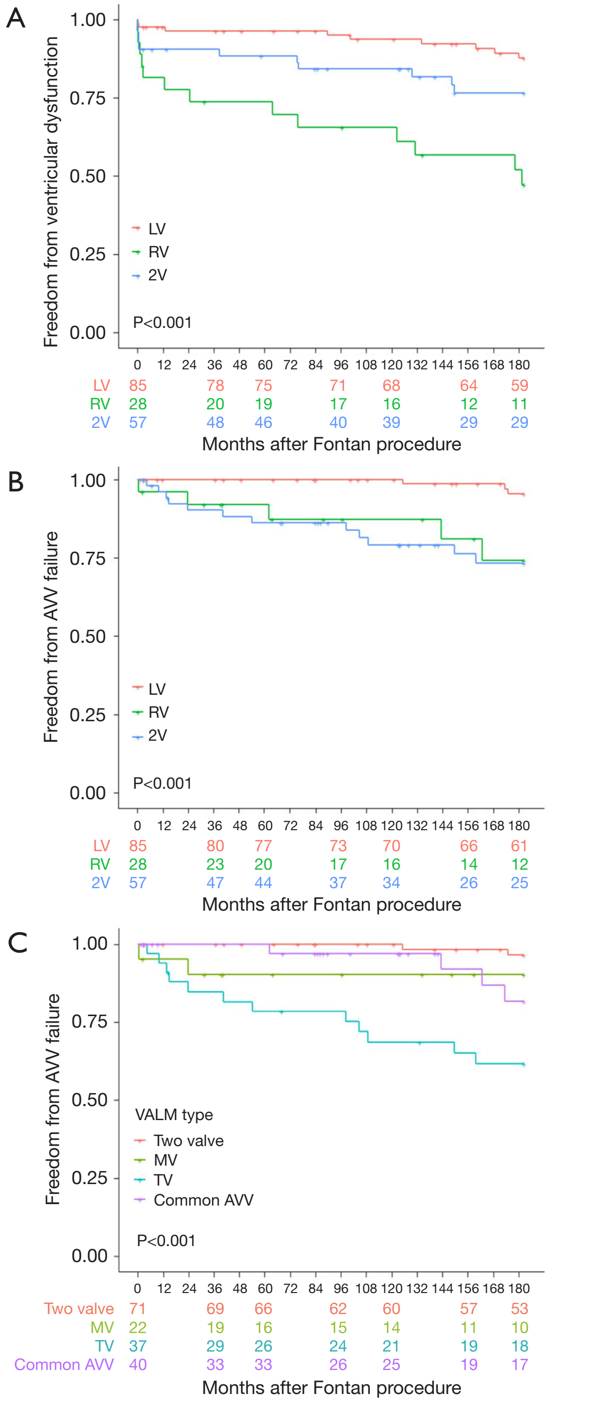

Figure 3 Freedom from ventricular dysfunction according to dominant ventricle morphology and AVV failure according to dominant ventricle morphology and AVV morphology. (A) Freedom from ventricular dysfunction according to dominant ventricle morphology and (B,C) AVV failure according to dominant ventricle morphology and AVV morphology are shown. AVV, atrioventricular valve; LV, left ventricle; MV, mitral valve; RV, right ventricle; TV, tricuspid valve; 2V, two-ventricular morphology.

paired testing and showed an overall decreasing trend in EF toward the mid-50s.

The impact of single ventricle morphology has been controversial, with short- and mid-term studies demonstrating no effect, whereas a recent longer-term study demonstrated worse outcomes with a single RV (21-24). Dominant RV and TV are recognized as adverse risk factors. Despite limited evidence, indications suggest that a dominant RV, characterized by increased dilation, higher native T1 values, and diminished torsion and untwisting compared to single LVs, may have detrimental effects, supported by our study results (13). Additionally, we also found that a substantial number of patients with 2V morphology experienced unfavorable outcomes than LV morphology group. While many previous studies from North America and Europe have reported a higher prevalence of hypoplastic left heart syndrome (HLHS), we observed a higher incidence of patients with the 2V morphology in our population. Oster and colleagues (24) demonstrated that of 3,807 subjects from 44 centers throughout the United States, 42% of systemic RV and also 20% of non-classifiable, which included single ventricles without clearly defined morphology or conditions with systemic support by two ventricles that were not amenable to ventricular separation, showed unfavorable survival than systemic LV. Marathe *et al.* (25) reported that the RV and 2V groups showed a trend toward worse outcomes in terms of freedom from Fontan failure, although the difference was not statistically significant. *Figure 4* illustrates the longitudinal trajectories of ventricular dysfunction and AVV failure across morphologic subtypes. To the best of our knowledge, this study represents the first identification of a heightened risk of ventricular dysfunction in both the RV and 2V groups compared to the LV group. Although the multivariate Cox regression did not yield statistically significant results, this may be due to the small sample size, and it suggests that further research is warranted to validate these findings.

