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Surgical results of patients with a functional single ventricle

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Abstract

Objective: Overall mortality of the patients with a functional single ventricle is still high, though excellent mid- and long-term results of the modified Fontan procedure have been reported. This study assessed the factors that affect the surgical outcomes mainly in the pre-Fontan stage and performed long-term survival analysis. **Methods**: Between January 1988 and December 2000, 405 patients with a functional single ventricle underwent surgical interventions and were followed up until June 2001. The mean follow-up period was 74.5 ± 69.4 months and 95% of the patients were followed up completely. Their median age was 2.5 months at the time of shunt or pulmonary artery banding (PAB), 8.6 months at BCPS, and 28.6 months at the Fontan operation. The variables of the anatomical lesions with single ventricle physiology, combined abnormalities, surgical pathways leading to the Fontan stage, age at operation, study periods, and type of the Fontan procedure were analyzed. The role of BCPS in the long-term results was evaluated. **Results**: Overall survival after birth was 60.1 ± 2.8% at 10 years. In multivariate analysis, complete atrioventricular septal defect-typed lesion, pulmonary venous obstruction, total anomalous pulmonary venous connection, and interruption of aortic arch were risk factors for long-term survival, while pulmonary stenosis was demonstrated as a favorable prognostic factor. In this study, there was no significant survival difference between the early and late study period. Actuarial mortality in the pre-Fontan stage was 41.3% in the non-BCPS group and 16.3% in the BCPS group ($P < 0.001$). The 10-year survival rates of the populations in staged and primary Fontan groups were not significantly different ($P = 0.24$). The long-term survival rate of the atriopulmonary Fontan group was significantly lower than that of lateral tunnel Fontan (60.3 ± 6.3% vs. 86.8 ± 3.1% at 10 years, $P = 0.0001$). **Conclusion**: This study revealed that the overall survival was disappointing and there were still problems that need to be solved in the pre-Fontan stage to improve the overall survival. The role of BCPS was not to contribute to the longer survival after Fontan operation, but to lower mortality in the pre-Fontan stage, which can offer a higher probability to proceed to the Fontan procedure successfully.

Keywords: Functional single ventricle; Long-term survival; Risk factors

1. Introduction

Patients with a functional single ventricle have various underlying cardiac anomalies and experience diverse surgical pathways leading to the Fontan operation. With the introduction of new surgical modifications of the Fontan procedure [1–3], excellent mid- and long-term results have been reported [4–7]. However, the long-term survival of patients with a functional single ventricle still remains to be improved. In this study, we performed the long-term survival analysis upon patients with a functional single ventricle with various surgical experiences prior to the Fontan operation and types of operation, and assessed the factors that influenced the results.

2. Patients and methods

Between January 1988 and December 2000, 405 patients with a functional single ventricle underwent surgical interventions at Seoul National University Children's Hospital and followed up until June 2001. The mean

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follow-up period was 74.5 ± 69.4 months and follow-up loss occurred in 21 patients (5%). Two hundred and sixty patients were male and 145 were female. The anatomical lesions with single ventricle physiology are listed in Table 1. Initial palliation included systemic to pulmonary shunt, pulmonary artery banding (PAB), bidirectional cavopulmonary shunt (BCPS), or the Norwood procedure. Patients' median ages at the time of palliations were 2.6 months (0–9.5 years) for shunt ($n = 156$), 2.6 months (8 days–5.2 years) for PAB ($n = 82$), 21 days (2 days–8.2 months) for Norwood procedure ($n = 17$), and 8.6 months (1.7 months–32.9 years) for the BCPS ($n = 125$). Median age at Fontan operation was 28.6 months (7.5 months-18.5 years).

The risk analysis for long-term survival was performed with respect to the anatomical lesions, combined morphologic anomalies, and various surgical pathways leading to the Fontan operation as well as whether bidirectional cavopulmonary shunt was performed as an interim procedure.

The long-term survival rates were compared between the two study periods (earlier period: 1988–1991; latter period: 1992–2000). The majority of the BCPS and lateral tunnel Fontan procedures in this study had been performed since 1992, which means major change in the surgical era and accompanied advance in perioperative management.

Surgical pathways to the Fontan operation were divided into four: (1) shunt or PAB followed by Fontan ($n = 81$), (2) shunt or PAB followed by BCPS and Fontan ($n = 29$), (3) BCPS as an initial palliation followed by Fontan ($n = 31$) and (4) direct Fontan ($n = 88$).

