Management of failing bidirectional cavopulmonary shunt: Influence of additional systemic-to-pulmonary-artery shunt with classic Glenn physiology

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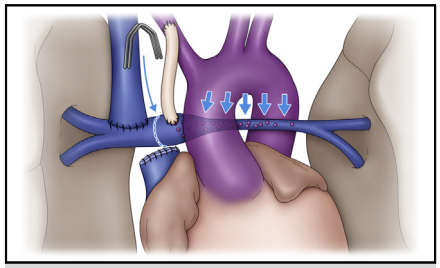
ABSTRACT

Objectives: Severe hypoxemia in the early postoperative period after bidirectional cavopulmonary shunt (BCPS) is a critical complication. We aimed to evaluate patients who underwent additional systemic to pulmonary shunt and septation of central pulmonary artery (partial takedown) after BCPS.

Methods: The medical records of all patients who underwent BCPS between 2007 and 2020 were reviewed. Patients who underwent partial takedown were extracted and their outcomes were analyzed.

Results: Of 441 BCPS patients, 27 patients (6%) required partial takedown. Most frequent diagnosis was hypoplastic left heart syndrome (n = 14; 52%). Additional complicating factors included pulmonary artery hypoplasia (n = 12) and pulmonary venous obstruction (n = 3). Thirteen patients (48%) underwent partial takedown on the same day of BCPS, and all of them survived the procedure. The remaining 14 patients (52%) underwent partial takedown between postoperative 1 to 64 days. The reasons for partial takedown were: postoperative high pulmonary vascular resistance (n = 4), early BCPS (<90 days) with PA hypoplasia (n = 3), mediastinitis/pneumonia (n = 3), pulmonary venous obstruction (n = 2), ventricular dysfunction (n = 1), and recurrent pneumothorax (n = 1). Four patients experienced hospital deaths. Six patients died after discharge, 10 achieved Fontan completion, and 6 were alive and waiting for Fontan. Overall survival after partial takedown was 54% at 3 years. The pulmonary venous obstruction ($P = .041$) and genetic/extracardiac anomalies ($P = .085$) were identified as risks for mortality after partial takedown.

Conclusions: The partial takedown resulted in a 3-year survival rate of more than 50%. Of these patients, a significant number underwent successful Fontan completion who would exhibit potential early death with conservative treatment. (JTCVS Open 2022;11:373-87)



Schematic drawing of additional systemic-to-pulmonary shunt with classic Glenn physiology.

CENTRAL MESSAGE

In 27 patients who underwent additional systemic-to-pulmonary shunt for failing bidirectional cavopulmonary shunt, survival at 3 years was 54% and Fontan completion was done or planed in 16 patients.

PERSPECTIVE

Additional systemic-to-pulmonary-artery shunt with classic Glenn physiology was performed for heterogeneous reasons in 27 patients with failing bidirectional cavopulmonary shunt. Although these patients frequently develop thrombus and need ECMO, survival and successful Fontan completion rate are encouraging. Refinement of this strategy might improve the survival of this high-risk population.

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Abbreviations and Acronyms

AVV = atrioventricular valve
BCPS = bidirectional cavopulmonary shunt
CPB = cardiopulmonary bypass
ECMO = extracorporeal membrane oxygenation
HLHS = hypoplastic left heart syndrome
PA = pulmonary artery
PAP = pulmonary artery pressure
PVO = pulmonary venous obstruction
SVC = superior vena cava
TCPC = total cavopulmonary connection
VF = ventricular function

Video clip is available online.

Because we actually experience no early mortality after stage III Fontan palliation, concerns have been focused on the indication, timing, and postoperative management of stage II palliation by means of bidirectional cavopulmonary shunt (BCPS) procedure in patients with functional single ventricle physiology.[1-3] Although low pulmonary artery pressure (PAP) is the most important criteria for successful BCPS, there are many other issues to be considered, such as pulmonary artery (PA) hypoplasia, pulmonary venous obstruction (PVO), reduced systemic ventricular function (VF), atrioventricular valve (AVV) regurgitation, and other underlying patient diseases.[4,5] Despite sophisticated preoperative patient selection, we encounter certain patients who demonstrated severe hypoxemia after BCPS, so called failing BCPS. A fundamental concern of BCPS is that pulmonary blood flow is entirely dependent on passive superior vena cava (SVC) flow, and the success of BCPS is considered to be exquisitely dependent on an unobstructed, low-resistance pulmonary vascular bed.[6] When a BCPS fails without any SVC-PA pathway obstruction, SVC flow is insufficient to supply pulmonary blood flow in both lungs to maintain BCPS circulation. Differential diagnoses include postoperative high pulmonary vascular resistance (occasionally due to pulmonary infection), PVO, AVV regurgitation, or systemic ventricular dysfunction. These conditions are difficult to manage.[7]

In this setting, complete takedown of BCPS to the shunt dependent pulmonary circulation is a standard option. However, previous studies have shown that this approach has a high mortality rate.[8,9] Since 2007, we have consistently performed another option, so-called partial takedown (ie, placement of additional systemic-to-pulmonary shunt and interruption of central pulmonary artery between BCPS and systemic-to-pulmonary shunt) to overcome this severe situation (Figure 1). Therefore, we undertook the present study to determine the short- to midterm outcomes of patients who underwent partial takedown for failing BCPS to evaluate the usefulness of this option and to analyze the risks for survival after partial takedown.

MATERIALS AND METHODS

We reviewed data on all patients who underwent staged II BCPS palliation at the German Heart Center Munich between January 2007 and December 2020 and extracted the patients who underwent partial takedown (additional systemic-to-pulmonary shunt with interruption of the PA continuity between BCPS and additional systemic-to-pulmonary shunt). The Institutional Review Board of the Technische Universität München approved the study and waived the requirement for patient consent (approved ID: 65/20 S-KH; February 18, 2020). Review of medical records, including in-hospital and outpatient notes, echocardiography, and cardiac catheterization was performed.

