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ORIGINAL ARTICLE

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Risk analysis for patients with a functionally univentricular heart after systemic-to-pulmonary shunt placement

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Key question

What are the risk factors for death of univentricular heart disease after systemic-to-pulmonary shunt placement?

Key finding(s)

Central shunt placement, small body size and complex cardiac anomalies were identified as risk factors for mortality.

Take-home message

A systemic-to-pulmonary shunt procedure carries a high mortality rate when it is performed in small patients with complex cardiac anomalies.

RF for 30-day mortality	OR (95% CI)	*P* value
central shunt	2.94 (1.17-7.36)	0.021

RF for 90-day mortality	OR (95% CI)	*P* value
Body Weight < 2.5 kg	3.58 (1.01-12.67)	0.048
Body Weight 2.5-3.5 kg	2.28 (1.04-4.99)	0.039
preoperative ventilator support	2.75 (1.20-6.30)	0.017
right atrial isomerism	2.38 (1.27-4.44)	0.007
MAPCAs	3.51 (1.07-11.52)	0.038
unbalanced AVSD	2.62 (1.01-6.79)	0.047
concomitant PDA closure	0.34 (0.11-0.98)	0.045

Abstract

OBJECTIVES: To investigate risk factors for mortality after systemic-to-pulmonary (SP) shunt procedures in patients with a functionally univentricular heart using the Japan Cardiovascular Surgery Database registry.

METHODS: Clinical data from 75 domestic institutions were collected. Overall, 812 patients with a functionally univentricular heart who underwent initial SP shunt palliation were eligible for analysis. Patients with pulmonary atresia with an intact ventricular septum and patients with a SP shunt as part of the Norwood procedure were excluded. Risk factors for 30- and 90-day mortalities were analysed using a logistic regression model.

RESULTS: Median age and body weight at SP shunt placement were 41 days and 3.6 kg, respectively. Modified Blalock–Taussig shunt, central shunt and other types of SP shunts were applied in 689 (84.9%), 94 (11.8%) and 30 (3.7%) patients, respectively. Cardiopulmonary bypass was utilized in 410 patients (51%) for 128 min (median, 19–561). There were 411 isolated SP shunt procedures. Median hospital stay was 27 days, and 742 (91.4%) patients were discharged. The 30- and 90-day mortality rates were 3.4% and 6.0%, respectively. Placement of a central shunt was identified as a risk factor for 30-day mortality, while lower body weight, preoperative ventilator support, right atrial isomerism and coexistence of major aortopulmonary collateral arteries and an unbalanced atrioventricular septal defect were identified as risk factors for 90-day mortality.

CONCLUSIONS: SP shunt carries a high mortality rate in patients with a functionally univentricular heart when it is performed in smaller patients with complex cardiac anomalies.

Keywords: Functionally univentricular heart • Systemic-to-pulmonary shunt • Mortality risk

ABBREVIATIONS

BT	Blalock–Taussig
OR	Odds ratios
PA	Pulmonary artery
PDA	Patent ductus arteriosus
RAI	Right atrial isomerism
SP	Systemic-to-pulmonary
TAPVC	Total anomalous pulmonary venous connection
UVH	Univentricular heart

INTRODUCTION

A systemic-to-pulmonary (SP) shunt is an established surgical procedure that continues to be indicated as palliative therapy to deliver a reliable source of pulmonary blood flow in patients with various congenital heart diseases [1, 2]. While primary corrective surgeries are preferential to palliative SP shunt procedures in patients with biventricular physiology, placement of SP shunts in patients with a functionally univentricular heart (UVH) has increased considerably, owing to advances in multistage palliation management strategies [3, 4]. However, the procedure has a relatively high morbidity and mortality, especially when applied to patients with a single ventricle [5–7]. To date, several single- and multicentre studies have investigated the risk factors for mortality after SP shunt procedures.

However, these studies included a relatively small number of patients with a functionally UVH or a prolonged study period with an inconsistent treatment policy [5–7]. In this study, we clarify the present state of SP shunt procedure for patients with functionally UVHs and investigate the risk factors for mortality based on the Japan Cardiovascular Surgery Database registry.

PATIENTS AND METHODS

This study was approved by the ethical committee of Kyoto University (8 October 2019, R2130). Due to the retrospective nature of the data collected, the need for informed consent was waived.

