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ORIGINAL ARTICLE

Midterm results and risk factors of functional single ventricles with extracardiac total anomalous pulmonary venous connection

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Abstract

Objectives To evaluate the clinical outcomes of surgical repair of an extracardiac total anomalous pulmonary venous connection (TAPVC) in a functional single-ventricle (f-SV) strategy.

Methods This was a retrospective analysis of 48 consecutive cases of extracardiac TAPVC repair, from 1998 to 2015. Demographic and clinical variables were as follows: median age, 24 (range 0–744) days; median weight, 3.1 (range 2.0–9.6) kg; type of TAPVC—supracardiac, 21 patients, infracardiac, 8, and mixed, 19; right atrial isomerism, 45 patients; pulmonary atresia, 24 patients; and obstructed TAPVC, 30 patients. Concomitant procedures included systemic-to-pulmonary shunting in 15 patients, pulmonary artery banding in 12, ventricle-to-pulmonary artery shunting in 2, a Norwood procedure in one, a bidirectional Glenn procedure in 16, and a Fontan procedure in 1.

Results The 1- and 5-year cumulative survival rates were 66.0% and 58.0%, respectively. Of the 28 survivors, 22 (78.6%) underwent Fontan completion and 4 (14.3%) a bidirectional Glenn procedure, and 1 (3.6%) was awaiting a bidirectional Glenn procedure. Recurrent pulmonary venous stenosis (PVS) was observed in 17 patients, with a 1- and 5-year rates of freedom from recurrent PVS 59.8% and 53.5%, respectively. Of the 20 post-operative deaths, only 4 were PVS related. Only pre-operative pulmonary atresia was identified as an independent risk factor of mortality after TAPVC repair.

Conclusions The midterm surgical outcomes of f-SV with extracardiac TAPVC were acceptable. Moreover, among survivors, Fontan completion can be sufficiently expected. Further improvement, with the development of a comprehensive treatment strategy, is required for this patient group.

Keywords Total anomalous pulmonary venous connection · Single ventricle · Pulmonary venous obstruction

Introduction

Since first being reported in 1951 [1], surgical treatment outcomes for a total anomalous pulmonary venous connection (TAPVC) have improved dramatically. However, in a functional single-ventricle (f-SV) strategy, TAPVC treatment is still challenging, due to the complexity of hemodynamic control and concomitant disease, including extracardiac malformations. Moreover, with an extracardiac TAPVC, surgical repair is often required during the neonatal phase due to pulmonary venous obstruction. Thus, the 5-year survival rate after extracardiac TAPVC repair with a f-SV remains low, ranging between 31.3 and 55.0% [2–6], with an early death rate of 54.0% identified in our previous review of 26 cases [5]. Here, we report on the early and midterm outcomes of patients with a f-SV and extracardiac TAPVC, and identify the risk factors for mortality and post-operative recurrence of pulmonary venous stenosis (PVS) in this complicated patient group.

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Methods

Patients

Our retrospective analysis was based on the data of 48 consecutive patients who underwent extracardiac TAPVC repair, with a f-SV strategy, between January 1998 and December 2015, at Mount Fuji Shizuoka Children's Hospital. The study was approved by the ethics committee of Mount Fuji Shizuoka Children's Hospital.

Patients' medical and surgical reports and peri-operative echocardiogram and catheterization examinations were reviewed. The mean duration of follow-up was 58.6 ± 61.1 months, with all patients being followed up. The study was divided into the following two periods for analysis: the period between 1998 and 2006, and the period between 2007 and 2015.

Surgical procedures

TAPVC repairs were performed via a median sternotomy, with cardiopulmonary bypass introduced under moderate hypothermia. Cross-clamping and crystalloid cardioplegia were used in all cases for myocardial protection. As necessary, short and intermittent circulatory arrest were used for precise anastomosis. Intervention for pulmonary veins was based on appropriate dissection and minimal manipulation. If the site of anastomosis was the wall of the right atrium, sectioning through the thick muscle layer of the right atrial wall was needed. Until 2010, anastomosis between the common pulmonary vein and the thin intimal layer of the atrial wall was performed using delicate continuous sutures (8–0 non-absorbable monofilaments). After 2010, we used a primary sutureless technique for selected patients with mixed-type TAPVC.