Table 1
Diagnosis and multivariable analysis for long-term survival after birth

Anatomical lesions associated with single ventricle physiology	*n* (%)	Multivariable analysis (Cox regression) Stepwise forward conditional method	
		P-value	Exp(B)
Double inlet ventricle	206 (50.9)	n.s.	
DIRV	140 (34.6)	n.s.	
DILV	48 (11.9)	n.s.	
DIUV	18 (4.4)	n.s.	
Tricuspid atresia	70 (17.3)	n.s.	
Mitral atresia	21 (5.2)	n.s.	
HLHS	14 (3.5)	<0.001	15.93
PA with IVS	7 (1.7)	n.s.	
DORV	52 (12.8)	n.s.	
Complete AVSD	51 (12.6)	0.002	1.94
AV valve straddling	12 (3.0)	n.s.	
Criss-cross heart	9 (2.2)	n.s.	
Ebstein anomaly	4 (1.0)	n.s.	

Condition of stepwise regression: entry by $P \leq 0.05$; removal by $P > 0.1$. n.s., not significant. DIRV, double inlet right ventricle; DILV, double inlet left ventricle; DIUV, double inlet undetermined ventricle; HLHS, hypoplastic left heart syndrome; PA, pulmonary atresia; IVS, intact ventricular septum; DORV, double outlet right ventricle; TGA, transposition of the great arteries; AVSD, atrioventricular septal defect; AV, atrioventricular.

Estimation of the survival rates at 5 years and 10 years were obtained by the Kaplan–Meier method with 95% confidence intervals. Survival rate comparisons between the two groups were carried out using the log-rank test. The role of the BCPS procedure was assessed by comparing the mortalities in the pre-Fontan stage and by comparing the long-term survival after Fontan operation between the BCPS and non-BCPS groups.

All of the analyzed data were selected on the basis of a review of medical records, operative notes, echocardiography and cardiac catheterization reports. Atrioventricular valve regurgitation of greater than mild degree in the echocardiography was considered to have significant regurgitation. The Cox proportional hazards regression model was used for multivariate analysis and the method of variable selection was 'Enter' or 'Forward stepwise: conditional' (entry by $P < 0.05$, removal by $P > 0.1$). Odds ratios (Exp(B)) were calculated for the significant multivariate predictors. Values of $P < 0.05$ were considered statistically significant. All statistical analyses were performed using the SPSS software package (version 10.0, SPSS Inc, Chicago, IL).

3. Results

3.1. Risk factor analysis

Among the anatomical lesions associated with single ventricle physiology, hypoplastic left heart syndrome ($P < 0.001$) and complete AVSD typed lesion ($P = 0.002$) were significant risk factors as a result of multivariate analysis (Table 1).

The results of the multivariate analysis for the morphologic risk factors for long-term survival was shown in Table 2. Total anomalous pulmonary venous connection (TAPVC), pulmonary venous obstruction, interruption of the aortic arch were identified as significant risk factors. Cox proportional hazards model revealed that single atrioventricular valve and pulmonary atresia were not independent anatomical risk factors. However, pulmonary stenosis was the only favorable prognostic factor ($P = 0.002$; relative risk ratio = 0.43).

3.2. Various surgical pathways and follow-up results

Fig. 1 shows various surgical pathways to the Fontan operation and the follow-up results. There were four surgical pathways to the Fontan procedure. Firstly, 63 patients among the 405 patients included in this study underwent BCPS and then 31 patients arrived Fontan stage. Secondly, 254 patients went through an initial palliation like shunt or banding, and then 62 patients underwent BCPS, and 29 patients finally arrived Fontan stage. Next third,

Table 2
Risk factor analysis in Single ventricle physiology ($n = 405$)

Variables	Multivariate analysis Cox proportional hazard model		
	n (%)	Exp(B)	*P*-value
Atrial situs			
Solitus	212 (52.3)	2.07	0.14
Inversus	54 (13.3)	1.46	0.46
Isomerism	43 (10.6)	0.47	0.08
Systemic venous anomaly			
Bilateral SVC	126 (31.1)	0.88	0.52
IVC interruption	9 (2.9)	0.71	0.61
AV valve regurgitation	31 (7.7)	0.99	0.97
Mode of AV connection			
Single AV valve	253 (62.5)	0.94	0.83
Two AV valve	62 (15.3)	0.48	0.16
Dominant ventricle			
Right ventricle	198 (48.9)	0.90	0.73
Left ventricle	97 (24)	0.66	0.21
Undetermined ventricle	15 (3.7)	0.73	0.60
Outflow obstruction			
Coarctation of aorta	23 (5.7)	1.48	0.47
Pulmonary stenosis	187 (46.2)	0.43	0.002
Pulmonary atresia	102 (25.2)	1.16	0.50
Interruption of aortic arch	2 (0.5)	13.55	0.001
Arch hypoplasia	15 (3.7)	2.55	0.14
Aortic stenosis	22 (5.4)	1.90	0.11
PV anomaly			
TAPVC	47 (11.6)	2.53	0.000
PAPVC	9 (2.2)	1.16	0.79
PV obstruction	5 (1.6)	4.71	0.001

SVC, superior vena cava; IVC, inferior vena cava; AV, atrioventricular; TAPVC, total anomalous pulmonary venous connection; PAPVC, partial anomalous pulmonary venous connection; PV, pulmonary vein.