One potential reason for the lack of superiority in the 2V Fontan group, despite having a larger myocyte mass, could be preload deficiency (26). As pulmonary circulation acts as a resistor in the circuit, the additional myocardium distal to the lungs may not make a difference and would have no impact on systemic venous hypertension, thereby continuing to pose problems. Penny and colleagues (27) demonstrated abnormal and asynchronous intraventricular and diastolic filling in this group of patients could be attributed to a combination of preload reduction and acquired ventricular

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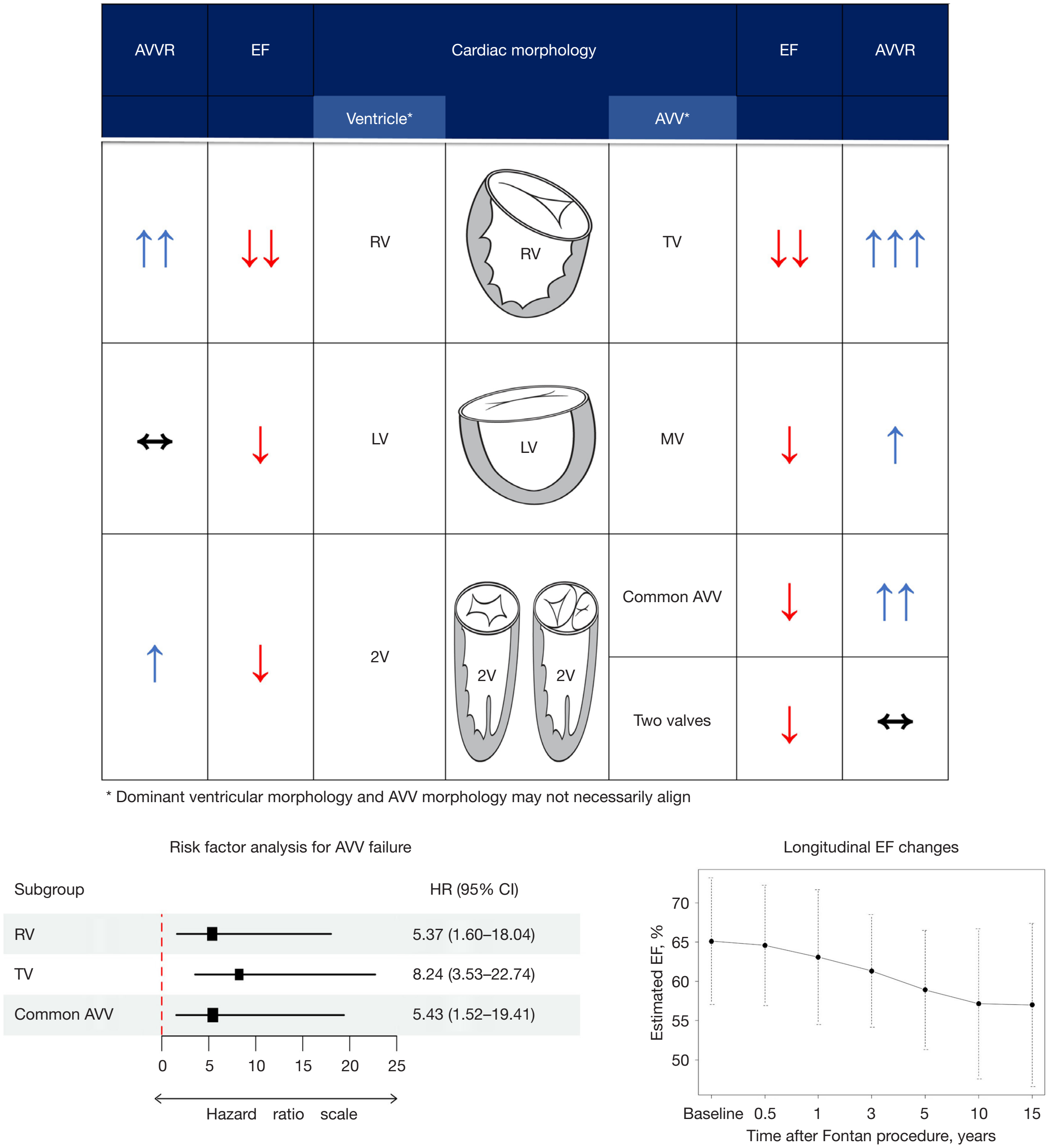