Data collection

Data were collected from the Japan Cardiovascular Surgery Database registry, which collects clinical data from 75 domestic institutions specializing in congenital heart diseases, covering almost all major congenital heart surgery programmes in Japan. Clinical data on preoperative, operative, and postoperative characteristics and survival within 30 and 90 days were available from the database. Inclusion criteria were [1] any type of initial SP shunt procedure performed between 2008 and 2017, regardless of age at the time of the procedure and concomitant cardiac procedures and [2] cardiac diagnosis of a single ventricle with or without double-inlet left ventricle, double-inlet right ventricle, mitral atresia (MA), tricuspid atresia and heterotaxy syndrome. Exclusion criteria were [1] diagnosis of pulmonary atresia with intact ventricular septum and Ebstein's anomaly or [2] SP shunt placement as a part of the Norwood procedure by following reasons. In patients with pulmonary atresia with intact ventricular septum, SP shunt procedure could be performed as initial palliation for one and a half or biventricular repairs [11, 12]. Although those patients should be excluded from our study, it is not possible in our database to distinguish them from patients treated along univentricular repair strategy. Similar situation may occur also in patients with Ebstein's anomaly. With regard to the Norwood procedure, its mortality rate is still higher than that of other congenital heart surgeries [1, 13, 14]. Therefore, results will be strongly affected by the number of Norwood procedures in cohort when included as a variable in the analysis.

Primary end-points

The primary end-points were mortality at 30 days and 90 days after the SP shunt procedure.

Statistical analysis

Categorical variables were expressed as numbers and percentages. Continuous variables were presented as medians with ranges or means with standard deviations. We conducted univariable logistic regressions between outcome and each variable and those results were shown crude odds ratios (OR) with 95% confidence intervals. In the univariable analysis of the outcomes and the candidate risk factors, no adjustment for multiple comparisons was made. This is because this univariable analysis is not intended to confirm the relationship between the candidate outcomes and risk factors. The independent variables examined included cardiac diagnosis, coexisting cardiac anomalies, chromosomal abnormality, early delivery (<37 weeks of gestation), preoperative mechanical ventilator support, age and weight at operation, utilization and duration of cardiopulmonary bypass (CPB) and concomitant operative procedures. Risk factors for 30- and 90-day mortality after the SP shunt procedure were

analysed. To develop each model, we conducted a multivariable logistic regression analysis with the independent variables and a forward–backward stepwise selection method using Akaike's information criterion. We chose the model that minimized Akaike's information criterion to predict the occurrence of the primary end-points. The step Akaike's information criterion function in the Modern Applied Statistics with S (MASS) package of R (R Foundation for Statistical Computing, Vienna, Austria) was used for this procedure. A P-value < 0.05 was considered statistically significant. All statistical analyses were performed using R software version 3.6 and later (R Foundation for Statistical Computing, Vienna, Austria).

RESULTS

Patient demographics

A total of 817 patients met the inclusion criteria for this study. Due to incomplete data input, 5 patients were further excluded, and 812 patients were eligible for analysis. There were 465 (57.3%) male patients. Prior to the SP shunt procedures, 17 surgical interventions were performed in 16 patients: main pulmonary artery (PA) banding in 4 patients, bilateral PA banding in 4 patients, repair of total anomalous pulmonary venous connection (TAPVC) in 2 patients, PA reconstruction in 2 patients, patent ductus arteriosus (PDA) closure in 2 patients and miscellaneous interventions in 3 patients. The median age and body weight at the time of the SP shunt procedure were 41 days (range 0 days–10 years) and 3.6 kg (range 1.0–22.7), respectively. Early delivery was observed in 51 patients (6.3%). Chromosomal abnormalities were identified in 25 (3.1%) patients. There were 278 (34.2%) neonates. The most common fundamental cardiac diagnosis was heterotaxy syndrome (n = 274, 33.7%), followed by single ventricle (n = 208, 25.6%), tricuspid atresia (n = 149, 18.3%), double-inlet right ventricle/double-inlet left ventricle (n = 124, 15.2%) and mitral atresia (n = 57, 7.0%). Of the cardiac anomalies present in the patient population, pulmonary atresia was the most common (n = 261, 32.2%), followed by pulmonary stenosis (n = 127, 15.7%), PA stenosis/discontinuity (n = 127, 15.7%), extracardiac TAPVC (n = 57, 7.0%), unbalanced atrioventricular septal defect (AVSD) (n = 50, 32.3%), cardiac TAPVC (n = 39, 4.8%) and major aortopulmonary collateral arteries (MAPCAs) (n = 24, 3.0%). Prior to the SP shunt procedure, 61 patients (7.5%) had mechanical ventilator support. Patient demographics are summarized in Table 1.