88 patients underwent Fontan operation directly. Lastly, 83 patients among 254 patients who underwent this initial palliation went to the Fontan without BCPS.

Seventy-four patients died after initial palliative procedures (shunt, PAB or Norwood) and 20 patients after BCPS before arriving Fontan stage. Though not indicated in Fig. 1, 44 early death occurred after palliative procedure and 12 early deaths after BCPS procedure. After Fontan operation, 49 patients died (with 29 early deaths) and eight patients were lost from the follow-up period.

3.3. Comparison of long-term survival rates

Overall survival after birth was 60.1 ± 2.8% at 10 years (95% confidence limit) using the Kaplan–Meier method (Fig. 2). Comparisons of long-term survival of the various subgroups are summarized in Table 3. In this study, no significant differences were found between the long-term survivals with respect to the types of the initial palliation (shunt vs. PAB), ages at BCPS (early vs. non-early), and the four surgical pathways to the Fontan procedure. Long-term survival rates after Fontan operation were not significantly different between staged Fontan (via BCPS) and primary Fontan (without BCPS) group. There were no significant survival difference between the two study periods, so the fact that operation was done in the earlier periods of this study is not a major confounding factor affecting the results.

There were no significant difference in number of major risk factors including TAPVC, PV obstruction, Interruption of aortic arch, complete AVSD typed lesion between non-BCPS and BCPS group (Table 4). The result was the same

