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Mid-term outcomes of a catheter-based right ventricular decompression strategy for pulmonary atresia with intact ventricular septum and critical pulmonary stenosis

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Abstract

Pulmonary atresia with intact ventricular septum (PAIVS) and critical pulmonary stenosis (cPS) span a spectrum of right ventricular hypoplasia in which the optimal initial pathway remains uncertain. We describe mid-term outcomes of an anatomy-guided percutaneous transcatheter pulmonary valvuloplasty (PTPV)-first strategy. We retrospectively reviewed 24 patients treated invasively from 2007 through 2025; three lacked measurable right ventricular end-diastolic volume (RVEDV) and were excluded, leaving an analytic cohort of 21 patients (PAIVS, n = 9; cPS, n = 12). Fifteen patients with a tripartite right ventricle and no major sinusoidal communication underwent PTPV first; six with an absent outlet component or major sinusoidal communication entered a single-ventricle pathway. All 15 PTPV-first patients achieved biventricular circulation; all three PAIVS patients in this group required subsequent, elective surgical right ventricular decompression, consistent with staged rather than failed decompression. All six single-ventricle patients completed Fontan circulation, and no PTPV-first patient required one-and-a-half ventricular repair or crossed over to Fontan. All 21 patients were alive after a median follow-up of 8 years [interquartile range, 6–11]. One patient with borderline RVEDV and tricuspid valve Z-score developed protein-losing enteropathy 11 years after Fontan completion. PTPV-first patients tended to have higher RVEDV and less negative tricuspid valve Z-scores than single-ventricle patients, reflecting selection criteria rather than validated thresholds. PTPV first permitted staged right ventricular recruitment and biventricular circulation with favorable mid-term outcomes, with surgical decompression as an effective subsequent step. Given the small, retrospective, single-center design, these findings are hypothesis-generating and warrant prospective reassessment of borderline right ventricles.

Introduction

Pulmonary atresia with intact ventricular septum (PAIVS) is characterized by complete obstruction of the right ventricular outflow tract without a ventricular septal defect and encompasses a broad range of right ventricular (RV), tricuspid valve (TV), and coronary morphologies. Critical pulmonary stenosis (cPS) represents an adjacent physiological spectrum in which severe valvar obstruction may produce duct-dependent pulmonary blood flow and varying degrees of RV hypoplasia. In both conditions, the initial intervention must address pulmonary blood flow while anticipating whether the RV can eventually support the entire pulmonary circulation [1–4].

Potential final pathways include biventricular repair (BVR), one-and-a-half ventricular repair (1.5VR), and single-ventricle palliation culminating in Fontan circulation. TV size, RV volume and function, and coronary anatomy—including right ventricle-dependent coronary circulation—have all been associated with pathway selection and outcome [1–4]. Nevertheless, treatment remains highly center dependent, and no single numerical threshold reliably separates patients who should undergo RV decompression from those who should enter a single-ventricle pathway [1, 2, 5–7].

Catheter-based pulmonary valve perforation followed by balloon dilation is an established initial option for anatomically suitable PAIVS, and balloon pulmonary valvuloplasty is the standard initial intervention

for cPS. Decompression of the RV by catheter-based intervention is safer and less invasive than surgical right ventricular outflow tract reconstruction; additional procedures may or may not be required to achieve biventricular circulation, and one-and-a-half ventricular repair is selected as the final pathway when they are not [8–13].

At our institution, the decision to attempt percutaneous transcatheter pulmonary valvuloplasty (PTPV)—including valve perforation followed by balloon dilation for membranous PAIVS—has been based primarily on qualitative anatomy: a tripartite RV and the absence of major sinusoidal communications (SCs). RV end-diastolic volume (RVEDV) and TV Z-score were not used as fixed prospective thresholds. This study aimed to describe pathway completion and mid-term outcomes under this anatomy-guided PTPV-first strategy and to explore the pretreatment distribution of RVEDV and TV Z-score according to the final circulation.

Patients and Methods

Study design and participants

This retrospective, single-center cohort study included patients with PAIVS or cPS who underwent invasive treatment at Saitama Medical University International Medical Center between January 2007 and December 2025. The study was performed in accordance with the Declaration of Helsinki and is reported in accordance with the Strengthening the Reporting of Observational Studies in Epidemiology recommendations [14]. The Institutional Review Board of Saitama Medical University International Medical Center approved the study (May 13, 2026; I2026-022). The requirement for individual written informed consent was waived, and information about the study was provided through an institutional opt-out process.