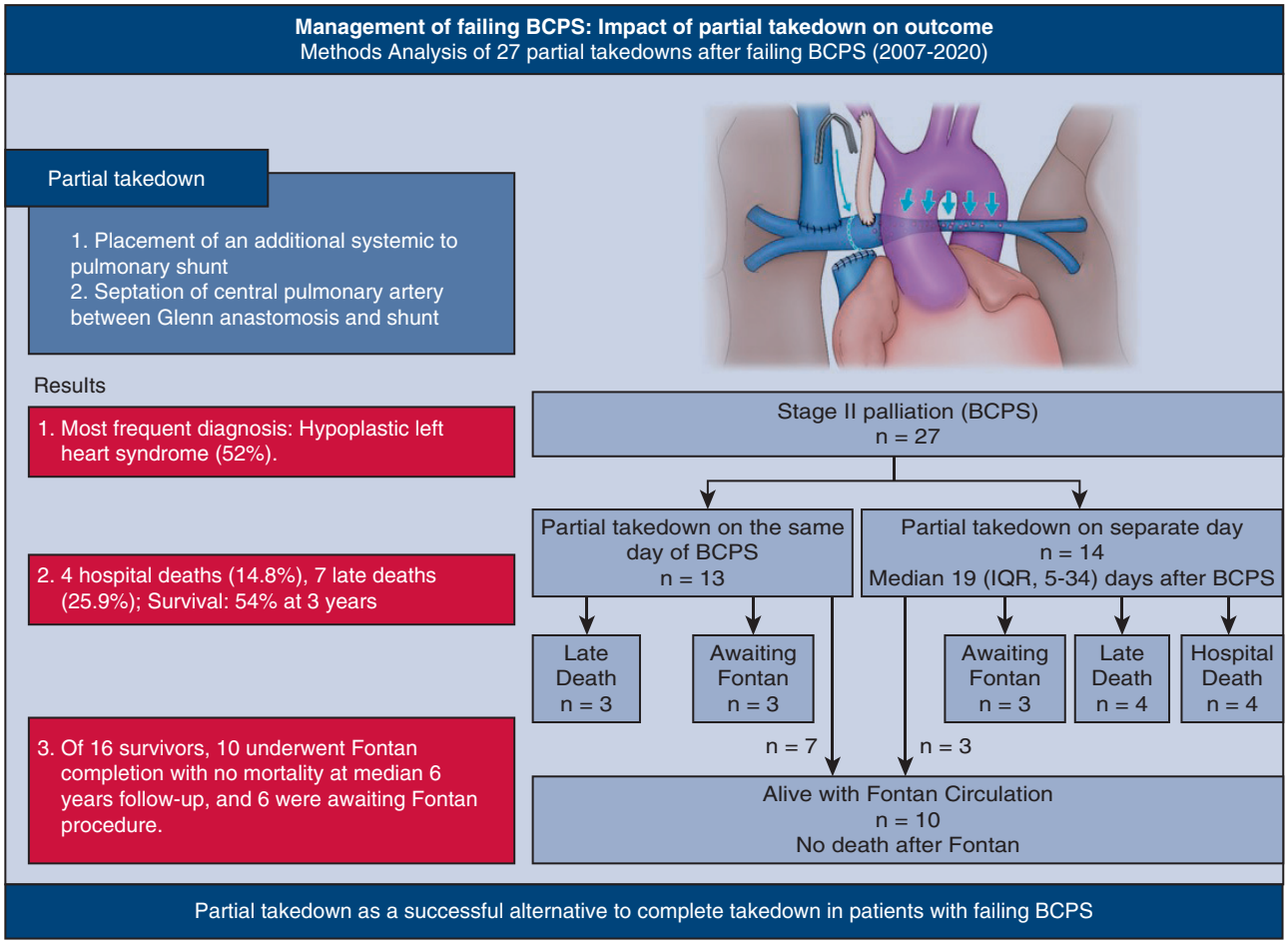
Pre-BCPS Assessment

In our institution, all patients routinely underwent cardiac catheterization and echocardiography as the preoperative evaluation before BCPS. Cardiac catheterization was performed to obtain hemodynamic data and angiographic assessment of PA, and echocardiography to assess systemic VF and AVV regurgitation. Systemic VF was assessed qualitatively, and reduced VF was defined as ejection fraction <50%. The AVV regurgitation was graded as previously described.[10] Moderate or more AVV regurgitation was defined as a significant AVV regurgitation.

Surgical Techniques

BCPS was performed with mild hypothermic cardiopulmonary bypass (CPB) and standard bicaval cannulation, as described in our previous reports.[1,11] A catheter to monitor central venous pressure was routinely inserted into the right internal jugular vein. Cardioplegic arrest was used only for patients who required intracardiac procedures. The azygos vein was routinely divided before initiation of CPB. The BCPS anastomosis and PA reconstruction were performed in an on-pump beating state. Any antegrade pulmonary blood flow was routinely eliminated at the time of BCPS by dividing the main PA and oversewing the pulmonary valve, or by dividing previous systemic-to-pulmonary shunt.[11] The SVC was anastomosed to the right PA in an end-to-side fashion using 7–0 or 8–0 polydioxanone continuous sutures (Ethicon Inc).

Partial takedown was performed in patients who demonstrated severe hypoxemia after BCPS (Figure 2). On CPB, an additional systemic-to-pulmonary shunt was placed to the left of BCPS anastomosis with 3.0 mm, 3.5 mm, or 4.0 mm polytetrafluoroethylene graft. Shunt size was determined by the surgeons according to patient's size, pulmonary artery size, and estimation of the pulmonary vascular resistance of the affected lung. Proximal anastomosis was performed in the innominate artery, or in the ascending aorta (central aortopulmonary shunt). In some patients, the preexisting shunt was applied for partial takedown. The interruption of PA continuity was accomplished by ligation, clipping, or patch insertion of the middle portion between the BCPS and the additional systemic-to-pulmonary shunt. Methods of PA interruption were chosen at the surgeon's preference.



BCPS: bidirectional cavopulmonary shunt, SPS: systemic to pulmonary shunt, PTD: partial takedown

FIGURE 1. Outcomes of partial takedown in 27 patients who presented with prohibitive cyanosis after bidirectional cavopulmonary shunt (*BCPS*) during the study period between 2007 and 2020. More than half of the patients were diagnosed with hypoplastic left heart syndrome. The causes of partial takedown were left pulmonary artery hypoplasia in 10 patients, elevated pulmonary vascular resistance in 5 patients, mediastinitis/pneumonia in 4 patients, early BCPS <90 days in 3 patients, pulmonary venous obstruction in 3 patients, ventricular dysfunction in 1 patient, and recurrent pneumothorax in 1 patients. There were 4 hospital deaths and 7 late deaths. Kaplan-Meier estimate transplant-free survival was 54% at 3 years. During the follow-up, 10 patients completed the Fontan procedure. *IQR*, Interquartile range.

Postoperative Management

As for the anticoagulation strategies, postoperative standard thrombosis prophylaxis after BCPS consisted of intravenous administration of unfractionated heparin (5000 IU/m^2/d) with a target partial thromboplastin time of 60 seconds, until all central lines (usually a 4.5Fr catheter was used) were removed (usually 4-5 postoperative days). Later, no routine antithrombotic prophylaxis was given after BCPS. Nevertheless, patients who underwent partial takedown received acetylsalicylic acid or warfarin due to a persistent aortopulmonary shunt. In patients who needed extracorporeal membrane oxygenation (ECMO) support, anticoagulation was performed using intravenous administration of unfractionated heparin with the target range of anti-Xa 0.35 to 0.50 U/mL, activated clotting time within 180 to 200 seconds and partial thromboplastin time of 60 to 80 seconds.

Follow-up Data

Patients were followed-up by pediatric cardiologists as outpatients, and follow-up times were defined per patient as the time from the day of partial takedown to the last visit. For the patients who died, the data were collected at the time of death. Because no heart transplantation was performed in the entire cohort during the study period, the primary outcome of interest was the composite end point of Fontan completion or death. The follow-up data from time of surgery until the last known record of the patients were regularly tracked using our institutional single ventricle patient database system.

Statistical Analysis

Categorical variables are presented as absolute numbers and percentages. A χ^2 test was used for categorical data. Continuous variables are expressed as mean $\pm$ SD or median with interquartile range (IQR), if appropriate. An independent sample *t* test was used to compare normally distributed variables. Mann-Whitney test was used for variables that were not normally distributed. Survival after BCPS partial takedown was analyzed using the Kaplan-Meier method. A univariable Cox proportional hazard regression model was used to assess the association between preoperative risk factors and mortality after partial takedown. The proportional hazard assumption was tested using Schoenfeld residuals. We did not

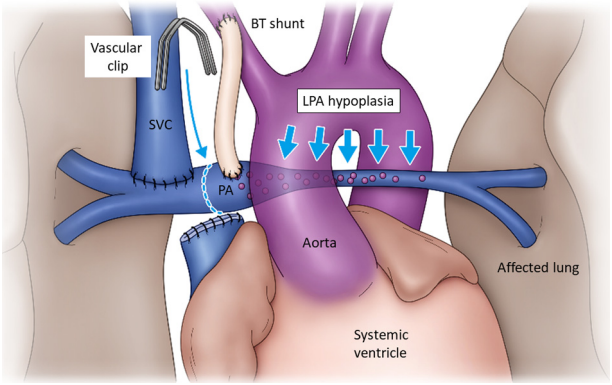


FIGURE 2. Schematic drawing of partial takedown (additional systemic to pulmonary shunt with classic Glenn physiology). *BT*, Blalock-Taussig shunt; *SVC*, superior vena cava; *LPA*, left pulmonary artery; *PA*, pulmonary artery.

correct for multiple testing in all tests and models we performed. Data analysis and graphing were performed with the Statistical Package for the Social Sciences version 28.0 for Windows (IBM-SPSS Inc) and state package and CMPRSK package in R version 4.0.3 (R Foundation for Statistical Computing).