Table 1: Patient characteristics

Total number of cases		812
Male patients		465 (57.3%)
Early delivery (<37 weeks)		51 (6.3%)
Chromosomal abnormality		25 (3.1%)
Preoperative ventilator support		61 (7.5%)
Age[a]		
	<2 weeks	67 (8.3%)
	<1 month	211 (26.0%)
	<3 months	342 (42.1%)
	<6 months	88 (10.8%)
	>6 months	103 (12.7%)
Body weight (kg)		
	<2.5	30 (3.7%)
	2.5–3.5	339 (41.7%)
	3.5–5.0	279 (34.4%)
	>5.0	164 (20.2%)
Fundamental cardiac diagnosis		
	Single ventricle	208 (25.6%)
	Heterotaxy	274 (33.7%)
	DIRV/DILV	124 (15.3%)
	Tricuspid atresia	149 (18.3%)
	Mitral atresia	57 (7.0%)
Coexisting cardiac anomaly		
	Pulmonary atresia	261 (32.1%)
	Pulmonary stenosis	127 (15.6%)
	Pulmonary artery stenosis/discontinuity	127 (15.6%)
	TAPVC (extracardiac)	57 (7.0%)
	TAPVC (cardiac)	39 (4.8%)
	Unbalanced AVSD	50 (6.2%)
	MAPCAs	24 (3.0%)

[a]Age is missing in 1 patient.

AVSD: atrioventricular septal defect; DILV: double-inlet left ventricle; DIRV: double-inlet right ventricle; MAPCAs: major aortopulmonary collateral arteries; TAPVC: total anomalous pulmonary venous connection.

Operative characteristics

CPB was utilized in 408 patients (50.2%) for 128 min (range 19–561). During CPB, the aorta was cross-clamped in 148 patients (18.2%) for 43.5 min (range 6–140). A modified Blalock–Taussig (BT) shunt, central shunt (a shunt whose inflow is ascending aorta or aortic arch) and other types of SP shunt were placed in 689 (84.8%), 94 (11.6%) and 30 patients (3.7%), respectively. In one patient, a modified BT shunt and central shunt were placed simultaneously. The isolated SP shunt procedure was performed in 411 patients (50.7%), while concomitant procedures were performed with the SP shunt procedure in the remaining 401 patients (49.3%). The most common concomitant procedure was the reconstruction of the PA (n = 194), followed by the closure of PDA (n = 147), enlargement or creation of atrial communication (n = 42), Damus–Kaye–Stansel anastomosis (n = 31), TAPVC repair (n = 26), unifocalization of MAPCAs (n = 21), superior cavopulmonary (Glenn) anastomosis (n = 20) and atrioventricular valve repair (n = 12). The operative characteristics are summarized in Table 2.

Postoperative events

Mechanical circulatory support was introduced in 34 patients (4.2%). There were 63 unplanned reoperations, not including exploration for haemostasis.

Hospital discharge and mortality

There were 741 (91.4%) hospital discharges 27 days (range 2–365) after the SP shunt procedure. Mortalities at 30 days and 90 days post-SP shunt procedure were 28 (3.4%) and 49 (6.0%), respectively. Within 90 days after operation, there were at least 12 deaths after hospital discharge, which accounted for 24% of all 90-day mortality. Figure 1 exhibits the Kaplan–Meier survival