PAIVS was defined by complete valvar or muscular pulmonary atresia with an intact ventricular septum. cPS was defined as severe valvar pulmonary stenosis presenting in infancy with critical obstruction, cyanosis or duct-dependent pulmonary blood flow, and a requirement for invasive relief of obstruction.

During the study period, 24 patients with PAIVS or cPS underwent invasive treatment. Three patients lacked angiograms from which RVEDV could be measured and were excluded from the prespecified anatomical cohort. The remaining 21 patients comprised the main analytic cohort.

Institutional treatment strategy

The institutional strategy is summarized in Fig. 1. Patients were considered for a PTPV-first approach when the RV was tripartite, with identifiable inlet, trabecular, and outlet components, and when major SCs were absent. In cPS, PTPV consisted of balloon pulmonary valvuloplasty. In PAIVS with membranous pulmonary atresia, the atretic valve was perforated and then dilated.

Major SC was an angiographically prominent RV-to-coronary communication judged to make RV decompression unsafe or unlikely to be effective. Patients with an absent outlet component or major SC entered a single-ventricle pathway. In the PTPV-first group, inadequate RV decompression, insufficient antegrade pulmonary blood flow, or inadequate RV growth prompted surgical RV outflow decompression. The final pathway was determined by serial assessment of RV and TV growth, coronary anatomy, oxygenation, and hemodynamics. BVR was defined as a circulation in which the RV supported the entire pulmonary blood flow without a superior cavopulmonary connection.

Anatomical and outcome assessment

Baseline variables were abstracted from the earliest pretreatment echocardiographic and catheterization studies available for pathway assessment. TV diameter and its Z-score were obtained from the contemporaneous echocardiographic record. RV morphology and SCs were assessed from echocardiography and angiography. RVEDV was abstracted from the catheterization record, in which it had been estimated from monoplane right ventriculography and expressed as a percentage of the body-surface-area-predicted normal value, calculated using normative monoplane angiographic reference data from the pediatric cardiac catheterization literature [15]; although this normative dataset predates the study period, it remains the standard reference for monoplane angiographic RV volume normalization in the PAIVS/cPS literature, as contemporary normative data are derived from three-dimensional echocardiography or cardiac magnetic resonance imaging and are not directly interchangeable with monoplane angiographic measurements. Because this retrospectively recorded measure depends on geometric assumptions and no reproducibility analysis was available, RVEDV was treated as an exploratory anatomical marker rather than a validated selection criterion.

Outcomes were final circulation (BVR, 1.5VR, or Fontan), requirement for surgical RV decompression after PTPV, survival, follow-up duration, latest B-type natriuretic peptide (BNP), clinically documented heart failure or cyanosis after BVR, oxygen requirement beyond infancy, residual interatrial shunt after BVR, and Fontan-associated complications. Follow-up was determined from the latest clinical assessment available through 2026. Residual interatrial shunt after BVR was assessed by transthoracic echocardiography at the latest available follow-up visit and was recorded qualitatively for presence and shunt direction. The PLE case was examined descriptively; no causal inference regarding the initial pathway was made.

Statistical analysis

Continuous variables are presented as median [interquartile range], and categorical variables as number (percentage). Analyses were descriptive. Formal between-group hypothesis testing was not performed because pathway groups were intentionally defined by nonexchangeable anatomical features and the sample was small. RVEDV and TV Z-score analyses, including the visual reference lines at RVEDV 60% of predicted normal and TV Z-score of -3, were post hoc and exploratory. Analyses were performed using R version 4.6.1 (R Foundation for Statistical Computing, Vienna, Austria).

Results

Cohort formation and baseline anatomy (Fig. 2)

The analytic cohort included 9 patients with PAIVS and 12 with cPS. Median body weight was 3.00 [2.75–3.21] kg. Fifteen patients had a tripartite RV and no major SC and were selected for PTPV first; this group comprised 12 patients with cPS and 3 with PAIVS. Six patients, all with PAIVS, entered a single-ventricle pathway because of an absent outlet component, major SC, or both. Baseline anatomy is summarized in Table 1.