RESULTS

Patient's Characteristics and Pre-BCPS Hemodynamic Assessment

Among 441 patients who underwent BCPS at our institution between January 2007 and December 2020, 27 patients (6%) needed partial takedown. Patient characteristics are shown in Table 1. Primary diagnoses included 14 (52%) hypoplastic left heart syndromes (HLHS), 4 (15%) tricuspid atresia, and 3 (11%) unbalanced atrioventricular septal defects. Genetic/extracardiac anomalies were associated in 9 patients (33%).

Pre-BCPS cardiac catheterization data are shown in Table 2. Hemodynamic assessment revealed mean PAP of 14.5 ± 3.9 mm Hg, mean transpulmonary gradient of 7.4 ± 2.7 mm Hg, systemic ventricular end-diastolic pressure of 9.5 ± 2.9 mm Hg, and arterial oxygen saturation of 75.5% ± 6.1%. Pulmonary vascular pathology included left PA hypoplasia in 12 patients (Figure E1) and PVO in 3 patients. In these 3 patients with PVO, total/partial anomalous pulmonary venous connection had been repaired at the time of stage I or II palliation, and PVO had at least 1 lobar pulmonary vein branch. No patient demonstrated abundant aortopulmonary collaterals, sizable systemic venous-venous collaterals, or re-coarctation of the aorta. A preoperative echocardiography performed immediately before BCPS showed reduced VF in 3 patients and significant AVV regurgitation in 5 patients.

Operative and Early Postoperative Outcomes

Patient-specific details and additional procedures at the time of BCPS are listed in Table 3. Median age and weight at BCPS were 4.1 months (IQR, 3.2-4.8 months) and 4.7 kg (IQR, 4.1-5.6 kg), respectively. Right BCPS was performed in 26 patients and left BCPS was performed in 1 patient with situs inversus and a single left SVC. No patient after bilateral BCPS demonstrated failing BCPS, so there was no patient who underwent bilateral BCPS. The median CPB time was 69 minutes (IQR, 52-107 minutes), and aortic cross-clamp was needed in 5 patients (19.2%) with a median duration of 18 minutes (IQR, 15-33 minutes). Intracardiac procedures included 11 PA reconstructions (including 6 out of 12 patients with left PA hypoplasia), 2 AVV repairs, 2 atrioseptectomies, (Blalock-Hanlon procedure), and 1 pulmonary valve closure. An autologous or xeno pericardium was used for PA reconstruction.

Partial takedown was performed either on the same day of BCPS in 13 patients or between postoperative days 1 and 64 in 14 patients (Table 1 and Figure 3). Among the 13 patients who underwent partial takedown on the same day as BCPS, there were 11 cases that were performed simultaneously with BCPS procedure, and 2 cases that were performed separately several hours after BCPS procedure. Notably, 11 of the 13 patients had previous Norwood procedure, and 9 patients demonstrated left PA hypoplasia (Tables 1 and 4). In 11 concomitant cases, systemic oxygen saturation after weaning from CPB and modified ultrafiltration varied between 40% and 60% even with nitric oxide inhalation therapy. An additional systemic-to-pulmonary shunt was constructed immediately. A new modified Blalock-Taussig shunt was placed in 4 patients, and the preexisting shunt was used in remaining 7 patients. Also, pulmonary septation was performed using a vascular clip (n = 6) or simple ligation (n = 3) or patch (n = 2) placed between the BCPS and the systemic-to-pulmonary shunt. Systemic oxygen saturation increased to 75% to 90%, and SVC pressure was acceptable (<20 mm Hg) after partial takedown in all patients. Another 2 patients underwent partial takedown as a separate operation. They demonstrated severe hypoxemia shortly after admission to intensive care unit, and were operated on again several hours later. A new, 3.5-mm modified Blalock-Taussig shunt was placed and septation was performed using a vascular clip in both cases. In the 13 patients who underwent partial takedown on the same day of BCPS, median stay in the intensive care unit was 7 days (IQR, 6-15 days), and all were successfully discharged from hospital a median of 18 days (IQR, 13-32 days) after partial takedown.

The remaining 14 patients underwent partial takedown on a different day, and they had complicated postoperative courses. Postoperative data of these patients are shown in Tables 4 and 5. The interval between BCPS and partial takedown was a median of 19 days (IQR, 5-34 days). Median stay in intensive care unit was 45 days (IQR, 30-81 days) and median hospital stay was 59 days (IQR, 42-101 days). These patients had initial arterial oxygen saturation more

TABLE 1. Patient characteristics and bidirectional cavopulmonary shunt (BCPS) data

Patient No.	Diagnosis	Genetic/ extracardiac anomalies	Stage I palliation	Shunt size	PA hypoplasia	LPA index	BCPS age (mon)	PCPC height (cm)	Interval (d)	Causes of hypoxemia	Additional shunt	Shunt size
1	HLHS		Norwood	5.0	LPA (2.6 mm)	38	5.0	61	Con	Hypo LPA	Sano LPA	5.0
2	DILV, TGA, IAA		Norwood	4.0	LPA (3.2 mm)	43	3.7	60	Con	Hypo LPA	BT LPA	3.5
3	HLHS	Dandy-Walker	Norwood	5.0	LPA (2.5 mm)	27	5.1	60	Con	Hypo LPA	Sano LPA	5.0
4	DORV, MS	Tetrasomie 18p	Norwood	3.5	LPA (1.2 mm)	31	3.8	56	Con	Hypo LPA	BT LPA	3.5
5	TA Ib	VACTERL	no			279	2.2	59	Con	Low SVC flow	BT LPA	3.5
6	HLHS	Cleft palate	Norwood	3.0	LPA (1.5 mm)	12	4.6	53	Con	Hypo LPA	BT LPA	3.0
7	HLHS		Norwood	3.5	LPA (1.9 mm)	35	4.7	60	Con	Hypo LPA	BT LPA	3.5
8	HLHS		Norwood	3.0		76	3.1	50	Con	Low SVC flow	BT LPA	3.0
9	DORV, MA		Norwood	3.5	LPA (3.0 mm)	29	4.2	54	Con	Hypo LPA	BT LPA	3.5
10	HLHS, TAPVC		Norwood	3.5		86	5.1	57	Con	Low SVC flow	CS LPA	3.5
11	HLHS		Norwood	3.5		86	3.6	52	Con	Low SVC flow	BT LPA	3.5
12	HLHS		Norwood	3.5	LPA (2.0 mm)	35	1.8	55	0	Hypo LPA	BT LPA	3.5
13	DIRV, TGA, PA		PDA stent	4.0	LPA (3.5 mm)	44	3.4	57	0	Hypo LPA	CS RPA	3.5
14	HLHS, PAPVC		Norwood	3.5		53	3.4	58	1	PVO	BT LPA	3.5
15	UAVSD, TGA, PA	CHARGE	PDA stent	3.5		148	7.5	64	3	high PVR	CS LPA	4.0
16	HLHS		Norwood	3.5		66	4.5	54	4	high PVR	BT LPA	3.0
17	HLHS		Norwood	3.5	LPA (3.0 mm)	52	2.7	54	5	Early BCPS	BT LPA	3.5
18	HLHS, TAPVC		Norwood	3.5		85	2.8	61	9	Early BCPS	BT LPA	3.5
19	UAVSD	Trisomy 21	PAB			336	12.8	78	13	high PVR	CS LPA	4.0
20	HLHS		Norwood	5.0		82	4.3	58	17	TR, reduced VF	BT LPA	3.5
21	PAIVS, Ebstein		MBTS	3.5		98	4.1	61	21	Mediastinitis	CS LPA	3.5
22	UAVSD, DORV, PS	Tracheal stenosis	MBTS	3.5		184	3.2	58	23	PVO	BT LPA	3.5
23	TA Ia		PDA stent	4.0	LPA (2.7 mm)	88	5.2	64	29	high PVR	CS LPA	3.0
24	TA Ib		PDA stent	3.5		66	2.9	56	33	Early BCPS	CS LPA	3.5
25	ccTGA, TA, IAA	Notch-Gen	Norwood	3.5	LPA (2.6 mm)	72	4.1	62	35	Mediastinitis	BT LPA	3.5
26	TA Ic	Renal anomalies	MBTS	3.5		107	4.1	62	62	Pneumothorax	BT LPA	4.0
27	HLHS		Norwood	5.0		86	3.5	60	63	Mediastinitis	BT LPA	3.5