Table 2: Operative characteristics

CPB utilization		408 (50.2%)
Aortic cross-clamp		148 (18.2%)
Duration of CPB (min)		128 (19–561)
Duration of aortic cross clamp (min)		43.5 (6–140)
Type of SP shunt		
	Modified BT shunt[a]	689 (84.9%)
	Central shunt[a]	94 (11.6%)
	Other	30 (3.7%)
Isolated SP shunt		411 (50.6%)
Concomitant procedures		
	Reconstruction of pulmonary artery	194 (23.9%)
	PDA closure	147 (18.1%)
	ASD enlargement	42 (5.2%)
	Damus–Kaye–Stansel anastomosis	31 (3.8%)
	TAPVC repair	26 (3.2%)
	Unifocalization of MAPCAs	21 (2.6%)
	Glenn anastomosis	20 (2.5%)
	Atrioventricular valve repair/ replacement	12 (1.5%)
	Pulmonary venous stenosis repair	5 (0.6%)
	Aortic coarctation repair	5 (0.6%)
	Other	29 (3.6%)

[a]In one patient, modified BT shunt and central shunt were placed simultaneously.

CPB: cardiopulmonary bypass; SP: systemic-to-pulmonary; BT: Blalock–Taussig; PDA: patent ductus arteriosus; TAPVC: total anomalous pulmonary venous connection; MAPCAs: major aortopulmonary collateral arteries.

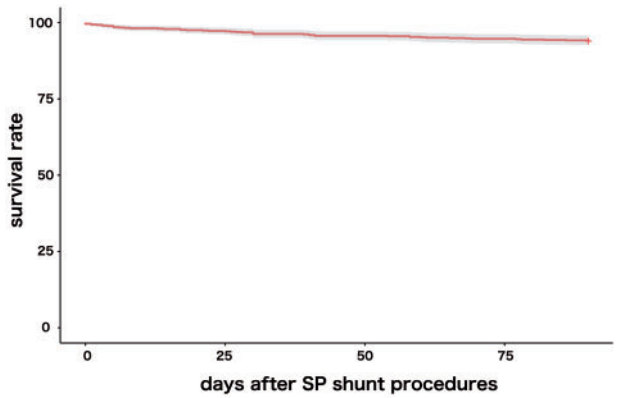

Figure 1: Kaplan–Meier survival curve within 90 days.

curve of total cohort within 90 days. Postoperative events, hospital discharge and mortalities are summarized in Table 3.

Risk factors for mortality

The results of the univariable logistic regressions between outcome and each categorical variable and the multivariable analysis are summarized in Tables 4 and 5, respectively. Placement of a central shunt (OR: 2.94, $P = 0.021$) was identified as a unique risk factor for 30-day mortality, while body weight < 2.5 kg (OR: 3.58, $P = 0.048$), body weight between 2.5 and 3.5 kg (OR: 2.28, $P = 0.039$), preoperative ventilator support (OR: 2.75, $P = 0.017$), diagnosis of right atrial isomerism (RAI) (OR: 2.38, $P = 0.007$), coexisting cardiac anomaly of MAPCAs (OR: 3.51, $P = 0.038$) and unbalanced AVSD (OR: 2.62, $P = 0.047$) were risk factors for 90-day mortality. Concomitant PDA closure was negatively associated with 90-day mortality (OR: 0.33, $P = 0.045$).

Table 3: Postoperative events and hospital discharge/ mortality

Postoperative events	
Mechanical circulatory support	34 (4.2%)
Unplanned reoperation	63 (7.8%)
Hospital discharge and mortality	
Hospital mortality[a]	49 (6.2%)
30-day mortality	28 (3.4%)
90-day mortality	49 (6.0%)
Hospital stay[b]	27 (2–365)

[a]Status at discharge is missing in 21 patients.

[b]Hospital stay is recorded among those who discharged alive.

DISCUSSION

SP shunt procedures continue to be an important palliative operation for various congenital heart diseases and are the fourth most commonly performed procedures in Japan in recent years [8]. Although surgical outcomes for most congenital heart disorders have improved over the years, SP shunt procedures are associated with a high mortality among patients with single ventricular physiology. There are some single- and multicentre studies that have identified functionally UVHs as a risk factor for mortality after SP shunt procedures [5, 6, 9]. However, functionally UVHs are characterized by a variety of fundamental cardiac diagnoses as well as coexisting cardiac anomalies. Because of the diversity of coexisting cardiac anomalies, other concomitant cardiac procedures, such as the closure of the PDA or PA plasty, are often mandatory when SP shunt procedures are performed.