Table 1
Baseline characteristics according to the initial institutional pathway

Characteristic	Overall	PTPV-first/BVR	Single-ventricle/Fontan
Patients, n	21	15	6
PAIVS	9 (43%)	3 (20%)	6 (100%)
Critical pulmonary stenosis	12 (57%)	12 (80%)	0
Assessment during neonatal period	17 (81%)	12 (80%)	5 (83%)
Body weight, kg	3.00 [2.75–3.21]	3.12 [2.88–3.56]	2.64 [2.27–3.07]
Tripartite RV	16 (76%)	15 (100%)	1 (17%)
Major sinusoidal communication	4 (19%)	0	4 (67%)
Tricuspid valve Z-score	−1.31 [− 2.46 to − 0.57]	−0.78 [− 1.42 to − 0.57]	−5.00 [− 6.00 to − 3.34]
RVEDV, % of predicted normal	76 [69–97]	92 [75.5–104]	40.5 [34.8–53.0]

Values are number (%) or median [interquartile range]. BVR, biventricular repair; PAIVS, pulmonary atresia with intact ventricular septum; PTPV, percutaneous transcatheter pulmonary valvuloplasty; RV, right ventricle; RVEDV, right ventricular end-diastolic volume.

Regarding the three out of 24 patients who underwent invasive treatment during the study period but not included in this study because of lack of RVEDV measurement, one later achieved BVR and was alive at the latest available assessment, one underwent a bidirectional Glenn procedure and died before Fontan completion, and one had insufficient follow-up information to determine the final circulation.

Treatment course and pathway completion (Fig. 2)

All 15 PTPV-first patients ultimately achieved BVR. All 12 patients with cPS reached BVR without surgical RV outflow decompression. Each of the three patients with PAIVS required additional surgical decompression after catheter intervention: two underwent a closed Brock procedure and one underwent RV outflow tract reconstruction. Thus, the initial catheter-based procedure did not achieve complete

standalone decompression in the PAIVS subgroup, but it preserved a staged route to BVR. No patient underwent 1.5VR.

All six patients assigned to the single-ventricle pathway completed Fontan circulation.

Mid-term outcomes (Table 2)

Table 2
Treatment course and mid-term outcomes

Outcome	Overall	PTPV-first	Single-ventricle
Biventricular circulation	15 (71%)	15 (100%)	0
Additional surgical RV decompression after PTPV	3 (14%)	3 (20%)	Not applicable
Closed Brock procedure	2 (10%)	2 (13%)	Not applicable
Surgical RVOT reconstruction	1 (5%)	1 (7%)	Not applicable
One-and-a-half ventricular repair	0	0	0
Fontan completion	6 (29%)	0	6 (100%)
Alive at latest follow-up	21 (100%)	15 (100%)	6 (100%)
Follow-up, years	8 [6–11]	7 [4–8.5]	11.5 [9–14]
Latest BNP, pg/mL	10.0 [7.9–30.8] (n = 17)	9.9 [6.2–31.9] (n = 11)	18.8 [10.3–28.2] (n = 6)
Residual interatrial shunt after BVR	11 (73%)	11 (73%)	Not applicable
Protein-losing enteropathy	1 (5%)	0	1 (17%)

Values are number (%) or median [interquartile range]. BVR, biventricular repair; BNP, B-type natriuretic peptide; PTPV, percutaneous transcatheter pulmonary valvuloplasty; RV, right ventricle; RVOT, right ventricular outflow tract.

All 21 patients were alive at the latest assessment after a median follow-up of 8 [6–11] years. Follow-up was 7 [4–8.5] years in the PTPV-first/BVR group and 11.5 [9–14] years in the Fontan group. Latest BNP was available in 17 patients and was 10.0 [7.9–30.8] pg/mL overall. No BVR patient had documented resting cyanosis or a requirement for home oxygen beyond infancy; one had clinically recorded exercise limitation. In none of the cases was tricuspid regurgitation (TR-PG > 35 mmHg) observed [16]. Among the 15 patients who achieved BVR, a residual interatrial shunt was present in 11 (73%); all demonstrated left-to-right shunting, and none had associated cyanosis.

One Fontan patient developed protein-losing enteropathy (PLE) at 18 years of age, 11 years after Fontan completion. The earliest available anatomical assessment showed RVEDV of 57% of predicted normal, a TV Z-score of − 1.6, absence of major SC, and absence of the RV outlet component. The patient had

undergone two systemic-to-pulmonary shunt procedures by 1 month of age at the prior institution, hemi-Fontan at 1.5 years, and a fenestrated lateral-tunnel Fontan at 7 years in our center. Subsequently, due to the development of PLE, a Fontan conversion to an extra-cardiac total cavopulmonary connection was performed at age 19. Before Fontan conversion, pulmonary vascular resistance index was 0.78 WU·m², pulmonary artery pressure was 13 mmHg, and pulmonary artery wedge pressure was 10–11 mmHg with significant cyanosis. After conversion, the corresponding values were 0.81 WU·m², 8 mmHg, and 6 mmHg, respectively, with improving cyanosis. Four months have passed since then, and serum albumin has been maintained at around 3.0 g/dL.