PA, Pulmonary artery; *LPA*, left pulmonary artery; *PCPC*, partial cavopulmonary connection; *HLHS*, hypoplastic left heart syndrome, *Con*, concomitant; *Sano*, Sano shunt (right ventricle to pulmonary artery conduit); *DILV*, double inlet left ventricle; *TGA*, transposition of the great arteries; *IAA*, interrupted aortic arch; *BT*, Blalock-Taussig; *DORV*, double outlet right ventricle; *MS*, multiple sclerosis; *TA*, tricuspid atresia; *VACTERL*, VACTERL syndrome; *SVC*, superior vena cava; *TAPVC*, total anomalous pulmonary venous return; *CS*, central shunt; *RPA*, right pulmonary artery; *DIRV*, double inlet right ventricle; *PA*, pulmonary artery; *PAPVC*, partial anomalous pulmonary venous connection; *PVO*, pulmonary venous obstruction; *UAVSD*, unbalanced atrioventricular septal defect; *PVR*, pulmonary vascular resistance, *LPA*, left pulmonary artery; *PAB*, pulmonary artery banding; *TR*, tricuspid valve regurgitation; *VF*, ventricular function; *MBTS*, modified Blalock-Taussig shunt; *CHARGE*, CHARGE association; *PAIVS*, pulmonary atresia with intact ventricular septum; *Ebstein*, Ebstein anomaly; *PS*, pulmonary stenosis; *PDA*, patent ductus arteriosus; *ccTGA*, congenitally corrected transposition of the great arteries.

than 70% but demonstrated progressive hypoxemia. All underwent postoperative cardiac catheterization at a median of 12 days (IQR, 2-19 days) after BCPS to confirm etiology of severe hypoxemia. The following causes of cyanosis were identified: early BCPS before 90 days with mild-to-moderate PA hypoplasia in 3 patients, PVO in 2 patients, post-BCPS high pulmonary vascular resistance in 4 patients, severe tricuspid valve regurgitation and systemic ventricular dysfunction in 1 patient, post-BCPS mediastinitis and pneumonia in 3 patients, and postoperative recurrent pneumothorax in 1 patient (Table 4). The postoperative causes in these patients are shown in Table E1.

There were 4 hospital deaths before discharge, and the cause of deaths was considered as 2 high pulmonary vascular resistance, 1 PVO, and 1 mediastinitis/pneumonia. As morbidity, a thrombus formation was detected in 11 of 14 patients (78.6%). Six out of 11 patients developed thrombus between initial BCPS and partial takedown, and the remaining 5 patients developed thrombus after partial takedown. Thrombolysis was done in all patients and operative removal of thrombus was performed in 5 patients. Of the 5 patients who required surgical removal of thrombosis, thrombus was located in SVC to PA in 4 patients and in right atrium in 1 patient. ECMO therapy was needed in 9 of 14 (64.3%) patients. ECMO was needed before partial takedown in 6 patients. One patient successfully weaned from ECMO before partial takedown and 3 weaned from ECMO at the time of partial takedown. ECMO support

TABLE 2. Pre-bidirectional cavopulmonary shunt catheterization and echocardiographic data

Variable	Result
Catheterization data	
Hemoglobin (g/dL)	13.8 ± 1.9
Mean pulmonary artery pressure (mm Hg)	14.5 ± 3.9
Mean left atrial pressure (mm Hg)	6.9 ± 2.5
Transpulmonary gradient (mm Hg)	7.4 ± 2.7
Systolic ventricular pressure (mm Hg)	78.0 ± 12.4
Ventricular endodiastolic pressure (mm Hg)	9.5 ± 2.9
Aortic oxygen saturation (%)	75.5 ± 6.1
Echocardiographic data	
Ventricular function	
Normal	23 (85.2)
Mildly impaired	2 (7.4)
Moderately impaired	2 (7.4)
Atrioventricular valve regurgitation	
None	3 (11.1)
Trivial	10 (37.0)
Mild	9 (33.3)
Moderate	4 (14.8)
Severe	1 (3.7)
(Neo) aortic insufficiency	
None	20 (74.1)
Trivial	6 (22.2)
Mild	1 (3.7)

Values are presented as n (%) or mean ± SD.

TABLE 3. Perioperative variables

Variable	Result
No. of patients	27
Operative data	
Age at BCPS (mo)	4.1 (3.2-4.8)
Weight at BCPS (kg)	4.7 (4.1-5.6)
Type of BCPS	
Unilateral	27 (100)
Right BCPS	26 (96.3)
Left BCPS	1 (3.7)
Bilateral	0 (0.0)
CPB time (min)	69 (52-107)
Aortic crossclamp	5 (18.5)
Aortic crossclamp time (min)	18 (15-33)
Concomitant procedure	
PA reconstruction	11 (40.7)
AVV procedure	2 (7.4)
Atrioseptectomy	2 (7.4)
Pulmonary valve closure	1 (3.7)
Partial takedown	
On same day of BCPS	3 (48.1)
Concomitant with BCPS	11 (40.7)
Separately after BCPS	2 (7.4)
On separate day after BCPS	14 (51.9)
Interval after BCPS (d)	19 (5-34)
Shunt size (mm)	
3.0	4 (14.8)
3.5	18 (66.7)
4.0	3 (11.1)
5.0	2 (7.4)
Septation of central PA	
Clip	16 (59.3)
Ligation	8 (29.6)
Patch	3 (11.1)

Values are presented as n (%) or median (interquartile range). *BCPS*, Bidirectional cavopulmonary shunt; *CPB*, cardiopulmonary bypass; *PA*, pulmonary artery; *AVV*, atrioventricular valve.

was needed after partial takedown in the remaining 3 patients. Median duration of ECMO support was 10 days (IQR, 5-19 days; minimum, 4 days; and maximum, 43 days).

Development of the left PA was evaluated in 24 patients who underwent postpartial takedown cardiac catheterization at the median of 5.9 months (IQR, 1.4-15.1 months) after partial takedown (Table E2). Left PA index was significantly increased (from 79 ± 58 to 104 ± 77 mm^2/m^2; $P < .001$). Of the 12 patients with left PA hypoplasia, we had an angiogram after partial takedown in 11 patients. Their left PA index also increased (from 43 ± 21 to 54 ± 20 mm^2/m^2; $P = .159$), but was not statistically significant.