The outcome of SP shunt procedures is strongly affected by the patient's fundamental cardiac diagnosis, coexisting cardiac anomalies and concomitant procedures. In the present study, we analysed these factors to assess the mortality risk after SP shunt procedures. The 30-day and 90-day mortality rates were 28 (3.4%) and 49 (6.0%), respectively. As the data at discharge were missing in 21 patients, the resulting hospital mortality was 49 (6.2%) out of 791 patients. These results are comparable to previous studies which reported 7–15% hospital mortality [5, 10, 14].

In a previous study, nearly 33% of fatalities after BT shunt procedures for neonates occurred within 24 hours and 75% within 30 days [5], while in our study, there was a discrepancy between 30-day mortality (3.4%) and hospital mortality (6.2%). This discrepancy may be caused by postoperative mechanical circulatory support introduced in 34 (4.2%) patients in our study cohort. Out of these 34 patients, 11 (32%) patients died within 30 days after the SP shunt procedure, while the remaining 23 (68%) patients survived beyond 30 days, with further five deaths within 90 days. From these data, we consider that the use of mechanical circulatory support improves the early results of the SP shunt procedure. Similarly, the use of mechanical circulatory support might

Table 4: Results of univariable logistic regression

Covariate	Odds ratio	95% CI lower	95% CI upper	*P*-value
Early delivery	1.77	0.67	4.68	0.25
Preoperative ventilator support	3.58	1.69	7.58	0.001
Asplenia syndrome	2.72	1.51	4.89	0.001
Body weight < 2.5 kg	5.60	1.77	17.73	0.003
Body weight 2.5–3.5 kg	2.32	1.10	4.88	0.027
Body weight >5.0 kg	1.20	0.45	3.21	0.72
Tricuspid atresia	0.49	0.19	1.25	0.14
Heterotaxia syndrome	1.65	0.92	2.96	0.091
Mitral atresia	0.54	0.13	2.27	0.40
Single ventricle, other	0.99	0.34	2.84	0.98
Unbalanced AVSD	2.28	0.92	5.65	0.075
Pulmonary atresia	1.79	1.00	3.20	0.051
Pulmonary stenosis	0.34	0.10	1.10	0.071
PA stenosis	0.43	0.13	1.42	0.17
Discontinuous PA	0.67	0.09	5.07	0.70
TAPVC, supracardiac type	0.89	0.21	3.79	0.87
TAPVC, cardiac type	2.44	0.91	6.54	0.077
TAPVC, mixed type	2.91	0.63	13.50	0.17
MAPCAs	4.45	1.59	12.48	0.005
Hypoplastic left ventricle	1.97	0.24	16.04	0.53
Central shunt	1.79	0.84	3.83	0.13
Modified BT shunt	0.59	0.30	1.20	0.15
Other type of SP shunt	1.78	0.52	6.08	0.36
No concomitant procedure (isolated SP shunt)	0.93	0.52	1.66	0.81
ASD creation or enlargement	0.77	0.18	3.28	0.72
PDA closure	0.39	0.14	1.09	0.072
PA reconstruction	0.92	0.46	1.83	0.81
TAPVC repair	2.99	0.99	9.06	0.052
Unifocalization of MAPCA	3.90	1.26	12.08	0.018
Glenn procedure	0.82	0.11	6.22	0.84

ASD: atrial septal defect; AVSD: atrioventricular septal defect; BT: Blalock–Taussig; CI: confidence intervals; MAPCA: major aortopulmonary collateral arteries; PA: pulmonary artery; PDA: patent ductus arteriosus; SP: systemic-to-pulmonary; TAPVC: total anomalous pulmonary venous connection.