Post hoc anatomical distribution (Fig. 3)

In the PTPV-first/BVR group, median RVEDV was 92 [75.5–104] % of predicted normal and median TV Z-score was − 0.78 [− 1.42 to − 0.57]. The minimum observed values were 69% of RVEDV predicted normal and − 2.46 of TV Z-score. In the Fontan group, median RVEDV was 40.5 [34.8–53.0] % of predicted normal and median TV Z-score was − 5.00 [− 6.00 to − 3.34]. All Fontan group patients had less than 60% of RVEDV predicted normal, except for one patient with major SC. Similarly, all Fontan group patients had less than − 3 of TV Z-score except for one who later developed protein-losing enteropathy after Fontan completion as described above.

Discussion

This study describes a 19-year single-center experience with an anatomy-guided PTPV-first strategy across the PAIVS/cPS spectrum. The principal findings were that all 15 patients who were anatomically suitable for PTPV to decompress RV and increase forward pulmonary blood flow eventually achieved biventricular circulation without any mortality. Of those, all three patients with PAIVS required subsequent surgical RVOT procedure to relieve residual pulmonary stenosis, whereas none of the 12 patients with cPS did. Retrospectively, patients selected for initial PTPV tended to have higher RVEDV and less negative TV Z-score than those directed to a single-ventricle pathway, consistent with the anatomical criteria used for selection rather than independently validated thresholds. The clinically relevant feature of this strategy is that PTPV was treated as the first step in RV recruitment rather than as a requirement for complete definitive decompression. Contemporary catheter-based RV decompression series support this staged framework (Table 3) [6, 10, 11, 13], with broadly comparable final-pathway and survival ranges.

Although all three patients with PAIVS required subsequent surgical RVOT procedure, initial perforation and balloon dilation achieved measurable RV pressure decompression and increased antegrade pulmonary flow in each patient; none required emergency conversion to surgery or sustained a procedure-related complication. The later closed Brock procedure or RVOT reconstruction was performed electively, after observed RV and TV growth, for residual infundibular or valvar stenosis rather than for a failed initial procedure. This pattern is consistent with the reintervention rates reported across

comparator series (Table 3). We interpret surgery in our three patients as an anticipated second stage rather than a rescue for technical failure.

Table 3: Comparison of catheter-based right ventricular decompression series.

Authors (published year)	Cohort (n)	PTPV attempt (%)	Surgical reintervention (%)	Final circulation (%)	Survival (%)	F/U (years)
Schwartz et al. (2014) [10]	23 PAIVS	23/23 (100%)	9/21† (42%)	BVR 18/19‡ (95%)	No deaths	3.9 §
Rathgeber et al. (2017) [11]	18 PAIVS	17/18 (94%)	1/18¶ (6%)	BVR 12/17†† (71%) 1.5VR 2/17 (12%) Fontan 2/17 (12%)	18/18 (100%)	4.9
Morgan et al. (2020) [13]	71 PAIVS	48/71# (68%)	36/45 (80%, any)	BVR 28/38# (74%) 1.5VR 9/38 (24%) Fontan 1/38 (3%)	40/48 (83%)	11.3
Manhem et al. (2025) [6]	67 screened	18/21 (86%) *	11/18 (61%)	BVR 16/18 (89%) 1.5VR 2/18 (11%)	18/18 (100%)	9
Present study	21 analytic	15/21 (71%)	3/15 (20%)	BVR 15/21 (71%) Fontan 6/21 (29%)	21/21 (100%)	8

1.5VR, one-and-a-half ventricular repair; BVR, biventricular repair; F/U, follow-up; PAIVS, pulmonary atresia with intact ventricular septum; PTPV, percutaneous transcatheter pulmonary valvuloplasty.
†Excludes 2 patients with <1-month follow-up (n=21). ‡Excludes 4 patients with insufficient follow-up (n=19); the other 1/19 had persistent right-to-left shunting (87% saturation), not meeting the BVR definition. §Mean ± SD, not median. ¶Emergency surgery for procedural complication only. #Of 71 total, 48 attempted PTPV and 45 succeeded; 7 of those 45 died, leaving the 38 survivors shown here. ††Of 18 attempted, 17 succeeded; the 1 failed case went to emergency surgery and is not included here. *Three failed mechanical perforations were reassigned to the surgical group.