Initial palliation for TAPVC repair included systemic-to-pulmonary shunting in 15 patients, ventricle-to-pulmonary shunting in 2, pulmonary artery banding in 12, a Norwood procedure in 1, and a bidirectional Glenn procedure in 16. In only one case was an initial Fontan procedure achieved with TAPVC repair.

Evaluation of the pulmonary venous stenosis

PVS was evaluated using echocardiography to assess the flow velocity within individual pulmonary veins (PVs). PVS was defined by a flow velocity ≥ 1.5 m/s which did not return to baseline, despite the stenosis site being limited to only one branch of the pulmonary veins.

Statistical analysis

Data were described using mean ± standard deviation. Statistical comparisons were performed using unpaired, two-tailed, Student's *t* tests, with a probability value < 0.05 indicating significance. The actuarial survival rate and freedom from post-operative PVS were evaluated using a Kaplan–Meier survival analysis, with between group differences evaluated using a log-rank test.

Primary outcomes of the study were mortality and post-operative recurrent PVS, with the following variables included in uni- and multivariable analyses to identify predictive variables: age at the time of TAPVC repair; body weight; sex; gestational age; type of TAPVC; pre-operative pulmonary atresia/stenosis; pre-operative pulmonary hypertension; pre-operative or post-operative PVS; pre-operative or post-operative atrioventricular valve regurgitation (AVVR); TAPVC repair with a systemic-to-pulmonary shunt or a ventricle-to-pulmonary shunt; abdominal visceral anomaly requiring surgical intervention; and era (early period, 1998–2006, or late period, 2007–2015). These clinical variables were analyzed using the log-rank test for univariate analyses, with variables having a *P* value < 0.1 and with < 5% missing data entered in the multivariate analysis, using the likelihood ratio test in a Cox proportional-hazard regression model. Variables with a *P* value < 0.05 on multivariate analysis were considered as independent predictors of clinical outcomes.

Results

Pre-operative data are presented in Table 1, with salient features summarized as follows. The sample group included 26 males (54.2%), with 54.2% of procedures (26 of the 48 cases) performed in the neonatal period, and 93.8% (45 of 48 cases) having a diagnosis of right atrial isomerism (RAI). Of the remaining three patients, two had a double-outlet right ventricle (DORV) and the other, hypoplastic left heart syndrome. The median age at the initial surgical intervention was 24 (range 0–744) days, with a median weight of 3.1 (range 2.0–9.6) kg. The type of extracardiac TAPVC was supracardiac in 21 patients, infracardiac in 8, and mixed in 19. The TAPVC in the mixed group included a combination of two different supracardiac types in eight, supracardiac and cardiac in six, and supra- and infracardiac in five.

Actuarial survival

The 1- and 5-year survival rates are 66.0% and 58.0%, respectively, across all patients (Fig. 1a). Of 31 patients

Table 1 Patients' characteristics

Type of TAPVC	*n*	New born	RAI	Pulmonary outflow obstruction		Obstructed TAPVC		Concomitant procedure					Survivors		
				Atresia	Stenosis	(+)	(−)	SP shunt	VPS	PAB	Norwood	BDG	Fontan	Waiting	BDG	Fontan
Supracardiac	21	15	19	11	6	17	4	7	1	6	1	6	0	0	3	9
Infracardiac	8	4	8	6	1	5	3	6	0	1	0	0	0	0	1	4
Mixed	19	7	18	7	10	8	11	2	1	5	0	10	1	1	2	8
Total	48	26	45	24	17	30	18	15	2	12	1	16	1	1	6	21

TAPVC total anomalous pulmonary venous connection, *n* number, *RAI* right atrial isomerism, *SP shunt* systemic to pulmonary shunt, *VPS* ventricular to pulmonary shunt, *PAB* pulmonary artery banding, *BDG* bidirectional Glenn

who underwent first-stage palliation with TAPVC repair, including 25 neonates, there were ten early (in-hospital) deaths and two late deaths (Fig. 2). Sixteen patients underwent a bidirectional Glenn procedure with TAPVC repair, with two early deaths and one late death in this group. Only one patient who underwent a Fontan procedure in combination with TAPVC repair at one time survived.