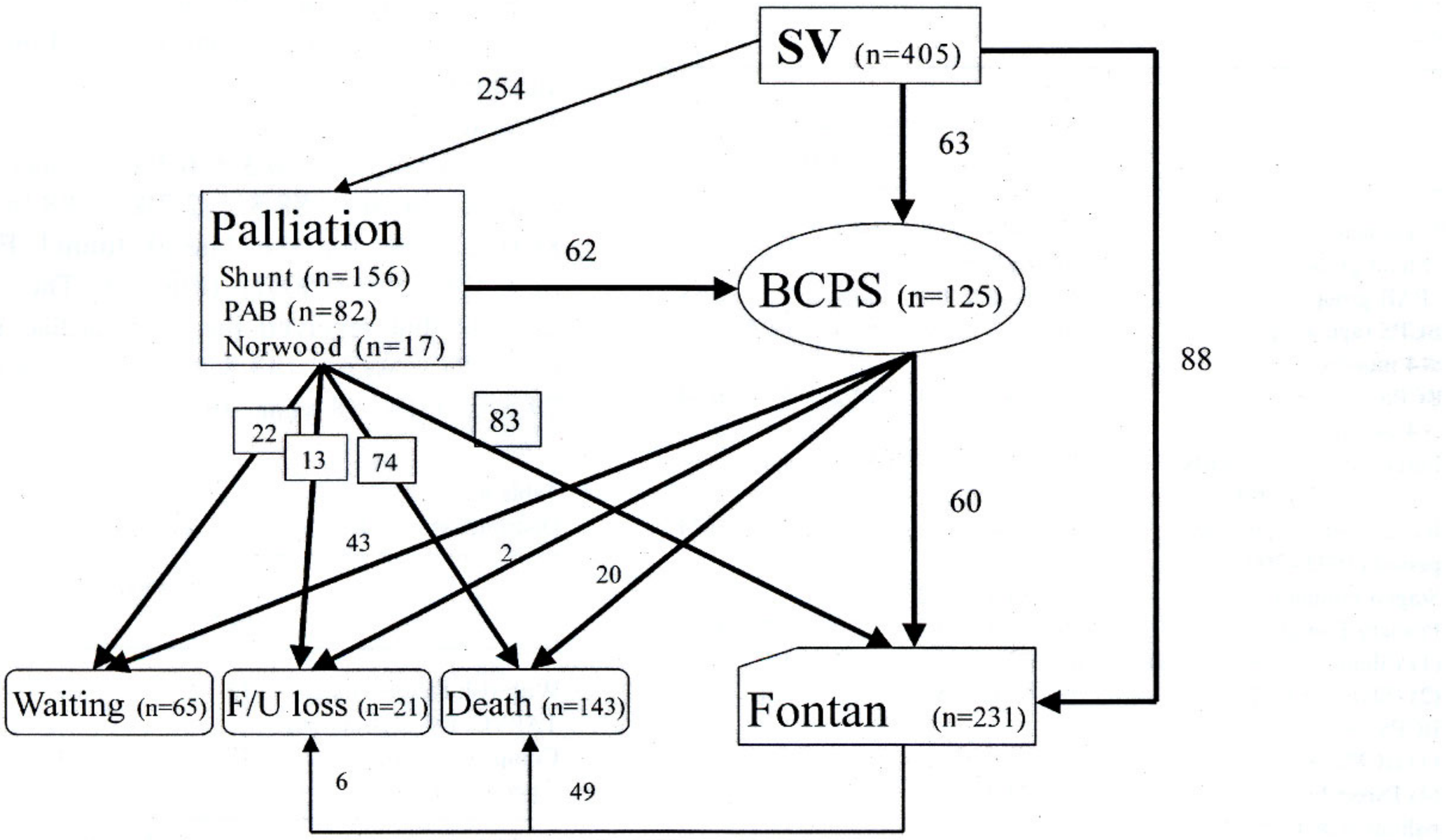


Fig. 1. Diagram of various Fontan pathways. SV, patients with a functional single ventricle; PAB, pulmonary artery banding; BCPS, bilateral cavopulmonary shunt; Waiting, waiting for Fontan operation; F/U, follow-up.

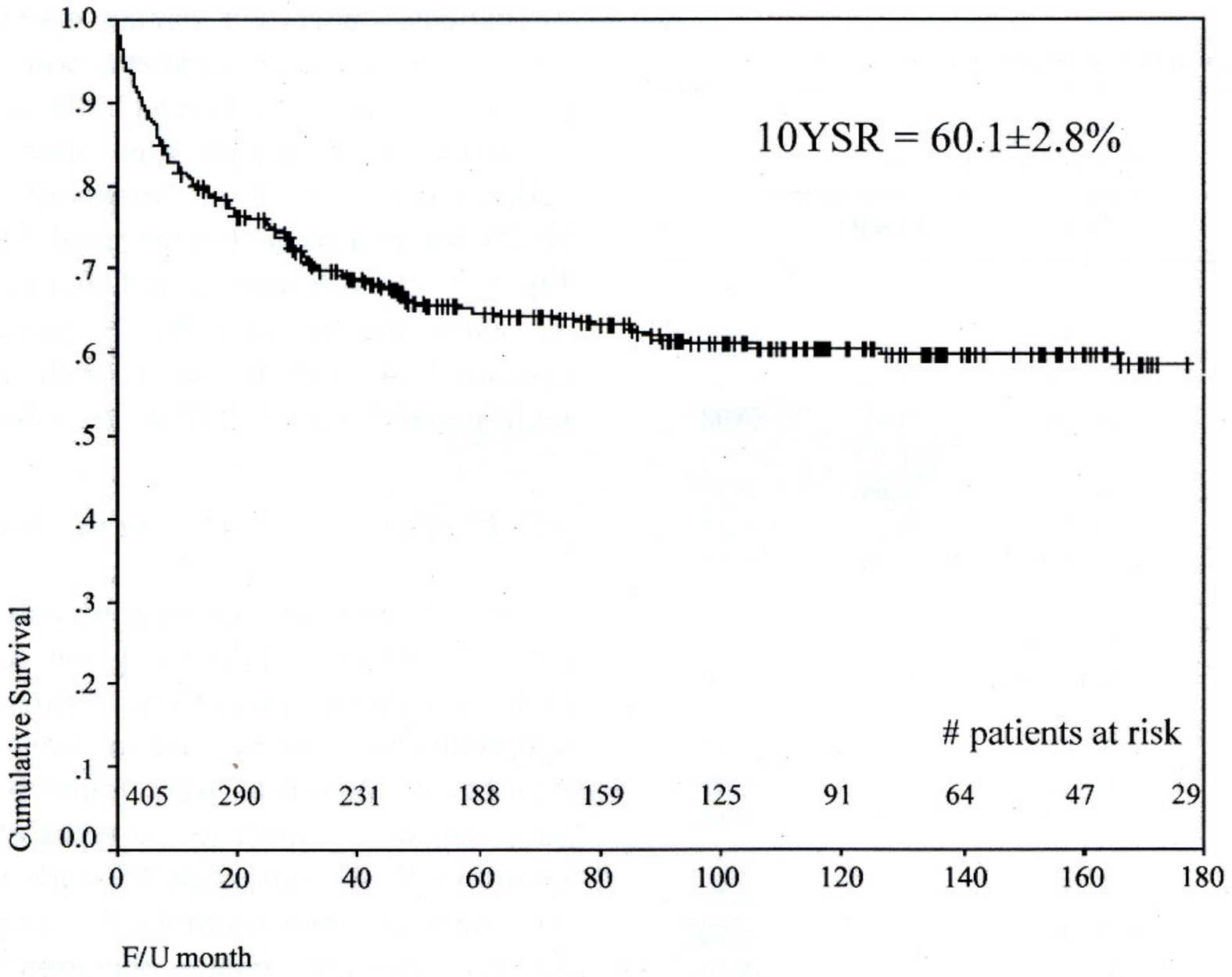


Fig. 2. Long-term survival of patients with a functional single ventricle. Starting point of the follow-up duration: from birth.

when the population between the two groups was limited to latter period of this study in which most BCPS were performed.