A second notable feature of our cohort is that 1.5VR was not selected in all patients, in contrast to the 11–24% 1.5VR rates reported in three comparator series (Table 3) [6, 11, 13]. Small RV size or TV diameter may identify patients unlikely to reach BVR, but published thresholds vary across eras, imaging methods, and treatment strategies [1–7]. All 15 patients who were treated by PTPV first strategy went on to BVR and have been free from RV failure during the entire follow-up period. In contrast, the anatomical features that guided assignment to the single-ventricle pathway appeared appropriate in all patients, each of whom had a diminutive tricuspid valve, markedly reduced RVEDV, major SC, or a combination of these findings that would have made RV decompression unsafe or physiologically inadequate, but one, who later developed PLE, had borderline RVEDV and TV Z-score. He was referred to our center after two neonatal shunt operations, then lack of outlet portion did not allow BVR or even 1.5VR. Although PLE could occur late after 1.5VR due to high venous angle pressure along with high SVC pressure, where thoracic duct drains, surgical right ventricular outflow tract reconstruction combined with additional procedure to increase RV cavity or improve TV competency and subsequent 1.5VR or BVR may avoid such a life-threatening late Fontan-related complication [17].

Long-term outcome after PAIVS is determined not only by the selected pathway but also by early and interstage hazards. A large registry study reported excellent survival among patients who reached

definitive repair, regardless of whether the final circulation was biventricular, one-and-a-half ventricular, or Fontan, while overall survival remained limited by early events [18]. Our 100% survival figure applies only to the 21 patients with measurable RVEDV and should not be interpreted as the survival of every invasively treated patient at the institution: one of the three patients excluded because of unavailable RVEDV died before Fontan completion and another lacked definitive follow-up. This transparent distinction is essential when comparing cohorts.

Study Limitations

First, this was a small, retrospective, single-center study without a concurrent comparison strategy, and treatment allocation was determined by anatomy; causal or comparative conclusions cannot be drawn. Second, the study combined PAIVS and cPS. Although these lesions form a clinical spectrum, cPS generally has more favorable RV morphology, and 12 of 15 PTPV-first patients had cPS. Diagnosis-specific outcomes are therefore reported, and the finding that all three PAIVS patients required surgery should not be obscured by the combined result. Third, RVEDV was obtained from retrospective monoplane angiographic records, TV Z-scores were derived from contemporaneous reports, and no interobserver reproducibility analysis was available. Finally, objective exercise testing, quality-of-life assessment, arrhythmia surveillance, and standardized longitudinal RV functional imaging were not routinely available.

Conclusion

In conclusion, an anatomy-guided PTPV-first strategy achieved biventricular circulation with favorable mid-term outcomes in all anatomically selected patients, with surgical RV outflow decompression serving as an effective subsequent step rather than a treatment failure when required.

Abbreviations

1.5VR = one-and-a-half ventricular repair

BVR = biventricular repair

cPS = critical pulmonary stenosis

PAIVS = pulmonary atresia with intact ventricular septum

PTPV = percutaneous transcatheter pulmonary valvuloplasty

RV = right ventricle

RVEDV = right ventricular end-diastolic volume

SC = sinusoidal communication

TV = tricuspid valve

Declarations

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Competing interests

The authors have no relevant financial or non-financial interests to disclose.

Author contributions

Kaito Sano conceived the study, collected and curated the data, performed the descriptive analysis, prepared the figures, and drafted the manuscript. Koichi Toda, Taisuke Nabeshima, Kota Nagaoka, and Takuro Kojima contributed to clinical data acquisition and interpretation. Akinori Hirano and Ryusuke Hosoda contributed surgical data and interpretation. Takaya Hoashi supervised the study, interpreted the findings, and critically revised the manuscript. All authors reviewed and approved the final manuscript.

Ethics approval

This study was performed in line with the principles of the Declaration of Helsinki. Approval was granted by the Institutional Review Board of Saitama Medical University International Medical Center (May 13, 2026; I2026-022).

Consent to participate

The requirement for individual written informed consent was waived. Information about the study was provided through an institutional opt-out process.

Consent to publish

Not applicable. The manuscript contains no directly identifying participant information or clinical photographs.

Data availability

The de-identified data supporting the findings of this study are available from the corresponding author upon reasonable request, subject to institutional and ethical restrictions.