The Kaplan–Meier survival analysis after TAPVC repair, according to the type of TAPVC, is shown in Fig. 1b. The survival rate of supracardiac TAPVC was 66.7% at 1 year and 55.0% at 5 years, compared to 75.0% and 56.3%, respectively, for patients with an infracardiac TAPVC and 61.6%, at both 1 and 5 years, for patients with mixed-type TAPVC. The survival rate was not significantly different across the different TAPVC types. With regard to time period, the overall 1- and 5-year survival rates in the early period were 54.6% and 50.0%, respectively, compared to 76.0% and 64.8%, respectively, in the late period ($p = 0.298$; Fig. 1c).

Freedom from recurrent pulmonary venous stenosis

The 1- and 5-year rates of freedom from PVS after TAPVC repair were 59.8% and 53.5%, respectively (Fig. 3a). Of the 48 cases included in the study, 30 presented with PVS before TAPVC repair, with 25 of these treated with first-stage palliation and the other five undergoing a bidirectional Glenn procedure. Of these 30 cases, post-operative PVS recurred in 12 (40.0%), of which 11 (44.0%) had been treated with first-stage palliation before TAPVC repair, with PVS recurrence identified in only one of the five cases (20.0%) treated using a bidirectional Glenn procedure before TAPVC repair. Of the 18 patients with no pre-operative PVS before TAPVC repair, post-operative PVS occurred in four cases (27.8%), including two of the six cases (33.3%) treated using first-stage palliation, and two of the 11 (18.1%) treated using a bidirectional Glenn procedure.

The PVS-free survival rate after TAPVC repair, according to the type of TAPVC, is shown in Fig. 3b. The PVS-free rate for supracardiac TAPVC was 72.4% at 1 year and 64.3% at 5 years, with rates of 66.7% and 50.0%, respectively, for infracardiac TAPVC and 44.3% at both 1 and 5 years for mixed-type TAPVC. The PVS-free survival rate was, therefore, not significantly different across the three types of TAPVC. With regard to the period of analysis, the PVS-free survival rate at 1 and 5 years was 61.7% and 54.5%, respectively, in the early period and 58.6% and 52.7%, respectively, in the late period. Therefore, PVS-free survival, at 1 and 5 years, was not significantly different between the periods (Fig. 3c).