Follow-up Outcomes

For the 23 hospital survivors, the median follow-up time after partial takedown was 2.0 years (IQR, 0.7-5.9 years). There were 7 late deaths (interstage mortality between discharge after partial takedown and before Fontan completion) on 61, 73, 96, 181, 599, 720, and 967 days after partial takedown. HLHS and its variant were the cause of death in 6 patients. The causes of deaths were ventricular dysfunction in 5 patients, progressive PVO in 1 patient, and hypoxia due to high pulmonary vascular resistance in 1 patient. The remaining 16 patients were alive at a median of 5.0 years (IQR, 1.2-8.3 years) after partial takedown, 10 patients underwent Fontan procedure at the median age of 1.9 years (IQR, 1.4-5.1 years) and median interval of 1.7 years (IQR, 1.1-4.8 years) after partial takedown. Total cavopulmonary connection before cardiac catheterization (pre-TCPC) data are shown in Table E3. There were no deaths after Fontan completion at a median follow-up period of 5.8 years (IQR, 2.0-10.6 years). The remaining 6 patients were waiting for Fontan completion. Overall survival after partial takedown was 69% at 1 year and 54% at 3 years (Figure 4).

As for the surgical intervention for AVV regurgitation, 5 patients underwent AVV repair (3 tricuspid valve repair concomitant with BCPS, 1 mitral valve repair 524 days after partial takedown, and 1 tricuspid valve repair concomitant with TCPC). One patient, who needed tricuspid valve replacement after tricuspid valve repair concomitant with BCPS, died as a result of ventricular dysfunction. The remaining 4 patients survived without significant AVV regurgitation.

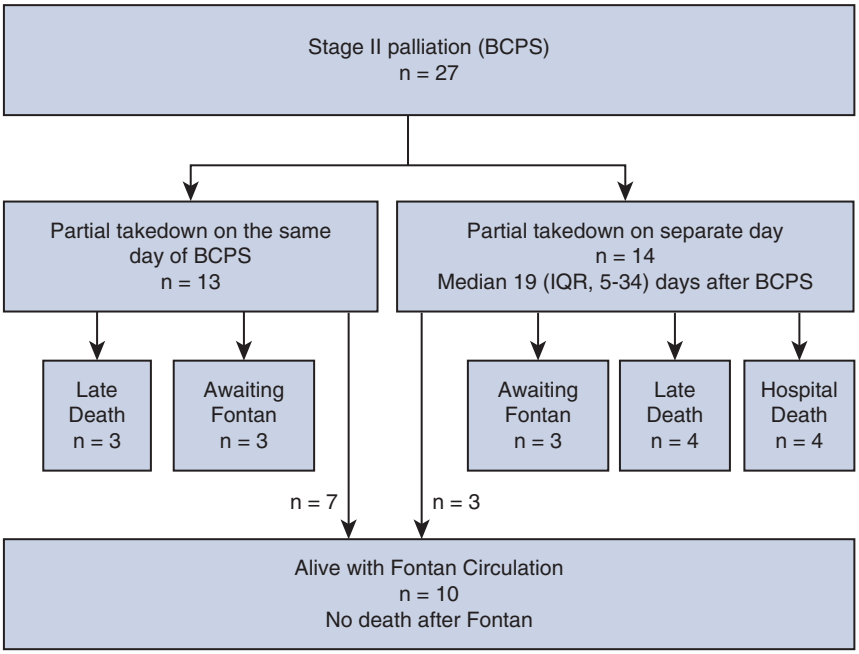


FIGURE 3. Flow chart of the 27 patients who underwent partial takedown. *BCPS*, Bidirectional cavopulmonary shunt; *IQR*, interquartile range.

Risk Factor Analysis for Mortality after Partial Takedown

Unbalanced atrioventricular septal defect (hazard ratio [HR], 4.913; $P = .026$), genetic/extracardiac anomalies (HR, 3.083; $P = .066$), and PVO (HR, 10.982; $P = .019$) showed P values $< .1$ for deaths after partial takedown (Table E4). Methods of PA interruption had no influence on outcomes.

DISCUSSION

Summary of the Results

The current study was undertaken to determine whether or not partial takedown was a reasonable procedure in 27 failing BCPS patients. The causes for partial takedown were: 10 left PA hypoplasia, 7 low SVC flow/high pulmonary vascular resistance, 3 early BCPS, 3 mediastinitis/pneumonia, 2 PVO, 1 pneumothorax, and 1 severe ventricular dysfunction. There were 4 hospital deaths and 7 interstage deaths. Survival at 3 years was 54%. Fontan completion was possible in 10 patients (Figure 1, Video 1, and Online Data Supplement).

Potential Therapeutic Options for Prohibitive Cyanosis after BCPS without Pathway Obstruction

In general, BCPS has a relatively low early mortality rate with survival to Fontan operation.[1,12-14] Nevertheless, certain patients develop significant hypoxemia and inadequate physiology after the procedure, even when the

TABLE 4. Causes of hypoxemia and outcomes of partial takedown

Group	n	Outcomes					
		ECMO	Thrombus	HD	LD	Fontan	Waiting Fontan
Partial takedown on the same day	13	0	0	0	3	7	2
LPA hypoplasia	9	0	0	0	3	5	1
High PVR/low SVC flow	4	0	0	0	0	2	2
Partial takedown on the different day	14	9	11	4	4	3	3
Early BCPS <90 d	3	1	2	0	0	1	2
Pulmonary venous obstruction	2	2	2	1	1	0	0
High PVR	4	3	4	2	1	1	0
TR and reduced VF	1	1	1	0	1	0	0
Mediastinitis and pneumonia	3	2	2	1	1	1	0
Pneumothorax	1	0	0	0	0	0	1

ECMO, Extracorporeal membrane oxygenation; *HD*, hospital death; *LD*, late death; *LPA*, left pulmonary artery; *PVR*, pulmonary vascular resistance; *SVC*, superior vena cava; *BCPS*, bidirectional cavopulmonary shunt; *TR*, tricuspid regurgitation; *VF*, ventricular function.

TABLE 5. Postoperative data in 14 patients with separate partial takedown (*PTD*)

Variable	Result
Timing of PTD	
Interval between BCPS and PTD (d)	19 (5-34)
Postoperative data	
ICU stay (d)	45 (30-81)
Hospital stay (d)	59 (42-101)
Hospital stay after PTD (d)	38 (18-60)
Possible causes of PTD	
High pulmonary vascular resistance	4 (28.6)
Early BCPS <90 d	3 (21.4)
Mediastinitis/pneumonia	3 (21.4)
Pulmonary venous obstruction	2 (14.3)
Systemic ventricular dysfunction	1 (7.1)
Repeat pneumothorax	1 (7.1)
Complications	
Reoperation with CPB	
Thrombectomy	4 (28.6)
BCPS pathway revision	2 (14.3)
AVV replacement	1 (7.1)
PV patch enlargement	1 (7.1)
Complete takedown	1 (7.1)
Reoperation without CPB	
Thoracic exploration	2 (14.3)
Pacemaker implantation	1 (7.1)
Diaphragm plication	1 (7.1)
Intervention	
Stent implantation in PA	4 (28.6)
v-v collateral coil closure	3 (21.4)
APCs coil closure	1 (7.1)
Complications	
Thrombus formation	12 (85.7)
Pleural effusion	7 (50.0)
Pneumothorax	5 (35.7)
Mediastinitis	4 (28.6)
Chylothorax	3 (21.4)
ECMO implantation	9 (64.3)

Values are presented as n (%) or median (interquartile range). *BCPS*, Bidirectional cavopulmonary shunt; *ICU*, intensive care unit; *CPB*, cardiopulmonary bypass; *AVV*, atrioventricular valve; *PV*, pulmonary vein; *PA*, pulmonary artery; *v-v*, veno-venous; *APC*, aorto pulmonary collaterals; *ECMO*, extracorporeal membrane oxygenation.