Code availability

The descriptive calculations used for this study are available from the corresponding author upon reasonable request.

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Figures

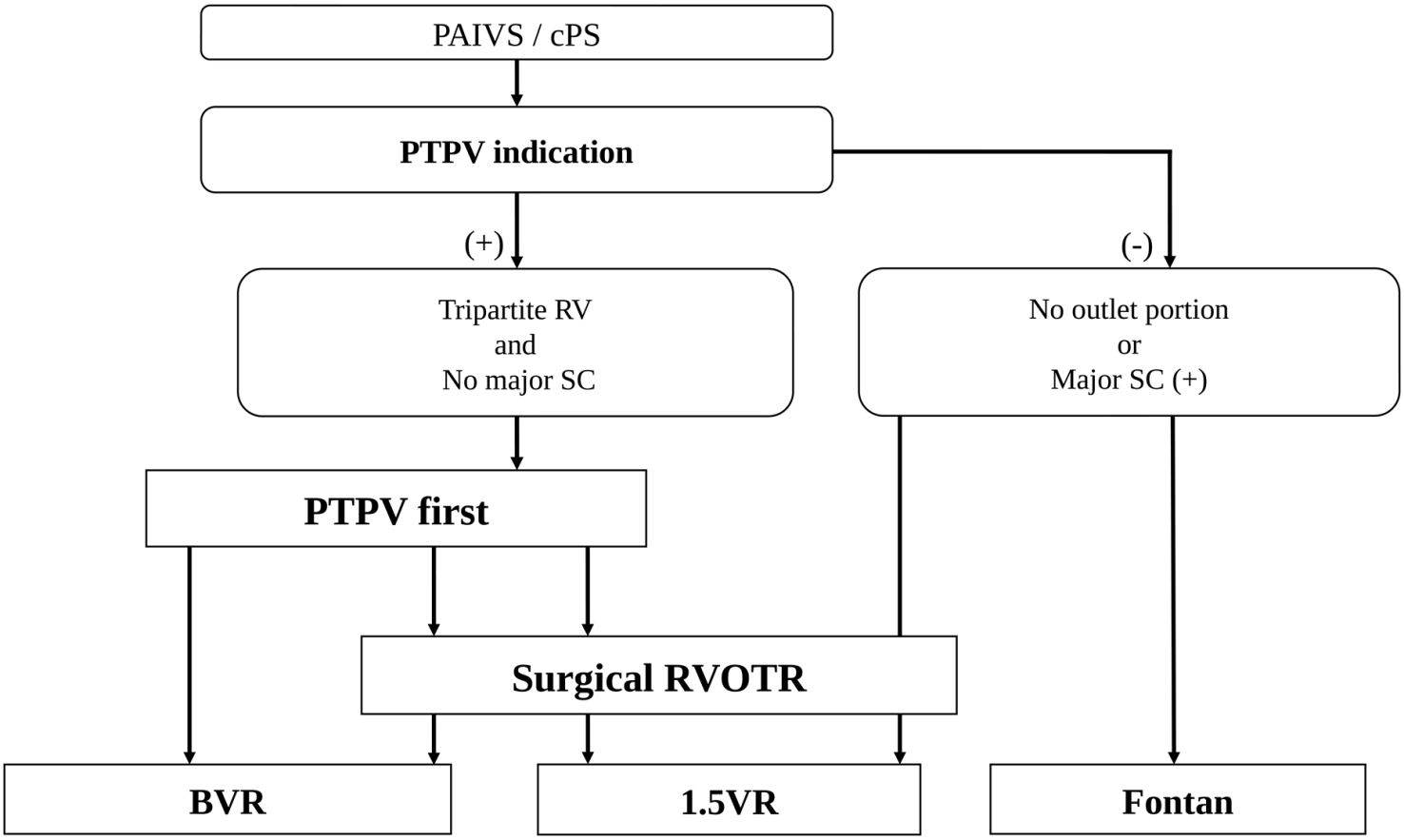


Figure 1

Institutional anatomy-guided treatment strategy. PTPV was attempted when the RV was tripartite and major SCs were absent. Inadequate catheter-based decompression prompted surgical RV outflow decompression, followed by reassessment of the final pathway. RVEDV and TV Z-score were analyzed retrospectively and were not prospectively defined selection thresholds. BVR, biventricular repair; cPS, critical pulmonary stenosis; PAIVS, pulmonary atresia with intact ventricular septum; PTPV, percutaneous transcatheter pulmonary valvuloplasty; RV, right ventricle; SC, sinusoidal communication; RVEDV, right ventricular end-diastolic volume; TV, tricuspid valve