Figure 4 Longitudinal changes in ventricular function and AVV failure according to cardiac morphology. The top matrix summarizes overall trends in ventricular EF and AVV regurgitation across various ventricular and valvular morphologies. Arrow indicates the trend and severity of changes in EF and AVV function by morphology (e.g., ↓↓ indicates marked decline, ↑↑ indicates marked improvement, ↔ indicates minimal change). The forest plot (bottom left) displays multivariate Cox regression analysis for risk factors predicting AVV failure. The line graph (bottom right) shows the estimated mean EF over time after the Fontan procedure. Note: dominant ventricular morphology and AVV morphology may not necessarily align in individual patients. AVV, atrioventricular valve; CI, confidence interval; EF, ejection fraction; HR, hazard ratio; LV, left ventricle; MV, mitral valve; RV, right ventricle; TV, tricuspid valve; 2V, two-ventricular morphology.

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hypertrophy. Yamamura *et al.* (28) similarly demonstrated that anomalous mechanics may not effectively lower end-diastolic pressure, thereby negating the potential advantage of a larger myocyte mass (29). Coupling the RV with the LV in 2V Fontan circulation may not circumvent this problem, and ventricular interaction could mean that right ventricular dysfunction negatively influences the mechanics of both ventricles.

Our study found a significantly higher occurrence rate of AVV failure in cases where the dominant RV and TV were present, consistent with the results of previous studies (6,10,30). We further investigated whether a dominant RV is negatively associated with long-term survival following the Fontan procedure, possibly due to a tendency toward progressive AVVR and deterioration of single ventricle function because of the vicious cycle of ventricular dilatation and worsening regurgitation (6,31). Although the causal relationship between ventricular dysfunction and AVV regurgitation remains incompletely understood (5,6,32), our data showed a statistically significant positive correlation between the two (Spearman's rank correlation coefficient =0.329, P<0.001) (Figure S1). In addition, CAVV in Fontan patients is known to be unfavorable (10,29,33), as in our data, and among RV and 2V patients with a high incidence of AVV failure compared to mitral valve and two-valve morphology, whereas CAVV is frequently observed with AVV failure. Nevertheless, cases with 2V morphology and CAVV also had higher occurrence rates than cases with LV dominance and MV. It can be noted that AVV morphology has a greater impact, independent of ventricle morphology. The cumulative incidence of valve intervention and significant regurgitation in our group steadily increased; therefore, with a longer follow-up period, the incidence of valve failure will continue to increase.

Study limitation

Our study has several limitations owing to its retrospective nature, which inherently renders it susceptible to certain biases. Echocardiograms were obtained at the discretion of the patient's providers rather than at predefined intervals, introducing a potential bias in EF assessment influenced by clinical status, age, and surgical era. Additionally, the 2D echocardiography used in the study was constrained in its ability to assess the functional single ventricle, lacking universally agreed-upon or readily available normal values for functional indices. As a result, complementary imaging techniques, such as MRI or catheterization, are necessary to supplement echocardiography results. Despite these limitations, we consistently obtained adequate echocardiographic data from the same patients, facilitating the monitoring of serial changes in ventricular function, as recommended by the present recommendations (34). Additionally, long-term data provide valuable insights into ventricular function progression in Fontan survivors, with reduced interobserver variability through consensus review further strengthening the reliability of our findings. A notable limitation of our study is the potential selection bias due to the inclusion of only long-term survivors of the Fontan procedure. Our cohort demonstrated a high rate of transplant-free and Fontan takedown-free survival, suggesting that these patients represent a relatively healthier subset of the broader Fontan population, often referred to as "well Fontans". As a result, the percentage of ventricular dysfunction or AVV failure may be underestimated, and the number of patients with each morphological variation may be limited. This selection bias may affect the generalizability of our findings to the entire Fontan population.

Conclusions

After Fontan palliation, a gradual decline in ventricular function was observed. The propensity for developing ventricular dysfunction and AVV failure was notably increased in cases with RV and 2V morphology. Specifically, TV and CAVV exhibited greater susceptibility to AVV failure. Prudent consideration of these findings underscores the importance of rigorous surveillance, close monitoring, and timely interventions. Subsequent investigations of the clinical implications of ventricular and AVV morphologies are warranted to validate and elucidate these observations.

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Footnote

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Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. This study was conducted in accordance with the Declaration of Helsinki and its subsequent amendments. The study was approved by the Institutional Review Board of Yonsei University Health System (No. 2023-1068-001). Individual consent for this retrospective analysis was waived due to the nature of the study using anonymized clinical data.

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