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

Caecilia Euringer, MS, Takashi Kido, MD, PhD, Bettina Ruf, MD, Melchior Burri, MD, PhD, Paul Philipp Heinisch, MD, PhD, Janez Vodiskar, MD, Martina Strbad, MSc, Bettina Ruf, MD, Melchior Burri, MD, PhD, Paul Philipp Heinisch, MD, PhD, Janez Vodiskar, MD, Bettina Ruf, MD, Melchior Burri, MD, PhD, Peter Ewert, MD, PhD, Jürgen Hörer, MD, PhD, and Masamichi Ono, MD, PhD

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)
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. 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. 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. 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.

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. 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. 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 bivacal cannulation, as described in our previous reports. A catheter to monitor central venous pressure was routinely inserted into the right internal jugular vein. Cardiopulmonary 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. 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.
Postoperative Management

As for the anticoagulation strategies, postoperative standard thrombosis prophylaxis after BCPS consisted of intravenous administration of unfractionated heparin (5000 IU/m²/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 χ² test was used for categorical data. Continuous variables are expressed as mean ± 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

| 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. |

| Partial takedown as a successful alternative to complete takedown in patients with failing BCPS |
|------------------------------------------------------------------------------------------------|
| Partial takedown on the same day of BCPS n = 13 | Partial takedown on separate day n = 14 Median 19 (IQR, 5-34) days after BCPS |
| Late Death n = 3 | Late Death n = 4 |
| Awaiting Fontan n = 3 | Awaiting Fontan n = 3 |
| Alive with Fontan Circulation n = 10 |
| No death after Fontan |

| Partial takedown |
|------------------|
| 1. Placement of an additional systemic to pulmonary shunt |
| 2. Septation of central pulmonary artery between Glenn anastomosis and shunt |
| 3. Of 16 survivors, 10 underwent Fontan completion with no mortality at median 6 years follow-up, and 6 were awaiting Fontan procedure. |

| Results |
|--------|
| 1. Most frequent diagnosis: Hypoplastic left heart syndrome (52%). |
| 2. 4 hospital deaths (14.8%), 7 late deaths (25.9%); Survival: 54% at 3 years |

| Management of failing BCPS: impact of partial takedown on outcome |
|------------------------------------------------------------------|
| Methods Analysis of 27 partial takedowns after failing BCPS (2007-2020) |

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Postoperative management of failing BCPS: impact of partial takedown on outcome

Methods Analysis of 27 partial takedowns after failing BCPS (2007-2020)
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 PAOP of 14.5 \pm 3.9 mm Hg, mean transpulmonary gradient of 7.4 \pm 2.7 mm Hg, systemic ventricular end-diastolic pressure of 9.5 \pm 2.9 mm Hg, and arterial oxygen saturation of 75.5\% \pm 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
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, 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 support 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

| 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                   | 5.0               | LPA (2.5 mm) | 27           | 5.1       | 60           | Con           | Hypo LPA     | Sano LPA         | 5.0            |
| 4          | DORV, MS  | Tetrakom     18p               | Norwood           | 3.5         | LPA (1.2 mm) | 31        | 3.8          | 56           | Con           | Hypo LPA         | BT LPA         | 3.5        |
| 5          | TA lb     | VACTERL                        | no                |            |              |           |              |              |              | Low SVC flow     | BT LPA         | 3.5        |
| 6          | HLHS      | Cleft palate                   | 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               |              |              |           |              |              |              | 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               |              |              |           |              |              |              | Low SVC flow     | CS LPA         | 3.5        |
| 11         | HLHS      | Norwood                        | 3.5               |              |              |           |              |              |              | 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               |              |              |           |              |              |              | PVO               | BT LPA         | 3.5        |
| 15         | UAVSD, TGA, PA | CHARGE         | PDA stent         | 3.5         |              |           |              |              |              | high PVR        | CS LPA         | 4.0        |
| 16         | HLHS      | Norwood                        | 3.5               |              |              |           |              |              |              | 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               |              |              |           |              |              |              | 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               |              |              |           |              |              |              | TR, reduced VF   | BT LPA         | 3.5        |
| 21         | PAIIS, Ebstein | MBTS                      | 3.5               |              |              |           |              |              |              | 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 lb     | PDA stent                      | 3.5               |              |              |           |              |              |              | 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 lc     | Renal anomalies                | MBTS             | 3.5         | 107         | 4.1       | 62           | 62           | Pneumothorax  | BT LPA         | 4.0            |
| 27         | HLHS      | Norwood                        | 5.0               |              |              |           |              |              |              | 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; PAR, 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.
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 to 104 mm²/m²; 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 to 54 mm²/m²; 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.