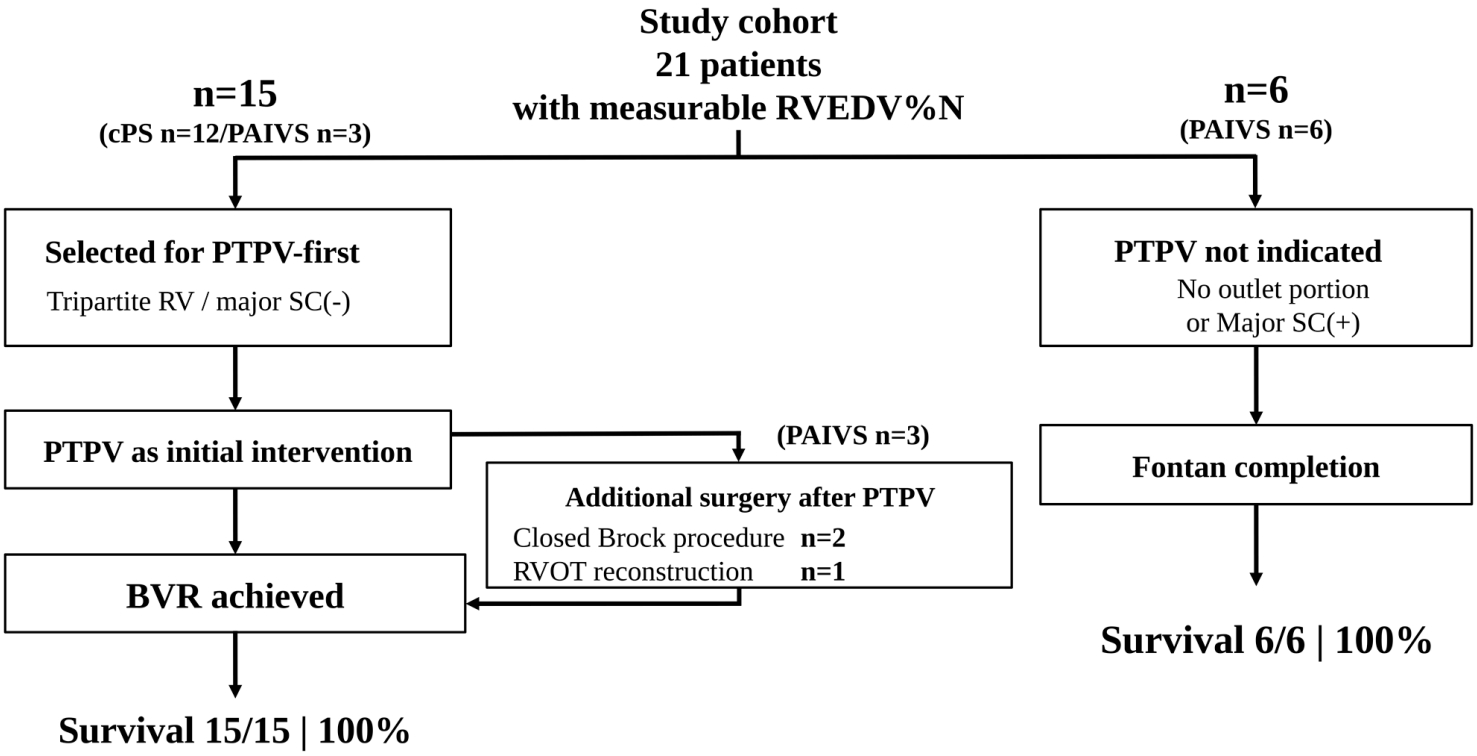


Figure 2

Study cohort, initial pathway selection, and final circulation. The outcome percentages refer to the 21 patients with measurable RVEDV. The clinical courses of the three excluded patients are described in the Results. All three PTPV-treated PAIVS patients required subsequent surgical RV decompression before achieving BVR. BVR, biventricular repair; cPS, critical pulmonary stenosis; PAIVS, pulmonary atresia with intact ventricular septum; PTPV, percutaneous transcatheter pulmonary valvuloplasty; RV, right ventricle; RVEDV, right ventricular end-diastolic volume; RVOT, right ventricular outflow tract; SC, sinusoidal communication