In the non-BCPS group ($n = 179$, follow-up loss = 13), 74 patients died before the Fontan operation, while 20 patients died in the BCPS group ($n = 123$, follow-up loss = 2). The actuarial mortalities in the pre-Fontan stage of the above two groups were 41.3% in the non-BCPS group and 16.3% in the BCPS group, and this difference was statistically significant by the Pearson chi-square test ($P < 0.001$) (Table 5).

In those patients who reached the Fontan stage, the types of Fontan operation influenced long-term survival regardless of the surgical route to the Fontan operation. In atriopulmonary Fontan procedure ($n = 65$), survival rates at 1 year, 5 years, and 10 years were 70.5 ± 5.8%, 65.5 ± 6.1%, and 60.3 ± 6.3%, respectively, while these figures were 88.8 ± 2.7%, 88.0% ± 2.8%, and 88.0 ± 2.8% for the lateral tunnel Fontan procedure ($n = 136$, $P = 0.0001$) (Fig. 3). The survival rates of patients that received the extracardiac Fontan procedure ($n = 28$) were 83.3 ± 7.6%, 74.6 ± 9.0%, and 74.6 ± 9.0% (8-year survival rate with a maximum

Table 3
Survival rate of various groups

	Survival rates (%)			*P*-value
	n	5 YSR	10 YSR	
Total survival	405	64.9 ± 2.6	60.1 ± 2.8	0.27
Shunt group	156	62.5 ± 4.3	57.0 ± 4.5	
PAB group	82	55.6 ± 6.1	47.4 ± 7.5	0.55
BCPS (age at op. ≤4 months)	6	66.7 ± 19.3	66.7 ± 19.3	
BCPS (age at op. >4 months)	119	74.2 ± 4.4	74.2 ± 4.4	0.64
Initial operation in early period (1988–1991)	138	64.0 ± 4.3	57.5 ± 4.4	
Initial operation in late period (1992–2000)	267	66.1 ± 3.1	63.1 ± 3.5	0.24
Staged Fontan (2) + (3)	60	84.1 ± 5.2		
Primary Fontan (1) + (4)	169	78.4 ± 3.3	74.4 ± 3.7	n.s.
(1) (Shunt or PAB) → Fontan	81	77.6 ± 4.8	75.5 ± 5.1	
(2) (Shunt or PAB) → BCPS → Fontan	29	92.6 ± 5.0	92.6 ± 5.0	
(3) BCPS → Fontan	31	78.0 ± 8.0		
(4) Direct Fontan (without palliation & BCPS)	88	78.8 ± 4.4	73.7 ± 5.1	

YSR, year survival rate; PAB, pulmonary artery banding; BCPS, bilateral cavopulmonary shunt.

Table 4
Distribution of risk variables for survival in BCPS and non-BCPS group

	BCPS group ($n = 125$)	Non-BCPS group ($n = 190$)	Pearson chi-square
With risk factors (IAA, TAPVC, PV obstruction)	13	27	$P = 0.32$
Complete AVSD typed lesion	17	24	$P = 0.80$

IAA, interruption of aortic arch; TAPVC, total anomalous pulmonary venous connection; PV, pulmonary vein; AVSD, atrioventricular septal defect.

Table 5
Cross table showing different mortality rates before Fontan operation between non-BCPS and BCPS group

	Alive or dead before Fontan operation		Total	Lost to follow-up
	Alive	Dead		
Non-BCPS group (mortality rate before Fontan operation)	105	74 (41.3%)	179	13
BCPS group (mortality rate before Fontan operation)	103	20 (16.3%)	123	2
Total	208	94	302	15