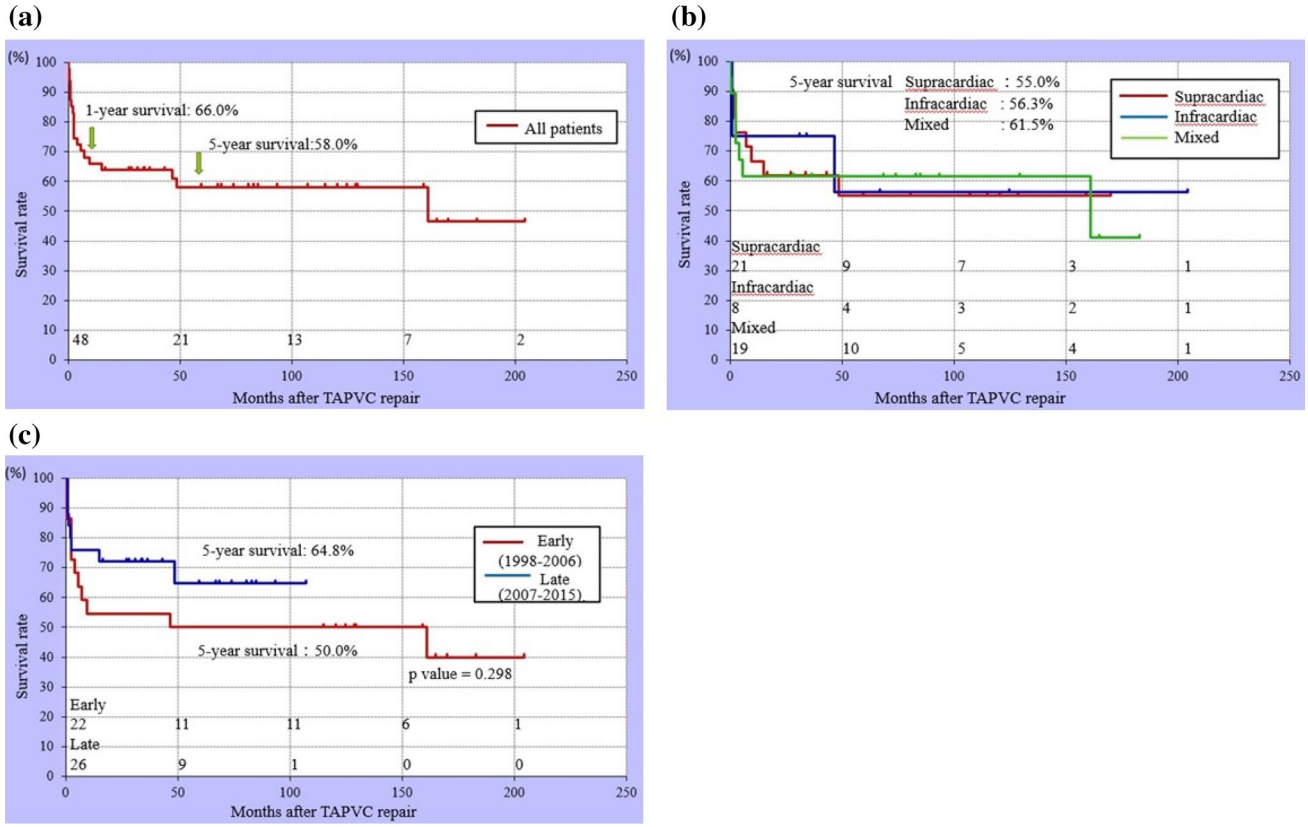
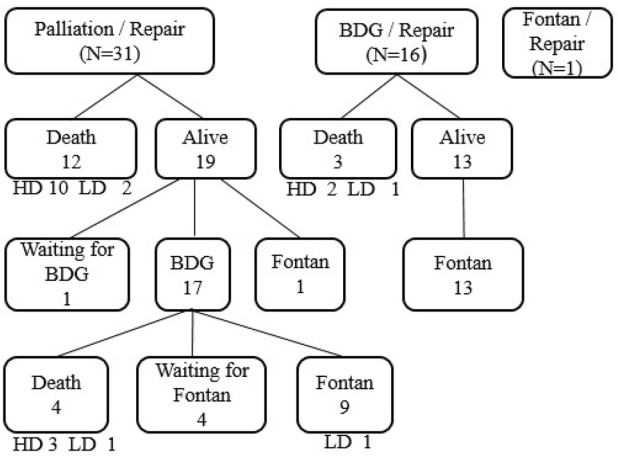


Fig. 1 Actuarial survival rate after TAPVC repair, estimated using the Kaplan–Meier method: **a** all patients, **b** type of TAPVC, and **c** operative period



TAPVC: total anomalous pulmonary venous connection, N: number, BDG: bidirectional Glenn, HD: hospital death, LD: late death