BCPS pathway is unobstructed. To rescue such patients, potential treatment options other than heart transplantation are complete takedown and partial takedown. Patients requiring complete takedown may be among the candidates at greatest risk for stage II palliation for single-ventricle anatomy. The mortality associated with complete takedown is high.[9,15,16] The concept of an additional aortopulmonary shunt to the targeted lung was reported by Sakamoto and colleagues in 2007.[17] They named this procedure intrapulmonary-artery septation and performed it in 20 patients with severe unbalanced PAs. Subsequently, they reported the efficacy of this technique in both unilateral PA hypoplasia[18] and pulmonary vein obstruction.[19] In 2018, Casella and colleagues[20] demonstrated the targeted increase of pulmonary blood flow at BCPS, and performed unilateral Glenn shunt, a systemic-to-pulmonary shunt in the contralateral lung, and a narrow banding between Glenn and systemic-to-pulmonary shunt in 20 patients with unilateral pulmonary vascular abnormalities. Recently, Turkoz and Dogan[21] reported 2 patients who underwent additional aortopulmonary shunt immediately after the BCPS procedure. In both cases, an aortopulmonary shunt was placed to the left lung and tight central pulmonary artery banding was performed between the BCPS and the aortopulmonary shunt. Our concept is nearly the same as this report. Although the reasons for performing partial takedown are different in this study, the rationale for partial takedown are the same: A unilateral Glenn shunt should be established; an additional customized aortopulmonary shunt may increase contralateral pulmonary blood flow to increase oxygen saturation. A complete septation of the central PA prevents SVC syndrome and provides adequate driving pressure to the affected PA. In this study, growth of affected PA was observed after partial takedown.

Surgical Options: Complete Takedown or Partial Takedown

We realized that the lack of a comparison group is a weakness of this study. In the study period, only 1 patient underwent complete takedown, and died 2 months postoperatively (mortality of 100%). In the literature, there are relatively scant descriptions of survival after complete takedown. Luo and colleagues[9] showed that 5 out of 7 (71.4%) patients died after complete takedown. Greenberg and colleagues demonstrated that 4 out of 6 (66.6%) patients died (n = 3) or needed heart transplantation (n = 1) after complete takedown.[22] We know that complete takedown is associated with substantial mortality, usually more than 50%. Additionally, we performed a total of 572 staged Fontan procedure between 1994 and 2021. There was no patient who experienced previous BCPS complete takedown. In the literature, we could not find a report where a successful Fontan procedure after BCPS complete takedown was described. Therefore, it can be said that partial takedown may be better tolerable than complete takedown and could keep good Fontan candidacy.

Our current determining criteria for a partial or complete takedown are as following: partial takedown is the first choice if unilateral Glenn shunt could be maintained. In these cases, additional systemic-to-pulmonary shunt improves hypoxemia, and partial takedown would be tolerable. Whereas complete takedown is indicated if the unilateral Glenn circulation could not be maintained, due to too high pulmonary vascular resistance, too-low SVC flow, or existence of PVO at Glenn side. Another option is the placement of larger systemic-to-pulmonary shunt and

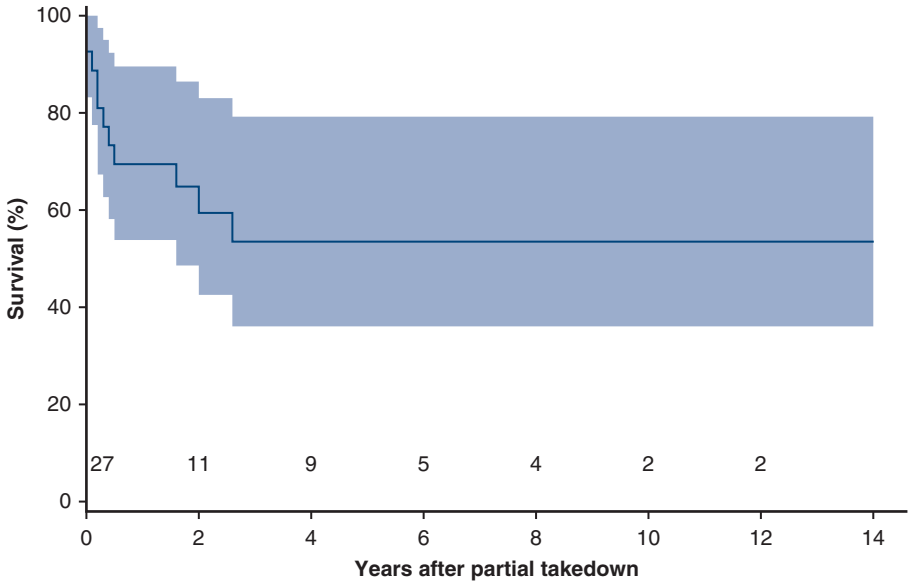


FIGURE 4. Transplant-free survival in patients after partial takedown. Kaplan-Meier estimate for freedom from death or transplantation after the partial takedown showed 53.5% survival at 3 years follow-up: The mean follow-up after the partial takedown was 1.8 days (interquartile range, 0.3-5.6 days). CI is 95%.

aggressive PA reconstruction. In patients with dominant left ventricle (such as tricuspid atresia or double inlet left ventricle) and good systemic ventricular function, this strategy might be the method of choice. However, in patients with dominant right ventricle, partial takedown might be indicated rather than upsizing the shunt. In contrast to complete takedown, partial takedown remains a step toward TCPC. All of our patients directly underwent TCPC following partial takedown without any additional procedure. In this study, 10 patients underwent Fontan palliation at a median of 1.7 years after partial takedown, and central PA could be reconstructed concomitantly in all patients.

Technical Aspect of Partial Takedown

After placement of an additional aortopulmonary shunt, whether or not to band or to septate the central PA is a great concern. When central PA is banded, a relatively small aortopulmonary shunt should be used as a protective measure against increased pressures in the Glenn pathway.[20] When central PA is interrupted, the unilateral Glenn works without any additional driving force.[17] A relatively large shunt can be placed to the affected lung. Either banding or septation is performed, this strategy might help patients with inadequate SVC flow to achieve reasonable saturation by providing an additional source of pulmonary blood flow.[23] The trade-off is an increased volume load on the systemic ventricle. However, apparent negative effects on systemic ventricular function were not observed in this study. Second concern is the shunt size. In this study, a 3.5-mm shunt was most commonly used. When central PA is interrupted, the shunt size can be chosen for the requirements of the affected PA. The amount of aortopulmonary collaterals flow to the affected lung, pulmonary vascular resistance, and size of the PA might be the decision-making components. As for interruption method, it was surgeon's preference and it did not affect the outcomes. Actually, we prefer to use clip because of the technical simplicity. As for the ECMO support, we used ECMO support as a bridge to weaning the decision for partial takedown in 6 out of 14 patients who underwent partial takedown on a separate day. For the remaining 8 patients, we decided directly to perform partial takedown without previous ECMO support.

VIDEO 1. The author explaining the importance and relevance of this study. Video available at: https://www.jtcvs.org/article/S2666-2736(22)00278-9/fulltext.

Etiology and Prognosis after Partial Takedown

There might be several etiologies of severe cyanosis after BCPS without pathway obstruction. One main reason is the left PA hypoplasia. In the present study, 12 patients had left

PA hypoplasia, which is a good indication for partial takedown. In patients with unilateral PA hypoplasia, shunt renewal (size-up) with extensive PA reconstruction is an alternative to perform early BCPS. In early era, we adopted this strategy. However, as the results of improved Norwood procedure and HLHS became the main diagnosis, we introduced early staged Fontan completion strategy because right ventricle failure and significant tricuspid regurgitation developed frequently after placement of a larger shunt. Younger age at BCPS is also a reason to demonstrate failing BCPS physiology. Patients who demonstrated mildly elevated pulmonary pressure are also good candidates for partial takedown. However, this approach does not seem to be effective in patients with PVO. The optimal treatment for patients with PVO remains unclear, and mortality remains high even after partial takedown. Genetic/extracardiac abnormalities were identified as another risk factor. In these patients, intrinsic pulmonary vascular disease associated with disorders might influence the BCPS physiology and postoperative outcome. Furthermore, we had patients who demonstrated mediastinitis/pneumonia after BCPS. These patients developed increased pulmonary vascular resistance and/or pulmonary venous desaturation. Another reason is low cardiac output due to ventricular dysfunction. We had 1 patient who required AVV replacement after BCPS and demonstrated severe systemic ventricular dysfunction.