Table 5: Risk factors for 30- and 90-day mortality

Risk factors for 30-day mortality	Odds ratio (95% CI)	*P*-value
Body weight < 2.5 kg	3.47 (0.65–18.60)	0.15
Body weight 2.5–3.5 kg	1.85 (0.70–4.86)	0.21
Body weight >5.0 kg	0.77 (0.21–2.86)	0.69
Pulmonary stenosis	0.16 (0.02–1.26)	0.082
Unbalanced AVSD	2.65 (0.85–8.26)	0.093
Central shunt	2.94 (1.17–7.36)	0.021
Concomitant PDA closure	0.31 (0.07–1.33)	0.11
Risk factors for 90-day mortality	**Odds ratio (95% CI)**	***P*-value**
Body weight < 2.5 kg	3.58 (1.01–12.67)	0.048
Body weight 2.5–3.5 kg	2.28 (1.04–4.99)	0.039
Body weight 5.0 kg <	1.01 (0.36–2.82)	1.00
Preoperative ventilator support	2.75 (1.20–6.30)	0.017
Right atrial isomerism	2.38 (1.27–4.44)	0.007
Pulmonary stenosis	0.36 (0.10–1.22)	0.10
MAPCAs	3.51 (1.07–11.52)	0.038
Unbalanced AVSD	2.62 (1.01–6.79)	0.047
TAPVC mixed type	3.88 (0.78–19.23)	0.097
Concomitant PDA closure	0.34 (0.11–0.98)	0.045

AVSD: atrioventricular septal defect; CI: confidence intervals; MAPCAs: major aortopulmonary collateral arteries; PDA: patent ductus arteriosus; TAPVC: total anomalous pulmonary venous connection.

reflect that no risk factors for 30-day mortality were identified except for placement of a central shunt.

In the present study, placement of a central shunt was identified as a unique risk factor for 30-day mortality. Indeed, the 30-day mortality was significantly higher in patients who underwent central shunt placement than in those who underwent modified BT shunt placement (7.4% vs 2.9%, $P = 0.037$). It is difficult to discern why the placement of the central shunt was identified as a unique risk factor for 30-day mortality. Judging from the percentage of modified BT shunt (84.9%), it is rational to consider that modified BT shunt is the first choice of SP shunt procedure in the majority of Japanese institutions. Under the situation, a central shunt could be an unusual procedure for high-risk patients not suitable for modified BT shunt, such as very hypoplastic PA. Although the prevalence of modified BT shunt may partly explain our result, further study using central shunt subgroup is necessary to elucidate the reason for the higher mortality of central shunt.

As described in other reports, lower body weight was also identified in this study as a risk factor for 90-day mortality [5, 7]. Interestingly, younger age at SP shunt placement was not recognized as a risk factor for mortality. This may simply imply that a small shunt size is difficult to manage, as the shunt size is determined based on the patient's body weight. Previous studies reported that a 3.0 mm shunt size was associated with a higher rate of shunt thrombosis and shunt reintervention [14, 15]. However, this could not be confirmed in our study due to lack of data regarding shunt size within the database. Our results suggest that SP shunt procedures for patients with a functionally UVH should be performed after patients gain 3.5 kg in body weight, or at least 3.0 kg if their conditions are stable. To achieve this goal, many patients would need prolonged prostaglandin E1 infusion or ductal stenting. The former frequently induces apnoea attack

which may necessitate ventilator support, another risk factor revealed in this study. In addition, prolonged hospitalization is another issue of this approach. The latter may involve aortic wall and complicate subsequent procedures. In Japan, continuous infusion of prostaglandin E1 is preferred partly because entire hospital charge is covered by national insurance. In any case, the timing of SP shunt procedure should be carefully decided in each patient, assessing whether the benefits of body weight gain surpass the risks of these approaches.

As several studies have reported, this study also revealed that the outcome of SP shunt procedures is affected by underlying cardiac anomalies and concomitant procedures [5, 10, 14]. There were 274 patients with heterotaxy, accounting for 33.8% of the study cohort. Using the database of the Society of Thoracic Surgeons, Jacobs *et al.* reported that patients with heterotaxy had higher discharge mortality for every cardiac procedure compared to those without heterotaxy. In particular, discharge mortality after SP shunt placement was 6.6% in a cohort of all patients with a single ventricle without heterotaxy, while it was 10.8% in those with heterotaxy [16]. Alsoufi *et al.* [10] also reported that the diagnosis of heterotaxy was significantly associated with worse survival rates after modified BT shunt procedures in neonates with single ventricular anatomy. The diagnosis of RAI, but not left atrial isomerism, was identified as a risk factor for 90-day mortality in our study. This result is consistent with the fact that RAI is a clinically more severe type of heterotaxy syndrome [16]. As patients with RAI are often accompanied by TAPVC, which requires surgical repair during the very early stage of life, we hypothesized that concomitant TAPVC repair would be a risk factor for mortality. However, TAPVC was not statistically significant in the univariable analysis ($P = 0.052$).