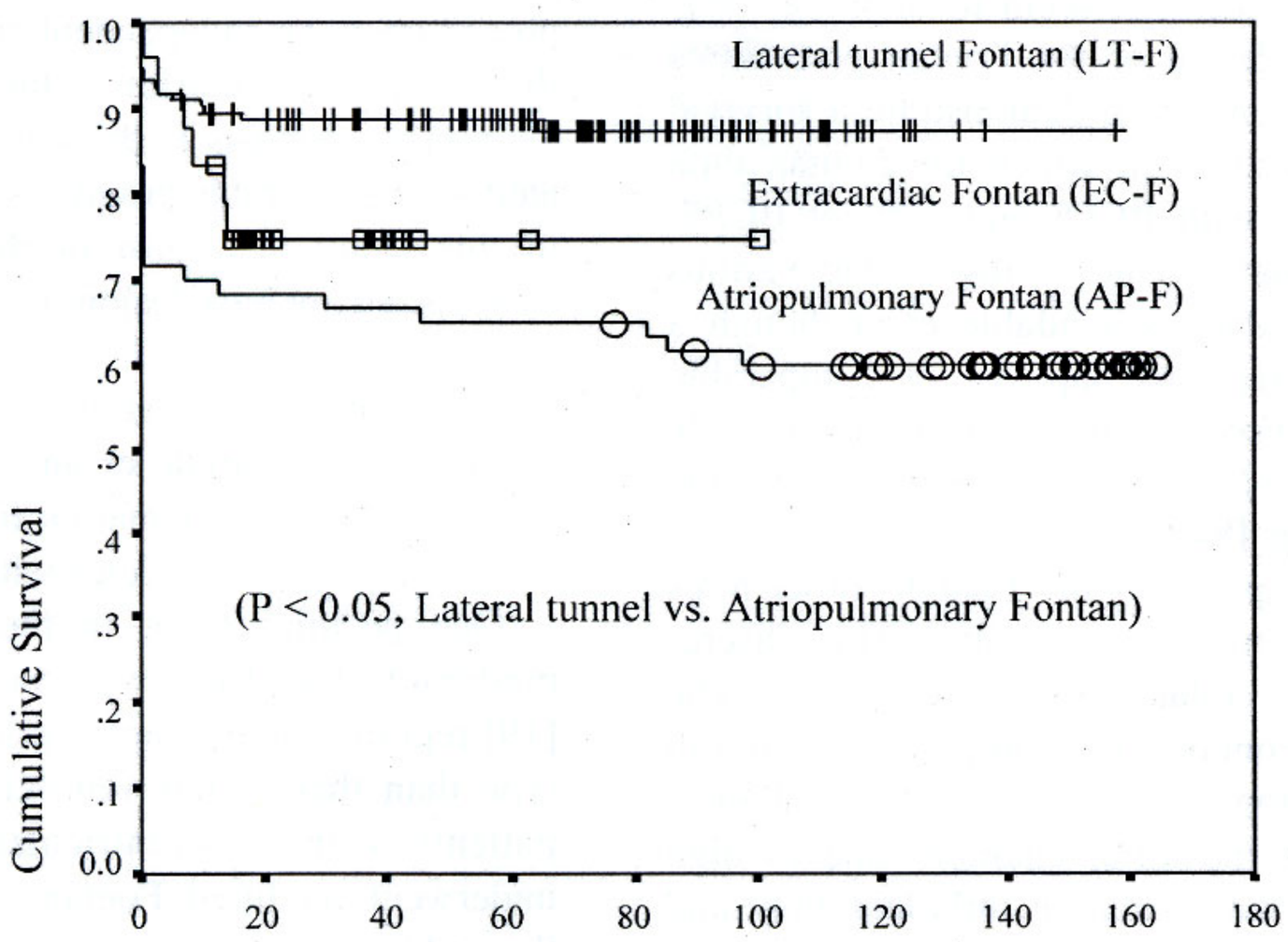
follow-up period of 100.3 months), respectively, without any significant difference compared with the other methods. In patients that received the lateral tunnel Fontan procedure, early mortality was 7.5% in the fenestrated group vs. 6.5% in non-fenestrated group, and 10-year survival rates were 85.0 ± 5.7 vs. $88.2 \pm 3.6\%$, respectively ($P > 0.05$). In patients that received the extracardiac Fontan procedure, there was no early mortality.

4. Discussion

This study focused on the long-term outcomes of various complex cardiac anomalies associated with single ventricle physiology, which requires a number of palliative procedures. It has been hypothesized that the problems in the pre-Fontan stage like variety of staged approaches and palliative procedures before the Fontan operation as well as anatomical risk factors exert an influence on the long-term outcome of the functional single ventricle.

Though many reports have shown excellent long-term results of the modified Fontan procedures, few have demonstrated the long-term course of the functional single ventricles. In our study, the 10-year survival rate of patients with lateral tunnel Fontan operation was 86.8%, while overall survival was only 60.1%. These figures show that a number of problems need to be solved in the pre-Fontan stages to improve long-term survival. Some of the problems may result from the inherent morphologic risk factors, and others from the surgical strategies used, surgical timing related obstacles, or initial learning periods.

The surgical goal to treat the patients with single ventricle physiology is reaching the Fontan type operation and establishing better long-term survival, and allowing patients a better quality of life. The present study demonstrates that actuarial mortality before the Fontan operation was lower in the group in which the Fontan procedure was finished via BCPS than in the group finished without BCPS (16.3 vs. 41.3%, $P < 0.001$). This means that the various surgical approaches or the different pathways to the Fontan procedure can affect the probability of



	# patients at risk									
LT-F	136	112	102	82	48	25	7	1		
EC-F	28	10	5	2	1					
AP-F	65	46	43	40	38	37	34	28	20	4

Fig. 3. Long-term survivals of the modified Fontan groups. Starting point of the follow-up duration: from the time of modified Fontan operation.

reaching the Fontan type operation. Though it is true that in this era of surgery, anatomical risk factors may affect the outcome of the above two groups, there were no significant difference in number of major risk factors including TAPVC, PV obstruction, interruption of aortic arch, complete AVSD typed lesion between the non-BCPS and the BCPS group in this study, and the study period was not a major confounding factor affecting the long-term survival rates after birth. Though not indicated in the table, a multivariate analysis for mortality before the Fontan operation using logistic regression with stepwise entered variables of the above four risk factors, study periods, and BCPS procedure, showed that BCPS was an independent favorable factor ($P < 0.001$, odds ratio = 0.33).