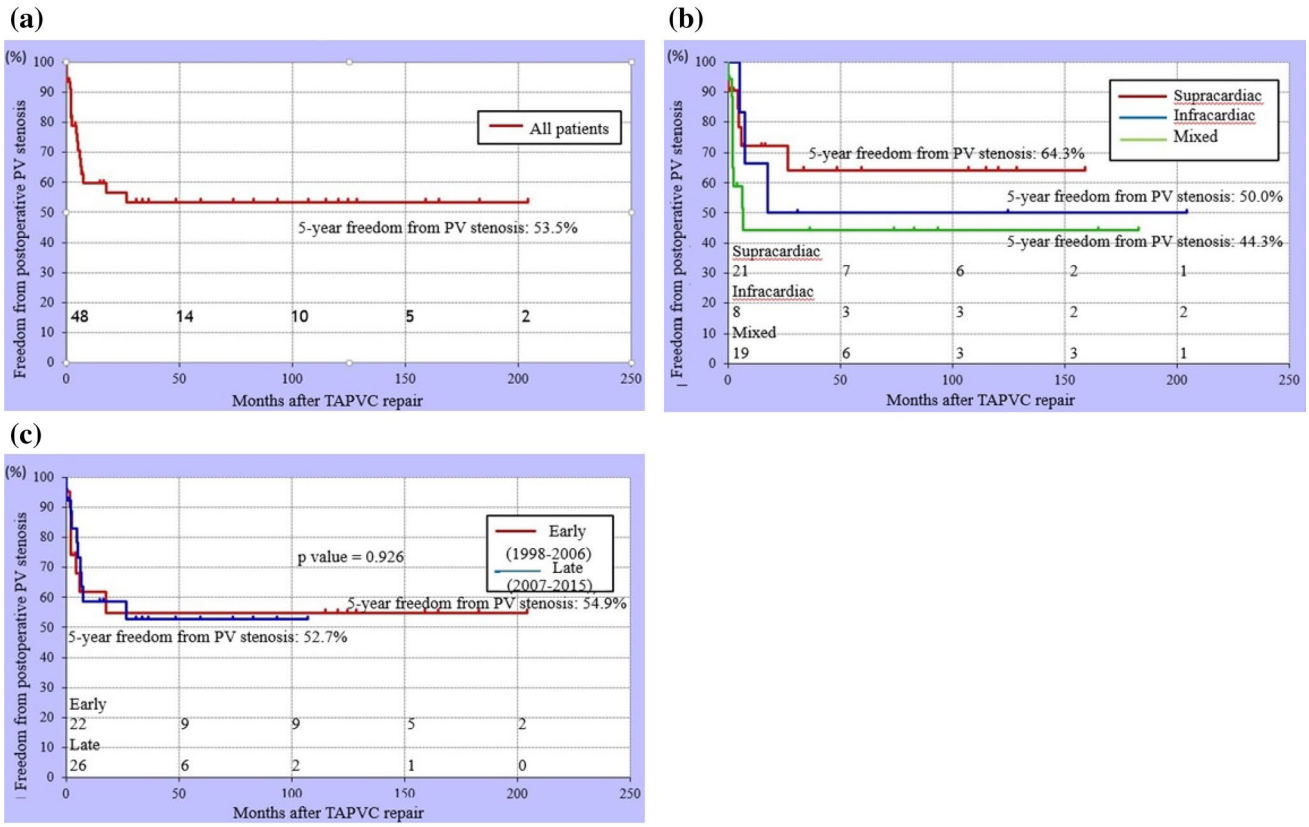
Fig. 2 Outcomes of patients with single ventricle after TAPVC repair

Death due to recurrent pulmonary venous stenosis

Among the 48 cases of TAPVC repair, 20 (41.7%) patients died after surgical repair, 6 of whom developed recurrent PVS, leading to death in four of these cases due to pulmonary bleeding. Among these latter four cases, three required frequent re-PVS release after TAPVC repair, including intrapulmonary venous stenting. The other two patients who experienced recurrent PVS died due to causes not related to post-repair PVS, one due to pulmonary bleeding not associated with post-repair PVS and the other due to protein-losing enteropathy, late after total Fontan completion. The cause of death in the 14 patients who did not have post-repair PVS included: heart failure in 5, respiratory complication in 4, systemic-to-pulmonary shunt occlusion in 2, infection in 2, and gastrointestinal perforation in 1.

Fontan completion of survival case

Among all cases in this study, 23 (47.9%) patients underwent Fontan completion, with no early deaths among these cases (Fig. 2). The median age at the time of Fontan



PV: pulmonary vein, TAPVC: total anomalous pulmonary venous connection

Fig. 3 Freedom from PV stenosis after TAPVC repair, estimated using the Kaplan–Meier method: **a** all patients, **b** type of TAPVC and **c** operative period

completion was 22 (range 14–44) months, and the median body weight was 9.3 (range 8.1–14.0) kg. Fenestrations were placed in four patients, of which, a fenestration was placed late after Fontan completion because of protein-losing enteropathy (PLE), in one patient. The death of this patient due to late PLE was the only cause of late death case after Fontan completion. Among the 28 patients alive at the end of this study, the Fontan completion rate was 78.6%, with follow-up cardiac catheterization required in 22 of these patients. For patients who underwent several follow-up catheterizations, the latest data were evaluated. The median interval between Fontan completion and the latest follow-up cardiac catheterization was 50 (range 11–161) months. The hemodynamic variables were as follows: central venous pressure, 11.2 ± 2.4 mmHg; cardiac index, 3.5 ± 0.8 L/min/m^2; ejection fraction, $53.4 \pm 8.0\%$; and O_2 saturation, $94.0\% \pm 2.0\%$. At the latest follow-up, all patients had a New York Heart Association class I.