When unilateral (usually right side) Glenn circulation can be maintained without SVC syndrome and an additional antegrade pulmonary blood flow in the contralateral (usually left) lung can increase the total pulmonary blood flow, partial takedown improves arterial oxygen saturation and rescues failing BCPS. If unilateral Glenn shunt could not be established due to too high pulmonary vascular resistance, SVC pressure remains high after partial takedown, and partial takedown could not rescue this situation. In this study, this was the case in patients with severe pulmonary vascular diseases with a genetic syndrome, patients with PVO, or patients with severe pneumonia. Complete takedown might be an alternative for these patients. If unilateral Glenn shunt could not establish due to too low SVC flow, partial takedown could not rescue this situation. In this study, this was the case in patients with severe AVV regurgitation and ventricular dysfunction. For these patients, heart transplantation or a new therapy such as assisted bidirectional Glenn may be more appropriate.[24]

The reasons why postoperative management was so complicated in patients who underwent partial takedown on a separate day are that patients' hemodynamic parameters, such as PAP, arterial oxygen saturation, and systemic arterial pressure, consistently changed during the postoperative days. It is not easy to decide whether or not BCPS hemodynamic parameters are established or not, and when partial takedown should be performed. Certain patients demonstrated relatively low arterial oxygen saturation during the early postoperative phase, and gradually increased their arterial oxygen saturation spontaneously. Effects of ECMO support and thrombus formation are also factors influencing the postoperative course. We routinely perform pulmonary angiography through central venous catheter on postoperative day 2 or 3 to rule out the technical problem (BCPS pathway obstruction). Therefore, it is still difficult to define the clear indication and management strategy of partial takedown. Further experiences and studies with large scale cohort are necessary to establish better management strategies for the treatment of failing BCPS patients.

In this study, patients who underwent immediate partial takedown showed better hospital survival. However, the reasons to perform partial takedown in late partial takedown group were different, so we cannot simply say that early timing to perform partial takedown is better.

Study Limitations

This study was limited by its retrospective, observational, and single-center design. This study was also limited by the heterogeneity of the patient cohort. Partial takedown was performed in different period after the initial BCPS. The indication for partial takedown was also different from patient to patient. Despite all efforts to conduct a large retrospective study, this study has inevitable limitations of small cohort size, no comparative group, and descriptive nature. Lack of any acceptable control group is also a significant limitation. No patients underwent complete takedown and so it is impossible to evaluate the merits or problems associated with this strategy. Another limitation was a lack of a quantifiable measurement of pulmonary blood flow. Given the recent application of this procedure, there was variable utilization of magnetic resonance imaging flow data pre- and postoperatively. Furthermore, certain patients underwent concomitant procedures on the primary vascular anomaly during preoperative catheterization and at the time of the operation, which inherently influences the outcome of this intervention. Because of the small size of cohort, results should be interpreted cautiously as only large effects could be identified. Finally, the follow-up period was not long enough to delineate the outcome after Fontan completion.

CONCLUSIONS

In 27 patients showing severe hypoxemia after BCPS without any stenosis at the BCPS anastomosis, partial takedown was performed to rescue the patients. There were 4 early and 7 late mortalities, and the 3-year survival was 54%. However, 10 patients underwent Fontan completion without postoperative mortality, and 6 patients were waiting for Fontan completion. Our results are encouraging in this inherently high-risk population. Future stratification of patients with intrinsic pulmonary vascular pathology and

sophistication of operative and postoperative management might improve outcomes as we continue to investigate this procedure.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

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Key Words: bidirectional cavopulmonary shunt, cyanosis, additional systemic-to-pulmonary-artery shunt, pulmonary artery hypoplasia, pulmonary venous obstruction, takedown

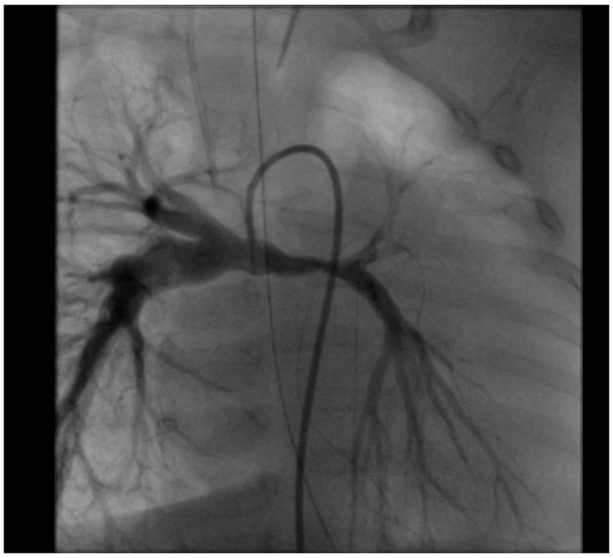

FIGURE E1. Pulmonary artery angiogram demonstrating left pulmonary artery stenosis before bidirectional cavopulmonary shunt.

TABLE E1. Postoperative outcome after partial takedown (*PTD*)

Patient No.	Diagnosis	ICU stay	Hospital stay	Hospital after PTD	ECMO	Thrombus	Chylothorax	SVC syndrome	Reoperation	TCPC	Age at TCPC	Interval after PTD	Outcome	Follow-up	Status
1	HLHS	6	15	15	0	0	0	1	1	1	3.5	3.1	Alive	14.0	fTCPC
2	DILV, TGA, IAA	6	16	16	0	0	0	0	0	1	0.9	0.5	Alive	12.5	fTCPC
3	HLHS	13	20	20	0	0	0	0	1	0			LD	0.5	
4	DORV, MS	42	42	42	0	0	0	0	1	0			LD	0.3	
5	TA Ib	6	18	18	0	0	0	0	0	1	1.5	1.4	Alive	9.9	TCPC
6	HLHS	12	29	29	0	0	0	0	0	0			Alive	4.5	Waiting
7	HLHS	16	30	30	0	0	0	0	0	0			LD	2.0	
8	HLHS	16	38	38	0	0	0	0	0	1	1.9	1.7	Alive	8.9	TCPC
9	DORV, MA	7	16	16	0	0	0	0	0	1	4.3	4.0	Alive	5.4	TCPC
10	HLHS, TAPVC	2	10	10	0	0	0	0	0	0			Alive	0.7	waiting
11	HLHS	7	34	34	0	0	1	0	0	0			Alive	0.7	waiting
12	HLHS	7	11	11	0	0	0	0	0	1	5.9	5.8	Alive	5.9	fTCPC
13	DIRV, TGA, PA	6	11	11	0	0	0	0	0	1	2.3	2.0	Alive	2.1	TCPC
14	HLHS, PAPVC	44	44	43	1	SVC, PA, PV	0	1	1	0			LD	0.2	
15	UAVSD, TGA, PA	161	161	158	1	SVC	0	1	1	0			HD	0.4	.
16	HLHS	16	16	12	1	PA	0	0	1	0			HD	0.0	
17	HLHS	32	118	113	1	SVC	1	0	1	0			Alive	6.6	Waiting
18	HLHS, TAPVC	15	25	16	0	0	0	0	0	0			Alive	0.0	Waiting
19	UAVSD	25	43	30	0	Aorta	0	0	0	0			LD	1.6	
20	HLHS	75	75	58	1	SVC	0	0	1	0			LD	0.2	.
21	PAIVS, Ebstein	40	40	19	1	0	0	0	1	1	5.9	5.5	Alive	5.6	TCPC
22	UAVSD, DORV, PS	63	63	40	1	RA	0	0	0	0			HD	0.1	.
23	TA Ia	39	55	26	1	PA	1	1	0	1	1.3	0.8	Alive	1.0	fTCPC
24	TA Ib	76	100	67	0	SVC	0	0	1	1	1.8	1.5	Alive	1.8	fTCPC
25	ccTGA, TA, IAA	46	46	11	1	SVC	0	0	1	0			HD	0.0	.
26	TA Ic	97	97	35	0	0	0	0	0	0			Alive	1.9	Waiting
27	HLHS	105	105	42	0	Aorta	0	0	1	0			LD	2.6	