The combination of MAPCAs was identified as one of the risk factors for 90-day mortality in our study. Some single-institutional studies have reported that the SP shunt procedure for patients with a single ventricle combined with MAPCAs was associated with high mortality. Patric *et al.* reported 33 such patients who underwent SP shunt procedures with or without unifocalization. In the report, 14 patients (42%) died before the second-stage palliation [17]. As the combination of MAPCAs and a functionally UVH is very rare, it would be difficult to prove that the combination is a risk factor without utilizing a nationwide database.

In our study, a coexisting cardiac anomaly of unbalanced AVSD was also identified as a risk factor for 90-day mortality (OR: 2.62, $P = 0.047$). Unbalanced AVSD constitutes approximately 10% of all AVSD cases [18]. Those patients often present with significant atrioventricular valve regurgitation, which causes morbidity and mortality [19, 20]. Our results may represent the deterioration of valve regurgitation caused by ventricular volume overload after the placement of SP shunts. Moreover, MAPCAs and unbalanced AVSD are often observed in patients with heterotaxy, and may represent severe forms of heterotaxy [17, 19, 20].

Whether or not a PDA should be closed concomitant with the SP shunt procedure remains controversial. Zahorec *et al.* [21] advocated that PDA closure during a modified BT shunt procedure was associated with early postoperative mortality. However, the dual source of pulmonary blood flow presents a risk of pulmonary over circulation, significant coronary steal during the diastolic phase and subsequent myocardial dysfunction, which are strongly associated with morbidity and mortality. Moreover, the competitive shunt flow makes the total pulmonary blood flow uncertain. The adverse aspects of a dual source pulmonary blood flow may explain why the concomitant PDA closure was negatively associated with the 90-day mortality after SP shunt procedures in the present study.

High inter-stage mortality after the SP shunt procedure is also problematic. Alsoufi *et al.* reported that hospital and inter-stage mortality after modified BT shunt procedures for neonates with single ventricle anomalies were 15% and 10%, respectively [10]. As such, approximately 25% of patients would not be able to undergo the Glenn operation. In another report by Fenton *et al.* [22], 14% of patients who underwent the SP shunt procedure died after hospital discharge and before follow-up surgery. In addition, most of them had unexpected sudden deaths, even though they were doing well at time of hospital discharge. These reports remind us of the instability and fragility of parallel circulation with SP shunts. As Ota *et al.* [23] reported, early escape from insecure circulation with SP shunts and achievement of the Glenn procedure may improve the inter-stage mortality among high-risk patients.

It is important to note that there are some limitations to our study. This is a retrospective study conducted with data from the national clinical database, in which data entry is completed voluntarily. The database lacks some important information, such as indication of SP shunt, shunt size, type of central shunt and postoperative anticoagulation therapy. Furthermore, institutional volume and surgeon's experience were not taken into account in the present study.

Another limitation is that the number of occurrences of outcome events of this study is small. We should carefully interpret the results of the multivariable logistic regressions as a small number of events could cause insufficiency of events per variable and may cause instability of estimation of parameters.

CONCLUSION

According to our study, after the initial SP shunt procedure for patients with a functionally UVH, approximately 90% of patients were discharged within a month. The placement of a central shunt is a unique risk factor for 30-day mortality, while lower body weight, preoperative mechanical ventilator support, diagnosis of RAI, coexisting cardiac anomaly of MAPCAs and unbalanced AVSD were identified as risk factors for 90-day mortality.

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Author contributions

Yujiro Ide: Conceptualization; Writing – original draft; Writing – review & editing. **Hisateru Tachimori:** Data curation; Methodology; statistical analysis. **Yasutaka Hirata:** Supervision. **Norimichi Hirahara:** Methodology; statistical analysis. **Noritaka Ota:** Conceptualization; Supervision. **Kisaburo Sakamoto:** Supervision. **Tadashi Ikeda:** Supervision; Validation; Writing – review & editing. **Kenji Minatoya:** Supervision; Validation.

Reviewer information

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