Masuda and colleagues [8] reported a 12.2% mortality rate (ten patients died among 82 bidirectional Glenn shunt patients) before the completion of the Fontan procedure. The lower mortality and the higher probability to complete the Fontan procedure of the BCPS group may be explained by the results of Masuda et al. In their high-risk patients, a bidirectional Glenn shunt resulted in a significant decrease in the mean pulmonary arterial pressure and ventricular end-diastolic pressure, which means a reduction in the pulmonary blood flow and ventricular volume without any progression of arterial oxygen desaturation. In the future, a greater number of high-risk patients will meet the selection criteria of the Fontan procedure as risk will be reduced.

Despite the higher probability of proceeding to Fontan, staged Fontan via BCPS does not seem to improve long-term survival after completion of the Fontan procedures because our results were unable to demonstrate a survival difference between the groups in which the Fontan type operation was completed with BCPS and without BCPS. In terms of long-term survival according to the various staged approaches, little data is available even though a number of series have reported superior or comparable results for the staged Fontan operation with an early mortality of less than 8% compared with that of the primary Fontan operation [8–11].

In our study, the types of the Fontan procedures showed different long-term survival rates. This discrepancy is believed to be related to many factors. The atriopulmonary type of Fontan operation is also limited in terms of maintaining long-term functional excellence. Progressive dilatation of the atrial chamber due to high central venous pressure may result in turbulent flow and late arrhythmias. The introduction of the lateral tunnel procedure theoretically and practically solved this turbulent flow related problem by allowing laminar flow along the Fontan pathway. The better long-term result observed for the lateral tunnel Fontan procedure as compared to atriopulmonary anastomosis can be explained on the basis of this hemodynamic superiority. The extracardiac Fontan procedure failed to show a significantly better outcome than atriopulmonary anastomosis, perhaps due to the relatively short follow-up period and the small number of patients. According to the recent report by Tokunaga and colleagues [12], the actuarial survival rate of the patients who underwent the extracardiac conduit Fontan was 93.9% at 5 years, which is similar to that of the lateral tunnel TCPC group, but the event-free rate of the extracardiac conduit Fontan was significantly superior to that of the lateral tunnel TCPC group (93.0 vs. 82.7% at 5 years, $P = 0.037$). However, the extracardiac conduit Fontan operation needs a longer-term follow-up to demonstrate its long-term hemodynamic superiority and better event-free rate. In the present study, fenestration was not found to be a determining factor of long-term outcome in either the lateral tunnel or extracardiac Fontan groups in spite of the fact that other report [13] found that baffle fenestration proved to improve short-term outcome in standard-risk patients by decreasing pleural drainage, length of hospital stay, and the need for additional postoperative procedures without contribution to the occurrence of systemic thromboembolism [13,14].

Some risk factors of mortality and morbidity have been identified for optimizing patient selection for the Fontan procedure [4,10,15–18]. In the present study, patient-related risk factors were limited to morphologic factors. TAPVC, PV obstruction and interruption of the aortic arch were found to be major morphologic risk factors of mortality. Aortic stenosis and coarctation of the aorta were risk factors only in univariate analysis. Stamm et al. [4] noted that coarctation repair prior to the Fontan procedure is an independent risk factor for late failure of the Fontan circulation. Our study demonstrated that pulmonary stenosis is the only independent factor associated with a favorable prognosis for survival. It was observed in this study that most of the patients with pulmonary stenosis did not have lesions causing obstruction of systemic outflow, including aortic stenosis, coarctation of the aorta, or interrupted aortic arch, which were found to be risk factors in multivariate or univariate analysis in this study. This may be the explanation for the good prognosis of the functional single ventricles with pulmonary stenosis.

One peculiar thing in the present study is that the predominant ventricle type was a right ventricle. Imai et al. [19] reported more a univentricular heart of right ventricle type than that of left ventricle type (31 vs. 14%) in the patients with atrioventricular valve regurgitation who underwent modified Fontan procedure. Predominancy of the right ventricle type may be a tendency in the Oriental population, which has not been proved clearly.

In conclusion, there are still problems that need to be solved in the pre-Fontan stage to improve the overall survival. The role of BCPS was not to contribute to the longer survival after Fontan operation, but to lower mortality in the pre-Fontan stage, which can offer a higher probability to proceed to the Fontan procedure successfully.