ICU, Intensive care unit; *ECMO*, extracorporeal membrane oxygenation; *SVC*, superior vena cava; *TCPC*, total cavopulmonary connection; *HLHS*, hypoplastic left heart syndrome; *fTCPC*, fenestrated total cavopulmonary connection; *DILV*, double inlet left ventricle; *TGA*, transposition of the great arteries; *IAA*, interruption of the aorta; *LD*, late death; *DORV*, double outlet right ventricle; *MS*, multiple sclerosis; *TA*, tricuspid atresia; *MA*, mitral atresia; *TAPVC*, total anomalous pulmonary venous connection; *DIRV*, double inlet right ventricle; *PA*, pulmonary artery; *PAPVC*, partial anomalous pulmonary venous connection; *UAVSD*, unbalanced atrioventricular septal defect; *PAIVS*, pulmonary atresia with in intact ventricular septum; *Ebstein*, Ebstein anomaly; *PS*, pulmonary stenosis; *TA*, tricuspid atresia; *ccTGA*, congenitally corrected transposition of the great arteries.

TABLE E2. Characteristics of patients with regurging pulmonary arteries (PA)

Patient No.	Diagnosis	Stage I palliation	Shunt size	PA hypoplasia	LPA index pre BCPS	BCPS age (mon)	Shunt reoperation	LPA index after PTD	TCPC	Age at TCPC	Interval after PTD
1	HLHS	Norwood	5.0	LPA (2.6 mm)	38	5.0	CS 4.0	35	1	3.5	3.1
2	DILV, TGA, IAA	Norwood	4.0	LPA (3.2 mm)	43	3.7		53	1	0.9	0.5
3	HLHS	Norwood	5.0	LPA (2.5 mm)	27	5.1	BT 5.0	38	0		
4	DORV, MS	Norwood	3.5	LPA (1.2 mm)	31	3.8	BT 5.0	70	0		
5	TA Ib	no			279	2.2		271	1	1.5	1.4
6	HLHS	Norwood	3.0	LPA (1.5 mm)	12	4.6		30	0		
7	HLHS	Norwood	3.5	LPA (1.9 mm)	35	4.7		.	0		
8	HLHS	Norwood	3.0		76	3.1		81	1	1.9	1.7
9	DORV, MA	Norwood	3.5	LPA (3.0 mm)	29	4.2		71	1	4.3	4.0
10	HLHS, TAPVC	Norwood	3.5		86	5.1		111	0		
11	HLHS	Norwood	3.5		86	3.6		78	0		
12	HLHS	Norwood	3.5	LPA (2.0 mm)	35	1.8		28	1	5.9	5.8
13	DIRV, TGA PA	PDA stent	4.0	LPA (3.5 mm)	44	3.4		88	1	2.3	2.0
14	HLHS, PAPVC	Norwood	3.5		53	3.4		75	0		
15	UAVSD, TGA, PA	PDA stent	3.5		148	7.5		337	0		
16	HLHS	Norwood	3.5		66	4.5		86	0		
17	HLHS	Norwood	3.5	LPA (3.0 mm)	52	2.7	BT 5.0*	48	0		
18	HLHS, TAPVC	Norwood	3.5		85	2.8		.	0		
19	UAVSD	PAB			336	12.8		.	0		
20	HLHS	Norwood	5.0		82	4.3		94	0		
21	PAIVS, Ebstein	MBTS	3.5		98	4.1	CS 4.0	170	1	5.9	5.5
22	UAVSD, DORV, PS	MBTS	3.5		184	3.2		218	0		
23	TA Ia	PDA stent	4.0	LPA (2.7 mm)	88	5.2		74	1	1.3	0.8
24	TA Ib	PDA stent	3.5		66	2.9		141	1	1.8	1.5
25	ccTGA, TA, IAA	Norwood	3.5	LPA (2.6 mm)	72	4.1		61	0		
26	TA Ic	MBTS	3.5		107	4.1		145	0		
27	HLHS	Norwood	5.0		86	3.5		87	0		

LPA, Left pulmonary artery; *BCPS*, bidirectional cavopulmonary shunt; *PTD*, partial takedown; *TCPC*, total cavopulmonary connection; *HLHS*, hypoplastic left heart syndrome; *CS*, central shunt; *DILV*, double inlet left ventricle; *TGA*, transposition of the great arteries; *IAA*, interruption of the aorta; *BT*, Blalock-Taussig shunt; *DORV*, double outlet right ventricle; *MS*, multiple sclerosis; *TA*, tricuspid atresia; *MA*, mitral atresia; *TAPVC*, total anomalous pulmonary venous connection; *DIRV*, double inlet right ventricle; *PAPVC*, partial anomalous pulmonary venous connection; *UAVSD*, unbalanced atrioventricular septal defect; *PDA*, patent ductus arteriosus; *PAB*, pulmonary artery banding; *PAIVS*, pulmonary atresia with in intact ventricular septum; *Ebstein*, Ebstein anomaly; *PS*, pulmonary stenosis; *MBTS*, modified Blalock-Taussig shunt; *ccTGA*, congenitally corrected transposition of the great arteries. *Concomitant with pulmonary artery reconstruction.

TABLE E3. Total cavopulmonary connection (TCPC) before catheterization and echocardiographic data (n = 10)

Variable	Result
Catheterization data	
Hemoglobin (g/dL)	16.0 ± 15
Mean right pulmonary artery pressure (mm Hg)	10.5 ± 2.7
Mean left pulmonary artery pressure (mm Hg)	10.0 ± 1.9
Mean left atrial pressure (mm Hg)	5.2 ± 1.5
Transpulmonary gradient (mm Hg)	5.2 ± 2.1
Systolic ventricular pressure (mm Hg)	83.7 ± 12.1
Ventricular endo-diastolic pressure (mm Hg)	7.2 ± 1.7
Aortic oxygen saturation (%)	80.9 ± 5.3
Echocardiographic data	
Ventricular function	
Normal	10 (100.0)
Mildly impaired	0 (0.0)
Moderately impaired	0 (0.0)
Atrioventricular valve regurgitation	
None	1 (10.0)
Trivial	4 (40.0)
Mild	5 (50.0)
Moderate	0 (0.0)
Severe	0 (0.0)
(Neo) aortic insufficiency	
None	8 (80.0)
Trivíal Trivial	2 (20.0)
Mild	0 (0.0)

Values are presented as n (%) or mean ± SD.

TABLE E4. Variables influencing survival after partial takedown

Variable	Univariate model	
	P value	Hazard ratio (95% CI)
HLHS	.871	1.104 (0.3-3.6)
UAVSD	.026	4.913 (1.2-19.9)
Genetic/extracardiac anomaly	.066	3.083 (0.9-10.2)
Norwood procedure as stage I	.626	1.395 (0.4-5.3)
PAP	.271	1.092 (0.9-1.3)
PAP >15 mm Hg	.180	2.850 (0.6-13.2)
PVO	.019	10.982 (1.5-81.3)

CI, Confidence interval; *HLHS*, hypoplastic left heart syndrome, *UAVSD*, unbalanced atrioventricular septal defect, *PAP*, pulmonary artery pressure, *PVO*, pulmonary venous obstruction.