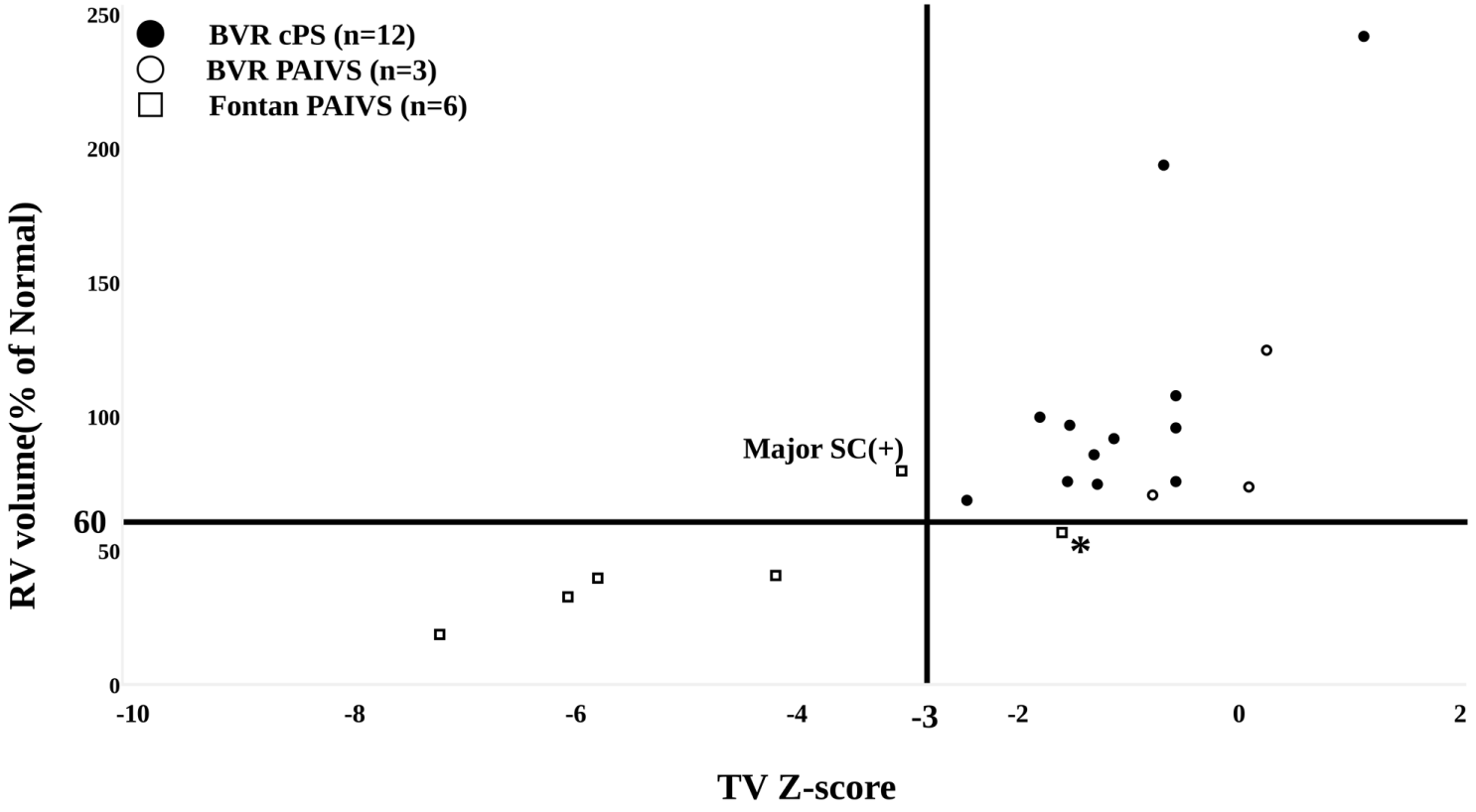


Figure 3

Post hoc distribution of RVEDV and TV Z-score according to diagnosis and final circulation. Filled circles indicate patients with cPS and BVR, open circles patients with PAIVS and BVR, and open squares patients with PAIVS and Fontan circulation. The approximate region of patients with major SC is labeled. The asterisk identified the Fontan patient who later developed protein-losing enteropathy. Dashed lines at RVEDV 60% of predicted normal and TV Z-score −3 are descriptive post hoc reference lines and not validated treatment cutoffs. BVR, biventricular repair; cPS, critical pulmonary stenosis; PAIVS, pulmonary atresia with intact ventricular septum; RVEDV, right ventricular end-diastolic volume; SC, sinusoidal communication; TV, tricuspid valve