### 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          |
|---------------------------------|-----------------|
| **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)**              |                 |
| Aortic crossclamp               | 69 (52-107)     |
| 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)**             |                 |
| 5.0                             | 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.
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  | 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, atroventricular 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...
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. When central PA is interrupted, the unilateral Glenn works without any additional driving force. 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. 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.

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 take-
down. 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 intro-
duced 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 re-
mains high even after partial takedown. Genetic/extracar-
diac abnormalities were identified as another risk factor.
In these patients, intrinsic pulmonary vascular disease asso-
ciated 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 (usu-
ally 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 resis-
tance, 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 pulmo-
nary vascular diseases with a genetic syndrome, patients
with PVO, or patients with severe pneumonia. Complete
takedown might be an alternative for these patients. If uni-
ilateral 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 regur-
gitation and ventricular dysfunction. For these patients,
heart transplantation or a new therapy such as assisted bidirec-
tional 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 parame-
ters, such as PAP, arterial oxygen saturation, and systemic
arterial pressure, consistently changed during the postoper-
ative days. It is not easy to decide whether or not BCPS he-
modynamic parameters are established or not, and when
partial takedown should be performed. Certain patients
demonstrated relatively low arterial oxygen saturation dur-
ing 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 pa-
tient to patient. Despite all efforts to conduct a large retro-
spective 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 asso-
ciated 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 under-
went 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 take-
down 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 pa-
tients 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.
| Patient No. | Diagnosis | ICU stay | Hospital stay PTD | Hospital stay ECMO | Hospital stay Thrombus | Hospital stay Chylothorax | SVC syndrome | Reoperation | TCPC Age at | Outcome | Follow-up | Status |
|-------------|-----------|---------|------------------|-------------------|------------------------|--------------------------|--------------|-------------|------------|----------|-----------|--------|
| 1           | HLHS      | 6       | 15               | 15                | 0                      | 0                        | 0            | 1           | 1          | 3.5      | 3.1       | Alive  |
| 2           | DILV, TGA, IAA | 6     | 16               | 16                | 0                      | 0                        | 0            | 1           | 1          | 0.9      | 0.5       | Alive  |
| 3           | HLHS      | 13      | 20               | 20                | 0                      | 0                        | 0            | 1           | 1          | 1.0      | 1.0       | LD     |
| 4           | DORV, MS  | 42      | 42               | 42                | 0                      | 0                        | 0            | 1           | 1          | 4.3      | 4.0       | Alive  |
| 5           | TA Ia     | 6       | 18               | 18                | 0                      | 0                        | 0            | 0           | 1          | 1.5      | 1.4       | Alive  |
| 6           | HLHS      | 12      | 29               | 29                | 0                      | 0                        | 0            | 0           | 0          | Alive    | 4.5       | Waiting |
| 7           | HLHS      | 16      | 30               | 30                | 0                      | 0                        | 0            | 0           | 0          | LD       | 2.0       |          |
| 8           | HLHS      | 16      | 38               | 38                | 0                      | 0                        | 0            | 0           | 1          | 1.9      | 1.7       | Alive  |
| 9           | DORV, MA  | 7       | 16               | 16                | 0                      | 0                        | 0            | 0           | 1          | 4.3      | 4.0       | Alive  |
| 10          | HLHS, TAPVC | 2     | 10               | 10                | 0                      | 0                        | 0            | 0           | 0          | Alive    | 0.7       | waiting |