Uni- and multivariable analyses of factors predictive of death and recurrent pulmonary venous stenosis

Significant risk factors of mortality after TAPVC repair with a f-SV are described in Table 2a. Univariate risk factors for mortality included: pulmonary atresia, TAPVC repair with systemic-to-pulmonary shunt, TAPVC repair with a bidirectional Glenn procedure, age at the time of TAPVC repair, and body weight at the time of TAPVC repair. Of these, only pulmonary atresia was retained as an independent risk factor of mortality after TAPVC repair on multivariate analysis ($p = 0.036$, hazard ratio 5.21; 95% confidence interval (CI) 1.12–22.57).

Significant risk factors for PVS after TAPVC repair are described in Table 2b. Univariate risk factors included atrioventricular regurgitation requiring intervention, TAPVC repair with pulmonary artery banding, TAPVC repair using a bidirectional Glenn procedure, age at the time of TAPVC

Table 2 Risk factors for mortality or recurrent pulmonary venous stenosis from multivariate analysis

(a) Mortality						
Variable	Alive $n = 28$	Death $n = 20$	Univariate p value	Cox multivariate analysis		
				Hazard ratio	95% CI	p value
Pulmonary atresia	10 (35.7%)	14 (70.0%)	0.012	5.21	1.12–22.57	0.036
TAPVC repair with SP	5 (17.9%)	10 (50.0%)	0.018	0.46	0.10–2.46	0.343
TAPVC repair with BDG	13 (46.4%)	3 (15.0%)	0.018	0.45	0.05–4.53	0.450
Age at TAPVC repair (days)	133.07 ± 166.67	49.00 ± 77.60	0.014	1.00	0.98–1.01	0.674
BW at TAPVC repair (kg)	4.78 ± 2.22	3.25 ± 1.33	0.004	0.61	0.22–1.49	0.283
(b) Recurrent pulmonary venous stenosis						
Variable	PVS + $n = 28$	PVS − $n = 20$	Univariate p value	Cox multivariate analysis		
				Hazard ratio	95% CI	p value
AVVR required intervention	6 (21.4%)	12 (60.0%)	0.003	2.42	0.77–8.04	0.132
TAPVC repair with PAB	3 (10.7%)	9 (45.0%)	0.003	2.11	0.55–9.44	0.295
TAPVC repair with BDG	13 (46.4%)	3 (15.0%)	0.014	0.65	0.07–6.81	0.696
Age at TAPVC repair (days)	127.90 ± 166.70	48.28 ± 64.05	0.008	1.00	0.98–1.01	0.571
BW at TAPVC repair (kg)	4.55 ± 2.27	3.45 ± 1.35	0.011	1.06	0.37–3.02	0.919

n number, *CI* confidence interval, *TAPVC* total anomalous pulmonary venous connection, *SP shunt* systemic to pulmonary shunt, *BDG* bidirectional Glenn, *BW* body weight, *PVS* pulmonary venous stenosis, *AVVR* atrioventricular valve regurgitation, *PAB* pulmonary artery banding

repair, and body weight at the time of TAPVC repair. On multivariate analysis, none of these were retained as an independent risk factor of PVS after TAPVC repair.