References

[1] de Leval MR, Kilner P, Gewilling M, Bull C. Total cavopulmonary connection: a logical alternative to atriopulmonary connection for complex Fontan operations – experimental studies and early clinical experience. J Thorac Cardiovasc Surg 1988;96:682–95.

[2] Jonas RA, Castaneda AR. Modified Fontan procedure: atrial baffle and systemic to pulmonary artery anastomotic techniques. J Card Surg 1988;3:91–6.

[3] Marcelletti C, Corno A, Giannico S, Marino B. Inferior vena cava-pulmonary artery extracardiac conduit: a new form of right heart bypass. J Thorac Cardiovasc Surg 1990;100:228–32.

[4] Stamm C, Friehs I, Mayer JE, Zurakowski D, Triedman JK, Moran AM, Walsh EP, Lock JE, Jonas RA, ddel Nido PJ. Long-term results of the lateral tunnel Fontan operation. J Thorac Cardiovasc Surg 2001; 121:28–41.

[5] Gentles TL, Mayer JE, Gauvreau K, Newburger JW, Lock JE, Kupferschmid JP, Burnett J, Jonas RA, Castaneda AR, Wernovsky G. Fontan operation in five hundred consecutive patients: factors influencing early and late outcome. J Thorac Cardiovasc Surg 1997; 114:376–91.

[6] Amodeo A, Galletti L, Marianeschi S, Picardo S, Giannico S, Di Renzi P, Marcelletti C. Extracardiac Fontan operation for complex cardiac anomalies: seven years' experience. J Thorac Cardiovasc Surg 1997;114:1020–30.

[7] Petrossian E, Reddy VM, McElhinney DB, Akkersdijk GP, Moore P, Parry AJ, Thompson LD, Hanley FL. Early results of the extracardiac conduit Fontan operation. J Thorac Cardiovasc Surg 1999;117: 688–96.

[8] Masuda M, Kado H, Shiokawa Y, Fukae K, Suzuki M, Murakami E, Yasui H. Clinical results of the staged Fontan procedure in high-risk patients. Ann Thorac Surg 1998;65:1721–5.

[9] Norwood WI, Jacobs ML. Fontan's procedure in two stages. Am J Surg 1993;166:548–51.

[10] Pridijan AK, Mendelsohn AM, Lupinetti FM, Beekman 3rd RH, Dick 2nd M, Serwer G, Bove EL. Usefulness of the bidirectional Glenn procedure as staged reconstruction for the functional single ventricle. Am J Cardiol 1993;71:959–62.

[11] Hawkins JA, Shaddy RE, Day RW, Sturtevant JE, Orsmond GS, McGough EC. Mid-term results after bidirectional cavopulmonary shunts. Ann Thorac Surg 1993;56:833–7.

[12] Tokunaga S, Kado H, Imoto Y, Masuda M, Shiokawa Y, Fukae K, Fusazaki N, Ishikawa S, Yasui H. Total cavopulmonary connection with an extracardiac conduit: experience with 100 patients. Ann Thorac Surg 2002;73:76–80.

[13] Lemler MS, Scott WA, Leonard SR, Stromberg D, Ramaciotti C. Fenestration improves clinical outcomes of the Fontan procedure: a prospective, randomized study. Circulation 2002; 105:207–12.

[14] Airan B, Sharma R, Kumar S, Mohanty SR, Bhan A, Kumar U, Juneja R, Kothari SS, Saxena A, Venugopal P. Univentricular repair: is routine fenestration justified? Ann Thorac Surg. 2000;69: 1900–6.

[15] Fishberger SB, Wernovsky G, Gentles TL, Gauvreau K, Burnett J, Mayer JE, Walsh EP. Factors that influence the development of atrial flutter after the Fontan operation. J Thorac Cardiovasc Surg 1997;113: 80–6.

[16] Gentles TL, Gauvreau K, Mayer Jr JE, Fishberger SB, Burnett J, Colan SD, Newburger JW, Wernovsky G. Functional outcome after the Fontan operation: factors influencing late morbidity. J Thorac Cardiovasc Surg 1997;114:392–403.

[17] Gelatt M, Hamilton RM, McCrindle BW, Gow RM, Williams WG, Trusler GA, Freedom RM. Risk factors for atrial tachyarrhythmias after the Fontan operation. J Am Coll Cardiol 1994;24:1735–41.

[18] Knott-Craig CJ, Danielson GK, Schaff HV, Puga FJ, Weaver AL, Driscoll DD. The modified Fontan operation: an analysis of risk factors for early postoperative death or takedown in 702 consecutive patients from one institution. J Thorac Cardiovasc Surg 1995;109: 1237–43.

[19] Imai Y, Takanashi Y, Hoshino S, Terada M, Aoki M, Ohta J. Modified Fontan procedure in ninety-nine cases of atrioventricular valve regurgitation. J Thorac Cardiovasc Surg 1997;113:262–9.