| 11          | HLHS      | 7       | 34               | 34                | 0                      | 0                        | 0            | 1           | 0          | Alive    | 0.7       | waiting |
| 12          | HLHS      | 7       | 11               | 11                | 0                      | 0                        | 1            | 0           | 0          | Alive    | 0.7       | waiting |
| 13          | DRV, TGA, PA | 6     | 11               | 11                | 0                      | 0                        | 0            | 1           | 1          | 5.9      | 5.8       | Alive  |
| 14          | HLHS, PAPVC | 44    | 44               | 43                | SVC, PA, PV             | 0                        | 1            | 1           | 0          | 2.3      | 2.0       | Alive  |
| 15          | UAVSD, TGA, PA | 161  | 161              | 158               | SVC                     | 0                        | 1            | 1           | 0          | HD       | 0.4       |        |
| 16          | HLHS      | 16      | 16               | 12                | PA                      | 0                        | 0            | 0           | 1          | LD       | 0.2       |        |
| 17          | HLHS      | 32      | 118              | 113               | SVC                     | 1                        | 0            | 1           | 0          | Alive    | 6.6       | Waiting |
| 18          | HLHS, TAPVC | 15    | 25               | 16                | 0                      | 0                        | 0            | 0           | 0          | Alive    | 0.0       | Waiting |
| 19          | UAVSD     | 25      | 43               | 30                | Aorta                   | 0                        | 0            | 0           | 0          | LD       | 1.6       |        |
| 20          | HLHS      | 75      | 75               | 58                | SVC                     | 0                        | 0            | 1           | 0          | LD       | 0.2       |        |
| 21          | PAIVS, Ebstein | 40    | 40               | 19                | 1                      | 0                        | 0            | 0           | 1          | 5.9      | 5.5       | Alive  |
| 22          | UAVSD, DORV, PS | 63   | 63               | 40                | RA                      | 0                        | 0            | 0           | 0          | HD       | 0.1       |        |
| 23          | TA Ia     | 39      | 55               | 26                | PA                      | 1                        | 1            | 0           | 1          | 1.3      | 0.8       | Alive  |
| 24          | TA Ib     | 76      | 100              | 67                | SVC                     | 0                        | 0            | 1           | 1          | 1.8      | 1.5       | Alive  |
| 25          | ccTGA, TA, IAA | 46   | 46               | 11                | SVC                     | 0                        | 0            | 1           | 0          | HD       | 0.0       |        |
| 26          | TA Ic     | 97      | 97               | 35                | Aorta                   | 0                        | 0            | 0           | 0          | Alive    | 1.9       | Waiting |
| 27          | HLHS      | 105     | 105              | 42                | 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; DRV, double inlet right ventricle; PA, pulmonary artery; PAPVC, partial anomalous pulmonary venous connection; UAVSD, unbalanced atrioventricular septal defect; PAIVS, pulmonary atresia with 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 regurgiting pulmonary arteries (PA)**

| Patient No. | Diagnosis               | Stage I palliation | Shunt size | PA hypoplasia | LPA index | 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                | 0.9                 | 0.5  | 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 lb                   | 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                | 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                | 5.9                 | 5.8  |             |                   |
| 13          | DIRV, TGA PA            | PDA stent          | 4.0        | LPA (3.5 mm)  | 44        | 3.4            | 88                | 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               | 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                | 1.3                 | 0.8  |             |                   |
| 24          | TA lb                   | 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 an intact ventricular septum; 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-diastatic 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)             |
| Trivial 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                      | P value | Hazard ratio (95% CI) |
|-------------------------------|---------|-----------------------|
| HLHS                          | .871    | 1.104 (0.3-3.6)       |
| UA VSD                        | .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; UA VSD, unbalanced atrioventricular septal defect; PAP, pulmonary artery pressure; PVO, pulmonary venous obstruction.