Discussion

Our study demonstrated that the management of infants with a f-SV and extracardiac TAPVC continues to be challenging, with 12 early (in-hospital) deaths and three late deaths after TAPVC repair among our 48 cases. Moreover, the survival rates at 1 and 5 years were 66.0% and 58.0%, respectively, and the findings are consistent with previous studies reporting a 5-year survival rate of 31.3–55.0% [2–6]. In the vast majority of patients (93.8%) in our cases series, TAPVC was associated with right atrial isomerism. Generally, patients with right atrial isomerism are at risk of atrioventricular valve regurgitation, cardiac dysfunction, arrhythmia, infection, and abdominal visceral anomaly [4, 7–9]. Alsoufi et al. reported heterotaxy syndrome as an independent risk factor associated with increased morbidity and mortality after single-ventricle palliation [7]. Additionally, in our case series, 26 patients were neonates at the time of TAPVC repair (54.2%), 24 had pulmonary atresia (50.0%), 30 obstructed TAPVC (62.5%), and 8 an infracardiac TAPVC (16.7%), with 19 presenting with mixed TAPVC (39.6%). Therefore, we propose that these complicated conditions are associated with a high rate of mortality and poor prognosis after TAPVC repair.

We previously reported mixed-type TAPVC as a significant risk factor for mortality [5]. However, there have been improvements in the surgical management of mixed-type TAPVC due to advancement in technique and accurate preoperative investigation of the pulmonary veins. In fact, in this study, we did not identify mixed-type TAPVC as an independent risk factor of mortality after TAPVC repair. However, we did identify pulmonary atresia as an independent risk factor of mortality for TAPVC repair using a f-SV strategy, indicative of the continued difficulty in establishing an appropriate regulation of pulmonary blood flow according to patients' cardiac function and atrioventricular valve function, as proposed in previous reports [10–12]. In this regard, the degree of peri-operative atrioventricular valve regurgitation should relate to the risk of mortality. However, atrioventricular regurgitation was not associated with an increased risk of mortality in our study, which could be explained, in part, by the small cohort size of our study.

A previous report has emphasized that the timing of TAPVC repair can affect the risk of mortality after repair [4]. Before a bidirectional Glenn procedure, the hemodynamic regulation associated with a f-SV is very unstable. Moreover, many of these patients in our study cohort were neonates or early infants and, thus, had immature and fragile tissue and organ function. Improving the outcomes in this high-risk group requires not only strict peri-operative intensive care to maintain cardiac function, but also rigid regulation of pulmonary blood flow and preservation of atrioventricular valve function. Of note, the mechanisms of

atrioventricular valve regurgitation with a f-SV are complex and multifactorial and can include chronic volume overload, structural atrioventricular valve anomalies, and impaired ventricular function. Several institutions have reported on successful atrioventricular valvuloplasty with a f-SV [13, 14], with successful repair being the key to improving surgical outcome and obtaining good prognosis.

Our surgical approaches were acceptable in avoiding post-operative PVS. We have no data regarding the superiority between our strategy and a primary sutureless repair. However, we did aggressively adopt a primary sutureless technique if there was a risk of stenosis or twisting due to a suture-related helical structural shift between the atrium and pulmonary veins.

Post-operative PVS occurred regardless of the type of TAPVC and the timing of the surgical repair. The 1- and 5-year rates of PVS after TAPVC repair were 59.8% and 53.5%, respectively. Reintervention for PVS was required in 16 cases, with five of these patients having undergone Fontan completion and doing well. Among the 20 cases of post-operative deaths, only four deaths were due to recurrent PVS, with three of these patients requiring multiple reintervention for PVS. All four patients died of pulmonary bleeding resulting from pulmonary hypertension. The association between post-operative PVS and both early and late deaths after TAPVC repair has previously been reported [6]. In our cases series, however, post-operative PVS did not influence hemodynamics to any great extent, with only four deaths due to recurrent PVS; other causes of death, not related to PVS, were more common.

Our Fontan completion rate among survivors was 78.6%, which is relatively high compared to the findings of previous studies [4, 6, 7]. Hemodynamic status after Fontan completion was acceptable in our study group, with a central venous pressure of 11.2 ± 2.4 mmHg, ejection fraction of $53.5 \pm 8.0\%$ and oxygen saturation of $94.0\ \mathrm{W} \pm 2.0\%$. We consider those outcomes to be excellent for this high-risk patient group, especially as a previous study reported significant risk of post-operative PVS after Fontan completion [4]. However, in our study, Fontan circulation was completed even in many cases with post-operative PVS recurrence, with the site of PVS being limited to the pulmonary venous branches. Overall, we consider that if issues of regulation of pulmonary blood flow, atrioventricular function and cardiac function could be solved, Fontan completion would be feasible in this high-risk patient group.

Conclusions

The surgical outcome of f-SV with extracardiac TAPVC was acceptable, with a rate of Fontan completion of almost 80% among survivors. Therefore, we believe that a good prognosis can be expected after successful treatment, even in this complicated patient group. In patients with pulmonary atresia, accurate regulation of pulmonary flow is required during first palliation, as this regulation can significantly affect cardiac and atrioventricular valve functions, which have a significant influence on prognosis. As such, a more comprehensive treatment strategy for hemodynamic control for this patient group is needed.

Compliance with ethical standards

Conflict of interest The authors declare that no conflict of interest exists.

References

1. Muller WH Jr. The surgical treatment of transposition of the pulmonary veins. Ann Surg. 1951;134:683–93.
2. Hancock Friesen CL, Zurakowski D, Thiagarajan RR, Forbess JM, del Nido PJ, Mayer JE, et al. Total anomalous pulmonary venous connection: an analysis of current management strategies in a single institution. Ann Thorac Surg. 2005;79:596–606.
3. Foerster SR, Gauvreau K, McElhinney DB, Geva T. Importance of totally anomalous pulmonary venous connection and postoperative pulmonary vein stenosis in outcomes of heterotaxy syndrome. Pediatr Cardiol. 2008;29:536–44.
4. Hoashi T, Kagisaki K, Oda T, Kitano M, Kurosaki K, Shiraishi I, et al. Long-term results of treatments for functional single ventricle associated with extracardiac type total anomalous pulmonary venous connection. Eur J Cardiothorac Surg. 2013;43:965–70.
5. Nakata T, Fujimoto Y, Hirose K, Osaki M, Tosaka Y, Ide Y, et al. Functional single ventricle with extracardiac total anomalous pulmonary venous connection. Eur J Cardiothorac Surg. 2009;36:49–56.
6. Nakayama Y, Hiramatsu T, Iwata Y, Okamura T, Konuma T, Matsumura G, et al. Surgical results for functional univentricular heart with total anomalous pulmonary venous connection over a 25-year experience. Ann Thorac Surg. 2012;93:606–13.
7. Alsoufi B, McCracken C, Schlosser B, Sachdeva R, Well A, Kogon B, et al. Outcomes of multistage palliation of infants with functional single ventricle and heterotaxy syndrome. J Thorac Cardiovasc Surg. 2016;151:1369–77.
8. Kim SJ, Kim WH, Lim HG, Lee CH, Lee JY. Improving results of the Fontan procedure in patients with heterotaxy syndrome. Ann Thorac Surg. 2006;82:1245–51.
9. Stamm C, Friehs I, Duebener LF, Zurakowski D, Mayer JE Jr, Jonas RA, et al. Improving results of the modified Fontan operation in patients with heterotaxy syndrome. Ann Thorac Surg. 2002;74:1967–77.
10. Honjo O, Atlin CR, Mertens L, Al-Radi OO, Redington AN, Caldarone CA, et al. Atrioventricular valve repair in patients with functional single-ventricle physiology: impact of ventricular and valve function and morphology on survival and reintervention. J Thorac Cardiovasc Surg. 2011;142:326–35.
11. Kotani Y, Chetan D, Atlin CR, Mertens LL, Jegatheeswaran A, Caldarone CA, et al. Longevity and durability of atrioventricular valve repair in single-ventricle patients. Ann Thorac Surg. 2012;94:2061–9.
12. Wong DJ, Iyengar AJ, Wheaton GR, Ramsay JM, Grigg LE, Horton S, et al. Long-term outcomes after atrioventricular valve

operations in patients undergoing single-ventricle palliation. Ann Thorac Surg. 2012;94:606–13.
13. Ando M, Takahashi Y. Edge-to-edge repair of common atrioventricular or tricuspid valve in patients with functionally single ventricle. Ann Thorac Surg. 2007;84:1571–6.
14. Naito Y, Hiramatsu T, Kurosawa H, Agematsu K, Sasoh M, Nakanishi T, et al. Long-term results of modified Fontan operation for single-ventricle patients associated with atrioventricular valve regurgitation. Ann Thorac Surg. 2013;96:211–8.

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